PSYCHOSOCIAL INFLUENCES ON OUTCOME AFTER PAEDIATRIC CARDIAC SURGERY, INCLUDING TRANSPLANTATION

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To my children: Leah and Anna

ABSTRACT

This thesis is based on a study that explored the impact of individual and family factors on medical outcome and psychological adjustment following heart and heart-lung transplantation. Thirty eight children referred for heart transplantation (age range: birth-17 yrs, mean: 8 yrs, 2 mths), sixty eight for heart-lung transplantation (age range: 9 mths-17 yrs, mean: 11 yrs, 1 mths) and fifty nine children requiring conventional cardiac surgery (age range: 2 yrs-17 yrs 9 mth, mean:8 yrs, 9 mths) were included in the study. Parents and children, aged 5 or older, underwent separate standardized interviews; and standardized questionnaires were completed by parents, schoolteachers and children, aged 7yrs 6 mths or older. Assessments were made when the children were put on the waiting list and one year post-operatively.

The groups did not differ initially with regard to child's mental status and psychosocial functioning or with regard to marital and family adjustment or attitudes to other family members (parent-patient and partner-partner). Overall, 24% of the children had a psychiatric diagnosis and 60% had impaired functioning; over 40% of the parents were psychiatrically distressed and half of the families showed adjustment difficulties. Post-operatively, there was a reduction in the proportion of children with psychiatric disturbance (p=.0005) and the children's psychosocial functioning was enhanced (p=.001). Parental distress lessened (p≤ .04) but there was deterioration in the marital relationship (p≤ .02) and less warmth towards the partner (p≤ .04). The overall levels of family adjustment were similar pre-and post-operatively.

Psychological measures were not associated with survival, but were associated with physical health pre and post-operatively. Heart-lung transplantation (p=.01), living in a single or reconstituted family (p=.05), critical comments towards partner (p≤ .05) and family maladjustment (p=.003) were associated with poor adherence to medical treatment which was found in a third of the transplant recipients.

The study has important clinical implications and highlights the need for effective psychosocial interventions in children undergoing heart and heart-lung transplantation.

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INTRODUCTION AND OUTLINE OF THE THESIS

This thesis focuses on children who required a heart or heart lung transplant and their families. The conditions for which these possible treatment options might be considered are (i) congenital or acquired heart disease, (ii) pulmonary-vascular defect, or (iii) parenchymal lung disease, the most common being cystic fibrosis. Once acquired, these diseases progress and become chronic and life long. For comparison purposes two groups of children with congenital heart disease (CHD) who were in need of high or low risk conventional cardiac surgery and their families were also studied.

Despite the increase in the number of children with cardio-respiratory disorders who receive heart or heart-lung transplantation, little is known about the adaptation process in these children and their families. There is a lack of documented research about the psychological implications of such procedures or about the effects of transplantation on the children's physical and psychological well being. The methods used in this study have been derived from family systems theory and the main aims were:

i) To explore the way in which the family organises itself at the time its members are informed about the child's need for transplantation, and

ii) To explore whether the way in which the family is organised has a long-term effect on the patient's medical outcome.

The terms disease, defect and illness in this manuscript refer to an ongoing problem of the physical functioning of the body; the term disability refers to what the children are prevented from doing as a result of disease or injury (Davis 1993). The terms 'conventional cardiac surgery' and 'non-transplant cardiac surgery' have been used interchangeably.

The Expressed Emotion (EE) terminology has been used throughout this manuscript. Some of these terms (e.g. overinvolved parents, overprotective parents etc.), may sound pejorative to the reader not familiar with the EE scales. The author questions the appropriateness of using such terms when describing the attitudes of parents whose children are not only disabled but facing life-threatening conditions. Nevertheless, the author felt that it would be inaccurate to use a different terminology at this stage.

This thesis contains 6 chapters and a section of appendices. Chapter 1 gives an overview of the clinical picture presented by children who are referred for heart (HTx) or heart-lung (HLTx) transplantation and by children who are in receipt of conventional cardiac surgery. The demands of the illness on the children and their families are also described. Chapter 2 addresses a number of psychosocial issues in the literature of paediatric transplantation. At the end of the chapter there are a series of tables summarising methods of the studies discussed. The first part of Chapter 3 illustrates the theoretical background and the context of this thesis. The second part describes the design, measures and the statistical methods. The results are conveyed in Chapter 4. This chapter comprises a section on the reliability of the measures used, the description of the sample, and a quantitative analysis of the data. Chapter 5 explores further information gathered from the family interviews. The results, the critical appraisal of this research and its theoretical, clinical and ethical implications as well as some suggestions for future research are presented in Chapter 6. The interviews used are presented in appendices 1 and 2; Appendix 3 contains information about the expressed emotion ratings and, finally, two case studies are given in appendix 4.

CHAPTER 1

CARDIAC SURGERY INCLUDING HEART AND HEART-LUNG TRANSPLANTATION: AN OVERVIEW

Most heart disease in childhood is of a congenital nature; this is the commonest single group of congenital abnormalities accounting for approximately thirty percent of the total. The incidence is approximately 8/1000 live births (Jordan and Scott 1989). The development of a congenital defect usually requires genetic susceptibility triggered by an environmental hazard occurring between the second and ninth week of pregnancy during organogenesis. However, a minority of cases seems to be inherited without environmental insult.

The most serious outcome of congenital heart defect in childhood is premature death. If these children were left untreated 85% would die during their first year of life or during infancy (Somerville 1990). Hoffman (1987) followed a group of cardiac patients who were not treated and found that by 15 years of age, 26-48% of all these children had died, as compared with 5% for the normal population.

Not all children with heart disease undergo cardiac surgery. Some minor conditions do not necessitate any sort of treatment, whereas for some complex conditions surgery is not possible. However, approximately 60% of all children with heart disease will require some form of surgery during their childhood (Jordan and Scott 1989). These authors suggest that "the earlier the defect is corrected the quicker will heart size and function return to normal". However, this has to be weighed against the rather higher mortality of open-heart surgery carried out under the age of a year; and the fact that any prosthetic conduits or valves will not grow as the child grows. Subsequently, many children will have to undergo two or more cardiac operations during their childhood. Due to the emphasis placed on palliative surgery in infancy followed by corrective surgery later on, the mortality rate in this population has significantly decreased (MaCartney et al 1980).

Although most congenital heart lesions are potentially correctable, between ten and twenty percent of patients have lesions for which there is no adequate "corrective" procedure (Addonizio et al 1987). These children may be candidates for heart transplantation. Heart transplantation is also an option for patients with congenital cardiac disease with end-stage

myocardial dysfunction, which sometimes occurs after unsuccessful palliative surgery (Addonizio et al 1987; Taggart and Dark 1991). Other suitable candidates for heart transplantation are previously healthy born children who develop myocardial disorders (cardiomyopathy) at some point during their childhood. Bacterial and viral infections may have a role to play in the development of the illness by weakening the heart muscle. The incidence of these conditions is difficult to estimate as they are not the same in every country.

Children with a congenital heart disease and associated pulmonary vascular disease, a primary vascular disease, end-stage lung disease or cystic fibrosis may be candidates for heart-lung transplantation (Smyth et al 1989, Whitehead et al 1991, Bolman 1994; Bolman et al 1994).

Conventional Cardiac Surgery: Treatment and Convalescence

In the case of a mild defect, the only management required may be an annual physical examination, an electrocardiogram and x-rays of the chest. In more severe cases the children may require medication, make frequent visits to the cardiac clinic, and undergo complex in-patient tests (Hoffman 1987).

While awaiting cardiac surgery most of these children can lead ordinary lives and attend ordinary schools. Usually, parents are advised that there is no need to restrain or limit their children's activities. Children are admitted to hospital 24-48 hours prior to surgery and during this time they undergo a series of routine examinations including blood tests, x-rays, and sometimes a cardiac catheter, etc. The children and the parents are also seen by the play-leader and nursing staff, who prepare them for the operation.

Post-operatively, many children are only in intensive care for 24 hours and then return to the main ward, requiring little specific post-operative treatment. Antibiotics are given for 48 hours to cover the perioperative risk of infective endocarditis. Children undergoing complex operations may require a more prolonged period of intensive post-operative care including assisted ventilation and the use of inotropic agents such as Dopamine. These children may also need to take other drugs (e.g. diuretics and anti-arrhythmic) for several weeks post-operatively, and may be on antibiotics for life (Hoffman 1987; Somerville 1990; Jordan and Scott 1989).

The total post-operative convalescence period varies according to the complexity of the operation (Jordan and Scott 1989). This may range from a brief 4-7 day stay in hospital, followed by 4 weeks off school for simple operations, to a week in intensive care, followed by several weeks in hospital and a more prolonged period off school for children undergoing complex operations. Those children who undergo complex operations need to be carefully followed by the cardiac team. In most cases this is indefinite care, as the heart condition is not static and changes take place throughout the patient's life. Patients who have palliative operations early in life will deteriorate requiring other operation(s) as they grow. In some cases, if conventional surgery is unsuccessful or not possible, the children may be suitable for transplantation (Hoffman 1987; Somerville 1989, 1990; Jordan and Scott 1989).

Transplantation: Treatment and Convalescence

The treatment of children who are candidates for transplantation varies according to the diagnosis and ranges from no treatment at all to very demanding treatment. In general, the treatment for children suffering from diseases other than cystic fibrosis involves regular and frequent hospital visits, and in some cases drug therapy. The treatment of children with cystic fibrosis involves antibiotics, bronchodilators, oxygen, steroids and vitamin therapy; intensive physiotherapy two to four times daily and breathing exercises, plus a special diet. In addition, but depending on the severity of the illness, the children with cystic fibrosis have to attend the clinic at regular intervals, from once a week to once every three months (Whitehead et al 1991; Madden et al 1993; Whitehead 1993).

Each transplant centre has its own protocol. At Great Ormond Street Hospital for children (GOSH), candidates for transplantation are admitted for a period of 4-5 days, during which time an intensive medical and psychological assessment takes place (Whitehead et al 1991). Medical tests include: x-rays blood tests, assessment of exercise tolerance, respiratory function at rest and on exercise and, for some children, a cardiac catheter.

The psychological evaluation involves a full evaluation of the patient and the family by a child psychiatrist, a clinical nurse specialist sister and a social worker. Once the evaluation process is completed, the transplant team meets to discuss the findings and to make recommendations. Candidates for transplantation are either considered medically unsuitable or accepted to a "provisional" or an "active" waiting list, depending of illness severity and/or expectancy.

Children considered suitable for transplantation are severely disabled or a life expectancy of less than two years, are placed on the active waiting list. Due to donor shortage, between 20% - 40% of children die while awaiting transplantation (Addonizio et al 1987; Steinberger et al 1993). Children considered suitable for transplantation but who are not severely symptomatic and whose life expectancy is greater than 24 months are placed on the provisional waiting list.

Many months may pass before suitable donor organ(s) become available. Candidates for heart transplantation wait an average of 6 months, while candidates for heart-lung transplantation wait an average of 12 months. It is essential that children on the active list can be contacted at any time in case suitable organ(s) become available; accordingly, their parents carry an air call bleep 24 hours a day.

On the day of the transplant only a limited amount of time is available. The potential recipient is usually called to start preparation before all the tests on the donor organ are completed. Occasionally, the donor organ(s) is/are found to be unsuitable and the children are discharged from hospital with great disappointment. When a correct match of donor organ/s and patient is made, the child is taken to the operating theatre where he/she may wait for several hours before surgery starts. Post-operatively, the patient will be moved from intensive care to a special cubicle where he/she will stay for a possible 4 to 6 weeks. During that time the children needs to exercise regularly (cycling, treadmills, and walking) in order to get the new heart or heart and lung working.

After transplantation, the children must have treatment for the rest of their lives to prevent their bodies from rejecting the new organs (Whitehead et al 1995). The main drugs used for this purpose are immunosuppressants, including Cyclosporine A, Prednisolone, Azathioprine and steroids. Serial endomyocardial or transbronchial biopsies (for heart/ heart-lung transplantation, respectively) are also performed on a regular basis. During the first 12 months post-transplantation, these children have an average of 18 routine admissions to Great Ormond Street Hospital. In addition, they have to attend regular appointments at their local hospital.

Possible signs of rejection or infection in children who have received a heart-lung transplant can be detected by a simple lung function test which the patient must perform twice daily at

home using a spirometer. No such test exists to detect rejection in heart transplant patients and thus rejection is more difficult to diagnose. A possible episode of rejection is confirmed or excluded by means of a biopsy, which is carried out under local anaesthetic. Any episode of rejection or infection requires rapid diagnosis and treatment (Whitehead 1993; Whitehead and De Leval 1994; Whitehead et al 1995).

The Effect of the Illness on the Children

The effect that heart disease has upon the children who are in need of conventional cardiac surgery varies depending upon the type of defect (Jordan and Scott 1989). Defects range from small and trivial to large and fatal. The children's complexion ranges from normal pinkish to bluish. Some babies and small children may have only minor heart problems and are completely asymptomatic. However, children with more complex conditions have symptoms that may include breathlessness, particularly with movement or exertion. Babies may even become breathless when feeding, which may lead to failure to thrive and slower growth. Thus it can be seen that children with minor heart defects may not have physical disability at all, while children with moderate or complex defects have minor or major physical disabilities.

There exists a spectrum of disability in patients receiving transplantation (Hutchings and Monett 1989; Whitehead 1993). Breathlessness, tiredness, major physical disability, including periods of invalidity and long periods of confinement at home and in hospital is the picture presented by those children awaiting transplantation on the 'active list'. Subsequently, the children's school and social life is severely impaired and these children become isolated from their peer group and school friends, which may cause them to become 'loners'. Those patients on a provisional waiting list, and thus less severely affected by their disease, may experience a more normal mobility (Whitehead 1993).

The Effect of the Illness on the Family

The demands of the illness and treatment upon the parents and siblings vary according to the children's symptomatology, disability and treatment (Glaser and Bentovim 1987; Bentovim 1980; Gottesfeld 1979). Generally, parents will accommodate first to the children having a disease and requiring treatment. Later on, they have to accept the need for cardiac surgery or transplantation. Then, they have to come to terms with the "wait" for an operation, even though the children's life may be at risk. At the time of surgery, the families not only have

to deal with the disruption of family life, separation and loss, but also with the anxiety associated with the risk of death during surgery and the uncertainty of the children's future.

On a day to day basis, annual hospital appointments serve as the only reminder of the illness for those parents whose children have minor defects and are asymptomatic. The impact of the illness is naturally greater upon those families whose children have severe and complex defects, and subsequently suffer greater disability. Parents of these children have to bear in mind the children's physical limitations whilst planning the families' daily activities. Time may also be lost from work and the parents' social life may be disrupted. The healthy siblings often lose parental attention (Glaser and Bentovim 1987).

The stresses for families whose children are awaiting transplantation are even greater (Hutchings and Monett 1989; Uzark and Crowley 1989; Whitehead and De Leval 1994). The initial referral may be characterised by high levels of anxiety for both the children and families. This may be related to the fear of being 'unacceptable' for transplantation, but also to the fear of the unknown (Whitehead and De Leval 1994).

Many believe that transplantation is simply a surgical procedure effecting a permanent cure, and are dismayed at the prospect of lifelong treatment. In addition, the realisation that the children's life expectancy is less than two years once accepted for transplantation may present emotional difficulties for the families (Whitehead and De Leval 1994).

Furthermore, due to the children's physical disability and the periods of confinement at home, these children may be completely dependent upon the parents and other family members, and are in need of constant attention and care. In the case of children with Cystic Fibrosis (CF), life expectancy is partly dependent on the ability of their families to follow through with a lifetime of daily therapy efforts on the children's behalf. The gradual deterioration in pulmonary function can be forestalled by chest physiotherapy administered several times a day by a parent. In some cases, parents spend up to six hours a day carrying out treatments. Failure of the patients and family to carry out this treatment is an important factor in progression of CF lung disease (Patterson et al 1990). Subsequently, parents have serious difficulties meeting the demands that work imposes on them resulting, in some cases, in having

to give up working life. In addition, due to the demands of the treatment and to the children's frequent hospitalisation, siblings may be emotionally neglected.

In summary, the waiting period for transplantation is very stressful for the children and their families. As the children continue to deteriorate, their families have to cope with doubts and uncertainties about whether it is right to put their children through a transplant, and to keep up the struggle and hope for life at the same time as having to come to terms with the possibility of their children's death. This stage has been referred to as "dancing with death" (Kuhn et al 1988; Kuhn et al 1990).

Whilst transplantation aims both to prolong the children's life and to improve the quality of life, it is not curative. Following transplantation, life remains stressful and full of events. The children and their families not only have to deal with the chronic anxieties and disruptions produced by frequent routine admissions for biopsies, and constant threat of complications, but also with the uncertainty of survival. These families have to cope with the ever-present threat of death and yet hold on to hope for the future.

CHAPTER 2

PSYCHOLOGICAL ASPECTS OF PAEDIATRIC TRANSPLANTATION

This chapter reviews the literature related to paediatric transplantation. The first section presents a general view of the surgical developments and discusses stressors specific to transplant surgery. The second section comprises a literature review on adaptation and outcome following paediatric renal, bone marrow, liver, heart and heart-lung transplantation. Finally, the last section of this chapter contains a review on adherence to treatment in children with life-threatening conditions.

1. Paediatric Transplantation: New developments and Stressors

Bone marrow, renal, heart and lung organ transplantation have become widely accepted procedures for several life-threatening conditions in childhood such as leukaemia, renal failure and severe heart and lung diseases. The number of children with biliary atresia and other advanced hepatic disorders who undergo liver transplantation is also increasing (Bradford 1991).

New treatments which are in an early experimental state are lobar-lung and partial-liver replacement from living family donors, for children with lung and liver diseases, and computer matching of living unrelated donors in bone-marrow transplantation (Starnes 1993, Goldman 1993, Hows et al 1986). In addition to the overall increase in the number of children who are undergoing transplantation, the age range has also widened with increasing number of infants and younger children now undergoing these procedures (Backer et al 1992; Beath et al 1993; Reynolds et al 1991).

There is now a significant body of descriptive literature and clinical papers addressing psychosocial aspects/care of adolescents and children undergoing transplantation (Bradford and Tomlinson 1990; Stuber 1993a, 1993b; Gold et al 1986; Shaben 1993; Andrykowski 1994). Similarities and differences between paediatric and adult transplantation have also been addressed (Stuber 1993b). The stresses that the

transplant itself imposes on the children and on their families are not universal, but are related to the following factors:

1) The nature of the illness from which the child is suffering, and procedures associated with the type of transplantation.

2) The child's mobility and psychological well being prior to the transplant and whether transplantation is the only life saving procedure available.

3) The length of time that the child can be kept artificially alive (e.g. ventilator, dialysis).

4) Whether there are difficulties obtaining the organ/s.

5) Whether there is a short or long waiting period until transplantation after acceptance to the transplant programme.

6) The type of donor: cadaveric, living related or living unrelated.

7) The degree of ambivalence about transplantation, in the patient and other family members.

8) The risk involved in the procedure for the patient and donor (if a living donor is used).

9) Whether there are alternative medical treatments prior to the transplant, and following unsuccessful transplantation.

10) Parents' ability to communicate, negotiate new roles and support each other during the child's admission for surgery.

11) The annual survival rate following transplantation and the way that parents and children process/adjust to such information.

12) The seriousness of complications and the child's physical and psychological well being after the transplant.

13) If a living related donor is used, the seriousness of the relative's complications and well being

14) Probably one of the most important factors is the patient's and family members' appraisal of the situation.

The weight of each one these factors vary across types of transplantation. Therefore it is an almost impossible task to compare the psychological impact of transplant surgery across paediatric organ transplantation. However, it might be possible to outline some similarities in the psychological processes (stages). In doing this, it is important to keep in mind that

a) not all of these factors apply to each individual family containing a child undergoing transplantation, and b) when specific factors apply, there is a great variation regarding the intensity. In addition, there are important physical and psychological differences across organ transplantation. A summary of the similarities and differences is presented in TABLES I and II (survival rates have been facilitated by U.K Transplant Service).

Stresses Prior to Transplantation

The period of waiting for transplantation causes significant psychological distress but there has been little evaluation of the difficulties faced by these children and their families at this time. Clinical data and early descriptive reports suggest that prior to transplantation, fear of death and uncertainty about the future, anxiety and depression in the patient leading to temporary withdrawal and/or demanding behaviour with increased dependency, patients' guilt related to being a financial and emotional burden on their family, patients' concerns about other patients in a similar situation, jealousy manifested by siblings, parent-child protection/rejection and dependency, fatigue and decreased social relations for the parents and the siblings are psychological reactions which are common to all transplantation procedures (Khan and Herndin 1971; Gardner et al 1977; Zitelli et al 1986; Korsch et al 1972; Bradford 1991).

One study showed a higher prevalence of behavioural problems in candidates for heart transplantation when compared to a matched group of healthy children (Wray et al 1992). A prospective study on families of children awaiting heart or heart-lung transplantation demonstrated that 25% of the children had an emotional disorder and over half showed impaired psychosocial functioning. Twenty per cent of the well siblings also manifested behavioural and emotional problems and more than 50% of the families appeared to have long-standing difficulties (Serrano-Ikkos et al 1997).

TABLE I : _PSYCHOLOGICAL PROCESSES COMMON TO ALL TYPES OF PAEDIATRIC ORGAN TRANSPLANTATION_

Pre-Transplant	Waiting for the Transplant	Hospitalisation	First four weeks after Tx	Returning Home
- Medical misinformation. - The child undergoes a full physical assessment: uncertainty and anxiety about the child being accepted in the Tx programme. Frustration while awaiting for the preliminary medical testing to be completed. - The following aspects might also be evaluated: a)the child and parents understanding of the procedure; b)their commitment to going through with the transplant; c)the amount of support available to them; d) history of adherence; e) the psychological state of the patient and psychopathology in the parents, and f) family functioning. - If using a living-related donor: there is an evaluation of the quality donor-patient relationship. - If the child is suitable for transplantation: further information and preparation for the procedure is given to the family,including meeting other transplanted children and their families. - Parents and child, if old enough, give legal written consent. - If the donor is a sibling, a specific legal evaluation or/ and a psychiatric evaluation of the donor may be required to ensure that the child has an advocate.	- Ambivalence about transplantation. - In some cases there is a decline in the child's health, increasing his dependency on the family and feelings of guilt about being a burden. - In some cases the child can be kept artificially alive using alternative treatments (eg. dialysis). - If there is no other treatment and the child is facing death: intense anxiety, family attempts to maintain hope plagued by fear and worry that the child may die. In addition, and due to the donor shortage,increased anxiety, depression,anger and competition among waiters and their families. - Fatigue and decreased social relations for the parents and siblings. - Disruption of family, school and parents'working life. Parents inability to control their own feelings of guilt and sympathy leading to rejection, inappropriate expectations and over-protectiveness. Siblings may feel resentful - If there is a living related donor: a) she/he may have ambivalent feelings about donation; b)parents may put pressure on siblings to donate (eg. bone-marrow Tx). Conflict if sibling expressed apprehension and resentment about participating in the procedure.	- Sense of relief,excitement and euphoria that the Tx is taken place, and an accentuated fear of death. - Disruption of family life. Siblings are usually left behind with neighbours or extended family. - Immediately following Tx: rapid fluctuation in the patient's medical condition leading to conflicts between fear and hope. Parents' experience fluctuations in mood from euphoria to depression. - Parents feel exhausted. - If there is a living related donor: increased anxiety about the member of the same family undergoing risky procedures.	- Process of identification and comparison with other children in similar circumstances. Survivors'guilt if any of the other patients die. - Anxiety associated with the 1st biopsy, rejection or complications. Conflict between fear and hope. - Changes in family structure: prolonged separation between the spouses leading to changes in the divisions of responsibilities within the family, forcing the parents to give up roles and to assume new ones. - Changes in spouses' self-image leading to disynchrony of coping between the parents, creating enormous tensions and distress between family members. - Establishment of a strong and intimate bond between the child and the parent who is in hospital, often excluding parent and siblings at home. Parent guilt about being devoted to the child in hospital to the detriment of siblings at home. - Grief for the loss of own organ/s. Fears and anxieties about the psychological effect of the new organ on the patient, especially if the donor is from opposite sex and different background. - If a living donor was used: intense feelings of guilt, anxiety and anger if either the patient or donor is not doing as well as expected.	- Struggle for acceptance of the new organ/s as part of body. If a family donor: changes in patient-donor relationship and feelings for each other. - Ambivalence about returning home and intense fear of rejection. Parents take responsibility for administration of immunosuppressive medication and other aspects of treatment and for detecting infection and rejection episodes. - The child and other family members have to adjust to changes in the child's physical appearance due to immunosuppressant therapy. - Renegotiation of roles between the parents and child's re-integration into family life: the family has to adjust to the change the patient's energy and behaviours, attempts for independence and organization of his/her social life. - Frequent follow-ups visits to hospital and local clinic. If Tx was unsuccessful, the family has to accept either that the child is back to a disabling life and artificial treatment or that the child's death is imminent. - Patient's, and donor's if applicable, re-adjustment to school and social life. - If living donor is unwell: he/she may undergo continuous medical procedures, this creating resentment and feeling of guilt.

TABLE II: MAIN   STRESSES   DISTINGUISHING   TYPES   OF   PAEDIATRIC   ORGAN   TRANSPLANTATION   (Tx)

Renal Transplantation	Bone-Marrow Transplantation	Liver Transplantation	Heart Transplantation	Lung Transplantation
* Some children may have a chromosomal disorder. Risk of a syndrome of progressive encephalopathy, growth etardation and mental delay.	* The child is facing a life-threatening condition.	* Progressive jaundice, risk of life-threatening bleeding, incapacitating hepatic encephalopathy, anorexia and growth failure despite aggressive nutritional support. Possible brain damage.	* Risk of cognitive impairment (cyanotic children). Life-threatening condition.	* Life-threatening condition. Children with CF may have other organs seriously damaged (pancreas, liver, kidney etc).
* Dialysis: to sustain life. Restrictive schedule, complicated and restrictive diet.			* Ventilator: to sustain life.	
* Donor: living related (parents) or unrelated and cadaveric.	* Donor: living related (siblings, parents)or unrelated.	* Donor: living related (parents) or cadaveric.	* Donor: cadaveric.	* Donor: cadaveric (UK) or living related (from parents; USA).
	* Intensive treatment prior to Tx: radiation, chemotherapy or both (nausea, vomiting and fatigue). Isolation in laminar air flow room: skin to skin contact not allowed.Bone-marrow suppression lasting 3 to 4 weeks (risk of infection,bleeding, anaemia).			
* Surgery: open. Replacement of 1 kidney.	* Surgery: it does not involved open surgery. It takes 2-4 weeks for the marrow stem cells of the donor to migrate and mature in marrow cavities of the patient.	* Surgery: open. Full or partial replacement of the liver, depending of donor used. Minimal immunosuppression.	* Surgery: open. Full replacement of the organ.	* Surgery: open. Full or partial replacement of the lung. Very risky procedure for living donors.
* If successful: discharge 7 days after the actual Tx. If unsuccessful: back to dialysis and possible re-transplantation.	* If successful: patient leave for home approximately 100 days after admission. If unsuccessful: re-transplantation may be possible.	* If successful: discharge 4 weeks after actual Tx. If unsuccessful:re-transplantation is unlikely. The child is facing death.	* If successful: discharge 4 weeks after actual Tx. If unsuccessful: The child is facing death.	* If successful: discharge 4-6 weeks after actual Tx. If unsuccessful: the child is facing death.
* Survival rate: 1 year Post-Tx= 75% 5 years Post-Tx= 62% Long-term outlook: very good.	* Survival rate: 1 & 5 years Post-Tx= Great variation depending of type of: disease, BMT, and donor used. Long-term outlook: great variation	* Survival rate: 1 year Post-Tx= 60% 5 years Post-Tx= 51% Long-term outlook: unknown.	* Survival rate: 1 year Post-Tx= 69% 5 years Post-Tx= 55% Long-term outlook: promising.	* Survival rate: 1 year Post-Tx= unknown 70% of recipients have ongoing rejection at 3 years post-Tx Long-term outlook: Poor.

Stresses after Transplantation

Identified psychological stressors requiring adaptation following successful transplantation are: a) the re-integration into the family of a child who had been chronically ill and who returns home with renewed mobility and vigor, and the return to school life; b) concerns about appearance, short stature and weight as a result of treatment; c) permissiveness or excessive restriction of the child by the parents and the re-organization of family roles; d) survivors' guilt after another patient dies; e) financial strains; f) uncertain future, fears of relapse, and fears and ruminations associated with thoughts about the experience of transplantation; g) emotional isolation, difficulties forming heterosexual relationships and depression as these young people move into late adolescence and young adulthood (Bernstein 1971; van der Wal et al 1988; Poznanski et al 1978;Patenaude and Rappeport 1982; Zamberlan 1992; Lesko 1994; Sormanti et al 1994; White 1994; Stuber and Nader 1995; Kanabar et al 1995).

It is to be expected that once a child is put on a transplant waiting list family roles will change temporarily or permanently, because transplantation adds a unique dimension to the stresses already imposed by a life-threatening condition. The psychological impact of transplantation on the family may be intensified if the donor is a living related person, such as a sibling or one of the parents. Even if the risk for the donor is statistically small, it is perceived as a threat by the family, affecting relationships within the family system (Wiley et al 1984; Hare et al 1989). Furthermore, ambivalence is common among donor and non-donor family members. Additional family conflicts requiring psychological intervention may rise when donors, such as siblings, express apprehension and resentment at having to participate in the procedure, and at being deprived of normal age-appropriate activities.

Furthermore, the donation may increase the emotional liaison between the donor and the recipient in such a way that it may constitute a real danger to the adequate future psychological performance of both individuals (Velasco de Parra 1982; Steele and Altholz 1987; Hare et al 1989). Following transplantation, the donor could become heavily invested emotionally in the recipient, and the recipient may experience guilt. Consequently, changes in family structure may have either a positive or negative effect on family functioning depending upon whether the

donor ranked highest in the family hierarchy and was already in a rigid alliance with the patient before the transplant (Velasco de Parra 1982). The donor-recipient relationship has been said to be especially traumatic for adolescents, jeopardizing their wishes for autonomy and independence (Zarinsky 1975; Velasco de Parra 1982; Stuber 1993b).

The integration of the new organ represents a fundamental way in which transplantation treatment constitutes a different stressor when compared to other treatments for life-threatening conditions. This is a complex process in transplant recipients regardless of the type of donor (cadaveric or living donors). Children, and adolescents in particular, have difficulties accepting that one has a part of someone else's body within one's own body, and that someone else had to die to give them this chance to live (Bernstein 1977; Stuber 1993a). The internalisation of the new organ is further complicated by what the organ represents (eg. heart as the symbol of emotions), and by the fantasies and attitudes attached to it (eg. sexual or gender attributes) (Riether 1990).

2. Outcome and Adaptation following Paediatric Transplantation

Cognitive, Intellectual and Motor Development

Few empirical investigations have assessed the effects of transplantation on the cognitive development and learning of paediatric organ transplant recipients. Rasbury et al (1983) assessed the cognitive development of 14 children with endstage renal disease, before and 1 month after the transplant. Their findings indicated that renal disease itself impairs cognition and learning, but improved problem-solving abilities were demonstrated post-transplant. However, in a later study the same authors reported that successful transplantation might not completely restore cognitive losses incurred by a chronic and debilitating disease (Fennel et al 1984). A summary of the studies discussed in this section is presented in TABLES III to VII (pages 36-51)

With regard to young patients awaiting liver transplants, Stewart et al (1987) found that infants and children with biliary atresia were subject to both mental and motor development delays. In a later paper, they demonstrated that although the cognitive functioning of 36% of children

improved to normal levels after transplantation, early onset of illness was associated with persistent delays in mental functioning (Stewart et al 1989). Zitelli et al (1988) found that despite an improvement in the child's physical health there were no changes in the child's cognitive abilities following liver transplantation.

Cranial radiation is an important component in the treatment regimen of paediatric bone marrow transplant recipients. The effect of such treatment on the psychological development of children has been a focus of attention. Pot-Mees (1989) found that the reading and arithmetic abilities of 26 children who survived 12 months post-bone marrow transplantation were lower than before the transplant. The child's age at the time of treatment has been associated with intellectual impairment. Whilst Smedler et al (1990) demonstrated a pronounced delay in motor development in children treated before their 3rd birthday, but not in older children, results of a meta-analysis study showed that greater decrements occur among children treated before five years of age compared with children receiving the same treatment after this age (Cousens et al 1988). However, these findings were not supported in a later study conducted by Kramer et al (1992), although examination of individual cases revealed a high degree of variability, with decrements in IQ of 10 points or more found in 7 children at one year follow-up.

Very little attention has been given to the cognitive development of children undergoing heart and heart-lung transplantation. Wray et al (1992, 1994) found that the intelligence quotient of children with heart and lung diseases who received transplantation was within the normal range, although transplanted children had lower scores on the developmental parameters than healthy controls (Wray et al 1994).

In summary, these studies suggest that children with renal and liver failure may be developmentally damaged prior to transplantation, because of the metabolic abnormalities associated with the disease, whilst children who are suitable for bone-marrow transplantation may be at risk of cognitive impairment as a result of the medical treatment, specifically cranial radiation. There is no evidence suggesting that the intellectual abilities of children undergoing heart or heart-lung transplantation are affected during the transplant process. Improvement in

cognitive functioning following transplantation may occur, but the characteristics which differentiate those who improve from those who do not improve/deteriorate remain unclear. Future studies on cognitive functioning should incorporate longer-term follow-ups, as evaluations up to 12 months post-transplant may not be sufficient to pick up deficits, especially in children receiving radiation.

Physical Status and Psychological Adaptation

Although the literature on adults has investigated psychological adjustment before and after transplantation, only a small number of prospective research studies address this topic in paediatric recipients. A summary of the studies discussed in this section is presented in TABLES III to VII (pages 36-51).

During the 90's, transplantation raised hopes for an increased survival rate for children with various forms of fatal diseases. In general, survivors of successful transplantation experience a much improved physical status compared to before transplantation (Bernstein 1977; Reynolds et al 1991; Colonna et al 1988; Zitelli et al 1988; Pot-Mees 1989; Samuelson et al 1984; Zamberlan 1992; Sharples et al 1994; Whitehead et al 1994; Petterson et al 1990; Backer et al 1992; Radley-Smith and Yacoub 1992). Regarding paediatric heart transplantation, there are reports suggesting that survivors returned to age-appropriate activities but these conclusions are not supported by solid data (Pahl et al 1988; Parness and Nadas 1988; Bailey et al 1992; Backer et al 1992; Dennis et al 1993; Smyth et al 1990a; Lawrence and Fricker 1987).

Although school attendance improves following transplantation surgery, recipients do have great difficulties re-adjusting to normal life and age-appropriate activities.. In an early report, Bernstein (1977) found that the school performance of renal transplant recipients was worse than the performance of other children with chronic conditions. Recently, Fukunishi and Kudo (1995) have reported that renal recipients had worse school adjustment than healthy peers, but better school adjustment than a control group of children receiving ambulatory dialysis. Regarding liver transplantation, Colonna et al (1988) found that 32 liver recipients had returned to school but no measures of school performance or adjustment was used. However, Zamberlan (1992) found delays in psychosocial functioning at school in this group of

recipients. Zitelli et al (1988) reported that 77% of 57 children who received liver transplantation were either at an age-appropriate grade or only one-year behind. In a longitudinal study of children who received a bone-marrow transplantation, Pot-Mees (1989) found that at 12 months post-surgery the number of children with a behaviour disorder in the school situation had dropped from 29.4% to 23.5% but this difference was not statistically significant. Van der Wal et al (1988) pointed out that bone marrow recipients, due to prolonged school absence and to the neurological and physical effects of the radiation procedure, may be specially vulnerable to school phobia and lower levels of self-esteem.

Overall, survivors' behaviours appear to improve following successful transplantation. Nevertheless, whilst some authors have found that behaviour scores are within the normal range (Korsch et al 1972; Korsch et al 1973), parents still do not perceive their children's behaviour as normal, the siblings are still resentful at 24 to 60 months post-transplantation (Zitelli et al 1988), and have emotional or behavioural difficulties (Stewart et al 1993). In addition, higher levels of depression and anxiety, behavioural problems, social difficulties and symptoms of post-traumatic stress disorder (PTSD) have been reported at 12 months post-transplantation (Khan and Herndin 1971; Gardner et al 1977; Korsch et al 1973; Pot-Mees 1989; Reynolds et al 1991; Stuber et al 1991; Windsorova et al 1991; Fukunishi and Kudo 1995). Regarding heart and heart-lung transplantation, Wray et al (1992) found that behavioural problems lessened following transplantation. More recently Wray et al (1994) reported that 24% of heart or heart-lung recipients had significant behavioural problems, but differences were not found between the recipients and 2 comparison groups, children who received conventional heart surgery and healthy children. Uzark et al (1992) also reported that heart transplant recipients were not different from their peers on self-concept and anxiety measures, but they showed significantly less social competence and more behaviour problems than children drawn from the general population.

The Family

Relatively, little attention has been given to the effect of transplantation on family relationships and on parental well being. Reynolds et al (1991) reported a lower percentage of intact families in parents of renal recipients when compared to parents of children receiving

dialysis or parents of healthy children. Uzark and Crowley (1989) described the effect of transplantation on the parents of 10 heart recipients. Uncertainty about the child's future, and extra demands on time with diminished physical energy, were the most serious concerns of these parents. The authors concluded that "parents experience chronic stress and tend to focus their attention, and direct their energy toward the child with a heart transplant. Role strain can occur when parents have difficulty fulfilling obligations of parents, spouse, and various other roles". Later on, Dermatis and Lesko (1990) evaluated the prevalence of psychological symptoms in parents of children assessed for bone-marrow transplantation. They found that 47% of the fathers and 60% of the mothers exhibited high levels of psychological distress. Reynolds et al (1991) reported that 60% of the mothers whose children were receiving haemodialysis, and 26% of the mothers whose children had undergone renal transplantation had mental health problems.

It is of interest that the parents' psychological well-being has been found to be affected by marital status, emotional coping styles and the quality of physician-parent communication, and to be unrelated to the characteristics of the child's disease and treatment (Dermatis and Lesko 1990). Early and more recent reports suggest that rehabilitation of the family to pre-illness levels of functioning is achieved by 12 months after transplantation (Korsch et al 1973; Bernstein 1977; Zamberlan 1992). However, Khan and Herndin (1971), found high levels of parental overprotection and moderate levels of parental rejection between 2 and 5 years after renal transplantation. The authors attributed these parental attitudes to their unrealistic expectations of transplantation. Using a standardized scale, Fukunishi and Kudo (1995) found that families of renal recipients scored less in the independence and achievement orientation scales than the families of healthy controls.

Given the impact that transplantation has on all family members, there is a surprising shortage of longitudinal studies evaluating changes in specific family domains after transplantation. Lee et al (1994) observed parent-child interaction in 6 families, prior and up to 24 months after bone marrow transplantation. They found that parental responsiveness remained fairly stable over time, despite new stressors and concurrent changes in the child's physical condition.

Factors associated with adjustment and medical outcome

Because of the high mortality rate prior to transplantation, transplant teams are constantly dealing with helplessness and grief. Due to the shortage of organs, one of the most difficult decisions faced by the teams is the selection of the candidate that will get maximum benefit and longevity. The selection of the candidate is conducted in the knowledge that the decision will mean probably death for one or more of the patients on the waiting list. Whether psychological factors have an effect on medical course or survival are important aspects, which need careful and detailed examination. Currently, many transplant centres around the World use psychosocial contraindications for transplantation[1]. Subsequently, patients with family instability, inadequate support, psychiatric illness or maladaptive illness behaviour, or unsatisfactory adherence to treatment are excluded from their programmes (Freeman et al 1992). This raises important ethical dilemmas.

Few studies have examined associations between demographic, and environmental factors, and children's psychosocial adjustment after transplantation. The following factors have been associated with behavioural or social problems at home or at school: severity of illness and complications; the children's age and developmental stage at the time of transplantation; dissatisfaction with physical appearance; friction between the parents; parental adjustment, coping style and family's expressiveness and cohesion; greater family stress and diminished family resources for managing stress ((Bernstein 1971; Poznanski et al 1978; Reynolds et al 1991; Stuber 1993b; Bernstein 1977; Korsch et al 1978; Pot-Mees 1989; Fukunishi and Kudo 1995; Uzark et al 1992).

Little attempt has been made to relate psychosocial factors to medical outcome, although poor medical outcome may be due to poor adherence with medical treatment (Whitehead et al 1994). Studies examining psychosocial predictors of poor adherence to treatment are described in the next section of this chapter. In an early report on bone marrow transplantation, Pfefferbaum et al (1977) examined the effect of parent-child relationships on the child's survival following transplantation. The physicians, nursing staff and social worker made retrospective subjective

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[1] This is not the case at Great Ormond Street Hospital. The selection criteria used by the team are as follows: 1:donor-recipient matching; 2: disease severity; and 3: length of time on waiting list.

ratings. Correlation between survival and psychosocial factors were not demonstrated. The authors postulated that a larger sample and a more detailed study using other measures might demonstrate a direct or indirect effect on mortality rates. However, and also using a retrospective design, McConville et al (1990) demonstrated that children who die unexpectedly or had unexpectedly high complication rates had shown more psychological distress patterns previously than those children surviving up to 12 months after bone-marrow transplantation. The child's functional impairment, family dysfunction and paternal psychopathology were associated with poor medical outcome. Nevertheless, a recent prospective study on renal transplant recipients failed to find significant associations between somatic indicators and psychosocial factors (Fukunishi and Kudo 1995).

Therefore, it appears that medical outcome is dependent to some extent on the psychological context in which ill health and treatment occur. During the process of transplantation an unusually close relationship between family and staff develops (Patenaude et al 1979). Clinical information suggests (Parsons et al 1996) that "the psychosocial context of ill health is not solely dictated by family response, but is also affected by interactions with the health care professionals through the process of mirroring. This process of mirroring can be described as the reflecting of behaviour, emotions or beliefs between at least 2 individuals, or groups, and forms a part of wider processes of reflexivity" (Parsons et al 1996, page 18). In that context, a number of maladaptive patterns may occur between the family and the teams of health professionals involved, with potentially damaging effects. Despite the clinical implications, the professionals rarely acknowledge the mirroring processes involved.

Despite the differences between various types of transplantation, the data suggest that there are common psychological reactions among the children and their families. However, one should be cautious in interpreting these findings due to the following methodological problems:
a) Most studies are cross-sectional.
b) The length of time since transplantation and severity of illness post-transplantation are not always controlled for. The follow-up assessments are carried out either too soon after the transplant or at any time between 3 months to 4 years post-transplantation.
c) Within the same study, retrospective and prospective data are presented together .

d) Most studies use only self-report measures.

e) The instruments used are not always standardized or validated.

f) Samples are often too small, and in order to increase the number of participants, patients are gathered from several transplant centers that might have used different techniques or procedures. In addition, patients who received just one or more transplants are pooled together.

g) Pre-transplant data is often missing.

The children's physical conditions, risk of complications and survival rate change over time since transplantation (Dennis et al 1993; Sharples et al 1994; Backer et al 1992; Whitehead et al 1994). Therefore, fluctuations in the children and family's adjustment and in family organization are expected in order to re-adjust to the continuous challenges in the child's physical condition. In the studies reviewed, families undergoing a crisis and families maintaining a transitory state of balance are mixed together, obscuring the validity of the results. The methodological limitations of the studies reviewed emphasized the need for prospective research on the impact that transplantation has on the children and other family members.

In summary, current understanding of the psychiatric and psychological aspects of paediatric transplantation is limited. Available studies indicate that despite an improvement in the child's physical health after transplantation, adjustment is difficult for the children and their families. The emotional cost of transplantation to a particular family depends upon the medical course, the individual and familial balance of strengths and weaknesses at the time of crises, and the extent of the changes in family organisation necessitated by the transplant. More research is required to evaluate how previous experience of illness and possible death of a sibling, disruption of family work and school life at the time of transplantation, unstable marital relationships and lack of support from extended family and community influence both the children's physical and psychological well-being, and the way the family copes with the process of specific types of transplantation. Psychological aspects of unrelated or living-related donors, and associations between the recipient's physical and psychological status and the donors' psychosocial reactions remain to be investigated.

TABLE III: RENAL TRANSPLANTATION (RTx)

AUTHOR/S	AIM OF STUDY	TIMING OF ASSESSMENT/S	GROUP(S)	INSTRUMENTS	FINDINGS AND CONCLUSION
Bernstein 1971	To examine the emotional and adaptational aspects of the transplanted child immediately following surgery and over a longer period of survival time	Retrospective Assessment - Prior to Rtx Prospective Assessment: a) 2 to 6 weeks post-RTx b) 4 to 11 mths post-RTx c) 1 to 6.8 yrs post-RTx	Group 1: 36 recipients Age Range: 6-18 yrs Mean: Unspecified	- Unspecified Psychiatric evaluations - Unspecified Questionnaires	- At discharge, areas of concern were the child's reintegration into the family, and the return to school. - 4 to 11 months post-RTx, the child was in a dependency role, with ensuing convert hostility. Patient-donor revealed ambivalence about his kidney loss. - 1 to 7 years post-RTx, the Tx still remained a pivotal area of many life situations, although survivors appeared to adapt well if there were few complication factors. Four of 32 patients (12%) had emotional reactions.
Kahn et al 1971	Identification of factors that contributed to the social and emotional adaptation of the children and their families	Crossectional Assessment: - Recipients: at any time between 2-5 yrs post- RTx; - Haemodialysis patients: at any time from 6 mths to several yrs since treatment started	Group 1: 7 recipients Age range: 6 - 19 Mean age : 12.8 Group 2: 7 children receiving haemodialysis 6 mths or longer Age range: 7 - 16 Mean age : 11.1	- Unstructured Interviews - Wechsler Intelligence Scale, WISC - Draw a Person Test	- 5 recipients (71.4%) and all children who were receiving hemodyalisis were depressed; and most children had social difficulties. All children's intelligence within normal range. - Overall: 9 families (64.2%) were overprotective, 2 families (14.2%) showed rejection of child, and only 3 families (21%) were supportive. Child and family had unrealistic expectations about transplantation.
Korsh et al 1973	To evaluate the impact of transplantation on the child and family	Crossectional Assessment: - At any time between 1 and 5 yrs post-RTx	Group 1: 35 recipients Age range: 1.5-20 yrs Mean age: unspecified Group 2: 20 children with CF Group 3: 200 well children and 200 chronically ill children	- The Rochester Parent Interview - Unstandardised Child Interview - California Personality Test -Saranson's General Anxiety Scale - Piers-Harris Self-Esteem Scale -Draw a Person Test	- Total personality scores were within normal range but the social adjustment scores were lower than normal in renal recipients. This group also scored higher in anxiety and lower in self-esteem than the controls. Personality disturbance was not greater in the renal recipients than in the controls and family equilibrium was restored 12 months after Rtx .

AUTHOR/S	AIM OF STUDY	TIMING OF ASSESSMENT/S	GROUP(S)	INSTRUMENTS	FINDINGS AND CONCLUSION
Bernstein 1977	To examine psychological adaptation following transplantation	Longitudinal Assessment: a) Prior to RTx b) 6 weeks post- RTx, and c) 12 mths post- RTx	Group 1: 100 recipients Age range: 6 wks to 18 yrs	- Psychiatric Evaluation - Draw a Person Test - Draw your Family Test - Sentence Completion Test - Wechsler Intelligence Scale, WISC	- At 12 mths post-RTx, 10% of children manifested behavioural & emotional problems requiring treatment, 12 to 18 years old were more at risk. -Adolescents from disrupted families had more difficulty adjusting. -Dissatisfaction with physical appearance was associated with lower sociability and self-esteem. -Recipients performance at school was worse than other children with chronic conditions. -The patient's level of adjustment was affected by the developmental stage.
Korsch et al 1978	To examine psychosocial predictors of poor adherence	Crossectional Assessment: -at 12 mths post-RTx Adherence was measured over a 10 yr period	Group 1: 80 recipients Age range: unspecified Mean age : unspecified	- Modified Rochester semi-structured Parent Interview - California Personality Test - Piers-Harris Self-Esteem Scale	- Total personality scores were no different from those of normal children, but there were persistent personality problems relating to low self-esteem and poor social adjustment. - 14 of 80 children (17.5%) interrupted medication at least once over a year period. -Family structure, gender, dysfunctional communication, friction between parents and low income following Tx were associated with unsatisfactory adherence.
Poznanski et al 1978	To examine quality of life and after transplantation	Crossectional Assessment: -2 to 10 yrs post-RTx	Group 1: 9 recipients with successful transplantation Group 2: 9 recipients with unsuccessful transplantation Age range: 9-16 (at Tx) Mean age : 12.3 (at Tx)	- Separated unstandardised interviews with patients and parents	- Patients in Group 1 had better school / job performance and better social life than patients in Group 2. - The depression rate in group 1 was of 22.2% and 87.5% in group 2. Negative self-image was associated with inability to relate to friends, interfering with early dating leading to social isolation.

AUTHOR/S	AIM OF STUDY	TIMING OF ASSESSMENT/S	GROUP(S)	INSTRUMENTS	FINDINGS AND CONCLUSION
Rasbury 1983	To examine the effects of chronic renal failure on cognitive functioning	Longitudinal Assessment: a) at onset b) 1 mths post-RTx	Group 1: 14 recipients Mean age: 11.2 yrs Group 2: 14 healthy children Mean age : 11 yrs	- Weschler Intelligence Scale, WISC - Peabody Individual Achievement Test - Halstead-Reitan Category test - Free Recall Memory Test	- Renal disease can impair cognition and learning, and successful transplantation results in improved problem solving behaviours. - Renal function and cognitive performance of patients significantly improved, indicating a relationship between cognitive & improved physiologic function.
Fennell 1984	To examine changes in Cognitive functioning following transplantation	Longitudinal Assessment: a) 1-3 weeks prior to haemodialysis, b) at 1 to 12 mths after RTx	Group 1: 20 candidates for RTx Age range: unspecified Mean age: 11.7 yrs Group 2: 18 children from public school Age range: unspecified Mean age: 12.6 yrs	- Weschler Intelligence Scale, WISC - Halstead-Reitan Categories Test - Free Recall Memory Test	- Base lines scores showed that candidates for RTx were not different in intelligence from the control group. - Tx recipients showed a significantly larger increase in performance IQ (non-verbal problem solving) than the control group. - Successful transplantation may not completely restore cognitive losses incurred by a chronic and debilitating disease.
Reynolds et al 1991	To examine changes in psychological adjustment following transplantation	Retrospective Assessment: a) prior to RTx Prospective Assessment: a) at any time between 8 mths to 6 yrs after RTx	Group 1: 29 recipients Age range: 1 mth to 17 yrs Mean age: 11.5 yrs Group 2: 22 children receiving haemodialysis Age range: 3-18 yrs Mean age: 13 yrs Group 3: 22 children with renal failure not receiving haemodialysis matched for age and sex with group 2 Group 4: 31 healthy children Age range: 2-18 yrs Mean age : 13 yrs	- A Questionnaire devised for this study measuring changes in physical and psychological health since Tx - Rutter's A and B Scales - Pre-school Behavioural Checklist - Parental perception of the Child's Behavioural and Emotional problems on a 3 point Scale - Birleson Depression Scale for Children - Lipsitt Self-Concept Scale for Children - General Health Questionnaire-28 (GHQ-28) for parents	- 75% of parents reported a pronounced improvement in child's physical health, whilst 25% reported a pronounced improvement in child's behaviours. - 37% of the parents reported a pronounced improvement in family life following RTx. - The prevalence of behavioural problems at home were twice as high in the Tx group (n=10, 38%) than in the control groups. - Mothers whose children were receiving haemodialysis had a higher incidence of mental health problems (60%) than mothers of recipients (26%). - The percentage of children with social adjustment difficulties was higher in the group of children receiving haemodialysis than in the Tx recipients group. No statistical differences between the groups were found in the depression or self-concept scales.

AUTHOR/S	AIM OF STUDY	TIMING OF ASSESSMENT/S	GROUP(S)	INSTRUMENTS	FINDINGS AND CONCLUSION
Foulkes et al 1993	To examine associations between psychosocial factors and adherence	Crossectional Assessment: - Post- RTx (range from Rtx to 31.3 mths)	Group 1: 32 recipients Age range: 6-21 yrs Mean age : 11.9 yrs	- Unstandardised child's perception of social support scale - Inventory of Socially Supportive Behaviours - Family Adaptability and Cohesion Evaluation Scale (FACES) - Family Inventory of Life Events and Changes (FILE)	- Informative and emotional support from fathers was found to be negatively related to compliance with azathioprine and cyclosporine . - The number of stresses experienced by the family was predictive of compliance with azathioprine. - Older children were more compliant than younger children with cyclosporine.

Fukunishi and Kudo 1995	To examine family environment and school adjustment in children with end-stage renal disease	Crossectional Assessment:
		 - Children with end-stage renal disease, receiving dialysis or transplanted, were evaluated over 3 months during routine monthly admissions
			Group 1: 26 children on continuous ambulatory peritoneal dialysis (CAPD)
			   Age range: 6-15 yrs
			   Mean age : 9.7 yrs
			 Group 2: 27 recipients
			   Age range: 6-14 yrs
			   Mean age : 9.4 yrs
			 Group 3: 27 healthy children
			   Age range: 6-14 yrs
			   Mean age : 7.4 yrs
				- Diagnostic Interviews for Children and Adolescents (DICA), DSMIII-R criteria
				 - Unstandardised parental interview to assess school maladjustment (school absenteeism and relationship with peers)
				 - Family Environment Scale (FES)
					- 88.4% of the children with CAPD, 55.5% of the recipients, and 3.7% of the healthy children, had a psychiatric disorder. Separation anxiety and adjustment disorders were, respectively, the most common diagnosis in children with CAPD and in transplant recipients.
					 - School absenteeism and problems in relationship with peers was significantly higher in CAPD children than in healthy children. Transplanted children also had a higher rate of problems with peers than healthy children.
					 - Regarding the family environment, the independence and achievement orientation scales were significantly lower in CAPD families than in healthy families. Scores on these two scales were also significantly lower in families of transplanted children than in healthy families.
					 - Significant negative correlations were found between school absenteeism, peers relationship and family's cohesion and expressiveness in both CAPD and transplanted children, but there were no associations between psychosocial (maladjustment, psychiatric disturbance or family environment) and physiological measures (blood urea and creatinine).

TABLE IV: LIVER TRANSPLANTATION (LTx)

AUTHOR/S	AIM OF STUDY	TIMING OF ASSESSMENT/S	GROUP(S)	INSTRUMENTS	FINDINGS AND CONCLUSIONS
Stewart et al 1987	To examine mental and motor development in infants and children with end-stage biliary atresia who were being evaluated for transplantation	Crossectional Assessment: - prior to LTx	Group 1: 41 candidates Age range: >30-61 mths Mean age (infants): 12.7 mths Mean age (children):42.8 mths	- Bayley Scales of Development - Stanford-Binet Intelligence Scale - Minnesota Child Development Inventory	- Growth, mental & motor development were delayed. - Infants development scores were related to growth but development scores in children tended to be related to measures of liver function rather to measures of growth.
Colonna et al 1988	To examine quality of life following transplantation	Retrospective Assessment: - 6 mths prior to LTx Prospective Assessment: - most recent 6 mths of survival (survival range: 6mths - 3 yrs)	Group 1: 32 recipients Age range: unspecified Mean age : 4.7 yrs	- Unstandardised questionnaires	- Accelerated weight and height gain in 87% and 60% of the children, respectively. - Improved activity tolerance, all children returning to school.
Zitelli et al 1988	To examine changes in life styles of children who had undergone LTx and who were observed for as many as 5 years	Crossectional Assessment: - 24-60 mths post-LTx Longitudinal Assessment: - A subgroup of 24 parents were interviewed before and after LTx	Group 1: 65recipients Age range: 3.6-21.3 yrs Mean age : Unspecified	- Bayley Scales of Development - Merrill-Palmer Mental Tests - Stanford-Binet Intelligence Scales - Wechsler for Children,WISC - Vineland Social Maturity Scale (for Parents) - Unstandardised questionnaires	As compared with pre-tx status, most children: - Had an improvement in physical health and gross motor functioning, and there were no changes in cognitive functioning. - were attending age-appropriate classes. - Behaviour improved but parents were still concerned about: separation-anxiety, aggression, enuresis and immaturity. - Siblings still resentful towards the patient.
Stewart et al 1989	To examine changes in mental and motor development, social function and growth at 12 mths post-LTx	Longitudinal Assessment: a) prior to LTx and b) at 12 mths post-LTx	Group 1: 29 recipients Age range: 3 mths to 15 yrs Mean age : 4.7 yrs	- Bayley Scales of Development - Stanford-Binet Intelligence Scale - Minnesota Child Development Inventory - Motor Age Quotient - Weschler Intelligence Scale ,WISC - Child Behaviour Checklist - Social Competence Scales	- Patients scoring less than 80 before LTx, gained an average of 8 points in intellectual and motor scores. - Onset of liver disease in the first year of life was associated with delayed development after LTx. - Older subjects improved in social competence. - Weight, head and arm circumference increased but there was no change in linear growth rate.

AUTHOR/S	AIM OF STUDY	TIMING OF ASSESSMENT/S	GROUP(S)	INSTRUMENTS	FINDINGS AND CONCLUSIONS
Windsorova et al 1991	To examine the adaptation of children who had received LTx, and to compare their performance with the performance of children with diabetes	Crossectional Assessment: - 1.2 to 5.5 yrs post-LTx	Group 1: 25 recipients Age range: 4-12.7 yrs Mean age : 7.9 yrs Group 2: 26 children with diabetes Age range: 4-12.4 yrs Mean age : 8.1 yrs	- Achenbach Child Behaviour Scale - Kovacs Depression Inventory - Children State-Trait Anxiety Scale - Piers-Harris Self-Concept Scale - Rorschach Inkblot Test	- On self-report measures, recipients were as well adjusted, as were children with diabetes or normative samples. - On projective measures, the liver transplant recipients had greater indication of depression, anxiety and lower self-concept than the normative samples.
Zamberlan 1992	To elicit information about the quality of life in children who had undergone LTx 3 to 6 years previously	Crossectional Assessment: - 3 to 6 yrs post-LTx	Group 1: 20 recipients Age range: 5.4-11.9 yrs Mean age : 7.8 yrs	- The Zamberlan Quality of Life Questionnaire for School-Age Recipients - The Pigem's Test (Three Wishes) - Piers-Harris Self-Concept Scale	- Psychosocial functioning at school and relationship with peers demonstrated delays in socialization, difficulties establishing peer relations, feelings of loneliness and vulnerability. - Family relationships were reported to be close and family functioning had returned to normal. - Children had all internalized the new liver as a part of them. - While physical functioning had improved, changes in physical appearance induced by post-Tx treatment had a negative impact on the child's psychological status. - All children denied worrying about rejection, and the further away the child was from the transplant date, the more optimistic the child was about survival. - Children were overall satisfied with their life and had positive thoughts about their future.

AUTHOR/S	AIM OF STUDY	TIMING OF ASSESSMENT/S	GROUP(S)	INSTRUMENTS	FINDINGS AND CONCLUSION
Stewart et al 1993	To examine psychological adaptation in siblings of children awaiting liver transplantation	Crossectional Assessment: - prior to LTx	Group 1: 64 siblings Age range: 4-16 yrs Mean age : 7.4 yrs	- Achenbach Child Behaviour Scale - Impact on Family Scale	- As a group, siblings of patients with end-stage liver disease were not maladjusted, socially or behaviourally. However, 23% of siblings presented behavioural or emotional difficulties. - Maternal stress appeared to relate to the presence of emotional and behavioural disturbance in siblings.
Goldman 1993	To examine the psychological adjustment of living related donors before and after transplantation	Longitudinal Assessment: a) at the time of medical investigation to evaluate the suitability of the donor b) at 12 mnths post-LTx	Group 1: 22 donors Age range: unspecified Age mean : unspecified	- Minnesota Multiphasic Personality Inventory (MMPI) - Symptom Checklist-90 (SCL-90) - Unstandardised Interview with the donor and spouse	- The decision to donate was generally made very quickly and further medical information had little impact. - Families seemed not to have any crisis or communication difficulties in reaching a decision. Nevertheless, the parental couple tended to make the decisions themselves, excluding or overriding any contrary opinions from other family members. - Prior to transplantation, 5 of 9 subjects who completed the MMPI scored abnormal in at least one of the scales; and 3 of 10 subject who completed the SCL-90 scored high on the anxiety scale. - Donors had fairly smooth post-operative courses and tended to fare well in the immediate post-operative period, with no evident depression or major regression. - Shortly after transplantation, 2 couples split and questions about spouse abuse were rise with a third patient.

TABLE V: BONE-MARROW TRANSPLANTATION (BMTx)

AUTHOR/S	AIM OF STUDY	TIMING OF ASSESSMENT/S	GROUP(S)	INSTRUMENTS	FINDINGS AND CONCLUSION
Pfefferbaum et al 1977	To examine whether emotional factors and family relationships had a direct bearing on survival	Crossectional Assessment: - retrospective ratings were conducted post-BMTx	Group 1: 19 recipients Age range: 7-16 years Mean age : unspecified	- Ratings on various aspects of family interaction: number of visits, support, enthusiasm and child's motivation	- No associations were found between psychosocial factors and survival rates or post-hospitalization course.
Gardner et al 1977	To examine the psychological and emotional problems experienced by bone marrow recipients and their families	Crossectional Assessment: - prior to BMTx	Group 1: 7 recipients Age range: 4-15 years Mean age : Unspecified	- Unstandardised interviews - Weschler Intelligence Scale - Rorscharch Test - Thematic Apperception Test - Minnesota Multiphasic Inventory - Figure Drawing - Sentence Completions	- Prior to Tx: anxiety, depression, overdependence, anger and reduced tolerance for medical procedure. - Depression persisted following Tx. - Honest discussions about possible complications, death and opportunity for child and family to discuss their fears seemed essential. - Continuous psychological support was needed.
Pot Mees 1989	To study the effect of transplantation on the child's psychosocial adjustment	Longitudinal Assessment: a) prior to BMTx b) 6 mths post-BMTx c) and at 12 mths after BMTx	Group 1: 26 recipients Age range: 5-18 yrs Mean age : 10.1 yrs Group 2: 28 children who underwent conventional heart surgery Age range: 5-18 yrs Mean age : 8.1 yrs Group 3: 26 healthy children Age range: 5-18 yrs Mean age : 10.1 yrs	- Rutter 's A and B Scales - British Abilities Scales - Griffiths Developmental Scales - Visual Grid (Self-Perception) - The General Health Questionnaire (GHQ-28) for Parents - Dyadic Adjustment Scales (DAS) - Locus of Control - Utrecht Coping List - Unstandardised parental interview	At 12 mths post-MBTx, recipients showed: - An improved physical health. - A raise in level of disturbance from 15% prior Tx to 35% at 12 mths post-Tx. - Lower abilities in reading and arithmetic skills than before Tx. - More disturbance than the groups of healthy children but less behaviour symptoms than the open-heart surgery group. - Children's adjustment following Tx was associated with the emotional state of the parents, especially mothers, and their marital happiness.

AUTHOR/S	AIM OF STUDY	TIMING OF ASSESSMENT/S	GROUP(S)	INSTRUMENTS	FINDINGS AND CONCLUSION
McConville et al 1990	To evaluate the effect of psychosocial factors on medical outcome (complications and survival)	Crossectional Assessment - patients' charts were retrospective reviewed	Group 1: 15 recipients who survived 12 mths post-tx Age range: unspecified Mean age : unspecified Group 2: 17 recipients who did not survived 12 mths post-tx Age range: unspecified Mean age : unspecified	- Severity of Illness and Complication Scales - Global Family Functioning - Brief Psychiatric Rating Scale for children - Brief Psychiatric Rating Scale for parents - Global Assessment of Functioning Scale (CGAS) for children - Global Assessment of Functioning Scale (GAS) for adults	- The non-survivors were more severily ill than the survivors and children who died unexpectedly or had unexpectedly high complication rates showed more psychosocial distress patterns than those surviving up to one year post-Tx . - Child functional impairment, family dysfunction and paternal psychopathology scores were associated with unexpected physical complications and death. - Mothers were more supportive than fathers.
Smedler et al 1990	To examine cognitive functioning following BMTx	Crossectional Assessment: - 1 to 6 yrs after BMTx	Group 1: 32 recipients Age range: 4-23.4 yrs Mean age : unspecified Group 2: 32 sibling-donors Age range: 1-23.4 yrs Mean age : unspecified	- Griffith's Development Scale II - Weschler Intelligence Scale, WISC - Weschler Adult Intelligence Scale, WAIS-R - Halstead-Reitan Neuropsychological Battery	- No neuropsychological deficits were observed in patients who were aged 12 or older at the time of the BMT. - A pronounced delay in motor development was found in children treated before 3 yrs of age. - Patients between 3 and 11 yrs of age at BMTx were at a slight disadvantage, compared to donors, on tasks involving perceptual and fine motor speed.

AUTHOR/S	AIM OF STUDY	TIMING OF ASSESSMENT/S	GROUP(S)	INSTRUMENTS	FINDINGS AND AUTHORS CONCLUSION
Dermatis and Lesko 1990	To examine the prevalence of psychological symptomatology in parents of children undergoing BMTx	Crossectional Assessment: - After consent was obtained for BMTx	Group 1: 61 parents	- The Brief Symptom Inventory (BSI) - Ways of Coping Checklist (WOC) - Unstandardised Quality of Communication Scale (Physician -Parent)	- 47% of father and 60% of mothers exhibited psychological distress. Mother exhibited broader symptomatology, more levels of depression and phobic anxiety than did fathers. - The level of parents' distress was unrelated to characteristics of the child's disease/treatment or information regarding BMTx. - Emotional coping and quality of communication with physician was associated with psychological distress. - Married parents exhibited lower levels of distress than widowed, separated or divorced parents.
Stuber et al 1991	To examine the long-term effect of BMTx and post-traumatic stress disorder (PTSD) following BMTx	Longitudinal Assessment: a) Prior to BMTx b) at 3 mnths post-BMTx, c) 6 mnths post-BMTx and d) 12 mths post-BMTx	Group 1: 6 recipients Age range: 3.7-6.7 yrs Mean age : 4.9 yrs	- Modified PTSD Pynoos and Eth's interview - PTSD Reaction Index - The Play Performance Scale	- Symptoms of PTSD were seen up to 12 mths after Tx. - Survivors demonstrated more denial and avoidance, and fewer arousal symptoms than has been noted in children traumatized by a violent life threat.
Kramer et al 1992	To investigate the intellectual and social development of bone marrow recipients	Longitudinal Assessment: a) Prior to transplantation and b) at 12 mths post-BMTx	Group 1: 22 recipients Age range: 1.1 to 14.1 yrs Mean age : 2.6 yrs	- Vineland Adaptive Behaviour Scales - Bayley Infant Development Scales - Stanford-Binet IV Scales - Weschler Intelligence Scale, WISC.	- There were no decrements in psychological functioning at 12 mths follow-up, regardless of the dose of cranial radiation received or the age at which radiation was administered.

AUTHOR	AIM OF STUDY	TIMING OF ASSESSMENT/S	GROUP(S)	INSTRUMENTS	FINDINGS AND CONCLUSIONS
Sormanti et al 1994	To examine the psychosocial status of parents of transplanted children	Crossectional Assessment: - 1 to 10 years post-BMTx (range since Tx: 12-190 mths)	Group 1: 70 parents Age range: 35-50 yrs Mean age : unspecified	- Unstandardised Family Assessment - Profile of Mood States (POMS)	- Parents reported distress with: fear of relapse, finances, respondent's mental & physical health, insurance, and employment .For each area, the amount of distress decreased as the time since the Tx increased. - Parents also reported improvements following Tx in: outlook of life, relationship with children, ability to handle everyday events and ability to express themselves. However, parents reported an increase in anxiety levels after tx.
Lee et al 1994	To examine parental responsiveness, the relationship between parental responsiveness and child's post-traumatic stress disorder, and the relationship between parental coping and parenting behaviour	Longitudinal Assessment: a) pre-BMTx b) 3 mths post-BMTx, c) 6 mths post-BMTx, d) 12 mths post-BMTx, and e) 24 mths post-BMTx	Group 1: 6 recipients Age range: 3.9-6.9 yrs Mean age : 4.9 yrs Group 2: 7 healthy children Age range: 3-6.3 yrs Mean age : unspecified	- Parent-Child Interaction from 20 minutes videotape (tasks free: play, decision-making and working together). Videotapes were scored using the Family Interaction Q-Sort - Ways of Coping Cancer Questionnaire - PTSD Reaction Index - Unstandardised Parental Interview	- Parental responsiveness remained fairly stable over time, despite new stressors and concurrent changes in the children's physical appearance and medication. - The actual number or type of life stressors the parents were experiencing was not indicative of their level of parental responsiveness. - The parents' educational level appeared to be a mediating factor in their responsiveness. Less well-educated parents had fewer resources for caring for their child than did better educated parents. - Children of less responsive parents scored lower in PTSD. The authors suggest that this was due to: inability to express feelings, avoidance and denial. - Fear and uncertainty was the most frequent problem reported by parents. Social support and focusing on the positive was the coping pattern used by most parents, while behavioural escape-avoidance was the coping style used least often.. No clear relation was found between parental responsiveness and coping.

AUTHOR	AIMS OF STUDY	TIMING OF ASSESSMENT/S	GROUP(S)	INSTRUMENTS	FINDINGS AND AUTHORS CONCLUSION
Stuber and Nader 1995	To provide descriptive data on adolescents' memories of transplantation and symptoms of post-traumatic stress	Crossectional Assessment: - 2 to 12 yrs after BMTx (mean since Tx: 7.5 yrs)	Group 1: 10 recipients Age range: 13-20 yrs Mean age : 16.3 yrs Group 2: 9 healthy friends Age range: 14-20 yrs Mean age : 15.7 yrs	- 90 minutes Clinical Interview - PTSR Reaction Index - Woodcock-Johnson Test of Academic Performance - Wechsler Intelligence Scale, WISC	- The patients reported relatively few symptoms of post-traumatic stress, suggesting that either did not experience Tx as traumatic or denied their traumatic symptoms. - Emotional isolation, fear at thoughts of the event, and intrusive mental images were more commonly reported by survivors than control subjects.
Kanabar et al 1995	To assess quality of life after BMTx	Crossectional Assessment: - 1 to 12 yrs post-BMTx	Group 1: recipients n=14; Age range :3-13 yrs Age mean : 8 yrs n=19;Age range: 15-26 yrs Age mean: 19 yrs	- Quality of Life Feeney et al's Scale (modified) - Open Ended Questionnaire	- 11 children (39%) showed no deficit in: sensation (sight, hearing and speech), mobility, emotions (happiness, worry and anxiety), cognition (ability to learn), self-care and pain. - 10 children (35%) showed some degree of deficit in sensation and mobility but did not required help from others. Only 1 child (3.5%) was unable to eat, drink, bathe, dress and use the toilet as peer group. - 5 children (17.8%) were learning slower than peers or had learning disabilities. - 7 children (25%) complained of occasional pain but not interfering with normal activities or requiring analgesics. Two other children (7%) complained of frequent pains interfering with normal activities, and medication was required. - 5 children (17.8%) were occasionally unhappy, and 2 children (7%) were often unhappy. 9 parents (32%) and 11 the children (39%) expressed anxiety about illness. 10 children (35.7%) lacked of self-confidence and energy, or were worried about their appearances because of either short statue or disfigurement as a result of radiation therapy and/or surgery.

TABLE VI: Heart Transplantation (HTx)

AUTHOR	AIM OF STUDY	TIMING OF ASSESSMENT/S	GROUP(S)	INSTRUMENTS	FINDINGS AND CONCLUSION
Samuelson 1984	To assess functional & social rehabilitation	Retrospective Assessment:	Group 1: 45 recipients Age range: 13-29 yrs Mean age : unspecified	- Unstandardised questionnaire	- HTx not only extends longevity but also provides genuine rehabilitation. - The majority of patients went back to work (39%) or to their studies (44%).
Lawrence and Fricker 1987	To assess quality of life after HTx	Crossectional Assessment: - 3 to 34 mths post HTx	Group 1: 7 recipients Age range: 6-15 yrs Mean age : unspecified	- Personality Inventory - Offer self image questionnaire - Eyberg Child Behaviour Inventory - Projective drawings - A parental interview	- HTx has enabled 7 children to return to normal life styles. In each area measured there was adequate or improved functioning.
Uzar et al 1989	To assess parents concerns and needs during the process of HTx	Crossectional Assessment: - 3 to 24 mths post HTx	Group 1: 10 recipients Age range: 6 mths to 16 yrs Mean age : 8.4 yrs	- Hymovich Chronicity Impact & Coping Instrument (CICI) - Feetham Family Function Survey (FFFS)	- Uncertainty about their child's future was the most serious concern of the parents but also: diminished physical energy, worry about their ability to provide care for the child, and worry about their ability to meet expectations of parents' and spouse' roles.
Uzark 1992	To assess the frequency of psychosocial problems/needs after HTx and to indentify factors associated with psychosocial adaptation.	Crossectional Assessment: - 3 mths post HTx	Group 1: 49 recipients Age range: 4 mths - 13 yrs Mean age : 10.4 yrs Group 2: Normative population	- Family Inventory of Life Events & Change - Family Inventory of Resources - Coping Health Inventory for Parents - Chronic Illness Coping Inventory - Child Behaviour Checklist (CBCL) - Children State Trait Anxiety Scale - Piers & Harris Self-Concept Scale - Adolescent Coping Orientation for Problem Experience	- Most children were active & able to participate in age appropriate activities following Tx.. - HTx recipients showed more emotional & behavioural problems than the normative population. - Low self-concept, increased anxiety and family stress and resources were associated with behavioural problems. Parental coping was associated with child's self-concept, anxiety and coping.

AUTHOR	AIM OF STUDY	TIMING OF ASSESSMENT/S	GROUP(S)	INSTRUMENTS	FINDINGS AND CONCLUSION
DeMaso et al 1995	To examine: a) psychological functioning prior and after Tx ; b) associations between psychological functioning prior to Tx, post-Tx medical severity, family functioning and post-Tx psychological functioning.	Longitudinal Assessment: a) prior HTx b) after HTx (mean time since HTx: 2.1 yrs; range: 1 to 5.2 yrs)	Group 1: 23 recipients Age range: 3 to 20 yrs Mean Age: 13.1 yrs old	- The Children's Global Assessment Scale (CGAS) - The Global Assessment of Relational Functioning Scale (GARF) - Side Effect Severity Scale	- After HTx, 78% of the patients had good psychological functioning. - Patients with psychological difficulties before and after Htx had more hospitalizations following Tx. -Pre-tx, emotional functioning and family functioning were more correlated with post-transplantation psychological functioning than medical severity.

TABLE VII: Heart (Htx) or Heart-Lung (HLTx) Transplantation

AUTHOR	AIM OF STUDY	TIMING OF ASSESSMENT/S	GROUP(S)	INSTRUMENTS	FINDINGS AND CONCLUSION
Wray et al 1992	Cognitive development	Longitudinal Assessment: a) prior to HTx or HLTx b) 3 mths post-HTx or HLTx	Group 1: 28 recipients Age range: 1 mth to 16 yrs Mean age : 8.6 yrs Group 2: 28 normal children Age range: 3 mths to 15.8 yrs Mean age : 8.5 yrs	- Ruth Griffiths Mental Developmental Scales - British Ability Scales - Rutter Scales A & B - Behaviour Checklist (RBCL)	- 45% of the children showed behavioural problems at home pre-Tx, whilst only 20% of the children showed behavioural problems post Tx.. - The incidence of behavioural problems was higher in the Tx recipients than in control group. No differences were found regarding behavioural problems at school between the groups. - Locomotor development was delayed prior to Tx but was significantly improved after surgery. Cognitive development pre & post Tx was within normal range.
Wray et al 1994	Impact of transplantation in cognitive development	Crossesctional Assessment - retrospective: 3 to 25 mths after HTx or HLTx	Group 1: 65 recipients Age range: 1 mth to 17 yrs Mean age : Group 2: 52 children who had conventional cardiac surgery Age range: unspecified Mean age : 6.2 yrs Group 3: 45 healthy children Age range: unspecified Mean age : 8.2 yrs	- Ruth Griffiths Mental Development Scales - British Ability Scales - Rutter Scales A & B - Behaviour Checklist (RBLC) - Semi-structured Interview for parents	- Although all the mean subtest scores & overall intelligence quotient were within the normal range in the Tx group, they had lower scores on the developmental parameters than the healthy group but did not differ from the open heart surgery group. - 33% of the heart recipients & 15% of the heart-lung recipients had behavioural problems at home. - Differences in the prevalence of behavioural problems were not found between the groups.

AUTHOR/S	AIMS OF THE STUDY	TIMING OF ASSESSMENT/S	GROUP(S)	INSTRUMENTS	FINDINGS AND CONCLUSION
Serrano-Ikkos et al 1997	To monitor psychosocial adjustment in a group of paediatric candidates for transplantation and their families.	Longitudinal Assessment: a) pre Htx or HLTx (at initial referral), b) 6 mths later (pre Htx or HLTx)	Group 1: 51 families Patients' age range: 5 - 16.7 yrs Mean age: 11.2 yrs Siblings' age range: 7.7 – 16.9 yrs Mean age: 10.7 yrs	- Index of Physical Ability - Rutter & Graham's Psychiatric Interview for Children - Global Assessment of Functioning Scale (GAF) - Birleson Depression Scale, - Rutter's A & B Scales - Piers & Harris Self-Concept Scale - Camberwell Family Interview - General Health Questionnaire (GHQ-28) for parents - Golombok-Rust Inventory of Marital State (GRIMS)	- At entry into the study, 25% of the patients had an emotional disorder, and over 50% showed impaired psychosocial functioning. - 20% of the well siblings manifested behavioural or emotional problems. Over 50% of the families had long-standing family conflicts. - Only 23 children were available for follow-up. Over 6 mths period, there were no changes in psychological health of the patient, well sibling or parents.

3. Adherence to treatment in Children with Life-Threatening Conditions

Although Hippocrates identified poor adherence to treatment as a difficulty faced by physicians during the 5th century BC (Chadwick and Mann 1950), there is as yet no universally accepted definition (Lask 1994). In general terms, adherence to treatment describes the extent to which the doctor's perception of the correct form of treatment is shared by the patient (Thompson 1984). This way of describing patient behaviour underlines the fact that both patient and doctor have their own ways of interpreting the 'facts' of illness and unless there is substantial agreement, successful treatment is unlikely.

Attention to adherence with medication as well as with behavioural treatment in the paediatric population is important if one is to evaluate a) the efficacy, b) the safety, and c) the side effects of the treatment; and if one is to obtain maximum therapeutic benefit (Dunbar 1983). Review studies have shown that overall non-adherence in the paediatric population varies from 20-80% (Litt and Cuskey 1980; Shope 1981). Similar rates of non-adherence have been found among children with life threatening conditions, such as cancer and cystic fibrosis (Tebbi et al 1986; Koocher 1986; Smith et al 1979; Muszynski-Kwan et al 1988; Passero et al 1981). These findings suggest that severity of illness and the seriousness of the implications of failing to take the medication have not been shown to influence adherence.

It is surprising that only a few studies have evaluated the prevalence of non-adherent behaviour with immunosuppressive therapy amongst child and adolescent transplant recipients. Most of these studies have been carried out following renal transplantation, and no attention has been paid to children undergoing heart and heart-lung transplantation.

Korsch et al (1978) suspected non-adherence following renal transplantation when diminution in cushingoid features, unexplained weight loss or changes in renal functioning occurred. In addition, the specialist nurse reviewed which medicines and how much the patient was taking and obtained verbal confirmation of non-adherence by interview. It was found that 14 of 80 children (10.7%) were non-adherent. In another study with paediatric renal recipients, non-adherence was assessed via interview, frequency of refills, and accuracy of pill counts (Beck et

al 1980). Forty three percent of the sample was found to be in some way non-adherent with the immunosuppressant therapy. Using more objective measures of adherence, such as the concentration of prednisone and cortisol in the serum, Hesse et al (1990) compared the incidence of adherence between a group of 7 children and adolescents who received renal transplantation, and 52 adult renal transplant recipients. The incidence of poor adherence was significantly higher in the paediatric population as compared with adults. Swanson et al (1991) has reported similar results.

Adherence problems following bone marrow transplantation have also been reported. In a retrospective study, Phipps et al (1990) reviewed the in-patient records on all patients admitted to the BMT unit during a 3-year period. Children were classified as having adherence problems if there were multiple references to episodes of unsatisfactory adherence in their notes. Significant adherence difficulties, for which intervention was required, were identified in 28 of 54 children (52%).

Maintenance of transplant recipients on immunosuppressive medication is an essential component of a successful outcome with the transplanted organ (Rodin and Abbey 1992). Several studies on paediatric transplant recipients have found an association between unsatisfactory adherence with immunosuppressive therapy and both an increased number of rejection episodes and allograft failure which has led, in some cases, to premature complications and death (Korsch et al 1978; Beck et al 1980, Hesse et al 1990, Dunn et al 1990; Whitehead et al 1994). Given this association, it is striking that there is only a small number of studies aiming at identifying pre-surgical risk factors associated with non-adherence following transplantation in the paediatric population. TABLE VIII (see page 54) summarises the risk factors highlighted by these studies.

Although these studies seem to indicate that demographic characteristic as well as psychosocial and illness related factors are mediators of poor adherence to treatment, some of the results shown on the table are contradictory. It needs to be said that the authors have used different methodologies and definitions of adherence. More research is required to investigate how all these factors interact with each other.

The aversive side-effects of treatment following transplantation, such as hypertrichosis, gingival hypertrophy, dizziness, and the effects of immunosuppressants on physical appearance (cushingoid features, puffy faces, hirsutism) have also been found to be associated

TABLE VIII

Predictors of Poor Adherence in Paediatric Recipients

Study	Transplant	Risk Factors
Korsch et al (1978)	Renal	Adolescence Female Absent Father Poor Family Communication Poor Understanding of Illness Cosmetic Side Effects Low Family Incomes Absent Support from Community
Beck et al (1980)	Renal	Adolescence Female Family Instability No Parental Involvement in Treatment
Phipps et al (1990)	Bone Marrow	Pre-school & School Age Children Aversive Side Effects of Treatment
Foulkes et al (1991)	Renal	Number of Stressful Family Events Emotional Support from Father Informative Support from Father

with poor adherence in children with cancer (Koocher 1986, Tebbi et al 1986). Other illness related factors associated with adherence to treatment, which have been mentioned in the literature of chronic and life-threatening conditions in childhood, are: the length and complexity of treatment (Haynes et al 1979), disease chronicity (La Greca 1988; Litt and Cuskey 1980; Shope 1981) and the degree to which the child's normal developmental behaviour is interrupted by treatment (Gordis et al 1969; Koocher 1986; Friedman and Litt 1986).

Attitudes and behaviours of family members can be crucial (Eiser 1993). Hanson et al (1987) found that positive family relationships were associated with good adherence behaviour in adolescents with diabetes. However, larger number of siblings (Gordis et al 1969), difficulties in family communication and negative mother-child interaction have been found to be associated with poor adherence in children and adolescents with cystic fibrosis (CF), diabetes, asthma, cancer and rheumatoid arthritis (Patterson 1985; Schobinger et al 1993; Tebbi et al 1986; Chaney and Peterson 1989). In contrast, Geiss et al (1992) reported higher levels of perceived compliance with CF treatment associated with less satisfactory marital relationships and, to a lesser degree, with less frequent social contacts. Different explanations may account for these results: a) when marital satisfaction and social contact are lower, mothers may become more involved with patient care. An extremely high level of involvement that entails rigid compliance with the child's medical treatment may be regarded as a substitute for positive marital and social interactions. But it could also be argued that b) maintaining what physicians considered to be high levels of compliance may interfere with the parents' ability to maintain satisfactory marital and social relations.

In an interesting study of children with rheumatoid arthritis, Chaney and Peterson (1989) showed that family life events as well as the mother's coping behaviour were significantly related to adherence. The authors indicated that as the number of family stressors increased, compliance decreased; and that families in which mothers reported a greater number of coping behaviours, had children who demonstrated higher levels of medication compliance.

Factors associated with non-adherence are not simply illness related or parent-child specific. Problems associated with patterns of interaction between professionals, the child and the parents are also of relevance. As Thompson (1984) has stated in his review on this topic, the parents' and child's satisfaction with a) the consultation, b) communication and c) medical care received, as well as the clarity of the physician's terminology, are all associated with patient's adherence to treatment. There is now some evidence suggesting that what happens between the parents and the professionals at the time of the child's diagnosis may be crucial for the short and long term adjustment of the child and family (Davis 1993). In a study on how much information parents recall of the period immediately before the diagnosis of cancer, Eiser et al

(1994) found that, on diagnosis, all parents recalled the information that was given about cancer and proposed treatment. Fewer recalled that any information was given about side effects, complications or the cause of illness. In addition, many parents felt unable to ask questions even when given the opportunity. In a study on patterns of communication among children with leukaemia, parents and physicians, Mulhern et al (1981) found major disagreement and misunderstanding between physician and parents with regard to the children's prognosis.

The results of these studies suggest that parents often do not understand what they are told by the physicians and that misunderstanding may have a detrimental effect on the child's adherence. Tebbi et al (1986) reported that children who were compliers with cancer treatment differed from non-compliers in how well they understood the instructions concerning how to take the medication. Recent research on adherence issues in the paediatric population is also pointing to the need to evaluate gender differences regarding parents' attributions about the illness, its implications for parents' relationships with medical staff and for the short and long-term acceptance of treatment. Eiser et al (1995) reported that fathers were more likely than mothers to accept that there was no known cause for the child's illness and that fathers who blamed doctors for failing to diagnose the condition found medical staff less helpful up to 2 years later. To what extent these parents may have more difficulties believing in the effectiveness of treatment, and administering it to their children has not been explored.

Future research aimed at identifying risk factors associated with unsatisfactory adherence to treatment should use longitudinal designs with a multivariate approach, using multiple measures for the assessment of adherence and taking into account the relationship between physiological, psychosocial and clinical variables.

CHAPTER 3

THEORETICAL BACKGROUND AND METHODOLOGY OF THE STUDY

In the previous chapter, I have highlighted the challenges faced by children with life-threatening conditions in need of organ transplantation as well as the challenges faced by their parents. In the light of these challenges it is not surprising that researchers have become increasingly interested in exploring the psychological sequelae of chronic conditions.

Using methods derived from the medical and health psychology approaches, studies on children's adjustment have found that as many as 30-35% of children with physical disabilities showed psychological disturbance, and that children with chronic disease but not physical disability, were less at risk for psychiatric disorder and poor social adjustment (Steinhausen et al 1981, 1983; Rutter 1989; Jessop and Stein 1985; Breslau 1985). However, not all children with physical disabilities develop adjustment difficulties. This raises the question of why some children and their families cope well with the stresses imposed by their condition while others have difficulties. Researchers have attempted to find an answer to this question but the results are contradictory. Wallander and colleagues (1989) suggest that adaptation to chronic disease and disability is determined by the interplay of risk and resistance factors. In their model, risk factors refer to disease and disability variables (e.g. severity, visibility, level of cognitive functioning), and psychosocial stresses (e.g. handicap problems, life events related to the disease), and resistance factors refer to intrapersonal variables (e.g. temperament, motivation), socio-ecological factors (e.g. family environment, support) and stress processing (cognitive appraisal and coping strategies). Whilst these authors argue that adaptation to disease can be predicted by exploring this balance, the review of their own series of studies indicates that 'several of the predictions made by their conceptual model have not been substantiated, whilst others have received only tentative support' (Bradford 1997, pp.139-153). In Wallander's studies it was found that the child's adjustment was not predicted by: child's diagnosis, severity of the condition, physical health and disability, a learning difficulty, the child's functional independence or maternal adaptation. Child's adjustment was associated with family conflict, poor family cohesion, poor parental control, financial hardship and maternal education level. Maternal adaptation was associated with social support, marital satisfaction and duration of marriage. However,

other researches using different methods have found that severity of illness, visibility, and socially embarrassing and humiliating symptomatology are directly related to an increased rate of psychiatric disorder and poor psychosocial adjustment (Steinhausen et al 1981, 1983; Rutter 1989; Jessop and Stein 1985; Breslau 1985; Cadman et al 1987; Pless and Satterwhite 1975; Perrin et al 1989; Engstrom 1992).

Evidence from studies on adaptation in children and adolescents with congenital heart disease and cystic fibrosis also reveals a contradictory picture. Some studies suggest that these children achieve an age-appropriate level of adjustment at home and school (Linde et al 1966; Linde 1982; Drotar et al 1981; Bywater 1981); whilst others show a higher rate of behavioural problems than expected from the test norms (Spurkland et al 1993; Cowen et al 1986). Associations between the children's psychosocial functioning and physical impairment in children with heart conditions and in children with cystic fibrosis have also been reported (O'Dougherty et al 1983; Kramer et al 1989; Steinhausen and Schindler 1981). Adjustment difficulties in paediatric heart recipients were associated with greater family stress and diminished family resources for managing stress; whilst open family communication and quality of family support were found to be protective factors (Uzark et al 1992).

Researchers in chronic illness have explained the discrepancies in terms of the different methods used and the diverse definitions given to the parameters measured (Eiser 1993; Bradford 1997). Whilst Bradford recognises that Wallander et al's (1989) multi-factorial model of adjustment to chronic illness 'represents the most coherent theory to date and that the model is unique in making specific predictions as to which children and families are most at risk of poor coping, as well as the reasons why' (Bradford 1997, p. 153), he also asserts that there is a need to re-formulate the model so that its predictive validity can be strengthened. Bradford goes on to suggest that "there is a need to re-define the socio-ecological factors, such as the family parameters, incorporating concepts and measures from family systems theory which will offer a way forward in thinking how family interactions mediate adjustment and what dimensions might be important to explore" (Bradford 1997, p.158). Bradford's suggestion implies an integration of paediatric health psychology and systems theory approaches.

The present study was developed within the systems theory approach. Systems theory provides a framework to explore the way in which people behave and the problems they have as being related to their family structure and organisation, which in turn is influenced by the wider environment. In systems theory, two of the key principles are that i) a change in one part of the system is associated with changes in all other parts, and ii) that systems maintain a balance between periods of change and stability. The confirmation of a medical diagnosis which suggests the child's need for transplantation represents a 'hazardous event', which the family needs to adjust to by establishing a new balance to cope with the short and long-term challenges it will face. Such an event will not only have an effect on all other family members but the way in which each family member attempts to adjust will, in turn, have a further impact on the family.

The main aims of the present research were:

i) To explore the way in which the family organises itself at the time its members are
   informed about the child's need for transplantation, and

ii) To explore whether the way in which the family is organised has a long-term effect on the
   patient's medical outcome.

The methods used to assess family organisation have been developed by researchers and clinicians working in social psychiatry, medical sociology and health problems. The theoretical issues that lead to the present investigation will be briefly described in the first section of this chapter. In the second section, the context and the methodological approach used in this study will be presented.

3.1 <u>FAMILY SYSTEMS THEORY</u>

"Family medicine research is just beginning to be influenced by the range of family theories available from other disciplines and to develop its own ideas about the importance of family functioning in relation to physical illness and health" (Baker and Patterson 1989, p. 57). Based on systems theory, Engel's concept of the biopsychosocial model (Engel 1977) has expanded our field of vision to include hierarchical, interdependent relationships of biological, psychological, individual, family and community systems. Since Engel's publication, family

therapists and researchers have developed theoretical models of illness and family systems. McDaniel et al (1992, pp. 32-35) have emphasised the major theoretical contributions to this growing field. These are: 'the psychosomatic family model (Minuchin et al 1978); the family paradigm model (Reiss et al 1986); the family system illness model (Rolland 1988); the biopsychosocial approach to mental illness and expressed emotion index (EE) (Brown et al 1962; Leff and Vaughn 1985); and finally the family adjustment and adaptation response (FAAR) (Patterson 1989).

Concepts from these different theoretical models, or from frameworks that have evolved from them, have been integrated creating the setting of the research presented in this thesis. Therefore, I will now discuss the most relevant aspects of those models in order to clarify the theoretical context in which the present research has evolved.

The Psychosomatic Model

The pioneering contribution to family systems and medical issues was Minuchin, Rosman and Baker's psychosomatic family model. This model is based on structural family therapy theory, and evolved from clinical and research observations of families of children with uncontrolled childhood diabetes. Minuchin et al (1978) showed that episodes of ketoacidosis and acetonuria in these children were associated with triangulation of the child in marital conflict. Interactional patterns of enmeshment, overprotection, rigidity, and poor conflict resolution/avoidance also co-occurred on those families. Similar patterns were found in families of anorexic girls (Minuchin et al 1978). This model postulates 'that certain types of family organisation are closely related to the development and maintenance of psychosomatic syndromes in children, and that the child's psychosomatic symptoms in turn play an important role in maintaining the family homeostasis' (Minuchin et al, 1978, p. 20). Whilst in this model identifies 'pathological ' patterns of interaction, and the family is seen as contributing to the onset of illness and unfavourable outcome, the model does not suggest that the observed patterns are necessarily always abnormal. Families that function well are also likely to show similar patterns of interaction at times of stress and conflict. What distinguishes these families from the 'psychosomatic' ones is the inability to move on from a pattern of interaction which may be adaptive at that time of crisis but becomes maladaptive

Kog et al (1985) criticised the lack of clear definitions of the psychosomatic family model and suggested that the psychosomatic family model could be reduced to 3 interactional dimensions: the intensity of intrafamilial boundaries (cohesion), the degree of the family's adaptability, and the family's way of handling conflicts. In a control study comparing 50 families with a hospitalised eating disorder patient to 220 nonpatient control families, Kog and colleagues found that all eating disorder families reported significantly more conflict and disorganisation but did not differ from the nonpatient families in cohesion (Kog et al 1985).

Clinical trials of family intervention devised from a structural family therapy model provide some indirect evidence for the validity of the model. The first controlled study on childhood asthma, showed that interventions in family interactions do have an effect on disease activity (Lask and Matthew 1979). Children with moderate to severe asthma were randomly allocated to one of two treatment groups: standard medical treatment or standard medical treatment as well as 6 hours of family treatment during a four-month period. Children in the family treatment group were significantly better in day-wheeze and thoracic gas volume than the children receiving conventional treatment. This study was replicated by Gustafsson et al (1986) obtaining similar results. The effectiveness of family interventions in improving anorexia nervosa symptoms (Le Grange 1993), and in improving and maintaining the levels of glycosylated haemoglobin in adolescents with diabetes (Satin et al 1989) have also been demonstrated.

However, Woods et al (1989) argued that the key features of the psychosomatic family model have not been investigated, since none of the studies have explored associations between family dysfunction, triangulation of the ill child to detour marital dysfunction, and disease activity. To test the model, forty families of children with Crohn's disease, ulcerative colitis, and functional recurrent abdominal pain syndrome were rated during standard, videotaped family interactional tasks, lunch, and interview. As expected from the model, greater degrees of certain psychosomatic family pattern were associated with greater disease activity within disease type. The psychosomatic family patterns associated with greater disease activity were rigidity, triangulation of the child and marital dysfunction; whereas enmeshment and overprotection were associated with less disease activity. This result suggests that some family patterns may be maladaptive while others may be adaptive. The authors concluded that overt symptomatology is crucial in organising family patterns of interaction, and that the family and

disease mutually affect one another. With a similar way of thinking, Lask and Fosson described the Illness Network Model (1989) in which the chronically ill child is placed in the context of the family. These authors developed a multifactorial approach to childhood illness in which biological, psychological, developmental and social factors are integrated, and changes in one factor lead to changes in another factor. This model proposes that adverse psychological consequences of an illness can play a key role in the perpetuation or aggravation of the illness, while optimal coping is likely to lead to better health care and outcome.

More recently, Wood has described the Biobehavioural Family Model of chronic illness in children and adolescents (Wood 1994). This model focuses on the dynamic pathways by which patterns of family interaction and physiological function influence each other, and assumes that the well-being of the patient depends on a dynamic balance among three levels of functioning: individual physical functioning, individual psychological functioning, and family-social functioning. The family has a pivotal role in achieving biopsychosocial balance. This model proposes that the "family systems properties of proximity, generational hierarchy, triangulation, parental relationship and interpersonal responsivity are factors that influence one another and interact with individual (family member) biobehavioural reactivity in ways which either buffer or activate psychobiological processes related to disease activity" (Wood 1994, p. 57). In this model, proximity is defined as the extent to which various domains of interaction (e.g. personal space, private information, emotions and decisions) are shared by family members. Generational hierarchy is defined as the extent to which parents are in charge of their children by providing them with nurturance and limits through a strong parental alliance. Responsivity is conceptualised as the degree to which people respond to one another physiologically, emotionally and behaviourally. High levels of interpersonal responsivity can be beneficial when the emotions are positive ones but it can also be detrimental if the emotions are primarily negative.

In an attempt to elaborate homeostatic processes within this model, the Kentucky group (Wilson et al 1996) developed the Family Homeostatic Model which postulates that:: 1) Families that have been coping with a chronic illness over a long period of time will have adapted to this strain by improving their cohesion, organisation, control, expressiveness and other important aspects of family environment. Therefore, these families will be functioning at or above population norms on measures of family environment. Similarly, chronically ill

children are expected to function above population norms (i.e. show higher self-esteem and less depression). 2) If physical functioning declines, family functioning will in the short term improve in response to that decline, in the same way that homeostatic alterations attempt to compensate for changes in blood pressure within a physiological system. Furthermore, improvements in family functioning would also help maintain children's psychological functioning. The last prediction is that 3) exogenous stressors will compromise the ability of the family to compensate for the physical decline. To test the model, this group conducted a two-year longitudinal study with 35 children with cystic fibrosis and their families (Wilson et al 1996). It was found that despite a decline in lung function measures over the course of the study, children with cystic fibrosis reported lower depression and higher self-esteem than population norms, and improvement in psychological functioning. A decline in biological indicators of health was associated with increases in family cohesion, expressiveness and organisation, and with maintenance of high self-esteem and low depression. As expected, additional major life events were associated with deterioration in pulmonary function, higher levels of depression and poor scores on the family environment measures.

The Family Paradigm Model

The second theoretical framework, the family paradigm model, comes from Reiss, Steinglass, Jacobs and colleagues' at the George Washington University. This model is based on an ecological approach and its emphasis is on describing differences in family style that may or may not be dysfunctional depending on particular challenges facing the family and the different settings in which they find themselves. Their model focused on how families organised themselves around health problems, and was built around the concept of a family paradigm in which family identity is based on three dimensions: configuration, coordination and closure. Configuration 'refers to the pattern or coherence families can recognise in complex and ambiguous situations. Families that can grasp complex patterns and connections probably have more confidence and optimism, and they can understand and master a broad range of surprising and challenging situations in everyday life. This is measured by the family's ability to recognise pattern concepts in problem-solving procedures. Coordination reflects the family's belief that they must, necessarily, face ambiguous situations as a unified group rather than as individuals. This is measured by the level of cooperation and integrated planning in the problem-solving setting. Finally, delayed closure reflects the family's openness to new

information and it is measured by their ability to change their problem solutions in accordance with new data (Reiss et al 1986).

This group showed that highly coordinated families adapted very successfully to the onset of psychiatric illness in an adolescent member (Costell et al 1981). However, in a second study, the researchers found that the survival of patients with unremitting chronic stage renal disease was shorter in high-coordination families (Reiss 1986). The author concluded that high coordination, though an effective response to an acute medical emergency, if it persists may make the family more vulnerable in the long run. Initially, these families focus heavily on the patient's needs, neglecting or excluding other family members. As the illness progresses and the patient becomes terminal, the family undergo a process of reorganisation (often protecting the healthy family system that remains) in which the patient is excluded from family decision-making. At the same time that family members emotionally distance themselves from the patient, reducing their involvement with his medical care. This mechanism of "walling off" may be mutual and has also been described by Bluebond-Langer (1978,1996). Using an anthropological model, this author demonstrated the active role played by children with terminal cancer or cystic fibrosis in the distancing and exclusion process. In this model adaptation is regulated by the fit between families of a particular type and situations of particular type (challenge or stressor faced by the family in specific settings such as neighbourhood, hospitals, work, characteristics of the extended kin etc.).

The Family Systems-Illness Model

The third theoretical contribution within the biopsychosocial framework is John Rolland's family system-illness model that describes illness and disability in psychosocial terms (Rolland 1984). This model has a psycho-educational emphasis, and predominantly evolved from Rolland and colleagues' clinical work at the Centre for Illness in Families in New Haven, Connecticut. The model has 3 dimensions:

a) Illness typology (onset, course, outcome, and degree of incapacitation); b) Time stages of the illness (crises: pre-diagnosis, diagnosis and initial adjustment; chronic: chronic long haul, and terminal: pre-terminal, death, mourning and resolution of loss); and c) Components of family functioning which refers to family structural/organizational patterns; communication processes; multigenerational patterns and family life cycle; and family

belief systems. Rolland (1994) has pointed out that physical illness may be grouped according to their psychosocial aspects as follows:

1. - Acute or gradual onset;

2. – Progressive, relapsing or constant;

3. - Incapacitation (none, mild, moderate, severe)

4. – Crisis, chronic or terminal phase; and

5. – Fatal, non-fatal or shorten life span

Families confronted with gradual, progressive, incapacitating but non-fatal disorders (e.g. rheumatoid arthritis) will have a different set of challenges than families facing an acute, fatal condition (e.g. acute leukaemia) . Rolland has pointed out that "in chronic and life-threatening disorders, the primary focus is the system created by the interaction of a condition (the psychosocial demands of the particular disorder over time) with an individual, the family, and with other biopsychosocial systems. This model takes the family as its central point because, from the perspective of clinical assessment and intervention, the family may provide the best lens through which to view these other systems" (Rolland 1994, p. 11).

The Expressed Emotion Index

A separate strand of work with elements of a family systems approach is that pursued by Brown et al (1962). This has postulated that the attitudes and behaviours of a close relative will influence the course of psychiatric illness. From this model, the expressed emotion (EE) index was developed. This is perhaps the best validated risk indicator for relapses of psycho-affective disorders (Brown et al 1962; Vaughn and Leff, 1976a,b).

The EE index scores the attitudes and behaviours that a relative expresses during a systematic interview (Camberwell Family Interview -CFI), and reported behaviour outside the interview, towards a symptomatic member of the family. Five scales comprise this measurement (critical comments, hostility, emotional overinvolvement, warmth, and positive remarks). It has been shown that hospitalised patients who are discharged to families with high levels of expressed emotion (i.e. with at least one relative scoring high in critical comments, overinvolvement or hostility) are two to three times more likely to experience relapse and re-hospitalisation at 9 months to 1 year periods of follow-up than those patients returning to families with low levels of expressed emotion. Despite the differences in the course of chronic psychiatric disturbance

or general cultural attitudes towards it, EE has been found to be a psychosocial risk marker for the exacerbation of severe psychopathology in many cultures (for literature review see Kavanagh 1992).

Studies on children and adolescents with depression, disruptive behaviours, obsessive compulsive, conduct and emotional disorders have also shown a correlation between the criticism element of EE and the child's behaviour (Asarnow et al 1993; Hibbs et al 1991; Vostanis et al, 1994; 1995). Furthermore, Schwartz et al (1990) demonstrated that a higher degree of maternal EE was associated with a three-fold increase in a child's risk for having a psychiatric diagnosis. High parental criticism has also been found to be associated with poor weight gain and an increased rate of dropout from treatment in studies involving families of young patients with eating disorder (Le Grange et al 1992; Szmukler et al 1985; Van Furth et al 1996).

It is unclear whether the development of high EE attitudes is the result of the disturbed family member's behaviours (family state) or whether high EE attitudes are parental attributes (family trait) which may contribute to the patient's behaviours. If EE are state variables, it can be hypothesised that they will recede in the face of improvements in the patient's clinical state. If EE are trait variables, high EE in the parents would not necessarily decrease once the burden of the patient's illness has lifted (Doane and Diamond 1994). A number of treatment studies have suggested that family intervention is successful in reducing relapse rates, improving the patient's psychosocial functioning and overall family functioning, and that this is associated with lower rates of EE (Doane et al 1986; Le Grange 1993; Rund et al 1995; Van Furth et al 1996). However, some researchers have shown that for some families, high EE attitudes persist or reassert themselves even after intensive family treatment (Doane and Diamond 1994; Tarrier et al 1988). This suggests that there are more complex factors underlying intrafamilial forces that persist even after the patient has stabilised and recovered. To clarify whether the EE components are trait or state attitudes, Schreiber and colleagues (1995) conducted a study in which they interviewed 17 families rating their attitudes towards two of their children, with and without a mental illness. Results showed that EE response patterns directed toward patients and siblings were significantly different on 2 measures. Parents showed significantly more overinvolvement with the mentally ill child and significantly more warmth towards the well child. The authors concluded that overinvolvement and warmth scales are related to the

state of the child, and the lack of significant differences in criticism suggests that this is a parental trait. There is little indication of what actually fuels high EE type behaviours.

Currently, researchers are trying to understand what sort of factors make a particular relative to be high or low EE. Attention has been drawn to the association of high EE with psychopathology in both the child and the parents (Hibbs et al 1991; Schwartz et al 1990; Dossetor et al 1994). Stressed mothers have been found to interact with their children using more negative attitudes when compared to mothers with lower levels of stress (Hammen et al 1987). It has also been reported that parents with high EE critical partners are subjected to higher levels of stress and that there is an association between criticism levels and marital satisfaction (Hooley and Teasdale 1989). Poor-quality marriage, family conflict, and poor practical support have also been associated with parental high EE (Dossetor et al 1994; Hibbs et al 1993). Recent work linking attachment theory and the expressed emotion constructs illuminates the relationship between high EE type behaviours and relative's characteristics. Wynne (1984) suggested earlier that emotional overinvolvement and criticism are special forms of attachment/caregiving that are likely to lead to dysfunctional communicating, problem-solving, and intimacy. Doane and Diamond (1994) explored this further and found that many parents with disturbed attachment histories persisted in expressing negative attitudes over the course of family treatment regardless of patient improvement. The linkages between disturbances in the parents' attachments to their own parents and the degree of negative attitudes that they display toward their disturbed child suggest a conceptual model for the transgenerational transmission of psychosocial risk factors related to attachment. For clinicians, because of the treatment implications, this issue of the underlying dynamics of EE remains an important one, still to be further investigated in the research literature.

Research on EE and illness outcome has extended well beyond mental disorders to include studies on relatives of nonpsychiatrically ill patients. Obese adult subjects living with a high EE relative were found to comply much less with diet treatment than those subjects living with a low EE relative (Flanagan and Wagner 1991). Similarly, Jadresic (1988) has shown associations between high parental EE and high seizure frequencies in young patients with epilepsy. Koenigsberg et al (1993; 1995) demonstrated that patients with insulin-dependent diabetes mellitus living with highly critical relatives had poorer metabolic control than those living with low or non-critical relatives. In addition, three independent studies have shown that

parents of asthmatic children were more likely to be rated higher in criticism when compared to parent of healthy controls; and that children with critical parents had more frequent and more severe asthma attacks, and worse adherence to medication, than children living in low EE criticism households (Hermanns et al 1989; Schobinger et al 1993; Wamboldt et al 1995).

If EE was first used to characterise how parents related to their adult schizophrenic children, in the medical illness context, some authors have speculated that high expressed emotion. is an indication of unhealthy parental and spousal interactions, or an indication of failure to cope with a disorder or chronic handicap within the family (Orford et al 1987; Dossetor et al 1994). Conceptualising EE as a measure of fusion and emotional reactivity in families, Fischmann-Havstad and Marston (1984) found that individuals from fused, reactive families (high EE criticism) were less successful in their attempts to maintain weight loss than those individuals with less reactive families (low EE criticism). Dossetor et al (1994) found that parents scoring high in EE overinvolvement tended to complain more about their difficulties coping with their child's intellectual impairment, have more psychological ill health, a worse quality marriage, and received less practical support from friends and relatives, but more professional support, than those parents scoring low EE overinvolvement. The children in the first group had greater impairment and more behaviour disturbances in public. Compared to those with low EE criticism, parents scoring high on this scale had adolescents with less severe intellectual impairment, more symptoms of high activity and agitation, and received less professional support. Nevertheless, the quality of the marriage, and social support from friends and relative were unaffected in this group of parents (Dossetor et al 1994). These findings suggest that the carer's own characteristics and those of the person with intellectual impairment influence and perpetuate the qualities in the other.

Findings reported by Stevenson et al (1991), Invernizzi et al (1991) and Blair et al (1995), shed new light on the EE index, suggesting that these scales may have a different predictive value for outcome when studying physical illness as compared to mental disorders. Blair et al (1995) questions whether high emotional overinvolvement in parents of severely ill children represents a natural reaction by parents to the gravity of the illness. In a controlled study, these researchers found that there were no differences in levels of EE criticism when comparing parents of healthy children, and parents of children with cystic fibrosis and anorexia nervosa. However, healthy young subjects were living in households which did not show high

overinvolvement, compared to over 30% of the children with anorexia nervosa and cystic fibrosis (Blair et al 1995), and this scale correlated with severity of illness. On the other hand, Stevenson et al (1991) found that high overinvolvement, which had generally been associated with poor prognosis in studies on mental illness, had a good impact on patients suffering from a physical disorder. Good glycaemic control in adolescents with type I diabetes was found to be associated with high emotional overinvolvement; and against expectations based on the EE literature, poor control was not associated with parental criticism or marital discord (Stevenson et al 1991). This protective value of the overinvolvement scale was confirmed by Invernizzi et al's study (1991) which showed that high overinvolvement and warmth in families of heart operated adult patients were associated with lower levels of anxiety and depression in the patient, and with better 12-months outcome of the illness. Positive correlations between warmth and metabolic control in adolescents and young adults with diabetes mellitus has also been reported (Koenigsberg et al 1995), but, in this study, the overinvolvement scale failed to show any association with glycosylated haemoglobin levels.

Studies that did not use the EE measure as such, but rather different methods of measuring parental attitudes have also found associations between warmth and overprotection and adjustment in adolescents with corrected bowel malformations and children with congenital heart disease (Diseth et al 1995; Linde et al 1966; 1970).

The role of the different EE components in predicting physical and psychological outcome with patients with chronic physical disorders needs further investigation and has implications for treatment.

The Family Adjustment and Adaptation Response (FAAR) Model

The fifth important contribution to medical family therapy comes from McCubbin et al's model (Patterson 1989), the family adjustment and adaptation response (FAAR) model which combines family stress theory and family systems theory. The focus is on family efforts to manage the demands (threats) it faces from life events (stressors) and strains (tensions associated with the desire to change something) with its capabilities for meeting demands (resources and coping behaviours), mediated by meanings so as to achieve a balance in family functioning (adjustment or adaptation). Because families do change as a result of their circumstances, this model places an emphasis on viewing the family's response over time. This

time perspective has given rise to a two-phase model: the adjustment phase (this is a period of stability in which the family meet demands with their existing capabilities) and the adaptation phase (the family tries to restore a balance involving changes in roles, rules, patterns of interaction and meaning). These phases are separated by a period of family crises which is characterised by disorganisation and disruption (old patterns and capabilities are no longer adequate) and change is called for. Three levels of systems are considered: individual family members, the family unit, and the community. Each of this is characterised by both demand and capabilities; and there are two types of capabilities: resources (what the family has) and coping behaviours (what the individual/ family does to reduce or manage the demand).

The FAAR model is useful for examining the impact of chronic illness on the family and for determining what resources, coping behaviours, and meanings in the family facilitate successful adaptation, and how this changes over time in response to the chronic strain of the illness (Patterson 1989). Within this framework, medical treatment is viewed as a 'demand' and whether the family can adhere to the treatment is viewed as one indicator of 'adaptation'. Patterson and colleagues have carried out a series of studies to test their model. Family resources, family coping, family stress and an accumulation of family life events have been found to be associated with compliance to treatment and with changes in the physical status of children with cystic fibrosis (Patterson 1985; Patterson et al 1983; Patterson et al 1990). In a study on the health status of 48 families providing home care for their medically fragile children it was found that the rate of family physical illness symptoms was associated with greater financial burden and with more strain with the professionals. Mothers, as the primary carers, experienced a greater decline in their physical health than did fathers; and the more hours of care provided by parents, the greater the negative impact of illness on the family (Patterson et al 1992).

In formulating my aims and hypotheses, I have used a theoretical framework that draws common themes from the different models assembled above, all of which derived from family systems approaches:
- The sick child is not independent from the family system;
- Family organisation (or re-organisation) take place around the illness;
- At times of crises, such as a 'diagnosis' in a family member, maladaptive patterns are common in healthy as well as in disturbed families;

- Adaptation to illness is multi-factorial, and the well being of the patient depends on the interaction between the illness typology, the individual, the family and other social systems (e.g. professionals);

- Family interactions, attitudes and emotions expressed towards the patient are not stable over time but fluctuate as the illness progresses, or as there are changes in the illness typology;

- The way family members respond to one another, including the patient, can have a beneficial or a detrimental effect, depending on whether the interactions or emotions are positive or negative;

- Interactions that are beneficial at one stage of the illness may have a detrimental effect at a different stage.

The methodological limitations of the studies on paediatric transplantation (see chapter 2) show a need for prospective research examining the effectiveness of transplantation surgery and its effect on the ill child and other family members.

Children referred for heart or heart-lung transplant assessments are dying. The acceptance for transplantation represents the entrance of the illness into its terminal phase, a confrontation with 'impending death'. Using John Rolland's model, this group of children presents a different psychosocial typology of illness than children with congenital heart disease who need to undergo conventional high or low-risk cardiac surgery (see FIGURE 1). Also, there are typological differences between children in need of heart and children in need of heart-lung transplantation. Although the outcome is fatal for all the children in these groups, some children have a life expectancy of less than 24 months whilst others have a longer life-expectancy. The level of incapacity is more severe in children with shorter life expectancy. Typological differences are also found between the groups of children undergoing conventional high and low-risk cardiac surgery (see FIGURE 1).

Wood and colleagues have pointed out that families of children with the most severe chronic disease activity are expected to be more disrupted and disrupting, and therefore it is necessary to take into account differences in disease severity when examining how families organised themselves around the illness (Wood et al 1989). Furthermore, family organisation is

Figure 1: Psychosocial Typology of Illness: Transplantation and Conventional Cardiac
  Surgery

a) Psychosocial Typology presented by Children referred for Transplantation

Illness Phase	Illness Type
Crisis, Chronic or Pre-Terminal	Onset: Acute or Gradual (Heart Tx) or Gradual (Heart-Lung Tx) Course: Progressive Outcome: Fatal Incapacitation: Moderate to Severe (active list) Mild to Moderate (provisional list)
	Family Elements

b) Psychosocial Typology presented by Children in need of High Risk Cardiac Surgery

Illness Phase	Illness Type
Crisis or Chronic	Onset: Acute or Gradual Course: Constant or Progressive Outcome: Shortened Life Span or Fatal Incapacitation: Mild to Severe
	Family Elements

c) Psychosocial Typology presented by Children in need of Low-risk Cardiac Surgery

Illness Phase	Illness Type
Crisis or Chronic	Onset: Acute or Gradual Course: Constant or Progressive Outcome: Non-fatal Incapacitation: None or Mild
	Family Elements

influenced by the daily demands of medical treatment (e.g. drugs, physiotherapy etc.) on family members, and by the family awareness of the stresses associated with child's prognosis (Patterson 1989).

The Hypotheses of the Study:

Based on these principles the following predictions were made:

1. Children referred for transplantation, and their parents, will demonstrate higher levels of adjustment problems than children admitted for high or low-risk conventional cardiac surgery, and their parents.

2. Families of children who are referred for transplantation assessment will demonstrate a different pattern of family organisation than families of children admitted for high or low risk conventional cardiac surgery. These will be manifested by higher levels of marital and family conflict and a higher rate of negative attitudes (hostility, number of criticisms and emotional overinvolvement) between family members.

3. Children's adjustment at entry into the study will be associated with parents' psychological and marital adjustment, and with family organisation.

4. As the children's health improves following surgery, changes would be expected at the individual and family levels. At 12 months post-surgery, children and parents in all groups would show better adjustment and better family organisation. These will be shown by lower levels of marital and family conflict and a lower rate of negative attitudes (less hostility, criticisms and emotional overinvolvement) between family members, than prior to surgery.

5. As the demands of daily treatment on the parents diminishes following transplantation, changes in the children's and parents'adjustment, and in the family organisation, will be greater in those families whose children received transplantation than in families whose children received conventional low or high risk cardiac surgery.

6. Poor children's adjustment, negative family organisation (shown by negative attitudes, poor marital and family adjustment) and poor parental adjustment will be associated with non-survival while awaiting transplantation.

7. Following transplantation or cardiac surgery, poor child's adjustment, negative family organisation (shown by negative attitudes, poor marital and family adjustment) and, poor parental adjustment will be associated with worse physical health than expected. Normal child's adjustment, positive family organisation (shown by positive attitudes, good marital and family adjustment) and, normal parental adjustment will be associated with better physical health than expected.

8. Whilst type of transplantation will not be associated with adherence levels, socio-economic status, having a chronically ill sibling and family composition will discriminate between levels of children's adherence. Higher rates of unsatisfactory adherence will be expected among children with a psychiatric disorder when compared to children with no psychiatric disorder. Higher rates of unsatisfactory adherence will be expected among children who are exposed to negative patterns of family organisation (negative attitudes between family members , poor marital and family adjustment) and to poor parental adjustment when compared with children whose families lack these characteristics.

3.2 METHODOLOGY OF THE STUDY

The research protocol used in the present study was integrated into the clinical transplant programme. For ethical reasons it was not always possible to maintain a strict boundary between the researcher and the clinical team. In case of concern about a specific child or family, the investigator passed the relevant clinical information to the clinical team. However, in order to help the investigator to keep her objectivity, members of the clinical team never fed back information to the investigator.

Subject Selection

Participation was sought from a consecutive series of parents whose child was referred to Great Ormond Street Hospital for heart (HTx) or heart-lung (HLTx) transplant assessment

between September 1988 and May 1993. Prior to the child's first admission to assess suitability for transplantation, a letter explaining the study, a consent-form and a booklet containing information regarding transplant assessment procedures were sent by post to the parents. During the child's admission, parents gave the consent-form back to the investigator who was available to clarify any queries and to move on with the assessment of those families willing to participate. The assessment procedure is discussed below.

Participation was also sought from a consecutive series of parents whose child was admitted to Great Ormond Street Hospital to undergo conventional cardiac surgery between September 1989 and November 1992. Prior to the child's admission for surgery, a letter explaining the study, a consent-form and a booklet with information regarding surgical and post-operative procedures were sent by post to the family. Families willing to take part returned the consent-form in a postage-paid envelope addressed to the investigator. The assessment procedure is discussed below.

Subjects referred for transplantation, or undergoing conventional cardiac surgery, were excluded from the study if:
  - The patient was 18 years or older at initial assessment;
  - The patient was unsuitable for transplantation, was brain damaged or had other chronic physical illness unrelated to the current surgical procedure;
  - Communication in English Language was difficult with the child and/or the parents, or if
  - The child was unable to attend the follow-up appointment.

Following acceptance into the transplant programme, those children whose life expectancy was of 24 months or less joined the active list for heart (HTx group) or heart-lung transplantation (HLTx group). If the children were suitable for transplantation but their life expectancy was more than 24 months, they joined the provisional waiting list (PVHL group).

Most children admitted to undergo conventional cardiac surgery were well known to the paediatric cardiologists who were following them up on an annual basis. Many of these children had undergone previous hospitalisations and/or palliative or corrective cardiac surgery at Great Ormond Street Hospital. Prior to admission, and in order to form two clinical subgroups, a cardiologist assessed the clinical picture of each child on the basis of previous

medical examinations, the case history and the type of surgery required. Children were then classified as undergoing either high (HR group) or low-risk (LR group) conventional cardiac surgery. High risk was defined as more than 5% risk (up to 30%) of operative mortality during hospital admission. Low-risk was defined as 5% or less risk of operative mortality during hospital admission. The children in the high risk group generally had cyanosis and physical disability, whilst the children in the low-risk group were generally non-symptomatic.

Total Number of Families invited to Participate

Figure 2: Total number of Families invited to participate in the study

398 Families were Invited

233 Families
did not participate

165 Families
participated
(47.5% of those eligible)

51 Families
were excluded

182 Families
were lost

127 unwilling
to participate   30 withdrew   25 were lost
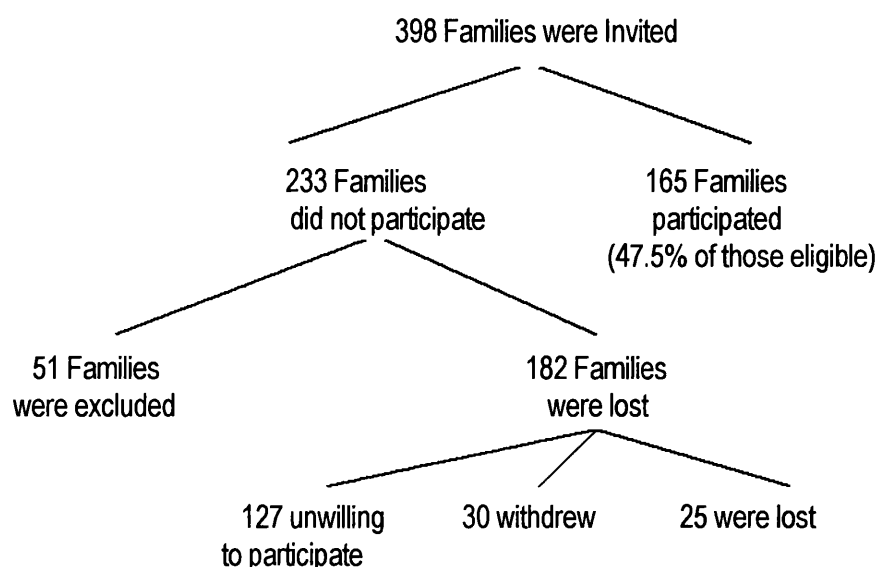
As shown in FIGURE 2, three hundred ninety eight families were invited to take part in the study. Of those, 165 participated.

A total of two hundred and thirty three families were not included: 51 were excluded after the first assessment because the child or parents did not fulfil the inclusion criteria. One hundred and eighty two families were either unwilling to participate, withdrew or were lost for various reasons. Parents unwilling to participate failed to reply to two letters asking for participation. Parents who withdrew either did not want their children to be interviewed due to their concerns about the content of the interview, said to be too tired, or too worried about the ill child and/or other family members left behind, wanting to be left alone. Ward closures, unexpected changes in the dates for surgery and difficulties co-ordinating physical and psychological assessments contributed to the loss of 25 families. Overall, the timing of the

psychological assessment was problematic, and at times inappropriate. Often there were big delays in X-rays and other hospital departments, the children were usually feeling tired and hungry following the medical investigations, and after catheterization children were asleep or somnolent for 12 hours.

FIGURE 3 shows that 106 of the 165 families participating contained a child who was suitable for transplantation (Tx). Of those, 38 children joined the active list for heart transplant (HTx group), and 68 children were accepted for heart-lung transplantation. In this latter group, 29 children joined the active list for heart-lung transplantation (HLTx group), and 39 children joined the provisional list for heart-lung transplantation (PvHL group). There were no children on the provisional list for heart transplantation.

Figure 3: Families participating in the study at Initial Assessment (T1)
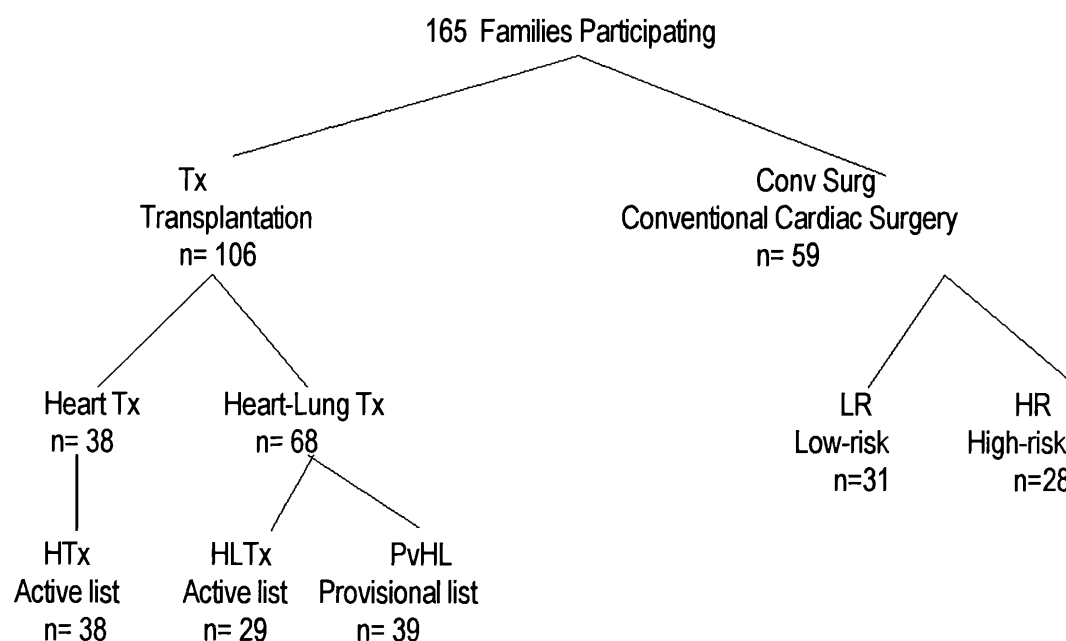
Fifty nine families contained a child who was in need of conventional cardiac surgery (Conv Surg). Of those, 31 acyanotic children were undergoing conventional low-risk cardiac surgery (LR group); and 28 cyanotic children were undergoing conventional high-risk cardiac surgery (HR group). FIGURES 4 and 5 show the children's diagnoses.

Figure 4: Diagnoses of Children assessed for Transplantation

Heart Transplantation	Heart-Lung Transplantation
Congenital Heart, n= 14 Cardiomyopathy, n= 20 Heart Tumour, n= 1 Hypoplastic left heart, n= 3	Cystic Fibrosis, n= 60 Eisenmenger, n= 7 Graft vs Host, n= 1

Figure 5: Diagnoses of Children undergoing Conventional Cardiac Surgery

Low-risk	High-risk
Atrial Septal Defect, n= 5, Aortic Valve Stenosis, n= 7, Coartation of the Aorta n= 3, Ventricular Septal Defect, n= 8, Persistent Ductus Arteriosis, n= 4, Supravalvar Aortic Stenosis, n= 3, Severe Aortic Incompetence, n= 1	Tetralogy of Fallot, n= 5, Pulmonary Atresian, n= 4, Tricuspic Atresian, n= 3, Fontan, n= 6, Severe Mitral Resurgitation, n= 3, Ebsteins Anomaly, n= 1, Transposition Great Arteries, n= 2, Double Imlet Left Ventricle, n = 3, Complex Congenital Disease, n= 1

Assessment Procedure and Instruments

In order to ascertain changes in the child's physical and psychological adjustment, as well as changes in parental and marital adjustment, family organisation and levels of expressed emotion, evaluations were carried out at two times: T1 and T2.

Different instruments and sources of information tend to identify disturbances in different children (Rutter et al 1970). For that reason, a combination of standardised self-report and observational measures, and multiple sources of information (such as parents, children and school-teachers) were used in the present study. At entry into the study (T1), all children underwent detailed physical investigation as in-patients. Regarding the psychological evaluation, children referred for transplant assessment and their families completed the questionnaires and were interviewed during a 4-5 day admission period to assess the child's suitability for transplantation. This assessment took place prior to the child's acceptance into the transplant programme. Children undergoing non-transplant cardiac surgery and their

The assessment procedure is presented schematically in FIGURE 6:

Independent Assessment

	Children's Physical Health	Children's Adjustment	Parents' Adjustment	Marital Adjustment and Family Organisation
Initial Assessment	T1	T1	T1	T1
Follow-up	T2	T2	T2	T2
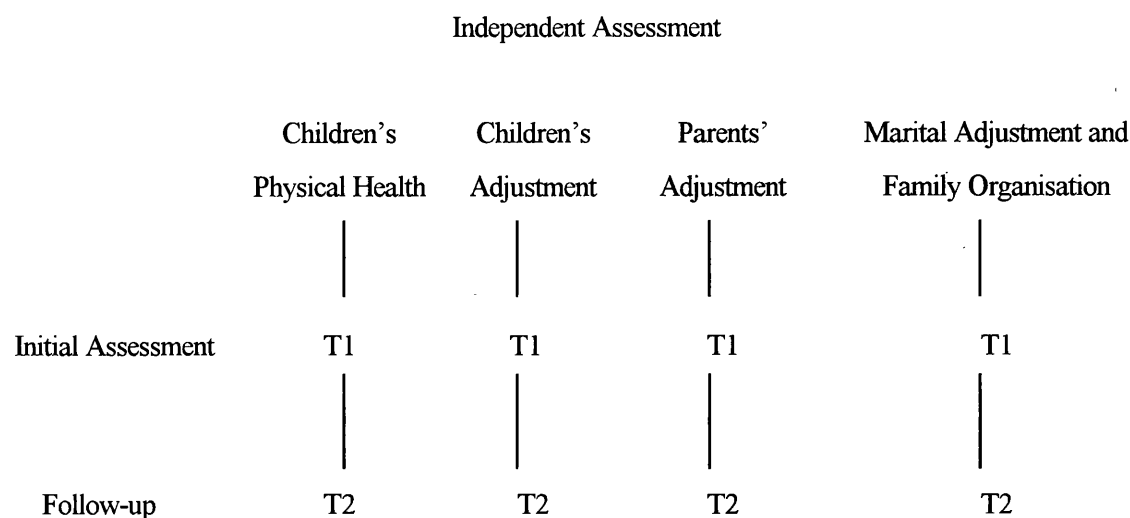
families received the questionnaires at home two to three weeks prior to the child's admission for surgery. Parents were requested to bring the completed questionnaires with them at the time of surgery and the interviews took place during the admission period, one-two days before the child's operation. Once medical and psychological evaluations were completed, a school report was sent to the school head teacher together with a postage-paid envelope, and a request to mail it back.

Transplant recipients and their families were followed up (T2) at a mean of 12 months post-operatively (range: 9 to 17 months), during a routine admission for a biopsy. This follow-up assessment took place at a mean of 22 months from the initial assessment (T1). Children on the provisional list (PvHL group) for transplantation and their families were also followed up at a mean of 22 months from the initial assessment (T1), during a planned admission to monitor changes in the child's physical health. The follow-up of children who received non-transplant cardiac surgery was conducted during a routine outpatient visit at 12 months post-operatively (mean 12 months; range 9 to 15 months).

The medical team obtained all medical investigations[1]. A child psychiatrist[2] or the investigator conducted the child psychiatric interviews. The investigator conducted the parental interviews. A student of psychology or the investigator administered and scored the questionnaires. The

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[1] Drs Phil Rees and Bruce Whitehead, and colleagues

consultant child psychiatrist, a trained Expressed Emotion (EE) rater [3], the transplant specialist nurse[4], and the liaison sister[5] conducted reliability tests with the investigator on several measures.

Child's Physical Health Measures

Prior to acceptance into the transplant programme all children underwent a detailed clinical assessment which included: percentage of ideal weight corrected for height (Tanner and Whitehouse 1986), routine blood investigations, chest x-rays, walking distance covered in twelve minutes, minimal oxygen saturation obtained during the exercise test, catheterization (heart transplant candidates) and pulmonary function testing (heart-lung transplant candidates).

Children undergoing non-transplant cardiac surgery underwent the following medical assessment: percentage of ideal weight corrected for height (Tanner and Whitehouse 1986), routine blood investigations, chest x-rays, walking distance covered in twelve minutes and catheterization.

Based on the above results, *a global rating of physical status* was made independently by the transplant consultant [6] and a consultant cardiologist [7] on a scale from 1 "well functioning" to 4 "critically ill and incapacitated" (Serrano-Ikkos et al 1997). The parental account of the child's *physical disability* was rated on a 6-point scale from 1 "no physical restrictions to 6 "needs to use a wheelchair inside the house". Again the higher the rating the more physically incapacitated was the child (see Appendix 1, page 266-267). This was rated by the interviewer during the parental interview. The teachers rated the child's *school attendance* on a scale from poor (1) to excellent (5) (Rutter 1967, Scale B (2)).

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[2] Dr Bryan Lask
[3] Mrs Rebecca Lewin
[4] Mrs Pauline Whitmore
[5] Mrs Mary Goodwin
[6] Dr Bruce Whitehead
[7] Dr Phil Rees

Child's Adjustment Measures

Child's Mental Status: Children, aged 5 or above, underwent a semi-structured psychiatric interview with a child psychiatrist or the investigator. This elicits information regarding the child's mental state (Rutter and Graham 1968). As well as information on psychiatric symptoms (such as worries, fears, unhappiness, irritability, tempers etc.), systematic data was collected on the child's feelings about the illness, difficulties with medical treatment, frequency of social contacts, peer relationships and hopes for the future. The ICD-9 classification (WHO 1978) was used to define any psychiatric disorder. Only data regarding the ICD-9 classification will be presented in this study. All psychiatric interviews were audio-recorded for subsequent reliability analysis.

Global Assessment of Functioning (GAF Scale DSM-III-R, 1987): This is a measure of overall psychosocial functioning. This global assessment scale condenses the total knowledge about psychiatric and social disturbances into a single clinically significant index. This scale allows comparison of children and adolescents over a wide age range. Ratings are made on a 0-90-point scale, with 0 indicating a child with the most severe psychosocial impairment and 90 indicating optimal level of functioning. Every tenth marker on the scale has an associated narrative that describes, in terms of function, the behaviour of the patient. A cut-off score of 70 was used in this study to distinguish normal from impaired functioning (0-50: severe impairment; 51-70: moderate impairment; 71-90: normal level of functioning).

A child psychiatrist and the investigator scored, independently of each other, the level of functioning of those children aged 5 or older. This was done from information a) collated during the psychiatric interview, b) provided by the parents, and c) obtained from standardised questionnaires completed by parents, school teacher, and the child if able to write and read independently. The following standardised instruments were used:

- *Children's Behaviour Questionnaire, Scale A(2) (Rutter et al 1970):* Parents were asked to complete this scale which provides valid and reliable screening measures of the child's behaviour at home. A score of 13 points or more indicates behavioural disturbance in a child; and sub-scales discriminate between neurotic, antisocial and mixed disorders.

- Children's Behaviour Questionnaire, Scale B(2) (Rutter 1967): School teachers were asked to complete this instrument which provides a valid and reliable screening measure of children's behaviour at school. A score of 9 or more indicates disturbance in the child; and subscales discriminate between neurotic, antisocial and mixed disorders.

- Children Self-Concept Scale (Piers and Harris 1984): Children above 7 years of age with adequate cognitive functioning completed this scale. It has 80 yes/no items and provides a self-description regarding physical appearance, social behaviour, academic status, dissatisfactions, depreciation and contentment with self. A high score indicates a more positive self-concept. This scale has been carefully developed and has been demonstrated to have high reliability and internal consistency. It is widely used with both healthy children and children with disabilities.

Parental and Marital Adjustment

The General Health Questionnaire (GHQ-28) (Goldberg 1978): This 28 item likert scale was completed by each parent. This instrument was designed to be a self-administered instrument aimed at detecting psychiatric disorders among respondents in community settings, such as primary care or among general medical outpatients. The questionnaire is easy to administer, acceptable to respondents and fairly short. It measures two major classes of phenomena: a) inability to continue to carry out normal healthy functions and b) the appearance of new phenomena of a distressing nature. A bimodal rating of each scale (0 0 1 1) is used to obtain an overall score and the higher the total scored the poorer the psychosocial functioning. A threshold score of 4/5 discriminates psychiatrically distressed cases from no psychiatric abnormality. This scale correlates highly with psychiatrists' rating of depression, anxiety and psychosomatic symptoms.

The Golombok Rust Inventory of Marital State (GRIMS) (Rust et al 1985): This inventory was designed for the assessment of the overall quality of the relationship between a man and a woman who are married or living together. It has 28 items, and the questions concern: interests shared, dependence/ independence, communication, warmth, love and hostility, decision making, trust and respect, coping with problems and crises, behaviour, attitudes and feelings about the relationship, and, commitment to a future together. It is used in research, either to assess the efficacy of different forms of therapy or to assess the impact of social,

psychological, medical or other factors on a relationship. A transformed score of 6 or more is recommended as a cut off point for identifying a poor relationship. The advantages of the GRIMS over other marital or relationship questionnaires are: a) the simplicity of administration, and, b) that it has been developed and standardised in the United Kingdom.

Family Organisation Measures

Attitudes towards the Child.-The Expressed Emotion Scales (EE). EE taps subjective attitudes and feelings (positive and negative) that a relative expresses in relation to a symptomatic member of the family and his/her illness during the process of a semi-structured interview, *The Camberwell Family Interview (CFI)* (Vaughn and Leff, 1976a;b). The CFI was derived from measures used to study the effects of psychiatric illness upon the family and the family relationships (Brown and Rutter 1966). Although shown to be a reliable and valid instrument, the original interview took as long as four or five hours to administer. This obstacle lead Vaughn and Leff (1976b) to develop an abbreviated version, The Camberwell Family Interview Schedule. EE is usually rated through the use of audio recordings of separate interviews with relatives of patients.

Five principal scales comprise this instrument: warmth (W), positive remarks (Pr), critical comments (CC), hostility (H), and emotional overinvolvement (EOI). Tone of voice, content of speech and gesture are used to assess the degree to which emotion is shown.

Warmth refers to enthusiasm, sympathy, concern and empathy shown by a relative when talking about the patient; and also about positive changes in manner and tone when the respondent switches from talking about neutral subjects to talking about the person. Warmth is measured on a 6 point global scale. A *positive remark* is a statement that expresses praise, approval or appreciation of the behaviour or personality of the person to whom it refers. This score is obtained by a frequency count of relevant positive remarks during the course of the interview. A *critical comment* is defined as a statement that by the manner in which it is expressed, constitutes an unfavourable comment upon the behaviour or personality of the person to whom it refers. This score is obtained by counting all such comments occurring at any point during the interview. *Hostility* is considered to be present when the patient is attacked for what he/she is rather than for what he/she does. It can be presented as a generalisation of criticism, as rejecting remarks, or both. Hostility is measured on a 4 point

global scale. A marked concern for the patient reflected in unusually self-sacrificing and devoted behaviour, extremely age-inappropriate overprotective behaviours, and exaggerated emotional responses are signs of the relative's *overinvolvement* with the patient. Emotional overinvolvement is measured on a 6 point global scale (Leff and Vaughn 1985).

A Pilot Study

Vaughn and Leff's abbreviated version of the CFI (1976b) was modified by the investigator to be used with the parents of children with cardio-respiratory disorders. The version used in the present study had previously been piloted in a sample of 20 families with a child admitted for transplant assessment or conventional cardiac surgery. Interviews from the pilot study were audiotaped and later transcribed for content and reliability analysis. Subsequently, in order to improve validity and reliability, a series of amendments were incorporated following discussions with Dr Vaughn. Each parent's interview was closely examined for systematic examination of text: identifying and grouping themes and coding, classifying, and developing categories.

As in the original CFI, the version used in this study aimed to obtain a) an account of the circumstances in the home in the 3 months preceding the patient's admission, b) details of the onset and the development of the illness, c) its impact on various aspects of family life, such as : change in family roles, participation in domestic tasks, the frequency of irritability and quarrelling, contact between the ill child and other family members, patient's social and academic life, and adherence to treatment. The modified version of the CFI used in this study is described in Appendix 1 (pages 259-276; 277-280). However, the following innovations in the content of the interview and in the EE methodology were introduced:

1. A section was added to the interview exploring: a) how much information the parents conveyed to the ill child, and to the siblings, regarding the hospital procedures, prognosis and the patient's life expectancy; b) to what extent parents protect the children from such threatening information; and c) who is responsible for different aspects of treatment.

2. Instead of interviewing the parents separately, parents were interviewed together for approximately 60 minutes. The rationale was, first of all, to gather information about how parents interact with each other and about family dynamics; secondly, to abbreviate as much as

possible the psychological assessment procedure in order to fit in with the timing of medical evaluation. Other authors have used similar methodology and obtained satisfactory results (Berkovitz 1987; Szmukler et al 1987; Le Grange 1989).

3. Parental attitudes to the child were rated separately. In order to facilitate these ratings, during the process of the interview the investigator had to ensure she obtained responses from both parents, not allowing one of them to dominate the conversation.

4. All interviews were audio-recorded and subsequently rated by two raters, a clinical psychologist and the author, both of who were EE trained. The procedure involved establishing criteria to rate parenting and discipline issues in families whose children suffer chronic and life-threatening conditions.

5. The clinical psychologist was blind to the child's severity of illness and rated the interviews in a consecutive order within 4 weeks following the interview. The research number given in the audio-tape was different at T1 and T2. This protocol was followed in order to help the blind EE rater to keep her objectivity by preventing her from scoring a specific group of relatives more or less favourably, and also to prevent her from identifying the family at T2, and from checking her own EE ratings at T1.

6. The investigator postponed the ratings until the data had been collected for all the families and for all the assessments. This protocol was followed to prevent the two raters from communicating about the ratings.

Attitudes towards the Partner.- Expressed Emotion between the Parents. Parental attitudes and feelings towards their partner were measured with The *Expressed Emotion Partner Interview Schedule (EEPIS).* This is a short semi-structured interview, devised by the investigator, which was also piloted with the same 20 couples who participated in the pilot CFI study (see Appendix 2, pages 281-283; 284-287). The interviews were audiotaped for subsequent content and for reliability analysis.

Following the CFI, parents were interviewed separately during an average of 15 minutes. The main topics covered by the EEPIS were: a) the amount of perceived practical and emotional

support given by the partner regarding the child's illness; b) satisfaction with the amount of affection shown by the partner; c) satisfaction with the partner's involvement in the child's care, and openness in the sharing of fears and painful feelings regarding the child's illness and life expectancy. All interviews were audiotaped for subsequent inter-rater reliability. From the information gathered during this interview, warmth, hostility, critical comments and positive remarks ratings were made using the criteria described in the EE scoring manual (Leff and Vaughan 1985).

Clinical Ratings on Family Adjustment: The modified version of the CFI and the EEPIS allowed information to be obtained regarding: the existence and severity of long-standing unresolved marital issues, coalitions (excluding) or rejection of family members by withdrawing emotional support or ignoring needs, long-standing blame of the other parent, unresolved long-standing difficulties and disagreement with regard to parental issues etc.

TABLE IX

FAMILY ADJUSTMENT-CLINICAL RATINGS

Well Adjusted Families	Moderately Adjusted Families	Poorly Adjusted Families
1.The relationship between family members is constructive and appropriate: mutual respect, support and affection.	1. Satisfactory relationships but with greater closeness/distance between some family members than others. Some deficiencies in support and/or working together.	1.Precarious marital relationship. Serious discord or distance between members or shifting alignments. Episodes of separation.
2. Parents share a set of goals, they work together as a team.	2. Not a strong sense of cohesive family unit / mutual respect.	2. Children detour parental tension/ conflict.
3. Parents show care, understanding and pay attention to the children.	3. Parents have minor or occasional problems relating to the children.	3. Parents show poor understanding of children.
4. Differences / conflicts are resolved constructively.	4. Crises highlight conflicts/ differences that are not always solved.	4. Crises leading to further disintegration.

This information permitted a broad rating of family adjustment on a 3 point scale: well adjusted, moderately adjusted, and poorly adjusted families (Schneiderman et al 1976), see TABLE IX. Inter-rater reliability test between the investigator and the clinical team ratings were conducted, see page 91.

Level of Adherence to Treatment Post-Transplantation

Following transplantation children need to adhere to a strict treatment regimen. This includes medication with cyclosporin A (CyA) (three times/day), Prednisolone (once/day) and Azathioprine (once/day), and the maintenance of a daily diary, which records doses of medication and the time taken. For heart-lung recipients peak flow measurements are also recorded. The completion of the medical diary is an important part of the regimen since it is helpful in identifying medical complications, and a failure to complete it regularly constitutes a degree of unsatisfactory adherence to the regimen.

Children are followed-up intensively by the transplant team during the first 6 months, and at monthly intervals thereafter. During the first 12 months post-transplantation, the children visit the hospital a minimum of 18 times. CyA levels are measured at each attendance. Those children who have received a heart-lung transplant also undergo respiratory function tests. The patients' diaries are checked for (i) medication dosage; (ii) peak flow measurement and (iii) daily completion. Any irregularities in the diary or unexplained lowering of CyA levels or unexpected discrepancies between the pulmonary function tests at home and in the laboratory (heart-lung transplants only), raise the possibility of incomplete adherence, which is then discussed with the children and their parents. Of all the medications, CyA is the most important to avoid organ rejection.

The criteria used for satisfactory/ unsatisfactory adherence to Cyclosporine A (CyA) and to the diary completion is as follows:

a) Satisfactory adherence to CyA if CyA levels were within 80-100% of the expected range whenever measured. Occasional levels below this range would be acceptable if there was a good clinical explanation. However, adherence was *unsatisfactory* if CyA levels were below 80-100% of expected range on more than one occasion, without a good clinical explanation.

b) Satisfactory adherence to diary completion if the diaries were completed in relation to lung function tests and medication 80% of or more of the time. However, adherence was *unsatisfactory* if the diaries were completed in relation to lung function tests and medication less than 80% of the time.

Based on this criterion, levels of adherence were determined as shown below.

TABLE X

LEVELS OF ADHERENCE TO TRANSPLANT TREATMENT

Good Adherence	Moderate Adherence	Poor Adherence
Both, CyA levels and diary completion were satisfactory	CyA levels were satisfactory, but diary completion was unsatisfactory	CyA levels were unsatisfactory regardless of diary completion

Reliability-Results: Child's Adjustment and Family Measures

Child's Mental Status (Rutter & Graham 1968).

All psychiatric interviews were audio-recorded and the child's mental status was rated (no abnormality/psychiatric illness) separately by a child psychiatrist (BL) and the author. Cohen's kappa for inter-rater reliability was 0.90. When disparity was found between the two raters consensus was reached by discussion.

Child's Global Assessment of Functioning (GAF Scale).

The scale has shown high inter-rater reliability, test-retest stability, and discriminant validity for children and adolescents (Shaffer et al 1983; Steinhausen 1987). The child psychiatrist and the investigator scored the Child's Global Assessment of Functioning independently. The mean score was calculated when the ratings between the two raters differed by ten points or less. If the difference between the raters was more than ten points consensus was reached by discussion. The intraclass correlation coefficient between the ratings was 0.73.

Attitudes towards the Child: The Expressed Emotion Scales (EE) (Camberwell Family Interview Schedule, CFI).

All interviews were audio recorded, transcribed and subsequently rated by two independent raters. The reliability between the raters' ratings was established. As EE ratings are on a quantitative scale, an analysis of variance (ANOVA) intraclass correlation approach was thought to be desirable (Bartko and Carpenter 1976; Dunn 1995, personal communication). The following coefficients were obtained regarding parental attitudes to the child:

Overinvolvement	Mother 0.91	Father 0.89
Warmth	Mother 0.93	Father 0.95
Hostility	Mother 0.95	Father 0.98;
Positive Remarks	Mother 0.88	Father 0.76
Critical Comments	Mother 0.97	Father 0.86

Prior to entering the scores in the computer, both raters discussed those ratings in which there was a disagreement, and consensus was reached by discussion.

During the pilot study conducted prior to the current research, the raters encountered some obstacles regarding the ratings of two scales, overinvolvement and warmth. This was discussed at length with an expert in the Expressed Emotion field, Dr. Christine Vaughn, and consensus about the criteria for rating these scales was reached prior to the collection of data of the present study. The difficulties were due to the following factors:

1) At the time of the first interviews, and in order to survive, many of the children had to be pampered and were completely dependent on their parents. Parents' own needs had to be 'on hold' in order to accommodate the child's needs. Sometimes it was difficult to assess to what extent parents' self-sacrificing behaviour was appropriate or exaggerated given the child's physical condition.

2) Some parents' emotional life was in a turmoil due to the fact that their children were in a pre-terminal state and they had to continue all their efforts to keep their child alive at the same time as having to come to terms with the likelihood of death in the face of progressive deterioration. This situation sometimes raised inner conflicts in the parents and ambivalence

towards the patient. At times, it was difficult to distinguish between parent's coping mechanisms, exaggerated emotional responses and their attitudes towards the child.

3) A significant number of these children were initially misdiagnosed and the parents' concerns about the child's health had not been taken seriously. At the time of the initial interview, parents had not ventilated their feelings of anger and frustration related to their earlier experiences of the child's illness, and those negative feelings were very vivid in their minds during the interviewing process.

4) Some parents felt ambivalence towards the demands of the child and his/her illness on them. This ambivalence manifested itself in a mixture of emotions such as overwhelming feelings of anger, denial, desperation and guilt about having given birth to an ill child and about having negative feelings regarding the child's condition and care. At the same time, parents had strong feelings of warmth, sympathy, care, and love. In addition, they were desperate to keep their disabled child alive. Understandably, parents had difficulties integrating all those feelings. Some parents coped by denying the existence of negative feelings and "idealising" the child. In these situations, it was difficult to distinguish between warmth and idealisation of the child. All these factors together made it difficult to rate warmth, and to judge to what extent parental devotion, self-sacrificing behaviour (putting the children's need ahead of their own), protectiveness and strong emotional responses were appropriate given the child's condition.

In order to rate overinvolvement, the patient's age and his/her degree of disability were taken into account. This applied to both the initial assessment and the follow-up. Individual parents' behaviours and emotions were judged against what was considered to be appropriate for the child's age, in the context of the child's physical status. For example, parental high involvement with a 15 years old (i.e. carrying him to the toilet, bathing him, feeding him, etc.) may be adaptive if the child is severely disabled and unable to do things by himself, but that same interaction style may become smothering after successful treatment, if the child is not disabled and has the ability to become more age-appropriate independent.

Attitudes towards the Partner (The Expressed Emotion Partner Interview Schedule EEPIS)):

As with the CFI, all interviews were audiotaped and scored by two raters. The following intraclass correlation coefficients between the two raters were obtained regarding parental attitudes to their partner:

Warmth	Mother 0.96	Father 0.94
Hostility	Mother 0.96	Father ; 1.00
Positive Remarks	Mother 0.81	Father 0.96
Critical Comments	Mother 0.91	Father 0.95

The high level of agreement across the raters, and the discrimination of these scales between well and moderate-poorly adjusted families (see TABLE XXV, page 115) at T1 are evidence of the concurrent validity of this new instrument. Furthermore, significant associations were found between warmth, critical comments and positive remark ratings at T1 and marital adjustment (GRIMS) at T1.

- Warmth :   mothers: $r=-.62$, $p<.001$;   fathers: $r=-.71$, $p<.001$
- Critical Comments : mothers: $r=.35$, $p<.001$;   fathers: $r=.27$, $p<.001$
- Positive Remarks: mothers: $r=-.24$, $p=.01$;   fathers: $r=.31$, $p=.001$

In order to explore whether these scales were predictors of future marital breakdown (predictive validity) a series of X^2 were conducted between the EE variables at T1 and the marital scale at T2. The variables were categorised as follows: Warmth: none-some/moderate-high; Positive remarks: present/absent; Critical comments: present/absent; Marital relationship: very good-average/poor-severe problems. Results showed that the scales at T1 discriminate between good and poor marital relationship (GRIMS) at T2:

- Warmth: mothers: $X^2=13.1$, $df=2$, $p=.001$;
                      fathers: $X^2=12.2$, $df=2$, $p=.002$,
- Positive Remarks: mothers: $X^2=5.3$, $df=1$, $p=.02$,
- Critical Comments: mothers: $X^2=4.4$, $df=1$, $p=.03$

Clinical Ratings on Family Adjustment (Schneiderman categories):

Using a clinical protocol, as part of the transplant programme routine assessment, families of children who were referred for transplantation were also evaluated by a child psychiatrist, nurse specialist, liaison sister or social worker. Following their assessment, the clinicians rated family adjustment using the same global scale as the investigator. The agreement between the clinicians and the investigator on this rating was 0.91 (intraclass correlations).

Analyses of Data

The statistical package for the social sciences (SPSS) programmes were utilised on an IBM computer system. Although non-parametric statistical tests have been used, parametric statistical methods were mainly used for the analysis as these are more powerful and can take account of more complex data structures. The data have been analysed as follows:

i) Differences in demographic characteristics, family organisation, children's and parents' adjustment between the groups presenting different psychosocial typology of illness (hypotheses 1 and 2):

A series of descriptive statistics, Chi-square tests and One-way analyses of variance (using an a priori contrast method), were conducted to identify differences in the demographic and psychosocial characteristics between the following five specified group combinations (see FIGURE 7) - in all contrasts children and family characteristics were compared.

With multiple tests of significance, the probability of making at least one type I error is higher than 1 in 20 (Fleiss 1986). To overcome the problem, the p value corresponding to the required level of significance can be lowered using the Bonferroni multiple comparison adjustment. Because this study is exploratory, attempts to control for such an error were not made; this means that extra caution is needed in interpreting the results.

Figure 7: Specified Group Comparisons
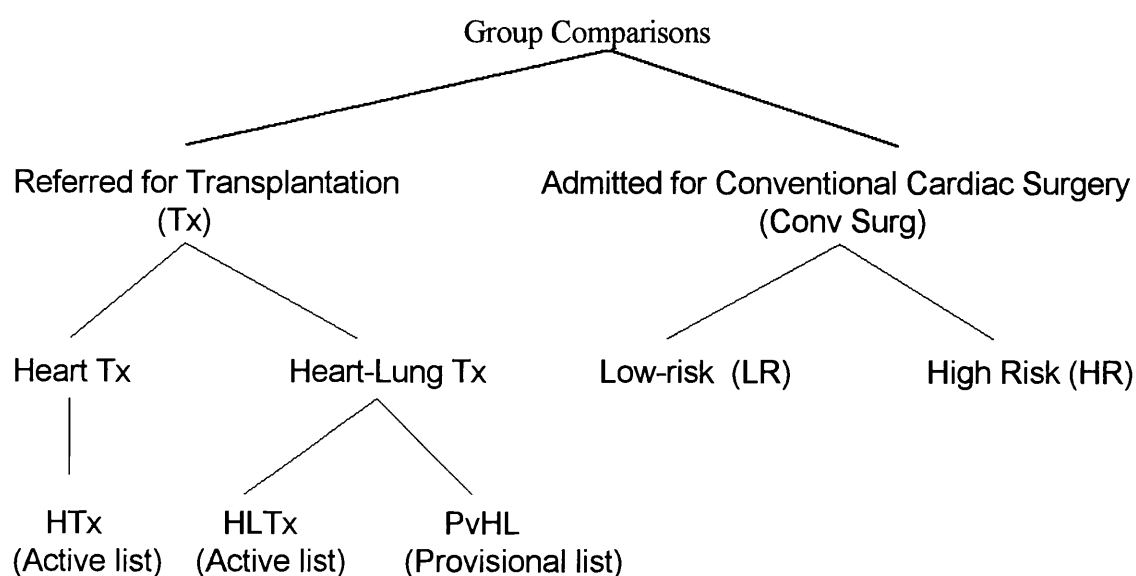
Contrast 1: Assessed for transplantation (Tx) Vs admitted for conventional cardiac surgery
(Conv. Surg).

Contrast 2: Heart transplant active list (HTx) Vs Heart-lung transplantation active and
provisional list (HLTx + PvHL).

Contrast 3: Heart transplantation active list (Htx) Vs Heart-lung transplantation active list
(HLTx).

Contrast 4: Heart-lung transplantation active list (HLTx) Vs Heart-lung transplantation
provisional list (PvHL).

Contrast 5: Conventional high-risk cardiac surgery (HR) Vs Conventional low-risk cardiac
surgery (LR).

Four of the five a priori planned contrasts are orthogonal (independent) and no adjustment for
multiple comparisons is required. A series of two tailed t-test or paired t-tests were performed
comparing mothers' and fathers' mean scores. In addition, a serie of McNemar tests and Chi
square tests were performed comparing the proportion of cases scoring above threshold
scores. Associations and intercorrelations between mothers' and fathers' expressed emotion
ratings were calculated using a series of Pearson correlation coefficients.

Finally, a series of t-test and McNemar tests were conducted to identify significant associations
between family adjustment and the expressed emotion scales.

ii) *Associations between Child Adjustment, Parental and Family Measures (hypothesis 3):*

A series of t-tests (two tailed probability), Chi-square tests, McNemar tests and one-way
analyses of variance (using the Scheffe multiple comparison adjustment method), were
conducted to compare differences between children with and without psychiatric disorder, and
between children with different levels of severity in their psychosocial functioning.

iii) *Changes in Children's Physical Status and in Psychological Measures following Surgery (hypotheses 4 and 5):*

A series of paired t-tests was conducted comparing the child's physical and psychological
mean scores measured at initial assessment and at the follow-up. The same procedure was
repeated for the family measures. In addition, a series of McNemar tests were carried out to
examine changes in the proportion of children with a psychiatric disorder between initial

assessment and the follow-up. The same procedure was used to examine changes in family adjustment, and in the number of parents scoring as psychiatrically distressed cases or as having serious marital difficulties.

New variables were created by subtracting the score at initial assessment from the score at the follow-up. This procedure was conducted for each one of the physical and psychological measures. To examine whether the degree and direction of 'change' in the physical and psychological measures was different in specified group combinations, a one-way analysis of variance with an a priori contrast was performed. The groups were compared as follows:

Contrast 1: Received transplant surgery (HTx + HLTx) Vs Received conventional
cardiac surgery (HR + LR).

Contrast 2: Heart transplant recipients (HTx) Vs Heart-lung transplant recipients (HLTx).

Contrast 3: Received low-risk conventional cardiac surgery (LR) Vs Received high risk
conventional surgery (HR).

Finally, an analysis was conducted to see whether changes in the marital relationship of the parents were associated with changes in the child's physical health. The GRIMS score at T1 was subtracted from the GRIMS score at T2 to obtain a GRIMS change score (a large negative score representing a large improvement; a large positive representing a large deterioration). This was done separately for the mothers and fathers. The same procedure was repeated with the child's health measures (physical status, disability and weight for height). Pearson correlations were conducted to relate changes in parental marital relationship with changes in the child's physical health.

iv) Associations between Psychosocial Factors and Medical Outcome (hypotheses 6 & 7):

In the first part of this section, a series of standard t-tests and Chi-square tests were conducted to identify physical and psychosocial differences between survivors and non-survivors while awaiting transplantation.

In the second part of this section, associations between psychosocial factors and the children being more or less healthy than expected at the follow-up, controlling for the children's health

at T1, were examined. Any improvement in the child's physical condition would be shown by a reduction in their scores on the physicians' and parents' ratings, and by an increase in their weight for height since prior to surgery most of the children were below normal weight. In order to avoid multiple tests of significance a composite index of physical health was derived from the cardiologist's ratings (physical status), the physical disability ratings and the weight for height. This was obtained from the first principal component for these three measures prior to surgery (T1, 1prefc) and after surgery (T2, 1 postfc). Taking the first principal components (Eigenvalue at T1: 1.83; at T2: 2.04) I then performed a varimax rotation and Kaiser normalization to obtain the factor loadings which are presented in TABLE XI. The factor loadings for the measures were very similar before and after surgery.

TABLE XI

INDEX OF PHYSICAL HEALTH: LOADING AND COMMUNALITY

	Pre-surgery		Post-surgery	
	Loading	Communality	Loading	Communality
Physical Status	.90	.81	.90	.82
Physical Disability	.88	.78	.88	.78
Weight for height	-.47	.22	-.66	.43
% variance explained	61.2		68.0	

The ideal weight corrected for height loadings are negative because a larger value indicates good health, whilst the other ratings are scored in the opposite direction. Communality refers to the proportion of the deviation in each one of the physical measures, which is explained by variability in a common factor. It is a measure of how well the factor score reflected each of the individual measures. The commonality is lower for weight for height. Genetic and nutritional factors as well as the child's physical health influence this measure.

In order to establish whether the children's post-surgery physical health (T2) was different from that expected from the children's pre-surgery physical health (T1), a regression analysis was performed between the children's composite index of physical health at T2 - dependent variable- and the children's composite index of physical health at T1. The equation obtained was: T2: .0491 + .217 xT1) See FIGURE 8

Figure 8: Regression Analysis : Expected Physical Health
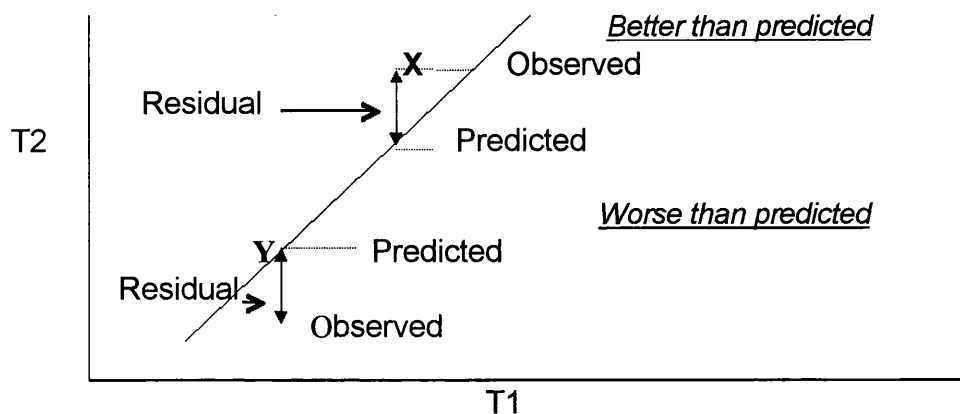
Subsequently, a new variable was created (chfc1) from the standardised (mean=0; standard desviation=1) residuals (the difference between the observed value at T2 and the value predicted from the regression). A series of correlations was then conducted between this new variable (chfc1) and the children's and families' characteristics. Due to the problem with interpreting multiple test of significance, there is a need for extra caution in interpreting these results.

v) Associations between Psychosocial Factors and Adherence to Medical Treatment following Transplantation (hypothesis 8):

A series of standard Chi-square tests was conducted in order to explore associations between the demographic and psychological characteristics at initial assessment and the children's adherence to treatment at 12 months post-transplantation.

CHAPTER 4

PRESENTATION OF THE RESULTS

4.1 CHILDREN'S AND FAMILIES' CHARACTERISTICS

4.1.1 Demographic Profile

The children and families' demographic characteristics at initial assessment (T1) are presented in TABLES XII and XIII. A series of one-way analyses of variance using a priori contrasts and Chi-square tests was conducted to identify differences in the children and family demographic characteristics between the 5 specified group combinations (see chapter 3, page 86). There were differences in sex ratio between the groups (X^2=14.4, df=4, p=0.006): the HTx group contained more males than the PvHL group; and the PvHL group contained a greater number of females than the other groups.

TABLE XII

CHILDREN'S DEMOGRAPHIC CHARACTERISTICS

	HTx	HLTx	PvHL	HR	LR
Gender	26 M	16 M	11 M	17 M	14 M
	12 F	13 F	28 F	11 F	17 F
Mean Age (s.d)	8.2(5.4)	11.6(3.6)	10.5(4.4)	8.8(3.6)	9.1(3.9)
Age Range	0.01-17.1	3.6-16.11	0.9-17.3	3.0-17.9	2.0-17
0-4.11 yrs	12(31.6%)	1(3.4%)	5(12.8%)	2(7.1%)	4(12.9%)
5-10.11 yrs	9(23.7%)	11(37.9%)	10(25.6%)	16(57.1%)	16(51.6%)
11-17.9 yrs	17(44.7%)	16(58.6%)	24(61.5%)	10(35.7%)	11(35.5%)
Mean Age at Diagnosis (s.d)	3.1 (3.9)	1.7 (1.1)	1.5 (1.0)	1.5 (1.6)	2.3 (2.8)
< Previous Surgery	12(31.5%)	12(41.3%)	15(38.5%)	26(92.9%)	5(16.1%)

HTx = Active list for heart transplantation   HR =High-risk cardiac surgery
HLTx = Active list for heart-lung transplantation   LR =Low-risk cardiac surgery
PvHL = Provisional list for heart-lung transplantation

Age differences were also found (F=3.14; df=4,165, p=0.01). The children on the active list for heart transplant (HTx) were significantly younger than the children on the active and provisional list (HLTx and PvHL together) for heart-lung transplantation (p=0.002).

Also, the children in the HTx group were significantly younger than the children in the HLTx group (p=0.002). There were no significant age differences when all the children assessed for transplantation (Tx group) were compared with all children undergoing conventional cardiac surgery (OFCS), nor between the children undergoing conventional high (HR) or low-risk cardiac surgery (LR).

Differences between the groups were also found regarding the number of previous operations (X^2=42.9, df=4, p<0.001). Children on the active list for heart transplantation (HTx) had fewer operations than the children assessed for heart-lung transplantation (HLTx and PvHL groups together). Nearly all children (92.8%) in the high-risk cardiac surgery group had previously undergone palliative or corrective cardiac surgery, whilst only 5 children (16.1%) in the low-risk group had experienced previous surgery. Most children were diagnosed as having a chronic condition during their first three years of life, but differences between the groups were found regarding child's age at diagnosis (F=2.93, df=4,160, p=0.02).

Children accepted for heart transplantation were older (mean age: 3.1 years) at the time of diagnosis than children accepted for heart-lung transplantation (mean age: 1.6 years) (p=0.01). Differences between the groups were also found regarding whether the child's illness was genetic or acquired (X^2=38.3, df=4, p< 0.001). All children in the conventional cardiac surgery groups (OFCS) and most children who were assessed for heart-lung transplantation had a congenital illness, whilst 13 children (34.2%) on the active list for heart transplantation (HTx) were healthy at birth and had only recently become ill, mainly with cardiomyopathies.

All children were Caucasian with the exception of three children who were referred for transplantation and two children in the conventional cardiac surgery group. All families were living within the U.K. or Ireland. Eighty percent of the families whose children were assessed for transplantation and 69% of the families whose children underwent conventional cardiac surgery had to travel a long distance to the hospital. All parents were accommodated in the hospital premises. TABLE XIII shows that a smaller number of children who were assessed for transplantation (n=75, 70.7%; HTx, HLTx and PvHL groups) were living in families with their married natural parents when compared to the children in the conventional cardiac surgery groups (n= 55, 93.2%, HR and LR) (X^2=11.7, df=4, p=0.01). No other differences between the groups were no found, except that greater number of children in the HLTx and PvHL groups had previously lost a sibling when compared to the other groups (X^2=9.5, df=4, p=0.03).

Differences between the groups regarding social class or employment status were not found.

TABLE XIII
FAMILIES' DEMOGRAPHIC CHARACTERISTICS

	HTx	HLTx	PvHL	HR	LR
Family Composition					
Natural Parents	27(71.0%)	21(72.4%)	27(69.2%)	27(96.4%)	28(90.3%)
Single Parent	3(7.8%)	2(6.9%)	6(15.4%)	1(3.6%)	2(6.5%)
Reconstituted	8(21.0%)	6(20.6%)	6(15.4%)	-	1(3.2%)
No. Children					
Range	1-4	1-5	1-6	1-5	1-5
Mean, (sd)	2.0(0.7)	2.6(1.0)	2.4(0.9)	2.5(1.0)	2.4(0.8)
No. Families with other Chronically ill Sibling/s	5 (13.1%)	7 (24.1%)	8 (20.5%)	4 (14.3%)	3 (9.7%)
No. Families who had lost a Child	3 (7.9%)	6 (20.7%)	5 (12.8%)	1 (3.6%)	2 (6.5%)
Social Class ✦*					
I	4(10.5%)	2(6.9%)	1(2.6%)	3(10.7%)	2(6.5%)
II	4(10.5%)	9(31.0%)	11(28.2%)	9(32.1%)	8(25.8%)
III	11(28.9%)	10(34.4%)	9(23.0%)	5(17.8%)	8(25.8%)
IV	14(36.8%)	8(27.6%)	14(35.9%)	10(35.7%)	13(41.9%)
V	5(13.2%)	-	4(10.3%)	1(3.6%)	-
Unemployed or other	4(10.5%)	4(13.8%)	7(17.9%)	2(7.1%)	5(16.1%)

HTx = Active list for heart transplantation   HR =High-risk cardiac surgery
HLTx = Active list for heart-lung transplantation   LR =Low-risk cardiac surgery
PvHL = Provisional list for heart-lung transplantation
✦Registrar General's Classification of Occupations and Coding Index (1980)
*Groups I & II, and groups III, IV & V were joined together for this analysis

4.1.2 Children's Physical Health Profile

The mean scores of the children's physical health measures are shown in TABLE XIV. Data were unavailable when children were too young to show physical disability (babies, toddlers) or when the child was too young to attend school. Children who were assessed for transplantation were rated by the physicians, parents and teachers as being more severely ill, having more physical disability, and having worse school attendance than the children who were in the conventional surgery group. Children who were assessed for transplantation also had a significantly lower percentage of weight for height.

TABLE XIV
CHILDREN'S PHYSICAL HEALTH MEASURES
n, Mean (s.d.)

	Transplantation	Cardiac Surgery	Sign of Diff[*]
Physical Status	n= 106, 3.6 (0.6)	n= 59, 2.0 (0.9)	0.0001
Weight for Height	n= 104, 84.9 (12.4)	n= 59, 90.3 (11.6)	0.0003
Physical Disability	n= 99, 3.7 (0.9)	n= 59, 2.0 (1.0)	0.0001
School Attendance	n= 86, 1.5 (1.2)	n= 55, 3.6 (1.0)	0.0001

* One-way analysis of variance, a priori contrast method.
Physical Status: 1= good functioning, 4= critically ill
Physical Disability: 1= no disability,   6= needs a wheelchair
School Attendance: 1= poor,   5 = excellent
Percentage of ideal Weight corrected for Height

TABLE XV shows the groups mean scores. Differences between the specified group combinations were found regarding the children's physical status (F=87.0, df=4,160, p=0.0001), percentage of weight for height (F=4.9, df=4,158, p=0.0008), physical disability (F=46.3, df=4,153, p=0.0001) and school attendance (F=32.2, df=4,136, p=0.0001). Children assessed for heart-lung transplantation (HLTx and PvHL groups together) had lower percentage of ideal weight corrected for height (p=0.05), and slightly better school attendance (p=0.05), than the children assessed for heart transplant (HTx). The weight for height of the children on the active list for heart-lung transplantation (HLTx group) was also significantly lower than the weight for height of the children on the active list for heart transplantation (HTx group) (p=0.004). Comparing the children on the active list (HLTx) with the children on the provisional list (PvHL) for heart-lung transplantation, it was found that the children on the HLTx group were more ill (p=0.0001) and had worse school attendance (p=0.004).

TABLE XV
CHILDREN'S PHYSICAL HEALTH MEASURES- Subgroups
Mean (s.d.)

	Heart Tx (HTx)	Heart-Lung Tx (HLTx)	Awaiting Tx (PvHL)	High-risk (HR)	Low-risk (LR)
Physical Status (Physicians' rating)	3.7 (0.5)	3.8 (0.3)	3.2 (0.8)	2.8 (0.5)	1.3 (0.6)
Weight for Height	87.5 (11.9)	78.9 (12.9)	86.8 (11.2)	88.3 (13.8)	92.1 (9.0)
Physical Disability (Parents' rating)	3.7 (1.0)	3.9 (0.7)	3.5 (1.0)	2.8 (0.8)	1.3 (0.4)
School Attendance (Teachers' rating)	1.1 (1.1)	1.2 (0.9)	2.0 (1.2)	3.3 (1.2)	3.8 (0.8)

* One-way analysis of variance, a priori contrast method.
Physical Status: 1 = good functioning; 4 = critically ill
Physical Disability: 1 = no disability;   6 = needs a wheelchair inside and outside
School Attendance: 1 = poor;   5 = excellent
Percentage of ideal Weight corrected for Height

The children in the first group also had lower percentage weight for height than the children in the PvHL group (p=0.007). With regard to the two groups of children undergoing conventional cardiac surgery, it was also found that children undergoing high-risk (HR) surgery were more ill (p=0.0001) and more physically disabled (p=0.0001) than the children undergoing low-risk surgery (LR).

4.1.3 Children's Adjustment Profile

The Children's Mental Status (ICD-9):

Prior to the child's admission, psychological problems had been detected in 5 children, requiring a professional consultation. However, none of the children was receiving long-term psychological treatment. Only children aged 5 years and older underwent the psychiatric interview. Therefore, data on the child's mental status and global assessment of functioning was available on 142 children. Overall, an average of 24% of the children had a psychiatric diagnosis. TABLE XVI shows the distribution of the children's mental status (ICD-9 Classification) in the group of children assessed for transplantation and in the group of children in need of conventional cardiac surgery.

TABLE XVI

CHILDREN'S MENTAL STATUS (ICD-9 Classification)

	Transplantation (n=89)	Conventional Surgery (n=53)
No Abnormality	69 (77.5%)	39 (73.5%)
Neurotic Disorder	8 (8.9%)	1 (1.1%)
Adjustment Reaction	1 (1.1%)	2 (3.7%)
Disturbance of Emotions	11 (12.3%)	10 (18.8%)
Disturbance of Conduct	-	1 (1.8%)

There were no differences in the number of children with psychiatric disturbance when the group of children assessed for transplantation was compared to the group of children in need of conventional cardiac surgery (X^2=0.28, df=1, p=0.59, NS). No other differences were found when the other specified group combinations were examined.

Children's Global Assessment of Functioning (GAF Scale)

A total of 54 children (38%) had no psychosocial impairment; 80 children (56.4%) had mild to moderate impairment; and 8 children (5.6%) were severely impaired in their psychosocial functioning.

Psychosocial functioning was not associated with psychosocial typology of illness as there were no significant differences between the mean scores of children assessed for transplantation and children undergoing conventional cardiac surgery (p=0.22), or between cyanotic (HR) and acyanotic children (LR) undergoing conventional cardiac surgery (p=0.35). However, children on the active list for heart-lung transplantation (HLTx) showed worse psychosocial functioning than the children on the provisional list (PvHL) for heart-lung transplantation (F=2.5, df=4,139, p=0.008).

TABLE XVII

SURGICAL OPERATION STATUS ♠ AND

CHILDREN'S GLOBAL ASSESSMENT OF FUNCTIONING (GAF Scale)

	N	Mean (s.d)	Scores Range
HTx	27	66.0 (11.0)	47 - 90
HLTx	29	64.4 (8.1) *	50 - 80
PvHL	35	71.8 (12.7) *	50 - 90
HR	26	68.4 (10.2)	55 - 85
LR	27	71.2 (11.9)	40 - 85

♠Relates to Psychosocial Typology of Illness, see page 72.
Oneway analysis of variance, a priory contrast method
GAF Scale: 71-90= no impairment; 51-70= mild-moderate impairment; 20-50= severe impairment
*Groups statistically different

TABLE XVII shows that the mean scores for children in the HTx, HLTx and HR groups indicated 'mild to moderate symptoms or some difficulty in several areas of functioning (school, social life etc), whilst the mean scores of children in the PvHL and LR groups are within the lower no impairment range, indicating no psychosocial impairment. Differences between the groups with regard to the number of children scoring above/below the threshold

score (70/71) were also examined. No differences were found regarding the proportion of children showing mild to severe impairment between the group of children assessed for transplantation and the group of children undergoing conventional cardiac surgery (X^2= 2.6, df=2, p=0.2). Neither were there differences between any other specified group comparison, including children on the active and provisional waiting lists for transplantation (surgery (X^2=5.4, df=2, p=0.06). FIGURE 9 shows the proportion of children with psychosocial impairment in each group.

Figure 9: Psychosocial Typology of Illness and Children's Global Assessment of
Functioning (GAF Scale)

%
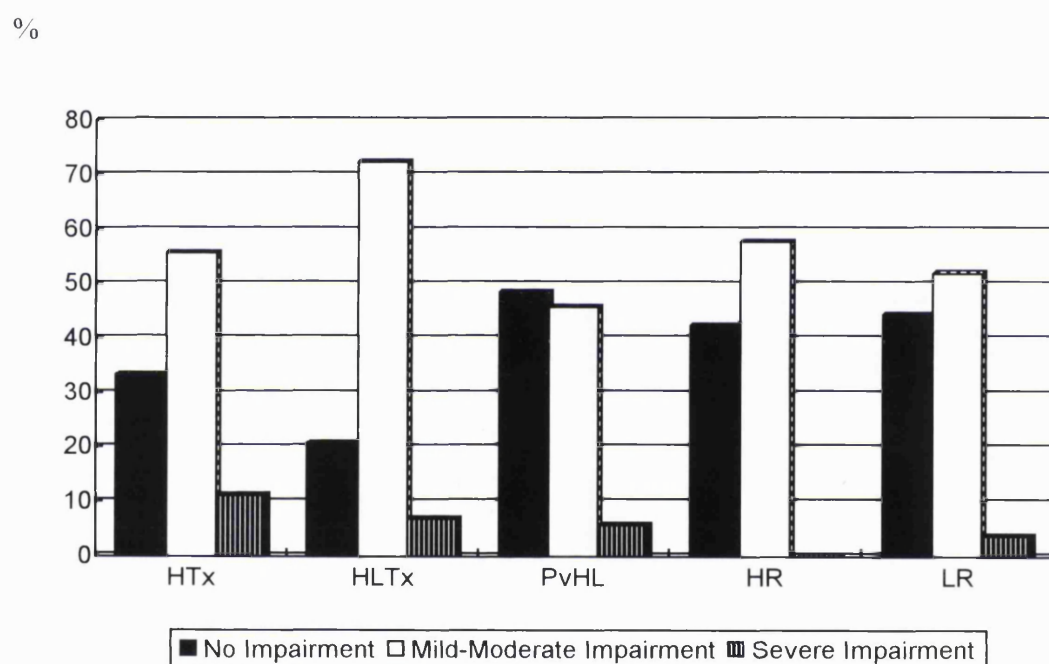
4.1.4 Parental and Marital Adjustment Profile

Parents' Psychological Adjustment (GHQ-28)

A total of 136 mothers and 126 fathers completed the GHQ-28 scale. Overall, mothers showed poorer functioning than fathers (Paired t-test, n=126; mothers' mean:5.1 s.d.:5.0; fathers' mean: 3.7, s.d.:4.2; t=2.9, p=0.004).

A summary of the parents' psychological adjustment is shown in TABLE XVIII. With the exception of the mothers whose children were on the provisional list for heart-lung (HLTx) transplantation, maternal mean scores were on or above the threshold score of 4/5 which

is indicative of psychiatric distress. The mean scores of the fathers in the heart transplant (HTx) and high-risk cardiac surgery (HR) groups were also on or above the threshold score. Nevertheless, differences in mean scores were not found between any of the specified group combinations (One-way anova, mothers: F=1.2, df=4,133, p=0.3; fathers: F=1.2, df=4,122, p=0.2).

The number of parents scoring as psychiatric cases was also examined. It was found that 40 to 64% of the mothers and 36 to 44.5% of the fathers rated above the threshold score (see TABLE XVIII). The differences in the number of mothers scoring as psychiatrically distressed between the transplantation and the conventional cardiac surgery groups were not statistically significant (mothers: X^2=4.4, df=1, p=0.5), but a higher number of fathers in the conventional cardiac surgery group rated above the threshold score when compared to the transplantation group (fathers: X^2=4.0, df=1, p=0.4). There were no other differences between the specified group comparisons with regard to the number of parents scoring on or above the threshold score on this scale. Permission was requested from these parents to inform their family doctor.

Marital Adjustment (GRIMS)

A total of 124 mothers and 125 fathers completed the GRIMS scale. TABLE XIX shows that the mean scores of both mothers and fathers indicated above average relationships, and there were no statistical differences between the mothers' and fathers' mean scores (Paired t-test, mothers' mean: 3.1, fathers' mean: 3.6, t=1.0, p=0.28, NS).

Differences were not found when comparing the mean scores of parents of children assessed for transplantation to those of parents of children undergoing conventional cardiac surgery. Nor there were significant differences between the other specified group combinations (mothers: F=0.41, df=4,119, p=0.79; fathers: F=0.85, df=4,120, p=0.49). No differences were found when parents of children assessed for transplantation were compared to parents of children undergoing cardiac surgery with regard to the number of parents rating above the threshold score (mothers: X^2= 0.3, df=1, p=0.5; fathers: X^2=2.6, df=1, p=0.1), or between the other specified group comparisons.

TABLE XIX

PARENTS MARITAL ADJUSTMENT (GRIMS)

N, Mean, (s.d.)

	Mothers'	Fathers'
HTx	n= 22 3.8 (2.1)	n=22 3.4 (2.0)
HLTx	n=21 4.2 (2.5)	n=21 4.4 (2.4)
PvHL	n=23 3.7 (2.5)	n=23 3.6 (2.1)
HR	n=27 3.8 (2.3)	n=27 3.5 (2.3)
LR	n=31 3.4 (2.2)	n=31 3.4 (2.2)

Paired T-test.

GRIMS scores:1-3= very good-good; 4= above average; 5= average; 6= poor; 7-9= severe problems.

Figure 10: Psychosocial Typology of Illness and Mothers' Marital Adjustment

%

■Very good- Average marital relationship □ Poor-Severe marital problems
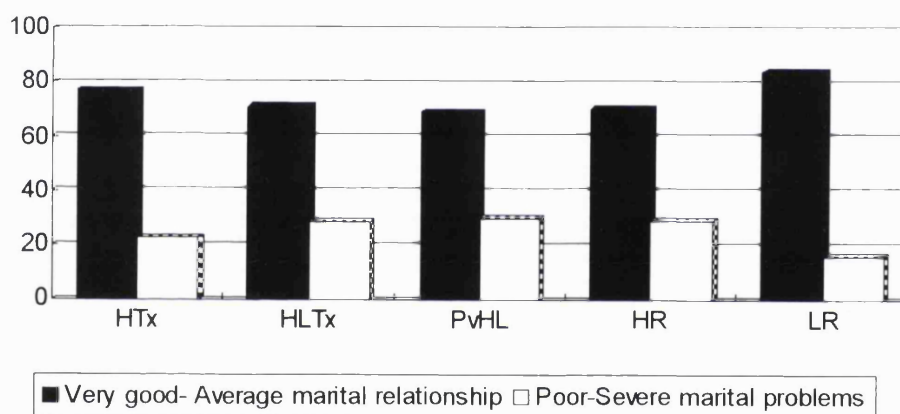
Figure 11: Psychosocial Typology of Illness and Father's Marital Adjustment

%

■ Very good- Average marital relationship □ Poor-Severe marital problems
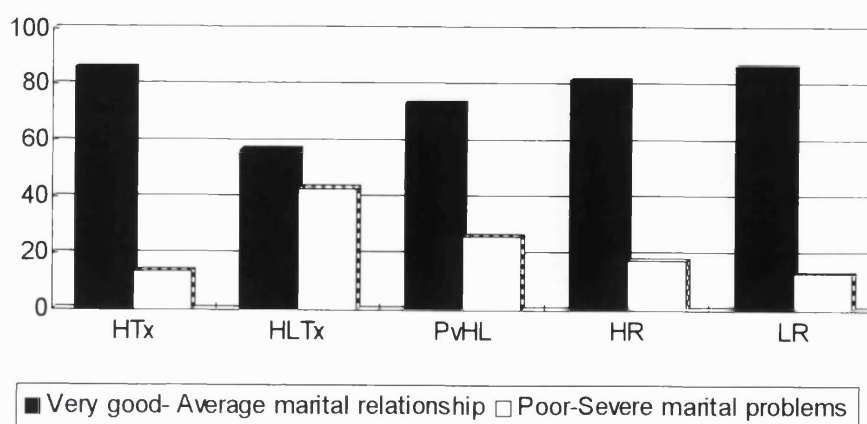
4.1.5 Family Organisation Profile

Attitudes towards the Child. The Expressed Emotion Scales (EE)

Overall, the parents' emotional profile regarding the child showed low levels of overinvolvement, high levels of warmth, very low hostility, low positive remarks and a virtual absence of criticism. See TABLE XX and Appendix 3.1 (page 288). As shown in TABLE XX, mothers rated higher than fathers on the emotional overinvolvement and positive remark scales. There were no significant differences in the parents' emotional expressiveness between the specified group combinations (see Appendix 3.2, page 289). Mean scores are presented in TABLE XXI.

TABLE XX

DISTRIBUTION OF PARENTS' ATTITUDES TOWARDS THE CHILDREN (EE)

Mean; (s.d.)

	Mothers (n= 138)	Fathers (n= 109)	Sign of Diff
Emotional Overinvolvement (EOI)	2.2 (1.3)	1.8 (1.3)	0.001*
Warmth (W)	4.1 (0.9)	4.1 (0.1)	NS*
Hostility (H)	0.1 (0.4)	0.0 (0.1)	NS**
Positive Remarks (PR)	2.1 (1.5)	1.7 (1.0)	0.03*
Critical Comments (CC)	0.4 (1.1)	0.2 (0.6)	NS**

* T-tests; ** McNemar test
EOI:none= 0; very little= 1; some= 2; moderate= 3; high= 4; mark= 5
W: none= 0; very little= 1; some= 2; moderate= 3; high= 4-5
H: none= 0; present= 1
PR: frequency of positive remarks
CC: frequency of critical remarks (analysis of data: none=0; one or more=1)

Attitudes towards the Partner. The Expressed Emotion Scales (EE)

With regard to the parents' emotional profile concerning their partner, TABLE XXII (page 111) shows an overall absence of overinvolvement, moderate warmth, and very low levels of hostility, positive remarks and criticism. Only two mothers and none of the fathers rated in the emotional overinvolvement scale (EOI). This scale was, therefore, not examined further.

TABLE XXI

PSYCHOSOCIAL TYPOLOGY OF ILLNESS AND PARENTS' ATTITUDES TOWARDS THE CHILDREN (EE)

N, Mean (s.d.)

	HTx	HLTx	PvHL	HR	LR
MOTHER - CHILD	n=27	n=24	n=38	n=21	n=28
Overinvolvement (EOI)	2.44 (1.4)	2.33 (1.5)	1.89 (1.2)	2.29 (1.4)	1.96 (1.1)
Warmth (W)	4.19 (.8)	3.96 (1.3)	4.34 (.9)	4.29 (.7)	4.25 (.8)
Hostility (H)	0.07 (.3)	0.17 (.4)	0.05 (.3)	0.14 (.6)	0.11 (.5)
Positive Remarks (PR)	1.52 (1.5)	2.25 (1.2)	2.26 (1.5)	2.62 (1.8)	2.54 (1.3)
Critical Comments (CC)	0.33 (.8)	0.42 (1.0)	0.7 (1.2)	0.52 (1.5)	0.54 (1.1)
FATHER - CHILD	n=19	n=19	n=29	n=17	n=25
Overinvolvement (EOI)	2.05 (1.4)	1.95 (1.5)	1.66 (1.0)	1.82 (1.5)	1.92 (1.1)
Warmth (W)	4.05 (.7)	3.74 (1.3)	4.17 (1.0)	4.35 (.7)	4.36 (.7)
Hostility (H)	0.00 (.0)	0.00 (.0)	0.00 (.0)	0.06 (.2)	0.00 (.0)
Positive Remarks (PR)	1.37 (1.3)	1.47 (1.3)	1.97 (1.6)	2.76 (1.8)	2.52 (3.0)
Critical Comments (CC)	0.05 (.2)	0.16 (.5)	0.38 (.7)	0.41 (1.0)	0.16 (.3)

EOI:none= 0; very little= 1; some= 2; moderate= 3; high= 4; mark= 5
W: none= 0; very little= 1; some= 2; moderate= 3; high= 4-5
H: none= 0; present= 1
PR: frequency of positive remarks
CC: frequency of critical remarks (analysis of data: none=0; one or more=1)

TABLE XXII

DISTRIBUTION OF PARENTS' ATTITUDES TOWARDS PARTNERS (EE)

Mean (s.d.)

	Mothers (n= 127)	Fathers (n= 110)	Sign of Diff
Warmth (W)	3.4 (1.1)	3.2 (1.0)	NS*
Hostility (H)	0.0 (0.2)	0.0 (0.1)	NS**
Positive Remarks (PR)	0.2 (0.6)	0.2 (0.4)	NS*
Critical Comments (CC)	0.2 (0.6)	0.0 (0.4)	NS**

* T-tests; ** McNemar tests
W: none= 0; very little= 1; some= 2; moderate= 3; high warmth= 4-5
H: none= 0; present = 1
PR: frequency of positive remarks
CC: frequency of critical remarks (analysis of data: none=0; one or more=1)

Significant differences were not found between the mothers' and fathers' ratings. When associations between the partners' attitudes towards their partners and psychosocial typology of illness were examined, there were no significant differences in any of the mothers' or fathers' EE mean scores between any of the specified group combinations (see Appendix 3.2, page 289). The mean scores are presented in TABLE XXIII.

Patterns in the Family's Emotional Climate: attitudes towards children and partners

The existence of patterns in the family's emotional climate was explored. As there were no differences in levels of expressed emotion towards the child or the partner between the specified group comparisons, data from all groups was gathered together. A series of Pearson correlations was conducted between the parents' attitudes towards the patient and the parents' attitudes towards their partner. Overall, these correlations indicated that those parents who had a warm and positive attitude towards each other also had a warm and positive attitude towards the patient. These findings are summarised in TABLE XXIV (page 113).

TABLE XXIII

PSYCHOSOCIAL TYPOLOGY OF ILLNESS AND PARENTS' ATTITUDES TOWARDS THEIR PARTNERS (EE)

N, Mean (s.d.)

	HTx	HLTx	PvHL	HR	LR
MOTHER - PARTNER	n=23	n=22	n=32	n=22	n=28
Warmth (W)	3.43 (1.1)	3.09 (1.2)	3.31 (1.3)	3.36 (1.2)	3.46 (.9)
Hostility (H)	0.00 (.0)	0.00 (.0)	0.13 (.4)	0.05 (.2)	0.04 (.1)
Positive Remarks (PR)	0.22 (.5)	0.36 (.9)	0.59 (.7)	0.09 (.2)	0.18 (.3)
Critical Comments (CC)	0.30 (.8)	0.18 (.6)	0.34 (.6)	0.23 (.6)	0.11 (.3)
FATHER - PARTNER	n=20	n=19	n=28	n=18	n=25
Warmth (W)	3.45 (.8)	3.21 (1.1)	3.07 (1.2)	3.56 (1.2)	3.64 (.7)
Hostility (H)	0.00 (.0)	0.00 (.0)	0.07 (.3)	0.00 (.0)	0.00 (.0)
Positive Remarks (PR)	0.05 (.2)	0.32 (.5)	0.29 (.4)	0.11 (.3)	0.24 (.4)
Critical Comments (CC)	0.05 (.2)	0.16 (.6)	0.14 (.4)	0.00 (.0)	0.08 (.2)

W: none= 0; very little= 1; some= 2; moderate= 3; high= 4-5
H: none= 0; present= 1
PR: frequency of positive remarks
CC: frequency of critical remarks (analysis of data: none=0; one or more=1)

TABLE XXIV
FAMILIES' EMOTIONAL CLIMATE: ATTITUDES TOWARDS THE CHILDREN AND PARTNERS (EE)

	Warmth		Hostility		Critical Comments		Positive Remarks	
	Mothers	Fathers	Mothers	Fathers	Mothers	Fathers	Mothers	Fathers
Mother's Child								
EOI	-.075	-.072	.099	.053	.023	.112	.116	-.037
W	.502***	.557***	-.256**	-.222*	-.319***	-.267**	.240**	.208*
H	-.158*	-.311***	-.032	-.020	.285**	.092	-.093	-.100
PR	.355***	.375***	-.215**	-.137	-.131	-.270**	.173*	.309***
CC	-.032	-.249**	.075	.132	.274**	-.056	.126	.101
Father's Child								
EOI	-.136	-.074	.065	.007	-.044	.144	.004	-.093
W	.443***	.438***	-.336**	-.323***	-.319***	-.137	.102	.235**
H	.054	.056	.016	-.009	.111	-.022	-.044	-.048
PR	.169*	.249**	-.173*	-.095	-.112	-.137	.059	.246**
CC	-.116	-.285***	.143	.273**	.310***	-.016	-.080	-.054

Pearson Correlation Coefficients, Levels of Significance: * 0.05; ** 0.01; *** 0.001

As mentioned in chapter 3, attempts to control for type I and II errors were not made because of the exploratory nature of this study, therefore, some significant associations might be due to chance. These results should be interpreted with caution.

Family Adjustment

Overall, 75 families (45.5%) were found to be well adjusted, 65 families (39.4%) were moderately adjusted and 25 families (15.2%) were poorly adjusted. FIGURE 12 presents family functioning in each group.

Figure 12: Psychosocial Typology of Illness and Family Adjustment

%
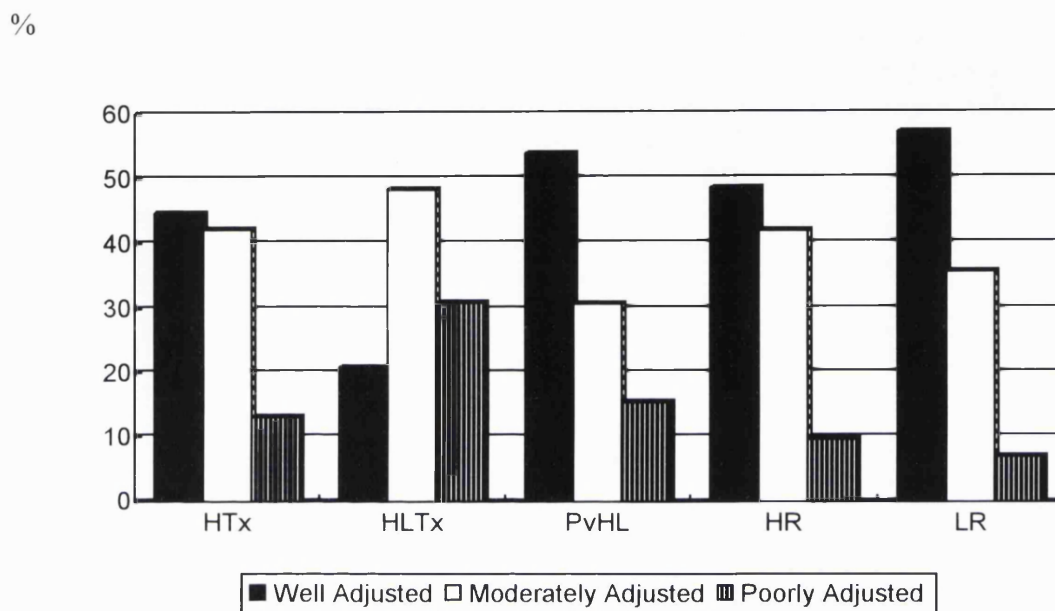
There were no significant differences in family adjustment between the transplantation and conventional cardiac surgery groups (X^2=3.7, df=2, p=0.15, NS). Nor were differences found when comparing the families of the two groups of children undergoing conventional cardiac surgery (p=0.78, NS). However, differences were found when comparing the specified transplant group combinations. The families whose children were on the active list for heart-lung transplantation (HLTx) had greater adjustment difficulties than the families whose children were on the provisional list for heart-lung transplantation (PvHL) (X^2=7.78, df=2, p=0.02). Family adjustment was also poorer in the HLTx group than in the HTx group, but not significantly so (X^2=5.42, df=2, p=0.06).

Associations between Family Adjustment and the Expressed Emotion Scales

Associations between family adjustment and the expressed emotion scales are summarised in TABLE XXV. As above, for this analysis families were grouped together.

TABLE XXV

FAMILY ADJUSTMENT AND THE EXPRESSED EMOTION SCALES
N, Mean (s.d.)

	Well Adjusted		Moderate-Poorly Adjusted	
Mothers - Children				
Overinvolvement (EOI)	n=64,	1.6 (1.0)	n=74,	2.5 (1.4) *
Warmth (W)	n=64,	4.6 (.6)	n=74,	3.8 (1.0) *
Hostility (H)	n=64,	0.3 (.2)	n=74,	0.1 (.6) **
Positive Remarks (PR)	n=64,	2.5 (1.6)	n=74,	1.9 (1.3) *
Critical Comments (CC)	n=64,	0.3 (.7)	n=74,	0.7 (1.4) **
Fathers - Children				
Overinvolvement (EOI)	n=49,	1.5 (1.1)	n=60,	2.1(1.3) *
Warmth (W)	n=49,	4.5 (.6)	n=60,	3.8 (1.0) *
Hostility (H)	n=49,	0.0 (.1)	n=60,	0.0 (.0) **
Positive Remarks (PR)	n=49,	2.5 (2.4)	n=60,	1.6 (1.5) *
Critical Comments (CC)	n=49,	0.1 (.4)	n=60,	0.3 (.7) **
Mothers - Partners				
Warmth (W)	n=56,	4.1 (.5)	n=71,	2.7 (1.1) *
Hostility (H)	n=56,	0.0 (.0)	n=71,	0.0 (.3) **
Positive Remarks (PR)	n=56,	0.4 (.7)	n=71,	0.2 (.5) *
Critical Comments (CC)	n=56,	0.0 (.2)	n=71,	0.3 (.8) **
Fathers - Partners				
Warmth (W)	n=48,	4.0 (.5)	n=62,	2.8 (1.0) *
Hostility (H)	n=48,	0.0 (.0)	n=62,	0.0 (.2) **
Positive Remarks (PR)	n=48,	0.2 (.4)	n=62,	0.1 (.4)
Critical Comments (CC)	n=48,	0.0 (.0)	n=62,	0.1 (.5) **

* T-test, **McNemar tests

Parents in well adjusted families showed less overinvolvement with the children (mother: t=-4.1, p=0.0001; father: t=-2.6, p=0.009) and rated higher in warmth (mother: t=5.2, p=0.0001; father: t=4.2, p=0.0001) and in positive remarks (mother: t=2.2, p=0.2; father: t=2.1, p=0.03) than parents in moderate-poorly adjusted families. The mothers in the former group also showed less hostility (p=0.006) and the fathers were rated lower on critical comments p=0.003). Figure 13 shows the association between family adjustment and mothers' warmth.

Figure 13: Family Adjustment and Mothers' Warmth towards the Children

%

■ Well Adjusted □ Moderately Adjusted ▥ Poorly Adjusted
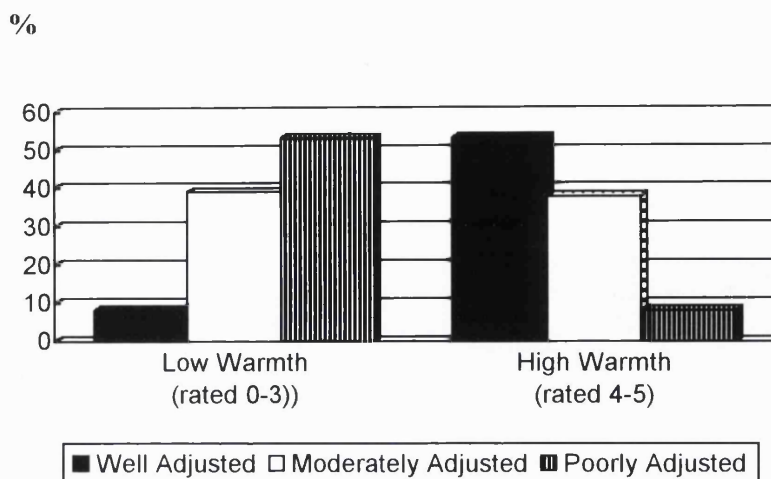
With regard to parents' attitudes towards their partner, parents in well adjusted families scored higher on warmth (mother: t=8.5, p=0.0001; father: t=8.4, p=0.0001) and made fewer critical comments (mother: p=0.001; father: p=0.03) than those parents in moderate-poorly adjusted families. Mothers in the well adjusted families group also rated higher on positive remarks (t=2.1, p=0.03) and lower in hostility (p=0.002) than the mothers in the other group. Figure 14 shows the association between family adjustment and mothers' warmth towards their partners.

Figure 14: Family Adjustment and Mothers' Warmth towards their Partners

%

■ Well Adjusted □ Moderately Adjusted ▥ Poorly Adjusted
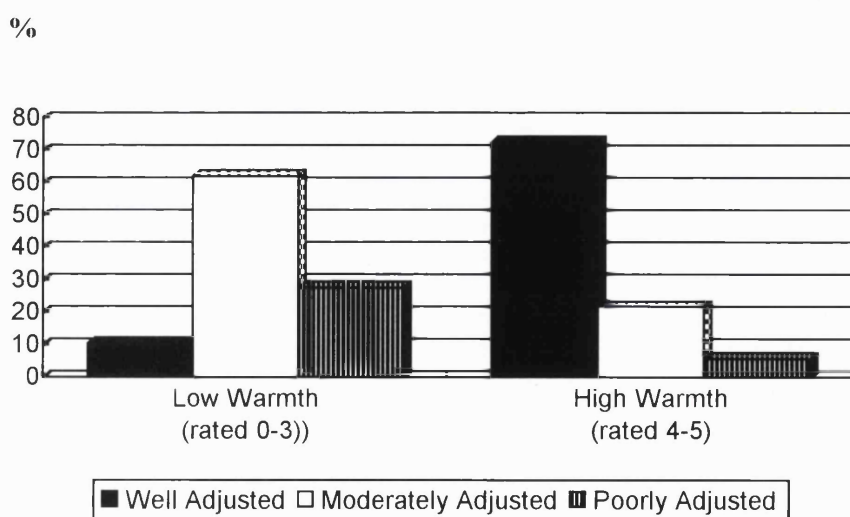
Box 1: Psychosocial profile presented by children referred for transplantation and children undergoing conventional cardiac surgery

- A total of 136 families participated in the study.

- Children assessed for transplantation (n=106) were more ill than children undergoing conventional cardiac surgery (n=59).

- 24% of the children had a psychiatric diagnosis, and over 60% had mild-moderate psychosocial impairment.

  Differences were not found between the groups.

- 40-64% of the mothers and 36-44.5% of the fathers scored as having a psychiatric diagnosis.

  Marital adjustment mean scores were within average-above average range.

  Differences between the groups were not found with regard to psychological or marital adjustment.

- Parents showed low levels of overinvolvement, hostility and critical comments, and high levels of warmth towards their children. They also showed moderate warmth and low levels of positive remarks, hostility, and criticisms towards their partners.

  Over half of the families were moderate-poorly adjusted.

  Differences between the groups were not found with regard to parents' attitudes or family adjustment.

4.2 ASSOCIATIONS BETWEEN CHILDREN'S ADJUSTMENT, PARENTS AND FAMILY MEASURES

Because no major differences between the specified groups were found in any of the previous analyses, the results are presented for all groups combined.

4.2.1 Associations between Children's Mental Status (ICD-9 Classification), Demographic, Parental and Family's Measures

Children's Mental Status (ICD-9 Classification) and Demographic Measures:

Family composition, social class, number of previous operations, child's sex and age were not related to the children's mental status.

Children's Mental Status (ICD-9 Classification), Parental and Marital Adjustment

TABLE XXVI compares parents' psychological (GHQ-28) and marital (GRIMS) adjustment mean scores of two groups of parents, those parents with and without a child suffering from a psychiatric disorder (ICD-9).

TABLE XXVI

CHILDREN'S MENTAL STATUS (ICD-9), PARENTS' PSYCHOLOGICAL AND MARITAL ADJUSTMENT

Mean (s.d.)

	No Abnormality		Psychiatric Diagnoses		Sign of Diff
Mother's GHQ-28[#]	n=94	4.9 (4.9)	n=25	5.2 (4.9)	NS
Father's GHQ-28[#]	n=86	3.4 (3.9)	n=22	3.7 (2.9)	NS
Mother's GRIMS[##]	n= 84	3.7 (2.2)	n= 21	4.2 (2.4)	NS
Father's GRIMS[##]	n= 84	3.5 (2.3)	n= 22	4.1 (1.8)	NS

T-tests, two tailed probability
[#] A score of 4 or more indicates parental psychiatric distress/poor adjustment
[##] A score of 6 or more indicates poor marital relationship

Parents of children who had a psychiatric diagnosis scored slightly higher in the GHQ-28 scale than the parents whose children did not have a psychiatric disorder. However, this difference did not reach statistical significance (mothers: p=0.7; fathers: p=0.6). Mothers' mean scores were above the threshold score indicating poor adjustment or possible psychiatric illness. A crosstabulation was conducted examining whether those parents scoring above the threshold score were more likely to have children suffering from a psychiatric disorder than parents scoring below the threshold score. Associations were not found between the parental psychiatric functioning and children's mental status (mothers: X^2=0.2, df=1, p=0.5, NS; fathers: X^2=0.1, df=1, p=0.7, NS).

With regards to the marital adjustment ratings, the parents whose children had a psychiatric disturbance showed a tendency towards more dissatisfaction with their marriage than the parents whose children were not disturbed. But this was not significant and the marital mean scores indicated average or above average relationships.

Children's Mental Status (ICD-9 Classification) and Family's Organisation Measures:

TABLE XXVII summarises the associations between the children's mental status and the parents' attitudes towards the children and their partner. Mothers of children with a psychiatric diagnoses were significantly less warm and more critical towards their children than the mothers whose children were not disturbed (although the levels of criticism were in fact low in both groups). Fathers of children with psychiatric problems made more critical comments about their children than the fathers whose children were not disturbed. There were no associations between the children's mental status and mothers' or fathers' levels of overinvolvement, hostility or positive remarks. Regarding parents' attitudes towards their partner, only mothers' and fathers' hostility scale was significantly associated with the children's mental status, though again hostility was rarely expressed in either group.

TABLE XXVII

CHILDREN'S MENTAL STATUS (ICD-9) AND PARENTS' ATTITUDES (EE)

N, Mean (s.d.)

	No Abnormality	Psychiatric Diagnoses	Sign of Diff
Mother - Child			
EOI	n= 91, 2.0 (1.3)	n= 27, 2.5 (1.4)	NS*
W	n= 91, 4.3 (0.8)	n= 27, 3.6 (1.3)	0.01*
H	n= 91, 0.0 (0.4)	n= 27, 0.2 (0.6)	NS**
PR	n= 91, 2.3 (1.6)	n= 27, 2.2 (1.2)	NS*
CC	n= 91, 0.5 (1.2)	n= 27, 0.9 (1.3)	0.01**
Father - Child			
EOI	n= 71, 1.8 (1.2)	n= 18, 2.0 (1.3)	NS*
W	n= 71, 4.1 (0.8)	n= 18, 3.6 (1.4)	NS*
H	n= 71, 0.0 (0.1)	n= 18, 0.0 (0.0)	NS**
PR	n= 71, 2.0 (2.1)	n= 18, 2.1 (1.9)	NS*
CC	n= 71, 0.2 (0.6)	n= 18, 0.6 (0.7)	0.004**
Mother - Partner			
W	n= 83, 3.4 (1.0)	n= 24, 3.0 (1.4)	NS*
H	n= 83, 0.0 (0.0)	n= 24, 0.1 (0.4)	0.01**
PR	n= 83, 0.3 (0.6)	n= 24, 0.2 (0.6)	NS*
CC	n= 83, 0.2 (0.6)	n= 24, 0.2 (0.6)	NS**
Father - Partner			
W	n= 73, 3.4 (1.0)	n= 17, 3.1 (1.1)	NS*
H	n= 73, 0.0 (0.0)	n= 17, 0.1 (0.4)	0.04**
PR	n= 73, 0.2 (0.4)	n= 17, 0.2 (0.5)	NS*
CC	n= 73, 0.0 (0.3)	n= 18, 0.0 (0.2)	NS**

* T-tests; ** McNemar tests
EOI: none= 0; very little= 1; some= 2; moderate= 3; high= 4; mark= 5
W: none= 0; very little= 1; some= 2; moderate= 3; high= 4-5
H: none= 0; present= 1
PR: frequency of positive remarks
CC: frequency of critical remarks (analysis of data: none=0; one or more=1)

An association was also found between family adjustment and children's mental status. As shown in FIGURE 15, there were fewer children with a psychiatric disturbance in the well-adjusted family group than in the poorly adjusted family group (X^2=9.80, df=2, p=0.007).

Figure 15: Children's Mental Status (ICD-9) and Family Adjustment

%

■ No Psychiatric Abnormality ☐ Psychiatric Diagnoses
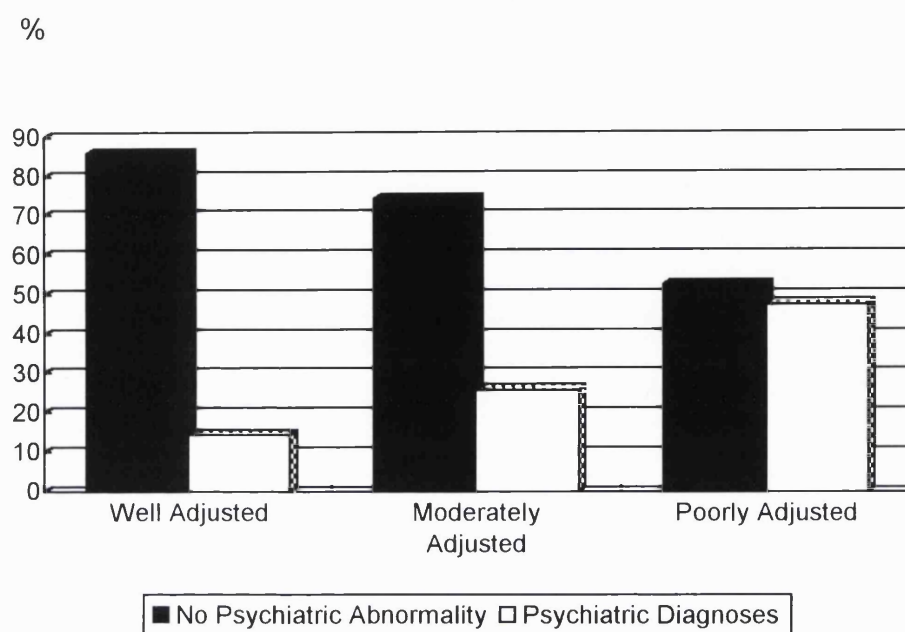
4.2.2 Associations between Children's Global Assessment of Functioning (GAF Scale), Demographic, Parental and Family's Measures.

Children's Global Assessment of Functioning (GAF Scale) and Demographic Measures:

Family composition, social class, number of previous operations and children's age was not associated with children's psychosocial functioning. However, gender was significantly associated with children's psychosocial functioning. The boys participating in this study (n= 72, mean: 66.7) had slightly poorer functioning than girls (n= 72, mean: 70.4) (p=0.04).

Children's Global Assessment of Functioning (GAF Scale), Parental and Marital Adjustment:

TABLE XXVIII compares the parents' psychological (GHQ-28) and marital adjustment mean scores (GRIMS) of three groups of parents: parents whose children showed no impairment, parents whose children had mild to moderate or severe psychosocial impairment.

TABLE XXVIII

CHILDREN'S GLOBAL ASSESSMENT OF FUNCTIONING (GAF SCALE),

PARENTS' PSYCHOLOGICAL AND MARITAL ADJUSTMENT

N, Mean (s.d.)

	No Impairment	Mild-Moderately Impaired	Severely Impaired
Mothers' GHQ-28[*]	n=48 3.6 (3.1) *	n=66 6.0 (5.7) *	n=6 5.8 (2.3)
Fathers' GHQ-28[*]	n=41 3.0 (2.9)	n=62 3.8 (4.3)	n=6 1.8 (1.8)
Mothers' GRIMS[**]	n=38 3.0 (1.9)**	n=62 4.3 (2.4) **	n= 6 3.8 (2.1)
Fathers' GRIMS[**]	n=38 2.8 (1.8)** *	n=63 4.1 (2.3)** *	n= 6 3.3 (2.0)

One-way analysis of variance, Scheffe procedure method.

*, **, ** *   groups significantly different at the .05 level

[*] A score of 4 or more indicates parental psychiatric distress/poor adjustment

[**] A score of 6 or more indicates poor marital relationship

This table shows that children with no psychosocial impairment had mothers with better psychological functioning than children who showed mild to moderate impairment (F=3.5, df=2,117, p=0.03). See FIGURE 16. TABLE XVIII also shows that parents of children with no psychosocial impairment were more satisfied with their marital relationship than the parents whose children showed mild to moderate impairment (mothers: F=3.8, df=2,103, p=0.02; fathers: F=4.1, df=2,104, p=0.01). Mean marital adjustment scores in all three groups were below the threshold indicating marital difficulties.

Figure 16: Children's Global Assessment of Functioning (GAF Scale) and Mother's Mental Status
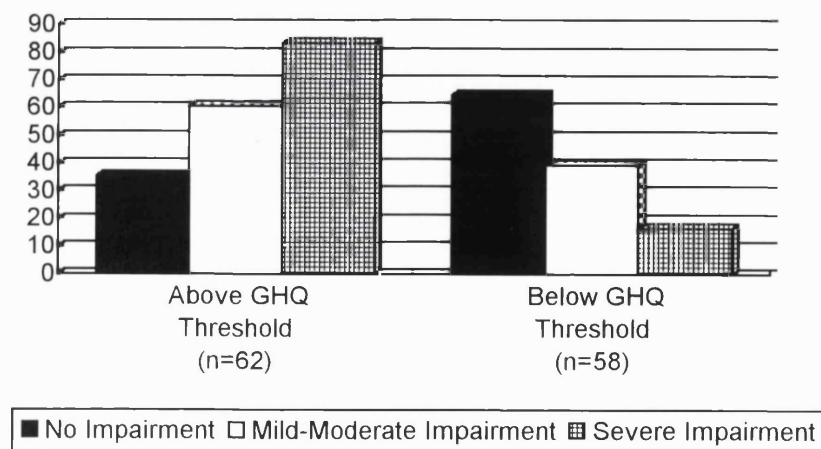
As shown in FIGURE 16, mothers scoring above the GHQ-28 threshold score had greater number of children with impaired psychosocial functioning than the mothers scoring below the GHQ-28 threshold score (X^2 =9.5, df=2, p=0.008).

Children's Global Assessment of Functioning (GAF Scale) and Family Organisation Measures:

Significant associations were found between the children's level of functioning and parental expressed emotion (EE). A summary is presented in TABLE XXIX. Mothers of children with no psychosocial impairment were less overinvolved (F=4.04, df=2,118, p=0.02) and showed higher levels of warmth (F=7.0, df=2,118, p=0.001) than the mothers whose children had a mild to moderate impairment. Mothers of children with severe impairment showed less warmth to their children than the mothers of good functioning children. In addition, the mothers of severely impaired children scored significantly higher in the hostility scale than the mothers of children in the other two groups (McNemar test, p=0.01).

Only one of the fathers' EE subscales was associated with the children's global assessment of functioning. Fathers whose children had good psychosocial functioning scored higher in warmth than the fathers of children who were severely impaired; and the fathers of children with mild impairment showed higher warmth than the parents of children who were severely impaired in their psychosocial functioning (F=6.9, df=2,89, p=0.001).

It is of interest that the levels of warmth shown by the parents of children with good functioning or by the parents of children with mild to moderate impairment, were high; whilst the levels of warmth shown by the mothers of severely impaired children were only moderate. In addition, none of the mothers whose children had normal levels of psychosocial functioning showed hostility towards their child, whilst a few mothers in the other two groups scored on this scale.

TABLE XXIX
GLOBAL ASSESSMENT OF FUNCTIONING AND PARENTS' ATTITUDES
Mean (s.d.)

	No Impairment	Mild-Moderately Impaired	Severely Impaired
Mother-Children	n=48	n=65	n=8
Overinvolvement	1.7 (1.1)*	2.4 (1.4)*	2.5 (2.5)
Warmth	4.5 (0.6)***	4.0 (1.0)**	3.5 (1.4)*
Hostility	0.0 (0.0)*	0.1 (0.5)**	0.6 (1.1)***
Positive Remarks	2.6 (1.6)	2.1 (1.3)	2.7 (2.3)
Critical Comments	0.3 (0.7)	0.6 (1.4)	1.2 (1.8)
Father-Children	n=33	n=52	n=7
Overinvolvement	1.5 (1.0)	2.0 (1.4)	1.8 (1.4)
Warmth	4.4 (0.6)*	4.0 (1.0)**	3.0(1.4)***
Hostility	0.0 (0.0)	0.0 (0.1)	0.0 (0.0)
Positive Remarks	2.2 (1.8)	2.1 (2.4)	1.4 (1.3)
Critical Comments	0.1 (0.4)	0.3 (0.7)	0.5 (0.7)
Mother-Partners	n=40	n=63	n=7
Warmth	3.7 (0.8)*	3.0 (1.2)*	3.2 (1.3)
Hostility	0.0 (0.0)	0.03 (0.1)	0.2 (0.7)
Positive Remarks	0.4 (0.6)	0.2 (0.6)	0.4 (0.7)
Critical Comments	0.07 (0.3)	0.3 (0.7)	0.7 (1.2)
Father-Partners	n=33	n=53	n=7
Warmth	3.6 (0.8)	3.2 (1.0)	3.0 (1.6)
Hostility	0.0 (0.0)*	0.0 (0.0)*	0.2 (0.7)**
Positive Remarks	0.3 (0.4)	0.1 (0.4)	0.1 (0.3)
Critical Comments	0.0 (0.1)	0.0 (0.4)	0.1 (0.3)

*, ** and ** denote groups significantly different at the 0.05 level

TABLE XXIX also shows that mothers of children with good psychosocial functioning were warmer towards their husbands than the mothers whose children had mild to moderate psychosocial impairment (F=4.9, df=2,107, p=0.008). Regarding the fathers' ratings, fathers of severely impaired children were more hostile towards their wives than the fathers in the other two groups (p=0.03). No other statistical differences were found between the groups.

Analysis of the family adjustment measure revealed that well adjusted families were less likely to have children who were psychosocially impaired than moderately or poorly adjusted families (F=12.8, df=2,141, p=0.001). This is shown in FIGURE 17.

Figure 17: Children's Global Assessment of Functioning (GAF Scale) and Family Adjustment

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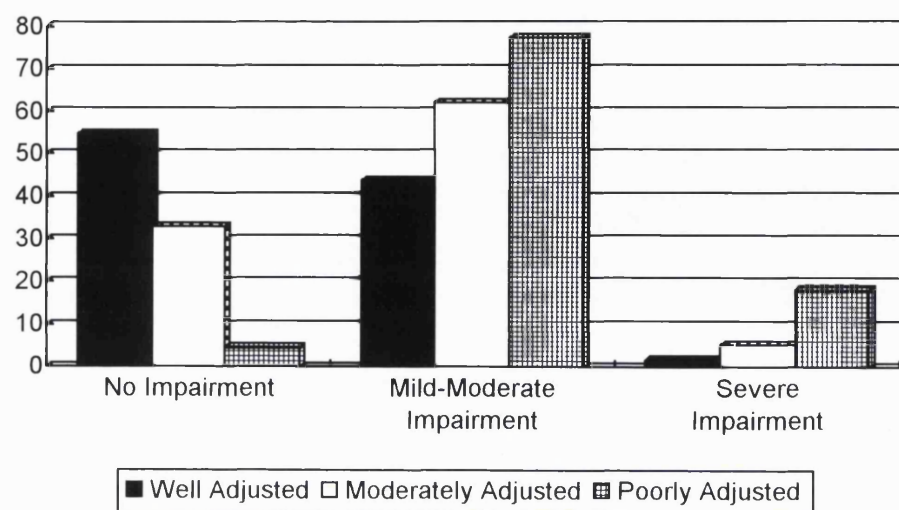
Box 2: Psychosocial Factors associated with Children's Adjustment

- Demographic characteristics, parents' psychological and marital adjustments were not associated with children's mental status.

  Low warmth, presence of critical comments or hostility and poor family adjustment were associated with children's psychiatric disturbance.

- Boys had poorer psychosocial functioning than girls. No other demographic characteristic was associated with the children's level of psychosocial functioning.

- Poor mothers' psychological adjustment, poor marital adjustment, high overinvolvement, low warmth, presence of hostility and poor family adjustment were associated with psychosocial impairment.

4.3 CHANGES IN THE CHILDREN'S, PARENTS' AND FAMILY MEASURES FOLLOWING SURGERY

4.3.1 Total Number of Families Participating at the Follow-up

One hundred and twenty six children underwent transplantation or conventional cardiac surgery and a total of 118 children were available for followed-up (T2). As shown in FIGURE 18, 8 children who received a heart transplant, 9 children who received heart-lung transplantation, and 3 children who underwent high-risk cardiac surgery died within 10 months post-operatively. All children who were initially placed on the provisional list (PvHL) for heart-lung transplantation had moved to the active list between the initial assessment and the follow-up. Twenty-six of those children (66.6%) died prior to the follow-up, and one child withdrew from the waiting list feeling unable to cope with the stresses associated with transplant treatment.

There was some variation in the amount of time that the children had to wait to receive transplantation. Heart recipients waited an average of 5 months (range: 1 day to 11 months), and heart-lung recipients waited an average of 11 months (range: 2 to 20 months). Survivors were followed up (T2) at a mean of 12 months (range: 9 to 16 months) post-operatively, and at a mean of 21 months from initial assessment. The twelve children who did not received transplantation (PvHL group) were also followed up at a mean of 21 months (range: 17 to 24 months) from the initial assessment. This mean was based on previous data and was chosen to make it comparable to the mean of the transplant recipients. Children who received conventional cardiac surgery were also followed up at a mean of 12 months (range 9 to 16 months) post-operatively.

4.3.2 Changes in the Children's and Families' Demographic Characteristics

Differences in the children's and families' demographic characteristics between the 3 specified group combinations (see chapter 3, page 88) were examined. There were no sex differences between the groups (X^2=7.2, df=4, p=0.12, NS); but age differences were found (F=3.1, df=4,100, p=0.01). The children who had conventional cardiac surgery (HR and LR groups together) were younger than the transplant recipients (p=0.008).

FIGURE 18: PARTICIPANTS AT T1 and T2

T1: Entry into Study   T2: Follow-up

165 Families

Tx Programme:
106 Children

HTx Group: 38 Children received Heart Tx
8 children died shortly post-operatively
30 children were followed up
18 boys, 12 girls
mean age: 9.3 years

HLTx Group: 29 Children received Heart-Lung Tx
9 children died shortly post-operatively
20 children were followed up
11 boys, 9 girls
mean age: 11.2 years

Pv HL Group: 39 children in provisional list for Tx
26 children died prior to the follow up and
1 child withdrew from the Tx programme
12 children were followed up
4 boys, 8 girls
mean age: 11.7 years

Conventional Cardiac Surgery:
59 Children

HR Group: 28 Children received High-risk Surgery
3 children died during surgery
25 children were followed up
16 boys, 9 girls
mean age:8.4 years

LR Group: 31 Children received Low-risk Surgery
All children survived
31 children were followed up
14 boys, 17 girls
mean age: 9.1 year

At the follow-up, a total of 81 children (75%) were living with both their biological parents. Regarding changes in family composition between initial assessment (T1) and the follow-up (T2), the parents of 3 transplant recipients had separated within 7 months post-transplantation, and the parents of another 4 transplant recipients were thinking about getting a divorce at the time of the follow-up. The mother of 1 child who received a heart transplant was diagnosed with cancer 3 months after the child's operation and died 6 months later. At 10 months post-surgery, one of the children who had received conventional high-risk cardiac surgery lost a sibling following a sudden onset of illness. When changes in family composition between the specified group combinations were examined no significant differences were found.

For the final assessment, home visits were arranged for 13 families who were either unable or unwilling to come to London to be followed up; these families were living in Yorkshire, Scotland and Wales. The medical evaluations were arranged at their local hospitals.

4.3.3 Changes in the Children's Physical Health Measures

Significant statistical changes in all the physical measures from T1 to T2 were found for those children who underwent transplantation or other forms of cardiac surgery. The changes in the health measures are summarised in TABLE XXX. Children who received heart (HTx) or heart-lung transplantation (HLTx), and conventional high (HR) or low-risk (LR) cardiac surgery showed an improvement in both their physical status and physical disability as rated by the physician and their parents. Their percentage of ideal weight also increased significantly. According to the teachers' report, heart (T1 mean:1.0; T2 mean:3.9; t=-10.1; p=0.001) and heart-lung transplant recipients (T1 mean:1.4; T2 mean: 3.8; t=-5.8; p=0.001), and children who received conventional low-risk cardiac surgery (T1 mean:3.8; T2 mean:4.3; t=-3.1; p=0.004) improved their school attendance following surgery. Nevertheless, there was no improvement in the group of children who received conventional high-risk cardiac surgery (T1 mean: 3.3; T2 mean:3.3; t=0.0; p=1.00).

Children who were initially on the provisional list (PvHL) for heart-lung transplantation and who had moved to the active list by the time of the follow-up, showed a deterioration in their physical status; but there were no significant changes in their ideal weight for height percentage, parental report on physical disability or in the teachers' report on school attendance.

TABLE XXX

CHANGES IN THE CHILDREN'S HEALTH MEASURES PROFILE

Mean (s.d.)

	Physical Status			Weight for Height			Physical Disability		
	T1	T2	Sign	T1	T2	Sign	T1	T2	Sign
HTx	3.7 (0.5)	1.1(0.3)	0.0001	88.4 (12.8)	104.6 (17.3)	0.0001	3.7 (1.0)	1.5 (0.9)	0.0001
HLTx	3.9 (0.3)	1.5(0.6)	0.0001	77.4 (12.4)	92.8 (14.7)	0.0001	3.7 (0.7)	1.2 (0.4)	0.0001
HR	2.7 (0.5)	1.4(0.8)	0.0001	91.0 (13.2)	95.0 (12.1)	0.02	2.9 (0.7)	2.2 (1.1)	0.009
LR	1.3 (0.6)	1.0(0.0)	0.003	92.2 (9.1)	95.7 (8.6)	0.0001	1.3 (0.4)	1.1 (0.3)	0.04
PvHL	2.8 (0.7)	3.2(0.7)	0.01	84.5 (6.1)	80.4 (9.6)	NS	2.9 (1.1)	3.5 (1.0)	NS

Paired T-tests
Physical Status (1-4):   1= well functioning, 4= critically ill.
Physical Disability (1-6): 1= no impairment,   6= needs a wheelchair.
Percentage of ideal Weight corrected for Height .

Differences between the specified groups were found in the direction and intensity of change of the following measures: children's physical status (One-way anova, F=73.6, df=3,100, p=0.0001), weight for height (F=10.4, df=3,97, p =0.0001), parental report on the children's physical disability (F=35.7, df=3,96, p =0.0001) and school attendance (F=20.6, df=3,85, p =0.001).

The children who received transplantation had a greater improvement in their physical status (p=0.001), physical disability (p=0.002), ideal weight for height percentage (p=0.001) and school attendance (p=0.0001) than the children who received conventional cardiac surgery. The degree of improvement, in all health measures, was similar for the heart and heart-lung transplant recipients. However, the children who received conventional high-risk surgery (HR) had a greater improvement in their children's physical status (p=0.001), and disability (p=0.02), than in the children who received conventional low-risk (LR) cardiac surgery.

4.3.4 Changes in the Children's Adjustment Measures

Changes in the Children's Mental Status (ICD-9 Classification)

Overall, there was a significant reduction in the proportion of children with a psychiatric disturbance following transplantation and other forms of cardiac surgery. Fifteen of 86 children (17.4%) who had a psychiatric disorder prior to surgery (T1) did not show a psychiatric disturbance at the follow-up (T2) (McNemar test, p=0.0005). Changes in the children's mental status were examined separately for each group. See FIGURE 19. A marked tendency towards a reduction in the number of children with an ICD-9 diagnosis was observed in all surgery groups, but was only significant in the conventional high-risk cardiac surgery group (HR) (McNemar test, p=0.03). There was an increase in the number of children with a psychiatric diagnosis in the group of non-transplanted children (PvHL group), but this did not reach statistical significance (McNemar test, p=0.50).

Figure 19: Changes in Children's Mental Health Status (ICD-9)

%
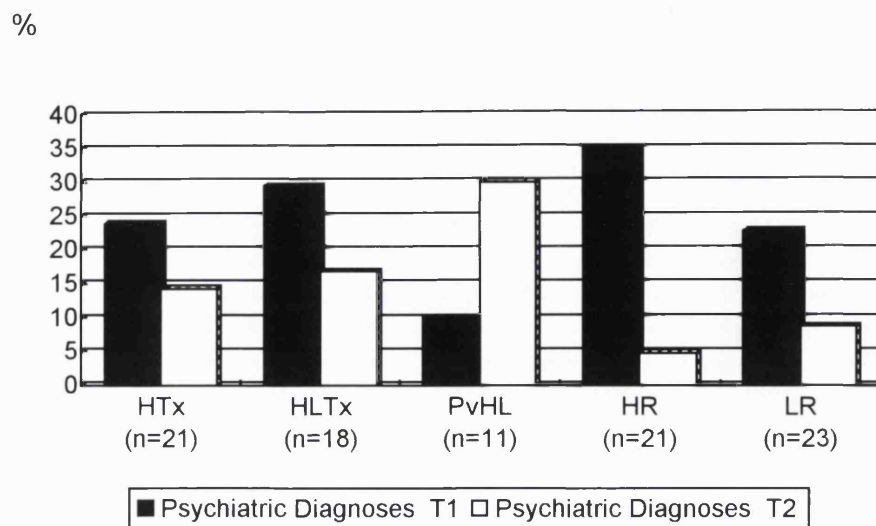
Changes in the Children's Global Assessment of Functioning (GAF Scale)

Overall, those children who received surgery had an improvement in their psychosocial functioning. As shown in TABLE XXXI, mean score comparisons indicated that heart (p= 0.009) and heart-lung (p=0.0001) transplant recipients, and children who underwent low-risk cardiac surgery (p=0.002) had a significant improvement in their psychosocial functioning post-operatively. The psychosocial functioning of the children who underwent high-risk surgery also improved but not significantly so (p=0.09). However, children awaiting heart-lung transplantation (PvHL) experienced a deterioration in their functioning (p=0.02).

TABLE XXXI

CHANGES IN CHILDREN'S GLOBAL ASSESSMENT OF FUNCTIONING

	HTx n=23	HLTx n=21	PvHL n=11	HR n=21	LR n=26
+T1: mean (sd)	66.8 (11.0)*	63.3 (8.4)*	81.8 (9.0)*	68.2 (9.8)	71.3 (12.5)*
+T2: mean (sd)	72.8 (10.6)*	70.8 (11.7)*	77.6 (12.1)*	71.7 (9.0)	76.0 (13.1)*
◆T1:. Impaired	63.6%	83.3%	10%	61.9%	54.5%
◆T2: Impaired	50%	50%	30%	52.4%	34.8%

+Paired T-tests; ◆McNemar tests
* Statistically significant mean differences

Changes in the individual global assessment of functioning ratings are shown in FIGURE 20. TABLE XXXI also shows the proportion of children, at T1 and T2, scoring below the threshold score (70/71), this indicating mild to severe psychosocial impairment. None of the McNemar tests were significant.

When the surgery groups were compared regarding the intensity of improvement in the children's functioning, no significant differences were found between any of the group combinations (One-way anova, F=0.90, df=3,83, p=0.44, NS). This means that the degree of improvement in the children's psychological functioning was similar in all groups.

Figure 20: Changes in the Children's Individual Global Assessment of Functioning Ratings (GAF Scale)

No Impairment (ratings range: 71-90); Mild-Moderate Impairment (ratings range: 51-70); Severe Impairment (ratings range:20-50)
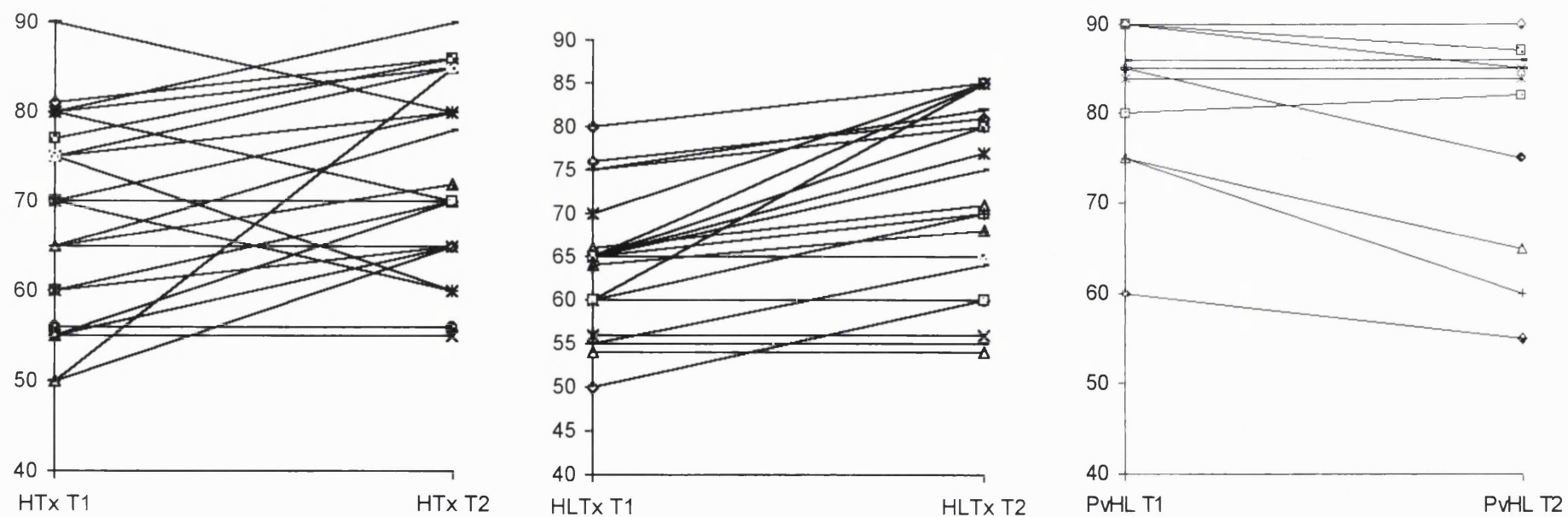
Figure 20: Changes in the Children's Individual Global Assessment of Functioning Ratings (GAF Scale)

No Impairment (ratings range: 71-90); Mild-Moderate Impairment (ratings range: 51-70); Severe Impairment (ratings range:20-50)
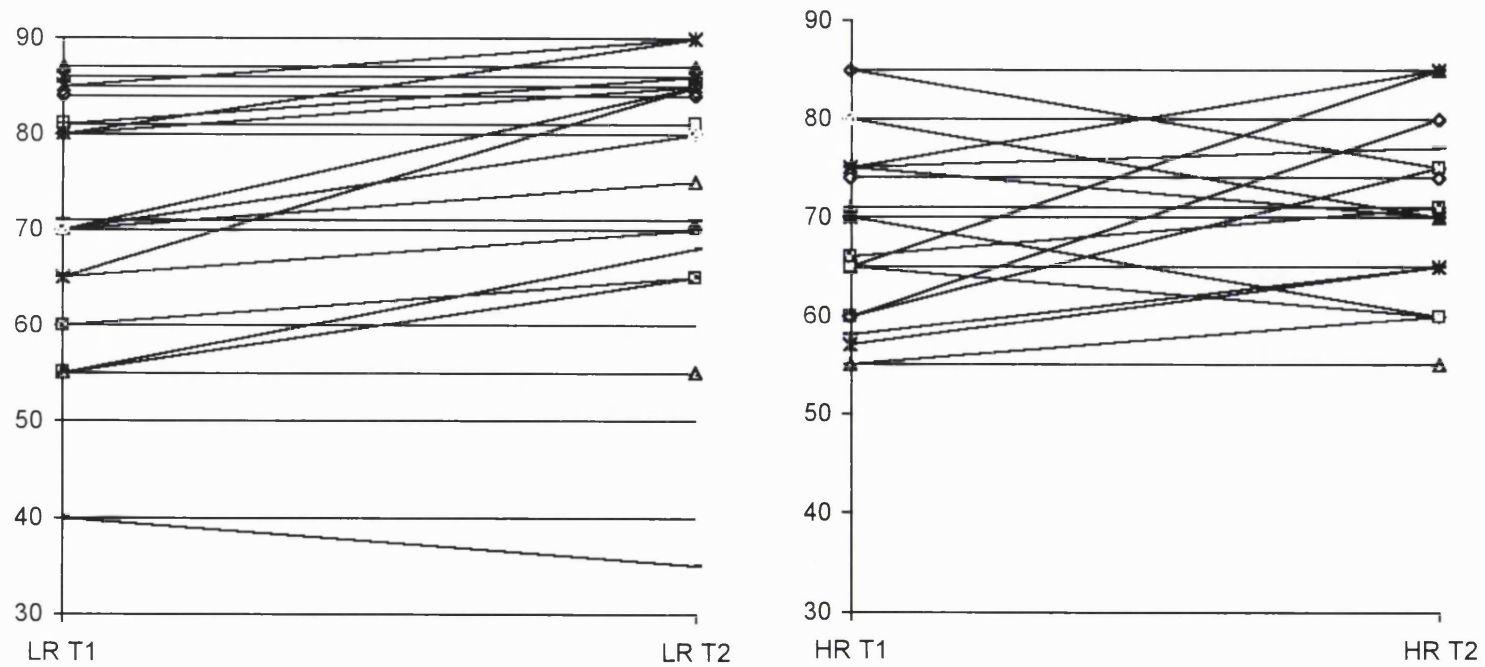
4.3.5 Changes in the Parents' Psychological and Marital Adjustment Measures

Changes in Parents' Psychological Adjustment (GHQ-28):

All surgery groups showed significant changes in parents' psychological adjustment (GHQ-28). A summary is presented in TABLE XXXII.

A comparison of the mean scores shows that both fathers and mothers of heart transplant recipients (HTx), and the fathers of the heart-lung transplant recipients (HLTx), but not the mothers, were better adjusted at the follow-up (T2) than at initial assessment (T1). Similar results were obtained for the parents of children who received conventional high-risk cardiac surgery (HR). Although the functioning of those parents whose children had received low-risk surgery (LR) was also improved at 12 months post-surgery, this only reached statistical significance for the mothers. However, the psychological functioning of the parents whose children were awaiting heart-lung transplantation (PvHL group) deteriorated between the initial assessment and the follow-up, although this was not statistically significant.

TABLE XXXII also shows changes in the number of parents scoring on or above the GHQ-28 threshold score. Overall, following surgery there was a reduction in the number of parents scoring as psychiatrically distressed (threshold score of 4/5) but this was only significant in the group of mothers whose children had received heart-lung transplantation.

When the specified group combinations were examined for differences in the degree of parents' psychological improvement, it was found that there were no significant differences between the groups (mothers: F=0.2, df=3,83, p=0.8; fathers:F=0.5, df=3,78, p=0.6).

TABLE XXXII

CHANGES IN THE PARENTS' PSYCHOLOGICAL ADJUSTMENT RATINGS (GHQ-28)

		Mean scores (sd) **			Psychiatric Distress n (%) *		
		T1	T2	Sign	T1	T2	Sign
Mothers'							
HTx	n=20	4.9 (5.7)	2.6 (4.6)	0.0001	7 (35.0%)	3 (15.0%)	NS
HLTx	n=18	4.7 (2.5)	2.7 (4.4)	0.06, NS	13 (72.2%)	6 (33.3%)	0.01
HR	n=22	6.0 (5.7)	2.7 (4.3)	0.04	11 (50.0%)	6 (27.2%)	NS
LR	n=27	6.1 (7.2)	2.7 (5.4)	0.04	13 (48.1%)	6 (22.2%)	0.06, NS
PvHL	n=12	2.8 (2.5)	3.0 (2.4)	NS	4 (33.3%)	4 (33.3%)	NS
Fathers'							
HTx	n=18	5.1 (7.6)	3.3 (6.5)	0.03	5 (27.7%)	4 (22.2%)	NS
HLTx	n=16	3.3 (2.2)	1.4 (1.9)	0.01	8 (50.0%)	3 (18.7%)	NS
HR	n=21	4.8 (4.1)	2.1 (3.0)	0.005	11 (52.3%)	5 (23.8%)	NS
LR	n=27	2.7 (3.0)	1.5 (4.2)	NS	8 (29.6%)	3 (11.1%)	0 .06, NS
PvHL	n= 7	3.5 (1.6)	4.0 (1.5)	NS	3 (42.8%)	4 (57.1%)	NS

** Paired T-test

* McNemar Test, Psychiatric Distress refers to number of cases scoring above the GHQ-28 threshold score of 4/5.

Changes in Parents' Marital Adjustment (GRIMS)

Significant differences between the mothers (mean 4.0) and fathers (mean 4.1) mean scores comparisons were not found (p= 0.6). Nevertheless, both parents showed more dissatisfaction with their marital relationship at the follow-up (mothers' means: T1=3.5; T2=3.9, p=0.02; fathers' means: T1=3.3, T2=4.0, p=0001). Summaries of the changes in marital adjustment scores are presented in TABLE XXXIII.

Parents of heart transplant recipients (HTx) were significantly more dissatisfied with their marital relationship at 12 month post-transplant than prior to transplantation. Similar findings were obtained for parents of children who received high and low-risk conventional cardiac surgery, but this was only statistically significant for the fathers. The number of parents rating above the cut off score indicating marital problems was also examined. With the exception of mothers whose children received conventional high-risk cardiac surgery (HR group), there was an overall increase in the number of parents scoring as having marital problems at the follow-up (T2) when compared to the initial assessment (T1). However, this was not significant in any of the groups.

As shown in the following correlations, changes in marital adjustment were not associated with changes in the children's physical status, ideal percentage of weight for height percentage or physical disability:

Physical status: mothers' n=76, r=.05, p=.3; fathers' n=74, r=.00, p=.4

Physical disability: mothers' n=76, r=.09, p=.2; fathers' n=74, r=.02, p=.4

Weight for height: mothers' n=76, r=.01, p=.4; fathers' n=74, r=.06, p=.2

These findings remained the same when each surgical group was analysed separately.

With regard to the parents of children awaiting heart-lung transplantation (PvHL group), only 6 mothers and 5 fathers completed this scale at T2, therefore no reliable conclusions could be drawn from this data.

The degree of deterioration in the parents' marital adjustment ratings was examined. No significant differences were found between the specified group combinations (mothers: F=0.6, df=3,75, p=0.6; fathers: F=0.4, df=3,74, p=0.7).

TABLE XXXIII
CHANGES IN THE PARENTS' MARITAL ADJUSTMENT RATINGS (GRIMS)

		Mean (s.d) **			Poor Marital Relationship (%) *		
		T1	T2	Sign	T1	T2	Sign
Mother's							
HTx	n= 17	3.5 (1.8)	4.1 (2.5)	0.01	17.6%	35.3%	NS
HLTx	n= 15	3.7 (2.0)	4.0 (2.6)	NS	13.3%	33.3%	NS
HR	n= 21	3.5 (2.0)	3.9 (1.8)	NS	28.6%	23.8%	NS
LR	n= 26	3.5 (2.3)	3.6 (2.4)	NS	19.2%	26.9%	NS
PvHL	n= 6	4.0 (1.5)	5.3 (2.0)	NS			
Father's							
HTx	n= 17	3.1 (1.7)	3.8 (2.5)	0.02	5.9%	17.6%	NS
HLTx	n= 15	3.7 (2.2)	4.6 (2.7)	(.06) NS	33.3%	40.0%	NS
HR	n= 21	3.1 (2.3)	4.0 (2.5)	0.01	19.0%	28.5%	NS
LR	n= 26	3.4 (2.2)	3.9 (2.3)	0.04	11.5%	26.9%	NS
PvHL	n= 5	3.4 (0.9)	5.4 (1.7)	(.07) NS			

** Paired T-test

* McNemar Test , Poor marital relationship refers to ratings above the threshold score of 5/6 as suggested in the GRIMS Manual.

4.3.6 Changes in the Family Organisation Measures

Changes in Attitudes towards the Child. The Expressed Emotion Scales (EE)

Differences in parental attitudes between initial assessment and follow-up were found. These are summarised in TABLES XXXIV (page 142).

At the follow-up, the mothers of heart (p=0.004) and heart-lung (p=0.003) transplant recipients, and the mothers whose children received low-risk conventional cardiac surgery (p=0.001) showed less overinvolvement with the ill child than prior to surgery. It is of interest that despite the physical deterioration suffered in the group of children awaiting transplantation (PvHL), these mothers also had a tendency to show less overinvolvement at T2 (p=0.08) than at T1.

The fathers of children in the HLTx (p=0.008) and LR (p=0.001) groups also showed less overinvolvement after surgery than before surgery. Only 3 fathers of children in the PvHL group were available to be interviewed at the follow-up. Therefore, this data was not analysed.

Mothers whose children received high-risk conventional cardiac surgery showed less warmth post-surgery (p=0.05); and the mothers of heart-lung transplant recipients expressed more criticisms post-surgery (p=0.03) than prior to the operation. More critical comments at the follow-up were also expressed by the fathers of children receiving low-risk cardiac surgery (p=0.04). There were no changes in the hostility and positive remark scales between T1 and T2.

There were some differences in the degree and direction of change between the specified group comparisons. This is relevant to: mothers' hostility scale (One-way anova, F=2.73, df=3,75, p=0.04), fathers' overinvolvement scale (F=4.08, df=3,50, p=0.01) and fathers' critical comments (F=2.71, df=3,50, p=0.05) scales. Mother's of heart-lung transplant recipients (HLTx) had become more hostile towards the children than the mothers of heart transplant recipients (HTx) (p=0.04). The fathers of heart-lung recipients had become less overinvolved with the children than the fathers of heart transplant recipients (p=0.01), whilst

fathers of children who received low-risk cardiac surgery (LR) became less overinvolved than the fathers whose children underwent high-risk cardiac surgery (HR) (p=0.02). Fathers of children who received conventional cardiac surgery had became more critical of the child than the fathers of transplant recipients (p=0.008); and the fathers of children who received low-risk cardiac surgery became more critical than the fathers of children who received conventional high-risk cardiac surgery.

Changes in Parents' Attitudes towards their Partner. The Expressed Emotion Scales (EE)

As shown in TABLE XXXV (page 143), mothers of heart (p=0.006) and heart-lung (p=0.001) transplant recipients were less warm towards their husbands at the follow-up. Nevertheless, mothers of the heart transplant recipients (HTx, p=0.05) expressed more positive remarks (PR) at the follow-up than at initial assessment. Similar findings were obtained for the mothers of children who received low-risk cardiac surgery (LR, p=0.008). The fathers of heart-lung transplant recipients were also less warm towards their wives (p=0.006); and the fathers whose children received low-risk cardiac surgery (LR) expressed more positive remarks (PR) towards their wives at the follow up than at initial assessment (p=0.04).

When the degree of change in the parents' attitudes towards their partner were compared in the specified groups, differences in the direction and the intensity were found in both mothers' (One-way anova, F=5.06, df=3,71, p=0.003) and fathers' (F=5.57, df=3,52, p=0.002) warmth scale. Following surgery, parents of transplant recipients (HTx and HLTx groups) became less warm towards their partners than the parents whose children received conventional cardiac surgery (mothers: p=0.01; fathers: p=0.04). Also, fathers whose children received high-risk conventional cardiac surgery showed less warmth towards their wives at the follow-up, whilst fathers whose children received low-risk conventional cardiac surgery showed more warmth towards their partners at the follow-up than prior to surgery (p=0.02).

TABLE XXXIV

CHANGES IN PARENTS' ATTITUDES TOWARD THEIR CHILDREN (EE)

N, means (s.d)

	HTx n, T1 T2	HLTx n, T1 T2	HR n, T1 T2	LR n, T1 T2	PvHL n, T1 T2
Mother	N= 21	N=17	N=17	N=24	N=12
Overinvolvement	2.5 1.9*	2.5 1.5*	1.8 1.6	2.0 1.4*	1.9 1.6
Warmth	4.3 4.4	4.1 4.0	4.4 4.1*	4.2 4.1	4.2 4.3
Hostility	0.0 0.0	0.0 0.3	0.0 0.0	0.1 0.0	0.0 0.3
Positive Remarks	1.5 1.5	2.7 3.1	3.1 2.4	2.6 2.2	2.7 2.7
Critical Comments	0.0 0.1	0.2 1.2**	0.2 1.0	0.5 0.7	0.8 1.0
Father	N=15	N=11	N=12	N=16	
Overinvolvement	2.0 2.0	2.3 1.3*	1.7 1.7	1.8 1.0*	-
Warmth	4.1 4.2	4.3 4.1	4.4 4.1	4.4 4.3	-
Hostility	0.0 0.0	0.0 0.1	0.0 0.0	0.0 0.0	-
Positive Remarks	1.1 1.2	1.6 2.0	3.2 2.8	2.8 1.6	-
Critical Comments	0.0 0.0	0.0 0.3	0.3 0.4	0.1 0.9**	-

*Paired T-test and ** McNemar test significant at the .05 level
Overinvolvement: none= 0; very little= 1; some= 2; moderate= 3; high= 4; mark= 5
Warmth: none= 0; very little= 1; some= 2; moderate= 3; high= 4-5
Hostility: none= 0; present= 1
Positive Remarks: frequency of positive remarks
Critical Comments: frequency of critical remarks

TABLE XXXV

CHANGES IN PARENTS' ATTITUDES TOWARD THEIR PARTNERS (EE)

N, means

	HTx		HLTx		HR		LR		PvHL	
	n, T1 T2		n, T1 T2		n, T1 T2		n, T1 T2		n, T1 T2	
Mother										
	N=18		N=15		N=18		N=24		N=7	
Warmth	3.6	2.8*	3.5	2.1*	3.7	3.3	3.6	3.6	4.0	2.7
Hostility	0.0	0.2	0.0	0.4	0.0	0.0	0.0	0.0	0.0	0.1
Positive Remarks	0.1	0.3*	0.4	0.4	0.1	0.3	0.1	0.5*	0.8	0.4
Critical Comments	0.2	0.5	0.2	0.4	0.1	0.3	0.1	0.3	0.2	1.0
Father										
	N=16		N=11		N=13		N=16		-	
Warmth	3.5	3.0	3.5	2.1*	3.7	3.1	3.6	4.0	-	
Hostility	0.0	0.0	0.0	0.1	0.0	0.0	0.0	0.0	-	
Positive Remarks	0.0	0.3	0.4	0.2	0.1	0.2	0.1	0.4*	-	
Critical Comments	0.0	0.0	0.0	0.3	0.0	0.2	0.0	0.0	-	

* Paired T-test, and McNemar test significant at the .05 level
Warmth: none= 0; very little= 1; some= 2; moderate= 3; high= 4-5
Hostility: none= 0; present= 1
Positive Remarks: frequency of positive remarks
Critical Comments: frequency of critical remarks (analysis of data: none=0; one or more=1)

Changes in Family Adjustment

Changes in family adjustment between the initial assessment (T1) and the follow-up (T2) are shown in FIGURE 21. In this figure, moderately and poorly adjusted families have been grouped together.

Figure 21: Changes in the number of Moderate-Poorly Adjusted Families
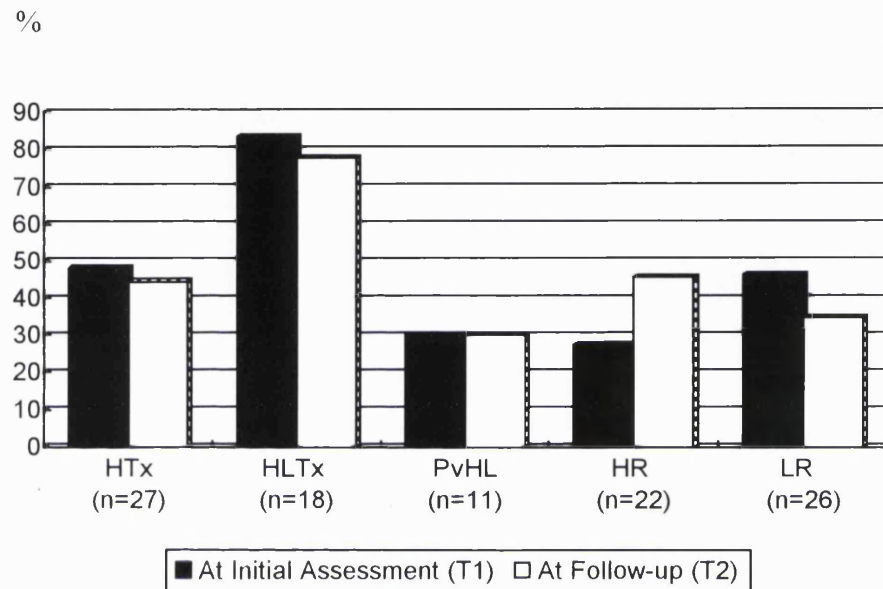
Following surgery, there were no significant changes in the proportion of moderately/poorly adjusted families. This finding holds true when the surgical groups were analysed together (McNemar test, p=0.12), and separately. When the mean scores were examined, some differences in the degree and direction of change were detected between the specified groups (One way-anova, F=3.65, df=3,93, p=0.01). It was found that the functioning of families whose children received heart-lung transplantation showed a tendency towards improvement (T1 mean:2.10; T2 mean:1.90), whilst in contrast the functioning of families whose children received a heart transplant showed a deterioration (T1 mean:1.51; T2 mean: 1.58) (p=0.03). Differences were also found when comparing changes between the two groups of families whose children received conventional cardiac surgery: the functioning of families whose children received high-risk surgery deteriorated (T1 mean: 1.27; T2 mean: 1.45) while the functioning of the families whose children received low-risk surgery showed a tendency towards improvement (T1 mean: 1.53; T2 mean: 1.42) (p=0.01).

Box 3: Changes following Transplantation and Conventional Cardiac Surgery

- A total of 118 families were available at the follow-up. Transplantation and conventional cardiac surgery improved the children's physical health status.

- There was a reduction in the number of children with a psychiatric disturbance and the children's psychosocial functioning also improved. There were no differences between the groups with regard to degree of change in the adjustment measures.

- Parents' psychological adjustment also improved but their marital relationship deteriorated. There were no differences between the groups with regard to degree of change in the parents' psychological and marital adjustment measures.

- Following transplantation or surgery, parents showed less overinvolvement with the children than before. The fathers of children who received conventional cardiac surgery became more critical of the children over time than the parents of transplant recipients.

- The parents of transplant recipients showed less warmth towards their partners over time than the parents of children who received conventional cardiac surgery.

- Following transplantation and cardiac surgery, there were no changes in the percentage of moderate-poorly adjusted families.

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4.4 ASSOCIATIONS BETWEEN PSYCHOSOCIAL FACTORS
AND OUTCOME

4.4.1 Physical and Psychosocial Factors Associated with Survival while awaiting Transplantation

Demographic Characteristics of the Survivors and Non-Survivors

The 39 children who were initially accepted onto the provisional list for heart-lung transplantation (PvHL group) were the focus of this part of the study. All these children had moved to the active list within 16 months from the initial assessment (T1). Twelve children (30.7%) survived at least 17 months from T1, and 27 died whilst waiting transplantation. The demographic characteristics of these children are shown in TABLE XXXVI.

TABLE XXXVI
CHARACTERISTICS OF THE CHILDREN ON THE PROVISIONAL LIST
FOR HEART-LUNG TRANSPLANTATION (PvHL)

	SURVIVORS	NON-SURVIVORS
N	12	27
Mean Age (s.d)	11.7 (4.6)	9.9 (3.7)
Gender	4 males, 8 females	9 males, 18 females
Previous Surgery	3 (25.0%)	12 (44.4%)
Natural Parents	4 (33.3%)	13 (48.1)
Social Class (n, %)*		
I & II	4 (57.2%)	8 (32.0%)
III	-	2 (8.0%)
IV	3 (42.8%)	15 (60.0%)
V	-	-
Unemployed or other	5 (41.7%)	2 (7.4%)

* Groups I & II, and groups III, IV & V were joined together for this analysis
*Registrar General's Classification of Occupations and Coding Index (1980)

Children in the survivors' group were slightly older than the children in the non-survivors' group, but this was not significant (p=0.1). The groups were not different in gender (p=0.9), number of previous operations (p=0.14) or family composition (p=0.14). No differences were found with regard to social class (p=0.23).

Survivors' and Non-Survivors' Physical Health

TABLE XXXVII shows the children's physical health measures.

TABLE XXXVII

SURVIVORS AND NON-SURVIVORS: PHYSICAL HEALTH MEASURES

	SURVIVORS	NON-SURVIVORS	Sign of Diff*
Physical Status	2.7 (0.8)	3.5 (0.7)	0.005
Physical Disability	2.9 (1.1)	3.8 (0.8)	0.03
School Attendance	2.5 (1.0)	1.6 (1.2)	0.03
Weight for Height	84.5 (6.1)	88.5 (10.0)	NS

* T-Tests
Physical Status: 1= good functioning; 4= critically ill
Physical Disability: 1= no disability; 6= needs a wheelchair
School Attendance:1= poor; 5= excellent
Percentage of ideal Weight corrected for Height

Overall, the children who survived had better health at initial assessment and were less disabled than the children who did not survive. School attendance was also better in the former group.

Survivors' and Non-survivors' Adjustment

A series of t-test and standard Chi-square tests were conducted to detect group differences in the children's adjustment measures. There were no significant differences between the groups regarding the children's mental status but differences were found regarding the Global Assessment of Functioning (GAF Scale) measure. See TABLE XXXVIII. The survivors scored higher on psychosocial functioning, obtaining a mean score within the 'no impairment' range (GAF Scale range: 71 - 90), whilst the non-

survivors group mean score indicated mild impairment. This difference between the groups reached statistical significance (t=-4.02, p=0.0001).

TABLE XXXVIII

SURVIVORS AND NON-SURVIVORS: PSYCHOLOGICAL MEASURES

	Non-Survivors	Survivors
Children's Mental Status ICD -9 (n, %)	4/23 (14.8%)	1/11 (8.3%)
Global Assessment of Functioning (GAF Scale) mean, (sd)	*67.3 (11.5)**	*81.8 (9.0)**
Mothers' Adjustment (GHQ-28); n, mean, (sd)	18, 25.0 (12.0)	11, 18.4 (9.1)
Fathers' Adjustment (GHQ-28); n, mean, (sd)	17, 19.5 (10.5)	6, 14.8 (7.1)
Family Adjustment; n, mean, (sd)	27, 1.7 (.7)	12, 1.3 (.6)
Mothers' Marital Adjustment (GRIMS); n, mean, (sd)	17, 3.7 (2.4)	6, 4.0 (2.7)
Fathers' Marital Adjustment (GRIMS); n, mean, (sd)	17, 3.8 (2.2)	6, 3.1 (1.9)
Mothers'/Child Overinvolvement; n, mean, (sd)	26, 1.8 (1.4)	12, 1.9 (.9)
Mothers'/Child Warmth; n, mean, (sd)	26, 4.3 (.9)	12, 4.2 (.9)
Mothers'/Child Hostility; n, mean, (sd)	26, 0.0 (.3)	12, 0.0 (.0)
Mothers'/Child Positive Remarks; n, mean, (sd)	26, 2.0 (1.6)	12, 2.7 (1.1)
Mothers'/Child Critical Comments; n, mean, (sd)	26, 0.7 (1.2)	12, 0.8 (1.0)
Fathers'/Child Overinvolvement; n, mean, (sd)	23, 1.5 (1.1)	6, 2.1 (.4)
Fathers'/Child Warmth; n, mean, (sd)	23, 4.0 (1.0)	6, 4.6 (.5)
Fathers'/Child Hostility; n, mean, (sd);	23, 0.0 (.0)	6, 0.0 (.0)
Fathers'/Child Positive Remarks n, mean, (sd)	23, 1.6 (1.7)	6, 3.0 (.8)
Fathers'/Child Critical Comments; n, mean, (sd)	23, 0.3 (.7)	6, 0.5 (.8)
Mothers'/Partner Warmth; n, mean, (sd)	25, 3.1 (1.3)	7, 4.0 (1.0)
Mothers'/Partner Hostility; n, mean, (sd)	25, 0.1 (.5)	7, 0.0 (.0)
Mothers'/Partner Positive Remarks.; n, mean, (sd).	25, 0.5 (.7)	7, 0.8 (.6)
Mothers'/Partner Critical Comments; n, mean, (sd)	25, 0.3 (.6)	7, 0.2 (.7)
Fathers'/Partner Warmth; n, mean, (sd)	22, 2.9 (1.2)	6, 3.5 (1.2)
Fathers'/Partner Hostility; n, mean, (sd)	22, 0.0 (.4)	6, 0.0 (.0)
Fathers'/Partner Positive Remarks; n, mean, (sd)	22, 0.2 (.4)	6, 0.5 (.5)
Fathers'/Partner Critical Comments; n, mean, (sd)	22, 0.1 (.5)	6, 0.0 (0.0)

* T-test significant at the 0.001 levels

Parent's Psychological and Marital Adjustment

Parents of survivors and non-survivors did not differ significantly in their psychological or marital adjustment (GHQ-28; GRIMS).

Family Organisation

Parents of survivors and non-survivors did not differ in their attitudes towards their children or their partner (EE). Nor were there any differences between the groups in family adjustment.

4.4.2 Psychosocial Factors Associated with Better or Worse Health than Expected following Transplantation or Conventional Cardiac Surgery

Children's and Families' Demographic Characteristics

The demographic characteristics of the children who survived transplantation and conventional cardiac surgery, and their families, have already been described in section 4.5 (see pages 127-129).

Heart Transplantation (HTx) Group

TABLE XXXIX shows that the following pre-operative variables were associated with the children being more healthy than expected at the follow-up: no impairment in the children's psychosocial functioning, parents' positive remarks and high warmth towards the children, and parents' high warmth towards their partner. TABLE XL shows that poor family and marital adjustment at initial assessment were associated with the children being more ill than expected from their physical health pre-transplantation.

Associations between the psychological characteristics at the follow-up and the children's physical health are shown in TABLES XLI and XLII. Again, no impairment in the children's psychological functioning, mothers' high levels of warmth and parents' positive remarks towards the children, parents' warmth towards their partner and mothers' expression of positive remarks about her partner were associated with better health. Nevertheless, hostility between the parents, poor family adjustment, and fathers' poor marital adjustment were associated with the children being more ill than expected.

Heart-Lung Transplantation (HLTx) Group

As shown in TABLE XXXIX fathers' positive remarks at initial assessment was the only variable associated with better health than expected; and mothers' critical comments

regarding her husband was associated with the children being more ill than would have been expected from the children's health before the transplant (TABLE XL).

At the follow-up, mothers' high levels of warmth towards her partner were associated with better health; but fathers' hostility towards mother and both parents' poor marital adjustment were associated with worse health than expected (TABLES XLI and XLII).

Low-risk Conventional Cardiac Surgery (LR) Group

None of the pre-surgery psychosocial variables were strongly associated with better health than expected from the children's health at T1. However, pre-surgical negative attitudes, such as fathers' overinvolvement and parents' critical comments about the children, were associated with the children having a worse medical outcome. The level of significance of these associations are shown in TABLES XXXIX and XL

At follow-up, mothers' high levels of warmth towards both their children and partners were linked to better health than expected. Negative attitudes between family members, such as parents' critical comments about the children, fathers' overinvolvement, mothers' hostility and critical comments about their partners, and poor family adjustment were all associated with worse physical health than expected from the pre-surgery physical health. TABLES XLI and XLII.

High-risk Conventional Cardiac Surgery (HR) Group

With regard to the psychosocial characteristics at initial assessment, it was found that fathers' high levels of warmth towards the children and mothers' high levels of warmth towards their partners were associated with the children being more healthy than expected. Fathers' overinvolvement with the children, and mothers' critical comments about their partners were associated with the children being more ill than expected from the children's health prior to surgery (TABLES XXXLX and XL).

When the psychological factors post-operatively were examined, only absence of impairment in the children's functioning was associated with better health; whilst mothers' critical comments about the children, fathers' poor marital adjustment and mothers' poor

psychological adjustment were associated with worse physical health than would have been expected (TABLES XLI and XLII).

Awaiting Heart-Lung Transplantation (PvHL) Group

An absence of children's psychological impairment, mothers' high warmth and positive remarks about the children, at the time of initial assessment, were associated with better health at the follow-up than expected (TABLE XXXIX); whilst mothers' overinvolvement, parents' critical comments about the children, poor family adjustment and children's psychiatric disturbance were associated with worse physical health than would have been expected at the follow-up (TABLE XL).

Regarding the psychological characteristics at the follow-up, lack of impairment in the children's functioning and mothers' high levels of warmth towards her partner were associated with better children health; whilst mothers' overinvolvement and hostility towards the children were associated with poorer physical health than expected after surgery (TABLES XLI and XLII).

At the follow-up only three fathers were available to be interviewed, so this data was not analysed.

TABLE XXXIX

PSYCHOSOCIAL FACTORS AT T1 ASSOCIATED WITH BETTER HEALTH THAN EXPECTED

	GAF	MW	MPR	FW	FPR	MFW	FMW
HTx	-.43**	-.49**	-.43*	-.77***	-.48*	-.54**	-.66**
HLTx					-.46*		
LR	-.31*	-.31*		-.36*		-.31**	-.32*
HR			-.39*	-.42*		-.41*	
PvHL	-.69**	-.69**	-.64**				

Correlations with residual for regression models (see page 90); Level of Significance: * 0.05, ** 0.01, *** 0.001

GAF= Children's global assessment of functioning
MW = Mother's warmth towards the child
MPR= Mother's positive remarks towards the child
FPR= Father's positive remarks towards the child

FW = Father's warmth towards the child
MFW= Mother's warmth towards father
FMW= Father's warmth towards mother

TABLE XL

PSYCHOSOCIAL FACTORS AT T1 ASSOCIATED WITH WORSE HEALTH THAN EXPECTED

	MEOI	MCC	FEOI	FCC	MFCC	FAMFUNCT	MGRIM	FGRIM	ICD-9
HTX						.51**	.55**	.64**	
HLTx					.44*				
LR	.31*	.42*	.58***	.52**		.38*	.29*		.37*
HR			.46*		.53**				
PvHL	.63**	.48*		.90**		.55*			.63*

Correlations with residual for regression models (see page 90); Level of Significance: * 0.05, ** 0.01, *** 0.001

MEOI= Mother's overinvolvement with child
MCC = Mother's critical comments about the child
FEOI= Father's overinvolvement with the child
FCC = Father's critical comments about the child
MFCC= Mothers' critical comments about father
FGHQ= Fathers' psychological adjustment

FAMFUNCT= Family adjustment
MGRIM   = Mothers' marital adjustment
FGRIM   = Fathers' marital adjustment
ICD-9   = Children's mental status
MGHQ   = Mothers' psychological adjustment

TABLE XLI

PSYCHOSOCIAL FACTORS AT T2 ASSOCIATED WITH BETTER HEALTH THAN EXPECTED

	GAF	MW	MPR	FPR	MFW	MFPR	FMW
HTx	-.58**	-.63**	-.50**	-.46*	-.60**	-.56**	-.77***
HLTx					-.46**		
LR	-.35*	-.42**			-.40*		
HR	-.50**						
PvHL	-.56*				-.49***		

Correlations with residual for regression models (see page 90); Level of Significance: * 0.05, ** 0.01, *** 0.001

GAF= Children's global assessment of functioning   MFW = Mother's warmth towards her partner
MW = Mother's warmth towards the child   MPRP= Mother's positive remarks about her partner
MPR= Mother's positive remarks about the child   FMW = Father's warmth towards his partner
FPR= Father's positive remarks towards the child

TABLE XLII

PSYCHOSOCIAL FACTORS AT T2 ASSOCIATED WITH WORSE HEALTH THAN EXPECTED

	MEOI	MCC	MH	FEOI	FCC	MFCC	MFH	FMH	FAMFUNCT	MGRIM	FGRIM	MGH
HTx							.47**	.77***	.52**		.52**	
HLTx								.74**		.48*	.55*	
LR		.44**		.63**	.56**	.40*	.58**		.45**			
HR		.53**									.40*	.63**
PvHL	.67**		.61*									

Correlations with residual for regression models (see page 90); Level of Significance: * 0.05, ** 0.01, *** 0.001

MEOI= Mother's overinvolvement with child
MCC = Mother's critical comments about the child
MH = Mother's hostility towards the child
FEOI= Father's overinvolvement with child
FCC = Father's critical comments about the child
MFCC= Mother's critical comments about father

MFH   = Mother's hostility towards father
FMH   = Father's hostility towards mother
FAMFUNCT= Family adjustment
MGRIM = Mother's marital adjustment
FGRIM = Father's marital adjustment
MGH   = Mother's psychological adjustment

Box 4: Associations between Psychosocial Factors and Medical Outcome

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SURVIVORS / NON-SURVIVORS

- Children who survived awaiting transplantation were slightly older than those children who did not survive.

- The survivors had better physical health status, less disability, better school attendance and better psychosocial functioning at the initial assessment than the non-survivors.

BETTER/WORSE PHYSICAL HEALTH THAN EXPECTED

- No presence of impairment in the children's psychosocial functioning and a positive family organization characterized by expression of warmth and positive remarks were associated with better health than expected.

- A psychiatric disorder in the children, poor family and marital adjustment, mothers' poor adjustment, and a negative family organization characterized by high overinvolvement with the patient, and the presence of criticism and hostility between family members, were associated with worse health than expected.

- There was variation across the groups with regard to the number of psychosocial factors associated with better/worse health than expected.

4.5 ASSOCIATIONS BETWEEN PSYCHOSOCIAL FACTORS AND ADHERENCE TO TREATMENT FOLLOWING TRANSPLANTATION

Demographic Characteristics of the Children participating in the Adherence Study and their Families

The characteristics of the children included in this part of the study, and their families, are presented in TABLE XLIII.

TABLE XLIII

ADHERENCE STUDY: CHILDREN'S AND FAMILIES' CHARACTERISTICS

GENDER	**32 Males (60.3%)** **21 Females (39.6%)**
AGE (mean, sd)	**10.2 (5.0)**

AGE RANGE (n, %)
    under 5 yrs
    5yrs to 9.11 yrs
    10 yrs and older
	
	8 (15.0%)
	10 (18.8%)
	35 (66.0%)

HEART RECIPIENTS (HTx) **HEART-LUNG RECIPIENTS (HLTx)**	**29 (54.7%)** **24 (45.2%)**
1≥ Previous Surgery; n (%)	**23 (43.4%)**

FAMILY COMPOSITION n (%)
    Natural Parents
    Reconstituted Family
    Single Parent Family
	
	39 (73.6%)
	9 (16.9%)
	5 (9.4%)

No. Siblings in Family
    Range
    mean (sd)
	
	1 - 5
	2.3 (0.9)

No. Families with 2 or more chronically ill children **No. Families in whom a child had died**	 **8 (15%)** **7 (13.2%)**

SOCIAL CLASS (n,%)*
    I & II
    III
    IV & V
	
	15 (33.3%)
	9 (20.0%)
	21 (46.7%)

Unemployed or other	**8 (15.1%)**

* Groups I & II, and groups III, IV & V were joined together for this analysis
*Registrar General's Classification of Occupations and Coding Index (1980)

Fifty-three children who survived at least 9 months post-transplantation were included in this study. The heart-lung transplant recipients were significantly older (age range: 3 yrs-17 yrs.5 mths; mean: 11.6) than the heart transplant recipients (age range: 1 mth -17 yrs; mean: 9.0) (t=-1.97, p=0.05). There were no sex differences between the 2 groups of recipients (X^2=9.28, df=1, p=0.59); nor were they different regarding the number of previous operations (p=0.60), family structure (p=0.67) or social class (p=0.45). However, a significantly greater number of recipients in the HLTx group had lost a sibling (X^2=5.32, df=1, p=0.02) or had a sibling with a chronic illness living in the household (X^2=5.21, df=1, p=0.05) when compared to the recipients in the HTx group.

Levels of Adherence to Treatment

Figure 22: Levels of Adherence to Medical Treatment (%)
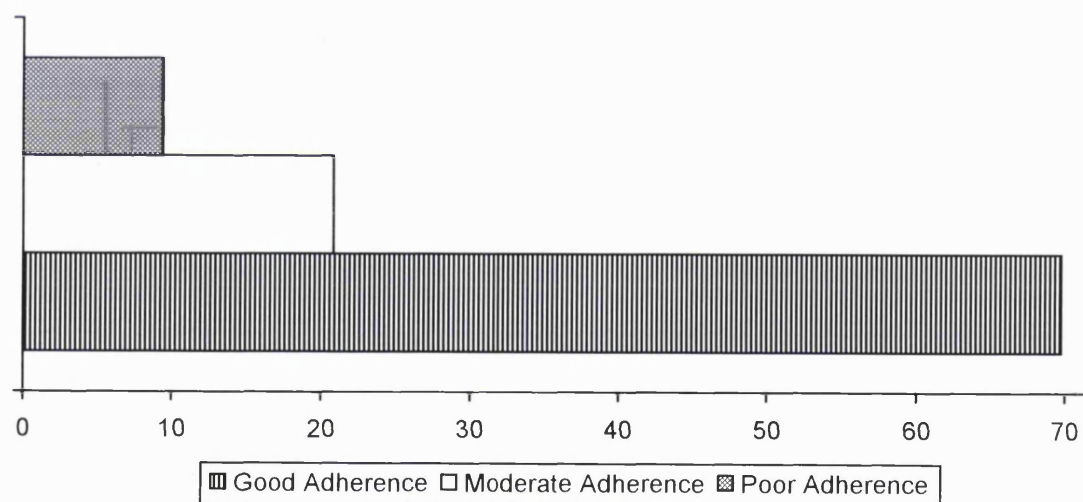
Thirty seven children (69.8%) had good adherence, 11 children (20.8%) showed moderate adherence (unsatisfactory diary completion), and 5 children (9.4%) showed poor adherence (unsatisfactory CyA levels).

Demographic Factors Associated with levels of Adherence

No significant associations were found between adherence to treatment and children's sex (p=0.99), number of previous operations (p=0.60), social class (p=0.16), having a chronically ill sibling (p=0.12) or having lost a sibling (p=0.12). A summary of the children's age, gender, and level of adherence to treatment is presented in TABLE XLIV.

TABLE XLIV
ADHERENCE LEVELS, CHILDREN'S AGE AND GENDER

	Good n=37	Moderate n=11	Poor n=5
Under 5 years **Gender**	7 (18.9%) 5 m, 2 f	1 (9.1%) 1 m	- -
5 to 9.11 years **Gender**	7 (18.9%) 2 m, 5 f	3 (27.3%) 1 m, 2 f	- -
10 years & older **Gender**	23 (63.2%) 16 m, 7 f	7 (63.7%) 5 m, 2 f	5 (100%) 3 m, 2 f

Nor was age correlated with adherence when the three groups were compared, although all children failing to take the immunosuppressant therapy were 10 years of age or older (One-way anova, - age by adherence: F=2.16, df=2,50, p=0.12). When this data was re-analysed comparing the group of children who seem to miss medication against the two groups of children who had good and moderate adherence, it was found that the former group had a mean of 13.8 years of age, whilst the latter group had a mean of 9.9 years of age. This difference between means reached statistical significance (p= 0.01). Clinical information gathered in the interviews suggests that the older recipients had more concerns about the changes in their physical appearance due to the side effects of the medication than younger recipients.

Family composition was associated with unsatisfactory adherence (X^2=5.65, df=2, p=0.05). This is shown in FIGURE 23. Children living with both their natural parents had better

adherence than children living with single or reconstituted families. However, this

Figure 23: Family Composition and Adherence

%
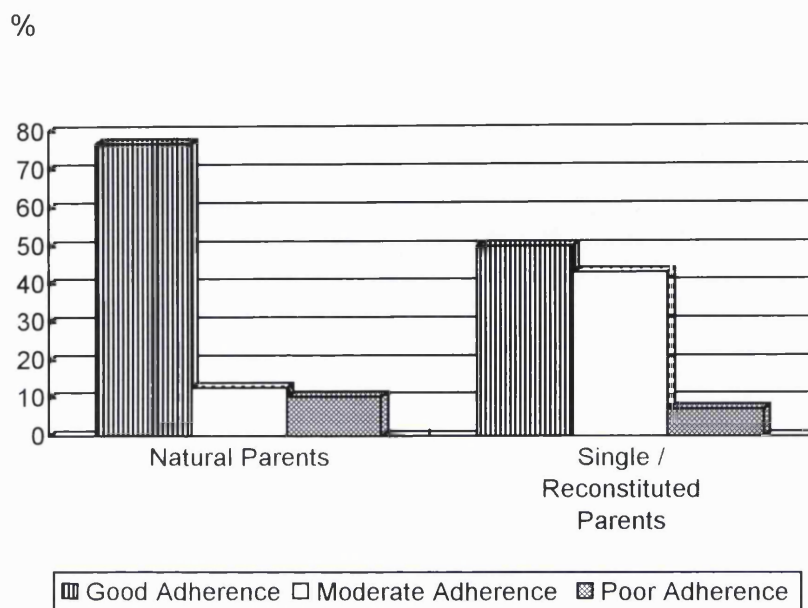
association vanished when the HTx and HLTx groups were examined separately. It was also found that heart transplant recipients were significantly more adherent than heart-lung transplant recipients (X^2=8.2, df=2, p=0.01). This finding is summarised in FIGURE 24.

Figure 24: Type of Transplantation and Adherence

%
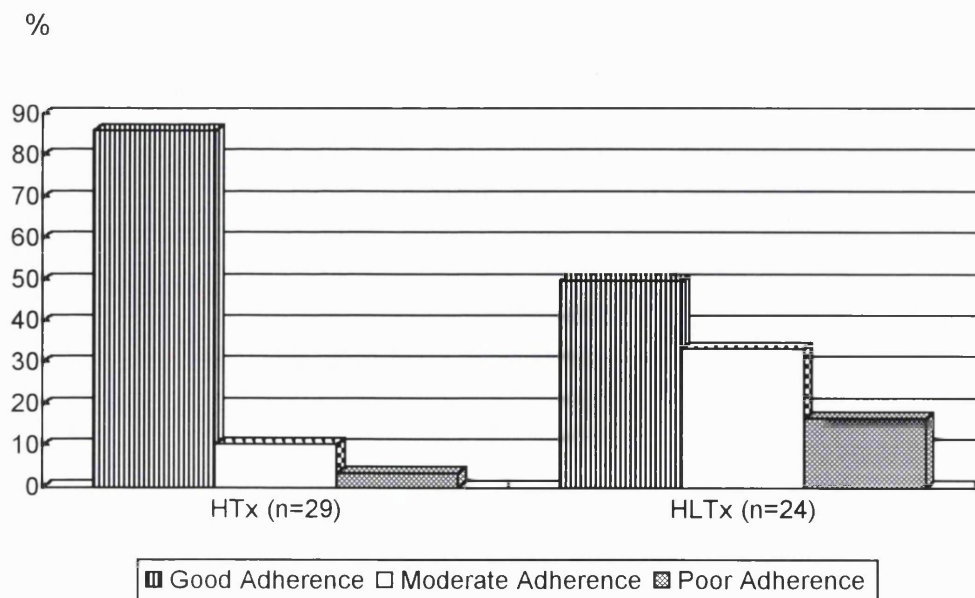
Children's Adjustment and Levels of Adherence

There were no statistical associations between children's mental status (p=0.74) or psychosocial functioning (p=0.15) at initial assessment (T1) and adherence to treatment. These findings held true when the HTx and the HLTx groups were analysed separately and together.

Parents' Psychological and Marital Adjustment and Levels Adherence

No significant associations were found between adherence and parents' psychological (mothers': p=0.5; fathers': p=0.6) or marital adjustment (mothers': p=0.14, fathers': p=0.14).

Family Organisation and Levels of Adherence

Detailed analysis of parental attitudes towards the children showed no association with adherence, with only two exceptions. Heart recipients, but not heart-lung recipients, had better adherence if their mothers showed high levels of warmth towards them (X^2=5.8, df=1, p=0.01), and independently, absence of hostility (X^2= 6.9, df=1, p=0.008). These findings are shown in FIGURES 25 and 26.

Figure 25: Mothers' Warmth (HTx group) and Adherence

%

Low Warmth (rated 0-3)   High Warmth (rated 4-5)

▥ Good Adherence ☐ Moderate Adherence ▩ Poor Adherence
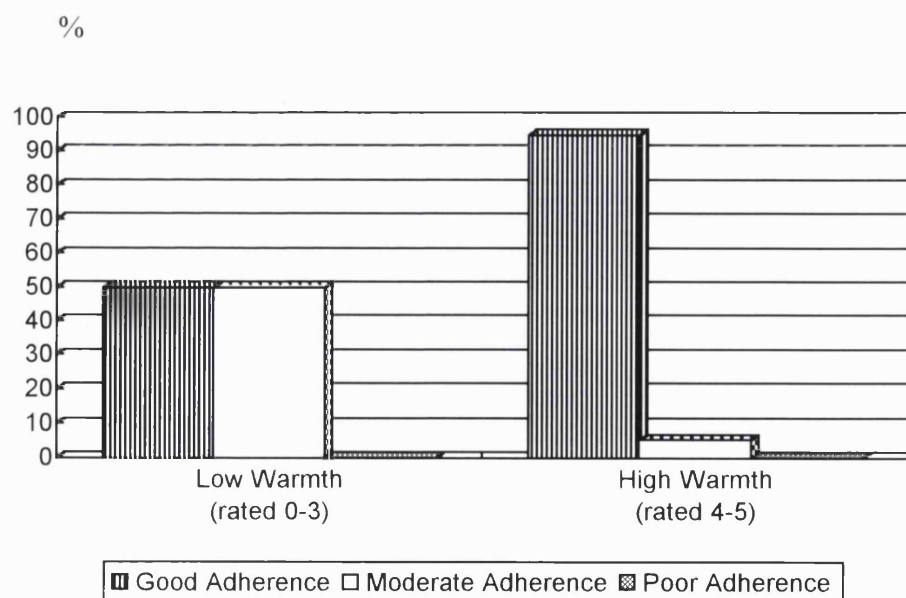
However, due to the small number of mothers' ratings on hostility and low warmth, and to the association vanishing when moderate/poor adherence groups were joined together (Fisher's test), these results should also be interpreted with caution.

Figure 26: Mothers' Hostility (HTx group) and Adherence
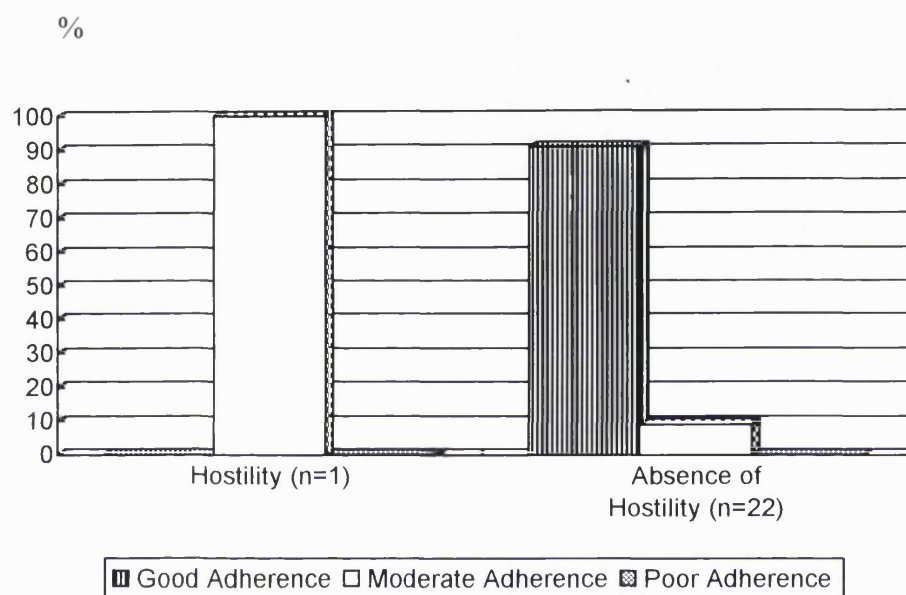
When the two groups of recipients were analysed together regarding parents' attitudes towards their partner, it was found that those children whose mothers (X^2=6.3, df=2, p=0.04) and fathers (X^2=10.3, df=2, p=0.05) were critical of their partner's emotional availability prior to transplantation were less adherent to treatment. When the two groups of recipients were analysed separately, the association was maintained for the mothers of heart recipients (X^2=8.9, df=1, p=0.02), but not for the mothers of the heart-lung transplant recipients (p=0.2). This finding should be interpreted with caution as only two mothers were critical of their partners and the association vanished when the moderate/poor adherence groups were joined together (see FIGURE 27). Regarding the fathers, it was found that the association faded when the groups were analysed separately.

Figure 27: Mothers' Critical Comments -Partner (HTx group) and Adherence

☐ Good Adherence ☐ Moderate Adherence ☒ Poor Adherence
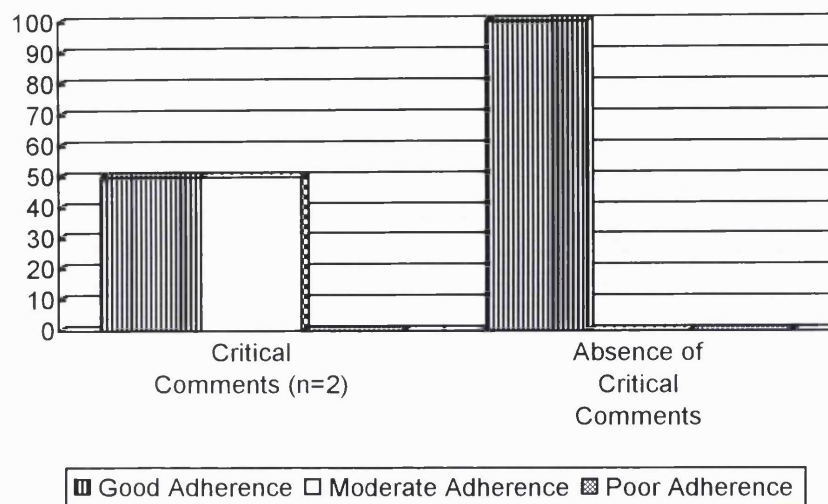
Most heart transplant recipients, but not heart-lung recipients, who had well-adjusted families showed good adherence ($X^2 = 10.2$, df=4, p=0.003). However, this result should be interpreted with caution as there is only one child in the poorly adjusted family group (see FIGURE 28), and the association vanished when moderate/poorly adjusted families and moderate/poor adherence groups were analysed together (Fisher's test).

Figure 28: Family Adjustment (HTx group) and Adherence

%

☐ Good Adherence ☐ Moderate Adherence ☒ Poor Adherence
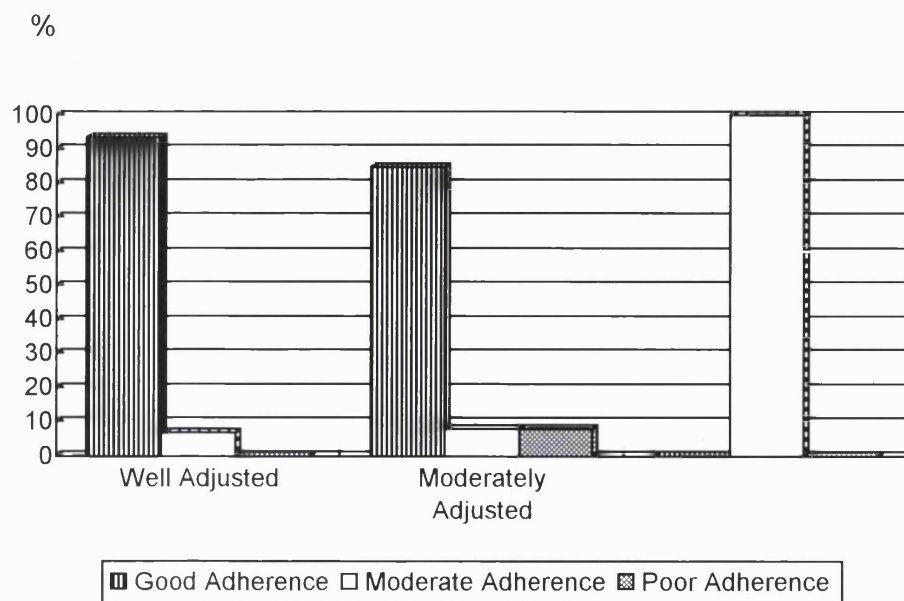
BOX 5: *Adherence, its Prevalence and associations with Psychosocial Factors*

- Overall 30.2% of the children showed unsatisfactory adherence to medical treatment following heart or heart-lung transplantation.

- Heart-lung transplant recipients were more at risk of unsatisfactory adherence than heart recipients.

- Greater number of children living with both natural parents showed good adherence when compared to children living in single or reconstituted families. However, this association was no longer significant when the heart and heart-lung recipients groups were analyzed separately.

- In the heart transplant recipients group, family adjustment, mother's warmth and hostility towards the child, and mother's critical comments about her partner were significantly associated with adherence. Nonetheless, these results should be interpreted with caution.

Chapter 5

FURTHER EXPLORATION OF FAMILY INTERVIEWS

In this chapter, the parents' interviews (CFI, EEPIS and family adjustment assessments) are examined more closely. In the parental interviews, there were three sources of information: a) the content of what was said, b) the observed interaction between the parents, and c) the changes between T1 and T2 elicited at interviews (in both the content and at the interactional level). This information is very valuable for the further development of clinical research studies and for the development of intervention programmes aiming at improving family relationships in this population. In addition, parent-child and parent-partner relationships in families containing young children and adolescents with cardio-respiratory disorders have not been described previously. To provide a structure for this exploration the EE scales and Family Adjustment Assessment measures have been used. All names in this chapter and details that might identify individual families have been changed.

EXPRESSED EMOTION SCALES (EE)

1. Description of Overinvolvement (EOI)

The EOI pattern across the groups was very similar. Parents who obtained a score of two or less on this subscale (indicating "some, little or no" overinvolvement) either had some reluctance about giving to the patient age-appropriate information about the illness and its prognosis, showed over-identification with the patient or gave some evidence of 'enmeshment' in the family. This is illustrated by the mother of Samantha, aged 16 and awaiting high risk cardiac surgery, when she remarked: "I know her inside out", or the father of Anthony who said: "If he cries, I cry; if he is moody, I am moody". Some of these parents could have earned a score of three or more (moderately high to high overinvolvement). However, they provided "alternative information" against EOI such as appropriate expectations for individuation (taking into account the child's physical condition and the limitations imposed by the illness), encouragement for independence and respect for the children's social and private lives.

There was a lot of variation regarding the extent to which parents communicated with the patient and siblings about the illness and forthcoming procedures (see Appendix 1, pages 270-271). The amount of information conveyed to the children was not related to severity of illness or child's prognosis, but to the parents' beliefs, to their ability to support each other consistently, and to share with each other fears about the possible death of the child. Some parents who scored between moderately high and an extreme manifestation of emotional overinvolvement, showed extreme overprotection regarding the illness. The parents of Ramon, aged 13, illustrate this. To the question: How have you prepared R. for the operation?. How much have you told him about the surgical procedures and about the recovery process?. His father remarked: " R. doesn't know that he is going to be operated on within the next 24 hours. We will tell him when the operation is over. Tomorrow, when he wakes up. We don't want to upset him now". These parents had planned very carefully all the practicalities associated with Ramon's admission. However, despite the fact their child's operation was associated with a 30% mortality risk they had never discussed between themselves how they felt about the operation, or the likelihood of things not going as planned.

Paradoxically, some highly overinvolved parents presented themselves as lacking in overprotective behaviours. These parents were not trying to lessen the child's autonomy and independence but, on the contrary, they were pushing their children to do things without acknowledging the child's illness. These parents were 'devoted' to deny the illness, and the major aim of their life was to prove that their child was 'normal'. They acted as a buffer between the patient and the illness. James, aged 14, was diagnosed with CF at the age of 2. He was admitted in acute respiratory failure. This was his first admission. His mother said: "This is a big shock for him. We never told him he had C.F. We have never talked about it at home... He has never read anything about CF. Every morning, my husband and I check all newspapers and the TV guide looking for articles or programs about CF, and we make sure that James doesn't have access to them. He has never met anyone with CF. For us, he has always been normal, healthy. Just like his brother!. We wanted him to grow up having a normal life. He has had all sort of experiences: climbing, parachuting, motor racing. He has travelled around Europe and America with friends. We allow him to do anything. We are always organising new and exciting things for him to do. He goes to night-clubs

every week. Sometimes we are up all night waiting for his call asking us to fetch him". Father interrupted saying with pride, "he has done more things than any other kid of his own age. I work three times harder than any other father to keep up his high life-style. I'm killing myself, but don't mind. No kid of his age has done as many things as he has".

Other parents were very open in their communication about the illness and possible death of their child. Paul, aged 14.5, was previously healthy until he developed an acute cardiac condition due to a viral infection requiring a heart transplant. His mother was interviewed within two weeks following the diagnosis. When she was asked about how much Paul knew about his condition, she replied "Paul knows everything, he is well prepared. We have read and discussed all the information available. He knows that if he doesn't get a new heart he will be dead in two months. Last week he told me: Mum, I'll be ok. if I die, but what would you do?. We had a very long conversation about it.... Then his brother arrived home and he (the brother) got very upset listening to our conversation, but we kept talking. The three of us cried together and cuddled each other. This is a very difficult situation, extremely painful...(mother cries), but we have to talk about these things together ...before is too late". Other parents, whose children were awaiting transplantation, had discussed with the children what they should do with their toys and belongings if they were to die.

For some parents the word transplantation was like 'magic'. The mother of Yvonne gave us an example when she said, "The chance of the transplant gave me a new life. It is very important to me, and to Yvonne. The fact that the transplant came on the scene saved my life. It saved my sanity anyway!".

There was variation regarding the extent to which parents took into account their children's own feelings about transplantation prior to deciding whether to put the child through a transplant or to leave the illness to follow its own course. The parents of Kate who was 8.9 years old, said: "We don't want to put her through a transplant if she doesn't want to. She is very mature for her age and has to decide what is best for her. We have to accept her decision... it would be very painful for us if she decides that she does not want to be transplanted, but we can not impose a transplant

on her". Low overinvolvement and high warmth was also clear when the father of Kate said: "We don't want to keep her alive just for our sake. We would rather she has a short good life than a long miserable one". The parents of Anton, who was 11 years old and had previously lost a sibling from the same disease, had a different approach. Mother said: "... He feels very negative about the transplant. We know he prefers to be left to die, but he has no choice. We owe it to his brother. His brother never had this opportunity".

For some families "togetherness" was important. Often parents who were highly overinvolved with their child were isolated and lacking 'social life'. These parents also suffered from separation-anxiety as illustrated in the following cases. The mother of Sibille, a 7 years old girl with CF, said: "I haven't had any social life since Sibille was diagnosed 6 years ago. I have lost all my friends... I'm afraid that if I go out, something will go wrong". The mother of Robert, who was undergoing conventional cardiac surgery, said: "We are a very close family, we do everything together. Some parents leave their children with grandparents or babysitters. Robert is now 13 years old and we have never ever separated from him. Only when he is at school. We are very close as a family... we don't feel comfortable if he is not with us. Robert is not the sort of boy who likes to be with friends; actually, he hasn't got any friends,... he prefers our company... at night Robert comes to our bed to sleep with us". The mother of Mary, aged 8.5 and who had tetralogy of Fallot, said: "We are together 24 hours a day. She does not go to school, Mary has a home-tutor. I don't like her mixing with other children, or going to other children's houses. ...Dr. P.R. has told me that she should go to school, but I prefer to have her at home...I don't think she misses the company of other children...".

Parents who scored low on the EOI scale were more aware of the healthy siblings' needs, and they treated the sick child with similar levels of discipline and control as the healthy siblings. The parents of Martha, aged 11 with C.F., noted, "We both treat her like a normal girl. She has to help in the house like her sister does. We tell her off and punish her just like her sister. We make no allowances for her temper and bad behaviour". In contrast, when talking about Christmas presents, the father of Josephine, who was 12 years old and also had C.F., said, 'we always spend more money on Josephine than the others. We find it difficult to say 'no' to her. She always gets

what she wants. This year we spent over £100 on her Christmas presents, and we spend a maximum of £10 on each of her sisters (he laughs). Regarding discipline it is the same old story: Josephine can get away with murder! We are quite strict with her sisters...on the whole we give her many more privileges, we treat her very differently. Her sisters are very jealous, but that's how it is. Josephine has a heart condition. Her sisters are healthy ".

Changes in Overinvolvement

Overall, as the child's physical health improved there was a reduction in parents' overinvolvement. At the follow-up, parents showed less self-sacrificing behaviour, and spent more time thinking about their own leisure activities and social life (see Appendix 1, pages 272-275). Usually, following surgery, illness issues were only discussed in families whose children had unsuccessful operations and/or the child's medical management was unclear.

Most parents described the experience of the child's surgery as 'traumatic' and they showed a resistance to talk about it at the follow up. Often, parents who were rated high on the EOI scale at the initial assessment showed exaggerated emotional responses during the follow-up interview when describing what happened on the day of the operation. However, most of these parents also expressed appropriate expectations for individuation and autonomy in their sick child.

Despite the dramatic change in the child's physical health, a few parents who showed high overinvolvement during the initial interview (T1) were also highly overinvolved at the follow-up. The mother of Thomas, aged 17 illustrates this, when she said, "I do everything for him. I even prepare his bath and help him to wash his hair. I give him the tablets four times per day, I fill the medical diary for him every night. ...He knows which tablets he has to take, and how to fill the diary in. That's not a problem, but I have always done everything for him and I don't see why things should be different now... He is now over 17 and he is very healthy since his transplant, but I am still his mother. I am just doing my job. I don't expect him to do anything. I have nothing else to do but look after him... I never go out without him. ....Oh, no I don't have time for friends, Thomas takes all my time".

Other parents acknowledged difficulties adjusting to a less demanding child. The mother of Terry was also rated as moderately high EOI when first interviewed. At the follow-up she remarked: "It has been a difficult year. During 8 years I did nothing but look after Terry ... appointments, tests, admissions etc. ... I used to go to his school to feed him. It gave me comfort. All my life was around him. I didn't have time for the girls. ... Now he doesn't need me. He doesn't want me to do things for him. It hurts!... it really does!. After the operation he rebelled against me. He wants to be independent. I know he is right!, but I find it difficult. I am not used to it. Our relationship has changed. And that's difficult for me. I wake up in the morning and I feel empty. ...I know I still interfere too much with Terry's life. I know I have to let him go, but it isn't easy. Perhaps, I should be looking for a job".

There were also examples of relatives who showed evidence against overinvolvement. For example, the father of Kate, a 15-year-old who underwent heart transplantation said: "She is now physically well, but she is still missing out a lot. I would like her to be more sociable, to go out more often, to have fun. Kids of her age go to parties... dancing... have fun. I would like her to be more outgoing... I don't know for how long she is going to be around. It would be nice if she enjoys life while she can, before things start going wrong again." Charles's mother felt differently about her 16-year-old son who received a heart transplant. She remarked, "When he is out, I can't cook, I can't go shopping, I can't do anything. I am confined to home waiting for his friends' to call and tell me that he has been taken to hospital, or that he has died. When I hear the police or the fire brigade sirens, I always think that they are coming to my house with bad news. ... Since he had the transplant, I can't relax. I don't want him to go anywhere. I want him with me all the time".

2. Description of Warmth (W)

On this scale, a rating of one or less indicates 'very little warmth or no warmth'; a rating of two is indicative of 'some warmth'. A rating of three denotes 'moderate warmth', whilst a score of more than three is regarded as 'moderately high and high warmth'. The importance of tone of voice for the rating of this scale makes it very difficult to provide case examples.

Parents scoring two or less on this scale were very unemotional about their child 's condition and possible death. They also showed lack of both sympathy and understanding regarding the child's feelings about his/her illness. Most of these parents had great difficulties describing their child as 'a person', and they were unable to mention even one activity that they enjoyed doing with their child. Simon's father expressed a lack of affective feelings about his 8-year-old son through the interview. When answering the question 'What is the most disturbing aspect of Simon's illness for you? He replied, "I can't play football with my friends. After work, I have to go home to help my wife with the physiotherapy. I hate it! I really do! I have lost my friends because of him!" Throughout the interview, the father of Simon seemed very detached from his son, showing no evidence of warmth.

Overall, parents who rated as 'moderate' on warmth scale were definitely sympathetic, understanding and caring about the child. However, there was an absence of evidence regarding the parent's interest in the child as a person and/or no enjoyment of the child's company was described. Moderate warmth was shown by Olivia's mother who, describing her feelings towards her 6 year old sick daughter and her other children, said, 'I care about her, but I don't feel the same warmth that I feel towards her siblings. I don't love her as much. I care for her. I feel sorry to see her like that. I really hope she gets a transplant. It would be nice to see her enjoying life".

Parents who were rated as 'moderately high or high warmth', clearly showed an interest and enthusiasm for the child's activities and achievements, they were concerned and sympathetic regarding their child's physical condition and gave clear evidence of enjoyment of the child's company. The parents of Alex said, "She is a joy to be with. She's got a special little character, everybody who knows Alex, loves Alex. Once you know her you never forget her. It is her nature, she's got a lovely character. We adore her!. Obviously, she is our child. She is a joy to be with. That's my opinion". Father said, "Not much I can add to that. She's very close to me, she is the apple of my eye. She's got that little thing that brings us together. I don't know what it is. There is a big strong bond between me and Alex. We both feel very glad to have been her parents. She's given us so much in the time we've had her. We just feel very privileged to have been her parents". The parents of Karin who was 8 years old, also gave evidence of high warmth

and low EOI in the following statement, "It has been wonderful to have her. It's been absolutely great!. We really enjoy every minute with her".

Warmth between the parents: Parents who expressed low warmth about their partner found it difficult to talk to each other about the implications of surgery. They also found it difficult to share feelings with each other. Often they expressed dissatisfaction with the support and availability given by their partner or they denied the need to be comforted by the other. Nearly all cases that were rated between 'none' and 'moderate' on warmth towards their partner also scored on the critical comment scale. In addition, a few of these parents also rated in the hostile scale. Mrs Roberts, whose 12 year old daughter was awaiting heart-lung transplantation, said:"I get more affection and sympathy from my friends. I can talk to them freely. He (her husband) doesn't want to know about my feelings...(she laughs) I can't remember the last time he tried to comfort me". The mother of Cyril, aged 6 who was undergoing high-risk cardiac surgery, remarked, "We never do things together. We never go out the two of us. We do not talk to each other. We sit together to watch TV., but we rarely talk. We are two separate people. We have nothing in common. We share nothing".

Couples expressing high warmth towards each other were aware of each others needs. During the interview, these couples kept eye to eye contact and invited each other to participate in the conversation. Ben's mother remarked, "I am concerned about Ben, but I am also concerned about John (her husband). I know how much he is suffering with Ben's operation... He is my strength (John). He is such a wonderful man... He usually knows how I feel, and he is always there for me. He is loving, very loving. I enjoy having him around. I am lucky in that respect".

Changes in Warmth

Overall, there was very little change in parental warmth towards the child between the initial assessment and the follow-up. All parents who showed 'moderately high' or 'high warmth' at T1 also showed the same amount of affection at T2. None of the parents who were rated as expressing 'none', 'little', or 'moderate' warmth towards the child at the initial assessment were rated as showing 'moderately high' or 'high warmth' at the follow-up.

Warmth between the parents: The parents' ability, or lack of it, to support each other through the child's admission for surgery seemed to be mediating both the positive and negative change in the amount of affection shown between the parents. This is illustrated in the following examples:

Arnon's mother said, "We couldn't talk to each other at the time of the transplant. It was very disappointing. He wasn't there for me. Since then our relationship has changed. Now, when he goes to work I hope he doesn't come back". The mother of Katty remarked, "He didn't help me. It was the most difficult time of my life. Now we are separating". The mother of Mike said, "When Mike was taken to the theatre I thought I would never see him again. I turned to James (husband) in tears. I needed him to hold me in his arms (she cries)... but he walked away. He left me there. Alone!. Now I feel nothing for him. I would like to forget what happened between us when Mike had his operation, but I can't. I cry every day since, ... I often think about dying".

Couples who showed an increase in warmth at the follow-up perceived each other differently after the child's operation. Mrs Gibson said: "At the time of the transplant, I needed Peter (step-dad) more than anything else. You don't do these things on your own. He was absolutely wonderful!. That experience brought us even closer". Mr Donoban, whose son underwent high-risk cardiac surgery also said, "you cope with the illness and surgery as a pair, not as individuals. We were there for each other, and we became even closer. We are now much more loving to each other. The operation was very upsetting for both of us, but I have to say that it had a very positive effect on our marriage". The father of Candy, who was 6 years old and underwent low-risk cardiac surgery, remarked, " Thanks to Candy's operation we have discovered each other. We are in love again". Candy's mother added, "I discovered new aspects of Dennis (husband). I never thought he could be so loving and caring. We both were feeling very vulnerable and frightened of losing her. When Candy was taken to the theatre, the feelings were so strong and painful!. I felt completely alone, and unable to cope with it. Then we started to share it. It was like a miracle!. I was not alone anymore. It is difficult to describe the comfort we got from each other. The experience is absolutely wonderful. We had a good relationship before, but now..., I don't know. Even our sexual relationship has changed. We are deeply in love".

3. Description of Critical Comments (CC)

A parent's mere recognition or description of characteristics of the patient, however unfavourable, is never in itself sufficient to define a statement as critical, unless a critical tone is present (Leff and Vaughn 1985). The families in this study are characterised by low levels of CC. The majority of the children were described as very well behaved, very caring and loving; often 'perfect' children.

In families containing a child with CF, the content of the CC regarding the child was often related to treatment management, as illustrated in the following examples: Ian's mother said, "The only thing he doesn't moan about is his physio. The rest of his treatment he moans: about the nebulizer, moans about medicines. It can be very annoying at times". The mother of Yvonne remarked, " She is brilliant taking her tablets, but the thing that bothers me most is when she doesn't eat, or doesn't do her physio properly. Then, I really get annoyed!. That really annoys me!... I would like her to take responsibility for the treatment. I can't trust her. I've got to keep checking she's done it. That annoys me. I say: why can't you help yourself and that will help me". Sam's mother also felt upset about the treatment when she said, "Having to remind him about the medications annoys me!. I get worked up because it's a routine you've got to stick to. This skipping and skipping is going to do him no good. Physiotherapy annoys him as well. It is annoying!. The problem is that I like it done here and now, whereas he takes his own time". Johnny's mother also said, " Food is always a struggle. I know he is not hungry, but if he doesn't put on weight he won't have a transplant. He is very reluctant to eat. He drives me mad!".

Some parents, whose children were too ill to attend school and were confined to the home most of the time, expressed some unhappiness about how the child was spending his/her time at home. Coming back to Yvonne's mother again, she said, "Sometimes she'll laze about all day which annoys me. She'll sit in front of the TV and won't want to do anything". Also, some parents seemed to feel that they were not getting back as much as they were giving to their child. Yvonne's mother said, "If she finds that Fred (husband) and I are having an argument, she will always take his side. Sometimes I get annoyed and think 'you little sod, you!'. I spend all my time

making sure you have everything... It does annoy me, but I do understand it". The mother of Kate said," It's annoying to cook a meal and then she'll only eat half of it".

Some parents were critical about disobedient behaviour, untidiness, social life, sibling relationship and how the ill child used the illness to get privileges. This is illustrated in following statements: Samuel's father said about S. "He is lazy and he is cheeky too. That gets me angry". James' mother said, "It is frustrating when he won't take any notice of the things I say, unless I shout. I don't expect a lot of help from him, but I do expect him to keep his room tidier than he does. I have to nag him all the time". The mother of Luig who was undergoing low-risk cardiac surgery, remarked " He goes upstairs and forgets what he is meant to be doing and starts arguing with his brother. Every day!, every morning!. It drives me mad!. I am constantly nagging. I say to myself 'I am not going to shout at him today' and then I am yelling every half an hour!. I don't know how people cope without yelling at their kids... Just lately he is breaking the rules all the time. He used to be quite good but lately he is damaging things, tearing things, he smashed all my pot plants down. I know children are meant to be naughty, but he is especially naughty, just a horror!. It drives me mad!". Marie's mother said about Marie and her relationship to her friends, "She'll invite her friends around, and they'll come and she's so bossy!". Manuel's mother said about his social life," He is not a really good mixer. He is a loner, not an easy child for other children to get on with. He likes adult company unfortunately". The mother of Allan said, "I think he uses his illness to get results which he couldn't get otherwise. He probably manages to get special treatment within the family by using an inability to walk very far or do things by himself. He doesn't like walking anywhere. So as soon as we go out, he starts to wheeze. He gets his brother very cross because he feels he gets away with things that he is not allowed to get away with. He is 14 and very immature, although he is bright."

The frequency of *critical comments between the parents* at the initial assessment was also relatively low. The content of these statements were directly related to the help offered, or the lack of it, with the child's treatment and house chores; or to different ways of dealing with the children and discipline issues. The following examples illustrate these sort of statements: Melanie's mother said," He leaves all the hard work to me. I could do with a little help from him. He never

comes to the hospital. He can't cope with it. I have to decide, alone, whether Melanie should have a transplant or not. He says that I am the mother, and it is up to me and Melanie to decide. He is washing his hands of the situation. It's a big decision, it's not fair on me!... He doesn't help with the physiotherapy either. I have to do it with both Melanie and her younger sister. There are two kids with C.F. in the house, to bring money home is not enough. Sometimes I get very annoyed". Mrs Frost whose boy was undergoing low-risk cardiac surgery, remarked about her husband, "He is too soft. Andy needs a bit of discipline. He never says 'no' to him. Even after I have said 'no', he contradicts me and says 'yes'. Andy has learnt the game and he always goes to his dad. He knows he is getting his way. I go mad at him (husband) sometimes. We are always arguing about it. He is damaging Andy".

Changes in Critical Comments

Parents were more critical of the child at the follow-up. There are several alternative explanations for this. It could be that because the child was healthier, the parents were less protective and felt less guilty about expressing unfavourable comment about the ill child. However, it could also be argued that the increase in the number of parents' critical comments was related to the parents' difficulties adjusting to the rapid changes in their child's behaviour and attitudes to life after successful surgery. The content of those critical comments was as follow: the mother of Anthony said, "Since the transplant, Anthony seems to play up. He was not like that before. It's infuriating". The mother of Steven also said, " He feels good now and he is getting very cheeky. Obviously, he feels a lot of aggression and before he didn't have the energy to let it out. I know he's got every right to be crabby, but it cracks me up. Just the tone of his voice, - no respect there. He angers me, he really does!. Neil's mother said, "It is frustrating having to push him when you know he's got more ability and energy than he is using".

On the whole, following the child's surgery, the number of *critical comments between the parents* also increased. This increase of CC was directly related to dissatisfaction with the partner's support and ability to give comfort at the time of surgery. This dissatisfaction has already been

described above, under changes in warmth. In a few cases, the parents were still very resentful about their partners and subsequently scored in the hostility scale, which is described below.

4. Description of Hostility (H)

A highly critical relative is not necessarily hostile. Hostility is considered to be present when the patient is attacked for what he or she is rather than for what he or she does. Generalisation of criticism and rejecting remarks are the criteria used for the rating of this scale (Leff and Vaughn 1985).

The following responses to the question 'What are the most disturbing aspects of (child's name)'s illness for you?' reflect the presence of one or more of these qualities: The father of Sean, aged 7, remarked, "We argue a lot about the treatment (he and his wife). Neither of us likes to do it, we both try to avoid it. We are always fighting and blaming each other. It is becoming unbearable! Things would be better if Sean dies". The mother of Geraint, aged 9, replied, "I feel embarrassed walking in the street next to him. People look at him and wonder what's wrong with him. I don't like it. I often pretend I am not his mother.... if I can, I avoid taking him out". The mother of Kelly, aged 8, remarked, "I don't like her being too close to me, her breathing annoys me... I never sit next to her". Later on in the interview and regarding adherence to medical treatment, she said, " A couple of times I've got really stroppy with her: 'if you are not going to have your treatment you are going to die anyway, so what's the point?". Another mother said about her 15 years old daughter with cystic fibrosis, "I don't want her near me, I can't stand her when she coughs". When the mother of Odette, aged 10, was asked: 'Are you satisfied with the amount of affection that Odette shows towards you?, she responded, "Oh, yes, she is very affectionate. Too much. She would like me to cuddle and tickle her, like I do to her brother and sister. I just can't do it to her. She is so skinny!. When I touch her, I feel yuuech!. No, I don't like it. I cuddle her now very rarely. I like to tell her as a joke: 'first you get your transplant, and then I'll cuddle you (mother laughs)'. Before I used to let her sit on my lap. It was very funny, I used to put two pillows on my lap and she sat on top of the pillows (mother laughs). But now she sits on the sofa".

Overall, *hostility* was nearly nil *between the parents*. The following examples illustrate the content of such remarks: Mrs Roberts, remarked, "I can do without him (her husband), but not without my friends". The mother of Libby, aged 7, said:"I don't want him around, I prefer to be on my own".

Changes in Hostility

Following surgery, there were few changes in the parent's hostility profile regarding the ill child. This is illustrated in the following passages: Martin's mother said when talking about her 4 years old son: "he has changed. My Martin was so sweet and beautiful!, but I do not recognise him anymore. I don't like him now. His face is puffy, he is heavy... he is not my boy any more. I don't like him any more". Mrs Sharpel said about her 16-year-old son, "Following the transplant, I was so glad to see him having a normal life. I know adolescents explore with drugs but I never thought that Steven (son) could behave like that. In one level I know it is a typical adolescent behaviour. At another level I regret keeping him alive. He says that he only tried them twice, but I don't want him in my house. I've told him, he was better dead. I don't trust him anymore. I feel like he has hit me with a sharp knife". The mother of Trevor, said about her son, "He has a total personality change. I think he's let everybody down, including himself. ... he is being naughty, very very naughty. I could have actually killed him the other day!... I feel like bashing his brains in for not eating. That really aggravates me!, infuriates me!".

Parents who showed *hostility towards their partners* at the initial interview, also rated on this scale at the follow-up. Parents scoring 'no hostility' at T1, but 'hostility' at T2, were either dissatisfied with the amount of support received from their partner, or were disappointed with their partner's behaviour at the time of the child's operation. In all cases but one, hostility took the form of rejection.

The following examples illustrate the parents' expression of hostility towards their partner. The mother of Nathan, aged 11, said about her husband, "He is a pain in the bottom. At the time of the transplant I had to keep him at home. He could have kept his mouth shout!. He created so

much trouble with the nurses and the doctors!. There is a part of him that I really dislike. Probably, he felt excluded; but there is no excuse. I never go out with him; he embarrasses me. We have separate social lives". Michael's mother, said, "I hate him, I really do (cries). He wasn't there for me when I most needed him". Another mother who also felt very unsupported by her husband at the time of the transplant remarked, "I don't want him any more. There is no point in us being together. I don't want to live with him any more. I have told him that I want a divorce". The mother of Aia described her husband as if he could do no right. Aia's father gave up his work following Aia's diagnosis with the intention of being near his son while awaiting transplantation. Following the transplant, he decided not to return to work and to help his wife out in the running of her own business. Aia's mother said, "He is lazy... He is using Aia's illness, but deep down he is a lazy person and doesn't want to have any responsibility. I am the breadwinner. He goes out spending my money. This is the way he gets me. He can't reason. It is just his presence... I can't breathe... I can't stand it any longer. His presence makes me sick!.

5. Description of Positive Remarks (PR)

A positive remark is a statement that expresses without ambiguity, praise, approval, or appreciation of the behaviour or personality of the person to whom it refers (Leff and Vaughn 1985).

Typical statements were along the following lines: The mother of Zoe said, "I like everything about her. She's very good. Pleasant. Easy going. A very loving child". Some parents felt very positive about their children's achievements at school, like Mr Carlton who said about his son, " He is very bright. Despite his illness, he has always been at the top. He is doing very well, extremely well". Mrs Jense, also talking about her son, said, "He is a genius. He really is".

Most parents praised their children for their courage and determination to survive the illness. The father of Alex talking about how Alex is coping with his illness, said, "He is very ill, but he never complains. He is very good in that way". Alex's mother added, "He is very strong. He has a very strong personality. If it hadn't been for Alex and his attitude about his illness we wouldn't cope

with it so well, but he is coping marvellously". He is lovely. I wouldn't change him at all". Terence's mother said, "He is a lot braver than most adults would be. I don't know what it is about children with C.F., but they have something special. It just seems they've got an inner sense to life. He is very, very adult in some ways". Libby's father, remarked, "she is a fighter, she really is. She would not give in to her illness. She is determined to survive".

Some parents felt positive about their children taking responsibility for their medication. Kattie's parents illustrate this in the following example, when father said, "She is very responsible with the medication. We trust her to take it herself. She is very good in that respect".

The tone of voice and body-movements of some parents while describing their ill child suggested the existence of strong idealisation of the child, as in the two following examples:
Sarah's father remarked, "Oh!, she is absolutely beautiful!. She is absolutely perfect!. She mixes well with other people, makes friends very easily. She is very, very caring. At school she is brilliant, really brilliant!. She is very intelligent!. I don't know where she gets it from". The mother of Deborah, who was 10 years old and required low-risk cardiac surgery said, "She is very intelligent. I am very proud of her. Whenever I go with her, everybody loves her. She is wonderful at school. I can't say anything bad about her. She's really... I'm really proud of her. She is what I wasn't. I was a good child but not brainy; Deborah is intelligent and educated. Whatever she puts in her mind to do, she will do it... She is a dream of a child!, she really is!".

The rate of *positive remarks between the parents* was very low, indeed. Most parents who made a positive comment about the other also rated high on the warmth scale. Most remarks expressed an appreciation for the help and support offered by the partner regarding the child's illness. For example, Mrs Peters said about her husband, "He is very good. He knows what I need. Laughing helps me to cope, and he knows how to make me laugh. He is very good with me, he is excellent"; or Mrs Voghn remarked, "He is brilliant. If I am busy, he does the physiotherapy with Neil, or he cooks dinner. He is really good." Mr White said about his wife, "She has a special sort of character that calms me down. She is also a wonderful mother".

Changes in Positive Remarks

There were no changes in the content of positive remarks regarding the child. Parents usually praised their children' academic achievements and how well they were coping with the hospital procedures and treatment.

Few parents increased the number of *positive remarks about their partner* following surgery. The content of the positive statements expressed showed appreciation for the partners' supportive behaviour. These statements have been already described above, under changes in warmth between the parents.

FAMILY ADJUSTMENT

The family's emotional expressiveness (EE scales) is an important aspect of family functioning. The examples given above not only show aspects of the mother-child, father-child, and mother-father relationships but also describe other aspects of family functioning such as communication, boundaries, marital conflicts, etc.. In this section, I will discuss other aspects associated with family adjustment, which have not been mentioned in the previous section.

Many families in this study showed marked difficulties in their relationship. This is not surprising given the extreme stress faced by these families. Most parents identified the time surrounding the diagnosis of the child as a critical time. In fact, many parents believed that their family difficulties started during the period in which their child was diagnosed. Mr and Mrs Jordan are a typical example.

Mrs Jordan noticed that something was wrong with Paula. Initially, Mr Jordan didn't notice anything. On a few occasions, when his wife was willing to take Paula to the hospital, he insisted that she was too anxious about Paula's health, and reassured her about their daughter's well-being.

When the diagnosis was made, Mrs Jordan felt extremely guilty about not having taken Paula to the hospital earlier. She was also very bitter with her husband for not having listened and trusted her judgement. On the other hand, Mr Jordan felt very guilty about both not recognising his daughter's symptoms and not having supported his wife. Believing that he failed as a father, and feeling rejected by Mrs Jordan, he put all his efforts into being the breadwinner; whilst his wife became Paula's only carer. Paula's dad was unhappy with his function in the family. However, due to his low self-esteem following his daughter's diagnosis, he didn't have the courage to discuss openly his role in the family. In this way, avoiding conflict and confrontation, the parents' roles were re-defined.

Although six years had passed since the diagnosis, 'that period' was still an unresolved issue for this couple. During all this time, Mr and Mrs Jordan had kept physically and emotionally distant from each other. When they were confronted with their daughter's need for transplantation, the feelings of the past became very vivid in the present. Mr and Mrs Jordan were very resentful and critical of each other. Subsequently, they were unable to discuss with each other the implications of their daughter having/not having a transplant. They were also unable to discuss the illness and prognosis with Paula, who wanted to discuss the likelihood of her own death.

Paula was this couple's first child. It is of interest that, in this study, a significant number of children with cystic fibrosis were living with single mothers. In 75% of the cases, the contact between the ill child and the biological father was non-existent. Nearly all mothers said that the biological fathers left the family at the time of diagnosis or soon afterwards. The most common reason given was that the father felt unable to bring up a child suffering from a life-threatening condition.

It should not be concluded that having a child with a chronic condition necessarily has a detrimental effect on the marital relationship. Only 30% of the parents participating in this study suggested that the illness had a negative effect on the quality of their marriage. Furthermore, some of these parents accepted they had some marital difficulties even before the child was conceived. An average of 50% of the parents reported that either the child's illness did not have any effect on

their relationship, or that they experienced a strengthening of their marriage following the child's diagnosis. The remaining 20% felt unable to comment on this matter.

The data collected during the interviews with the parents suggest that mothers have different ways of coping with the crises than fathers. Mothers seemed to relieve anxiety by talking and sharing their feelings with others, preferably their husbands. However, if their husband was absent or unavailable, the nurses or the mothers of other children who were also admitted on the ward also had a buffering effect on mother's distress.

The fathers were more introverted, and found it more difficult to talk about their concerns and feelings. Few fathers found it easier to talk to someone else other than their spouse. These fathers perceived their partners as weak and in need of constant support; they were worried about jeopardising their partners' coping abilities if they were to admit to them their own distress.

Whatever differences the coping mechanisms were between the husband and wife, parents in well adjusted families accepted their partner's ways of coping. Often these parents were able to create an environment in which their partner's needs were met. Some parents would have liked their partner to behave differently. Nevertheless, they did not feel resentful but sympathetic and understanding towards their partner.

On the contrary, parents in poorly adjusted families were more focused on their own feelings and needs, without being aware of their partners' necessities. Often they were resentful and critical of their partner's behaviour.

Although in some families parents shared 'equally' the practical and emotional demands of treatment, overall, mothers were mainly responsible for the burden of the child's treatment. However, clinical impressions seem to indicate that mother's subjective perception of father's emotional availability was more strongly associated with family adjustment than the father's lack of involvement with the child's care and treatment. Those families in which the parents had not organised their life and roles in a manner that was acceptable to both of them were also more

prone to have difficulties in their relationship than families in which parents had reached a satisfactory agreement about their roles and tasks. This suggests that it is not the behaviour in itself what it is important, but the context and the recognition of what a specific behaviour means.

Changes in Family Adjustment

There were few changes in family adjustment following the improvement in the child's physical condition. The parents of those children, whose family adjustment improved, attributed positive characteristics to the experience of surgery (such as personal growth or the discovery of new qualities in their partner).

On the contrary, parents whose family adjustment deteriorated were still resentful and angry about what their partner did or did not do at the time of surgery. All of them described their partner as unsupportive and suggested that the surgery had accentuated the negative aspects of family relationships. In summary, most families who were poorly adjusted prior to the child's admission for surgery remained poorly adjusted following successful treatment.

CHAPTER 6

DISCUSSION OF THE RESULTS, IMPLICATIONS OF THE STUDY
AND CONCLUDING STATEMENTS

6.1 SUMMARY OF THE RESULTS

Using a framework derived from family systems theory, the main aims of the present research were: i) to explore the way in which the family organises itself at the time its members are informed about the child's need for transplantation, and ii) to explore whether the way in which the family is organised has a long-term effect on the patient's medical outcome. In this process, differences in both children's and parents' adjustment, and family organisation, between transplants candidates and children admitted to undergo conventional cardiac surgery were explored.

A total of 165 children and families (transplant programme: n=106; conventional cardiac surgery: n=59) were initially assessed, of whom 118 were available at the follow-up (transplant programme: n=62; conventional cardiac surgery: n= 56). This study showed that, at entry into the study, over 60% of the children and over 40% of the parents had significant adjustment difficulties, and that half of the families were moderate or poorly adjusted. This study failed to show an association between psychosocial typology of illness, individual and family adjustment. It also failed to show that the families of children with the most severely chronic disease activity –candidates for transplantation– were more disrupted that families of children with less disease activity –children undergoing conventional cardiac surgery. Nor were there differences in family organisation between these two groups.

The study did show that, pre-operatively, parental adjustment, marital relationship and family organisation were associated with the child's adjustment. As the children's physical health improved post-operatively there was a reduction in the number of children with a psychiatric disturbance and the children's psychosocial functioning was also enhanced. Post-operatively, the parents' psychological adjustment also improved but there were

some changes in the family organisation. This was shown by a deterioration in the marital relationship and a reduction in warmth between partners, less parent-child overinvolvement, and higher rates of parent-child critical comments. Post-operatively, changes over time in family adjustment were expected. However this study showed that overall the levels of family adjustment were fairy similar pre-and post-operatively.

Children who survived on the waiting list for transplantation twenty-two months or longer, had better physical health, better school attendance and better psychosocial adjustment than children who did not survive this length of time. Parents and family's characteristics did not discriminate between survivors and non-survivors. However, this study supported the view that the children's characteristics, family and disease mutually affect one another (Woods et al 1989). Good child's psychosocial functioning and positive family organisation, shown by expression of warmth and positive remarks, were found to have a beneficial effect on the children's physical health. The presence of a psychiatric disturbance in the child, poor family and marital adjustment, mothers' poor adjustment, and negative family organisation (shown by high overinvolvement with the patient, and the presence of criticism and hostility between family members) were found to have a detrimental effect on the children's physical health.

Unsatisfactory adherence was found in a third of the transplant recipients, and heart-lung transplant recipients were more at risk of poor adherence than the heart recipients were. Living in a single or reconstituted family, the absence of warmth, expressions of hostility and criticism, and family maladjustment were associated with unsatisfactory adherence. The clinical significance of these associations is unclear.

6.2 DISCUSSION OF THE RESULTS

6.2.1 DEMOGRAPHIC CHARACTERISTICS OF THE PARTICIPANTS

There are inevitable limitations in studies of this nature making it impossible to obtain a comprehensive and representational sample. Some of the possible biases introduced by the nature of the sample included in this study will be briefly discussed first. Other factors affecting

the sample studied are discussed later on in this chapter, under "critical appraisal of the study" –see pages 221-225-.

Many non-Caucasian parents who were asked to participate in the study had difficulties with spoken and / or written English. This resulted in the exclusion of 49 families, 12.3% of the 398 families originally invited. Furthermore, prior to the request for participation, the physicians had already removed those families known to require an interpreter to communicate. This explains why there was a small proportion of non-Caucasian families participating in the study.

Three families (2.75%) whose children were assessed for transplantation and 213 families (27.6%) whose children were admitted to undergo conventional cardiac surgery refused participation. These parents were concerned about their children getting upset during the interviews, just before their operations. Many of these parents were clearly very distressed about their circumstances, needing space to keep themselves together. This may mean that families who participated and families who refused participation used different ways of dealing with medical stressful events. Interestingly, a few of the parents whose children were assessed for transplantation revealed at the follow-up their fears about their children not being accepted into the transplant programme, if they had refused participation, even though they had been given re-assurance about this not happening.

Although the vast majority of the children were being brought up in two-parents families (natural or reconstituted families), there were significant differences in family composition between the candidates for transplantation and the group of children admitted for conventional cardiac surgery. The lower number of children living with both natural parents in the former group showed this. In interviews, at least half of the single or re-married mothers whose children had cystic fibrosis attributed the break-up with their children's natural father to these fathers' non-acceptance of their ill children. It was also revealed that most of these natural fathers were not keeping regular contact with their children. It is of interest that the prevalence of single parents, particularly in the group of families whose children were awaiting heart-lung transplantation (15.4%, PvHL group), was higher than the estimated figures (4 %; the figure including ethnic minority groups is of 20.9%) from the Office of Population and Census Surveys for households headed by a person from the white population (Fundudis 1997).

The distribution of social class was also intriguing. Overall, the number of families from social class III was much lower than the estimated figures from the National Statistics (52%) whilst the number of families from social class IV and V seem much greater (Social Class IV: 14%, Social Class V: 4%; OPCS General Household Survey, 1988)

From an anecdotal point of view, low socio-economic status, lack of a supportive network, and difficulties in family relationships are seen as psychosocial contraindications for transplantation at other transplant centres in the U.K.. This results in a greater number of children from the disadvantaged families referred to Great Ormond Street for transplantation treatment.   This probably explains the social class bias in the transplant group sample. Nevertheless, the reasons for the bias in the group of children undergoing conventional cardiac surgery is not clear.   Once a month, the senior cardiologists from Great Ormond Street Hospital have a clinic at other hospitals or GP surgeries within the U.K.. These community clinics are in areas where local hospitals do not have the resources to assess and/or follow-up children with congenital heart abnormalities. When heart surgery is required, these children are operated at Great Ormond Street Hospital. It could be that the children who are referred to those clinics are a selected population.

6.2. 2 PROFILE OF THE CHILDREN AND FAMILIES PARTICIPATING IN THE STUDY

Children's Physical Health Profile

The results showed that the children assessed for transplantation were more seriously ill and more disabled than the children admitted to undergo conventional cardiac surgery.

Children's and Parents' Adjustment Profile

The psychosocial typology of illness was expected to be associated with the children's and parents' psychological adjustment.   It was hypothesised that children referred for transplantation and their parents would demonstrate higher levels of adjustment problems than children admitted for high or low-risk conventional cardiac surgery and their parents.

Overall, 24% of the children had a psychiatric diagnosis and over 60% of the children showed mild to severe psychosocial impairment in the Global Assessment of Functioning (GAF Scale) measure. Contrary to expectations, there were no differences in the number of children with a psychiatric diagnosis between the group of children assessed for transplantation and the group

of children admitted for conventional cardiac surgery. Furthermore, no differences between the groups were found in this parameter when the other specified group combinations were examined. Neither did the children referred for transplantation demonstrate higher levels of adjustment problems in the Global Assessment of Functioning measure (GAF scale) than children admitted for conventional cardiac surgery. Nevertheless, when the mean scores were compared between the specified groups, it was found that the children on the active list for heart-lung transplantation had poorer functioning than the children on the provisional list. If the children's level of functioning was related to the psychosocial typology of illness, differences between the other specified group comparisons should also be expected. That was not the case, and there were no differences between the groups when the numbers of children with mild to severe impairment were examined. Therefore, the hypothesis studied was not confirmed.

Comparison of the incidence of psychiatric disorders with other studies is difficult due to the lack of studies exploring this within the context of life threatening conditions in children. While some studies on adults have found that 52-60% of the candidates for transplantation had a psychiatric diagnosis, and 32% of the candidates showed adherence problems with treatment, (Chacko et al 1996; Mai et al 1990), Rodin and Voshart (1987) found that only 25% of the candidates for renal transplantation were mentally ill.

The number of children with impaired psychosocial functioning in the present study seems higher than that reported in other studies with children undergoing heart and heart-lung (45%) or bone- marrow (20%) transplantation (Wray et al 1994; Pot-Mees 1989). The reasons for this discrepancy are complex. As mentioned earlier in this chapter, the psychosocial contraindications for transplant treatment implemented at other transplant centers contributes towards producing a sample with different characteristics at Great Ormond Street Hospital. Possibly this has an effect on the prevalence rates. In addition, questionnaires were used in the studies mentioned above, whilst in the present study a combination of standardised clinical interviews and self-reports measures completed by different informants were utilised. Despite this, some of the findings in the present study are in consonance with those of Wray et al's (1992; 1994) who found no differences in adjustment levels between a group of candidates for heart transplantation, children undergoing conventional cardiac surgery and healthy controls. Some studies on children with congenital heart diseases, cystic fibrosis, bowel malformations,

children surviving malignant neoplasms, and children receiving glucocorticoid treatment have obtained similar results (Linde et at 1966; Linde 1982; Drotar et al 1981; Bywater 1981; Diseth et al 1995; Satel 1990; Copeland and Davidson 1989); whilst others have found greater incidence of behavioural problems in this population (Spurkland et al 1993; Cowen et al 1986).

As mentioned previously, mean score comparisons showed that the children in the active list for heart-lung transplantation had poorer levels of psychosocial functioning than the children on the provisional list. Although, the two groups were homogeneous with regard to the children's diagnosis of cystic fibrosis, the two groups differed in physiological functioning, required number of hospitalisations and stresses associated with provision of treatment. This has an impact on the amount of social contact with peers and demands imposed on other family members. Associations between the children's adjustment, physiological variables and number of hospitalisations had been previously reported (Steinhausen and Schindler 1981; Cowen et al 1986). If there was a strong link between illness variables and the children's adjustment, differences in adjustment levels between the children assessed for transplantation and the children undergoing conventional cardiac surgery would also be expected. However, this study failed to show such an association.

The process of grieving and mourning might be an alternative explanation. As mentioned above, nearly all children in the active or provisional lists for heart-lung transplantation had cystic fibrosis. The mortality rate of patients on the active list is around 60%, whilst for those on the provisional list it is under 15%. While critically ill, the children on the active list could not cope with the illness by denying the ultimate outcome of their disease. Information from the psychiatric interviews suggests that, at this stage the children were aware and coming to terms with their possible imminent death; and that bereavement was part of these children's lives as nearly all of them had recently experienced the death of a close friend from cystic fibrosis, usually another Great Ormond Street patient. Sadness, grief and identification with the dead child were additional aspects of their lives. Although children on the provisional list were also aware of the fatal outcome of their illness, and some had previously lost a sibling, these children are more protected from the experience of dying.

The admission of any child to hospital is a crisis for both the child and his/her family (Bentovim 1980). As expected, parents participating in the present study showed great levels of distress,

particularly mothers. With the exception of the mothers, whose children were on the provisional waiting list for transplantation, mothers' mean scores in all groups were above the threshold score in the GHQ-28, these indicating a possible psychiatric disturbance. Similar results were obtained for fathers of children assessed for heart transplantation and for fathers of children undergoing high-risk cardiac surgery.

Using the same methods, the parents of the children studied showed more adjustment difficulties in comparison to a random community sample (Goldberg 1978). Previous research on parents whose children are assessed for transplantation has found an incidence of psychiatric disorders of 60-69% for mothers, and of 47-58% for fathers (Pot-Mees 1989; Dermatis and Lesko 1990). The present study replicates the higher incidence of psychiatric problems in mothers when compared to the fathers. This is also in consonance with a large body of literature on chronic conditions in childhood (Goldberg et al 1990; Bywater 1981; Pot-Mees 1989). Information gathered from the parental interview gives some light into this gender difference. Mothers are more affected by the children's illness because of their role as main carers and, consequently, their closer relationship. In addition, women's behaviour is more emotion-orientated or expressive, whilst men's behaviour is more task-orientated or instrumental (Notarius and Pellegrini 1987). The combination of the father's instrumental behaviour and his lesser involvement with the child on a day-to-day basis, may have a buffering effect on his level of adjustment.

Parents, whose children were on the active list for heart or heart-lung transplantation were aware that time 'was running out' for their children, had the burden of extra care and frequent hospitalisations, and the complexities of the medical treatment (Goldberg et al 1990; Cowen et al 1986). Furthermore, many of these parents had previously lost another child, and were struggling with emotions associated with past experiences of illness and death. Therefore, parents of children assessed for transplantation were expected to show greater adjustment difficulties than the parents of children undergoing conventional cardiac surgery. This hypothesis was not supported by the results obtained, as the GHQ-28 comparisons did not reveal discrepancies between the transplantation and conventional cardiac surgery groups, nor were there differences found between the other specified group comparisons. This study also failed to show a higher incidence of psychiatric cases in mothers of candidates for transplantation when compared to mothers of children undergoing other forms of cardiac

surgery. Furthermore, it showed that fathers of children undergoing cardiac surgery had a higher incidence of psychiatric disorders than the fathers of children assessed for transplantation. This was due to the high number of fathers in the high-risk cardiac surgery group scoring above the threshold score. These results are contrary to all predictions. The role played by illness factors upon the parents is unclear. While some researchers have found associations between the level of parents' adjustment and the amount of time invested in daily care, frequent hospitalisations, severity of illness, and the complexities of the medical treatment (Goldberg et al 1990; Cowen et al 1986; McConville et al 1990), other have found no associations between parents' psychological symptoms and medical aspects (Kovacs et al 1990).

It is striking that a greater number of mothers in the low-risk cardiac surgery group (acyanotic children with no disabilities) had a psychiatric disturbance when compared to the mothers whose children were undergoing conventional high-risk cardiac surgery (cyanotic, disabled children). Differences in ratings of parental anxiety between mothers of acyanotic and mothers of cyanotic children have been reported (Linde et al 1966). It could be that the presence of the heart condition, and the meaning attributed to it, rather than its severity are the main determinant in mothers' adjustment. Information from the parental interviews suggests that there is no such thing as high and low-risk surgery for the parents. Cardiac surgery is always perceived as high-risk. Furthermore, due to the absence of physical symptoms or disability, parents of children undergoing low-risk cardiac surgery had greater difficulties accepting the need for surgery, and often they expressed a strong wish against surgery treatment. Surgery represented an act of taking a healthy child and making him sick. They experienced guilt as if they were inflicting unnecessary pain and suffering upon their children. On the contrary, parents of children undergoing high-risk cardiac surgery expected a decrease in cyanotic symptoms and improvements in the children's disabilities after surgery. Therefore, it is possible that due to the different experience of illness, parents of children undergoing high or low-risk cardiac surgery differ in their cognitive appraisal, meaning and beliefs about surgery, and consequently they have developed different coping strategies. This needs further exploration.

Results from these various measures indicate that the children studied were rated as having more psychiatric problems and poorer adjustment than that reported in normative samples. However, the level of difficulty was unrelated to the children's psychosocial typology of

illness. Parents of the children studied also had more psychiatric disturbance and adjustment difficulties than that reported in normative data, however the levels of maladjustment were similar to those reported in previous studies on parents of children with life-threatening conditions. The results also indicate that factors related to the children's disease or to the level of disability (psychosocial typology of illness) do not impact on parental adjustment.

Marital Adjustment and Family Organisation Profile

It was hypothesised that families of children who are referred for transplantation assessment would demonstrate a different pattern of family organisation than families of children undergoing conventional cardiac surgery. This was expected to be by higher levels of marital and family conflict and a higher rate of negative attitudes between family members.

The results showed that the mean marital adjustment scores of the parents studied were within an above average range, and that the level of marital adjustment was not mediated by the psychosocial typology of illness. This finding was confirmed when group differences were explored with regard to the number of parents rating above the threshold score indicating poor marital relationships. Although marital strain is greater in families containing a child with a chronic condition (Eiser 1990; Patterson and McCubbin 1983), this study failed to show an association between the seriousness of the illness and marital maladjustment. The rate of parents with marital conflicts in the present study is consistent with Sensky et al's findings that 30% of the parents of adolescents with diabetes had antagonistic-apathetic marriages (Sensky et al 1991). This study also showed that more than half of the families studied had adjustment difficulties. Other studies on chronic illnesses have reported similar results (Diseth et al 1995; Spurkland et al 1993).

The present study also failed to show that families of children with the most severe chronic disease activity, and with more daily demands of medical treatment (transplantation group), were more disrupted than families of children with lesser disease activity and smaller demands upon family members (conventional cardiac surgery group). Having said that, differences in the level of family adjustment were found between the transplantation groups. Children on the active list for heart transplantation had similar disease activity compared to children on the active list for heart-lung transplantation; but a greater number of families in the former group were well adjusted when compared to families in the latter group (although this difference did

not reach statistical significance). The families of children on the active heart-lung transplantation list showed significantly more disturbances than the families of children on the provisional list. The children on the provisional list for heart-lung transplantation had slightly less disease activity than the former group. The families of children on the active list were different to the families of children in the provisional list in the following parameters: they had greater family disruptions and more accumulative stresses due to the amount of time needed to provide medical treatment; the children had a greater number of medical complications requiring frequent admissions to hospital; and a larger number of families had previously lost other children. During the parental and children interviews, family members disclosed unresolved grief and guilt about the dead children. Many parents were resentful towards their partners for the way they coped with the loss. Clinical impressions suggested, that the way in which family members had organised themselves in the past was conflicting with the way in which family members were communicating during this new situation, the children's assessment for transplantation. This seemed to have a direct effect on the family members' ability to agree about whether transplantation was a treatment they could cope with. In fact, all families, who prior to the assessment for transplantation had lost one or more children, were referred for family counselling to deal with the unresolved grief and to assist them with their ambivalence about transplantation. Although further exploration is required, it is possible that the traumatic experience of having lost other children in the family, increases the family members' vulnerabilities to deal with traumatic events in the future. This may result in inadequate coping in new traumatic situations, and may give rise to patterns of distorted family communication.

Families of children assessed for transplantation were expected to show a different pattern of attitudes towards the children than families whose children were undergoing conventional cardiac surgery. This study showed that disease activity did not influence the parents' attitudes towards the ill children. Only recently, researchers have started to explore the attitudes of relatives towards the physically ill children. The attitude profile presented by parents of children with cardio-respiratory disorders has some similarities with the attitudes profile described by Senky and colleagues (1991) in their study with children with diabetes. This included low levels of hostility, positive remarks, critical comments and overinvolvement, and moderate to high warmth. However, there were some important differences. In the present study, over 73% of the mothers' and 84% of the fathers' made no critical comments, while

Sensky et al (1991) found that only 24% of their interviews contained no critical comments. Moreover, Sensky et al (1991) found a higher percentage of hostility (10%) than in the present study (mothers: 5%; fathers: .9%).

Previous research has also found that parents of children with cystic fibrosis expressed higher levels of overinvolvement than a sample of parents with healthy children (Blair et al 1995), concluding that overinvolvement represents a natural parental reaction to the gravity of the illness. The present study failed to replicate such findings. In addition, the profile displayed by parents of children with cardio-respiratory disorders is dissimilar to the profile described by Invernizzi et al (1991) in families of heart operated adult patients. These researchers found high levels of both warmth and overinvolvement , and there was a positive association between these two parameters. In the present study, levels of parental overinvolvement with the children were much lower and there was no association with levels of warmth. Invernizzi and colleagues' sample was collected in Italy and age as well as cultural differences may account for the discrepancy in the results. Another difference between the present study and previous research is that other studies have found no differences between mothers' and fathers' EE ratings (Cook et al 1989; Sensky et al 1991; Le Grange 1989). Yet, in the present study mothers showed more overinvolvement with the children and rated higher on positive remarks than did fathers.

There are no studies exploring levels of expressed emotion towards the partner in parents of physically ill children. Again, the specific predictions made were not confirmed, as parents' attitudes towards their partners were quite similar across the groups. Because of the way in which parents were invited to talk about their partners during the interviews, the low mean scores in positive remarks and warmth are striking. Despite the reported positive associations between stress, poor family adjustment and critical comments (Hammen et al 1987; Hooley and Teasdale 1989), the parents' ratings in critical comments were also low. It seemed as if these parents were holding back both positive and negative emotions towards their partner. It could be that the pattern of attitudes shown represented the way in which these parents usually behaved towards each other. On the other hand, feeling vulnerable and overwhelmed by the stresses associated with the transplant assessment or with conventional cardiac surgery, parents may have become detached from their emotions in order to protect themselves and to cope with their stressful situation, hence distancing themselves from their partner.

The present study showed relatively small differences between mothers' and fathers' attitudes to their partners. During the interviews, mothers of transplant candidates expressed more dissatisfaction with their partners' emotional availability and involvement in the children's treatment than the mothers of children undergoing conventional cardiac surgery. However, these mothers' comments were seldom sufficiently critical in tone to rate them formally as critical comments. There are several reasons to explain the differences between mothers and fathers as well as between the two groups:

a) The burden of the illness on the mother was greater in families of transplant candidates than in families of children in need of conventional surgery. Therefore, mothers in the former group may be more isolated and in need of more support than the mothers in the other group. Other studies have found increased dissatisfaction in Alzheimer disease relatives with little contact with friends, and lacking social support (Gilhooly and Whittick 1989). Unfortunately, a measure of global perceived support was not used in this study.

b) Clinical data from the parents' interviews showed that parents use dissimilar coping mechanisms, and have different needs, styles and desires. Nearly all mothers responded with a need for intimacy and closeness in response to their children's surgery. They wanted their partners near them, listening to their feelings and sharing their fears about losing the child. In contrast, fathers often reacted by detaching themselves from the experience of surgery and had difficulty expressing their feelings. Furthermore, many fathers said that they did not know what their wives were expecting them to do, and felt puzzled about their partners' needs and demands to be comforted.

In consonance with Sensky et al's (1991) findings, this study has also shown associations between the parents' attitudes (EE) towards their children and family adjustment. Furthermore, this study incorporated a measure of parents' attitudes towards their partners, showing that: a) high warmth and positive feelings between the parents were associated with high levels of warmth and positive feelings in the parent-child dyad; and b) parents in well adjusted families showed more positive attitudes towards both their children and partners than parents in poorly adjusted families.

In summary, this is the first report on the levels of adjustment in children referred for heart and heart-lung transplantation and in their families. The findings indicate that: 1) The levels of

children and parents' maladjustment prior to transplantation are high. Similar levels of maladjustment were found in the families of children undergoing conventional cardiac surgery.

2) There is a need to monitor children and families levels of adjustment while waiting transplantation and to identify risk factors associated with it; and 3) There is a need to provide ongoing psychological support for the children and their families throughout this very difficult period of their life.

As Bradford and Tomlison have stated 'there are critical transition points in the cycles of families that can delay successful adaptation, and consultation needs to focus not only on medical aspects but on how the families are coping. It is only by integrating psychological and medical considerations that optimal care can be provided' Bradford and Tomlison (1990, page 1003).

6.2.3 ASSOCIATIONS BETWEEN CHILDREN'S ADJUSTMENT, PARENTAL AND FAMILY MEASURES

It was hypothesised that the children's adjustment would be associated with parental and marital adjustment, and with aspects of family organisation parameters.

This study found no significant associations between demographic characteristics, parents' psychological or marital adjustment and the children's mental status. However, there were associations between the children's' mental status and the following family characteristics: warmth, critical comments hostility and family adjustment. Families whose children had a psychiatric disorder had more adjustment difficulties than the families whose children did not meet the ICD-9 diagnostic criteria. When compared to the parents of children with no psychiatric diagnoses, parents of psychiatrically disturbed children were also more critical of the children, the mothers scored less in warmth, and both parents were more hostile towards each other. These associations are in consonance with previous reports on children with psychiatric conditions (Vostanis et al 1994, 1995; Hibbs et al 1992; Stubbe et al 1993; Asarnow et al 1993). The methods used in these other studies are similar to the methods used in the present study in that expressed emotion scales are the main measure.

This study also showed that gender was the only demographic characteristics associated with the children's level of psychosocial functioning and that, as expected, the children's psychosocial functioning was associated several parents' and family's characteristics. The mothers of children with impaired functioning had more difficulties functioning on a day to day basis than the mothers of children with no impairment. The parents of children with impaired functioning also had more marital difficulties than the parents of children with no impairment. Some attitudes between family members were also related to the children's functioning. Low levels of warmth, the presence of hostility and overinvolvement, and poor family adjustment were associated with impairment in the children's functioning.

Regarding gender differences, the psychosocial functioning of the boys participating in this study was poorer than the psychosocial functioning of the girls. La Greca has noted that 'conditions that limit physical activity might produce greater social consequence for boys, given the athletic and activity-orientated nature of boys' peer contact' (in Eiser et al 1992, page 263). Clinical information from the children's interview seems to agree with this statement.

As McFarlane et al (1995) have recently demonstrated, family adjustment is strongly associated with parental style. In fact, partner-partner and parent-child style of attitudes are different aspects of family functioning, (Stubbe et al 1993). Hence it is not surprising to find so many interactional variables associated with the children's psychiatric and psychological functioning. These results suggest that despite the anxiety provoking circumstances in which the parents were interviewed, the parents of well adjusted children were less distressed and experienced a greater sense of emotional closeness, empathy and enjoyment of their children's company than the parents of children with impaired functioning. These parents were also more aware of their partners' needs and were able to offer them their emotional support, this is shown by the higher ratings on the warmth scale and lower ratings on the hostility scale. The results also showed a tendency in these parents to express more positive remarks about their partners. Not surprisingly these parents reported greater satisfaction with their marriage and had better functioning families than the parents of children with impaired psychosocial functioning. This study showed that poorly adjusted children came from families where there was inadequate or inconsistent parenting and the children commonly got caught up in marital problems. There was an overall inability to respond appropriately to emotions and, often, the parents expressed their resentment about the illness by withholding expression of warmth towards the children.

These results are consistent with previous work in samples of young transplant recipients, children with congenital heart diseases, children with cystic fibrosis, bowel malformations, cancer, diabetes and other chronic conditions in childhood (Bernstein 1977; Pot-Mees 1989; Uzark et al 1992; Phipps and Mulhern 1995; Linde et al 1966; 1970; Garson et al 1978; Kong et al 1986; Cappelli et al 1989; Perrin et al 1989; DeMaso 1991; Wells and Schwebel 1987; Spurkland et al 1993, Steinhausen and Schindler 1981; Lewis and Khaw 1982; Lask 1994; Diseth et al 1995; Wallander 1989; Overholser and Fritz 1990; Hanson et al 1992; Wolman et al 1994). These authors found that the presence of chronic family difficulties were predictors of psychosocial outcome in physically ill children.

Associations between parental psychiatric illness and poor children's adjustment have been reported in the literature. The mechanism by which parental characteristics and children's adjustment influences each other is not clear. Linde et al suggested that distressed parents

transmit their anxieties to their children (Linde et al 1966, 1970). This phenomenon has been called by Wolfer and Visintainer 'the emotional contagion hypothesis' (quoted in Weichler 1993, page 135). In an interesting study, Kovacs et al (1990) found that the levels of mothers' depression and overall emotional distress were not related to the levels of depression or anxiety reported by children, but the degree to which mothers perceived the illness difficult to handle was associated with the children's adjustment. The role-played by parental coping has also been emphasised in the literature. In a study on the impact of childhood cancer on the family, Overholser and Fritz (1990) found that parental coping was associated with the long-term adjustment of both parents and children, and that the amount of emotional distress that parents experienced was related to their ability to establish and maintain supportive ties with other people. On the other hand, a negative parents-children interactional style such as pampering, overprotection, and unnecessarily restricting the range of experiences to which the children are exposed, diminishes the children's opportunities for learning. This interferes with the children's normal development and jeopardises their psychosocial adjustment (Linde et al 1966, 1970; Davis 1993).

The work of Hammen et al (1987) is relevant. Coding observation of mother-children interactions, these authors demonstrated that distressed mothers used negative patterns of interaction with their children. A higher rate of psychopathology was found in children of both depressed and highly distressed mothers (whether distressed by medical illness, psychiatric condition or environmental conditions not associated with mother's psychiatric disturbance) than in children of healthy controls. Children's psychiatric diagnoses and impaired functioning were especially strongly associated with the extent of the mothers' negative interaction with the children. Highly distressed women interacted with their children in relatively critical ways, with less use of positive reinforcement.

It has been suggested that children's disturbance and impaired functioning is not only related to the presence of negative emotional attitudes within the family but to the absence of parental warmth (Vostanis et al 1995). Furthermore, the effect of parental warmth upon children's development has been highlighted. In a recent review of the literature, Cummings and Davies noted that "parental warmth and responsiveness facilitate the development of secure attachments and internal working models characterised by positive self-concept and secure confidence in the availability and responsiveness of the parents and the larger world. On the

other hand, negative, insensitive styles of parenting, foster insecure parent-child attachments and internal working models in which children view their parents as unreliable, and themselves as unworthy and undeserving of love and affection" (Cummings and Davies 1994; page 83), which has an effect on the children's self-concept and psychosocial functioning.

Some of these studies are misleading in the sense that they do not take into account the effect that the children's behaviour may have on their parents or on other family members. The children's psychiatric symptoms and maladjusted behaviours may trigger negative parent-children interactions, these affecting indirectly parental and marital levels of adjustment.

The family's previous experience of illness, and self-efficacy with regard to treatment management (Kovacs et al. 1990), are important factors to consider when dealing with the physically ill. Clinical impressions gathered during the parental interviews give us some insight into how family and children factors have mutually reinforcing influences. The amount of parents' previous difficulties and dissatisfaction with medical professionals, and the way in which parents had organised themselves to deal with the extra demands imposed by the illness in the past, seemed to have an effect on how the parents were accepting this new medical event: transplantation or conventional cardiac surgery. Parents with greater difficulties in the past seemed more distressed at the time of initial assessment. Mothers with poor psychological adjustment (more distress) may have less individual resources, and therefore feel less able to cope with the medical and emotional needs of their children. This may lead to feelings of guilt and overinvolvement with the children. In order to find a balance, it could be that these mothers became more demanding of their partners' time and availability, seeking reassurance and support.

Concurrently, their partners may have difficulties achieving those requirements due to a combination of factors: previous unresolved issues which may be re-enacted before or during their children's admission, parents' role and gender differences. These perhaps result in withholding of warmth, disagreements about the management of the children, and increase marital and family difficulties. See FIGURE 29.

FIGURE 29: Factors affecting Psychological Outcome during a Medical Crisis
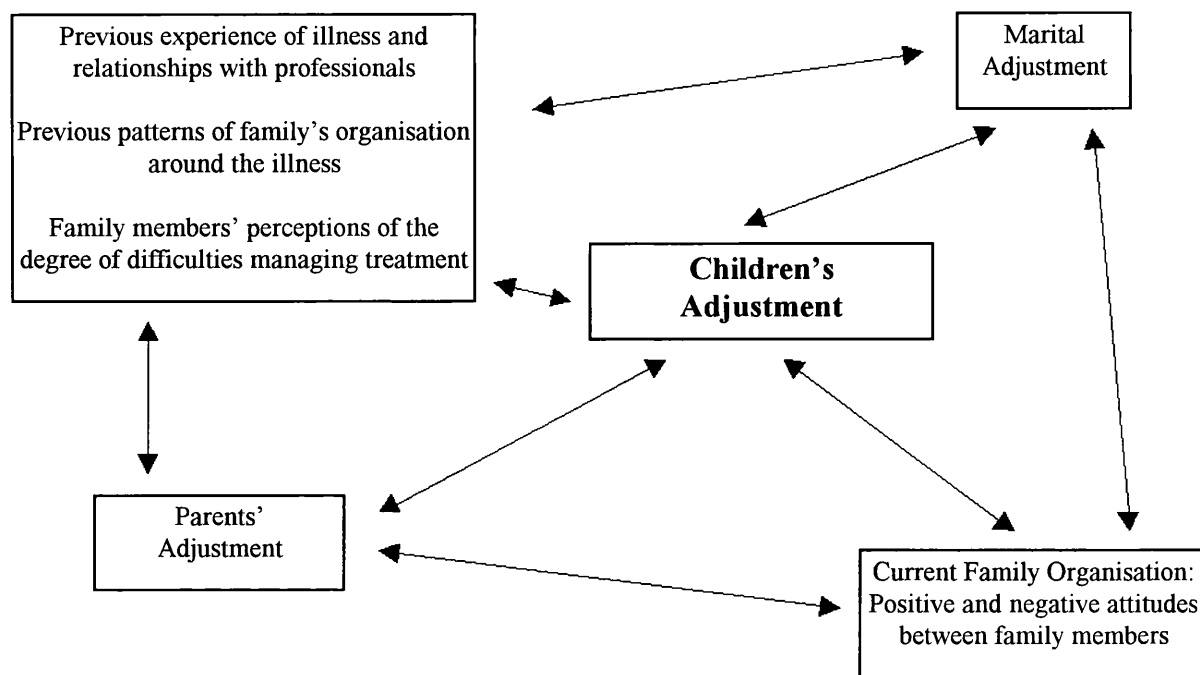
In summary, parents' level of distress, the existence of negative patterns of family organisation, and the extra strains on family relationships associated with the medical procedures and, independently, previous family patterns related to the illness and its course, are associated with the children's psychological adjustment. These results suggest that optimal treatment of children with cardio-respiratory conditions require somatic care as well as individual psychological and family care. Empirical reports suggest that children's adjustment can be improved by modifying the family organisational patterns (Kinston and Bentovim 1978). Therefore, family counselling should be available to children with poor adjustment and their families. This may be particularly important when children are confined at home due to physical disability, as the amount of contact with their peer group is often insignificant. Such children become very dependent on their parents and on other family members to retain a sense of self-efficacy and a positive self-concept.

6.2.4 CHANGES IN CHILDREN'S ADJUSTMENT, PARENTS' PSYCHOLOGICAL AND MARITAL ADJUSTMENT AND IN FAMILY ORGANISATION FOLLOWING SURGERY

It was hypothesised that as the children's health improves following surgery, changes would occur at the individual and family levels. At 12 months post-surgery, children and parents in all groups were expected to show better adjustment and better family organisation. It was also hypothesised that as the demands of daily treatment on the parents diminishes following transplantation, a greater improvement in the children's and parents' adjustment measures, and in family organisation, would be found in those families whose children received transplantation when compared to families whose children received conventional low or high cardiac surgery.

Changes in Physical Status

The effects of heart and heart-lung transplantation on the children have not been previously reported. This study shows that heart and heart-lung transplantation significantly improve the children's physical status, this leading to improvements in both school attendance and psychosocial functioning. As expected, transplant recipients had greater improvement in their physical health and in school attendance than the children who received conventional cardiac surgery.

With regard to the group awaiting transplantation there was a tendency towards deterioration in the children's physical and psychological health. Since at follow-up these children were more ill than the transplant recipients, we would have expected a higher rate of psychosocial impairment in this group. It may be that the condition of these children had not changed dramatically, while at one year post-transplantation children are facing new stressful events such as changes in their physical appearance due to the immunosuppressive medication, difficulties being accepted by their peer group, and anxiety related to possible medical complications.

Changes in Children's Adjustment

Although there was an improvement in the children's psychiatric status, this study failed to show a greater reduction in the number of children with psychiatric diagnosis in the group of transplant recipients when compared to the group of children who underwent

conventional cardiac surgery. The children experienced significant improvement in their psychosocial functioning but this was not greater (as expected) in the group of transplant recipients when compared to the conventional cardiac surgery group.

There is an absence of reports examining changes in the incidence of psychiatric illness following paediatric transplantation. In the present study, the number of children who were psychiatrically disturbed one year after transplantation was greater than in the normal population (Rutter 1989), but the incidence of psychiatric disorder post-transplantation was lower than that reported in studies on adult heart recipients (20.8% - 54%; Mai et al 1990; Maricle et al 1989). The drop in the number of patients with a psychiatric diagnosis was also less dramatic than that reported in studies with adult patients (pre-transplant: 51.8%; 12 mths post-transplant: 20.8%; Mai et al 1990).

In the present study, the frequency of major psychiatric problems following conventional cardiac surgery was similar to that found in the general population (6-10%) (Rutter 1989), and to that found by Spurkland et al (1993) in children with congenital heart disease.

While reductions in both social competence and self-concept following bone marrow transplantation have been reported (Phipps and Mulhern 1995), recipients participating in the present study showed an improvement in their psychosocial functioning. Nevertheless, despite this improvement, and in agreement with Poznanski et al's (1978) findings in children undergoing renal transplantation, half of the children showed mild to moderate psychosocial impairment at 12 months post-transplantation. Because of the methodological differences it is difficult to compare our results with those from other studies, but the prevalence of psychosocial impairment following heart and heart-lung transplantation in the children participating in the current study seems higher than the findings reported in previous studies (Wray et al 1992, 1994; Pot-Mees 1989; Reynolds et al 1991). Using Rutter's scales, the above authors found that between 15-39% of the heart, heart-lung, bone-marrow and renal recipients had behavioural problems at home post-transplantation. The differences in these results may be due to the different instruments used. All these studies employed parental self-reports and it is possible that parents underestimate the degree of their children's difficulties. Regarding the heart and heart-lung recipients, Wray et al assessed their population 3 months after the transplant while we did it at 12 months. It is possible that the longer the follow-up

from the time of transplantation the higher the incidence of psychological disturbance. This might be because the children are no longer living through a 'euphoric post-transplantation' stage, but have experienced the first complication and rejection episodes and realise that, although free of symptoms, they still suffer from a life-threatening condition and their future is still uncertain. However, the results in the present study are not consistent with Pot-Mees's (1989) findings in that she found a significant increase in the rate of disturbance from 15% pre bone-marrow transplantation to 40% at 12 months post-surgery. The reasons for this discrepancy are unclear but there were differences in the methods used and, of course, the illness factors and medical course could be very different in these samples. Also, while Wray et al's (1994) study showed differences in psychological disturbance between the heart and heart-lung transplant recipients, the present study failed to show differences in psychosocial functioning at 12 months post-transplantation between the two groups. This discrepancy may also be due to the different methods used.

Some studies have shown that paediatric transplant recipients have fewer problems maintaining peers relationships, and showed higher rate of scholastic difficulties than children who are chronically ill or medically well children (DeBolt et al 1995; Phipps and Mulhern 1995; Fukunishi and Kudo 1995; Zamberlan 1992; Melzer et al 1989). Clinical impressions from the interviews with the children and parents seem to support these findings, but this was not empirically explored in the present study.

This study also showed that 34-52% of the children who received conventional cardiac surgery had some degree of psychosocial impairment at the follow-up. These results are in consonance with previous reports on children with congenital heart diseases (Spurkland et al 1993; Utens et al 1993; Kramer et al 1989; Garson et al 1974). However, this investigation does not confirm the Linde et al (1970) findings that cyanotic children (high-risk surgery group) had greater gains in general adjustment, attention and willingness, whilst acyanotic children (low-risk surgery group) remained the same.

Changes in the Parents' Adjustment Measures

Regarding the parents' measures, this study showed that the parents' psychological adjustment is also enhanced following transplantation or conventional cardiac surgery. However, the expected differences in the degree of improved functioning between parents of

transplant recipients and parents of children who underwent conventional cardiac surgery were not established. Furthermore, this study failed to show an improvement in marital and family adjustment at 12 months post-surgery. In addition, and with the exception of overinvolvement, this study also failed to show a lower rate of negative attitudes between family members at the follow up. In fact, parents showed lower levels of warmth towards their partners, and greater dissatisfaction with their marital relationship at 12 months post-surgery than before hand.

These findings are striking. Lower levels of parental distress following organ transplantation or conventional heart surgery have been previously reported (Pot-Mees 1989; Sormanti et al 1994; Linde et al 1970; Bentovim 1980). The parental prevalence of psychiatric disorders post-surgery is still higher than in the normal population (Goldberg 1978), and the results obtained regarding parents of transplant recipients are comparable with those reported by Reynolds et al (1991) with parents of renal recipients. Spurkland et al found that 50% of the families with a child with congenital heart-disease had chronic difficulties shown by somatic or mental health problems in the parents or siblings or intra-familial conflicts (Spurkland et al 1993). With regard to the group awaiting transplantation there was a tendency towards deterioration in the parents' psychological functioning.

Following a liberation from the demands of treatment after the transplant, a greater improvement in psychological functioning was expected in the group of parents whose children had received transplantation when compared to the group of parents whose children underwent conventional cardiac surgery. As the groups were no different with regard to the degree of improvement in parents' psychological functioning, this study showed that treatment demands were not associated with the parents' psychological functioning. Other factors may account for this, such as perceived stress, concerns about the children's future, finances, or other life events not related to the children's health.

Contrary to expectations, the empirical data gathered in the present study show that there was more marital dissatisfaction post-surgery than prior to it, and that this was unrelated to the changes in the children's physical status. There may be three alternative explanations acting independently or in combination. First, as the children become more independent and healthy, the parents also become more energetic and have more time for themselves, developing an awareness of longstanding unresolved marital difficulties, leading to greater marital

dissatisfaction. This means that prior to transplantation, and in order to cope with their dreadful circumstances, parents denied or ignored marital difficulties. The children's illness and disability had a buffering effect on the marital conflicts. Following changes in the child's role, from sick to healthy, marital difficulties intrude and come to the fore (Glaser and Bentovim 1987).

Secondly, during the transplantation and recovery period, there is an accumulation of psychosocial, interpersonal and individual factors, which affect the marital and family adjustment. Furthermore, transplantation confronts parents with existential issues, and tense differences between the parents emerge at a time of emotional pressure and intense need for the partner's support and availability. The geographical distance is an obstacle for resolution. Disillusionment with or misinterpretation of the partners' behaviours or attitudes during the process of transplantation may have a longstanding detrimental effect on the couple.

Thirdly, the way in which the family has got organised at the time of the transplant assessment has become rigid. This stops family members from acquiring new patterns of organisation to adjust to their new circumstances and, indirectly, makes the marital relationship vulnerable in the long run.

Parents of children receiving conventional surgery go through a similar but less traumatic process. However, the parents' expectations about the effect of surgery seem to affect, indirectly, family adjustment. The majority of the parents whose children received conventional high-risk cardiac surgery were expecting a full recovery following the operation. At 12 months post-operatively, a significant number of children were symptomatic, ten requiring further conventional surgery and two needing transplantation. Many parents had difficulties accepting these facts, and some blamed their partners for having put the children through surgery. In some cases the children were caught up in the parental conflicts. Subsequently, difficulties in family relationships significantly increased in this group. As pointed out by Jessop and Stein (1985), 'uncertainty' appears to be associated with more disturbances and greater perceived impact of the illness on the family.

Changes in Family Organisation

Major changes in the family organisation measures were expected between initial assessment and the follow-up. This study showed that the expressed emotion scales (EE) are not a static measures, but are sensitive to changes in the patients' physical condition and disability. This has been previously reported in studies using paediatric and adult samples (Vostanis and Nicholls 1995; Vaughn 1989). As previously noted (Linde et al 1970; Bentovim 1980), an improvement in the children's physical health following surgery was accompanied by changes in the levels of parental overinvolvement with the children. Parents were less overinvolved with their children at the follow-up, letting them be more independent and giving them more responsibility. However, mothers whose children were awaiting transplantation showed a tendency towards less overinvolvement when their children's physical health was rapidly deteriorating. It could be that, as the children's illness progresses, mothers deny the seriousness of their condition or emotionally distance themselves, as a defence mechanism to cope with their realistic fears about losing the child (Garson et al 1978; Reiss 1986). Nevertheless, it is also possible that mothers try to help their children, both in their fight for survival and in keeping normal levels of self-esteem, by doing fewer things for them and expecting the children to gain control over their lives.

Over time, fathers of heart-lung recipients became less overinvolved with their children than fathers of heart recipients. The difference in degree of less overinvolvement between these two groups of fathers is explained by the changes in medical treatment. Most heart-lung recipients had cystic fibrosis and prior to transplantation were dependent of their parents for the provision of a demanding treatment. However, heart recipients did not have such a demanding treatment prior to transplantation. This study also showed that the fathers of children who received low-risk cardiac surgery had become less overinvolved with their children over time, when compared to fathers of children who received high-risk cardiac surgery. Medical course mediates this difference between these two groups of fathers. At the follow-up, children in the low-risk group were healthy and there was no need for a long-term medical follow-up; whilst the children in the high-risk group had to be monitored at regular intervals, and some were still disabled and in need of further surgery.

As parents were less distressed at the follow-up than prior to surgery, lower rates of critical comments or hostility were expected. In fact, there was a tendency towards an increase in the

number of critical comments across the groups. This was statistically significant in the group of mothers whose children received heart-lung transplantation, and in the group of fathers whose children underwent conventional cardiac surgery. There are alternative explanations for these findings. It could be that the disclosure of the need for transplantation or cardiac surgery suppresses the parents' expressions of anger and disappointment in order to protect the children from getting upset or protect the parents themselves from their own guilt, or both. Prior to transplantation most children were confined at home with no energy to spare or misbehave. The mothers' main concern was to keep the children alive as long as possible at the same time as they were accepting the children's possible death. Under such tense circumstances discipline issues were unimportant and family members actively avoided conflicts and disagreements with the dying patient. Following a remission in the children's illness, mothers may have been less anxious about criticising them. On the other hand, it may be that the children had now adopted age-appropriate activities and the mothers are still re-adjusting to the changes in the children's behaviour and new energy. At the follow-up, fathers of children who received low-risk cardiac surgery were also more relaxed about the children's physical health, which allowed them to be more strict and objective about their children.

Over time, the fathers of children who received conventional cardiac surgery became more critical of the children than the fathers of transplant recipients. Similarly, fathers of children who received low-risk cardiac surgery had become more critical than fathers of children who received high-risk cardiac surgery. This suggests that changes in children's behaviours or illness factors following transplantation or cardiac surgery do have an effect on the fathers' response to the children's behaviours.

Surprisingly, it was also found that mothers' warmth decreased following high-risk cardiac surgery. Despite the improvement in physical health, these children still have physical symptoms and most of them might need further surgery in the near future. It may be that the uncertainty about future surgery creates a cumulative stress response in the mothers, reducing their resources for coping (Jessop and Stein 1985). These mothers might feel resentful about having to care for fragile children as opposed to the mothers of transplant recipients who, having faced the possibility of their children's death beforehand, are delighted to see him/her accomplishing age-appropriate activities, which they were unable to conduct prior to transplantation.

This study showed that not only was there an increase in negative attitudes towards the children over time, but also there was a decrease of warmth between the parents. This is not surprising, as parents were more dissatisfied with their marital relationship over time. The parents of transplant recipients become less warm towards their partners than the parents of children who received conventional cardiac surgery. Similar findings were also found when parents of children who received high-risk surgery were compared to parents of children who received low-risk surgery. These findings seem to suggest that there is a link between the levels of expressed warmth and the stresses associated with medical procedures or children's prognosis. The mechanisms explaining this link are not clear, but information from the parental interviews suggest that those mothers who were less warm towards their husbands at follow-up expressed disappointment about the amount of support and care received from their partner during the surgery and recovery period. On the other hand, those mothers, who showed more warmth and made more positive remarks at the follow-up were very appreciative and full of praise for their husbands' behaviour towards them at such a difficult time. This data seem to indicate that the parents' interaction during and after transplantation do have a long-term effect, and may be a predictor of the parents' future relationship. This needs further exploration.

Clinical observations and quantitative data from this study seem to suggest that latent problems in family dynamics may be exacerbated during the surgery process, and that transplantation may accentuate both positive and negative aspects of family relationships. Furthermore, conflicts between the parents at the time of surgery or transplantation, may still be unresolved one year later. To monitor changes over time in the siblings' psychological morbidity was not the purpose of this study. Nonetheless, during the process of the interviews, parents often gave spontaneous information regarding the high levels of disturbance in these children. They reported detrimental changes in siblings-parents relationships, and admitted feelings of guilt about it. Future research in this area should explore the effect of transplantation in the healthy siblings.

In summary, this study shows that even after successful transplantation and conventional cardiac surgery, these children are at risk of emotional problems. There are also changes in family organisation over time; this is shown by a decrease of parental overinvolvement with the children and an increase of parents' criticisms, and marital disharmony. Psychological

intervention at an early stage is recommended since, in this population, psychopathology during childhood has been associated with psychological disturbance during adult life (Baer et al 1984; Garson et al 1974; Shampaine et al 1990).

Factors associated with Children's Survival

It was hypothesised that while awaiting transplantation, children's poor adjustment, negative patterns of family organisation (negative attitudes, poor marital and family adjustment) and, poor parental adjustment would be predictors of non-survival. .

This study showed that the group of children who survived 17 months or longer while awaiting to receive heart-lung transplantation, were slightly older and had a better psychosocial level of functioning, than the group of children who did not survive. The survivors also had better physical health status and were less disabled than the group of children who passed away. This was reflected in the children's level of functioning in different areas. The children in the first group were able to attend school and to participate in greater number of social activities than the children in the later group.

There were no other associations between demographic characteristics and survival rate. Contrary to expectation, this study failed to show associations between survival, parents' psychological or marital adjustment, and patterns of family organisation. As there is an absence of prospective studies examining factors associated with survival prior to paediatric organ transplantation, comparison of the results with previous studies is not possible. However, there are two retrospective studies examining the effect of psychosocial characteristics on survival following paediatric bone-marrow transplantation. The results are contradictory. Pfefferbaum et al (1977) found no association between family relationships and survival. But McConville et al (1990) showed that the child's functional impairment, family dysfunction and paternal psychopathology were associated with unexpected physical complications and premature death. Research on adult organ transplantation has also found significant associations between depression, lack of family support, main coping mechanisms (denial or repression) and survival following renal and liver transplantation (Schulman et al 1989; Kober et al 1990; Kuchler et al 1991). However, the effect of both i) the patient's own vulnerability and protective mechanisms and, independently, ii) the quality of family relationships, on the patient's survival might be different in children and in adults. This implies that comparison of studies across different age-groups might be inadequate and misleading.

Factors associated with Better or Worse Health than expected at the Follow-Up

It was also hypothesised that following transplantation or cardiac surgery, poorly adjusted children, and children who had been exposed to negative patterns of family organisation (little or no warmth, criticisms, hostility, overinvolvement, poor marital and family adjustment), and to poor parental adjustment, would have worse physical health than expected at the follow-up, whilst well adjusted children, and children who had been exposed to positive patterns of family organisation (high warmth, lack of critical comments or hostility; little overinvolvement, and good marital or family adjustment) and to well adjusted parents would have better physical health than expected at the follow-up.

The results from the current study show that, overall, an absence of impairment in the child's psychosocial functioning and positive attitudes between family members as manifested by high warmth and positive remarks was associated with better health than expected following transplantation whilst poor marital and family adjustment, and negative attitudes between family members were associated with worse health than expected. A link between specific negative and positive patterns of family organisation and children's health were also found in the two groups of children who received conventional cardiac surgery. These findings support Wood's (1994) biobehavioural model of chronic illness which assume that the well-being of the patient depends on the physical functioning, the psychosocial functioning and the family-social functioning; and that the degree in which people respond to the patient can be beneficial when the emotions are positive ones but can be detrimental if the emotions are primary negative.

These results are also in consonance with previous research on children and adolescents with cardiac conditions, asthma, diabetes, and cystic fibrosis (Wells and Schwebel 1987; Liebman et al 1976; Hermanns et al 1989; Garrison et al 1990; Schafer et al 1986; Patterson et al 1990; Fosson et al 1991). Although using different methodologies and evaluating different aspects of family functioning, the findings of all these authors reinforce the importance of the family and its influence on the physical well being of the patients.

The number of factors associated with better or worse health than expected varies across the groups. Regarding the children who joined the active list for transplantation, the way in which the families are organised at the time of the heart transplant assessment seem to have a greater

impact on the children's health than the families' organisation at the time of the heart-lung transplant assessment. Similarly, the impact on the children's health was greater in the group of children undergoing low-risk cardiac surgery than in the group of children undergoing high-risk cardiac surgery. The reasons for this are not clear but it may be that chance, small sample size, the differences between the groups in the typology and time stages of the illness (Rolland's model, 1988), differences in the families' ability to master challenging or ambiguous situations and to change their problem solution in accordance with new data (Reiss et al 1986) may contribute to these findings. Unfortunately, these parameters were not measured in the present study.

The mechanisms mediating the link between the families' characteristics and children's health have not been established. It might be that the interrelations between the different components of the families organisation, marital adjustment, and parents psychological functioning have an impact on the quality of the parents' support and care of their children. In addition, the quality of parental care may be influenced by the children's physical status as well as their psychosocial adjustment and by the families' relationship with the professionals involved. These other parameters were not measured in the present study but they require further exploration.

Furthermore, information gathered during the parental interviews suggests that it is too simplistic to say that the children and families' psychosocial characteristics have an effect on the children's physical health. The illness has a variety of effects on the family, depending on its developmental stage, the family life-cycle stage and the developmental needs. An example of this was illustrated in chapter 5, when describing how Mr and Mrs Jordan became organised around their daughter's initial diagnosis (see pages 181-183). In addition, it has been suggested that outcome is also affected by the family members' appraisal of the situation and capacity to process the experience and attach meaning to it (Rutter 1985). Clinical experience suggests that these families need to develop new patterns of interaction to manage the demands of the chronic illness and children's disability. Subsequently, the family members attribute 'meaning' to their behaviours in the context of the children's physical condition. However, as the context changes due to the progressive course of the illness, some families failed either to acquire new interactional patterns to manage the new demands of the illness, or to attribute new meaning to their behaviours. Subsequently, family disharmony increases.

Moreover, clinical information from the parental interviews suggest that most families whose children were facing a life-threatening condition received little or no help from health professionals to understand the child's medical condition and treatment, and to make the appropriate life-style changes. Given the high levels of stress faced by these families, the genetic component of the illness and the associated parental feelings of guilt, and the lack of support available, it is not surprising that many families developed difficulties in their relationship. Perhaps, the psychosocial outcome of these families would be different if they had received more professional help to integrate the children's illness with their families' developmental needs, and to change the family members' appraisal of the situation and capacity to process the experience and attached meaning to it.

As suggested by Rolland (1988), medical outcome may also be affected by other psycho-environmental factors such as the professionals' appraisal of the situation, their message transmitted to the child and to the family, and the quality of their relationship with the family, and with other team-members (professionals' dynamics). A summary of the factors which may influence medical outcome have been summarised in FIGURE 30.

FIGURE 30: Factors Influencing Medical Outcome
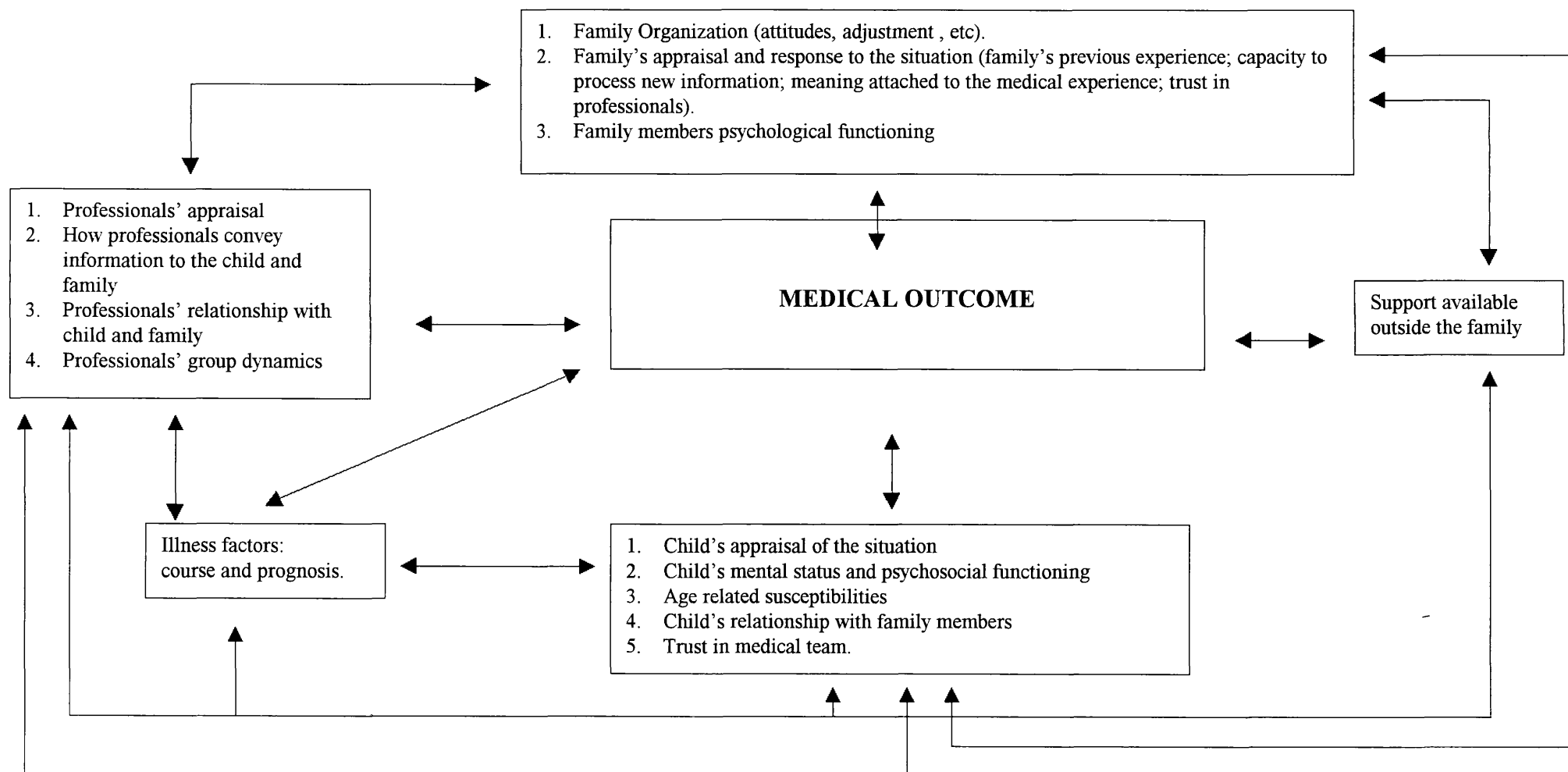
This is the first prospective study examining the incidence of unsatisfactory adherence following paediatric heart and heart-lung transplantation, and exploring psychosocial risk factors for poor adherence. Approximately one third of the children showed unsatisfactory adherence to medical treatment. Five children (9.4%) had poor adherence to immnunossuppressant therapy and 11 children (20.8%) manifested moderate adherence with other aspects of the treatment. Satisfactory adherence was found in 37 children.

These findings are consistent with previous reports on paediatric transplant recipients (Phipps and DeCuir-Walley 1990) and children with other life-threatening conditions (Tebbi et al 1986; Gordis et al 1969; Smith et al 1979). However, the prevalence of poor-adherence is much lower than that reported by Beck and colleagues (1980; 43% of paediatric renal recipients failed to take their medication). This discrepancy may be explained by the differences between subject groups and methodologies. It could also be that, in the present study, the follow-up and the reminder at each visit of the importance of medication intake and diary completion may had a positive effect in facilitating adherence to different aspects of the treatment. Longitudinal studies with children with cancer (Tebbi et al 1986; Gordis et al 1969), and a report on adult heart transplant recipients, shows that patients' adherence with annual diagnostic testing decreases significantly over 5 post-operative years (Grady et al 1993). It remains to be determined whether satisfactory adherence after paediatric HTx and HLTx decreases over time, and whether those children who failed to complete the medical diary during the first 12 post-operative months would become poorly adherent with their immunosuppressive medication later.

It was hypothesised that whilst type of transplantation would not be associated with adherence levels, socio-economic status, having a chronically ill sibling and, independently, family composition were associated with adherence. In addition, higher rates of unsatisfactory adherence were expected among disturbed children when compared to children with no psychiatric difficulties. Similarly, higher rates of unsatisfactory adherence were expected among children who were exposed to negative patterns of family organisation (negative

attitudes, poor marital and family adjustment) and to poor parental adjustment when compared with children whose families lack these characteristics.

Contrary to our expectations, there was a striking difference between the two transplant groups in that there was a much lower prevalence of adherence problems in the HTx than in the HLTx recipients. There were more adolescents in the heart-lung group, and the results showed that all children who failed to take their medication regularly were within the adolescent age range. This may explain the higher rate of unsatisfactory adherence in the HLTx recipients when compared to the HTx recipients. This association has been previously reported (Beck et al 1980; Tebbi et al 1986; Gordis et al 1969; Smith et al 1979). Age differences may be related to adolescent issues and to parents of younger children taking more responsibility for the administration of treatment. This finding emphasises the importance of bearing in mind developmental issues when working clinically with adolescents and young adults.

However, there may be more complex explanations for the difference in adherence levels between the heart and heart-lung recipients. As revealed during the interviews, the two groups were different with regard to timing prior to diagnosis. Delays in the diagnosis of children with cystic fibrosis -the majority in the HLTx group- may have jeopardised the parents' trust in the medical profession, increasing the likelihood of unsatisfactory adherence. The groups also differed with regard to the experience of illness and to the demands of treatment upon family members. Cystic fibrosis makes for greater demands, which are likely to set up a pattern of poor adherence, which in turn is maintained. Nevertheless, it is also possible that children who show poor adherence after transplant were also poor adheres beforehand (Tebbi et al 1986; Gordis et al 1969, Abbot et al 1994). In the present study, it was not possible to evaluate adherence behaviour prior to transplantation given the lack of satisfactory pre-operative measures. These alternative explanations require further exploration.

Previous studies have found that females were more at risk of unsatisfactory adherence (Korsch et al 1978; Beck et al 1980). Also contrary to expectations, gender differences were not significant. Both boys and girls during the interviews expressed distress at the side effects of the medication. It may be that with the current trend of changes in social values, and the

emphasis on body shape, adolescent boys are becoming as preoccupied about their appearance as girls.

Nearly half of the parents studied had unskilled manual occupations and some of these families had more than one child who was chronically ill. However, this study failed to show associations between levels of adherence, socio-economic status, and numbers of children living in the household with a demanding chronic condition (such as cystic fibrosis).

The hypothesised association between family structure and adherence was confirmed. Single parent families have previously been identified as a risk factor associated with unsatisfactory adherence to immunosuppressant therapy (Korsch et al 1978). This study confirmed this in that there was a lower prevalence of adherence problems in the group of children who were living with both natural parents when compared to children living with single or reconstituted families. It is striking that none of the children in this latter group showed good adherence levels. An explanation for this could be that children living with both natural parents received more support. In addition, biological fathers may take more responsibility for the administration of the children's treatment than stepfathers, and subsequently the children and their mothers may feel more supported. Nevertheless, the effect of fathers' support upon the children's adherence to treatment is unclear and the findings in the literature are contradictory (Korsch 1978; Foulkes et al 1993). The mothers participating in our study were the main carers for the children, and objective measures of fathers' involvement with the treatment-administration were not used. Nor did we explore how stepfathers were coping or how they felt about the burden of the illness.

It was found that unsatisfactory adherence in heart transplant recipients was associated with maternal hostility and low levels of expressed warmth towards her child, and with criticisms about the partner's emotional support and availability. However, due to the small number of mothers showing negative attitudes, the clinical significance of these findings remains unclear.

The families studied were not selected in any way, and represented a wide range with regard to psychosocial situations. Furthermore, due to the selection process in the other transplant centres in the U.K., we had a significant number of socially disadvantaged families. Contrary to expectations, our study failed to show that adverse psychosocial factors such as poverty,

psychiatric illness in family members, marital or family disharmony, were associated with adherence levels post-transplantation. These findings suggest that, with the current confusion in results regarding how adherence behaviour is affected by psychosocial factors, it would be wrong to use adverse psychosocial factors as a contraindication for transplantation until a better understanding is available.

Future research in this field should: a) aim to develop reliable measures of adherence prior to transplantation; b) examine whether the family's experience of illness and its meaning prior to transplantation do have an effect on adherence post-transplant; and c) include both objective and subjective measures of mothers and fathers involvement in their children's treatment.

Meanwhile, the results from this study suggest that although the rate of unsatisfactory adherence after transplantation is high there is no strong evidence to help transplant teams in the selection of the best candidates. Psychosocial consultations to support and help team members with difficult ethical dilemmas should be available. Transplant candidates and their parents need to be monitored rigorously, reinforcing the importance of medication and diary keeping, and helping the parents to transfer gradually the responsibility for medical treatment to the growing child. Support, guidance, and if necessary intervention, are particularly relevant for adolescents and single parents. Psychosocial intervention should be offered for families having difficulties accomplishing these tasks.

6.3 CRITICAL APPRAISAL OF THE STUDY

A) The strengths of the study

To my knowledge, this is the first study exploring changes over time in adjustment levels and in family organisation in a sample of candidates for heart or heart-lung transplantation, and their families. The methodology of this study is strengthened by the following factors:

- The use of standardised clinical interviews with children and parents, and the use of standard operational diagnostic criteria.

- The use of standardised self-reports completed by multiple respondents (children, parents and teachers).

- The method used to rate the Global Assessment of Functioning Scale involved the use of information gathered from both interviews (child and parent), and from the standardised questionnaires. The Rutter A-scale contains items of a physical nature ('headaches; stomach-aches; eating and sleeping habits'). Children suitable for transplantation, particularly children with cystic fibrosis, experience a number of physical symptoms and their eating and sleeping habits are disturbed due to their physical illness. Therefore, their scores could be elevated in comparison to their well peers or to the group of children undergoing low risk cardiac surgery. This could give false positive ratings suggesting that they have more behavioural or emotional problems. In this study, an attempt was made to exclude those items that might have been affected by the physical condition from which the child suffered (e.g. poor appetite, sleeping difficulties), this enabling a rating on psychosocial functioning uncontaminated by physical symptoms.

- The use of two raters to score the children's mental status, children's global assessment of functioning, family adjustment and the parental interviews, and the high inter-rater reliability. With regard to the Camberwell Family Interview, the high reliability between the two raters indicates that this instrument can be adapted and be used satisfactorily not only with relatives of physically ill children, but also with relatives of children who are approaching terminal illness.

- The use of the full Camberwell Family Interview: a growing number of studies on children with physical conditions have used the Five Minute Speech Sample as a means of assessing EE. This measures only two dimensions of the EE index: critical comments and emotional overinvolvement, and it is less reliable than the full CFI. The findings from this study suggest that the warmth scale mediates both the children's psychological adjustment and their physical well being. Therefore, studies attempting to measure attitudes of relatives of chronically ill children should attempt to use the Camberwell Family Interview rather than the Five Minutes Speech Sample so that certain mediating factors, such as warmth, are not left out.

- The design used was longitudinal and prospective. The great majority of existing studies are cross-sectional which limits the reliability of diagnostic conclusions, the examination of changes over time and risk factors.

- Within each group, the children were similar regarding the effects of severity of illness, extent of restrictions or amount of self-care involved. Furthermore, all children received transplantation in the same Transplant Centre, ensuring that the medical care and the psychological support provided by the Transplant Team was similar for all children and their families.

B) The limitations of the study

However, there were certain limitations in the methodology of this study. Due to this, we have to be careful with the interpretation of the results:

- The sample was uneven in that the proportion of families willing to participate in the study varied between the subgroups. A greater number of families whose children were assessed for transplantation agreed to participate when compared to the number of families in the conventional cardiac surgery group. Results may be biased by this, as non-participating families may have different properties to those of the families that co-operated with the research. Reasons for refusal to participate were lack of time and inconvenience in getting parents together when one of them had to be looking after the other siblings. Other families were too distressed or did not want to expose their child to further stress. The families who did not agree to participate or dropped out of the study may not be comparable to those who took part. Data on these families was not available to make any systematic comparisons.

- The sample used was not representative of children in need of transplantation or conventional cardiac surgery, as families with difficulties in English language were excluded from the study. This explains the non-representation of ethnic minorities. Therefore, we should be careful not to generalise the results to other cultural groups.

- The selection process in different transplant centres in the U.K. varies considerable. Our sample turned out to contain a higher proportion of children from lower social classes than would have been expected in an illness known not related to social disadvantage. Strong anecdotal evidence suggests that the other paediatric cardiac surgery centres were excluding children on social and psychosocial grounds, so that the Great Ormond Street group was left with an excess of children from underprivileged backgrounds. Therefore, we should be careful not to generalise the results to the population in need of paediatric heart or heart-lung transplantation.

- There were differences in the timing of the initial assessment for the different subgroups. Although candidates for transplantation and children undergoing conventional cardiac surgery, and their families, were evaluated at a time of high distress, there was a difference between the groups with regard to the timing of this initial assessment. It was not possible to assess the candidates for transplantation the day prior to surgery, as was the case with the children undergoing conventional cardiac surgery. This was due to the unpredictability of transplantation and to the impossibility of keeping the new organs for more than 5 hours once removed from the body. How the timing factor may have affected the results obtained is not known.

- The meaning of surgery was different for transplant candidates and for children undergoing conventional cardiac surgery, and their families. The experience of receiving an organ, which had belonged to somebody else, is very different from having a hole in your own heart repaired. Clinical information reveals that following the acquisition of an organ, children have all sort of fantasies about their donor, and that they are fearful of acquiring some of the donor's habits. Parents report similar fears. Whether this has any effect on the level of children's functioning, or on family organisation, remains unclear. Children receiving conventional cardiac surgery, and their families, were not exposed to these experiences.

- There were age differences between the groups. The age-range was greater in the transplantation group when compared to the conventional cardiac surgery group.

Although, all measures used were appropriate for the age-range of all participants, it is unknown whether family organisation and parents' marital and psychological adjustment was affected by the age of the children.

- The results obtained may be distorted by the following difficulties with some of the measures used:

I) The Global Assessment of Functioning Scale (GAF Scale). The Rutter Scales and the Piers & Harris Self-Concept Measure were used to obtain this rating. However, these scales were not developed to capture the 'adjustment' relevant to chronically sick children. In addition, the Piers & Harris Scale contains a number of questions regarding participation in the class, in sports etc. Children with chronic conditions and, particularly the children with cystic fibrosis participating in this study, had restricted or no opportunities to join such activities. Therefore, there is a risk their scores were artificially lowered, suggesting a worse self-concept in comparison to their well peers or to the group of children undergoing low risk cardiac surgery. The Piers & Harris Scale also failed to pick up issues that were relevant to the population assessed. For example, although clinical information revealed that following transplantation the children had serious difficulties integrating the changes in 'body image' due to the side effects of the medication, the physical appearance subscale does not have the appropriate items to assess this. This may have led to a misinterpretation of the children's ratings.

II) Regarding the ratings of family adjustment, some clinically significant changes occurred over time in some dimensions of family adjustment (communication, boundaries, cohesion, parental discord, etc.) but these were not picked up by the global Schneiderman categories used in this study. If the different components of family adjustment were rated separately, it is possible that a) the families of children undergoing transplantation would have differed in their configuration from families of children undergoing conventional cardiac surgery; and b) changes in some dimensions of family adjustment may have taken place following surgery.

III) In addition, the ratings of family adjustment were based on clinicians' judgements. Future research in this area should also take into account the family members' perception of family adjustment.

IV) The children's global rating of physical health measure used was chosen in order to compare the children' physical status across the different surgery groups

(transplantation and conventional cardiac surgery). However, in some cases, and due to the globality of the rating, this measure failed to pick up significant clinical changes in the children's health. For example, following transplantation, a few children showed a slow but progressive drop in their pulmonary function, whilst there were no changes in other health measures such as walking distance covered in 12 minutes, routine blood samples etc. The deterioration in the lung function tests was not apparent when all the results were put together.

V) Although the levels of Cyclosporine A (CyA) in blood are considered an objective measure of adherence to medical treatment, the CyA levels can oscillate because of changes in the children's immune and physiological system. Furthermore, children not taking their medication regularly can still show expected levels of CyA in blood for some time. All children who were in the poor adherence group gave verbal confirmation of not taking the medication regularly. However, it is possible that in the good and moderate adherence groups there were children who were also missing medication but this was not picked up by their CyA levels, or by the professionals who were following up these children.

- Beyond the variables studied, other variables could explain the findings. Even after successful transplantation or high risk cardiac surgery, some children and their families continued to face overwhelming medical crises, frequent hospitalisations, and reminders of the children's uncertainty about their future. Some children were more exposed to unpleasant events related to the illness than others. The present study did not control for other illness variables that could affect medical course and psychological adjustment.

- There are reports on chronic illness suggesting that parent-child communication, conflict resolution, parental attitudes and beliefs, pre-existing sources of stress, the meaning that the family ascribes to the illness, family resources in managing stressors associated with the course of illness, and life events that are unrelated to the illness have an effect on the physical well being of the children (Wilson et al 1996; Gotz 1996; King et al 1994; Wysocki 1993; Tse et al 1987; Koch-Hattem 1987). The present study did not control for such variables.

6.4 THE IMPLICATIONS OF THE STUDY

- Theoretical Implications

The theoretical framework of this study is an attempt to view the illness process in the context of the family as a psychosocial system. Many of the findings can be best understood as being part of an interactive process in which physical, psychological and family factors have a mutually reinforcing effect on each other. For intance, changes over time in the children's physical and psychological well being were associated with changes in the parents' adjustment levels, and with changes in the family organisation. This is shown by the changes found in the marital adjustment ratings and in parental attitudes towards both the children and partners. At the same time, children and parents' adjustment, and the families' patterns of organisation were shown to have an effect on disease activity over time. Positive patterns of family organisation were associated with better health than expected; while children and parent adjustment and negative patterns of family organisation were associated with worse health than expected. Subsequently, associations between psychosocial factors and disease activity were found in all surgical groups.

These results also suggest that there is a mutual influence of psychosocial and physical factors, regardless of the particular psychosocial and developmental challenges and sequelae of the cardio-respiratory illnesses (Rolland's psychosocial typology; 1994). The results from this study support the Biobehavioural Family Model of chronic illness in children and adolescents (Wood 1994) in that they suggest that the well-being of the patient depends on a dynamic balance between individual physical functioning, individual psychological functioning, and the family-social functioning (see Figure 31).

The results are consistent with Wood's hypothesis, which suggests that some interactions can either buffer or activate psychobiological processes related to disease activity. According to the biobehavioural model, the boundary dimensions of proximity and hierarchy are shaped by and enhance or modulate responsivity in families. This model predicts that high proximity, weak hierarchy, high responsivity, triangulation, and parental discord will

FIGURE 31: Biopsychosocial Approach: Levels of Functioning
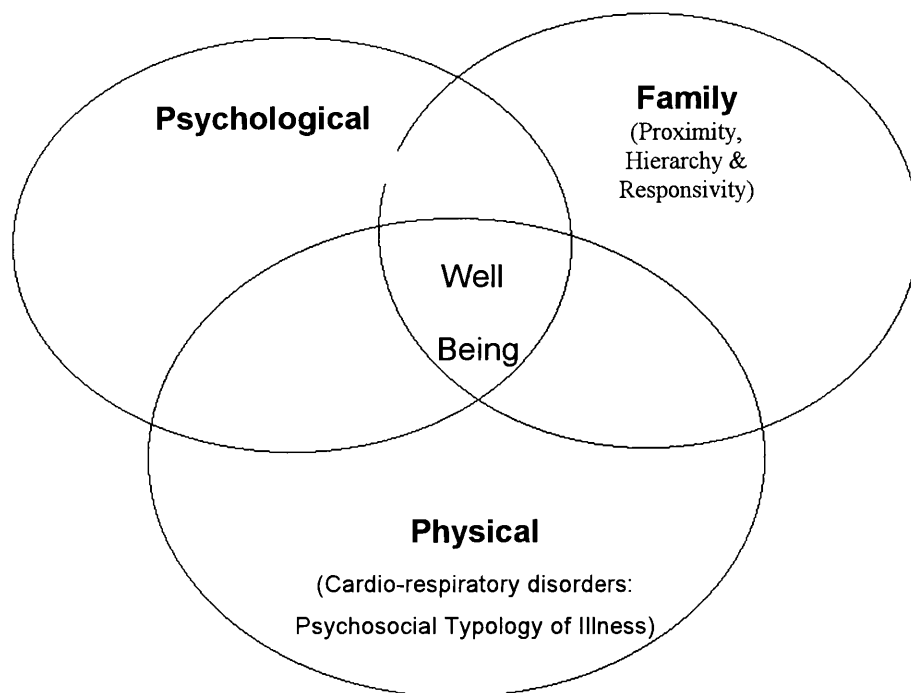
undermine physical and psychosocial well being (Wood 1994). There are similarities between these parameters and that used in the present study. Wood's definition of high proximity is similar to that of high overinvolvement (expressed emotion scales), as both connote a high degree of involvement, invasion of personal space and overprotection of the patient, this interfering with normal development and autonomy. In the present study, weak generational hierarchy, triangulation and parental discord were part of the global rating of family adjustment. Unfortunately, due to the way the study was set up, it was not possible to measure these domains separately, measuring their reliability and validity. Positive attitudes of warmth and positive remarks, and negative attitudes such as criticism and hostility are different manifestations of responsivity between family members.

According to this model, at times there is competition between these three levels of functioning, which may result in an imbalance between these factors. Within this framework, quantitative and clinical information obtained suggests that biopsychosocial imbalances might occur in several ways in the families studied. Examples of these are given below:

1.- Balance between physical functioning and family adjustment:

1.1 - Physical functioning, and its associated stresses and demands, can compromise family boundaries (this is defined as "rules defining who participates and how"), having an effect on the overall family adjustment. Families rated as moderately-poorly adjusted presented at least one of the following interactional patterns: enmeshment, overprotection, rigidity, triangulation and poor conflict resolution. At initial assessment, there were a high number of families showing one or more of these sequences. In the framework of this study, these sequences were not seen as pathological as these families were attempting to adjust to the tremendous stresses associated with the illness stage.

1.2 - Families were expected to change their interactional patterns or sequences as the children's physical health improved following surgery. However, there was a tendency for family adjustment to remain unchanged over time. The data seem to suggest that, overall, these families failed to accommodate to the changes in the children's illness. An adaptive family response at an earlier stage of the illness may become maladaptive if it remains unchanged during further stages. This had a negative effect on the children's disease activity. The occurrence of new traumatic events (rejections, infections, biopsies, etc) was specific to the families of transplant recipients; these promoting further family disorganisation and disruptions. Because of the continuous need to accommodate to new events and situations, it is not surprising that these families had difficulties achieving more adaptive sequences.

1.3 - Patterns that optimise the children's medical management and psychosocial adjustment may have negatively affected marital adjustment. The deterioration that was found in the parents' marital adjustment could be the result of this. It could be that the parents participating in the study neglected their marriage in order to provide intensive physical and emotional care of their ill children. The neglect may have happened prior to the referral or could be a consequence of the stresses associated with medical procedures at the time of the children's admission for surgery.

2. Balance between individual psychological and family adjustment:

2.1 - Family patterns that improve physical well being can undermine the children's psychosocial functioning and autonomy. This was particularly relevant for children with cystic fibrosis. Physiotherapy three times a day can help in containing the disease activity but may also be a burden for these children's social life, jeopardising their social and

personal development. This also applies to good adherence with hospital appointments, and is particularly relevant for those children who needed frequent follow-ups. Similarly, some parents, regardless of the children's typology of illness, who were concerned about their children getting infections and diseases, restricted their children's social activities, including school attendance.

2.2 - Family patterns that may improve adherence to treatment can undermine the child's mastery over the illness and their bodies, jeopardising the children's active responsibility in the management of and adaptation to their chronic illness. Most parents of transplant recipients were retaining full responsibility for the treatment provision (administration of tablets and completion of diaries), regardless of the children's age.

2.3 - Family patterns of overprotection, cross-generational coalitions of the child with one parent against the other, or conflict detouring through attention to the child or illness, and child's dependency would undermine psychosocial functioning.

3. Balance between physical and individual psychological adjustment:

3.1 - Activities that improved the children's psychological functioning may have a detrimental effect on the physical well being. Medical treatment can be neglected in favour of other functions. Post-transplantation and avoiding the side effects of medication, some adolescents regularly postponed taking drugs that made them feel dizzy in order to participate in physical activities with their peers.

This study failed to support the notion of families of children with different severity of illness would present different family configurations. While it is possible that the family measures used did not assess the relevant domains, the data indicate that the way families adjust to chronic or life-threatening illness is more dependent on family characteristics than on specific demands made by the illness. Future research on chronic physical illness using the biopsychosocial model should explore how different components of family functioning activate psychophysiological pathways to influence the disease process.

- Clinical Implications

It is clear from the discussion of the findings of this research that there are important implications for clinicians involved with children referred for transplantation assessment. The

disclosure of the need for transplantation represents a significant life crisis and is a devastating event for all family members. The interaction between the developmental stages of the children, the illness and the families is crucial in inducing maladaptive family interactions. Families are confronted with multiple and complex dilemmas and challenges; with physical, emotional, social and financial demands. When the children are transferred from the provisional to the active waiting list for transplantation, the children and their families are facing a new stage in the development of the illness: the anticipation of inevitable death. Even after successful transplantation, the uncertainty about the children's future remains.

The quantitative analysis of data has shown that adjustment difficulties in children referred for transplantation or conventional cardiac surgery, and in their parents, are common. It has also shown that there is a link between parental adjustment and the children's psychosocial functioning. Parental distress, and mothers' distress in particular because of her role and involvement with the children, can jeopardise parental ability to deal with the children's illness, and may bring on denial, minimisation, anger, withdrawal, and difficulties administering medical treatment. The identification of poorly adjusted children and parents is important in order to provide preventive psycho-educational counselling to reduce levels of stress and to enhance children's and parents' coping skills prior to medical interventions. Of course, the effectiveness of such counselling requires evaluation and monitoring.

The deterioration in the marital relationship over time, and the high rate of family maladjustment both before and after surgery, suggest the need for a systematic monitoring of family relationships and for family intervention. Clinical impressions gathered during the interviews seem to indicate the existence of problematic areas presented by these families before, during and after transplantation. These are shown in TABLE XLV. Treatment efforts should focus on loosening inappropriate coalitions, re-defining the patients' role, dealing with marital conflict and maintaining appropriate parenting. Furthermore, due to the associations found between negative patterns of family organisation –that is, negative attitudes between family members- and the children's physical well being over time, the clinician should make an effort to identify families at risk, referring them to a liaison mental health professional for preventive family work.

This study also showed that adolescents are more at risk of unsatisfactory adherence post-transplantation than younger groups. Changes in body image and appearance, their need for more independence, and peer pressure to 'be', 'look' and 'behave normally' are important factors. Preparation for the changes in body shape and appearance should commence while awaiting transplantation. Group counselling sessions before and after transplantation could be an effective way to provide education and support for candidates and recipients. In situations where independent issues are a source of conflict for the family, or if there are indications of psychiatric disorder, prompt and intensive intervention is warranted, as this can be a very dangerous situation for the ill adolescent.

Adolescence is a particularly crucial stage for subjects who are chronically ill because this is the time when identity formation is a primary task. Care must be taken to encourage this population to develop a firm boundary between the disease and their identity. Parents, health professionals and school staff can assist healthy identity formation by striking a balance between firm limit setting and nurturance, while expecting the ill adolescent to function as normally as possible.

No doubt all families, whether single or two-parent, are likely to encounter stresses in their attempts to cope with the demands of transplantation. However, the evidence of the present study indicates that single parents and parents in stepfamilies have greater difficulties than natural two-parent families in coping with the demands of treatment. This is probably due to poorer resources. It is important that the professionals guard against prejudice; and given the dependence of transplanted children on their parents for adequate nurturing and care, the need for effective intervention should be recognised. Clinicians can play a key role in evaluating parental resources, offering guidance and extra support, or arranging adequate support in the community (e.g. regular home visits from a health

TABLE XLV: FAMILY DIFFICULTIES-CLINICAL IMPRESSIONS

The Illness Phases (acute, chronic and terminal) and Family Transitions	Each deterioration in the child's physical status means more disability and altered quality of family life requiring constant re-adaptation. It is not uncommon for families to cling to a process they used in the past. Healthy coalitions, which become established in response to the illness at one stage, may become frozen in time, preventing the family from moving on and changing (transition from illness to remission), affecting family relationships negatively.
Gender Differences	Overall, fathers prefer to work through painful issues quietly on their own; while mothers wish to talk at length about their thoughts, feelings, anxieties. Some fathers may feel pushed to focus on the illness more than they want/can cope with. Some mothers may interpret their partners' attitude as a personal rejection. These differences in style may lead to marital difficulties.
Secrecy between Family Members	Secrecy about the poor prognosis may act as a barrier between them at the time when all members feel most distressed, vulnerable and in need of supporting each other.
Focusing on Illness	Excessive focusing of the illness may lead to unnecessary restriction of enjoyable activities. The needs of other family members can become neglected, which results in frustration, resentment, misbehaviour (siblings) and poor communication.
Illness Progression	Even if transplantation has been successful, families may feel increasingly overwhelmed by the continuing medical monitoring, frequent hospitalisations and continuing life-threatening condition. Families with unresolved conflicts have more difficulties adjusting to the illness progression than healthy functioning families.

Overprotection	Protectiveness is necessary to care for children facing a life- threatening condition. Parents need to keep a balance between appropriate protectiveness and family overprotection, which may lead, to less personal autonomy, loss of self-esteem and more disability in the child.
Isolation	Family's isolation in coping with the illness may maintain rigid patterns of interaction. Some families have great difficulties sharing their concerns about the illness between: family members, extended family and/or other important sources of support in the community.
Unresolved grief	Families who have already lost a child may have unresolved issues regarding their previous loss. This may interfere with their current family interactions, attitudes and expectations about transplantation. Furthermore, in same cases the dead sibling has been idealised affecting parent-ill child relationship.
Anticipation of Death	Following successful transplantation, the child's development and the quality of the family relationships may be jeopardised when families have organised themselves not only around the child's illness but also around the anticipated death of the child. This pattern may be seen in some families whose child was actually dying when, unexpectedly, organs become available and the child survived. The family had already said good bye to the child and was coming to terms with his/her loss.
Dealing with Death Issues	Many parents have great difficulties communicating about death. Often, the dying child is aware of what is happening but avoids asking questions about the future in an attempt to protect his/her family from the distress. The child has no one with whom to share his fears and feels desperately lonely. This pattern prevents families from dealing with unfinished emotional concerns, be intimate with one another and, say good-bye.

visitor or community nurse). Effective preventive and support programmes should be implemented prior to the transplant.

Early detection of both children at risk and highly distressed parents is desirable. Although the focus of attention in this study has been the ill child and their parents, the clinical information obtained from the parental interviews at 12 months post-transplantation suggests that some siblings are badly affected. The parent-sibling relationship may undergo a dramatic change during the process of transplantation. Health professionals should also be aware of the siblings' difficulties and address transplantation as a 'family affair', including when possible all members of the family in the discussions. How can professionals help these families to respond in a healthy way to the challenges of the illness?. TABLE XLVI contains some strategies for managing families of children referred for transplantation

TABLE XLVI: CLINICAL INTERVENTIONS

1. Provide a psychoeducational approach educating the family about the illness, prognosis and treatment. Helping the family to take responsibility for treatment management and encouraging open communication with medical team.

2. Provide social support and positive connotation for the adaptations they have made, emphasizing their strengths, enhancing self-esteem and respecting their defences.

3. Remove feelings of guilt and blame and normalize unacceptable feelings (anger, resentment, bitterness, etc.).

4. Encourage honest communication throughout the family.

5. Help the families to attend to the fit between 'stage of illness' and 'stage of family development', discussing this relationship with them. Help families to attend to the needs of the illness but not allowing the illness to dominate the families' schedules or organise the family's emotional life.

6. Prepare the child and family to face the stresses of the transplant process and to anticipate the changes in the family system prior and post-transplantation.

7. Assist the parents in anticipating changes in the child's energy/behaviour following the transplant.

8. Help parents to communicate, support and be aware of each other's needs, before, during and after transplantation. This is particularly important when one parent is living in the hospital and the other is at home with the siblings.

9. To help the child and parents to anticipate medical events and changes in physical appearance due to the medication.

10. Help families to communicate openly about death when appropriate.

11. Helping the medical team to discuss the illness and course when there is ambiguity and uncertainty

12. Liaising with all agencies involved in the child's care in the community.

Due to the fact that most children live far from the hospital, it might be appropriate to organise ongoing support locally. Therefore, close liaison between the transplant centre and the local professionals is required. There is extremely limited scientific evidence that the measures suggested here are effective, but my own clinical experience and that of others suggest that they are worth undertaking and deserve further evaluation.

The findings from this research, together with the clinical expertise developed since 1989 by the Transplant Team at Great Ormond Street Hospital, have lead to considerable changes in the clinical service offered to children referred for transplantation and their families. The modifications introduced are as follows:

1. Currently, all children accepted into the transplant programme have a mental health key-worker allocated who will follow the child and his/her family before, during and after transplantation. The key-worker also organises support for the family in the community and liaises with the school personnel.

2. The provision of education about the illness and the side effects of the medication have been strengthening. Special emphasis is given to adherence to treatment issues and to the associated changes in body appearance. Children, from 9 years of age, are encouraged to take an active part in the management of their treatment, shifting gradually the responsibility towards self-management.

3. The medical and the mental health members of the transplant team work more closely together, assisting each other and seeing the child and the family together when appropriate.

In summary, possible candidates for transplantation and their families are in considerable emotional distress. The effect of the illness on the family is profound, but equally the family

relationships can have both a positive and a negative effect on both adjustment and the course of illness. In addition, the experience of transplantation is frightening for both children and parents and all may benefit from the expression and sharing of their thoughts, feelings and concerns, reducing anxiety and strengthening good family functioning. This research has highlighted the importance of monitoring the psychological well being of children referred for transplantation as well as monitoring the well being of other family members. Professionals working with these families should offer both ongoing systematic assessments of the family situation and support before, during and after transplantation. to help the families to adjust to new medical events and uncertainty about the child's future.

- Ethical Implications

The psychological assessment of children seeking organ transplantation presents a potential for ethical conflict. The number of children dying while awaiting transplantation is rapidly increasing. This is because the number of patients who may potentially benefit from transplantation far exceeds the supply of organs available. Because of these factors, there is pressure on the transplantation teams to be more selective and choose to operate on the patients with a better prognosis. Current thought appears to encourage health choices based on the personal and moral responsibility of physicians and health care workers. In fact, many rehabilitation programmes now use psychosocial assessments as part of their eligibility criteria. Some candidates are given low priority by virtue of psychosocial impairment (Surman and Purtilo 1994), and in some countries, personal and family responsibility are now playing a role in government decisions for health care budgets (Caplan 1994). This raises some important questions: on which principles can life-preserving resources be provided to some while being denied to others? And, if there are principles, do we have data that either support them or demand that we revise them? Do psychosocial data have a place in making choices and assessing principles? Four interconnected principles have been described in Medical Ethics: respect for autonomy, beneficence, nonmaleficence and justice (Beauchamp and Childress 1994).

The results from the present study are of relevance to the principles of beneficence, nonmaleficence and justice. The present study has shown that, overall, transplantation is beneficial to children and their families on both physical and psychosocial grounds.

However, some issues remain unclear, such as the probability of both physical benefit and harm that transplantation offers to each individual child, and whether transplantation induce a lot of psychosocial pathology in siblings or other family members.

This study has failed to show that psychosocial factors are associated with survival rate. With regard to the principle of justice, therefore, it is hightly questionable to decline transplantation to a particular child because of adverse individual or family factors. Associations were found between the recipient's physical well being and psychosocial factors, and between psychosocial factors and adherence to treatment. However, even if one were to accept in principle that this could be a factor in deciding about the suitability for treatment, the results of the present study are not sufficiently robust to warrant any simple conclusion and need to be treated with considerable caution. Furthermore, clinical data have shown that some children from seriously disturbed families do really well after transplantation. It has also shown that well adjusted children and families prior to transplantation may become chaotic following transplantation, and that members of the transplant team are unable to predict which children and families will do well or become disorganised following transplantation (Serrano-Ikkos 1996). In addition, associations between the children's physical health and families' characteristics were also present in the two groups of children undergoing conventional cardiac surgery. This raises another the ethical question: is it right to treat children in need of transplantation and children in need of conventional cardiac surgery, and their families differently? In conclusion, empirical data and clinical information have shown that psychosocial factors do not have sufficient predictive value with respect to maleficence. Thus, even putting aside the need for respect for peoples moral and legal rights, the only justifiable approach at the present state of knowledge is to focus on physical aspects of the disease and not to include psychosocial factors in deciding how to allocate the resources. More studies are required to look into:

a). Long-term qualitative and quantitative follow-up of both transplant candidates and recipients, with and without adverse psychosocial factors, to detect discrepancies between these two groups; and

b) Intervention studies aimed to evaluate the effectiveness of psychological interventions in diminishing negative interactional patterns and helping parents to support each other and the child. A number of studies have shown the effectiveness of family intervention in introducing positive changes in the parent-child relationship, and in other aspects of family functioning and

that this has a beneficial effect on the physical well being of children (Lask and Matthew 1979; Koblenzer and Koblenzer 1988; Liebman et al 1976; Lask 1988).

In summary, while the clinical and research experience at Great Ormond Street Hospital for Children brings into question the use of psychosocial and social variables in selecting candidates for transplantation, it demonstrates clearly that ongoing psychological support should be available for all the family, regardless of whether the family has adverse psychosocial factors or not. Special attention should be paid to the marital relationship and to other children in the family. Until further data to the contrary is available, scrupulous attention to ethical considerations should be given priority. During the selection process we should bear in mind the 4 principles in medical ethics: respect for autonomy, beneficence, nonmaleficence, and justice. Unless this is done, the professional's prejudice may interfere and lead to more harm than good and neglect of simple but powerful ethical principles.

6.6 CONCLUDING STATEMENTS

Paediatric transplantation is now a treatment of choice for children with a variety of life-threatening conditions. As survival rates are improving because of advanced technology, the shortage of donor organs is a major impediment and the number of children dying while waiting for suitable donors is rapidly increasing. Currently, it is estimated that 60-70% of the children waiting to be transplanted at Great Ormond Street will die prior to transplantation.

Uncertainty regarding the duration of the illness or its ultimate outcome probably remains the greatest single psychosocial stressor facing children and adolescents who are candidates for heart/heart-lung transplantation, and their families. As Koocher (1986) has pointed out, this is more difficult for the paediatric patient whose developmental changes are progressing at a more rapid pace than for an adult patient who has established basic life activity patterns. This study highlights the importance of multidisciplinary teams, and their key role in detecting children and families at risk. The clinical objectives of the team working with these families are effective prevention and therapeutic intervention.

This study shows that, as one might expect, no family member is left untouched by the transplant experience regardless of outcome. The long-term effects of transplantation and cardiac surgery may be profound and there is a need for prospective studies in this field. It also suggests the need: a) to monitor ongoing changes in family relationships at the time of surgery and during the recovery period, and b) for an intervention trial aimed at ameliorating the adverse effects of transplantation and conventional surgery on both the children's psychological well-being and the family relationships by helping parents in their negotiation of new roles, to support and communicate with each throughout the process as well as preparing them for future medical events and for the children's return home and school life.

Although children with cardio-respiratory diseases have unique characteristics associated with their diseases, there are features that are common to other chronic and life-threatening illnesses, such as:

1. The tendency of chronic illness to jeopardise psychosocial well being and development.

2. Additional demands (emotional, financial, time, etc) on the carers.

3. The need for changes in family structure and in family organisation to accommodate to the changes in illness typology.

4. The tendency for negative patterns of family organisation and poor parental adjustment conflicting directly or indirectly with disease management.

5. The tendency of the illness to interfere with school attendance, patients' and all family members' social life, and parents' working life.

6. The tendency of the chronic illness to interfere with the parents-sibling relationships.

Improvement in clinical care of children with chronic diseases often goes hand-in-hand with research developments. Research is this field is difficult because of the methodological problems created by rapid changes in pharmacological treatments and surgical techniques. In addition, the mortality rate in populations suffering with life-threatening disorders is very high. Despite these difficulties, multidisciplinary teams should attempt to integrate clinical and research methods into routine practice, providing the best possible care for the children and their families. Mental health professionals should play a key role assisting members of the medical team in developing a better understanding of how physical and psychological factors interact with each other.

Future research in paediatric organ transplantation should attempt to answer key clinical questions such as:

1. The role of behavioural and psychosocial factors in graft rejection and the mechanisms by which these factors may contribute to tissue rejection.

2. The effect of infant, child and adolescent organ transplantation on long-term growth and psychosocial development.

3. The relationship between pre-transplant psychiatric and psychosocial status and specific post-transplant medical outcome, including survival status.

4. What the child knows and understands about the illness and its treatment, and the way treatments and their consequences interact with different levels of intellectual, emotional and social development?

5. Early identification of non-compliers?

6. Which factors differentiate those families who cope better with the adversity of illness from those families whom cope badly?

7. Identification of effective methods of providing psychological support for these children and their families. Intervention studies should aim to examine whether a significant proportion of long-term psychiatric morbidity and secondary medical morbidity and mortality in paediatric transplant recipients is potentially preventable through psychosocial intervention.

REFERENCES

Abbott J., Dodd M., Bilton D. and Webb A.K. (1994). "Treatment compliance in adults with cystic fibrosis". Thorax, 49, 115-120.

Addonizio L.J. and Rose E. A. (1987). "Cardiac transplantation in children and adolescents". Journal of Pediatrics, 111, (part 2); 1034-1038.

Anderson B.J., Auslander W.F., Jung K.C., Miller J.P. and Santiago J.V. (1990). "Assessing family sharing of diabetes responsibilities". Journal of Pediatric Psychology, 15, 4, 477-492.

Andrykowski M.A. (1994). "Psychiatric and psychosocial aspects of bone marrow transplantation". Psychosomatics, 35, 13-24.

Asarnow J.R., Goldstein M.J., Tompson M. and Guthrie D. (1993). "One-year outcomes of depressive disorders in child psychiatric in-patients: Evaluation of the prognostic power of a brief measure of expressed emotion". Journal of Child Psychology and Psychiatry, 34, 129-137.

Backer C.L., Zales V.R., Idriss F.S., Lynch P., Crawford S., Woodrow M. et al (1992). "Heart transplantation in neonates and in children". Journal of Heart and Lung Transplantation, 11, 311-319.

Baer P.E., Freedman D.A. and Garson A. (1984). "Long-term psychological follow-up of patients after corrective surgery for Tetralogy of Fallot". Journal of the American Academy of Child Psychiatry, 23, 622-625.

Bailey L.L., Wood M., Razzouck A., Van Arsdell G. and Gundry S. (1989). "Heart transplantation during the first 12 years of life". Archives of Surgery, 124, 1221-1226.

Bailey L., Gundry S., Razzouk A., Wang N. and the Loma Linda University Pediatric Heart Transplant Group (1992). "Pediatric heart transplantation: issues relating to outcome and results". Journal of Heart and Lung Transplantation, 11, S267-S271.

Baker L.C. and Patterson J.M. (1989). "Introduction to family theory". In C N Ramsey, editor. Family Systems in Medicine. Guilford Family Therapy Series, Guilford; 1989:57-62.

Baker L.C. "Critical issues in family theory in family medicine research". In: CN Ramsey, editor. Family Systems in Medicine. Guilford Press, New-York-London; 1989:150-163.

Bartko J. and Carpenter W.T. (1976). "On the methods and theory of reliability". Journal of Nervous and Mental Disease, 163, 5, 307-317.

Beath S.V., Brook G.D., Kelly D.A., Cash A.J., McMaster P., Mayer A.D. and Buckels J.A.C. (1993). "Successful liver transplantation in babies under 1 year". British Medical Journal, 307, 825-828.

Beachamp T.L. and Childress J.F. Principles of Biomedical Ethics. New York, Oxford. Oxford University Press, 1994.

Beck D.E., Fennell R.S., Yost R.L., Robinson J.D., Geary D., and Richards G.A. (1980). "Evaluation of an educational program on compliance with medication regimens in pediatric patients with renal transplants". Journal of Pediatrics 96, 1094-1097.

Bentovim A. (1980). "Psychological and social aspects of cardiac disease in children". In: G Graham and E Rossi, editors. Heart Disease in Infants and Children. Edward Arnold; 1980:179-185.

Berkowitz R. (1987). "Rating expressed emotion from initial family therapy sessions (a pilot study): Journal of

Family Therapy, 9, 27-37.

Bernstein D.M. (1971). " After transplantation - the child's emotional reactions". American Journal of
    Psychiatry, 127, 1189-1193.

Bernstein D.M.. "Psychiatric assessment of the adjustment of transplanted children". In: RG Simmons, SD
    Klein, RL Simmons, editors. Gift of life: the social and psychological impact of organ transplantation.
    Wiley, New York; 1977:119-147.

Blackwell B. (1973). "Drug therapy: patients compliance". New England Journal of Medicine, 289, 249-252.

Blair C., Freeman C. and Cull A (1995). "The families of anorexia nervosa and cystic fibrosis patients".
    Psychological Medicine, 25, 985-993.

Blotcky A.D., Cohen D.G., Conatser C., Klopovitch P. (1985). "Psychosocial characteristics of adolescents who
    refused cancer treatment". Journal of Consulting Clinical Psychology, 53, 729-731.

Bluebond-Langer M. The private worlds of dying children. Princeton, NJ, Princeton University Press, 1978.

Bluebond-Langer M. In the shadow of illness: parents and siblings of the chronically ill child. Princeton, NJ,
    Princeton University Press, 1996.

Bolman III R.M., Braunlin E., Shumway S.J., Hertz M.I. and Warwick W. (1994). "Pediatric lung and heart-
    lung transplantation at the University of Minnesota". Transplantation Proceedings, 26, 203-204.

Bolman III R.M. (1994). "Pediatric lung and heart-lung transplantation". Transplantation Proceedings, 26, 211-
    213.

Bradford R. and Tomlison L. (1990). "Psychological guidelines in the management of paediatric organ
    transplantation". Archives of Disease in Childhood, 65, 1000-1003.

Bradford R. (1991). "Children's psychological health status - the impact of liver transplantation: a review".
    Journal of the Royal Society of Medicine, 84, 90-94.

Bradford R.. Children, families and chronic disease. Psychological models and methods of care. Routledge,
    London and New York, 1997.

Breslau N. (1985). "Psychiatric disorder in children with physical disabilities". Journal of the American
    Academy of Child Psychiatry, 24, 87-94.

Brown G.W., Monck E.M., Carstairs G.M. and Wing J.K. (1962). "Influence of family life on the course of
    schizophrenic illness". British Journal of Preventive and Social Medicine, 16, 55-68.

Brown G.W. and Rutter M. (1966). "The measurement of family activities and relationships: a methodological
    study". Human Relations, 19, 241-263.

Bywater E.M. (1981). "Adolescents with cystic fibrosis: psychosocial adjustment". Archives of Disease in
    Childhood, 56, 538-543.

Cadman D., Boyle M., Szatmari P. and Offord D.R. (1987). "Chronic illness, disability and mental and social
    well-being: findings of the Ontario child health study". Pediatrics, 79, 5, 805-813.

Caplan A.L. (1994). "Ethics of casting the first stone: personal responsibility, rationing, and transplants".
    Alcoholism-Clinical and Experimental Research, 18, 219-221.

Cappelli M., McGrath P.J., MacDonald N.E., Katsanis J. and Lascelles M. (1989). "Parental care and
    overprotection of children with cystic fibrosis". British Journal of Medical Psychology, 62, 281-289.

Chacko R.C., Harper R.G., Kunik M. and Young J. (1996). "Relationship of psychiatric morbidity and

psychosocial factors in organ transplant candidates". Psychosomatics, 37, 100-107.

Chadwick J. and Mann W.N. (1950). The medical works of Hippocrates. Springfield, Ill, Thomas.

Chaney J.M. and Peterson L. (1989). "Family variables and disease management in juvenile rheumatoid arthritis". Journal of Pediatrics and Psychology, 14, 3, 389-403.

Colonna J.O.H., Brems J.J., Hiatt J.R., Millis J.M., Ament M.E., Baldrich-Quinones W.J. et al (1988). "The quality of survival after liver transplantation". Transplantation Proceedings, 20, supplement 1, 594-597.

Cook W.L., Strachan A.M., Goldstein M.J. and Miklowitz D.J. (1989). "Expressed emotion and reciprocal affective relationships in families of disturbed adolescents". Family Process, 28, 337-348.

Copeland D.R. and Davidson E.R. "Comprehensive treatment of the child with cancer". In :MC Roberts and CE Walker editors. Casebook of Child and Paediatric Psychology. Guilford Press, New York, 1989:319-345.

Costell R., Reiss D., Berkman H. and Jones C. (1981). "The family meets the hospital: predicting the family's perception of treatment programme from its problem solving style". Archives of General Psychiatry, 38, 567-577.

Cousens P., Waters B., Said J.and Stevens M. (1988). "Cognitive effects of cranial irradiation in leukemia: a survey and meta-analysis". Journal of Child Psychology and Psychiatry, 29, 839-852.

Cowen L., Mok J., Corey M., MacMillan H., Simmons R. and Levison H. (1986)."Psychologic adjustment of the families with a member who has cystic fibrosis". Pediatrics, 77, 745-753.

Cummings E.M. and Davies P.T. (1994). "Maternal depression and child development". Journal of Child Psychology and Psychiatry, 35, 73-112.

Davis H. (1993). "Counselling parents of children with chronic illness or disability". British Psychological Society.

DeBolt A.J., Stewart S.M., Kennard B.D., Petrik K., et al (1995). "A survey of psychosocial adaptation in long-term survivors of pediatric liver transplants". Children's Health Care, 24, 268.

DeMaso D.R., Campis L.K., Wypij D., Bertram S., Lipshitz M. and Freed M. (1991). "The impact of maternal perceptions and medical severity on the adjustment of children with congenital heart disease". Journal of Paediatric Psychology, 16, 137-149.

Dennis C., Caine N., Sharples L., Smyth R., Higenbottam T., Stewart S. et al (1993). "Heart-lung transplantation for end-stage respiratory disease in patients with cystic fibrosis at Papworth Hospital". Journal of Heart and Lung Transplantation, 12, 894-902.

Dermatis H. and Lesko L.M. (1990). "Psychological distress in parents consenting to child's bone marrow transplantation". Bone Marrow Transplantation, 6, 411-417.

Diseth T.H., Emblem R. and Vandvik I.H. (1995). "Adolescents with anorectal malformations and their families: examples of hidden psychosocial trauma". Family System Medicine, 13, 215-231.

Doane J.A.; Goldstein M.J.; Miklowitz D.J. and Falloon I.R.H (1986). "The impact of individual and family treatment of the affective climate of families of schizophrenics". British Journal of Psychiatry, 148:279-287.

Doane J.A. and Diamond D. " A review of previous studies on expressed emotion, affective style, and

attachment". In: JA Doane and D Diamond editors. Affect and attachment in the family. A family-based treatment of major psychiatric disorder. BasicBooks, Harper Collins Publishers, New York; 1994:11-37.

Dossetor D.R., Nicol A.R., Stretch D.D. and Rajkhowa S.J. (1994). "A study of expressed emotion in the parental primary carers of adolescents with intellectual impairment". Journal of Intellectual Disability Research, 38,487-499.

Drotar D., Doershuk C.F., Stern R.C., Boat T.F., Boyer W. and Matthews L. (1981). "Psychosocial functioning of children with cystic fibrosis". Pediatrics, 67, 338-343.

Dunbar J.. "Compliance in Paediatric Populations: A Review". In: PJ McGarth and P Firestone editors. Pediatric and Adolescents Behavioural Medicine. Springer, New York; 1983:211-230.

Dunn J., Golden D., Van Buren C.T., Lewis R.M., Lawen J. and Kahan B.D. (1990). "Causes of graft loss beyond two years in the cyclosporine era". Transplantation, 49, 349-353.

Eiser C. (1990)."Psychological effects of chronic disease". Journal of Child Psychology and Psychiatry, 31, 85-98.

Eiser C., Havermans T., Pancer M., and Eiser J.R. (1992). "Adjustment to chronic disease in relation to age and gender: mothers' and fathers' reports of their childrens' behavior". Journal of Pediatric Psychology, 17, 261-275.

Eiser C. "Consequences of parenting styles for children's adjustment to treatment". In: Eiser C, editor. Growing up with a chronic disease, Jessica Kingsley Publishers, London-Philadelphia;1993:155-175.

Eiser C., Parkyn T., Havermans T. and McNinch A. (1994). "Parents' recall on the diagnosis of cancer in their child". Psycho-Oncology, 3, 197-203.

Eiser C., Havermans T. and Eiser J.R. (1995). "Parents' attributions about childhood cancer: implications for relationships with medical staff". Child: care, health and development, 21, 31-42.

Engel G.L. (1977). "The need for a new medical model: a challange for biomedicine". Science, 196, 129-136.

Engstrom I. (1992)."Mental health and psychological functioning in children and adolescents with inflammatory bowel disease: a comparison with children having other chronic illness and with healthy children". Journal of Child Psychology and Psychiatry, 33, 563-582.

Fennell R.S.III, Rasbury W.C., Fennell E.B. and Morris M.K. (1984). "Effects of kidney transplantation on cognitive performance in pediatric population". Pediatrics, 74, 273-278.

Fischmann-Havstad L. and Marston A.R. (1984). "Weight loss maintenance as an aspect of family emotion and process". British Journal of Clinical Psychology, 23, 265-271.

Flanagan D.A.J. and Wagner H.L. (1991). "Expressed emotion and panic-fear in the prediction of diet treatment compliance". British Journal of Clinical Psychology, 30, 231-240.

Fleiss J.L. . "Bonferroni criterion". In: JL Fleiss, editor. The design and analysis of clinical experiments. John Wiley and Sons; 1986:103.

Fosson A., Wilson J., Kanga J. and D'Angelo S. (1991). "Interplay of physical, psychological and family factors in cystic fibrosis: Treating variables in 35 children for 24 months". Pediatric Pulmonology, Supplement, 7, 351.

Foulkes L.M., Boggs S.R., Fennell R.S. and Skibinski K. (1993). "Social support, family variables, and

compliance in renal transplant children". Pediatric Nephrology, 7, 185-188.

Freeman A., Davis L., Libb J.W. and Craven J. "Assessment of transplant candidates and prediction of
outcome". In: J Craven and GM, editors. Psychiatric Aspects of Organ Transplantation. Oxford
University Press; 1992:9-21.

Friedman I.M. and Litt N.F. (1986). "Promoting adolescents' compliance with therapeutic regimen". Pediatric
Clinics of North America, 33, 955-973.

Frist W.H., Fox M.D., Campbell P.W., Fiel S.B., Loyd J.E. and Merrill W.H. (1991). "Cystic fibrosis treated
with heart-lung transplantation: North American Results". Transplantation Proceedings, 23, 1205-
1206.

Fukunishi I. and Kudo H. (1995). "Psychiatric problems of pediatric end-stage renal failure". General Hospital
Psychiatry, 17, 32-36.

Fundudis T. (1997). "Single parents: risk or resource". Child Psychology and Psychiatry Review, 2, 2-14.

GAF SCALE (1987). Quick reference to the diagnostic criteria from DSM-III-R. The American Psychiatric
Association.

Gardner G.G., August C.S. and Githens J. (1977). "Psychological issues in bone marrow transplantation".
Pediatrics, 60, 625-631.

Garrison W.T., Biggs D. and Williams K. (1990)."Temperament characteristics and clinical outcomes in young
children with diabetes mellitus". Journal of Child Psychology and Psychiatry, 31, 1079-1088.

Garson A., Williams R.B. and Reckless J. (1974). "Long term follow-up of patients with tetralogy of fallot:
physical health and psychopathology". Journal of Pediatrics, 85, 429-433.

Garson A., Benson S., Ivler L. and Patton C. (1978). "Parental reactions to children with congenital heart
disease". Child Psychiatry and Human Development, 9, 86-94.

Geiss S.K., Hobbs S.A., Hammersley-Maercklein G., Kramer J.C. and Henley M. (1992). "Psychosocial factors
related to perceived compliance with cystic fibrosis treatment". Journal of Clinical Psychology, 48, 99-
103.

Gilhooly M.L.M. and Whittick J.E. (1989). "Expressed emotion in caregivers of the dementing elderly". British
Journal of Medical Psychology, 62, 265-272.

Glaser D. and Bentovim A. "Psychological aspects of congenital heart disease". In: RH Anderson, J Macartney,
EA Shinebourne and M Tynan, editors. Paediatric Cardiology. Churchill Livingstone; 1987: 1371-
1383.

Gold L.M., Kirkpatrick B.S., Fricker F.J. and Zitelli B.J. (1986). "Psychosocial issues in pediatric organ
transplantation: The parents' perspective". Pediatrics, 77, 738-744.

Goldberg I. (1978). "General health questionnaire (GHQ-28)". NFER-NELSON.

Goldberg S., Morris P., Simmons R.J., Fowler R.S. and Levison H. (1990). "Chronic illness in infancy and
parenting stress: A comparison of three groups of parents". Journal of Paediatric Psychology, 15, 347-
358.

Goldman L.S. (1993). "Liver transplantation using living donors: preliminary donor psychiatric outcomes".
Psychosomatics, 34, 235-240.

Gordis L., Markowitz M., and Lilienfeld A.M. (1969). "Studies in the epidemiology and preventability of

rheumatic fever. Part IV. A quantitative determination of compliance in children on oral penicillin prophylaxis". Pediatrics, 43, 173-182.

Gotz I (1996). "Psychosocial background of families with a child with cystic fibrosis". Israel Journal of Medical Sciences, 32, S66-S67.

Gottesfeld I.B. (1979). "The family of the child with congenital heart disease". American Journal of Maternal Child Nursing, March/April, 101-104.

Grady, K.L., Russell, K.M., Srinivasan S., Costanzo M.R., and Pifarre R. (1993). "Patient Compliance with Annual Diagnostic Testing After Heart Transplantation". Transplantation Proceedings, 25, 2978-2980.

Gustafsson P., Kjeliman N. and Cederblad M. (1986). "Family therapy in the treatment of severe childhood asthma". Journal of Psychosomatic Research, 30, 369-375.

Hammen C., Gordon D., Burge D., Adrian C., Jaenicke C. and Hiroto D. (1987). "Communication patterns of mothers with affective disorders and their relationships to children's status and social functioning". In: K Hahlweg and MJ Goldstein, editors. Understanding Major Mental Disorder. The Contribution of Family Interaction Research. Family Process, New York; 1987:103-119.

Hanson C.L., Henggeler S.W., and Burghen G.A. (1987). "Model of assocations between psychological variables and health outcome measures of adolescents with I.D.D.M". Diabetes Care, 10, 752-758.

Hanson C.L., de-Guire M. J., Schinkel A.M., Henggeler S.W., et al (1992). "Comparing social learning and family systems correlates of adaptation in youths with IDDM. Special issue: Theory-driven research in pediatric psychology". Journal of Pediatric Psychology, 17, 555-572.

Hare J., Skinner D. and Kliewer D. (1989). "Family system approach to pediatric bone marrow transplantation". Children Health Care, 18, 30-36.

Haynes R.B., Taylor D.W. and Sackett D.L. (1979). Compliance in health care. Baltimore: Johns Hopkins University Press.

Hermanns J., Florin I., Dietrich M., Rieger C. and Hahlweg K. (1989). "Maternal criticism, mother-child interaction, and bronchial asthma". Journal of Psychosomatic Research, 33, 469-476.

Herrick C.M., Mealey P.C., Tischner L.L. and Holland C.S. (1987). "Combined heart failure transplant program: advantages in assessing medical compliance". Journal of Heart Transplant, 6: 141-146.

Hesse U.J., Roth B., Knuppertz G., Wienand P., and Lilien T. (1990). "Control of patient compliance in outpatient steroid treatment of nephrologic disease and renal transplant recipients". Transplantation proceedings, 22, 1405-1406.

Hibbs E.D., Hamburger S.D., Lenane M., Rapoport J.L., Kruesi M.J.P., Keysor C.S., and Goldstein M.J. (1991). "Determinants of expressed emotion in families of disturbed and normal children". Journal of Child Psychology and Psychiatry, 32,757-770.

Hibbs E.D., Zahn T.P., Hamburger S.D., Kruesi M.M.P. and Rapoport J.L. (1992). "Parental expressed emotion and psychophysiological reactivity in disturbed and normal children". British Journal of Psychiatry, 160, 504-510.

Hibbs E.D., Hamburger S.D., Kruesi M.S.P., Lenane M.S.W. (1993). "Factors affecting expressed emotion in parents of ill and normal children". American Journal of Orthopsychiatry, 63, 103-112.

Hodes M. and Le Grange D. (1993). "Expressed emotion in the investigation of eating disorders: A review". International Journal of Eating Disorders, 13, 279-288.

Hoffman J.I.E.. "Incidence, mortality and natural history". In: RH Anderson, FJ Macartney, EA Shinebourne and M Tynan, editors. Paediatric Cardiology, Volume 1. Churchill Livingstone;1987:1-14.

Hooley J.M. (1986). "Expressed emotion and depression: interactions between patients and high-versus-low-expressed-emotion spouses". Journal of Abnormal Psychology, 95, 237-246.

Hooley J.M. and Teasdale J.D. (1989)."Predictors of relapse in unipolar depressives: expressed emotion, marital distress, and perceived criticism". Journal of Abnormal Psychology, 98, 229-235.

Hows J.M., Yin J.L., Marsh J., Swirsky D., Jones L., Apperley J.F. et al (1986). "Histocompatible unrelated volunteer donor with HLA nonidentical family donors in marrow transplantation for aplastic anemia and leukemia". Blood, 68, 1322-1328.

Hutchings S.M. and Monett Z.J. (1989). "Caring for the cardiac transplant patient". Critical Care Nursing Clinics of North America, 1, 245-261.

Invernizzi G., Bressi C., Bertrando P., Passerini A., Giannelli A., Clerici M., Biglioli P. and Cazzullo C.L. (1991). "Emotional profiles of families with a heart-operated patient: A pilot study". Psychotherapy and Psychosomatics, 55, 1-8.

Jadresic E (1988). "Expresividad emocional familiar y epilepsia". Revista Chilena de Neuro-Psiquiatria, 26,26-31.

Jessop J.D. and Stein R.E.K. (1985). "Uncertainty and its relation to the psychological and social correlates of chronic illness in children". Social Sciences and Medicine, 20, 993-999.

Jordan S.C. and Scott O. (1989). "Heart disease in paediatrics". Butterworths.

Kanabar D.J., Attard-Montalto S., Saha V., Kingston J.E., Malpas J.E. and Eden O.B. (1995). "Quality of life in survivors of childhood cancer after megatherapy with autologous bone marrow rescue". Pediatric Hematology and Oncology, 12, 29-36.

Kavanagh D.J. (1992). "Recent developments in expressed emotion and schizophrenia". British Journal of Psychiatry, 160,601-620.

Khan A.V., Herndon M.A. and Ahmadian S.Y. (1971). "Social and emotional adoptions of children with transplanted kidneys and chronic hemodialysis". American Journal of Psychiatry, 127, 1194-1198.

King M., Speck P. and Thomas A. (1994). "Spiritual and religious beliefs in acute illness: is this a feasible area for study? ". Social Science and Medicine, 38, 631-636.

Kinston W. and Bentovim A. (1978). "Brief focal family therapy when the child is the referred patient: II. Methodology and results". Journal of Child Psychology and Psychiatry and Allied Disciplines, 19, 119-143.

Kober B., Kuchler T., Broelsch C., Kremer B. and Henne-Bruns D. (1990). "A psychological support concept and quality of life research in a liver transplantation program: an interdisciplinary multicenter study". Psychotherapy and Psychosomatics, 54, 117-131.

Koblenzer C.S. and Koblenzer P.J. (1988). "Chronic intractable atopic eczema". Archives of Dermatology, 124, 1673-1677.

Koch-Hattem A. (1987). "Families and chronic illness". Family Therapy-Collections, 22, 33-50.

Koenigsberg H.R., Klausner E., Pelino D., Rosnick P. et al (1993). "Expressed emotion and glucose control in insulin-dependent diabetes mellitus". Americal Journal of Psychiatry, 150, 1114-1115.

Koenigsberg H.R., Klausner E., Chung H., Pelino D. et al (1995). "Expressed emotion and warmth: extending the EE contrast to insuin-dependent diabetes mellitus". International Journal of Mental Health, 24,50-63.

Kog E., Vertommen H. and Degroote T (1985). "Family interaction research in anorexia nervosa: the use and misuse of self-report questionnaire". International Journal of Family Psychiatry, 6, 227-243.

Kong S.G., Tay J.S.H., Yip W.C.L. and Chay S.O. (1986). "Emotional and social effects of congenital heart disease in Singapore". Australian Paediatric Journal, 22, 101-106.

Koocher G.P. (1986). "Psychosocial issues during the acute treatment of paediatric cancer". Cancer 58: 468-472

Korsch B.M., Gardner J.E., Fine R.N. and Negrete V.F. (1972). "Long-term follow up on kidney transplant patients and their families". Proceedings of the European Dialysis and Transplant Association, 9, 359-363.

Korsch B.M., Negrete V.F., Gardner J.E., Weinstock C.L., Mercer A.S., Grushkin C.M. et al (1973). "Kidney transplantation in children: psychosocial follow-up study on child and family". Journal of Pediatrics, 83, 399-408.

Korsch B.M., Fine R.N., and Negrete V.F. (1978). "Noncompliance in children with renal transplants". Pediatrics, 61, 872-876.

Kovacs M., Iyengar S., Goldston D., Obrosky D. et al (1990). "Psychological functioning among mothers of children with insulin-dependent diabetes mellitus: A longitudinal study". Journal of Consulting and Clinical Psychology, 58, 189-195.

Kramer H.H., Awiszus D., Sterzel U., van Halteren A. and Claßen R. (1989). "Development of personality and intelligence in children with congenital heart disease". Journal of Child Psychology and Psychiatry, 30, 299-308.

Kramer J.H., Crittenden M.R., Halberg F.E., Wara W.M. and Cowan M.J. (1992). "A prosceptive study of cognitive functioning following low-dose cranial radiation for bone marrow transplantation". Pediatrics, 90, 447-450.

Kuchler T., Kober B., Brolsch C., Henne-Bruns D. and Kremer B. (1991). "Quality of life after liver transplantation: can a psychosocial support program contribute?". Transplantation Proceedings, 23, 1541-1544.

Kuhn W.F., Davis M.H. and Lippmann S.B. (1988). "Emotional adjustment to cardiac transplantation". General Hospital Psychiatry, 10, 108-113.

Kuhn W.F., Brennan A.F., Lacefield P.K., Brohm J., Skelton V.D. and Gray L.A (1990). "Psychiatric distress during stages of the heart transplant protocol". Journal of Heart Transplantation, 9, 25-29.

La Greca A.M. . Adherence to prescribed medical regimens. In: D Routh, editor. Handbook of Pediatric Psychology. Guilford Press, New York; 1988:299-320.

La Greca A.M. (1990). "Issues in adherence with pediatric Regimens". Journal of Pediatric Psychology, 15, 4, 432-436.

Lask B. (1988). "Psychosocial factors in childhood diabetes and seizure disorders". Paediatrician. 15, 95-101.

Lask B. (1994)."Non-Adherence to treatment for Cystic Fibrosis". Journal of the Royal Society of Medicine, 87, 25-27.

Lask B. and Fosson A. Childhood illness: the psychosomatic approach. Children talking to their bodies. John Wiley and Sons Ltd., Chichester, England, 1989.

Lask B. and Matthew D. (1979)."Childhood asthma: a controlled trial of family psychotherapy". Archives of Disease in Childhood, 54, 116-119.

Lawrence K.S. and Fricker F.J. (1987). "Pediatric heart transplantation: quality of life". Journal of Heart Transplantation, 6, 329-333.

Lee M.L., Cohen S.E., Stuber M.L. and Nader K. (1994). "Parent-child interactions with pediatric bone marrow transplant patients". Journal of Psychosocial Oncology, 12, 43-60.

Leff J. and Vaughn C. "Expressed emotion in families". The Guildford Press. New York-London, 1985.

Lesko L.M. (1994). "Bone marrow transplantation: support of the patient and his/her family". Support Care Cancer, 2, 35-49.

Lewis B.L. and Khaw K.T. (1982). "Family functioning as a mediating variable affecting psychosocial adjustment of children with cystic fibrosis". Journal of Pediatrics, 101, 636-640.

Le Grange P.D. (1989). "Anorexia nervosa and family therapy: a study of changes in the individual and the family during the process of body weight restoration". PhD dissertion.

Le Grange P.D. (1993). "Family therapy outcome in adolescent anorexia nervosa". South African Journal of Psychology, 23, 174-179.

Le Grange D., Eiser I., Dare C. and Hodes M. (1992). "Family criticism and self-starvation: a study of expressed emotion". Journal of Family Therapy, 14, 177-192.

Liebman R., Minuchin S., Baker L. and Rosman B.L.. "The role of the family in the treatment of chronic asthma". In: PJ Guerin, editor. Family Therapy, Theory and Practice. Gardner Press; 1976:309-324.

Linde L.M. (1982). "Psychiatric aspects of congential heart disease". Psychiatric Clinics of North America, 5, 399-406.

Linde L.M., Rasof B., Dunn O.J. and Rabb E. (1966). "Attitudinal factors in congential heart disease". Pediatrics, 38, 92-101.

Linde L.M., Rasof B. and Dunn O.J. (1970). "Longitudinal studies of intellectual and behavioral development in children with congenital heart disease". Acta Paediatrica Scandinavica, 59, 169-176.

Litt I.F. and Cuskey W.R.(1980). "Compliance with medical regimens during adolescence". Pediatric Clinic of North America, 27, 3-15.

Macartney F.J., Taylor J., Graham G.R., De Leval M., Stark J. (1980). "The fate of survivors of cardiac surgery in infancy". Circulation, 62, 80-91.

Madden B.P., Kamalvand K., Chan C.M., Khaghani A., Hodson M.E. and Yacoub M. (1993). "The medical management of patients with cystic fibrosis following heart-lung transplantation". European Respiratory Journal, 6, 965-970.

Mai F.M., McKenzie F.N. and Kostuk W.J. (1990). "Psychosocial adjustment and quality of life following heart transplantation". Canadian Journal of Psychiatry, 35, 223-227.

Maricle R.A., Hosenpud J.D., Norman D.J., Woodbury A. et al (1989). "Depression in patients being evaluated for heart transplantation". General Hospital Psychiatry, 11, 418-424.

Mascaretti L., Pappalettera M., Pizzi C., Bossi G., Scalamogna M. and Sirchia G. (1991). "Four years of heart transplantation in the North Italy transplant program (NITp)". Transplantation Proceedings, 23, 1186-1188.

Melzer S.M., Leadbeater B., Reisman L., Jaffe L.R., et al (1989). "Characteristics of social networks in adolescents with end-stage renal disease treated with renal transplantation". Journal of Adolescent Health Care, 10, 308-312.

Metras D., Kreitmann B., Shennib H. and Noirclerc M. (1992). "Lung transplantation in children". Journal of Heart and Lung Transplantation, 11, S282-S285.

McConville B.J., Steichen-Asch P., Harriw R., Neudorf S., Sambrano J., Lampkin B. et al (1990). "Pediatric bone marrow transplants: psychological aspects". Canadian Journal of Psychiatry, 35, 769-775.

McDaniel S.H., Hepworth J. and Doherty W.J.. Medical family therapy: a biopsychosocial approach to families with health problems. BasicBooks, Haper Collins Publishers, Inc., New York, 1992.

McFarlane A.H., Bellissimo A. and Norman G.R. (1995). "Family structure, family functioning, and adolescents well-being: the transcendent influence of parental style". Journal of Child Psychology and Psychiatry, 36, 847-864.

Minichin S., Rosman B.L. and Baker L. . Psychosomatic families: anorexia nervosa in context. Cambridge: Harvard University Press, 1978.

Mulhern R.K., Crisco J.J. and Camitta B.M. (1981). "Patterns of communication among pediatric patients with leukemia, parents, and physicians: Prognostic disagreements and misunderstandings". Journal of Pediatrics, 99, 480-483.

Muszynski-Kwan A.T., Perlman R., and Rivington-Law B.A. (1988). "Compliance with and effectiveness of chest physiotherapy in cystic fibrosis: a review". Physiotherapy Canada, 40, 28-32.

Notarius C.I. and Pellegrini D.S.. "Differences between husbands and wives: Implications for understanding marital discord". In: K Hahlweg and MJ Goldstein, editors. Understanding Major Mental Disorder. The Contribution of Family Interaction Research. Family Process Press, New York; 1987:231-249.

O'Dougherty M., Wright F.S., Garmezy N., Loewenson R.B. and Torres F. (1983). "Later competence and adaption in infants who survive severe heart defects". Child Development, 54, 1129-1142.

Orford J., O'Reilly P. and Goonatilleke A. (1987). "Expressed emotion and perceived family interaction in the key relatives of elderly patients with dementia". Psychological Medicine, 17, 963-970.

Overholser J.C. and Fritz G.K. (1990). "The impact of childhood cancer on the family". Journal of Psychosocial- Oncology, 8, 71-85.

Pahl E., Fricker F.J., Trento A., Griffith B., Hardesty R., Gold L. et al. (1988). "Late follow-up of children after heart transplantation". Transplantation Proceedings, XX, 1, 743-746.

Parness I.A. and Nadas A.S. (1988). "Cardiac transplantation in children". Pediatrics in review, 10, 111-118.

Parsons R., Goodwin M., Bickerton A. and Lask B. (1996). "Mirrored grief : the systemic context of paediatric heart/lung transplantation". Accepted for publication in Clinical Child Psychology and Psychiatry, 1(2).

Passero M.A., Remor B., and Salomon J. (1981). "Patient-reported compliance with cystic fibrosis therapy". Clinical Pediatrics, 20, 264-268.

Patenaude A.F., Szymanski L. and Rappeport J. (1979). "Psychological costs of bone marrow transplantation in children". American Journal of Orthopsychiatry, 49, 409-422.

Patenaude A.F. and Rappeport J. (1982). "Surviving bone marrow transplantation: the patient in the other bed". Annals of Internal Medicine, 97, 915-918.

Patterson J.M. (1985). "Critical factors affecting family compliance with home treatment for children with cystic fibrosis". Family Relations 34, 79-89.

Patterson J.M.. "A family stress model: the family adjustment and adaptation response". In: CN Ramsey, editor. Family Systems in Medicine. Guilford Press, New-York-London; 1989:95-118.

Patterson J.M. and McCubbin H.I. . "Chronic illness: Family stress and coping". In: CR Figley and HI McCubbin, editors. Stress and the Family. Volume II, Coping with Catastrophe. Brunner/Mazel, New York; 1983:21-36.

Patterson J.M., McCubbin H.L. and Hamilton I. (1983). "The impact of family life events and changes on the health of a chronically ill child". Journal of Applied Family and Child Studies, 32, 255-264.

Patterson J.M., McCubbin H.I. and Warwick W.J. (1990). "The impact of family functioning on health changes in children with CF". Social Sciences and Medicine, 31, 159-164.

Patterson J.M., Leonard B.J., Titus J.C. (1992). "Home care for medically fragile children: impact on family health and well-being". Journal of Developmental and Behavioural Pediatrics, 13, 248-255.

Perrin J.M., MacLean W.E. and Perrin E.C. (1989). "Parental perceptions of health status and psychological adjustment of children with asthma". Pediatrics, 83, 26-30.

Petterson G., Ahren C., Berggren H., Berglin E., Edvardsson N., Hoyer S. et al (1990). "Heart transplantation in Gothenberg, 1988 through 1989". Transplantation Proceedings, 22, 188.

Pfefferbaum B., Lindamood M.M. and Wiley F.M. (1977). "Pediatric bone marrow transplantation: psychosocial aspects". American Journal of Psychiatry, 134, 1299-1301.

Phipps S. and DeCuir-Whalley (1990). "Adherence Issues in Pediatric bone marrow transplantation". Journal of Pediatrics and Psychology, 15, 459-475.

Phipps S. and Mulhern R (1995). "Family cohesion and expressiveness promote resilience to the stress of pediatric bone marrow transplant: A preliminary report". Journal of Developmental and Behavioural Pediatrics, 16, 257-263.

Piers V.E. and Harris D.B. (1984). Piers and Harris children's self-concept scale. Western Psychological Services, Los Angeles, California.

Pless I.V. and Satterwhite B.B. ."Family functioning and family problems". In: RJ Haggerty, KJ Roghmann and IB Pless, editors. Child Health and the Community. A Wiley-Interscience Publication, London, New-York, Sydney, Toronto; 1975:41-54.

Pot-Mees C.. "The psychosocial effects of bone marrow transplantation in children". Eburn Delft, 1989.

Poznanski E.O., Miller E., Salguero C. and Keish R.C. (1978). "Quality of life for long-term survivors of end-stage renal disease". Journal of the American Medical Association, 239, 2343-2347.

Radley-Smith R.C. and Yacoub M.H. (1992). "Long-term results of pediatric heart transplantation". Journal of

Heart and Lung Transplantation, 11, S277-S281.

Rasbury W.C., Fennell R.S. and Morris M.K. (1983). " Congnitive functioning of children with end-stage renal disease before and after successful transplantation". Journal of Pediatrics, 102, 589-592.

Reither A.N.. "Psychiatric aspects of transplantation". In: SL Smith editor. Issues and organ transplantation. American Association of Care Nurses. Masby-Year book; 1990:117-143.

Reiss D., Gonzalez S. and Kramer N. (1986). "Family process, chronic illness, and death". Archives of General Psychiatry, 43, 795-804.

Reiss D. "Families and their paradigms: an ecologic approach to understanding the family in its social work". In CN Ramsey, editor. Family systems in medicine. Guilford Publications; 1989: 119-133.

Reynolds J.M. , Garralda M.E., Postlethwaite R.J. and Goh D. (1991). "Changes in psychosocial adjustment after renal transplantation". Archives of Disease in Childhood, 66, 508-513.

Rodin G. and Abbey S. . "Kidney transplantation". In: J Craven and G Rodin, editors. Psychiatric aspects of organ transplantation. Oxford Medical Publications; 1992:145-164.

Rodin G and Voshart K. (1987). "Depressive symptoms and functional impairment in the medically ill". General Hospital Psychiatry, 9, 251-258.

Rodriguez A., Diaz M., Colon A. and Santiago-Delpin E.A. (1991). "Psychosocial profile of noncompliant transplant patients". Transplant Proceedings, 23, 1807-1809.

Rolland J.S. (1984). "Towards a psychosocial typology of chronic and life-threatening illness". Family System Medicine, 2, 245-263.

Rolland J.S. Families, illness, and disability: an integrative treatment model. BasicBooks, HarperCollins Publishers, Inc., 1994.

Rund B.J., Oie M., Borchgrevink T.S. and Fjell A. (1995). "Expressed emotion, communication deviance and schizophrenia". Psychopathology, 28, 220-228.

Rust J., Bennun I., Crowe M., Golombok S. (1985). "The Golombok Rust Inventory of Marital State (GRIMS)". NFER-NELSON.

Rutter M. (1967). "A childrens behaviour questionnaire for completion by teachers: preliminary findings". Journal of Child Psychology and Psychiatry, 8, 1-11.

Rutter M. (1985). "Resilience in the face of adversity. Protective factors and resistance to psychiatric disorder". British Journal of Psychiatry, 147, 598-611.

Rutter M. (1989). "Isle of Wight revisited: twenty-five years of child psychiatry epidemiology". Journal of the American Academy of Child and Adolescent Psychiatry, 28, 633-653.

Rutter M. and Graham P. (1968). "The reliability and validity of the psychiatric assessment of the child: interview with the child". British Journal of Psychiatry, 114, 563-579.

Rutter M., Tizard J. and Whitmore K. (1970). "Education, health and behaviour". Longmans: London (reprinted Krieger: New York 1981).

Samuelson R.G., Hunt S.A. and Schroeder J.S. (1984). "Functional and social rehabilitation of heart transplant recipients under age thirty". Scandinavian Journal of Thoracic and Cardiovascular Surgery, 18, 97-103.

Satel S.L. (1990). "Mental status changes in children receiving glucocorticoids: review of the literature".

Clinical Pediatrics, 29, 382-388.

Satin W., la Greca A.M., Zigo M.A. and Skyler J.S (1989). "Diabetes in adolescence: effects of multifamily group intervention and parent simulation of diabetes. Journal of Pediatric Psychology, 14, 259-275.

Schafer L.C., McCaul K.D. and Glasgow R.E. (1986). "Supportive and nonsupportive family behaviours: Relationships to adherence and metabolic control in persons with type I diabetes". Diabetes Care, 9, 179-185.

Schneiderman G., Lowden J.A. and Rae-Grant Q. (1976). "Family reactions, physicians responses, and management issues in fatal lipid storage diseases". Clinical Pediatrics, 15, 887-890.

Schobinger R., Florin I., Reichbauer M., Lindemann H. and Zimmer C. (1993). "Childhood asthma: mothers' affective attitude, mother-child interaction and children's compliance with medical requirements". Journal of Psychosomatic Research, 37, 697-707.

Schreiber J.L., Breier A. and Pickar D. (1995). "Expressed emotion: trait or state?". British Journal of Psychiatry, 166, 647-649.

Schulman R., Price J.D.E. and Spinelli J. (1989). "Biopsychosocial aspects of long-term survival on end-stage renal failure therapy". Psychological Medicine, 19, 945-954.

Schwartz C.E., Dorer D.J., Beardslee W.R. Lavori P.W. (1990). " Maternal expressed emotion and parental affective disorder: Risk for childhood depressive disorder, substance abuse, or conduct disorder". Journal of Psychiatric Research, 24, 231-250.

Sensky T., Stevenson K., Magrill L. and Petty R. (1991). "Family expressed emotion in non-psychiatric illness: adaptation of the Camberwell Family Interview to the families of adolescents with diabetes". International Journal of Methods in Psychiatric Research, 1, 39-51.

Serrano-Ikkos E. (1996). "Who should live when all can not? ". Israel Journal of Medical Sciences, 32, S103-S105. Paper presented at the XIIth International Cystic Fibrosis Congress. Jerusalem, Israel.

Serrano-Ikkos E., Lask B., Whitehead B. (1997). "Psychosocial morbidity in children and their families, awaiting heart or heart-lung transplantation". Journal of Psychosomatic Research, 42, 253-260.

Shaben T.C. (1993). "Psychosocial issues in kidney-transplanted children and adolescents: literature review". ANNA Journal, 20, 663-668.

Shaffer D., Gould M.S., (1983). "A children's global assessment scale". Archives of General Psychiatry,40, 1228-1231.

Shampaine E.L., Nadelman L., Rosenthal A., Behrendt D. and Sloan H. (1990). "Longitudinal psychological assessment in Tetralogy of Fallot". Paediatric Cardiology, 10, 135-140.

Sharples L.D., Caine N., Mullins P., Schofield P. M., Large S. R. and Wallwork J. (1991). "Analysis of survival data after transplantation and its risk factors for graft loss". Transplantation Proceedings, 23, 1167-1169.

Sharples L.D., Scott J.P., Dennis C., Higenbottam T.W., Stewart S., Wreghitt T. et al (1994). "Risk factors for survival following combined heart-lung transplantation". Transplantation, 57, 218-223.

Shope (1981). "Medication Compliance". Pediatric Clinics of North America, 28, 5-21.

Smedler A.C, Ringden K., Bergman H. and Bolme P. (1990). "Sensory-motor and cognitive functioning in children who have undergone bone marrow transplantation". Acta Paediatrica Scandinavica, 79, 613-

621.

Smith S.D., Rosen D., Trueworthy R.C., and Lowman J.T. (1979). "A reliable method for evaluating drug compliance in children with cancer". Cancer, 43, 169-173.

Smyth R.L., Higenbottam T.W., Scott J.P., McGoldrick J.P., Whitehead B., Helms P. et al (1989). "Early experience of heart-lung transplantation". Archives of Disease in Childhood, 64, 122-1230.

Smyth R.L., Scott J.P., Higenbottam T.W., Whitehead B., Helms P., Sharples L. et al (1990a). "The use of heart-lung transplantation in management of terminal respiratory complications of cystic fibrosis". Transplantation Proceedings, 22, 1472-1473.

Smyth R.L., Scott J.P., Whitehead B., Fradet G., Helms P., Higenbottam T., de Leval M. and Wallwork J. (1990b). "Heart-lung transplantation in children". Transplantation Proceedings, 22, 1470-1471.

Somerville J. (1989). "Congenital heart disease in the adolescent". Archives of Disease in Childhood, 64, 771-773.

Somerville J. (1990). "Grown-up survivors of congenital heart disease: Who knows? Who cares?". British Journal of Hospital Medicine, 43, 132-136.

Sormanti M., Dungan S. and Rieker P.P. (1994). "Pediatric bone marrow transplantation: psychosocial issues for parents after a child's hospitalization". Journal Of Psychosocial Oncology, 12, 23-42.

Spurkland I., Bjornstad P.G., Lindberg H. and Seem E. (1993). "Mental health and psychosocial functioning in adolescents with congenital heart disease. A comparison between adolescents born with severe heart defect and atrial septal defect". Acta Paediatrica, 82, 71-76.

Starnes V.A. (1993). "Lobar transplantation: indication and outcome". Journal of Heart Lung Transplantation, 12, S94.

Steele C.I. and Altholz J.A.S. (1987). "Donor ambivalence: a key issue in families of children with end-stage renal disease". Social Work in Health Care, 13, 47-57.

Steinberger J., Haines H.C., Shumway S.J., Bolman III R.M., Rocchini A.P. and Braunline E.A. (1993). "Outcome after referral for pediatric transplantation". Journal of Heart and Lung Transplantation, 12, 766-769.

Steinhausen H. (1987). "Global assessment of child psychopathology". Journal of the American Academy of Child and Adolescent Psychiatry, 26, 203-206.

Steinhausen H. and Schindler H. (1981). "Psychosocial adaption in children and adolescents with cystic fibrosis". Developmental and Behavioural Pediatrics, 2, 74-77.

Steinhausen H., Schindler H. and Stephan H. (1983). "Correlated of psychopathology in sick children and an empirical model". Journal of the Americal Academy of Child and Adolescent Psychiatry, 22, 559-64.

Stevenson K., Sensky T. and Petty R. (1991). "Glycaemic control in adolescents with type I diabetes and parental expressed emotion". Psychotherapy and Psychosomatics, 55, 170-175.

Stewart S.M., Vavy R., Waller D.A. (1987). "Mental and motor development correlates in patients with end stage biliary atresia awaiting liver transplantation". Journal of Pediatrics, 79, 882-888.

Stewart S.M., Vavy R., Waller D.A. (1989). "Mental and motor development, social competence and growth one year after successful paediatric liver transplantation". Journal of Pediatrics, 114, 574-581.

Stewart S.M., Kennard B.D., DeBolt A., Petrik K., Waller D.A. and Andrews W.S. (1993). "Adaptation of

siblings of children awaiting liver transplantation". Children's Health Care, 22, 205-215.

Stubbe D.E., Zahner G.E.P., Goldstein M.J. and Leckman J.F. (1993). "Diagnostic specificity of a brief measure of expressed emotion: A community study of children". Journal of Child Psychology and Psychiatry, 34, 139-154.

Stuber M.L., Nader K., Yasuda P., Pynoos R.S. and Cohen S. (1991). "Stress responses after pediatric bone marrow transplantation: preliminary results of a prospective longitudinal study". Journal of the American Academy of Child and Adolescent Psychiatry, 30, 952-957.

Stuber M.L. (1993a). "Psychological care of adolescents undergoing transplantation". In: ER McNarney, RE Kreipe, DP Orr et al, editors. Texbook of Adolescent Medicine. Philadelphia, PA, WB Saunders;1993:1138-1142.

Stuber M.L. (1993b). "Psychiatric aspects of organ transplantation in children and adolescents". Psychosomatics, 34, 379-387.

Stuber M.L. and Nader K.O. (1995). "Psychiatric sequelae in adolescent bone marrow transplantation survivors". Journal of Psychotherapy Practice and Research, 4, 30-42.

Surman O.S. and Purtilo R. (!992). "Reevaluation of organ transplantation criteria: allocation of scarce resources to borderline candidates". Psychosomatics, 33, 202-212.

Swanson M., Palmeri D. Vossler E.D., Bartus S.A., Hull D. and Schweizer R.T. (1991). "Noncompliance in organ transplant recipients". Pharmacotherapy, 11, 173S-174S.

Szmukler G.I., Eisler I., Russell G.F.M. and Dare C. (1985). "Anorexia nervosa, parental "expressed emotion" and dropping out of treatment". British Journal of Psychiatry, 147, 265-271.

Szmukler G.I., Berkowitz R., Eisler I., Leff J. and Dary C. (1987). "Expressed emotion in individual and family settings: a comparative study" British Journal of Psychiatry, 151, 174-178.

Taggart D.P. and Dark J.H. (1991). "Heart transplantation in children. Short term results are encouraging". British Medical Journal, 302, 1035-1036.

Tanner J.M. and Whitehouse R.H. (1986). "Growth and development: a book of reference charts" Tanner-Whitehouse Standards. Herts: Castlemead Publications.

Tarrier N., Barrowclough C., Vaughn C.E., Bamrah J.S., Porcedu K., Watts S., and Freeman H. (1988). "The community management of schizophrenia: a controlled trial of a behavioral intervention with families to reduce relapse". British Journal of Psychiatry, 153,532-542.

Tebbi C.K., Cummings K.M., Zevon M.A., Smith L. Richards M., and Mallon J. (1986). Compliance of pediatric and adolescent cancer patients. Cancer, 58, 1179-1184.

Thompson J. ."Compliance". In: R Fitzpatrick, J Hinton, S Newman, G Scambler, and J Thompson, editors.The Experience of Illness. Tavistock Publications, London & New York; 1984:109-132.

Tse A.M., Perz-Woods R.C. and Opie N.D. (1987). "Children's admission to the intense care unit: parents' attitudes and expectations of outcome". Children's Health Care, 16, 68-75.

Utens E.M. Verhulst F.C., Meijboom F.J., Duivenvoorden H.J., Erdman R.A., Bos E. et al (1993). "Behavioural and emotional problems in children and adolescents with congenital heart disease". Psychological Medicine, 23, 415-424..

Uzark K. and Crowley D. (1989). "Family stresses after pediatric heart transplantation". Progress in Cardiac

Nursing, 4, 23-27.

Uzark K.C., Sauer S.N., Lawrence K.S., Miller J., Addonizio L. and Crowley D.C. (1992). "The psychosocial impact of pediatric heart transplantation". Journal of Heart Lung Transplantation, 11, 1160-1167.

Van der Wal R., Nims J. and Davies B. (1988). " Bone marrow transplantation in children: Nursing management of late effects". Cancer Nursing, 11, 132-43.

Van Furth E.F.; Van Strien D.C.; Martina L.; Van Son M. et al (1996). "Expressed emotion and the prediction of outcome in adolescent eating disorders". International Journal of Eating Disorders, 20:19-31.

Vaughn C.E. (1989). "Annotation: Expressed emotion in family relationships". Journal of Child Psychology and Psychiatry, 30, 13-22.

Vaughn C.E. and Leff J. (1976a). "The influence of family and social factors on the course of psychiatric illness: A comparison of schizophrenic and depressed neurotic patients". British Journal of Psychiatry, 129, 125-137.

Vaughn C.E. and Leff J. (1976b). "The measurement of expressed emotion in the families of psychiatric patients". British Journal of Social and Clinical Psychology, 15, 157-165.

Velasco de Parra M.L. (1982). "Changes in family structure after a renal transplant". Family Process, 21, 195-202.

Vostanis P., Nicholls J. and Harrington R. (1994). "Maternal expressed emotion in conduct and emotional disorders of childhood". Journal of Child Psychology and Psychiatry, 35, 365-376.

Vostanis P. and Nicholls J. (1995). "Nine-months changes of maternal expressed emotion in conduct and emotional disorders of childhood: a follow-up study". Journal of Child Psychology and Psychiatry, 36, 833-846.

Wallander J., Varni J., Babanis L., Banis H. and Wilcox K. (1989). "Family resources as resistance factors for psychological maladjustment in chronically ill and handicapped children". Journal of Paediatric Psychology, 14, 157-173.

Wamboldt F.S., Wamboldt M.Z., Gavin L.A., Roesler T.A. and Brugman S.M. (1995). "Parental expressed emotion and treatment outcome in adolescents hospitalized for severe, chronic asthma". Journal of Psychosomatic Research, 39, 995-1005.

Weichler N.K. (1993). "Caretakers' informational needs after their children's renal or liver transplant". ANNA Journal, 20, 135-140.

Wells R.D. and Schwebel A.I. (1987). "Chronically ill children and their mothers: predictors of resilience and vulnerability to hospitalisation and surgical stress". Developmental and Behavioural Paediatrics, 8, 83-89.

White A. (1994). "Parental concerns following a child's discharge from a bone marrow transplant unit". Journal of Pediatric Oncology Nursing, 11, 93-101.

Whitehead B. (1993). "Heart-lung transplantation in patients with cystic fibrosis". Paediatric Respiratory Medicine, 2, 16-18.

Whitehead B., Helms P., Goodwin M., Lask B., Serrano-Ikkos E., Scott J.P. et al (1991). "Heart-lung transplantation for cystic fibrosis : 1. Assessment". Archives of Disease In Childhood, 66, 1018-1021.

Whitehead B., Rees P., Sorensen K., Bull C., Higenbottam T.W., Wallwork J. et al (1994). "Incidence of

obliterative bronchiolitis after heart-lung transplantation in children". Journal of Heart Lung Transplantation, 13, 903-908.

Whitehead B. and De Leval M. (1994). "Paediatric lung transplantation: the agony and the ecstasy". Thorax, 49, 437-439.

Whitehead B., Rees P.G., Sorensen K., Bull C., Fabre J., De Leval M. and Elliot M. (1995). "Results of heart-lung transplantation in children with cystic fibrosis". European Journal of Cardio-thoracic Surgery, 9, 1-6.

WHO (1978). The ICD 9 classification of mental and behavioural disorders. World Health Organisation, Geneva.

Wiley F.M., Lindamood M.M. and Pfefferbaum-Levine B. (1984). "Donor-patient relationship in pediatric bone marrow transplantation" Journal of the Association of Pediatric Oncology Nurses, 1, 8-14.

Wilson J., Fosson A., Kanga J.F., and D'Angelo S.L. (1996). "Homeostatic interactions: a longitudinal study of biological, psychosocial and family variables in children with cystic fibrosis. Journal of Family Therapy,18: 123-139.

Windsorova D., Stewart S.M., Lovitt R., Waller D.A., Andrews W.S. (1991). "Emotional adaptation in children after liver transplantation". Journal of Pediatrics, 199, 880-887.

Wolman C., Resnick M.D., Harris L. J. and Blum R. W. (1994). "Emotional well-being among adolescents with and without chronic conditions". Journal of Adolescent-Health, 15, 199-204.

Wood B.L., Watkins J.B., Boyle J.T., Nogueira J. et al (1989). " The psychosomatic family model: an empirical and theoretical analysis". Family Process, 28, 399-417.

Wood B.L. (1994). "One articulation of the structural family therapy model: a biobehavioural family model of chronic illness in children". Journal of Family Therapy, 16, 53-72.

Wray J., Radley-Smith R. and Yacoub M. (1992). "Effect of cardiac or heart-lung transplantation on the quality of life of the paediatric patient". Quality of Life Research, 1, 41-46.

Wray J., Pot-Mees C., Zeitlin H., Radley-Smith R. and Yacoub M. (1994). "Cognitive function and behavioural status in paediatric heart and heart-lung transplant recipients: the Harefield experience". British Medical Journal, 309, 837-841.

Wynne L. (1984). "The epigenesis of relational systems: a model for understanding family development". Family Process, 23, 297-318.

Wysocki T. (!993). "Associations among teen-parent relationships, metabolic control, and adjustment to diabetes in adolescents". Journal of Pediatric Psychology, 18, 441-452.

Zamberlan K.E. (1992). "Quality of life in school-age children following liver transplantation". Maternal-Child Nursing Journal, 20, 167-229.

Zarinsky I. (1975). " Psychological problems of kidney transplanted adolescents" Adolescence, 10, 101-109.

Zitelli B.J., Malatack J.J., Gartner J.C., Urbach A.H., Williams L., Miller J.W. et al (1986). "Evaluation of the pediatric patient for liver transplantation". Pediatrics, 78, 559-565.

Zitelli B.J., Miller J.W., Gartner J.C., Malatack J.J., Urbach A.H., Belle S.H. et al (1988). "Changes in life-style after liver transplantation". Pediatrics, 82, 173-180.

APPENDIX 1

CAMBERWELL FAMILY INTERVIEW SHEDULE

Notes to the Interviewer

1. Some parts of these interviews may be perceived as painful and threatening by some parents. It is important that the interviewer is respectful, sympathetic and uncritical with the parents at all times.

2. Do not allow one of the parents to take over. Make sure you get satisfactory replies from both parents.

3. It is important to distinguish whether family/marital difficulties are longstanding or whether the child's current admission and the associated changes in parental roles trigger them.

4. During the process of the interview some parents may become aware of disturbing feelings. If that occurs, it is imperative to ask a member of the psychosocial team not involved with the research, to help parents to ventilate such feelings. Under no circumstances should parents leave the hospital upset.

5. If you think that a specific family needs help from health professionals, please <u>DO NOT FORGET TO DISCUSS</u> your concerns with the psychiatrist, psychologist or liaison nurse attached to the unit.

THE CONTENT OF THE FOLLOW UP INTERVIEW WAS THE SAME AS IN THE INITIAL INTERVIEW. HOWEVER, SOME NEW QUESTIONS WERE ADDED.AT THE FOLLOW-UP SEE PAGES

CAMBERWELL FAMILY INTERVIEW SCHEDULE

MODIFIED VERSION FOR PARENTS OF CHILDREN WITH

CARDIO-RESPIRATORY DISEASES

(Serrano-Ikkos E. 1990) Adapted from Camberwell Family Interview
Schedule: Abbreviated version (Vaughn and Leff 1976).

RESEARCH NUMBER: [ ] [ ] [ ]

AGE: [ ] [ ] YEARS [ ] [ ] MONTHS

SEX: [ ]   Masculine 1; Feminine 2

The interview has taken place:

[ ] [ ] weeks before surgery

[ ] [ ] months before surgery

The interview was attended by: [ ]

1. Mother alone   3. Both parents

2. Father alone   4. Other (specify)

Interviewer:

Date:

Audiotape: Yes   No

Videotape: Yes   No

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DEMOGRAPHIC DATA

1. **Are you working?**   **Briefly, what sort of work do you do?**

   Full time: What sort of hours?   What about weekends?

   Part time: What sort of hours?   What about weekends?

   From home: What sort of hours?   What about weekends?

   Mother's Occupation: [ ]

   Housewife   1

   Working full time   2

   Working part time   3

   Not Working   4

   Current Work: [ ][ ]

Socio-economic group I	01	Socio-economic group V	06
Socio-economic group II	02	Student, retired, other (specify)	07
Socio-economic group III	03	Unemployed	08
Socio-economic group III (manual)	04	Not applicable	88
Socio-economic group IV	05	Missing	99

   Father's Occupation: (as above)   [ ]

   Current Work: (as above)   [ ][ ]

   # Ethnic Background-Culture: Mother [ ][ ]

       Father [ ][ ]

United Kingdom & Ireland	01	Africa	06
North & Centre Europe	02	Asia	07
South Europe	03	Australia	08
North America & Canada	04	Not applicable	88
South America	05	Missing	99

2. **FAMILY STRUCTURE**

   Who lives in the household with you?   [ ]

   The child is living with:

Both biological parents	1	Foster/adoptive parents	4
Single mother/single father	2	Other (specify)	5

First generation of immigrants will be scored by Country of Origin

Biological parent and step-parent   3   Missing   9

Family composition:

No. of children living in the household   [ ]

No. of children from the current relationship.   [ ]

No. of children with a chronic illness living in the household   [ ]

How long have you been married or living together   [ ][ ] yrs [ ][ ] mths

3.   **If the child is not living with one/both biological parents:**

How much contact does (patient's name) have with his/her father/mother?   [ ]

Regular contact   1   No contact at all   3

Irregular contact   2   Not applicable   4

If there is contact, this is every   [ ] [ ] wks, [ ] [ ] mnths

THE CHILD AND HIS/HER ILLNESS

4.   **(EE- FMSS) DESCRIPTION OF THE CHILD**

Before we start talking about (patient's name) and the effect of the illness on him/her "I'd like your thoughts about (patient's name).   In your own words, and without me interrupting you with any questions or comments, I'd like each one of you to speak for 2 minutes telling me what kind of person (patient's name) is and how the two of you get along together."

BOTH PARENTS should give a description of the child in their own words [Ψ].

MOTHER:

FATHER:

DURING THE LAST 3 MONTHS

PATIENT'S ROUTINES / INDEPENDENCE / EXPECTATIONS

5.   **I would like to get a picture of how (patient's name) usually spends a day [1].**

What time would ....

-      get up?

[Ψ] IF ONE OF THE PARENTS HAS DIFFICULTIES with this task, the interviewer can help by saying: "Often parents experience their own children differently. Parents' expectations about the child and their relationship with the child, are quite dissimilar." "I would like you to tell me what kind of person (patient's name) is and how the two of you get along together."

[1] **NOTE:** Obtain Details of How the Patient's Routine Relates to Contact With Other Family Members

-      have breakfast?
-      go to school?
-      return from school/eat supper?
-      spend the evening?
-      go to bed?
-      and who would be there at the time?

PROBE:

How Much Time the Child Spends with Mother/Father/Siblings.

MOTHER:

FATHER:

6.   **I would like you to tell me what things (patient's name) does by himself/herself and what things you do for him/her.**

- How much help does (patient's name) need to get ready in the morning?

- How often do you help him/her get dressed/washed?

- For what sort of things would (patient's name) ask for help?

PROBE :

Whether given the Child's age and Physical Condition, it is Necessary for the Parents to do those things for the Child:

- Regarding self-care and the things you have just mentioned: Are you satisfied with the number of things which (patient's name) does by him/herself?

- Bearing in mind (patient's name) 's physical limitation, is there anything which he/she could do differently? Could he/she be more independent?

IF POSITIVE:

How?

When this sort of thing happens, how do you react?

If necessary. How do you feel about that?

MOTHER:

FATHER:

7.   **Often children are expected to help in various jobs that have to be done in the family/ in the house (like shopping for things, laying the table......etc.). Taking into account that (patient's name) is .... years old, what sort of help do you expect from him/her?**

- Does he/she ever do it?

- **IF NOT**: what happens then?

- How do you react to that?

- How do you feel about it?

IF NO EXPECTATIONS:

- Is he/she tidy?

- Who clears his/her room?

- And what about..........(name the siblings).

PROBE:

Different expectations about the ill child & siblings?

MOTHER:

FATHER:

DISCIPLINE & RELATIONSHIPS

8.   **All parents have rules about how their children should behave and what they should and should not do:**

- How often does (patient's name) break the rules?

- What sort of things would he/she do which make you cross?

- Or, in what way does he/she get on your nerves?

PROBE:

- How often would that happen (once a week/month)? - How do you react?

- How do you tell him/her off?

MOTHER:

FATHER:

And what about (name the siblings)?

- How do you tell them off?

- How do you treat (patient's name) and his/her siblings differently?

- With whom are you more strict? If with siblings: why is so?

PROBE:

- Do parents give more privileges to ill child than to the siblings [τ]

- Are parents softer with the patient than with sibling?

[τ] The interviewer should be aware of different treatment due to sex and age differences between the patient and the siblings

MOTHER:

FATHER:

Parental account on STRICTNESS AT HOME   [ ]

Equal treatment   1   Not applicable: no siblings   8

More strict with siblings 2   Missing   9

More strict with ill child 3

9.   **Can you get close to (patient's name)**

       MOTHER:

       FATHER:

10.   **Is he/she easy to get along with? How does the child get along with siblings?**

       MOTHER:

       FATHER:

11.   **In what way would you like (patient's name) to be different?**

       MOTHER:

       FATHER:

12.   **Are you satisfied with the amount of affection/interest (patient's name) shows you?**

       MOTHER:

       FATHER:

13.   **What sort of things you do enjoy doing with (patient's name)?**

       MOTHER:

       FATHER:

       PROBE:

       How much each parent enjoys, appreciates and value the child's company

THE EFFECT OF THE ILLNESS ON THE CHILD

14.   **I would like to check with you some events related to your child illness:**

- How old was (patient's name) when first diagnosed   [   ]   [   ] yrs   [   ]   [   ] mth

- Has he/she undergone any previous operation?   [   ]   [   ] **number of operations**

- Has she/he had any previous admissions during the last year?

IF POSITIVE:

- When? How many?   [   ]   [   ] **previous admissions**

- How has the illness shown itself during the last months?

PROBE:

- Severity of symptoms (cyanosis, breathlessness, fatigue)

- Child's physical disability

- Parents' reaction to the child's symptomatology

MOTHER:

FATHER:

Parental account of CHILD'S SEVERITY OF SYMPTOMS   [   ]

- No symptoms at all	1	- Symptoms on rest	4
- Occasional breathlessness, fatigue, tiredness	2	- Not applicable	8
- Frequent breathlessness, fatigue, tiredness	3	- Missing	9

Parental account of the CHILD'S COLOUR   [   ]

- Normal colour	1	- Not applicable	8
- Light cyanosis	2	- Missing	9
- Severe cyanosis	3		

Parental account of DEGREE of CYANOSIS   [   ]

- Local cyanosis	1	- Not applicable	8
- General cyanosis	2	- Missing	9

Parental account of CHILD'S PHYSICAL DISABILITY   [   ]

- No limitations: the child does not have any physical restrictions at all   1

- Minor limitations: normal physical activity, but occasionally gets breathless

  and has to stop for a while   2

- Moderate limitations: minor restrictions, e.g. avoids walking fast, staircases   3

- Major limitations: major restrictions, e.g. difficulties walking flat	,	4
- Severe limitations: needs a wheelchair to go out		5
- Invalid: uses a wheelchair to move outside/inside the house		6
- Not applicable: the child is a baby		8
- Missing		9

15. **Now, I would like you to tell me about his/her school life. Which sort of school is the child attending?**

The child is attending   [ ]

- Normal day school	1	- Other (specify)	4
- Special day school	2	- Not applicable	5
- Home tuition	3	- Missing	9

IF attending school: Full-Time [1]   Part-Time [2]

How much school has the child missed in the last 3 months because of: admissions to hospital, appointments, feeling unwell etc.

[ ] [ ] Wks missed

How is (patient's name) doing academically?

Are you satisfied about his/her performance at school?

IF NECESSARY:

- Academically, could he/she do better?

- What could he/she do differently?

IF POSITIVE:

- How do/did you react to that?

- How do you feel about it?

MOTHER:

FATHER:

16. **Can you tell me about his/her social life?**

- How often would he/she meet friends outside school hours, or bring friends home, or go for a ride?

- How does (patient's name) get along with other people outside the family?

- Do people like him/her? Is he/she popular?

- How do you feel about his/her social life?

- How would you like his/her social life to be different?

PROBE:

- Has he/she ever spent a night in his/her friend's house?

- Are the parents too anxious to let him/her be away from home

- If (patient's name) was not ill, would his/her social life be any different?

IF POSITIVE:

- How would it be?

MOTHER:

FATHER:

Parental account on CHILD'S SOCIAL LIFE   [ ] [ ]

The child meets friends outside school hours:

- More than once/week	1	- Less than once in 3 months	6
- Once a week	2	- Never	7
- Between once a wk and once fortnight	3	- Not applicable	88
- Between once/week and once/month	4	- Missing	99
- Between once/month and once/3 months	5		

How many close friends the child has   [ ] [ ]

Now I would like to ask some questions about how (patient's name) may have been affected by this illness. Of course, many will not apply, but I should like to run through these quickly and perhaps you can tell me whether or not (patient's name) has been like this **particularly in the last 3 months.**

17.   **Irritability: Children with this condition may feel irritable. I mean snappy...raising their voice...getting impatient...quarrelling etc.**

-     How often has (patient's name) been like this with: mother, father, siblings?. Can you describe what happened?

PROBE:

What sorts of things make him/her irritable with (name family members)?

- Does this usually happen at any particular time of the day?

- Who else would be there?

- How would (named all people present) react?

IF DENIAL:

- Has he/she ever made you cross?

- Can you remember a time when he/she lost his/her temper in the past year?

If yes: why do you think this has changed?

MOTHER:

FATHER:

18.   **How has his/her sleep been like? (in the past 3 months)**

PROBE:

Difficulty getting to sleep? Nightmares? Waking very early?

IF POSITIVE:

- How much of a problem is that for you?

- How do you deal with it? How do you react? What do you do?

PROBE:

- Does the child sleep in own or parents bed/bedroom? How often the child sleeps with either of the parents?

- Taking into account the child's physical condition, is it necessary for the child to sleep with parents?

MOTHER:

FATHER:

19.   **I would like you to tell me about his/her eating habits and whether this is a problem for you.** - What has his/her appetite been like?

PROBE:

Do parents cook different meals for (patient's name)?

IF POSITIVE: Taken into account the child's physical condition, is that necessary?

MOTHER:

FATHER:

20.   **Has he/she been worrying about his/her health?**

- About coming to hospital?

- And about his/her physical appearance?

PROBE: what makes you think he/she is worrying about it?

- How much does he/she talk about it?

- How often? To whom?

- How do you react/ deal with it?

MOTHER:

FATHER:

21.   **Has he/she been depressed/miserable?**

How do you react?

PROBE:

- What makes you think that he/she is depressed/miserable?

- Has he/she been worrying about dying?

MOTHER:

FATHER:

COMMUNICATION ABOUT THE ILLNESS/SURGERY

22.   **I would like you to tell me about (patient's name)'s knowledge of his/her medical condition:**

- In your view, what is (patient's name) understanding of his/her physical condition?. What does (patient's name) think is wrong in his/her body?

- How long ago did you inform the child about the need for this operation?

- How much does (patient's name) know about what going to happen to him/her during this admission?

-     How have you prepared (patient's name) for the operation? Who has taken the responsibility for preparing the child?

- What have you said to (patient's name) about the surgery and the recovery period?

-     (if applicable) And what about his/her prognosis and life expectancy? Have you discussed this issue at all with him/her?

-     Has (patient's name) ever asked about the possibility of things not going as planned or about the possibility of him/her dying?

IF POSITIVE:

-     These sorts of questions are very difficult for parents to handle. What happened when (child's name) asked you (repeat parents' reply to the above question)? How did you manage that?

- What about his/her sibling? Are they worried about (patient's name) dying?

PROBE:

- How much/often have the parents discussed these issues at home with patient's or siblings?.

- What about with grandparents, extended family, or between themselves?

- Do parents know how their partner is feeling about surgery/operation?

MOTHER:

FATHER:

Parents account of CHILD'S PREPARATION FOR SURGERY   [ ]

Parents share equal responsibility	1	Share responsibility but not equally	4
Mother alone	2	Not applicable: there is only one parent	8
Father alone	3	Missing	9

FAMILY COMMUNICATION ABOUT SEVERITY OF ILLNESS: [ ]

Open Communication [1]	1
Intermediate Communication [2]	2
Poor Communication [3]	3

MANAGEMENT OF CHILD'S MEDICAL TREATMENT

23.   **Now, I'd like you to tell me how (patient's name) is accepting the treatment.**

As I understand it, during the last 3 months (patient's name)'s medical treatment consists of. (the interviewer mentions all different aspects of treatment).   In some families, one of the parents takes the responsibility for providing the treatment. In other families, both parents take responsibility for the daily administration of the treatment.

-        I am wondering who does what in your family. Who is responsible for providing (patient's name) with the different aspects of treatment?

MOTHER:

FATHER:

---

[1] If the parents have discussed openly the child's illness and the possibility of the child's death between themselves and the children (if age-appropriate) and children feel free to ask questions which parents answer openly and appropriately. Both parents are involved in the preparation of the child for surgery.

[2] Parents have some difficulties talking between themselves or with the children about illness/death. Children's questions are not openly answered. Severity of illness may be acknowledged but possibility of death is denied.

[3] Parents have major difficulties talking between themselves about illness/death. Parents have not informed the ill child or the siblings about the severity of illness/possibility of death. The child has not been prepared for transplantation/hospital admission.

PROBE:

- Each parent's role regarding the administration of treatment

- How involved each one of the parents is with the administration of each component of the treatment ?

- Parents' beliefs about the treatment

If the child is 8 years or older:

- Does (patient's name) need to be reminded / take any responsibility for:

a) taking the medication?   b) doing the physiotherapy?

c) having the nebulizer?   d) following the diet?

PROBE:

- How often do the parents have difficulties administering the treatment (patient refuses or is not happy about it)?

- What do you do when (patient's name) behaves like that?

MOTHER:

FATHER:

Parents account of DIFFICULTIES ADMINISTERING THE TREATMENT

Medication	Mother [ ][ ]	Father [ ][ ]
Physiotherapy	Mother [ ][ ]	Father [ ][ ]
Nebulizer	Mother [ ][ ]	Father [ ][ ]
Diet	Mother [ ][ ]	Father [ ][ ]

- Not at all	01	- More than daily	06
- Once a month or less	02	- Parent not involved	07
- Between once/week and once/month	03	- N/A	08
- Between 2-4 times a week	04	- Missing	99
- Between 5-7 times a week	05		

EFFECT OF THE ILLNESS ON PARENTS & SIBLINGS

You have given me a very good picture of (patient's name) and how he/she is being affected by his/her medical condition. Now, I would like you to tell me about yourselves and the other children.

24. **Briefly I would like you to tell me about the effect that the illness is having on you, as individuals.**

- How much time do you have for yourselves?. How often do you do things just for yourself, for your own enjoyment?

IF NECESSARY:

I mean, forget your responsibilities and duties for a while and .....how often do you sit and listen to music ....or you meet a friend for lunch/drink ... or you go to the gym...etc?

IF NEGATIVE:

- If (patient's name) was healthy, would you have time for your hobbies, social life etc.?
- How would you like things to be different?

PROBE:

- Whether parents difficulties leaving the children with other carers, such as grandparents or babysitter. are understandable given the child's physical condition or whether parents do not leave the children with others because of separation-anxiety/overprotection etc.

MOTHER:

FATHER:

25. **What about the two of you as a couple:**

- How much time do you have for each other?
- Which sort of things do you enjoy doing together?
- How often the two of you go out for a meal/to the cinema/socialise with friends etc?
- How do you imagine that your social life would be if (patient's name) were healthy?

IF NEGATIVE:

- How would you like you social life to be different
- What is stopping you from (the interviewer mentions the things they have just said above)?

MOTHER:

FATHER:

26. **Most husbands and wives have quarrels from time to time:**

-What sorts of things do you complain/nag/grumble about to each other?
- How often has this occurred in the last week ... month ... 3 months?

IF POSITIVE:

- Would you like it to be any different? - How?

PROBE:

- Disagreement about how to handle the patient and/or the other children.

- Do the children play off one against the other?

- How much do they support each other in front of the children?

MOTHER:

FATHER:

27.   **When there is an ill child who needs so much parental attention, it is sometimes difficult for the parents to have enough time for the other children:**

- In which way have (siblings names) been affected by (patient's name)'s illness?

- How much time have you got for (patient's name)'s sibling/s?.

- Do/does (the interviewer mentioned the sibling/s by name) understand why you give so much of your time to (patient's name) or is this a conflictual family issue? How do you react to that?

- Are you satisfied with the time available for them?

- Are (sibling's names) missing out appropriate activities for their age?

IF POSITIVE:

- How do you deal with that?

- How do you feel about it?

-    How would you like it to be different?

-

PROBE:

- Siblings emotional deprivation

- Social life (friends, holidays etc)

- School life (academic performance, attendance)

MOTHER:

FATHER:

28.   **We have nearly finished this interview. I would like you to tell me a bit about:**

- What sort of things do you enjoy doing together as a family?

- How do you decide what to do/not to do as a family, and

- What happens when (sibling's name) wants to do things that (patient's name) cannot do because of his medical condition? How do you solve the problem?

- If (patient's name) was healthy, would your family life be any different?

PROBE:

Holidays, activities together etc.

IF NEGATIVE:

- Are you satisfied with your current family style of life?

- Does it bother you?

- How would you like your family life to be different?

MOTHER:

FATHER:

RATINGS-SUMMARY

MOTHER'S SCORES (EE)

Overinvolvement:

Warmth:

Critical Comments:

Positive Remarks   :

Hostility:

FATHER'S SCORES (EE)

Overinvolvement:

Warmth:

Critical Comments:

Positive Remarks:

Hostility:

FAMILY ADJUSTMENT

Schneiderman's Ratings:

1- Well adjusted

2- Moderately adjusted

3- Poorly adjusted

Longstanding family difficulties:

- Roles

- Boundaries

- Communication

- Problem solving

- Affective responsiveness/involvement

- Marital functioning

COMMENTS REGARDING MANAGEMENT OF TREATMENT

RECOMMENDATIONS

If this family needs to be referred to the Department of Family & Child Psychiatry or similar, please state reasons for referral:

AT THE FOLLOW-UP AND UNDER SPECIFIC SECTIONS, FEW QUESTIONS WERE ADDED TO
THE CAMBERWELL FAMILY INTERVIEW (See beneath).

The interview has taken place: [ ] [ ] mths after surgery

[ ] [ ] mths from initial

assessment

DEMOGRAPHIC DATA

A.   **Have you changed your job/occupation during the last 12 months?**

IF POSITIVE PROBE:

Whether changes in occupation were related to changes in child health.

THE EFFECT OF THE ILLNESS ON THE CHILD

B.   **I would like to check some events related to your child's illness.**

- Has he/she had any other operation during the last 12 months?

[ ][ ] **Number of operations**

- Has he/ she had any other admissions than the expected ones?

-       [ ][ ] **Number of unexpected**

IF POSITIVE : When?   **admissions**

- How has the illness shown itself during the last months?

PROBE:

- Severity of symptoms since conventional surgery/transplantation

- Child's physical disability

- Complications (e.g. transplanted children: rejection, infections)

- Parents' reaction to the child's symptomatology

MOTHER

FATHER

C.   Has (patients name) school performance changed since he/she received surgery/transplantation?

IF POSITIVE:

- In what way?

COMMUNICATION ABOUT THE ILLNESS

D.   **I would like you to tell me about (patient's name)'s knowledge of his/her medical condition.**

- In your view, what is (patient's name)'s understanding of his/her physical condition?

- What does (patient's name) think is wrong with his/her body?

- (If applicable) Does (patient's name) know that he/she needs another operation in the near future?

- How often do you talk about the illness or about the previous operation with (patient's name) or other family members?

- (For parents of transplanted children only.) What happens when (patient's name) is coming for a biopsy, do you talk about what would happen if the results are not as expected?

- (For parents of transplanted children only.) As you know, the transplant is not curative, how often do you discuss this with (patient's name)?

PROBE (Parents of transplanted children only)

Whether life expectancy and the patient's short life span has been discussed with the patient, siblings, extended family, and between parents.

MOTHER

FATHER

MANAGEMENT OF CHILD'S MEDICAL TREATMENT

E. **Now I would like you to tell me how (patient's name) is accepting the treatment. As I understand it, during the last 3 months (patient's name)'s medical treatment consists of ... (the interviewer mentions all different aspects of treatment.** There is a variation in how families organise themselves. In some families, one of the parents takes full responsibility for Providing the treatment; however in other families that responsibility is equally or partially shared between the parents:

- I am wondering who does what in your family regarding the administration of the treatment?

- Who is responsible for providing (patient's name) with his/her treatment?

PROBE:

Each parent role regarding the administration of the treatment.

- How involved are each one of the parents with the administration of each one of the treatment-components?

If the child is 8 years or older:

- Does (child's name) need to be reminded/take any responsibility for:

a/ taking medication?

b/ completing the medical diary?

c/ doing the lung function test?

PROBE:

- How often do the parents have difficulties administering the treatment (child's refusal)?

- (Older children) How much parents allow the patient to take responsibility for his/her treatment?

MOTHER

FATHER

Who is usually responsible for administering:

Medication   [ ][ ]

Completing the medical diary   [ ][ ]

Lung function test   [ ][ ]

Mainly the mother	01	Mainly the patient (without reminder)	05
Mainly the father	02	Equally: patient and parents	06
Both parents jointly	03	N/A	88
Mainly the patient (with a reminder)	04	Missing	99

(For parents of transplanted patients only)

- During the last 12 months, (patient's name) has between 14 and 18 routine admissions for biopsies, check ups, etc. How does he/she respond to all these admissions? Is she happy about it or does he/she get upset?

MOTHER

FATHER

The child gets upset about admissions/check-ups   [ ]

- Always	1	- Never	5
- Often	2	- NA	8

| - Sometimes | 3 | - Missing | 9 |
| - Rarely | 4 | | |

SIDE EFFECTS OF TREATMENT

- Is (patient's name) bothered about the scars of surgery?

- What does he/she say when other kids/people ask about it?

- What about you, are you bothered about it?

- (For parents of transplanted children only)

   Does (child's name) get upset about the side effects of the medication such as getting hairy/puffy-face, etc?

- What about you, are you concerned about it?

MOTHER

FATHER

EFFECT OF THE ILLNESS ON PARENTS AND SIBLINGS

F. (Patient's name) is now more independent and less demanding of your time [1]. These changes are due to the fact that he/she is getting older but also to the improvement in health following surgery.

   - How easy is it for you to adjust to these changes? (Patient's name) needed you before but now he/she is more independent. How are you adjusting to this major change?

IF NECESSARY

- Do you ever get upset about it?

- How do you feel about not being needed to the same extent as before?

MOTHER

FATHER

- Has the amount of time you have for the other kids changed since the operation?.

---

[1] The interviewer goes on to give specific examples such as ' before F. was confined to bed,. now F. attends school, plays football ...etc.'.

APPENDIX 2

THE EXPRESSED EMOTION PARTNER INTERVIEW SCHEDULE (EEPIS)

(Serrano-Ikkos E, 1990)

AT INITIAL ASSESSMENT

I find it helpful to see both of you together. It is also important for me to talk to you separately for 5-10 minutes, to get a full picture, just to get your own impressions, your different perspectives/points of view as individuals.

1. **We have talked a lot about (patient's name) illness, and its effect on the family.**
   - Is there something which you would like to add or to ask me?

2. **From you own point of view, what has been the most disturbing aspect of (patient's name)'s trouble?**

3. **What is helping you the most to cope with all this?**

4. **What is the most effective way that (partner's name) has found to support you?**
   - What does he/she do which helps you to cope with this difficult situation?

5. **If something is worrying you about your work/your children. or if you are feeling unhappy about something, does (partner's name) help you to deal with that?**

   IF POSITIVE:
   - How?
   -

   IF NEGATIVE:
   - In general, how do you get along with (partner's name)?
   - What sort of things do you enjoy doing together?
   - How would you like things to be different?

6. **In general, would you like to lean more or less on him/her or is it about right?**

   PROBE:
   Support/help with practical issues : taking the child to the doctor, looking after the other kids, staying in hospital.

- Emotional support: How much?

- Can they lean on/comfort each other?

- Can they talk frankly with each other about their fears/anxieties of the operation/possible loss of the child?

- Do you feel that the other is emotionally available?

- Do you have other people around you to whom you can talk frankly?

Does he/she lean   **Wife**   **[ ]**

                                                        Husband   **[ ]**

- More on partner than on others   1

- More on others than on partner   2

- Equally   3

7.   **Has the affection your husband has shown you, changed at all over the last 9 months?**

- What about the interest in you?.... or in the things you do? Does he/she show much interest?

- Are you satisfied with the affection and interest he/she shows in you?

IF NECESSARY:

- In what ways would you like to be different?   OR

- How do you feel about the change?

- Does it bother you much?

Change in Affection shown by spouse: **Wife**   **[ ]**

                                                 Husband   **[ ]**

- Definite increase	1	- Definite decrease	5
- Possible increase	2	- Marked fluctuations	6
- No change	3	- N/A. No partner	8
- Possible decrease	4	- Missing	9

8.   **All couples have rows from time to time, they nag each other, or they have disagreements....**

  - Have the amount of rows, nagging or disagreements with your partner, changed at all over the last 9 months?

IF POSITIVE:

- In what way?

- Does it bother you much?

IF NECESSARY:

- Have you ever considered to ask for help from a professional?

Change in Disagreements: **Wife**   [ ]

                                            Husband   [ ]

- Definite increase	1	- Definite decrease	5
- Possible increase	2	- Marked fluctuations	6
- No change	3	- N/A. No partner	8
- Possible decrease	4	- Missing	9

9.   **To have a child with a physical condition like (child's name) puts a lot of strain on the parents. Some parents feel that their child's illness brings them together, other parents feel that the pressure of illness separates them while a third group of parents feel that the illness doe not have any effect on the way they relate/feel to/about each other. - How do you see yourself in that spectrum? [1]**

The child's illness has   **Wife**   [ ]

                                          Husband   [ ]

Brought them together	1
Separated them	2
Had no effect	3

EE HUSBAND AND WIFE RATINGS

Husband to wife:

Critical comments:

Hostility:

Emotional Overinvolvement:

Warmth:

Positive remarks:

Wife to husband:

Critical comments:

Hostility:

Emotional Overinvolvement:

Warmth:

Positive remarks:

---

[1] Before they leave the interviewing room: This short interview was meant to find out about how couples cope with the operation and the impact that the stress of surgery may have on their relationship. If **after you leave this room**, there is something which bothers you related to what we have been talking about, **PLEASE DO NOT HESITATE TO GET IN TOUCH WITH ME AS SOON AS CONVENIENT.**

THE EXPRESSED EMOTION PARTNER INTERVIEW SCHEDULE (EEPIS)

AT THE FOLLOW-UP

1. **We have talked a lot about (patient's name) and what happened to him/her after the operation. Is there anything else you would like to add or ask me?**

2. **From your own point of view, what was the most difficult aspect while (patient's name) was in hospital having his/her operation?**

   PROBE:

   - How did you cope with the stresses of the operation?

   - Did you talk to people about it?

   - What about your partner? How much have you talked to each other about what was happening?

Communicating with partner/people:		Wife	[ ]
		Husband	[ ]
- Talked mostly with partner	1	- N/A. There is no partner	8
- Talked mostly to other people	2	- Missing	9
- Talked equally to partner and to other people	3		

3. **Did you have enough time together, you and your partner?**

   - What was the best way (partner's name) found to help you during that difficult period?.

   - How would you have liked things to be different?. Is there something that you would have like her/him to do differently?.

   IF NECESSARY:

   - At the time, were you able to talk frankly to him/her about your fears, anxieties and possible loss of the child?.

   - To what extent you felt emotionally supported by (partner's name).

Dissatisfaction with time spent together:		Wife	[ ]
		Husband	[ ]
- Yes, definitely	1	- N/A. No partner	8
- Yes, to some extent	2	- Missing	9
- Not at all	3		

| They would have liked : | | Wife | [ ] |
| | | **Husband** | [ ] |

- More time together   1   - N/A. No partner   8

- Less time together   2   - Missing   9

- About right   3

| Feeling supported by partner: | | **Wife** | [ ] |
| | | **Husband** | [ ] |

- Yes, definitely   1   - N/A. No partner   8

- Yes, to some extent   2   - Missing   9

- No   3

| They would have liked to get: | | **Wife** | [ ] |
| | | **Husband** | [ ] |

- More support   1   - N/A. No partner   8

- Less support   2   - Missing   9

- About right   3

4.   **Couples do differ in how much they need to share feelings / be close or distant. Looking back to the time when (patient's name) had the operation:**

- Did you become closer to your partner?.... or ....

- Did you need to be more distant?.... or ....

- The operation didn't make any difference in this respect?

| During the Child's Admission the Couple became: | | **Wife** | [ ] |
| | | **Husband** | [ ] |

- Much closer   1   - Much distant   5

- Mildly closer   2   - N/A. No partner   8

- No change   3   - Missing   9

- Mildly distant   4

4.1   **- Would you have liked it to be different in any way?.**

IF POSITIVE:

- How?.

- How did you feel about that at the time?.

- How do you feel about it now?.

- **Has the way you feel about your partner changed at all since (patient's name) had his her operation?.**

IF POSITIVE:

- How?. In what way?.

5.   **Has the affection your partner has shown you changed at all over the   last 9 months?**

- What about the interest in you?.... or in the things you do? Does he/she show much interest?

- Are you satisfied with the affection and interest he/she shows in you?

IF NECESSARY:

- In what ways would you like to be different? OR

- How do you feel about the change?

- Does it bother you much?

Change in Affection shown by spouse: **Wife**   [ ]

                                                       Husband   [ ]

- Definite increase	1	- Definite decrease	5
- Possible increase	2	- Marked fluctuations	6
- No change	3	- N/A. No partner	8
- Possible decrease	4	- Missing	9

6.   **All couples have rows from time to time, they nag each other, or they have disagreements....**

- Has the amount of rows, nagging or disagreements with your partner, changed at all over the

         last 9 months?

IF POSITIVE:

- In what way?

- Does it bother you much?

IF NECESSARY:

- Have you ever considered asking for help from a professional?

Change in Disagreements:		Wife	[ ]
		Husband	[ ]

- Definite increase	1	- Definite decrease	5
- Possible increase	2	- Marked fluctuations	6
- No change	3	- N/A. No partner	8
- Possible decrease	4	- Missing	9

EE HUSBAND AND WIFE RATINGS[1]

Husband to wife:

Critical comments:

Hostility:

Emotional Overinvolvement:

Warmth:

Positive remarks:

Wife to husband:

Critical comments:

Hostility:

Emotional Overinvolvement:

Warmth:

Positive remarks:

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[1] This short interview was meant to find out about how couples cope with the operation and the impact that the stress of surgery may have on their relationship. If **after you leave this room**, there is something which bothers you related to what we have been talking about, **PLEASE DO NOT HESITATE TO GET IN TOUCH WITH ME AS SOON AS CONVENIENT.**

APPENDIX 3.1

Distribution of the Expressed Emotion Ratings by Rating Categories

EE Towards the Children	Mothers	Fathers
Overinvolvement:		
None - Little	n=90 (65.2%)	n=77 (70.6%)
Moderate	n=20 (14.5%)	n=18 (16.5%)
High - Mark	n=28 (20.3%)	n=14 (12.8%)
Warmth:		
None - Some	n=26 (18.8%)	n=22 (20.2%)
High	n=112 (81.2%)	n=87 (79.8%)
Hostility:		
None	n=131 (94.9%)	n=108 (99.1%)
Present	n= 7 (5.1%)	n= 1 (.9%)
Positive Remarks:		
None	n=18 (13.0%)	n=23 (21.1%)
1 or 2	n=66 (47.8%)	n=50 (45.9%)
More than 2	n=54 (39.1%)	n=36 (33.0%)
Critical Comments:		
None	n=102 (73.9%)	n=92 (84.4%)
1 or 2	n=27 (19.6%)	n=15 (13.8%)
More than 2	n=9 (6.5%)	n=2 (1.8%)

EE Towards their Partners	Mothers	Fathers
Warmth:		
None - Some	n=58 (45.7%	n=50 (45.5%)
High	n=69 (54.3%)	n=60 (54.5%)
Hostility:		
None	n=123 (96.9%)	n=109 (99.1%)
Present	n= 4 (3.1%)	n= 1 (.9%)
Positive Remarks:		
None	n=98 (77.1%)	n=88 (80.0%)
1 or 2	n=27 (21.3%)	n=22 (20.0%)
More than 2	n=2 (1.6%)	n=0 (0.0%)
Critical Comments:		
None	n=108 (85.0%)	n=103 (93.6%)
1 or 2	n=16 (12.6%)	n=6 (5.5%)
More than 2	n=3 (2.4%)	n=1 (.9%)

APPENDIX 3.2

One-way Analyses of Variance (priori contrast)

Mothers' Overinvolvement (EOI):	$F= 0.93$, $df= 4,133$, $p=0.44$
Mothers' Warmth (W):	$F= 0.63$, $df= 4,133$, $p=0.63$
Mothers' Hostility (H):	$F= 0.27$, $df= 4,133$, $p=0.89$
Mothers' Positive Remarks (PR):	$F= 2.11$, $df= 4,133$, $p=0.08$
Mothers' Critical Comments (CC):	$F= 0.62$, $df= 4,133$, $p=0.64$
Fathers' Overinvolvement (EOI):	$F= 0.30$, $df= 4,104$, $p=0.87$
Fathers' Warmth (W):	$F= 1.47$, $df= 4,104$, $p=0.21$
Fathers' Hostility (H):	$F= 1.37$, $df= 4,104$, $p=0.24$
Fathers' Positive Remarks (PR):	$F= 1.79$, $df= 4,104$, $p=0.13$
Fathers' Critical Comments (CC):	$F= 1.31$, $df= 4,104$, $p=0.26$

Mothers' Warmth (W) /partner:	$F= 0.35$, $df= 4,122$, $p=0.83$
Mothers' Hostility (H) /partner:	$F= 0.96$, $df= 4,122$, $p=0.42$
Mothers' Positive Remarks (PR) /partner:	$F= 1.86$, $df= 4,122$, $p=0.12$
Mothers' Critical Comments (CC)/partner:	$F= 0.61$, $df= 4,122$, p 0.65

Fathers' Warmth (W) /partner:	$F= 1.27$, $df= 4,105$, $p=0.28$
Fathers' Hostility (H) /partner:	$F= 0.72$, $df= 4,105$, $p=0.57$
Fathers' Positive Remarks (PR) /partner:	$F= 1.48$, $df= 4,105$, $p=0.21$
Fathers' Critical Comments (CC) /partner:	$F= 0.54$, $df= 4,105$, $p=0.70$

APPENDIX 4

TWO CASE EXAMPLES DEMONSTRATING DIFFERENCES IN THE FAMILIES' ORGANISATION
AND MEDICAL COURSE

CASE 1

C.W., 9 years old boy who was referred for heart-lung transplantation on 9.10.89. C. was diagnosed as having cystic fibrosis (CF) at the age of 4 months. His progress was steady, but following his 8th birthday he suffered a sudden deterioration becoming quite cyanosed, breathless, and unable to attend school. C. was living with his mother, his stepfather and two sisters from this new marriage, aged 2 years and 6 weeks. C. was adopted by Mr W. and had no contact at all with his natural father, who left home when C. was just under 6 years of age. The break-up of Mrs W. with C.'s natural father was traumatic and involved a lot of physical violence, which C. had witnessed.

C. was attending the local Child and Family Centre prior to his referral to GOSH. As C. became severely ill he also became non co-operative with his treatment, felt threatened, miserable and confused about his physical condition and uncertain future. He was feeling responsible about his mother's separation from his natural father. He was biting his nails excessively and had developed a bilateral eye tic. A child psychiatrist was offering ongoing support for C. and his mother, and was helping them to work through their feelings about the past.

Both parents accompanied C. during his initial admission to assess suitability for transplantation. They presented themselves as very caring and warm towards C. and towards their partner. They did not express any criticism or hostility about the patient, nor about each other. However, Mrs W. was highly overprotective and emotionally overinvolved with C.. Mrs and Mr W. feared that C. would have a short life, therefore, they both tended to respond to his demands on most occasions. Occasionally, this created some difficulties between Mrs W. and her husband, who tried to be more strict with C.. Their ratings on the marital adjustment scale were above average, and the ratings on the psychological functioning were below the cut-off point, which indicates the possible existence of a psychiatric disorder. There was no known history of psychiatric illness in the family, although Mrs W. was said to feel very low at the end of her previous marriage. During the assessment process, Mr W. expressed on various occasions his concerns about the amount of time his wife might be absent from home if C. was to be transplanted and its effect on the siblings, specially the 6 weeks old baby. In contrast, Mrs W. main concern was C.'s well being. C. was accepted into the transplant programme. It was agreed that, at each hospital-visit while awaiting transplantation, a child psychiatrist (B.L.) would meet C. and his mother to monitor progress, and to give recommendations to the psychiatrist working with the family locally. A good liaison GOSH-local Child and Family Centre was established.

C. received heart and heart-lung transplantation 8 months from the initial assessment. His post-operative course was smooth, but his lung function (FEV1) dropped at 5 weeks post-transplant. During the following 7 weeks, C. had a series of unexpected admissions to both the local hospital and GOSH because of recurrent problems of infections and rejection (but not seriously so). At 15 weeks post-transplantation, he appeared slightly withdrawn; the importance of discipline became important on the ward when Mrs W. was unable to make C. take his medication. She felt a need for help with general management and in particular to encourage him to eat. In addition, Mrs C. complained about her husband and felt the need to talk about changes in her marital relationship since the transplant. Due to her fears or anxieties about C., and her wish to do the best possible for him, Mrs W. had started excessive house cleaning not having any time for the other two siblings. She recognised that this was a pointless occupation but felt compelled to do it (ICD-10: obsessive-compulsive), and her husband was feeling so exasperated that, on occasions, he was blaming C. for destroying his family life. Therefore, family and marital intervention was requested from the local team.

Because C. had 15 admissions between the 4th and the 8th month post-transplantation, the local team had tremendous difficulties scheduling the family sessions. His mother accompanied most of the time C.. When Mr W. visited them at the hospital, arguments between the couple were common. Mr W. always complained about the other children growing up without a mother and the poor quality of his family life. On the ward, Mr W. was able to show some affection and care towards C., but he was feeling very ambivalent towards him. On several occasions, he expressed his wish for this child to die (rejection), so he could have his family life and his wife back. During this period the following pattern happened on several occasions: C. received intensive treatment in hospital and following his recovery the medical staff tried to send him directly home. C. became quite breathless and complained of headaches and stomach pains either on his way to the train station or on the train in the way home. Mrs W. would phone her husband to inform him that C. was unwell and she was taking him back to the hospital from the train station. Often the medical staff felt that C. symptoms were more related to anxiety, about being away from the hospital (fear of complications/dying) or about feeling unwanted by his stepfather, than to clinical problems.

At 10 months post-transplant, C. was admitted with acute rejection, infection and tracheal stenosis. He was very lethargic and refusing to take any medication or having further treatment. When the rejection was irreversible, he was discharged for home-care. Shortly after, he passed away with his family by his side.

In summary, in this case there appeared to be a consistent deterioration in the family relationships and mother's psychiatric functioning (as manifested by marital distress, decrease in warmth, a marked increase in the levels of criticism as well as hostility, and mother's rituals), which paralleled the child's physical deterioration.

CASE 2

J.D., 8 years old boy who was referred for heart-lung transplantation on 10.5.1990. J. was diagnosed as having cystic fibrosis (CF) at the age of 18 months. He was attending normal school on a regular basis. Although J. was living with both natural parents, only his mother accompanied him during his admission. His parents were in the process of separating. J. had previously lost a 3 year old sister who also had CF.

The psychiatric assessment showed no obvious abnormality but it was felt that he should be encouraged to express his feelings. His schoolteacher was happy about his academic performance, and his behaviour at school did not show any abnormality either. Mrs D. appeared withdrawn, depressed, tearful and very distressed. She scored moderately high on the GHQ-28 (score: 31) but below the cut-off point which might indicate a psychiatric diagnosis. Due to the marital problems, Mrs D. appeared to have difficulties managing J. and asked for psychological support. Therefore, they were referred to the local psychiatric services. Regarding the Expressed Emotion ratings, Mrs D. showed little overinvolvement towards J., high warmth, absence of criticism and hostility, and made two positive remarks. Surprisingly, she rated nought on critical comments and in hostility towards her husband, but none on the warmth scale.

Since J.'s mobility was good, he was initially accepted on the provisional list. However, when he was reviewed on the 26.2.91 and due to a significant deterioration (local admissions, poor quality of life, loss of weight, and missing quite a lot of school) he was moved to the active list. During this visit his mother again accompanied him. Mrs D. was very anxious and concerned about J., but appropriately so given his physical condition. J.'s natural father was not living in the household anymore; but J. had daily contact with him, and Mr D. was being very supportive to both J. and his former wife. Mrs D. described him as a 'good friend'. Mrs D. and J. had stopped attending the local Child and Family Centre.

J. received heart-lung transplantation on the 1.11.91. Both natural parents were at his bedside on this occasion. He recovered very well from the operation but developed diabetes. At 12 months post-transplant, physically, J. was remarkably well, and was taking responsibility for all aspects of medical treatment. The diabetes was under control. J. appeared very well adjusted both at home and at school and there were no psychiatric abnormalities. He had a wonderful social life and a good quality of life (very sportive and active). J.'s mother was also more relaxed and happy in herself, she scored very low on the GHQ-28 (score= 9). There were no changes in the Expressed Emotion ratings, other than at the follow-up Mrs D. made 1 critical comments about J. (untidiness), but immediately excused him because of his age.

At 3 years post-transplantation, J. was doing well physically but appeared slightly depressed and Mrs D. was said to have difficulties handling him. This was related to mother's recent splitting up with her fiancé to whom J. had

become very close. Following one joint session (Mrs D. and J. together) with the local psychiatrist who saw them prior to transplantation, this issue was sorted out.

J. is now 14 years old and has just had his 4th post-transplant annual review. Physically, he is very fit. Psychologically very well adjusted and, academically, he is doing extremely well. J. has a good social life and a warm and happy family life. J.'s mother has adjusted very well to the changes associated with J.'s age; she describes him as 'a proper teenager', enjoys his company (when he is around!) and to see him growing up and getting on with his life. She has a life of her own, very good social life, and currently, she is looking for a job. J.'s natural father has re-married but he is still a very 'good friend' and very supportive to both, J. and his mother.

In summary, in this case there appeared to be consistent improvement in the child's and mother's functioning and family relationships despite the child's physical deterioration prior to transplantation and the development of a new illness (diabetes) post-transplantation. Many transplant Centers, especially in U.S.A., would have denied transplantation to J. because of the initial marital difficulties, Mrs D.'s depression and her difficulties managing J. during the pre-transplant period.