Patient-reported quality of life outcomes for children with serious congenital heart defects

Rachel L Knowles, Thomas Day, Angie Wade, Catherine Bull, Christopher Wren, Carol Dezateux
On behalf of the UK Collaborative Study of Congenital Heart Defects (UKCSCHD)

ABSTRACT

Objective To compare patient-reported, health-related quality of life (QoL) for children with serious congenital heart defects (CHDs) and unaffected classmates and to investigate the demographic and clinical factors influencing QoL.

Design Retrospective cohort study.

Setting UK National Health Service.

Patients UK-wide cohort of children with serious CHDs aged 10–14 years requiring cardiac intervention in the first year of life in one of 17 UK paediatric cardiac surgical centres operating during 1992–1995. A comparison group of classmates of similar age and sex was recruited.

Main outcome measures Child self-report of health-related QoL scores (Pediatric Quality of Life Inventory, PedsQL) and parental report of schooling and social activities.

Results Questionnaires were completed by 477 children with CHDs (56% boys; mean age 12.1 (SD 1.0) years) and 464 classmates (55%; 12.0 (SD 1.1) years). Children with CHDs rated QoL significantly lower than classmates (CHDs: median 78.3 (IQR 65.0–88.6); classmates: 88.0 (80.2–94.6)) and scored lower on physical (CHDs: 84.4; classmates: 93.8; difference 9.4 (7.8 to 10.9)) and psychosocial functioning subscales (CHDs: 76.7; classmates: 85.0; difference 8.3 (6.0 to 10.6)). Cardiac interventions, school absence, regular medications and non-cardiac comorbidities were independently associated with reduced QoL. Participation in sport positively influenced QoL and was associated with higher psychosocial functioning scores.

Conclusions Children with serious CHDs experience lower QoL than unaffected classmates. This appears related to the burden of clinical intervention rather than underlying cardiac diagnosis. Participation in sports activities is positively associated with increased emotional well-being. Child self-report measures of QoL would be a valuable addition to clinical outcome audit in this age group.

INTRODUCTION

Increasing numbers of children operated in infancy for serious congenital heart defects (CHDs) are surviving through childhood and into adulthood. As mortality falls for these children, broader health outcomes of significance to children and their families, such as health-related quality of life (QoL) and the capacity for social and educational participation attain greater importance. Over a decade ago, the Bristol Inquiry report into the care of children undergoing cardiac surgery highlighted the lack of long-term outcome data for children with CHDs and underlined the need for a national monitoring system. Although short-term cardiac surgical outcomes for UK children are now comprehensively collected by the Central Cardiac Audit Database and published through the NICOR-Congenital Heart Disease Portal, the quality of long-term survival at a national level is not routinely captured.

Patient-reported experiences and outcomes are central to quality improvements within the National Health Service (NHS) and are increasingly being advocated for monitoring individual clinical care, which has led to an expansion in instruments designed to measure well-being, QoL and healthcare experience. Patient-reported outcome measures (PROMS) ascertain the patient’s own assessment of their health, functional status and QoL. The application of PROMS in clinical

What is already known

▸ Health, educational and quality of life (QoL) outcomes have increasing relevance to children with serious congenital heart defects (CHDs), who can now expect to survive into adulthood.

▸ Studies comparing health-related QoL outcomes of school-age children with serious CHDs and their classmate peers are lacking.

▸ Children’s own self-reported views are important outcome measures as the child’s perspective often differs from parents.

What this study adds

▸ Ten to 14-year-olds with serious congenital heart defect (CHDs) report significantly lower health-related quality of life than unaffected classmates.

▸ This reduction is related to the burden of clinical intervention and on-going care rather than cardiac diagnosis; sports participation offers positive benefit to psychosocial functioning.

▸ Collection of child-reported outcomes for CHDs is practicable and its inclusion in routine national clinical outcome monitoring and audit should be considered.
practice and outcomes research is pertinent to all chronic child-
hood disorders, of which CHDs are a key example, as outcomes
will vary across the lifetime influenced by children’s adaptation
to their changing environment and particularly the transition
to adulthood. While age-adapted questionnaires specifically suited
to self-reporting of health outcomes by children have multiplied,
there remains a reliance on proxy reporting by parents. Although parental perspectives are useful, QoL and patient
experience are subjective concepts. Consequently children’s own
views on their health and well-being should be assessed.
Moreover, evidence suggests that children’s views are reliable
and can differ greatly from the views of their parents or education
and health professionals.8

QoL measures focus on daily life experiences and outcomes
during childhood and adolescence and facilitate the develop-
ment of interventions to support families and promote resilience,
or positive adaptation, in long-term survivors with chronic
disorders.9 A limited number of multidimensional patient self-
report instruments have been validated to explore the child’s perspective on health and well-being in paediatric cardiac popu-
lations, including the impact of a CHD on current lifestyle, past experience and future expectations.2 10-12 The Pediatric Quality
of Life Inventory (PedsQL 4.0)13 is a widely employed generic
QoL instrument for children aged 2–18 years; the questionnaire
may be self-completed by children aged 5 years or over.

Our aims were to estimate and compare QoL scores reported
using the PedsQL 4.0 by a UK-wide cohort of school-age chil-
dren with serious CHDs, with a comparison group comprised
of their unaffected classmates and to investigate the factors that influence self-reported outcomes for children living with
chronic or congenital conditions.

METHODS
The UK Collaborative Study of CHDs (UKCSCHD) is a multi-
centre prospective study of almost 4000 children, born 1992–
1995 with serious CHDs requiring intervention within the first
year of life and involving all 17 UK paediatric cardiac centres
operating at that time. The cohort excluded children with
minor defects not requiring intervention,14 but included around
one-third of all children with CHDs (2/1000 live births). Information, including sex, cardiac diagnosis and cardiac proce-
dures, was obtained from individual case notes review. Each
child was assigned a primary cardiac diagnosis using Wren’s
hierarchy14 and a diagnosis-derived cardiac prognostic severity
(CPS) score, adapted from Lane15 (see online supplementary
table S1). Research ethics approval (Trent MREC 04/04/017) was
given for local cardiologists to contact surviving children with
an invitation to participate in the postal questionnaire follow-up
of QoL, health and educational outcomes.

During 2004–2007, local collaborating cardiologists
attempted to contact 2963 surviving children, then aged 10–
14 years (figure 1). Due to restrictions imposed by ethics and
governance approvals,16 families could only be contacted by the
local clinical care team after approval by the child’s cardiologist
and general practitioner; we estimate that around 70% of eli-

gible children received an invitation to participate. Of 853 chil-
dren who agreed to take part, 515 (60.3%) returned a ques-
tionnaire; 38 were subsequently excluded due to incom-
plete responses (figure 1). In addition, parents returned a parent
questionnaire for 19 children who did not return the child ques-
tionnaire; 12 of these children were reported by parents to have
significant learning difficulties which may account for non-
completion. Participants were asked to recruit a comparison
group by giving additional questionnaires to their two class-
mates closest in age and of the same sex. We received question-
naires from 479 classmates; 15 were excluded as incomplete/ineligible.

Each child completed the PedsQL 4.0 questionnaire (UK
English version). Items are scored on a Likert scale from 0 (never
a problem) to 4 (almost always a problem) then transformed to
a 0–100 scale to provide physical functioning, psychosocial
(school, social and emotional) functioning and summary scores;
higher scores represented a better QoL.13 Scores were not calcu-
lated if more than half the scale items were incomplete (physical
scale incomplete (n=3); emotional scale incomplete (n=1)). We
estimated the minimal clinically important difference (MCID),
defined as the minimum score increase on a QoL scale for a treat-
ment to be considered of patient benefit and compared this with

Figure 1 Flow diagram of
recruitment and response to
questionnaires. *Contact with families by researchers was dependent on the
local clinician obtaining consent to
contact from each child’s general
practitioner (GP). If a GP did not
provide written consent or did not
respond, questionnaires were not sent
to the family. This procedure was
mandated by the research ethics
committee in order to protect patient
confidentiality and the central study
team did not receive details of local
response rates. On this basis of the
London mailing, which did involve
members of the study team, we
estimate that only 70% of survivors
were sent an invitation to participate,
but a proportion of these were
addressed incorrectly and/or returned
unopened. Throughout the UK, 853
families agreed to receive a
questionnaire and 515 (60.3%) of
these were returned completed.

Surviving children
N=2963

Family contacted and
consented to take part
N=853 *

Families returned
questionnaires
N=515

Exclusion of incomplete
questionnaires
N=38

Affected children in final
dataset
N=477

Families returned
questionnaires
N=479

Exclusion of
questionnaires
N=14

Unaffected classmates in final
dataset
N=464

Excluded as child had
congenital heart defect
N=1

Unaffected classmate peers
recruited by families of children
with CHDs
the MCID of 4.4 points attributed to the unadjusted Child Report PedsQL Summary Scale.13

Parents (including carers or guardians acting in a parental role) completed a questionnaire, providing information about their employment, education, family and their child’s health, schooling and daily activities. Parental employment was full time (1.0) or part-time (0.5) for each parent, then summed. Parents reported regular medications (classified as cardiac or non-cardiac), vision, hearing or speech problems, special educational needs provision, school absences and participation in sporting and social activities for their child. Parents were asked whether they considered their child to have a long-standing non-cardiac illness, and if so, whether this limited their child’s activities.17

Statistical analysis
Participant characteristics were examined for response bias. As PedsQL scores were not normally distributed, median scores were compared between groups and 95% CIs for the difference in medians estimated.18 To explore factors influencing outcome, we developed univariable and multivariable regression models, using Generalised Additive Models for Location, Scale and Shape (GAMLSS) based on the Sinh-Arcsinh (SHASH) distribution, to take account of the non-normal outcome distribution.19 20 A forward variable-selection approach was used and variables retained if they improved goodness-of-fit, based on the Akaike information criterion. We assessed the need for a multilevel model to account for correlation within cardiac centres and case-control clusters; no evidence of correlation within cardiac centres was found and we adjusted for clustering of cases and controls by including three levels of school-type factors (mainstream school, mainstream school with learning support or special school/unit) in regression models.

Sensitivity analyses explored the effect of excluding children who did not recruit classmates, as these children were more likely to attend special schools. Statistical analyses were performed in R V12.2.1 (R Foundation for Statistical Computing, Austria).

RESULTS
Data from 477 affected children (268 boys (56%); mean age 12.1 (SD 1.0) years) and 464 classmates (255 boys (55%); mean age 12.0 (SD 1.1) years) were analysed. Characteristics of children with CHDs were compared with non-responding survivors (n=2486) in the UKCSCHD cohort (see online supplementary table S1). Children returning questionnaires were representative of all primary cardiac diagnoses, although children with more severe CPS scores (palliated CHDs) appeared more likely to return questionnaires. Characteristics of affected children and classmates were compared (table 1); children with CHDs were on average 200 g lighter at birth, more likely to take regular medications, have associated health problems and health-related absences from school and participated less frequently in sport and social activities.

Unadjusted median PedsQL physical functioning, psychosocial functioning and summary scores for children with CHDs and unaffected classmates differed significantly (figure 2). The unadjusted median summary score was 78.3 (IQR 65.0–88.6) for the affected children, compared to 88.0 (80.2–94.6) for unaffected classmates (difference 9.8 (95% CI 7.1 to 12.4)). Unadjusted median physical and psychosocial functioning scores for children with CHDs were (84.4 (63.6–93.8) and 76.7 (62.5–86.7)), respectively; difference 9.4 (95% CI 7.8 to 10.9) and significantly lower than for unaffected classmates (93.8 (84.0–100.0) and 85.0 (76.7–93.3), respectively; difference 8.3 (95% CI 6.0 to 10.6)).

In univariable regression models (see online supplementary table S2), factors significantly associated with lower scores included worse CPS score, increasing number of cardiac interventions, long-standing limiting non-cardiac illness, regular medications, longer school absence, and vision or speech problems. Hearing difficulties were associated with worse psychosocial functioning and summary scores only. Increasing frequency of sporting and social activities was associated with higher PedsQL scores, and higher psychosocial functioning and summary scores were associated with both parents living in the family home and parents in employment.

In multivariable models (table 2), adjusting for school-type and sociodemographic factors such as parent education or employment, having a CHD remained an independent predictor of significantly worse summary, physical and psychosocial functioning scores. Non-cardiac comorbidities, indicated by limiting long-standing illness and regular medications, were also independently associated with poorer outcomes. Visual or hearing difficulties were associated with lower summary scores only, whereas school absence was associated with poorer summary and psychosocial, but not physical, functioning scores. Increased frequency of sports participation remained associated with better summary and psychosocial, but not physical, functioning scores (table 3A). Individual child age, sex or ethnicity, and parental educational or employment, did not independently influence self-reported QoL for children in our study. These findings persisted in sensitivity analyses.

For children with CHDs physical functioning, psychosocial functioning and summary scores decreased significantly as the number of cardiac interventions increased; conversely the diagnosis-based CPS score was not associated with outcome (table 3B).

DISCUSSION
Our study demonstrates that children with serious CHDs report significantly impaired QoL compared with their unaffected classmates, with unadjusted median scores 8–10 points lower on the summary, physical and psychosocial functioning PedsQL scales. The difference we observed between affected children and their classmates exceeds the MCID for the PedsQL summary scale and therefore has clinical relevance. Significant differences persisted after adjustment for age, sex, ethnicity and sociodemographic factors. Specific CHD diagnosis was not associated with QoL, whereas a higher cumulative burden of cardiac interventions over the lifetime had a significant negative impact. Additional factors associated with reduced QoL independently of having a CHD were non-cardiac comorbidities, specifically long-standing limiting non-cardiac illness, vision or hearing difficulties, and the need to take regular medications or time off from school for health reasons. Our findings suggest that children whose full inclusion in school and social activities is limited by a chronic disorder, whether this is cardiac or non-cardiac, experience lower subjective QoL.

Latal2 identified seven studies in which QoL after cardiac surgery was measured using child self-report instruments. Study findings were inconsistent, which could be related to instrument heterogeneity or variable participant selection. In studies of children with chronic conditions, worse PedsQL scores were reported by children with cardiac defects in comparison with population norms10 21–23 or ‘healthy’ comparison groups.24–26 Children with more ‘severe’ CHDs rated QoL lower, although ‘severe’ was variously defined as cyanosis,24,25 or type of intervention.10 21
Many studies involved only a single CHD diagnosis; therefore, generalisability is limited. Although methodological differences limit direct comparison with our findings, studies involving paediatric chronic disease groups have demonstrated reductions in PedsQL scores comparable with CHDs, for children with end-stage renal disease, epilepsy and cystic fibrosis, while children with diabetes experience less reduction in QoL.

Table 1  Characteristics of affected children and their classmates participating in the study

<table>
<thead>
<tr>
<th></th>
<th>Children with CHDs N=477</th>
<th>Unaffected classmates N=464</th>
<th>p Value for difference</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>N (%)</td>
<td>Missing (N (%))</td>
<td>N (%)</td>
</tr>
<tr>
<td><strong>Individual factors</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>268 (56)</td>
<td>0</td>
<td>255 (55)</td>
</tr>
<tr>
<td>Age (years)</td>
<td>12.1 (1.0)*</td>
<td>0</td>
<td>12.0 (1.1)*</td>
</tr>
<tr>
<td>Ethnicity</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>White</td>
<td>443 (93)</td>
<td>7 (2)</td>
<td>420 (90)</td>
</tr>
<tr>
<td>Non-white</td>
<td>30 (6)</td>
<td></td>
<td>37 (8)</td>
</tr>
<tr>
<td>Birth weight (g)</td>
<td>3228 (673.5)*</td>
<td>16</td>
<td>3443 (603.0)*</td>
</tr>
<tr>
<td><strong>Parental and family factors</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Parental education level</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>None/General Certificate of Secondary Education</td>
<td>168 (35)</td>
<td>142 (31)</td>
<td>0.14</td>
</tr>
<tr>
<td>A level</td>
<td>140 (29)</td>
<td>129 (28)</td>
<td></td>
</tr>
<tr>
<td>Degree</td>
<td>169 (36)</td>
<td>193 (41)</td>
<td></td>
</tr>
<tr>
<td>Number of full-time equivalent working parents†</td>
<td>1.2 (0.58)*</td>
<td>1.3 (0.56)*</td>
<td>0.19</td>
</tr>
<tr>
<td>Number of siblings at home</td>
<td>2.0 [2.0–2.0]†</td>
<td>0</td>
<td>2.0 [2.0–2.0]†</td>
</tr>
<tr>
<td>Two parent(s) at home at birth</td>
<td>452 (95)</td>
<td>0</td>
<td>444 (96)</td>
</tr>
<tr>
<td>Two parent(s) at home now</td>
<td>385 (81)</td>
<td>0</td>
<td>364 (78)</td>
</tr>
<tr>
<td>Mother’s age at child’s birth (years)</td>
<td>29.6 (5.1)*</td>
<td>6</td>
<td>29.8 (4.8)*</td>
</tr>
<tr>
<td>Father’s age at child’s birth (years)</td>
<td>32.1 (6.1)*</td>
<td>11</td>
<td>32.0 (5.9)*</td>
</tr>
<tr>
<td><strong>Comorbidities</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Non-cardiac long-standing illness</td>
<td>20</td>
<td>13</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Yes, not limiting</td>
<td>66 (14)</td>
<td>48 (10)</td>
<td></td>
</tr>
<tr>
<td>Yes, limiting</td>
<td>111 (23)</td>
<td>31 (7)</td>
<td></td>
</tr>
<tr>
<td>Uses regular non-cardiac medications</td>
<td>170 (36)</td>
<td>3</td>
<td>40 (9)</td>
</tr>
<tr>
<td><strong>School and daily life activities</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Type of schooling</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mainstream school</td>
<td>326 (68)</td>
<td>418 (90)</td>
<td></td>
</tr>
<tr>
<td>Mainstream with assistance</td>
<td>112 (23)</td>
<td>40 (9)</td>
<td></td>
</tr>
<tr>
<td>Special school/unit</td>
<td>39 (8)</td>
<td>6 (1)</td>
<td></td>
</tr>
<tr>
<td>School absence in last year</td>
<td>4</td>
<td>5</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Never</td>
<td>105 (22)</td>
<td>148 (32)</td>
<td></td>
</tr>
<tr>
<td>&lt;1 week</td>
<td>193 (40)</td>
<td>227 (49)</td>
<td></td>
</tr>
<tr>
<td>1–2 weeks</td>
<td>102 (21)</td>
<td>56 (12)</td>
<td></td>
</tr>
<tr>
<td>2 weeks–1 month</td>
<td>46 (10)</td>
<td>20 (4)</td>
<td></td>
</tr>
<tr>
<td>&gt;1 month</td>
<td>28 (6)</td>
<td>8 (2)</td>
<td></td>
</tr>
<tr>
<td>Frequency of sport§</td>
<td>1.0 [0.5–3.0]†</td>
<td>0</td>
<td>2.0 (1.0–3.0)†</td>
</tr>
<tr>
<td>Frequency of social activities¶</td>
<td>5.0 [2.5–6.5]†</td>
<td>0</td>
<td>6.0 (4.0–7.2)†</td>
</tr>
<tr>
<td><strong>Cardiac factors</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No cardiac disorder</td>
<td>0</td>
<td>464 (100)</td>
<td>0</td>
</tr>
<tr>
<td>Cardiac prognostic severity (CPS)</td>
<td>1 (&lt;1)</td>
<td></td>
<td>N/A</td>
</tr>
<tr>
<td>Curative</td>
<td>102 (21)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Corrective</td>
<td>272 (57)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Palliative</td>
<td>103 (22)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Number of cardiac interventions</td>
<td>2.0 [1.0–3.0]†</td>
<td>52</td>
<td>0</td>
</tr>
</tbody>
</table>

*Mean (SD).
†Includes carers/guardians.
‡Median [IQR] based on parent questionnaire (parents reported all interventions where hospital case notes were incomplete and missing data reflects where parents could not provide data).
§Number of occasions child takes part in a sporting activity (not school PE) per week.
¶Number of occasions outside of school hours that a child takes part in sport, plays with friends or attends non-sport clubs per week.
CHD, congenital heart defect.

CHD, congenital heart defect; PedsQL, Pediatric Quality of Life Inventory; QoL, quality of life.
Figure 2  Unadjusted median PedsQL scores for children with CHDs (n=477) compared with unaffected classmates (n=464).

Table 2  Multivariable models to investigate factors associated with PedsQL outcome scores

<table>
<thead>
<tr>
<th></th>
<th>PedsQL summary score</th>
<th>Physical functioning</th>
<th>Psychosocial functioning</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Est.</td>
<td>SE</td>
<td>p Value</td>
</tr>
<tr>
<td><strong>Individual factors</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Presence of CHD</td>
<td>−2.58</td>
<td>0.77</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Female</td>
<td>−0.29</td>
<td>0.77</td>
<td>0.71</td>
</tr>
<tr>
<td>Age</td>
<td>−0.30</td>
<td>0.38</td>
<td>0.42</td>
</tr>
<tr>
<td>White</td>
<td>−0.12</td>
<td>1.70</td>
<td>0.95</td>
</tr>
<tr>
<td>Birth weight (per kg)</td>
<td>−0.32</td>
<td>0.57</td>
<td>0.58</td>
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<tr>
<td><strong>Parental and family factors</strong></td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>Parental education</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>None/General Certificate of Secondary Education</td>
<td>ref</td>
<td>0.94</td>
<td>0.73</td>
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<tr>
<td>A level</td>
<td>0.33</td>
<td>0.89</td>
<td>0.83</td>
</tr>
<tr>
<td>Degree</td>
<td>0.19</td>
<td>0.42</td>
<td></td>
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<tr>
<td>Number of full-time equivalent working parents†</td>
<td>−0.09</td>
<td>0.81</td>
<td>0.91</td>
</tr>
<tr>
<td>Number of siblings at home</td>
<td>6.09</td>
<td>8.16</td>
<td>0.46</td>
</tr>
<tr>
<td>Two parents† at home now</td>
<td>−4.63</td>
<td>8.20</td>
<td>0.57</td>
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<tr>
<td>Two parents† at birth</td>
<td>−1.23</td>
<td>1.75</td>
<td>0.48</td>
</tr>
<tr>
<td>Mother’s age at birth</td>
<td>0.09</td>
<td>0.11</td>
<td>0.42</td>
</tr>
<tr>
<td>Father’s age at birth</td>
<td>−0.01</td>
<td>0.10</td>
<td>0.94</td>
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<tr>
<td><strong>Comorbidities</strong></td>
<td></td>
<td></td>
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</tr>
<tr>
<td>Non-cardiac long-standing illness</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>None</td>
<td>ref</td>
<td>1.17</td>
<td>0.88</td>
</tr>
<tr>
<td>Yes, not limiting</td>
<td>−0.17</td>
<td>1.04</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Yes, limiting</td>
<td>−5.71</td>
<td>1.38</td>
<td></td>
</tr>
<tr>
<td>Uses regular medications</td>
<td>−3.74</td>
<td>0.98</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Problems with vision</td>
<td>−1.90</td>
<td>0.85</td>
<td>0.03</td>
</tr>
<tr>
<td>Problems with hearing</td>
<td>−2.32</td>
<td>0.97</td>
<td>0.02</td>
</tr>
<tr>
<td>Problems with speech</td>
<td>2.04</td>
<td>1.58</td>
<td>0.20</td>
</tr>
</tbody>
</table>

Adjusted for school type, that is, mainstream school, mainstream school with learning support or special school/unit.

† Includes carers/guardians.

CHD, congenital heart defect.
Our prospective population-based study included all UK paediatric cardiac surgical centres and is representative of children born during the 1990s with a CHD requiring intervention. Only children who had a cardiac intervention before the age of 1 year were included, nevertheless few children with complex and significant CHDs will have been excluded by this pragmatic definition of severity. Although governance restrictions limited the questionnaire to two-thirds of survivors, we achieved 60% response rate from families and comparison of respondent and non-respondent characteristics did not indicate systematic bias. A limitation of our postal survey method is that we are unable to verify whether children completed their questionnaires without support or influence from others and, as noted above, some children with learning difficulties were unable to complete their questionnaire. Children with palliated defects appeared more likely to respond; however, these children also had more frequent outpatient visits so their contact details were more likely to be current. Over 400 classmates of survivors designed specifically to capture the child’s perspective has.

PROMs are a key development as many healthcare outcomes, such as reduced symptoms or improvements in functional status and QoL, can only be assessed by patients. PROMs are increasingly being used to support shared decision-making between patients and clinicians. We have demonstrated the feasibility of using the PedsQL to obtain patient-reported outcomes for UK children living with a CHD. The added benefit of using measures designed specifically to capture the child’s perspective has significantly reduced emotional and behavioural problems. It is clear that participation in sporting or social activities represents a complex mediator of risk, influenced partly by the physical limitation imposed by a CHD. It is conceivable that some children with CHDs who participate fully in school and sports rate their psychosocial QoL high despite scoring their physical functioning lower, because they understand implicitly that they are ‘successfully’ negotiating the physical limitations of their condition.

Our study explores the impact of living with a CHD for children who have a range of defects, many of which might be considered surgically ‘corrected’ in infancy. Crucially, the mediating factors that might protect children from adverse QoL outcomes are likely to differ between individuals and may change over time. Experiencing recent or frequent health interventions may increase awareness of CHD as an ongoing health burden, with a consequent negative impact on QoL. We found decreased QoL associated with regular medication or cardiac interventions; in contrast, diagnostic severity was not an independent predictor of QoL. Greater attention may therefore need to be paid to the cumulative burden of interventions and medical care experienced by young patients.

Table 3 Multivariable models to investigate the additional effect on PedsQL outcome scores of daily life activities and cardiac severity (children with CHDs only)§

<table>
<thead>
<tr>
<th>PedsQL summary score</th>
<th>Physical functioning</th>
<th>Psychosocial functioning</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>A: Daily life activities</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>School absence in last year</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Never</td>
<td>ref</td>
<td>1.07</td>
</tr>
<tr>
<td>&lt;1 week</td>
<td>−1.37</td>
<td>1.23</td>
</tr>
<tr>
<td>1–2 weeks</td>
<td>−4.46</td>
<td>1.65</td>
</tr>
<tr>
<td>2 weeks–1 month</td>
<td>−5.29</td>
<td>1.81</td>
</tr>
<tr>
<td>&gt;1 month</td>
<td>−10.12</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td><strong>B: Cardiac severity (children with CHDs only)</strong>§</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cardiac factors</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Curative</td>
<td>ref</td>
<td>1.55</td>
</tr>
<tr>
<td>Corrective</td>
<td>−1.17</td>
<td>1.35</td>
</tr>
<tr>
<td>Palliative</td>
<td>0.14</td>
<td>0.09</td>
</tr>
<tr>
<td>Number of cardiac interventions</td>
<td>−0.74</td>
<td>0.16</td>
</tr>
</tbody>
</table>

*Number of occasions child takes part in a sporting activity (not school PE) per week.
†Number of occasions outside of school hours that a child takes part in sport, plays with friends or attends non-sport clubs per week.
‡Adjusted for school type, individual factors, parent and family factors and comorbidities.
§Adjusted for school type, individual factors, parent and family factors, comorbidities and activities of daily life.

Est., estimate.

...
been clearly highlighted.35 Paediatric patient-report measures should be considered for integration into routine monitoring of chronic childhood disorders, and specifically to enrich cardiac audit and provide an additional source for evaluating and effecting improvements in care. A child-centred approach is fundamental to communication between children, families, health and education professionals about individual care, as well as to promoting good coping strategies and social inclusion to enhance the lives of children with CHDs for whom long-term survival in adulthood is now a realistic expectation.

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Contributors All authors have contributed substantially to the study, approved the final version of the manuscript and share responsibility for the results. RLK, CD, CB and CW conceptualised and designed the study, designed the data collection instruments and undertook data collection, TD and AKH undertook analyses; all authors reviewed the analyses. RLK and TD drafted the manuscript. All authors critically reviewed and approved the final manuscript. The guarantor for this study is RLK.

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Competing interests None.

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Data sharing statement Unpublished data are not available for sharing outside of the UKCSCHD collaborating centres.

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