Medically unexplained visual loss in children and young people: an observational single site study of incidence and outcomes

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Conflict of interest

The authors declare no conflict of interest.

Abstract

Aims To determine the incidence of medically unexplained visual loss (MUVL) in children in an open access children's eye casualty.

Methods We collated demographic and clinical data of consecutive patients younger than 16 years who presented to the children's eye casualty at Moorfields Eye Hospital over a 12-month period and were diagnosed with MUVL or suspected MUVL. We reviewed the clinical records at least three months after initial presentation. We calculated the incidence using the number of "new patient" attendances over the same period as denominator (n=2 397). We used descriptive analysis. Main outcome measures: number of patients diagnosed with MUVL, proportion of patients with a history of or present psychological problems, recovery rate, improvement in visual acuity.

Results We identified 85 cases of MUVL (54 females; mean age: 9 years (IQR 7 to 12)). The median duration of follow-up was 1.2 months (IQR 0 to 4.3). The estimated annual incidence was 3.5% (95% confidence interval 2.9 to 4.4%). 33% of children had a history of psychiatric disorders, reported a stressful life event or showed signs of psychiatric disorder at the time of first presentation. The recovery rate was 25%. Median improvement in best corrected visual acuity from presentation to last appointment was 0.22 (IQR 0.06 to 0.43) logMAR.

Conclusions The incidence of MUVL is higher and the rate of resolution lower than previously reported. MUVL may be associated with mental health problems. We recommend screening for psychological problems to facilitate access to psychological treatment**.**

Introduction

Medically unexplained visual loss (MUVL) describes visual loss or visual symptoms in the absence of any medically detectable eye, visual pathway or brain condition. It is classified as a conversion disorder, in DSM-5, that is, a functional neurological symptom disorder resulting in loss of function. As with other medically unexplained symptoms, there is no universally accepted definition.[1] A number of different terms are used to describe the condition, and terminology has evolved over time (medically unexplained visual loss, non-organic visual loss, functional visual loss, hysterical visual loss, malingering, non-physiologic visual loss, factitious visual loss, psychogenic visual loss, hypochondriasis, and conversion disorder of vision).[2-5] We will use the term medically unexplained vision loss throughout this manuscript as this is the term families have told us is most acceptable as it makes no assumptions about cause.

In children, MUVL is not uncommon. The reported prevalence ranges from 1 to 9%.[6-8] The incidence of MUVL has been estimated at 1 to 1.75%.[9, 10] As in adults with MUVL[11] and other medically unexplained symptoms [12], socio-economic factors may also contribute to MUVL in children .

All previous studies indicate that girls are more commonly affected than boys.[9, 13-15] The reported mean age at presentation ranges from 9.0 to 13.4 years, but younger children can also be affected.[9, 10, 13-18] The most common complaints are deterioration of visual acuity, visual field defects and double vision.[5, 9, 14, 15] In the majority of cases both eyes are affected.[9, 13-15] Other symptoms are photopsia (perception of flashes of light which are usually brief and intermittent), perception of phosphenes (light perceptions of any colour or shape other than intermittent flashes which are not induced by light entering the eye), photophobia (light hyper-sensitivity), dyschromatopsia (altered perception of colours), amblyopia, voluntary nystagmus, accommodation weakness, ptosis, blepharospasm and painful eyes.[3, 5, 14, 15, 19] Some children have a history of previous eye diseases and treatment.[13, 20] MUVL in the presence of known eye diseases and/or non-ocular conditions such as

asthma, autoimmune diseases and accidental[13, 21] or surgical trauma is referred to as functional overlay.[20]

Children with MUVL are more likely to also report other medically unexplained physical symptoms such as headaches and abdominal or limb pain.[14] MUVL is also associated with factors similar to those underlying other medically unexplained physical symptoms. For example, 40 to 90%[9, 19, 22] of children with MUVL also report psychological stressors such as family problems, problems at school or bullying.[10, 19, 23] High rates of mental health problems have been reported in adults with MUVL[14] , and some research has indicated that young people with MUVL are more likely to report symptoms such as depression and attention deficit hyperactivity disorder.[16,25-28] As with other medically unexplained symptoms, there are likely to be multiple interacting causal factors, and the presence of comorbid mental health disorders does not suggest that symptoms are 'all in the mind'. In other medically unexplained symptoms (e.g. headache, stomach pains etc.), as many as 30-50% of children have associated mental health disorders.[24] Screening for, and detecting mental health problems in children with MUVL may facilitate access to appropriate services.[2, 4, 14, 15]

The rate of spontaneous resolution of MUVL in children has been reported to be high, particularly in studies with long follow-up data, ranging from 37% at 12 months[10] to 100%[9] (unknown duration of follow-up); the management of MUVL therefore often focuses on providing reassurance to the child and family that the visual prognosis is excellent.

There are no management recommendations for ophthalmologists, beyond the establishment of the diagnosis. In order to establish current practice and outcomes and to facilitate service planning and the development of future research projects we carried out a retrospective observational study to describe incidence, clinical characteristics of patients, current diagnostic workup and outcomes of MUVL in children.

Subjects and Methods

This service evaluation had Trust approval (CA16/ONSP/91). A research fellow (MCD) ~~We~~ collated demographic and clinical data of consecutive patients younger than 16 years who presented to the children's eye casualty at Moorfields Eye Hospital over a 12-month period and were diagnosed with MUVL or suspected MUVL. The research fellow ~~We~~ reviewed clinical records at least three months after the initial presentation. All information was gathered from the clinical notes. We calculated the incidence using the number of "new patient" attendances over the same period as denominator (n=2 397). Patients were included into the study if a diagnosis of "MUVL" or "functional visual loss" was documented in the medical notes and was not revised over subsequent clinic visits.

We recorded any history of previous eye problems that had occurred at least four weeks before the presentation which led to a diagnosis of MUVL and could therefore be reasonably assumed to be unrelated. Children were considered as having fully recovered if they felt the eye problems had completely resolved and visual acuity was at least 0.1 logMAR in the initially affected eye. In cases where visual acuity at first presentation could not be determined in logMAR values ("hand movements" or "perception of light"), we did not quantify the change in vision between visits. The main outcome measures were the number of patients diagnosed with MUVL, the proportion of patients with a history of or present psychological problems, the recovery rate and the improvement in visual acuity. All data were analysed using descriptive statistics.

Results

Incidence

We identified 88 cases of suspected MUVL. Three children were subsequently found to have isolated optic disc atrophy, macular dystrophy, or optic neuropathy, confirmed by abnormal electrophysiological findings. These children were excluded from the analysis. 85 cases were

included in the analysis. We estimated the annual incidence in our setting to be 3.5% (95% confidence interval 2.85 to 4.35%). The number of children diagnosed with MUVL peaked during the winter months (Fig. 1).

Patient characteristics

The median age at presentation was 9 years (IQR 7 to 12). 54 patients were girls. The median number of appointments was 2 (IQR 1 to 3). 28 children only attended once. The median time from onset of symptoms until initial presentation was 1 week (IQR 0.14 to 2); the median duration of follow-up was 1.2 months (IQR 0 to 4.3) (Table 1).

Diagnostic workup and findings

There was considerable variability in diagnostic investigations (Tab. 1). 31% of all children were diagnosed with orthoptic abnormalities or refractive errors, 12% showed abnormal visual field test results. 57% had a previous history of contact with eye health professionals, for glasses or surgical procedures.

None of the children seen during this period were referred for psychological assessment or intervention.

Patient history and presentation

64% of all children had bilateral symptoms. 36% had a history of eye problems or ocular surgery. 41% had glasses at first presentation. The most common complaints were deterioration of visual acuity (68%), painful eyes (24%), photopsia or perception of phosphenes (19%) and diplopia (19%). Complete loss of vision (13%), photophobia (9%), visual field loss (7%) and swollen lids (7%) were less common.

Ocular symptoms were associated with non-ocular symptoms in 35% of all cases, headache being the most common complaint (93%).

48% of all children had non-ocular health problems such as allergies, asthma and hypothyroidism. Rare diagnoses were complex regional pain syndrome, lactose intolerance, Marfan syndrome, migraine and thalassaemia. A brief behavioural and emotional symptom history and a history of previous clinical service use was taken as is usual in any paediatric consultation. 33% of all children

had a history of psychiatric disorders, or showed signs of psychiatric disorder at the time of first presentation. One child currently reported current clinical levels of depression (under psychiatric care), three children had a history of psychiatric problems but no longer showed symptoms at the time of presentation and 28% reported stressful live events. 24% reported recent injuries.

Clinical course and resolution of symptoms

At last follow-up 21 children (25%) had fully and 12 (14%) had partially recovered (resolution rate at 3 months after first presentation: 13%, resolution rate after at least three months of follow-up: 34%). The median improvement in best corrected visual acuity (worse affected eye) was 0.22 logMAR (IQR 0.06 to 0.43). When tested for visual acuity, four children claimed not to be able to see anything or to perceive light or hand movements only; we excluded these from the analysis. Three of these children had normal visual acuities at the last follow-up. One child did not report any clinically significant improvement of visual acuity.

Discussion

Our principal finding of a 3.5% incidence of MUVL in children and young people attending a specialist ophthalmological hospital is two to three times higher than previously reported [10,11]. Our study also challenges the commonly held belief that - in children - MUVL has a high rate of spontaneous resolution. We report here that three months after presentation, 87% of those children who had at least three months of follow-up still experienced vision problems.

The high incidence of MUVL in this study may be explained in part by the setting, a walk-in clinic in a specialist ophthalmological hospital providing tertiary-care level workup for patients who often have not consulted their general practitioner or local optometrist before attending our service. We observed that a high proportion of children with MUVL had previous experience with eye care providers; this may have raised their awareness of the possibility of having eye problems and direct access to eye care providers. The number of children diagnosed with MUVL peaked during the winter months, indicating possible seasonal variation. However, our sample size is small and limited to one calendar year only. In addition, we did not systematically ask children about any seasonal stressors/relieving factors (e.g. school examinations/school holidays). Further research is required to establish any seasonal variation in incidence of MUVL.

MUVL may have commonalities with other medically unexplained symptoms such as headaches, abdominal pain and non-epileptic seizures, including comorbidity with common mental health disorders including anxiety, depression and behavioural problems.[24] Currently there is little known about the mental health of young people with MUVL and there is no routine screening, established referral pathways or guidelines for the detection and management of mental health difficulties associated with MUVL. It has been suggested that an absence of mental health disorders may be associated with faster resolution of symptoms,[25] though this finding is controversial.[14, 19] Though psychiatric consultation has not yet been shown to improve final visual outcome,[7] patients may benefit from addressing psychological issues.[13] Cognitive behavioural and whole system approaches

to the management of other medically unexplained symptoms and associated mental health difficulties are successful for the management for both adult patients and children and young people.[26-30] Screening for psychiatric comorbidity in MUVL in young people, will allow early detection of emotional and behavioural problems, and facilitate access to evidence based psychological therapies. Therefore, a comprehensive multidisciplinary assessment of these children is likely to include, in addition to the ophthalmological and medical history, a mental health review, family history and social and educational history.

The low rate of recovery in this study compared with other publications (93%[14] to 100%[9]) may in part be explained by the lack of a standardized definition of "complete resolution", and by the relatively short follow-up in our study. Ophthalmologists are often satisfied when good vision can be demonstrated, and limit management to providing a "strong dose of reassurance" that symptoms will resolve.[7-9, 13] Some discuss psychological aspects with the family and the general practitioner.[15] Few refer children for neuropsychological evaluation.[5] In other medically unexplained conditions, the presence of an unrecognised comorbid mental health problem can impact negatively on the symptom trajectory.[24, 31] In recent years there has been an emphasis on integrating mental and physical healthcare therefore it is necessary to ensure that young people with MUVL are referred to appropriate evidence based services for treatment if a psychiatric comorbidity is identified.

Limitations of our work include data collection at a single site in a highly urbanised area and the relatively short follow-up duration. The present study does not allow conclusions on the long-term course of MUVL. Longitudinal studies with a longer follow-up duration could provide valuable information on the fluctuation of symptoms and the likelihood of relapses and or the simultaneous or delayed manifestation of other types of medically unexplained symptoms. However, our setting caters for a multi-ethnic urban population and we expect our findings could be replicated in similar settings. A further limitation is the current lack of a "positive diagnosis" and a lack of consensus in terminology. A recent qualitative study of non-epileptic seizures highlighted the importance of families and young people having ownership over the terminology used to describe their

condition,[32, 33] and the field is likely to be advanced through qualitative studies to explore and examine the experiences of young people with MUVL.

To achieve optimum and rapid recovery in paediatric MUVL it is likely that integrated ophthalmological and mental health assessment and treatment will be needed. The low rate of full recovery of MUVL with ophthalmological approaches alone, suggests that additional interventions may be needed. Identifying, understanding and alleviating psychosocial stressors may be important as they may be precipitants or causes of MUVL. In addition, establishing rates of psychiatric comorbidity (for example anxiety, depression etc) in these children will improve understanding of mechanisms and identify additional treatment targets.

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355 **Titles and legends to figures**

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357 **Table 1.** Incidence, clinical work-up and demographical and clinical characteristics of children
358 diagnosed with MUVL.

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360 **Figure 1.** Seasonal variation of the number of children diagnosed with MUVL. Peak in the winter
361 months.