

Paediatric MUVL: incidence, diagnosis, outcome

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

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34 Conflict of interest

35 The authors declare no conflict of interest.

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38 **Abstract**

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

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

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

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

58 **Introduction**

59

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

70 In children, MUVL is not uncommon. The reported prevalence ranges from 1 to 9%.⁶⁻⁸ The incidence
71 of MUVL has been estimated at 1 to 1.75%.^{9,10} As in adults with MUVL¹¹ and other medically
72 unexplained symptoms¹², socio-economic factors may also contribute to MUVL in children .

73 All previous studies indicate that girls are more commonly affected than boys.^{9, 13-15} The reported
74 mean age at presentation ranges from 9.0 to 13.4 years, but younger children can also be affected.^{9,}

75 ^{10, 13-18} The most common complaints are deterioration of visual acuity, visual field defects and

76 double vision.^{5,9, 14, 15} In the majority of cases both eyes are affected.^{9, 13-15} Other symptoms are

77 photopsia (perception of flashes of light which are usually brief and intermittent), perception of

78 phosphenes (light perceptions of any colour or shape other than intermittent flashes which are not

79 induced by light entering the eye), photophobia (light hyper-sensitivity), dyschromatopsia (altered

80 perception of colours), amblyopia, voluntary nystagmus, accommodation weakness, ptosis,

81 blepharospasm and painful eyes.^{3, 5, 14, 15, 19} Some children have a history of previous eye diseases and

82 treatment.^{13, 20} MUVL in the presence of known eye diseases and/or non-ocular conditions such as

83 asthma, autoimmune diseases and accidental^{13, 21} or surgical trauma is referred to as functional
84 overlay.²⁰

85 Children with MUVL are more likely to also report other medically unexplained physical symptoms
86 such as headaches and abdominal or limb pain.¹⁴ MUVL is also associated with factors similar to
87 those underlying other medically unexplained physical symptoms. For example, 40 to 90%^{9, 19, 22} of
88 children with MUVL also report psychological stressors such as family problems, problems at school
89 or bullying.^{10, 19, 23} High rates of mental health problems have been reported in adults with MUVL¹⁴,
90 and some research has indicated that young people with MUVL are more likely to report symptoms
91 such as depression and attention deficit hyperactivity disorder.^{16, 25-28} As with other medically
92 unexplained symptoms, there are likely to be multiple interacting causal factors, and the presence of
93 comorbid mental health disorders does not suggest that symptoms are 'all in the mind'. In other
94 medically unexplained symptoms (e.g. headache, stomach pains etc.), as many as 30-50% of children
95 have associated mental health disorders.²⁴ Screening for, and detecting mental health problems in
96 children with MUVL may facilitate access to appropriate services.^{2, 4, 14, 15}

97 The rate of spontaneous resolution of MUVL in children has been reported to be high, particularly in
98 studies with long follow-up data, ranging from 37% at 12 months¹⁰ to 100%⁹ (unknown duration of
99 follow-up); the management of MUVL therefore often focuses on providing reassurance to the child
100 and family that the visual prognosis is excellent.

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

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109 **Subjects and Methods**

110 This service evaluation had Trust approval (CA16/ONSP/91). A research fellow (MCD) ~~We~~ collated
111 demographic and clinical data of consecutive patients younger than 16 years who presented to the
112 children's eye casualty at Moorfields Eye Hospital over a 12-month period and were diagnosed with
113 MUVL or suspected MUVL. The research fellow ~~We~~ reviewed clinical records at least three months
114 after the initial presentation. All information was gathered from the clinical notes. We calculated the
115 incidence using the number of "new patient" attendances over the same period as denominator
116 (n=2 397). Patients were included into the study if a diagnosis of "MUVL" or "functional visual loss"
117 was documented in the medical notes and was not revised over subsequent clinic visits.
118 We recorded any history of previous eye problems that had occurred at least four weeks before the
119 presentation which led to a diagnosis of MUVL and could therefore be reasonably assumed to be
120 unrelated. Children were considered as having fully recovered if they felt the eye problems had
121 completely resolved and visual acuity was at least 0.1 logMAR in the initially affected eye. In cases
122 where visual acuity at first presentation could not be determined in logMAR values ("hand
123 movements" or "perception of light"), we did not quantify the change in vision between visits. The
124 main outcome measures were the number of patients diagnosed with MUVL, the proportion of
125 patients with a history of or present psychological problems, the recovery rate and the improvement
126 in visual acuity. All data were analysed using descriptive statistics.

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129 **Results**

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131 **Incidence**

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

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

138

139 **Patient characteristics**

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

144

145 **Diagnostic workup and findings**

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

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

152

153 **Patient history and presentation**

154 64% of all children had bilateral symptoms. 36% had a history of eye problems or ocular surgery.
155 41% had glasses at first presentation. The most common complaints were deterioration of visual
156 acuity (68%), painful eyes (24%), photopsia or perception of phosphenes (19%) and diplopia (19%).
157 Complete loss of vision (13%), photophobia (9%), visual field loss (7%) and swollen lids (7%) were
158 less common.

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

161 48% of all children had non-ocular health problems such as allergies, asthma and hypothyroidism.

162 Rare diagnoses were complex regional pain syndrome, lactose intolerance, Marfan syndrome,
163 migraine and thalassaemia. A brief behavioural and emotional symptom history and a history of

164 previous clinical service use was taken as is usual in any paediatric consultation. 33% of all children

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

169

170 **Clinical course and resolution of symptoms**

171 At last follow-up 21 children (25%) had fully and 12 (14%) had partially recovered (resolution rate at
172 3 months after first presentation: 13%, resolution rate after at least three months of follow-up:
173 34%). The median improvement in best corrected visual acuity (worse affected eye) was 0.22
174 logMAR (IQR 0.06 to 0.43). When tested for visual acuity, four children claimed not to be able to see
175 anything or to perceive light or hand movements only; we excluded these from the analysis. Three
176 of these children had normal visual acuities at the last follow-up. One child did not report any
177 clinically significant improvement of visual acuity.

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181 **Discussion**

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

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

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

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

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

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

233 condition,^{32, 33} and the field is likely to be advanced through qualitative studies to explore and
234 examine the experiences of young people with MUVL.
235 To achieve optimum and rapid recovery in paediatric MUVL it is likely that integrated
236 ophthalmological and mental health assessment and treatment will be needed. The low rate of full
237 recovery of MUVL with ophthalmological approaches alone, suggests that additional interventions
238 may be needed. Identifying, understanding and alleviating psychosocial stressors may be important
239 as they may be precipitants or causes of MUVL. In addition, establishing rates of psychiatric
240 comorbidity (for example anxiety, depression etc) in these children will improve understanding of
241 mechanisms and identify additional treatment targets.

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354

355 **Titles and legends to figures**

356

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

358 diagnosed with MUVL.

359

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

361 months.