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Ocular Myositis

Congenital Fibrosis of the Extraocular Muscles

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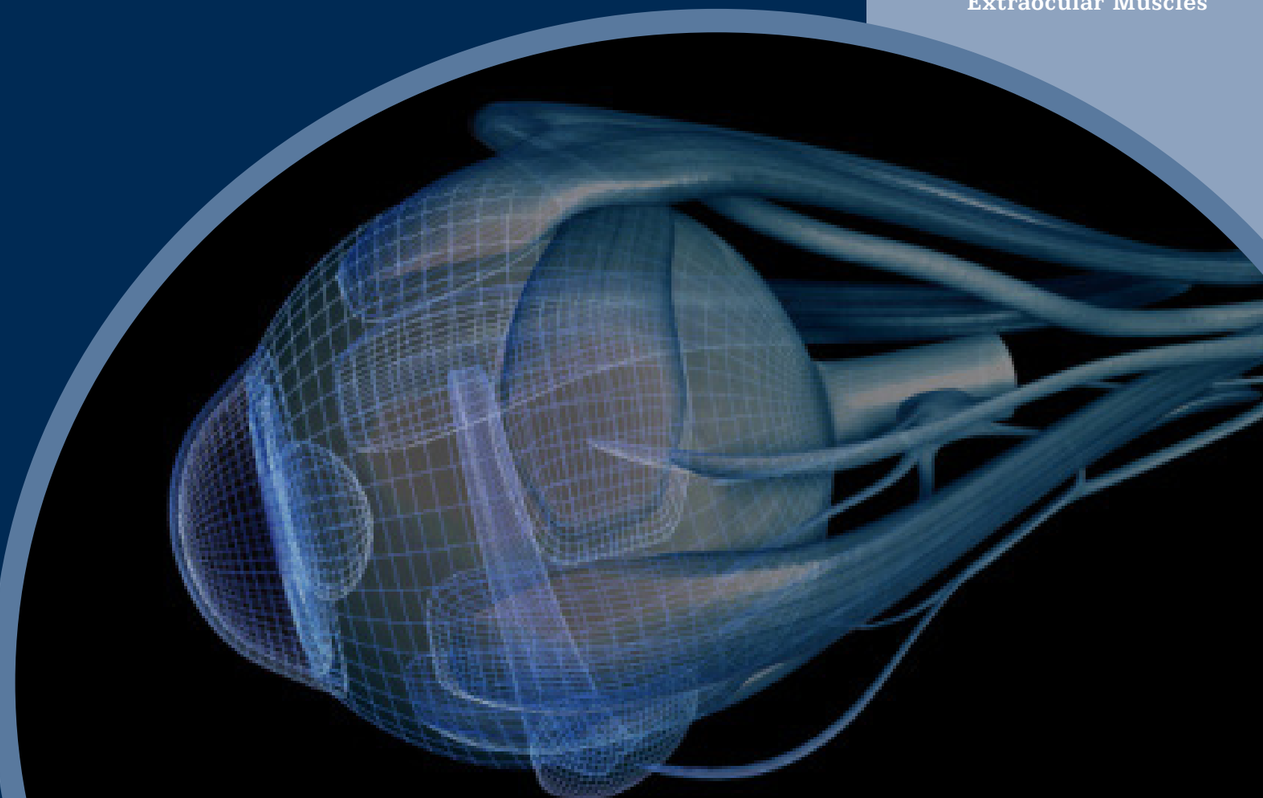

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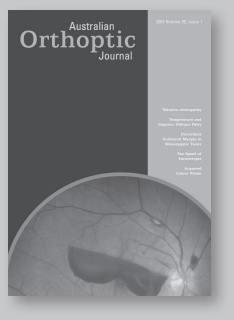

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Ocular Complications of Mucopolysaccharidoses

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ABSTRACT

Purpose: To study the extent of ocular involvement among children with mucopolysaccharidoses (MPS) at The Children's Hospital at Westmead, Sydney, Australia.

Methods: This study consists of a retrospective consecutive case series, with review of medical records of children with confirmed diagnosis of MPS from 1997 to 2009.

Results: Forty-five children had MPS but only 29 had a record of previous formal ocular assessment. Of these, more than half had documented ocular involvement, including corneal clouding, common among the MPS I subtypes and MPS VI (Maroteaux-Lamy) patients. Posterior segment changes, including pigmentary retinopathy, epiretinal membranes and optic disc changes were more common in MPS II (Hunter). Two children with MPS VI were also noted to have epiretinal membranes and this is likely to be a previously unrecognised association of MPS VI. Only 7 out of 18 children with MPS III (Sanfilippo) were examined, and clinically none were found to have retinopathy. Among those who were cooperative for vision assessment, four were found to see 6/12 or better, while the majority had best corrected vision between 6/15 and 6/60. Three patients had documented disease progression leading to blindness. All four MPS VI patients receiving enzyme replacement therapy (ERT) had stable visual acuity with no ocular progression (6.5 years mean follow-up). However progression of corneal clouding was noted in the only MPS I patient receiving ERT.

Conclusion: Ocular involvement in MPS may cause significant vision impairment. Formal ophthalmic review is important for early detection and treatment to help achieve the best visual outcome.

Keywords: mucopolysaccharidoses, Hurler, Scheie, Hunter, Sanfilippo

INTRODUCTION

The mucopolysaccharidoses (MPS) are a group of inborn errors of metabolism resulting from a deficiency of specific lysosomal enzymes necessary to break down complex carbohydrates called glycosaminoglycans (GAG). As a result glycosaminoglycans and other substances including glycosphingolipids, accumulate within various tissues and organs of the body. The most common MPS disorders are Type I (encompassing the severe Hurler and the attenuated Hurler-Scheie and Scheie spectrum), Type II (Hunter), Type III (Sanfilippo), Type IV (Morquio) and Type VI (Maroteaux-Lamy). Affected patients typically have coarse facial features, skeletal dysplasia, joint contractures and hepatosplenomegaly. Some will have significant cardiac and respiratory disease, intellectual impairment and neurological involvement. Ocular deposition leads to corneal clouding, glaucoma, pigmentary retinopathy and optic nerve involvement. These findings can result in significant vision impairment.[1]

There have been recent advances in the treatment of MPS. Although bone marrow transplantation (BMT) has known mortality and morbidity, in its current regimen as haematopoietic stem cell transplantation (HSCT) it is an increasingly effective treatment for some MPS disorders. It is a particularly effective treatment for children with MPS I (Hurler syndrome) transplanted less than two years of age. Successful engraftment replaces the relevant deficient enzyme, allowing for biochemical and clinical improvement and increased lifespan.[2,3] More recently, enzyme-replacement therapy (ERT) has been shown to decrease the level of urinary GAG, decrease hepatosplenomegaly and increase joint mobility.[4,5] Combined ERT/HSCT have been reported as efficacious therapy for young patients with MPS I (Hurler) with much reduced mortality and morbidity[6] and is now the treatment of choice. ERT alone is now available for MPS I (Hurler-Scheie), MPS II (non-neurological) and MPS VI of intermediate or milder type. Whether or not these treatment modalities alter long-term ocular progression remains uncertain. The full extent of ocular and visual morbidity from MPS disorders is not well documented. The

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objective of this study was to review the extent of ocular complications among children diagnosed and treated for MPS at a tertiary paediatric hospital.

METHODS

We conducted a retrospective review of the medical records of children diagnosed with MPS who received treatment at The Children's Hospital from July 1997 to June 2009. Diagnosis of the MPS subtypes was confirmed through their clinical phenotypes and biochemical assays. The children who underwent formal ophthalmic assessment within the study period were included. All ocular findings and conditions noted in the medical records were documented including visual acuity, presence of corneal clouding, glaucoma, pigmentary retinopathy and optic nerve changes. Progression of ocular complications during the course of follow-up and refractive status were also assessed. Visual outcome was determined by the best corrected visual acuity in the better eye (if unequal) at the most recent eye review, to reflect the child's functional vision. Institutional Ethics Committee approval was obtained for this study.

RESULTS

A total of 45 children (ranging from 0 - 18 years) were diagnosed with MPS during the 12-year study period, with MPS III being the most common (40%) followed by MPS I (24.5%), MPS II (24.5%), MPS VI (9%) and MPS IV (2%). Only twenty-nine children (64%) received formal ophthalmic review, of whom 18 (62%) children had significant ocular involvement. Table 1 summarises the number of children with each type of MPS and their ocular findings.

Table 1. Distribution of MPS patients and their ocular complications

MPS types N (%)	Those who underwent ocular examination	Those with ocular involvement	Corneal clouding	Glaucoma	Pigmentary retinopathy	Optic nerve involvement	Epiretinal membrane
MPS I 11 (24.5%)	8	8	8	0	2	2	0
MPS II 11 (24.5%)	9	5	0	0	2	3	2
MPS III 18 (40%)	7	0	0	0	0	0	0
MPS IV 1 (2%)	1	1	1	0	0	0	0
MPS VI 4 (9%)	4	4	4	1	0	1	2

Ocular findings in MPS I

Eight out of eleven children with MPS I sub-types underwent ophthalmic review. All of these children had significant ocular involvement with corneal clouding, which varied in severity according to the subtype present, being most advanced in the Hurler phenotype and least advanced in the Scheie phenotype. Pigmentary retinopathy was noted in two patients with one confirmed by an abnormal electroretinogram (ERG). One child had papilloedema whereas another child had marked elevation of the optic discs without evidence of raised intracranial pressure (pseudopapilloedema).

Ocular findings in MPS II

Five out of the nine MPS II children reviewed had significant ocular involvement. Posterior segment involvement was more commonly seen in the children with MPS II compared to other types of MPS. Pigmentary retinopathy and epiretinal membranes were found in two children, papilloedema in one and optic atrophy in two. In one boy the optic atrophy was associated with marked thickening of the posterior sclera on ocular ultrasound. No MPS II children were diagnosed with corneal clouding, as expected.

Ocular findings in MPS III

Eighteen children were diagnosed with MPS III, however, only seven underwent formal ophthalmic examination. None of them were known to have night blindness or were diagnosed with ocular involvement including the anticipated pigmentary retinopathy.[1] Unfortunately, none of these children underwent ERG.

Ocular findings in MPS IV

One child was diagnosed with MPS IV and was noted to have mild corneal clouding which did not significantly affect vision. Fundoscopy did not show any evidence of retinopathy or optic disc changes.

Ocular findings in MPS VI

All four children diagnosed with MPS VI underwent ophthalmic examination. All had corneal clouding of varying degrees, with one severe enough to warrant a corneal graft. None of the MPS VI children had pigmentary retinopathy; this was supported by a normal ERG in one child. Papilloedema was noted in one child. Interestingly, epiretinal membranes were also found in two children with MPS VI. One patient was documented to have glaucoma,

with intraocular pressures medically controlled by topical anti-glaucoma medication.

Visual acuity status

Visual acuity was assessed in 20 children using an age and intellectually appropriate vision test, including Teller Acuity Cards, Kay Picture test or the Snellen chart. Four children had a visual acuity of 6/12 or better while the majority (11 children) had vision between 6/15 and 6/60, and five had vision less than 6/60. Analysis of visual acuity over time showed most children's vision remained relatively stable. Unfortunately, three children (two with MPS I, one with MPS II) showed significant visual deterioration leading to severe vision impairment (defined as visual acuity worse than 6/60) during the study period. One MPS I child had severe hydrocephalus and optic atrophy associated with no perception of light in one eye and perception of light only in the fellow eye. In the other MPS I child the cause of the severely impaired vision was progression of corneal clouding. The MPS II child suffered from hydrocephalus with resulting optic atrophy and optic nerve compression from posterior scleral thickening around the optic nerve.

Refractive status

Cycloplegic refraction was successfully performed in 20 children. Seven had normal refraction for their age, while the remaining thirteen had refractive errors. Eight children had significant hypermetropia, three had myopia and two had myopic astigmatism. Glasses were prescribed for four children in whom the refractive error was considered to be the main cause of reduced vision.

Bone marrow transplant, enzyme replacement therapy and ocular findings

Two MPS I children underwent BMT, with one child receiving ERT prior to BMT. Both had no documented progression of their ocular findings over a mean follow-up period of 6.5 years. Vision remained stable (6/12 equivalent) in one child and stable at 6/75 in the other due to pigmentary retinopathy. Neither child developed severe dry eye (keratoconjunctivitis sicca) or cataract as a consequence of BMT. However, one child with MPS I (Hurler-Scheie subtype) experienced a deterioration of vision due to the progression of corneal clouding despite ERT (5 years follow-up). Four children with MPS VI received ERT and had stable vision and ocular findings whilst undergoing therapy, with a mean follow-up period of 6.5 years.

DISCUSSION

Ocular findings in MPS are frequent and may lead to significant vision impairment due to corneal clouding, glaucoma, pigmentary retinopathy and optic nerve involvement. Similar to previous studies, corneal clouding in this study was more common in the MPS I subtypes and MPS VI[1]; no children with MPS II or MPS III in this study showed corneal clouding. A favourable prognosis for corneal transplant has been previously documented in MPS disorders, including MPS VI.[7,8,9,10,11] In our study, the one child who underwent corneal transplant for severe corneal clouding had MPS VI. The donor cornea remained clear for a three-year follow-up period with best corrected vision of 6/20.

Glaucoma in MPS results from GAG accumulation within the anterior segment structures of the eye, including the trabecular meshwork, causes narrowing of the drainage angle and obstruction of the aqueous outflow.[1] In a study of 121 patients with MPS VI (estimated to be about 10% of MPS VI patients globally) 10% were on anti-glaucoma treatment.[12] In our study, the only child diagnosed with glaucoma also had MPS VI. However, the prevalence of glaucoma in our study could have been underestimated due to the complex nature of examining for glaucoma in a cohort of MPS children who often had intellectual impairment and ocular complications such as corneal clouding which prevented a clear view of the relevant ocular structures.

Retinopathy was noted in two children with MPS I and two children with MPS II. Previous studies have documented moderate to severe pigmentary retinal degeneration associated with ERG abnormalities as a prominent feature of MPS III.[1,13] In our study, no MPS III children were found to have any ocular complications. However, the onset of pigmentary retinal degeneration may be delayed until adolescence and our subjects were too young for this to be detected. Furthermore ERG had not been performed to detect the presence of retinal changes. Retinopathy was also not found in any of the children with MPS IV or MPS VI. This was confirmed in one individual with MPS V1 who had a normal ERG. Retinopathy has not been described among MPS VI children in the literature apart from one child who had reduced dark-adapted amplitude on ERG.[1,9]

Four children (two with MPS II and two with MPS VI) were found to have epiretinal membranes. This layer of tissue overlying the macula has been previously reported in MPS II[14] and Type III and IV mucolipidoses.[15,16] To our knowledge, epiretinal membranes have not been previously reported in MPS VI and this finding is suggestive of a new ocular association in this MPS subgroup. Whether or not these membranes represent deposition of GAG within the retinal layers is unclear.

In our series six children had optic nerve involvement including papilloedema, elevation of the optic nerves due to GAG deposition, and optic atrophy. Three of these children had MPS II, and optic nerve involvement in MPS II has been well documented.[17,18] This occurs as a consequence of raised intracranial pressure or direct GAG accumulation within the optic nerve and surrounding

meninges compressing the optic nerve. Increased scleral thickness, especially posteriorly, appears to be a common association in MPS II and no doubt contributes to the compression of the optic nerve at its exit from the eye.[18,19]

Bone marrow transplantation (BMT) has been reported to reduce corneal clouding,[20,21] to contribute to resolution of optic nerve swelling[8,20] and improvement in the retinal function as measured by ERG in MPS I.[20] In a study of 23 patients (19 with MPS I)[8] it was shown that a reduction of corneal clouding was seen in approximately one-third of patients, while a similar number showed worsening of corneal clouding (mean follow-up period of 6.1 years). Also in this study, approximately 80% of patients showed initial ERG improvement within one year of BMT followed by a decline after this time.[8] Interestingly, 30% of patients had papilloedema and raised intracranial pressure prior to BMT, which resolved following BMT.[8] No ocular improvement occurred in our two MPS I children following BMT although their visual acuity, refractive status and ocular findings remained stable. In one child however, significant corneal clouding precluded any view of the optic discs.

ERT has now become available for the treatment of MPS I, MPS II and MPS VI. Kakkis and colleagues found no improvement in corneal clouding over the course of a year despite the normalisation of liver size and near normalisation of urinary GAG and improvement in other systemic symptoms in eight MPS I children who received ERT.[4] Similar findings were also found among MPS I patients receiving ERT in another study with vision stable in five out of eight patients over a four-year follow-up period and vision deteriorating in three patients, two due to progressive corneal clouding.[10] In a more recent study of seven patients with MPS VI, ocular findings remained stable in five patients with no substantial change in corneal clouding or any improvement in the optic nerve pathology (mean follow-up period of 44 months).[11]

The MPS I child in our study who received ERT had progressive corneal clouding. Ocular findings remained stable throughout the course of follow-up in all our four MPS VI patients who received ERT (mean follow-up of 6.5 years). There was no evidence of improvement in visual acuity or reduction of the corneal clouding.

This study was a retrospective analysis, and shortcomings are acknowledged. At times medical record documentation was incomplete and variability of visual assessment techniques made analysis difficult. Ophthalmic investigation of MPS children was challenging and limited by the nature of their capabilities. Tests such as ocular ultrasound and electroretinogram were often not conducted which hindered the detection of compressive optic neuropathy and pigmentary retinopathy respectively.

CONCLUSION

Ocular involvement is common in MPS and in some individuals it may be severe enough to lead to significant vision impairment. This study has revealed that MPS children may not necessarily access regular ophthalmic care. To ensure MPS sufferers enjoy the best possible vision, formal ophthalmic review for the early detection of treatable ocular conditions is vital. Health professionals need to be aware of the likelihood of ocular involvement in MPS.

In view of the findings of this study, we propose that MPS children undergo comprehensive ophthalmic review on a yearly basis, to detect potentially treatable causes of vision impairment. Children with more severe ocular manifestations and progression should be seen more frequently. A prospective study at our centre has recently commenced and will gather detailed data on the ocular manifestations, progression and visual outcome of children diagnosed with MPS. The findings of this study will be used to establish guidelines regarding frequency of ophthalmic review and efficiencies in assessment of children with MPS.

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Juvenile Idiopathic Arthritis and Uveitis in a Paediatric Sydney Population

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ABSTRACT

Juvenile idiopathic arthritis (JIA) is an inflammatory condition that affects 1 in 1,000 children in Australia. JIA can be defined by inflammation in one or more joints for a period of at least six weeks, with an onset younger than 16 years of age. JIA is sub-classified into different types depending on the number of joints affected, the rheumatoid factor and whether other systemic conditions are present.

JIA can be associated with uveitis, a serious and chronic ocular complication which is often difficult to manage and can result in visual loss. The risk of development of uveitis differs dependent on the type of JIA present. An ophthalmology assessment forms a vital part of the assessment for children with JIA.

The aim of this study was to ascertain the prevalence of visual complications associated with children who have a diagnosis of JIA. A retrospective review of children presenting to the Eye Clinic at The Children's Hospital Westmead with JIA over a twelve-month period between 2009 and 2010 was performed. This paper emphasises the need for ophthalmology review in this cohort of children.

Keywords: juvenile idiopathic arthritis, uveitis, paediatric

INTRODUCTION

Juvenile idiopathic arthritis (JIA) is defined as idiopathic arthritis of greater than six weeks duration with onset before sixteen years of age. It is a chronic inflammatory joint disease and is the most common rheumatic disease in children and adolescents. The incidence of JIA is 10 in 100,000 children worldwide.[1] In Australia at least 5,000 children are affected by JIA at any one time[2] with an incidence of between 1 and 4 cases per 1,000 children.[3] The cause of JIA is currently unknown.

The International League of Associations for Rheumatology (ILAR) sub-classifies JIA into different categories depending on the number of joints involved and associated systemic conditions.

- Persistent or extended oligoarticular arthritis is the most common type of JIA and is defined by the involvement of up to four joints at the onset of the disease.
- Rheumatoid factor positive and rheumatoid factor negative polyarthritis occurs when five or more joints are affected.
- Systemic arthritis is a chronic arthritis, associated with systemic features.
- Enthesitis-related arthritis (previously known as juvenile spondyloarthropathy) is a chronic arthritis associated with enthesitis, or with lower axial skeletal involvement.
- Psoriatic arthritis is a chronic arthritis usually with asymmetrical involvement of small and large joints and evidence of psoriasis or a psoriatic diathesis.
- Undifferentiated arthritis which is arthritis that fulfils criteria in no category or in two or more categories.

The presence of antinuclear antibodies (ANA) is common in inflammatory disease and is detected on a blood sample. JIA may be diagnosed with or without detection of an increased level of ANA (ANA positive or negative respectively).

Symptoms of JIA include swelling of the affected joints, commonly the knee, ankle and wrist, along with pain, joint stiffness and possible joint contracture and joint damage. Extra-articular features are common, such as fever, rash, pleuritis, pericarditis, lymphadenopathy and hepatosplenomegaly. Ocular inflammation and uveitis are the most common extra-articular manifestations. Uveitis occurs when inflammation arising from the iris, ciliary body or choroid is present. It is a serious and chronic condition which is often difficult to manage and can result in severe visual loss and other ocular complications such as glaucoma, cataract, hypotony, cystoid macula oedema, band keratopathy, and amblyopia.[4]

The incidence of JIA-related uveitis is 1 in 100,000 worldwide and it accounts for 80% of all childhood uveitis.[5]

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The severity of uveitis in children with JIA is vast. The risk factors for developing uveitis include an early diagnosis of JIA, female, oligoarticular form and ANA positive. Children developing arthritis below the age of three years are at risk for up to seven years. Children who develop arthritis after the age of six are at risk for up to three years.[1] Chronic anterior uveitis is most commonly associated with JIA. Approximately 20% to 30% of children with JIA and uveitis will have their vision significantly affected.[6]

Clinically, the ocular signs and symptoms associated with JIA-related uveitis can include redness, pain, blurring of vision, and photophobia. It is important to mention that although redness can be a clinical feature of uveitis, the eye is often white, with no obvious sign of inflammation. Some children with mild disease can be asymptomatic. Uveitis can at times be the first sign of JIA.[5]

The severity of the uveitis present and treatment methods used will determine the risk of developing other ocular complications. The use of topical and systemic corticosteroids in the management of this disease can induce adverse ocular effects such as cataract and glaucoma. Surgical intervention is often necessary in these cases. These factors determine the visual prognosis of the patient, which is often poor and can result in irreversible blindness. Children diagnosed with JIA will require regular rheumatology reviews and ophthalmology screening. If uveitis associated with JIA is detected, the frequency of ophthalmology reviews will increase and may be as regular as fortnightly for a duration of years.

METHODS

A retrospective analysis of the medical records of patients with a diagnosis of JIA, seen in the eye clinic at The Children's Hospital at Westmead over a twelve-month period, between June 2009 and June 2010 was performed. Ethical approval was obtained for this study.

The data retrieved from medical records included age, gender, presenting symptoms, diagnosis and sub-classification of JIA, age at onset of JIA, presence of antinuclear antibodies, presence and classification of uveitis, age at onset of uveitis, secondary ocular complications and final visual acuity.

Children presenting with uveitis without a diagnosis of JIA were excluded from the study.

RESULTS

A total of 57 patient files were included in the study, with the characteristics presented in Table 1. JIA was more prevalent in females than males with 65% of patients reviewed being female.

Table 1. Characteristics of children with juvenile idiopathic arthritis

	All JIA patients (N = 57)	Patients with JIA and uveitis (N = 20)	Patients with JIA without uveitis (N = 37)
Patients, %	100	35.7	64.3
Female Number (%) {% of all female patients}	37 (64.9)	8 (40) {21.6}	29 (78.4) {78.4}
Age at diagnosis of JIA (years) Mean (range)	3.9 (1-13)	4.7 (1-13)	3.5 (1-10)
Age at diagnosis of uveitis (years) Mean (range)		5 (3-13)	N/A
ANA positive* Number (%)	39 (68)	18 (90)	21 (57)

* Five patients did not have an ANA analysis available

Oligoarticular JIA was the most prevalent subtype, being identified in 30 patients (53%). Polyarticular JIA was present in seven patients (12%), systemic JIA in four patients (7%) and one patient reviewed had psoriatic JIA (2%). Fifteen patients (26%) were identified as having undifferentiated arthritis or nonspecific JIA, and no patients reviewed were diagnosed with enthesitis-related arthritis (Figure 1).

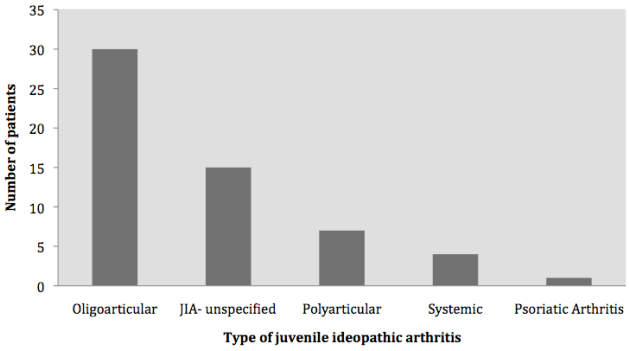

Figure 1. The distribution of the sub-classifications of JIA (N = 57).

A positive ANA factor was prevalent across the group. Thirty-nine patients (68%) were identified as ANA positive and 18 patients (32%) ANA negative. The age of patients at onset of JIA ranged from 12 months to 13 years with a mean of 47 months (SD ±32.06), showing no apparent pattern in the age of diagnosis (Figure 2).

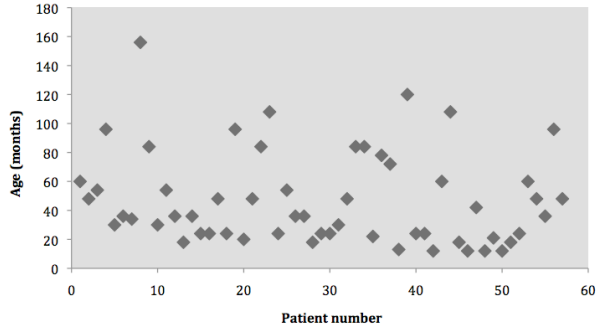

Figure 2. The age of patients at the time of diagnosis of JIA (N = 57).

The presence of uveitis was reviewed. Twenty patients (36%) had uveitis, while 37 patients (64%) had not developed uveitis at the time of the review. Bilateral uveitis was most prevalent and identified in 85% (n = 17) of patients while only 15% (n = 3) had unilateral disease. The age of onset of uveitis ranged from 3 years to 13 years with a mean of 65 months (SD ±27.39). Fifteen patients (75%) had an onset of uveitis before 7 years of age. Two patients reviewed with uveitis did not have an age of onset documented (Figure 3, Table 1).

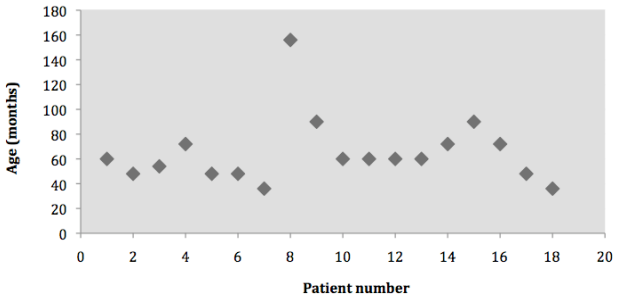

Figure 3. The age of patients at the time of diagnosis of uveitis (N = 18).

The type of uveitis present was reviewed. Anterior uveitis was most common, and was detected in 18 patients (90%) and panuveitis was identified in two patients (10%). No cases of intermediate uveitis were identified.

Patients with oligoarticular JIA had a high incidence of ANA positivity (76%, n = 23) and uveitis was present in 23% (n = 7). Unclassified JIA also had a high incidence of ANA positivity at 60% (n = 9) and uveitis at 67% (n = 10) (Table 2).

Table 2. Relationship of ANA factor and uveitis to type of JIA

JIA sub-classification	Number of patients	ANA positive N (%)	Patients with uveitis N (%)
All subgroups	57	39 (68.4)	20 (35.1)
Oligoarticular	30	23 (76.7)	7 (23.3)
Polyarthritis	7	3 (42.9)	1 (14.2)
Systemic	4	1 (25.0)	2 (50)
Psoriatic	1	1 (100)	0
Enthesitis-related	0	0	0
Unclassified	15	9 (60.0)	10 (66.7)

The initial symptoms of uveitis varied across the group. Interestingly, eight patients (40%) were asymptomatic. Ocular symptoms included red eyes 35% (n = 7), photophobia 15% (n = 3) and reduced vision 5% (n = 1). In one patient the initial symptoms were not documented as this patient had been transferred from another eye care centre (Figure 4).

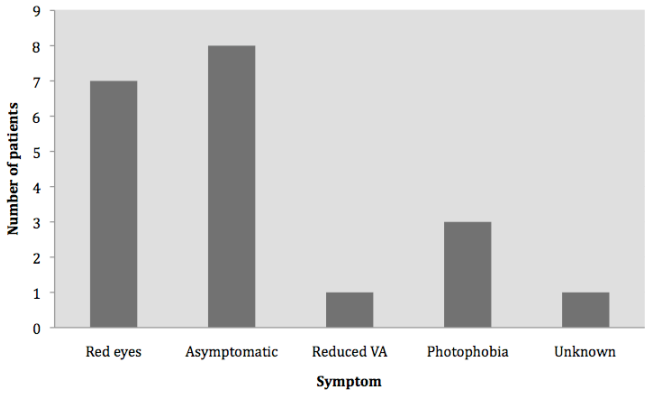

Figure 4. The distribution of the initial presenting symptoms of uveitis (N = 20).

Visual outcome in patients with JIA and uveitis ranged from 6/4.5 to no light perception. Of the 37 eyes with uveitis 62% (n = 23) had visual acuity of 6/9.5 or better, 11% (n = 4) had visual acuity of 6/12 to 6/18, 11% (n = 4) had visual acuity of 6/18 to 6/60 and 16% (n = 6) had visual acuity worse than 6/60.

Associated ocular complications in these patients were diverse. Commonly, glaucoma (60%, n = 12) and cataracts (40%, n = 8) were identified. Other complications identified in the group were band keratopathy (15%, n = 3), and anterior or posterior synechiae (15%, n = 3). 50% (n = 10) of these patients with ocular complications required surgical intervention.

Of the patients reviewed, six had a diagnosis of uveitis that preceded a diagnosis of JIA. For these patients, the visual outcome was poorer than those initially diagnosed with JIA who then went on to develop associated uveitis. Two of patients initially diagnosed with uveitis had a final visual acuity of no light perception in at least one eye, one had 6/18 or worse vision and three had a visual acuity of 6/9.5 or better. Five had an associated ocular complication, which included glaucoma (n = 4), cataract (n = 3), posterior or anterior synechiae (n = 2), and band keratopathy (n = 1). Surgical intervention was required in four patients with associated ocular complications required surgical intervention.

DISCUSSION

Early presence of uveitis in JIA is an important prognostic factor associated with adverse visual outcomes.[7] Early detection and treatment of uveitis is mandatory in enabling the best possible visual outcome for these patients. Regular follow-up and review for the duration of their childhood, and sometimes into the adult years, is required due to the chronic nature of the condition. While not all children with uveitis will go on to develop JIA, it is important that it is investigated throughout the course of the disease.

Uveitis can be the initial presentation of JIA. Children with signs of uveitis preceding signs of arthritis may have a poorer visual outcome.[8] Patients reviewed in this study with a diagnosis of uveitis prior to a diagnosis of JIA showed a poorer visual outcome and majority of these patients had an associated ocular complication, such as glaucoma or cataract. A poorer visual outcome in these patients may be a result of later presentation to an ophthalmologist, and therefore further progression of the disease.

Symptoms of uveitis may be nonspecific, such as intermittent red eyes, epiphora and reduced vision. Some children may even be asymptomatic. Interestingly, in this study 40% of patients reviewed were asymptomatic. It is imperative that all children with idiopathic uveitis are screened for JIA and early treatment is commenced to achieve the best visual outcome.

The type of subclassification of JIA and the ANA factor determine the risk of development of uveitis. Oligoarticular and unclassified JIA with an ANA positive factor are at high risk for the development of uveitis. In this study 23% of patients with oligoarticular JIA were ANA positive and had a diagnosis of uveitis, and 67% of patients with unclassified JIA were ANA positive and had a diagnosis of uveitis.

The exact visual prognosis of children with JIA is not known. Rates of visual impairment ranging from 6% to 25% have been published for JIA-associated uveitis.[9,10] In this study visual acuity ranged from 6/4.5 to no light perception. This emphasises the importance of regular eye reviews for patients with JIA and the significance of comorbidity in children with JIA. All patients included in this study required frequent ophthalmic review over the twelve-month period. Review periods included fortnightly, monthly, three-monthly, six-monthly and yearly. The frequency of review depended upon the type of JIA, level of uveitis and current ocular associations and varied for patients throughout the course of their disease. The Royal College of Ophthalmologists and the British Paediatric Association have compiled a summary of recommendations regarding the frequency of eye review for these patients. The recommended schedule ranges from three to twelve months depending on whether the patient is considered high or low risk.[11]

The disease course of JIA is prolonged and can continue into adulthood. Although JIA becomes less inflammatory with age, it has been reported that up to 50% of adults who suffered JIA in childhood will continue to experience the effects of the disease, such as joint deformity, growth abnormalities, osteoporosis, pain and difficulties with daily living.[12] They may also suffer a visual impairment as a result of uveitis, or the secondary complications of cataract and glaucoma. The transition from paediatric health services to appropriate adult health services must be considered for these patients.

CONCLUSION

Juvenile idiopathic arthritis is a serious and chronic condition that has a known association with uveitis. It is more common in females than males. The type of sub-classification of JIA and the ANA factor determine the risk of development of uveitis with oligoarticular and unclassified subgroups with ANA positivity being the highest risk factor.

Uveitis is an ocular condition that can be difficult to manage and results in visual loss and other ocular complications such as glaucoma and cataract, which often require surgical intervention. Visual prognosis in children with JIA is influenced by the age at which uveitis is detected. Children with a diagnosis of uveitis preceding a diagnosis of JIA have a poorer visual outcome.

All children who are diagnosed with JIA will require a full ophthalmology screening as well as regular eye reviews. Similarly, all children who present with uveitis will require a full rheumatology assessment as uveitis may be the first sign of juvenile idiopathic arthritis.

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Ocular Myositis: A Case Study

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ABSTRACT

Orbital myositis is an uncommon inflammatory condition resulting in variable degrees of restriction of the extraocular muscles. A case of a 15-year-old girl is presented, highlighting the importance of differential diagnosis from other ocular conditions that can cause extraocular muscle restrictions.

Keywords: orbital myositis, extraocular muscles, inflammation, restriction, diplopia

INTRODUCTION

Ocular myositis is an idiopathic inflammatory condition in which orbital inflammation is confined to the extraocular muscles and occurs in the absence of inflammation of other orbital or peri-orbital tissues.[1,2] It is classified as one of the patterns of clinical presentation of orbital pseudotumour, a condition distinguished by inflammation of any orbital soft tissue, including orbital fat, lacrimal gland and connective tissue.[3]

The condition is characterised by an acute onset of orbital pain, often exacerbated by eye movement, diplopia, proptosis, duction restrictions, ptosis and conjunctival injection. Visual acuity and optic nerve function remain intact in the presence of the extraocular muscle inflammation.[4,5]

Cases of ocular myositis may be acute or chronic in presentation. Acute or isolated cases of ocular myositis are those that present with a recent onset of symptoms, normally less than two weeks, including pain and/or diplopia. The more chronic cases of orbital myositis include episodes that continue for a period of more than two months, or recurrent acute episodes which can lead to long term extraocular muscle restriction. Atypical cases of orbital myositis have also been reported.[6] These cases include those with uncharacteristic presentations including lack of pain or optic nerve dysfunction.

The cause of ocular myositis at this stage is unknown but it is hypothesised that an immune-mediated process may be involved following reports of associations between systemic conditions (sarcoidosis and Crohn's disease) and the development of ocular myositis.[5,6]

This paper will present a case study of a 15-year-old girl diagnosed with chronic orbital myositis.

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CASE REPORT

In January 2010 a 15-year-old girl, Miss J, attended the Sydney Children's Hospital eye clinic following a three-week history of variable right ptosis and diplopia on down gaze. These symptoms were accompanied by a right-sided temporal headache and pain behind her right eye that had been present for the previous three months, with minimum relief from paracetamol.

Miss J had a history of similar symptoms with the exception of diplopia on two previous occasions. These episodes were investigated by an ophthalmologist and paediatrician respectively and no abnormality found on examination or on her computed tomography (CT) or medical resonance imaging (MRI) scans.

Interestingly, there was a strong family history of conditions affecting the extraocular muscles. Miss J's maternal aunt was diagnosed with myasthenia gravis and four male members of her family (both immediate and extended) were diagnosed with ocular myositis.

On initial observation Miss J displayed a right partial ptosis, which was confirmed by measurement of the palpebral aperture with the right being 7 mm and the left being 14 mm. Her visual acuity without glasses was right 6/6 and left 6/5. On cover test at 6 metres she demonstrated orthophoria. Cover test at 1/3 metre revealed a small

exophoria with an accommodative target. Cover test at 1/3 metre repeated with a non-accommodative target (a torch) showed decompensation of the latent deviation to a small right exotropia with a small right hypertropia. On measurement, this deviation was neutralised with 4 prism dioptres base-in with the vertical measurement varying between 14 and 18 prism dioptres base-down, fixing left, restoring binocularity. Binocularity was tested using the Lang Stereotest, on which Miss J demonstrated a positive response achieving 550 seconds of arc.

On testing ocular movements, there was marked restriction of movement of the right eye in most positions of gaze. There were -4 underactions of the right superior rectus, inferior rectus and superior oblique. There were -3 underactions of the right inferior oblique and a mild underaction of the right medial rectus of -1.

The patient was referred to the immunology and neurology departments for multidisciplinary review. All investigations by immunology and neurology departments were found to be unremarkable. She was also sent for CT of the orbits and CT angiogram to rule out possible cerebral aneurysm. CT of the orbits revealed marked enlargement of the right superior and medial rectus muscles with involvement of the myotendinous junctions.

Miss J was treated promptly with a high dose of oral prednisone, with 50 mg for three days, tapered to 25 mg for a further three days. A week after high-dose steroid treatment saw an improvement in the patient's signs and symptoms, with almost complete resolution of diplopia and minimal extraocular muscle restriction remaining two months later.

Over the following year, the patient's condition improved but did not completely resolve. She had two episodes of relapse in which there was deterioration and changes in the affected extraocular muscles. Following each episode, maximum improvement in signs and symptoms occurred three to four weeks after commencing treatment. Each recurrence of ocular myositis coincided with reductions in the patient's steroid dosage below 5 mg. As a result of the patient's condition deteriorating with reductions in steroid dose, she has remained on a constant low-dose of steroids since initial onset of her condition, with increases in dosage when she has a replapse episode. Due to the long-term use of steroids the patient commenced treatment with methotrexate (a steroid-sparing agent) to help reduce the side effects of long-term steroid use.

DISCUSSION

Ocular myositis is a distinct clinical entity however extraocular muscle enlargement is a clinical feature also seen in other conditions including thryoid orbitopathy, carotid cavernous fistulas, metastases and infiltrative conditions.[6] Careful investigation of clinical characteristics is necessary for correct differential diagnosis to be made. Modern ultrasound and radiological techniques, allow the differential diagnosis of enlargement of the extraocular muscles to be promptly narrowed to ocular myositis and thyroid orbitopathy.

Thyroid orbitopathy is reported as the most common cause of enlargement of the extraocular muscles.[5] For cases of non-thryoid-related extraocular muscle enlargement, inflammation (classified as idiopathic orbital inflammatory disease or orbital pseudotumour) has been reported as one of the most common causes of extraocular muscle enlargement,[5,6,7] followed by vascular and neoplastic causes.[6,7]

The presenting signs and symptoms of thyroid eye disease and ocular myositis differ despite both having characteristic enlargement of the extraocular muscles. Patients with thyroid orbitopathy often present with a gradual onset of bilateral problems, often asymmetric, including dry eye, irritation, proptosis, and diplopia. Patients with ocular myositis will present with a more acute onset of symptoms, often unilateral, including pain on or exacerbated by eye movement, swelling and diplopia. Characteristically, in ocular myositis inflammation is isolated to extraocular muscles, whilst in thyroid orbitopathy a characteristic increase in orbital fat, causing exophthalmos, occurs in conjunction with enlarged extraocular muscles.

Diagnostic imaging (computed tomography) allows for differentiation between the two conditions as there is a distinct difference in the pattern of extraocular muscle enlargement and muscle involvement. In thyroid orbitopathy, bilateral asymmetric multiple muscle involvement is observed with regular muscle enlargement confined to the muscle belly, sparing the tendinous insertions. The inferior rectus is the muscle most frequently involved, followed by the medial, superior and lateral recti muscles.[2,7] In contrast, the most common presentation of ocular myositis is unilateral with only a single muscle affected.[6] Ocular myositis can also present with multiple muscle involvement and it has been suggested that multiple muscle involvement at initial presentation may be a risk factor for recurrent episodes or chronic cases of the condition.[6] The horizontal recti muscles tend to be most commonly involved in cases of ocular myositis,[2,4] with vertical recti muscles and obliques less commonly involved.[2,8,9] Miss J's case of chronic ocular myositis which initially presented with unilateral enlargement of the medial rectus and superior rectus of the right eye, agrees with the literature in demonstrating cases of ocular myositis that initially present with multiple muscle involvement can be associated with recurrent episodes or chronic ocular myositis.

Diagnostic imaging performed on Miss J also revealed inflammation of the myotendinous junction in addition to enlargement of the extraocular muscles. The pattern of

muscle enlargement in ocular myositis seen on imaging tends to be irregular often with inflammation of the tendinous insertion on the globe. Although the 'tendon sign' has been identified as a reliable indicator of ocular myositis, with 40-53% of cases by Mannor et al,[4] and 70% of cases by Zulfiqar et al[2] reporting tendon involvement, an absence of tendon involvement does not disclude a diagnosis of ocular myositis.

A pathognomonic sign indicative of ocular myositis is a rapid and dramatic improvement in signs and symptoms once treatment has commenced with high doses of systemic corticosteroids.[4,6,10] The most notable improvement in symptoms occurs within a period of 3 to 5 days,[1,9,10] after which the high steroid dosage is tapered. Similar to Miss J, the majority of patients experience a complete resolution of signs and symptoms approximately one month following initial onset. However, as seen in our case study, tapering the dosage of systemic steroids has been reported to coincide with recurrent episodes of myositis.[10] Long-term systemic steroid use in some patients becomes intolerable and other treatment options require consideration. These include treatment with steroid-sparing agents (or adjuvant drugs) such as methotrexate,[6] which was used as an adjunctive therapy for Miss J due to her long-term steroid use, or more radical treatment with radiation therapy.[3,11] It has also been reported that initial stages of treatment with non-steroidal anti-inflammatory drugs is also an option, however this treatment option tends to lend itself to patients with a non-recurrent or acute ocular myositis.[4]

Although it is suggested that ocular myositis is an immune-mediated process, familial influence on the development of this condition may also be important to consider. Our patient reported four of her male family members had been diagnosed with ocular myositis. At this stage, it seems only one other study has reported a family with multiple members demonstrating symptoms suggestive of ocular myositis.[12] Therefore it could be hypothesised that genetic predisposition may play a part in the development of the ocular myositis. However, it is important to consider equally that other events may influence the expression of the condition even in the presence of genetically predisposing factors.[12]

CONCLUSION

Ocular myositis is characterised by enlargement of extraocular muscles visible on medical imaging and rapid improvement of symptoms following treatment with high dose systemic corticosteroids. Although it is a distinct clinical entity, thorough clinical investigation is required for this condition to be differentially diagnosed from other causes of enlarged extraocular muscles including thyroid eye disease, vascular disorders and neoplastic disease.

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A Case Study: Management Options for a Patient with Congenital Fibrosis of the Extraocular Muscles

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ABSTRACT

Congenital fibrosis of the extraocular muscles is a relatively static congenital disorder leading to restrictive extraocular movements. The need for early intervention is vital to alleviate the development of an abnormal head posture and to lower the risk of amblyopia. A case of an 18-year-old male with congenital fibrosis, bilateral blepharoptosis, chin-up head posture, and external ophthalmoplegia is presented. His mother and older brother also exhibited similar clinical signs, thereby suggesting a familial pattern. Surgical management is discussed in light of the patient's presentation..

Keywords: congenital fibrosis of the extraocular muscles (CFEOM), bilateral ptosis, congenital external ophthalmoplegia, rotary nystagmus

INTRODUCTION

Congenital fibrosis of the extraocular muscles (CFEOM) is a rare non-progressive disorder characterised by ophthalmoplegia, bilateral blepharoptosis, abnormal head posture (AHP) and possible amblyopia.[1-9] CFEOM is broadly known as one of the congenital cranial dysinnervation disorders (CCDDs) due to its orbital innervational defects.[3,6,8,10] Muscle restriction is variable depending on the isolation or spread of fibrosis amongst healthy contractile muscle tissue. Earlier literature reported five clinical sub-classifications of CFEOM depending on the extent of fibrosis (Table 1).[11]

As the characteristics of the various forms of CFEOM may overlap, CFEOM is now categorised under three clinical phenotypes (Table 2).[3-4,7-9,12-14] The most common form of CFEOM is CFEOM1, with a 1/230,000 prevalence rate in the Western world.[3-4,14-15] CFEOM1 and CFEOM3 have been reported worldwide, however CFEOM2 has only been noted in people of Middle Eastern and Turkish descent.[3,14-16]

These genetic changes lead to an absence of the oculomotor and/or trochlear nucleus in the brainstem, agenesis of the superior division of the oculomotor nerve and motor neurons in the brainstem, a decrease in large motor axons and/or abnormal motor neurons, all of which result in atrophy and fibrosis of the extraocular muscles predominantly innervated by the oculomotor and trochlear nerves.[3-6,12-14,16-18]

Table 1. The five clinical sub-classifications of CFEOM[2,11]

General fibrosis syndrome (autosomal dominant > autosomal recessive > idiopathic)	Most severe form affecting all muscles bilaterally. Most severely the inferior recti and the levator palpebrae superioris.
Congenital fibrosis of inferior rectus with blepharoptosis (sporadic or familial)	Only the inferior rectus is affected. Mostly unilateral rather than bilateral. Ptosis, enophthalmos and unilateral fibrosis. Considered non-familial.
Strabismus fixus (sporadic)	Affects bilateral horizontal recti. Lateral rectus affected less than the medial rectus. Results in severe esotropia.
Vertical retraction syndrome	Bilateral vertical muscle restriction of superior and inferior recti. Most severely the superior recti, causing restriction of downgaze.
Congenital unilateral fibrosis with blepharoptosis and enophthalmos (sporadic)	All muscles are affected unilaterally, causing ptosis and enophthalmos.

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The levator palpebrae superioris and superior rectus muscles are particularly affected by atrophy due to agenesis of the superior division of the oculomotor nerve, thereby causing bilateral ptosis and hypotropia.[3,5-6,12,13,16,18]

Table 2. The three clinical phenotypes of CFEOM[2-4,6,10,12-16]	
CFEOM1	*Autosomal Dominant;* related to gene *KIF21A*/12q11-q12 of chromosome 12cen.
CFEOM2	*Autosomal recessive;* related to gene *PHOX2A/ ARIX* 11q13.2 of chromosome 11.
CFEOM3	*Classic CFEOM3;* related to gene 16q24.2-q24.3/*TUBB3* of chromosome 16. *Subtypes of CFEOM3;* related to gene *KIF21A*/12q11-q12 of chromosome 12cen.

CASE REPORT

An 18-year-old male presented to clinic for a pre-military ocular assessment, with signs of CFEOM. He had bilateral blepharoptosis (R>L), restrictive external ophthalmoplegia with the eyes fixed in infraduction, and a chin-up AHP markedly increased at distance. Bilateral eye movement restrictions with hypoglobus were similarly exhibited in his mother and older brother. In addition, he had fine manifest rotary nystagmus, with no history of surgery undertaken in the past and no associated learning barriers.

As he was unable to alternate fixation without moving his head, due to the restrictive ophthalmoplegia, the Krimsky test was performed instead of the prism cover test. This is because the Krimsky test relies on centring corneal reflections with a prism, either the non-fixing or fixing eye, while the prism cover test relies on the ability of the eyes to alternate fixation; something that he cannot do due to the restrictive external ophthalmoplegia.[19] The primary Krimsky method cannot be used in this instance as it relies on placing the prism on the fixing eye and Hering's Law, while watching for the corneal reflections to centralise in the deviating eye. Therefore the secondary Krimsky method was used, with the prism placed in front of the non-fixing left eye.[19] The Krimsky test performed with AHP revealed a right exotropia (RXT) of variable angle (approximately 20-30 prism dioptres), though appearing esotropic at times for near, and right hypertropia of 10 prism dioptres at both near and distance.

Assessment of his ocular movements showed bilateral superior recti restrictions of -5 resulting in an inability to elevate the eyes. There were however, less bilateral restrictions (-1) on downgaze/depression. The movement restriction of the left eye on laevoversion was worse than for the right, with the left lateral rectus not able to abduct the eye past midline (-4) and the right medial rectus unable to adduct by -2. On dextroversion, the left medial rectus was unable to adduct by -2½ and the right lateral rectus was unable to abduct by -½.

His distance visual acuity (VA) tested on the Snellen chart with his glasses (RE +1.50/-3.00x165°, LE +1.50/-3.25x10°) was reduced: RE 6/21 and LE 6/45, and no improvement was achievable with pinhole. Auto-refraction was performed (RE +1.50/-4.25x20°, LE unable to take measurement), but a retinoscopy was not performed as a manifest refraction taken from auto-refraction results could not improve VA. Near VA however was good at N5, when tested using the Moorfield's Bar Reading Book. The Ishihara test revealed no colour vision defect, and pupils were equally reactive to light. Fundal examination appeared normal with no pathological changes. Stereoacuity was also assessed, but no stereopsis was demonstratable, using the near Frisby real-depth and distance Mentor BVAT contour-line stereotests.

DISCUSSION

CFEOM is a disease which truly debilitates the functionality of the eyes. Therefore, the need for appropriate management at an early age is vital, in order to minimise the AHP and to lower the risk of developing amblyopia.[3,5,7,9] Interventions include surgical correction of the blepharoptosis and strabismus for cosmesis, as well as correction of any refractive error using glasses, due to the likely presence of significant astigmatism and amblyopia.[3-4,6,8,16,20] Early detection of amblyopia should be treated aggressively[4] through occlusion therapy for best visual outcome.[20-21] Although studies have shown the possible improvement of VA in amblyopic eyes with compliant full-time occlusion in children aged 7 to 17 years old,[21-22] our patient was not keen on occlusion, thus it was not prescribed. As his condition was long-standing from early childhood, binocular functions were absent and he did not suffer from diplopia due to suppression. Surgical treatment would therefore result in a cosmetic, rather than functional outcome.[5]

Before surgery is undertaken, forced duction testing should be performed to reveal the true extent of the extraocular muscle restrictions.[8,14] Management should be individually tailored due to the differing nature and extent of ocular fibrotic muscle involvement, which in turn, is dependent on the type of genetic loci involved.[3,6-7] Any history of previous extraocular muscle surgery needs to be taken into consideration because scarring can result from repeated surgical procedures and this can affect the treatment outcome.[3,6] Surgery should be sequenced in order of vertical, horizontal and lastly, ptosis correction to reduce lid alteration from precedent strabismus surgery.[3,7,11,14]

All adhesions or fibrotic bands need to be removed from the muscles before any surgical recessions and resections are performed.[2,14] Maximal inferior rectus recessions are very popular for relief of AHP and hypotropia.[3-4,7,11,14,20] Superior rectus resections may also be performed, however they are only used to enhance inferior rectus recessions if needed such as in cases of bilateral involvement.[4,14] Resections are usually avoided no matter the action of the muscle, as CFEOM is a CCDD and there is fear of creating

or worsening the enophthalmos.[8] If there is fibrotic superior rectus involvement, resection and transposition of the superior oblique muscle to the superior rectus insertion is an option.[3,16,20] Inferior rectus recessions are preferred to tenotomies.[11] A silicon plate can also be inserted on the orbital floor, which may improve the hypotropic deviation in primary position, as well as ptosis and palpebral retractions.[20]

With associated horizontal deviations, very large recessions (often greater than 10 mm) are the preferred treatment option, with only occasional resections performed alongside stay sutures and bare sclera conjunctival closures.[6,16] For exotropia, lateral rectus recessions are most popular surgically.[4,7,11,20] They are only accompanied by medial rectus resections if the recession did not have a significant enough impact.[4] However, due to the variability of the horizontal angle in this case, this may be problematic to correct.[4,7] The accuracy of the strabismus angle measurement may be compromised by the use of the Krimsky test due to the inability of the patient to maintain a repeatable and consistent AHP, so care should be taken with the choice of surgical correction.[23] The Krimsky test may therefore be repeated on future visits to eliminate any clinician errors such as placement, size of prism and Krimsky method used and to factor in any possible variability of AHP upon testing.[19] The presence of nystagmus could also have affected the measurements due to the constant oscillation of the eyes.[24] Although not typically a reported factor of CFEOM, nystagmus alongside astigmatic and amblyopic symptoms may coincide with familial CFEOM, as well as some neurological diseases.[4,7,9-11]

A sliding suture or hang-back method is another method that may be used to move the recti muscles as far back as needed for alignment purposes.[4] A traction suture is then used so that the globe maintains its position postoperatively.[4] A conjunctival recession over any recessed muscles may also be performed to enhance weakening.[4,7] The combined correction of hypotropia and exotropia would be required to achieve the best ocular alignment for our patient.

As many extraocular muscles are involved, the aim of the recessions is mainly to shift the eyes to a more appropriate position to relieve the AHP, rather than being effective in treating the extraocular muscle restrictions.[2-3,11,14,25] Full ocular rotations are difficult to restore, usually with unpredictable outcomes.[2-3,19,25] Subsequent surgeries may therefore be required.[3-4,7,11,20] If the AHP is not severe and the patient is not concerned about cosmesis, surgery may be deferred due to the unpredictability of the surgical outcome.[2-3,8,20,25] This information should be provided to the patients and/or their parents preoperatively so that there are no unrealistic postoperative expectations.[3,8,11,14,20]

Treatment of blepharoptosis, may be corrected by frontalis sling suspension and autologous fascia lata or brow alongside inferior rectus surgery if levator action is absent.[4,7,11,14,16,20] In some cases of moderate to severe lid ptosis, a resection of the levator muscle by skin approach rather than conjunctival may be preferred.[3,7] In mild cases, levator resection is effective.[4,7,14] It should be noted that ptosis surgery runs the risk of overcorrection, possibly resulting in exposure corneal keratitis.[6,8,14] Therefore, the aim is to slightly under-correct the ptosis by placing the lid 1 - 2 mm above the pupil in primary position, allowing the visual axis to remain clear and also possibly reducing the AHP.[8,11,14] Lubricant eye drops may be given to those with a higher risk of corneal exposure keratitis both preoperatively and postoperatively.[14]

CONCLUSION

CFEOM is a rare, non-progressive congenital disease, resulting in restrictions of movement of the eye/s, blepharoptosis, AHP and possible amblyopia.[2-3,5-7] Therefore, early interventions such as surgery at a young age is of great importance.[2-3,5-7] Surgical aims include the achievement of improved lid positions, cosmetic or even functional adjustments of the eyes (depending on the length of presentation of the condition), and a reduction or elimination of AHP.[11] Amblyopia management upon early detection, should be carried out through refractive and occlusive treatment regimes.[20] The management options for our patient may include inferior recti recessions to correct the bilateral hypoglobus, lateral recti recessions to correct the exotropia, and frontalis sling suspension to correct the bilateral blepharoptosis. However, as the patient is not concerned with the cosmesis of the strabismus and as the longstanding AHP is not severe, he is reluctant to proceed with any surgical management at this stage.

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2005	Kathryn Rose	The Sydney Myopia Study: Implications for evidence based practice and public health
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2008	Stephen Vale	A vision for orthoptics: An outsider's perspective
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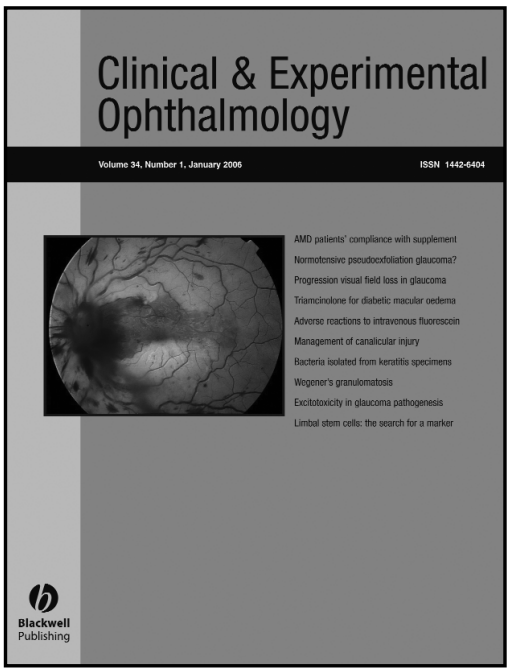

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