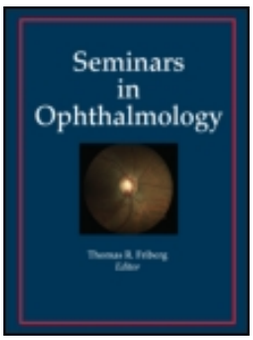

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Presentation of Suspected Pediatric Uveitis

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ABSTRACT

Presentation of suspected pediatric uveitis: Pediatric uveitis is usually managed in specialized ophthalmic centers in the UK. Meaningful data acquisition in these clinics may be helpful in clinical governance, and healthcare planning in a specialty that is gradually changing due to changes in treatment choices. *Methods*: Retrospective analysis of prospectively acquired data in the Liverpool pediatric uveitis database was performed. *Results*: Analysis of our data, based on 147 patients, with a mean age of 10 years, indicated a female to male ratio of 2:1. 99% of patients were Caucasian. Our data indicates 86% of all patients attending the uveitis clinic were diagnosed with juvenile idiopathic arthritis, followed by intermediate uveitis 5% and idiopathic uveitis 4%. 46% of patients required treatment. Systemic treatment included methotrexate (34%), prednisolone (14%), etanercept (6%), ciclosporin (6%), mycophenolate (3%), and infliximab (1%). Severe visual loss (defined by counting fingers or below vision) was seen in 10 eyes despite appropriately treated chronic uveitis. *Conclusion*: Our data shows uveitis-related ocular morbidity in a predominantly pediatric Caucasian population. Patients with severe and chronic uveitis may experience significant uveitis-related complications and subsequent visual loss despite aggressive treatment.

Keywords: Arthritis, childhood, complications, juvenile, treatment, uveitis, vision

INTRODUCTION

Pediatric uveitis may be a difficult entity to recognize and treat due to difficulties in examining young, uncooperative children. Asymptomatic chronic pediatric uveitis may be another problem, which may lead to delay in medical therapy. We report ocular findings in patients presenting with uveitis or conditions predisposing to uveitis in a pediatric population in Liverpool.

MATERIALS AND METHODS

Prospective data collection of all patients attending the pediatric uveitis clinic in Royal Liverpool Children's Hospital during September 2005 to October 2006 was performed. Inclusion criteria included all children with uveitis (past or present) and all children at risk of uveitis. This included children with juvenile idiopathic arthritis and related entities associated with uveitis in a screening program.[1] Patients with traumatic uveitis and postoperative uveitis were excluded. The hospital ethics committee, data protection officer, and research manager were approached and appropriate permissions were secured. Data was added to a custom-built electronic patient record (EPR) database for uveitis patients. Data collected included age, sex, race, diagnosis, visual acuity, uveitis activity, topical or systemic treatment, complications of uveitis, and any risk factors, including appropriate investigations like Rh factor, ANA, and HLA B27, etc. Results were analyzed using ''queries'' in Microsoft Access.

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RESULTS

A total of 147 patients were seen in the uveitis clinic between October 2005 and October 2006. 66% were female ($n = 95$) and 33% were male ($n = 48$). The age distribution was one year (minimum) and 19 years (maximum) with a mean of 10 years. 99% patients were Caucasian ($n = 145$), one was Asian ($n = 1$), and one was Afro-Caribbean ($n = 1$).

82% of all patients seen were diagnosed with juvenile idiopathic arthritis, 5% with intermediate uveitis, 2% with HLA-B27-related uveitis, 1% with sarcoidosis, and 4% with idiopathic uveitis (Table 1).

Antinuclear antibodies were positive in 32% ($n = 47$) and HLA B27 was positive in 6% ($n = 9$) of all patients investigated. Serum ACE was found to be elevated in 3% patients ($n = 5$), but only one was diagnosed as sarcoidosis.

Visual acuities were recorded separately for right and left eyes on a logmar chart. 294 eyes of 147 patients were considered. 66% of eyes had a visual acuity equal to or better than 6/12. Nine eyes were recorded to have poor vision (6/36 or worse). Approximately 30% of patients could not have formal assessment of visual acuity (Table 2).

A total of 27% patients ($n = 40$) had a prior ocular history of anterior uveitis and 7% ($n = 10$) had a prior ocular history of vitreous activity. The remainder had no previous history of uveitis and were being screened for any sign of uveitis (JIA associated uveitis screening). 33 patients (22%) had at least one episode of active uveitis at any stage between October 2005 and October 2006.

In total, 14% of all patients were noticed to have long-term complications of uveitis ($n = 21$). These included 14 patients with posterior synaechiae, eight with band keratopathy (minimum age five years), two with hypotony, one patient with active CMO (both eyes), and two with secondary glaucoma. Cataracts were identified as a complication of uveitis or its treatment (steroid-related, etc). 21 eyes of 19 patients were identified to have cataracts (minimum age five years).

Treatment was noted in 46% patients ($n = 68$). Some of these required treatment for their joint disease, only with no uveitis. Patients required treatment for their active uveitis and joint disease with few being on low-dose maintenance treatment with topical and systemic treatment. Breakdown of those requiring treatment includes 22% topical steroids only ($n = 15$), 41% systemic treatment ($n = 28$), and 37% systemic + topical treatment ($n = 25$). Topical treatment included steroid eye drops (prednisolone or dexamethasone) with or without cycloplegic drops (cyclopentolate 1% or atropine 1%). Systemic methotrexate (34%), prednisolone (14%), etanercept (6%), ciclosporin (6%), mycophenolate (3%), and infliximab (1%) were used in patients requiring systemic therapy.

DISCUSSION

Childhood uveitis may be a difficult disease to monitor and treat because of difficulties associated with examination of young, uncooperative children. Early diagnosis may be particularly difficult if symptoms are not detected or defined and inflammatory signs are unnoticeable, absent, or incorrectly interpreted.[2] This may lead to the insidious development of complications and permanent visual loss due to

TABLE 2. Vision in patients in the pediatric clinic.

Logmar vision Snellen equivalent	Eyes	Percentage*
6/4	3	1
6/5	28	9.5%
6/6	100	34%
6/7.5	26	8.8%
6/9	18	6.1%
6/12	15	5.1%
6/15	4	1.7%
6/18	2	0.6%
6/36	2	0.6%
6/48	1	0.3%
6/60	2	0.6%
CF	1	0.3%
Hand movement	1	0.3%
PL	2	0.6%
Not assessed/assessable**		30%

*Percentages have been rounded up for sake of simplicity.
**Vision was not assessable due to young age, uncooperative behavior, and lack of concentration in these cases.

TABLE 1. Diagnosis of patients in the pediatric uveitis clinic.

Condition	Number of patients	Percentage	Comments
Juvenile idiopathic arthritis	96	86%	
Intermediate uveitis	5	5%	
Idiopathic uveitis	4	4 %	
HLA B27-related uveitis	2	2 %	
Crohn's disease	1	1 %	
CT disorder	1	1 %	No uveitis
Tuberculous uveitis	1	1 %	African patient, old tuberculosis with inactive disease and chorio-retinal scars
Sarcoid uveitis	1	1 %	Systemic sarcoidosis

lack of early diagnosis and treatment. We report the patterns of uveitis, manifest complications, and visual loss in a predominantly Caucasian cohort from a pediatric uveitis clinic in Liverpool.

Most patients at our clinic presented between the ages of four and 13 years of age. However, children as young as two were seen, mainly due to associated juvenile idiopathic arthritis. In cases such as these, perseverance and occasionally an examination under anesthetic may be required for an adequate ocular examination. Kadayifcilar et al. in Turkey reported on a much larger series on an age group of five years and above, presumably because of difficult examination in younger children and social circumstances.[3] However, de Boer et al. in Holland report a series with an age group similar to ours.[4] The differences in these populations may be explained by the differences in geographical, genetic, and racial factors.

Our series shows a much higher female to male ratio than de Boer's group, with 69% female to 31% males. This is presumably due to a higher number of female patients presenting with juvenile idiopathic arthritis in our cohort.

In our series, three patients were recorded to have raised serum ACE levels, but only one was diagnosed with sarcoidosis. Although the raised serum ACE levels are of diagnostic importance in adults, their value in pediatric uveitis is limited.[5]

Compared to the series reported by Kadayifcilar et al., patients in our series did not show any significant features suggestive of Behcet's or active toxoplasmosis. This may be due to the genetic and environmental differences between Turkish and English populations.

The majority of our patients with uveitis had non-granulomatous iridocyclitis. Dana et al. reported similarly high figures for patients with juvenile chronic arthritis and uveitis with non-granulomatous iridocyclitis.[6]

A high proportion of our patients (46%) required treatment. This is because of the referral of screen-positive patients to our unit, who will often require treatment because of uveitis or arthritis or both. As parents and patients have to travel to the hospital, we run joint rheumatology-ophthalmology clinics to give these patients a one-stop service, which includes an ophthalmic and pediatric rheumatology consultation in one room. This also improves the communication between the clinicians to make the best possible decision considering medical and ophthalmic facets of the disease.

Vision was affected in a significant proportion of our patients (see Table 2). The significant ocular morbidity and visual loss are despite an active and robust screening and appropriately aggressive treatment program for children at risk of uveitis in this community. Appropriate and aggressive treatment to control uveitis was prescribed in our patients. This included topical steroids and cycloplegics, systemic prednisolone, methotrexate, etanercept, ciclosporin, and mycophenolate mofetil in appropriate patients based on the clinical course. However, a significant number of children developed long-term complications of uveitis and its treatment. We report a 15% occurrence of at least one complication (excluding cataract). Edelstein et al., in 2002, reported 21% incidence of at least one uveitis-related complication in their cohort.[7] The most significant predictor of complications in this study was severe disease at onset.

Similar rates of ocular morbidity have been reported by other studies, except the incidence of macular oedema. The author feels the incidence of uveitis-associated macular oedema in our study is probably underreported. This may be due to media opacities (cataracts), band keratopathy, or active uveitis being attributed as major causes of visual loss. It may be possible that cystoid macular oedema associated with uveitis is underdiagnosed in children.

Pediatric and juvenile uveitis can be caused by a heterogeneous group of conditions. These conditions may require frequent observation or aggressive topical and systemic treatment, including immune suppression. This may be ideally managed with close input from a pediatric rheumatologist. Our data shows patterns of uveitis-related ocular morbidity in a predominantly pediatric Caucasian population. Patients with severe and chronic uveitis may experience significant uveitis-related complications, which may result in moderate to severe visual loss and may be severe in a small proportion of patients with chronic juvenile uveitis.

We plan to study this cohort as they attend our clinics as per routine treatment and monitoring programs and plan to report on the five-year and 10-year visual results.

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DECLARATION OF INTEREST

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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