



Guidelines for Screening for Uveitis in Juvenile Idiopathic Arthritis (JIA) Produced jointly by BSPAR and the RCPOphth 2006

Aim of the screening programme

To reduce the incidence of visual impairment among children and young people with juvenile idiopathic arthritis (JIA) by early detection through screening allowing for early intervention.

Background

The prevalence of uveitis in JIA overall is approximately 8-30%, but in young oligoarticular onset group (ie arthritis in which up to 4 joints are involved) it may be as high as 45-57%. The annual incidence of JIA in the UK is 1:10,000 with a prevalence of 1:1000. The type of arthritis and age at onset dictates the **risk** of developing uveitis. Only the highest risk groups are included in the regular screening recommendations below. However, late onset of first uveitis can occur even in young adults and cases have been reported in systemic JIA so it is important to make clinical referrals for ophthalmology assessment in patients where there are clinical concerns even if the patient is not specifically covered by these screening guidelines.

The uveitis in JIA is asymptomatic and therefore screening by slit-lamp is essential for diagnosis. Visual impairment arises mainly from complications of the uveitis including cataract, glaucoma, macular oedema and hypotony. Once complications have arisen they are often irreversible. Early detection and treatment can prevent the development of complications and can prevent permanent visual impairment. These complications are more frequent and more severe in younger children and are often asymptomatic. **The most frequent cause of avoidable morbidity remains missed or inadequate examinations in the first year of disease and all efforts must be made to achieve early and thorough early examinations.**

Principles

- 1) **Initial screening examination** Uveitis often starts soon after onset of arthritis but may also start before the arthritis. **The initial screening examination is therefore a clinical priority** and should occur as soon as possible and no later than **6 weeks** from referral. (Reference the BSPAR Standards of Care.)
- 2) **Symptomatic patients** or patients suspected of cataracts or synechiae should be seen within **a week** of referral.

- 3) **Difficult examinations** If the patient is uncooperative at initial screening or for an urgent symptomatic examination in a young child an examination under anaesthetic should be considered.
- 4) **Parent information** Parents and carers of children with JIA need to be fully informed about the possibility of uveitis and that this is usually an asymptomatic condition until complications arise. They need to be told that a high street optician assessment is not an adequate examination to exclude uveitis. They should be instructed to seek medical assessment **urgently** if their child develops visual symptoms or signs. These include red eyes, photophobia, abnormal pupils, corneal clouding, or visual impairment. In younger children this may be manifest by unusual blinking, eye rubbing, visual inattention or preferential attention on auditory signals, or a new onset squint.
- 5) **Missed appointments** Parents and carers must be fully informed about the method of screening and the need to attend for specific uveitis screening examinations on a regular basis. Arrangements need to be in place to give priority to rebooking of any missed appointments in this group with a system of contacting non-attenders.
- 6) **Training** Ophthalmologists and other health professionals carrying out uveitis screening should be appropriately trained and experienced. They should have facilities to audit the outcomes of their screening program.
- 7) **Older patients** Older teenage patients need to be told to return quickly should they become symptomatic. If there is concern about their reliability then they should be considered for longer term less frequent screening. After discharge from the screening programme an annual check by an optometrist is a useful adjunct to self-monitoring for symptoms.
- 8) **On stopping immunosuppressant treatment such as Methotrexate**
Patients who have been treated with methotrexate for their arthritis may not have developed uveitis due to drug suppression . However after methotrexate is stopped uveitis may flare. Screening should therefore restart at 2 monthly intervals after stopping Methotrexate or any other immunosuppressant therapy during the period of maximum risk for 6 months before reverting to the previous screening arrangements.

Specific Screening Schedules

These schedules are the best recommendation possible with current data and are focused on the highest risk groups.

First screening within 6 weeks of referral

Two monthly intervals **from onset of arthritis** for 6 months

Then 3-4 monthly screening for time outlined below:

a) Oligoarticular JIA, Psoriatic arthritis onset and Enthesitis related arthritis (ERA) irrespective of ANA status onset under 11 years

Age at onset	Length of screening
<3 yrs	8 years
3-4yrs	6 yrs
5-8yrs	3 yrs
9-10yrs	1 yr

b) Polyarticular, ANA+ JIA onset < 10 years

AGE at onset	Length of screening
<6 yrs	5 yrs
6-9 yrs	2 yrs

c) Polyarticular, ANA- JIA, onset <7years

All children need 5 yrs screening

This more complicated screening regime above is for high volume centres to safely concentrate their efforts on the highest risk groups. The data is based on refs 1,2. An alternative is to screen all the above groups until their 11th/ 12th birthday, and to continue to screen polyarticular patients presenting older than 7 yrs as well. Older patients presenting for the first time after the age of 11, should undergo one year of screening.

When a patient is discharged from the regular screening program it is vital to stress to them that they and the family are now deemed able to detect any changes in their vision which may signify a new onset or flare of uveitis. It does NOT mean that their risk of uveitis has gone completely. A tip for family self monitoring is to remind the young person to check his or her vision unilaterally- eg by reading small print with each eye once a week.

Monitoring may need to continue indefinitely if there are other reasons such as learning difficulties or treatment non-compliance when the young person may be unable to detect a change in vision or unwilling to seek re-referral.

Systemic onset JIA and definite rheumatoid factor positive polyarticular JIA patients are at very low risk of uveitis. However, there may be delay in being certain about the diagnosis or exact category of JIA, and overlaps between groups do occur. For this reason an initial screening examination may be indicated.

Treatment

Referral to a tertiary referral centre for advice about specific immunosuppression is recommended if complications are present at onset or if the disease is active (without complications) after two years of topical

treatment. Management of complex cases is optimized in tertiary centres with joint clinics between a paediatric rheumatologist and specialist ophthalmologist. Treatment options include topical steroid eye drops mydriatics, sub-tenon injections of steroids, orbital floor injections occasionally systemic steroids intravenous methylprednisolone pulses or oral prednisolone, and increasingly with weekly methotrexate. Cyclosporine, mycophenolate and Infliximab may also be considered.

Some important facts about uveitis in JIA

Boys do get severe uveitis

ANA negative patients can get significant uveitis

Uveitis does occur in young patients with psoriatic arthritis

Patients with polyarticular JIA do get uveitis.

Patients with enthesitis related arthritis may get chronic as well as acute iritis.

Refs:

Symmons; J Rheum 1996;23:1975-80

Edelstein C, Lee V, Bentley CR, Kanski JJ, Graham EM. An evaluation of baseline risk factors predicting severity in juvenile idiopathic arthritis associated uveitis and other chronic anterior uveitis in early childhood. Br J Ophthalmol 2002;86:51-6.

Zulian F, Martini G, Falcini F et al. Early predictors of severe course of uveitis in oligoarticular juvenile idiopathic arthritis. J Rheumatol 2002;29:2446-53.

Rosenberg AM. Uveitis associated with juvenile idiopathic arthritis: envisioning the future. J Rheumatol 2002; 29:2253-5.

Kanski JJ Uveitis in juvenile chronic arthritis :incidence, clinical features and prognosis . Eye 1988,;2:641-5

Kotaniemi K, Arkela- Kautianen M , Haapasaari J, Lerisalo-repo M et al Ann Rheum Dis 2005 Jun 64 (6):871-4

Kotaniemi K, Kotaniemi A, Savolainen A. Uveitis as a marker of active arthritis in 372 patients with juvenile idiopathic seronegative oligoarthritis or polyarthritis. Clin Exp Rheumatol 2002;20(1):109-12

Kotaniemi K, Kautianen H, Karma A, Aho K. Occurrence of uveitis in recently diagnosed juvenile chronic arthritis; a prospective study. Ophthalmology 2001;108(11):2071-5

Kodsi SR, Rubin SE, Milojevic D, Ilowite N, Gottlieb B. Time of onset of uveitis in children with juvenile rheumatoid arthritis. JAAPOS 2002;6(6):373-6.

Heiligenhaus A, Niewerth M, Mingels A, Ganser G ,Thon A , Pleyer U Greiner K, Minden K. Epidemiology of uveitis in juvenile idiopathic arthritis from a national paediatric rheumatology and ophthalmologic database. Klin monatsbl Augenheilkd 2005 Dec ;222(12):993-1001.

Berk AT, Kocak N, Unsal E Uveitis in juvenile arthritis . Ocul Immun Inflamm 2001;9 (4):243-51

Foeldvari I, Wierk A. Methotrexate is an effective treatment for chronic uveitis. J Rheumatol 2005 Feb;32 (2):362-5.

Schmeling H , Horneff G. Rheumatology 2005 Aug ;44 (8): 1008-11. Etanercept and uveitis in patients with juvenile idiopathic arthritis.

Carvounis PE et al 2006. Graefes Arch Clin Exp Ophthalmol 2006 Mar;244 (3) 281-90.

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