Guidance for Screening for Uveitis in Juvenile Idiopathic Arthritis (JIA)  
June 2018

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 is approximately 8-30%, but in a young oligo-articular onset group (i.e. arthritis in which up to four 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 diagnosis can occur even in young adults and cases have been reported in systemic JIA. Therefore, it is important to make clinical referrals for ophthalmology assessment in patients where there are concerns, even if the patient is not specifically covered by these screening guidelines.

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 therefore 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 is 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 six weeks from referral.
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 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 re-booking of any missed appointments in this group with a system of contacting non-attenders.
6. Ophthalmologists and other health professionals carrying out uveitis screenings should be appropriately trained and experienced. They should have facilities to audit the outcomes of their screening programme.
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. 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 two-monthly intervals after stopping methotrexate or any other immunosuppressant therapy during the period of maximum risk for six 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 six weeks of referral
- Two monthly intervals from onset of arthritis for six months
- Then three to four monthly screenings for times outlined below:

  a) Oligoarticular JIA, psoriatic arthritis onset and enthesitis-related arthritis (ERA) irrespective of ANA status onset under 11 years.

<table>
<thead>
<tr>
<th>Age at onset</th>
<th>Length of screening</th>
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<tbody>
<tr>
<td>&lt;3 years</td>
<td>8 years</td>
</tr>
<tr>
<td>3-4 years</td>
<td>6 years</td>
</tr>
<tr>
<td>5-8 years</td>
<td>3 years</td>
</tr>
<tr>
<td>9-10 years</td>
<td>1 year</td>
</tr>
</tbody>
</table>

  b) Polyarticular, ANA+ JIA onset < 10 years.

<table>
<thead>
<tr>
<th>Age at onset</th>
<th>Length of screening</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;6 years</td>
<td>5 years</td>
</tr>
<tr>
<td>6-9 years</td>
<td>2 years</td>
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</tbody>
</table>

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

All children need five years of 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 references 1 and 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 years 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 programme, 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 uni-ocularly- e.g. 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 optimised 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, e.g. intravenous methylprednisolone pulses or oral prednisolone, and increasingly with weekly methotrexate. Ciclosporine, 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
References

- Adapted from combined documents produced by the Royal College of Ophthalmologists and the Council of the paediatric and adolescent rheumatology section.

This guidance document was ratified by the Council of the paediatric and adolescent rheumatology section within the British Society for Rheumatology and is designed to support the delivery of paediatric and adolescent care in rheumatology. It was produced jointly with the Royal College of Ophthalmologists.

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Last reviewed: June 2018