Guidelines for Ophthalmologic Examinations in Children With Juvenile Rheumatoid Arthritis

Section on Rheumatology and Section on Ophthalmology

Chronic, nongranulomatous iridocyclitis is an important complication of juvenile rheumatoid arthritis (JRA). First reported by Ohm in 1910, the association between iridocyclitis and JRA has become well established. The intraocular inflammation referred to as iridocyclitis primarily affects the iris and ciliary body. Overall, the reported incidence of iridocyclitis varies from 2% to 21% in children with JRA. The morbidity of iridocyclitis includes cataracts, glaucoma, band keratopathy, and loss of vision. Diagnosis of early iridocyclitis is usually not possible by routine direct ophthalmoscopy. Slitlamp examination detects the signs of active anterior chamber inflammation. Guidelines for the schedule of routine serial slitlamp examination are suggested for early detection of iridocyclitis.

The presentation of eye involvement in JRA may be asymptomatic or of an insidious onset. The outcome has improved in the past 20 years. The majority of children have a relatively good visual prognosis if the iridocyclitis is detected and treated early.

RISK FACTORS FOR IRIDOCYCLITIS

Articular Features

The diagnosis of JRA describes a heterogeneous group of arthritic conditions with onset of disease before age 16 years. There are three major subtypes of JRA: systemic onset, polyarticular onset, and pauciarticular onset, defined by the clinical manifestations in the first 6 weeks of the disease. Fewer than 2% of children with systemic-onset JRA have iridocyclitis. Children with polyarticular disease are at moderate risk, with 7% to 37% incidence of iridocyclitis. The majority of children with iridocyclitis have pauciarticular disease. The onset of the iridocyclitis may precede the onset of the arthritis in approximately 6% of cases. Rarely, it occurs in the absence of arthritis after long-term follow-up. Iridocyclitis may be detected at the time of initial diagnosis of arthritis; however, it most often presents over the next 5 to 7 years. The highest risk of iridocyclitis is most commonly within 2 years of the onset of arthritis. Children with JRA remain at risk for iridocyclitis into adulthood. There are reports of iridocyclitis diagnosed more than 20 years after the onset of the arthritis. Iridocyclitis does not usually parallel the activity of joint disease.

Age, Gender

The majority of children at risk for developing iridocyclitis are young females with pauciarticular-onset JRA. The peak age of onset of the arthritis in this group is age 2 to 5 years, with subsequent development of iridocyclitis within the next 5 to 7 years.

Immunogenetic and Serologic Markers

The serologic marker most strongly associated with iridocyclitis is the presence of antinuclear antibodies. Antinuclear antibodies are present in 65% to 88% of these children. They are usually present in low titer and are of unknown specificity. Rheumatoid factor is not usually present in children with iridocyclitis. Rheumatoid factor positivity is most commonly seen in older children with polyarticular disease that is clinically similar to adult rheumatoid arthritis.

Genetic factors may predispose to the development of iridocyclitis. Recent data show that HLA-DR5 is correlated with the presence of eye disease, and HLA-DR1 with its absence; HLA-DRw8, which strongly predisposes to pauciarticular-onset JRA, was neutral with respect to eye disease.

Clinical Characteristics of Iridocyclitis

The ocular inflammation is insidious in onset and asymptomatic in the majority of patients. Due to the lack of symptoms, the exact time of onset of ocular involvement is frequently difficult to determine. This emphasizes the need for slitlamp examination by an ophthalmologist. Signs or symptoms in children with iridocyclitis may include red eyes, decreased vision, unequal pupils, ocular pain, and headaches. Most cases of iridocyclitis are bilateral; children with unilateral iridocyclitis may progress to bilateral iridocyclitis after the initial 12 months of eye disease. Visual prognosis is improved by early detection of iridocyclitis.

In 25% of iridocyclitis cases, the prognosis is very good. Twenty-five percent of children respond poorly to treatment and may require surgery for cataracts and/or glaucoma. This group is at risk for loss of vision and may experience more ocular than articular morbidity. Approximately 50% of patients require prolonged treatment for moderate to severe chronic inflammation; however, the visual prognosis in these patients is generally good. Functional blindness has been reported in 15% to 40% of affected eyes.

The recommendations in this statement do not indicate an exclusive course of treatment or serve as a standard of medical care. Variations, taking into account individual circumstances, may be appropriate.
GUIDELINES FOR THE FREQUENCY OF OPHTHALMOLOGIC EXAMINATIONS IN CHILDREN WITH JUVENILE RHEUMATOID ARTHRITIS

The suggested frequency of ophthalmologic visits for children with JRA without known iridocyclitis is presented in the Table. Once iridocyclitis is diagnosed, the treating ophthalmologist will determine the frequency of visits.

The subtype of juvenile arthritis is determined by the systemic features of the illness and the number of joints with arthritis during the first 6 weeks of the illness. Pauciarticular JRA is defined by involvement of four or fewer joints, polyarticular JRA is defined by involvement of more than four joints, and systemic JRA is defined by a characteristic rash associated with spiking fevers during the first 6 weeks of the illness. Initial referral to an ophthalmologist should be made at the time of diagnosis of JRA.

TABLE. Frequency of Ophthalmologic Visits for Children With Juvenile Rheumatoid Arthritis (JRA) and Without Known Iridocyclitis*

<table>
<thead>
<tr>
<th>JRA Subtype at Onset</th>
<th>Age of Onset</th>
<th>&lt;7 y†</th>
<th>≥7 y‡</th>
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</thead>
<tbody>
<tr>
<td>Pauciarticular</td>
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<tr>
<td>+ ANA</td>
<td>H§</td>
<td>M</td>
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</tr>
<tr>
<td>- ANA</td>
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<td>M</td>
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<tr>
<td>Polyarticular</td>
<td></td>
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<tr>
<td>+ ANA</td>
<td>H§</td>
<td>M</td>
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</tr>
<tr>
<td>- ANA</td>
<td>M</td>
<td>M</td>
<td></td>
</tr>
<tr>
<td>Systemic</td>
<td></td>
<td>L</td>
<td>L</td>
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</tbody>
</table>

* High risk (H) indicates ophthalmologic examinations every 3 to 4 months. Medium risk (M) indicates ophthalmologic examinations every 6 months. Low risk (L) indicates ophthalmologic examinations every 12 months. ANA indicates antinuclear antibody test.
† All patients are considered at low risk 7 years after the onset of their arthritis and should have yearly ophthalmologic examinations indefinitely.
‡ All patients are considered at low risk 4 years after the onset of their arthritis and should have yearly ophthalmologic examinations indefinitely.
§ All high-risk patients are considered at medium risk 4 years after the onset of their arthritis.

REFERENCES

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