CARING FOR PATIENTS WITH JUVENILE IDIOPATHIC ARTHRITIS

Know the ocular manifestations and screening protocol for this autoimmune disease.

BY KATHRYN ANDREWS, OD, FAAO

As primary eye care providers, optometrists are on the front lines of diagnosing and treating ocular diseases in pediatric patients that may have systemic origins. It is therefore crucial that we are able to rapidly identify conditions that may have underlying consequences beyond the visual system and to communicate effectively with the child’s pediatrician.

Many systemic conditions can present first with ocular manifestations. They range from diabetes mellitus, which can lead to retinopathy, to more obscure conditions such as Wilson disease, which can result in a Kayser-Fleischer ring in the cornea.

Another is juvenile idiopathic arthritis (JIA), also known as juvenile rheumatoid arthritis. Left untreated, JIA can have sight-threatening complications. This article reviews the classifications of JIA and presents appropriate screening measures and treatment strategies.

CLASSIFICATIONS

JIA is one of the most common inflammatory autoimmune conditions in children. JIA describes a group of chronic joint disorders, and it may be divided into five subtypes: systemic, oligoarticular, polyarticular, juvenile psoriatic, and enthesitis-related JIA.

Oligoarticular JIA presents as arthritis in four or fewer joints, most often in large joints such as the knee or elbow. Polyarticular JIA presents as arthritis in five or more joints, usually smaller joints of the hands and fingers. Juvenile psoriatic arthritis occurs in conjunction with psoriasis that typically precedes the onset of joint inflammation. Enthesitis-related JIA occurs at the junction where a tendon meets a bone.

JIA-associated uveitis is the most common extraarticular manifestation of JIA. The type of uveitis typically associated with JIA is a recurrent non-granulomatous anterior uveitis. In 3% to 7% of children with JIA, diagnosis of uveitis can precede diagnosis of the systemic condition.

Several risk factors make a child with a diagnosis of JIA more likely to develop anterior uveitis. The patient’s sex, JIA subcategory, age of onset, and results of antinuclear antibody and human leukocyte antigen B27
blood testing are useful in determining appropriate follow-up. In female patients with JIA, younger age of onset, oligoarticular subtype, and the presence of antinuclear antibodies are risk factors for the development of chronic anterior uveitis.1-5 In male patients with JIA, the presence of human leukocyte antigen B27 and enthesitis-related arthritis are risk factors for the development of uveitis.1

The polyarticular JIA subtype may be subdivided into positive and negative rheumatoid factors based on blood testing.1 Children with a positive rheumatoid factor test result are far less likely than those with a negative result to develop uveitis.1-5

Clinicians must bear in mind that children with uveitis are usually asymptomatic. They rarely present with symptoms of adult uveitis such as limbal hyperemia, pain, or severe photophobia. Often, children remain asymptomatic even after their VA has been drastically compromised. For this reason, we must routinely examine

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**TABLE 1. Recommended Screening Protocol in Patients With JIA**

<table>
<thead>
<tr>
<th>JIA SUBTYPE</th>
<th>ANA RESULTS</th>
<th>AGE AT JIA ONSET, Y</th>
<th>JIA DURATION, Y</th>
<th>RECOMMENDED SCREENING INTERVAL, MO</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Oligoarticular arthritis</td>
<td>+</td>
<td>≤ 6</td>
<td>≤ 4</td>
<td>3</td>
</tr>
<tr>
<td>• RF-negative polyarticular arthritis</td>
<td>&gt; 6</td>
<td>≤ 2</td>
<td>&gt; 4</td>
<td>6</td>
</tr>
<tr>
<td>• Psoriatic arthritis</td>
<td>-</td>
<td>≥ 4</td>
<td>≥ 7</td>
<td>12</td>
</tr>
</tbody>
</table>

**Enthesitis-related arthritis**

N/A

**RF-positive polyarticular arthritis**

N/A

**Abbreviations:** ANA, antinuclear antibody; JIA, juvenile idiopathic arthritis; MO, months; NA, not applicable; RF, rheumatoid factor; Y, years

**TABLE 2. Recommended Length of Screening**

<table>
<thead>
<tr>
<th>UVEITIS SUBTYPE</th>
<th>AGE AT ONSET OF JIA, Y</th>
<th>LENGTH OF SCREENING PROTOCOL, Y</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Oligoarticular arthritis</td>
<td>&lt; 3</td>
<td>8</td>
</tr>
<tr>
<td>• Psoriatic Arthritis</td>
<td>3-4</td>
<td>6</td>
</tr>
<tr>
<td>• Enthesitis-related arthritis</td>
<td>5-8</td>
<td>3</td>
</tr>
<tr>
<td></td>
<td>9-10</td>
<td>1</td>
</tr>
<tr>
<td>Polyarticular, ANA +JIA</td>
<td>&lt; 6</td>
<td>5</td>
</tr>
<tr>
<td></td>
<td>6-9</td>
<td>2</td>
</tr>
</tbody>
</table>

**Abbreviations:** ANA, antinuclear antibody; JIA, juvenile idiopathic arthritis; Y, years
 pediatric patients with a diagnosis of JIA at the slit lamp.

**SCREENING GUIDELINES**

Tables 1 and 2 present screening guidelines for children with a diagnosis of JIA who do not have a diagnosis of uveitis. Children should have their first screening within the first few weeks of their JIA diagnosis. The screening recommendation for enthesitis-related arthritis is 12 months because patients typically have acute anterior uveitis with symptoms that mimic those of adult uveitis, which would prompt a visit secondary to the appearance of the symptoms.

**TREATMENT**

The first line of treatment for anterior uveitis includes topical glucocorticoids and cycloplegic agents. Long-term steroid use can lead to cataract formation, increased IOP, and secondary glaucoma, but the benefits of short-term therapy with a glucocorticoid outweigh its risks. Patients with severe or sight-threatening uveitis may require a systemic steroid to control ocular inflammation.

If topical and systemic steroids fail to control ocular inflammation, the child’s rheumatologist may initiate a nonbiologic disease-modifying anti-rheumatic drug. A combination of methotrexate and a monoclonal antibody tumor necrosis factor inhibitor is frequently recommended over monotherapy. In the Systemic Immunosuppressive Therapy for Eye Diseases (SITE) cohort study, the use of nonbiologic disease-modifying anti-rheumatic drugs and immunosuppressant medications decreased the risk of VA loss in patients with uveitis. Before systemic therapy is tapered, the child’s uveitis should be well controlled for at least 2 years.

**AN OPPORTUNITY FOR COLLABORATIVE CARE**

JIA presents as an opportunity for optometry to work closely with rheumatology to monitor and comanage the health of pediatric patients. JIA-associated uveitis is a common manifestation of the disease that can lead to severe vision loss if patients are not observed closely.

Our communication with the pediatric rheumatologist is pivotal to these young patients’ ocular and systemic outcomes. JIA management highlights the importance of routine eye examinations and demonstrates the potential for ocular health evaluations to lead to systemic diagnoses in a pediatric population.

**AT A GLANCE**

- Juvenile idiopathic arthritis (JIA), one of the most common inflammatory autoimmune conditions in children, describes a group of chronic joint disorders.
- JIA-associated uveitis is a common manifestation of the disease that can lead to severe vision loss if patients are not observed closely.
- Routine eye examinations are important in the management of patients with JIA. This is an example of the potential for ocular health evaluations in the pediatric population to lead to systemic diagnoses.


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