The iris, ciliary body and choroid make up the uveal tract. The uveal tract is the middle coat of the eye, lying between the white outer covering of the eyeball (sclera) and the retina, which lines the interior of the posterior segment of the globe (Figure 1). Uveitis is an inflammation of any part or all of the uvea. Panuveitis is when the entire uvea is inflamed. Iritis (iritis ciliare) inflammation of the iris and ciliary body. Iritis is characterized by a breakdown of the natural barrier between the blood vessels and the fluid (aqueous humor) that fills the space between the iris and the cornea (the anterior chamber). As a result, protein, fibrin, and white blood cells enter the aqueous. The cells and debris circulating in the anterior chamber can be deposited on the inner (interior) surface of the cornea (the corneal endothelium). These deposits, called keratic precipitates (KP), can be fine or take the form of larger greasy clumps called mutton-fat KP. A distinction is made between non-granulomatous iritis (with fine KP) and granulomatous iritis (with mutton-fat KP). Although more severe uveitis, including panuveitis, can be a manifestation of juvenile rheumatoid arthritis (JRA), it is the non-granulomatous iritis that is most characteristic of JRA.

Chronic anterior non-granulomatous uveitis in children is most often caused by JRA (known as juvenile chronic arthritis [JCA] in the European literature). Roughly 6% of all cases of uveitis occur in childhood and of these, 80% are associated with juvenile rheumatoid arthritis. The incidence of iritis in patients with JRA ranges from 8% to 24%, but certain subgroups are at high risk. Of all cases of JRA-associated uveitis, 78% to 90% will have pauciarticular JRA (and 90% of these will be antinuclear antibody [ANA] positive and rheumatoid-factor negative), 7% to 14% will have polyarticular disease, and 2% to 6% will have systemic JRA (Still’s disease). Young girls with pauciarticular onset, ANA seropositivity and rheumatoid factor negativity are most likely to develop uveitis. Many children who are rheumatoid factor positive and have polyarticular disease uncommonly develop uveitis. The majority of children who develop uveitis do so within four to seven years of the onset of JRA; the peak is within two years of onset. The disease is bilateral in 75% to 80% of children. Common symptoms and signs of other forms of iritis are pain, photophobia, and red eye; however, the anterior uveitis seen in JRA is almost entirely asymptomatic. To detect uveitis, an ophthalmologist must examine the anterior chamber aqueous with a slit lamp to see if cells, keratic precipitates and/or proteinaceous exudate (called flare) are present. Because flare can persist despite treatment in some children with JRA, the focus of treatment is to eliminate cells. Even low-grade uveitis left untreated can lead to ocular damage including retinal swelling (macular edema), glaucoma and cataract. Up to 10% of patients with JRA will develop iritis before joint disease.
There is no correlation between joint activity and eye disease. To prevent ocular damage, children with JRA must be screened regularly by an ophthalmologist who is familiar with the disease and comfortable performing thorough eye examinations on sometimes-uncooperative children.

The mainstay of treatment is corticosteroid medication, which can be administered topically, by regional injection, orally or parenterally. The first-line treatment is usually topical steroid drops or ointments. Administration as often as every hour may be needed to control the inflammation. Although many children will tolerate a short course of treatment followed by a rapid tapering of their eyedrops, approximately 10% to 15% of children with JRA iritis will require topical steroids to be tapered slowly. In our experience, tapering too quickly is the most common cause of recurrent chronic disease. Some children may require tapering that takes many months, continuing with doses as low as one drop per week. It is this unique feature of JRA that makes it essential that children be cared for by an ophthalmologist well versed in this disease.

Even though many children with pauciarticular onset develop polyarticular disease, it is the original presentation as pauciarticular that confers a higher lifetime risk. Some authors have suggested decreasing screening frequency seven years after onset in children older than seven if an eye disease has never occurred.6

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If the adverse effects of steroids such as glaucoma do not develop, a periocular injection may allow for reduction in topical dose frequency. The injection, which in young children may require a general anesthetic, is given in the sub Tenon’s space. Tenon’s fascia lies outside the eyeball underneath the conjunctiva. In more resistant cases, oral or parenteral steroids may be needed. When steroids fail or are poorly tolerated, methotrexate (oral or subcutaneous) can be a valuable adjunct.7–9 Nonsteroidal anti-inflammatory medications appear to have a limited role both topically and orally. Other immunosuppressive medications might sometimes be necessary.

During active uveitis, a topical dilating drop is usually used to prevent scar tissue (posterior synechiae) from forming between the iris and the pupil. These synechiae may cause the border of the pupil to stick down to the lens causing an irregular and poorly dilating pupil.

Figure 1

The main ocular complications of JRA-associated iritis are cataract, glaucoma and macular edema. Despite all the advances in the management of this condition, up to 22% of children with pauciarticular
JRA-associated uveitis still develop legal blindness secondary to chronic low-grade intraocular inflammation. The prognosis is more favourable for males, for those with unilateral disease, for older children, for those in whom the disease is milder at presentation, and for children without posterior synechia or cataract at presentation. Cataract and glaucoma can both also be caused by steroids. When vision is threatened, cataract may require surgery, although this can further aggravate iritis. Therefore, attempts are made to delay surgery when possible until the iritis is well controlled. Unlike adults, intraocular lens implantation in children with JRA can have serious complications. Glaucoma can occur in children who have had cataract removal and in those who have not. It is difficult to treat and a combination of medication (topical and systemic) and surgery are usually needed. Glaucoma may be due to clogging of the eye’s drainage system (trabecular meshwork) by cells and protein in the aqueous, excessive posterior synechia blocking the flow of aqueous through the pupil, or synechia which cause the iris tissue to scar over the trabecular meshwork. Macular and optic nerve edema are very difficult to treat and often lead to irreversible loss of vision.

JRA-associated uveitis is a chronic asymptomatic condition that can lead to severe blinding ocular complications. Early detection through routine screening and appropriate management is essential.

**TABLE 1**

<table>
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<tr>
<th>Onset:</th>
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<th>Kanski</th>
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<tr>
<td>&lt; 7 y/o</td>
<td>&gt;7 y/o</td>
<td></td>
</tr>
<tr>
<td>Pauciarticular at onset &amp; ANA positive</td>
<td>3–6 months*</td>
<td>6 months</td>
</tr>
<tr>
<td>Pauciarticular at onset &amp; ANA negative</td>
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<td>6 months</td>
</tr>
<tr>
<td>Polyarticular onset &amp; ANA positive</td>
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<td>6 months</td>
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<tr>
<td>Polyarticular onset &amp; ANA negative</td>
<td>6 months</td>
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</tr>
<tr>
<td>Systemic onset</td>
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<td>12 months</td>
</tr>
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</table>

† Suggested frequency of visits for a child without a history of iritis.
* After four years and no eye involvement, decrease to 6 months.

References: