AMERICAN UVEITIS SOCIETY

Juvenile Idiopathic Arthritis–associated Uveitis

A Patient Education Monograph prepared for the American Uveitis Society **January 2003**

by C. Stephen Foster, M.D., F.A.C.S. and Saadia Rashid
Ocular Immunology and Uveitis Service
Massachusetts Eye and Ear Infirmary
Harvard Medical School
Boston, MA, USA

Introduction

Juvenile Idiopathic Arthritis (JIA), [formerly known as Juvenile Rheumatoid Arthritis or JRA] is a chronic arthritis with onset prior to the age of 16 years. JIA is the most common cause of arthritis in children. In the United States, one in every 1000 children develops JIA. Of all cases of uveitis, approximately 6 out of every 100 occur in children, with up to 80% of these associated with JIA, making this disease the most commonly identified cause of uveitis in children. JIA is three times more likely to affect girls than boys, but has no geographical or racial predilection. The uveitis associated with JIA typically causes inflammation of the anterior part of the eye, affecting the iris and the ciliary body (and therefore termed an iridocyclitis).

History

JIA has existed since ancient times. An early adolescent skeleton with changes compatible with JIA was entombed in the Peruvian Andes between AD 900 and 1050. Although children with inflammation of multiple joints were first described by Cornil in 1864 and by Diamentberger in 1890, it was not until 1897 that George Frederick Still provided the clinical description that formed the basis for the diagnosis of JIA. Eye inflammation in JIA has been recognized since Ohm's first description in 1910.

Course of Disease

The course of uveitis depends upon the way in which the arthritis begins. JIA is classically divided into three subtypes, based upon the number of joints involved at the onset of the disorder: (1) oligoarticular (or pauciarticular), which involves four or fewer joints; (2) polyarticular, which involves five or more joints; and (3) systemic onset, which involves any number of joints along with systemic symptoms. These categories account for 50%, 40% and 10% of all cases of JIA, respectively.

(1) Oligoarticular onset JIA occurs in early childhood, usually around 2 years of age. The knees and less frequently the ankles and wrists may exhibit painless swelling. The arthritis is evanescent and rarely destructive. Twenty percent of children with oligoarticular onset JIA develop uveitis.

(2) Polyarticular onset JIA can occur throughout childhood, but the peak age of onset is 3 years. Small joints of the hands are typically involved. Joint involvement may be destructive in 15% of patients with polyarticular onset JIA. Systemic symptoms such as loss of appetite and slowing of growth may be present. Five percent of children with polyarticular onset JIA develop uveitis.

(3) Systemic onset JIA can occur at any time during childhood. Both large and small joints can be affected. In addition, the skin, liver, spleen, and coverings of the heart and lungs can become involved. The arthritis of systemic onset JIA is destructive in 25% of the patients; uveitis however is very rare. Only 6% of patients with systemic onset JIA develop uveitis.

The majority of JIA-associated uveitis patients have oligoarticular onset JIA (78 to 90%) while 7-14% have the polyarticular variety. JIA–associated uveitis is chronic and usually involves both eyes, either simultaneously or within a few months of each other. The majority of patients develop uveitis within 4 to 7 years of joint involvement; the average age at diagnosis is 6 to 8 years. Uveitis begins before arthritis in about 6% of cases. The greatest risk of uveitis development occurs within the first 2 years after the onset of JIA, and the risk declines considerably by 8 years after arthritis onset.

Eye symptoms are non-specific and may include pain, light sensitivity and blurring of vision. Unfortunately, the disease does not cause obvious symptoms in about 50% of patients and symptoms only appear once the vision robbing complications such as cataract, band keratopathy, glaucoma, and hypotony have already occurred. There is no correlation between the severity of uveitis and that of the arthritis, thus a patient with mild arthritis may have a severe uveitis that produces no warning signs until late in the disease.

Diagnosis, Testing and Screening

The diagnosis of JIA-associated uveitis is based upon history, signs of inflammation detected on a thorough eye examination using the slit lamp, and laboratory tests. Initial testing includes blood tests for a special antibody called antinuclear antibody (ANA). Other blood tests and a chest X-ray may be performed to ensure that other diseases, which may look similar, are not present.

Screening for patients with newly diagnosed JIA is mandatory, because small visual changes due to active inflammation are not usually noticed or reported by young children. All newly diagnosed JRA patients should be referred immediately to an eye doctor for a complete eye examination. If uveitis is present, then the eye doctor determines the frequency of visits depending upon the severity of the disease and the type and response to treatment. If no uveitis is present, then the following screening algorithm may be used:

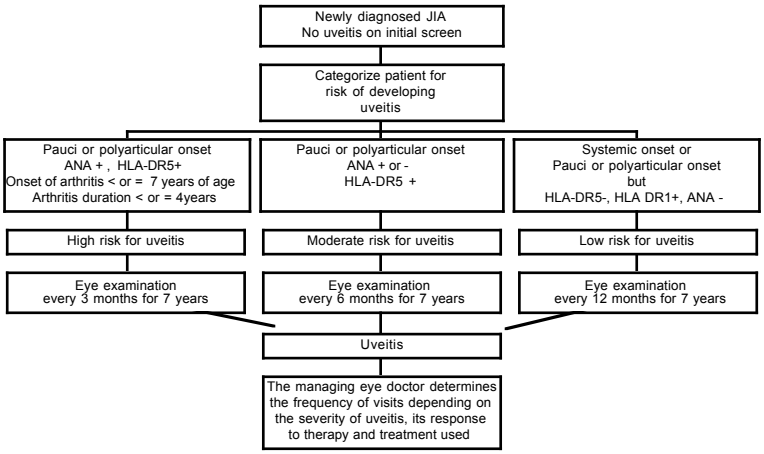

Figure 1. Screening algorithm for monitoring for presence of eye disease in patients with Juvenile idiopathic arthritis. (See the glossary for a definition of the HLA system. Note that while ANA testing is typically performed, not all doctors perform HLA-DR5 testing.)

Treatment

The aim of treatment is complete elimination of all active inflammation in the eye at all times. In aspiring to this goal, the patient, family, and physician attempt to balance the risk of the medications with the intensity of the inflammation. An aggressive "step-ladder" approach is used to achieve this. Initial management consists of topical corticosteroid therapy, sometimes combined with regional corticosteroid injection and corticosteroid pills taken by mouth. In the case of treatment failure after 90 days of compliant therapy or relapse when corticosteroids are tapered, oral non-steroidal anti-inflammatory drugs may be administered by mouth. Approximately 70% of the patients respond to this strategy. The remaining 30% generally require immunomodulatory therapy to accomplish the goal of freedom from uveitis and freedom from chronic steroid use.

Cause of Condition

The cause of uveitis in JIA is unknown. Some scientists believe that it might be caused by an attack mounted by the patient's own white blood cells against certain substances present in the patient's iris. However, it is not clearly understood whether the immune reaction is the cause or merely an effect of damage to the eye.

Prognosis

In children with JIA associated uveitis, 10% have mild uveitis, 15% have moderate uveitis, 50% have severe uveitis and 25% are unresponsive to therapy. Overall, 75% of children with severe uveitis experience visual loss resulting from various eye complications. This means that overall up to 12% of children with JIA-associated uveitis will develop permanent, profound loss of vision as a result of persistent low grade inflammation. This is preventable through early diagnosis and prompt treatment.

Research and future outlook

The prevalence of blindness in children with JRA associated uveitis has decreased from 50% in 1950 to its current level of approximately 12%. Further reduction of this prevalence figure can be achieved by early referrals for newly diagnosed JIA patients, screening programs for early detection of uveitis and development of collaborative liaisons between ophthalmologists and rheumatologists to reduce ocular damage and blindness.

Close Window