Complete Summary

GUIDELINE TITLE

Ophthalmologic examinations in children with juvenile rheumatoid arthritis.

BIBLIOGRAPHIC SOURCE(S)

Cassidy J, Kivlin J, Lindsley C, Nocton J, Section on Rheumatology, Section on Ophthalmology. Ophthalmologic examinations in children with juvenile rheumatoid arthritis. Pediatrics 2006 May;117(5):1843-5. [19 references] PubMed

GUIDELINE STATUS

This is the current release of the guideline.

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COMPLETE SUMMARY CONTENT

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SCOPE

DISEASE/CONDITION(S)

Ocular diseases, including chronic uveitis, in juvenile rheumatoid arthritis

GUIDELINE CATEGORY

Risk Assessment
Screening

CLINICAL SPECIALTY

Ophthalmology
Pediatrics
Rheumatology

INTENDED USERS

Physicians

GUIDELINE OBJECTIVE(S)

To provide recommendations on ophthalmologic examinations in children with juvenile rheumatoid arthritis

TARGET POPULATION

Children with juvenile rheumatoid arthritis

INTERVENTIONS AND PRACTICES CONSIDERED

1. Risk assessment by:
    - Articular features
    - Age
    - Immunogenetic and serologic markers (e.g., antinuclear antibodies)
    - Clinical characteristics
2. Regularly scheduled slit-lamp examinations

MAJOR OUTCOMES CONSIDERED

Risk for chronic uveitis

METHODOLOGY

METHODS USED TO COLLECT/SELECT EVIDENCE

Searches of Electronic Databases

DESCRIPTION OF METHODS USED TO COLLECT/SELECT THE EVIDENCE

Not stated

NUMBER OF SOURCE DOCUMENTS

Not stated

METHODS USED TO ASSESS THE QUALITY AND STRENGTH OF THE EVIDENCE

Not stated

RATING SCHEME FOR THE STRENGTH OF THE EVIDENCE

Not applicable

METHODS USED TO ANALYZE THE EVIDENCE

Review

DESCRIPTION OF THE METHODS USED TO ANALYZE THE EVIDENCE

Not stated

METHODS USED TO FORMULATE THE RECOMMENDATIONS

Not stated

RATING SCHEME FOR THE STRENGTH OF THE RECOMMENDATIONS

Not applicable

COST ANALYSIS

A formal cost analysis was not performed and published cost analyses were not reviewed.

METHOD OF GUIDELINE VALIDATION

Not stated

DESCRIPTION OF METHOD OF GUIDELINE VALIDATION

Not applicable

RECOMMENDATIONS

MAJOR RECOMMENDATIONS

Risk Factors for Chronic Uveitis

Articular Features

The classification of juvenile rheumatoid arthritis (JRA) describes a heterogeneous group of disorders of predominantly peripheral arthritis with onset of disease before 16 years of age. The 3 major onset types defined by clinical manifestations in the first 6 months of the disease are oligoarticular (pauciarticular), polyarticular, and systemic. The onset type is determined by the systemic features of the illness and the number of joints with arthritis at diagnosis. Oligoarticular JRA is defined by involvement of 4 or fewer joints; polyarticular JRA is defined by involvement of >4 joints (usually 10-20); and systemic-onset JRA is

defined by quotidian fevers during the first 6 weeks of the illness, almost always associated with a characteristic rash. Less than 1% of children with systemic-onset JRA develop chronic uveitis. Most children with uveitis have an oligoarticular onset.

Chronic uveitis may be detected at the time of initial diagnosis of arthritis; however, if not present at onset, it most often presents during the next 4 to 7 years. The period of highest risk is within 4 years of onset of arthritis, although the risk is never entirely absent. Eye involvement precedes involvement of the joints in approximately 5% of cases.

Children with JRA remain at risk of developing uveitis into adulthood. There are reports of uveitis diagnosed initially more than 20 years after onset of arthritis. The activity of the uveal inflammation does not parallel that of the joint disease.

Age

Children at greatest risk of developing uveitis are those with oligoarticular-onset JRA. The peak age of onset of arthritis in oligoarthritis is 1 to 5 years.

Immunogenetic and Serologic Markers

The serologic marker most strongly associated with chronic uveitis is the presence of antinuclear antibodies. Antinuclear antibodies are present in 65% to 90% of children with chronic uveitis and are a major risk factor for its development. They are usually detected in low to moderate titers on HEp-2 cells and are of unknown antigenic specificity. Rheumatoid factor is not usually present in children with JRA, including those with uveitis. Immunogenetic factors may predispose to the development of chronic uveitis. The associated alleles are located predominantly in the major histocompatibility complex (MHC) region on chromosome 6 and involve specificities in the human lymphocyte antigen (HLA)-DR, DP, and DQ regions.

Clinical Characteristics

The onset of ocular inflammation is insidious and asymptomatic in most young children. Because of the lack of symptoms or the cognitive recognition by the child, the exact time of onset of ocular involvement is frequently difficult to determine. This observation emphasizes the requirement for slit-lamp examination by an ophthalmologist at diagnosis of JRA and periodically thereafter.

Signs or symptoms in older children, rare as they are, may include a red eye, decreased vision, unequal pupils, ocular pain, and headaches and should prompt an urgent eye examination. Most cases of uveitis are bilateral (70% to 80%); unilateral disease may progress to bilateral involvement.

Data compiled before widespread therapy with methotrexate and tumor necrosis factor blockers indicated that the prognosis was good in 25% of cases, and 25% of children responded poorly to treatment and/or might require surgery for cataracts or glaucoma. Approximately 50% of patients required prolonged treatment for moderate to severe chronic inflammation; the visual prognosis in

these patients remained guarded. Early and aggressive treatment of intraocular inflammation has helped to reduce the morbidity of the ocular disease.

Frequency of Ophthalmologic Examinations in Children with JRA

The suggested frequency of ophthalmologic visits for children with JRA without known uveitis at diagnosis and during follow-up is presented in the table below. Once uveitis is diagnosed, the pediatric ophthalmologist will determine the frequency of examinations on the basis of response to therapy and complications. Because a substantial number of patients may have the eye disease before or shortly after their arthritis is diagnosed, they should have their initial eye examination within 1 month of the diagnosis of arthritis rather than waiting for the first available appointment.

Table: Frequency of Ophthalmologic Examination in Patients with JRA

Type	ANA	Age at onset, y	Duration of disease, y	Risk Category	Eye Examination Frequency, mo
Oligoarthritis or polyarthritis	+	≤6	≤4	High	3
	+	≤6	>4	Moderate	6
	+	≤6	>7	Low	12
	+	>6	≤4	Moderate	6
	+	>6	>4	Low	12
	-	≤6	≤4	Moderate	6
	-	≤6	>4	Low	12
	-	>6	NA	Low	12
Systemic disease (fever, rash)	NA	NA	NA	Low	12

ANA indicates antinuclear antibodies; NA, not applicable

Recommendations for follow-up continue through childhood and adolescence.

CLINICAL ALGORITHM(S)

None provided

EVIDENCE SUPPORTING THE RECOMMENDATIONS

TYPE OF EVIDENCE SUPPORTING THE RECOMMENDATIONS

The type of evidence supporting the recommendations is not specifically stated.

BENEFITS/HARMS OF IMPLEMENTING THE GUIDELINE RECOMMENDATIONS

POTENTIAL BENEFITS

Scheduled slit-lamp examinations by an ophthalmologist at specific intervals can detect ocular disease early, and prompt treatment can prevent vision loss.

POTENTIAL HARMS

Not stated

IMPLEMENTATION OF THE GUIDELINE

DESCRIPTION OF IMPLEMENTATION STRATEGY

An implementation strategy was not provided.

INSTITUTE OF MEDICINE (IOM) NATIONAL HEALTHCARE QUALITY REPORT CATEGORIES

IOM CARE NEED

Living with Illness
Staying Healthy

IOM DOMAIN

Effectiveness
Timeliness

IDENTIFYING INFORMATION AND AVAILABILITY

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ADAPTATION

Not applicable: The guideline was not adapted from another source.

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2006 May 1

GUIDELINE DEVELOPER(S)

American Academy of Pediatrics - Medical Specialty Society

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American Academy of Pediatrics

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FINANCIAL DISCLOSURES/CONFLICTS OF INTEREST

Not stated

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GUIDELINE AVAILABILITY

Electronic copies: Available from the American Academy of Pediatrics (AAP) Policy Web site.

Print copies: Available from American Academy of Pediatrics, 141 Northwest Point Blvd., P.O. Box 927, Elk Grove Village, IL 60009-0927.

AVAILABILITY OF COMPANION DOCUMENTS

None available

PATIENT RESOURCES

None available

NGC STATUS

This NGC summary was completed by ECRI on May 23, 2006. The information was verified by the guideline developer on June 9, 2006.

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