

Subspecialty Referrals for Pauciarticular Juvenile Rheumatoid Arthritis

Inocencio Antonio Cuesta, MD; Karen Kerr, RN, MSN; Pippa Simpson, PhD; James N. Jarvis, MD

Objectives: To examine referral patterns from primary care physicians for children with pauciarticular juvenile rheumatoid arthritis (JRA) and to determine whether children with pauciarticular JRA referred to pediatric rheumatologists differ in clinical presentation from children referred to other specialists.

Design: A retrospective records review of 49 patients with pauciarticular JRA was performed. Records were reviewed to determine the specialty of the referring physician and whether the children referred had symptoms and signs compatible with a synovitis at the time primary care was sought.

Setting: Inner-city tertiary pediatric rheumatology referral center.

Participants: Children with pauciarticular JRA.

Main Outcome Measures: Identification of referral patterns of primary care physicians. Associated mor-

bidity owing to JRA was ascertained at the time of referral.

Results: Most children with pauciarticular JRA (62%) were referred to orthopedic surgeons prior to referral for pediatric rheumatology care. No differences in clinical symptoms were seen between children referred to pediatric rheumatologists and those referred to orthopedic surgeons. Children referred initially to orthopedic surgeons were younger than those referred to pediatric rheumatologists.

Conclusion: A notable number of children with pauciarticular JRA are referred to orthopedic surgeons prior to the establishment of that diagnosis, even when such children present with unequivocal signs of synovitis. This may be owing to the misconception that arthritis is rare in preschool-aged children or to the difficulty of ascertaining the presence of synovitis in younger children.

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Editor's Note: So why did generalists (and especially general pediatricians) refer so many of these children to orthopedists? Was it access, availability, nonexposure to pediatric rheumatologists during training, or . . . ?

Catherine D. DeAngelis, MD

From the Division of Immunology and Rheumatology (Drs Cuesta and Jarvis and Ms Kerr), and the Department of Pediatrics (Dr Simpson), the Children's Hospital of Michigan, and the Department of Pediatrics (Dr Simpson) and Division of Immunology (Dr Jarvis), Wayne State University School of Medicine, Detroit, Mich. Dr Jarvis is now with the Department of Pediatrics, Oklahoma University Health Sciences Center, Oklahoma City.

JUVENILE RHEUMATOID arthritis (JRA) is a chronic disease of childhood characterized by inflammation of the synovial membranes of joints. Three subtypes (polyarticular, pauciarticular, and systemic onset) are recognized based on clinical presentation.^{1,2} As a group, the rheumatic diseases of childhood are among the most common chronic disorders of childhood. While reliable prevalence data are difficult to obtain, some estimates suggest that JRA is more common than better recognized illnesses such as diabetes mellitus or static encephalopathies.³ Therefore, it

is highly desirable that primary care physicians learn to recognize JRA and related disorders. The diagnosis of JRA is made almost entirely on the basis of findings from patient history and physical examination. Compatible historic features of chronic synovitis in children include morning stiffness, fatigue, and limping that is lessened by activity and worsened with rest. Pain can be conspicuously absent as a presenting complaint.⁴ The sine qua non in the diagnosis of any chronic form of arthritis is the presence of physical signs of inflamed synovial membranes in 1 or more joints. The presence of warm, thickened synovial membranes with or without effusion is the classic sign of synovitis in both adults and children.

Pediatric rheumatology is a relatively young subspecialty with board certification available only since 1992. Therefore, relatively few pediatric rheumatologists are practicing in the United States (149 in 1996,

PATIENTS AND METHODS

STUDY POPULATION

Patients were seen in the rheumatology clinic at the Children's Hospital of Michigan, Detroit, with initial visits occurring between January 1987 and May, 1996. Patients were identified by *International Classification of Diseases, Ninth Revision (ICD-9)*⁷ diagnostic coding for pauciarticular JRA, with each patient fulfilling the diagnostic criteria for the disease.¹

DESIGN

Each patient's history and findings from physical examination at the time of the initial visit were reviewed. A total of 49 patients were identified (40 girls; 9 boys). Of these, 45 (92%) were white; and 4 (8%), African American. Patient's ages ranged from 1 to 16 years at the time of diagnosis. The median age of JRA onset was 4.0 years (age range, 1-15 years).

DATA ANALYSIS

Records were examined to determine whether the child saw other specialists (eg, orthopedic surgeons or adult rheumatologists) prior to being evaluated at the Children's Hospital of Michigan rheumatology clinic. Records were reviewed to determine whether any of the following symptoms and signs were present at the time of evaluation by the physician who initially evaluated the patient (ie, prior to referral to orthopedic or other specialty care): morning stiffness, joint swelling without fever, or limping (and whether this complaint was lessened or worsened by activity). None of the 49 children had suffered trauma in the 24 hours preceding their initial evaluation by a physician for musculoskeletal complaints. The initial joint involved was noted as either an upper or lower extremity or both. The presence or absence of antinuclear antibodies was noted. Records were reviewed for the presence of uveitis at the time of diagnosis, hospitalizations, and casting and surgical procedures excluding arthrocentesis (eg, synovial biopsies). Board certification status was obtained from the referring physician. Insurance information was ascertained for each patient at the initial consultation and classified as managed care, commercial, Medicaid, and no insurance.

STATISTICAL ANALYSIS

Initial plotting of the variables of interest suggested a nonparametric distribution. Since data were skewed, we used the nonparametric Mann-Whitney *U* test to compare pairs of groups. For proportions, a χ^2 or Fisher exact test was used.

with $\geq 90\%$ practicing full time in academic medical centers).⁵ Forty-five medical schools in the United States have no pediatric rheumatologist and another 45 have only 1 pediatric rheumatologist.⁶ Thus, it seems likely that a considerable number of primary care physicians may have

Table 1. Referral Patterns and Board Certification*

Referral	Pediatrician Certified (n = 27)	Pediatrician Noncertified (n = 11)	Family Physician Certified (n = 4)	Family Physician Noncertified (n = 6)
Orthopedic surgeon	17 (63)	8 (73)	2 (50)	3 (50)
Rheumatologist	10 (37)	3 (27)	2 (50)	3 (50)

*Values given as number (percentage) of patients.

developed referral patterns that do not include pediatric rheumatologists as a "first line" of referral for children with musculoskeletal complaints.

An understanding of how and when primary care physicians use pediatric rheumatologists is essential in estimating future personnel needs in this area and, therefore, in planning how and where training programs in pediatric rheumatology can be supported. This study was undertaken to determine the referral patterns of pediatricians, family practitioners, and other subspecialty physicians with respect to the most common form of chronic synovitis in childhood: pauciarticular JRA. We specifically sought to determine how frequently children with this illness were referred to other subspecialists (eg, orthopedic surgeons) before the diagnosis was made and care was undertaken by rheumatologists, and whether there were predictable clinical patterns that resulted in referral to a pediatric rheumatologist rather than other specialists.

RESULTS

Records from 49 patients with pauciarticular JRA were examined. Of these children, 30 (62%) were referred to orthopedic surgeons before referral was made to either adult or pediatric rheumatologists. The other 19 (38%) were referred directly from the physician initially consulted to either pediatric or adult rheumatologists.

Regarding physicians' referral patterns for patients with pauciarticular JRA based on their practice specialty, of the 49 children, 38 (76%) were initially seen by pediatricians. Of these 38, 25 (63%) were referred initially to orthopedic surgeons. Of the remaining 13, 8 (24%) were referred to pediatric and 5 (13%) to adult rheumatologists. In contrast, family practitioners referred a higher percentage of their patients directly to pediatric rheumatologists. Of 10 patients initially seen by a family physician, 5 (50%) were referred to orthopedic surgeons. The remaining patients were referred either to pediatric (n = 4) or adult (n = 1) rheumatologists for further evaluation of their musculoskeletal complaints. One patient was seen in the emergency department and was referred to an orthopedic surgeon. Board certification status did not predict the referral patterns of either pediatricians or family physicians, as given in **Table 1**. That is, board certified pediatricians and family physicians were as likely to refer to orthopedic surgeons as noncertified physicians.

Table 2 gives the clinical features of the 49 patients evaluated. Most patients presented to their primary care physicians with signs and symptoms characteristic of

Table 2. Signs and Symptoms of Juvenile Rheumatoid Arthritis*

Physical Findings	Present	Not Present	Not Noted
Morning stiffness	46 (94)	1 (2)	2 (4)
Joint swelling without fever	47 (96)	2 (4)	...
Limp or stiffness	42 (86)	3 (6)	4 (8)
Limp lessened with activity	39 (78)	4 (8)	6 (14)
History of trauma within 24 h	...	49 (100)	...
Antinuclear antibodies	30 (60)	19 (40)	...

*Patient population was $N = 49$. Values are given as number (percentage) of patients. Ellipses indicate none.

chronic synovitis in children: morning stiffness, joint swelling, or limping that was lessened by activity and worsened with rest. Pain was conspicuously absent as a presenting complaint. All but 1 of the children had synovitis of the lower extremity (knee or ankle). The single child presenting with synovitis of the wrist was referred to an orthopedic surgeon. All but 2 of the children had joint swelling noted by their primary care physician at the time of presentation. Family physicians and pediatricians were not more likely to refer patients with any particular 1 of the 4 symptoms (limping, limping lessened with activity, morning stiffness, and joint swelling) to either an orthopedic surgeon or rheumatologist ($P > .05$).

Children referred by their primary care physicians to orthopedic surgeons were younger (median age, 2.0 years; age range, 1-12 years) compared with children referred to rheumatologists (median age, 6.5 years; age range, 1-15 years). This difference was statistically significant ($P < .038$). Uveitis was not present at the time of diagnosis. One child was placed in a cast, one had a synovial biopsy performed, and another was admitted to the hospital and given intravenous antibiotics.

Insurance status was analyzed as a separate variable determining referral patterns. Thirty-one of the patients were covered by traditional commercial health insurance plans. Of these, 18 (58%) were referred first to orthopedic surgeons and 13 (42%) to pediatric or adult rheumatologists. Twelve children were covered by the Michigan Medicaid program. Of these, 7 (58%) were referred to orthopedic surgeons and 5 (42%) to rheumatologists. Six children were in managed care organizations. Of these, 5 (83%) were referred to orthopedic surgeons while 1 was referred to a rheumatologist. A single child, who was referred initially to an orthopedic surgeon, carried no insurance coverage. Although the data from the managed care group are intriguing, they were not statistically significant when compared with the other groups, in all likelihood owing to the small numbers of health maintenance organization patients. Four African American children were identified in this study. Of these, 2 were referred to orthopedic surgeons and 2 to rheumatologists.

COMMENT

Pauciarticular JRA represents one of the most common chronic disease conditions in children. Although there has been substantial growth in the number of physi-

cians limiting their clinical practices to children with rheumatic disease, it is still reasonable to hypothesize that most children with this illness will be seen and evaluated by primary care physicians at the time of initial symptom development. In planning for future pediatric rheumatology workforce needs, it is therefore of considerable importance to know how and when primary care physicians seek the consultative services of pediatric rheumatologists compared with other specialists with expertise in the area of musculoskeletal problems in children. This is especially true in managed care environments where access to pediatric subspecialty care may be stringently limited.⁸

Our study demonstrated that children with pauciarticular JRA who are eventually referred for pediatric rheumatology subspecialty care are likely to have been evaluated by orthopedic surgeons prior to referral to pediatric rheumatologists. The reasons for this pattern were not immediately obvious. We found no differences in the presenting signs and symptoms between children referred to orthopedic surgeons and children referred to pediatric rheumatologists. In both groups, the presenting complaint was the one most commonly seen in children presenting with this illness: limping or morning stiffness lessened with activity and worsened with rest. Limping is a common complaint, particularly in preschoolers,⁹ and invites a broad differential diagnosis that includes malignant, mechanical, and infectious disease processes. We found the absence of verbalized pain to be an important clue in identifying those children who limp and who have pauciarticular JRA, as others have also reported.⁴

It is possible that primary care physicians perceive that orthopedic surgeons are better able to handle the breadth of potential diagnoses in a child who limps, especially trauma. Trauma, however, was as conspicuously absent in the histories of children sent to orthopedic surgeons as it was in children referred directly to pediatric rheumatologists. Thus, it is reasonable to ask whether the important clinical clue, that the limping or morning stiffness was lessened with activity and worsened with rest (a phenomenon sometimes referred to as *gelling*), was fully appreciated by the primary care physician. Similarly, joint swelling associated with warmth was noted with equal frequency between the 2 groups and was present in 96% of the children at the time of initial presentation to the primary care physician. The joint swelling associated with rheumatoid arthritis in adults and children represents proliferative synovial tissue, and its presence is the sine qua non for diagnosing active synovitis. Joint swelling can also be present in trauma (eg, traumatic hemarthrosis) although the patient's history and findings from physical examination usually distinguish such cases from children with pauciarticular JRA.

One significant difference between children referred to orthopedic surgeons and children referred directly to pediatric rheumatologists was the patient's age. Children referred to orthopedic surgeons were significantly younger than children referred to pediatric rheumatologists. This fact may point to the difficulty of detecting objective signs of synovitis in younger children. However, it may also reflect the misconception that arthritis is rare in preschoolers. This should not be so, as

pauciarticular JRA is, by and large, a disease of preschool-aged children, at least in white populations.¹⁰

We have considered that the greater availability of orthopedic surgeons might account for the preference of primary care physicians to use them instead of rheumatologists. However, the usual waiting time for an outpatient appointment in our clinic is between 1 and 6 days, and patients are almost always seen within 24 to 48 hours if there is direct telephone contact between the referring physician and one of the rheumatologists at our hospital. Therefore, a long lag time between referral and the actual patient visit seems an unlikely explanation for the data. The referral pattern may, however, reflect the preferences of adult rheumatologists rather than primary care physicians. A recent survey in the state of Washington demonstrated that adult rheumatologists strongly prefer not to care for children younger than 6 years.¹¹ Thus, it is possible that the primary care physicians chose orthopedic surgeons for children with pauciarticular JRA only after unsuccessful attempts to make a referral to an adult rheumatologist.

Specific findings on laboratory evaluation were non-discriminatory in determining whether children with pauciarticular JRA were referred to orthopedic surgeons or pediatric rheumatologists. This is predictable. There is no single laboratory finding that will allow practitioners to include or exclude JRA from the differential diagnosis of a child with limping and joint swelling. The IgM rheumatoid factors (detected by latex agglutination), which are sometimes useful when found in adults, have little diagnostic utility in children¹² as the results are positive in only a small fraction of children. Furthermore, children with arthritis who test positive for IgM rheumatoid factors have such prominent and diffuse synovitis on physical examination that the diagnosis is usually apparent.¹² Similarly, although the presence of antinuclear antibodies in children with established pauciarticular JRA is useful in identifying children at risk for developing chronic uveitis,¹³ their presence is by no means an indication that a child with musculoskeletal complaints has JRA or any other form of chronic rheumatism.¹⁴ Anywhere between 2%¹⁵ and 9%¹⁶ of healthy children may have antinuclear antibodies.

We conclude that primary care physicians frequently use the services of orthopedic surgeons in the evaluation of children subsequently shown to have pauciarticular JRA. Records review demonstrated that classic symptoms and signs of chronic synovitis were present in most of these children when seen by their primary care physicians. A better understanding of how and when primary care physicians use orthopedic surgeons and rheumatologists would be extraordinarily helpful in planning health care delivery in managed care environ-

ments. We also believe that greater awareness of the pediatric rheumatic diseases and the expertise of pediatric rheumatologists may reduce the multiple referrals that seem to be a common pattern for children with pauciarticular JRA.

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Reprints: James N. Jarvis, MD, Department of Pediatrics/Division of Rheumatology, Basic Science Educational Bldg No. 235A, Oklahoma University Health Sciences Center, Oklahoma City, OK 73104.

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