Acute Rheumatic Fever and Poststreptococcal Reactive Arthritis: Diagnostic and Treatment Practices of Pediatric Subspecialists in Canada

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ABSTRACT. ***Objective.*** We conducted a survey of pediatric specialists in rheumatology, cardiology, and infectious diseases to ascertain present Canadian clinical practice with respect to diagnosis and treatment of acute rheumatic fever (ARF) and poststreptococcal reactive arthritis (PSReA), and to determine what variables influence the decision for or against prophylaxis in these cases.

Methods. A questionnaire comprising 6 clinical case scenarios of acute arthritis occurring after recent streptococcal pharyngitis was sent to members of the Canadian Pediatric Rheumatology Association, and to heads of divisions of pediatric cardiology and pediatric infectious diseases at the 16 university affiliated centers across Canada.

Results. There is considerable variability with respect to diagnosis in cases of ReA following group A streptococcal (GAS) infection both within and across specialties. There is extensive variability regarding the decision to provide prophylaxis in cases designated as ARF or PSReA. Findings indicated that physicians are most comfortable prescribing antibiotic prophylaxis in the presence of clear cardiac risk and are less inclined to such intervention for patients diagnosed with PSReA. When prophylaxis was recommended for cases of PSReA, the majority of respondents prescribed longer term courses of antibiotics.

Conclusion. The lack of observed consistency in diagnosis and treatment in cases of reactive arthritis post-GAS infection likely reflects the lack of universally accepted criteria for diagnosis of PSReA and insufficient longterm data regarding carditis risk within this population. There is a need for clear definitions and treatment guidelines to allow greater consistency in clinical practice across pediatric specialties. (J Rheumatol 2001;28:1681–8)

Key Indexing Terms:
ACUTE RHEUMATIC FEVER POSTSTREPTOCOCCAL REACTIVE ARTHRITIS
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Reactive polyarthritis following Group A beta-hemolytic streptococcal (GAS) infection is among the cardinal manifestations of acute rheumatic fever (ARF) designated by Jones' criteria for diagnosis of the disease[1] (Table 1). This arthritis classically involves large joints, is migratory in 50% of cases, and usually responds to salicylates[2]. Other major symptoms of ARF include carditis, chorea, erythema marginatum, and subcutaneous nodules. While there appears to be consensus regarding the diagnosis of ARF according to Jones' criteria, there is controversy concerning the appropriate designation of reactive arthritis (ReA) in the absence of carditis, or other major criteria, and with supporting evidence of prior streptococcal infection. Several authors have suggested that poststreptococcal reactive arthritis (PSReA) represents a distinct clinical entity to be distinguished from ARF based on differences in both symptomatology and course, while others consider it to be part of the same spectrum of disease[3-7].

Review of the early literature (1938–1963) examining cases of "ARF without carditis" at initial presentation indicates a risk of carditis at recurrence or longterm followup of between 0 and 44%[8-14]. To the extent that polyarthritis may be the sole major manifestation of ARF, investigators have suggested that careful assessment for the presence of minor criteria and documentation of a recent GAS infection is required to prevent potential underdiagnosis of ARF[15]. Some patients designated as having PSReA do indeed fulfill Jones' criteria for ARF at initial presentation. It is argued that the term PSReA should be applied only to cases of

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Table 1. Diagnosis of acute rheumatic fever: 1992 Revised Jones' criteria — presence of 2 major or one major and 2 minor criteria indicates a high probability of ARF.

Major Manifestations	Minor Manifestations
Carditis	Arthralgia
Polyarthritis	Fever
Chorea	Elevated acute phase reactants
Erythema marginatum	Prolonged PR interval
Subcutaneous nodules	

Plus: supporting evidence of preceding streptococcal infection: increased titer of streptococcal antibodies, positive throat culture for Group A streptococcus, recent scarlet fever.

inflammatory arthropathy post-group A streptococcal infection that do not fulfill Jones' criteria for a diagnosis of ARF[15].

Accepted treatment for patients with ARF includes longterm antibiotic prophylaxis to prevent recurrences of ARF and carditis[12,16,17]. The risk of rheumatic heart disease for the child with PSReA has not been clearly determined, but there are infrequent reports of clinical carditis occurring with a subsequent streptococcal infection in patients initially diagnosed with PSReA (and not fulfilling Jones' criteria for ARF)[6,18,19]. There are also data suggesting a genetic link between PSReA and ARF as these patients appear to share the same B cell surface marker, D8/17[19-21]. Considerable controversy exists regarding the need for antibiotic prophylaxis in children with PSReA and no guidelines have yet been determined for this practice.

We conducted a clinical scenario based survey of pediatric specialists in rheumatology, cardiology, and infectious diseases to ascertain present Canadian clinical practice with respect to the diagnosis and treatment of children with ARF and PSReA, and to determine what variables influence the decision for or against antibiotic prophylaxis in these cases. Management of cases of PSReA (with insufficient Jones' criteria) was an area specifically targeted in the survey.

MATERIALS AND METHODS

A questionnaire was sent to members of the Canadian Pediatric Rheumatology Association (CPRA), and to the heads of divisions of pediatric cardiology and pediatric infectious diseases at the 16 university affiliated centers across Canada. Heads of infectious diseases and cardiology were asked to meet with the members of their divisions and respond by consensus.

The questionnaire comprised 6 distinct clinical scenarios of acute inflammatory arthritis occurring after a recent streptococcal pharyngitis. The scenarios were developed by consensus among the investigators (one infectious diseases specialist, 3 rheumatologists, and one cardiologist) and ranged from afebrile monoarthritis to classic ARF with carditis. Full descriptions of each scenario are presented in Table 2. For all scenarios respondents were instructed to assume definite serologic confirmation of recent streptococcal infection with rising titers of antistreptolysin-O (ASOT) and anti-DNase B. For each scenario physicians were asked for their clinical diagnosis, whether antibiotic prophylaxis would be recommended and for what duration, and the rationale for or against prophylaxis. In addition, physicians were asked to rate, in order of importance, the

Table 2. Clinical scenarios of acute arthritis following recent streptococcal pharyngitis with serologic confirmation.

Case 1 —	ARF with carditis: A 12-year-old girl presents with 1 week history of fever and migratory polyarthritis 3 weeks following the onset of streptococcal pharyngitis. She has a cardiac murmur consistent with mitral regurgitation (confirmed by echocardiography) and subcutaneous nodules. Her white blood cell count (WBC) is 17,000 and erythrocyte sedimentation rate (ESR, mm/h Wintrobe) is 55.
Case 2 —	Afebrile monoarthritis: An 8-year-old boy presents with an acute painful arthritis of his left knee and refusal to bear weight. He had streptococcal pharyngitis 10 days ago. He is afebrile and has no murmurs. The left knee is warm, effused, and tender (not felt to be a bacterial septic arthritis clinically). Complete blood cound (CBC) is normal and ESR is 47. Electrocardiogram (ECG) and echocardiogram are normal.
Case 3 —	Monoarthritis and positive family history of ARF*: Same clinical scenario as Case 2 above, but this patient has 2 family members with prior history of ARF, one of whom has cardiac sequelae.
Case 4 —	Monoarthritis and fever: Same clinical scenario as Case 2 above, but this patient has documented fever of 38.5˚C po for the last 3 days.
Case 5 —	Monoarthritis, recurrent episodes: A 6-year-old girl presents with an acutely painful left hip 1 week following onset of streptococcal pharyngitis. On examination she is afebrile and has pain and loss of range in her left hip. Cardiovascular examination is normal. WBC is 15,000 and ESR is 48. ECG and echocardiogram are normal. On further questioning, she has a history of streptococcal pharyngitis (culture proven) 3 or 4 times a year, and had a previous episode one year ago of hip pain following streptococcal pharyngitis with difficulty bearing weight that lasted one week.
Case 6 —	Afebrile migratory polyarthritis: A 15-year-old boy presents with migratory polyarthritis involving 4 joints, each one lasting about a week. This started 3 weeks following streptococcal pharyngitis. He has had no fever. On examination, there is no evidence of carditis, rash, or nodules. ECG and echocardiogram are normal.

*New diagnosis not requested for this scenario.

degree of influence of certain clinical features in determining the need for antibiotic prophylaxis in cases of clearly documented PSReA and ARF.

RESULTS

Participants. In total, 16 pediatric infectious disease centers, 16 pediatric cardiology centers, and 25 pediatric rheumatologists were asked to participate in the survey. Six (38%) infectious disease centers, 7 (44%) pediatric cardiology centers, and 23 (92%) pediatric rheumatologists returned the completed survey.

Diagnosis of ARF and PSReA. Table 3 presents a summary of diagnoses made by specialists for each clinical case scenario. Scenario 1 was included as a classic case of ARF with carditis with which other scenarios could be compared. A high degree of agreement was observed for scenarios involving afebrile monoarthritis (Scenario 2) and recurrent monoarthritis (Scenario 5), with 82% and 78% of physicians indicating a diagnosis of definite or probable PSReA for each of these cases, respectively. Greater variability in responses was observed within each of the remaining scenarios, with about one-quarter of respondents conferring a diagnosis of definite or probable ARF for cases involving monoarthritis with fever (Scenario 4) and afebrile migratory polyarthritis (Scenario 6). In these cases PSReA was diagnosed by about one-half of physicians and other diagnoses (e.g., septic arthritis for Scenario 4, arthritis not yet diagnosed) made up the remaining responses.

Although the small number of participants in each group precludes definite conclusions regarding the comparability of diagnostic practices across specialties, it appeared that, after case 1, cases 2 and 5 elicited the greatest degree of diagnostic consistency, with the majority of respondents in each group identifying these as cases of PSReA. No further consistent similarities or differences were discernible across the 3 groups with respect to diagnostic category.

Antibiotic prophylaxis. Table 4 presents data summarizing use of antibiotic prophylaxis. All physicians recommended prophylaxis for the case of classic ARF with carditis (Scenario 1). In cases where the diagnosis was less clearly defined, however, family history of ARF appeared to exert the greatest influence on the decision to give prophylaxis, with 47% of specialists recommending antibiotic intervention in case 3. About one-third of physicians prescribed antibiotic prophylaxis in scenarios involving monoarthritis with fever (Scenario 4), recurrent episodes of monoarthritis (Scenario 5), or afebrile migratory polyarthritis (Scenario 6). Afebrile monoarthritis without a family history of ARF (Scenario 2) was the least likely scenario to elicit a recommendation for prophylaxis, with 19% of respondents choosing antibiotic intervention. Despite considerable variability in responses both within and across specialties, in general, rheumatologists appear to prescribe antibiotic prophylaxis for a diagnosis of PSReA more often than infectious disease specialists, who prescribe more often than cardiologists.

Table 3. Diagnoses for clinical case scenarios by pediatric specialty.

Case Scenarios	Infectious Diseases, n* (%)	Cardiology, n* (%)	Rheumatology, n** (%)	All Specialties Combined, n*** (%)
Case 1: (n = 36) ARF with carditis				
ARF	6 (100)	7 (100)	23 (100)	36 (100)
PSReA	0 (0)	0 (0)	0 (0)	0 (0)
Other diagnosis	0 (0)	0 (0)	0 (0)	0 (0)
Case 2: (n = 34) Afebrile monoarthritis				
Definite/probable ARF	0 (0)	0 (0)	0 (0)	0 (0)
Definite/probable PSReA	3 (50)	4 (67)	21 (96)	28 (82)
Other diagnosis	3 (50)	2 (33)	1 (4)	6 (18)
Case 3: Scenario 2 + family history — diagnosis not requested on survey				
Case 4: (n = 29) Monoarthritis and fever				
Definite/probable ARF	0 (0)	3 (43)	4 (21)	7 (24)
Definite/probable PSReA	1 (33)	2 (29)	12 (63)	15 (52)
Other diagnosis	2 (67)	2 (29)	3 (16)	7 (24)
Case 5: (n = 32) Monoarthritis, recurrent episodes				
Definite/probable ARF	0 (0)	0 (0)	1 (5)	1 (3)
Definite/probable PSReA	4 (80)	5 (71)	16 (80)	25 (78)
Other diagnosis	1 (20)	2 (29)	3 (15)	6 (19)
Case 6: (n = 32) Afebrile migratory polyarthritis				
Definite/probable ARF	4 (80)	0 (0)	5 (25)	9 (28)
Definite/probable PSReA	1 (20)	3 (43)	13 (65)	17 (53)
Other diagnosis	0 (0)	4 (57)	2 (10)	6 (19)

*Refers to number of infectious disease and cardiology centers, **refers to individual rheumatologists, ***refers to infectious disease and cardiology centers combined with individual rheumatologists.

Table 4. Percentage of respondents recommending antibiotic prophylaxis and recommended duration of prophylaxis by clinical case scenario and pediatric specialty.

Case Scenarios	Infectious Diseases, n* (%)	Cardiology, n* (%)	Rheumatology, n** (%)	All Specialties Combined, n*** (%)†
Case 1: Recommend antibiotic prophylaxis	6 (100)	7 (100)	23 (100)	36 (100)
Lifelong/indefinitely	1 (17)	1 (14)	12 (52)	14 (39)
Early adulthood or 3–5 yrs	5 (83)	6 (86)	9 (39)	20 (55)
5 yrs then reassess	0 (0)	0 (0)	1 (4)	1 (3)
3–6 mo	0 (0)	0 (0)	1 (4)	1 (3)
Case 2: (n = 8) Recommend antibiotic prophylaxis	2 (40)	0 (0)	5 (23)	7 (19)
Lifelong/indefinitely	0 (0)	0 (0)	0 (0)	0 (0)
Early adulthood or 3–5 yrs	1 (50)	0 (0)	4 (67)	5 (62)
5 yrs then reassess	0 (0)	0 (0)	1 (17)	1 (13)
3–6 mo	1 (50)	0 (0)	1 (17)	2 (25)
Case 3: (n = 16) Recommend antibiotic prophylaxis	2 (40)	3 (43)	12 (57)	17 (47)
Lifelong/indefinitely	0 (0)	0 (0)	0 (0)	0 (0)
Early adulthood or 3–5 yrs	1 (50)	3 (100)	8 (73)	12 (75)
5 yrs then reassess	0 (0)	0 (0)	1 (9)	1 (6)
3–6 mo	1 (50)	0 (0)	2 (18)	3 (19)
Case 4: (n = 11) Recommend antibiotic prophylaxis	1 (20)	3 (43)	9 (43)	13 (36)
Lifelong/indefinitely	0 (0)	0 (0)	2 (29)	2 (18)
Early adulthood or 3–5 yrs	1 (100)	3 (100)	4 (57)	8 (73)
5 yrs then reassess	0 (0)	0 (0)	0 (0)	0 (0)
3–6 mo	0 (0)	0 (0)	1 (14)	1 (9)
Case 5: (n = 12) Recommend antibiotic prophylaxis	2 (40)	1 (14)	9 (45)	12 (33)
Lifelong/indefinitely	0 (0)	0 (0)	1 (11)	1 (8)
Early adulthood or 3–5 yrs	1 (50)	1 (100)	8 (89)	10 (83)
5 yrs then reassess	0 (0)	0 (0)	0 (0)	0 (0)
3–6 mo	1 (50)	0 (0)	0 (0)	1 (8)
Case 6: (n = 13) Recommend antibiotic prophylaxis	1 (17)	1 (14)	8 (36)	10 (28)
Lifelong/indefinitely	0 (0)	0 (0)	1 (13)	1 (8)
Early adulthood or 3–5 yrs	1 (33)	2 (100)	6 (75)	9 (69)
5 yrs then reassess	1 (33)	0 (0)	0 (0)	1 (8)
3–6 mo	1 (33)	0 (0)	1 (13)	2 (15)

*Refers to number of infectious disease and cardiology centers, **refers to individual rheumatologists, ***refers to infectious disease and cardiology centers combined with individual rheumatologists. †The number of respondents recommending antibiotic prophylaxis for each scenario differs from the total number providing a duration in instances where respondents indicated that they were "not sure" if they would provide prophylaxis but indicated a duration of treatment or in instances where respondents indicated that they would provide prophylaxis but did not indicate for how long.

Rationales for and against prophylaxis. Table 5 indicates considerable inter and intra-specialty variability with respect to rationales for and against prophylaxis, and within the context of very small sample sizes, any assertions regarding possible trends by group is impossible. When specialties were considered together, prevention of recurrence and carditis emerged as the most frequently stated reason for advocating antibiotic prophylaxis in most cases. In case 3, family history of ARF/genetic predisposition took precedence by physicians in all specialties. This clinical factor was noted by physicians to place children at increased risk for disease recurrence.

The most common reason cited for taking a decision not to recommend antibiotic prophylaxis was insufficient Jones criteria for a diagnosis of ARF. The absence of data proving efficacy of such intervention in cases of PSReA and the absence of cardiac involvement were also provided as reasons not to prescribe antibiotic prophylaxis.

Recommended duration of antibiotic prophylaxis. As Table 4 indicates, 94% of physicians suggested longterm antibiotic administration (to early adulthood or lifelong) in Case 1 of ARF with carditis. Among those who recommended antibiotic prophylaxis for the remaining scenarios, the duration also tended to be prolonged with between 75% and 92% of respondents suggesting that prophylaxis last 3 to 5 years or longer. In contrast, brief courses of antibiotics were advocated least often, only 8% to 25% of respondents suggesting a duration of treatment between 3 and 6 months for scenarios 2 through 6.

Factors influencing the decision to recommend antibiotic prophylaxis for PSReA. Table 6 summarizes the extent to which respondents considered various clinical features

Table 5. Rationales provided for and against prophylaxis by clinical case scenarios and pediatric specialty.

Case Scenarios	Infectious Diseases n* (%)	Cardiology, n* (%)	Rheumatology, n** (%)	All Specialties Combined, n*** (%)
Case 1				
Rationale for prophylaxis (n = 33)				
— Prevent recurrence and carditis	7 (100)	6 (100)	20 (100)	33 (100)
Case 2				
Rationale for prophylaxis (n = 6)				
— Prevent recurrence and carditis	1 (50)	0 (0)	4 (100)	5 (83)
— Diagnosis of PSReA	1 (50)	0 (0)	0 (0)	1 (17)
Rationale against prophylaxis (n = 19)				
— Does not fulfill criteria for ARF	1 (33)	6 (86)	8 (89)	15 (79)
— No proven efficacy	2 (67)	0 (0)	1 (11)	3 (16)
— Other	0 (0)	1 (14)	0 (0)	1 (5)
Case 3				
Rationale for prophylaxis (n = 16)				
— FH/genetic predisposition — increased risk	1 (50)	2 (67)	8 (73)	11 (69)
— Prevent recurrence and carditis	0 (0)	1 (33)	1 (9)	2 (12)
— Other	1 (50)	0 (0)	2 (18)	3 (19)
Rationale against prophylaxis (n = 8)				
— Does not fulfill criteria for ARF	1 (50)	2 (50)	1 (50)	4 (50)
— No evidence of cardiac involvement	0 (0)	2 (50)	1 (50)	3 (38)
— Other	1 (50)	0 (0)	0 (0)	1 (12)
Case 4				
Rationale for prophylaxis (n = 10)				
— Prevent recurrence and carditis	2 (100)	1 (33)	2 (33)	4 (40)
— Diagnosis tends toward/meets ARF criteria	0 (0)	2 (67)	3 (50)	5 (50)
— Other	0 (0)	0 (0)	1 (17)	1 (10)
Rationale against prophylaxis (n = 11)				
— No proven efficacy	2 (100)	0 (0)	0 (0)	2 (18)
— Does not fulfill criteria for ARF	0 (0)	1 (25)	2 (40)	3 (27)
— No evidence of cardiac involvement	0 (0)	3 (75)	2 (40)	5 (46)
— Other	0 (0)	0 (0)	1 (20)	1 (9)
Case 5				
Rationale for prophylaxis (n = 9)				
— Prevent recurrence and carditis	1 (50)	1 (100)	4 (67)	6 (67)
— Other	1 (50)	0 (0)	2 (33)	3 (33)
Rationale against prophylaxis (n = 12)				
— Does not fulfill criteria for ARF	1 (50)	4 (67)	1 (25)	6 (50)
— No proven efficacy	1 (50)	0 (0)	0 (0)	1 (8)
— Other	0 (0)	2 (33)	3 (75)	5 (42)
Case 6				
Rationale for prophylaxis (n = 7)				
— Prevention of recurrence and carditis	1 (100)	1 (100)	1 (20)	3 (43)
— Diagnosis tends toward/meets ARF criteria	0 (0)	0 (0)	3 (60)	3 (43)
— Other	0 (0)	0 (0)	1 (20)	1 (14)
Rationale against prophylaxis (n = 7)				
— Does not fulfill criteria for ARF	0 (0)	3 (75)	2 (67)	5 (71)
— Other	0 (0)	1 (25)	1 (33)	2 (29)

*Refers to number of infectious disease and cardiology centers, **refers to individual rheumatologists, ***refers to infectious disease and cardiology centers combined with individual rheumatologists. FH: family history.

important or very important in determining the need for antibiotic prophylaxis for ARF and PSReA. Ranked in order of importance, they were arranged as follows: (1) presence of carditis; (2) recurrent episodes of PSReA; (3) presence of polyarthritis; (4) presence of fever; (5) presence of family history of ARF. However, it is interesting that, after carditis, all clinical features appeared to exert a comparable degree of influence, with about half of all specialists considering the specified factors important or very important in deciding to institute antibiotic prophylaxis. The influence a positive family history of ARF appeared to have in the case scenarios suggests that the relative importance of this factor may be enhanced within a clinical setting.

Overall, infectious disease specialists considered each of

Table 6. Percentage of respondents considering factors important or very important in determining necessity for antibiotic prophylaxis in PSRA.

Factor	Infectious Disease, n* (%)	Cardiology, n* (%)	Rheumatology, n** (%)	All Specialties Combined, n*** (%)
Presence of polyarthritis (n = 35)	5 (83)	5 (71)	8 (36)	18 (51)
Presence of fever (n = 34)	4 (67)	3 (50)	9 (41)	16 (47)
Presence of carditis (n = 35)	6 (100)	7 (100)	22 (100)	34 (100)
Presence of family history of ARF (n = 35)	3 (50)	2 (29)	11 (50)	16 (46)
Recurrent episodes of PSReA (n = 33)	4 (80)	1 (17)	13 (59)	18 (55)

*Refers to number of infectious disease and cardiology centers, **refers to individual rheumatologists, ***refers to infectious disease and cardiology centers combined with individual rheumatologists.

the various clinical factors more important than rheumatologists, who considered them more important than cardiologists. Cardiologists appeared notably less influenced by a family history of ARF or by recurrent episodes than either of the other specialties.

Same spectrum or separate entities. About 55% of physicians were of the opinion that PSReA and ARF should be considered part of the same spectrum of disease, 18% considered them to be separate entities, and another 18% stated that they were not sure how ARF and PSReA should be classified. The remaining 9% suggested that they should be considered both separate entities and part of the same spectrum of disease, as viewing ARF and PSReA as falling along the same clinical spectrum does not necessarily mean that treatment requirements are the same for both conditions.

DISCUSSION

It is clear from the assessment of current Canadian clinical practice that there exists considerable variability with respect to the diagnosis of ARF/PSReA both within and across specialties in infectious disease, cardiology, and rheumatology. The variability within the infectious disease and cardiology divisions is particularly noteworthy, insofar as responses from these centers represent consensus within their groups. To the extent that the process of arriving at agreement likely resulted in an averaging of opinions we would have expected a greater degree of consistency within these specialties.

There also appears to be extensive variability regarding the decision to institute antibiotic prophylaxis in patients with PSReA. Not surprisingly, findings indicated that physicians are most comfortable prescribing antibiotic prophylaxis in the presence of clear cardiac risk and are less inclined to such intervention for diagnoses of PSReA. Interestingly, in cases where prophylaxis was recommended for PSReA, the majority of respondents adopted longer term courses of at least 3 to 5 years, or longer. The lack of observed consistency in diagnostic and treatment decisions may reflect current problems in the definition and nomenclature of PSReA as well as the fact that there is insufficient data regarding longterm cardiac outcome within this population. Clearly, there is a need for unambiguous definitions and treatment guidelines as well as further research aimed at determining carditis risk associated with PSReA. The variability in item response rates, highest among rheumatologists who had variable unanswered questions around cases 2, 4, 5, and 6, further highlights the need for clearer guidelines based on the available data.

We suggest (as others have[15]) that to avoid confusion in clinical diagnosis and nomenclature the term PSReA be reserved for those cases of well documented poststreptococcal inflammatory arthritis that do not fulfill Jones criteria for a diagnosis of ARF. In addition, patients presenting with polyarthritis and sufficient minor criteria to fulfill Jones' criteria can be diagnosed with ARF (without carditis) and be given prophylaxis accordingly, regardless of the duration of arthritis or response to nonsteroidal agents.

That ARF and PSReA may be conceptualized as part of the same spectrum of disease is supported by evidence that patients not fulfilling Jones' criteria initially may develop cardiac sequelae with recurrences. Moreover, the immunogenetic marker D8/17, which is found to be present in the majority of patients with acute rheumatic fever, has also been identified in children with PSReA[19,21]. Zemel and colleagues report 8 of 11 (73%) patients with PSReA to be D8/17 positive versus only 17% of controls. However, in a recent article Ahmed, *et al* report that while PSReA patients demonstrated a higher frequency of HLA DRB1*01 alleles, ARF patients had a higher frequency of DRB1*06 compared to controls[22].

The paucity of data on the exact risk of carditis and longterm cardiac sequelae in a given clinical situation among children with PSReA has resulted in a lack of agreement regarding what course of antibiotics, if any, is appropriate for this population[6,18,19]. This confusion, clearly reflected in clinical practice, speaks to the need for the development of practical guidelines to aid physicians in

making treatment decisions. The risk of cardiac sequelae among PSReA patients may be deemed too small to warrant standard ARF prophylaxis, and thus remains to be ascertained as further information becomes available.

The American Heart Association and the American Academy of Pediatrics Red Book guidelines suggest that prophylaxis be considered for up to one year in cases of PSReA and then suspended if carditis is not observed[23,24]. Others have supported the use of antibiotic prophylaxis similar to that prescribed for ARF patients until cardiac risks are further delineated[7,18]. The Canadian Pediatric Society statement on poststreptococcal arthritis[25] suggests antibiotic prophylaxis may be considered for all cases of PSReA and discontinued after 3 months if there is no evidence of carditis. The prevailing view among physicians surveyed who advocated for antibiotic prophylaxis in patients with PSReA was to institute a longer term course of antibiotics. This may be because of data indicating that most recurrences in patients with ARF occur within 3 to 5 years after the initial episode.

Evaluating the significance of "silent carditis" (patients with echographic evidence of valvular involvement without auscultatory findings) represents another area of controversy. There are no universally accepted criteria for echographic diagnosis of carditis at this time and the longterm cardiac outcome of patients with subclinical carditis is unknown. Until such criteria are established and additional prognostic value is confirmed, the American Heart Association does not recommend the use of echographic evidence of carditis in the absence of auscultatory findings as a criterion for ARF[26].

Insofar as PSReA may be considered a "forme fruste" of ARF, in our practice, longer term prophylaxis is carefully considered in all cases of PSReA and the decision to initiate treatment is individualized depending on the specific circumstances of the case. We have found it helpful to consider where along the spectrum of PSReA–ARF a particular case lies, ranging from afebrile monoarthritis through monoarthritis with family history/monoarthritis and fever/recurrent episodes of PSReA, afebrile polyarthritis, polyarthritis with fever, ARF without carditis, and to ARF with carditis. Depending on the specific additional risk factors that are present, we may be less inclined to recommend prophylaxis for cases at the far left of the continuum and are more inclined to prescribe prophylaxis for those approaching the far right. Cases fulfilling Jones' criteria should be diagnosed with ARF and treated accordingly. In addition to assessment for the presence of minor criteria and careful interpretation and documentation of antistreptolysin-O (and one other streptococcal antibody) results to ensure unequivocal documentation of recent streptococcal infection, physicians should consider factors such as the child's age, risk of exposure to streptococcus, access to medical care, and family history when deciding whether to recommend antibiotic prophylaxis. Finally, a discussion with the family regarding the potential risks and benefits associated with and without prophylaxis should be included as part of the decision making process.

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