Access to Pediatric Rheumatology Care — A Major Challenge to Improving Outcome in Juvenile Idiopathic Arthritis

Juvenile idiopathic arthritis (JIA) is a heterogeneous group of diseases and is one of the most common chronic rheumatic diseases in children. Improving the outcome for children with JIA remains an important goal, with current management of severe JIA involving increasingly aggressive immunosuppressive agents.

Undoubtedly, improving outcome in JIA depends on the availability of effective treatments, but if a child presents late in their disease course or receives inappropriate treatment or does not have access to treatment at all, then outcome is likely to be suboptimal. There is an increasing body of evidence (Table 1) that many children with JIA have a prolonged interval from disease onset to pediatric rheumatology care, and despite there being no published data from developing countries, anecdotal observations suggest that poor access to optimal care is likely to be a global issue.

The longterm impact of delay is unknown, but a longer interval from disease onset to definitive treatment is likely to adversely affect clinical outcomes. In one study, many children with delay (defined as > 10 weeks from symptom onset to first pediatric rheumatology assessment) had prolonged untreated active disease, many presented with multiple restricted joints, none had been referred for eye screening (to detect chronic anterior uveitis), and the median interval from onset of symptoms to starting methotrexate was 10 months. Such delay is likely to have a profound effect on the potential for functional disability and psychosocial impact as a result of poorly controlled symptoms and the family living with an uncertain diagnosis.

The article by Shiff, et al in this issue of The Journal describes a large incident cohort of children with JIA from across Canada, again reporting prolonged interval to pediatric rheumatology care, but also describes factors that may hinder or facilitate referral. Both this and many other studies refer to children presenting to pediatric rheumatology services, and there are no data on comparative outcomes between children managed by specialist and nonspecialist centers. However, inequity in access to most appropriate care exists, with reports of children with incident JIA being subjected to multiple, often invasive and unnecessary investigations (such as arthroscopy or synovial biopsy) before a pediatric rheumatology referral and this likely contributes to the reported delay; further, anecdotally, children with JIA are managed by doctors or other healthcare professionals who are not optimally trained or resourced to deliver best practice. The British Society for Paediatric and Adolescent Rheumatology (BSPAR) Standards of Care for Children with JIA are a step in the right direction for equitable access to high quality care, with emphasis on early recognition of JIA and prompt referral (target of within 10 weeks of disease onset), and highlight the need for high quality clinical trials to increase the evidence base to inform best practice; further the BSPAR Position Statement describes the training and experience required for members of the pediatric rheumatology specialist team.

Although Shiff, et al present limited data on the referral pathways, their findings are important as they corroborate other studies that give some insight into the complexity of patient journeys to pediatric rheumatology care. We propose that barriers and drivers to care can be divided into broad themes, as follows.

Physical Factors

Physical factors inherent in the disease influence the time to referral. For example, the ill child with severe JIA subtype who presents with raised inflammatory markers, often with fever or acute limp, is more likely to present quickly to healthcare; whereas more indolent presentations may be overlooked, such as the well child with normal inflammatory markers and intermittent limp (a common finding in oligoarticular JIA) or the child with chronic heel pain due to enthesitis, and it will often take longer for the diagnosis to be made. Differentiating early forms of JIA from reactive or transient forms of inflammatory arthritis remains a diagnostic challenge. Screening questionnaires have been described but lack sensitivity and specificity to detect JIA.

See Factors associated with longer time to specialist access for Canadian children with JIA, page 2415
Table 1. Studies reporting interval from onset of symptoms to pediatric rheumatology care. Adapted from Foster, et al, Rheumatology 2010;49:401-3.

<table>
<thead>
<tr>
<th>Study</th>
<th>Country</th>
<th>n</th>
<th>Time from Onset to First Pediatric Rheumatology Assessment median, mo (range, yrs)</th>
<th>Type of Study</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tzaribachev, 200921</td>
<td>Germany</td>
<td>132</td>
<td>7.2 (0–1.67)</td>
<td>Retrospective cohort, regional service</td>
</tr>
<tr>
<td>Shiff, 200929</td>
<td>Canada</td>
<td>35</td>
<td>4.2 (0–13.6)</td>
<td>Prospective cohort, regional service</td>
</tr>
<tr>
<td>Adib, 200812</td>
<td>UK</td>
<td>507</td>
<td>4.6 (0–2.75)</td>
<td>Prospective cohort, multicity center</td>
</tr>
<tr>
<td>Foster, 20077</td>
<td>UK</td>
<td>152</td>
<td>5 (0–7.9)</td>
<td>Retrospective cohort, regional</td>
</tr>
<tr>
<td>Shapiro, 200730</td>
<td>Canada</td>
<td>836</td>
<td>2.57 (0–5)</td>
<td>Retrospective cohort, regional</td>
</tr>
<tr>
<td>Arguedas, 200231</td>
<td>Costa Rica</td>
<td>47</td>
<td>3.7 (0–2.75)</td>
<td>Prospective cohort, population based</td>
</tr>
<tr>
<td>Guillaume, 200032</td>
<td>France</td>
<td>207</td>
<td>3 (0–7.98)</td>
<td>Retrospective, longitudinal, regional; oligoarthritis-onset JIA only</td>
</tr>
<tr>
<td>Minden, 200033</td>
<td>Germany</td>
<td>171</td>
<td>10 (0–7.7)</td>
<td>Retrospective, longitudinal, regional</td>
</tr>
<tr>
<td>Manners, 199913</td>
<td>Australia</td>
<td>42</td>
<td>10 (0–4)</td>
<td>Retrospective cohort, regional</td>
</tr>
<tr>
<td>Andersson Gare, 199534</td>
<td>Sweden</td>
<td>124</td>
<td>3.5 (0–3.4)</td>
<td>Prospective cohort, population based</td>
</tr>
</tbody>
</table>

early14; in the absence of a diagnostic test, careful clinical assessment, knowledge, and exclusion of other diagnoses remain key to early recognition of JIA.

Recognition of JIA
Pathways of care for children with suspected JIA are not reported in detail by Shiff, et al, but undoubtedly they are complex and vary across healthcare systems. Children with JIA often present to healthcare professionals who are not expert in pediatric rheumatology; many doctors in family practice and hospital care (such as general pediatrics and orthopedics) are not confident in their musculoskeletal assessment of children15,16 — perhaps not surprising, given that pediatric musculoskeletal medicine is not core teaching in medical schools17,18, which may contribute to the reported delay in JIA and other conditions presenting with musculoskeletal features19,20. Notably, children with JIA presenting via orthopedics are reported to have the longest interval to pediatric rheumatology care7,13,21. Clearly, improved education and training to improve awareness and skills to recognize JIA are important22; these should be acquired by all doctors who are likely to encounter children (especially doctors working within orthopedics, emergency medicine, general pediatrics, and family medicine); and such training needs to start at medical school.

The development of a simple validated musculoskeletal screening examination (pGALS25) increasingly taught to medical students and in the UK, the introduction of competency frameworks for postgraduate training for all general pediatricians with musculoskeletal themes incorporated in the mandatory assessments (www.rcpch.ac.uk/training) will hopefully facilitate early recognition of JIA.

Healthcare Provision
Conditions of healthcare provision, such as the geographical distance to pediatric rheumatology specialist services and the availability of medicines, are major influences on access to high quality care. In the study by Shiff, et al across Canada, long distance to the nearest pediatric rheumatology clinic was not a significant factor to explain the delay, although this finding conflicts with data from Germany21 and from the US, where the greater distance from pediatric rheumatology services, the more likely services are delivered by adult rheumatologists24. The paucity of appropriately trained pediatric rheumatologists and multidisciplinary teams24 is likely to contribute to inequity in delivery of appropriate care, and increased recruitment to the specialty is needed. Access to appropriate care may be limited by healthcare costs such as in the USA, where children with JIA covered by basic healthcare insurance are less likely to have eye screening25. Biological agents are not available in many parts of the world, and may be subject to “rationing” based on cost-effectiveness (www.nice.org.uk), and availability may be geographically dependent on local health budgets.

Sociocultural Factors
Sociocultural factors may influence health-seeking behavior. Shiff, et al report that children of South Asian ethnicity and parents with higher levels of education are likely to present more quickly and suggest that this may reflect greater parent empowerment or may be due to ethnic variation in the epidemiology of severe JIA subtypes. Such observations are important and need to be explored further; we can learn much from parent and child experiences. There is increasing need, given the great advances in management, to understand the factors that drive or hinder the referral to pediatric rheumatology. Irrespective of the ultimate diagnosis, the visibility, persistence, and perceived severity of symptoms are central to a parent’s actions around help-seeking behavior26. Parents will notice a problem, but may not be able to judge the actual severity and need to seek expert advice27. In consultations with health professionals, parents often act as advocates for their sick child26, and have to balance being seen as responsible and vigilant against being labeled as overly neurotic, protective, or even exploitative.
When parents of children with cancer first reported that their child was unwell, their concerns were often disregarded and they struggled to obtain further investigations and a diagnosis of cancer\textsuperscript{28}. Anecdotally, parents of children with JIA are sometimes spurred on to seek healthcare by observations from teachers or nursery workers who may have noticed altered function in the child during school or play activities. Centrally, the inequalities of referrals to appropriate care often stem from inequalities of knowledge and experience of the health professionals to recognize JIA; experience with a prior case or contact with a specialist service, or both, are likely key drivers in triggering concern and a prompt referral.

In summary, while the reasons for delay, inequity, and inappropriate care are likely to be multifactorial, they are increasingly important to address as a major challenge to improve outcome for children with JIA. More work is required to identify and understand the barriers and drivers in the referral process, identify and address the training needs of doctors likely to encounter children presenting with suspected JIA, and identify strategies to empower parents and caregivers to seek appropriate healthcare attention.

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REFERENCES


