

EDITORIAL

Oligoarticular JIA: Is it a benign disease?

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I see the issues of oligoarticular JIA in the context of the differences between “textbook answers” and “clinical reality.” We all often refer to the need for published and refereed information. What better guarantee of “truth” can there be than peer review and publication? Yet I believe that evidence-based medicine can have its limits. For example, which of us publishes our mistakes? Or repeat the same mistake several times to prove that the unfortunate outcome was not just a coincidence? Despite admonitions for journals to publish negative research results, those brave or foolish enough to attempt to publish such information so that others might learn from this mistakes often have these efforts rejected.

Physicians who have been in clinical practice for a prolonged period may become aware that there is a body of information which has never been formally codified in journals or textbooks, but is often accepted as “truth.” It may range from the simple statement by an attending to a fellow that, “We use or don’t use that approach here,” to an informal comment at a national meeting by a fellow physician who says simply, “We have had the same experience many times. So we just don’t do that anymore.” Yet textbooks and the literature may not mention these observations at all.

Our knowledge of oligoarticular JIA (persistent and extended) may be one of the best examples of this type of information. After all, a few textbooks and informational resources for parents may still group HLA B27-positive, late onset oligoarticular JIA in a teenager with a 3 year old young child with oligoarticular JIA who has a single swollen knee and who is ANA-positive and HLA-B27 negative. In practice, most of us believe that a HLA-B27 positive teenager with arthritis as having a totally different disease (spondyloarthropathy) than the 3 year old with arthritis of one knee (oligoarticular JIA). Over-reliance on “textbook information” can lead to problems in the clinical setting.

Like many of us, I’ve occasionally seen a child for a second opinion who has been diagnosed to have oligoarticular JIA and who is not doing well. The parents have been told, have read, believe, and hope that the disease is self-limited and mild. They may experience a disconnect when their rheumatologist makes an assessment that the disease is no longer benign and wants to start methotrexate, a drug they hear might have a lot of possible side effects. The parents may wonder how the doctor can recommend methotrexate when the child has a benign disease that they are going to grow out of. Often the answer may be that the child really does need to be on methotrexate. Their original doctor may have lost some credibility by not have advising the parents initially and frequently thereafter that not all pauciarticular children do well.

It's important to be clear. True oligoarticular JIA, persistent or extended, is not always a benign disease. Every textbook agrees the most common complications of true oligoarticular JIA are leg length discrepancy and uveitis. But let's not forget that the typical case of oligoarticular JIA is a young child (typically 2 – 6 years of age) with a rapidly developing body image, and a variety of important physical and mental developmental milestones to accomplish. Impose pain, swelling, and limitation of motion, morning stiffness and gel effect in this setting and there may be nothing benign about the condition at all. It is likely that the arthritis had been ongoing for weeks to months before the child reached a pediatric rheumatologist. Even if no bony overgrowth is present, these effects will have a significant psychosocial impact and a developmental impact if the arthritis is left untreated. When bony overgrowth has developed because the disease has been untreated for too long there will be an aberrant gait, possible flexion contractures, and perhaps the need for a lift. These may be minor issues to the physician who is caring for a possibly mild to minimal arthritis problem but may be major issues for the family and child which can have life long consequences.

The consequences of uveitis and other joint involvement are even more profound. Fortunately most children with oligoarticular JIA do not develop uveitis, but for those who do develop it may have a major, life-changing complication. Yes, minor uveitis may respond well to topical steroids. But ever try putting drops in a small child's eyes four to six times a day? It's often a major stress. Are all the struggling and worry and exasperation that may transpire benign for the child or parents? Not likely. We are not even considering the consequences for children who do have visual damage. The articles in this issue of PROJ illustrate other systemic disease that may occur in a child with oligoarticular JIA. The clinician should watch for involvement of other joints, even the TMJ and atlo-epistropheal.

What does all this mean? I would suggest that if we take oligoarticular JIA disease lightly, we may convey the wrong information to the family, verbally and non-verbally. If we seem unconcerned and overly optimistic, why should the parents be conscientious about giving the medicine and getting the eye exams? Furthermore, we need to be conscientious about making sure you get the disease under control. Controlling the information will control the bony overgrowth and minimize the pain, swelling, limitation of motion, and discomfort. All of these issues have a major and long lasting impact on the family and child. There is also plenty of experience to suggest that vigorous NSAID therapy may help to control uveitis. It may not be enough, but it is incorrect to assume it has no role.

I favor an aggressive approach to treatment of these children. For example, children whose oligoarticular disease does not promptly come under control need to be treated aggressively to bring the disease under control. In my opinion, if you've been treating the child for three months and tried more than one NSAID without an adequate response, this is not a typical oligoarticular response, no matter what joint is involved. (Note: We are also only too aware that NSAIDs in the 2005 are not necessarily benign drugs.) It may be time to get more aggressive with a remissive drug and not allow the inflammation to continue. I do occasionally hear rheumatologists say things like, "I would never use methotrexate in an oligoarticular child." I agree that it is unusual for us to have to use methotrexate or biologics in a "true oligo", e.g., with one to three joints involved, but it may be crucial to that child's well-being. Joint injections with corticosteroids may be helpful, but seldom stop aggressive disease like a remissive drug. An occasional child may have only one or two knees involved, but still develop a destructive arthritis in a knee that requires a remissive drug. Some

oligoarticular children may have major functional problems that interfere with their normal lives and may need methotrexate or etanercept. This is a simple message: Don't rule out aggressive treatment.

I would also caution against defining oligoarticular JIA by counting joints. It may fit the original JRA criteria, but it suits the 1970's and 1980's better than 2005. Long ago I showed my mentor Dr. Virgil Hanson a child with a diffusely swollen ring finger – three joints involved total. When he called it “pauciarticular JRA” I commented that it didn't look like any “pauci” I'd ever seen. “Well, it probably isn't the same thing,” he promptly agreed. “But it fits the criteria and we don't know what else to call it.” Well that was almost thirty years ago. Now we do know a bit better what to call it (psoriatic, extended oligo) and we know it behaves very differently and needs to be treated very differently. If the child's disease in front of you is not behaving like typical oligoarticular JIA, I would suggest that you don't just count joints and think that the resultant number limits your therapeutic options. You are responsible for getting the best result for the child in front of you and kids always don't read the book or follow perfectly criteria written down by a committee. The rheumatologists on those committees (including the ones that established the JRA and the JIA criteria) knew that there were exceptions and expected rheumatologists to allow for exceptions, too. Some of these children will develop extended oligoarticular disease, develop complications, or never had oligo disease in the first place.

Let me suggest some guidelines for predicting that a child may turn out to be an “exception”:

- 1) ESR great than 40 mm/hr or high platelet count (e.g., $>600,000/\text{mm}^3$)
- 2) Hemoglobin less than 10.5
- 3) Involvement of any joint other than the knee raises the possibility of an exception:
 - a) ankles—could be an oligoarticular but could be polyarticular, systemic, or psoriatic.
 - b) toes—very unusual to be an oligo—more likely psoriatic or poly JIA.
 - c) wrists—unusual—think poly, systemic, or psoriatic.
 - d) finger PIP's or MCP's—rare in oligos—think polyarticular, systemic, or psoriatic disease also.
 - e) elbows—unusual—think psoriatic, systemic, or polyarticular disease.
 - f) Hips or lumbar spine—never in oligo's—suggests spondyloarthropathy or psoriasis early in disease course; for the hips alone, if hip problems occur late, it may suggest systemic or polyarticular disease.
 - g) cervical spine—uncommon—think extended oligoarticular disease or polyarticular or systemic.

In conclusion, I would suggest that we all maintain a health respect for oligoarticular JIA. True, it does not have the risk of polyarticular or systemic JIA or most spondyloarthropathies, but it is not a benign condition in some children. Be cautious in your predictions of early remission and an excellent prognosis. Be aware of the complications of uveitis, unusual involvements like TMJ disease or cervical spine disease, or destructive arthritis. Most important, be ready to treat aggressively if the child's arthritis starts to deviate from the typical oligoarticular course.