Editorial

Pediatric Rheumatology: Old Diseases, But A New Division!

Pediatric rheumatology is considered to be the newest subspecialty developed in pediatrics. Indeed, the American Board of Pediatrics and the American Board of Medical Specialists recently secured the board status of pediatric rheumatology in 1989, and its first board examination took place in 1992. To date, there are probably less than 200 board certified pediatric rheumatologists distribute through out the world.[1]

Pediatric rheumatology is the clinical discipline dedicated to the study and care of children with both inflammatory and non-inflammatory connective tissue disorders. Most of the diseases in this category have multisystemic involvement, thus the term "autoimmune diseases" is applied by some authorities. Since there is no clear understanding of the extent, etiology and pathogenesis of childhood rheumatic diseases, it is hard to classify these diseases into groups like other disorders, for instance, infectious diseases. The diagnosis is based largely on carefully developed diagnostic criteria so that comparisons in different aspects can be made between institutions. Since this subspecialty was established recently, some classifications may be changed as we learn more about the disorders; for example, the classification of juvenile arthritis was recently modified in 1997 as our understanding of the characteristics, natural history and more importantly, the response to therapy has become better.

It has been widely accepted that children are not small versions of adults. Rheumatic disorders that develop during the childhood period are clearly different from those of adults although they do bear the same names. As experience in taking care of this group of patients accumulates, differences in the clinical presentation, natural history, and response compared with the same diseases in adults have been noticed. Together with the nature of pediatric patients in term of their physical and mental immaturity, specially trained pediatricians and a unique approach are required to deal with children with these complicated diseases. Factors that modify rheumatic diseases of childhood are :[2]

1. Growth and development potential.
2. Immaturity of the gonad.
3. Immune system immaturity.
4. Less complex antigenic explosure.
5. Early expression of immunogenetic predisposition.

Childhood is not a constant period of life. Several continuous changes are taking place in both anatomical and physiological aspects. The influence of growth and development may in part effect the pathogenesis and manifestation of disease. The degree of skeletal maturity influences the expression of rheumatic disease is poorly understood. It seems probable that physical and biochemical differences between young cartilage and bone and older cartilage and bone profoundly influence the effect of an inflammatory process involving those structures. The physes are not fused in children, local growth disturbance may occur in untreated arthritis in children, and generalized growth failure or short stature may result in children with widespread, uncontrolled arthritis. This certainly does not happen in an adult with arthritis. Age-specific diseases have also been identified only in children, for example, Kawasaki disease, and several types of juvenile idiopathic arthritides. Most of the time, the manifestations of the same adult rheumatic diseases early in life are not typical to those of adults and at times, totally different, as in the seronegative spondyloarthropathy group, as this group is classified into "enthesitis-related juvenile idiopathic arthritis, JIA" according to their own characteristics.

The immaturity of gonadal development may affect the manifestation of disease in children. The influence of sex hormone in the expression of some of the childhood rheumatic disorders is less apparent. For example, SLE presenting in early childhood or prepuberty has a lower female:male ratio than that seen in adults.

Genetically determined diseases are expressed early in life as in many skeletal dysplasia and biochemical disorders. The immature or developing immune system may also play a part in

the pathogenesis and expression of the disease. Limited exposure to environmental antigens may explain the less frequent association of some genetic predisposition markers with childhood rheumatic diseases, such as only 10% HLA-B27 positivity in juvenile ankylosing spondylitis patients. Also in some diseases such as persistent oligoarticular JIA and Kawasaki disease, occur only in early childhood.

The response to treatment and selecting appropriate agents in the management of childhood autoimmune disorders are also different from adult rheumatic disease. Anecdotally, it seems that the pediatric age group may be more sentitive to some of the immunosuppressive drugs. Growth and development, risk of malignancy and organ complications secondary to treatment are among the major concerns in choosing appropriate agents as children still have many years ahead of them.

As mentioned earlier, most of the rheumatic diseases of childhood are multisystemic diseases. Pediatric rheumatologist will need to work and coordinate with other pediatric subspecialists and a wide variety of medical personals as followed:

1. Pediatric PM & R
2. Child psychiatrist and psychologist
3. Pediatric ophthalmologist
4. Nutritionist
5. Pediatric orthopaedist
6. Pediatric dentist
7. Social worker

It has been well demonstrated where inflammation takes place, if left untreated or uncontrolled, fibrosis is the ultimate result and at this point, recovery is difficult and usually irreversible! It is crucial to identify this group of patients, diagnose and more importantly introduce specific treatment in order to prevent severe complications and disabilities. As there is only a window in time when active inflammation takes place. If we miss this period, then the fibrosis will start and then it may be too late to reverse it. Children with inflammatory diseases are all in pain and have limited locomotion as part of their inflammatory signs + symptoms (pain, swelling, erythema, warmth and loss of function.), especially a child with arthritis. Children need to mobilize toward the appropriate stimulus in order to gain or develop optimal or age-appropriate physical skills and mental development. Once this ability is taken away by their disability, their development will slow or stop and their quality of life will eventually be limited. It is our job as pediatricians to prevent disability from happening to the next generation so they can: live as normally as possible, develop physically and mentally to their maximum potential and remain healthy so they can eventually become productive adults in the future.

There are some circumstances in which a pediatric rheumatologist may be able to offer help in the evaluation of patients who present with:

1. Fever of unknown origin. Apart from infectious causes (especially in our country), malignancies and autoimmune/ connective tissue disease are always included in the differential diagnosis.
2. Musculoskeletal complaints.
3. Rashes or nodules of unknown etiology, especially painful ones.
4. Diseases with evidence of reticuloendothelial system activation i.e. generalized lymphadenopathy, hepatosplenomegaly.
5. Other organ-specific diseases that required potent immunosupressive agents other than corticosteroids, i.e. uveitis.
6. Any disorders in which the diagnosis remains unclear or unknown.

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References

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