

Pediatric rheumatology: A special issue from the European Journal of Rheumatology

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Childhood-onset rheumatic diseases are generally chronic and are associated with significant morbidity and sometimes mortality in affected children, adolescents, and young adults. The appreciation of rheumatic diseases during childhood, aside from rheumatic fever, dates back to the late 19th century (1). It was not until the mid-1900s that centers dedicated to the care of children with chronic arthritis were established (2). In 1976, the American Rheumatism Association (predecessor of the American College of Rheumatology) organized a meeting on rheumatic diseases of childhood in Park City, Utah (3). This meeting marked the first step towards developing pediatric rheumatology as a specialty, which was formally recognized by the American Board of Pediatrics in the early 1990s. It has since been increasingly appreciated that while basic disease mechanisms might be similar between children and adults for some autoimmune diseases, implications, complications, and management of these chronic disease conditions are often different in the young. In lupus for example, disease severity and the extent of major organ involvement are more pronounced in childhood-onset compared to adult-onset disease (4). Some of these differences in disease manifestations have been attributed to higher genetic risk when complex polygenic autoimmune diseases start during childhood (4). Monogenic forms of these conditions are usually characterized by an early disease onset (5). Therefore, it is likely that differences in basic pathogenic mechanisms and in interactions between genetic factors and environmental triggers could explain, at least in part, clinical variability between pediatric and adult-onset rheumatic diseases.

We are pleased to put together this special issue on *Pediatric Rheumatology* for the first time for readers of the *European Journal of Rheumatology*. This issue includes a number of invited review articles by leaders in the field, original research articles, and case discussions. Invited reviews were carefully solicited to be authoritative and to focus on topics less addressed in the current literature.

Glaser et al. (6) performed a systematic review on linear scleroderma of the head, commonly referred to as En coup de sabre, which is French for “by strike of a sword”, due to resemblance of lesions to the scar of a sword wound. The condition is commonly associated with Parry Romberg Syndrome, which is characterized by progressive unilateral atrophy of the face, involving the skin and underlying soft tissues, muscles, and osteo-cartilagenous structures. They reviewed 215 manuscripts including 1430 patients, and provided a flow chart summarizing recommendations for comprehensive evaluation, monitoring, and treatment for patients with En coup de sabre/Parry Romberg Syndrome.

Yıldız et al. (7) reviewed Behçet’s disease in childhood, and highlighted similarities and differences in classification criteria, clinical presentation, and manifestations between children and adults with Behçet’s disease. Management options in Behçet’s disease were also discussed. In yet another type of vasculitis, Aeschlimann et al. (8) provided a comprehensive review on Takayasu arteritis during childhood, a rare large vessel vasculitis. They discussed recent literature summarizing what is currently known about the pathogenesis of Takayasu arteritis, epidemiology, clinical presentation, angiographic patterns, differential diagnosis, classification criteria, and current treatment options, with a focus on recent observations in pediatric patients. Importantly, they summarized detailed clinical and laboratory characteristics of 599 pediatric Takayasu arteritis patients. In a similarly rare inflammatory vascular disease, Madison et al. (9) comprehensively discussed antiphospholipid syndrome in childhood, and provided guidelines for approach and therapy in the pediatric patient population based on current up to date literature. They also discussed pathogenic mechanisms and important considerations for neonates with the disease, which reflect unique characteristics of the coagulation system in this age group.

Tille et al. (10) described a case of pediatric IgG4-related disease with ophthalmic involvement and colitis. They performed a detailed review of current literature pertinent to disease epidemiology, pathophysiology,

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diagnosis, and management. A number of flow charts and tables summarizing current recommendations for the diagnosis and treatment of IgG4-related disease are provided. An update and a comprehensive review of pediatric macrophage activation syndrome is presented by Crayne et al. (11) The authors compare the various diagnostic criteria for macrophage activation syndrome, discuss the genetic basis and pathophysiology, and provide an update on treatment options, including the role of cytokine-specific therapies in this life-threatening condition.

The utilization of musculoskeletal ultrasound is expanding in rheumatology. Brunner et al. (12) discussed the role and applications of musculoskeletal ultrasound in pediatric rheumatology. They provided a comparison between the utility of musculoskeletal ultrasound versus MRI, and highlighted the potential utility of musculoskeletal ultrasound in predicting disease flares and assessing damage in rheumatic diseases during childhood. Key sonographic features of joints in healthy children and in various pathologies were also discussed and demonstrated using example images.

This special issue also includes original research articles. Reiff et al. (13) performed a retrospective chart review and report that 8 out of 87 (9%) patients with pediatric localized scleroderma had co-existing inflammatory arthritis. Their data suggest that inflammatory arthritis in this group of patients was less likely to respond to therapy with methotrexate and other disease modifying anti-rheumatic agents than the co-existing skin involvement. Another original research article by Rai et al. (14) provided a perspective for clinical, microbiological, and imaging features of neonatal septic arthritis in India. The series included a cohort of 43 patients, a relatively large number for reports of

this condition, most were followed for at least 12 months.

Several case reports and discussions were also included in this issue. Wiener et al. (15) shared their experience with pediatric Tolosa-Hunt syndrome, a rare granulomatous disease of the cavernous sinus that presents with headache and unilateral painful ophthalmoplegia. They reported the successful treatment of this condition with adalimumab in one patient. Krutzke et al. (16) described a patient with COPA syndrome, a relatively newly recognized interferonopathy characterized by autoantibody production, arthritis, interstitial lung disease, and kidney involvement. COPA syndrome is believed to be caused by autosomal dominant mutations in the gene encoding coatmer complex I, subunit alpha (*COPA*). The authors reported successful treatment of a 15-yr old girl with COPA syndrome using the JAK1/2 inhibitor baricitinib. They also reviewed the literature on 31 additional COPA syndrome patients published to date.

We hope you enjoy reading this special issue.

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