



EDITORIAL

## Paediatric Rheumatology: Where we are coming from and where we are going<sup>☆</sup>



### Reumatología Pediátrica, de dónde venimos y a dónde vamos

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In 1483, Sandro Botticelli depicted what may be one of the earliest representations of juvenile idiopathic arthritis (JIA) in ‘Portrait of a Youth’, in which he showed the swelling of the wrist, metacarpophalangeal and proximal interphalangeal joints of one hand. But it was in 1897 that George Frederic Still, a paediatrician and pathologist at the Hospital for Sick Children at Great Ormond Street, London, published an article describing 22 children with chronic arthritis and highlighting that rheumatic manifestations in children differed from those in adults. Some of these patients had what years later came to be known as Still’s disease and is currently known as systemic-onset JIA. It was not until 1940 that the idea emerged again that children could have rheumatic diseases other than rheumatic fever.

Paediatric Rheumatology is a subspecialty of paediatrics focused on the diagnosis and management of a broad range of diseases: musculoskeletal disorders such as JIA, systemic autoimmune diseases such as systemic lupus erythematosus SLE, juvenile dermatomyositis or juvenile scleroderma, autoinflammatory diseases such as periodic fever, apht-

ous stomatitis, pharyngitis and cervical adenitis PFAPA syndrome or hereditary periodic fever syndromes, disorders manifesting with increased pain such as fibromyalgia or chronic fatigue syndrome, or secondary osteoporosis, to name a few. To further knowledge in this field, in 1992 a group of paediatricians and rheumatologists in Spain created the Grupo Español de Reumatología Pediátrica (Spanish Group of Paediatric Rheumatology, GERP), which in 1995 became the Section of Paediatric Rheumatology of the Asociación Española de Pediatría (Spanish Association of Pediatrics, AEP) and in 1998 gave rise to the Sociedad Española de Reumatología Pediátrica (Spanish Society of Paediatric Rheumatology, SERPE).

The most frequent chronic rheumatic disease in the paediatric population is JIA, formerly known as juvenile rheumatoid arthritis or juvenile chronic arthritis; JIA ultimately is a consensus-based term that encompasses several subtypes of disease. Few studies have been conducted in Spain to analyse its epidemiology, and a study conducted in Catalonia reported an incidence of 6.9 per 10 000 children aged less than 16 years and a prevalence of 39.7 per 10 000 children aged less than 16 years.<sup>1</sup> On the other hand, it is estimated that 20% of cases of SLE have onset in childhood or adolescence, and most autoinflammatory disorders have onset in childhood. As physicians, we are all familiar with the saying that ‘one only diagnoses what one knows’, something that is particularly true in the field of paediatric rheumatology. The study published by Udaondo Gascón et al.

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in the current issue of *Anales de Pediatría* corroborates the perception that training in paediatric rheumatology must be improved at every level (undergraduate and postgraduate).<sup>2</sup> It is essential that training programmes for medical students, residents in paediatrics and continuous education include not only information on this group of diseases but also on the evaluation of the musculoskeletal system. Nobody would think that it is acceptable for paediatricians not to know how to examine an ear or perform auscultation of the chest, but even the most recent evidence today shows that the physical examination often does not include a musculoskeletal evaluation and that many medical providers lack confidence in assessing joints.<sup>2,3</sup>

Advances in imaging techniques along with the increased availability of ultrasound technicians in many clinics and in genetic diagnosis, which guides the management of newly recognised autoinflammatory diseases, combined with the development of new treatments, are some of the factors that have contributed to improvements in the management of these diseases that we would not even have dreamt of until a few years ago. Despite advances in treatment, methotrexate, which has been in use for more than 30 years, continues to be the main systemic drug used for the management of diseases like JIA. For this reason, we consider that the work of Barral Mena et al. analysing the factors associated with adverse reactions to methotrexate, published in this issue of *Anales de Pediatría*, is certainly relevant. It shows that despite its efficacy, up to one third of patients need to discontinue methotrexate due to adverse effects.<sup>4</sup> Fortunately, alternative options are available today, such as selective immunosuppressive agents (biologic drugs) or Janus kinase (JAK) inhibitors. The days when children frequently ended up with significant limitations or physical sequelae, or even dependent on a wheelchair, are behind us. Today, the vast majority of patients lead normal lives and participate in sports activities as do children of the same age.

On the other hand, the follow-up of patients in specialty clinics has allowed participation in collaborative networks, registers, studies and clinical trials at the national and international levels. Institutions such as the Pediatric Rheumatology Trials Organization (PRINTO), the Pediatric Rheumatology Society (PReS) or SERPE have invested on promoting research and the establishment of working groups to join efforts and progress in the field. In 2013, the Single Hub and Access point for paediatric Rheumatology in Europe (SHARE) initiative was launched with the participation of European paediatric rheumatology networks with the aim of describing current needs for the optimal management of children and youth with rheumatic disease.<sup>6</sup> This initiative has given rise to numerous publications in recent years with recommendations for the management of the most common paediatric rheumatic diseases, the performance of collaborative studies or the establishment of biobanks. Particular emphasis has been placed on the needs of adolescents with chronic disease and how to facilitate transitioning to adult care, which often involves a switch in the provider in charge of the patient.<sup>7</sup>

We ought to highlight the important and novel role taken in recent years by patient organizations. For instance, since 2015 and with the support of the SERPE, a yearly day-long "symposium for the families of children

and adolescents with rheumatic diseases" is held simultaneously in several cities throughout Spain in collaboration with local patient organizations and the specialists that manage these patients. In these symposiums, health professionals, parents and children share experiences, questions and hopes. Starting this year, this symposium will be held on March to join the celebration in multiple countries of the World Young Rheumatic Diseases day (WORD). Furthermore, some of these organizations make altruistic contributions to research in paediatric rheumatology, offering research grants to further knowledge on these diseases.

However, many challenges remain, for instance at the educational level, since, as is the case in other paediatric subspecialties, adequate training in the field needs to be guaranteed. In 1992, the American Board of Pediatrics held the first paediatric rheumatology subspecialty exam, and in 1994, specialist training was approved in the United Kingdom. Since then, numerous countries have recognised it as a specialty: Germany, the Netherlands, Italy, France, Austria, Sweden and Finland, among others. Furthermore, the European Union of Medical Specialists (UEMS) and its paediatric section, the European Academy of Paediatrics (EAP), have acknowledged this specialty and the specialty training curriculum at the European level. Spain should promptly recognise paediatric subspecialties officially, including paediatric rheumatology.

At the level of care delivery, new communication technologies must be considered and applied to facilitate access of patients and their families to the providers involved in their care. In this regard, specialty nurses and case managers can play an essential role in contributing to the education, self-care and follow-up of these patients.

When it comes to research objectives, it is important to continue to develop new drugs and to conduct trials with those that already exist, as well as to perform studies with the aim of identifying biomarkers (genetic, molecular, indicative of disease recurrence or treatment response) to aid prognosis and thus guide the selection of appropriate, individualised and targeted treatment, which not only would increase its effectiveness and decrease the probability of recurrence, but also improve the adverse reaction profile.<sup>5,8</sup>

As health care providers, we must strive to ensure access to all patients to the best possible care, to specialised care and to appropriate treatment for their disease. As paediatricians, we should be able to demand from our managers, coordinators, administrators and politicians that children with suspected non-infectious arthritis or autoimmune disease be assessed by a paediatric rheumatologist. There is still a long way to go, but the subspecialty of Paediatric Rheumatology is now a reality established on solid foundations.

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