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Perspective

Paediatric rheumatology: What has changed in last 10 years?

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The last decade has witnessed rapid growth in awareness of the rheumatic diseases of childhood, an enhanced understanding of many of the unique aspects of their presentation, diagnosis, treatment, and outcome, the application of dramatically effective therapies, and the emergence of increasingly widespread clinical expertise in patient care. Importantly, the discipline, initially centred in Europe and North America, is now emerging in parts of the world, such as the Indian subcontinent, where most of the world's children live.

With paediatric rheumatology now established on every continent, the pace of national and international collaboration is accelerating. This welcome development facilitates sophisticated clinical and basic research on much larger populations which have heretofore been inadequately studied, and the application of the technical expertise available in a limited number of laboratories to the investigation of a much broader patient community. In addition to promoting communication among the widely dispersed members of our discipline, such collaboration produces information of much greater significance.

Many milestones could be cited. The publication of the International League of Associations for Rheumatology (ILAR) criteria for classification of juvenile idiopathic arthritis (JIA)[1] has facilitated communication and systematisation of the diagnosis of this complex group of diseases. An increasing number of clinical drug trials in many of the rheumatic diseases of childhood have contributed to better outcomes. Evaluation and comparison of results of clinical trials in JIA has been facilitated by the application of the "Core Set" criteria.[2] The fact that disease control can now be achieved in JIA has required the adoption of criteria for remission.[3] Increasing recognition of the genetic basis of the autoinflammatory diseases has added a new dimension to the discipline and will illuminate the pathogenesis of many inflammatory diseases.[4] Most of these disorders are responsive to anti-IL1R therapy.[5] The similarity of many of the autoinflammatory diseases to systemic JIA suggests a reconsideration of the place of that disease within the JIA umbrella.[6] The macrophage activation syndrome (MAS), a potentially fatal complication of several rheumatic diseases, especially systemic JIA, can now be recognised in its early stages and effectively treated.[7] Increasing recognition of the central nervous system vasculitis has broadened the spectrum of vasculitides with which the paediatric rheumatologist is confronted.[8]

Advances in drug therapy have radically changed the management and outcome of children with a range of rheumatic diseases. The newest additions to the therapeutic armamentarium, the biologic response modifiers, may mark the most important therapeutic advance since the introduction of corticosteroids. Inhibition of the interaction of tumour necrosis factor (TNF)-α with its receptor has been demonstrated to dramatically improve the outcome of children with polyarthritis.[9] Systemic JIA, a disease that has often been very difficult to control, usually responds very well to anti-IL1R or IL6R biologics.[10,11] Resistant uveitis often responds to the anti-TNF-α biologic infliximab,[12] adalimumab,[13] or the immunomodulatory drug mycophenolate mofetil (MMF).[14] The MMF has also found a significant role in the treatment of systemic lupus erythematosus (SLE), reducing the requirement for more toxic drugs such as cyclophosphamide.[15] Treatment of some of the vasculitides such as granulomatous polyangiitis with biologics that act on CD20 receptor on pre-B and mature B lymphocytes results in a favourable response and diminished corticosteroid requirement.[16] These agents are also effective in the management of patients with resistant dermatomyositis[17] and some aspects of SLE.[18] The biologics have changed the practice of paediatric rheumatology in the last decade, and give the promise of much improved outcome for our patients, although the long-term risks of infection, autoimmune and malignant disease (not to mention their high cost) necessitate caution in their use.

Awareness of the importance of outcome measures such as function and quality of life has increased considerably in the last decade, and such measures are now incorporated into many clinical studies.

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Children with rheumatic diseases throughout the world can look forward to vastly improved outcomes as a result of extensive basic research, translated to the bedside by well-trained and highly motivated clinicians. We are witnessing an era of great excitement and optimism in this discipline. If the last decade is any indicator, progress in the next decade will astonish us!

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