



## Editorial

# Juvenile localized scleroderma: Is it a benign disease? ☆



## Esclerodermia localizada juvenil: ¿es una enfermedad benigna?

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Scleroderma is an autoimmune polymorphic disease characterized by the presence of cutaneous sclerosis secondary to excessive accumulation of collagen. It can be classified into systemic, localized, and scleroderma-like syndromes.<sup>1,2</sup>

It is called localized scleroderma when the skin commitment is not accompanied by involvement of internal organs. According to the classification criteria published by the European Society of Pediatric Rheumatology, it includes 5 subtypes: circumscribed morphea, linear scleroderma, generalized morphea, pansclerotic morphea and mixed scleroderma (when there is a combination of 2 or more of the above subtypes).<sup>2,3</sup>

In this issue of the Journal, Arango et al.,<sup>4</sup> through a descriptive cross-sectional study, describe 88 children from 10 centers specialized in Pediatric Rheumatology of Colombia. The study included patients with a diagnosis of juvenile localized scleroderma with a minimum of one year of evolution and 6 months of follow-up, with a higher prevalence in women, an average age at the onset of the disease of 7.1 years (0–14), and a mean duration of the disease at diagnosis of 16.5 months (1–96). The distribution by subtypes was circumscribed morphea (32.9%), mixed (31.8%), linear (21.5%, it rises to 55% when mixed forms with linear lesions are included) generalized (11.4%) and pansclerotic (2.3%). Esthetic alterations were detected in 91%, growth alterations in 41%, and functional commitment of neighboring joints in 32%. Extradermal commitment was present in 22.7% and polyautoimmunity in 12.5%. The authors conclude that juvenile localized

scleroderma is a polymorphic and unpredictable disease, with late diagnosis, in which the rate of extradermal commitment suggests that it is not a disease limited to the skin.

In a study conducted in Mexico, we found 44 cases of juvenile localized scleroderma, with an average time of evolution of the pathology of months (range between 7 months and 10 years).<sup>5</sup> Zulian et al.<sup>6</sup> described 750 children from 70 centers coming from Germany, Argentina, Brazil, Canada, USA, Spain, England, Israel, Italy and the Netherlands, with insidious evolution of the disease, delay in diagnosis, and recognition of the mixed subtype in its presentation. Wu et al.<sup>7</sup> reported delays in the referral to the pediatric rheumatologist of up to almost 2 years and a limitation in functional ability of 30% in the record of the Childhood Arthritis and Rheumatology Research Alliance of USA. Mertens et al.<sup>8</sup> warn of the recurrence of the morphea lesions in up to 27% of pediatric cases.

The results of the work of Arango et al.<sup>4</sup> coincide with others from other centers around the world, with different populations, and raising the same problems, which suggests that both in Colombia and throughout Latin America, doctors and healthcare authorities must join our efforts to achieve early diagnosis, the understanding of the disease by the patients and caregivers, the importance of adherence to treatment, the guaranteed access of the patients to specialized consultation and medications, and a close follow-up that allows early prevention and detection of complications derived from the disease.

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