Arch Argent Pediatr. 2024;122(6):e202410317

Childhood leprosy in Buenos Aires: four cases report

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ABSTRACT

Childhood leprosy, which affects children up to 14 years old, is characterized by a delay in diagnosis since it is usually confused with other dermatoses. Its presence in a child is a relevant epidemiological indicator since it signals active disease transmission.

We present 4 patients between 5 and 14 years old who attended a public hospital in Buenos Aires —two patients with borderline tuberculoid leprosy, one with lepromatous leprosy, and one with indeterminate leprosy.The World Health Organization provides therapy for people between 10 and 14 but does not consider children under 10. This difficulty implies adapting the dosage and pharmaceutical form to each patient under this age.

Finally, it should be noted that the diagnosis of the patients led to the diagnosis and treatment of the disease in adult cohabitants.

Keywords: *leprosy; child;* Mycobacterium leprae*; reveral reaction; hypoesthesia.*

doi: http://dx.doi.org/10.5546/aap.2024-10317.eng

To cite: Merenzon S, Costa AL, Frare C, Tiscornia JE. Childhood leprosy in Buenos Aires: four cases report. *Arch Argent Pediatr.* 2024;122(6):e202410317.

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Funding*: None.*

Conflict of interest*: None.*

Received*: 1-9-2024*
Accepted*: 5-17-2024*

INTRODUCTION

Leprosy is one of the typical neglected tropical diseases in developing countries.[1] It is a chronic infectious-contagious disease caused by the bacterium *Mycobacterium leprae* that can leave disabling sequelae if not treated promptly. Visible deformities are a significant cause of stigma affecting the social inclusion of those with the disease. In 2021, globally, there were 8490 new cases with grade 2 disability (deformity or visible damage to eyes, hands, or feet), of which 4% were children.[2] Within Latin America, Brazil is the country with the highest incidence of childhood leprosy. Children are more susceptible than adults, due to their developing immunity and possible intrafamilial contact.[3,4] During 2022, 343 people in treatment were registered in Argentina, of whom 1.41% were under 14.[5] Currently, Argentina has reached the leprosy elimination target, with a prevalence since 1996 below 1 per 10,000 inhabitants.[6] The endemic regions are the Autonomous City of Buenos Aires (CABA, by its acronym in Spanish), Greater Buenos Aires, Chaco, Formosa, Mesopotamia, Santa Fe, Córdoba, Santiago del Estero, Tucumán, Salta and Jujuy.[7] Leprosy mainly affects the skin and peripheral nerves but may also involve internal organs. It has low pathogenicity and high infectivity. It has an incubation period of between 2 and 7 years, and its main route of infection is through nasal secretions.[1] It is estimated that between 3% and 5% of exposed persons develop different forms of clinical presentation depending on their immunological status.[6] In childhood, it manifests differently than in adults: it usually presents lesions in exposed areas of the skin, and sensory nerve alterations are difficult to evaluate.[6] In early and middle childhood, paucibacillary forms (PB) predominate (tuberculoid leprosy [TL], including infantile nodular and indeterminate [IL] [*Figure 1*]) and in adolescence, multibacillary forms (MB) (borderline leprosy, tuberculoid borderline [TB] [*Figure 2*], lepromatous borderline [LB] and lepromatous [LL]).[7] This is probably because the more contagious MB forms require a longer incubation period to manifest.[5] Diagnosis is often delayed because it shares clinical features with other dermatoses.[7] When it presents as hypoesthetic hypopigmented macules in the case of IL (a common childhood-onset form that can later change to other forms within the spectrum), a differential diagnosis should be made with acromion eczema, post-inflammatory hypopigmentation, acromic nevus, vitiligo, pityriasis versicolor, hypopigmented mycosis fungoides. If it manifests as an anesthetic erythematous plaque (up to 5), as in TL, it should be differentiated from granuloma *annulare* or *tinea corporis*.[7]

CLINICAL CASE 1

A 5-year-old male patient from San Miguel, province of Buenos Aires, consulted for lesions on the face and trunk, which had been treated with antifungal agents without response. He presented hyperpigmented plaques and other erythematous plaques with clear limits, some with a lighter border, giving them an annular appearance. Complementary studies were performed, and TB was diagnosed. During the control of cohabitants, the father and uncle were diagnosed with LL, and the paternal grandfather with LB, all of them them were born in another province (Tucumán) and now live in the province of Buenos Aires. During the clinical evolution, he presented a type I reactional episode (RE) in the second month of treatment with cutaneous and neural involvement (left ulnar); he was treated with corticosteroids, and improvement was observed in the third month.

CLINICAL CASE 2

A 14-years-old male patient, from Paraguay, has lived in CABA since childhood. He consulted two hospitals in CABA due to thigh nodules and arthralgias, which were interpreted as lupus panniculitis, for which he received corticosteroids. He was referred to our department due to positive Ziehl-Neelsen staining of skin biopsy. On physical examination, he presented multiple erythematous-violaceous nodules with increased local temperature in the lower limbs, predominantly in the thighs and upper limbs, hypoesthesia in feet and hands (in a glove and stocking distribution), poor general condition, and fever. LL and type II RE were diagnosed. During the examination of cohabitants, the father was diagnosed with LL.

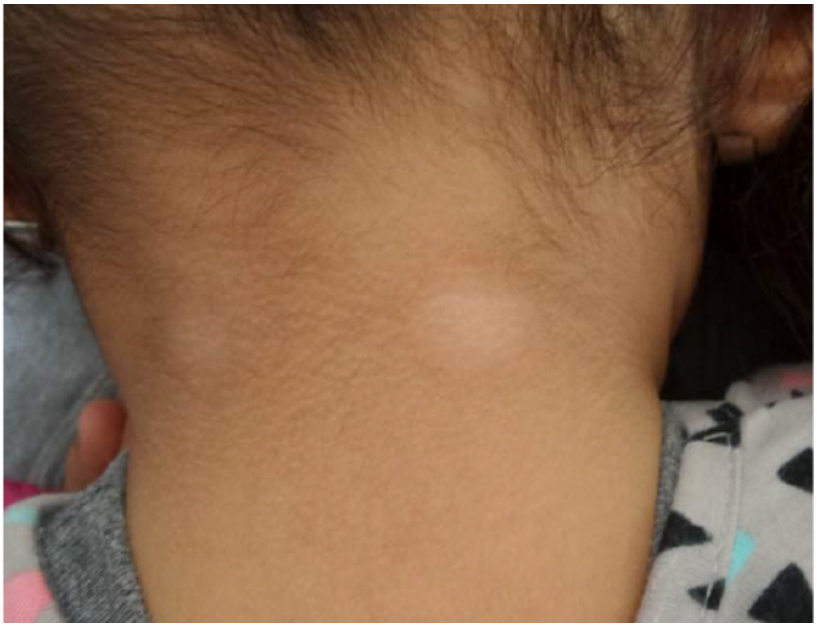
CLINICAL CASE 3

A 5-years-old female patient from CABA, whose mother had BL, presented with hypochromic macules in the occipital and abdominal, regions along with altered thermal sensitivity. The diagnosis was LI.

CLINICAL CASE 4

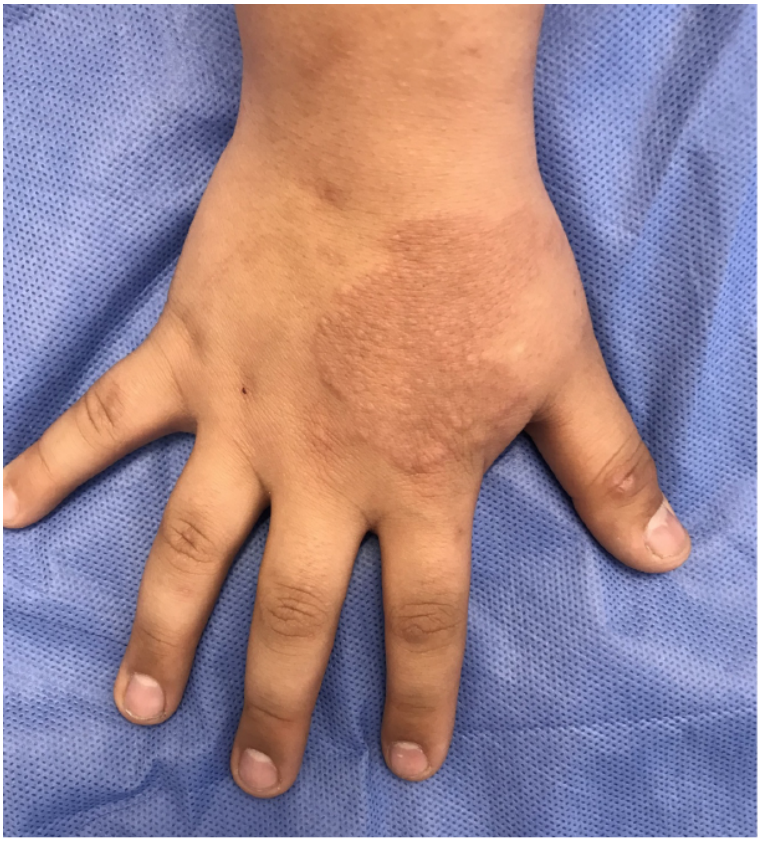
A 10-year-old male patient, whose father has LL, was from Lanús, province of Buenos Aires. He presented with an erythematous plaque with

Figure 1. Photograph of lesions of clinical case 3



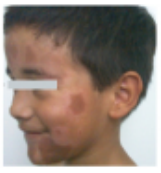
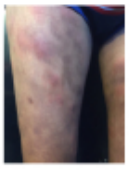
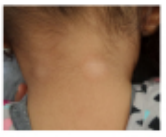
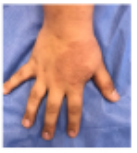
Annular hypopigmented macules in the occipital region with altered thermal sensitivity.

Figure 2. Photograph of the lesion of clinical case 4



Erythematous infiltrated plaque with polycyclic hypo-esthetic borders on the dorsum of the right hand.

Table 1. Clinical data of four patients with childhood leprosy

	Case 1	Case 2	Case 3	Case 4
Age/sex	5 yrs/M	14 yrs/M	5 yrs/F	10 yrs/M
Origin	Province of Buenos Aires	Paraguay	CABA	Province of Buenos Aires
Family history	No	No	Mother BL	Father LL
Time of evolution	1 year	1½ year	10 months	3 years
Clinical presentation	Hyperpigmented and other erythematous plaques with clear borders, some with lighter and annular borders on the face and trunk.	Multiple erythematous-violaceous nodules with increased local temperature on lower limbs, predominantly on thighs and upper limbs. Hypoesthesia in "boot and glove". Poor general condition. Arthalgias. Fever.	Hypochromic macules in occipital and abdominal region with alteration of thermal sensitivity.	Hypoesthetic erythematous plaque with polycyclic borders on the dorsum of the right hand. Hypoesthetic hyperpigmented macule on the left hand.
Clinical picture				
Biopsy	Macrophage infiltrates with vacuolated cytoplasm. Non-necrotizing granulomas with epitheloid cells and lymphocytes in the periphery. ZN: scarce AFB.	Compatible with LL and type II RE.	Lymphohistocytic inflammatory infiltrate and monocytes in papillary and periannexal follicular dermis. ZN: negative.	Compatible with granuloma *annulare*.
Bacilloscopy	Negative	BI 3+. MI is 80% solid and 20% fragmented. Globis regular to abundant.	Negative	Negative
Treatment	Pediatric WHO MB scheme. Prednisone 1 mg/kg/day in gradually descending doses.	WHO MB without dapsone and with minocycline. Thalidomid acenocoumarol for prothrombotic risk.	WHO scheme PB pediatric..	WHO pediatric PB PB scheme.
Complications	Reversal type I RE. Postinflammatory and clofazimine hyperpigmentation.	Type II RE WBC: 45,700 (82% N), PT: 67% (70-120), KPTT: 63" (24-37). Lupus anticoagulant positive. anti-B2 glycoprotein 1 IgM antibodies >150.	No	No
Epidemiologic screening	The father and uncle are with LL, and the grandfather is with BL.	Father with LL.	No new cases	No new cases
Follow-up and evolution	Discharge after one year of treatment and annual follow-up for 5 years with no recurrences.	Same as case 1	Discharge after 6 months of treatment and annual follow-up for 5 years without recurrences.	Same as case 1

AFB: acid-fast bacilli, BI: bacteriological index, BT: borderline tuberculoid leprosy, CABA: Autonomous City of Buenos Aires (by its acronym in Spanish), F: female, KPTT: kaolin-activated partial thromboplastin time, LB: lepromatous borderline leprosy, LI: leprosy indeterminate, LL: lepromatous leprosy, M: male, MB: multibacillary, MI: morphological index, N: neutrophils, PB: paucibacillary, PT: prothrombin time, RE: reactional episode, WBC: white blood cells, WHO: World Health Organization, ZN: Ziehl-Neelsen.

polycyclic borders on the *dorsum* of the right hand that had been evolving for three years, with a biopsy compatible with granuloma *annulare*. The lesion persisted, and a hyperpigmented macule was on the left wrist. Both lesions presented hypoesthesia. The diagnosis of TB was made. The detailed clinical cases are reported in *Table 1*. Monthly supervised intake was conducted at the service office, and to facilitate daily intake, the responsible adults were instructer (based on a color code) on which pill or syrup to administer to the children.

DISCUSSION

In the cases presented, there was a 3:1 male-to-female ratio, similar to that reported in the literature, although some publications report a ratio of 1:1.[8-12]. The most affected pediatric age range is between 10 and 15 years; six cases were identified in this series, with half were under 10 years old. Diagnosis requires at least one of the three cardinal signs: 1) definitive loss of sensation in a macule or hypopigmented or erythematous plaque; 2) thickened peripheral nerve with loss of sensation and muscle weakness; or 3) presence of acid-fast bacilli in a skin smear.[13] Sensory alteration is key to diagnosis, though unfortunately rarely perceived by the child. Screening is complex, and given the country's low incidence of this disease, is often overlooked.[7] Because infection in children reflects a recent infection, a high proportion of childhood leprosy cases is a sign of active transmission and an important epidemiological indicator.[4] REs are less frequent in children than in adults due to the predominant types of leprosy in each age group. Between 5 to 9 years old, the most common RE is type I mediated by the cellular immune response (seen in patients with BB, TB, and LB), presenting as new papules or plaques or enlargement of pre-existing ones, and potentially causing severe neuritis of peripheral nerves that can lead to deformities such as claw hand or foot drop, among others.[14] After the age of 10, type II reactions secondary to immune complexes and inflammatory cytokines (seen in patients with LL) such as erythema nodosum leprosum (ENL), may occur. Unlike erythema nodosum from other causes, which is limited to the anterior aspect of the legs, ENL nodules can appear on the trunk, face, and limbs, and are accompanied by poor general condition with altered erythrocyte sedimentation rate, leukocytosis, and visceral involvement,[7] iridocyclitides, as well as some cases of isolated pure neuritis.[15] ENL can sometimes be confused with classic erythema nodosum or lupus panniculitis.[7] RE in pediatric patients typically occurs in those older than 9 years, likely due to increased exposure time, making timely diagnosis crucial to prevent disabling sequelae.[15] Type I RE is treated with oral prednisone at 1 mg/kg/day with gradual tapering, while type II is managed with thalidomide at 100-200 mg/day.[14] Treatment typically includes rifampicin, dapsone, and clofazimine.[13] There is a critical need for pediatric syrup formulations, suitable for children under 10 years of age as current options are lacking. The treatment regimen is summarized in *Table 2*.

Chemoprophylaxis with single-dose rifampicin is recommended for contacts of patients over 2 years old. The treatment regimen is 2 years after ruling out of leprosy and tuberculosis. This work aims to raise awareness of leprosy among

Table 2. Treatment regimens recommended by the World Health Organization, 2018

Age group	Drug	Dosage and frequency	MB	PB
Children 10-14 years	Rifampicin + clofazimine + dapsone	450 mg once a month 150 mg once a month, 50 mg every other day 50 mg/day	12 months	6 months
Children under 10 years of age or under 40 kg weight	Rifampicin + clofamizine + dapsone	10 mg/kg once a month 100 mg once a month, 50 mg twice per week 2 mg/kg/day	12 months	6 months

MB: multibacillary, PB: paucibacillary.

pediatricians and integrate it into medical practice. Clinical suspicion should be raised for children presenting with hypo- or hyperpigmented macules and nodules with altered sensitivity; as these is the key diagnostic indicator. The dual epidemiological significance in pediatrics should be emphasized: it involves tracing contacts of diagnosed children to adults and vigilantly monitoring children living with adults affected by MB. Early diagnosis, prompt treatment, and active contact tracing are essential to further reduce incidence and prevent disability. ■

Acknowledgments

Margarita Larralde and Roberto Schroh (Hospital General de Agudos José María Ramos Mejía); Marianela Palmieri, Jesica Vivoda, Ana Bellelli, Daniel Navacchia and Susana Grees (Dermatology Unit, Hospital de Niños Pedro Elizalde), City of Buenos Aires, Argentina.

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