

# Infantile acropustulosis

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## ABSTRACT

Infantile acropustulosis (IA) is a benign cutaneous disease that affects infants in their first years of life and is characterized by recurrent crops of very pruriginous vesicles, papules and pustules with a mainly acral distribution. It is self-resolving. Its etiology is unknown but it has been associated to scabies. There are many different therapeutic options being topical steroids the most widely used. We performed a retrospective study of 22 patients diagnosed as IA at our institution during 12 years. We analyzed epidemiologic, clinical and therapeutic characteristics. 63.64% were female, symptoms started in a median of around 11 months old. Eight patients had been previously treated as scabies, with only three patients having a confirmed diagnosis. Treatment consisted of topical steroids plus antihistamines during each outbreak (Dermatol Argent 2010;16(4):268-271).

## Keywords:

*infantile acropustulosis, infantile pustulosis, sterile pustulosis.*

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## Introduction

Infantile acropustulosis (IA) described simultaneously by Kahn and Rywlin<sup>1</sup> and Jarratt and Ramsdell<sup>2</sup>, in 1979, is characterized by recurrent eruptions of erythematous papules that progress to vesicles and pustules of 1 to 4 mm, localized mainly on palms and soles, accompanied by intense Pruritus.<sup>3</sup> The dorsum aspect of hands and feet may also be affected; limbs, trunk, face and scalp are less frequently affected. Outbreaks occur every 2 to 3 weeks and each episode lasts 3 to 14 days. Episodes become less frequent and intervals between them become longer while the disease progresses. Typically begins between 2 and 24 months of life, however, cases in recently born infants and children up to 9 years old have been reported.<sup>4</sup>

Several authors have described that scabies frequently precedes IA and suggested a relationship between both diseases.<sup>5</sup>

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There are controversial opinions regarding treatment. Oral dapsone was considered to be the most effective agent, producing improvement in symptoms between 24 to 72 hours after administration; nowadays, due to its significant toxicity and the self-resolving nature of injuries, it is reserved for recalcitrant cases. Other studies support the use of topical glucocorticosteroids for their antiinflammatory activity.

The aim of our study is to determine the sex and age distribution of patients diagnosed with IA in our department and its relationship with scabies and its response to treatment administered.

## Materials and methods

We performed a retrospective, descriptive and transverse study over 22 patients with a clinical diagnosis of IA, who were treated in the ambulatory of the Pediatric Dermatology Area of Ramos Mejia Hospital, Buenos Aires, Argentina, from August 1997 to July 2009. The diagnosis was based on the presence of pruritic vesicles or vesiculopustules with acral location, combined with a history of recurrent similar episodes and lack of clinical and laboratory findings suggestive of other vesiculopustular infantile pathology. We performed a histopathological examinations of the lesions. Patients whose medical records had insufficient information were excluded.

We analyzed the epidemiologic characteristics of the disease, such as sex, age of onset of lesions and age at the time of dermatological examination on the affected patients, clinical characteristics, such as type and location of lesions, history of scabies, and type of undertaken treatment and the patient's response.

## Results

Of the 22 diagnosed patients, 14 were female (63.64%) and 8 male (36.36%). The onset age of symptoms ranged from 7 days to 36 months of age (with a mean of 11 months) and the initial clinical dermatological examination was carried out between 3 and 42 months of age (average 13 months) (**Table 1**). Pustules as well as vesiculopustules rashes were the most frequent clinical presentation (**Photo 1**). Pruritus was positive in 100% of patients. In 11 patients (50%) injuries committed both palms and soles and in 9 patients (41%) only soles were affected (**Photo 2**), the remaining 9% (2 patients) had in addition to palms and soles lesions (**Photo 3**), involvement of the scalp and upper trunk with a histological diagnosis of eosinophilic folliculitis (**Photo 4**).

Of all patients, 8 (40%) reported a previous diagnosis of scabies and had been treated with 5% permethrin cream (7 patients) or 6% sulfur petroleum jelly (1 patient). However, the diagnosis was confirmed only in 3 patients.

Biopsies and histopathological examination confirmed the clinical diagnosis.



**PHOTO 1.** Injuries affecting the internal side and sole of the foot.



**PHOTO 2.** Pustules on the lateral side of the foot.



**PHOTO 3.** Papules and pustules on palms.



**PHOTO 4.** Papules and pustules on the edge of the scalp.

On each outbreak, treatment consisted of medium and high power topical glucocorticoids (twice a day) in combination with oral antihistamines (carbinoxamine maleate 0.4 mg/kg) for 7 days in all classes. In 4 patients topical antibiotics were added to the basic treatment. There were no side effects to the medication.

## Comments

IA is a benign disorder with unknown exact incidence. The cases included in published papers worldwide are similar to that included in this study (22 patients over a period of 12 years). Some years ago, it was considered a male predominance, but nowadays, larger series of cases tend to show an equal distribution between both genders.<sup>6</sup> Our study showed a female predominance (63.64%) compared to males (36.36%). Early reports suggested a higher incidence in African Americans, however, the current communications describe IA in all races equally. Usually it begins between the first 2 to 24 months of life, although there have been cases reporting onsets by age of 9.<sup>4,5</sup> Spontaneous resolution is observed around the age of 3. In our study, the onset age ranged from 7 days to 36 months.

The etiology and pathogenesis of IA remains still unclear. In our series of cases, 8 patients had a previous diagnosis and specific treatment for scabies, but only in 3 patients the diagnosis was confirmed, therefore, it is possible that the other 5 patients had IA from the beginning. It is important to underline that the differentiation between these diseases is difficult. Possibly, some patients are overdiagnosed as scabies because not taking into account IA, since there is often great similarity in both clinical symptoms;

however, many authors suggest a relationship between these two diseases and concluded that possibly IA represents a cyclic hypersensitivity cutaneous reaction against antigens or antigenic components associated with a history of scabies infestation. On this basis, it is established the possibility of an idiopathic or *de novo* form and a form related to a previous infection by *S. escabiei*.<sup>7</sup>

The histology is typical. Intraepidermal vesicles filled with polymorphonuclear leukocytes located in upper epidermis and extending into the stratum corneum are observed, while in the papillary dermis there is a permeation of a perivascular lymphohistiocytic infiltrate with some neutrophils and eosinophils.<sup>8</sup> However, the diagnosis is mainly based on clinical manifestations, including the characteristic rash of papules, vesicles and/or pustules and their characteristic recurrence, as well as their location almost exclusively on palms and soles. In this paper, the results of histopathological studies coincided with the literature.

Some publications say that IA and eosinophilic pustular folliculitis of infants present clinical and histopathological characteristics that lead to the conclusion that they consist of different manifestations of the same entity, since the first sets mainly in areas lacking hair follicles, such as palms and soles, and the second, is seen mainly on the scalp and other hairy areas; the authors rescue that the difference would be caused, among other things, on the anatomical site where the biopsy is performed.<sup>9</sup> In our series of cases we found two patients with associated clinical and histological lesions compatible with both eosinophilic folliculitis and infantile acropustulosis.

The differential diagnosis should also include other vesicular and pustular pathologies of childhood such as neonatal toxic erythema, transient neonatal pustular melanosis, impetigo, candidiasis, herpes simplex infection, Langerhans cell histiocytosis, congenital syphilis, pustular psoriasis and eczema Dyshidrotic.<sup>10</sup>

Despite being a benign and self-limited pathology, each episode may be associated with irritability, sleep disturbances, excoriations and secondary infections, so it is essential to opt for an effective symptomatic treatment. Dapsone (4,4-diaminodifenilsulfona) has been considered in early publications by many authors, as the most effective treatment for severe IA at a dose of 2 mg/kg/day.<sup>11-13</sup> Apart from its antimicrobial action, dapsone has an anti-inflammatory action mechanism, as it is effective in skin diseases with abnormal accumulation of polymorphonuclear (PMN).<sup>14</sup> This drug is currently reserved only for serious cases due to its toxicity, cyclic evolution and self-resolving outbreaks characteristic of IA, and is used only in cases of severe and recalcitrant symptoms. Antihistamines are useful in the treatment for their sedative effects. It is also recommended the use of oral antibiotics and even oral glucocorticoids.<sup>2</sup>

Topical glucocorticoids of medium and high power are

**TABLE 1. Epidemiologic characteristics of patients with AI.**

<b>Sex</b>	Female	14
	Male	8
<b>Age of onset</b>	1 day to 11 months	15
	12 to 23 months	5
	24 to 35 months	1
	More than 36 months	1
<b>Age of consult</b>	1 day to 11 months	12
	12 to 23 months	8
	24 to 35 months	1
	More than 36 months	1
<b>History of scabies</b>	Yes	8
	no	14
<b>Associations</b>	Eosinophilic pustular folliculitis	2

very useful in the treatment of IA. Their antiinflammatory effects include inhibition of dermal edema, of capillary dilation and migration of inflammatory cells in the skin, with decreasing of vascular permeability and reduced migration of leukocytes through the vascular walls. Side effects such as skin atrophy and systemic absorption, would be diminished by reduced absorption in areas of thick skin (such as palms and soles), in addition, an intermittent use due to the ongoing outbreaks, helps to reduce their toxicity. The goal of treatment is, in all cases, improve symptoms and avoid complications such as secondary infections, therefore glucocorticoids of medium and high power associated with antihistamines were administered in all cases. Topical antibiotics were added to patients who presented secondary infection.

In conclusion, the IA is a self-limiting and benign disease, which should be considered in the differential diagnosis of rashes during childhood which present papules, vesicles and/or pustules as primary lesions; and it should be mainly differentiated from scabies, which is so frequent in our environment and with the greatest relationship that it has been

established, especially if the lesions are located only in acral regions. We also note the presentation of two patients sharing clinical lesions of IA and infantile eosinophilic pustular folliculitis.

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