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**Case report**

# Infantile perianal protrusion

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## Abstract

Infantile perianal protrusion is characterized by a skin fold located in the perianal area. It is a relatively recent reported condition and affects both infants and prepubertal children with a clear female predominance. Three types are recognized: constitutional/congenital, acquired, and associated with lichen sclerosus et atrophicus. We report eleven new cases, three of whom have the defect in locations that have been reported only once before. We would like to increase the awareness of this condition to avoid erroneous diagnostic and therapeutic procedures.

**Keywords: Infantile perianal protrusion, lichen sclerosus et atrophicus**

## Introduction

Infantile perianal protrusion (IPP) is a benign condition, clinically characterized by a solitary, pyramidal protrusion covered by red to rose-colored skin located in the perianal region [1]. These protrusions can be located in the midline, anterior and/or posterior to the anus [1].

IPP is often found in young girls and can be confused with evidence of sexual child abuse or genital warts [1]. There are three different types of IPPs: constitutional, which sometimes may be familial and / or congenital, acquired or functional, and a form associated with lichen sclerosus et atrophicus (LSA).

To date, 108 cases have been published and most of them have been female (98/99%) [2, 3]. Nevertheless, the incidence of this disorder might be underestimated [4].

## Case synopsis

A retrospective observational study was conducted at Ramos Mejía and Alemán Hospital, Buenos Aires, Argentina. The study included patients younger than 14 years who were diagnosed with IPP between March 1, 2005 and March 1, 2014. Eleven patients

between the ages of 0 to 7 years with IPP were included: 9 girls and 2 boys. Eight of our patients were breast-feeding, 2 of whom had the protrusion from birth, whereas the others began to notice it between 15 days old and 12 months of life (Table 1).

**Table 1.** Clinical features of patients with Infantile Perianal Protrusion

| Patient | Gender | Age of onset | IPP family history | Localization | Type of IPP |
|---|---|---|---|---|---|
| 1 | female | At birth | none | anterior and posterior midline | Constitutional |
| 2 | female | 11 months | none | midline posterior to the anus | Constitutional |
| 3 | male | 12 months | none | midline posterior to the anus | Constitutional |
| 4 | female | At birth | none | right perianal region | Constitutional |
| 5 | female | 15 days | Cousin with IPP | right perianal region | Functional (related to constipation) |
| 6 | female | 3 months | none | midline anterior to the anus | Functional (related to constipation) |
| 7 | female | 5 months | none | left perianal region | Constitutional |
| 8 | female | 7 years | none | midline anterior to the anus | Associated to LEA |
| 9 | female | 3 years | none | midline anterior to the anus | Constitutional |
| 10 | male | 12 months | none | midline anterior to the anus | Constitutional |
| 11 | female | 5 years | none | midline anterior to the anus | Associated to LEA |

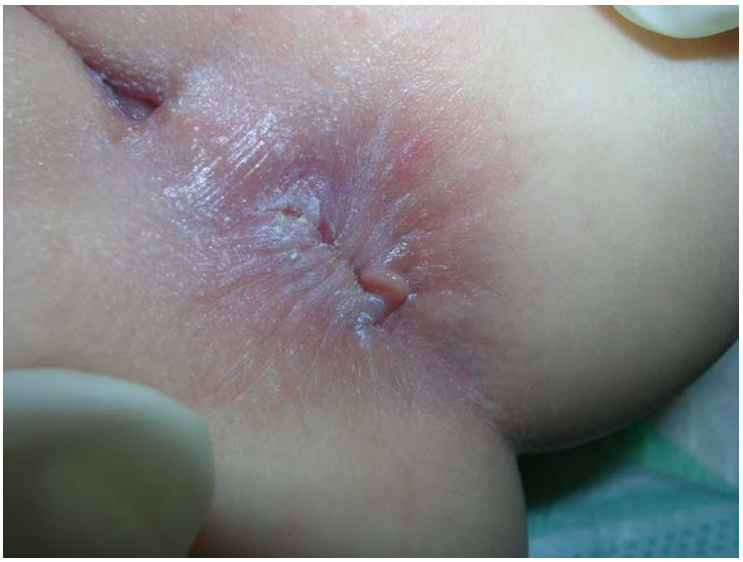

**Figure 1.** Infantile perianal protrusion located in the midline posterior to the anus.

The protrusions of the 5 and 7-year-old patients were noted about 2 weeks prior to presentation. In the case of the 3 years old patient, the IPP was an incidental finding while being examined for another issue.

Seven of our patients had the constitutional type of IPP, one of whom also had a cousin with IPP. Two patients showed the functional/acquired type, both appeared to be related to constipation. Finally, the other 2 exhibited associated LSA. In 2 cases the defect was located in the midline posterior to the anus (Figure 1). In 3 others the defect was located in the lateral perianal region, two on the right side and one on the left (Figure 2 a and b). Five patients had a protrusion in the midline anterior to the anus (Figure 3 a and b), and the last one displayed the defect in both the anterior and posterior midline (Figure 4). Nine of our patients had conservative management. In the IPP cases associated with LSA (Figure 3 a) initial topical corticosteroid treatment and then topical immunomodulators were employed.

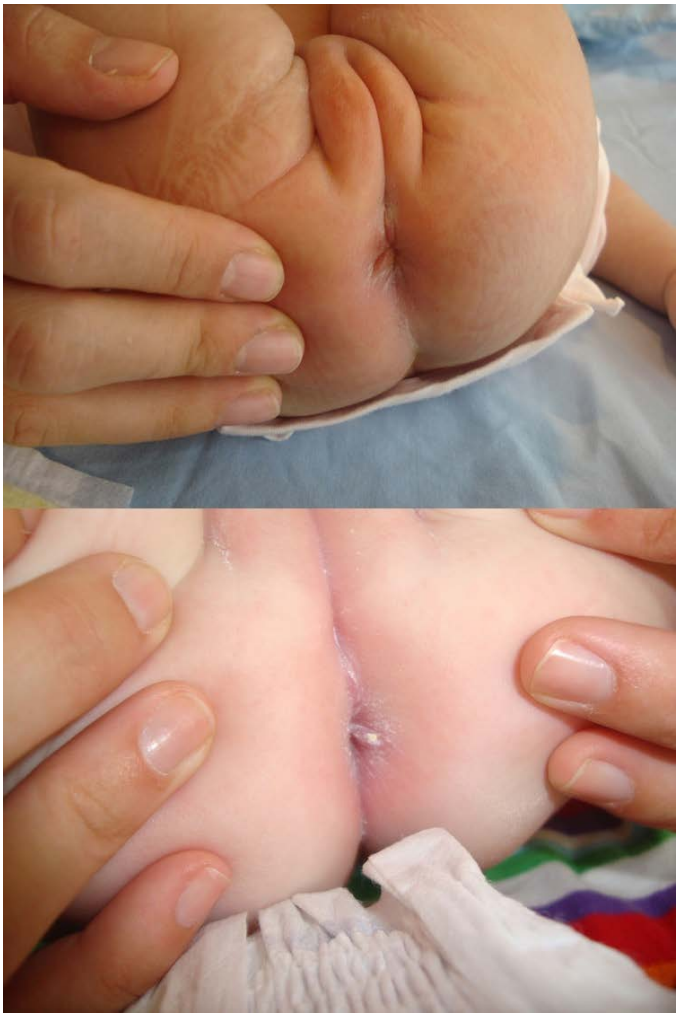

**Figure 2.** Infantile perianal protrusion in the a) right and b) left perianal region.

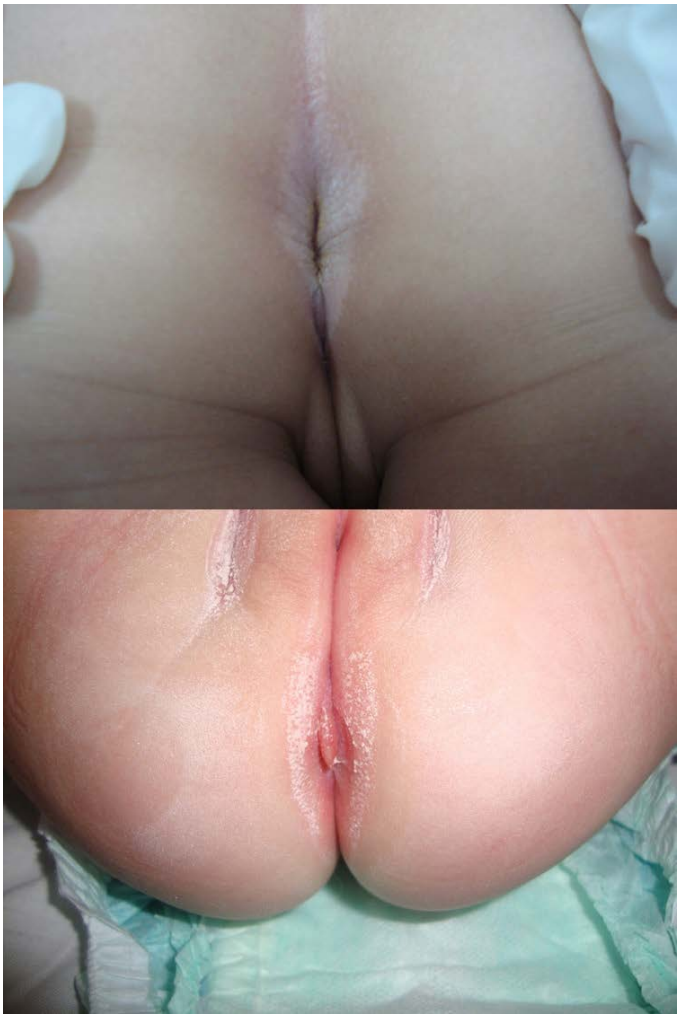

**Figure 3.** Infantile perianal protrusion in the midline anterior to the anus a) associated to LSA and b) without associations.

None of our patients had spontaneous resolution of IPP during the follow-up period.

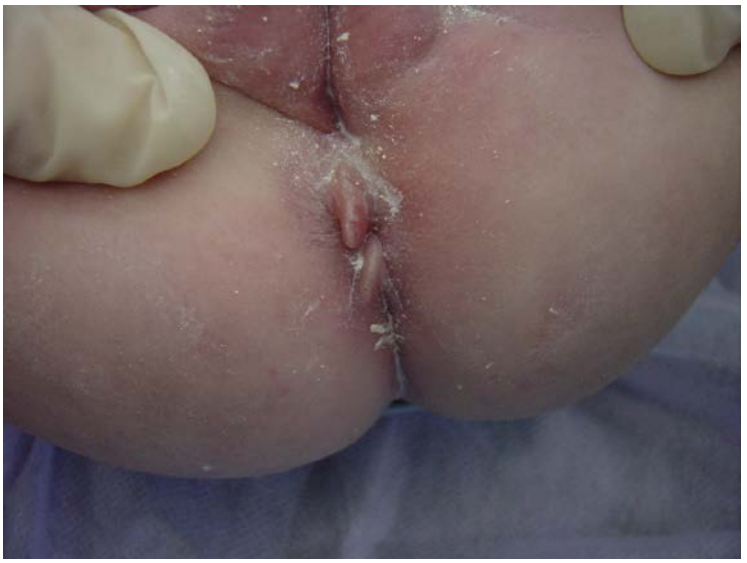

**Figure 4.** Infantile perianal protrusion in the anterior and posterior midline.

# Discussion

IPP is a benign condition characterized by a solitary protrusion covered by red to rose-colored skin located in the perianal region, usually anterior to the anus [5]. It affects both infants and prepubertal children [6]. This entity is also known as skin fold and perineal nodule [2, 7].
It was described for the first time by McCann et al in 1989 [1] in 18 prepubertal girls with no history of sexual abuse. The name infantile perianal pyramidal protrusion (IPPP) was coined in 1996 by Kayashima [5]. Cruces et al, in 1998, proposed to avoid the term pyramidal, owing to the variability in the shape of IPPs [8].

In 2002, Patrizi et al. reported the location of the protrusion not only in the midline anterior to the anus but also in the posterior midline [1]. We include in our series three patients with a lateral location of the entity, reported only once before in the literature [3].

**Pathogenesis**

The pathogenesis of the constitutional type is unknown, although it was suggested that it is related to the constitutional anatomic weakness in the perineum of females. This may explain the prevalence of this condition among girls and its presentation at birth [1, 5, 9]. In addition, the medial perineal raphe is a known site of potential weakness in both sexes [4, 9].

Other authors have proposed that the congenital type may be a remnant of a projected tip of the urogenital sinus, because the perineum is made of a lengthening of the urogenital septum during the growth of the fetus [7].

Most of the time IPP is congenital [2]. One study found that 93% of lesions were present at birth. The authors concluded that this disorder is a type of developmental abnormality that occurs primarily in children, with an incidence of 13% in the female population and a tendency to decrease in size with age [7]. This variant has also been reported to remain unchanged over the years and may have a papillomatous leaf-like shape with a pink surface [1]. Although most of our patients were female, only 2 patients in our series showed the lesion at birth. When a family history of IPP exists, it may be considered a constitutional type [1]. Only one of our cases had a positive family history.

In acquired cases of IPP (functional IPP) constipation with or without perianal fissures, diarrhea, and perianal fistulas, as well as mechanical stimulation of the perineum from wiping after defecation seems to play a role in the pathogenesis [1, 4, 5, 10]. It has been reported that functional/acquired IPP patients experienced complete resolution after cessation of constipation [1]. In these cases, the IPP presents as an expanded pyramidal skin protrusion covered by pink or light red skin and may show recurrent episodes of inflammation after defecation. Two patients of our case series presented with constipation.

Regarding the cases of IPP associated with LSA, simultaneously or asynchronously [1, 8, 11], there have been reported patients in whom IPP was the first manifestation of LSA and the diagnosis was made by histopathology. Patrizi et al. suggested that IPP may represent an early manifestation of LSA in a small group of patients with recurrent episodes of perianal erythema or may be associated with pre-existing LSA. In both cases, this may relate to the restructuring of the fibrous tissue caused by the disease [1]. It was also suggested that IPP is sometimes the main manifestation of LSA in girls, similar to the appearance of LSA in boys as phimosis. Hernandez Machin et al reported a case of a child with IPP with histologic LSA with an anal and vulvar localization [11]. Two of our patients had associated LSA.

**Diagnosis**

The diagnosis of IPP is clinical [2]. A complete physical examination should be performed focusing on signs of lichen sclerosus et atrophicus [4]. Patients with IPP, or their parents, should be questioned about both dietary and bowel habits [4].

Histologically, the epidermis and dermis are essentially normal [7], although in some cases acanthosis in the epidermis and dermal inflammation have been described [5, 7]. None of our patients underwent biopsy.

The differential diagnosis of IPP includes a sentinel fold of anal fissure [1], skin tags, genital warts, sexual abuse, rectal prolapse, perianal lesions of Crohn's disease, hemangiomas, and hemorrhoid disease [2, 4, 7, 9].

**Treatment**

Conservative management is indicated in the constitutional type of IPP. Some authors documented spontaneous resolution a few weeks after initial presentation [4, 5, 8], but that is not a common occurrence [12]. In the case of coexisting constipation, appropriate treatment can result in regression of the skin lesion [2, 9, 12]. IPP associated or caused by LSA can be treated with topical steroids [2, 12], although spontaneous regression has also been observed [2].

**Conclusion**

We consider it to be important that pediatricians, pediatric dermatologists, and pediatric surgeons are aware of this entity so that erroneous diagnosis and aggressive treatment is avoided. IPP is probably a more common condition than the number of publications would suggest.

We report a very unusual patient with a lateral perianal location of IPP and a patient with two protrusions, in the midline anterior and posterior to the anus.

# References

1. Patrizi A, Raone B, Neri I, D'Antuono A. Infantile perianal protrusion: 13 new cases. Pediatr Dermatol. 2002;19:15-8. [PMID: 11860563]
2. Zavras N, Christianakis E, Tsamoudaki S, Velaoras K. Infantile Perianal Pyramidal Protrusion: A Report of 8 New Cases and a Review of the Literature. Case Rep Dermatol 2012; 4: 202-206. [PMID: 23271995]
3. Song HS, Kim YC. Infantile Perianal Pyramidal Protrusion. Ann Dermatol. 2014 Apr; 26(2):278-9.
4. Fleet S, Davis L. Infantile Perianal Pyramidal Protrusion: Report of a Case and Review of the Literature. Pediatr Dermatol. 2005;22:151-152. [PMID: 15804306]
5. Kayashima K, Kitoh M, Ono T. Infantile perianal pyramidal protrusion. Arch Dermatol. 1996;132:1481-4. [PMID: 8961878]
6. Khachemoune A, Guldbakke K, Ehrsam E. Infantile perineal protrusion. J Am Acad Dermatol 2006;54:1046-1049. [PMID: 16713460]
7. Konta R, Hashimoto I, Takahashi M, Tamai K. Infantile Perineal Protrusion: A Statistical, Clinical, and Histopathologic Study. Dermatology 2000;201:316-320. [PMID: 11146341]
8. Cruces MJ, De La Torre C, Losada A, Ocampo C, García-Doval I. Infantile pyramidal protrusion as a manifestation of lichen sclerosus et atrophicus. Arch Dermatol. 1998;134:1118-20. [PMID: 9762025]
9. Mérigou D, Labreze C, Lamireau T, Vergnes P, Taïeb A. Infantile perianal piramidal protrusión: A marker of constipation? Pediatr Dermatol 1998;15:143-144. [PMID: 9572703]
10. Miyamoto T, Inoue S, Hagari Y, Mihara M. Infantile perianal piramidal protrusión with hard stool history. British Journal of Dermatol 2004;151: 229. [PMID: 15270897]
11. Hernández Machin B, Almeida P, Dunia L, Montenegro T, Borrego L. Infantile piramidal protrusión localized at the vulva as a manifestation of lichen sclerosus et atrophicus. J Am Acad Dermatol 2007;56: 49-50. [PMID: 17224385]
12. Kim BJ, Woo SM, Li K, Lee DH, Cho S. Infantile perianal pyramidal protrusion treated by topical steroid application. JEADV 2007, 21:247-289. [PMID: 17243973]