CASE REPORT

# Congenital perineal grove: an unusual Anorectal anomaly in females—report of two cases and literature review

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## Abstract

Congenital perineal grove (CPG) is a rare anorectal anomaly; only 65 cases have been reported in literature. Two cases who were referred for evaluation of a lesion in the perineum are reported here. The patients were diagnosed clinically as CPG in neonatal period and were initially managed conservatively. Surgery was required in one case as the lesion was persistent and symptomatic. A high index of suspicion is required for diagnosis of CPG to avoid parental anxiety and unnecessary diagnostic work-up and surgery. Surgery is required only in cases where the lesion persists or there is infection, pain, and ulceration.

**Keywords:** Anorectal malformations, Perineal cleft, Perineal grove.

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## Introduction

Congenital perineal grove (CPG) is a rare anomaly of the perineum, manifested by a reddish wet groove extending from the posterior fourchette to the anterior margin of the anus. Urethra, vagina, and anus are otherwise normal.[1,2] The paediatric fraternity is usually not familiar with the entity and misdiagnosis as anal fissure, perineal trauma, or sexual abuse is common and may lead to parental anxiety and superfluous diagnostic work-up and surgery.[3] However, CPG usually resolves spontaneously by 1-2 years of age and only a few cases may require surgical intervention.[4] About 65 cases have been reported in literature as case reports and a large case series (26 cases) by Kyong et al. [2] Two cases of CPG seen at The Aga Khan Hospital, Karachi are reported here and relevant literature is reviewed to highlight the significance of correct diagnosis, treatment, and follow-up.

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## Case 1

A two-month-old healthy baby girl (birth weight of 3.5 kg) was referred by the paediatrician in Apr 2017 as a case of constipation and fissure in Ano. Examination of the perineum revealed a wet erythematous lesion extending from the posterior fourchette to the anterior margin of the anus (Figure 1). The urethra, vagina, and anus were normal. A diagnosis of CPG was made clinically. Constipation responded to conservative measures and on follow-up at one year of age, though the perineal lesion persisted, it had narrowed. On subsequent visit at two years of age, the parents complained of remnant perineal lesion, itching, and pain on sitting.

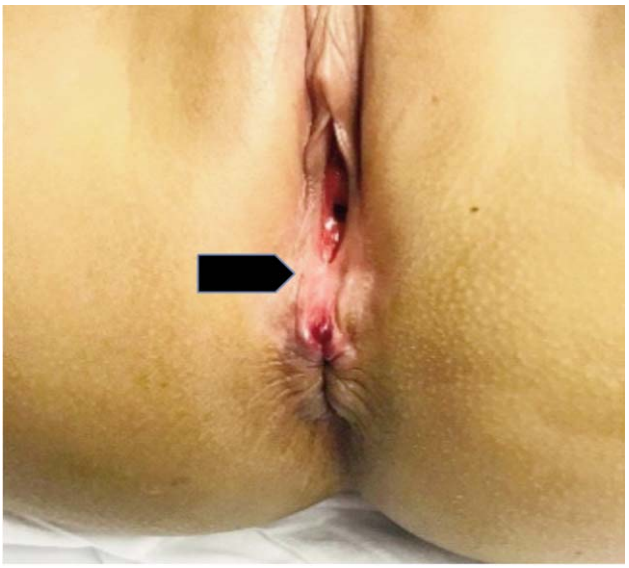

**Figure-1:** Photograph showing persistent Perineal Cleft in a two-year-old girl (black arrow)

Under general anaesthesia and antibiotic cover, the perineal lesion was excised completely to muscles and the wound was closed in layers with 4/0 PDSR (Polydioxanone) suture. Histopathology of specimen revealed stratified squamous epithelium covering, fibro collagenous tissue, and dilated thin-walled vascular channels. There was no evidence of granulomatous inflammation or malignancy. At an outpatient visit after nine months, the wound had healed, and she was asymptomatic.

## Case 2

A two-day-old female was referred by neonatology service (in late 2020) for evaluation of reddish wet lesion extending from the posterior fourchette to the anus. The introitus and anus were normal (Fig. 2). She was a product of pregnancy complicated by gestational diabetes and was born at full-term (weight 3.2 Kg) via uncomplicated

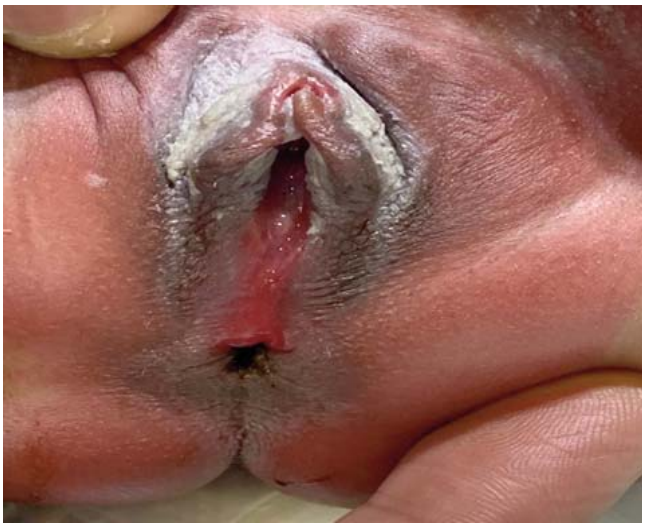

**Figure-2:** A two-day old female baby with a wide Perineal Groove.

caesarean section. She was admitted to the neonatal intensive care unit for management of hypoglycaemia. A diagnosis of perineal groove was based on physical examination. She was asymptomatic so conservative management was started with application of petroleum jelly with diaper change. Hypoglycaemia resolved in a week, and she was discharged for home. At six-months follow-up, the baby was asymptomatic and the perineal groove had almost closed. Conservative management was continued. At one and half year of age (July 2022), the perineal groove was barely visible, and she was clinically well.

## Discussion

Perineal grove or cleft (CPG) is a rare Anorectal malformation. In 1968, Stephens et al described perineal cleft as a congenital anomaly with three features: (a) a wet groove in the perineum between the fourchette and the anus; (b) normal introitus including the urethra and vagina; and (c) hypertrophy of the labia minora tails which courses posteriorly around the perineum to join the anus or encircle it.[5] Females are commonly affected by the anomaly. [6] Only three cases have been reported in males, and the anomaly was associated with hypospadias, bifid scrotum, urinary tract anomalies, and anteriorly placed anus.[5,7,8] The pathogenesis of the perineal groove remains unclear. However, there are several embryological hypotheses: (a) a relic of the open cloacal duct,[3,4] (b) failure of medial genital folds fusion in midline,[5,6] or (c) a uro-rectal septum defect during cloacal embryogenesis at fifth to eighth week of gestation.[4] Histology of the resected specimen showed the epithelium varying from non-keratinized stratified squamous epithelium, simple columnar, stratified columnar, or cuboidal epithelium of rectal mucosa with intervening area of a non-keratinized stratified squamous epithelium as seen in the reported cases.[4]

Perineal cleft or groove (CPG) is a clinical diagnosis and requires knowledge and a high index of suspicion on part of the physician. Shen et al, described two types of perineal groove: partial and complete perineal groove. The complete type is more common than the partial type and usually extends from the anus towards the perineum for a variable distance without involving the vagina and may mimic anal fissure.[1,2] Radiological imaging, e.g., ultrasound examination of the abdomen and pelvis is required in males to rule out associated urological anomalies.[5,9,10]

We reviewed all the published articles in English literature. Most of the lesions healed spontaneously, suggesting that CPG cases should be followed for one to two years of age before deciding to perform surgery. In patients who present late (> 2 years of age) surgery can be offered at presentation.

## Conclusion

CPC is a rare anal anomaly. A high index of suspicion is required for diagnosis, parental counselling, and avoiding unnecessary diagnostic work-up. It heals spontaneously as seen in one of the reported cases (Case 2). However, surgical excision may be required in cases (case 1) where the lesion is persistent, or it is complicated by infection, pain, or ulceration.

**Consent:** Written consent for using the patients' pictures was taken from the parents.

**Disclaimer:** None.

**Conflict of Interest:** None.

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