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# *Management Quandary*

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## Extensive Perineal Infantile Hemangioma with Associated Congenital Anomalies: An Example of the PELVIS Syndrome

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**Key Words.** Infantile hemangioma—Perineal hemangioma—Tethered cord—Genitourinary malformation

### Case Presentation

A 2.9-kg fullterm female was noted to have a plaque-type infantile hemangioma (IH) involving the left labial, perineal, and coccygeal skin at birth. She also had a horizontally divided right labia, anteriorly-displaced anus, and sacral dimple. Cardiac examination revealed a 2/6 systolic ejection murmur at the left upper sternal border radiating to the left axilla, consistent with peripheral pulmonary stenosis. Physical examination was otherwise normal. Abdominal, pelvic, and spinal ultrasonography on day 1 revealed a low-lying conus medullaris consistent with a tethered cord. Chromosome analysis was normal. Her bowel and bladder functions were normal.

*Discussants' comments:*

Infantile hemangiomas are the most common benign tumors of childhood. The prevalence of IH is 5−10% in the general population with a female predominance of approximately 4:1.[1] Although most IH are benign and cause no permanent defects, complications may occur depending on hemangioma size, location, and subtype (localized, segmental, indeterminate, or multifocal).[2] Ulceration is the most common complication of IH. Other complications (depending on location) include visual compromise, obstruction of airway or auditory canal, and cardiac compromise.[2] Ulcerated IH may cause bleeding, pain, infection, scarring, and/or disfigurement. Ulceration is more common in large plaque-type lesions and usually occurs during the first few months of life when IH growth is most rapid.[3] Most IH occur on the head and neck (fewer than 10% are perineal). However, the perineum is the most common location of ulcerated IH because of irritation from stool, urine, friction, and maceration.[2,3] Because perineal IH ulcerate so frequently, perineal IH tend to have the highest rate of complications and be the most likely (by anatomic location) to receive treatment.[2]

While the association between lumbosacral IH and occult spinal dysraphism (OSD) has been well documented,[1,4−6] perineal and genital IH may also show OSD, particularly when the sacral region is also involved.[1,3,4,7−10] In addition to OSD, perineal IH may be associated with numerous other congenital anomalies, including malformations of the external genitalia, urinary tract, and anus. Recently, Girard et al[10] proposed the acronym PELVIS syndrome (perineal hemangioma, external genitalia malformations, lipomyelomeningocele, vesicorenal abnormalities, imperforate anus, and skin tag) to describe these patients.

### Case

Examination in pediatric gynecology clinic confirmed the presence of a patent vagina. Examination in

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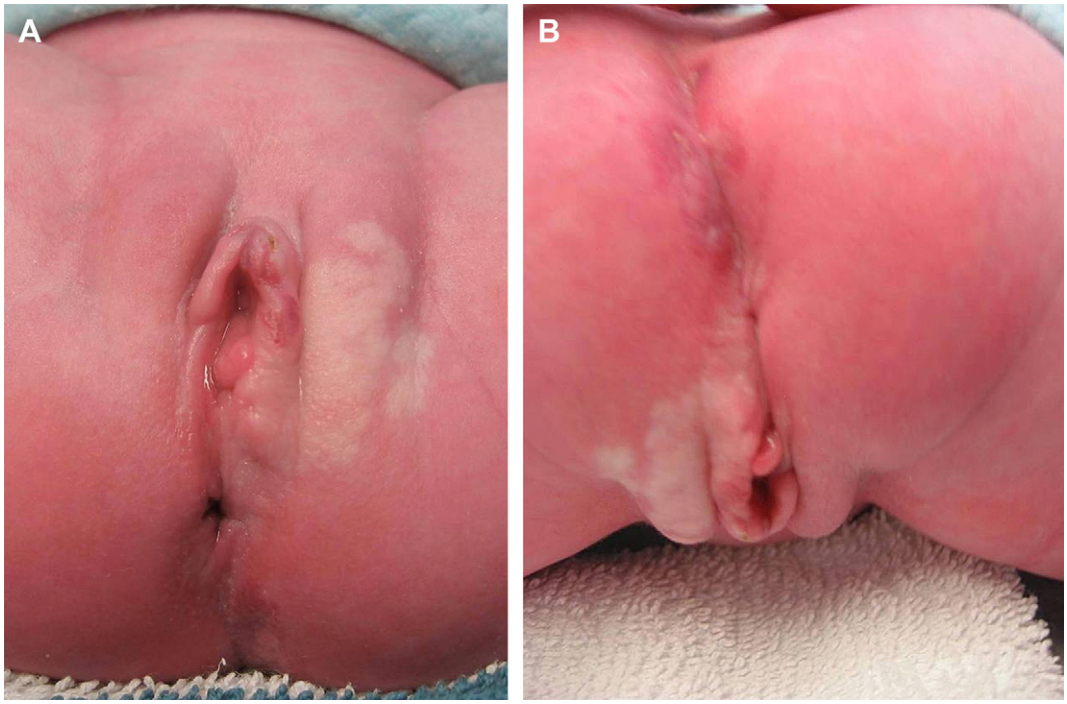


**Fig. 1. (A)** Patient at age 4 weeks demonstrating a plaque-type infantile hemangioma, horizontally divided right labia, and a hypopigmented precursor lesion on the left. **(B)** Posterior view demonstrating sacral and coccygeal infantile hemangioma, and rightward deviated anus.

pediatric dermatology clinic during the fourth week of life demonstrated progression of the hemangioma of the labia, coccyx, and sacrum since birth, without ulceration (Figure 1). She was treated with topical clobetasol cream 3 times daily. Improvement of the IH was noted at 7 weeks of age (Figure 2). Spinal magnetic resonance imaging (MRI) at 3 months of age showed a tethered cord, with filum terminale lipoma, incomplete coccyx, and small syrinx between C1-T2 and T11-L3.

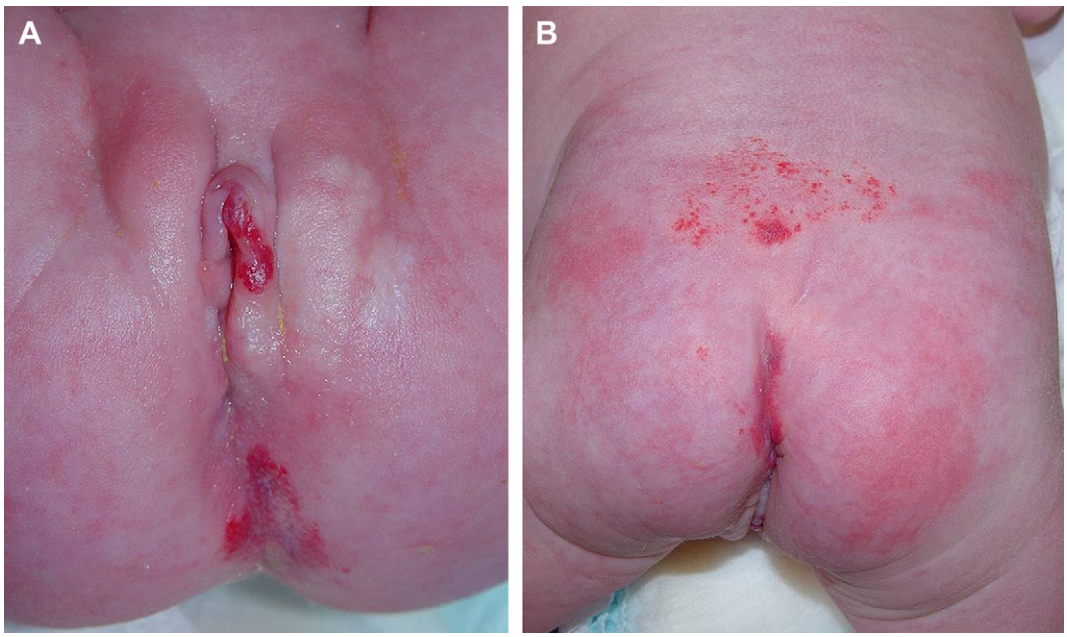


**Fig. 2.** Patient at 7 weeks demonstrating no progression of the hemangioma on anterior **(A)** and posterior **(B)** views.