



Clinical Image

Pseudo prune belly syndrome: a female case report

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1. CASE REPORT

A 14-month-old girl presented with a history of gross motor development delay. She was born full-term with adequate weight for gestational age. She had pneumonia without sequelae at 11 months. Her physical development was normal (weight, length, and head circumference of 9,800 g, 79 cm, and 48 cm, respectively). The Denver Developmental Screening Test showed 8-month-old gross motor development milestones. The physical examination highlighted a loose and globular abdomen with a small umbilical hernia and diastasis recti; superficial veins and intestinal peristalsis were observed, especially in the right abdomen; there was no visceromegaly or palpable masses; when sitting up the girl, these signs were magnified showing a prune abdomen (Figure 1); the rest of the exam was normal. Upon clinical suspicion of prune belly

syndrome (PBS), a pelvic, renal, and urinary tract ultrasound was performed, which was normal.

PBS is a rare congenital disease of unknown cause, classically defined by a triad of cardinal features including deficient or absent abdominal wall musculature, bilateral intra-abdominal cryptorchidism, and urinary tract anomalies. By strict definition, PBS affects boys, with an incidence of 3.6 to 3.8 per 100,000 live male births. Females represent less than 5% of all PBS cases, and they present with abdominal wall deficiency and a dysmorphic urinary tract without any associated gonadal anomaly. PBS may present a wide range of severity, distinguishing three major categories of presentation. Category III, also called pseudo PBS, includes patients with mild triad features or incomplete forms and whose renal function is normal or mildly impaired, and females with abdominal wall laxity [1, 2].

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Figure 1: 14-month-old girl showing a loose, globular abdomen with a small umbilical hernia and diastasis recti. Superficial veins and intestinal peristalsis are seen, especially in the right abdomen. When sitting up the girl, these signs are magnified showing a prune abdomen.

In female cases of pseudo PBS, the abdominal muscles deficiency is not only an aesthetic problem. It can also predispose, among other problems, to gross motor development delayed, because it does not allow maintaining the levels of force required to maintain balance and stabilization of the spine when performing various activities such as sitting, walking or running [2, 3]. Therefore, together with the eventual abdominal wall reconstruction, a comprehensive approach that includes rehabilitation, early stimulation and psychological support should be considered in these patients.

2. REFERENCES

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