LETTER TO EDITOR

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A Female Neonate with Pseudo-Prune Belly Syndrome

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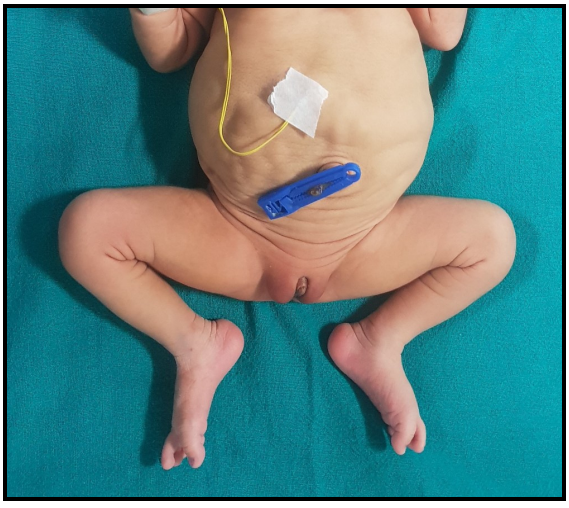
Dear Editor:

We present a case of female neonate born vaginally at 40 weeks of gestation with birth weight 2425 grams, head circumference 34.5 cm, length 49.5 cm, low birth weight and small for gestational age. Antenatal level II ultrasonogram report for congenital anomalies in baby was not available but the antenatal period was otherwise uneventful. Labor progressed normally and the baby had unassisted perinatal transition with normal Apgar score at birth. On examination, there was deficiency of abdominal wall musculature bilaterally giving it a wrinkled and dried prune like appearance (Figure 1).

Fig-1: Deficient abdominal wall musculature giving a dried prune like appearance



Facies, oral cavity, genitalia, spine and skeletal examination were normal, i.e., there was no evidence of any other gross congenital anomaly. Chest X-ray, skeletal survey, renal scan and skull ultrasound, echocardiography, detailed eye examination and hearing screen were normal. Initially, a provisional diagnosis of Prune belly syndrome (PBS) was made which was finally changed to Pseudo-prune belly syndrome (PPBS) with the fact that this female baby had normal kidneys and urinary tract [1-3]. The parents decided to postpone surgical consultation for abdominoplasty in view of ongoing COVID-19 pandemic and the baby was discharged on day 8th of life after an uneventful early neonatal period.

Prune Belly syndrome (PBS), a rare entity with an overall incidence of 1:35000-50000 live births, predominantly seen in males, is also known as Eagle-Barrett syndrome, Osler-Parker syndrome, abdominal muscles deficiency syndrome and mesenchymal dysplasia syndrome. The classical triad of PBS consists of abdominal wall muscles deficiency or hypoplasia, urological anomalies and bilateral cryptorchidism. Up to 75% of patients with PBS have associated pulmonary, skeletal, cardiac, and gastrointestinal defects [1-6].

Pseudo-prune belly syndrome (PPBS), a furthermore rare entity comprises only 3-5% of all patients with PBS. An article published in 1996 described eight patients who did not classify as classical PBS. PPBS patients may have normal abdominal wall, absent or incomplete cryptorchidism and urinary tract anomalies like that seen in PBS [1]. Patients with partial or unilateral abdominal wall deficiency, or those with unilateral undescended testis, as well as female neonates with abdominal wall laxity are also classified as PPBS [2-3].

Patients with PPBS could be presumed prognostically as mild. However, this is not so and the urinary system anomalies in PPBS

may be as severe as PBS [1-3]. This necessitates for an increased awareness of this entity among physicians because the associated anomalies guide the management and predict the prognosis.

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