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Prune Belly Syndrome: Incomplete Clinical Triad

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Clinical Image

During the pregnancy of a caucasian woman, were detected polyhydramnios, omphalocele and pyelectasis. The karyotype was normal and both parents were not non-consanguineous. The baby was delivered vaginally at 37+5/7 weeks of gestational age. Apgar score was 5I and 6V while body weight, length and head circumference were 3000 gr, 48 cm and 34 cm, respectively.

Newborn was intubated and assisted with mechanical ventilation due to respiratory distress for 30 days. A large hernia of the abdominal viscera was also observed (**Figure 1A**). The infant showed a loose not hyperelastic skin, wrinkled at the root of the limbs, micrognathia, blepharophimosis, low-set ears with hypoplastic pavilions and ante-rotated ear lobes, the left lobe with dimples overlying and mild bladder distention.

The thoraco-abdominal X-ray revealed a malformation of the ribcage with curved ribs (**Figure 1B**). Magnetic resonance image (MRI) showed thinning of the abdominal wall (**Figure 2A**) and the ultrasonography detected the hypotrophy of the rectus, oblique muscles and diastasis recti with stretching of the linea alba (**Figure 2B**). At 5 months old, the infant clinical conditions became stable; therefore, it was decided to postpone the operation.

Furthermore, Chan and Bird [1] and Digilio et al. [2] reported cases similar to Prune Belly phenotype with vertical transmission and isolated abdominal defect. We considered Prune Belly syndrome also if it was characterized by triad: cryptorchidism, abdominal wall and urinary tract's abnormalities. Our patient, due to the incomplete triad, could be considered a PBS type III according to Woodard classification [3].



Figure 1 (A) The large abdominal hernia was noted on clinical examination and (B), the malformation of the rib cage was detected by thoraco-abdominal X-ray.

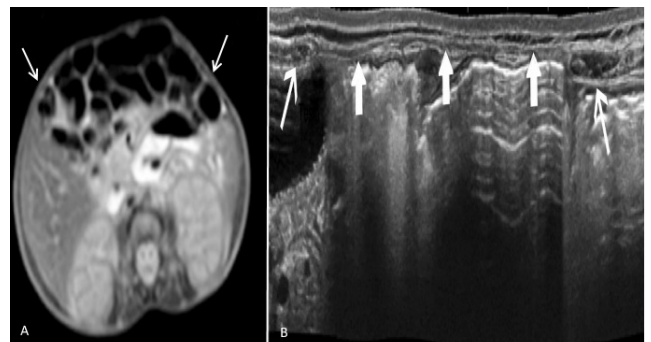


Figure 2 (A) T2-wighted MRI imaging showed significant thinning of the abdominal wall (thin long arrow) and (B), ultrasonography (LongView reconstruction) detected diastasis of hypotrophic recti (thin long arrow) with stretching of the linea alba (solid arrow).

References

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