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Interest of Antenatal Diagnosis of Prune Belly Syndrome

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1. Clinical Image

We present the case of a 2-month-old male infant born to non-consanguineous parents. He was delivered prematurely at 35 weeks of gestation by cesarean section due to severe pre-eclampsia, with a birth weight of 2400g, a height of 45cm, and a head circumference of 34cm. The mother, aged 33, has had three pregnancies with three healthy siblings. An antenatal ultrasound performed at 26 weeks of gestation revealed a distended bladder with left urethral dilation and left ureterohydronephrosis with junction syndrome. Clinical examination at birth revealed hypoplasia of the abdominal musculature and bilateral cryptorchidism, without other signs of skeletal deformities, thoracic, gastrointestinal, or cardiac anomalies related to the syndrome. Abdominopelvic ultrasound confirmed moderately differentiated kidney on the right and poorly differentiated on the left, right pyelocaliceal dilation with tortuous ureter, and left pyelocaliceal dilation with junction syndrome. The bladder wall appeared scalloped and thickened in places, with poor individualization of its anterior wall. Bilateral undescended testicles were also detected, with a voiding cystourethrogram showing passive vesico-urethral reflux grade V. Laboratory tests, including renal function, were unremarkable and within normal limits. Based on the triad of hypoplasia of the anterior abdominal wall muscles, urinary tract dilations, and bilateral cryptorchidism, the diagnosis of Prune Belly syndrome was made. The infant underwent vesicostomy and circumcision, and the postoperative course was uneventful. The infant is scheduled for regular medical follow-up to ensure appropriate monitoring and possible surgical intervention.

The term Prune Belly syndrome was first described in 1839 by Fröhlich, and in 1895, Parker provided the initial description associating urinary tract abnormalities with the syndrome. In 1950, Osler coined the term "Prune Belly" due to the appearance of the abdominal wall. Its incidence is estimated at one case in 40,000 births, with a male predominance. The exact etiology of this syndrome remains unknown.

Diagnosis of Prune Belly syndrome can be established antenatally between the 12th and 22nd week of gestation. In early pregnancy, the syndrome should be suspected in a male fetus presenting with a cystic intra-abdominal image, which corresponds to bladder dilatation. Prune Belly syndrome is typically detected in its complete form from the 20th week of gestation onwards. It is a rare congenital anomaly characterized by a triad of aplasia of the anterior abdominal wall muscles, cryptorchidism, and urinary tract abnormalities. Urinary system malformations include renal dysplasia, hydronephrosis, dilated and tortuous ureters, bladder enlargement, and sometimes diverticulum near the vesicoureteral junction, as well as urethral obstruction. Up to 75% of patients also have as-

sociated pulmonary, skeletal, cardiac, and gastrointestinal abnormalities. Despite medical and surgical interventions, mortality in Prune Belly syndrome remains high. Prenatal diagnosis plays a crucial role in the early detection of PBS.

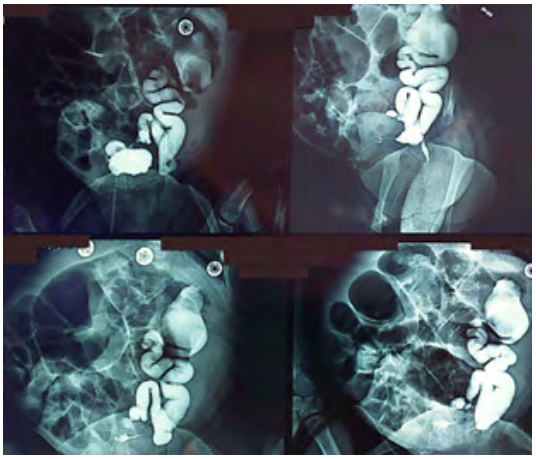

Figure1: UCG showing passive vesicourethral reflux grade V

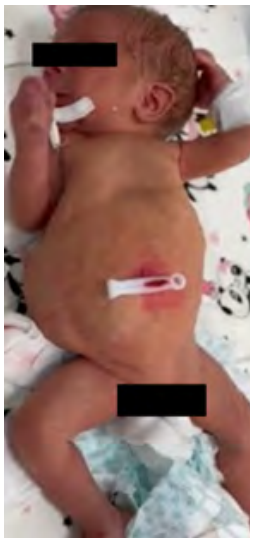

Figure 2: Image of our patient showing the wrinkled and thinned appearance of the abdominal wall

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