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Prune Belly syndrome: A case report

ABSTRACT :
Prune Belly Syndrome (PBS) is a rare congenital malformation that primarily affects boys. It is characterized by hypoplasia of the abdominal wall muscles, urological anomalies, and bilateral cryptorchidism. Although the etiology remains unclear, genetic factors and chromosomal abnormalities are suspected. Early diagnosis is essential, as the prognosis ranges from unfavorable to near-normal survival. We report the case of a premature neonate diagnosed with PBS at birth, who required surgical intervention but unfortunately passed away at 8 months due to urinary tract infections and severe dehydration. A multidisciplinary approach is crucial to improve patient outcomes.

Key Words: prune belly syndrome, abdominal wall musculature deficiency, bilateral cryptorchidism, urinary tract anomalies

INTRODUCTION:

Prune Belly Syndrome (PBS), also known as Triadic Syndrome, Eagle-Barrett Syndrome and Abdominal Muscle Deficiency Syndrome, is a rare congenital malformation, occurring in around 1 in 30,000 births, and mainly affecting boys in 96% of cases. Although the etiology of Prune Belly syndrome remains largely unknown, several hypotheses suggest a genetic origin, as well as a possible association with chromosomal abnormalities, such as trisomy 18 and 21.(1)(2)
This syndrome is characterized by a triad of clinical anomalies: hypoplasia of the abdominal wall muscles, resulting in a wrinkled, plum-like abdominal skin appearance, bilateral cryptorchidism; and urinary tract anomalies, such as bilateral hydronephrosis, megacyst and megaureter. In addition, it is often associated with pulmonary, cardiac, skeletal and gastrointestinal malformations, affecting up to 75% of patients.(3)
Early diagnosis of Prune Belly Syndrome (PBS) is crucial, ideally from birth or antenatally, to ensure prompt care and improved management of complications. (4)
The prognosis of infants with PBS is generally unfavorable, with frequent in utero or neonatal death, although less severe forms may allow near-normal survival. This variability in prognosis, combined with the diversity of malformations, makes the clinical management of PBS complex and requires a therapeutic approach tailored to the severity of each case.(5)
We report here the clinical case of a premature male neonate diagnosed with Prune Belly Syndrome on clinical examination at birth. This case is presented because of the rarity of this congenital anomaly.

CASE REPORT:

This was a male newborn admitted at birth for the management of prematurity at 32 weeks of gestation, associated with an abdominal wall anomaly. The infant was born to a non-consanguineous couple, the mother aged 36, with a gravida of 2 and para of 3. She had no history of diabetes, took no medication during this pregnancy, and had no family history of genetic or congenital anomalies. The pregnancy was a bichorionic, biamniotic twin pregnancy, with an antenatal diagnosis of abdominal malformation and malformative uropathy in the male fetus, and no notable

morphological anomalies in the female fetus. The delivery was vaginal, with a birth weight of 1800 g and Apgar scores of 9 and 10 at 1 and 5 minutes, respectively.
Clinical examination of the newborn revealed a distended abdomen, aplasia of the anterior abdominal wall muscles, palpation of the intestinal loops under the skin, thin, wrinkled skin, and visible peristalsis. The kidneys and bladder were palpable (figure 1), suggesting a diagnosis of Prune Belly syndrome. The examination also revealed hypotonia and facial dysmorphia, characterized by a triangular face, a large nasal pyramid, low-set ears, and bilateral cryptorchidism, reinforcing the diagnosis of Prune Belly syndrome. The rest of the somatic examination was unremarkable.
Abdominal ultrasound revealed a massive unilateral megaureter, a dilated bladder, poorly differentiated hyperechoic kidneys, and bilateral hydronephrosis. Given the renal anomalies, a vesicostomy was performed during hospitalization, with a straightforward postoperative course. The infant was stabilized and discharged after appropriate management, with close medical follow-up planned.
The patient was rehospitalized several times at 1.5 months, 3 months, 4 months, and 5 months of age, for resistant urinary tract infections and episodes of dehydration. Finally, at the age of 8 months, he died of severe dehydration and sepsis.

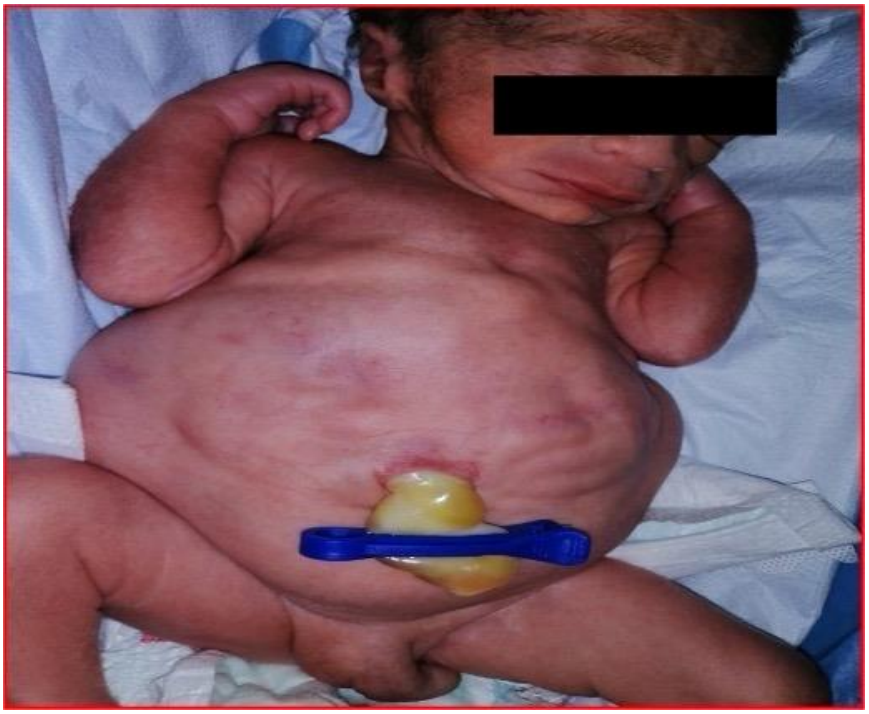

Figure 1: Image of our patient showing the prune-like appearance of the abdomen.

DISCUSSION:

Prune Belly Syndrome (PBS), also known as Eagle-Barrett syndrome, is a rare and complex condition affecting multiple organ systems. It is primarily characterized by a triad of symptoms: urinary tract anomalies, abdominal muscle deficiency, and bilateral cryptorchidism. This triad was first described by Parker in 1895, but since then, other musculoskeletal, cardiovascular, pulmonary, and genital malformations have been observed, illustrating the diversity and complexity of this syndrome.(1)

The etiology of PBS remains largely unknown, although genetic hypotheses have been proposed. Family cases have been reported, suggesting possible hereditary transmission. However, the exact mutations responsible for PBS are still not fully understood. This lack of genetic knowledge makes both diagnosis and prognosis of the syndrome particularly challenging.(6)

The prognosis for infants with PBS varies considerably depending on the severity of the malformations. In the most severe forms, complications such as renal failure, pulmonary hypoplasia, or cardiac malformations can lead to early death, sometimes before or shortly after birth. In contrast, in less severe cases, patients may have a near-normal life expectancy with appropriate medical follow-up. This variability in prognosis presents a major challenge for clinicians, as each case requires a precise assessment of the severity of the anomalies.(7)(8)

In our case, the patient exhibits the classic features of PBS, including abdominal hypotrophy, renal malformations, and urinary anomalies. This case highlights the importance of early detection, especially in severe forms where major complications such as renal failure or recurrent urinary tract infections may arise. The diagnosis is primarily clinical, but additional investigations such as ultrasound and magnetic resonance imaging (MRI) are essential for assessing the extent of renal malformations and urinary function, which guides treatment.(9)

Abdominal malformations are another key aspect of PBS. In our case, the absence of abdominal muscles led to visible distension of the abdominal wall, which places the patient at increased risk of hernias and respiratory complications due to the weakness of the muscle wall. Managing these complications requires close monitoring and, in some cases, surgical interventions.(10)

Treatment of PBS requires a multidisciplinary approach. The medical team should include neonatologists, nephrologists, urologists, and other specialists depending on the anomalies present. Therapeutic strategies vary according to the severity of the malformations. For example, close monitoring is crucial in mild forms, while more complex interventions, such as voiding cystourethrography and placement of a suprapubic catheter, may be necessary in cases of suspected urinary obstruction or renal insufficiency. Orchidopexy is commonly performed to address cryptorchidism, while chest X-rays are conducted to exclude pulmonary complications such as pneumothorax or pulmonary hypoplasia.(10)(11)

Despite the significant challenges associated with managing PBS, there are prospects for improving prognosis thanks to advances in diagnostic and treatment options. Cases of patients who have survived into adulthood after abdominal reconstruction surgery and urinary tract repair have been reported. These advances, while encouraging, should not overshadow the persistent challenges clinicians face. Early intervention and a personalized approach are essential for improving the quality of life for patients with this rare disease.(12)(13)

CONCLUSION:

Prune Belly syndrome is a rare condition primarily observed in boys. Renal failure and pulmonary hypoplasia are the major causes of mortality associated with this

syndrome. A better understanding of this disease, as well as the possibility of antenatal diagnosis, could improve a prognosis that is still often unfavorable. It is also crucial to consider a multidisciplinary approach involving both the pediatrician and pediatric surgeon to optimize care and improve the prognosis for patients.(14)

Conflicts of Interest:
The authors declare no conflicts of interest.

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