

Renal complication due to prune belly syndrome: A case report

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ABSTRACT

Introduction: Prune belly syndrome (PBS) is a congenital disorder characterized by injury or hypoplasia of the abdominal wall musculature, severe urinary tract abnormalities, and bilateral cryptorchidism in males. It has an estimated incidence of 1 in 40,000 live births. Life expectancy is low, and most affected individuals do not survive for more than one year. About 30% of survivors develop chronic renal failure and require a kidney transplant. **Case Report:** A male patient, delivered by cesarean section at 39 weeks of gestation, presented with respiratory failure, anuria, flaccid abdomen, bilateral cryptorchidism, and heartbeat with absence of abdominal muscle movements at delivery. Ultrasound of kidneys and urinary tract showed bilateral obstructive hydronephrosis, dilated ureters with tortuous aspect, and bladder partially full with normal wall thickness, suggesting low obstruction. In

addition, polycystic kidneys with reduction in renal parenchyma were observed bilaterally. Based on the signs and imaging exam, the patient was diagnosed with PBS. Peritoneal dialysis and ureterostomy were performed. At two years of age, ureteroplasty and orchiopexy were performed, and spontaneous urethral diuresis occurred. After the surgical procedure, the patient underwent hemodialysis for one year before kidney transplant. **Conclusion:** Prune belly syndrome is a rare, complex disease with high morbidity and mortality rates. Therefore, early diagnosis before birth during a routine fetal ultrasound or recognition at birth, as well as choice of the most suitable treatment requires greater knowledge of PBS and its complications.

Keywords: Abdominal wall, Cryptorchidism, Prune belly syndrome, Renal insufficiency, Urinary tract

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INTRODUCTION

Prune belly syndrome (PBS), also known as Eagle-Barret syndrome, triad syndrome, or Obrinsky syndrome, is a congenital disorder with an estimated incidence of

1 in 40,000 live births, and males account for 95% of the cases. It is characterized by absence or hypoplasia of the abdominal wall musculature, severe urinary tract abnormalities, and bilateral cryptorchidism in males [1, 2]. Patients have periumbilical skin folds, giving rise to a wrinkled or prune-like appearance. The testes are close to the ureteral segments at the height of the intra-abdominal iliac vessels. In addition, these individuals may have renal displacement and hydronephrosis [3].

It is considered a multisystemic disease, since it may present with concomitant anomalies, including pulmonary and renal hypoplasia, cardiac and musculoskeletal anomalies, imperforate anus, and varying degrees of intestinal malformation [4, 5]. In addition to the physical problems, this syndrome also negatively affects the emotional, social, and school functioning of these patients, deeply impairing their quality of life [6]. In cases this syndrome affects females, it is considered a pseudosensory syndrome, because they lack the cryptorchidism criterion [5, 7].

Patients with PBS generally have a poor prognosis, and up to 30% of those that suffer from chronic renal failure need kidney transplantation [7]. Therefore, this study aimed to report a case of a 4-year-old patient diagnosed with PBS who underwent a kidney transplant.

CASE REPORT

A male patient, delivered by cesarean section at 39 weeks of gestation, was referred to the neonatal intensive care unit (ICU), since he presented with respiratory failure at the time of delivery, requiring orotracheal intubation. In addition, he had anuria, fluid in the abdomen, bilateral cryptorchidism, and heartbeat with absence of abdominal muscle movements. Family members reported that no complications were noticed during prenatal care, except in the last trimester, when oligohydramnios was diagnosed.

In the neonatal ICU, ultrasound of kidneys and urinary tract revealed bilateral obstructive hydronephrosis, dilated tortuous ureters, and bladder partially full with normal wall thickness, suggesting low obstruction. Moreover, polycystic kidneys with reduction in renal parenchyma were observed bilaterally. Therefore, a suprapubic cystostomy was indicated and performed. Based on the signs and imaging exam, the patient was diagnosed with PBS.

During the first months of life, he had recurrent urinary tract infections, requiring the use of an oral outpatient protocol with cephalexin. At eight months of age, laboratory tests showed chronic renal failure and anemia, and the patient needed a blood transfusion. During this procedure, he had urticarial plaques and the transfusion was suspended. The child was treated with erythropoietin, hixizine, and prednisone.

He underwent a urethrocytography that identified the partially filled, elongated, thick-walled bladder, but did not identify the ureters. In addition, he had episodes

of hyperkalemia and uremia. A Tenckhoff catheter was placed and peritoneal dialysis and ureterostomy were scheduled. Three months later, he underwent bilateral nephrostomy, and since the left kidney presented ureteropelvic junction stenosis, it was resected and sent for analysis. After that, the patient started home peritoneal dialysis five times a day.

At two years of age, ureteroplasty and orchiopexy were performed, and spontaneous urethral diuresis occurred. After this surgical procedure, he underwent hemodialysis for one year before a kidney transplant was performed. Currently, at four years of age, he presents with distended abdomen (Figure 1), hypertrichosis, and cushingoid facies (Figure 2). The patient takes doxazosin mesylate, cyclosporine, mycophenolate mofetil, amlodipine,

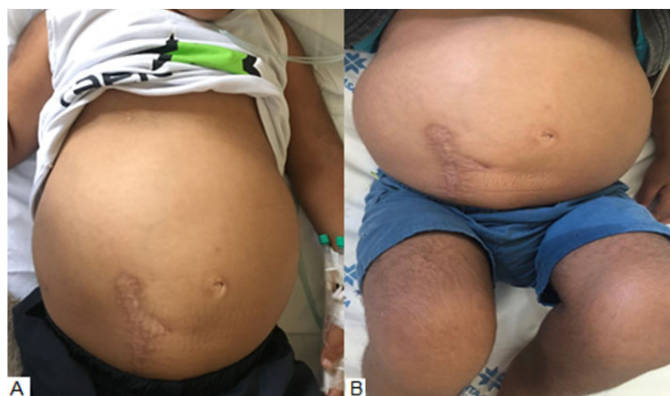


Figure 1: (A) Globose abdomen due to absence of abdominal wall musculature and a scar after kidney transplant. (B) Note the hypertrichosis in the lower limbs.



Figure 2: Face with cushingoid aspect, swollen, with hypertrichosis.

prednisone, and bactrim. He continues being followed up as a pediatric nephrology outpatient.

DISCUSSION

Prune belly syndrome is characterized by a classic triad: deficient or absent abdominal muscles, urinary tract abnormalities, and bilateral cryptorchidism [5, 8]. The genetic basis of this condition remains unknown. However, some authors affirm that it is the result of an abnormal development of the middle and lateral plate mesoderm during the third week of gestation, responsible for the formation of the urinary tract, kidneys, and abdominal wall [9–11].

Life expectancy of PBS patients is low, and up to 30% of them do not survive for more than one year. Neonatal mortality rate reaches 23%, 20% are stillborn, and 43% are premature [12].

Laxity of the abdominal wall musculature can lead to several effects, such as difficulty in urinating, impaired bowel movements, and postural changes. In addition, due to the absence of intra-abdominal pressure, the individual is more prone to develop respiratory tract infections secondary to impaired cough mechanism, and to chronic constipation because of ineffective Valsalva maneuver [12, 13]. Another frequent symptom is recurrent urinary tract infections due to incomplete bladder emptying leading to urinary stasis [14].

The diagnosis of PBS can be made using intrauterine ultrasound, usually in the 13th week of gestation. In general, ultrasound findings include bulging abdomen, lack of abdominal muscles, dilated bladder and ureters, hydronephrosis, polycystic or dysplastic kidneys, fetal ascites, oligohydramnios, and hypoplastic lungs [15, 16]. Nevertheless, if it is not diagnosed during pregnancy, at birth the above mentioned triad helps the clinical identification of PBS.

The management of PBS depends on the severity of the clinical findings. In some cases, prenatal intervention using vesicoamniotic shunt therapy is required to repair severe obstructive uropathies, which otherwise lead to oligohydramnios and pulmonary hypoplasia [17, 18]. After birth, it is generally necessary to perform a surgical procedure, such as a button cystostomy for adequate bladder drainage to prevent complications [19]. For patients with end-stage renal disease, hemodialysis or continuous peritoneal dialysis is indicated before kidney transplantation [20–23].

In male subjects, Fowler-Stephens orchiopexy should not be delayed until adolescence, so that spontaneous onset of puberty can be reached, resulting in satisfactory sexual function in adulthood [24]. Additionally, in many cases, abdominoplasty is performed for aesthetic or psychosocial reasons, aiming to improve intra-abdominal pressure. As a consequence, pulmonary, intestinal, bladder, and postural functions are also improved [6, 25–28].

CONCLUSION

Prune belly syndrome is a rare, complex disease with high morbidity and mortality rates. For a better prognosis, PBS diagnosis should be made early, preferably in utero. Physicians and pregnant women should be alerted to the importance of prenatal care, inasmuch as the early detection and identification of congenital abnormalities allows health professionals to define proper treatment. Further studies on the pathogenesis, diagnosis, and management of patients with PBS are required to acquire greater knowledge of this disease and its complications since the literature on the syndrome is scarce.

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Guarantor of Submission

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Written informed consent was obtained from the patient for publication of this article.

Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.

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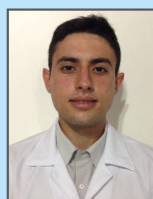
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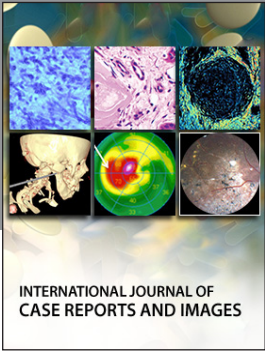
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