Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Mother-Child Radiology

A Special Case of Prune Belly Syndrome

H. El Madkouri[1*], I. Azzahiri[1], I. DAHA[1], B. Zouita[1], D. Basraoui[1], H. Jalal[1]

[1]Department of Mother-Child Radiology, CHU Mohammed VI of Marrakech, Faculty of Medicine and Pharmacy, Marrakech Cadi Ayad University, Morocco

DOI: 10.36347/sjmcr.2022.v10i10.001 | **Received:** 22.08.2022 | **Accepted:** 29.09.2022 | **Published:** 04.10.2022

***Corresponding author:** H. El Madkouri
Department of Mother-Child Radiology, CHU Mohammed VI of Marrakech, Faculty of Medicine and Pharmacy, Marrakech Cadi Ayad University, Morocco

Abstract **Case Report**

Prune Belly syndrome (SPB) is an extremely rare and complex anatomo-radiological entity with a clear male predominance, characterized by the triad made up of aplasia or hypoplasia of the muscles of the anterior wall of the abdomen, dilatations of the urinary tract and bilateral cryptorchidism. This condition is associated in 75% of cases with pulmonary, skeletal, cardiac and gastrointestinal malformations. There are also so-called incomplete or partial forms that are more common in females and are known as Pseudo-Prune Belly Syndrome. The evolution of this entity is characterized by several complications and the prognosis depends mainly on the severity of the renal damage as well as the presence of pulmonary hypoplasia. Imaging confirms the observed abnormalities. We report the case of a 4-year-old male child presenting with sudden onset abdominal distension evolving for 7 days prior to admission in which the clinical examination found a crumpled and loose wrinkle of the abdominal wall more marked on the right. Abdominal wall muscle hypoplasia was confirmed by abdominal ultrasound and computed tomography.

Keywords: Prune Belly syndrome (SPB), hypoplasia, bilateral cryptorchidism, gastrointestinal malformations.

INTRODUCTION

Prune Belly syndrome (PBS) or Eagle-barrett syndrome is a rare congenital condition with a clear male predominance (95%) [1]. which typically associates aplasia or great hypoplasia of the muscles of the anterior wall of the abdomen, urinary malformations and bilateral cryptorchidism [2-4]. It is associated in 75% of cases with pulmonary, skeletal, cardiac and gastrointestinal malformations.

There are also so-called incomplete or partial forms that are more common in females, where hypoplasia of the abdominal wall is partial or unilateral, associated or not with renal, testicular and osteo-articular malformations, which are generally on the same side. These forms are known as Pseudo-Prune Belly Syndrome (PPBS) [1, 2, 5].

The clinical forms can be very variable, and the clinical evolution is characterized by several complications. In addition, pulmonary hypoplasia due to oligohydramnios secondary to renal dysplasia is a potential cause of death after birth, but the prognosis depends mainly on the severity of the renal damage [3].

Diagnosis must be made early at birth or even better antenatally, to allow early management of this condition, the prognosis of which remains poor.

OBSERVATION

We report the case of a 4-year-old male child from a non-consanguineous marriage. Admitted to the pediatric emergency room with abdominal distention with acute urinary retention.

On clinical examination, a conscious child was found hemodynamically and respiratory stable. Abdominal examination found a thinned and wrinkled abdominal wall with a more marked saggy appearance on the right side (Figure 1), the rest of the clinical examination was unremarkable.

An abdominal ultrasound performed showed major bilateral ureterohydronephrosis associated with a mega bladder seat of urinary sediment with hypoplasia of the muscles of the anterior abdominal wall and empty testicular bursae.

Complementary abdominal CT revealed major bilateral ureterohydronephrosis associated with a

megabladder with a hypoplastic aspect of the muscles of the anterior abdominal wall.

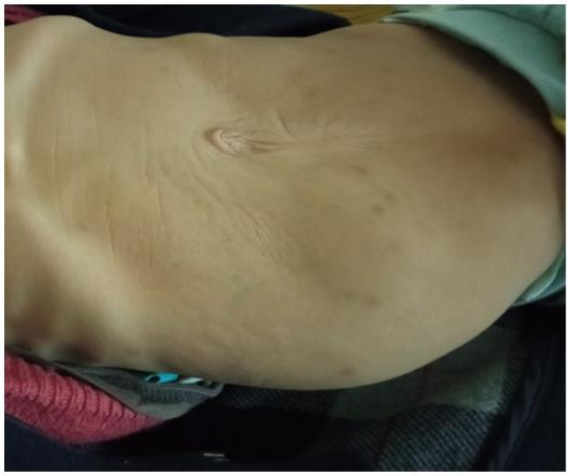


Figure 1: Thin, wrinkled and flaccid appearance of the abdominal wall

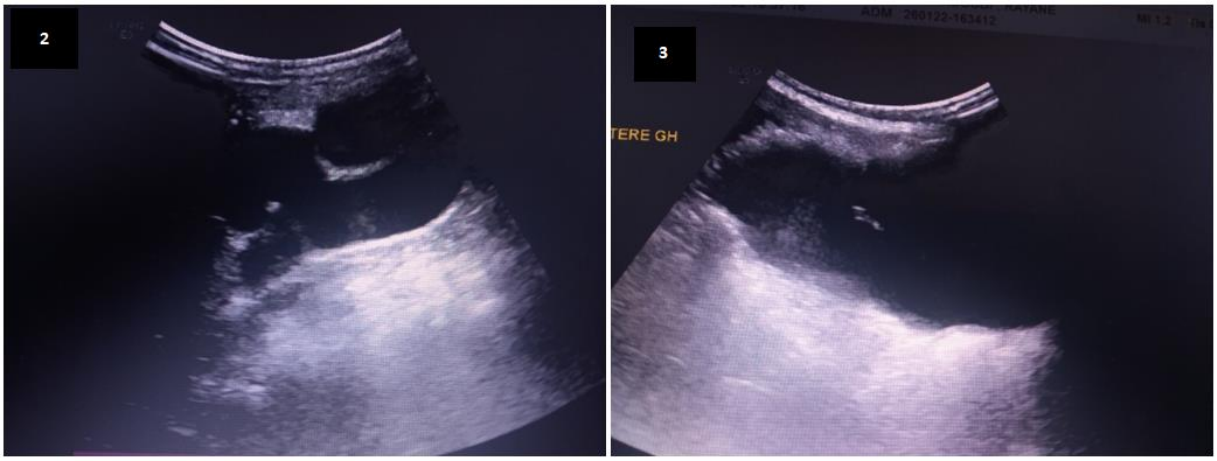


Figure 2 and 3: Abdominal ultrasound showing major ureteral dilation with tortuous appearance of both ureters

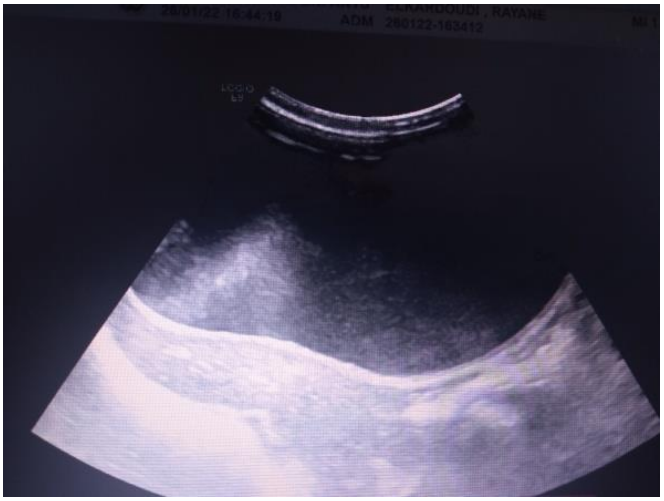
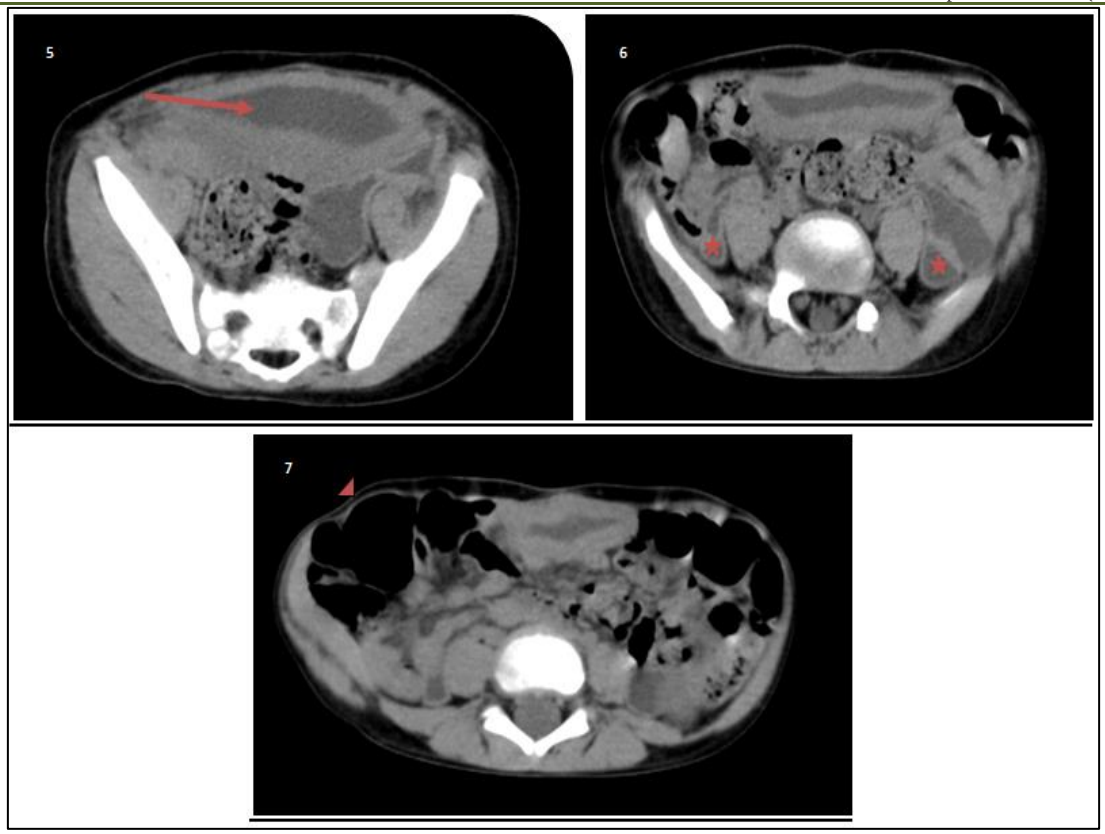


Figure 4: Abdominal ultrasound showing a mega bladder with finely echogenic content



Figures 5, 6 and 7: Abdominal CT scan without injection of contrast product in axial sections reveals a mega bladder with a thickened wall (arrow), associated with more marked mega ureters on the left (star) with hypoplastic aspect of the muscles of the anterior abdominal wall more marked on the right side (head arrows)

DISCUSSION

Prune Belly syndrome (SPB) or Eagle-Barrett syndrome was first described by the German Frolich in 1839. And it was not until 1950 that Osler named it "Prune Belly" before the appearance of the abdominal wall [6]. This syndrome is characterized by the triad: abdominal wall musculature defect (of varying degrees), abnormalities of the urinary tract and bilateral cryptorchidism.

According to various studies, the incidence of SPB is estimated at 1 case out of 40,000 births with a clear male predominance (in >95% of cases) as well as the rarity of complete forms in girls [6]. This clear male predominance has evoked a possible genetic involvement with an autosomal recessive transmission linked to the sex [7].

The exact etiology of SPB remains unknown. However, 3 theories predominate: that which proposes a prenatal obstruction of the urine which is based on embryology (proposing a failure of migration or differentiation of the lateral mesoderm at the level of the abdominal musculature) and that of the urinary tract between the 6th and 10th week of amenorrhea, and then the 3rd theory; that of the bladder sacs, which suggests dysgenesis of the velum sac and the allantois [4].

Clinically, the aplasia of the abdominal wall is by definition constant, it appears responsible for the wrinkled, crumpled and loose appearance of the skin of the abdomen, which resembles a dried plum, from which comes the name of Prune Belly syndrome [8]. This anomaly is manifested on ultrasound by an increase in abdominal circumference, thus reflecting a thinned and prominent abdominal wall.

Bilateral cryptorchidism is one of the three cardinal signs defining SPB, it is found in 95% of patients [8] and its absence casts doubt on the diagnosis.

Urinary tract abnormalities are the 3rd major feature of BPS, namely renal dysplasia, megabladder, and dilated ureters, polycystic kidney disease, hydronephrosis, and sometimes diverticulum near the vesicourethral and ureteral junction [1, 2, 8].

The diagnosis is based prenatally on obstetric ultrasound which is able to detect abnormalities of the urinary tract associated with the typical appearance of the abdominal wall as well as on the ultrasound signs which are the oligo- amnios, the mega bladder occupying entire contents of fetal abdomen, unilateral or bilateral pyelocalicial dilation, prominent thinned abdominal wall in sagittal sections [2, 9]. Post-natally, the diagnosis is essentially based on the clinic and the abdomino-pelvic ultrasound supplemented by an

abdomino-pelvic CT scan, a trans-thoracic ultrasound in search of cardiac malformation, a renal assessment to assess renal function, an ultrasound hips with a skeletal x-ray looking for skeletal malformations as well as a karyotype looking for nuclear factor 1-beta (HNF1beta) suppression [2, 10].

Therapeutic management is essentially based on surgery: abdominoplasty, orchidopexy and reconstruction of the urinary tract. For patients with mild abdominal wall dysplasia, postures are acceptable and do not require abdominoplasty. However, for severe cases, surgical treatment is discussed on a case-by-case basis. Pyelostomies, ureterostomy and cystostomies are methods undertaken in unstable patients. Kidney transplantation is inevitable for patients with kidney failure. In addition, patients with SPB require, in addition to surgical treatment, permanent multidisciplinary medical care and close follow-up [2, 3,11].

There is great heterogeneity in the prognosis of this syndrome ranging from forms incompatible with life to less severe forms whose prognosis is conditioned by the severity of pulmonary hypoplasia and urinary tract abnormalities. Pulmonary hypoplasia is the main cause of mortality in the neonatal period. Severity of urinary tract abnormalities and kidney function determine not only mortality but also long-term prognosis [2, 3].

CONCLUSION

SPB is a rare disease mainly affecting boys. It is characterized by its classic clinical triad associating abdominal muscular aplasia, bilateral cryptorchidism as well as a variety of urinary malformations. Prenatal diagnosis is now possible from an early gestational age thanks to ultrasound. Kidney failure and pulmonary hypoplasia are the main causes of death. The surgery of urinary malformations requires a case-by-case approach and its realization must be entrusted to an informed team.

REFERENCES

1. Zugor, V., Schott, G. E., & Labanaris, A. P. (2012). The Prune Belly syndrome: urological aspects and long-term outcomes of a rare disease. *Pediatr Rep*, 4, e20.
2. Soukaina, B., Houria, K., & Amina, B. (2018). Prune Belly syndrome: a special case. *The Pan African Medical Journal*, 29.
3. Xu, W., Wu, H., Wang, D.-X., & Mu, Z.-H. (2015). A Case of Prune Belly Syndrome.*Pediatrics & Neonatology*, 56, 193-196.
4. Fette, A. (2015). Associated rare anomalies in prune belly syndrome: A case report.*Journal of Pediatric Surgery Case Reports*, 3, 65-71.
5. Grover, H., Sethi, S., Garg, J., & Ahluwalia, A. P. (2017). Pseudo Prune Belly Syndrome: Diagnosis Revealed by Imaging - A Case Report and Brief Review. *Pol J Radiol*, 82, 252-257.
6. Boukoffa, S. (2019). Prune Belly syndrome illustrated by a clinical case. *Morphology*, 103, 93.
7. Masson, E. Prune Belly syndrome: epidemiological, clinical and therapeutic aspects.*EM-Consult*https://www.em-consulte.com/article/177377/syndrome-de-prune-belly-aspects-epidemiologiques-c.
8. Prune-Belly syndrome: about 2 cases. 1.
9. Pomajzl, A. J., & Sankararaman, S. (2022). Plum Belly Syndrome. In *StatPearls*, StatPearls Publishing.
10. Granberg, C. F., Harrison, S. M., Dajusta, D., Zhang, S., Hajarnis, S., Igarashi, P., & Baker, L. A. (2012). Genetic basis of prune belly syndrome: screening for HNF1β gene. *The Journal of urology*, *187*(1), 272-278.
11. Dénes, F. T., Park, R., Lopes, R. I., Moscardi, P. R. M., & Srougi, M. (2015).Abdominoplasty in Prune Belly Syndrome. *Journal of Pediatric Urology*, 11, 291-292.