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Antenatal Diagnosis of Prune Belly Syndrome

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Abstract **Case Report**

Prune Belly Syndrome (PBS) or Eagle-Barrett Syndrome is a congenital disorder that typically associates aplasia or hypoplasia of the anterior wall muscles of the abdomen, urinary malformations, and bilateral cryptorchidism. Other malformations such as pulmonary, skeletal, cardiac and gastrointestinal malformations may also be associated. Antenatal diagnosis can be done by echography. We report the case of a 32-year-old patient, referred in our formation at the CHU Hassan II of Fez at 24SA for a severe oligoamnios. Obstetrical ultrasound showed a bilaterally uretral pyelocaliciel dilatation associated with mega-bladder and dilatation of the ureter in a male fetus. An abnormality of the anterior abdominal wall was suspected with an ultrasound appearance suggestive of agenesis of the right muscles of the abdomen. The plum belly syndrome has been suspected. The patient presented a premature delivery at 32SA. Examination of the new born finds a thin, flaccid, wrinkled skin appearance, with empty purses. NN died from respiratory distress 2 hours after delivery. We discussed through this observation the various malformations found in Belly Prune Syndrome, the prenatal and postnatal ultrasound diagnosis, the fate of these children and their therapeutic management.

Keywords: Antenatal Diagnosis, Prune Belly Syndrome, congenital disorder.

INTRODUCTION

Prune Belly Syndrome is an extremely rare anatomo-radiological syndrome that associates aplasia of the muscles of the anterior wall of the abdomen, dilations of the urinary tract and testicular malformations, thus forming the classic triad of the syndrome. However, up to 75% of patients with Prune Belly syndrome have pulmonary, skeletal, cardiac and gastrointestinal malformations. We have reported the case of a Prune Belly syndrome diagnosed prenatally in our obstetrics and gynecology department I at Hassan II CHU in Fez.

We have discussed through this observation the various malformations found in Prune Belly Syndrome, prenatal and postnatal ultrasound diagnosis, the fate of these children and their therapeutic management.

OBSERVATION

We report the case of a 32-year-old patient, without significant pathological antecedent, G3P2, with two normal vaginal deliveries without incident. The patient was referred to obstetrics and gynecology obstetrics department of the CHU Hassan II of Fez at 24SA for a severe oligoamnios.

Obstetrical ultrasound showed a mono-fetal pregnancy. The fetus was eutrophic. A severe oligoamnios was. The ultrasound revealed a bilateral pyelocalicoid urethral dilatation associated with mega-bladder and dilatation of the ureter. The fetus was a male.

An anomaly of the anterior abdominal wall was suspected with an ultrasound appearance suggestive of agenesis of the right muscles of the abdomen.

Morphological ultrasound did not show any associated cardiac or pulmonary malformation. The plum belly syndrome has been suspected. Fetal MRI was performed at 31SA showing an appearance of polycystic kidney disease and confirming agenesis of the right abdominal muscles.

The patient presented a Preterm labor at 32SA, a tocolysis and first dose of corticosteroid therapy were administered. But the patient escaped to the tocolysis

with normal vaginal delivery of a male new born. The new born weight 2kg. He initially have a 7/10 apgar with severe respiratory distress. The pediatric examination revele a thin, flaccid, wrinkled skin appearance, with empty purses. NN died from respiratory distress 2 hours after delivery.

DISCUSSION

Prune Belly syndrome is a complex and rare malformation [1]. Congenital absence of the abdominal wall was described in 1839 by Fröhlich for the first time [2]. Later in 1895, Parker made the first association with urinary tract abnormalities, and Osler was credited to name it "plum-belly." In 1950, Eagle and Barrett defined this syndrome as the deficient abdominal wall musculature, undescended testes and urinary tract abnormalities [3, 4]. For the next half century, the syndrome is considered to be most common, and most of the patients died from overwhelming infections and destruction of their urinary tract [5].

The clinical forms can vary widely from born dead, to renal and major respiratory dysplasia, to almost normal children. Sometimes, it's called the "Eagle-Barrett Syndrome".

The estimated incidence of Prune Bellu syndrome in live births is 1 in 29,000 to 50,000, respectively 3.8 per 100,000 male live births. Prune belly syndrome is almost exclusively in boys, reports in anecdotal evidence in medical literature reviews [2]. Affected girls usually do not exhibit the characteristic urogenital dysplasia, nor, of course cryptorchism [6].

This clear predominance of men evoked a possible genetic participation with autosomal recessive transmission related to the sex [1].

The exact etiology of Prune Belly syndrome is unknown. Three theories predominate [7]. the first one that proposes a prenatal obstruction of urine. The seconde one that is based on embryology and that proposes the failure of primary mesodermal differentiation between the 6th and 10th week of gestational, leads to defective bodybuilding of the abdominal wall and urinary tract.Thee third one of vesical bags, which proposes a dysgenesis of the velum bag and the allantois.

Clinically the main components of this syndrome are urinary malformations. We offen find a mega bladder, ureter and ureters dilatation, polycystic kidney disease, hydronephrosis, and sometimes a diverticulum near vesico-ureteral and urethral junction [8-10]. The state of renal function is an important prognostic determinant [8, 9].

However more than 75% of patients with Prune Belly syndrome associate other malformations, such as lung, heart, skeletal, gastrointestinal and genital malformations. These malformations were reported by Routh et al with an incidence of 25% for cardiovascular, 24% for gastrointestinal, 23% for musculoskeletal, 58% for respiratory and 15% for genital [7].

Respiratory malformations have been found in pulmonary hypoplasia and cystic adenomatosis, which may be caused differently by respiratory failure, the main cause of neonatal mortality.Gastrointestinal malformations such as mesenteric malrotation, atresia, stenosis, volvulus, anal imperforation, splenic torsion, Hirschsprung's disease and gastroschisis. Osteo-articular malformations such as clubfoot, hip dysplasia, vertebral malformations, and scoliosis. Cardiovascular malformations of permeable arterial type and tetralogy of Fallot [8].

Genital malformations like cryptorchid are present in almost all male patients, however, cavernous body abnormalities or prostatic hypoplasia has also been reported. In women, genital malformations include vaginal atresia, bicornous uterus, and urogenital sinus. There has never been a case of infertility for both women and men [11, 12].

In rare cases, abdominal wall muscle hypoplasia is unilateral, with other associated renal, testicular, and bone malformations generally found on the same side as abdominal muscle hypoplasia, thus describing the incomplete form usually more common in the female. It is also called Pseudo Prune Belly Syndrome (PPBS) [13, 14].

The diagnosis is prenatal based on obstetrical ultrasound, which can detect abnormalities of the urinary system associated with the typical appearance of the abdominal wall. The antenatal diagnosis in our observation was suspected in association with an anomaly of the anterior abdominal wall associated with bilateral urethral pyelocalicoid dilatation, mega-bladder and dilatation of the ureter.

In post-natal, it is based on abdomino-pelvic ultrasound with abdomino-pelvic scan, trans-thoracic ultrasound for cardiac malformation, renal assessment for renal function, ultrasound of the hips, with skeletal radiography for skeletal malformations and a karyotype for suppression of nuclear factor 1-beta (HNF1beta) [8,15]. The treatment is mainly based on surgery: abdominoplasty, orchidopexy and reconstruction of the urinary tract [7].

For patients with mild dysplasia of the abdominal wall, postures are acceptable and do not require abdominoplasty. However, for severe cases, surgical treatment is discussed on a case-by-case basis. Pyelostomies, ureterostomies and cystostomies are also being used to temporarily shunt urine in some unstable infants who cannot tolerate surgery. Sometimes kidney

transplantation is unavoidable for patients with kidney failure. Whether surgical treatment is undertaken or not, patients with Prune Belly Syndrome require ongoing multidisciplinary medical care and close monitoring.

If multi-stage surgery is advocated by some authors such as Woodhouse *et al*. [1, 16] This is because surgery in a time is difficult to perform. Other authors like Timoty *et al*. [17], opt for surgery in a time because:

- It is relatively easy when it is done early and completely;
- It also allows a better exploration of all the lesions, especially for the testicles which are sometimes in the upper abdominal position therefore often difficult to find in an inguinal approach;
- It saves patients and parents the psychological weight of repeating surgical procedures.

The complexity of urinary malformations means that conservative treatment keeps a good place in the therapeutic arsenal. Surgery for urinary malformations requires a case-by-case approach (both for indication and timing of surgery) and should be performed by a knowledgeable team. Testicular lowering should be done more frequently in the neonatal period to increase the chances of paternity, as well as the abdominoplasty, whose aesthetic and functional benefit is real.

Pulmonary hypoplasia is the leading cause of death in the neonatal period. The severity of urinary tract abnormalities and renal function determine not only mortality, but also long-term prognosis [7]. The overall mortality rate varies between 10 and 50%, of which 20% are stillbirths, and 50% of deaths consecutive sepsis or treatment failure in the second year of life [1].

CONCLUSION

Belly Prune Syndrome is rare and affects mostly male subjects. Renal failure and pulmonary hypoplasia are the leading causes of death. Given the absence of the classic triad it is imperative to look for other malformations given the existence of atypical forms, to undertake adequate and immediate care.

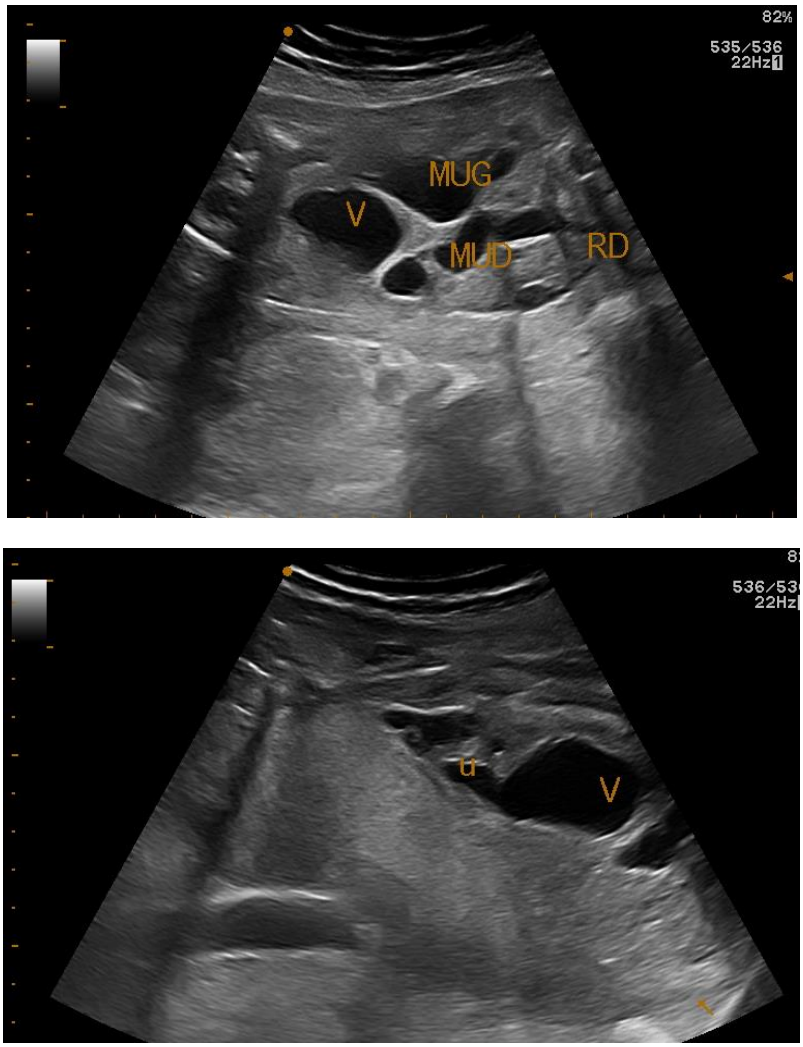
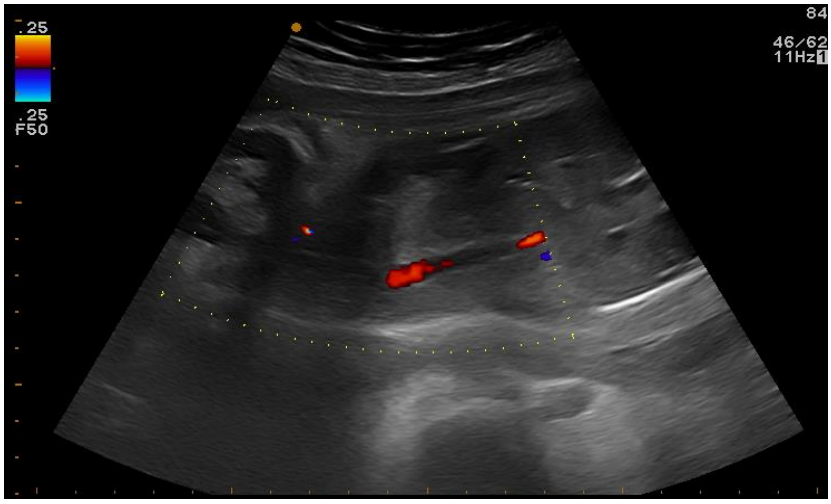
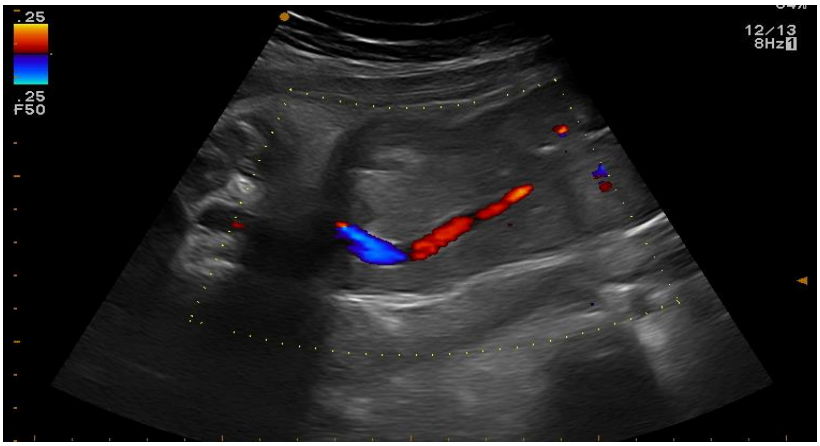


Fig-1 and 2: Ultrasound images showing ureteropyelocaliciel dilatation with mega-bladder and dilation of the ureter





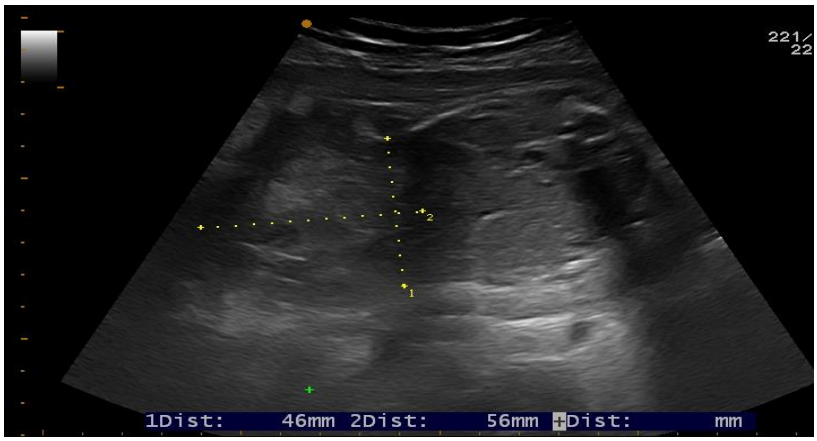
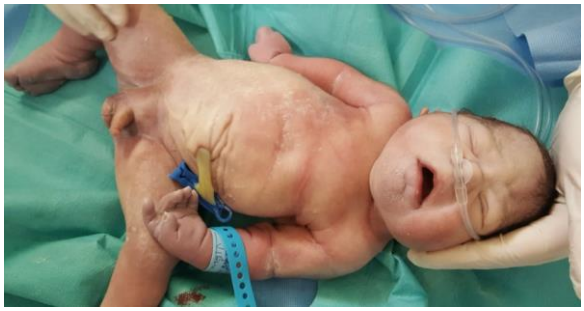


Fig-3, 4, 5: Ultrasound and color Doppler images in favor of agenesis of right abdomen muscles



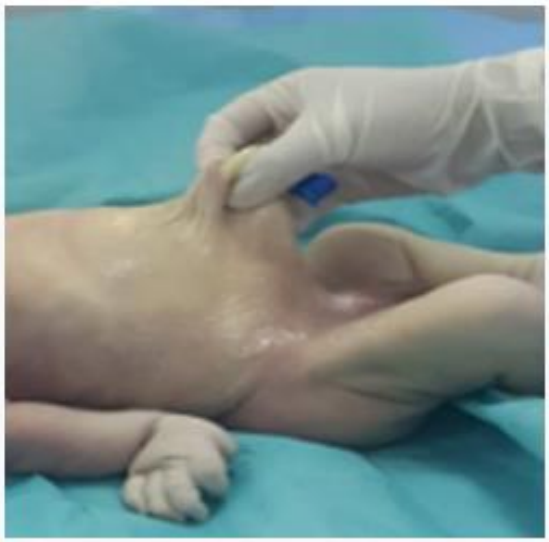


Fig-5, 6 and 7: Image of the NN with a thin, flaccid and wrinkled aspect of the skin with empty purses

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