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# Prenatal diagnosis of Prune Belly Syndrome in a case report

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***Abstract:*** *Prune Belly syndrome is a rare, predominantly male congenital malformation [1], characterised by a clinical triad of absent or hypoplastic abdominal muscles, cryptorchidism and urinary tract anomalies. In 75% of cases, it is associated with pulmonary, osteoarticular, cardiac and gastrointestinal malformations [2]. Prognosis depends essentially on urological involvement. We report a clinical case of Prune Belly syndrome diagnosed at the University Hospital of fes*

***Introduction:***

Prune Belly syndrome, also known as 'plum syndrome', is a congenital anomaly mainly seen in boys, with an estimated incidence of around 1 in 40,000 births. The typical presentation includes a poorly developed abdominal wall, undescended testes and various urinary tract anomalies (ureteral dilatation, vesico-ureteral reflux, renal dysplasia). Management requires a multidisciplinary approach, involving paediatricians, urologists and paediatric surgeons.

***Keywords:*** prenatal diagnosis , cryptorchidism , ultrasound scan

***Clinical Case :***

Our patient is 32 years old, with no particular history, third pare with vaginal delivery of two male babies with good psychomotor development, current pregnancy estimated at 22 months, well monitored, morphological ultrasound shows a Meg bladder with moderate dilatation of the left pelvis and abdominal bloating, with a male sex.

At birth, the infant had an abdominal wall that was visibly flaccid and not very muscular, giving the abdomen a 'wrinkled' appearance reminiscent of the skin of a plum.

Physical examination confirmed the presence of abdominal muscle hypoplasia. Genital evaluation revealed bilateral cryptorchidism, with the testes located in the inguinal region. No other dysmorphic signs were noted. Neonatal function was stable overall, although the enlarged abdomen required close monitoring for urinary complications.

A renal and pelvic ultrasound scan was performed on the second day of life. This revealed moderate dilatation of the urinary tract, with enlarged urethras and mild renal dysplasia, particularly on the left side. Subsequent cystography confirmed the absence of severe anatomical obstruction, but revealed mild vesico-ureteral reflux. These examinations contributed to the clinical diagnosis of Prune Belly syndrome.

Initial management was conservative. Close monitoring of renal function and antibiotic prophylaxis were introduced to prevent recurrent urinary tract infections. In addition, a paediatric surgical consultation was organised to plan the surgical management, including correction of the cryptorchidism and possible rehabilitation of the abdominal wall. Long-term follow-up was set up to assess the progression of renal function and the need for further interventions.

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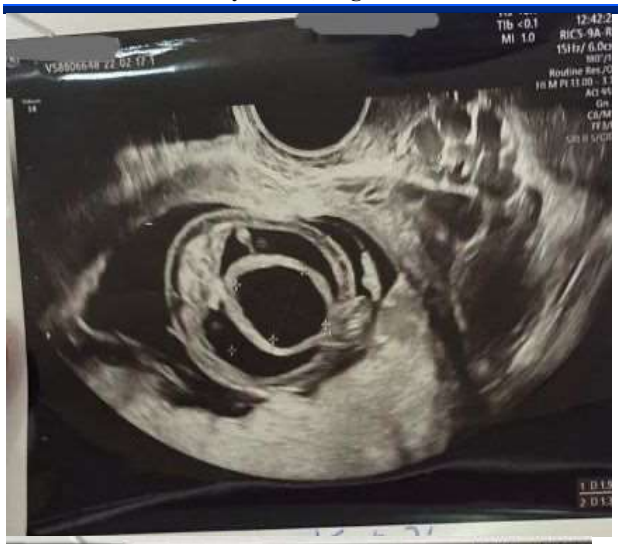
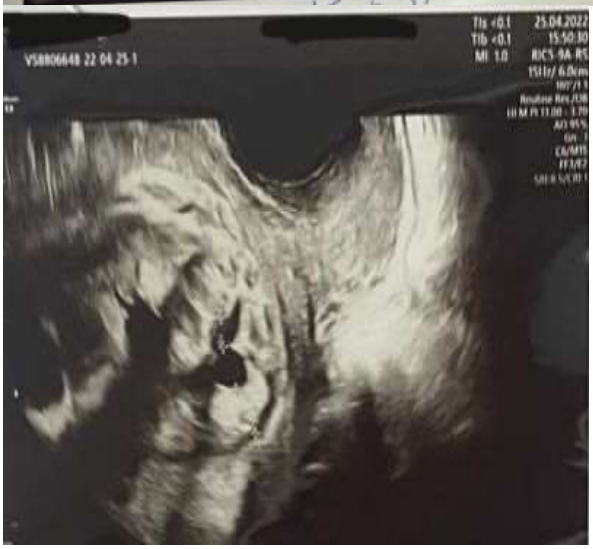
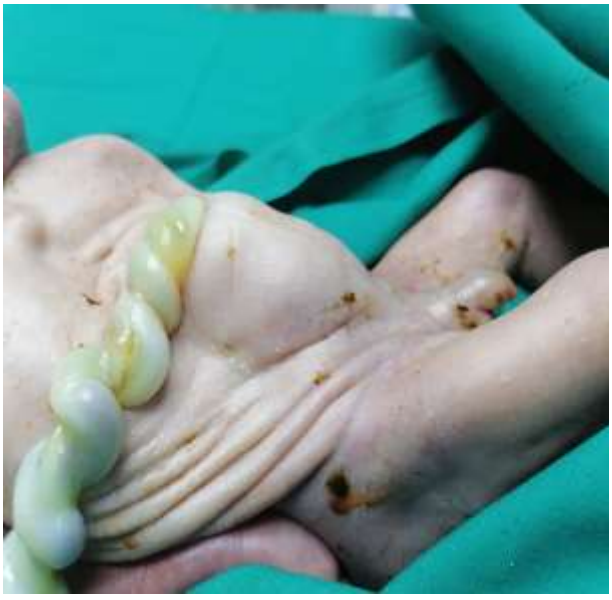




B. Dilatation du bassinet



***Discussion***

Described for the first time in 1839 by Frohlish, and defined as a triad by Parker in 1895, prune belly syndrome is a very rare anatomo-clinical entity (1/40000 births) which combines malformative muscular, urinary and genital lesions(3).
Its pathophysiology is still only partially understood, although several hypotheses have been put forward, including a malformation of the mesoderm of the lateral plate occurring between the 6th and 10th week of gestation, i.e. at the precise moment when the ureteral bud induces renal maturation. The result is maldevelopment of the abdominal wall musculature and early urinary obstruction, leading to intrauterine bladder distension.

The clinical triad - hypoplasia of the abdominal muscles, cryptorchidism and abnormalities of the urinary tract - generally allows the diagnosis to be made in the neonatal period. In our case, the diagnosis was made antenatally by morphological ultrasound and confirmed postnatally.
From birth, urological lesions dominate the vital prognosis, and are classified in 3 stages of severity: in stage 1 renal function is normal, in stage 2 renal failure is in its early stages and the course is unpredictable, in stage 3 renal failure is already established and may be accompanied by fatal septicaemia.

The routine implementation of maternal sonography has rendered prenatal diagnosis of PBS the most common presentation.(4) PBS presents prenatally with many sonogram findings comparable to that of lower urinary tract obstruction (LUTO). Although accurate diagnosis has been reported as early as 11–12 weeks of gestation, the classical findings of hydroureteronephrosis, megacystis, irregular abdominal wall circumference and/or oligohydramnios may not be consistently identified until later in pregnancy.(5) Differential diagnosis includes causes of LUTO including posterior urethral valves, ureterocele and urethral atresia as well mimics such as megacystis-microcolon-intestinal-hypoperistalsis syndrome (MMIHS).(6) Prenatal diagnosis of PBS should be considered whenever the following ultrasound anomalies are clearly identified: oligohydramnios, urinary abnormalities (dilatation of the urinary tract, megacystis, bilateral hydroureteronephrosis), and the absence of abdominal musculature. Early, accurate diagnosis allows not only for prompt multidisciplinary management of newborns in a tertiary center at birth, resulting in improved survival, but also allows for the option of voluntary termination if desired.

Management is multidisciplinary. On the one hand, monitoring of renal function is essential, as associated urinary anomalies may develop into chronic renal failure if not treated appropriately. Secondly, surgical correction of cryptorchidism must be carried out within the recommended timeframe in order to reduce the risk of complications (in particular testicular cancer in adulthood and infertility). Rehabilitation of the abdominal wall is envisaged to improve respiratory dynamics and the patient's quality of life.

The course of Prune Belly syndrome is variable and depends essentially on the severity of associated anomalies. Regular, individual follow-up is essential to adapt the treatment and anticipate potential complications. Advances in paediatric surgery and imaging have improved long-term outcomes, although the prognosis remains guarded in severe forms with advanced renal dysplasia.

***Conclusion***

Prune Belly syndrome represents a rare clinical challenge, the early diagnosis of which makes it possible to initiate appropriate multidisciplinary management. This case illustrates the importance of careful assessment in the neonatal period to identify the characteristic clinical triad and rapidly initiate a therapeutic strategy aimed at preserving renal function and correcting anatomical abnormalities. Coordination between paediatricians, urologists and paediatric surgeons remains essential to improve the prognosis and quality of life of patients with this condition.

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