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Prune-Belly Syndrome: Diagnostic and Management Challenges in Sub-Saharan Africa An Illustrative Clinical Case at CSVH "la croix" Zinvié

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Abstract: - Prune-Belly Syndrome (PBS) is a rare condition characterized by a triad of symptoms including abdominal laxity, variable urinary tract dilation, and cryptorchidism. This study presents the clinical case of a 20-year-old patient diagnosed with PBS, showing bilateral scrotal emptiness, bilateral hydronephrosis, and abdominal wall abnormalities. Complementary tests, including urogenital ultrasounds and a Uro-CT scan, revealed typical features of the syndrome. Conservative management was chosen due to the no history of recurrent urinary tract infections, nor renal dysplasia, and a huge financial constraint for more complex surgical interventions. This approach aimed to address the patient's aesthetic and psychological needs while considering economic realities. Continuous monitoring was recommended to prevent future complications.

Keywords: Prune-Belly Syndrome, Cryptorchidism, hydronephrosis, Conservative Management.

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Introduction

Prune-Belly Syndrome (PBS) is characterized by a triad of symptoms: abdominal laxity, variable urinary tract dilation, and cryptorchidism [1, 2]. This condition is mostly sporadic, with an incidence estimated at 1 in 29,000 to 40,000 births, and affects almost exclusively males [1, 2]. The syndrome may be associated with orthopedic, pulmonary, and gastrointestinal anomalies in more than 60% of cases [3, 4]. Recently, mutations in genes regulating embryonic urogenital myogenesis have been identified, leading to advances in genetic counseling [5]. A multidisciplinary approach and individualized management based on phenotypic severity are necessary. Major treatment goals include preserving renal function, managing polyuria, ensuring adequate bladder emptying, and improving body image and quality of life [5].

Observation

A 20-year-old patient, the third of five children, without significant medical history. Consulted in 2024 at the CSVH " la croix" Zinvié urology department for bilateral scrotal emptiness since birth. Parents observed this condition at birth and were advised to wait until age five and then adolescence. The patient also noticed a pot-bellied appearance after meals, visible intestinal peristalsis, and increased abdominal skin folds during fasting. Furthermore, no history of recurrent urinary tract, polyuria, dysuria, or respiratory difficulties were noted. Maternal pregnancy at age 18 was normal, and no other family members had expressed similar symptoms.

Clinical examination showed good general condition but dysuric micturition. Abdominal examination revealed notable laxity of the anterior abdominal wall muscles and bilateral scrotal emptiness. The lumbar fossae and ureteral points were normal.

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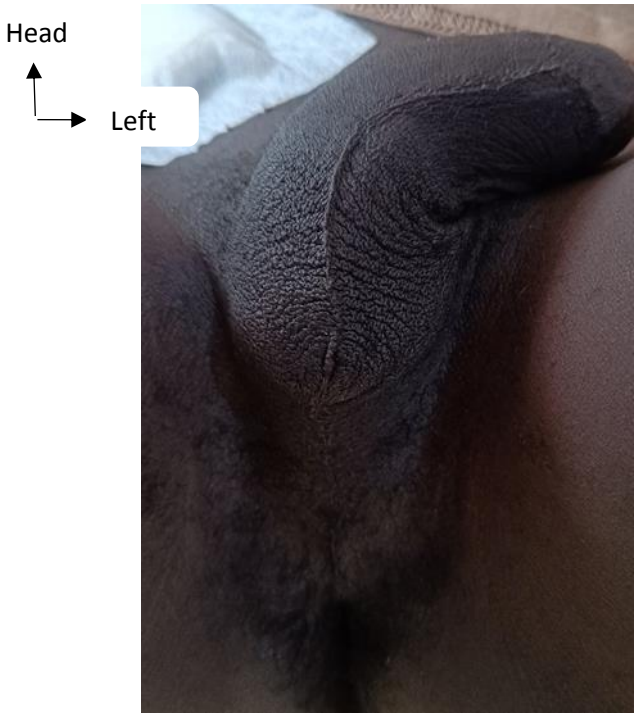

Fig 1: Bilateral scrotal emptiness

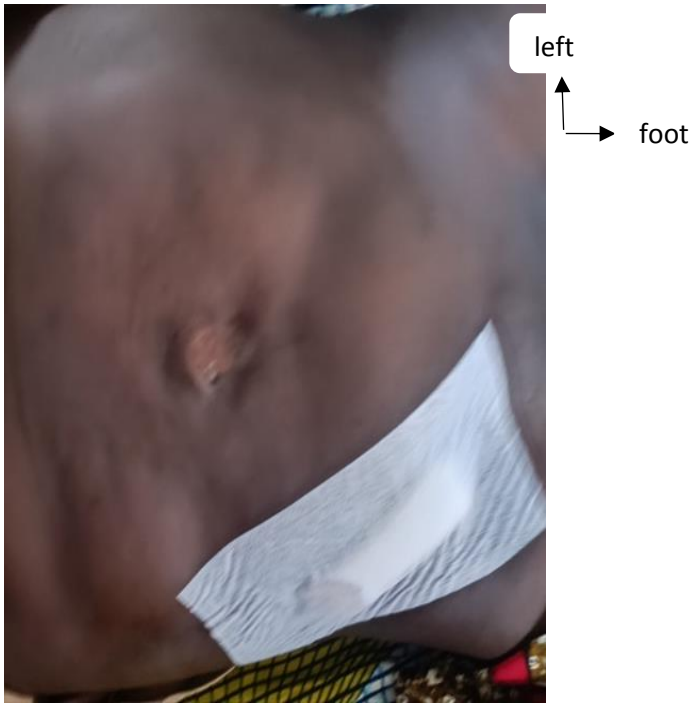

Fig 2: Flaccid and wrinkled-skin aspect of the abdominal wall

Biological analyses showed normal testosterone levels at 6.14 ng/ml, uremia at 0.1 g/l, and creatinine levels at 9.37 mg/l. Other parameters, including glucose, blood electrolytes, and urinalysis, were normal.

Inguino-scrotal ultrasound confirmed scrotal emptiness with no testicles present. Abdominal ultrasound revealed homogeneous splenomegaly, homogeneous hepatomegaly, and non-visualized testicles. Urinary tract ultrasound showed bilateral hydronephrosis with no visible obstacle. Uro-CT scan confirmed kidneys with normal size and topography, normal excretion, and moderate bilateral hydronephrosis. Cystoscopy revealed a normal-appearing urethra, visible volcano-shaped ureteral meatus, and a normal bladder wall without signs of bladder outlet obstruction.

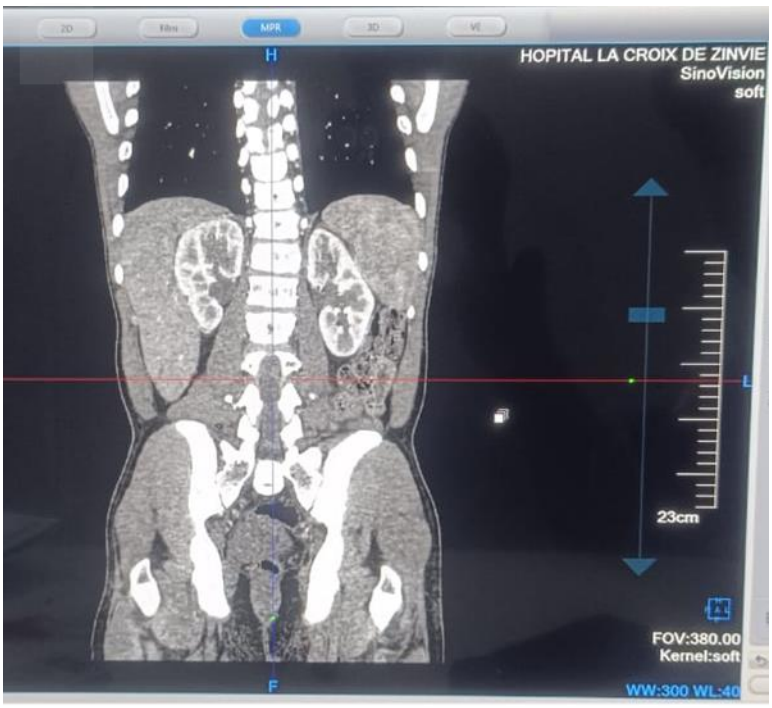

Fig 3: Bilateral hydronephrosis on Uro-CT

The diagnosis of Prune-Belly Syndrome, category III according to Woodard, was confirmed.

Management and Follow-up

A bilateral orchidoplexy attempt failed. McBurney incision revealed loose and hypotrophic anterior-lateral abdominal wall muscles and a totally atrophied right testicle. The left-side attempt was not performed after this finding.

Due to the absence of history of recurrent urinary tract, no signs of bladder outlet obstruction, moderate bilateral hydronephrosis with preserved renal function, the absence of renal dysplasia, as well as the patient's age, no abdominal muscle repair or bilateral ureteral reimplantation was performed. After consulting with the patient and parents, continuous monitoring of renal function, future loin pain, and urinary tract infection was recommended. Preventive measures were proposed to avoid fatal abdominal shock due to hypotrophy of the anterior abdominal wall muscles.

Discussion

Prune-Belly Syndrome manifests as hypoplasia of abdominal muscles, bilateral cryptorchidism, and urinary tract anomalies. The case of our patient illustrates these features with abdominal wall anomalies, undescended testicles, and non-obstructive bilateral hydronephrosis [1, 4]. The incidence of Prune-Belly Syndrome ranges from 1 in 29,000 to 40,000 births, with a 95% prevalence in males [4]. Genetic mutations involved in the syndrome are increasingly understood, enabling more precise genetic counseling [5]. Management of patients with this syndrome must be individualized based on the severity of anomalies and specific needs [4]. In our case, conservative management was motivated by the need to address aesthetic and psychological needs, as the patient wanted to feel testicles in his scrotum, and financial limitations for abdominal plastic surgery and ureteral reimplantation, a common reality in developing countries [5]. The absence of severe urinary tract complications allowed this approach, with careful monitoring to detect any future changes or complications [5].

Conclusion

Early diagnosis of Prune-Belly Syndrome is crucial for adequate management. Although the prognosis can be grim, appropriate management and continuous monitoring can improve patients' quality of life. The case of our patient highlights the complexity of this condition and the need for individualized approaches based on specific clinical manifestations [1, 4, 5].

Conflict of Interest

The authors declare no conflicts of interest.

Funding

None declared

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