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ORIGINAL ARTICLE

Challenges in the surgical treatment of a 8-year old child living with Prune Belly syndrome: case report

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Abstract

Backgroung: Prune Belly syndrome, also known as Eagle-Barret syndrome, is a rare disease, with a prevalence of 3.8 live births per 100,000 births. Its main characteristic is the hypoplasia of the abdominal muscles, giving rise to the name “prune belly syndrome”. The gold standard treatment is surgery, ideally with correction of cryptorchidism and phimosis between 6 to 18 months of life. Correction of urinary malformations and abdominoplasty should be performed up to 4 years of age. Little evidence exists in the literature about late treatment and its implications for prognosis. The context mentioned above led us to present an uncommon case of a six years old child in which a surgical approach was performed later than usual.

Keywords: Prune Belly Syndrome, eagle-barrett syndrome, Monford abdominoplasty, case report.

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Authors summary

Why was this study done?
There is little published evidence on the challenges of treating Prune Belly syndrome, also known as Eagle-Barrett syndrome, especially in late treatment cases.

What did the researchers do and find?
A ureteroscopy was performed, and there was a significant narrowing in the left ureter. Therefore, left ureteral reimplantation was chosen. The patient presented anatomical alterations: bilateral megaureter, megabladder, persistent urachus, cryptorchidism, hypoplasia of the rectus abdominis, and phimosis.

What do these findings mean?
The earlier the malformations are treated, the better clinical outcomes are obtained. Studies with a more substantial number of patients and longer follow-up times are needed to understand better and manage these patients, seeking to enhance the quality of care delivered.

INTRODUCTION

Prune Belly Syndrome, also referred to as Eagle-Barret syndrome, is a rare disease that affects about 3.8 live births per 100,000 births in the United States. It is more common in boys, rare in girls, in only 3 to 5% of cases[1]. Parker and Guthrie first described it in 1895 and Osler in 1901. However, the term "prune belly syndrome" was given by Eagle and Barrett in 1950[2].

This disease is characterized by the triad: abdominal wall defect, cryptorchidism, and urinary malformations, such as ureterohydronephrosis, renal dysplasia, and megabladder[1-3]. The etiology is not entirely understood; however, it is known that there is a defect in the mesenchymal differentiation between the 6th and 10th week of gestational age[1-3].

The earlier the malformations are corrected, the better clinical outcomes are expected[4]. Here we present a report of a late correction due to loss of patient follow-up and the possible negative implications of this delay[5].

Patient information and diagnostic evaluation

A 6-year-old male patient. Accompanied in the hospital service since birth. At birth, hy- poplasia of the abdominal muscles was observed and confirmed by a new ultrasound of bilateral ureterohydronephrosis associated with megabladder. The diagnosis was made through gestational ultrasound, which showed bilateral ureterohydronephrosis, bladder wall thickening, and abdominal wall muscle hypoplasia, characterizing the syndrome triad.

Initially followed up by the nephrology and urology team, the patient suffered recurrent urinary infections, most febrile and requiring hospitalization. The patient was scheduled for surgical treatment but lost outpatient follow-up, thus delaying definitive treatment. Figures 1 (A and B) show the patient's abdominal physical condition when returning to hospital follow-up, evidencing the hypoplasia of the abdominal muscles.

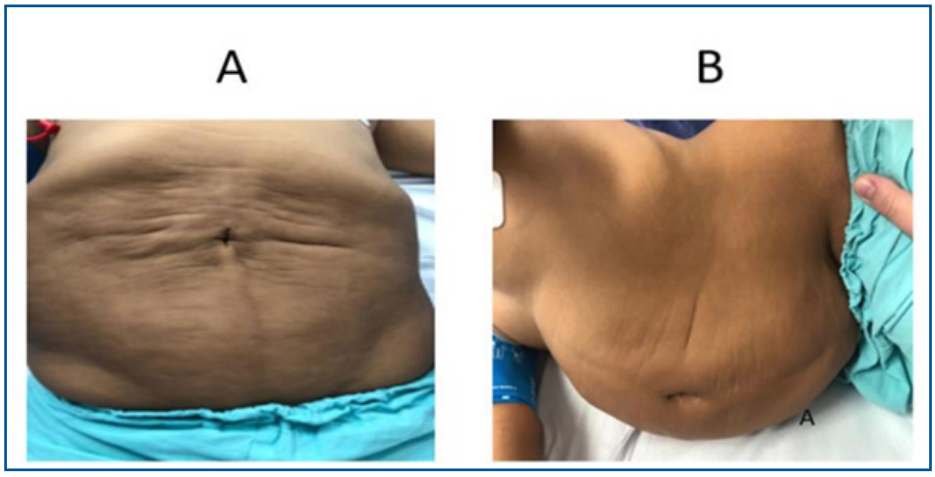

Figure 1: (A and B): Hypoplasia of the patient's abdominal muscles

Therapeutic and surgical approach

After returning from hospital follow-up at six years of age, the patient showed the fol- lowing anatomical alterations: bilateral megaureter, megabladder, persistent urachus, cryptorchidism, hypoplasia of the rectus abdominis, and phimosis.

Surgical treatment was proposed, the gold standard for correcting urinary anatomical alterations and abdominal wall defects.

Intraoperatively, a ureteroscopy was performed, which showed significant narrowing in the left ureter. Therefore, left ureteral reimplantation was chosen. Associated with ura- chal excision, bilateral orchidopexy, postectomy, and Monford abdominoplasty, as shown in figures 2 (A and B) and 3. The immediate postoperative appearance is shown in figure 4

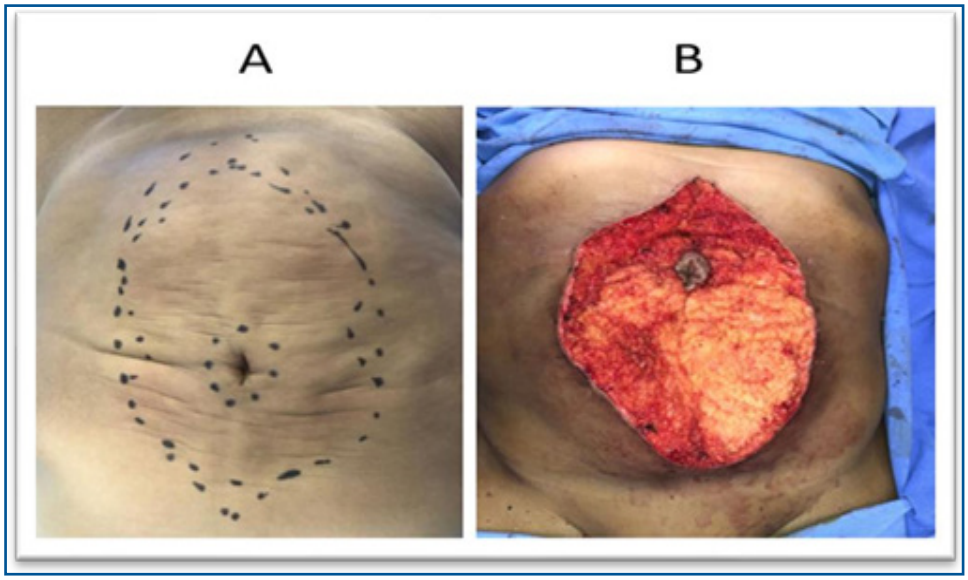

Figure 2: (A and B) shows: intraoperatively, the marking of the skin flap to be removed for the reconstruction of the patient's abdominal wall is evidenced

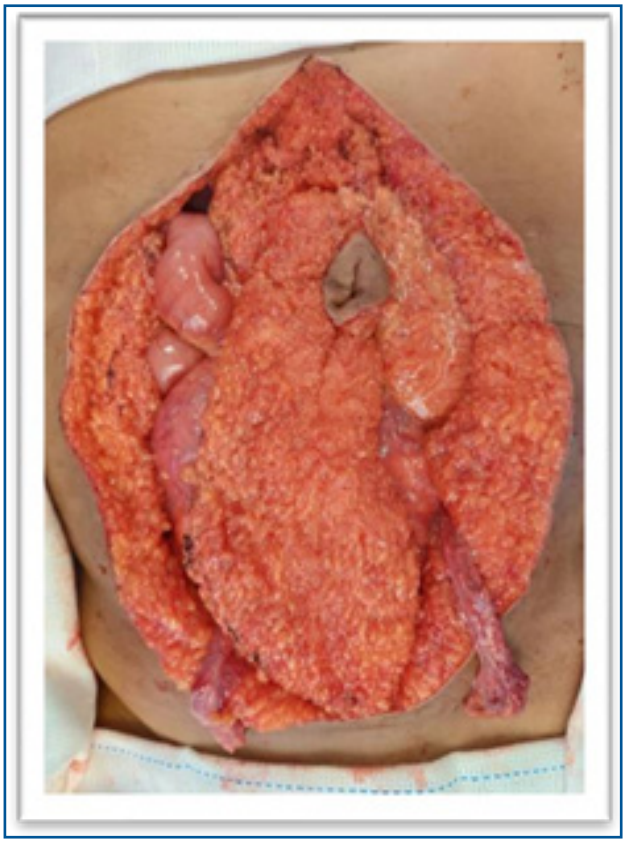

Figure 3: Incisions in the aponeurosis for intra-abdominal access and carrying out the descent of the testes into the scrotum

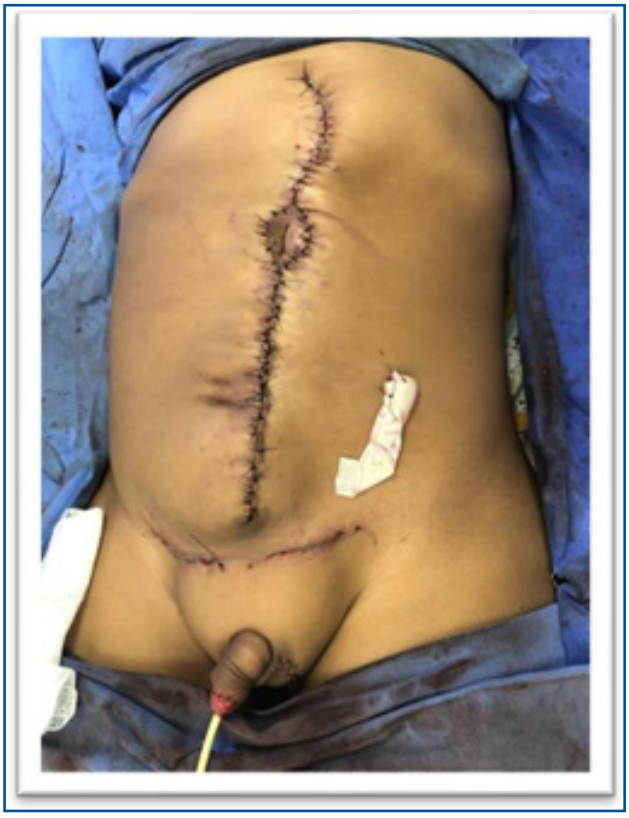

Figure 4: Immediate postoperative period, showing the appearance after Monford abdominoplasty, orchidopexy, and postectomy

Postoperative treatment and follow-up

The patient had an excellent initial clinical evolution with satisfactory postoperative di- uresis and stable renal function. A Penrose-type abdominal drain was left, removed on the second postoperative day due to low output, and the indwelling urinary catheter was removed after five days.

The patient presented adequate healing of the inguinal surgical wound and postectomy, but evolved with periumbilical skin necrosis and partial suture dehiscence on the sev-enth postoperative day.

On the 40th postoperative day, the patient underwent a skin graft in the region of the surgical wound, with the donor region on the right thigh, a procedure performed by the plastic surgery team of this institution.

He is currently under outpatient follow-up by both teams and has adequate closure of the abdominal surgical wound and good graft healing, as well as stable renal function and absence of postoperative urinary tract infection.

DISCUSSION

Prune Belly syndrome is a complex disease due to the broad spectrum of clinical symptoms. The syndrome can manifest with signs of the genitourinary, cardiopulmonary, musculoskeletal, or gastrointestinal tract4,5. Therefore, multidisciplinary management is necessary, as presented in this case, involving also different medical subspecialties such as nephrology, urology, pediatric surgery, and plastic surgery. It was also monitored by the physiotherapy and social assistance team.

The disease has different spectra of presentation ranging from I to III stages. Subtype I has high perinatal morbidity due to cardiopulmonary malformations, mainly pulmonary hypoplasia. Most children are stillborn, and newborns remain alive for a few days until death. Patients with subtype II present the full spectrum of the syndrome with the classic triad. Subtype III presents an incomplete phenotype with less severe renal alterations[4,5]. This group is the most benefited in indicating surgical approaches to correct malformations.

Surgical management with correction of cryptorchidism and phimosis should ideally occur between 6 to 18 months due to recurrent urinary tract infections and deterioration of renal function, consequently increasing the morbidity and mortality of these patients[4,5]. The period for performing abdominoplasty and correction of urinary tract malformations remains uncertain.

Seidel *et al.* report indicating corrections in children under four years old before entering school. Some reports have already shown that the combined approach with correction of urinary malformations, orchidopexy, and abdominoplasty is feasible without increasing perioperative morbidity[6].

Delayed correction of cryptorchidism is known to contribute to infertility. However, other mechanisms are associated with infertility in these patients, such as retrograde ejaculation and prostatic, seminal, and vas deferens malformations[7].

Abdominoplasty is indicated to improve aesthetics, and intra-abdominal pressure improves bladder emptying. Several techniques are described in the literature, with the Monford technique being the most used with good results[8]. Fearon et al. bring, as a result of their review, the high need for reoperation to bring the suture edges closer to the skin after correction, reaching up to 20%, as occurred in the case in question[9].

The diagnosis of rare congenital diseases is not simple and often occurs late, especially in developing countries such as Brazil, and even if it occurs promptly, treatment is often delayed due to lack of access to the health service, which may compromise the results. It is known that even when correction occurs on time, about 40 to 50% of patients will need renal replacement therapy, and 15% of them will progress to renal transplantation due to renal dysplasia[10].

CONCLUSION

Pune Belly syndrome is a disease with a broad spectrum of symptoms; therefore, it needs an individualized approach.

The literature shows that even in high-volume hospital settings, the number of cases is low, making it difficult to standardize treatment. Even with early treatments, the rate of renal dysfunction requiring replacement therapy and renal transplantation is high. Studies with a more significant number of patients and longer follow-up times are needed to better understand and manage these patients, aiming to enhance the quality of care delivered.

REFERENCES

1. Tonni, G. et al. Prune-Belly Syndrome: Case Series and Review of the Literature Regarding Early Prenatal Diagnosis, Epidemiology, Genetic Factors, Treatment, and Prognosis. Fetal and Pediatric Pathology, Guastalla, v. 31, n. 1, p. 13-24, 2012.
2. Routh, JC. et al. Contemporary Epidemiology and Characterization of Newborn Males with Prune Belly Syndrome. Urology, Boston, v. 76, n. 1, p 44-48, 2010.
3. Lopes, RI. et al. 27 years of experience with the comprehensive surgical treatment of prune belly syndrome. Journal of pediatric urology, São Paulo, v. 11, n. 5, p 1-7, 2015.
4. Lopes, RI.; Baker, LA.; Denes, FT. Modern management of and update on prune belly syndrome. Journal of pediatric urology, São Paulo, v. 17, n. 4, p 548-554, 2021.
5. Nogueira, PCK.; Paz, IP. Signs and symptoms of developmental abnormalities of the genitourinary tract. Jornal de pediatria, São Paulo, v. 92, n. 3, p S57-S63, 2016.
6. SEIDEL, N.E; ARLEN, A M; SMITH, E.A; KIRSCH, A. J. Clinical Manifestations and Management of Prune-belly Syndrome in a Large Contemporary Pediatric Population. Pediatric Urology, v.85, p.211-215, 01 de jan. de 2015.
7. Lopes, RI; Tavares, A; Denes, FT; Cocuzza, M. Gonadal function and reproductive system anatomy in post- pubertal prune-belly syndrome patients. Urology. v 145, p. 292-296, 1 de nov.de 2020.
8. Dénes, FT et al. Abdominoplasty in Prune Belly Syndrome. Journal of Pediatric Urology, v.11(5), p.291-292, 21 de jun. de 2015.
9. Fearon, JA.; Varkaraski, G. Dynamic abdominoplasty for the treatment of prune belly syndrome. Plastic and reconstructive surgery, Dallas, v. 130, n. 3, p 648-657, 2012.

10. Denes, FT; Arap, MA; Giron AM; Silva FAQ; Arap, S. Comprehensive surgical treatment of prune belly syndrome: 17 years' experience with 32 patients. Pediatric urology, v. 64, p. 789-794, mai. 2004.

Resumo

Introdução: A síndrome de Prune Belly é uma doença rara, com prevalência de 3,8 nascidos vivos a cada 100 mil nascimentos. Tem como principal caraterística a hipoplasia da musculatura abdominal gerando a origem do nome "síndrome da barriga de ameixa". O tratamento padrão ouro é cirúrgico, idealmente com correção da criptorquidia e fimose entre 6 a 18 meses e a correção das malformações urinárias e abdominoplastia devem ser realizadas até os 4 anos. Aqui descreve-se um caso ainda menos comum, em que foi realizada abordagem cirúrgica de forma mais tardia que o habitual. Há pouca evidência na literatura acerca do tratamento tardio e suas implicações em relação ao prognóstico.

Palavras-chave: Síndrome de Prune Belly, síndrome da barriga de ameixa, abdominoplastia a Monford, relato de caso.