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27 years of experience with the comprehensive surgical treatment of prune belly syndrome

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Summary

Introduction
Prune belly syndrome (PBS) presents with three main features: abdominal wall flaccidity, urological abnormalities and cryptorchidism. As a result, urologists must consider the eventual repair of the abdominal wall flaccidity and urinary tract abnormalities, and the mandatory correction of cryptorchidism, as well as decide whether to perform the procedures in a single comprehensive approach or in multiple steps.

Objectives
To report experiences with comprehensive surgical management of prune belly syndrome.

Material and methods
From 1987 to 2014, 46 children with PBS were submitted for comprehensive surgical treatment. According to individual needs, treatment aimed to correct the abdominal flaccidity, reconstruct the urinary tract, and perform bilateral orchiopexy and circumcision, which were performed in one procedure. Urinary tract reconstruction was indicated whenever pyelo-ureteral dilatation with evidence of significant stasis and/or vesicoureteral reflux was associated with recurrent urinary tract infections (UTI). Treatment for this cohort included: 44 abdominoplasties, 40 upper urinary tract reconstructions, 44 cystoplasties associated with three appendico-vesicostomies, 46 bilateral orchiopexies and 36 circumcisions. The median age at surgery was 16 months and children were followed for a median of 143 months.

Results
Abdominal appearance and tonus were improved in 90% of the children after the primary surgery and 100% after reoperation. Upper urinary tract reconstruction was performed in most children and long-term follow-up showed functional stabilization of the urinary tract in about 90% of the children, with progression to renal failure in 10%. Lower urinary tract reconstruction was performed in most children (95.6%); on late follow-up, continence was observed in 81% of them, while incontinence was present in 19% and usually associated with polyuria. Adequate bladder emptying was possible in most boys (82.6%), while the remaining required clean intermittent catheterization. Pre-operative UTI was present in 89.1% and urinary sepsis in 15.2%. Postoperatively, the incidence of laboratorial UTI was significantly reduced to 39.1%, while urinary sepsis was absent. Bilateral orchiopexy was performed in all children, with 85% of the testes becoming normal in size and well located in the scrotum.

Conclusions
Comprehensive surgical treatment is feasible and has good long-term results. A considerable incidence of reoperations due to complications or progression of the disease was observed. The long-term results for reno-ureteral anatomy and function, bladder function, infection, testicular size and location, as well as abdominal aspect and tonus, show that comprehensive surgery is an adequate method for managing children with PBS.

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Introduction

Prune belly syndrome (PBS) has an incidence of 1:35,000 to 1:50,000 live births and mainly occurs in boys. It presents with three main features: abdominal wall flaccidity, urological abnormalities and cryptorchidism [1—5]. Its characteristic, but variable, abdominal flaccidity may cause physical limitations and compromised self-image for these children. The variable anomalies of the urinary tract, which are not always proportional to the abdominal flaccidity, affect the kidneys and bladder in a non-uniform fashion [1—6]. Renal dysplasia, ureterohydronephrosis, megaureter, VUR, and bladder dysfunction are associated to recurrent UTI, which when untreated or not prevented further impair renal function in most children [1—6]. Intra-abdominal cryptorchidism is uniformly associated with infertility in these children [1—7].

Management of these children is controversial. Urologists must consider the eventual repair of the abdominal wall flaccidity and urinary tract abnormalities, and the mandatory correction of cryptorchidism, as well as decide whether to perform the procedures in a single comprehensive approach or in multiple steps.

Non-operative treatment of the urinary tract anomalies and the abdominal wall has been proposed, assuming that renal function will stabilise with prevention of UTI and the spontaneous improvement of the reno-ureteral anatomy [8,9]. Some studies have recommended urinary tract reconstruction (UTR) only in cases of confirmed obstruction and/or untreatable UTI [1,8,9]. This approach requires close surveillance and the long-term results have not been homogeneous [1,8,9].

Other studies have advocated UTR whenever stasis, VUR or UTI are present because these common factors put children with PBS at risk of progressive renal deterioration [10,11]. Because long-term continuous surveillance of these children is difficult in Brazil, due to social and geographical conditions, comprehensive surgical management of these children with PBS is preferred [6]. The aim of this study was to present the long-term results of this approach. It is believed that, to date, this experience is one of the largest reported in the literature.

Patient and methods

At initial presentation, all children with PBS underwent clinical evaluation followed by functional evaluations with ultrasonography or intravenous urography, as well as radioisotopic renography. VCUG was performed to assess the presence of urethral athresia and megalourethra in cases with severe UTI, marked ureteral dilatation or in those who pre-operatively presented with abnormal voiding patterns or urachal fistula [12—14].

Urodynamic evaluation was only performed for older children who could not spontaneously void and/or had episodes of urinary retention. Renal function was evaluated by blood urea and creatinine levels. Urinalysis and urоculture were performed in all children.

From 1987 to 2014, 46 children with PBS were submitted for comprehensive surgical treatment. Children were classified according to disease severity using the Woodard scale [1], which classifies children in three groups. All groups have the characteristic, but variable, abdominal flaccidity. In Group 1, significant renal dysplasia and pulmonary hypoplasia are present, and the children have a high rate of perinatal mortality resulting from pulmonary and/or renal insufficiency. In Group 2, despite the significant anatomical and functional abnormalities of the urinary tract, renal function is initially stable, but may progress to renal insufficiency due to recurrent obstruction or infection. In Group 3, mild urinary tract abnormalities may be present and most children present with stable renal and pulmonary function, with high survival rates without intervention.

Forty children of the cohort were classified as Woodard Grade 2, while the remaining six had Grade 3 disease. The median age at surgery was 16 months (25 days—10 years) and children were followed for a median of 143 months (15 months—26 years). Two of the children had loop ureterostomies and one had a vesicostomy performed elsewhere, while three others had patent urachuses draining the bladder. Some children were performing CIC, while others were referred with bladder catheters. In one patient with malnutrition and a single functioning kidney associated with a severely dilated ureter and recurrent UTIs, a terminal loop ureterostomy was performed before UTR in order to improve general conditions.

According to individual needs, surgical treatment aimed to correct the abdominal flaccidity, reconstruct the urinary tract and orchiopexy, preferably in one procedure. Treatment for this cohort included: 44 abdominoplasties, 40 upper urinary tract reconstructions, 44 cystoplasties associated with three appendico-vesicostomies, 46 bilateral orchiopexies and 36 circumcisions.

Abdominoplasty and laparotomy

Forty-four of the 46 children received abdominal wall reconstruction. The timing of this procedure was dictated by the need for other surgical procedures, particularly orchiopexy and urinary tract reconstruction. Three children were initially submitted to Erlich's technique and two to Randolph's repair [1], but in all 39 subsequent children, abdominoplasty was performed with the modified technique, which has previously been described [15].

Upper urinary tract reconstruction

Upper urinary tract reconstruction was indicated whenever pyelo-ureteral dilatation was associated with significant stasis or obstruction, as well as to VUR, recurrent UTI and scarring of the ipsilateral kidney (Fig. 1A). Significant stasis or obstruction was considered when progressive dilatation with worsening renal or differential renal function was observed. Reconstruction of the upper urinary tract [16] required either unilateral or bilateral distal ureterectomy and ureterocystoneostomy (Fig. 1B and C). Non-functioning units due to dysplasia or hydronephrosis were submitted to nephroureterectomy. Distal ureterectomy with ureterocystoneostomy was performed due to redundancy or dilatation, as well as VUR. Uretero-