CASE REPORT

# *Urinary ascites secondary to forniceal rupture in a child with the Prune Belly Syndrome*

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*Despite adequate bladder catheterization, a neonate with Prune Belly Syndrome developed urinary ascites secondary to forniceal rupture. Treatment consisted of bilateral cutaneous pyelostomies. Even though most children with Prune Belly Syndrome respond to lower urinary tract drainage, a cutaneous pyelostomy may be necessary when the ureters are tortuous and do not drain adequately following bladder decompression.*

**Key Words:** bladder cancer, genitourinary malignancy, cigarette smoking

## Introduction

Prune Belly Syndrome is a congenital disorder characterized by hypoplastic abdominal wall musculature that occurs in approximately 1 in 50 000 live births.[1] The syndrome is often associated with undescended testis and non-obstructive dilatation of the bladder and upper urinary tract. In neonates, bladder emptying is often incomplete and drainage can be accomplished by catheter or cutaneous vesicostomy. Many reports demonstrate that high urinary diversion is rarely indicated since most patients do well upon adequate bladder drainage. This is the first case report of a neonate with Prune Belly Syndrome who developed urinary ascites secondary to forniceal rupture despite adequate bladder drainage.

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## Case report

JB is a 2-month old infant born at 35 weeks gestation who was diagnosed perinatally with Prune Belly Syndrome. Physical examination revealed a massively distended abdomen with a prune-like appearance and partial absence of the abdominal wall musculature. His testes were undescended bilaterally. Pulmonary examination demonstrated clear breath sounds but the patient intermittently went into respiratory distress over the course of his hospitalization secondary to pulmonary

hypoplasia. A basic metabolic panel showed a blood urea nitrogen 12 mg/dl, and creatinine of 1.1 mg/dl. Renal ultrasonography revealed massive bilateral hydroureteronephrosis with moderately echogenic renal parenchyma. Voiding cystourethrogram after prophylactic antibiotics showed poor bladder emptying with no urethral obstruction and no evidence of reflux. Since the patient had poor bladder emptying and respiratory distress an 8 French foley catheter was placed for bladder drainage, which resulted in adequate decompression of bladder with significant improvement in the respiratory status. Clean intermittent catheterization was attempted but the patient was difficult to catheterize due to prostatic urethral dilation. A bladder scan demonstrated complete bladder emptying with the catheter in place. Over the next 3 weeks his abdominal distention and girth gradually increased, his creatinine rose to 2.5 mg/dl, negative urine culture and a repeat ultrasound revealed worsening hydroureteronephrosis despite an empty bladder. On day 24 the patient developed sudden increased abdominal distention and respiratory distress. Analysis of an abdominal paracentesis revealed urinary ascites. Urinary diversion was promptly performed with bilateral pyelostomies. We did not feel that a vesicostomy would be effective, as we had already accomplished complete bladder emptying with the catheter. Bilateral percutaneous nephrostomies were not performed as infants of this age tolerate tubes poorly and very difficult to manage the nephrostomies in this age group. Operative findings included a dilated right ureter and confirmed urinary ascites secondary to a ruptured renal fornix. The right kidney was decompressed due to the rupture and there was surrounding soft tissue inflammation. Post-operatively the patient's abdominal distention and urine output improved and his creatinine stabilized at 1.0 mg/dl.

## Discussion

The exact cause of Prune Belly Syndrome is unclear, but it may be secondary to a primary mesodermal abnormality. Both the abdominal wall defects and genitourinary abnormalities arise from the intermediate and lateral plate mesoderm.[1] Some authors contend that urethral obstruction that occurs early in fetal development produces megalocystis, which in turn impedes the descent of the testes and distends the abdominal wall, thus preventing its complete formation.[2] In addition, genetic factors have been sought without any identification of clear inheritance patterns.

Prune Belly syndrome is often associated with severe disorders of the genitourinary tract, including a large, hypotonic bladder, dilated and tortuous ureters, and bilateral cryptorchidism. The resulting upper urinary tract stasis and poor bladder emptying may contribute substantially to the morbidity and mortality associated with this condition. In most cases, bladder emptying will be adequate and no intervention is necessary. Adequate bladder drainage can be achieved by either clean intermittent catheterization or cutaneous vesicostomy. In our patient, a foley catheter was placed since the patient was unstable and clean intermittent

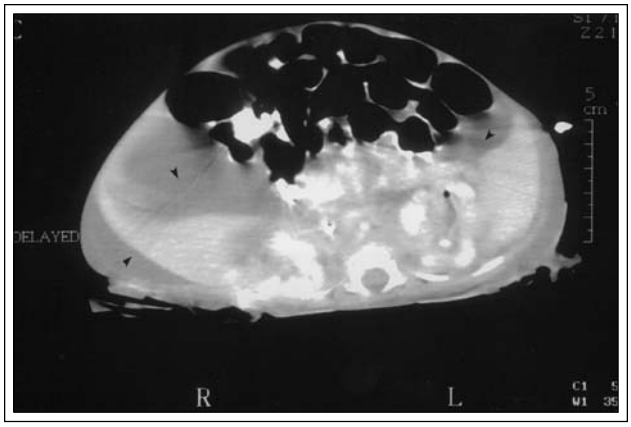


**Figure 1.** CT scan demonstrating extensive abdominal ascites (arrowhead).

catheterization was not feasible. It should be noted, however, that inserting a Foley catheter in a child with Prune Belly Syndrome might increase the risk of urinary tract infection in their dilated systems in instances where patients do not respond to bladder drainage, it is thought that the ureters can become mechanically obstructed due to their redundancy and tortuosity. Upper urinary tract diversion may be necessary in these cases.

In our patient urinary ascites resulted due to forniceal rupture despite having the bladder totally decompressed with an indwelling catheter. This event has not been previously reported in a child with Prune Belly Syndrome. Indeed, these patients are often considered to have dilated but non-obstructed systems. In conclusion, if there is clinical evidence of ongoing upper urinary tract obstruction following adequate bladder drainage in a child with Prune Belly syndrome, cutaneous pyelostomy may be a necessary intervention. □

References

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