Journal Pre-proof

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PII: S1477-5131(26)00085-9

DOI: https://doi.org/10.1016/j.jpurol.2026.105806

Reference: JPUROL 105806

To appear in: *Journal of Pediatric Urology*

Received Date: 8 October 2025

Revised Date: 2 January 2026

Accepted Date: 4 February 2026

Please cite this article as: Chu E, Press B, Weinstein C, Arlen AM, Smith EA, Kirsch AJ, Clinical Manifestations and Management of Prune-Belly Syndrome: A 20-Year Single Center Experience, *Journal of Pediatric Urology*, https://doi.org/10.1016/j.jpurol.2026.105806.

Clinical Manifestations and Management of Prune-Belly Syndrome: A 20-Year Single Center Experience

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Ethical Approval

This study was approved by the IRB at Children's Healthcare of Atlanta.

Abstract

Introduction:

Prune Belly Syndrome (PBS) is a rare congenital disorder characterized by abdominal wall musculature deficiency, cryptorchidism, and urinary tract abnormalities. Clinical presentation is highly variable, with multisystem involvement and a wide range of urologic and renal outcomes. Long-term data from contemporary pediatric cohorts remain limited.

Methods:

We conducted a retrospective review of pediatric patients diagnosed with PBS and followed at a tertiary pediatric urology center between 2002 and 2024. Demographic, clinical, surgical, and urodynamic data were extracted from electronic records. Renal function was assessed by estimated glomerular filtration rate and CKD staging. Comparative analyses were performed between subgroups using Student's t-test or Fisher's exact test, where appropriate. Multivariate logistic regression was performed to evaluate predictors of progression to CKD and renal transplant.

Results:

Sixty-four patients (61 males, 3 females) with a median follow-up of 12.6 years were included. Hydronephrosis (87.5%), vesicoureteral reflux (73.4%), and recurrent UTIs (76.6%) were common. Nearly all patients underwent surgical intervention, most frequently orchidopexy (83.6%), vesicostomy (23.4%), and appendicovesicostomy (35.9%). Abdominal wall reconstruction was performed in 45.3% of patients. CIC was utilized by 35.9%, typically via a continent catheterizable channel. Urodynamic studies demonstrated large, compliant bladders

with impaired emptying. CKD developed in 48.4% of patients, and 17.2% required renal transplantation, with nadir serum creatinine in infancy predictive of long-term outcomes.

Conclusion:

PBS requires lifelong, multidisciplinary care. Early identification of patients at risk for CKD, proactive bladder management, and coordinated reconstructive surgery are critical to optimizing outcomes. These results reinforce the importance of integrating functional and quality-of-life goals into care planning. Future prospective studies are needed to refine surgical timing, preserve renal function, and better characterize long-term quality-of-life outcomes.

Keywords: Prune Belly Syndrome, Chronic Kidney Disease, Bladder Dysfunction, Renal Transplantation, Abdominal Wall Reconstruction

Introduction

Prune Belly Syndrome (PBS) is a rare congenital multisystem disorder characterized by a triad of abdominal wall musculature deficiency, bilateral cryptorchidism, and urinary tract abnormalities, including megacystis, hydroureteronephrosis, and renal dysplasia [1-3]. PBS predominantly affects males, with a reported incidence of 3.6–3.8 per 100,000 live male births in the United States [2]. Females account for fewer than 5% of cases and typically present with abdominal wall and urinary tract anomalies in the absence of gonadal involvement [1].

The clinical presentation of PBS is heterogeneous, ranging from mild urinary tract dilation to severe renal dysplasia and pulmonary hypoplasia. Many patients have associated anomalies involving the gastrointestinal, musculoskeletal, and cardiopulmonary systems [4, 5].

Management often requires multiple surgical interventions throughout childhood, including bilateral orchidopexy, individualized urinary tract reconstruction, and abdominoplasty [6].

Initial postnatal care is focused on cardiopulmonary stabilization and establishing bladder drainage to prevent renal deterioration. Historically, instrumentation of the urinary tract such as clean intermittent catheterization (CIC) and videourodynamic studies (UDS) was discouraged due to concerns about high rates of urinary tract infections (UTIs) and associated complications [7]. However, the paradigm has shifted in recent years. The adoption of CIC and continuous antibiotic prophylaxis is now more common, reflecting efforts to reduce urinary stasis and mitigate infection risk while better managing lower urinary tract dysfunction [8].

Despite advances in care, long-term outcome data for PBS remain limited, particularly in contemporary cohorts managed at high-volume pediatric centers. This study aims to characterize the urologic, surgical, and renal outcomes of a large cohort of PBS patients followed over two decades at a tertiary referral center, with a focus on identifying factors associated with chronic kidney disease progression and reconstructive intervention.

Methods

This study was approved by the institutional review board (STUDY00002351). A retrospective chart review was performed on 92 pediatric patients diagnosed with PBS who were followed at our tertiary pediatric urology center between 2002 and 2024. Patients who were lost to follow-up before adolescence or who underwent renal transplantation at outside institutions with incomplete records were excluded. Missing date were managed using available-case analysis.

Patients were initially identified through diagnostic coding (ICD-9: 756.71 and ICD-10: Q64.1) and cross-referenced with the our prior institutional cohort to ensure completeness [8]. The following data was collected from the electronic medical record:

- Demographics: age at last follow-up, sex, race/ethnicity, duration of follow-up.
- Urologic history: presence of hydronephrosis (SFU), vesicoureteral reflux (VUR), febrile and afebrile UTIs, urinary incontinence, and nephrolithiasis.
- Bladder management: use of CIC, vesicostomy, or continent catheterizable channel
- Surgical history: procedures including orchidopexy, ureteral reimplantation, bladder augmentation, appendicovesicostomy, abdominal wall reconstruction, and renal transplantation
- UDS: bladder capacity, compliance, detrusor activity, VUR, and post-void residual (PVR)
- Renal function: nadir and longitudinal serum creatinine, estimated glomerular filtration rate (eGFR), and chronic kidney disease (CKD) staging per 2024 KDIGO guidelines. End-stage renal disease (ESRD) was defined as the need for dialysis or kidney transplantation.

Descriptive statistics were used to summarize patient characteristics and clinical outcomes. Comparative analyses between subgroups (e.g., transplanted vs. non-transplanted patients) were performed using Student's t-test or Fisher's exact test, where appropriate. Multivariate logistic regression was performed to evaluate predictors of progression to CKD and renal transplant. A two-sided p-value of <0.05 was considered statistically significant. Statistical analyses were conducted using SPSS.

Results

Patient Demographics

A total of 64 patients with clinically confirmed PBS met all inclusion criteria, including 61 males (95.3%) and 3 females (4.7%). Median age was 14.31 years with a median duration of clinical follow-up of 12.6 years. Race/ethnic distribution included 29 Caucasian (45.3%), 24 African American (37.5%), 7 Hispanic (10.9%), 1 Asian (1.6%), and 3 patients (4.7%) of mixed ethnicity. (Table 1)

Urologic Diagnoses

Hydronephrosis, was present in 56 patients (87.5%). A total of 47 patients (73.4%) had a history of vesicoureteral reflux (VUR), with 46 cases being bilateral and one unilateral. Nonfebrile urinary tract infections (UTIs) were observed in 49 patients (76.6%), and 30 (46.9%) had experienced at least one febrile UTI. Nephrolithiasis occurred in 3 patients (4.7%). Among the 64 patients over age 3 years, 14 (24.1%) experienced urinary incontinence beyond the expected developmental stage. Of these, 2 were continent during the day but reported nocturnal enuresis.

Comorbidities

Extra-genitourinary anomalies were present in in 30 patients (46.9%). These included cardiac anomalies in 8 (12.5%) and musculoskeletal abnormalities in 12 (18.8%), with scoliosis (n = 4) and clubfoot (n = 5) being most common. Gastrointestinal diagnoses were present in 37 patients (57.8%), of whom 23 (35.%) had chronic constipation. Pulmonary insufficiency was reported in 6 patients (9.4%), and 1 child (1.6%) required supplemental oxygen. Documented psychiatric conditions were present in 6 patients (9.4%), including attention-deficit/hyperactivity disorder,

autism spectrum disorder, anxiety, adjustment disorder, and pervasive developmental disorder. Among the 3 female patients, 2 had a cloacal anomaly.

Urologic Surgeries

All patients underwent at least one urologic or reconstructive surgical procedure. Orchidopexy was the most frequently performed operation, with 51 of 61 eligible males (83.6%) undergoing bilateral orchidopexy by age 3. An additional 5 males (8.2%) underwent unilateral orchidopexy. Of the 5 patients in our cohort who did not undergo orchiopexy, 2 are awaiting surgery and 3 did not have undescended testicles. Ureteral surgery was performed in 26 patients (40.6%), with 23 (35.9%) undergoing ureteral reimplantation. Thirty-three patients (51.6%) had bladder procedures: vesicostomy in 15 (23.4%), appendicovesicostomy (Mitrofanoff) in 23 (35.9%), reduction cystoplasty in 4 (6.2%). (Table 2)

Renal surgery was performed in 12 patients (18.8%), including 11 renal transplantations (17.2%). Fourteen patients (21.9%) underwent urethral surgery, most urethral dilations (n = 9, 14.1%). Other urethral procedures included posterior urethral valve ablation (n = 3), urethrostomy (n = 1), and urethrotomy (n = 1). Abdominal wall reconstruction (abdominoplasty) was performed in 29 patients (45.3%), with a mean age at surgery of 4.91 ± 2.9 years. Fifteen of these patients also underwent appendicovesicostomy, typically in a staged approach.

Bladder Management

Twenty-three patients (35.9%) performed CIC, including 19 via a continent catheterizable channel and 4 via the urethra. UDS were performed in 18 patients (28.1%). Indications for UDS

included preoperative planning in nine patients (50%), continued incontinence in 4 patients (22.2%), urinary retention in 3 patients (16.67%), and recurrent UTI in 1 patient (5.56%). Sixteen out of the 18 patients (88.9%) had a normal to supranormal bladder capacity with a mean 2.07 times capacity for estimated bladder capacity. Estimated bladder capacity was calculated as (age + 2) x 30 mL per Koff's formula [9]. Post-void residual was calculated as a percentage of the PVR measured on urodynamic study in relation to each child's estimated bladder capacity. Bladder compliance was considered normal or supranormal if it is greater than 10 mL/cm H2O [10]. All but two patients had good bladder compliance with a mean post-void residual of 67.9%, having the ability to only empty one-third of their bladder volume.

Chronic Kidney Disease and Transplantation

Chronic kidney disease (CKD) was documented in 31 patients (48.4%), and 11 patients (17.2%) underwent renal transplantation. The mean age at transplantation was 7.0 years. One patient experienced graft failure and subsequently underwent successful re-transplantation. The mean nadir serum creatinine in infancy among children with CKD was 0.77 mg/dL (range: 0.2–3.0 mg/dL), compared to 0.26 mg/dL (range: <0.2–0.5 mg/dL) in those without CKD ($p < 0.001$). There was no difference in presence of hydronephrosis between those with CKD and those without (90.3 vs 84.8%, $p = 0.399$). (Figure 1). When considering 13 patients who developed advanced CKD (Stages 4 and 5), many had congenital genitourinary anomalies. Five (38.5%) had urethral hypoplasia and two (13.3%) had posterior urethral valves. Two (13.3%) patients had ureterovesical junction obstruction and 5 (38.4%) had VUR. Among the transplant recipients, 1 (9.1%) had undergone vesicostomy in infancy and 8 (72.7%) were performing CIC via appendicovesicostomy at last follow-up. Mitrofanoff creation preceded transplantation in all

cases. Ten of 11 transplant recipients (90.9%) had a history of VUR and 9 (81.8%) had hydronephrosis. Six patients (54.5%) underwent abdominoplasty prior to transplantation; no complications related to abdominal wall reconstruction were reported in the peri-transplant period. On multivariate logistic regression, neither age, VUR, hydronephrosis, race, or nadir Cr was predictive of progression to CKD or progression to renal transplant.

Discussion

In this large cohort of 64 PBS patients followed over two decades, we observed a high burden of surgical intervention, significant rates of CKD, and diverse patterns of bladder dysfunction with respect to bladder emptying, catheterization requirements, continence, and need for reconstruction. Nearly half of the cohort developed CKD, with 17.2% requiring renal transplantation. These findings reinforce the complexity of PBS and the need for early, multidisciplinary intervention. PBS presents with significant phenotypic variability, complicating efforts to establish uniform treatment pathways. Multisystem involvement, including gastrointestinal, musculoskeletal, cardiopulmonary, and neuropsychiatric manifestations, further diversifies clinical presentation [4, 5]. In our cohort, the incidence of pulmonary insufficiency (9.4%) was markedly lower than previously reported rates approaching 58% [2, 4], likely due to our outpatient-based population. Critically ill neonates with life-limiting cardiopulmonary disease were likely underrepresented in our patient population.

Bladder dysfunction in PBS is typified by preserved compliance and capacity but impaired emptying due to poor detrusor contractility [7, 11]. In our cohort, urodynamic findings confirmed large-capacity, hypotonic bladders in patients with voiding dysfunction, aligning with prior

studies [8]. This functional impairment contributes to urinary stasis, predisposing patients to infections, bladder stones, and renal deterioration. CIC used in over one-third of our cohort, remains a mainstay for bladder management. Our findings support early CIC and urodynamic assessment as key components of renal preservation.

Although polyuria was not systematically assessed in this retrospective cohort, it may plausibly contribute to urinary symptoms and bladder management strategies in patients with PBS. High urine output related to congenital renal dysplasia and impaired concentrating ability may increase the risk of urinary leakage or nocturnal enuresis and, in some patients, necessitate more frequent CIC to maintain low bladder volumes and protect upper tract function. These factors should be considered when interpreting continence outcomes and catheterization burden in this population.

Bilateral cryptorchidism is nearly universal in male PBS patients and is presumed to result from mechanical obstruction caused by an enlarged urinary tract and underdeveloped abdominal musculature [4, 12]. Surgical approaches are selected based on testicular position and viability [13-15]. Orchidopexy is often the earliest surgical intervention and may be performed in conjunction with other reconstructive procedures, optimizing anesthetic efficiency. However, while combining procedures can be practical, orchidopexy should not be deferred beyond the AUA-recommended window solely to coordinate with other anticipated surgeries, as delays risk compromising outcomes.

All patients in our cohort underwent at least one urologic surgery, with orchidopexy, vesicostomy, and Mitrofanoff creation being most common. Vesicostomy was performed in

15.6% of our patients, consistent with our previous cohort [7], and remains a reliable, reversible option in neonates with obstructive uropathy or infection risk [4, 16]. For long-term management, continent catheterizable channels such as the Mitrofanoff appendicovesicostomy provide a discreet and effective means of CIC [17]. In our cohort, 28.1% of patients underwent Mitrofanoff creation, which a notably higher rate than earlier series, where PBS accounted for a minority of cases [18]. This may reflect evolving surgical preferences and increased emphasis on quality-of-life considerations. The Mitrofanoff has been shown to promote independence and self-esteem, particularly in adolescents, though complications such as stomal stenosis and leakage require ongoing surveillance [19, 20]. Preoperative urodynamic evaluation is essential for case selection and optimization of surgical outcomes [19]. Importantly, no major complications were noted in our cohort, supporting its safety and efficacy in PBS.

Renal preservation remains a central goal in PBS management, with approximately 40–50% of patients at risk of progressing to CKD or ESRD [15]. In our study, 48.4% developed CKD, and 17.2% underwent renal transplantation. Nadir serum creatinine in infancy was predictive of long-term renal function, consistent with previous reports [15], with higher values associated with eventual transplantation. Among transplant recipients, the majority had VUR and hydronephrosis, and many had already undergone urinary reconstruction. These findings suggest that early markers of obstruction and reflux may help identify patients at highest risk for progressive renal dysfunction. Detailed upper tract imaging with magnetic resonance urography may assist with these diagnoses. [21]

Abdominoplasty addresses one of the defining physical deformities of PBS and is typically offered in early childhood. [22] In our cohort, 45.3% underwent abdominal wall reconstruction at a mean age of 4.9 years. While historically performed for cosmetic reasons, evidence suggests that abdominoplasty may also improve voiding mechanics by enhancing intra-abdominal pressure [23]. Long-term outcomes have demonstrated durable cosmetic and functional benefits, with minimal morbidity [24].

This study is limited by its retrospective design and reliance on outpatient records, which excluded patients who died neonatally or were lost to follow-up. As such, our findings may underestimate the prevalence of severe comorbidities and early mortality. Urodynamic evaluation was not uniformly performed, as only 18 of 64 eligible patients underwent UDS. This does introduce possibility for selection bias, which may limit generalizability. Surgical decisions were individualized, limiting standardization across the cohort. The absence of statistically significant associations on multivariable analysis likely reflects limited statistical power related to the modest number of outcome events, rather than definitive absence of clinically meaningful effects. Nevertheless, this remains one of the largest single-center longitudinal studies of PBS and provides valuable insights into evolving management strategies. Overall, our findings emphasize the importance of early identification of high-risk patients, timely bladder management, and coordinated surgical planning to optimize long-term outcomes in PBS. Those requiring CIC may be at higher risk for ESRD necessitating close surveillance. Importantly, fertility potential in this population remains incompletely defined: while orchiopexy can preserve testicular position in most males, studies demonstrate high rates of impaired spermatogenesis and azoospermia, though assisted reproductive technologies have occasionally led to successful

conceptions. [25-27] Female fertility appears better preserved, with several reported cases of natural pregnancy, though abdominal wall and pelvic anomalies may complicate labor and delivery. [28-30] Looking ahead, we aim to expand longitudinal follow-up to address broader quality-of-life concerns which remain critical but underexplored domains in this patient population.

Conclusion

PBS is a rare and heterogeneous condition requiring multidisciplinary, lifelong care. Our longitudinal institutional experience highlights the high burden of surgical intervention in this population, particularly for cryptorchidism, urinary tract reconstruction, and abdominal wall repair. While individualized treatment remains essential, our findings support early identification of risk factors for renal deterioration, proactive bladder management, and the integration of reconstructive surgery into a coordinated care strategy. The frequent use of continent catheterizable channels and abdominal wall reconstruction in our cohort highlights evolving priorities toward functional outcomes and quality of life. These findings underscore the importance of multidisciplinary, longitudinal care for patients with PBS. Future prospective studies are needed to evaluate long-term quality-of-life outcomes, optimize timing of surgical interventions, and reduce the burden of renal deterioration in this vulnerable population.

Conflicts of Interest

The authors have no conflicts of interest to declare.

Acknowledgements

The authors have no acknowledgements to declare.

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Table 1 - Demographics and Comorbidities

Sex	**n**	**(%)**
Male	61	(95.3)
Female	3	(4.7)
Age (years)	**n**	**(IQR)**
Median age	14.3	(5.9 – 19.4)
Median age at last follow-up	12.7	(5.1 – 15.9)
Urological Diagnoses	**n**	**(%)**
VUR	47	(73.4)
Non-febrile UTI	49	(76.5)
Febrile UTI	30	(46.8)
Hydronephrosis	56	(87.5)
Other congenital malformations	**n**	**(%)**
Yes	30	(46.9)
Cardiac anomalies	8	(12.5)
Musculoskeletal anomalies	12	(18.8)
Spinal deformity	5	(7.8)
Club-foot	5	(7.8)
Gastrointestinal anomalies	37	(57.8)
Pulmonary insufficiency	6	(9.4)
Oxygen-dependent	1	(1.6)
No	34	(53.1)

Table 2 - Urologic and Reconstructive Surgeries

	N	(%)
Urethral surgery	**14**	**(21.8)**
Urethral dilation	9	(14.0)
Valve ablation	3	(4.6)
Urethrotomy	1	(1.5)
Bladder surgery	**33**	**(51.6)**
Vesicostomy	15	(23.4)
Appendicovesicostomy	23	(35.9)
Reduction Cystoplasty	4	(6.2)
Tumor resection	1	(1.5)
Casale procedure	1	(1.5)
Orchidopexy	**56**	**(91.8)**
Renal surgery	**12**	**(18.8)**
Nephrectomy	7	(10.9)
Transplant	11	(17.4)
Ureteral surgery	**26**	**(40.6)**
Reimplantation	23	(35.9)
Cutaneous Ureterostomy	4	(6.2)
Pyeloplasty	2	(3.1)

Figure 1 – (A) Nadir serum creatinine in infancy and (B) prevalence of hydronephrosis among patients with and without CKD. Error bars represent 95% confidence intervals.