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Detailed evaluation of the upper urinary tract in patients with prune belly syndrome using magnetic resonance urography

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Summary

Introduction
Magnetic resonance urography (MRU) has proven to be useful in the setting of complex urologic anatomy. Prune belly syndrome (PBS) patients are known to have malformed and highly variable urinary tract anatomy due to significant dilation and renal dysplasia.

Objective
To further characterize the renal and ureteral anatomy and renal function in patients with PBS via MRU.

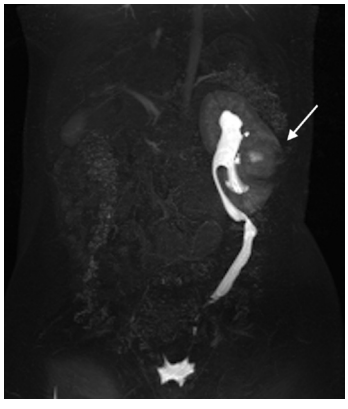
Study design
Children with PBS undergoing MRU (2006–2011) were identified. Studies were performed to evaluate severe hydronephrosis in all patients. Demographics, previous imaging, and MRU findings were collected. A single radiologist reviewed all studies.

Results
MRU was performed on 13 boys, with a median age of 29.3 months (IQR 6–97). Two patients underwent >1 study for ureteropelvic junction obstruction (UPJ obstruction) and calyceal diverticulum with a solitary kidney, respectively. Hydroureteronephrosis (HUN) was identified in 12 boys (92%), while one (8%) did not have ureteral dilation. All patients demonstrated morphologic abnormalities beyond HUN as follows: five (38%) renal dysplasia; five (38%) scarring; four (31%) calyceal diverticula; and three (23%) thickened bladder. The median renal transit time (RTT) was 6 min (IQR 3.5–10.5), and >8 min (range 8.5–35) in six patients; one patient was ultimately diagnosed with obstruction. The mean serum creatinine was 0.5 ± 0.3 mg/dl. This summary figure is a coronal excretory phase T1 MRU image demonstrating absence of well-defined calyces and a 5-cm calyceal diverticulum (white arrow).

Discussion
This study reports significant anatomic and functional findings on MRU that were not readily apparent when using standard imaging for children with PBS. The high-resolution images and functional data obtained with MRU allowed for visualization of calyceal diverticula and abnormal renal pelvic anatomy not previously described in PBS. In addition, renal dysplasia could be identified with MRU, which is badly characterized in the PBS population outside of renal biopsy studies.

Potential limitations of the study included its nature as a small retrospective case series, which limited the ability to compare imaging modalities. Imaging modalities were based on individual clinical needs; therefore, comparison with diuretic renal scintigraphy was limited.

Conclusion
MRU provided anatomic and functional details of the urinary tract in children with PBS that allowed for characterization of new renal anatomic abnormalities, including the incidence of calyceal diverticula and renal dysplasia, which have not been previously described. While renal scarring, dysplasia and calyceal diverticula were easily discerned on MRU in ten patients, their clinical significance requires longer follow-up in a larger patient population.



Figure

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Introduction

Prune belly syndrome (PBS) involves a classic triad of bilateral undescended testes, abdominal wall laxity, and urinary tract abnormalities. There is often severe dilation of the urinary tract that is non-obstructive [1]. Current evaluation involves ultrasound (US) and diuretic renal scintigraphy (DRS), most commonly a mercaptoacetyltriglycine (MAG-3) scan. However, these imaging modalities may be limited due to low resolution and inability to characterize obstruction in dilated and poorly functioning systems [2]. Detection of obstruction and the need for operative management can be challenging as the urinary tract may be massively dilated, leading to prolonged drainage of contrast material in otherwise unobstructed low-pressure systems.

In the setting of complex urinary tract anatomy, magnetic resonance urography (MRU) has proven to be useful as it allows real-time integration of renal anatomy and function [2,3]. Diagnosis of urinary tract obstruction on MRU is highly accurate when based on peak renal enhancement and washout [4,5]. DMSA is effective in identifying renal scars by presence of photopenic defects, with similar accuracy on MRU [2]. It was felt that the combination of anatomic detailed and functional assessment in the same study would allow for identification of previously unreported uro-anatomic abnormalities in this population. This study sought to further characterize the renal and ureteral anatomy and renal function of patients with PBS through the use of MRU.

Methods

Upon receiving institutional review board approval, the medical charts of 13 patients with a diagnosis of PBS who underwent MRU between May 2006 and February 2015 were reviewed. Patients were not included in this report if they did not have PBS and did not undergo an MRU during the study interval. Studies were performed to evaluate severe hydronephrosis in all patients, in an attempt to differentiate obstruction from dilatation in challenging cases. The MRU was performed according to the institution's previously described protocols, including use of sedation [6,7]. Ultrasound and DRS are the imaging modalities of choice to follow PBS patients and were ordered in these children before and after MRU, based on clinical indications and individual physician practice patterns.

Magnetic resonance urography techniques for image analysis and interpretation, evaluation of differential renal function, and renal transit time (RTT) have been extensively detailed [8,9]. Briefly, patients were sedated with propofol under the supervision of a dedicated physician trained in pediatric sedation. A Foley catheter was inserted into the bladder for all patients during MRU. Standard 2-dimensional T1 and T2 weighted anatomical images were acquired. Intravenous furosemide and gadolinium-based contrast (Magnevist®) were used to evaluate renal function and drainage. Drainage was determined by RTT.

During DRS studies, obstruction was evaluated by the time (minutes) to half contrast radiotracer drainage ($T^1/_2$) from the renal pelvis after the administration of furosemide. Similarly, during MRU, obstruction was evaluated as the time to contrast drainage from the renal cortex to the ureter just below the lower pole of the kidney. An RTT on MRU <4 min excludes obstruction, 4—8 min is equivocal, and >8 min suggests obstruction [10]. A final diagnosis of obstruction on MRU takes into account RTT, and renal parenchymal and collecting system morphology.

Calyceal diverticula were defined as a cavity within the renal parenchyma that filled passively with contrast during the excretory phase of imaging. Renal dysplasia on MRU was defined as disorganized renal architecture, loss of corticomedullary differentiation, and T2 hyperintensity [11]. Renal scarring was defined as an area of decreased perfusion located in the renal cortex, or transmurally if associated with a dilated calyx and adjacent renal contour defect. Relevant clinical data were taken from patient charts, including: demographics, clinical information, creatinine, and associated relevant imaging prior to MRU. The reported creatinine levels were obtained within 30 days prior to MRU. A single radiologist (JDGS) reviewed all imaging studies.

Results

Magnetic resonance urography was performed in 13 boys at a median age of 29.3 months (IQR 6—97 months). Two patients underwent more than one study for the evaluation of UPJ obstruction and calyceal diverticulum in the setting of a solitary kidney. The median clinical follow-up from the time of MRU was 82.1 months (50.3—61.3). One patient had a concomitant diagnosis of pulmonary hypoplasia and two others ultimately progressed to end stage renal disease.

Renal morphology

Hydronephrosis was present in all boys and hydroureteronephrosis was present in 12 (92%). Unilateral UPJ obstruction was diagnosed by MRU in a single patient (8%) with a solitary kidney and no hydroureter. A pyeloplasty was performed after follow-up MRU revealed progressive obstruction. On MRU, all patients demonstrated additional morphologic renal abnormalities, including: five (38%) with dysplasia; five (38%) with scarring; and four (31%) with calyceal diverticula. Four patients had a mean 1.5 ± 1.0 calyceal diverticula, measuring a median of 11 mm (10.3—12.5). Calyceal diverticula were not found on any pre-MRU imaging studies and ultimately did not require surgical treatment. Infundibular stenosis was not detected in any patients. Additionally, a thickened bladder wall was identified in three MRU studies (23%). The individual findings on MRU are detailed in Table 1, and representative images of renal parenchymal morphology can be found in Fig. 1.

Renal calyceal morphology was markedly abnormal in all patients. Calyces were blunted, few in number with an abnormal branching pattern, or absent with only a renal pelvis visible on imaging. Fig. 2 highlights these findings.

Renal dysplasia was noted in five (35%) patients and scarring in an additional five (38%) patients. Both scarring