Percutaneous nephrolithotomy in an 8-week-old infant

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We report successful percutaneous nephrolithotomy (PCNL) in an 8-week-old, 4.12 kg infant with a combined stone burden of > 2 cm in a solitary kidney. The patient was born with thoracolumbar myelomeningocele and had developed recurrent urinary tract infections. Her size precluded retrograde intrarenal surgery and shockwave lithotripsy would be unlikely to clear the stone burden. Stone analysis revealed hydroxyapatite and carbonate apatite stones, and metabolic work up revealed hypercalciuria for which chlorothiazide was started. To our knowledge, this is the youngest patient to undergo PCNL reported in the literature.

Key Words: percutaneous nephrolithotomy, infant, myelomeningocele, nephrolithiasis, pediatric, urology

Introduction

The cumulative incidence of nephrolithiasis during childhood has nearly doubled over the last 15 years.[1] Previous studies have shown 1%5-28% of children with nephrolithiasis require surgical treatment.[2] The need for surgery may be higher in the pediatric patients with neurogenic bladder. Stephany et al reported 60% of patients with myelomeningocele (MMC) required surgical intervention for stones.[3] Percutaneous nephrolithotomy (PCNL) have been shown to be an effective method for stone clearance in infants, and its safety in children as young as 5 months of age has been previously reported.[4] Here, we present a case demonstrating PCNL can be safely performed in a 8-week-old infant.

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Patient presentation

A female infant born full-term with T12-L1 MMC resulting in severe kyphoscoliosis underwent postnatal MMC closure on day of life 6. A routine renal bladder ultrasound (RBUS), performed 48 hours post repair per standard of care for patients with MMC, showed right renal agenesis and a left kidney with mild pelviectasis. She was started on clean intermittent catheterization (CIC) every 4 hours with overnight drainage following our institute's clinical pathway as she had elevated post-void residual (> 1.5 estimated bladder capacity (EBC)). She was discharged to home at 2 weeks of life on amoxicillin prophylaxis and CIC. She was readmitted at 5 weeks of age with her first febrile urinary tract infection (UTI). Her urine culture grew *Klebsiella pneumoniae* for which she was treated with 1 week course of cephalexin. Two weeks later, she developed an afebrile *Pseudomonas* UTI. Her urine pH at the time of the *Pseudomonas* infection was 7.0. An RBUS demonstrated multiple new non-obstructing stones in her solitary kidney with total stone burden > 2 cm, Figure 1.

After admission for intravenous antibiotic (ceftriaxone and subsequent cefepime after urine culture resulted) and clinical monitoring, a PCNL was performed when she was 8 weeks of age. PCNL was chosen because it

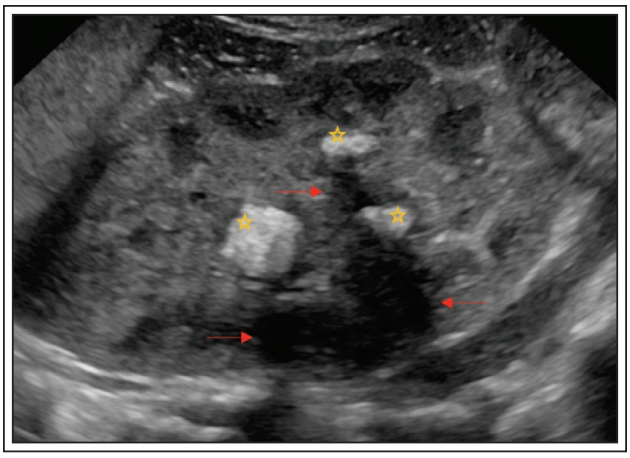

Figure 1. Renal ultrasound obtained at five weeks of life demonstrating multiple new large non-obstructing stones in her solitary kidney. (Star indicating hyperechoic foci with red arrows pointing out posterior acoustic shadowing).

had the highest probability of stone clearance. Her size precluded retrograde intrarenal surgery. Shockwave lithotripsy (SWL) would be unlikely to clear the stone and this modality has been associated with an increased lifetime risk of hypertension. Initial cystoscopy was limited by her lower limb contractures and scoliosis, and the left ureteral orifice was not able to be identified. After placing the patient prone, a mid-pole calyx was accessed with a 5Fr micropuncture set under ultrasound guidance. Once the collecting system was accessed, a nephrostogram was performed using a second generation ultrasound contrast agent, Figure 2. Using Amplatz dilators, the tract was dilated under fluoroscopic guidance from 6Fr to 10Fr enabling placement of a 8/10Fr dual-lumen catheter and safety

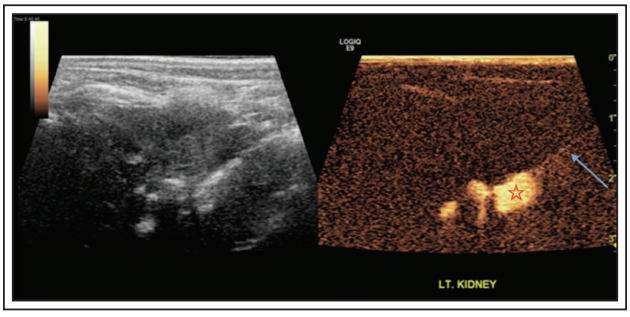

Figure 2. Percutaneous nephrostogram utilizing a second generation ultrasound contrast agent defining the collecting system and confirming needle access into the collecting system. (Star indicating interpolar minor calyx, arrow indicating the access needle).

wire across the ureteropelvic junction (UPJ) and into the bladder, Figure 3. Subsequently, the tract was dilated to 12Fr and a 14Fr ureteral access sheath was advanced under fluoroscopic guidance. An 8Fr Storz FlexEx-C flexible ureteroscope was introduced through the sheath. The UPJ appeared funnel-like but open. Seven stones were found in the interpolar calyx, the pelvis, and lower pole. The stones were extracted using the 1.9Fr nitinol basket without the need for lithotripsy. At the conclusion of the procedure, an 8Fr Malecot catheter was placed in the renal pelvis as a nephrostomy tube.

Her stone analysis returned calcium phosphate, comprised of hydroxy-apatite and carbonate apatite. Her serum creatinine was 0.2 mg/dL, potassium 5.1 mmol/L, and CO_2 23 mmol/L. Her subsequent 24 hour urine collection demonstrated polyuria (105 mL/kg/day), pH of 7.0, and hypercalciuria with a calcium to creatinine ratio of 650000 mg/kg. Chlorothiazide 10 mg/kg twice daily was initiated. RBUS performed at 2 months and 4 months postoperatively showed no stone recurrence. However, stones were visualized on surveillance ultrasound 8 months after surgery (2 mm calculi in the lower and interpolar calyces, and a 6 mm stone in the lower pole calyx). After counseling, the family chose close surveillance. Her videourodynamics demonstrated that she had low bladder pressure (3 cm) at 100% of EBC with an early grade 1 vesicoureteral reflux. She was continued on CIC every 4 hours with overnight drainage and daily 2 mg/kg/day trimethoprim/sulfamethoxazole prophylaxis.

Discussion

To the best of our knowledge, this report describes the youngest patient to have undergone PCNL. Current American Urological Association (AUA) guidelines on the treatment of nephrolithiasis in the pediatric

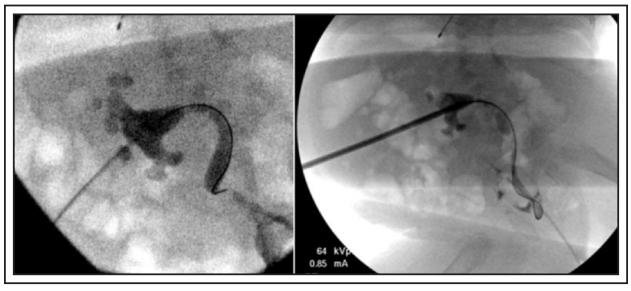

Figure 3. a) Intraoperative antegrade nephrostogram illustrating the renal pelvis and lower pole during the PCNL surgery and **b)** with 14Fr ureteral access sheath in place.

population recommend SWL or PCNL as first line therapy for patients with renal stones greater than 2 cm.[5] Due to the small size of her ureter and large stone burden, as well as her kyphoscoliosis causing positioning difficulty, retrograde intrarenal surgery would not have been possible. Furthermore, SWL was not a viable option given its lower efficacy and higher retreatment rates, particularly for larger stone burdens. Additionally, SWL to the kidney has been associated with a 40% increased hazard of hypertension,[6] which is of particular concern for a child with a solitary kidney and spina bifida. Although the stones were non-obstructive, surgical intervention was choses to eliminate the nidus for her recurrent urinary tract infections, and prevent ureteral obstruction and potential renal failure in a solitary kidney.

Since Woodside et al reported the first series of seven successful percutaneous treatments of stone in a pediatric population in 1985,[7] PCNL has been an option for treating children with large renal stone burdens. Guven et al compared 107 pediatric PCNLs, including six infants with an average age of 7 years versus 2666 adult PCNLs using their international database and showed no difference in overall complication rates and stone clearance.[8] Although pediatric PCNL is safe and effective, several technique and equipment modifications are necessary for the procedure to be successful. Due to the size and anatomy of our patient, we did not use a traditional nephroscope. Instead, we dilated the percutaneous tract to accommodate a 14Fr ureteral access sheath and a 8Fr flexible ureteroscope was used. Although the current AUA guidelines recommend a preoperative CT scan prior to PCNL,[5] we chose to avoid the risks of ionizing radiation in an 8-week-old baby as contrast-enhanced ultrasound allowed us to directly visualize the peri-renal anatomy during percutaneous access.

Our patient had multiple risk factors that contributed to her significant stone burden at an extremely young age, including infection (one with a urease-producing organism), urinary stasis, and hypercalciuria, to which the thoracic level spinal defect and associated immobility possibly contributed. The urine pH of 7 alludes to alkalinization possibly from the urease-producing bacteria in her urinary tract, facilitating calcium phosphate formation. Additionally, our patient has a high spinal cord defect which prior studies have shown is associated with kidney stone incidence.[3,9] Raj et al demonstrated the highest incidence of nephrolithiasis within the MMC population was found in the patient with thoracic defects(14.8%), followed by 4.5% lumbar spina bifida and 4.3% with lipomeningocel.[9] This patient's postoperative 24 hour urine collection revealed hypercalciuria, highlighting a metabolic component to stone development. Matlaga et al noted a shifting stone composition of their adult neurogenic population undergoing PCNL, in that the majority (62%) were metabolic (uric acid or calcium stone) versus traditional infection stones.[10] This highlights the importance of metabolic work up and stone prevention in patients at high risk within the MMC population. Lastly, her 24 hour urine also revealed mild polyuria that improved on subsequent 24 hour urine collection. It should be noted that her 24 hour urine collections had excessive variation in creatinine which raised the possibility of over-collection as a cause of her elevated urine volume.

Conclusion

The increasing incidence of pediatric nephrolithiasis necessitates treatment using modalities traditionally employed in adult stone disease, including PCNL. Our case presents a unique situation of an 8-week infant with a solitary kidney who developed stones that required surgical removal. We demonstrate that it is feasible to safely perform PCNL in an infant at 8 weeks of age, adapting the size of access and instrumentation to the size of the child and stone burden. □

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