Neonatal Obstructive Uropathy

11

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Core Messages

- Obstructive uropathy describes a condition where renal function has been adversely affected by impairment of flow of urine within the collecting system. This condition can be congenital, acquired, reversible, and irreversible.
- The majority of neonatal obstructive uropathy is diagnosed based upon findings on prenatal ultrasounds; however, the majority of fetuses with evidence of prenatal renal/ureteral dilation will not require any type of surgical intervention.
- Percutaneous nephrostomy tube is an effective means of decompressing the obstructed urinary tract in the clinically unstable neonate while awaiting definitive surgical reconstruction at a later time.

Case Vignette

A 25-day-old male presents to the emergency room with history of fever and repetitive emesis for the past 2 days. The mother states he has not been feeding well and his emesis occurs shortly after feedings. He was born full term via Cesarean section for breech presentation. His neonatal course has been uncomplicated. There is no family history of urologic problems.

The patient had a history of unilateral left sided prenatal hydronephrosis. This was detected at 20 weeks and was observed on every ultrasound obtained throughout the pregnancy. The hydronephrosis increased in its severity with an anterior posterior renal pelvic diameter of 22 mm at 34 weeks. There was no evidence of ureteral dilation or bladder distension. He was lost to follow-up postnatally until presenting to the emergency room with the above complaints.

His physical examination reveals a child that is responsive but not overly active. His temperature is 102 °F with a pulse of 170 bpm and blood pressure of 100/50. His abdomen reveals no any palpable mass in the right upper quadrant, but there is tenderness to palpation and fullness to the left upper quadrant. He is circumcised with an orthotopic urethral meatus and normal penis. Both testicles are descended. The lumbosacral region is normal to inspection and palpation.

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A.S. Chishti et al. (eds.), *Kidney and Urinary Tract Diseases in the Newborn*,
DOI 10.1007/978-3-642-39988-6_11, © Springer-Verlag Berlin Heidelberg 2014

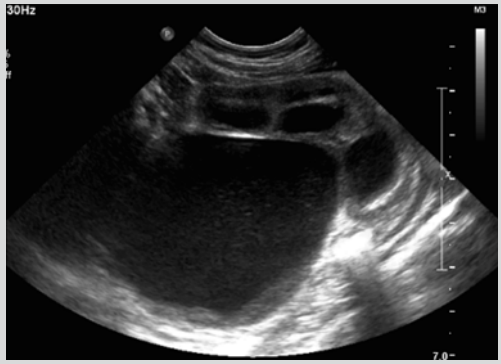

Fig. 11.1 Ultrasound appearance of severe left hydronephrosis and layering echogenic debris within the renal pelvis in a febrile neonate

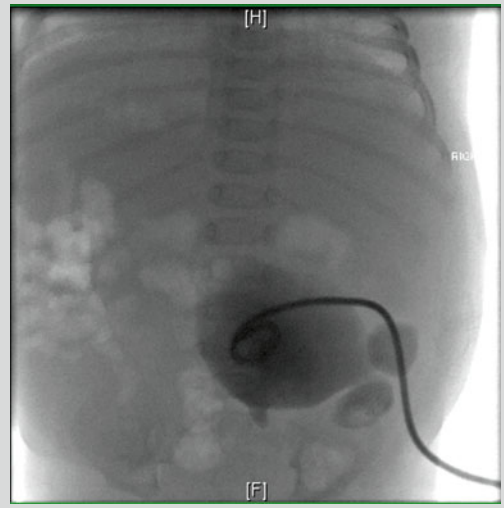

Fig. 11.2 Percutaneous nephrostogram performed at the time of urgent nephrostomy tube placement to treat obstructive pyelonephritis. This revealed high-grade UPJ obstruction that was treated with open pyeloplasty after successful treatment of infection

An abdominal radiograph shows normal air gas pattern and no signs of intestinal obstruction. There is medial deviation of the left upper quadrant abdominal contents. An abdominal ultrasound was performed given concern over possible pyloric stenosis and there was no evidence of pyloric hypertrophy. There was severe left hydronephrosis and layering echogenic debris within the renal pelvis (Fig. 11.1). There was no left ureteral dilation. The right kidney and ureter were normal. The bladder was not distended. A catheterized urine specimen is sent for urinalysis and is without abnormality. A CBC shows a leukocyte count of 20,000. Serum creatinine is 0.3 mg/ dL.

The diagnosis of left ureteropelvic junction (UPJ) obstruction with obstructive pyelonephritis is made. IV fluid resuscitation is begun and broad-spectrum antibiotics administered. A percutaneous nephrostomy is placed which reveals high-grade UPJ obstruction and turbid fluid is drained (Fig. 11.2). The patient defervesces within 24 h of the procedure and is discharged home with nephrostomy tube to complete a 2-week course of antibiotics. A diagnostic nephrostogram confirms the initial findings of stenosis at the UPJ. He underwent a dismembered pyeloplasty 4 weeks after initial presentation. His surgery was uncomplicated; he made a full recovery and has done well without any recurrent clinical problems.

11.1 Introduction

Obstructive uropathy describes a condition where renal function has been adversely affected by impairment of flow of urine within the collecting system. This condition can be congenital, acquired, reversible, and irreversible. Peters proposed a similar definition for congenital urinary obstruction in children of "a condition of impaired urinary drainage which, if uncorrected, will limit the ultimate functional potential of a developing kidney" [33]. Obstructive uropathy is an important cause of end-stage renal disease in newborns and children [39]. Identification of obstruction and relief of it can minimize its deleterious effect on renal function. This seems obvious, however current clinical criteria, in the form of laboratory or radiographic investigations, to recognize this category of patients is challenging.

Table 11.1 Society of fetal urology hydronephrosis grading system

SFU grade	Central renal complex	Renal parenchymal thickness
Grade 1	Slight splitting	Normal
Grade 2	Evident splitting, complex confined within renal border	Normal
Grade 3	Wide splitting pelvis dilated outside renal border, calices dilated	Normal
Grade 4	Further dilatation of pelvis and calices	Thin

Contemporary congenital cases of obstructive uropathy are diagnosed based upon the widespread use of prenatal ultrasonography. Ultrasound abnormalities of the urinary tract have reportedly been detected in up to 4.5 % of prenatal ultrasounds performed [11, 16]. The majority of these radiographic anomalies are hydronephrosis [29]. The severity of the hydronephrosis can vary, and the Society of Fetal Urology (SFU) has developed a grading system (Table 11.1) [27]. Additional obstructive uropathies diagnosed based upon prenatal ultrasounds, besides hydronephrosis secondary to ureteropelvic junction (UPJ) obstruction, can be megaureters as a result of ureterovesical junction (UVJ) obstruction, ectopic ureter, or ureterocele.

The majority of prenatally detected urinary tract abnormalities will not require any surgical intervention [28]. However, there can be cases of prenatally detected unilateral or bilateral severe urinary tract dilation that can cause concern in the neonatal period where any intervention aside from observation may be appropriate.

11.2 Diagnosis

11.2.1 History

The initial step in the evaluation of the neonate with potential obstructive uropathy will be obtaining a prenatal history. If obstructive uropathy is suspected as a result of prenatal ultrasonography, there are important data variables from the ultrasound to guide in diagnosis and subsequent management of the neonate. Specifically answers to the following questions can be critical to arriving at the correct diagnosis and subsequent appropriate treatment:

- What is degree of dilation?
  Prenatal transverse anteroposterior (AP) renal pelvic diameter has predictive value in determining the likelihood of postnatal surgery. Fetuses with an AP renal pelvic diameter of greater than 20 mm have a greater than 90 % of intervention, whereas fetuses with a diameter less than 10 mm have low probability [9].
- Are one or both kidneys hydronephrotic?
  Bilateral hydronephrosis can obviously be more challenging because there is no contralateral "normal" kidney to perform compensatory renal function when a unilateral obstruction is present. However, bilateral hydronephrosis is not an absolute indication for operative intervention. Close monitoring urine output and serum creatinine is essential in the scenario of severe bilateral hydronephrosis as prompt intervention may be necessary.
- Is there associated ureteral dilation?
  Ureteral dilation may signify a transient physiologic process as a result of diuresis, vesicoureteral reflux, or obstruction. Neonatal ureteral obstruction is most commonly the result of the presence of a ureterovesical junction obstruction or ureterocele. A ureteral duplication can be seen and if seen usually associated with hydroureteronephrosis of the upper pole moiety as a result of a distal ureteral obstructive process such as ectopic ureter or ureterocele. However, a ureterocele can be observed in single or duplex collecting systems.

  Knowledge of the sonographic appearance of the bladder is critical in interpreting the pathophysiology of the dilated ureter. For example, a distended bladder with accompanying ureteral dilation can be due to vesicoureteral reflux; however, obstruction can also be the cause. Conversely, an empty bladder does not rule out reflux to be the cause of the hydroureter unless the fetus voided shortly before image acquisition and the dilated ureter a result of refluxed urine from the bladder.

- What is the status of the bladder? Is it empty, thickened, and distended? Is an ureterocele identified?
  As stated in the previous section, knowledge of the bladder is crucial to correctly interpret the collecting system dilation. Bilateral hydroureteronephrosis with a distended and thickened bladder in a male fetus is highly compatible with a diagnosis of posterior urethral valves.
- What is the amniotic fluid volume? Is there oligohydramnios?
  Children with a history of oligohydramnios may have pulmonary difficulties as a result of poor lung development. Respiratory assistance is given as clinically indicated. A frequent etiology of oligohydramnios is renal dysplasia, and thus oligohydramnios can be a predictor of poor postnatal renal function in addition to poor lung function.

  A postnatal history of feeding intolerance may be related to severe hydronephrosis that can result in recurrent emesis from either direct compression of gastrointestinal tract or further distention of collecting system. In this uncommon neonatal scenario, a palpable mass is nearly universal. The neonate with an obstructed upper urinary tract can present with urinary tract infection manifested by fever, lethargy, and at times turbid fluid exiting the introitus of female representing infected urine.

11.2.2 Physical Examination

Inspection of the neonate will be the first portion of the physical examination. The infant with significant renal dysfunction as a result of an obstructive uropathy and accompanying oligohydramnios can result in labored breathing. Hydronephrosis can be appreciated as a palpable abdominal mass in severe cases as described in the previous section. An interlabial mass in the female may be a result of a prolapsed ureterocele (Fig. 11.3). Bladder outlet obstruction as a result of an ureterocele can result in bladder distension and a palpable suprapubic mass.

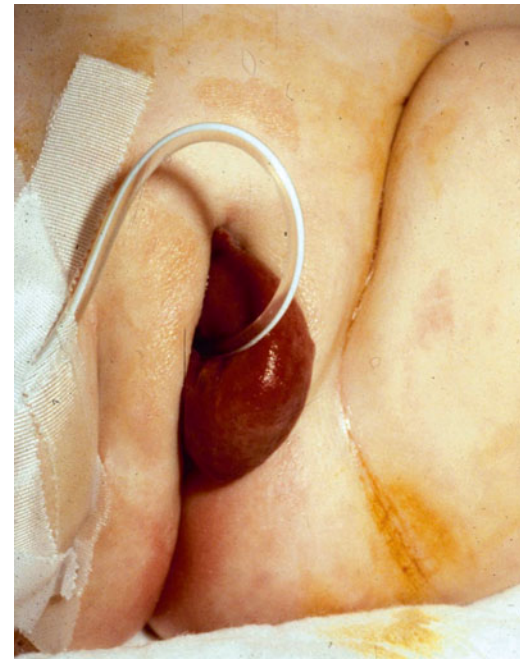

Fig. 11.3 Photograph of female neonate that presented with vomiting and interlabial mass secondary to vascular congested prolapsed ureterocele. The catheter is draining the bladder placed adjacent to the prolapsed ureterocele and reduction of the ureterocele with transurethral incision performed after adequate resuscitation

11.3 Imaging

A renal ultrasound is the preferred initial radiographic imaging test of choice to assess the urinary tract because of its widespread availability and lack of radiation. There is a physiologic degree of intravascular volume depletion in the immediate newborn period. Therefore, in certain cases of prenatally detected unilateral hydronephrosis, it is recommended to delay the initial postnatal ultrasound until at least 48 h to minimize false-negative findings or understaging which has been reported to occur in up to 44 % [44]. The severity of neonatal hydronephrosis and the appearance of the renal parenchyma are easily assessed using ultrasound and graded using the SFU grading system [27]. Differentiation of severe hydronephrosis from cystic dysplasia can be challenging at times, and additional imaging such as nuclear scan can clarify this. As outlined

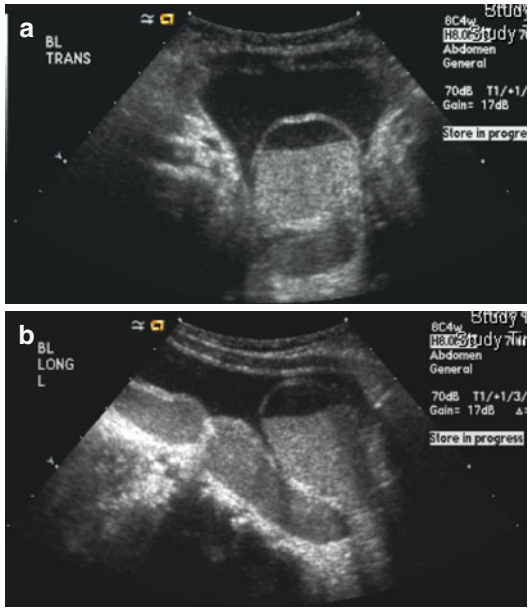

Fig. 11.4 (**a**) Transverse and (**b**) longitudinal ultrasound appearances of echogenic debris within an infected obstructed ureterocele

above it is essential to obtain images of the bladder as well as kidneys to determine if the bladder is distended, if a ureterocele is present, or if a dilated ureter is present. The neonate with an infected obstructed ureterocele can demonstrate hyperechoic debris on sonographic images of the bladder (Fig. 11.4). The ureterocele found with a duplex system is associated with the upper pole, and the renal parenchyma can have a cystic dysplastic appearance on ultrasound which may help determine if extirpative or reconstructive surgical approach is selected.

A voiding cystourethrogram (VCUG) is mandatory in cases of megaureter to exclude the presence of vesicoureteral reflux (VUR). However, there is the scenario of an obstructed, refluxing megaureter where both processes can be present and beneficial to know prior to treatment. Identification of coexistent VUR and excluding urethral anomalies is another reason to perform VCUG.

Nuclear imaging renogram provides valuable information with respect to differential function of the kidneys. A decision to intervene may be based upon decreased differential function of the hydronephrotic renal unit and concern that the hydronephrosis may be a result of significant obstruction. Diuretic renography is an extremely useful imaging investigation to discern whether a dilated collecting system is a result of an obstructed or nonobstructed process. Hydrational status of the patient, urethral catheterization, the timing of diuretic, dosage of diuretic, and data analysis can all affect the results or interpretation of the renogram. Therefore, the SFU has proposed the "well-tempered" renogram to standardize the procedure in an attempt to minimize these technical variables and create a uniform [8].

Due to their dependence on glomerular filtration, radionucleotide tracers have been deemed to be unreliable in the neonatal period due to immature renal function, a reduced glomerular filtration rate, and reduced responsiveness to a diuretic stimulus. Several investigators have assessed the reliability of a diuretic renogram in the neonatal period. Chung et al. found that in 17 neonates that underwent diuretic renal scans and followed clinically [7], there was no statistically significant difference between neonatal and follow-up differential function and response to diuretic stimulation revealed no statistically significant difference as the patients aged. Similarly the mean drainage times for normal non-hydronephrotic kidneys were similar when comparing those performed as neonates and at follow-up. The authors concluded to refute each of the criticisms against the use of diuretic renography to evaluate neonatal hydronephrosis and demonstrate its reliability in neonates. While maturation of the glomerular filtration takes several weeks, delaying the diuretic renogram, when clinically appropriate, may eliminate any confounding variables. However, the renogram should not be delayed when its results will impact the clinician's decision to intervene based on possibly diminished differential function that could be ameliorated with relief of obstruction.

Percutaneous antegrade pyeloureterography is helpful to define the anatomy, however mandates the use of sedation or general anesthetic to be safely performed. The length of stenotic segment can be accurately delineated prior to

embarking upon operative intervention with either retrograde ureteropyelography or antegrade nephrostogram.

11.4 Treatment

The hydronephrotic kidney or dilated ureter is a radiographic finding that must be interpreted to determine whether a surgical procedure is appropriate. These imaging results are not absolute indications for intervention. A simple classification scheme of collecting system dilation would be obstructing or nonobstructing. Unfortunately in the clinical setting, this classification scheme is not this dichotomous or simplistic. The "holy grail" of urology is identifying the neonate with asymptomatic urinary tract dilation who will benefit from intervention with respect to limiting, or improving, renal function as a result of correcting the obstruction from those patients that will not benefit.

Unilateral ureteral obstruction during early development in the experimental setting has been shown to result in impaired growth and development of the kidney [6, 35]. Experimentally, relief of the obstruction has been shown to attenuate, but unfortunately not reverse the observed decrease in number of nephrons in the affected renal unit [5]. Clinically a similar lack of significant improvement in differential renal function following pyeloplasty has also been shown to be true [26, 30].

11.4.1 Nonoperative Management of Hydronephrosis

The observation of similar preoperative and postoperative differential renal function has led many to advocate for an observational approach to the management of neonatal hydronephrosis as there appears to be limited "recoverable" function. Proponents of this surveillance strategy publish their experience in nonoperative treatment and have shown that approximately 75 % of neonates with history of prenatal hydronephrosis will not require immediate or any surgery [18, 42]. There is an essential requirement for regular follow-up when embarking upon an observational protocol. During this observational period, renal ultrasound and diuretic renograms are used to identify as early as possible of renal deterioration reflected as either progression of hydronephrosis or decreasing differential function, respectively. Prompt surgical intervention is necessary for these signs of deterioration to prevent progressive deterioration and potentially restore function.

11.4.2 Nonoperative Management of Hydroureteronephrosis

Similar principles apply to the management of antenatally detected dilated ureter as are applied to the management of hydronephrosis. Initial attempts at observation can be entertained when clinically appropriate. Several reports have shown that resolution of the hydroureteronephrosis and/or avoidance of surgery can be expected in up to 80 % of patients whose megaureters were identified prenatally [4, 31]. According to Lee et al. the risk of urinary tract infection during the first year of life is higher in cases of obstructive hydroureteronephrosis (47 %) compared to obstructive hydronephrosis (13 %) [25]. Periodic renal ultrasound and diuretic renograms are once again used to identify signs of renal deterioration reflected as either progression of hydronephrosis or decreasing differential function, with surgical intervention enacted swiftly.

The patient who has a megaureter associated with an ureterocele is also a candidate for a nonoperative approach [10, 13]. Ureteroceles with nonobstructed duplex systems have better preservation of renal function and a high rate of natural resolution of hydronephrosis and reflux. Ureteroceles associated with MCD or completely nonfunctioning upper pole moieties may never require surgical management.

11.4.3 Operative Management of Hydronephrosis Without Hydroureter

In the neonate with severe hydronephrosis secondary to UPJ obstruction and decreased differential renal function is a clinical situation that

warrants serious consideration of surgical correction. Pyeloplasty can be safely performed in the neonatal period; however, deferring the procedure may decrease the risk of general anesthesia as the child continues to mature from a cardiovascular and pulmonary perspective. Success rates and the incidence of complications have not been reported to be higher when the pyeloplasty has been performed in the neonatal period compared to when the child is older [19, 21]. Hanna commented that frequently encountered wide caliber of the neonatal ureter below the UPJ may contribute to a technically easier pyeloplasty to perform relative to the older child where the ureteral diameter is not as wide [14]. Surgical complications of urine leak, persistent/recurrent obstruction have been reported to be less than 5 % of patients undergoing open pyeloplasty in modern series [2, 38].

At times the differential function of the hydronephrotic obstructed kidney may reveal extremely poor function, 10 % or less. Management of the kidney with UPJ obstruction and associated poor function in the neonate is controversial and options include reconstruction or removal of the affected kidney. Reconstruction becomes a viable alternative because of reports of significant increase in differential renal function with relief of the obstruction [1, 12, 43]. Percutaneous nephrostomy tube placement is an easy and effective means to decompress the kidney and assessing the kidney's function devoid of obstruction while avoiding the risks of a potentially ineffective pyeloplasty. Gupta et al. described their experience of placing percutaneous nephrostomy in 20 patients who presented with UPJ obstruction and a split differential renal function of less than 10 % [12]. The nephrostomy tube remained in situ for at least 4 weeks and renography repeated 4 weeks after decompression. If no improvement in the split renal function had occurred, nephrectomy was performed; otherwise, pyeloplasty was performed. Twelve of 17 kidneys with unilateral UPJO improved after PCN drainage and underwent pyeloplasty with the split differential renal function increasing to 29 % from baseline where all kidneys were ≤10 %.

The scenario of infection and urinary tract obstruction is a potentially lethal one and merits emergent treatment. Decompression of the obstructed renal pelvis in the form of percutaneous nephrostomy tube is the most expeditious technique to achieve relief of the obstruction [23, 41]. Broad-spectrum intravenous antibiotics and aggressive intravenous resuscitation are essential components to proper treatment to minimize the systemic effects of this serious infection.

11.4.4 Operative Management of Hydroureteronephrosis

11.4.4.1 Operative Management of Primary Obstructed Megaureter in the Neonate

Once again similar principles apply to the clinical decision-making process on whether surgery is necessary for cases of obstructed megaureter. Accepted indications for immediate intervention include the presence of symptoms or infection, whereas poor drainage and decreased renal function can reasonably be initially observed. Operative treatment options are more numerous with an isolated distal obstructive process compared to obstruction at the level of the UPJ.

Definitive surgical treatment of primary obstructed megaureters typically involves ureteral reimplantation, which is technically challenging in the neonate because of the significant size discrepancy between the dilated distal ureter, small bladder template, and need for ureteral tailoring [34]. Cutaneous ureterostomy is an effective method of alleviating the distal obstruction and allowing the neonate to grow to adequate size while awaiting definitive reconstruction at a later time. End cutaneous ureterostomy has been used and can minimize the need for ureteral tailoring in subsequent ureteral reimplantation in approximately 75 % of cases [20, 36]. Loop cutaneous ureterostomy is another surgical technique performed through a Gibson incision that does not disturb the distal ureteral anatomy near the ureterovesical junction [32].

Surgical complications of ureterostomy include stomal stenosis and inadequate drainage. The incidence of stomal stenosis following end cutaneous ureterostomy has been reported to occur more frequently than loop cutaneous

ureterostomy because of the division of the ureter from the bladder with end ureterostomy. Kaefer and colleagues have proposed an alternative surgical treatment strategy that simultaneously avoids the problem of stomal stenosis while addressing the distal ureteral obstruction with the creation of an intentional refluxing ureteral reimplant as an internal diversion of the primary obstructed megaureter [17, 24]. The definitive non-refluxing ureteral reimplantation is deferred until after the child is older than 1 year and is maintained on daily antibiotic prophylaxis for high-grade reflux. Kaefer described this technique in 10 children who underwent surgery at 2 months of age with all patients demonstrating improved drainage postoperatively. Five patients have subsequently undergone uncomplicated ureteral reimplantation with the remainder awaiting reconstruction.

11.4.4.2 Operative Management of Primary Obstructed Ureterocele in the Neonate

Ureteroceles can be associated with either single or duplicated ureteral collecting system. The ureterocele that is obstructed and non-refluxing can adversely affect the renal function of the moiety it is associated with. Presenting symptoms of an infected ureterocele can be fever, lethargy, and even sepsis. Emergent decompression in the form of transurethral incision is preferable as this avoids the challenges of maintaining a percutaneous nephrostomy tube in the neonate. A low transverse incision achieves decompression while minimizing the risk of de novo vesicoureteral reflux [37, 40]. However, there may be technical challenges of transurethral instrumentation in the patient population with the smallest commercially available resectoscope being too large for the male neonatal urethra. In this circumstance percutaneous drainage of the obstructed renal moiety is always an alternative method of drainage in the septic patient to eliminate causing iatrogenic injury to the urethra.

The obstructed ureterocele associated with ureteral duplication can be unobstructed by performing ipsilateral ureteroureterostomy [3, 22]. The ureteroureterostomy can be performed either proximally or distally. The advantage of performing an ipsilateral ureteroureterostomy is avoiding operating on the small bladder of the neonate or infant while eliminating the need for ureteral tailoring and its attendant operative risks. The ability to treat bladder pathology in the older through the same incision is one advantage of a distal, or low, ureteroureterostomy compared to a proximal ureteroureterostomy. Surgical complications include urine leak, persistent obstruction of the hydronephrotic upper pole moiety, or de novo obstruction of the lower pole ureter. The success rate of this type of "upper tract" approach as a single procedure for the successful treatment of obstructed ureterocele has been reported to range from 100 to 4 % in one study [15]. The presence of high-grade reflux into one moiety or vesicoureteral reflux into more than one moiety, regardless of the grade of reflux, was strongly associated with the need for further surgery.

Conclusion

Neonatal hydronephrosis and hydroureter is diverse spectrum that poses significant diagnostic dilemma for the clinician. The overwhelming majority of these patients are asymptomatic with a partial obstruction. Differentiating between the children who benefit from aggressive surgical intervention from those patients that may experience spontaneous resolution and observation appropriate is inadequate based upon current diagnostic imaging modalities.

However, when the patient has been diagnosed with obstructive uropathy, there should be no delay in seeking relief of the obstructive process and minimize its deleterious effects on kidney function regardless of the patient's age. A variety of methods to decompress the hydronephrotic collecting system exist each with their advantages and disadvantages. Temporary urinary diversion may be indicated prior to pursuing more definitive surgical reconstruction based upon the clinical scenario.

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