Management of hydronephrosis: a comprehensive review in pediatric urology perspective

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Hydronephrosis, characterized by dilation of the renal pelvis and calyces due to urine flow obstruction, poses a significant clinical challenge. Although often asymptomatic and capable of spontaneous resolution, surgical intervention is necessary for specific scenarios such as febrile urinary tract infections, deteriorating hydronephrosis, or declining renal function. The efficacy of continuous antibiotic prophylaxis in preventing urinary tract infections remains controversial. Yet, it may benefit high-risk patients, emphasizing the importance of individualized patient selection, as surgical treatment methods for patients with hydronephrosis have become less invasive than in previous decades. However, long-term follow-up outcomes are lacking, necessitating further clarification. This review presents an overview of the etiology, natural progression, and modern management of hydronephrosis, encompassing advancements in minimally invasive procedures.

Keywords: Child; Hydronephrosis; Ureteral obstruction

Introduction

Congenital abnormalities of the kidney and urinary tract include a spectrum of malformations that can occur at the level of the kidney, ureters, bladder, and/or urethra. Hydronephrosis, often considered a marker of congenital abnormalities of the kidney and urinary tract, is defined as dilation of the collecting system in the upper urinary tract (i.e., the ureter, renal pelvis, major calyces, and minor calyces) and is the most frequently detected abnormality on prenatal ultrasonography (US), occurring in 1% to 5% of all pregnancies [1,2]. Consequently, it is a common reason for pediatric urology referrals before birth or early in infancy.

Hydronephrosis represents a wide spectrum of urological conditions ranging from mild anomalies such as transient dilation of the collecting system to more important anomalies such as high-grade congenital vesicoureteral reflux (VUR), ureteropelvic junction obstruction (UPJO), and primary obstructive megaureter (POM) (Table 1). Accordingly, the underlying etiology is well-known for determining outcomes.

Among the aforementioned causes, except for VUR, the two most common conditions that cause symptoms and require surgical treatment are UPJO and POM. In this review, we discuss the pathological etiology, natural history, use of prophylactic antibiotics, and surgical treatment of these two conditions to provide a comprehensive and practical overview for pediatricians from the pediatric urology perspective.

Etiology

Ureteropelvic junction obstruction

Congenital UPJO commonly arises from an aperistaltic ureter

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Table 1. Differential diagnosis of antenatal hydronephrosis

Transient hydronephrosis
Ureteropelvic junction obstruction
Vesicoureteral reflux
Ureterovesical junction obstruction (primary obstructive megaureter)
Posterior urethral valve/urethral atresia
Ureterocele
Ectopic ureter
Duplex system
Others: prune belly syndrome, tumors

segment, which hinders the formation of efficient peristaltic waves. This abnormality leads to the replacement of the spiral musculature with longitudinal muscle or fibrous tissue in that specific area. These causes are thought to result from inadequate recanalization in utero at 10 to 12 weeks of gestation. Additionally, conditions such as abnormal secretion of compounds like transforming growth factor-β, epidermal growth factor, cytokines, nitric oxide, and neuropeptide Y have been identified to be critical in developing UPJO [3]. The significance of interstitial Cajal cells in UPJO remains controversial. Studies examining the distributional variance of Cajal cells in the obstructed and unobstructed ureteropelvic junction have yielded conflicting results across various publications [4].

The involvement of crossing vessels in the development of the UPJO remains a contentious issue. Although these vessels are found in approximately 20% of the general population and in up to 38%–71% of UPJO cases [5,6] the exact relationship between crossing vessels and UPJO remains uncertain. Some cases suggest that crossing vessels pose a purely mechanical obstacle owing to the absence of histopathological changes [7,8]. In contrast, in other instances, crossing vessels trigger inflammation, fibrosis, and smooth muscle hypertrophy at the ureteropelvic junction, leading to obstruction. Not all UPJO cases are linked to silent crossing vessels. Instead, they may result from different tissue pathologies. Other extrinsic causes include congenital kidney abnormalities such as horseshoe kidneys, duplex kidneys, and fibroepithelial polyps [9,10].

Primary obstructive megaureter
POM is characterized by functional obstruction due to an aperistaltic segment near the bladder, hindering normal urine flow. Histological studies revealed elevated collagen levels (predominantly type I) in this segment, contributing to the disruption of communication between cells, ureteroarrhythmias, and

obstruction [11-14]. Other theories propose that factors such as muscle atrophy of the inner longitudinal layer, hypertrophy of the outer layer, and compressive circular muscles lead to obstruction [15,16]. Additionally, histologic evidence points to different structural abnormalities like fibrotic terminal ends or excessive muscle responsiveness causing contraction [17,18]. Another hypothesis proposes that obstruction reflects the developmental progression of the distal ureter from a single circular muscle layer to the child's dual-layer (circular and longitudinal) structure [19,20]. The dilated proximal ureteral segment is observed to contain altered connective tissue, contributing to fibrosis, dilation, and potential ureteroarrhythmias with impaired peristaltic wave transmission [19-21].

Natural history

Studies specifically examining the natural progression of isolated UPJO are limited, with a focus primarily on hydronephrosis in general, encompassing transient cases. Based on the current research, a significant percentage of cases of isolated hydronephrosis resolved, ranging between 50% and 70%, irrespective of the severity [22-25]. Time to resolution was assessed, with patients having a smaller anteroposterior diameter (<10 mm) showing complete resolution in a median of 5 months compared to those with diameters of 10–20 mm resolving in 11 months [26]. A recent prospective study reported a resolution rate of 82% during a mean follow-up of 24 months, with notable differences in resolution times based on the initial hydronephrosis grade: 98% at 3 years for the Society for Fetal Urology (SFU) grade I versus 57% for grade IV. Similarly, urinary tract dilation grades 1, 2, and 3 showed resolution rates of 90%, 81%, and 71%, respectively, at 3 years.

Primary obstructive megaureter
Prenatal US allows earlier detection of the megaureter, with many cases remaining symptom-free and resolving without surgery [27]. Spontaneous resolution typically occurs within the first 2 years of life but can extend to young adulthood [27,28]. In literature, spontaneous resolution rates vary between 34% and 88% [29-31], with approximately 24% of megaureter cases requiring surgery, especially those with larger ureteral diameters [27]. Factors like baseline ureteral dilation, washout pattern, and timing of presentation can influence the need for intervention or potential resolution, emphasizing the importance of long-term ultrasound monitoring until puberty [32].

Role of continuous prophylactic antibiotics

Hydronephrosis may increase the risk of urinary tract infections (UTI) in children, especially during the first 2 years of life [33]. However, the debate over continuous antibiotic prophylaxis (CAP) versus observation for antenatal hydronephrosis (ANH) remains contentious, with limited conclusive evidence from two randomized controlled trials (RCTs) [34]. The available studies contained incomplete data and outcomes. A systematic review assessed the literature from 1980 onwards, highlighting the challenges in drawing strong conclusions regarding the superiority of CAP over observation alone for children with ANH [35]. Although one RCT identified factors such as female sex, uncircumcised male sex, lack of CAP, high-grade hydronephrosis, hydroureteronephrosis, and VUR as predictors of UTI development, another RCT without detailed data suggested no clear benefit of CAP on UTI rates for patients with ANH and VUR [34]. Considering these results and risk factors for UTI, it is advisable to consider CAP specifically for this subgroup of children who have been identified as at high risk [35].

Indications for surgical intervention

The European Association of Urology guidelines recommend surgery as the treatment of choice in patients with symptomatic UPJO. Other indications for surgery included impairment of split renal function (<40%) and a >10% decrease in split renal function in subsequent studies. Poor drainage function after diuretic administration, increased anteroposterior diameter on US, and grade III or IV hydronephrosis, as defined by the SFU, can be considered relative indications for intervention [36].

In POM, the primary indications for intervention include febrile UTI, urolithiasis, and abdominal symptoms. Other relative surgical indications are considered similar to those of UPJO, such as ureteral diameter >10 mm with split renal function <40% on initial Tc-99m MAG3 scintigraphy, split renal function with delta >10% on subsequent Tc-99m MAG3 scintigraphy, or worsening of dilation on repeat ultrasounds (>14 mm) [36,37].

However, except when symptomatic, surgery is typically considered when multiple criteria, and not only a single parameter, are aligned. Considering factors such as bladder fullness and hydration status, it is crucial to confirm the deterioration of ureteral dilation or hydronephrosis using repeat US before deciding on surgery. In addition, reduced split renal function may be due to hydronephrosis but may also be related to kidney dysplasia combined with hydronephrosis. Therefore, surgical decisions should rely on a comprehensive assessment of various clinical and imaging factors rather than a single criterion.

In summary, symptomatic patients (such as UTI and flank pain) in both of UPJO and POM are absolute indications for surgery, and in addition, if there is a decrease in renal function more than 10% during follow-up, worsening hydronephrosis, or poor renal function from the initial examination, a referral to a urologist is required.

Surgical options for UPJO

Endoscopic intervention (endopyelotomy)

Endoscopic endopyelotomy, introduced by Ramsay et al. in 1984 [38], offers benefits such as shorter hospital stays and quicker recovery. Endopyelotomy is based on the principle of Davis intubated ureterotomy, where a full-thickness incision through a strictured segment of the ureter heals over a stent to a larger caliber within weeks [39]. This procedure involves a full-thickness incision on the lateral side to avoid crossing the vessel on the ureteral lumen using a cold or hot knife or holmium lasers, followed by ureteral stenting. However, the success rate of endopyelotomy is reportedly worse than that of open, laparoscopic, or robotic pyeloplasty. The available studies on antegrade endopyelotomy have shown success rates of 65% to 93% [40]. Therefore, to increase the success rate of the procedure, careful patient selection is warranted, and patients with a less severe grade of hydronephrosis, better renal function, and short stricture length may be good candidates for this procedure [41]. In the pediatric population, the median success rate was 71% (46%–100%) in the primary group and 75% (25%–100%) in the secondary group which failed in primary procedures, while complications were reported in 14% of both primary and secondary procedures [42]. Considering these results, careful patient selection is important for successful procedures, and endopyelotomy can be considered a secondary rather than a primary operation.

Pyeloplasty

Pyeloplasty is considered the gold standard for UPJO treatment and can be divided into dismembered and flap procedures. Dismembered pyeloplasty, also referred to as the Anderson-Hynes operation, is a versatile surgical technique characterized by complete disconnection of the ureter and removal of the affected segment. This procedure allows for the correction of issues

such as redundant pelvis and transposition of the UPJ in cases where crossing vessels obstruct urinary flow. Flap procedures such as Foley Y-V plasty present benefits such as reduced operative time and decreased risk of devascularization of the UPJ, making them suitable for addressing long ureteral strictures. Comparative studies investigating surgeries, such as Anderson-Hynes and Y-V plasty, have revealed variations in success rates and ease of surgery [43,44]. These findings underscore the importance of tailoring the choice of procedure to the specific characteristics of each case, ensuring the best possible outcomes for patients undergoing pyeloplasty.

Laparoscopic and robotic-assisted approaches

Laparoscopic treatment of UPJO, pioneered in 1993 by Schuessler et al. [45] and Kavoussi and Peters [46], has become the standard for UPJO management because of its superior outcomes compared to endourologic techniques. Laparoscopic pyeloplasty (LP) offers excellent success rates ranging from 85% to 100% and is suitable for various patient groups, including infants and older individuals [47,48]. Although LP requires advanced skills for intracorporeal suturing, its benefits include reduced pain, decreased blood loss, and improved aesthetics. Due to the larger working space and more familiar anatomy, retrocolic and transmesenteric transperitoneal routes are more frequently chosen. However, the choice between transperitoneal, anterior extraperitoneal, or retroperitoneal approaches can be made by considering factors such as operative time and anatomy familiarity. The success rates of LP through the retroperitoneal and transperitoneal approaches were not significantly different; however, the operative time for the retroperitoneal approach was longer. Novel approaches, such as mini-laparoscopy and one-trocar-assisted pyeloplasty, provide safe and effective alternatives to traditional LP [49,50].

Robotic systems enhance laparoscopy by using a three-dimensional magnified view, instrument mobility, and tremor elimination. However, they lack tactile feedback, have higher costs, and require longer procedure times than traditional laparoscopy, offering no significant clinical advantage to experienced laparoscopic surgeons [51]. Despite this, robotic pyeloplasty is likely to be favored when technology is available. Recent meta-analyses comparing robotic, laparoscopic, and open pyeloplasty in the pediatric population have shown that robotic pyeloplasty reduces analgesia requirements, estimated blood loss, and hospital stay compared with LP, with similar success and complication rates [52]. Recently, single-port robot-assisted

LP using the da Vinci SP system has been introduced, showing a success rate similar to that of conventional robotic systems with fewer incision sites, and increasing application is expected [53].

Surgical options for POM

Traditionally, ureteral reimplantation with or without ureteral tailoring using an open technique has been the mainstay of POM treatment. The objectives of surgical correction are the resection of the obstructive lesion and the narrowing of the ureter to achieve an adequate length-to-diameter ratio. This can be achieved by intravesical or extravesical techniques or by combining both. Although open surgery remains the gold standard for the treatment of POM with an excellent success rate [29,54], successful results of laparoscopic ureteral reimplantations have been reported with the rapid advancement of laparoscopic surgery [55-57]. Robotic surgery, similar to laparoscopy but with enhanced precision and instrument mobility, is increasingly utilized for complex procedures [58]. Overall, ureteral reimplantation demonstrated success rates of 90% to 96% in the treatment of primary non-refluxing megaureter [59].

However, challenges in younger patients due to ureter-bladder size disparities may lead to complications such as secondary obstruction, secondary VUR, or transient bladder dysfunction [60]. Considering these problems, in young patients aged <6–12 months, temporary diversion, such as cutaneous ureterostomy or endoscopic balloon dilation, may be considered as an alternative treatment to surgery [60].

Cutaneous ureterostomy is the preferred procedure for severe acute septic complications, particularly in young patients. It is a practical method for temporary urinary diversion in neonates and infants with uncontrolled hydronephrosis symptoms, including compromised renal function and/or uncontrolled UTI. Refluxing ureteral reimplantation can also be considered a viable option for temporary diversion [61]. These procedures ensure efficient renal drainage, allow for stepwise reconstruction 12 to 18 months after the initial operation, and help alleviate the inconvenience for the child and their family [61].

Endoscopic balloon dilation can be proposed as an initial step for patients aged <12 months with POM before potential reimplantation, with a success rate of 67% to 95% [62]. Initially suggested as an interim measure for patients aged <12 months before safe reimplantation [63], it was later proven successful in certain cases, resulting in the avoidance of open surgery [64]. If

endoscopic treatment is unsuccessful, open reimplantation remains an option. Postoperatively, complications ranging from 23% to 60% have been observed, largely comprising temporary hematuria, UTIs, and concerns related to stent movement or intolerance [65]. Although endoscopic balloon dilation is considered the primary treatment for symptomatic POM, further comparative studies are needed to evaluate its effectiveness in infants and its long-term impact owing to the complex nature of POM.

Conclusion

POM and UPJO share a similar pathophysiology but demonstrate slight variations in their natural progression. With advances in knowledge and the accumulated outcomes of surgically treated cases, it has become feasible to predict the natural progression of prenatally detected hydronephrosis. Based on this predictability and knowledge, surgical methods and the use of prophylactic antibiotics tend to lean toward a more conservative and less invasive approach than before. Although many patients with hydronephrosis experience spontaneous improvement, some may encounter symptomatic issues, such as UTIs. Therefore, through a comprehensive analysis of the current literature and emerging practices, we aimed to assist clinicians in selecting the most suitable management strategies and surgical interventions tailored to each patient's unique requirements, ultimately aiming to enhance outcomes and preserve renal function.

Conflicts of interest

No potential conflict of interest relevant to this article was reported.

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