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Current Role of PCNL in Pediatric Urolithiasis

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Abstract The incidence of pediatric stone disease has been increasing. Though there are geographical variations, there remains a common theme in that this is a high-risk population with regard to stone formation and recurrence. Consequently, it is important to keep the number of procedures performed to a minimum and to save the developing kidney from the deleterious effects of intervention. Of the number of available treatment options, Percutaneous Nephrolithotomy (PCNL) offers a number of distinct advantages albeit with its own set of concerns. In the present article, the current literature on PCNL in pediatric urolithiasis was reviewed by a MEDLINE/PubMed search that was limited to literature in the English language, with emphasis on the current state of art in PCNL. Within the last few years, there have been improvements in radiological techniques, particularly computed tomography (CT), with dedicated reconstructions and development of scoring systems leading to better preoperative planning. Also, there has been miniaturization of instruments, particularly smaller nephroscopes, innovative sheaths and the potential to use lasers as well as smaller pneumatic and ultrasound probes. The combination of these has led to various modifications, such as miniperc, microperc and ultra-mini perc techniques. These modifications have been shown to be associated with a decrease in morbidity and high clearance rates. In this article, we analyze the current role of PCNL and its modifications, in terms of the indications, techniques, results, and complications in management of pediatric urolithiasis.

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Introduction

The surgical management of pediatric urolithiasis has undergone significant changes over the last few years. Of the number of available treatment options—extra corporeal shockwave lithotripsy, retrograde intra-renal surgery, percutaneous nephrolithotomy (PCNL) and open /laparoscopic/ robotic surgery—PCNL offers a number of distinct advantages, albeit with its own set of concerns in the form of parenchymal damage and the associated effects on renal function, radiation exposure, and the risk of bleeding, hypothermia and sepsis. The current literature on PCNL in pediatric urolithiasis was reviewed by a MEDLINE/PubMed search. The search was limited to literature in the English language, with due emphasis on the current state of art in PCNL. The publications considered were from peer-reviewed journals. We have used in our search the following keywords: urolithiasis, nephrolithiasis, pediatric, children and percutaneous nephrolithotomy.

Epidemiology and Clinical Presentation

Children represent 2–4.3 % of the total population of stone formers [1, 2]. The incidence of urolithiasis in children has increased globally over the last few decades. The pattern of stone disease has also changed, with an increase in kidney stones secondary to calcium oxalate or calcium phosphate, as well as marked regional variations in the stone prevalence [3]. Pediatric patients with urinary stones have a high risk of recurrence; therefore standard diagnostic procedures for high-risk patients apply to all children with urolithiasis [4]. Stone disease in this patient population is known to have association with urinary infection and anatomic and metabolic abnormalities. It is likely that all these play an important role in causing recurrence as well. Thus, it becomes prudent to have complete clearance, along with eradication of urinary infection and appropriate correction of any underlying metabolic or anatomical abnormalities. Karabacak et al. [5] have compared metabolic risk factors in children and adults having stone disease. Whereas hypercalciuria was the most common metabolic risk factor in the adult group, hypocitraturia was the most common metabolic abnormality in the pediatric population, found in 57.7 % of patients. Hypercalciuria was present in 27 % of the children evaluated. Recently, two studies [6, 7] assessed the association of body mass index (BMI) in the pediatric population and stone formation. Surprisingly, in both the studies, higher BMI was not associated with urolithiasis in children. This is in contrast to that seen in adult obese patients, where the nephrolithiasis development rate is significantly higher than in the general population.

As far as the gender distribution is concerned, there are differing reports, with some stating the male to female ratio of 1.2-4:1 [8] and others quoting almost equal gender distribution [9].

The clinical presentation of urinary stones in children is different from adults [10, 11]. Flank pain is the predominant symptom in adults, while abdominal pain (55–70 %) is the most common presenting symptom in children, followed by gross hematuria (14–33 %) and urinary tract infection (UTI) (8–46 %).

Imaging

Because of the above presenting clinical features, these patients frequently undergo ultrasound study [12, 13]. Though very effective in identifying renal stones including the radiolucent ones, ureteral evaluation is suboptimal. Nevertheless, for accurate treatment planning, digital radiography is required. The presence of a large amount of bowel gas in children and infants makes plain X-ray KUB less reliable for the diagnosis of urolithiasis in pediatric population, as shown by Diament et al. [14]. However, non-contrast helical computed tomography (CT) remains the most sensitive test for the detection of urolithiasis, with a sensitivity and specificity of 97 % and 100 %, respectively [15, 16]. CT, along with its urography, is of paramount importance in treatment planning, as it allows three-dimensional (3D) visualization of the urinary system, relationship to surrounding structures and identification of renal lesions and blood vessels [17, 18]. Recently, a number of morphometric or scoring systems have been designed, so as to aid in better planning, assessing the risk of perioperative complications and predicting the stone free rates following PCNL [19–22]. However, radiation exposure during CT always remains a concern and may exceed that of intravenous urography (IVU). This is of utmost importance in this specific patient population, which is at high risk of developing recurrent stones in future. The principle of ALARA (as low as reasonable achievable) should always be observed while performing diagnostic study in children [4]. So far as IVU is concerned, though rarely used in developed countries, it still continues to be used in many centers of the developing world.

Procedure Planning

As with any surgical procedure, meticulous planning is required before embarking on the procedure. The patient and the parents must be fully informed about the likelihood of success and complications. The latter comprise of bleeding (early and delayed), which may or may not require transfusion or angioablation; pneumothorax, hemothorax, urinothorax, incomplete clearance, sepsis and injuries to organs adjacent to each respective kidney. All the pertinent investigations, specifically the urography films, must be thoroughly reviewed. Every attempt should be made to treat a urinary tract infection and /or minimize bacteriuria prior to the procedure. Broad-spectrum intravenous antibiotics should be given at the time of the procedure.

Operative Considerations

Pediatric PCNL is performed under general anesthesia. After induction of anesthesia with the patient in the lithotomy position, a retrograde pyelogram is performed to outline the collecting system, and a 5-Fr open-ended ureteral catheter is left in situ to opacify the collecting system during percutaneous access. The patient is then repositioned in the prone position with the torso elevated at 30 degrees from the table surface with towel rolls acting as bolsters to support the chest and the lower abdomen. Adequate padding of pressure points should be done to prevent pressure induced injuries and neuropraxias. Circumstances that require special consideration involve children with congenital anomalies such as spina bifida, spinal cord injuries and implants or prosthesis in spine following a previous surgical intervention. Positioning can be a challenge under these circumstances, but needs special and extra padding. Nevertheless, renal anatomy can be altered as well, with an increase in the incidence of complications [23].

Access to the Collecting System

The success and treatment outcomes of the surgery are very well known as being highly dependent on the precision and accuracy of the puncture step (because it must reach the calculi with a precise and direct path), making this step the most challenging task for the urologist to master [17]. Hence, the most important step in the entire procedure is the correct access to the collecting system, which encompasses puncture and tract dilatation. As mentioned, a well-obtained urography study at the outset of the procedure and a retrograde pyelography help in achieving these goals. The number and sites of puncture, infundibular width, length and angle of entry into the pelvis can be discerned in this manner, and also guides as to the size of amplatz sheath to be used.

Ultrasonography Guided Puncture

Osman et al. [24] suggested that PCNL punctures could be safely carried out under ultrasound guidance. The advantages of this are avoidance of radiation, localization of even radiolucent stones, a shortest straight tract to the desired calyx and avoidance of visceral injury. The ultrasound probe can be with puncture guide or without puncture guide [25]. The scan starts posteriorly and proceeds anteriorly; the first calyx to be seen is the posterior calyx, which is the calyx of choice to gain access. The needle should traverse the electronic dotted line to achieve this goal. The relevant points to achieve this are to jiggle the needle in the subcutaneous plane, use a echo tip needle (Cook Urological Inc. Spencer IN), or keep the serrated margin of the needle facing the crystals of the probe [26••].

Fluoroscopic Guided Puncture

In pediatric patients, the technique remains the same as in adults, be it the bull's eye or the triangulation. However, the urologist should take into consideration the short distance between the skin and the kidney. It is worthy enough to have in mind that the depth the needle has to negotiate before it punctures the calyx of interest. The rest of the standard PCNL technique follows suit.

As to the choice of ultrasonographic or fluoroscopic puncture, the authors believe that it is institutional and urologist preference, so long as accurate puncture can be achieved.

Apart from the standard PCNL, which has been in vogue for years, there are *modifications* that have been introduced in recent years, whereby the efforts have been to decrease the tract size and the associated morbidity. These are the "mini", "micro" and "ultra mini" PERC techniques.

"Miniperc" (Mini PCNL) and "Microperc" (Micro-PCNL)

Traditionally, PCNL required a 30 Fr nephrostomy sheath for renal access. With the development of smaller sheaths, it was found that Mini-PCNL or "Miniperc" (tract size ≤20 F) could be performed with minimal damage to renal parenchyma, thereby reducing the procedure related morbidity without diminishing its therapeutic efficacy [27, 28]. Micro-PCNL or "microperc" is a recently described minimally invasive PCNL technique, which is performed using a 16 G microperc needle [29] (Fig. 1). A three-way connector is attached to the latter, which admits a saline irrigation tube, a 272 μm laser fiber and a 0.9 mm flexible microperc telescope (Fig. 2).

Modifications

Because of the risk of bending and parenchymal injury arising out of manipulation, recently an 8 F metallic "mini-microperc" sheath has been developed (Fig. 3). This allows utilization of the same three-way connector with accessories as in standard microperc. In addition, this also lends the ability to use a 1.6 mm ultrasonic lithotripter, for fragmenting and sucking out the fragments. Penbegul et al. described the use of a 14 G (6.6 Fr) angiocath, which can be used as an amplatz sheath in pre-school children [30].

Current role of Microperc

Microperc is still in its early years of application. It is currently used to manage single renal calculus or multiple renal calculi, which can be accessed with a single puncture and cumulative diameter of less than 1.5 cm in diameter. Recent series, however, have also seen a broader application of the technique to treat even the larger and complex stones with good clearance rates [31]. It can be safely used in the pediatric age group.

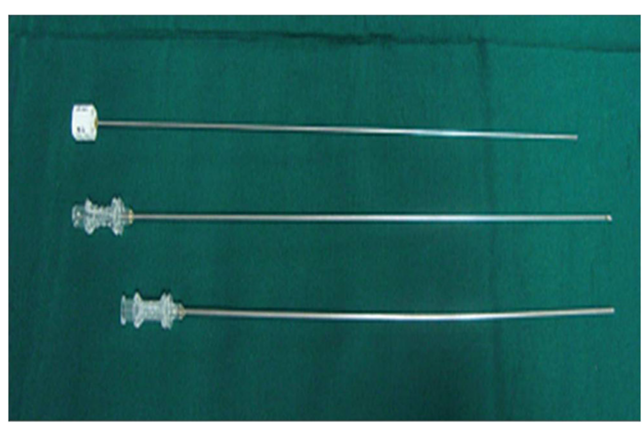

Fig. 1 Three-part Microperc needle

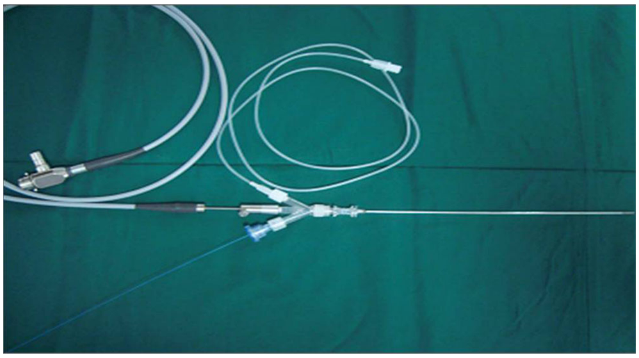

Fig. 2 Setup of microperc armamentarium

Ultra Miniperc

Recently, Desai et al. [32] described their novel modified technique of ultra-mini-percutaneous nephrolithotomy using of a novel 6 Fr mini nephroscope through an 11–13 Fr metal sheath to perform holmium: YAG laser lithotripsy. The authors successfully performed the procedure in 36 patients with a mean stone size of 14.9 mm, with no need for blood transfusion. Of these, two were preschool children. The technique was described as being safe and efficacious for moderate-sized renal stones with an advantage of high stone-free rates and low complication rates. Though it was used in both the pediatric and adult population with an age range of 2–79 years, its role and indications remain to be seen in larger prospective studies in exclusive pediatric populations.

Review of Current Literature

The options for management for upper tract urolithiasis include shock wave lithotripsy (SWL), percutaneous nephrolithotomy (PCNL), retrograde intrarenal surgery (RIRS), and open / laparoscopic / robotic approach (e.g., anatrophic nephrolithotomy).

Nevertheless, despite a wide array of management options, there needs to be a balance between stone clearance and morbidity related to the procedure. As a low-risk procedure with a high retreatment rates (18–67 %), ESWL often leads to persistent residual stones [33]. The developing RIRS can minimize the risks associated with bleeding and visceral injury, but there are times when the pelvicalyceal anatomy [34] is not ideal, and because of the anatomical delicacy of pediatric ureter, flexible ureteroscopy may not be an ideal option.

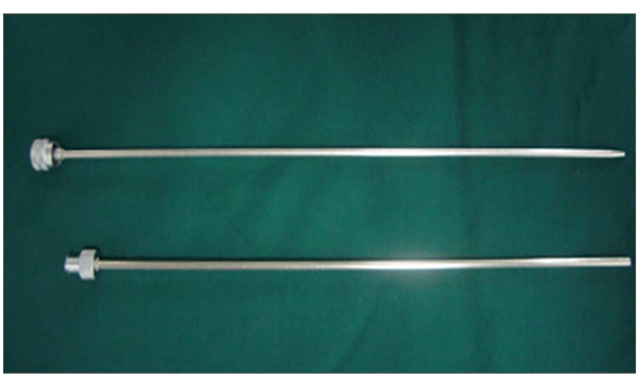

Fig. 3 Mini-microperc sheath with obturator

Being a safe procedure in adults, PCNL was first described in children by Woodside et al. in 1985, having been performed in seven children, with 100 % stone removal in one session, using standard adult instruments (24–34 F) [35]. Ever since then, there have been multiple studies by various authors in pediatric PCNL (Table 1).

Various imaging techniques have been used to define stone clearance and success rates. Most of these studies, however, have utilized either plain X-ray abdomen alone [41–43], or a combination of the former with ultrasonography [36, 38, 47••, 59] or a nephrostogram [39, 40, 45, 48]. Recently, there are authors [57] who have also utilized non-contrast CT for evaluation of stone clearance. As far as the intra-corporeal lithotripters are concerned, ultrasonic, pneumatic and lasers all have been used successfully in this patient population. The authors believe that so long as judiciously utilized, any of these can be used with good outcomes.

Although there is no current international consensus, relative indications for PCNL as the primary treatment modality in children include large upper tract stone burden (>1.5 cm), lower pole calculi greater than 1 cm, concurrent anatomic abnormality impairing urinary drainage and stone clearance, or known cystine or struvite composition [60].

Despite the initial skepticism associated with PCNL in pediatric patients arising as a result of larger sized instrumentation causing renal trauma and the associated effects of the same on renal function, radiation exposure with fluoroscopy, and the risk of complications including sepsis, hypothermia and bleeding, it continues to be increasingly utilized as monotherapy and in combination with SWL (sandwich therapy) in children and adults [61]. As it was realized that morbidity of PCNL is related to the tract size and number, this led to the introduction of the concept of sandwich therapy [62].

The concern in these small patients relates to the potential for renal damage due to percutaneous access tracts. This led to evolution of technique and technology in pediatric percutaneous surgery, wherein the tract size has reduced and energy sources have become efficacious and safe. This in turn has given an impetus to developing techniques such as microperc and miniperc. As a result, PCNL has now replaced open surgery as the treatment of choice for large stone burdens in children of all ages. Recently, Zeng et al. [59] reported their experience with infants having a mean age of 20.6 months (range 7–36 months) and all the procedures had been performed with 14 to 16 F percutaneous access and a stone free rate of 95 %. Guven [63] et al. reported high stone free rate in

Table 1 Outcomes in various series

Study	Renal units	Mean age (years)	Stone size	Tract	Complications	Success rate
Mor et al. [36]	25	8	Not available	26	Fever, sheath dislodgement, Steinstrausse	68 %
Jackman et al. [37]	11	3.4	12 mm	11	None	85 %
Badawy H et al. [38]	60	6	1.8– 3.7 cm^2	28	Fever, colon injury, urine leak, open conversion	90 %
Al-Shammari et al. [39]	10	6.4	17.4 mm	24	Hypothermia	87.5 %
Zeren et al. [40]	62	7.9	283 mm^2	18–30	Fever, hemorrhage requiring transfusion, open conversion	86.9 %
Rizvi et al. [41]	62	6-10	47 mm	22	Fever, hematuria requiring transfusion, open conversion, urine leak, hydrothorax	67.7 %
Desai et al. [42]	128	9.1	246 mm^2	20–24	Urine leak	93.7 %
Salah et al. [43]	138	8.9	507 mm^2	26	Urine leak	98.6 %
Holman et al. [44]	138	8.9	22.5 mm	28	Fever , urine leak	98.5 %
Raza et al. [45]	43	6.4	40 mm	24	Fever , pain, abscess and fistula	79 %
Samad et al. [46]	188	8.2	27.2 mm	28	Fever, hyponatremia, hypokalemia, transfusion	47-90 %
Shokeir et al. [47••]	82	6.6	14.4 mm	30	Urine leak	95.1 %
Bilen et al. [48]	46	9.6	18.2 mm	26	Fever, hydrothorax urine leak	87.8 %
Nouralizadeh et al. [49]	20	3.1	20–46 mm	26	Overall -15.38 %	91.67 %
Onal B et al. [50••]	1,205	8.8±4.7	4.09±4.06 cm^2	Cutoff size-20 F	Overall -27.7 % Bleeding requiring transfusion -10 %	81.6 %
Goyal NK et al. [51]	158	10.03±4.51	376.68±265.23 mm^2	24–30 F	Bleeding	93.7 %
Veeratterapillay R [52]	32	10.8	19 mm	28 F	No significant complications reported	91 %
Wah TM et al. [53]	23	1.6 to 14.6	3.44 cm^2	16 F	Postoperative hydrothorax	90.5 %
Yan X et al. [54]	27	42.6 months (range 14-68 months)	1.85 cm	14–16 F	Bleeding	92.6 %
Etemadian M et al. [55]	38	8.4±4.24	2.93±0.89 cm	26–30 F	Bleeding hyponatremia	67 %
Kumar R et al. [56]	34	11.7	848 mm^2	24–30	Fever Abdominal collection	-
Anand A et al. [57]	27	11.12	200 to 1150 mm^2	24–30	Abdominal collection, hydrothorax	96.1 %
Unsal A et al. [58]	45	≤16	2.37–3.41 cm	12–26	Bleeding requiring transfusion Pleural effusion Nephrostomy site leak	91.7 % - 94.1 %
Zeng et al. [59]	20	20.6 months	2.2 cm	14–16	No significant complications reported	95 %

infants and small children (5–36 months) with large renal calculi who underwent PCNL without major complications.

As far as the issue of kidney function is concerned in these cases, Mor et al. [36], studied radioisotope scans on children before and after PCNL, and showed no change in differential function. Similarly, Dawaba et al. [64] found no decrease in GFR following PCNL.

Earlier, authors have reported that dilatation even up to 26 Fr does not cause significant morbidity in children, and has been shown in animal models that there is no advantage in using a small access based on renal scarring alone [65•]. However, this issue has been challenged by later studies [26••]. Desai et al. [42] assessed percutaneous nephrolithotomy in children and found that the caliber and number of tracts were significantly correlated with the intra-operative hemorrhage and hemoglobin drop. However, because of the small size of the kidney and the more compact collecting system of infants, it seems logical that the use of the smallest and least traumatic instruments with least number of renal accesses may reduce the likelihood of major complications, including bleeding.

Bilateral renal stones in children are generally treated in a staged manner. The safe application of bilateral simultaneous PCNL was reported for the first time by Salah et al. [66] and later by Samad et al. [46]. These studies recommended simultaneous bilateral procedures because it reduced psychological stress, necessitated only one cystoscopy and anesthesia, exposed the children to less radiation, enabled less medication, and needed a shorter hospital stay and convalescence, and hence led to considerable savings in costs. Recently, Guven et al. [67] described their experience and feasibility in five patients with mean age of 6.28 years with high success rate and no significant complications.

Future Perspectives [68•]

Newer non-X-ray-based protocols are being investigated. These are based either on ultrasound or magnetic resonance (MR) guidance. The use of these appears to be appealing, as these are non-ionizing and give real time image acquisition. New three-dimensional ultrasonography provides volumetric measurements and 360-degree analyses of anatomic structures. Ultrasound–Doppler integration has been investigated as a means to prevent vascular complications during the initial access [69]. MR-based protocols allow for construction of images in specific planes, which provides a complete view of the total needle length and its spatial relationship relative to the puncture target [18, 70]. As with the application of robotics in other fields of urology, researchers are endeavoring to assess the role of novel robotic systems in PCNL, such as the PAKY-RCM, the AcuBot Robot and the MrBot [71]..

Conclusions

Due to the high incidence of predisposing factors for urolithiasis in children and high stone recurrence rates, every child with urinary stones should be made to undergo a complete metabolic evaluation. Because of its recurrent nature, every effort should be made to reduce the number of procedures performed to a minimum and to save the developing kidney from the deleterious effects of the intervention. From a review of the available studies, it can be said that PCNL can be performed safely and effectively in children to achieve high stone-free rates and allow a short treatment period in most cases. Despite the well-known hazardous and serious complications, most of which are related to tract formation and size, the rates of these are small, albeit, of real existence. However, with the combination of increasing surgical experience in this patient population and technological innovations, the outcomes are continuing to improve.

Compliance with Ethics Guidelines

Conflict of Interest Dr. Ravindra B. Sabnis, Dr. Jaspreet S. Chhabra, Dr. Arvind P. Ganpule, Dr. Sachin Abrol, and Dr. Mahesh R. Desai each declare no potential conflicts of interest.

Human and Animal Rights and Informed Consent This article does not contain any studies with human or animal subjects performed by any of the authors.

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- • Of importance
- •• Of major importance

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