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EDUCATIONAL REVIEW

Pediatric urolithiasis: the current surgical management

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Abstract Children represent about 1% of all patients with urolithiasis, but 100% of these children are considered high risk for recurrent stone formation, and it is crucial for them to receive a therapy that will render them stone free. In addition, a metabolic workup is necessary to ensure a tailored metaphylaxis to prevent or delay recurrence. The appropriate therapy depends on localization, size, and composition of the calculus, as well as on the anatomy of the urinary tract. In specialized centers, the whole range of extracorporeal shock-wave lithotripsy (ESWL), ureterorenoscopy (URS), and percutaneous nephrolithotomy (PCNL) are available for children, with the same efficiency and safety as in adults.

Keywords ESWL · PCNL · URS · Urolithiasis · Children · Pediatric urolithiasis · Endoscopy

Introduction

Children presenting with urolithiasis pose a challenge to the treating doctor, as they are part of the high-risk group for stone recurrence [1]. Although a rare disease in children living in developed countries, with a prevalence of between 1:1,000 and 1:7,600 in different parts of the USA, the number of pediatric patients per capita is increasing [2, 3]. For the pediatric patient, especially being a high-risk patient, it is of utmost importance that no stone material is left behind, no matter what therapy is employed, as recurrence rates are higher than in adults. Thirty-three percent of patients with small remaining stone fragments after extracorporeal shock-wave lithotripsy (ESWL) (<3 mm), which were formerly called clinically insignificant residual fragments (CIRFs) in the early era of ESWL, had an increasing stone mass on median follow-up of 24 months [4]. In a study conducted by Afshar et al., 69% of patients with residual fragments ≤5 mm on mean follow-up of 48 months after ESWL had symptomatic episodes or an increase in stone size [5]. Compared with stone-free individuals, patients with residual fragments had an increased risk for adverse clinical outcome, with an odds ratio (OR) of 3.9. If an underlying metabolic disorder was existent, the OR for growth of residual fragments was 11.4 [5]. This begs the question: Is ESWL still the best treatment for urolithiasis, or could other—i.e. endourological therapy—be better suited?

The standard procedures to treat pediatric urolithiasis do not differ from those used for adults: ESWL, ureterorenoscopy (URS), percutaneous nephrolithotomy (PCNL), and—in selected cases—laparoscopic surgery. Open surgery is reserved for selected cases, especially those with the need for anatomical correction of the urinary tract. Before choosing the appropriate treatment, it is indispensable to know the number, size, location, and composition of a stone, and in addition, any information about the urinary tract below the stone. Therefore, in our opinion, adequate imaging is obligatory.

Extracorporeal shock-wave lithotripsy

ESWL was first reported by Chaussy et al. in 1980 and has since changed the management of stone disease in adults

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and children, including infants [6]. ESWL was successfully introduced into the pediatric setting after Newman et al. reported its safety and effectiveness in 1986. At the time, ten of the 15 children were stone free, and three had small residual fragments [7]. The underlying function of all types of ESWL machines is to generate and focus shock-wave energy at a focal point that is clustered at the calculus. Ideally, the impact of the shock wave disintegrates the stone so the fragments can pass the ureter. It is known that ESWL is safe in adults, but there has been concern in the past that the high-energy impact may negatively affect kidney development [8]. Brinkmann et al. found no effect of ESWL on renal function or blood pressure, and there was no sign of renal scarring in children [9]. In addition, hematuria and proteinuria resolved after patients were stone free [9]. Lottmann and colleagues analyzed the treatment of 15 children aged 9 months to 15 years. They found no changes in blood pressure or signs of acquired parenchymal scarring 6 months after ESWL using 99 m-Tc dimercapto-succinate (DMSA) renal scans for evaluation [10]. Vlajkovic et al. evaluated glomerular filtration rate (GFR) using 99 m-Tc diethylenetriamine pentaacetic acid (DTPA) before, directly after, and 3 months after ESWL. The first measurement after ESWL showed a significant decrease in GFR, from 118 ± 7 ml/min pretreatment to 107 ± 6 ml/min posttreatment. On further follow-up, GFR normalized or even improved to 121 ± 6 ml/min 3 months after ESWL. GFR was 131 ± 10 ml/min at the end of the observation period (average of 3 years). The authors concluded that ESWL is a safe treatment in children [11].

Studies conducted to investigate short- or long-term effects demonstrate that there are short-term effects such as perirenal hematomas, hematuria, and reduced GFR directly after ESWL therapy. However, there has been no evidence for long-term damage in children [12–15]. One long-term study reported in adults by Krambeck et al. needs to be mentioned. Krambeck and colleagues conducted an epidemiologic study based on questionnaires 19 years after ESWL, the longest available follow-up after ESWL to date. According to the results, patients treated with ESWL were more prone to develop hypertension (OR 1.47), especially after bilateral treatment ($p=0.033$), than controls with nephrolithiasis not treated surgically. There was also an increased risk of developing diabetes mellitus compared with controls (OR 3.23). The risk of diabetes mellitus was directly related to treatment intensity and the amount of shock waves delivered ($p=0.007$; $p=0.005$, respectively) [16]. Results of Krambeck et al.'s retrospective study are questionable, as the control group had less severe urolithiasis and no individuals in the control arm with bilateral ESWL treatment [17]. Also, the authors grouped patients who had ESWL for kidney stones with patients who received ESWL for proximal ureter stones, which could make a significant difference considering the different focus of the shock waves. Knoll and colleagues prospectively examined 12 patients to determine whether they developed acute endocrine pancreatic injury after ESWL for renal stones and could not detect any effect on endocrine or exocrine pancreatic function. They therefore concluded it is unlikely that ESWL and diabetes mellitus are related [18].

ESWL is the preferred treatment in pediatric urolithiasis patients with calculi <20 mm. Stone-free rates after ESWL in children range between 57% and 92% [19–21]. In suspected cystine stones, the maximum diameter should not exceed 15 mm because of the hardness of the stone [18]. With growing endourologic options and the known hardness of cystine stones, a ureteroscopic or minipercutaneous nephrolithotomy (mini-PCNL, or mini-Perc) approach is coequal, if not the new first-line therapy. Thus, large and hard stones such as cystine and whewellite decrease ESWL success rates [22].

Another important aspect in treatment planning is urinary tract anatomy and stone location. ESWL treatment of stones in lower calices has a lower success rate due to the special anatomy and gravity situation, as noted by Sampaio and Aragao in 1992, anatomists, who suggested including the knowledge of the specific anatomy of a patient into treatment planning [23]. Tan and colleagues examined the radiological anatomy of their pediatric patients. They demonstrated that there is a significant difference between mean lower pole infundibular length ($p=0.0032$) and lower infundibulopelvic angle between stone-free cases and others. Interestingly, stone burden or infundibular width did not play a role in stone-free rates. A cutoff angle of 40° for the infundibulopelvic–ureteropelvic angle was an excellent method to predict the clearance of lower pole fragments after ESWL. The stone-free rate in this series of 34 patients was 55.9% (the average stone-free rate for lower caliceal stones being 50–62% in other representative series) [24–27].

Spontaneous stone passage is easier in children compared with adults, and ureter stenting is not needed as often (only 5–37%) as it is in adults [28–30]. Nevertheless, these patients should receive pharmacological support to pass their stone fragments. For pain control, acetaminophen and ibuprofen are the medication of choice, as passing fragments can cause colic. Alfa 1a adrenergic receptor antagonist, such as tamsulosin, can be used (off label) to facilitate the passage of fragments through the intramural part of the ureter [31, 32]. Even though ESWL can cause minor complications, including hematuria, bruising, renal colic, and perirenal hematoma, it is important to be aware that in children (up to the age of 14), all therapeutic approaches require anesthesia. Therefore, if a therapy is likely to be unsuccessful the first time, it may be better to use another technique that assures that the patient is stone free after one procedure [33–35].

Minipercutaneous nephrolithotomy

According to the guidelines of the European Association of Urology, ESWL should be the first choice for most renal pediatric stones, but a percutaneous approach could be used for bigger and more complex calculi [20]. To gain access to the renal pelvis or renal calices for a percutaneous nephrolithotomy, it is necessary to puncture a renal calyx under ultrasound guidance. Fluoroscopy is generally employed as soon as the puncture is successful. The puncture channel is dilated and the instrument inserted to start fragmentation and elimination of the stone under permanent videoscopy. For stone fragmentation, pneumatic lithoclast probes, ultrasound probes, or holmium:yttrium aluminum garnet (Ho:YAG) laser probes can be used. Stone fragments can then be captured with special wire baskets or graspers, which can be inserted through the working channel of the instrument.

Being a safe procedure in adults, PCNL is a safe procedure in adults and 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) [36]. PCNL in children has since become easier, after Jackman et al. introduced the mini-perc technique using an instrument with only 11 F compared with the 24- to 34-F sized instruments [37]. In that series, the stone-free rate was 85%, whereas the numbers in the current literature range between 86.9% and 98.5% [38–40]. With this new technique, it is even possible to render 89% of patients with staghorn calculi stone free in one session [41].

Although there are no international guidelines as to when PCNL should be the primary treatment in children, there are relative indications, such as large stones (>1.5 cm) or >1 cm for lower-pole concrements. Especially if there are anatomical abnormalities that prevent good fragment clearance (i.e. ureteropelvic junction obstruction, calyceal diverticulum, ureter stricture), and depending on stone composition, PCNL can be the treatment of choice [22, 42].

As was the case with ESWL, there was concern about renal scarring and loss of renal function (or other long-term effects) due to PCNL. Dawaba et al. used DMSA renal scans to search for renal scarring after PCNL but found none, whereas others found minimal scarring [40, 43]. Dawaba et al. also used DTPA renal scans to follow-up renal function after PCNL and noted that except for one patient (out of 65), renal function improved or stabilized after percutaneous procedure [40]. Reported complications of PCNL include postoperative fever (30%) and bleeding, the latter being the most reported complication associated with PCNL and consecutive transfusion rates ranging from 0% to 23.9% [37, 38, 44–48]. The risk for blood transfusions is generally very low, with the report of one transfusion in 287 procedures in four pediatric studies [38, 44, 45, 47]. The highest reported number of transfusions needed (23.9%, or 16 out of 67) was by Zeren et al., which occurred mainly during their early PCNL procedures. The authors compared the 16 patients who received blood transfusions to the those who did not and found a significant difference between stone burden, sheath size, and operative time [46]. Although PCNL is an invasive treatment, it achieves excellent stone-free rates and comes with a relatively low risk in experienced hands, especially if performed as mini-perc. It is always necessary to keep in mind that the child should be stone free after one treatment; therefore, a good preselection to every treatment is the key to success.

Ureterorenoscopy

URS for concrements in the distal ureter in children was first reported by Ritchey et al. in 1988, with stone-free rates in the early series of 86–100% [48–54]. URS has become available in children due to the invention of smaller diameter ureteroscopic instruments. Today, pediatric rigid endoscopes of 4.5 F and good applicable tools are routinely available. Rigid and flexible endoscopes are on the market, with rigid endoscopes providing a bigger working channel, whereas flexible endoscopes can bend up to 270° and are especially useful for removing stones in the lower calices. URS is ideally suited for calculi in the mid and distal ureter. De Dominicis et al. compared URS and ESWL in 31 children in a randomized fashion. After one treatment, 94% (16 of 17) of patients were stone free in the URS-treated group compared with 43% (6 of 14) in the ESWL group (64% after two sessions) [51]. There was a concern that due to the instrumentation—dilation of the ureteral orifice—vesicoureteral reflux and strictures might follow stone removal via URS. However, Schuster et al. found in a literature review of 221 URSs in children that only two patients had ureteral strictures and eight had low-grade vesicoureteral reflux [50].

As complications do not seem to differ compared with URS in adults, this procedure (including in the proximal and intrarenal calculi) has become a first-line treatment for stone disease in pediatric patients in many clinics [50, 52, 53, 55–57]. All those series reported excellent stone-free rates. Unfortunately, it was not possible to examine the stone-free rate for proximal or intrarenal calculi separately, as the number of reported cases is limited. Cannon et al. treated 21 patients with a mean age of 15 years. The success rate of lower-pole calculi removal was 76% [57]. Tanaka et al. evaluated 52 URS procedures for intrarenal calculi [58] and found that retrograde endoscopy for intrarenal calculi was safe, and there was a primary stone-free rate of 50% after one procedure. With a mean follow-up of 246 days, the stone-free rate increased to 58%. Predictors for further treatment were younger age and preoperative stone size >6 mm. Re-treatment was not necessary if the stone size ≤6 mm. That study clearly demonstrates that treatment of lower-pole stones often requires additional treatment, no matter what kind of therapy is first used. Pearle et al. reported a prospective randomized trial comparing ESWL and URS for lower-pole calculi up to 1 cm [59]. On radiographic follow-up 3 months after the procedures, 35% of patients with ESWL were rendered stone free, whereas this was true for 50% of patients in the URS group. This difference did not achieve statistical significance, but there were more complications in the URS group (1 vs. 7). Thus, ureteroscopy has become a first-line treatment for ureteral stones and can even be considered a good treatment option for renal calculi.

Laparoscopic surgery/open surgery

In developed countries, open surgery remains the treatment of choice for 0.3–5.4% of children. In general patients with anatomical abnormalities—i.e. ureteropelvic junction obstruction, obstructive megaureter, urolithiasis—will receive open surgery if stone removal and anatomical correction can be combined in one operation [60]. In developing countries, open surgery is used in 14% of cases, which is likely due to the fact that open surgery is more cost effective in those countries [61]. Zargooshi et al. reported a 95.4% stone-free rate in the treated children and even 100% for single stones. Casale et al. successfully used a transperitoneal laparoscopic pyelolithotomy in eight children, with a mean stone burden of 2.9 cm after percutaneous access had failed [62]. One hundred percent of their patients were stone free afterward, although one patient had a recurrence at 12 month follow-up. Between 2002 and 2005, five adolescents (mean age 16.6 years) underwent robot-assisted laparoscopic pyelolithotomy. One patient had to be converted to an open procedure due to inability to remove the staghorn stone. Three of the four other patients were stone free after surgery, and one had a remaining lower-pole calculus of 6 mm. The authors stated that the procedure is safe and efficacious but that its role in stone management needs to be determined [63].

Conclusion

Due to the miniaturization of instruments and better optic and ancillary instruments, the use of ESWL is decreasing. With the primary goal of complete stone removal in one session to prevent recurrence from residual fragments, endoscopic treatment is on the rise.

Questions

(Answers follow the reference list)

1. The incidence of stone disease in children is increasing. How many of them are considered high risk for recurrence?
   a. 25%
   b. 50%
   c. 75%
   d. 95%
   e. 100%
2. What is not a typical side effect of ESWL?
   a. Hematuria
   b. Proteinuria
   c. Increased GFR
   d. Renal colic
   e. Perirenal hematoma
3. For a 2-year-old child who presents with a 1-cm stone in the left renal pelvis and no known anatomical problems, the treatment of choice is:
   a. PCNL
   b. ESWL
   c. URS
   d. Mini-Perc
   e. Laparoscopic surgery
4. For the treatment of an intrarenal stone, the following variables need to be considered. Which one is NOT one of them?
   a. Gender
   b. Stone size
   c. Patient age
   d. Urinary tract anatomy
   e. Stone location
5. Anesthesia in children is not required for:
   a. ESWL
   b. URS
   c. PCNL
   d. Mini-Perc
   e. Metabolic intervention

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Answers

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