PERCUTANEOUS NEPHROLITHOTOMY IN CHILDREN

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ABSTRACT

Although urolithiasis is considered as a rare entity in children, it is an important health problem in the world, especially in endemic areas. When the long lifespan expectation for children is considered, minimally invasive surgical procedures such as percutaneous nephrolithotomy (PNL) have become important recently. Despite the amount of PNL procedures that have taken place in the literature and guidelines, with enormous clinical knowledge, there are inadequate quantitative data concerning the impact of this procedure and radiation on growing kidneys and children. Within this review, we aim to identify the effects and side-effects of PNL on children, and to present the importance, and difficulties, as well as some tricks of the procedure.

Keywords: Percutaneous nephrolithotomy, children.

INTRODUCTION

The prevalence of urolithiasis disease has been increasing because of changing dietary habits, obesity, and lifestyle factors.[1] The prevalence of stone disease was reported as 11.1% in the adult population. While the prevalence in children varies with age, it is approximately 2-3%.[2,3] The increased prevalence of paediatric stone disease has also been on the rise due to increased utilisation of imaging modalities in children. It has been reported that patients with early onset of urolithiasis (especially children and teenagers) are high-risk stone formers.[4] Stone disease in children could also be associated with systemic disease that could put their general health at risk. Thereby, children need full metabolic and systemic evaluation. When the expectation of a long lifespan for children is considered, it is of utmost importance to consider the possible need for surgery in the future. Thus, for the surgical treatment, minimally invasive techniques have become more important in children, and with technological advancements, instruments have become miniaturised with high-quality imaging.

While open surgery was the only surgical treatment option in the past, stones can be managed today with minimally invasive procedures, such as extracorporeal shock wave lithotripsy (ESWL), retrograde intrarenal surgery (RIRS), percutaneous nephrolithotomy (PNL), or laparoscopy. The current treatment recommendations for kidney stones in children are similar to those in adults. For kidney stones <2 cm, ESWL has been recommended as the first-line treatment.[5-9] There are various factors affecting the stone-free rates of ESWL such as stone size, composition, and location. European Association of Urology (EAU) Guidelines recommend ESWL as a first-line treatment option in children with upper ureter or renal pelvis stones <2 cm, and with lower pole calyx stones <1 cm.[10] An increase of the stone size results in a decrease of the stone-free rates and an increase of retreatment rates. Regardless of the location, stone-free rates for <1 cm, 1-2 cm, and >2 cm, were reported as nearly 90%, 80%, and 60%, respectively. The main disadvantage of the ESWL is the need for anaesthesia. But due to new advances in medication, anaesthesia does not seem to be a problem anymore, except in infants.[10]

RIRS, which can be considered as a new approach for renal stones, has greatly improved as a result of advances in the technology of flexible ureteroscopes.[11-13] However, because it is a new approach in children, it is not recommended under the EAU guidelines as a first-line treatment procedure for any of the kidney stones yet. It is mentioned as a secondary treatment option to ESWL in children with stones <1 cm.[10]

For the treatment of kidney stones >2 cm, it is recommended that PNL be used as first-line treatment as it returns high success rates. According to EAU guidelines, PNL is the primary treatment option in children with kidney stones >2 cm or with staghorn calculus, and EAU guidelines also recommend PNL for lower pole calyx stones >1 cm in children.[10] Furthermore, obstructed kidneys, hard stones such as cysteine and calcium oxalate monohydrate, and ESWL-failed kidney stones are other indications for PNL.[10,14-16] For proximal ureteral stones, there is no evidence about the impact of PNL in children. So for proximal ureteral stones in children, while ESWL is the primary treatment option, PNL is recommended as a secondary treatment option such as ureteroscopy and open surgery.[10]

There are many factors affecting the success rate of PNL, such as the anatomy of the kidneys, stone burden, and localisation. It has been reported in the literature that the stone-free rate is between 73-96%.[16-18] Although the success rate with PNL is similar between children and adults, small kidneys and large instruments make this surgery difficult in children. All the complications that occur in adults can also be experienced by children. Children have less tolerance for bleeding, and this lack of tolerance can cause an anxiety in surgeons when performing PNL. The impact of tract dilatation on growing kidneys and the side-effects of radiation are also important topics that must be defined. And before performing PNL in children, we should be aware of the effects of the procedure on kidney and patient.

There are many articles which reveal that ESWL has no long-term morbidity on kidneys.[10] But there are insufficient data and few articles regarding the impact of PNL on growing kidneys in the international literature. Unsal et al.[19] evaluated 50 patients by quantitative single-photon emission computed tomography (QSPECT) of (99 m) Tc-dimercaptosuccinic acid (DMSA) before and 3-6 months after surgery. They found that six of the patients had new focal cortical defects corresponding to the access site for tract formation during PNL. In contrast, they demonstrated that renal function was preserved after percutaneous stone removal. Wadhwa et al.[15] showed that PNL did not cause adverse renal morphologic or functional alteration in children. Reisiger et al.[20] calculated the expected renal length using Chen's nomogram. They observed that ESWL, ureteroscopy,

and PNL did not impair renal growth with a 6-year follow-up period.

In contrast, Moskovitz et al.[21] reported adult patients whose functional volumes from surgically treated regions statistically significantly decreased, although their global uptake was not affected by PNL. It is understood from the passage that they performed standard PNL procedure, and any of the patients underwent mini or micro PNL, which are described recently. There is plenty of evidence that the complication rate declines with the decrease in size of instruments. But there is a lack of quantitative knowledge regarding whether the size of instruments is associated with the global and regional renal functions. Despite there being a number of articles demonstrating that PNL does not reduce renal functions and that instruments have become smaller, we still need further prospective studies to investigate the impact of PNL on growing kidneys in children.

Another important topic about PNL is radiation hazards in children. The International Commission on Radiological Protection (ICRP) reported that the safe annual dose was 150 mSv for the eyes and 500 mSv for the hands, skin, and other organs. But the total radiation dose of >50 mSv is the recommended annual dose limit for occupational exposure by ICRP.[22-24] Kumari et al.[25] showed that patients receive 0.56 mSv of radiation with a mean fluoroscopy time of 6 minutes per surgery. Bush et al.[26] documented that the skin on the flank area, the testes, and the ovaries received 0.25 mSv, 1.6 mSv, and 5.8 mSv, respectively. They found that gonadal doses from PNL were similar to those with 7-view excretory urography. Using ultrasonography, while performing renal access, reduces the radiation exposure in children. Radiation-free PNL should be standard, especially in children, while there are many series presently on how easy, safe, and successful the procedure is.[27,28] But patients are exposed to radiation, not only during the PNL procedure, but also during the diagnosis and follow-up period. After an acute stone episode, a mean of 1.2 (0-7) plain abdominal films, 1.7 (0-6) computerised tomography, and 1 (0-3) excretory urogram are required during the first year of follow-up. The median total effective radiation dose per patient is 29.7 mSv, and 20% of patients receive potentially significant radiation doses (>50 mSv) in the short-term follow-up of an acute stone event.[24]

The biological effects of radiation on tissue have been examined under two headings, as deterministic

and stochastic effects. Deterministic effects include cataracts, infertility, and damage to the skin, gastrointestinal tract, and haematopoietic tissue. Deterministic effects are dose-dependent, and their intensity increases with increasing doses. In contrast, the stochastic effects of radiation include genetic changes, such as cancer, and these effects are not associated with the dose of radiation. There is no threshold value, and these effects can occur after exposure to low doses of radiation. But if a tumour in the future is detected, it is impossible to say whether the radiation is responsible or not with the available technology.[29,30]

The risk of radiation-induced stochastic effects is greater in children than in adults.[31] Therefore, radiation should be avoided in paediatric patients as much as possible. Ultrasound (US) should be preferred for diagnosis and follow-up, and also for performing renal access during PNL procedure. If the surgeon provides renal access via fluoroscopy, he/she has to decide before and after shooting fluoroscopy, not while shooting, to keep the fluoroscopy time at a minimum.

PERFORMING PNL IN CHILDREN

PNL has a learning curve, just as other surgeries and surgical competence in PNL can be obtained after 60 cases and excellence after 115.[32,33] After the first 35 paediatric PNL experiences, the surgeon can perform PNL safely, even on very young children, and paediatric PNL should be performed after 120 adult PNL procedures.[34] In other words, paediatric PNL should be performed after the surgeon has become an expert at performing it in adults.

PNL in paediatrics is similar to adult PNL. The procedure is performed following the standard procedures, starting with insertion of a 4 Fr or 5 Fr ureteral catheter in the lithotomy position under general anaesthesia. Renal access can be obtained with ultrasonographic or fluoroscopic guidance, based on the surgeon's experience. If fluoroscopic guidance is preferred, lead aprons should be laid under the patient, on all areas except the flank area, to protect him/her from radiation exposure (Figure 1). Percutaneous access should be transparenchymal toward the posterior calyces. Access should not be directly into the renal pelvis and should be parallel to the infundibulum. It should always be borne in mind that the length between the parenchyma and the calyx is shorter in

Figure 1: Percutaneous nephrolithotomy performed on a 9-month-old male child.

Figure 2: A paediatric mini-percutaneous nephrolithotomy performed dilatation of up to 16 Fr.

children. Following the appropriate calyx entrance, a guidewire is placed into the collecting system.

Then, the nephrostomy tract is dilated to an appropriate size. Because the kidneys are smaller in children, a 24 Fr dilation in infants corresponds to a 72 Fr dilation in adults.[35] It has been reported in some literature that the risk for bleeding increases after 24 Fr tract dilation, and in children younger than 3-years-old, tract dilation should be restricted to 20 Fr or smaller. Moreover, in preschool-aged children, adult instruments are not recommended.[18,22,35] In light of this information, mini-PNL, ultra-mini-PNL, and micro-PNL procedures to avoid bleeding complications, have recently been described, based on miniaturisation of the

instruments and tract dilation. In standard adult PNL surgery, renal access is obtained through 24-30 Fr access sheaths. However, although it has not yet been standardised in children, the definition of mini-PNL has included a low dilatation size of the tract. In several studies, authors have accepted 16-22 Fr for mini-PNL.[36-39] However, in the 2013 EAU guidelines, dilatation of up to 18 Fr was described as mini-PNL4 (Figure 2). The most recent development is micro-PNL which includes 5-8 Fr seeing needles. Experience with micro-PNL has rapidly increased in a short period. Silay et al.[40] demonstrated the first series of micro-PNL on 19 children. It is understood that this procedure is effective and safe for small and moderate kidney stones. Micro-PNL procedure has been described to reduce overall kidney trauma, but it has limited applicability for large stones and for stones located in anterior calicies. On the other hand, it is superior to RIRS in patients with lower pole stones and with acute infundibular angle or with calyceal diverticulum.[40,41] Although this procedure is becoming popular in children, more evidence needs to be obtained by a large series.

The tract size should be sufficiently large to remove fragments, and sufficiently small to decrease the risk of bleeding. Fragmentation can be accomplished using pneumatics, US energy, or holmium laser. After this stage, the procedure is the same as with adult PNL. A nephrostomy tube should be placed routinely at the end of the surgery, and an antegrade double J (DJ stent) can be inserted if necessary. Although tubeless PNL can be performed in selected adolescent and adult patients, a nephrostomy tube of the smallest diameter should be placed in infants and preschool–aged children. The need for another cystoscopy under general anaesthesia to remove the DJ stent is another limitation of tubeless PNL.[42,43]

The difficulties of paediatric PNL are mostly associated with the small size of the kidney. Because of the hypermobility of the kidney in children, it can be difficult to enter the kidney, and mucosal bleeding can occur easily. The access sheath can be removed in children from the kidney during PNL, because the renal parenchyma is thinner, and the line between the parenchyma and mucosa is often unclear.[22] Therefore, it can be useful to perform a second guidewire replacement in children.

Although PNL is an effective minimally invasive treatment, it can result in some serious complications. Because of the complicated classification systems, such as Clavien and Satava, including minor incidents, complication rates of up to 83% have been reported in the literature. However, major complications, such as sepsis, bowel, and colon injuries, have been extremely rare. Complication rates in children have been similar to those in adults, and all complications in adults may occur in children.[44-47] But the possibility of the rapid deterioration of haemodynamics in children and their lower tolerance than adults for complications, such as bleeding, are the most important factors that can cause anxiety in surgeons.

The most important complication is bleeding, and it can occur during any step of the operation. In the literature, the blood transfusion rates have been similar in children and adults at <10%. Renal haemorrhaging requiring intervention is a rare complication, with a frequency of 0.6-1.4%. The risk of bleeding can be reduced by creating a posterior calix entrance with fewer tracts and less dilation.[10,22,45,46,48-51] Kapoor et al.[18] recommended not dilating more than 24 Fr and not increasing the number of tracts to reduce bleeding. Bilen et al.[52] reported that blood transfusion rates were similar between 20 Fr and 26 Fr dilation, but 14 Fr dilation resulted in a significant reduction in the rate of transfusion. Güneş et al.[53] reported that using adult instruments in children younger than 7 years old with staghorn calculi significantly increased the rates of complications. In several studies, it has been shown that intraoperative bleeding is associated with the degree of tract dilation.[35,54,55] A decrease in haemoglobin can occur after all PNL operations; however, because of the restrictive effect of the retroperitoneum, these decreases are mostly self-limiting. Therefore, bleeding is often controlled by conservative measures, such as monitoring of vital signs and fluid resuscitation therapy, or by blood transfusion if necessary. If the bleeding is not self-limiting, insertion and clamping of a nephrostomy tube is often adequate to control bleeding that derives from the tract location. In cases of haemorrhaging that cannot be controlled with a clamped nephrostomy tube, tamponade with a nephrostomy balloon catheter can be placed in the tract. In a recent study, we evaluated the emerging intervention criteria for bleeding after PNL.[48] In this study, we demonstrated that blood transfusion and fluid replacement were not sufficient to stabilise patients if the decrease in haemoglobin was associated with anuria/oliguria and metabolic acidosis. If emergent surgical

intervention is not performed, vascular collapse is the next step before cardiovascular arrest. When we reviewed the literature regarding this topic, it was the only study in adult patients, and there were no similar studies in children.

Children cannot tolerate acute and severe bleeding like adults can, and their haemodynamics can deteriorate suddenly. Therefore, postoperative vital signs, urine output, and the parenteral fluid amount and rate should be monitored closely. Postoperative haemogram control should be performed routinely. In self-limited bleeding and slight asymptomatic decreases in haemoglobin, the red blood cells will recover quickly with iron therapy and without transfusion. If there is not acute massive blood loss, crystalloid and/or colloid solutions should be used to increase tissue perfusion. However, if there is a serious decrease in haemoglobin, erythrocyte suspension should be administered to the patient. Before major surgery, haemoglobin value in children must be >10 g/dl. However, blood transfusion should not be performed in children unless necessary. If there is no massive postoperative haemorrhaging but an asymptomatic decline in haemoglobin, a blood transfusion is not recommended. If symptoms such as tachypnoea, dyspnoea, apnoea, tachycardia, bradycardia, difficulty with feeding, or lethargy are detected, transfusion should be performed, and the haemogram value is the threshold value for that child. The most important problem is that this threshold value is different based on age, and in any child. A 3 ml/kg erythrocyte transfusion can increase the haemoglobin value by approximately 1 g/dl. The amount and speed of transfusions are determined by the patients' clinic and can increase to 10-15 ml/kg. In patients at risk of heart failure, transfusions must be performed at a rate of 2 ml/kg/hour after the administration of diuretics.[56] In cases in which the clinical findings are deteriorated, surgical intervention should be performed immediately.

Postoperative sepsis has varied between 0.3% and 4.7% in the international literature.[44-46] Bayrak et al.[51] reported in their series that the postoperative fever rates in children and adults were 5.4% and 5.6%, respectively. Chan et al.[57] defined the risk factors for sepsis after PNL, such as the number of tracts, stone volume, presence of pyelocaliectasis, and blood transfusions. The most important preventive measure to reduce the risk of sepsis is to render the patient's urine sterile before surgery. Preoperative urinalysis and urine culture should be performed routinely in all patients. If bacteria are determined by urine culture, an appropriate antibiotic therapy should be administered, and the operation should be performed after confirmation that the urine is sterile. In the light of this information, some studies have shown that preoperative nitrofurantoin treatment for 1 week in adult patients reduced the risk of sepsis.[58,59] However, in paediatric patients, there has been no research into the use of long-term preoperative prophylactic antibiotics. In the presence of pyonephrosis, drainage should be provided first by placing a ureteral DJ stent or nephrostomy tube; then, infection should be treated with antibiotic therapy. The PNL procedure must be performed when the patient is stable and the urine is sterile.[60] If pyonephrosis is detected during the procedure, PNL should be postponed after placing a nephrostomy tube.

Another important point is that hypothermia can develop in children very quickly. Children are much more prone to hypothermia than adults, and this risk is further increased, especially in infants, due to their relatively large body surface area. Brown adipose tissue is used for thermal control, and breathing acceleration is used for increased oxygen requirements. If hypothermia deepens further, respiratory depression and metabolic acidosis can occur. In hypothermia, the anaesthesia requirement decreases so that the anaesthetic in circulation increases, and overdose, deep anaesthesia, and delays in awakening can occur.[61] In addition, it should be borne in mind that hypothermia can result in complications such as platelet dysfunction, decrease in drug metabolism, increased risk of infection, and cardiac arrhythmia.[35] It is possible to prevent hypothermia by taking the necessary measures. In this context, regulation of room temperature, the use of heaters, extremity binding with cotton, and heating of fluid before use as methods of prevention are usually sufficient. Additionally, the PNL overlay used during the procedure should not be allowed to leak water beneath it. Care should be taken not to allow the child to become wet.

Different distributions of fat mass and varying intracellular and extracellular fluid rates make fluid-electrolyte and volume balances more sensitive in children.[61] Some of the fluid leaking out to the retroperitoneum during PNL is absorbed, so saline solution must be used during the procedure. A disadvantage of paediatric PNL is the extended operative time due to the small tract size, particularly in patients with a greater stone burden.

Especially in extended cases, increased pressure can cause tears of the collecting system and increase the amount of absorbed liquid. These tears will close spontaneously within 1-2 days, so it should be sufficient to keep the nephrostomy tube in for a few days. During the operation, if it is noticed on fluoroscopy that the kidney is pushed medially due to fluid in the retroperitoneum, such tears should be suspected. Intraperitoneal extravasation is a rare entity. Because the patient is in the prone position during the procedure, it is difficult to detect extravasations in these cases. However, anaesthetists should be aware in such cases of increases in diastolic blood pressure and decreases in ventilation. In such situations, inserting a drain into the peritoneum is usually sufficient.[62]

Apart from these complications, the complications seen in adults, such as adjacent organ injury, hydropneumothorax, postoperative fever, and leakage from the nephrostomy tract, can also occur in children. The treatment algorithms for these complications are similar to those in adults.

Another important topic is metabolic evaluation of children after PNL and its importance as a successful surgery. Although PNL is an effective treatment for renal calculi, it is not sufficient for the continuation of a stone-free condition due to underlying metabolic abnormalities. It is well-known that patients with early onset of urolithiasis are high-risk stone formers.[1,4] Increased recurrence of urolithiasis is expected in children with metabolic abnormalities, and stone disease may be a sign of a systemic disease which can cause other health problems just as in primary hyperoxaluria, Lesch-Nyhan syndrome, or Xanthinuria. A genetic disorder, cystinuria, is another important reason for repeated stone formation and should be taken into consideration, especially in children and the younger population (<30 years).[63-65] Hypocitraturia and hypercalciuria are the most common metabolic disorders in children reported in literature.[66,67] According to our previous research, hypomagnesaemia was the most common metabolic abnormality in children with hypocitraturia and hypercalciuria. Another important finding of the study was that all children had at least one metabolic disorder.[1] Therefore, a full evaluation is necessary in children and metabolic evaluation should be performed after surgery, when the patient is stone-free, and the urine is sterile without haematuria.[1,4,68]

CONCLUSION

In conclusion, before performing PNL in children, sufficient surgical experience must be obtained in adults. During the procedure, radiation should be avoided as much as possible. The importance of radiation and the impact of surgery on growing kidneys must become clearer, with further well-designed, prospective studies. Although surgical complications are similar in adults and children, the hypermobility of the kidney in children can make it difficult to enter the kidney and can cause complications. The basic treatment principles for complications are similar to those with adults. To reduce serious complications such as bleeding, the mini-PNL and micro-PNL concepts have been introduced, which reduce tract dilatation. Mini-PNL is becoming increasingly common, especially in children, and paediatric PNL tools are widely available on the market. In addition, metabolic evaluation in children is another important procedure that should be performed after surgery.

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