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REVIEW

The management of staghorn calculi in children

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KEYWORDS

Child;
Kidney calculi;
Staghorn;
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ABBREVIATIONS

ESPU, European Society of Paediatric Urology; PCNL, percutaneous nephrolithotomy

Abstract ***Objectives:*** To review reports focusing on the surgical treatment of staghorn stones in children, as despite all the improvements in the surgical treatment of paediatric urolithiasis the management of staghorn calculi still represents a challenging problem in urology practice.

Methods: To evaluate current knowledge about treating staghorn calculi in children, we searched PubMed for relevant articles published between 1991 and 2011, using a combination of related keywords, i.e. staghorn stone, child, kidney calculi, surgical treatment, electrohydraulic shockwave therapy (ESWL), percutaneous nephrolithotomy (PCNL), and open surgery. Reports relating to the treatment of paediatric stone disease in general (open surgery, PCNL, ESWL) were also searched with the same method. Additional references were obtained from the reference list of full-text reports.

Results: Although open surgery had been widely used in the past for treating such stones in children, currently it has only limited indications in highly selected patients. Current published data clearly indicate that, in experienced hands, both PCNL and ESWL are now effective methods for treating staghorn calculi in children.

Conclusions: Due to advanced techniques and instrumentation, it is now possible to successfully treat staghorn calculi in children, with very limited safety concerns.

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Currently, while PCNL is recommended as the first-line surgical treatment, ESWL, open surgery and/or combined methods are valuable but secondary options in the treatment of paediatric staghorn calculi.

Introduction

Of the human population, 5–10% have stone disease during their lifetime, and of these cases only 2–3% are children [1,2]. The incidence of the pathology has been found to show marked epidemiological variations between developed and developing countries, with a prevalence of 1–5% and 5–15%, respectively [3]. Although the disease has been reported to be particularly rare in some countries, such as Scandinavia, it is still an endemic problem in countries such as Turkey, Iran, Pakistan and the Far East [1,2]. As for staghorn stones in this specific population, ≈19% of children have been reported to present with such stones at first referral [4].

Staghorn stones

Staghorn calculi are branched stones that occupy a large portion of the collecting system. Although there has been no consensus on the exact definition of 'staghorn stone', typically they are defined as kidney stones that fill the renal pelvis and branch into the calyces. The terms 'partial' or 'complete' staghorn are used when a calculus occupies the pelvicalyceal system partly or entirely, but the designation of 'partial' or 'complete' staghorn calculus does not imply any specific volume criteria [5]. As to chemical composition, most of these stones are composed of magnesium ammonium phosphate (struvite) and/or calcium carbonate apatite. While some other stones, e.g. cystine or uric acid stones, can also form a 'staghorn' configuration, calcium oxalate or phosphate stones rarely grow in this configuration. As struvite/calcium carbonate apatite stones have a strong association with UTIs caused by specific organisms that produce urease, they are generally referred to as 'infection stones' [6]. Repeated UTIs with urea-splitting bacteria can result in stone formation, and once an 'infection stone' is present, infections tend to recur [7,8].

As in all patients with stone, the ultimate goal of a surgical treatment should also be to maintain the patient free of staghorn stones, as the presence of residual stone fragments, regardless of treatment method, is associated with an adverse clinical outcome [9]. Although some studies in adults suggest that it might be possible to sterilise small residual struvite fragments and limit subsequent stone activity [5,10], most studies indicate that residual fragments can grow and be a source of recurrent UTI [11,12]. An untreated staghorn calculus could risk damaging the kidney as a result of not only obstruction but also, and more importantly, infection-related problems. Thus, when treating such calculi the ultimate aim of management must be to remove all existing stones, leaving no residual fragments and aiming to free the patient of stone as much as possible, particularly in children. Last but not least, in addition to the eradication of the causative micro-organisms, relief of any obstruction, and appropriate and adequate management of UTI, the correction of any underlying metabolic or anatomical abnormalities are also crucial factors to prevent stone recurrence and preserve kidney function in this specific population [5,13].

Acquiring evidence

To evaluate the current knowledge about the treatment of staghorn calculi in children, we searched PubMed for relevant articles published between 1991 and 2011, using a combination of related keywords, i.e. staghorn stone, child, kidney calculi, surgical treatment, ESWL, percutaneous nephrolithotomy (PCNL) and open surgery. Reports related to the treatment of paediatric stone disease in general (open surgery, PCNL, ESWL) were also sought by the same method. Additional references were obtained from the reference list of full-text reports.

Evidence synthesis

During the last three decades the management of urinary stone disease has developed dramatically, with the clinical introduction of PCNL and ESWL for treating this problem [14,15]. Minimally invasive methods have grown rapidly and have replaced open procedures in the management of urolithiasis at any age, with the first cases of paediatric PCNL and ESWL reported in 1984 and 1986, respectively [15,16].

Despite the widespread use of open surgery before the 1990s, throughout the world, currently minimally invasive methods have replaced this approach for removing staghorn calculi, even in children. In particular, while the recently updated European Guidelines (European Society of Paediatric Urology, ESPU) recommend PCNL as the first-line treatment in children with staghorn calculi, open surgery is recommended among the secondary options, with ESWL [17]. However, as in adults, the location, composition and size of the stone, anatomy of collecting system, and presence of obstruction and/or infection, are important factors to be considered in selecting the most appropriate method to be used.

Last but not least, the preservation of renal development and function, limitation to radiation exposure, and the need for re-treatment are particular considerations well before any surgery for stones in children. To select the most effective treatment in each child, the decision process must be individualised for the age of the patient, anatomy of the kidney and composition of stone burden.

PCNL for staghorn calculi in children

Since the first report of percutaneous stone surgery for upper-tract stones in adults in 1976 [14], experience has dramatically increased with the increasing application of PCNL in subsequent years. Nine years later, in 1985, the first paediatric series of PCNL was reported [16]. By contrast to the easy and widespread acceptance of this method among adults, the use and popularisation of PCNL were somewhat delayed in children because of some concerns about acute or long-term parenchymal damage, radiation exposure, and risk of major complications such as haemorrhage, intestinal trauma and sepsis [18]. However, with increased experience and more cases treated, several studies showed minimal scarring and no significant loss of renal function after PCNL; this convinced urologists of the safety and efficacy of the procedure [19]. In the study of Dawaba et al. [20], no renal scarring was reported in 65 patients on a long-term follow-up, indicating the long-term safety of this procedure in children.

Initial publications of paediatric PCNL focused on the complications of PCNL, where the use of 'adult-sized' instruments was the main concern as a potential cause of higher complication rates. Related to this, Desai et al. [21] found that intraoperative haemorrhage during PCNL in children was related to the calibre and number of tracts. Also, Zeren et al. [22] reported a significant association between the complications (particularly intraoperative bleeding) and sheath size, as well as operative time and stone burden.

Although PCNL was initially not rapidly accepted as a safe and effective management alternative for removing stones in children, after refinements in access techniques and instrumentation, and technological advances in energy sources for lithotripters, PCNL is now considered as a safe and efficient method compared with open surgery (even for staghorn stones) in children, either as monotherapy or as part of a combined approach with ESWL. Although the published data on PCNL for staghorn calculi in children are still limited, increasing experience of PCNL in children, along with accumulated experience of PCNL in adults, has contributed to the acceptability of this procedure for staghorn calculi. As a result, the high efficacy of PCNL in large renal calculi has promoted it to a first-line treatment for staghorn calculi in adults, and it is now recommended as the standard management for staghorn calculi in adults [5,17]. With the experience accumulated from adult patients, PCNL is now used successfully in children [23]. Indeed, it is now recommended as the primary treatment option in paediatric staghorn calculi in the guidelines of the ESPU and the AUA [5,17].

Published data on the use of primary PCNL monotherapy in staghorn stones report overall stone-free rates for different sized kidney stones to be 60–100% in populations of different ages [24–26]. Reported success rates for PCNL in children with staghorn stones are also within the same range, e.g. Aron et al. [27] found PCNL monotherapy to be highly effective, with stone-free rates approaching 90% in pre-school-aged children with staghorn calculi. Romanowsky et al. [28] reported complete clearance in eight of nine children (seven of whom had staghorn calculi) after a single stage of PCNL. In 2004, Desai et al. [21] published their results of PCNL for complex calculi (either staghorn or with a large bulk and involving more than one calyx, the upper ureter, or both) in children, and they reported that the complete clearance rate was 90% with PCNL monotherapy among 56 patients.

Even though it is well confirmed that PCNL is an effective method for removing staghorn stones in children, with certain significant advantages (e.g. direct visualisation of the fragmentation, clearance of the fragments under vision, minimising the need for recurrent visits and multiple/ancillary procedures [29]), the possible complications are still a major concern. Zeren et al. [22] reported a 24% incidence of haemorrhage requiring transfusion after 67 PCNL procedures in 55 children with renal stones. They found a close correlation between transfusion and operative time, stone burden and sheath size. Although much lower transfusion rates (< 5%) were reported in subsequent studies, these reports also indicate an association between both tract number and size and the need for transfusion [26,30]. Other commonly reported notable complications include transient fever and urine leaks [26]. The most serious complication of PCNL in children with staghorn calculi (struvite) is sepsis, which can result in death [31].

ESWL for staghorn calculi in children

Although the application of ESWL in children was first reported in 1986 [15], ESWL monotherapy for managing staghorn calculi in children was first reported in 1999 by Orsola et al. [32] where 11 of 15 children were rendered stone-free. In recently published studies the reported stone-free rates after ESWL increased to 95% for infants and 71–87.5% for children with staghorn calculi [33,34].

When considering ESWL for kidney stones in children, unlike in PCNL, it appears to be a different condition from that in adults, and recommendations for children cannot be directly inferred from those for adults. This is mainly because staghorn calculi in children represent a smaller stone burden, and that the small body volume of children facilitates better shock-wave transmission, both resulting in superior stone fragmentation and clearance rates [32,34]. In addition, the shorter duration of the disease, greater stone fragility and lower impedance to shock waves might be the possible reasons for better stone fragmentation. Furthermore, these stones are often present in undilated collecting systems, occupying a relatively smaller volume even for complete staghorn calculi [32,34]. Therefore, unlike in adults, ESWL monotherapy has the potential to be accepted as a suitable treatment option in children with staghorn calculi.

Although ESWL has been shown to be effective and safe in the short term in large series of children [35–37], the long-term safety in children is still debated. However, despite the ongoing theoretical discussions about the long-term safety and effects of ESWL on the function of growing kidneys, no evidence of renal scarring, change in blood pressure or renal functional loss after ESWL has been shown in several studies [37–43]. In fact, this issue has been well supported by recently updated European Guidelines (ESPU) where they stated that 'many reports confirm that shock-wave lithotripsy can be performed in children, with no suspicion of long-term morbidity of the kidney' [17].

However, for the acute/subacute period of ESWL, it is known that ESWL is a well-tolerated procedure with minimal morbidity. Minor complications such as bruising, ecchymosis and renal colic are reported in 11–50% of cases. Studies focusing on the management of large stone burdens in children have reported the rate of 'steinstrasse formation' to be 1.9–5.4% [42,44]. While the incidence of haematuria (40%) is less than that in adults, ureteric obstruction or sepsis requiring stenting or percutaneous drainage might occur in such cases, and the need for ancillary procedures is directly proportional to the size of the stone treated particularly when the reported complications of PCNL are considered, together with the fact that ESWL has the potential to give an 80% success rate for staghorn calculi in several studies [45], it might appear to be logical to prefer ESWL as a less-invasive treatment alternative at least in some selected cases. However, some factors, e.g. the need for several sessions (mostly under anaesthesia), prolonged clearance time, risk of residual fragments (of greater relevance in struvite stones) and obstruction resulting in colic attacks, constitute major limitations of ESWL in children with staghorn calculi. Moreover, higher rates of re-admission and re-treatment, with an increased need for auxiliary procedures, are other critical points to be considered. For example, although a success rate of 79% was reported by Al-Busaidy et al. [45], where 42 children (9 months to 12 years old) with staghorn calculi were treated with ESWL, they also reported a significant number of ancillary procedures. In the light of all these concerns, with its highly effective and safe results, currently PCNL is a valuable alternative in the management of children with staghorn calculi. Table 1 compares the reports on PCNL and ESWL for staghorn stones in children.

Table 1 Comparison of ESWL and PCNL results for staghorn calculi in children.

References (no. of cases)	Stone/patient characteristics (*n*)	% Complete clearance
ESWL		
[32] (15)	Age 14 months to 13 years	73.3
[34] (23)	Age 5.5 months to 2 years (16)	87.5
	6–11 years (7)	71.4
[45] (42)	Age 9 months to 12 years	79
PCNL		
[21] (56)	Complex calculi, either staghorn (complete or partial) or those with a large bulk and involving more than one calyx	89.8
[23] (12)	A staghorn calculus, defined as a branched stone occupying more than one part of the collecting system	91.6
[24] (31)		67.7
[27] (19)	Pre-school children	89

Despite acceptable success rates and with high stone-free rates, ESWL for staghorn calculi can require several sessions, higher ancillary procedures and ureteric stenting. As to the use of stents, in a study of ESWL in children with staghorn calculi it was reported that there was no difference in stone-free rates for stented and unstented groups, but complication rates were significantly higher in the latter group [45]. Thus, stenting patients with staghorn calculi before ESWL provides a greater margin of safety, decreases the incidence of ureteric obstruction and shortens the hospital stay [45].

The use of ultrasonography and digital fluoroscopy has significantly decreased the radiation exposure during ESWL sessions, and it has been shown that children are exposed to significantly lower doses of radiation than are adults [37,44]. The type of anaesthesia should be general or dissociative for children aged <10 years, whereas conventional intravenous sedation or patient-controlled analgesia is an option for relatively older cooperative children [46].

Knowing that the vast majority of staghorn calculi are composed of struvite, residual fragments after ESWL, particularly in children, constitute a major risk for infection and recurrences, and that should be accepted as a major disadvantage of ESWL. The cumulative risk of stone recurrence is higher in children than in adults. Afshar et al. [9] reported that 34.5% of fragments grew at a mean follow-up of 48 months, and a similar number of patients developed clinically significant symptoms. Nijman et al. [47] also reported that

33% of children with small fragments had evidence of calculus growth at 24 months.

Open surgery for staghorn calculi in children

Although open surgery is currently known to be indicated only in a few selected cases with staghorn calculi, its reported incidence could be as high as 14% in developing countries [48]. Good candidates for open stone surgery include very young children with large stones and/or a congenitally obstructed system which also requires surgical correction, and children with severe orthopaedic deformities that might limit positioning for endoscopic procedures.

Although it is reported that open stone surgery has some favourable aspects in children when compared to adults, such as a rapid healing and with lower complication rates [48], it is still difficult to perform (removal of the staghorn calculi often necessitates several as well as extended nephrotomies) and difficult to repeat in case of recurrence. More importantly, it can compromise renal function in the developing kidneys of these special cases. In the study by Gough and Baillie [49] there was a significant decrease in renal function after anatrophic nephrolithotomy in five of nine children with staghorn stones, with a blood transfusion needed in three of these cases. However, Assimos et al. [50] reported successful paediatric anatrophic nephrolithotomies in 10 of 11 patients, with minimal morbidity but with a high recurrence rate.

Conclusion

Despite the evident and dramatic changes in stone management in the last two decades, the management of staghorn calculi in children constitutes a major challenge to urologists. The aims of management in these cases should be the complete clearance of stones, preservation of renal function and prevention of future recurrences. Although open surgery was widely used in the past for treating such stones in children, currently it has only limited indications in highly selected cases. However, following significant improvements in endoscopic technology, and the application of these techniques after accumulated experience from adult cases, PCNL has become a safe and effective accepted primary treatment in children. ESWL could be considered as the second effective option for the minimally invasive management of staghorn calculi, with its significantly successful and safe results, at the expense of higher retreatment rates, additional procedures, likelihood of residual fragments and longer time to stone-free status. To select the most appropriate method among these alternatives, each case should be evaluated well and individualised for the factors related to the patient, stone and body habitus.

Conflict of interest

No conflict of interest to declare.

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