



Review

# Management of staghorn stones in special situations



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Received 27 March 2019; received in revised form 5 August 2019; accepted 18 October 2019  
Available online 30 December 2019

## KEYWORDS

Staghorn stone;  
Horseshoe kidney;  
Ectopic kidney;  
Paediatric kidney;  
Solitary kidney;  
Management

**Abstract** Staghorn stones have always been a challenge for urologists, especially in some special situations, such as horseshoe kidney, ectopic kidney, paediatric kidney, and solitary kidney. The treatment of these staghorn stones must be aggressive because they can lead to renal function loss and serious complications. The gold-standard management for staghorn stones is surgical treatment with the aim of clearing the stones and preserving renal function. Treatment methods for staghorn stones have developed rapidly, such as extracorporeal shock wave lithotripsy, retrograde intrarenal surgery, percutaneous nephrolithotomy and laparoscopy and open surgery. Whether the standard procedures for staghorn stones can also apply to these stones in special situations is still not agreed upon. The decision should be made individually according to the circumstances of the patient. In this review, we evaluate the previous studies and comments on the management of staghorn stones under special situations in the hope of guiding the optimal choice for urologists.

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## 1. Introduction

According to the definition in Campbell's urology and related literature, staghorn stones are defined as large and branched stones that occupy part or all of the

collecting system. They can be partial or complete, depending on the level of occupation of the renal pelvis and renal calyces. That is, staghorn stones occupy the renal pelvis as well as at least two calyces of the kidney [1,2].

The ideal treatment for staghorn calculi involves removing the whole stone by operation, improving the metabolic abnormalities and dealing with anatomic variation. Conservative treatment of staghorn stones has a tight relationship with renal loss and urosepsis, with an approximate mortality rate of 30% [3–5]. Therefore,

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Peer review under responsibility of Second Military Medical University.

staghorn stones should be managed actively by surgical treatment [6,7]. Traditional treatment options for staghorn stones include extracorporeal shock wave lithotripsy (ESWL), retrograde intrarenal surgery (RIRS), percutaneous nephrolithotomy (PCNL), laparoscopy and open surgery [8]. For its high rate of stone clearance and low incidence of complications, PCNL has been recommended as the most suitable operation method for staghorn stone by the American Urological Association (AUA) since 2005 [9]. Mini-PCNL is becoming a prevalent way to manage staghorn stones around the world, and it can reduce the complication rate by using a smaller tract [10,11].

Surgical approaches for the management of staghorn stones in special situations are still confusing. Common special situations include horseshoe kidney, ectopic kidney (cross-fused ectopic kidney and pelvic ectopic kidney), paediatric kidney and solitary kidney. In current guidelines, there is no clear suggestion or preferred treatment modality regarding stone treatment in these situations.

Due to the abnormal anatomy and the overburdened stone, these situations are even more of a challenge for urologists. The most appropriate treatment should be chosen according to the patient's specific conditions so that the patient can benefit the most. In this article, we review the experience in handling staghorn stones under the above circumstances.

## 2. Acquiring evidence

A literature review was performed by using PubMed. The following terms and combinations of terms were searched in English-language publications: "staghorn stone" or "staghorn calculi" combined with the terms "horseshoe kidney" or "fused" or "pelvic ectopic kidney" or "paediatric kidney" or "solitary kidney". This search returned 182 articles in total, and we excluded those articles that were not appropriate for our review by reading the title, abstract, and full text. In addition, reports related to the treatment of stone disease in these special situations were sought by the same method. Additional publications were obtained from the reference lists of full-text reports.

## 3. Evidence synthesis

### 3.1. Staghorn stones in horseshoe kidney (HK)

HK, as the most common congenital malformation, is one of the renal fusion anomalies. It is thought to be induced by inadequate head displacement and poor renal rotation due to the clamping of the lower mesenteric artery under the isthmus in the early stages of pregnancy [12]. It was reported to have a prevalence of one in 400–800 in the normal population [13]. Due to malrotation of the kidney, the ureter, which links with the renal pelvis, is forced superiorly and laterally. One-third of cases of HK are associated with ureteropelvic obstruction. Patients with HK are very susceptible to urinary tract infection, renal calculi, and obstruction. Kidney stone is the most common

complication in HK, and the incidence of stone disease in HK is approximately 20% [14]. This may be due to stasis and infection caused by impaired drainage of the urinary tract, which results in stone formation. The management of staghorn stones in HK is challenging because of the variable vascular and anatomic abnormality [15]. The abnormal position of the renal pelvis also increases the complexity of stone treatment. HK is often accompanied by ureteropelvic junction obstruction and abnormal orientation of the calyces, which prevent the passage of the stones.

The treatment of staghorn stones in HK should follow the same guiding principles as for staghorn stones in normal kidney stones. PCNL is suggested as the first-line treatment method for staghorn stones in patients with HK [16]. The usefulness of a percutaneous method in the management of kidney stones in HK has been reported in several studies [17–21]. In 1973, Fletcher and Kettlewell [22] reported the first PCNL in HK. From then on, percutaneous puncture of the HK has been found to be relatively safe because of favourable calyceal orientation and vascularity [23]. PCNL showed a better stone-free rate (SFR) than ESWL, and it was found that the risk of arterial bleeding did not increase in HKs compared with normal kidneys [17,23,24]. A higher success rate with minimal complications has been observed in PCNL with HK in numerous studies [17,18,25,26]. The SFR of the primary PCNL procedure in HK with stones ranges from 81% to 87% [14,16,27,28]. Most studies have been focused on the management of large stones in HK, which include only a few cases of staghorn stones [14,27,28]. Raj and his colleagues [14] managed three staghorn calculi among 37 cases of HK stones, and the SFR reached 87%. Most procedures in the study were performed with both rigid and flexible nephroscopes through a single access. Mosaxi-Bahar and his colleagues [28] managed two staghorn stones in HK using both PCNL and SWL, and multiple access tracts were needed for the complete stone clearance. Liatsikos et al. [16] managed 17 staghorn calculi in HK patients with PCNL, and the SFR was 82%, and mean hospital stay were 4.4 days. The primary and secondary complications rates were 20% and 46.6%, respectively. Ding et al. [29] reported that in the management of staghorn stones in HK, PCNL combined with antegrade flexible ureteroscopy (FURS) is a safe and effective way to significantly reduce blood loss compared with PCNL alone.

How to do PCNL in HK with staghorn stones remains a challenge. Percutaneous puncture in HK is relatively safe. Usually, it is best to go through the superior poles. Janetschek and Kuanzel [24] pointed out that the puncture should be made below the 12th rib on the posterior axillary line with caudad angle of puncture. The risk of major blood vessel bleeding associated with access is no higher than that of normal kidneys. Symons et al. [30] performed PCNL in six patients with staghorn calculi in HK. They performed upper pole access in half of the cases, while the remaining cases needed multiple approaches. The major complication rate was 3%. Miller and co-workers [31] reported a primary SFR of 84.1% in 35 patients with HKs, and 97.7% of the cases were handled with only one access, including 82.2% of the kidneys accessed by the upper pole. Gupta et al. [32] treated 31 kidney stone patients with HK and observed that the medial tracts were more difficult to

dilate, and the tracts were usually longer. However, HK-related colon injury can be avoided by puncturing more medially because the colon has a more posterolateral relationship with the kidney. Recently, a multi-institution study reported their experience with the use of various techniques of PCNL [16]. The SFR of primary PCNL was 82%. The upper pole access was used in all cases but one. The approach was feasible in most cases because of the lower position of the HK. They proposed angular access in a previous study with a single incision, which achieved a SFR of 85% [33], and this subcostal triangulation technique also proved to be effective in HKs with staghorn stones. The management of large stones in HKs requires an unusual long tract for approaching the medial and lower calices [14]. The flexible nephroscope can provide better manoeuvrability in medial and lower calices. The upper pole access facilitates the entry of the instrument into all anatomic sites of the collecting system [14]. No studies have reported the use of ESWL and RIRS in the management of staghorn stones in HK.

In HK, PCNL seems to be the most suitable surgical management technique for staghorn stones. It can achieve a relatively higher SFR, and some techniques, such as upper puncture and multiple accesses, are necessary.

## 3.2. Staghorn stones in ectopic kidney

### 3.2.1. Staghorn stones in cross-fused ectopic kidney

Crossed fused ectopic kidney refers to one kidney being shifted abnormally from one side to the other side and the two sides fusing together during the period of growth [34]. The disease is an uncommon congenital deformity. Some literature reports that its incidence is approximately 0.1% of live births and 1 in 7 500 in autopsy series [35,36]. Like HK, crossed fused kidney is a common renal fusion abnormality in the urinary system, which has a predominance in males over females (3:2) and right over left (3:1) [35]. Because it is related to recurrent urinary tract infections, stone formation, hydronephrosis and abdominal mass, these malformations are clinically important and should be paid much attention to [37–39]. There are six main types of crossed fused kidneys. However, there are no guidelines for the treatment of staghorn stones in crossed fused kidney.

The formation of staghorn calculi within crossed fused kidneys has been reported in few studies. Stubbs and Resnick [40] described two cases of crossed fused renal ectopia with struvite staghorn calculus. Both patients had a successful outcome from the treatment of nephrolithotomy. All the stones were successfully removed. So far, no one has relapsed from stone disease.

Amin et al. [41] reported that a young man was hospitalized with haemorrhagic urine and right abdominal pain. Intravenous pyelography imaging shows the crossed fused ectopic kidney with staghorn stones in the upper part. After excluding other congenital abnormalities, patients received conservative treatment. At present, the patient is asymptomatic with no further consultation [41]. A woman of 68 years was reported to have L-shaped crossed renal fusion (left to right) with staghorn stone. She underwent supine PCNL in three stages and three renal tracts. Three-

dimensional reconstruction by computerized tomography urography, correct preoperative preparation, suitable positioning, an ultrasound-guided approach, traction in operation, and flexible nephroscopy may completely remove the stones in renal abnormalities [42].

Decisions for staghorn stones in crossed fused ectopic kidneys must be managed individually. Image examinations such as urography, suitable positioning, an ultrasound-guided approach and traction in operation, will increase the SFR.

### 3.2.2. Staghorn stones in ectopic pelvic kidney

Pelvic kidney is a rare congenital anomaly with an incidence between 1/2 200 and 1/3 000 [43]. The ectopic pelvic kidney is more susceptible to developing nephrolithiasis because of the position of the renal pelvis, ureteral insertion, and kidney malrotation, which can lead to urine stasis [44]. The treatment methods for the stones in pelvic kidneys include ESWL, PCNL, RIRS, laparoscopy and open surgery. However, choosing the best modality for the treatment of staghorn stones in ectopic pelvic kidney is still confusing [45].

Articles related to staghorn stones in ectopic pelvic kidney are very limited. Theiss et al. [46] reported that ESWL is an effective method in stone fragmentation, but abnormal drainage of pelvic kidney reduces SFR and increases residual fragments. For RIRS, it is difficult for the ureteroscopy to pass the tortuous ureter. Eshghi et al. [47] described laparoscopy-assisted PCNL for the treatment of stones in a pelvic kidney, and this modality had the advantages of enhancing safe puncture and correct tract placement [44]. In recent years, successful laparoscopic pyelolithotomy (LPL) for the management of renal stones has advanced rapidly, and compared with PCNL, its risks of bleeding and nephron injury are lower, and the SFR of LPL is very high [48]. It is worth mentioning that LPL is an appropriate approach for kidneys with anteriorly or laterally positioned extrarenal pelvis [49] and Soltani et al. [50] reported a patient with staghorn stones in ectopic pelvic kidney, which was managed with LPL and they concluded LPL is a safe and effective treatment modality and can be proposed as the first line treatment for such situation.

For ectopic pelvic kidney with staghorn stones, the therapeutic opinion varies from person to person, and laparoscopic pyelolithotomy is feasible.

## 3.3. Staghorn stones in paediatric kidney

Stones in children is a common problem [51]. The incidence is approximately 1%–5% in developed countries and 5%–15% in developing countries [52,53]. The risk factors of stone formation in children include anatomical defects, metabolic, urinary tract infections, and especially relevant genetic factors [54]. Staghorn stones in children were once reported to account for approximately 19% of the specific population [55]. However, the management of this type of stone is also a challenge for urologists.

### 3.3.1. PCNL for staghorn stones in paediatric kidney

PCNL is suggested as the first choice for the management of staghorn stones in adult patients due to its high efficacy [9],

and also for children. The small kidney often has a highly vascular nature and variable vascular anatomy, and the use of adult-sized instruments often brings potential complications, such as haemorrhage and intestinal trauma. However, with advances in instruments and improvements in techniques, PCNL has become safer and more effective. The advantages of PCNL for staghorn stones in children include a higher SFR and lower requirement for ancillary procedures; thus, it could reduce the need for multiple procedures [56]. Many studies have shown that PCNL is an effective and safe method in the management of paediatric kidney stones, and it is also recommended by the AUA as the primary option for the treatment of paediatric staghorn calculi [9].

The success rate of PCNL for staghorn stones ranges from 60% to 100% in different age groups [57–59]. Aron et al. [60] reported a complete SFR of 89% in children with staghorn stones using PCNL. Horuz and Sarica [61] found that for preschool children with staghorn stones, PCNL monotherapy was highly effective, and the SFR reached 90%. Kumar et al. [62] reported an SFR of 92% in 12 paediatric patients with staghorn stones. Desai et al. [63] reported that the total clearance rate of PCNL single treatment was 90% in 56 paediatric patients, and the clearance rate increased to 96% after adding SWL. Romanowsky et al. [64] reported a complete SFR in eight of nine children (seven had staghorn calculi) without major complications or blood transfusion after a single stage of PCNL.

The complications after PCNL for the management of paediatric staghorn stones are also a focus. Studies have found that the size and number of puncture channels are significantly correlated with intraoperative bleeding and postoperative decreased haemoglobin in children with PCNL. The collection system is more compact in children because of the small kidney size. Therefore, using instruments with minimal trauma and minimal renal access may reduce the likelihood of major complications. El-Nahas et al. [65] reported a complication rate of 28.6% in 28 children with staghorn stones after PCNL. Guven et al. [66] concluded that complication rates seen in children were comparable to those in adults. Earlier, the authors reported that dilation of up to 26 Fr did not result in significant morbidity in children, and it has been shown in animal models that the use of small channels alone has no advantage over models with renal scarring. Desai et al. [63] reported that intraoperative haemorrhage in children was associated with PCNL tracts. Later studies also showed a link between transfusion and tract numbers and size [59,67].

Because the paediatric kidney is small, PCNL will be challenging for the management of staghorn stones. Due to the high clearance rate and the need to avoid multiple operations and hospitalizations, PCNL has become increasingly popular in the management of staghorn stones in children [56,68]. PCNL in 3-month-old children was reported to be safe and effective [69]. Adult-size instruments were not suitable for children [59]. In recent years, the improvement of endoscopes in urology has made great progress. Currently, minimally invasive PCNL (mini-PCNL) is used for clinical applications. In 1998, Jackman et al. [70] developed the mini-PCNL device and applied it to children. Mini-PCNL is now proven to be an effective method for

paediatric staghorn stones. The utilization of smaller instruments for PCNL (mini-PCNL) may limit the risk of haemorrhage in this population [71–73]. Rashid et al. [74] used mini-PCNL for the management of complex staghorn stones in children, and they found it was safe and efficient, with an SFR of 78%, which reached 89% after a few ancillary treatments, and the complication rate was only 14%. Bleeding complications are associated with tract diameter, and mini-PCNL seems to be the best method in children [58,75].

### 3.3.2. ESWL for staghorn stones in paediatric kidney

The advent of ESWL in 1978 brought dramatic changes in urolithiasis treatment. Eight years later, Newman et al. [76] recorded the efficacy of ESWL for the first time in children. In 1999, Orsola et al. [77] recorded the efficacy of ESWL for staghorn calculi. They treated 15 children with the Lithostar Ultra, and the SFR reached 73%. Later, Lottmann et al. [78] used the Sonolith 3000 to describe the efficacy of ESWL in the treatment of 20 children with staghorn stones. Another study reported seven child cases of ESWL treatment, and the staghorn stones were cleared successfully after 2 months without complications [79].

ESWL has proven effectiveness and safety in children [80–82]. However, growing kidneys were more susceptible to the influence of shock waves. Several studies reported no complications of renal scarring or kidney damage after ESWL for children with staghorn stones [83–86]. Complications were reported in 11%–50% of cases, such as bruises, blood stasis and renal colic. The incidence of steinstrasse formation was 1.9%–5.4% in the management of large stones in children. The incidence of haematuria was lower than that in adults, and ureteral stenosis or sepsis may occur, which requires stenting or percutaneous drainage [86,87].

The staghorn stones of children have smaller volume loads, thus leading to smaller stone fragments and higher removal efficiency. The shorter course, the less brittle stones, and the lower impedance to the shock wave might explain the better stone fragmentation. Thus, unlike in adults, ESWL alone has the potential to be accepted as an appropriate method in children.

### 3.3.3. Open and laparoscopy for staghorn stones in paediatric kidney

Though open surgery is only reported in a few specific cases of staghorn stones, it is reported that the application rate can be up to 14% in some developing countries. The indications include children with large and complex stones or congenital malformations, which also require surgical correction, as well as those with severe orthopaedic malformations, which may limit the application of endoscopic surgery. Open surgery was reported to be beneficial for children due to fast healing, fewer complications, and the advantage of avoiding multiple procedures [88]. Jurkiewicz et al. [89] also used the combination of pyelolithotomy and endoscopy to manage the staghorn stones in children and achieved a completely stone clearance without causing parenchymal damage in one single procedure. They concluded that this method is safe and efficient, and does not require blood transfusion.

There are still some difficulties in performing open surgery for staghorn stones. Open surgery usually requires



several extended nephrectomies when there is difficulty in repeating operations in recurrent cases. More importantly, it can harm the renal development of children. Gough and Baillie [90] reported apparent renal function loss in five of nine children with staghorn stones who underwent nephrectomy after open surgery. However, Assimos and his colleagues [91] reported successful anatomic nephrolithotomy in 10 children with minimal postoperative morbidity.

Similarly, very few studies have reported laparoscopic lithotomy for the management of staghorn stones in children. Most of these studies focussed on the procedures and not the stone size. However, researchers have proven that it is a safe and feasible method for the management of paediatric renal calculi, even not for staghorn stones. It can be an alternative to ESWL and PCNL when these are not feasible or possible.

In summary, we suggest that the aim for treatment of paediatric staghorn stones should be completely removing the stones, retaining kidney function and preventing future recurrence. Open surgery has been widely used to treat staghorn stones in children in the past, but the indications are limited to highly selected cases. With the substantial improvement in endoscopic techniques and the experience in adult cases, PCNL has come to be regarded as a safe and effective primary treatment for children. In particular, the utilization of mini-PCNL has become increasingly popular, as it may limit the risk of haemorrhage. ESWL can be used as the second effective treatment option for minimally invasive treatment of paediatric staghorn stones. Though with higher retreatment rates, additional operations, the possibility of residual stone and more time to become complete stone-free, overall it is significantly successful and safe.

### 3.4. Staghorn stones in solitary kidney

The European Association of Urology (EAU) guidelines point out that solitary kidney patients are at high risk of stones [92,93]. Solitary kidney and staghorn stones are both very difficult cases for urological surgeons. Untreated staghorn stones may destroy the entire kidney and may cause life-threatening sepsis, so eliminating the calculus completely and removing the obstruction is the key to the treatment. Solitary kidney patients with kidney stones have higher susceptibility to the risk factors of kidney disease, even including small complications, and if the treatment is not good, renal function can be obviously damaged, and the patients' quality of life will be reduced. For a solitary kidney patient, uncontrollable renal haemorrhage may be life-threatening.

#### 3.4.1. PCNL for staghorn stones in solitary kidney

The AUA guidelines recommend PCNL as the preferred treatment for staghorn stones based on the high SFRs and acceptable rate of complications. With the improvement in the technique and increase in the experience of urologists, PCNL can now remove staghorn stones from certain patients with solitary kidney [94]. Postoperative complications in solitary kidney stones are the most common problems in urological surgery, while staghorn stones are the most difficult to

treat by PCNL. When these two factors are combined, it becomes one of the most complex and extremely high-risk clinical challenges in urological operation [95].

With the tremendous improvements both in equipment and in technique, PCNL has proven safer than open surgery. Most importantly, PCNL resulted in an improvement of kidney function, while kidney function worsened after open surgery [96]. In a large-scale study, patients with solitary kidney stones were more likely to have bleeding (10.2% vs. 5.7%) than those with double-sided kidney stones, and the likelihood of blood transfusions was higher (10.1% vs. 5.6%) [94]. Most of the postoperative bleeding in PCNL was due to the large number of puncture channels and the inability of the kidneys to self-repair. The flow of blood around the renal collection system accounts for 20% of the cardiac output; the blood vessels (including the arteries and veins) around the renal collection system are the most important structures; and the solitary kidneys have a greater percentage of their blood come from the collection system due to the increased compensation. Therefore, it is necessary to avoid the injury of these blood vessels during the puncture [97]. Resorlu et al. [94] reported their experience of PCNL for staghorn stones in solitary kidney, and their 1-year follow-up showed that patients had significant improvement of renal function. Multi-channel PCNL was thought to come with more complications, which could be particularly important factors for solitary kidney patients. Liu et al. [98] used minimally invasive PCNL (MPCNL) to treat 105 solitary kidneys with staghorn kidney stones, and the results showed that the single-channel MPCNL complication rate was lower, while the SFR was comparable to that of multi-channel PCNL. Haberal et al. [99] achieved a SFR of 73% in 91 patients (14 staghorn stones) with solitary kidney by PCNL. The presence of a staghorn stone was found to have a negative impact on success.

#### 3.4.2. RIRS for staghorn stones in solitary kidney

In recent years, with the rapid growth of minimally invasive methods, increasing numbers of urologists have tried to use RIRS to treat large kidney stones, but there are few reports about RIRS alone in the management of solitary kidney with staghorn stones. The effect of RIRS on the management of large solitary-kidney calculi (>2 cm) was compared with that of PCNL. The results showed that the number of operations for PCNL was lower, the efficacy quotient (EQ) was higher, and the total cost was comparable to that of RIRS, so they recommended PCNL as the first choice for large solitary kidney stones. For patients with special diseases, such as those requiring anti-coagulation, RIRS is a good choice for urologists because of its safety and acceptable SFR [100].

At present, the clinical application of RIRS to solitary kidney is mainly focused on subsequent treatment after PCNL. The combined treatment by the retrograde and antegrade approaches can obviously improve the SFR and reduce the damage to renal function. Xu et al. [101] reported on their application of single-channel MPCNL combined with flexible ureteroscopy to treat 20 cases of solitary kidney staghorn calculi. The stone-clearing rate was 83.3%, and the follow-up mean serum creatinine value decreased from  $1.7 \pm 0.5$  mg/dL to  $1.3 \pm 0.4$  mg/dL ( $p < 0.05$ ). In a study reported by Atis and his colleagues [102], 15 RIRS-treated single-kidney stones showed a fairly

high stone removal effect, with an initial SFR of 83.3% and a final SFR of 95.8%.

### 3.4.3. ESWL for staghorn stones in solitary kidney

As with RIRS, there are few reports of ESWL alone for treating solitary kidney staghorn stones. Among the limited studies, the conclusions are similar: ESWL for staghorn stones in a solitary kidney is troublesome [103,104]. The main reason may be that ESWL is not very effective in the treatment of staghorn stones, the SFR is low, and the patient often needs multiple rounds of treatment. The easy-to-form steinstrasse leads to ureteral obstruction, especially after repeated or high-energy lithotripsy, which is more prone to renal atrophy. It is often not acceptable to solitary kidney patients.

Strem and Geisinger [105] combined PCNL with ESWL in the management of solitary kidney staghorn calculi with the aim of first removing as many stones as they could by PCNL, followed by the treatment of residual stones by ESWL, and a final PCNL examination and removal of residual stone fragments (*i.e.*, PCNL + SWL + PCNL sandwich therapy). The results indicated that the outcome of the combined therapy was comparable to that of separate surgical treatments, and postoperative kidney function was stabilized for a long period of time.

Above all, the postoperative complications of PCNL in solitary kidney stones are the most common problems in urological surgery, and uncontrollable renal haemorrhage may be life-threatening. With the tremendous improvement in both equipment and technique, PCNL has demonstrated significantly better safety results and comparable effectiveness compared with open surgery and the long-term stone recurrence rate is lower as well. Combined treatment by RIRS and PCNL can obviously improve the SFR and reduce the damage to renal function.

## 4. Conclusion

The management of staghorn stones in special situations should follow the principles above, and urologists should make decisions according to the patient's individual situation.

## Author contributions

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*Drafting of manuscript:* Xiaofeng Gao, Ziyu Fang.

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## Conflict of interest

The authors declare no conflict of interest.

## Acknowledgement

This work was financed by a grant from the National Natural Science Foundation of China (No. 81800624).

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