Percutaneous Nephrolithotomy (PCNL) in Kidneys with Fusion and Rotation Anomalies

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Objectives: *Fusion and rotation anomalies are among the most common anomalies of the kidneys especially the horseshoe kidneys that have abnormal mixtures of malrotation, ectopia and fusion. Percutaneous nephrolithotomy (PCNL) in horseshoe kidney and malrotation kidney is challenging. We report PCNL in 10 horseshoe kidneys and 4 kidneys with congenital malrotation.*

Material and Method*: PCNL was done in 14 kidneys of 12 patients who had horseshoe kidneys (6 patients) and malrotation kidneys (4 patients). Two patients with horseshoe kidney had bilateral calculi. One patient had simultaneous PCNL. Upper pole access was done in 12 kidneys under fluoroscopic guidance with modified PCNL technique. Tubeless PCNL was done in 7 kidneys.*

Results: *Stone-free rate was 86% and the fragment of stones less than 4 mm was 14%. Only minimal pleural effusion was found in one patient that no intervention was needed. Average hospital stay was 3.3 days.*

Conclusion*: Percutaneous nephrolithotomy (PCNL) in horseshoe kidney and malrotation kidney is effective and a safe method in wish only a minor modified technique is needed. Upper pole access was used in most of the cases. No increasing of the complications of PCNL in horseshoe kidneys and malrotion kidneys is found comparing to the same procedure done in the normal kidney.*

Keywords: *Fusion and rotation anomalies, Percutaneous nephrolithotomy(PCNL)*

J Med Assoc Thai 2005; 88 (10): 1426-9
Full text. e-Journal: *http://www.medassocthai.org/journal*

Percutaneous nephrolithotomy is accepted as the treatment of choice for the large renal and upper ureteral stone[1]. Fusion and malrotation are among the most common anomalies of the kidney especially the horseshoe kidney which usually presents with calculi. Percutaneous nephrolithotomy in horseshoe kidney is challenging due to the mixtures of malrotation, ectopia and fusion of the lower pole[2]. Minor modifications of percutaneous nephrolithotomy in horseshoe kidney are required. Report of the technique and the outcome of percutaneous nephrolithotomy in the treatment of the calculi in malrotated and horseshoe kidney will be described.

Material and Method

The study group consisting of 8 horseshoe kidneys with 10 renal units and 4 malrotated kidneys were treated with percutaneous nephrolithotomy. Mean age of the patients was 44.6 years old (range from 20 to 61 years old). Gender and lateralization were the same (6 male, 6 female, 7 right side, 7 left side). Three patients had a history of previous open nephrolithotomy. Mean stone size was 2.82 cm (range from 1.5 to 4.6 cm). The positions of the stones were 8 pelvic stones, 2 partial staghorn stones, 3 pelvic stones with lower caliceal stone and 1 lower caliceal stone. Simultaneous percutaneous nephrolithotomy was done in 1 patient (Table 1).

After the induction of the general anesthesia, a 6 F open-ended ureteral catheter was placed in the upper ureter. The access tract in the horseshoe kidney and malrotated kidney was more vertical and lower than the access in a normal kidney. Percutaneous access was obtained via a single tract through the upper pole posterior calix in 12 (86%), a middle pole posterior calix in 2 (14%) .The upper pole access was done in all cases of horseshoe kidney. All access tracts were subcostal. Dilation was done with fascial dilators up to 30 F with Amplatz sheath in place. The 24 F nephroscope with

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ultrasonic lithotripsy and pneumatic lithotripsy for the disintegration of the stone was introduced. After completing the percutaneous procedure and stone fragments removal, 20 F nephrostomy tube was placed in 7 cases, and 7 cases had tubeless PCNL. Nephrostomy tubes were removed 48 hours after the surgery. In addition, chest x-ray films were performed in all patients to rule out pneumothorax or hydrothorax. Plain KUB and nephrostogram were done in all cases before nephrostomy tube removal.

Results

The stone free rate of percutaneous nephrolithotomy in malrotated and horseshoe kidney was 86% (12 in 14 cases). Two (14%) retained small fragments of stone (less than 4 millimeter). Average hospital stay was 3.3 days (range from 3 to 4). No major complication was found. Minimal hydrothorax was found in one patient who had upper pole access and could be managed with conservative treatment (Table 2).

Discussion

Fusion and rotation anomalies of the kidney are among the most common urological anomalies in humans especially the horseshoe kidney. The first diseased horseshoe kidney was reported by Morgagni in 1820. The incidence is 1 to 4 in 1000 cases with a male to female ratio of 2 to 3:1. The horseshoe kidney is an anomaly of the mixture of malrotation and ectopia with fusion of the lower pole of the kidney[2]. Failure of ascent and rotation is due to the fusion of the metanephric blastema which is seen in the isthmus that contains connective tissue[3]. The isthmus contains parenchyma it can explained by abnormal migration of cells of the posterior nephrogenic area. Fusion of the lower poles prevents normal ascent and causes malrotation with anterior displacement of the collecting system. Associated anomalies with the horseshoe kidney are anomalies of skeletal system, cardiovascular system (ventricular septal defect), central nervous system (neural tube defect) and anorectal malformation.

The mixtures of malrotation and ectopia with fusion of the lower pole in a horseshoe kidney cause abnormalities of the arteries, collecting systems[5] and also the position of the horseshoe kidney that is lower than the normal kidney. Horseshoe kidneys usually have multiple renal arteries and aberrant vessels with the abnormality of the insertion of the ureter. Three types of arteries in a horseshoe kidney are normal renal arteries; accessory arteries that originating from the distal aorta, bifurcation of aorta, common iliac artery and aberrant arteries that originating from therenal artery, aorta, bifurcation of aorta, middle sacral and the common iliac artery. The caliber of the artery in a horse-

Table 1. Patient profile

Kidneys:	Horseshoe kidney (patient)	10 (8)
	Malrotation	4
Gender:	M:F	6:6
Laterality:	R:L	7:7
Previous surgery		4
Stone size (cm)		2.82 (1.5-4.6)
Position	: Partial staghorn	2
	: Pelvic stone	8
	: Pelvic stone + lower caliceal stone	3
	: Lower caliceal stone	1

Table 2. Methods and results

Access	: Upper pole	12
	: Middle calix	2
Hospital stay (day)		3.3 (3-4)
Stone free (%)		12 (86%)
Stone fragment < 4 mm		2 (14%)
Tubeless PCNL		7
Complication	: Hydrothorax[a]	1

(a) minimal hydrothorax in whish no intervention was needed shoe kidney is always smaller than the artery in a normal kidney and no vessel is found on the dorsal aspect of the kidney except for some arteries to the isthmus. In the horseshoe kidney, the anterior row of calices points dorsomedially and the ventral row points dorsolaterally and also the renal pelvis is in a ventral position.

High insertion of the ureter is common in the horseshoe kidney and the ureters pass over the isthmus anteriorly resulting in poor drainage from the kidney. Seventy percents of horseshoe kidneys have symptoms due to urinary stasis, hydronephrosis, infection or urolithiasis. Prevalence of stone ranges from 20% to more than 60% is secondary from infection and poor drainage from the renal pelvis(2,4). The indication for operation in a horseshoe kidney is mainly due to calculus disease.

Various treatments have been used for the management of calculi in the horseshoe kidney, including extracorporeal shock wave lithotripsy (ESWL), ureterorenoscopy (URS), percutaneous nephrolithotomy (PCNL) and open surgery. Due to the altered anatomy of the horseshoe kidney, ureterorenoscopy is technically challenged and not universally recommended. ESWL and PCNL are the treatments of choice depending on the stone burden. Open surgery is reserved only for those in whom PCNL fails.

The limitations of extracorporeal shock wave lithotripsy (ESWL) for treatment of the stone in a horseshoe kidney are the distance between the flank to stone that fail to focus stone at the focal point (F2) that providing the difficulty in accurate stone position. The medial calices may be obscured by the spine and stone in lower calices may be obscured by the bony pelvis and the impairing of the passage of stone fragments(6,7). ESWL is associated with a low clearance rate and the frequent need for auxillary treatments. Some cases need insertion of a retrograde catheter and injection of contrast media for localization of the stone. Forty seven to eighty seven percent of patients were rendered free of stone after ESWL treatment alone(4,6,8). ESWL is likely to requiring multiple sessions. A good result can be achieved if good patient selection is done i.e the small (less than 15 mm) or moderate (15 to 25 mm) stone volume. The stone free rate of these stones is 76% which is not different from the result of ESWL in a normal kidney (77%). Adjunctive procedures such as preoperative retrograde catheter placement, ureteroscopy and placement of a double pigtail stent may be needed before or after ESWL(6) renders a more stone free rate.

Percutaneous nephrolithotomy is accepted for the treatment of large renal and upper ureteric stone. Due to the abnormal anatomy of the horseshoe kidney, minor modifications to the standard technique are required(7,9). In lower position and malrotation of the kidney, the access is usually lower and more vertical. Dilation of the access tract is more difficult for screening by fluoroscopy because of the vertical puncture. The renal pelvis of the horseshoe kidney is deeper than the normal kidney. The extra-long Amplatz sheath and nephroscope may be needed especially in obese patients.

Despite the abnormal vessels in horseshoe kidney, no vessel is found on the dorsal aspect of the kidney except at the isthmus(5). All blood vessels except to the isthmus enter the kidney from the medial aspect and dorsal arteries to isthmus are protected by the spine. Percutaneous access via upper pole is ideal with no risk of arterial bleeding. An upper pole nephrostomy tract allows for intrarenal access to the upper pole calices, renal pelvis, lower pole calies, ureteropelvic junction and proximal ureter. The upper pole access can also decrease blood loss since the long axis of the nephroscope is aligned with the long axis of the kidney, minimized nephroscope torque on renal parenchyma during the procedure.

Even with the upper pole access, the incidence of pneumothorax or hydrothorax is not increased because of the low position of the horseshoe kidney. CT scan before PCNL is recommended due to the ability to detect small or radiolucent stones and the relative anatomy of the kidney and colon(2).

Renal pelvis in a horseshoe kidney is in the ventral position. It is easy to reach with nephroscope during the PCNL. Percutaneous nephrolithotomy has been shown to be highly successful with 89% overall stones - free rate and 12.5% of major complications, including significant bleeding with post operative blood transfusion, nephropleural fistula and pneumothorax(8).

Conclusion

Percutaneous nephrolithotony in the horseshoe kidney and malrotated kidney appears to be an effective and safe method of treatment for renal calculi in which only a minor modified technique needed and usually requires upper pole access. No increase in the complication of arterial bleeding was found and the abnormal orientation of the collecting system offers the good access with a good result.

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การผ่าตัดส่องกล้องเพื่อรักษานิ่วในไตที่มีความผิดปกติด้านการเชื่อมตัวและการหมุนตัวตั้งแต่กำเนิด

บรรณกิจ โลจนาภิวัฒน์

ความผิดปกติด้านการเชื่อมตัวและการหมุนตัวของไตตั้งแต่กำเนิด เป็นความผิดปกติที่พบบ่อย โดยเฉพาะไตรูปเกือกม้า ซึ่งความผิดปกตินี้จะเป็นความผิดปกติของการหมุนตัว การอยู่ผิดที่ และการเชื่อมกัน ทำให้การรักษานิ่วที่อยู่ในไตที่มีความผิดปกติประเภทนี้มีความยุ่งยากและท้าทาย รายงานการรักษาผู้ป่วยที่มีนิ่วในไตรูปเกือกม้า 8 คน และผู้ป่วยที่มีความผิดปกติด้านการหมุนตัวของไตตั้งแต่กำเนิด 4 คน โดยวิธีผ่าตัดส่องกล้อง ผู้ป่วยที่มีไตเกือกม้า 2 ราย มีนิ่วทั้ง 2 ข้าง เทคนิคการส่องกล้องในไตประเภทนี้ จำเป็นต้องเจาะเข้าบริเวณขั้วไตส่วนบน และต้องดัดแปลงเทคนิคการทำผ่าตัดส่องกล้องต่างไปจากการผ่าตัดส่องกล้องในไตปกติเล็กน้อย ระยะเวลานอนโรงพยาบาลเฉลี่ย 3.3 วัน นิ่วในไต 12 ราย (ร้อยละ 86) สามารถเอานิ่วออกได้หมด และมี 2 ราย ที่มีเศษนิ่วเหลือเล็กกว่า 4 มิลลิเมตร พบภาวะแทรกซ้อนในผู้ป่วย 1 ราย ที่มีน้ำในช่องปอดจำนวนเล็กน้อยหลังการผ่าตัด ซึ่งไม่ได้รับการผ่าตัดเพิ่ม

การผ่าตัดส่องกล้องในผู้ป่วยที่มีความผิดปกติด้านการเชื่อมตัวและการหมุนตัว พบผลสำเร็จสูง และภาวะแทรกซ้อนหลังผ่าตัดพบน้อย ดังนั้นการผ่าตัดส่องกล้องน่าจะเป็นการผ่าตัดทางเลือกลำดับแรกของการรักษานิ่วในไต ที่มีความผิดปกติด้านการเชื่อมตัวและการหมุนตัวของไตตั้งแต่กำเนิด