Managing urologic disease in the horseshoe kidney

Richard Link, MD, PhD

SCOTT DEPARTMENT OF UROLOGY Division of Endourology and Minimally Invasive Surgery

Baylor College of Medicine



Targets for today

General overview of horseshoe kidneys in adults.

- Critical anatomic features.
- The special case of the horseshoe kidney in:
 - Ureteral obstruction
 - Nephrolithiasis
 - nephrectomy, donor nephrectomy)





Infection

Surgical management (heminephrectomy, isthmus division, partial

Horseshoe kidney demographics

- The most common of all renal fusion anomalies.
- Occurs in 0.25% of the population (1 in 400).
- Abnormality occurs between the 4th and 6th week of gestation.
- ► Kidneys are <u>not</u> ectopic.





Higher frequency in children with vertebral and neural tube defects.



Crossed fused ectopic kidney

90% of crossed ectopic kidneys are fused with the contralateral kidney

Ureter is usually orthotopic



Ectopic kidneys





Jimenez and Morant, 2011



98% of horseshoe kidneys are fused at the lower pole



Notocord and floor plate of the neural tube play an important role in kidney development.

Disruption of these structures during development results in renal fusion anomalies.





Neural Tube Somite

pV0 pV1 pV2 pMN

pV3

Sonic Hedgehog (Shh) protein forms a gradient in the neural tube and has particularly high expression in notocord and floor plate

Developmental Biology 340 (2010) 518–527 Contents lists available at ScienceDirect Developmental Biology journal homepage: www.elsevier.com/developmentalbiology

Midline signaling regulates kidney positioning but not nephrogenesis through Shh

Piyush Tripathi ^a, Qiusha Guo ^a, Yinqiu Wang ^a, Matthew Coussens ^a, Helen Liapis ^b, Sanjay Jain ^{a,b}, Michael R. Kuehn ^c, Mario R. Capecchi ^d, Feng Chen ^{a,e,*}

^a Internal Medicine, Renal Division, Washington University School of Medicine, St. Louis, MO, USA

^b Department of Pathology and Immunology, Washington University School of Medicine, St. Louis, MO, USA

^c Laboratory of Protein Dynamics and Signaling, NCI, NIH, Frederick, MD, USA ^d Human Genetics, University of Utah, HHMI, Salt Lake City, UT, USA

^e Department of Cell Biology and Physiology, Washington University School of Medicine, St. Louis, MO, USA

Selective inactivation of Shh (sonic hedgehog) in notocord and floor plate resulted in renal fusion but not agenesis. Fusion



Shh+/Shh+ control



Shh-/Shh-









Horseshoe kidney

> 50% of patients with horseshoe kidneys are asymptomatic.

> 10 years remained asymptomatic.

Glenn, 1959. Glenn JF: Analysis of 51 patients with horseshoe kidney. N Engl J Med 1959; 261:684.

Even when anatomic features appear dramatic, they may not be mag3 renal scan).





Even during an era (the 1950s) prior to routine abdominal ultrasound and CT imaging, 58% of patients with horseshoe kidneys observed for

functionally relevant (e.g. hydronephrosis with normal drainage on





Watch out for anatomic/functional correlation

684

THE NEW ENGLAND JOURNAL OF MEDICINE

ANALYSIS OF 51 PATIENTS WITH HORSESHOE KIDNEY*

JAMES F. GLENN, M.D.† DURHAM, NORTH CAROLINA

51 consecutive horseshoe kidney patients monitored over a 10 year period.

58% remained asymptomatic.

13% had recurrent infections. 13% had episodic pain. 16% % had symptomatic stones.

Datum	No. of Patients	Percentage		
Sex:				
Males	28	54.9		
Females	23	45.1		
Major presenting signs & symptoms:				
Pain	17	33.3		
Infection	12	23.5		
Incidental finding (asymptomatic)	12	23.5		
Passage of calculi	7	13.8		
Diagnosis elsewhere	3	5.9		
Method of initial diagnosis:				
Radiologic (intravenous or retrograde pyelograms)	42	82.3		
Anatomic (autopsy)	6	11.8		
Clinical (palpation)	3	5.9		
Accessory features:				
Infection	21	41.2		
Obstruction	18	35.3		
Calculi	16	31.4		
Uremia	3	5.9		
Rovsing syndrome	3	5.9		
No complications	14	27.5		

TABLE 1. Data in 51 Patients with Horseshoe Kidney.

Horseshoe kidney: symptomatic

- Most symptoms are related to hydronephrosis, infections or nephrolithiasis.
- Solve of patients have recurrent UTIs, "20% 80%" have nephrolithiasis (as compared to 9-10% of the general population).
- Classic constellation of symptoms: vague abdominal pain radiating to the lower lumbar region, nausea and vomiting. Exacerbated by hyperextension of the spine.
 - Does not appear to be resolved by division of the isthmus.



Horseshoe kidneys lie outside the pelvic inlet and do not confer an increased risk during pregnancy or delivery.

Horseshoe kidneys do not appear to have a higher progression rate to renal insufficiency.

Review of the UNOS database for transplant recipients in 2000 showed no horseshoe kidney patients who underwent renal transplantation.





Targets for today

General overview of horseshoe kidneys in adults.

Critical anatomic features.

- The special case of the horseshoe kidney in:
 - Ureteral obstruction
 - Nephrolithiasis
 - Surgical management (heminephrectomy, isthmus division, partial nephrectomy, donor nephrectomy)





Infection

Relationship to inferior mesenteric artery.

"Renal ascent is stopped by the junction of the aorta and inferior mesenteric artery."

Kidneys are generally lower in the abdomen.





In truth, the kidneys are lower than normal in roughly 60% of cases.

The isthmus lies just below the IMA in 40%.



3) Horseshoe kidney with isthmus lying at normal position of lower poles (40%). L4 below inferior mesenteric artery (40%).



4) Horseshoe kidney with isthmus in the pelvis (20%).

Journal of Pediatric Urology (2016) 12, 275e280



Trocar shifts lower and away from umbilicus

Anteriorly oriented renal pelvis and ureters.





Multiple renal vessels in 70% and variable vessels to isthmus. Vessels are smaller and originate lower.









Watch out for arteries entering the isthmus directly from the aorta or the iliacs.

Gupta T., et. al.: JPMER. Dec 2016,



Figure 3. Diagram presenting the proposed levels of origin of renal arteries.

					of renar arteries according to branching ieven.							
	Arteries Supplying Horseshoe Kidneys						Arteries Supplying Separated Kidneys					
Level of Origin of Renal Artery	Ν	Mean (mm)	Media: (mm)	n Min (mm)	Max (mm)	SD (mm)	Ν	Mean (mm)	Media: (mm)	n Min (mm)	Max (mm)	SD (mm)
Ι	230	4.54	4.60	0.60	8.50	1.64	596	5.53	5.70	1.00	9.80	1.61
II	108	4.28	4.20	1.40	7.40	1.35	2	4.45	4.45	2.80	6.10	2.33
III	57	3.41	3.30	1.10	6.30	1.11	0	0	0	0	0	0
IV	3	3.43	3.20	2.80	4.30	0.78	0	0	0	0	0	0

Retrospective CT angiogram study: 248 normal kidney patients vs. 88 horseshoe kidney patients.

Horseshoe kidneys:

Smaller arteries Arise lower on the aorta, common iliac or iliac Smaller in caliber

Table 2. The diameters of renal arteries according to branching level.

Majos M, et. al.: J. Clin. Med. 2019, 8, 464





Tricky vascular anatomy



Significant variation in isthmus thickness and presence of collecting system in the isthmus.

The isthmus is usually bulky and located at the L3 or L4 level.

Occasionally it is just a thin fibrous band.

In some cases, the lowermost calyces overlie the vertebral column.













Watch Out!

Asymmetrical systems are LEFT side dominant in 70%.



Case Report

A Case of Incomplete Removal of Horseshoe Kidney by Laparoscopic Nephrectomy in an Adult Leading to Urinary Leak: An Eye Opener

S. Venkat Ramanan (), P. Velmurugan (), A. R. Bhaskar Prakash, Anuj Arora, and LeelaKrishna Karri

Department of Urology. Sri Ramachandra Medical College and Research Institute. Chennai. India



Residual right lower pole with fusion well to left of aorta.



Hindawi Case Reports in Urology Volume 2019, Article ID 4132521, 3 pages

Laparoscopic right Nx



Left lower pole

The kidneys do not rotate completely antero-medially due to the fusion event.

This results in kidneys that are posteriorly rotated.





Horseshoe kidneys have the same number of calyces as normal kidneys but these calyces are atypical in orientation.



The ureter often inserts high into the renal pelvis.

The lower ureter usually enters the bladder normally.



Targets for today

- General overview of horseshoe kidneys in adults.
- Critical anatomic features.

The special case of the horseshoe kidney in:

- Ureteral obstruction
- Nephrolithiasis
- nephrectomy, donor nephrectomy)







Infection

Surgical management (heminephrectomy, isthmus division, partial



Anteriorly oriented renal pelvis and ureters.



Increased risk of urinary stasis









Increased UTI risk



Nephrolithiasis







Difficulty targeting during bodies, bony pelvis, bowel

approach.

Any modality that leaves fragments behind may be less effective in the horseshoe kidney due to suboptimal urine drainage.



ESWL



Ureteroscopy




ESWL in horseshoe kidney

Traditionally has been the mainstay of therapy for stone burden \leq 1.5 cm

Table 1					
Results of	SWL	patients	with	horseshoe	kidneys

Results of SWL patients with horseshoe kidneys						
Investigator, Y	Mean Stone Diameter, mm	No. of Patients	Fragmentation Rate, %	Stone- Free Rate, %	Retreatment Rate, %	
Esuvaranathan, ¹²² 1991	12	7	Not Reported	59	50	
Kirkali et al, ⁵ 1996	24	18	78	28	57	
Smith, ¹²³ 1989	Not Reported	14	Not Reported	79	29	
Bhatia and Biyani, ² 1994	28	27	Not Reported	70	48	
Sheir et al, ⁷ 2003	13.5	49	Not Reported	71	71	
Tunc et al, ⁶ 2004	22	46	Not Reported	66	Not Reported	
Ray et al, ⁹ 2011	9.5	41	Not Reported	39.1	71	

Tan YK: Management of stones in abnormal situations. Urol Clin N. Am. 40, 2013

Total stone free rate = 60% (39 to 79%) Retreatment rate = 61% (29 to 71%)



PCNL in the horseshoe kidney

JOURNAL OF ENDOUROLOGY Volume 22, Number 6, June 2008 © Mary Ann Liebert, Inc. Pp. 1219–1226 DOI: 10.1089/end.2008.0051

The Presence of Horseshoe Kidney Does Not Affect the Outcome of Percutaneous Nephrolithotomy

Nicole L. Miller, M.D.,¹ Brian R. Matlaga, M.D.,² Shelly E. Handa, R.N., B.S.N.,³ Larry C. Munch, M.D.,³ and James E. Lingeman, M.D.³

JOURNAL OF ENDOUROLOGY Volume 24, Number 4, April 2010 © Mary Ann Liebert, Inc. Pp. 531–536 DOI: 10.1089/end.2009.0264

Percutaneous Management of Staghorn Calculi in Horseshoe Kidneys: A Multi-Institutional Experience

Evangelos N. Liatsikos, M.D., Ph.D.,¹ Panagiotis Kallidonis, M.D.,¹ Jens-Uwe Stolzenburg, M.D., Ph.D.,² Micheal Ost, M.D., Ph.D.,³ Frank Keeley, M.D., Ph.D.,⁴ Olivier Traxer, M.D., Ph.D.,⁵ Norberto Bernardo, M.D., Ph.D.,⁶ Petros Perimenis, M.D., Ph.D.,¹ Arthur D. Smith, M.D., Ph.D.³

Consensus: Stone clearance rates generally equivalent to PCNL in normal kidneys.

n = 44 Primary stone free rate: 84% Second look in 11% Overall stone free: 93%

n = 17 Primary stone free rate: 82%

PCNL in horseshoe: caveats #1

Percutaneous access can be more technically difficult due to malrotation, lower renal units.

Critical Features # I

Relationship to inferior nesenteric artery.

Renal ascent is stopped by the junction of the aorta and inferior mesenteric artery.

Kidneys are lower in the



Critical Features #5

The kidneys do not rotate completely antero-medially due to the fusion event.

This results in kidneys that are posteriorly rotated.

Almost all require upper pole, posterior calyceal access.

Some require CT guided or laparoscopic-assisted access.



Critical Features #6

Horseshoe kidneys have the ne number of calyces as al kidneys but these alyces are atypical in







Good news and bad news....



PCNL in horseshoe: caveats #2

Multiple vessels and UPJ obstruction can also complicate access to the calyx of entry and the ureter.





Critical Features #7

he ureter often inserts n into the renal pelvi

- Posterior vessels into the isthmus preclude access there.
- entry point of vessels..





This is a factor almost exclusively when attempting lower pole access.

Bleeding risk is not increased in horseshoe kidney PCNL due to medial

Janetschek, G. and Kunzel, K. H. Br J Urol: 62, 1988



Due to kidney location, the access tract can be very long in horseshoe kidney patients, requiring special instrumentation and technique.

long nephroscopes flexible nephroscopes buried sheath





Low stone clearance rate High retreatment rate



ESWL

0



Ureteroscopy in the horseshoe kidney

- Technically challenging.
- Usually requires:
 - flexible instrumentation
 - access sheath
- Not all calyces can be accessed.
- Staged procedures are common.
- Subsequent drainage remains a factor.



Critical Features #7

The ureter often inserts into the renal pelvis

The lower ureter usually , rs the bladder normal



Critical Features #6







Flexible Ureterorenoscopy With **Holmium Laser in Horseshoe Kidneys**

Benoit Molimard, Saeed Al-Qahtani, Amine Lakmichi, Majed Sejiny, Sixtina Gil-Diez de Medina, Xavier Carpentier, and Olivier Traxer

UROLOGY 76 (6), 2010

- ► 17 patents
- Average stone burden 1.6 cm.
- ► 42% of patients required a second procedure.
- On average 1.5 procedures per patient (total of 25).
- Overall stone free rate was 88%.
- No major complications, no transfusions.
- One patient treated for pyelonephritis.



Ureteroscopic Urinary Stone Treatment Among Patients With Renal Anomalies: Patient Characteristics and Treatment Outcomes

Jaap D. Legemate, Barbaros Baseskioglu, Jakub Dobruch, Jorge Gutierrez-Aceves, Oscar Negrete, Carlos Rioja Sanz, Muharrem Murat Yildiz, and Jean J.M.C.H. de la Rosette

UROLOGY 110, 2017

Outcomes

Stone-free Renal stor ≤80 mn >80 mr

Ureteral st ≤80 mm

Evaluation

Intraopera

Postopera

Horseshoe Kidney n = 43
17 (77.3)
10 (83.3)
7 (70.0)
(n = 22)
17 (85.0)
15 (93.8)
2 (50.0) (n = 20)
(1 - 20)
4 (9.3)
20 (46.5)
21 (48.8)
1 (2.3)
1 (2.3)
6 (14.0)
2 (4.7)
5 (11.6) 3 (7.0)
1 (2.3)
1 (2.3)
O (O)
0(0)
O (O)
(n = 43)
3 (7.0)
1 (2.3)
0 (0) 1 (2.3)
0 (0)
1 (2.3)
(n = 43)
1 [1-2]
(n = 43)
7 (16.3)
(n = 43)
5 (12.5)
(n = 40)



UPJ obstruction in horseshoe kidneys

Incidence is 15 - 30% in the horseshoe population.

Critical Features #7

The ureter often inserts high into the renal pelvis.

he lower ureter usually nters the bladder normally.



Critical Features #3

Multiple renal vessels in 70% and variable vessels to isthmus.



- <u>Conventional</u> open management included dismembered pyeloplasty, division of isthmus and nephropexy.
- More recent studies confirm that division of isthmus and nephropexy are not necessary in most cases.









Success Rates (|° UP|)

- Acucise endopyelotomy 56 77 %
- Endopyelotomy 62 94%
- Laparoscopic pyeloplasty 89 98%
- Open pyeloplasty 86 93%

Also....with longer followup (5-10 yrs), we are seeing lots of late failures with endopyelotomy...

Crossing vessels can be seen in 87% of endopyelotomy failures.



selection criteria:

Degree of hydronephrosis Presence of crossing vessel Degree of renal function



Tips and tricks

- Lower placement of trocars.
- Transperitoneal, and sometimes transmesenteric, approach is favored due to anterior location of pelvis and ureter relative to isthmus.
- A preoperative CT angiogram may be helpful to provide a roadmap of aberrant vasculature.
- Pelvic reduction is at the discretion of the surgeon. Correction of high insertion is not....

Critical Features # I

Relationship to inferior mesenteric artery.

Renal ascent is stopped by the junction of the aorta and inferior mesenteric artery.

Kidneys are lower in the abdomen.







Don't be a T-rex, spread out the trocars away from target.

Case example

31 year old woman with right upper quadrant pain and microscopic hematuria.

Pain generally worse with alcohol intake.

No prior surgery.









- Robotic dismembered pyeloplasty and pyelolithotomy performed.
- Multiple aberrant vessels and high insertion present.
- Large stone placed into LapSac.
- LapSac exteriorized at umbilicus and stone fragmented and evacuated with nephroscope and Cyberwand.
- Smooth postop recovery. Symptom free without new stone formation at 4 years postop.





Heminephrectomy

- In some cases (malignancy, infection), heminephrectomy is indicated and can be performed using minimally invasive approaches (laparoscopic, single site laparoscopic or robotic) in horseshoe kidney patients.
- Issues:
 - Positioning of trocars.
 - Anticipate and control multiple vessels
 - Divide the isthmus
 - Be careful for short arteries feeding the isthmus from the aorta.



Heminephrectomy

- Skill set is a fusion of skills used for laparoscopic donor nephrectomy and laparoscopic/robotic partial nephrectomy.
- Generally favor a top down approach, dealing with the isthmus as the last step once everything else is mobilized.







Not so effective with horseshoe kidney due to tethering at isthmus.

Dividing the isthmus

- Methods depend on thickness of isthmus.
 - Harmonic scalpel or ligasure for thin fibrous isthmus. Stapler also an option for medium thickness without collecting system.
 - Scissors with oversew/reconstruction for thick, "meaty" isthmus in fashion similar to MIS partial Nx.
 - Laparoscopic Satinsky clamp can be helpful for contralateral parenchymal control. In my experience, bulldog clamping of contralateral lower pole vessels is unnecessary.
 - Watch out for short posterior vessels!



ignificant variation in sthmus thickness.

The isthmus is usually bulky and located at the L3 or L4

Occasionally it is just a thin fibrous band.

most calyces overlie the vertebral colum









Case example

24 year old patient with horseshoe kidney and recurrent episodes of pyelonephritis.

Severe episode of urosepsis necessitating PCN placement.

CT scan shows burned out, decompressed right renal moiety.







Elevation of right renal moiety.

Placement of laparoscopic Satinsky clamp across isthmus.

Transection of isthmus.





Oversewing the isthmus

Use of Lapra-Ty to tension suture line after clamp removal.





Alternative technique

Renal cell carcinoma risk is not increased by presence of horseshoe kidney.

Not infrequently, isthmus division is necessary to facilitate partial or radical nephrectomy.

- ▶ The presence of a tumor in the isthmus can be a challenging situation as the blood supply can be indeterminate.
- Depending on the size and location of the tumor, bilateral lower pole vessels and/or posterior branches off the aorta may need to be controlled.
- Would be an excellent indication for fluorescence imaging on the robotic platform or intraoperative doppler ultrasound.







Integrating all the special characteristics of horseshoe kidneys.

Living donor heminephrectomy





USRDS annual report 2015



Sobering statistics

Kidney waiting list patients who died prior to transplantation: 2016







Since 1995, 84,695 patients on the kidney waiting list died before being transplanted.

Deceased donors



OPTN data 3/19/17

Transplants by donor type

Living donors

Waiting list





OPTN and NKF data 3/19/17



Relationship to inferior mesenteric artery.

"Renal ascent is stopped by the junction of the aorta and inferior mesenteric artery."

Kidneys are generally lower in the abdomen.



Critical Features #2

Anteriorly oriented renal pelvis and ureters.



Critical Features #4

Significant variation in isthmus thickness and presence of collecting system in the isthmus.

The isthmus is usually bulky and located at the L3 or L4 level.

Occasionally it is just a thin fibrous band.

In some cases, the lowermost calyces overlie the vertebral column.







Critical Features #6

Horseshoe kidneys have the same number of calyces as normal kidneys but these calyces are atypical in orientation.







Critical Features #3

Multiple renal vessels in 70% and variable vessels to isthmus.





Transplant Donor Nx

Critical Features #5

The kidneys do not rotate completely antero-medially due to the fusion event.

This results in kidneys that are posteriorly rotated.





Critical Features #7

high into the renal pelvis.

The lower ureter usually enters the bladder normally







52 year old woman donating to her father.





Single-site technique + robotics



Compression of the renal parenchyma at the narrowest portion of the isthmus using standard partial Nx techniques.

Transection of the ureter.

Transection of the lower pole artery arising from the IMA.











Stapling across the isthmus.

Transection of the main renal artery.

Transection of the main renal vein.



CONCLUSIONS

Overview of horseshoe kidneys in adults, incidence, embryology and prognosis.

7 anatomic features of horseshoe kidneys that impact on disease and treatment.

Broad look at stone disease in horseshoe kidneys and technical considerations.

UPJ obstruction is common in horseshoe kidneys but management is fairly standard.

Tips and tricks for managing heminephrectomy and isthmus division for Nx and partial Nx.





