JOURNAL Of Medical Science And Clinical Research
IGM Publication   An Official Publication Of IGM Publication

The Morphology and Treatment of Coexisting Pelvi-Ureteric Junction Obstruction In Horseshoe Kidney

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Abstract
Background: *The most common congenital abnormality of urinary tract is duplex kidney but horseshoe kidney (HSK) represent the most common congenital renal fusion anomaly.[1] Pelvi-ureteric junction obstruction (PUJO) is a relatively common finding during urological investigation. The combination of HSK and PUJO occurs in 15-33% of patients. Surgical treatment can be challenging in such cases. Herein, we report our experience of management of PUJO in HSK.*
Materials and Methods: *In between July 2012 and April 2017, fourteen patients were identified with the horseshoe kidney having PUJO. There demographic, diagnostic, and procedural data were recorded. All cases were investigated with renal ultrasonography, computed tomography (CT) and renal scintigraphy. Surgical repair of PUJO by pyeloplasty was done in nine cases. Hellstrom repair was done in three cases and two cases underwent nephrectomy. Treatment outcome was assessed in terms of quality of life, recurrence and complications.*
Results: *Median age of presentation was 30.78 years (9-51 years) . At a mean follow-up of 22 months all were symptomatic free. Out of fourteen, ten patients presented with flank pain. Incidental hydronephrosis was detected in two patients, and two had a recurrent febrile urinary tract infection. Out of nine patients who underwent pyeloplasty, Anderson-Hynes dismembered pyeloplasty was performed in five and foley y-v plasty was performed in four patients. Hellstrom repair was done in three cases and two patients underwent nephrectomy. Laparoscopy surgey was done in two patients each of Anderson-Hynes dismembered pyeloplasty and Hellstrom repair. Nephrectomy was done in one patient by laparoscopy who developed chyluria later on and was managed conservatively. No complications were detected in others during follow-up and all had a good quality of life.*
Conclusions: *PUJO in horseshoe kidneys is a surgical challenge. Treatment should be individualized depending upon the clinical profile of patient.*
Keywords: *Horseshoe kidney (HSK); Pelvi-ureteric junction obstruction (PUJO); Pyeloplasty; Laparoscopy.*

Introduction

Horseshoe kidney (HSK) is one of the major congenital anomaly of the upper urinary tract similar to the duplication of collecting system.[1] The fusion of the lower poles of each metanephros leads to formation of a parenchymal or fibrous

isthmus between the kidneys resulting in HSK. Incidence of HSK is 1 in 400–1000 births and have a male predominance.[2] Pelvi-ureteric junction obstruction (PUJO) is found in one third of HSK cases.[3,4,6] In the literature , factors predisposing to obstruction in horseshoe kidneys described include: a high insertion of the ureter with an acute PUJ angle; an isthmus over which the ureter must pass; aberrant blood vessels in the region of the PUJ; a relatively intrarenal pelvis with a medially placed inferior pole and congenital PUJ narrowing.[7,8] The diagnosis and management of such cases can be difficult and complicated because of the high anatomic variability, the degree of obstruction and clinical aspects. Surgical management of PUJO in HSK includes various techniques of the reconstruction such as pyeloplasty, ureterocalycostomy or hellstrom repair. The Anderson–Hynes pyeloplasty is still the more common primary procedure to manage PUJ obstruction in the horseshoe kidney.[9]

Aim and Objectives

The aim of this study was to review the morphology and treatment of pelvi-ureteric junction obstruction (PUJO) in horseshoe kidney (HSK) in our institute.

Material and Methods

Patients who underwent various surgical procedures for PUJO in horseshoe kidney at our institution between July 2012 and June 2017 were evaluated. Their demographic, diagnostic and procedural data were recorded. Ultrasonography was used for initial evaluation. Computed tomography (CT) with urography was required in all patients to demonstrate the precise anatomy before the surgical procedure. Full written informed consent was obtained from each patient after explanation of all the available techniques and risk of surgical procedures.

The surgical outcome was evaluated in terms of, operative duration, surgical findings, volume of bleeding, length of hospital stay, perioperative complications, postoperative symptoms, postoperative structural change of hydronephrosis, and postoperative renal function of the side with hydronephrosis. After 6 months stent was removed and repeat USG and diethylene triamine pentacetic acid (DTPA) renal scintigraphy was done. A successful outcome was defined as the subjective and objective improvement in symptoms. Objective outcomes was evaluated postoperatively with USG by assessing the changes in degree of hydronephrosis using Society for Fetal Urology (SFU) grade. DTPA assessed postoperative split renal function and drainage. Furosemide as diuretric was administrated (1 mg/kg intravenous [i.v.]) based on visually confirmed, complete filling of the dilated pelvis. Standard computer generated curves was used for drainage evaluation. Functional delay was defined as a rapid washout of over 50% accumulated nuclide after furosemide administration.[3] Patients were also followed up for any urinary infection and any new complications.

Results

Between July 2012 and April 2017, fourteen patients with a mean age of 30.78 years (9 to 51 years) underwent various surgical procedures for PUJO in horseshoe kidney at our institution. The mean follow-up period was 22 months (range, 6–30 months; median, 24 months).The major presenting symptoms were flank pain in ten patients. HSK with PUJO was incidentally detected in USG in two patients and two patients presented with recurrent urinary tract infection (UTI). There was no history of haematuria in any of the patients. All patients had unilateral pathology, ten left-sided and four right-sided. All patients had obstructive drainage pattern in DTPA scan and pre-operative stenting was done in two patients as shown in Table 1. Obstruction was confirmed in all cases by progressive accumulation of the nuclide or less than 50% washout in the diuretic renal scan taking into consideration the results of ultrasound grading.

Eleven patients had SFU grade 4 hydronephrosis and three patients had grade 3 hydronephrosis. None of the patients had dilatation of ureters. Intraoperative findings included aberrant crossing vessels in seven cases, high ureteral insertion in four and congenital narrowing of PUJ in three cases in which histology revealed intrinsic ureteropelvic obstruction. Out of nine patients who underwent pyeloplasty, Anderson-Hynes dismembered pyeloplasty was performed in five and foley y-v plasty was performed in four patients. Hellstrom repair was done in three cases and two patients underwent nephrectomy.

TABLE 1 : Patient characteristics and preoperative configuration and function of the affected kidney

Sr.No	Gender	Age	Side	Symptoms	Preop-Stenting	Preop-Sfu Grade Of Hn	Preop-Df	Preop-Drainage
1	M	27	L	Flank Pain	Yes	4	12	Obstructive
2	F	23	L	Flank Pain	No	4	24	Obstructive
3	M	17	L	Flank Pain	Yes	4	10	Obstructive
4	M	43	L	Flank Pain	No	4	27	Obstructive
5	F	34	R	Flank Pain	No	3	34	Obstructive
6	M	32	L	Incidental	No	4	26	Obstructive
7	F	30	R	Flank Pain	No	4	-	Obstructive
8	M	28	R	Flank Pain	No	4	36	Obstructive
9	M	29	L	Recurrent Uti	No	3	30	Obstructive
10	F	37	L	Flank Pain	No	4	24	Obstructive
11	M	45	R	Incidental	No	4	-	Obstructive
12	M	51	L	Flank Pain	No	4	26	Obstructive
13	F	26	L	Flank Pain	No	4	36	Obstructive
14	M	9	L	Recurrent Uti	No	3	28	Obstructive

M-MALE ; F-FEMALE ; L-LEFT ; R-RIGHT

Laparoscopy surgey was done in two patients each of Anderson-Hynes dismembered pyeloplasty and Hellstrom repair as shown in Table 2. Nephrectomy was done in one patient by laparoscopy. Mean operation time was 111.07 minutes with average amount of bleeding of 41.42 ml. Mean duration of stay in hospital post surgery was 4.42 days.

Table 2 : Perioperative findings

Sr.No	Cause Of The Obstruction	Surgery	Operative Duration (Min)	Volume Of Bleeding (Ml)	Length Of Hospital Stay (Day)
1	Aberrant Vessels	Ah Pyeloplasty	125	40	6
2	High Insertion Of Ureter	Foley Yv Plasty	150	50	6
3	Aberrant Vessels	Hellstrom Repair	70	30	4
4	Aberrant Vessels	Ah Pyeloplasty	120	60	5
5	High Insertion Of Ureter	Foley Yv Plasty	115	60	5
6	Aberrant Vessels	Laparoscopyah Pyeloplasty	150	50	4
7	Congenital Pujo	Laparoscopynephrectomy	90	40	3
8	High Insertion Of Ureter	Foley Yv Plasty	110	50	4
9	Aberrant Vessels	Laparoscopyhellstrom Repair	85	30	3
10	Congenital Pujo	Ah Pyeloplasty	110	40	5
11	Congenital Pujo	Nephrectomy	90	30	4
12	Aberrant Vessels	Laparoscopyah Pyeloplasty	140	30	5
13	High Insertion Of Ureter	Foley Yv Plasty	110	40	5
14	Aberrant Vessels	Laparoscopyhellstrom Repair	90	30	3

All patients had an ultrasound examination 6 months after the operation, and hydronephrosis had either disappeared or was downgraded as shown in table 3. Four patients demonstrated functional delayed drainage and ten had total washout without furosemide. None of the patients had obstruction in post-operative renal dynamic scans. Surgical outcomes are shown in Table 3. None of the patients had flank pain during the postoperative follow-up period. Chyluria was seen in one case of nephrectomy done by laparoscopy and it was later managed conservatively. No one had a urinary tract infection or recurrence of symptomatic HN during the follow-up period.

Table 3 : Therapeutic outcome of all cases

Sr.No	Post Op Symptoms	Post Op - Sfu Grade Of Hdn	Post Op-Df	Post Op-Drainage
1	None	1	24	Free
2	None	2	35	Delayed
3	None	1	30	Free
4	None	2	40	Free
5	None	1	45	Free
6	None	No Hdn	40	Free
7	Chyluria	2	20	Delayed
8	None	1	46	Free
9	None	No Hdn	45	Free
10	None	No Hdn	40	Free
11	None	2	23	Delayed
12	None	1	38	Free
13	None	No Hdn	47	Free
14	None	1	38	Delayed

Discussion

Berengario da Carpi in 1522 gave first time the description of a patient with horseshoe kidney (HSK).[10] It is characterized by the aggregation, usually at the lower pole, of the two kidneys over the spine across the midline. Its incidence is 1 in 400–1000 births and about 0.1 % in intravenous pyelogaphies and represent the most common fusion anomaly of the kidney.[11] A slight male predominance is seen in HSK. Most patients with horseshoe kidneys are asymptomatic, but can be associated with PUJO in about 14% to 35% of cases and is one of its most common complications.[12,13,14] The management and surgical indications for treating these patients follow the same criteria as for patients with normal kidneys.

Horseshoe kidney in children differs from that in adults: 80% of patients requiring surgery reported by Sharma et al.[15] Renal calculi is presented in 21-60% as reported by Pitts et al.and Culp et al.[14,16] In our study, none of the patients had calculi. In our study, obstruction is was due to high insertion of the ureter, congenital PUJO and crossing vessels and crossing vessels contributed to the obstruction to a higher degree than in PUJO without a fusion anomaly.[17] A narrow PUJ was found during surgery in 21.4%of our patients; this is reported to occur in 78% of PUJO in normal kidneys.[17]

Hydronephrosis in HSK is due to the kinking of ureter at isthmus as they are anteriorly placed.[18] Dismembered pyeloplasty is usually the procedure of choice in PUJO. Basic principles of surgical management include excision of stenotic ureteropelvic segments, trimming of a redundant pelvis, transposition of any aberrant vessels, and ureteropelvic anastomosis. Success rates of 90–100% for patients with normal kidneys and 55–80% for patients with horseshoe kidneys has been reported for open pyeloplasty.[13,14,19] In 1980 minimally invasive techniques were developed and applied in these particular patients with variable results. Jabbour *et al.* reported retrograde endopyelotomy treatment in the presence of kidney anomalies in nine patients (four

horseshoes, three lumbar and two pelvic kidneys), with a long-term success rate of 78% and no major bleeding complications.[20] Successful use of endopyelotomy was reported by Nakamura *et al.* in three patients with horseshoe kidneys.[21] A favourable outcome was reported with endopyelotomy via percutaneous approach by Bellman in one patient with a horseshoe kidney.[22] Laparoscopy pyeloplasty since its introduction in 1993 by Schuessler *et al.*, has rapidly become part of the urological options for treating primary PUJO.[23] Long learning curve is one of the hurdle in its widespread clinical use but its success rates appear to be similar to those for open PUJO repair.[24] Few cases have been reported for laparoscopic pyeloplasty in PUJO with horseshoe kidney.[25] The difficulty of this technique and the prolonged operative duration are still disadvantages although improvements in laparoscopic instrumentation have been introduced recently in an attempt to reduce the training and experience required.[23] The integration of computer-enhanced robotic technology with the technical skill of the surgeon by da Vinci System may reduce some of the disadvantages of laparoscopy. Sung *et al.* first reported the feasibility of robotic pyeloplasty using female pigs randomized to surgery with or without use of the Zeus robot.[26] No significant differences was found in operative duration, suturing time, or number of suture-'bites' per ureter between the robotic and non-robotic procedures. The initial capital cost of the equipment, together with the costs of the associated disposables is an important issue in robotic pyeloplasty but its advantages may outweigh this. Inorder to draw such conclusions prospective studies addressing cost-benefit in high-risk populations are needed.

Isthmusectomy followed by nephropexy of the ipsilateral kidney and dismembered pyeloplasty have been described by some authors for open surgical repair of PUJO in horseshoe kidneys. Isthmusectomy may allow the kidneys to lie in a more dependent position that maintains the patency of newly reconstructed ureteropelvic regions, as reported by Culp et al.[16] Electrocautery under direct vision, microwave coagulation, ultrasonic shears, or with the argon beam can be used for division of isthmus.[10] Other authors proposed a different approach suggesting that this division is unnecessary and consider Anderson–Hynes pyeloplasty with no division of the isthmus.[3] Our present study did not involve division of the isthmus, and had good clinical and radiological results.

Dismembered pyeloplasty was one of the major surgery performed in our study but three patients successfully underwent a Hellstrom vascular hitch repair, as first described in 1949.[27] Theoretically, a crossing vessel causing obstruction in isolation can be treated with either a vascular hitch or dismembered pyeloplasty. For hellstrom repair , prior studies have suggested that the best candidates are those with a crossing vessel and a normal caliber ureter with good peristalsis across the UPJ.[28] A success rate of 95% was reported at a mean follow up of 22 months in a series of 20 patients undergoing an MIS vascular hitch procedure.[28] Its application remains controversial but our experience and the experience of others suggests it is a reasonable approach in well selected patients.[29,30]

The retroperitoneal approach via a flank incision provides excellent exposure of the PUJ even in a horse shoe kidneys although we used a transperitoneal approach via lower midline or transverse incision for exposure of the PUJ in horseshoe kidney.[3] The reported use of an extraperitoneal approach to laparoscopic pyeloplasty in horseshoe kidney is limited to one description of a patient by Hsu et al.[25] The median hospital stay of 7.6 days was longer than in other series with laparoscopic management of PUJO (3.2–3.3 days).[5,31]

Various reports on outcome of pyeloplasty for horseshoe PUJO comes from series of open Foley Y-V plasty and they generally lack reporting of both radiologic and clinical follow up which is now standard.[14] Success rates of open

surgery in horseshoe kidneys were less favorable than in orthotopic kidneys, ranging from 55 to 80% compared to 90% respectively.[13,14,32,33] Similarly success rates of endopyelotomy in horseshoe kidneys range from 66-75% compared to rates in orthotopic kidneys, which range from 77 to 89% with absent crossing vessels, and less than 70% in the presence of a crossing vessel. [20,21,34-36] The overall radiographic success rate of 78% is thus comparable to success rates reported for open pyeloplasty in horseshoe kidneys.[13,14] Post-operatively, the differential function increased in all cases in our study. After Anderson-Hynes pyeloplasty the decrease in hydronephrosis in ultrasound and adequate drainage in renal dynamic scan were comparable to those in normal kidneys. For the diagnosis, management and follow-up of patients of horseshoe kidney with PUJO ultrasound and renal dynamic scan are usually sufficient. Ultrasound reduces the need for cross-sectional imaging in most cases as it is very accurate and sensitive in these patients.[37]

Conclusions

PUJO in horseshoe kidneys is a surgical challenge. Success rates of MIS pyeloplasty in horseshoe kidneys may be inferior to those in heterotopic kidneys and so treatment should be individualized depending upon the clinical profile of patient. Pyeloplasty, hellstrom repair and nephrectomy are feasible treatment depending upon the cause of PUJO and renal function. With the technical improvements all procedures can be performed using laparoscopy but. This study is limited by its small number and due to the rarity of horseshoe kidney, large case series will take time to occur, which limits our ability to interpret the outcomes and/or advantages of various techniques.

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