Original Article

Etiological Determinants of Congenital Pelviureteric Junction Obstruction

M. Irfan Munir, Zahid Iqbal, Safdar Hassan Javed, G.M. Subhani, Khalid Mehmood, Tahir Bashir Malik, Brig. Sajjid Mushtaq, Fayyaz Ahmed Malik

ABSTRACT

Congenital ureteropelvic junction obstruction is an important and treatable cause of neonatal hydronephrosis. It may be due to congenital absence of nerves leading to adynamic segment causing functional obstruction. It may also be due to defective muscular arrangement and replacement of muscles by fibrosis leading to anatomical obstruction or it may be caused by extrinsic compression by aberrant lower polar vessel. The aim of the present study is to define the etiological determinants of congenital PUJ obstruction. **Objectives:** To study the macroscopic abnormalities of congenital PUJ obstruction and correlating theses abnormalities with the microscopic and immunohistological findings. **Material and Methods:** It was a cross sectional observational study and patients presenting to outpatients department, irrespective of age and sex, with the diagnosis of PUJ obstruction and needing surgery were included in the study. Anderson Hynes Pyeloplasty was done in all cases and resected portion of redundant pelvis and narrow segment was submitted for histopathological and immunohistological examination. **Results:** Congenital PUJ obstruction was more common in males with a male to female ratio of 2:1 and it was common on left side in 55.55% cases. Presentation was in wide age range patients (2-40 years). In 33.33% patients it was structural abnormality where we were unable to pass feeding tube and in 66.66% patients it was functional abnormality where it was distensible PUJ. Predominantly circular muscle arrangement was seen in 15(57.69%) cases. Varying degree of replacement of muscle fibers with fibrous tissue was seen in all cases and total replacement in those where kidney was nonfunctional due to PUJ obstruction. Nerves were present in 5/26 (19.23%) cases. In 4/5 (80%) cases of crossing vessels, nerves were present. **Conclusion:** Most cases of congenital PUJ obstruction are due to functional obstruction but anatomical obstruction also has a significant contribution (33.33%).Predominant circular muscle arrangement is the abnormality leading to impaired peristalsis. Absence of nerves leading to functional obstruction is the major defect in congenital PUJ obstruction. Crossing vessel is the real cause of PUJ obstruction mechanically compressing the PUJ in vascular tangle cases. **Key Words:** PUJ, Pyeloplasty, IVP, DTPA renal scan.

INTRODUCTION

Ureteropelvic junction obstruction is the most common cause of significant dilatation of pelvicalyceal system in fetal kidneys accounting for 44-65% of all dilatation of collecting system[1]. Pelviureteric junction obstruction may be congenital or acquired. Patients present with urinary infection, renal mass, heamaturia, failure to thrive, persistent vomiting or incidental diagnosis on ultrasonography[2]. Different treatment options for congenital pelviureteric junction obstruction are available including watchful waiting, antegrade or retrograde endopyelotomy with cold knife, diathermy, holmium laser[3]or acucise, balloon dilatation, laparoscopic pyeloplasty, open Pyeloplasty and robotic Pyeloplasty.[4,5] Open pyeloplasty has two types: membered and dismembered; Anderson Hynes pyeloplasty whether intubated or nonintubated[6] is the classical example of dismembered type and Culp deweerd's is a membered type of pyeloplasty. Anderson Hynes pyeloplasty is the preferred method

of treatment as this has >90% success rate[7]. In the present study, Anderson Hynes pyeloplasty through a small 2-3 inches incision and without mobilizing the kidney will be done. Indications of surgery will be symptoms associated with obstruction, impairment of overall renal function or progressive impairment of >5% ipsilateral renal function, development of stones in cases of pelviureteric junction obstruction or rarely, causal hypertension.[8] If untreated obstruction leads to renal damage as shown by studies on animals[9] and can lead to end stage renal disease. To treat any disease its etiology must be understood. The pathophysiology of congenital PUJ obstruction is unknown.[10] The pathology is characterized by a persistent, narrow, aperistaltic segment of ureter, 2-10 mm long, immediately below the pelviureteric junction[2]. The abnormalities described are:

1. Absent or deficient muscles at ureteropelvic junction.
2. Abnormal muscle orientation: circular muscle arrangement rather than the normal predominant longitudinal/spiral/helical muscle arrangement.[2, 10]
3. Excess collagen.[11]
4. Crossing vessels obstructing the pelviureteric junction.
5. Absent or deficient nerves

The abnormalities described are not consistent and to our knowledge no work has been done on it in Pakistan. So the present study is being undertaken to demonstrate the exact etiology of congenital pelviureteric junction obstruction in Pakistani population.

Objectives

1. To detect the macroscopic abnormalities of pelviureteric junction and involved ureters in patients of pelviureteric junction obstruction.
2. To correlate these macroscopic features with the histopathological and immunohistochemical features for determination of etiology of congenital pelviureteric junction obstruction.

Operational definition

Macroscopic features include:

1. Crossing vessels
2. Length of narrow segment
3. Calibre of narrow segment

Histopathological features include:

1. Absent or deficient muscles at ureteropelvic junction.
2. Muscle fiber arrangement
3. Presence of excess collagen

Immunohistopathological features include:

Presence or absence of nerves detected by S-100 staining in the narrow segment of PUJ.

a) Setting

The study was conducted at department of Urology Allied Hospital Faisalabad. Allied Hospital Faisalabad is a tertiary care center attached with Punjab Medical College Faisalabad.

a) Duration of study

Duration of study was two years

c) Ethical consideration

Informed consent was taken from all patients after full explanation of the procedure to the patients. Approval of the project was taken from ethical committee of Punjab Medical College Faisalabad.

d) Sample size

A total of 27cases of pelviureteric junction obstruction were studied. Histopathology of the specimens was compared with the normal histology mentioned in literature.

e) Sampling technique

It was non probability consecutive sampling

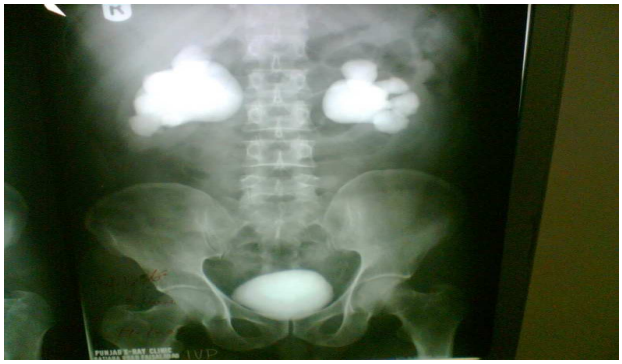
f) Sample selection

Inclusion criteria

All the consecutive patients presenting to Urology department, irrespective of age and sex with the

diagnosis of congenital pelviureteric junction obstruction as per Ultrasound/Intravenous urography/99mTc DTPA renal scan (if necessary) showing hydronephrosis without hydroureter and level of obstruction at pelviureteric junction and where surgery was indicated were included in the study.

Figure-1
IVP Showing the Bilateral PUJ Obstruction



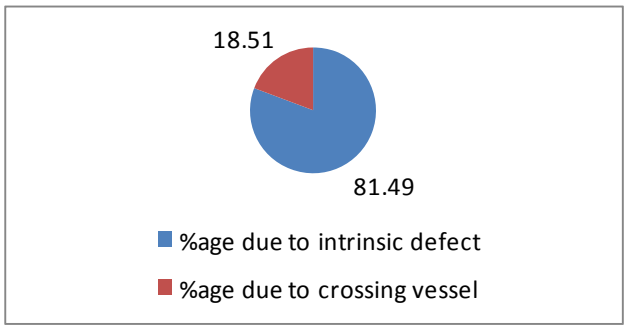
Exclusion criteria
Any patient having the diagnosis of pelviureteric junction obstruction with previous history of renal pelvic surgery or stone disease secondarily leading to pelviureteric junction obstruction was excluded from the study.

g) Study design
It was a cross sectional study.

DATA COLLECTION
All the consecutive patients irrespective of age and sex presenting to us with the diagnosis of congenital pelviureteric junction obstruction as diagnosed on the basis of inclusion criteria were included in the study. Informed consent was taken from all the patients. All the information regarding patient's bio data, clinical examination findings, investigations, macroscopic features, histopathological findings and immunohistopathological findings were recorded on a specially designed Performa.

RESULTS
A total of 27 patients fulfilled the inclusion criteria and were included in the study. Among these 18 (66.66%) were male, 9(33.33%) were female and male to female ratio was 2:1. Age range was between 2-40 years; average being 14.6 years. PUJ obstruction involved left side in 15 (55.55%) and right side in 11 (40.74%) patients and bilaterel in one (3.70%).Intravenous Pyelography showed left sided hydronephrosis in 13 (48.14%) and right sided in 11 (40.74%) patients, one (3.70%) patient showed faint nephrogram and 2(7.40%) were non functioning kidneys as contrast was not taken up by the kidneys even on delayed films. Narrow segment of PUJ was visible in 3(11.11%) patients and dye did not enter the ureter in 21(77.77%) patients. Tc 99m DTPA scan showed nonfunctioning kidney in two patients (7.40%). All 27 patients were explored and 5/27 (18.51%) patients were found to have lower polar aberrant crossing vessels crossing the PUJ and compressing it.

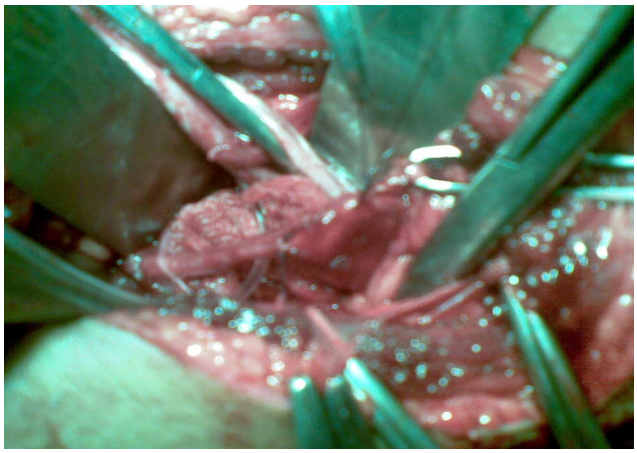
Figure-2
Showing the percentage of PUJ Obstruction due to crossing vessel and due to intrinsic defect



Anderson Hynes Pyeloplasty was done in 24 patients and three patients (11.11%) underwent nephrectomy due to very poorly functioning or nonfunctioning kidney. Crossing vessel was found in 5/27 (18.51%) patients. Crossing vessel was transposed to the opposite side after doing dismembered Anderson Hynes Pyeloplasty. We were able to pass 8 Fr feeding tube in 7(25.92%) patients and 6 Fr feeding tube in 11(40.74%) patients. In 9 (33.33%) patients, we were unable to pass the feeding tube. Length of narrow segment was measured and it ranged from 4 mm to 20mm, average being 7.19mm. Histopthalogist found 1 specimen to be autolysed due to leaking of formalin and it was naturally excluded from the histological examination. Rests of the specimens were processed and histological examination was done under light microscopy.

There was predominately circular muscle arrangement at the narrow segment of PUJ in 15/26 (57.69%) patients, and both circular and longitudinal muscle arrangement in 6 (23.07%), spiral arrangement in 2 (7.69%), helical in 1 (3.8%) and mixed pattern in 2(7.69%). Varying degree of fibrosis was seen in all patients. Varying degree of congestion (presence of inflammatory cells) was seen in 6 (23.07%) specimens. Nerves were present on Immunohistochemistry in 5/26(19.23%) patients and among them in four out of five (80%) patients of crossing vessels.

Figure-3
Showing the completed Anderson Hynes Pyeloplasty with funneling of newly built pelvis



DISCUSSION

PUJ obstruction in the present study occurred more on left side with a male prepondrance and a wide age range which coincides with the literature.[2,12] Bilateral disease was found in one patient which is below the bilaterality described in literature.[13,14] Presentation was usual in all patients. Aberrant lower polar crossing vessel was found in 18.51 % of our patients and it is found less frequently than mentioned in literature where about 42-60 % patients of PUJ obstruction were found to have crossing vessels.[15,16] Macroscopically anatomical obstruction was found in 33.33 % patient in which we were unable to pass the 6Fr feeding tube and it is in contrast with the literature review where it is mentioned that it is functional obstruction and is distensible.[12,15] Length of Narrow segment varied between 4-20mm, average being 7.19mm. On microscopy of the narrow segment there was predominantly circular muscle arrangement in the 57.69 % patients like the microscopic description by G. A. Maranya and colleagues[2] but unlike the finding of Sankar Kausik and colleagues and star NJ and colleagues [17, 18] who say that it is predominantly abnormal longitudinal muscle arrangement. Varying degrees of fibrosis was found in all specimens and this fibrosis replaced the normal smooth muscle to varying degrees and this is consistent with literature review.[19,15] Fibrosis replaced the smooth muscles completely in cases where kidney were severely hyrdonephrortic and especially in those where kidney had become nonfunctional due to severe hydronephrosis and this is consistent with literature review.[19] Immunohistochemistry with S-100 was done in all cases to detect the presence of nerves in the narrow segment of PUJ. Nerves were present in 5 cases and absent in 21 cases and this finding is showing similar results as shown in literature [20] it means that congenital PUJ obstruction is caused mostly by adynamic segment of the PUJ. Among the 5 cases where nerves were present, four were those where crossing vessel was found. It means that in most cases of crossing vessel there is no intrinsic defect (adynamic segment) present and compression by the crossing vessel may be the real cause of obstruction. This is in contrast with the finding of Stern JM, and colleagues who found that no histological abnormalities were found in only 43 % cases of crossing vessels[21] meaning there by that57% patients of crossing vessels were having histological abnormalities. Our finding of normal histology and presence of nerves in 80% patients of crossing vessel is consistent with the finding of Kajbafzadeh AM and colleagues and Yiee and colleagues[22,23] This also means that it may not be necessary to excise the narrow PUJ as done in Anderson hynes Pyeloplasty and only transposition of the vessel cephalad and burying the vessel in the peripelvic fat may only be necessary.

CONCLUSION

- Most cases of congenital PUJ obstruction are due to functional obstruction but anatomical obstruction also has a significant contribution (33.33%).

- Predominant circular muscle arrangement is the abnormality leading to impaired peristalsis.
- Absence of nerves leading to functional obstruction is the major defect in congenital PUJ obstruction.
- Crossing vessel is the real cause of PUJ obstruction mechanically compressing the PUJ in vascular tangle cases.

ACKNOWLEDGMENT

- Institutional Research Committee Punjab Medical College Faisalabad for the grant of funds for this project.
- Brig. Sajjid Mushtaq head of Armed forces institute of pathology who did the histopathology and Immunohistochemistry of the specimens.
- Biostatition Ms. Qurratulain and Haq Nawaz of CPSP regional center Faisalabad regarding their help on statistical issues and references writing respectively.

REFERENCES

1. Lim DJ, Park JY, Kim JH, Paick SH, Oh SJ, Choi H. et al. Clinical characteristics and outcome of hydronephrosis detected by prenatal ultrasonography. J Korean Med Sci 2003; 18: 859-62.
2. Maranya, G.A Odour PR. Pelviureteric junction obstruction: a surgical experience. East central Afr J Surg 2004; 9: 21-4.
3. Minervini A, Davenport K, Keeley FX Jr, Timoney AG. Antegrade versus retrograde endopyelotomy for pelviuretric junction (PUJ) obstruction. Eur Urol 2006; 49:536-43
4. Hubert J. Robotic pyeloplasty. Curr urol Ref 2003; 4:124-9.
5. Bhayani SB, Link RE, Varkarakis JM, Kavoussi LR. Complete da Vinci versus laparoscopic Pyeloplasty: cost analysis. J Endo Urol 2005; 19:327-32.
6. Aminul Haq, Khan I. Tubeless and stentless pyeloplasty. J Postgrad Med Inst 2003; 17:124-7.
7. Gogns C, Karamursel T, Tokati Z, Yaman O, Ozdiler E, Gogus O. Long term results of Anderson Hynes pyeloplasty in 180 adults in the era of endourologic procedures. Urol Int 2002; 73:11-4.
8. Streem SB, Franke JJ, Smith JA Jr. Management of upper urinary tract obstruction. In: Walsh PC, Retik AB, Vaugan ED, Wein AJ, Karousi LR, Novick AC, et al. Campbell's urology Vol.1.8th ed. Philadelphia: Saunders; 2002. p.463-512.
9. Waris AS, Khaleeq ur Rehman, Khan FA. Evolution of deterioration in renal function during 30 days of partial ureteric obstruction. Ann KE Med Coll 2001; 7:41-3.
10. Car MC. Anomalies and surgery of ureteropelvic junction in children. In: Walsh PC, Retik Ab, Vaugan ED, Wein AJ, Karousi LR, Novick AC, et al. Campbell's Urology vol.3 . 8th edition Philadelphia: Saunders;2002.p.1995-2006.
11. Hosgor M, Karaca I, Ulukus C, Ozer E, Ozkara E, Sam B, et al. Structural changes of smooth muscle in congenital ureteropelvic junction obstruction. J Pediatr Surg 2005; 40: 1632-6.
12. Ahmad N, Manan A, Akbar A, Chughtai MN. Pyeloplasty: 8 years experience at Lahore and Larkana, Pakistan. Pak J Med Res 2007;46.
13. Yusef H, Saleem, Massoud M. Outcome analysis of peadiatric pyelopalsty as a function of patient's age, presentation and differential renal function. J Urol 1995;154:1889-93
14. Rarive G, leibovitch I, Jonas P, Gold Vasser B. Ureteropelvic junction obstruction, relation of etiology and age at surgical repair to clinical outcome. Urol Int 1994;52:135-9.
15. Braink. Auge, DM, Blean M, Preminger, MO. Uretropelvic junction repair;planning the approach. Atos urol clin 2003;11:141-148.
16. El-Nahas AR, Abou El-Ghar ME, Refae HF, Gad HM, El-Diasty. Magnetic resonance imaging in the evaluation of pelvi-ureteric junction obstruction a all in one approach BJU Int 2007 ;99:641-5.
17. Kausik S, Segura JW. Surgical management of ureteropelvic junction obstruction in adults. Int braz J Urol 2003;21:3-10.

18. Starr NT, Maizels M, Chou P, Brannigan R, Shapiro E. Microanatomy and morphometry of the hydronephrotic "obstructed" renal pelvis in asymptomatic infants. J Urol 1992;148:519-24.
19. Kim DS, Noh JY, jeong HJ, Kim MJ, Jeon HJ, Han SW. Elastin content of the renal pelvis and ureter determines post pyeloplasty recovery. J Urol 2005;173:962-6.
20. Murakumo M, Nonomura K, Yamashita T, Ushiki T, Abe K, Koyanagi T. Structural changes of collagen component and diminution of nerves in congenital ureteropelvic junction obstrution. J Urol 1997; 157:1963-8.
21. Stern JM, Park S, Anderson JK, Landman J, Pearle M, Cadeddu JA. Functional assessment of crossing vessels as etiology of ureteropelvic junction obstruction. J Urol 2007; 69:1022-4.
22. Kajbafzadeh AM, Payabvash S, Salmasi AH, Monajemzadeh M, Tavangar S. Smooth muscle cell apoptosis and defective neural development in congenital ureteropelvic junction obstruction. J Urol 2006; 176:718-23.
23. Yiee JH, Johnson-Welch S, Baker LA, Wilcox DT. Histologic differences between extrinsic and intrinsic ureteropelvic junction obstruction. J Urol 2010; 76:181-4.

AUTHORS

- **Dr. Muhammad Irfan Munir**
  MBBS, FCPS
  Senior Medical Officer Urology Department
  Allied Hospital, Faisalabad
- **Prof. Dr. Zahid Iqbal**
  Professor of Urology (MS Urology)
  PMC, Allied Hospital, Faisalabad
- **Dr. Safdar Hassan Javed**
  MS Urology
  Associate Professor
  Urology Department
  Allied Hospital Faisalabad
- **Dr. G. M. Subhani**
  MS Urology
  Assistant Professor
  Urology Department
  Allied Hospital Faisalabad
- **Dr. Khalid Mehmood**
  FCPS Urology
  Senior Registrar
  Urology Department
  Allied Hospital Faisalabad
- **Dr. Tahir Bashir**
  FCPS Urology
  Medical Officer
  Urology Department
  Allied Hospital Faisalabad
- **Brig. Sajjid Mushtaq**
  M. Phil Histopathology
  Head of Pathology Department AFIP
  Rawalpindi.
- **Dr. Fayyaz Ahmed Malik**
  MCPS, M. Phil Histopathology
  Associate Professor Pathology
  Independent Medical College Faisalabad

Contact Address:

- **Dr. Muhammad Irfan Munir**
  MBBS, FCPS
  Senior Medical Officer Urology Department
  Allied Hospital Faisalabad
  03336701970,drirfan_munir@yahoo.com