Case Collection

A Retrospective Study of Management of Congenital Hydronephrosis, is Early Detection Necessary ?

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Introduction:

Hydronephrosis due to congenital ureteropelvic junction obstruction is one of the most common anomalies seen in childhood. It is defined as an impediment to urinary flow from the renal pelvis into the ureter. Inefficient drainage at this point leads to progressive dilatation of collecting system and a further diminution in the efficiency of pelvic emptying. Initially the muscle of renal pelvis hypertrophies and glomerular filtration diminishes to accommodate of obstructive process. Eventually, when the limit of kidney compromise, the structure of renal parenchyma changes and impaired of function ensues.

It is seen in all ages. In the 1st year, 25% of cases can be detected but ultrasonogrmam can diagnosed perinatally. Relatively few cases are noted after puberty and into adulthood, of those detected aberrant vessel crossing over the pelvi-ureteric junction is the common. The Male: Female ratio is 2:1 with the left side being more common.

The most common cause is obstruction at the PUJ1, 2. The two main causes of PUJ (Pelvis-ureter junction) obstruction are an intrinsic muscular defect causing impaired peristalsis and urine drainage, or an extrinsic obstruction caused by an aberrant or accessory vascular stalk leading to the lower pole of the kidney and crossing anteriorly to the PUJ or upper ureter 3–5, controversies still exist the cause or effect.

More than 90% of PUJ obstructions managed during infancy involve asymptomatic cases, detected by prenatal ultrasonogram. The indications for conservative or surgical therapy of PUJ obstruction are still debatable.

As most of the cases are asymptomatic in early life the role of perinatal sonography is of paramount importance. The pyramid and medullary areas remain sonulucent in the developing kidneys until full maturity of 3 months of age and this makes early detection difficult. Many cases which show mild to moderate (< 2 cm) dilatation of renal pelvis show resolution of hydronephrosis within first 2 years of life. Doppler ultrasonogram with measurement of Resistive Index can aid in diagnosis.

The controversies exist in timing of surgeries in between conservative watchful observation and Pyeloplasty Vs Nephrectomy. Kidneys that provide at least one third of overall renal function can be preserved.

Out of many pyeloplasties, the Anderson-Hyne (Dismembered Pyeloplasy) is considered to be the most time tested.

The purpose of this study is to present our experience with PUJ obstruction of different etiologies managed mostly by Anderson-Hyne Pyeloplasty.

Objective:

To evaluate the causes of Congenital Hydronephrosis, their presentation, the surgical management approached and their outcome.

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Material and Methods:

Total of 06 cases managed between January 2005 to December 2005 were retrospectively studied.

All the cases underwent routine renal function test, Ultrasonography and intravenous urograpy. The patient selectively underwent CT scan, retrograde pyelography and renogram. All the cases were operated and routine follow up was done.

Results:

The patient age varied from 8 years to 40 years. (Mean age of 19.66 years and median age of 16.5 years). Out of them 4 were male and 2 were female, showing the male predominance (66.66%).Four of them were on the right (66.66%), one on the left(16.66%) and one on the both sides(bilateral).

Figure 1. *Non visualized right kidney with normal contralateral kidney.*

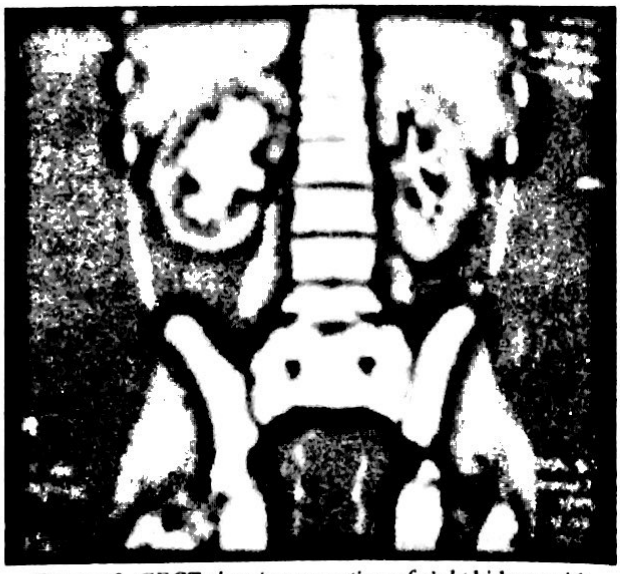

Figure 2. *CECT showing excretion of right* kidney with hydronephrotic changes.

The most common symptoms being loin pain in 3 patients (50%) and the other each case present with recurrent Urinary tract infections, abdominal mass and incidentally diagnosed respectively.

All the cases revealed normal blood urea and creatinine levels. One case had recurrent E. coli in the urine culture and sensitivity. USG of abdomen/ pelvis and IVU (Intravenous Pyelogram) were performed in all cases. The CT scan was performed selectively in 3 of the cases to rule out the other causes and to access the function of the other contralateral kidney.

All patients underwent exploratory retroperitoneal approach under GA (General Anesthesia) with pre operative one dose of intravenous antibiotics and routine catheterization. Four of them were approached with subcostal incision (66.66%) and 2 with (33.33%) 12th rib resection.

Figure 3. *Anderson-Hyne being performed.*

Figure 4. *Aberrant vessel compressing at anterior part of pelvi-ureteric junction.*

Per-Operative Findings:

In the retroperitoneal exploration, 3 (50%) had intrinsic cause of congenital HDN with marked constriction at the PUJ (Pelvi-ureteric Junction) which were sent for the Histopathological examination. One had extrinsic cause (16.66%), an aberrant vessel compressing the Pelvi-ureteric junction. In the other 2 cases (33.33%) the kidneys did grossly hydronephrotic with parenchymal loss resemble the kidney into a bag of water.

Four of them underwent Dismembered Anderson-Hyne Pyeloplasty (66.66%) and nephrectomies were performed in 2 cases (33.33%).

In the intrinsic causes, the constriction parts were excised making sure the healthy end were there for the anastomosis after placing the standard Double-J stent.

One case which had an aberrant vessel compressing the Pelvi-ureteric junction in the anterior part underwent dismembered Pyeloplasty from the posterior with a Double-J stenting, salvaging the kidney and the constricted part was sent for the Histopathological examination.

Two of them had secondary causes (33.33%). One of them had interurteric content at the pelvi-urteric junction and proximal to it there was chronic pyelonephritis leading to severe thinned out cortex not in position of salvaging the kidney, so nephrectomy was performed. The interureteric content was sent for histopathological examination. The other had constriction at the Pelvi-ureteric junction and proximal to it there was a growth at the pelvis obstructing the flow of urine with severe pyelonephritis with thinned out cortex. Nephrectomy was done as the kidney was compromised and the specimen together with pelvi-ureteric constriction was sent for histopathological examination.

Histopathological Examination Report

In the histopathological examination of 4 constricted segments 3 (75%) of them had intrinsic defect of the wall of the ureter leading to adynamic segment and 1 (25%) of them had hypertrophied and hyperplastic smooth muscle cells. All of them had finding suggestive of acute on chronic inflammation.

One of them had interureteric content and the findings were suggestive of hyperplastic fibro muscular tissue and the other had mass in the pelvis and the findings were consistent with inflammatory myofibroblastic changes with hypertrophied smooth muscle at pelvi-ureteric junction.

Follow up

All the patients are on regular follow-up on OPD (Out Patient Door of Surgery) basis. The patients who underwent nephrectomies are maintaining normal renal functions tests.

Four of them who underwent Anderson-Hyne Pyeloplasty, their Double-J Stent were removed after 8 weeks as a day case surgery basis. Their follow-up sonography of showed increase of the cortical thickness (> than 3mm) suggesting the functional gain of the kidneys. In the course of their follow up none of them had symptoms related to urinary tract.

Discussion

The age group of detection of congenital hydronephrosis in early infancy in our set up is very difficult as we do not perform perinatal sonogram and this lead to the late detection of cases. The nephrectomies could have been avoided if detection were early.

The Male: Female ratio is comparable with the existing literature above i.e. 2:1.15. However, right side hydronephrosis is seen more common in our small study, the greater preponderance being on the left side.

The detection of Congenital Hydronephrosis in our group is only on symptomatic patient, the reason being not performed perinatal ultrasonogram as a screening programme which is understandable to the above mentioned literature which said that it is detected more in asymptomatic group16.

The most common causes of congenital hydronephrosis in our group were intrinsic causes with histopathological findings suggestive of defect of wall of ureter with no hypertrophy of smooth muscle.

The follow up period of our study is short (longest being 9 months and shortest being 1.5 months) so the long term results are still awaited.

Conclusion

Early detection of cases can avoid the nephrectomies. Therefore, perinatal and early childhood screening with ultrasonogram is recommended. Any case of the unexplained hydronephrosis should be thoroughly investigated in the line of possible ureteropelvic junction obstruction.

Dismembered Anderson-Hyne Pyeloplasty still holds best in an established case of congenital ureteropelvic junction obstruction where facilities of endopyelotomy are lacking.

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