

A SHORT TERM STUDY ON PELVIURETERIC JUNCTION OBSTRUCTION IN CHILDREN

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CERTIFICATE

This is to certify that this is the bonafide dissertation work done by **Dr.A.RAVIKUMAR, MS.**, submitted for the MCh Paediatric surgery examination held on August 2006 under the title of “**A SHORT TERM STUDY ON PELVIURETERIC JUNCTION OBSTRUCTION IN CHILDREN**” under my guidance and supervision.

Prof. **Dr.RAMESH RATHINAM, MS., MCh**
Professor and Head,
Department of Peadiatric Surgery,
Madurai Medical College,
Madurai.

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INTRODUCTION

Hydronephrosis is defined as dilatation of the renal collecting system as a result of either inadequate drainage or retrograde flow of urine. Pelviureteric junction obstruction (PUJ stenosis) represents 44% of all postnatal causes of hydronephrosis.

Congenital hydronephrosis, caused by pelviureteric junction obstruction, has challenged the ingenuity of paediatric surgeons and remains one of the most enigmatic clinical problems today. Historically, pain, infection, stone and hematuria have been the classic and compelling indications for intervention.

Currently with the increased widespread use of high-resolution real time fetal ultrasonography, antenatal hydronephrosis with minor dilatation of the upper urinary tract is being detected with increased frequency.

The challenge for today's Paediatric surgeons in the management of hydronephrosis is to decide which patients can be observed, which can be medically managed and which require surgery.

MATERIALS AND METHODS

60 cases of hydronephrosis were studied in the department of Paediatric surgery in Madurai medical college, Madurai during the period from July 2003 to September 2005. Out of 60, 41 had Pelviureteric junction obstruction.

All cases were evaluated by recording antenatal history, age, sex, presenting symptoms, signs and if there is any previous treatment.

All the cases were investigated with urine culture, blood urea, creatinine estimation, ultrasonogram abdomen and intravenous urogram. Some were subjected to CT scan abdomen with contrast and Diuretic renogram.

The conventional open Anderson Hynes pyeloplasty was done in 39 cases. All the patients were followed regularly with ultrasonogram and few of them with intravenous pyelogram and diuretic renogram.

AIM OF THE STUDY

1. To analyse the incidence of pelviureteric junction obstruction
2. To analyse the clinical presentation
3. To analyse the various modalities of treatment
4. To analyse the pre and postoperative function of the affected kidney.

REVIEW OF LITERATURE

EMBRYOLOGY

At around the 5th week of intrauterine life, the ureteral bud develops as a diverticulum from the caudal segment of the mesonephric duct near its entry into cloaca. The ureteral bud grows cephalad and penetrates the metanephric blastema resulting tissue induction of the undifferentiating mesenchyme and transforming it into the functioning nephrons of the metanephric kidney.

The ureteric bud undergoes a series of approximately 15 generations of divisions and by 20 weeks of gestation form the entire collecting system that is, the ureter, renal pelvis, calices, papillary ducts and collecting tubules.

At one time it was thought that the ureter forms a solid cord of tissue by the 6th week of intrauterine life and undergoes canalisation that begins in the midureteric segment and that extends bidirectionally.(1) The pelviureteric junction and the ureterovesical junction (UVJ) are the last segments to canalise. More recently, however, it has been shown that bidirectional recanalisation does not occur. Recanalisation occurs only at the middle portion of the ureter.(2)

The membranes that develop during canalisation at the pelviureteric junction or vesicoureteric junction (Chwalla's membrane) may lead to transient

physiological hydronephrosis or hydroureteronephrosis, respectively. A delay in the resolution of these membranes can lead to upper urinary tract dilatation.(1,3)

The ability of the kidneys to make urine and to maintain adequate amniotic fluid volume has a profound influence on the growth and development of the fetus, particularly after the 18th week of gestation. Under the inductive influence of ureteric bud, nephron differentiation begins during the seventh week.

By 20 weeks, when the collecting system is completely developed, approximately one third of nephrons are present, nephrogenesis is complete by 36 weeks.

HISTOLOGY

The renal pelvis is composed of three layers; mucosa, muscularis and adventitia. The inner layer is mucosal and lined by transitional epithelium supported by a lamina. The epithelium is two to three cells thickness in the renal pelvis and four to five cells thickness in the ureter. The epithelium sits on a thin basal lamina which rests in the lamina propria composed of dense fibro connective tissue with prominent elastic fibres.(95)

The muscular layer is composed of interdigitating fibres of smooth muscle separated by strands of connective tissue. These muscle fibres are arranged into an inner longitudinal layer and an outer circular layer.(40)

The adventitia is external to the muscular layer and is composed of fibroelastic tissue, which is continuous with the capsule of the kidney. The ureter has rich blood supply with extensive vascular and lymphatic plexus within the muscularis and lamina propria. Ganglion cells are present within the ureter and supply motor function to the muscularis. Sensory fibers within the muscularis penetrate between the cells of epithelium.

PHYSIOLOGY

Urine produced in the glomerulus flows into the pelvis via the calyces. At physiologic rates of urine production, the calyces and renal pelvic musculature contracts at a frequency greater than that of the upper ureter.

Morita has demonstrated a relative block of electrical activity at the pelviureteric junction during normal diuresis.⁽⁴⁾ At these flow rates; the pelvis gradually fills with a concomitant rise in intrapelvic pressure. This continues until the pelvic pressure exceeds upper ureteric pressure. Urine then flows into the upper ureter. A ureteral contraction then propels the bolus of urine distally while the closed pelviureteric junction proximally blocks back flow and prevents backpressure from the ureteral contraction. Successful conduction of urine from the renal pelvis requires both anatomic patency and undisturbed transmission of peristaltic contractions.

PATHOPHYSIOLOGY

Anatomic Response to Urine Flow Impairment

Parenchymal Response

Dilatation of the pelvis and calyces is the first anatomic response to urine flow impairment. It may lead to significant and long-standing histological damage of the renal parenchyma and changes in the renal function. This damage is related to the degree and duration of the urine flow impairment.

The ultimate response to urine flow impairment is renal atrophy due to programmed cell death called **apoptosis**.⁽⁵⁾ **The SGP – 2-gene** expression located in the adventitial layer of the hilar arteries and intrarenal arterioles is responsible for apoptosis.⁽⁶⁾

Histological Changes

Unilateral ureteral obstruction is characterised by infiltration of renal cortical interstitial macrophages, an increase in cortical transforming growth factor – Beta-1 gene expression. Renal cortical TGF- Beta-1 derived from the infiltrating macrophage, contributes to the formation of the interstitial fibrosis. The release of prostaglandin E2 and thromboxane A2 may be the responsible for the proliferation of fibroblast like cells and mononuclear cells.⁽⁷⁾ The types I, III, V collagen are

found increased levels in the band of interstitium. Type IV collagen is found in the tubular basal membrane of the obstructed kidneys.(8)

Histological Changes in the Contralateral Kidney

1. Immunoglobulin G deposits.
2. Increased in size of the glomerular corpuscle.(9)

Timing of Urinary Flow Impairment

The parenchyma response to urine flow impairment is closely related to the gestational age or the severity of impairment. If it occurs in the early in pregnancy dysplasia will be formed.(10,11) When urine flow impairment occurs in the later gestation, it dilates excretory system without affecting the renal parenchyma.(12,13)

Response of the Ipsilateral Excretory System to Urinary Flow Impairment

Ureteral obstruction leads to dilatation and hypertrophy of the pelvis.

Functional Response to Urinary Flow Impairment

A reduction of the ipsilateral glomerular filtration rate and an increase of the contralateral glomerular filtration rate is the ultimate response to, significant and durable unilateral urine flow impairment.

Ipsilateral Vascular Response to Urinary Flow Impairment

The nitric oxide is the prime response for the initial increase in renal blood flow after acute unilateral obstruction.(14) After the acute phase, the Angiotensin II, Platelet activating factor and Thromboxane A are responsible for decrease blood flow and glomerular filtration rate.(15,16) This vascular changes lead to renal hypoxia, which reflected by an increased level of renal Lactate dehydrogenase in the obstructed kidney.(17)

Ipsilateral Glomerular and Tubular Response to Urinary Flow Impairment

The affected kidneys produce a larger volume, lose more sodium and have lower creatinine clearance. The concentrating capacity is also decreased.(18)

Ipsilateral Pelvic Contractility and Pressure Response to Urinary Flow Impairment

Normal human kidney pelves have uniform peristaltic pressure waves of 1 to 4 mm Hg amplitude and 5 to 10 seconds duration. In patients with mild to moderate hydronephrosis but well-preserved function, the pressure wave amplitude and frequency vary only minimally.(19) When function is poor, however, the amplitude and frequency may drastically change. The baseline pressure of the normal pelvis also depends on the urine flow rate; the normal range is 5 to 25 cm of H₂O.

Johnston noted that most obstructed kidneys maintain a low intrapelvic pressure, usually in the normal range.(20) The ability of the extra renal pelvis to

dilate and maintain physiologically normal pressures may help to preserve the renal parenchyma. This pelvic compliance is effective until a critical volume or capacity is reached. At this point, smooth muscles, elastin, and connective tissue were become overstretched. At and above this volume, pressure may increase sharply.

Koff and Thrall established pelvi -metric curves that correlate pelvic volume and pelvic pressure and show two phases in cases of urine flow impairment ; the accommodation phase (pelvic compliance) and the over distention phase.(21)

Koff (22) describes a pressure dependent flow pattern, seen when intrinsic pelviureteric junction obstruction is present and characterised by increased flow with increased intrapelvic pressure. This intrinsic obstruction is caused by a narrowing or amuscular segment and is associated with the pressure-dependent restriction to urinary outflow. There is a linear relationship between pressure and flow such that as pressure in the pelvis increases, so does flow across the pelviureteric junction.

Koff has also described a volume-dependent flow pattern, seen in extrinsic pelviureteric junction obstruction. In these patients, flow initially increases with increased pressure, but this is quickly followed by an abrupt decrease in flow

caused by increasing pelviureteric junction angulation with intrapelvic volume expansion. As the pelvic volume expands, the pressure also increases, resulting in increased resistance and even more severe obstruction. Flow remains constant or declines.

Contralateral Response to Unilateral Urinary Flow Impairment

Due to some biological events after unilateral obstruction, compensatory hypertrophy occurs in the opposite kidney. (23)

The total number of filtering nephrons however, is decreased in the post-obstructed kidney indicating a significant increase in the single nephron GFR (**SNGFR**) of the remaining nephron units. This is because due to the local production of prostacyclin and PGE₂ and their total effect on the afferent and efferent glomerular arteriolar vascular resistance. It is obvious that these changes, in turn determine the net effect on renal clearance or GFR.

ETIOLOGY

The etiologies for the pelviureteric junction obstruction are:

1. Intrinsic abnormalities.
2. Extrinsic abnormalities.

Intrinsic Abnormalities

This is the most common causes of obstruction. They are usually congenital. The valve like process and polyps may also associate with obstruction at pelviureteric junction. These prevent urine physically from entering the upper ureter from renal pelvis. In this abnormality the pelviureteric junction neither was nor anatomically narrowed and there is no demonstrable extrinsic obstruction.

Histology

Murnaghan noted muscular bundles with abnormal configurations at the site of obstruction.(24) Decreased musculature was noted. All of these are consistent with an aperistaltic segment often associated with replacement of the normal circular musculature with muscle having a primarily longitudinal orientation. In such instances when the renal pelvis is distended, the ureter will elongate and the longitudinal bundles will produce narrowing instead of the normal widening that occurs when circular fibres relax.

Runano-Gil demonstrated that the ureter is the result of the recanalisation of a tube that is closed during early development. They hypothesized that pelviureteric junction may be secondarily deformed or incomplete recanalisation. This theory supported for the hydronephrosis in the fetus that often resolves over time.(1)

Tainio et al have shown the abnormalities of peptidergic innervation with dense innervation of neuropeptide Y and vasoactive intestinal peptide and proposed that these may have a role in intrinsic obstruction (25)

Hanna reported that electron microscopy often reveals disruption of the intercellular relationship between the muscle cells at the pelviureteric junction.(26)

Extrinsic Abnormalities

Aberrant vessels are seen in about one third of cases.

Stephen coined the term ureterovascular tangle, to describe the condition where in the proximal ureter is seemingly angulated and obstructed by the aberrant renal vessels supplying the lower pole of the kidney. (27)A lower hilar segmental vessel may arise from any point along the course of the main renal artery, aorta and iliac artery. In 25% of the cases of obstructive hydronephrosis are associated with this ureterovascular relationship.

This anomaly was uniformly on the left side and the pelvis bulged anteriorly between the lower and middle hilar segmental vessels. The expanding pelvis seemed to angulate the adjoining ureter at pelviureteric junction and hook the ureter over the lower segmental vessels creating a partial obstruction. The angulated ureter is usually bound by fascial adhesions to the pelvis.

During fetal development the maturing kidney ascends to its position in the upper part of the retroperitoneum as the renal pelvis changes its orientation from the anterior to medial. During this stage the kidney acquires its permanent blood supply. The vessels are arranged in a "ladder" pattern from the aorta to the involuting mesonephros, and the maturing metanephros tunnels its way toward an upper retroperitoneal position posterior to these vessels. As the kidney ascends, it sequentially derives its blood supply from the higher vessel and sheds the lower one.(28) It is conceivable that abnormal spatial or temporal progression of renal ascent/ rotation in combination with renal vascular formation may lead to an unfavorable ureterovascular configuration leading to partial obstruction.

Kinks, bands and adhesions are often intraoperative findings even in the absence of inflammation or prior infection. These anomalies cause volume dependent obstruction. As the pelvis distends, the angulation becomes more severe and the ureter may be folded in such a way that most dependent portion of the pelvis does not drain. This anomaly then often becomes an apparent high insertion of the ureter into the pelvis.

CLINICAL PRESENTATION

Pelviureteric junction is the most common site of obstruction in the urinary collecting system.

Incidence

The incidence of pelviureteric junction obstruction is 1 in 1500 of live births. The boys are more commonly affected than girls (65%: 35%). (29) In 60% of cases, the left side is predominate. The bilateral involvement is about 10%. Today, pelviureteric junction obstruction is manifested in less than 19% of neonates with mass abdomen.

Before the advent of prenatal ultrasonography most infants presented with

1. Abdominal mass
2. Hematuria
3. Urinary tract infection
4. Gastrointestinal discomfort (30)

Most of the cases now detected during prenatal ultrasonographic screening. The fetal kidney and collecting system are often detectable by USG scanning as early as 15 weeks of gestation. In those children with detectable abdominal masses on physical examination 50% of masses are genitourinary in origin and of these 40% are secondary to hydronephrosis caused by a pelviureteric junction obstruction. Older children usually present with abdominal discomfort, hematuria following minimal trauma, nausea or vomiting or urinary tract infection. (31) **Dietl's crisis**, which refers to episodes of pain, vomiting, oliguria with abdominal lump, is due to intermittent hydronephrosis.(32)

ASSOCIATED ANOMALIES

1. Multicystic dysplastic kidney
2. Duplication of collecting system
3. Imperforate anus
4. Congenital heart disease
5. VATER association
6. Oesophageal atresia

INVESTIGATIONS

1. ULTRASONOGRAPHY

This is the first radiological investigation of choice in a child suspected pelviureteric junction obstruction. No special preparation is required prior to the study. In ultrasonogram the following information should be obtained. (Fig no.4&5)

1. Degree or severity of hydronephrosis

Anteroposterior diameter of pelvis <12mm-mild, 12-20mm-moderate,
>20mm-severe

2. Size of renal pelvis
3. Calyceal dilatation
4. Renal length, ipsilateral and contralateral kidney
5. Cortical thickness
6. Status of the ureter

7. Bladder wall thickness and bladder volume at the time of study

The USG is simple to perform, noninvasive, easily reproducible. It is important that the child is well hydrated prior to the study. The classical picture seen in pelviureteric junction obstruction is the **Mickey Mouse appearance**.

Dhillon et al have measured the anteroposterior diameter of renal pelvis in the transverse plane in all children with an antenatal diagnosis of hydronephrosis. This has been unique parameter in the management of antenatally diagnosed problems.(33)

He also maintain that children with renal pelvis of anteroposterior diameter <20mm did not come for surgery and progressive increase in the degree of hydronephrosis may precede functional deterioration on renography after several years.

Ransley et al showed that none of the children being followed for asymptomatic hydronephrosis with an AP renal pelvis of <12mm, required a pyeloplasty.(34)

Koff and co workers used compensatory changes in the opposite normal kidney as parameter of obstruction in unilateral hydronephrosis.(35)

On USG the longitudinal scan of normal kidney appears as an oval organ with the echo poor (black) parenchyma and white central echogenic area. The parenchyma includes both cortex and medulla. Central echogenic area contains fibro fatty, vessels, lymphatic and most important of all the pelvicalyceal system.

Normally the pelvicalyceal system is collapsed and is not separated. When there is hydronephrosis, fluid distended pelvicalyceal system is seen on longitudinal scan as separation of central echogenic area by lucency of fluid. This separation will increase with increasing hydronephrosis.

On seeing this, next step will be a coronal scan of kidney where the fluid filled calyces will be seen to join the pelvis. After this, trace the pelvis to continue as the ureter if obstruction is beyond PUJ. Visualizing the proximal ureter on one side of the image the ureter is traced down by tilting the probe to find out the level and cause of obstruction.

Sometimes a vessel crossing the PUJ and causing obstruction can be visualised on colour Doppler study. A calculus at PUJ will be seen as a bright echogenic mass in the lumen with acoustic after shadow.

Hydronephrosis is the most common anomaly diagnosed in utero. With advent of maternal ultrasonography, hydronephrosis is frequently detected in utero and the diagnostic work up is completed shortly after birth. Before the use of maternal sonography, infants would present with urosepsis, abdominal masses, failure to thrive and hematuria. Pelviureteric junction obstruction was the most common cause of hydronephrosis (22%).

Pelviureteric junction obstruction is suspected when there is dilatation of the renal pelvis and no visualisation of the ureter. On ultrasonography the dilated pelvis should also communicate with calices. Lack of communication suggests a multicystic dysplastic kidney.

It is generally agreed that dilation of the urinary tract as seen in ultrasonography, IVP or Retrograde pyelography alone is insufficient to diagnose obstruction of the upper urinary tract. Attempt to use Doppler ultrasound for renal blood flow studies have been made. For example, the renal blood flow decreases with obstruction, a parameter called the resistive Index have been studied.

The resistive index is defined as the peak systolic velocity minus the lowest diastolic velocity divided by the peak systolic velocity.⁽³⁶⁾ Normal kidneys reliably demonstrate resistive index < 0.70 , and obstructed kidneys show higher values. Renal resistive index of less than 82% correlate well with intrarenal pressures of

less than 14cm H₂O . To expand the utility of this study further, furosemide can be administered in the course of this study. An increase of 15% in the postfurosemide resistive index over the prefurosemide resistive index does correlate with the pressure of obstruction with 88% specificity and 76% sensitivity. Thus far, the use of resistive index to define the presence of obstruction has not gained widespread acceptance in paediatric age group.

2. VOIDING CYSTOURETHROGRAPHY

In 9%-14% of patients with pelviureteric junction obstruction have vesicoureteric reflux. Conversely, 1% of patients found to have PUJ obstruction. These associations, albeit low, can affect renal function and change management. Consequently, voiding cystourethrography is the standard of practice for the clinical evaluation of all infants with prenatal hydronephrosis, regardless of age or gender.(37,38)

Recently, several investigators have challenged this standard. They concluded that VCUG should be limited to children with pelviureteric junction obstruction who also have a dilated ureter on ultrasound.(39)

3. INTRAVENOUS PYELOGRAHY

In general IVP is delayed until after the first month of life because renal visualisation may be poor when the renal function is poor. The diagnosis of obstruction is established by IVP when there is dilatation of the renal pelvis, caliectasis and non-

visualization of the ipsilateral ureter. Delayed films are important because with better pelvic filling they tend to confirm the diagnosis. In established cases, thickness of the renal parenchyma is reduced and in severe hydronephrosis, a sliver of opacification known as “**Shell Nephrogram**” or “**Rim sign**” is seen.(Fig No.11) In severe hydronephrosis, a crescentic collection of contrast medium “**Crescent Sign**” is observed in the collecting ducts overlying nonopacified calyces. It indicates that some renal function is present.(40)

The drawbacks of IVP include the necessity of dehydration even in infants, which makes it a relatively risky procedure. Of course, a risk of radiation exposure exists, which can be minimised by limiting number of films taken. Problems associated with contrast media exist, such as nephrotoxicity and anaphylactic reactions. The newer nonionic contrast agents that are currently available can reduce these problems.

4. RETROGRADE PYELOGRAPHY

Retrograde pyelography is rarely needed to diagnose pelviureteric junction obstruction because the diagnosis is ascertained by ultrasonogram and diuretic renogram. The value of retrograde pyelography lies in the preoperative evaluation of the ureter to rule out missed ureterovesical obstruction and to anatomically characterise the pelviureteric junction obstruction.

5. DIURETIC RADIONUCLIDE RENOGRAPHY

Diuretic renogram is a provocative method of evaluating patients found to have dilation of the upper urinary tract in which an obstructive process is suspected. (Fig No.18)

The theoretical basis of this test is two fold: if an obstructive lesion is present, then (a) renal function, more specifically glomerular function may be impaired and (b) a dilated upper urinary tract will retain a larger amount of radionuclide that will not wash out if increased urine flow is generated by the administration of a diuretic.

Three radio pharmaceuticals are primarily used in diuretic renography and their characteristics are linked to their biological activity. Technetium 99m Diethylenetriaminepentaacetic acid (^{99m}Tc -DTPA) and technetium 99m-Mercaptoacetyltriglycine (MAG3) are preferentially concentrated by the kidney and filtered by the glomerulus.(41)

DTPA is neither secreted nor resorbed by the renal tubules, where as MAG3 is secreted by the tubules. Because each is nearly completely excreted, they can be used to estimate differential function and urinary drainage. The third agent, Tc99 dimercaptosuccinic acid (^{99m}Tc DMSA), is tightly bound to renal

tubular cell and is therefore, useful for the detection of differential renal function and clinically significant cortical lesions such as renal scars.

In general, neonates and young infants are placed supine for the study. Ideally, the child should be well hydrated because relative dehydration prolongs parenchymal transit and delays urinary excretion.

MAG-3 or DTPA is injected intravenously as a bolus. The paediatric dose of MAG-3 is 50mcu/ kg. (43) Subsequently, 4- second posterior images are recorded with a high-resolution collimator. One minute after injection, images of the kidneys are obtained each minute.

Normally, the renal parenchyma is well visualised during the first minute; by 2 (or) 3 minutes, activity seen in collecting system and by 6 to 9 minutes the bladder is visualised.

The differential renal function of each kidney is calculated between 60 and 180 seconds after injection of the tracer, which represents parenchymal transit and reflects glomerular function. On computer images, region of interest (ROI) are selected from each kidney and background activity is subtracted.

The activity within each kidney is expressed as a percentage of the total renal counts, and this differential activity is used to compute differential renal function.

To determine whether significant obstruction is present, furosemide is administered intravenously. There are 3 variations in the furosemide administrations.

1. F +20 - furosemide is injected 20 minutes after the injection of tracer.
2. F – 15 - furosemide is injected 15 minutes prior to the tracer.
3. F – 0 - furosemide is injected at the beginning of the study

The most commonly used protocol for diuretic renogram is that in which the furosemide is administered 20 minutes after tracer injection the so-called F+20 protocol. The dose of furosemide in paediatric is 1mg/kg in infants, 0.5mg/kg in children aged 1-16 years.

Furosemide is a potent loop diuretic that induces acutely increased urine flow, reaching maximal effect 15 to 18 minutes after intravenous administration. The furosemide is injected slowly over 3 minutes. Images are acquired for at least 20 minutes following injection.

At completion of the study, a static upright post void image is obtained, especially if the study has been performed supine and persistent tracer pooling seen within the renal pelvis.

Image acquisition must be carried out for at least 20 minutes following furosemide injection to avoid missing flow- dependent intermittent obstruction the so called “ **beer drinker’s kidney**”.

The renogram curve in this setting may appear to fall normally. In response to furosemide early on, but, as urine flow rates reach maximum, the curve again deflects upward in an obstructive pattern. This produces the “**delayed double peak**” pattern – **Homsy’s sign** of intermittent obstruction.⁽⁴²⁾ Premature termination of the test may result in missing this second upward curve deflection. In these patients, a follow up F – 15 renogram is helpful in confirming the true obstruction. ⁽⁴³⁾. F - 15 renography can be used in patients in whom the diagnosis of significant obstruction is equivocal by F + 20 protocol.

A combined approach has also been recommended in patients in whom obstruction is clinically suspected but in whom most of the tracer activity has drained from the collecting system at the end of the F + 20 renogram. In such instances, a repeat injection of the same dose of tracer is immediately made and another 20 minutes study acquired.

The additional information, which can be obtained, includes clearance half time of the tracer from the pelvicalyceal system, which is used by some workers as a parameter to diagnose obstruction.

The renogram curve

Diuresis renography was pioneered by **PH 'O' Reilly (1978)** and associates as a means of distinguishing between obstructive and non obstructive dilatation.(44)

The characteristics of the uptake and drainage curves fall into four patterns;

Type 1: Normal uptake with prompt washout.

Type 2: Rising uptake curve, no response to diuretic.

Type 3a: An initially rising curve, which falls rapidly in response to injection of lasix.

Type 3b: An initially rising curve, which neither falls promptly

Following the injection of lasix nor continues to rise.

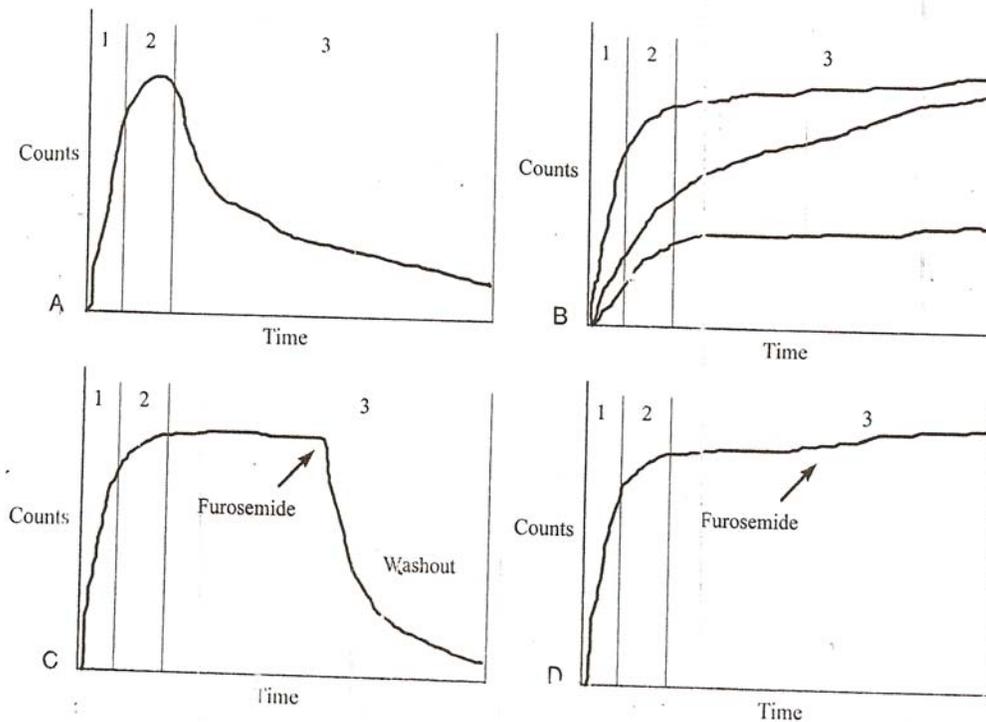
This renogram pattern was defined by O'Reilly and

Colleagues as equivocal and much of their subsequent

work has been aimed at eliminating the type 3b curve

by modifying the technique and timing of lasix administration

The renogram curve



- A. Normal curve shows the three renogram phase.
- B. Abnormal curves. These curves are typical for obstructive or nonobstructive dilatation with accumulation of tracer in the collecting system. Impaired renal function with cortical retention of tracer can produce a similar appearance, although the peak parenchymal activity will be lower than normal.(bottom curve)
- C. In non obstructed collecting system, there is prompt washout of pelvicaliceal activity after intravenous furosemide.
- D. In case of obstruction or impaired renal function,poor or no washout is seen.

The renogram curve is a time activity curve describing the transit of tracer through the kidney. The curve is obtained by placing a computer assisted region of interest over the whole kidney or cortex obtaining the counts in the ROI for each period of data acquisition and plotting these counts as a function of time.

The renogram curve is often divided into the period of tracer appearance; tracer extraction and tracer elimination denoted as phase 1, 2 and 3 respectively. Tracer appearance describes the period of blood flow beneath the detector; tracer extraction is proportional to renal plasma flow or glomerular filtration rate according to used tracer. The curve peaks when tracer exits from the kidney at the same rate it is entering the kidney. Normally the intrarenal transit of tracer is less than 5 minutes.

The excreting part of the curve is measure of drainage from the pelvis. Various quantitative parameters are available but the simple one is measuring the half time ($T_{1/2}$) of excretion. It is generally recognized that a $T_{1/2} < 10$ minutes is considered normal, a $T_{1/2}$ between 10-20 minutes is considered indeterminate and a $T_{1/2}$ of >20 minutes is likely caused by an obstruction.

However, interpretation of the $T_{1/2}$ should not be attempted without evaluation of the dynamic images, renal curves and other quantitative factors. The

technique was also compared with other modalities like perfusion studies described by Whitaker where 85% correlation was obtained. When compared with morphological studies (**Goslings and Dixon 1978**) the correlation rate was 88%.

Factors affecting diuretic renography in the neonate are

Renal maturity	Volume of urine in the bladder
Renal functioning	Outlined regions of interest
Hydration status	Patient position
Type and dose of tracer	Patient movement
Dose of diuretic	Capacity of upper tract
Timing of diuretic administration	Severity of obstruction
Vesicoureteric reflux	Site of obstruction
Method of data interpretation	

It has become the procedure of choice in the evaluation of suspected obstruction because it is relatively noninvasive, providing information about differential function and reliably diagnosing functional obstruction.

Glomerular filtration can be measured by determining the rate of its removal from the blood. The ^{99}Tc emits a 140Kev gamma ray with no harmful beta emission. The estimated absorbed dose by the kidney is 0.04 mrad and total body absorption is 16 mrad, which is lesser than intravenous pyelogram..

To reduce inter institutional difference the society for fetal urology and society of nuclear medicine council proposed a uniform methodology for performing the diuretic renogram called **well - tempered renogram**.⁽⁴⁵⁾

To reduce the likelihood of immature renal function interfering with interpretation, infants should be older than 2 weeks. Oral hydration is begun 2 hours before study. Intravenous hydration of 15ml /kg over 30minutes is started as a bolus 15 minutes before injection of the isotope followed by maintenance fluid at a rate of 200ml /kg/24hours. The bladder is catheterised throughout the study.

Diuretic renogram is well tolerated, easily repeatable and appropriate for paediatric patients. It should be done after 4-6 weeks of life.⁽⁴⁶⁾

6. CT SCAN

CT scan has been used to diagnose pelviureteric junction obstruction in children, especially in association with the abdominal trauma. CT scan, like IVP studies, shows the dilatation of the kidney and collecting system well, and it may be used to estimate the differential renal function by measuring the cortical thickness.(Fig No.9&13) CT angiogram, Helical CT scans are used to evaluate the presence of lower pole vessels preoperatively.

7. MAGNETIC RESONANCE IMAGING

New developments in MRI technology have made it possible to image kidneys while assessing intracellular metabolic parameters independent of blood flow and tubular function. Relative high cost and the noise during the procedure limit the routine use of MRI for evaluating urinary obstruction in children.(Fig No.15)

8. ENDOURETERAL SONOGRAPHY

Bagley and co workers have used endoureteral sonography in the evaluation of the obstructed pelviureteric junction.(47) Catheter based ultrasound probes with single crystal transducers (12.5 Mhz/ 20Mhz) are available in size ranging from 3.5 F to 6.2 F. The transducer is densely radio opaque and can be seen fluoroscopically. The device can be used in conjunction with standard endoscopic equipment. It can provide accurate information of pelviureteric junction anatomy. It can visualise vessels adjacent to the PUJ/ ureter; define the presence of high ureteral insertion including the direction, length and thickness of the septum.

With increasing popularity of endoscopic treatment techniques, this modality may ultimately provide the key to the selection of the optimal therapy for the obstructed pelviureteric junction.

The obstructed PUJ has three distinct patterns seen sonographically. The area of the PUJ can be quite narrow, with a stenotic segment ranging from 1 or 2 to several millimeters.

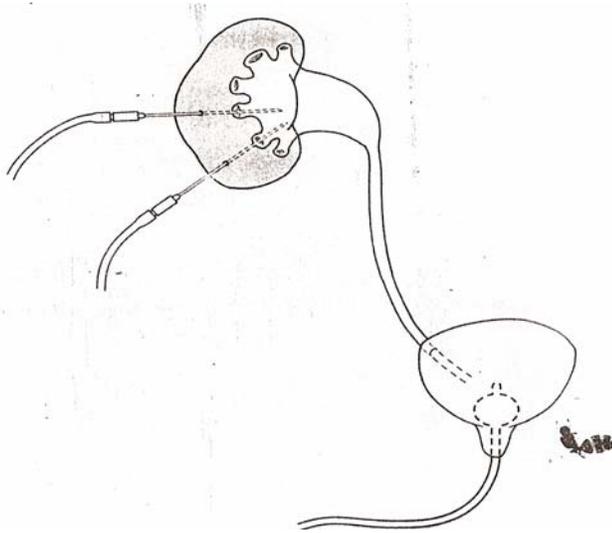
This can be the only finding or may be associated with the other two patterns of crossing vessels or high insertion, as the presence of one pattern does not exclude the others.

9. PRESSURE PERFUSION STUDY

Whitaker's test was considered the gold standard for the evaluation of upper urinary tract dilatation.⁽⁴⁸⁾ It provides urodynamic evidence of a mechanical obstruction of the upper urinary tract at a given flow rate. With the advent of the diuretic renogram and some of the newer radiopharmaceutical agents, the Whitaker test is not often utilised clinically.

Whitaker test is performed with the patient placed on a fluoroscopy table in the prone position. Before the patient is positioned prone, a bladder catheter is placed and connected to a pressure transducer for continuous monitoring of intravesical pressures with changes in renal pressure. A renal cannula (18-gauge) is then inserted and connected to a pressure transducer. A combination of saline solution and contrast material is administered by way of the renal cannula at a rate of 10 ml/min. Bladder pressure is monitored throughout the

procedure, and its relationship to changes in renal pressure can be significant. Contrast material is given along with the saline solution, making fluoroscopic monitoring of the anatomic site of the obstruction possible. **Whitaker's test**



Results are separated into three categories

1. Pressure less than 15 cm H₂O = non obstructed
2. Pressures of 15 to 22 cm H₂O = equivocal
3. Pressures greater than 22 cm H₂O = obstructed

Because the diuretic renogram is noninvasive, easily reproducible and provides quantitative evaluation of the split and total renal function with minimal radiation exposure, it is clinically utilised more today than the Whitaker test. However, when there is extreme upper tract dilatation or poor renal function or both, precluding an adequate diuretic response the Whitaker's test may still have clinical utility.

10. URINARY MARKERS PELVIURETERIC JUNCTION OBSTRUCTION

1. Urinary Beta 2 Microglobulin

Disruption of proximal tubular integrity leads to increased urinary concentrations of beta-2-microglobulin (B₂M), which normally reabsorbed from the tubular lumen via phagocytosis and lysosomal digestion.⁽⁴⁹⁾

An increase in urinary concentrations of B₂M may indicate tubular dysfunction as a result of the obstructive insult. Functionally significant obstruction and recovery from obstruction may be determined by following the urinary concentration of B₂M.

The potential for B₂M to be a marker for significant obstruction is quite appealing; however, the determination of its levels in obstructed kidneys is not routine, and many different insults other than pelviureteric junction obstruction can lead to increased levels of B₂M in the urine. In addition, the immaturity of the nephron and the high fractional excretion of water in neonates contribute to elevated B₂M levels in the absence of any identifiable renal stress.

2. N-Acetyl- β - Glucosaminidase

N-acetyl- β -glucosaminidase (NAG) is a tubular lysosomal enzyme present in the urine of children who have various renal diseases. This is currently experimental.⁽⁵⁰⁾

In rats with experimental partial ureteral obstruction, the urinary concentration of NAG increases in the first 2 weeks of obstruction and decreases with the relief of obstruction.

Urinary biochemical markers of renal damage sometimes may aid the diagnosis of clinically significant urinary obstruction.

The assessment of urine for growth factors (eg, epidermal growth factor [EGF], platelet-derived growth factor [PDGF], TGF β , cytokines, p53, p21) and vasoactive substances may be an important adjunct in evaluating obstructive uropathy in the future.⁽⁵¹⁾

TREATMENT

Goals

The goals of the treatment of pelviureteric junction obstruction are to improve urine flow to prevent further parenchymal damage and to alleviate

symptoms when they exist. There are four therapeutic approaches available for the treatment of PUJ obstruction

1. Conservative management
2. Temporary diversion of urine (percutaneous nephrostomy)
3. Surgery or endoscopic treatment
4. Fetal surgery or urinary diversion

Conservative Management

Conservative management of pelviureteric junction obstruction is justified in most of the cases during the first year of life. Three conditions are required to treat a unilateral pelviureteric junction obstruction conservatively.

1. The asymptomatic child
2. The pelvic dilatation should be stable or decrease on repeated ultrasonogram
3. The relative function on repeated isotopic studies should be stable or increase.

Temporary Diversion

Temporary diversion of the urine is indicated in infants with severe unilateral pelvic dilatation and poor relative function. The percutaneous nephrostomy is recommended for 3 to 4 weeks, followed by reassessment of relative function. At this point, if the function is improved, pyeloplasty will be considered or the function remains poor, nephrectomy should be discussed. In case

of bilateral dilated pelvis, unilateral nephrostomy usually improves drainage from both kidneys.

Surgical or Endoscopic Treatment

Indications

1. A symptomatic pelviureteric junction obstruction
2. Declining function of the dilated kidney
3. Increasing pelvic dilatation
4. Bilateral, moderate to severe dilatation of the pelvis

Surgical Treatment

PYELOPLASTY

Historical review

The first PUJ repair was performed by Trendelenburg in 1886. The first successful pyeloplasty is credited to Kuster in 1891. In 1894 Fenzer applied the Heineke- Mickulicz principle to reconstruct the pelviureteric junction. The dismembered procedure of Kuster was modified by Nesbit in 1949 and further modified by Anderson and Hynes by spatulating the ureter and excising the redundant pelvis.

The flap procedure was introduced by Schwyzer in 1823. This is further modified by Foley who described the Y-V plasty in 1937. This technique was best

applicable to cases of high insertion and unsuitable for pelviureteric junctions, which were dependent.

Four criteria for success in the repair of a pelviureteric junction obstruction were defined by Foley in 1937 as follows (52)

1. Formation of a funnel
2. Dependent drainage
3. Water tight anastomosis
4. Tension free anastomosis

The following procedures are available to treat pelviureteric junction obstruction

1. Dismembered Procedures

- Open Anderson Hynes pyeloplasty
- Laparoscopic pyeloplasty

2. Flap Procedures

- Foley Y plasty
- Spiral flap
- Scardino Prince Vertical flap pyeloureteroplasty

3. Ureterocalycostomy

4. Nephrectomy

Dismembered Procedures

Dismembered procedures involve excision of pelviureteric junction with an anastomosis of the upper ureter to the dependent renal pelvis. Success rate of this procedure ranges between 90-95%, with failure often due to fibrosis secondary to urinary extravasation or to poor tissue handling.

The **Anderson-Hynes pyeloplasty** has become the most commonly employed "open" surgical procedure for the repair of pelviureteric obstruction.⁽⁵³⁾The principal reasons for the universal acceptance of the dismembered pyeloplasty are

1. Broad applicability, including preservation of anomalous vessels
2. Excision of the pathologic pelviureteric junction and appropriate repositioning
3. Successful reduction pyeloplasty.

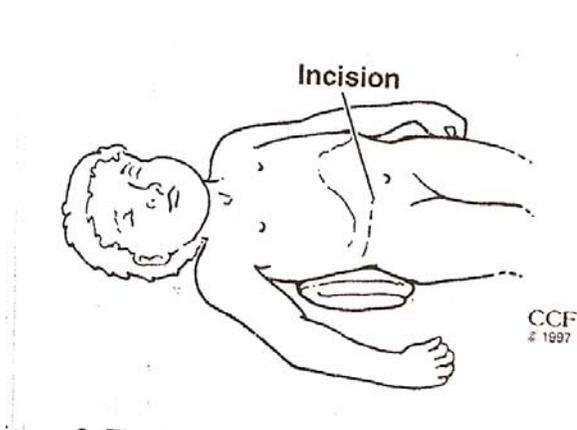
This operation is generally easy to perform and can be accomplished by a number of surgical approaches including

1. Anterolateral extraperitoneal approach
2. Posterior lumbotomy approach
3. Flank approach

Anterolateral extraperitoneal approach

Anderson-Hynes dismembered pyeloplasty, as performed through an anterolateral approach, is as follows (Fig No.20)

1. The anterolateral incision is a muscle-splitting incision that is made with the patient supine and a roll placed transversely beneath the patient to elevate the flank.

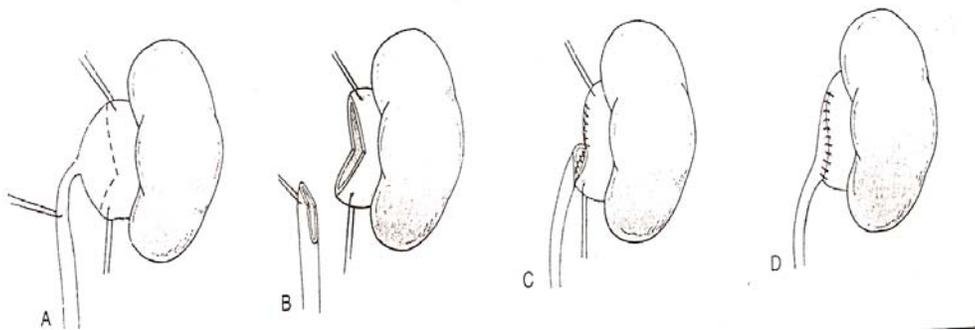


2. Each muscle layer encountered is split in the direction of the muscle fibers until Gerota's fascia is identified by sweeping the peritoneum medially. The fascia is then incised posteriorly over the lateral aspect of the kidney.
3. The renal pelvis is identified by medial retraction of the peritoneum and lateral traction of the kidney.
4. Anterior exposure is usually better when a dismembered pyeloplasty is being performed. Once the ureter and pelviureteric junction are identified, a traction suture is displaced anteriorly through the proximal ureter to minimise subsequent handling.

5. The area of pelviureteric junction is dissected free to allow for a clear area in which to perform the anastomosis. Traction sutures may be placed in the renal pelvis superiorly, medially, laterally, and inferiorly to the pelviureteric junction. Once adequate ureteral length is confirmed and the pathology of pelviureteric junction identified, the ureter can be transected at this level.
6. The ureter is spatulated on the side opposite to the traction suture using Potts tenotomy scissors. The distance over which the ureter is opened is variable, until healthy ureter is encountered, which springs open when forceps are placed into it.
7. A portion of pelvis is excised. It is better to leave too much renal pelvis than too little, especially when resecting along the medial aspect of the renal pelvis. The ureter and renal pelvis are aligned to ensure that the anastomosis can be accomplished without tension. If a nephrostomy tube is to be used, it is placed at this time. An inferior calyx is chosen, preferably where the overlying parenchyma is not too thick.
8. The anastomosis is started by placing the first suture at the apex of the "V" in the ureter and into the tip of the inferior pelvic flap. As the suture is tied down, the ureter and renal pelvis are brought together to minimise tension on the repair. A small feeding tube is placed into the ureter; it can be used to stabilize the ureter during the anastomosis. A "no-touch" technique is employed with the ureter to minimise trauma and edema to the ureteral

- tissue. The area of the initial anastomosis is critical to ensuring a watertight closure.
9. Before the repair is completed, the renal pelvis is irrigated to remove any blood clots or debris that could obstruct the pelviureteric junction. A drain is placed adjacent to the repair and brought out through a separate stab wound.
 10. The kidney is returned to its native position, and perinephric fat if available, is placed over the anastomosis. Wound closed in layers.
 11. The transanastomotic stent removed usually after 48 hours. The drain removed on 4th day.

Anderson-Hynes pyeloplasty



In the following situations, postoperative temporary nephrostomy is advisable.

1. Inflamed renal pelvis (prior nephrostomy, presence of calculus, infection)
2. Thin or hypoplastic ureter.
3. Operative difficulties.

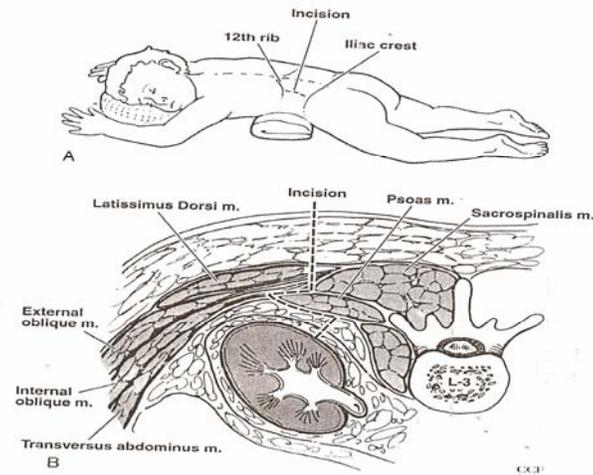
4. Poor renal function.
5. Solitary kidney or bilateral disease.
6. Transabdominal pyeloplasty.
7. Long segment stricture or distal ipsilateral pathology (VU reflux and when PU junction edema anticipated)

Posterior lumbotomy approach

Posterior lumbotomy approach is more commonly used in infants, small children and lean older preadolescent with normally located operative kidney.⁽⁵⁴⁾ The use of muscle splitting rather than cutting makes it almost a minimal invasive procedure. The location of the incision is posterior and in the crease line has a cosmetic advantage.

Procedure

The child is placed in a prone position with a roll under the chest and midthigh regions. A transverse skin incision is made just under and parallel to the 12th rib, with one third of the incision over the paraspinal muscle and two third lateral to the skin. Scarpa's fascia is sharply incised and a vertical incision is made through the lumbodorsal fascia (posterior lamella).



The lateral edge of the lumbodorsal fascia is elevated, and the sacrospinalis muscle is medially retracted. An incision is made through the middle and anterior lamella of the lumbodorsal fascia, taking care not to injure the ileohypogastric nerve. The quadratus lumborum muscle is retracted, exposing Gerota's fascia beneath the paranephric fat, and then this fascia is opened.

The renal pelvis is identified, and several holding stitches are placed in the pelvis. The ureter is identified, a holding stitch is placed in the ureter, and the surgeon proceeds with the dismembered pyeloplasty as usual manner.

After pyeloplasty, a single muscle fascia layer bringing the lumbodorsal fascia back together again does closure.

The bilateral procedure can be done successfully under same anesthesia without position changes or redraping. Pain is minimal, morbidity reduced and mobilisation is almost immediately.

Flank Approach

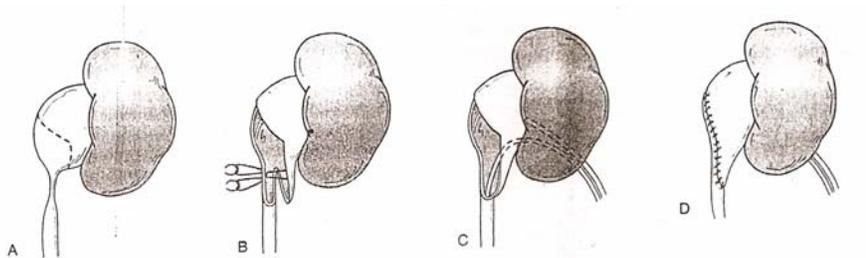
The patient is placed over the kidney rest in a flank position; the kidney rest is elevated and the operating room table is flexed. The skin incision is made off the tip of the 12th rib, or, if necessary, a supracostal 12th rib incision is made.

The external oblique and latissimus dorsi muscles are divided. Next the internal oblique and serratus posterior inferior muscle are divided. The transversalis muscle is often thin and can be divided with digital dissection. The peritoneum is identified and retracted medially.

Gerota's fascia is then encountered and opened longitudinally to gain exposure to the perinephric space. After identification of the renal pelvis and the ureter, a dismembered pyeloplasty can be performed as described earlier.

Flap Procedures

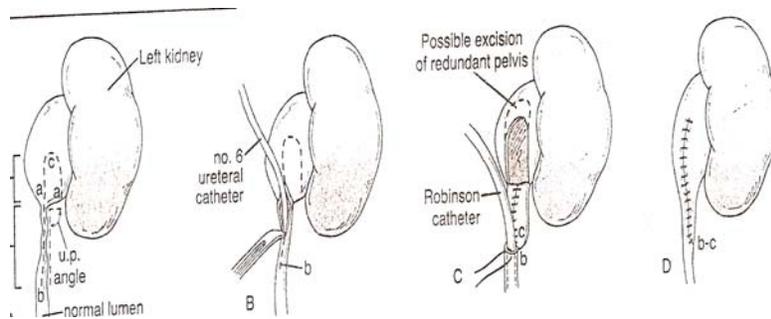
Culp and DeWeerd's spiral flap (55) can be used for patients with a dependent pelviureteric junction and a relatively long area of proximal ureteral obstruction.



The spiral flap is outlined with its base situated obliquely on the dependent aspect of the pelvis. The base of the flap is positioned anatomically lateral to the pelviureteric junction and should lie between the ureteral insertion and the renal parenchyma. The flap is then spiraled posteriorly to anteriorly or vice versa. The medial line of incision through the flap is carried down completely through the obstructed proximal ureteral segment into normal caliber ureter.

The length of the flap is determined by the site of the apex, which in turn should be a function of the length of proximal ureter to be traversed. In order to insure vascular integrity of the flap, however, the ratio of flap length to width should not exceed 3-1. The flap is developed with fine scissors and the apex then rotated down to the inferior most aspect of the ureterotomy. The anastomosis is completed over an internal stent, using fine absorbable sutures.

Scardino and Prince described a vertical flap that can be used in the situation of a dependent pelviureteric junction with a large, square-shaped extrarenal pelvis. (56)

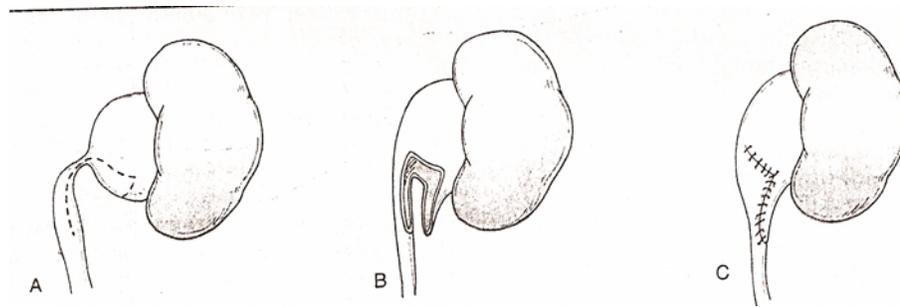


In contrast to a spiral flap, however, the base of the vertical flap is situated horizontally rather than obliquely between the pelviureteric junction and renal parenchyma. The flap itself is formed by a convergence of two straight lines from the base vertically to the apex on either the anterior or posterior aspects of the renal pelvis.

As for the spiral flap, the height of the apex determines the length of flap obtained which again is a function of the length of proximal ureter to be bridged. Medially, the ureterotomy is carried through the proximal ureter completely through the strictured area, continuing a few millimeters into normal caliber ureter. The apex of the flap is rotated down and joined to the inferior most aspect of the ureterotomy. The flap is closed over an internal stent by approximating the edges with fine absorbable suture.

Foley -Y- Plasty

This is useful technique when the obstructed segment is more than 2cm in length a flap of renal pelvis is rotated inferiorly to restore ureteral caliber.(52)



In this procedure, the limbs of the Y are widely separated one on the anterior and one on the posterior aspect of the renal pelvis. The laterally placed portion of the pelvis then drops inferiorly. The lateral border of the ureter is opened longitudinally through the pelviureteric junction to at least 1cm below the obstruction where the ureteral caliber is normal. The midportion of the pelvic flap is sutured to the inferior margin of the ureterotomy converting the incision to a V.

Ureterocalicostomy

This procedure generally reserved for situations in which dependent renal drainage cannot be established by conventional techniques. It is primarily used for horseshoe kidney, fusion anomalies and in the repair of secondary pelviureteric junction obstruction. The inferior calyx is more dependent than pelvis in the Horseshoe kidney and it may be a more physiologic procedure than pyeloplasty.

According to **Hawthorne et al** cardinal features of a successful ureterocalicostomy should include amputation of enough lower pole cortex to free

the ureter from entrapment by contracting fibrosis of the renal cortex. (57) There should be no tension on the anastomosis and there should be no renal parenchyma below the level of the repair.

Nephrectomy

Because the recovery potential of the kidney is greater in children, extreme conservation is justified. Salvage pyeloplasty should be considered as renal function shown in renal scintigraphy can recover.(58) During surgery, the renal cortex should be assessed. The severe cystic dysplasia is an indication for nephrectomy, otherwise every effort be made to salvage the kidney.

In the bilateral anomalies, unilateral pyeloplasty on the most severely dilated pelvis is usually recommended. Unilateral pyeloplasty often improves the dilatation of the contralateral side as well. If the dilatation of the contralateral side does not improve, pyeloplasty should then be considered. If nephrectomy is considered on one side, the pyeloplasty should precede this.

ENDOSCOPIC MANAGEMENT OF PELVIURETERIC OBSTRUCTION

Historical review

The idea of endopyelotomy was conceived in 1909 by **Albarran** in France describing the technique of incising the narrowed pelviureteric junction as **URETEROTOME EXTERNE** in which a stent was left in.⁽⁵⁹⁾ The incised ureter heals over time, which is something akin to internal urethrotomy in stricture urethra. **Davis 1943** popularised this technique as intubated ureterostomy in which few loose stitches were placed.⁽⁶⁰⁾

Wickham and Miller first described a percutaneous approach to incising the pelviureteric junction termed pyelolysis. Subsequently **Arthur smith** popularized this technique renaming it endopyelotomy.^(61,62)

With the advent of endourological techniques the following minimal invasive procedures are being applied to pelviureteric junction obstruction

1. Antegrade nephroscopic endopyelotomy
2. Retrograde ureteroscopic endopyelotomy
3. Balloon dilatation with cutting device

1. Antegrade Endopyelotomy

Open surgery has been the optimal therapy for pelviureteric junction obstruction, and the dismembered pyeloplasty remains the gold standard.

Percutaneous techniques were introduced in the early 80's for the management of nephrolithiasis. Subsequently in 1983 reports began to appear in the literature for endoscopic treatment of pelviureteric obstruction. Success rates vary from 50% to 95% and remain inferior to that of open pyeloplasty.⁽⁶³⁾ Success appears to be dependent on correct patient selection rather than the type of operation.

Patient Factors

1. Age

Neonates and infants may not be suitable candidates for endourologic management. In addition to technical difficulties, the radiation exposure may be extensive. Children in the adolescent age group are more suitable as the anatomy and caliber of the ureter is similar to the adult urinary tract.

2. Hydronephrosis

Van Cangh ⁽⁶⁴⁾ in his series of adult patients found the success rate of endopyelotomy fall from 95% to 77% in the presence of massive hydronephrosis.

3. Crossing Vessels

Van Cangh reported the success rate of endopyelotomy was only 42% in the presence of crossing vessels and dropped to 39% in the presence of both crossing vessels and massive hydronephrosis.

4. Primary versus Secondary PUJ Obstruction

A review of most recent series including that of **Van Cangh** and co workers, it appears that secondary UPJ obstruction responds better to antegrade pyelotomy.

In older child or adolescent patient with primary pelviureteric junction obstruction and favorable factors (e.g. good ipsilateral renal function and only mild to moderate hydronephrosis), endopyelotomy is the preferred procedure. Endopyelotomy is the initial therapy for secondary PUJ obstruction in all-pediatric patients

Endopyelotomy

The pelviureteric junction can be incised with various devices.

1. Cold knife.
2. Electrocautery.
3. Laser – Nd YAG / KTP / Holmium

The UPJ is assessed to determine whether it is wide enough to accommodate subsequent instrumentation. If it is stenotic it can be dilated using a balloon.

The Incision

This is made on the lateral side and extends down the ureter for a centimeter beyond the area of narrowing. Proximally it extends 1-2 cm up the pelvis and should go through all layers till the retroperitoneal fat is visualised. Either a graduated or a standard stent is passed down the guide wire into the bladder. A nephrostomy tube is placed in the pelvis and remains for 48 hours. A nephrostogram is usually performed prior to removal and the stent is left indwelling for 3-6 weeks.

2. Retrograde Ureteroscopic Endopyelotomy

A 5 or 6 F indwelling ureteral stent is placed 1 to 2 weeks prior to planned endopyelotomy. This stent drains the obstructed kidney and stabilises renal function. This maneuver facilitates passage of the ureteroresectoscope and allow for the preoperative evaluation of stent intolerance. Under flouroscopic control retrograde pyelography indicates the level and extension of pelviureteric narrowing using ureteroscope of appropriate size according to size of ureter. The PUJ narrowing is incised at the lateral position (8to9 0'clock on the right, 3to4 0'clock on the left) and stent is left indwelling. Incision at the lateral position minimises the risk of injury to any possible crossing vessels. The success rate is 75% in primary and 82% for secondary PUJ narrowing.(65)

3. Retrograde Balloon Cautey Incision of the Pelviureteric Junction (Acucise Device)

A special balloon catheter with an inbuilt cutting device is used in this technique. Once the waist disappears (after 10 minutes of balloon dilatation) cutting current is activated for 3 seconds to get the desired endopyelotomy. Overall success in this method is around 78% and hospital stay is around 1 day only. In this method the retrograde balloon is used to define the area of stenosis and to carry the cutting wire into the area to be incised. It is designed to accept a maximum of 2.5ml of fluid. The electrically active surface on the cutting wire is 2.8cm in length and 150 μ m in diameter.

Balloon inflation with contrast indicates the narrowed PUJ segment by presenting as a waist. Then the cutting device which is positioned across the PUJ in such a manner that it points posterolaterally, is activated by cutting current for 3 seconds, so as to cut the stenosed PUJ precisely but surely in one plane only avoiding the vessels.

After completion of the incision, retrograde pyelography is performed through the retrograde balloon catheter to confirm extravasation at the incision site. The ureteral stent should be kept for a period of 6 weeks postoperatively.

Patients return 4 weeks following stent removal for postoperative intravenous pyelography to confirm efficacy of the endopyelotomy.

Postoperative bleeding, necrosis of ureter, urinoma, hematoma and urinary tract infections are the frequent complications of this procedure.

Advantages of this technique include

1. Short operating time.
2. Ability to perform it as a day care procedure.
3. No special instrumentation required. The balloon can be passed through a standard cystoscope.
4. The device can also be used through an antegrade percutaneous nephrostomy tract.

4. Balloon Dilatation / Endoburst

The advantages of this procedure are similar to that of the acucise device; however one cannot incise the pelviureteric junction. Cystoscopy is performed and a 0.035 – inch guide wire passed up the affected ureter and coiled in the renal pelvis. An appropriately sized dilating balloon catheter (12F to 24F) is passed over the guide wire and positioned across the PUJ fluoroscopically. The balloon is inflated for 3 minutes and the PUJ ruptured. Extravasations of contrast confirms

the rupture and a 6 to 8 F stent is positioned over the guide wire on removal of the balloon catheter.

Tan reported on the early results of balloon dilatation for primary pelviureteric junction obstruction in 10 children. The age range was 3 months to 9 years and at 22 months follow up had a success rate of 70%.⁽⁶⁶⁾

Pros and cons in endopyelotomy

The concern in high insertion pelviureteric junction endopyelotomy may not yield good results. It is not a suitable method in vessels crossing pelviureteric junction where there can be torrential bleeding. **Lingman et al** 1993 reported antegrade approach with 100% success when followed for one year. **Figenshau's** 4 years follow up shown 100% success. ⁽⁶⁷⁾

In children with secondary pelviureteric junction obstruction, the results are more encouraging. It approaches nearly 100% as against available (70 to 95% success) results in primary pelviureteric junction obstruction.

Endopyelotomy has proven to be preferred method in failed pyeloplasty where the success rate is nearly 95 to 100%.

For primary pelviureteric junction obstruction with pelvic size less than 60ml volume, if vessel crossing pelviureteric junction is ruled out by spiral CT scan or endoluminal US scan, definitely endopyelotomy can be attempted.

LAPAROSCOPIC PYELOPLASTY

The latest technique is laparoscopic pyeloplasty where dismembered pyeloplasty is done as in open with the advantage of minimal invasive nature. Laparoscopic pyeloplasty was first reported by **Schuesler**.⁽⁶⁸⁾

Approach of laparoscopic pyeloplasty

The exposure of pelviureteric junction can be done either by

1. Transperitoneal approach
2. Retroperitoneal approach

By transperitoneal approach pelviureteric junction is exposed by mobilising the colon. But problems in transperitoneal approach are

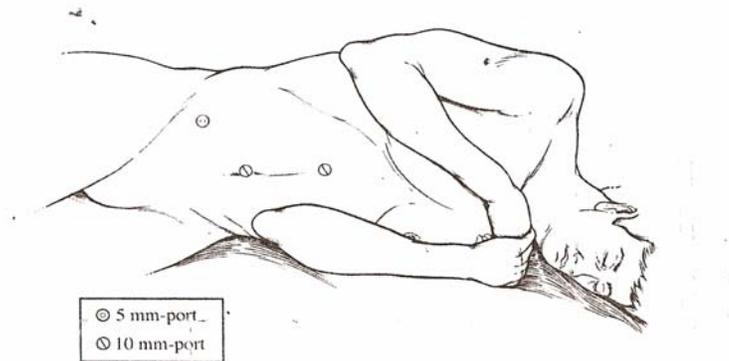
1. Transgressing the peritoneal cavity carries the potential risk of adhesion.
2. Anastomotic leak can cause intraperitoneal spillage, paralytic ileus and septicemia.

Peters and associates reported the first laparoscopic dismembered pyeloplasty in a child, using a transperitoneal approach with four ports and interrupted sutures tied intracorporeally.⁽⁶⁹⁾

Technique

The principle of the dismembered pyeloplasty is followed in general. The preoperative enema to empty the colon, nasogastric tube to decompress the stomach and Foleys catheter to keep the bladder empty are helpful to improve the available space in the peritoneal cavity in children.

Following induction with general anaesthesia, cystoscopy and retrograde pyelography are performed to confirm the diagnosis and a long indwelling ureteral stent is passed



The patient is placed in a 45-degree lateral decubitus position and secured to the operating table. Insufflation is performed through a veress needle and three laparoscopic ports are passed into the peritoneal cavity.

The ipsilateral colon is reflected and the proximal ureter and renal pelvis are identified and fully mobilised. If a crossing vessel is present, fibrotic bands

between the vessels and collecting system should be divided. The renal pelvis is transected circumferentially above the PUJ and the proximally spatulated through the level of the PUJ laterally. Care should be taken not to cut the ureteral stent.

If the crossing vessels are present, the ureter and renal pelvis are transposed to the opposite side of the vessels prior to completion of the anastomosis. All intracorporeal suturing is performed using the endostich device and 4'0' poyglycolic acid suture. A corner stitch is placed through the most dependent portion of the renal pelvis and through the corresponding corner of the spatulated ureter. The posterior anastomosis is then performed using multiple interrupted sutures followed by the anterior anastomosis.

As in true open pyeloplasty, the goal of surgical repair is to create a dependent, tension free, water tight anastomosis.

Following completion of anastomosis, a 5mm closed suction drain is placed through a posterior stab incision into the perinephric space adjacent to the PUJ. Hemostasis is confirmed, the CO2 is evacuated and port sites are closed.

RETROPERITONEAL PYELOPLASTY

With the patient in the lateral position tip of 12th rib is identified and iliac crest is outlined. Between these two points an incision is made in the middle. It is

deepened by a blunt dissection with a hemostat and index finger to reach the peritoneum.

Then, peritoneum is pushed away from abdominal wall either with index finger or Hegar dilator (size 12 or 14). The inflating balloon is inserted through this and kept between the peritoneum and muscles. It is inflated with CO₂ or saline is put to create a space beneath the peritoneum. Balloon is kept for few minutes and is deflated.

A trocar either a Hassons type or a trocar with self-retained balloon at the tip, is chosen to prevent leak from the trocar site. After verifying the correct entry of retroperitoneal space by telescopic examination, two additional ports are made one near the costal margin and another near ileac crest. By blunt and sharp dissection pelviureteric junction is exposed and pyeloplasty is done in the usual way.(100)

In retroperitoneal approach, the main advantage is avoiding entry of peritoneal cavity and thereby urinary leakage related problems can be avoided.

The major disadvantage is very much limited retroperitoneal space in children and hence there is difficulty in suturing and knotting. In addition creating retroperitoneal space is difficult when compared to transperitoneal approach.

Complications of laparoscopic pyeloplasty.

The complications are similar to open pyeloplasty like anastomotic leak, septicemia in the immediate postoperative period and anastomotic stenosis later.

With experience and improvements in instrumentation and innovative suturing techniques laparoscopic approach will become the procedure of choice.

ROBOTIC PYELOPLASTY

Robotics is another exciting and evolving area for minimally invasive surgery in children. This new technique provides excellent three dimensional visualisation, unprecedented control of endocorporeal instruments, and an ergonomic surgeon's position. Robots may advance minimally invasive surgery by allowing paediatric urologist with limited laparoscopic experience, to rapidly master the endocorporeal skills necessary to treat pelviureteric junction obstruction.(70,71)

FETAL INTERVENTION

Currently it is accepted that antenatal therapeutic intervention is beneficial only in bilateral, moderate or severe hydronephrosis, hydronephrosis detected

around 24 to 28 weeks, fetal renal function is adequate, and there are no associated serious anomalies. The male child with posterior urethral valves with good renal function is the usual candidate. Mild uropathy which worsens on serial scans as shown by increasing dilatation or decrease of renal function is also an indication for antenatal decompression. The percutaneous vesicoamniotic shunting (in bladder outlet obstruction), open vesicostomy and open pyelostomy can be used for antenatal decompression.

PYELOPLASTY FOR SPECIFIC ANATOMIC DERANGEMENT

1. Pelviureteric junction obstruction in the Horseshoe Kidney

Pelviureteric junction obstruction is the most common congenital genitourinary abnormality found in association with a horseshoe kidney, occurring in approximately in 15% of cases.^(72,73) The horseshoe kidneys frequently demonstrate pyelocaliectasis without obstruction. This may be the result of a mild resistance to flow where the ureter crosses the isthmus. Once the diagnosis of functionally significant PUJ obstruction is made, the options for correction are the same as in normally rotated kidneys. Most dismembered pyeloplasties in otherwise normal kidneys are performed through an extraperitoneal flank or posterior approach.

Because of concern, regarding angulation and obstruction at the isthmus, division of the isthmus and nephropexy to allow more dependent drainage of the PUJ have been proposed.

Ureterocalicostomy may be considered as an alternative to dismembered pyeloplasty for repair of even primary PUJ obstruction in horseshoe kidneys. It permits dependent drainage and avoids any problems related to the isthmus.⁽⁷⁴⁾

Newer minimally invasive techniques have been successfully utilised for PUJ obstruction in horseshoe kidneys, albeit, in small numbers of patients. Antegrade endopyelotomy has been used successfully in seven of eight reported cases.

Laparoscopic pyeloplasty has been successfully applied to horseshoe kidneys (**J.H.Ross,H.Winfield 1997**). The horseshoe kidney is particularly amenable to laparoscopic approach; because the anteriorly placed PUJ is easily exposed and manipulated with this technique.

2. Pelviureteric junction obstruction in the lower segment of duplex collecting system.

Vesicoureteric reflux is often seen in the lower segment of a completely duplicated system and is some times associated with pelviureteric junction obstruction. The following procedures can be done

1. When the duplication is complete dismembered pyeloplasty may be performed.
2. When the lower pole ureter of a partially duplicated system is short, the entire lower pole ureter is excised and a lower to upper pole pyeloureterostomy is performed.
3. When there is marked hydronephrosis of the lower-pole moiety, particularly if the pelviureteric junction obstruction is difficult to expose, then an ureterocalicostomy may be performed.
4. If the lower pole is poorly functioning, a lower-pole heminephrectomy is appropriate.
5. If a dysplastic upper pole segment is present, owing to an ectopic ureter or ureterocele, it may be excised at the time of the lower pole repair. Alternatively, an upper to lower pole ureteropyelostomy may be performed at the time of the lower pole pyeloplasty.

3. Pelviureteric junction obstruction in the pelvic kidney

Renal ectopy refers to a kidney outside the renal fossa. It is a rare finding, occurring in 0.01 to 0.08 percent of patients.⁽⁷⁵⁾ According **Gleason** the sites

include pelvic (55%)(Fig No.14), crossed (32%), lumbar (12%), and thoracic (1%). Approximately half of such kidneys are hydronephrotic. This results from a variety of causes, including PUJ problems (37%), VUR and lower tract problems (26%) and hypoplastic adynamic segments at the UVJ (15%). All cases require lower tract imaging and accurate anatomical upper tract imaging. Surgical correction requires special considerations. Position, incision, and approach must be individualized; in a majority a modified Gibson incision allows an extraperitoneal approach. Open pyeloplasty is usually successful. Experience with less invasive techniques is limited.

ANTENATALLY DETECTED HYDRONEPHROSIS

The clinical presentation of the pelviureteric obstruction has dramatically changed since the advent of maternal ultrasonographic screening. Before the routine fetal ultrasound the commonest presentation was abdominal flank mass.⁽⁷⁶⁾ 50% of abdominal masses in newborns are of renal origin with 40% being secondary to pelviureteric obstruction. The newborns are also present with abdominal pain, urinary tract infections, irritability, vomiting, and failure to thrive.

Fetal urinary tract dilatation is present approximately 1 in 100-200 pregnancies. Male to female ratio is 3: 1 and usually sporadic. Left kidney is commonly involved.

Associated Anomalies

10% of the PUJ is associated with vesicoureteric reflux. The association of multicystic dysplastic kidney and contralateral pelviureteric junction obstruction is well known. The dysplastic kidney reflects the extreme end of the clinical spectrum of pelviureteric junction obstruction. Bilateral pelviureteric junction obstruction is 10-36% of cases.(77)

Differential Diagnosis

Dilatation of urinary tract can be secondary to obstructive or nonobstructive causes. The various obstructive causes include pelviureteric junction obstruction (44%), VUJ obstruction (21%), multicystic dysplastic kidney, ureterocele/ectopic ureter, duplicated collecting system (12%), posterior urethral valves (9%) and hydrometrocolpos.

The non obstructive causes include physiological dilatation, vesicoureteric reflux (14%), prune belly syndrome, renal cyst and megacalycosis.

Indications for maternal fetal ultrasonography

Whenever the discrepancy in the expected fundal height for gestational age should undergo prompt an examination to detect multiple fetus, oligohydromnios and polyhydromnios. A history of previous pregnancies associated with congenital anomalies is a specific indication for ultrasonographic screening during pregnancy.(Fig no.1)

A systematic approach to the prenatal diagnosis of urinary tract abnormalities improves the yield of the ultrasonographic examination. (**Schlüssel RN, MandellJ et al**) (78)

This includes the following steps

1. Assessment of fetal size and maturity.
2. Assessment of amniotic fluid volume (AFV), which is a semi quantitative technique. Currently, there are two methods to estimate AFV; amniotic fluid index (AFI) or measurement of the single deepest vertical pocket of amniotic fluid. Current ultrasonographic criteria for oligohydramnios (actual AFV < 500 ml) include an AFI of 5.0 cm or less and a two - diameter amniotic fluid pocket of less than 15 cm. Both methods are poor predictors of oligohygramnios.
3. Identification of the gender of the fetus.
4. Localisation and characterisation of urinary tract abnormalities (bladder volume, kidney size, anteroposterior diameter of the renal pelvis, echotexture of the kidney, and the presence of cortical cysts).
5. Identification of associated abnormalities.
6. Monitoring the detected lesions and their impact on the overall health of the fetus.

Dilatation has been considered significant when AP diameter of renal pelvis measured 10mm or more irrespective of gestational age. In addition a ratio

of renal pelvis to AP diameter of kidney > 0.5 has been considered significant.

Grigon et al (79) graded the fetal hydronephrosis into 5 grades as follows

Grade I – detectable renal pelvic dilatation.

Grade II – dilatation greater than 1 cm.

Grade III- IV – further degree of pyelectasis with dilatation greater than 1.5 cms.

Grade V – association with atrophic cortex.

Dilatation of the collecting system can occur in the absence of obstruction and is termed as physiological hydronephrosis.

Typically, the ureter is of normal caliber and is not seen.(80) But if it dilated the size of ureter is also assessed ultrasonographically and graded 1-3 according to ureteral size width $< 7\text{mm}$, $7\text{-}10\text{mm}$, $>10\text{mm}$ respectively.(81)

Harrison et al suggested that proportion of more than 1:2 of the pyelon width to the kidney width is pathological and may be diagnosed as fetal hydronephrosis.(82)

In order to standardize postnatal evaluation of prenatal hydronephrosis, a grading system of postnatal hydronephrosis was implemented in 1993 by society for fetal urology.

The Society for fetal urology grading system is as follows

Grade 0 – Normal kidney with no hydronephrosis.

Grade 1 – Slightly dilated renal pelvis without caliectasis.

Grade 2 – Moderately dilated pelvis with mild caliectasis.

Grade 3 – Large renal pelvis, dilated calyces, and normal renal parenchyma.

Grade 4 – Very large renal pelvis, large dilated calyces, with
thinning of the renal parenchyma.

P. A. Dewan et al describe a new sign in ultrasound examination that may help to identify those fetuses who have high intrarenal pressure and therefore justify more aggressive management, while obviating the need for intervention for those in whom it is not present. The egg-shell sign consists of a thin crescent of increased echogenicity over a distended calyx and, in this case, was documented to be associated with other features of raised intrarenal pressure.(101)

**AMNIOCENTESIS AND ASSESSMENT OF FETAL URINE
FUNCTION**

This is an invasive procedure to be done only by experienced personal in patients in whom renal ultasound has revealed several problems like bilateral hydronephrosis or silidary kidney, progression of severity, oligohydromnios or

absence of corticomedullary junction differentiation, which suggest bilateral hydronephrosis and dysplasia of the kidneys.

The goal is to identify fetuses that are risk while in utero, of total renal destruction and pulmonary hypoplasia when there is a reasonable hope of beneficial treatment. This test currently carries of two types.

1. Evaluation of amniotic fluid fetal urine to assess fetal tubular function.
2. Evaluation of proteins in fetal serum or urine to assess fetal glomerular filtration rate.

Between 16 and 21 weeks of gestation, the fetal urine normally becomes progressively more hypotonic because of selective tubular reabsorption of sodium and chloride in excess of free water. The most quoted values for fetal urinary electrolytes and osmolality abnormalities indicative of an impaired renal function in the fetus with detectable upper tract dilatation are as follows

1. Osmolality of less than 210mosm/l
2. Urinary sodium concentration of less than 100 meq/l
3. Urinary chloride concentration of less than 90meq/l
4. Urinary output of less than 2ml/hr

5. Urinary calcium levels greater than 8mg/dl (normal <8mg/dl) are the most sensitive indication of renal dysplasia(100%) but have a demonstrated specificity of only 60%.
6. Beta 2 microglobulin elevation in fetal urine of greater than 4mg/l is pathological.

Management

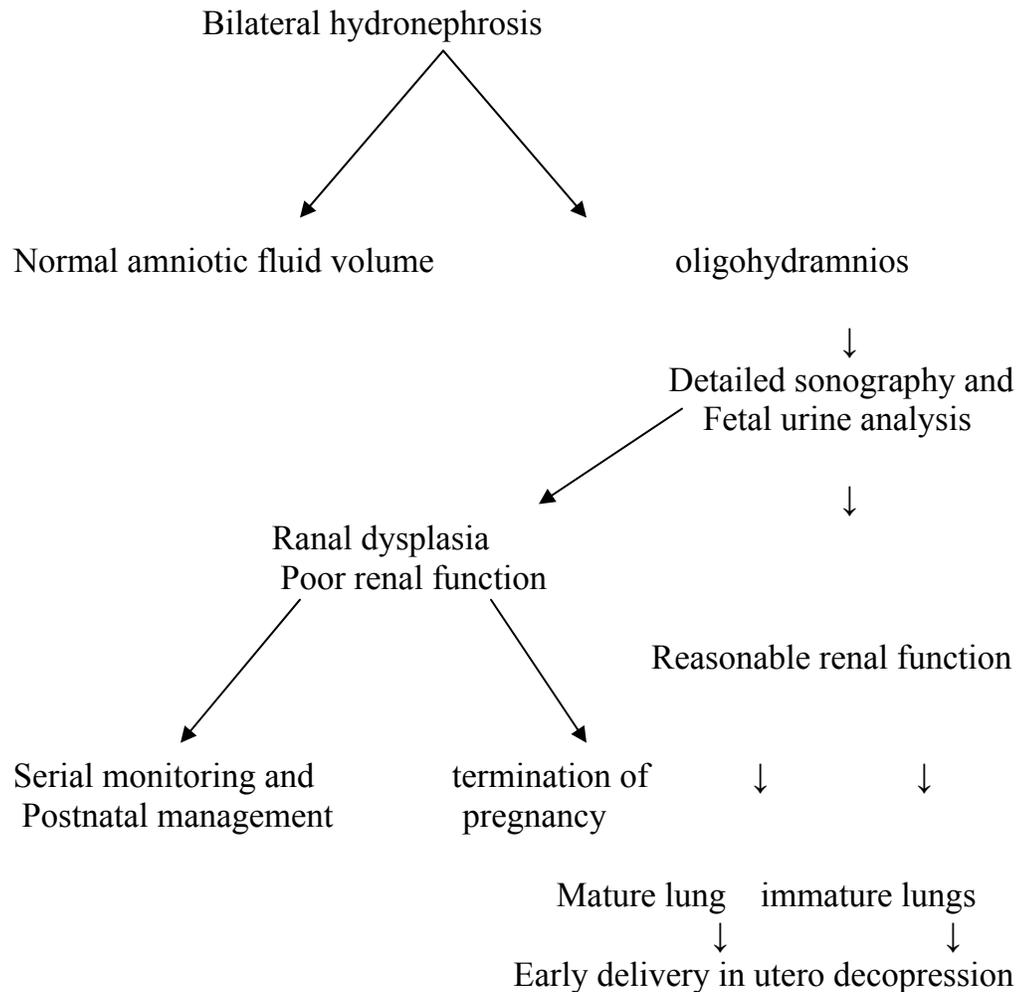
Antenatal Management

Fetuses with unilateral dilatation of upper tracts and a normal contralateral kidney are simply observed with serial ultrasound examinations at 1-2 weeks intervals till term.

Fetuses with bilateral hydronephrosis who are at higher risk for obstructive uropathy and in whom the Amniotic fluid index should be carefully monitored. Once the oligohydromnios develops, fetal urine sampling is the next step to be evaluated.

Once a fetus with salvageable function is selected, early delivery is considered if it is more than 32 weeks of gestation. If it is between 20-32 weeks various fetal interventions are considered. It is less than 20 weeks with oligohydramnios termination of pregnancy to be considered.(96)

Scheme of management for antenatal diagnosed bilateral hydronephrosis



Postnatal evaluation and Management

All the newborns with a prenatal diagnosis should undergo physical examination at birth to rule out associated anomalies. Prenatally detected hydronephrosis should be confirmed postnatally on day 2 or 3 of life(Fig no 2),because neonates may have transient oliguria and dilated obstructed collecting system may appear for 24-48 hours of life.(83) The ultrasound evaluates pelvic dilatation, which is graded according to the SFU grading system. Ureteric

dilatation is specifically looked for. The bladder is evaluated for size, wall thickness, presence of diverticulum or ureterocele and dilatation of the posterior urethra.

Next VCUG should be performed to rule out posterior urethral valve, a bladder diverticulum, or vesicoureteric reflux. Even if the ultrasound is normal a VCUG should be performed, because reflux may be the cause of fetal hydronephrosis.

If the ultrasound and VCUG are normal, then only a follow up ultrasound in 6-8 weeks is necessary.

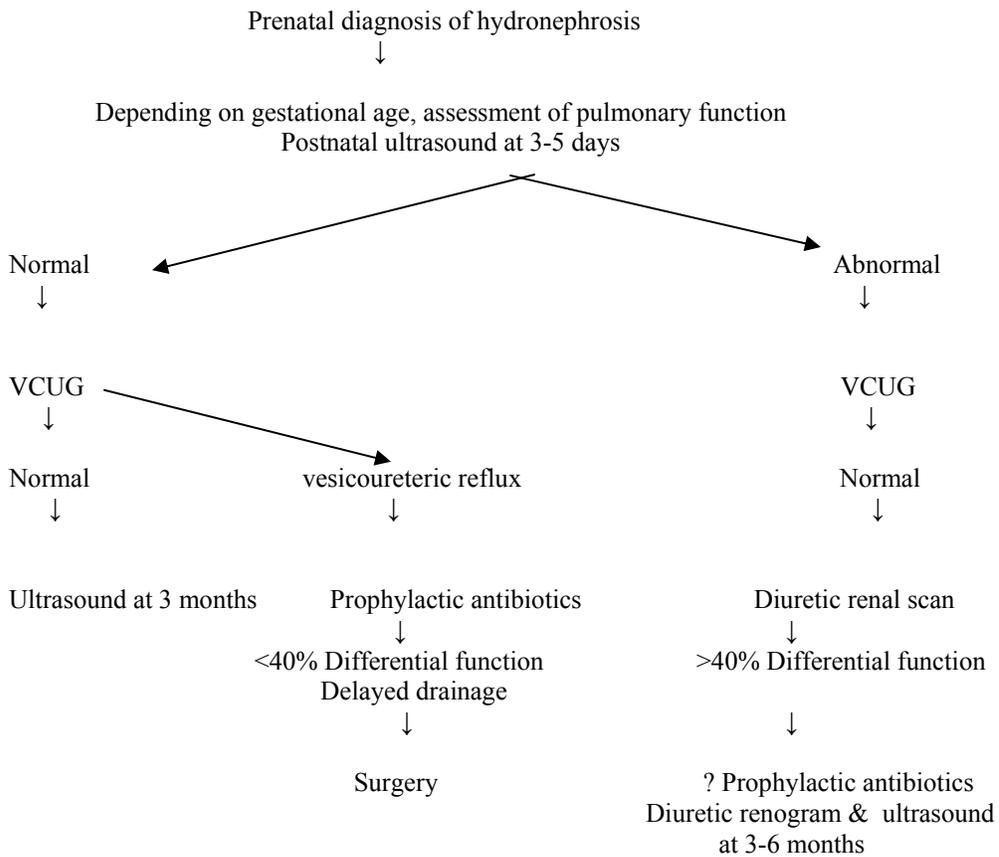
If the postnatal sonogram shows grade 1 or 2 hydronephrosis and the VCUG is normal, then the pelvicalyceal dilatation is due to physiological. These children should follow with ultrasonogram in 3-6 months period.

If the sonogram shows grade 3 or 4 hydronephrosis and there is no reflux, the upper tract must be evaluated further with T99 MAG 3 or DTPA diuretic renogram at 4-6 weeks of age.

The goal of early evaluation is to determine whether a true anatomic obstruction is present that should be repaired or whether it is safe to follow the infant nonoperatively.

Some authors advocate early surgical intervention to prevent damage to matured nephrons. While others feel that early surgery carries no specific benefit.

Algorithm for the the postnatal evaluation of infants with prenatally detected hydronephrosis



Koff and Campbell suggested that immediate postnatal surgical intervention is unnecessary in the majority of newborn child with pelviureteric junction obstruction. These babies should be followed up several examinations to observe anatomical and functional improvement. Surgery is undertaken in infant with deteriorating renal function.(84,85)

Follow up ultrasound may be performed 3-6 months after operation when maximum improvement can be seen. Radionuclide scan are useful to monitor the post pyeloplasty function and drainage.

Bilateral Hydronephrosis

If the ultrasound finding is suggestive of posterior urethral valve, an early voiding cystourethrography is performed to confirm the diagnosis and treat accordingly. All male babies with bilateral hydronephrosis are evaluated with voiding cystourethrography to rule out bladder outlet obstruction and reflux. In female babies direct radionuclide cystography is considered only if ureter is dilated on ultrasound.

In bilateral pelviureteric obstruction the symptomatic side or the side with better function should be operated first. If nephrectomy is considered on one side, the pyeloplasty should precede this.

Unilateral Hydronephrosis

The work up depends on the status of the opposite kidney on ultrasound. If the opposite kidney is normal, and the ureter is not seen on ultrasound, the most probable diagnosis is pelviureteric junction obstruction and the baby only requires a diuretic renogram at 4-6 weeks of age for further evaluation.

If the opposite kidney shows multicystic dysplastic kidney, voiding cystourethrography should be done to rule out vesicoureteric reflux. If it is present, aggressive management should be taken to save the single functioning kidney.

At least 35 to 50% of antenatally detected hydronephrosis reveals an apparent obstruction at the pelviureteric junction.

Two schools of management have been emerged. Early intervention is supported by the fact that the functional recovery is better with early relief of obstruction and there is a significant risk of irreversible functional loss while on observation alone. Where as, delayed intervention until the obstruction is proved on serial assessment is lead onto renal damage. Recent evidence suggested that early pyeloplasty improved the renal growth and function. Diuretic renography is gold standard to diagnose obstruction. This should be done after 4-6 weeks of age.

Ransley et al proposed treatment protocols based on the split renal function on the first diuretic renogram done after 4 weeks of age. Kidneys with SRF > 40% are observed and SRF <20% undergo operation or percutaneous drainage. Kidneys with SRF 20-39% undergo repeat scan at 3 months of age and those kidneys with SRF <40% then undergo pyeloplasty while the kidneys that improved good function are observed.(85)

COMPLICATIONS OF PYELOPLASTY

1. Persistent Urinary Leakage

If the persistent urinary leakage occurs beyond 10 days, evaluation with Ultrasonogram and antegrade or retrograde pyelography should be entertained. Conservative measures may preserve the repair but if hydronephrosis persist, reexploration should be considered.

2. Delayed Opening of Anastomosis

This is usually indicated by increased nephrostomy drainage following removal of a splint or in patients without splint, by persistent large volume of drainage. It is caused by postoperative edema at suture line or by blood clot. If the persistent high volume drainage continues, appropriate measures should be undertaken to evaluate the patency of the anastomosis.

3. Urinoma Formation

Collection of urine around kidney caused by anastomotic leakage or may follow the removal of a nephrostomy tube. In most cases it results from inadequate placement or early drain removal. The most frequent cause of an urinoma seems to result from premature removal of the flank drain in the absence of nephrostomy. The presence of urinoma should be suspected in a patient who suddenly ceases urinary leakage as opposed to those who have a gradual diminution and cessation of drainage. Should an urinoma be suspected, ultrasound

is helpful to ascertain diagnosis. Many small urinomas resolve spontaneously while other must be drained surgically.

4. Infection, Anastomotic Disruption and Stone Formation

In pyeloplasty if the urine becomes infected with E.coli, proteus the absorbable sutures may either become a nidus for stone formation or reabsorb too quickly, causing leakage or disruption of the suture line. The faulty suture technique or vascular compromises, the presence of infected urine are the other factors responsible for anastomotic disruption. Prevention and treatment of the infection may improve the outcome.

5. Hemorrhage

In most cases it is related to the presence of a nephrostomy. It can be controlled with hydration and transfusion.

6. Ureteral Kinking or Angulation

This complication occurs when the kidney is returned to the renal fossa, which can alter the relationship of the renal pelvis to the newly anastomosed ureter. To prevent this occurrence, **Hendren** suggest pexing the kidney to the psoas muscle.

7. Anastomotic Fibrosis and Stenosis

It may be as a result of vascular compromise, persistent urinary leakage, extravasation of infected urine, or any combination thereof. By keeping these events to a minimum, the incidence of anastomotic failure will be reduced.

8. Persistent Hydronephrosis and Secondary Nephrectomy

The term persistent hydronephrosis should probably be replaced by radiological deterioration. This may be occurring in a small group of patients.

Preservation of the ureteral vasculature, a watertight anastomosis, prevention of infection and care the placement and management of drains enhance the opportunity for a favorable outcome.

REDO PYELOPLASTY

The failure rate of pyeloplasty is 5 %.(86) The causes of failed pyeloplasty after allowing 3 months waiting period to allow for anastomotic edema to settle are

1. Dense scarring around the anastomosis in all cases.
2. A high non dependent anastomosis in some cases.

The anastomosis is however almost always proved patent. The points that need attention during pyeloplasty are (87)

1. Meticulous technique avoiding uerteral ischemia.
2. Avoid an undrained urinary extravasation

Both are chief causes of perianastomotic scarring .A well placed drain close to the pyeloplasty is essential. The use of nephrostomy also reduces the chance of urinoma and associated with extremely low redo rate.

Indications for redo pyeloplasty

1. Recurrence of palpable lump and pain
2. Un resolving urosepsis
3. Lack of drainage in nephrostogram if nephrostomy is in place.

While a successful pyeloplasty should result in diminishing renal dilatation in ultrasound and improving renal function in radionuclide scan, the following points need to be remembered.

1. The ultrasound scan show increased dilatation in the first few months after pyeloplasty, which then resolves gradually.(88)
2. The function on radionuclide study remains unchanged in many pyeloplasties and can even show some deterioration in 7% of successful pyeloplasties.(89)

MANAGEMENT OF FAILED PYELOPLASTY

Surgery for failed pyeloplasty should not be considered for at least 2 months. There are five basic approaches to the treatment of secondary PUJ obstruction are (90)

1. Redopyeloplasty
2. Ureterocalicostomy
3. Laparoscopy
4. Endopyelotomy
5. Nephrectomy

Open procedure

The principles of open redo pyeloplasty are excision of the scarred reanastomosis in tension free dependent position and routine use of stent and nephrostomy.

When tensions free anastomosis is not possible, the lower calyx denuded of overlying renal parenchyma, and anastomosed to the spatulated ureter as a ureterocalicostomy.

Endourological procedure

Endopyelotomy can give satisfactory results in failed pyeloplasty cases provided

1. The stenotic segment is not long
2. The kidney is not very hydronephrotic
3. In older children
4. There is no vessels lateral or posterolateral to the PUJ where the incision will be made.
5. The stenotic PUJ can be cannulated.

The results of endopyelotomy for failed pyeloplasty (100%) seem to be better than endopyelotomy done as a primary procedure (62%) for children.⁽⁹¹⁾

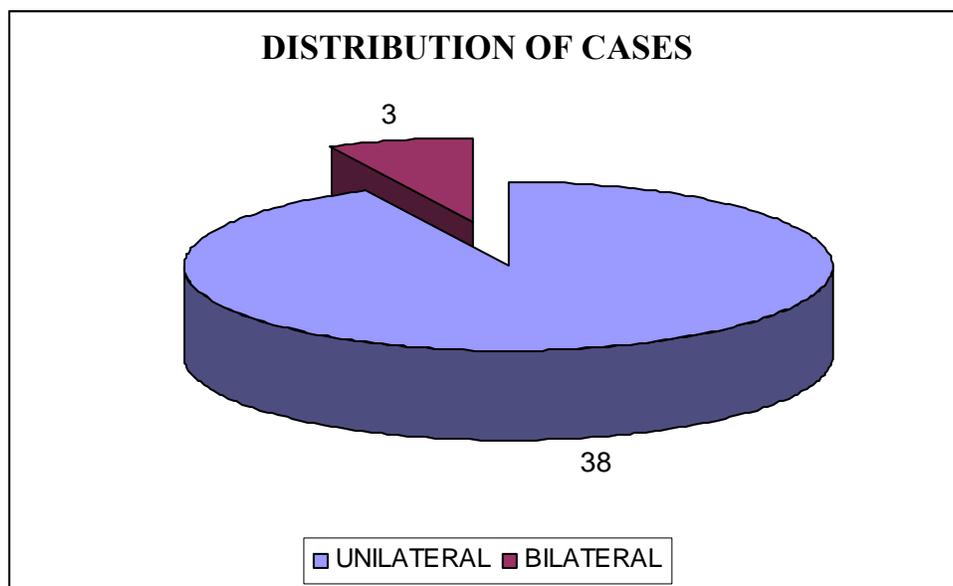
6. RESULTS OF STUDY AND DISCUSSION

In this study 41 cases of the pelviureteric junction obstruction were studied from the period of July 2003 to September 2005.

Distribution of Cases

Among the 41 cases unilateral 38 cases, bilateral 3 cases.

SIDE	NO OF CASES	PERCENTAGE
UNILATERAL	38	93%
BILATERAL	3	7%



Among the 38 cases of unilateral, 13 (32%) were on right side and 25 were on left (61%) and 3 cases (7%) were having bilateral obstruction. This is

consistent with observation of Johnston JH. J Urol 1977:117:97& Snyder.Urol Clin North Am 1980:7; 273.

SIDE	NO OF CASES	PERCENTAGE
RIGHT	13	32%
LEFT	25	61%
BILATERAL	3	7%

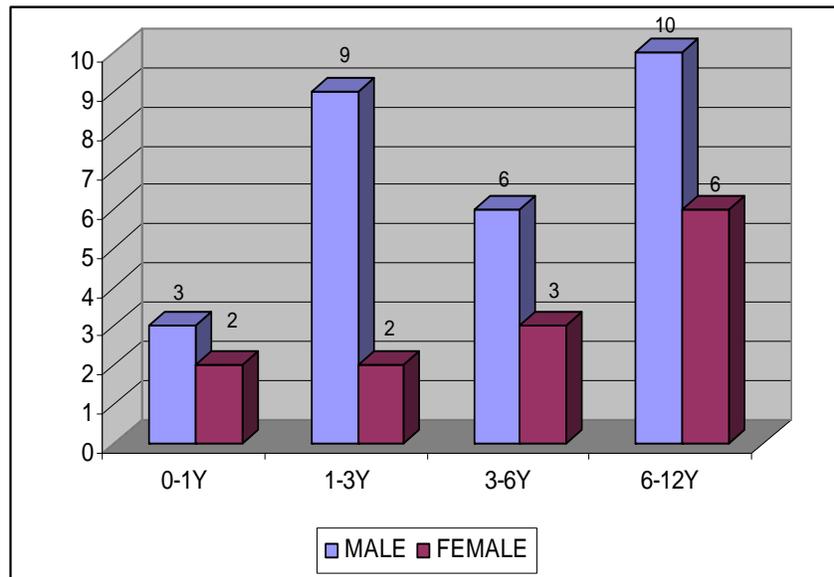
Age and Sex Incidence

Age

In this series commonest age group is 6-12years. 16 cases (39%) are in this age group. Only 3 cases presented with antenatal diagnosis.

AGE	MALE	FEMALE	TOTAL	PERCENTAGE
0-1Y	3	2	5	12%
1-3Y	9	2	11	27%
3-6Y	6	3	9	22%
6-12Y	10	6	16	39%

AGE INCIDENCE



Sex Ratio

In this series Boys were commonly affected than girls.. Male to female ratio is 2:1. Of 41 cases studied 28 cases are male (68%) and 13 cases are female (32%). Williams and Karlaftis, 1966, Kelalis et al 1971 also observed the same incidence.

Clinical Presentation

Before the advent of antenatal ultrasonography, most infants with pelviureteric junction obstruction present with abdominal masses ,Urinary tract infection, Hematuria, and gastrointestinal discomfort.

Most of the cases now are being detected during antenatal ultrasonographic screening. But in this study only 3 cases presented with antenatal diagnosis.

In this series the main presenting symptom was mass per abdomen. (Fig No.16&17). 10 presented with mass and pain abdomen. 11 cases presented with only urinary tract infections. Most of the patients had associated gastrointestinal symptoms like abdominal discomfort,nausea,vomiting. Hematuria occurred in only 2 cases.

Associated Anomalies

In this series 6 of the patients had other congenital anomalies namely

1. Malrotation
2. Hemivertebra
3. Situs inversus with dextrocardia
4. Unilateral renal agenesis (Fig No:8&9)
5. Glanular hypospadias
6. Right undescended testis

Pelviureteric junction obstruction in Other Anomalies of Kidney

In this series pelviureteric junction obstruction occurred in an ectopic kidney (left pelvic kidney) (Fig No.12&14) and another occurred in crossed ectopia.(Fig No.16)

Etiology

Among the 41 cases studied, 37 had intrinsic cause for the obstruction. The length of the narrowed segment was ranging from 0.5cm to 2cm in length. There was one case with polar vessel. In this series pelviureteric junction obstruction was present in a dysplastic kidney for which nephrectomy was done. All the excised specimen were sent for histopathological examination.

TREATMENT

Conservative Management

In this series only 3 patients were treated conservatively. Of 3, 2 were diagnosed antenatally.

Surgical Treatment

In 37 patients 39 Anderson Hynes pyeloplasty was done including 3 bilateral cases. Of these
Nephrostomy tube + Stent + Drainage tube in 4 cases,
Stent with drainage tube in 19 cases,
Nephrostomy tube with drainage tube in 1 case,
Drainage tube alone kept in 13 cases.
Nephrectomy was done in 1 case, in which the kidney was dysplastic.

In this series the transanastomotic stent was removed after 48 hours, followed by the removable of nephrostomy tube on the 5th postoperative day. The extraperitoneal drain was removed on 8 th PO day.

Post Operative Complications

Mild urinary leak occurred through drainage site in most of the cases, which subsided slowly over a period of 7 to 10 days. One patient developed perinephric urinoma which was treated by percutaneous drainage. Another had intestinal obstruction in the postoperative period. On laparotomy a loop of small bowel was found to herniate through the ureteric mesentery and gangrenous. He also had malrotation. Resection of the gangrenous bowel loop along with Ladd's procedure was done.

Outcome

Of 39 Anderson Hynes pyeloplasties done, the success rate was 95 % in this series.

Follow up Study

All the patients were followed over a period of one month to two years. During the follow up most of cases were examined clinically and radiologically with ultrasound and few cases with intravenous urogram and isotope renogram .In the ultrasound examination the pelvicaliceal dilatation and ureter caliber were assessed(Fig No.). The size of the kidney was found to be without much alteration. In postoperative intravenous pyelogram, dye was seen in early pictures which were not present in preoperative intravenous pyelogram(Fig No. 21)

CONCLUSION

The incidence and presentation in this study are consistent with the most of the reports given in the literatures. Eventhough the other series show the incidence of antenatally diagnosed hydronephrosis is more, in this series ironically only 3 cases had antenatally diagnosed hydronephrosis. The results of the dismembered pyeloplasty (Anderson Hynes pyeloplasty) are comparable with any other reports in the literature. In conclusion the Anderson Hynes pyeloplasty is the most preferred method to treat pelviureteric junction obstruction.

PROFORMA FOR PELVIURETERIC JUNCTION OBSTRUCTION IN CHILDREN

Name of the patient:

IP No :

Age :

sex:

DOA:

DOD :

Diagnosis :

PRESENT HISTORY

1. abdominal mass
2. abdominal pain
3. Hematuria
4. urinary tract infection – dysuria
pyuria
5. gastrointestinal symptoms – nausea
Vomiting
6. Antenatally diagnosed
7. Hypertension

Side Right Left Bilateral

PAST HISTORY

Previous surgery
H/o Trauma

FAMILY HISTORY

ANTENATAL HISTORY

POSTNATAL HISTORY

INVESTIGATIONS:

Urine- culture and sensitivity

Blood – urea

- creatinine

1. Ultrasonogram
2. Intravenous pyelogram
3. Micturiting cystourethrogram
4. Diuretic renogram
5. CT scan

TREATMENT

1. Conservative management
2. Pyeloplasty
 - a) Anderson Hynes pyeloplasty
 - with nephrostomy tube and drain
 - with nephrostomy and stent and drain
 - with stent alone and drain
 - with extraperitoneal drain alone
 - b)Foley Y plasty
 - c)Spiral flap procedure
 - d) Vertical Flap procedure
 - e)Ureterocalycostomy
3. Pyeloplasty for UPJ obstruction in
 - 1.Horseshoe kidney
 - 2.Lower segment of duplex system
 3. Pelvic kidney
4. Laparoscopic dismembered pyeloplasty
5. Nephrectomy

Postoperative complications

Persistent urinary leakage

Delayed opening of anastomosis

Formation of urinoma

Infection

Anastomotic disruption

Stone formation

Hemorrhage

Ureteral kinking or angulation

Anastomotic fibrosis

Persistent hydronephrosis

Secondary nephrectomy

Follow up

24	Meena	6 y	F	339432	UPJ				+		R-AHP			+					
25	Buvaneswari	3 y	F	342682	UPJ			+			AHP		+				+		
26	Ajithpandi	5 y	M	350619	UPJ			+			AHP		+					+	
27	Sivasankar	4 y	M	351736	UPJ in single kidney			+			AHP		+				+		+
28	Balamurugan	1 ½ y	M	353547	UPJ				+		AHP		+						+
29	Sivanesan	1 ½Y	M	360657	UPJ				+		AHP		+				+		+
30	Naveenkumar	11	M	360664	UPJ			+			AHP		+					+	+
31	Praveen	6 y	M	364475	UPJ	+		+			AHP			+					
32	Nagarajan	7 y	M	365605	UPJ					+	R-AHP								
33	Aswin	5 y	M	369265	UPJ				+		AHP			+					+
34	Muthukumar	1 y	M	369472	UPJ			+			AHP		+				+		
35	Veerapathiran	11 y	M	373142	UPJ				+		AHP			+					+
36	Muthukumaran	11 y	M	381779	UPJ			+			AHP			+					+
37	Abdulsamed	3 Y	M	384317	UPJ				+		AHP			+					+
38	Chandru	4 y	M	384321	UPJ				+		AHP		+	+			+		+
39	Periyakaruppan	1 1/4y	M	267235	UPJ	-		+			AHP		+						+
40	B/O guruvammal	6/365	F	380448	UPJ	+		+											+
41	Ismail	5y	M	386471	L-pelvic kidney +UPJ				+										+

L- left side

BIL- Bilateral

SUR- Surgery, AHP-Anderson Hynes Pyeloplasty

S+D - Stent + Drainage Tube

NEPH- Nephrectomy

M+P – Mass + Pain

GIT- Gastrointestinal symptoms

R- right side

CON- Conservative management

N+S+D- Nephrostomy tube + Stent + Drainage tube

D – Drainage tube alone

M- Mass

UTI – Urinary tract infection

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ANTENATALLY DETECTED RIGHT HYDRONEPHROSIS



FIG NO- 1 - Antenatal USG shows increased pelvic AP diameter of right kidney

FIG NO- 2- Postnatal USG at 48 hr shows AP diameter of the pelvis is 1.7cm in right kidney

FIG NO-3 - The contralateral kidney of the same patient showed normal kidney

ULTRASONOGRAM OF PELVIURETERIC JUNCTION OBSTRUCTION

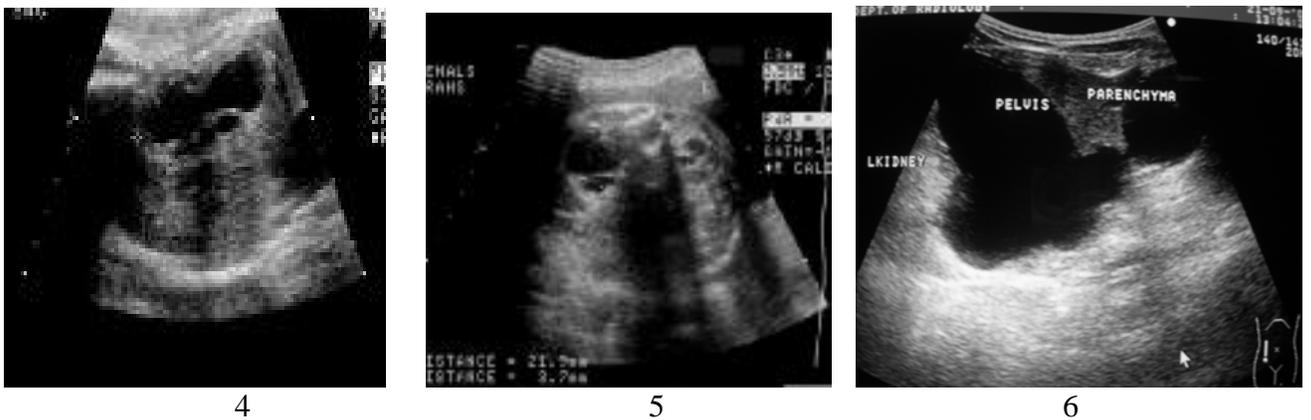


Fig No-4 & 5- USG of right kidney shows that increased pelvicalyceal system due to pelviureteric junction obstruction

Fig No-6- USG shows that dilated pelvicalyceal system with intervening parenchyma suggestive of crossed fused kidney with pelviureteric junction obstruction



7



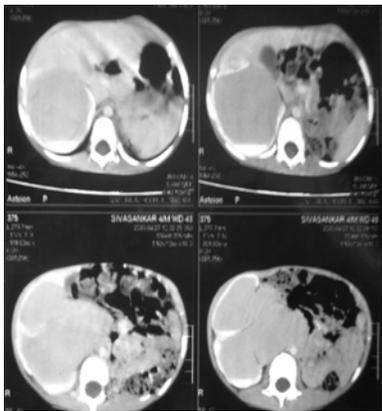
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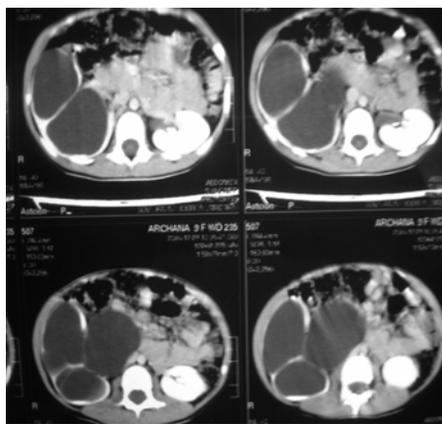
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FIG NO -7- LOIN MASS DUE TO LEFT CROSSED FUSED LEFT KIDNEY WITH HYDRONEPHROSIS

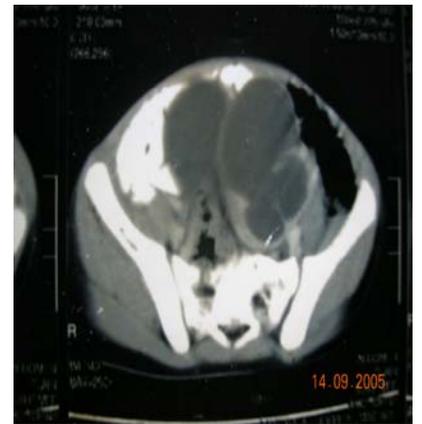
FIG -NO- 8&9 - INTRAVENOUS PYELOGRAPHY OF SOLITARY RIGHT KIDNEY WITH HYDRONEPHROSIS SHOWS DILATED PELVICALICEAL SYSTEM AND URETER NOT SEEN ON RIGHT SIDE.ON LEFT SIDE THERE IS NO CONTRAST SEEN



9



11



12

FIG NO – 10- computed tomography with contrast shows solitary right kidney with hydronephrosis with thin renal parenchyma and absent left kidney

FIG NO – 11- Abdominal CT with contrast of a patient with PUJ obstruction demonstrates massive dilation of the right renal pelvis and calyces, with thinning of the right renal cortex. The left kidney is normal in size and appearance.

FIG NO – 12-CT Scan of the left pelvic kidney with hydronephrosis

INTRAVENOUS PYELOGRAPHY WITH CONTRAST



13

FIG NO- 13 - INTRAVENOUS PYELOGRAPHY OF ANTENATALLY DIAGNOSED CHILD SHOWS FAINT NEPHROGRAPHIC EFFECT ON RIGHT SIDE DUE TO RITHT PUJ OBSTRUCTION



14

FIG NO- 14 - INTRAVENOUS PYELOGRAPHY SHOWS LEFT HYDRONEPHROSIS WITH RIM SIGN.



15

FIG NO-15 - INTRAVENOUS PYELOGRAPHY SHOWS RIGHT SIDE GRADE III HYDRONEPHROSIS WITH NO URETER SEEN. - RIGHT PELVIURETERIC OBSTRUCTION, LEFT KIDNEY NORMAL



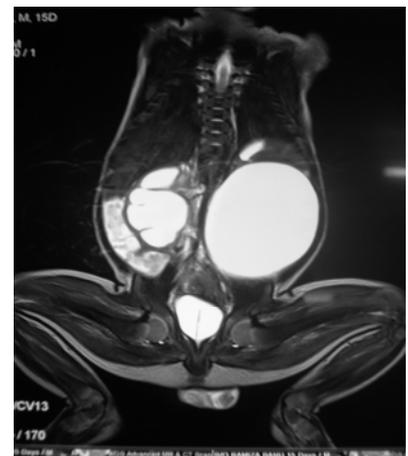
16

FIG NO - 16 - INTRAVENOUS PYELOGRAPHY SHOWS LEFT PELVIC KIDNEY WITH HYDRONEPHROSIS



17

FIG NO -17&18 - MR UROGRPHY SHOWS BILATERAL PELVICALICEAL SYSTEM DILATATION DUE TO BILATERAL PELVIURETERIC OBSTRUCTION



18

Fig No-19- Pre operative diuretic renogram

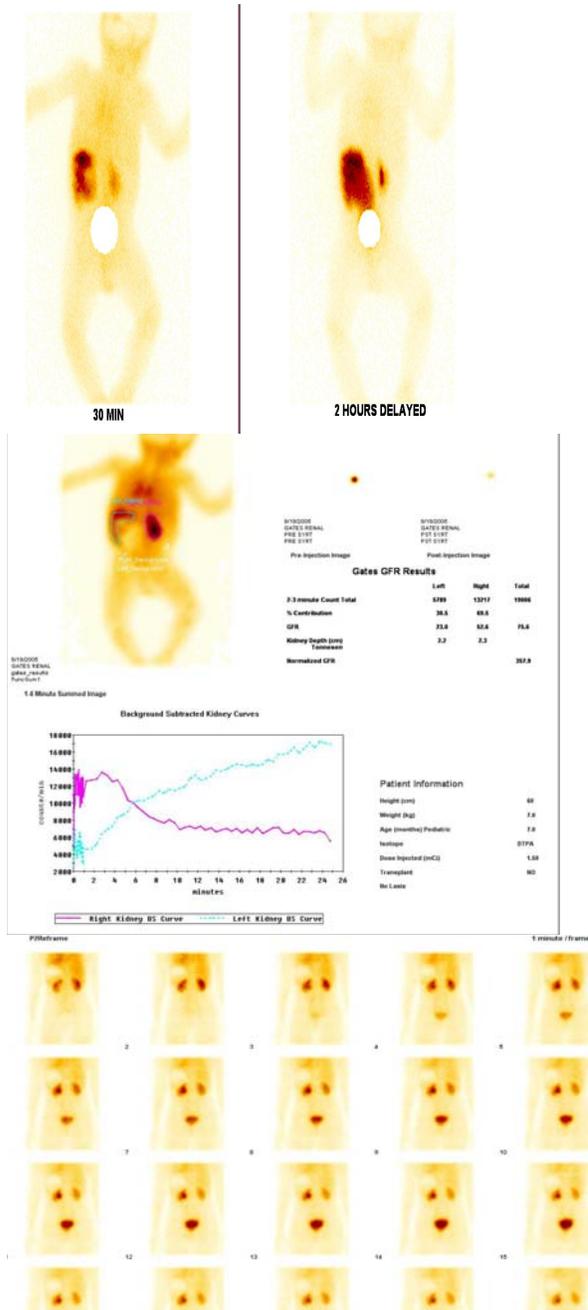


Fig No-20- Post operative diuretic renogram



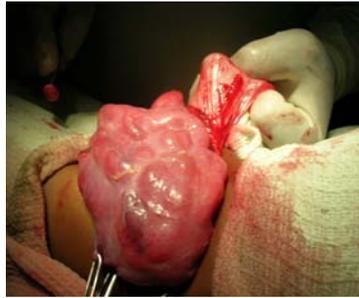
Preoperative Diuretic renal scan shows normal uptake and excretion of radiotracer from the right kidney into the right ureter and bladder. The progressive uptake of contrast material into the left renal collecting system without excretion is consistent with pelviureteric junction obstruction

Postoperative follow up renogram demonstrates renal tracer accumulation and excretion of left kidney. The normal right kidney demonstrates prompt uptake and excretion.

FIG NO-21- OPERATIVE STEPS OF ANDERSON HYNES PYELOPLASTY



1



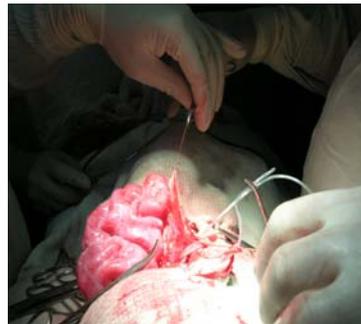
2



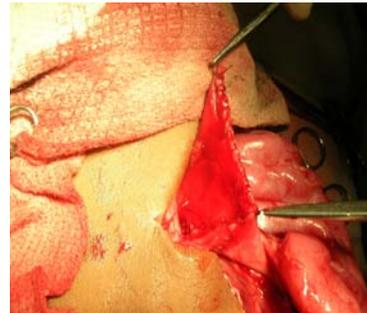
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5



6



7

Fig No-1and 2 - Through anterolateral incision left kidney approached extraperitoneally
Fig No- 3-Dilated Pelvis and ureter were identified and narrowed PUJ was dismembered.
Fig No-4, 5 &6- Ureter was spatulated and dependent site anastomosis completed.
Fig no-7- wound closed in layers with drainage tube.

FIG NO-22-POST OPERATIVE FOLLOW UP INTRAVENOUS PYELOGRAM



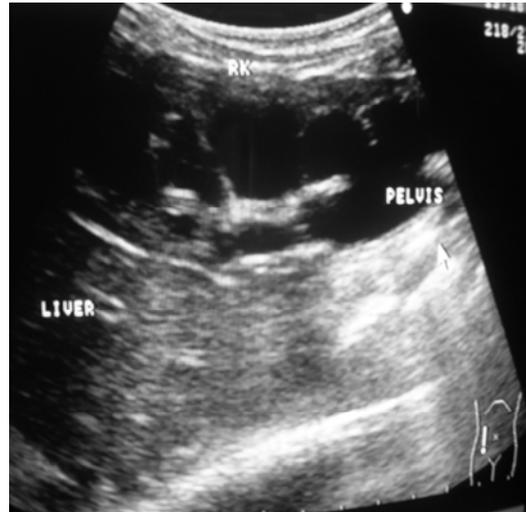
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4

Intravenous pyelogram shows

1. Preoperative film shows left sided hydronephrosis with no ureter seen
2. Postoperative 1 hour picture shows dye in the left pelvicaliceal system.
3. Post op 3 hour picture shows the dye is completely drained from the left pelvis
4. post pyeloplasty USG shows dilated pelvicaliceal system right kidney