INTRODUCTION

Definition:

Uretro-pelvic junction obstruction (UPJ obstruction) is restriction to urine flow from renal pelvis to ureter which, if left uncorrected, leads to progressive renal deterioration[1].

Incidence:

Obstruction of the uretro-pelvic junction (UPJ) is the most common congenital abnormality of the ureter with reported incidence of 5/100.000 annually[1].

UPJ obstruction responsible for about 48% of significant dilatation of the fetal kidney. UPJ obstruction more common in boys than girls, especially in the newborn period, when the ratio exceeds 2:1[1].

Left sided UPJ obstruction predominant, particularly in the neonate (67%) and bilateral in10-40% whether synchronous or asynchronous[1].

AIM OF THE WORK

Is to evaluate the value of adding fibrin glue as a sealant material to the anastomotic line during the procedure of laparoscopic non-stented pyeloplasty.

Chapter One

ETIOLOGY

Embryological background:

During nephrogenesis, the ureteral bud forms the ureter and the ureter then ascends towards the area of parenchyma that will become the kidney. The UPJ is formed at the fifth week. It's thought that the induction of the metanephric blastema is mediated by the ureteral bud through transforming growth factor β (TGFβ) and by growth factors such as c-ret, kdn-1, and wt1, as well as by transcription factors such as Pax2. The initial tubular lumen of the ureteric bud becomes canalized by the weeks 10-12 of gestation. Beginning from the midsection canalization proceeds to the UPJ and the vesico-ureteric junction (VUJ) with the UPJ being the last to canalise. Inadequate canalization of this area is thought to be the main embryological explanation for UPJ obstruction [(1)].

Intrinsic factors:

In all likelihood, intrinsic UPJ obstruction may be caused by a variety of factors, which may have both biochemical and mechanical etiologies.

1. presence of an aperistaltic segment

The most favorable theory is that the obstruction is secondary to muscular discontinuity, which disrupts the

coordinated motion of smooth muscle cells and may cause impaired peristalsis propagation across the UPJ and interference with the formation of urine bolus in the proximal ureter. This absence or disorientation at the UPJ is clearly evident on electron microscope examination.

Another theory suggests premature arrest of development of ureteral wall musculature leading to the persistence of an aperistaltic segment at the level the UPJ, thus preventing normal propulsion of urine down the ureter[2].

2. Persistent Valvular mucosal folds (Ostling's folds)or polyp:

Occasionally, the proximal ureter is folded on itself and persistence of the infolding may contribute to the kinked appearance of the upper ureter[3].

3. High insertion ureter:

Abnormal insertion of the ureter into the pelvis instead of a dependent position may cause a similar type of obstruction. During surgery, intrinsic obstruction is found to be a ureteral narrowing with angulation and the lumen is, in general, narrowed but open, allowing the probing of the lumen of the ureter[2].

4. Neurogenic theory.

It suggests that abnormal neuronal signaling with diminished synaptic vesicles may play a role in the

development of UPJ obstruction. Factors, such as protein gene product (PGP) 9.5 (a general neuronal marker), S100 protein (a nerve supporting cell marker), synaptophysin (a synapse vesical marker), and nerve growth factor receptor have been evident in decreased amounts in resected specimens of UPJ[2].

Extrinsic

1. Crossing vessel:

Extrinsic obstructions secondary to fibrous bands, kinks, and aberrant crossing vessels are also encountered. The incidence of that the proximal ureter is angulated, distorted, and compressed by vessels that supply the lower pole of the kidney in about 25% of UPJ obstructions.

During ascent and rotation of the fetal kidney, the kidney has segmented vessel from the aorta arranged in a ladder pattern. As the kidney ascends further, its blood supply is derived from higher vessels, while the lower ones disappear[5].

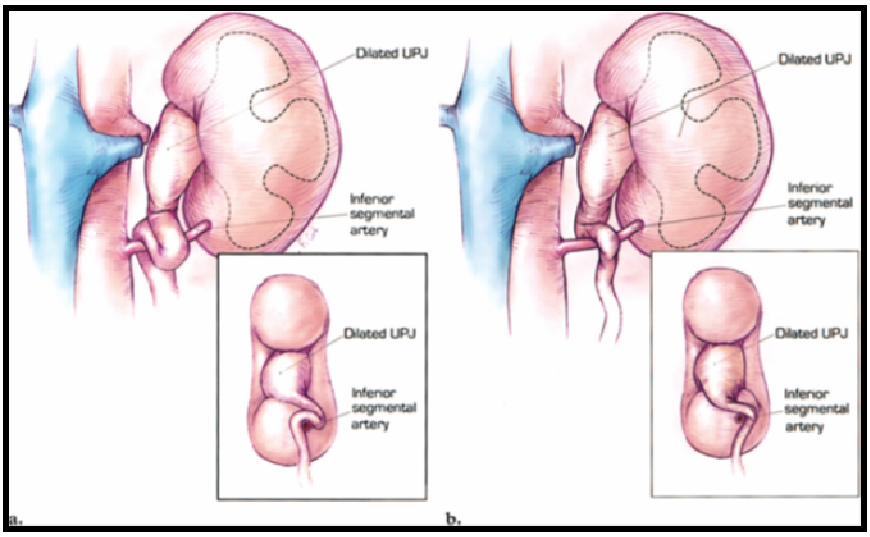

Figure (1): Drawings show left UPJO with hydronephrosis in which the dilated extrarenal pelvis and proximal ureter bow anteriorly (a) or posteriorly (b) over the lower-pole segmental artery. Note that, when there is significant hydronephrosis, the UPJO site is frequently more cephalad behind the overgrown pelvis[4].

2. Secondary to iatrogenic trauma of the UPJ:

Secondary causes can be due to upper tract infection, stones, trauma (eg, instrumentation), previous surgery which can lead to reactive fibrosis and formation of stricture[5].

3. *Secondary to VUR*:

In10% of patients with severe VUR due to elongation and tortuosity of the ureter at the level of UPJ[2].

4. Abnormal rotation of the kidney:

May be associated with ectopic kidney or hypermobile kidney, which can cause intermittent obstruction that is solely dependent on the position of the kidney relative to the ureter[6].

Unusual cause UPJ obstruction

Congenital lumbar hernia is rare in the pediatric age group. A case report describes an infant with a superior lumbar hernia with herniation of the kidney that resulted in UPJ obstruction. After surgical repair of the hernia and reduction of the herniated kidney, resolution of the UPJ obstruction achieved[7].

Chapter Two

PRESENTATION

Usually the presentation in most infants with UPJ obstruction is asymptomatic while in older age groups it's discovered because of their symptoms and signs.

The widespread of use of antenatal ultrasonography has played an important role in the discovery of the UPJ obstruction[3].

Infants

They may be present with hydronephrosis. UTI is the presenting sign in 30% of affected children beyond neonatal period.

Occasionally, infants may present with failure to thrive, feeding difficulties and sepsis secondary to urinary tract infection (UTI), pain or hematuria related to nephrolithiasis[3].

Older children

Older children may present with urinary tract infection (UTI), a flank mass or intermittent flank pain secondary to a primary UPJ obstruction. Hematuria may also be a presenting sign if it is associated with infection[3].

Young adult

In contrast to prenatal screening, adult UPJO may be discovered in several ways: (a) evaluation following symptoms such as back and flank pain, UTI, and/or pyelonephritis, hematuria, and (b) accidentally during diagnostic imaging of the abdomen or spinal area for other problems such as abdominal or back pain. A detailed history may reveal that the pain correlates with periods of increased fluid intake or ingestion of a food with diuretic properties (Dietl's crisis)[8].

With other associated urologic abnormalities UPJ obstruction is often associated with some congenital anomalies, such as imperforate anus, congenital heart disease, contralateral multicystic kidney, **VATER syndrome** which are (vertebral defect, imperforate anus, trachea-esophageal fistulae and radial and renal dysplasia), UPJ was noted in 21% of children with VATER in older reports, whereas more recently only 9% of VATER are diagnosed with UPJ, and esophageal atresia. Ultrasound is a must in such patients with the established diagnosis[9].

UPJ obstruction may be also presented with other congenital abnormality in almost 50% of patients, and 10% of patients with UPJ obstruction show ipsilateral reflux. SO, Voiding cystourethrography (VCUG) is mandatory as in such patients, more severe anomalies should be treated first[3].

Chapter Three

TECHNIQUES TO DIAGNOSE UPJ OBSTRUCTION

The challenge in the management of dilated upper urinary tract is to decide which child can be observed, which one should be managed medically and which one requires surgical intervention. There is no single definitive test to distinguish obstructive from non-obstructive cases[10].

1- Antenatal U/S:

The routine use of prenatal ultrasonography has made ultrasonographic findings the primary presentation of congenital ureteropelvic junction (UPJ) obstruction. Initial prenatal assessment typically occurs at 16-20 weeks' gestation. On the basis of those findings, serial follow up and postnatal studies are performed[3].

Criteria for fetal hydronephrosis

The criteria used to grade fetal hydronephrosis are the Society of Fetal Urology (SFU) consensus guidelines, which are based on pelvic dilation and caliectasis. They include the following:

Table (1): Society of Fetal Urology grading system for hydronephrosis[11].

Grade	Central renal complex	Parenchyma
0	Intact	Normal
1	Slight splitting of pelvis	Normal
2	Evident splitting of intra-renal pelvis or dilated extra-renal pelvis major calyces dilated	Normal
3	Wide splitting of pelvis major and minor calyces dilated	Normal
4	Wide splitting of pelvis major and minor calyceal dilatation	Thinned or reduced

Grade 3-4 hydronephrosis is 88% sensitive and 95% specific for obstruction on isotope scanning. However, because of the variability in assessing the degree of dilatation in ultrasonography as it depends on the operator skills, many centers continue to rely on the AP diameter of the renal pelvis to diagnose hydronephrosis.

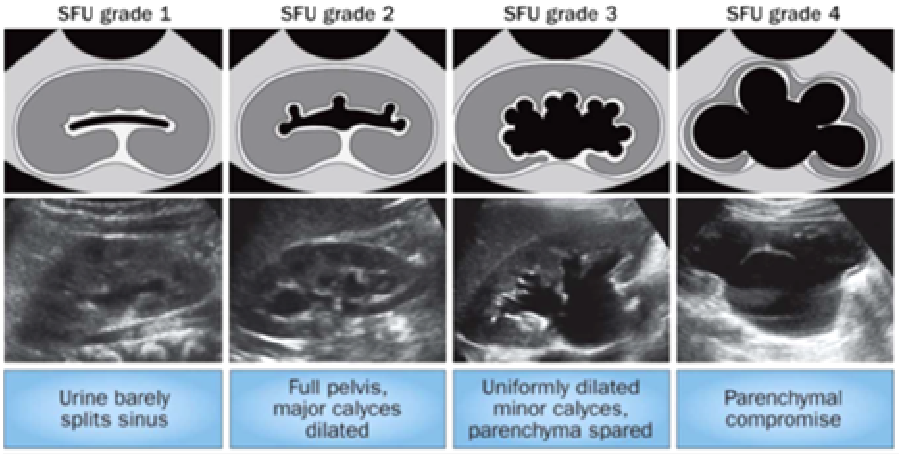

Figure (2): Ultrasound pictures of the various grades of hydronephrosis by SFU [11].

Hydronephrotic kidneys are now defined by the current radiologic standards as those with an antero-posterior (AP) diameter of the renal pelvis of greater than 4mm at a gestational age of less than 33 weeks and an AP diameter of greater than 7mm at a gestational age of 33 weeks or older. An abnormal initial ultrasonography should be followed up with another study after 4 weeks in severe cases or after 33-34 weeks in mild to moderate cases[11].

2- Postnatal U/S:

For any pre-natal u/s with renal pelvis antero-posterior diameter 5 mm or more.

Timing of postnatal U/S

Optimum timing for follow up postnatal imaging has much controversy. Some prefers delaying imaging at least 48 hours after birth to minimize false negative findings, owing to the infant's relative state of dehydration and decreased glomerular filtration rate (GFR). Others have found no great difference between early and delayed ultrasonography. It depends mainly on the severity of antenatal hydronephrosis or affection of solitary kidney as urgent evaluation will help in the decision for early intervention[3].

3- Intravenous pyelography (IVP):

Intravenous pyelography (IVP) has been the standard investigation to confirm UPJ obstruction in older children and adult patients. It has both functional and anatomic values. IVP findings suggestive of ureteropelvic junction obstruction include significant dilatation of the renal calyces and pelvis, funneling down to a narrow beak end, with non-visualization of the ureter[3].

4- Radionuclide renography:

Diuretic renography is usually performed to assess total and relative kidney function[2].

It measures the drainage time from the renal pelvis and assesses total and relative kidney function.

The rate at which tracer leaves the renal pelvis after diuretic injection, reflected in the slope of the drainage curve and often reported as T1/2 (the time required for 50% of the isotope to exit), is generally viewed as a reflection of the patency of the UPJ. Rapid drainage indicates no obstruction, while impaired drainage or slow or no washout (T1/2 > 20 min) indicates obstruction[2].

Some standardization is needed to increase detection of asymptomatic patients and this includes:

- **Radionuclide material**

99mTc-MAG3 (mercaptoacetyltriglycine) is the subtance of choice. It is cleared by the kidney through secretion in proximal convoluted tubules with extraction coefficient of 50%, giving excellent images, including ureteric visualization and produce clear accurate values, especially in low function or immature kidneys.

Other radionuclide materials can be used like 99mTc-DTPA (diethylenetriaminepentaacetic acid) with extraction coefficient of only 20%. It has the advantage of being cheap material. 123I-hippuran has short half-life and expensive[(2)].

The most useful measure in diuretic renography is the estimate of differential renal function. This is considered significant when it is less than 40%. It's usually correlated with the half-life (T1/2) washout curve.

Variables (as mentioned in the table below) include the use of intravenous fluids, the dosage and timing of administration of diuretic, the degree of pelvic dilatation, the severity of outflow obstruction, and the method of calculating the clearance after the administration of diuretic. For this reason, the T1/2 of the diuretic renogram cannot be the only indicator to determine surgery, especially in the neonate[(2)].

- **Diuresis renography pitfalls**

1. Severity of obstruction
2. Variable impairments of renal function
3. Volume capacity of the pelvis, ureter or bladder
4. Hydration state
5. Bladder fullness
6. Patient positioning
7. Radiopharmaceutical
8. Region of interest
9. Patient movement
10. Dose and choice of diuretic
11. Time at which diuretic is given

12 Method of data interpretation

(Presented by M. Majd at the American Academy of Pediatrics Meeting, Chicago, IL, 1989)

5- Voiding cystourethrogram (VCUG)

In neonates with known upper tract dilatation, it's important to exclude presence of other factors. This includes vesicoureteral reflux in up to 25% of affected children, presence of urethral valves, ureteroceles, diverticula and neurogenic bladder. VCUG is the investigation of choice for primary diagnostic procedures[10].