INTRODUCTION

Pelvi-ureteric junction obstruction (PUJO) is the most common pathologic cause of antenatal hydronephrosis (ANH) and occurs sporadically in 1 in 750-1500 live births, with a ratio of (2:1) in boys compared with girls, and the left side is affected in approximately two-thirds of cases. The condition occurs bilaterally in (10-46%).There are several theories about the development of PUJO, which may be congenital or acquired. The causes can be divided into intrinsic and extrinsic ***(Hashim and Woodhouse, 2012)***.

Renal ultrasound (US) is a widely available, relatively inexpensive, non-invasive, safe test that provides adequate anatomic visualization without radiation exposure. Given these exceptional qualities, it is widely used both in maternal-fetal sonography and for postnatal imaging ***(Groth and Mitchell, 2012)***.

Measuring the renal pelvis anterior-posterior diameter (APD) and Society for Fetal Urology (SFU) grading system are the most commonly used methods in the published data for determining the severity of prenatal hydronephrosis ***(Oktar et al., 2012)***.

If hydronephrosis is detected in the second trimester with APD (> 4mm), then repeated sonography is performed in the

third trimester to assess progression. For moderate to severe hydronephrosis discovered early in gestation with APD (>1cm), more frequent examination may be appropriate. If renal APD is (>7mm) in the third trimester, many recommend newborn evaluation ***(Pates and Dashe, 2006)***.

Most limiting is the fact that US cannot provide information regarding renal function. Technetium 99M renal scintigraphy is the present study of choice for estimation of overall and differential renal function. DTPA and MAG-3 can be used to estimate differential renal function and urinary drainage. DMSA is useful for the detection of differential renal function and clinically significant cortical lesions. Recently magnetic resonance urography (MRU) has been considered by some investigators to be a more reliable determinant of renal anatomy and function ***(Groth and Mitchell, 2012)***.

Most urologists manage the majority of the cases of fetal hydronephrosis due to PUJO by non-operative observation, reserving surgery only for patients with deterioration of renal function or clinical symptoms ***(Chertin et al., 2006)***.

Traditionally, the surgical repair involved an open surgical procedure that was performed retroperitoneally. Recently, a mini-incision approach has been described with equal results when compared to a more traditional standard incision. However, laparoscopy with and without robotic

assistance has become more prevalent for neonatal and infant PUJ repair ***(Herndon and Kitchens, 2009)***.

However, the natural history of fetal hydronephrosis, the optimal time for surgery, the ability to define which kidney will benefit from surgical intervention, and which children will have deterioration in renal function while on surveillance are still matters of controversy ***(Chertin et al., 2006)***.

AIM OF THE WORK

The aim of this work is to discuss the different diagnostic modalities of postnatal PUJO and the different options of treatment with emphasis on the indications of conservative and/or the operative management of this condition.

Methodology:

This will be done through review of recent literature discussing different lines of management.

ETIOLOGY AND PATHOPHYSIOLOGY

Pelviureteric junction obstruction (PUJO) is defined as a blockage or obstruction of urine flow from the kidney into the proximal upper ureter. This obstruction can lead to an increase in backpressure on the kidney, hydronephrosis, and progressive damage to the kidney function. The most common causes of antenatal hydronephrosis (ANH) are either transient or physiologic. PUJO is the most common pathologic cause of ANH ***(Hashim and Woodhouse, 2012)***.

Etiology:

It is sporadically in 1 in 750-1500 live births, but familial inheritance has been reported. PUJO has a ratio of (2:1) in boys compared with girls, and the left side is affected in approximately two-thirds of cases. The condition occurs bilaterally in (10-46%) of cases ***(Hashim and Woodhouse, 2012)*.**

There are several theories about the development of PUJO, which may be congenital or acquired. The causes can be divided into intrinsic and extrinsic.

1. Intrinsic abnormalities:

In the embryogenesis, the pelvi-ureteric junction (PUJ) is formed during the fifth week. By weeks 10-12 of gestation, the initial tubular lumen of the ureteric bud becomes recanalized, and the PUJ area is the last to recanalize. Inadequate canalization of this area is the main embryological explanation of PUJO. Researchers propose that improper innervations with diminished synaptic vesicles may be a factor in the development of PUJO.

This intrinsic obstruction is evident as the ureteral narrowing with angulation is found. During exploration, a catheter usually is passed to the renal pelvis without resistance, and this is evidence of the fact that the true narrowing is not a main pathologic change in PUJO. Some claimed the presence of remnant valvular mucosal folds, while others postulate the disproportionate abundance of longitudinal muscles as the cause of this condition.

The most attractive theory is that the obstruction is secondary to muscular discontinuity, which disrupts the coordinated motion of smooth muscle cells and may result in impeded transport of urine and blockage of the downward transmission of ureteral peristalsis. This absence or disorientation of smooth muscle fibers at PUJ is clearly evident on electron microscope evaluation with the findings of hypotrophy/hypertrophy of the smooth muscle and its

replacement with excessive collagen, combined with diminution of nerve terminals and nerves at the stenotic portion. ***(Lee and Han, 2009)***.

A study made by Solari V et al. identified altered expression of interstitial Cajal cells in obstructed PUJ specimens, which are normally intercalated between nerve terminal and smooth muscle cells providing a means of transducing signals from neurotransmitters and mediating neurotransmission. This suggests that PUJO may cause the failure of transmission of peristaltic waves across the PUJ, resulting in the failure of urine to be propelled from the renal pelvis into the ureter ***(Solari et al., 2003)***.

2. Extrinsic abnormalities:

Extrinsic obstructions secondary to bands, kinks, and aberrant vessels also are commonly encountered. In (40%) of cases, an aberrant, accessory, or early-branching lower pole segment vessel is found and observed to compress the ureter, causing mechanical obstruction. In this case, with the increased urine volume, the PUJ angulation with intrapelvic volume expansion causes increased resistance and obstruction. Further angulation may occur as it becomes adherent to an inflammatory process.

The presence of such a vessel in the vicinity of PUJ has gained recent attention after the advent of the endourological management. The anterior surface of the renal pelvis is

associated with a lower pole vessel in (65%) of cases, whereas the posterior surface is in contact with a vessel in (6%) of the kidneys examined. This information is relevant for the endoscopic incision of PUJ, making lateral incision the only safe option.

Patients with extrinsic obstructions present rather late in childhood, with intermittent abdominal or flank pain. Horseshoe or pelvic kidney, duplex collecting systems, and other rotational abnormalities also may cause PUJO. Cases of so-called high inserted ureter-to-renal pelvis exist, but this is presumed to be a secondary phenomenon to obstruction because the ureteral insertion seems to be higher in cases of dilated renal pelvis ***(Lee and Han, 2009)***.

Pathophysiology:

The urinary drainage from renal pelvis to ureter is determined by many factors. Urine volume and flow, the degree of PUJO, the functional capacity of glomerulus & collecting system, and the compliance of renal pelvis are the 4 main variables determining the pelvic pressure.

Koff proposed the concept of pressure- or volume-dependent flow. In instances of intrinsic obstruction, at low urinary flow rates, no obstruction exists; however, as the flow rate increases, the urinary bolus is not conducted, causing the renal pelvis to distend. This concept is called a pressure-dependent flow pattern.

On the contrary, in cases of extrinsic compression usually caused by aberrant vessels, urine flow is impeded only after a definite amount of urine is collected in the renal pelvis. This is an example of volume-dependent flow, and the pressure damage is only evident intermittently; thus, the degree of damage generally is less than that of intrinsic obstruction ***(Lee and Han, 2009)***.

Obstruction of the upper tracts in the acute phase leads to an increase in ureteric & renal pelvic pressures and renal blood flow. As ureteric pressure continues to rise, the renal pelvis dilates and renal blood flow decreases as a result of efferent arteriole vasoconstriction. In the long run, the ureteric pressure falls and renal blood flow decreases because of afferent arteriole vasoconstriction, leading to a decrease in the overall glomerular filtration rate. The dilation of the renal pelvis dampens the effect of the increase in pressure and results in tubular dilation, glomerulosclerosis, inflammation, and fibrosis of the kidney secondary to PUJO ***(Hashim and Woodhouse, 2012).***

PRENATAL MANAGEMENT

Prenatal upper tract dilatation may be a response to various events occurring throughout the developmental period and does not necessarily imply significant urinary flow impairment (UFI), which may affect the renal development and the renal function. The only difficulty is being able to identify during the prenatal and the neonatal periods those dilatations that may be the consequence of a permanent pathological event (i.e. significant UFI) requiring early specific treatment ***(Mure and Mouriquand, 2008)***.

A- Diagnosis of Antenatal hydronephrosis

Four main sources of information are available to identify prenatal urological anomalies which are:

1- Ultrasonography
2- Prenatal magnetic resonance imaging (MRI) scans
3- Prenatal biochemistry
4- Fetal karyotyping

(Mure and Mouriquand, 2008)

1- Ultrasonography:

Ultrasound (US) is a widely available, relatively inexpensive, Non-invasive, safe test that provides adequate anatomic visualization without radiation exposure. Given these exceptional qualities, it is widely used both in maternal-fetal

sonography and for postnatal imaging. Consequently, renal sonography has resulted in the increased diagnosis of prenatal and postnatal dilation of the upper urinary tract. It is highly accurate in the diagnosis of hydronephrosis ***(Groth and Mitchell, 2012)***.

- **Besides renal pelvis dilatation, prenatal sonography also gives other indications concerning the urinary tract as the:**

  (i) Presence of renal pelvis dilatation with or without ureteric dilatation.

  (ii) Echogenicity of the renal parenchyma. Bright parenchyma, cystic parenchyma, small kidneys and poor corticomedullary differentiation may reflect a structural anomaly of the kidney and may lead to an impaired renal function when bilateral.

  (iii) Amniotic fluid (AF) volume. Fetal urine production accounts for the majority of AF production in the second and third trimesters. Severe loss of urine production may lead to an oligohydramnios, which may be the consequence of either:

    (a) Mechanical urinary flow impairment

    (b) Deficient renal function as found in bilateral renal agenesis

    (c) Placental failure.

- The evaluation of the amount of AF around the child is subjective and very much examiner-dependent.

(iv) Visualization of the fetal bladder. Dilated upper tracts when associated with a persistent large bladder have a poor prognosis. They can reflect urethral UFI (posterior valves or urethral atresia), neuropathic bladder or complex cloacal malformations ***(Mure and Mouriquand, 2008)***.

During the antenatal period, at (16-20 weeks), US is performed to assess the AF volume to rule out oligohydramnios or any associated abnormalities and to measure bladder volume, kidney size, and the anteroposterior diameter of the renal pelvis (APDRP). The most sensitive time for urinary tract evaluation is (28 weeks). ***(Hashim and Woodhouse, 2012)***.

Sonographically, the fetal kidneys appear as two small circular structures adjacent to the lumbar spine in the transverse plane (Fig. 1) and are elliptical in shape when viewed longitudinally, spanning 4 to 5 spinal segments. With advancing gestation, the relatively echogenic retroperitoneal fat that surrounds the kidneys makes their visualization even clearer. The renal pyramids are echogenic as compared with the surrounding cortex, and they are arranged in anterior and posterior rows. The pyramids may be differentiated from visible or dilated calyces, which signal an obstructive process, in that the latter can be demonstrated to connect with the renal pelvis ***(Pates and Dashe, 2006)***.

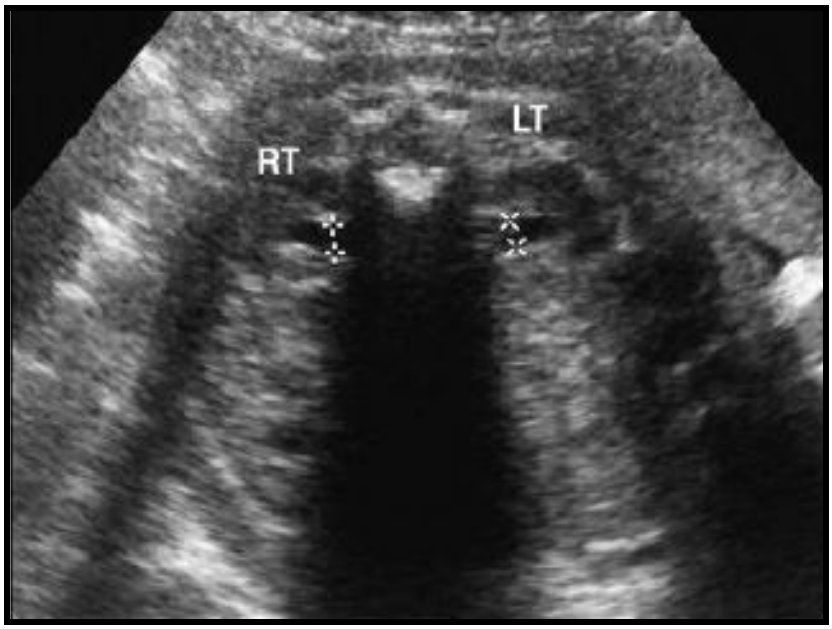

Figure (1): Transverse view of the fetal kidneys at 25 weeks of gestation, with calipers positioned to demonstrate how to measure the anterior-posterior diameter of the renal pelvis, taken from Early Human Development ***(Pates and Dashe, 2006).***

Even a small amount of fluid may be easily appreciated within the renal pelvis on routine ultrasound examination (Fig. 1). Differentiating a normal or physiologic accumulation of fluid from a pathologic process is done by measuring APDRP of the renal pelvis in the transverse plane. Measurements of (3mm) or less are considered normal at any gestational age. As renal pelvis measurements increase beyond this, prenatal evaluation of suspected hydronephrosis may be accomplished through a series of steps (Table 1) ***(Pates and Dashe, 2006)***.

Table (1): Sonographic approach to fetal hydronephrosis, taken from Early Human Development

1	How dilated is the renal pelvis?
2	Is the dilation progressively increasing?
3	Is the process unilateral or bilateral?
4	Is the amniotic fluid volume normal, increased, or decreased?
5	Are other structures, bladder or ureters, dilated as well?
6	Is there just one renal pelvis on longitudinal images, or is there a duplication of the collecting system?
7	Are calyces visible or even dilated, and if so, are we certain they connect to the renal pelvis (and that this is not a cystic renal abnormality)?
8	If there is dilation of calyces, is there cortical thinning?
9	If there concern, based on maternal age, serum screening, or other ultrasound findings, that this finding may raise the risk of fetal Trisomy 21 sufficiently, offering fetal karyotype analysis may be warranted.
10	If this is a complex genitourinary anomaly that cannot be completely characterized with ultrasound parentally, would fetal MRI be beneficial?

(Pates and Dashe, 2006)

According to Leung et al. there have been great inconsistencies in defining the maximum APD for physiological fetal pyelectasis as many factors such as growth, maternal hydration, maternal hormonal effect, fetal urinary

bladder fullness and temporal variation cause an increase in the APD of fetal renal pelvis. No systematic database has addressed the normal range of fetal renal pelvis in relation to quantifiable factors such as gestational age and fetal bladder fullness ***(Leung et al., 2009)***.

Although the classification of ANH remains somewhat subjective, several systems have been proposed in attempts to standardize this grading system, and these are used in varying degrees.

I. A qualitative scale:

Currently, the system most widely used to grade hydronephrosis is a qualitative scale, in which the degree is characterized as being mild, moderate, or severe.

II. Measuring the anterior-posterior diameter (APD):

The APD measurement is the most reproducible and most commonly used method of assessing renal pelvic dilation, most probably because of the sense of quantification it provides. Assessing collecting system dilation numerically might enable more objective characterization of hydronephrosis ***(Oktar et al., 2012)***.

If hydronephrosis is detected in the second trimester with APD (>4mm), then repeated sonography is performed in the third trimester to assess progression. For moderate to severe hydronephrosis discovered early in gestation with APD (>1cm),