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Leading Article

Dilema in Managing Children Born with Antenatally Detected Ureteropelvic Junction Obstruction (UPJO)

MD. HABIBUR RAHMAN

Abstract:

Now-a-Days, attitudes of paediatric urologists have been changed from surgical to observational regarding definite management of congenital ureteropelvic junction obstruction (UPJO), because many cases of UPJO goes away on its own before baby is born. In the last decade, this type of management trend has become progressively increased despite the lack of precise predictors of outcome. But these patients should be brought into regular follow-up by physical examinations, ultrasonography and nuclear imaging to observe resolution events. In some clinical scenario surgical intervention is inevitable such as solitary kidney or child with infection.

Now-a-days, it is a good practice by the modern pediatric urologists frequently consults with the guardians about advantage and disadvantage of observational and surgical management of antenatally detected hydronephrosis due to UPJO of their babies and their future outcome. As a result guardians feel comfortable in decision making.

Key words: *Ureteropelvic Junction Obstruction (UPJO), Hydronephrosis, Ultrasonogram, Diuretic renogram.*

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Introduction:

Ureteropelvic Junction Obstruction (UPJO) is a blockage that connects the renal pelvis to the ureter which drains urine to the bladder. In fetal life, congenital UPJO impair natal growth and development with baby is still growing in the womb, which is later on diagnosed as hydronephrosis in the second trimester by the ultrasonogram. Prior to fetal or prenatal ultrasonography, patients with congenital UPJO were usually diagnosed between 6 to 15 years of age due to their signs and symptoms. Only 14% of UPJO patients are less than 1 year of age.[1,2]

However, prenatal imaging has increased the rate of diagnosis of asymptomatic hydronephrosis due to UPJO. In this regard, it may be mentioned that previously corrective surgery was done to every child who presented with UPJO irrespective of functional status of the patient. But now a days, a multitude of surgeons have questioned the earlier operative procedure by demonstrating a high rate of spontaneous resolution of ureteropelvic junction obstruction. Unfortunately, still has no clear solution, which asymptomatic infants with hydronephrosis will lose renal function if left untreated.

Incidence of ureteropelvic junction obstruction: Ureteropelvic junction obstruction is the most common cases of antenatal hydronephrosis and has an incidence of 1 in 100-1500 newborns. Ureteropelvic junction obstruction is more common on the left side (2:1) and more common in boys than girls at a ratio 2:1. Bilateral UPJO is very rare. It is usually caused by stenosis of the proximal ureter at the ureteropelvic junction due to failure of development of renal smooth muscles, abnormal pyeloareterial intervention and impaired smooth muscles cell differentiation, less commonly by extrinsic compression of an aberrant accessory renal artery. More than 80% hydronephrosis are detected prenatally and out of them 39-64% are ureteropelvic junction obstruction (UPJO).[3-5]

1. Professor (Retired) of Pediatrics, Bangabandhu Sheikh Mujib Medical University, Dhaka

Correspondence: Prof. Md. Habibur Rahman, Professor (Retired) of Pediatrics, Bangabandhu Sheikh Mujib Medical University, Dhaka. Phone: 01711-381693. E-mail: adiba@dhaka.net

How UPJO Presents?

Presenting features of UPJO depends on degree of obstruction. Many mild cases do not have any symptoms at all. UPJO usually detected by prenatal hydronephrosis and this condition usually goes away on its own after birth[2]. Moderate to severe UPJO may be associated with pain, hematuria fever due to urinary tract infection, severe vomiting, anuria or polyuria, edema, palpable abdominal mass and later on systemic hypertension.[2,6]

Natural history of UPJO: Previously it was believed that most of the cases of UPJO with severe dilatation detected prenatally required surgical intervention, but later on several studies have shown high rate of spontaneous resolution or improvement even for grade 3 or 4 and it dilatation as defined by the Society for Fetal Urology (SFU) varied from 33 to 70%. Another observation of these studies was that a change in SFU grade 3-4 to 1-2 was considered significant and kidney may be without functional deterioration in future[7]. Data from previous studies shows that between 10 to 39% children with SFU grade 3 or 4 have reduced Differential Renal Function (DRF) <40 needs early pyeloplasty.[8-12]

How UPJO affects on the kidney: Obstruction begins to develop in the intrauterine life when organogenesis begins, which ultimately hampers growth of the number of nephrons which usually completed by 36 weeks of gestation. Normally no more nephrons are formed after birth. That is why premature babies are born with lower number of nephrons compared with babies born at term.[12]

So any insult leads to renal injury will be replaced by adaptive changes of the remaining nephrons to maintain Glomerular Filtration Rate (GFR) at first, but in the long term it lead to renal damage.[13] Moreover obstruction that originate harmful effects by altering the pathway of development of kidneys.[14] It has been observed on autopsy that fetuses due to UPJO on prenatal ultrasound have reduced number of glomeruli, reduced cortical thickness and increased fibrosis when compared to specimens from age matched fetuses with normal kidneys which is consistent with increased hyperechogenicely on prenatal ultrasound. But not all hydronephrosis will persist and impose same damage. Therefore primary responsibility lies on clinically to identify infants the more at risk of renal function impairment by applying all clinical and diagnostic tools.

Clinical risk factors of impaired renal function: It has been observed from many studies that a child with Differential Renal Function (DRF) has not deteriorated yet, will have batter long term result after operation, even lost function may be regained and may persist upto adult life. On the other hand early detection of surgical candidates is essential to reduce costs as well as stress for the families. The prediction of surgical candidate depends on parameters obtained from ultrasound and nuclear medicine imaging. Regarding renal function status difference more than 10% in hydronephrotic kidney at initial evaluation has 3 times more renal function deterioration and 2 times more symptoms development.[16]

Delay cortical transit time is also an independent risk factor for impaired renal function. Anterior posterior diameter (APD) is an independent predictor of resolution of hydronephrosis.[17] An APD of 24mm on initial evaluation along with 10% or more reduction of DRU or worsening hydronephrosis on are highly specific and sensitive predictions for surgical treatment.[18]

Shapiro and co-workers also introduced that hydronephrosis is selected with selecting the patients who will undergo resolution or worsening of hydronephrosis.[19] Rickard et al[20] showed that renal parenchymia to hydronephrosis are a ratio correlates with DRU and T ½ is very strong predictor in selecting children who will require surgery.[20]

How UPJO is diagnosed postnatally?

All patients who have symptoms of UPJO should have full set of complete blood count, serum creatinine, estimation of GFR and Blood Urea Nitrogen (BUN). A few patients may present with high level of serum creatinine and decreased (GFR). Neutrophil leukocytosis is present in case of infection. Urine routine test and culture should be done as recurrent infection is not uncommon in UPJO.

Imaging: Hydronephrosis is an incidental findings of ultrasonogram in the antenatal period which reflects underlying UPJO. Who have mild to moderate hydronephrosis on an antenatal scan, a follow up ultrasonography should be done after 48 hours to avoid transient neonatal dehydration period, but in severe cases, a scan should be performed within 48 hours which may be needed for urgent intervention.[21] Severity of hydronephrosis is evaluated based on the grading system of the Society for Fetal Urology (SFU) which varies from grade 0 (no hydronephrosis, intact central

renal complex) to grade 4 (Severe dilatation of renal pelvis and calyces with parenchymal atrophy and thinning) It may be mentioned that 20% of antenatally detected hydronephrosis is not found after birth[21].

Diuretic Renography: Renography is one of the most important diagnostic tool is used to determine the split function of each kidney to identify any renal evidence of obstruction. It is the gold standard to evaluate the severity of obstruction. The agent which is mostly preferred for paediatric population is technetium 99m mercapto acetyl triglycine (99m Tc-MAG3). The kidney is considered significantly damaged if the split renal function in one kidney is less than 40% of the total kidney function, correlating to the half-life (T 1/2) of the agent [22,23]

Intravenous pyelography (IVP): IVP gives information about the degree of dialatation of pelvis in comparison to the contralateral side. By IVP during the excretory phase, the excretory function of the affected kidney is compared with the normal kidney. But now a days IVP is not commonly practiced except in few candidates like urolithiasis and pre and operative evaluation where others diagnostic tools are not available

Voiding cystourethrogram (VCUG): VCUG is an essential part of basic evaluation of patients with hydronephrosis. It is a gold standard diagnostic tool for detecting posterior urethral valve, vesicoureteral reflux ureterocic and bladder diverticula. VUR can contribute to additional information due to ureteral dilatation. So VCUG should be done as early after correction of any existing infection.

Magnetic resonance Imaging (MRI): MRI is considered as an ideal investigation for assessment of detecting anatomy of ectopic ureteral infection, complicated duplex system or when other complex condition are suspected. It provides excellent anatomic and functional detail of the collecting system and ureteral structure in children can also be diagnosed accurately by MRI[24].

How UPJO is managed?

Though majority of UPJO patients will have resolution of their hydronephrosis, however a minority will not improve and may be best managed with surgical intervention to preserve the renal function. Early signs of worsening obstruction should be promptly diagnosed by the experienced paediatric nephrologist Passoniet at and weitz et al showed in their series that majority of the patients with UPJO can be managed conservatively with close observation and imaging. They advised postnatal ultrasound should be done between 48 to 72 hours of their birth in a well hydrated infants to avoid masking of hydronephrosis. In some special situation like bilateral hydronephrosis, solitary kidney, history of maternal oligohydramnios earlier ultrasonography is recommended. Change et al observed in their study that up to 17% UPJO under go spontaneous mutation. In first week of life among all patients of their series only 2% had severe dilatation (715mm) and remaining were in mild (5-9mm) and moderately dilated and 90% of them either resolved up to 2 years of age and 10% of them had persistent moderate to severe dilatation.

It has been observed that 25% infants with severe dilatation (715 mm) on initial evaluation will remain stabilized or resolve to normal or mildly dilated (5-9m), 50% stabilized with moderate dilatation with their first 2 years and remaining 25% severely dilated[27]. So, these patients should be closely followed up by serial ultrasonogram and duratic sonogram for assessment of improvement or deterioration.

Question may arise:

Who is the absolute candidate for surgical treatment? The patient who is symptomatic like recurrent urinary tract infection despite antibiotic prophylaxis, hematuria, kidney stones, mass effect due to severely dilated kidney, solitary kidney with reduced renal function prior to surgery, children with confirmed severe hydronephrosis on postnatal imaging should undergo MAG3 study which will dictate the future plan of treatment by evaluations differential renal function. If initial function is above 45%, ultrasound evaluation should be repeated at 3 and 12 months. If initial differential function is below 35%, obstruction should be released by surgery within first 2 years of life [28]. It has been observed that early surgical treatment in newborn with severe UPJO is safe and improve renal function status [29]. Surgical treatment for UPJO is still considered as the gold standard method and it was observed in a retrospective analysis of 343 those who were initially treated conservatively 50% of them later on required pyeloplasty [28].

Other newer surgical methods practiced now a days for the treatment of UPJO are neonatal pyeloplasty, endoscopic endoluminal pyeloplasty and robotic pyeloplasty [25,26,30]. All the newer methods of surgery showed good results, low morbidity, resolution or

improvement of hydronephrosis. But limitation of these methods are higher grade technology and non-availability in many countries. Another method is percutaneous nephrology (PCN) as bridging until surgery. Today PCN is restricted to untreatable hydronephrosis severe biletrial obstruction with renal insufficiency and in a severely obstructed single kidney with elevated serum creatinine.

How UPJO patients are followed up?

Though long term outcome of UPJO patients are good but it is essential every patient should be under follow-up for long term has been observed that a few patients after pyeloplasty may develop hypertension and chronic renal insufficiency. Boubaker et al [31] observed that out of 33 patients 14 patients had persistent abnormal renal function on radionuclide scan after pyeloplasty due to UPJO.

They also observed that improvement of renal function was better in less than 1 year compared with more than 1 year [31]. Valler PG et al[32] observed that surgery for grade IV hydronephrosis or delay in surgery develop intestinal fibrosis and reduced glomerular rate and hypertension. Interstitial fibrosis found in kidney biopsy in UPJO patients and hypertension co-relates significantly with the differential renal function of the affected kidney is also determinant of progressive renal damage[33]. It is critical to monitor blood pressure and treat properly. So this data strongly support the needs for long term follow up in less severe cases of UPJO to determine the long term prognosis.

During follow-up patients family should also be counseled to maintain adequate hydration, sodium repletion during hot weather. Parents should also be advised to avoid nephrotoxic drugs and non-steroidal anti inflammatory to prevent progression of the renal disease and development of CKD, regular of the time of diagnosis and surgical intervention.

So it can be summarized whole principles of follow-up rules as follows:

- Monitor blood pressure and prevent hypertension
- Monitor urinary protein excretion and urinalysis
- Monitor renal function
- Renal ultrasound
- Prevent urinary tract infection
- Avoid dehydration
- Avoid nephrotoxic drugs
- Diuretic renogram if required

Conclusion:

Due to broad spectrum of UPJO in children and different variables like age of the patient, degree of severity, ranging from mild asymptotic to severe kidney injury before and after birth, unilateral or bilateral obstruction, mild, moderate and severe degree of antero-posterior dilatation, impaired renal function status each patient required individualized clinical management. Because morbidity of mild cases may not manifest until later in life. These patients should be closely followed up throughout the life with proper diet, maintaining nutritional status, growth blood pressure monitoring, assessing renal function, proteinuria and regular ultrasound 3 to 4 times in a year.

Close partnership between paediatric nephrologist and paediatric urologist is essential to integrating medical, observational, robotic and surgical management of the patient as well as transitive to the appropriate specialties when the patient reaches adulthood. It is very essential and mandatory that every step of present and future management and their outcome of the patient should be informed to the parents by proper counseling.

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