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Review Article

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A Narrative Review of Contemporary Challenges and Controversies in the Management of Congenital Pelvi-Ureteric Junction Obstruction (PUJO)

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Abstract

Pelvi-ureteric junction obstruction (PUJO/UPJO) is a relatively common presentation which may be identified at any age. The detection of this condition has grown exponentially with the widespread increase of imaging. This new surge in detection presents a unique challenge to the treating clinician to determine the optimal strategy for evaluation and intervention. The challenge is balancing early intervention whilst minimizing overtreatment. We present a narrative review of existing evidence on management of PUJO from detection, evaluation pathway, intervention, follow-up and management of complications with special emphasis on technical nuances on surgical management of PUJO for the practicing urologist.

Keywords: Pelvi ureteric junction obstruction, pyeloplasty, surveillance

1. Introduction

Pelvi-ureteric junction obstruction (PUJO/UPJO) is a relatively common presentation with an incidence of approximately 1 in 1000 to 1500 live births (1). This condition may be diagnosed with antenatal to geriatric age groups. Although most commonly congenital in etiology there are acquired causes that lead to delayed PUJO (2). Delayed presentation of congenital PUJO is likely due to initial partial obstruction progressing into a complete obstruction either acutely (such as infection) or chronically (chronic inflammation or hypertrophy). Optimal management is complex and depends on multiple variables, including age, degree of obstruction, risk of obstructive uropathy at the time of detection, symptomatology, and complications such as infection or stone formation.

The major concern with any obstruction to the urinary tract is deterioration of renal function. The objective of current management of PUJO is to identify the patients at risk of deterioration of the renal function and prevention of complications whilst avoiding overtreatment based on mere image-based diagnosis of this entity. Once decided on active surgical intervention the options are open, laparoscopic (with or without robotic assistance) and endoscopic. Pyeloplasty which is currently considered the standard of care with success rates over 90% it is a challenge to the operator to decide which technique would give the best result on a particular variant of PUJO. We plan to discuss the controversies, diagnostic evaluation and personalized surgical and non-surgical management options based on existing evidence from the perspective of a practicing urologist.

2. Evaluation pathway

Once presented with loin pain (exacerbated with fluid intake or diuretics), superadded pyelonephritis or incidental renal stones and rarely unexplained young hypertensive or as an incidental diagnosis, apart from basic urinalysis and global renal function assessment, all international guidelines approve the following special investigations.

2.1. **Focused ultrasonography (US -KUB)**

Ultrasonography (US-KUB) is the recommended first-line noninvasive investigation for detecting hydronephrosis based on age-specific renal pelvic diameters. It can also provide additional information, such as the presence of ureteric jets (frequency, duration, and velocity) with color Doppler. Several studies have demonstrated the utility of USS alone to reliably distinguishing obstructive from non-obstructive hydronephrosis (3,4).

2.2. **Contrast enhanced urography (CT-U)**

Contrast-enhanced urography (CT-U) with an angiography phase provides detailed anatomical information, including cortical thinning, calyceal clubbing, and the location of the PUJ (dependent vs high insertion). It also identifies duplex and horseshoe kidney variants and the presence of aberrant vessels, which are vital considerations in surgical planning.

Moreover, CT (and MRI) can demonstrate secondary PUJO caused by underlying primary conditions such as retroperitoneal tumors, fibrosis, calculi, or PUJ ureteric kinks secondary to reflux uropathy, which can significantly alter the management approach. Information regarding differential renal function obtained from CT can serve as a surrogate for isotope studies (5). This is particularly true in cases of nonfunctioning renal units with severe cortical thinning. However the reverse is not always true as passage of contrast into the ureter in the pyelography suggests some degree of patency it doesn't safely rule out clinically significant obstruction and functional imaging is usually recommended (6).

2.3. **Nuclear renography- Tc 99 DTPA/ MAG-3**

This is considered as the gold standard in functional evaluation of urinary obstruction. Nuclear renography diagnoses renal drainage and obstruction which could exclude the diagnostic dilemmas such as extrarenal non obstructed “Baggy pelvis”. Out of the two radiotracers Tc 99-MAG-3 is actively secreted from the proximal renal tubules, and safer for patients with lower renal function than DTPA (55% of MAG-3 is excreted vs 20% excretion in DTPA). Tc^{99m}-DTPA is the most used radionucleotide filtered from the glomerulus and better for calculating fractional eGFR than MAG-3. Interpretation of the nuclear renal imaging should be done with due consideration for the tracer used as well as the protocol followed.

The addition of diuretics (e.g. Frusemide -F) improves the sensitivity but the optimal timing of administration (F+20, F+0 or F-15) remains controversial. It must be kept in mind that later administration(F+20) of diuretics allows for visualization of the whole response curve whilst earlier administration (F+0 or F-15) increases the sensitivity of the study to differentiate equivocal cases. It is agreed currently that failure to clear 50% of the tracer (T1/2) by 20 minutes is suggestive of urinary obstruction. Interpreters should be aware of false positive results caused by dehydration, massively dilated renal pelvis, high-grade reflux, pelvic kidneys and in cases where a distended thickened bladder which cause false obstruction.

2.3. **MR-urography- (MRU)**

MRU can give excellent anatomical and functional images and lacks the risk of ionizing radiation. There is a strong body of evidence suggest MRI done with administration of Gadolinium-diethylenetriamine-pentaacetate-dimeglumine (Gd-DTPA) in conjunction with Frusemide could replace both CTU and radionucleotide imaging as a single comprehensive diagnostic modality in evaluation of PUJO (7). Other studies have demonstrated MRU to be equivalent to renal scintigraphy in assessing differential function (8).

It is evident that the failure of MRI to replace the conventional imaging is largely due to the cost, availability and complexity of software required for this analysis, with some concern about rare but lethal complications of Gadolinium (nephrogenic systemic fibrosis) especially in patients with significantly impaired renal function. However, the strong tendency in future is for MRI to replace conventional imaging as the single modality of evaluation of all upper urinary tract obstructions.

3. Indications for intervention

It is interesting that the even centuries after identifying the entity no consensus statement or universally accepted guidance could be found on absolute indications of intervention for PUJO. The goal of management is to personalize the treatment to detect and prevent continuous renal parenchymal injury and complications whilst avoiding overtreatment.

All the clinicians managing the PUJO must be thoroughly knowledgeable about the following generally accepted guidelines to counsel their patients properly to avoid overtreatment and the serious postoperative complications associated with the threat of litigation and negligence.

3.1. **Intervention based on symptomatology**

Attributable loin pain-especially if related to fluid or alcohol intake is an indication to intervene. However, it is important to exclude other significant causes of loin pain such as myo-fasciitis, disk prolapse especially in adults with congenital PUJO with well-preserved differential renal function.

3.2. **Deterioration of differential function**

Less than 40% differential function (i.e., >10% drop) or a progressive deterioration of function of the affected kidney on serial imaging. Certain authors advocate using the individualized GFR of the affected renal unit for serial monitoring as the differential function is confounded by the state of the other kidney. There is no consensus on the interval of serial assessments but generally accepted as 24-36 months.

3.3. **Development of complications**

Secondary stone formation. The evaluation for primary PUJO in the presence of stones is difficult. It is also difficult to establish the relationship of the cause and the effect. When the patient presents with stones and hydronephrosis disproportionate to the size and location of stones, the stone shape (globular smooth stone) and presence of high riding PUJ gives a hint for preexisting occlusion leading to the calculus.

The current recommendation is to initially manage stones and evaluate the PUJO once stone-free. However, there is increasing evidence for the safety and efficacy of concomitant stone removal with PUJ repair if it is highly suggestive of PUJO as the primary disease (9).

3.4. **Recurrent urinary tract infections and unexplained or refractory hypertension**

Except episodes of pyelonephritis, non-ascending unrelated recurrent bladder infections especially in females does not warrant PUJO intervention. PUJO is a known rare cause of hypertension due to an unknown mechanism. PUJ repair may help in a minority of patients with unexplained hypertension (other causes like renal artery stenosis and medical diseases are excluded) who have become refractory to standard antihypertensives once.

4. Controversies in surgical interventions

4.1. **Active surveillance vs Surgical intervention**

In a large series evaluating outcomes of surveillance, only 30% of adults undergoing non-operative management needed subsequent intervention (10). There is a serious paucity of evidence and guidance on the conservative management strategies including the diagnostic test used, as well as the interval and total duration of surveillance. This is particularly true for children and younger adults planned for surveillance as the risk of repeated testing and the harm of radiation should be balanced with the risk of missing a potentially curable condition at the stage where further progression can be prevented. However, close observation at a specialist centre has been described as a suitable option in adult as well as pediatric populations when there are no strong indications to intervene.

4.2. **Surgical approach of PUJ repair**

4.2.1. **Open vs Laparoscopic vs Robotic assistance**

Historically open dismembered (Anderson-Hynes) pyeloplasty was considered the standard of care in the management of PUJO with all subsequent treatments being compared against it. With the development of minimally invasive technologies, laparoscopic and robotic assistance has posed strong evidence-based challenge to take up this position with non-inferiority and added advantages of minimal access surgery.

The laparoscopic and robotic approaches have shown to be superior with regards to blood loss, analgesic requirement and length of stay than open approach in systematic reviews looking at both retrospective and prospective studies (11,12). Current evidence proves similar long-term outcomes in all three approaches in most of the large-scale studies. With the limiting factors of cost, training and expertise becoming challenges in minimally invasive methods.

Interestingly several studies done in the west have shown when considering a comprehensive cost analysis, laparoscopy is more cost effective than robotic as well as open approaches (13). Reduction in analgesic requirement, shortened hospital stay, reduced incision associated complications (early and late) are likely to be the contributor to laparoscopic approach being the most cost effective for this particular procedure (12).

4.2.2. **Trans peritoneal vs Retro-peritoneal approach**

Both transperitoneal and retroperitoneal approaches are employed for minimally invasive pyeloplasties. The retroperitoneal approach theoretically mirrors the open approach while reducing the potential risks associated with bowel handling and peritoneal complications, such as postoperative ileus and adhesions. Additionally, it offers the advantage of confining any potential urine leaks to the retroperitoneum, thereby minimizing the need for further interventions. However, the retroperitoneal approach is technically challenging due to limited working space. Another drawback is the difficulty in cases involving crossing vessels, which require suturing behind the vessel after transposition of the PUJ, given the posterior approach.

Despite these considerations, evidence has not demonstrated a definitive benefit of one approach over the other. A recent meta-analysis revealed that most outcomes, including postoperative complications, hospital stay, and success rates, are comparable between the two approaches. However, the transperitoneal approach was associated with significantly shorter operative times, while the retroperitoneal approach had shorter durations of external drainage (14). In light of this and other studies, the retroperitoneal approach appears to offer marginal benefits in pain scores and a slight reduction in urine leak complications, albeit at the expense of a steeper learning curve and greater technical difficulty.

4.2.3. **Dismembered vs non dismembered (Culp, Fengers vs. Foley's Y-V vs Flap) pyeloplasty**

The gold standard reconstructive procedure has been the dismembered technique described by Anderson-Hynes initially in 1949 which is now effectively replicated with minimally invasive techniques (15). Non dismembered techniques have been described including the Y-V plasty and flap techniques (vertical or spiral). Limited evidence comparing these techniques to each other slightly favours dismembered techniques (16). Y-V plasty is practically more useful for high insertion of the ureter or intra pelvis. Flap techniques retain a place in instances of a large renal pelvis with longer segments of narrowing as in cases of recurrent PUJO and small caliber ureters. Urologists need to be knowledgeable and conversant in all these techniques and indications to prevent post operative disasters such as restenosis and prolonged urine leakage.

4.2.4. **Suturing method- Interrupted vs. continuous suturing**

Another contentious topic in PUJ repair is the choice between continuous and interrupted suturing techniques. Traditional open surgical repair utilized interrupted suturing, but continuous suturing has gained popularity due to its reduced operative time and fewer surgical knots. A recent meta-analysis found a significant reduction in operative time (mean difference ~33 minutes) and length of stay (mean difference ~1.08 days) favoring continuous suturing, with similar complication rates and overall outcomes for both techniques (17).

Recently, special emphasis has been placed on preventing ischemia at the apex of the anastomosis, which is at the highest risk of stenosis. The practice of using two separate sutures on either side of the distal apex and continuing them proximally on the anterior and posterior walls of the anastomosis is now recommended by experts in the field.

4.2.5. **Suture material used for anastomosis**

Various absorbable suture materials have been utilized and compared in the literature, without demonstrating clear superiority of any specific type. Commonly used materials include absorbable monofilament sutures (AMS) such as Monocryl and PDS, which slide through tissue with minimal risk of damage. Conversely, barbed sutures, also known as self-retaining sutures (SRS), offer the perceived advantage of maintaining tension and reducing the need for knots Braided sutures, which predate AMS and SRS, have proven their efficacy over time, and

some studies advocate for their continued use in contemporary practice due to their favorable properties in upper tract reconstruction (18).

In the absence of definitive evidence favoring any particular suture material, it is reasonable to conclude that the surgical suturing technique is the most critical factor influencing outcomes, rather than the individual suture material (19). However, it is important to recognize the subtle technical nuances associated with each type of suture material for PUJ repair, as they are not necessarily interchangeable.

4.2.6. **Post-operative drainage**

The optimal method and duration of drainage of the collecting system post-repair remain subjects of debate. The three primary options include internal stenting, external drainage via nephrostomy, and no stenting. Several studies have investigated the optimal drainage method but have failed to reach a consensus. A network meta-analysis in the pediatric population found no significant differences in operative time, success rates, hospital stay, renal function improvement, or overall complications among the three methods. However, the analysis recommended the use of an indwelling DJ stent due to its efficacy in reducing postoperative urine leaks, albeit with a slight increase in postoperative pain compared to the stentless technique (20).

The duration for stent placement is also contentious, with the standard practice being 4-6 weeks. Recent studies are exploring early stent removal, with a recent randomized controlled trial demonstrating equivalent outcomes and improved postoperative symptom control with stent removal as early as one week postoperatively (21). The drain is recommended to be removed once the output is <50cc/ 24 hours.

4.3. **Place of percutaneous or ureteroscopic endopyelotomy**

A recent systematic review of published data on endopyelotomy reported an overall success rate of 75% at a median follow-up of 29 months (22). However, comparative studies between endopyelotomy and surgical repair have demonstrated a preference for surgical correction due to superior overall success and durability (23). Additional studies have identified that high-grade obstruction (severe hydronephrosis), longer segments of occlusion, and the presence of crossing vessels are factors associated with a higher risk of failure of ureteroscopic endopyelotomy (24,25). Despite these limitations, endopyelotomy remains a viable option for highly selected patients, particularly those with recurrent PUJO with periureteric fibrosis who are not ideal candidates for reconstructive surgery.

5. Controversies in management of specific complications

5.1. **Postoperative urine leak**

When managing post-operative high drain outputs, it is crucial to differentiate between collected intra-operative irrigation fluid or peritoneal fluid from a true urine leak. This is done by checking drain fluid creatinine to serum creatinine ratio (drain fluid value of at least 10-30 times the serum value confirms urinary in the drain).

Once confirmed, supra-anastomotic drainage with a percutaneous nephrostomy ensuring the least resistance to anatomical drainage gives the best chance of healing. The first step will be IDC insertion, then a nephrostomy may be attempted (though it may technically be difficult due to the non-dilated system) to reduce the leak and ensuring healing. Other tricks of the trade include inserting a second parallel DJ stent under fluoroscopic guidance across the anastomosis and gradual shortening of the peritoneal drain when there is direct adhesion of it to the suture line.

5.2. **Challenge of restenosis and recurrent PUJO**

Recurrence can be symptomatic, radiological, or functional. Functional obstruction is defined as post-operative diuretic renogram has not returned to baseline despite contrast drainage in the urogram with symptomatic improvement. It is hypothesized this atonic non-obstructed renal pelvis is probably a result of renal pelvis denervation caused by prolonged dilatation, extensive stripping of the adventitia with neuro-vascular lining of the renal pelvis in preparation for anastomosis may contribute to the denervation of the renal pelvis. The operators should be aware of the recommendation to strip the adventitia only at the anastomotic surfaces to prevent renal pelvic neuropathy.

Retrograde pyelography and diagnostic ureteroscopy are recommended to confirm structural obstruction and restenosis as imaging results could be equivocal. The recommendation for structural restenosis is redo surgical pyeloplasty. Some authors suggest endo-pyelotomy as an initial treatment given the technical difficulty of redo surgery. However, it has shown inferior outcomes compared to redo pyeloplasty in some good quality comparative studies (26). However, redo pyeloplasty with either dismembered or utilizing a flap technique can be performed safely and with good outcomes though more technically challenging than primary cases (27). In cases of severely fibrosed, recurrent un-reconstructable PUJO, a uretero-calycostomy provides a viable and versatile salvage option.

6. Postoperative follow-up

No consensus has been reached in any guidelines at present regarding the optimal follow-up regime. Most guidelines recommend follow-up for 2 years after surgical repair since it is evident that the majority of recurrences occur within that period (28). However, others have reported late failures even up to 10 years.

The initial follow-up is usually with renal imaging (ultrasound or CT-Urogram) plus a radionucleotide renal scan at 3 or 6 months. However, there is significant heterogenicity of the interval for repeat scans as well as a threshold for further investigation and treatment.

Given the paucity of evidence, a reasonable approach is to follow up for up to 2 years and investigate if symptomatic afterwards.

7. Conclusion

Pelviureteric junction obstruction (PUJO) is a prevalent clinical condition detectable at any age. Due to the increased availability of diagnostic imaging, a significant number of asymptomatic PUJOs are detected. No clear evidence is present to predict the patients who are likely to progress into renal injury secondary to the obstruction which makes the decision to intervene less straightforward. In the absence of strong indications to intervene close follow-up is paramount to prevent silent deterioration of the renal parenchyma. Emerging evidence suggests that combination of conventional contrast and radionuclide imaging may soon be replaced by MR-urography with a functional protocol, offering a single comprehensive diagnostic test.

In cases of symptomatic or complicated PUJO, where surgical correction is planned minimally invasive pyeloplasty with or without robotic assistance using a dismembered technique seems to offer the best outcomes in the current context. Recurrent or failed repairs can be managed with operative revision which shows superior outcomes compared to less invasive endopyelotomy. Despite the technical challenges, the outcomes of redo repairs in expert hands are comparable to primary repairs, underscoring the importance of surgical expertise in achieving optimal results.

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