

# Clinical, surgical and morphological assessment of the pyeloureteral syndrome

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## Abstract

Obstruction of the pyeloureteral junction (PUJ) is by far the most common cause of hydronephrosis in children, with an incidence of one in 1000–2000 newborns. Also, the obstruction of the PUJ is the most common cause of prenatal hydronephrosis, accounting for 80% of the cases. The aim of this study is to observe and discuss first the efficacy of described surgery procedures and second the microscopic modifications of the PUJ (abnormalities of smooth muscle tissue, inflammation and fibrosis). One hundred and eleven children with a diverse urological pathology with an average age of 11.57 years were operated between 2011 and 2015 in Urology Clinic of Oradea, Romania. Of these, 20 children (11 boys and nine girls) with congenital hydronephrosis by junction syndrome required surgical correction. The surgical techniques used were Anderson–Hynes dismembered pyeloplasty, non-dismembered Scardino procedure and the Hellström procedure. Operator interventions were performed by subcostal laparotomy with or without partial XII rib resection. The average operator time was between 40 and 50 minutes. None of the patients required blood transfusions. Average hospitalization was seven days. All patients were monitored through the Ambulatory Pediatric and Urological Service. Anderson–Hynes operation is the main procedure to solve the obstructive syndromes of the PUJ. It can be performed without stentings of the ureter as originally described by the author, but also by protecting anastomosis with a urinary diversion, such as JJ catheter, ureteronephrostomy or ureteropyelostomy. Even though clinical and imaging studies are sufficient for diagnosis of PUJ syndromes, morphology and histology bring essential data regarding the age of the lesions.

**Keywords:** congenital hydronephrosis, pyeloureteral junction, pyeloplasty, Anderson–Hynes, junction syndrome.

## Introduction

Hydronephrosis is a term often used by specialists for a distension of the pelvis and calyces due to an obstruction which untreated leads to a progressive deterioration of the kidneys [1, 2]. There is still discussion about how dilation of the prenatal and postnatal urinary system should be classified [3]. The most frequently found cause of hydronephrosis is ureteropelvic junction (UPJ) obstruction with an estimated incidence of one in 1000–1500 cases [3]. UPJ obstruction is mostly considered as a functional obstruction originating from abnormalities in the smooth muscle of the pelvis and ureter [4, 5]. The UPJ by crossing vessels is also observed but whether the vessel alone causes obstruction or whether there is also a functional component is still debated [6].

In the usual practice, information about the condition of the prenatal urinary system is known by the urologist only when the pregnant woman has a urological problem. The information is provided by the gynecologist who can detect by ultrasound in the week 15 bladder and kidney, and in weeks 18–20 an echogenic central zone (renal sinus) [7].

The urologist should be present at birth for each patient known with prenatal hydronephrosis, especially if it is a single congenital and/or obstructed kidney. In such

cases, urinary drainage is mandatory by a minimally invasive method (pyelostomy or nephrostomy) [8, 9].

## Aim

The aim of this study is to observe and discuss first the efficacy of described surgery procedures and second the microscopic modifications of the pyeloureteral junction (PUJ) (abnormalities of smooth muscle tissue, inflammation and fibrosis).

## Patients, Materials and Methods

Between 2011 and 2015, 111 children with a diverse urological pathology with an average age of 11.57 years were operated in Urology Clinic of Oradea, Romania, 20 children for congenital hydronephrosis due to obstructed PUJ. All 20 patients were operated in general anesthesia by lumbar approach. In three patients, the approach was on the XII rib with partial rib resection. There were 13 interventions on the left side and seven interventions on the right side (11 boys and nine girls). There have been 14 Anderson–Hynes (AH) interventions, two Scardino & Prince procedures, two combined-ureter surgery (Hellström surgery and AH surgery).

All children were enrolled after parental consent. A detailed clinical assessment including history and relevant

physical examination was done as initial step followed by laboratory tests including complete blood picture, urinalysis, serum creatinine, urea, electrolytes and ultrasound kidneys, ureters and bladder (USKUB) in all cases.

All of these patients were recommended by our pediatric colleagues, most of the interventions were performed in teams with pediatric surgeons in the Operating Room and with the tools of the Pediatric Surgery Clinic at "Dr. Gavril Curteanu" Municipal Hospital of Oradea. In all patients, retrograde voiding urethrocytography was performed under fluoroscopic guidance to exclude associated malformations, such as reflux and bladder and ureteral abnormalities. Retrograde ureteropyelography (RUP) was performed in only three cases.

In two patients, surgical specimens were collected for histopathological examination. Tissue fragments undergo immediately fixation in 10% buffered formalin for at least 24 hours and were processed histological using paraffin embedding technique. With a manual rotational Leica microtome, 4  $\mu$ m-thick tissue sections were displayed on regular slides, stained using Hematoxylin–Eosin (HE) and examined with Leica DM1000LED optical microscope. Gross pictures were taken using Nikon D810 camera and histology pictures were taken with the camera attached to the microscope.

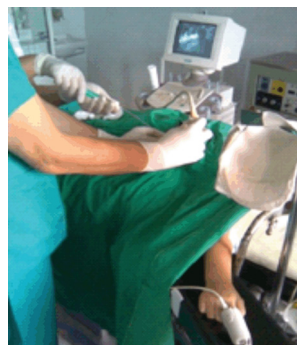
## Results

All patients undergoing dismembered and non-dismembered pyeloplasty have required a surgery time of 40 to 50 minutes. No patient had surgery complications and did not require blood transfusion. Hospitalization in the vast majority of cases lasted no longer than seven days. All patients were monitored and followed-up by pediatric and urology personal. We had a patient who developed a postoperative urinoma due to the clogging of the retroperitoneal drainage tube that required percutaneous drainage under ultrasound control (Figure 1). Another patient required a post-surgery ultrasound-guided percutaneous nephrostomy due to the jug of the JJ catheter on the ureter.

A third patient required a repeated insertion of the nephrostomy under fluoroscopic control after ureter dislocation. It required a longer period of hospitalization due to febrile syndrome and required antibiotic therapy.

Another patient operated from other hospital required

secondary ureterolysis due to the persistence of hydronephrosis. Intraoperative dissected and isolated the ureter from the scar tissue probably resulting from extravasation from the first intervention, with the protection of the ureter's adventitia in order not to compromise its circulation. In addition, the ureter has been fattened to provide protection and mobility. The majority of patients had a good post-operative progression. Dilatation of the pelvis and renal function improved over the next three months.

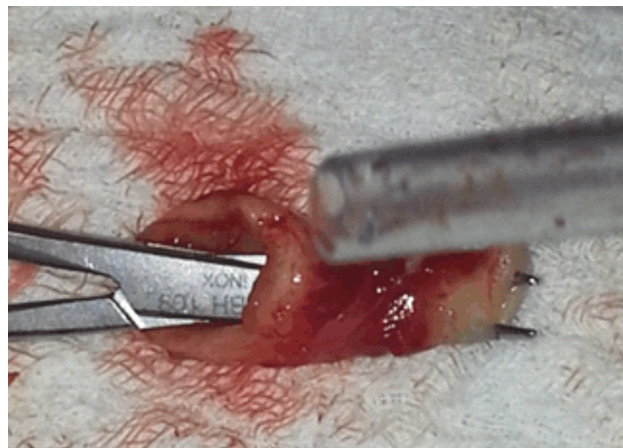


**Figure 1 – Post-surgery ultrasound-guided seroma drainage after Anderson–Hynes dismembered pyeloplasty.**

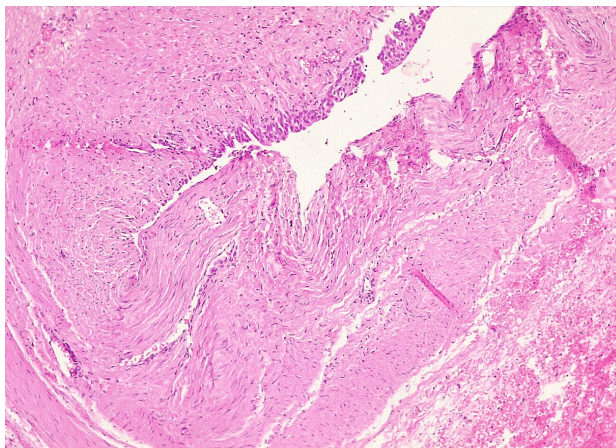
Except for two interventions without drainage in two girls, in all other cases we used (most often) drainage with ureteral JJ catheters, but also nephrostomy or pyelostomy were used.

Both tissue specimens examined morphological had threadlike appearance of 2/1.5 cm and 2/1 cm with elastic structure. On cut surface, both were hemorrhagic (Figure 2). Both fragments were processed entirely.

Microscopic examination confirmed signs of obstructive uropathy on transversal sections with reactive and disordered transitional epithelium with foci of ulceration (Figure 3). Beneath the urothelial layer, we can appreciate recent hemorrhage, signs of acute (represented clearly with neutrophil margination phenomenon, as seen in Figure 4 and subacute inflammation, proliferation of collagen bundles in muscle layer (Figure 5), fibrosis, hypertrophic muscle fascicles. In the adventitial space, we found abundant hemorrhage (Figure 3). On longitudinal sections (Figure 6) of the proximal ureter, we can highlight the loss of surface epithelium with sharp demarcation excluding dysplastic features that can appear in some transversal sections due to some technique difficulties as shown in Figure 7.

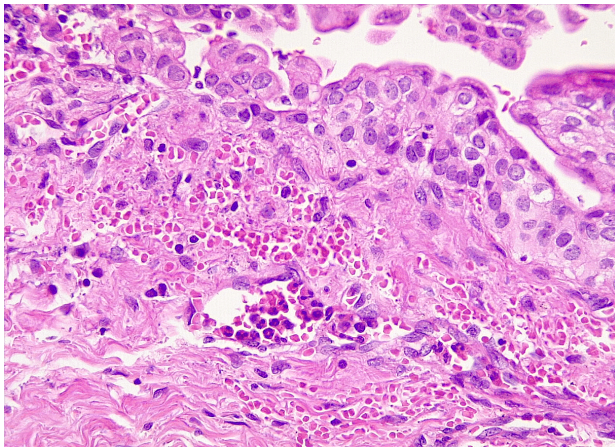


**Figure 2 – Excised pyeloureteral junction with gross examination on fresh tissue, having a hemorrhagic outer surface.**

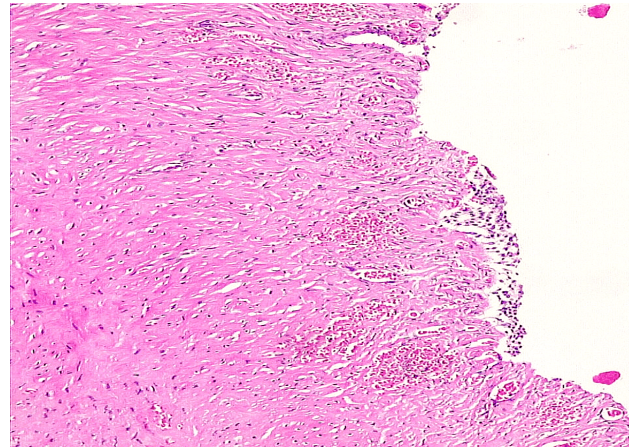


**Figure 3 – Cross-section from proximal ureter showing loss of urothelium, fibrosis in the muscle layer, and blood extravasation in the adventitia (HE staining,  $\times 100$ ).**

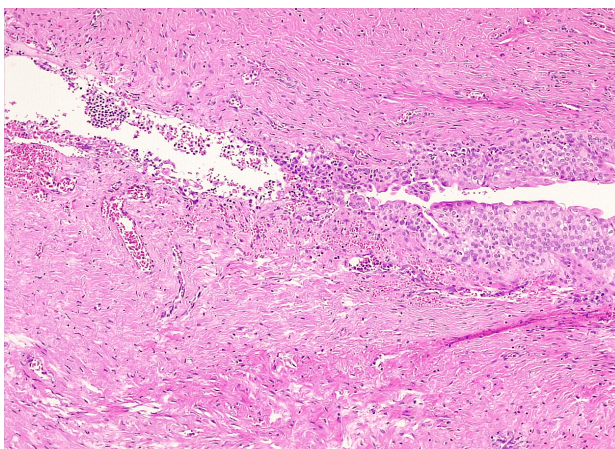




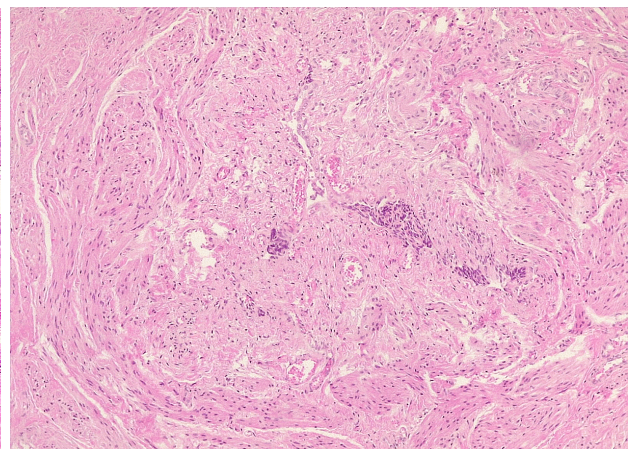
**Figure 4** – Close examination of the transitional epithelium from the proximal ureter revealing disordered reactive epithelium, acute inflammation, with neutrophil margination, and recent hemorrhage (HE staining,  $\times 400$ ).



**Figure 5** – Cross-section from the pelvic junction with loss of mucosa, abundant collagen bundles, mixed inflammatory infiltrate and recent hemorrhage in the wall (HE staining,  $\times 100$ ).



**Figure 6** – Longitudinal section from proximal ureter showing erosion of the urothelium, hemorrhage and fibrosis in the wall (HE staining,  $\times 100$ ).



**Figure 7** – On cross-section, we can find misplaced urothelium nests in the muscle wall of the ureter misleading a proliferation at this level (HE staining,  $\times 100$ ).

## Discussions

In the therapeutic indication of PUJ syndrome patients, we start from the idea that there is no medical treatment. However, we can watchful waiting these patients when to the obstruction is not symptomatic, there are no elements suggesting a deterioration of the kidney function without infection or associated lithiasis. We also need to keep in mind that in selected cases obstruction may regress as the baby grows [1].

The success rate of the different surgical methods of the PUJ obstruction depends on a number of factors including the degree of obstruction, whether primary or secondary obstruction is, the function of the incriminated kidney, anatomical considerations, such as junction compression vessels, infrastructure, intermediate and auxiliary personnel instruction and last but not least, the surgeon's experience.

An intervention without urinary drainage would be the best option given concerning difficulties we encounter in handling the urethral and ureteral catheters. These difficulties can be due to the size of the urethra that makes it unfeasible for instruments, or the lack of instruction and equipment required (C-arm).

That is why the qualification of the staff available in

the care of small patients is very important. There are patients with multiple venous, urethral, intrarenal catheters. All the maneuvers required for mounting a stent, removing a nephrostomy, extracting a JJ catheter, aspirating the catheters, not to mention the repeated mount of a leaked nephrostomy, should be done by the surgeon who has to be helped by trained personnel and an adequate infrastructure.

The association of ureteral reflux requires a certain strategy and raises the question of which of these conditions should be treated first. As a rule, ureterovesical reflux is of a small degree; therefore, the junction syndrome must be corrected first. When the reflux is high, things are the opposite. It is then necessary to distinguish between changes in the secondary junction to reflux by a true obstructive junction syndrome [10, 11].

For the surgery indication, in addition to the other normal limits, the urinalysis must be sterile. It is not always true that urography is sufficiently informative considering the functional immaturity of the kidney and/or intestinal gas overlapping. This is why we, in selected cases, required abdominal tomography even if it is an expensive radiological exploration that requires anesthesia assessment [12–14].

The vast majority already had the ultrasound exam and the urography performed. If renal ultrasound shows moderate or severe hydronephrosis without a dilated ureter, we should only think of an obstructive syndrome of PUJ.

For a successful intervention, we have followed a series of steps such as a broad approach that allows a good isolation and inspection of the kidney, sufficient urethral spatulation for a wide anastomosis, correct and symmetrical repair (without folds) of the renal pelvis were essential for a final funnel configuration that ensures good drainage. We have always checked if the ureter is open by injecting physiological serum into the ureter lumen. It is also very important to mark a ureter's margin with a guide suture in order to avoid anastomosis with a twisted ureter, but also the edges of the projected basinetal for the symmetrical excision of the walls. After the repair of the renal pelvis, we have always controlled the cavities knowing the fact of the association of obstruction with secondary lithiasis [15, 16]. It is disappointing to have radiotransparent calculi and not extract this.

Particular attention should be paid to each encountered blood vessel and its protection if possible. Under no circumstances will a compression vessel be cut on the front of the junction known to be important vessels for the lower renal artery in the renal artery or even the aorta. It will be done by cutting the ureter as we did in three cases. The cuts were made with 3/0 or 4/0, separate threads and a surge in a linear fashion. Load tissue should be equal at each step and the suture should not be in tension. We performed the AH operation and in the situation where the apparently intraoperative cause of the obstruction was only extrinsic (two cases of vascular compression, one case of fibrous bump compression). The Hellström operation of the crossing may be insufficient in certain situations.

Traditionally, in adults and adolescents, the election treatment after junction relapse is antegrade endopyelotomy, whereas the open endopyelotomy is common only after endoscopy was ineffective [17, 18].

The technique of solving the PUJ obstruction evolved much from the pioneering attempts of Trendelenburg & Foley. When Anderson-Hynes described the dismembered technique (1949 for the retroreflective ureter) it became the procedure of choice that can be done through open surgery, laparoscopic or robotic assisted [19]. The patient's position was in lateral decubitus, which allows the retroperitoneal lumbar approach of the kidney and ureter.

Intraoperatively after decompressing the ureter and dividing the ureter from the vessel, one can observe emptying or missing the renal pelvis. Lack of emptying is also the sign of intrinsic obstruction and proof of AH need operation. On the other hand, when the operation is limited to a decrossing, in order to avoid an anastomosis in tension, excision in the subconjunctive ureter (associated urethral atresia) must be well calculated for an anastomosis without tension.

A vascular compression (which hurries the operative indication) can be suspected after urographic clustering (a procubitus X-ray, the contrast substance being heavier than the urine will better emphasize the junction) and confirmed by Doppler ultrasound. Endoluminal ultra-

sound can more accurately identify compression vessels at the junction [20].

It should be said that the presence of a lower polar vessel as well as a sub-junctional ureteral stiffness of more than 2 cm is a contraindication to the percutaneous (endopyelotomy) approach of the PUJ obstruction, the minimally invasive procedure we do in adults. In complex kidney abnormalities (kidney in the horseshoe, etc.), a very good knowledge of the vasculature is needed. In such situations, the three-dimensional reconstruction of computed tomography (CT) or magnetic resonance imaging (MRI) can be used. The non-dismembered Scardino procedure was performed in a case of sub-junctional ureteral atresia associated with junctional obstruction. Double kidney systems, ectopic kidneys and other rotating abnormalities can also be the cause of PUJ obstruction [21, 22].

Pyeloureteral anastomosis may or may not be protected by a urinary drainage system (JJ, nephrostomy, pyelostomy), depending on the surgeon's experience and preferences, but it also depends on the technical possibilities he has. It is known that the original technique did not predict urethral stenosis. Subsequently, over the decades, there has been a dispute of stenting or not [23].

## Conclusions

Even though clinical and imaging studies are sufficient for diagnose of PUJ syndromes, morphology and histology bring essential data regarding the age of the lesions. AH dismembered operation is the princeps surgery to solve the obstructive syndromes of the PUJ. It can be performed by open, laparoscopic but also robotic-assisted surgery. The urologist finds his place in the treatment of this condition provided he/she has received additional training in a specialized service, the organization of a specific infrastructure and the use of qualified staff.

## Conflict of interests

The authors declare that they have no conflict of interests.

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