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DEPARTMENT OF MEDECINE

THEME:

MEGAURETER IN CHILDREN :

A RETROSPECTIVE STUDY

OF 11 CASES FROM THE MOTHER AND CHILD EHS OF TLEMCEN

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INTRODUCTION

INTRODUCTION:

The practice and science of pediatric urology have changed rapidly in the last few years. New surgical techniques, better and more informative prenatal evaluations, and better biomedical substances continue to arrive on the scene.

The term megaureter does not define a specific pathological condition, because it can be due to different underlying abnormalities.

The most used classification includes three groups: refluxing megaureter, associated with vesicoureteral reflux (VUR); obstructive megaureter, associated with urine flow impairment at the vesicoureteral junction; non-refluxing non-obstructive megaureter if neither obstruction nor reflux can be identified.

Each group can be divided into two subgroups: primary megaureter; secondary megaureter.

As the use of fetal ultrasonography has expanded, the majority of children with megaureters are now diagnosed early in their development and an increased number of cases are identified before the onset of symptoms.

Physicians are faced with the complex task of distinguishing which children need medical intervention and which do not.

Nowadays non-operative management is preferred (a "wait and see" approach).

The surgical treatments of megaureters are well established, relatively simple, and effective if performed on the correct candidates.

Therefore, research efforts in this field have recently focused on improving our ability to diagnose clinically relevant obstructive uropathy and examining the developmental causes of megaureter, and how this disorder may be prevented.

The pediatric surgeon together with the pediatric radiologist and nephrologist should pose the indication for surgery. Only in these cases: significant impairment to urine flow; worsening renal function during the observation time; recurrent UTI despite adequate antibiotic prophylaxis.

PATIENTS AND METHODS

PATIENTS AND METHODS:

I. Aim of the Study:

The main purpose of our work is to report the experience of the Pediatric Surgery service, at the mother and child EHS of Tlemcen in the diagnosis and management of megaureters.

II. Type of study:

It's a retrospective descriptive study, involving a series of 11 ***children*** and 11 cases of megaurter(MU). These series are collected at the Pediatric Surgery service at the mother and child EHS of Tlemcen over 20 months, from January 2021 to July 2022.

III. Selection Criteria:

1. Inclusion criteria:

They are included in our study, any hospitalized child under 15 years old with a megaureters, taken care of at the pediatric surgery service during the period between January 2021 to July 2022.

2. Exclusion criteria:

We excluded from this study:

- All patients with ureteropelvic junction (UPJ) obstruction, bladder or urethral malformation.
- Lost-to-follow-up patients.
- those whose file was inexploitable.

IV. Data collection :

A data sheet (Appendix) produced for this purpose made it possible to collect the various epidemiological, clinical, para-clinical, therapeutic and evolutionary data from the patient files.

V. Statistical analysis:

Data entry was carried out using Word and Excel. In order to compare our results with those of the literature, we have carried out a bibliographic search, the analysis of theses and the study of pediatric urology books on MU.

VI. Ethics:

Retrospective file analysis does not require patient consent and this type of work does not require formal submission to an ethics committee.

However, in order to respect medical confidentiality, anonymity was maintained in the exploitation sheets.

VII. Evaluation criteria :

The patients in our series had an evaluation of the clinical state and the biological renal function.

A radiological A review including:

- An ultrasound of the urinary tract.
- A renal scintigraphy with DTPA or DMSA.
- A retrograde cystography.
- Intravenous urography.

RESULTS AND ANALYSIS

I. Epidemiology :

1) Gender:

A clear male predominance was noted in our series with 8 cases, for 3 girls. That's a sex ratio of 2.66.

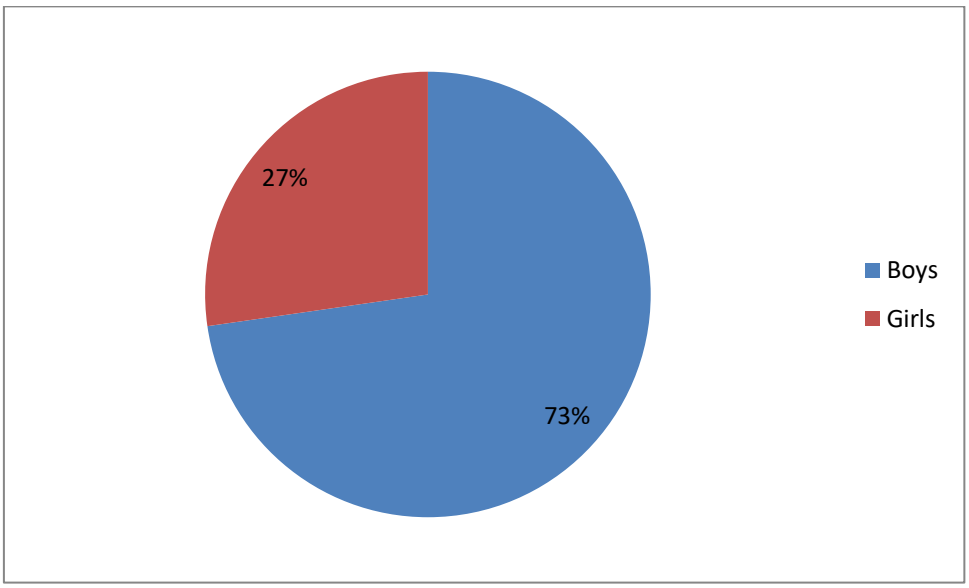

Figure 1: Gender distribution.

2) Age :

The age of our patients at the time of hospitalization ranged from 18 months to 11 years, with an average of 5 and ½ years.

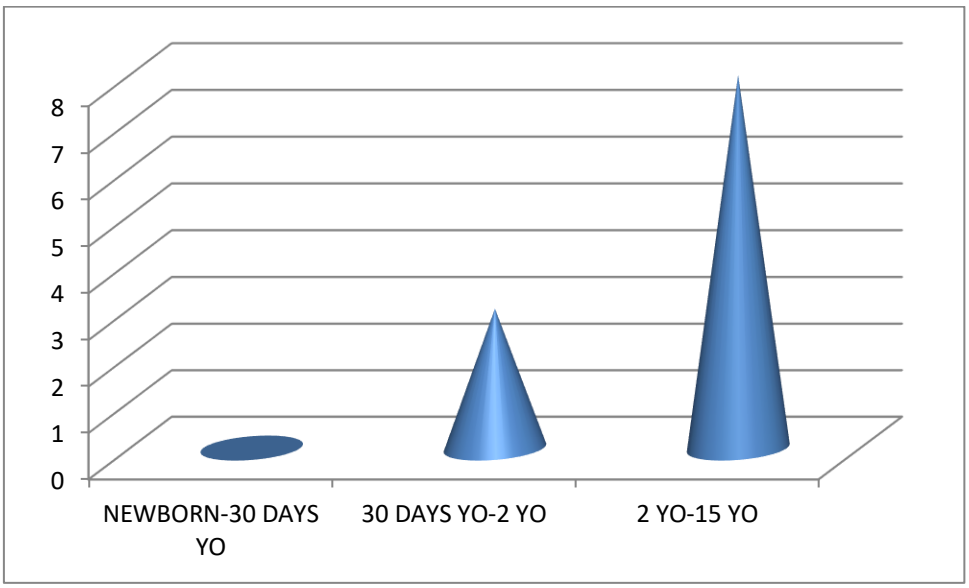

Figure 2:Age distribution.

3) Antecedent:

First degree inbreeding was not found in our study, however, no family cases were noted in our study.

4) Discovery Circumstance:

The reasons for consultation leading to diagnosis varied for the patients in our study as described in Table 1.

Table 1: Discovery Circumstance.

RFC	Antenatal	Lower back pain	Urinary infection	Acute pyelonephritis	Fever	Renal insufficiency	Abdominal pain	Hematuria	Urinary disorders
N°	3	5	6	0	7	0	4	2	3
%	27,27%	45,45%	54,55%	0%	63,64%	0%	36,36%	18,18%	27,27%

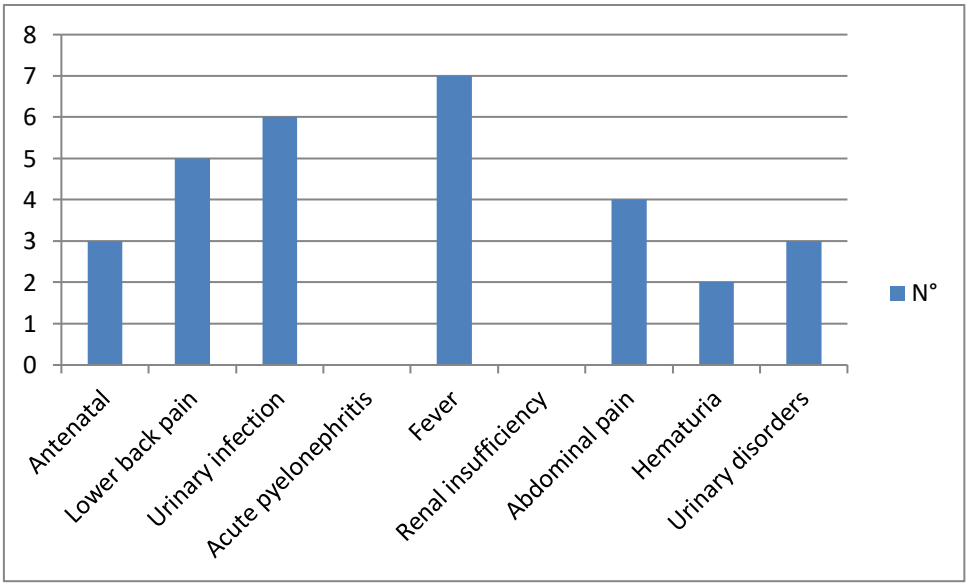

Figure 3: Breakdown of patient consultation reasons..

II. Clinical study :

The clinical examination in our patients was poor and there was no palpation of abdominal mass or abnormal EGO.

However, we've noted the presence of a lumbar contact in 2 cases.

III. Paraclinical Study:

1) Biological exams:

a) Cytobacteriologic examination of the urine:

We only found 2 cases of urinary tract infection in our series, one in **E. Coli** and other in **Klebsiella**; both patients received parenteral antibiotic therapy during hospitalization with good progress (sterilization of urine at the end of treatment).

Germ-free leukocytosis was found in one patient, who was not treated with antibiotics due to the absence of accompanying clinical signs.

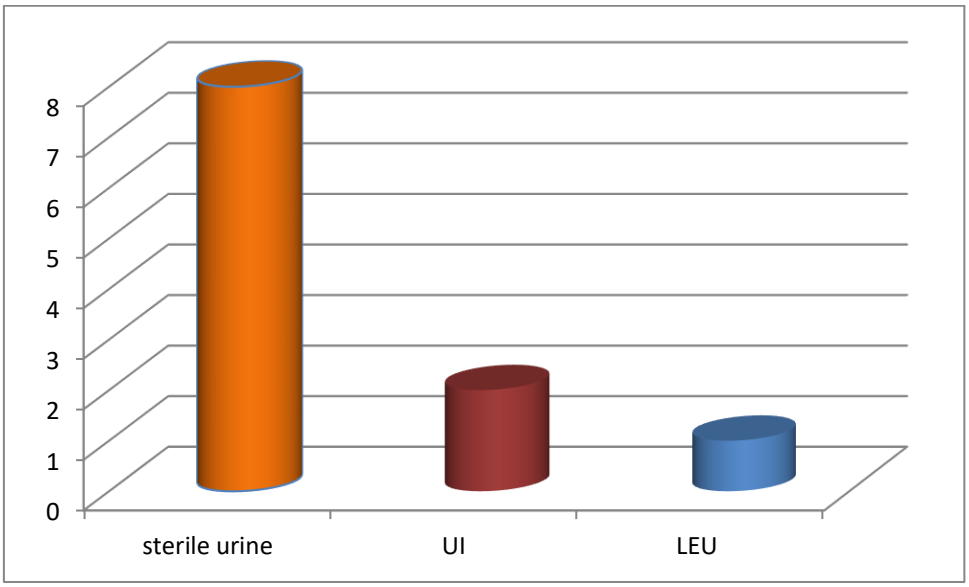

Figure 4:CBEU results.

b) Renal function:

It was found that only one patient who presents kidney failure with glomerular filtration rate at 35,26 mL/min/1.73m (SERUM CREATININE: 60,11 umol/l).

2) Radiological exams:

a) Pelvic abdominal ultrasound:

Ultrasound of the urinary tract is a fundamental examination in the diagnosis of MU by objectifying ureterohydronephrosis, its laterality, the quality of the renal parenchyma as well as the cortical index and the demonstration of an associated anomaly.

In our study the right side was more affected than the left side and we also noted a high frequency of cases of bilaterality as described in the following diagram;

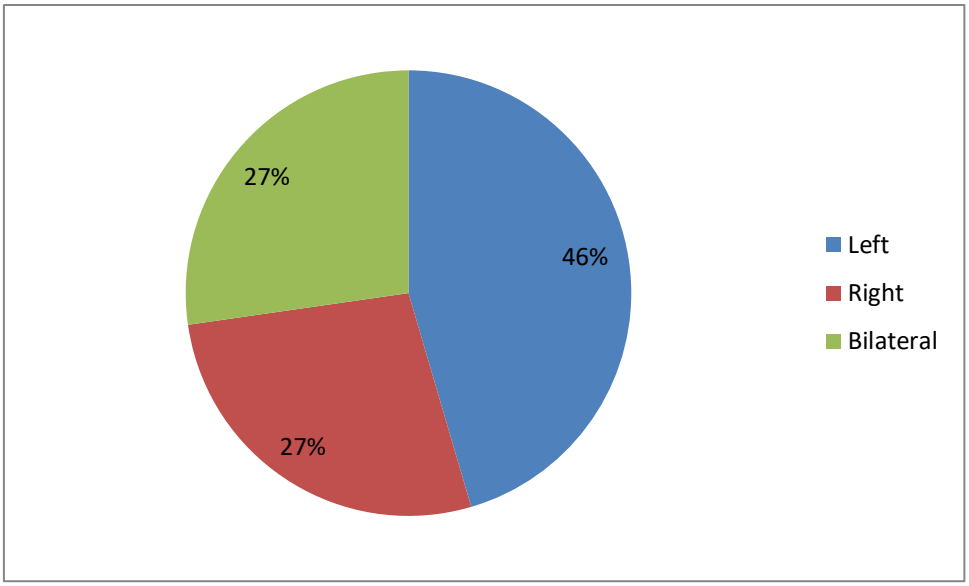

Figure 5: Affected side according to ultrasound.

The cortical index was reduced in 2 cases, i.e. 18,18% of cases with a lower extreme of up to 3mm.

The ureter diameter varied between 7 mm and 28 cm.

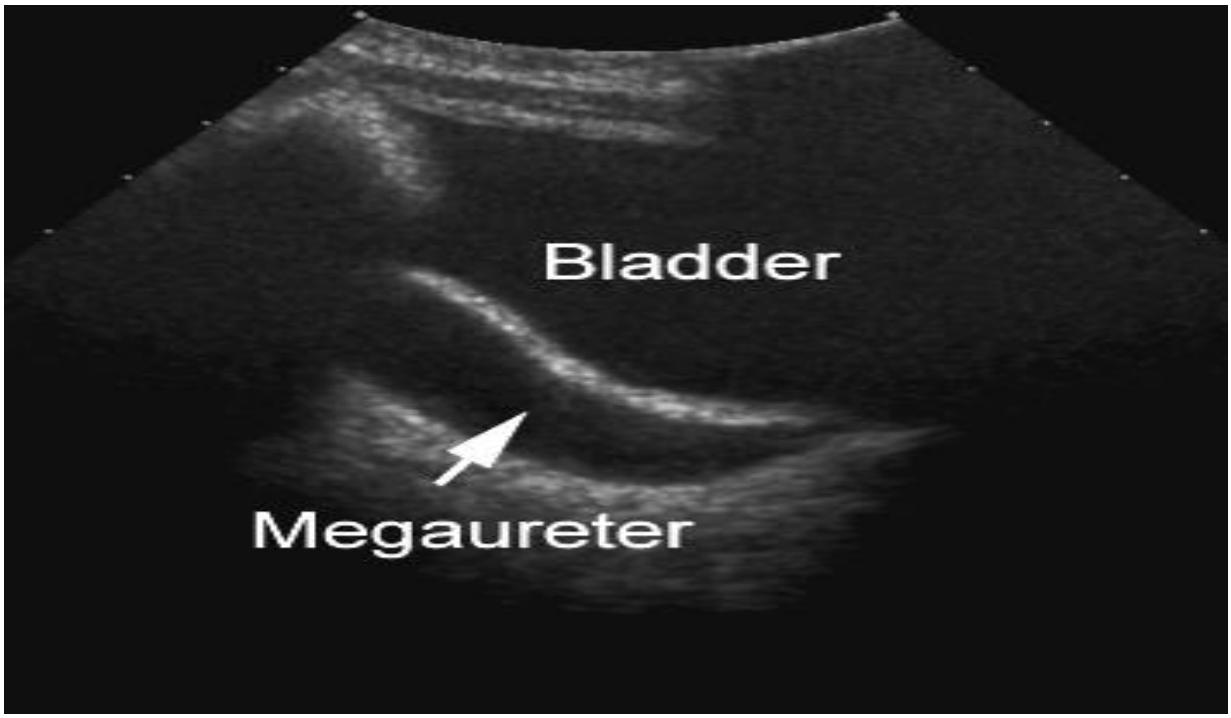

Figure 6: Ultrasound from obstructed megaureter demonstrating dilated collecting system and proximal ureter .

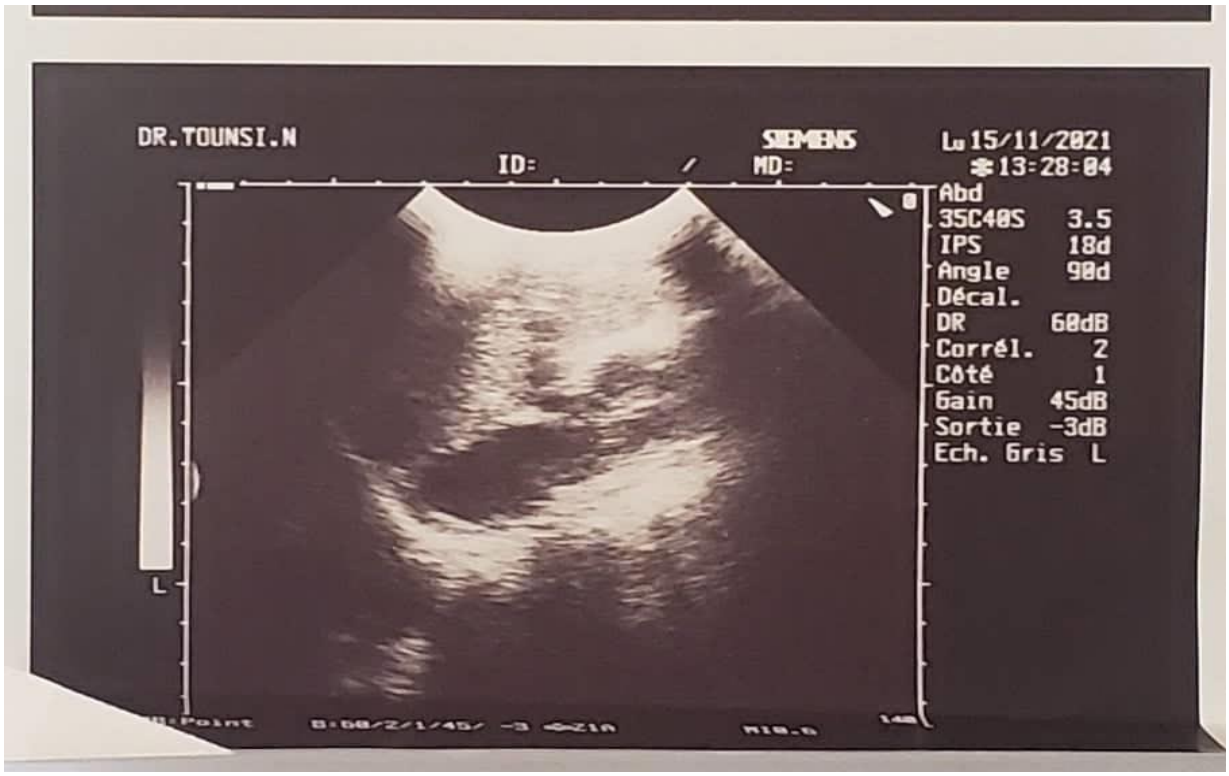

Figure 7: Ultrasound showing ureteral dilation in a patient in our series.

b) Voiding Cystourethrogram (VCUG):

It makes it possible to specify the refluxing character or not of the MU. In our series it was performed in only 1 patient.

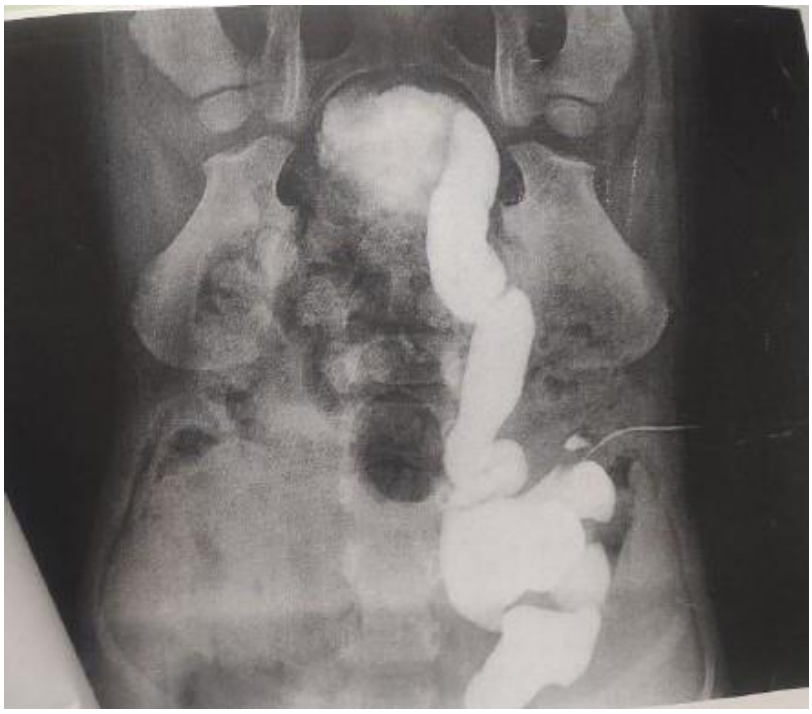

Figure 8: VCUG showing a grade V reflux in a patient in our series

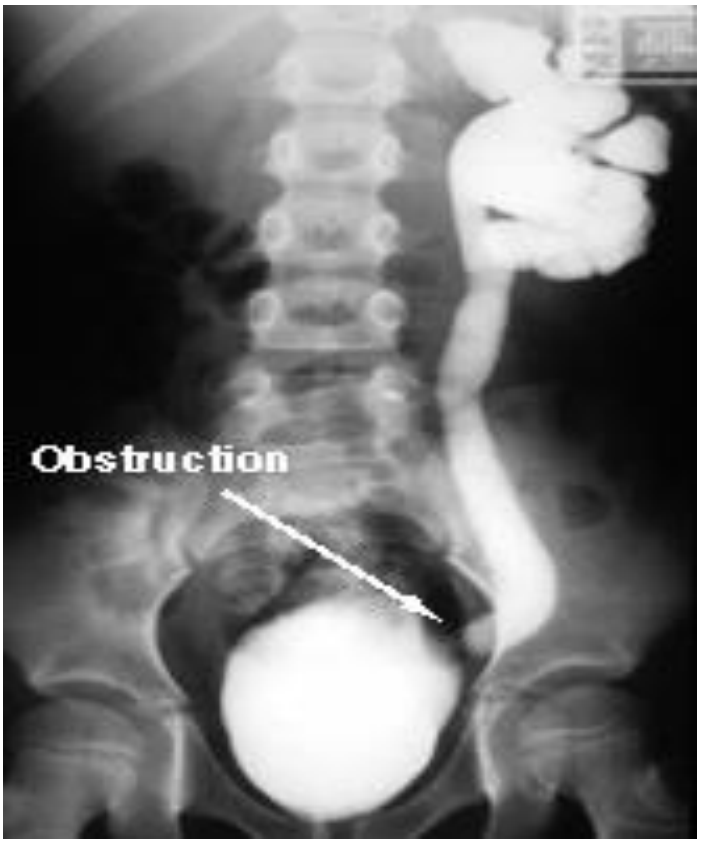

Figure 9: VCUG with a left reflux.

c) Intravenous urography:

It was performed in only 1 patient. We were able to visualize an obstacle upstream of the bladder"radicelle".

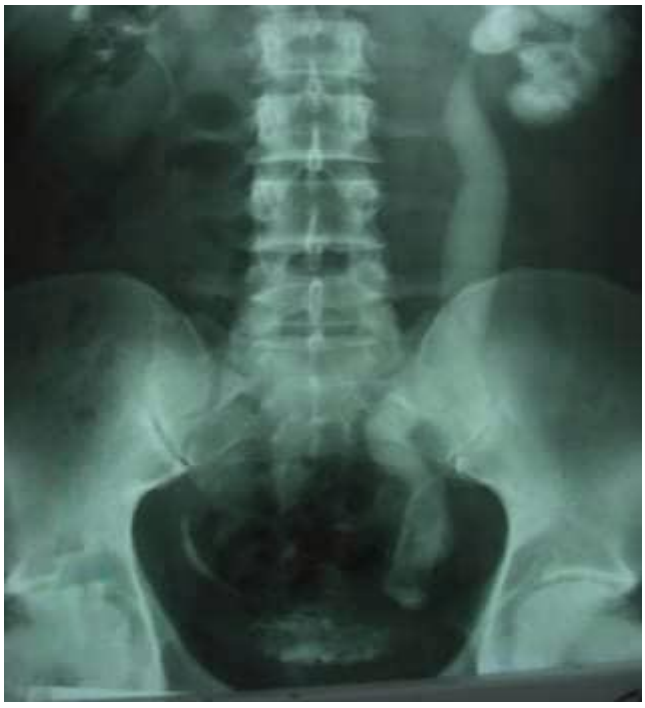

Figure 10:IVU demonstrating left megaureter with ipsilateral hydronephrotic kidney.

d) Renal scintigraphy:

The renal scintigraphies performed were with DMSA or DTPA. They were performed in 6 patients, 2 of whom had bilateral involvement.

There were 6 cases of drainage alteration including 2 on the right and 4 on the left and only 2 cases had no drainage alteration.

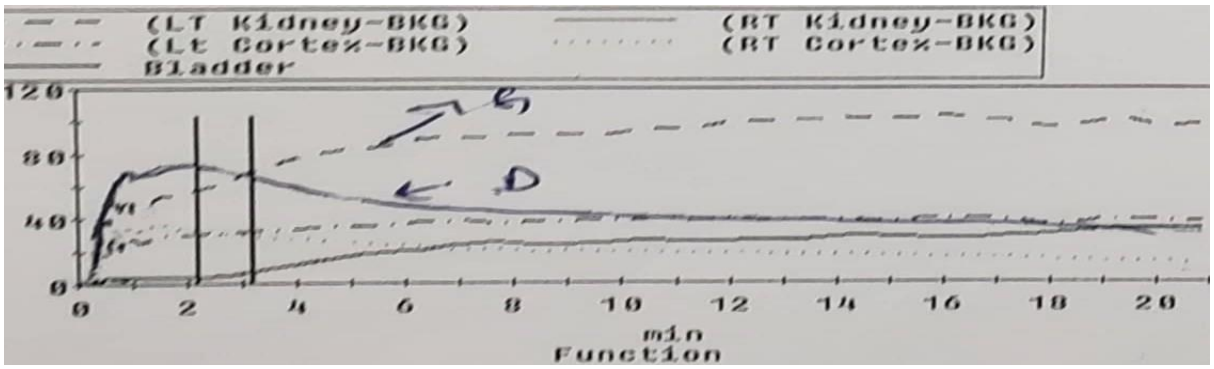

Figure 11:DMSA scintigraphy objectifying a non-functional right kidney in a patient in our series.

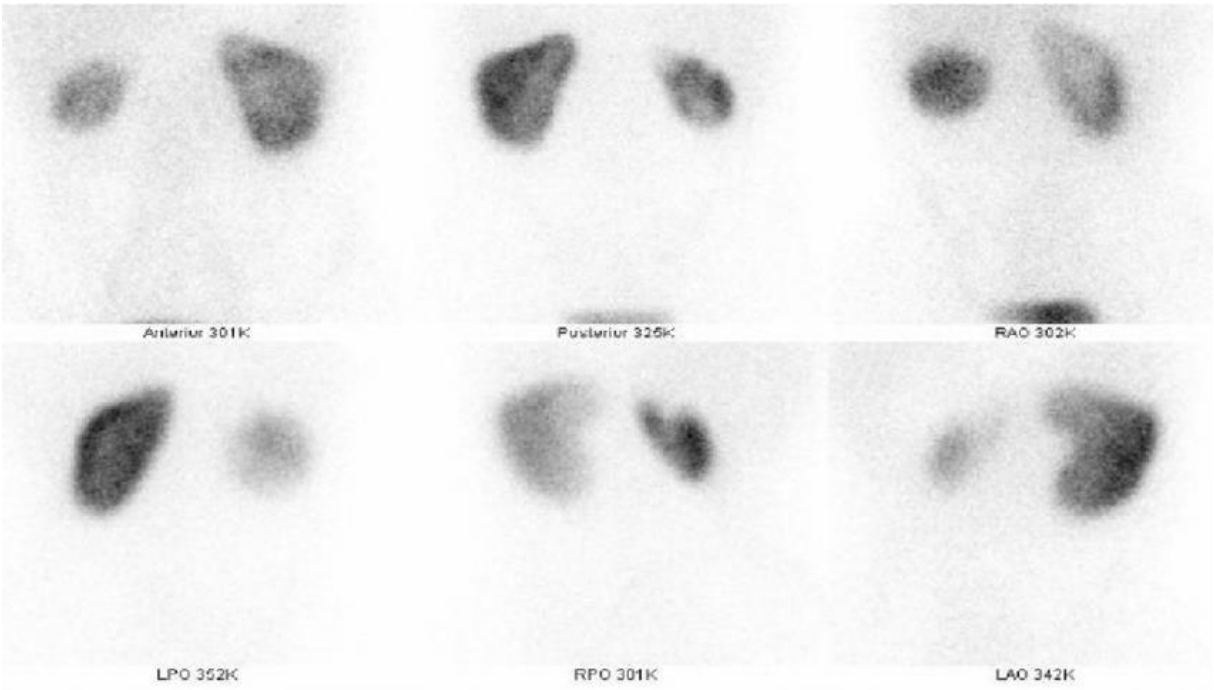

Figure 12 DTPA scintigraphy showing a:split renal function of 69% on the left and 31% on the right kidne in a patient in our series.

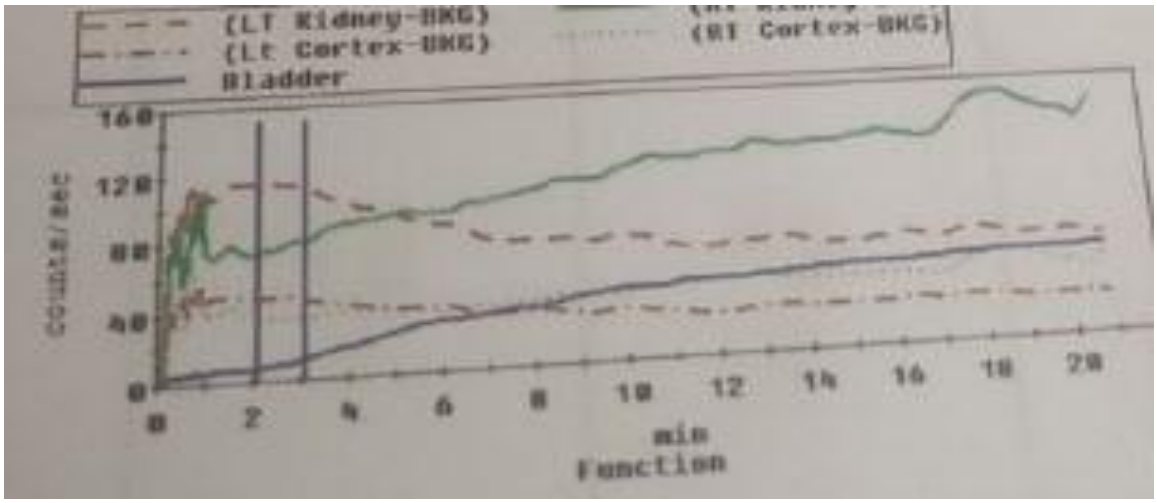

Figure 13: DMSA scintigraphy showing a non-functional left kidney with compensation of the right kidney in a patient in our series.

e) CT urology scan:

Although the CT-scan has less and less indication in the context of malformative uropathies, we have performed it in two patients, one of them has an estimated renal function at 2% in scintigraphy.

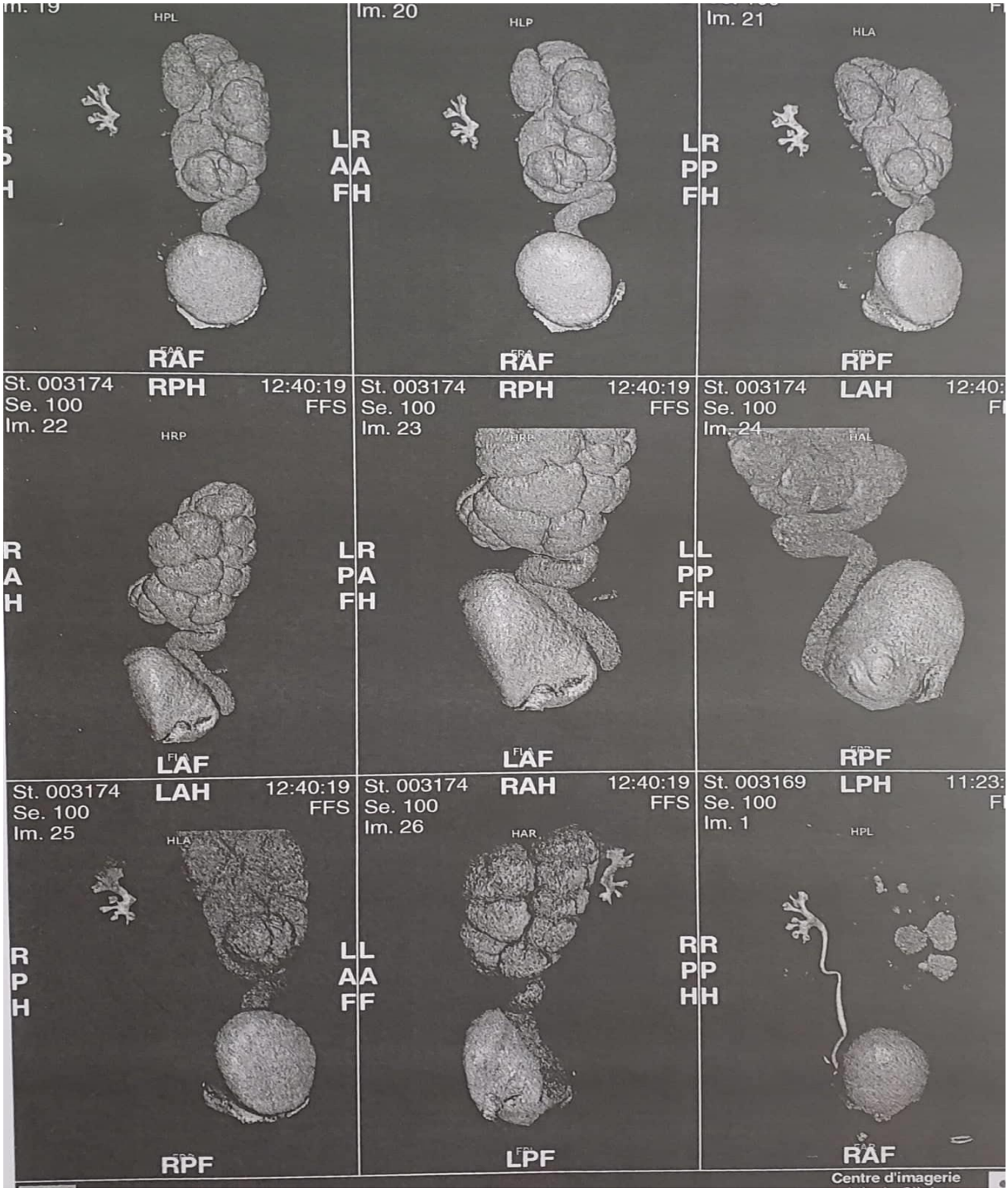

Figure 14a:CT scan 3D reconstruction showing a right hydronephrosis with a major pyelocaliciel dilation with MU in a patient in our series.

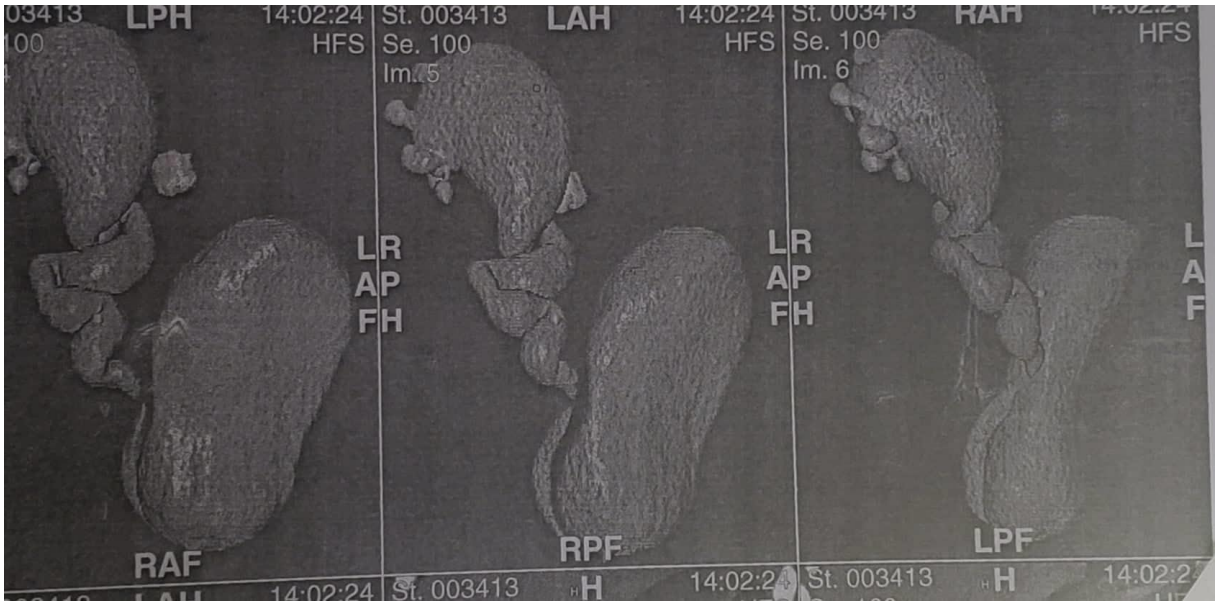

Figure 15b:CT scan 3D reconstruction showing a right hydronephrosis with a major pyelocaliciel dilation with MU in a patient in our series.

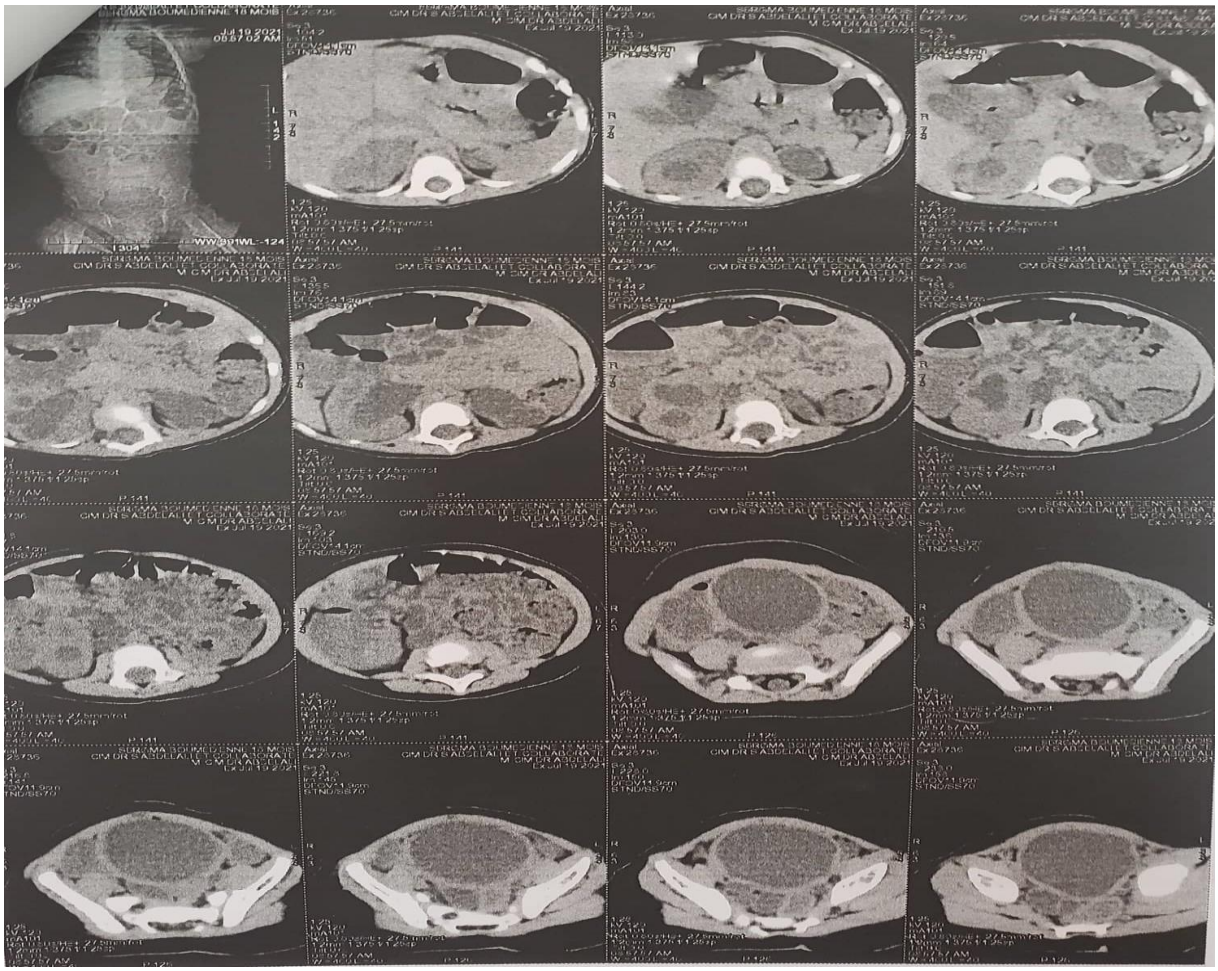

Figure 16a: CT scan showing a major left hydronephrosis.

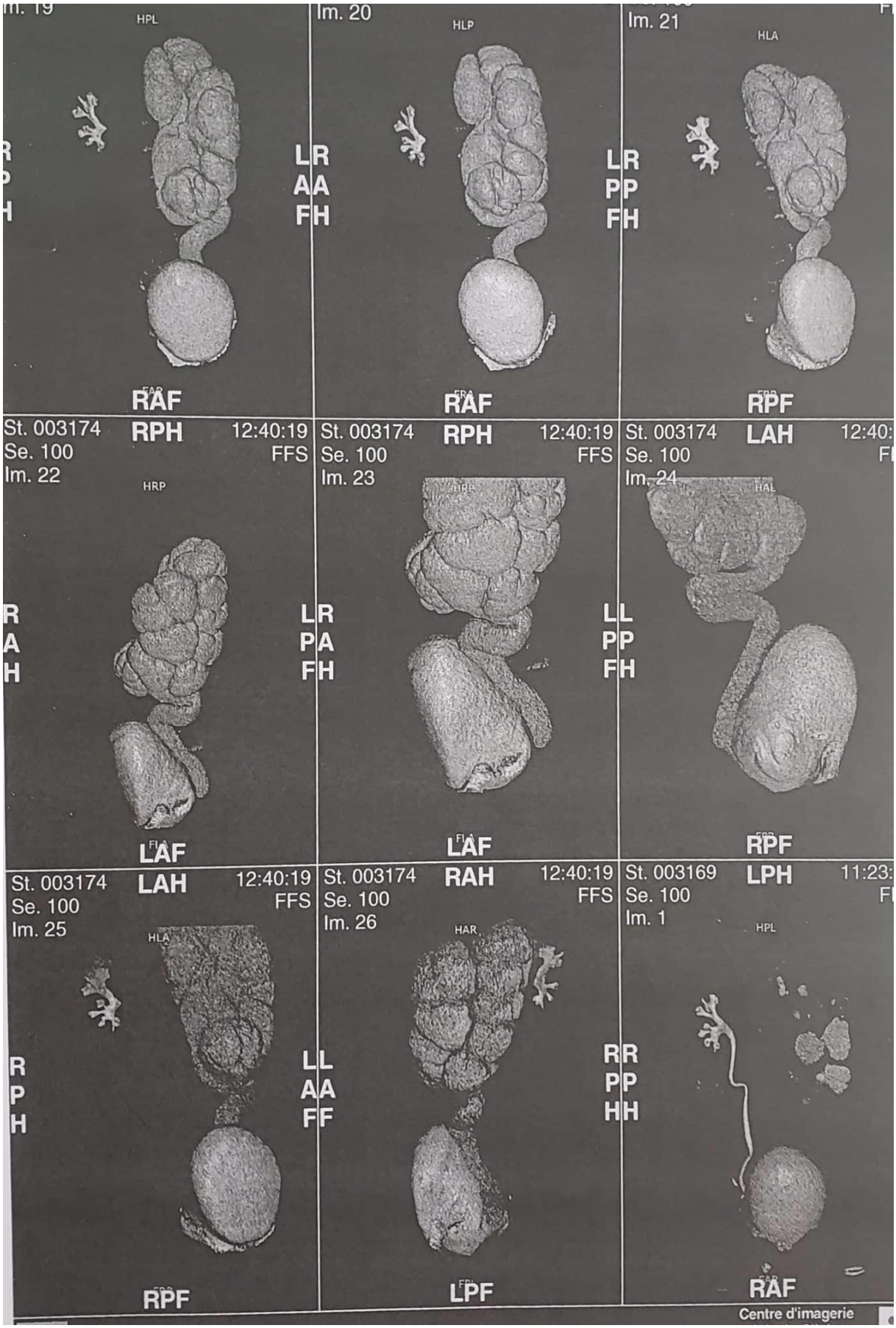

Figure 17b: a 3D reconstruction showing a mojor left hydronephrosis with a MU.

3) Final diagnosis and classification:

At the end, we were able to classify our patients into 4 groups according to **King**'s classification. They are distributed as follows:

Groupe 01: OBSTRUCTIVE MEGAURETER; 4 cases i.e. 36,36%

Groupe 02: REFLUXING MEGAURETER; 6 cases i.e. 54,54%

Groupe 03 : REFLUXING OBSTRUCTED MEGAURETER ; 1 case i.e. 09,09%

Groupe 04 : NONOBSTRUCTIVE NONREFLUXING MEGAURETER ; 0 case.

IV. Treatment and results:

Due to lack of data, a lot of files were without an operational protocol, which was difficult for us to fully grasp the care. Despite everything, we managed to understand the approach followed.

- No medical treatment is mentioned in the records we have.
- out of 11 patients ; 8 patients benefited from a surgical intervention that consists of:
  - Reimplantation of the ureters using the COHEN technique; 4 cases i.e. 36.36% of cases.
  - Modelling intervention using Kalicinski and Starr's technique; 2 cases i.e. 18.18% of cases.
  - Nephrectomy was observed in 2 patients cases i.e. 18.18% of cases.

The ureteral probes were kept in our patients for 10 to 15 days, All patients were treated with antibiotics (C3G) postoperatively and for the duration that drainage probes are in place, and then relayed with a urinary antiseptic.

Unfortunately, the follow-up of these patients is not mentioned in the files. Although, we were able to ask the residents about the rhythm of survival and the examinations to be requested.

The follow-up of operated patients included monitoring by biological renal function and ultrasound of the urinary tract at each consultation. The monitoring rhythm was a control at 1 postoperative month then at 3 months, 6 months then annually.

In our series, the evolution after surgical treatment was favorable for all our patients, who did not show any IU during the whole period of surveillance and with normal renal function.

To conclude The evolution was favorable for all the patients who had undergone surgical treatment. The complication rate with surgical treatment is nil in our series.

DISCUSSION

DISCUSSION

I. DEFINITION:

1. What is a MEGAURETER?

Much of the debate surrounding the diagnosis and treatment of "megaureter" stems from the confusion in the nomenclature, as the term could be applied to any dilated or big (mega) ureter. Although the term megaureter means a single specific disease to some, it carries a mere connotation to others. Many classifications were proposed for megaureter, but clinicians should use the system that seems most simple and useful.

The term "megaureter" is simply a descriptive term about the diameter of the ureteral lumen. It means exactly what it says, "large" ureter. It implies no description of function of the ureter in terms of obstruction or reflux. A megaureter on imaging studies can be quite impressive, and of concern to physicians and parents alike. And, since the surgical techniques to repair megaureters are relatively simple and successful, it is tempting to try to improve surgically abnormal appearing ureters in the presence of marked dilation; however, the data have shown us that surgical intervention even in the setting of some pathology, such as obstruction, is not always warranted. Indeed, the major challenge in the management of megaureters is the clinical decision to intervene or not (1–3).

A ureter is considered a megaureter if the lumen is dilated. Although the dilation is rarely subtle, for the terms of definition any diameter larger than 07 mm is considered abnormal. Megaureters can be classified as obstructed, refluxing, obstructed and refluxing, or neither obstructing nor refluxing, using the international classification of Smith et Al(4). Typically, if there is an obvious cause for the enlarged ureter, such as elevated bladder pressure due to a neuropathy, the term megaureter is not used. Rather, it is reserved for cases of hydroureteronephrosis and otherwise normal bladder and outlet function. The pathology of the ureter can also be primary (or intrinsic) or secondary (due to another urinary tract pathology). It is important to distinguish between cases of primary or secondary causes of the pathology, as in the case of secondary causes, the treatment is directed at the initiating pathology and not at the ureter (1–5).

The question of obstruction is a difficult one in cases of megaureter, much as in the children with ureteropelvic junction (UPJ) obstruction. Indeed, any megaureter labeled "obstructed" has, by definition, a delay in drainage of radiotracer on nuclear medicine renogram beyond the normal t½ of 20 min commonly used as a cutoff for urinary tract blockage. However, as Koff and Campbell described years ago, the true definition of obstruction lies not in the t½, but in the determination of the degree of obstruction that will lead to renal injury if it is not relieved (6).

2. Incidence:

MU is the second common cause of obstructive uropathy, represents 23% after JPU 44% in a study that concerned 185 born having obstructive uropathy in 2000 by SHOKIER et al(1).

The diagnosis is more common in boys than girls, and more often is on the left side. It can be bilateral in 25% of cases, and the contralateral kidney is absent or dysplastic in 10–15% of cases.

There is no clear genetic pattern of inheritance, although some cases do appear to run in families. Rarely, a UPJ obstruction will be present in conjunction with a ureterovesical junction (UVJ) obstruction in cases of megaureter (1–3).

Most cases of megaureter are now first detected with prenatal ultrasound and then diagnosed after birth. Some cases present clinically during childhood with abdominal pain, hematuria, and/or urinary tract infections. Megaureter may also be incidentally discovered later in life on imaging studies. It rarely leads to renal insufficiency (1–3).

3. Types of MEGAURETER :

A megaureter means “big ureter” and it is a descriptive term, not a diagnosis. The two important questions about megaureter are whether there is reflux (backwash) of urine causing the megaureter or whether there is blockage at the ureterovesical junction causing megaureter (Figure 1). If there is reflux, the diagnosis is “refluxing megaureter” or “megaureter from reflux”. If there is obstruction, the diagnosis is “obstructed megaureter” or “primary obstructed megaureter”. If there is neither reflux nor obstruction, then the diagnosis is “primary non-obstructed megaureter”. In very rare circumstances, there can be reflux and obstruction and the diagnosis is “refluxing obstructed megaureter”. The two most common types of megaureter by far are the “primary non-obstructed megaureter” and the “refluxing megaureter”.

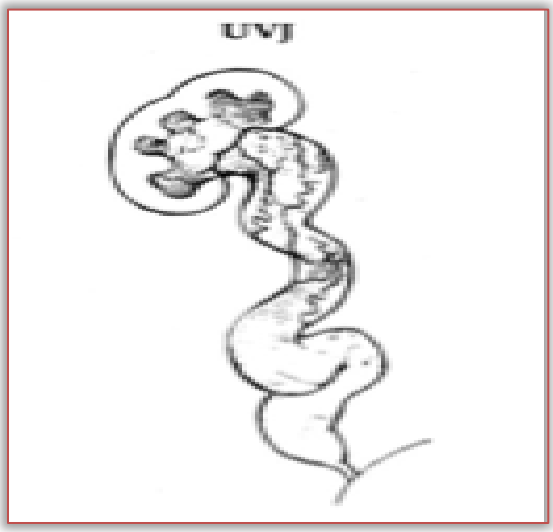

Figure 18 :Image of a ureterovesical junction obstruction.

a) Primary Obstructive Megaureter:

Primary obstructive megaureter is considered a functional obstruction. There is thought to be an aperistaltic juxtavesical (adynamic) segment in the ureter, leading to a lack of propagation of the ureteral peristalsis and therefore urine flow. This distal segment has been examined histologically and has been found to contain increased levels of collagen type I and III (predominantly type I). It is this increased fibrosis that is implicated in the disruption of intercellular communications and leads to uretero arrhythmias and obstruction (7–10).

There are many other theories regarding the development of obstructive megaureters, however. Some scientists have shown evidence of atrophy of the inner longitudinal muscles in these ureteral segments (the longitudinal muscles are the ones that transmit peristalsis) and hypertrophy of outer, compressive circular muscle, leading to obstruction (11,12).

The fact that so many obstructive megaureters resolve and develop into normal collecting systems over time has pushed many to define a maturational cause of obstructive megaureters, signifying that perhaps the renal urine production began slightly prematurely, before the ureter fully cannulated at its caudal end, leading to hydroureter. The full canalization of the mature ureter could then explain the resolution of the obstructive appearance of the ureter. Another maturational theory is that the obstruction represents a developmental evolution of the distal ureter from a single, circular muscle layer to the double layer (circular and longitudinal) of the child (1,3).

Other histologic findings claiming to display the causative aspect of the obstructive megaureter include distal ureteral segments with no muscle tissue present, but simply a fibrotic, static terminal end.

Yet others have documented distal ureteral segments with a nonureteral, nondetrusor muscle that is excessively responsive to nonadrenergic stimulus, leading to almost tonic contraction (1,13–15).

Interestingly, the proximal, dilated ureteral segment has also been found to be composed of altered connective tissue, and this fibrosis and the dilation itself can lead to uretero arrhythmias and poor peristaltic wave transmission. It is important to note that the upper tract dilation (while appearing to be asignificant pathology in and of itself) does play an important role in the urinary tract response to the presence of obstruction. The infant collecting system is more pliable than in more mature patients and this dilation allows for the dampening of pressure, allowing the kidneys to produce urine into a collecting system at close to physiologic pressures (1–3).

Other than the adynamic segment described above at the terminal ureter in the obstructive megaureter, other anatomic causes can lead to a similar clinical scenario. Both congenital distal ureteral strictures and distal ureteral valves can be almost indistinguishable from the classic obstructive megaureter (16–18).

b) Secondary Obstructive Megaureter :

Secondary obstructive megaureter represents an obstructive process secondary to elevated intravesical pressure of some other cause. Common causes include spinal dysraphism and neurogenic bladder, which may elevate detrusor pressure to over 40 cm H_2O, causing a physiologic obstruction and hydronephrosis in the collecting system. Non-neurogenic voiding dysfunction, if severe enough to elevate bladder pressure above the safe range, may also be a cause. Posterior urethral valves, or other causes of infravesical obstruction, can also lead to similar findings (1,17,18).

Other anatomic causes of secondary, distal ureteral obstruction include ureterocele, ectopic ureter, bladder diverticula, periureteral fibrosis, and external compression by retroperitoneal tumor, masses, or aberrant vessels (1,17,18).

c) Primary and Secondary Refluxing Megaureter :

Refluxing megaureters simply represent a refluxing ureter that happens to be dilated. The pathology mimics that of any refluxing ureter, with a short intravesical ureter and submucosal tunnel. They may be associated with abnormalities of the UVJ, making reflux more likely, such as periureteral diverticula.

Some children present with megacystis megaureter syndrome, in which the bladder is markedly distended and thin walled, in addition to the ureters (17).

The distal segment of refluxing megaureters also shows histologic derangement with increased fibrosis (much like the obstructive megaureters); however, in these cases, the predominant collagen is collagen type III (10).

Refluxing megaureters can be a characteristic of Prune Belly syndrome as well. These ureters also often demonstrate increased collagen deposition distally, with the clinical manifestation ranging from inefficient peristalsis to distal obstruction (18).

d) Refluxing Obstructed Megaureter :

In refluxing megaureters, 2% also present with some degree of obstruction.

Although not intuitively apparent, refluxing ureters may lead to obstruction when the distal ureter that fails to coapt (reflux) also does not transmit peristalsis (obstruction). Alternatively, the ureter may have an ectopic insertion at the bladder neck, which refluxes when relaxed and obstructs when tightened (19).

e) Primary Nonobstructive, Nonrefluxing Megaureter ;

Most cases of megaureter end up being of the nonobstructive, nonrefluxing variety. This is very heartening, as it confirms that simple observation will serve as the therapy for most children. However, as mentioned, the lack of obstruction can be difficult to prove (1,17).

Certain important points should be kept in mind when evaluating a megaureter that may help to prevent unnecessary intervention. First of all, the fact that an infant is born with a functioning kidney provides evidence that any degree of ureteral obstruction is not complete, as the kidney would not have formed normally in the setting of early or very high-grade obstruction. Also, as touched on earlier, the complex orchestration of embryologic development may have many variations that create an appearance of anomalous development, only to improve as the necessary steps of development are completed. The fetus makes larger volumes of urine compared to the infant, and if this diuresis precedes the natural canalization of the distal ureter, a megaureter may develop (maturational delay hypothesis). Since the ureter in the fetus is so compliant, small increases in urine flow can induce a ureteral dilation, even in the absence of obstruction and reflux. It is this compliant collecting system that allows the infant kidney to continue to function in the setting of varying degrees of obstruction or reflux without suffering pressure injury, so dilation may be beneficial and not harm the child, and is therefore not necessarily an indication for repair(1–3,17).

f) Secondary Nonobstructive, Nonrefluxing Megaureter :

The cases of nonobstructive and nonrefluxing megaureter due to a cause unrelated to ureteral anatomy are termed secondary. It is in this category that dilation due to high fetal urine output, increased compliance of fetal ureter (due to extracellular matrix composition, including elevated collagen type II, and elastin

concentration), or a partial or transient obstruction during development (such as ureteral folds or delays in the development of normal peristalsis) occur (17).

There are many other relatively benign causes of secondary megaureter. For example, urinary tract infections can lead to temporary ureteral dilation due to the presence of bacterial endotoxins that can inhibit peristalsis. As mentioned, any increase in urine output can cause dilation of the fetal/infant collecting system. Some possible causes of diuresis include lithium toxicity, diabetes insipidus or mellitus, sickle cell nephropathy, or psychogenic polydipsia (17).

II. ANATOMOPHYSIOLOGICAL REMINDERS :

1. Anatomical recall: (20)

The ureters are bilateral, muscular, tubular structures, responsible for taking urine from one kidney to the urinary bladder, thanks to its peristaltic activity for storage, prior to excretion. After blood has been filtered in the kidneys, the filtrate undergoes a series of reabsorptions and exudation throughout the length of the convoluted tubules. The resulting liquid then passes to the collecting tubules, after which it enters the collecting duct. From the collecting ducts, the urine passes from the calyces to the renal pelvis, which marks the beginning of the ureters. The arterial supply to the ureters comes directly and indirectly from the abdominal aorta. There are no ganglia on the ureters; however, it receives both sympathetic and parasympathetic innervation.

a) **Location**: retroperitoneal location, it occupies successively the lumbar and pelvic regions
b) **Shape and size**:

Shape: cylindrical

Length: 25-30cm.

Average diameter: 4mm.

c) the ureter is described as having **three** anatomic sites of narrowing:

-The ureteropelvic junction (UPJ), the ureteral crossing of the iliac vessels, and the ureterovesical junction (UVJ).

d) It is described in 4 portions:

- Lumbar portion : lumbar ureter
- Iliac portion: iliac ureter
- Pelvic portion: pelvic ureter
- Intramural or intravesical portion: within the bladder wall.

e) Course:

The ureters leave the kidneys posterior to the renal vessels. Both ureters pass inferiorly over the abdominal surface of the psoas major, with the genitofemoral nerve behind it and the vessels of the gonads in front. As the right ureter travels towards the bladder, it travels posterior to the duodenum and further down it is crossed by branches of the superior mesenteric vessels.

The left ureter, however, travels laterally to the inferior mesenteric vessels and is subsequently crossed by its branches. Eventually, the vessels leave the psoas major as the common iliac arteries bifurcate to enter the true pelvis. The ureter pierces through the wall of the urinary bladder from lateral to medial and posterior to anterior. So, this entrance is oblique. It forms the orifice of the ureter in the urinary bladder at the ureterovesicular junction.

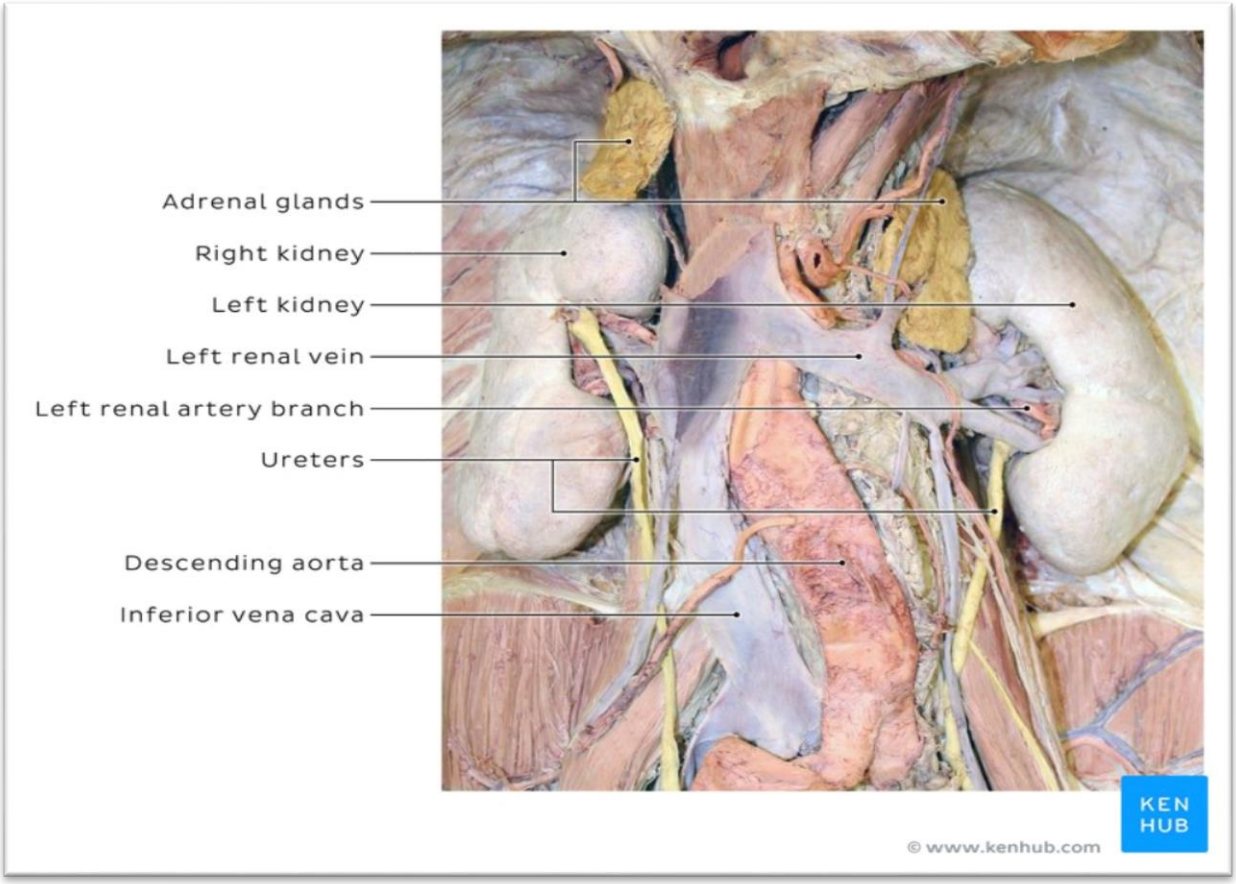

Figure 19: 2 Kidneys and ureters in cadavers: Ureters are continuations of the renal pelvis, which is located posterior to the renal artery and renal vein (acronym 'AVP'). The ureters can be confused with the inferior mesenteric artery. The distinguishing feature .

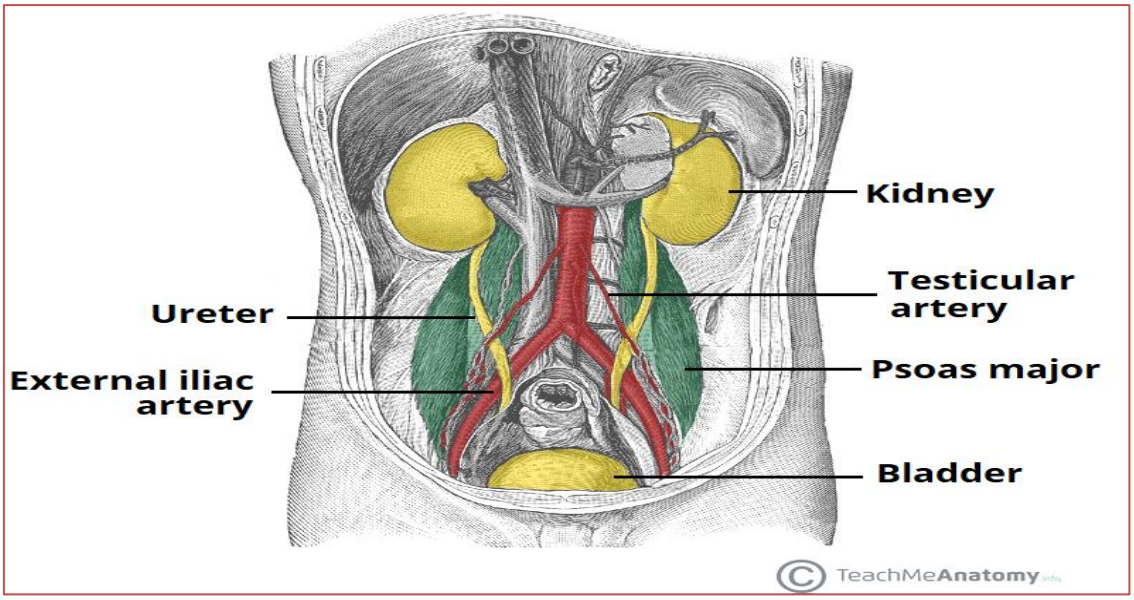

Figure 20:the anatomical course of the ureters from the renal pelvis to the bladder(20).

2. Neurovascular supply and lymphatic drainage:

The ureter is a structure that has developed via the ureteric bud from the mesonephric duct, and then followed the kidney during its ascend to the final lumbar position in the retroperitoneum.

The ureters have an expansive anastomosing network of arterial supply and venous drainage along their length. The proximal end receives arterial supply from the ureteric branch of the renal artery.

Contributions from the ovarian artery (testicular artery in males) as well as a direct ureteric branch from the abdominal aorta supply the middle segment. The distal portion receives its arterial supply from ureteric branches from both the superior and inferior vesical arteries. They are drained by accompanying veins.

Neuronal supply to the ureters comes from both divisions of the autonomic nervous system. Thoracolumbar outflow from T10-L1 provides sympathetic innervation via the:

- renal plexus and ganglia
- renal and upper ureteric branches from the intermesenteric plexus proximally
- middle ureteric branch of the intermesenteric plexus in the middle segment

In the true pelvis, the ureter receives parasympathetic supply from the pelvic splanchnic nerves and from the inferior hypogastric plexus. These innervations are not pertinent to the generation and maintenance of peristaltic action along the ureters; as this arises from pacemaker cells in the renal pelvis and calyces. However, both the sympathetic and parasympathetic divisions are capable of increasing ureteric peristalsis.

The lymphatic drainage of the proximal part of the ureters is similar to that of the kidneys, and therefore drains into the lateral caval nodes on the right, or lateral aortic nodes on the left. Distally, they drain to the internal and external iliac nodes. The middle segment drains to the common iliac and precaval/preaortic nodes. All lymph drained from the ureters eventually makes its way to the cisterna chyli and thoracic duct before returning to systemic circulation.

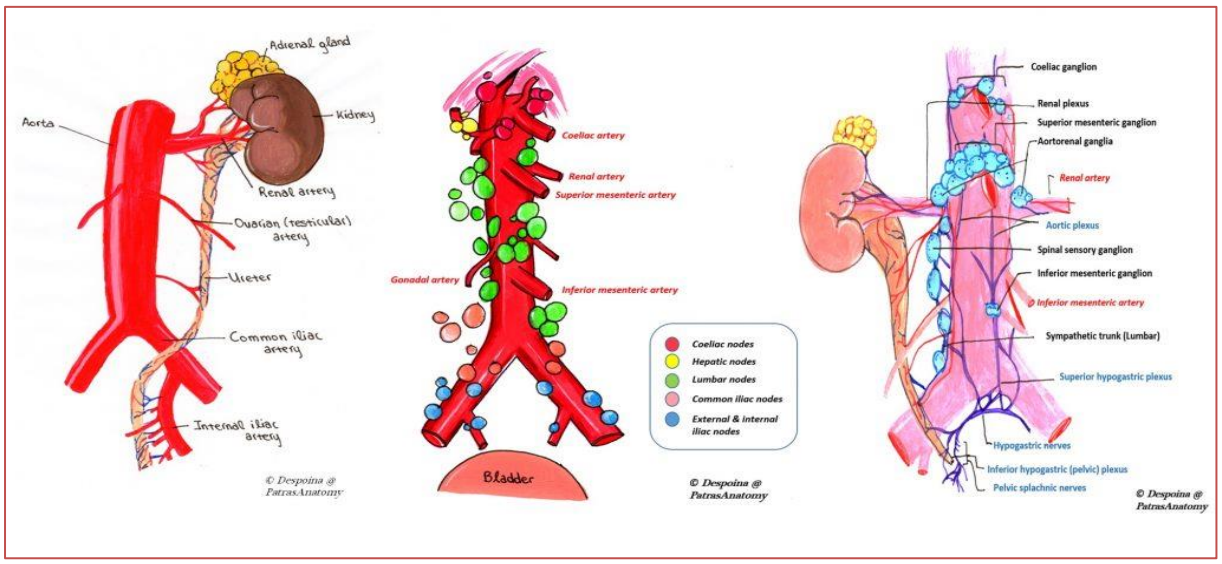

Figure 21:The neurovascular and lymphatic supply to the ureters.

3. Anatomy of the ureterovesical junction:

The ureterovesical junction is located where the ureter (the tube that drains urine from the kidney) meets the bladder. Ureterovesical junction (UVJ) obstruction refers to a blockage to this area. The obstruction impedes the flow of urine down to the bladder, causing the urine to back up into and dilate the ureters and kidney (megaureter and hydronephrosis).

The terminal ureter passes obliquely downwards, anteriorly and medially through the bladder wall , this ureter has 2 muscle systems:

- **Urethro-trigonal musculature:**

It consists of 3 parts: [figure 22].

- ✓ Juxta-vesical ureter: extends 3 cm above the ureteral hiatus and is formed mainly of longitudinal fibres.
- ✓ Intravesical ureter: Consists of 2 segments:

o Intramural: surrounded by the detrusor, 9 mm long, consisting of longitudinal muscle fibres only.

o Submural: rests on the detrusor, covered by the bladder mucosa formed by longitudinal fibres. At the level of the meatus, a contingent of these muscle fibres extends into the upper edge of the trigone while another descends towards the bladder neck, so there is no interruption between the ureteral and trigonal musculature.

- ✓ The superficial trigone: Formed by the extension of the ureteral musculature itself, enveloped in dense connective tissue. Its upper border is the densest formation of the trigone.

- **Peri-ureteral musculature and the deep trigone:**

The ureter is surrounded by two sheaths, a deep sheath and a superficial sheath, known as Waldeyer's sheath.

The deep sheath is made up of longitudinal ureteral muscle fibres, which surround the ureter and extend towards the bladder neck to form the superficial trigone

The superficial sheath is derived from the detrusor (bladder muscle) which surrounds the juxta and intra-vesical ureter and inserts below the bladder neck to form the deep trigone[figure 23] **(21).**

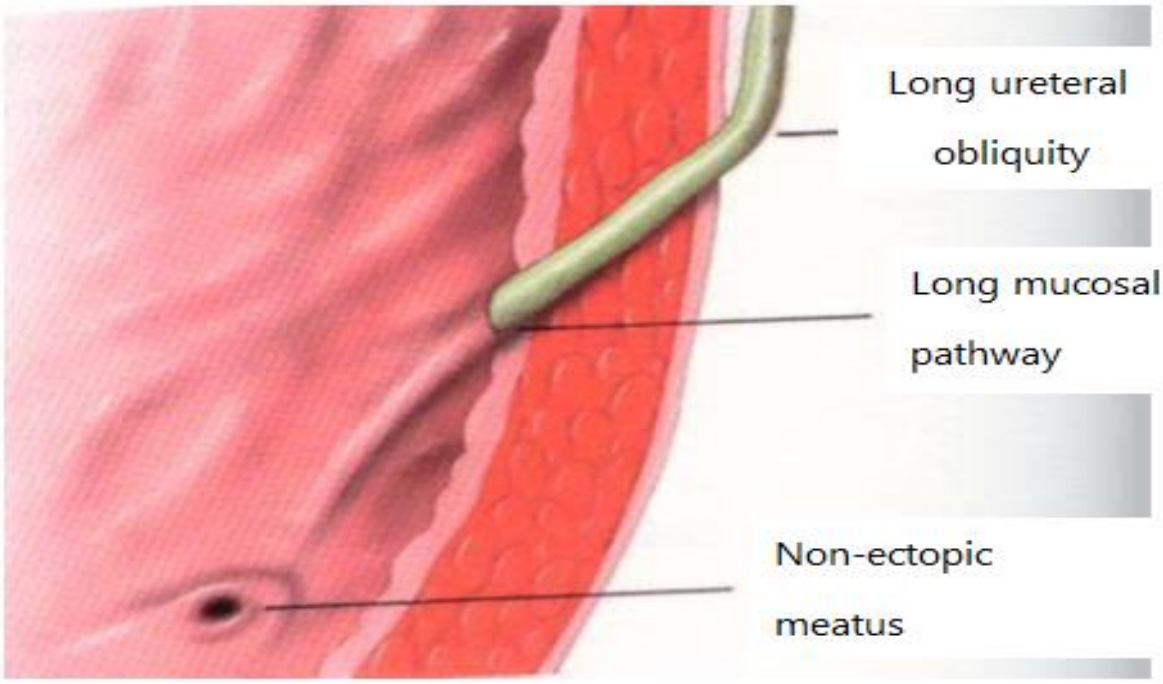

Figure 22: ANATOMY OF THE URETEROVESICAL JUNCTION.

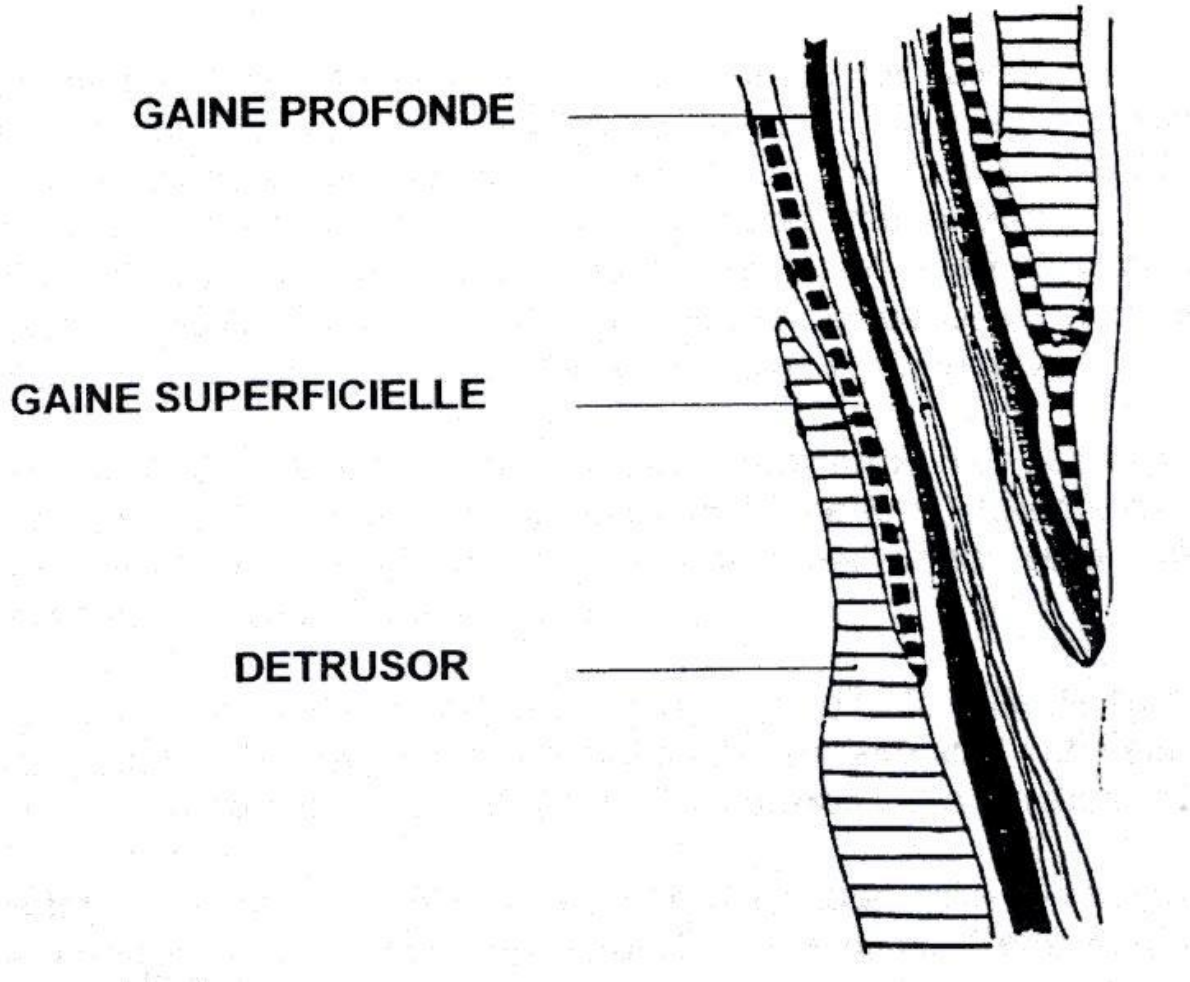

Figure 23: Schematic representation of inter-trigonal musculature.

4. Embryology:(22)

At the fifth week of development, the ureteric bud arises as a diverticulum from the mesonephric (Wolfian) duct. The bud grows laterally and invades the center of the metanephrogenic blastema, the primordial renal tissue. The meeting of these two tissues causes changes in the bud and the metanephros. The metanephrogenic blastema forms glomeruli, proximal tubules and distal tubules. The ureteric bud divides and branches forming the renal pelvis, infundibulae, calyces, and collecting tubules which will provide a conduit for urine drainage in the mature kidney. This process is known as the induction of the kidney.

From 28 to 35 days of development, the ureter is patent, probably as a result of the mesonephros producing urine which fills the tube. From 37 to 40 days of development the ureter loses its lumen. At 40 days of development the ureter regains a lumen. Starting at the midpoint and progressing in both directions toward the developing kidney and the urogenital sinus, the lumen of the ureter reforms. The last segments of the ureter to gain a lumen are at either end (kidney or urogenital sinus).

As development of the bladder progresses the mesonephric duct and the attached ureter are incorporated into the base of the bladder and the proximal urethra. In males, the mesonephric duct drains into the prostatic urethra as the ejaculatory duct. In females, the mesonephric duct regresses and the ureter alone remains. As the mesonephric duct and ureter are absorbed into the base of the bladder, they rotate so that the ureter meets the bladder cephalad to the point at which the mesonephric duct meets the urethra.

At the point where the ureter joins the urogenital sinus, a thin membrane (Chawalla's membrane) develops which separates the two lumens. This membrane then ruptures allowing passage of fetal urine into the urogenital sinus.

At nine weeks of development the metanephros, which will become the mature kidney, starts to produce urine. As this fetal urine drains into the kidney, patency of the ureter is maintained. Smooth muscle develops in the ureteric wall. Later, this muscle will generate and propagate peristaltic contractions to conduct urine from the kidney to the bladder.

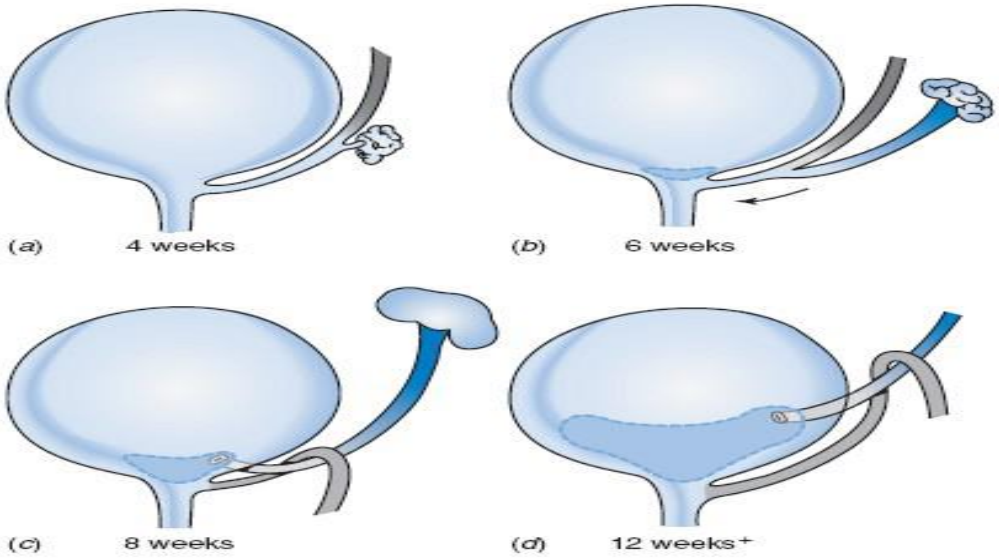

Figure 24: Normal Ureter Development.

The first signs of ureteral muscularization are seen at 12 weeks of gestation. Smooth muscle differentiation is first noted at the ureterovesical junction (UVJ) and then ascends cranially toward the upper collecting system(23). With time, these fibers, which are initially randomly arranged in the wall ofthe ureter, become more numerous and assume a more specific orientation(24). The epithelium attains atransitional configuration by 14 weeks.

Proper development and spatial orientation of the ureter as it combines with the bladder are dependent on a number of molecular factors. The renin– angiotensin system appears to play a major role in ureteral development. AT2 is expressed in high concentration by the mesenchymal cells that are positioned adjacent to the ureteric bud at early developmental stages. The fact that expression of this receptor markedly decreases after birth supports the contention that AT2 might play an important role in ureteral development.(25) Mice in whom the AT2 gene has been knocked-out demonstrate a high incidence of congenital anomalies of the urinary tract, including VUR, UVJ obstruction, and megaureter(26). These structural anomalies may result from delayed apoptosis of the mesenchymal cells surrounding the ureteric bud. The clinical relevance of these experimental findings has been underscored by the identification of a select group of patients with mutations at this locus who possess a syndrome that includes uteropelvic junction (UPJ) obstruction and megaureter(27).

Proper spatial orientation of the ureter is probably dependent on genes responsible for cell specification and body segmentation. The PAX family of genes has been extensively studied in this regard(28). These genes appear to play a role in the interplay between the ureter and the developing kidney(29). PAX mutations have been implicated in syndromes with VUR and renal anomalies, and are inherited in an autosomal dominant manner(30). However, to date, neither mutations in the PAX genes nor in other genes involving body segmentation have been demonstrated to any significant degree in patients with familial VUR(31).

5. Abnormal Ureter Development:(32)

The normal ureter is a conduit to drain urine effectively at low pressures from the kidney to the bladder. At several points along its development abnormalities can occur which impair its function.

Renal agenesis.

If the ureteric bud fails to arise from the mesonephric duct the metanephros will not be induced to form a kidney (renal agenesis). In some males, the mesonephric duct fails to form. In this case, neither a kidney nor a vas and epididymis will form although the testis may descend to its normal position in the scrotum.

In some females, neither the mesonephric (Wolfian) duct nor the paramesonephric (Müllerian) duct develop. In these girls, a kidney and oviduct are absent on the same side. This combination of renal, ureteric and genital abnormalities is known as the Meyer-Rokitansky Syndrome.

Renal dysplasia.

If the ureteric bud arises in an abnormal position on the mesonephric duct, it will not invade the center of the metanephrogenic blastema. Although contact with the periphery of the blastema may still result in the induction of a kidney, the induced renal tissue may be dysplastic. Although such a kidney will produce urine, it will not function normally.

Vesicoureteric reflux.

The ureteric bud may arise from the mesonephric duct nearer to the urogential sinus (medial) than it should. This may result in renal dysplasia. In addition, as the rotational incorporation of the mesonephric duct and ureter into the developing bladder progresses, the ureteric orifice (where the ureter drains into the bladder) will finally lie lateral to and cephalad to the normal position in the developed bladder. In such cases, the tunnel through which the ureter travels into the bladder will be abnormally short. Vesicoureteric reflux (abnormal flow of urine from the bladder up the ureter to the kidney) may result because the tunnel is too short to make a competent distal ureteric valve. This may explain why many kidneys with high grade reflux have abnormal function. The abnormal position of the ureteric bud results both in renal dysplasia (poor induction of the kidney) and also in reflux (abnormal position or ureteric orifice in the bladder).

Megaureter

If the musculature of the distal ureter is deficient or abnormal, that portion of the ureter will not conduct a peristaltic wave normally. Although the lumen of the ureter is of normal calibre, the lack of peristalsis through this portion effectively obstructs the flow of urine. The back-up of urine proximal to this aperistaltic segment causes dilation of the ureter and renal pelvis. This condition is referred to as megaureter for obvious reasons.

Ureterocele

As the ureter develops, the distal end of the ureter (the point at which it meets the urogenital sinus) is covered with a thin layer of tissue (Chawalla's membrane). Normally the membrane ruptures allowing free passage of urine. If rupture of Chawalla's membrane is delayed, drainage of urine from the fetal kidney will be obstructed. Because Chawalla's membrane lies on the interior of the musculature of the developing bladder, the distal ureter (that portion inside the urogenital sinus) will be enlarged. This distal enlargement of the ureter within the bladder is called a ureterocele.

Ectopic Ureter

The ureteric bud may arise farther from the urogenital sinus than it should. This may result in renal dysplasia as described above. In this case, however, the ureter will come to lie medial and distal to the normal position in the mature bladder. Should the ureteric bud arise still further lateral on the mesonephric duct, the mature ureter may open at the bladder neck, in the urethra, in the vagina (in females, of course) or on the vas deverens (in males).

The lumen of each ureter is lined by a mucosal layer of transitional epithelium, which accommodates the increase in pressure that accompanies increases in the volume of urine leaving the kidney; thereby aiding to minimize the risk of rupturing the ureters. These conduits have several in-folding caused by multiple layers of smooth muscle throughout the ureteral wall.

From a histological perspective, there are two muscular layers in the wall of the ureter: a longitudinal and a circular layer. In the lower segment of the ureters, another longitudinal layer can be found proximal to the bladder. Also interesting to note is that urine is propelled along the ureters by peristaltic motions initiated by pacemaker cells in the proximal renal pelvis. The whitish, non-pulsatile exterior along with the peristaltic waves helps to distinguish between ureters and blood vessels in vivo.

6. Histolgy:(20)

The ureters are collapsible S-shaped channels, each about 25 cm in length. They are widest at the renal pelvis and narrow progressively as they enter the urinary bladder in the concavity of the true pelvis.

The ureter is composed of 3 different layers from the surface to the depth:

- The adventitia.

- The muscularis.

- The mucosa.

a) THE ADVENTICE :

It consists of bundles of collagen fibres and some elastic fibres, fibrocytes, vessels and nerves.

b) THE MUSCULARIS :

It is formed of 2 layers:

- An inner layer with a longitudinal direction.

- A circular outer layer.

In its abdominal part, the muscular bundles of the ureter form a helical path, while in its pelvic part, the external spirals become horizontal.

In the juxta vesical part of the ureter, there are more prominent internal longitudinal fibres, whereas in the intra vesical course, the ureter contains mainly longitudinal fibres which will continue with the vesical wall to form the upper part of the trigone separated from the detrusor.

The superficial trigonal muscular part, thin from the ureter, ends below at the level of the supramental part of the urethra.

c) THE MUCKLES :

Consists of:

The epithelium: It is stratified squamous type excreto-urinary, consisting of 5 layers when collapsed and 2 layers when distended.

Chorion: It is made up of connective tissue that is denser on the surface than in depth, in contact with the smooth muscle, with elastic and collagen fibres, the latter being more abundant in depth.

The longitudinally oriented folds are prominent and are responsible for the star-like appearance of the ureteral lumen on transverse section.

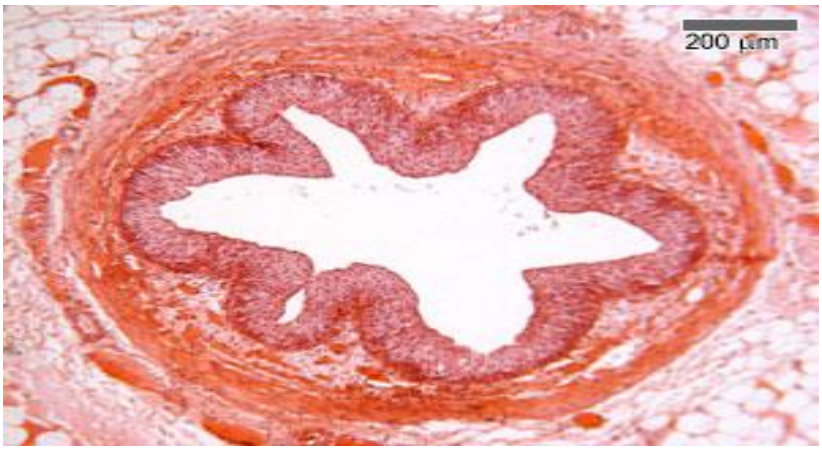

Figure 25: cross section of uretere under optical microscope.

7. Structure and ultrastructure of normal and MEGAURETER:

The muscular arrangement of the ureter is best observed under the light microscope, but its low resolution makes for difficulties in the interpretation of detailed cellular morphology. The higher resolution of the electron microscope allows for a more accurate description of the fine cellular structure, but the minuteness of the sample to be examined makes an organ survey impractical by this means. The arrangement of the muscle fibers of the ureter varies. The renal pelvic muscle fibers run obliquely and the muscle bundles are separated by connective tissue. the fixation of the ureter to the trigonal muscle, the support of the detrusor posteriorly and the compressibility of the ureter, account for the one-way action of the ureterovesical valvular mechanism. The periureteral sheath is largely composed of connective tissue but also contains some muscle fibers that are "shed" into the bladder musculature. There is a potentially dissectable space beneath the adventitia and the longitudinal muscle fibers of the intravesical section of the ureter (Figure 26). This space accounts for the ability of the bladder wall to slide proximally over the terminal ureter during bladder filling. This acts as another mechanism of reflux prevention by lengthening the intravesical ureter as the bladder distends. The normal human cadaveric bladder does not allow reflux even when distended until it ruptures. Thus the mechanism for reflux prevention is passive and depends upon the intravesical portion of the ureter being compressed against the detrusor muscle behind it.

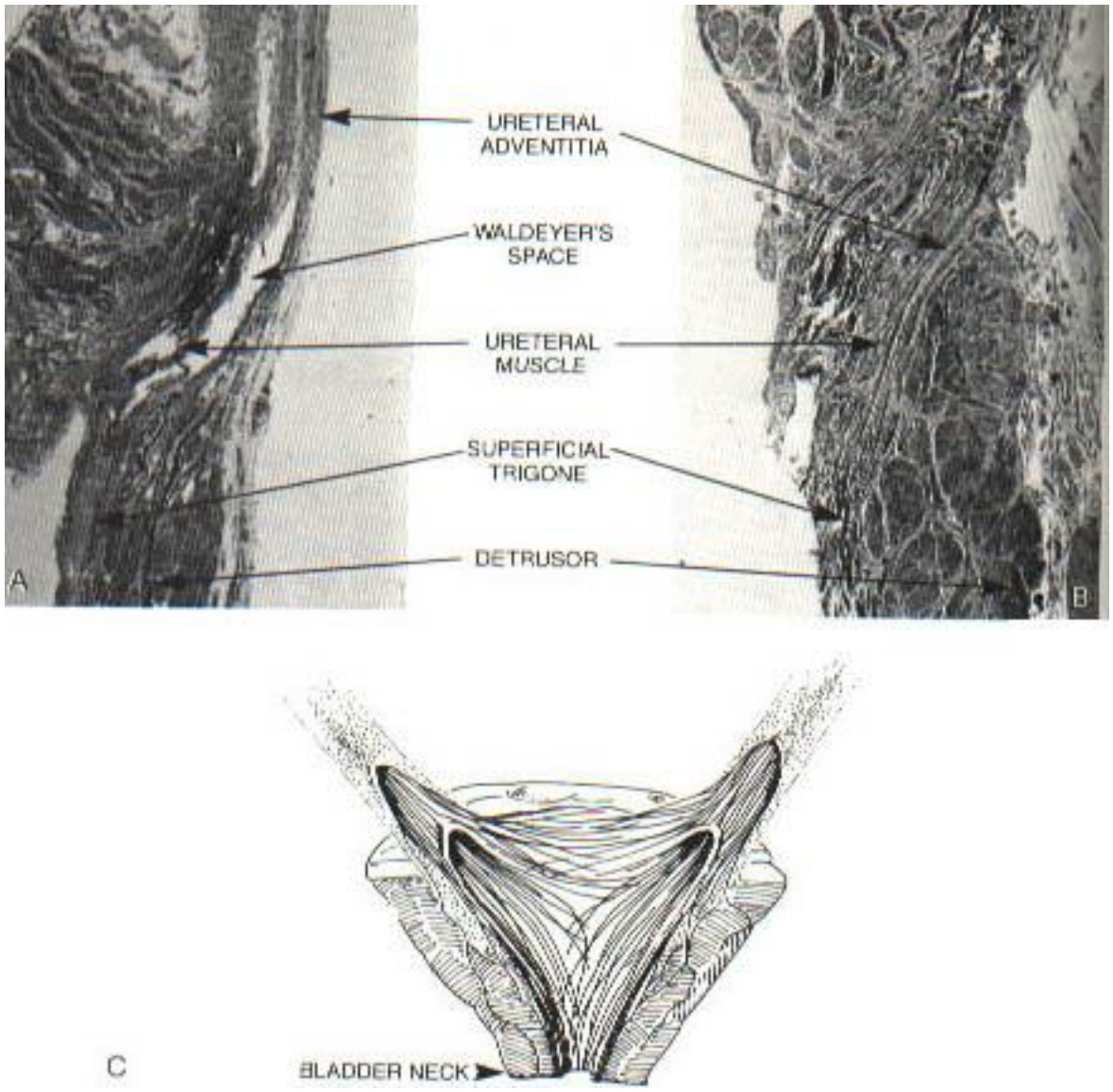

Figure 26: Distal ureter and trigone. A, Sagittal section of ureterovesical junction. B, Sagittal section of intravesical ureter. C, Diagrammatic representation of ureterovesical junction.

Cussen(33) studied the morphology of congenitally dilated ureters. He measured the muscle population and the size of smooth muscle cells of megaureters. In obstructed megaureters (both primary and secondary types) he noted muscle hypertrophy and hyperplasia. These changes were absent or minimal in refluxing megaureters and ureters of prune belly syndrome.

Under electron microscopy the muscle cells are grouped in bundles, each bundle containing 50 to 100 cells.(34) Collagen fibers bundle. The space between the muscle cells is filled with ground sub elastic fibers, and occasional collagen fibers. The neonatal ureter contains, many more elastic fibers than the adult ureter, hence the enormous propensity of the infantile ureter for dilatation and tortuosity. Some extend from one bundle to the other, maintaining the continuity of ureteral musculature. The cell cytoplasm of the muscle cells is filled with electron-dense myofilaments. There are two types of filaments-the thin filaments (actin) and the much smaller population of thick and electron-dense filaments (myosin). In some regions the plasma membranes of the muscle cells are closely opposed, forming close junctions or nexi where the outer basal laminae continue from one cell to another and

the inner plasma membranes are intimately opposed. The structural organization of the nexus suggests that it represents a pathway for the transmission of impulses from one cell to another. The intracellular mitochondria are the powerhouse of the cell and contain all the enzymes necessary for Krebs' cycle. The mitochondria use oxygen to produce the energy-rich compound adenosine triphosphate (ATP). The endoplasmic reticulum and ribosomes represent the sites for protein synthesis. A Golgi apparatus, consisting of lamellae, vesicles, and vacuoles is usually oriented within the cell. Its main function is the addition of carbohydrates to the protein synthesized by the endoplasmic reticulum, and the subsequent concentration, packaging, and transportation of secretory material to the cell surfaces. The nucleus contains chromatin and the nucleolus, which is an intranuclear electron-dense structure in nuclear membrane. The nucleus reflects the level of cellular activity and maturity. Dead or dying cells have shriveled or ruptured nuclei.

Congenital ureteral muscle abnormalities may be quantitative, qualitative, or both. Many wide ureters contain adequate muscle cells and are capable of contraction despite their large diameter. However, there is a group of megaureters that are anatomically and functionally so severely compromised as to warrant the term "dysplasia."4 They have poor or absent ureteral motility. Under light microscopy these ureters are noted to be poorly muscularized and their muscle/connective tissue ratio is decreased. Electron microscopy reveals small deformed muscle cells with nexi that are markedly decreased or absent. The intracellular organelles are poorly defined, and excessive collagen and ground substance are present throughout the dysplastic ureter. (35) Subsequent studies have confirmed these findings and re-corded the incidence of muscle dysplasia in various types of megaureter. (36) Dysplastic changes varied from 24 per cent in nonrefluxing megaureters to 44 per cent among obstructed ectopic ureters. (36)

It is therefore clear that the dilated ureter exhibits a spectrum under electron microscopy as shown in Figure 27 that is expressed primarily by the degree of muscle cell disorder. Muscle cell contraction is a complex process that demands energy supplied by the adenosine triphosphate from intracellular mitochondria. The energy causes interdigitation of the actin and myosin proteins within the cell to form the complex responsible for muscle fiber shortening. The dysplastic ureter lacks the essential ingredients for impulse propagation and muscle contraction. Effective peristalsis is apt to be compromised in dilated ureters with poor quality musculature, which unable to coapt adequately around a bolus or urine. The muscle abnormalities cannot be corrected, but the deficiency of the bolus propulsion may be improved by reduction of the ureteral lumen. Ureteral tapering may allow even the somewhat compromised contractile elements to propel the urine more efficiently although usually below normal capacity. Ureters with severely damaged muscle cells are morphologically end-stage organs usually draining severely dysplastic kidneys and therefore do not lend themselves to reconstructive surgery.

Figure 27: Spectrum of megaureter under electron microscopy. A, Megaureter with normal muscle cells. B, Megaureter containing separated muscle cells and ruptured nexus. C, Megaureter containing excessive collagen and ground substance with dying muscle cells.

8. Physiolàgycal recall: [6] [7] [8]

The principal function of the ureter is to transport urine from the kidney to the bladder. The transport mechanism includes distention and myogenic and neurogenic activities. The law of Hagen-Poiseuille may be applied to urine flow through an aperistaltic dilated ureter. This law implies that the flow may increase 10-fold for every 1-mm increase in diameter of a tube. Thus ureteral dilatation may be a mechanism for maintaining a low intraluminal pressure. It has been demonstrated that the muscle cell integrity is adequate in many uninfected and massively dilated ureters.(37) However, since the ureteral lumen is incompletely obliterated by contraction, the peristalsis is not bolus-propelling and retrograde regurgitation of the urine occurs (Fig. 11-4).

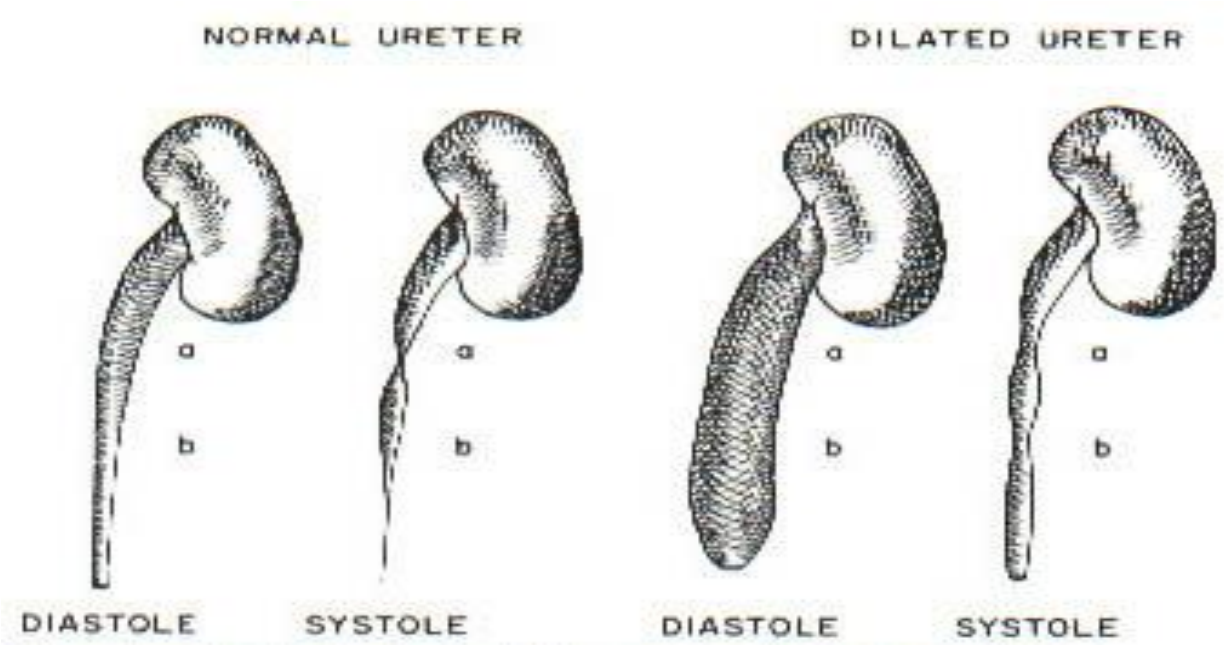

Figure 28: Transmission of bolus of urine in normal and dilated ureters. In the latter, incomplete obliteration of the lumen accounts for ineffective propulsion of the bolus from a to b..

The Laplace equation expresses the mathematical relationship of the variables relevant to our discussion.

$$\text{pressure} = (\text{tension} * \text{wall thickness}) / \text{radius}$$

a) The mechanism of ureteral peristaltic activity:

Ureteral peristaltic activity is the most important function of the ureter. It originates frompace-makers cells: interstitial myoblastic cells grouped in clusters in the region of insertion of the small calyces and then become rarer as we move away from the calyces.

The electrical activity of the smooth muscle cell is related to the movement of ions across the cell membrane which allows the propagation of ureteral peristaltic activity by simple contiguity, thanks to the nexuses (specialized areas of fusion between muscle cells) with a speed of 2 to 5cm/second.

The coordination between the peristaltic contractions allows the transport of urine from the pelvis to the bladder Thus, the entire upper urinary tract is excitable, the contractile wave arises from the calyces (Pace-Maker activity) and is transmitted through the pelvis to the ureter. This ureter is crossed by one to two waves per minute in basal operation (figure 29).

b) Pressure in the ureter:

The basal pelvis pressure is less than 10 cmH2O, the ureter pressure varies between 2 and 6 cmH2O, the contraction pressures of the ureter are estimated to be 10-15 cmH2O at the lumbar level and 25-30 cmH2O at the ureterovesical junction.

If the bladder pressure increases, there will be an increase in ureteral pressure with an increase in the frequency of its contractions.

- Under normal diuretic conditions, the frequency of contractions decreases from the calyx to the ureter to 1 or 2 per minute. The amplitude of contractions increases along the ureter.
- In hyperdiuresis, the frequency of contractions in the ureter increases as does the volume of the bolus, and to a lesser extent, the amplitude of contractions. Transport is still active through ureteral peristalsis.
- For a higher diuresis, the boluses merge, the basal pressure rises and equals over the entire height of the upper excretory pathway, while the contraction pressure is damped; the transport of urine depends only on hydrostatic pressure.

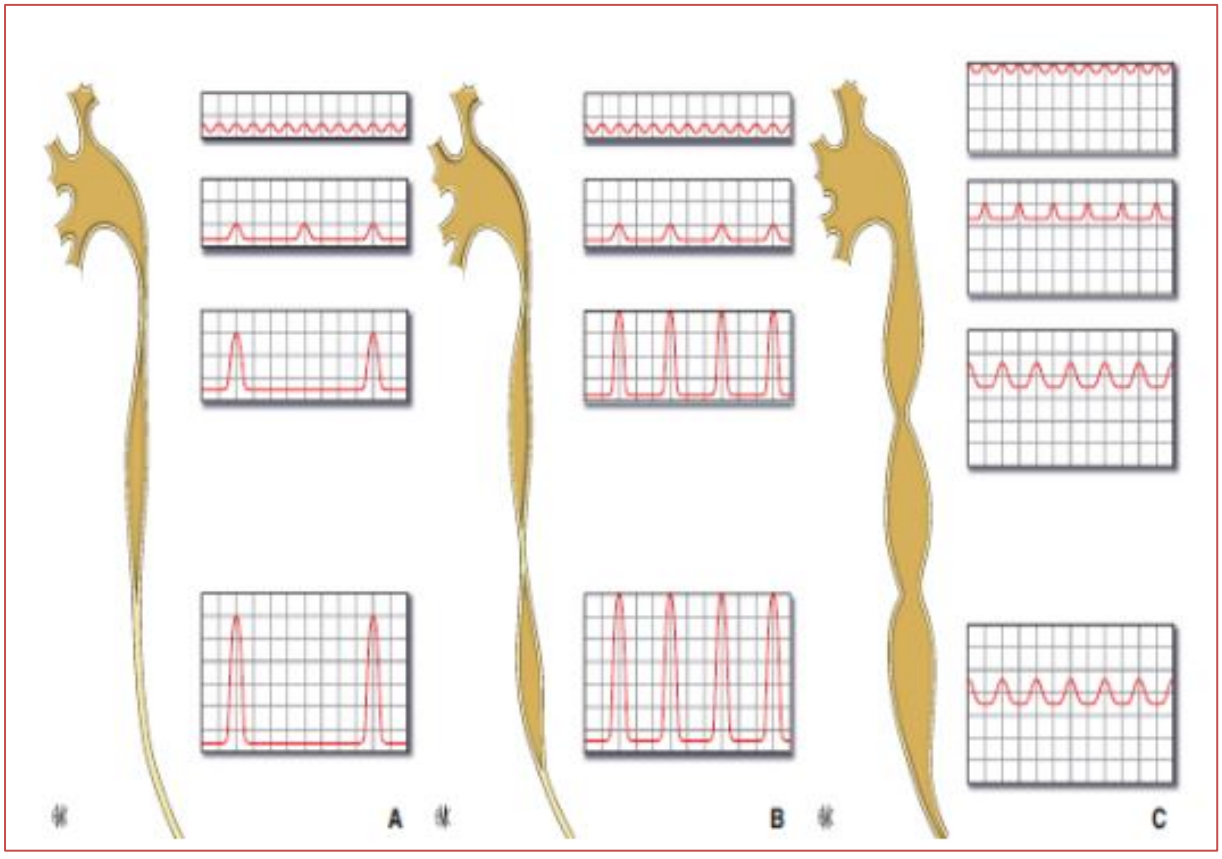

Figure 29: Urinary truct function.

9. Physiopath(38) :

Increased ureteral diameter decreases the intraluminal pressure. Low basal pressures have been recorded in most hydronephrotic pelves(39) and megaureters.(40) The equation indicates that surgical reduction of the diameter of the ureter should result in an increase in its ability to generate the pressure necessary to propel the urine distally.

A. Refluxing PM:

Refluxing PM is caused by a short or absent intravesical ureter, congenital para-ureteric diverticulum or other derangement of the VUJ. Lee et al.(41) assumed that the marked increase in collagen and significant decrease in smooth muscle could be a major contributing factor in the pathogenesis of refluxing PM.

They also expected that this group of patients would have a lower chance of surgical success even with a technically sound reimplantation. There are a few patients with refluxing PM who have the megacystis-megaureter association, with the characteristics of bilateral hydroureteronephrosis, a large thin-walled bladder and bilateral high-grade VUR. The cause of the upper tract dilatation and the enlarged bladder is the constant recycling of massive amounts of refluxed urine. Another special group of patients with dilated ureters are those with prune-belly syndrome, in which the ureterectasis may be caused by reflux or VUJ obstruction, or may be not refluxing and unobstructed, and categorized according to these characteristics.

In a few patients an element of obstruction may be combined with reflux. In a series of 400 refluxing renal units, Weiss and Lyton (42) found concomitant VUJ obstruction in nine (2%). A dysgenetic distal ureteric segment that not only fails to cope within the intramural tunnel but also has ineffective peristalsis is implicated. Identification is important because the management of obstruction often differs from that of reflux alone.

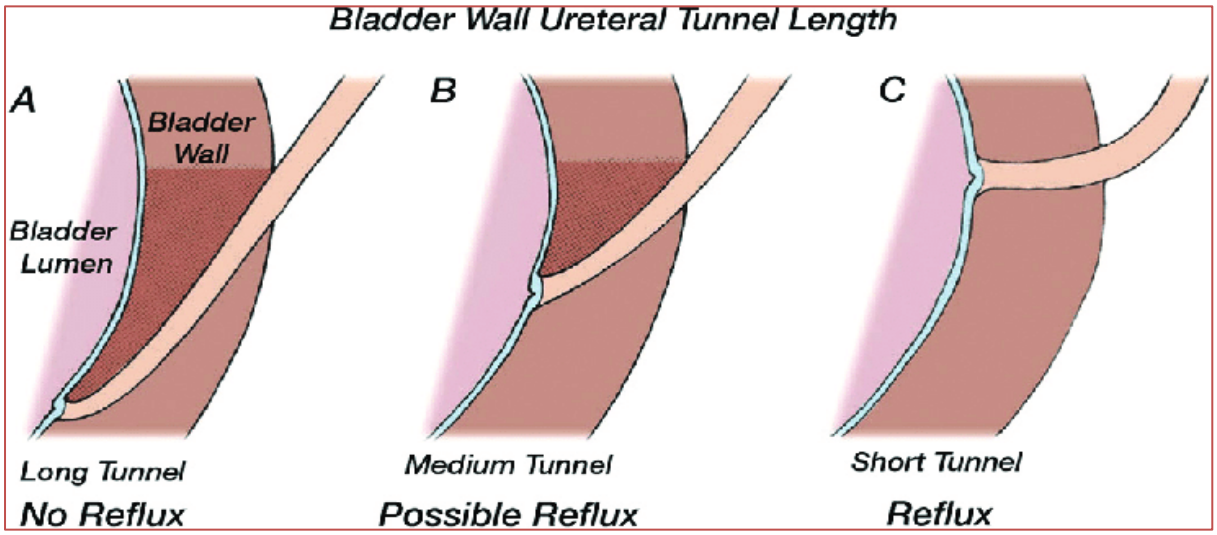

Figure 30: Schematic representation of intravesical ureter length

B. Obstructed PM : (43)

There is a general agreement that there is no true narrowing at the VUJ, but a functional obstruction arising from an aperistaltic juxtavesical segment 0.5±4 cm long that is unable to transport urine at acceptable rates. The pathogenesis of aperistalsis remains controversial and several explanations have been proposed. Excess collagen deposition has been a common finding. In theory, increased matrix deposition alters cell-to-cell junctions and disrupts myoelectrical propagation and peristalsis. Segmental changes of muscle cells were also highlighted in adynamic segments of obstructed PM, where there is atrophy of the inner longitudinal layer that conducts the peristaltic waves and hypertrophy of the outer circular layer that causes obstruction. Why the distal ureter is usually involved is explained by the embryological studies of Tanagho, who noted that the distal ureter is the last portion to develop its muscular coat and that early muscular differentiation is primarily of the circular muscles. The theory of segmental changes of muscle cells has been recently supported by Nicotina et al., who detected TGF-b activity in the narrowed segments of PM but not in the dilated segments. TGF activity is not expected in the ureter postnatally, suggesting that there is some form of developmental arrest of the ureter at 20±28 weeks of gestation. This may explain the frequent spontaneous resolution of PM within the first 2 years of postnatal life, where the circular muscular pattern, which is typical of the fetal ureter, changes progressively into the double muscle layers of the full-term infant.

A band of circumferential tissue devoid of muscle in the most distal portion of the narrowed segment is also suggested as a cause of functional obstruction in some cases. Tokunaka and Koyanagi also suggested that a generously developed bulky peri-ureteric sheath appeared to contribute to obstruction in some cases of nonrefluxing megaureters. Dixon et al. recently showed another cause of obstructive PM associated with ectopic ureteric insertion. There is a thick sleeve of muscle forming a continuous layer surrounding the muscle bundles of the terminal ureter. The muscle forming this outer layer is distinct in its histological appearance and arrangement from the muscle bundles which form the ureteric muscle of the VUJ. Furthermore, this muscle layer is very densely innervated by nonadrenergic (immunoreactive to dopamine betahydroxylase) nerves when compared with the smooth muscle coat of the ureter. The precise mechanism controlling this additional muscle layer remains unknown. This muscle collar has no continuity with either detrusor or ureteric muscle and thus myotonic influences such as those associated with ureteric peristalsis will not be transmitted to this region. Furthermore, the dense nonadrenergic innervation of this muscle collar might cause it to constrict inappropriately, impeding urinary flow, and ultimately lead to the development of megaureter.

Regardless of the cause, altered peristalsis prevents the free outflow of urine and functional obstruction results. Retrograde regurgitation occurs as successive boluses of urine are unable to traverse fully the aberrant distal segment. The degree of ureteric dilatation that results depends on the amount of urine forced to aggregate proximally because it is passed incompletely. This in turn is determined by the degree of distal obstruction and urinary output. This disruption in ureteric dynamics has obvious implications for the renal parenchyma if the collecting system is unable to ameliorate the proximal pressure that can develop. Theoretically, it might be suspected that a compliant dilated ureter would provide a larger reservoir and hence a lower pressure buffering system than with dilatation caused by PUJ obstruction. Thus, it would be less likely for the dilatation caused by VUJ to transmit damaging pressure to the kidney. In contrast, a comparable PUJ obstruction would be likely have more of a direct damaging effect on the kidney with no more than the renal pelvis to diffuse the pressure.

C. Nonrefluxing unobstructed PM :

Most PM in neonates fall into this category; there is neither reflux nor stenosis of the juxtavesical ureter, but the ureter is dilated beginning at a point just above the bladder. The cause of this phenomenon is unknown, but several theoretical explanations were given based on transitional (antenatal to postnatal) renal physiology and histo-anatomy of the developing ureter. Fetal urine production is 4±6 times greater before than after delivery, because of differences in renal vascular resistance, GFR and concentrating ability. This high outflow might contribute to ureteric dilatation in the absence of functionally significant obstruction, as is occasionally seen with diabetes insipidus. Another contributing factor appears to be increased compliance of the fetal ureter because of differences in the deposition of type III collagen, elastin and other extracellular matrix proteins. Finally, partial or transient anatomical or functional obstructions that spontaneously improve with postnatal development could contribute to proximal dilatation. Persistent fetal ureteric folds or delays in the development of normal peristalsis may represent such transient obstructions.

This group of dilated ureters could includes not as primary MU, those with transient dilatation encountered during urosepsis. The bacterial toxins paralyze the muscle cells and render the ureter temporarily atonic. Impressive recovery following antibacterial therapy is expected and often ensues. Nonetheless, chronic low-grade bacteriuria may cause irreversible damage to the ureteral musculature. This has been documented by electron microscopy.(9)

D. MEGAURETERS IN PRUNE BELLY SYNDROME :

Prune belly syndrome presents as a spectrum of abnormalities of the anterior abdominal wall and the urinary tract and in the degree of testicular descent. A high mortality, with almost 50 per cent of the babies dying by two years of age as a result of renal failure, urosepsis, or both.. The abdominal wall defect varies from virtually complete absence of contractile muscle to a limited muscular defect with a patch of wrinkled skin overlying the large bladder. The degree of abdominal wall involvement does not correlate with the seriousness of the uropathology. Various degrees of ureteral dilatation and tortuosity as well as renal dysplasia are encountered. From the pathologic standpoint the prune belly ureter demonstrates gross, microscopic, and electron microscopic abnormalities. There may be massive dilatation and tortuosity. Microscopically, there is a decrease in the muscle bundles and an increase in the amount of connective tissue. When the cells are viewed by electron microscopy there appears to be a decrease in the thick and thin filaments of the smooth muscle cells and an increase in the homogeneous ground substance. Thus the defect in prune belly megaureter is a distinctive myopathy, with both quantitative and qualitative deficiencies of the musculature.

III. CLASSIFICATIONS :

The megaureter poses not only a problem of terminology but also of classification. There are several classifications, varying from simple to complex:

1) International classification:

The international classification, which appears to be gaining acceptance, is shown in Figure 31 and is based on evaluation of the urinary tract by intravenous pyelography and voiding cystourethrography. The dilatation is essentially due to obstruction or reflux or neither (nonrefluxing, nonobstructed). Each group is further subdivided into primary and secondary. In the former the defect lies in the megaureter itself, in the latter the dilatation is secondary to another disorder, e.g., distal urethral obstruction. In some instances obstruction of the terminal ureter may coexist with reflux, a group that is not included in the above classification. It is important to bear in mind the above basic factors that may lead to ureteral dilatation and to thoroughly evaluate the urinary tract

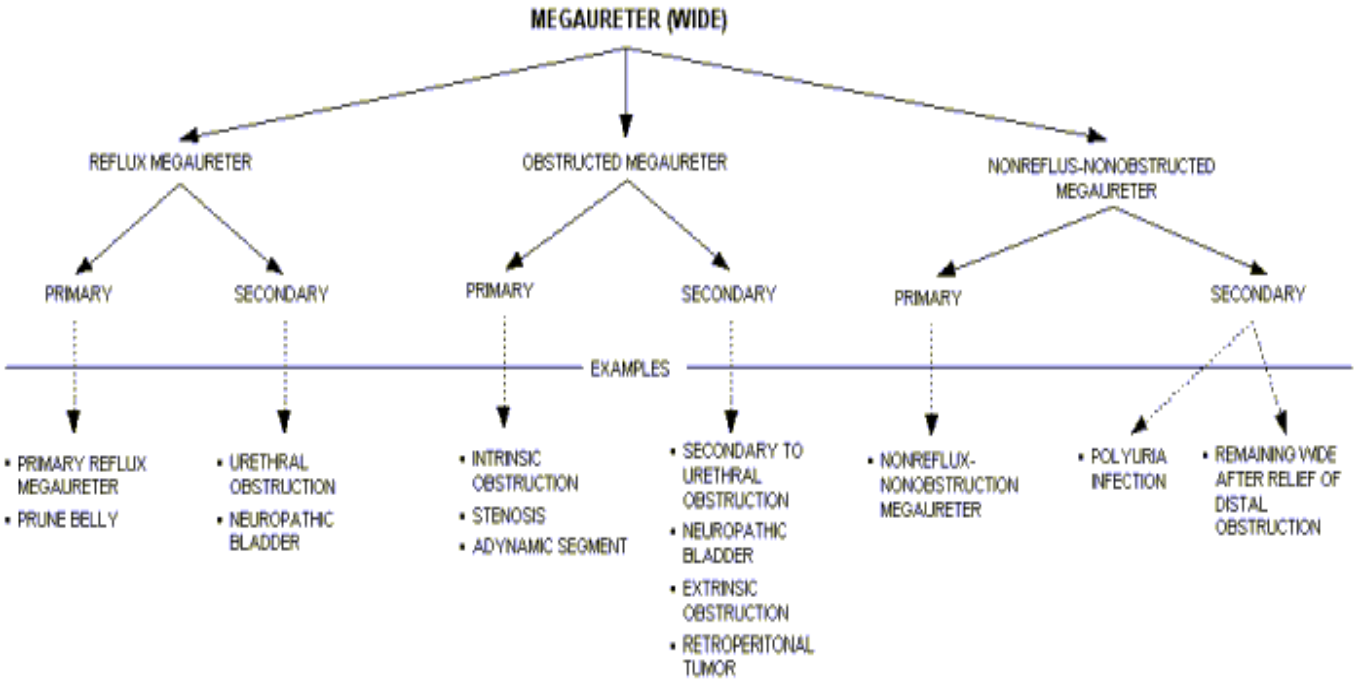

Figure 31: International classification.

2) Functional classification :(44)

King in 1980 criticized the international classification, and proposed a functional classification based on the existence of reflux or an obstruction, we find four groups:

-Non-refluxing non-obstructive megaureter.

-Non-refluxing obstructive megaureter.

-Refluxing non-obstructive megaureter.

-Refluxing obstructive megaureter.

3) Anatomic classification :

The different anatomical classifications of the MUs are based on morphological analyzes provided essentially by intravenous urography.

a) BEURTON's classification :

It is a urographic classification based on the degree of pyelo-caliceal dilatation, the renal impact and the type of mega-ureter:

Type 1A: pelvic mega ureter.

Type 1B: subtotal mega-ureter respecting the sub-pyloric ureter.

Type 2: total mega ureter without sinuosities.

Type 3: sinuous total mega-ureter

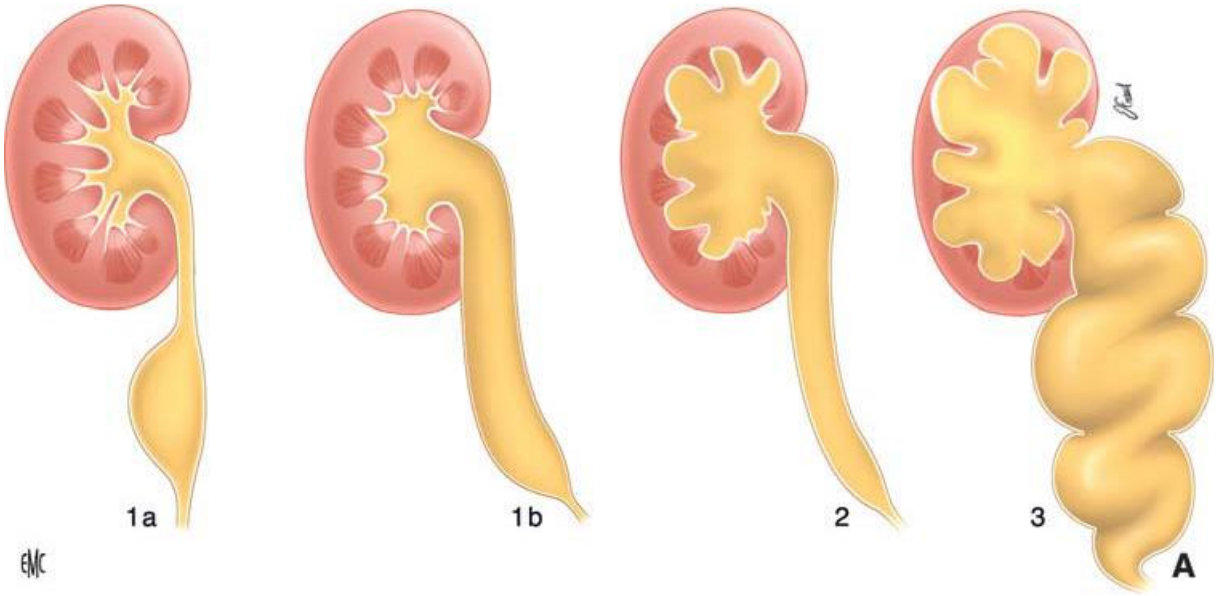

Figure 32:BEURTON's classification.

b) MACLELLAN's classification :

For MacLellan, the classification is proposed in five grades and only concerns mega-ureters associated with pyelocaliceal dilatation based on the ultrasound appearance of the dilatation and the renal parenchyma:

Grade1: pyloric dilatation without dilatation of the calyces.

Grade2: very moderate dilatation of the calyces which remain concave.

Grade3: moderate dilatation of the calyces which keep their shape.

Grade 4: important dilatation of the calyces "in ball" with a renal parenchyma of normal appearance.

Grade 5: important dilatation of the calyces "in ball" with a thinned renal parenchyma.

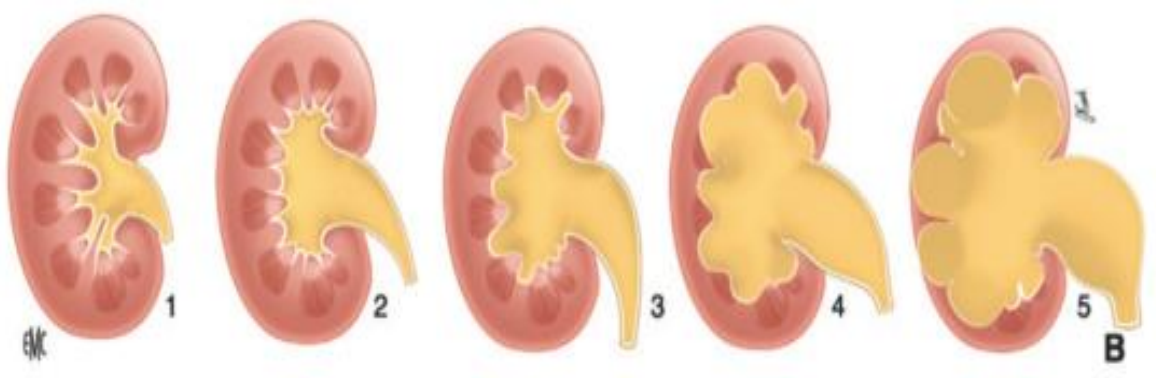

Figure 33: MACLELLAN's classification.

c) The PFISTER and HENDREN classification:(45)

The morphological classification of PFISTER and HENDREN characterizes the degree of ureteral dilatation, it is the simplest in current practice:

Grade 1: normal kidney, with dilatation predominantly in the distal part and may involve the entire ureter.

Grade 2: moderately altered kidney, usually associated with greater ureteral dilatation

Grade 3: atrophy of the renal parenchyma, associated with a significant and total dilatation of the ureter.

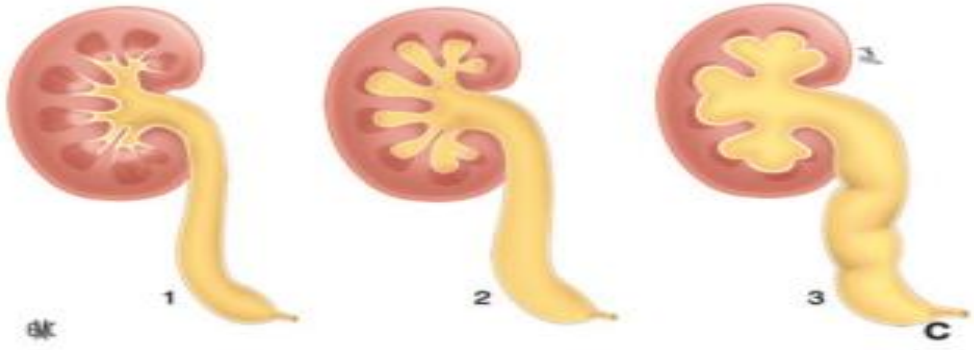

Figure 34: The PFISTER and HENDREN classification.

In daily practice, the severity of mega-ureters is classified according to the dilatation. It is easier to speak of a "very dilated" or "large" mega-ureter (ureteral diameter > 10-15 mm with dilatation of the renal cavities) versus "minimally dilated" or "minor" mega-ureter (ureteral diameter <10 mm with little or no dilated renal cavities).

IV. DEMOGRAPHICS: (1)

Primary MGU is a relatively common finding in neonates and infants referred for urologic evaluation.

Prenatal ultrasound series suggest UVJ obstruction in up to 23% of patients with urinary tract dilatation (Brown et al, 1987). Primary MGU is two to four times more common in **boys** than girls, has a slight predilection (1.6 to 4.5 times) for the **left side**, and is bilateral in approximately 25% of patients (James et al, 1998 ; Shokeir and Nijman, 2000 ; Mouriquand et al, 2001). In up to 10% to 15% of children the contralateral kidney may be absent or dysplastic (Shokeir and Nijman, 2000), and concomitant obstruction of the ipsilateral UPJ area has been described on rare occasion (McGrath et al, 1987). There is no clear evidence for a hereditary predisposition, although some families with more than one affected member have been described (Shokeir and Nijman, 2000).

V. CLINICAL STYDY:

1) Circumstances of discovry :

A. Antenatal Diagnosis:

MUP has become a common cause of prenatally discovered fetal hydronephrosis. The gestational age of discovery of fetal hydronephrosis varies from one series to another; the majority of MUPs are discovered in the 2nd trimester of pregnancy.(46)

In the study made by Ghanmi et al (47), the mean age of antenatal diagnosis of hydronephrosis was 26 weeks of amenorrhea. In the study by Diakité et al(48), the gestational age at the time of screening was 32.3±6.5 SA .

Fetal ultrasound routinely performed during pregnancy allows the identification of 90% of fetal kidneys by the 17th week and 95% by the 22nd week. Dilatations of the upper excretory tract are therefore easily detectable. However, one should be wary of misdiagnosis due to a lack of specificity of fetal ultrasound(49). Antenatal ultrasonography can also be used to assess the extent of the renal impact of the malformation by evaluating the degree of parenchymal atrophy. In case of bilateral involvement, the degree of renal insufficiency will also be assessed by the decrease in the quantity of amniotic fluid. However only morphological examinations performed after birth will make it possible to relate this dilatation to a mega-ureter.

Di Renzo et al (50) noted in their series 56% of MUP discovered antenatally out of 75 children studied, Hosquétis (51) had 20/41 patients diagnosed antenatally.

The antenatal degree of renal pelvis dilatation is graded according to the ultrasound appearance of the renal parenchyma and renal pelvis system according to SFU (Society for Fetal Urology grading system). Consideration of the appearance of the renal parenchyma and not only the measurement of the AP diameter of the renal pelvis is important since the ultimate prognosis of the renal function depends on it.

There isn't a one single definition of renal pelvis dilatation in pregnancy, but Blachar et al. defined CPP when the anteroposterior (AP) diameter of the renal pelvis is 9 mm or greater. Grignon et al. reviewed 34,000 obstetric ultrasounds (after 20 weeks of gestation), and found that an AP diameter of the renal pelvis of 10 mm, or progressive calyx dilation , were predictive radiological signs of postnatal renal pelvix dilatation

In practice, we retain the following:

- An AP(anteroposterior) measurement of the renal pelvis
- An AP dilatation of the renal pelvis between 5 and 10 mm, or classified as grade I or II according to the SFU, is considered as moderate dilatation;
- An AP dilation of the renal pelvis of 10 mm, or classified as SFU grade III or IV, is considered as significant dilation.

The antenatal detection of renal pelvis dilation allows for:

- Screening for possible associated anomalies
- The rapid initiation of uro-radiological examinations in the postnatal period .
- The initiation of antibiotic prophylaxis from age of birth
- A possible urological intervention in utero.

According to the British Association of Paediatric Urologists (BAPU) a diameter > 7mm antenatally is considered abnormal and should be explored postnatally. The consensus is as follows:

- All newborns with antenatal Hydroureteronephrosis should be started on antibiotic prophylaxis for the first 6 to 12 months of life.
- A post-natal ultrasound should be performed between the first and third day of life. This ultrasound should be repeated at six to eight weeks of age and then every three to six months to monitor progress. Persistent ureteral dilatation greater than 10 mm is predictive of surgical treatment.
- Any newborn with bilateral ureteral dilatation or bilateral Hydroureteronephrosis should have cystography routinely within three months of birth to rule out a secondary cause of megaureter or Vesicoureteral Reflux .
- Once a secondary cause has been ruled out, a MAG-3 scan is indicated as a second-line procedure; it should be performed at about six weeks of age and not before to determine whether the MUP is obstructive or not and to decide on the subsequent management

Antenatal prognostic factors for renal failure in the postnatal period are:

- renal dysplasia characterized by the abnormal constitution of the renal blastema and a reduction in the number of functional nephrons. It is objectified by renal hyperechogenicity, Loss of corticomedullary differentiation and the appearance of cortical renal cysts .
- A major dilatation of the renal pelvis, greater than 20 mm from the 30th week of gestation

These indices of renal dysplasia are found in massive dilatation SFU III or IV.

B. Postnatal diagnosis:

Rarely, the mega-ureter is discovered at birth in the presence of renal distress. More frequently mega-ureter manifest in young children in the form of febrile or non-fevery urinary tract infections, which may sometimes take the form of an isolated fever, enuresis, lumbar or abdominal pain.

The MUP is discovered postnatally in front of clinical signs that will lead the investigations to confirm it, but in some cases the MU remains asymptomatic and it is of fortuitous discovery.

MU may manifest in later stages with pain or an upper urinary tract infection or more rarely with renal colic or hematuria, often revealing an associated lithiasis.

2) Clinical examination:

The physical examination is often normal, except for severe forms in newborns and infants where abdominal distension may be noted. In older children, there may be attacks of pyelonephritis with fever, pyuria and pain on palpation of the lumbar and iliac fossa.

The existence of renal insufficiency will be suspected in front of a staturo-ponderal delay. It is sometimes possible to palpate a mass in the iliac fossa or in the flank of an infant .

In severe bilateral forms, symptoms may appear early, such as deterioration of general condition and digestive disorders due to massive dilatation and deterioration of renal function .

3) Paraclinic:

1) RADIOLOGICAL EXAMINATIONS :

The radiological explorations make it possible to establish the diagnosis of the primary megaureter, to eliminate the secondary megaureter, the research of a repercussion on the kidney and the possible complications , They also make it possible to search for other urogenital malformations associated with the megaureter such as:

- Contralateral renal agenesis

- Homo or contralateral pyeloureteral duplication

- Pyeloureteral junction syndrome

- Contralateral vesico-renal reflux

- Posterior urethral valve

- Bladder extrophy

The association of mega ureter with other urogenital malformations shows the interest to systematically search for them in order to prioritize their therapeutic management.

a. Renal and pelvic ultrasound:

It is the key examination for the diagnosis and follow-up of mega ureters (52,53)

Technique: Ultrasound exploration can be performed without special preparation. However, it is advisable to avoid performing this examination during periods of hyperhydration to avoid deceptive effects on the urinary tract [pseudo-dilatation]. The equipment used is a sectorial real-time ultrasound scanner with a frequency of 3.5 MHz, allowing the entire kidney to be explored on one incidence. The sections are longitudinal and transverse along the axes of the kidney, taking into account its triple obliquity in space.

Benefit: It is a painless, non-irradiating, easily reproducible examination with high specificity and sensitivity for the diagnosis of mega ureter(54–56).Ultrasound provides multiple information on the morphology of the upper and lower urinary tract (4,57):

- Measurement of the ureteral diameter in lumbar and pelvic on transverse and longitudinal sections with a full bladder and the appreciation of the sinuous or rectilinear character of the ureter and its peristalsis. It allows the value of the peristalsis to be known before the operation, and to specify the topography and the mode of abduction of the ureter at the level of the bladder (4).
- Search for pyelocalic dilatation with measurement of the anteroposterior diameter of the renal pelvis and the appearance of the calyces. The normal diameter of the ureter in children is 5mm [70]. According to the 2014 Multidisciplinary consensus on the classification of prenatal and postnatal urinary tract dilation (UTD classification system) [147], the anteroposterior diameter of the renal pelvis is 3mm in 1-year-old infants, and it is 6mm at 18 years of age, with an extreme that does not exceed 10mm in children under 5 years of age.
- Study of the renal parenchyma, its echogenicity and its dimensions. This is an important step because it allows to look for indirect signs in favor of a severe obstruction that may lead to or increase the alteration of the renal function [72].
- Study of the bladder: it allows to explore the bladder content and the bladder wall and to appreciate the post-mictional residue and the influence of the bladder emptying on the dilation of the upper excretory tract.

Limitations :

- It is an operator dependent examination.

- The technological level of the equipment.

- The patient's weight can alter the quality of the result

b. Antenatal ultrasound (58,59):

Advances in obstetrical ultrasound have changed the prognosis of neonates with pyelocalic dilatation (PCD) by allowing early management. A study conducted by Czarmiak et al in 2009 revealed that 56.2% of newborns with neonatal PCD could be detected by antenatal ultrasonography.

In practice, the diagnosis of MUP is when dilationof the anterior-posterior renal pelvis on antenatal ultrasonography is greater than or equal to 7mm in the 3rd trimester of pregnancy or SFU II, III or IV according to the classification of the Society of Fetal Urology (SFU).

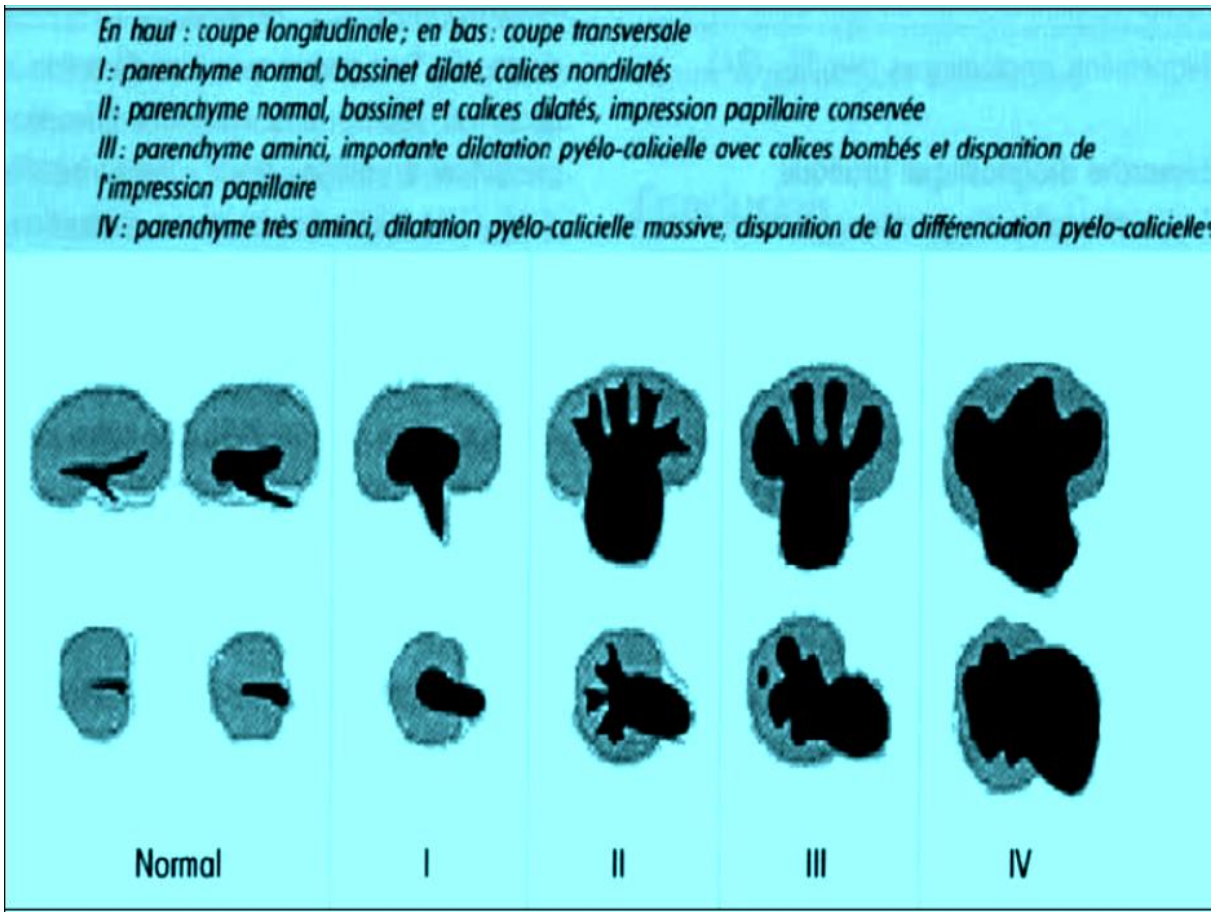

Figure 35: SFU classification.

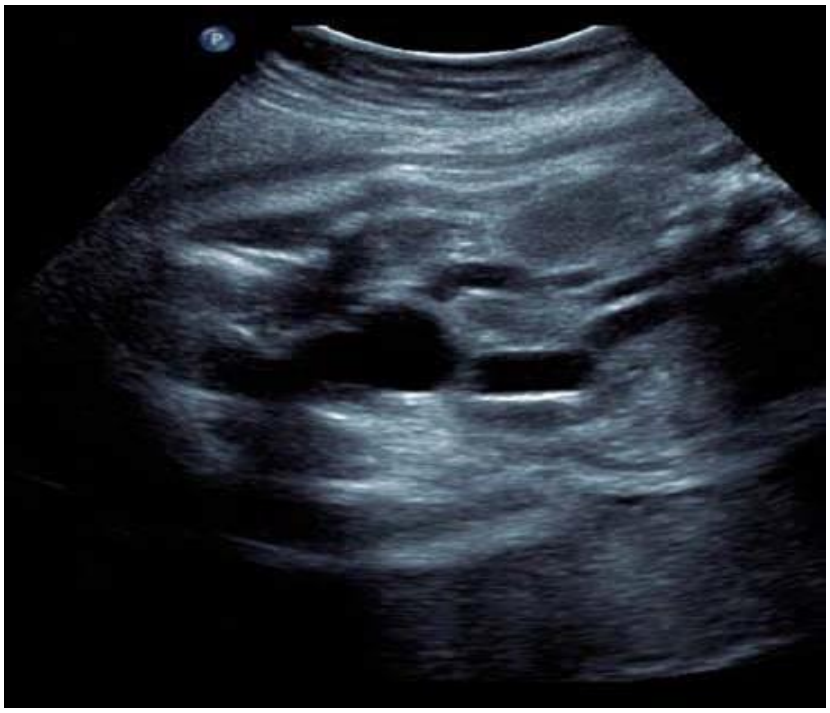

Figure 36: Coronal abdominopelvic section at 32 weeks of amenorrhea: dilated ureters and bladder. The ureters appear as tubular trans-sonorous liquid structures with peristaltic movements; in the antenatal period, there is no certainty about the mechanism: either either primary or by vesico-ureteral reflux.

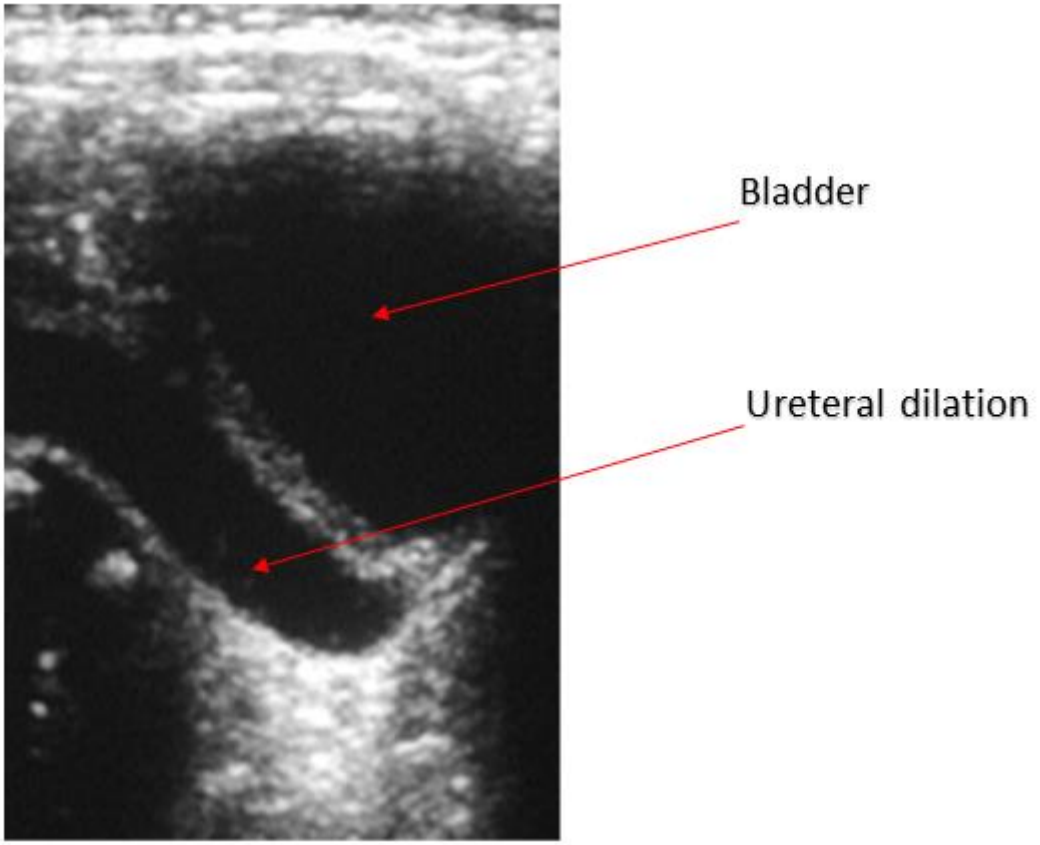

Figure 37: postnatal ultrasound that shows ureteral dilatation.

c. Cystography:

Technique: It is most often performed by bladder catheterization or, more rarely, if catheterization is not possible, by suprapubic puncture of the replenishing bladder. The bladder is filled at low pressure with a water-soluble iodinated contrast medium diluted in serum. The risk of sepsis requires sterilization of the urine before the examination. Televised fluoroscopy monitoring is essential to detect passive reflux during filling.

The most important images are taken during micturition, and should allow visualization of the entire urinary tract. Pictures at the beginning of micturition, but especially at the end of micturition, must be taken, as reflux frequently occurs at the end of micturition, when intra-vesical pressure is at its maximum. A post-voiding film is systematic and in case of reflux, it is necessary to evaluate the possibilities of evacuation of the urine flowing back into the ureter and the ureteral contractility.

Benefit: It is performed to look for a vesico-sphincter abnormality or vesico-renal reflux, not only in the dilated ureter but also in the contralateral excretory tract(57). The presence of reflux does not eliminate the possibility of uretero-vesical junction obstruction. Its role is essential in the detection of malformative uropathy in children with recurrent UTIs (60).

The exploration should be complete with pre-, per- and post-voiding films from the front to detect vesico-renal reflux and from 3/4 to visualize the urethral tract and ensure its normality(61).

BLICKMAN and LEBOWITZ(62) described the cystographic criteria of a refluxing megaureter in order to differentiate it from a simple ureter dilated by a medium or high grade reflux.

These criteria are:

- A dilated ureter upstream of a distal segment of normal caliber.

- A clear delay in the evacuation of the contrast medium having flowed back into the ureter, the contrast medium appears diluted.

- Absence of drainage of the contrast product into the bladder after miction and emptying of the bladder: the urine appears trapped in the ureter.

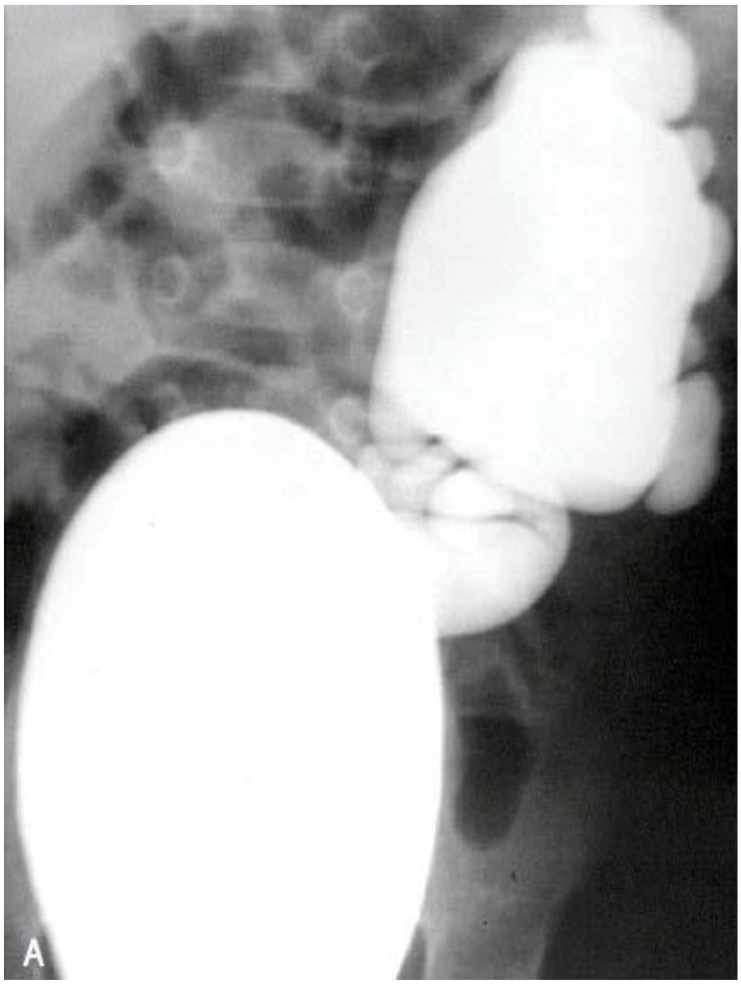

Figure 38: Retrograde cystography of a newborn showing a refluxing megaureter.

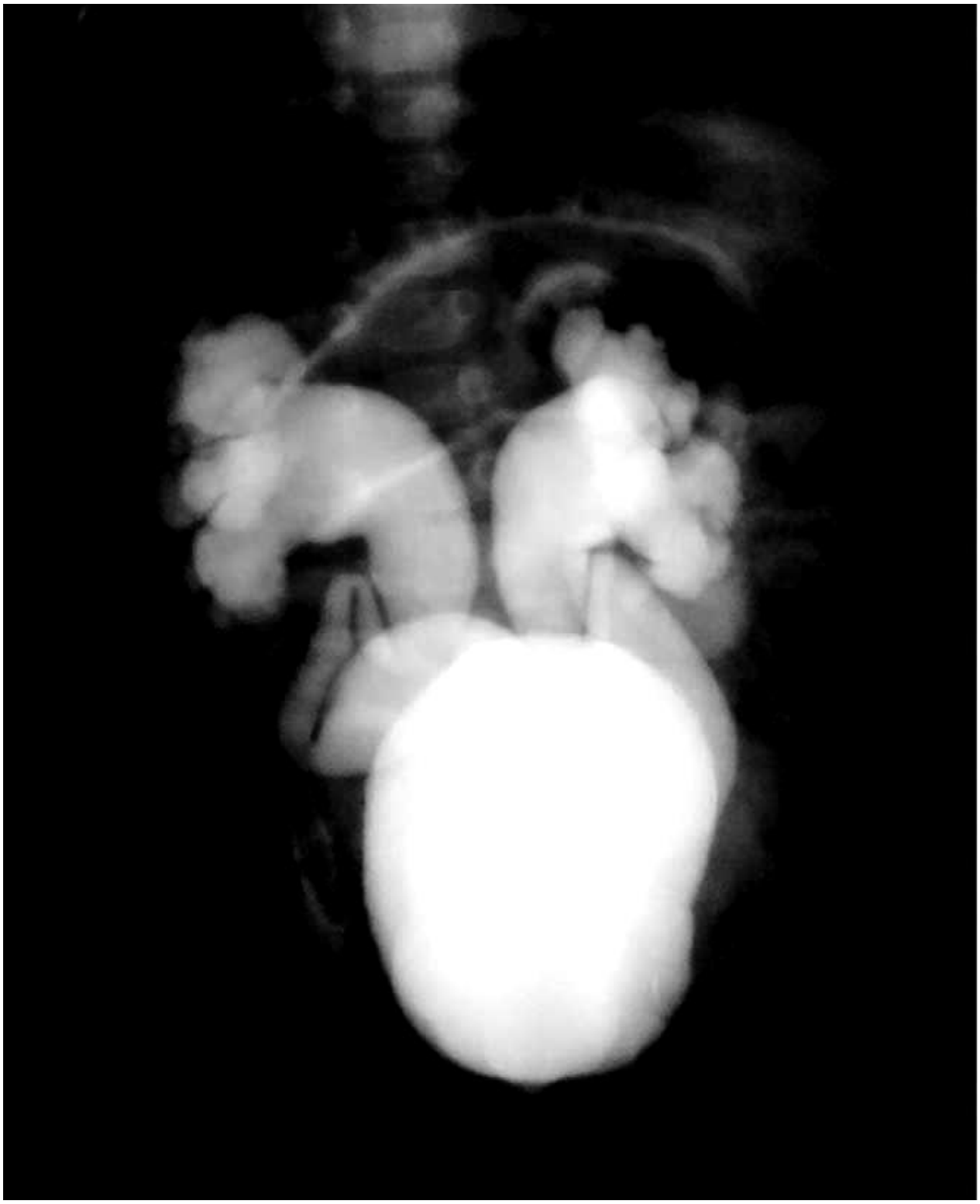

Figure 39: Retrograde cystography (bilateral refluent mega-ureter grade V).

d. Dynamic renal scintigraphy:

Technique: It is a low-radiation examination that is performed after intravenous injection of a molecule marked with a radioactive marker. There is no risk of allergy. For urine flow abnormalities, the most commonly used product is MAG3 [mercapto-acetyl-triglycine] labeled with technetium 99m. The renal uptake of MAG3 allows the study of relative renal function expressed as a percentage and its rapid urinary elimination allows dynamic imaging by analysis of urinary excretion kinetics. The injection of furosemide intravenously 15min after the injection of MAG3 allows the quantification of its urinary excretion to be made more sensitive(63). The results are given in the form of a time/activity curve. The first part of this curve shows the renal uptake and spontaneous elimination of MAG3, the second part shows the elimination of MAG3 under furosemide.

DMSA (dimercapto-succinic acid) scintigraphy labeled with technetium-99m allows an analysis of the functional nephron mass in each kidney. It has replaced mercury bichloride scintigraphy which, although reliable, posed dosimetry problems. However, the interpretation must avoid certain pitfalls:

- 12% of the DMSA is recovered in the urine of the 6 hours following the injection. Significant urinary stasis will delay this elimination and the fixation may be overestimated. It is therefore preferable to carry out the measurement 24 hours after the injection(64).
- The sensitivity of the examination is poor in the case of severe impairment of renal function (clearance less than 30 ml/min).
- Fixation may be transiently lowered in the weeks following an episode of acute pyelonephritis.
- There is a relatively high variability of normal values [30%] (64).

Measurement of clearance from each kidney can be done by isotope nephrogram curve analysis using technetium-99m-labeled DTPA or iodine-131 or 123-labeled hippuran. These same scans can be used for the diagnosis of obstruction by inducing hyperdiuresis (60).

Benefit: The interpretation of this furosemide drainage curve is controversial in the literature. Some authors consider that the absence of elimination of the marker during the time of the examination or that an elimination half-life of the marker greater than 20 min is synonymous with obstruction and that surgical treatment is necessary to preserve renal function, whereas a normal curve proves the absence of obstruction (65,66).

Other authors are more reserved about the value of the marker elimination study because the results may vary depending on the vacuity of the bladder and the position of the subjects at the time of recording [gravity effect]. In the case of the mega ureter, the isotopic study will focus not only on the pyelo-caliceal cavities but also on the height of the ureter.

Thus, the renal scintigraphy allows to confirm the obstructive character of the mega ureter, it is an examination which finds its utility during the various times of the follow-up of a mega ureter. It also has its place during the monitoring of patients postoperatively to assess the functional quality of the result without systematically resorting to repeated urograms. Coupled with ultrasound, it can even supplant postoperative urography.

Limitations: The diagnostic value of dynamic scans has been extensively studied and appears to be good within certain limits.

The diuretic response to furosemide may be greatly impaired if there is a significant decrease in kidney function. Therefore, results should be interpreted with caution.

The replenishing bladder overlaps with the terminal portion of the ureter and interferes with the analysis of ureter activity. Thus, obtaining bladder emptying during the examination is essential(67).

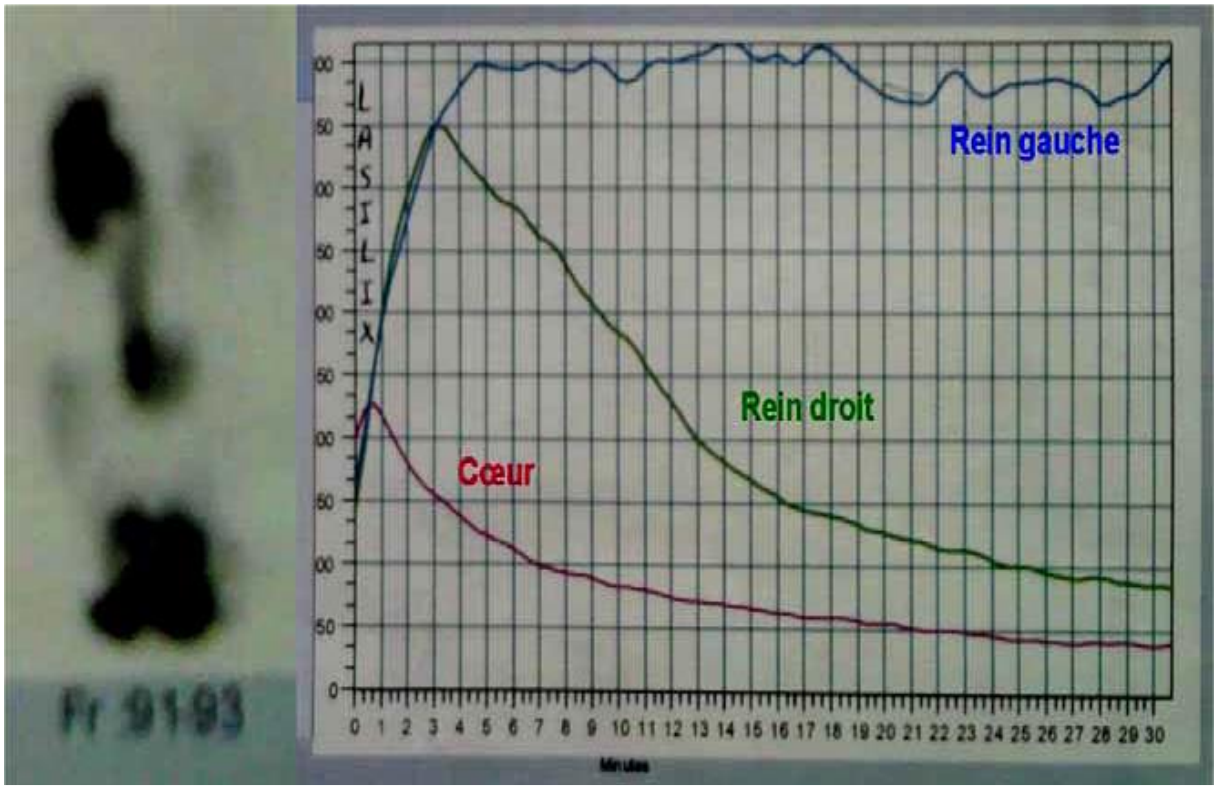

Figure 40: renal scintigraphy (left obstructive ureteropyelocal stasis.

Figure 41: MAG3 in a 3-month-old girl showing isotopic accumulation at the dilated ureter.

e. Intravenous urography [IVU]:

Technique: After the unprepared film is taken, the contrast medium is injected at a rate of 10 ml/s. The early films are taken after 15 to 30 seconds, the secretion film which is imperative must be taken at the 3rd minute, while the morphological films must be repeated between the 5th and 15th minute. Late films should be requested in case of delayed excretion.

Benefit : For some Intravenous urography has a full place in the preoperative workup because it specifies the state of the parenchyma, the type of mega ureter, the abnormality of the uretero-vesical junction and the normality of the lower ureter, and the diagnosis and location of urinary lithiasis (68).

It is interesting to couple IVUS with a furosemide test [IVUS under hyperdiuresis] to confirm or deny the existence of ureteral obstruction in doubtful cases.

IVU remains essential to establish the diagnosis of mega ureter and to assess its impact on the urinary tract.

Limitation: During the last twenty years the place of IVUS for the diagnosis and follow-up of mega ureter has clearly decreased due to the performance of ultrasound and scintigraphy.

The morphological information provided by ultrasound is very often sufficient, whereas renal function and the importance of the obstruction to urine flow are better assessed by scintigraphy(65).

In addition, IVUS is a much more irradiating and time-consuming examination [late films] than renal scintigraphy and requires the use of a potentially allergenic contrast medium.

The interpretation of the results may be hampered by the presence of intestinal gases or by a dilated kidney that does not concentrate the contrast medium well, or in the case of significant alteration of the functional value of the kidney leading to a contrast insufficiency (65).

All these arguments mean that IVUS is no longer a routine part of the initial workup and follow-up of mega ureters.

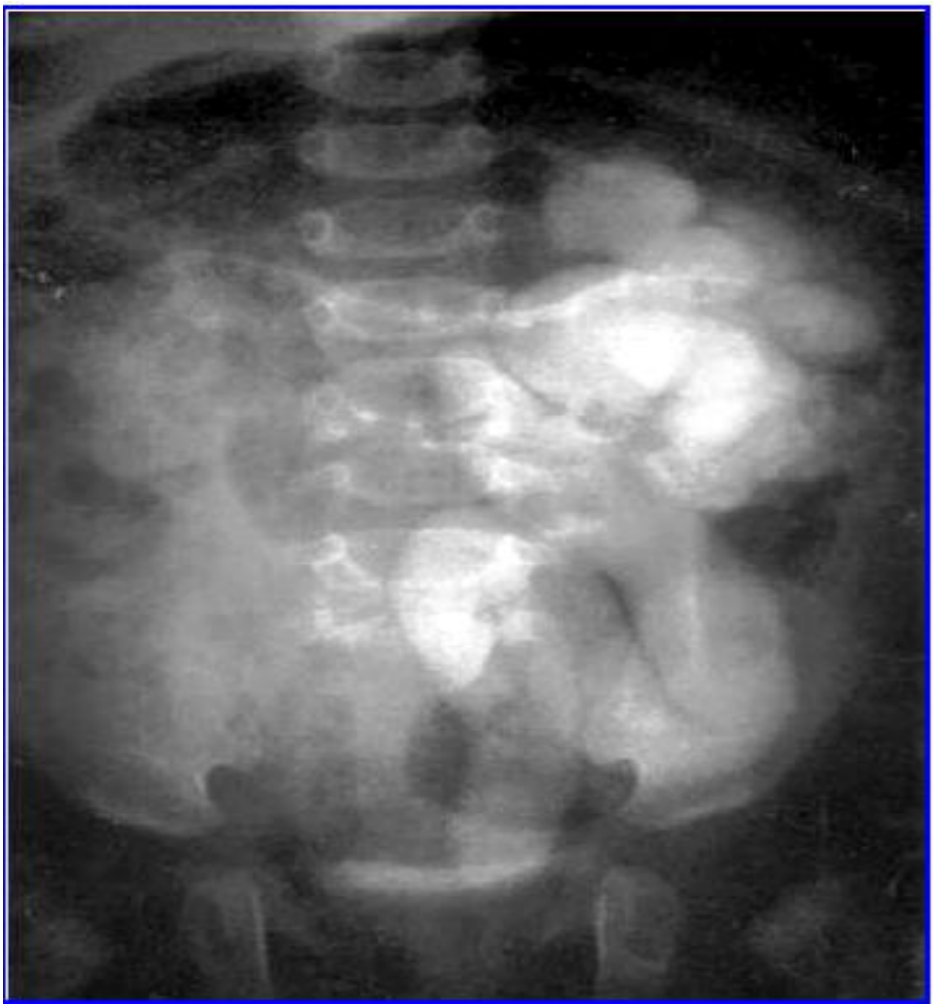

Figure 42: Intravenous urogram showing a tortuous appearance.

f. Magnetic resonance urography [URO-IRM]:

Technique: Magnetic resonance imaging derives from a magnetic signal emitted by the atoms of the organism, under particular conditions of stimulation. Progress in computer science has made it possible to collect information from physical phenomena giving a very weak signal, represented, in the case of imaging, by the magnetic resonance of the atomic nuclei of hydrogen.

Benefit: UROIRM is an excellent modality to obtain an accurate radiological image of the urinary tract. It has the advantage of being non-irradiating, the gadolinium (contrast medium) is not nephrotoxic and the quality of the radiological image is better. In addition, UROIRM is reproducible and is not disturbed by the superposition of digestive gases. In the case of mega ureter, UROIRM allows to recognize ureteral obstruction, to determine its exact location and to evaluate the impact on the upper urinary tract. It easily detects hydronephrosis. It also allows to differentiate the refluxing mega ureter from the non refluxing mega ureter.

Limitation: The very high cost of UROIRM limits its use.

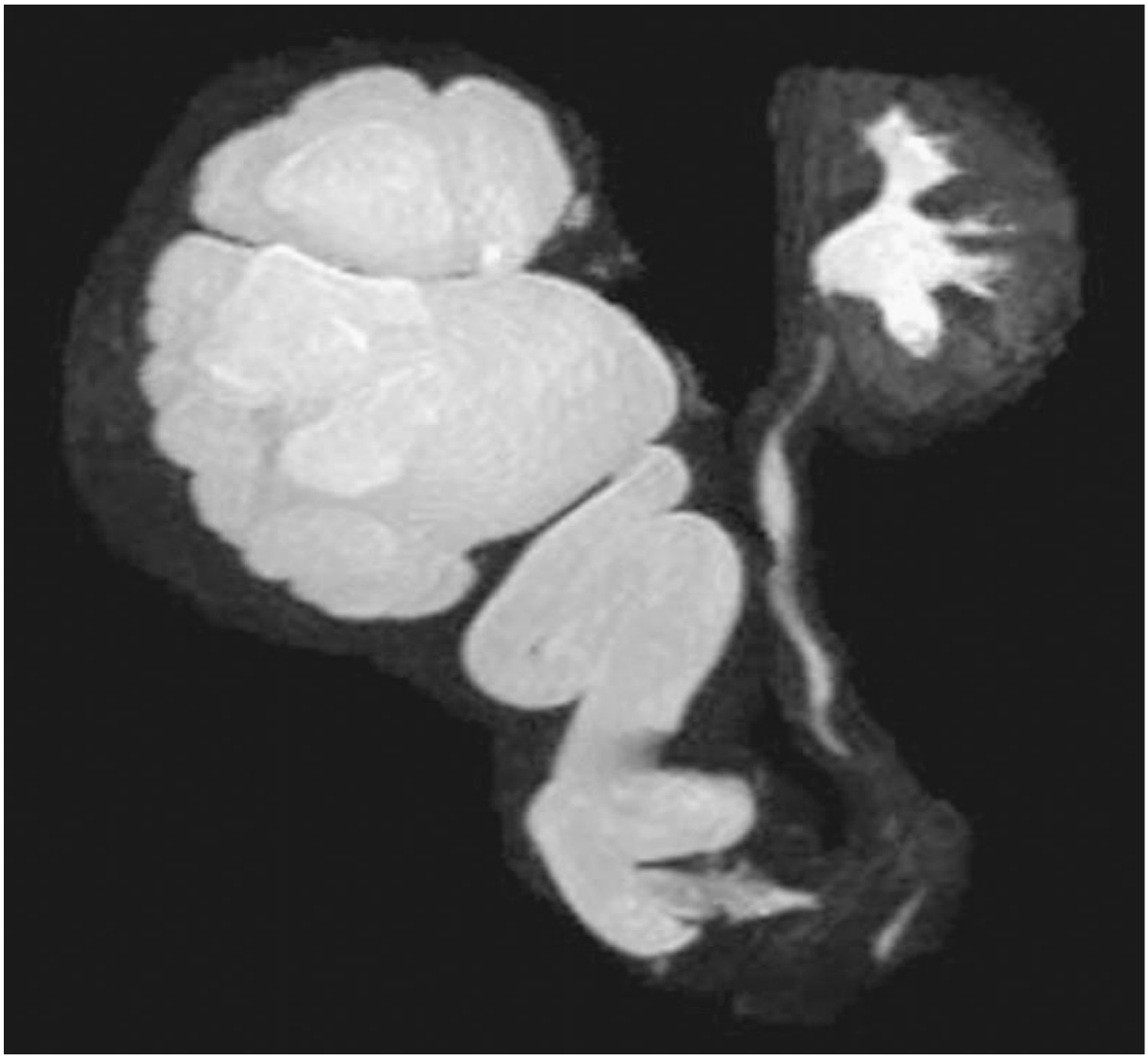

Figure 43: Uro-MRI in a boy with MUP.

2) BIOLOGICAL EXAMINATIONS:

a) Renal function:

The alteration of renal function in obstructive mega ureter in older children is rare, it represents 0% to 10% of patients, it is related to the existence of bilateral mega ureter, mega ureter on single kidney.

It conditions the prognosis of this pathology, hence the importance of exploring the renal function by looking for possible renal insufficiency.

Urea, creatinine and creatinine clearance measurements are used to assess renal function. Creatinine clearance is important for judging the severity of renal failure.

b) Cytobacteriological examination of urine:

Urinary tract infection represents a frequent reason for consultation in MU, it is characterized by the recurrence of infectious episodes, complicating this pathology.

The cytobacteriological examination of the urine is essential to eliminate any urinary infection before starting the radiological investigations that approach the diagnosis. It is also carried out as part of the surveillance from the 3rd month (52,53,69).

The germs most often responsible for urinary tract infection in cases of mega ureter are Gram-negative germs.

The practical conduct in case of discovery of antenatal hydronephrosis or in case of postnatal ureterohydronephrosis can be concluded as follows:

Antenatal Hydronephrosis

(Antenatal ultrasonography: 30thWeeks of amenorrhea):

DAPPR > or = 7mm

Postnatal ultrasound: D1-D3

Cystography (to rule out a secondary cause/reflux)

Scintigraphy > 6 weeks

IVUS / Uro-MRI

VI. DIAGNOSIS:

1) Positive diagnosis:

The diagnosis of mega ureter is a **radiological diagnosis**.

The diagnostic criteria for primary mega ureter is: A dilated ureter seen on:

- Intravenous urogram.

- Pelvic ultrasound.

- URO-MRI.

2) Diagnosis of obstruction:(43)

Obstruction is the inability of the ureter to ensure an increase in urinary flow other than by increasing the basal pressure, unlike stasis where the increase in diuresis does not lead to any increase in basal pressure. Here we find the classic pressure/flow relationship which in urodynamic sense defines any obstruction.

The different examinations used for the diagnosis of obstruction use hyperdiuresis to unmask the obstruction

- **Renal and pelvic ultrasound under diuretic conditions:** (70,71)

It allows to look for indirect signs in favor of a severe obstruction that can lead to or increase an alteration of the renal function:

- Hyper-echogenicity of the renal cortex with decreased cortico-medullary differentiation.

- Thinning of the parenchyma, small kidney size

- Compensatory hypertrophy of the contralateral healthy kidney

- **IVU under hyperdiuresis:**

The delay in excretion is as much related to stasis in dilated cavities as to obstruction. Stasis is normally relieved by the furosemide test, whereas obstruction will lead to an inability of the excretory pathway to ensure a high flow rate, which will result in an increase in the volume of the collecting system. To demonstrate this obstruction, 40mg of furosemide is injected 20 minutes after the beginning of the examination. An obstruction is considered to exist when the percentage increase in the excretory tract, 15 minutes after the injection of diuretic, exceeds 22%, and there is no obstruction if this increase in dilatation remains below 15%.

- **Dynamic scintigraphy:**

Obstruction is characterized by delayed elimination of a radioactive tracer under furosemide-induced hyperdiuresis. Results are obtained in the form of curves reflecting urinary elimination. Obstruction is considered to occur when furosemide-induced activity increases even after injection of the diuretic.

3) **Differential diagnosis:**

❖ Reflux:

Reflux is defined as the abnormal retrograde passage of bladder urine into the ureter. Physiologically, there is an anti-reflux system at the uretero-vesical junction, based on the submucosal pathway of the ureter and the trigonal musculature.

Depending on the cause, a distinction is made between primary refluxes, due to an insufficiency of the physiological anti-reflux system, and refluxes secondary either to a local malformation (duplicity, ureterocele, diverticulum), or to an organic subvesical obstacle (urethral valves) or finally to a functional vesico-sphincter disorder (neurological bladder, bladder instability).

Primary refluxes frequently disappear with growth, by maturation of the uretero-vesical junction.

Reflux of sterile urine has no consequences for the kidneys, but the reflux of infected urine into the kidneys can lead to acute pyelonephritis, which may leave permanent sequelae (chronic pyelonephritis, reflux nephropathy) Epidemiologically, age and sex are important: boys often have significant reflux, discovered early, before the age of one, whereas after the age of one, reflux clearly predominates in girls, often in less significant forms than in boys. Overall, the prevalence at birth is estimated at 0.5% of newborns. In children explored for urinary tract infection, it can reach 50%.

Urinary tract infection and antenatal diagnosis are the usual circumstances of discovery. Ascending lumbar pain is only seen (rarely) in older children. When questioning, one should always look for continence or micturition abnormalities in order not to ignore a functional vesico-sphincter disorder; bladder instability is very often associated with reflux.

Retrograde cystography is the key examination. It is the only way to confirm the reflux and to classify it. It should be done at least one month after a urinary infection. Ultrasound is mandatory, it allows to assess the renal impact.

Intravenous urography is no longer of much use, except if reflux secondary to duplicity is suspected.

DMSA renal scintigraphy is the reference examination to assess the existence or not of renal sequelae.

Primary refluxes of low grade (1 and 2) disappear spontaneously with growth in three quarters of cases, compared to only one quarter of cases for primary dilating refluxes (grades 3 and higher). Secondary refluxes rarely regress without surgery, except in cases of bladder instability where medical treatment of the functional disorder often makes the reflux disappear.

Reflux-related renal damage is permanent and may sometimes progress on its own to renal failure and/or hypertension, even after the reflux has disappeared.

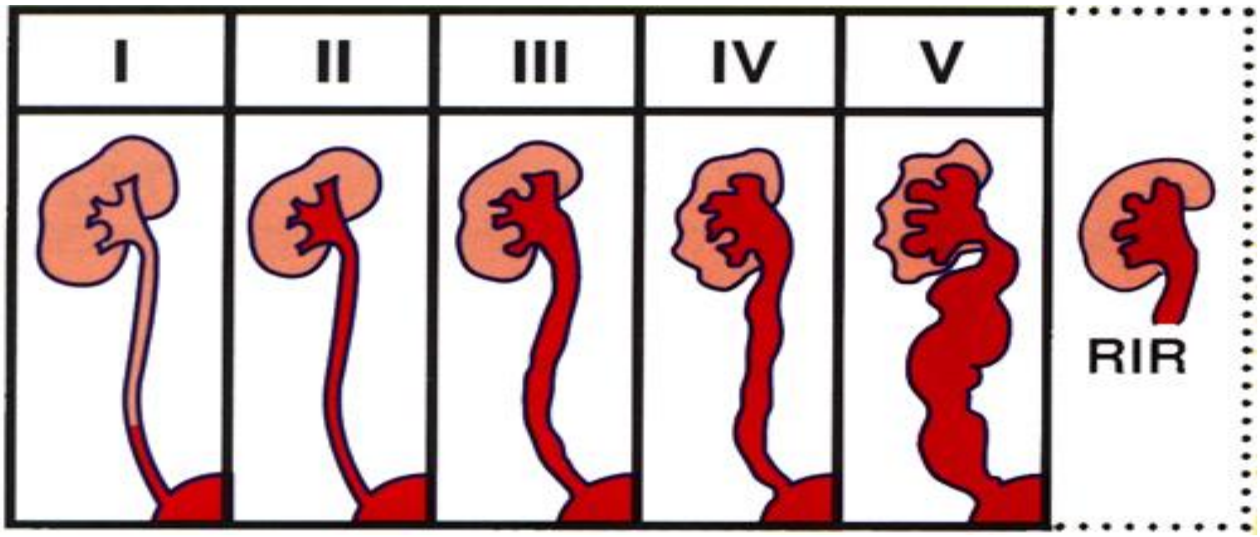

Figure 44: International classification of reflux according to Duckett-Bellinger.

Grade I: reflux into the pelvic ureter.

Grade II: ureteropyelocele reflux without dilatation.

Grade III: idem with moderate dilatation of the ureter.

Grade IV: global uretero-pyelocecal dilatation, but the papillae remain marked.

Grade V: important dilatation, tortuous ureter, ball-shaped calyces.

Intra-renal reflux (IRR) is not taken into account in this classification. It is an additional severity factor.

❖ Valve of the posterior urethra:

Posterior urethral valves (PUV) are the most common obstructive uropathy in boys (72). It is a serious malformation because it can lead to end-stage renal failure by destruction of the renal parenchyma.

The seriousness of this malformative uropathy lies in the importance of its repercussion on the upper urinary tract with a significant risk of end-stage renal failure (25 to 40% of cases), and 1% of children awaiting renal transplantation are carriers of PUV(72).

Posterior urethral valves (PUVs) are the most common obstructive uropathy of the lower urinary tract. Its incidence varies from one in 8,000 to one in 25,000 live births (72). It consists of an abnormal membrane that radiates forward from the veru montanum to the membranous urethra. This valve has a small posterior opening so that during micturition the fused part bulges forward. It develops early, during intrauterine life, thus impeding the normal development of the kidneys and bladder.

With the increasing use of prenatal ultrasound, most patients with this malformation are identified early in intrauterine life with uni- or bilateral dilatation of the upper urinary tract, a large bladder

with a thick wall and poor emptying, and sometimes a dilated posterior urethra. Oligohydramnios and a cortical hyperechoic appearance may be associated with dysplastic lesions. Ultrasound thus allows the antenatal diagnosis of UPV, but also to evaluate its impact by studying the quantity of amniotic fluid, the appearance of the kidneys and by taking fetal samples. The latter are taken in fetal blood and/or fetal urine (renal pelvis) and allow the determination of microglobulin, the protein that best reflects fetal renal function.

From the second trimester onwards, several situations can be distinguished according to the amount of amniotic fluid and its evolution, and, to a lesser extent, the ultrasound appearance of the kidneys.

Indeed, in the presence of an amniotic fluid, the prognosis is poor since it reflects an absence of renal filtration and raises fears of the appearance of pulmonary hypoplasia. Fetal urine sampling is not essential, but can sometimes help with prenatal counseling or if the question of medical termination of pregnancy is discussed.

If the fluid is not abundant for the term, and regardless of the appearance of the kidneys, fetal sampling is of most interest. Prenatal counseling will depend on the microglobulin level and possibly its evolution.

❖ **Ureterocele:**(73,74)

Ureterocele is a pseudo-cystic dilatation of the distal portion of the ureter in its trigonal submucosal course, which lacks sufficient musculature and thus becomes distended forming a rounded lacuna in the bladder. The two most frequent types of ureterocele are opposed point by point: the ectopic ureterocele, developed at the termination of a superior polar ureter of ureteral duplication, and the orthotopic ureterocele at the termination of a simplex ureter.

❖ **Ectopic implantation of the ureter:**

Ectopic abruption is the second pathology that can involve the upper pyeloneal ureter with ureterocele in the context of ureteral duplications. Like ureterocele, ectopic abruption can also involve a simple non-duplicated ureter.

By definition, this is a ureter that does not terminate at the level of the bladder trigone: in girls, it may be a termination below the bladder neck with a picture of urinary incontinence (distal urethra, vagina, rectal vestibule, etc.) and may also be associated with a cyst of Gartner's duct, a remnant of Wolff's duct. In boys, the ectopic ureter terminates in the urogenital system above the external sphincter and perineum and usually in the Wolffian structures such as the vas deferens, seminal vesicles, ejaculatory ducts or even the posterior supramontane urethra.

The revealing picture is then not one of incontinence but of infection or pain in the organs concerned (orchiepididymitis).

In 80% of cases, the ectopic ureteral outlet involves a duplicated ureter, and in this case it is the ureter of the upper pyelo. In 20% of cases, the ectopy concerns a simple ureter, whether it is unilateral or bilateral.

It can be associated with certain malformations such as homolateral renal malformations in case of simple ureter (ectopy, horseshoe kidney) or contralateral (ureteral duplication), epididymotesticular anomalies, anovagino-uterine malformations (vaginal duplications, hemivagina, bicornuate uterus, etc.) and anorectal malformations, as well as esophageal atresia.

❖ **Prune belly syndrome:** (75)

Prune Belly syndrome (PBS) or Eagle-Barrett syndrome is the association of a muscle deficit of the abdominal wall, dilatation of the renal pelvis associated or not with renal dysplasia, absence of prostate tissue and cryptorchidism. Persistence of the urachus is possible. Uropathy is dysfunction or obstruction of the urethra causing megavessia and mega-ureters. Hypoplasia of the prostate is common in boys. This sequence is much rarer in girls (2% of patients) where it combines uropathy and abdominal wall anomaly with vaginal or rectovaginal atresia, rectovesical fistulas or a bicornuate uterus. The prognosis depends mainly on the severity of the renal involvement. The complexity of urinary tract malformations means that conservative treatment remains an important part of the therapeutic arsenal. Surgery for urinary malformations requires a case-by-case approach (both for the indication and the timing of surgery) and should be performed by an experienced team. Testicular lowering should be performed more frequently in the neonatal period to increase the chances of paternity, as should abdominoplasty, which has real aesthetic and functional benefits.

VII. Treatment:

Since its development in 1974, antenatal ultrasound has profoundly changed the treatment of mega-ureters. Indeed, the frequent observation of spontaneous resolution **(80% of cases)** after a few months or years of surveillance has led to the proposal of conservative treatment in the most of cases. (76)

Surgical treatment is then proposed in case of failure of the conservative treatment. Its indications are:

- Recurrence of symptoms (urinary tract infection, pain) despite adequate antibiotic prophylaxis.
- Ureteral stones.
- Deterioration of kidney function.

1) Conservative therapy:

It's based on **Surveillance** and **Antibiotic prophylaxis;**

A. Surveillance:(68)

The objective of this surveillance is to act before the alteration of the renal parenchyma and not to act on children who would have spontaneously evolved towards recovery. Surveillance is organised through rigorous clinical, ultrasound and, if necessary, scintigraphic monitoring.

It will be more frequent during the first year of life due to the spontaneous regression of the majority of forms during this period but also because of the high frequency of appearance of urinary infections. Parental education about the risk of infection is essential.

B. Antibiotic prophylaxis:

Antibiotic prophylaxis is indicated in cases of febrile urinary tract infection and is prescribed routinely for the first six months of life, or even the first year if the dilation is important. (77)

The basis of antibiotic prophylaxis:

- ✓ Be active on E. coli, the main bacteria causing recurrent UTIs.
- ✓ Be orally administered and well tolerated.
- ✓ Have predominantly urinary elimination.
- ✓ Be different from those suggested for curative treatment: if an infection occurs, the same antibiotic cannot be used as a curative treatment because the bacteria involved are likely to be resistant to it.
- ✓ Have a minimal ecological effect on the digestive flora.

The two most commonly used antibiotics are **cotrimoxazole** and **nitrofurantoin;**

- Cotrimoxazole (contraindicated before 1 month of age) is used at a dosage of 1-2

mg/kg/day of trimethoprim and 5-10 mg/kg/day of sulfamethoxazole

- Nitrofurantoin at a dosage of 1-2 mg/kg/day.

These are prescribed once daily, at the time of maximum osmolar concentration in the urine for better effectiveness, in the morning for children under 2 years of age, or in the evening for the others.

The optimal duration is a unanimous:

- 6 to 12 months.
- Until the dilation disappears.
- Until the age of 5 years.

C. Surgical indications and criteria for failure of conservative treatment:

- Relative renal function <40%.
- 5% loss on follow-up scans.
- Clinical complications: recurrent or severe UTI.
- Persistence of dilatation >15mm beyond the first year of life.*

D. Circumcision:

It is indicated in boys with Megaureter who have been treated by conservative treatment. It has the advantage of significantly reducing the rate of urinary tract infections according to the Braga study with a rate of 19% of UTI in circumcised patients against 41% of UTI cases in uncircumcised ones.

2) Surgical treatment: (68)

A. Temporary diversions:

a) Indications:

This treatment option was for a long time the first-line technique (1960s-1970s). Nephrostomy or ureterostomy was performed in the case of any very dilated and symptomatic mega-ureter. This type of treatment is still valid in an emergency in case of septic choc by retention of infected urine at the level of the ureter.

The indication for a derivation can also be given in the case of major dilatation in infants to facilitate reimplantation (78).

The recent consensus conference of the British Association of Pediatric Urologists (BAPU) recommends percutaneous nephrostomy as the first line of treatment, followed by the double J (79). Cutaneous ureterostomy is progressively being considered as a last resort because of the morbidity of the procedure.

b) <u>Methods:</u>

- Nephrostomy.
- Ureterostomy.
- Non-continental ureterovesicostomy.
- double j ureteric stent.

B. Curative treatment:(80)

a) Open surgery:

Definitive treatment consists of **excising the obstructing distal segment and reimplanting the ureter into the bladder** by a technique which facilitates good upper tract drainage without permitting vesicoureteric reflux. However, these twin goals may be difficult to achieve, particularly in infants with grossly dilated ureters and relatively small bladders.

Wherever possible, reimplantation of a megaureter should be avoided in the first year of life, because of these technical difficulties and growing anecdotal evidence that dissection in the region of the distal ureter and bladder neck can result in a degree of neurological impairment, with a possible impact on continence in later childhood.

b) Alternatives to open surgery in the first year of life:

Successful medium-term drainage with a paediatric in-dwelling ureteric JJ stent has been reported from a number of centres. If endoscopic insertion is not feasible, open cystostomy, dilatation of the ureteric orifice and insertion of a JJ stent is well tolerated. The stent can usually be safely left in situ for up to 6 months, after which time it may be feasible to proceed to definitive surgery. However, it is apparent that, in some infants, the combination of dilatation and prolonged in-dwelling stent drainage is sufficient to correct the obstruction without the need for further surgery. Although upper tract diversion by terminal or ring ureterostomy was once widely practised in infants with obstructed megaureter, there are now very few indications for this approach. Ureteric reimplantation is best deferred until 12 months of age upwards, but, depending on the circumstances, may be considered from 6 months upwards, according to the anatomical features and the experience of the surgeon.

c) Surgical treatment from 1 year of age upwards:

- **Moderate dilatation (ureteric diameter approximately 1 cm or less):**

In these cases, the ureter can be mobilised intra-vesically, as for a conventional ureteric reimplantation, and the ureter reimplanted in a conventional **Cohen cross-trigonal tunnel** after excision of the stenotic distal segment.

❖ **Severe dilatation:**

The dilated ureter is identified extra-vesically and mobilised down to the bladder, where the stenotic segment is identified and excised. In older children, it may be feasible to reimplant the ureter via the original hiatus, but because of the difficulty in creating the optimum ratio of ureteric width to length of submucosal tunnel required to prevent reflux, plication or tapering of the distal ureter is usually advisable. Where the ureter is grossly dilated, plication or tapering of the distal ureter is combined with **a Politano–Leadbetter reimplantation,** which creates a longer anti-reflux submucosal.

A psoas hitch procedure is also performed to anchor the bladder to the psoas muscle in the region of the entry point of the ureter into the bladder. This manoeuvre minimises the risk of kinking and obstruction during bladder filling. Although tapering "remodeling" of the distal ureter has been claimed to restore ureteric peristalsis, this is very doubtful. Plication by **the Starr or Kalicinski technique** is more straightforward but creates a bulkier ureter and, theoretically, a greater risk of postoperative reflux. The presence of bilateral obstructed megaureters represents an indication for **transureteroureterostomy**, in which one ureter is reimplanted using **the Leadbetter–Politano** procedure and **psoas hitch** while the contralateral ureter is disconnected from the bladder, rerouted across the midline and anastomosed to the reimplanted ureter.

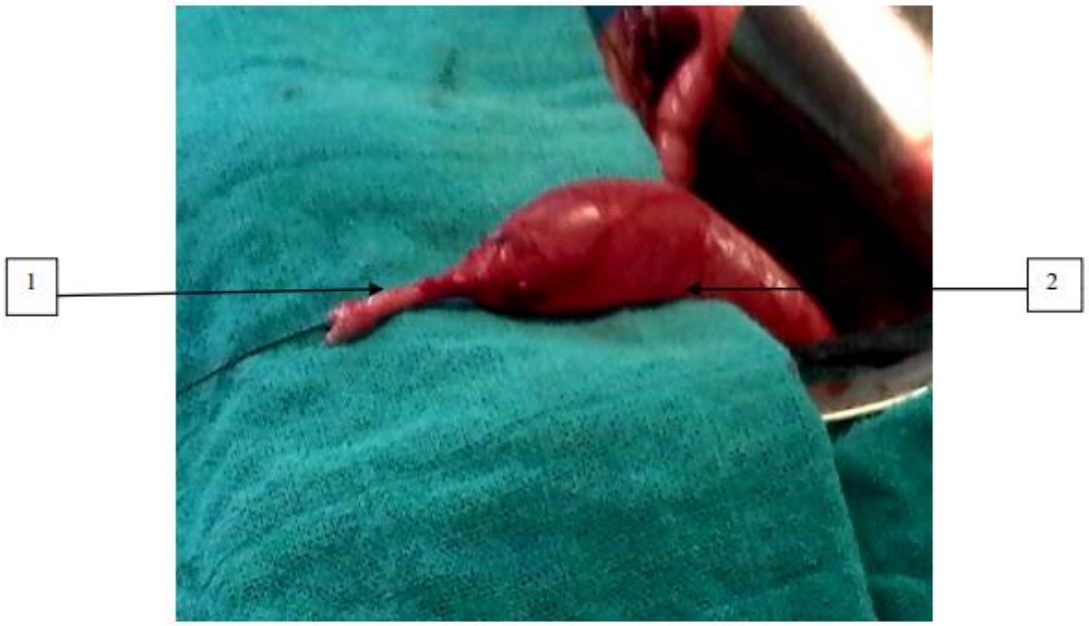

Figure 45: intraoperative photo showing a radish tail appearance: a non-functioning area is shrunken (1) and a dilated area (2).

d) Techniques:

❖ Cohen's technique:

- Unilateral or minimally dilated ureter, with sufficient length of ureter to pull it through its hiatus without traction and sufficient trigone width to accept the transverse submucosal tunnel.
- It is avoided in cases of bilateral involvement because the presence of a retro-trigonal bar formed by the two transverse submucosal tunnels may disturb bladder function.

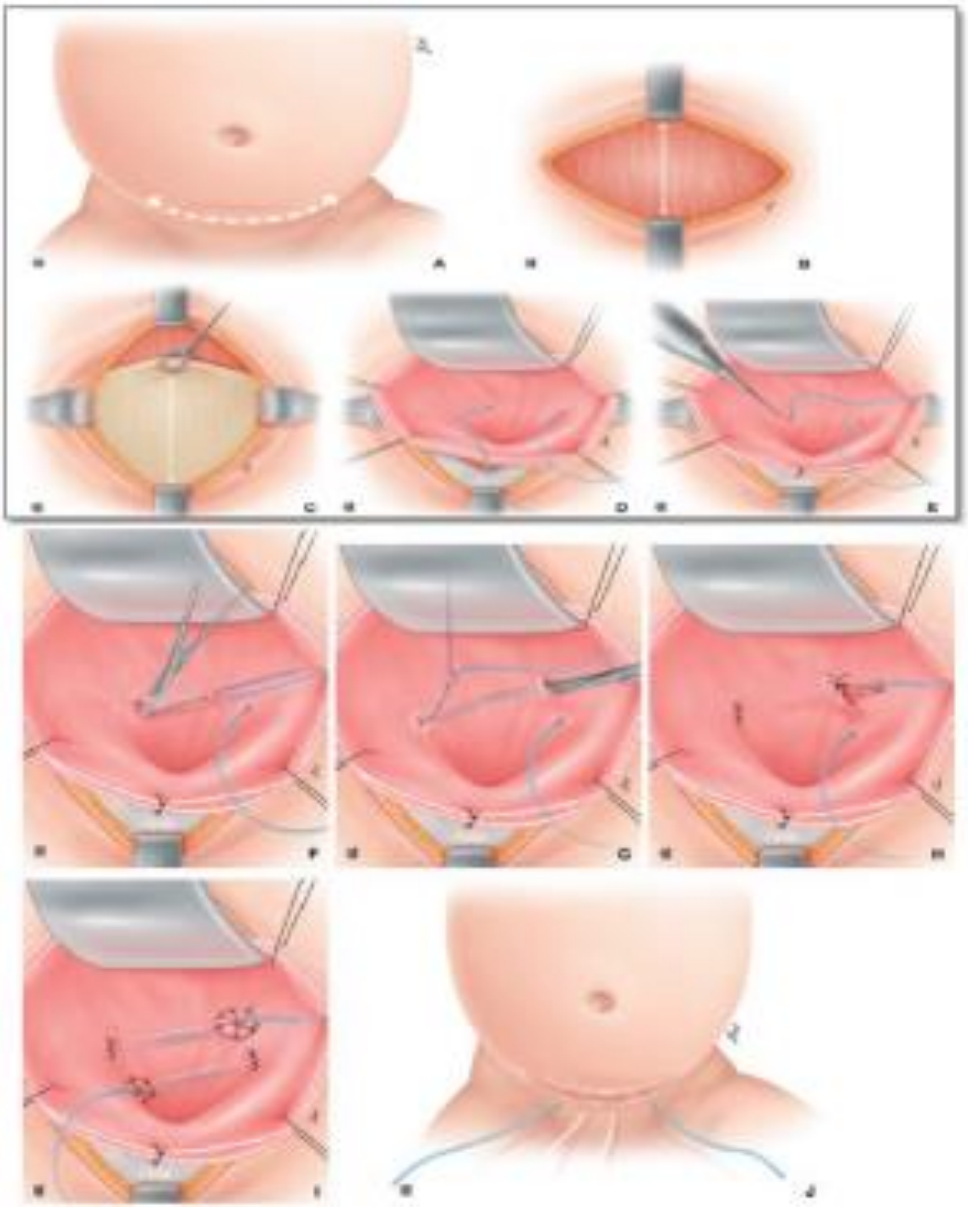

Figure 46: Ureteral reimplantation according to Cohen

A.Skin incision layout. B.Layout of the fascia incision C.Bladder opening D.Location of the ureters by catheterisation.

E.Incision of the right ureteral meatus flange F.Ureteral dissection. G.Making the submucosal pathway.

H.Right ureteral reimplantation - meatus docking and closure of the right ureteral hiatus I.End of procedure J.Skin closure and draining.

❖ Politano-Leadbetter technique:

- It is preferred in cases of bilateral involvement or a very dilated ureter.

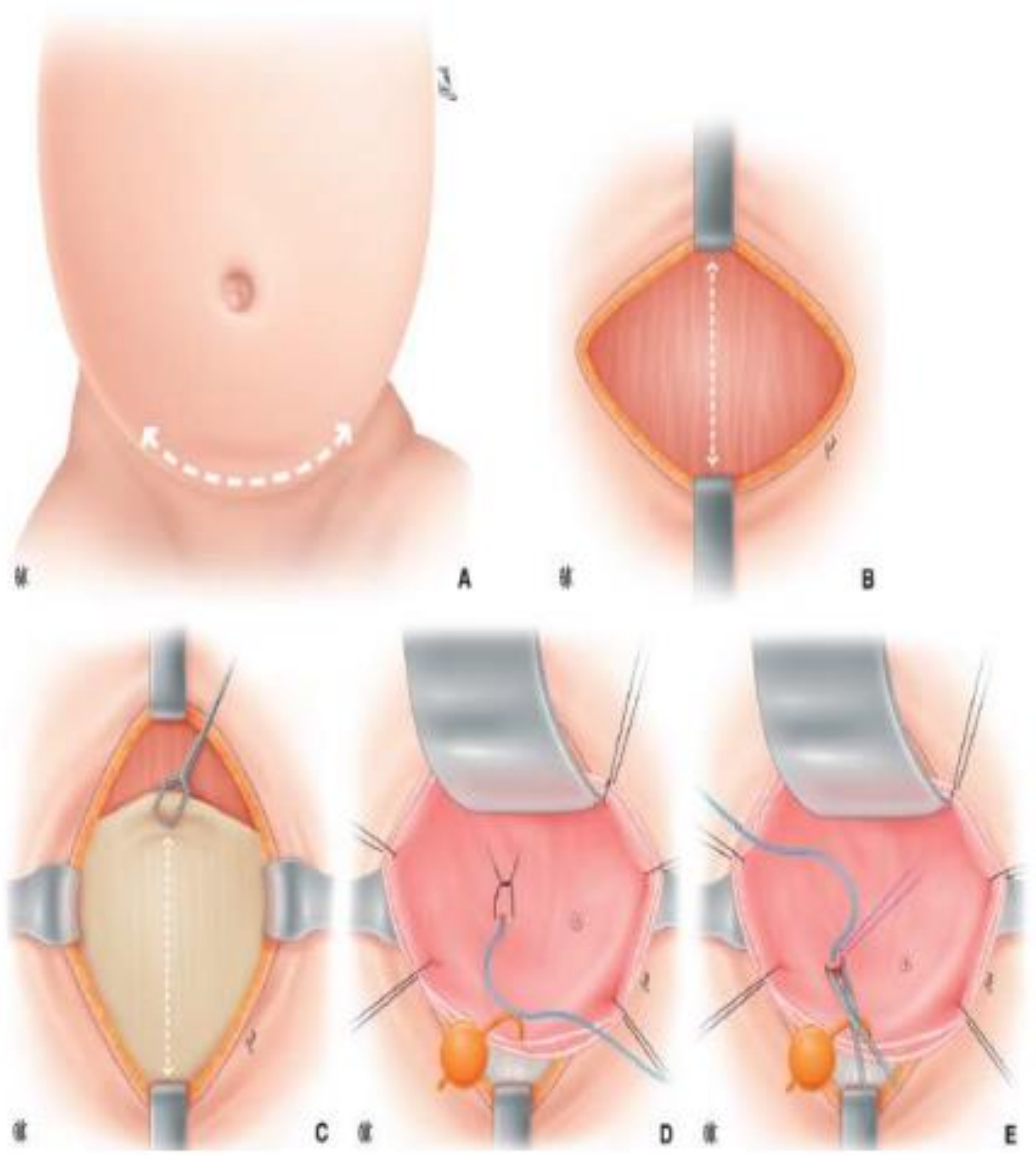

Figure 47: Ureteral reimplantation according to Politano-Leadbetter

A.Skin incision layout B.Layout of the fascia incision.

C.Bladder opening D.Location of the ureters by catheterization

E.Incision of the right ureteral meatus and start of dissection of the ureter.

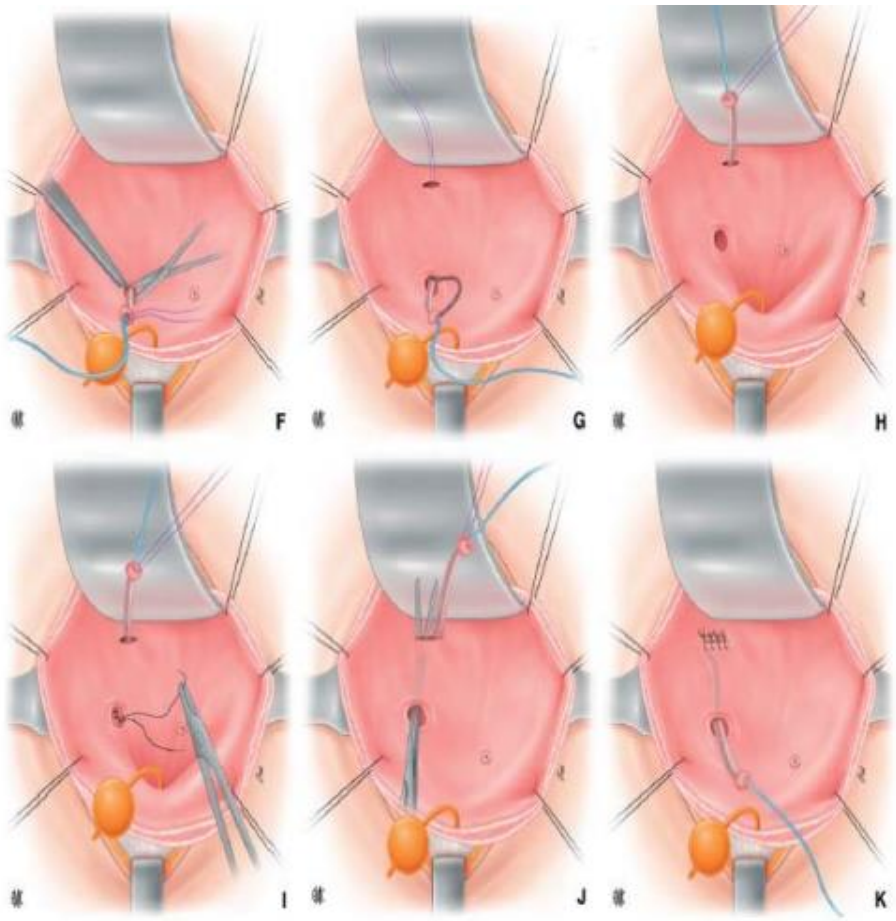

Figure 48 : Ureteral reimplantation according to Politano-Leadbetter;

F.Dissection of ureter. G.Suprahiatal counter-incision

H.Reintroduction of the ureter through the neo-orifice. I.Closure of the old ureteral hiatus.

J.Making the submucosal pathway.K.Right ureteral reimplantation - mucosal closure of the neohiatus.

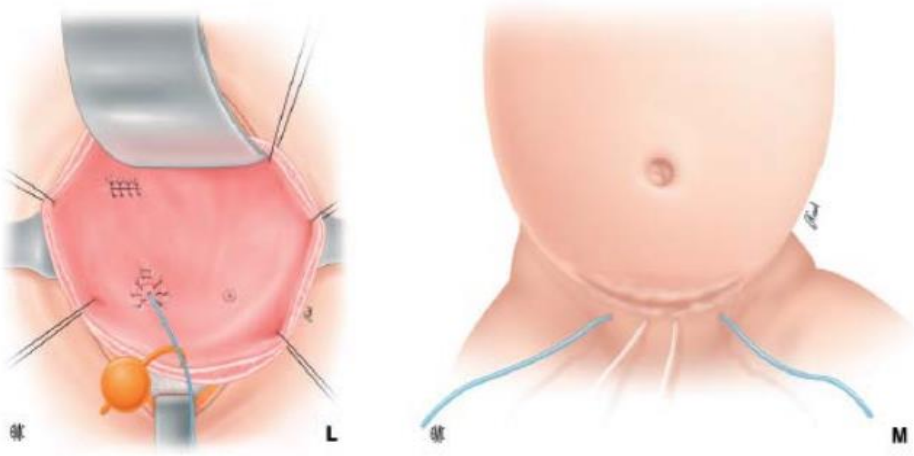

Figure 49 : Ureteral reimplantation according to Politano-Leadbetter

L.Docking the meatus M.Skin closure and draining

❖ A psoas hitch:

- It can help in cases of significant loss of substance (important resection of the dilated ureter or ureterostomy) to allow reimplantation without traction.
- In the case of mega-dolicho-ureters.
- Re-operation for secondary stenosis
- It can also be used on one side in case of unilateral damage.
- Can only be applied to vertical reimplantations, Politano -Leadbetter type.

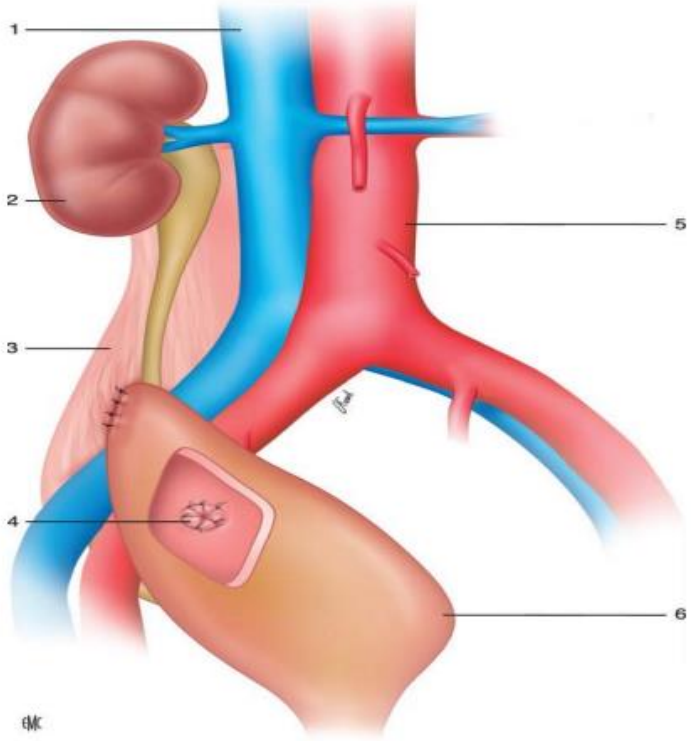

Figure 50 : A psoas hitch:

1. Inferior vena cava 2. Right kidney 3. Psoas muscle

4. Ureterovesical junction 5. Abdominal aorta 6. Bladder

e) Mini-reimplantation:

The mini-reimplantation performed by **Ramesh Babu** (81) consists of dissecting the terminal part of the ureter after opening the bladder. then the narrowed segment and the very dilated part of the ureter will be resected (about 3-5 cm). Support to the ureter is provided by the closure of the detrusor behind. No attempt to remodel was made and the ureter is re-implanted in its original position by separated points without a sub-mucosal pathway. The bladder mucosa is closed in a vertical direction to the new ureteral meatus and a JJ stent is left in place for 6 weeks.

The aim of this technique is to operate on children under 1 year old.

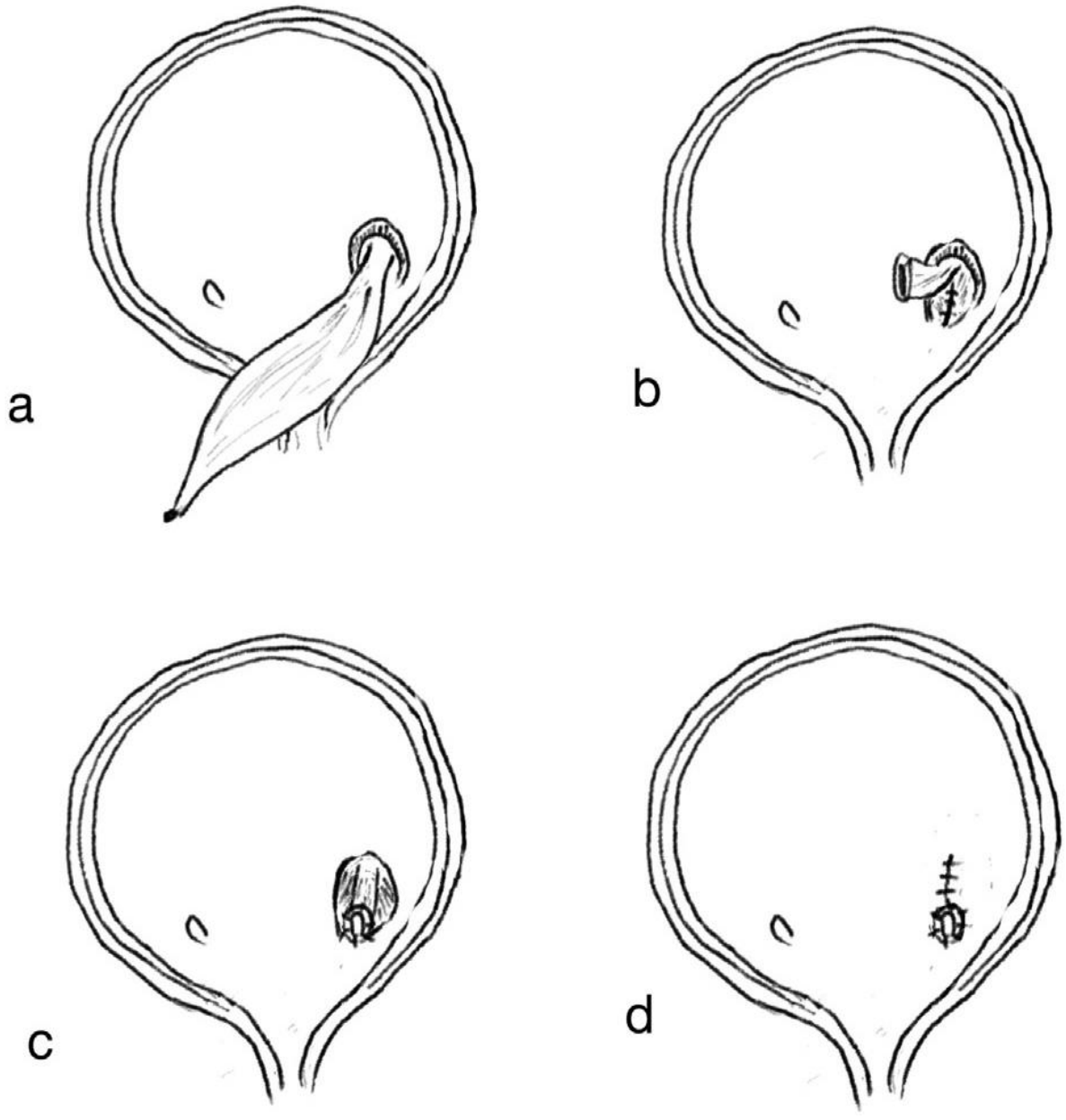

Figure 51: Mini reimplantation: (a) removal of the distal ureteral portion; (b) detrusor closure behind the ureter; (c) reimplantation of the ureter (d) closure of the bladder mucosa.

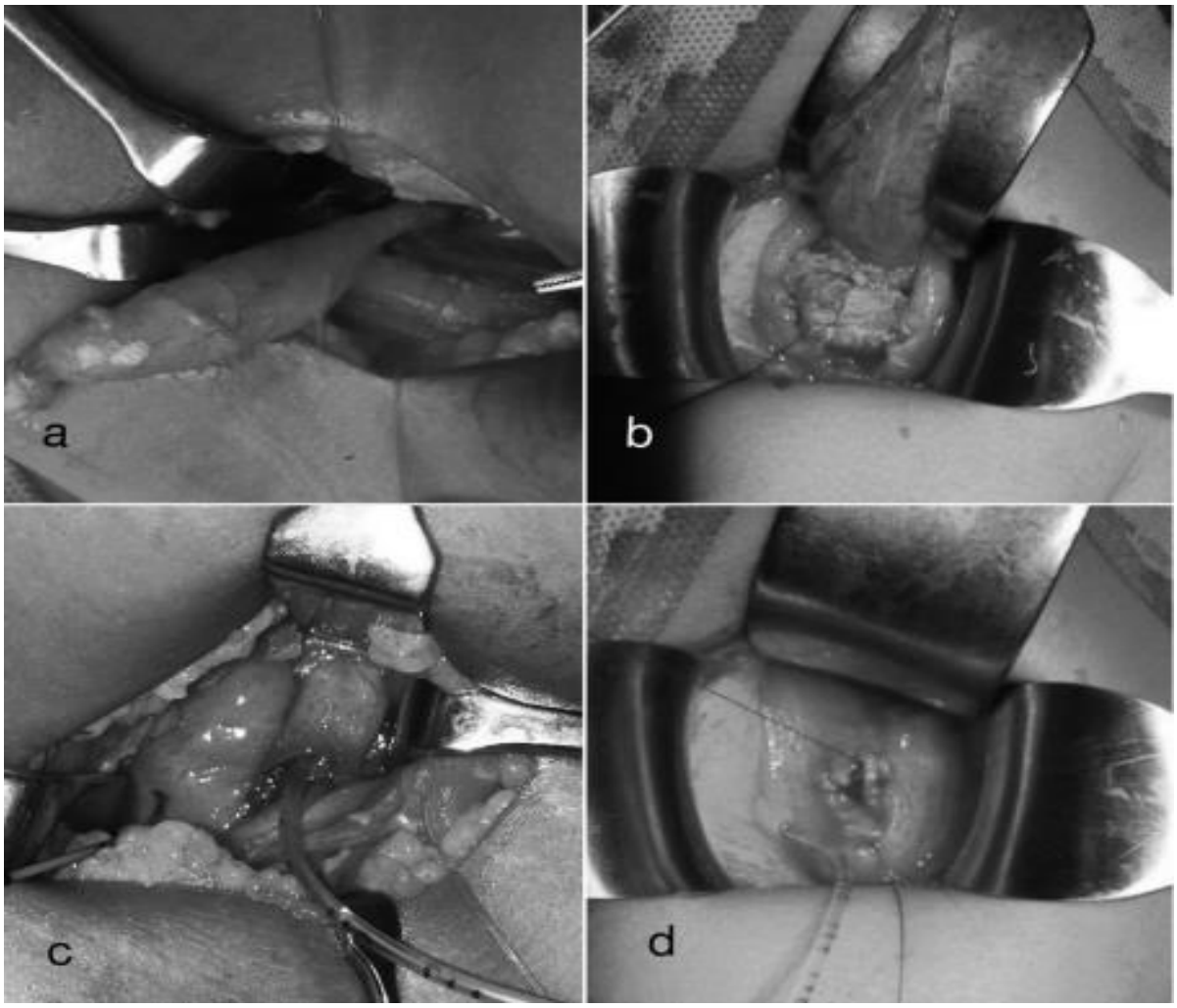

Figure 52: steps of the mini-reimplantation; (a) After opening the bladder, the MUP is dissected; the Distal narrow segment and coarsely dilated distal segment (approximately 3-5 cm) are excised. (b) detrusor closure behind the ureter.(c) the ureter is reimplanted to its original position by stitches separated at PDC 6/0 without submucosal trajectory or crossing or remodeling. (d) The bladder mucosa is closed in a cranial direction up to the new ureteral meatus.

f) Modelling:

❖ **Hendren's technique:**(82)

It consists of a resection of a longitudinal ureteral strip at the level of the "anti-mesenteric" edge of the ureter, followed by a longitudinal suture. This method allows thinner ureters to be obtained but presents a danger for the vascularisation in case of excessive resection.

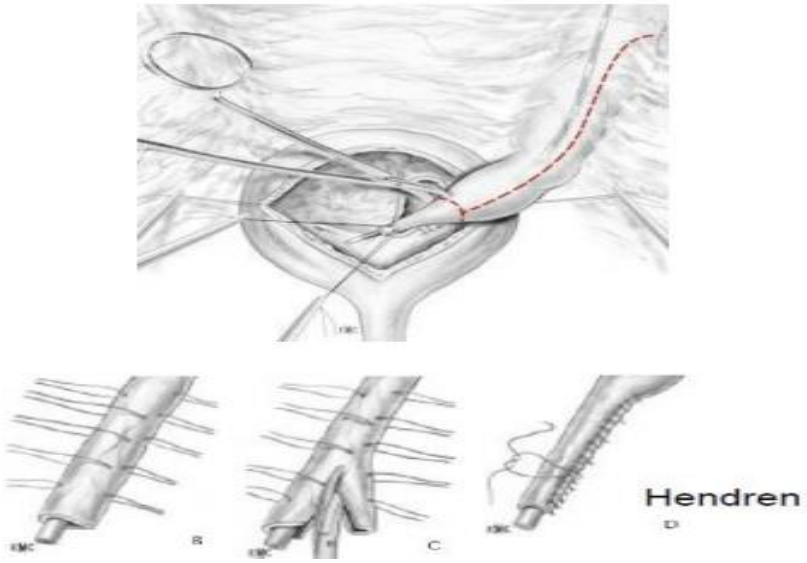

Figure 53: Hendren's Modelling.

❖ **Kalicinski and Starr's technique:** (83,84)

It consists of a paletot plication of the ureter. This method respects the vascularisation but gives wider ureters.

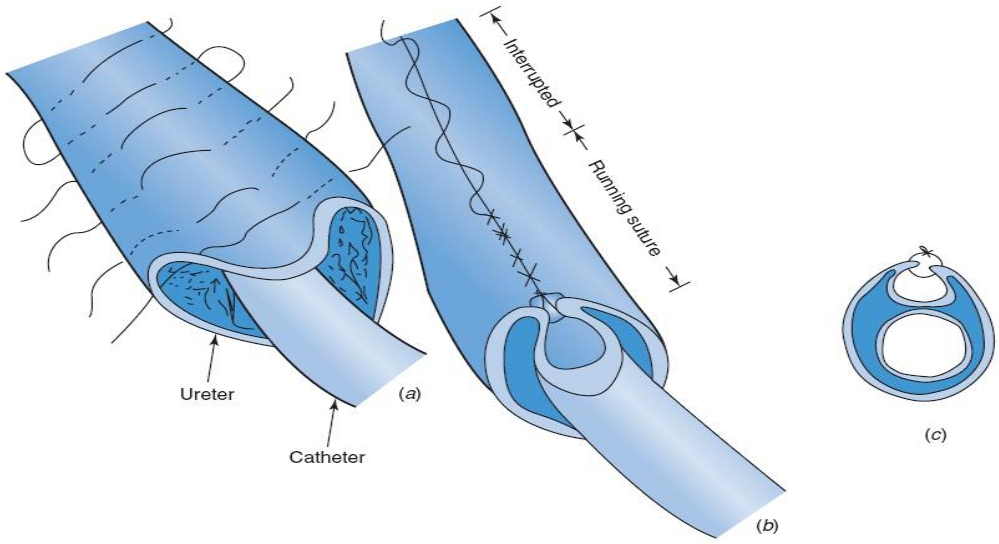

Figure 39.8 Starr technique of ureteral imbrication. (*a*) The ureter is folded inward over a 10 or 12 Fr catheter as a guide. (*b*) The distal portion is secured with interrupted sutures to allow for transection without compromising the running suture line. (c) Cross section of (b).

Figure 54: Starr Technique of ureteral imbrication.

g) Post-Surgery Drain:

A ureteral catheter is left in place for 4 to 12 days depending on whether or not modelling is performed.

h) Complications of open surgical treatment:(83,85)

1. Immediate:

- Uretero-vesical fistulas which may be expressed secondarily and require revision surgery.
- Stenosis of the re-implanted ureter due to postoperative oedema can be corrected by temporary drainage, nephrostomy or double J Stent.
- Problems with the removal of drains can occur.

2. **Late**:

- The para-ureteral diverticulum.
- Secondary reflux linked to a submucosal pathway that is too short or to retraction of the of the ureter.
- Organic stenosis secondary to ischaemia in most cases by non-respect of the ureteral vascularization.

C. Endoscopic treatment:

Today, double J tube placement is widely accepted as the first line of treatment in infants, although it is not clear whether the key to success is the dilation of the urethral meatus or prolonged use of the double J catheter (79).

It was in the 1990s that double J catheter treatment was first attempted as an alternative to the classic ureterostomy, to temporarily relieve the obstruction (Ransley). In addition to having many advantages such as the absence of stomy and the preservation of a functional bladder, this treatment has led to a regression of the obstruction in many cases, obviating the need for subsequent reimplantation. These results have been confirmed by several series with a success rate ranging from 50 to 70% and a morbidity ranging from 30 to 70% .

Balloon dilatation of the ureterovesical junction and endoureterotomy alone or in combination are more recent and show equivalent results in many small retrospective series, but with a risk of stenosis not reported in other series.

1. Indications:

Endoscopic treatment of the mega-ureter with the double J catheter is a mini-invasive technique that allows internal drainage of urine in symptomatic children with obstructive mega-ureter.

The three factors we consider when proposing endoscopic treatment of megaureter are:

The occurrence of complications (severe febrile urinary tract infections or stones).

The obstructive and non-refluxing nature of the mega-ureter with an appearance of ureteritis on ultrasound during fever episodes (diameter of ureteral dilatation on ultrasound greater than 8 mm).

A frankly obstructive emptying curve or an alteration of the renal function between two scintigraphic controls.

2. Technique:

Under general anaesthesia, in the gynaecological position, the surgical procedure begins by an exploratory cystoscopy with location of the two ureteral meatus. Once the ureteral orifice has been located, a hydrophilic guide is inserted, followed by a ureteral catheter to perform a retrograde ureteropyelogram which confirms the diagnosis. Thereafter, ureteral dilators adapted to the age of the child are fitted to the guide in succession (6-8 CH).

At the end of the procedure, the double J catheter adapted to the child's age (3 to 7CH) is then placed on the hydrophilic guide and under fluoroscopic control. The choice of the catheter can sometimes lead to the use of "blue stent" type catheters, of which is crossed out. The cystoscope is used as a pusher for the double J stent and allows the positioning of its lower end to be checked.

A final fluoroscopic check verifies the correct position of the two ends of the double J stents. Bladder drainage is not systematic, unless there is significant haematuria intraoperatively. Intraoperative retrograde cystography is generally not indicated.

D. Uretero-nephrectomy:

It should be reserved for destroyed kidneys and should involve total removal of the ureter and kidney. This solution can only be considered if the contralateral kidney is healthy.

Ureteronephrectomy for obstructive mega ureter is currently rare and has been performed in 7% of cases in some series(53,86).

E. Mega ureter complicated by lithiasis: (82)

Concomitant treatment of ureteral lithiasis and mega ureter is the ideal attitude. In some cases, there may be uncertainty as to whether ureteral lithiasis is the cause of the dilation or secondary to stasis, and if in doubt, simply remove the stone and insert a nephrostomy. In the following weeks, the indication for a re-implantation can be safely made.

If the stone is clearly a stasis stone, the mega ureter is treated at the same time as the stone. If the degree of inflammatory damage to the ureter caused by the presence of the stones makes immediate reimplantation dangerous a standby nephrostomy may be necessary if the obstruction is severe.

If the stone is renal, there may be some doubt as to whether it is a mega-ureter or an ureteral hypotonia induced by infection. If the stone is terminal pelvic, molded by the ureter, obstructive stone is the most likely hypothesis and it is best to treat the stone first and see what happens to the upper excretory tract once the stone is removed(69).

In many series, treatment of ureteral lithiasis was performed during uretero-vesical reimplantation, whereas 50% of renal lithiasis was treated by pyelolithotomy during the surgical cure of the megaureter. The rest of the renal lithiasis were treated with extracorporeal lithotripsy.(68)

F. Mega ureter complicated by renal failure:

In the case of severely degraded renal function, a temporary urine diversion in the form of a percutaneous nephrostomy will be performed to relieve the kidney and to judge the recovery capacity of the kidney, on which the further surgical treatment will depend.

Once advanced renal failure has set in, uretero-vesical reimplantation seems unnecessary. In this case, symptomatic treatment of renal failure should be started waiting for a renal transplantation (53).

VIII. MONITORING AND PROGRESS: (53,69):

The effectiveness of surgical treatment is judged by the anatomical and functional improvement of the upper excretory tract.

Thus, in case of a favourable evolution, an IVU is requested; six weeks after the surgical treatment. Ultrasound and Cytobacteriological urinalysis are performed every 3 months for 1 year and every 6 months for 2 years.

Cystography is performed at 6 months to rule out a vesico-renal reflux.

Dynamic scintigraphy is an excellent complementary monitoring test, which is repeated every 12 to 24 months for 5 years and then every 5 years if the results remain unchanged.

The average postoperative surveillance period varies between 6 and 7 years.

IX. PROGNOSIS:

The prognosis of megaureter is conditioned by the degree of renal involvement, hence the importance of early diagnosis and a good therapeutic management strategy (87).

Surgical treatment is effective if the indication has been made early and correctly and if the indications for the ureterovesical reimplantation technique have been respected. The long-term results are excellent in more than 93% of cases.

CONCLUSION

CONCLUSION:

At the end of our study, we retain the following definitions and recommendations:

- The patients with a megaureter are noted predominantly in infants (72.72%) with maximal incidence between 4 and 5 years old (5 out of 11 infants)
- In this study, the sex distribution is found as male : female – 2,66:1, with definitive male preponderance.
- The laterality distribution is noted as the right side was more affected than the left side and we also noted a high frequency of cases of bilaterality.
- the most common symptom at presentation is febrile UTI (63,64%), followed by lower back pain (45.45%).
- In this study, one patient with is noted to have a kidney failure.
- Cohen's Uretero-neocystostomy with intra-vesical approach is the surgical procedure of choice, in majority of the case, in this study.
- out of 11 patients ; 8 patients benefited from a surgical intervention that consists of: Reimplantation of the ureters using the COHEN technique; 4 cases i.e. 36.36% of cases, Modelling intervention using Kalicinski and Starr's technique; 2 cases i.e. 18.18% of cases., Nephrectomy was observed in 2 patients cases i.e. 18.18% of cases.

- Faced with this pejorative development of uropathies, management must be as early as possible through antenatal screening and good follow-up to avoid deterioration of renal function.

ABSTRACTS

ABSTRACT

Introduction: Megaureter (MU) is defined as a congenital dilatation of the ureter > or = 7 mm, it can be associated with vesico ureteral reflux. It is often diagnosed after recurrent urinary tract infection (UTI), but currently, it can be diagnosed since the 3rd trimester of pregnancy thanks to antenatal ultrasound.

Material and methods: Our retrospective study was managed in the Pediatric Surgery service at the mother and child EHS of Tlemcen about 11 ***children*** and 11 cases of megaurter(MU) between January 2021 and July 2022.

Results: In our series we noted a male predominance (72,72% of cases), the mean age of diagnosis was about 5 and ½ years, and three cases was antenatally diagnosed. Eight out of 11 cases were unilaterale and 5 were right sided. Clinical examination of our patients was poor.Six units were refluxing confirmed with retrograde cystography. IVU and scintgraphy were used to confirm the obstructed character of MU. The obstructed non refluxing MU was confirmed in more than 36% of cases. Surgery was performed for 8 patients.

The surgical treatment consisting to the resection of the zone narrows and antireflux re-imlpantation with remodeling of the ureter. We used Cohen procedure in 36.36% of cases, modelling intervention using Kalicinski and Starr's technique procedure in 18.18% of cases and the nephrectomy procedure in 18.18% of cases. After surgery no patient presented a urinary tract infection and renal dilatation tended to regression or stabilization.

Conclusion: The management of PMU is now well codified especially for patients diagnosed prenatally, and surgical treatment appears to have good results when it is indicated.

Résumé

Introduction: Le mégauretère (MU) est défini comme une dilatation congénitale de l'uretère > ou = 7 mm, il peut être associé à un reflux vésico-urétéral Il se révèle dans notre contexte surtout par des infections urinaires à répétition, actuellement grâce à l'échographie obstétricale le diagnostic de MU devient anténatal.

Materiel et méthode: notre étude est rétrospective à propos de 11 patients ayant un mégauretère entre janvier 2021 et juillet 2022, au service de Chirurgie Pédiatrique de l'EHS Mère et Enfant de Tlemcen.

Résultats: Dans notre série nous avons noté une prédominance masculine (72,72% des cas) avec un âge moyen au diagnostic d'environ 5 ans et demi dont trois cas ont été diagnostiqués en anténatal. Huit cas sur 11 étaient unilatéraux dont 5 ont une atteinte du côté droit. L'examen clinique de nos patients était pauvre. Six unités avaient un reflux confirmé par cystographie rétrograde. UIV et scintigraphie avaient confirmées le caractère obstructif du MU. L'UM obstruée sans reflux a été confirmée dans plus de 36 % des cas. La chirurgie a été réalisée pour 8 patients.

Le traitement chirurgicale consistait à la résection de la portion urétérale rétrécit, la réimplantation antireflux après un remodelage urétéral .Nous avons utilisé la procédure de Cohen dans 36,36 % des cas, la modélisation de l'intervention par la technique de Kalicinski et Starr dans 18,18 % des cas et la néphrectomie dans 18,18 % des cas. Après la chirurgie aucun patient n'a présenté d'infection urinaire avec une nette amélioration ou stabilisation de la dilatation urétérale.

Conclusion: Le traitement du MU est bien codifié surtout pour les patients diagnostiqués en anténatale avec possibilité de choix, selon un algorithme bien établit de critères, entre un traitement conservateur ou un traitement chirurgical.

ملخص

المقدمة : يتم تعريف توسع الحالب على أنه توسع خلقي للحالب يتجاوز أو يعادل 7 ملم، يمكن أن تترافق مع الجزر المثاني الحالبي. كثيرا ما يتم تشخيصه بعد التهابات متكررة في المسالك البولية ، لكن حاليا و بفضل الموجات فوق الصوتية خلال فترة الحمل اصبح من الممكن تشخيصه ابتداءا من الاسبوع الثلاثين

المواد والطرق: : دراسة استعادية ولقد عكفت على فحص 11 مريضا من يناير/كانون الثاني 2021 إلى يوليو/تموز 2022 في قسم جراحات طب الأطفال في مدرسة طب الأم والطفل في تلمسان.

النتائج: في سلسلتنا, لاحظنا غلبة للذكور (72.72٪ من الحالات) بمتوسط عمر عند التشخيص حوالي 5.5 سنوات ، تم تشخيص ثلاثة منها قبل الولادة. كانت ثماني حالات من أصل 11 حالة أحادية الجانب ، 5 منها انطوت على تورط الجانب الأيمن. كان الفحص السريري لمرضانا ضعيفًا.

العلاج الجراحي يتجلى في استئصال المنطقة وإعادة زرع مضاد للجزر و اعادة عرض الحالب. استخدمنا إجراء كوهين في 36.36٪ من الحالات ، ونمذجة التدخل باستخدام تقنية كاليسينسكي وستار في 18.18٪ من الحالات واستئصال الكلية في 18.18٪ من الحالات.

بعد الجراحة، لم يظهر أي مريض مصاب بعدوى في المسالك البولية مع تحسن واضح أو استقرار في تمدد الحالب.

الخلاصة: إن علاج التوسع الحالبي البدائي مقنن بشكل دقيق و خاصة للمرضى المشخصين قبل الولادة وفقا لمعايير راسخة سواء عن طريق علاج محافظ أو جراحي.

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APPENDIX

Technical sheet

I. Civil status:

Card number:

File number:

Name:

First name:

Age:

Gender:

☐ Male Female ☐

Age at diagnosis

Address:

Entry date:

Release date:

reason for consultation

☐ Abdominal pain	Lumbar pain	☐
☐ Weak urinary stream	Urinary incontinence	☐
☐ Drop urination	Retention	☐
☐ Dysuria	Hematuria	☐

☐ Urinary tract infection (ECBU+)

☐ Antenatal diagnosis

Medical history

Personal history

Vaccination:

Medical: YES ☐ ☐ NO

if yes, specify:

Surgical: YES ☐ ☐ NO

if yes, specify:

II. Family history:

Consanguinity between parents: ☐YES ☐ NO

Notion of uropathy/lithiasis:

Mother : YES ☐ NO ☐

Father : YES ☐ NO ☐

Physical examination:

- General condition: Good ☐ Altered☐
- size: -weight:
- temperature : Febrile☐ not febrile☐

III. Physical signs:

1. Inspection:

Presence of scar: Yes ☐ No☐ if yes, where:

Abdominal aspect:

☐ flat ☐ symmetrical ☐ asymmetric
☐ bow ☐ bloated ☐ abdominal distension

Urine aspects:

☐ Normal ☐ hematuria ☐ pyuria

If other signs to be specified:

2. Palpation:

☐ Pain caused ☐ abdominal mass
☐ Abdominal swelling ☐ bladder globe
☐ Lumbar contact

If other signs to be specified:

IV. Neurological exam

spine exam

☐ Normal ☐ Abnormal

Cremasterian reflex:

☐ Present ☐ abolished

V. Supplementary Examination:

- Uremia : ☐ normal ☐ IR
- Creatinemia : ☐ Normal ☐ IR
- Creatine clearance:
-CBC : ☐ Anemia ☐ hyperleukocytosis
- Urine report:

ECBU :

☐ Normal ☐ Abnormal ☐ not done

VI. Imaging exams:

1. Non-contrast abdominal X-ray film:

☐ Normal ☐ Abnormal ☐ not done

If pathological to be specified:

2. Intravenous urography:

Morphology:

☐ Normal ☐ hydronephrosis ☐ uretero-hydronéphrosisUrethral ☐ Dilation

Function :

☐ Normal secretion ☐ Delayed secretion

3. Pelvic abdominal ultrasound:

☐ Normal ☐ Abnormal ☐ not done

If pathological to be specified:

4. Voiding Cystourethrogram (VCUG):

-The cervico-urethral parade: ☐ Normal ☐ Spinning top sign

-bladder condition:

-Vesical reflux:

-Ureteral dilatation:

5. Renal scintigraphy:

- DMSA :
- DTPA :

VII. Diagnosis :

- preoperative diagnosis:

☐ Hydronéphrosis ☐ multicystic dysplasia
☐ vesicoureteral reflux

☐ Polycystic kidney disease ☐ Renal agenesis
☐ Megaureter

☐ Seat anomalies ☐ Rotation anomalies
☐ Fusion anomalies ☐ Posterior urethral valve

If other signs to be specified:

- postoperative diagnosis :

VIII. TREATMENT:

Medical :

☐ Anti-inflammatory ☐ painkillers
☐ antibiotics

If other signs to be specified:

Surgical :

Procedure:

IX. Recovery:

after a week :

☐ Simple ☐ complicated ☐ Death