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Antenatal hydronephrosis

Hidronefrose antenatal

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ABSTRACT: Genitourinary anomalies can be detected in the antenatal period with incidence of 0.5 a 1% in gestational population; 20 a 30% of these anomalies involve the urinary tract. Hydronephrosis is the most frequent urinary tract anomaly followed by cystic anomalies. Currently, technical advances of high-resolution ultrasound identify, in a non-invasive way, the gestational anatomy. The evaluation of the urinary tract, by means of complementary examinations: precocious abdominal and pelvic ultrasound (US), laboratory analysis, functional evaluation (DTPA99 -glomerular function; DMSA99 -tubular function and MAG99 -MAG-3), voiding cystourethrography and others if necessary. *Conclusion* - Every pregnant woman should undergo at least one morphological ultrasound examination carried out in satisfactory conditions and by qualified professional during the prenatal follow-up to identify possible malformations of the fetus. Following a pre-established roadmap of complementary examinations, it is possible to treat the pathology safely, allowing the newborn to receive hospital discharge in good condition, with a mandatory multidisciplinary outpatient follow-up with pediatric, nephrologist and pediatric urologist's consultations.

Keywords: Hydronephrosis; Urinary tract/abnormalities; Urinary tract/diagnostic imaging; Prenatal care; Ultrasonography.

RESUMO: As anomalias do trato geniturinário podem ser detectadas no período ante natal com incidência de 0.5 a 1% na população gestacional; 20 a 30% envolvem anomalias do trato urinário. Hidronefrose como consequência é mais frequente, seguida anomalias císticas. Atualmente com avanços técnicos de ultrassonografia de alta resolução podem identificar de modo não-invasivo, a anatomia gestacional. A avaliação do trato urinário segue com exames complementares: ultrassonografia pós-natal, análises laboratoriais, avaliação funcional (DTPA 99- função glomerular; DMSA 99 – função tubular e MAG 3), cistouretrografia miccional e outros se necessário. *Conclusão* – A gestante deveria submeter-se, pelo menos a uma ultrassonografia morfológica em condições adequadas para identificar malformações fetais. Seguindo um protocolo pré-estabelecido, é possível tratar patologias com segurança e ao recém-nascido quando liberado do hospital, em boas condições; acompanhamento de equipe multidisciplinar com pediatra, nefrologista e urologista pediátrico.

Descritores: Hidronefrose; Sistema urinário/anormalidades; Sistema urinário/diagnóstico por imagem; Cuidado pré-natal; Ultrassonografia.

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INTRODUCTION

Genitourinary anomalies can be detected in the antenatal period with incidence of 0.5 a 1% in gestational population; 20 a 30% of these anomalies involve the urinary tract; 50% correspond to defects in the central nervous system, 15% of the gastrointestinal system and 8% are cardiopulmonary abnormalities. Hydronephrosis is the most frequent urinary tract anomaly followed by cystic anomalies – multicystic and polycystic kidney – and others like agenesis, renal hypoplasia and dysplasia, obstructive uropathies – Ureterocele, ureteral ectopy, posterior urethral valve – later identified already from the 15 to 16^a gestation week^{1,2}.

The Gestational ultrasound has made a major impact on the detection of fetal anomalies, mainly fetal hydronephrosis, allowing in addition to the presumptive diagnosis, the treatment of asymptomatic obstructive uropathies in the neonatal period even before the installation of infection in the tract urinary.

Currently, technical advances of high-resolution ultrasound identify, in a non-invasive way, the gestational anatomy. The ultrasound detects the Fetal hydronephrosis, with access to the echogenicity of the parenchyma alerting the possibility of renal dysplasia; congenital anomalies of other systems are also detectable by the ultrasound: hydrocephalus, fusion defects in the vertebral arches, diaphragmatic hernias, intestinal obstruction and defects of the abdominal wall, among others. The following parameters should be evaluated by the doctor in the face of any urologic anomaly detected in the gestational period: fetal sex, amount of amniotic fluid, unilateral or bilateral pathology, evidence of ureteral dilation, existence of a bladder with thickened wall, distended or empty bladder and posterior urethral dilation in males.

Etiology

Hydronephrosis corresponds to the physiological or organic dilation of the renal excretory pathways and the degree of dilation is variable with gestational age; It is usually unilateral, but in 20% of the cases it is bilateral, and the majority may decrease by the end of gestation or in the first year of life. The Ureteropelvic Junction (UPJ) is the most common place of congenital obstruction; the ultrasound shows pyelocaliceal dilation and eventual tapering of the renal parenchyma. Normal ureters are not visible in the ultrasound; the bladder appears after the 13th e 16th week and at the end of the 32th week it may have a measured capacity of 10 ml.

Other urinary tract dilation conditions and non-urinary pathologies (digestive, neurological, gynecological) may mislead to an equivocal hydronephrosis diagnosis:

multicystic kidney, polycystic kidney, Renal cyst, megaureter, pyeloureteral duplicity, Ureterocele, vesicoureteral reflux, posterior urethral valve, ovarian cyst, urachal cyst, hydrocools, sacrococcygeal teratoma, meningocele, duodenal atresia, intestinal duplication, mesentery cyst. The antenatal ultrasound describes only anatomical data, with little indication for renal function and without any information about histopathology.

The fetal diuresis constitutes a parameter in clinical evaluation; in evaluating the possible commitment of renal function, Ultrasonography continues to be useful by directing the needles for urine collection in the bladder or renal pelvis; Normal fetus produces hypotonic urine (osmolarity < 210/mEq/L; Na⁺ < 100 mEq/L and Cl⁻ < 90mEq/L). When renal function is compromised, reabsorption ff Na⁺ is altered, increasing the urinary concentration of this electrolyte and the osmolarity. This increase in levels between the 16th and 20th fetal week may be associated with the spina bifida or anencephaly, omphalocele as well as oligohydramnios and can correspond to severe obstructive uropathies. The microglobulin beta² is the most measurable protein and is entirely absorbed by the proximal fetal tubule after glomerular filtration; uropathies severely increase the urinary concentration of beta microglobulin², in such a sort that urinary levels above 13mg/L may define fetal inviability by renal failure³.

It is important in history, the investigation of renal background in the family; there's a high incidence of hereditary genetic syndromes with renal impairment. In pregnancies interrupted with fetal death or after birth, the fetus should be subjected to autopsy for diagnostic conclusion, besides the possibility of genetically targeting parents when necessary. The dilation of the urinary tract is not always justified as obstruction of the excretory pathways, i.e. they are not juxtaposed and remains still great dilemma today.

For the pediatric urologist confronting different spectra or degrees of hydronephrosis, the challenge is for identifying and selecting obstructive systems for possible surgical decompensation. Association of severe dilation of the urinary tract and oligohydramnios, detected by the Gestational ultrasound, are predictive factors for high-risk gestation.

The approach of urological malformations should be multidisciplinary, involving the participation of obstetrician, neonatologist/intensivist, nephrologist and pediatric urologist, participating in both diagnosis and treatment as guiding parents and mainly in genetic anomalies. Similarly, government institutions should be asked to participate in early diagnosis by means of standardization and/or compulsory in performing gestational ultrasound, aiming at preventive treatment (Figure 1).

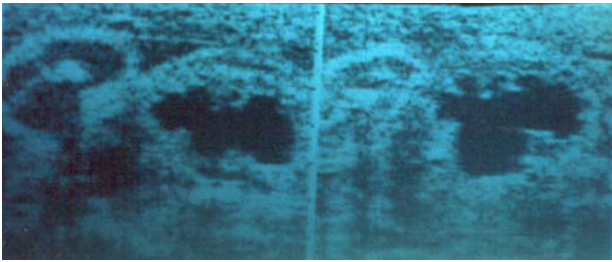


Figure 1 - Ultrasonography antenatal showing hydronephrosis of left kidney, pelvis and dilated calyces with discrete tapering of renal parenchyma

Diagnosis

Imaging methods

The first evaluation of the newborn is made by the neonatologist, who must be informed of all history of the antenatal investigation of the urological malformation; obstructions/dilations detected early on; and when there is a progression of such alterations, demanding detailed research in the post-natal period. It is important to assess the first voiding stream by observing if the urinary voiding is weak, normal or if urination occurs by dripping.

The obstructive uropathies, particularly the most serious, may present clinical and laboratory manifestations soon after birth, such as presence of abdominal mass located on the flanks (Figure 2), palpable bladder that does not reduce with urination, in addition to urinary tract infection, septicemia, hematuria and/or renal failure. The presence of genital defects also makes it suspicious of concomitant anomalies in the urinary tract or endocrinopathies.



Figure 2 - Abdominal mass in the left hypochondriac, cystic and mobile consistency with respiration = hydronephrosis

The evaluation of the urinary tract, by means of complementary examinations, will depend basically on the nature of the lesion and the clinical condition of the newborn. In the nursery attached to the maternity (BAM)

Hospital of the University of São Paulo, from 1990 on, we have established a Research Protocol of the urogenital anomaly with antenatal diagnosis and we follow the according script with some flexibility^{2,4}:

1) Precocious abdominal and pelvic ultrasound (US)

After stabilization of the newborn (NB): evaluation of the thickness of the renal cortex, quantification of pyelocaliceal dilation, evaluation of the ureter, bladder and urethra⁵. The age of the NB to perform the neonatal ultrasound to determine whether the time affects the accuracy of the examination, was the reason for prospective study comparing the ultrasound after 48 hours and with 7 days of life. Neonatal follow-up with a serial ultrasound and evaluation of hydronephrosis (classified according to the Society of Fetal Urology). There was an increase in hydronephrosis in 44% of the renal units between the 1st and 2nd Ultrasound. They concluded that the hydronephrosis is underestimated after 48 hours of life of the NB whose difference clinically is not significant; They recommend initial ultrasound between 7 and 10 days of life and may be more early in selected cases (Figure 3)⁶.

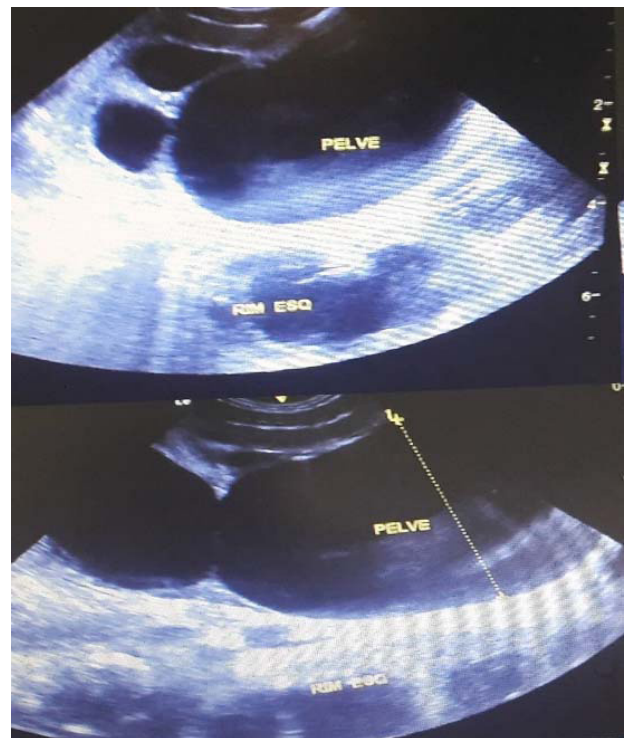


Figure 3 - The postnatal ultrasound in the first week: defines the size of the kidney, shows the dilation and thickness of the renal parenchyma (severe hydronephrosis)

2) Laboratory

Analysis: Urea, creatinine, venous gasometry and serum electrolytes in the first week; clearance of creatinine is calculated from the serum and urinary dosage collected in a period of 6 hours. Analysis of the urinary sediment and culture of urine collected by means of collector's bag or supra-pubic puncture.

3) Functional evaluation

From the urinary tract via radio isotopes from the 4th week: the main pharmacological drugs used are: DTPA⁹⁹ Dietilenotriaminopentacético acid; (Glomerular function); DMSA⁹⁹ Dimercaptosuccinic acid (Tubular function) and MAG⁹⁹ Mercuroacetiltriglicina (MAG-3), which measures tubular function between 1 and 3 minutes and used to measure renal elimination.

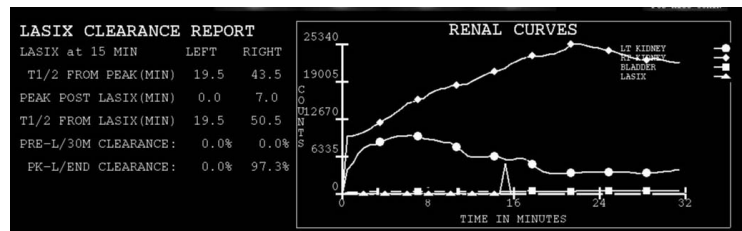
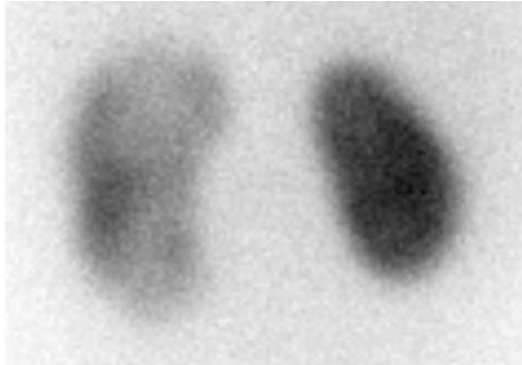


Figure 4 – A: DMSA – Clear areas of hypo uptake of the drug; It measures the renal function separately. B: DTPA - the downward curve corresponds to the left kidney, discreetly slow, though normal; the superior line corresponds right kidney obstructed, lowered function and which does not change after the diuretic injection

- Renogram with diuretic - DTPA and MAG-3: A bladder catheter is recommended in the presence of vesicoureteral reflux (VUR) and in small children. The Time Curves X Activity obtained after injection of the radio drug presents three distinct phases: arrival to the kidney (ascension curve), peak of 60 seconds (renal cortical) and decline curve (excretion in the collector system). The provocative test with diuretic (furosemide) is usually quantified by T1/2, that is, time required to eliminate 50% of the activity of the substance in excretory system. Normal standards: T1/2<15minutes (normal) and T1/2>20minutes (obstruction) and undeterminate between 15 and 20 minutes (Figure 4B). The current consensus is that the MAG 3 constitutes the agent of choice⁸.

4) Voiding cystourethrography

Last evaluation exam, held in the 4th week under antibiotic prophylactic. When it comes to ureteral hydronephrosis diagnosed in postnatal ultrasound, it is required anticipated to suspected by vesicoureteral reflux or obstructive ureteral dilation (megaureter and/or urethral obstructions).

5) Intravenous pyelography (IVP)

It is not routinely used and is performed in specific situations, for example, to assist in the overall morphological analysis of the urinary tract and in the presence of important dilations of the excretory pathways.

6) Anterograde percutaneous pyelography

In selected cases, when the previous exams were insufficient to characterize important anatomical aspects for the definition of the therapeutic strategy; In the face of the need for anesthesia, its realization may be concomitant to the surgical act. Often, following the percutaneous approach it is performed a perfusion test with pelvic

- Renal Scan-DMSA: It is a static renal assessment; the Tc⁹⁹-DMSA maintains connection to the cells of the proximal tubule after several hours of intravenous administration. The image represents the functioning cortical mass; the normal differential renal function varies between 45 and 55%. It has greater sensitivity than ultrasound and Intravenous pyelography (IVP), in the definition of acute and chronic pyelonephritis (Figure 4A).

pressure assessment (Whitaker test)⁹, to better quantify the degree of UJP obstruction or uretero-vesical obstruction; It intends to artificially reproduce the urinary flow resistance to the ureter or the ureter dilated into the bladder; it is indicated when the eventual obstruction was not defined by the radioisotopes test - DTPA.

7) Computer tomography (CT) / Magnetic resonance imaging (MRI)

The role of TC/MRI is limited and they are not routinely used, being employed in more complicated situations where the previous methods did not define the diagnosis (Figure 5). IVP with MRI using gadolinium contrast is being used in children with hydronephrosis, providing superior images of both kidney and ureter, with excellent spatial and three-dimensional resolution, when compared with renal diuretic renografin (MAG-3, DTPA)¹⁰.

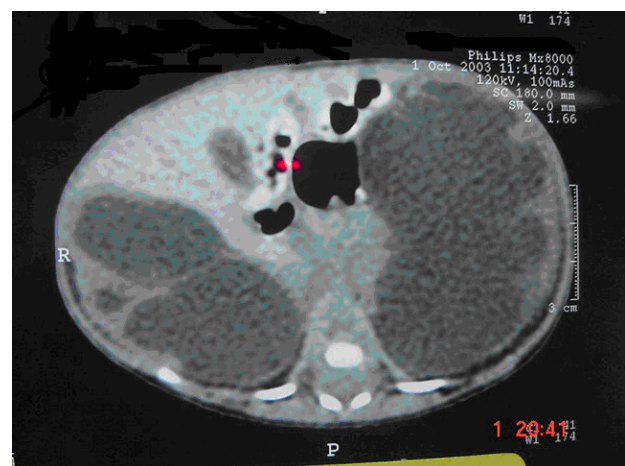


Figure 5 – Computed tomography of the abdomen identifying the bilateral hydronephrosis

Clinical picture

In the nursery attached to maternity (BAM) of the Hospital das Clínicas of São Paulo-USP, Sector of Perinatal Urology, there were 146 Cases/6.635 births (= 2,2% “High risk gestation”) with the diagnoses of the urinary tract malformation. They had their first care in this maternity that only receives patients with high risk gestation (diabetes, arterial hypertension, antenatal congenital anomalies) (Table 1).

Table 1 - Type and frequency of malformation's in newborns

Type	N	(%)
Hidronephrosis (UPJ)	40	(32.0%)
Myelomeningocele	19	(15.2%)
Multicystic kidney	14	(11.9%)
Posterior urethral valve	08	(6.4%)
Vesicoureteral reflux	04	(5.6%)
Megaureters	05	(4.8%)
Duplicity with ureterocele	05	(4.0%)
Unilateral renal agenesis	07	(4.0%)
Prune - belly	04	(3.2%)
Anorectal anomaly	03	(2.4%)
Multicystic e UPJ	02	(1.6%)
Pyeloureteral duplicity	02	(1.6%)
Others	12	(9.6%)
Total	146	(100%)

Anomalies and surgical considerations

1- Ureteropyelic Junction Stenosis (UPJ): It is the most frequent anomaly, with incidence of 1/1000 newborns, predominating in boys. Antenatal diagnostic suspicion initially can be confirmed by physical examination that often presents palpable mass in the palpable mass on the flank. The postnatal US shows kidneys with images that may suggest cysts, but unlike these, are communicating cavities among themselves; the presence of a larger central cyst renal and smaller peripherally cysts suggest hydronephrosis. The evidence of renal parenchyma, with varying thickness, reinforces the ultrasound diagnosis of hydronephrosis.

Renal scan with DTPA (glomerular function) is the ideal examination to evaluate the renal function of filtration of both kidneys; the criteria for surgical correction present many variables, for example, kidney function less than 40%; surgery to fix the stenosis should be recommended in the first six months of life. If renal function is above this value, clinical follow-up with repeated scintigraphy after 3, 6 and 12 months is suggested, and surgical correction

may be performed if renal function deteriorates. Currently, unilateral hydronephrosis has been considered a benign pathology in the face of the recovery of renal function observed in large series of newborns, subjected to expectant clinical treatment. Antibiotic prophylactic and systematic monitoring of renal function should be administered¹¹. Antenatal UPJ stenosis and Renal pelvis: <15mm, light = note/regress; 15-30mm, moderate = 70% can progress; if >30mm, severe = usually surgical.

2- Multicystic kidney: The ultrasound image of multiple renal cysts, with no visible renal parenchyma and unidentified ureter induces the diagnosis of rim multicystic (Figure 6); the differentiation with hydronephrosis by UPJ stenosis is difficult, especially when the number of cysts is small and can be confused with dilated calices. Unlike the stenosis of UPJ, the evaluation with radioactive isotopes (DMSA) demonstrate a lack of function of this kidney. Management should be discussed with parents: When the contralateral kidney is normal, clinical follow-up with US is the universal approach adopted, until 4 to 5 years old. The multicystic kidney tends to diminish in size and behaves silently. Another proposed approach to parents would be nephrectomy without the need for periodic evaluations, and without potential risks of hypertension, infection and/or malignant degeneration justify it is justified in some cases¹².

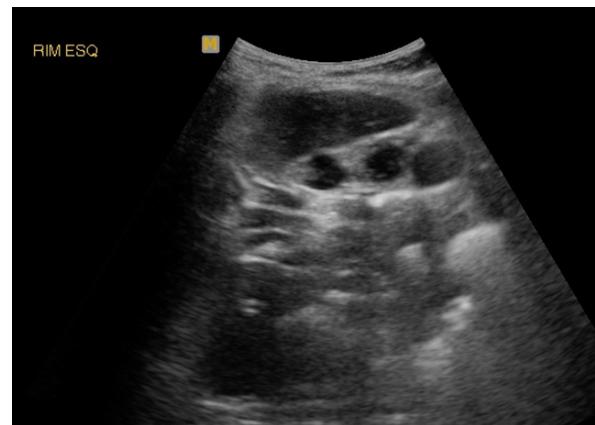


Figure 6 – Multicystic kidney

3- Megaureter: 50% of newborns, mainly those with antenatal diagnosis, are asymptomatic and will have spontaneous regression of the dilation of the excretory pathways and improvement of the renal function that must occur until 2 years of age. The megaureter with the worst evolution is the one with a larger diameter of 10 mm, whose dilation is consequent to an obstructive component, to the point of imposing surgical treatment (urinary derivation) in the first months of life. Renal functional evaluation by means of radioisotopes and renal scan can define the degree of commitment of renal function and confirm that ureteral dilation is obstructive or not (Figure 7)^{13,14}.



Figure 7 - Unilateral Megaureter, important dilation of excretory pathways, the caliber of the ureter exceeds 10mm (intravenous pyelography)

4- Ureterocele and ureteral duplication: In childhood the ureterocele is usually associated with the duplication of the pyelocaliceal system but it can also occur in simple unit of the excretory system; by definition, ureterocele corresponds to the congenital dilation of the intra-vesical ureter with stenosis of the ureteral orificile and variable ureterohydronephrosis. In addition to the risk of urinary infection caused by obstruction and urinary tract,

ureterocele can hinder vesical emptying even in women. In newborns, intervention is occasionally indicated for decompression of the obstructed unit already in the first weeks of life; it can be performed by nephrostomy, ureterostomies or preferably by incision of ureterocele, endoscopically. These procedures facilitate the treatment of infection, the functional and anatomical recovery of the compromised renal unit.

5 - Vesicoureteral Reflux: ureteral dilations arising from the vesicoureteral reflux (VUR) are generally smaller than those consequent to the megaureter. The neonatal vesicoureteral reflux associated with hydronephrosis, has a predominant impact on boys and is usually associated with high-grade reflux. Normal postnatal ultrasound should not exclude cystourethrography (Figure 8A and 8B). High-grade reflux in the newborn may be associated with the deterioration of renal function even before the urinary infection is installed. Even in the presence of significant dilation and absence of recurrent urinary infection, most of this reflux improves and can resolve spontaneously. Also, differential diagnosis should be made between primary and secondary VUR, consequent to a neurogenic bladder, posterior urethral valve, among others. Vesicoureteral reflux with good vesical emptiness should be managed expectantly with prolonged chemoprophylaxis; Only in massive ureteral dilations with severe functional deterioration a temporary urinary derivation should be performed (vesicostomy or cutaneous ureterostomy). The natural history of VUR reveals a spontaneous improvement and healing with age, which is why surgical treatment is indicated only in very special situations and rarely before the first year of life^{15,16}.

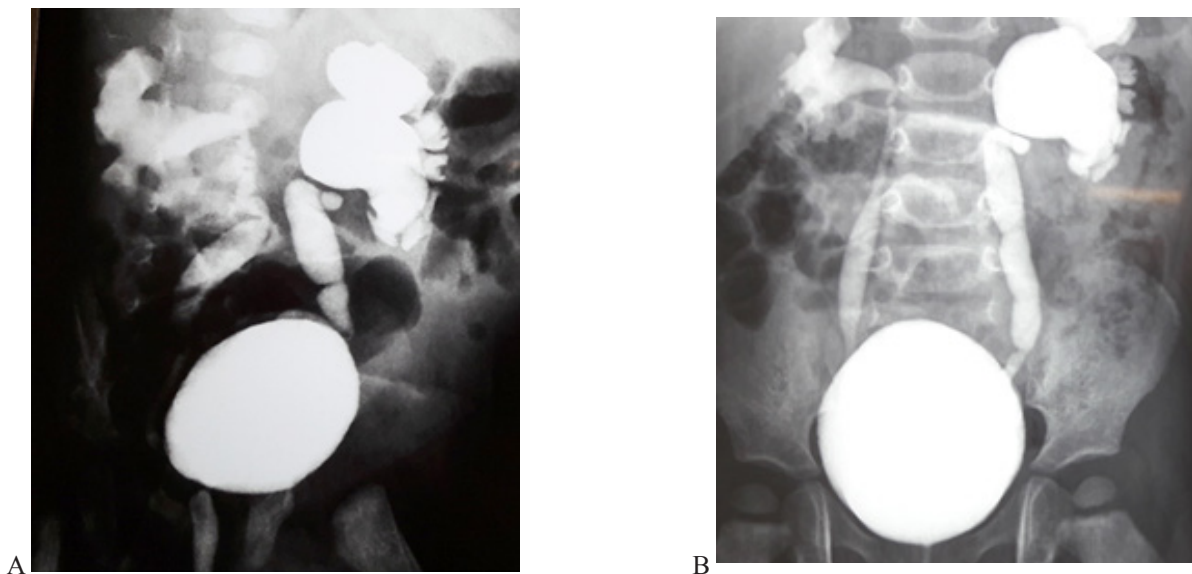


Figure 8 – A: Cystourethrography - vesicoureteral reflux bilateral in newborn, one month age, Grade V; B: Vesicoureteral reflux bilateral, grade III/IV, after 18 months of chemoprophylaxis, same child

6 - Posterior urethral valve (PUV): It is the most common and most serious cause of infravesical obstruction in boys. The presumptive diagnosis with antenatal ultrasound of posterior urethral valve is reinforced in physical abdominal examination when the presence of hypogastric “tumor” which reduces little with urination. The general state of the newborn may be compromised and aggravated by antenatal factors related to oligoamine (respiratory failure) and obstructive renal failure. The US examination demonstrates dilation of the urinary tract, with eventual presence of urinary ascites; renal function

may be compromised in the early days of life and, not rarely, these children develop urinary sepsis when urinary infection it is not treated in time. The diagnosis of certainty of PUV is obtained by voiding cystourethrogram, with the identification of the dilation characteristic of the posterior urethra and the secondary hypertrophy of the vesical lap (Figure 9). If an emergency vesical urinary drainage is necessary, it can be accomplished by urethral catheter or by supra-pubic puncture. It should be established antibiotic monitored by renal function, pending the anatomical and functional recovery of the urinary tract.

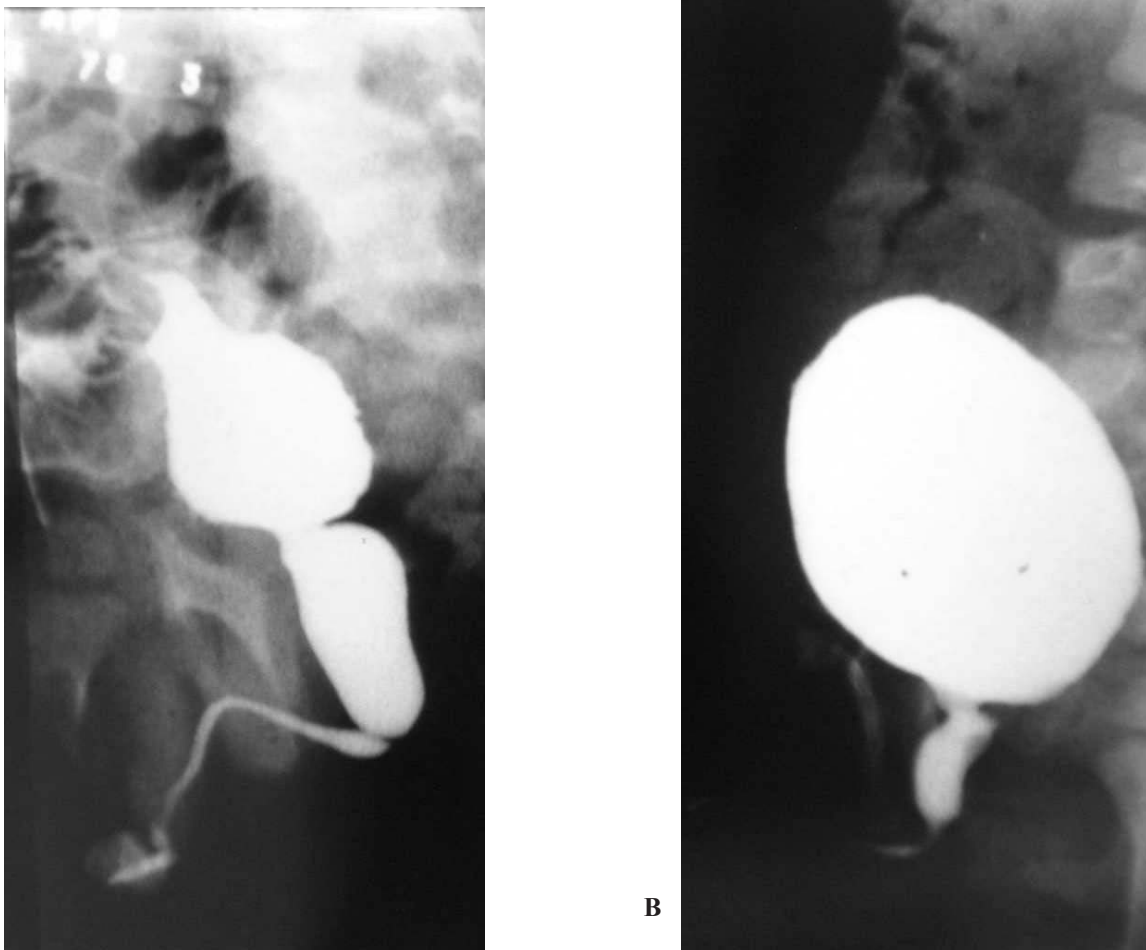


Figure 9 - A - Cystourethrography shows obstruction in the posterior urethra (posterior urethral valve). The posterior urethra is quite dilated and obstructed. B - post-ablation valve: absence of reflux and important decrease of urethral dilation

The definitive treatment is endoscopic valve ablation. In newborns with severe metabolic decompensation, “urosepsis” or massive dilation of the upper urinary tract, it is indicated high urinary derivation (ureterostomies) to make it easier to drain urine in the initial period of treatment. Vesicostomy, until recently indicated as a satisfactory technique of derivation, is subject of controversy and has been avoided by the fact that it defunctionalizes the bladder and it does not often adequately drain the upper urinary tract. With the antenatal diagnosis and early institution of clinical and/or surgical treatment, soon after birth, the

prognosis of these patients has improved considerably, allowing a good anatomical and functional recovery in most patients¹⁷.

CONCLUSION

Every pregnant woman should undergo at least one morphological ultrasound examination carried out in satisfactory conditions and by qualified professional during the prenatal follow-up to identify possible malformations of the fetus. Following a pre-established roadmap of

complementary examinations, it is possible to treat the pathology safely, allowing the newborn to receive hospital discharge in good condition, with a mandatory multidisciplinary outpatient follow-up with pediatric, nephrologist and pediatric urologist's consultations.

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