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RESEARCH ARTICLE

ANTENATAL DIAGNOSIS OF POSTERIOR URETHRAL VALVE: A CASE REPORT.

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Abstract

The posterior urethral valve (PUV) is the most frequent cause of end-stage renal disease in male children. The echographic appeals that may suspect this malformation are variable. We report the case of a 33 year old patient, without a specific antecedent. In whom the T1 ultrasound screening reveals an aspect of megavessy with renal dilation. The T2 ultrasound shows an enlarged bladder with a thickened wall. Both kidneys have a visible cortex, abnormal echostructure marked by bilateral ureter-pyelo-calicial dilatation. Amniotic fluid index is normal. The T3 Ultrasound reveals a persisting bilateral ureter-pyelo-calcicial dilatation and a large bladder with thick walls. The Delivery was natural at 37 SA + 5J and the development of the baby was normal.

Introduction:-

The posterior urethral valve (PUVs), also referred as congenital obstructing posterior urethral membranes, are the most common congenital obstructive lesion of the urethra and a common cause of obstructive uropathy in infancy [1]. This rare anomaly is usually isolated (some cases associated with a Prune-Belly syndrome).

Antenatal diagnosis is possible in more than 80% of cases. The PUV is the most frequent cause of kidney failure in male kids.In less severe cases,the diagnosis is often not apparent until early infancy.

Observation:-

The clinical case:-

Mrs. B is 33 years old, with no specific antecedents.
First trimester ultrasound finds a distended bladder with bilateral hydronephrosis.
Nuchal translucency measurement is 1.2mm and Crown-rump length is 55.3mm.
The remainder of the morphological assessment is normal.

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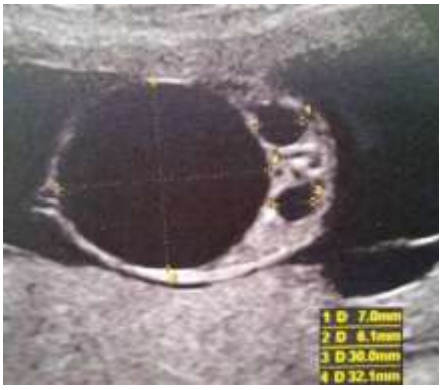


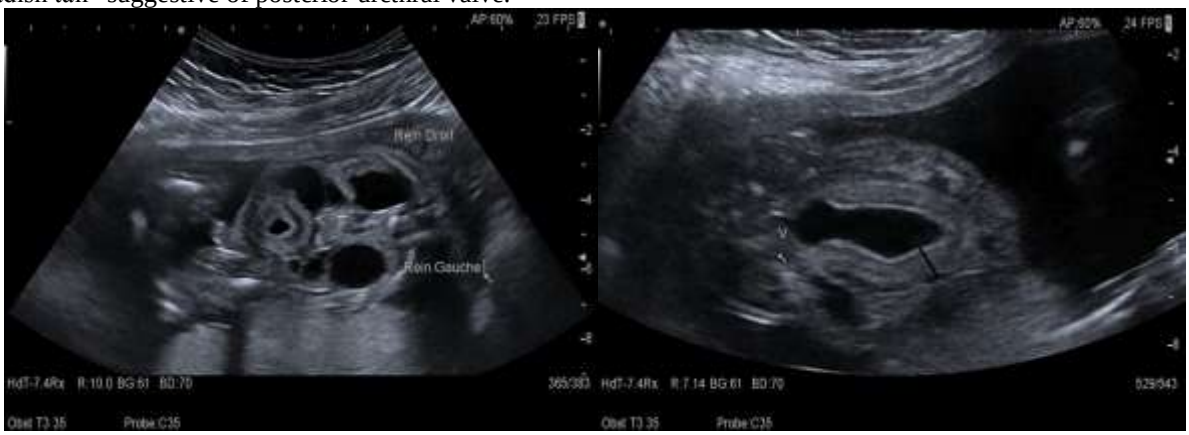
Figure 1: A cross section showing a distended bladder with bilateral ureteral dilatation

Ultrasound follow up:-

At 14 WA: The sonographic control shows that the distention and hypertrophy of bladder worsened (30 x 38mm) also the bilateral pyelectasia (7 and 6 mm) with the presence of a urethral recess.

At 20WA+ 4D:-

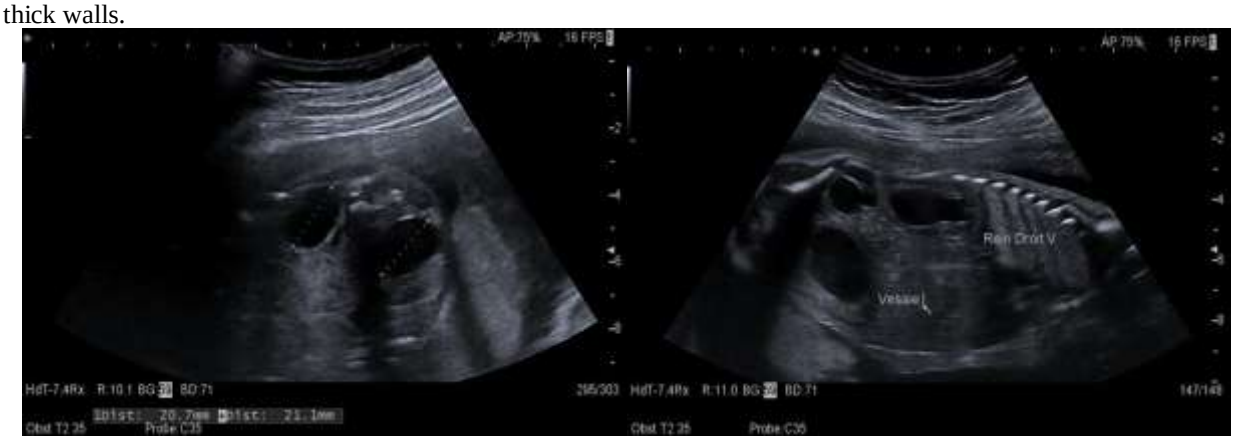
Fetal growth is normal. The Bladder size is 37.8 x 10.5 mm and thick is 8 mm, with a vesical recess similar to "radish tail" suggestive of posterior urethral valve.



The two kidneys keep a visible cortex measuring 3.5 mm with normal echostructure marked by bilateral ureter-pyelo-calcial dilatation, right pyelon measuring 20.7mm and left pyelon 21.1mm.. Amniotic fluid index in normal.

At 34SA + 5D:-

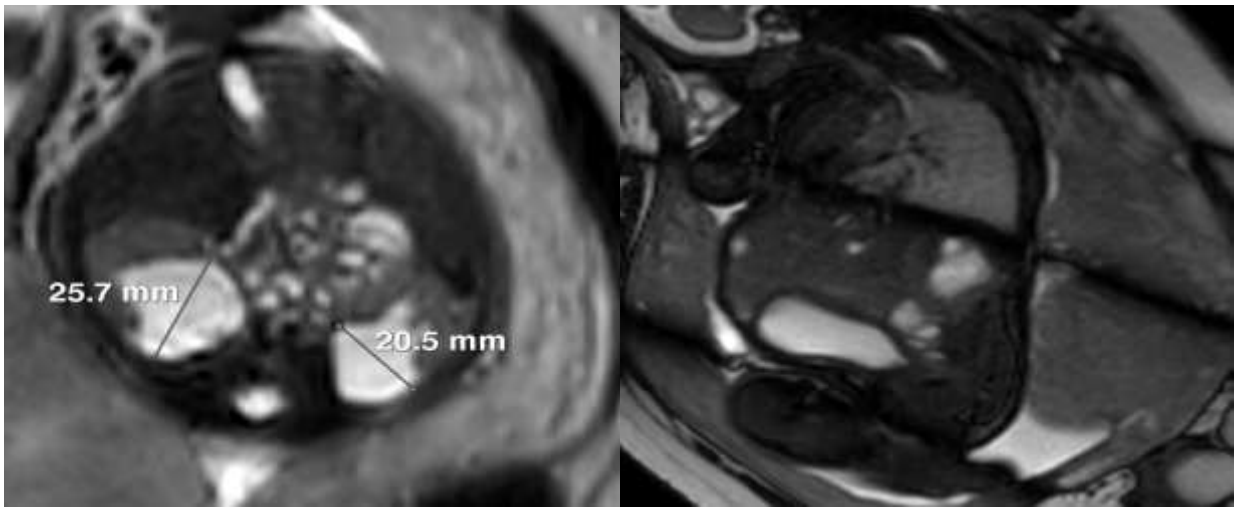
Persistent Bilateral hydroureteronephrosis. Right pyramid measures 19mm and left 25mm.Bladder was large with thick walls.



Fetal MRI:-
A fetal MRI was realized at 31WA that confirmed the diagnosis of PUV with dilatation of the subvesicular ureter.

The right kidney measured 40 mm and the left one 47.5 mm with a normal peripheral renal parenchyma.

There were a bilateral hydroureterodronephrosis with renal malrotation, measured 25 mm to the right and 20 mm to the left.



Delivery and postnatal evolution:-
Labor was inducted at 37SA + 5J for oligohydramnios. Delivery was vacuum-assisted and gave birth to a male newborn, weighed 2680g, Apgar 7 then 10/10 transferred to neonatal care unit.

The day of birth:-
Biological evaluation:-
Urea 4 mmol / l Creatinine 77 mmol / l Sodium 142 mmol / l Potassium 3.3 mmol / l

Malformation assessment:-
1. Abdominal-pelvic ultrasound: Kidneys' echo structure was good, the right one measured 44x19mm and the left one 42x19mm. Pyelic ectasia was bilateral between 7 and 8mm. Bladder was large with a thick wall and cryptorchidism was bilateral. Liver, spleen and pancreas were normal.
2. Echocardiography : No cardiac disease.
3. Transfontanelle ultrasound - Lumboscral spine Xray - Spinal cord sonography : normal.

Day one of life:-
Retrograde cystography :-
The posterior urethra was dilated. Large diverticular bladder integrated with a pathological cervix and urachus residue in a Belly Prune syndrome.

Surgical management :-
Endoscopic section of the posterior urethral valve.

One month of life:-
Good evolution

Renal ultrasound:-
The right kidney measured 65mm and the left one 69mm with a minor dilatation of the pyelocalyceal system.

Discussion:-

Definition and Epidemiology:-

Posterior urethral valve is an obstructive developmental anomaly in the urethra and genitourinary system of male newborns and the estimated incidence is at l 2.1 per 10 000 live births [2]. It was first described by Hugh Hampton Young and coworkers in 1919. Antenatal diagnosis is possible in more than 80% of cases and can sometimes be done from 12 weeks of amenorrhea (WA).

Clinical presentation:-

Clinical presentation depends on the severity of obstruction. In severe obstruction, the diagnosis is usually made antenatally. The fetus will be small for gestational age and ultrasound examination will demonstrate ligohydramnios and associated abnormalities (see below). In less severe cases, the diagnosis is often not apparent until early infancy. Urinary tract infections are common in this group.

Antenatal diagnosis:-

Ultrasound is a good tool for screening for VUP syndrome with good sensitivity but a specificity that remains low.
On antenatal ultrasound, the appearance is that of:

1. Marked distention and hypertrophy of the bladder.
2. Hydronephrosis and hydroureter may or may not be present.
3. In severe cases oligohydramnios and renal dysplasia (assessing the degree of renal dysplasia is difficult antenatally, although some authors believe that significantly increased echogenicity of the kidneys is an indication of poor function) [3].
4. Keyhole sign may be seen on ultrasound due to the distention of both the bladder and the urethra immediately proximal to the valve [4].

Treatment:-

Antenatal treatment is possible, consisting of vesicoamniotic shunting (allowing urine to exit the bladder via the shunt, bypassing the obstructed urethra). Essentially this procedure consists of a supra-pubic catheter performed under ultrasound guidance. The efficacy of this procedure is controversial, as often despite this significant renal and pulmonary morbidity exist [5].

Postnatally, definitive treatment involves transurethral ablation of the offending valve,correction of hydroelectrolytic disorders and fighting against the urinary infection.

Prognosis:-

Prognosis is most affected by the degree and duration of obstruction. Severe cases with obstructive cystic renal dysplasia, oligohydramnios and pulmonary hypoplasia are often incompatible with life.Up to 3% of newborns with VUP die during the neonatal period due to respiratory insufficiency caused by pulmonary hypoplasia with a mortality risk of around 50% [6 - 7].

Good Factors are: Nadir creatinine < 0.8 mg/dl, S. creatinine < 1 mg/dl, Pop-off mechanism that joins the VURD (Posterior urethral valve, Unilateral vesicoureteral reflux, Renal dysplasia) syndrome, ascitis and large bladder diverticulum[8].

Conclusion:-

Posterior urethral valve (PUV) remains the most frequent cause of subvesical obstruction of the child. It is also the most serious malformatif uropathy. Ultrasound is the first-line examination in case of suspicion of VUP. It allows the antenatal diagnosis of VUP, but also the evaluation of its repercussion by the study of the quantity of amniotic fluid and the aspect of the kidneys. Initial management at birth consists of ensuring bladder drainage, correction of hydroelectrolyteic disturbances and then endoscopic section of valves. In severe forms a voluntary termination of pregnancy is accepted in Western countries.

Conflict of interest statement:-

There is no conflict of interest between the authors.

Compliance with Ethical Standards:-

Ethical Approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. Informed Consent Informed consent was obtained from participant included in the study.

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