

OUTCOME OF ANTENATAL INTERVENTION FOR POSTERIOR URETHRAL VALVE

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Abstract

Posterior Urethral Valves (PUV) are the most common cause of lower urinary tract obstruction in male fetuses, eventually leading to compromised kidney function as well as pulmonary hypoplasia due to oligohydramnios. A 34-year-old primigravida nullipara in her 18th gestational week was referred to the Elias Emergency University Hospital in Bucharest for further second trimester morphology investigations. The prenatal course was uncomplicated prior to admission. The ultrasound examination revealed fetal megacystis, severe oligohydramnios and a dilated urethra giving the appearance of the “keyhole” sign. These findings were consistent with the diagnosis of posterior urethral valves. In an attempt to temporize the unfavorable evolution, our patient decided to undergo a percutaneous vesicoamniotic shunting in order to bypass the obstructed urethra through a pig-tailed catheter placed between the fetal bladder and the amniotic cavity. Although the procedure has improved the ultrasonographic appearance of the fetal kidneys and the existing oligohydramnios was corrected, the long term evolution was eventually poor because of the previously compromised renal function.

Keywords: posterior urethral valve, vesicoamniotic shunting, urethral obstruction, antenatal

Introduction

Posterior urethral valves (PUV) are the most common congenital cause of lower urinary tract obstruction diagnosed in the male fetus, occurring sporadically with an incidence of 1 in 8000 pregnancies [1]. Familial cases have also been reported.

The etiology of PUV is not yet clearly established, although proposed theories include hypertrophy of the urethral mucosal folds, persistence and continuation of the urogenital membrane and the abnormal development of the Wolffian duct [2]. The result is an obstructive

intraluminal membrane situated proximally to the verumontatum.

The antenatal diagnosis of PUV is mostly based on maternal ultrasound and fetal urinary biochemical assessment for the renal function [3]. Although the ultrasonographic visualization of the valve is not possible because of its small size, consequences such as fetal megacystis along with a dilated urethra in a male fetus raise the suspicion of PUV. In severe cases of obstruction, oligohydramnios may also be present [4].

The differential diagnosis is made with Prune belly syndrome, urethral atresia and bilateral vesico-ureteric reflux.

Presentation of the case

A.P., a 34-year-old primigravida nullipara in her 18th gestational week was referred to the Elias Emergency University Hospital in Bucharest for further second trimester morphology investigations. The prenatal course was uncomplicated prior to admission and she had no past relevant obstetrical history.

The ultrasonography revealed fetal megacystis with bladder measurements of 41/31 mm (Figure 1). The thickened bladder wall along with a dilated proximal urethra gave the appearance of the “keyhole” sign, which is suggestive for posterior urethral valve in a male fetus (Figure 2). Furthermore, severe oligohydramnios was present, with the deepest vertical pocket of 1 cm.



Figure 1 - Ultrasonography - Fetal megacystis with bladder measurements of 41/31 mm



Figure 2 – Ultrasonography - the “Keyhole” sign; the proximal urethra is distended, appearing as a small extension of the bladder

Fetal bilateral ureterohydronephrosis was identified, as well as an increase in the cortical echogenicity of both kidneys (Figure 3).



Figure 3 - Ultrasonography - Echogenic kidneys with a dilated, tortuous ureter

At this stage, possible options for intervention include vesicoamniotic shunting (from the bladder to the amniotic fluid) and endoscopic ablation of the valve, both applying exclusively when the process of kidney damage is reversible and therefore its function is salvageable. Another option would be the medical termination of pregnancy [3].

In this case, despite the echogenic appearance of the kidneys, which was highly suggestive of renal function compromise, the placement of a shunt was decided considering the early gestational age.

The patient underwent a percutaneous vesicoamniotic shunting which provided an alternate passageway for the fetal urine. Under maternal anaesthesia and antibiotic cover, with mother in supine position, the procedure consists of introducing an ultrasound-guided trocar through the maternal and fetal abdomens in order to place a double pig-tailed suprapubic catheter (Figure 4). Over a guide wire, the distal end was settled inside the fetal bladder and the proximal end in the amniotic cavity. The guide wire was then withdrawn.

The fetal bladder pressure was relieved, therefore allowing the drainage of the urine after bypassing the obstructed urethra. As a result, three days after the intervention the bladder dimensions decreased to 16/14 mm and the amniotic fluid index (AFI) increased to 5 cm (Figures 5, 6).

Follow-up is required to assess fetal renal function, which is at risk of being compromised.



Figure 4 - Percutaneous Vesicoamniotic Shunting under ultrasonographic guidance



Figure 5 - Postprocedural ultrasonography reveals smaller bladder, detectable catheter inside the bladder (white lines) and increased amniotic fluid.



Figure 6 - Postprocedural ultrasonography - Improved aspect of the kidneys due to pressure relief

Despite the apparent improvements that have been noticed, two weeks after the procedure (20 weeks gestational age) there was absent amniotic fluid, the kidneys maintained their echobright appearance, the ureters appeared dilated and tortuous. The urinary bladder was of

normal size, but without visible dynamics (emptying/filling sequence) on repeated, prolonged examinations. The catheter was visible within the bladder, but the outer end could not be detected. The image was suggestive of total renal compromise with anuria.

Considering the lethal prognosis, the patient decided to undergo a medical termination of the pregnancy. The second trimester pregnancy interruption was carried out using Mifepristone, an antiprogesterone steroid and Misoprostol (Figure 7,8).

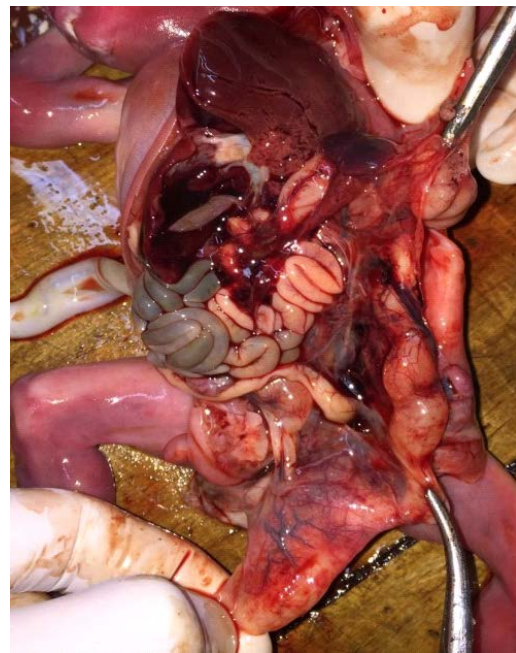


Figure 7 – Dilated ureter

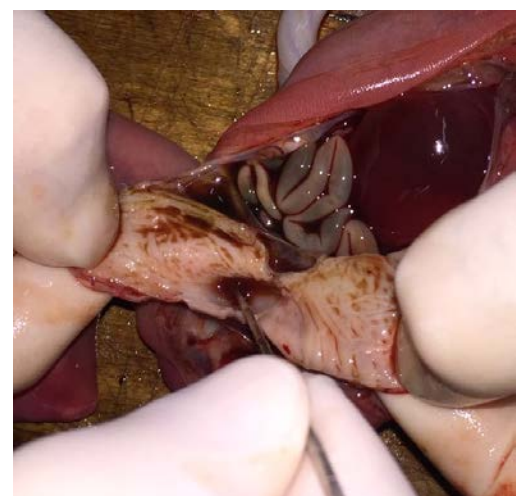


Figure 8 - Thickened smooth muscle trabeculations; the urethra could not be passed through to the meatus due to the existing obstruction.

Discussions

Due to the mechanical obstruction of the urethra, the entire urinary tract development is altered by the increased voiding pressures.

With the aim to prevent this, early antenatal intervention by vesicoamniotic shunting improves the outcome in terms of renal function and bladder dynamics after birth, as the overall prognosis is mostly affected by the degree (volume of amniotic fluid) and duration (gestational age at diagnosis) of the obstruction [3,5,9].

The placement of the vesicoamniotic shunt is used as a temporizing measure until delivery.

If successful, postnatally, definitive treatment involves the transurethral ablation of the offending valve. This is performed within the first several days of life, along with the removal of the catheter [6].

Nonetheless, the risk of perinatal mortality and postnatal chronic kidney failure are increased if the diagnosis is made before 24 weeks of gestation, or if findings consistent with renal dysplasia are present [7].

Post-procedural complications include shunt blockage, preterm delivery and urinary ascites.

A promising alternative is the fetoscopic laser ablation of the valve, which can achieve bladder decompression and amniotic fluid normalization with a single procedure in selected cases with anhydramnios. However, there is a significant risk of progression to renal failure pre- or postnatally [8].

Conclusions

The efficacy of antenatal intervention by percutaneous vesicoamniotic shunting is questionable, as the long term survival remains poor. Although the procedure has initially improved the ultrasonographic appearance of

the fetal kidneys, it has failed to compensate the unfavorable evolution of the pregnancy.

In order to improve the survival rates as well as the preservation of fetal renal function, the technique and timing of the procedure need to be reevaluated.

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