Fetal Ascites and Posterior Urethral Valves. A Case Report and Literature Review

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Abstract

Abnormality of posterior urethral valves is a common cause of obstructive uropathy during fetal life. The prognosis depends on the degree of oligohydramnios, with which posterior urethral valves are always associated; moreover, it is a factor that significantly affects the intrauterine development of the lungs as well as the severity of the kidney impairment. Nowadays, diagnosis is prenatally possible, which leads to early intrauterine treatment with urinary decongestion, thus prognosis is significantly improved. Nevertheless, chronic renal insufficiency occurs later to a significant number of neonates. In this paper, a case of a male fetus with posterior urethral valves manifested as fetal ascites after rupture of the urinary bladder wall, as well as bilateral hydronephrosis is described. The parents decided, after having been thoroughly counseled, to terminate the pregnancy due to the risk of severe chronic renal failure, based on amniotic fluid electrolyte tests.

Introduction

The presence of posterior urethral valves is a congenital malformation of the urinary system that threatens the life of the neonate. Despite optimal treatment provided, posterior urethral valves can lead to kidney failure in approximately 17% of the cases. Posterior urethral valves occur in 1 in every 5,000-8,000 births [1].

The Hampton Young classification, describes three types of urethral valves. According to the current scientific point of view, type II is not considered obstructive, because it is a simple fold and therefore is not considered a valve at all. Type I (90-95% of cases) is the most common type of urethral valves, consisting in two occlusive folds in the area of the bulbous urethra. Type III valves are located at different levels of the posterior urethra, consisting in a fold attached to the entire perimeter of the urethra, with a small hole in the center [2].

In males, an obstruction at the level of the urethra affects the entire urinary system, resulting in dilation of the prostatic urethra, while ejaculation may be impeded due to urinary reflux [3,4].

When the bladder neck is overfed and hardens, it affects the bladder, which becomes dilated with hypertrophic and hyperplastic response of bladder wall and may sometimes present diversions.

Upper urinary tract dilatation occurs in almost all patients, due either to the presence of valves and high pressure in the bladder, or to the obstruction of the cystourethral junction by the hypertrophic bladder wall [4].

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Keywords: fetal ascites; congenital abnormality; posterior urethral valves

Received: 30 October 2020; Accepted: 20 November 2020; Published: 30 November 2020
In the case of secondary urinary reflux, the functionality of the corresponding kidney is low. In the frame rate of the prenatal ultrasound examination, bilateral hydronephrosis, dilated ureters and urinary bladder are signs of urethral valves. The occurrence of this congenital anomaly may also be suspected when a dilated posterior urethra and bladder with thickened walls are observed, as well as when increased echogenicity of the kidneys in the presence of hydronephrosis and oligohydramnios are identified. Surgical treatment is indicated, while the long-term prognosis depends on the degree of renal dysplasia and the severity of the lung hypoplasia. This emphasizes the importance of prenatal diagnosis for a timely management [2-4].

If immediate intrauterine treatment is performed, the deterioration of the renal function and impaired lung development are prevented. It is of great importance to highlight the need for suspecting the occurrence of posterior urethral valves in the presence of a urinary tract obstruction, mainly in male fetuses with dilatation of the lower part [5,8].

However, the present case confirms that this type of congenital malformation is associated with risks of renal impairment until failure, which is also the basis of the parents’ decision to terminate the pregnancy.

Case report

A 39-year old pregnant woman, IIG / 0P with the following medical history was introduced for the 2nd trimester anatomical ultrasound examination at 22 + 0 weeks of gestation.

Patient with primary infertility (increased FSH and LH, low AMH, normal hysterosalpingography imaging, unremarkable sperm count).

In 2018, a missed abortion was reported during the 7th week of gestation following spontaneous conception. The current pregnancy resulted after treatment with clomiphene citrate, induction of ovulation and homologous insemination.

The course of the pregnancy until then presented no complications. Pregnancy multivitamin agents were administered. Normal findings of the 1st trimester’s detailed ultrasound screening examination (12+3 GW) were reported.

Current ultrasound findings: male fetus in vertex presentation and appropriate for gestational age intrauterine growth. Fundal-posterior placenta, degree of maturity 0 according to Gramann classification, normal amount of amniotic fluid.

Umbilical cord with central, normal bulge and 3 vessels, cervical length 38 mm, internal cervical orifice closed.

The following pathological sonographic findings were noted:

Fetus with significant intraperitoneal fluid accumulation and moderate bilateral dilatation of the renal pelvies. Fetal bladder with irregularly thickened walls and blurred borders. Absence of signs of fetal anemia (normal maximal Peak Systolic Velocity PSV in the Middle Cerebral Artery-Doppler).

Urinary ascites was suspected after rupture of the urinary bladder due to overfilling (megacyst) in the presence of posterior urethral valves in a male fetus (Figure 1 A,B).

Testing for congenital infections (TORCH, Parvo virus B19), antibody test and indirect Coombs test were found to be negative. Rh (+) positive blood type.

After amniocentesis, a normal molecular karyotype of a male fetus was identified. Ultrasound re-examination was performed in 23 + 0 GW which showed a normal amount of amniotic fluid, an increase in ascites accumulation, a severe dilation of the pelvic and calyx system in the fetal kidneys (figure 2 A,B), dilation of both ureters, enlarged “keyhole” urinary bladder with thickened and irregular walls, as well as rectal dilatation (figure 3 A,B). The pregnant woman was referred to a special center for intrauterine surgery for the placement of a vesicoamniotic shunt. The plan was to insert an intravesical pigtail catheter to relieve the urinary bladder and drain the urine into the amniotic cavity. Ascites drainage should also be performed.

Biochemical tests were carried out on the fetal ascites fluid. Based on the trace elements measurements (sodium levels 110 mmol/l, chloride levels 100 mmol/l, calcium 8.5 mg/dl, B2–microglobulin 11,2 mg/l, total protein 43,1 mg/l, osmolality 202 mOsm/l), a poor prognosis for the kidneys function was revealed (possible renal insufficiency or future necessity of kidney transplantation). After consultation with a pediatric urologist, the parents decided to terminate the pregnancy due to high probability of severe fetal renal failure. Fetal cystoscopy or vesicoamniotic shunting were rejected by the parents, due to high complication rate: 74% and 92%
respectively [9, 12].

Figure 1A: 23 GW; fetal urinary ascites (yellow arrow) after rupture of the urinary bladder wall in a male fetus with posterior urethral valves.

Figure 1B: 23 GW; fetal urinary ascites (yellow arrow) after rupture of the urinary bladder wall in a male fetus with posterior urethral valves.
Figure 2A: 23 GW: fetal hydrenephrosis - compressed renal parenchyma (yellow arrows) in a male fetus with posterior urethral valves.

Figure 2B: 23 GW: fetal renal pelvis & calyces dilatation (yellow arrows) in a male fetus with posterior urethral valves.
Discussion

The posterior urethral valves consist of an abnormal relative obstructive membrane which is located inside the posterior urethra in male fetuses [2-4]. During embryogenesis, the caudal end of the Wolff pore is absorbed into the primordial cloaca at the site of the future seminal vesicle in the posterior urethra. The residues of this process form the folds of the posterior urethra (plicae colliculi) [2-4].

Histological studies have shown that the posterior urethral valves are formed during the 4th week of gestation, when Wolff's pore fuses with the developing...
cloaca. They were first described by Young in 1919, who classified them into 3 types [4]. Type I is a two-way valve, extending from the posterior end of the seminal vesicle to the anterior end of the proximal membrane clause (95% of cases). Type II is no longer considered a blockage valve and it is described as a hypertrophic muscle band extending from the posterior wall of the urethra to the seminal vesicle. Type III is a circular membrane located in the membranous urethra. This type is considered to be the result of incomplete absorption of the urogenital membrane during embryogenesis (5% of cases) [2,10-12].

Neonates with low urinary tract obstruction (LUTO) due to posterior urethral valves may develop severe respiratory distress immediately after birth, as a result of pulmonary hypoplasia, a consequence of oligohydramnios [11-14].

Perinatal mortality’s rate was formerly 50% due to pulmonary hypoplasia renal insufficiency and sepsis. This percentage decreased to <3% due to the improvement of perinatal care and prenatal diagnosis. The use of ultrasound examination in fetuses has led to the ability of an early detection [15].

In 1989, a study by Thomas et al reported that 10% of patients with a prenatal diagnosis of hydronephrosis had posterior urethral valves [16]. Later, in 1993, Dinneen et al reported low sensitivity of prenatal ultrasound for the diagnosis of posterior urethral valves (only 45%) [17]. The sensitivity of the method has increased over the last 10 years [15]. Body fluids, such as blood and amniotic fluid, behave, to some degree, like water solutions. The skin of the fetus behaves like a semi-permeable membrane until at least 25 weeks of gestation, when a process called "keratinization" is completed; this process starts at the 19th week of gestation and makes the skin impermeable [3,6,17-19]. The fetus grows protected inside the amniotic sac, the walls of which consist of two semi-permeable membranes, filled with amniotic fluid. These membranes also cover the surface of the umbilical cord and the placenta. Through these semi-permeable membranes and due to the phenomenon of osmosis, exchanges of liquids take place.

In the first half of gestation, the amniotic fluid likely comes from fluid, which, due to the osmosis, penetrates the membranes of dermis and the amniotic sac, that separate the fetus from the mother. Amniotic fluid reaches the fetus through the semi-permeable covering membranes, the first of which is the skin, until its keratinization at the 25 weeks of gestation [3,6,17-19].

Through these membranes, fluid enters the placenta and the umbilical cord and is then infused into the fetus. Amniotic fluid appears to exit the amniotic cavity through the amniotic sac and the membranes, which are in contact with the surface of the endometrium. It seems that the amniotic fluid that exits the mouth and nose of the fetus originates from lung and tracheal secretions. The production of these secretions is achieved through the so-called "breathing movements" of the fetus, which are lung movements, following chest activity. Respiratory movements are recorded by ultrasound since the 11 weeks of gestation. The contribution of these secretions to the production of amniotic fluid is rather small, since the fetus swallows part of these secretions [3,6,17-19].

The fetal urination is the main mechanism, which increases the amount of amniotic fluid. From the 8th week of gestation onwards, the kidneys produce urine and the fetus urinates through its urethra. In fact, the bladder can be imaged on ultrasound, with the use of transvaginal probes, from just the 9th week of gestation and with the use of transabdominal probes from the 11th week of gestation onwards [15].

The reduction of amniotic fluid is mainly due to its ingestion by the fetus, a highly important procedure for the maturation of the lungs and the digestive system of the fetus, which starts shortly after the 8th week of gestation and can be imaged on ultrasound.

The posterior urethral valve is a membrane that covers the posterior urethra either in part or totally [1].

Prenatal ultrasound examination may show hydronephrosis, dilatation of the ureters, enlargement and thickening of the bladder, proximal urethra and oligohydramnios.

Prenatal intervention is mainly limited to a few facilities; moreover, it is not clear whether it has a significant benefit in long-term prognosis. Pulmonary function appears to be improving but no benefit has been found in terms of renal function [12,18,19].

After birth, there may occur neonatal edema, palpable dilated bladder, small radius of urination, signs and symptoms of renal and respiratory failure.

In neonates with suspected posterior urethral valves, abdominal x-ray, ultrasound and cystourethrography should be performed. Ascending cystography and cystourethrography during urination lead to the diagnosis, enhanced by the characteristic dilation of the posterior
urethra, and provide indications of possible coexistence of
cystourethral reflux.

Secondary cystourethral urinary reflux occurs in at
least 50% of the patients with posterior urethral valves.
In these patients, reflux is consistently associated with
renal dysplasia.

Kidney scintigraphy to assess renal function is
essential. Creatinine, urea and blood electrolytes should be
monitored in the first days of life. Low creatinine levels
(≤80 μmol/l) are associated with a better prognosis [18-20].

If the diagnosis is confirmed, a catheter should be
inserted to decompress the bladder.

After stabilization of the patient, the valves are
removed cystoscopically. In difficult cases, a cystostomy
is performed first and the valves are subsequently
removed. The prognosis is constantly improving in recent
years. In the past, the diagnosis was made postnatally, after
the onset of a urinary tract infection or progressive renal
failure. Today, in most cases, the urethral valves are
suspected after hydrenephrosis is detected in the prenatal
ultrasound examination [15]. Early treatment of bladder
obstruction and improved surgical techniques have
reduced neonatal mortality to <3%, but about 1/3 of
patients will develop kidney failure [18-21].

A good prognostic element at birth is the drop in blood
creatinine after decongestion of the bladder to levels below
0.8mg/dl.

Posterior urethral valves are a condition that requires
lifelong monitoring for early diagnosis and treatment of
complications, such as chronic renal failure (30%),
inability to condense urine and renal tubular acidosis
[20,21].

**Treatment**

**A. Prenatal treatment**

Nowadays, about 40-60% of the cases of posterior
urethral valves are identified prenatally. The obstruction
leads to reduced urine production by the fetus and
oligohydramnios. Amniotic fluid is essential for the
normal development of the lungs and its deficiency can
lead to lung hypoplasia, which increases the perinatal
mortality and morbidity. Attempts have been made for
intrauterine treatment of the posterior urethral valves.

As renal dysplasia is not reversible, it is important to
identify affected fetuses with sufficiently good renal
function (Sodium levels below 140 mmol/l, chloride
below 90 mmol/l and osmolality below 200 mOsmol/l,
found in three fetal urine samples, taken on three different
days), owing to the fact that only those fetuses are
associated with a better prognosis.

The intrauterine placement of a catheter which drains
the urine from the fetal bladder directly into the amniotic fluid,
presents a complication rate of 21-59%. Catheter
movements occur in up to 44% of cases, while mortality
rates extend between 33% and 43% and renal failure over
50% [12]. Although the catheter treats oligohydramnios,
there are no differences in the long-term outcomes of
patients with posterior urethral valves [18-21].

**B. After birth treatment**

Bladder drainage: If a male neonate is born with
suspected valves of the posterior urethra, a catheter is
immediately inserted into the bladder and a
cystourethrography is performed to determine if the
diagnosis is correct.

Alternatively, a supravesical catheter may be inserted
into the bladder where it remains until a
cystourethrography is performed, and an endoscopic
resection of the valves is carried out.

Valves excision: When the condition of the neonate
stabilizes and creatinine levels drop, the next step is to treat
the obstruction. With the use of special small cystoscopes,
cross-section and excision of the valves is possible [18-21].

Cystostomy: If immediate endoscopic surgery is not
possible, because of prematurity or co-morbidities, a
bladder drainage is used as a temporary treatment. An
intravesical catheter is initially inserted, which can remain
at the site for 6-12 weeks. Otherwise, a cystostomy can be
performed (creating an opening in the skin and bladder
through which urine is drained), which improves or
stabilizes kidney function in 90% of the cases [18-21].

There are, of course, concerns that the cystostomy may
affect the capacity or elasticity of the bladder. If the
drainage of the bladder cannot relieve the upper urinary
tract, then an ureterostomy or a pyelostomy can be
performed (direct drainage of urine from the ureter or the
pelvis of the kidney).

High deviation may be necessary when urinary tract
infections occur and no improvement in renal function or
upper urinary distension is observed, despite cystostomy.
The restitution of the urinary tract after diversion occurs when renal function is improved as much as possible.

About 72% of the patients will develop cystourethral reflux, which will be bilateral in about 1/3 of the cases. Decreased renal function is commonly linked to severe cystourethral reflux.

Lifelong monitoring of these patients is extremely important because bladder dysfunction may occur in many cases, while controlling urine on a 24/7 basis can be challenging. Bladder dysfunction that may occur is due to decreased sensitivity and elasticity of bladder’s wall. Moreover, the bladder’s extruder muscle is highly unstable, while polyuria is an additional important factor. Renal insufficiency may occur in about 10%-47% of affected patients and transplantation of kidney may be necessary to them [18-21].

**Conclusion**

The occurrence of posterior urethral valves can lead to a massive dilatation of the entire fetal urinary tract and is associated with loss of nephrons, starting from the intrauterine life due to renal dysplasia. This condition results in a primary or subsequent chronic renal failure. The extent of nephron loss determines the renal prognosis. Furthermore, lung hypoplasia on the basis of oligohydramnios during pregnancy leads to a dramatic increase in perinatal morbidity and mortality rates.

Early prenatal sonographic diagnosis is nowadays possible. The intrauterine placement of an intravesical catheter creating a cysto-amniotic drainage of fetal urine can help decongest the kidneys and preserve renal function. Nevertheless, prenatal intervention is mainly limited to a few facilities; moreover, it is not clear whether it has a significant benefit in long-term prognosis. Pulmonary function appears to be improving but no benefit has been found in terms of renal function.

**References**


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