

Prenatal Diagnosis of Posterior Urethral Valves

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Posterior urethral valves are a urologic deformity involving a narrowing of the prostatic portion of the male urethra. This causes a bladder outlet obstruction. The sonographic criteria include hydronephrosis, bilateral ureterectasis, and enlarged bladder, with a “keyhole” appearance in the region of the obstruction of the prostatic urethra. Varying degrees of oligohydramnios will be present due to the inability of the fetus to release urine. Additional findings may include pulmonary hypoplasia due to lack of amniotic fluid. In severe cases, the bladder may rupture and cause urinary ascites within the fetal pelvis. A case study demonstrating this condition is presented.

Key words: posterior urethral valves, bladder outlet obstruction, hydronephrosis, pulmonary hypoplasia, oligohydramnios

Case Report

A multigravida patient in her late 20s was referred to the prenatal detection center due to an abnormal obstetrical sonogram at 18 weeks’ gestation demonstrating fetal hydronephrosis. The Siemens Sequoia imaging unit was used with a 6-MHz curved linear array transducer. The sonographic findings included enlarged hydronephrotic kidneys (Figs. 1-2), hydroureterectasis, severely enlarged bladder, and bladder outlet obstruction. The sonographic “keyhole” pattern was well demonstrated at the prostatic portion of the fetal urethra (Fig. 3). A male fetus was confirmed by visualizing the male genitalia. There was severe oligohydramnios with an amniotic fluid index of 4 cm. The sagittal image of the fetal abdomen displayed significant abdominal distension caused by the enlarged kidneys and bladder (Fig. 4). The cardiothoracic ratio was increased to 45%, presumably due to pulmonary hypoplasia (Fig. 5). A

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FIG. 1. Bilateral hydronephrosis.



FIG. 2. Enlarged kidneys.



FIG. 3. Bladder outlet obstruction at posterior urethra.

small circumferential pericardial effusion was also demonstrated (Fig. 6). The diaphragm was elevated due to the enlarged bladder and kidneys. No other fetal anomalies were identified.

Discussion

Posterior urethral valves are a rare condition affecting only males. The abnormality occurs in

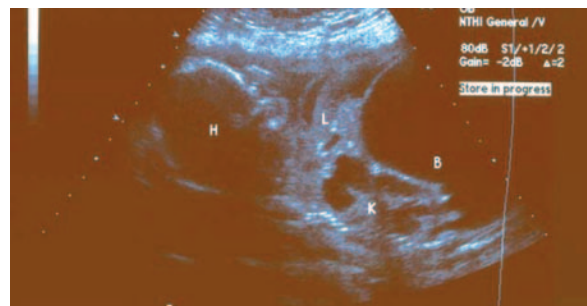


FIG. 4. Anyhydramnios. H = head; L = liver; K = kidney; B = bladder.



FIG. 5. Increased cardiothoracic ratio secondary to pulmonary hypoplasia.



FIG. 6. Small pericardial effusion.

about 1 in 8000 births.¹ Posterior urethral valves are formed within the sixth to eighth weeks of gestation.² Thickening of these valves may cause proximal urethral distension, a bladder outlet obstruction, and subsequent hydroureteronephrosis. Pulmonary hypoplasia may develop secondary to oligohydramnios.² The prognosis is dependent on the renal function and degree of pulmonary hypoplasia.³ Perinatal mortality has decreased

from approximately 50% to < 10%.⁴ This is due to improvements in prenatal and perinatal care and the increased incidence of the elective termination of pregnancies following prenatal detection. Pregnancies with oligohydramnios have a greater risk of morbidity and mortality.⁵ Because chromosomal abnormalities, including trisomies 21, 13, and 18, have been reported in up to 20% of the cases,² fetal karyotyping is recommended in cases with sufficient amniotic fluid. It is also possible to check the fetal renal function by aspirating urine from the enlarged bladder. When sodium, chloride, and osmolality are elevated, poor renal function is suggested.

The distinguishing feature of posterior urethral valves is the presence of urethral dilatation and an enlarged bladder. The typical keyhole appearance separates this urologic deformity from other conditions such as prune belly syndrome, ureterovesicular junction (UVJ) obstruction, severe ureterovesicular reflux, large ureterocele, or severe hydronephrosis. Sonographic evaluation should include the presence of urethral obstruction, enlarged bladder, variable degree of hydronephrosis and ureterectasis, amniotic fluid index, and the presence or absence of suspected pulmonary hypoplasia using the cardiothoracic ratio. Pericardial effusion, although present in this case, is not specifically associated with either posterior urethral valves or pulmonary hypoplasia.⁶

Summary

In fetuses such as the one described in this case study, the prognosis is dismal due to inevitable

pulmonary hypoplasia. The outcome of this pregnancy was infant mortality on day one of life due to pulmonary hypoplasia. Early sonographic detection is essential to ensure that a physician will promptly evaluate the newborn or, in some cases, seek prenatal intervention. Prenatal vesicoamniotic shunting has been used to therapeutically reduce the bladder volume and increase the amniotic fluid content. This has been met with variable success, using strict prognostic criteria on a case-to-case basis. When successful, amniotic fluid volume is temporarily restored, and the chance of pulmonary hypoplasia is reduced. In less severe cases, however, postnatal treatment may include supportive care such as a catheterization regimen, endoscopic ablation (removal of valve leaflets), or vesicostomy (small opening made in the bladder from the abdomen).

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