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# POLYCYSTIC HORSESHOE KIDNEY WITH MEGACYSTIS AND ANHYDRAMNION AS A RARE FETAL MALFORMATION

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Polycystic horseshoe kidney is thought to represent two separate renal diseases. Horseshoe kidney is a renal fusion anomaly during embryogenesis. Autosomal dominant polycystic kidney disease (ADPKD) is a hereditary disorder due to mutations in the genes responsible for the expression of the proteins polycystin 1 and polycystin 2. Polycystic horseshoe kidney is very rare with incidence ranges of 1 in 134 000 to 1 in 8 000 000 live births.

We present a case of an 18-year-old primigravida at 22 weeks of gestation with ultrasonographically detected cystic formation in the abdomen (a dilated bladder) and anhydramnion. Screening for chromosomal abnormalities in the first trimester (PRISCA 1) found a low risk. Labor was induced with oxytocin and a stillborn male fetus weighing 605 grams was delivered. A complex congenital malformation of the urinary system was detected on autopsy – a dilated bladder, membrane of the urethral valve, and polycystic kidneys that were fused at their lower pole with a horseshoe appearance. On cross-section, the renal parenchyma was reduced due to the presence of numerous cysts with a diameter of 1-6 mm, filled with clear content. Microscopic examination confirmed the macroscopic finding of polycystic kidneys, with a reduction of the renal parenchyma and the presence of numerous cysts lined with low cylindrical to cuboidal epithelium. Analysis of the bladder sample showed hypertrophy of the detrusor muscle. This malformation led to impaired urination and absence of amniotic fluid.

The aim of this case presentation is to emphasize that prenatal ultrasound examination allows the detection of major fetal defects and pregnancy can be terminated if the fetus is severely affected. Fetal autopsy is a method of vital importance in complementing prenatal diagnosis. It may add valuable information that may improve future pregnancy management and hence prenatal ultrasound and fetal autopsy should be regarded as complementary techniques.

**Keywords:** polycystic kidney disease, horseshoe kidney, megacystis, anhydramnion