Large fetal pulmonary arteriovenous malformation with severe cardiomegaly and favorable postnatal outcome.

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Introduction: Pulmonary arteriovenous malformation (PAVM) is a rare finding in a fetus and may lead to cardiac failure and hydrops. Because of the variable outcome of the few previously reported cases, perinatal management and parent counseling are difficult. The case presented a large PAVM complicated by severe cardiomegaly with a successful postnatal treatment.

Case: A 20-year-old woman was referred at 23w+1d of gestation due to suspicion of congenital heart malformation. US scan revealed normal fetal growth and amniotic fluid. The heart was in normal location with a severe cardiomegaly (CTAR: 0.628). Pulmonary trunk (PT) and right pulmonary artery (RPa) were dilated with retrograde perfusion through the ductus arteriosus. Turbulent flow was found in the right lung connecting the RPa with a dilated right pulmonary vein (RPv), and left-to-right shunt through the foramen ovale (FO). Doppler of the PT revealed a velocity of 72 cm/s and high velocity at the fistula (260 cm/s) with low pulsatility. The descending aorta and umbilical artery showed high pulsatility but normal peak velocity, with no reversal end-diastolic flow. The ductus venosus had no reverse “a” wave. No signs of hydrops or other malformations were found. The fetus was monitored every 2 weeks, cardiac function remained unchanged. The mother was sent to a center with pediatric cardio-surgery option. At 38 weeks a female fetus was vaginally delivered (weight 3270g; Apgar score 8-8 at 1-5 min) and then referred to a neonatal intensive care unit at a pediatric hospital. The image studies confirm a fistula, then, a surgical excision was performed and the fistula was closed. The newborn evolved favorably, without hemodynamic or neurological alterations at six months of age.

Conclusion: This is one of the few published cases of prenatal diagnosis of a large PAVM with successful postnatal treatment resulted in survival with adequate life quality. Close monitoring of the fetus and planned multidisciplinary management seems to be essential for the outcome.