Sudden Intrapartum Fetal Death in Fetuses with Absent Pulmonary Valve Syndrome: Report of Two Cases

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Abstract

Objective To describe potential intrapartum complications for fetuses affected by absent pulmonary valve syndrome.

Study Design Two cases of intrapartum fetal death at full term were collected from our institution’s labor and delivery unit records.

Results In both cases of intrapartum fetal death, the fetuses had been diagnosed with absent pulmonary valve syndrome and had likely experienced acute cardiac events during labor. Both were delivered as stillbirths despite emergency cesarean delivery.

Conclusion Patients should be counseled prior to labor about potential intrapartum complications for a fetus with absent pulmonary valve syndrome. Plans for fetal monitoring and the extent of aggressive intervention should be in place before labor in case sudden complications occur.

Keywords► absent pulmonary valve syndrome ► intrapartum fetal demise ► high-risk pregnancy

Absent pulmonary valve syndrome (APVS) is a rare congenital condition of uncertain etiology often associated with tetralogy of Fallot (TOF). First described by Cheevers in 1847, malformations include an absent or dysplastic pulmonary valve, annular pulmonary stenosis with regurgitation, and aneurysmatic dilation of the main pulmonary artery. Historically, the majority of cases have been diagnosed postnatally as a result of dilated pulmonary arteries compressing bronchial airways, with a risk of perinatal death as high as 60%.1,2 Few studies have followed the antenatal course of fetuses affected by APVS, and no study to date has described intrapartum fetal death.

Case Reports

Our first case was a nulliparous 16-year-old woman with abnormal fetal cardiac findings on an 18-week ultrasound. Fetal echocardiogram at 20 weeks visualized a nubbin-like pulmonary valve with a small 2.5-mm annulus, mild pulmonary stenosis with severe pulmonary regurgitation, enlarged main and branch pulmonary arteries (6 to 7 mm), mild right ventricular hypertrophy (RVH) and dilation, and an absent ductus arteriosus (DA). Subsequent echocardiograms displayed enlarging pulmonary arteries (9 to 11 mm), one small perimembranous and one midmuscular ventricular septal defect (VSD), and normal aortic architecture. Evaluation of the fetus included a normal karyotype (46, XX). The patient had no significant past medical history, and the fetus had no extracardiac abnormalities.

Labor was induced at 38 weeks and 6 days for intrauterine growth restriction [estimated fetal weight (EFW) 2620 g (< 10th percentile); 2 weeks prior the EFW = 2524 g (21st percentile)]. Doppler studies and amniotic fluid index (AFI) were in the normal range. Cervidil (Forest Pharmaceuticals, St. Louis, MO) (10 μg) was placed vaginally, followed by a standard oxytocin...
infusion. Nubain (APP Pharmaceuticals, Schaumburg, IL; nalbuphine hydrochloride) and promethazine were given for patient discomfort at midnight. After the medications were administered, there was decreased fetal heart rate (FHR) variability on the external fetal monitor (EFM) but no decelerations were noted, findings commonly seen in the FHR after Nubain administration. At 2:50 AM, the patient was disconnected from the EFM for 10 minutes to void. Upon reconnection, no FHR could be detected. Ultrasound showed fetal cardiac asystole. The cervix was 1 to 2 cm dilated, 50% effaced. Fetal membranes were intact. There was no cord prolapse. The patient had no vaginal bleeding or abdominal pain, and no contractions were palpable or demonstrated on the monitor. An emergency cesarean delivery was performed under general anesthesia, and a still-born infant female was delivered. There was no nuchal or body bleeding or abdominal pain, and no contractions were palpable or demonstrated on the monitor. An emergency cesarean delivery was performed under general anesthesia, and a still-born infant female was delivered. There was no nuchal or body delivery was performed under general anesthesia, and a still-born male infant was delivered in the vertex position. There was no nuchal or body cord and no evidence of a placental abruption. The birth weight was 3282 g. Aggressive resuscitation efforts were abandoned after 30 minutes. Cord venous blood gas was pH 7.31/pCO₂ 47/pO₂ 41. The 491-g placenta (75th to 90th percentile) contained intervillous fibrin deposition and a three-vessel umbilical cord. Cytogenetic analysis detected normal male karyotype 46, XY. The patient desired no autopsy to be performed.

**Discussion**

Improvements in obstetric ultrasound have allowed for earlier and more precise prenatal diagnosis of APVS. Recent studies have diagnosed APVS at 11 to 13 weeks’ gestation and have found associations with an increased first-trimester nuchal translucency, congenital diaphragmatic hernia, nonimmune hydrops, polyhydramnios, and the chromosomal 22q11 microdeletion. Varying presentations of APVS have also been documented. APVS is most commonly associated with TOF and often accompanying VSD. Regurgitant flow results in volume overload of both ventricles, especially in fetuses with a patent DA. The severe chronic volume overload makes this combination incompatible with fetal life. APVS with an absent DA (10 to 20% of these patients) may allow the fetus to reach the second trimester. In these fetuses, the pulmonary trunk and branches are often severely dilated, as in our first case. The rarer variant is APVS with tricuspid atresia, an intact interventricular septum, saclike dilation of the right ventricle, and a patent DA. This variant, due to increased aortic flow, results in more aortic and less pulmonary artery branch dilation. Some features of this condition are noted in our second case.

The detection of structural details by ultrasound has also been shown to have prognostic significance. Fetuses with hydrops or larger right pulmonary artery volume and annulus size have been linked with poorer prognosis. Infants with an intact interventricular septum and/or diagnosed after 6 months of life have a more benign prognosis than those with a VSD or who develop respiratory symptoms in infancy. The more frequent variant of APVS may also have slightly better outcomes.
Consequently, precise sono graphical evaluation of the fetal heart and vessels in APVS may be of prognostic benefit, as suggested in a recent study on the use of three- and four-dimensional ultrasonography in APVS. 

The intrapartum monitoring and clinical findings in our cases suggest acute fetal cardiac events when the patients were in early labor. The cause may have been electrophysiological as there were no findings to suggest progressive mechanical cardiac failure prior to labor and no evidence to suggest other common causes of intrapartum fetal death, such as abruptio placentae, cord accident, tetanic uterine contraction, or ongoing hypoxia/acidaemia. The advantages and timing of early amniotomy and fetal scalp electrode placement to more accurately monitor and intervene in the labor of fetuses with APVS is a question to consider.

Acute intrapartum fetal demise with APVS has not been previously reported and further adds to the well-accepted poor prognosis of affected fetuses. Larger studies of the intrapartum course of fetuses affected by APVS are needed to determine if factors that may increase sudden fetal death exist. In the meantime, in-depth parental counseling with maternal–fetal medicine and pediatric cardiology should include the possibility of acute fetal cardiac events in labor and the still uncertain fetal benefit of emergency cesarean delivery.

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