

# INTRAUTERINE DIAGNOSIS OF CORONARY ARTERY FISTULA: CASE REPORT



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**Introduction:** Coronary artery fistula (CAF) represent abnormal communications between a coronary artery and a cardiac chamber, great vessel, or the coronary sinus. Prenatal diagnosis of isolated CAF is extremely rare. We report a case of a large symptomatic CAF diagnosed at 21 weeks' gestation and surgically treated at neonatal period.

**Case:** A 31-year-old woman was referred for fetal echocardiography at 21 weeks' gestation due to gestational diabetes. Echocardiography showed normal segmental anatomy. There was right ventricular dilatation and moderate tricuspid insufficiency at the initial scan. On detailed examination, the left coronary artery was markedly dilated and color flow Doppler examination revealed the draining of the circumflex coronary arterial blood into the right ventricle just below the septal leaflet of tricuspid valve posteriorly (Figure 1 and 2). High velocity bidirectional flow, characteristic of a coronary fistula was detected. Follow-up visits were scheduled at 25, 29 and 35th weeks of gestation. The size of the fistula increased during follow-up. A female infant was delivered at 37th gestation weeks via elective Caesarean section with a birth weight 2,56 kg. A postnatal transthoracic echocardiography with 2-dimensional imaging, pulsed Doppler, and color-flow mapping demonstrated what appeared to be a giant proximal circumflex coronary artery to right ventricular fistula (Figure 3). Patient developed congestive cardiac failure soon after birth despite adequate medical treatment and required surgery for fistula at 17 days after birth. The patient was returned to the intensive care unit in satisfactory condition. Mechanical ventilatory support was weaned, and the patient was extubated on the fourth postoperative day. Follow-up echocardiogram following the procedure showed improved forward flow in the descending aorta with decreased RV size. She was discharged home 8 days after surgery. She appeared well at monthly follow-up appointments, with adequate weight gain.

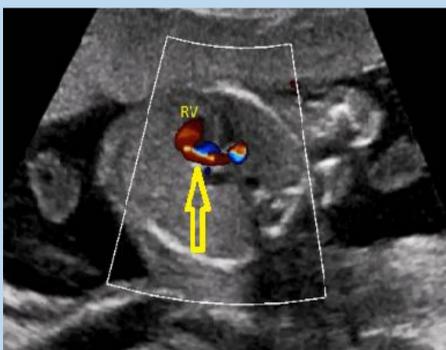


Figure 1. Prenatal view of coronary artery fistula at the 21 weeks.

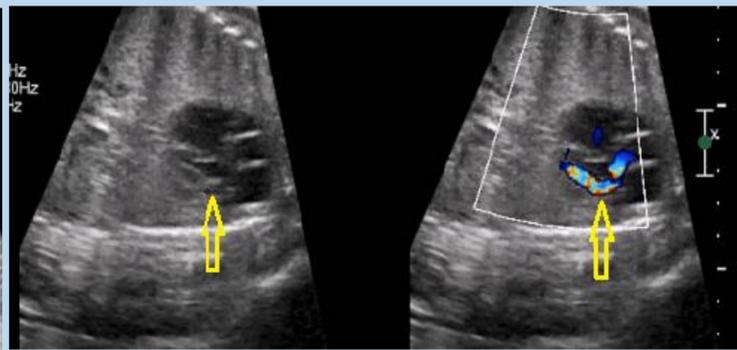


Figure 2. Prenatal view of coronary artery fistula at the 29 weeks

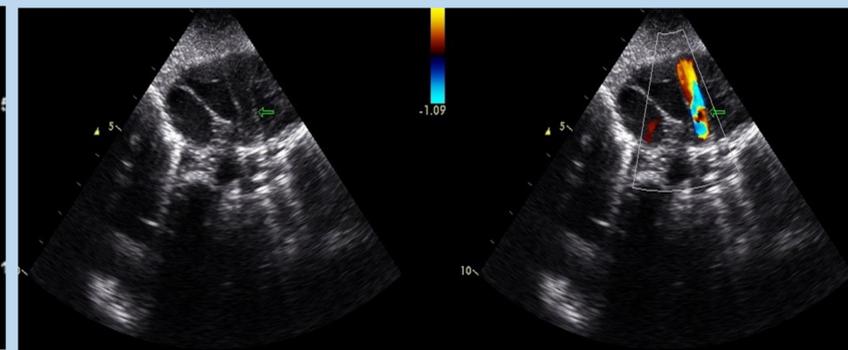


Figure 3. Postnatal view of coronary artery fistula

**Discussion:** Coronary artery fistula is observed in 0.002 % of the general population and accounts 0.08–0.4% among all congenital heart diseases. Majority of coronary artery fistulas are congenital and may occur due to persistence of sinusoidal connections between the lumens of the primitive tubular heart that supply myocardial blood flow in the early embryologic period. Coronary artery fistulas can be diagnosed accurately intrauterine period, but it is unclear whether these fetuses are at higher risk for developing symptoms earlier after birth. CAFs can be seen on color Doppler from 31th weeks onwards in the normal fetus. In case of dilated coronary artery it can be seen more early period. If the signs of heart failure (such as cardiomegaly, hydrops) appear earlier and progress during the fetal period, some authors recommend termination of a pregnancy. It is unusual for coronary artery fistulas to produce symptoms in the neonatal period or early infancy. Coronary artery fistula can manifest with myocardial ischemia resulting from coronary steal or with congestive heart failure due to substantial systemic-to pulmonary shunt. At the literature five of the nine cases (55.6%) presented with symptomatic congestive heart failure in neonatal period. It's recommended to close CAFs early in symptomatic patients or in asymptomatic patients with large fistulas originating from the proximal segments of coronary arteries. These fistulas are usually closed either in the catheterization laboratory or surgically.

**Conclusion:** Coronary artery fistulas can be diagnosed accurately during fetal life. Some babies may develop congestive cardiac failure soon after birth requiring early treatment. The correct prenatal diagnosis enabled close perinatal follow-up, prompt clinical evaluation without diagnostic delay and optimal management.