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Giant Coronary Artery Fistula: Prenatal Diagnosis, Newborn Manifestation

Fístula arterial coronaria gigante: diagnóstico prenatal y manifestación neonatal

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A rare case of a large left coronary artery fistula is presented. Prenatal echocardiography at our institution since 22 week of gestation was significant for an atypical diastolic high flow communication from the ascending aorta (Ao asc) to the right ventricle (RV) (Figures 1A-1B). There were no signs of ventricular dysfunction during prenatal follow-up.

A female neonate was delivered at 38 weeks of gestation. Already during first hours of life progressive tachy-/dyspnea developed, and noninvasive ventilation and oxygen supplementation was initiated. Prenatal diagnosis was confirmed by postnatal echocardiography (Figures 1C-1D). Angiography (Figures 2A-2D) had shown a giant coronary fistula arising from the dilated left aortic sinus at the site of the left coronary artery origin, represented by a tunnel-like structure with its narrowest part at the proximal end 4 mm and distally enlarged to 12 mm, draining into the right ventricle apex. Right ventricular dilatation but without ventricular dysfunction and no signs of myocardial ischemia were found. Due to progressive respiratory failure from a large left-to-right shunt, surgical fistula closure at both ends was performed.

Coronary artery fistulas may originate from left, right or both coronary arteries and may drain into any heart chamber. Clinically, they usually manifest with myocardial ischemia resulting from coronary steal or with congestive heart failure due to substantial systemic-to-pulmonary shunt. Although such early postnatal presentation (within hours) due to hemodynamic significance is extremely rare. The correct prenatal diagnosis, such as in our case, enabled close perinatal follow-up, prompt clinical evaluation without diagnostic delay and optimal management, including early intervention.

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