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The Window Ductus: a Rare Finding, and the Importance of Perinatal Echocardiography

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Background

Fetal echocardiography provides important data for postnatal management of complex cardiac defects. There must be appropriate interpretation of fetal images to allow correlation with neonatal findings in order to prevent adverse outcomes associated with unclear interpretation. We present a case of an unusual form of the ductus arteriosus termed "window ductus," and hope to highlight the importance of early recognition.

Embryologically and by location the window ductus does not meet criteria for an aortopulmonary window, and yet does not constitute a true ductal arch. Rather it is a side-to-side communication between the base of the left pulmonary artery and descending aorta. This anomaly has most often been found in the setting of abnormal cardiac anatomy where surgical intervention is warranted. It is typically described as wide, short, and without flow restriction and it was, therefore, hemodynamically significant. Typically, the ductus arises from the arterial duct which is initially a bilateral structure that develops from the 6th aortic arch. The right side normally atrophies, whereas the left sided arterial duct goes on to develop into the ductus arteriosus, connecting the aorta and pulmonary artery.

Histologically the ductus is composed of smooth muscle in the media of the vessel and an internal elastic lamina with intimal cushions. In regards to the "window ductus," it is unclear if it originates in the same manner, or is composed of the same tissue as a true ductus, but it is a similar origin is suggested.

While the "window ductus" defect is controversial in that some deny its existence, several case reports exist highlighting this finding on fetal echo. In some of these cases, failure to recognize the "window ductus" contributed to increased mortality in cases of surgical intervention.

Case Report

The patient presented as a 22 week fetus due to advanced maternal age, maternal cardiomyopathy, and an abnormal 4-chamber view on the fetal anatomical ultrasound. Fetal images were consistent with polyvalvar disease with redundancy of the mitral, tricuspid and aortic valves. There was an extremely abnormal pulmonic valve that was thickened, stenotic, and regurgitant due to relative immobility of the cusps, creating a narrow, tunnel-like orifice and post-stenotic dilated main pulmonary artery. The right ventricle was hypertrophic and stiff with decreased systolic function; the left ventricle appeared normal.

There was concern for a ductal-dependent lesion given the narrow diameter of the effective pulmonary valve orifice. The ductal arch was not well-identified antenatally, but assumed to be patent due to a normal Doppler flow.

The infant was born at 35 weeks due to maternal pre-eclampsia, and significantly cyanotic.

Present an unusual cardiac defect

Postnatal echo confirmed the aforementioned findings, with the additional finding of a direct communication between the descending aorta and left pulmonary artery. The communication had the appearance an aortopulmonary (AP) window, although in the wrong location, making this connection embryologically different than the usual AP window (neural crest abnormalities) or PDA (abnormality of the sixth aortic arch).

Results

The patient was evaluated with transillumination of the chest wall after delivery. The lesion was isolated and approached via the left chest wall. After delivery prostaglandin therapy was initiated. The infant was stable at our facility and then urgently transferred to a tertiary center offering surgical repair. At surgery, the lesion was identified as a window ductus, was separated and patched.

Discussion

While the ductal window nomenclature is controversial and reported sparingly in the literature, it’s existence has important implications for management. If the window is derived from ductal tissue, it should respond to an infusion of prostaglandins in the same manner. Applying this knowledge would be necessary in cases of ductal dependent congenital heart disease, to hopefully prevent window ductus closure and precipitous deterioration. Regardless, these patients would benefit from being at a surgical center where management of the window could be performed while correcting other cardiac defects.

Conclusions

This case underscores the importance of appropriately identifying cardiac abnormalities on pre-natal ultrasound anatomy, and even more the importance of a pre-natal echo for proper identification of cardiac abnormalities. It is also essential to identify the ductal arch on every fetal echocardiogram to allow appropriate planning in the event of ductal-dependency. Early detection of abnormalities in this infant allowed for delivery at a tertiary care center with access to adequate resources. As such, prostaglandin therapy was administered in a timely manner, and post-natal echo was obtained almost immediately after delivery.

Pediatric Cardiology was involved during pregnancy, and was readily available at time of delivery. Further, the infant was able to be transferred to a tertiary surgical center for further management after initial stabilization. Awareness of this unusual ductal lesion, likely assisted and in surgical management, and helped prevent mortality.

References


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