

iMAGE

LETTERS TO THE EDITOR

Prenatal Visualization of Persistent 5th Aortic Arch Artery

The embryonic arches develop in the fourth and fifth gestational weeks with the corresponding pharyngeal arch arteries connecting the aortic sac with the dorsal aorta. The fourth arch forms the aorta; the left sixth arch forms the left pulmonary artery and duct. The fifth aortic arch is a transient developmental structure in humans as there is no 5th pharyngeal arch; indeed, its existence has, in the past, been disputed (1). There are some reports of persistence of the 5th embryonic arch artery (2,3), but it is rare. This may, in part, be due to under-recognition and misdiagnosis (2). Further insight into the development and possible persistence of the 5th arch arteries have been gained from 2 recent studies that have described the presence of vascular channels thought to represent putative 5th arch arteries in mice (4,5). These studies are of particular interest because cardiac morphogenesis in mice closely resembles that of humans. Two forms of persistent 5th arch are proposed: a systemic-to-systemic connection, and a systemic-to-pulmonary connection. It may be associated with other cardiac pathology. The latter form requires that it be differentiated with a short wide arterial duct (window duct) or an aortopulmonary (AP) window.

We describe the imaging and outcomes in 2 children in whom a vascular channel was diagnosed prenatally and likely represented remnants of the 5th aortic arch artery. At 22 weeks' gestation, fetus 1 was referred for echocardiography, which showed normal situs and concordant connections with critical pulmonary stenosis. The pulmonary valve was dysplas-

tic with moderate regurgitation and flow reversal in the ductal arch. The aortic arch was thought normal and left sided during serial scans. The right ventricle was hypoplastic, but the eventual size was compatible with a biventricular circulation.

Postnatal echocardiography confirmed this diagnosis and, in addition, identified a broad communication between the aortic arch and pulmonary trunk that was interpreted as an AP window (Online Video 1). Similar images had been obtained prenatally but not recognized as unusual (Fig. 1A). A substantial communication between the aorta and the pulmonary trunk, adjacent to the pulmonary end of the duct, was identified at surgery. The aortic arch terminated at the communication, and the descending aorta was supplied via the duct, suggesting the alternative diagnosis of a persistent 5th arch with a Type A interrupted aortic arch. Repair of the arch was performed during a long and complex procedure, and the infant required post-operative venoarterial extracorporeal membrane oxygenation support. Unfortunately, ventricular function did not recover, and support was withdrawn after 7 days. Postmortem could not confirm AP window or arch discontinuity because of prior surgery. Review of imaging at our institutional morbidity and mortality conference concluded that echocardiographic features were compatible with the intraoperative diagnosis (Online Video 2).

Fetus 2 was referred at 21 weeks' gestation with sonographic findings of normal situs and concordant cardiac connections, but with a broad connection between the pulmonary artery to the underside of the aortic arch, proximal to the origin of the arch arteries (Fig. 1B, Online Videos 3 and 4). The images were reminiscent of those examined retrospectively from fetus 1 and the abnormal connection suspected to represent a remnant of the 5th arch artery or an unusual

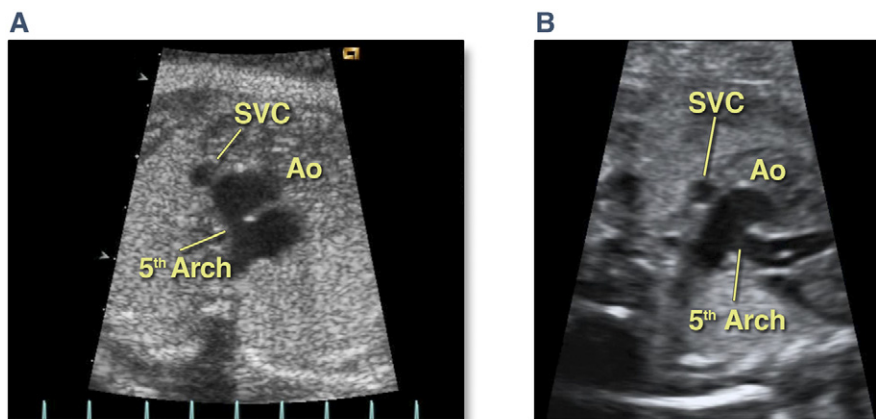


Figure 1. Fetal Echocardiogram

Three-vessel and tracheal view. (A) Case 1. This transverse view, high in the fetal chest, demonstrates the broad connection (the 5th arch artery) between the arterial duct and transverse aortic arch. (B) Case 2. This oblique view shows the insertion of the 5th arch artery into the transverse aorta. This site of insertion is more proximal than that normally seen when the ductal arch inserts into the descending aorta. See accompanying Online Videos 1, 2, 3, and 4. Ao = aorta; SVC = superior vena cava.

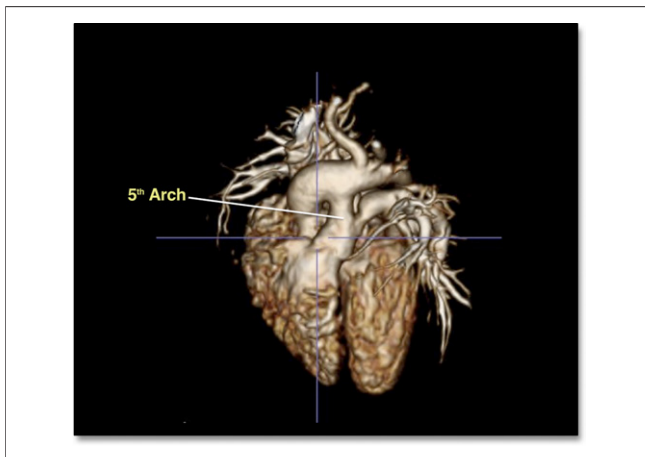


Figure 2. Postnatal CT Angiogram of Case 2

This shows the 5th arch artery connecting the pulmonary trunk and aorta. The communication with the aorta is proximal to the origin of the head and neck vessels. CT = computed tomography.

AP window. The baby was born at term in good condition, and postnatal echocardiography was consistent with prenatal diagnosis. Computed tomography angiography (Fig. 2) was consistent with the remnant of a persistent 5th arch artery, and successful surgery was performed.

The features in case 1 suggested the likely diagnosis of persistent 5th arch rather than an AP window because the AP communicating artery was connecting distal to the origin of the pulmonary arteries. In an AP window, this would be positioned along the pulmonary trunk and bifurcation of the pulmonary arteries. A high AP window may extend to the origin of the right pulmonary artery, but the window is not in the form of a channel with a length resembling a vessel. Moreover, the characteristic wall structure of the arterial duct identified at surgery in our case supported the likely origin of the vessel as a remnant of the fifth arch artery. For the second case, the differentiating point from an arterial duct was the location of the communicating artery connecting the aorta proximal to the origin of the head and neck vessels; an arterial duct would communicate distal to the arch arteries.

Both fetuses reported here likely had vascular channels that represented remnants of a 5th aortic arch artery. Failure to diagnose aortic interruption with persistence of the 5th arch arterial channel in our first case resulted in prolonged surgery and subsequent demise. Lessons learned from this case led to appropriate considerations in the second fetus, confirmation by postnatal imaging, and appropriate surgery that resulted in a good outcome.

Although postnatal reports of a persistent 5th arch artery exist (2), we have demonstrated that fetal diagnosis is possible and that additional imaging, such as computed tomography angiography, is useful to confirm prenatal diagnosis after birth and aid its successful surgical management.

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Repeat Routine Differential Pulmonary Blood Flow Measurements in Congenital Heart Disease by MR

Interstudy Variability and Benchmark of a Clinically Relevant Change

Phase-velocity magnetic resonance (PV-MR) quantifies differential pulmonary blood flow as accurately as the previous gold standard lung perfusion scintigraphy in patients with a single pulmonary blood source supplied by a subpulmonary ventricle (1–3). Therefore, in routine clinical practice, PV-MR has taken on the role of quantifying differential pulmonary blood flow in congenital heart disease (4). However, the interstudy variability and a benchmark of a clinically relevant change of repeat routine differential pulmonary blood flow measurements in congenital heart disease using PV-MR are still unknown.

To test the interstudy variability, we studied 80 consecutive routine clinical cases without intervention or pathological event of the pulmonary branch arteries between 2 consecutive PV-MR measurements (control group). Therefore, in this group, no change of the measured differential pulmonary blood flow ratio was to be expected. This group consisted of 56 cases with repaired tetralogy of Fallot, pulmonary atresia, or truncus arteriosus communis, 7 cases with atrial switch operation, 6 cases with arterial switch operation, 5 cases with Ross operation, and 6 cases with other congenital heart defects. Mean age was 23.2 ± 9.8 years at the time of the first measurement. Mean time between the 2 measurements was 1.2 ± 0.6 years.

To establish a benchmark of a clinically relevant change of repeat routine differential pulmonary blood flow measurements using PV-MR, we studied 13 consecutive routine clinical cases with an explicit unilateral intervention or morphological change to 1 of the pulmonary branch arteries between 2 PV-MR measurements (intervention group). Therefore, in this group, a clear change of the measured differential pulmonary blood flow ratio was to be expected. It is important to note that in our center, PV-MR measurements are not