Persistent 5th Aortic Arch

with Coexisting Discontinuous Pulmonary Arteries, Double-Outlet Right Ventricle, and Complete Atrioventricular Septal Defect

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A diabetic mother's newborn female infant, delivered at 34 weeks of gestation at a birth weight of 4.8 lb, was transferred to our cardiac intensive care unit for treatment. The neonate had a heart rate of 141 beats/min, a respiratory rate of 33 breaths/min, and an oxygen saturation of 100% on room air. She had been diagnosed prenatally with a complete unbalanced atrioventricular septal defect and double-outlet right ventricle. A transthoracic echocardiogram confirmed the diagnosis of complete unbalanced, right-dominant atrioventricular septal defect, double-outlet right ventricle with the aorta to the right of the pulmonary artery (PA), a subaortic ventricular septal defect, and moderate-to-severe pulmonary stenosis. Additional echocardiographic evaluation revealed a left aortic arch with a persistent 5th aortic arch, and the left PA was found to be discontinuous with the main PA (Figs. 1–4). Results of cardiac catheterization confirmed the diagnosis of a persistent 5th aortic arch and a left PA arising from the ascending aorta (Fig. 5).

Comment

A persistent 5th aortic arch with an ipsilateral 4th arch is a rare anomaly.^{1,2} In addition, anomalous origin of the left PA from the ascending aorta is unique. From an embryologic standpoint, the anomalous left PA is theorized to represent the persistence of bilateral 5th aortic arches coupled with agenesis of the left 6th arch.³ The

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Fig. 1 Transthoracic echocardiogram (long-axis view from the suprasternal notch) shows the persistent 5th aortic arch (A) and the origin of the left pulmonary artery (B) from the ascending aorta (C).

Real-time motion image is available for Figure 1.

Fig. 2 Color-flow Doppler echocardiogram (long-axis view from the suprasternal notch) shows the persistent 5th aortic arch (A) and the origin of the left pulmonary artery (B) from the ascending aorta (C). There was turbulent flow from the ascending aorta into the left pulmonary artery, and diastolic flow reversal in the proximal descending aorta.

Real-time motion image is available for Figure 2.

Fig. 3 Transthoracic echocardiogram (modified long-axis view from the suprasternal notch) reveals the persistent 5th aortic arch (A) and left aortic arch (B). The origin of the left pulmonary artery from the ascending aorta (C) is seen.

Real-time motion image is available for Figure 3.

Fig. 4 Color-flow Doppler echocardiogram (modified long-axis view from the suprasternal notch) reveals the persistent 5th aortic arch (A) and left aortic arch (B). The origin of the left pulmonary artery from the ascending aorta (C) is seen. There was turbulent flow from the ascending aorta into the left pulmonary artery, and diastolic flow reversal in the proximal descending aorta.

Real-time motion image is available for Figure 4.

coexistence of these arch anomalies has been reported only once⁴; however, the constellation of cardiac anomalies presented here has not been reported. Similar to a previous report,² our patient's 5th arch was sensitive to prostaglandin E₁ therapy (Fig. 6). She subsequently underwent unifocalization of the left PA and placement of a right modified Blalock-Taussig shunt.

References

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Fig. 5 Angiogram of the ascending aorta (anteroposterior view with ascending aortic injection) shows the persistent 5th aortic arch (A), and the anomalous left pulmonary artery (B) arising from the ascending aorta (C).

Fig. 6 Transthoracic echocardiogram (long-axis view from the suprasternal notch) shows a membrane-like appearance to the involution of the 5th aortic arch (arrow) upon the discontinuation of prostaglandin therapy.

* = left pulmonary artery

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