Isolated Left Subclavian Artery, Complete Atrioventricular Block, and Tricuspid Atresia in a Neonate

Isolated left subclavian artery is one of the rarer aortic arch anomalies. It has been associated with other congenital heart diseases, typically tetralogy of Fallot, double-outlet right ventricle, and atrial and ventricular septal defects. Its significant clinical implications include a left-to-right shunt from the vertebrobasilar system, which causes pulmonary overcirculation and subclavian steal. We present an unusual case of a premature infant who was diagnosed prenatally with congenital complete atrioventricular block and tricuspid atresia and was found to have an isolated left subclavian artery postnatally. The patient underwent implantation of a permanent single-chamber epicardial pacing system. To our knowledge, this combination of lesions has not been reported—and in our case, it influenced our surgical planning. (Tex Heart Inst J 2016;43(6):546-9)

Isolated left subclavian artery (LSA), an infrequently encountered aortic arch anomaly, can be seen in cases of right-sided aortic arch. It can be explained by embryologic involution of the distal left dorsal aorta, which creates a connection between the LSA and the pulmonary artery (PA) through the ductus arteriosus. We report the unusual case of a neonate with tricuspid atresia, isolated LSA, and congenital complete atrioventricular block (AVB). This unusual anatomy included a natural systemic-to-PA shunt, which influenced our surgical planning.

Case Report

In September 2014, a female infant was delivered by emergency cesarean section at 32 weeks’ gestation because of a fetal heart rate of 30 beats/min. Before birth, she had been diagnosed with tricuspid atresia, normally related great arteries, a large ventricular septal defect, and congenital complete AVB. Upon delivery, the 2-kg infant was intubated. Epinephrine, given through the endotracheal tube, transiently increased her heart rate to 60 beats/min. An umbilical venous line was placed, an epinephrine drip was started, and cardiopulmonary resuscitation was performed on the way to the operating room. Because of the neonate’s size and clinical status, temporary pacing wires were placed on the surface of the ventricle and the pacing rate set at 120 beats/min.

The patient was transported to the neonatal intensive care unit for stabilization, was given surfactant, and was placed on continuous positive airway pressure. A complete echocardiogram confirmed the diagnosis of tricuspid atresia with normally related great arteries (type IC), nonrestrictive atrial septal defect, ventricular septal defect, antegrade flow across the pulmonary valve, mild subpulmonary stenosis, and a right aortic arch with the suspicion of an isolated LSA arising from the PA through a ductus arteriosus (Fig. 1). Clinical findings of diminished left-arm pulses and a lower blood pressure in the left arm than in the right furthered this suspicion. A cardiac computed tomographic angiogram confirmed the diagnosis of right aortic arch with an isolated LSA arising from the ductus arteriosus off the main PA (Fig. 2).

When the patient was one month old, her respiratory status had stabilized, and she had gained sufficient weight to undergo catheterization. There, test-occlusion of the LSA was performed, with evaluation of the antegrade flow across the pulmonary valve, PA pressures, and cerebral perfusion. During angiography, a vast network...
of collateral vessels from the vertebral arteries was seen supplying the LSA (Fig. 3).

Balloon occlusion of the LSA caused no change in the PA pressures; however, the patient’s cerebral oxygen saturation level decreased, as documented on near-infrared spectroscopy. Because the LSA arose from the PA, we thought that this drop in cerebral saturation was due to occluded ductal flow, which in turn decreased pulmonary venous return and cardiac output. Therefore, we decided not to occlude the LSA. The patient was taken to the operating room, and a permanent single-chamber (ventricular) epicardial pacing system was implanted. At 5 months of age, she was referred for a bidirectional Glenn procedure with reimplantation of the aberrant LSA to the base of the left common carotid artery. As of October 2016, the patient was doing well, and she will undergo a Fontan procedure in the future.

**Discussion**

To our knowledge, the combination of isolated LSA from the ductus arteriosus, tricuspid atresia, and congenital complete AVB has not been reported.
Isolated LSA from the ductus arteriosus has been described in patients with and without congenital heart disease and has frequently been associated with right aortic arch.\(^1\) Approximately 60% of such patients have tetralogy of Fallot, double-outlet right ventricle, atrial and ventricular septal defects, or a combination of these.\(^2\)

Embryologically, the left 7th intersegmental artery migrates cephalad to the level of the ductal arch, after which the distal left dorsal aorta involutes, isolating the LSA from the aortic arch but maintaining its connectivity to the PA through the ductus arteriosus.\(^3,4\) Thus, the LSA receives blood flow retrograde through the vertebral system, leading to a left-to-right shunt from the vertebralbasilar system to the PA.\(^5\)

After birth, a natural physiologic decrease in pulmonary vascular resistance occurs, and reversal of flow through the LSA results in subclavian steal, reversing blood flow from the brain and increasing blood flow into the lungs.\(^6,7\)

Computed tomographic angiography noninvasively enables extensive anatomic and functional evaluation of aortic arch anomalies. In our patient, the results confirmed our suspicion of the isolated LSA off the PA.\(^8\)

In these rare cases, cardiac catheterization can reveal collateral blood supply between the left and right vertebral arteries and retrograde flow through the isolated LSA. Catheterization also enables the evaluation of pulmonary vascular resistance, which is crucial to surgical decision-making.\(^9\)

The management of an isolated LSA often depends on the patient’s associated congenital heart defects, symptoms, and physiologic status. Ductal ligation and reimplantation of the LSA, as well as coil occlusion of the ductus arteriosus, have yielded good results.\(^10\)\(^11\) The importance of early recognition of the isolated LSA cannot be overstated, because this knowledge can influence the choice of surgical procedure—in our patient, a Blalock-Taussig operation could be avoided because of this natural systemic-to-PA shunt.

This abnormal-yet-natural shunt helped to maintain reliable pulmonary blood flow in our patient. However, substantial flow reversal from the isolated LSA into the PA can create risks of vertebralbasilar insufficiency and pulmonary hypertension that can jeopardize the future management of a congenital heart disease. The cardiac catheterization findings helped our decision-making process by revealing our patient’s normal pulmonary vascular resistance. Accordingly, we performed no intervention on the isolated LSA at the time.

The patient also had complete AVB. This conduction abnormality prompted an emergency premature delivery and necessitated permanent pacemaker implantation. Like isolated LSA, complete AVB has been associated with various congenital heart diseases—in as many as 50% of patients, in one study.\(^12\) We found one description of complete AVB with double-outlet right ventricle, moderate hypoplasia of the left ventricle, and an isolated LSA,\(^13\) and a single report of complete AVB associated with tricuspid atresia.\(^14\) However, to our knowledge, neither the precise combination in our patient nor any particular genetic association between tricuspid atresia and congenital complete AVB has been reported.

### References