

Anomalous Origin of the Right Coronary Artery

from the Pulmonary Artery in a Neonate with Turner Syndrome and Aortic Arch Hypoplasia

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Anomalous origin of the right coronary artery from the pulmonary artery, a rare congenital cardiac defect, is typically not diagnosed during infancy. On the other hand, Turner syndrome is usually diagnosed early, and it is classically associated with bicuspid aortic valve and aortic coarctation. Individuals with Turner syndrome are also at increased risk for coronary artery anomalies. We present a case of anomalous right coronary artery from the pulmonary artery in a week-old neonate who also had Turner syndrome, patent ductus arteriosus, transverse aortic arch hypoplasia, and impaired ventricular function. Prostaglandin therapy through the ductus increased the patient's myocardial perfusion. Four months after corrective surgery, she was doing well. We discuss the reperfusion phenomenon in our patient's case, as well as other considerations in this combination of congenital defects. (Tex Heart Inst J 2019;46(3):225-8)

Key words: Abnormalities, multiple; cardiac surgical procedures; coronary vessel anomalies/diagnostic imaging/physiopathology/surgery; ductus arteriosus, patent/physiopathology; heart defects, congenital; infant, newborn; pulmonary artery abnormalities; treatment outcome; Turner syndrome/surgery/therapy

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Anomalous origin of the right coronary artery from the pulmonary artery (ARCAPA) is a rare congenital heart defect (estimated incidence, 1 in 50,000).¹ Turner syndrome (monosomy X) has classically been associated with bicuspid aortic valve and aortic coarctation; in addition, patients are at increased risk for coronary artery anomalies.²⁻⁴ We present the case of a newborn with Turner syndrome, bicuspid aortic valve, patent ductus arteriosus (PDA), and transverse aortic arch hypoplasia who had impaired ventricular function and was found to have ARCAPA.

Case Report

In November 2016, a female neonate prenatally diagnosed with Turner syndrome and congenital heart disease was born at 37 weeks' gestation (weight, 2.5 kg; length, 46 cm). She was admitted to our Neonatal Intensive Care Unit. Postnatal transthoracic echocardiograms showed a mildly hypoplastic proximal transverse aortic arch with distal narrowing and tortuosity. The aortic isthmus appeared mildly hypoplastic in the presence of a large PDA. A bicuspid aortic valve with horizontally oriented commissures was noted. The dimensions of her aortic annulus, aortic root, and ascending aorta were normal. Bilateral superior venae cavae (SVC), a small perimembranous ventricular septal defect, and a patent foramen ovale were seen. An unexpected finding was an apparent ARCAPA originating from the right pulmonary sinus (Fig. 1). An anomalous accessory coronary branch also originated from the left side of the mid main pulmonary artery (PA) (Fig. 2). Computed tomographic angiograms showed an 8-mm ascending aorta (normal range), a 3 × 4-mm transverse arch (mildly hypoplastic), a 5-mm isthmus (normal range), a 4-mm PDA, and post-stenotic dilation of the mid descending aorta to 9 mm (Fig. 3). The aortic diameter at the level of the diaphragm was 7 mm (top of normal range). The anomalous connection between the RCA and PA could not be identified definitively as a coronary artery fistula or ARCAPA. The patient had palpable lower-extremity pulses with upper- and lower-extremity systolic blood pressure gradients of 10 mmHg. Her urine output was adequate. Nonurgent surgical repair was planned, and her condition was observed.

When the patient was one week old, echocardiograms showed decreased biventricular systolic function, decreased peak descending aortic velocities, and a pulsatile abdominal aortic Doppler pattern without diastolic flow continuation. Flow through the PDA was right to left in systole and left to right in diastole. The myocardial

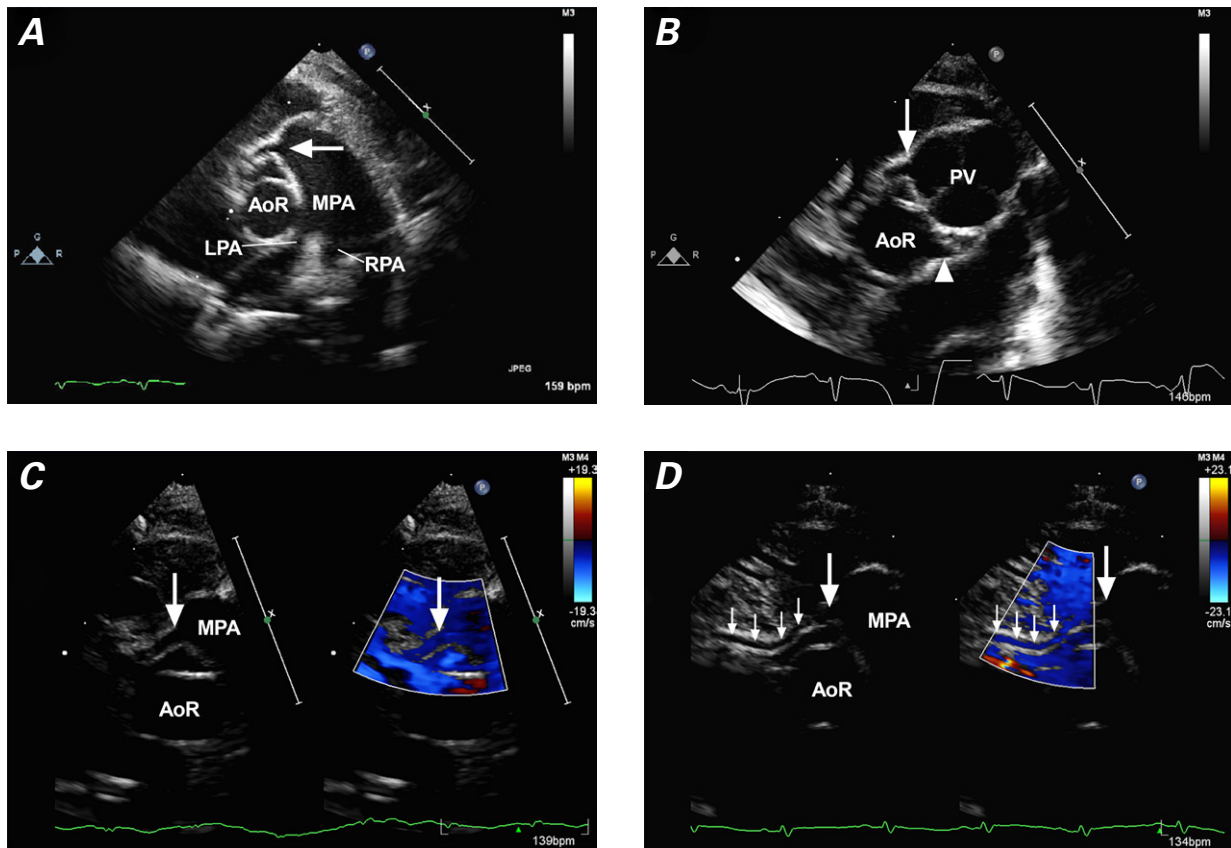


Fig. 1 Two-dimensional transthoracic echocardiograms show **A**) the main pulmonary artery and an anomalous coronary artery (arrow), and **B**) confirmation of an anomalous right coronary artery originating from the right sinus (arrow) of the pulmonary artery (*en face* view) and normal origin of the left main coronary artery from the left aortic sinus (arrowhead). Color-flow Doppler mode reveals **C**) antegrade flow at the origin of the anomalous right coronary artery (arrow), and **D**) antegrade flow along the length of the artery (small arrows) as it courses toward the right atrioventricular groove.

AoR = aortic root; LPA = left pulmonary artery; MPA = main pulmonary artery; PV = pulmonary valve; RPA = right pulmonary artery

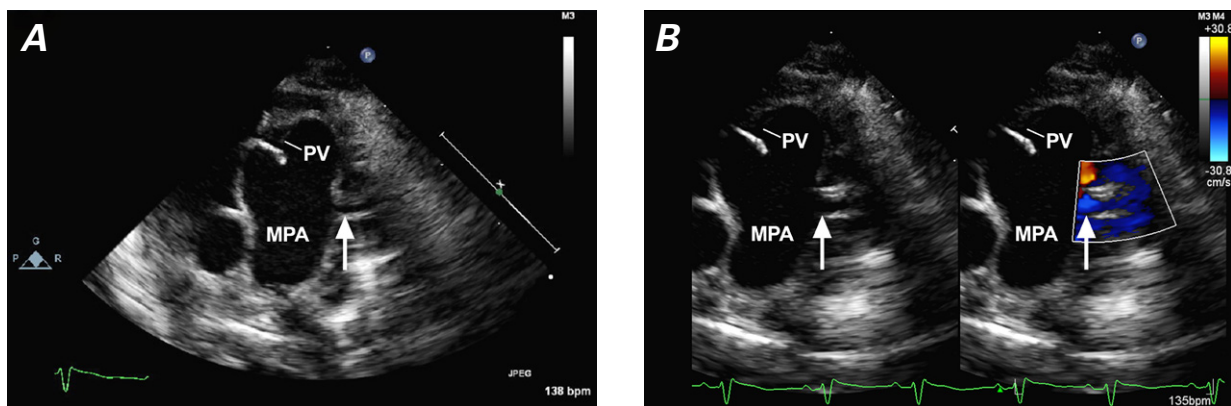


Fig. 2 Two-dimensional transthoracic echocardiograms show **A**) the origin of a small accessory coronary artery (arrow) from the left side of the mid main pulmonary artery (arrow) and **B**) antegrade flow within the accessory artery (color-flow Doppler mode).

MPA = main pulmonary artery; PV = pulmonary valve

dysfunction was thought to result from decreased pulmonary vascular resistance and PA pressures, leading to decreased RCA perfusion pressure. The patient's lower-extremity pulses were unchanged, her blood pressure

gradients remained stable, and her urine output was maintained.

To promote more antegrade flow in the suspected ARCAPA, we initiated continuous prostaglandin

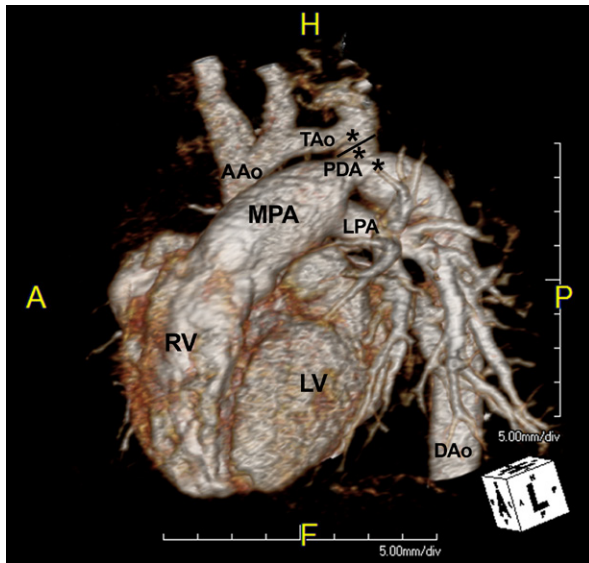


Fig. 3 Computed tomographic angiogram (3-dimensional reconstruction) shows a hypoplastic transverse aortic arch (TAo) with distal tortuosity (asterisks) and the aortic isthmus (black line). The coronary arteries are not shown.

AAo = ascending aorta; DAo = descending aorta; LPA = left pulmonary artery; LV = left ventricle; MPA = main pulmonary artery; PDA = patent ductus arteriosus; RV = right ventricle

therapy. Subsequent echocardiograms revealed that the PDA had enlarged to 6 mm, the diastolic left-to-right ductal velocities had decreased, and ventricular function had improved. Preoperative cardiac catheterization, performed to characterize the anomalous vessel, enabled us to diagnose ARCAPA conclusively (Fig. 4). A left-sided SVC draining to the coronary sinus was also confirmed. At age 13 days, the neonate underwent aortic arch repair and reimplantation of the RCA into the aorta. In addition, the anomalous accessory coronary branch from the mid PA was empirically ligated, to prevent late coronary steal. After a postoperative course complicated by capillary leak, lymphedema, and prolonged mechanical ventilation, the patient was discharged from the hospital in good condition on day of life 55. At 4 months of age, she was doing well without arch obstruction and with normal-appearing coronary anatomy.

Discussion

Turner syndrome is the most prevalent sex chromosome abnormality in females, seen in approximately 1 in 2,000 live female births.^{2,3} Patients with Turner syndrome are at elevated risk for congenital heart disease (incidence, approximately 40%).^{2,3,5} Associated left-sided obstructive lesions include bicuspid aortic valve, aortic coarctation, and aortic arch obstruction.^{2,3} Also notable is the association between this syndrome and congenital anomalies of the coronary arteries. In a study of 50 patients who had Turner syndrome,⁴ computed

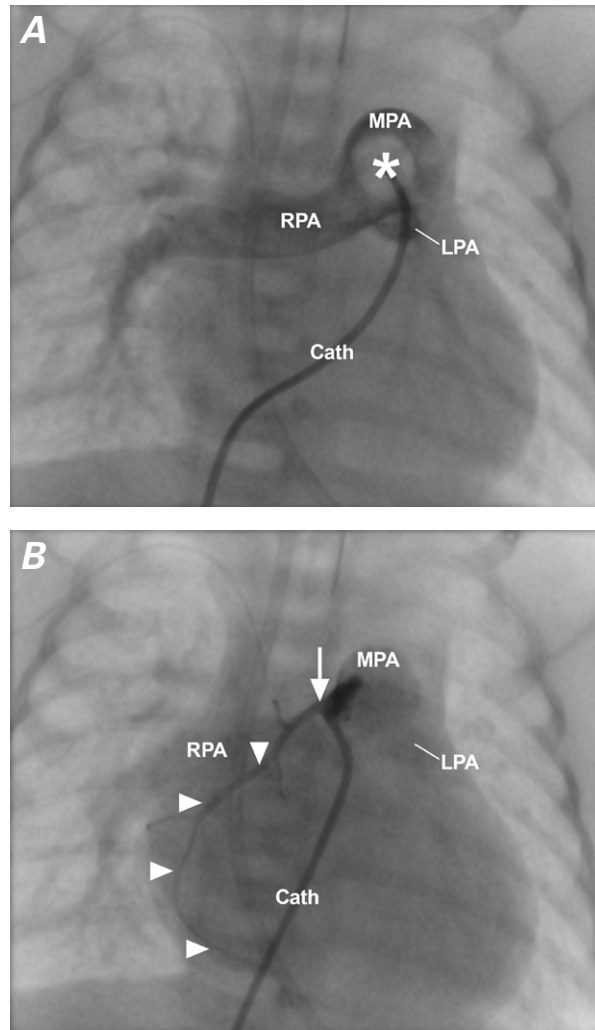


Fig. 4 Fluoroscopic angiograms conclusively diagnose anomalous origin of the right coronary artery from the pulmonary artery. **A)** The main, left, and right pulmonary arteries are normal; the inflated balloon catheter tip (*) is in the main pulmonary artery. **B)** Selective injection shows the anomalous right coronary artery originating from the right pulmonary sinus (arrow) with usual course through the right atrioventricular (AV) groove (arrowheads).

Cath = catheter; LPA = left pulmonary artery; RPA = right pulmonary artery

tomographic angiography showed that 10 had coronary anomalies, including 7 instances of absent left main coronary artery. In another study of 173 patients who had Turner syndrome,³ 2 had coronary anomalies: one was an anomalous left coronary artery arising from the PA (ALCAPA); the other, a left coronary artery-to-PA fistula. Outcomes of congenital heart surgery in patients with Turner syndrome have been mixed; they are reportedly at increased risk for prolonged hospitalization and death after repair of some defects.^{2,3}

The combination of Turner syndrome, aortic arch coarctation, and anomalous coronary artery has rarely been reported. In one case, a girl with Turner syndrome,

coarctation, and ARCAPA was diagnosed at 4 years of age; 2 years later, she underwent repair involving coronary reimplantation.¹ In another case, a 9-day-old infant with Turner syndrome was undergoing aortic arch repair and was found to have an RCA-to-PA fistula, which was proximally ligated at the PA.⁶ Although these cases reinforce the importance of accurately evaluating coronary artery anatomy preoperatively in patients with Turner syndrome, definitive diagnosis by means of noninvasive imaging may be difficult. In the case of an adult patient with Turner syndrome,⁵ echocardiograms showed 2 abnormal color-flow jets entering the PA but did not conclusively delineate the anatomy; in contrast, results of cardiac catheterization showed separate right and left coronary artery fistulas to the PA.

Because ARCAPA poses less risk of myocardial ischemia, congestive heart failure, and sudden death, patients are typically older at diagnosis than are those who have the more prevalent ALCAPA.¹ In previous reports, ARCAPA has been found in patients ranging in age from 6 weeks to adulthood, and 90% of diagnoses were in noninfants.¹ Our patient presented unusually early with depressed ventricular function, presumably due to decreased RCA perfusion pressure after a decrease in pulmonary vascular resistance. We thought that prostaglandin therapy would improve myocardial perfusion by increasing blood flow through the PDA, thus increasing PA pressures and antegrade flow through the ARCAPA. This result had been described in the presence of ALCAPA with PDA: robust blood flow through the ductus supports myocardial perfusion through the anomalous artery by elevating PA pressure.⁷ Increased PA pressure may either support antegrade flow through the anomalous artery or limit retrograde flow or coronary steal if there is collateralization. When a PDA is closed percutaneously or surgically, PA pressure may decrease substantially and result in acute myocardial ischemia.⁷ In infants with unrecognized ALCAPA, PDA ligation has been associated with sudden myocardial ischemia, ventricular fibrillation and failure, and cardiac arrest.^{8,9}

To our knowledge, the beneficial physiologic effect of a PDA on myocardial perfusion in the presence of ARCAPA has not been described. This case—perhaps

in the youngest patient reported to date—highlights the importance of accurately delineating the coronary arteries in Turner syndrome, and it emphasizes the potential for myocardial dysfunction caused by decreased PA pressure associated with closing a PDA.

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