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Case Report

Diagnosis and follow-up of neonatal tetralogy of Fallot and hemitruncus with discontinuous pulmonary arteries noninvasively using awake ultra low-dose computed tomographic angiography

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ABSTRACT

We present a case of an ex-30 week premature male infant diagnosed postnatally with Tetralogy of Fallot, hemitruncus, and discontinuous pulmonary arteries (PAs) at 6 days of life. The patient was diagnosed by echocardiography, and the diagnosis was confirmed on subsequent dual-energy chest CT angiogram. In our patient, the left PA arose directly from the aorta, whereas the right PA originated normally from the right ventricular outflow tract. At 9 days of life, he underwent successful surgical palliation with placement of a modified Blalock–Taussig (aortopulmonary) shunt from the base of the left subclavian artery to the anomalously connected left PA along with anastomosis together of the right and left branch PAs to establish continuity with the main PA. Such cases have been described and are rare. The specific aim of this case report is to illustrate the added benefit of dual-energy electrocardiographically-triggered computed tomographic angiography (CTA) along with standard echocardiography. In addition, high quality images useful in preoperative planning were obtained noninvasively using an ultra low radiation dose without the need for sedation. The information obtained proved essential for confirmation of the diagnosis, preoperative planning, and post-surgical monitoring of branch PA development.

Introduction

Hemitruncus (HT) refers to a form of congenital heart disease in which a PA branch (usually the right) arises directly from the aorta, whereas discontinuous PAs describes a condition where one or both PAs originate from the ductus arteriosus or a major aortopulmonary collateral artery. Both are uncommon with only a fraction of a percent of HT cases being

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associated with TOF [1]. This patient was considered to have discontinuous pulmonary arteries (PAs) since the left PA arose from the ascending aorta at the base of the innominate artery instead of the normal arrangement where both PAs arise from the main PA. While similar cases have been described on a few occasions in the literature, to our knowledge the use of awake ultra low radiation dose chest computed tomographic angiography (CTA) to confirm the cardiac diagnosis and perform postsurgical follow-up in this lesion has not been reported.

Case report

A child born by urgent Caesarian section at 30 weeks gestation secondary to maternal preeclampsia was admitted to the Neonatal Intensive Care Unit. The patient had been in the newborn nursery when, at 6 days of life, repeated cyanotic episodes prompted further evaluation with 2D/Doppler echocardiogram (echo). The respiratory rate was 59 breaths per minute with oxygen saturation by pulse oximetry in the low 90s. The hemoglobin and hematocrit were within normal limits for age (15.7 and 47.1, respectively). Physical examination revealed a mildly cyanotic neonate in no acute distress with no dysmorphic features. A 2 out of 6 continuous cardiac murmur was auscultated throughout the precordium and the infant demonstrated normal peripheral perfusion. A chest X-ray indicated a mildly enlarged "boot-shaped" cardiomediastinal silhouette and the 12-lead electrocardiogram (ECG) showed right ventricular hypertrophy with prolonged QT interval.

After the diagnosis of Tetralogy of Fallot (TOF), HT, and discontinuous branch PAs was established by echo, a chest CTA with contrast was obtained to further delineate branch PA origin and size. The CTA also helped confirm that the right PA arose normally from the main PA and the left PA arose anomalously from the ascending aorta near the base of the innominate artery. There was no evidence of native right or left branch PA stenosis. The arch was right-sided.

At 9 days of life, the patient underwent surgical placement of a 3-mm modified Blalock–Taussig (BT) aortopulmonary shunt from the proximal left subclavian artery to the anomalously connected left PA along with anastomosis of the right and left branch PAs together to establish continuity with the native right ventricular outflow tract (Fig. 1). Postsurgically, the patient was admitted to the Cardiovascular Intensive Care Unit and maintained briefly on an intravenous milrinone infusion and then placed on daily aspirin therapy when able to tolerate enteral feedings.

The branch PAs were subsequently surveyed for growth and stenosis with echo. Follow-up echo done 2 weeks after surgery suggested that the right PA was fed by antegrade flow from the main PA and the left PA was supplied by flow from the left BT shunt with limited main PA contribution, raising the suspicion of the recurrence of branch PA discontinuity. Image quality was not sufficient to prove this given several technical factors. Thus, follow-up chest CTA was obtained which revealed a patent BT shunt to the left PA connecting to the left subclavian artery but with occlusion of the surgically created PA anastomosis. There was also mild hypoplasia of both branch PAs. At that time, the PA anastomosis was not repaired as the child had an acceptable source of pulmonary blood flow and would need full repair of tetralogy of Fallot in the relatively near future so a more definitive source of pulmonary flow could be established.

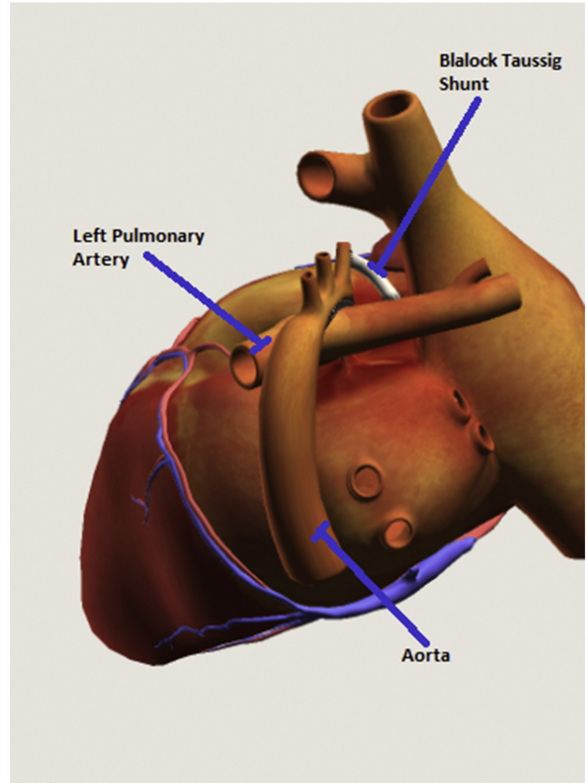

Fig. 1 – Blalock–Taussig shunt mirroring the repair completed in the case. Although this figure depicts a right-sided aortopulmonary shunt, our patient underwent the same procedure with connection to the left pulmonary artery instead [2].

Meanwhile, the patient remained clinically stable with oxygen saturation in room air by pulse oximetry in the upper 80s to low 90s, which is considered adequate for his cardiac physiology. He was gradually advanced to full oral alimentation and discharged home in good condition.

Discussion

HT is most often encountered as an isolated cardiovascular finding, first described by Fraentzel in 1868. Building on this work, Kutsche and Van Mierop [3] published the largest case series and review of this diagnosis in 1988 encompassing 108 cases of which 89 involved anomalous right PA origin from the aorta, whereas in 19 it was the left PA which arose directly from the aorta. Discontinuous PAs can be a characteristic of HT. Specifically, HT refers to a PA branch that arises from the aorta and discontinuous PAs describe a case in which one or both PAs originate from the ductus arteriosus or a major aortopulmonary collateral artery [4]. Embryologically, an anomalous origin of the left PA is most likely from persistence of the left fifth branchial arch and hypoplasia of the left sixth

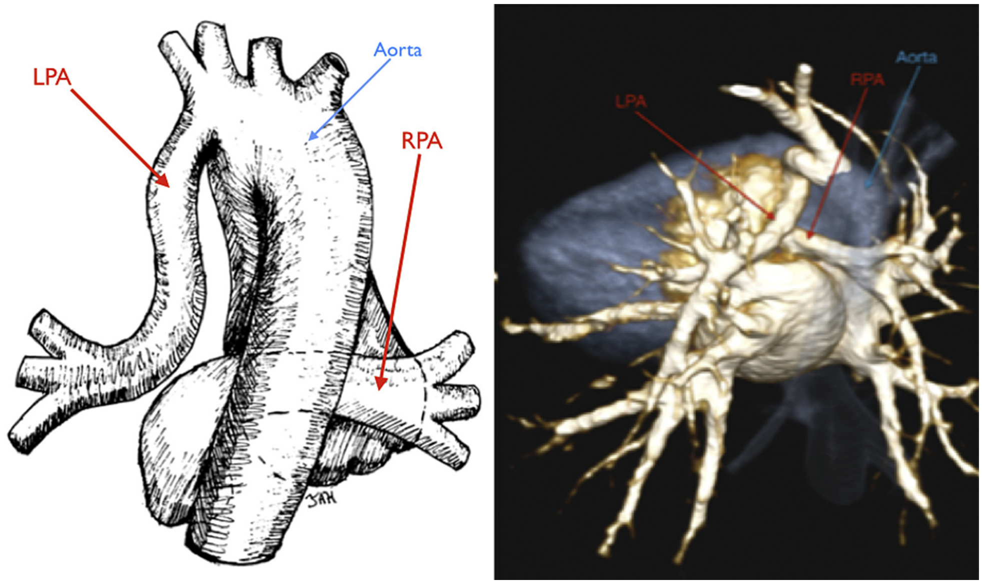

Fig. 2 – Preoperative anatomy by computed tomographic angiography and hand-drawn anatomic counterpart. LPA = left pulmonary artery; RPA = right pulmonary artery. (Original drawing by Dr. Jamil A. Aboulhosn, used with permission.)

branchial arch [1]. Conversely, abnormal origin of the right PA is more a problem with migration of the branchial arches, suggesting a delay in migration of the right sixth arch [4].

This condition necessitates surgical correction as differential main PA flow to one PA and relatively high pressure aortic pulsatile flow to the other branch PA has been associated with pulmonary vascular congestion and arteriopathy as well as ventilation perfusion imbalances. In cases that are not promptly corrected with surgical intervention, there is a less than 30% survival rate [4]. In addition, once the branch PAs have been anastomosed together, a full TOF repair with ventricular septal defect closure and augmentation of the right ventricular outflow tract and pulmonary annulus with a transannular patch would have to be undertaken to complete the repair of this cyanotic defect.

Although there are many cases of solitary HT described, to our knowledge there have only been 17 rare cases of HT with TOF described to date. This has led to calculation of an association between the 2 conditions in approximately 0.4% of TOF cases. Right HT is generally more common while left HT is more often discovered in association with TOF and right aortic arch, as is the case with our patient. Other common

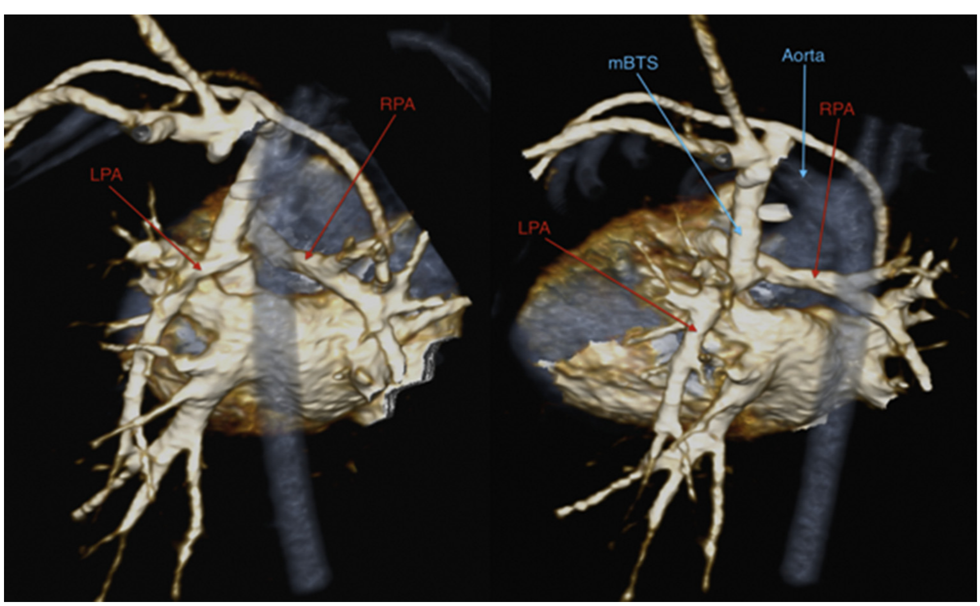

Fig. 3 – Postoperative anatomy by computed tomographic angiography. mBTS = modified Blalock–Taussig shunt; LPA = left pulmonary artery; RPA = right pulmonary artery.

associations with HT include interrupted aortic arch type A, coarctation of the aorta, and aortopulmonary window [1].

We present a case of TOF in a premature male neonate in combination with left HT and discontinuous PAs. To our knowledge, there have not been any cases of this constellation of defects reported in which advanced cardiovascular imaging has been an important component of preoperative planning and short-term follow-up [4].

Our patient underwent surgery to unite the 2 discontinuous branch PAs by reimplantation of the left PA into the main PA with placement of a 3-mm BT shunt from the left subclavian artery to the left PA. In addition, extensive patch reconstruction of the branch PAs was performed during the surgery to establish continuity between the branches and the BT shunt. Shunt placement was required to establish adequate pulmonary blood flow due to the fact that the native pulmonary valve and artery were hypoplastic. Most important, the BT shunt is expected to better control blood flow to the left lung and decrease the risk of pulmonary arteriopathy.

Postsurgical follow-up CTA imaging revealed a patent BT shunt to the left PA from the aorta but with occlusion of the surgically created PA anastomosis. Subsequently, the patient underwent an extensive patch augmentation of the branch PAs and placement of a larger left-sided BT shunt. Then, at 12 months of age, he underwent successful complete TOF repair with ventricular septal defect closure, BT shunt takedown, and repeat patch augmentation of the branch PAs resulting in a normal circulatory pattern.

In this patient, both chest CTAs obtained were performed in less than 1 second using a dual-source CT scanner and an ECG-triggered (not gated) mode with the patient awake and spontaneously breathing. Dual-source CT technology combines 2 sources of radiation simultaneously, which allows for rapid data acquisition. Besides a marked increase in temporal resolution with decrease in motion artifacts, this scan protocol allows for ultra low-dose radiation exposure without significantly compromising image quality or diagnostic efficacy (Figs. 2 and 3). The effective radiation dose for each of the scans was estimated at 0.22 mSy using a standardized conversion coefficient for term neonates and multiplying by a factor of 2 [5,6]. By comparison, the effective radiation dose from a typical neonatal chest CTA is in the range from 1.2 to 4.2 mSy and that of a diagnostic catheterization is 0.6-23.2 mSy [6,7]. This dose is equivalent to approximately 10 neonatal chest X-rays. Further, CTA and yields a three dimensional data set with good vascular and tissue definition as compared to the two dimension images acquired during cardiac catheterization, leading to more comprehensive preoperative planning with a significant reduction in radiation dose without compromising image quality. Historically, presurgical confirmation of anatomy and post-surgical follow up of branch PAs in such a patient has required invasive cardiac catheterization.

This case highlights the utility of noninvasive advanced cardiovascular imaging such as dual-source ECG-triggered chest CTA as a safe and important adjunct to standard 2D/Doppler echocardiography and an alternative to invasive cardiac catheterization in the preoperative planning and postoperative follow-up of patients with complex cyanotic congenital heart disease.

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