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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 preecclampsia 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.

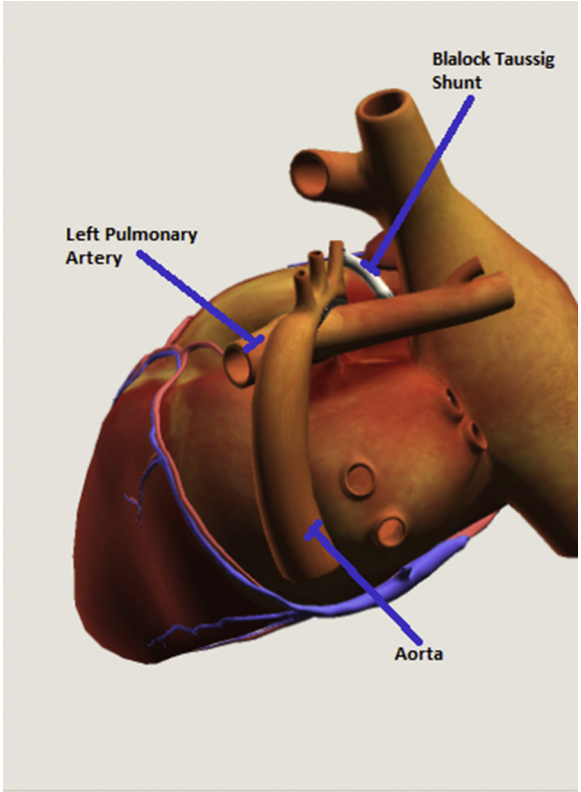

Fig. 1 – Blalock–Taussig shunt mirroring the repair completed in the case. Although this figure depicts a right-sided aortopulmonary shunt, our patient underwent the same procedure with connection to the left pulmonary artery instead [2].

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