Type B Interrupted Right Aortic Arch: Diagnostic and Surgical Approaches

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Interrupted right aortic arch is a rare congenital cardiovascular anomaly typically associated with other forms of congenital heart disease. We report two cases of interrupted right aortic arch associated with isolated left pulmonary artery and bilateral ductus arteriosus in the first case and with truncus arteriosus in the second case. Computed tomography was complementary to echocardiography in delineating the anatomic details in both cases, informing surgical repair with reconstruction of a left aortic arch in the first case and right arch in the second case.

(Ann Thorac Surg 2019;107:e41–3)

Interrupted right aortic arch (IRAA) is a rare congenital cardiovascular anomaly with only 30 reported cases [1, 2]. Surgical treatment is challenging because the right bronchus is higher than the left bronchus and is prone to compression after repair [1]. We report the diagnostic features and surgical approaches in two cases of IRAA associated with major cardiac anomalies.

Accepted for publication May 31, 2018.

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Case Reports

Patient 1

A 6-day old female newborn presented with cyanosis and tachypnea to an outside hospital and was later transferred to our institution. An echocardiogram and computed tomographic scan showed type B IRAA with mirror image branching. The aortic arch was interrupted between the right common carotid and right subclavian arteries. A large right-sided patent ductus arteriosus (PDA) supplied the descending aorta, whereas a restrictive left-sided PDA, which originated from the base of the left innominate artery, supplied a discontinuous left pulmonary artery (LPA) (Fig 1). Doppler interrogation of the left-sided PDA showed continuous flow with a maximal gradient of 75 mm Hg. A large conoventricular septal defect (VSD) with posterior malalignment of the conal septum was noted (Fig 2).

Surgical repair included aortic arch reconstruction with uncrossing to the left of the trachea and esophagus, pericardial patch closure of the VSD, bilateral PDA ligation, and LPA anastomosis to the main PA. Because of the proximal LPA stenosis, a stent was placed intraoperatively. The decision to reconstruct the aortic arch to the left of the trachea and esophagus was made after bronchoscopy before cardiopulmonary bypass showed complete obstruction of the right main bronchus where the ductal arch crossed over it. The uncrossing included extensive mobilization of the right ductal arch under and around the esophagus, bringing it to the left side of the

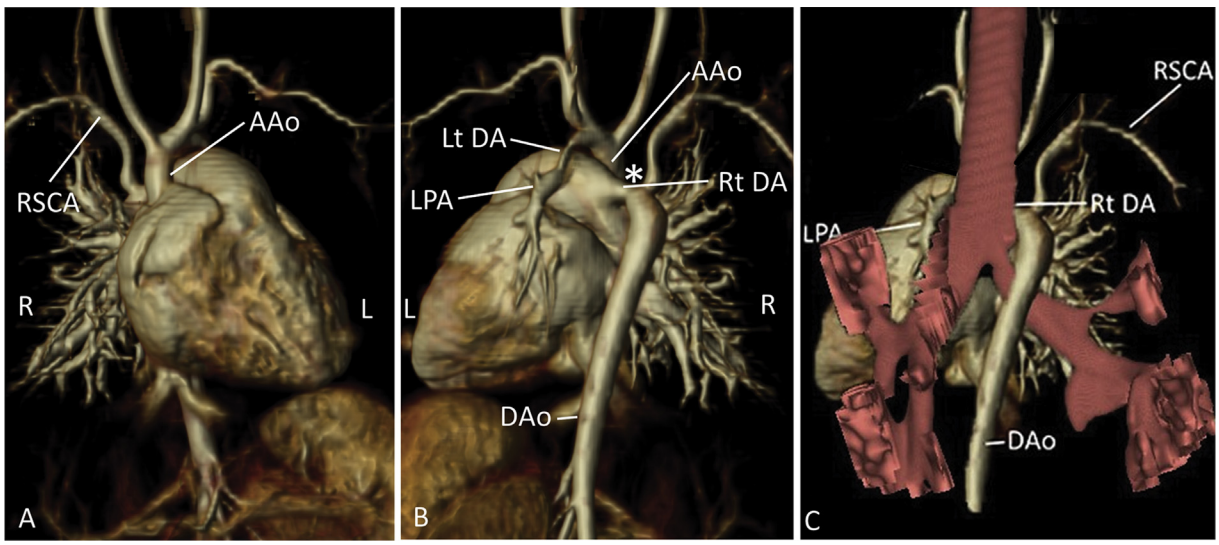

Fig 1. Computed tomographic angiogram image of the right-sided interrupted aortic arch in case 1. (A) Anteroposterior and (B) posteroanterior views with the site of interruption (*). (C) Posteroanterior view showing right bronchial compression before repair. (AAo = ascending aorta; DAo = descending aorta; L = left; LPA = left pulmonary artery; Lt DA = left ductus arteriosus; R = right; RSCA = right subclavian artery; Rt DA = right ductus arteriosus.)

0003-4975/$36.00
https://doi.org/10.1016/j.athoracsur.2018.05.098

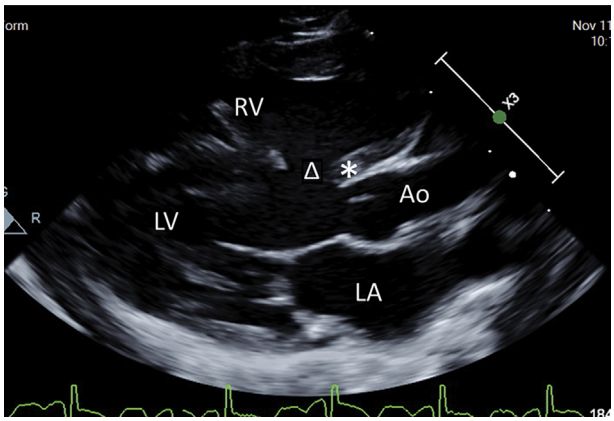

Fig 2. Echocardiographic image of the large conoventricular ventricular septal defect (arrowhead) with posterior malalignment of the conal septum (*). (Ao = aorta; LA = left atrium; LV = left ventricle; RV = right ventricle.)

trachea, and augmenting the arch with the use of a pulmonary homograft. Bronchospcopy after cardiopulmonary bypass showed remarkable improvement of the right bronchus compression. Postoperative imaging showed unobstructed left arch and mild LPA stenosis with 32% flow to the left lung by a lung perfusion scan. Patient was discharged home on postoperative day 15 and is clinically well. A follow-up lung perfusion scan at age 4 months showed 39% flow to the left lung. At latest follow-up at age 7 months the patient had no symptoms.

Patient 2

A male newborn had prenatal diagnosis of truncus arteriosus (TA). An echocardiogram showed type 4 TA with good-sized PAs arising separately from the posterior aspect of the trunk and likely type B IRAA. A computed tomographic scan showed IRAA with the left subclavian artery originating immediately above the takeoff of the branch PAs, followed by both carotids originating together through a common trunk. The aortic arch was interrupted between the carotid trunk and the right subclavian artery, which arose from the descending aorta. A large right-sided ductus arteriosus supplied the descending aorta (Fig 3).

At day of life 7 the patient underwent surgical repair with patch closure of the VSD, IRAA repair, placement of an 11-mm pulmonary homograft from the right ventricle to the PAs, and LeCompte maneuver (to facilitate the approximation between the ascending and descending aorta without compressing the right main bronchus).

Postoperative imaging showed an unobstructed right aortic arch and unobstructed branch PAs. Patient had poor oral feeding postoperatively and was discharged on postoperative day 60 with nasogastric tube feeding.

Comment

The reported two cases highlight the diagnostic challenges and the surgical approach to type B IRAA. To our knowledge, the combination of IRAA, bilateral ductus, and isolated LPA as described in the first case and the uncrossing of the aortic arch in these settings were not reported before. The surgical approach aimed at reconstructing the aortic arch while avoiding compression of the right main bronchus.

Isolated branch PA is a consequence of involution of the proximal sixth pharyngeal arch with persistence of the ipsilateral distal sixth arch, resulting in a ductus arteriosus on the same side of the isolated branch PA [3].

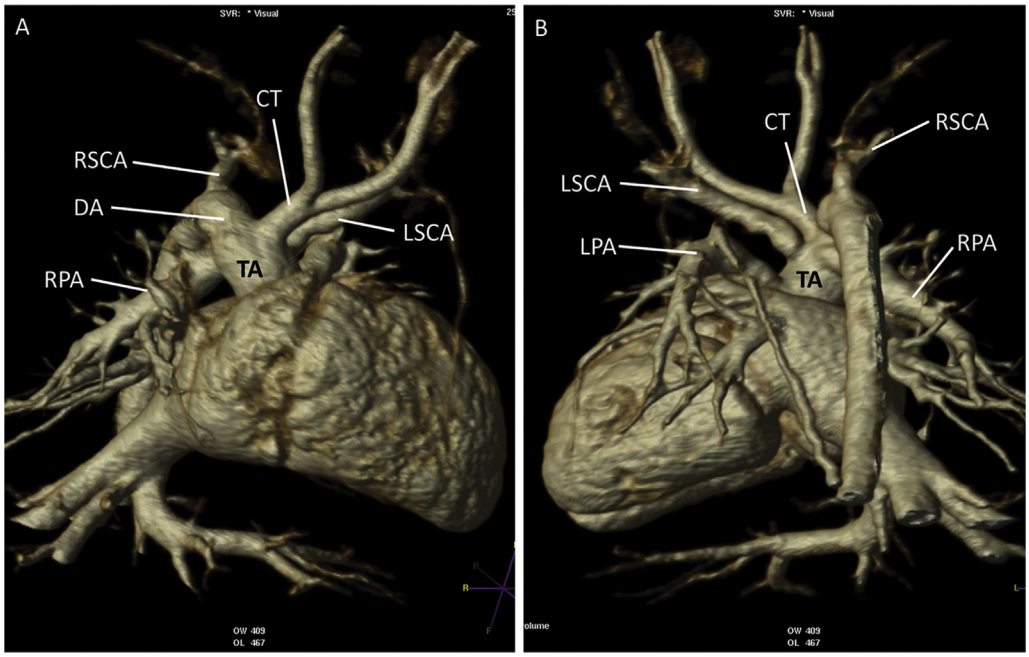

Fig 3. Computed tomographic angiogram image of the right-sided interrupted aortic arch in case 2. (A) Anteroposterior and (B) posteroanterior views. (CT = common carotid trunk; DA = ductus arteriosus; LPA = left pulmonary artery; LSCA = left subclavian artery; RPA = right pulmonary artery; RSCA = right subclavian artery; TA = truncus arteriosus.)

Our second case was associated with TA. Although interrupted aortic arch is seen in 15% of cases of type 4 TA, most exhibit interruption of a left aortic arch. In a current series, among 50 patients with TA and interrupted aortic arch only 3 had IRAA and 2 of these had died [4].

Surgical repair of IRAA is challenging because the right bronchus is higher than the left bronchus, and limited space is available for aortic arch reconstruction [1]. Direct anastomosis of a right aortic arch carries a risk of compression of the right bronchus and trachea in contrast to direct anastomosis of a left arch [1]. Therefore, uncrossing the arch to create a left arch was used in the first case, and LeCompte maneuver was performed in the second case to create more space and to prevent airway obstruction. The uncrossing procedure is reported in cases of circumflex aortic arch where there is a right aortic arch that passes over the right bronchus, and then takes a retroesophageal course, then becoming a left-sided descending aorta. The procedure involves recreating the transverse arch in front of the trachea [5]. In our first case we used that concept to mobilize the aorta and to create a left aortic arch, avoiding right bronchial compression.

The LeCompte maneuver was used in cases of TA and interrupted arch to avoid branch PA and left bronchus compression and was used in our case to avoid right bronchial compression [6].

With the use of echocardiography and computed tomography (CT) in concert proved helpful in our cases to provide comprehensive three-dimensional understanding of vascular and airway anatomy. CT is fast (approximately 200 milliseconds), can be performed without sedation, provides high spatial resolution (in-plane resolution 0.5 mm), and has modest radiation exposure (0.3 to 0.6 mSv). This makes CT an attractive method to evaluate complex aortic arch anatomy in the neonate [7].

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