*Case Reports*

# Importance of Absent Ductus Arteriosus

## in Tetralogy of Fallot with Absent Pulmonary Valve Syndrome

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*Tetralogy of Fallot without pulmonary valve syndrome is almost always associated with an absent ductus arteriosus. Patients with right aortic arch and retroesophageal left subclavian artery have a vascular ring if the left ductus arteriosus or its remnant and the Kommerell diverticulum are present. We report the cases of 2 infants in whom the role of an absent ductus arteriosus or its remnant is noteworthy. Both patients had a combination of tetralogy of Fallot with absent pulmonary valve syndrome and right aortic arch with retroesophageal left subclavian artery without a vascular ring. The absence of the ductus arteriosus has a role in the pathogenesis of tetralogy of Fallot with absent pulmonary valve syndrome. The absence of a ductus arteriosus in the right aortic arch with retroesophageal left subclavian artery precludes a vascular ring.* ***(Tex Heart Inst J 2014;41(6):664-7)***

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Tetralogy of Fallot (TOF) with absent pulmonary valve syndrome (APVS) usually causes substantial respiratory symptoms, due to bronchial narrowing of severely dilated pulmonary arteries (PAs). A vascular ring, if present, adds more problems. Tetralogy of Fallot with APVS is almost always associated with an absent ductus arteriosus (DA).[1,2] Right aortic arch with a retroesophageal left subclavian artery (LSA) typically constitutes a vascular ring when a left DA or its remnant is present along with the Kommerell diverticulum. In reporting the cases of 2 infants, we highlight the implications of the absence of a DA: the DA has a role in the pathogenesis of TOF with APVS, and the absence of a DA precludes a vascular ring in right aortic arch with retroesophageal LSA.

## Case Reports

*Patient 1.* The results of a fetal echocardiogram at 19 weeks of gestation led to a diagnosis of TOF with APVS. There was massive dilation of the PAs. The DA was never seen on any fetal echocardiogram. Delivery was by repeat cesarean section at 36 weeks of gestation. The female infant was intubated at birth. A transthoracic echocardiogram, obtained in the delivery room, confirmed the cardiac diagnosis. Massively aneurysmal PAs compressed the left atrium and compromised the cardiac output. The infant underwent surgical repair within 2 hours of delivery. The main PA was resected and replaced by a PA homograft. The dilated portions of the branch PAs were resected and reconstructed. The PA confluence was brought anterior to the ascending aorta by means of the LeCompte maneuver, to avoid airway compression from the PAs. No DA was identified intraoperatively by the surgeon. Several days later, because of persistent respiratory compromise, a computed tomographic evaluation of the chest was performed to clarify the vascular and airway anatomy. The ascending aorta arched over the right mainstem bronchus and was to the right of the trachea. The descending aorta was not retroesophageal. All 4 brachiocephalic arteries originated separately from the aortic arch in the following order from anterior to posterior: left common carotid artery, right common carotid artery, right subclavian artery, and LSA (Fig. 1). The LSA coursed from the right-sided descending aorta toward the left arm, crossing the midline posterior to the esophagus. The caliber of the proximal LSA was not substantially greater than that of its distal segment, and no Kommerell diverticulum was evident. Therefore, despite the right arch with retroesophageal LSA, a vascular ring was not present. The patient was later confirmed to have 22q11 deletion.

*Patient 2.* A 6-month-old girl presented with severe respiratory distress. She was known to have 22q11 deletion and TOF with APVS. As a neonate at another institution, she had undergone surgical closure of a ventricular septal defect, insertion of a pulmonary conduit, and plication of the PAs. She continued to have substantial respiratory problems, caused by persistent narrowing of the tracheobronchial tree. Computed tomograms of the chest clarified the vascular and airway anatomy. A right aortic arch crossed over the right mainstem bronchus, passing to the right of the trachea. The descending aorta

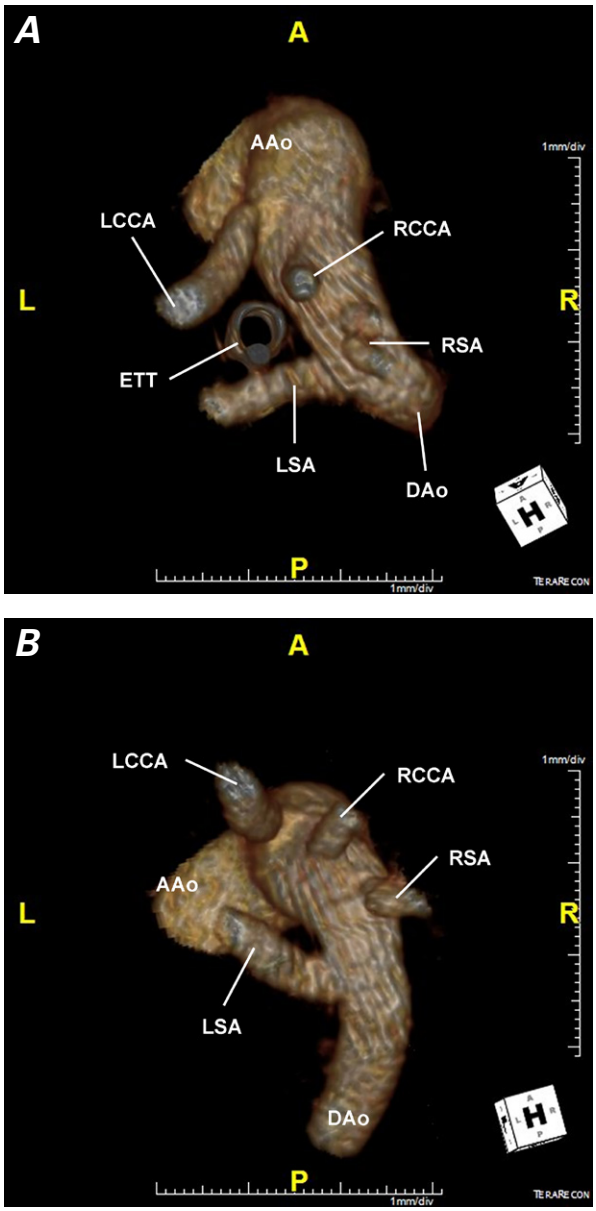


***Fig. 1*** *Patient 1. Computed tomogram (volume-rendered 3-dimensional reconstruction).* ***A****) View from the head shows the origin of the brachiocephalic vessels and an incomplete vascular ring.* ***B****) Posterolateral view of the origin of the left subclavian artery shows a nondilated proximal portion, excluding the Kommerell diverticulum. Absence of a left-sided ductus arteriosus precludes a vascular ring. Endotracheal tube (center) marks the tracheal position.*

*AAo = ascending aorta; DAo = descending aorta; ETT = endotracheal tube; LCCA = left common carotid artery; LSA = left subclavian artery; RCCA = right common carotid artery; RSA = right subclavian artery*

*Supplemental motion image is available for Figure 1.*

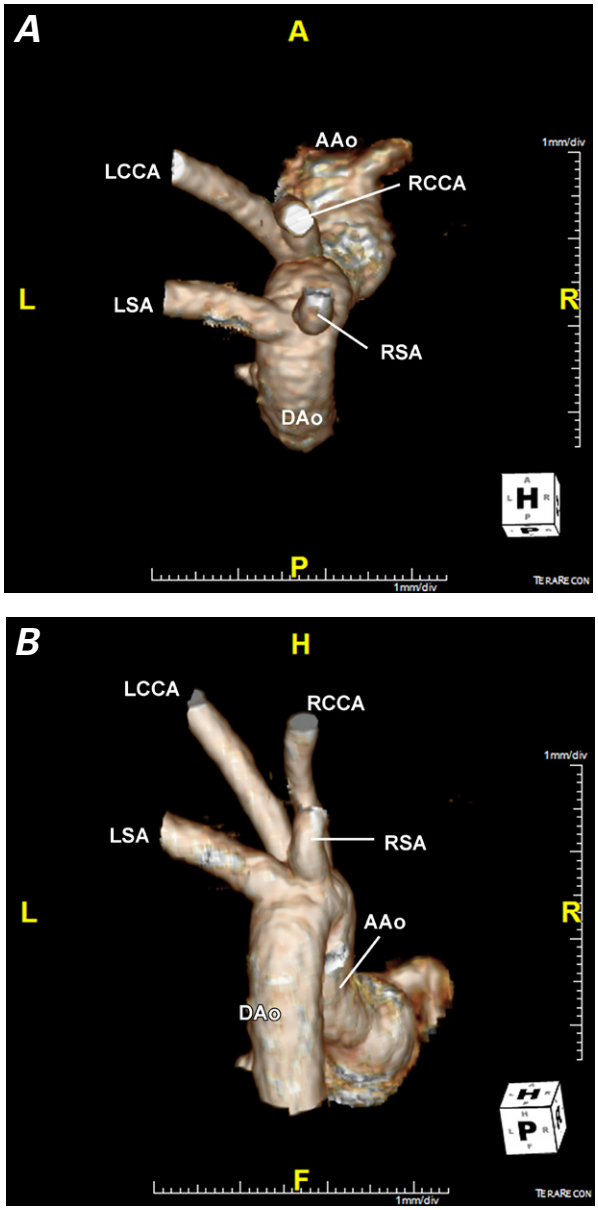


***Fig. 2*** *Patient 2. Computed tomogram (volume-rendered 3-dimensional reconstruction).* ***A****) View from the head shows the origin of the brachiocephalic vessels and an incomplete vascular ring.* ***B****) Posterior view of the origin of the left subclavian artery shows a nondilated proximal portion, excluding the Kommerell diverticulum. Absence of a left-sided ductus arteriosus precludes a vascular ring.*

*AAo = ascending aorta; DAo = descending aorta; LCCA = left common carotid artery; LSA = left subclavian artery; RCCA = right common carotid artery; RSA = right subclavian artery*

did not cross the midline and was not retroesophageal in position. Three brachiocephalic arteries arose from the aortic arch (Fig. 2): a common origin of the left and right common carotid arteries, a right subclavian artery, and a retroesophageal LSA. The LSA originated from the left anterior aspect of the upper descending aorta, crossed the midline behind the esophagus, and coursed toward the left arm. The proximal portion of the LSA was not excessively dilated and did not constitute a Kommerell diverticulum. There was no dimple or bump on the descending aorta that would mark the presence of retroesophageal ligamentum arteriosum. Because of persistent tracheobronchial obstruction from aneurysmal PAs, the patient underwent repeat median sternotomy. The LeCompte maneuver was performed, along with repeat plication of the PAs. During the operation, there was no evidence of a ligamentum arteriosum connecting to the descending aorta.

## Discussion

Absent pulmonary valve syndrome is a rare form of congenital heart disease. The absence of the pulmonary valve causes to-and-fro flow between the right ventricle and the PAs, which leads to severe dilation of the PAs. These aneurysmal vessels can compress the tracheobronchial tree, causing respiratory compromise. Absent pulmonary valve syndrome can occur with or without a ventricular septal defect. Absent pulmonary valve syndrome with ventricular septal defect (also known as TOF with absent pulmonary valve) is commonly associated with absence of the DA.[1,2] Conversely, when the ventricular septum is intact, the DA is usually present.[3-6] The absence of the DA has a role in the pathogenesis of APVS.[1,5,7] In utero, the absence of the DA prevents the exit of blood from the PA into the descending aorta. This causes increased PA pressure and consequent severe pulmonary regurgitation, which interferes with normal development of the pulmonary valve.[7] The hypoplastic or absent pulmonary valve along with severe pulmonary regurgitation causes the PAs to dilate. In contrast, pulmonary regurgitation acquired in postnatal life usually does not cause dilation of the PAs as is seen in APVS. This is because low PA resistance enables the volume of blood entering the main PA to flow easily through the pulmonary vascular bed. It is important to note that, if the DA is present in cases of APVS with ventricular septal defect, there are discontinuous PAs, and the ductus supplies only the left PA.[8-10]

Right aortic arch has been seen in 13% to 34% of cases of TOF.[11] Right aortic arch with retroesophageal LSA can create a vascular ring if the left DA is present.[12,13] To complete the ring, a left-sided DA or ligamentum must connect either to the Kommerell diverticulum at the base of the LSA, or directly to the descending aorta. The Kommerell diverticulum is a dilated proximal portion of the LSA, which is a remnant of the left dorsal aortic root.[14] It is dilated because it receives all cardiac output from the right ventricle during fetal life. If the ductus has closed, the presence of a ligamentum cannot be ascertained by means of echocardiographic evaluation. Computed tomograms and magnetic resonance images can help to identify the ligamentum. However, the Kommerell diverticulum can be identified via any imaging method as a dilated proximal portion of the LSA. This dilated portion can be equal to the descending aorta in caliber and tapers down to the subclavian artery at the attachment of the ligamentum. In the absence of a left DA, the proximal portion of the LSA is not dilated, similar to the findings in our 2 patients. In addition, neither computed tomography nor inspection yielded evidence of a left ligamentum arteriosum attached to the descending aorta. In such situations, in the absence of a left DA or its remnant, a vascular ring is precluded in the right aortic arch with retroesophageal LSA. This is true whether a right DA is present or there is no ductus at all. A right aortic arch is also more likely to have a retroesophageal LSA without a vascular ring in cases of 22q11 deletion, rather than a right aortic arch with a mirror-image branching pattern.[15] This was true in both our patients.

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