

# Asymptomatic Right Circumflex Aortic Arch Associated with Ventricular Septal Defect and Bicuspid Aortic Valve

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## ABSTRACT

**Introduction:** Anomalies of the aortic arch are frequent congenital malformations, which rarely form partial or complete vascular rings. A rare form of vascular ring is the encircling, or circumflex, aortic arch.

**Case Report:** A 19-month-old boy, with no respiratory symptoms, was referred for ventricular septal defect (VSD) repair. Cardiac magnetic resonance imaging and echocardiography confirmed the perimembranous VSD, a bicuspid aortic valve with normal function, and showed a right-sided ascending aorta, bifurcating to the left behind the esophagus and trachea above the tracheal bifurcation, with a left-sided descending aorta, a left ligamentum arteriosum and aberrant left subclavian artery, realizing a circumflex aortic arch. The child underwent successful VSD repair and ligamentum arteriosum division, with an uneventful postoperative course.

**Conclusions:** Previous reports have described the association of circumflex aortic arch with VSD, but there is no previous report of its association with VSD and bicuspid aortic valve. Patients are usually symptomatic either preoperatively, or after VSD repair. For this reason, division of the ligamentum arteriosum, to open the vascular ring and free the trachea and esophagus from compression, should be performed in patients undergoing cardiac surgery for associated malformations.

## INTRODUCTION

Anomalies of the aortic arch are frequent congenital malformations, which rarely form partial or complete vascular rings. A rare form of vascular ring is the encircling, or circumflex, aortic arch. We report this incidental finding and association with other congenital cardiac malformations in a young boy, as well as its surgical management.

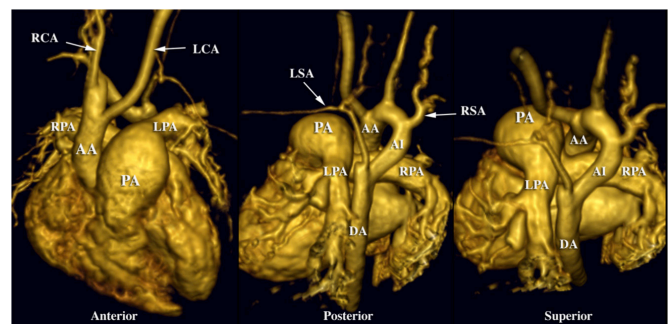
## CASE REPORT

A 19-month-old boy was referred for ventricular septal defect (VSD) repair. On physical exam, the child presented

mild tachypnea with no signs of upper airways obstruction. Echocardiography showed a large perimembranous VSD with left heart dilatation. The aortic valve was bicuspid with neither stenosis nor regurgitation. The aortic arch anatomy was difficult to delineate by echocardiography, but the aortic arch appeared right-sided with suspicion of a left aberrant subclavian artery. Cardiac magnetic resonance imaging (Figure) confirmed the right-sided ascending aorta, bifurcating to the left behind the esophagus and trachea above the tracheal bifurcation, with a left-sided descending aorta, a left ligamentum arteriosum and aberrant left subclavian artery, realizing a circumflex aortic arch [Knight 1974]. The child underwent successful VSD repair and ligamentum arteriosum ligation and division, with an uneventful postoperative course and no respiratory symptoms, allowing extubation on postoperative day one.

## DISCUSSION

Circumflex or encircling aortic arch is a rare form of vascular ring, usually associated with severe tracheobronchial compression [Bruniaux 1984; Minami 1986; Traxer 1998; Watanabe 1995], created by a ligamentum arteriosum or by the retroesophageal aorta itself. The treatment in symptomatic patients consists of surgical ligation and division of the



Volume rendering of 3D Gadolinium-enhanced magnetic resonance angiography of the aortic arch, showing a right-sided aortic arch bifurcating to the left behind the esophagus and trachea to form a left-sided descending aorta (encircling circumflex aortic arch) with a retroesophageal left subclavian artery. AA indicates ascending aorta; DA, descending aorta; AI, aortic isthmus; PA, pulmonary artery; RPA, right pulmonary artery; LPA, left pulmonary artery; LSA, left subclavian artery; LCA, left carotid artery; RCA, right carotid artery; RSA, right subclavian artery.

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ligamentum arteriosum, which usually allows the trachea and esophagus to be freed of compression by this vascular ring. If compression persists from the segment of aorta which courses around and posterior to the esophagus and trachea, some authors have proposed uncrossing the aorta [Bruniaux 1984], although this is an elaborate procedure requiring deep hypothermic circulatory arrest and is seldom necessary.

Previous reports have described the association of circumflex aortic arch with VSD [Bruniaux 1984; Watanabe 1995], but there is no previous report of its association with VSD and bicuspid aortic valve. Patients are usually symptomatic either preoperatively, or after VSD repair. For this reason and despite the absence of respiratory symptoms, surgical ligation and division of the ligamentum arteriosum, to open the vascular ring and free the trachea and esophagus from compression, should be performed in patients undergoing cardiac surgery for associated malformations.

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