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Images in Congenital Cardiac Disease

Right aortic arch with retroesophageal brachiocephalic artery

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Keywords: Multidetector computed tomography; vascular ring; congenital heart disease

A 3-MONTH OLD MALE INFANT WITH DYSPHAGIA and respiratory difficulty was referred for investigation of a possible vascular ring. Transthoracic echocardiography revealed a persistently patent arterial duct without intracardiac anomalies. He had no finding to suggest 22q11 deletion. Volume-rendered images from multidetector computed tomography (Figs 1 and 2; AA = ascending aorta; DA = descending aorta; MPA = pulmonary trunk) showed a right aortic arch with aberrant, retroesophageal brachiocephalic artery. The connection between the pulmonary trunk and the left-sided brachiocephalic artery was interpreted as an abnormally positioned arterial duct, which more usually would connect to the proximal part of the left pulmonary artery. Surgical observations confirmed these findings, although there was additionally a fibrous ligament connecting the base of the left subclavian artery to the left pulmonary artery. Through a left thoracotomy, the patent arterial duct and fibrous ligament were divided, and the child remains well one year after operation. Figure 3 shows a postoperative image.

While difficult to explain strictly according to the hypothetical model of the double aortic arch

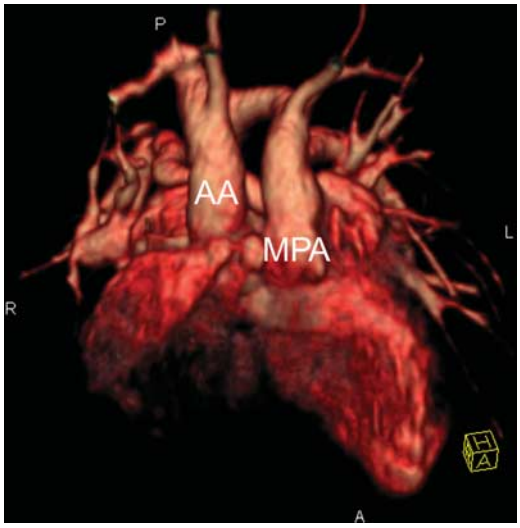


Figure 1.

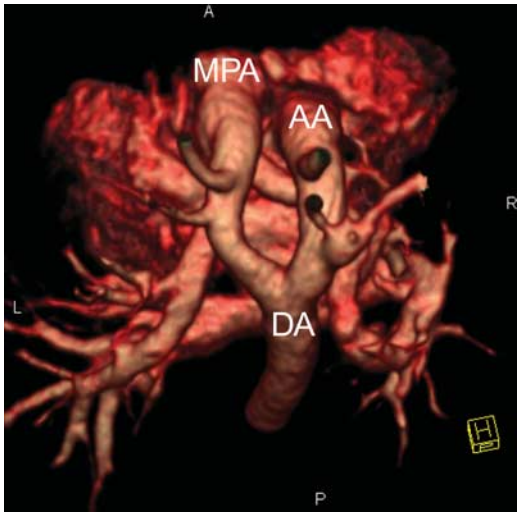


Figure 2.

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Figure 3.

propounded by Edwards,[1] this rare type of right aortic arch could result from regression of the right distal sixth primitive pharyngeal arch, the arterial duct, and the proximal part of the left arch between the origin of the left common carotid artery and the right arch. Unlike right aortic arch with aberrant left subclavian artery, Kommerell's diverticulum is not associated with this anomaly.

Reference

1. Edwards JE. Anomalies of the aortic arch system. Birth Defects 1977; 13: 47–63.

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