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An 18-year-old woman with known DiGeorge syndrome presented with dyspnoea; the chest radiograph suggested a right-sided arch (panel A). 64 Detector row ECG-gated thoracic aortic computed tomography (General Electric Healthcare Technologies, Waukesha, USA) was performed to exclude a vascular ring. A 1.25 mm collimation was used, with 100 ml intravenous iodinated contrast and 50 ml saline bolus chaser. Images were reconstructed on a GE workstation.

This demonstrated a widely patent double aortic arch (DAA), mild hypoplasia of the left arch, with left and right subclavian and common carotid arteries arising from their respective arches panel B and a small ductus diverticulum/ligamentum arteriosus (starred).

DiGeorge syndrome, also known as 22q11 deletion syndrome or CATCH 22 (conotruncal cardiac defects, abnormal facies, thymic hypoplasia, cleft palate and hypocalcaemia) has a prevalence of 1:4000. Congenital cardiac defects occur in 75–80% of patients, most commonly tetralogy of Fallot, interrupted aortic arch, truncus arteriosus, ventricular septal defect and aortic arch anomalies.

Twenty-four per cent of patients with an isolated aortic arch abnormality are found to have 22q11 deletion. DAA is rarely associated with congenital heart disease, but when present tetralogy of Fallot is the most common. The two separate arches arise from a single ascending aorta, and form a single descending aorta. Typically the right-sided arch is more superiorly located—usually the left arch is either hypoplastic or atretic; the
smaller arch is anterior in 80% of cases. DAA is associated with a right-sided descending aorta in 25% of cases.

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