IMAGES in PAEDIATRIC CARDIOLOGY

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Abstract
We present a rare case of isolated right subclavian artery arising from a right-sided patent arterial duct in a patient with DiGeorge syndrome, diagnosed on cardiac CT, along with potential complications and management approaches.

Introduction
An aberrant right subclavian artery occurs in approximately 0.5–1.0% of the population.[1] This is the most common aortic arch anomaly that originates from a left sided aortic arch.[2] This anomaly results from interruption of the dorsal segment of the right arch between the right carotid artery and right subclavian artery, with regression of the right ductus arteriosus in the developing double aortic arch.[3]

Isolation of the subclavian artery, on the other hand, is the rarest arch anomaly.[4] In this situation, the subclavian artery is completely separate from the aortic arch and arises from a ductus or ligamentum arteriosus connected to the same sided pulmonary artery. This anomaly can cause pulmonary or subclavian steal with perfusion of the arm or lung via the vertebrobasilar system.[4]

This abnormality is seen most commonly in association with 22q11 deletion.[4] We report a case of a boy diagnosed with DiGeorge syndrome with 22q11 deletion who has an aberrant right subclavian artery arising from a right patent ductus arteriosus.

Case Report
Our patient was born at 40+1 gestation via normal delivery to a 45 year old healthy mother. APGAR scores were 9, 9 at 1 and 5 minutes respectively. Birth weight was 3.31kg.

At 10 hours of age he was transferred to Neonatal Intensive Care (NICU) in view of feeding difficulty due to nasal congestion with cyanotic spells. On examination he was found to have a possible right choanal atresia, left sided structural talipes, high arched palate and bilateral single palmar creases. Occasional oxygen desaturations were noted and CT brain, EEG and echo were normal (except for a left-sided patent ductus arteriosus and patent foramen ovale – physiological for age).

Following discharge from NICU, self-resolving cyanotic episodes recurred. Bronchoscopy excluded compression of the trachea or of proximal main bronchi. Genetics studies confirmed 22Q11.2 microdeletion. No hypocalcaemia was detected.

During his stay it was noted that the right brachial pulse was not palpable whilst normal on the left side. A non-ECG gated, contrast-enhanced CT was performed which demonstrated an aberrant vessel coursing anterior to the trachea interpreted as arising from the region of the right pulmonary artery and coursing towards the expected location of the right subclavian artery. A separate right

subclavian artery was not identified at the expected origin from the brachiocephalic artery. The aortic arch is left sided with the usual branch pattern and architecture, with the exception that the first branch is a right common carotid artery rather than an innominate artery.

The patient is currently 3 years old and is well, with repeated echocardiograms showing the aberrant vessel causing a small left to right shunt at great artery level with no signs of ventricular dysfunction. An ECG-gated contrast enhanced CT was performed and again showed isolation of right subclavian artery, which arises via a right-sided PDA from the right pulmonary artery. Collaterals from right intercostal arteries provide arterial blood to the right vertebral artery which, in turn, is connected to the aberrant right subclavian artery. Arterial phase contrast-enhanced blood appears to flow from the aberrant subclavian artery to the low-pressure pulmonary artery, analogous to the situation with a left-sided PDA. The pulmonary arteries were seen to be dilated and mildly ectatic. Also noted was a probably absent inferior vena cava. No other cardiovascular anomalies were demonstrated (figures 1 and 2).

Figure 1: Volume rendered reconstruction of contrast-enhanced 3D angiogram of thoracic aorta obtained by ECG-gated computed tomography, viewed from posterior aspect. A large right-sided patent ductus arteriosus is seen connecting the isolated right subclavian artery to the right pulmonary artery. In addition, there is a plexus of collaterals in the neck supplying arterial blood to the right subclavian and right vertebral arteries, arising from right intercostal arteries coursing upwards towards the neck (full course not shown).

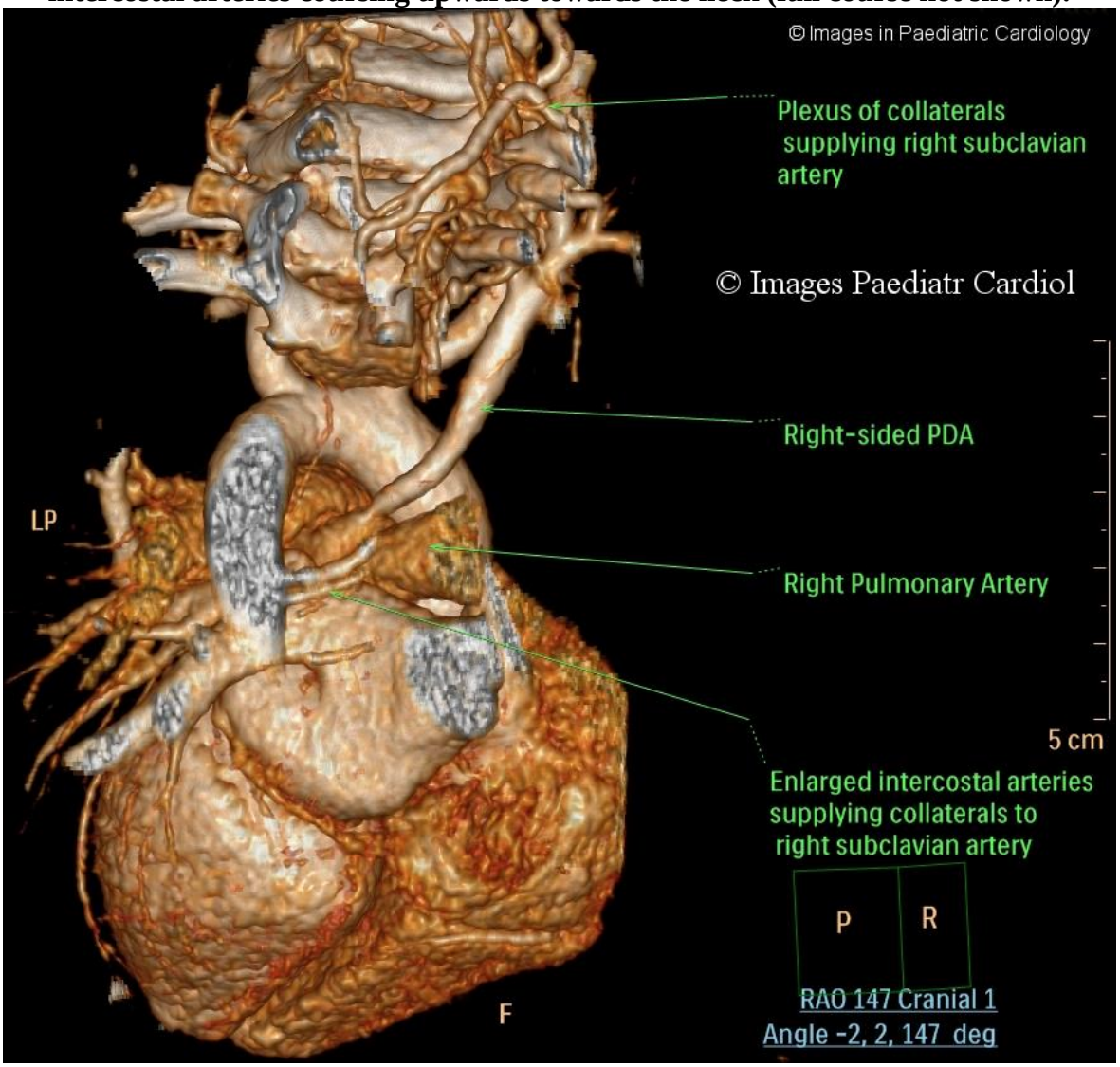
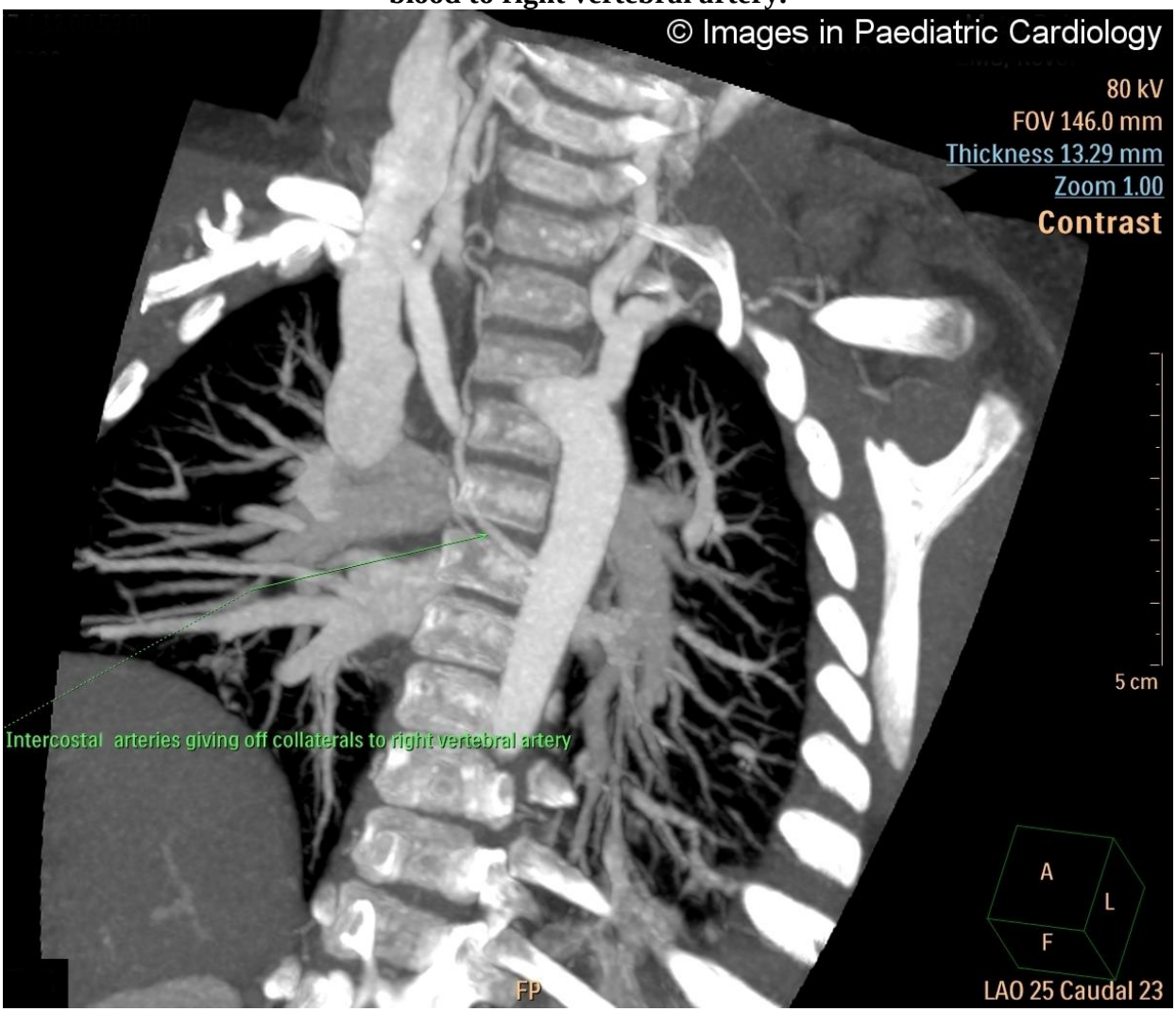


Figure 2: Same acquisition as in Figure 1, reconstructed as a thick 2D slab viewed from anterior aspect, showing enlarged right intercostal from descending aorta, supplying arterial blood to right vertebral artery.



Discussion

An aberrant right subclavian artery is generally asymptomatic and is diagnosed incidentally, but this anomaly may present with dysphagia in view of extrinsic compression of the esophagus. Rarely, with failure of regression of the right ductus, a loose vascular ring may be formed by the left aortic arch with an aberrant right subclavian artery, right pulmonary artery and right ductus arteriosus.[5]

Our patient has an isolated right subclavian artery which is the rarest described arch anomaly.[6,7] He had no vascular ring as the ductus is ipsilateral to the isolated subclavian artery and is not attached to the aorta. Pulmonary steal may occur from the circle of Willis leading to pulmonary hypertension.[4] Thus, the primary aim of any intervention is to limit pulmonary overcirculation from a left to right shunt, and this can be surgically.

Isolation of the subclavian artery is seen in association with DiGeorge syndrome and with conotruncal cardiac defects.[2–4] Therefore it is important to remember to screen for this deletion when a patient presents with an arch anomaly, as earlier other DiGeorge signs may be subtle and missed.[8] Our patient does not demonstrate any signs of pulmonary steal and is well in himself.

We thus present a rare case of isolated right subclavian artery arising from a right-sided PDA in a patient with DiGeorge syndrome, diagnosed on cardiac CT, along with potential complications and management approaches.

References

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