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IMAGING VIGNETTE

BEGINNER

CLINICAL VIGNETTE

Truncus Arteriosus and Absent Ascending Aorta With Unusual Head and Neck Vessel Origins

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ABSTRACT

We describe a neonate with a unique variant of truncus arteriosus with interrupted aortic arch, an absent ascending aorta, persistent right dorsal aorta, and an unusual brachiocephalic artery pattern in which all head and neck vessels were supplied from the ductal arch-descending aorta continuum. (**Level of Difficulty: Beginner.**) (J Am Coll Cardiol Case Rep 2023;14:101839)

We describe a unique variant of truncus arteriosus communis (TA) with interrupted aortic arch (IAA) and absent ascending aorta, and a unique, retrograde brachiocephalic artery supply. Three-dimensional anatomy was elucidated by using computed tomography (CT) imaging.

Complex congenital heart disease was identified prenatally. Technically limited fetal echocardiograms suggested a diagnosis of aortic atresia with extreme ascending aortic hypoplasia vs TA with IAA type B and aberrant subclavian artery. The postnatal echocardiogram of the genetically normal, term, male infant displayed a trileaflet, normally functioning truncal valve entirely committed to the right ventricle, a large malalignment ventricular septal defect, and a single, anteriorly arising coronary artery. Both pulmonary artery branches arose separately from the posterior aspect of the truncus. The ascending aorta was absent. The arterial trunk above the pulmonary artery ostia continued into a mildly restrictive, left-sided, ductal arch that enlarged after prostaglandin infusion. A left subclavian artery and a tortuous, right dorsal aorta arose from the proximal descending aorta; other details of retrograde brachiocephalic artery supply were unclear, however.

A gated CT scan with contrast confirmed TA-IAA with absent ascending aorta and separate branch pulmonary artery ostia from the posterior truncus with a more superior left pulmonary artery takeoff. Above this, the truncus continued as a left ductal arch to the left descending aorta. All head and neck vessels were supplied postductally, with the first branch being the left subclavian artery, immediately followed by a larger posterior and rightward vessel that continued superior-anteriorly to give off the right subclavian artery. This vessel became tortuous, wrapping anteriorly around the right superior vena cava, forming a “U”-shaped termination parallel to and behind the left innominate vein that supplied the right and left common carotid arteries (**Figure 1**).

The neonate underwent surgical palliation with extensive reconstruction of the left arch and placement of a right ventricular-pulmonary artery shunt. Intraoperative inspection confirmed the detailed CT scan findings.

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ABBREVIATIONS AND ACRONYMS

CT = computed tomography

IAA = interrupted aortic arch

LSA = left subclavian artery

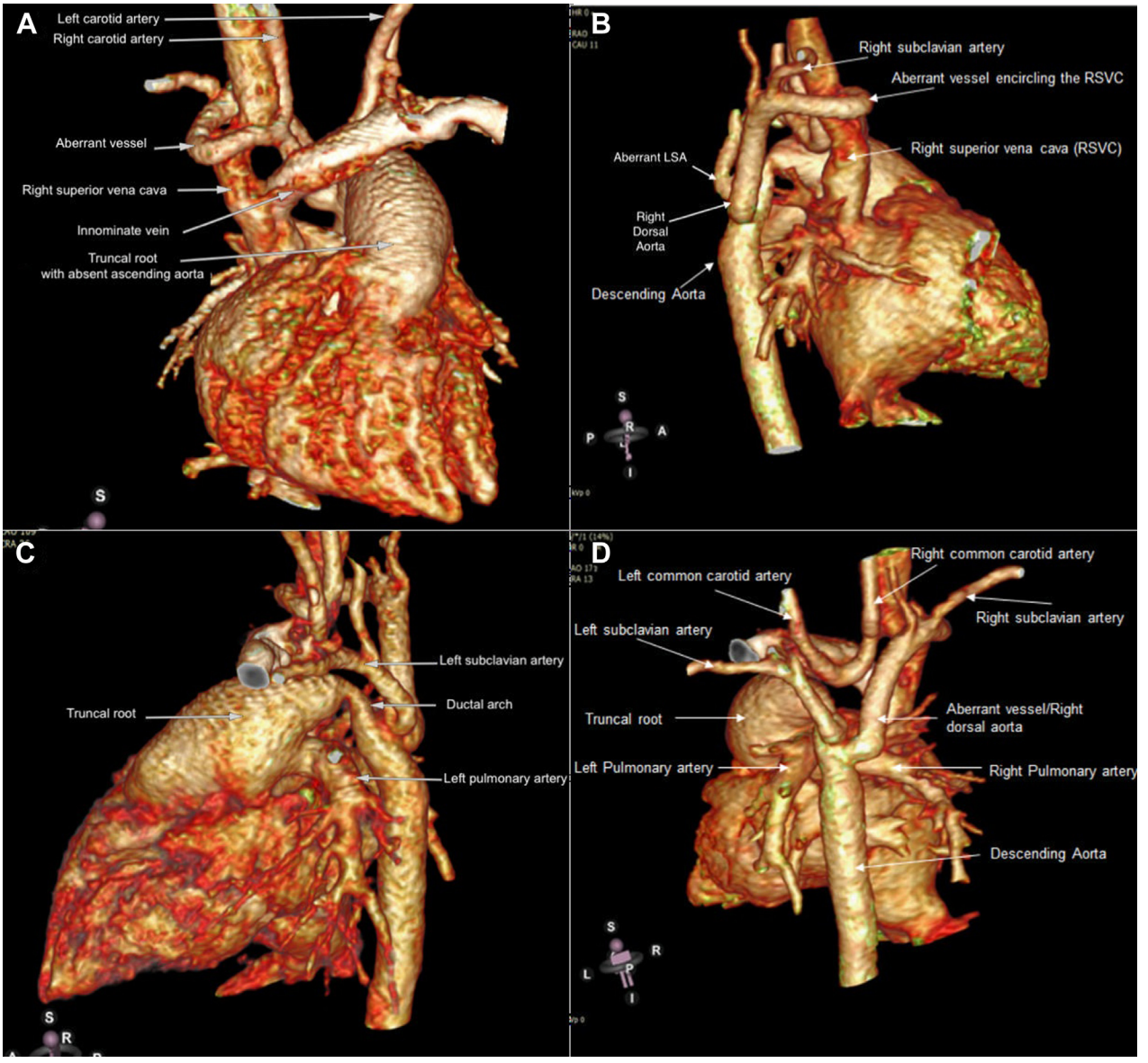
RSVC = right superior vena cava

TA = truncus arteriosus communis

The patient recovered without major complications and was discharged home 4 weeks later but succumbed to sudden death at 11 weeks of age.

TA is a rare congenital malformation and, when associated with IAA, has worse outcomes compared with patients with other types of IAA.[1] Previous reports of TA-IAA have documented IAA types A and B, aberrant subclavian artery origin, and variably hypoplastic ascending aorta and arch.[1] A rare case of retrograde supply of a hypoplastic ascending aorta and aortic arch due to preferential truncal valve streaming has been reported.[2] To our knowledge, there is only 1 other reported case of TA-IAA with absent ascending aorta.[3] Our TA-IAA case was unique in that the right subclavian and both carotid arteries were supplied by a persistent, tortuous (embryonic) right dorsal aorta that wrapped around

FIGURE 1 Three-Dimensional CT Imaging (after prostaglandin)



(A) Anterior view of the truncal root with absent ascending aorta and postductal, right-sided aberrant vessel encircling the right superior vena cava (RSVC) from behind, after supplying the right subclavian artery. The "U" shaped terminus supplying the right and left common carotid arteries is partly obscured by the overlying innominate vein. Right sagittal view **(B)** and left sagittal view **(C)** of the ductal arch from which the right dorsal aorta arises to supply all but the left subclavian artery (LSA). **(D)** Posterior view of the ductal arch, descending aorta and "U" shaped termination of the right dorsal aorta giving rise to both carotid arteries. CT = computed tomography.

the superior vena cava and innominate vein. Our experience reinforces the significant benefit of CT angiography in elucidating complex congenital extracardiac anatomy to aid in neonatal surgical planning.

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KEY WORDS computed tomography, congenital heart defect, persistent truncus arteriosus