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Right Aortic Arch with Isolated Left Subclavian Artery in Association with Double Outlet Right Ventricle and Cor Triatriatum Sinister: Hitherto Unreported Association

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Right-sided aortic arch with isolation of left subclavian artery (LSCA) is associated with a spectrum of congenital heart diseases, most commonly tetralogy of Fallot (TOF). However, to the best of our knowledge, its association with cor triatriatum has not been reported in literature so far. Moreover, present case highlights the rare concomitant presence of double outlet right ventricle (DORV) and cor triatriatum sinister with vertebral segmentation anomalies in association with isolation of LSCA.

An 11-month-old infant with cyanosis and echocardiographic findings of DORV, ventricular septal defect (VSD) without pulmonary stenosis underwent computed tomography (CT) angiography to assess the great vessels anatomy and status of branch pulmonary arteries. Additionally, echocardiography revealed dilated right-sided cardiac chambers with tricuspid annulus measuring 19mm and presence of mild tricuspid regurgitation. Left ventricle was smaller with mitral annulus measuring 8mm (Z score= −2.8). CT angiography confirmed the echocardiographic findings and additionally showed the presence of cor triatriatum sinister with thin hypodense membrane dividing the left atrium into proximal (receiving all pulmonary veins) and distal (containing left atrial appendage) chambers. Double superior vena cava (SVC) was seen with left SVC seen draining into the right atrium via coronary sinus (►**Fig. 1**). Interestingly, it showed presence of right sided aortic arch with no communication of LSCA from the aortic arch. However, LSCA was seen to arise from the pulmonary artery via patent ductus arteriosus (caliber 2.9 mm). Scoliotic curvature of spine with segmentation anomalies at multiple vertebral levels was also seen.

Right-sided aortic arch with isolated LSCA is an uncommonly reported arch anomaly where the LSCA arises from the pulmonary artery via the ductus arteriosus with no aortic origin, as seen in our case (►**Fig. 2**). It is seen often associated with a spectrum of congenital heart diseases (CHD), of which TOF is the most common.[1,2] Association with VSD, dextrotransposition of great arteries, interrupted aortic arch, and DORV has also been reported in literature.[1–4] Patients with associated CHD generally present early in the form of cyanosis as in case of TOF or failure to thrive secondary to heart failure as in case of large patent ductus. Isolated cases may present late in the adulthood with features of vertebrobasilar insufficiency secondary to the subclavian steal.

Nearly one third cases may have syndromic association, most commonly 22q11 mutation (DiGeorge syndrome), followed by trisomy 21.[1] Very rarely, VACTERL association has also been described in a single case report so far with primum atrial septal defect, common atrioventricular valve and indeterminate common ventricle being the intracardiac associations and presence of thoracic hemivertebrae, left lung and renal agenesis along with absence of left thumb and radius being the extracardiac associations.[5] The association with cor triatriatum has not been reported in literature so far. Though there are cases of

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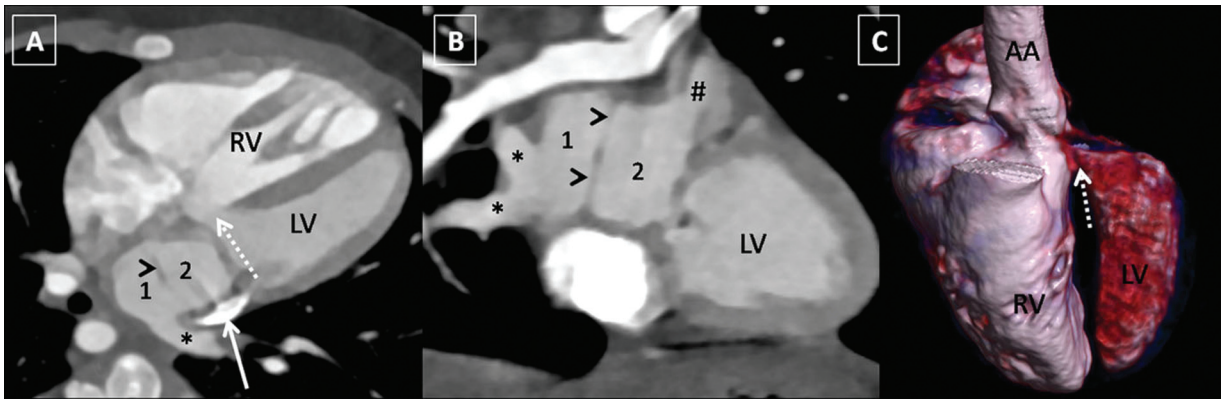

Fig. 1. Computed tomography angiography images (**A–C**) showing dilated right-sided cardiac chambers with presence of hypodense membrane (arrowheads) dividing left atrium into proximal (1) and distal (2) chambers receiving pulmonary veins (*) and left atrial appendage (#), respectively. Associated double outlet right ventricle (RV) with perimembranous ventricular septal defect (dashed arrow) and left superior vena cava (solid arrow) are also seen. AA, ascending aorta; LV, left ventricle.

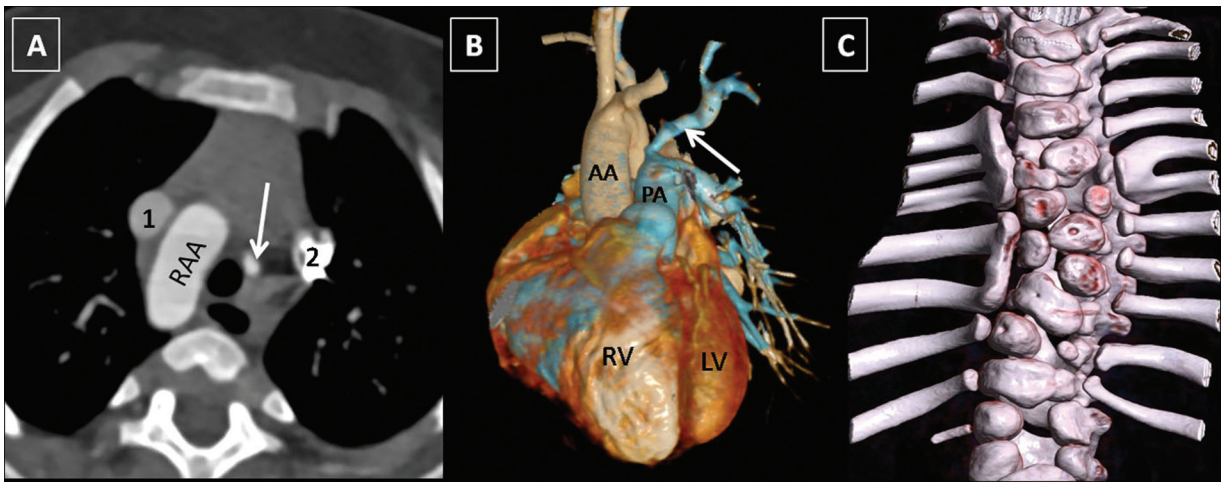

Fig. 2. Computed tomography angiography images (**A–C**) showing right-sided aortic arch (RAA) with isolated left subclavian artery (arrow), double superior vena cava (SVC; 1—right SVC; 2—left SVC) and multiple vertebral and rib anomalies. AA, ascending aorta; LV, left ventricle; PA, main pulmonary artery; RV, right ventricle.

simultaneous occurrence of cor triatriatum with TOF, in which its preoperative identification is of great importance as surgical repair of TOF may result in worsening of the previously unsuspected pulmonary venous obstruction associated with cor triatriatum, literature describes very few isolated reports of DORV with cortriatriatum.[6,7] However, its association with right sided aortic arch and isolated LSCA, as seen in present case, with vertebral segmentation anomalies has not been reported so far.

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Conflict of Interest
None declared.

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