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Ductus arteriosus location in relation to aortic arch position, branching pattern, and viscero-atrial situs

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ARTICLE INFORMATION

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AIM: To investigate ductus arteriosus (DA) location in relation to viscero-atrial situs and branching pattern of the right aortic arch (RAA).

MATERIALS AND METHODS: Computed tomography (CT) images of paediatric patients (May 2015 to July 2017, n=352) referred for evaluation of cardiovascular malformations were reviewed retrospectively. Patients with RAA were identified and images were analysed for documentation of the viscero-atrial situs, the branching pattern, DA location, and associated malformations.

RESULTS: The study included 64 RAA patients, 45 situs solitus, 11 situs ambiguous, and eight situs inversus. Mirror-image branching was observed in 34 situs solitus, 11 situs ambiguous, and seven situs inversus patients. Aberrant left subclavian artery (ALSCA) was present in 10 situs solitus and one situs inversus patient. One patient with situs solitus had isolated left subclavian artery. The location of the DA was identifiable in 32 patients (19 left and 13 right). In situs solitus with mirror-image branching, the DA was left in 10, right in four patients. In situs inversus and ambiguous with mirror-image branching, the DA was right in eight and left in two patients. In patients with ALSCA, the DA was left in seven and right in one patient. The location of the DA was identifiable in 85 patients with left aortic arch (left-sided in 84, right-sided in only one patient).

CONCLUSION: The location of DA in RAA varies according to viscero-atrial situs and branching pattern. In mirror-image branching, the DA is more commonly left-sided with situs solitus and more right-sided with abnormal viscero-atrial situs.

Introduction

The estimated prevalence of right aortic arch (RAA) is about 0.05–0.1% in the general population.[1] It may be an asymptomatic incidental finding during imaging, or present with tracheal and/or oesophageal compression due to a vascular ring formation. Certain cardiac malformations as well as viscero-atrial situs anomalies are associated with increased prevalence of RAA.[1,2] There are two main types of RAA: mirror-image branching and RAA with aberrant left subclavian artery (ALSCA).[3]

The ductus arteriosus (DA) is an important fetal vascular channel connecting the pulmonary to systemic circulations, rerouting blood away from the lungs. Normally, it starts to close soon after birth.

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The DA is typically located on the left side in the presence of a left-sided aortic arch (AA), connecting the proximal descending aorta and left pulmonary artery (PA)[3]; however, when the aortic arch is right-sided, the location of the DA becomes more variable (Fig 1); it could be on the left side, right side, or even bilateral as well as anterior or posterior.[3,4]

Clinically, the DA location in association with RAA is very important because it is the determinant factor in the formation of vascular rings,[3–6] and cross-sectional imaging, mainly by computed tomography (CT), is ideal for evaluation of vascular structures and airway compression. Imaging of the DA location in association with RAA is also important in the presence of a pathological patent DA (PDA) before surgical closure. In children with RAA and duct-dependent circulation, palliative stenting of the DA may be performed to maintain its patency before corrective surgery,[7] and CT may be urgently requested to assess the location and morphology of the DA. Follow-up after DA stenting may also be done using CT.

Interestingly, DA location and side in association with RAA has been also linked to the presence or absence of associated intra-cardiac shunts.[6] Another interesting point is the effect of viscero-atrial situs on the location and side of the DA, which remains fairly unclear. Although the imaging findings of RAA were discussed in several previous articles,[3–6,8–11] the variability of DA location in the presence of RAA is poorly described in the literature. The present CT angiography study was undertaken to assess DA location in relation to viscero-atrial situs and branching pattern of RAA.

Materials and methods

Study population

The institution's review board approved this retrospective study and patients' consent was waived. CT images were reviewed of all paediatric patients who were referred for cardiac CT angiography for the evaluation of aortic or cardiovascular anomalies in a tertiary referral university hospital between May 2015 and September 2017. All patients were referred to CT after abnormal fetal or post-natal echocardiography. Out of 352 patients with CT angiography in this time period, 64 patients had RAA, 283 had left aortic arch (AA), and five patients had double AA. The study population is part of a larger study on AA variations and anomalies in congenital heart disease.[12]

Cardiac CT technique

Cardiac CT angiography examinations were performed using a 128-multidetector CT machine (Philips Ingenuity Core[128], Philips Healthcare, The Netherlands). Patients were examined supine, during free breathing. Children >4 years were not sedated; children <4 years were sedated using chloral hydrate (0.5 ml/kg) 30 min prior to the scan. Intravenous anaesthesia was administered in few patients by a paediatric anaesthesiologist. The scan extended from the base of the neck to the upper abdomen. Low-dose retrospective electrocardiogram (ECG)-gated CT was acquired with the following parameters: 80 kV tube voltage, 150–200 mA tube current, 128×0.625 mm collimation, 0.67 mm section thickness, 1 mm increment, 0.2 mm/s table

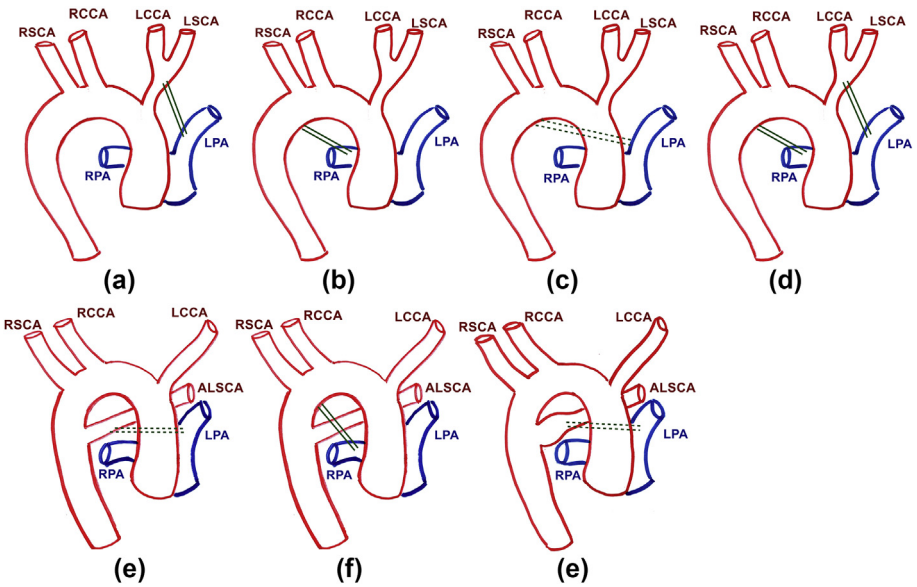

Figure 1 DA locations in RAA (green lines; dotted lines represent posterior location or retro-oesophageal course). (a–d) Mirror-image branching pattern: (a) left anterior DA; (b) right DA (true mirror image); (c) left retro-oesophageal DA; (d) bilateral DA. (e–g) Right aortic arch with aberrant left subclavian artery (ALSCA): (e) left posterior DA arising from ALSCA; (f) right DA; (g) left posterior DA from Kommerell's diverticulum. LPA, left PA; RCCA, right common carotid artery; RPA, right PA; RSCA, right subclavian artery.

feed, 0.4 seconds rotation time. Non-ionic iodinated contrast agent (iohexol, 350 mg iodine/ml; Omnipaque, GE health care, Ireland) was injected intravenously with a dose of 0.5–2 ml/kg at a rate of 1–2 ml/s. The scanning delay was 10 seconds. A delayed phase was performed in selected cases according to the indication. Images were processed on a separate workstation (Intellispace portal, version 5, Philips Healthcare) with multiplanar reformatted, volume-rendered, maximum and minimum intensity projection images.

Image analysis

CT images were analysed in consensus by two radiologists experienced in cardiac CT. All images were analysed for confirmation of the presence of RAA. The viscero-atrial situs was reported according to the segmental approach for imaging of congenital cardiac disease.[13] The aortic arch branching pattern of RAA was assigned to one of three types: mirror image, ALSCA, or isolated left subclavian artery (LSCA). A brachiobicephalic trunk was defined as a common origin of the left brachiocephalic and right common carotid arteries.[14] The origin of vertebral arteries was recorded in all cases. The location of the DA/ligamentum arteriosum, vascular ring formation, and associated cardiac malformations were recorded. The presence of a diverticulum, focal bulge, or tenting on CT was considered an indirect indicator of the site of the ligamentum arteriosum.[5] In cases where neither a PDA nor indirect signs were identifiable, the DA was considered absent. For the purpose of the present study, both the DA and ligamentum arteriosum are referred to as DA. A vascular ring was defined as the presence of vascular or ligamentous structures encircling the trachea and oesophagus with compression on the trachea on CT.

Results

Sixty-four patients were identified with RAA comprising 41 male and 23 female patients. The age ranged between 7 days and 11 years. The branching pattern was mirror image in 52 patients (Figs 2 and 3), ALSCA in 11 (Fig 4), and LSCA in one patient (Fig 5). The viscero-atrial situs and branching patterns are listed in Table 1. In addition, Kommerell's diverticulum was observed in three out of 11 patients with ALSCA, and a circumflex aorta was present in two patients (Fig 6).

In half the patients with RAA, the location of the DA could be identified as either a PDA (Fig 2), a DA with stent (Fig 3), or indirect signs indicating the ligamentum arteriosum location (Fig 4). The DA was non-identifiable in the remaining half. The locations of the DA in relation to the situs and branching pattern are listed in Table 2. A right DA connects the distal aortic arch and right PA in both the mirror-image branching pattern (true mirror image, Fig 2) and the ALSCA pattern. A left DA connects the left brachiocephalic artery near the origin of LSCA and either the left PA or the dome of the main PA in mirror-image

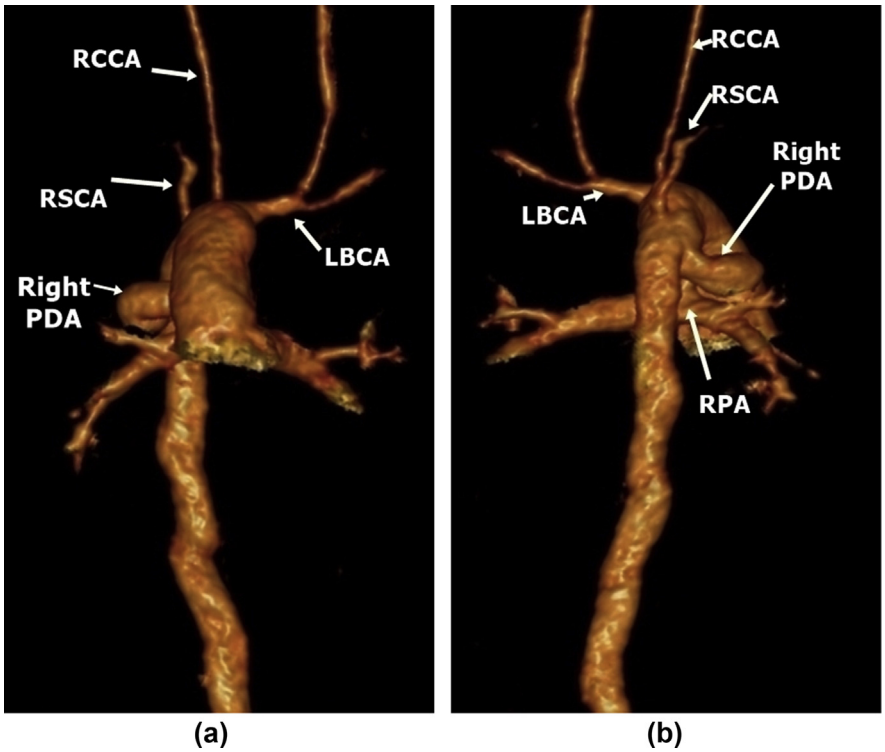

Figure 2 A 3-month-old boy with situs inversus. (a,b) Volume-rendered images showing RAA with mirror-image branches; left brachiocephalic (LBCA), right common carotid (RCCA) and right subclavian (RSCA) arteries. Note the right-sided PDA connecting the distal AA to the right PA.

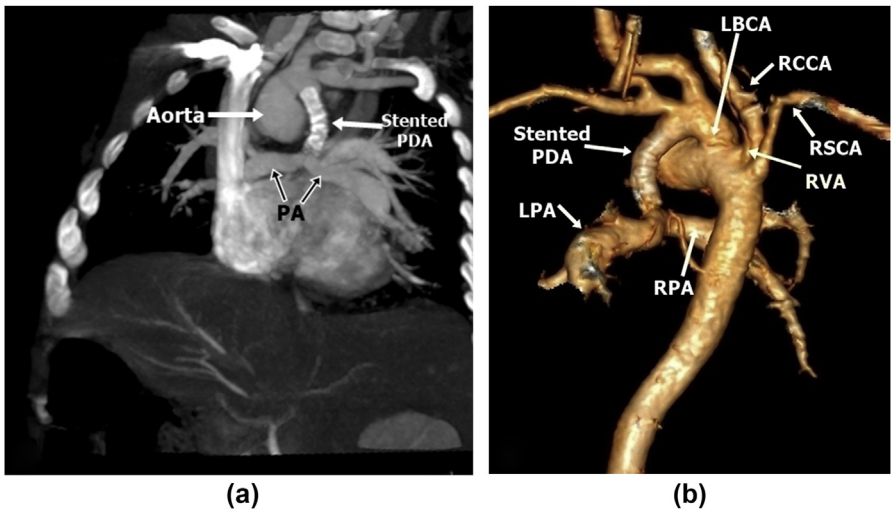

Figure 3 A 4-month-old boy with situs ambiguous (right isomerism). (a) Coronal MIP CT image. (b) Volume-rendered image. Right-sided arch with mirror-image branching pattern and direct origin of right vertebral artery (RVA). Stented left PDA arises from the left brachiocephalic artery (LBCA). LSCA, left subclavian artery; RCCA, right common carotid artery; RSCA, right subclavian artery; RPA, right PA; LPA, left PA.

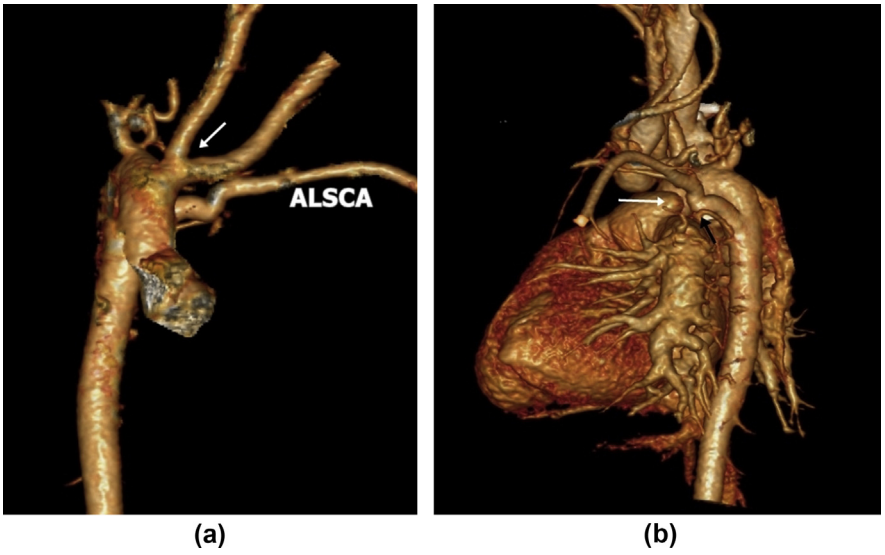

Figure 4 A 6-month-old boy with situs solitus. (a,b) Volume-rendered CT images showing RAA with ALSCA and bicarotid trunk (arrow in a). Note the indirect signs of ligamentum arteriosum in (b), with opposing tenting in the left subclavian artery and focal bulge in the dome of the main PA (black and white arrows, respectively).

branching (Fig 3), or Kommerell's diverticulum/LSCA to the left PA or main PA in the ALSCA branching pattern.

In the remaining 283 patients with left AA, the DA was identified in 85 patients (78 patients with situs solitus and seven with abnormal situs). In 84 patients, the DA was on the left side connecting the proximal descending aorta and left PA. In only one patient with situs solitus and tetralogy of Fallot, the DA remnant was right-sided and identified as a focal bulge in the right PA and another in the base of the right brachicephalic artery (Fig 7). In the five patients with double AA, the DA was not identifiable.

There was a vascular ring in four patients, in three of them the vascular ring was formed by an RAA and ALSCA and a left DA arising from Kommerell's diverticulum to join the left PA, findings were confirmed by operative reports. In the fourth patient, the ring was formed by a circumflex aorta (Fig 6).

Most of the patients had associated congenital cardiac malformations identifiable on cardiac CT (Table 3). Only five patients had no additional cardiac malformations.

Associated variants of the AA branching were observed in nine patients. In four patients, a left brachiobicephalic trunk or a common origin of the left brachiocephalic and right common carotid arteries was present. A right vertebral artery arising directly from the AA was observed in three patients (Fig 3). In one patient, there was a left brachiobicephalic trunk combined with a direct aortic origin of the right vertebral artery. In one patient with RAA and ALSCA,

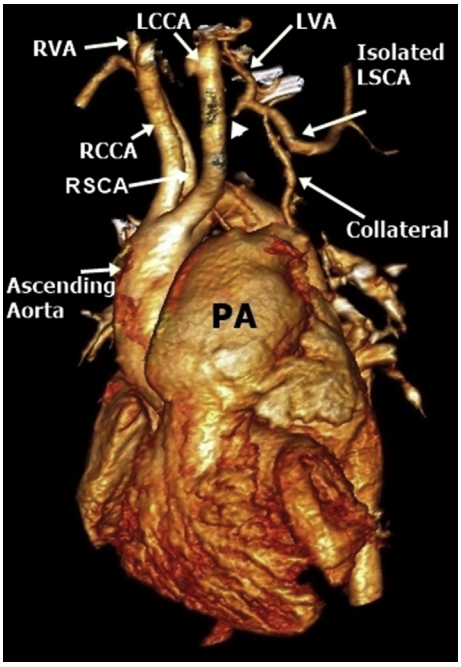

Figure 5 A 1-year-old boy with situs solitus. Volume-rendered CT image demonstrating RAA with isolated left subclavian artery (LSCA) supplied by a large aortic collateral. Arrowhead points to a presumed atretic cord between left common carotid artery (LCCA) and the isolated LSCA. The RAA gives origin to the LCCA, right common carotid (RCCA), and right subclavian artery (RSCA).

Table 1
Viscero-atrial situs and branching pattern in 64 patients with right aortic arch (RAA).

Branching pattern	Situs solitus *n*=45	Situs inversus *n*=8	Situs ambiguous *n*=11
Mirror-image branching	34 (76%)	7 (88%)	11 (100%)
Aberrant left subclavian artery	10 (22%)	1 (13%)	0
Isolated left subclavian artery	1 (2%)	0	0

there was a bi-carotid trunk (common trunk for both common carotid arteries; Fig 4).

Discussion

Branching patterns of RAA

The two common RAA types are mirror-image branching (type I) and RAA with an ALSCA (type II).[1] The ALSCA pattern is generally more common than mirror-image branching, but the latter is far more commonly associated with cardiac malformations.[3] Mirror-image branching was more common in the present study because most of the patients had associated malformations.

The uncommon RAA with isolated LSCA is classified as type III[1] (Fig 5). Typically, an isolated LSCA has no connection to the aorta or the left common carotid artery (LCCA), and arises from the left PA through a left DA.[15] Two more variants were described due to a persistent atretic cord between the LSCA and either the aorta or the LCCA. The former results in an intermediate variant between an isolated and an aberrant LSCA, and the latter results in an intermediate variant between an isolated LSCA and mirror-image branching.[16] Clinically, the three forms may be indistinguishable on angiography and could result in congenital subclavian steal syndrome if the LSCA is supplied by the left vertebral artery,[16] but only the first form could also result in pulmonary steal. The LSCA may also be supplied by collaterals from other vessels or from the aorta.[17]

Viscero-atrial situs and branching pattern in RAA

Mirror-image branching is the dominant type in association with abnormal viscero-atrial situs. In our own experience as well as in the literature, an RAA with ALSCA branching is rare in association with viscero-atrial situs anomalies. Only one patient in the present study had situs inversus and ALSCA pattern. Similarly, Tyczyński *et al.* reported only one patient associated with right isomerism in their series of 16 RAA with ALSCA patients, all the others were situs solitus.[18] In a previous autopsy study, all patients with RAA and asplenia had mirror-image branching pattern.[19]

DA location in RAA compared to LAA

In the 32 patients with RAA and identifiable DA, the DA was left-sided in 19 and right-sided in 13 patients. On the other hand, in 85 patients with LAA and identifiable DA, the DA was on the left side in 84, and only one patient had right-sided DA. It is worth noting that the presence of a right DA in the setting of a left AA is extremely rare, previously reported in few case reports only.[20]

DA location and branching pattern in RAA

In the mirror-image branching pattern, there are four possibilities for DA location (Fig 1). The commonest is a left DA connecting the anteriorly positioned left brachiocephalic artery near the base of left subclavian to the left PA, less commonly a right DA connects the distal aortic arch and the right PA (true mirror-image).[5] Rarely a retro-oesophageal DA connects the proximal descending aorta and the left PA.[3] Another rarely reported form is the presence of bilateral DA.[19]

In RAA with ALSCA, three possibilities for the DA have been described (Fig 1); the commonest is a left DA arising from Kommerell's diverticulum to join the left PA.[5] In cases with no Kommerell's diverticulum, the DA may be on the left connecting the ALSCA to the left PA or less commonly on the right connecting the distal aortic arch and the right PA.[19]

DA location and viscero-atrial situs in RAA

In the present series, the DA was more common on the left side in situs solitus cases. On the contrary, in association with abnormal viscero-atrial situs, the DA was more

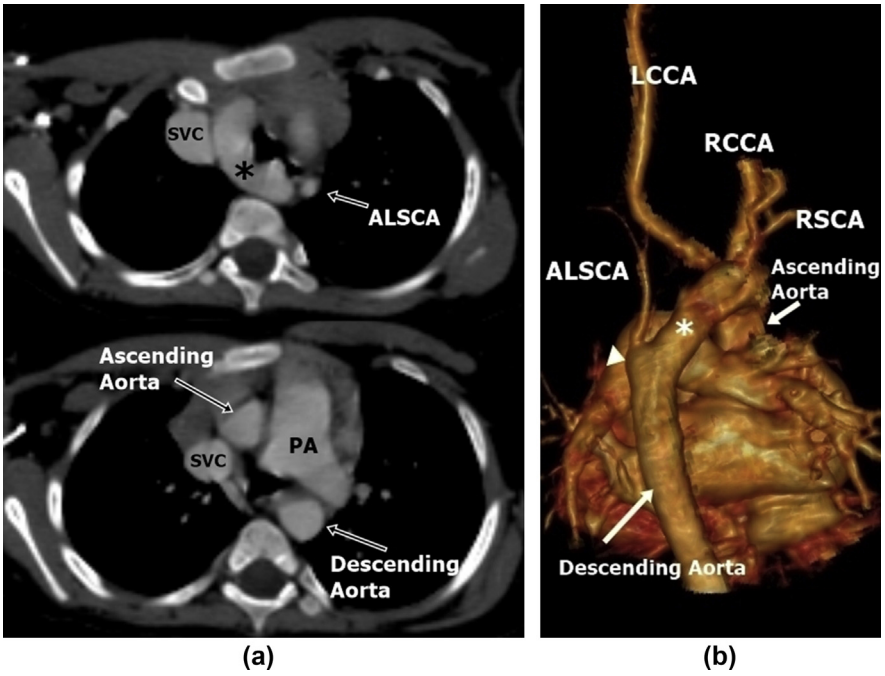

Figure 6 A 3-year-old girl with situs solitus. (a) Axial CT images showing RAA (asterisk) with retro-oesophageal course crossing to the left side above the carina "circumflex aorta". Note the ALSCA. (b) Volume-rendered image demonstrating the AA branching pattern and circumflex aorta crossing from right to left. Note the small left diverticulum (arrowhead) at the origin of the ALSCA. RCCA, right common carotid artery; RSCA, right subclavian artery.

Table 2
Location of the ductus arteriosus/ligamentum arteriosum (DA) in 32 cases with right aortic arch and identifiable DA.

Branching pattern	Situs solitus n=22		Situs inversus n=5		Situs ambiguous n=5	
	Right DA	Left DA	Right DA	Left DA	Right DA	Left DA
Mirror-image n=24	4	10	5	0	3	2
ALSCA with Kommerell's diverticulum n=3	0	3	0	0	0	0
ALSCA without Kommerell's diverticulum n=5	1	4	0	0	0	0

ALSCA, aberrant left subclavian artery.

common on the right side. To the authors' knowledge, there have been no reports on the relation between situs and DA location.

DA location and cardiac malformations in RAA

An interesting point is the relation between DA location and associated cardiac malformations. In the mirror-image branching pattern, a left DA is associated with cardiac malformations in 90% of cases while a right DA is usually not.[10] According to Weinberg,[6] a right DA is the only possibility for occurrence of isolated mirror-image branching, while a left DA can only occur in the presence of a significant defect that shunts fetal blood from right to left. Otherwise, most fetal right ventricular cardiac output would have been forced to take a long tortuous course passing through a left DA arising from the left PA to join the aorta at the base of the left brachiocephalic artery.[6] All patients with mirror-image branching in the present study had cardiac malformations, except one who had a right DA.

It was suggested that a congenitally absent DA must be associated with a major cardiac shunt to maintain fetal circulation.[6] Most of the present patients with absent DA on CT had associated shunts; however, two of the present patients had no DA without associated cardiac malformations. Other than true absence, an obliterated DA with no indirect signs will not be visualised on CT.

Right aortic arch and vascular rings

Vascular rings are more common in isolated RAA than in cases with associated cardiac malformations,[4,21] because the commonest form of vascular rings in RAA (ALSCA branching pattern with a Kommerell's diverticulum and a left DA),[22] is much less associated with cardiac malformations than mirror-image branching patterns.

Another form of vascular rings in an RAA is mirror-image branching with retro-oesophageal left DA,[3,4,23] which was not observed in this study. The third type of RAA associated with VR is a circumflex aorta, where the aorta crosses the midline taking a retro-oesophageal course above the level of the carina (Fig 6).[9] On the other hand, mirror-image branching is usually not associated with vascular rings

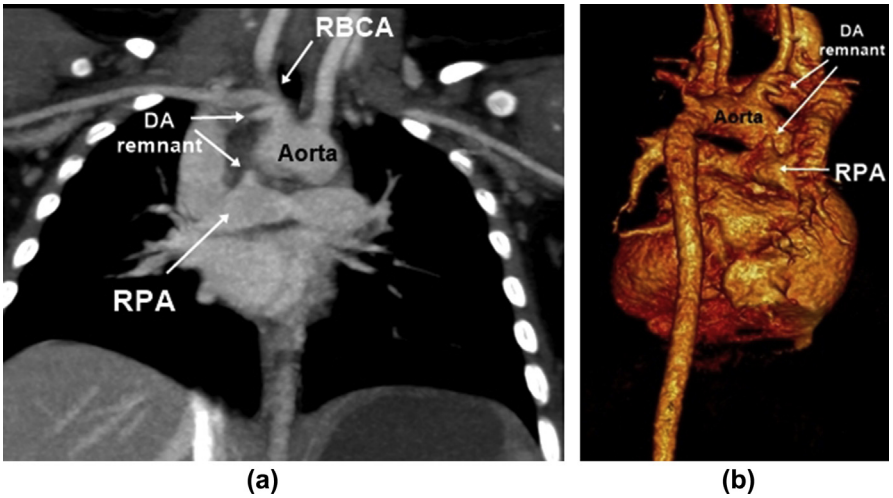

Figure 7 A 6-month-old girl with situs solitus. (a,b) Coronal MIP and volume-rendered CT images showing left-sided AA with evident DA remnant as opposing focal bulges in the right PA (RPA) and the base of the right brachicephalic artery (RBCA).

Table 3
Associated congenital cardiac malformations in 64 cases with RAA.

Congenital heart disease	Situs solitus n=45	Abnormal situs n=19
Tetralogy of Fallot n=32	32	0
Double outlet right ventricle n=10	1	9
Transposition of great arteries n=4	1	3
Hypoplastic left heart n=1	0	1
Total anomalous pulmonary venous drainage n=1	0	1
Pulmonary artery anomalies n=3	2	1
Cortriatrium n=1	0	1
Septal defects (atrial, ventricular, atrioventricular) n=5	3	2
Double inlet right ventricle n=1	1	0
Common atrium n=1	1	0
No cardiac malformations n=5	4	1

when the DA originates from the anteriorly positioned left brachiocephalic artery or is present on the right side.[4,5]

Associated cardiovascular malformations

An RAA is more commonly associated with cardiac malformations than isolated, most commonly tetralogy of Fallot, truncus arteriosus, tricuspid atresia, and transposition of the great arteries.[3,5] In the present series, the commonest was tetralogy of Fallot in situs solitus patients, and double outlet right ventricle in patients with abnormal viscero-atrial situs.

RAA branching variants

A left brachiobicephalic trunk[14] (common origin of the left brachiocephalic and right common carotid arteries) was observed in 8% of RAA. Direct aortic origin of the right vertebral artery was present in 6%. To the authors' knowledge, the prevalence of these variants in association with RAA has not been reported previously. The prevalence of their counterparts in left-sided AA has been reported in several studies, and showed significant ethnic variation,[24] yet it did not increase in association with other cardiac anomalies.[12]

Limitations

There are several limitations to this study: (a) the retrospective nature of the study; (2), the high prevalence of cardiac malformations, because most of the present patients were referred to CT after abnormal echocardiography; (3) the number of RAA patients with abnormal viscero-atrial situs is relatively small, so that the RAA in subgroups of situs anomalies could not be evaluated.

In conclusion, with increased use of CT in congenital cardiac and aortic anomalies, radiologists should be aware of the variable DA locations in RAA, their clinical significance, and the relation to branching pattern and viscero-atrial situs.

Conflicts of interest

The authors declare no conflict of interest.

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