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Review Article

Congenital abnormalities of the aortic arch: revisiting the 1964 Stewart classification☆

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Aberrant right innominate artery

ABSTRACT

The traditional classification of congenital aortic arch abnormalities was described by James Stewart and colleagues in 1964. Since that time, advances in diagnostic imaging technology have led to better delineation of the vasculature anatomy and the identification of previously unrecognized and unclassified anomalies. In this manuscript, we review the existing literature and propose a series of modifications to the original Stewart classification of congenital aortic arch abnormalities to incorporate this new knowledge. In brief, we propose the following modifications: (1) In Group I, we further divide subgroup B into left arch atretic and right arch atretic; (2) In Group II, we add three more subgroups, including aberrant right innominate artery, "isolated" right innominate artery (RIA), "isolated" right carotid artery with aberrant right subclavian artery; (3) In Groups I, II, and III, we add a subgroup of absence of both ductus arteriosus; and (4) In Group IV, we add three subgroups, including circumflex retro-esophageal aorta arch, persistent V aortic arch, and anomalous origin of pulmonary artery from ascending aorta.

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Abbreviations: AAO, ascending aorta; ARCH, aortic arch; CHD, congenital heart disease; DA, ductus arteriosus; DAO, descending aorta; IA, innominate artery; IAA, interrupted aortic arch; LCA, left common carotid artery; LDA, left ductus arteriosus; LDAO, left dorsal aorta; LIA, left innominate artery; LPA, left pulmonary artery; LPPA, left primitive pulmonary artery; LSA, left subclavian artery; MPA, main pulmonary artery; PDA, patent ductus arteriosus; RCA, right common carotid artery; RDA, right ductus arteriosus; RDAO, right dorsal aorta; RECA, right external carotid artery; RIA, right innominate artery; RICA, right internal carotid artery; RPA, right pulmonary artery; RPPA, right primitive pulmonary artery; RSA, right subclavian artery; SA, subclavian artery; VSD, ventricular septal defect.

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1. Introduction

Congenital abnormalities of the aortic arch refer to abnormal embryologic development of the aorta and its branches. Depending on the course and position of the aortic arch, anomalies may be characterized as interrupted, right-sided, left-sided, or double in configuration. They can also be characterized based on the order and branching pattern of the great vessels. The traditional classification of congenital aortic arch abnormalities was described by James Stewart and colleagues in 1964 "(Table 1)" [1]. This classification divided aortic arch anomalies into four major groups: (1) Group I, double aortic arch; (2) Group II, left aortic arch; (3) Group III, right aortic arch; and (4) Group IV, other and rare malformations of the aortic arch system. Since that time, major advances in diagnostic imaging technology have been made, including the introduction of thin-section computerized tomography (CT) with a shortened scanning time, multidetector computerized tomography (MDCT), and magnetic resonance imaging (MRI). This has led to improved diagnostic accuracy of congenital aortic arch abnormalities and better delineation of the vasculature anatomy. It has also led to the identification of previously unidentified anomalies not included in the original Stewart classification [2–8]. Moreover, advances in prenatal ultrasound has led to more cases of congenital aortic arch abnormalities being detected antenatally, providing opportunities for antenatal counseling and establishment of plans of care that include close monitoring of symptoms in the immediate postnatal period, early consultation with pediatric surgery, and early surgical intervention, if needed [9–11]. Taken together, these developments

Table 1
Traditional Stewart and Modified Classification of the Aortic Arch and its Branches

1964 Stewart classification*		Modifications of the Stewart classification			
Groups	**Subgroups**	**LDA**	**RDA**	**Bilateral DA**	**Absence of bilateral DA**
Group I (Double aortic arch)	A. Both arches patent	I A1	I A2	I A3	I A4
	B. Atresia in left arch	I B1	I B2	I B3	I B4
	C. Atresia in right arch	I C1	I C2	I C3	I C4
Group II (Left aortic arch)	A. Normal branch	II A1 (normal)	II A2	II A3	II A4
	B. Aberrant RSA	II B1	II B2	II B3	II B4
	C. Aberrant RIA	II C1	II C2	II C3	II C4
	D. Isolated RSA	--	II D2	II D3	--
	E. Isolated RIA	--	II E2	II E3	--
	F. Isolated RCA with aberrant RSA	--	II FB2	II FB3	--
Group III (Right aortic arch)	A. Mirror image branching	III A1	III A2	III A3	III A4
	B. Aberrant LSA	III B1	III B2	III B3	III B4
	C. Aberrant LIA	III C1	III C2	III C3	III C4
	D. Isolated LSA	III D1	--	III D3	--
	E. Isolated LIA	III E1	--	III E3	--
	F. Isolated LCA with aberrant LSA	III FB1	--	III FB3	--
Group IV (Other anomalies of the aortic arch system)	Circumflex retro-esophageal aortic arch	--			
	Coarctation of the aorta	--			
	Interrupted aortic arch	--			
	Persistent aortic arch V	--			
	Cervical aortic arch	--			
	Aberrant LPA (pulmonary artery sling)	--			
	Anomalous origin of pulmonary artery from ascending aorta and branching	--			

Black color designates original Stewart classification; blue designates modifications (additional subgroups).
* From reference [1] Stewart JR, Kincaid OW, Edwards JE. An atlas of vascular rings and related malformation of the aortic arch system. Springfield, IL: Charles C. Thomas, 1964. pp. 1–170.

make it necessary to revisit and modify the original 1964 Stewart classification of congenital aortic arch abnormalities.

In this manuscript, we review the existing literature and propose a series of modifications to the original Stewart classification of congenital aortic arch abnormalities to incorporate the new developments in the field. These modifications are summarized in (Table 1). In brief, we propose to: (1) In Group I, further divide subgroup B into left arch atretic and right arch atretic; (2) In Group II, add three more subgroups, including aberrant right innominate artery (RIA), "isolated" RIA, "isolated" right carotid artery (RCA) with aberrant right subclavian artery (RSA); (3) In Groups I, II, and III, add a subgroup of absence of both ductus arteriosus (DA); and (4) In Group IV, add three subgroups, including circumflex retro-esophageal aorta arch, persistent V aortic arch, and anomalous origin of pulmonary artery (PA) from ascending aorta (AAO). Throughout the manuscript, we include diagrams to describe the embryonic origins and anatomy of each of the congenital lesions.

2. Normal embryonic development of the aortic arch

The normal embryologic development of the normal aorta and its branches are shown in (Fig. 1). (Fig. 1A) shows the hypothetical configuration of the cardiothoracic vasculature of the early embryo as proposed by Congdon [12] and Barry [13], and is made up of 6 pairs of pharyngeal arches (analogous to the branchial arches in fish, designated I through VI) connecting the two primitive ventral and dorsal aortas. The development of branchial arches begins by the second week of gestation and continues till the seventh week. The two dorsal aortas fuse to form a single vessel, whereas the 6 pairs of aortic (pharyngeal) arches appear and regress at different times. Aortic arches I and II form and disappear first. Aortic arch III and portions of the ventral and dorsal aortic arches form the common carotid arteries, external carotid arteries, and internal carotid arteries, respectively. Aortic arch IV becomes the aortic arch. Aortic arch V becomes atretic or never fully develops. The distal portion of the right aortic arch VI disappears, while the proximal portion forms the right pulmonary artery (RPA). The proximal portion of the left aortic arch VI forms the left pulmonary artery (LPA), while the left distal portion persists as the ductus arteriosus (DA). The intersegmental arteries migrate and form the subclavian arteries. The normal aortic arch and its branches (Fig. 1C) result from regression of regions 8 and 9 (right descending aorta) and region 10 (RDA which is the distal portion of the right aortic arch VI) and persistence of the left aortic arch, which gives rise to the right innominate artery (RIA), left carotid artery (LCA) and left subclavian artery (LSA), in that order, along with the left ductus arteriosus (LDA). The normal aortic arch is classified as subgroup IIA1 (see Table 1).

3. GROUP I: Double aortic arch malformations

According to the double aortic arch hypothesis proposed by Edwards [14] two aortic arches (designated LARCH and RARCH) exist in early embryonic development that encircle the trachea and esophagus (Fig. 1B). Each arch has its own DA, common carotid artery, and subclavian artery. The DAO forms in the midline by fusion of the two arches. Specific congenital aortic arch abnormalities can be explained by interruption of the double aortic arch at different locations (summarized in Fig. 1B). If the RARCH does not regress as expected, a double aortic arch malformation or complete vascular ring will result. In such instances, the RARCH is typically dominant and larger than the LARCH (75% of cases) [15]. Each arch develops a common carotid artery and subclavian artery,

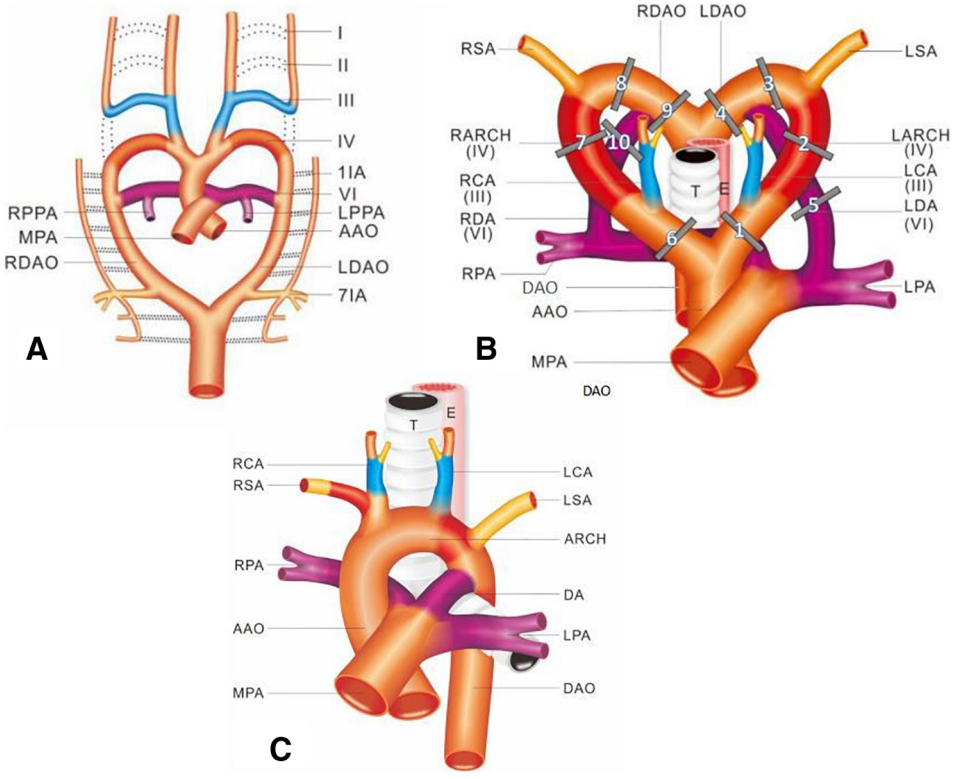

Fig. 1. Normal embryologic development of the aortic arch and its branches. **A:** Hypothetical configuration of the cardiothoracic vasculature in early embryogenesis showing the 6 pairs of pharyngeal (aortic) arches, labeled I through VI. **B:** Template showing the arrangement of the major vessels making up the aortic arch and its branches in the absence of regression. **C:** The normal aortic arch and its branches result from regression of regions 8 and 9 (RDAO) and region 10 (RDA) which is the distal portion of the right aortic arch (VI). The normal aortic arch is classified as subgroup IIA1 (see Table 1). Colors reflect the embryologic origins of different portions of the aortic arch and its branches (blue = aortic arch III; red = aortic arch IV; purple = aortic arch VI; yellow = seventh dorsal intersegmental artery). Description: The DAO is located behind the E and T. In the template absence of regression, an aortic arch can be seen on both sides of the E and T, known as the right arch and left arch, which arise from a single vessel, later to become the AAO, and end in a single vessel, later to become the DAO. Each of the arches has a separate DA connecting the ipsilateral PA and DAO. The gray regions in Fig. 1B identify the 10 key locations where regression or atresia occurs, and includes: (1) the root of LARCH; (2) the middle of LARCH; (3) the proximal portion of LDAO; (4) the distal portion of LDAO; (5) LDA (the distal portion of left aortic arch VI); (6) the root of RARCH; (7) the middle of RARCH; (8) the proximal portion of RDAO; (9) the distal portion of RDAO; and (10) RDA (the distal portion of the right aortic arch VI).

Table 2
Classification of Double Aortic Arch Abnormalities

Sub-group	Aortic Arch	Ductus arteriosus	Embryonic development	Vascular ring
I A1	Both arches patent	LDA patent	Regression of distal portion of right VI ARCH	complete
I A2	Both arches patent	RDA patent	Regression of distal portion of left VI ARCH	complete
I A3	Both arches patent	Bilateral DA patent	No regression of VI ARCH	complete
I A4	Both arches patent	Bilateral DA regression	Regression of distal portions of both VI ARCHES	complete
I B1	Left arch atretic	LDA patent	Regression of distal portion of right VI ARCH and atretic zone of distal LDAO, or proximal LDAO, or left IV ARCH, or root of LARCH	complete
I B2	Left arch atretic	RDA patent	Regression of distal portion of left VI ARCH and atretic zone of distal LDAO, or proximal LDAO, or left IV ARCH, or root of LARCH	complete
I B3	Left arch atretic	Bilateral DA patent	Atretic zone of distal LDAO, or proximal LDAO, or left IV ARCH, or root of LARCH	complete
I B4	Left arch atretic	Bilateral DA regression	Regression of distal portions of both VI ARCHES and atretic zone of distal LDAO, or proximal LDAO, or left IV ARCH, or root of LARCH	complete
I C1	Right arch atretic	LDA patent	Regression of distal portion of right VI ARCH and atretic zone of distal RDAO, or proximal RDAO, or right IV ARCH, or root of RARCH	complete
I C2	Right arch atretic	RDA patent	Regression of distal portion of left VI ARCH and atretic zone of distal RDAO, or proximal RDAO, or right IV ARCH, or root of RARCH	complete
I C3	Right arch atretic	Bilateral DA patent	Atretic zone of distal RDAO, or proximal RDAO, or right IV ARCH, or root of RARCH	complete
I C4	Right arch atretic	Bilateral DA regression	Regression of distal portions of both VI ARCHES and atretic zone of distal RDAO, or proximal RDAO, or right IV ARCH, or root of RARCH	complete

Black color designates original 1964 Stewart classification; blue designates modifications (additional subgroups).

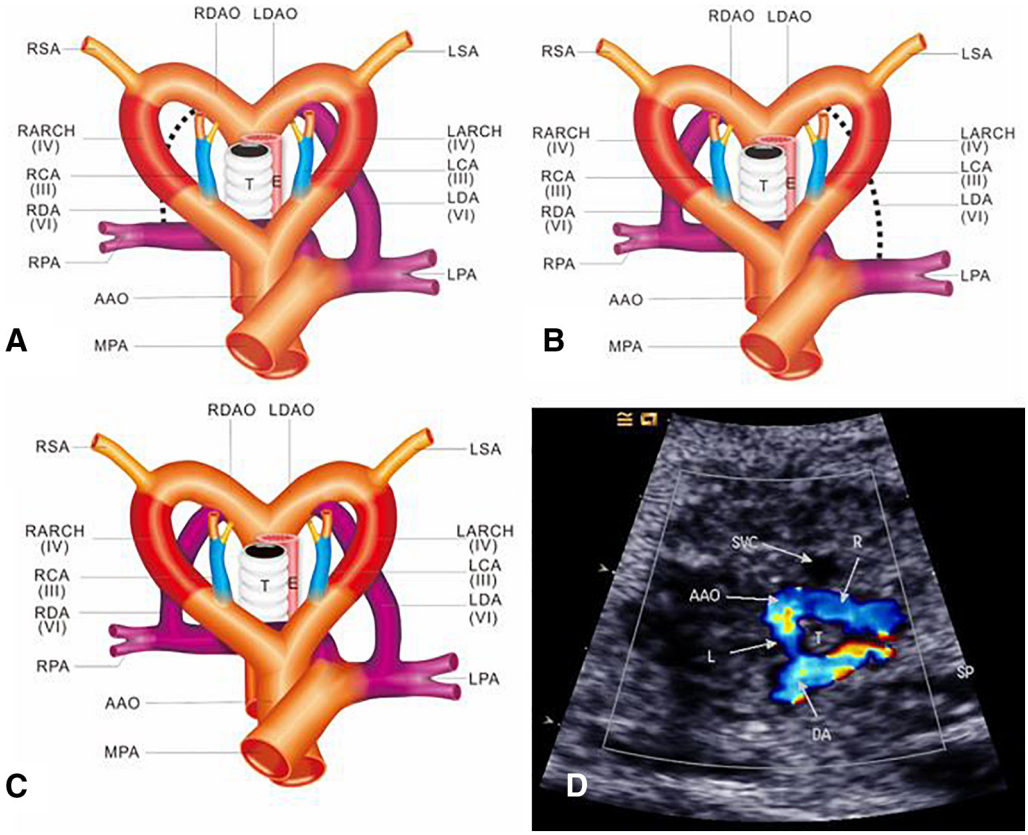

Fig. 2. GROUP I, Subgroup A: Persistence of both aortic arches. Congenital abnormalities shown in these figures include subgroups IA1 (**A**), IA2 (**B**), IA3 (**C**) and IA1 (**D**) (Table 2). Double aortic arches were shown on 3VT view (**D**) in a fetus at 24 weeks gestation. Color Doppler showed the left (L) and right arches (R) encircled the trachea and esophagus forming a complete vascular ring. The DA was on the left of the LARCH.

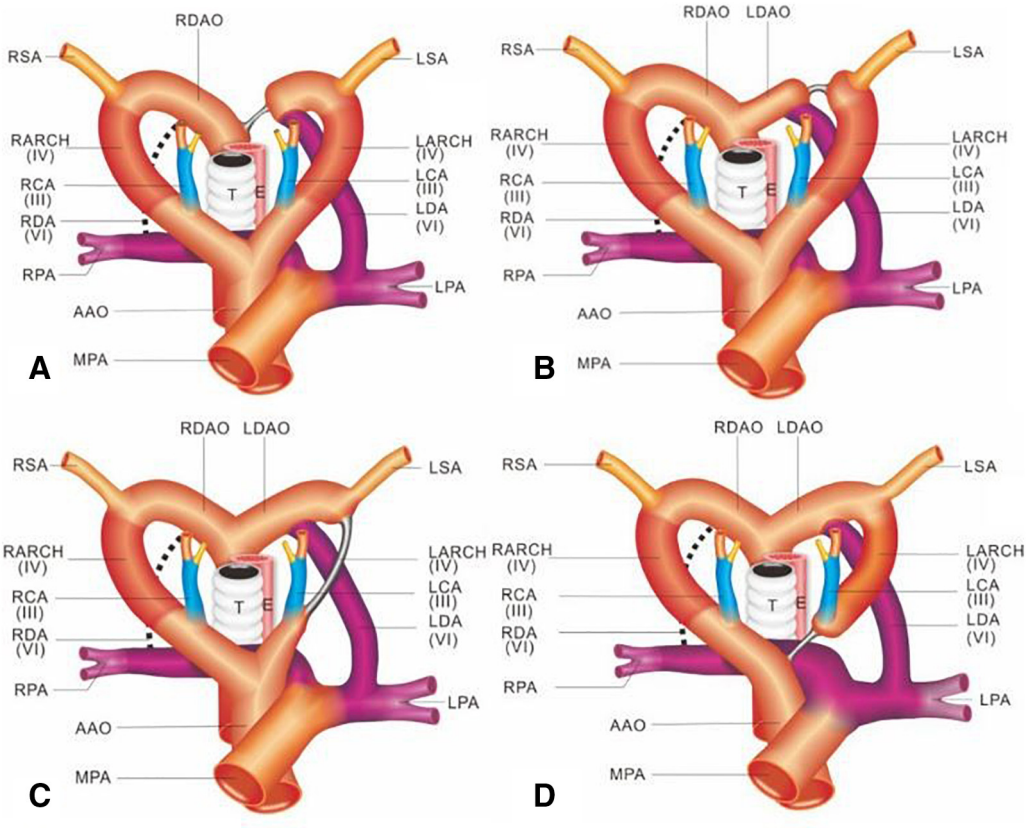

Fig. 3. GROUP I, Subgroup B: Double aortic arches with atresia in the left arch. Congenital abnormalities shown in these figures are all included under subgroup IB1, which has 4 subtypes based on the location of the atretic zone: $IB1_1$ (**A**), $IB1_2$ (**B**), $IB1_3$ (**C**), and $IB1_4$ (**D**) (Table 2).

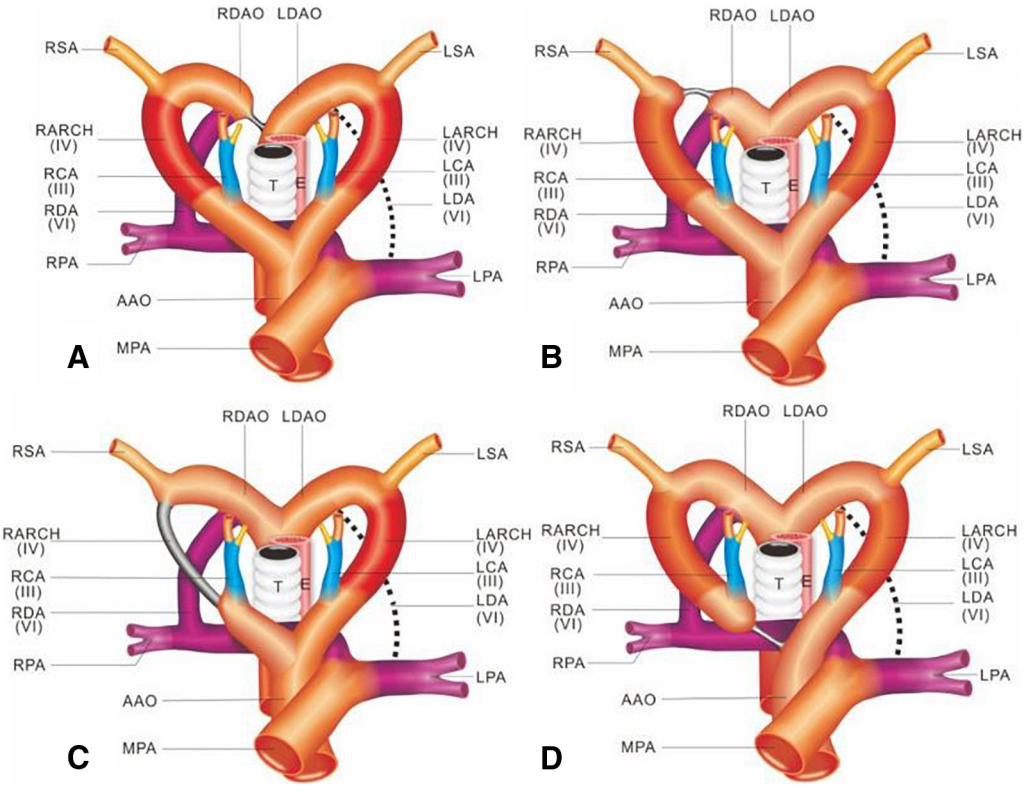

Fig. 4. GROUP I, Subgroup C: Double aortic arches with atresia in the right arch. Congenital abnormalities shown in these figures are all included under subgroup IC2, which has 4 subtypes based on the location of the atretic zone: $IC2_1$ (**A**), $IC2_2$ (**B**), $IC2_3$ (**C**), and $IC2_4$ (**D**) (Table 2).

Table 3
Classification of Left Aortic Arch Abnormalities

Sub-group	Branch	Ductus arteriosus	Embryonic development	Vascular ring
II A1 (Normal)	Normal branch	LDA patent	Regression RDAO and distal portion of right VI ARCH	No
II A2a	Normal branch	RDA patent	Regression of distal portion of RDOA (a) and distal portion of left VI ARCH	No
II A2b	Normal branch	RDA patent	Regression of proximal portion of RDOA (b) and distal portion of left VI ARCH	complete
II A3a	Normal branch	Bilateral DA patent	Regression of distal portion of RDAO (a)	No
II A3b	Normal branch	Bilateral DA patent	Regression of proximal portion of RDAO (b)	complete
II A4	Normal branch	Bilateral DA regression	Regression of RDAO and distal portions of both VI ARCHES	No
II B1	Aberrant RSA	LDA patent	Regression of right IV ARCH and distal portion of right VI ARCH	incomplete
II B2	Aberrant RSA	RDA patent	Regression of right IV ARCH and distal portion of left VI ARCH	complete
II B3	Aberrant RSA	Bilateral DA patent	Regression of right IV ARCH	complete
II B4	Aberrant RSA	Bilateral DA regression	Regression of right IV ARCH and distal portions of both VI ARCHES	incomplete
II C1	Aberrant RIA	LDA patent	Regression of root of right ARCH and distal portion of right VI ARCH	incomplete
II C2	Aberrant RIA	RDA patent	Regression of root of right ARCH and distal portion of left VI ARCH	complete
II C3	Aberrant RIA	Bilateral DA patent	Regression of root of right ARCH	complete
II C4	Aberrant RIA	Bilateral DA regression	Regression of root of right ARCH and distal portions of both VI ARCHES	incomplete
II D2	Isolated RSA	RDA patent	Regression of right IV ARCH, distal portion of RDOA, and distal portion of left VI ARCH	No
II D3	Isolated RSA	Bilateral DA patent	Regression of right IV ARCH and distal portion of RDAO	No
II E2	Isolated RIA	RDA patent	Regression of root of right ARCH, distal portion of RDAO, and distal portion of left VI ARCH	No
II E3	Isolated RIA	Bilateral DA patent	Regression of root of right ARCH and distal portion of RDAO	No
II FB2	Isolated RCA with aberrant RSA	RDA patent	Regression of root of right ARCH, proximal portion of RDAO, and distal portion of left VI ARCH	incomplete
II FB3	Isolated RCA with aberrant RSA	Bilateral DA patent	Regression of root of right ARCH and proximal portion of RDAO	incomplete

Black color designates original 1964 Stewart classification; blue designates modifications (additional subgroups).

and these are usually symmetrically arranged. Although the DA may remain patent on both sides, it is most common for only one DA to remain patent, usually the left. Rarely, both DA will regress and become atretic. The DAO usually develops towards one side, typically the side with patent DA. The classification of congenital double aortic arch abnormalities is summarized in (Table 2), and are discussed in detail below. In cases of double aortic arch with atresia in one arch, the atretic zone can be the distal portion of the dorsal aorta, the proximal portion of the dorsal aorta, the middle of ARCH, or the root of ARCH.

Double aortic arch is rarely associated with CHD. When present, tetralogy of Fallot is the most common disorder followed by transposition of the great arteries [16]. Infants born with a double aortic arch typically present early in life with a barking cough and respiratory stridor. In cases of double aortic arch in which one arch is atretic, the atretic arch can persistent as a fibrous or ligamentous remnant, which needs to be differentiated from another less common lesion, the circumflex retro-esophageal aortic arch. Imaging studies can usually only identify patent vessels and, as such, are not capable of directly visualizing the ligamentous atretic segment.

3.1. GROUP I, Subgroup A: Both aortic arches are patent

3.1.1. Both aortic arches are patent with a patent LDA (Subgroup IA1, Fig. 2A). Such cases are characterized by the presence of both aortic arches, left and right dorsal aortas, and a patent LDA, which results from regression at region 10 only.

3.1.2. Both aortic arches are patent with a patent RDA (Subgroup IA2, Fig. 2B). Such cases are characterized by the presence of both aortic arches, left and right dorsal aortas, and a patent RDA, which results from regression at region 5 only.

3.1.3. Both aortic arches are patent with bilateral patent DA (Subgroup IA3, Fig. 2C). Such cases are characterized by the presence of both aortic arches, left and right dorsal aortas, and bilateral patent DA.

3.1.4. Both aortic arches are patent with regression of the DA bilaterally (Proposed Subgroup IA4). Such cases are characterized by the presence of both aortic arches, left and right dorsal aortas, and regression of both DA, which results from regression at region 5 and 10 only.

3.2. GROUP I, Subgroup B: Double aortic arches with atresia in the left arch

3.2.1. Double aortic arches with atresia of LARCH and regression of RDA, resulting in a patent RARCH and LDA (Subgroup IB1). In such cases, embryologic regression occurred at the distal portion of right VI ARCH. The atresia of LARCH can occur at distal portion of LDAO (Subgroup $IB1_1$, Fig. 3A), the proximal portion of LDAO (Subgroup $IB1_2$, Fig. 3B), the left IV ARCH (Subgroup $IB1_3$, Fig. 3C), or at the root of LARCH (Subgroup $IB1_4$, Fig. 3D).

3.2.2. Double aortic arches with atresia of LARCH and regression of LDA, resulting in a patent RARCH and RDA (Subgroup IB2). In such case, the embryologic regression occurred at distal portion of left VI ARCH. The atresia of LARCH can occur at the distal portion of the LDAO (Subgroup $IB2_1$), proximal portion of LDAO (Subgroup $IB2_2$), the left IV ARCH (Subgroup $IB2_3$), or the root of LARCH (Subgroup $IB2_4$).

3.2.3. Double aortic arches with atresia of LARCH, resulting in a patent RARCH with bilateral DA (Subgroup IB3). The atresia of LARCH can occur at the distal LDAO (Subgroup $IB3_1$), proximal portion of LDAO (Subgroup $IB3_2$), the left IV ARCH (Subgroup $IB3_3$), or the root of LARCH (Subgroup $IB3_4$).

3.2.4. Double aortic arches the atresia of LARCH and regression of bilateral DA, resulting in a patent RARCH (Subgroup IB4). Both the left and right VI ARCHES regressed embryonically. The atresia of LARCH can occur at the distal portion of LDAO (Subgroup $IB4_1$), the proximal portion of LDAO (Subgroup $IB4_2$), the left IV ARCH (Subgroup $IB4_3$), or the root of LARCH (Subgroup $IB4_4$).

3.3. GROUP I, Subgroup C: Double aortic arches with atresia in the right arch

3.3.1. Double aortic arches with atresia of RARCH and regression of RDA, resulting in a patent LARCH and LDA (Subgroup IC1). The atresia of RARCH can occur at the distal of RDAO (Subgroup $IC1_1$), the proximal portion of RDAO (Subgroup $IC1_2$), the right IV ARCH (Subgroup $IC1_3$), or the root of RARCH (Subgroup $IC1_4$).

3.3.2. Double aortic arches with the atresia of RARCH and regression of LDA, resulting in a patent LARCH and RDA (Subgroup IC2). The atresia of RARCH can occur at the distal portion of RDAO (Subgroup $IC2_1$, Fig. 4A), the proximal portion RDAO (Subgroup $IC2_2$, Fig. 4B), the right IV ARCH (Subgroup $IC2_3$, Fig. 4C), or the root of RARCH (Subgroup $IC2_4$, Fig. 4D).

3.3.3. Double aortic arches with atresia of RARCH, resulting in a patent LARCH and bilateral DA (Subgroup 1C3). The atresia of RARCH can occur at the distal portion of RDAO Subgroup $1C3_1$), the proximal portion RADO (Subgroup $1C3_2$), the right IV ARCH Subgroup $1C3_3$), or the root of RARCH Subgroup $1C3_4$).

3.3.4. Double aortic arches with atresia of RARCH and regression of bilateral DA, resulting in a patent LARCH (Subgroup 1C4). Both the left and right VI ARCHES regressed embryonically. The atresia of RARCH can occur at the distal portion of RADO (Subgroup $1C4_1$), the proximal portion of RADO (Subgroup $1C4_2$), the right IV ARCH (Subgroup $1C4_3$), or the root of RARCH (Subgroup $1C4_4$).

4. GROUP II: Congenital Malformations of the Left Aortic Arch (LARCH)

The classification of congenital abnormalities of the LARCH based on aberrant embryologic development of the heart is summarized in (Table 3), and is discussed in detail below. Some of these have not been previously reported in the literature. The most common type of LARCH malformation is the aberrant RSA with patent LDA. Normal embryologic development results in formation of a dominant LARCH with regression of the RARCH at its root, its middle, and its proximal segment along with regression of the distal portion of the RDAO.

Congenital malformations of the left aortic arch do not typically result in a complete vascular ring as there is no persistent vessel on the right side of the trachea. LARCH with an aberrant RSA is usually an isolated lesion, but can rarely be seen in association with other congenital anomalies, including aortic coarctation, patent ductus arteriosus (PDA), ventricular septal defect (VSD), and carotid or vertebral artery anomalies [17]. It may result in esophageal compression due to an aberrant retroesophageal course. In cases with LARCH and isolation of the subclavian artery, patients are usually asymptomatic, although the may occasionally develop features of left upper limb ischemic pain [18].

4.1. GROUP II, Subgroup A: Left aortic arch with normal arch and branches

4.1.1. Subgroup IIA1 represents the normal aortic arch with normal branches (Fig. 1C).

4.1.2. Subgroup IIA2 represents LARCH malformations with patent RDA. It can be subdivided into: (i) LARCH with a patent RDA (Subgroup IIA2a) and results from regression of the distal portion of the RDAO and LDA, with embryologic regression occurred at regions 5 and 9. The aortic arch passes in front of the left bronchus and the trachea. From proximal to distal, the aortic arch gives rise to the IA, LCA and LSA. The distal portion of the RDA connects the RIA at the origin of the RSA; as such, a vascular ring is not formed. (ii) LARCH with a patient RDA (Subgroup IIA2b) that results from regression of the proximal portion of the RDAO and LDA, with embryologic regression occurred at region 5 and 8. The aortic arch passes in front of the left bronchus and trachea. It gives rise to the IA, LCA and LSA. The distal portion of the RDA connects with the distal portion of the RDAO. Therefore, the LARCH, RDAO and RDA encircle the trachea and esophagus forming a complete vascular ring.

4.1.3. Subgroup IIA3 represents LARCH malformations with bilateral patent DA. It can be subdivided into: (i) LARCH with bilateral DA (Subgroup IIA3a) which results from regression of the distal portion of the RDAO, with embryologic regression occurred at region 9. The aortic arch passes in front of the left bronchus and trachea. From proximal to distal, the arch gives rise to the RIA, LCA, and LSA. The RDA connects to the origin of the RSA and RIA, and the LDA connects to the origin of the DAO; therefore, no vascular ring is formed. (ii) LARCH with bilateral DA (Subgroup IIA3b) which results from regression of the proximal portion of the RDAO, with embryologic regression occurred at region 8. The aortic arch passes in front of the left bronchus and trachea. From proximal to distal, the arch gives rise to the RIA, LCA, and LSA. The RDA connects to the distal portion of RDAO, and the LDA connects the origin of the DAO. As such, the LARCH, the distal portion of the RDAO, and the RDA encircle the trachea and esophagus forming a complete vascular ring.

4.1.4. LARCH with regression of bilateral DA (Proposed Subgroup IIA4), which is due to regression of the RDAO and bilateral DA, with embryologic regression occurred at region 5, 8, 9 and 10. The aortic arch passes in front of the left bronchus and trachea. From proximal to distal, the arch gives rise to the RIA, LCA, and LSA; there is no DA. As such, no vascular ring is formed.

4.2. GROUP II, Subgroup B: Left aortic arch with aberrant RSA

4.2.1. LARCH with aberrant RSA and LDA (Subgroup IIB1) results from regression of the middle portion of the RARCH and RDA, with embryologic regression occurred at regions 7 and 10. The aortic arch passes in front of the left bronchus and trachea. From proximal to distal, the arch gives rise to the RCA, LCA, LSA, and the aberrant RSA. The LDA passes in front of the left bronchus and connects to the origin of the DAO. The RSA connects to the RDAO. As such, the LARCH and RSA surround the esophagus and trachea, forming an incomplete vascular ring.

4.2.2. LARCH with aberrant RSA and RDA (Subgroup IIB2) results from regression of the middle portion of the RARCH and LDA, with embryologic regression occurred at regions 5 and 7. The aortic arch passes in front of the left bronchus and trachea. From proximal to distal, the arch gives rise to the RCA, LCA, LSA, and the aberrant RSA. The RDA passes in front of the right bronchus and connects to the RSA, which connects in turn to the RDAO. Therefore, the LARCH, RSA and RDA encircle the trachea and esophagus, forming a complete vascular ring.

4.2.3. LARCH with aberrant RSA and bilateral DA (Subgroup IIB3) results from regression of the middle portion of the RARCH, with embryologic regression occurred at region 7. The aortic arch passes in front of the left bronchus and trachea. From proximal to distal, the arch gives rise to the RCA, LCA, LSA, and aberrant RSA. The RDA passes in front of the right bronchus and connects to the RSA, which connects in turn to the RDAO. The LDA passes in front of the left bronchus and

connects to the origin of the DAO. Therefore, the LARCH, RSA, and RDA encircle the esophagus and trachea, forming a complete vascular ring.

4.2.4. LARCH with aberrant RSA and regression of both DA (Proposed Subgroups IIB4) results from regression of the middle portion of the RARCH and both DA, with embryologic regression occurred at regions 5, 7 and 10. The aortic arch passes in front of the left bronchus and trachea. From proximal to distal, the arch gives rise to the RCA, LCA, LSA, and aberrant RSA. The RSA connects to the RDAO. The LARCH and RSA surround the esophagus and trachea, forming an incomplete vascular ring.

4.3. GROUP II, Subgroup C: Left aortic arch with aberrant RIA

4.3.1. LARCH with aberrant right innominate artery (RIA) and LDA (Proposed Subgroup IIC1) results from regression of the root of the RARCH and RDA, with embryologic regression occurred at region 10. The aortic arch passes in front of the left bronchus and trachea. From proximal to distal, the arch gives rise to the LCA, LSA, and aberrant RIA. The RIA connects to the RDAO. The LDA passes in front of the left bronchus and connects to the origin of the DAO. Therefore, the LARCH and RIA surround the esophagus and trachea, forming an incomplete vascular ring.

4.3.2. LARCH with aberrant RIA and RDA (Proposed Subgroup IIC2) results from regression of the root of the RARCH and LDA, with embryologic regression occurred at region 5. The aortic arch passes in front of the left bronchus and trachea. From proximal to distal, the arch gives rise to the LCA, LSA, and aberrant RIA. The RIA connects to the RDAO. The RDA passes in front of the right bronchus and connects to the RIA. Therefore, the LARCH, RIA, and RDA encircle the esophagus and trachea, forming a complete vascular ring.

4.3.3. LARCH with aberrant RIA and bilateral DA (Subgroup IIC3) results from regression of the root of the RARCH. The aortic arch passes in front of the left bronchus and trachea. From proximal to distal, the arch gives rise to the LCA, LSA, and aberrant RIA. The RIA connects to the RDAO; the RDA passes in front of the right bronchus and connects to the RIA. The LDA passes in front of the left bronchus and connects to the origin of the DAO. As such, the LARCH, RIA, and RDA encircle the esophagus and trachea, forming a complete vascular ring.

4.3.4. LARCH with aberrant RIA and regression of both DA (Proposed Subgroup IIC4) results from regression of the root of the RARCH and

Table 4
Classification of Right Aortic Arch Abnormalities

Sub-group	Branch	Ductus arteriosus	Embryonic development	Vascular ring
III A1a	Mirror image branching	LDA patent	Regression of distal portion of LDOA (a) and distal portion of right VI ARCH	No
III A1b	Mirror image branching	LDA patent	Regression of proximal portion of LDOA (b) and distal portion of right VI ARCH	complete
III A2	Mirror image branching	RDA patent	Regression of LDAO and distal portion of left VI ARCH	No
III A3a	Mirror image branching	Bilateral DA patent	Regression of distal portion of LDAO (a)	No
III A3b	Mirror image branching	Bilateral DA patent	Regression of proximal portion of LDAO (b)	complete
III A4	Mirror image branching	Bilateral DA regression	Regression of LDAO and distal portions of both VI ARCHES	No
III B1	Aberrant LSA	LDA patent	Regression of left IV ARCH and distal portion of right VI ARCH	complete
III B2	Aberrant LSA	RDA patent	Regression of left IV ARCH and distal portion of left VI ARCH	incomplete
III B3	Aberrant LSA	Bilateral DA patent	Regression of left IV ARCH	complete
III B4	Aberrant LSA	Bilateral DA regression	Regression of left IV ARCH and distal portions of both VI ARCHES	incomplete
III C1	Aberrant LIA	LDA patent	Regression of root of left ARCH and distal portion of right VI ARCH	complete
III C2	Aberrant LIA	RDA patent	Regression of root of left ARCH and distal portion of left VI ARCH	incomplete
III C3	Aberrant LIA	Bilateral DA patent	Regression of root of left ARCH	complete
III C4	Aberrant LIA	Bilateral DA regression	Regression of root of left ARCH and distal portions of both VI ARCHES	incomplete
III D1	Isolated LSA	LDA patent	Regression of left IV ARCH, distal portion of LDOA, and distal portion of right VI ARCH	No
III D3	Isolated LSA	Bilateral DA patent	Regression of left IV ARCH and distal portion of LDAO	No
III E1	Isolated LIA	LDA patent	Regression of root of left ARCH, distal portion of LDAO, and distal portion of right VI ARCH	No
III E3	Isolated LIA	Bilateral DA patent	Regression of root of left ARCH and distal portion of LDAO	No
III FB1	Isolated LCA with aberrant LSA	LDA patent	Regression of root of left ARCH, proximal portion of LDAO, and distal portion of right VI ARCH	incomplete
III FB3	Isolated LCA with aberrant LSA	Bilateral DA patent	Regression of root of left ARCH and proximal portion of LDAO	incomplete

Black color designates original 1964 Stewart classification; blue designates modifications (additional subgroups).

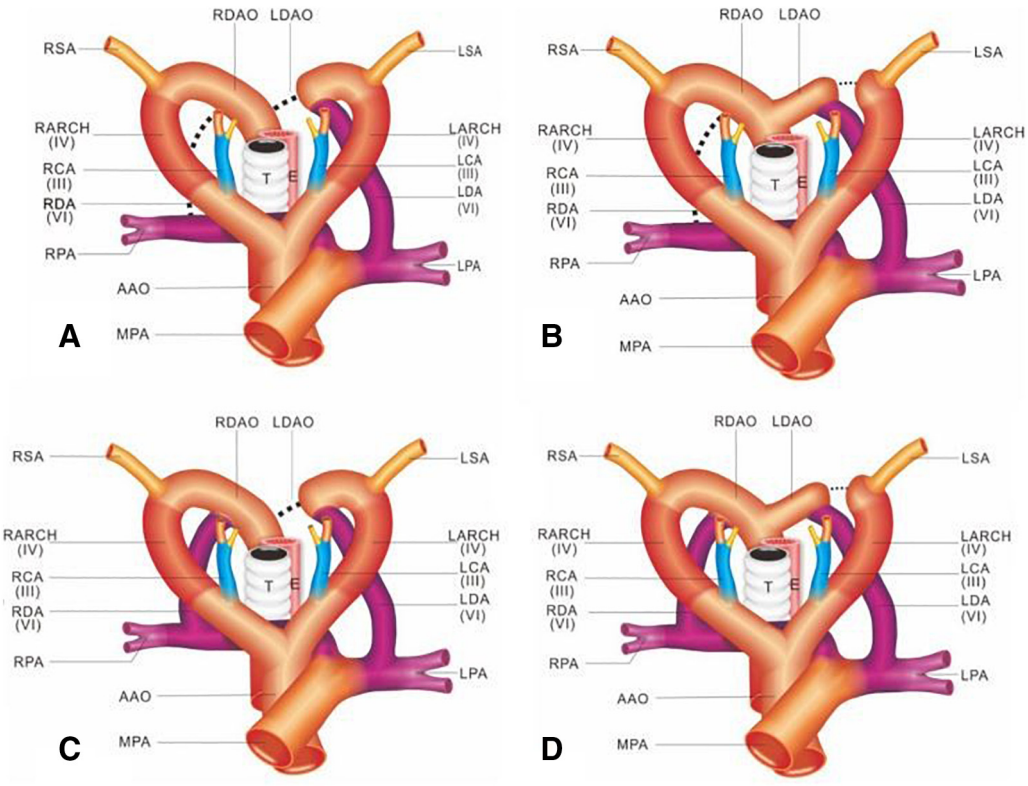

Fig. 5. GROUP III, Subgroup A: Malformations of the Right aortic arch. Congenital abnormalities shown in these figures include subgroups IIIA1a (**A**), IIIA1b (**B**), IIIA3a (**C**), and IIIA3b (**D**) (Table 4).

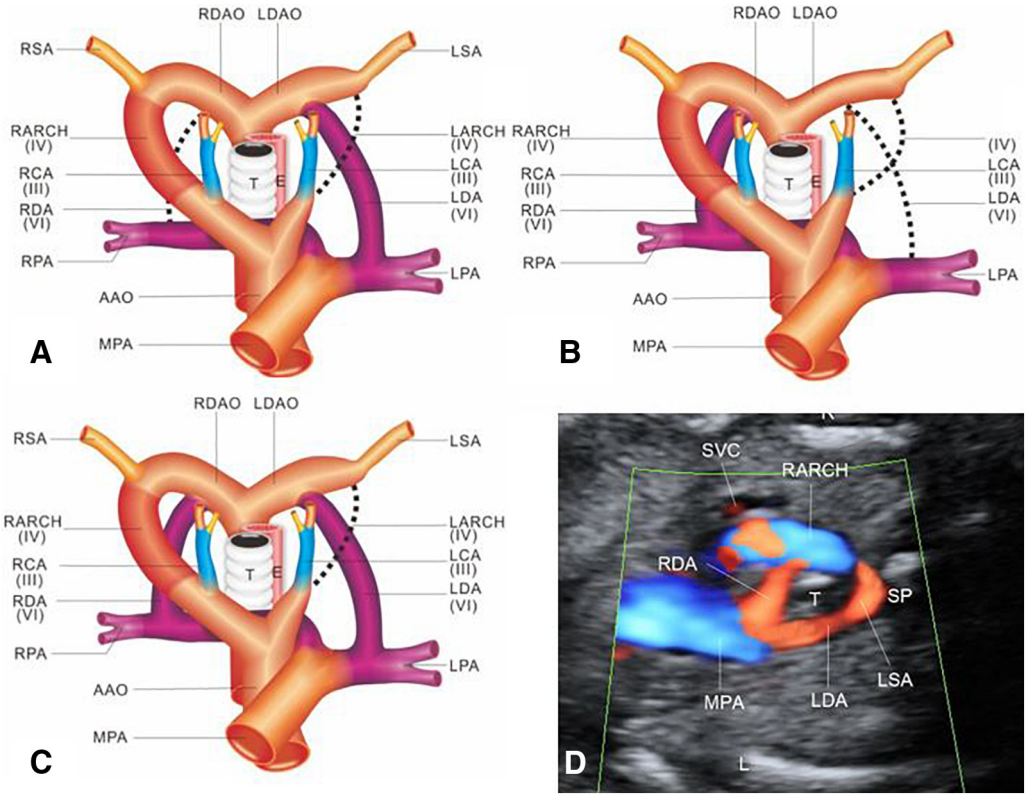

Fig. 6. GROUP III, Subgroup B: Right aortic arch with aberrant LSA. Congenital abnormalities shown in these figures include subgroups IIIB1 (**A**), IIIB2 (**B**), IIIB3 (**C**) and IIIB3 (**D**) (Table 4). RARCH with double DA and aberrant LSA was shown on 3VT view (**D**) in a fetus at 24 weeks gestation. Color Doppler showed RARCH, aberrant LSA, LDA and RDA encircle the trachea and esophagus forming a complete vascular ring.

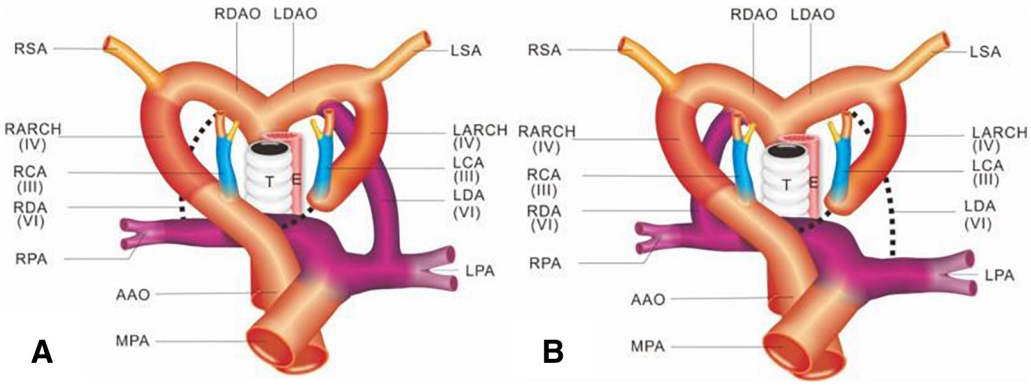

Fig. 7. GROUP III, Subgroup C: Right aortic arch with aberrant LIA. Congenital abnormalities shown in these figures include proposed subgroups IIIC1 (**A**) and IIIC2 (**B**) (Table 4).

both DA, with embryologic regression occurred at regions 5, 6 and 10. The aortic arch passes in front of the left bronchus and trachea. From proximal to distal, the arch gives rise to the LCA, LSA, and aberrant RIA. The RIA connects to the RDAO. The LARCH and RIA surround the esophagus and trachea, forming an incomplete vascular ring.

4.4. GROUP II, Subgroup D: Left aortic arch with isolated RSA

Isolated RSA is most commonly associated with bilateral DA (Subgroup IID3), and only rarely associated with regression of the LDA (Subgroup IID2). Type IID3 results from regression of the distal portion of the RDAO and the middle portion of the RARCH, with embryologic regression occurred at regions 7 and 9. The aortic arch passes in front of the left bronchus and trachea. From proximal to distal, the arch gives rise to the RCA, LCA, and LSA. The RSA connects to the RPA through the RDA. The LDA passes in front of the left bronchus and connects to the origin of the DAO; therefore, no vascular ring is formed. However, after birth, due to the contraction of the RDA, this lesion can lead to decreased blood supply to the right upper extremity of the affected child.

4.5. GROUP II, Subgroup E: Left aortic arch with isolated RIA

Isolated RIA is most commonly associated with bilateral patent DA (Proposed Subgroup IIE3), and only rarely associated with regression of the LDA (Proposed Subgroup IIE2). Type IID3 results from regression of the distal portion of the RDAO and the root of the RARCH, with embryologic regression occurred at regions 6 and 9. The aortic arch passes in front of the left bronchus and trachea. From proximal to distal, the arch gives rise to the LCA and LSA. The RIA connects to the RPA through the RDA; the LDA passes in front of the left bronchus and connects to the origin of the DAO. Therefore, no vascular ring is formed.

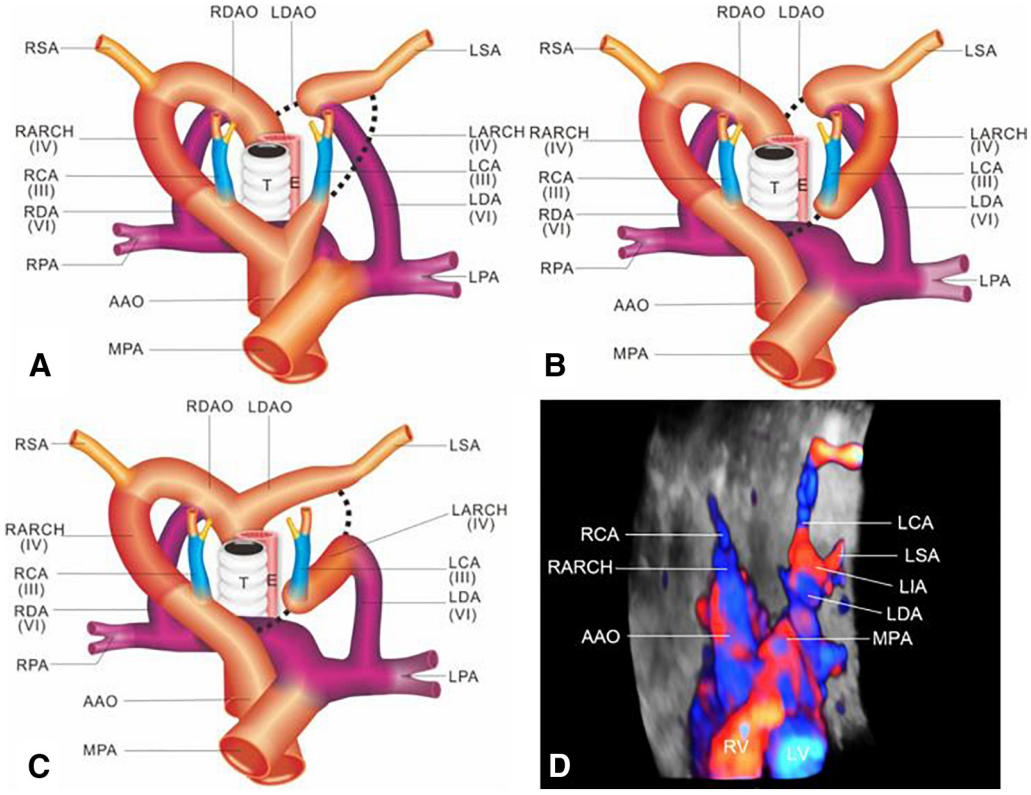

Fig. 8. GROUP III, Subgroup D, E, FB: Right aortic arch with isolated LSA or isolated LIA or isolated LCA and aberrant LSA. Congenital abnormalities shown in these figures include subgroups IIID3 (**A**), IIIE3 (**B**), IIIFB3 (**C**) and IIIE1 (**D**) (Table 4). RARCH with LDA and isolated LIA was shown on color Doppler 3D image with i-STIC HD flow mode (**D**) in a fetus at 26 weeks gestation. Isolated LIA originates from LDA and RARCH gives out RCA and RSA.

4.6. GROUP II, Subgroup F: Left aortic arch with isolated RCA (F) and aberrant RSA (B)

Isolated RCA with aberrant RSA occurs most commonly in association with bilateral patent DA (Proposed Subgroup IIFB3), and is only rarely seen in association with regression of the LDA (Proposed Subgroup IIFB2). Type IIFB3 results from regression of the root of the RARCH and the proximal portion of the RDAO, and from failure of cephalic migration of the RSA. In such cases, embryologic regression occurred at region 6 and 8. The aortic arch passes in front of the left bronchus and trachea. From proximal to distal, the arch gives rise to the LCA, LSA, and aberrant RSA. The RCA connects to the RPA through the RDA. The RSA connects to the distal portion of the RDAO. The LDA passes in front of the left bronchus and connects to the origin of the DAO. As such, the LARCH and RSA surround the esophagus and trachea, forming an incomplete vascular ring.

5. GROUP III: Congenital malformations of the right aortic arch (RARCH)

The classification of congenital abnormalities of the RARCH based on aberrant embryologic development of the heart is summarized in (Table 4). The formation of subgroups of RARCH congenital malformations (Group III) are similar to that seen with LARCH (Group II) in terms of the course of the individual vessel, and position and location of regression. The detailed subgroups are summarized in (Table 4) and representative schematic diagrams in (Figs. 5, 6, 7, and 8).

Right sided aortic arch is a rare aortic anomaly with a reported incidence of 0.1% [19]. As with congenital malformations of LARCH, malformations of RARCH do not result in a complete vascular ring as there is no persistent vessel on the left side of the trachea. However, RARCH malformations are commonly associated with other cardiovascular abnormalities [20]. Right aortic arch with mirror image branching is associated with CHD in up to 98% of cases, including tetralogy of Fallot, truncus arteriosus, tricuspid atresia, and transposition of the great arteries with pulmonary valve stenosis [21].

5.1. GROUP III, Subgroup A: Right aortic arch with mirror image branching

5.1.1. RARCH with patent LDA (Subgroup IIIA1,Fig. 5). It can be subdivided into: i) Subgroup IIIA1a with embryologic regression occurred at regions 4 and 10 and no vascular ring; ii) Subgroup IIIA1b with embryologic regression occurred at regions 3 and 10 and forming a complete vascular ring.

5.1.2. RARCH with patent RDA (Subgroup IIIA2). The embryologic regression occurred at regions 3, 4 and 5, and no vascular formation.

5.1.3. RARCH with patent DA bilaterally (Subgroup IIIA3,Fig. 5). It can be subdivided into: i) Subgroup IIIA3a with regression of the distal portion of the LDAO (region 4) and no vascular formation; ii) Subgroup IIIA3b with regression of the proximal portion of the LDAO (region3) and forming a complete vascular ring.

5.1.4. RARCH with regression of both DA (Subgroup IIIA4). This subgroup is secondary to regression of the LDAO and both DA occurred at regions 3, 4, 5 and 10, and no vascular ring formation.

5.2. GROUP III, Subgroup B: Right aortic arch with aberrant LSA

5.2.1. RARCH with aberrant LSA and LDA (Subgroup IIIB1, Fig. 6). This subgroup is due to regression of the middle portion of the LARCH and RDA occurred at regions 2 and 10, and as a result forming a complete vascular ring.

5.2.2. RARCH with aberrant LSA and RDA (Subgroup IIIB2, Fig. 6). This subgroup is due to regression of the middle portion of the LARCH and LDA occurred at regions 2 and 5, as a result forming an incomplete vascular ring.

5.2.3. RARCH with aberrant LSA and bilateral DA (Subgroup IIIB3,Fig. 6). This subgroup is due to regression of the middle portion of the LARCH occurred at region 2, and as a result forming a complete vascular ring.

5.2.4. RARCH with aberrant LSA and regression of both DA (Subgroup IIIB4). This subgroup is due to regression of the middle portion of the LARCH and both DA occurred at regions 2, 5 and 10, and as a result forming an incomplete vascular ring.

5.3. GROUP III, Subgroup C: Right aortic arch with aberrant LIA

5.3.1. RARCH with aberrant LIA and LDA (Subgroup IIIC1, Fig. 7). This subgroup is due to regression of the root of the LARCH and RDA occurred at regions 1 and 10, as a result forming a complete vascular ring.

5.3.2. RARCH with aberrant LIA and RDA (Subgroup IIIC2, Fig. 7). This subgroup is due to regression of the root of the LARCH and LDA occurred at regions 1 and 5, as a result forming an incomplete vascular ring.

5.3.3. RARCH with aberrant LIA and patent bilateral DA (Subgroup IIIC3). This subgroup is due to regression of the root of the LARCH occurred at region 1, as a result forming a complete vascular ring.

5.3.4. RARCH with aberrant LIA and regression of both DA (Subgroup IIIC4). This subgroup is due to regression of the root of the LARCH and both DA occurred at regions 1, 5 and 10, as a result forming an incomplete vascular ring.

5.4. GROUP III, Subgroup D: Right aortic arch with isolated LSA

Isolated LSA is most commonly associated with bilateral patent DA (Subgroup IIID3, Fig. 8), and is only rarely associated with regression of the RDA (Subgroup IIID1). Type IIID3 results from regression of the distal portion of the LDAO and the middle portion of the LARCH occurred at regions 2 and 4, and no vascular ring is formed. However, due to the contraction of the LDA after birth, blood supply to the left upper extremity may be reduced in affected child. Clinically, this anomaly can present with congenital subclavian steal syndrome and/or vertebrobasilar insufficiency [22]. RARCH with isolation of the LSA is the most common form, and is associated with CHD in more than 50% of cases, most commonly tetralogy of Fallot [23].

5.5. GROUP III, Subgroup E: Right aortic arch with isolated LIA

Isolated LIA is most commonly associated with bilateral patent DA (Subgroup IIIE3, Fig. 8), and is only rarely associated with regression of the RDA (Subgroup IIIE1). This subgroup has been associated also with VSD, cervical aortic arch, or double right ventricle [24]. Type IIIE3 results from regression of the distal portion of the LDAO and the root of the LARCH occurred at regions 1 and 4, and no vascular ring is formed.

5.6. GROUP III, subgroup F: Right aortic arch with isolated LCA (F) and aberrant LSA (B)

Isolated LCA with aberrant LSA is most commonly associated with bilateral patent DA (Subgroup IIIFB3, Fig. 8), and rarely associated with regression of the RDA (Subgroup IIIFB1). Subtype IIIFB3 is due to regression of the root of the LARCH and the proximal portion of the LDAO occurred at region 1 and 3, and failure of cephalic migration of the LSA, and forming an incomplete, and as a result forming an incomplete vascular ring.

6. GROUP IV: Other aortic arch abnormalities

6.1. Circumflex retro-esophageal aorta arch refers to the anatomic arrangement in which the distal portion of the aortic arch crosses the midline behind the trachea and esophagus and culminating in the opposite DAO. When LARCH persists, the branches are usually normal. When the RARCH persists, the branches usually reflect the mirror image; however, there can be an associated aberrant LSA or RSA. The DA can be on

either the left or the right side. When the DA is on the contralateral side of the aortic arch (i.e., located between the DAO and PA), a complete vascular ring is formed. This type of aortic arch abnormality is frequently associated with other congenital cardiac anomalies. Neither prenatal ultrasound examination nor postnatal imaging can accurately differentiate between circumflex retro-esophageal aortic arch and double aortic arch with one atretic arch.

6.2. Coarctation of the aorta refers to a congenital narrowing of the thoracic portion of the aorta, usually adjacent to the insertion of the DA (at the aortic arch isthmus). It is one of the more common congenital cardiovascular malformation, accounts for 5–7% of all cases of congenital heart diseases [25]. Different types of coarctation are described elsewhere [26]. Coarctation may occur as an isolated defect or in association with other CHD (most commonly bicuspid aortic valve and VSD). In rare instances, it may be associated with right-sided aortic arches, with a reported prevalence of 0.1% [27].

6.3. Interrupted aortic arch refers to a congenital vascular malformation in which there is discontinuity between the AAO and DAO. It is a rare congenital abnormality, accounting for only 1% of all congenital heart disease. There are three distinct subtypes depending on the location of the interruption in neonates: [28] (i) Type A, where the interruption occurs at the level of the isthmus between the LSA and LDA, accounts for about 43%; (ii) Type B, where the interruption occurs between the LCA and LSA. This is a more common type, accounting for 53%, and can be associated with an aberrant RSA; and (iii) Type C, where the interruption occurs between the LIA and LCA. This type is rare and accounts for only 4% of cases. IAA usually occurs in association with a large VSD and PDA, or less commonly with a large aortopulmonary window or truncus arteriosus [29].

6.4. Persistent fifth aortic arch is a rare abnormality resulting from persistence of embryonic aortic arch V. Persistent fifth aortic arch can be to the left or right, and multiple types have been described. The three most common types include: (i) Persistent fifth aortic left arch with normal LARCH; (ii) Persistent fifth aortic left arch with interrupted LARCH; (iii) Systemic-to- pulmonary connection resulting from an aberrant connection between the fifth and sixth aortic arches; the fifth arch may also connect with the MPA or LPA. Persistent fifth aortic arch is usually associated with intracardiac malformations, most commonly VSD [30].

6.5. Cervical aortic arch is a rare aortic arch abnormality in which the position of the aortic arch is shifted cephalad and extends above the clavicles, with the top of the arch extending into the supraclavicular fossa. Several theories have been proposed in an effort to explain the formation of a cervical aortic arch: (1) The arch may be derived from the second or third embryonic arch, instead of the fourth arch; (2) there may be failure of the normal caudal migration of the fourth arch; or (3) the third and fourth arches may fuse resulting in a failure of caudal migration. Cervical aortic arch is not usually seen in association with other structural heart lesions. Cervical aortic arch can be left-sided or right-sided. Right-sided cervical aortic arch is more common (80%), and 80% are associated with abnormal aortic branch anatomy. Haughton et al. [31] proposed five types of cervical aortic arch abnormalities based on the configuration of the aorta, the sequence of brachiocephalic branching, and embryogenesis. These include: (i) Type A, where the external and internal carotid artery branches arise separately from the aortic arch, the aortic arch is on the opposite side of the DOA, and there is an aberrant subclavian artery; (ii) Type B, where both common carotid arteries arise from the aortic arch, the DOA is on the opposite side of aortic arch, and there is an aberrant subclavian artery; (iii) Type C, characterized by a left-sided cervical arch with a right DAO and bicarotid trunk; (iv) Type D, which has an ipsilateral DAO with normal sequence of brachiocephalic branching; and (v) Type E, which has a right-sided cervical aortic arch with a right DAO and an aberrant left subclavian artery. Hirao and colleagues [32] reported that type D is the most common subtype and may be seen in association with aneurysm.

6.6. Left pulmonary artery sling (also known as an aberrant LSA) occurs when the LPA originates from the RPA instead of from the MPA. In these cases, the LPA courses around the right main bronchus and between the trachea and esophagus, forming a sling that compresses the distal trachea and right main bronchus. If the RPA is absent, the LPA may originate from the right lateral wall of the MPA.

6.7. Anomalous origin of pulmonary artery aorta and its branches refers to the situation in which one of the pulmonary arteries, either the right or left, arises directly from the aorta, commonly from the AAO. This is a rare abnormality and is often seen in association with other congenital cardiovascular diseases. Based on the distance from the origin of the PA to the aortic valve and IA, these lesions can be divided into proximal (close to the aortic value) and distal subtypes (close to the IA). If the RPA originates from the proximal portion of the AAO, it is usually associated with an aortic-pulmonary window. Several subtypes are described: (i) Anomalous origin of the LPA from the proximal portion of the AAO, which results from failed embryonic development of the left VI ARCH, and causes the left opisthobranchia pulmonary artery to connect to the proximal portion of the AAO; (ii) Anomalous origin of the LPA from the distal portion of the AAO, which results from failed embryonic development of the left VI ARCH, and causes persistence of the proximal portion of the left fifth aortic arch with the left opisthobranchia pulmonary artery connecting to the proximal portion of the left fifth arch; (iii) Anomalous origin of the LPA from the origin of the DAO, which is due to failed embryonic development of the proximal portion of the left VI ARCH, and causes the left opisthobranchia pulmonary artery to connect to the distal portion of the left VI ARCH; and (iv) Anomalous origin of the LPA from the LSA, which is due to failed embryonic development of the proximal portion of the LARCH, and causes the left opisthobranchia pulmonary artery to connect to the distal portion of the left VI ARCH, regression of the distal portion of the left dorsal aorta, persistent of the right dorsal aorta, and aberrant formation of the RARCH.

7. Current Conclusions

Congenital abnormalities of the aortic arch and its branches are complex, depending on the position, course, order, and pattern of the major as well as branching vessels. Recent advances in diagnostic imaging technology have led to better delineation of the vasculature anatomy, the identification of previously unrecognized and unclassified anomalies, and improved antenatal diagnosis. For all these reasons, we believe that the time is right to revisit and modify the original Stewart classification of congenital aortic arch abnormalities. Our proposed modifications are summarized in Table 1. These changes are important not only to facilitate more accurate antenatal counseling, but also to optimize postnatal care to include close monitoring of symptoms in the immediate postnatal period, early consultation with pediatric surgery, and early surgical intervention, if indicated. Moreover, congenital aortic arch abnormalities can exist in isolation or in associated with other intra- or extra-cardiac malformations and chromosomal abnormalities, such as chromosome 22q11 deletion syndrome (DiGeorge syndrome). Identification of an aortic arch abnormality should therefore prompt a more detailed evaluation to exclude other structural malformations (such as a fetal echo) and a referral for genetic counseling.

Conflicts of interest

None.

Acknowledgments

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