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Developmental Hypothesis

The left-sided aortic arch in humans, viewed as the end-result of natural selection during vertebrate evolution

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Abstract At some point during vertebrate evolution from species dwelling in water to living on land, the ancestral double or right aortic arches became single and left-sided in mammals, including humans, as the result of synchronous developments in cardiovascular and respiratory embryogenesis. Since left-sided aortic arches are unique to mammals, hemodynamics related to the placenta, specifically the requirement for a large arterial duct connecting to the descending aorta, may have led to switching from the right-sided to the left-sided arch. Additionally, development of a trilobar right lung and its bronchial tree, also unique to mammalian evolution, restricted the space above the high eparterial bronchus to a single large vessel. Consequently, mammals that mutated to the left-sided aortic arch avoided respiratory, digestive or circulatory problems that are often associated with an isolated right-sided aortic arch – something which could be considered a successful mistake. Due to natural selection, and survival of the fittest, the left-sided arch became the norm in mammals.

In congenital cardiac malformations where a large arterial duct is not mandatory in fetal life, as in Fallot's tetralogy or common arterial trunk, a right-sided aortic arch continues to occur, perhaps as an atavistic reversion to the anatomy seen in ancestral vertebrates.

Keywords: Aortic arches; evolution of vertebrate cardiovascular system; comparative anatomy of vertebrate aortic arches

In humans, as in most mammals, the aortic arch and the descending thoracic aorta are normally located in the left upper mediastinum (Fig. 1). When associated with cardiovascular malformations such as Fallot's tetralogy (Fig. 2), the aortic arch and the descending aorta are found in the right mediastinum, with an incidence as high as 50 per cent.[1] Since having the aortic arch on the 'wrong' side has no adverse consequences in certain types of cardiac anomalies, distinct anatomic, physiologic or genetic characteristics may exist that foster compatibility with the right-sided aortic arch. The incidence of a right-sided arch unassociated with cardiovascular anomalies is reported only as 0.1% in human adults.[2] Double aortic arches, with a right-sided descending aorta (Fig. 3), are rare. They are invariably associated with problems of respiration or swallowing. In contrast, double aortic arches are the norm in modern reptiles, which have no difficulties with swallowing, and right-sided aortic arches are the normal occurrence in modern birds. In 1962, Blake and Manion[3] compared the diverse anatomy of human thoracic arterial arches with those in other mammals, lower vertebrates, and birds and found that a single left-sided aortic arch is unique to mammals. In this study we discuss the rationale for laterality of the aortic arch in the context of cardiovascular and respiratory evolution from primitive vertebrates to modern mammals.

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Occurrence of a right-sided aortic arch in mammals

Both double and right-sided aortic arches in humans and other mammals can be regarded a reversion to the morphology of our vertebrate ancestors (Fig. 4).[4] During evolution, such double or right-sided arches may have become incompatible with asymptomatic survival due to a host of anatomic and physiologic developments unique to placental mammals. These include:

- The placental circulation, which requires the presence of a large systemic arterial duct for the return of systemic venous drainage to the placenta.
- Left-sidedness of the heart, which limits broncho-pulmonary expansion to the right hemithorax.
- Shortening of paired dorsal aortic roots which encircle the trachea and esophagus, producing problems with swallowing and respiration.
- Development of alveolar lungs with bi-directional flow of air; the addition of the right upper lobe and eparterial bronchus; the shortening of the trachea and bronchuses; and the separation of pleural cavities from the coelom by the muscular diaphragm.
- Presence of a large fetal thymus in the anterior mediastinum[5]

As a result of adaptation and natural selection, mutants with left-sided aortic arches prevailed as the dominant mammalian form, something which could be viewed as a successful mistake. The sporadic recurrence of double or right-sided aortic arches in modern mammals implies that the predisposition to the ancestral pattern has not been entirely suppressed. Right-sided aortic arches, without producing symptoms, often resurface in fetuses with cardiovascular malformations whose viability does not depend on the function of a systemic arterial duct.

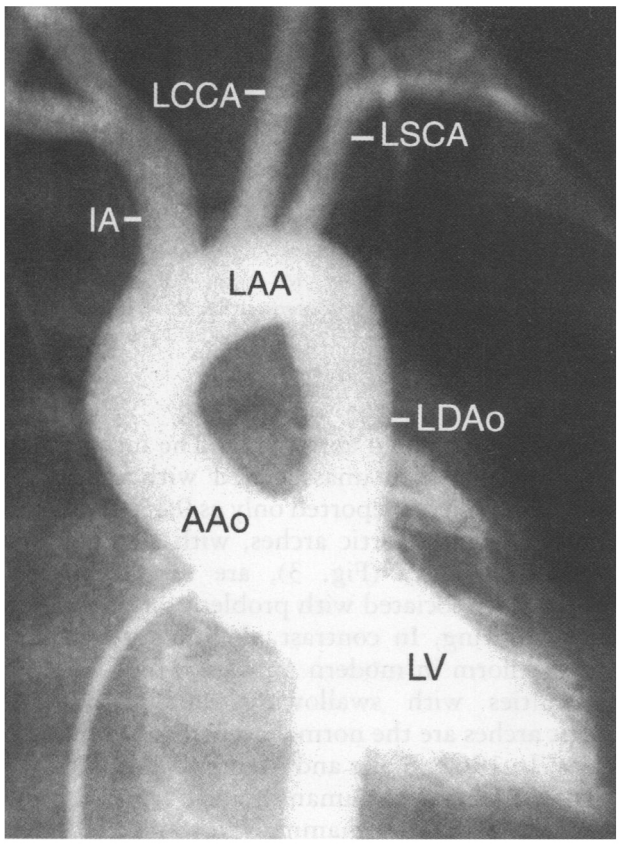

Figure 1.
Angiogram of a normal left-sided aortic arch. Legend: AAo = ascending aorta; IA = innominate artery; LAA = left-sided aortic arch; LCCA = left common carotid artery; LDAo = left-sided descending aorta; LSCA = left subclavian artery; LV = left ventricle.

Anatomic considerations

The right-sided aortic arch is not an exact mirror image of the normal left-sided aortic arch (Fig. 5A).[6] The right-sided arch extends more cephalad to straddle the high right eparterial bronchus. For added length, the proximal segment of the embryonic right anterior arch, located between the

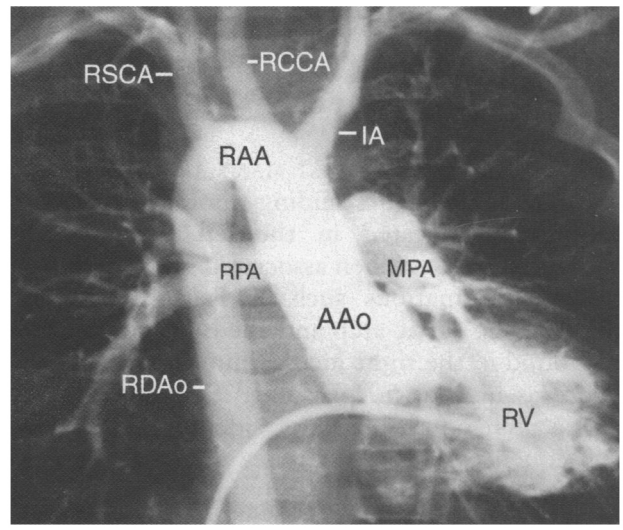

Figure 2.
Angiogram of right-sided aortic arch in Fallot's tetralogy. Legend: A Ao = ascending aorta; IA = innominate artery; MPA = pulmonary trunk; RAA = right-sided aortic arch; RCCA = right common carotid artery; RDAo = right-sided descending aorta; RPA = right pulmonary artery; RSCA = right subclavian artery; RV = right ventricle.

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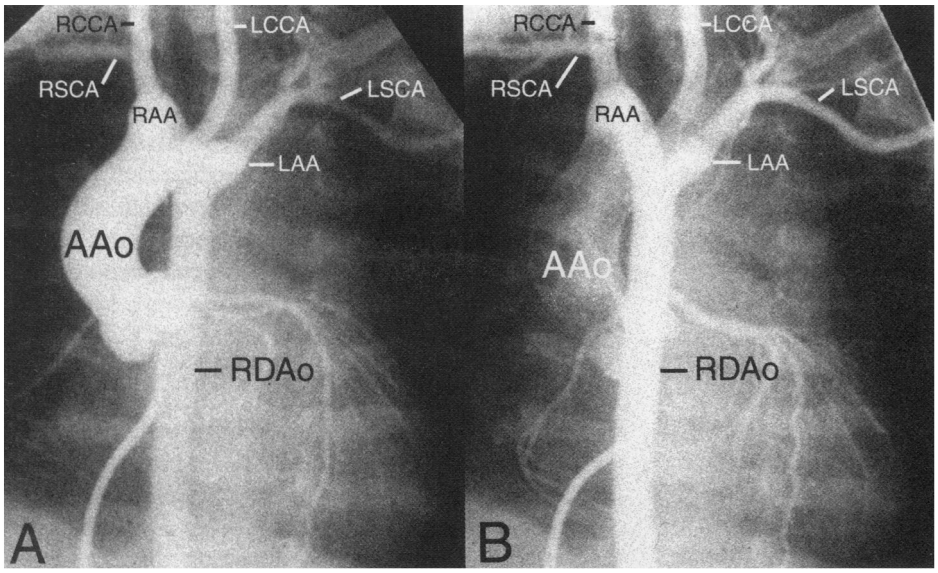

Figure 3.
Angiogram of anterior (A) and posterior (B) aspects of double aortic arch with right-sided descending aorta. In the usual arrangement, the right-sided arch is always more craniad than the left-sided arch. Legend: AAo = ascending aorta; LAA = left-sided aortic arch; LCCA = left common carotid artery; LSCA = left subclavian artery; RAA = right-sided aortic arch; RCCA = right common carotid artery; RDAo = right-sided descending aorta; RSCA = right subclavian artery.

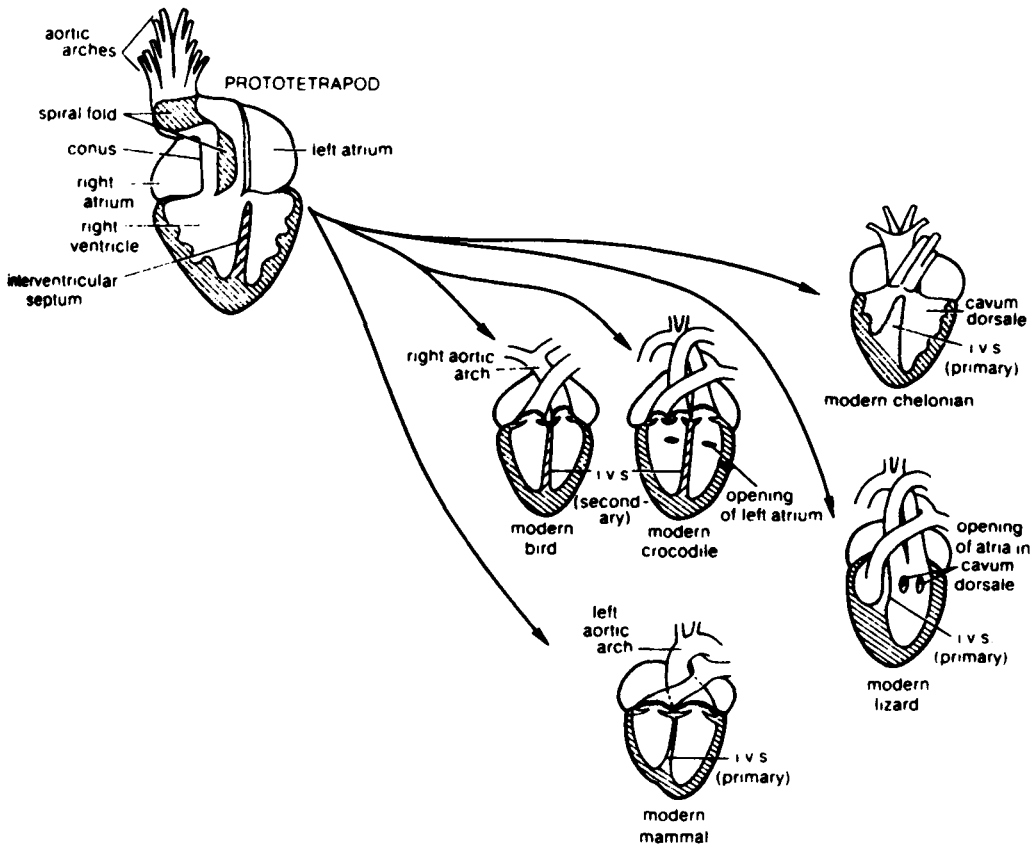

Figure 4.
Evolution of the great arteries in vertebrates from the common ancestor (Prototetrapod). With the exception of mammals, which possess a left-sided aortic arch, modern vertebrates have either double aortic arches, as in reptiles, or a right-sided aortic arch, as in birds. (Modified from Wake, ed., Hyman's Comparative Vertebrate Anatomy, 3rd ed., Fig. 11.30, p 499, with permission). Legend: i.v.s. = interventricular septum.

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left brachiocephalic and right common carotid arteries, does not involute (Fig. 5B). The large aortic arch constrained in the tight right paratracheal space produces the characteristic indentation in the lower trachea, a feature not found with the left-sided aortic arch. The only vessel normally found in this space is the azygos vein (Fig. 6A). In some mammals with right-sided aortic arches (Fig. 6B), the azygos drainage is switched to the left hemiazygos vein.[3,7]

Evolution of the mammalian respiratory system

Respiratory evolution from fish-like vertebrates to land-dwelling mammals began with addition of lungs, evolved from swim bladders, to gills,[8] both these organs being found together in modern lungfish and larval salamanders. Next, the gills were lost, as in adult salamanders, but the lungs remained primitive, and the skin had to be utilized for additional blood-gas exchange, as seen in the pulmo-cutaneous arch of modern frogs. In birds, which evolved in parallel with reptiles, small flow-through lungs developed without alveoluses that required large air sacks for ventilation. In land-dwelling mammals, the lungs grew in size and became alveolar, with bidirectional flow of air requiring a powerful diaphragm for efficient function.

Evolution of the vertebrate aortic arches

Evolution[9] (Fig. 7) began with the ancestral symmetrically paired double arch prototype (Fig. 7a). Such symmetry is preserved in modern fish (Fig. 7b) and amphibians (Fig. 7c,d). Double arches became asymmetric with dominance of the right side in reptiles (Fig. 7e,f), solitary and right-sided in birds (Fig. 7g), and solitary and left-sided in

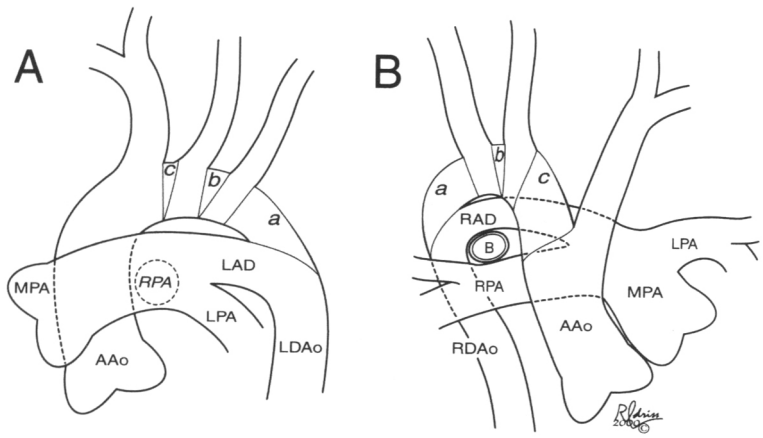

Figure 5.
Diagram of definitive aortic arch segments. A. In left-sided arch, the proximal embryonic arch segment (c) involutes. B. In right-sided arch, segment 'c' persists and enables the ascending aorta to span over the right eparterial bronchus (B). An associated right-sided arterial duct requires the arch to extend cervically. Arch segments: a = isthmic; b = distal; c = proximal. Legend: AAo = ascending aorta; B = eparterial bronchus; LAD = left-sided arterial duct; LDAo = left-sided descending aorta; LPA = left pulmonary artery; MPA = pulmonary trunk; RAD = right-sided arterial duct; RDAo = right-sided descending aorta; RPA = right pulmonary artery. (Modified from Moullaert[6]).

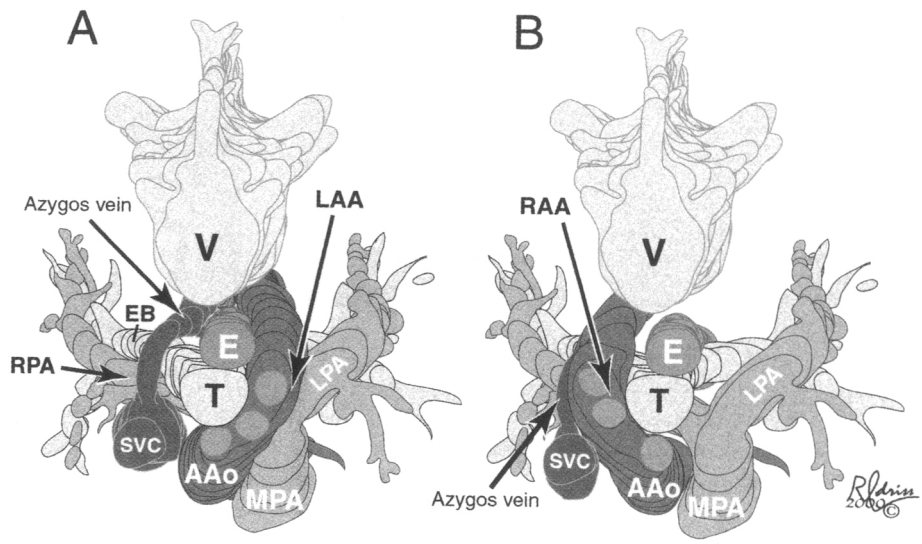

Figure 6.
Crossections of mediastinal structures (reconstructed from CT scan). A. With left-sided aortic arch, only the azygos vein passes over the eparterial bronchus. B. With right-sided aortic arch and right-sided descending aorta, the arterial duct and azygos vein tend to switch to the left mediastinum. Legend: AAo = ascending aorta; E = esophagus; EB = eparterial bronchus; LAA = left-sided aortic arch; LPA = left pulmonary artery; MPA = pulmonary trunk; RAA = right-sided aortic arch; RPA = right pulmonary artery; SVC = superior caval vein; T = trachea; V = vertebral column.

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mammals (Fig. 7h). In reptiles, who have two ascending aortas, two aortic arches, and two dorsal aortic roots (Fig. 7e,f), the dominant right aorta and arch supply the brain, forward limbs and the thorax, whereas the left aorta and its arch do not branch until they reach the abdomen and the hind limbs.[10] As a possible atavistic reversion to such ancestral pattern, an infant was born with pulmonary atresia, a large ventricular septal defect, a left-sided aortic arch, and persistent dorsal aortic roots bilaterally (Fig. 8). The larger left-sided root forms the descending aorta, which is a non-branching conduit until it passes through the diaphragm. The smaller right-sided root, reminiscent of the ancestral dominant aorta, gives rise to all the intercostal arteries and an aberrant right subclavian artery.

The human utero-placental circulation is established by the end of the 3rd week.[11] During the 5th week (Fig. 9), as bilateral bronchial buds begin to develop,[12] the aortic arches are symmetric. By the 6th week the aortic arches become asymmetric as three bronchuses sprout on the right, and two on the left. To accommodate the high right eparterial bronchus, the 4th right-sided aortic arch stretches craniad[13] over the eparterial bronchus, and the right-sided arterial duct disappears. By the 7th week, there is involution of the elongated right-sided dorsal aorta, the right-sided anterior 4th arch becomes the brachiocephalic artery and the definitive pattern of a left-sided arch with a left-sided systemic arterial duct is established. Anatomically and physiologically, a systemic arterial duct acts as direct extension of the pulmonary trunk configured to merge with the left-sided descending aorta as a continuous channel, the ductal arch (Fig. 10).

Incidence of right-sided aortic arch

The incidence of a right-sided aortic arch as an isolated malformation in children and adults as encountered in two series of necropsies on 2000 normal cadavers[14,15] was 0.1%. Analysis of 20.000

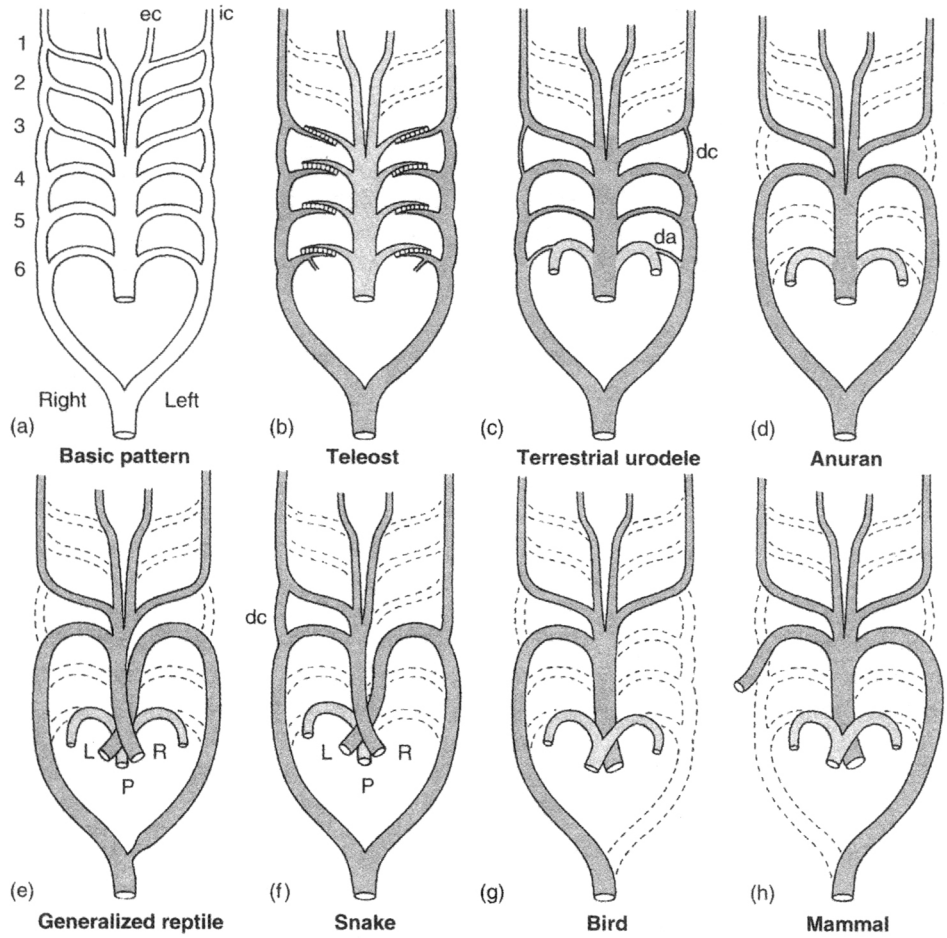

Figure 7.
Evolution of vertebrate aortic arches began with ancestral double symmetric arches (a) preserved in modern fish (b), adult salamanders (c) and frogs (d). The double arches are asymmetric in reptiles (e, f) with dominant right arch, single and right in birds (g) and single and left in mammals (h). (Original source: J. S. Kingsley, Outline of Comparative Anatomy of Vertebrates, The Blakiston Company, Philadelphia 1920).

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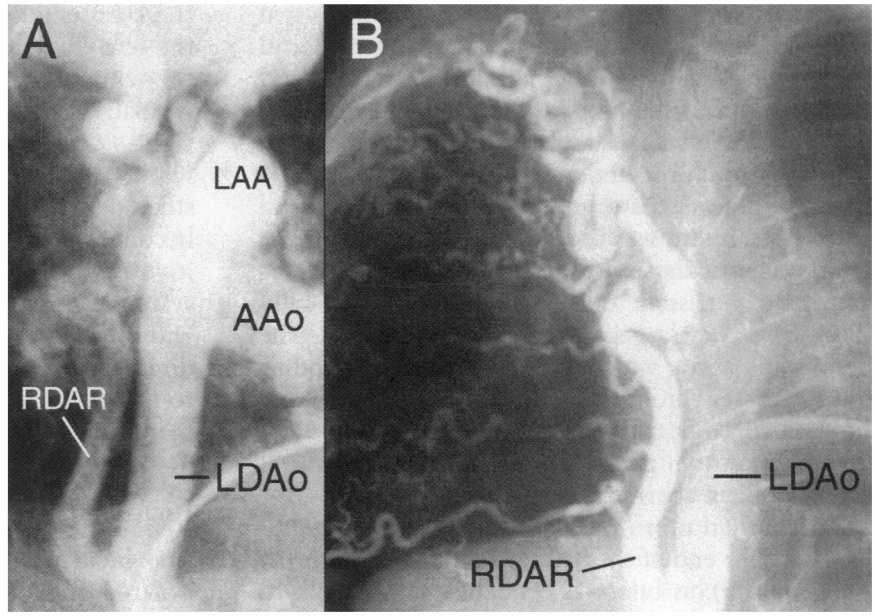

Figure 8.
A. Angiogram from an infant with pulmonary atresia, large ventricular septal defect, left aortic arch and left descending aorta. B. A right-sided dorsal aortic root, the remnant of the ancestral right-sided dominant aorta emerging from the descending aorta, gives rise to all intercostal arteries and right subclavian artery. Legend: AAo = ascending aorta; LAA = left-sided aortic arch; LDAo = left-sided descending aorta; RDAR = right-sided dorsal aortic root.

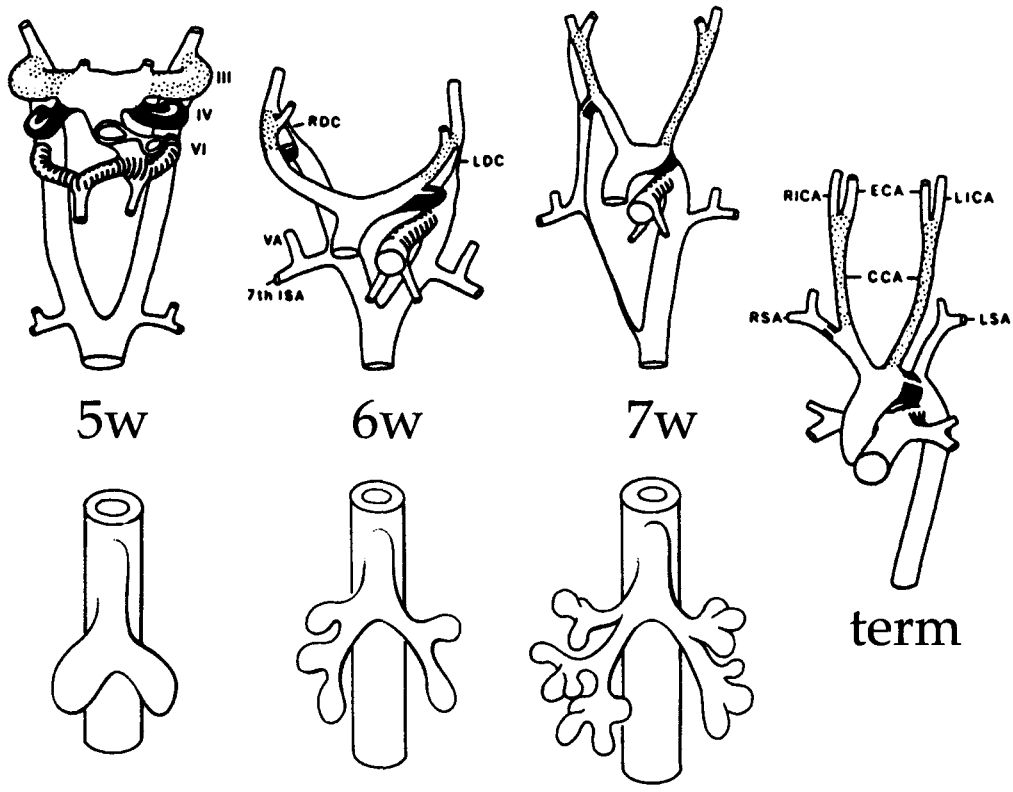

Figure 9.
Synchronous development of aortic arches (top panel) and bronchuses (bottom panel) in the human embryo. In the 5th week, a single right and left bronchial bud emerges and the aortic arches are symmetric. In the 6th week, an eparterial bronchus develops on the right, the right-sided 4th aortic arch extends craniad over the bronchus and the right-sided arterial duct disappears. During the 7th week, the definitive left-sided aortic arch is nearly complete. (Top panel: Modified from Am J Cardiol: 54: 832, Van Mierop and Kutsche, Interruption of the aortic arch and coarctation of the aorta, 1984, with permission from Excerpta Medica Inc.). (Bottom panel: Modified from W. J. Larsen, Essentials of Human Embryology 1998, p 82, with permission from Harcourt-International). Legend: III, IV, VI = aortic arches; CCA = common carotid artery; ECA = external carotid artery; ISA = intersegmental artery; LDC = left carotid duct; LICA = left internal carotid artery; LSA = left subclavian artery; RDC = right carotid duct; RICA = right internal carotid artery; RSA = right subclavian artery; VA = vertebral artery; w = weeks.

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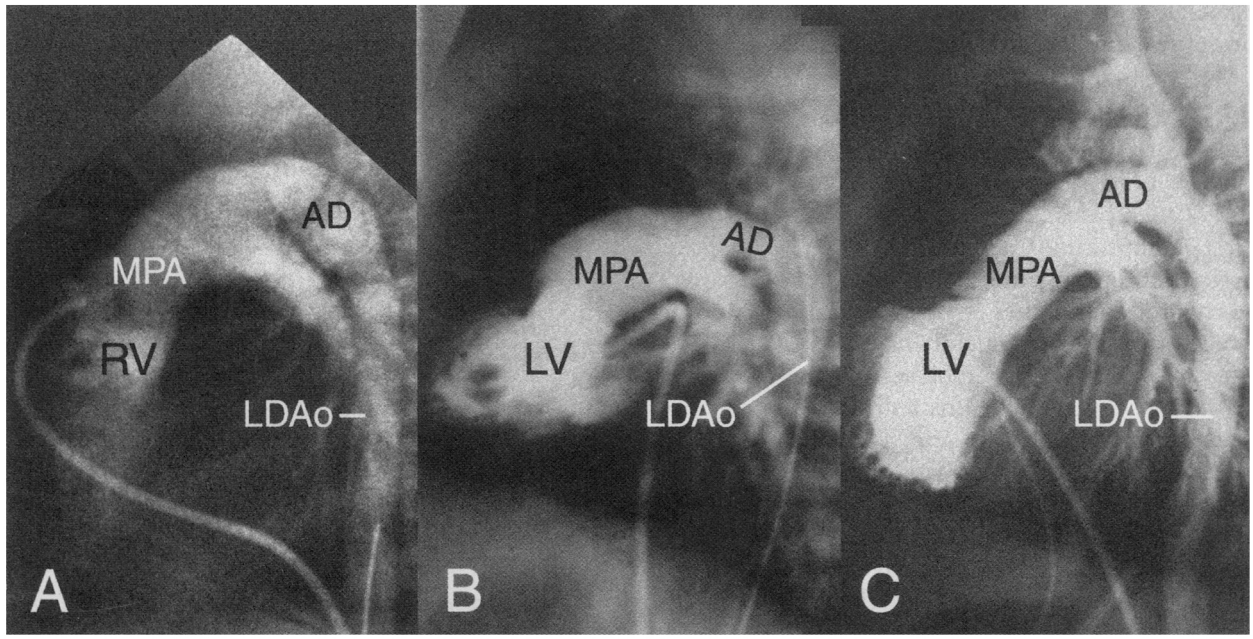

Figure 10.
Angiograms of neonatal ductal arches with flow from the pulmonary trunk through the arterial duct to the descending aorta. A. Normally related great arteries {S.D.S} B. Transposition {S.D.D} and C. Transposition {S.L.L}. Legend: AD = arterial duct: LDAo = left-sided descending aorta: LV = left ventricle: MPA = pulmonary trunk: RV = right ventricle.

chest radiographs for the characteristic right tracheal indentation from persons without apparent cardiac abnormalities[16] also revealed an incidence of 0.1%. These reports confirm the extreme rarity of the right-sided aortic arch as an isolated finding in humans. Perhaps some embryos destined to develop with an isolated right-sided aortic arch and right-sided descending aorta are unable to form a functional arterial duct and undergo spontaneous abortion, a form of natural selection. Cardiovascular malformations in spontaneously aborted human embryos have been reported to occur in up to two-fifths of cases prior to ten weeks gestation,[17] but the incidence of isolated right aortic arch has not been specifically addressed in these reports.

In complete mirror-imaged arrangement, the eparterial bronchus is left-sided. The aortic arch has been reported to be right-sided in four-fifths of such cases.[18]

In left isomerism, with bilateral hyparterial bronchuses, the side of the aortic arch has yet to be systematically evaluated.

Co-morbidity with isolated right-sided aortic arch

Two adults with a right-sided aortic arch, arterial duct and descending aorta were reported by Swan and his collegues in 1963.[19] Both had absence of the left pulmonary artery, one had a ventricular septal defect, neither had respiratory obstruction, and both underwent successful division of the patent arterial duct. Our survey of literature failed to find a report on a human neonate born with right-sided aortic arch, right-sided systemic arterial duct, right-sided descending aorta and normal cardio-pulmonary anatomy.

Dodge-Khatami et al.[20] reported on an infant with right-sided aortic arch and right-sided descending aorta, bilateral ductal ligaments, atretic left pulmonary artery and no intracardiac defects, who presented with severe respiratory symptoms. At angiography at 5 months, the right-sided arterial duct was found to be closed (Fig. 11A). There was a remnant of a left-sided duct. The right pulmonary artery had a tri-lobar distribution (Fig. 11B). Simultaneous bronchography showed a severely obstructed right main bronchus, with no visualization of an eparterial branch (Fig. 11C). At surgery, the right ductal ligament was divided with relief of the bronchial obstruction. Figure 11D depicts the anatomy in the right mediastinum during fetal life.

These case reports suggest that there is a cause and effect relationship between atresia or absence of the left pulmonary artery, right-sided aortic arch and right-sided systemic arterial duct, an association that can lead to prenatal origin of a right-sided bronchial obstruction.

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Right-sided aortic arch in association with congenital cardiac malformations

In a typical common arterial trunk, when flow of blood to the lungs is usually increased, a systemic arterial duct is not required in fetal life. The aortic arch has been reported to be right-sided in one-third of such cases.[1]

In malformations associated with a small or atretic pulmonary trunk, such as Fallot's tetralogy and related anomalies with reduced pulmonary blood flow, a systemic ductal arch cannot form. The systemic venous return reaches the placenta via intracardiac communications and via the ascending aorta. The point of origin and the configuration of arterial duct is altered to facilitate flow of aortic blood into the pulmonary arteries, the duct serving a pulmonary arterial function (Fig. 12, top). Although such a vessel of small caliber may fit into the space next to the right-sided aortic arch, it nevertheless emerges preferentially from the left subclavian artery (Fig. 12, bottom). When a pulmonary duct arises directly from a right-sided aortic arch, it is to supply discontinuous pulmonary arteries, or to supplement an inadequate left-sided arterial duct.

When the right-sided aortic arch is interrupted (Fig. 13), a right-sided ductal arch takes its place above the eparterial bronchus. Such a solitary ductal arch, however, tends to be short and, unlike a solitary aortic arch, it can cause bronchial compression.[21]

An isthmic aortic arch of reduced caliber associated with the rare right-sided tubular hypoplasia can coexist with a right-sided ductal arch (Fig. 14) during fetal and early neonatal life.

Aternatives to the right-sided arterial duct in association with a right-sided aortic arch

Human embryos destined to develop with an isolated right-sided aortic arch enhance their viability by activating alternative pathways in the early aortic arch template:[22]

- A right cervical aortic arch develops from the 3rd embryonic arch and extends to the level of the clavicle and beyond. In its isolated form (Fig. 15A), its craniad position allows for a large fetal arterial duct to connect to the right descending aorta without obstructing the eparterial bronchus. Nevertheless, most right cervical arches preferentially take a leftward retroesophageal course.[23]

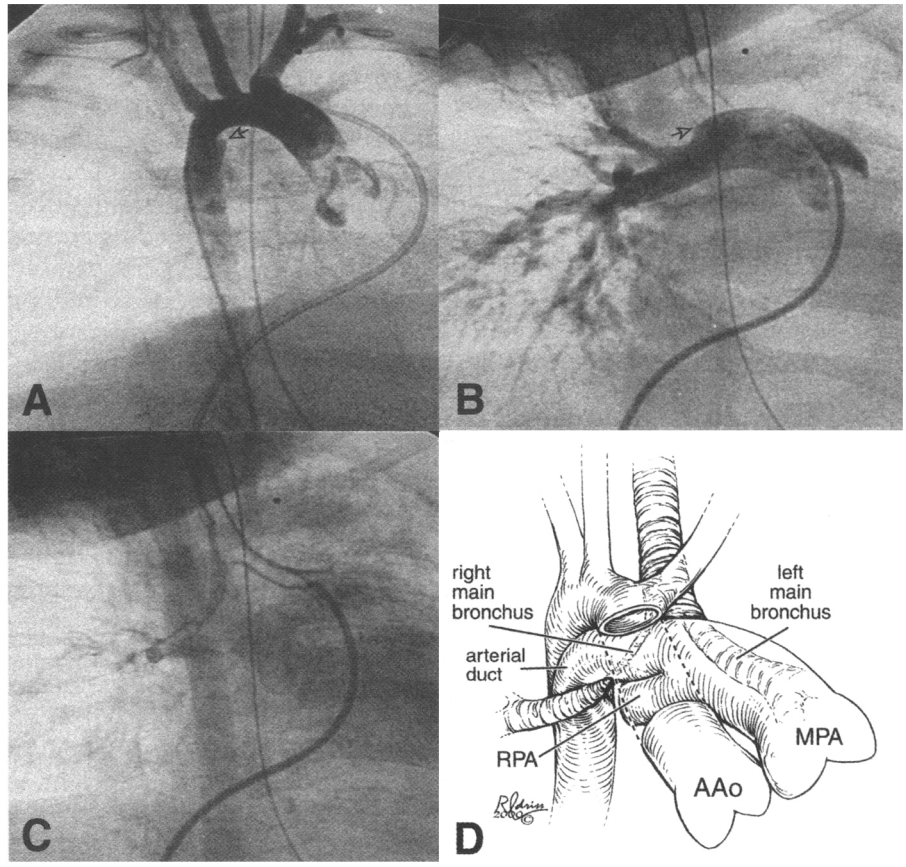

Figure 11.
A 5-month-old infant with severe respiratory symptoms. A. Aortogram showing right-sided aortic arch with remnant of right-sided systemic arterial duct (arrow). B. Pulmonary arteriogram showing absence of the left branch, trilobar right branch, and a ductal ampulla (arrow). C. Simultaneous bronchogram showing tubular compression of the right main bronchus with non-visualization of the eparterial bronchus. D. Artist's depiction of fetal anatomy with the large systemic arterial duct compressing the bronchus. (Ascending aorta and right pulmonary artery shown transsected). Legend: AAo = ascending aorta; MPA = pulmonary trunk; RPA = right pulmonary artery. (Figures A, B, C reprinted with permission from the Society of Thoracic Surgeons, Ann Thorac Surg 1999; 67: 1473).[20]

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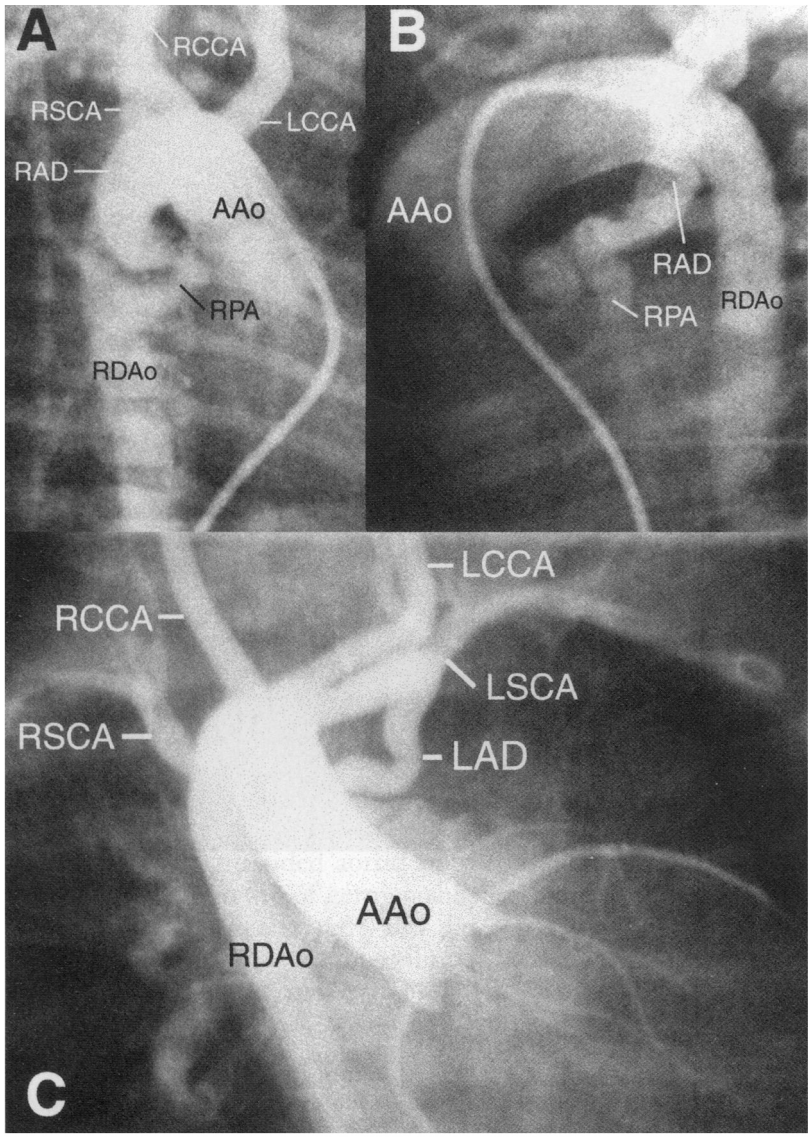

Figure 12.
Angiograms of right-sided aortic arches with flow through the arterial duct from the aorta to the pulmonary arteries. Top: Frontal (A) and lateral (B) view of the duct originating from the aortic arch (uncommon). C. Origin of pulmonary duct from the left subclavian artery (common). Legend: AAo = ascending aorta; LAD = left-sided arterial duct; LCCA = left commmon carotid artery; LSCA = left subclavian artery; RAD = right-sided arterial duct; RCCA = right common carotid artery; RDAo = right-sided descending aorta; RPA = right pulmonary artery; RSCA = right subclavian artery.

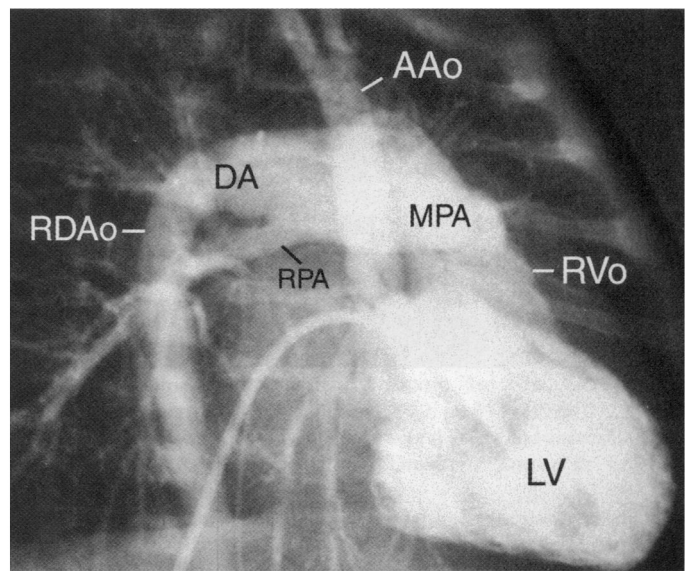

Figure 13.
Interrupted right-sided aortic arch. The right-sided arterial duct replaces the aortic arch. Legend: AAo = ascending aorta; DA = ductal arch; LV = left ventricle; MPA = pulmonary trunk; RDAo = right-sided descending aorta; RPA = right pulmonary artery; RVo = right ventricular outflow tract.

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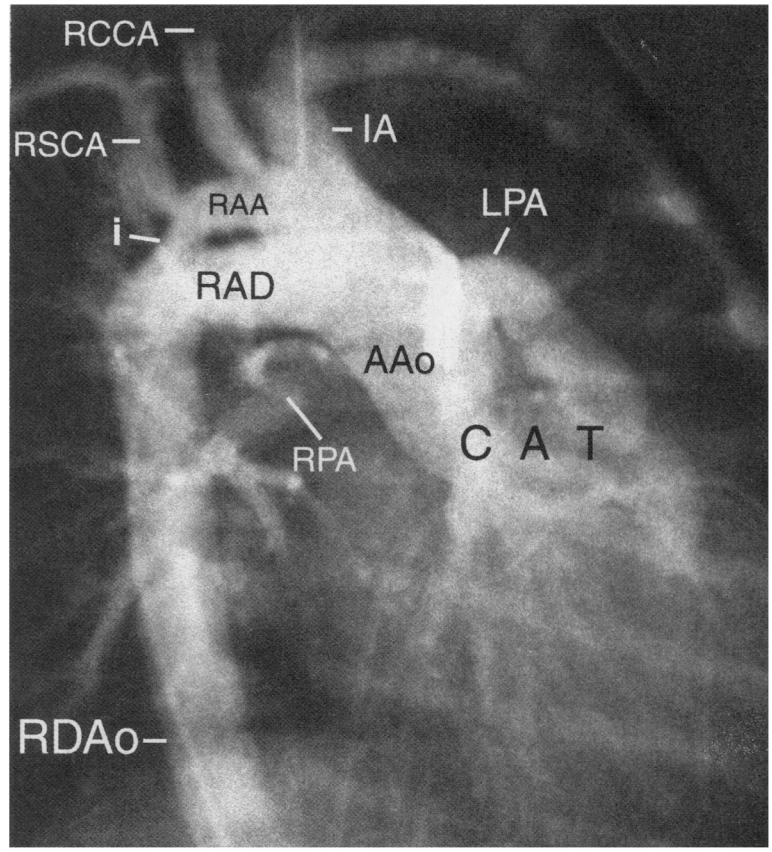

Figure 14.
Hypoplasia of the right-sided aortic arch in association with common arterial trunk. Legend: AAo = ascending aorta; CAT = common arterial trunk; i = isthmic arch; IA = innominate artery; LPA = left pulmonary artery; RAA = right-sided aortic arch; RAD = right-sided arterial duct; RCCA = right common carotid artery; RDAo = right-sided descending aorta; RPA = right pulmonary artery; RSCA = right subclavian artery.

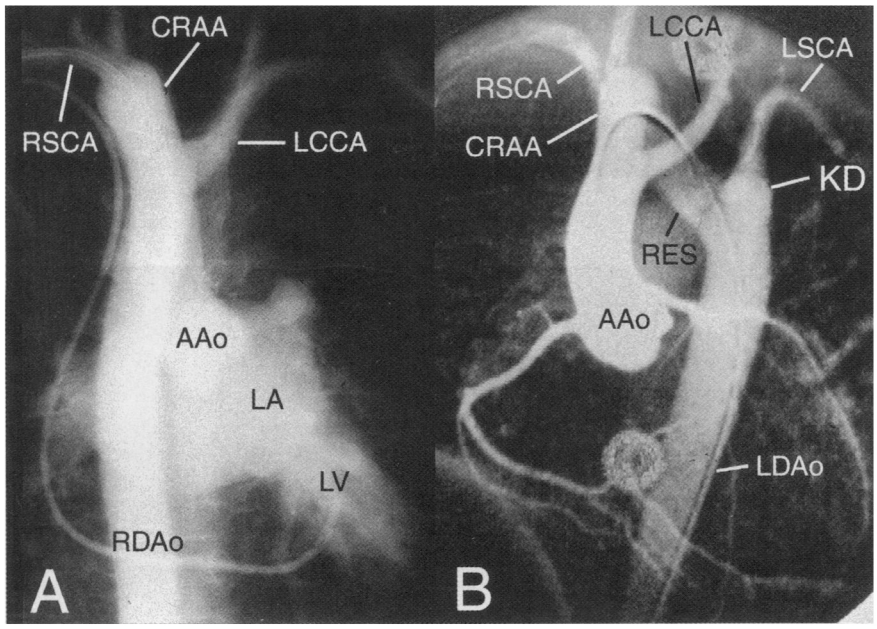

Figure 15.
Angiograms of two variants of isolated right-sided aortic arch. A. Cervical right-sided arch with right-sided descending aorta (rare). B. Circumflex right-sided arch with left-sided descending aorta (common). The arch is cervical and retroesophageal. There is a left diverticulum of Kommerell with a small arterial duct. Legend: AAo = ascending aorta; CRAA = cervical right-sided aortic arch; KD = Kommerell's diverticulum; LA = left atrium; LCCA = left common carotid artery; LDAo = left-sided descending aorta; LSCA = left subclavian artery; LV = left ventricle; RDAo = right-sided descending aorta; RES = retroesophageal segment; RSCA = right subclavian artery.

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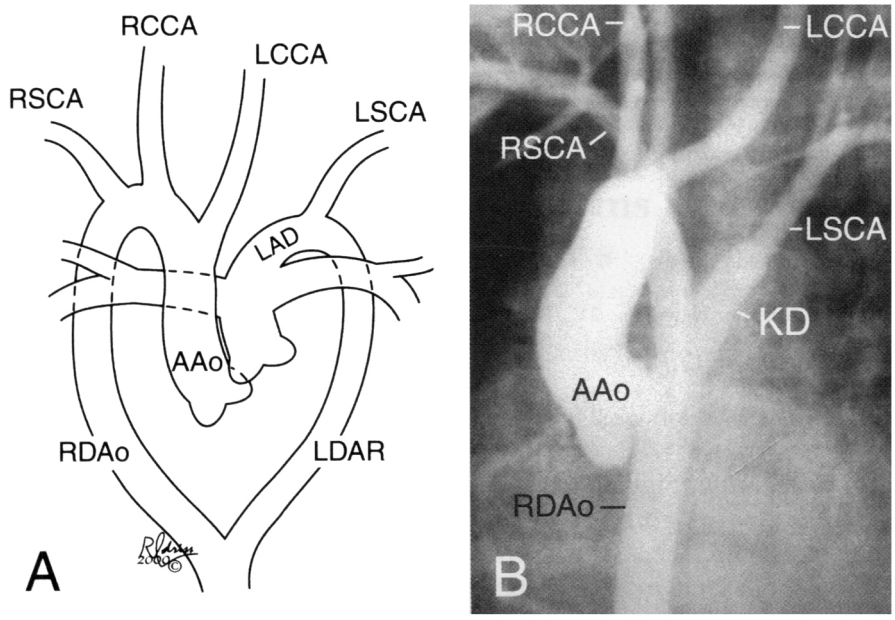

Figure 16.
A variant of isolated right-sided aortic arch with right-sided descending aorta and persistent left-sided dorsal aortic root. A. Drawing of prenatal anatomy with persistent left dorsal aortic root, left systemic arterial duct and aberrant left subclavian artery. B. Angiogram showing the postnatal anatomy with left dorsal aortic root converted to Kommerell's diverticulum and aberrant left subclavian artery. Legend: AAo = ascending aorta; KD = Kommerell's diverticulum; LAD = left-sided arterial duct; LCCA = left common carotid artery; LDAo = left-sided dorsal aorta; LDAR = left-sided dorsal aortic root; LSCA = left subclavian artery; RCCA = right common carotid artery; RSCA = right subclavian artery.

- The circumflex right aortic arch (Fig. 15B) takes a retroesophageal right-to-left course to join the left-sided descending aorta and arterial duct. One report[24] found that four-fifths of isolated right-sided aortic arches take this route. Dogs born with a right-sided aortic arch, accounting for almost one-tenth of german shepherds, have the retroesophageal type[3,7] and develop severe swallowing problems at weaning. These dogs often have hemiazygos drainage into the left superior caval vein.
- Persistence of the left-sided dorsal aortic root can serve as a conduit between the right-sided descending aorta and the left-sided arterial duct in prenatal life (Fig. 16A). Distally, it forms an aberrant left subclavian artery, while proximally, after ductal closure, it may form a diverticulum of Kommerell[25] (Fig. 16B).

Discussion

In the early stages of the normal human embryogenesis, the aortic arches are part of a symmetric system that gradually undergoes involution, persisting in the fetus as a single left-sided aortic arch, arterial duct and descending aorta. Embryogenesis of aortic arches is a period in human ontogeny that reflects the entire vertebrate evolutionary history. There is considerable evidence supporting the hypothesis that the uniqueness of placental circulation, which is dependent on a large caliber systemic arterial duct, and the right-sided tri-lobar lung with an eparterial bronchus, may have led to the change from right-sided to left-sided aortic arch. The crowding of the right upper mediastinum may have rendered untenable coexistence of a right-sided aortic arch, right-sided descending aorta and right-sided arterial duct. Mammals that mutated to a left-sided aortic arch proved more viable and, through natural selection, the left-sided aortic arch became the norm for the species. In situations where the right eparterial bronchus is absent, such as mirror imaging or bilateral leftsidedness, or where there is no necessity for a prenatal systemic arterial duct, as seen in a variety of cardiac malformations, the right-sided aortic arch remains a viable alternative. It is plausible that even in modern mammals, including humans, the ancestral tendency for formation of a right-sided aortic arch has not been entirely suppressed.

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