Cardiol Young 2003; 13: 537–543

ISSN 1047-9511

Original Article

The implications of common brachiocephalic trunk on associated congenital cardiovascular defects and their management

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Abstract A common brachiocephalic trunk is an anatomic variant in which both common carotid arteries and the right subclavian artery arise from the aortic arch via a single trunk. The impact of this condition on associated congenital cardiac malformations is presently unknown. Out of a total of 1480 cardiac catheterizations performed in children over a period of 10 years, we discovered 48 patients (3.2%) to have a common brachiocephalic trunk, of whom 98% had associated congenital cardiac malformations. A spectrum of associated lesions was identified, including left-to-right shunts in 19 patients, right-sided anomalies in 18 patients, left-sided obstructive lesions in 12 patients, and coronary arterial abnormalities in 10 patients, eight of whom had other cardiac defects. Genetic syndromes were present in one-fifth of the cases. When found with left-sided malformations, the common trunk was associated with persistent hypoplasia of the aortic arch, likely related to diminished flow through the arch during development. In each of four patients in whom the brachiocephalic trunk had been used during construction of a palliative shunt, we observed inadequate growth and deformation of the pulmonary arteries. Thus, angiographic identification of a common brachiocephalic trunk may be a marker for the presence of accompanying congenital cardiac defects and coronary arterial abnormalities. Understanding the pathophysiologic effects of the common trunk is important when planning the palliative or corrective procedures, and when assessing the potential benefit of the surgical repair over the long term.

Keywords: Common innominate artery; anomalous coronary artery; congenital heart defects; angiography; embryology

A COMMON BRACHIOCEPHALIC TRUNK IS AN anatomic vascular variant in which both common carotid arteries, together with the right subclavian artery, originate from the aortic arch via a single trunk. The condition was discovered in one-tenth of the specimens of congenital heart defects reported by Edwards,[1] but was found in up to one-quarter of the patients in the radiologic series reported by Bosniak.[2]

Developmental determinants of growth of the aortic arch and isthmus include the placental and cerebral circulations with their resultant ventricular output and afterload.[3] Since diminished flow to the ascending aorta produced during fetal life by lesions such as aortic stenosis is associated with coarctation of the aorta and isthmic hypoplasia,[4] a question arises as to whether a common brachiocephalic trunk may similarly affect distal flow through the arch due to preferential proximal shifting of the distribution of flow of blood to the brain.

Thus, we hypothesized that the anatomic severity of congenital cardiac malformations may be altered in the presence of a common brachiocephalic trunk. Specifically, we sought to investigate clinically whether left-sided cardiac defects, in the presence of a common brachiocephalic trunk, are associated with hypoplasia of the arch and isthmus, and whether the presence of a common brachiocephalic trunk affects the management of accompanying congenital cardiac defects.

Methods

We derived our cohort of patients from the registry of congenital heart diseases in the Division of Pediatric Cardiology, The Children's Heart Center, Medical

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Accepted for publication 2 July 2003

College of Virginia,Virginia Commonwealth University. Cardiac catheterizations and angiograms performed from 1992 to 2002 were reviewed by two experienced cardiologists who reached an agreement concerning the diagnosis of common brachiocephalic trunk. The presence of associated congenital cardiac malformations, and other clinically relevant information, were recorded and documented. We then reviewed the clinical details of the patients found to have a common brachiocephalic trunk, including cardiac surgical procedures and related results.

Results

A total of 1480 cardiac catheterizations had been performed in children over the period of study. A common brachiocephalic trunk was identified in 48 (3.2%) individuals, evenly distributed between females and males, with a ratio of 1.1 : 1, respectively. The age at the time of the cardiac catheterizations ranged from newborn infants to 12 years. Genetic syndromes were identified in ten (21%) individuals, of whom five had Down's syndrome. There was one individual each with CHARGE, Goldenhar, fetal alcohol, Williams, and Turner syndromes.

Of the patients, 47 (98%) had associated cardiac malformations (Fig. 1). All categories of lesions were represented, with left-to-right shunts, and obstructive lesions of the right and left hearts predominating. We also discovered patients with discordant ventriculo-arterial connections, of both the usual and congenitally corrected varieties, as well as finding a left-sided common brachiocephalic trunk in two patients with right aortic arch. The only example of a common brachiocephalic trunk without associated cardiac malformations was found in a patient who underwent cardiac catheterization for evaluation of a possible arteriovenous malformation in the neck. Among patients with atrioventricular septal defect and common atrioventricular junction guarded by a common valve, we found no examples of obstruction within the left heart.

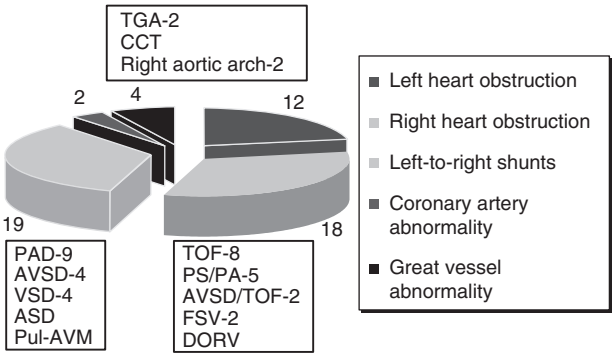

Figure 1.
Distribution of congenital cardiovascular defects accompanying a common brachiocephalic trunk. ASD: atrial septal defect; AVSD: atrioventricular septal defect with common valve; CCT: congenitally corrected transposition; DORV: double outlet right ventricle; FSV: functionally single ventricle; PA: pulmonary atresia; PAD: patent arterial duct; PS: pulmonary stenosis; Pul-AVM: pulmonary arteriovenous malformation; TGA: discordant ventriculo-arterial connections; TOF: tetralogy of Fallot; VSD: ventricular septal defect.

The majority (58%) of patients with obstructive left heart defects had hypoplasia of the aortic arch, including all patients with coarctation of the aorta, and those with interruption of the aortic arch at the isthmus (Fig. 2). The hypoplasia of the arch persisted following surgical repair of the coarctation or interruption, or balloon angioplasty, in all patients, including a 10-year-old who had undergone successful repair of coarctation by means of the subclavian flap angioplasty at the age of 7 months (Fig. 3). In one patient with isolated aortic valvar stenosis, hypoplasia of the arch was observed in the absence of obstruction distally with the arch (Fig. 4).

Coronary arterial abnormalities were seen in 21% of the patients (Table 1). In 5 patients, there was anomalously high take-off above the sinuses of Valsalva of one or both coronary arteries. In one patient, who had a combination of aortic stenosis, coarctation of the aorta and hypoplasia of the arch, the right coronary artery arose anomalously above the left sinutubular junction and coursed between the arterial trunks (Fig. 5).

Among our 18 patients with right-sided obstructive defects, 6 (33%) underwent balloon dilation of the pulmonary valve or arteries and/or primary surgical repair. Various palliative procedures were performed on the other 12 patients, including construction of palliative shunts. Interposition of a tube graft from the subclavian artery arising from the common brachiocephalic trunk in 4 patients to the pulmonary

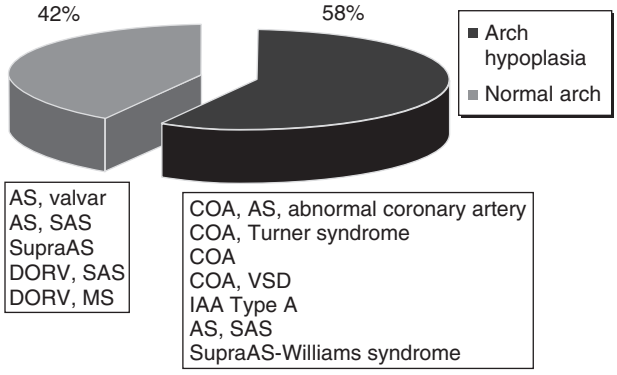

Figure 2.
Left heart obstruction defects and aortic arch hypoplasia. AS: aortic stenosis; COA: coarctation of the aorta; DORV: double outlet right ventricle; IAA: interrupted aortic arch; MS: mitral stenosis; SAS: subaortic stenosis; VSD: ventricular septal defect.

https://doi.org/10.1017/S1047951103001136 Published online by Cambridge University Press

artery on the same side was associated with poor growth of the pulmonary arteries (Fig. 6A), subsequent thrombosis of the shunt and need for construction of a second shunt, or distal stenosis at the anastomosis and tenting upwards of the pulmonary artery (Fig. 6B). Pulmonary arterioplasty was required in each of these four patients at the time of complete repair. In two patients, one with tetralogy of Fallot and right aortic arch (Fig. 7), it had been possible to construct an interposition graft from the base of the common trunk, or a central shunt, without complication.

No complications of catheterization had occurred due to the presence of the common brachiocephalic trunk.

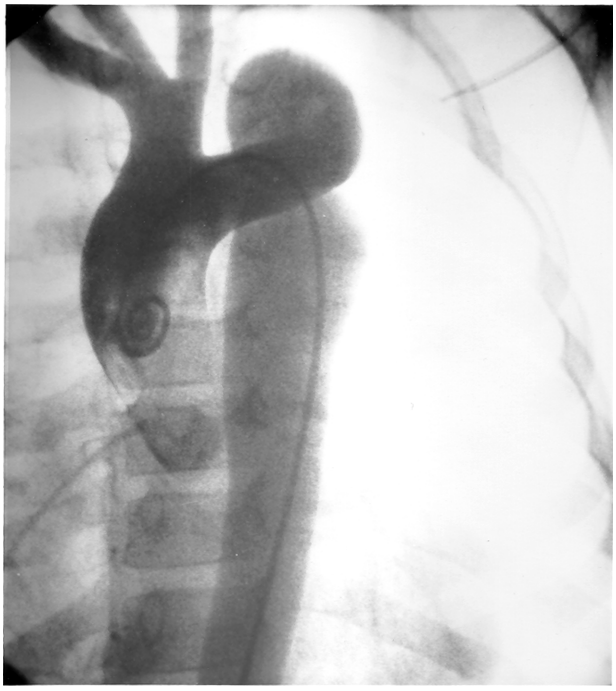

Figure 3.
Aortic angiogram of 10-year-old with coarctation of the aorta repaired by subclavian flap angioplasty at 7 months of age. Residual arch hypoplasia was associated with a resting gradient of 20 mmHg. The common brachiocephalic trunk is demonstrated.

Table 1. Coronary arterial abnormalities accompanying common brachiocephalic trunk.

Coronary abnormality	Cardiovascular defect
Congenital AV fistula: RCA to RA	Isolated
Congenital AV fistula: high RCA to RA	Tetralogy of Fallot
Left CxCA from right aortic sinus	Isolated
LAD from RCA	Tetralogy of Fallot
LAD from RCA	Double outlet RV
Anomalous RCA from left STJ	Coarctation of aorta, AS
Hypoplastic coronary arteries	Williams syndrome, SupraAS
High RCA take-off, dilated LCA	IAA Type A, post-repair
High RCA and LCA take-off	Down's syndrome, AVSD
High RCA take-off	VSD

Abbreviations: AS: aortic stenosis; AV: arteriovenous; AVSD: atrioventricular septal defect with common valve; CxCA: circumflex coronary artery; IAA: interrupted aortic arch; LAD: left anterior descending coronary artery; LCA: left coronary artery; RA: right atrium; RCA: right coronary artery; RV: right ventricle; STJ: sinutubular junction; VSD: ventricular septal defect

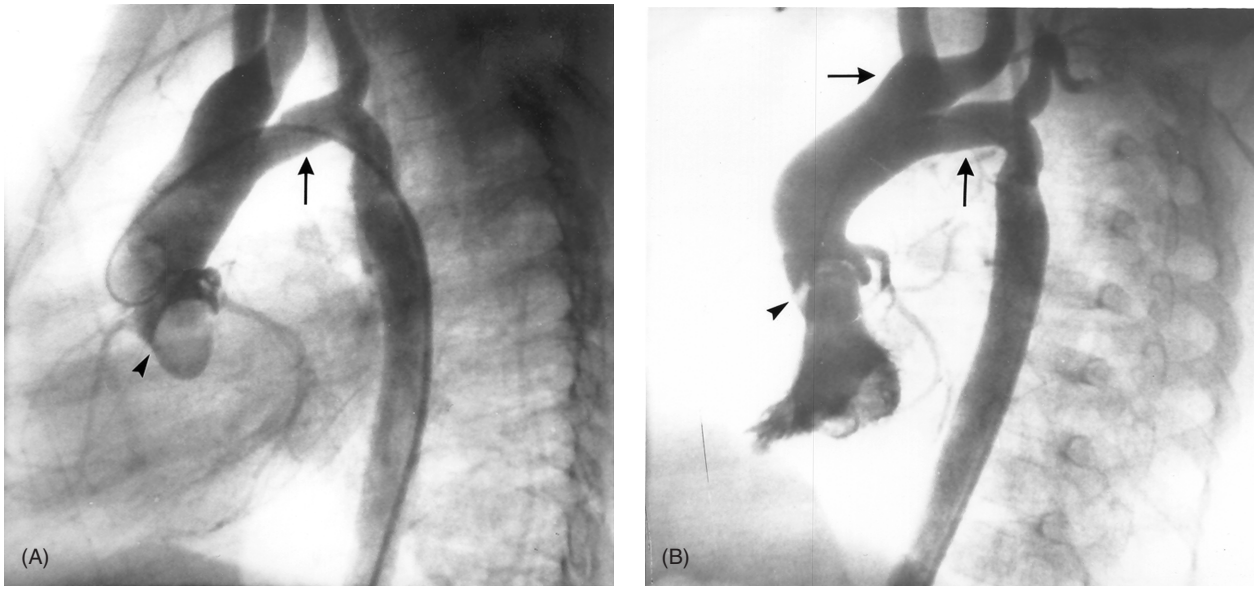

Figure 4.
Aortogram (A) and left ventriculogram (B) of 1-year-old with common brachiocephalic trunk (horizontal arrow), aortic valvar stenosis (arrow head) and significant hypoplasia of the aortic arch (vertical arrow) without coarctation of the aorta. The total gradient from the left ventricle to the descending aorta was 85 mmHg.

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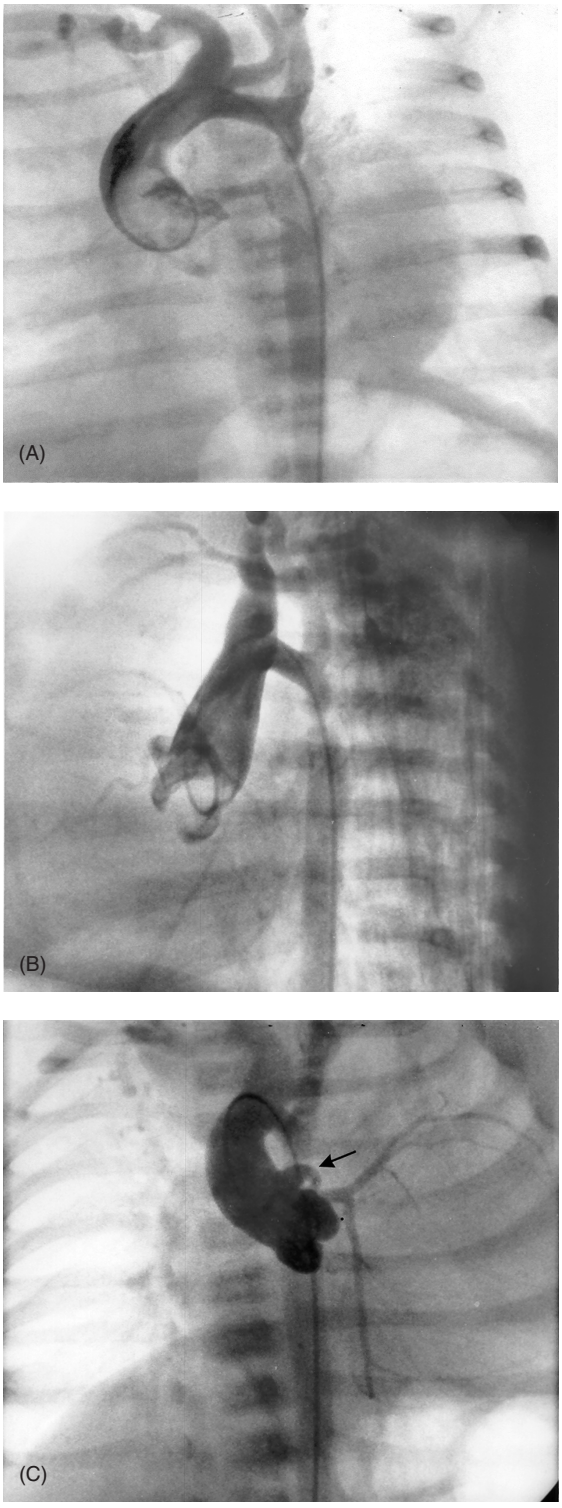

Figure 5.
Aortogram of 1-month-old infant with bicuspid aortic valve, coarctation of the aorta, and hypoplasia of the aortic arch and isthmus. (A) Anterior projection shows common brachiocephalic trunk, hypoplasia of the arch, and coarctation of the aorta. (B) Lateral projection revealing bicuspid aortic valve and significant hypoplasia of the arch. (C) Anterior projection of aortic root angiogram demonstrating anomalous origin of the right coronary artery above the left sinutubular junction (arrow).

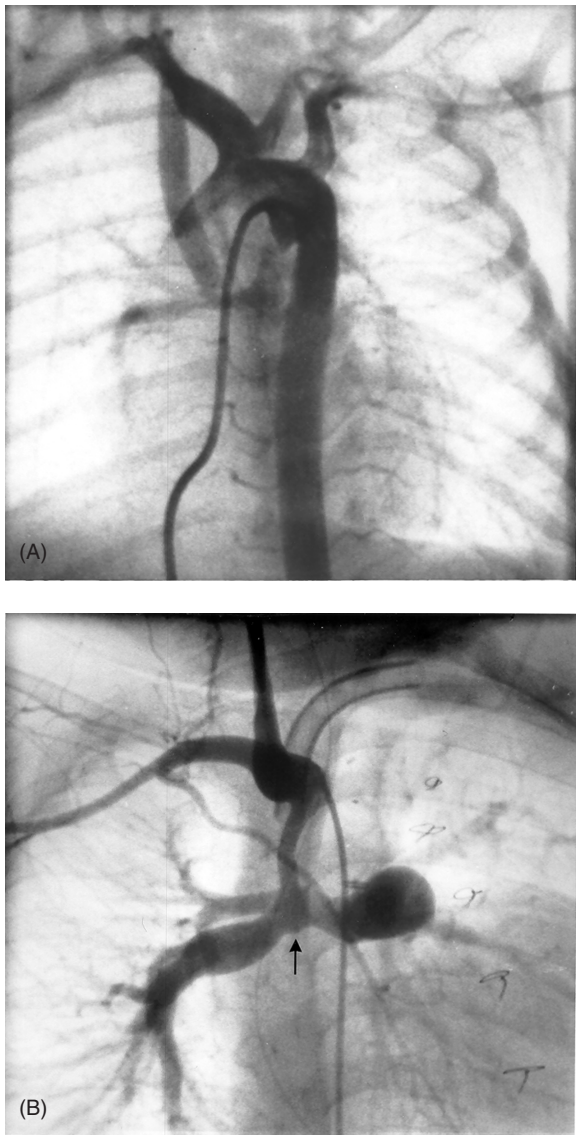

Figure 6.
(A) Three week old infant with 4 mm interposition graft from the subclavian artery arising from a common brachiocephalic trunk to the right pulmonary artery. Despite patency of the shunt, the pulmonary arteries fill poorly. Note the length of shunt (5 rib spaces). (B) 10-month-old with pulmonary atresia, and intact ventricular septum who, as a neonate, had an interposition graft placed from the distal common brachiocephalic trunk to the right pulmonary artery. Stenosis and upward tenting of the right pulmonary artery at the shunt site (arrow) is evident.

Discussion

The findings of our study are three-fold:

- First, when present and accompanied by congenital cardiac defects, especially left-sided defects, a common brachiocephalic trunk impacts on the severity and complexity of the associated lesions.
- Second, palliation of obstructive right-sided malformations incorporating a common brachiocephalic

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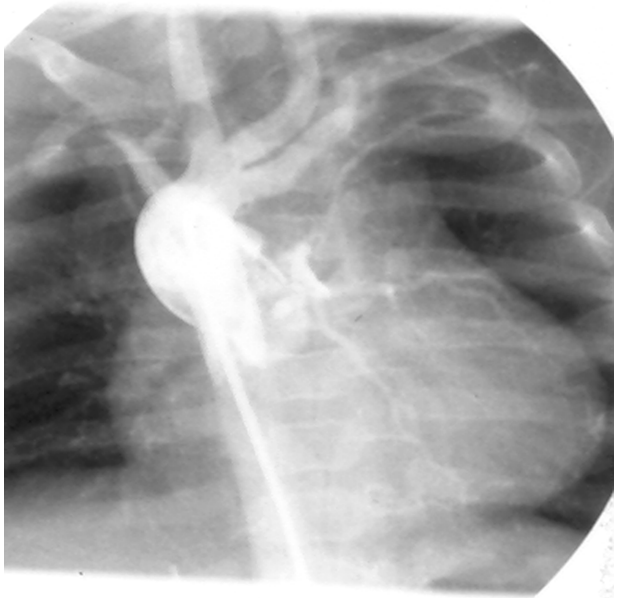

Figure 7.
A four month old with severe tetralogy of Fallot and right aortic arch. A left common brachiocephalic trunk is demonstrated.

trunk may be inadequate, or cause deformation of the pulmonary arteries requiring additional surgical procedures.

- Third, the presence of a common trunk may be a marker for important coronary arterial abnormalities.

Our finding of an angiographic incidence of 3.2% supports the notion that the anomaly itself is a normal variant of branching not typically associated with congenital cardiac anomalies. Importantly, a significant number of our patients had genetic syndromes, including several with Down's syndrome. This relationship has not been reported previously. Genetic syndromes may play a role in the development of the common brachiocephalic trunk, especially in those cases in which hemodynamic alterations cannot account for its formation. Variants in the configuration of the vessels arising from the arch are multiple. Common brachiocephalic trunk is commonly seen in mammals such as dogs, cats, pigs and rabbits. An arterial trunk giving rise to four arteries is seen in hoofed mammals. These evolutionary patterns may re-emerge in humans in association with genetic syndromes. In that regard, unusually close anatomic proximity between the brachiocephalic and left common carotid arteries or a common trunk were present in almost half of an autopsied series of cases of Trisomy 18.[5] Moreover, four-fifths of these cases had high take-off of an anomalous right coronary artery. This suggests, as we observed in our patients, that there may be a common relationship between migration of vessels along the aortic arch and the process which triggers ingrowth of the coronary arteries into the aorta at specific anatomic locations.[6]

Crucial developmental determinants of growth of the aortic arch and isthmus include the dynamic balance of distribution of the flow of blood between the upper and lower parts of the body.[3] The normally smaller diameter of the arch in a newborn as compared with the sizes of the ascending and descending aorta is a result of this balance.[7] All arteries arising from the aortic arch are dedicated to the upper body. Normally, the smallest segment of the arch is the aortic isthmus. The flow of blood in the subclavian artery approximates that of the common carotid artery. With common brachiocephalic trunk, the distal arch, representing the persistent fourth arch, carries a reduced flow, more so if the subclavian artery is aberrant. The distal arch may lack the capacity to grow even if the flow through it is enhanced.

Diminished flow to the ascending aorta in fetal life due to cardiac defects may result in coarctation of the aorta, along with hypoplasia of the arch and isthmus.[4] Conceivably, in a similar mechanism, a common brachiocephalic trunk may reduce flow through the distal arch by preferentially shifting the distribution of flow of blood to the brain within the proximal portion of the arch. Indeed, we found that a common trunk was associated with significant hypoplasia of the arch with or without discrete coarctation of the aorta. We speculate that the degree of hypoplasia was worsened by the common trunk. This effect on the arch appears to be permanent, as demonstrated in our patients where the hypoplasia persisted, with residual gradients, following successful intervention on left-sided obstructive defects.

As we have shown, the common brachiocephalic trunk may also occur in the setting of a right aortic arch. Normally, the brachiocephalic artery is the first vessel to arise from the aortic arch. When the arch is left-sided, the right brachiocephalic artery is joined by the left common carotid artery to form the common brachiocephalic trunk. In the human, the common brachiocephalic trunk may cause persistence of the embryonic segment of the fourth aortic arch that separates the subclavian artery from the common brachiocephalic trunk. Indeed, in some mammals, both subclavian arteries originiate from the common brachiocephalic trunk. The final configuration of the aortic arch can also be affected by abnormal intrauterine hemodynamics. When a small arterial duct supplies the pulmonary circulation, giving aortic to pulmonary flow, the fourth aortic arch becomes incorporated into the definitive transverse aortic arch. This large caliber channel facilitates both the systemic venous return towards the placenta, and the ductal supply to

the pulmonary circulation, producing the so-called "tetralogy physiology". When the ascending aortic inflow is compromised as described above, the large arterial duct carries the systemic venous return towards the placenta. The fourth aortic arch, as well as the aortic isthmus, is then often hypoplastic. When flow through the ascending aorta is critically compromised in early development, ductal flow must divide to supply the subclavian and vertebral arteries, or in case of aortic atresia, the entire aortic arch, as well as the descending aorta. The posterior shelf dividing the flows, itself incorporating ductal tissue, may develop into a shelf-like coarctation following ductal closure.

With right aortic arch, the sequence of arch arteries is the mirror-image of the configuration seen in the left arch. The left brachiocephalic artery and the right common carotid artery are joined at the base, and form the common brachiocephalic trunk. The right subclavian artery originates separately. Among our 18 patients with reduced pulmonary blood flow (Fig. 1), two had right aortic arch, one with tetralogy of Fallot (Fig. 7). With "tetralogy physiology", the arterial duct supports the pulmonary circulation. In the majority of cases of right aortic arch, the arterial duct is small, and originates from the left subclavian artery. As the only path to the placenta, the right aortic arch distal to the common brachiocephalic trunk cannot be significantly hypoplastic. Thus, in the presence of such a duct, tubular hypoplasia of the arch, and/or aortic coarctation, should be extremely rare. A right "systemic" arterial duct connected to the right descending aorta occurs only with severe coarctation or interruption in the right arch, and with common arterial trunk. The right subclavian artery is often aberrant. With interruption of the aortic arch, the ascending aorta resembles a common brachiocephalic trunk.

An interesting finding in our study was the relatively high incidence of coronary arterial abnormalities. Such anomalies occurred in association with specific cardiac defects, and should be considered of surgical importance. Examples are the anomalous left anterior descending coronary artery originating from the right coronary artery in tetralogy of Fallot,[8,9] ostial stenosis or proximal atresia of a coronary artery in Williams syndrome with supravalvar aortic stenosis,[10] and origin of the left circumflex coronary artery from the right sinus of Valsalva as an isolated variant.[11] In our series, the identification of five cases of anomalous high origin of a coronary artery, two cases of the right coronary artery involved in an arteriovenous fistula, and a case of anomalous right coronary artery arising high above the left sinus of Valsalva, altogether suggest an abnormal embryologic process of migration or ingrowth of the coronary vessels.

The processes which trigger the ingrowth of the coronary arteries into the aortic sinuses are still under investigation. Descriptive and experimental studies show that coronary endothelial and smooth muscle precursors self-organize in the subepicardial, or proepicardial, space, forming a vascular plexus that encircles the base of the aorta. Arterioles and arteries form only after this plexus penetrates the aorta to give rise to the roots of the two main coronary arteries.[6,12] Mechanical factors, such as shear stress, wall tension, and stretch have long been implicated in vascular growth.[13] During embryonic cardiac development, before formation of the coronary arteries, the major mechanical influences on the vascular tubes are most likely stretch imposed by diastolic filling and stretch associated with the expanding myocardium. Stretch of cardiac myocytes and endothelial cells has been shown to serve as a stimulus for increases in growth factors and receptor proteins that result in increases in proliferation, migration, and tube formation.[14] We speculate that early in embryologic development, the presence of a common brachiocephalic trunk in association with other cardiac defects alters diastolic filling within the base of the aorta because of an increased runoff to the cerebral circulation. This altered filling pattern may modulate both mechanical and metabolic factors influencing epicardial coronary arterial migration and ingrowth into the aorta that may explain the high incidence of coronary arterial abnormalities observed in our study.

Knowledge of the presence of a common brachiocephalic trunk prior to surgical palliation or repair of congenital heart defects is important for planning the conduct of the procedure, and for improving long term results. Our observations suggest that a more aggressive surgical approach at the time of coarctation repair may be warranted in patients with common brachiocephalic trunk in order to address the permanent hypoplasia of the transverse and distal aortic arch that we observed subsequent to repair. Patch aortoplasty, or implantation of stents, may be useful in older patients. Suboptimal palliation of right-sided defects by interposition of a graft from a subclavian artery originating from a common brachiocephalic trunk may be a result of either cerebral steal or exceeding length of the tube. Thus, we would suggest construction of a central interposition graft in this setting. Alternatively, placement of an interposition graft very proximal on the common trunk may provide better palliation while minimizing deformation of the pulmonary arteries. Thorough evaluation of the location, anatomy, course and drainage of the coronary arteries is also required prior to surgical intervention in any congenital cardiac defect associated with a common brachiocephalic trunk.

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