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Single Origin of Right and Left Pulmonary Artery Branches from Ascending Aorta with Nonbranching Main Pulmonary Artery: Relevance to a New Understanding of Truncus Arteriosus

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Abstract. We report the third known case of origin of the right and left pulmonary artery branches from the ascending aorta via a short common pulmonary artery. A large unbranching main pulmonary artery opened through a patent ductus arteriosus into the descending thoracic aorta. Preductal coarctation of the aorta and multiple congenital anomalies were also present. This rare cardiovascular malformation facilitates a new anatomic and developmental understanding of truncus arteriosus.

Key words: Origin of pulmonary artery branches — Embryology — Truncus arteriosus

Origin of the right and left pulmonary artery branches via a short common pulmonary artery from the posterior wall of the ascending aorta, with a large nonbranching main pulmonary artery opening through a patent ductus arteriosus into the descending thoracic aorta, has been reported only twice previously [1, 2]. This rare anomaly, of considerable interest *per se*, also appears to clarify what truncus arteriosus really is both anatomically and developmentally.

Case Report

A 2-day-old female infant weighing 2200 g was admitted to the Federico Gomez Children's Hospital in Mexico City with a history of vomiting, no bowel movements, tachypnea, hypotension, and cyanosis since birth. The abdomen was distended, without peristaltic sounds, and a nasogastric tube drained serous fluid. The patient suffered a cardiac arrest 10 hours after admission. Although resuscitated, the infant sustained severe brain damage and expired 38 hours after admission.

A complete autopsy showed that the segmental anatomy of the heart was {S,D,S} (normal), with concordant atrioventricular and ventriculoarterial alignments (Fig. 1). Septum primum was almost totally absent, resulting in a common atrium. The atrioventricular valves, ventricular septum, and ventricular outflow tracts were unremarkable.

The main pulmonary artery was of good size (9 mm in external diameter) but gave origin to no pulmonary artery branches (Figs. 1A and 1B). The only outlet from the main pulmonary artery was a patent ductus arteriosus (external diameter 6 mm, internal diameter 3 to 4 mm) that opened into the descending thoracic aorta (external diameter 9 mm) (Fig. 1B).

The right and left pulmonary artery branches both originated from the posterior wall of the ascending via a single ostium (internal diameter 3 mm) (Fig. 1C) that led into a short "common" pulmonary artery (Fig. 1D). The ostium of the common pulmonary artery lay 8 mm above an unremarkable aortic valve and 7 mm below a normal appearing innominate artery. The common pulmonary artery proceeded posteriorly (dorsally) for 5 mm and then bifurcated into left and right pulmonary artery branches (Fig. 1D). The common pulmonary artery measured 4 mm in external diameter, whereas the left and right pulmonary artery branches both measured 3 mm in external diameter.

Both right and left main stem bronchi were hyparterial (Fig. 1D), both lungs were bilobed, and polysplenia was present (two small splenuli, each weighing 1.5 g). The liver was bilaterally symmetrical, the gall bladder was absent, and the inferior vena cava was interrupted with an enlarged azygos vein draining into the right superior vena cava.

Marked congenital stenosis was present of the lower trachea (Fig. 1D), with complete tracheal rings. Severe airway stenosis also involved the carina and the proximal main stem bronchi, with luminal narrowing to a diameter of less than 1 mm.

The left-sided aortic arch had marked tubular hypoplasia of the aortic isthmus with severe preductal coarctation (internal diameter 1 mm) (Fig. 1E). Biventricular hypertrophy and enlargement were present (Table 1).

Additional associated anomalies included tracheomalacia, stomach in the right upper quadrant, atresia of the first portion of the duodenum with annular pancreas, intestinal malrotation with cecum and appendix in the left upper quadrant, horseshoe kidneys,

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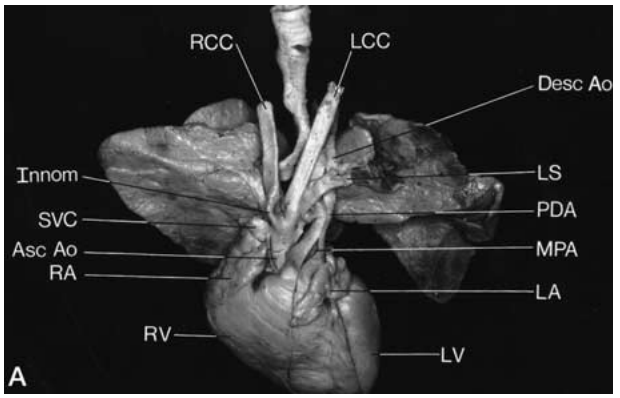

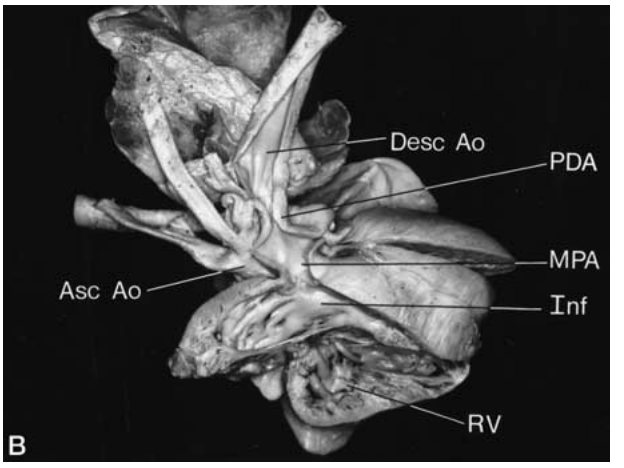

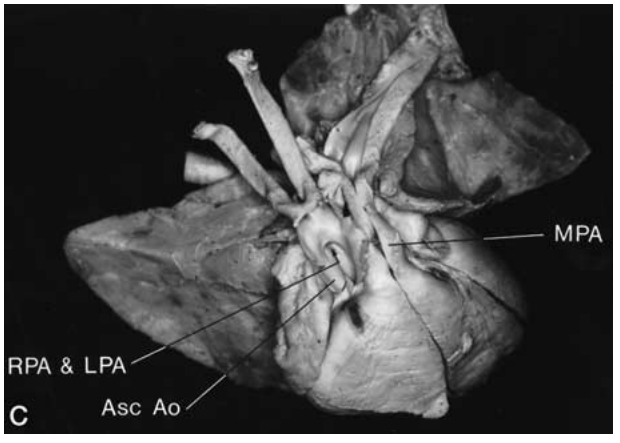

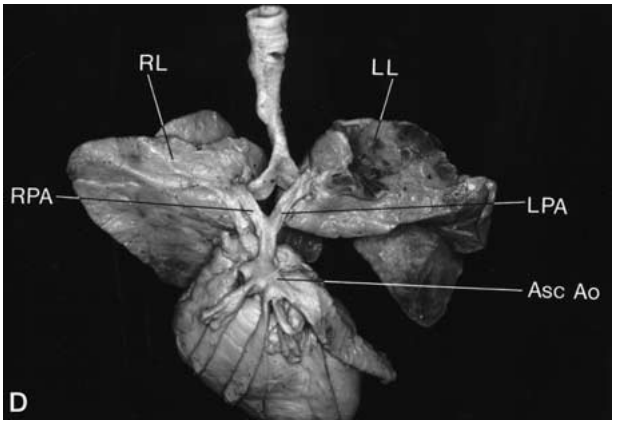

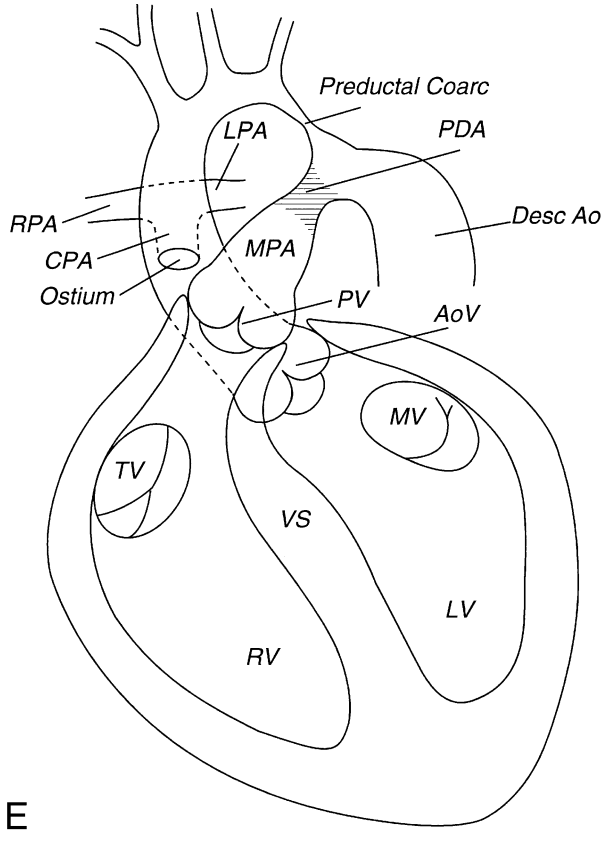

Fig. 1. (**A**) Heart and lungs viewed from the front. The atria are *in situs solitus* (*S*), with the morphologically right atrium (*RA*) lying to the right and the morphologically left atrium (*LA*) to the left. D-loop ventricles (*D*) are present, with the morphologically right ventricle (*RV*) being right-sided and the morphologically left ventricle (*LV*) being left-sided. The great arteries are solitus normally related (*S*), with the main pulmonary artery (*MPA*) arising anteriorly and to the left of the origin of the ascending aorta (*Asc Ao*). Hence, the segmental anatomy is {S,D,S} with concordant atrioventricular and ventriculoarterial alignments. A left aortic arch is present and the lungs are bilaterally bilobed. *Desc Ao*, descending aorta; *Innom*, innominate artery; *LCC*, left common carotid artery; *LS*, left subclavian artery; *PDA*, patent ductus arteriosus; *RCC*, right common carotid artery; *SVC*, superior vena cava. (**B**) Opened RV, MPA, PDA, and Desc Ao. The ventricular septum is intact. No pulmonary artery branches arise from the MPA. A narrowed PDA leads into the Desc Ao. *Inf*, infundibulum. (**C**) Opened Asc Ao showing single opening leading to the right pulmonary artery (*RPA*) and the left pulmonary artery (*LPA*). (**D**) Looking down from above, one sees a short common pulmonary artery that arises from the posterior surface of the Asc Ao and then bifurcates into RPA and LPA. *LL*, left lung; *RL*, right lung. Note the marked right-sided concavity and narrowing of the lower trachea in **A** and **D**. (**E**) Diagram of the salient cardiovascular anomalies. *AoV*, aortic valve; *Coarc*, coarctation; *CPA,* common pulmonary artery; *MV*, mitral valve; *PV*, pulmonary valve; *TV*, tricuspid valve; *VS*, ventricular septum.

agenesis of the ovaries and fallopian tubes, atresia of the inferior one third of the urinary bladder, atresia of the inferior one third of the vagina, the absence of the urethral meatus, and the absence of the hymen and clitoris.

Thus, although this patient had the heterotaxy syndrome with polysplenia, she also had additional severe malformations, some of which were consistent with the XO Turner syndrome; however, the karyotype was not determined.

Discussion

This rare patient and those of Beitzke and Shinebourne [2] and Aotsuka and colleagues [1] are consistent with the view that the normal pulmonary artery and its branches have a bipartite origin: main pulmonary artery from the truncus arteriosus and both right and left pulmonary artery branches from the aortic sac [5].

Although standard embryology textbooks state that the right and the left pulmonary artery branches arise from the sixth aortic arches [7], in order to understand the anomaly being presented, it is necessary to know that the pulmonary artery branches originate initially from the ascending aorta (aortic sac) at the 4-mm stage (24–26 days of age), before the sixth arches have been completely formed [5]. Completion of both sixth arches can occur as early as the 5-mm stage (26–28 days of age) and usually occurs by the 6-mm stage (28–30 days of age) [5]. As soon as both sixth arches have been completely formed, they enlarge considerably and both pulmonary artery branches appear to migrate out onto the sixth arches: this is where the standard account of the development of the pulmonary artery branches in embryology textbooks comes from [7].

Occasionally, the right or left pulmonary artery branch remains arising from the ascending aorta, resulting in "hemitruncus" [8]. Only very rarely do *both* pulmonary artery branches remains arising from the ascending aorta (Fig. 1) [1, 2].

The embryonic morphogenesis of origin of both pulmonary artery branches from the ascending aorta remains speculative. Our hypothesis is that, for reasons unknown, the ventral or proximal portions of both sixth arches fail to form [8], thereby preventing the normal migration of both pulmonary artery branches onto the sixth arches. Other hypotheses include abnormal growth of the wall of aortic sac [8] leaving both pulmonary artery branches originating from the ascending aorta following truncal septation, or abnormal truncal septation [4, 6] leaving both pulmonary artery branches originating from the ascending aorta. There is very little evidence to support truncal malseptation [4, 6]; almost always, the aortopulmonary septum appears to have been formed normally [8].

Table 1. Morphometry

	Case (average)$^{(mm)}$	Controls (average)$^{(mm)}$	% change
RV thickness	4	2	100
LV thickness	4.5	3	50
RV inlet length	34	28	21
RV outlet length	35	31	13
LV inlet length	36	34	6
LV outlet length	37	34	9

From the surgical standpoint, origin of both pulmonary artery branches from the ascending aorta can be treated with success [1], as long as lethal associated malformation do not coexist, as they did in our patient.

Origin of right and left pulmonary artery branches from the ascending aorta suggests that typical truncus arteriosus type A2 may well be a solitary aortic trunk, with origin of both pulmonary artery branches from the ascending aorta (not from a common aortopulmonary trunk or truncus arteriosus communis). This hypothesis may explain the absence of an aortopulmonary septal remnant in truncus type A2: no aortopulmonary septal remnant may be present because there is no main pulmonary artery component in truncus type A2. In contrast, an aortopulmonary septal remnant is present in truncus type A1 because there is a main pulmonary artery component in this type [3, 10, 12].

Our hypothesis is that, as classically conceived, there is no such thing as truncus arteriosus communis, i.e., a failure of septation at the great arterial, semilunar valvar, and conal levels. Instead, we hypothesize that truncus arteriosus type A1 is tetralogy of Fallot with infundibular atresia and an aortopulmonary septal defect [3, 12]. The new aspect of our developmental hypothesis is that truncus arteriosus type A2 is tetralogy of Fallot with infundibular atresia, *absence of the main pulmonary artery*, and origin of both pulmonary artery branches from the ascending aorta. The very rare case reported herein and two others like it [1, 2] prove (1) that the right and left pulmonary artery branches can both arise from the ascending aorta, and (2) that origin of the right and left pulmonary artery branches from the ascending great artery does not mean that this great artery must be a truncus arteriosus communis, which is the conventional assumption in typical truncus arteriosus type A2. This rare anomaly clearly does not have truncus arteriosus communis because a separate ascending aorta and main pulmonary artery are definitely present (Fig. 1).

Hence, the hypothesis is that in so-called typical truncus arteriosus communis type A2, origin of the

right and left pulmonary artery branches from the ascending great artery does *not* prove that a common aortopulmonary trunk is present. Instead, the hypothesis is that in truncus type A2, the ascending great artery is the aorta only, from which both pulmonary artery branches originate, as in the present rare anomaly.

In so-called truncus arteriosus communis type A2, why is there typically no remnant of the aortopulmonary septum? We think that the answer may be because in truncus type A2, the main pulmonary artery is absent and consequently there can be no aortopulmonary septal remnant.

In truncus arteriosus type A1, why is the truncus broader below the origin of the pulmonary artery branches than above the pulmonary artery branches, whereas in truncus type A2 the width of the truncus below and above the origin of the pulmonary artery branches is the same? We think that the answer may be because a main pulmonary artery component is present in truncus type A1 but absent in truncus type A2.

Much remains to be learned concerning the anatomy, morphogenesis, and etiology of truncus arteriosus. For example, it may be said that there are two large groups of truncus arteriosus with ventricular septal defect [3, 10–12]: *the large aortic arch group* (types A1–A3) and *the large pulmonary artery and ductus arteriosus group* (type A4). In truncus arteriosus, the development of aortic arch 4 and pulmonary arch 6 varies inversely: (1) large aortic arch, with small or partly absent pulmonary artery (types A1–A3), and (2) large pulmonary artery and ductus arteriosus, with aortic arch hypoplasia, coarctation, atresia, or interruption (type A4). The reasons for this inverse development of the aorta and the pulmonary artery in truncus arteriosus are unknown.

It is noteworthy that the aforementioned morphogenetic hypothesis concerning truncus arteriosus (i.e., that truncus arteriosus is subpulmonary infundibular atresia, with partial or total absence of the pulmonary valve leaflets and with partial or total absence of the main pulmonary artery) appears to apply only to the large aorta and small or absent pulmonary artery group of truncus (types A1–A3) but not to the large pulmonary artery and small or interrupted aortic arch group (type A4).

We suspect that the large aorta group of truncus and the large pulmonary artery group may be significantly different malformations, not only anatomically but also morphogenetically and etiologically. In contrast, the classical nonseptation hypothesis of the morphogenesis of truncus arteriosus assumes that all the aforementioned anatomic differences between the large aortic types (Al–A3) and the large pulmonic type (A4) of truncus are merely differences in associated malformations. Hence, as mentioned previously, truncus arteriosus appears to be much more interesting developmentally than is generally understood.

In conclusion, the very rare anomaly reported herein is not just of great interest *per se*. It also sheds a strong light on the morphogenesis of truncus arteriosus type A2, in which it has long been assumed that a main pulmonary artery component must be present—even though one cannot point to where it is because there is no aortopulmonary septal remnant in truncus type A2. There is also no aortopulmonary septal remnant in truncus types A3 and A4.

The malformation presented in this article proves that the right and left pulmonary artery branches can arise from the ascending aorta, which contains no component of the main pulmonary artery, because a large and separate main pulmonary artery is clearly present (Fig. 1).

Our current hypothesis concerning the morphogenesis of truncus arteriosus is as follows: First, truncus arteriosus type Al is a truncus arteriosus communis (a common arterial trunk) in the sense that both aortic and main pulmonary artery components are present anatomically, demarcated by an aortopulmonary septal remnant. However, at the valvar level, a common semilunar valve typically is not present in truncus type Al; instead, an aortic valve appears to be present. Also, at the conal level, a typical conal septal defect is not present; instead, atresia of the subpulmonary infundibulum appears to be present, as in tetralogy of Fallot with pulmonary outflow tract atresia. Consequently, the classical nonseptation hypothesis is thought to be erroneous.

Second, truncus arteriosus types A2 and A3 may have truncus aorticus solitarius (solitary aortic trunk), with no main pulmonary artery component, because there is no demarcating aortopulmonary septal remnant or other definite anatomic evidence of a main pulmonary artery component in these two types of truncus arteriosus.

Finally, truncus arteriosus type A4 may have truncus pulmonicus solitarius (solitary pulmonic trunk), with no ascending aortic component, because there is no demarcating aortopulmonary septal remnant or other definite anatomic evidence of an ascending aorta component in this anatomic type of truncus arteriosus.

Truncus arteriosus communis persistens (persistent common arterial trunk) is the only form of congenital heart disease that has been defined, described, and classified in terms of an unproved embryologic hypothesis. The purpose of presenting the previously mentioned relatively new hypothesis is to stimulate further research concerning the pathologic anatomy, morphogenesis, and etiology of truncus arteriosus.

If this relatively new concept of the morphogenesis of truncus arteriosus is correct, then there is no such thing as "true" truncus arteriosus communis persistens (all have "pseudo" truncus). Currently, we call this anomaly truncus arteriosus, not truncus arteriosus *communis,* because we strongly suspect that *communis* is not anatomically accurate in types A2, A3, and A4.

Finally, it should be emphasized that we continue to make the *anatomic diagnosis of truncus arteriosus* just as everyone else does. Anatomically, truncus arteriosus is defined as one great artery arising from the base of the heart, giving origin to the coronary arteries (at least one), to the pulmonary artery branches (at least one), and to the systemic arteries. It is not our intent to change diagnostic anatomic terminology. Indeed, we think, as a practical matter, that diagnostic anatomic terminology should be kept as constant as possible. Instead, our hope is to contribute to the growing understanding of the anatomy, morphogenesis, and etiology of congenital heart disease. The "price" of advances in anatomic and developmental understanding should not be changes in diagnostic anatomic terminology. To minimize confusion, the old terms should be used, insofar as possible, informed with new anatomic and developmental understanding.

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