MALPOSITION OF PULMONARY ARTERIES (CROSSED PULMONARY ARTERIES) IN PERSISTENT TRUNCUS ARTERIOSUS*

By ANTON E. BECKER, M.D., MIES J. BECKER, M.D., and JESSE E. EDWARDS, M.D.
MINNEAPOLIS, MINNESOTA

An uncommon arterial anomaly is that in which the ostium of the left pulmonary artery lies to the right of and above that of the right pulmonary artery. From these positions, the 2 pulmonary arteries cross each other as they proceed to their respective lungs.

Jue and associates proposed the name "crossed pulmonary arteries" for this condition and described an example in a newborn infant with the trisomy 18 syndrome. In addition to the pulmonary arterial malformation in question, their case exhibited an atrial septal defect of the so-called sinus venosus type associated with anomalous connection of the right upper pulmonary vein to the right atrium. The great arteries were otherwise normally developed.

The present report deals with 3 cases, each with persistent truncus arteriosus, in which the ostia of the pulmonary arteries were malpositioned. In 2 of the cases, the pulmonary arteries crossed each other and there was interruption of the aortic arch; in the third case, there was atresia of the aortic arch, but without crossing.

Knowledge of the occurrence of "crossed pulmonary arteries" in the presence of persistent truncus arteriosus associated with malformations of the aortic arch may aid in the interpretation of angiocardograms in appropriate cases.

DESCRIPTION OF SPECIMENS

An analysis was made of 13 specimens with persistent truncus arteriosus, in each of which the pulmonary arteries arose from the truncus (Types i and ii). In 10 of these cases, the left and right pulmonary arteries arose next to each other, the ostium of the left artery lying to the left of the ostium of the right (Fig. 1).

In 3 cases, there were abnormalities in origin of the pulmonary arteries and, in 2 of these, in the courses that all the arteries took. These 3 cases form the basis for the descriptions below.

ILLUSTRATIVE CASES

Case 1. The specimen was from a newborn infant, of whom no other information was available to us. The heart exhibited a classic example of persistent truncus arteriosus, Type i. The branches of the aortic arch were normal.

The segment of the aorta between the left subclavian artery and the ductus arteriosus was represented by an atretic strand. The pulmonary arterial ostia which were malpositioned arose from the pulmonary aspect of the persistent truncus. The orifice of the left pulmonary artery lay 2 mm. superior to and at the right of the orifice of the right pulmonary artery (Fig. 2; and 3, a and b). From these origins, the 2 pulmonary arteries crossed each other as they coursed toward their respective lungs. In crossing, the left pulmonary artery was superior to the right. The ductus arteriosus was patent although narrow. Its pulmonary end appeared to begin from the superior aspect of the pulmonary aspect of the truncus near the left pulmonary artery.

Case 2. A newborn female infant died at the age of 1 week following a surgical procedure for congenital atresia of the duodenum. The heart showed a Type i persistent truncus arteriosus, with complete interruption of the aortic arch. The pulmonary portion of the truncus was dilated and communicated directly through a

---

* From the Departments of Pathology, The Charles T. Miller Hospital, St. Paul, Minnesota and The University of Minnesota, Minneapolis, Minnesota.

This study was supported by Public Health Service Research Grant 5 RO1 HE05694 and Research Training Grant 5 T01 HE05570 from the National Heart Institute and by The Netherlands Organisation for the Advancement of Pure Research (Z.W.O.).
FIG. 1. An example of normally oriented pulmonary arteries in persistent truncus arteriosus (Type I). The ostia of the pulmonary arteries lie next to each other, with the ostium of the left pulmonary artery (L.P.) to the left of that of the right pulmonary artery (R.P.). R.V. = right ventricle; Ao. = ascending aorta beyond truncus.

widely patent ductus arteriosus with the descending aorta. The left subclavian artery arose from the descending aorta opposite the ductus arteriosus. The ostia of each of the pulmonary arteries were separated by 9 mm. The orifice of the left pulmonary artery was located superiorly and slightly to the right of the ostium of the right pulmonary artery. There was, as in Case 1, a typical pattern of "crossed pulmonary arteries," with the left pulmonary artery crossing superior to the right (Fig. 4; and 5, a and b).

Case III. A female infant, born at 32 weeks gestational age, died 1 hour after birth with multiple congenital malformations associated with the trisomy 18 syndrome. The heart showed a classic example of persistent truncus arteriosus, classified as Type I and associated with interruption of the aortic arch. There was no continuity between the walls of the aortic arch and descending aorta. The short pulmonary trunk, after its origin from the persistent truncus, was dilated and connected through a patent ductus arteriosus with the descending aorta. The left subclavian artery originated from the descending aorta, immediately distal to the orifice of the ductus arteriosus. Each of the 2 main pulmonary arteries originated from the pulmonary portion of the truncus. The orifice of the left pulmonary artery, which was narrow, was positioned superior to that of the right pulmonary artery (Fig. 6, a and b). From these origins, the vessels proceeded to their respective lungs without crossing.

DISCUSSION

Although congenital anomalies of major pulmonary arteries are uncommon, a variety of entities come under this category. Included are such conditions as: (1) localized stenosis or hypoplasia of 1 or both branches; (2) absence of 1 or each artery; (3) anomalous origin of either 1 or both branches from a vessel other than the pulmonary trunk; and (4) malposition of the origins of the pulmonary arteries, either

Fig. 2. Case 1. Schematic drawing of vascular abnormalities. Persistent truncus arteriosus with atresia of aortic arch and patent ductus arteriosus (P.D.). The ostium of the left pulmonary artery (L.P.) lies to the right of and above that of the right pulmonary artery (R.P.). The branches cross as each proceeds to its respective lung.
FIG. 3. Case I. (a) Interior of persistent truncus arteriosus. Vessel overrides a ventricular septal defect (D). The orifice of the left pulmonary artery (L.P.) lies to the right and above that of the right pulmonary artery (R.P.). Ao. = ascending aorta. (b) Posterior view of exterior of great vessels showing the left pulmonary artery (L.P.) crossing superiorly to the right pulmonary artery (R.P.). Ao. = ascending aorta.

from the pulmonary trunk or from a persistent truncus arteriosus.

In the case with the latter condition, reported by Jue and associates,2 each pulmonary artery arose from the pulmonary trunk. The origin of the left pulmonary artery lay to the right of and above the orifice of the right pulmonary artery. Each artery crossed the other as it coursed to its respective lung. Jue and his group2 proposed the term “crossed pulmonary arteries” for this anomaly.

In our study, there were 3 cases in which the pulmonary arteries arose anomalously

FIG. 4. Case II. Schematic drawing of vascular abnormalities. Persistent truncus arteriosus with interruption of aortic arch and patent ductus arteriosus (P.D.). The left (L.P.) and right (R.P.) pulmonary arteries show abnormalities similar to that in Case I.
and, in 2 of these, the pulmonary arteries crossed each other. These cases were found among 13 specimens with persistent truncus arteriosus (Types 1 and 11). In the 1 case with anomalously positioned ostia but without crossing (Case III), the malformation is considered a lesser form of classical “crossed pulmonary arteries.” It is of interest that, in each of our cases of persistent truncus with this pulmonary arterial anomaly, interruption or atresia of the aortic arch occurred.

To our knowledge, there have been no reported cases of “crossed pulmonary arteries” in persistent truncus arteriosus, although an anomalous position of the orifices of the pulmonary arteries has been described in this condition. In several reports, a superiorly located orifice of the left pulmonary artery has been mentioned.45 Keith and associates3 described, in persistent truncus arteriosus, the angiocardiographic phenomenon of a high arching left pulmonary artery. Van Praagh and Van Praagh5 attributed this sign to the superior position of the orifice of the left pulmonary artery in persistent truncus. Although mentioning that the left pulmonary artery may arise above the right, the latter authors did not indicate how common this change was among the 57 specimens with persistent truncus that they reviewed. They did not mention the phenomenon of crossing. Victorica and associates,6 in a clinical study of 14 cases of persistent truncus arteriosus in infancy, mentioned that, in roentgenograms, an abnormally high left hilum was an un-
common finding. This could indicate the rarity of malpositioned orifices of the pulmonary arteries. It is of interest that no cases with interrupted aortic arch were encountered in their series. In each of our cases with crossed pulmonary arteries, interruption or atresia of the aortic arch was present.

The developmental basis for the anomalous position of the ostia of both pulmonary arteries is not certain. Jue and associates suggested that this condition could result from faulty differential growth during the process of partitioning of the truncus arteriosus into the aorta and pulmonary trunk.

Although the condition of "crossed pulmonary arteries" is of no hemodynamic significance, knowledge of its occurrence, whether or not in combination with persistent truncus, may aid in the interpretation of certain peculiarities encountered in the angiograms.

**SUMMARY**

Among 13 specimens with persistent truncus arteriosus, there were 3 cases with malposition of the pulmonary arteries. In 2 specimens, the ostium of the left pulmonary artery lay to the right of and above that of the right pulmonary artery. The 2 pulmonary arteries crossed each other as they proceeded to their respective lungs: "crossed pulmonary arteries." The third specimen presented anomalously positioned ostia of the pulmonary arteries but without crossing. This condition is considered to be a lesser form of classical "crossed pulmo-
nary arteries.” Each of these cases also exhibited interruption or atresia of the aortic arch.

Jesse E. Edwards, M.D.
Department of Pathology
The Charles T. Miller Hospital
125 West College Avenue
Saint Paul, Minnesota 55102

REFERENCES
This article has been cited by:

1. Elizabeth Caris, Bhawna Arya. Abnormalities of the Ductus Arteriosus and Pulmonary Arteries 360-381. [Crossref]
6. Feng Li, Jiahui Yang, Jianjun Zhang, Yue Feng. 2016. Crossed pulmonary arteries associated with single atrium in an adult: a case report. *BMC Cardiovascular Disorders* 16:1. [Crossref]
8. Bhawna Arya, Craig A. Sable. Abnormalities of the Ductus Arteriosus and Pulmonary Arteries 317-335. [Crossref]
10. Madhan Kumar Murugan, Priya Jagia, Anita Saxena. 2015. Crisscross pulmonary arteries with partial anomalous pulmonary venous drainage on multislice cardiac CT. *Journal of Cardiovascular Computed Tomography* 9:1, 71-73. [Crossref]
12. Ryo Abea, Soshu Kotani, Kentaro Yamabe, Ryoei Yozu. 2013. Bilateral Banding of Malpositioned Pulmonary Artery Branches. *Pediatric Cardiology* 34:8, 1938-1940. [Crossref]
15. Goran Cuturilo, Danijela Drakulic, Aleksandar Krstic, Marija Gradinac, Tamara Ilisic, Vojislav Parezanovic, Milena Milivojevic, Milena Stevanovic, Ida Jovanovic. 2013. The role of modern imaging techniques in the diagnosis of malposition of the branch pulmonary arteries and possible association with microdeletion 22q11.2. *Cardiology in the Young* 23:2, 181-188. [Crossref]
16. Constantine Mavroudis, Carl L. Backer. Truncus Arteriosus 361-375. [Crossref]
20. Daniel J. Penny, Robert H. Anderson. Common Arterial Trunk 859-874. [Crossref]


29. R. M. Freedom. Anomalies of Aortopulmonary Septation: Persistent Truncus Arteriosus, Aortopulmonary Septal Defect, and Hemitruncus Arteriosus 429-452. [Crossref]