CLINICOPATHOLOGIC CORRELATIONS

Absence of Anatomic Origin from Heart of Pulmonary Arterial Supply

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SUMMARY
Among certain patients with cardiovascular malformations, a common phenomenon is a ventricular septal defect and absence of anatomic origin of the pulmonary arterial supply from the heart. In this situation, true pulmonary arteries may be present or absent. In the latter case, the pulmonary arterial supply comes through bronchial arteries. When true pulmonary arteries are present, the left and right pulmonary arteries either may be in continuity at their origins (confluence) or they may arise from separate sites (nonconfluence).

When confluence is present, the two pulmonary arteries may arise either from an atretic pulmonary trunk or from certain forms of persistent truncus arteriosus. Pulmonary atresia may be localized to the proximal end of the pulmonary trunk or diffuse in this vessel. When nonconfluence is present, each pulmonary artery arises separately from one of three possible sources as follows: (1) a form of persistent truncus arteriosus, (2) through ducti arteriosi from the aortic arch system, or (3) through bronchial arteries from the descending aorta.

Additional Indexing Words:
Pseudotruncus arteriosus Collateral circulation to lungs Bilateral ducti arteriosi

Among some cyanotic patients, there is a common phenomenon of a ventricular septal defect and no direct connection with the heart of the pulmonary arterial supply.

Identification of anatomic detail is possible through selective arteriography.1 From the surgical point of view, these cases represent highly challenging problems. In the latter regard, one crucial point is whether pulmonary arteries are present.2 If pulmonary arteries are present, it is important to determine whether the two are connected with each other (confluence) or whether each arises independently of the other (nonconfluence). With this pivotal point in mind, a classification is presented (Table 1) of those conditions in which there is no direct connection between the heart and the pulmonary arterial system.

In all cases except that in which there is free flow of blood to the lung (persistent truncus types I, II, and III to follow), bronchial and other collateral arteries supplying the lungs are prominent. In some cases, such vessels are present in addition to true pulmonary arteries. In others, true pulmonary arteries are not present.

Pulmonary Arteries Present
When the arterial supply to the lungs is by way of true pulmonary arteries, one must determine whether the two pulmonary arteries display continuity or confluence of their origins or whether each pulmonary artery has a separate source of origin (nonconfluence).

Confluent Origin of the Pulmonary Arteries
When there is confluence of the origins of the left and right pulmonary arteries, these vessels may either arise from an atretic pulmonary trunk or from certain forms of persistent truncus arteriosus.

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Table 1

Classification of Conditions Characterized by Absence of Direct Connection of Pulmonary Arterial Supply with the Heart

I. Pulmonary arteries present
   A. Confluence of pulmonary arterial origins
      1. Atresia of pulmonary trunk (with patent ductus arteriosus or bronchial arterial collaterals)
         a. Proximal
         b. Diffuse
            1. Fibrouslike remnant
            2. No remnant identifiable
      2. Persistent truncus arteriosus, type I or II
   B. Noneonfluence of pulmonary arterial origins
      1. Origins from persistent truncus arteriosus, type III
      2. Independent ductal origins from aortic arch system
      3. Independent bronchial arterial origins from descending aorta

II. Pulmonary arterial agenesis or atresia (persistent truncus, type IV)

III. Mixed types

Atresia of Pulmonary Trunk

Atresia of the pulmonary trunk may be represented by one of two basic forms as follows: (1) atresia of the proximal end of the pulmonary trunk while the distal end is patent, or (2) atresia of the entire length of the pulmonary trunk. In some cases, the pulmonary trunk may be identified as a fibrouslike remnant while, in others, no tissue is identifiable as that of pulmonary trunk.

Proximal Atresia

Atresia of the proximal end of the pulmonary trunk with patency of the distal end is usually associated with the intracardiac characteristics of the tetralogy of Fallot, the condition commonly called pseudotruncus arteriosus (fig. 1). There is atresia at the anticipated level of the pulmonary valve while, more distally, the pulmonary trunk is patent, though variable in diameter from case to case. The pulmonary trunk bifurcates normally into the right and left pulmonary arteries. A right aortic arch is commonly associated (30%). The ductus arteriosus may be absent or present and, if present, closed or patent. If a ductus is present in association with a right aortic arch, it usually runs between a left-sided innominate artery and the left pulmonary artery. Less commonly, an anomalous left subclavian artery is present. In such cases, if a ductus is present, it runs between the left pulmonary artery and the anomalous left subclavian artery.

Diffuse Atresia

When there is diffuse atresia of the pulmonary trunk, the intracardiac malformations may either be like those in tetralogy or more complicated, including various types of transposition of the great vessels (fig. 2). When a fibrouslike remnant of the pulmonary trunk is present, the cord runs from the base of the right ventricle to confluent origins of the two pulmonary arteries. The ductus may be absent or present, either patent or closed. If a ductus is present, its pulmonary arterial insertion is at the

Figure 1

Confluence of pulmonary arteries. Proximal atresia of pulmonary trunk.
Confluence of pulmonary arteries. Diffuse atresia of pulmonary trunk. (a. and b.) Pulmonary trunk identifiable as a fibrouslike remnant. (a.) Ductus absent. (b.) Ductus present and patent. (c. and d.) Pulmonary trunk not identifiable. (c.) Ductus absent. (d.) Ductus present and patent.

Persistent Truncus Arteriosus, Types I and II

Persistent truncus arteriosus is characterized by one arterial vessel leaving the heart and from which the coronary and pulmonary arteries as well as the aorta arise (fig. 3). There are several subdivisions or types of this condition according to the classification of Collett and Edwards.

In that form of diffuse atresia of the pulmonary trunk in which no remnant of a pulmonary trunk is identifiable, the patent aspects of the pulmonary arterial system are like those in diffuse atresia with an identifiable remnant of a pulmonary trunk. The right and left pulmonary arteries are represented by a horizontal arterial channel, the peripheral ends of which proceed into the respective lungs. The channel may be connected to the aorta by a ductus arteriosus which is either patent or closed (fig. 2c and d).

Confluence between the two pulmonary arteries (fig. 2a and b).

Persistent truncus arteriosus type I is characterized by partial septation of the truncus so that a pulmonary trunk of varying length, usually very short, arises from the truncus and, in turn, the pulmonary arteries originate from the vestigial pulmonary trunk (fig. 3a). The origins of the pulmonary arteries are confluent in that each arises from the pulmonary trunk.

In persistent truncus arteriosus type II, each pulmonary artery arises separately but close to the other from the posterior aspect of the truncus.
Confluence of pulmonary arteries. (a.) Persistent truncus arteriosus, type I. (b.) Persistent truncus arteriosus, type II.

Although the origins of the arteries are separate from each other, their closeness favors a view of “confluence” from a surgically technical viewpoint (fig. 3b).

Nonconfluent Origins of the Pulmonary Arteries

When the origins of the right and left pulmonary arteries are separate from each other, nonconfluence may be said to be present. The right and left pulmonary arteries may arise separately from (1) a form of persistent truncus arteriosus, (2) the aortic arch system through ducti arteriosi, or (3) from the descending aorta through bronchial arteries.

Persistent Truncus Arteriosus, Type III

In some instances of persistent truncus arteriosus, each pulmonary artery arises from the truncus arteriosus rather than from a pulmonary trunk (fig. 4). Two types fulfill this definition, one being type II, wherein each pulmonary artery arises from the posterior aspect of the truncus, and type III, in which each pulmonary artery arises from the lateral aspect of the truncus. Type II has been considered among the conditions manifesting confluence. In type III, on the other hand, the distance between the pulmonary arterial origins serves to favor considering this form of persistent truncus arteriosus as showing nonconfluence of the pulmonary arteries (fig. 4).

Independent Ductal Origins from Aortic Arch System

The condition sometimes referred to as “absence of a pulmonary artery” is characterized by the pulmonary trunk being present but having only one branch, while the contralateral lung is supplied either by a pulmonary artery which arises by way of a ductus arteriosus from the aortic arch system or from bronchial arteries, or the supply to the lung may be through bronchial arteries (fig. 5). Rarely, as in cases of Murray and associates, the condition “absence of a pulmonary artery” may occur bilaterally. In this situation when pulmonary arteries are present the pulmonary artery on the side of the aortic arch arises from the ductus originating from the aortic arch (fig. 5a), while the contralateral pulmonary artery arises from the innominate artery through the ductus of that side. This condition may be termed “independent ductal origins of pulmonary arteries from the aortic arch system.” A variation of this occurred in a case observed by Dr. Michael Shea and one of us (J.E.E.). In this situation wherein there was a left aortic arch, the left ductus arteriosus gave rise to the left pulmonary artery. An anomalous right subclavian artery was present as the fourth branch of the aorta and, from this, through a right ductus arteriosus, the right pulmonary artery arose (fig. 5b).

Independent Bronchial Arterial Origins from Descending Aorta

Hypothetically, a situation may occur in which true pulmonary arteries arise from the upper descending thoracic aorta, the stems of origin for the vessels being bronchial arteries (fig. 6). The anatomic details with regard to the pulmonary arterial vessels are those of true pulmonary arteries. The condition functionally is very similar to that condition in which no pulmonary arteries are present and in which the pulmonary arterial supply is from bronchial arteries (see below).
Nonconfluence of pulmonary arteries. Independent ductal origins from aortic arch system. (a.) The left pulmonary artery arises from the left ductus arteriosus which communicates with the aortic arch, while the right pulmonary artery arises from a right ductus arteriosus originating from the base of the innominate artery. (b.) A variant of a in which, in association with anomalous right subclavian artery ductal origin of the right pulmonary artery is through an anomalous right subclavian artery.

Nonconfluence of pulmonary arteries. Independent bronchial arterial origins of pulmonary arteries from descending aorta.

Pulmonary arterial agenesis or atresia (persistent truncus arteriosus, type IV). Arterial supply to lungs through bronchial arteries.
Pulmonary Arterial Agenesis or Atresia
(Persistent Truncus Arteriosus, Type IV)

In the condition termed pulmonary arterial agenesis or atresia, no pulmonary arteries are identifiable (fig. 7). Such vessels had either never formed or had deteriorated so early in development as not to be identifiable. The arterial supply to the lungs is through true bronchial arteries. A synonym for the condition is persistent truncus arteriosus, type IV. While the accompanying diagram (fig. 7) shows one bronchial artery to each lung, the usual state is that multiple vessels arising from the descending aorta proceed to the lungs. Some bronchial vessels may proceed to one lung, while others may divide and supply both lungs. Atria or agenesis of the pulmonary arteries may be considered a nonconfluent type of pulmonary arterial supply.

Mixed Types

Within nonconfluent origin of the pulmonary arterial supply, several variations may occur from the foregoing conditions in which the arterial supply to one lung exhibits the same type of origin as that to the other. Exclusive of those conditions in which one lung is supplied by a pulmonary artery arising from the pulmonary trunk while the other lung is supplied by a pulmonary artery arising either from the aortic arch of the upper descending aorta, there are other types. This relates to nonconfluence of pulmonary arterial origin but of a different type for one lung than for the other. Many types are hypothetically possible, examples being the situation wherein, on one side, a true pulmonary artery arises from the aortic arch system through a ductus while the supply to the other lung is by way of bronchial arteries (fig. 8).

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