Anomalous Coronary Arteries with Special Reference to Arteriovenous-like Communications

Anomalies of the coronary arteries have generally been classified on anatomic rather than on functional grounds. This has led to separation of several anatomic entities which, from a functional viewpoint, should be brought together. The most important anomalies of the coronary arterial system, regardless of their anatomic details, have in common the features of arteriovenous-like communications. Recognition of this fact should lead to a more realistic approach in the functional evaluation of these anomalies and thence to the inevitable uniformity in principle regarding definitive surgical therapy.

The anomalies of the coronary arterial system fall into 3 major groups. The first group represents those anomalies that, under usual circumstances, are of minor significance. The second group is composed of coronary anomalies secondary to the effects of certain primary cardiac malformations. The third group includes anomalies that are not secondary to other conditions and that are of primary and major significance.

The coronary anomalies of minor significance involve those conditions in which the coronary arterial system arises entirely from the aorta and in which the ramifications of the arteries make no unusual communication with the cardiac chambers, the coronary veins, or the pulmonary trunk. In this group are such anomalies as both coronary arteries originating above one aortic sinus, the left circumflex artery originating from the right coronary artery, and the several varieties of single coronary artery. It is well to emphasize that a congenitally single coronary artery is capable of supplying adequately the entire heart, unless, by chance, it becomes the site of acquired disease.

Though the function of the coronary arterial system is essentially normal in patients with these minor anomalies, peculiar courses of the vessels may be responsible for troublesome surgical problems when the anomalies happen to co-exist with certain intracardiac malformations. This is true in some patients with ventricular septal defect, but it is even commoner among patients who have ventricular septal defect associated with pulmonary stenosis (tetralogy of Fallot).

In an occasional case of this type the origin of one coronary artery may be stenotic or atretic. Under these circumstances the other coronary artery sends wide collateral branches to the coronary arterial bed of the stenotic or atretic artery. The troublesome aspect is that some of these branches extend across the anterior aspect of the right ventricle. Because of their location they may interfere with performance of right ventriculotomy and may introduce problems in performance of procedures designed to relieve infundibular stenosis.

The rare secondary anomalies of the coro-
b. Major anomalies of the coronary arteries represented diagrammatically. Sites of communication of coronary arteries with cardiac chambers are portrayed. The termination of the right coronary artery into right-sided chambers, and of the left coronary artery into left-sided chambers is simply displayed for diagrammatic convenience. Either of the coronary arteries may communicate with any of the cardiac chambers.

c. Anomalous communication of coronary arteries arising from the aorta with the pulmonary
nary arterial system are observed in cases of congenital atresia of either the aortic or pulmonary valve in the presence of an intact ventricular septum and of a competent corresponding atroventricular valve. Such a case was described for the first time, as far as I am aware, by Blakeway in 1918. Under these circumstances blood entering the ventricle proximal to the atretic semilunar valve cannot escape in either a forward or a backward direction. It is driven from the ventricle through myocardial sinusoids toward the epicardium (fig. 1a). In the peripheral aspect of the myocardium the enlarged sinusoids converge to form a gross vessel that emerges from the ventricular myocardium into the epicardium, where it unites with branches of the coronary arteries. Anomalies of the secondary type, while of considerable interest to students of embryology of the coronary circulation, are, nevertheless, only of incidental significance in hearts with serious and underlying malformations.

The anomalies of major significance have varied anatomic patterns; but all have in common the functional characteristic of abnormal communication of the coronary arterial system with its high pressure on one hand and a cardiac chamber or vessel with either constant or intermittent low pressure on the other. This naturally sets the stage either for a true arteriovenous shunt or a shunt that is arteriovenous-like in nature. This functional disturbance makes the patient susceptible to cardiac failure, myocardial ischemia, and bacterial endarteritis. In isolated instances, surgical interruption of the shunt has been successfully accomplished.

The major anomalies of the coronary arterial system may be subdivided anatomically into 2 groups depending on the site that is anomalously connected with the coronary arterial system, as follows (figs. 1b and c); (1) communication of the coronary arterial system with the right side of the circulation, including the coronary sinus or a tributary vein thereof, the right atrium, the right ventricle, or the pulmonary trunk, yielding a left-to-right shunt that is demonstrable both by cardiac catheterization and by aortography, and (2) anomalous communication of the coronary arterial system with the left atrium or ventricle, causing a functional disturbance similar to that of aortic insufficiency.

In each of these conditions there is tortuosity and dilatation of the portion of the coronary arterial system which is part of the abnormal communication. The generalized dilatation, at times, becomes complicated by formation of a localized saecular aneurysm. The latter feature, especially, and the uniform dilatation, to some extent, have been over-emphasized in many reports on these conditions and the primary problem, that is, the arteriovenous communication, has been given secondary consideration. This practice also is responsible for the misleading titles of a number of reports that, although primarily concerned with anomalous arteriovenous communications, have titles that solely concern or prominently display such terms as "coronary aneurysm" or "congenital coronary aneurysm." The reader will realize that arterial dilatation is simply a feature common to all arteries proximal to abnormal arteriovenous communications. It is recognized that the literature contains reports of coronary arterial aneurysms that are claimed simply to be examples of localized arterial disease on a congenital basis. In some of these reports the picture as presented is consistent with that of an anomalous communication although no

trunk. In some instances, both coronary arteries make such a connection, while in others, only one coronary artery does so. Mediastinal vessels may join in the anomalous communication. These are represented by cut ends of vessels below the arrows.

d. Anomalous origin of one coronary artery from the pulmonary trunk while the other artery arises from the aorta. Displayed are communications between branches of the 2 arteries. It is through such communications that blood derived from the aorta is considered to enter the coronary artery that arises from the pulmonary trunk, and through the latter coronary artery, aortic blood is shunted into the pulmonary trunk.
such communication is described. One needs to recognize that the dramatic appearance of an aneurysm involving a coronary artery might draw the examiner’s attention away from a search for an additional (and underlying) lesion in the form of an abnormal communication. In some instances a ruptured aortic sinus aneurysm may be confused with an anomalous communication of a coronary artery with a cardiac chamber.

Some further remarks are pertinent concerning the anatomic details of those anomalies that involve communication between a coronary artery and the pulmonary trunk.

Anomalous connection of the coronary arterial system with the pulmonary trunk takes 1 of 2 forms. The less common form is that in which the 2 coronary arteries originate normally from the aorta and, in addition, an accessory coronary artery arises from the pulmonary trunk as described by Krause\(^1\) and in case 2 of Brooks\(^1\)\(^2\) report. The latter vessel makes gross communication with the proximal portions of one or both aortic coronary arteries, either directly\(^1\)\(^3\) or through a plexus of vessels situated at the anterior aspect of the base of the heart (fig. 1c).

It is now generally agreed that in these cases the abnormal communication is associated with an arteriovenous-like condition in which there is a left-to-right shunt from the coronary arteries to the pulmonary trunk. In some cases mediastinal arteries also join in the anomalous communication and in the shunt, a condition noted by Biörck and Crafoord\(^1\)\(^4\) and in case 2 by Brooks.\(^1\)\(^2\)

The commoner variety of anomalous communication between the coronary arterial system and the pulmonary trunk is the one usually classified as “anomalous origin of a coronary artery from the pulmonary trunk.” In this type of anomaly only 1 of the 2 coronary arteries arises from the aorta while the other arises from the pulmonary trunk (fig. 1d). Often each of the 2 arteries is wider than normal, and tortuous. The reader who is under the influence of prevailing teaching may readily wonder why this condition should be classed as an arteriovenous communication.

The associated myocardial ischemia has been documented electrocardiographically many times since the report in 1933 of Bland, White, and Garland.\(^1\)\(^4\) Nevertheless the cause of the myocardial ischemia has not met uniform recognition. The basis for disagreement revolves around the question, In what direction does the blood flow in the artery arising anomalously from the pulmonary trunk?

Currently what seems to be the view most commonly held is that blood flows from the pulmonary trunk into the anomalous coronary artery and thence to the myocardium. Indirect evidence for rejecting this theory is supplied by cases of cyanotic congenital cardiac disease. In some of these cases the level of oxygen saturation of the arterial blood is far below that of the venous blood of patients without right-to-left shunts. Yet cyanotic patients do not manifest clinical or pathologic signs of myocardial ischemia. Those who hold this view would answer this argument with the point that it is not so much the level of oxygen saturation but the low perfusion pressure supplied to the anomalous artery by the right ventricle and the pulmonary trunk that makes the myocardium ischemic.

My associate, Dr. H. B. Burchell, and I\(^1\)\(^5\) have been more attracted by the theory that blood flows from the anomalous coronary into the pulmonary trunk. In regard to the latter theory H. St. John Brooks,\(^1\)\(^2\) Demonstrator of Anatomy in Trinity College, Dublin, reported in 1886 on finding, in a dissecting room specimen (case 1), the origin of the right coronary artery from the pulmonary trunk while the left coronary artery arose normally from the aorta. Brooks noted that branches of the anomalously arising right coronary artery anastomosed, over the cardiac wall, with branches of the normally arising left coronary artery. His consideration of the direction of blood flow in the anomalous right coronary artery is significant and of historic importance. It is quoted as follows:

“A consideration of this case will show that a very interesting question is connected with it. Here are two arteries belonging to the different circulations—the pulmonary and the
systemic—anastomosing with each other. In these circulations, as is well known, the arterial pressure is very much greater in the systemic than in the pulmonary; how then did the blood flow in the anomalous coronary artery? There cannot be a doubt that it acted very much after the manner of a vein, and that blood flowed through it towards the pulmonary artery, and from thence into the lungs."

In 1927 Maude Abbott,16 in describing the case of a 60-year-old woman with origin of the left coronary artery from the pulmonary trunk, agreed with Brooks’ suggestion regarding direction of blood flow in the anomalous artery.

The following statements from several sources provide evidence that supports the view that blood in the anomalous artery is derived from the coronary artery that arises from the aorta and, further, that the blood then flows into the pulmonary trunk:

1. In both normal and anomalous arteries, anastomoses exist between branches of the 2 coronary arteries.
2. In the anomaly under consideration both coronary arteries have been observed to be extremely dilated and tortuous in a manner similar to what occurs in other situations in which arteriovenous communications are known to be present.
3. Clinical evidence of myocardial ischemia is usually not apparent until several months after birth, when the adult type of difference between systemic arterial and pulmonary arterial pressures becomes established. At this time the maximal tendency for flow of blood from the artery arising from the aorta into branches of the artery arising from the pulmonary trunk would be expected to become manifest.
4. Perfusion studies on postmortem specimens show evidence of free communication between branches of the 2 arteries17, 18 and, through these pathways, communication of the aorta with the pulmonary trunk.
5. A fortuitous observation was made by Apley and associates19 that when the coronary artery that arose from the pulmonary trunk was divided at operation, bright red blood flowed freely from its distal end.

Thus in anomalous origin of 1 coronary artery from the pulmonary trunk, not only theory but also observations are in strong support of the concept that there is run-off of blood from the normally arising artery into the anomalous one, and thence into the pulmonary trunk. Anomalous origin of a coronary artery from the pulmonary trunk, therefore, logically seems to be classed along with other anomalies in which an arteriovenous fistula-like condition exists and is undisputed.

JESSE E. EDWARDS

REFERENCES

5. JOHNSON, J.: Quoted by Davis and associates.6
8. MOWE, A. G.: Personal communication.
12. BROOKS, H. ST. J.: Two cases of an abnormal coronary artery of the heart arising from
pulmonary artery: With some remarks upon the effect of this anomaly in producing cirrhotic dilatation of the vessels. J. Anat. 20: 26, 1886.


Medical Eponyms

By Robert W. Buck, M.D.


“A systolic tug of the left false ribs posteriorly communicated by the diaphragm may be conspicuous. The recoil from the drag may be so distinct as to look and feel to the hand like pulsation, and in the first case in which I observed it, now more than 20 years since—a case of left empyema—it was taken for pulsation, and it was supposed that a pulsating tumour of some kind underlay the empyema. A post-mortem examination showed that the cause was adherent pericardium. I have often seen this tugging since, and in some cases it can be made to affect the right false ribs by causing the patient in the sitting position to lean over to the left so as to throw the drag of the heart upon the right half of the diaphragm. It must be added that this indication is not infallible, as the tugging has been observed when the heart was hypertrophied without adhesions.”
Editorial: Anomalous Coronary Arteries with Special Reference to Arteriovenous-like Communications
JESSE E. EDWARDS

_Circulation_. 1958;17:1001-1006
doi: 10.1161/01.CIR.17.6.1001

_Circulation_ is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1958 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/17/6/1001.citation

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in _Circulation_ can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to _Circulation_ is online at:
http://circ.ahajournals.org/subscriptions/