Correction of Shunt from Right Conal Coronary Artery to Pulmonary Trunk with Relief of Symptoms

By Gerald B. Lee, M.D., Fredrick L. Gobel, M.D., C. Walton Lillehei, M.D., Walter S. Neff, M.D., and Robert S. Eliot, M.D.

SUMMARY
A case of a small right coronary conal branch to pulmonary trunk shunt with surgical correction is reported. The patient, a 45-year-old housewife, had chest pain, labile hypertension, and intermittent left bundle-branch block with normal serum cholesterol and triglyceride levels. Selective coronary arteriography was necessary to demonstrate the abnormal communication between the conal branch of the right coronary artery and the pulmonary trunk. Symptoms disappeared after surgical correction of the shunt. Transient left bundle-branch block appeared during exercise after surgery, but the patient was improved subjectively and resumed all household chores without difficulty. Shunting of oxygenated blood away from the myocardium thereby decreasing coronary blood flow to a specific area may have been responsible for significant symptomatology.

Additional Indexing Words:
Coronary arteriovenous fistula
Coronary arteriography

Coronary arteriovenous fistulas are relatively uncommon and therefore account for few cases of myocardial ischemia in adults. Anomalous communications between the conus artery and the pulmonary trunk are even more unusual. When such a shunt is small, it may be overlooked clinically or at necropsy. That such an apparently insignificant shunt might bring about major manifestations of myocardial ischemia is of significant interest.

Case Report
A 45-year-old housewife was hospitalized in November 1965 with the initial episode of severe retrosternal chest pain. An electrocardiogram showed left bundle-branch block. Serial enzyme determinations did not suggest acute myocardial necrosis. The blood pressure was 160/85 mm Hg.

After discharge the electrocardiogram changed to normal intraventricular conduction but was suggestive of left ventricular hypertrophy. During a two-step exercise test left bundle-branch block reappeared transiently and was accompanied by chest pain and dyspnea. Because of increasing symptoms she was referred to the University of Minnesota Hospitals. The patient related that she was unable to perform her household duties because of chest pain.

She gave a 9-year history of labile hypertension. She stated that her menses were normal and regular. There was no known history of premature cardiovascular disease in the family although her mother also has hypertension. Physical examination revealed a slender white female. The blood pressure was 150/90, and the pulse was 82 and regular. The retinal vessels appeared normal. The neck veins were not distended, and the heart was normal in size. A grade II/VI early systolic murmur was audible at the left sternal border.

Laboratory data included a normal hemoglobin, white blood cell count and differential. A urine
culture grew *Escherichia coli* in significant numbers and then became sterile after treatment with tetracycline. Urinalysis, blood urea nitrogen, serum creatinine, LDH, blood glucose, and electrolytes were normal. Serum cholesterol was 124 mg%. The serum triglycerides were 108 mg%. The chest x-ray and cardiac fluoroscopy were normal. The electrocardiogram showed nonspecific ST-T changes (fig. 1). Vectorcardiogram revealed a QRS loop that was slightly increased in size but otherwise was normal.

**Coronary Arteriography**

Two days after admission, a bilateral selective coronary arteriogram was performed. The left coronary artery and major branches were normal (fig. 2). The right coronary artery (RCA) was widely patent and preponderant. A moderately large conal branch originated from the proximal portion of the RCA, deviated cephalad and to the left, and communicated with an accessory coronary artery from the pulmonary trunk (fig. 3).

**Figure 1**

The preoperative vectorcardiogram (VCG) shows a slight increase in QRS loop size. (System used was that of Schmitt-SVEC III.) An electrocardiogram (ECG) recorded on the same day shows nonspecific ST-T changes.
Figure 2
Selective arteriography of the left coronary artery demonstrates widely patent major branches. The left anterior descending (LAD) and left circumflex branch (LCB) both appear normal.

Figure 3
Left. Using the same view as the x-ray (lateral) (right panel), a sketch shows the right coronary conal branch to pulmonary trunk fistula. Right. The conal branch arises from the proximal right coronary artery (RCA) and joins an anomalous branch that communicates with the pulmonary trunk. Three carets are used to outline a faintly opacified pulmonary trunk.

Right and Left Heart Catheterization
Hemodynamic studies were done to evaluate the size of the shunt, to determine the presence or absence of failure, to document any associated anomalies, and to evaluate the patient’s response to exercise. An oximetry series failed to demonstrate an increase in oxygen saturation in the pulmonary trunk. Injection of 50 mg of indocyanine green dye into the left ventricle with sampling from the pulmonary trunk also failed to give clear-cut evidence of a shunt. Methyl 131I and Freon inhalation studies2, 3 with sampling from the pulmonary trunk failed to demonstrate a left-to-right shunt. Failure to detect a shunt by these techniques indicates that the shunt was probably less than 4% of the cardiac output. During exercise the patient was able to increase her cardiac output from 4.8 to 7.1 liters per minute, with a rise in oxygen consumption from 183 to 425 cc O2 per minute. The pressures in all four chambers were normal at rest and during exercise. The patient had no chest pain during the procedure. The electrocardiogram during the entire procedure showed left bundle-branch block (fig. 4). Catheterization then con-
CORONARY ARTERY TO PULMONARY TRUNK SHUNT

Figure 4

Electrocardiogram during the time of heart catheterization showed persistent left bundle-branch block (record was taken at a paper speed of 25 mm per second).

firmed the facts that the left-to-right shunt demonstrated angiographically was small, that there was no failure, that there were no associated anomalies, and that the exercise response was normal.

Surgery

One week following catheterization a left thoractomy was performed. The communicating vessel between the conal branch of the RCA and the pulmonary trunk measured 2 to 3 mm in diameter. After transection of this vessel, pulsatile bright red blood came from the conal branch, whereas venous blood appeared from the portion communicating with the pulmonary trunk. There were no complications in the postoperative period.

During the 8 months following operation she had no recurrence of chest pain and resumed all her household chores without difficulties. The blood pressure remained labile but averaged 165/100 mm Hg. The murmur at the left sternal border could no longer be heard. Her resting electrocardiogram showed nonspecific ST-T changes. A left bundle-branch block appeared transiently during a two-step exercise test, but dyspnea and chest pain were not present.

Discussion

A shunt from the right conal branch of the RCA to the pulmonary trunk is another unusual cause of nonatheromatous myocardial ischemia. The true incidence of this shunt is unknown but it is probably uncommon. Such a shunt is clinically elusive, as there may be no characteristic historical or physical findings. In the case presented, the symptoms were suggestive of angina pectoris, and the murmur appeared innocent in type. The laboratory studies may also be noncontributory as evidenced in this case by normal serum cholesterol, serum triglycerides, and myocardial enzymes. Because of its small size, the left-to-right shunt may be readily missed at routine heart catheterization. Aortic root arteriography probably would not lead to adequate visualization of such a small vessel because of inadequate filling of the coronary arteries. Lack of awareness of the condition combined with its small size may lead to its being completely overlooked at necropsy.

Normally about 4 to 6% of the cardiac output is diverted to the coronary arterial system. We do not know the actual blood flow through this anastomosis or how much the remaining vascular bed was compromised in this patient. The fact that the shunt was small, however, was documented by cardiac catheterization, by coronary arteriography, and by direct visualization of the small vessel at the time of surgery. That such a small shunt could produce such dramatic symptoms is provocative. One may only speculate that oxygenated blood was intermittently diverted away from a portion of myocardium and from the left bundle branch. Studies by James and Schlesinger have shown that the right coronary artery—when preponderant—does supply the proximal few millimeters of the left bundle by terminal branches of the A-V node artery.

In the case reported, postoperative exercise caused a transient reappearance of left bundle-branch block. Furthermore, subjective relief of anginal pain following operation cannot be considered proof of the importance of the shunt. The possibility of “small vessel disease” always exists among hypertensive patients. When small vessel disease does exist, a small shunt might have a more significant and deleterious hemodynamic effect than in the normal coronary vascular bed. It is possible that pathological changes in the conduction system had become fixed or that disease in the coronary vessels was present but unrecognized.

In approximately half of the specimens examined by James and Schlesinger and associates, the conus artery was a direct
branch of the aorta; in these cases the conus artery ostium was located usually near the ostium of the RCA. In the remainder of the specimens the conus artery was the first ventricular branch of the RCA (conal branch of RCA). Regardless of the origin, the conus artery normally supplied the conus arteriosus and, in addition, anastomosed with a branch of the left coronary artery to form VIEUSSENS' ring.\(^8\) We were unable to visualize this ring by selective coronary arteriography. It is possible that, in early development of the heart, anastomosis occurred between the conal branch of the RCA and an accessory coronary artery arising from the pulmonary trunk.

A review of the literature failed to reveal a case of an extensively studied and surgically corrected conal artery to pulmonary trunk shunt. In most reported cases, the anomalous coronary artery is found to enter one of the four cardiac chambers, the pulmonary trunk,\(^1\),\(^7\) or the bronchial circulation.\(^9\) The vessel is usually enlarged and elongated, with focal saccular aneurysms. An increase in oxygen saturation in the chamber receiving the anomalous vessel has been reported.\(^1\) In 1958, Edwards and associates\(^1\) described at necropsy the communication of a branch of the right coronary artery with the right atrium. In 1960, Amplatzt and associates\(^1\) demonstrated by aortography a left coronary artery to main pulmonary artery fistula. In 1961, Neufeld and associates\(^1\) summarized six cases of "coronary artery fistula" diagnosed at the University of Minnesota Hospitals; two of the six had anomalous right coronary arteries that terminated in the right atrium and right ventricle. These authors described six previously studied cases in the literature with anomalous termination of the RCA into the pulmonary trunk. In four of the six cases reported by Neufeld, there was an increase in oxygen saturation in the cardiac chamber in which the anomalous vessel terminated. In no case reported is there a description of a communication between an aberrant artery between the pulmonary trunk and the conal branch of the RCA demonstrated by arteriography.

**Conclusion**

It is difficult to be certain that the patient's symptoms were related to the small shunt demonstrated by arteriography and at surgery. Relief of chest pain postoperatively is common even in patients who have had shunt operations for coronary artery disease. The left bundle-branch block could well be due to undiagnosed small vessel disease or secondary to systemic hypertension. It is possible that a specific portion of cardiac tissue was intermittently "robbed" of oxygenated blood that was diverted to the pulmonary circulation.

The risk of surgical correction of the shunt described is that of thoracotomy alone. When the shunt is larger and leads to cardiac failure the hazard is, of course, greater. The relief of myocardial ischemia by a corrective operation warrants such a risk.

**References**

Correction of Shunt from Right Conal Coronary Artery to Pulmonary Trunk with Relief of Symptoms

GERALD B. LEE, FREDARICK L. GOBEL, C. WALTON LILLEHEI, WALTER S. NEFF and ROBERT S. ELIOT

Circulation. 1968;37:244-248
doi: 10.1161/01.CIR.37.2.244

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1968 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/37/2/244

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/