Coronary Arteriovenous Fistula Between the Left Coronary Artery and Perisistent Left Superior Vena Cava Complicated by Bacterial Endocarditis *

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RECENT REVIEWS of coronary arteriovenous fistulas [1, 15] attest to the increasing frequency of recognition of this lesion. The clinical manifestations are often compatible with other defects producing continuous murmurs. Indeed, the first successful surgical correction of this malformation by Bjork and Crafoord in 1947 [3] was in a patient explored with the preoperative diagnosis of patent ductus arteriosus. However, the development of retrograde aortography and improved catheterization technics have provided the methods whereby one may obtain an accurate preoperative diagnosis.

To date, some 30 patients have been operated upon and, although the majority were asymptomatic, cardiac enlargement, congestive heart failure and bacterial endocarditis have been documented.

The present report illustrates the confusion this lesion may produce and also documents a previously undescribed site of venous communication, a persistent left superior vena cava.

Case Report

J. B., GNH No. 44-11-67, a 35-year-old woman, was first admitted to the Yale-New Haven Medical Center on January 15, 1956, because of increasing fatigability and shortness of breath. The presence of heart disease was first suspected three years prior to admission when a murmur was detected during the course of her first pregnancy. This pregnancy continued to term without incident and a second pregnancy the following year was also successfully concluded.

One year before admission, she noted the onset of intermittent fever, for which she was eventually hospitalized. At that time, a diagnosis of subacute bacterial endocarditis was confirmed by multiple blood cultures positive for Streptococcus viridans. The infection was successfully treated with antibiotics and she was discharged. Digitalis and prophylactic penicillin were prescribed.

In retrospect, she realized that she had always tired easily and that this symptom had progressed in recent years. At the age of five she developed pneumonia and subsequent left empyema which required thoracostomy tube drainage. She denied any history of rheumatic fever, cyanosis, orthopnea, hemoptysis, chest pain or peripheral edema.

On physical examination the pulse was 78 and regular, the blood pressure 120/70 and she appeared as a well-developed female in no distress. The only pertinent physical findings were limited to the chest. A Grade III/VI continuous murmur was heard in the second and third intercostal spaces along the left sternal border. There was also a soft, blowing systolic murmur audible at the cardiac apex. A roentgenogram of the chest (Fig. 1) demonstrated minimal cardiomegaly with definite dilatation of the ascending aorta. The lung fields were clear with normal vascular markings. The electrocardiogram revealed abnormally depressed ST segments in leads 1, 2, AVF and V_3 through V_5. The T-waves were abnormally low or biphasic in leads 2 and V_4 through V_6.

On January 24, 1956, a left thoracotomy was performed with a preoperative diagnosis of patent ductus arteriosus. A persistent left superior vena cava was found over which a thrill was thought to be maximum. Temporary occlusion of this vessel had no effect upon the intensity of the thrill,

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and division of the ligamentum arteriosus produced no change in the continuous thrill. With the use of a sterile stethoscope it was thought that the murmur was maximum over the pulmonary artery. This observation was interpreted to represent a probable aortico-pulmonary window and because of the left thoracotomy approach no further dissection was performed. The postoperative course was uneventful and two months later on April 29, 1956, right heart catheterization and angiocardiography were performed. The catheter was passed from the left antecubital vein into the right superior vena cava. An oxygen step-up from 74 per cent saturation in the right superior vena cava to 85 per cent in the right atrium was noted. The arterial oxygen saturation, breathing room air, was 96 per cent (Table 1). A venous angiocardiogram confirmed the presence of the persistent left superior vena cava, dilatation of the ascending aorta and enlargement of the right atrium (Fig. 2). These findings were interpreted as probably due to anomalous pulmonary venous connection to the left superior vena cava draining into the coronary sinus, although the continuous murmur was unexplained.

TABLE 1. *Right Heart Catheterization—April 29, 1956*

Catheter Position	Blood O_2 Content vpc.	Sat. %	Pressures
SVC	14.52	74.1	
RA—mid.	14.26	72.8	5/2
RA—outflow	16.71	85.2	
RV—inflow.	15.50	79.1	22/1
RV—outflow.	15.48	79.0	
PUL. Artery	15.79	80.5	20/7
Arterial Blood:			
Femoral Artery	18.99	96.3	126/70
After Breathing O_2	20.77	103.8	

For the next three and a half years she was lost to follow up. However, because of increasing fatigue and inability to carry out her normal activities, she again presented herself for further studies in September of 1959. An attempted cardiac catheterization was terminated because of extreme cardiac irritability and a severe coughing episode. She again was discharged and followed for three years with essentially no symptoms. However, in early 1962, she noted the onset of intermittent fever, easy fatigability, and palpitations. In April 1962 a retrograde aortogram demonstrated a large, tortuous coronary artery communicating with the left persistent superior vena cava, with progressive opacification of the coronary sinus and right atrium (Fig. 3).

In September of 1962, she was admitted for

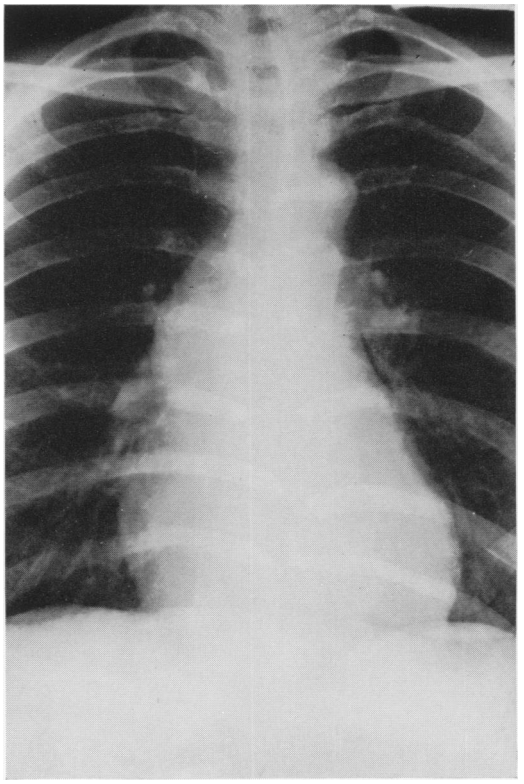

FIG. 1. Roentgenogram of the chest demonstrating the dilatation of the ascending aorta.

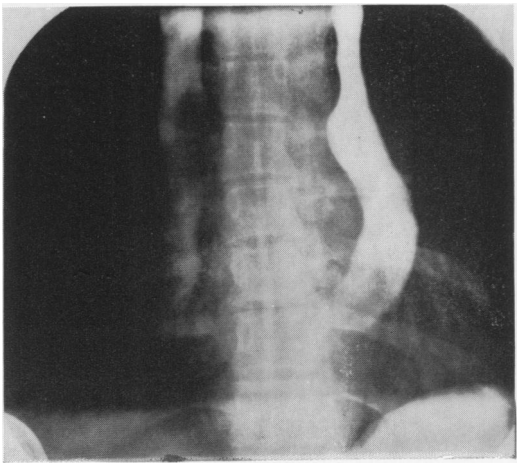

FIG. 2. Venous angiocardiogram with opacification of both right and left superior venae cavae.

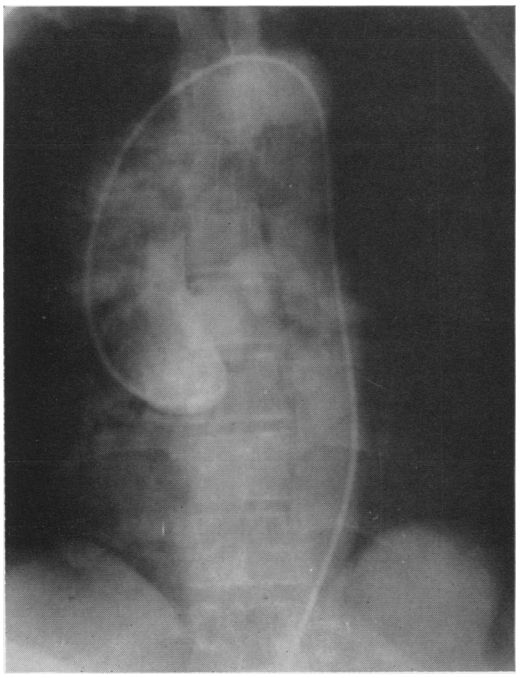

FIG. 3. Retrograde aortogram demonstrating the dilated and tortuous left coronary artery.

definitive operation. On the first hospital day she developed a fever of 38° C. which persisted for over five days. Multiple blood cultures were sterile, but because of the past history of subacute bacterial endocarditis it was felt that operation should be temporarily deferred.

She was readmitted one month later, and on October 30, 1962, operation was carried out. The chest was entered through the fifth left intercostal space. On opening the pericardium, the large anomalous coronary artery was immediately apparent just posterior and inferior to the tip of the left atrial appendage (Fig. 4). The continuous thrill was maximum at the junction of the persistent left superior vena cava and the anomalous coronary artery. After dissection of the fistula it became obvious that it was a branch of the circumflex division of the left coronary artery about 8 or 9 mm. in diameter and was in itself an end artery (Fig. 5). Temporary occlusion of the fistula resulted in a marked Nicoladoni-Branham sign but no change in the electrocardiogram. The fistula was divided and the ends oversewn with 5-0 silk. The postoperative course was uneventful and she was discharged three weeks later. She has now been followed for 20 months and remains well, with a marked increase in exercise tolerance.

Discussion

The recognition of a coronary arteriovenous fistula is dependent upon a high degree of suspicion. Except for rare reports, the venous drainage of the fistula has been into the right side of the heart.[1, 15] This presents the typical manifestations of a left-to-right intracardiac shunt associated with a continuous precordial murmur. The continuous murmur usually associated with this lesion is easily confused with a patent ductus arteriosus, as it was in this patient. Gasul[6] has emphasized the use of phonocardiography as a diagnostic tool, stating that "phonocardiographic demonstration of

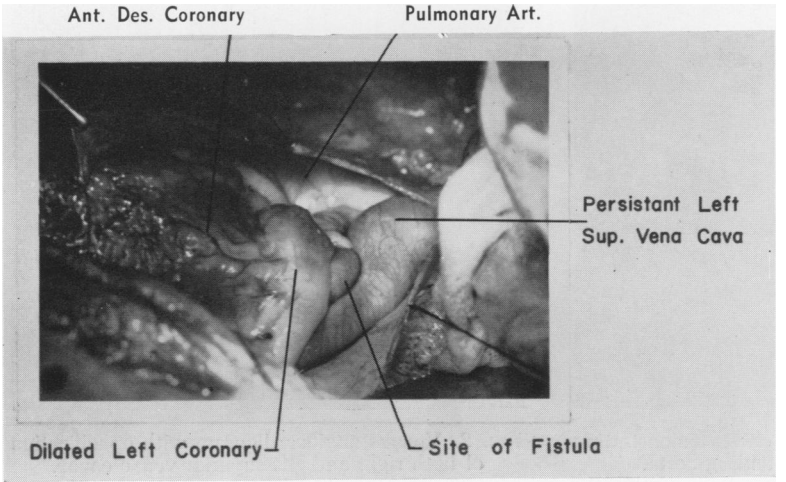

FIG. 4. Photograph demonstrating the relationship of the involved structures.

a louder diastolic than systolic component is very suggestive of a coronary arteriovenous fistula communicating with the right ventricle." However, as pointed out by Abbott,[1] in a review of the literature, the murmur is occasionally indistinguishable from a patent ductus, even by phonocardiography.

The conventional chest roentgenogram may be entirely normal unless there is a significant left-to-right shunt. Even in the presence of a large shunt, however, the changes associated with increased pulmonary flow or chamber enlargement are of no specific diagnostic importance. Dilatation of the ascending aorta is extremely common, however, as pointed out by Gasul,[6] Bjork[3] and Knoblich,[7] and was the only significant abnormality noted in our case.

The electrocardiogram also is of limited value as a diagnostic tool. Changes of ischemia or ventricular strain pattern may suggest which coronary artery is involved and certainly should be considered as an indication for surgery.

Cardiac catheterization, although expected to confirm the left-to-right shunt, may be misleading. The data obtained is usually nonspecific and entirely consistent with more common lesions producing left-to-right shunts. As in our patient, the small shunt detected was originally considered due to anomalous pulmonary venous connection to the persistent left superior vena cava.

The definitive diagnostic tool is unquestionably retrograde aortography. The supravalvular injection of contrast media should in every instance opacify the dilated and usually tortuous coronary artery and often delineate the site of venous communication.

The prognosis of untreated coronary arteriovenous fistulae is largely unknown, but prolonged and useful life appears common. Taussig, quoted by Nadas,[9] studied 13 patients at autopsy, nine of whom had lived over 53 years. A patient reported by Yenel[16] died at 75 years of age with congestive heart failure and recurrent chest pain. At autopsy, a typical arteriovenous fistula between the right coronary artery and coronary sinus was found. The oldest and even more remarkable patient was described by Scott,[12] dying at the age of 84.

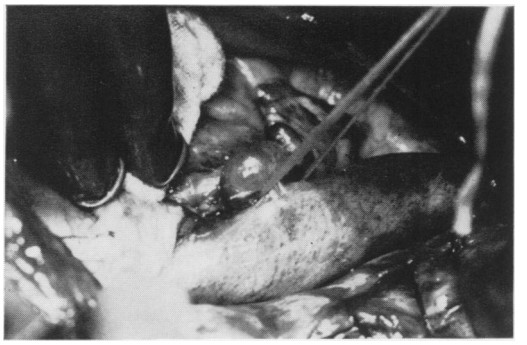

FIG. 5. Appearance of the operative field after dissection of the fistula with an umbilical tape around the site of communication.

It is not surprising to find bacterial endocarditis as a complication of untreated coronary arteriovenous fistula. Indeed, a review of the literature reveals that our patient is the sixth example of this complication among the 81 reported cases.[5, 11, 13, 14] As suggested by Gasul, we would agree that the risks of bacterial endocarditis alone justifies the surgical correction of symptomatic coronary arteriovenous fistula.

The surgical management of this lesion ideally consists of obliteration of the fistulous communication while preserving normal coronary arterial flow. Ligation of the coronary artery may produce significant myocardial ischemia and two such patients have been described by Cooley.[4] Michaud has described a death following division of a coronary artery proximal to the terminal branches in an infant with a coronary artery-right ventricular fistula.[8] The end artery character of the feeding vessel in our patient permitted this objective to be achieved. As noted by Abbott,[1] however, there are only three other patients in which

this was done. It is of interest that of the 35 reported surgical corrections, only three patients failed to survive.[2, 4, 8]

Summary

A case of a congenital arteriovenous fistula between the left coronary artery and a persistent left superior vena cava is reported. The complication of subacute bacterial endocarditis has been documented and the diagnostic confusion that this lesion may produce is well illustrated. In addition, this is the first report of a coronary arteriovenous fistula involving a persistent left superior vena cava.

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