Congenital Cirsoid Aneurysm of a Coronary Artery with Associated Arterio-atrial Fistula, Treated by Operation: A Case Report *

HERSCHEL E. MOZEN, M.D.
Cleveland, Ohio

From the Department of Surgery, Western Reserve University School of Medicine and the University Hospitals of Cleveland

INTRODUCTION

CONGENITAL anomalies of the coronary arteries are unusual and congenital aneurysms of these vessels are especially rare. Scott,[9] reviewing the literature on all aneurysms of the coronary arteries, reported a total of 47 cases. He classified them as congenital, mycotic-embolic, arteriosclerotic and syphilitic. He believed that 15 of the reported cases were congenital in origin. These congenital aneurysms were further classified as localized or diffuse. The localized ones were either saccular or fusiform, whereas the diffuse ones were dilated and tortuous for a distance of several centimeters along the involved vessel.

Since Scott's review, four additional case reports of an aneurysm of a coronary artery have appeared.[1, 7, 8, 10] This present case is the fifty-second one to be reported. It is the first case in which the diagnosis was made at operation. In all the other cases, the diagnosis was made at autopsy.

The occurrence of arterio-atrial or arterio-venous fistulae, accompanying these aneurysms, has been noted previously. Harris,[6] Essenberg [2] and Halpert [5] each have reported such a lesion. The present case is the first one in which such a fistula has been cured by operation.

CASE REPORT

G. D. K., a 10-year-old white boy, was admitted to the University Hospitals of Cleveland with a chief complaint of "heart murmur." The murmur had been present since infancy. Growth and development had been normal. There was no history of neo-natal distress, cyanosis, hemoptysis or edema. Except for slight dyspnea on extreme exertion, the child was asymptomatic. He was considered to be "high-strung and nervous."

He was well-developed and well-nourished. The blood pressure varied between 120/80 and 130/60. The pulse rate was 84 per minute. The peripheral pulses were normal. There was no abnormal precordial activity. There was a soft systolic thrill over the apex of the heart. The heart size was normal. A loud, continuous to-and-fro murmur with systolic accentuation was heard in the pulmonic area. This murmur radiated along the left border of the sternum. It also was heard in the left supra-clavicular area. A lower-pitched systolic murmur also was heard at the apex. P_2 was normal.

Blood and urine studies were normal. The electrocardiogram indicated a vertical heart. The right ventricle was questionably enlarged on cardiac fluoroscopy. There was no "hilar dance" of the pulmonary arteries. There was questionable diminution of the peripheral pulmonary vascular markings. Cardiac catheterization was performed and was technically satisfactory. Pressures and oxygen concentrations in the pulmonary artery, right ventricle, right atrium and venae cavae were normal. No diagnosis of cardiac abnormality could be made.

Operation was performed on August 5, 1955 by Dr. Claude S. Beck. The mediastinum was explored first. A patent ductus was not found. The ligamentum arteriosum was ligated and divided. Further palpation revealed a harsh, continuous thrill over the base of the heart. When the pericardium was opened, a tremendously dilated and tortuous vessel was found overlying the interventricular septum in the usual location of the anterior descending branch of the left coronary artery (Fig. 1). The aneurysmal dilatation extended for a distance of approximately 5 to 6 cm. from the base of the heart towards the apex. This abnormal vessel varied from 0.5 to 1.3 cm. in diameter. In many places, large branches disappeared abruptly

* Submitted for publication January, 1956.

into the myocardium of the left ventricle. One branch, which was 1.5 cm. in length and 0.5 cm. in diameter, appeared to empty directly into the left atrium. Compression of this vessel obliterated the harsh thrill. Evidently this was a fistula which was carrying large amounts of blood from the high pressure aneurysm into the low pressure left atrium. The fistula was doubly ligated. The aneurysm immediately became smaller in diameter. The thrill disappeared completely. The pulse pressure became narrower. The heart was observed for 20 minutes and no cardiac irregularities appeared. The electrocardiogram remained unchanged. The pericardium and chest wall were closed in the usual manner.

The postoperative course was uncomplicated. The murmurs disappeared and the heart sounds were normal. The child was discharged from the hospital on the eleventh postoperative day. The heart sounds have remained normal.

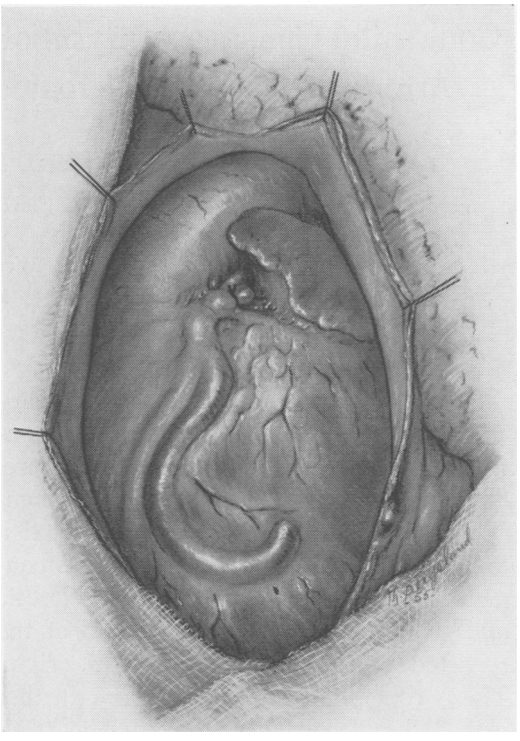

FIG. 1. The heart at operation, with the pericardium opened. The dilated and tortuous aneurysm of the coronary artery lies over the anterior surface of the heart. Two silk ligatures occlude the arterio-atrial fistula.

DISCUSSION

This case is an example of a rare congenital anomaly of the coronary arteries. The underlying embryologic deviations which cause such abnormalities remain obscure. Forbus [3] has studied the development of the cerebral and coronary arteries. He believes that congenital dilatations which occur at vascular bifurcations are due to a lack of elastic tissue in these regions. Grant [4] has described "inter-trabecular" spaces within the myocardium of the developing embryo. An abnormal coalescence of these spaces could result in a large, dilated and tortuous coronary artery as well as an abnormal drainage into a cardiac chamber.

The possibility of making a preoperative diagnosis of this lesion is considered. The harsh, continuous to-and-fro murmur in the pulmonic area could be due to a patent ductus arteriosus. However, the presence of an additional apical systolic murmur should suggest a different diagnosis. The slight widening of the pulse pressure and actual decrease in the pulmonary vascular markings are against the diagnosis of patent ductus. Normal findings during a satisfactory cardiac catheterization eliminate the possibility of any significant aortico-pulmonary artery communication. Some other type of arteriovenous fistula should be considered.

SUMMARY

Aneurysms of the coronary arteries, particularly the congenital ones, are rare.

The fifty-second case of an aneurysm of a coronary artery is reported. This is the first time that such a lesion has been recognized at operation. This is also the first case in which an associated arterio-atrial fistula has been cured by operation.

The possibility of making a correct preoperative diagnosis is considered.

BIBLIOGRAPHY

1. Colbeck, J. C. and J. M. Shaw: Coronary Aneurysm with Arterio-Venous Fistula. American Heart Journal, **48**: 270, 1954.

2. Essenberg, J. M.: An Anomalous Left Coronary Artery in a Human Foetus: Its Passage through the Left Atrium and Possible Discharge into the Right Atrium. Anatomical Record, **108**: 709.
3. Forbus, W. D.: On the Origin of Miliary Aneurysms of Cerebral Arteries. Bulletin of the Johns Hopkins Hospital, **47**: 239, 1930.
4. Grant, R. T.: Development of Cardiac Coronary Vessels in Rabbits. Heart, **13**: 261, 1926.
5. Halpert, B.: Arterio-venous Communication Between the Right Coronary Artery and the Coronary Sinus. Heart, **15**: 129, 1930.
6. Harris, P. N.: Aneurysmal Dilatation of the Cardiac Coronary Arteries. Review of the Literature and Report of a Case. Amer. Journal of Pathology, **13**: 89, 1937.
7. Rukstinat, G. J.: Multiple Aneurysm of the Right Coronary Artery. J. A. M. A., **149**: 1129, 1952.
8. Sarkisian, S.: Aneurysm of the Coronary Artery. United States Armed Forces Medical Journal, **1**: 1281, 1950.
9. Scott, D. H.: Aneurysm of the Coronary Arteries. American Heart Journal, **36**: 403, 1948.
10. Tellem, M. and A. T. Rubenstone: Aneurysm of the Left Coronary Artery. American Journal of Medicine, **19**: 652, 1955.