

ANEURYSMAL DILATATION OF THE CARDIAC
CORONARY ARTERIES *

REVIEW OF THE LITERATURE AND REPORT OF A CASE

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Except for small accessory coronary vessels, anomalies of the coronary arteries unassociated with abnormalities of the heart or great vessels are uncommon. In 1922 Plaut¹ reported a case of absence of the right coronary artery, and in 1930 Petré² reported a 2nd case. Several other cases of "absence" of a coronary artery have been reported, but in all the course of the "absent" vessel was at least approximated by a branch of the other. These cases have been discussed by Petré,² and by Bland, White and Garland.³ Of interest in this connection are the subsequently reported cases of Born⁴ and Hall.⁵ In Born's Case I both arteries arose from the right coronary ostium, but shortly after its origin the left artery sank into the interventricular septum and later emerged and followed its normal course. In Hall's case the right coronary artery was short and imperforate.

Origin of the left circumflex branch from the right coronary artery or from the right aortic sinus of Valsalva has been recorded with sufficient frequency to warrant its consideration as a distinct entity. Antopol and Kugel⁶ reported 4 such cases, Born mentions 2 (Cases II and III), and Plaut 1. In Plaut's case the right and posterior aortic cusps were replaced by a single large cusp, the right coronary artery arose from the normal site, and the left circumflex branch arose in the middle of the large sinus.

Of more importance are those cases in which one of the two coronary arteries arises from the pulmonary artery. Bland, White and Garland found reports of 8 such cases and added another. Additional cases have been described by Sanes and Kenny,⁷ Bartsch and Smekal,⁸ Kockel,⁹ and Mönckeberg.¹⁰ Wölffhugel¹¹ described a beef heart in which the right coronary artery arose from the pulmonary artery. In only 2 of the human cases (those of Mönckeberg¹⁰ and Schley¹²) has the right coronary artery been the anomalous one, and in neither was there damage of the myocardium supplied by the

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anomalous vessel. Mönckeberg's case was that of a 30 year old male who died of a fractured skull; Schley's was that of an 81 year old male. Nine of the other human cases form a distinct group, characterized among other things by death within the 1st year of life, and degeneration and fibrosis of the left ventricular myocardium. The remaining 2 cases are those of Abbott¹³ and Kockel.⁹ Abbott's case was that of a 60 year old female. The right coronary artery had a thick wall and the vessel and its ostium were much dilated. The left coronary artery arose from the pulmonary artery but had the normal distribution. Its wall was thin and Abbott thought the circulation might have been toward the pulmonary artery instead of away from it. Kockel's case was that of a 38 year old male who had had symptoms of heart disease for 10 years and who died suddenly. The left coronary artery was dilated and arose from the pulmonary artery. The left ventricular myocardium showed no degenerative changes. Bland, White and Garland mention 3 instances in which an accessory third coronary vessel arose from the pulmonary artery.

In 1929 Packard and Wechsler¹⁴ collected from the literature 29 cases of aneurysm of the coronary arteries, and described another. These were nearly all of arteriosclerotic or infectious origin. In 1934 Snyder and Hunter¹⁵ collected the subsequently reported cases and added 1 of syphilitic aneurysm.

An interesting, but heterogeneous, group of cases is characterized by dilatation of one or more branches of the coronary arteries. Of these, 7 have occurred in cattle and 4 in man. In some the vessels were cirroid. Although none of these cases is identical with the case to be described, they are of sufficient similarity to warrant consideration. In 5 of the cattle hearts the dilated vessel communicated with a ventricle (right 4, left 1), but all the animals were healthy.

In Schauder's¹⁶ case, that of a 6 or 7 year old ox, the left coronary artery and its ostium were much enlarged. The circumflex branch and radicles of the anterior descending branch were normal. The anterior descending branch was much dilated and soon left its normal course to enter a multilocular saccular structure measuring 7.5 by 3.8 by 4.2 cm., situated in the interventricular septum. There was no communication with either ventricle. The dilated vessel had a good internal elastica. The wall was thickened, mainly by an increase of the medial musculature, although elastic tissue was also

increased and formed a many layered external elastica. The internal elastica of the sac was poorly defined. The media contained elastic tissue, but little muscle or collagen. Masses of cartilage were present, displacing and compressing the elastic tissue. The wall of the dilated main stem near the aorta was twice as thick as normal, but the constituent elements were present in the normal proportion.

In Reid's¹⁷ case the animal was a 3 year old ox. The left coronary artery and its ostium and anterior descending branch were dilated. The circumflex and other branches of the dilated vessel were normal. The descending branch terminated just above the apex of the heart in a roughly conical, cyst-like structure with base outward and apex on a level with the inner surface of the left ventricle. The wall over the base of the cyst was exceedingly thin. The cyst communicated with the left ventricle by a circular aperture guarded by a valve-like structure composed of an inner fibrous ring 15 mm. in diameter, and an outer fibrous ring 35 mm. in diameter. The rings were connected by a fibrous tissue membrane thickened by several bands running radially between the rings. The wall of the dilated vessel was composed mainly of collagenous tissue with but little muscle and elastic tissue in the media.

Schöndube¹⁸ described the heart of a 7 year old cow in which there was an accessory ventricle formed by a transverse septum across the apical portion of the right ventricle. This contained an opening 4 mm. in diameter. The left coronary ostium and both main branches of the artery were much dilated. The vessels had the usual course and gave off normal branches. At the apex of the heart each vessel showed a region of stenosis and then emptied into a space, which in turn opened into the accessory ventricle. All three layers of the walls of the enlarged arteries were hypertrophic and resembled those of a normal, large systemic artery.

Rubli's¹⁹ case was that of a 62 day old calf. The left coronary ostium was enlarged and situated above the cusp. The diameter of the left coronary artery diminished rapidly, and the vessel gave off normal anterior and circumflex branches. Two cm. from its origin the circumflex branch gave off an anomalous vessel 14 mm. in diameter which, after several angulations and a horseshoe bend near the apex, ascended the interventricular septum posteriorly, and 2 cm. from the coronary sulcus disappeared into the septum, only to bend again and open into the right ventricle 5 cm. above the apex. This

dilated vessel gave off two normal branches which anastomosed with branches of the anterior descending branch. The first portion of the left coronary artery showed increase of elastic tissue at the expense of muscle. The dilated branch showed increase of elastica in the adventitia and less than the normal amount of muscle in the media. The internal elastic membrane was missing except at the origin of the vessel, and here it was incomplete. The right coronary artery was normal except for its origin craniodorsal to the left artery and above the anterior margin of the left posterior cusp. Such displacement of the right coronary ostium has been recorded by several authors and explained by Geipel²⁰ as depending on the spiral rotation which occurs in the truncus arteriosus and its bulbar swellings.

Joest²¹ described an aneurysm of the left coronary artery of a calf's heart communicating with the right ventricle, and Raschke²² described a cow's heart with an aneurysm of the right coronary artery opening into the right ventricle.

Schlegel²³ described a case of widespread aneurysmal dilatation of both coronary arteries and their branches in a beef heart. The arteries were in places thick walled and narrow, in other places thin walled and dilated, and contained saccular outpouchings. Anastomoses were seen between the vessels, but not in communication with the ventricles. The thickened vessels contained an abundant muscular layer.

Trevor's²⁴ case was that of an 11 year old female who died of streptococcus septicemia. Five days before death a to-and-fro murmur appeared over the precordium. The right coronary ostium was 1 cm. in diameter and the artery was dilated. The descending branch formed a thin walled aneurysm lying within the right ventricular wall. This communicated with the ventricle by a ragged opening 0.5 cm. in diameter, the margins of which were covered by blood clot. The aneurysm was undoubtedly congenital and rupture probably occurred when the murmur appeared.

Halpert²⁵ described an interesting arteriovenous communication in the heart of a 54 year old male who died of carcinoma of the stomach. The coronary sinus was dilated to a maximum diameter of 2.5 cm. and was connected to the right coronary artery by a vascular loop 1 cm. in diameter which began at the posterior longitudinal sulcus. The right coronary ostium was 5 mm. in diameter. The right coronary artery was 1.5 to 2 cm. in diameter, elongated and

tortuous. The wall showed atheromatous change with focal calcification. The vessel gave off a normal posterior descending branch and several smaller, apparently normal branches. Microscopically the anastomotic loop showed a structure intermediate between that of an artery and of a vein.

Löwenheim's²⁶ case was that of a 62 year old female who had had cardiac symptoms for 2 years. The blood pressure was 260/100. The heart was uniformly enlarged and weighed 835 gm. The coronary orifices were normal. Two cm. from the aorta the right coronary artery suddenly dilated to a vessel 5 cm. in circumference, which gave off normal branches. At the interventricular septum posteriorly it ended abruptly as a sac with a narrow connection to a similarly dilated 4 cm. segment of right coronary vein. The dilated part of the vein also terminated abruptly and had normal branches. The layers of the walls of both vein and artery were thickened, but there was an irregularity of distribution of the components, particularly of the muscle. There was no evidence of inflammatory changes.

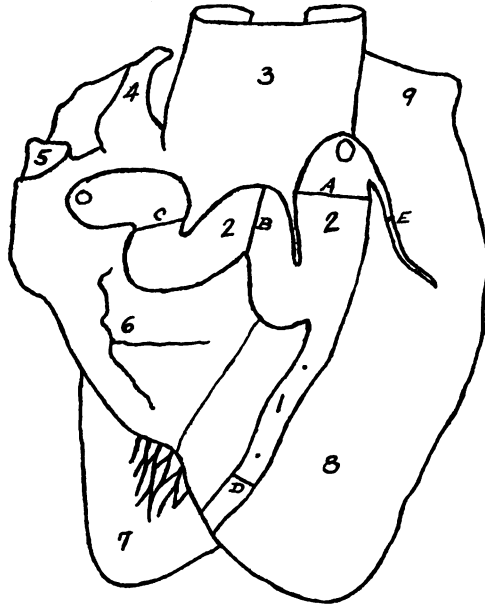
In Kockel's⁹ previously mentioned case the right coronary artery and both main branches of the left were greatly dilated and tortuous. The ostium of the right was 6 mm. in diameter. Immediately beyond the ostium the diameter of the vessel was doubled. The left arose from the pulmonary artery and had an ostium 5 by 10 mm. in diameter. The vessels had the normal distribution and did not anastomose with each other or empty directly into veins.

REPORT OF CASE

Clinical History: The patient was a 43 year old white male who died of hemorrhage into a brain tumor. No cardiac symptoms had been noted.

Autopsy Report: The heart weighed 380 gm. and the right side was much dilated. The left coronary artery showed moderate arteriosclerosis and the circumflex branch supplied less than the usual proportion of the posterior left ventricular wall. The left coronary ostium was 4 mm. in diameter and the right coronary ostium 5 mm. in diameter. Immediately after its origin the right coronary artery dilated to a circumference of 2 cm. The first branch of the vessel was slightly dilated. The second branch was dilated to a circumference of 2 cm. and with the dilated main stem of the artery formed a tortuous vessel of uniform caliber 13 cm. long, which ran

between the right auricular appendage and the aorta. After an abrupt diminution of caliber at its very end, 8 cm. from the aorta, it terminated in the right auricle through a 1 mm. opening in a soft nodule 4 mm. in diameter, which lay 1.5 cm. superior to the upper



TEXT-FIG. 1. Diagram of anterior superior surface of heart showing right coronary artery. The letters A to E indicate situations from which were taken the sections illustrated in the plate.

- 1 = main branch of right coronary artery
- 2 = dilated portion of the artery
- 3 = aorta
- 4 = tip of left auricular appendage
- 5 = superior vena cava
- 6 = tip of right auricular appendage
- 7 = left ventricle (internal surface)
- 8 = right ventricle
- 9 = pulmonary artery

margin of the foramen ovale. At a point 2.8 cm. from the aorta the main stem of the vessel continued in its normal course as an artery 1 cm. in circumference which, because of the difference in caliber, appeared to be merely a branch of the dilated sinuous vessel. Its branches were normal and it supplied somewhat more than the usual portion of the posterior left ventricular wall. A few atheromatous

plaques were present in both normal and dilated portions of the artery. The variation from the usual distribution of the terminal portion of the left circumflex and right coronary arteries was no greater than is seen in about 20 per cent of all human hearts.

Microscopic Examination: Microscopically the myocardium is not remarkable. The intima of the left coronary artery shows atherosclerosis with focal calcification. The internal elastic lamella is reduplicated and in places is broken. The elastic tissue and muscle of the media are normal except beneath the largest intimal plaques, where there is considerable thinning of the media. The outer portion of the media contains an abundance of elastic tissue forming a broad internal elastic lamella (Fig. 6).

The right coronary artery distal to the dilated tortuous portion shows intimal atherosclerosis, reduplication and focal breaking of the internal elastic lamella, a normal media, and well defined external elastic lamella (Fig. 4). The first branch of the artery shows moderate, uniform intimal thickening, reduplication of the internal elastic lamella, which is broken in a few foci, normal media, and a well defined external elastic lamella which is thinned in a few foci (Fig. 5).

The main stem of the right coronary artery between its first two branches shows slight, uniform intimal thickening. The internal elastic lamella is not reduplicated, but is thoroughly fragmented and contains large gaps. The elastic tissue of the media is diminished in places and many of the fibers are nearly straight. The muscle appears normal. The external elastic lamella shows some thinning and fragmentation (Fig. 1).

The dilated second branch shows slight intimal atherosclerosis. The internal elastic membrane shows some reduplication and much fragmentation, with large gaps in the lamella in some segments of the artery. There is very little elastic tissue in the media and some of the fibers are nearly straight (Fig. 2). The muscle cells in some places are hypertrophic and in places are grouped in bundles with intervening bands of collagen (Fig. 3). In all sections the external elastic lamella is thin and broken and in some places it contains large gaps.

DISCUSSION

The distribution of the dilatation of this vessel and its emptying into the right auricle, together with absence of convincing evidence

of any former infectious process, indicates that the anomaly is a congenital one. The reason for the dilatation is not so obvious. Deficiency in the elastic tissue is the most significant microscopic abnormality. Whether or not dilatation of the vessel occurred because of deficiency in the quality or quantity of the elastic tissue is problematical, but not improbable. It is, of course, possible that destruction of the elastica is a secondary change.

Communication of the artery directly with the right auricle is best explained on the basis of Grant's observations on a child's heart²⁷ in which blood-filled spaces in the ventricular muscle communicated freely with the coronary vessels and the cavity of the ventricle, and on his study of the development of the coronary vessels in rabbit embryos.²⁸ The latter study disclosed the presence of endothelial outgrowths between the forming auricular and ventricular myocardial trabeculae. These outgrowths extend out to the epicardium and develop as vessels, which in the auricles ultimately connect with the auricular branches of the coronary vessels and form the thebesian vessels. In the ventricles they join each other and the ventricular branches of the coronary vessels. For the most part, these intertrabecular spaces are reduced to capillaries, but persist as an integral part of the coronary circulation. Abnormal development of one of the auricular outgrowths would produce communication of the auricle and coronary artery, as seen in our case, but the reason for the abnormal development is obscure.

SUMMARY AND CONCLUSION

1. A case of cirroid aneurysm of the right cardiac coronary artery communicating with the right auricle is described and explained as a congenital anomaly probably due to deficiency in elastic tissue. Communication with the auricle is attributed to development of a fetal communication which is ordinarily reduced to capillary size.

2. The literature concerning certain anomalies of the coronary arteries is reviewed.

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DESCRIPTION OF PLATES

PLATE 12

All photographs were taken at a magnification of 150 diameters. The sections were stained with Verhoeff's elastic tissue stain.

FIG. 1. (A) Main stem of right coronary artery. The internal and external elastic lamellae are broken, the elastic tissue of the media is diminished and some of the fibers are straight.

FIG. 2. (B) Second branch of right coronary artery. The internal elastic lamella is broken and there is hardly any external elastic lamella. There is but little elastic tissue in the media and the fibers are straight or only slightly wavy.

FIG. 3. (C) Second branch of right coronary artery. There is a distinct internal elastic lamella and the intima contains much elastic tissue. The external elastic lamella is broken. The media contains only a few short elastic fibers. Hypertrophy of the muscle fibers in the outer portion of the media is manifest.

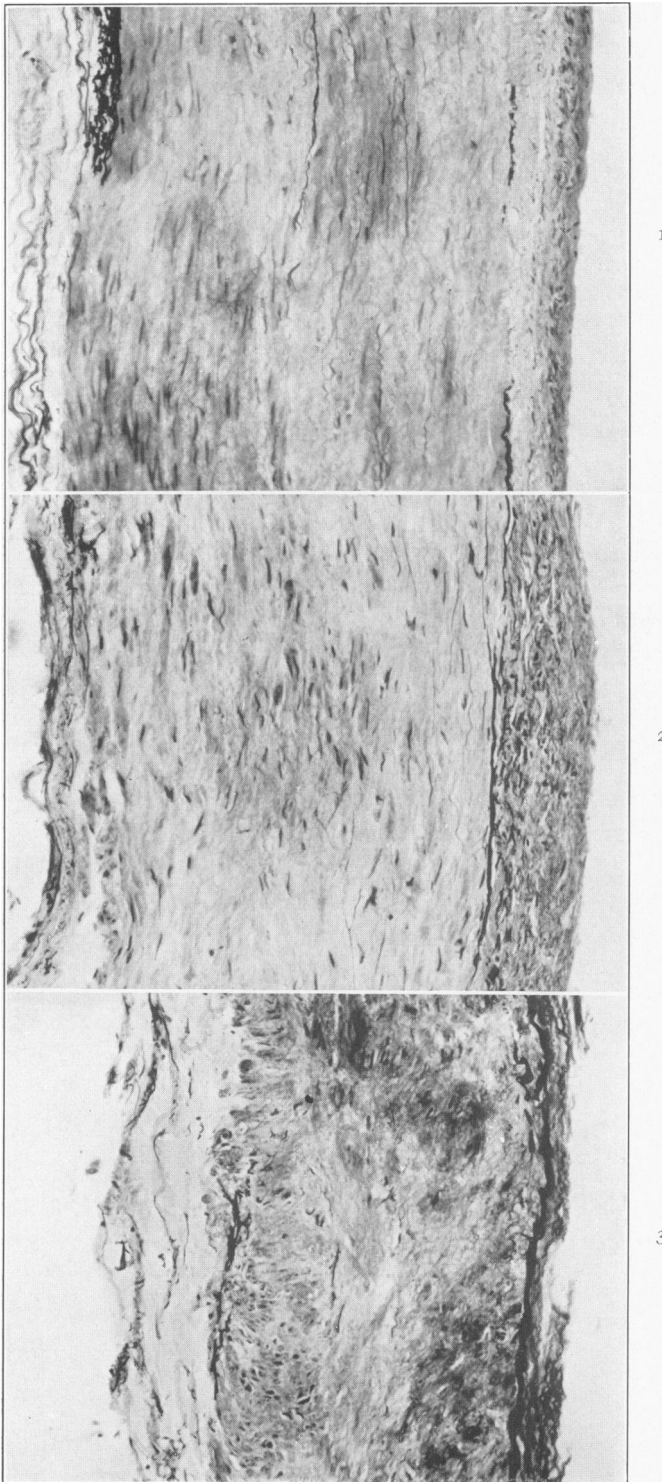


PLATE 13

- FIG. 4. (D) Main stem of right coronary artery. The intima shows atherosclerosis. The internal elastic lamella is broken and reduplicated. There is a broad external elastic lamella and the elastic tissue of the media is abundant.
- FIG. 5. (E) First branch of right coronary artery. There is reduplication of the internal elastic lamella and some thinning of the external elastic lamella. The elastic tissue of the media is abundant.
- FIG. 6. (F) Left coronary artery. The internal elastic lamella is reduplicated and broken, there is a very heavy external elastic lamella and an abundance of elastic tissue in the media.

