CLINICAL PROGRESS

Coronary Artery Anomalies

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CORONARY artery anomalies were formerly regarded as having slight clinical significance. This attitude reflected the little that could be accomplished in the way of diagnosis and treatment. Today, however, recognition seems imperative for four reasons. First, corrective surgery is available in certain instances. Secondly, there are cases in which correction is contraindicated. Thirdly, the presence of even minor anomalies may lead to considerable increased mortality and morbidity when encountered surgically. Fourthly, knowledge of these anomalies may aid in direct coronary cannulation for opacification studies and in subsequent roentgenologic interpretation of the films obtained.

Although elaborate studies of isolated aspects of this problem appear, few studies encompassing the entire spectrum of coronary artery anomalies are available. Twenty-two selected instances of coronary anomalies encountered in Walter Reed General Hospital, Brooke General Hospital, and the Registry of Cardiovascular Pathology of the Armed Forces Institute of Pathology have been reviewed and correlated with related cases from the literature. Better known anomalies are briefly mentioned, whereas less well-described anomalies are discussed in detail.

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In two of the earliest coronary artery descriptions, Fallopius in 1562 and Riolanus in 1649 held that normally a single vessel existed. It remained for Morgagni in 1761 to show that two such arteries are usually present. Unfortunately, the accuracy of much currently used material is not impressively greater than these early descriptions. Accurate knowledge concerning all aspects of the circulation of the heart is badly needed at the present time.

Wearn et al. described three types of intracardiac coronary artery terminations: namely, into the conventional capillary plexus, into a myocardial sinusoid, or into a communication connected directly with the lumen of the ventricle (fig. 1). These latter direct vascular connections between the coronary arteries and the ventricular cavities were first described by VieuSSens and are called arterioluminal by Wearn. They are frequently seen in the ventricles, but only rarely in the atria. Interposed between these various terminations is a rich intramyocardial anastomotic network, both homocoronary and intercoronary in type. Only rarely have subepicardial anastomotic channels been observed.

Venous channels, called Thebesian veins for their observer, are found frequently in the atria and less often in the ventricles. Small in diameter and consisting essentially of small endothelium-lined spaces, these vessels have been considered by some as a possible source of retrograde myocardial nourishment. Small endothelium-lined pockets, particularly likely to be found in the right
Contrast studies, secondly, chlianinels. for tant they tions which fr-omI-thlese, singge ventrictlar ouitflo Arttr.io.sinnisdal. e, No.- -2825. Thethesian 1TI/p).' of J§nt,lothelizarc-linied pockets may Sting artery often initerpreted as vascular channels. Secondly, during angiographic studies, contrast material may briefly hang up, suggesting an intramyocardial dissection by the injected material.

Edwards grouped the anomalies of the coronary arterial system into those of minor and those of major significance. Those of major significance he further subdivided into primary and secondary varieties. This most useful classification has been but slightly modified here.

The anomalies of minor significance are those that under usual circumstances cause no physiologic disturbance. These are generally anomalies of number, anatomic course, and distribution. The anomalies of major significance are those producing some degree of physiologic disturbance. Their subdivision into primary and secondary varieties indicates their respective independence of, or dependence upon, a coexisting cardiac anomaly. The major secondary group of anomalies is discussed first.

Major Secondary Anomalies
type Consideration of Retrograde Coronary Flow

These anomalies are found among seriously malformed hearts and are compensatory to the coexisting cardiac pathologic condition. The anomaly exists because it is needed. Consequently, isolated surgical correction is contraindicated. Figure 3 depicts a major secondary anomaly (communication between the right ventricle and the left coronary artery) found in a cyanotic 7-month-old infant. Additionally, the heart showed three other essential features: pulmonary atresia, an intact ventricular septum, and a competent tricuspid valve. In the presence of this complex, blood entering the right ventricle has no means of egress except through the communication between the ventricular cavity and a branch of the coronary artery. Figure 4, based on Grant's studies, attempts to show the evolution of this channel by persistence and coalescence of the primitive myocardial sinusoids and intratrabecular spaces. The irregularly enlarged, dilated channel joins the anterior descending coronary artery directly with the ventricle. An analogous situation

Figure 1
Types of coronary artery endings: a, Arterioluminal. b, Arteriosinusoidal. c, Into a capillary plexus, the vein from which may drain either into the coronary or Thebesian venous systems.

Figure 2
Endothelium-lined pockets in outflow tract of right ventricle. Radiopaque material may be held up in these, suggesting intramyocardial extravasation. AFIP Neg. No. 61-2825.
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Figure 3

Major secondary anomaly. (1) Marks point of pulmonary atresia. The right ventricular cavity (with an intact septum) is labeled (2). The aneurysmally dilated coronary branch is marked (3). A probe (4) lies in the communication between the dead-ending right ventricle and the coronary artery.

may develop on the left with the triad of aortic atresia, intact ventricular septum, and a competent mitral valve. More commonly, this decompressing communication is, however, not seen and the triads described are then found in association with a small, nonfunctioning, blood-filled ventricle.\textsuperscript{10} Comparatively infrequent, such secondary anomalies are described by Blakey et al.\textsuperscript{11} and Williams et al.\textsuperscript{12} More important clinically, however, will be the gradations encountered. Anselmi et al. have reported a complicated case in which there was severe pulmonary valvular and infundibular stenosis, an atrial septal defect, and an intact ventricular septum.\textsuperscript{13} Secondary to the above was a communication between the cavity of the right ventricle and a branch of the single coronary artery. During total repair, the single (its singularity being unrecognized at the time) coronary artery was sectioned. In spite of this, the anoxic, arrested heart, wounded by both ventriculotomy and atriotomy, was subsequently temporarily restored to a satisfactory pumping status. Resumption of beating seems unlikely without assuming either that the ligated single coronary artery received blood directly from the ventricle or that homocoronary anastomoses from branches above and below the site of division constituted an effective alternate channel. It is known that these channels may assume increased size under the influence of both hypoxia and hypertrophy.\textsuperscript{9}

Gouley, describing an anomalous left cor-

Figure 4

Drawing (after Grant) of evolution of coronary ventricular communication. a, Early embryonic myocardial sinusoids. b, Coalescence into a channel. c, Channel linking the coronary artery and ventricular lumen. d, Potentiality for bidirectional coronary flow.
ory artery arising from the pulmonary artery, described the occurrence of retrograde flow in stating, "the anterior wall of the left ventricle was nourished by a rich intramural circulation ... with the blood flow reversed, originating at the endocardial surface of the left ventricle." 14

The basis for such pathophysiologic changes is best understood by again referring to figure 1, illustrating the manner of coronary artery termination. While normally present in every heart, the first two forms, particularly when exaggerated, may constitute an anomaly in their own right. This is illustrated by figures 5 and 6. In figure 5 the sponglike myocardial spaces, in a patient with a coronary artery-ventricular fistula, constitute an exaggerated normal arteriosinusoidal type of ending. In figure 6, the postmortem injection of contrast material into the right coronary of an infant who had had surgical ligation of an anomalous left coro-

![Figure 6](image_url)

Postmortem injection study. Injection through catheter (1) tied into the aortic right coronary artery, results in immediate opacification of right ventricle (2) with spillover into the pulmonary artery (5). Sponglike collateral branches (3) passing from right toward the interrupted left pulmonary coronary are seen. The proximal stump of the interrupted left coronary (4) is also evident. The coronary sinus was never seen. This is an exaggeration of the normal arterioluminal termination (fig. 1a).

![Figure 5](image_url)

Venous angiogram (left-sided phase) showing rim of sponglike myocardial opacification, an exaggeration of the normal arteriosinusoidal type of ending (fig. 1b).

Major Primary Anomalies

The major primary anomalies in contradiction to the major secondary anomalies are independent of other cardiac pathology. Common to all are features of an abnormal communication between the high-pressure coronary system on the one hand and a cardi-
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ac chamber or vessel with a constant or intermittently lower pressure on the other. All are thus arteriovenous or arteriovenous-like shunts. Such shunts may cause myocardial failure, myocardial ischemia, or be the site for the development of bacterial endocarditis.

Anatomically and clinically this group is divisible into subgroups, depending upon the site of the artery's termination. Should the artery empty into the right atrium, ventricle, coronary sinus, or pulmonary artery, the features of a true left-to-right shunt are seen, a condition not incompatible with prolonged life. Should the artery communicate with the left atrium or ventricle, a functional disturbance resembling aortic insufficiency results. These anomalies have been extensively described and seem well understood.

Discussion of the major primary anomalies is thus limited to communications of the coronary with the pulmonary trunk.

The coronary arteries may be connected with the pulmonary artery in four ways. First, the two major coronary arteries may originate from the aorta, but with a third smaller accessory one arising from the pulmonary artery. Because of the pressure differential, this accessory artery will often eventually form a sizable connection with an aortic coronary. Retrograde flow into the pulmonary artery results in the ductus-like murmur. The characteristic gross appearance of this malformation is shown in figure 7. Repair consisted of ligation of the fistulous connection in one case and complete excision of the plexiform vascular mass in another. Others have similarly mistakenly suspected this malformation of being a ductus. There are three reports of this as an incidental autopsy finding, with that of Baylis and Campbell occurring in a 76-year-old man.

A second anomaly, both coronary arteries originating from the pulmonary artery, will likely remain of limited clinical interest with death occurring before serious study can be undertaken. Should the pulmonary artery, however, be subjected to systemic pressures, survival may be enhanced. In the type-I truncus shown in figure 8, the pulmo-

Figure 7
Fistulous malformation between a branch of the right aortic coronary (1) and an accessory third pulmonary coronary (2). Ties as shown placed about the collateral channels (3) between the two.

Figure 8
Opened truncus. Aortic side (1). Pulmonary side (2). The right coronary artery orifice (3) is located just above the valve raphe toward the pulmonary side. AFIP Neg. No. 61-2830.
ly similar cases with the right coronary arising from the anterior and pulmonary end of a aorticopulmonary window.

An infant’s heart with both coronaries arising from the pulmonary artery, living a mere 2 hours after birth, is shown in figure 9. A widely patent ductus was present as the only other cardiac finding. Fortunate enough to make an antemortem diagnosis, the banding procedure for production of a pulmonary hypertension would seem logical.36

Of more practical interest, is the origin of but one major coronary artery from the pulmonary trunk. When this is the right, the patient may do well with little disability, and with discovery often being made as an incidental autopsy finding. The excellent prognosis without surgery is attributable to the greater physiologic reserve of the right ventricle. This reserve has been demonstrated by Starr et al.,37 Bakos,38 and Kagan,39 who showed that near complete destruction of the free right ventricular wall produces little immediate hemodynamic effect.

In contrast, the physiologic disarrangements from an anomalous left coronary artery are severe, poorly understood, and the results of surgery far from satisfactory. As most emphasis has been placed on recogni-

tion and surgical treatment of this anomaly in the infant, one loses sight of numbers of these patients surviving to adulthood.40, 41 One of us (H.A.B.) has corrected this anomaly in a 33-year-old woman presenting with angina-like chest pain. Three years after surgery the patient remains well, is free from angina, and has completed an uneventful pregnancy and delivery.42 Others emphasize the coexistence of this coronary artery anomaly with mitral insufficiency.43 Correction of this insufficiency has been reported.43, 44

Adult patients with an anomalous origin of the left coronary artery present a unique opportunity to evaluate the relative importance of several factors in the genesis of coronary atherosclerosis. Both arteries possess common genetic, hormonal, and dietary backgrounds. Flows, pressures, and, by inference, trauma to the two, however, differ widely. The right coronary artery usually shows considerable atherosclerotic change, the left little, confirming the relative importance of flow, pressure, and trauma as causes of such changes.

**Minor Coronary Artery Anomalies**

In these, the anatomic abnormality produces no physiologic disturbance, as the artery arises from the aorta and makes no unusual connections. With development of open-heart surgery, these abnormalities in number, origin, size, course, and distribution began to cause technical problems, thereby increasing the morbidity and mortality of repair of certain cardiac defects.45-49 The question as to the number of coronary arteries is discussed first.

Three coronary arteries are found frequently.50 The third, or “conus” artery, arises separately in the right aortic sinus and courses to supply the conal area of the right ventricle (fig. 10). Present in from 23 to 50 per cent of all hearts, its intermediate position may lend importance as a collateral pathway in blockage of either major coronary. If present, the first right coronary artery branch usually supplying the conus will noteworthy be either small or absent.51, 52 Accessory third ar-
Enlarged conus artery passing to the outflow tract of the right ventricle. When present and this enlarged, the first branch of the right coronary is usually absent. Surgical interruption may be dangerous.

A single coronary artery may or may not be a benign anomaly. Figure 11 shows the excellent state of the interior of the aorta and orifice of the single coronary artery found in an 80-year-old patient dying a noncardiac death.53

Upon opening the aorta with a single coronary artery, one may find a tiny dimple marking the site of an involuted coronary artery anlage. With involution, collateral channels from the opposite coronary must now supply the deficient area. The heart of a 20-year-old soldier who died suddenly while hiking with full field equipment is shown in figure 12. The right coronary has disappeared, with only a prominent dimple remaining to mark its atretic orifice. One presumes that the collateral supply to the area normally supplied by the right coronary artery was insufficient, with death the result of the unusual physical exertion. This situation should be compared with that illustrated in figure 24.

The single coronary artery has additionally resulted in death following open-heart surgical procedures.13,47 The singularity may go unrecognized owing to the obscuring peri-vascular fat, and when a ventriculotomy incision is made, indispensable collateral vessels may be sacrificed. Baffes54 has commented on the particularly troublesome single coronary artery encountered in transpositions.

We have twice encountered four independent coronary artery orifices. Figure 13 is an artist's interpretation of the distribution of these orifices in one such case. Others have reported as many as three separate branches arising from within a single sinus.55-57 The greatest number of coronary arteries reported has apparently been six, found in a description byEssenberg.58

The mode of origin warrants consideration.
Figure 13
Aorta with four separate coronary artery ostia.

Figure 14 depicts two coronary artery branches arising at the lateral extremes of a shallow oval depression representing their common orifice. Closely related is the variation seen in figure 15 where only a slight bridge or spur exists between the two orifices. Whether these are separate coronary arteries or in reality a quickly branching single coronary artery, as reported by Bland, is debatable.

The importance of these anomalies relates to the surgical difficulties encountered in cannulating these vessels during open aortic surgery or in performing coronary arteriography. In the latter, to prevent overlooking one of several closely spaced orifices, a preliminary aortic root injection outlining the number of coronary arteries present is necessary. Such a diagnostic pitfall is illustrated by figure 16. If only the independently arising circumflex branch were cannulated and outlined, the unfilled anterior descending branch might erroneously be regarded as obstructed.

Whereas certain orifices are difficult to cannulate, entry into others can hardly be prevented. Figure 17 depicts a dilated funnel-like sinus of Valsalva with the left coronary artery exiting from its apex. Blind aortogram catheter positioning with subsequent injection of contrast media is hazardous because of the easy entry into the coronary arteries such modified sinuses encourage.

Figure 15
Close-spaced separate orifices of right and left coronary. Arrow indicates narrow septum between the two.

Figure 14
Looking from above toward the aortic valve, the two arrows mark the separate coronary artery ostia lying at the lateral extremes of the shallow common orifice. AFIP Neg. No. 61-2832.

Figure 16
Independent origin from the left aortic sinus of the two branches of the left coronary artery.
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Figure 17

Newborn's heart with the left coronary artery arising from a diverticulum of the left sinus. Such funnel-like configurations encourage misplacement of blindly positioned aortic catheters.

There is little agreement as to the normal height of "take-off" of the coronaries from the aorta. In normal hearts, in the experience of Banchi, the left coronary artery arose at the level of the free margin of the aortic cusps in 48 per cent, above in 34 per cent, and below in 18 per cent. The right coronary artery arose at the same level in 71 per cent, above in 19 per cent, and below in 10 per cent.

An extreme example of a high take-off occurs when the coronary artery seemingly arises from the arch itself. Such cases are suspect in the absence of careful histologic study. The schematic drawing in figure 18 would seemingly represent an instance of high take-off of a single coronary artery from the arch in a case of truncus. In reality, the small vessel coursing from the arch to the heart and having a coronary type of distribution is a small ascending aorta giving rise to coronary branches. Histologic studies provide verification showing it to be an elastic rather than muscular artery. The large vessel off the heart is the main pulmonary artery. The widely patent ductus passing to the arch of the aorta was not recognized as such, although its wrinkled intimal surface probably should have identified it. These cases, interesting academically, are overshadowed in terms of practical importance, however, by the coronary artery with lesser degrees of high take-off. Figure 19 is an interior ascending aortic view showing a left coronary arising within its sinus, whereas the right orifice is located well up onto the anterior wall of the ascending aorta.

When viewed from without, this artery is almost invariably obscured by the overlying periaortic fat. By referring to figure 20, three surgical misadventures can be anticipated: (1) An aortotomy veering laterally might divide such a hidden highly placed coronary. (2) In dissecting out the aortic root for valve

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replacement, this misplaced artery might be accidentally detached. (3) Low cross-clamping of the aorta to obtain a drier intracardiac field might damage an atheromatous, highly placed coronary with an unfavorable surgical result.

The tendency to emerge at these higher levels has been noted in several cardiac malformations. In our series of truncus arteriosus, high take-off occurred in 19 per cent of the left and 9 per cent of the right coronary arteries. It is also a frequent finding in the transpositions. Idriss et al. have suggested taking surgical advantage of this by isolating and mobilizing this segment and placing it in a correct position above the left ventricle. The very frequency of high take-offs seen in cases of anomalous left coronary artery suggests its being a possible causative feature of the anomaly. As illustrated diagrammatically in figure 21, a low-originating left coronary will arise from the aorta normally, whereas a high-originating left coronary may arise from the pulmonary artery portion of the divided common trunk above the spiral septum as it passes to the left. Baffes' proposed operation is facilitated by this situation.

The anomalously placed coronary artery

**Figure 19**

*Interior view of opened aorta, showing high take-off of a right coronary artery. AFIP Neg. No. 61-2827-2.*

**Figure 20**

*Drawing illustrating the danger in high take-off of right coronary artery. Injury may occur in aortotomy, in dissecting out the root of the aorta, or in low cross-clamping.*

**Figure 21**

*a and b. Cylinders representing primitive undivided truncus. c. The spiral septum has divided the truncus. If the left coronary artery arises low, its origin will lie in the aortic segment. d. If the take-off is high, it will originate from the pulmonary side.*

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Figure 22
Coronary artery distribution found in a tetralogy. The right coronary (1) gives rise to an anterior descending branch (3) which passes across the right ventricular outflow tract below an infundibular chamber (4). The arrow (2) points to a portion of the left coronary. Two small separate incisions provided reasonable intracardiac exposure.

Figure 23
A quickly branching single coronary artery. The two stem branches result in a good vascular distribution. The left anterior passing branch, however, proved troublesome at ventriculotomy.

and then in front of the pulmonary artery in a heart with two coronary arteries. 45

Closely related physiologically to the above is hypoplasia of one of the coronary arteries. Figure 24 shows the coronary pattern in a 20-year-old man dying suddenly while playing basketball. The left coronary artery is normal in take-off, size, and distribution. The right artery, however, is extremely tiny (2 mm.) with its branches disappearing quickly and being untraceable because of their size. Death during stressful exercise was stated to

Figure 24
Hypoplasia of the right coronary. Compare this with its physiologic equivalent seen in figure 12.

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be the result of myocardial insufficiency, secondary to this hypoplastic artery. Jokl et al. report an almost identical circumstance. Caution should be exercised in equating death from coronary insufficiency with small vessel size. When the "sudden death" rates in a large series of patients having a "normal" coronary distribution are compared to those with this type of distribution, the rates are seen to be nearly identical.

The last condition to be described is that of the hidden or mural coronary artery. Instead of occupying a surface position covered by a thin layer of fat, the artery dips into the myocardium, either to reappear shortly or to remain hidden for much of its course. Myocardial fibers cross the artery at right angles in bridge fashion. Such mural arteries were originally described by Crainicianu and Spalteholz. Edwards et al. found a 5-per cent incidence, Geiringer a 23-per cent incidence, and Poláček an 85 per cent incidence. Figure 25 shows a branch of the right coronary artery dipping beneath a thick myocardial bridge. Located higher and having a more extensive covering, ventriculotomy in

this child with a tetralogy might have proved dangerous. Derrick et al. report such an operative death. Bloor and Lowman emphasize the importance of these structures in the interpretation of coronary arteriograms. Distortion by this muscular bridge may simulate a focal stenosing process within the artery.

Summary

Twenty-two selected anomalies of the coronary arteries are related to cases previously reported in the literature. These are presented with simplified drawings and photographs in an attempt to provide a workable understanding of the practical problems such create for both the internist and surgeon.

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Figure 25
Mural or hidden coronary. The bridging, here quite obvious, is not always so.


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