Absence of one of the coronary arteries is a relatively frequent variation of the coronary circulation. It is usually found during postmortem examination, but the increasing number of cases found during diagnostic procedures and surgery illustrates the significance of this variation as a clinical entity.

It should be emphasized that this lesion is not one of absence of a complete coronary artery, but rather is the absence of a portion of one of the coronary arteries at the aortic root. This initial segment is referred to as the “proximal coronary artery.” The more distal portion of the “absent” coronary artery is found in the usual epicardial location (i.e., the atrioventricular or interventricular grooves). This distal segment communicates freely with the normally arising coronary artery, and thereby creates a coronary circulatory pattern with a single aortic source—a “single” coronary artery.

Ten cases of single coronary artery were found in a recent survey of congenital variations of the coronary arteries. These cases and those cases previously reported in the literature will be reviewed with regard to the anatomical patterns of distribution and clinical significance, and a new, comprehensive classification scheme will be presented.

CASE MATERIAL

Ten cases of single coronary artery were found in this survey. Data concerning these cases have been summarized in Table 1. Nine cases were found during autopsy and one case was found during evaluation of coarctation of the aorta. The ages ranged from one day to eighty-four years. There were six males and four females. A single coronary artery was present in five cases and a single left coronary artery in five cases. The single left coronary artery in one case arose from the pulmonary artery rather than the aorta. Associated cardiovascular anomalies were found in three cases: truncus arteriosus (case 7) and transposition of the great vessels (cases 8, 9).
Table 1. Summary of 10 Cases of Single Coronary Artery Found in the Current Case Review

| Case no. | Age | Sex | Classification | Comment |
|----------|-----|-----|----------------|---------|
| 1        | 70y | F   | L-1            | Small branch arose from anterior descending artery and crossed the right outflow tract. |
| 2        | 68y | M   | L-1            | ——        |
| 3        | 84y | M   | L-1            | Dimple in right aortic sinus: fibrous cord from dimple to distal right coronary artery. |
| 4        | 6mo | F   | R-4a           | ——        |
| 5        | 10y | M   | L-4            | Found during evaluation of coarctation of the aorta. |
| 6        | 11d | M   | R-4a           | ——        |
| 7        | 6mo | M   | R-5a           | Truncus arteriosus. |
| 8        | 1d  | F   | R-2a           | Transposition of the great vessels. |
| 9        | 42d | M   | R-4a           | Transposition of the great vessels. |
| 10       | 6d  | F   | L-3            | Single left coronary artery arose from the pulmonary artery. |

Case 3 had a unique variation. The heart was supplied by a single left coronary artery. A short proximal right coronary artery (dimple) was found in the right aortic sinus. This structure terminated shortly after the aortic origin, with a nonpatent, fibrous segment connecting the blindly ending proximal right coronary artery with the distal right coronary artery. The distal right coronary artery was a continuation of the left circumflex artery (Figure 1a).

Case 10 had an extremely unusual lesion. A single left coronary artery was present. However, this single artery arose from the left sinus of the pulmonary artery rather than the aorta. No coronary arteries originated from the aorta. The single coronary artery divided into three branches. The right coronary branch coursed between the aorta and pulmonary artery to reach the right atrioventricular groove, while the circumflex and anterior descending branches assumed their normal courses of distribution (Figure 1b).  

**DISCUSSION**

The single coronary artery was first reported by Hyrtl in 1841 when he described an absent right coronary artery in a seven-month old fetus.  


Fig. 1. A. Case 3—an outpouching (PR) extended from the right aortic sinus and ended abruptly. A non-patent, fibrous cord (F) connected this outpouching (proximal right coronary artery) with the distal right coronary artery (DR), which was a direct continuation of the circumflex branch of the left coronary artery (C). B. Case 10—a single coronary artery arose from the left pulmonary arterial sinus (0). The anterior descending (AD) and circumflex (C) branches were normal. The branch that became the right coronary artery (R) coursed between the aorta and pulmonary artery to reach the right atrioventricular groove.

Together 142 cases have been found in the literature and the present case survey.1 Of the 142 cases, 63 were males and 46 females. The sex was not given in 33 cases. From these figures it appears that the variation is slightly more common in males (1.4 to 1). This concurs with a male-female ratio of 1.4:1 reported by Allen in a review of 69 cases.4

The single coronary artery arose from the right aortic sinus in 70 cases and from the left aortic sinus in 64 cases. In four cases the single left coronary artery arose from the left sinus of the pulmonary artery. Four cases could not be classified. Thus, the variation affected the right and left coronary arteries with approximately the same frequency.

The cases ranged in age from a seven month old fetus to an 84-year old male. Sixty of the patients were less than twenty years of age. Thirty-two were fifty years of age or older, with six patients surviving to the ninth decade. The single coronary artery appears to be compatible with longevity.

Fifty-six cases were associated with other congenital cardiovascular anomalies.5 Only five of these cases were more than 20 years of age. Thus,
of the 60 reported cases under 20 years of age, 51 were associated with other congenital cardiovascular lesions of varying severity. Previous reports have stressed that the early deaths of patients with a single coronary artery were related to the primary cardiac pathology, rather than the coronary variation.6

The most frequent concurrent cardiac anomaly was transposition of the great vessels (17 cases). The next most common anomaly was a coronary artery-cardiac chamber fistula (seven cases of primary fistula and three cases of secondary fistula). Hallman, et al. corrected a fistulous single coronary artery by initially closing the fistula and then anastomosing a graft from the aorta to the transected end of the coronary artery, thereby creating a normal, two coronary system. A complete listing of the cardiac anomalies found in conjunction with the single coronary artery is found in Table 2.

While many reports have stressed that the single coronary artery is a relatively benign lesion, others have given evidence of coexistent myocardial ischemia.6,7 Twenty-seven cases had some evidence of coronary insufficiency.6 Further, sudden death at an early age during heavy exercise has been reported.6,8

Most examples of single coronary artery were discovered at autopsy or surgery. However, four cases have been diagnosed during clinical evaluation. The first case was a 24-year old male with hypertension and electrocardiographic evidence of an old infarction. Aortography demonstrated a

Table 2. Congenital Cardiovascular Defects Found in Hearts with a Single Coronary Artery

| Defect                                      | No. of cases |
|---------------------------------------------|--------------|
| Transposition of the great vessels          | 17           |
| Coronary (arteriovenous) fistula            | 10           |
| Bicuspid aortic valve                       | 7            |
| Tetralogy of Fallot                         | 5            |
| Origin from the pulmonary artery            | 4            |
| Truncus arteriosus                         | 3            |
| Pulmonary valvular atresia                  | 2            |
| Anomalous anterior descending branch        | 2            |
| Aortic valvular atresia                     | 1            |
| Ventricular septal defect                   | 1            |
| Patent ductus arteriosus/patent foramen ovale | 1       |
| Aortic insufficiency                        | 1            |
| Tricuspid atresia                           | 1            |
| "Congenital heart disease"                  | 1            |
single left coronary artery. Halperin reported a 27-year old female with congestive heart failure during pregnancy. Coronary arteriography in the postpartum period showed a single left coronary artery, which was confirmed by autopsy a few months later. The third case was a 10-year old male with coarctation of the aorta. Retrograde aortography demonstrated a single left coronary artery. The fourth case was quite unusual. Arteriography revealed a single left coronary artery with circumflex and right coronary branches, but without evidence of an anterior descending branch. Instead, the anterior descending artery and vein arose from the left internal thoracic (mammary) vessels and crossed the pericardial cavity in a fibrous band to the apex of the heart.

Hyrtle defined the single coronary artery as being present when one artery supplied the entire heart. He further stated that this anomaly must not be a matter of "a common aortic orifice of the two arteries or an unusual origin of the missing one." Such a definition excluded several variations of the single coronary artery found in this review.

Smith was the first to attempt an anatomical classification of the single coronary artery. He proposed three categories. Type 1 was a single coronary artery that followed the distribution pattern of only one coronary artery. He hypothesized that this situation was due to the congenital absence of one of the embryonic buds of the coronary arteries. This type probably corresponded to Hyrtle's definition of a single coronary artery. Type 2 was a single coronary artery that followed the distribution of both coronary arteries. This variety was associated with almost immediate division into right and left branches. Smith felt that this variety arose by a misplacement of a coronary artery bud such that the misplaced bud fused with the initial portion of the normally arising artery. Type 3 comprised those cases that had an atypical distribution of the coronary arteries. Longnecker enlarged upon this classification scheme by adding type 4—cases for which insufficient data were given.

However, these classification schemes are incomplete, since they fail to define precisely what variations should be included in each category. Because of these inadequacies of definition, a new scheme has been devised in which the arterial patterns of the single coronary artery will be classified according to several basic anatomical distributions.

This new scheme is based upon (1) the number of initial major divisions of the single coronary artery, and (2) the subsequent course of these major branches. The initial (proximal) segment of the coronary artery may not divide at all, or it may divide into two or three major branches. These major branches may assume a normal course of distribution, or may course in the area normally supplied by the "absent" coronary artery. In the latter
SINGLE CORONARY ARTERY
BASIC DISTRIBUTION PATTERNS

RIGHT CORONARY ARTERY

| TYPE 1 | NORMAL
|--------|--------|
| R-1    | (3)    |

| TYPE 2 |
|--------|
| R-2a   | (4)    |
| R-2b   | (0)    |

| TYPE 3 |
|--------|
| R-3a   | (11)   |
| R-3b   | (3)    |

| TYPE 4 |
|--------|
| R-4a   | (12)   |
| R-4b   | (4)    |

| TYPE 5 |
|--------|
| R-5a   | (5)    |
| R-5b   | (1)    |
| R-5c   | (1)    |

LEFT CORONARY ARTERY

|        |        |
|--------|--------|
| L-1    | (31)   |
| L-2    | (17)   |
| L-3    | (1)    |
| L-4    | (2)    |

Fig. 2. Projectional diagrams of the base of the heart illustrating the basic anatomical distribution patterns of the single right coronary artery and the single left coronary artery. The normal situation is depicted as a reference (Ao— aortic valve; P— pulmonar y valve; T— tricuspid valve; M— mitral valve; RCA— right coronary artery; LCIRC— left circumflex artery; LADCA— left anterior descending artery). The text should be consulted for detailed descriptions of each type.
situation the branches reach the area of distal distribution by crossing behind the aorta, between the great vessels, anterior to the pulmonary artery (across the right ventricular outflow tract), or as a continuation of a vessel on the posterior surface of the heart. These patterns are depicted in Figure 2 and will be described in more detail in the ensuing paragraphs.

The patterns in Figure 2 are derived from the ten cases being reported from the current study and those cases in the literature that gave sufficient description of the course of the single artery to allow graphic reconstruction. Using these criteria 95 cases were available for the compilation of Figure 2.

Type 1—one major vessel: The single right coronary artery continues on the posterior surface as the circumflex branch and terminates as the anterior descending branch (3 cases). In the corresponding single left coronary artery, the circumflex branch continues as the distal right coronary artery (31 cases).

Type 2—two major branches, with one branch coursing posterior to the great vessels: There are two variations of the single right coronary artery. In type 2a there is a normal distal right coronary artery, while the branch that courses posteriorly around the aorta is the main left coronary artery, which subsequently divides into circumflex and anterior descending branches (4 cases). In type 2b the circumflex branch is a direct continuation of the right coronary artery, while the vessel coursing posteriorly around the great vessels becomes the anterior descending branch (hypothetical case). In the corresponding single left coronary artery, the artery bifurcates into a normal main left coronary artery and a branch that loops posteriorly around the great vessels to become the distal right coronary artery (17 cases).

Type 3—two major branches, with one branch coursing between the great vessels: There are two variations of the single right coronary artery. In type 3a there is a normal distal right coronary artery, while the main left coronary artery arises immediately from the right artery, courses between the aorta and pulmonary artery, and subsequently divides into circumflex and anterior descending branches (11 cases). In type 3b the distal right coronary artery continues as the circumflex branch, while the branch coursing between the great vessels becomes the anterior descending branch (3 cases). In the corresponding single left coronary artery the vessel bifurcates into a normal main left coronary artery and a branch that crosses between the great vessels to become the right coronary artery (one case). The course of the branch between the great vessels varies. The branch may course superficially between the aorta and pulmonary artery, being surrounded only by epicardial fat (Figure 3a), or the involved
branch may immediately enter the upper portion of the interventricular septum, cross within the septum, and subsequently emerge on the opposite epicardial surface to assume a normal distribution pattern (Figure 3b).

Type 4—two major branches, with one branch coursing anterior to the great vessels: There are two variations of the single right coronary artery. In type 4a there is a normal distal right coronary artery, while the main left coronary artery arises as one of the initial branches of the right artery, crosses the ventricular outflow tract anterior to the great vessels, and subsequently divides into circumflex and anterior descending branches (12 cases). In type 4b the distal right coronary artery continues as the circumflex branch, while the branch crossing the right ventricular outflow tract becomes the anterior descending branch (4 cases). In the corresponding single left coronary artery the distal right coronary artery arises from the left anterior descending artery and crosses the ventricular outflow tract (2 cases).

Type 5—three major branches: There are three variations, all involving the right coronary artery. The distal right coronary artery has a normal course of distribution in each case. In type 5a the circumflex branch
courses posteriorly around the great vessels, while the anterior descending branch courses across the right ventricular outflow tract (5 cases). In type 5b the circumflex branch courses posteriorly around the great vessels while the anterior descending branch crosses between the aorta and pulmonary artery (one case). In type 5c the circumflex branch crosses between the aorta and pulmonary artery, while the anterior descending branch courses anterior to the great vessels (one case).

Thus, in future case descriptions, the following pattern should be used. The single coronary artery should be indicated by the letter “R” or “L” to indicate whether the vessel arises from the right or left aortic sinus. This letter should be followed by a number, 1-5, to indicate the pattern of anatomical distribution of the branches according to the initial divisions. In cases of single right coronary artery, types 2-5, the appropriate letters “a,” “b,” or “c” should be added to indicate the courses of the branches. Thus, a single right coronary artery that divided into two branches, with one branch coursing between the great vessels and dividing into circumflex and anterior descending branches, would be classified R-3a.

In some cases one of the main branches may be absent. Such cases should be classified according to those major branches present. For example, in the case reported by Robicsek, a single left coronary artery divided into circumflex and right coronary branches, with the right coronary branch crossing the ventricular outflow tract. The anterior descending vessel arose from the internal thoracic (mammary) artery. This variation should be classified L-4, with a notation as to the site of origin of the anterior descending branch.

Those rare cases in which the single left coronary artery arises from the pulmonary artery instead of the aorta can be treated similarly, with classification based upon the patterns of branching, and a notation to indicate that the origin is from the pulmonary artery. Thus the case found in the current series would be classified L-3 because the right coronary branch courses between the aorta and pulmonary artery.

While this classification scheme may appear complicated, it is necessary for an adequate understanding of the varied anatomy present in this lesion. Functionally there does not appear to be any significant difference, except in type 3, in which the course of the left artery between the aorta and pulmonary artery has been implicated as a factor in the ischemia reported in a few patients.

The cardiovascular surgeon must be aware of these variations and revise his approach accordingly when he is aware that a single coronary artery is present. In particular, ventriculotomy incisions should parallel the possible course of a major vessel crossing the right ventricular outflow tract (types
R-4a, R-4b, L-4, R-5a and R-5c). Further, if a single ostium is encountered at the time of perfusion of the coronary arteries, care should be taken that the perfusion cannula does not occlude a major branch.

In 15 cases there was an outpouching (dimpling) in the aortic coronary sinus of the "absent" coronary artery. No distal artery arose from this outpouching. In two cases the outpouching was connected to the distal segment of the "absent" coronary artery by a fibrous cord (Figure 1a). These outpouchings represent proximal anlagen (buds) of the "absent" coronary artery that either failed to develop appropriately, failed to canalize, or became atretic during an early phase of the development of the coronary arteries.

In the development of the single coronary artery, whatever the final pattern of distribution of the coronary branches, all probably arise by a common mechanism—the developmental failure of the proximal anlage of the absent coronary artery in conjunction with the enlargement of various anastomoses between the "single" coronary artery and the distal portion of the "absent" artery. These anastomoses then enlarge to the same caliber of the main epicardial vessels.

SUMMARY

Ten cases of single coronary artery were found in a recent review of congenital anomalies of the coronary arteries. Clinical considerations regarding these cases, as well as those previously reported in the literature, are discussed. Because of the lack of a uniform, precise method of classification, a new scheme based upon the anatomical distribution patterns has been devised. Basically the single coronary artery patterns are separated into five types. The tentative embryological basis of these types is discussed.

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