

Sudden Death as a Complication of Anomalous Left Coronary Origin From the Anterior Sinus of Valsalva

A Not-So-Minor Congenital Anomaly

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SUMMARY

Both coronary arteries arising as a single or double vessel from the same sinus of Valsalva has been considered a minor congenital anomaly not affecting longevity. There are rare case reports in the literature of sudden death in young males with this anomaly of coronary origin. We have reviewed 51 such cases from the Armed Forces Institute of Pathology congenital heart disease accessions. There were 33 patients in whom both coronaries arose from the anterior sinus of Valsalva either as a single or double vessel and 18 in whom they arose from the left coronary sinus of Valsalva. Out of the 33 patients in whom the coronaries arose from the anterior sinus of Valsalva, 9 (27.3%) died sudden, unexplained deaths. There were no sudden unexplained deaths among the patients in whom both coronaries arose from the left sinus of Valsalva. It is evident that where the left coronary artery does not pass acutely posterior and leftward between the pulmonary artery and the aorta there is no risk of sudden death. All patients who died suddenly were male. The suggested mechanism for sudden death in these cases is that the acute leftward passage of the coronary artery along the aortic wall causes the entrance into the left coronary system to be slit-like. Under circumstances of increased cardiac activity with increased expansion of the pulmonary artery and aorta with exercise, there is stretching of the left coronary artery and a flap-like closure of the orifice of the left coronary with sudden, fatal myocardial ischemia.

We also present the first case where this anomaly was recognized in a 14-year-old boy and a surgical attempt was made to correct the problem by creating a non-collapsible funnel-like opening into the left coronary artery.

Additional Indexing Words:

Coronary artery anomalies

SINGLE CORONARY ARTERY and origin of both coronary arteries from the same sinus of Valsalva have traditionally been regarded as having little clinical significance and being compatible with a long and active life.¹ In Ogden's classification of congenital coronary anomalies these variations of origin are listed as minor congenital anomalies.² Patients have been described as living into the eighth and ninth decades of life and finally dying of unrelated diseases, thus reinforcing the opinion that these anomalies are benign.^{3, 4}

With the advent of coronary arteriography and cardiopulmonary bypass surgery, anomalies of coronary origin and distribution have taken on increased significance as relates to the interpretation of coronary arteriograms and to the technical problems of open heart surgery.⁴

There are, however, sporadic case reports of sudden death described in patients in whom both coronary arteries arose as a single or double vessel from the anterior sinus of Valsalva which suggest that these anomalies are not always without clinical importance.^{5, 6, 7} Benson and Lack have collected three instances of sudden death with this anomaly, two occurring in teenage boys after exercise, and have warned that this appears to be one cause of sudden death with exertion.⁵

In the course of reviewing all congenital coronary anomalies accessioned at the Armed Forces Institute of Pathology (AFIP), we have collected all cases of single coronary artery or both coronary arteries arising

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from the same sinus of Valsalva. It is the purpose of this paper to describe these cases and to show that when the coronaries arise from the anterior sinus of Valsalva and the left coronary artery passes obliquely between the aorta and the pulmonary artery, a significant number of patients die suddenly in their early years. Furthermore, the sudden death almost always occurs during or after physical activity. This group is compared to the patients in whom either a single coronary artery or both coronary arteries arise from the *left* sinus of Valsalva with the right coronary artery passing between the aorta and the pulmonary artery. In this circumstance, we found no association with sudden death.

We will also briefly report the first instance in which this anomaly was recognized by coronary angiography and a surgical approach to correction was made in a 14-year-old symptomatic boy. A detailed report of this case is in preparation.

Methods

The entire collection of cases with coronary artery anomalies at the AFIP was reviewed. Patients who had either a single coronary artery or both coronary arteries from the same sinus of Valsalva without other congenital cardiac anomalies were included. All patients had clinical and autopsy summaries. All available specimens were examined by the authors. Where specimens were not available, the autopsy description and photographs of the specimens, when available, were reviewed.

The AFIP receives its specimens from physicians and hospitals all over the world. Its major source of referrals are service and government hospitals including the Veterans Administration hospitals. Specimens are received also from universities and private hospitals.

Because of the large contribution from the military and the Veterans Administration hospitals, there is a large bias in favor of males. This is not true for the congenital hearts accessioned under the age of 20 years. For instance, in the

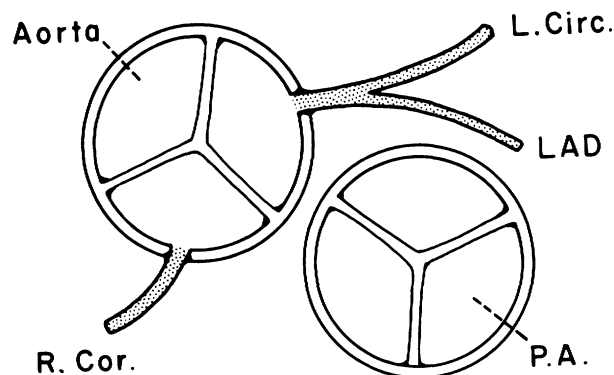


Figure 1

Normal relationship of pulmonary artery (P. A.) and aorta with origin of the left coronary artery from the left sinus of Valsalva and the right coronary artery (R. Cor.) from the anterior sinus of Valsalva. LAD = left anterior descending coronary artery; L. Circ. = left circumflex coronary artery.

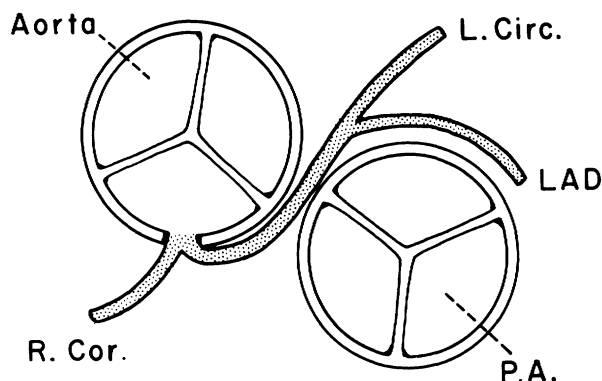


Figure 2

Both coronaries from anterior sinus of Valsalva. Notice leftward posterior passage of the left coronary artery.

age group 0 to 20 years there were a total of 151 hearts with coronary anomalies, 81 from males and 63 from females. Over the age of 20 there was a heavy male bias throughout the congenital heart collection.

Figure 1 is the diagram which will be used throughout the paper to illustrate the usual relationship of coronary arteries to aorta and pulmonary artery.

Results

There were 51 patients with the coronary arteries arising either as a single or double vessel from the same sinus of Valsalva without other congenital cardiac anomalies. In 33 hearts the coronary arteries arose from the anterior sinus of Valsalva (fig. 2), and in 18 they arose from the left sinus of Valsalva (fig. 3).

Table 1 lists all 33 patients with coronaries arising from the anterior sinus of Valsalva. The first 9 patients (27.3%) had sudden, unexplained deaths. With the exception of case 9, a 36-year-old man who died suddenly apparently of an arrhythmia, the patients were between 13 and 22 years of age, and 7 of the 9 died during or immediately after exercise. The

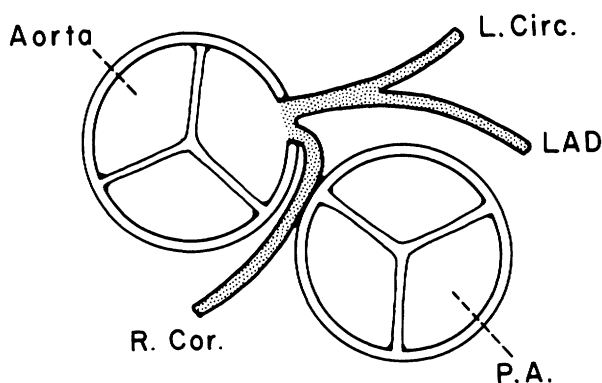


Figure 3

Both coronaries from the left sinus of Valsalva. Note passage of right coronary artery anterior and rightward.

Table 1

Both Coronaries from Anterior Sinus of Valsalva

No.	Age	Sex	Number of coronary ostia	Cause of death and comments
<i>Sudden death</i>				
1.	13	M	Single	Massive myocardial infarction. Died 4 months later*
2.	14	M	Double	Died suddenly after exercise
3.	18	M	Double	Died while running
4.	17	M	Double	Died while running
5.	18	M	Double	Died while running
6.	22	M	Double	Died while running†
7.	20	M	Double	Died during forced march. Had myocardial infarction
8.	22	M	Double	Died while running
9.	36	M	Single	Died suddenly after clutching chest and falling into river. Documented ventricular tachycardia in past‡
<i>Known cause of death</i>				
10.	28	M	Single	Multiple pulmonary emboli‡
11.	54	F	Single	Cirrhosis — gastrointestinal hemorrhage‡
12.	37	M	Single	Meningitis after nasal operation. Left anterior descending coronary anterior to pulmonary artery
13.	87	F	Double	Pulmonary emboli‡
14.	48	M	Double	Cancer of larynx
15.	47	M	Single	Postoperative thromboemboli
16.	49	M	3 ostia	Coronary artery disease. Myocardial infarction
17.	40	M	Double	Aortic stenosis. Bacterial endocarditis
18.	49	M	3 ostia (left coronary small)	Coronary artery disease. Myocardial infarction
19.	65	M	Single	Luetic aortic insufficiency
20.	66	M	Single	Coronary artery disease. Dissection of aorta
21.	64	M	Double	Coronary artery disease
22.	66	M	Double (right posterior cusp)	Cirrhosis. Acute myocarditis
23.	67	M	Double	Cancer of the lung
24.	70	M	Single (right coronary distribution)	Cancer of the cecum
25.	72	M	Single	Cerebrovascular accident
26.	69	M	Double	Coronary artery disease. Myocardial infarction
27.	82	M	Double	Coronary artery disease
28.	81	M	Double	Coronary artery disease. Myocardial infarction‡
29.	62	M	Single (right coronary posterior)	Cancer of the lung
30.	24	M	Single	Died at surgery for umbilical hernia. Left anterior descending artery anterior to pulmonary artery
31.	42	M	Double	Myocardiopathy
32.	54	M	Single	Coronary artery disease
33.	53	M	Single	Coronary artery disease

*USHER B, McGRANAHAN G: personal communication.

†Ref (8), Roberts and Loube, case #5

‡Left coronary artery tunnels through myocardium to reach septum. See figure 5.

remaining 24 patients each died of known causes unrelated to the coronary artery anomaly.

When the coronaries arose from the anterior sinus of Valsalva the distribution of both right and left coronaries with the exception of the initial portion of the left coronary was entirely normal in 27 of 33 patients (table 1). In patient 12 the anterior descending coronary passed anteriorly across the right ventricular outflow tract. In patients 9, 10, 11, 13, and 28 the left

coronary artery tunneled into the myocardium shortly after its origin to become subendocardial for a portion of its path. The incidence of sudden death in the patients with the intramyocardial tunnel (1 of 5) is not significantly different from that of patients where the left coronary artery passed posterior to the pulmonary artery (8 of 28).

Table 2 lists 18 patients with both coronaries arising from the left sinus of Valsalva. All died of known, un-

Table 2*Both Coronaries from Left Sinus of Valsalva*

No.	Age	Sex	Number of coronary ostia	Cause of death and comments
1.	76	M	Single	Coronary artery disease. Heart block and uremia
2.	28	M	Single	Coronary artery disease Thrombus in left main coronary artery and left anterior descending
3.	80	F	Single	Cancer of the rectum. Coronary artery disease
4.	61	M	Double	Coronary artery disease
5.	69	M	Double	Lutetic aneurysm. Infected thrombus
6.	65	M	Single	Cancer of the colon
7.	68	M	Double	Lutetic aortic. Hypertension
8.	61	M	Double	Coronary artery disease
9.	63	F	Double	Bacterial endocarditis mitral valve
10.	77	F	Single	Coronary artery disease. Myocardial infarction
11.	70	M	Double	Dissection of the aorta
12.	72	M	Double	Cerebrovascular accident. Pulmonary embolus
13.	25	M	Single	Rupture posterior sinus of Valsalva
14.	59	M	Double	Emphysema
15.	45	M	Single	Coronary artery disease
16.	59	M	Double	Cancer of the esophagus
17.	53	F	Single	Aneurysm ascending aorta
18.	28	M	Single	Navigator killed in air crash

related causes. Table 3 summarizes the age range, the mean ages, the sex ratio and the number of coronary ostia in each group. The mean age of patients dying suddenly, 20.0 years, is significantly younger than those dying of known causes, 57.3 years ($P < 0.001$).

All patients who died suddenly were males, both in the AFIP series and in the cases collected from the literature (table 4).

Discussion

The single coronary artery and other minor variations of distribution and origin have been regarded as congenital anomalies of little clinical significance. It is evident from a review of the literature (table 4) as well as the data presented that this is not always so. There is a significant number of young patients who die suddenly during or after exercise. In these cases the coronary arteries arise either from the same vessel or as separate vessels from the anterior sinus of Valsalva (fig. 2), with the main left

coronary artery passing posterior and leftward between the pulmonary artery and the aorta before bifurcating.

Where the single coronary or both coronaries arise from the left coronary sinus and the right coronary artery passes anteriorly between the pulmonary artery and the aorta (fig. 3), there is no instance of unexplained sudden death in either the literature or the experience of the AFIP attributable to this anomaly. Because it appeared to contradict this finding, case 5 of Roberts and Loube (AFIP Accession #96957) was reviewed.⁸ They described a 22-year-old man (case 6 in table 1) who died after a double-time march. His single coronary was reported to arise from the *left* sinus of Valsalva. Reviewing the heart it is evident that both coronary arteries arose from the anterior sinus of Valsalva.

The Walter Reed Cardiology Service has noted one instance in which both coronary arteries arising from the left sinus of Valsalva has been associated with

Table 3*Both Coronaries from Same Sinus of Valsalva*

No.	Mean Age (Range)	Sex M/F	Number of coronary ostia single/double	Sudden unexplained deaths
<i>Both from anterior sinus of Valsalva</i>				
9	20.0 (13 - 36)	9/0	2/7	9
24	57.3 (24 - 87)	22/2	12/12	0
<i>Both from left sinus of Valsalva</i>				
18	59.1 (25 - 80)	14/4	9/9	0

Table 4

Literature Review

No.	Author	Age	Sex	Number of coronary ostia	Originating from sinus of Valsalva	Cause of death
1.	Nicod ¹¹	21	M	Single	Anterior	Died after exercise
2.	Jokl, McClellan, and Ross ⁷	14	M	Single	Anterior	Died after running
3.	Cohen and Shaw ⁶	11	M	Double	Anterior	Died after running. Had myocardial infarction
4.	Jokl et al. ¹²	16	M	Double	Anterior	Died after basketball
5.	Benson ¹⁰	54	M	Double	Anterior	Died while shaving
6.	Benson and Lack ⁵	13	M	Double	Anterior	Died while running
7.	Benson and Lack ⁵	13	M	Double	Anterior	Died while playing basketball

myocardial infarction. A 30-year-old man (D. M. WRAMC #3175-022) sustained an inferior subendocardial myocardial infarction documented by ECG and enzyme changes. Coronary arteriography demonstrated that both coronary arteries arose from the left sinus of Valsalva and were patent vessels of normal caliber and free of any luminal narrowing.

Death is usually sudden in the cases described, often with a history of exertional syncope just before death. In those patients who lived long enough after the acute episode, and especially in those few who have been studied clinically, it is evident that severe, extensive myocardial ischemia or myocardial infarction occurred in the distribution of the entire left coronary system.

Case 1: (WRAMC #3813,018) E. J., a 14-year-old boy, had two episodes of exertional syncope, one in August and the other in November 1973, both without prodromal symptoms and both with transient ST-T wave changes consistent with antero-lateral wall myocardial ischemia. Unlike the first syncopal episode which resolved spontaneously, the second exertional collapse required immediate resuscitation for cardiac arrest with ventricular fibrillation upon hospital arrival. On both occasions, he made a full recovery without any symptoms or physical signs of neurologic or cardiac deficit. A submaximal treadmill test performed in October 1973 achieved a heart rate of 150 beats/minute and was normal. Cardiac catheterization with coronary arteriography in January 1974 revealed both coronary arteries to originate from the anterior sinus of Valsalva with subsequent initial course of the left coronary artery to the left and posteriorly between the aorta and the pulmonary artery (fig. 4). In February 1974 the patient underwent open heart surgery to enlarge the observed slit-like orifice of the left coronary artery. He has done well since surgery.

The pathophysiologic mechanism of sudden myocardial ischemia in this anomaly is not clear. There are certain observations from this study which

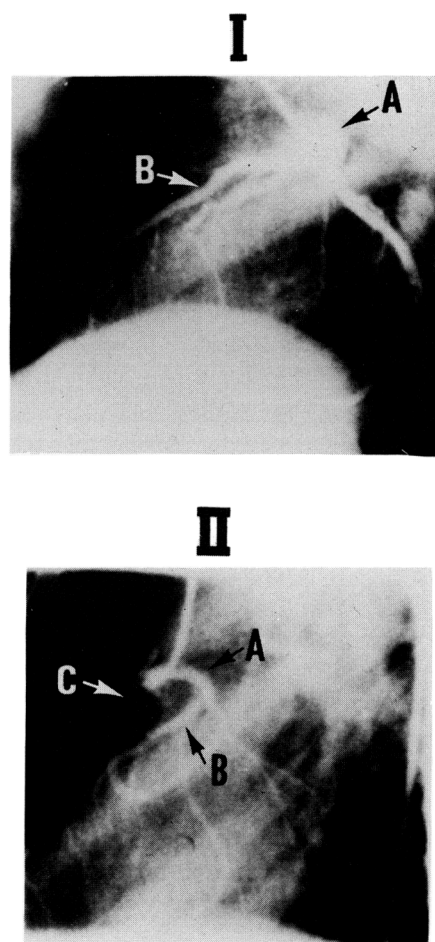


Figure 4

I) Coronary arteriogram, left lateral view, of normal vessels. Injection into left coronary artery. Note posterior position of catheter tip and slightly posterior passage of the main left coronary artery before bifurcation. Contrast material has spilled into the left sinus of Valsalva. A = main left coronary artery; B = left anterior descending coronary artery. II) Coronary arteriogram, left lateral view, of case 1 (WRAMC #3813, 018). Injection into left coronary artery. Note anterior position of catheter tip and the markedly posterior sweep of the main left coronary artery (A) before bifurcation. B = left anterior descending coronary artery; C = balloon catheter in main pulmonary artery.

must be considered in any explanation for the sudden development of myocardial ischemia.

1. Only cases with the left coronary artery arising as a single or double vessel from the anterior sinus of Valsalva (fig. 2), where the left coronary artery passes leftward and posteriorly between the aorta and the pulmonary artery, are associated with sudden death.

2. When the left anterior descending coronary artery passes anterior to the pulmonary artery, a condition seen most often in tetralogy of Fallot when the anterior descending artery arises from the right coronary artery, there has been no report of sudden, unexplained death.

3. There are specimens where the left coronary artery, immediately after arising from either the aorta or the common coronary artery, becomes intramyocardial, traversing the superior aspect of the crista supraventricularis in a subendocardial position (fig. 5). At the septum the artery again becomes epicardial and continues as the left anterior descending artery in the normal position. There are five such specimens in our laboratory, all occurring in cases where the coronaries arose from the anterior sinus of Valsalva. One of these was the heart of a 36-year-old man who had had a documented episode of ventricular tachycardia and who later died suddenly, probably related to the recurrence of the arrhythmia.

4. Every patient with this anomaly who died suddenly and unexpectedly was male. Although the anomaly occurs in females, there is no instance in the literature of a female with this anomaly dying

suddenly and unexpectedly. In spite of the male bias of the AFIP material, there are 6 females in the 51 cases (11.8%) we are presenting. Smith collected 27 adult patients with single coronary arteries; 8 of these were female, 18 were male, and in one the sex was unknown.⁹

5. Most of the sudden deaths from both the literature review and the present series occurred during or immediately following physical exercise. Of the total of 16 patients who died suddenly and unexpectedly, 13 (81.3%) were engaged in or had just stopped physical exercise.

6. Of the 16 patients who died suddenly 10 were in the second decade and four were between the ages of 20 and 22 years at the time of death. Only two were older than 22 years of age. There is no reported instance of a child less than 11 years of age without other congenital cardiac defects having died suddenly with this coronary anomaly.

There have been several mechanisms proposed to explain the cause of sudden death in the cases previously reported. Cohen and Shaw⁶ and Benson and Lack⁵ related the sudden death to the position of the left main coronary artery between the aorta and the pulmonary artery. They postulated that a squeezing action on the left coronary artery occurred during exercise with sudden interference with left coronary arterial flow. In the absence of lesions causing pulmonary hypertension it seems unlikely that a low pressure pulmonary artery could compress a coronary artery distended with systemic pressure.

Benson and Lack⁵ and Jokl et al.⁷ found the left coronary artery smaller than the right in some of their cases and suggested that the smallness of the artery limited coronary blood flow during exercise.

Jokl et al.⁷ also suggest that the mechanism of death might be sudden kinking of the long left coronary artery. In their case there was a sharp hairpin bend in the left main coronary artery which might have kinked with exercise and vigorous expansion of the pulmonary artery and aorta. The hearts we have examined, however, show only a long left main coronary artery which is usually closely applied to the aortic wall without unusual bending or kinking.

Finally, we propose that as the left coronary artery arises from the anterior sinus of Valsalva, it forms an acute angle at its origin which may compromise its lumen as it follows the contour of the aorta leftward and posteriorly. With increased expansion of the aorta during exercise it is possible that the already slit-like opening of the left coronary artery is actually occluded by a flap-like closure of the orifice (fig. 6). Supporting this theory is the fact that in two cases in the literature,^{5, 10} as well as in our clinical case, the left coronary artery adheres so closely to the aorta that the

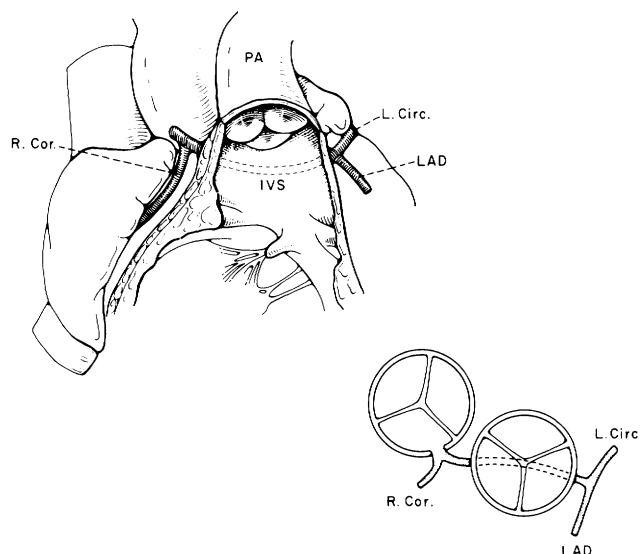


Figure 5

Origin of both coronary arteries from the anterior sinus of Valsalva with left coronary subendocardial passage (left coronary tunnel) through the interventricular septum (IVS).

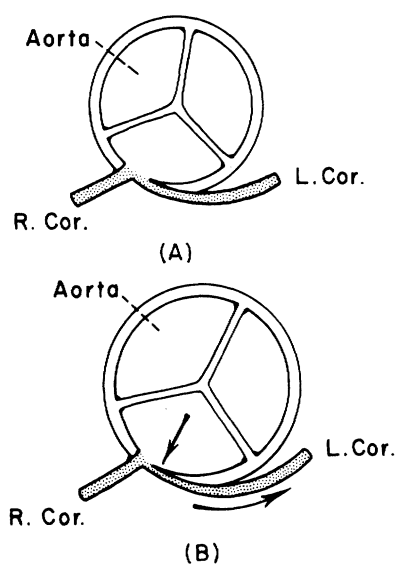


Figure 6

Suggested mechanism of ostial closure with expansion of the aorta and pull on the left coronary artery. Flap-like closure of the ostium by the coronary arterial wall is postulated. L. Cor. = left main coronary artery.

initial part of the coronary artery is actually incorporated in the wall of the aorta. The coronary artery and the aorta share the same media without an adventitia separating them. To show that this is not an invariable sign of poor prognosis, however, we have one specimen similar to this from a 67-year-old man who died of bronchogenic carcinoma (case 23, table 1).

The following case illustrates the importance of the left coronary artery originating from the anterior sinus of Valsalva to the sudden death syndrome.

Case 2: (AFIP #1,478,156) The patient was a 17-year-old male athlete with a previous history of syncope on exercise which had required resuscitation. Immediately after a competitive race, he collapsed and died. At autopsy, the left coronary artery was seen to arise from the anterior sinus of Valsalva and immediately divide into the anterior descending and circumflex arteries. The circumflex artery passed acutely leftward while the anterior descending passed leftward and inferiorly. The right coronary artery arose from the noncoronary sinus of Valsalva. On inspection, the ostium of the left coronary artery appeared to be narrowed and obstructed by the oblique course of the artery; by moving the aortic wall outward for only a distance of 1-2 mm, the aortic wall and the wall of the left circumflex could easily be approximated, effectively closing the orifice (fig. 6). It would therefore appear that the compromise of the left coronary artery produced by the acute angulation which occurs at its origin from the anterior sinus is basic to the mechanism of sudden death and un-

related to the site of origin of the right coronary artery.

In the clinical case (case 1) there was noted at surgery an acutely angulated left coronary artery with a slit-like orifice. Before surgery, because of the suspected inaccessible position of the left coronary artery posterior to the pulmonary artery, it was thought that the only procedure possible would be a saphenous vein or internal mammary artery bypass of both the anterior descending coronary artery and the circumflex artery. Fortunately, the takeoff of the left coronary artery was high enough to be above the sinus of Valsalva and far enough removed from the commissure not to involve the aortic valve. The initial part of the left coronary artery was applied very closely to the aorta and was essentially in the wall of the aorta. It was therefore possible to enlarge the opening into the left coronary artery by wedging out the common wall and constructing a funnel-like opening to the left coronary system. Whether or not this operation will be effective in preventing difficulty in the future depends on the accuracy of the mechanism of sudden death postulated. At the present time in the short term follow-up, the patient continues to do well.

Our patient represents the second clinically recognized case of single coronary artery associated with myocardial ischemia studied during life (Usher and McGranahan, personal communication), and the first where an attempt was made to correct the anatomy which had resulted in intermittent myocardial ischemia.

This is a rare anomaly. In the AFIP accessions, which from 1917 to the present approximate 475,000 autopsy cases, there are 33 cases of both coronary arteries arising from the anterior sinus of Valsalva. Nine of these patients died suddenly and unexpectedly.

This anomaly does represent a cause of sudden, unexpected death and should be considered in any young male with exertional syncope or chest pain accompanied by unexplained QRS or ST-T wave changes.

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