

CASE REPORT

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Sudden Death Due to Coronary Artery Anomalies: A Case Report and Clinical Review

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ABSTRACT: Congenital anomalies of the coronary arteries are relatively uncommon conditions with an incidence of approximately 2% in the adult population. Although less common than acquired coronary artery disease, these anomalies may be associated with myocardial ischemia and its consequences; angina, arrhythmia, infarction, and sudden death. A 21-year-old female patient with no significant prior medical history was found dead at home. Postmortem examination revealed high take-off of the right coronary artery with acute downward angulation of the proximal right coronary artery and acute downward angulation of the left main coronary artery. Microscopic examination revealed global myocardial ischemia consistent with a terminal ventricular dysrhythmia. There was no evidence of any other disease processes. Detailed toxicological investigation was negative. The Regional Forensic Pathology Unit experience with sudden death due to congenital coronary artery anomalies is presented along with a review of the current literature.

KEYWORDS: pathology and biology, sudden death, coronary artery, congenital anomalies

Congenital anomalies of the coronary arteries are relatively uncommon conditions with a reported incidence of approximately 2% in the adult population [1–3]. The true incidence is probably higher and varies with different series depending on whether they are based upon clinical, angiographic or pathological studies. These anomalies are commonly reported in the pediatric population [4–6] but their significance in the adult population has only recently been recognized. Although less common than acquired coronary artery disease, these anomalies can be associated with myocardial ischemia and its consequences; angina, arrhythmia, infarction, and sudden death. This paper presents a previously healthy 21-year-old female athlete who was found dead at home. The Regional

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Forensic Pathology Unit experience with sudden death and anomalous coronary arteries is discussed along with a review of the literature.

Case Report

D. K., a previously healthy 21-year-old woman had returned home after participating in a basketball game and had retired to bed at approximately 11 p.m. She was found at 7 a.m. the following morning by her common-law husband who was unable to awaken her. No medications were found at the scene.

Postmortem Findings

At autopsy the body was clothed in a nightgown and underwear. There was no external evidence of violence. Gross and microscopic examination was unremarkable with the exception of the heart. Detailed toxicological examination was negative.

The 245 g heart was of normal size and configuration. There was no chamber enlargement and all four valves appeared free of any disease process. However, there was an abnormal high take-off of the right coronary artery (RCA). The inferior margin of the right coronary ostium was situated 1.3 cm above the sinotubular junction of the right aortic sinus of Valsalva (Fig. 1). The ostium was also displaced anteriorly such that its margin lay above the commissure between the right and left coronary cusps of the aortic valve. Ostial ridge formation was also identified at the lower limbus of the orifice. There was a steep downward angulation of the proximal 1 cm of the RCA, which then made a sharp lateral, right angle turn into the right atrioventricular groove (Fig. 2). From this point the RCA followed a normal course.

The left coronary artery arose in the center of the left aortic sinus of Valsalva immediately below the sinotubular junction. The proximal left main coronary artery (LMCA) also followed a steep downward angle of approximately 30° between the LMCA and aortic sinus wall. The distribution of the left coronary was otherwise normal. All vessels were free of atherosclerosis. There were no focal lesions seen in the myocardium.



FIG. 1—High take-off of the right coronary artery above the sinotubular junction of the right aortic sinus of Valsalva.

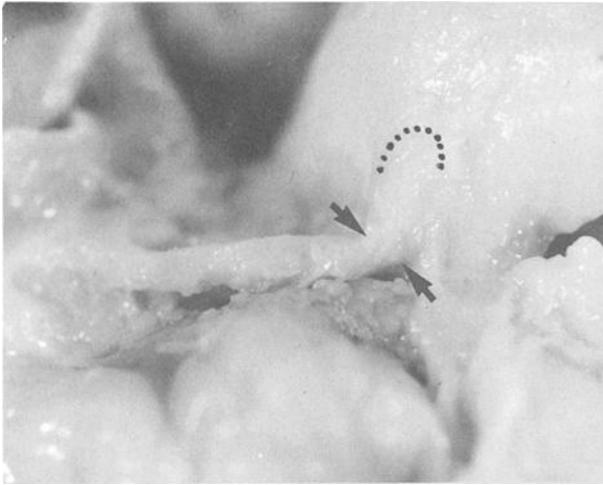


FIG. 2—External view of aortic root. The proximal right coronary artery follows a steep downward angle from the ostium (dots) before making a sharp right angle turn (arrows) into the atri-ventricular groove.

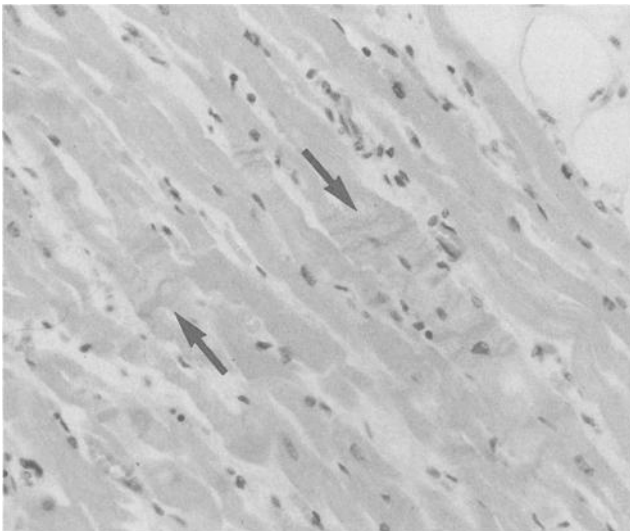


FIG. 3—Photomicrograph of myocardium showing multiple foci of contraction band necrosis (arrows) reflecting terminal ischemia. Hematoxylin and eosin.

Microscopically, there was no established myocardial infarction, myocarditis, or scarring. There was evidence of widespread global ischemia in the form of intense eosinophilia, wavy degeneration and contraction band necrosis consistent with a terminal ventricular dysrhythmia (Fig. 3).

In the absence of any other natural disease process and/or trauma in addition to negative toxicology we concluded that the cause of death was due to congenital anomaly of the coronary arteries.

Discussion

There have been many reports of sudden death attributed to congenital anomalies of the coronary arteries [1,5,6,10-13]. Although more commonly described in the pediatric population the importance of such anomalies as a cause of sudden death in the adult population has been recently recognized.

Coronary anomalies can be classified on the basis of their origin, their course and their termination. During routine postmortem examination it is important to note the location and the angle of take-off of the coronary arteries as well as identify the presence of ostial valvelike ridges. The relationship between these anatomical structures may be a triggering mechanism of angina and sudden death.

High take-off of the coronary artery occurs when the coronary artery arises above the sinotubular junction. There is controversy as to whether this anomaly is associated with sudden cardiac death.

The coronary artery is considered to have an acute angle take-off if the angle between the proximal coronary artery and the aortic wall is less than 45° [12]. An ostial valvelike ridge was considered to be significant if the surface area of the ridge exceeded 50% of the coronary ostial luminal area [12]. This case illustrates all three of the above anomalies, although the area of the ridge was less than 50%.

Virmani and colleagues examined the hearts of 41 patients consisting of 22 cases of sudden death and 19 controls with known causes of death [12]. The incidence of acute angle take-off was 59% in those patients who died suddenly versus 21% in controls ($P = 0.015$). The incidence of ostial valvelike ridges was 41% in cases of sudden death versus 11% in the control group ($P = 0.031$).

It has been postulated that aortic root dilatation may compress coronary arteries with acute angle take-off and that ostial valvelike ridges may act as occlusion valves. Either mechanism can cause acute obstruction of the proximal coronary artery and lead to sudden death. Table 1 reveals the incidence of coronary artery anomalies identified at postmortem examination at our center between January 1990 and January 1992. In five of the nine cases, the cause of death was attributed to congenital anomalies of the coronary arteries with two out of the five cases showing concurrent finding of coronary artery disease. This represents an incidence of 0.8% among 1050 consecutive autopsies.

These findings and those of other authors indicate that these congenital anomalies of the coronary arteries are an important cause of sudden death in otherwise healthy young patients. As such, their presence should be suspected and with careful inspection the incidence of such anomalies will likely be found to be higher than currently reported in the literature.

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TABLE 1—Incidence of anomalous origin of coronary arteries at the regional forensic unit—H.G.H. January 1990–January 1992.

Case no.	Age/yrs Sex	Clinical history	Circumstances	Coronary anomaly	Coronary and/or other heart disease	Cause of death
1	67 F	Emphysema	Pedestrian struck by a vehicle Died four hours after accident	LMCA from RSV coursing posterior	CAD	Trauma
2	61 M	Flu-like illness	Found dead in bed	LMCA from RSV coursing posterior with acute angle take-off	None	Coronary anomaly
3	57 F	Diabetes, hypertension and unstable angina	Died on the OR table following aortic coronary bypass surgery	LMCA—acute angle take-off with crescentic ridge	CAD	Acute myocardial ischemia (postoperative)
4	63 M	Chronic alcoholism and heavy smoking	Found dead on the kitchen floor lying on his back	RCA and LMCA. Both high take-off	Calcific aortic stenosis	Acute alcoholic poisoning
5	13 M	Tourette's syndrome	Found wedged inside an outhouse toilet located in a conservation park	RCA, LMCA from LSV with high take-off of RCA and acute downward angulation	Interstitial myocarditis	Suspension by steel bar
6	55 M	Manic depression	Found dead in hallway of apartment building	RCA—high take-off with acute angle take-off. LMCA—acute angle take-off with ostial valve ridge	Moderate CAD	Coronary anomaly and CAD

7	28 M	None	Sudden collapse following emotional and physical stress Found dead in bed	RCA, LMCA—high take-off with obstruction of ostia of LMCA LMCA—acute angle take-off with ostial ridge. RCA—slitlike ostia	Unicuspid and moderately stenosed aortic valve CAD	Arrythmia due to anomalies of coronaries and aortic valve Coronary anomaly and CAD
8	36 F	None	Found dead in bed	LMCA—acute angle take-off with ostial ridge. RCA—slitlike ostia	CAD	Coronary anomaly and CAD
9	45 F	None	Sudden collapse on entering the water (an attempt for scuba diving)	RCA—acute angle take-off with ostial stenosis. LMCA—acute angle take-off with ostial valve ridge	None	Coronary anomaly

NOTE: F—female. M—male. LMCA—left main coronary artery. RSV—right aortic sinus of valsalva. RCA—right coronary artery. CAD—coronary artery disease.

References

- [1] Kragel, A. H. and Roberts, W. C., "Anomalous Origin of Either the Right or Left Main Coronary Artery from the Aorta with Subsequent Coursing Between Aorta and Pulmonary Trunk: Analysis of 32 Necropsy Cases," *American Journal of Cardiology*, Vol. 62, No. 10, 1988, pp. 771-777.
- [2] Teplitsky, L., Wurzel, M., Melamed, R., and Aygen, M., "Anomalous Origin of the Coronary Arteries," *Angiology*, Vol. 38, 1987, pp. 128-132.
- [3] Yamanaka, O. and Hobbs, R. E., "Coronary Artery Anomalies in 126, 595 Patients Undergoing Coronary Arteriography," *Catheterization and Cardiovascular Diagnosis*, Vol. 21, No. 1, 1990, pp. 328-340.
- [4] McManus, B. M., Gries, L. A., Ness, M. J., and Galup, L. N., "Anomalous Origin of the Right Coronary Artery from the Left Sinus of Valsalva," *Pediatric Pathology*, Vol. 10, No. 6, 1990, pp. 987-991.
- [5] Lauridson, J. R., "Sudden Death and Anomalous Origin of the Coronary Arteries from the Aorta. A Case Report and Review," *American Journal of Forensic Medicine and Pathology*, Vol. 9, No. 3, 1988, pp. 236-240.
- [6] Ness, M. J. and McManus, B. M., "Anomalous Right Coronary Artery Origin in Otherwise Unexplained Infant Death," *Archives of Pathology and Laboratory Medicine*, Vol. 112, No. 6, 1988, pp. 626-629.
- [7] Greenberg, M. A., Fish, B. G., and Spindola-Franco, H., "Congenital Anomalies of the Coronary Arteries," *Radiologic Clinics of North America*, Vol. 27, No. 6, 1989, pp. 1127-1146.
- [8] Cheitlin, M. D., "Coronary Arterial Anomalies: Clinical and Angiographic Aspects," *Nonatherosclerotic Ischaemic Heart Disease*, R. Virmani and M. B. Forman, Eds., Raven Press, New York, 1989, pp. 125-152.
- [9] Virmani, R., Rogan, K., and Cheitlin, M. D., "Coronary Arterial Anomalies: Pathologic Aspects," *Nonatherosclerotic Ischaemic Heart Disease*, R. Virmani and M. V. Forman, Eds., Raven Press, New York, 1989, pp. 153-183.
- [10] Hanzlick, R. L. and Stivers, R. R., "Sudden Death Due to Anomalous Right Coronary Artery in a 26-year-old Marathon Runner," *American Journal of Forensic Medicine and Pathology*, Vol. 4, No. 3, 1983, pp. 265-268.
- [11] Tuna, I. C., Blanton-Bessinger, F., Ophoven, J. P., and Edwards, J. E., "Acute Angular Origin of Left Coronary Artery from Aorta: An Unusual Cause of Left Ventricular Failure in Infancy," *Pediatric Cardiology*, Vol. 10, 1989, pp. 39-43.
- [12] Virmani, R., Chun, P. K. C., Goldstein, R. E., Robinowitz, M., and McAllister, H. A., "Acute Take-offs of the Coronary Arteries Along the Aortic Wall and Congenital Coronary Ostial Valve-like Ridges: Association with Sudden Death," *Journal of the American College of Cardiology*, Vol. 3, No. 3, 1984, pp. 766-771.
- [13] Taylor, A. J., Rogan, K. M., and Virmani, R., "Sudden Cardiac Death Associated with Isolated Coronary Artery Anomalies," *JACC*, Vol. 20, No. 3, Sept. 1992, pp. 640-647.

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