## CASE REPORT

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# Sudden Unexpected Death Resulting from an Anomalous Hypoplastic Left Coronary Artery*

**REFERENCE:** McConnell, Collins KA. Sudden unexpected death resulting from an anomalous hypoplastic left coronary artery. J Forensic Sci 1998;43(3):708–711.

**ABSTRACT:** We present a case of sudden death in a 24-year-old, healthy white female who was physically active and participated in sports, including soccer. Two weeks prior to her death, an insurance physical examination revealed an abnormal electrocardiogram which demonstrated flipped T waves in the anterior leads. There was no other remarkable medical history.

At autopsy, only one coronary ostium was demonstrated and it originated from the right aortic sinus. Approximately 0.8 cm from this right coronary artery (RCA) ostium, a left coronary artery (LCA) branched off the RCA at a 90-degree angle. The LCA had luminal diameter of 0.4 m but the LCA had a luminal diameter of only 0.1 cm. The LCA coursed anterior to the base of the pulmonary artery and down the anterior ventricular septum reflecting the usual course of the left anterior descending (LAD) coronary artery. The LCA and RCA paths appeared to merge or terminate at the anterior left ventricular myocardium which was discolored gray, a process that involved the inner and middle thirds of the myocardium.

Based on the autopsy findings, we certified the cause of death as a probable arrhythmia due to myocardial fibrosis and dystrophic calcification resulting from complications of an anomalous hypoplastic left coronary artery.

Anomalies of coronary arteries have been classified and studied at autopsy and by clinical angiography. Coronary artery anomalies can be divided into minor and major forms with major anomalies often resulting in cardiac dysfunction that may cause failure and death. Minor anomalies, in general, have no pathophysiological significance and are compatible with life. Minor anomalies include variations in number and location of coronary ostia. A single coronary ostia is exceedingly rare in hearts with no other congenital malformations. The prognostic significance can be unpredictable. A single coronary artery has the potential to be dangerous if obstructed at its main stem, or if it branches at an acute angle. Additionally, hypoplasia of one or more coronary arteries has been found to be associated with sudden death.

**KEYWORDS:** forensic science, forensic pathology, sudden death, anomalous coronary artery, hypoplastic coronary artery

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*Presented at the 49th annual meeting of the American Academy of Forensic Sciences, New York, New York, 17–22 February 1997.

Received 25 July 1997; and in revised form 4 Sept. 1997; accepted 8 Sept. 1997.

### Case History

The decedent was a 24-year-old woman who collapsed while drinking coffee with her friends at 8:50 p.m. She reportedly experienced seizure-like activity followed by labored breathing. EMS was called at 8:53 p.m., and cardiopulmonary resuscitation was begun after she became unresponsive. EMS arrived at 9:05 p.m., and the decedent was noted to be in ventricular fibrillation; advanced cardiac life support was initiated. The decedent was defibrillated three times and given epinephrine, naloxone, bicarbonate, lidocaine, and atropine en route. She arrived at the hospital at 9:29 p.m. and was in asystole. Despite continued aggressive resuscitative efforts, she was pronounced dead at 9:50 p.m.

Two weeks prior, the decedent underwent an insurance physical examination. An echocardiogram performed at that time was abnormal, demonstrating flipped T waves in the anterior leads. The decedent was otherwise healthy with no history of headaches, seizures, chest pain, or exertional angina. She was physically active and participated in sports, including soccer. There was questionable history of marijuana use; however, no other drugs, including cocaine and heroin, were reportedly abused.

### Autopsy Finding

#### *Gross*

The decedent was approximately 152 cm in height and 53 kg. She exhibited mild head and neck cyanosis; the external examination was otherwise unremarkable.

The heart weighed 300 grams. One coronary ostium was within the right aortic sinus (Fig. 1). A left ostium was not present, neither in its expected position nor in an anomalous one. The left aortic sinus was smooth, with no dimple nor blind pouch.

The right coronary artery (RCA) had a luminal diameter of 0.4 cm. The RCA followed its usual course, running between the pulmonary trunk and right auricle, descending in the atrioventricular groove, and supplying branches to the right atrium and ventricle. It continued to the apex of the heart with branches to the right marginal coronary artery then became the posterior descending coronary artery.

Approximately 0.8 cm from the RCA ostia, an anomalous, hypoplastic left coronary artery (LCA) with a luminal diameter of 0.1 cm branched off the RCA at an angle of 90°. The LCA traveled anterior to the base of the pulmonary artery and down the anterior ventricular septum, reflecting the usual path of the left anterior

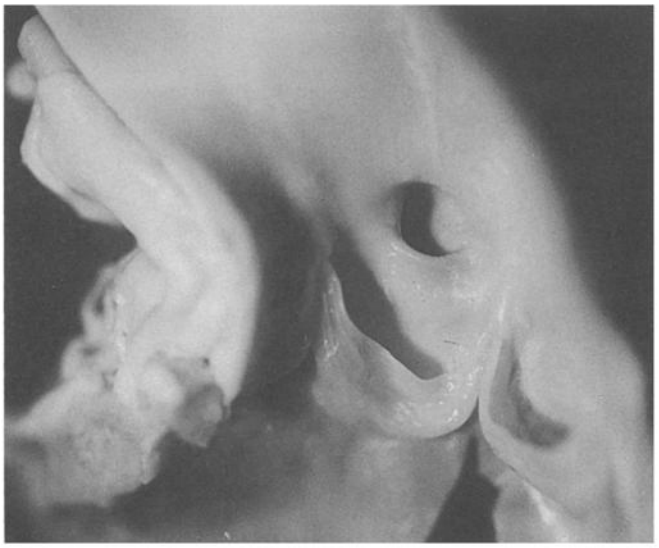

FIG. 1—*Right coronary ostium originating in the right aortic sinus. Note absence of a left coronary ostium.*

FIG. 2—*Anterior left ventricle with fibrosis and hypoplastic left coronary artery.*

descending artery. The left circumflex artery distribution appeared to be supplied by small branches from the LCA and posterior descending artery. No evidence of coronary atherosclerosis was identified.

The inner and middle thirds of the anterior left ventricular myocardium, where the LCA and RCA circulatory paths appeared to merge or terminate, were discolored gray (Fig. 2).

No other congenital anomalies were identified. The atria were normally formed with a probe patent foramen ovale. The papillary muscles were well developed. The left and right ventricles measured 1.4 cm and 0.4 cm in thickness, respectively. The valves were normally formed with slight thickening of the mitral valve. The great vessels arose from the heart in a normal fashion. The descending aorta was unremarkable.

### Microscopic

Multiple histologic sections of the heart confirmed the gross findings of a hypoplastic LCA with an abnormally small luminal diameter. Sections from the left ventricle showed large areas of fibrosis replacing the myocardium, a finding highlighted by a Mason's trichrome stain (Fig. 3). Surrounding myocytes exhibited hypertrophic changes with large hyperchromatic nuclei (Fig. 4*a* and *b*). Several foci of dystrophic calcification with fibrosis were noted in the anterior left ventricle (Fig. 5). Intimal proliferation of intramyocardial vessels was evident.

Additional findings included accumulation of hemosiderin-laden macrophages within pulmonary alveoli. Sections of the cerebrum revealed neuronal subacute hypoxic changes.

### Toxicology

Toxicological analysis of blood was negative for ethanol and other volatiles. A general urine drug screen was positive only for caffeine.

### Conclusion

The cause of death for this 24-year-old white female was cardiac arrhythmia due to regional myocardial fibrosis and calcification resulting from an anomalous, hypoplastic left coronary artery. In this case, the abnormal left coronary artery was an isolated congenital anomaly. The manner of death was natural.

### Discussion

Anomalies of the coronary arteries, which have been classified and studied at autopsy and by clinical angiography, can be divided into minor and major forms. Major coronary anomalies often result in cardiac dysfunction and may cause failure and death. Minor anomalies tend to have no pathophysiological significance and most are compatible with normal longevity. Minor anomalies include alterations in the number and location of coronary ostia. A single coronary artery is rare in hearts without other congenital malformations (1). The incidence of anomalous origin of the LCA has been estimated at 1/300,000 births (2).

The prognostic significance can be unpredictable. Although a coronary artery anomaly may be an incidental finding, it is also an acknowledged cause of sudden death (3). A single coronary artery has the potential to be dangerous if obstructed at its main stem or if it branches at an acute angle. However, acute angulation is not common when there is a single coronary ostium or when the LCA branches from the RCA as compared to when there is anomalous origin of a coronary artery from the aorta (4). Sudden deaths and exercise related deaths involving the LCA arising from the RCA are often associated with leftward passage of the LCA between the aorta and pulmonary artery, resulting in a slit-like lumen (1,5). Furthermore, compression of the LCA between the expanding aorta and pulmonary arteries during exercise could exacerbate this condition (6). However, these physical and anatomical changes were not present in this case. There was no evidence of increased physical activity prior to the death, and, more importantly, the anomalous LCA passed anterior to the pulmonary artery. Furthermore, no acute angulation of the LCA was identified. However, in the decedent's heart, the LCA luminal diameter was hypoplastic with a luminal diameter of 0.1 cm. The normal adult LCA luminal diameter ranges from 0.2 to 0.55 cm with an average of 0.4 cm (7). In this case, the myocardium with the poorest circulatory supply from the left and right coronary arteries, the anterior left ventricle, demonstrated the chronicity of this poor circulation both grossly and microscopically as fibrosis, dystrophic calcification, and myocyte hypertrophy. It was apparent that the myocardium suffered long-standing hypoxic and ischemic damage, even

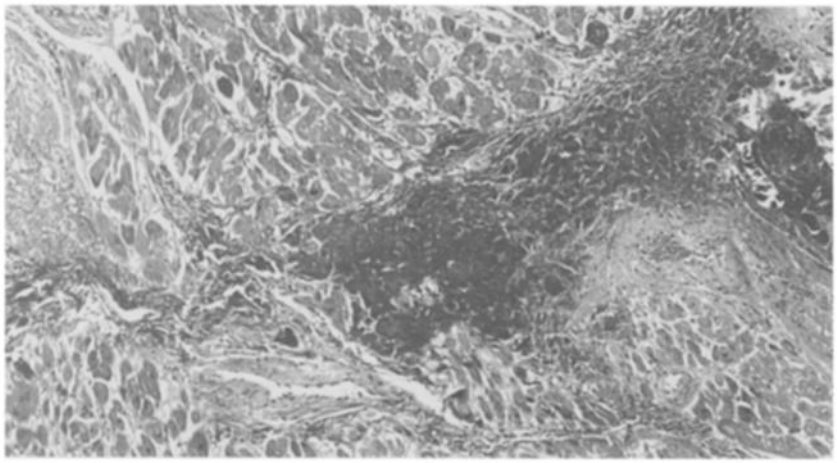

FIG. 3—*Masson's Trichrome stain, counterstained with hematoxylin and eosin, demonstrating myocardial fibrosis (×100).*

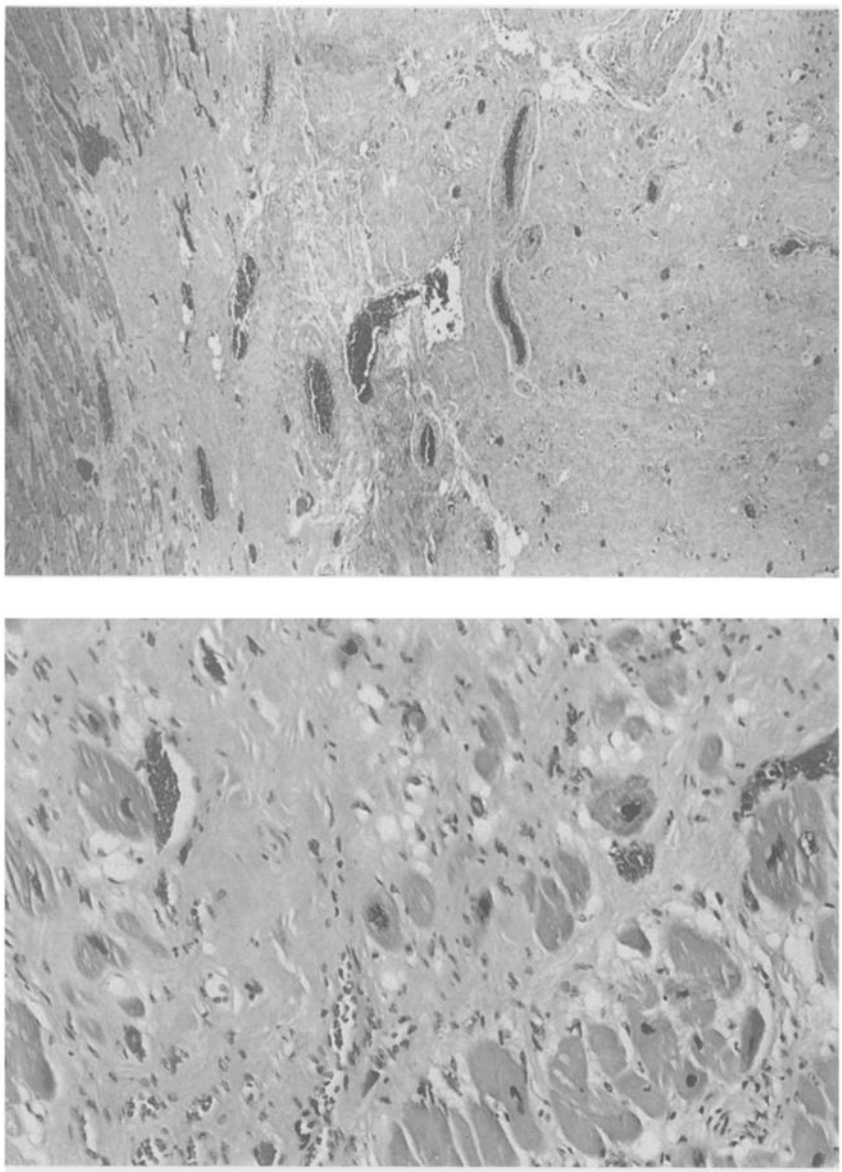

FIG. 4—*Myocardial fibrosis and myocyte hypertrophy within sections of the anterior left ventricle (4a ×40; 4b ×200).*

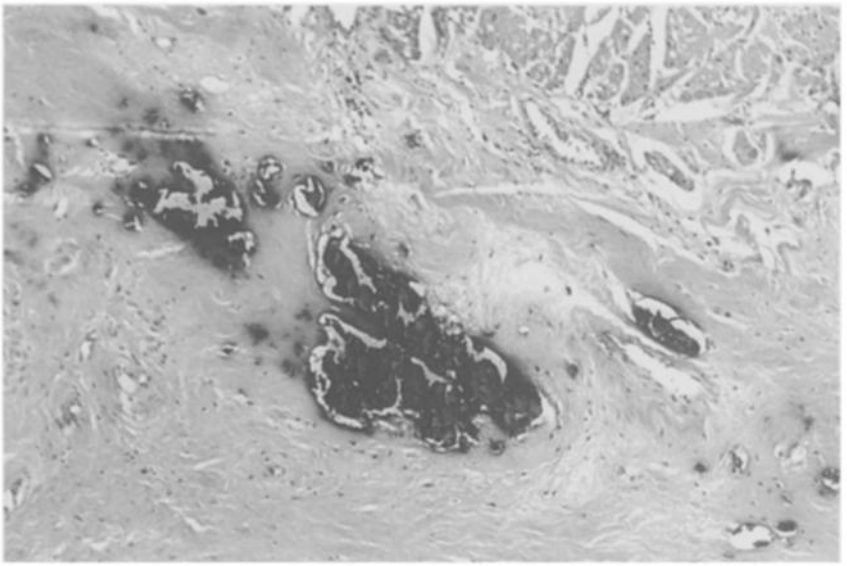

FIG. 5—*Intramyocardial dystrophic calcification and areas of myocardial fibrosis (×100).*

though, as with many cases of sudden death, the decedent was asymptomatic (4). Little has been published on the postmortem assessment of hypoplastic coronary arteries. It has been recently stated that "a general definition of coronary artery hypoplasia might simply be a markedly diminutive artery and artery ostium or segment of artery that is inadequate for sufficient blood flow to dependent myocardium" (8). To designate coronary artery hypoplasia as responsible for a death, there should be histologic signs of myocardial ischemia and other causes of death must be excluded (8).

Unexpected death associated with coronary artery hypoplasia is thought to be underreported and may be easily overlooked (9). Therefore, it is vitally important to be aware of the less frequent causes of sudden death, especially in young adults who have no history of significant medical disease. Overall, the more commonly recognized causes of sudden cardiac death include occlusive coronary thrombi, atherosclerotic heart disease, and cardiac conduction defects. In young adults, the predominating causes of sudden death are also of cardiovascular origin (9) and include myocarditis, mitral valve prolapse, and hypertrophic cardiomyopathy. Coronary artery anomalies in particular account for only a small percentage of sudden death, although within young athletes, they are found to account for a much larger percentage (10).

Encountering sudden death in a young adult is always a tragic event and a challenge for the forensic pathologist. Often, the decedent has no preceding medical history to guide the examination. Cardiac disease is quickly considered in both the older and middle aged cases, and it should not be overlooked in the younger group. Additionally, the coronary artery circulation should be meticulously examined for anomalies and hypoplastic artery segments in these cases so as to prevent unnecessarily listing the etiology of death as undetermined, to document clinically undetected disease, and to contribute to the more accurate assessment of the prevalence of these abnormalities.

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