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# Clinical Communications

## EXERCISE-UNRELATED SUDDEN DEATH AS THE FIRST EVENT OF ANOMALOUS ORIGIN OF THE LEFT CORONARY ARTERY FROM THE RIGHT AORTIC SINUS

Gabriella De Rosa, MD,* Marco Piastra, MD,† Manuela Pardeo, MD,* Elena Caresta, MD,† and Arnaldo Capelli, MD‡

*Pediatric Cardiology, Institute of Pediatrics, †Pediatric Intensive Care Unit, and ‡Department of Pathology, Catholic University of Sacred Heart, Rome, Italy

*Reprint Address*: Gabriella De Rosa, MD, Sezione di Cardiologia Pediatrica, Policlinico Universitario "Agostino Gemelli", Università Cattolica del Sacro Cuore, Largo Agostino Gemelli 8, I-00168 Roma, Italy

□ **Abstract—Congenital anomalous origins of the coronary arteries represents a rare but well-described cause of myocardial ischemia and sudden death. Left coronary artery (LCA) arising from the right sinus of Valsalva is a rare congenital coronary anomaly that seems to be commonly associated with sudden death in young trained athletes. The possibility of a coronary artery anomaly should always be considered in young individuals with a history of chest pain or syncope, particularly if the episodes are triggered by exercise. We describe a case of congenital LCA anomaly in an asymptomatic 10-year-old girl with no family history of sudden death; no previous unexplained syncopal episodes or exercise-induced symptoms were reported. She experienced a cardiac arrest while she was resting at school and was not recoverable despite early emergency department admission and intensive prolonged cardiopulmonary resuscitation attempts. Post-mortem pathological findings revealed a single origin from the right sinus of Valsalva for both right and left coronary arteries. The LCA was compressed between the aorta and the pulmonary trunk. Histologic features suggested recent ischemia. Although sudden death can be the first manifestation of this condition, it is important to be particularly aware of prodromic symptoms: exertional dyspnea, chest pain, syncope or dizziness. Recognition during life of this coronary anomaly is mandatory to prevent the risk of sudden death and to plan surgical correction if clinically indicated. © 2005 Elsevier Inc.**

□ **Keywords—anomalous origin of coronary artery; sudden cardiac death; children; pediatrics**

### INTRODUCTION

Sudden cardiac death (SCD) is relatively rare in children, adolescents, and young adults. However, its occurrence has a great impact on the families involved and on the whole community. Congenital anomalous origin of the coronary arteries is a rare but well-described cause of myocardial ischemia and sudden death in young people (1–3).

Left coronary artery arising from the right sinus of Valsalva (ALMCA) is a rare congenital coronary anomaly, with an incidence of 0.017% to 0.03% of angiographic series and 1.3% of the total coronary anomalies (2,4,5). Although this anomaly can be an incidental post-mortem finding and often permits survival into advanced age, it seems to be commonly associated with sudden death, most notably in young athletic individuals (1,6–9).

We present a rare case of such a congenital coronary anomaly in a young girl and discuss the clinical importance of this anomaly and its association with SCD.

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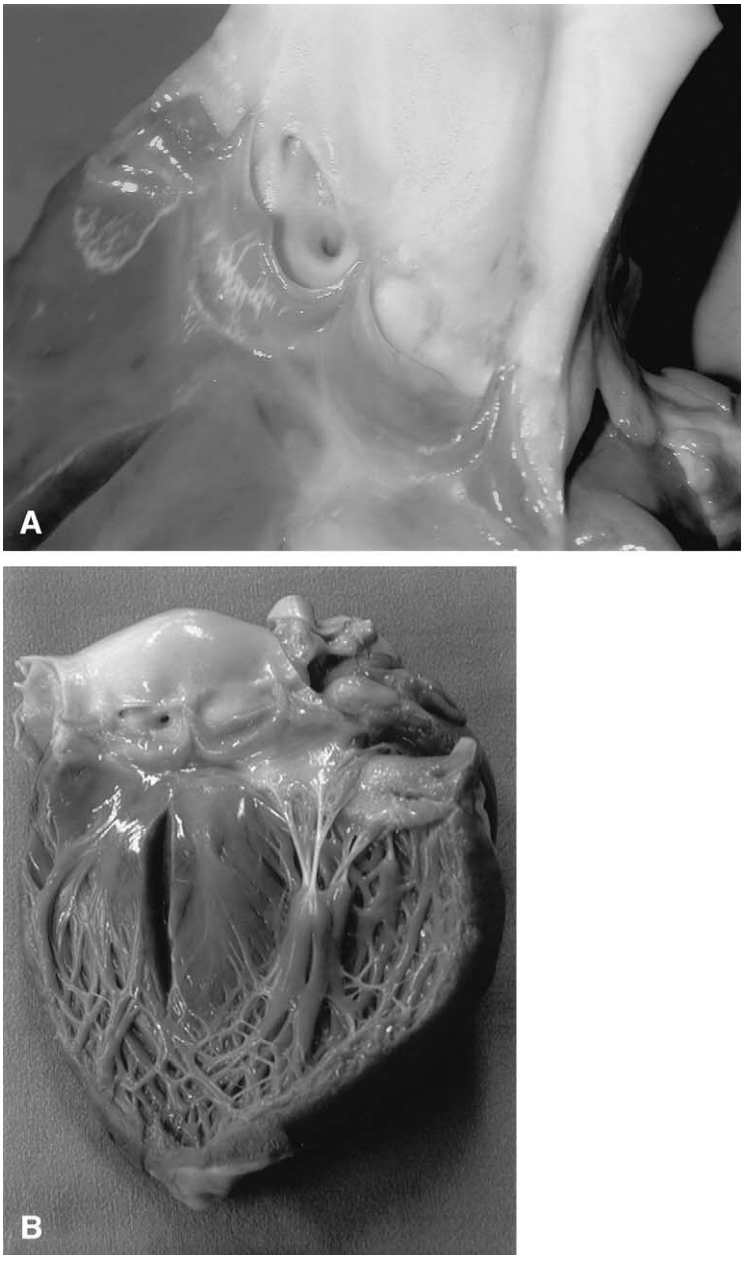


**Figure 1. (a,b) Views of the aorta showing left and right coronary arteries arising from the right coronary sinus.**

## CASE REPORT

A 10-year-old healthy girl from Pakistan, with no previous family history of juvenile sudden death or syncope episodes or exercise-induced symptoms, suffered an out-of-hospital cardiac arrest. She collapsed and became unresponsive and pulseless at school (while she was attending lessons for several hours as confirmed by teachers and bystanders). No signs of electrical burns or toxic ingestion were found by emergency care personnel. Resuscitation by bag-mask ventilation ($FiO_2$ 100%) and chest compressions were immediately started by teachers and then by paramedic staff and she was admitted to the Hospital Emergency Department 15 min later.

On admission, the patient was apneic, unconscious, and unresponsive (Glasgow Coma Scale score 3) with a bradycardic rhythm on the monitor of 30 beats/min; peripheral and central pulses were undetectable, as was blood pressure. Endotracheal intubation and assisted ventilation were rapidly performed together with epinephrine (0.1 mL/kg) administration by the endotracheal route and chest compressions were continued. Foamy

hemorrhagic secretions were suctioned from the tracheal tube. The first available arterial blood gas showed a remarkable metabolic acidosis and hyperkalemia; further laboratory parameters revealed a worsening metabolic acidosis and lactate, and increased transaminases and LDH/CPK levels. A femoral vein was cannulated and epinephrine (0.1 to 0.5 mg/kg) and sodium bicarbonate (1–2 mEq/kg) were administered. Electrocardiographic tracing initially showed severe sinus bradycardia with transient episodes of pulseless electrical activity evolving into ventricular fibrillation episodes requiring various attempts of cardioversion at 2 J/Kg and then at 4 J/Kg; subsequently, persistent asystole developed, unresponsive to repeated epinephrine and bicarbonate infusions. Cardiopulmonary resuscitation was ceased after 40 min of proven asystole.

## PATHOLOGIC FINDINGS

The girl was well developed, well nourished, and without external evidence of congenital anomalies. No effusions were present. Intrathoracic petechial hemorrhages and a subpleural hematoma were found behind the IV–V ribs. Both lungs were increased in weight and consistency due to pulmonary edema. On gross inspection, the heart weight was 150 g and examination revealed biventricular dilatation. Both right and left coronary arteries arose from a single right sinus of Valsalva (Figure 1a and 1b). The right coronary artery was normal in size and supplied the posterior wall of both ventricles. The left coronary artery arose with a normal size but was compressed, passing between the aorta and the pulmonary trunk. This abnormal course led to a loss of its lumen diameter, resulting in a circumflex and anterior descending artery hypoplasia. The septum and the anterior ventricular wall area appeared to have a pale color. Histologic features of the heart included undulated myofibers with nuclear inclusions from both the interventricular septum and the left ventricular anterior wall (Figure 2). Their section revealed a thickened endocardium with areas of fibrosis and vacuolation of the cytoplasma, suggesting recent ischemia. The coronaries presented restricted lumen and adventitial fibrosis without thrombotic phenomena (Figure 3). The screening for hyperhomocysteinemia, thrombophilic states, antiphospholipid syndrome, and hypercholesterolemia were negative.

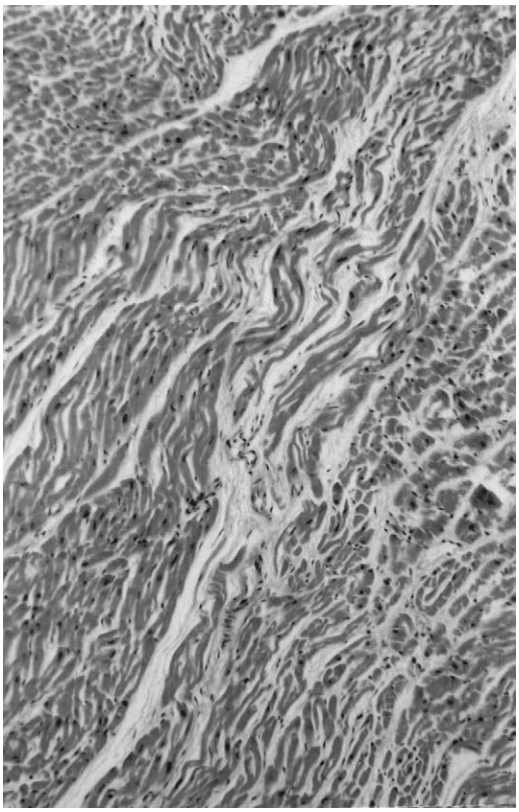

**Figure 2. Histologic section of the region of left ventricular myocardium supplied by the anomalous LCA showing diffuse undulated fibers (25×).**

## DISCUSSION

Sudden unexpected cardiac death in a child or adolescent is a rare but devastating event with serious impact on the family, the school, and the community. Various retrospective reviews were undertaken to determine the incidence of SCD in children. The reported incidence varied from 0.8% to 6.2 cases/100,000 population per year (5,10–13). Recent studies suggest that the incidence of SCD in children and adolescents is increasing (14,15). The reasons for this increase in the frequency are not clear.

Despite numerous reports, there is a paucity of demographic data on sudden death from cardiac causes in the young. Among infants (<1 year old) SCD is usually due to ductus dependent complex congenital cardiac lesions; after the first year and through the third decade of life the most common causes of SCD include myocarditis, hypertrophic cardiomyopathy, coronary artery disease, congenital coronary artery anomalies, conduction system abnormalities, mitral valve prolapse, and aortic dissection (8,11,12).

The most frequent congenital coronary artery anomalies causing sudden death in the young involve an ectopic origin of either the right or left coronary artery in the left or the right aortic sinus of Valsalva, respectively (2).

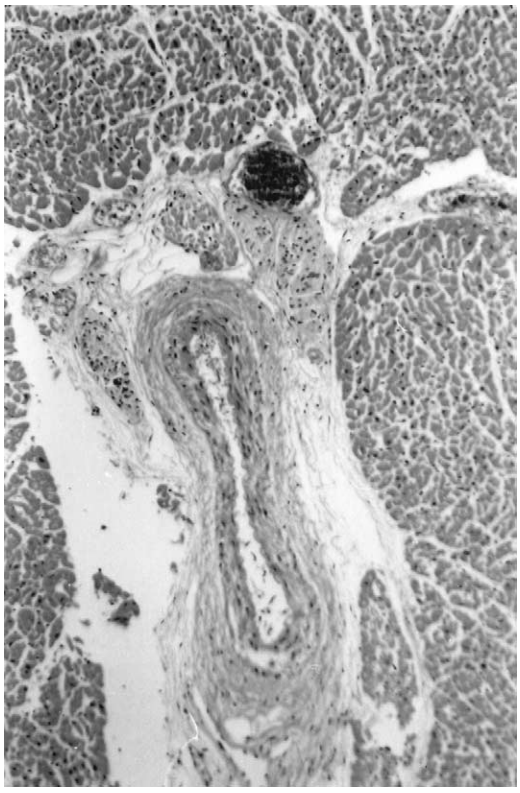

**Figure 3. Histologic section of the left coronary branches showing restricted lumen and adventitial fibrosis without thrombotic phenomena (10×).**

ALMCA is categorized among the potentially serious anomalies. Many autopsy studies have revealed a high coincidence of sudden death due to this anomaly in young athletic males (6–8,16). Virmani et al. reported 49 deaths due to an aberrant LCA from the Right Sinus of Valsalva. Sudden death occurred in 57% of these patients more frequently (64% of cases) during or soon after exercise. Younger patients (<30 years old) were significantly more likely to die suddenly or during exercise than older ones (>30 years old); however only a few patients were <20 years old, and they had higher rates of exercise-related deaths (17).

Three different anatomic types were identified according to the relation of the anomalous LCA with the aorta and pulmonary artery; Type 1 with the aorta passing anterior of the pulmonary artery, Type 2 in which the LCA passes between the aorta and the pulmonary artery with further classification into "interarterial" Type 2A or "septal or intramyocardial" Type 2B, and Type 3 with the LCA passing to the posterior of the aorta. Types 1 and 3 are almost completely benign, and reported SCD is extremely rare. The most serious type of these anomalies is type 2, often associated with a significant risk of SCD (2,9,18).

Because most patients are asymptomatic, this condition is often diagnosed post mortem (18). Cardiovascular symptoms (chest pain, exertional dyspnea, syncope or dizziness) occur in only 18% to 30% of cases (8,16).

An early diagnosis of ALMCA requires clinical suspicion and is particularly important, because surgical correction is feasible (6). The possibility of a coronary artery anomaly should always be considered in a young individual with a history of chest pain or syncope, particularly if triggered by exercise (19). Recent autopsy studies indicate that symptoms such as exertional chest pain or syncope can represent the principal indicators of anomalous origin of a coronary artery from the wrong sinus before death (6). Sudden death is probably related to a blood flow reduction in the anomalous vessel, resulting in myocardial ischemia or infarction, ventricular tachycardia, and fibrillation (2,9).

The most likely ischemic mechanism is an acute angled kinking at the origin of the coronary artery or compression of the anomalous artery between the aorta and pulmonary trunk (2,3). Transthoracic two-dimensional echocardiography and magnetic resonance imaging can be used to reveal the anomaly and diagnostic coronary arteriography ultimately can be performed (6). Patients usually do not have abnormalities on standard 12-lead electrocardiograms, because the myocardial ischemia is episodic, thereby limiting the value of random screening (6).

In a recent prospective study, Davies determined the incidence and clinical significance of major coronary artery anomalies in asymptomatic children using transthoracic two-dimensional echocardiography (20). A higher prevalence was found than in other prospective studies; therefore, identification of coronary artery origin should be included as part of any echocardiographic examination. However, negative findings in patients referred for cardiovascular symptoms do not preclude the need for further evaluation.

Our case represents a rare congenital coronary anomaly in which sudden death occurred at a very young age, unrelated to exercise and without evidence of prodromic symptoms attributable to myocardial ischemia.

In addition, histological examination failed to reveal chronic myocardial ischemic damage, suggesting that SCD may be the first event of this disease. Transthoracic echocardiography with focused two-dimensional and color Doppler imaging of the coronary arteries is an important non-invasive tool for identifying anomalous coronary origin. On the other hand, because a large population screening is not applicable, it is important to be aware of such anomalies in order to suspect an ALMCA in an otherwise healthy young patient presenting with a cardiac complaint.

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