CASE REPORT

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Association of Right Coronary Artery Hypoplasia with Sudden Death in an Eleven-Year-Old Child

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ABSTRACT: Congenital coronary artery abnormalities are a rare but well-documented cause of sudden and unexpected death in the pediatric age group. Most reported cases involve both an aberrant origin and course of the abnormal vessel. A case of unexpected death occurring in an otherwise healthy eleven-year-old boy, who had been previously investigated for sudden collapse, is described. The major finding at autopsy was marked disparity in diameter between the coronary arteries due to diffuse hypoplasia of the right coronary artery. This case demonstrates the importance of meticulous examination of the coronary artery system in cases of sudden death in childhood so that significant reduction in luminal cross section will be adequately documented. The possible role played by this finding in the etiology of sudden death is discussed.

KEYWORDS: pathology and biology, coronary artery system, coronary artery hypoplasia, sudden death, children

Cardiac causes of sudden and unexpected death in childhood most often are related to congenital structural defects of the heart, to cardiomyopathy or to myocarditis [*1*,*2*]. Coronary artery abnormalities may be caused by vasculitis, as in Kawasaki disease, or may be due to congenital structural abnormalities [*3*,*4*]. Although congenital anomalies of the coronary arteries most often involve ectopic coronary ostia that can be located anywhere from the pulmonary artery circulation to a shared sinus of Valsalva, occasionally there is an intrinsic abnormality of an otherwise normally placed vessel [*5*]. The following report describes a case of sudden death occurring in an eleven-year-old boy in whom marked hypoplasia of the right coronary artery was found at autopsy.

Case Report

An apparently healthy eleven-year-old boy suddenly lost consciousness while swimming in a public pool under the observation of a training instructor. When lifted to the side

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of the pool he was found to be cyanosed, pulseless, and not breathing. There was no evidence of convulsion, frothing at the mouth, or tongue biting. Resuscitation was begun immediately and continued in the ambulance, where spontaneous respiratory movements and irregular peripheral pulses were noted prior to final cardiorespiratory arrest. Death was certified soon after admission to the Adelaide Children's Hospital (Adelaide, Australia) following further failed resuscitation attempts.

The most significant past history concerned an admission to the hospital eight months earlier for investigation of sudden collapse. The episode occurred at school, where he had collapsed while sprinting. A teacher who witnessed the episode thought that he was asystolic and not breathing, and so resuscitation was begun immediately. He quickly recovered and was fully alert, with no evidence of postictal drowsiness or confusion. There had been no convulsion, frothing at the mouth, or tongue biting witnessed, although there had been urinary incontinence. Following this episode, he was admitted to the hospital for investigation. There was no history of exertional chest pain, although on specific questioning it was found that he had "fainted" once before at school, again while running. Physical examination results were unremarkable, except for previously documented congenital nerve deafness due to maternal rubella exposure, and laboratory test results, in particular electrocardiography and chest and skull X-rays, were all within normal limits. An electroencephalograph was normal, with no focal, asymmetric, or epileptogenic features. He had been quite well prior to the fatal episode, with no other features of congenital rubella infection evident.

Autopsy Findings

The body was that of a normally formed boy of eleven years of age. The upper airway showed hyperemia of the mucosa and a small amount of gastric contents within the major bronchi. The lungs weighed 150 g (right) and 138 g (left) (normal = 140 to 240 g/lung) and showed partial atelectasis with light red cut surfaces that failed to exude fluid. The heart weighed 130 g (normal = 100 to 160 g) and was anatomically normal except for the right coronary artery, which was markedly reduced in caliber in comparison with the left. The circulation was left dominant with the left circumflex artery running into the posterior descending coronary artery. Serial sectioning revealed widely patent left main, circumflex, anterior descending, and posterior descending coronary arteries. In contrast, the lumen of the proximal right coronary artery was markedly narrowed throughout its length, reducing to a pinpoint approximately 2.5 cm from the normally placed ostium. Microscopic examination confirmed the disparity in size of the two main coronary arteries, which were histologically otherwise unremarkable (Figs. 1 and 2). The myocardium was normal with no evidence of acute or chronic ischemic damage. There were no features of myocarditis or of any other occult disease process. Examination of the sinoatrial (SA) node revealed no ischemic changes and a normal-sized artery, which suggests that it arose from the left coronary system. No other significant pathological findings were present; in particular, there were no cerebral or cardiac manifestations of congenital rubella infection. Toxicological analyses of the serum, gastric contents, liver, and kidney were negative. Death was, therefore, thought to be due to a sudden cardiac episode and not to drowning. A possible likely terminal sequence was cardiac arrhythmia, secondary to coronary circulatory insufficiency (exacerbated by strenuous exercise) caused by the markedly hypoplastic right coronary artery.

Discussion

Congenital abnormalities of the coronary arteries are a well-recognized cause of sudden and unexpected death in children and young adults [*3,6–10*]. While angiographic studies in selected populations of symptomatic adult patients have shown an incidence of between

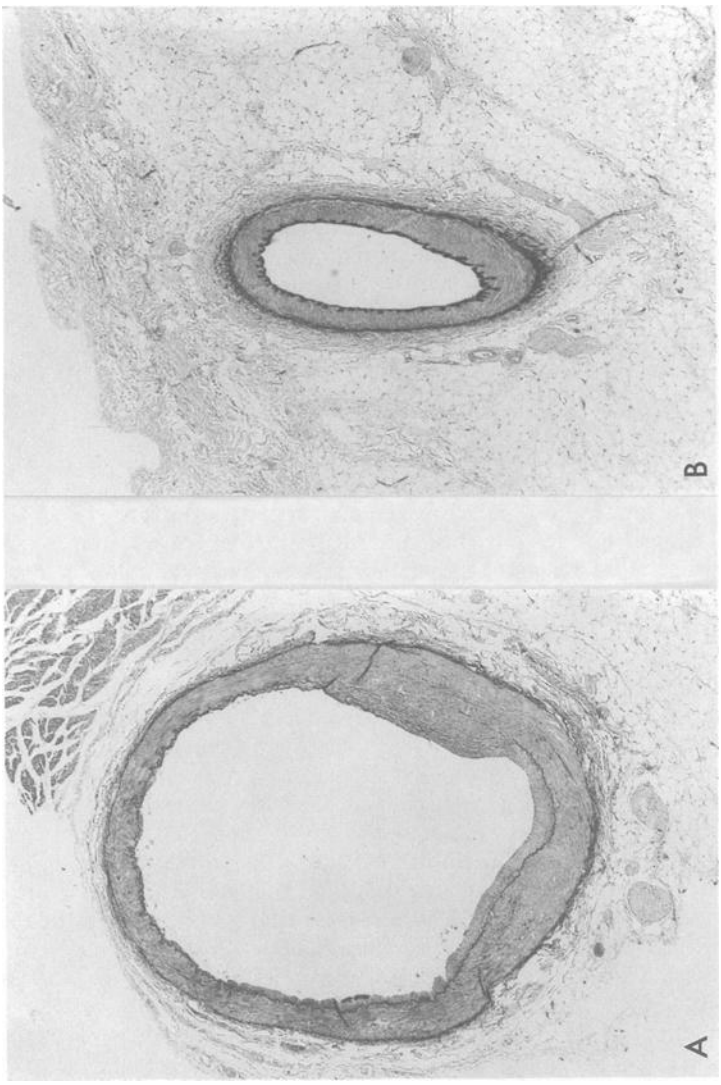

FIG. 1—*Marked reduction in cross-sectional area between* (a) *the proximal left coronary artery (section taken adjacent to a branch point) and* (b) *the proximal right coronary artery (magnification in both, ×15; Movat pentachrome stain).*

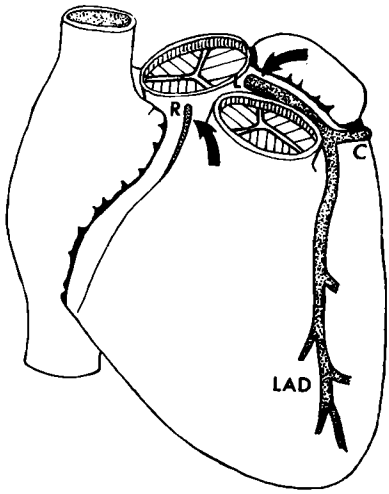

FIG. 2—*Diagrammatic representation of the heart, illustrating the marked disparity between the hypoplastic right coronary artery (R) and the left coronary (L), circumflex (C), and left anterior descending (LAD) arteries. Arrows show the position of the sections in Fig. 1.*

0.3 and 1.2% [*11,12*], the number associated with unexpected death in childhood appears to be less than this. The most commonly described anomalies that cause lethal problems are aberrant origin of the coronary arteries from variable sites, such as the pulmonary trunk, the aortic arch, or a shared sinus of Valsalva [*6–10*]. Death is believed to be due to compression or kinking of the abnormally placed artery as it courses around the roots of the major vessels. An acute angle of takeoff of the artery from its ostium and ridges in the intima are also features which are thought to interfere with the normal flow of blood [*3,6*]. Reduction in the blood supply to the dependent areas of the myocardium then results in ischemia, arrhythmia, and death. Often there is no evidence of any recent or preexisting myocardial damage, so the terminal event is presumed to be acute, possibly related to a sudden increase in physical activity [*6*]. Young athletes who have unexpectedly died and are found at autopsy to have aberrant coronary arteries demonstrate this point [*6–8*].

While aberrantly situated coronary arteries may be hypoplastic [*3*], cases of sudden death in childhood associated with a marked disparity in the sizes of the coronary arteries, which in all other respects are normal, are extremely rare [*13*]. Most documented cases have described isolated stenosis of an ostium, rather than diffuse narrowing of the entire vessel, as was present in this case [*10*]. In fact, it is sometimes the practice not to routinely examine normally situated coronary arteries in detail in children because of the relatively low yield of abnormal findings. The case reported, therefore, demonstrates the importance of detailed transverse sectioning of all of the major coronary trunks at autopsy. Had this not been done, death might have been attributed to the clinical diagnosis of drowning, rather than to the underlying congenital cardiovascular defect, as the coronary arteries, which were embedded in a moderate amount of epicardial fat, appeared externally unremarkable, following the normal distribution from their respective sinuses of Valsalva.

Familial congenital deafness has a well-documented association with sudden and unexpected death, as in the Jervell and Lange-Nielsen syndrome, in which there is a prolonged QT interval on electrocardiography [*14,15*]. Our patient's normal electrocardiograph and negative family history made this diagnosis unlikely, particularly as the deafness

was most probably due to the known *in utero* exposure to rubella virus. Diffuse coronary artery hypoplasia, unlike pulmonary artery stenosis, atrial and ventricular septal defects, valve stenosis, and tetralogy of Fallot, is not a recognized complication of congenital rubella infection [*16*].

The absence of definite histological evidence of myocardial ischemia in this case is not surprising given the rapid clinical history and does not preclude the diagnosis of sudden cardiac death [*6,9,10*]. The well-documented episode eight months before in which the victim was observed by a teacher to suddenly collapse during a fast sprint suggests an underlying cardiovascular problem. A similar exercise-associated collapse on at least one other occasion also supports an underlying cardiac abnormality. The absence of an abnormal electroencephalogram soon after the collapse is against a diagnosis of epilepsy. Although extensive clinical investigation was undertaken at the time of hospital admission, this did not include an exercise tolerance test or ambulatory electrocardiographic monitoring, two investigations which might have unmasked possible episodic cardiac arrhythmias or ischemia, or both, due to critical coronary artery blood flow problems. It is of note that the patient was engaging in quite strenuous exercise at the time of the second collapse, which may have predisposed him to focal myocardial hypoxia. Although a history of angina might have been expected given the proposed underlying mechanism of death [*10*], it is not essential, as a number of cases in the literature of sudden death due to aberrant coronary arteries with ostial narrowing have been completely asymptomatic until the lethal episode occurred [*7,10*].

One difficulty that arises is the lack of a normal control population with which to compare the diameters of the coronary vessels. Further study is, therefore, being performed to help define an age-related normal range and ratio of major coronary artery diameters in childhood.

In conclusion, this case demonstrates sudden death in an otherwise completely healthy boy, associated with marked hypoplasia of the right coronary artery. The absence of any other significant findings at autopsy, coupled with the previous episodes of exercise-related collapse, point strongly to a cardiac problem. The authors, therefore, stress the importance of meticulous examination of the coronary artery system in all children who present with sudden and unexpected death, even in circumstances in which the coronary arteries appear externally normal or in which the cause of death appears initially straightforward.

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