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Sudden death in a youth

A case of quadricuspid aortic valve with isolation of origin of left coronary artery

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SUMMARY A youth of 16 years of age died suddenly and quite unexpectedly while walking to school. The necropsy disclosed a quadricuspid aortic valve with complete isolation of the left coronary artery by an adherent aortic valve cusp. The left ventricular myocardium showed subendocardial contraction band necrosis suggesting that critical ischaemia had triggered a state of hypercontraction, in keeping with ventricular fibrillation as the immediate cause of death. The site of the anomaly in the aortic root showed dysplasia of the aortic wall and the affected valve cusp, histologically similar to the changes that characterise supravalvar aortic stenosis. The findings suggest a developmental anomaly, possibly a *forme fruste* of supravalvar aortic stenosis, rather than a postnatally acquired condition.

Sudden and unexpected death in adolescence is a rare event, but when it occurs the cause is often difficult to identify. In a number of cases a cardiovascular abnormality is found, such as hypertrophic cardiomyopathy, aortic coarctation, aortic stenosis, or congenital anomalies of the coronary arteries. It is a sad experience under such circumstances to realise that the underlying anomaly is often amenable to surgical intervention.

This report documents such a case in which the necropsy disclosed isolation of the ostium of the left coronary artery by adhesion of an aortic cusp, itself part of a quadricuspid valve.

Case report

The patient, a boy of 16 years, had been well until a few months before death, when he gradually noticed shortness of breath on exertion. He began to walk to school, instead of cycling, and it was noted by others that his enthusiasm for gymnastics had lessened. Otherwise, nothing unusual was noted. He never complained of chest pain and did not consult a doctor. While strolling to school he suddenly collapsed and died.

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NECROPSY

The relevant findings all relate to the heart, which weighed 267 g. There was a quadricuspid aortic valve with dysplastic features. Three of the cusps were of fairly uniform size. These were an anterior non-coronary cusp, a right coronary cusp, and a posterior non-coronary cusp (Fig. 1). The measurements of these three cusps were (width versus height) 14×12 mm, 14×11 mm, and 16×12 mm, respectively. The leaflets were thin and the commissures were normally formed. The fourth cusp, however, was thick though clearly separated from the adjoining cusps by well delineated commissures. The measurements of this additional cusp were 9×8 mm (width versus height). This additional cusp was totally adherent to the aortic wall, so that its true nature was only discovered when a section through the aortic root was made, parallel with the level of the valve (Fig. 1). The abnormal cusp produced a dome-like leaflet with a dimple on its luminal aspect (Fig. 1A). The "dome"

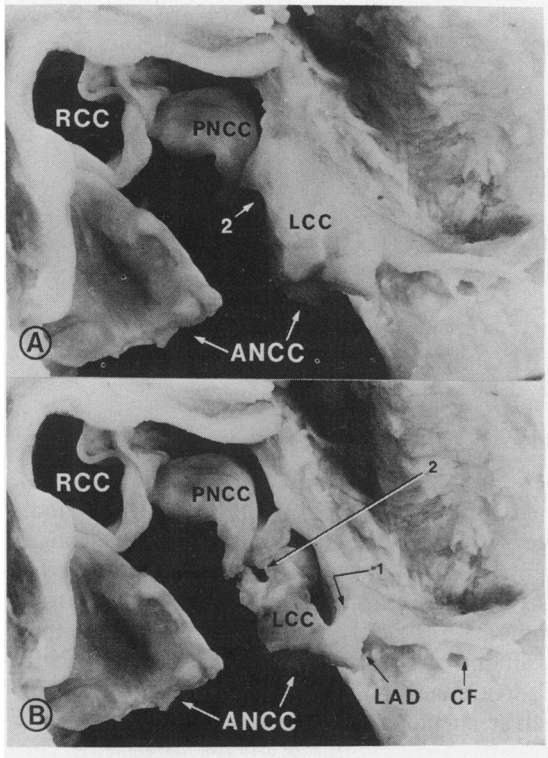

Fig. 1 *Superior view of the quadricuspid aortic valve, opened through the anterior non-coronary cusp (ANCC). (A) The left coronary cusp (LCC) presents as a dome-like structure with a dimple at the aortic site (arrow 2). (B) The same site after an incision parallel to the plane in (A). It now becomes evident that the left coronary cusp had completely isolated the ostium (arrow 1) of the left coronary artery. The latter immediately divides into small-sized and thin-walled anterior descending (LAD) and circumflex (CF) arteries. The sinus of Valsalva of the right coronary cusp (RCC) is widely opened. PNCC, posterior non-coronary cusp.*

completely isolated the ostium of the left coronary artery from the aortic lumen (Fig. 1B). The true ostium of the left coronary artery was of normal size, but the main stem and major branches appeared small. Immediately beyond the ostium, the bifurcation into anterior descending and circumflex coronary arteries was present. Each of the two vessels had an external diameter of approximately 1·5 mm, with a thin wall. Histological sections of these arteries showed no structural abnormalities (Fig. 2B and C). The right coronary artery was dominant and showed a normal topography. The artery was not obviously dilated and did not show a tortuous course. Its external diameter in the proximal segment measured 3 mm. Histology showed a normal architecture (Fig. 2A).

The heart showed no other congenital malformations and there were no gross signs of supravalvar aortic stenosis. Myocardial infarction was not identified on the gross specimen.

Microscopical examination disclosed signs of ischaemia in the subendocardial zone of both right and left ventricles (Fig. 3A). The immediate subendocardial layer of myocardial cells showed hydropic cell swelling with patches of contraction band necrosis, bordering upon a deeper zone of early coagulation necrosis. The latter zone was distinct only because of cytoplasmic eosinophilia and waviness in fibre architecture (Fig. 3A). There was no active inflammatory infiltrate and no signs of other disease processes, such as rheumatic fever.

Sections through the site of adhesion of the left coronary cusp to the aortic wall, which also encompassed the commissural site with the posterior non-coronary cusp, disclosed a distinct abnormality in aortic wall architecture (Fig. 3B). The normal parallel arrangement of the elastin fibres had been replaced by a "mosaic pattern" (Fig. 3C), extending beyond the region of the commissure into the valve itself. The

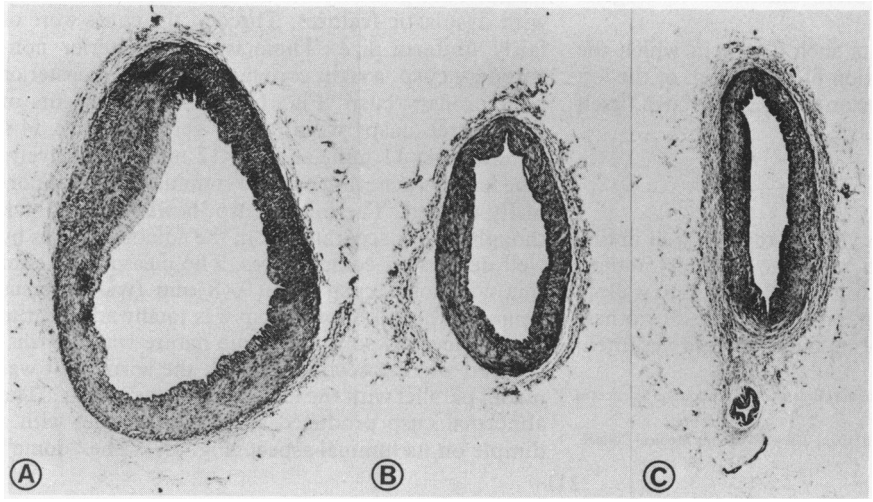

Fig. 2 *Histological cross-sections through the proximal right coronary artery (A), the left anterior descending coronary artery (B), and the left circumflex artery (C), shown at the same magnification. The wall structure of these arteries is basically normal. (Elastic tissue stain, counterstained with van Gieson's stain. ×18.)*

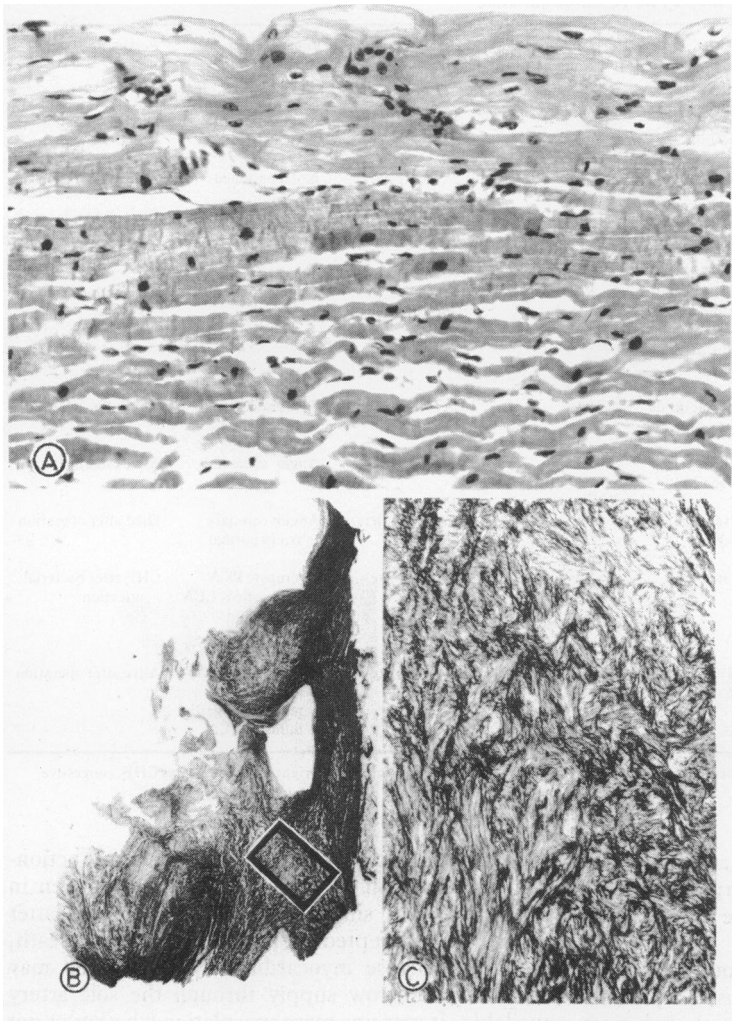

Fig. 3 *(A) Histological section of myocardium. The immediate subendocardial zone contains cells with mild hydropic cell swelling. Deeper, contraction bands are present, bordering upon an area with coagulation necrosis characterised by waviness of the fibres. (Haematoxylin and eosin stain. ×230.) (B) Histological section through the anterior wall and adhesion site of the left coronary cusp. The boxed area, enlarged in (C), shows a "mosaic" pattern, typical for supravalvar aortic stenosis.*

mosaic structure gradually blended with the more central area of the valve cusp which showed a haphazard intermingling of components which normally form well-delineated fibrosa and spongiosa layers. The whole area thus showed signs of dysplasia. There was no inflammatory infiltrate present.

Discussion

As far as we are aware, a case of quadricuspid aortic valve with adhesion of the left coronary cusp to the aortic bar, resulting in complete isolation of the orifice of the left coronary artery, has not previously been documented. Isolation of a coronary artery ostium by adherent aortic valve leaflets has always been reported with a tricuspid aortic valve and, with one exception, always in conjunction with supravalvar aortic stenosis (see Table). The one exception is a 16-year-old girl, described by Waxman and associates[9] in whom adhesion of the left aortic cusp occurred as an isolated lesion. This patient had experienced infrequent but typical angina pectoris and syncopal attacks since the age of 6 years. Selective coronary angiograms showed that the ostium of the left coronary artery was completely sealed off by an adherent left coronary cusp. The left coronary arterial system was filled through collaterals derived from the right coronary artery. At operation, the adherent cusp was incised and freed from its site of aortic fusion. Fifteen months after operation the patient was still free of symptoms.

Our patient was less fortunate in having no serious warning symptoms. Even in retrospect we have to conclude that his symptoms over the few months before death were far from alarming and did not

Table *Reported cases of coronary artery isolation by aortic valve adhesion*

Author	*Age (y)*	*Sex*	*Complaints*	*Associated malformations*	*Coronary artery involved*	*Intercoronary collaterals*	*Follow-up*
Denie and Verheugt[1]	25	M	Fatigue since age 19; angina for 9 mth	SAS	LCA (complete)	Not mentioned	Died at operation
Kreel *et al.*[2]	12	M	Chest pain for 9 mth	SAS	RCA (complete)	Not mentioned	Died at operation
Morrow *et al.*[3]	8	F	Failure to grow; respiratory infections, fatigue since age 5	SAS; PDA	LCA; RCA (incomplete)	Not mentioned	Died suddenly awaiting operation
Wooley *et al.*[4] Case 1	13	M	Progressive fatigue and exercise intolerance for years; recent onset of chest pain after exercise	SAS	RCA; LCA (incomplete)	Not mentioned	Died at operation
Bourassa and Campeau[5] Case 2	21	F	Dyspnoea and angina on exertion for 4 mth	SAS	LCA (incomplete)	Operation: RCA "quite large"	Alive after operation
Beuren *et al.*[6] Case 2	7½	M	Reduced exercise tolerance since age 2	SAS	LCA (complete)	Angio: collaterals	Alive after operation
Farrehi *et al.*[7] Case 1	14	M	Exertional dyspnoea and fatigue for years	SAS	RCA (incomplete)	Angio: coronary artery normal	Died after operation
Edwards[8]	23	F	(Not clearly stated)	SAS	RCA (complete)	Necropsy: RCA thin walled; LCA dilated and tortuous	CHF after bacterial infection
Waxman *et al.*[9]	16	F	Syncope and chest pain since age 6	—	LCA (complete)	Angio: large RCA, multiple collaterals, retrograde filling of LCA	Alive after operation

Abbreviations: SAS, supravalvar aortic stenosis; PDA, persistent ductus arteriosus; LCA, left coronary artery; RCA, right coronary artery; CHF, congestive heart failure.

indicate a pressing need to consult a doctor. His sudden death came as a complete surprise and the necropsy was the first indication that he was suffering from a serious congenital disorder.

A review of the published reports of other instances of cusp adhesion disclosed that almost all patients complained of dyspnoea and chest pain, though most of these symptoms may have been induced by the supravalvar aortic stenosis rather than by coronary ostial occlusion (Table). The patient reported by Waxman and associates,[9] who had no other congenital malformation had experienced attacks of angina pectoris and syncope for as long as 10 years before operation, but these symptoms may well have been aggravated by the fact that the aortic root anomaly had caused additional aortic stenosis and regurgitation. We found no evidence of these lesions in our patient. Isolation of a coronary artery orifice may cause the development of collaterals between the two coronary arteries (see Table). We had no opportunity to study this particular aspect adequately in our case but the heart showed a non-dilated and non-tortuous right coronary artery, suggesting absence of significant collateral flow. This is surprising in view of the uneventful clinical history and the fact that functionally this situation can be compared to that seen in patients with a single coronary artery. The latter condition is accepted as a cause of sudden death, probably because myocardial oxygen demand may ultimately outgrow supply through the sole artery available. It remains mere speculation whether or not such a mechanism operated in our patient but the presence of contraction band necrosis and early coagulation necrosis of the myocardium suggests that ventricular fibrillation induced by ischaemia was the immediate cause of death.[10] The myocardium did not contain appreciable fibrosis, suggesting that myocardial cell death before the fatal episode had been minimal or absent.

It might be argued, therefore, that isolation of the ostium of the coronary artery was recently acquired. There were no signs, however, of an inflammatory process or any other circumstance that could account for cusp adhesion, so that this possibility lacks further support. The alternative is that the abnormality was congenital. The mosaic tissue architecture at the site of the deformity and the disorderly arrangement of tissue components in the valve cusp indicate dys-

plasia, as in a developmental abnormality and therefore the lesion cannot be regarded as "acquired" in the sense of a postnatally acquired disease. Dysplasia of the media of arteries has previously been described as a form of generalised fetopathy by Becu and Gallo[11] and has also been documented as an aortic wall abnormality in patients with "isolated" pulmonary valve stenosis.[12] Moreover, the mosaic pattern has been described in the aortic wall at the site of atresia of the ostium of the left coronary artery, without aortic valve deformities.[13 14] In the latter instances the anomaly had been compared to supravalvar aortic stenosis, since the mosaic pattern of the aortic wall is generally accepted as the histological hallmark of that particular condition. It is of interest that Waxman and associates,[9] observing that all previous reported cases of coronary arterial ostial isolation were associated with "classical" supravalvar aortic stenosis, suggested that the anomaly in their case could represent a *forme fruste* of that condition. In our case we have histological evidence to support this concept.

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