

Coronary sinus orifice atresia and persistent left superior vena cava

A report of two cases, one associated with atypical coronary artery thrombosis

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SUMMARY Fourteen previously reported cases of atresia of the coronary sinus ostium, in which the coronary venous flow entered the right atrium by a persistent left superior vena cava, the innominate vein, and the right superior vena cava, are reviewed and two new cases reported. The first new case was in a 43 year old woman with atypical chest pains in whom investigations for suspected ischaemic heart disease, including coronary arteriography, yielded normal results. She died suddenly from massive myocardial infarction due to extensive old thrombotic occlusion of a major coronary artery without any appreciable underlying disease of the vessel wall. Exercise testing is considered to be advisable in symptomatic patients with normal coronary angiograms and attention to the venous phase might be informative. The second new case occurred in a child with an atrial septal defect and complete transposition of the great arteries, who died aged 1 month.

Atresia of the opening of the coronary sinus into the right atrium is a rare malformation and is often associated with anomalous coronary venous drainage into the left atrium through abnormal coronary veins or through an abnormal opening of the coronary sinus.¹⁻⁶ In a few instances, atresia of the coronary sinus orifice has been accompanied by drainage into the right atrium through a persistent left superior vena cava, the innominate vein, and the right superior vena cava. The first such case was reported in 1738 as quoted by Marshall.⁷ Since then 11 similar cases have been described^{1 8-15} together with one in a young cat¹⁶ and one in which the orifice showed severe stenosis rather than atresia.¹⁷ The Table gives details of these. We now report two further cases.

The diversion of the coronary blood flow is said not to be of clinical importance because it is physiologically corrected in that blood enters the venous atrium, albeit by a circuitous route. One of our cases, how-

ever, was associated with unexplained chest pains, presumably of cardiac origin, and unexpected death due to myocardial infarction after thrombotic obstruction of a major coronary artery in the absence of gross disease of the coronary artery wall.

Case reports

CASE 1

A 43 year old woman presented with a nine month history of precordial chest pain described as both stabbing and aching and sometimes associated with discomfort in the left elbow; each episode lasted for up to one hour. The pain often occurred at rest but was occasionally precipitated by exertion, when she obtained some relief 10 minutes after taking sublingual tablets of glyceryl trinitrate. Over the preceding few weeks she had been woken from sleep several times a night by similar pains and was consuming as many as 20 tablets of glyceryl trinitrate daily as well as frusemide, potassium supplements, and slow release aminophylline. She smoked 40 cigarettes a day, and her mother had died at the age of 43 of a "fatty heart." Both her sisters (aged 41 and 50) suffered from inter-

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Table Reported cases, including those in present paper, of coronary sinus atresia with drainage into a persistent left superior vena cava

Author	Year	Sex	Age	Other cardiac malformations	Notes
Le Cat	1738	F	8 d		
Gruber	1885	M	50 yr		
Siding	1886				
Beyerlein	1914	M	1 yr, 3 mnth		Died of diphtheria
Hutton	1915				
Harris <i>et al</i>	1927	M	1 yr, 10 mnth	Transposition of great arteries	
Reed	1938	M	77 yr		Drug addict; died of pulmonary oedema
Edwards	1960			Univentricular heart	
Edwards	1960			Atrioventricular defect	
Edwards	1960			Ebstein's malformation of tricuspid valve	
Fudemoto <i>et al</i>	1976	F	70 yr		Ischaemic heart disease
Grosse-Heitmeyer <i>et al</i>	1982	M	18 yr	Partial anomalous pulmonary venous connexion	
Grant	1960				Cat
Peele	1932	M	Adult		Negro; small coronary sinus orifice present
Gerlis <i>et al</i>	1984	{ F F	43 yr 1 mnth	Transposition of great arteries	Ischaemic heart disease

mittent chest pains (one was hypertensive) and had normal electrocardiograms.

On examination she appeared to be older than her years. Pulse rate was 80 beats/minute, blood pressure was 140/80 mm Hg, and there were no abnormal clinical signs. Chest radiographs showed normal lung fields with a cardiothoracic ratio of 12 to 30 cm, and electrocardiography showed sinus rhythm with planar inferior ST segments but otherwise yielded results within normal limits. She was admitted for coronary angiography, and treatment was initiated with a calcium channel blocker. Blood count, erythrocyte sedimentation rate, and concentrations of urea, electrolytes, and glucose were all within normal limits. Blood lipid concentrations were not measured. Left heart catheterisation showed an aortic pressure of 140/85 mm Hg and a left ventricular pressure of 145 mm Hg at systole and 8 mm Hg at end diastole. Left ventriculography showed a normally contracting ventricle. Selective coronary artery injections were performed and showed a dominant left coronary system. Stenoses were not visible in any part of the coronary arterial tree with the exception of a short muscle bridge across the left anterior descending artery distal to the origin of the principal diagonal branch. She was duly reassured that her coronary arteries were normal and was discharged from the clinic, her symptoms having slightly improved with antacids, but she remained convinced that her heart was abnormal. Four months later she was found dead in bed. After this her younger sister underwent cardiac catheterisation. There were normal arterial and venous phases of coronary angiography, and a persistent left superior vena cava was excluded by left subclavian venography.

Findings at necropsy

Postmortem examination did not show any significant abnormalities other than those of the cardiovascular system. There were extensive old pericardial adhesions anteriorly. The heart was moderately enlarged and weighed 475 g. Bilateral superior vena cavae were found. The right sided vein was 12 mm in diameter and opened normally into the right atrium; the left sided vein was 4 mm in diameter and drained into the right vein through the innominate vein. The lower part of the left sided vein passed medially and to the right, in the posterior atrioventricular groove, between the left atrium and the left ventricle. The lowest part was dilated up to 8 mm in diameter, received various coronary veins, and was in all respects morphologically identical to a normal coronary sinus except that it terminated blindly at the junction with the middle cardiac vein and had no opening into the right atrium or into any other cardiac chamber (Figs. 1-3).

The atria were of normal solitus morphology, and the venous connexions were normal apart from the coronary sinus anomaly. There were several openings of thebesian veins, up to 2 mm in diameter, in both atria. The atrioventricular valves, ventricles, and great arteries were of normal morphology. The ventricular myocardium showed extensive old fibrosed full thickness infarction of most of the septum with extension into the anterior wall at the apex and the posterior wall in the mid-zone. There was extensive endocardial fibrosis within the left ventricle, with adherent old thrombus at the apex where the wall consisted of scar tissue only 3 mm thick. The aortic and pulmonary valves were normal. The aorta was free from gross atheroma. The left coronary artery

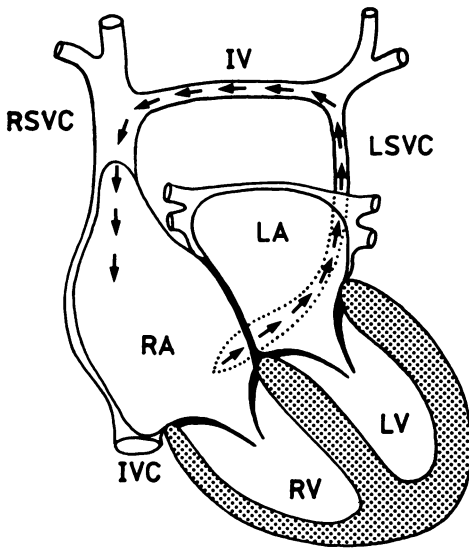


Fig. 1 Diagram of the cardiac chambers and systemic veins seen in cases 1 and 2. Coronary venous flow is indicated by arrows; coronary sinus is outlined by a dotted line. LSVC and RSVC, left and right superior vena cava; IVC, inferior vena cava; IV, innominate vein; LA, left atrium; RA, right atrium; LV, left ventricle; RV, right ventricle.

orifice was normal and fully patent (3 mm in diameter). The main left coronary artery appeared to be normal. The circumflex branch arose after 6 mm, had a normal distribution, and continued into the pos-

terior descending vessel. It was of normal width (roughly 2–2.5 mm in diameter) and macroscopically fully patent and free from atheroma.

The anterior descending coronary artery continued normally from the main left artery and then divided into two branches: a larger medial vessel and a slightly smaller vessel lying to the left. The larger medial branch passed obliquely downwards into the ventricular muscle of the anterior wall of the left ventricle and interventricular septum and showed extensive occlusion by greyish brown, dense, firmly adherent "thrombus"; this extended from just distal to the origin of the artery for a distance of 25 mm. There was no gross evidence of atheroma. The lateral anterior descending vessel was normal except for a small localised area of atheroma affecting two thirds of its circumference and extending for a length of about 3 mm with minimal luminal stenosis and with no evidence of mural thrombosis or haematoma. There were two small right coronary artery orifices 1 mm and 1.5 mm in diameter; these opened into small but quite patent and apparently normal arteries. The coronary veins were of normal size and appearance.

Histology

Numerous blocks were taken from the main coronary arteries at intervals of 3 mm. The left main coronary artery and the first part of the anterior descending artery were normal. The medial branch showed complete occlusion by organised old thrombosis with extensive revascularisation. There was slight concen-

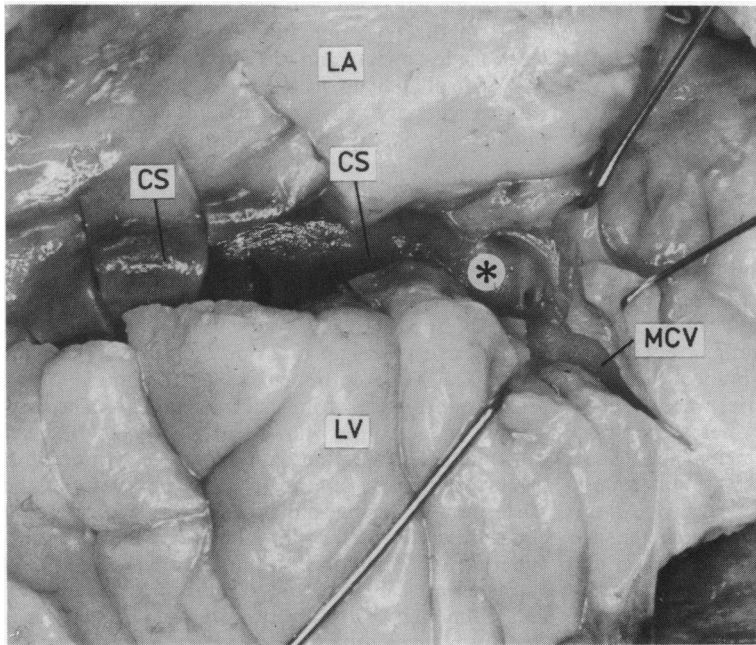


Fig. 2 Posterior aspect of heart (case 1) showing partially opened coronary sinus (CS). Asterisk indicates position of orifice atresia. LA, left atrium; LV, left ventricle; MCV, middle cardiac vein.

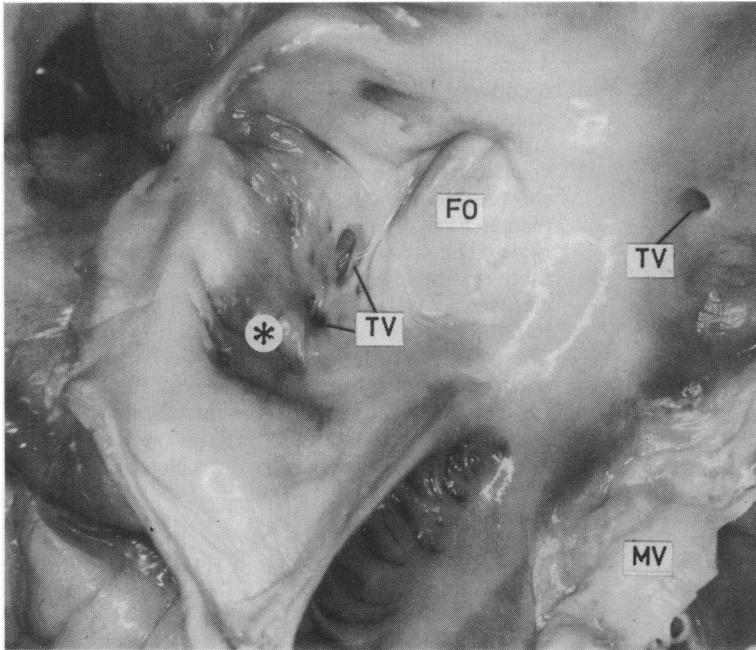


Fig. 3 Interior of right atrium (case 1). Asterisk indicates site of coronary sinus orifice atresia. FO, fossa ovale; TV, openings of thebesian veins; MV, mitral valve.

tric intimal hyperplasia, which merged with the occluding tissue, but no localised plaques, lipoid deposits, or calcification were found. The occluded portion dipped downwards obliquely to penetrate the muscle of the anterior wall, which bridged over it, but there was no suggestion that this had caused any stenosis or kinking (Fig. 4). The lateral descending branch showed only slight intimal thickening with the exception of the previously mentioned short segment, in which there was a localised zone of eccentric narrowing of the lumen to 1.5 mm in diameter, representing roughly 60% stenosis. This lesion consisted of intimal proliferation with slight lipoid deposition in the deeper aspect forming an early atheromatous plaque. No luminal thrombus was seen. The circumflex and posterior descending arteries showed only slight, mainly concentric, intimal hyperplasia and were fully patent. The right main coronary artery was rather small but was generally patent except for a localised area of eccentric intimal thickening that had narrowed the lumen to 0.8 mm in diameter (roughly 70% stenosis) (Fig. 5). There was no evidence of active or previous arteritis or of coronary artery embolism.

The coronary veins were histologically normal compared with those in a normal subject of the same age and sex. The myocardium showed extensive old full thickness infarction and fibrosis with antemortem endocardial thrombosis at the apex. The atrioventricular node was examined by serial sections; it was found to be small (3.5×3×1.0 mm) compared with

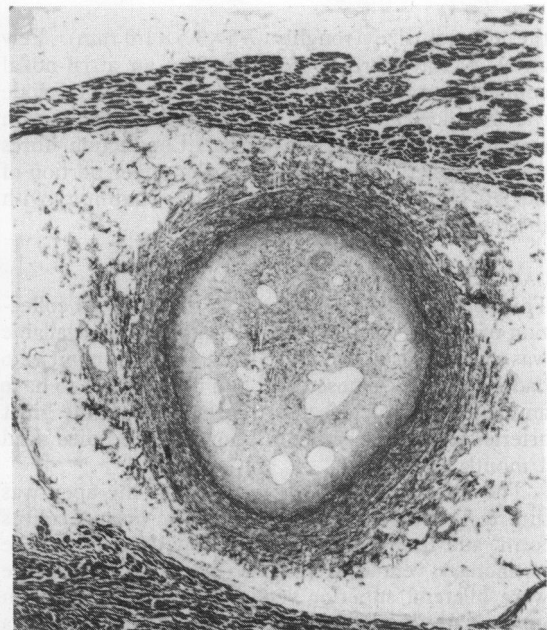


Fig. 4 Transverse section of affected portion of left descending coronary artery (case 1) showing overlying muscle bridge and organising thrombus occluding lumen. The arterial wall appears to be normal. (× 15. Haematoxylin and eosin stain.)

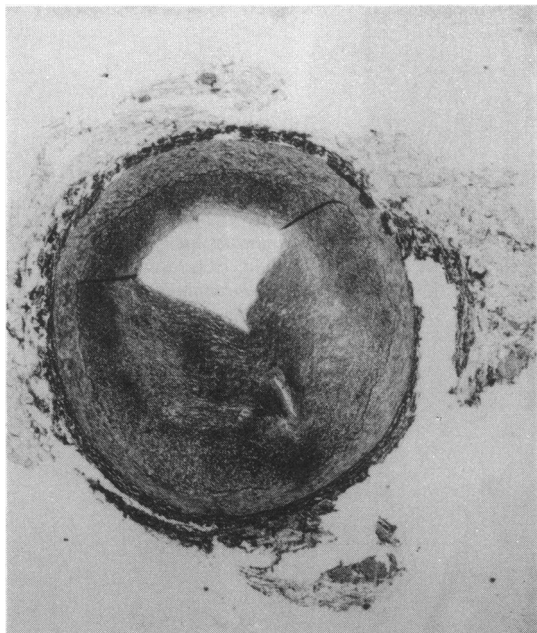


Fig. 5 Transverse section of a portion of right main coronary artery (case 1) showing considerable localised stenosis due to eccentric intimal thickening. ($\times 15$. Masson's trichrome stain.)

the normal size (roughly $7.5 \times 3.5 \times 1.0$ mm). Few atrial overlay fibres were seen, but an atrial-nodal connexion was present, although limited. The node was histologically normal. The penetrating portion and bundle of His were normal. Conduction fibres were considerably attenuated in the upper portion of the left bundle branch, but there was continuity with the main bundle.

CASE 2

This specimen was discovered in the personal collection of LMG. The only clinical information available was that the heart had come from a female infant who had been very cyanosed and had been shown to have an atrial septal defect and transposition of the great arteries by cardiac catheterisation. She had died aged 1 month.

The heart was of normal size, and the apex was directed to the left. The atria were of normal solitus form, and the right atrium was slightly dilated. The pulmonary venous connexions were normal. There were bilateral superior vena cavae connected by an innominate vein. The right sided superior vena cava was 5 mm in diameter, received a normal azygos vein, and opened normally into the right atrium. The left sided vein was narrow, 2 mm in diameter, passed downwards on the lateral wall of the left atrium, and

then curved backwards to enter the posterior atrioventricular groove, where it became dilated to 4 mm in diameter and received the coronary veins, occupying an identical position to a normal coronary sinus. No orifice, however, was present into the right atrium or any other cardiac chamber. The inferior vena cava was normal. Interiorly the right atrium showed a prominent right venous valve and a 5 mm in diameter valvar opening in the upper part of the fossa ovale. The atrioventricular valves were normal, and there was atrioventricular concordance. The ventricles were of normal morphology, with an intact septum and moderate right ventricular hypertrophy. There was ventriculoarterial discordance (d loop transposition of the great arteries) with the aorta arising from the right ventricle and the pulmonary artery arising from the left. The arteries were otherwise normal. The ductus arteriosus was closed.

Discussion

In all the 16 reported cases of coronary sinus ostial atresia with persistent left superior vena cava, including the present two, the ages ranged from 8 days to 77 years, and in half the 10 cases in which age was stated death occurred under the age of 2 years. Edwards considered that the condition was commonly associated with other cardiac malformations,¹ but these were present in only six (37.5%) of the total series (Table).

From descriptions of the published cases this would appear to be a benign condition as the coronary flow reaches the venous atrium, albeit by a circuitous route. Edwards commented that in such cases the left superior vena cava may be so narrow that it constitutes an ineffective channel,¹ and Falcone and Roberts pointed out that the haemodynamic consequences of coronary venous hypertension would be reflected largely in the left coronary artery as 75% of its flow enters the coronary sinus.² Apart from the case of our patient (case 1), nothing has really suggested that the coronary sinus orifice atresia interfered with myocardial perfusion. Ischaemic heart disease was mentioned in only one case,¹⁴ a 70 year old woman who had a left ventricular aneurysm and narrowing of the right coronary artery as shown by angiography. In both our cases the persistent left superior vena cava was small, and, although there was no evidence that the coronary venous flow was restricted on morphological examination, it remains possible that during life the extended venous pathway could have been submitted to obstructive pressure from various mediastinal structures.

Review of the angiograms of our patient (case 1) showed that the coronary venous system was not well seen, perhaps an unusual finding with a normal coro-

nary blood supply. The films were of excellent quality, and, even with hindsight, no coronary stenosis was visible. A mild degree of coronary atheromatous disease was undoubtedly present at death and was probably present but undetected at coronary angiography; that coronary arterial narrowing assessed by coronary angiography may be less than when assessed in fixed pathological specimens has been well described.^{18,19} Unfortunately, the patient did not undergo exercise testing or thallium scanning which might have provided important information before the case came to postmortem examination (although the clinical course would probably not have been changed).

The influence of the atretic coronary sinus on the development of the occlusion of the coronary artery remains a matter for conjecture; the coronary arterial stenoses found at necropsy were not of the degree usually associated with precipitation of acute myocardial infarction, and, in particular, there was no evidence of atheromatous plaque in the vessel that was thrombosed. The muscle bridge across the anterior descending artery seems an unlikely precipitant of thrombosis as the vessel was narrowed only during systole, as is usual with this phenomenon. Coronary arterial spasm has been accepted as a cause of myocardial infarction and of angina²⁰ but as yet has not been put forward as a cause of coronary thrombosis, clearly the ultimate cause of death in this patient (case 1). She had no evident predisposition to thromboembolic disease and was not taking the contraceptive pill.

It has been suggested that patients with chest pain and normal coronary arteriograms enjoy a benign prognosis,²¹ yet in some series as many as 7% of patients with myocardial infarction are later found to have normal or near normal coronary arteries^{22,23} and increasing evidence is being published that casts doubt on the prognostic value of a normal coronary angiogram. In most cases these patients may safely be reassured that their hearts appear to be normal and that the outlook is good. It may, however, be both interesting and prudent to subject patients with pain and normal angiograms to exercise testing either before or after invasive investigation, and more conscious attention to the venous phase of coronary angiograms may perhaps show that abnormalities of venous drainage are not as rare as they have so far appeared to be.

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