Isolated congenital absence of coronary sinus

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SUMMARY A case is reported in which contrast angiography showed complete absence of the coronary sinus, the cardiac veins draining separately into left and right atria. Normal systemic venous drainage was demonstrated by contrast cross-sectional echocardiographic methods and later confirmed by conventional angiographic injections. In addition, angiography showed a stenosis in the anomalous cardiac vein draining into left atrium accounting for the presence of a continuous murmur for which the patient was initially referred.

Absence of the coronary sinus is a rare congenital anomaly which has previously been described in association with other cardiovascular abnormalities. In particular, Raghib *et al.* (1965) described a developmental complex consisting of persistent left sided superior vena cava draining into left atrium, an atrial septal defect, and absence of the coronary sinus. Congenital absence of this structure has also been described in association with atrioventricular canal defects and other complex forms of congenital heart disease (Mantini *et al.*, 1966). To our knowledge no case of complete absence of the coronary sinus occurring as an isolated anomaly has been previously reported.

Case report

A 59-year-old woman was referred for evaluation of a cardiac murmur, her only symptom being fatigue. Twelve months previously mild hypertension had been noted and cyclopenthiazide was administered, with satisfactory blood pressure control. There was no past history of rheumatic fever, respiratory disease, or chest trauma. However, a cardiac murmur had been noted during her first pregnancy at the age of 23.

On examination the pulse rate was 80/minute, regular with a normal waveform. Blood pressure was 140/80 mmHg in both arms, supine and standing. The jugular venous pulse was normal. There was no abnormal praecordial impulses or thrills. Auscultation revealed normal heart sounds and at the second left intercostal space a continuous soft high-pitched murmur with late systolic accentuation. The murmur was unaffected by posture, respiration, or occlusion of the neck veins. The remainder of the physical examination was normal.

The resting electrocardiogram showed symmetrical T wave inversion in lead aVL with an upright P wave. The plain chest x-ray film was normal. The phonocardiogram using a high frequency filter with the transducer in the second left intercostal space documented the continuous murmur which was unaffected by respiration. Single crystal M-mode echocardiogram was normal. A 12 lead maximal treadmill exercise test was performed using the Bruce *et al.* (1973) procedure, during which an ear lobe oximeter¹ measured peripheral arteriolar oxygen saturation continuously. The patient achieved stage two with a heart rate of 140/minute, the test being limited by tiredness. A distinct increase in the intensity of the murmur was observed immediately after exercise. Oximetry revealed a gradual fall in oxygen saturation from a resting desaturated level of 93 to 86 per cent two minutes after exercise (Table). The phonocardiogram was repeated with ear lobe oximetry measurements after sublingual administration of 1.0 mg glyceryl trinitrate. The intensity of the murmur gradually increased to a peak at three minutes (in agreement with the auscultatory findings), there being no change in peripheral oxygen saturation.

In order to exclude arteriovenous fistula cardiac catheterisation with coronary arteriography was performed (Sones technique). No left-to-right shunt was detected on oxygen saturations and the atrial septum appeared intact. There was mild arterial desaturation at rest of 93 per cent, in agreement with the ear lobe oximetry measurements. Left ventriculography and coronary arteriography were normal. After the left coronary artery injection,

¹ Hewlett-Packard 47201A.

Table Oximetry measurements during treadmill test

Stage (Bruce)	Heart rate	12 lead electrocardiogram	Blood pressure (mmHg)	Arteriolar SO ₂
Rest	88	N	130/80	93%
Stage 1	134	N	160/85	—
Stage 2	142	N	170/90	—
Immediate PE	138	N	165/85	88%
2 minute PE	118	N	140/80	86%
4 minute PE	106	N	135/80	91%
6 minute PE	104	N	130/70	94%

Decrease in arteriolar ear-lobe oxygen saturation caused by increased right-to-left shunting (coronary venous efflux to left atrium) after exercise.

PE, postexercise; N, normal.

however, follow-through to the venous phase failed to opacify the coronary sinus, the left venous system

draining into the great cardiac vein which had a fixed stenosis near its origin (Fig. 1a). This vein then drained into a normal sized left atrium. Right coronary arteriography in the venous phase opacified the posterior interventricular vein draining into a normal right atrium (Fig. 1b). Between these veins draining separately into left and right atrium no coronary sinus was seen.

Cross-sectional echocardiography was performed using an 81° mechanical sector scanner¹ and three bolus injections of indocyanine green (Sahn *et al.*, 1974) given separately via left and right antecubital veins and right femoral vein. Systemic venous return was shown to be normal by the rapid appearance of 'contrast echoes' within the right

¹ Smith Kline Instrument Corporation.

Fig. 1 (a) Venous phase of left coronary artery injection demonstrating stenosis at origin of great cardiac vein (arrow), filling of left atrium (LA), and catheter tip in left coronary artery (LCA). The coronary sinus is absent in the posterior left atrioventricular groove. (b) Right coronary artery (RCA) injection with venous efflux into the right atrium (RA).

atrium only (Foale *et al.*, 1979). This was subsequently confirmed by conventional injections of radio-opaque contrast medium.

The patient's symptoms remain stable and have required no treatment.

Discussion

Although coronary venous drainage is usually fairly constant, several anomalies have been described (Mantini *et al.*, 1966; Helseth *et al.*, 1974). Absence of the coronary sinus is rare and is usually associated with other abnormalities, the commonest being an atrial septal defect with persistent left sided superior vena cava terminating in left atrium.

The coronary sinus is the major channel of cardiac venous return and 96 per cent of all veins which drain left ventricular myocardium (including the interventricular septum) efflux into this structure (Hood, 1968). In the posterior atrioventricular groove the coronary sinus receives the great cardiac vein which drains the anterolateral aspect of the heart via anterior interventricular and left marginal (diagonal) veins. The coronary sinus is formed laterally in the atrioventricular groove by the junction of the great cardiac vein with the oblique vein of the left atrium (vein of Marshall), the embryonic remnant of the left superior cardinal vein. This junction is often marked by an indentation produced by the venous valve of Vieussens. The coronary sinus transports 85 per cent of total coronary blood flow (Gensini *et al.*, 1965) and normally terminates on the inferomedial wall of the right atrium.

Anomalies of the coronary sinus are usually described as benign conditions or as part of more complex congenital cardiac malformations. Enlargement with persistence of a left sided superior vena cava is the most frequent anomaly, occurring in between 0.5 and 4 per cent of patients with congenital heart disease (Campbell and Deucher, 1954). Rarely, enlargement of the coronary sinus may be the result of communication between left atrium and coronary sinus ('unroofed coronary sinus'), with a resulting low pressure left-to-right shunt, or between coronary artery and coronary sinus (coronary artery fistula) resulting in a high pressure shunt. Very rarely, the ostium of the coronary sinus may be atretic, with the sinus ending in a blind sac. Drainage back to the right atrium is usually via an associated left sided superior vena cava, left innominate vein, and right superior vena cava. Complete absence of the coronary sinus has not hitherto been described as an isolated anomaly. When previously reported it has been in association with other anomalies, most commonly persistent left sided superior vena cava

terminating in left atrium with an atrial septal defect (located posteroinferiorly to the fossa ovalis). In these cases the cardiac veins drain separately into their corresponding atria (Raghib *et al.*, 1965).

In the case reported, no coronary sinus was seen during the venous phase of either coronary artery injection, left and right venous channels draining into their respective atria. The stenosis present in the venous channel draining into left atrium was best seen in the left anterior oblique projection, and corresponded to the site of the valve of Vieussens, where the oblique vein of Marshall joins the great cardiac vein to form the coronary sinus. We propose that this venous stenosis (in a major vein draining left ventricular efflux) was the cause of the continuous murmur in the patient. The distinct increase in intensity of the murmur after exercise and glyceryl trinitrate was, we believe, caused by increased venous return across the stenosis consequent upon an increase in myocardial blood flow. The further decrease in systemic oxygen saturation on exercise reflects both increased myocardial oxygen extraction and increased myocardial blood flow with an increase in right-to-left shunting. After administration of glyceryl trinitrate, however, systemic arterial desaturation remained the same, probably because increased myocardial blood flow was accompanied by a concomitant decrease in myocardial oxygen extraction.

This case also demonstrates the application of contrast cross-sectional echocardiography in patients suspected of having anomalous systemic venous return. It is unique in that congenital absence of the coronary sinus is described as the sole cardiac abnormality.

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