

Congenital diverticulum of the heart arising from the coronary sinus

ELIO DI SEGNI,* ANNETTE SIEGAL,† MICHAEL KATZENSTEIN‡

*From the Department of *Cardiology and of †Pathology, and ‡The Neonatal Unit, Meir General Hospital, Sapir Medical Center, Kfar-Saba, and Sackler School of Medicine, Tel-Aviv University, Israel*

SUMMARY A diverticulum originating from the coronary sinus was diagnosed by echocardiography in a two day old baby with hypoplastic left heart. At necropsy the diverticulum was seen to arise from the coronary sinus and it penetrated the right ventricular posterior wall.

Congenital diverticula of the heart have been reported to arise from the atria¹ or from the ventricles.^{2,3} We report on a diverticulum arising from the coronary sinus at its junction with the right atrium in a neonate with a hypoplastic left heart.

Case report

A 3.55 kg girl was born by normal delivery at the 39th week of a normal gestation. When she was two days old she was referred for cardiological evaluation because of mild cyanosis. Arterial PO_2 was 42 mm Hg. Physical examination showed a grade 2/6 high pitched pansystolic murmur and a single second heart sound. The peripheral pulses were normal at the first examination, but became weak on the third day of life. An electrocardiogram showed right ventricular hypertrophy and a chest x ray showed a bulge at the left cardiac border.

Cross sectional echocardiography was performed when she was two days old. The left atrium, the left ventricle, the aortic root, the aortic valve, and the ascending aorta were all hypoplastic. The mitral valve was atretic and a small interventricular septal defect was also seen. In the parasternal long axis view a large cavity was seen behind the left ventricle; above this was a large coronary sinus (fig 1a). The external wall of the cavity was approximately the same thickness as the atrial wall and did not contract appreciably; its floor was formed by ventricular wall. In the subcostal view we identified a hypoplastic left

atrium receiving the pulmonary veins. The right atrium was enlarged, and a very large coronary sinus was demonstrated. The ostium of the coronary sinus was widely patent. A standard four chamber posterior view showed that the accessory cavity was caudal to the coronary sinus and the right atrium (fig 1b). This diverticulum was divided into two by a membrane; the left part of the diverticulum was about twice the size of the right one and was connected to the coronary sinus by an opening of about 6 mm in diameter located on the floor of the coronary sinus at its junction with the right atrium. Contrast echocardiography was undertaken after injection of 2 to 3 ml of 5% dextrose into a left antecubital vein. Because the contrast echoes entered the heart from the coronary sinus a persistent left superior vena cava connected to the coronary sinus was diagnosed. The contrast material then entered the right atrium and the left portion of the diverticulum and thereafter the right part of the diverticulum.

Cardiac failure with cardiogenic shock progressively developed and the infant died when she was four days old.

PATHOLOGY

Macroscopic examination of the heart showed a bulge 2.5 × 2 cm on the posterior aspect of the ventricles (fig 2a). The right atrium, the right ventricle, and the pulmonary artery were enlarged; the ductus arteriosus was widely patent; the left atrium was severely hypoplastic, but normally connected to four pulmonary veins; the foramen ovale was patent. The mitral valve was atretic and the hypoplastic left ventricle communicated through a small ventricular septal defect with the right ventricle. The ascending aorta up to the isthmus was also hypoplastic. A

Requests for reprints to Dr Elio Di Segni, Department of Cardiology, Meir General Hospital, Kfar-Saba 44281, Israel.

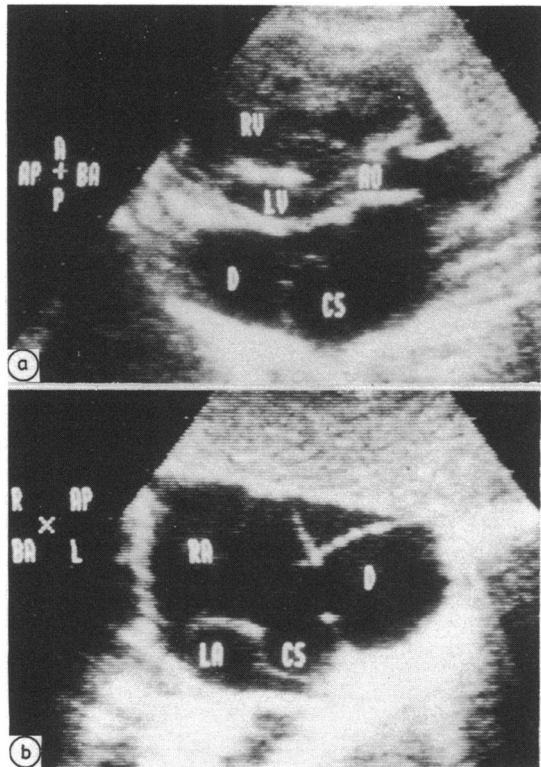


Fig 1 Cross sectional echocardiograms. (a) In the parasternal long axis view a diverticulum is seen behind the hypoplastic left ventricle and below the enlarged coronary sinus. (b) In the transversal subcostal view with posterior orientation of the transducer the large diverticulum is seen to be divided into two portions by a membrane. The left atrium is hypoplastic and the coronary sinus is enlarged. There is a connection between the diverticulum and the coronary sinus. AO, aorta; CS, coronary sinus; D, diverticulum; LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.

persistent left superior vena cava entered a very large coronary sinus. The ostium of the coronary sinus was patent and the thebesian valve was absent. An opening of about 6 mm in diameter on the floor of the coronary sinus just before its junction with the right atrium led into the left chamber of a diverticulum (fig 2b). The diverticulum lay behind the right ventricle and behind and to the right of the hypoplastic left ventricle; its superior border reached the atrioventricular groove below the coronary sinus (fig 2a) and its inferior border was 8 mm from the cardiac apex. The external surface of the diverticulum was covered by epicardium and by epicardial vessels continuous with the epicardial vessels of the ventricles. The diverticulum was divided into two cavities by a fenestrated membrane (fig 2c); the

left cavity was about twice as big as the right, being connected to the coronary sinus through a hole in the upper portion of its floor; the only openings of the right cavity were the fenestrations of the diverticular septum. The floor of the diverticulum was formed by trabeculated muscle covered by a smooth endocardium; the free wall was 0.5 mm thick.

Microscopic examination showed that the floor of the diverticulum was formed by normal myocardium covered by endocardium. Its free wall was composed of a layer of thin myocardial fibres, normal endocardial lining, and normally vascularised and innervated epicardium on the external surface (fig 2d). The septum of the diverticulum was formed by thin myocardium lined on both sides by endocardium.

Sections of the wall of the coronary sinus itself showed a well developed and oriented myocardial layer with a thin myocardium and a well vascularised epicardium with very scanty fatty tissue.

Discussion

Congenital diverticula of the heart are rare anomalies that usually arise from the left ventricle. They have been found on the basal aspect of the left ventricle⁴ or on the apex.^{2,5} Diverticula on the apex usually occur as part of a syndrome of cardiac anomalies and midline defects. The walls of these congenital left ventricular diverticula are formed by normal ventricular myocardium. Subvalvar left ventricular aneurysms, principally occurring in the Bantu and characterised by extensive fibrosis of the walls, with a suggested congenital^{6,7} or acquired aetiology have also been described.⁷

Right ventricular diverticula are extremely rare.³ They are occasionally associated with a left ventricular diverticulum.³ Only three cases of a congenital diverticulum arising from the right ventricle alone have been reported.^{3,8} These diverticula appeared as large outpouchings of the right ventricular cavity at the apex⁸ or at the base of the ventricle.³

Congenital aneurysms of the atria have also been reported.^{1,9,10} These are usually found in the appendage of the left⁹ or of the right atrium.¹⁰

Congenital anomalies of the coronary sinus include enlarged coronary sinus as a result of anomalous connection to the coronary sinus of systemic or pulmonary veins and fenestration or absence of the coronary sinus.¹¹⁻¹³ Fenestration of the coronary sinus ("partially unroofed coronary sinus") creates a communication between the atria and provides a route for blood to flow from one atrium to the other when an atretic atrioventricular valve is present.¹²

The diverticulum in our patient differs from the

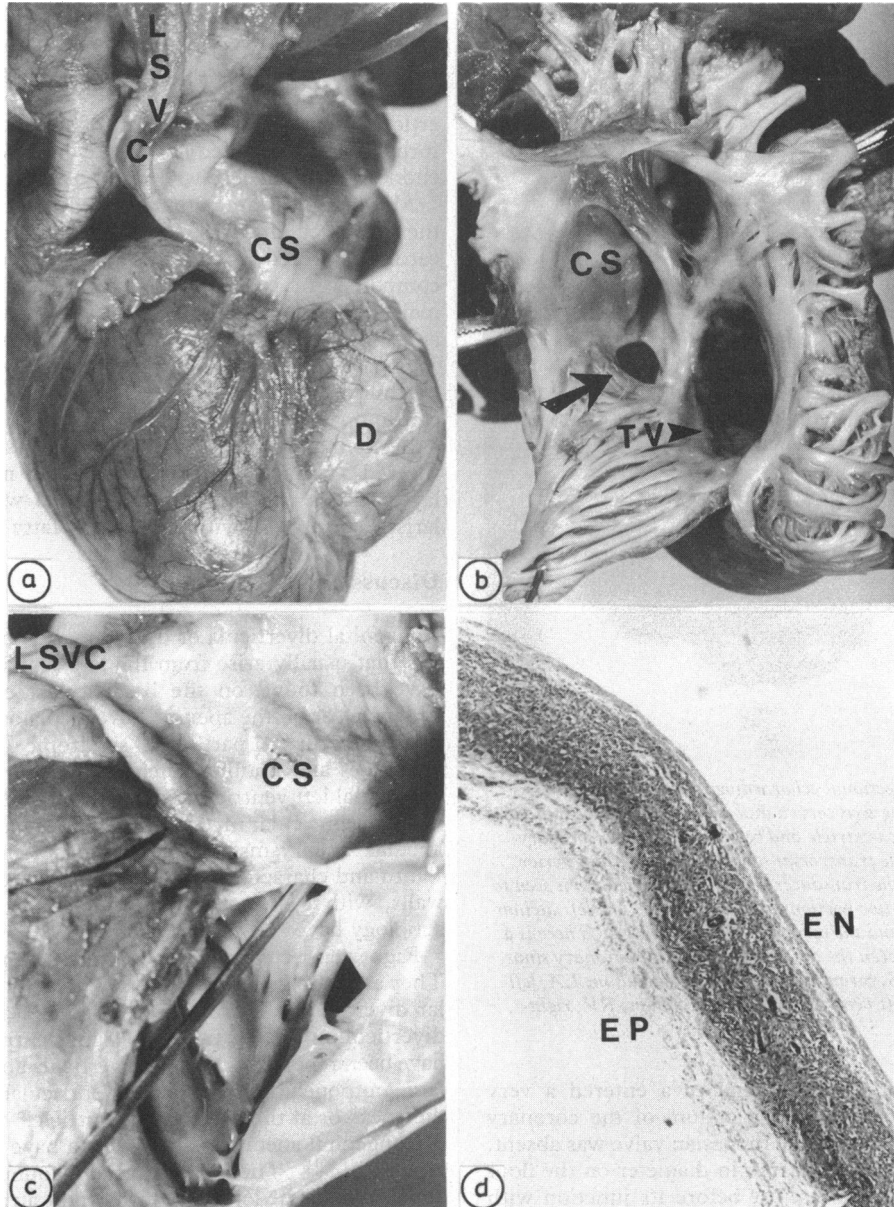


Fig 2 Pathology of the heart. (a) External view of the heart from its left side showing a 2.5 cm long diverticulum located below the atrioventricular groove and behind the ventricles. The large structure above the diverticulum is the coronary sinus, which is connected to the left superior vena cava. (b) Internal view of the right atrium. The ostium (arrow) of the diverticulum (6 mm in diameter) is within the coronary sinus immediately before its connection to the right atrium. (c) Internal view of the right and larger portion of the diverticulum. The floor of the diverticulum is formed by trabeculated right ventricular muscle lined by endocardium. The septum of the diverticulum is fenestrated. The free wall of the diverticulum is 0.5 mm thick. A probe is inserted in the opening between the diverticulum and the coronary sinus. (d) Microscopic appearance of the free wall of the diverticulum. The myocardial layer is covered by epicardium and internally by endocardium. Masson stain. EN, endocardium; EP, epicardium; LSVC, left superior vena cava; TV, tricuspid valve; other abbreviations as in fig 1.

anomalies described above and probably resulted from a developmental defect. It can be distinguished from a false aneurysm of inflammatory or traumatic origin because there were well developed striated myocardial fibres and no fibrous tissue in all portions of its walls. It may be a true diverticulum of the coronary sinus: the presence of myocardial fibres in its walls does not rule out this diagnosis because the coronary sinus is part of the cardiac venous system and, as with other juxtacardiac segments of veins (and as found in our case), myocardial fibres may be present in its walls.¹⁴ If this anomaly does indeed represent a true diverticulum of the coronary sinus one would expect to see similar histological components through the entire circumference of its walls. In our case, however, the floor of the diverticulum clearly belonged to the right ventricular wall. The free wall of the diverticulum was formed by thinned myocardium normally overlaid by pericardium and internally lined by endocardium. Its floor was formed by the normal right ventricular myocardium lined by endocardium; we believe the diverticulum was the result of a dissection within the right ventricular external wall.

A dissection of the right ventricular free wall could be the result of a primary event leading to the formation of a cavity that tracks to the coronary sinus and penetrates into the heart cavities. Cases of subvalvar aneurysm originating within the left ventricular wall and extending into the left atrium have been reported.⁷ In these cases extensive fibrosis, calcification, and even ossification of the left ventricular wall were found. In our case, however, there was no evidence of degenerative or inflammatory processes affecting the myocardium and leading to the formation of a cavity. There are no reported examples of a primary congenital dissection of the myocardium. Thus it seems most probable that this diverticulum originated from the coronary sinus and then penetrated into the right ventricular wall. Defects in the walls of the coronary sinus, although rare, have been reported,^{11 13} and we assume that a deficiency in the floor of the coronary sinus during fetal life by an undetermined cause resulted in the formation of a sac which made its way through the right ventricular myocardium.

In our case the coronary sinus was enlarged and was connected to a persistent left superior vena cava; mitral atresia with a hypoplastic left ventricle and aorta were also present. Previously reported fenestrations of the coronary sinus associated with mitral atresia occurred on the floor of the coronary sinus and allowed an accessory route for blood to flow from the left atrium.^{11 12} In our case the opening was on the free wall of the coronary sinus, which is at variance with other reported defects; it did not con-

nect the left atrium to the right atrium and so did not play any functional role in relieving the left atrio-ventricular obstruction.

The diverticulum in our patient was clearly diagnosed by cross sectional echocardiography. The echocardiographic appearance of the diverticulum exactly matched the pathological features and allowed the in vivo diagnosis of this bizarre anomaly. Clinical recognition of this anomaly did not influence the management of our case because of the severe associated anomaly of hypoplastic left heart, for which only experimental treatment is available in few selected centres throughout the world. It is, however, possible that an isolated diverticulum could be surgically repaired. Successful resection of cardiac diverticula has been reported.^{2 9} Untreated diverticulae may rupture, be a source of embolisation, or produce cardiac failure and arrhythmias.

We have described a previously unreported congenital cardiac anomaly—a diverticulum originating from the coronary sinus. The excellent correlation between the echocardiographic and pathological findings means that this anomaly may be readily diagnosed once the possibility of its existence is known.

We thank Dr Jesse E Edwards for his advice in interpreting the pathological findings.

References

- 1 Bramlet DA, Edwards JE. Congenital aneurysm of left atrial appendage. *Br Heart J* 1981;45:97-100.
- 2 Mady C. Left ventricular diverticulum. Analysis of two operated cases and review of the literature. *Angiology* 1982;33:280-6.
- 3 Carter JB, Van Tassel RA, Moller JH, Amplatz K, Edwards JE. Congenital diverticulum of the right ventricle. Association with pulmonary stenosis and ventricular septal defect. *Am J Cardiol* 1971;28:478-82.
- 4 Franco-Vazquez S, Sutherland D, Fowler M, Edwards JE. Congenital aneurysm of left ventricular base. *Chest* 1970;57:411-5.
- 5 Cantrell JR, Haller JA, Ravitch MM. A syndrome of congenital defects involving the abdominal wall, sternum, diaphragm, pericardium and heart. *Surg Gynecol Obstet* 1958;107:602-14.
- 6 Chesler E, Joffe N, Schamroth L, Meyers A. Annular subvalvular left ventricular aneurysm in the South African Bantu. *Circulation* 1965;32:43-51.
- 7 Edington GM, Olufemi Williams A. Left atrial aneurysm associated with annular subvalvular left ventricular aneurysm. *J Pathol Bacteriol* 1968;96:273-83.

- 8 Cumming GR. Congenital diverticulum of the right ventricle. *Am J Cardiol* 1969;23:294-7.
- 9 Tanabe T, Ishikaza M, Ohta S, Sugie S. Interpericardial aneurysm of the left atrial appendage. *Thorax* 1980;35:151-3.
- 10 Sheldon WC, Johnson CD, Favaloro RG. Idiopathic enlargement of the right atrium. Report of four cases. *Am J Cardiol* 1969;23:278-84.
- 11 Mantini E, Grondin CM, Lillehei CW, Edwards JE. Congenital anomalies involving the coronary sinus. *Circulation* 1966;33:317-27.
- 12 Freedom RM, Culham JAG, Rowe RD. Left atrial to coronary sinus fenestration (partially unroofed coronary sinus). Morphological and angiographic observations. *Br Heart J* 1981;46:63-8.
- 13 Raghieb G, Ruttenberg HD, Anderson RC, Amplatz K, Adams P Jr, Edwards JE. Termination of left superior vena cava in left atrium, atrial septal defect and absence of coronary sinus. A developmental complex. *Circulation* 1965;31:906-18.
- 14 Czyba JC, Girod C. *Histologie*. 3rd ed. Villeurbanne: Simep, 1979:41.