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Unroofed Coronary Sinus and Coronary Sinus Orifice Atresia

Implications for Management of Complex Congenital Heart Disease

IAN ADATIA, MRCP, ADRIANA C. GITTENBERGER-DE GROOT, PHD*

Freiburg, Germany and Leiden, The Netherlands

Objectives. **The aim of this study was to assess the morphology of the coronary sinus, its drainage and associated cardiac malformations when there is either complete unroofing of the coronary sinus or atresia of its connection to the right atrium.**

Background. **As more children with complex cardiac anomalies are accepted for primary surgical repair or palliation with cavopulmonary anastomoses, a knowledge of coronary sinus and systemic venous anomalies is important if coronary venous return is to be preserved and residual shunts avoided.**

Methods. **Twenty-six heart-lung specimens without a coronary sinus draining to the right atrium were identified from the Leiden collection of congenital heart malformations. These were classified into specimens with an unroofed coronary sinus and those with atresia of the coronary sinus orifice. Attention was paid to the associated cardiac malformations.**

Results. **In 14 (54%; confidence limits [CL] 35%, 73%) of 26 specimens, there was an unroofed coronary sinus, associated with persistence of the left superior caval vein. An inferoposterior location of an atrial septal defect was detected in 2 (14%; CL −4%, 33%) of 14. Atrial appendage anomalies were seen in 13 (93%; CL 79%, 106%) of 14 specimens, exemplified by both right and left isomerism. These were frequently associated with an atrioventricular septal defect (12 [86%; CL 67%, 104%] of 14). An atretic coronary sinus orifice was seen in 12 (46%; CL 27%, 65%) of 26. Atrial appendage anomalies (2 [17%; CL −4%, 38%] of 12) were rare in these cases. The drainage was then by way of a left superior caval vein or, in its absence, a coronary sinus to left atrial window. Ventricular hypoplasia was seen in both categories of coronary sinus abnormalities. Important ventricular hypoplasia was seen in 12 cases (46%; CL 27%, 65%).**

Conclusions. **These findings emphasize the need to study coronary sinus drainage before procedures such as ligation or transcatheter coil embolization of a left superior caval vein, venous redirection or closure of a dorsal atrial septal defect are contemplated. These procedures might inadvertently lead to impairment of coronary venous return or persistence of an intracardiac shunt.**

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A postmortem investigation was performed of hearts without a coronary sinus draining into the right atrium. These were classified into those with complete unroofing of the coronary sinus and those with atresia of the orifice of the coronary sinus.

It is important to define the coronary sinus from a morphologic point of view. The coronary sinus is located posteriorly in the left atrioventricular (AV) groove. It is a saclike structure with its distalmost end delineated by the connection to the ligament of Marshall (the embryonic remnant of the left superior caval vein). Usually at this site a more or less recognizable valve of Vieussens is present; thereafter, the lumen continues as the greater cardiac vein. The middle and smaller cardiac veins drain into the coronary sinus along its length (1). The coronary sinus opens into the right atrium through a coronary sinus orifice that may be guarded by a thebesian valve of variable size. This orifice is located in the posterior and inferior walls of the right atrium superior to the tricuspid valve (Fig. 1).

The definition of a coronary sinus that does not drain into the right atrium includes cases with an absence of the partition between the coronary sinus and the left atrium. This condition is also referred to as complete unroofing of the coronary sinus (2). Sometimes the site of the coronary sinus orifice is still detectable as a posteroinferiorly located defect in the typical site of a coronary sinus orifice in the atrial septum. This anomaly has been well described by Raghib et al. (3). The site of the coronary sinus orifice may not be detectable in cases complicated by AV septal defect with or without isomerism of the atrial appendages (4,5). Anomalies of the coronary sinus may occur as isolated curios of little functional importance (6). However, when they occur in combination with other congenital cardiac defects, they assume greater significance, particularly if surgical intervention is contemplated (2).

Less attention has been paid to cases comprising atresia of the right atrial coronary sinus orifice and their implications for the management of children with congenital heart disease (7).

The coronary sinus can be well visualized with two-dimensional cross-sectional echocardiography, particularly in young infants, and its pattern of drainage can be ascertained by Doppler study. As more infants with complex cardiac anoma-

From the Department of Paediatric Cardiology, Universitätskinderklinik, Freiburg, Germany and the *Department of Anatomy and Embryology, University of Leiden, Leiden, The Netherlands.

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Address for correspondence: Dr. Adriana C. Gittenberger-de Groot, Department of Anatomy and Embryology, University of Leiden, P.O. Box 9602, 2300 RC Leiden, The Netherlands.

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Figure 1. View into the right atrium (RA) indicating the site of a normal right atrial orifice of the coronary sinus (CS, **arrowhead**). There is a muscle band between the closed foramen ovale (FO, **asterisk**) and the coronary sinus. Muscle tissue is also present between the coronary sinus and the tricuspid valve (TV).

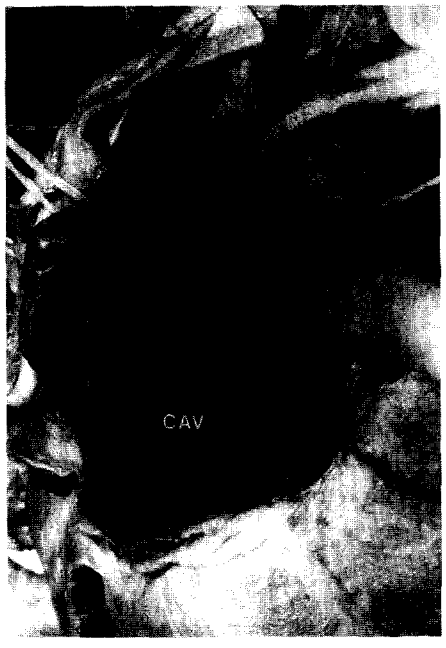

Figure 2. Heart with completely unroofed coronary sinus and no discrete inferoposterior atrial septal defect, in the setting of an atrioventricular (AV) septal defect. View from the right atrium (RA). The lower rim of the atrial septum (AS) merges with the posterior wall of the atrium. The common AV valve (CAV) is seen. There is a left-sided atrium with right atrial morphology ("RA"); in this structure, into which the left superior vena cava opens directly, there is no indication of a coronary sinus. SVC = superior caval vein.

lies are accepted for cavopulmonary connection and operations of the Fontan type, knowledge of coronary sinus and venous anomalies will become more important (8). We therefore undertook a pathologic study to assess the morphology of the coronary sinus, its drainage and associated cardiac malformations when there was either complete unroofing of the coronary sinus or atresia of its connection to the right atrium.

Methods

From the records of the collection of congenital heart malformations of the Department of Anatomy in Leiden, we identified 26 specimens without identifiable drainage of the coronary sinus into the right atrium. The specimens were classified into two major groups. In group I (14 of 26), all specimens had a completely unroofed coronary sinus. Two of these 14 were identified as having an inferoposteriorly located atrial septal defect at the site of the expected coronary sinus orifice. In group II (12 of 26), all specimens had atresia of the coronary sinus orifice. Four of the 12 specimens within this group had partial unroofing of the coronary sinus or left atrial window. We paid particular attention to the associated right and left superior caval vein drainage, the presence of atrial appendage anomalies, the anatomy of the atrial septal defect and imbalance in ventricular size. The results are presented as percentages with 95% confidence limits (CL), as well as the number of cases in each group.

Results

Group I (14 hearts with no recognizable coronary sinus). In 12 (86%; CL 67%, 104%) of 14, there was a complete unroofing of the coronary sinus and no discrete right atrial orifice (Fig. 2). Bilateral superior caval veins were present in 11 (92%; CL 76%, 107%) of these 12 specimens; the remaining specimen had an atretic right superior caval vein in combination with left atrial isomerism. The persistent left superior caval vein drained in all cases into the left-sided atrium at the junction of the atrial appendage and roof of the atrium anterior to the pulmonary vein connection. Atrial isomerism was present in 10 (83%; CL 62%, 104%) of 12, with 6 specimens (50%, CL 22%, 78%) showing left isomerism and 4 (33%; CL 7%, 60%) showing right isomerism. The diagnosis of left or right isomerism was based on the characteristic shape of the atrial appendages. Additionally, a broad internal entrance to the appendage was diagnosed as typical of a right atrial appendage. Juxtaposition of the left atrial appendage was seen in one case as well as situs solitus in another. All hearts had an AV septal defect, always involving the posteroinferior atrial rim; that is, in the expected position of the coronary sinus orifice, which could not be identified. In all cases the defect in the atrial septum was always more extensive, merging with the ostium primum defect of the AV septal defect. Thus, all hearts in this group had as unifying features a persistent left superior caval vein draining into the left-sided atrium, a defect in the posteroinferior portion of the atrial septum, an abnormality of atrial appendage lateralization with either isomeric or juxtaposed appendages (except for one case with situs solitus) and an AV septal defect. Within this group of 12 cases, ventricular imbalance occurred in 5; in 4 (33%; CL 7%, 60%) of the 12, the left ventricle was hypoplastic and in 1 (8%; CL −7%, 24%) of the 12, there was a small right ventricle.

Two of 14 hearts (14%; CL −4%, 33%) were identified with

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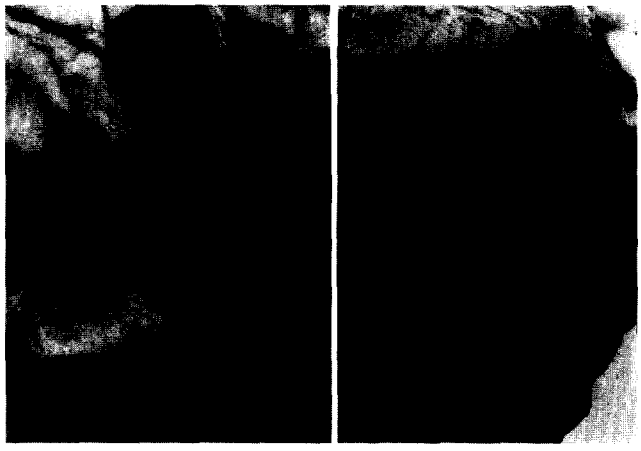

Figure 3. a, Unroofed coronary sinus with a discrete atrial septal defect in an inferoposterior position in the setting of situs solitus. Posterior view of the atria with the left superior vena cava (LSCV) entering the left atrium (LA) at the junction with the left atrial appendage (LAA). There is a large posteriorly located atrial septal defect (**arrow**) in a characteristic site posteroinferior to the fenestrated foramen ovale (FO). **b,** Right atrial view of the same specimen. Discrete inferoposterior atrial septal defect (**arrow**). The foramen ovale is patent and is also fenestrated. Between the foramen ovale and the atrial septal defect, an atrial septal muscle band is visible (**asterisk**), which is also indicated in **panel a**. TV = tricuspid valve.

Figure 4. Posterior view of a heart with coronary sinus orifice atresia. The left superior caval vein (LSVC) is patent. The coronary sinus (CS) is dilated, and the site of the atretic orifice is indicated by a dimple (**arrow**). LA = left atrium; LV = left ventricle; RA = right atrium.

a left superior caval vein, complete unroofing of the coronary sinus but with a discrete inferoposterior atrial septal defect circumscribed by atrial septal tissue. Unlike the hearts in group I, these two hearts did not have an AV septal defect. The atrial appendage morphology showed juxtaposition in one and situs solitus in the other case. Thus, recognition of a right atrial coronary sinus orifice when there is complete unroofing of the sinuatrial wall seems to be dependent on the extent of deficiency of the atrial septum. This is illustrated in Figure 3, which shows a heart with a normal atrial appendage arrangement. The site of the atrial septal defect was inferoposterior; the only other interatrial communication occurred through a fenestrated foramen ovale.

Group II (12 hearts with atresia of the coronary sinus orifice). In this group, when there was a left superior caval vein (8 [67%; CL 40%, 93%] of 12), the major route of coronary sinus drainage was through this vein (Fig. 4). In 4 (33%; CL 7%, 60%) of the 12 hearts, a left atrial coronary sinus window was present; that is, the coronary sinus was partially unroofed (Fig. 5, a and b). In three of these four cases, the left superior caval vein was absent; in one (25%; CL −17%, 67%) of the four, it was extremely diminutive (Fig. 5c). In none of these specimens was an atrial septal defect present in the characteristic position seen with complete unroofing of the coronary sinus. Rather, the expected site of the coronary sinus orifice was marked by a small dimple or depression or there was no recognizable vestige. The coronary sinus itself was easily recognizable in all cases and in 3 (25%; CL 0.5%, 50%) of the 12, it was dilated (Fig. 4). In 10 (83%; CL 62%, 104%) of 12 hearts, the right atrial coronary sinus orifice was absent in the setting of a probe patent oval foramen or intact atrial septum. Atrial isomerism (left-sided in 1 and right-sided in 1) was seen in 2 (17%; CL −4%, 38%) of 12 hearts. These were the only two specimens in this group with an AV septal defect. In no case did atresia of the coronary sinus appear to be due to an imperforate or obstructing thebesian or persistent right venous valve.

Ventricular imbalance occurred in 7 (58%; CL 30%, 86%) of the 12 hearts with atresia of the coronary sinus orifice. Hypoplasia of the right ventricle was present in 4 (33%; CL 7%, 60%) of the 12, and hypoplasia of the left ventricle in 3 (25%; CL 0.5%, 50%).

Discussion

Terminology. This study focuses on some important considerations in detecting coronary sinus anomalies. However, the terminology poses difficulties, because complete unroofing and absence of the coronary sinus are sometimes used as synonyms. It is more difficult to define the absence than the presence of the coronary sinus. If the orifice of the coronary sinus is identifiable, as in two of our specimens, and if the orifice is considered part of the sinus, one is not wholly justified in regarding this condition as complete absence of the coronary sinus, even though Raghib et al. (3) chose this term to describe a similar morphologic complex. It is impossible to judge by gross anatomic inspection whether the anterior or the posterior wall of the coronary sinus is absent. Therefore, when there is no identifiable coronary sinus orifice or wall separating the coronary sinus and the left atrium, and a left superior caval vein is present, the term complete absence of the sinoatrial wall would be applicable. However, this terminology is based on the embryologic concept of incorporation of the sinus venosus into the atrium in which a sinoatrial wall between the left atrium and coronary sinus is formed. Because knowledge of the development of the coronary sinus and its orifice wall is still incomplete, we have chosen a pure morphologic descrip-

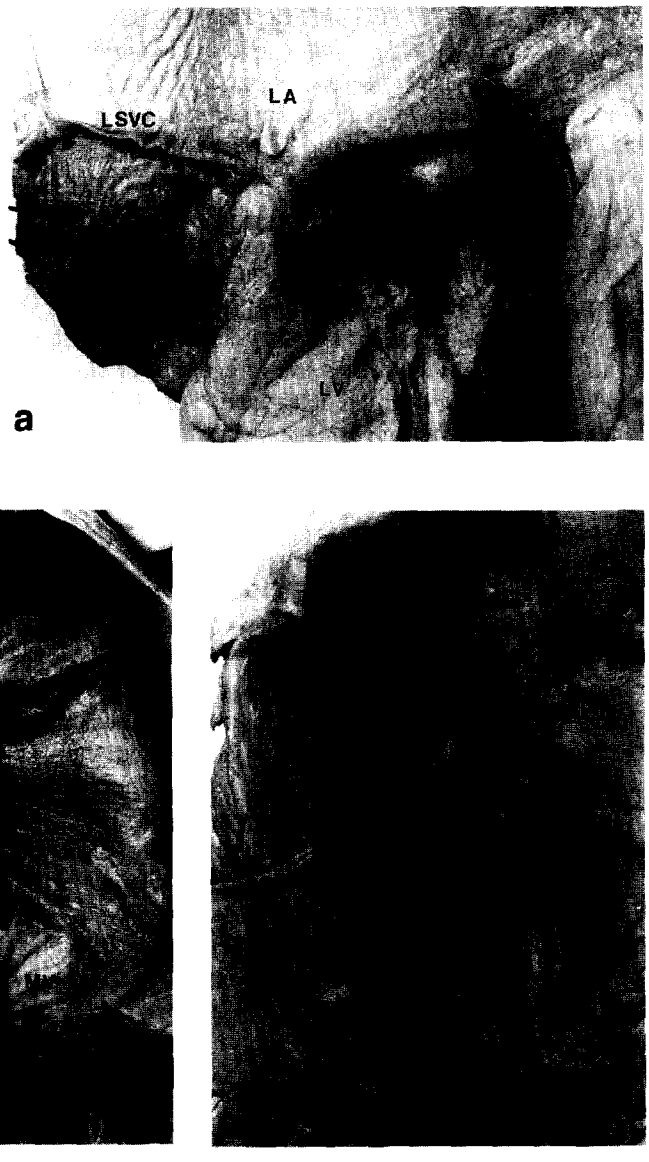

Figure 5. Heart with coronary sinus orifice atresia. **a,** View of the posterior wall of the left atrium (LA) and left ventricle (LV). The coronary sinus (CS) is connected to several veins (V) that course over the posterior surface of the ventricles. The left superior caval vein (LSVC) is diminutive. **b,** View into the left atrium. The wall of the coronary sinus shows a window (**arrow**). MV = mitral valve; VFO = valve of the foramen ovale. **c,** View into the right atrium (RA). The **asterisk** indicates the site of the atretic orifice of the coronary sinus. There is an Ebstein's anomaly of the tricuspid valve (TV). The atrialized part of the ventricle is indicated by **arrows**. FO = foramen ovale; SVC = superior caval vein.

tion, that is, a partial or complete unroofing of the coronary sinus. In the complete form it may still be possible to distinguish a discrete inferoposteriorly positioned atrial septal defect that would indicate the site of the coronary sinus orifice. This nomenclature has been used in the clinically relevant published reports (2).

Relation to AV septal defect and atrial isomerism. The possible developmental relation between an AV septal defect, atrial appendage anomalies such as isomerism and juxtaposition and coronary sinus abnormalities are still unclear. However, it is evident from our data that atrial situs is not a reliable method of distinguishing between unroofed coronary sinus and coronary orifice atresia. Nevertheless, both AV septal defects and atrial isomerism are far more obvious in the unroofed coronary sinus group. Our two specimens with a discrete coronary sinus orifice had no AV septal defect, a finding that suggests that the "atrial ostium primum anomaly" in AV septal defects renders a discrete coronary sinus defect unrecognizable.

In all but one specimen with right isomerism, there was a completely unroofed coronary sinus. It is debatable whether, from a developmental point of view, it is possible for a coronary sinus to be present with right isomerism. Nevertheless, the one specimen in our series clearly had abnormal atrial appendage lateralization and could not be described as an AV septal defect with situs solitus. In addition, the clinical management of children rarely, if ever, depends on recognition of atrial appendage morphology but rather on the constellation of particular cardiac and other somatic defects. The presence of a coronary sinus in right isomerism has been reported before (4,9).

Atresia of the coronary sinus orifice. Atresia of the coronary sinus orifice has been commented on only rarely, and in at least a third of cases it is associated with other cardiac

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malformations (10–12). Atresia of the coronary sinus orifice in the absence of a left atrial window is associated with persistence of the left superior caval vein, and ligation or coil embolization of this vein would impair coronary venous return and lead to cardiac dysfunction, in the absence of sufficient additional thebesian veins draining directly into the cardiac chambers. This view is supported by a recent report (7) of the death of a child with progressive cardiac dysfunction and eventual myocardial infarction after an arterial switch procedure with concomitant ligation of a left superior caval vein. At postmortem investigation the coronary sinus orifice proved to be atretic. The only possible route of egress from the sinus would have been through the left superior caval vein, which had been ligated. Watson (13) demonstrated that obstruction of the left superior caval vein with a catheter led to bradycardia and cardiac dysfunction in a patient with atresia of the right atrial coronary sinus orifice, emphasizing the need for unrestricted drainage of a coronary sinus.

When a left superior caval vein is found, it is important to determine the patency of the coronary sinus orifice by two-dimensional echocardiographic imaging and to interrogate the direction of flow in the superior caval vein with Doppler ultrasonography. When anomalous pulmonary venous drainage or left AV valve atresia and a levoatriocardial vein have been excluded (14), upward flow in the left superior caval vein would indicate this to be the only egress of blood from the coronary sinus. Preservation of the left superior caval vein would then be mandatory. The absence of dilation of the coronary sinus in the presence of a left superior caval vein to indicate atresia of the orifice is an unreliable finding. Figure 4 illustrates atresia of the coronary sinus orifice, a left superior caval vein and dilation of the sinus. It has been well documented (15,16) that anomalous coronary sinus connections may be important alternative and beneficial routes of atrial decompression in hearts with AV valve atresia and restrictive interatrial communications. However, these alternative routes of atrial decompression may also mask atrial hypertension, tempting deferment of a balloon septostomy or atrial septectomy. A patent foramen ovale or secundum atrial septal defect should be distinguished from the defect that is seen in cases of complete unroofing of the coronary sinus. Closing the latter would leave a left superior caval vein draining to the left atrium.

We encountered, as mentioned, atresia of the coronary sinus orifice (with atrial drainage through a left superior caval vein or coronary sinus atrial window) with atrial appendage anomalies in both cases accompanied by an AV septal defect.

Decompression of the coronary sinus. In our series a left superior caval vein was an unusual finding in hearts with atresia of the coronary sinus ostium and a window from coronary sinus to left atrium. It was found in only one of our four hearts with these features and was extremely diminutive. Similarly, Falcone and Roberts (17) reported that in three of their four hearts with atresia of the right atrial orifice of the coronary sinus, there was a communication between the left atrium and the coronary sinus but no left superior caval vein. In contrast, Freedom et al. (16) reported a case with pulmonary atresia and intact ventricular septum with severe stenosis of the coronary sinus orifice, a coronary sinus to left atrial window and a left superior vena cava, and Rose et al. (15) reported a similar case with mitral rather than pulmonary atresia. Shinpo et al. (12) described three cases of coronary sinus atresia with mitral atresia and a large left ventricle with drainage of the cardinal veins through a left superior caval vein. It is conceivable that decompression of the coronary sinus through the more proximal fenestration is related to obliteration of the left superior caval vein more distally unless atrial pressures are unusually elevated because of AV valve atresia.

Development of coronary orifice atresia. We are not able to speculate on a mechanism for the development of coronary sinus orifice atresia as a result of our study. Prominent venous valves were not a notable finding in any of our specimens, even in the heart with pulmonary atresia with intact ventricular septum. Abnormalities of venous valves are either an incidental finding or are rarely the cause of inferior vena cava obstruction in adults (18,19). In contrast, persistent right venous valves or large eustachian and thebesian valves in children are often associated with developmental complexes involving underdevelopment of the right heart chambers and vessels (19–22). In only one report (23), of a child with tricuspid atresia who developed signs of poor cardiac output after a Glenn anastomosis, was obstruction to coronary sinus outflow from venous valves suggested. In that child, the major hemodynamic consequence appeared to be due to restriction of interatrial shunting by the spinnakerlike venous valve. Obstruction to the great cardiac vein by a thickened valve of Vieussens was reported (24) in an adult with an intact atrial septum and absence of the coronary sinus without a left superior caval vein.

Important hypoplasia of a ventricle was a frequent feature of our hearts with coronary sinus anomalies. Because patients with such anomalies are those most likely to undergo operations of the Fontan type, it is important that abnormalities of systemic venous return be carefully considered in this group.

References

1. Maros TN, Rácz L, Plugor S, Maros TG. Contributions to the morphology of the human coronary sinus. Anat Anz 1983;154:133–44.
2. Quaegebeur J, Kirklin JW, Pacifico AD, Bargeron LM. Surgical experience with unroofed coronary sinus. Ann Thorac Surg 1979;27:418–25.
3. Raghib G, Ruttenberg H, Anderson RC, Amplatz K, Adams P, Edwards JE. Termination of left superior vena cava in left atrium, atrial septal defect, and absence of coronary sinus. Circulation 1965;31:906–18.
4. Rose V, Izukawa T, Moes CAF. Syndromes of asplenia and polysplenia. Br Heart J 1975;37:840–52.
5. Macartney FJ, Zuberbuhler JR, Anderson RH. Morphological considerations pertaining to recognition of atrial isomerism. Br Heart J 1980;44:657–67.
6. Mantini E, Grondin CM, Lillehei CW, Edwards JE. Congenital anomalies involving the coronary sinus. Circulation 1966;33:317–27.
7. Yokota M, Kyoku I, Kitano M, et al. Atresia of the coronary sinus orifice. J Thorac Cardiovasc Surg 1989;98:30–2.
8. Rumisek JD, Pigott JD, Weinberg PM, Norwood WI. Coronary sinus septal defect associated with tricuspid atresia. J Thorac Cardiovasc Surg 1986;92:142–5.

9. Kirklin JW, Barratt-Boyes BG. Cardiac Surgery. 2nd ed. New York: Churchill Livingstone, 1993:1586.
10. Edwards JE. Anomalies of the coronary sinus. In: Gould SE, editor. Pathology of the Heart. 2nd ed. Springfield (IL): Charles C Thomas, 1960:431–2.
11. Gerlis LM, Gibbs JL, Williams GJ, Thomas GDH. Coronary sinus orifice atresia and persistent left superior vena cava. Br Heart J 1984;52:648–53.
12. Shinpo H, Van Praagh S, Parness I, Sanders S, Molthan M, Castaneda A. Mitral atresia with a large left ventricle and an underdeveloped or absent right ventricular sinus: clinical profile, anatomic data and surgical considerations. J Am Coll Cardiol 1992;19:1561–76.
13. Watson GH. Atresia of the coronary sinus orifice. Pediatr Cardiol 1985;6: 99–102.
14. Edwards JE, DuShane JW. Thoracic venous anomalies. Arch Pathol 1950: 517–37.
15. Rose AG, Beckman CB, Edwards JE. Communication between coronary sinus and left atrium. Br Heart J 1974;36:182–5.
16. Freedom RM, Culham JAG, Rowe RD. Left atrial to coronary sinus fenestration (partially unroofed coronary sinus). Br Heart J 1981;46:63–8.
17. Falcone MW, Roberts WC. Atresia of the right atrial ostium of the coronary sinus unassociated with persistence of the left superior vena cava: a clinicopathologic study of four adult patients. Am Heart J 1972;83:604–11.
18. Rosall RE, Caldwell RA. Obstruction of inferior vena cava by a persistent Eustachian valve in a young adult. J Clin Pathol 1957;10:40–5.
19. Yater WM. Variation and anomalies of the venous valves. Arch Pathol 1929;7:418–41.
20. Freedom RM, Patel RG, Bloom KR, et al. Congenital absence of the pulmonary valve associated with imperforate membrane type of tricuspid atresia, right ventricular tensor apparatus and intact ventricular septum: a curious developmental complex. Eur J Cardiol 1979;10:171–96.
21. Hansing CE, Young WP, Rowe GR. Cor triatriatum dexter. Persistent right sinus venosus valve. Am J Cardiol 1972;30:559–63.
22. Kauffmann SL, Andersen DH. Persistent venous valves, maldevelopment of the right heart, and coronary artery-ventricular communications. Am Heart J 1963;66:664–9.
23. Jones RN, Niles NR. Spinnaker formation of sinus venosus valve. Circulation 1968;38:468–73.
24. Foale RA, Baron DW, Rickards AF. Isolated congenital absence of coronary sinus. Br Heart J 1979;42:355–8.