Inferior sinus venosus defect associated with incomplete cor triatriatum dexter and patent foramen ovale

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Received 24 February 2005; accepted 20 April 2005
Available online 25 May 2005

KEYWORDS
Sinus venosus defect; Cor triatriatum dexter; Patent foramen ovale

Abstract Sinus venosus atrial septal defect (SVD) is a rare cardiac abnormality in adults. Particularly, the inferior type is difficult to depict by transthoracic echocardiography because of its infero-posterior location to the fossa ovalis. We describe the case of a 33-year-old woman whose chest X-ray taken during bronchopneumonia revealed a cardiomegaly. Further echocardiographic investigations showed an underlying inferior SVD, an incomplete cor triatriatum dexter and a large patent foramen ovale (PFO). The diagnosis was confirmed by cardiac magnetic resonance imaging (CMR) and during surgical repair.

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Case report
A 33-year-old woman, with a highly active and athletic lifestyle (e.g. she is an avid mountaineer) never experienced any symptoms. Because of suspected bronchopneumonia she underwent chest X-ray which unexpectedly showed an increased cardiothoracic ratio. Subsequently, she was transferred for further cardiologic investigations. A wide and fixed splitting of the second heart sound and a grade 3/6 crescendo—decrescendo systolic murmur at the second left intercostal space were the outstanding findings during the otherwise normal physical examination. A 12-lead electrocardiogram showed a normal sinus rhythm, a frontal plane QRS axis of >120° and an incomplete right bundle branch block.

The transthoracic echocardiography revealed a dilatation of the right side of the heart and an eccentric hypertrophy of the right ventricle. A fenestrated membrane divided the right atrium into two chambers. A minimal pulmonary
regurgitation and a mild tricuspid regurgitation were found. The systolic pressure gradient over the tricuspid valve was 27 mmHg. Left and right ventricular systolic functions were normal. From the subcostal view there was already strong evidence for an inferior interatrial communication with left-to-right shunt by colour flow imaging. The pulmonary artery was dilated and pulmonary to systemic flow ratio was estimated to be >2.0.

To confirm this diagnosis and to exclude other associated congenital abnormalities (e.g. anomalous pulmonary venous connections), a transoesophageal echocardiography (TEE) was performed. The large membrane in the right atrium was seen which extended from the crista terminalis to the inferior-lateral rim of IVC and sinus coronarius and to the infero-posterior part of the proximal ascending aorta (Fig. 1a, b). Colour flow and contrast imaging demonstrated a communication between the two resulting cavities expressing an incomplete cor triatriatum dexter or extensive Chiari network (Fig. 2a, b). A large PFO (about 6 mm of separation between septum primum and secundum) was detected with spontaneous left-to-right shunt and intermittent right-to-left shunt, especially during Valsalva’s maneuver (Figs. 1a, b and 2b). All of the four pulmonary veins were correctly connected to the left atrium. However, the close position of the incoming right lower pulmonary vein to the inferior interatrial defect lead to a drainage of pulmonary venous blood into the right atrium. Finally, after extensive search, the inferior SVD with an IVC overriding the interatrial communication could be visualized (Fig. 3).

Subsequently performed cardiac magnetic resonance imaging (CMR) confirmed the anatomical findings (Fig. 4). Right heart catheterization demonstrated an oxygen saturation of 66% in the SVC, 70% in the IVC and 85% in the pulmonary artery. This resulted in a Qp/Qs of 2.6.

Because of this large left-to-right shunt and the marked volume-overloaded right ventricle we recommended surgery to the patient.

The operation was performed with a minimal invasive approach through a right axillary incision. The cardiac defect was confirmed. The intra-atrial membrane was resected, the SVD was closed directly over the inferior pulmonary vein without compromising its flow, and the PFO was closed directly. The patient tolerated the operation well and had an uneventful recovery.

Discussion

Sinus venosus atrial septal defects (SVD) represent 5–10% of atrial septal defects (ASD). In contrast to the more common superior type in which the superior vena cava overrides the pathological interatrial communication, the inferior type with overriding inferior vena cava (IVC) represents an extreme rarity.

The diagnostic accuracy of SVDs by TEE is limited in adult patients. Reasons are the posterior location of the defect and the often impaired transthoracic view, particularly from subcostal — the major and reliable entry for diagnosis in children. Due to the proximity of the transducer to the posterior atrial region, TEE is clearly superior and became the diagnostic procedure of choice. Associated cardiac abnormalities like anomalous pulmonary venous connections can be identified at the same time. Therefore, TEE is recommended in any patient with unexplained dilatation of the right side of the heart.

ASDs are classified according to their location relative to the fossa ovalis, their proposed

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**Figure 1**  (a) Mid-oesophageal view. Diastolic stop frame at 66°. The large intra-atrial membrane (†). No fenestration is seen at this view. Large patent foramen ovale (+). (b) Mid-oesophageal longitudinal view. Diastolic stop frame at 89°. There is a clear gap (arrow) between the membrane (†) and the infero-posterior part of the ascending aorta. Ao: Ascending aorta, IVC: inferior vena cava, LA: left atrium, PA: pulmonary artery, RA: right atrium, S: interatrial septum.
embryogenesis, and their size. Those posterior to the fossa ovalis are SVDs.1 Regarding the anatomic structure and morphologic development of the heart, SVDs can hardly be defined as ASDs. Some studies investigated the anatomic features of this infrequent cardiac disorder. At autopsy studies Anderson and Brown established the definition of the true atrial septum as that part of the atrial wall which directly separates the two atria. In a normal heart this criterion is only fulfilled by the floor of the fossa ovalis together with its inferior rim. A true ASD accordingly occurs within the confines of the fossa ovalis.2,3 The key to diagnosis of an SVD, however, is an overriding SVC or IVC across the intact muscular rim of the fossa ovalis resulting in an interatrial communication beyond the confines of the true atrial septum.4 A defect in the wall that normally separates the right pulmonary veins from the SVC and the right atrium (RA) is echocardiographically detected in superior SVDs.5 Thus, the right pulmonary veins are unroofed and may drain into the SVC or the RA. This explains the common coincidence of superior SVD and anomalous right pulmonary venous connection or drainage (malconnection in up to 85%). Anomalous right pulmonary venous drainage is unusual in the few reported patients with inferior SVD.4 The wall that separates the IVC from the left atrium normally does not form a roof over the pulmonary veins.7 A deficiency in this wall, therefore, results only in an overriding IVC and not in an unroofing of pulmonary veins. In our patient with inferior SVD we could also demonstrate a normal connection of all pulmonary veins to the left atrium, but an anomalous drainage of the right lower pulmonary

Figure 2  Mid-oesophageal short axis view at 32°. Injection of agitated saline into the left antecubital vein. (a) Contrast solely in the anterior part of the right atrium which is divided by the membrane (+). (b) Same view. Filling of the entire right atrium after injection of a greater amount of contrast. (c) Demonstration of a right-to-left shunt through the patent foramen ovale (+). See Fig. 1 for abbreviations.

Figure 3  Mid-oesophageal view. Diastolic stop frame at 44°. Inferior sinus venous defect (○) with “overriding” inferior vena cava. See Fig. 1 for abbreviations.

Figure 4  Cardiac magnetic resonance imaging. A modified longitudinal view demonstrates the intra-atrial membrane (+) and the inferior sinus venous defect (○). PA: Pulmonary artery, RLPV: right lower pulmonary vein, RUPV: right upper pulmonary vein. See Fig. 1 for further abbreviations.
vein via the inferior interatrial defect into the right atrium.

The additional finding of an incomplete or partial division of the right atrium is also a relatively rare cardiac abnormality. The so called "cor triatriatum dexter" represents a remnant of the right valve of the sinus venosus. During the third embryonic month this structure divides the right atrium into two chambers: (1) the smooth sinus portion to which the vena cavae and the coronary sinus are connected, and (2) the muscular primitive atrial portion including the right atrial appendage and associated with the tricuspid valve. It normally regresses between the 9th and 15th week of gestation. Dependent on the degree of regression the patient can keep a complete division of the right atrium (cor triatriatum dexter), a fenestrated membrane or a network of threads and fibers (called Chiari's network). Minimal remnants are the eustachian and thebesian valves that guard the orifice of the IVC and the coronary sinus, as well as the crista terminalis. The relatively common Chiari's network was found in 1.3—4% of autopsy studies. It is defined as coarse or fine fibers in the right atrium extending from the orifice of IVC and/or coronary sinus to the upper wall of the right atrium or the interatrial septum. Association with arterial embolic events had been discussed but remain uncertain. Generally, the diagnosis of this congenital abnormality is not of clinical significance. There is a fluid transition between Chiari's network and incomplete cor triatriatum. In our patient we found a membrane in typical position. Fenestration of this membrane allowed flow between the two cavities as expression of an incomplete cor triatriatum. A large PFO could also be demonstrated. A frequent association between Chiari's network and PFO was previously reported (PFO in 24 (83%) of 29 patients). This may be due to the anomalous, embryonic-like flow pattern in the right atrium, where the network or membranes direct the blood flow from the IVC preferentially towards the interatrial septum. Thus, paradoxical embolism may be promoted.

To our knowledge a combination of inferior SVD and incomplete cor triatriatum has not yet been reported before. An embryonic malformation of the sinus venosus and an incomplete resorption of its right valve lead to this patho-anatomic picture.

References