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REVIEW

The Sinus Venosus Veno-Venous Bridge

Not a septal defect

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abstract: This review provides an update on the morphology of the sinus venosus defect. It was earlier believed that a 'common wall' separated the right pulmonary veins from the superior caval vein. In the sinus venosus defects, this wall was absent. Current evidence shows that the superior rim of the oval fossa, rather than forming a second septum or representing a common wall, is an infolding between the walls of the caval veins and the right pulmonary veins. The sinus venosus defect is caused by the anomalous connection of one or more pulmonary veins to a systemic vein. However, the pulmonary vein(s) retain their left atrial connections, leading to a veno-venous bridge that allows interatrial shunting outside the oval fossa. True atrial septal defects are located within the oval fossa or in the anteo-inferior buttress, while sinus venosus defects, ostium defects and coronary sinus defects are morphologically distinct from them.

Keywords: Heart Septal Defects, Atrial; Diagnostic Imaging; Sinoatrial Node; Vena Cava, Superior.

The incidence of congenital heart disease is approximately 9 per 1,000 live births. Defects providing the potential for interatrial shunting account for about one-tenth of such malformations.[1] Among these defects, approximately three-quarters are true deficiencies of the atrial septum, with the shunt usually being across the oval fossa, although a proportion are within the antero-inferior buttress—the so-called vestibular defects. About another sixth of the defects are atrioventricular, rather than atrial, septal defects. In these instances, shunting is confined at the atrial level because of the fusion of the bridging leaflets of the common atrioventricular valve to the crest of the scooped-out ventricular septum. Most of the remaining defects are made-up of the sinus venosus variety, although rarely interatrial shunting can be through the mouth of the coronary sinus. The latter arrangement is found when its walls and those of the left atrium are deficient. The key to understanding the lesions not found within the oval fossa or its antero-inferior buttress is that the shunting, although unequivocally interatrial, is not across an atrial septal defect.[2]

The typical sinus venosus defect is usually found at the superior cavoatrial junction, more rarely at the inferior cavoatrial junction, or sometimes intermediate between these variants. The defect itself was initially described as long ago as 1858, but the term "sinus venosus" did not appear until 1956.[3] It has now become accepted that this particular arrangement, involving interatrial shunting outside the confines of the atrial septum, is the consequence of anomalous connection of one, or more, of the pulmonary veins to a systemic vein, but with the pulmonary vein, or veins, themselves retaining their left atrial connection. It had earlier been suggested that the defects existed because of the "absence" of a purported "common wall" between the right pulmonary and superior caval veins.[5] This explanation, however, falls at the first hurdle. This is because the pulmonary and systemic veins, in the normal heart, have their own discrete walls.[6] The extraseptal channel that permits the shunting is a veno-venous bridge produced by the anomalous connections of the pulmonary veins.[6] The presence of the venous bridge also explains why the orifice of the superior caval vein does not always override the intact superior rim of the oval fossa. Such overriding is usually found but can vary such that, in some instances, the superior caval vein can retain its exclusive connection with the right atrium or, in other instances, be exclusively connected to the morphologically left atrium.[7] In this review, we summarise the evidence showing that the lesions are the consequence of anomalous systemic connection of pulmonary veins that retain their left atrial connections, this explanation accounting for all the various sinus venosus defects.[8]

THE ANATOMICAL BASIS OF THE DEFECTS

The essence of the defects is that they do not involve a deficiency of the oval fossa. Nonetheless, such so-called "ostium secundum" defects can co-exist with the sinus venosus variety. It follows, therefore, that a knowledge of normal atrial septal anatomy is the key to appreciating the abnormal morphology. The basis of the atrial septum is the floor of the oval fossa.

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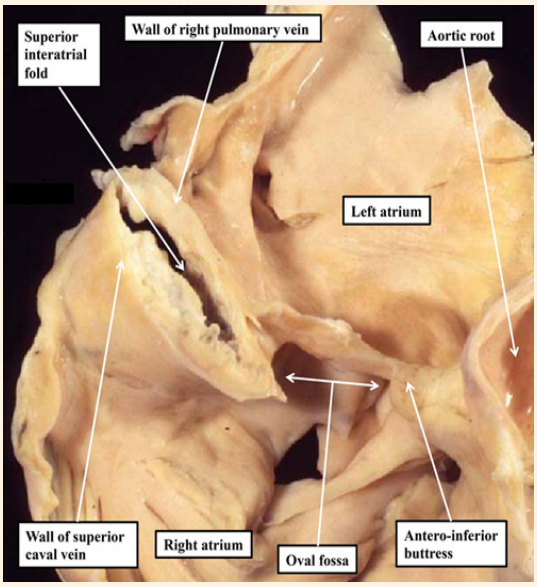

Figure 1: The image shows a "four-chamber" section through the atrial chambers of an adult heart. It shows the location of the oval fossa and its antero-inferior buttress adjacent to the aortic root. The buttress is the true second atrial septum, with the floor of the fossa derived from the first, or primary, atrial septum. The superior rim of the fossa, often described as the 'septum secundum', is no more than a deep infolding between the walls of the superior caval vein and the right pulmonary veins.

This structure is derived from the primary atrial septum, or 'septum primum'. It is anchored to the septal atrioventricular junction by the muscularised vestibular spine, with this component forming the antero-inferior buttress of the fossa. The antero-inferior buttress is the true second atrial septum. It is often thought that the superior rim of the fossa is the 'septum secundum'; this is not true. The rims of the fossa, apart from the antero-inferior buttress, are no more than infoldings of the walls of the right and left atrial chambers, with the superior rim being a deep fold between the attachments of the superior caval vein to the right atrium, and the right pulmonary veins to the left atrium [Figure 1].

In the normal heart, of course, the caval veins are exclusively connected to the morphologically right atrium, while the pulmonary veins are connected to the four corners of the roof of the morphologically left atrium. It is well-recognised, however, that the pulmonary veins can be anomalously connected to systemic sites. Such anomalous connections do not always involve all four of the pulmonary veins. It has now been recognised that, on occasion, one or more of the right pulmonary veins can be anomalously connected to a caval vein, usually the superior caval vein, while retaining their left atrial connection. This then produces a veno-venous bridge, which permits interatrial shunting outside the confines of the oval fossa. The presence of the veno-venous bridge is the essence of the sinus venosus defect. In most hearts with such anomalous connections of one or more of the right pulmonary veins, the superior caval vein attains a left atrial connection, with its orifice overriding the superior rim of the oval fossa [Figure 2A]. In a minority of cases, however, the superior caval vein can retain its exclusive connection to the right atrium, with the defect then being seen as a deficiency of its left wall [Figure 2B].

In still other instances, the caval vein can be almost exclusively connected to the morphologically left atrium, producing anomalous systemic rather than pulmonary venous connections. Nonetheless, the channel permitting interatrial shunting is always outside the confines of the normal atrial septum [Figure 3]. Hence, it is an interatrial communication rather than an atrial septal defect. This distinction obviously does not change the basic physiological situation, but as

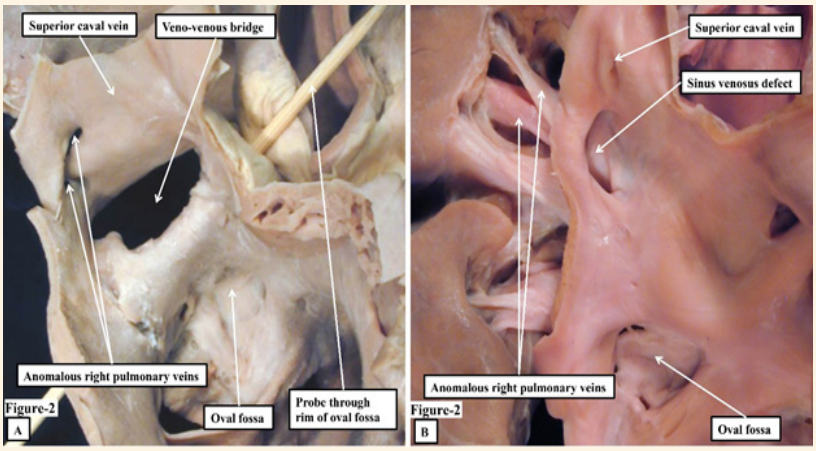

Figure 2: The images show typical sinus venosus defects as seen from the right side. **A:** The orifice of the superior caval vein is overriding the superior rim of the intact oval fossa, with a probe placed through the rim. **B:** The superior caval vein has retained its right atrial connection, with the defect distant from the intact oval fossa.

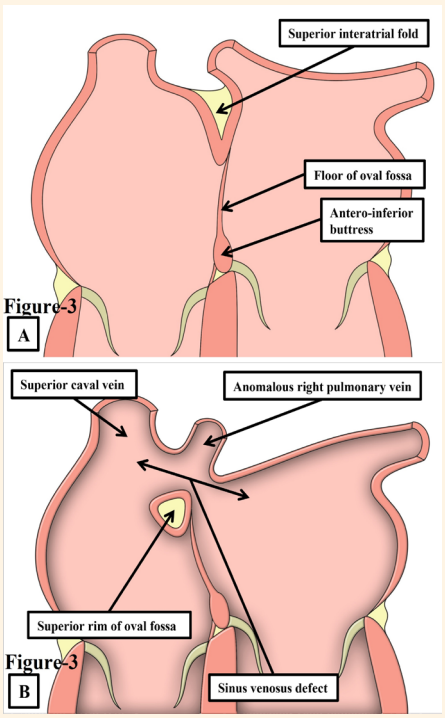

Figure 3: A: A diagram showing the normal anatomy of the atrial septum. **B:** A diagram showing how the anomalous connection of the right pulmonary vein, by retaining its left atrial connection, produces a veno-venous bridge outside the confines of the septum itself.

we will explain, it produces some nuances in clinical presentation. Understanding the true anatomical arrangement, however, is the key to determining the appropriate strategies for management.

CLINICAL FEATURES

Clinical presentation is frequently delayed, with some patients remaining asymptomatic until their third to fifth decade of life.[9,10] When the clinical manifestations appear, they are usually non-specific. The patients may present with exertional dyspnoea, easy fatiguability, arrhythmias, paradoxical embolism while straining, etc. Some patients have only come to attention when pacing leads or catheters were noted to enter the left-sided chambers during cardiac interventions.[11] Significantly, the left-to-right shunt caused by the veno-venous bridge is usually larger when compared to that produced by deficiencies within the oval fossa. This is due, in part, to the anomalous pulmonary venous connection contributing to the magnitude of the shunt. The extent of such additional shunting depends on the number of pulmonary veins anomalously connected, and the number of lung segments drained by the anomalous veins. Due to the large shunt, there is frequently significant dilation of the right-sided chambers and pulmonary arteries. As with any significant left-to-right shunt, this can lead to pulmonary vascular disease, pulmonary hypertension and Eisenmenger's syndrome. Again, because of the increased magnitude of the shunt, these complications occur earlier compared to isolated atrial septal defects, another reason for distinguishing between interatrial communication and septal defects.[12]

DIAGNOSIS

Diagnosis is primarily made by echocardiography. The correct decision cannot be reached without differentiating between the presence of a veno-venous bridge as opposed to a defect within the oval fossa, or its antero-inferior buttress. Nowadays, any patient with an incidentally detected cardiac murmur, presence of a fixed split-second heart sound, right heart enlargement on chest x-ray, or an abnormal electrocardiogram should undergo transthoracic echocardiographic examination. Although, when compared to the defects within the oval fossa, diagnosis of the sinus venosus defects is more challenging, it should not be difficult once their anatomical basis is understood. This explains both their location, and

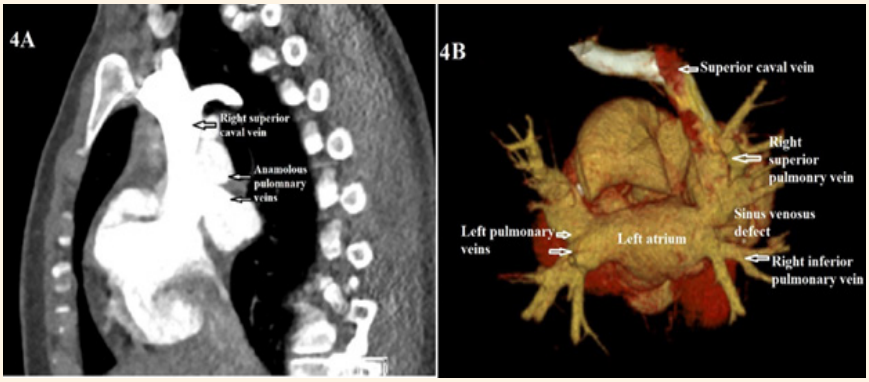

Figure 4: A: A computed tomography image in sagittal plane showing the superior pulmonary veins anomalously connected to the right superior caval vein. **B:** 3-D reconstruction of computed tomography image from the posterior aspect showing the right superior caval vein, right superior pulmonary veins draining to the right superior caval vein and a sinus venosus defect.

the associated pulmonary venous malformations.[13] Transthoracic echocardiography, nonetheless, should always demonstrate the haemodynamic effects of the shunt, with obvious dilation of the right-sided chambers and the pulmonary arteries.

Multimodal techniques such as cardiac computed tomography, cardiac magnetic resonance imaging, and cardiac catheterisation are now more frequently available. The anatomy can also be demonstrated precisely through the use of cardiac computed tomography with 3-dimensional reconstruction, cardiac magnetic resonance imaging, and transesophageal echocardiography [Figure 4; Supplementary Figure 1]. These techniques ensure accurate diagnosis before any attempted surgical or percutaneous intervention. They also provide additional information by the identification of anomalously connected pulmonary veins and demonstrating the precise connection of the superior vena cava. Cardiac magnetic resonance imaging has the added advantage of accurately quantifying ventricular volumes and the magnitude of the interatrial shunt.[14]

TREATMENT OPTIONS

Traditionally, surgical repair was the only treatment option for the defects. Its basic principle was to redirect the anomalous pulmonary venous drainage through the interatrial communication into the left atrium.[15] Although the operative mortality is very low, surgical repair is complex, and carries the risk of producing stenosis of either the superior caval vein or the pulmonary veins. Additional recognised problems are residual shunting, and sinus nodal dysfunction [Supplementary Figure 2].[16] These complications are likely to be minimised in future by use of robotic surgical repair.[17] The technique can involve use of a single patch or double patches, while the Warden technique can also be used.[18] It is now recognised, however, that repair can also be achieved by trans-catheter insertion of devices to separate the systemic and pulmonary blood streams.[19,20] The technique, however, is markedly different from the interventional approach used for defect within the oval fossa. This again emphasises the need to distinguish true atrial septal defects from the lesions that permit interatrial shunting outside the confines of the oval fossa. The first successful catheter-based repair of the sinus venosus defect had been reported by Butts *et al*.[6] In this report, they emphasised that the lesion, in essence, was a veno-venous bridge.[6] By 2021, an international registry reported the outcomes for 75 patients undergoing closure using covered Cheatham-platinum stents. The patients were aged between 11 and 76 years, with a median age of 45 years. In just over two-fifths, additional stents were required, either to anchor the first stent placed in the superior caval vein, or to close residual shunts. In two patients, embolisation of the stent had required surgical removal, while another patient needed drainage of cardiac tamponade. Follow-up studies showed occlusion of the right upper pulmonary vein in one patient. Overall, nonetheless, the results of interventional treatment were excellent.[21]

Conclusion

The understanding of the sinus venosus defects depends on an appreciation that they are not true defects of the atrial septum. Significantly, the shunting produced by the veno-venous bridges may remain undiagnosed for many years, with some patients only presenting in later life with symptoms of increased pulmonary cardiac output or pulmonary hypertension. The phenotypic feature is the presence of an anomalous pulmonary venous connection, but with the pulmonary vein, or veins, retaining their left atrial connections. This produces an interatrial communication, with increased left-to-right shunting, which leads, in turn, to dilation of the right-sided chambers and early onset of pulmonary vascular disease. A correct appreciation of the underlying anatomy is the basis not only of successful surgical repair but also for promoting the option for transcatheter intervention.

AUTHORS' CONTRIBUTION

MMM, RHA and SNM conceptualised the work. MMM and FK acquired the images. MMM, RHA and FK analysed and interpreted the images. MMM and RHA drafted the manuscript and contributed to the intellectual content. RHA, SNM and HNK critically reviewed and edited the manuscript. All authors approved the final version of the manuscript.

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