

SURGICAL TECHNIQUE I

Management of Sinus Venosus Defects

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Sinus venosus defects are not atrial septal defects, but are intra-atrial communications outside of the boundaries of the atrial septum. The superior type is located above and separate from the fossa ovalis, usually adjacent to the superior vena cava and the right upper pulmonary vein. The inferior type is located near the orifice of the inferior vena cava and the right lower pulmonary vein. The goal of surgical repair is closure of the defect with unobstructed drainage of the pulmonary veins to the left atrium and of the vena cava to the right atrium. Numerous techniques have been described, particularly for the repair of the superior vena cava type of defect. Mortality and morbidity should be minimal. The risk of either vena cava or pulmonary vein obstruction is low. Sinus node dysfunction can occur postoperatively, particularly when an incision has been made across the superior vena cava/right atrial junction. There is little long-term data on the functional outcomes following repair of these defects.

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Embryology and Anatomy

The exact nature of sinus venosus defects remains controversial. Sinus venosus defects are not atrial septal defects, but rather intra-atrial communications.^{1,2} The superior type, or superior vena cava (SVC) type is located above and separate from the oval fossa. It is usually associated with anomalous drainage of the right superior, and occasionally other, pulmonary veins to the SVC. Inferior, or inferior vena cava (IVC) type, defects are located near the orifice of the IVC.

The rim of the fossa ovalis, below the SVC orifice, represents an in-folding of the walls of the right atrium (RA) and left atrium (LA). This is the inter-atrial groove, also known as Waterston's groove, which can be dissected to improve access to the LA. Anderson et al⁴ suggested that superior sinus venosus defects result from a failure of in-folding of the atrial walls between the SVC and the right pulmonary veins. Van Praagh et al,² however, have suggested that the sinus venosus defects result from a deficiency in a common muscular wall that normally separates the right pulmonary veins from the

SVC, resulting in un-roofing of the right pulmonary veins allowing them to drain into the SVC or the RA. They contend that this mechanism can explain the etiology of both superior and inferior type sinus venosus defects. Immunohistochemical studies in human embryos using HNK-1 antigen expression as a marker for sinus venosus myocardium demonstrate that there is a common muscular wall between the sinus venosus and the common pulmonary vein during cardiac development.³

Despite the controversy over the embryology of the defects, the most important consideration for surgical correction of superior defects is the intimate relationship of the orifices of the SVC and the right upper pulmonary vein; or in the case of inferior defects, the orifice of the IVC and the lower pulmonary vein. Closure of these defects may result in obstruction of either pulmonary venous drainage to the LA or caval drainage to the RA. A second important anatomic concern is the location of the sinus node and the sinus node artery. Anatomy studies have shown that the most common location for the sinus node is the groove between the RA appendage and the lateral atrial wall, the sulcus terminalis (Fig 1).⁴ Some proposed techniques for repair of these defects incorporate incisions across the junction of the SVC and the RA, which could potentially injure the sinus node or the sinus node artery resulting in sinus node dysfunction. Ana-

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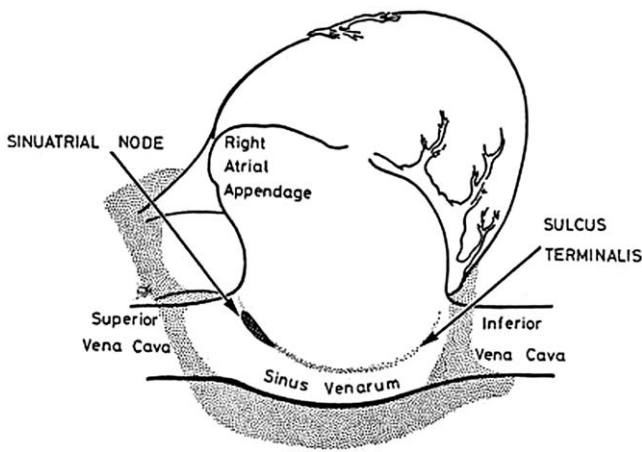


Figure 1 Diagram of the heart viewed from the right side of the chest showing the most common location of sinus node in the sulcus terminalis between the RA appendage and the lateral atrial wall. (Reprinted with permission from The BMJ Publishing Group.⁴)

atomic studies in humans have shown considerable variation in the anatomy of the arterial supply to the sinus node.⁴ The artery most commonly arises from the right coronary artery and approaches the sinus node either on the leftward medial aspect, or courses posteriorly around the SVC approaching the sinus node from the rightward or lateral side of the SVC (Fig 2).⁴ Depending on a patient's specific anatomy, an incision across the SVC/RA junction, either medially or laterally, might result in damage to the sinus node artery.

Diagnosis and Indications for Surgery

Transthoracic echocardiography is usually adequate for clearly defining the anatomy of sinus venosus defects and the associated anomalous pulmonary venous drainage as well as evaluating associated anomalies. The uppermost pulmonary veins may be difficult to visualize, but can be identified at the time of surgical repair. Use of magnetic resonance imaging or cardiac catheterization is rarely necessary. Sinus venosus defects do not close spontaneously. The presence of the defect is an indication for repair. Except in rare cases, elective repair should be performed to prevent long-term complications of RA and right ventricular volume overload.

Techniques of Surgical Repair

Surgical repair of sinus venosus defect consists of closure of the inter-atrial communication to create unobstructed drainage of the right pulmonary veins to the LA and of the SVC to the RA. Three common techniques have been utilized for repair of sinus venosus defects: 1) simple patch closure or baffling of the defect between the SVC/RA and right pulmonary veins. 2) patch closure of the defect with augmentation of the SVC-right atrial junction with a second patch and, 3) division of the SVC above the veins with re-implantation to the RA appendage and patch baffling of the SVC orifice, and

thus the anomalous pulmonary veins, to the RA. This is often termed the "Warden repair," although similar techniques have been independently described by others.^{5,6}

Simple patch closure of the defect may result in obstruction of the SVC. Because of this concern, use of a second patch placed across the SVC/RA junction has been advocated to augment this area and reduce the incidence of SVC obstruction. However, an incision across the SVC/RA junction may injure either the sinus node or the sinus node artery. In addition, use of a second patch does not eliminate the risk of SVC obstruction. An alternative repair is division of the SVC above the right pulmonary veins and re-implantation onto the RA appendage.⁵⁻⁷ The SVC is closed above the pulmonary veins and the orifice is closed with a separate patch (or an atrial wall flap), or by direct apposition of the inferior border of the defect to the SVC orifice, baffling the pulmonary venous drainage to the LA. Repair is usually performed through a median sternotomy. If the right pulmonary veins connect to

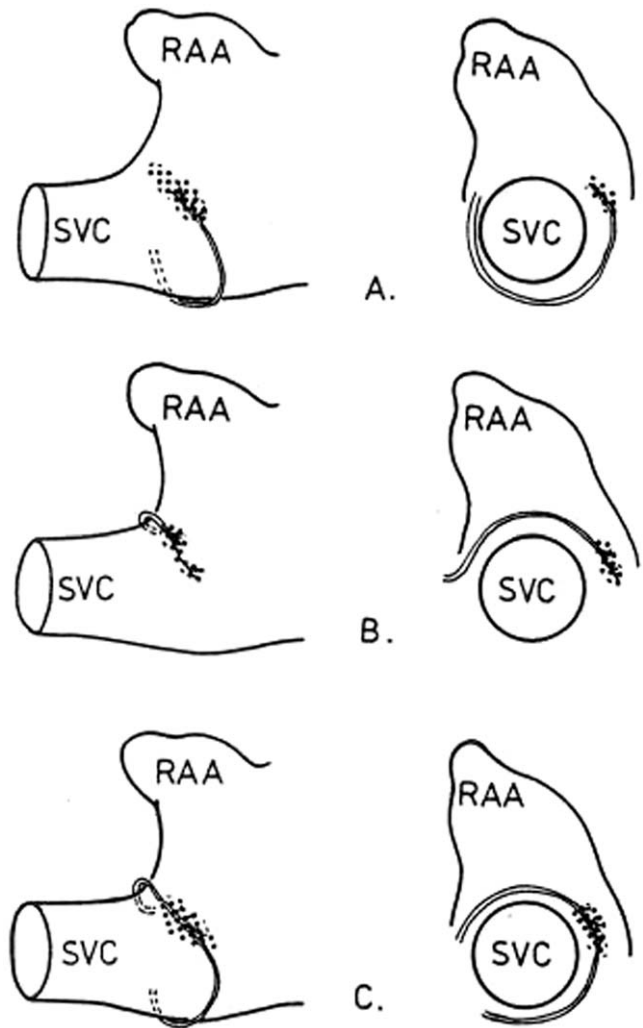


Figure 2 Diagrams illustrating the variable arterial supply of the sinus node. (A) Coursing posterior to the SVC; (B) coursing anterior to the SVC; (C) anterior and posterior arteries forming anatomic rings. SVC, superior vena cava; RAA, right atrial appendage. (Reprinted with permission from The BMJ Publishing Group.⁴)

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