Pediatric Cardiac Surgery Annual

SURGICAL TECHNIQUE I

Management of Sinus Venosus Defects

J. William Gaynor

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 fossal 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.

Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 9:35-39 © 2006 Elsevier Inc. All rights reserved.

KEYWORDS: Sinus venosus defect, anomalous pulmonary venous return, superior vena cava, heart surgery

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 al[4] 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-

Division of Cardiothoracic Surgery, The Children's Hospital of Philadelphia, Philadelphia, PA.

Address reprint requests to J. William Gaynor, MD, The Children's Hospital of Philadelphia, Division of Cardiothoracic Surgery, Suite 8527, Philadelphia, PA 19104; e-mail: Gaynor@email.chop.edu

1092-9126/06/$-see front matter
doi:10.1053/j.pcsu.2006.02.008

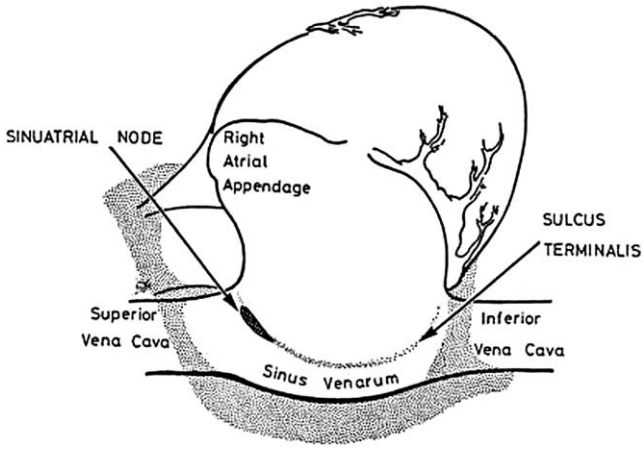

Figure 1 Diagram of the heart viewed from the right side of the chest showing the most common location of sinus node in the sulcus terminalis between the RA appendage and the lateral atrial wall. (Reprinted with permission from The BMJ Publishing Group.[4])

tomic 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 patients' 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

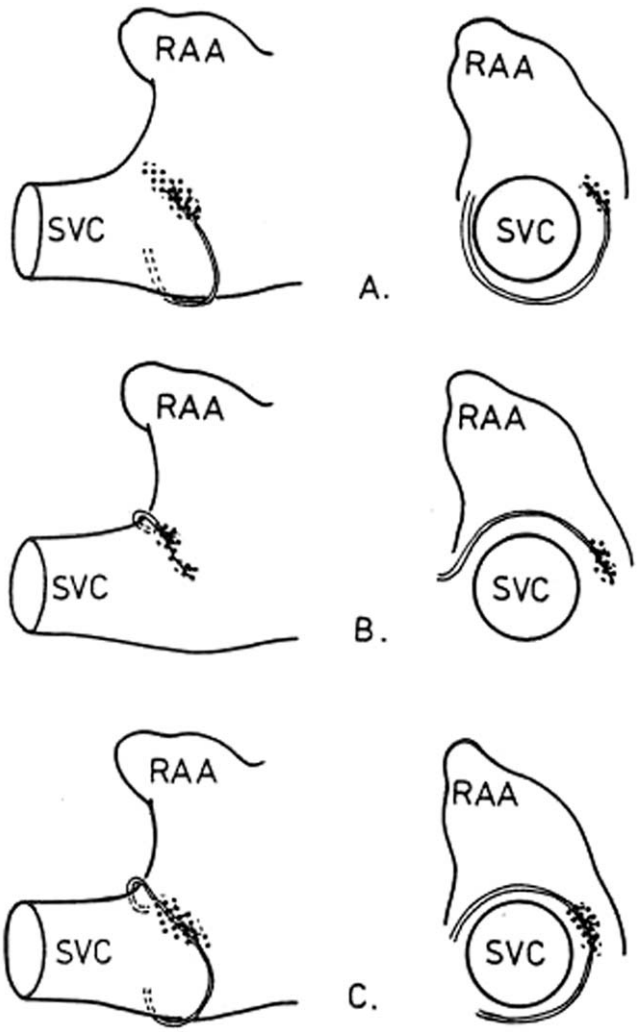

Figure 2 Diagrams illustrating the variable arterial supply of the sinus node. (A) Coursing posterior to the SVC; (B) coursing anterior to the SVC; (C) anterior and posterior arteries forming anatomic rings. SVC, superior vena cava; RAA, right atrial appendage. (Reprinted with permission from The BMJ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

the upper SVC, it may be useful to remove the thymus to improve exposure. The pericardium is opened and a portion is harvested for use as patches. If the defect is the superior type, it is necessary to mobilize the SVC superiorly above the insertion of the pulmonary veins to identify the uppermost vein. Care must be taken to avoid injury to the right phrenic nerve. Occasionally, a single pulmonary vein will insert very high on the SVC and maybe be excluded from the repair. If SVC division and reimplantation is planned, the azygous vein is doubly ligated and divided to improve mobility. The repair is performed using cardiopulmonary bypass at normothermia or mild hyperthermia (32° to 34°C) with bicaval cannulation. For repair of most superior type defects, the SVC cannula must be placed well above the insertion of the uppermost pulmonary vein, frequently at the junction of the jugular and innominate vein. However if a single patch repair is planned, it may be possible to position the SVC cannula via the right atrial appendage. The cannula functions as a stent to help prevent SVC narrowing during placement of the patch. After cannulation and initiation of cardiopulmonary bypass, tourniquets are placed around both cavae. Cardioplegic arrest is induced and the caval tapes are tightened. A right atriotomy is performed and the anatomy inspected. Additional secundum atrial septal defects are occasionally present. The position of all of the pulmonary veins must be carefully identified. Occasionally, the sinus venosus defect is restrictive and should be enlarged towards the fossa ovalis to prevent pulmonary vein obstruction. It may be possible to close the defect with a single patch baffling the pulmonary veins to the LA and avoiding SVC obstruction; however, this scenario is uncommon. More commonly, the right pulmonary veins insert well up on the SVC and placement of a single patch within the SVC would result in either pulmonary vein or SVC obstruction. There are two commonly used surgical options. One is to extend the atriotomy across the lateral aspect junction of the SVC/ RA junction to above the pulmonary vein insertion (Fig 3). A pericardial patch is used to close the defect baffling the pulmonary veins to the LA (Fig 4). A second patch is used to augment the SVC/RA junction (Fig 5).

An alternative approach is division and reimplantation of the SVC.[6-8] The SVC is divided above the insertion of the uppermost pulmonary vein. The cardiac end of the SVC is over sewn (Fig 6). The SVC is mobilized to allow anastomosis to the RA appendage. A pericardial patch is used to close the sinus venosus defect and the ostium of the SVC so that the stump of the vena cava and the pulmonary veins will drain in an unobstructed fashion to the LA. Alternatively a flap of atrial wall may be used to baffle the SVC orifice to the LA (Fig 7).[6] The tip of the RA appendage is incised. It is very important to divide all the trabulae within the appendage to ensure an unobstructed pathway. The divided SVC is attached to the tip of the RA appendage (Fig 8). If there is any concern over adequacy of the pathway, augmentation of the anastomosis of the SVC and appendage with a pericardial patch may be useful to prevent obstruction. If necessary to reduce anastomotic tension, a flap of atrial wall can be used to create a posterior wall for the pathway from the SVC to the RA.[5] Use

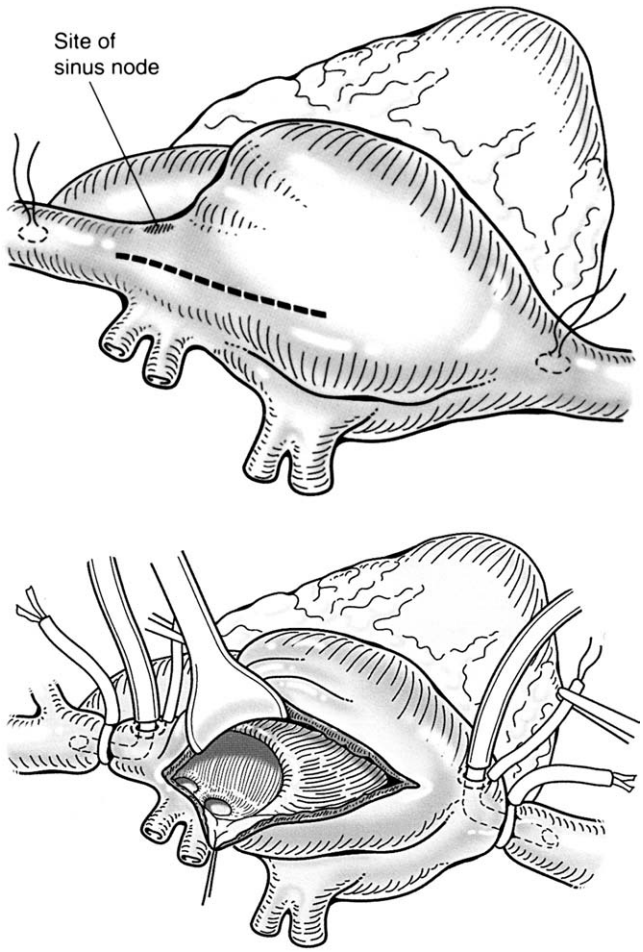

Figure 3 (A) The RA incision is oriented longitudinally and extended along the SVC/RA junction onto the SVC. The incision is carried to the upper limit of any anomalously connected pulmonary veins. (B) The anatomy is inspected to locate the pulmonary veins and any additional septal defects. (From Manning PB, Atrial Septal Defect, in *Operative Cardiac Surgery*, 5th edition, Arnold, 2004, reprinted with permission.)

of interrupted sutures may help prevent anastomotic narrowing if the SVC is small. The remainder of the pathway is constructed with a pericardial patch. The atriotomy is repaired and the patient is separated from cardiopulmonary bypass. Transesophageal echocardiography following separation from cardiopulmonary bypass is useful to ensure that there is no residual atrial shunting, as well as unobstructed drainage of both the pulmonary veins and the SVC. Sinus node dysfunction in the early postoperative period may occur even with SVC re-implantation, and it is helpful to place both atrial and ventricular pacing wires.

Inferior sinus venosus type defects can usually be managed with simple patch closure. It is important to realize that the defects may arise very close to the IVC. The IVC cannula should be positioned just above the insertion of the hepatic veins to improve access to the most inferior part of the defect near the IVC orifice. Following initiation of cardiopulmonary bypass and cardioplegic arrest, a right atriotomy is performed. The anatomy is inspected to evaluate possible addi-

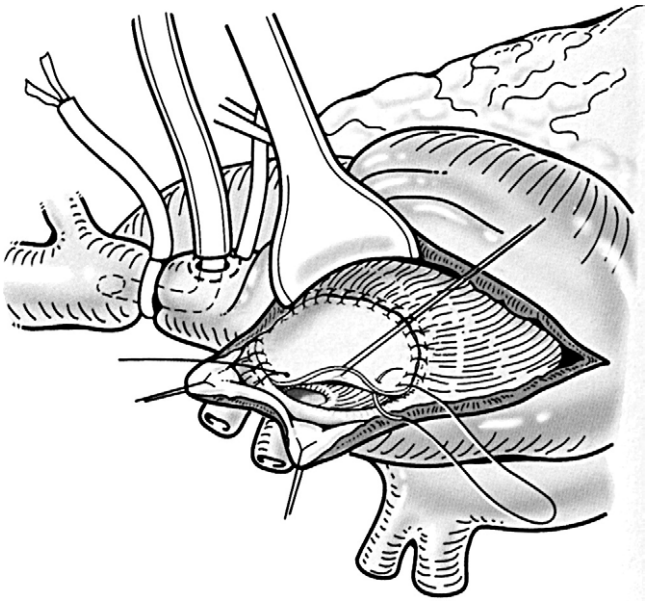

Figure 4 A pericardial patch is used to close the lower edge of the septal defect, and then continued up and around the superior edge of the highest pulmonary vein directing the pulmonary venous flow to the LA. (From Manning PB, Atrial Septal Defect in *Operative Cardiac Surgery*, 5th edition, Arnold, 2004, reprinted with permission.)

tional defects, the location of the IVC orifice, the sinus venosus defect, and the pulmonary vein insertion. It is rarely necessary to enlarge the IVC/RA junction. The defects can usually be closed with a single patch baffling the pulmonary veins to the RA. Care must be taken to avoid placing sutures in the eustachian valve, which can result in either IVC obstruction or diversion of the IVC to the LA.

A variety of other techniques have been described including patch closure via an incision in the SVC (cavotomy), avoiding incision in the atrium or SVC/RA junction.[8] There have been reports of minimally invasive approaches and robotic-assisted techniques.[9] Indications for these techniques have not been determined. Transcatheter techniques are not used because of the proximity of the pulmonary veins and the SVC to the defect.

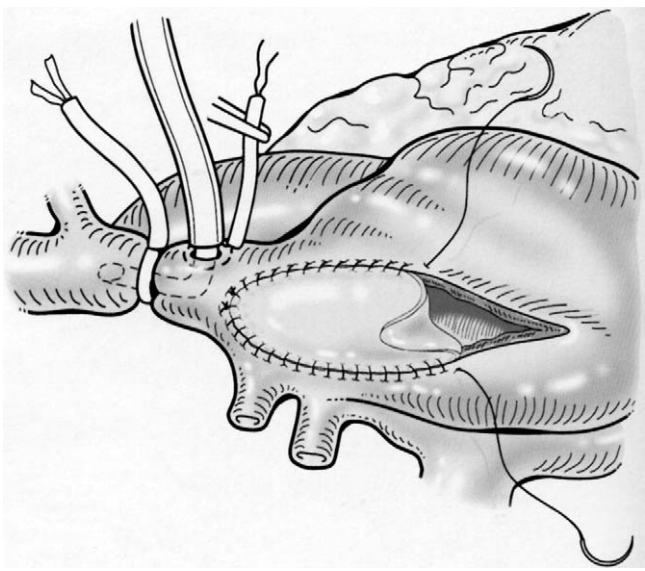

Figure 5 A second patch is used to augment the SVC/RA junction to ensure unobstructed drainage of the SVC to the RA. (From Manning PB, Atrial Septal Defect in *Operative Cardiac Surgery*, 5th edition, Arnold, 2004, reprinted with permission.)

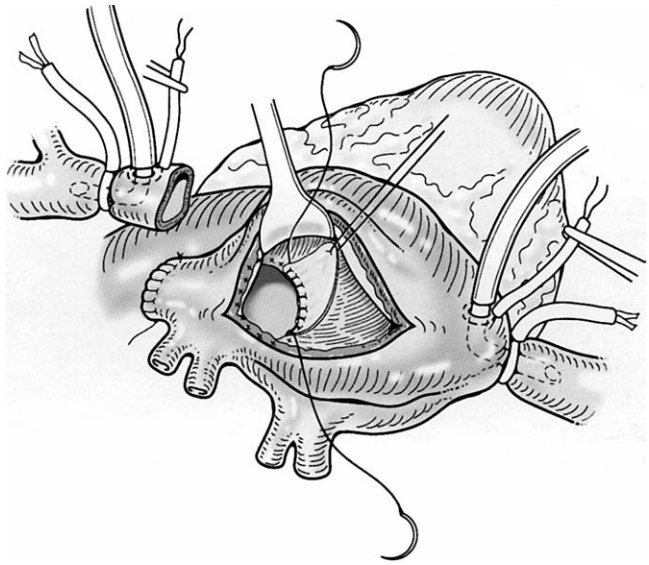

Figure 6 The SVC is divided and the cardiac end is over sewn. A pericardial patch is used to close the stump of the SVC and the sinus venosus directing pulmonary venous flow to the LA. (From Manning PB, Atrial Septal Defect in *Operative Cardiac Surgery*, 5th edition, Arnold, 2004, reprinted with permission.)

Outcomes

Mortality and morbidity should be minimal following repair of sinus venosus defects. Gaynor et al[6] reported repair of a variety of forms of anomalous pulmonary venous connection to the SVC by using diversion of the pulmonary venous drainage to the RA with an atrial wall flap with division and reimplantation of the SVC. Total anomalous pulmonary ve-

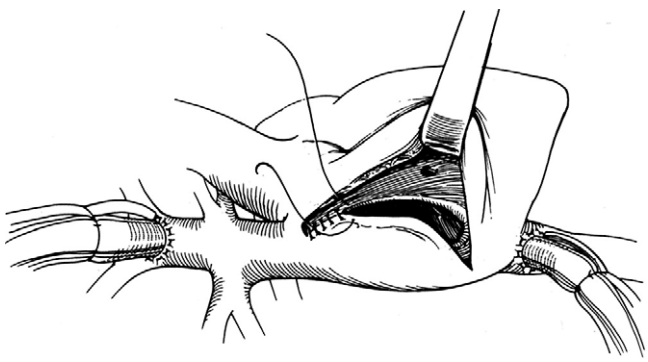

Figure 7 The posterior edge of the atriotomy is sutured around the SVC orifice and the anterior and inferior border of the defect creating an atrial wall baffle. (Reprinted with permission from The Society of Thoracic Surgeons.[6])

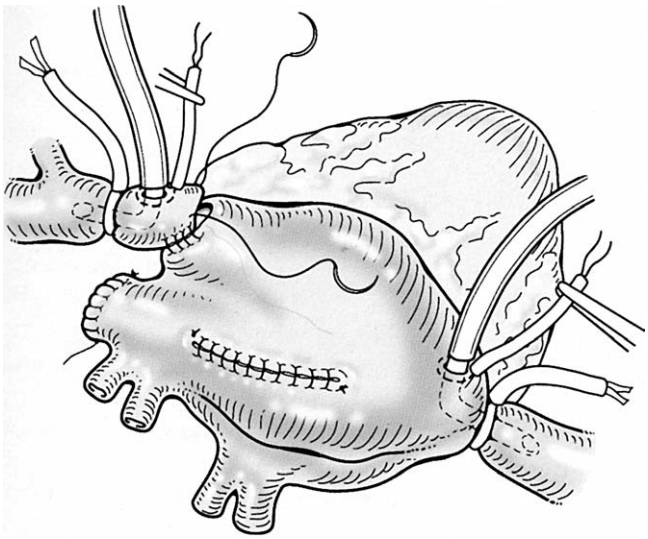

Figure 8 The tip of the RA appendage is incised. It is important to divide all trabeculae to ensure an unobstructed pathway. An anastomosis is performed between the divided SVC and the RA appendage. (From Manning PB, Atrial Septal Defect in *Operative Cardiac Surgery*, 5th edition, Arnold, reprinted with permission.)

nous connection was present in three patients and partial anomalous venous connection in eight. There were no operative deaths and no late deaths at a mean follow-up of 2.3 years. One patient required reoperation for pulmonary venous obstruction. No patient developed evidence of SVC obstruction and all were in sinus rhythm. Nicholson et al[8] reported outcomes of 66 patients undergoing repair of superior sinus venosus defect using a lateral transcaval approach. There were no operative deaths and no late deaths at a mean follow-up of more than 4 years. No patient had a residual atrial septal defect or evidence of vena cava or pulmonary venous obstruction. All patients had normal sinus node function at a mean of 7.3 years postoperatively. DiBardino et al[7] reported the outcomes of 16 patients undergoing repair of partial anomalous pulmonary venous connection to the SVC with the Warden procedure. There were no operative deaths and no late deaths. No patient developed sinus node dysfunction or obstruction to the SVC. A major concern following repair of these defects has been sinus node dysfunction, especially when an incision has been made across the SVC/RA junction. However, recent studies suggest that the risk of sinus node dysfunction is low for all of the described techniques.[10] Despite numerous reports on new operative techniques and early outcomes, there is very little long-term data on functional outcomes, the risk of venous obstruction, and late arrhythmias.

References

1. Al Zaghal JAM, Anderson RH: The nature of the superior sinus venosus defect. Clin Anat 11:349-352, 1998
2. Van Praagh S, Carrera ME, Sanders SP, et al: Sinus venosus defects: Unroofing of the right pulmonary veins – Anatomic and echocardiographic findings and surgical treatment. Am Heart J 128:365-379, 1994
3. Blom NA, Gittenberger-de Groot AC, Jonggeneel TH, et al: Normal development of the pulmonary veins in human embryos and formulation of a morphogenetic concept for sinus venosus defects. Am J Cardiol 87:305-309, 2001
4. Anderson KR, Ho SY, Anderson RH: Location and vascular supply of sinus node in human heart. Br Heart J 41:28-32, 1979
5. Warden HE, Gustafson RA, Tarnay TJ, et al: An alternative method for repair of partial anomalous pulmonary venous connection to the superior vena cava. Ann Thorac Surg 38:601-605, 1984
6. Gaynor JW, Burch M, Dollery C, et al: Repair of anomalous pulmonary venous connection to the superior vena cava. Ann Thorac Surg 59: 1471-1475, 1995
7. DiBardino DJ, McKenzie ED, Heinle JS, et al: The Warden procedure for partially anomalous pulmonary venous connection to the superior caval vein. Cardiol Young 14:64-67, 2004
8. Nicholson IA, Chard RB, Nunn GR, et al: Transcaval repair of the sinus venosus syndrome. J Thorac Cardiovasc Surg 119:741-744, 2000
9. Bacha EA, Bolotin G, Consillo K, et al: Robotically assisted repair of sinus venosus defect. J Thorac Cardiovasc Surg 129:442-443, 2005
10. Walker RE, Mayer JE, Alexander ME, et al: Paucity of sinus node dysfunction following repair of sinus venous defects in children. Am J Cardiol 87:1223-1226, 2001