TRANSCAVAL REPAIR OF THE SINUS VENOSUS SYNDROME

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Background: Surgical correction of the sinus venosus syndrome has been associated with sinus node dysfunction and venous obstruction postoperatively. We present the long-term follow-up of a lateral transcaval approach, which closes the atrial communication and corrects the partial anomalous pulmonary venous connection to the superior vena cava with the use of a simple pericardial patch.

Methods: The records of 66 patients undergoing repair between April 1981 and April 1997 were examined. Mean age at repair was 10.2 years (range, 1.5-65 years; median, 5 years). Six patients had a left superior vena cava, 4 had an additional atrial septal defect, and 2 had coronary artery bypass grafts. Immediate and long-term follow-up included physical examination, electrocardiography, transthoracic echocardiography, and use of a 24-hour ambulatory Holter monitor. Sinus node function, incidence of significant arrhythmia, and evidence of mechanical venous obstruction were assessed.

Results: Follow-up data were available for 64 (97%) patients for a mean follow-up of 4.1 years (range, 1-9 years). There were no deaths. No evidence of residual atrial septal defect, superior vena cava, or venous obstruction were found by echocardiography. On electrocardiography all patients were in sinus rhythm, with no arrhythmia seen. Holter monitoring was performed at a mean of 7.3 years postoperatively. All patients had normal sinus node function, and no sustained atrial arrhythmia was seen.

Conclusion: Transcaval repair is a simple technique that does not interfere with sinus node function. There is no evidence to suggest that this approach leads to venous obstruction. (J Thorac Cardiovasc Surg 2000;119:741-4)

Sinus venosus syndrome is defined as partial anomalous pulmonary venous connection of the right lung to the superior vena cava (SVC) with an associated sinus venosus atrial septal defect (ASD). This accounts for approximately 10% of patients presenting for surgery with an ASD. Various surgical techniques have been used to repair this defect involving incisions across the cavoatrial junction,[1,2] right atrial free wall muscle flaps,[3] and transection and relocation of the SVC to the right atrial appendage.[4] Surgical treatment has been associated with sinus node dysfunction and new and persistent atrial arrhythmia in up to 30% of patients.[2,5] Obstruction to the SVC and pulmonary veins has also been reported with these techniques.[4]

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Patients and methods

Between April 1981 and April 1997, 66 patients had transcaval repair of the sinus venosus syndrome. Patients with partial anomalous pulmonary venous connection to the right atrium or with inferior sinus venosus ASD were excluded from the study. Mean age at repair was 10.2 years (range, 1.5-65 years; median, 5 years). There were 36 male and 30 female subjects. Additional procedures included coronary artery bypass grafting in 2 patients and additional secundum ASD closure in 4 patients; 6 patients had a left SVC. Four patients required enlargement of the sinus venosus ASD so as not to restrict pulmonary venous return after repair.

The pattern of anomalous pulmonary venous connection to the SVC was determined by inspection at the time of repair and is summarized in Table I.

Follow-up included physical examination, electrocardiography, and transthoracic echocardiography as part of a routine postoperative protocol. To extend this follow-up, patients were asked to participate in further late follow-up involving a

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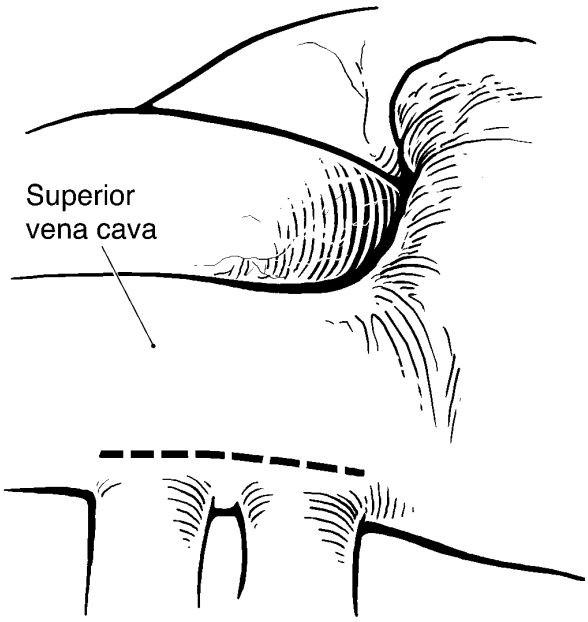

Fig 1. An incision is made in the lateral SVC wall at the junction with the anomalous pulmonary veins. This incision in wholly in the SVC and not the atrium.

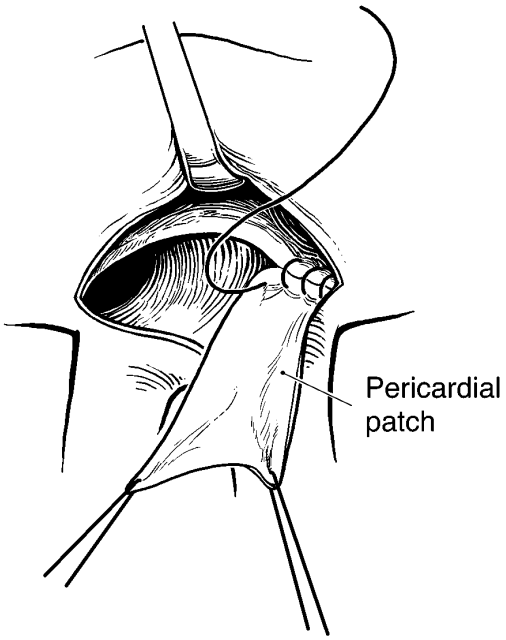

Fig 2. The pericardial patch is sutured to the medial, superior, and inferior margins of the defect within the SVC.

Table I. *Partial anomalous pulmonary venous connection to the SVC*

Origin of pulmonary vein	
RUL	17 (26%)
RUL, RML	39 (59%)
RUL, RML, RLL	10 (15%)

RUL, Right upper lobe; *RML,* right middle lobe; *RLL,* right lower lobe.

24-hour ambulatory Holter monitor to further assess sinus node function. This latter group of 38 patients represented patients greater than 4 years after surgical repair. This protocol was approved by the hospital ethics committee.

The venous structures were assessed for obstruction by 2-dimensional echocardiographic visualization and Doppler flow signal pattern. Three adult patients underwent thoracic computed tomographic scanning in addition to echocardiographic scanning to better visualize the repair site.

Operative technique. All patients had a median sternotomy approach, and continuous cardiopulmonary bypass was used. The aortic cannula was placed in a convenient position on the ascending aorta. Separate venous cannulas were placed, with the SVC either cannulated directly and high near the innominate vein junction or through the right atrial appendage, depending on the surgeon's preference. It is important to control the SVC venous return well above the region of the defect. A blood cardioplegic solution was given antegradely through the aortic root, and usually only one dose was necessary.

An incision was made in the lateral SVC wall over the insertion of the anomalous pulmonary veins (Fig 1). This incision was extended in a cephalad manner and also inferiorly to incorporate the junction of all anomalous pulmonary veins with the SVC in continuity. The incision was not taken across the cavoatrial junction. This gives excellent exposure both to the anomalous pulmonary venous orifices in the SVC and to the ASD.

There are three important landmarks in the repair. These are the ASD rim, the junction of the most superior anomalous pulmonary vein with the SVC, and the junction of the most inferior anomalous pulmonary vein with the SVC.

An untreated segment of autologous pericardium was used to baffle the anomalous drainage through the ASD into the left atrium. A double-armed 5-0 Prolene suture (Ethicon, Inc, Somerville, NJ) was used, beginning at the distant (medial) border of the ASD and continuing superiorly along the ASD rim to the superior junction of the anomalous vein and lateral caval wall and then exteriorized. The other needle was run in a similar manner along the inferior extent of the ASD rim to the inferior junction of the anomalous veins with the lateral caval wall and then exteriorized (Fig 2). The patch was then trimmed to allow approximately 3 mm of redundancy, which made a generous conduit for pulmonary venous return. It was important not to make the patch too redundant to avoid narrowing the effective SVC (systemic venous) dimensions after completion of the repair. The patch was then incorporated in the caval closure suture line as a "sandwich" (Fig 3). The venous return has then been efficiently partitioned with pulmonary and systemic venous components (Fig 4).

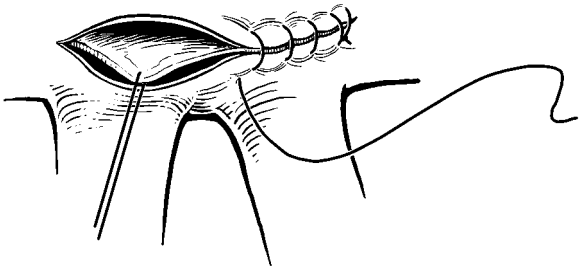

Fig 3. The SVC is closed by using the pericardial patch as a sandwich, thus completing the baffle and closing the defect.

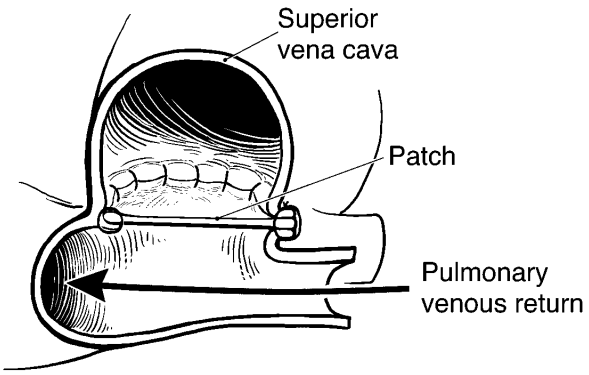

Fig 4. Looking into the right atrium from above, the SVC has been partitioned by the pericardial baffle into systemic and pulmonary venous channels.

Venous return from a left SVC was managed either by suction through the caval incision or a separate small stab incision in the right atrial free wall. The presence of an additional secundum ASD required a separate atrial incision in 3 patients.

The procedure was then completed with routine de-airing, and the patient was weaned from cardiopulmonary bypass.

Results

There were no deaths in the series. One patient had a neurologic episode thought to be related to air embolism, which resolved. All other patients had an uneventful postoperative course. Clinically, all patients were free of symptoms and had normal findings on physical examination at the time of follow-up.

Electrocardiography was performed at a mean follow-up of 4.1 years (range, 1-9 years). All patients were in sinus rhythm, and no new persistent arrhythmias were seen. There were 38 patients identified in the late follow-up group (>4 years postoperatively), and 30 (79%) patients agreed to have 24-hour ambulatory Holter monitoring performed at a mean of 7.3 years (range, 4-12 years) postoperatively. All patients maintained normal sinus node function, and there was no evidence of sustained atrial arrhythmia in any patient during the 24-hour Holter monitoring period.

Transthoracic echocardiography was performed on 64 (97%) patients at a mean follow-up of 4.1 years (0.25-9 years) postoperatively. There was no evidence of systemic or pulmonary venous obstruction with either 2-dimensional or Doppler flow studies, which confirmed findings on clinical examination. Right ventricular dimensions had returned toward normal limits in all patients, and no residual atrial level shunts were seen. Three adult patients underwent additional spiral computed tomographic scans to visualize the SVC–right atrial junction, which was not well seen on their transthoracic echocardiographic study. No evidence of SVC or pulmonary venous obstruction was seen on these studies.

Discussion

The repair of sinus venosus syndrome has involved the use of various techniques that have been associated with sinus node dysfunction, persistent atrial arrhythmias, and mechanical venous obstruction at the site of the repair. The ideal repair should not diminish SVC or pulmonary venous flow and should avoid injury to the sinus node and its artery.

Atrial arrhythmias are commonly reported both early and late after closure of an isolated ASD.[6,7] These arrhythmias are often asymptomatic in up to 45% of patients.[7] Interference with sinus node function may occur from either placement of an incision in the sinus node directly or from damage to the sinus node artery, which has a varied course in the region of the cavoatrial junction or in the lateral free wall of the right atrium.[8] Therefore incisions that cross the cavoatrial junction and involve the atrial appendage or the free wall of the right atrium have the potential to damage the sinus node arterial supply, and this may be important in the incidence of sinus node dysfunction recorded in some surgical series. Retraction of the sinus node without vascular injury may also contribute to postoperative sinus node dysfunction.

A lateral transcaval approach to repair of the sinus venosus syndrome avoids an incision in the region of the sinus node artery and still allows excellent visualization of the vital anatomic landmarks of the repair. In the majority of cases, there is minimal direct retraction on the sinoatrial node. The absence of atrial arrhythmia and the maintenance of normal sinus node function seen in our group of patients may be due to the

avoidance of the above factors by using the lateral transcaval pericardial baffle approach. All of our patients tested had normal sinus node function on echocardiography postoperatively, with no evidence of sustained arrhythmia, and the patients that underwent late Holter monitoring confirmed these findings. There are also other factors that may contribute to our observed improvement in postoperative sinus node function and rhythm disturbance unrelated to the technique of our repair. In the modern era, earlier diagnosis, a younger age at operation, and improvements in operative and perioperative management are likely to improve results compared with earlier series regardless of the technique used.

Additional secundum ASD can be repaired either through the transcaval approach if all edges are well seen or through an additional incision in the lower right atrial free wall.

The transcaval approach provides a simple approach for the baffling of anomalous pulmonary venous return through the sinus venosus defect to the left atrium with a pericardial patch. It is important not to make the pericardial patch too redundant because that may obstruct systemic venous return in the SVC. Closing the caval incision as a sandwich, incorporating the pericardial patch with approximately 3 mm of redundancy, appears to produce a repair with the correct patch dimensions. We did not augment the SVC with a separate patch, as advocated by some authors,[9,10] and have not experienced any late systemic or pulmonary venous obstruction in our patients clinically or on echocardiographic assessment. The SVC is usually enlarged in the region of the anomalous pulmonary veins, and the pericardial patch used as a baffle simply replaces the deficient posterior wall of the SVC. On echocardiography some patients had a slight “waisting” of the SVC reported, but no hemodynamic evidence of obstruction was seen, and similar techniques that do not enlarge the SVC have not been associated with venous obstruction on follow-up.[11]

This minimal lateral transcaval approach is simple and reproducible, does not cause venous obstruction, and maintains normal sinus node function in the long term.

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