Sinus Venosus Atrial Septal Defect
Long-Term Postoperative Outcome for 115 Patients

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Background—Sinus venosus atrial septal defect (SVASD) differs from secundum atrial septal defect by its atrial septal location and its association with anomalous pulmonary venous connection (APVC). Data on long-term outcome after surgical repair are limited.

Methods and Results—We reviewed outcomes of 115 patients (mean age ±SD 34 ±23 years) with SVASD who had repair from 1972 through 1996. APVC was present in 112 patients (97%). Early mortality was 0.9%. Complete follow-up was obtained for 108 patients (95%) at 144 ±99 months. Symptomatic improvement was noted in 83 patients (77%), and deterioration was noted in 17 patients (16%). At follow-up, 7 (6%) of 108 patients had sinus node dysfunction, a permanent pacemaker, or both, and 15 (14%) of 108 patients had atrial fibrillation. Older age at repair was predictive of postoperative atrial fibrillation (P = 0.033). No reoperations were required during follow-up. Survival was not different from expected for an age- and sex-matched population. Clinical improvement was more common with older age at surgery (P = 0.014). Older age at repair (P = 0.008) and preoperative New York Heart Association class III or IV (P = 0.038) were independent predictors of late mortality.

Conclusions—Operation for SVASD is associated with low morbidity and mortality, and postoperative subjective clinical improvement occurs irrespective of age at surgery. Postoperative atrial fibrillation appears to be related to older age at operation. SVASD repair achieves survival similar to that of a matched population and should be considered whenever repair may impact survival or symptoms. (Circulation. 2005;112:1953-1958.)

Key Words: heart defects, congenital ■ heart septal defects ■ surgery ■ survival

Sinus venosus atrial septal defect (SVASD), originally described in 1858, encompasses approximately 4% to 11% of atrial septal defects (ASDs).1,2 The typical malformation is an interatrial communication caused by a deficiency of the common wall between the superior vena cava (SVC) and the right-sided pulmonary veins.2,3 SVASD is commonly associated with anomalous pulmonary venous connection (APVC) of some or all of the pulmonary veins,2,3,4 which produces additional left-to-right shunting. The basic principle of repair is redirection of the APVC through the interatrial communication into the left atrium. In contrast to operative repair of secundum ASD, the surgical approach for SVASD is more complex and carries the risk of stenosis of the SVC or pulmonary veins, residual shunting, and sinoatrial node dysfunction (SND).4

The present study reviews outcomes for patients who underwent repair of SVASD at Mayo Clinic (Rochester, Minn) and focuses on patient survival and development of arrhythmias.

Methods

Patients

We reviewed 131 consecutive patients who underwent surgical repair of SVASD at Mayo Clinic between January 1972 and December 1996; these patients comprised 4.0% of all 3277 patients having an operation for ASD during this period. We excluded 16 patients with SVASD associated with severe congenital heart disease. Thus, the study cohort included 115 patients (mean age ±SD 34 ±23 years; range 1.5 to 80 years). Four patients (3%) were older than 70 years at operation, the oldest being 80 years. Six patients (5%) were operated on when they were younger than 5 years. Typical superiorly located SVASD was present in 109 patients, and 6 had an atypical inferior SVASD. Typical SVASD results from a deficiency in the wall that normally separates the right pulmonary veins from the superior vena cava and the right atrium.2,3 Rarely, there is absence of only the posterior or inferior portions of the atrial septum (or both), and 1 or more of the right pulmonary veins enters the right atrium anterior to the atrial septum; this is called atypical inferior SVASD. In the present series of patients because it is not typical SVASD.5 In 111 patients, the operation was the first attempt at repair. In 4, the operation was for failed repair performed elsewhere, including failure to divert 1 or more APVCs (n = 3) and recurrent SVASD (n = 1). The study was approved by the Mayo Clinic Institutional Review Board.

Data from 108 patients (95%) are available for follow-up, which was 23 years; range 1.5 to 80 years). Four patients (3%) were older than 70 years at operation, the oldest being 80 years. Six patients (5%) were operated on when they were younger than 5 years. Typical superiorly located SVASD was present in 109 patients, and 6 had an atypical inferior SVASD. Typical SVASD results from a deficiency in the wall that normally separates the right pulmonary veins from the superior vena cava and the right atrium.2,3 Rarely, there is absence of only the posterior or inferior portions of the atrial septum (or both), and 1 or more of the right pulmonary veins enters the right atrium anterior to the atrial septum; this is called atypical inferior SVASD. In the present series of patients because it is not typical SVASD.5 In 111 patients, the operation was the first attempt at repair. In 4, the operation was for failed repair performed elsewhere, including failure to divert 1 or more APVCs (n = 3) and recurrent SVASD (n = 1). The study was approved by the Mayo Clinic Institutional Review Board.

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The defect was exposed through the right atrium. The defect was exposed through an incision in the right atrium that extended across the cavoatrial junction into the superior vena cava. After right atrial incision shown in B. The pericardial patch redirected the anomalous pulmonary venous flow to the left atrium and closed the SVASD. AO indicates aorta; CS, coronary sinus; FO, foramen ovale; IVC, inferior vena cava; PT, pulmonary trunk; RLPV, right lower pulmonary vein (which connects appropriately to the left atrium in this diagram); RMPV, right middle pulmonary vein; and RUPV, right upper pulmonary vein. Reproduced with permission from the Mayo Foundation for Medical Education and Research.

ECG Findings

All ECGs were reviewed, and SND was defined as persistent sinus bradycardia (<50 bpm), ectopic atrial rhythm, junctional/nodal rhythm, or a wandering pacemaker (<60 bpm); pauses of more than 3 seconds; or evidence of SND on an electrophysiological study. New SND was defined as SND that was present at hospital dismissal or at late follow-up when sinus rhythm was present preoperatively. For patients with no SND at dismissal but no follow-up, the data were not analyzed and were considered missing.

Follow-Up Data

The date of latest follow-up for retrospective review was July 1, 1997. Postoperative follow-up information was obtained for 108 (95%) of the surviving 114 early survivors from a subsequent visit to Mayo Clinic before July 1, 1997 (n=60); phone call from the survey research department (n=28); visits to other clinics before July 1, 1997 (n=20); and information from the National Death Index (n=6). Six patients were lost to follow-up. The functional classification was assessed by use of the New York Heart Association (NYHA) classification. For the present analysis, clinical improvement was defined as subjective improvement reported by the patient. Subjective improvement included any improvement in exercise capacity or energy level or a decreased frequency of cardiovascular symptoms.

Statistical Analysis

Discrete variables were summarized as percentages, and continuous variables were summarized as mean±SD. Differences between the characteristics of 3 patient age groups were tested for significance with the χ² test or Fisher exact test when appropriate for discrete variables and with the 2-sample t test or rank sum test for continuous variables. Trends in baseline characteristics with age were assessed by the 2-sample t test (on age) for binary variables and the Spearman rank correlation for continuous variables. Simple and multiple logistic regression analyses were used to assess patient and surgical factors related to postoperative improvement. Simple and multiple Cox regression analysis was used to assess predictors of survival. Survival after the date of operation was estimated with the Kaplan-Meier method and was compared with the age- and sex-matched survival for the US population overall, as well as being stratified by 3 age groups: 40 years or younger, 41 to 60 years, and 61 years or older. All tests of significance were 2-tailed, with P<0.05 assumed to indicate significance. “Early death” was defined as occurring within 30 days after SVASD repair or during the index hospitalization. Other outcomes, including postoperative improvement in dyspnea and atrial fibrillation, were compared between the 3 age groups. The rates of these outcomes were compared between the 3 age groups by the Pearson χ² test of independence.

Results

The study group included 61 women (53%). Preoperatively, 50 patients (43%) were in NYHA functional class I. The diagnosis of ASD was suspected on the basis of a systolic murmur in the pulmonary area in 111 patients (97%), cardiomegaly, or recurrent pulmonary infections. NYHA functional class III or IV dyspnea was present in 20% of patients. Apart from dyspnea, the following symptoms and history were
reported: palpitations (31%), angina (19%), history of congestive heart failure (9%), and history of stroke (3%).

Preoperative hemodynamic data are shown in Table 1. Systolic pulmonary artery pressure (n=103) was >50 mm Hg in 21 patients (20%). Total pulmonary vascular resistance (n=45) was >5 U in 12 patients (27%) and >8 U in 6 of these patients (13%).

Forty-six patients (40%) were older than 40 years at the time of repair; 17 (15%) were older than 60 years, and 4 (3%) were older than 70 years. Patients older than 40 years at operation were significantly more likely to have preoperative dyspnea (P<0.0001), atrial fibrillation or flutter (P<0.0001), and a higher pulmonary vascular resistance (P<0.0001) and pulmonary arteriolar resistance (P=0.001).

**Operations**

The SVASD was classified according to the surgical findings into the typical superior type (n=109) or the atypical inferior type (n=6). In 112 patients, there was associated APVC, with insertion of pulmonary veins into 1 or more of the following: SVC, cavoatrial junction, and right atrium. The pulmonary vein anatomy was not described for 4 patients.

The mean diameter of the SVASD intraoperatively was 22±11 mm (range 5 to 60 mm). A persistent left SVC to the coronary sinus was found in 17 patients (15%). An associated secundum ASD was present in 10 patients (9%), and a patent foramen ovale was present in 20 (17%). In addition to SVASD repair, the following surgical procedures were performed: CABG (n=2), tricuspid valve replacement (n=1), tricuspid valve annuloplasty (n=5), excision of benign pericardial tumor consisting of mesothelial cells (n=1), cryoablation for arrhythmias (n=1), and division of the left vertical vein with anastomosis to the left atrial appendage for repair of anomalous pulmonary venous return from the left lung (n=1). All secundum ASDs and patent foramina were closed.

**Early Complications**

There was 1 early death (0.9%) in a 76-year-old woman with preoperative NYHA class IV who underwent patch closure of SVASD, suture closure of a patent foramen ovale, and insertion of a 31-mm Hancock tricuspid valve in 1974. The preoperative systolic pulmonary arterial pressure was 35 mm Hg. Six days postoperatively, she died of right-sided heart failure. Serious postoperative morbidity occurred in 2 patients. A 59-year-old woman with chronic atrial fibrillation and a history of multiple strokes preoperatively had a large nonhemorrhagic stroke on postoperative day 6. A 36-year-old woman developed an embolic left femoral artery occlusion on postoperative day 2 while in sinus rhythm; embolectomy was successful.

**ECG Findings**

The preoperative ECG was normal in 7 patients (6%). It demonstrated right bundle-branch block, right ventricular hypertrophy, or right axis deviation in 82 patients (71%) and SND in 2 (2%). First-degree atroventricular block was seen in 5 patients (5%). Paroxysmal or chronic atrial fibrillation or flutter was present in 12 (10%). Complete atroventricular block was rare and was found in 1 patient. One patient had a permanent pacemaker implanted preoperatively. New postoperative SND occurred in 6 patients and was not related to the presence of persistent left superior vena cava or APVC but was marginally (P=0.07 for each) related to age and presence of NYHA functional class III or IV symptoms.

Sixty patients had predismissal and late ECG assessment. Four patients in whom SND developed before dismissal had late follow-up; SND resolved in 2, atrial flutter developed in 1, and 1 patient received a pacemaker 6 years after surgery. Two patients required early postoperative permanent pacemaker implantation for slow ectopic atrial rhythm, and 3 required late permanent pacemaker implantation. Overall, 6 patients had permanent pacemaker implantation postoperatively.

Twelve patients had atrial fibrillation preoperatively. Of the remaining 103 patients, new-onset atrial fibrillation occurred in 7 patients postoperatively. Univariable predictors for new-onset postoperative atrial fibrillation by Cox regression were older age at repair (P=0.033) and preoperative palpitations (P=0.086).

**Postoperative Echocardiographic Findings**

Postoperative echocardiography was performed at Mayo Clinic in 67 patients; small persistent defects (residual defect <5 mm by echocardiography) were detected in 5 patients (7%). Pulmonary vein and SVC stenoses were not identified by follow-up echocardiography.

**Follow-Up**

Long-term follow-up was possible for 108 (95%) of the 114 early survivors at an average of 144±99 months postopera-
Postoperatively (median 138 months; range 6.8 to 394 months). Postoperative follow-up data and outcomes according to age are shown in Table 2. Improvement in symptoms (ie, decrease in NYHA class or improvement in exercise capacity if preoperative NYHA class was I) occurred in 83 patients (77%) who were symptomatic or in NYHA class I preoperatively and was more common with older age at operation ($P_{H11005}<0.014$), presence of symptoms preoperatively ($P_{H11005}<0.04$), and higher preoperative pulmonary artery pressure ($P_{H11005}<0.01$).

Despite immediate improvement in postoperative symptoms in the majority of patients, 17 (16%) demonstrated symptomatic deterioration during long-term follow-up. Deterioration in functional class occurred in 12 patients with preoperative NYHA class I, 4 patients with preoperative NYHA class II, and 1 patient with NYHA class III. In multivariable analysis, higher preoperative pulmonary artery pressure ($P_{H11005}<0.022$), but not age, was associated with a higher probability of postoperative symptomatic improvement. The incidence of postoperative dyspnea ($P<0.0001$) and atrial fibrillation ($P<0.0001$) also increased with age. Among 14 patients with preoperative chronic or paroxysmal atrial fibrillation, 2 maintained sinus rhythm late after SVASD repair. In 7 other patients (average age at repair 44.7 ± 20.9 years), atrial fibrillation or flutter developed postoperatively. The mean time from repair to the onset of atrial fibrillation was 9.4 ± 9.2 years.

Follow-up ranging from 2 months to 21 years was available for 5 of the 6 patients with preoperative pulmonary vascular resistance >8 U. All of these patients reported improvement in symptoms. Estimated pulmonary artery systolic pressure at follow-up was available for 2 patients (41 and 77 mm Hg).

Sixteen patients died late during follow-up (mean age 69 ± 19 years). The cause of death was unknown for 9 patients. Five patients died of vehicular accident or carcinoma. Two deaths were possibly related to the SVASD. A 65-year-old man with atrial fibrillation died suddenly 41 months postoperatively. A 60-year-old woman with hypertension had sinus bradycardia 12 months postoperatively and died suddenly 9 months later. Both patients had normal coronary arteries preoperatively.

In the forward stepwise multivariable Cox regression analysis, postoperative mortality ($n=17$) was related to older age at repair ($P=0.008$) and preoperative NYHA class III or IV ($P=0.038$). However, survival of these patients after SVASD repair was not significantly different from the expected survival for the US white population either overall or within any of the 3 age strata ($P=0.31$ for overall; 1-sample log-rank test; Figure 2; Table 3).

Among 108 patients with long-term follow-up, reoperation was not required during the follow-up period. Subsequently, a patient who had SVASD repair in 1976 underwent reoperation for bidirectional shunting.

**Discussion**

Patients with SVASD demonstrate unique developmental, anatomic, and surgical features and are at risk for postoperative complications.3–9,12 The decision to repair any kind of ASD is based on clinical and compiled echocardiographic information, including (1) size and location of the ASD, (2)
hemodynamic impact of the left-to-right shunt and associated right-sided cardiac volume overload, and (3) the presence and degree of pulmonary hypertension. Data from the present series of patients included early and late results of operation for SVASD. We found a low rate of perioperative morbidity, mortality, and need for reoperation. In addition, the overall survival was similar to that of a matched population. However, older age at operation and NYHA functional class III or IV symptoms were independent predictors of late mortality. Older age at operation was also the best independent predictor of new-onset postoperative atrial fibrillation. Postoperative symptomatic improvement was noted in the majority of the patients.

Surgical Considerations
In SVASDs, the complex anatomy with APVCs poses a challenge to the surgeon. The results of the present study cannot prove which method of surgical repair is the best. Current methods of surgical repair, which include a trend favoring 2-patch repair and incisions away from the cavoatrial junction at our institution, as well as intraoperative transesophageal echocardiography, may have been successful in eliminating venous pathway stenosis and residual ASDs, and they may affect the incidence of SND.13,14 Late problems may occur from contraction of the pericardial patches, resulting in stenosis of the venous pathways or recurrent ASD, but such problems are rare.

Postoperative Symptoms and Long-Term Follow-Up
Postoperative symptomatic improvement occurred in most (77%) of the patients, especially older patients. Improvement also occurred in patients who were in NYHA class I before surgery. Functional improvement of previously asymptomatic patients has been reported after secundum ASD closure.15 Lack of preoperative symptoms is not a contraindication to the current practice of secundum ASD repair and should not be a contraindication for repair of SVASD, even in adults.

Reports suggest that ASD closure in patients in their 20s is associated with survival similar to age- and sex-matched controls, but closure after age 41 years is associated with a substantial increase in late mortality.16 The present data do not support these findings. Although ASD closure should be considered in select symptomatic patients older than 60 years, in the absence of serious comorbidities or pulmonary hypertension, it is preferable that ASDs be closed as early as possible. Older patients are reported to deteriorate symptomatically without ASD repair, because the age-related decrease in left ventricular compliance augments the left-to-right shunt and because secondary pulmonary hypertension develops.17 The frequency of atrial arrhythmias increases after the fourth decade, which also contributes to functional deterioration.

Postoperative improvement in functional class in patients older than 60 years at the time of ASD repair has been reported from our institution.18 Despite the improvement in symptoms, patients in the present study who had repair after age 40 years demonstrated persistent dyspnea on exertion, possibly due to accelerated late diastolic dysfunction; the dyspnea was more pronounced than in patients younger than 40 years. This finding underscores the importance of early surgical intervention.

Sinus Node Dysfunction
Postoperative SND is more common in patients after SVASD repair than after secundum ASD repair.19 Potential mechanisms for SND in SVASD include anatomic anomaly of the sinus node (eg, from a persistent left SVC),19 intrinsic SND, or surgical trauma caused by proximity of the SVASD to the sinus node, the internodal tracts, and the blood supply to the sinus node. In the present study, only 6 patients had newly documented early postoperative SND independent of an incision across the cavoatrial junction. Because of the small number of patients with SND, it is difficult to determine whether any surgical factors play a role in the development of postoperative SND. Only 5 patients required postoperative pacemaker implantation. The presence of atrial arrhythmias decreases the ability to detect SND; thus, the frequency of SND may be underestimated in the present series of patients.

Atrial Fibrillation or Flutter
Older age at operation was the best independent predictor of new-onset atrial fibrillation during follow-up in the present series of patients. Potential mechanisms for occurrence of atrial fibrillation or flutter in SVASD patients include SND with bradycardia-dependent atrial arrhythmias, scar-dependent multiple reentries, and increased atrial size or atrial fibrosis due to increasing pulmonary venous pressure with exercise.

In some of the patients in the present series, new atrial fibrillation developed during long-term follow-up. Other studies have also demonstrated that late repair of secundum ASD does not impact the development of atrial arrhythmias.20 The significant association of older age and postoperative atrial fibrillation raises the question of whether a maze procedure should be considered routinely in this subgroup.

Study Limitations
A limitation of the present study is its retrospective design. We could not assess the natural history of SVASD and do not have a historical control group for comparison. The natural
history of unoperated SVASD is unknown but is likely similar to that in patients with large ASDs. In addition, postoperative clinical improvement is problematic, because it is difficult to quantify; however, this subjective information is the only method to determine clinical outcome in this retrospective series.

Not all patients had ECG follow-up. Therefore, the frequency of SND or occurrence of long-term atrial arrhythmias cannot be stated with certainty. The presence of atrial arrhythmias decreases the ability to detect SND; thus, the frequency of SND detection may be underestimated. Finally, the cause of death in the majority of patients is unknown; therefore, mortality due to SVASD sequelae may be underestimated.

The decision for operative intervention for SVASD should be individualized. No definite recommendations about upper and lower age limits for surgery can be made from these data; however, operation for SVASD is rarely necessary or advisable in an infant younger than 1 year or in the very elderly.

Recommendations with regard to surgery in asymptomatic patients are hampered by the fact that during long-term follow-up, symptomatic deterioration was noted in 24% of patients who were in functional class I before surgery. We cannot determine whether this was due to surgery or despite surgery or whether this was only a normal deterioration paralleling the long follow-up. In patients who were in functional class I before surgery, death at last follow-up was less likely than in the other patients (P = 0.0005), and there was no significant difference in the occurrence of sinus node dysfunction, atrial fibrillation, or pacemaker implantation.

Conclusions
Despite the complexity of the lesion, repair of SVASD with associated APVC is associated with low morbidity and mortality even in patients older than 40 years. In our experience, severe complications are rare, and development of SND and the need for pacemaker implantation are uncommon. Functional improvement is expected irrespective of age at repair, but postoperative atrial fibrillation appears to be related to older age at operation. SVASD repair achieves survival rates similar to those of a matched population, and although repair is suggested as early as possible, it should be considered whenever repair may impact survival or symptoms.

Disclosure
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References
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