Outcomes of inferior sinus venosus defect repair

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Objective: Inferior sinus venosus defect is an unusual form of interatrial communication with few published data on surgical outcomes. We sought to compare outcomes of surgical repair of inferior sinus venosus defect with those of large secundum atrial septal defects.

Methods: Patients undergoing surgical closure of an isolated interatrial defect were reviewed, and those with inferior sinus venosus defect were identified on the basis of predetermined anatomic criteria. For each case, 2 controls with secundum atrial septal defect, matched for age and year of surgery, were selected. Technical outcome scores and other perioperative outcomes were compared.

Results: Compared with the secundum atrial septal defect group (n = 90), the inferior sinus venosus defect group (n = 45) had worse technical outcome scores ($P = .02$), a higher rate of reintervention (9% vs 1%, $P = .04$), longer median total cardiopulmonary bypass (48 vs 39 minutes, $P < .001$) and crossclamp (29 vs 20 minutes, $P < .001$) times, and were more likely to stay more than 1 day in the intensive care unit (20% vs 8%, $P = .04$) and more than 3 days in the hospital (29% vs 13%, $P = .03$). Only 16 (36%) of the patients with inferior sinus venosus defect had a correct diagnosis preoperatively. Patients with an incorrect diagnosis had worse technical outcome scores than the secundum atrial septal defect group ($P = .003$), whereas those with a correct diagnosis had scores similar to those of the secundum atrial septal defect group ($P = .55$).

Conclusions: Compared with patients with secundum atrial septal defect, patients with inferior sinus venosus defect have more residual defects and longer durations of cardiopulmonary bypass and hospitalization. Rates of misdiagnosis of inferior sinus venosus defect are high and associated with worse technical outcome scores. Accurate preoperative diagnosis of this lesion may lead to improved outcomes. (J Thorac Cardiovasc Surg 2011;142:517-22)

Sinus venosus defect (SVD) is an uncommon form of interatrial communication involving the common wall between the superior vena cava (SVC) and the right upper and middle pulmonary veins (superior sinus venosus defect [SSVD]),[1] or between the right lower pulmonary veins and the right atrium or at the inferior caval-right atrial junction (inferior sinus venosus defect [ISVD]).[1,2] In contrast with secundum atrial septal defect (ASD), which is located within the fossa ovale, SVDs do not typically involve septum primum, the valve of the fossa ovale.[3] SVDs comprise 4% to 11% of isolated interatrial shunts, with the majority of cases being the superior or SVC type.[2] ISVD, also called ''right atrial type SVD,'' is less common and published data regarding outcomes of surgical repair are scarce.[4] On the basis of our clinical impression that ISVD might be confused with secundum ASD and that surgical repair of ISVD might be associated with a higher rate of residual defects, this study was undertaken to compare the frequency of misdiagnosis and surgical outcomes in consecutive patients undergoing surgical repair of ISVD with those of matched patients undergoing surgical repair of large secundum ASD.

MATERIALS AND METHODS

Subjects

A database search identified all patients who underwent surgical repair of an isolated interatrial defect, other than a primum ASD or coronary sino-septal defect, between January 1998 and March 2009. Patients with associated congenital heart disease other than patent ductus arteriosus were excluded. Demographic, imaging, and operative data were abstracted from echocardiogram and surgical reports, and outcome data were ascertained from medical records. Echocardiographic images for patients with defects described as an ISVD or as a posterior and inferior secundum ASD were reviewed by one of the authors (P.B.).

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Abbreviations and Acronyms

ASD = atrial septal defect
ICU = intensive care unit
ISVD = inferior sinus venosus defect
SVC = superior vena cava
SVD = sinus venosus defect
SSVD = superior sinus venosus defect

The study was approved by the Scientific Review Committee of the Department of Cardiology and by the Children's Hospital Boston Committee on Clinical Investigation.

Diagnostic Criteria of Inferior Sinus Venosus Defect

The diagnosis of ISVD was based on the following criteria: (1) posterior or inferior defect confluent with the posterior wall of the atria with no posterior or inferior rim; (2) confluence of the defect with the right pulmonary veins(s) or the inferior vena cava-right atrial junction; and (3) presence of a well-developed septum primum covering the fossa ovale without fenestrations (Figure 1).

Outcomes

Patients who satisfied all 3 criteria comprised the ISVD group. For each patient with ISVD, 2 control patients, matched for age at surgery and time of surgery (within 1 year), undergoing repair of an isolated large (>8 mm) secundum ASD were randomly selected.

The ISVD and secundum ASD groups were compared for differences in perioperative outcomes, including correct preoperative diagnosis, technical outcome score, rate of reinterventions, total cardiopulmonary bypass and crossclamp times, and lengths of intensive care unit (ICU) and hospital stay. A technical outcome score, modified from Larrazabal and colleagues,[5] was assigned to each patient for whom postoperative imaging was available as follows: (1) optimal, no residual shunt; (2) adequate, small residual shunt < 3 mm; (3) inadequate, moderate or greater residual shunt > 3 mm, or requiring surgical or transcatheter reintervention.

Interobserver Reproducibility of Diagnosing Inferior Sinus Venosus Defect

Two observers independently reviewed a randomly selected sample of ISVD and secundum ASD patient echocardiograms and labeled them as ISVD or secundum ASD using the above criteria.

Inferior Sinus Venosus Defect in Conjunction With Other Congenital Heart Diseases

In addition to isolated interatrial defects, a separate database search was conducted for patients with ISVD in conjunction with other forms of congenital heart disease. These patients were not included in the comparison of outcomes between isolated ISVD and secundum ASD.

Statistical Analyses

For descriptive statistics, normally distributed continuous variables are presented as mean ± standard deviation, non-normally distributed continuous variables are presented as median (range), and categoric variables are presented as frequencies. Nominal data (eg, frequency of reintervention) were compared using the Fisher exact test, and non-normally distributed ordinal and continuous data (eg, technical outcome score and cardiopulmonary bypass time) were compared using the Mann–Whitney *U* (Wilcoxon rank-sum) test. Log-rank test was used to compare hospital and ICU length of stay for the 2 groups. The interobserver reproducibility of diagnosis of ISVD was assessed with the Kappa statistic. All statistical tests were 2-sided. All data analysis was performed using PASW Statistics version 17.0.2 (SPSS Inc, Chicago, Ill).

RESULTS

Patient Characteristics

Of 3750 patients followed in our department from January 1998 to March 2009 with isolated interatrial defects (excluding primum atrial septal and coronary sino-septal defects), 700 were referred for surgical closure and 620 were referred for catheter device closure. Of the 700 surgical patients, we identified 45 who had undergone surgical closure of an ISVD and 90 matched controls.

The ISVD and secundum ASD groups were similar in age at surgery (median 2.7 vs 3.1 years, $P = .51$), although they were younger than the overall group of patients with isolated interatrial defect referred for surgical closure during this time period (median 3.0 vs 4.6 years, $P < .001$). There was no difference in percentage of female patients between the ISVD and secundum ASD groups (42% vs 59%, $P = .099$) or in weight at surgery (mean 17.5 vs 18.2 kg, $P = .799$). The 2 groups were also well matched in year of surgery ($P = 1.0$). None of the patients in the ISVD group had undergone an attempted catheter closure of the defect, whereas 4 patients in the secundum ASD group had undergone an unsuccessful attempt at device closure of the defect before surgery.

Outcomes

All patients survived to hospital discharge. Among those with available postoperative imaging (ISVD n = 33, 73%; secundum ASD n = 49, 54%), patients in the ISVD group had worse technical outcome scores ($P = .021$) with a greater percentage of patients receiving an inadequate score (11% vs 2%, $P = .041$) and requiring reintervention (9% vs 1%, $P = .042$) (Figure 2).

The ISVD group had longer total cardiopulmonary bypass (median 48 vs 39 minutes, $P < .001$) and crossclamp (median 29 vs 20 minutes, $P < .001$) times compared with the secundum ASD group (Figure 3). The ICU length of stay was also longer for the ISVD group (median 1 day, range 1–4 vs 1–2 days, $P = .011$), with a greater percentage of the ISVD group spending more than 1 day in the ICU (20% vs 8%, $P = .049$). Similarly, hospital length of stay was longer in the ISVD group (median 3 days, range 2–12 vs 2–5 days, $P = .01$), with a greater percentage of the ISVD group spending more than 3 days in the hospital (29% vs 13%, $P = .036$). Among the ISVD group with postoperative imaging, only 2 patients were noted to have inferior vena cava obstruction, mild in both, and none had pulmonary vein obstruction on their most recent echocardiograms.

Residual Defects and Reinterventions

Of the 45 patients in the ISVD group, 5 (11%) had significant residual defects or reintervention. In contrast, only

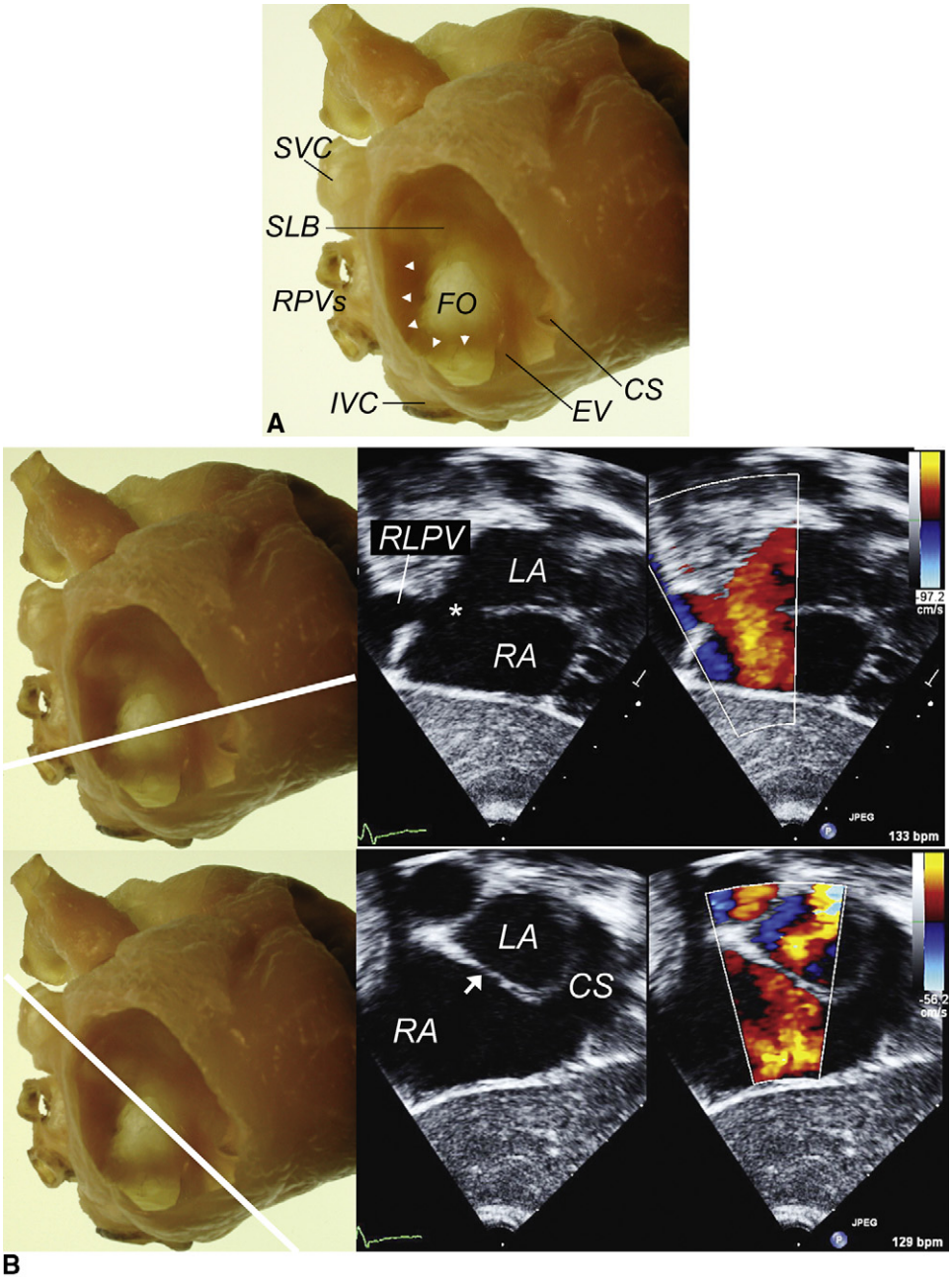

FIGURE 1. Atrial septal anatomy: location and appearance of ISVD. A, Wax model of a normal heart with the sinus venosus septum marked by *white arrowheads*. B, Echocardiographic 2-dimensional and color Doppler images in a subcostal long-axis plane (*top*) show the typical appearance of an ISVD (*star*) and, in a subcostal short-axis plane, (*bottom*) demonstrate an intact fossa ovale and septum primum (*arrow*) in the same patient. *CS,* Coronary sinus; *EV,* eustachian valve; *FO,* fossa ovale; *IVC,* inferior vena cava; *LA,* left atrium; *RLPV,* right lower pulmonary vein; *RPV,* right pulmonary vein; *SLB,* superior limbic band; *SVC,* superior vena cava.

2 of the 90 patients in the secundum ASD group (2%) had residual defects ($P = .041$): One patient had a 4-mm residual patch margin defect on postoperative transesophageal echocardiogram, and one patient had complete closure of the secundum ASD documented on postoperative imaging, but returned later with patch dehiscence requiring reoperation.

In the ISVD group, 3 patients, all with an incorrect diagnosis of secundum ASD, were noted on postoperative echocardiography to have residual defects with inferior vena caval flow into both atria. All 3 patients underwent reoperation for closure of the residual defect. A fourth patient was noted to be cyanotic immediately after surgical closure of a presumed secundum ASD. A postoperative echocardiogram,

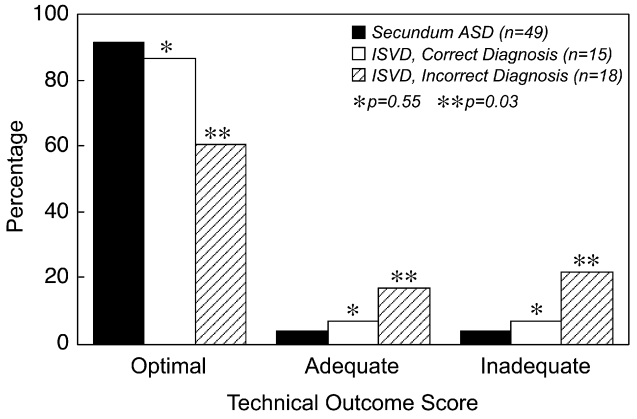

FIGURE 2. Technical outcome score: effect of preoperative diagnosis comparison of technical outcome scores between ISVDs correctly or incorrectly diagnosed before surgery and secundum ASDs. Technical outcome score: optimal = no residual shunt; adequate = small (<3 mm) residual shunt; inadequate = significant (>3 mm) residual shunt or need for reintervention. *ASD,* Atrial septal defect; *ISVD,* inferior sinus venosus defect.

including agitated saline contrast injection through an intravenous cannula in the foot, revealed that the ASD patch had been attached to the eustachian valve inferiorly, with resultant baffling of the inferior vena cava flow to the left atrium. Reoperation was subsequently performed to revise the patch with no significant residual defects. The fifth patient received a correct diagnosis of ISVD on preoperative imaging. No intraoperative imaging was performed, and postoperative transthoracic echocardiography demonstrated a 6-mm residual defect. The patient was discharged from the hospital and has not had subsequent interventions.

Misdiagnosis of Inferior Sinus Venosus Defect

Compared with the secundum ASD group, the ISVD group was more frequently misdiagnosed by both preoperative imaging (64% vs 0%, $P < .001$) and operative assessment (40% vs 1%, $P < .001$), with only 41% of the ISVDs correctly identified by preoperative imaging or direct intraoperative assessment by the surgeon. In the last 2 years, the rate of correct preoperative diagnosis of ISVD improved from 36% (16 patients) to 60% (27 patients).

As seen in Figure 3, patients with a correct diagnosis of ISVD by preoperative imaging (n = 16) had similar technical outcome scores compared with the secundum ASD group ($P = .551$), with similar rates of reintervention (0% vs 1%, $P = .849$). By contrast, patients with an incorrect ISVD diagnosis had worse technical outcome scores compared with the secundum ASD group ($P = .003$), with a higher frequency of reintervention (14% vs 1%, $P = .012$). Of the patients with ISVD, those with a correct diagnosis by direct intraoperative inspection had similar technical scores as those with an incorrect diagnosis ($P = .596$), with a similar frequency of inadequate scores (11% vs 11%, $P = .675$).

Effect of Surgical Incision

The frequency of limited sternotomy (mini-sternotomy) was similar in the ISVD and secundum ASD groups (82% and 89%, respectively, $P = .297$). Although the difference in technical outcome scores between the patients with ISVD who underwent mini-sternotomy and the patients who underwent full sternotomy did not reach statistical significance ($P = .076$), all 7 patients who underwent a full sternotomy had an optimal technical outcome score with no residual defects or reinterventions.

Interobserver Reproducibility of Diagnosing Inferior Sinus Venosus Defect

Two independent observers reviewed 9 ISVD and 14 secundum ASD studies with good agreement of diagnosis (kappa 0.81, $P < .001$). Both observers correctly identified all 14 secundum ASDs. One observer correctly identified all 9 ISVDs, and the other observer correctly identified 7 of the 9 ISVDs.

Inferior Sinus Venosus Defect in Conjunction With Other Congenital Heart Disease

In addition to the 45 patients with isolated ISVD described, we identified 8 patients with ISVD in conjunction with other forms of congenital heart disease: 4 with D-looped transposition of the great arteries, 2 with tetralogy of Fallot, 1 with L-looped transposition of the great arteries, and 1 with a membranous ventricular septal defect (Table 1). Only 2 of the patients had a correct diagnosis of ISVD at the time of their preoperative imaging, and both of these patients underwent surgical closure of the ISVD at their initial operation with no residual defects. The remaining 6 patients did not have an ISVD diagnosis before their initial surgical repair and were later found to have significant residual interatrial defects consistent with ISVD. All 6 patients required reoperation for ISVD repair.

DISCUSSION

Unlike published data on SSVDs,[1,2,6-13] published data regarding the diagnosis and treatment of ISVDs are scarce and consist mostly of anecdotal reports.[2,6,13-15] In this single institution study of the largest series of patients with ISVD to date, only one third of ISVDs were diagnosed correctly before surgical repair. Compared with patients undergoing secundum ASD repair, they had worse technical outcomes, longer cardiopulmonary bypass and crossclamp times, and longer ICU and hospital length of stay. Correct preoperative diagnosis of ISVD was associated with better surgical outcomes, comparable to those of secundum ASD.

Previously, the largest published series of ISVD consisted of 11 patients and found similarly poor diagnostic accuracy by echocardiography, which improved over time.[4] Although this series reported good surgical outcomes with no reoperations or reinterventions, the number of patients

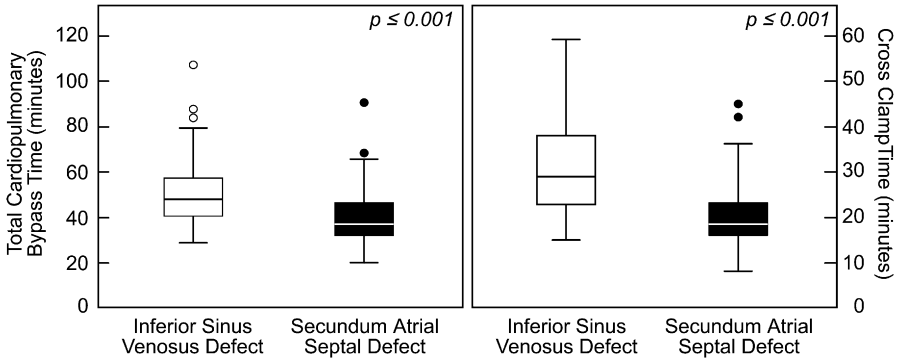

FIGURE 3. Surgical repair: total cardiopulmonary bypass and crossclamp times. Comparison of total cardiopulmonary bypass and crossclamp times during surgical repair of ISVDs versus secundum ASDs.

was small and it is unclear from the data how many patients had residual defects. Furthermore, the study relied on echocardiographic reports and surgical notes without review of the primary images. Given the poor diagnostic recognition of ISVDs by echocardiography and at surgery, this may have underestimated the number of ISVDs and confounded the assessment of complication rates.

Even with a high index of suspicion and clearly defined diagnostic criteria, there was imperfect agreement between our independent observers on the diagnosis of ISVD, highlighting the challenges in distinguishing ISVD from secundum ASD. Diagnostic accuracy has important implications for the perioperative management of these patients, and it is because of the difficulties distinguishing ISVD from secundum ASD that we chose the latter group as controls.

Proper identification of patients with ISVD before surgery allows adequate operative planning and discussion with the family. We speculate that cannulation of the inferior vena cava at the usual site, that is, at the junction between the right atrial free wall and the inferior vena cava, followed by placement of a caval occluding tape may have contributed to poor visualization of its proper inferior border. With correct preoperative diagnosis of ISVD, the inferior vena cava can be cannulated more caudally, thereby further improving visualization of the proper inferior border of the defect (Figure 4). The surgeon may also consider the inferior extent of the sternotomy incision to optimize exposure of the defect and neighboring structures. In this study, none of the patients with a full median sternotomy had any residual defect or need for reintervention, possibly because of better visualization of the defect's margins. Furthermore, intraoperative imaging can assist with both operative planning and allow for detection and correction of residual defects at the time of initial operation, as can injection of agitated saline contrast via intravenous cannulation in a lower extremity.

TABLE 1. Inferior sinus venosus defects in association with other forms of congenital heart disease

Patient	Associated CHD	Preoperative diagnosis of ISVD	ASD closure at first surgery	Reoperation for ISVD
1	Membranous VSD	Yes	Yes	No
2	Tetralogy of Fallot	No	No	Yes
3	Tetralogy of Fallot	No	Yes	Yes
4	L-looped TGA	Yes	Yes	No
5	D-looped TGA	No	Yes	Yes
6	D-looped TGA	No	Yes	Yes
7	D-looped TGA	No	Yes	Yes
8	D-looped TGA	No	Unknown*	Yes

ASD, Atrial septal defect; *CHD,* congenital heart disease; *ISVD,* inferior sinus venosus defect; *VSD,* ventricular septal defect; *TGA,* transposition of the great arteries. *Initial care for this patient was at another institution, and details of the first surgery are unknown.

In this study, patients with an incorrect diagnosis of ISVD before surgery had worse technical outcomes and greater frequency of reinterventions than patients with secundum ASD. Conversely, those who were correctly identified before surgery had technical outcomes similar to those of the secundum ASD group, and none required reintervention. Notably, only 1 of the patients with a correct diagnosis had a significant residual shunt, and no intraoperative imaging was performed in this patient. Furthermore, among the 8 patients with ISVD in the setting of other cardiac defects, 6 required reoperation for the ISVD, none of whom had a correct diagnosis before their initial surgical repair. The 2 patients with a correct diagnosis preoperatively had no significant residual defects.

Greater awareness of the distinguishing features of ISVD from those of secundum ASD may lead to improved preoperative diagnostic accuracy. In our practice, this is likely the explanation for the improved rate of detection in the last 2 years. Furthermore, these data suggest that with accurate ISVD diagnosis and careful preoperative planning, the incidence of residual defects can be reduced. Careful examination of the posterior-inferior portion of the interatrial septum should be a part of every study in patients with isolated interatrial shunts and in patients with concomitant congenital heart defects.

Even with appropriate preoperative identification and planning, surgical repair of ISVD may be more challenging than

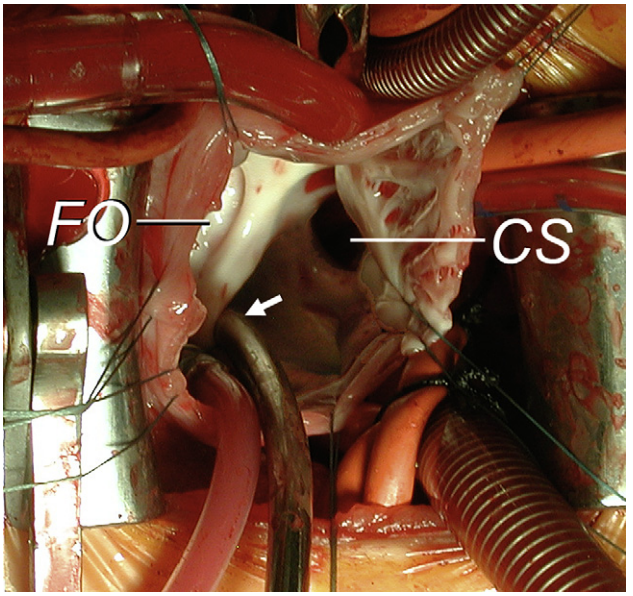

FIGURE 4. Surgical repair of ISVDs. Intraoperative photograph of an ISVD showing an intact fossa ovale with a suction catheter through the defect (*arrow*). The inferior vena cava cannula is placed caudally to allow complete visualization of the defect margins. *CS,* Coronary sinus; *FO,* fossa ovale.

secundum ASD. In our cohort, the patients with ISVD were younger than the overall group with isolated interatrial defects referred for surgical closure, possibly reflecting a greater degree of shunting. They also had longer bypass times and length of stay in the ICU and hospital than the age-matched controls, suggesting that the surgical repair may be more complex in this population. These data should be considered when counseling patients before surgical repair of ISVDs.

Limitations

Because of variable practices surrounding perioperative and postoperative imaging, follow-up imaging data were not available for all patients. Thus, the true incidence of residual shunts in both groups is not known. However, clinical evaluations in these patients were not suggestive of important residual shunt.

CONCLUSIONS

ISVD is a diagnosis that carries greater perioperative and postoperative morbidity than previously recognized. Compared with patients with secundum ASD, patients with ISVD have more residual defects, a higher rate of reinterventions, and longer durations of cardiopulmonary bypass and hospitalization. Rates of misdiagnosis of ISVD are high, even in the current era, and incorrect diagnosis is associated with worse technical outcomes. Given that correct preoperative diagnosis is associated with outcome comparable to those undergoing surgical repair of isolated secundum ASD, heightened diagnostic and intraoperative awareness of this defect are needed.

References

1. Van Praagh S, Carrera ME, Sanders SP, Mayer JE, Van Praagh R. Sinus venosus defects: unroofing of the right pulmonary veins—anatomic and echocardiographic findings and surgical treatment. *Am Heart J*. 1994;128:365-79.
2. Jost CHA, Connolly HM, Danielson GK, Bailey KR, Schaff HV, Shen W-K, et al. Sinus venosus atrial septal defect: long-term postoperative outcome for 115 patients. *Circulation*. 2005;112:1953-8.
3. Geva T. Anomalies of the atrial septum. In: Lai WW, Mertens LL, Cohen MS, Geva T, eds. Echocardiography in Pediatric and Congenital Heart Disease. Oxford, UK: Wiley-Blackwell; 2009:158-74.
4. Crystal MA, Al Najashi K, Williams WG, Redington AN, Anderson RH. Inferior sinus venosus defect: echocardiographic diagnosis and surgical approach. *J Thorac Cardiovasc Surg*. 2009;137:1349-55.
5. Larrazabal LA, del Nido PJ, Jenkins KJ, Gauvreau K, Lacro R, Colan SD, et al. Measurement of technical performance in congenital heart surgery: a pilot study. *Ann Thorac Surg*. 2007;83:179-84.
6. Davia JE, Cheitlin MD, Bedynek JL. Sinus venosus atrial septal defect: analysis of fifty cases. *Am Heart J*. 1973;85:177-85.
7. Ettedgui JA, Siewers RD, Anderson RH, Park SC, Pahl E, Zuberbuhler JR. Diagnostic echocardiographic features of the sinus venosus defect. *Br Heart J*. 1990;64:329-31.
8. Muhler EG, Engelhardt W, Von Bernuth G. Detection of sinus venosus atrial septal defect by two-dimensional echocardiography. *Eur Heart J*. 1992;13:453-6.
9. Sharma S, Krishnan U. Diagnosis of the sinus venosus defect by echocardiography and angiocardiography. *Indian Heart J*. 1992;44:395-8.
10. Agrawal SK, Khanna SK, Tampe D. Sinus venosus atrial septal defects: surgical follow-up. *Eur J Cardiothorac Surg*. 1997;11:455-7.
11. Russell JL, LeBlanc JG, Deagle ML, Potts JE. Outcome following repair of sinus venosus atrial septal defects in children. *Asian Cardiovasc Thorac Ann*. 2002;10:231-4.
12. Iyer AP, Somanrema K, Pathak S, Manjunath PY, Pradhan S, Krishnan S. Comparative study of single- and double-patch techniques for sinus venosus atrial septal defect with partial anomalous pulmonary venous connection. *J Thorac Cardiovasc Surg*. 2007;133:656-9.
13. Valente A, Sena L, Powell A, Del Nido P, Geva T. Cardiac magnetic resonance imaging evaluation of sinus venosus defects. *Pediatr Cardiol*. 2007;28:51-6.
14. Pai RG, Pai SM, Ortega V, Lopez A. A rare case of inferior venacaval type of atrial septal defect in an adult: Echocardiographic features. *J Am Soc Echocardiogr.* 1996;9:819-21.
15. Chen CA, Wang JK, Hsu JY, Hsu HH, Chen SJ, Wu MH. Diagnosis of inferior sinus venosus atrial septal defects using transthoracic three-dimensional echocardiography. *J Am Soc Echocardiogr.* 2010;23:457:e4-6.