A Poorly Prognostic Case of Renal Dialysis in a 20-Year Case Series of Adult Coronary Sinus Atrial Septal Defect Repair at New Tokyo Hospital

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Keywords
Coronary sinus atrial septal defect • Unroofed coronary sinus syndrome • Poor prognosis • Renal dialysis • Adult congenital heart disease
Abstract

**Background:** Isolated coronary sinus atrial septal defect (ASD) is defined as a coronary sinus unroofed in the terminal portion without a persistent left superior vena cava and other anomalies. This defect is rare and part of a wide spectrum of unroofed coronary sinus syndromes. Recently, several reports have described this finding. We searched the hospital’s database to determine the incidence of this defect, and to raise awareness of this condition, we discussed the findings from five patients with coronary sinus ASD who underwent surgical repair.

**Case presentation:** The patients were three women and two men with an age range of 63–77 years. All patients underwent transthoracic echocardiography and computed tomography, and one underwent magnetic resonance imaging. In two patients, the defect was found unexpectedly intraoperatively; left-to-right shunting was apparent in the other three patients preoperatively. The pulmonary-to-systemic blood flow ratio ranged from 1.42 to 3.1 following cardiac catheterization, and oxygen saturation step-up was seen on the right side of the heart. Valvular regurgitation was seen in 4/5 patients with the mitral, tricuspid, and aortic valves involved in different combinations and to different degrees. Right atrial and ventricular dilation were seen in 4/5 patients; three patients had left atrial dilation. Three patients experienced atrial fibrillation, and one of these also experienced paroxysmal ventricular contractions. All patients underwent surgical repair, and some underwent multiple procedures. One patient who had previously undergone kidney transplantation died approximately 1 year postoperatively; the remaining four patients are currently experiencing good activities of daily living without symptoms.

**Conclusions:** Coronary sinus ASD (Kirklin and Barratt–Boyes type IV unroofed coronary sinus syndrome) comprised 1.3% of adult congenital heart surgeries and 0.07% of adult open-heart surgeries in our hospital from 1999 to 2019. Our hospital performs cardiac surgery mainly for patients with acquired cardiac disease, and coronary sinus ASD is rare. Early diagnosis and, in symptomatic patients (especially those with blood access shunts, which may overload the heart), early surgical repair are important. The poorly prognostic case in our series is noteworthy, as similar cases have not been reported previously.
Background

Unroofed coronary sinus syndrome is a rare congenital heart disease.\textsuperscript{1, 2} Coronary sinuses (CSs) unroofed in the terminal portion (Kirklin and Barratt-Boyes type IV)\textsuperscript{3} without a persistent left superior vena cava (PLSVC) and other anomalies are classified as a type of atrial septal defect (ASD) ("isolated coronary sinus ASD"), which comprises less than 1\% of all ASDs in the literature.\textsuperscript{3-5} In general, the prognosis after repair is considered to have a favorable and uneventful course. Although some surgeons specializing in congenital heart disease may have the impression that coronary sinus ASD is not a concern, especially in hospitals performing surgery mainly for adult patients with acquired heart diseases, coronary sinus ASD is very rare.

Several recent reports have discussed this anomaly to raise awareness of its existence.\textsuperscript{6-9} When we identify CS enlargement, we must rule out not only PLSVC but also other anomalies. Otherwise, it can be difficult to reach the true diagnosis. Especially in isolated coronary sinus ASD, there are often almost no symptoms, and transthoracic echocardiography (TTE) can miss the defect,\textsuperscript{10, 11} although right-to-left shunting due to PLSVC or other more complex congenital anomalies can result in cyanosis, hypoxia, transient ischemic attacks, paradoxical embolism, and cerebral abscess. Current imaging modalities such as transesophageal echocardiography (TEE), magnetic resonance imaging (MRI), or three-dimensional computed tomography (CT) are quite helpful and can clearly reveal this anomaly from various view angles.\textsuperscript{4, 12-14}

At New Tokyo Hospital, we began scheduling operations for repairing this congenital anomaly in three adult patients in 2016. Unfortunately, one of the patients died approximately 1 year after the operation. Reviewing past reports, we could not find a patient with a poor prognosis after repair of the same anomalous type. Therefore, we conducted this investigation of our medical database and discussed our experience with the repair of coronary sinus ASD with a retrospective review of our hospital’s surgical records from the past 20 years, which confirmed the existence of and survival outcomes associated with this anomaly. Reviewing the surgeries performed for congenital heart
diseases in our hospital, where mainly surgeries for adult acquired heart diseases are performed, we consider it valuable to discuss the epidemiology of and clinically important points related to this anomaly.

**Case presentation**

All included patients underwent surgical repair of a coronary sinus ASD between January 1999 and December 2019. When we reviewed our hospital’s surgical records from the past 20 years, we identified five cases (as described below chronologically) and confirmed the survival of patients with coronary sinus ASD. Background and preoperative laboratory data are shown in Table 1 and Table 2, respectively.

We also investigated surgical cases of adult congenital heart disease and the number of open-heart surgeries. We included off-pump coronary artery bypass grafting cases as open-heart surgeries and excluded patent foramen ovale as a congenital heart disease from our investigation (Supplemental Table 1).

This research was approved by our hospital’s ethics committee and the president of the hospital. The ethics committee waived the need to obtain consent to use patients’ data except for that of the five cases in this retrospective study.

**Case 1**

A 63-year-old man experienced paroxysmal atrial fibrillation (AF) from the age of 33 years. Ostium secundum ASD was identified at age 42 years at another hospital, and the ASD was monitored over time because of his asymptomatic status. He had been followed for 11 years at another hospital, and when chest X-ray (CXR) showed cardiomegaly with a cardiothoracic ratio (CTR) of 70%, he was referred to our hospital. TTE showed dilation of the right atrium (RA), right ventricle (RV), and left atrium (LA); an ASD measuring 30 mm near the atrioventricular valves; and
left-to-right intracardiac shunting. The pulmonary-to-systemic blood flow ratio (Qp/Qs) was 4.2, but this was calculated as a reference regarding AF. Moreover, TTE showed moderate mitral regurgitation (MR) due to prolapse of the anterior mitral leaflet and severe tricuspid regurgitation (TR) with a TR pressure gradient of 54 mmHg (Table 3). Cardiac catheterization revealed a Qp/Qs of 3.1, a pulmonary-to-systemic pressure ratio (Pp/Ps) of 0.5, and no significant stenosis in the coronary arteries (Table 4). The transcatheter pressure study and blood oxygen saturation study results are summarized in Table 4 and Table 5, respectively. We observed oxygen saturation step-up on the right side of the heart. He was admitted for surgery on 2 November 2005.

Case 2

A 71-year-old woman was followed up regularly by one of our cardiologists to monitor AF. She underwent radiofrequency catheter ablation (RFCA) for frequent paroxysmal ventricular contractions (PVCs) focused on the right ventricular outflow tract (RVOT) in April 2011. Subsequently, she developed dyspnea with New York Heart Association (NYHA) class II heart disease, and she was referred to our hospital for progression of aortic regurgitation (AR) in October 2012. Her electrocardiography (ECG) showed AF but a stable heart rate of approximately 70 bpm. CXR showed cardiomegaly, with a CTR of 60%, and TTE showed moderate AR and mild-to-moderate MR, and dilation of the RA and LA. Although the TTE findings indicated CS dilation measuring 23 mm, no intracardiac shunt was identified preoperatively (Table 3). She was admitted for surgery on 5 January 2013.

Case 3

A 77-year-old woman experienced discomfort during calm physical exercise (yoga) for 2 years. Subsequently, she experienced episodes of presyncope, and she was referred to her local cardiologist. TTE and CT detected an unroofed coronary sinus. She was referred to our hospital to confirm indications for surgical repair. CXR images showed no significant signs of heart failure,
such as cardiomegaly, pulmonary congestion, or pulmonary effusion. Her heart rate was stable, with normal sinus rhythm. TTE showed a large defect between the left atrium and the CS, measuring 16 mm in width. Qp/Qs was estimated at 2.1, and severe TR was seen. The RA and RV were relatively dilated (Table 3); however, there was no significant stenosis in her coronary arteries on CT. She was admitted for surgery on 14 March 2016.

Case 4

A 65-year-old man was referred to our hospital with NYHA class II dyspnea. CXR showed an almost normal cardiac size, with a 51% CTR and no pulmonary congestion or pulmonary effusion. His heart rate was approximately 70 bpm with sinus rhythm during ECG. TTE showed a dilated CS orifice measuring 24 mm × 18 mm, and shunt flow was visible at almost the same point, which suggested blood flow from the LA through the unroofed CS to the RA. Qp/Qs was 2.0, and the RA and RV were relatively dilated (Table 3). Cardiac catheterization resulted in a Qp/Qs of 1.42 and a Pp/Ps of 0.10, with no significant coronary stenosis (Table 4). He was admitted for surgery on 23 July 2016.

Case 5

A 74-year-old woman was referred to our hospital with complaints of palpitation and dyspnea. Renal dialysis was started in her 40s because of chronic glomerulonephritis, and at 46 years of age, she underwent renal transplantation from a deceased donor. However, she subsequently required constant renal dialysis. After the transplantation, she began taking cyclosporine and was followed up medically after the age of 63 years to monitor pancytopenia. On admission for heart surgery, she was taking 100 mg of cyclosporine once per day, and fortunately, her blood cell counts were in the normal ranges (Table 2). CXR showed cardiomegaly, with a CTR of 59% and pulmonary congestion but no pulmonary effusion. Her cardiac rhythm showed AF, and her heart rate was approximately 70 bpm on ECG. TTE showed a dilated CS orifice measuring 28 mm × 27 mm and a
13-mm wide defect on the roof of the CS. Qp/Qs was estimated at 2.7, and moderate regurgitation in both the mitral and tricuspid valves was detected. The RA, RV, and LA were relatively dilated, and the left ventricle was mildly hypertrophic, but with a normal ejection fraction and mild-to-moderate MR. Cardiac catheterization revealed a Qp/Qs of 1.82 and a Pp/Ps of 0.31, with no significant coronary stenosis (Table 4). She was admitted for surgery on 1 October 2016.

TEE (iE33; Philips Medical Systems, Bothell, WA, USA) clearly showed a defect in the roof of the CS (Figure 1a for case 5), with clear left-to-right shunting dynamically through the defect (Figure 1b and c for case 3). MRI (Achieva 3.0T; Philips Inc., Best, The Netherlands) and multidetector-row CT (SOMATOM Definition AS+; Siemens Medical Systems, Forchheim, Germany) confirmed this congenital disease (Figure 2 for case 4 and Figure 3 for case 5, respectively). We identified no PLSVC or other congenital cardiovascular anomalies.

**Surgical procedure**

On 7 November 2005, the patient in case 1 underwent secundum ASD patch closure with autologous pericardium, mitral annuloplasty (MAP), tricuspid annuloplasty (TAP), a biatrial maze procedure, and RA plication due to secundum ASD, moderate MR, severe TR, and chronic AF. Cardiopulmonary bypass was commenced by ascending aortic cannulation near the aortic arch and both superior vena cava (SVC) and inferior vena cava (IVC) cannulation. We performed aortic cross-clamping (AXC), and antegrade cardioplegia (CP) was infused through the aortic root. Under cardiac arrest, we found that the secundum ASD measured 30 mm × 40 mm, and we identified an unroofed CS to the LA. An LA vent tube was inserted from the right upper pulmonary vein (RUPV), and a retrograde CP cannula was inserted from the CS directly. After opening the right side of the LA, we observed the mitral valve. We found dilation of the mitral annulus, mild myxomatous degeneration on the leaflets, and a relatively normal undervalvular apparatus. We performed maze cryoablation, MAP with a 30-mm ring, and TAP with a 36-mm band. Finally, we
closed the secundum ASD with an autologous pericardial patch using 5-0 polypropylene (ppp) running sutures. The coronary sinus ASD was also closed by direct suture or an autologous pericardial patch from the left atrial side.

On 15 January 2013, the patient in case 2 underwent aortic valve replacement with a 25-mm bioprosthesis, mitral valve repair (plasty) (MVP) with anterior commissurotomy, anterior mitral leaflet thinning by peeling off the thickened intima, MAP with a 28-mm ring, TAP with a 30-mm band, the biatrial maze procedure, and RA plication due to moderate AR, mild-to-moderate rheumatic MR, mild TR, and chronic AF. Cardiopulmonary bypass was established by ascending aortic cannulation and both SVC and IVC cannulation. After the LA vent tube was inserted from the RUPV, we performed AXC, and antegrade CP was infused. Under cardiac arrest, after opening the RA, we found a 10-mm communication between the LA and CS when the retrograde CP cannula was inserted from the CS directly. We diagnosed coronary sinus ASD, which we closed with direct suturing from the left atrial side through the opening of the right side of the LA.

In 2016, operations for repairing coronary sinus ASD diagnosed preoperatively were performed for case 3 on 16th March, case 4 on 25th July, and case 5 on 4th October, described as follows.

Under general anesthesia and with a median sternotomy, cardiopulmonary bypass was performed between the ascending aorta and both vena cavae. Myocardial protection was achieved with cold-blood CP through the aortic root. After AXC and cardioplegic arrest, the RA was opened from the right atrial appendage toward the IVC, obliquely and transversely, across the crista terminalis. With a trans-interatrial septal approach, the septum was retracted to clearly observe the mitral valve, and we identified the CS defect from the left atrial side. The defect was observed near the mitral posterior commissure (Figure 4). The upper limbus was fibrous tissue, and the lower limbus was the muscular line, as seen under guidance from the retrograde cardioplegia cannula in the CS. The defect was closed using a fresh autologous pericardial patch (3 cm × 2.5 cm) with 5-0
ppp continuous suture. To open the coronary sinus, we placed a Nelaton tube in the CS when we sutured the deep side of the patch edge.

For case 3, TAP with a 26-mm band was added due to moderate to severe TR. For case 5, we performed MAP with a 28-mm ring, valvuloplasty on both the P1–P2 and P2–P3 indentations, TAP with a 30-mm band, and a left atrial maze procedure due to mild to moderate MR, moderate TR, and chronic AF.

**Outcomes**

For cases 3 and 4, after aortic declamping, TEE showed a residual shunt at the patch intraoperatively. Therefore, we immediately performed secondary AXC and reversed the cardioplegic arrest, observed the patch site closure carefully and found leakage near the mitral posterior commissure. At the site, a small fold was present in the running suture line, which we corrected with additional sutures, closing the leak.

The AXC times were 146 min (99 min for the first procedure and 47 min for the second procedure to close the residual shunt) in case 3, 143 min (95 min for the first procedure and 48 min for the second procedure to close the residual shunt) in case 4, 114 min in case 1, 223 min in case 2, and 147 min in case 5 (Table 6).

These patients had uneventful postoperative courses and were discharged approximately 2–4 weeks after the operations (Table 6). All patients except for the patient in case 5 are currently experiencing very good activities of daily living without symptoms. Although the patient in case 1 has been on renal dialysis due to renal sclerosis since April 2016, i.e., 5 months and 10 years after the operation, he has been living in a very good condition for 15 years after the repair. Unfortunately, the patient in case 5 developed sustained chronic AF, anemia, hypoproteinemia, progression of disuse syndrome from cervical spondylotic myelopathy, and difficulties during renal dialysis due to hypotension despite good ventricular contraction. The patient died approximately 1 year after the operation at another hospital.
From January 1999 to December 2019, we identified five patients with coronary sinus ASD: one in 2005, one in 2013 and three in 2016. In the past 20 years, 371 adult congenital cardiac surgeries (median age: 61 years, range: 13–88 years; males: 210; females: 161) and 7314 open-heart surgeries were performed (Supplemental Table 1). Coronary sinus ASD comprised 1.3% of adult congenital heart surgeries and 0.07% of adult open-heart surgeries (Supplemental Table 1). For adult congenital surgeries, those for acquired cardiovascular diseases, such as valvular surgeries, coronary artery bypass grafting, and aortic surgeries, are usually performed concomitantly.

Discussion and conclusions

We found five cases of coronary sinus ASD repaired at New Tokyo Hospital from 1999 to 2019. In the last 20 years at the hospital, where cardiac surgeons perform operations mainly for adult acquired heart diseases, coronary sinus ASD comprised 0.07% of open-heart surgeries, so we consider this condition to be a very rare anomaly at our hospital. These cases were also the simplest type of unroofed coronary sinus syndrome; therefore, in general, the prognosis after repair is considered to have a favorable and uneventful course. Unfortunately, the patient in case 5 died approximately 1 year after the operation. Reviewing past reports, we could not find a patient with a poor prognosis after coronary sinus ASD repair (Kirklin and Barratt-Boyes type IV unroofed coronary sinus syndrome) without other complex congenital anomalies, i.e., case 5 provided us with novel insights. In case 5, volume overload to the cardiac chambers resulted in AF, annular dilatation of the atroventricular valves, and atrial functional TR/MR. Furthermore, a blood access shunt for renal dialysis overloaded the patient’s heart. Although renal failure and disuse syndrome caused systemic failure, leading directly to the patient’s death, surgical intervention before the progression of arrhythmia and/or valvular dysfunction is still recommended. Therefore, it is very important to make a true diagnosis as early as possible.

Optimal timing for surgical intervention is also very relevant. The indications for the operation in case 4 may be controversial because the Qp/Qs ratio was 1.42 according to the catheter analysis,
while a Qp/Qs ratio of 2.0 was determined using echocardiography. Some may suggest that it was too early to decide to perform surgical intervention in case 4. However, we opted to close the isolated coronary sinus ASD because of dyspnea and dilation of the RA and RV. As noted above, in case 5, the blood access shunt for renal dialysis had been overloading the patient’s heart before the operation for 34 years, which may have been an additional and fatal load for case 5 specifically. In contrast to case 5, the patient in case 1 lived in very good health for 15 years after the operation, even with renal dialysis, which was introduced due to renal sclerosis 10 years after the repair.

It is often difficult to diagnose coronary sinus ASD. Notably, the patients in cases 1 and 2 had no preoperative diagnosis of coronary sinus ASD. In case 1, because of the large secundum ASD, the echocardiographer might have disregarded other congenital anomalies. Intraoperatively, the initial CP was infused anterograde from the aortic root, and cannulation for retrograde CP was performed from the CS ostium directly, so blind cannulation injury on the CS roof was impossible. In case 2, preoperative TTE detected CS enlargement but did not identify the defect on the CS roof or the intracardiac shunt. Retrospectively, preoperative CT scans were performed only in the axial view, so the defect could have been overlooked. The patient in case 2 previously underwent RFCA to treat frequent PVCs associated with the RVOT; the catheter was not inserted into the CS, so it was impossible that the catheter damaged the roof of the CS iatrogenically. TEE, MRI, and three-dimensional CT are quite helpful for diagnosing both coronary sinus ASD and other concomitant congenital anomalies.4, 12-14

In this case series, we found no PLSVC and closed the CS roof defect from the left atrial side through a right-side LA approach in 2005 and 2013 and a trans-interatrial septal approach in 2016. Unroofed coronary sinus syndrome is a rare congenital heart disease,1-2 and the syndrome also has a wide spectrum of anomalies in the literature,5 from total to partial defects of the CS roof with or without PLSVC and with or without other congenital anomalies. Quaegebeur J and colleagues reported that 75% of 24 unroofed CS patients treated over 10 years had PLSVC, which returns venous blood correctly into the RA with the roofing technique or baffle rerouting technique; in
Quaegebeur J and colleagues study, these defects were corrected in the LA but can also be treated with simple ligation following pressure monitoring, as shown in Ootaki Y and coworkers’ report. Through the left atrial approach, we can confirm the heart morphology, especially the LA interior, to identify partial or total roof defects and examine the PLSVC orifice. Readers may question whether two of the three cases in 2016 required additional aortic cross-clamping to close the residual shunt intraoperatively. The defects were not small, as shown in Figure 1a and Figure 3, and the surgical view was also spatially limited, as shown in Figure 4. Even with a very experienced adult cardiac surgery team, it is important to obtain a sufficient surgical view and appropriate suture technique to avoid stenosis of the CS and residual shunting, especially in atypical surgical cases, such as with this anomaly.

We performed the repairs from the left atrial side through standard and orthodox approaches, although simple closure of the CS orifice is also indicated for unroofed CS without PLSVC, with permissible right-to-left shunts. Some innovative reports are described as follows. Takahashi and colleagues reported an anatomical correction of an isolated coronary sinus ASD from the right atrial side and closure of the defect through the CS orifice. In addition, Bozso SJ and coworkers reported that through a periareolar approach, an isolated coronary sinus ASD was closed successfully with minimally invasive endoscopic repair. Current state-of-the-art endovascular procedures can be indicated for various intracardiac structural diseases. An isolated coronary sinus ASD was also closed successfully with a percutaneous device closure technique by Sandeep N et al. However, the safety and efficacy of these approaches have not yet been established because of limited clinical experience. Therefore, open-heart surgery is often still essential in repairing coronary sinus ASD as well as ostium secundum ASD without a rigid limbus, sinus venosus ASD, or ostium primum ASD.

In conclusion, coronary sinus ASD was very rare at New Tokyo Hospital from 1999 to 2019. We found five adult patients (Kirklin and Barratt-Boyes type IV unroofed coronary sinus syndrome) whose ASDs were surgically repaired in the past 20 years. One of the patients died approximately 1
year after the operation. Reviewing past reports, we could not find a patient with a poor prognosis after repair of the same anomalous type. Blood access shunts may overload the heart, especially in patients receiving renal dialysis, and we recommend not hesitating in performing early surgical repair.

**List of abbreviations**

AF, atrial fibrillation  
AR, aortic regurgitation  
ASD, atrial septal defect  
AXC, aortic cross-clamping  
BA V, bicuspid aortic valve  
CoA, coarctation of the aorta  
CP, cardioplegia  
CPAF, coronary artery-pulmonary artery fistula  
CS, coronary sinus  
CT, computed tomography  
CTR, cardiothoracic ratio  
CXR, chest X-ray  
ECG, electrocardiography  
IVC, inferior vena cava  
LA, left atrium  
MAP, mitral annuloplasty  
MR, mitral regurgitation  
MRI, magnetic resonance imaging  
MVP, mitral valve repair  
NYHA, New York Heart Association
PA, pulmonary artery

PAPVR, partial anomalous pulmonary venous return

PDA, patent ductus arteriosus

PLSVC, persistent left superior vena cava

ppp, polypropylene

Pp/Ps, pulmonary-to-systemic pressure ratio

PVC, paroxysmal ventricular contraction

QAV, quadricuspid aortic valve

Qp/Qs, pulmonary-to-systemic blood flow ratio

RA, right atrium

RFCA, radiofrequency catheter ablation

RUPV, right upper pulmonary vein

RV, right ventricle

RVOT, right ventricular outflow tract

SVC, superior vena cava

TAP, tricuspid annuloplasty

TEE, transesophageal echocardiography

TR, tricuspid regurgitation

TTE, transthoracic echocardiography

URCS, unroofed coronary sinus syndrome

VSD, ventricular septal defect

Declarations

Ethics approval and consent to participate

This research was approved by the New Tokyo Hospital Research Ethics Committee and the president of the hospital (committee reference number 0188). The ethics committee waived the
need to obtain consent to use patient data except for the five patients (cases 1-5) described in this retrospective study.

**Consent for publication**

Written informed consent was obtained from the patients (case 1, case 2, case 4, and case 5) or the patient’s family (case 3) for publication of this case report and any accompanying images. A copy of the written consent form is available for review by the Editor of this journal.

**Availability of data and materials**

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

**Competing interests**

The authors declare that they have no competing interests.

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**Authors’ contributions**

HS was a major contributor in writing the manuscript and drafting the tables and figures and was an assistant surgeon. TN is the head of the division of cardiovascular surgery in the hospital and was the chief surgeon for these procedures. YI was an assistant surgeon for these operations. HS, YI, DI, ST, and NK were engaged in preoperative and postoperative management and the surgical operations. SN is the president of the hospital and gave final approval for this case report and research work. All authors read and approved the final manuscript.

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Figure legends

**Figure 1** Echocardiographic findings in cases 3 and 5

a. Three-dimensional transesophageal echocardiography in case 5 clearly showing a defect (white arrow) near the posterolateral commissure of the mitral valve (MV). b, c. Dynamic left-to-right shunting is visible through the defect in case 3.

CS coronary sinus, LA left atrium, MV mitral valve, RA right atrium

**Figure 2** Magnetic resonance imaging in case 4

Magnetic resonance imaging (sagittal view) in case 4 showing the morphology of the patient’s congenital heart disease. We found no persistent left superior vena cava or other congenital cardiovascular anomalies. The arrow indicates the coronary sinus defect.

Ao aorta, CS coronary sinus, LA left atrium, PA pulmonary artery, RA right atrium

**Figure 3** Sixty-four-row multidetector computed tomography scan in case 5

Sixty-four-row multidetector computed tomography scan in case 5 showing the morphology of the patient’s congenital heart disease. The arrow indicates the coronary sinus defect.

Ao aorta, MV mitral valve, PA pulmonary artery, TV tricuspid valve

**Figure 4** Coronary sinus defect in case 3

We confirmed communication between both atria through the defect (arrow) by inserting a catheter through the coronary sinus orifice. The image is a surgical photograph from case 3.
**Table legends**

**Table 1** Background characteristics of patients with coronary sinus atrial septal defect

- BSA: body surface area,
- NSR: normal sinus rhythm,
- AF: atrial fibrillation,
- HT: hypertension,
- DM: diabetes mellitus,
- DL: dyslipidemia,
- OMI: old myocardial infarction,
- COPD: chronic obstructive pulmonary disease,
- CKD: chronic kidney disease,
- HD: hemodialysis,
- PCI: percutaneous coronary intervention,
- RFCA: radiofrequency catheter ablation,
- cesarean sec: cesarean section

**Table 2** Preoperative laboratory data

- WBC: white blood cells,
- RBC: red blood cells,
- Hb: hemoglobin,
- Plt: platelets,
- TP: total protein,
- AST: aspartate transaminase,
- ALT: alanine transaminase,
- BUN: blood urea nitrogen,
- Cre: creatinine,
- eGFR: estimated glomerular filtration rate,
- HbA1c: hemoglobin A1C,
- BNP: brain natriuretic peptide

**Table 3** Preoperative transthoracic echocardiography data

- BSA: body surface area,
- CS: coronary sinus,
- Qp/Qs: pulmonary-to-systemic blood flow ratio,
- RA 4ch: right atrium diameter on four-chamber view,
- RV 4ch: right ventricular diameter on four-chamber view,
- TR: tricuspid regurgitation,
- TRPG: tricuspid regurgitant pressure gradient,
- LVIVST: left ventricular interventricular septal thickness,
- PWT: LV posterior wall thickness,
- Dd: LV end-diastolic dimension,
- Ds: LV end-systolic dimension,
- EF: LV ejection fraction,
- MR: mitral regurgitation,
- AR: aortic regurgitation,
- PR: pulmonary regurgitation

**Table 4** Cardiac catheter flow study and pressure study results

- Qp/Qs: pulmonary-to-systemic blood flow ratio,
- L to R: left to right,
- R to L: right to left,
- PAP s/m: pulmonary artery pressure systolic/mean,
- PCWP: mean pulmonary capillary wedge pressure,
- C.I.: cardiac output index
Table 5 Cardiac catheter oxygen saturation study results

SVC superior vena cava, RA right atrium, RV right ventricle, PA pulmonary artery, PCW pulmonary capillary wedge, Ao aorta

Table 6 Operative and postoperative results in five cases of coronary sinus atrial septal defect

The morphology of URCS was classified as reported by Kirklin and Barratt-Boyes as follows: type I, completely unroofed with PLSVC; type II, completely unroofed without PLSVC; type III, partially unroofed midportion; and type IV, partially unroofed terminal portion.3

URCS unroofed coronary sinus syndrome, PLSVC persistent left superior vena cava, AXC aortic cross-clamp, re-AXC repeat aortic cross-clamp, CPB cardiopulmonary bypass, TAP tricuspid annuloplasty, MVP mitral valve repair (plasty), LAAA left atrial appendage amputation, AVR aortic valve replacement, RA right atrium, MAP mitral annuloplasty, POD postoperative day, ICU intensive care unit, HD hemodialysis, CSM cervical spondylotic myelopathy

Supplemental Table 1 Number of congenital heart disease and open-heart surgeries performed in the past 20 years

The numbers in parentheses represent the number of concomitant anomalies, as described precisely below.

We identified one case of VSD (type I) with concurrent RVOT stenosis in 2002; one case of secundum ASD with PLSVC in 2003; one case of secundum ASD with sinus venosus ASD and one case of secundum ASD with PDA in 2005; one case of BAV with PDA in 2006; one case of secundum ASD with VSD (type II) and one case of sinus venosus ASD with PAPVR in 2007; one case of sinus venosus ASD with PAPVR and PLSVC in 2008; one case of BAV with PDA in 2009; one case of PDA with CoA in 2010; one case of secundum ASD with PA stenosis in 2012; one case of secundum ASD with PA aneurysm in 2013; one case of BAV with CPAF in 2014; one case of VSD (type II) with a double-chamber RV in 2015; and one case of sinus venosus ASD with concomitant PAPVR in 2017. The details of “others” in Table 6 are as follows: one case of QAV in 1999 and one in 2016, two cases of QAV in 2005, one case of aorto-pulmonary arterial fistula in 2008, and one case of PA aneurysm in 2013. In the past 20 years, the redo operations were as follows.
Secundum ASD patch reclosure was performed because of detachment of a previous ASD patch in a 62-year-old woman in 2005, an 81-year-old woman in 2012, and a 45-year-old man in 2015. MVP and TAP were performed because of recurrent MR and TR after atrioventricular septal defect repair was performed in a 68-year-old woman in 2008. Mitral valve replacement and MVP were performed because of recurrent MR after primary ASD repair in a 73-year-old man in 2009 and a 70-year-old woman in 2013. PDA division and closure because of recurrent PDA was performed in a 73-year-old woman in 2010.

ASD atrial septal defect, URCS unroofed coronary sinus, VSD ventricular septal defect, I type I VSD, II type II VSD, PAPVR partial anomalous pulmonary venous return, AVSD atrioventricular septal defect, ToF tetralogy of Fallot, BAV bicuspid aortic valve, CoA coarctation of the aorta, PDA patent ductus arteriosus, Ebstein Ebstein anomaly, CPAF coronary artery–pulmonary artery fistula, OHS open-heart surgery, PA pulmonary artery, QAV quadricuspid aortic valve