

A case report of an elderly male with isolated coronary sinus atrial septal defect

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Background

Isolated coronary sinus atrial septal defect (CSASD) is a rare congenital cardiac anomaly, comprising <1% of atrial septal defects. Elderly patients with this anomaly are even more uncommon and sometimes overlooked.

Case summary

A 73-year-old man with a history of electrical defibrillation therapy for atrial flutter presented with worsening exertional dyspnoea. Cardiac examination revealed CSASD without persistent left superior vena cava, showing only moderate tricuspid regurgitation. Surgical repair of the defect and regurgitant valve improved symptoms dramatically.

Discussion

Elderly patients with atrial arrhythmias might show uncommon presentations of congenital heart disease. Cardiologists should pay attention to enlarged right ventricle, pulmonary artery, and, in particular, enlarged coronary sinus. Direct closure with interrupted sutures secured with pledgets is effective in some case of terminal type of CSASD.

Keywords

Case report • Unroofed coronary sinus • Interatrial communication • Direct closure • Elderly patient

Learning points

- Coronary sinus atrial septal defect (CSASD) without persistent left superior vena cava is exceptionally rare among the elderly.
- Close observation by transoesophageal echocardiography is useful for diagnosing CSASD.
- Even in elderly patients, CSASD can be treated effectively with surgery.

Introduction

Coronary sinus atrial septal defect (CSASD) is the rarest form of interatrial communication.¹ It is a type of unroofed coronary sinus (CS), with the absence of part or the entirety of a common wall between the CS and left atrium (LA), and is usually associated with persistent left superior vena cava (LSVC).² This anomaly is sometimes difficult to diagnose and may be overlooked.¹ We report the case of an elderly man with isolated CSASD and moderate tricuspid regurgitation (TR), who underwent surgical repair.

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Timeline

| | |
|-------------------------------|--|
| Background | Suggested as having ventricular hypertrophy |
| 10 years prior to first visit | Atrial flutter, pulmonary hypertension on echocardiography Successful electrical defibrillation therapy Medical treatment for resistant atrial flutter |
| 2 years prior to first visit | Healthy enough to go hiking without atrial flutter |
| 5 months prior to first visit | Palpitations |
| First visit to our hospital | Exertional dyspnoea after 50 m walk |
| 3 months after first visit | Surgical treatment |
| 23 days after surgery | Discharge home |
| 12 months after surgery | Latest follow-up |

Case presentation

A 73-year-old Asian man with hypertension was referred to our hospital with exertional dyspnoea. He underwent successful cavotricuspid isthmus ablation therapy for atrial flutter at another institution 10 years earlier. They noticed a large LA and pulmonary hypertension (PH), but did not find any shunt flow (Table 1). He had no limitations until 2 years before this presentation, when he felt palpitations. He was in New York Heart Association Class III and was treated with diuretics by a family physician, but treatment failed to markedly improve symptoms.

He had a Grade 2 systolic murmur in the third intercostal space on the left sternal border. There was no extremity oedema. Electrocardiography showed sinus rhythm with first-degree atrioventricular block, complete right bundle branch block, and ST-T abnormalities. Transthoracic echocardiography (TTE) revealed mild ventricular septal hypertrophy, mild PH, moderate TR, and left-to-right shunt. Tricuspid valvular annulus was dilated to 52.2 mm (normal $28 \pm 5 \text{ mm}^3$) or 25.8 mm/m^2 in diameter. Transoesophageal echocardiography (TOE) showed an enlarged CS. Catheterization study showed O2 step-up between right atrium (RA) and right ventricle (Table 2). Left atrium angiography suggested the defect represented inferior sinus venous-type atrial septal defect (ASD).

Intraoperative TOE showed a posteromedial left atrial wall defect communicating with the dilated CS draining into the RA through a Thebesian valve, establishing the diagnosis of CSASD (Figure 1).

Surgery was performed through a midline sternotomy under standard cardiopulmonary bypass. Via right atriotomy, through the dilated CS orifice (30 mm × 30 mm), a defect in the septum between the CS and LA was 2 cm × 2 cm in diameter and clearly identified (Figure 2A). No other anomalies existed. The defect was closed directly with interrupted Teflon-pledgeted mattress sutures, in

Table 1 Findings reported before catheter intervention 10 years before presenting to our hospital

| Modalities | Findings |
|------------|---|
| TTE | LAD 55 mm (normal 3.0–4.0 mm) RV basal diameter 44 mm (normal 25–41 mm) ³ Right atrium enlarged TR 2/4 degree with peak PG 42 mmHg IVC 28 mm with good respiratory fluctuation Estimated PA pressure of 52/14, mean 27 mmHg |
| TOE | Atrial septal defect was not found PFO was not found Shunt flow was not found |
| 3D CT | Normal pulmonary veins and left atrium |

3D CT, three dimensional computed tomography; IVC, inferior vena cava; LAD, Left atrium diameter; PA, pulmonary artery; PFO, patent foramen ovale; RV, right ventricular; TOE, transoesophageal echocardiography; TR, tricuspid regurgitation; TTE, transthoracic echocardiography.

combination with tricuspid annuloplasty using a 29 mm SJM Tailor ring (Figure 2B and C).

The postoperative course was uneventful. The patient was discharged on postoperative day 23. Transthoracic echocardiography at hospital discharge showed a left ventricular ejection fraction of 56%, trivial regurgitation of the tricuspid valve and no residual interatrial shunting. Warfarin was administered for 3 months. At the follow-up 12 months after the surgery, he was without functional limitations in normal sinus rhythm without conduction disturbance.

The patient gave written informed consent for publication of this report.

Discussion

Coronary sinus atrial septal defect is a deficiency of the wall between the CS and LA and also described as unroofed CS. It is the rarest type of interatrial communication¹ and is often accompanied by a persistent LSVC and other congenital diseases.² Our case had a partially unroofed terminal portion of the CS without LSVC and can be considered even rarer for the patient's age at identification. We searched through the full text of PubMed for 'coronary sinus unroofing' and 'adult' and identified only 10 cases of lone CSASD in patients aged over 60. Most elderly cases were reported after 2008, thanks to the evolution of modern imaging modalities, which include TOE, magnetic resonance imaging, and multidetector-row computed tomography (MDCT).

Coronary sinus atrial septal defect was first found incidentally during a dissection in the 1950s when catheter technique had generated interest in CS anatomy. The term 'unroofing' first meant the absence of the CS,⁴ but now also includes partial defects or fenestration of the CS. The CSASD, once seen as embryologic failure of fusion between left atriovenous fold and septum primum, is now considered as a fenestration resulted from resorption of walls between the CS and the LA.²

Table 2 Preoperative catheterization result shows O₂ step-up between right atrium and right ventricle

| Heart chamber | Oxygen saturation | Pressure (mmHg) | Estimated pressure (mmHg) |
|--------------------|-------------------|-----------------|---------------------------|
| Inferior vena cava | 75% | | |
| Superior vena cava | 66.5% | | |
| Right atrium | | | |
| High | 76.2% | 4/3/3 | 8 |
| Low | 77.8% | | |
| Right ventricle | | | |
| Apex | 82.7% | 32/6 ed 8 | |
| Outflow | 84.5% | | |
| Pulmonary artery | | | |
| Main | 83.8% | 19/13/15 | 46/13 |
| Right | 81.7% | PCW 11/9/9 | |
| Left atrium | 93.7% | 9/5/5 | |
| Left ventricle | 92.1% | 137/-9 ed 9 | |
| Aorta | 92.1% | 134/60/93 | |
| Qp/Qs | 2.27 | | 2.6 |

Pulmonary pressure from catheterization was not high from unknown reason. PCW, pulmonary capillary wedge.

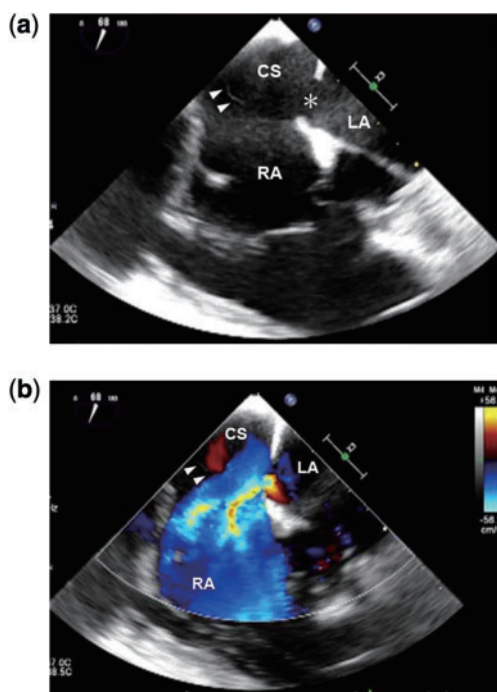


Figure 1 Intraoperative transoesophageal echocardiography shows communication between the left atrium and right atrium via coronary sinus unroofing. Note that both anterior and posterior edges are thick. (A) Thebesian valve indicates the coronary sinus. (B) Colour Doppler image shows left-to-right shunt flow. CS, coronary sinus; LA, left atrium; RA, right atrium; *, unroofed portion of the coronary sinus; arrowheads, Thebesian valve.

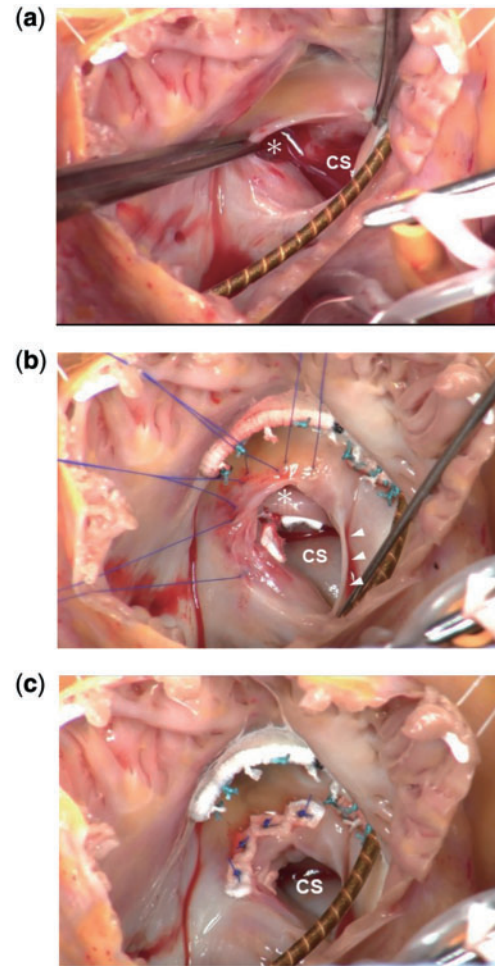


Figure 2 Intraoperative findings. (A) Coronary sinus orifice was dilated (30 mm × 30 mm), entire boundary of unroofing of the coronary sinus was clearly seen (20 mm × 20 mm) through the orifice. (B) The defect was directly closed after ring annuloplasty of the tricuspid valve. (C) Completion of the repair. CS, coronary sinus; *, unroofed portion of the coronary sinus; arrowheads, Thebesian valve.

Exact diagnosis before surgery is often difficult, however, without previous diagnosis, a CSASD is hard to diagnose during the open heart surgery.² A dilated CS is an important finding⁵ and suggests that the CS grows larger by receiving shunt flow, increasing shunt ratio.

In our case, we suspected interatrial communication on Qp/Qs obtained from TTE, but had difficulty in locating it. Reviewing preoperative diagnostic imaging, MDCT shows interatrial communication through a dilated CS (Figure 3A). Preoperative TTE shows terminal dilatation of CS in short-axis view (Figure 3B) and a considerable blood flow from near tricuspid valve in apical four-chamber view (Figure 3C). Local dilatation of CS is a typical finding. Contrast echocardiogram by infusion from left upper limb can identify the presence of LSVc.

Surgical indications are the same as those for ASD closure in patients >60 years old,² include an enlarged RA and ventricle,

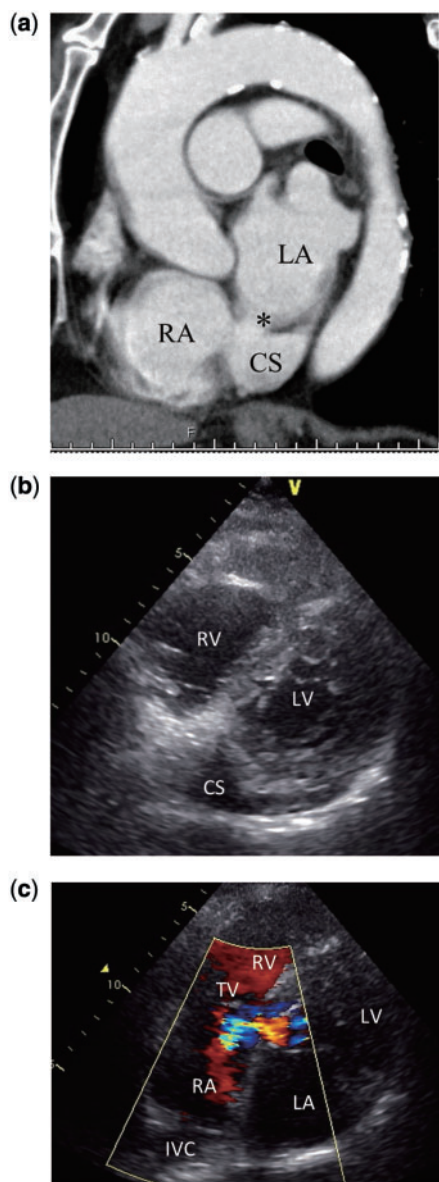


Figure 3 Reviewing of preoperative diagnostic imaging. (A) Arterial phase sagittal oblique reformatted CT scan shows a dilated coronary sinus communicate with left atrium. (B) Transthoracic echocardiography in short-axis view shows partially dilated coronary sinus tadpole-like shape. (C) Colour Doppler image in posteriorly directed apical four-chamber view shows considerable blood flow from near the septal cusp of the tricuspid valve perpendicularly to the atrial septum. CS, coronary sinus; IVC, inferior vena cava; LA, left atrium; RA, right atrium; RV, right ventricular; TV, tricuspid valve; *, unroofed portion of the coronary sinus; arrowheads, Thebesian valve.

pulmonary-systemic flow ratio ≥ 1.5 (normal 1.0), and symptoms such as paradoxical embolism or dyspnoea.⁶ Recent reports demonstrate survival rates of surgical closure are not worse, though complication rates are higher compared to younger individuals.⁶ Transcatheter therapy is also an option.

For a surgical treatment, patch closure with continuous suture is common, because avoiding bundle of His may meet fragility of superior rim.⁷ The fragility of anterior edge depends mainly on muscular thickness of left atrial-CS myocardial connection and varies a lot.⁸ The AV node is located at the apex of the Koch's triangle in a terminal type CSASD as expected in normal heart, but may be closer to the enlarged orifice.⁹ Conduction abnormalities would be avoided on the assumption that the atrioventricular (AV) node and His bundle are in the normal location.¹⁰ In our case, anterior edge was not thin and depressed, so that it satisfied both strength and distance from His bundle. We adopted interrupted mattress sutures with pledgets to avoid tension at sutures. Posterior edge was easily mobilized to side by side with the anterior edge by pulling the interrupted sutures. Enlarged orifice made the stitches from RA easier. If the anterior edge is thick and not pushed away, the posterior edge is mobile enough, and the CS orifice is large, the defect can be closed directly using interrupted pledgeted sutures.

Among patients without LSVC, the interatrial communication can be safely interrupted by closure of the CS orifice, because CS blood flow is no more than 4% of normal cardiac output; however, CS flow impairment could deteriorate cardiac function.¹¹

Tricuspid regurgitation was moderate but annulus was dilated. Annuloplasty was done according to indication for TR treatment concomitant with left heart surgery.¹²

As for anticoagulant therapy for intracardiac repair, our policy is treatment with warfarin for 3 months, that is estimated period for endothelium to cover the inner surface of heart.

Isolated CS terminal unroofing is very rare and has difficulty in diagnosis. Advances in cardiac imaging modalities have allowed diagnosis more easily. A dilated CS offers clues to a correct diagnosis. Surgical treatment represents a feasible option even in elderly patients. Direct closure can be safely performed in some terminal types of isolated CSASD.

Conclusion

Clinicians should be alert to the implications of a dilated CS. Transoesophageal echocardiography is quite useful to diagnose an unroofed CS. Isolated CSASD can be treated surgically, even in elderly patients. Some cases of terminal type of CSASD are suitable for direct closure with interrupted mattress sutures with pledgets.

Lead author biography



Junko Okamoto is a cardiovascular surgeon who works for Osaka Medical College Hospital. She graduated from Osaka Medical College in 1988. She majored in cardiovascular surgery and obtained her medical degree in 1988 from Osaka Medical College in autologous transfusion during cardiovascular surgery. After leaving for years, she currently works in the Department of Thoracic and

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Supplementary material

[Supplementary material](#) is available at *European Heart Journal - Case Reports* online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as [Supplementary data](#).

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: None declared.

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