

Anatomical Repair of Partially Unroofed Coronary Sinus Syndrome through the Coronary Sinus Orifice

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A 15-year-old girl was admitted for repair of partially unroofed coronary sinus syndrome. A preoperative echocardiographic study disclosed the dilated coronary sinus and a draining blood flow into the right atrium from both the left atrium and the end of the coronary sinus. At surgery, the distal portion of the coronary sinus roof was found to be absent. The defect was repaired through the coronary sinus orifice with a patch. No electrophysiological conduction abnormalities developed and follow-up echocardiography showed neither residual shunts nor coronary sinus blood flow disturbances. (Ann Thorac Cardiovasc Surg 2005; 11: 208–10)

Key words: congenital heart disease, atrial septal defect, unroofed coronary sinus syndrome, echocardiogram

Introduction

Unroofed coronary sinus syndrome is usually associated with a persistent left superior vena cava and other cardiac anomalies. This study reports a rare case of unroofed coronary sinus syndrome without a persistent left superior vena cava, in which the distal portion of the coronary sinus roof was fenestrated into the left atrium.

Case Report

A 15-year-old girl was admitted for an elective operation for unroofed coronary sinus syndrome. A systolic heart murmur of Levine II was found without arrhythmias. Electrocardiography showed a sinus rhythm with an incomplete right bundle branch block and no left axis deviation of the QRS complexes, and chest radiography revealed slight pulmonary congestion without an increased cardiothoracic ratio. The echocardiographic study disclosed a dilated coronary sinus and blood flows from both the coronary sinus and the left atrium draining into the right atrium

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(Fig. 1). Cardiac catheterization revealed no persistent left superior vena cava, a left-to-right shunt ratio of 37%, and a pulmonary/systemic blood flow ratio of 1.56. From these findings partially unroofed coronary sinus syndrome was diagnosed.

The operation was performed following standard procedures, and antegrade cardioplegic solution was used for myocardial protection with moderate core cooling. After right atriotomy, the fossa ovalis was found to be intact, though the coronary sinus orifice was enlarged and located more anteriorly than usual. Through the coronary sinus orifice, a round fenestration, 8 mm in diameter, was found in the coronary sinus, and fibrous tissue 3-4 mm wide was recognized between the coronary sinus orifice and the fenestration (Fig. 2). The defect of the roof was repaired without difficulty through the coronary sinus orifice using a bioprosthetic patch and interrupted sutures and the residual tissue of the coronary sinus.

No electrophysiological conduction abnormalities developed and the patient's postoperative course was uneventful. Follow-up echocardiographic examinations showed neither residual shunts nor coronary sinus blood flow disturbances.

Discussion

Unroofed coronary sinus syndrome is a rare congenital cardiac anomaly that was classified by Quaegebeur and

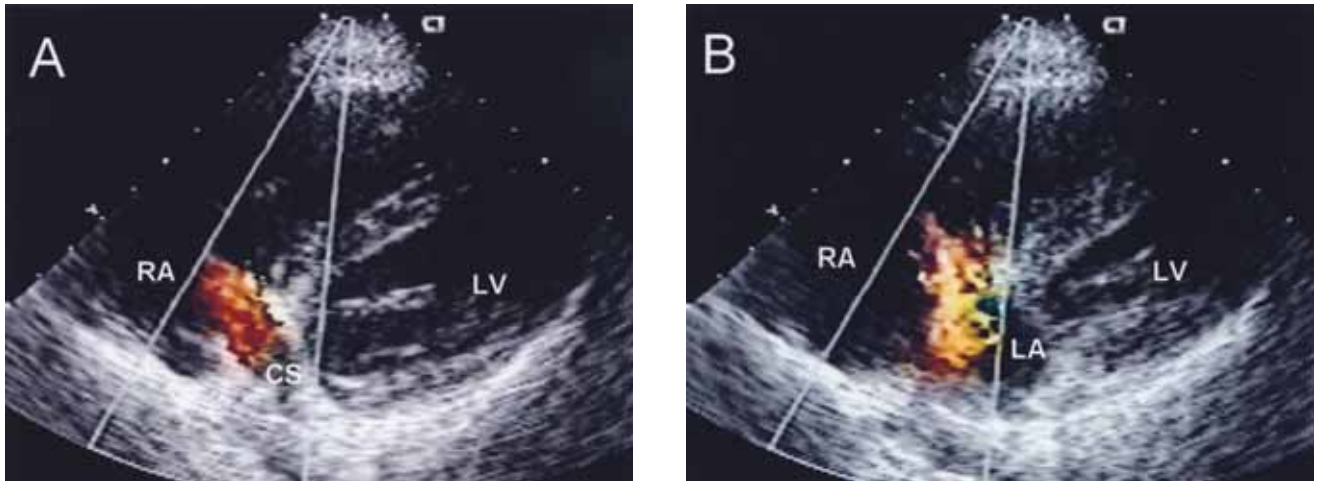


Fig. 1. Preoperative echocardiographic findings.

- A: Blood flow from the dilated coronary sinus draining into the right atrium.
- B: Blood flow from the left atrium draining into the right atrium through a defect of the coronary sinus roof.

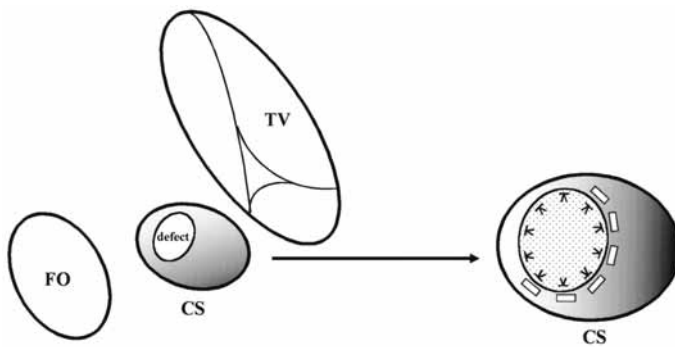


Fig. 2. Operative findings.

The fenestration in the sinus roof into the left atrium was recognized through the dilated coronary sinus orifice. The rim of the fenestration consists of some fibrous tissue. The defect was repaired with a bioprosthetic patch.

colleagues.¹⁾ The pure form of the syndrome is associated with a left superior vena cava and the total absence of a partition between the coronary sinus and left atrium. Partially unroofed coronary sinus syndrome is not associated with a persistent left superior vena cava, and defects are recognized at different sites of the coronary sinus. Unroofed coronary sinus syndrome might be associated with other cardiac anomalies such as anomalous pulmonary venous return, cor triatriatum, tetralogy of Fallot, and atrioventricular canals.¹⁻⁵⁾ Echocardiographic studies and magnetic resonance imaging are helpful for obtaining preoperative information about unroofed coronary sinus syndrome;^{6,7)} however, sometimes remains to be diagnosed intraoperatively.

Surgical procedures for unroofed coronary sinus syndrome vary depending on the associated cardiac anomalies. With partially unroofed coronary sinus syndrome without the left superior vena cava, surgery involves roof-

ing the unroofed portion or allowing the coronary sinus to drain into the left atrium by patch closure of the coronary orifice.^{1,8-10)} The former is usually employed when the mid-portion of the sinus roof is fenestrated, while the latter is employed when the downstream portion of the roof is fenestrated. Conduction disturbances including complete atrioventricular blocks can lead to serious post-operative complications after downstream portion repair. To avoid these complications, care is required during the procedures for repair of the downstream portion.

In the case study documented here, preoperative echocardiographic and operative findings confirmed the diagnosis of partially unroofed coronary sinus syndrome. The unroofed portion was located and found to be limited to the distal portion of the coronary sinus roof, thus resembling a downstream portion defect. However, the patient was diagnosed as having a mid-portion defect because fibrous tissue was found between the coronary

sinus orifice and the coronary sinus fenestration. In such cases, allowing the coronary sinus to drain into the left atrium is a possible strategy; however, we chose anatomical repair of the defect, which was easily accomplished without any postoperative complications. It is suggested therefore, that anatomical repair should be the initial strategy for all types of unroofed coronary sinus syndrome.

References

1. Quaegebeur J, Kirklin JW, Pacifico AD, Barger LM Jr. Surgical experience with unroofed coronary sinus. *Ann Thorac Surg* 1979; **27**: 418–25.
2. van Son JA, Autschbach R, Mohr FW. Repair of cor triatriatum associated with partially unroofed coronary sinus. *Ann Thorac Surg* 1999; **68**: 1414–6.
3. Watanabe H, Hayashi JI, Sugawara M, Yagi N. Complete unilateral anomalous connection of the left pulmonary veins to the coronary sinus with unroofed coronary sinus syndrome: a case report. *Thorac Cardiovasc Surg* 1999; **47**: 193–5.
4. Alexi-Meskishvili V, Dahnert I, Beyer E, Hetzer R. Successful total correction of complete atrioventricular canal, total anomalous pulmonary venous drainage and unroofed coronary sinus in an infant. *Eur J Cardiothorac Surg* 1999; **15**: 95–6.
5. van Son JA, Hamsch J, Mohr FW. Modified repair of mixed anomalous pulmonary venous connection. *Ann Thorac Surg* 1998; **65**: 1441–2.
6. Hahm JK, Park YW, Lee JK, et al. Magnetic resonance imaging of unroofed coronary sinus: three cases. *Pediatr Cardiol* 2000; **21**: 382–7.
7. Vargas-Barron J, Espinola-Zavaleta N, Romero-Cardenas A, et al. Two- and three-dimensional echocardiographic unroofed coronary sinus. *J Am Soc Echocardiogr* 2001; **14**: 742–4.
8. Nakano H, Sato T, Chin Y, Uetsuka Y, Tuzuku Y. Surgical management of an unroofed coronary sinus: a case report. *Kyobu Geka* 1987; **40**: 507–12.
9. Inoue T, Arimitsu K, Honda S, Yamabe K, Imachi T. A case report of coronary sinus atrial septal defect. *Nippon Kyobu Geka Gakkai Zasshi* 1993; **41**: 474–8. (in Japanese)
10. Kawahito T, Kitagawa T, Hori T, Chikugo F, Shimoe Y, Katoh I. Surgical experience of two different types of unroofed coronary sinus. *Nippon Kyobu Geka Gakkai Zasshi* 1996; **44**: 1015–21.