Unroofed coronary sinus syndrome: Diagnosis, classification, and surgical treatment

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Unroofed coronary sinus syndrome (URCS) is a rare cardiac anomaly in which a communication occurs between the coronary sinus and the left atrium as a result of the partial or complete absence of the roof of the coronary sinus. This entity is strongly associated with a persistent left superior vena cava (LSVC), with or without a connection between both superior venae cavae.[1] The diagnosis of this lesion is important to the prognosis of the patient because of the consequences of brain abscess or cerebral emboli that may result from a right-to-left shunt. However, the diagnosis is often difficult because of nonspecific clinical features. In this report we present our experience with the diagnosis, classification, and surgical treatment of 11 cases of URCS.

Patients and Methods

Between June 1986 and August 2000, a total of 11 consecutive patients with URCS underwent surgery (Table 1). The patients with URCS who had atrial isomerism were excluded from this study. Mean age was 8.0 ± 6.3 years (1 month–16.5 years).

The morphologic type of URCS was classified as Kirklin and Barratt-Boyes[1] reported: (type I, completely unroofed with LSVC; type II, completely unroofed without LSVC; type III, partially unroofed midportion; type IV, partially unroofed terminal portion). Preoperative transthoracic echocardiography and cardiac catheterization were performed in all cases.

During the operation, all patients who were found to have an LSVC were monitored with bilateral blood pressure readings of the superior venae cavae. Before cardiopulmonary bypass, the clamped pressure of the LSVC was measured. When the pressure was greater than 16 mm Hg, we decided not to ligate the LSVC but to reroute the LSVC to the right atrium, and an intracardiac baffle repair (ICBR) was performed with antegrade cardioplegia. In the 3 patients who had an atrioventricular septal defect, the LSVC was directly cannulated. In 2 patients the LSVC was cannulated through an intracardiac orifice of the LSVC. Autologous pericardium or expanded polytetrafluoroethylene was used to create the baffle. The ICBR was constructed along the superior wall of the left atrium and ended with an opening at the level of the atrial septal patch. When the pressure of the LSVC was less than 15 mm Hg, the LSVC was ligated, and then the atrial septal defect was closed avoiding the atrioventricular node and bundle of His, and the main drainage of the small coronary vein was left-sided. Closure of the atrial septal defect was performed for those patients with type II URCS; therefore the main drainage of coronary vein was left-sided. For the patients with type III and type IV URCS, roof repair or closure of the coronary sinus with or without LSVC ligation was performed. The operative method should be based on the anatomic findings around the atrioventricular node and bundle of His.

Postoperative echocardiography was performed in all cases. Postoperative cardiac catheterization was also performed in all cases except that of patient 9.

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Results

URCS was confirmed by preoperative diagnosis in 6 cases, and we had suspicions of URCS in 3 cases with a final diagnosis made during the operation. Two cases of URCS were diagnosed after the definitive operation of the heart defect. Postoperative transthoracic echocardiography revealed type III URCS. The patients in whom the diagnosis was not made perioperatively had type III URCS. URCS of these patients was easily defined by angiography of the LSVC or contrast echocardiography with an injection of saline solution into a vein of the left arm.

There were no operative deaths. No patients had perioperative complications related to ICBR or the ligation of the LSVC. No patients had preoperative or perioperative arrhythmia, and all patients were in sinus rhythm after the repair. Postoperative echocardiography and cardiac catheterization demonstrated normal anatomy, patent LSVC, and the absence of a residual shunt, pulmonary venous obstruction, or stenosis of the baffle. The mean follow-up time was 85.5 months (minimum 22 months, maximum 191 months), and there were no late deaths.

Discussion

Before the era of echocardiography, precise diagnosis of this anomaly was only possible during a surgical procedure or at autopsy. Since then, several studies have reported the usefulness of 2- and 3-dimensional transthoracic and transesophageal echocardiography in diagnosing URCS.

TABLE 1. Patient data, diagnosis of URCS, and operations

Case	Age (y)	Sex	Diagnosis	Previous operation	LSVC	Type	Timing of diagnosis			Operation	Main drainage of coronary vein
							Preoperative	Intraoperative	Postoperative		
1	3.0	M	AVSD	PAB	+	I	+			Two-patch repair, ICBR	LA
2	1	M	VSD(I), MSR	−	−	IV	+			VSD closure, MVR, CS closure	LA
3	8.7	F	AVSD, PS	−	+	IV	−	+		Two patch repair, TAP, LSVC ligation	RA
4	4.6	F	VSD, ASD, cor triatriatum	−	+	IV	+			VSD closure, roof closure, diaphragm excision	RA
5	15	M	Cor triatriatum, ASD	−	+	I	+			Diaphragm excision, ASD closure, ICBR	LA
6	10	M	PA, VSD, ASD	Left modified BT	+	IV	−	+		Rastelli type repair, CS closure, LSVC ligation	LA
7	16.5	F	TOF, cor triatriatum	Right BT, TC	+	III	−	−	+	Diaphragm excision, CS closure, LSVC ligation	LA
8	0.8	F	AVSD, cor triatriatum	PAB, PDA ligation	+	I	+			Two-patch repair, diaphragm excision, ICBR	LA
9	12.6	F	ASD, PS	−	−	IV	+			Roof closure, commisurotomy	RA
10	0.1	M	Cor triatriatum, ASD	−	−	II	−	+		Diaphragm excision, roof closure	RA
11	15.6	M	PA, VSD, ASD	Bilateral BT, TC	+	III	−	−	+	CS closure, LSVC ligation	LA

AVSD, Atrioventricular septal defect; *PAB*, pulmonary artery banding; *LA*, left atrium; *VSD*, ventricular septal defect; *MSR*, mitral stenosis and regurgitation; *MVR*, mitral valve replacement; *CS*, coronary sinus; *PS*, pulmonary stenosis; *TAP*, transannular patch repair; *RA*, right atrium; *ASD*, atrial septal defect; *PA*, pulmonary atresia; *BT*, Blalock-Taussig shunt; *TOF*, tetralogy of Fallot; *TC*, total correction of the heart defect; *PDA*, patent ductus arteriosus.

In our series, preoperative diagnosis was made in 6 cases, and we had suspicions of URCS before the operation in 3. However, a residual shunt remained in 2 patients after the total correction of the heart defect. These 2 patients had type III URCS with LSVC. Although there has been a report of successful preoperative diagnosis of type III URCS,[2] our results demonstrated that the diagnosis of this type III URCS is still difficult, especially in the children with complex cardiac malformations. After this experience, we routinely injected contrast material into the upper LSVC in the patients with heart defects associated with LSVC. When URCS was suspected by angiography of the LSVC, the injection of saline solution into a vein of the left arm provided contrast in the LSVC and made it possible to see that the microbubbles dispersed throughout the atrium because of the absence of the coronary sinus roof.

With our criteria for LSVC ligation, cerebral and upper limb edema or petechiae were not seen in the patients with LSVC ligation. Postoperative echocardiography and cardiac catheterization confirmed the patency in the baffle, and there was no evidence of pulmonary venous congestion in the patients with ICBR. The treatment for LSVC associated with URCS is still controversial.[3-5] However, measuring the pressure of the LSVC may be a helpful procedure to decide whether a surgeon should ligate it.

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