Ventricular Septal Defect of the Atrioventricular Canal Type—Report of a Surgically Treated Case—

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Summary: A 19-month-old male infant with Down syndrome was referred for investigation of heart murmur. An electrocardiogram showed normal axis (+100°) of the QRS complex, incomplete right bundle branch block, and biventricular hypertrophy. Echocardiography revealed a ventricular septal defect (VSD) and a cleft of the anterior mitral leaflet with mild valve regurgitation. The anterior mitral leaflet and the septal tricuspid leaflet attached to the same level of the ventricular septum were also delineated, however, no atrial septal defects were detected. Cardiac catheterization demonstrated pulmonary hypertension and a left to right shunt at the right ventricle. During operation, no defects in the atrial septum and no cleft of the septal tricuspid leaflet were found. Cleft of the anterior mitral leaflet, continuity between the mitral anterior leaflet and the tricuspid septal leaflet, and a VSD of the atrioventricular canal type were confirmed. The cleft of the anterior mitral leaflet and the VSD were repaired with the aid of cardiopulmonary bypass. The patient recovered uneventfully.

Key words ventricular septal defect, persistent common atroventricular canal, cleft of the anterior mitral leaflet, atrioventricular canal type ventricular septal defect

INTRODUCTION

Ventricular septal defect (VSD) is the most common congenital heart anomaly, and is found either as an isolated lesion, or in combination with other cardiac anomalies. The ventricular septum is embryologically composed of the endocardial cushions of the atrioventricular (A-V) canal, the conus septum, and the muscular septum, therefore, the defect may appear in a variety of areas in the ventricular septum. The majority of VSDs generally develop adjacent to the membranous septum, however, a VSD beneath the septal tricuspid leaflet, the so-called A-V canal type of VSD, is rare, usually comprising 5% or less of surgically treated VSDs [1-6]. In Japan, only 6 cases of the A-V canal type of VSD have been described, to our knowledge [4-6]. In addition, from the surgeon’s point of view, the A-V canal type VSD has several distinctive anatomical and clinical characteristics, compared with the common types of VSD which arise in other areas in the ventricular septum.

In this paper, we present a case of the A-V canal type of VSD with a cleft of the anterior mitral leaflet and discuss the diagnostic and surgical characteristics of the A-V canal type VSD.

CASE REPORT

A 19-month-old male infant, who had a mongolism-face appearance, was referred to our hospital for investigation of heart murmur and a failure to thrive. His body weight was 7605 g, heart rate was 120/min with regular rhythm, and respiratory rate was 50/min. Cardiac examination revealed a grade 4/6 systolic regurgitant murmur with thrills over the third intercostal space at the left sternal border. An electrocardiogram (ECG) showed normal axis (+100°), PR interval of 0.14 sec, incomplete right bundle branch block, and biventricular hypertrophy (Fig. 1), while a chest X-ray revealed moderate
cardiomegaly and prominent pulmonary vascularity. Echocardiography demonstrated dilatation of both ventricles, a perimembranous VSD of 10×14 mm (Fig. 2-A), and a cleft of the anterior mitral leaflet with mild valve regurgitation (Fig. 2-B). The anterior mitral leaflet and the septal tricuspid leaflet attached to the same level of the ventricular septum were also delineated, however, no atrial septal defects were detected by echocardiography (Fig. 2-A). Cardiac catheterization revealed severe pulmonary hypertension (Pp/Ps=0.87) and a left to right shunt at the right ventricle (Qp/Qs=2.64). Pulmonary vascular

Fig. 1. The preoperative ECG showing normal electrical axis (+100°) of the QRS complex, incomplete right bundle branch block, and biventricular hypertrophy.

Fig. 2. The preoperative echocardiograms showing a large perimembranous VSD of 10×14 mm (arrows), the anterior mitral leaflet and the septal tricuspid leaflet attached to the same level of the ventricular septum, and no atrial septal defects (A), a cleft of the anterior mitral leaflet (arrow head) with mild valve regurgitation (B). LA: left atrium; LV: left ventricle; RA: right atrium; RV: right ventricle; M: mitral valve; T: tricuspid valve

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TABLE 1.
Cardiac catheterization

<table>
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<tr>
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<th>Pressure (mean) mmHg</th>
<th>O₂ Saturation (%)</th>
<th>100% Oxygen Inhalation</th>
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<td></td>
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<tr>
<td>SVC</td>
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<td>66</td>
<td>68</td>
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<td>IVC</td>
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<td>74</td>
<td></td>
</tr>
<tr>
<td>RA</td>
<td>a=10, v=8 (6)</td>
<td>98/12</td>
<td>95/28 (61)</td>
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<td></td>
<td></td>
<td>63</td>
<td>83</td>
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<td></td>
<td></td>
<td>94</td>
<td>92</td>
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<tr>
<td>Aorta</td>
<td>124/65 (92)</td>
<td>120/62 (88)</td>
<td>99</td>
</tr>
<tr>
<td>LV</td>
<td>125/13</td>
<td>84</td>
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Qp/Qs=2.64, PVRI=7.6 unit Qp/Qs=4.0, PVRI=3.92 unit

Fig. 3. The preoperative left ventriculograms showing a cleft of the anterior mitral leaflet (arrows) with mild valve regurgitation (A), and a large amount of left to right shunt through the defect (B).

resistance index (PVRI) was calculated as 7.6 units. After administration of 100% oxygen for 10 min, Qp/Qs increased to 4.0, and PVRI decreased to 3.92 units as shown in Table 1. Left ventriculography demonstrated a large amount of shunt through the VSD and mild mitral regurgitation via the cleft of the anterior mitral leaflet (Fig. 3-A, B). On the basis of these findings, the diagnosis of a perimembranous VSD combined with severe pulmonary hypertension, mild regurgitation due to the cleft of the anterior mitral leaflet, and Down syndrome was made.

During operation, the right atrium was opened
under total cardiopulmonary bypass and myocardial protection with cold blood cardioplegia. There were neither ostium secundum type defects nor ostium primum type defects in the atrial septum. No deformity or cleft of the septal tricuspid leaflet were found. The left atrium was approached through the atrial septum. The anterior mitral leaflet was cleft from base to apex, and the continuity between the mitral anterior leaflet and the tricuspid septal leaflet resembling the A-V valve of incomplete forms of mitral anterior leaflet and the tricuspid septal leaflet from base to apex, and the continuity between the atrial septum. The anterior mitral leaflet was cleft found. The left atrium was approached through the tricuspid valve. The anterior mitral and the septal tricuspid leaflets were attached to the crest of the ventricular septum by short, thick chordae tendineae. The cleft of the anterior mitral leaflet was repaired with interrupted sutures and the VSD was closed with a dacron patch. After weaning off cardiopulmonary bypass, systolic pressure in the pulmonary artery dropped to 37 mmHg, when systolic pressure in the left ventricle was 92 mmHg.

The patient recovered uneventfully and is doing well at present 14 months after surgery.

DISCUSSION

Ventricular septal defect is the most common congenital heart anomaly, and the defect may appear in a variety of areas in the ventricular septum. Soto and colleagues [3] categorized VSDs into four types; (1) perimembranous, (2) muscular, (3) subarterial infundibular, and (4) mixed defects. According to this classification, the A-V canal type VSD is included among the perimembranous defects, particularly perimembranous inlet defects. Although perimembranous VSDs are most frequently observed at autopsy or during surgery, the A-V canal type VSD is rare. They found the A-V canal type VSD in only 4 (1.8%) of 220 necropsy hearts with isolated or multiple VSDs [3]. Similarly, Neufeld and colleagues [1] found the A-V canal type VSD in 15 (4.2%) of 360 cases with VSDs at necropsy and surgery. On the other hand, only 6 operated cases of the A-V canal type VSD have been reported in the literature in Japan, to our knowledge [4-6].

The ventricular septum is embryologically composed of the endocardial cushions of the A-V canal, the conus septum, and the muscular septum. Among these three elements, deficiency of the endocardial cushions of the A-V canal results in a defect beneath the septal tricuspid leaflet, the so-called A-V canal type of VSD. Furthermore, the endocardial cushions of the A-V canal also contribute to the formation of the A-V valves and the atrial septum. Therefore, deficiency of the endocardial cushions of the A-V canal may develop an isolated atrial septal defect of the ostium primum type, an isolated cleft of the mitral or tricuspid valve or of both, an isolated VSD with or without cleavage of the A-V valves, or a combination of these anomalies. In our patient, the deficiency occurred at the junction of the lower portion of the A-V endocardial cushions, and it apparently involved the left A-V valve, but not the atrial septum.

The electrocardiographic findings in the A-V canal type VSD are generally characteristic. Left axis deviation of the QRS complex, right ventricular volume overloading or right ventricular hypertrophy, and a prolonged PQ interval, are similar to the findings in persistent common A-V canal [1-6]. These electrocardiographic changes have been considered to result from the presence of a congenital anomaly or distortion of the conduction system. The elongation and looping of the bundle around the defect may be the cause of the alteration in excitation that gives the characteristic pattern on ECGs [1]. Among these findings, left axis deviation of the QRS complex was noted in most of the cases previously reported, and has been emphasized to be the most diagnostic finding [1,2,5]. However, Char and colleagues [7] observed left axis deviation of the QRS complex between 0° and −180° in 16% of 100 children with isolated VSD. On the other hand, Hosoi and colleagues [4] reported 2 autopsied cases with the A-V canal type VSD, in which preoperative ECGs showed no left axis deviation of the QRS complex. In our patient, right ventricular hypertrophy was found, however, the mean electrical axis of the QRS complex was +100°. These facts suggest that left axis deviation of the QRS complex is a very valuable electrocardiographic finding, but it is not a change specific to ECGs in patients with the A-V canal type VSD.

From the surgical standpoint, repair of the defect is technically difficult because the chordae tendineae of the mitral and tricuspid valves attached to the crest of the ventricular septum obscure the area to be stitched and make it difficult to secure placement of a patch. In addition, the anatomical characteristics of the conduction system, particularly the bundle of His in the proximity of the posteroinferior rim of the
defect, occasionally lead to injury of the bundle by stitches to repair the defect resulting in occurrence of complete atrio-ventricular block. In our patient, we were able to avoid complete atrioventricular block by placing the stitches on the right surface of the ventricular septum 3-5 mm away from the rim of the defect. However, postoperative left ventriculography showed a minor residual shunt through the defect.

In summary, we described a 19-month-old infant with the A-V canal type VSD, severe pulmonary hypertension, a cleft of the anterior mitral leaflet and Down syndrome, and discussed the diagnostic characteristics of ECGs and some surgical challenges faced when repairing A-V canal type VSD.

REFERENCES