Atroventricular Septal Defects*
— Angiographic Analysis of 31 Cases —

Shi Joon Yoo, M.D., Cheong Hee Park, M.D., Kyung Mo Yeon, M.D.,
Man Chung Han, M.D.

Department of Radiology, College of Medicine, Seoul National University

心房室中隔缺損
心血管映畵擺影術所見에 關한 考察

<国文抄録>

先天生性心臓病の手術前 正確な形態学的診断は 未だ完璧な状況を与えるのは 常に 重要
な要素である 知知的事実である。最近のaxial cineangiographyの登場と心臓の観察部位を X -
透の走行に垂直面を主に先天生性心臓病の全曲正確な手術前 形態学的評価を可能に
した。著者等は 1980年から 1984年まで 首爾大學校病院 放射線科にて 心導子術と 心血管映畵
撮影術を施行し 心房室中隔缺損を診断した 31例を対象に 分析を 進行して axial viewにての心
血管造影所見を 前後 側面像にての所見と 連関せり 心房室中隔缺損の 手術前 評価に 必須
の造影剤注入部位と 撮影方法を決定した。

1. 側面の 前後・側面像にて 観察せる能できる左心室流出路の "가위목 变形 (goose-neck deformity)") , 婢側面の畸形 , 左心室流入部の流出路の disproporti
on 等の 所見は axial viewにての 所見と 一致せり。

2. 房室口 (atrioventricular orifice)の partitioning 與否は 4-chamber viewにて 左心室造影
を施行して 心室の拡張期に 房室間隔に 依ず 所有な 関連を 観察して 区分が可能となり 4-chamber viewにての 右心室造影を 施行することにより 見 ものの正確性を 期待せり。

3. 4-chamber viewにて 左上肺動脈造影を 施行することにより 心房間の 靜流の 有無 , 位相 , 周期の 判定
が可能せり。

4. 心室間の 転流と 房室間隔と 心室 中隔の 関係の 判定は 4-chamber viewにての 左心
室造影が 見当正確さを 瑕接せり。

5. 手術後 視野に 心室瓣弁不全の 関連を 有する 逆行性 左心室造影の 必要性が 切迫せり。

6. 心房室中隔缺損の 手術前 心血管造影術にて 逆行性 左心室造影, 4-chamber viewにての 右心室造影 及び 4-chamber viewにての 右上肺動脈造影が 必須せり。

*이 논문은 1985 년도 서울대학교병원 특별연구비조로 이루어진 것임
이 논문은 1985 년 2월 8일에 접수하여 1985 년 2월 25일에 수락되었음。
**Introduction**

Although there has been no unanimous opinion on the basic pathologic features of the atrioventricular septal defects (AVSD's), "goose-neck" deformity of the left ventricular outflow tract in a frontal angiogram has been regarded as an angiographic denominator of the AVSD's. Advancements in cardiac surgical capability have prompted the more accurate angiographic demonstration of the features of surgical importance in addition to the generalized diagnosis. Recent use of axial views has permitted the demonstration of the more detailed pathologic anatomy by placing the regions of interest at a right angle to the X-ray beam. The purposes of this report are to correlate the angiographic findings demonstrated in axial views with those demonstrated in the conventional views and to provide the essential injections and projections in the preoperative evaluation of the AVSD's.

**Materials and Methods**

31 patients of AVSD's in which cardiac catheterization and cineangiography were done at the Department of Radiology, Seoul National University Hospital between 1980 and 1984 were selected for the angiographic analysis. Patients with an AVSD coexisting with cardiac malposition and single ventricle were excluded.

Biplane 35mm cineangiograms were made at 60 frames/sec after the cardiac catheterization. The cineangiographic unit used was the fixed tube system, GE MSI-1250-biplane X-ray Generator Unit. The injections and projections used are presented in Table 1. The 4-chamber view was obtained by placing the patient in a 45° left anterior oblique and 35-45° sitting-up position with or without slanting on the table. The complementary right anterior oblique (RAO) view was made by a lateral tube during the 4-chamber projection. The elongated RAO view was obtained by placing the patient in a 30° RAO and 35-45° sitting-up position using a vertical tube.

Each angiogram was interpreted independently and then correlated with the clinical and laboratory findings. Efforts were made to correlate the angiographic findings with the operative findings in 19 patients.

**Results**

10 of the 31 patients were male and 21 female. They ranged in age from 7 months to 25 years. Angiographic findings were:

1. Absent atrioventricular septum (AVS); The straight segment between the noncoronary cusp and the crux cordis in the left ventriculogram, representing the foreshortened profile of the AVS in the 4-chamber view (Fig. 2) and the upper and single ventricle were excluded. margin of the en face of the AVS in the elongated RAO plane 35mm cineangiograms were made at RAO view of the normal heart, was deficient or deformed (Fig. 3, 4). The right margin of the conventional AP left ventriculogram between the aortic valve and the crux cordis was concave (Fig. 1). The aortic valve-crus cordis distance was increased, together with the downward displacement of the crux cordis. It occupied the 34.7-61.1 (av. 47.4%) of the apex-aortic valve distance in the 4-chamber view. It occupies only a short subaortic segment in the 4-chamber view of the normal heart.

2. Disproportion between the lengths of the inflow and outflow tracts of the left ventricle; The posterior aspect of the left ventricular inlet septum was short and the outflow tract was

<table>
<thead>
<tr>
<th>Table 1. Injections and Projections Used</th>
</tr>
</thead>
<tbody>
<tr>
<td>Conventional AP and lateral LV</td>
</tr>
<tr>
<td>with 4-chamber LA</td>
</tr>
<tr>
<td>4-chamber LV</td>
</tr>
<tr>
<td>with complementary RAO</td>
</tr>
<tr>
<td>with 4-chamber LA</td>
</tr>
<tr>
<td>with 4-chamber RV</td>
</tr>
<tr>
<td>with elongated RAO LV</td>
</tr>
<tr>
<td>Conventional AP and lateral LV and</td>
</tr>
<tr>
<td>4-chamber LV</td>
</tr>
<tr>
<td>with 4-chamber LA</td>
</tr>
<tr>
<td>LV=left ventricular injection</td>
</tr>
<tr>
<td>RV=right ventricular injection</td>
</tr>
<tr>
<td>LA=left atrial or right upper pulmonary vein injection</td>
</tr>
</tbody>
</table>

---

- 300 -
Fig. 1. Conventional AP left ventriculograms of a patient with AVSD. A. Diastolic phase; Typical "goose-neck" deformity of the left ventricular outflow tract is formed between the ventricular free wall and the mural attachment (arrows) of the mitral valve or component. Notice the disproportion between the apex (A)-crux cordis (CC) distance and the apex (A)-aortic valve (NC) distance. The aortic valve (NC)-crux cordis (CC) distance which is the AP diameter of the atroventricular annulus is increased together with the downward displacement of the crux cordis. The left ventricular aspect of the atroventricular orifice is crescentic and in the more sagittal plane between the mural attachment (arrows) and the septal aspect (arrow heads) of the mitral valve or component. The ventricular free wall except the short subaortic segment shows the outward bulging (open arrow). AO=aorta, LV=left ventricle, NC=noncoronary cusp of aortic valve. B. Systolic phase. The right margin of the LV between the aortic valve and the crux cordis is the coat septal aspect(s) of the mitral valve or component. A jet of regurgitant contrast medium (R) escapes from the left ventricle into the atrial cavity through the negative defect or cleft (arrow) at the center of the coat mitral valve or component. Elongated (Fig. 1, 3). The percentage of the inflow length (apex-crux cordis) to the outflow length (apex-aortic value) was between 58.8 and 78.6 (av. 73) in the conventional AP view and between 38.9 and 65.3 (av. 52.7) in the 4-chamber view. It was reported that it ranged from 93% to 100% (av. 98%) in the control study of 10 normal cardiac specimens.5

3. Deformity of the left ventricular outflow tract (LVOT); A tunnel of opacified blood was present between the mitral valve or mitral component of the common atroventricular valve and the left ventricular free wall in the conventional AP view. Its right wall was serrated septal attach-

Fig. 2. Levophase of pulmonary arteriogram in a normal child. The straight line between the noncoronary cusp (upper arrow) and the crux cordis (lower arrow) is the foreshortened profile of the atroventricular septum. LA=left atrium.
ment or septal aspect of the coapt atrioventricular AP view. Its right wall was serrated septal attachment or septal aspect of the coapt atrioventricular valves in the systolic phase (Fig. 1-B). In the diastolic phase, the tunnel of the LVOT was encroached upon from its right side by the smooth mural attachment of the mitral valve or component above and by the nonopacified blood entering the left ventricular cavity below (typical “goose-neck” deformity) (Fig. 1-A). The left wall of the left ventricle except the short subaortic segment was bulged outwardly. A narrow tunnel of the LVOT was also appreciated in the diastolic phase of the 4-chamber left ventriculogram, between the mural attachment and the left ventricular free wall (Fig. 3) although it was less impressive than was demonstrated in the conventional AP view, it was a constant finding.

4. Deformity and malorientation of the atrioventricular and was oriented in the more sagittal plane. The right margin of the crescentic left atrioventricular orifice was apically displaced septal aspect of the mitral valve or component on the deficient ventricular septal crest. It was serrated with a negative defect (cleft) at its center (Fig. 1). The left margin was smooth mural attachment. The catheter course in the lateral view was from above and in front, instead of normal course from above and behind. Whether the atrioventricular valve was partitioned or common could not be determined by the conventional views only. In the 4-chamber left ventriculogram, the mitral valve or component was well visualized along the deficient atrioventricular septum (Fig. 3). The diagnosis of the common atrioventricular valve could be made in 4 of 22 4-chamber left ventriculograms, by noticing the bridging leaflet after opacification of the right and left ventricles (Fig. 5, Table 2). The common leaflet was identified as a negative shadow during

---

**Fig. 3.** Left ventriculograms in 4-chamber view from a patient of AVSD with partitioned atrioventricular orifice. A. Diastolic phase. Loss of the straight line between the noncoronary cusp (NC) and the crux cordis (CC) indicates the absence of the AVS. Note the short posterior inlet septum (A-CC) and the wide distance between the aortic valve and crux cordis. The narrow tunnel of contrast medium (goose-neck) is formed between the ventricular free wall and the mural attachment of the mitral valve (open arrows). B. Systolic phase. Serrated margin between the aortic valve and the crux cordis is formed by the coapt anterior and posterior components of the septal leaflet of the mitral valve. The right atrial cavity is faintly opacified due to the regurgitation through the cleft (arrow head). No interventricular shunt is present.
the diastole. Further subclassification of the common atrioventricular valve could not be made. The diagnosis of the partitioned atrioventricular orifice could not be made by the 4-chamber left ventriculogram alone, because the failure of demonstration of the common valve and the

<table>
<thead>
<tr>
<th></th>
<th>IVS(+)</th>
<th>IVS(-)</th>
<th>IVS(?)</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>partitioned AV orifice</td>
<td>2</td>
<td>9</td>
<td></td>
<td>11</td>
</tr>
<tr>
<td>common AV orifice</td>
<td>4</td>
<td></td>
<td>3</td>
<td>7</td>
</tr>
<tr>
<td>undetermined</td>
<td>2</td>
<td>3</td>
<td>2</td>
<td>7</td>
</tr>
<tr>
<td>Total</td>
<td>8</td>
<td>12</td>
<td>2</td>
<td>22</td>
</tr>
</tbody>
</table>

Fig. 4. Left ventriculogram in elongated RAO view from a patient of AVSD. The absence of the normal AVS results in the concave defect extending from the noncoronary cusp (NC) to the crux cordis (CC).

Fig. 5. Left (A) and right (B) ventriculograms in 4-chamber view from a patient of AVSD with common atrioventricular orifice. The continuous lines (arrow heads in A) of the common leaflets extend across the ventricular septal crest. The interventricular shunt beneath the anterior common leaflet (AC) is related to the aortic valve and the shunt beneath the posterior common leaflet (PC) is related to the crux cordis. The interventricular shunt is reconfirmed by the 4-chamber right ventriculogram. SC is the stout filling defect of the upper margin of the ventricular septal crest.
absence of the interventricular shunt do not always indicate the partitioned atrioventricular orifice. Of 12 four-chamber right ventriculograms, the separate tricuspid valve was identified in nine and the common leaflet was reconfirmed in three. In one of 19 four-chamber left atriograms, the diagnosis of the separate mitral orifice could be made by noticing the round negative shadows of tricuspid and mitral valves at the time of atrial wash-out of contrast (Fig. 6). The differentiation of the partitioned and common atrioventricular orifice could not be made in seven patients.

5. Interatrial shunt: The interatrial shunt was visualized in 19 patients by the selective opacification of the left atrium in the 4-chamber view. The defect was between the apically displaced atrioventricular valves inferiorly and the atrial septum superiorly (Fig. 7). The septal aspects of the floors of the two atria (i.e., the atrioventricular valves) were continuous below the defect. There was no spatial interface between the left ventricle and the right atrium. Associated ostium secundum defect was identified in one patient.

6. Interventricular shunt and relation between the atrioventricular valves and the ventricular septum; In the conventional AP left ventriculogram, the only reliable evidence of the interventricular shunt was the immediate opacification of the right ventricular outflow tract and the pulmonary artery during the first ventricular systole after the injection, and was found in one patient. The presence of the interventricular shunt could not be evaluated by the conventional AP and lateral views, as the regurgitant contrast medium in the right atrium and ventricle could not be differentiated from the direct interventricular shunt. In the 4-chamber left ventriculogram, the presence of the interventricular shunt and the relation between the atrioventricular valves and the ventricular septum could be evaluated, as the ventricular septum was in profile. Interventricular shunt was present in eight equivocal in two and absent in 12 of 22 four-chamber left ventriculograms (Table 2). In one patient with interventricular shunt, the shunt was through the space beneath the anterior leaflet only and the posterior leaflet had the fibrous attachment on the ventricular septal crest. The diagnosis of chordal insertion of the atrioventricular valves on the septal crest could be made in two patients, by noticing the linear defect in the contrast medium beneath the atrioventricular valve leaflet. The interventricular shunt was reconfirmed by the 4-chamber right ventriculogram in three patients (Fig. 5). No new diagnosis of the interventricular shunt was made by right ventriculogram.

7. Atrioventricular regurgitation; In all patients, direct or indirect evidences of the regurgitation through the mitral valve or component were demonstrated (Fig. 1, 2, 8). The direction of the regurgitation was more accurately evaluated in the 4-chamber view. The regurgitation into the right atrium was predominant in 11 patients. The regurgitation into the left atrium was exacerbated by the effect of the antegrade catheter on the mitral valve or component.

8. Associated malformations; Left side superior
Fig. 7. A. Right upper pulmonary vein (RUPV) injection in 4-chamber view from a patient with secundum ASD. The left atrial floor (closed mitral valve, longer arrows) is a little higher than the right atrial one (closed tricuspid valve, shorter arrows), with a small gap of the intact AVS. ASD=secundum ASD. 
B. RUPV injection in 4-chamber view from a patient of AVSD. The septal aspects of the floors of the two atria are apically displaced and are continuous below the defect(D).

Fig. 8. Left ventriculogram in 4-chamber view from a patient of AVSD. A jet of contrast medium (arrow) is directed toward the left atrium (LA). Compare with Fig. 3-B.

Discussion

A generalized diagnosis of AVSD can be made by the conventional AP and lateral left ventriculograms because the deformity of the LVOT and the disproportion between the left ventricular inflow and outflow lengths are characteristic.4,6,11-16 But the ability of the conventional views in evaluating the variable features of surgical importance is far from satisfaction. Recent development of axial views has permitted increased accuracy in defining the more important surgical features in addition to the common basic morphology.7-10 The steps in preoperative angiographic evaluation of the AVSD's should
include;

1) identification of the common features that are essential in making the diagnosis of AVSD.

2) subclassification of the AVSD's into the partial (with partitioned orifice) and complete (with common orifice) types,

3) evaluation of the variable features that are of surgical importance, i.e., the interatrial shunt, the interventricular, the relation between the atrioventricular valves and the atrial and ventricular septa and the function of the atrioventricular valves,

4) identification of the associated anomalies.

Common Angiographic Features:4-6,11-17 Common angiographic features are deformed or deficient AVS with wide separation of the aortic valve and crux cordis, disproportion between the lengths of the inflow and outflow tracts, deformity of the LVOT and deformity and malorientation of the atrioventricular valves. These pathognomonic findings of the AVSD's are readily appreciated in the conventional AP, 4-chamber and elongated RAO views.

Partitioned or common atrioventricular orifice; The diagnosis of the common atrioventricular orifice can be obtained from the 4-chamber left ventriculogram after the opacification of the right ventricle.17 The negative shadow of the common atrioventricular valve extends across the ventricular septum during the diastole. The anterior common leaflet is related to the aortic valve and the posterior leaflet to the crux cordis. It is reported that right and left anterior leaflet can be identified and the subclassification into the Rastelli's subtypes are possible.17 But we could not make the differentiation. The absence of the interventricular shunt usually indicates the presence of partitioned orifice, as the majority of the partitioned atrioventricular valves have fibrous attachments on the ventricular septal crests.2,17 But the failure to demonstrate the common atrioventricular valve and the absence of the interventricular shunt dose not always mean the partitioned orifice, because there are a few cases in the literatures in which the common atrioventricular valves are attached to the ventricular septal crest and no true interventricular shunt is present.1 The 4-chamber right ventriculogram provides the additional clues in differentiating the partitioned orifice from the common orifice in the large number of cases.

Interventricular shunt; In the normal heart, the septal aspect of the left atrial floor (anterior leaflet of the mitral valve) is a little higher than that of the right atrium (septal leaflet of the tricuspid valve), with a small gap of AVS between the left ventricle and the right atrium (Fig. 7-A).1,16 In the AVSD's, the atrioventricular valves are displaced apically on the ventricular septal crest and the floors of the two atria are at the same level (Fig. 7-B). The shunt is through the deficient AVS converted into the interatrial communication.1 The shunt between the atrial septum and the atrioventricular valves is best demonstrated in the 4-chamber right upper pulmonary vein injection. A few cases are reported in the literature in which the atrioventricular valves are attached to the lower margin of the atrial septum and there is no interatrial communication.2 The 4-chamber right upper pulmonary injection is mandatory for the evaluation of the size of the defect and the associated defects of the interatrial septum.17

Interventricular shunt and relation between the atrioventricular valves and the ventricular septum; Four possibilities of relation between the atrioventricular valves and the ventricular septum exist; 1) free-floating leaflets with large interventricular shunt, 2) leaflets of chordal insertion on the ventricular septal crest with small interventricular shunt, 3) leaflets related to the atrial septum with no or small interatrial communication.1,2,9,10,17 The 4-chamber left ventriculogram is the best method in the evaluation of the interventricular shunt. The shunt beneath the anterior component of the atrioventricular valve is related to the aortic valve and the shunt beneath the posterior component to the crux cordis. The angiographic distinction of the shunt beneath the free-floating leaflet from the shunt beneath the tethered always can be made.17 But the filling defects
in the contrast medium passing from one ventricle to the other is the reliable evidence of the chordal insertion.\textsuperscript{17}

Function of the atrioventricular valves; The presence of regurgitation into the atrial chamber may be demonstrated in the elongated RAO, view of conventional AP and lateral views. But the direction of the regurgitation into the right or left atrium can be evaluated more accurately by the 4-chamber view. The negative defect (cleft) of the septal aspect of the mitral valve or component in the protosystole is more commonly into the right atrium.\textsuperscript{17} For the more accurate evaluation of the regurgitation, the 4-chamber left ventriculogram should be made by the retrograde through the aorta.

Summary

The cineangiograms of 31 patients with an atrioventricular septal defects (AVSD's) were reviewed to correlate the angiographic findings demonstrated in the axial views with those demonstrated in the conventional views and to provide the essential injections and projections in the preoperative evaluation of the AVSD's. It is concluded that the axial views provide the more accurate features of surgical importance as well as the common pathologic features than are demonstrated from the conventional frontal and lateral views and that the axial views should replace the conventional frontal and lateral views. The injections and projections in the preoperative evaluation of the AVSD's should include 1) 4-chamber left ventriculography, through the retrograde catheter, for the evaluation of the interventricular shunt, the variation of the morphology of the atrioventricular valves and the atrioventricular regurgitation, 2) 4-chamber right ventriculography, for the better evaluation of the variation of the morphology of the atrioventricular valves, and 3) 4-chamber right upper pulmonary vein injection, for the evaluation of the presence or absence, location and size of the interatrial shunt.

References