Cineangiography of the Perimembranous Ventricular Septal Defect With Left Ventricular-Right Atrial Shunt

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Nine cases are reported of perimembranous ventricular septal defect associated with left ventricular to right atrial shunting. Cineangiographic findings included an aneurysm of the membranous septum in all patients; two patients had obvious adherence of deformed tricuspid valve leaflets to the membranous septum. The location of the ventricular septal defect was confirmed at surgery or cardiac endoscopy in seven patients. In the presence of a perimembranous ventricular septal defect, left ventricular to right atrial shunting is usually the result of tricuspid valve abnormalities, including clefts or perforations of the septal leaflet, deformity or adherence of valve tissue to the margins of the septal defect and widening of the anteroseptal commissure. Biplane left ventriculography, using the long axial oblique and reciprocal right anterior oblique projections, may best demonstrate the pathologic anatomy, although the hepatoclavicular projection is a useful alternative, particularly when an atrioventricular canal defect is a diagnostic consideration.

The anatomic basis for left ventricular to right atrial shunting in the presence of a perimembranous ventricular septal defect has been described previously (1–4). Briefly, the left ventricular-right atrial shunt is caused by an anomaly or deformity of the tricuspid valve leaflets adjacent to the defect in the membranous septum. We undertook this study to determine the cineangiographic features of this abnormality and the optimal projections for demonstrating the pathologic anatomy.

Methods

Study patients. Nine patients with a perimembranous ventricular septal defect and left ventricular-right atrial shunting underwent cardiac catheterization and cineangiography between January 1973 and December 1981 at Children's Hospital Medical Center, Boston. The patients' ages ranged from 9 months to 14.6 years (mean 6.9 years). We examined the cardiac catheterization data, cineangiograms, medical records and electrocardiograms of all patients. In addition, videotaped two-dimensional echocardiograms of four patients and the chest radiographs of eight patients were reviewed. One patient who was referred from another institution did not have a preoperative chest radiograph at our center. The left ventricular-right atrial shunt was diagnosed angiographically when contrast material injected into the left ventricle entered the right atrium before, or at the same time as it entered the right ventricle, and physiologically by an atrial left to right shunt in the absence of an atrial septal defect. Patients with atrioventricular (AV) canal, ventricular septal defect of the AV canal type or obvious tricuspid valve regurgitation were excluded from this study. The presence of contrast material in the right atrium during ventricular arrhythmia was not considered to represent left ventricular-right atrial shunting.

Angiography. Cineangiograms included left ventricular injections in all patients. Six patients studied after 1977 had biplane left ventriculography in the right anterior oblique and long axial oblique (70° left anterior oblique with 20° cranial angulation) projections. The series also included left ventriculograms in the hepatoclavicular (40° left anterior oblique, 40° cranial angulation) projection (two patients), 40° cranially angled frontal projection (one patient) and right and left anterior oblique projections without axial angulation (one patient).

Six children had confirmation of the location of the ventricular septal defect during surgical repair. An additional patient underwent fiberoptic endoscopic examination of the heart in 1975.

Findings

Catheterization findings. Eight of the nine patients had a left to right shunt with pulmonary/systemic flow ratios (Qp:Qs) ranging from 1.5 to 3.5 (mean = 2.0). The other child had subinfundibular and valvular pulmonary stenosis

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in addition to a perimembranous ventricular septal defect and had a net right to left shunt (Qp:Qs = 0.8). Pulmonary artery pressures were normal in eight of the nine patients and elevated (85/10 mm Hg) in one patient who developed left ventricular-right atrial shunting after patch closure of a large perimembranous ventricular septal defect.

Left ventriculographic findings. Left ventriculography demonstrated a perimembranous ventricular septal defect with aneurysm of the membranous septum in all patients. The lateral projection and the left anterior oblique projection without axial angulation were the least useful views for demonstration of these abnormalities. Posteroanterior and right anterior oblique projections confirmed the left ventricular-right atrial shunt and usually demonstrated a jet of contrast material that reached the right atrium before, or simultaneously with, the right ventricle (Fig. 1). The long axial oblique projection best demonstrated the location of the ventricular septal defect, the aneurysm of the membranous septum and the relation of these abnormalities to the tricuspid valve leaflets (Fig. 2). The hepatoclavicular projection adequately demonstrated the ventricular septal defect and aneurysm of the membranous septum, and also showed the left ventricular-right atrial shunt clearly (Fig. 3). The tricuspid valve leaflets appeared to be deformed and adherent to the margins of the ventricular septal defect in two patients. This was confirmed in one patient by a right ventricular injection that demonstrated a pouch-like deformity of the septal leaflet of the tricuspid valve (Fig. 4).

Additional angiographic abnormalities were identified in four of the nine patients and included persistent left superior vena cava, mitral regurgitation, valvular aortic and pulmonary stenosis and anomalous muscle bundles in the right ventricle.

Echocardiographic findings. Two-dimensional echocardiography in four patients demonstrated a perimembranous ventricular septal defect in all four patients and, in addition, an aneurysm of the membranous septum in two. The patient who developed left ventricular-right atrial shunting after surgical closure of the perimembranous ventricular septal defect had echocardiographic demonstration of detachment of the superior margin of the patch. There were no specific echocardiographic features that predicted left ventricular-right atrial shunting in any of the four patients.

Radiographic findings. Chest radiographic findings were similar to those previously described (3–5); they included increased pulmonary blood flow, globular cardiomegaly and a particularly prominent right atrial border in the majority of patients. In addition to right atrial enlargement, the left atrium appeared large in half of the patients.

Electrocardiographic findings. Analysis of the electrocardiograms also revealed findings similar to those previously reported (6). In three of the nine patients, the frontal plane QRS rotation was counterclockwise and superior ($-15°$, $-15°$, $-75°$).

Surgical findings. The location of the perimembranous ventricular septal defect was confirmed at surgery in all six patients who underwent operation, and by cardiac endoscopy in one patient. A fibrous rim or true aneurysm was associated with the margin of the ventricular septal defect in three patients. The surgeons noted that the ventricular

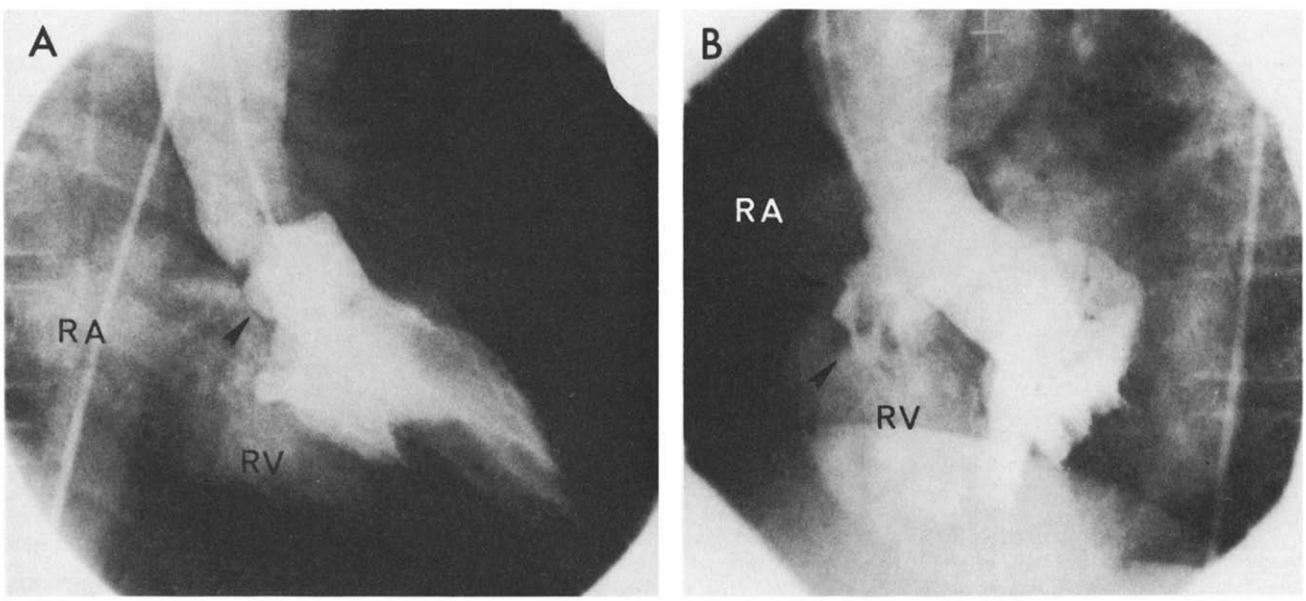

Figure 1. Biplane left ventriculogram demonstrating perimembranous ventricular septal defect, aneurysm of the membranous septum (**arrowheads**) and left ventricular-right atrial shunt. **A,** Right anterior oblique projection shows a jet of contrast medium from the left ventricle into the right atrium (RA). Note proximity of the aneurysm of the membranous septum to the tricuspid valve. **B,** Long axial oblique projection. A small amount of contrast medium can be identified in the right atrium. RV = right ventricle.

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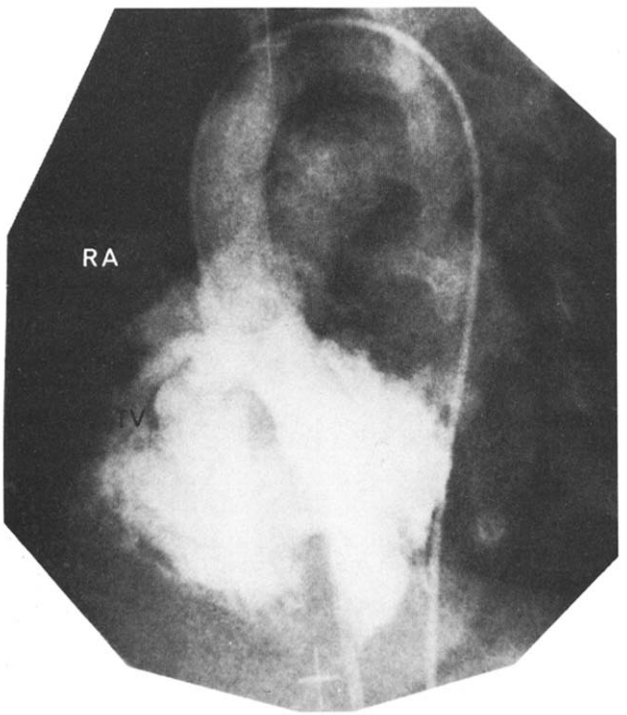


Figure 2. Left ventriculogram in long axial oblique projection. Tricuspid valve (TV) tissue is demonstrated adjacent to the perimembranous ventricular septal defect. A small amount of contrast medium is present in the right atrium (RA)

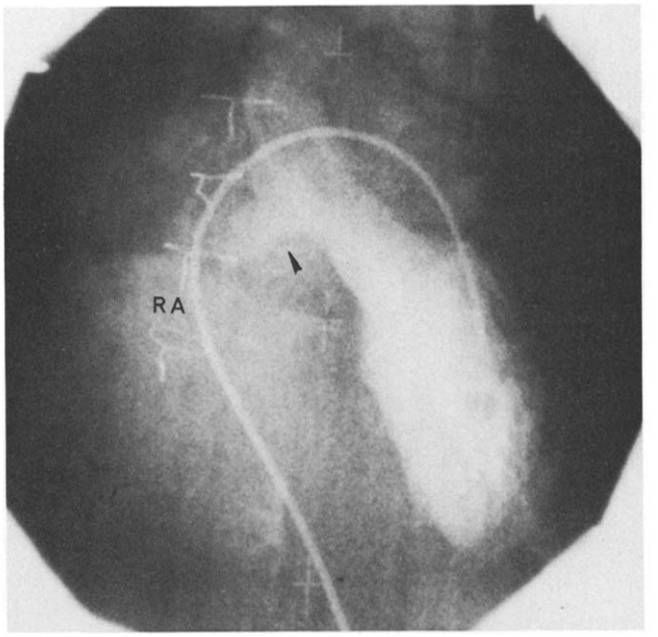


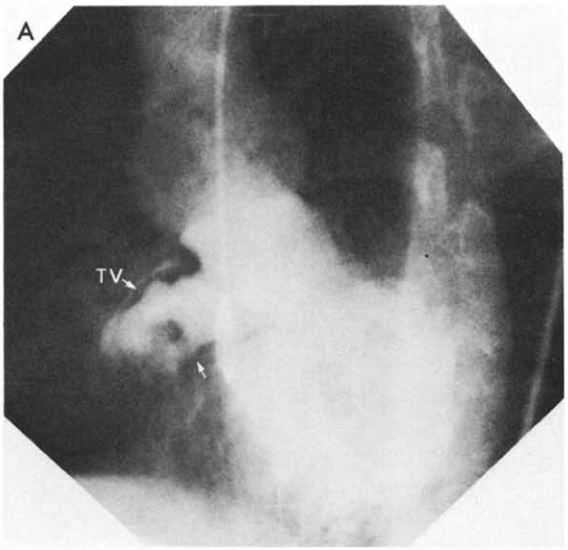
Figure 3. Left ventriculogram in the hepatoclavicular projection in a patient with left ventricular-right atrial shunt after surgical closure of a perimembranous ventricular septal defect. The free margin of the patch (**arrowhead**) and contrast medium in the right atrium (RA) are demonstrated.

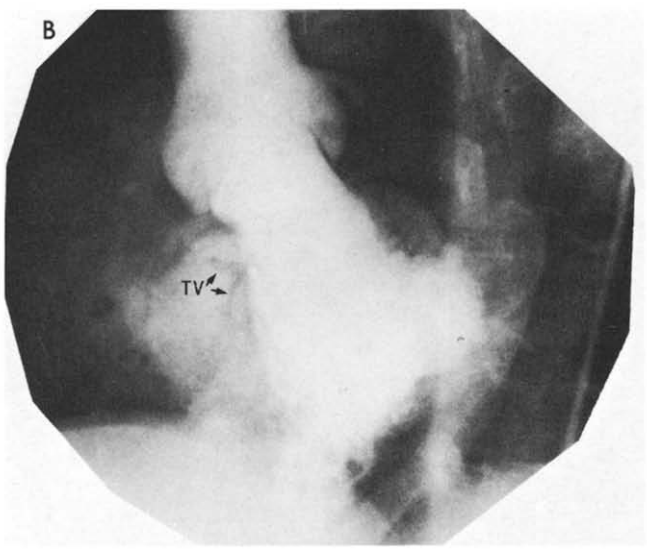
septal defect was adjacent to a widened posteromedial tricuspid valve commissure in one patient and adherent to a pouch-like deformity of the septal leaflet (Fig. 4) in another. The tricuspid valve was not described in the operative reports of the four other patients. At endoscopic examination of one patient, the septal leaflet of the tricuspid valve was noted to have a rolled edge and bright red blood was seen to enter the right atrium through the deformity in the valve leaflet.

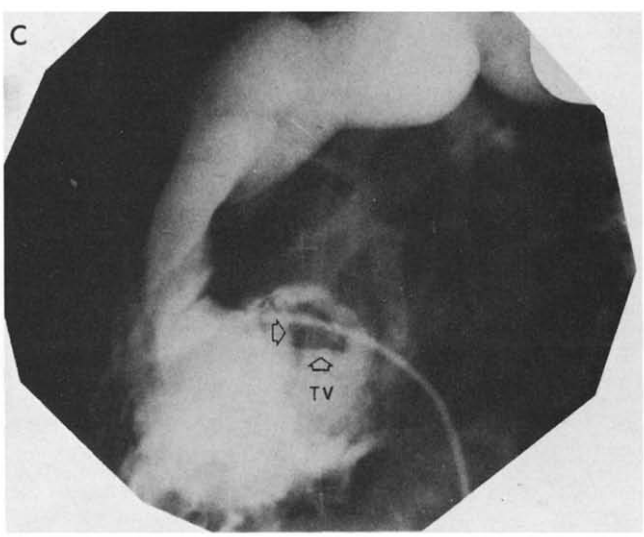
Discussion

Left ventricular-right atrial shunt: role of tricuspid valve anomalies. Left ventricular-right atrial shunting is a feature of several cardiac anomalies, including atrioventricular (AV) canal and isolated defects of the septum between the left ventricle and right atrium (1). This report describes the most common type of left ventricular-right atrial shunt, that associated with the combination of perimembranous ventricular septal defect and anomalies of the tricuspid valve (1–4). The valvular abnormalities most frequently implicated (Fig. 5) include perforations or clefts of the septal leaflet (3), widening of the anteromedial commissure (3,4), thickening and distortion of the septal or anterior leaflets secondary to hemodynamic trauma (2,4) and adherence of the tricuspid leaflets to the margins of the ventricular septal defect (1,3). Incomplete attachment of the tricuspid valve to the membranous ventricular septum at the anteromedial commissure has been found in 39% of normal hearts and may contribute to left ventricular-right atrial shunting in the presence of a perimembranous ventricular septal defect, with or without additional tricuspid valve abnormalities (3,7).

Aneurysm of membranous septum. A feature that was observed on left ventriculography in all of our patients with left ventricular-right atrial shunting and that has been noted at postmortem and surgical inspection by others (4,8) is the presence of an aneurysm of the membranous septum. In cardiac angiography, the term aneurysm of the membranous septum refers to alteration of the margins of a perimembranous ventricular septal defect producing a structure with definite but often irregular margins that protrude into the right ventricle during systole (9,10). The term describes a radiologic finding; a true aneurysm may or may not be found at surgery or postmortem examination (10). Anatomically, a true aneurysm of the membranous septum is a fibrous walled sac that usually has an opening at its apex and that protrudes into the right ventricle under the septal leaflet of the tricuspid valve (8,11). Uncommonly, true aneurysms may herniate through the tricuspid valve during systole, leading to tricuspid regurgitation or left ventricular-right atrial shunting, or both (8). Alternatively, the angiographic finding of an aneurysm may correspond to ballooning of the tricuspid septal leaflet caused by the jet through the ven-







tricular septal defect, or to adherence of valve tissue to the margins of the defect (4,9–11). The latter probably occurs commonly in patients with perimembranous ventricular septal defect and is thought to play an important role in the spontaneous closure of the defect (9). A third cause of the angiographic demonstration of an aneurysm of the membranous septum is a pouch-like malformation of the tricuspid valve caused by the presence of redundant valve tissue, often with abnormal chordal attachments to the membranous septum (8). Because the underlying pathologic anatomy in the left ventricular-right atrial communication includes the combination of a perimembranous ventricular septal defect and tricuspid valve abnormalities, the high incidence of aneurysm of the membranous septum on left ventriculography in our series is not surprising.

Differentiation from atrioventricular canal defects. Left ventricular-right atrial shunting associated with perimembranous ventricular septal defect is easily distinguished angiographically from that associated with AV canal defects. In the latter, left ventricular-right atrial shunting results from regurgitation from the left ventricle through a cleft in the mitral valve into the right atrium through the primum atrial septal defect. Left ventriculography demonstrates deficiency of the posterior or basilar part of the interventricular septum, a cleft and abnormally attached anterior leaflet of the mitral valve and a divided or common superior AV valve leaflet (12), none of which is present in left ventricular-right atrial shunting with a perimembranous ventricular septal defect. The hepatoclavicular projection is the most useful view for demonstration of these abnormalities, as it separates the four cardiac chambers and places the posterior or basilar portion of the interventricular septum in profile (Fig. 6) (12–15).

Angiographic considerations. The long axial oblique projection places the anterior interventricular septum in profile and demonstrates defects as well as aneurysms of the membranous septum to the best advantage (12–15). The course of the right coronary artery and the radiolucency produced by the fat in the AV groove provide an estimation of the plane of the tricuspid valve anulus, and are useful in identifying the left ventricular-right atrial shunt in this projection. If biplane cineangiography is used, the reciprocal right anterior oblique projection nicely demonstrates the jet

Figure 4. Perimembranous ventricular septal defect with left ventricular-right atrial shunt and adherence of tricuspid valve tissue to the margins of the ventricular septal defect. **A** and **B,** Left ventriculogram, long axial oblique projection. Deformity of the tricuspid valve (TV) (**arrows**) adherent to the membranous septum is seen in two phases of the cardiac cycle (**A,** diastole; **B,** systole). The left ventricular-right atrial shunt is not well seen in these frames. **C,** Right ventriculogram, lateral projection, showing pouch-like deformity of the tricuspid valve (**arrowheads**), caused by its adherence to the margins of the ventricular septal defect. (Additional abnormalities include anomalous muscle bundles in the right ventricle and valvular pulmonary stenosis.)

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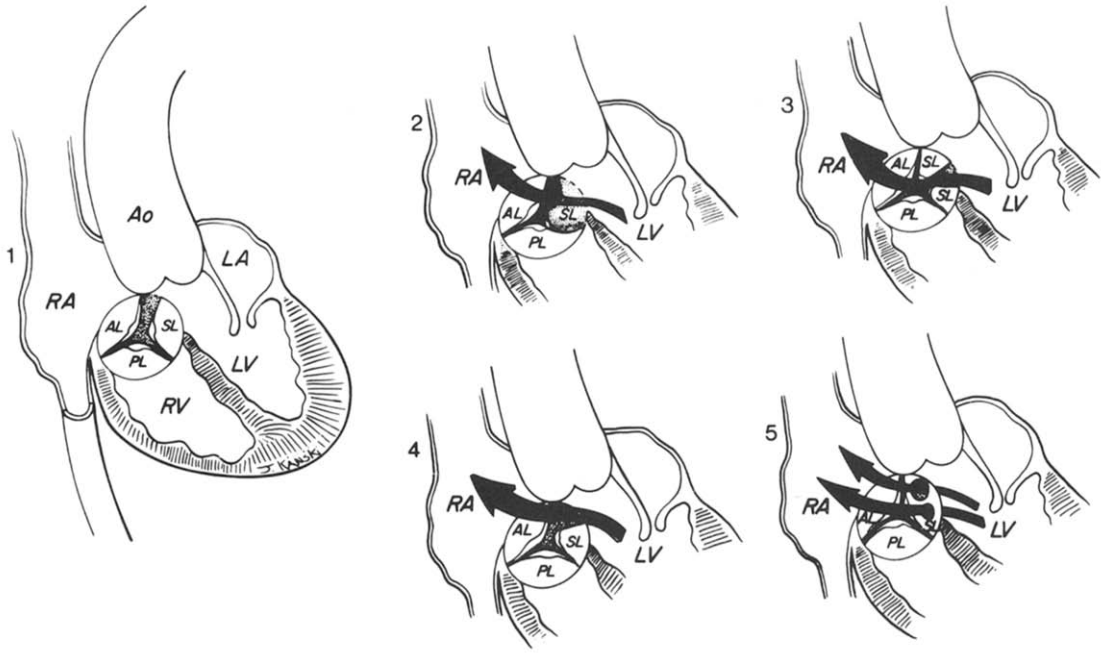


Figure 5. Diagrammatic illustration of the anomalies of the tricuspid valve associated with left ventricular-right atrial shunting in patients with perimembranous ventricular septal defect. **1**, Location of perimembranous ventricular septal defect adjacent to the septal leaflet (SL) of the tricuspid valve. **2**, Adherence of septal leaflet to the margins of the ventricular septal defect, producing an "aneurysm." **3**, Cleft septal leaflet. **4**, Widened anteromedial commissure. **5**, Perforations in the septal leaflet. Ao = aorta; AL = anterior leaflet of tricuspid valve; LA = left atrium; LV = left ventricle; PL = posterior leaflet; RV = right ventricle.

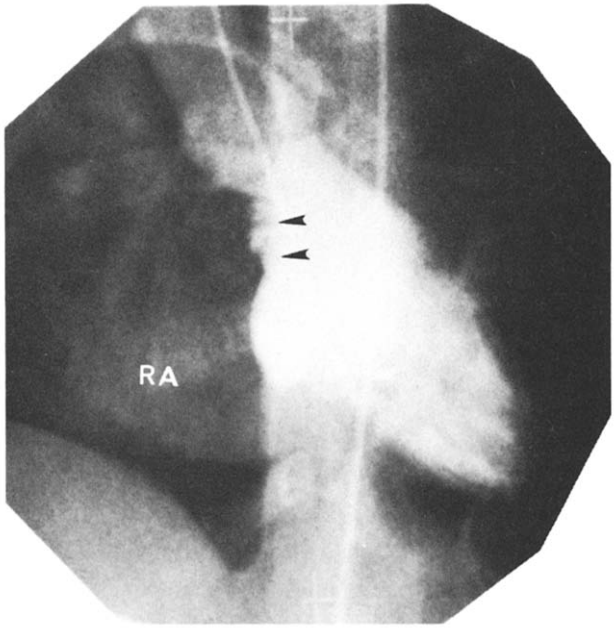
of contrast medium from the left ventricle to the right atrium (Fig. 1).

Although it does not profile the anterior interventricular septum, the hepatoclavicular projection is the best single view to demonstrate the combination of perimembranous ventricular septal defect, aneurysm of the membranous septum and left ventricular-right atrial shunt. We believe that the long axial oblique projection better demonstrates the pathologic anatomy of this anomaly, but the hepatoclavicular view is a useful alternative, especially if the diagnosis of an AV canal defect is in question.

Echocardiography. In our experience, two-dimensional echocardiography accurately demonstrated the location of the ventricular septal defect, but did not predict the presence of left ventricular-right atrial shunting. Saline contrast echocardiography was not performed in our patients and should be useful in the detection of the left ventricular-right atrial shunt. Others (16) have reported the presence of high frequency, low amplitude systolic flutter of the tricuspid valve on M-mode echocardiography in the presence of shunting.

Clinical diagnosis. The clinical data of the patient with a left ventricular-right atrial shunt may be confusing. Physical examination reveals a harsh systolic murmur and a thrill at the lower left sternal border, suggesting the presence of a ventricular septal defect (2,5). The second heart sound may be widely split, but varies normally with respiration (2,5). The chest radiograph typically demonstrates a globular cardiac configuration with a prominent right atrial border similar to the findings seen in the presence of a common AV canal with AV valve regurgitation (4). Cardiac catheterization data demonstrate an increase in oxygen saturation

Figure 6. Left ventriculogram in the hepatoclavicular projection in a patient with an atrioventricular canal defect (primum atrial septal defect). Abnormal attachments of the mitral valve (**arrowheads**), deficiency of the basilar part of the ventricular septum and contrast material in the right atrium (RA) constitute the abnormal findings.



at the atrial level, often without a further rise at the ventricular level (2,5). Therefore, angiography is the most definitive investigative procedure in patients with left ventricular-right atrial shunt. With the appropriate projections this anomaly can be readily distinguished from the other diagnostic possibilities, that is, complete common AV canal and primum atrial septal defect with mitral regurgitation.

The anatomic basis for the left ventricular-right atrial shunt is useful preoperative information. Surgical repair of this defect by a right atriotomy has been performed successfully since 1957 (17,18).

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