Complete Atrioventricular Canal with Survival to the Eighth Decade*

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Survival to the eighth decade of patients with atrioventricular canal is extremely rare. A patient is presented with such survival of the complete form of the defect. This possibly represents the first such report in the medical literature. The value of 2-dimensional echocardiography, particularly with venous contrast studies, in establishing the diagnosis, is shown.

Survival of patients with atrioventricular canal beyond the seventh decade of life is extremely rare. Hynes et al described six such patients and commented that the total number of patients reported surviving into the eighth decade was five. These five patients had the partial form of the disorder. We report a patient with the complete form of the defect, proved by 2D echocardiography with venous contrast studies. To the best of our knowledge, this is the first such patient reported with survival to the eighth decade.

CASE REPORT

This 73-year-old man was seen in consultation to assess his fitness for prostatectomy. He had known of heart disease since adolescence, which was diagnosed as rheumatic valvular disease. In recent years, two cardiologists had suspected the lesion as being congenital. He had no symptom of cardiac disease and was leading a normal life, but he had had one attack of paroxysmal atrial tachycardia in 1971. Physical examination revealed normal pulse rate. Blood pressure was 160/90 mm Hg. There was no evidence of congestive heart failure. There was clinical evidence of right ventricular enlargement. Cardiac auscultation revealed a grade 2/6 ejection systolic murmur over the pulmonary area. There was wide, fixed splitting of the second heart sound. A short, scratchy, mid-diastolic murmur at the lower left sternal border was regarded as a tricuspid flow murmur. At the apex, there was a grade 3/6 holosystolic murmur suggestive of mitral incompetence.

His electrocardiogram showed right bundle branch block with left axis deviation (mean frontal plane axis minus 90°; axis of first 0.06 second minus 45°).

M-mode echocardiography (Fig 1) showed a dilated right ventricle and paradoxic movement of the interventricular septum. The

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FIGURE 1. Composite M-mode echocardiogram of the aortic, pulmonic and mitral valves. The aortic valve shows early, abrupt, partial systolic closure (arrow) followed by fine flutter. The pulmonic valve shows a large "A" dip (arrow) excluding pulmonary hypertension and suggesting a large left-to-right shunt. At the ventricular level, the right ventricle is dilated, septal motion is paradox and the anterior mitral valve leaflet moves into the septum. Ao = aorta; MV = mitral valve; PA = pulmonary artery; RV = right ventricle; S = interventricular septum.

FIGURE 2. Two-dimensional echocardiogram (apical four-chamber view), showing a defect in the upper part of the interventricular septum and the lower part of the interatrial septum (between upper and lower arrows). In this picture, in systole, the tricuspid and mitral valve tips (?) are seen crossing each other. LV = left ventricle; MV = mitral valve; RA = right atrium; RV = right ventricle; S = interventricular septum; TV = tricuspid valve.
anterior leaflet of the mitral valve moved into the interventricular septum during early diastole. There was partial closure of the aortic valve in early systole with fine flutter of the leaflets. Pulmonary valve movement was normal, with no features of pulmonic hypertension.

Two-dimensional echocardiography performed with an apical four-chamber view (Fig 2) showed a defect in the lower part of the interatrial septum overlying a defect in the upper part of the interventricular septum. Separate mitral and tricuspid components of the anterior common leaflet were present, with probable chordae appearing to cross over just above the interventricular septum.

Venous contrast angiography (bubbled saline) showed positive contrast shadowing between the right heart chambers and left ventricle in early diastole (Fig 3) and negative contrast shadowing between the left atrium and right ventricle about 60 milliseconds later (Fig 4). A deficiency in the upper part of the interatrial septum was thought to represent a possible separate secundum defect, as negative contrast shadowing was seen in the right atrium (Fig 3 and 4).

**DISCUSSION**

The M-mode echocardiographic findings of paradoxic interventricular septal movement with apparent opening of the anterior mitral leaflet into the interventricular septum in early diastole, coupled with the electrocardiographic features of right bundle branch block and left anterior hemiblock, enabled us to make a diagnosis of atrioventricular canal. The 2-dimensional finding of a defect of the lower interatrial septum overlying a defect of the upper interventricular septum, coupled with contrast echocardiographic evidence of direct shunting from left atrium to right ventricle (negative contrast effect, Fig 4) and of right-to-left shunting from right atrium to left ventricle, during diastole, (positive contrast effect, Fig 3) indicated the complete form of atrioventricular canal.

There was an apparent cross-over of chordal attachments from the mitral and tricuspid components of the anterior common leaflet. It was felt that this was probably related to the chordae attaching to the upper part of interventricular septum (type A), although such attachment could not be directly demonstrated. On M-mode echocardiography, there was partial closure of the aortic valve in early systole. This finding has been reported in patients with subaortic membrane, but no evidence of such a defect was found. It is possible that deformity of the left ventricular outflow tract, which causes the swan-neck appearance on angiography, could explain this finding.

**Clinical Implications**

This case adds further evidence that occasional prolonged survival is possible in patients with atrioventricular canal, even the complete form. Echocardiography, especially the 2D mode, with the additional application of venous contrast angiography, enables the diagnosis to be made by noninvasive means.

**REFERENCES**