Unusual Longevity in Persistent Common Atrioventricular Canal

By Rajendra Tandon, M.D., James H. Moller, M.D., and Jesse E. Edwards, M.D.

SUMMARY

Five of 139 patients with persistent common atrioventricular canal (AVC) confirmed at necropsy had lived to the age of 46 years or older. The complete type of the malformation was present in one, the transitional type in one and the partial type in three. The oldest of the latter group was 79 years of age at the time of death.

In a study of ages at death among 139 cases of AVC the following observations were made. Of the 91 patients with the complete type of AVC, 37 percent survived more than one year and 17.6 percent more than five years. Of the 48 patients with the transitional and partial types of the defect, 67 percent lived more than one year and 37.5 percent beyond five years.

One of our cases was highly unusual in that AVC was associated with corrected transposition.

Additional Indexing Words:
Endocardial cushion defect Ostium primum atrial septal defect Corrected transposition of great vessels
Left-to-right shunts Prognosis in congenital heart disease

Persistent common atrioventricular canal (AVC) is a complicated anomaly which involves the atrial and ventricular septa and the atrioventricular valves. The condition is commonly associated with death in infancy. In only exceptional cases does the patient survive to adult life. This communication describes five patients with this malformation who lived from 46 to 79 years.

It is commonly accepted that three anatomic types of AVC may be recognized. These are the complete, transitional and partial types. In each there is a defect in the lowermost part of the atrial septum representing a persistent ostium primum. The ventricular septum is deficient in the subaortic area. Involvement of the valves varies and forms the basis for subdivision of AVC into three types.

The specimens were classified according to the nomenclature of Wakai and Edwards into three forms: complete, transitional and partial (fig. 1). The complete type is characterized by cleft anterior mitral and septal tricuspid valve leaflets, forming a large anterior and a smaller posterior leaflet of a valve common to both sides of the heart. The ventricular septum is deficient and a large interventricular communication is usually present.

In the transitional type, the anterior mitral and septal tricuspid leaflets are each cleft but a bridge of tissue extends between the anterior and the posterior leaflets dividing the atrioventricular canal into two halves. The partial type has a cleft in one of the atrioventricular valves, usually the mitral. While an interventricular communication is usually present in the complete type, it may be present or absent in the transitional type. Usually the partial type is not associated with an interventricular communication in spite of deficiency in ventricular tissue.

An uncommon forme fruste of AVC is that in which only the characteristic atrial septal defect is present while the atrioventricular valve and ventricular septum are normal.

Description of Cases

Of the five cases to be described from subjects 46 years of age and older, in one the AVC was of the complete type, in one of the transitional type and in three of the partial type.

Case 1 (Complete Type of AVC)

The patient was a 46-year-old woman who, since
childhood, suffered from dyspnea and cyanosis. Physical examination showed normal first and second cardiac sounds without murmurs. The electrocardiogram showed right axis deviation without evidence for hypertrophy of either ventricle. The vectorcardiogram was read as within normal limits. Thoracic roentgenograms showed marked scoliosis and increased pulmonary vasculature. Because of marked scoliosis, she was considered to represent an example of cor pulmonale until cardiac catheterization data were obtained at the age of 45 years. At that time, a left-to-right shunt at the atrial level with systemic pressures in the right ventricle were found. The site of a coexisting right-to-left shunt was not specifically defined. She died at 46 years of age while angiocardiography was being performed. These studies in-

Figure 1

a. Diagrammatic portrayal of a normal heart showing the relationship of the atrial and ventricular septa and the tricuspid and mitral valves as a reference for the diagrams to follow. b. Complete type of AVC. The atrial and ventricular septa are deficient allowing left-to-right shunts. The anterior mitral and septal tricuspid valve leaflets are cleft. The adjoining parts are continuous forming a valve common to both sides of the heart. c. Transitional type. The atrial septum is deficient inferior to the fossa ovalis. The anterior mitral and septal tricuspid leaflets are cleft but a bridge of tissue divides the atrioventricular orifice into mitral and tricuspid components. d. Partial type. The atrial septum is deficient inferior to fossa ovalis. The anterior mitral leaflet is cleft while the tricuspid valve is not cleft.
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Figure 2
Case 1. Corrected transposition and the complete type of AVC. a. External view of the heart and transposed great vessels. The aorta (A.) arises anterior and to the left of the pulmonary trunk (P.T.). b. Interior of the right atrium (R.A.) and anatomic left ventricle (L.V.). Anterior leaflet (A.L.) and posterior leaflet (P.L.) of the common atrioventricular valve below persistent ostium primum. The upper edge of the latter (single arrow) forms the lowermost aspect of atrial septum. Deficiency of ventricular septum (between paired arrows). c. Aorta (A.) arising from the infundibulum (I.) of the anatomic right ventricle (R.V.) that is inverted.

The essential necropsy findings were those of corrected transposition associated with the complete type of AVC (fig. 2). The upper ventricular septum was markedly deficient and a ventricular septal defect was present. The morphological right ventricle was hypoplastic. An atrial septal defect of the fossa ovalis type was also present and the aortic valve was bicuspid (fig. 3).

Case 2 (Transitional Type of AVC)
In a woman who died at the age of 65 years the clinical history available to us was in summary form, indicating that the results of physical examination and electrocardiography were consistent with a diagnosis of AVC.

Figure 3
Case 1. Diagrammatic portrayal of the anomaly. The right-sided ventricle ("R.V.") is morphologically the left ventricle and the left-sided ventricle ("L.V.") is morphologically the right ventricle. The transposed aorta (A.) arises anterior and to the left of the pulmonary trunk. The complete type of common AVC is shown posterior to the roots of the great vessels. The atrial and ventricular septa are deficient allowing left-to-right shunts. S.V.C. = superior vena cava; I.V.C. = inferior vena cava; R.A. = right atrium; L.A. = left atrium; R.P.A. = right pulmonary artery; L.P.A. = left pulmonary artery; L.P.V. = left pulmonary vein.
Pathologic examination of the heart showed the transitional type of AVC in which the septal leaflet of the tricuspid valve was thickened and showed a small cleft. The mitral valve showed a more prominent cleft.

The ventricular septum showed a deficiency characterized of AVC but there was no interventricular communication as the anterior mitral leaflet was adherent to the superior rim of the ventricular septum (fig. 4).

Cases 3-5 (Partial Type of AVC)

The oldest patient in this series was a man who died at the age of 79 (case 3). In this patient a cardiac murmur had been known to be present throughout his life. His first symptom occurred at the age of 64 years and was characterized by thoracic pain diagnosed as a “heart attack.”

Although he denied paroxysmal nocturnal dyspnea or orthopnea, physical examination showed signs of congestive cardiac failure and auscultatory findings of an atrial septal defect. The electrocardiogram showed atrial fibrillation and antero-septal infarction with right bundle branch block. The mean QRS axis of the unblocked portion of the QRS complex was +75 degrees (fig. 5). A thoracic roentgenogram showed a large heart with right ventricular enlargement. The main pulmonary arterial segment was enlarged and the vasculature increased. There was also right pleural effusion (fig. 6). He died in a state of intractable congestive cardiac failure.

The next case (case 4) was that of a man who died at the age of 48 years. When a child, he was considered to have rheumatic heart disease. At the age of 27 years, a diagnosis of atrial septal defect was made but the patient remained asymptomatic until the age of 37 years, when, following a number of episodes of syncope, he was found to have atrial fibrillation. This was converted to sinus rhythm with quinidine therapy.

The electrocardiogram at the age of 39 years showed a frontal plane QRS axis of −60 degrees. There was first degree atrioventricular block with right ventricular hypertrophy. Thoracic roentgeno-

Figure 4

Case 2. Transitional type of AVC. a. Interior of the right atrium (R.A.) and right ventricle (R.V.). The defect (D.) in the atrial septum lies below the fossa ovalis (F.O.) and is a persistent ostium primum. The septal tricuspid leaflet is cleft (arrow) and the right ventricle dilated. b. Interior of the left atrium (L.A.) and left ventricle (L.V.) showing the persistent ostium primum (D.) and a cleft in the anterior leaflet of the mitral valve (arrow). c. Outflow tract of the left ventricle (L.V.) showing attachment of the anterior leaflet of the mitral valve (A.M.) to the deficient ventricular septum preventing interventricular communication. A. = aorta.
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Figure 5
Case 3. Electrocardiogram of the 79-year-old man showing complete right bundle branch block and signs of anteroseptal myocardial infarction.

Figure 6
Case 3. Thoracic roentgenogram of the 79-year-old man showing cardiomegaly and right pleural effusion.

gram showed mild cardiac enlargement involving the right ventricle. The pulmonary arterial segment and the pulmonary vasculature were prominent.

When the patient was 39 years old cardiac catheterization showed a large left-to-right shunt at the atrial level. The pulmonary arterial pressure was 37/11 mm Hg. Following these observations the patient was operated upon, at which time an atrial septal defect of the AVC type was closed. A cleft found in the mitral valve was also repaired. The patient remained well for eight years following operation but died suddenly and unexpectedly when 48 years old.

The last patient (case 5) was a woman who died at the age of 49 years. During pregnancy at the age of 30 years a cardiac murmur was first noted. At age 42 years physical examination was consistent with atrial septal defect. The electrocardiogram showed an indeterminate axis, first degree atroventricular block and incomplete right bundle branch block with right ventricular hypertrophy. Thoracic roentgenograms showed marked cardiac enlargement, a prominent main pulmonary arterial segment and increased vasculature. Cardiac catheterization showed a large left-to-right shunt at the atrial level with the pulmonary arterial pressure being 58/20 mm Hg. Corrective operation was recommended. At the age of 49 years, during induction of anesthesia, she developed ventricular fibrillation and died.

In cases 3 and 5, pathologic examination showed essentially similar malformations characteristic of the partial type of AVC (figs. 7-9). The ventricular septum was deficient in the subaortic area but adhesion of the anterior mitral leaflet to the rim of the deficient septum prevented interventricular communication. In case 4, wherein operation had been done eight years before death, pathologic examination indicated total correction of the type of malformation observed in cases 3 and 5 (fig. 8).

In only one patient was there significant coronary atherosclerosis. This was observed in the 79-year-old man (case 3) along with signs of healed infarction.
Case 3. Partial type of AVC.  

a. External view of the heart and great vessels showing the wide pulmonary trunk (P.T.).  
b. Interior of the right atrium (R.A.) and right ventricle (R.V.) showing the ostium primum (D.).  
c. Interior of the left ventricle (L.V.) showing cleft anterior leaflet of the mitral valve (A.M.) attached to the deficient ventricular septum (arrows).  
A. = aorta.

Case 4. Partial type of AVC (8 years postoperative).  

a. Interior of the right atrium (R.A.) and right ventricle (R.V.) Arrow points to the site of the endothelialized prosthetic patch used to close the ostium primum 8 years previously.  
b. Interior of the left atrium (L.A.) and left ventricle (L.V.). The endocardium covering the prosthetic patch (P.) has been removed. The anterior leaflet of the mitral valve is cleft (arrows).
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Figure 9

Case 5. Partial type of AVC. a. Interior of the right atrium (R.A.) and right ventricle (R.V.) showing the ostium primum (D.) and normal tricuspid valve. b. Left atrium (L.A.) and left ventricle (L.V.) showing the ostium primum (D.) and thickened cleft (arrow) anterior leaflet of the mitral valve.

Comment

This communication presents five examples of patients with AVC who survived to the age of 46 years or longer. It was of interest to determine how unusual such longevity is. For this reason, we reviewed the ages at death of the 139 cases at our disposal of which pathologic specimens of heart show the AVC malformation. These were divided into the three anatomic

Figure 10

Age at the time of death in 139 patients of persistent common AVC. The complete type of the defect was present in 91 cases and the transitional or partial types of the defect in 48 patients.
types of the malformation as defined in the main body of this paper. A review of this material (fig. 10) shows that of the 139 cases of AVC 91 were of the complete type and 48 of the transitional or partial types.

It is generally accepted 2-4 that longevity is less in the complete type than in the transitional and partial types. This view is supported by our material in that death under one year of age occurred in 65 per cent of the cases with the complete type and in 33 per cent of the partial and transitional types grouped together. The latter figure, nevertheless, indicates a significant problem in the infant with the partial and transitional types.

Further support of the better prognosis for the partial and transitional types, as compared to the complete, is given by our material. Of the 21 patients who survived ten years or longer, in nine the AVC was of the complete type and in 12 of the partial or transitional types. Moreover, in the group of five patients who survived to the age of 46 years or older only one was of the complete type. As far as we are aware, our 46-year-old patient with the complete type of AVC is the oldest with this form of the malformation. With regard to long survival in the partial and the transitional types, Somerville 5 referred to reported cases and indicated that in her series there were 14 of 122 patients who survived to ages between 40 and 69 years. In four of these, there was necropsy confirmation, the oldest being 56 years.

In the adult, the clinical suspicion that the AVC malformation is present depends, as in the young, upon signs of an atrial septal defect coupled with the electrocardiographic signs typical of this malformation.

Attention is drawn to our case 1 in which AVC was associated with corrected transposition. That this is an unusual association is borne out by the fact that it was observed only once in our series of 139 cases of AVC and this association is usually not mentioned in reported series. In our case the electrocardiographic pattern did not suggest either of the two malformations which were present.

References

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RAJENDRA TANDON, JAMES H. MOLLER and JESSE E. EDWARDS

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