Mitral Atresia with Normal Aortic Valve
A Study of Eighteen Cases and a Review of the Literature

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SUMMARY Eighteen cases of mitral atresia with normal aortic valve plus 68 cases from the literature are analyzed. A new classification based on anatomical findings is proposed. Pulmonary stenosis or atresia is frequent in the type with transposition of the great arteries. The rare instances of normal or large left ventricle are due to a large ventricular septal defect, or to straddling or displaced tricuspid valve.

The clinical, radiologic and electrocardiographic findings have been summarized and correlated with the different physio-

MITRAL ATRESIA with normal aortic valve is an uncommon cardiac lesion,1,2 which has been included by some in the spectrum of hypoplastic left heart syndrome.3,4 However, because it tends to have a longer natural history, a different pathophysiology, and a different clinical picture, it seems appropriate to consider it separate from hypoplastic left heart syndrome.5,6

The purpose of this study is to propose a new classification of this entity together with a description of the anatomical findings, clinical manifestations, hemodynamics and angiographic features. The importance of selective left atrial injection for precise diagnosis is emphasized.

Patient Material

The patient material includes 18 cases from our institution confirmed by autopsy or angiocardiography with 14 pathologic specimens available for examination. All patients had a complete physical examination, electrocardiogram and teleoenterogram. Cardiac catheterization with angiocardiography was performed in nine cases. Sixty-eight cases in the literature with autopsy confirmation were reviewed and the data available are included.2,5,6,8-48

Cases with L bulboventricular loop and atresia of the left ativoventricular valve are not included, because from a strictly anatomical point of view they represent cases of tricuspid atresia. Within the subgroup of mitral atresia with single left ventricle we have only considered cases with the characteristics previously described by one of the authors.22 Other cases reported as mitral atresia with a single or a common ventricle23-26 are included in the subgroup with an absent left ventricle.

Results

Anatomic Findings

Of the 82 cases with autopsy data, 52 had normally related great vessels with varying degrees of dextroposition of the aorta. Transposition of the great arteries was present in the remaining 30.

The left ventricle was absent in 19 cases, hypoplastic in 50, normal in ten, and single in three.22,26,27 Pulmonary stenosis or atresia was found in 24 cases, of which 16 were type II (table 1).

The left atrium was small in all but three cases where it was normal or large.26 It was absent in one.9 The left atrial wall was often thick and occasionally associated with fibroelastosis. A dimple was frequently observed in the floor of the left atrium. In one instance22 valvular tissue was encountered and in one of our own cases a completely formed but closed mitral valve with minute chordae tendineae and papillary muscle was present (fig. 1). A patent foramen ovale was observed in 46 cases and a true atrial septal defect in another 25 (table 2). In seven specimens there was no interatrial communication, although in some a levoatriocardinal vein was described.29 This was demonstrated by angiocardiography in one of our cases (fig. 6). In one case, there was a communication between the left atrium and coronary sinus.30 In another there was an aneurysm of the foramen ovale,31 and in another the margins of the foramen ovale were calcified.4 The right atrium and tricuspid valve were consistently enlarged. The right ventricle also was large in cases with absent or hypoplastic left ventricle; in the remainder it was small or even reduced to an outlet chamber. In 58 specimens there was a ventricular septal defect, which was usually small and high. In a few cases the defect was large or multiple,29,32 some being muscular. In five instances with straddling of the tricuspid valve, the defect was posterior as in common ativoventricular canal.32,33,34 There was one case of hypoplastic left ventricle without any interventricular communication.35

Patent ductus arteriosus was present in 56 cases, coarctation of the aorta in 17, and interruption of the aortic isthmus in two.36,37 A left superior vena cava was observed in 11 cases, anomalous pulmonary venous drainage in eight, and right aortic arch in six of the specimens with pulmonary stenosis. Cor triatriatum was found in three instances.38-40 Bilateral conus was observed in four of our own specimens. Eight patients had asplenia6,10,12,19,20,30,31,42 and four had polysplenia.5,36,37,43

Clinical Manifestations

Clinical data were available in 76 patients. Forty-seven were male and 35 were female. The sex was not specified in

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four. Symptoms appeared at an early age, the majority of patients having cyanosis, tachypnea or respiratory difficulty within the first week of life. Patients who survived longer periods had recurrent respiratory infections with failure to thrive. Five of our cases with pulmonary stenosis had one or more cyanotic spells. Cyanosis was more severe in cases with pulmonary outflow tract obstruction (table 3).

The arterial pulse was normal in the majority of cases with the femoral pulsations being absent in patients with associated coarctation of the aorta. Hepatomegaly was often present and in our series was more pronounced in patients without pulmonary outflow obstruction. In 35 cases, a systolic murmur was described, and in seven (five of our own) there was a mid-diastolic murmur. In one of our patients with pulmonary atresia a continuous murmur was heard in the back. A case with a continuous murmur in the low sternal area due to flow of blood from the left to the right atrium has been reported. In 23 cases, a murmur was either not noted or was described as insignificant. Gallop rhythm was present in six. The second sound was single in almost all cases of severe pulmonary stenosis and in the remaining cases the most frequent finding was a loud second sound. In three of our cases there was a constant split second sound.

Radiological Findings

Radiographic findings were available in 62 cases. Cardiomegaly was usually present and in our series was less severe in the patients with associated pulmonary stenosis. The cardiac silhouette was generally nonspecific, but in many cases with pulmonary stenosis the coeur en sabot configuration was present (table 3). Radiographic evidence of left atrial enlargement was present in only two cases.

In the absence of severe pulmonary outflow tract obstruction, pulmonary arterial and venous markings were increased. They were usually decreased with severe pulmonary stenosis, although in some of our cases pulmonary venous stasis was seen (fig. 2a). In one of our patients a pattern of acute pulmonary edema was observed shortly before death (fig. 2b).

Electrocardiographic Findings

Electrocardiographic data were available in 53 cases. The QRS axis was to the right in 48 and to the left in five (fig.

<table>
<thead>
<tr>
<th>Table 1</th>
<th>Classification of Mitral Atresia with Normal Aortic Valve*</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Total</td>
</tr>
<tr>
<td>I. Normally related great arteries</td>
<td></td>
</tr>
<tr>
<td>Absent left ventricle</td>
<td>15 (4)</td>
</tr>
<tr>
<td>Hypoplastic left ventricle</td>
<td>5 (1)</td>
</tr>
<tr>
<td>Normal left ventricle</td>
<td>1 (0)</td>
</tr>
<tr>
<td>Single left ventricle</td>
<td>1 (0)</td>
</tr>
<tr>
<td>II. Transposed great arteries</td>
<td></td>
</tr>
<tr>
<td>Absent right ventricle</td>
<td>15 (4)</td>
</tr>
<tr>
<td>Hypoplastic right ventricle</td>
<td>5 (1)</td>
</tr>
<tr>
<td>Normal right ventricle</td>
<td>1 (0)</td>
</tr>
</tbody>
</table>

*Figures in parentheses represent our own cases.

Autopsy proven cases only.

Table 2. Associated Lesions

<table>
<thead>
<tr>
<th>Lesion</th>
<th>Number</th>
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<tbody>
<tr>
<td>Interventricular communication</td>
<td>71 (14)*</td>
</tr>
<tr>
<td>Patent foramen ovale</td>
<td>46 (9)</td>
</tr>
<tr>
<td>Patent ductus arteriosus</td>
<td>56 (11)</td>
</tr>
<tr>
<td>Coarctation of aorta</td>
<td>17 (4)</td>
</tr>
<tr>
<td>Left superior vena cava</td>
<td>11 (2)</td>
</tr>
<tr>
<td>Anomalous pulmonary venous drainage</td>
<td>8 (1)</td>
</tr>
<tr>
<td>Right aortic arch</td>
<td>6 (2)</td>
</tr>
<tr>
<td>Straddling tricuspid valve</td>
<td>5 (0)</td>
</tr>
<tr>
<td>Con tricuspid apparatus</td>
<td>3 (0)</td>
</tr>
<tr>
<td>Bilateral conus</td>
<td>(4)</td>
</tr>
</tbody>
</table>

*Figures in parentheses represent our own cases.

3) Right atrial and right ventricular hypertrophy were the most usual findings. Two cases of left ventricular hypertrophy were reported. Initial q waves in V1 were frequently present (44% in our own cases, table 3).

In two of the patients with overriding tricuspid valve, there was evidence of biventricular hypertrophy, and in another there was left ventricular predominance. In two of the cases with a single left ventricle, electrocardiographic signs suggesting left ventricular hypertrophy were observed.
### Table 3. Clinical Data

<table>
<thead>
<tr>
<th>Case</th>
<th>Great arteries</th>
<th>Left ventricle</th>
<th>Pulmonary outflow</th>
<th>Sex</th>
<th>Age at death (d)</th>
<th>Cyanosis</th>
<th>Liver edge (cm)</th>
<th>Peripheral arterial pulses</th>
<th>Murmurs</th>
<th>2nd sound</th>
<th>Chest X-ray</th>
<th>ECG findings</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>CTR</td>
<td>Heart shape</td>
</tr>
<tr>
<td>1. HJC</td>
<td>N</td>
<td>H</td>
<td>N</td>
<td>M</td>
<td>2</td>
<td>mild</td>
<td>3</td>
<td>N (FP a)</td>
<td>No</td>
<td>split</td>
<td>0.70</td>
<td>Inespec.</td>
</tr>
<tr>
<td>2. JCM</td>
<td>N</td>
<td>H</td>
<td>N</td>
<td>M</td>
<td>46</td>
<td>mild</td>
<td>2</td>
<td>N (FP a)</td>
<td>No</td>
<td>?</td>
<td>0.65</td>
<td>C en S</td>
</tr>
<tr>
<td>3. HJP</td>
<td>N</td>
<td>H</td>
<td>N</td>
<td>F</td>
<td>2</td>
<td>—</td>
<td>3</td>
<td>N (FP a)</td>
<td>systolic fixed split</td>
<td>0.65</td>
<td>Inespec. Increased</td>
<td>100° qRs RAE, RVE</td>
</tr>
<tr>
<td>4. FCG</td>
<td>N</td>
<td>H</td>
<td>N</td>
<td>M</td>
<td>15</td>
<td>mild</td>
<td>3</td>
<td>N (FP a)</td>
<td>systolic split</td>
<td>0.75</td>
<td>Inespec. Increased</td>
<td>130° qR RVE</td>
</tr>
<tr>
<td>5. DFI</td>
<td>N</td>
<td>H</td>
<td>N</td>
<td>M</td>
<td>3</td>
<td>mild</td>
<td>4</td>
<td>W (FP a)</td>
<td>systolic single</td>
<td>0.62</td>
<td>Inespec. Increased</td>
<td>105° R RVE</td>
</tr>
<tr>
<td>6. AMC</td>
<td>N</td>
<td>?</td>
<td>N</td>
<td>F</td>
<td>50</td>
<td>mild</td>
<td>4</td>
<td>W (FP a)</td>
<td>No</td>
<td>split</td>
<td>0.60</td>
<td>Inespec.</td>
</tr>
<tr>
<td>7. JPV</td>
<td>N</td>
<td>?</td>
<td>N</td>
<td>M</td>
<td>33</td>
<td>mild</td>
<td>4</td>
<td>N (FP a)</td>
<td>systolic fixed split</td>
<td>0.63</td>
<td>Inespec. Increased</td>
<td>130° R RVE</td>
</tr>
<tr>
<td>8. JFS</td>
<td>N</td>
<td>N</td>
<td>N</td>
<td>S</td>
<td>20</td>
<td>mild</td>
<td>3</td>
<td>N (FP a)</td>
<td>systolic fixed split</td>
<td>0.68</td>
<td>Inespec. Increased</td>
<td>150° Rs RAE, RVE</td>
</tr>
<tr>
<td>9. MSL</td>
<td>N</td>
<td>Single</td>
<td>N</td>
<td>F</td>
<td>3</td>
<td>severe</td>
<td>1</td>
<td>N (FP a)</td>
<td>ejectional single</td>
<td>0.58</td>
<td>Inespec. Decreased</td>
<td>70° rS LVE</td>
</tr>
<tr>
<td>10. SC</td>
<td>T</td>
<td>A</td>
<td>N</td>
<td>M</td>
<td>75</td>
<td>—</td>
<td>4</td>
<td>W (FP a)</td>
<td>systolic split</td>
<td>0.70</td>
<td>Inespec. Increased</td>
<td>180° qR RAE, RVE</td>
</tr>
<tr>
<td>11. HES</td>
<td>T</td>
<td>A</td>
<td>S</td>
<td>M</td>
<td>1</td>
<td>severe</td>
<td>1</td>
<td>N (FP a)</td>
<td>systolic single</td>
<td>0.57</td>
<td>C en S Decreased</td>
<td>180° qR RAE, RVE</td>
</tr>
<tr>
<td>12. HBG</td>
<td>T</td>
<td>A</td>
<td>S</td>
<td>M</td>
<td>2</td>
<td>severe</td>
<td>3</td>
<td>W (FP a)</td>
<td>systolic single</td>
<td>0.60</td>
<td>C en S Increased</td>
<td>180° R RVE</td>
</tr>
<tr>
<td>13. ALC</td>
<td>T</td>
<td>A</td>
<td>S</td>
<td>M</td>
<td>4</td>
<td>severe</td>
<td>1</td>
<td>N (FP a)</td>
<td>ejectional ?</td>
<td>0.58</td>
<td>Inespec. Increased</td>
<td>130° R RAE, RVE</td>
</tr>
<tr>
<td>14. YCC</td>
<td>T</td>
<td>H</td>
<td>S</td>
<td>F</td>
<td>10</td>
<td>severe</td>
<td>—</td>
<td>N (FP a)</td>
<td>No single</td>
<td>0.55</td>
<td>C en S Decreased</td>
<td>170° qRs RAE, RVE</td>
</tr>
<tr>
<td>15. PML</td>
<td>T</td>
<td>H</td>
<td>S</td>
<td>M</td>
<td>78</td>
<td>severe</td>
<td>3</td>
<td>N (FP a)</td>
<td>systolic fixed split</td>
<td>0.60</td>
<td>Inespec. Decreased</td>
<td>140° Rs RVE</td>
</tr>
<tr>
<td>16. FFB</td>
<td>T</td>
<td>H</td>
<td>S</td>
<td>M</td>
<td>15</td>
<td>mild</td>
<td>1</td>
<td>N (FP a)</td>
<td>systolic fixed split</td>
<td>0.62</td>
<td>Inespec. Increased</td>
<td>170° Rs RAE, RVE</td>
</tr>
<tr>
<td>17. ERL</td>
<td>T</td>
<td>?</td>
<td>N</td>
<td>F</td>
<td>a-2½ yr</td>
<td>moderate</td>
<td>3</td>
<td>N (FP a)</td>
<td>systolic split</td>
<td>0.65</td>
<td>Round Increased</td>
<td>150° Rs RAE, RVE</td>
</tr>
<tr>
<td>18. FRT</td>
<td>T</td>
<td>?</td>
<td>S</td>
<td>M</td>
<td>a-2 yr</td>
<td>severe</td>
<td>1</td>
<td>N (FP a)</td>
<td>continuous split</td>
<td>0.65</td>
<td>C en S Increased</td>
<td>130° R RAE, RVE</td>
</tr>
</tbody>
</table>

Abbreviations: M = male; F = female; N = normal; T = transposed; H = hypoplastic; A = absent; S = stenotic; d = days; m = months; a = alive; yr = years; FP a = femoral pulses absent; W = weak; C en S = Coeur en sabot; RAE = right atrial enlargement; RVE = right ventricular enlargement; LVE = left ventricular enlargement; ? = unknown.
MITRAL ATRESIA WITH NORMAL AORTIC VALVE/Moreno, Quero, Diaz

FIGURE 2. a) Teleradiogram in a case with associated transposition of the great arteries and pulmonary atresia. There is cardiomegaly with coeur en sabot configuration. Lung fields show increased pulmonary venous markings. b) Roentgenogram of thorax in a case with small foramen ovale. Butterfly pattern of acute pulmonary edema is present.

FIGURE 3. Electrocardiogram of an infant with type I mitral atresia. Note left axis deviation (-100°) and right ventricular enlargement.

In our series the T wave was positive in V1 in eight cases, negative in seven and flat in three.

Hemodynamic Data (table 4)

Very few of the cases from the literature include hemodynamic data. In eight of our nine catheterized patients there was a step-up in oxygen saturation in the right atrium. Peripheral arterial oxygen saturations were obtained in four cases and were decreased in three. Left atrial hypertension was present in seven of the eight cases in which this chamber was entered, with interatrial gradients between 1 and 20 mm Hg (fig. 4). The a wave was the most prominent wave in the left atrial pressure curves. Right ventricular hypertension was found in seven cases, and was systemic in the four cases in which the aorta was entered. The pulmonary artery was

<table>
<thead>
<tr>
<th>Table 4. Hemodynamic Data</th>
<th>Oxygen saturation (%)</th>
<th>Pressure (mm Hg)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patient</td>
<td>SVC</td>
<td>RA</td>
</tr>
<tr>
<td>FCG</td>
<td>77</td>
<td>97</td>
</tr>
<tr>
<td>DFIP</td>
<td>53</td>
<td>76</td>
</tr>
<tr>
<td>AMCC</td>
<td>50</td>
<td>66</td>
</tr>
<tr>
<td>JPVE</td>
<td>36</td>
<td>48</td>
</tr>
<tr>
<td>HESA</td>
<td>50</td>
<td>65</td>
</tr>
<tr>
<td>ALG</td>
<td>52</td>
<td>66</td>
</tr>
<tr>
<td>FFB</td>
<td>58</td>
<td>75</td>
</tr>
<tr>
<td>ERL</td>
<td>58</td>
<td>69</td>
</tr>
<tr>
<td>FRDT</td>
<td>40</td>
<td>51</td>
</tr>
</tbody>
</table>

Abbreviations: SVC = superior vena cava; RA = right atrium; RV = right ventricle; PA = pulmonary artery; LA = left atrium; SA = systemic artery; Ao = aorta; m = mean pressure.
catheterized in five patients and the pressures were equal to those of the right ventricle in four. A transpulmonary gradient of 60 mm Hg was recorded in one case. In two patients the pulmonary artery pressure was only slightly higher than the left atrial pressure. In three, the interatrial pressure gradient decreased following balloon atrial septostomy.

**Angiocardiographic Findings**

In six of our cases selective left atrial contrast injection was done. In all, the contrast passed to the right atrium and then to the right ventricle, and then the great vessels were filled (fig. 5). In one case, contrast was injected in the pulmonary artery and this same filling sequence was seen in the recirculation phase. In the two remaining cases right ventricular injection was done and the mitral valve area was not visualized. In three cases, angiocardiography performed in the right ventricle showed a hypoplastic left ventricular cavity (fig. 6a).

In one case a vein connecting the left atrium with the left innominate vein was visualized (levoatriocardinal vein, fig. 6b). In five cases, transposition of the great arteries was observed, with bilateral conus in three. Pulmonary stenosis was seen in two patients and pulmonary atresia with bronchial collateral circulation in one.

**Clinical Course**

The average age at death was six months. In our series, 13 patients (72%) died before the age of three months. One of our patients and seven from the literature survived the first year of life, with a maximum life span of 16 years. Of the patients who lived beyond one year, four had an atrial septal defect with pulmonary stenosis, two had an atrial septal defect, and one had pulmonary stenosis. Two of our patients are still alive; one at two years of age with a surgical atrial septostomy and the other at two and one-half years with a balloon atrial septostomy. Two of our patients died after surgical aortopulmonary anastomosis, one from acute pulmonary edema and the other from hypoxemia. Another three patients reported in the literature with surgical aortopulmonary anastomosis died postoperatively.6, 41, 45

![Figure 4](http://circ.ahajournals.org/)

**Figure 4.** Pressure withdrawal tracing from left atrium to right atrium showing marked left atrial hypertension with interatrial pressure gradient of 20 mm Hg.

![Figure 5](http://circ.ahajournals.org/)

**Figure 5.** Left atrial angiocardiogram showing (a) no filling of left ventricle, and (b) contrast passing to right atrium and then to right ventricle. Note the small size of left atrium.

**Discussion**

From the data recorded in the literature and our own material, a new classification based on the position of the great arteries, the size of the left ventricle, and the presence or absence of pulmonary outflow tract obstruction is proposed (table I). In type I the relationship of the great arteries is basically normal, with varying degrees of dextroposition of the aorta. Type II is characterized by transposition of the great arteries with or without bilateral conus.

In type I, which is more frequent than type II, 75% of cases have a hypoplastic left ventricle. Absent, normal or single left ventricle is rare in this type. Pulmonary stenosis is infrequent (14% of cases) and although not considered in this study, aortic stenosis or atresia has been reported to be frequent with mitral atresia and normally related great arteries.7, 51, 52

In type II, 50% of patients have an absent left ventricle and in the remainder this chamber is hypoplastic or normal except in one case associated with a single left ventricle.57 Fifty-three percent of cases have associated pulmonary outflow tract obstruction. This finding, together with the fact that obstructive anomalies of the aortic outflow tract are very frequent in cases with normally related great arteries, leads to the conclusion that the great artery which is nearer to the atretic mitral valve is more prone to develop obstructive lesions.

In some cases the normal size of the left ventricle has been
attributed to the existence of a large or multiple ventricular septal defects. In our case with a formed mitral valve, the normal size of the left ventricle was probably due to delayed mitral atresia. In other cases, the normal or enlarged left ventricle is due to straddling or displacement of the tricuspid valve.

It seems preferable to separate the group of mitral atresia with normal aortic valve from the broad spectrum of hypoplastic left heart syndrome because of basic differences between the two. In aortic atresia, the most representative anomaly of the syndrome, there is retrograde flow in the ascending aorta with a ductal right-to-left shunt. Clinically, cardiac failure with absent or very weak peripheral arterial pulses is more pronounced and the natural history is shorter.

Mitrail atresia represents an obstacle to the pulmonary venous return, and the whole pulmonary blood flow has to be driven up to the right heart chambers through some form of interatrial communication. Consequently, pulmonary venous congestion and pulmonary hypertension ensue, relating to the size of the atrial septal defect and to the amount of pulmonary blood flow. Symptomatology and prognosis depend on the interrelation between these two factors. In cases with pulmonary atresia, hypoxemia also plays an important role. The association of a large atrial septal defect and a moderately severe pulmonary stenosis would be the ideal situation for survival of mitral atresia cases. The position of the great arteries does not modify either the hemodynamic or the clinical features, because functionally both situations can be considered as a double outlet right ventricle. Nevertheless, pulmonary outflow tract obstruction is more frequently associated with cases with transposition which accounts for the fact that in these cases, cyanosis and hypoxemia are more usually found. When the left ventricle is of normal size, it can contribute to the heart work. These cases and those with single left ventricle show an electrocardiographic pattern rather different from the usual one. In cases with straddling or displaced tricuspid valve, evidence of left or biventricular hypertrophy was seen in the electrocardiogram, rather than the usual finding of right ventricular hypertrophy.

In two of our cases, pulmonary artery pressure was slightly higher than left atrial pressure. In both, left atrial pressure was 20 mm Hg higher than right atrial pressure and both had severe pulmonary venous congestion on X-ray examination. At autopsy, one of these had a small patent foramen ovale. In this case, acute pulmonary edema developed (fig. 2b), suggesting that the pulmonary hypertension was passive with failure of the protective mechanism of the pulmonary arteriolar vasoconstriction.

Selective contrast injection either into the left atrium or into the pulmonary artery is necessary for diagnosis of this malformation. Mitrail atresia should be suspected when an oxygen saturation step-up is found at the level of the right atrium together with left atrial hypertension. Precise diagnosis is important because of therapeutic implications and, if confirmed, a balloon atrial septostomy can be performed at catheterization. In cases with pulmonary stenosis in which the mitral atresia is overlooked, acute pulmonary edema may be precipitated by a shunt procedure, as occurred in one of our cases.

At present there is no corrective surgery for this malformation. Palliative procedures include atrial septostomy with aorticopulmonary anastomosis in cases with pulmonary stenosis, or atrial septostomy with pulmonary artery banding in cases with increased pulmonary blood flow. The possibility of corrective surgery in cases with overriding or displaced tricuspid valve has been considered.

The embryological origin of this malformation is not well established. It has been attributed to displacement or malalignment of the atrial and/or ventricular septum, but cases with those septal malpositions have been described without mitral atresia. Exaggerated growth of the endocardial cushions and displacement of the atrioventricular canal to the right have also been proposed as etiological factors. Watson et al. have suggested that fusion of the central endocardial cushions with the left lateral cushion at or before six weeks of embryological life may give rise to this defect. Hast et al. produced hypoplastic left heart syndrome in chick embryos by placing a device in the left atrioventricular canal that changed the course of the blood.

**Acknowledgment**

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