Ebstein's Anomaly of the Left Atrioventricular Valve with Congenital Corrected Transposition of the Great Arteries*

Diagnosis by Intracavitary Electrocardiography

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We present the findings in a 13-month-old infant with angiographically confirmed congenital corrected transposition of the great arteries and insufficiency of the left atrioventricular valve. Simultaneous intracavitary electrocardiographic and pressure recordings across the left atrioventricular valve were similar to those obtained in Ebstein's anomaly and suggested Ebstein's disease of the left atrioventricular valve. To our knowledge, this is the first reported case with intracavitary electrocardiograms in a patient with congenital corrected transposition of the great arteries with Ebstein's malformation of the left atrioventricular valve. The usefulness of the simultaneous recording of the intracavitary ECG and pressure in the diagnosis by catheterization of Ebstein's anomaly of the left atrioventricular valve in patients with congenital corrected transposition of the great arteries is emphasized.

Intracavitary electrocardiographic studies are widely used at cardiac catheterization to confirm the presence of Ebstein's anomaly of the tricuspid valve when this malformation occurs in a patient with normally positioned great arteries and ventricles; however, its use in the diagnosis of Ebstein's anomaly of the left atrioventricular valve in patients with congenital corrected transposition of the great arteries has not been previously reported. The purpose of this communication is to report the simultaneous recordings of the intracavitary electrocardiogram and pressure in a patient with congenital corrected transposition of the great arteries and Ebstein's anomaly of the left atrioventricular valve and to demonstrate the usefulness of intracavitary electrocardiographic studies in the diagnosis of this lesion.

Case Report

An infant girl was the product of a normal pregnancy, labor, and delivery and had a birth weight of 3.3 kg (7 lb 4 oz). At two weeks of age, she had a cardiac murmur, tachypnea, and poor feeding. Examination at that time revealed physical findings suggestive of a large ventricular septal defect and moderate congestive heart failure. The chest x-ray film showed marked cardiomegaly and increased pulmonary vascular markings. The ECG was interpreted as showing right ventricular hypertrophy. The initial forces of the QRS vector were directed to the left, suggesting congenital corrected transposition of the great arteries. After improvement of the congestive heart failure following therapy with digoxin and diuretics, cardiac catheterization (Table 1) and selective cineangiographic studies were performed, at which time the following diagnoses were made: congenital corrected transposition of the great arteries, large ventricular septal defect, subvalvular pulmonic stenosis, and a stretched patent foramen ovale. The left-sided ventricular cineangiogram revealed significant regurgitation across the left atrioventricular valve, suggesting the possibility of Ebstein's anomaly of the left atrioventricular valve.

During the subsequent 12 months, the patient did well, with good control of her congestive failure and normal linear growth, but weight gain was slow. At 13 months of age, physical examination revealed hyperdynamic left and right ventricular impulses and systolic thrills at the apex and lower left sternal border. The second heart sound was physiologically split, with a normal pulmonary component. A grade 5/6 holosystolic murmur was heard at the apex, with radiation into the axilla. An additional grade 4/6 holosystolic murmur was heard at the lower left sternal border. There was also a grade 3/6 middiastolic murmur at the apex. The ECG revealed left atrial enlargement, biventricular hypertrophy, and leftward initial QRS forces in the horizontal plane. The chest x-ray film was similar to that obtained previously, except for

<table>
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<tr>
<th>Site</th>
<th>Saturation (percent)</th>
<th>Pressure (mm Hg)**</th>
<th>Site</th>
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<th>Pressure (mm Hg)**</th>
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<tbody>
<tr>
<td>Superior vena cava</td>
<td>69</td>
<td>. . .</td>
<td></td>
<td>63</td>
<td>. . .</td>
</tr>
<tr>
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<td>77</td>
<td>a = 10; v = 6 (4.5)</td>
<td>Right atrium</td>
<td>66</td>
<td>a = 9; v = 6 (5)</td>
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<td>Right-sided ventricle</td>
<td>84</td>
<td>75/9</td>
<td>Pulmonary artery</td>
<td>90</td>
<td>36/8</td>
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<tr>
<td>Pulmonary vein</td>
<td>91</td>
<td>30/16 (22)</td>
<td>Pulmonary vein</td>
<td></td>
<td>32/17 (23)</td>
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<tr>
<td>Left atrium</td>
<td>95</td>
<td>a = 9.5; v = 13 (10)†</td>
<td>Left atrium</td>
<td>93</td>
<td>a = 18; v = 22 (16)‡</td>
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<tr>
<td>Left-sided ventricle</td>
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<td>65/10</td>
<td>Left-sided ventricle</td>
<td>96</td>
<td>78/16</td>
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<tr>
<td>Aorta</td>
<td></td>
<td>. . .</td>
<td>Aorta</td>
<td>92</td>
<td>76/50 (62)</td>
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Table 1—Data from Cardiac Catheterisation

*Ratio of pulmonary to systemic blood flow was 5.4/1 at 16 days and 2/1 at 13 months.
**Numbers within parentheses indicate mean pressures.
†Left atrium was entered via patent foramen ovale.
‡Left atrium was entered from left-sided ventricle.
additional left atrial enlargement.

Data from cardiac catheterization (Table 1) suggested that the ventricular septal defect had become smaller, when compared to the first study. There was absence of the previously demonstrated pulmonary outflow tract gradient and a significantly decreased ratio of pulmonary-to-systemic flow, when compared to the initial study. Cineangiographic studies demonstrated the typical findings of congenital corrected transposition of the great arteries and significant insufficiency of the left atrioventricular valve (Fig 1).

Because of the suspected presence of Ebstein’s anomaly of the left atrioventricular valve, simultaneous intracavitary electrocardiographic studies and pressure recordings were performed. A No. 5 French unipolar platinum-tipped electrode catheter (USCI 5610) was advanced retrograde to the ascending aorta, where a small loop was formed. The loop was advanced across the aortic valve into the left-sided ventricle, and the tip was successfully manipulated across the left atrioventricular valve into the left atrium. Pressure and the intracavitary ECG were then recorded simultaneously as

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**Figure 1.** Cineangiograms. (a) Selected frame from cineangiogram of right-sided ventricle (RSV), demonstrating typical features of morphologic left ventricle from which arises pulmonary artery (PA). (b) Cineangiographic frame of left-sided ventricle (LSV), demonstrating typical features of morphologic right ventricle from which arises aorta (Ao). Opacification of markedly enlarged left atrium (LA) occurred due to insufficiency of left atrioventricular valve. Pulmonary artery (PA) was also opacified due to left-to-right shunt through ventricular septal defect.

**Figure 2.** Simultaneous intracardiac ECGs (IECG) and pressure tracings. At left is atrial pressure curve and atrial electrogram in left atrium (LA). At center is atrial pressure curve and ventricular electrogram in “atrialized” portion of left-sided ventricle (LSV), due to Ebstein’s malformation of left atrioventricular valve. At right is ventricular pressure curve and ventricular electrogram in body of left-sided ventricle (LSV). Ventricular pressure curve is damped due to small size of lumen of catheter. Pressure is in millimeters of mercury. These tracings were recorded while catheter was withdrawn slowly from left atrium to left-sided ventricle.
the catheter was withdrawn from the left atrium into the left-sided ventricle. This demonstrated a ventricular electrical complex at a time when an atrial pressure tracing was present (Fig 2), thus confirming the presence of Ebstein’s anomaly of the left atrioventricular valve.

**Discussion**

Corrected transposition of the great arteries was originally described by von Rokitansky in 1875. Although only a few cases were reported by 1950, several large series of cases have since been published and have defined the pathologic, clinical, and hemodynamic features of this entity. The case presented herein demonstrates the typical angiographic findings of congenital corrected transposition of the great arteries (Fig 1). The coarsely trabeculated morphologic right ventricle is left-sided and gives rise to the aorta. The finely trabeculated morphologic left ventricle is right-sided and gives rise to the pulmonary artery. The decrease in the pressure in the right-sided ventricle at 13 months of age, when compared to the initial study (see Table 1), indicates partial closure of the ventricular septal defect, with a resultant decrease in the ratio of pulmonary-to-systemic flow. The absence of the subpulmonary gradient at the second study suggests that the original gradient at 16 days of age may have been due in part to the large pulmonary blood flow and may be explained on the basis of a kinetic energy-related gradient.

In congenital corrected transposition of the great arteries, the atrioventricular valves correspond to the ventricular chamber. Therefore, the left atrioventricular valve is the morphologic tricuspid valve. Malformations of this left-sided tricuspid valve are very common and were considered an integral part of this pathologic entity by Paul and associates. The occurrence of an anomaly of Ebstein’s type in the left atrioventricular valve is well described however, its incidence is difficult to determine. Schiebler et al examined 13 specimens of hearts with congenital corrected transposition of the great arteries and visceral situs solitus and found a deformed left atrioventricular valve in 11. In seven cases, the deformity assumed the pattern of Ebstein’s anomaly.

The use of intracavitary electrocardiographic studies to confirm the diagnosis of the usual type of Ebstein’s anomaly of the tricuspid valve was first suggested by Sodi-Pallares and Marsico. The simultaneous recording of the intracavitary ECG and pressure at cardiac catheterization to confirm the diagnosis of Ebstein’s anomaly was first reported by Hernandez et al. In three patients with this malformation, these investigators showed that the recording of typical right ventricular intracavitary electrocardiographic patterns with simultaneously obtained atrial-type pressure curves was diagnostic of this condition. This was confirmed by subsequent reports by other investigators.

Schiebler et al suggested that this technique might be valuable in the diagnosis of left-sided Ebstein’s malformation in congenital corrected transposition of the great arteries if the electrode catheter were introduced into the left side of the heart. Kupersmith et al reported a patient with dextrocardia, visceral situs solitus, and congenital corrected transposition of the great arteries, in whom the systemic atrioventricular valve was abnormal at surgery. During surgery, Kupersmith et al recorded a ventricular electrogram from an area above this abnormal valve and confirmed the presence of an Ebstein-like deformity; however, to our knowledge, the use of the intracavitary ECG with simultaneously recorded pressures during cardiac catheterization to diagnose Ebstein’s anomaly of the left atrioventricular valve in congenital corrected transposition of the great arteries has not been previously reported. In the present case, the intracavitary ECG was recorded simultaneously with the curves for pressure while a unipolar platinum-tipped catheter was withdrawn from the left atrium into the left-sided ventricle. The following sequence was recorded (Fig 2): (1) atrial pressure and atrial ECG; (2) atrial pressure and ventricular ECG; and (3) ventricular pressure and ventricular ECG. The recording of a ventricular ECG simultaneously with an atrial pressure curve thus confirmed the presence of Ebstein’s anomaly of the left atrioventricular valve.

The presence of an Ebstein’s anomaly of the left atrioventricular valve should be suspected in any patient with congenital corrected transposition of the great arteries if there is evidence of insufficiency of the valve. We suggest that simultaneous intracavitary electrocardiographic studies and pressure recordings be performed routinely in all patients with congenital corrected transposition of the great arteries, especially if there is evidence of insufficiency of all left atrioventricular valve, in an attempt to confirm the presence of an anomaly of Ebstein’s type. If an atrial septal defect or patent foramen ovale is present, this could be easily performed antegrade. If there is no interatrial communication, the retrograde approach with appropriate looping of the electrode catheter, as in this report, can be used.

Diagnosis by catheterization of Ebstein’s anomaly of the left atrioventricular valve in congenital corrected transposition of the great arteries is important because of its high incidence and possible surgical implications. Van Mierop et al reported a patient whose death was probably related to unrecognized Ebstein’s anomaly of the left atrioventricular valve. Preoperative recognition of Ebstein’s disease may result in dealing with the diseased left atrioventricular valve at the time of surgical correction of the associated defects.

**References**

4. Watson H: Electrode catheters and the diagnosis of

**EBSTEIN’S ANOMALY**
Bilateral Sequestrations of Different Type with Mirror-Image Vascularization*

Jules A. Trudel, M.D., F.C.C.P.; Guy Lemtre, M.D.; and Antoine G. Rabbat, M.D.

The case of a 15-year-old boy with coexistent bilateral intralobar and extralobar sequestrations is reported. The interesting variant lies in the vascularization of each sequestration as a mirror-image of the other, the arterial supply originating from a common trunk of the thoracic aorta and the venous drainage converging to a unique channel into the ayzygos system.

The coexistence of intralobar and extralobar sequestrations is a rarity. Carter reports four cases, while reports by Kafka and Beco and by Pendse et al each describe one case. Ennis and associates present a case where the arterial supply to a sequestration in the left lower lobe originates from a common trunk low on the thoracic aorta, with a branch going to the right lower lobe. The venous drainage is to the pulmonary vein on the left, while there is no mention of the right side.

The interesting variant in our case report lies in the existence of common trunks, arterial and venous, in relation to a right intralobar and a left extralobar sequestration.

**Case Report**

A previously healthy 15-year-old boy was brought for consultation because of fever, cough, purulent sputum, and...

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![Figure 1. Preoperative chest x-ray film, showing cavitations and infiltration of right lower lobe and retrocardiac mass on left.](http://journal.publications.chestnet.org/pdfaccess.ashx?url=/data/journals/chest/20996/ on 04/08/2017)