An Unusual Cardiac Anomaly: Ebstein-like Malformation of the Left Atrioventricular Valve, Masquerading as a Dextrocardia*

Report of a Case

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EBSTEIN’S ANOMALY, first described in 1866, is relatively rare. It is a downward displacement of the tricuspid (right atrioventricular) valve below its annulus, dividing the right heart into a relatively large atrium and a small, at times functionless, right ventricle. The foramen ovale is usually patent and there may be other intracardiac anomalies.1–4 We are describing a similar lesion involving the left atrioventricular valve, seen in an adult who seemed to have dextrocardia.

CASE REPORT

The patient was a 35-year-old Caucasian, first seen at the City of Hope Medical Center in January, 1956. Although cyanosis had been present since birth, and he had been told he had a “right-sided heart,” the patient managed to live a relatively normal life with only moderate restriction of activity. In November, 1955, the first ominous symptoms were noted: cardiac palpitation and episodic throbbing, lasting for approximately 15 minutes. A few weeks later, right anterior chest “pressure” was noted, lasting only a few moments. The episodes increased in severity and duration. On admission, the pressure lasted up to several hours, and was relieved only upon lying down. Occasional episodes of exertional dyspnea and anxiety also occurred.

Physical examination: He appeared robust, was 6 feet tall and weighed 162 pounds. Except for poor dental hygiene, findings on examinations of the ears, nose, throat and lungs were normal. Blood pressure was 116/78 mm.Hg in the right arm, and 118/84 mm.Hg in the left arm. The peripheral pulse was 78 per minute and regular. The apical heart beat was palpable in the right fourth intercostal space in the mid-clavicular line in expiration; in the right fifth intercostal space in the mid-clavicular line in inspiration.

A grade II/IV systolic thrill, a grade IV/VI systolic murmur, and an accentuated second sound were present at the second, left anterior intercostal space. The murmur was transmitted into both sides of the neck and over the entire chest and back. The first sound was accentuated at the area of apical palpation (Fig. 1A). Femoral arterial pulsations were normal. The left testicle was atrophic, palpable in the left inguinal area. Except for clubbing of digits and cyanosis of the fingernail beds and tips of the fingers and toes, the remainder of findings on physical examination were essentially within normal limits.

Laboratory findings: Hematocrit was 72 per cent, with hemoglobin 24.7 mg. per cent. The total white count was 11,650 per mm.³ with 87 per cent polymorphonuclear leukocytes, 7 per cent lymphocytes, and 6 per cent monocytes.

(a) Electrocardiograms: The electrocardiograms (Fig. 1B) had pronounced right axis deviation of the QRS with a mean electrical axis of +160°; biphasic P-wave in lead I, with a mean electrical axis of 0°. The precordial transition was reversed, so that leads over the right precordium recorded upright QRS deflections and upright T waves, while downwardly directed ventricular deflections were recorded over the left precordium. Leads V₅ and V₆ were similar to the leads in V₅₆ and V₆₆ of normal hearts, and lead V₅₂ was similar to V₅ of a normal heart except for slight ST sagging.

When the left and right lead waves were reversed and the precordial leads were recorded from left to right over the right precordium, the electrocardiogram appeared normal. The electrocardiographic findings were consistent with the diagnosis of dextrocardia.

(b) Direct spatial vectorcardiograms were obtained utilizing Kimura, Grishman and Frank’s lead systems (Fig. 2). The initial forces of depolarization were oriented to the right, anteriorly and slightly superiorly. Progression was at a uniform rate to the posterior and inferior octants so that orientation of the mean depolarization force was in the inferior, posterior, right octant. The direct spatial vectorcardiogram demonstrated right ventricular hypertrophy.

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(c) X-ray films (Fig. 1C): A dextrocardia of normal size and configuration, with a left aortic arch and descending aorta were described. The pulmonary vascularity appeared slightly increased. A right asygos lobe was present. The lung fields suggested bronchiectasis. The stomach was on the left. Skull x-ray films showed a thickened right antral membrane as well as slight thickening of the right frontal membrane.

(d) Cardiac catheterization: The right ventricle was entered from the right atrium via the inferior vena cava. An enlarged right atrium was demonstrated. The catheter could not be advanced beyond the apex of the right ventricle because of runs of ventricular tachycardia. Elevated pressures were demonstrated in the right atrium (22/11) and right ventricle (98/4-7), with radial artery pressure of 100/70. Dye dilution studies showed a large left-to-right, as well as a smaller right-to-left shunt.

(e) Angiograms showed the heart in the right chest with a left aortic arch and a left descending aorta, a huge right auricle, or possible common auricle, and a moderate right ventricular hypertrophy. An interatrial septal defect was also suggested. Early visualization of the aorta was noted and its position anterior to the origin of the pulmonary artery was seen.

Course of Illness: A second hospital admission was necessary in September, 1958, because of episodes of fever, chills and occipital headaches which began two weeks after a tooth extraction. At that time, the physical findings were essentially the same as before. Treatment with antibiotics was instituted. Phlebotomy was performed because of his polycythemia. He was discharged eight days after admission as improved, only to be re-admitted three days later for lethargy, nausea, vomiting and severe headache. His temperature was 102°F. He appeared critically ill and in moderate distress. Nuchal rigidity and bilateral positive Kernig's signs were elicited. There was some question of weakness of the right leg and lower facial musculature.

![Figure 1: (a) Phonocardiogram: Right=50-200 frequency; Left=200-500 frequency; Line 1=right second intercostal space; Line 2=left second intercostal space; Line 3=apex. (b) Electrocardiogram. Normal progression of R waves across right precordium. (c) P-A chest x-ray film; consistent with dextrocardia and left aortic arch.](image)
Figure 2: (See text) (a) Kimura lead system; (b) Grishman lead system; (c) Frank lead system.

Figure 3: A=Aorta; P=Pulmonary artery; C=Posterior descending coronary artery. Figure 4: RA =Right atrium; RV=Right ventricle.
The total white blood count at this time was 26,000 per mm.\(^2\), and a skull x-ray film showed a shift of the pineal gland to the right. A left frontal lobe brain abscess was tapped through a left frontal burr hole. When the catheters were inserted, yellow ventricular fluid admixed with purulent material was obtained. Despite slight postoperative improvement, he became comatose and expired.

**Necropsy Findings**

The pertinent anatomic findings were confined to the heart, great vessels and brain. The abdominal viscera were normally situated and showed gross abnormalities. Other findings will be described briefly.

**Heart:** The aortic arch and aorta were to the left of the spine. The heart weighed 490 grams, was rotated counter-clockwise almost 180°, and the apex situated in the right thorax. The superior and inferior venae cavae were on the right side of the thoracic column and entered a large right atrium. The anterior aspect of the heart consisted almost entirely of right ventricle, with the left anterior descending coronary artery sulcus situated posteriorly and the posterior descending coronary artery situated on the anterior aspect (Fig. 3). Extremely prominent, the aorta appeared to arise from the left border and base of the heart. Its origin was to the left and slightly anterior to the pulmonary artery.

A superior, anterior septal defect, measuring approximately 1.5 cm. in diameter, was present between the atria. It resembled a widely patent foramen ovale (Fig. 6). The valve leaflets of the foramen could not be identified. The ostium of the coronary sinus was immediately to the right of the rudimentary interauricular septum. Between the right atrium and the right ventricle was a valve 14.0 cm. in diameter having two cusps (Fig. 4). The right ventricular chamber had an 11 mm. thick wall, approximately one-third of the thickness was composed of trabeculae carneae. A probe could easily be passed through a superiorly situated, 3 cm. interventricular septal defect and thence either into the aorta or into the pulmonary artery. A thin predominate fibrous band, measuring approximately 10 mm. in length and 5 mm. at its greatest width, separated the origins of the aorta and pulmonary arteries. The crista supraventricularis began at the anterior border of the ridge separating the pulmonary artery and the aorta and coursed along the anterior border of the interventricular septal defect into the lateral wall of the left-sided ventricle (Figs. 5A and B). A crista could not be identified in the right-sided ventricle. The pulmonary artery was moderately stenosed, had one rudimentary and one large, thick cup; was displaced to the left, and overrode the interventricular septal defect. A pulmonary conus could not be identified.

Two pulmonary veins, one from the left and the other from the right lung, entered the left atrium which was approximately one-fifth of volume of the right atrium. The right vein overlay the interatrial septal defect.
Between the left atrium and a small, almost rudimentary left-sided ventricle, was a deformed valve (Fig. 5). This measured 4.0 cm. in greatest dimension, was thick, had a single, web-like leaf, and only vaguely defined webbed chordae tendinae. The medial portion was attached to the inferior border of the interventricular septal defect. This valve was obliquely situated and appeared to form a portion of the inferior and posterior wall of the interventricular septal defect. The aortic valve was 10.0 cm. in circumference; the three aortic cusps were slightly thickened but otherwise normal. The anterior coronary artery had two ostia and the posterior, one large ostium. The left ventricular wall did not exceed 8 mm. in thickness.

The rudimentary pulmonary artery originated posteriorly and slightly to the right of the aorta, with the arch of the aorta and the descending aorta left of the vertebral column. A rudimentary ligamentum arteriosum replaced the ductus arteriosus.

Microscopically, the myocardial fibers appeared larger than normal. There was interstitial and perivascular edema. In some of the perivascular areas were small numbers of mononuclear cells. The endocardium was thickened.

Within the subendocardial fibrous tissue was a moderate sized artery which showed marked medial proliferation and luminal narrowing with early necrosis of a portion of its wall. A section of the deformed left atrioventricular valve showed an orderly arrangement of dense connective tissue throughout which were interspersed elastic fibers. Several vasa vasorum of the ascending aorta had thickened medias. There was no other significant abnormality.

Lungs: Bilateral hydrothorax, pulmonary hyperemia, and acute bronchial pneumonia were present. Thickening of alveolar walls was noted.

Abdominal organs: Hyperemia with chronic phlyonephritis.

Brain: Hyperemia and 3 x 3 cm. left frontal lobe abscess.

Discussion

This 35-year-old man had congenital heart disease characterized by isolated dextroversion associated with multiple intracardiac abnormalities, one of which was a complex consisting of deformity of the left ventricle, and the atrioventricular valve; a left-sided complex which is similar to the right-sided complex known as Ebstein's anomaly.

Ebstein's anomaly probably is related to the formation of the proximal bulbar cushion. In reviews of examples of Ebstein's anomaly, there are no reports of an Ebstein-like deformity of the mitral valve. Schiebler, et al. reviewed 40 cases of cor-
rected transpositions of great vessels and found no instances of a "left-sided Ebstein anomaly." It is expected, however, that there should be rare Ebstein-like malformations of the left A-V valve. Similar to ours are the cases reported by Edwards, Becu, et al., Kraus, and van Mierop, et al. As in our case, there was transposition of the great vessels and the right atrioventricular valve resembled a mitral valve. It was their opinion that the deformed left atrioventricular valves were actually transposed tricuspid valves.

It is important to differentiate mirror image dextrocardia which may be associated with some degree of abdominal situs inversus, dextroposition which is in effect a normal heart shifted to the right by some extracardiac factor such as a tumor, diaphragmatic hernia, etc., and dextroversion, which we believed was present in our patient and which is closely related to the embryogenesis of the conotruncal region of the heart. In dextroversion, although the heart lies to the right, the atria have a normal relation to the ventricles, spinal column and great veins. It is as if the ventricular heart had been swung like a pendulum through an arch of 120° with simultaneous counter-clockwise rotation on its long axis. In this anomaly, a commonly associated intracardiac abnormality is transposition of the great vessels. This was also present in our patient. Transposition signifies an anomalous course of formation of the spiral septum during division of the conus and truncus into the aorta and pulmonary artery. In normal development, the spiral or bulbar septum divides the conus and truncus into the pulmonary artery and the aorta in spiral fashion so that they grow and meet the normally situated ventricles. The spiral septum, in addition, aids in the completion of the interventricular septum. In transposition of the great vessels, however, the spiral septum does not develop normally and may follow a straight course, result in an unequal division of the conus and truncus and a pulmonary stenosis, as in our patient. It would appear that his primary embryogenetic fault lay in development and partitioning of the conotruncal region with a compensatory dextroversion and counterclockwise rotation of ventricles. However, the left atrium emptied into a rudimentary left-sided ventricle via an atrioventricular valve which was abnormal in size, shape and origin; a portion of the ventricle appearing in the atrium. The abnormality of the left atrioventricular valve, therefore, caused an Ebstein-like malformation of the left chambers.

The embryogenesis of Ebstein's anomaly is unknown, but there are several possible explanations. One of these suggests that there may be an abnormal downward displacement of atrioventricular cushion material during the second period of valve development. Another, which may apply to the anomaly in our patient, suggests that the right proximal bulbar cushion could form a downward displaced lateral endocardial cushion of the tricuspid valve. If this explanation is valid, it may explain the anomaly of the left atrioventricular valve in our patient. Since there was an abnormality in formation and course of the bulbar septum associated with transposition of the great vessels, and secondary dextroversion of the heart, it is entirely conceivable that the proximal bulbar cushion would, instead of fusing with the endocardial cushions which form the right atrioventricular valve, fuse with the partially formed cushions or valve structures of the left atrioventricular valve. The crista bordered the anterior portion of the interventricular septal defect and appeared to touch the most medial and inferior portion of the deformed mitral valve. The levoposition of the aorta and pulmonary artery, the septum dividing these vessels, and the crista supraventricularis to the left of the large interventricular septal defect lend support to this suggestion.

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REFERENCES

PREDNISONE IN PULMONARY EMPHYSEMA

The effect of prednisone on air-flow obstruction in ten patients with severe emphysema was evaluated in a double-blind, controlled manner. The volume expired during the first second of the vital capacity was used as an index of air flow obstruction. Variance analysis of the control data showed that each patient was stable, with a small standard deviation of one observation. Prednisone did not produce a statistically significant increase in timed vital capacity when the results in the ten patients were analyzed together. Two patients of the ten, however, appeared to behave as if they were members of a different population in that they showed significant increases in one-second vital capacity during steroid treatment. A comparison between the results of prednisone treatment and a standard history obtained during the control week suggested that steroids might benefit the patient who had had his disease for a long time, who began with asthmatic symptoms and who did not exhibit a relentless downhill course.


ASPERGILLOMA OF LUNG WITH SARCOID REACTION IN LYMPH NODES

A case is described of a man, aged 35, with a healed primary complex. aspergilloma of the lung and bilateral enlargement of hilar nodes. Right upper lobectomy was done. In the specimen, an aspergillum and fungus in the small broncholites and in pulmonary parenchyma were found. Six months after the operation, bronchoscopy revealed numerous colonies of A. fumigatus in the stump of the right upper bronchus which were the cause of repeated hemoptysis. A stude was removed bronchoscopically from the bronchial stump and a series of intrabronchial instillations of Nystatin were made. The patient recovered. It is suggested that sarcoid reaction found in the lymph nodes was a reaction to the fungal infection.


BRONCHIAL POLyps

Two patients are presented in whom acute respiratory illness was due to obstruction of a lower lobe bronchus by a polyp. Both tumors were removed at bronchoscopy with good clinical results. In one patient, the polyp was regarded as solitary phenomenon of unknown etiology and the prognosis was assessed as good. In the other, the lesion was regarded as an unusual manifestation of chronic inflammatory disease of the bronchial tree.