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Disturbance of Conduction System in Corrected Transposition of the Great Vessels

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Introduction

Numerous reports have been made on corrected transposition of the great vessels since it was first described by Von ROKITANSKY[14] in 1875. In Japan, this cardiac anomaly has been arousing interest, though thought to be of rare occurrence.

Cases of corrected transposition of great vessels requiring treatment have concomitant cardiac malformations that give rise to hemodynamic abnormalities, i. e. ventricular septal defect, pulmonary stenosis, left-sided atrioventricular valve insufficiency and atrial septal defect, together with disturbances of conduction system, such as arrhythmias and atrioventricular block.

Among cases of corrected transposition of the great vessels treated by us recently, 2 with complete atrioventricular block had a favorable course with an implanted pacemaker, while in another with first degree atrioventricular block the patient died suddenly while under medical supervision. The present study explored impairment of conduction system associated with corrected transposition of the great vessels.

Report of cases

Case 1, H. K., a 49-year-old woman. The patient was notified of her having a suspected congenital heart disease, as a primary school-girl. However, as she felt no difficulty in her daily activities, she underwent no further exploration.

In 1965 she began to feel out of breath on going upstairs and developed cyanosis of lips and fingertips during cold seasons.

Key words　Corrected transposition of great vessels, Arrhythmia, Complete A-V block, Implantation of pacemaker, Left-sided atrioventricular valve insufficiency.
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In November, 1974 she was admitted to hospital for thorough examination. Examination on admission revealed a moderately nourished, middle-aged woman whose lips and fingertips were not cyanotic. There was no discernible deformity of the chest nor were any thrills palpable. Levine 2nd degree systolic and diastolic murmurs were audible over an area extending from the sternal border of the left 4th intercostal space to the apex of heart and accentuated 2nd heart sounds were heard at the sternal border of the left 2nd intercostal space. The pulse rate was 74 per minute, regular; the blood pressure was 130/90 mmHg.

On chest X-ray the left 1st cardiac arch was bulged and the left hilus was overshadowed for the most part by the heart, with the cardiothoratic index being 62%. No displacement of the esophagus was noted on oblique view.

The ECG pattern was typical of first degree atrioventricular block with the predominance of sinus rhythm, mean electrical axis $-30°$ and a prolonged PQ interval of 0.24 sec (Fig. 1).

Attempt to catheterize the pulmonary artery was unsuccessful (Table 1). On cardioan-

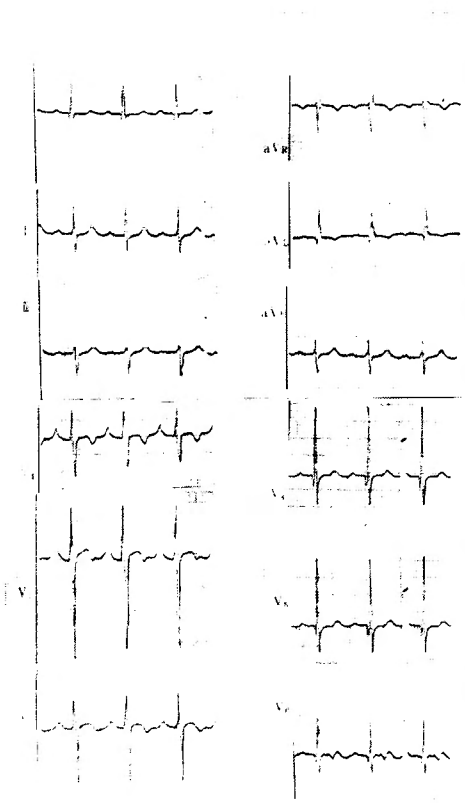

Fig. 1 ECG showed a prolonged PQ interval of 0.24 sec.

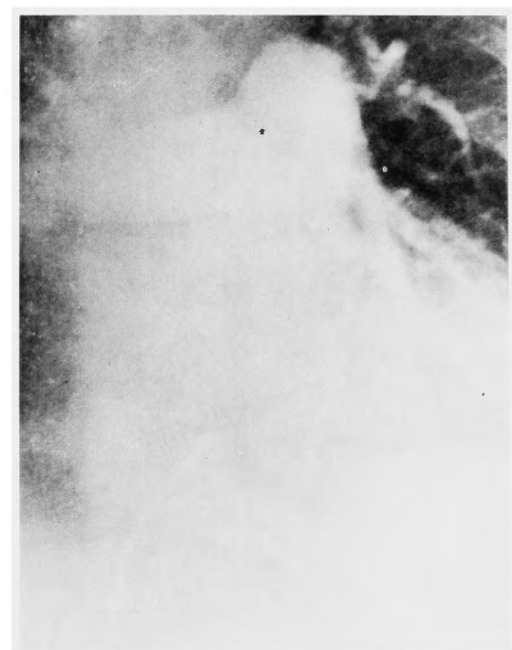

Fig. 2 the right-sided ventricle was smooth-walled and looked like an anatomical left ventricle.

Table 1 Cardiac catheterization of case 1

	O_2 Vol %	Pressure mmHg
SVC	12.60	4
IVC	12.72	4
RA	12.72	4
RV	11.63	40/4
PA		
FA	14.23 (90.7%)	120/74

giography the origin of the pulmonary artery was displaced centrally and the ventricle was smooth-walled and looked like an anatomical left ventricle (Fig. 2).

At night of April 8, 1978 she developed a syncopal attack. On the following day three similar, but mild attacks occurred. She was noticed as having a bradycardia of 30-40 beats per minute by a neighbouring doctor. She died of a subsequent syncopal attack and dyspnea early in the morning of April 10.

Case 2 : Y. U., a 56-year-old man. Two years previously the patient began to have palpitation and dyspenea during exercises and while going up and downstairs. Afterwards his condition became progressively worsened to cause orthopnea usually at night. At the time of hospitalization, Levine 2-3 degree systolic and diastolic murmurs were heard at the

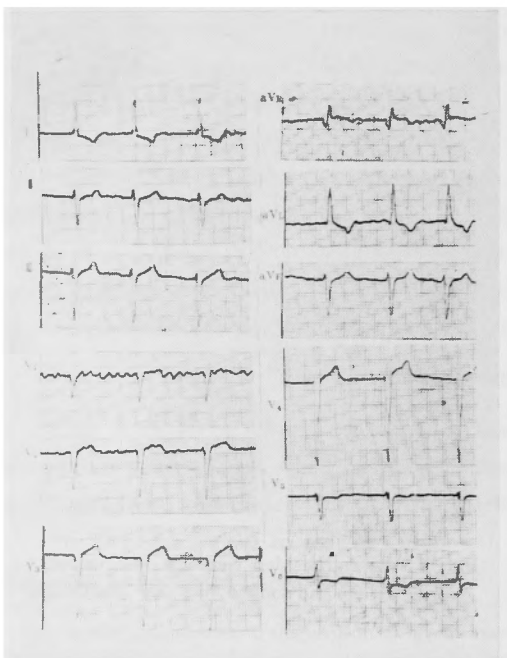

Fig. 3 ECG demonstrated atrial fibrillation and complete A-V block.

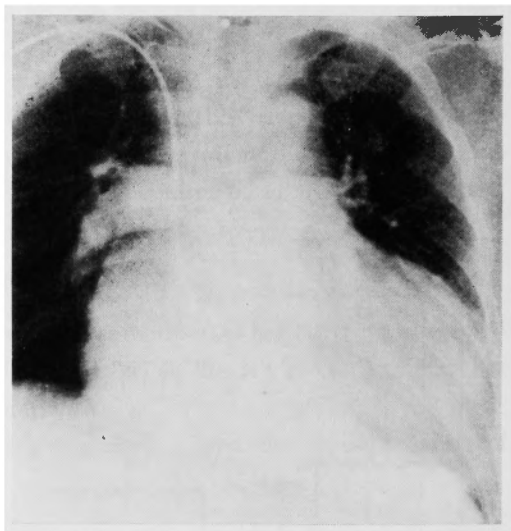

Fig. 4 the right-sided ventricle

apex. The liver was palpable 4 finger-breath below the costal margin. There was no thoracic deformity.

On chest X-ray the left and right inferior cardiac arches were markedly enlarged and no pulmonary arterial conus was noticeable on the left margin of the cardiac silhouette. Pulmonary vascular markings were increased. ECG study demonstrated atrial fibillation and left axis deviation with QRS axis located at $-30°$; the pattern was of QS type in right precordial leads, while the Q wave was absent in tracings obtained in left precordial leads (Fig. 3).

Cardioangiography disclosed an enlarged left atrium and a right ventricle, structurally identical with the anatomical left ventricle (Fig. 4). On coronary arteriography the right coronary artery was found to be bifurcate while the left coronary artery was non-bifurcate (Fig. 5).

Sellers 2 to 3 degree atrioventricular valve insufficiency was noted.

For the control of bradycardia an artificial pacemaker was used, which brought about improvement of cardiac failure. As the placement of a catheter electrode of the pacemaker via the vena cava was unfeasible, a myocardial electrode had to be employed instead. After improvement of cardiac failure, annuloplication was performed for the left atrioventricular valve insufficiency.

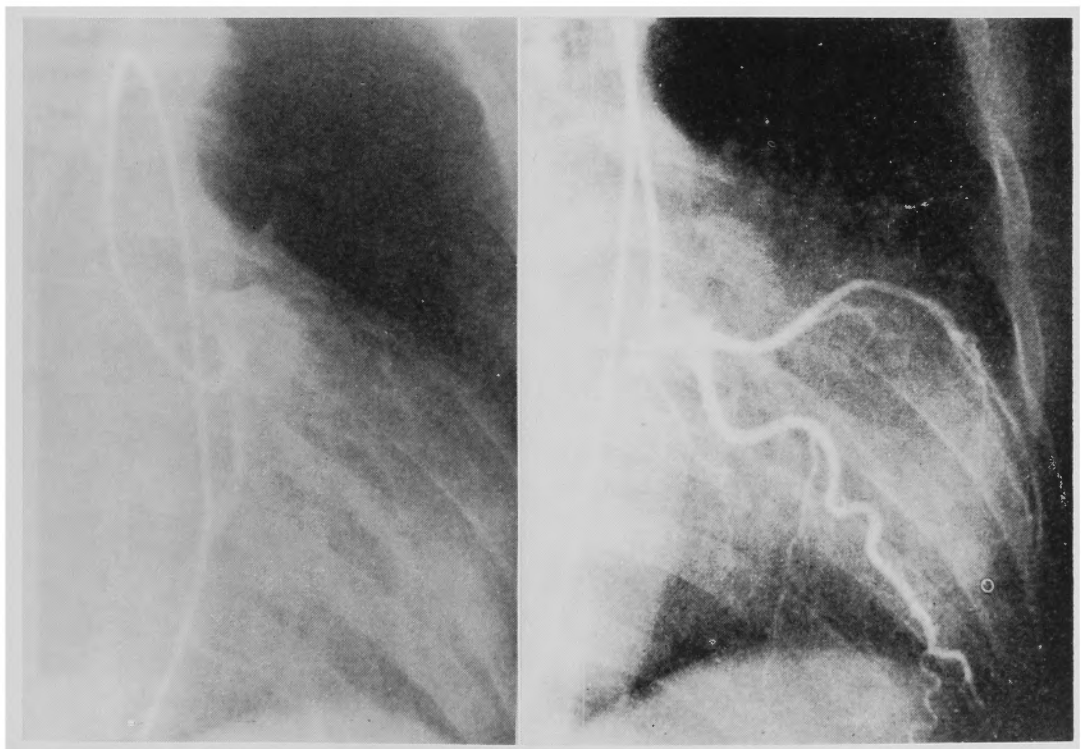

Fig. 5 the right coronary artery was found to be bifurcate while the left coronary artery was non-bifurcate.

Case 3 : T. H., a 51-year-old man. Twelve years ago the patient was first noticed of his having cardiac murmurs and bradycardia by a doctor, who diagnosed syncope the patient had frequently developed to be due to Adams-Stokes attacks. At the time of hospitalization, there were cyanosis, hepato- and splenomegaly, but edema was absent.

On auscultation Levine 4th degree pansystolic murmurs, the loudest at the apex and radiating to the axilla, were audible. Accentuated 2nd pulmonary arterial sounds were heard along the left 2nd intercostal space. ECG study demonstrated complete A-V block with atrial fibrillation (Fig. 6).

Cardioangiography revealed a displacement of the origin of the pulmonary artery toward the midline and a smooth-walled right-sided ventricle which structurally resembled the anatomical left ventricle (Fig. 7), A pacemaker was used with an electrode sutured to the myocardium. Left-sided ventriculography demonstrated Sellers 3rd degree atrioventricular valve insufficiency, for which valve replacement was performed after symptomatic improvement was obtained.

In case 2 and 3, as mentioned before, the placement of a catheter electrode in the right-sided ventricle for the implantation of a pacemaker was impossible due to a structural similarity of the ventricle to the anatomical left ventricle.

Failure of electrode onto the endocardium may thus be regarded as presumptive evidence of the disease in question.

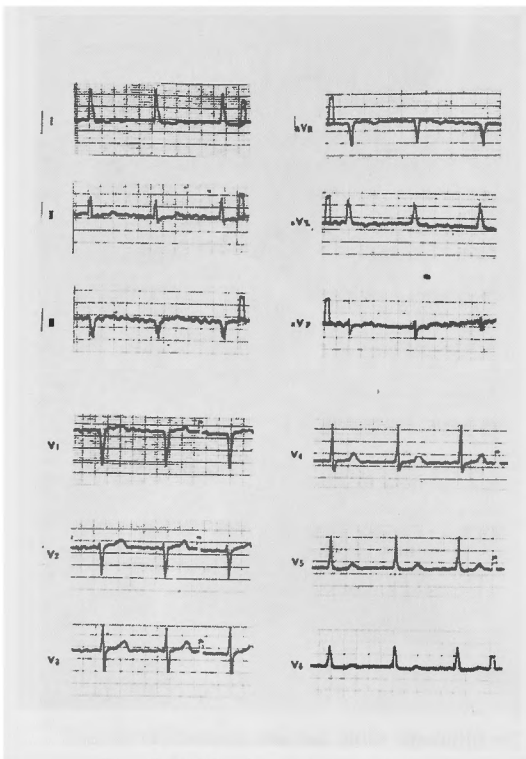

Fig. 6 ECG of complete A-V block

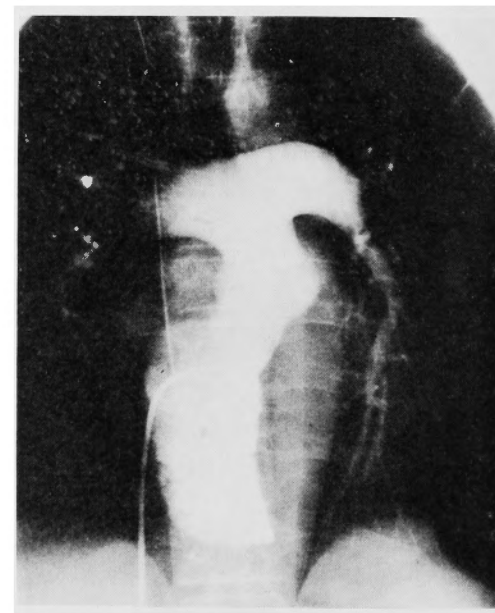

Fig. 7 the right-sided ventricle

Discussion

Corrected transposition of the great vessels was classified into 4 types by CARDELL[5], i. e., B 1 (with bulbus inversion), B 2 (with sinoatrial and ventricular inversion), B 3 (with bulboventricular inversion) and B 4 (with sinoatrial inversion). It is a congenital cardiac anomaly in which the inverted aorta receives arterial blood and the inverted pulmonary artery receives venous blood, thereby the transposition being compensated for physiologically. Therefore, this anomalous condition does not become a therapeutic problem unless there are cardiac complications that give rise to hemodynamic abnormalities.

One of the anatomical features of the condition is the right-sided atrioventricular valve being bicuspid. Its anterior cusp is attached to the pulmonary artery valve annulus and constitutes the anterior walls of the in- and out flow tracts in the right-sided ventricle (the outflow tract thus being extremely short). Relatively few trabeculae are formed on the inner surface of the ventricle as in the normal left ventricle and the surface of the interventricular septum is smooth.

On the other hand, the left-sided atrioventricular valve is tricuspid and there are large trabeculae within the ventricle. The supraventricular crista and infundibulum are also present.

The upper end of the upper margin of the interventricular septum, which normally is located beneath the right atrium, is situated beneath the left atrium in this anomalous condition. The transposition of this structure has a great influence upon the conduction system. Thus, in this anomaly, whereas the atrioventricular node and the bundle of His are located on the side of the right atrium as they are in the normal heart, the bundle branches are inverted together with the ventricles; as a consequence, the course of the conduction system is very much lengthened. The anatomical left bundle branch is located on the right side and hence the periphery of the bundle of HIS and right bundle branch have to run a long course to reach the left sided interventricular septum (Fig. 8).

The coronary artery also runs in a peculiar pattern of course which is just a mirror-image of that of the normal heart (Fig. 9).

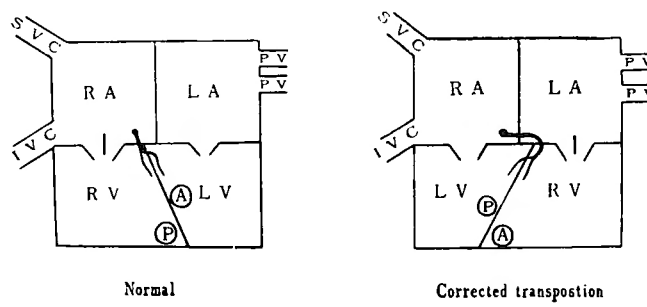

Fig. 8 (SAYED : J. Thorac. Cardiovasc. Surg., 44 : 446, 1962.)

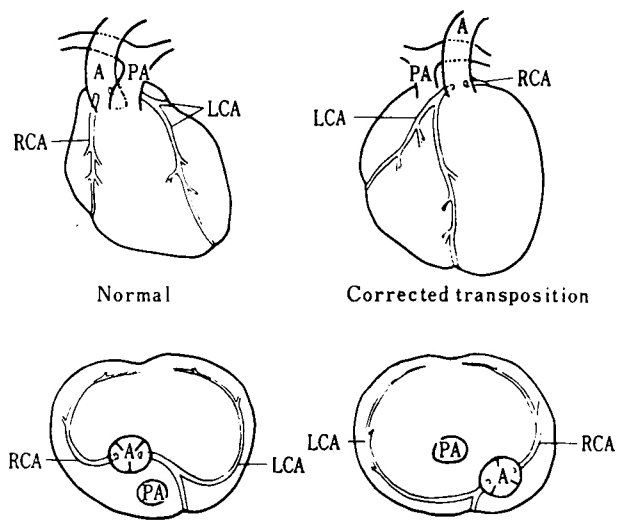

Fig. 9 (CARDELL[5] : Brit. Heart J., **18** : 186, 1957.)

Disturbance of conduction and arrhythmias in corrected transposition of the great vessels have been investigated by many researchers. ANDERSON[1] and SHEM-TOV[2] described the appearance of Q wave in right precordial leads, reversal of the normal precordial Q wave pattern, tall P wave in standard lead Ⅱ and positive T wave in all precordial leads as specific ECG correlates of inversion of the ventricles.

Positive T wave occurring in right precordial leads are seen in diseases with right ventricular overload also and reversal of the normal precordial Q wave pattern in leads Ⅰ, aVL and V5, V6 is reported to be seen in 0.5% of healthy individuals[3,4]. Accordingly, these two ECG features, only when combined, are of diagnostic evidence.

The pathology and pathogenesis of the impairment of conduction still remains obscure since even the anatomical exploration of the conduction system per se is fraught with many difficulties. YATER[17] recognized rupture of the atrioventricular node in one case of corrected transposition of the great vessels with associated atrioventricular block. WALMSLEY [15] noted fibrosis of the bundle of HIS which he implicated as a cause of the disturbance of conduction in the congenital anomaly. He suggested that elongation of the conduction system resulting from inversion of the ventricles might also be a possible cause of the impairment of conduction.

ANDERSON, et al.[1], implicated elongation of the conduction system due to ventricular inversion as the main cause of disturbance of conduction. LEV, et al.[9,10], pointed to the possibility that the inverted or bilateral conus might cause enlargement of the membranous portion of the ventricular septum which in turn results in elongation of the conduction system with a consequent occurrence of atrioventricular block.

According to Friedberg and NADAS[6], impairment of conduction was noted in 19 cases (32%) in their series of 60 cases, with 2 of these 19 having second degree atrioventricular

block and 7 cases with complete atrioventricular block, 3 had a fatal outcome; one of the remaining 4 had a pacemaker implanted. In this series there were cases in which heart block, progressing from first to third degree with advancing age, occurred in patients who were apparently normal at birth. Six cases had intermittent episodes of wandering atrial pacemaker.

FONTANA, et al.[7] reported the occurrence of complete atrioventricular block in 2 (with one of them having onset immediately after birth) of 6 cases of corrected transposition of great vessels with no other concomitant anomalies. Atrioventricular block was present in 12 of 45 cases so far reported in Japan[2].

The atrioventricular block may be permanent or paroxymal. Cardiac catheterization in patients with the anomaly may give rise to supraventricular tachycardia and ventricular fibrillation which may develop into complete atrioventricular block[6,8,13,16]. Fatal complete atrioventricular block may reportedly occur during or after surgery for some concomitant anomaly and various arrhythmias may also become a clinical problem[11]. Different types of disturbance of conduction may occur at different times in the same case. Disturbance of atrioventricular condition and heart block are said to occur and progress with advancing age. The mechanism underlying a sequence of events from a progressing block to Adams-Stokes attack or even sudden death is not as yet clearly understood.

However, it is assumed that the diminished function of the anatomical right ventricle disproportionate to left-sided ventricular work causes low cardiac output due to bradycardia to be lessened further, thereby leading to aggravation of the disease state.

Under such circumstances the use, therapeutic or prophylactic of an artificial pacemaker is highly desirable. In the three cases presented here, the patients were middle-aged or presenile and had diminished cardiac function with 2 of them having a concomitant left-sided atrioventricular valve insufficiency.

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和文抄録

修正大血管転位症に合併する刺激伝導障害

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高令者に属する修正大血管転位症3例で，2例には完全房室ブロックを認めペースメーカーを移植し良好な経過をとったが，1例は1度の房室ブロックから完全房室ブロックに移行したためか，4年後に突然死をみた．

修正大血管転位症には，房室ブロック，その他多様な不整脈が発作性にまたは永久的にあらわれる．しかも加令に従い完全房室ブロックなどに進行するといわれている．

厳重な監視を要するとともに積極的なペースメーカーの使用が望まれる．