A Case of Interventricular Septal Defect with Dextrocardia and Situs Inversus Treated by Surgery

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A case of interventricular septal defect associated with situs inversus and dextrocardia in twenty year old male patient was presented along with discussion on three types of dextrocardial complexes and with emphasis on the rarity of occurrence of the intracardiac anomalies in true "mirror-image" dextrocardia with total situs inversus. A comment was also made on Kartagener's syndrome which was excluded by appropriate radiographic procedures in this case.

The closure of the interventricular septal defect with extracorporeal circulation was successfully performed under moderate hypothermia.

DEXTROCARDIA WITH complete situs inversus is considered by some not to be an anomalous condition, since there is only a reversal of the circulatory path with no interference in function.

The occurrence of acquired heart disease in these individuals probably is the same as in the general population, but they are very often accompanied with other congenital cardio-vascular anomalies.

The following case illustrates an interventricular septal defect associated with dextrocardia and complete situs inversus, and the successful closure of the interventricular septal defect under direct vision.

CASE REPORT

The patient, a 20 year old male, was well until 12 years of age, when he consulted a doctor about slight fever and cough, and was told to have a cardiac failure.

In July 1960, he was hospitalized, because of fever, fatigability, headache, palpitation, cyanosis and edema. He was found to have situs inversus and congenital cardiac

disease, and was referred to our clinic.

On admission, physical examination revealed no cyanosis, no anemia and no clubbing of fingers. The right anterior chest was slightly protruded. The apical pulsations were visible in the fourth intercostal space in the right mid-clavicular line at a rate of 100 with occasional irregularity. A loud harsh systolic murmur was audible at the right fourth intercostal space, left parasternal margin, and a coarse systolic thrill was palpated at the same place. The second pulmonary sound was not accentuated. Neither rales nor wheezes were heard in both lungs. There was no peripheral edema or palpable liver.

The blood pressure was 142/70 mm Hg. Routine blood examination revealed slight anemia, but both urinalysis and biochemical blood examinations were within normal limits. Serological tests were negative.

Electrocardiograms on admission and during the hospital stay were consistent with dextrocardia and showed bilateral ventricular hypertrophy. (Fig. 1)

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PRE-OPERATIVE E.C.G.

Fig. 1 Preoperative Electrocardiogram: Lead I complexes are inverted; the patterns in II-III and $aV_R - aV_L$ are the reverse of the normal. Left precordial leads reflect right ventricular epicardial potential; right precordial leads reflect left ventricular epicardial potential. Both right and left ventricular hypertrophies are noted.

Fig. 2. a. Posteroanterior chest roentgenogram. b. Right anterior oblique projection. c. Left anterior oblique projection. d. Lateral projection.

Four views of the chest demonstrate right-sided position of the cardiac silhouette. Moderate cardiomegaly is noted with definite enlargement of the right ventricle. There appears to be some enlargement of the left ventricle as well. No appreciable enlargement of the left atrium is noted. There seems to be slight prominence of the pulmonary artery segment. Pulmonary vascularity is not definitely increased. Aorta is hypoplastic.

X-ray films revealed dextrocardia and situs inversus. The apex was to the right, slightly down above the apex of the diaphragm. The pulmonic area in the postero-anterior views was straight. The lung fields were clear and no congestive changes were noted. The aortic knob was on the right. Cardiac silhouette was moderately enlarged to the right. (Fig. 2) Bronchography showed that right lung was consisted of three lobes,

Fig. 3. Bronchogram demonstrated mirror image of the normal bronchial distribution of the lung without evidence of bronchiectasis.

Fig. 4. The pattern of the stomach and small intestine forms a mirror image of the normal. No radiographic evidence of intrinsic or extrinsic organic lesion can be seen.

Fig. 5. Examination of the colon also shows a mirror image of the normal. Some redundancy of the lower descending and sigmoid colon is incidentally noted.

Fig. 6. Caldwell's (a) and Water's (b) projections for the paranasal sinuses show the maxillary, ethmoid, frontal and sphenoid sinuses to be well pneumatized without radiographic evidence of sinusitis.

left lung two lobes, and the aortic arch rode over the right main bronchus. There was no sign of bronchiectasis. (Fig. 3)

Barium swallow showed the stomach on the right side, and barium enema showed mirror image position of the large bowel

and no intrinsic organic lesions of the colon, (Fig. 4, 5)

Various projections of nasal sinusis showed no radiographic evidence of sinusitis. And Kartagenar's syndrome was ruled out. (Fig. 6)

Right cardiac catheterization showed moderate hypertension and high blood oxygen capacity of venous chambers suggesting presence of left to right shunt. (Fig. 7)

From these examinations, diagnosis of interventricular septal defect associated with dextrocardia and situs inversus was made.

Under extracorporeal circulation with mild hypothermia with the aid of our Thermo-Disc Oxygenator, cardiectomy was carried out in June, 1962, by Professor J. Wada.

Fig. 7. Posteroanterior chest roentgenogram taken during the right heart catheterization shows the catheter tip situating in the right ventricle. The presence of left-to-right shunt at the ventricular level was demonstrated with higher oxygen saturation in the right ventricle as compared to the right atrium, and increased right ventricular and pulmonary artery pressure.

(The above descriptions for Fig. 2 & 7 are given as in a case of usual levocardia for convenience' sake.)

Sternal splitting and pericardiotomy revealed dextrocardia and moderate dilatation of the right ventricle with systolic thrill in its out-flow tract radiating to the pulmonary artery. The main pulmonary artery was larger than the aorta, but did not show any evidence of pulmonic valvular or infundibular stenosis.

Right ventricular incision showed a membranous type interventricular septal defect of about 1.0 cm in diameter. This was completely closed with interrupted silk sutures. Cardiomyotomy was then sutured, followed by termination of the extracorporeal circulation.

During the procedure, normal sinus rhythm was maintained. Immediately after the total bypass, spontaneous respiration was recovered, systolic blood pressure was over 80 mmHg and clear consciousness returned.

Postoperative course was essentially uneventful and he was discharged back to his home with no abnormal cardiac murmur on the 28th postoperative day.

DISCUSSION

Dextrocardia, right-sided position of the heart, has been recognized for centuries.

There are three types of dextrocardial complexes, and the commonest and most familiar is the mirror image dextrocardia. This inversion of the cardiac chambers is such that the normal anterior position of the right atrium and right ventricle is preserved while there is a complete inversion of the relationships in the frontal plane. It is practically always a part of total situs inversus, in which the position of all the viscera is inverted from right to left. Situs inversus may be partial and the heart may be inverted with only a few of the other viscera.

The second type of dextrocardia is dextroposition, in which an otherwise normal heart is shifted to the right by some associated extracardiac lesions such as eventration of the left diaphragm, fibrosis of the right lung, etc., and should not present any great diagnostic problems.

The final type is the dextrocardia consisting of a rotation of the ventricular part of the heart to the right with the atria remaining in normal position. This type of dextrocardia is frequently accompanied by other intracardiac abnormalities, usually transposition of the great vessels and a ventricular septal defect.

Rarely one sees a levocardia in situs inversus. The heart is then the seat of a severe combination of anomalies.

On the other hand, mirror image dextrocardia for the most part is unassociated with other intracardiac anomalies, and a case of interventricular septal defect associated with situs inversus and dextrocardia such as the case presented in this paper seems to be exceedingly uncommon.

Not infrequently, the combination of dextrocardia with hypertrophic rhinitis, nasal polyposis, chronic sinusitis and bronchiectasis has been referred to as Kartagener's syndrome which was ruled out by appropriate radiographic procedures in our case.

SUMMARY

A case of interventricular septal defect associated with situs inversus and dextrocardia in an adult male is presented with particular emphasis on the rarity of occurrence of the intracardiac defects in true "mirror image" dextrocardia with total situs inversus.

The successful closure of the interventricular septal defect with extracorporeal circulation was carried out under moderate hypothermia.

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