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Surgical Treatment of Common Atrium Associated with Asplenia, with Special Reference to the Abnormal Conduction System

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The wide variety and the severity of congenital heart diseases associated with asplenia are well known. In most cases death occurs in th neonatal period or during infancy. Reports of successful surgery are very rare[1]. Recently we performed open heart surgery on a 3 year-old boy with asplenia. He had a common atrium, visceral situs anomaly, abnormal entry of the infenior vena cava and an abnormally situated atrioventricular node which was detected by His bundle mapping tecnique during open heart surgery.

As far as we know, there has been no report of such an abnormal conduction system. This paper describes this case in detail

Case report

A 3 year-old boy was first seen at the Kyoto University Hospital in November, 1975. Delivery had been normal after a full term pregnancy. There was no cyanosis, but he had had frequent upper respiratory tract infections from birth on.

Growth was retarded, physical activity was moderately limited and cyanosis appeared gradually. A primum atrial septal defect was suspected, and he was digitalized and followed in the out-patient clinic until 3 years of age when he was first catheterized and refered to our surgical department.

Key words : Asplenia, common atrium, abnormal conduction system, Howell-Jolly bodies, bundle of His.
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Preoperative study

The height of this boy was 97.4 cm and the body weight was 13.5 kg. This small boy had mild cyanosis. Hematological study showed a increased red cell count (605 $\times 10^4/mm^3$) and hemoglobin value (14.7 g/dl). HOWELL-JOLLY bodies were seen in the peripheral blood smear (Fig 1). Other laboratory tests, including liver and renal function, were within normal limits.

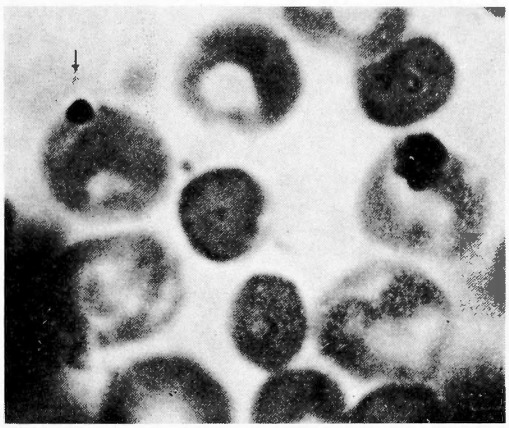

Fig 1 HOWELL-JOLLY Bodies in the Peripheral Blood Smear. Arrow shows HOWELL-JOLLY bodies in erythrocytes

The liver was palpable 2 cm below the left costal margin. The lungs were clear. The heart was hyperactive. There was a grade 3/6 systolic ejection murmur along the left sternal border.

The 2nd heart sound was split, and the pulmonary component was pronounced. A roentgenogram of the chest showed pulmonary congestion and cardiac enlargement. The cardio-thoracic ratio was 0.65 (Fig. 2). There was evidence of situs ambiguus with levocardia. The stomach bubble was on the right side and the liver shadow was rather symmetrical.

The electrocardiogram showed arrhythmia with atrio-ventricular junctional beats. The electrical axis of QRS was -110 degrees.

Technesium scanning proved the presence of the liver, but not of the spleen (Fig 3).

Right heart catheterization via the saphenous vein showed absence of the inferior vena cava with azigos connection (Fig 4), moderate pulmonary hypertension ($P_p/P_s=0.5$) and dominant left to right atrial shunt ($Q_p/Q_s=2.0$). Arterial blood oxygen saturation was 80%. The branching of the pulmonary arteries demonstrated by selective right ventroculography strongly suggested bilateral trilobulated lungs (Fig 4), although bronchography was not done.

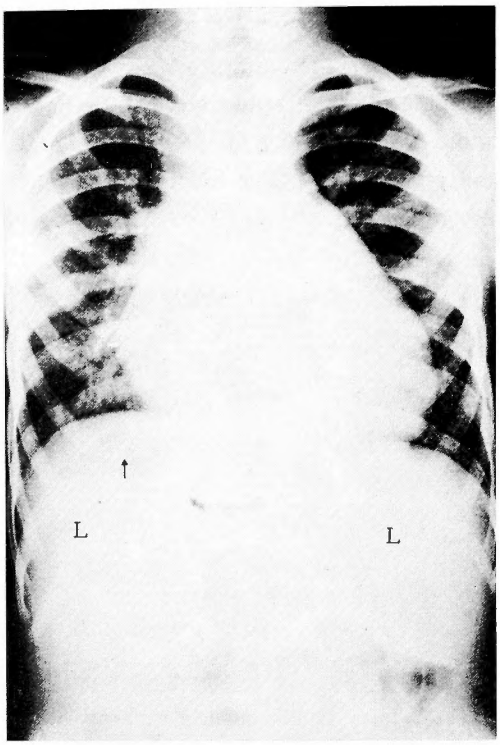

Fig 2 Preoperative Chest x-ray. Increased pulmonary vasculature, enlarged heart (Cardio-thoracic ratio =0.65) and levocardia are seen. The stomach bubble is on the right side (arrow). Liver shadow is symmetrical (L.).

The levogram of this series showed a large single atrium (Fig 5). Selective left ventriculograms showed slight mitral regurgitation and goose neck appearance of the left ventricular outflow tract. The aortic arch was on the left side (Fig 6).

The final preoparative diagnosis was asplenia associated with situs ambiguus with levocardia, absent inferior vena cava, azigos connection, common atrium, atrioventricular canal defect and supraventricular arrhythmia of unknown origin.

Operative method and Finding

Operative repair via median sternotomy under extracorporeal circulation was performed on August 2, 1978. The atrial septum was completely absent, and a clef anterior mitral cusp was seen. The abnormally located coronary ostium drained into the hepatic vein ostium below the right atrium. It was very difficult to guess the course of the conduction system in this situation. Therefore, we attempted to identify the exact location of the conduction system by an electrophysiological mapping technique. This revealed that the atrioventricular node was in the hepatic vein ostium, and the

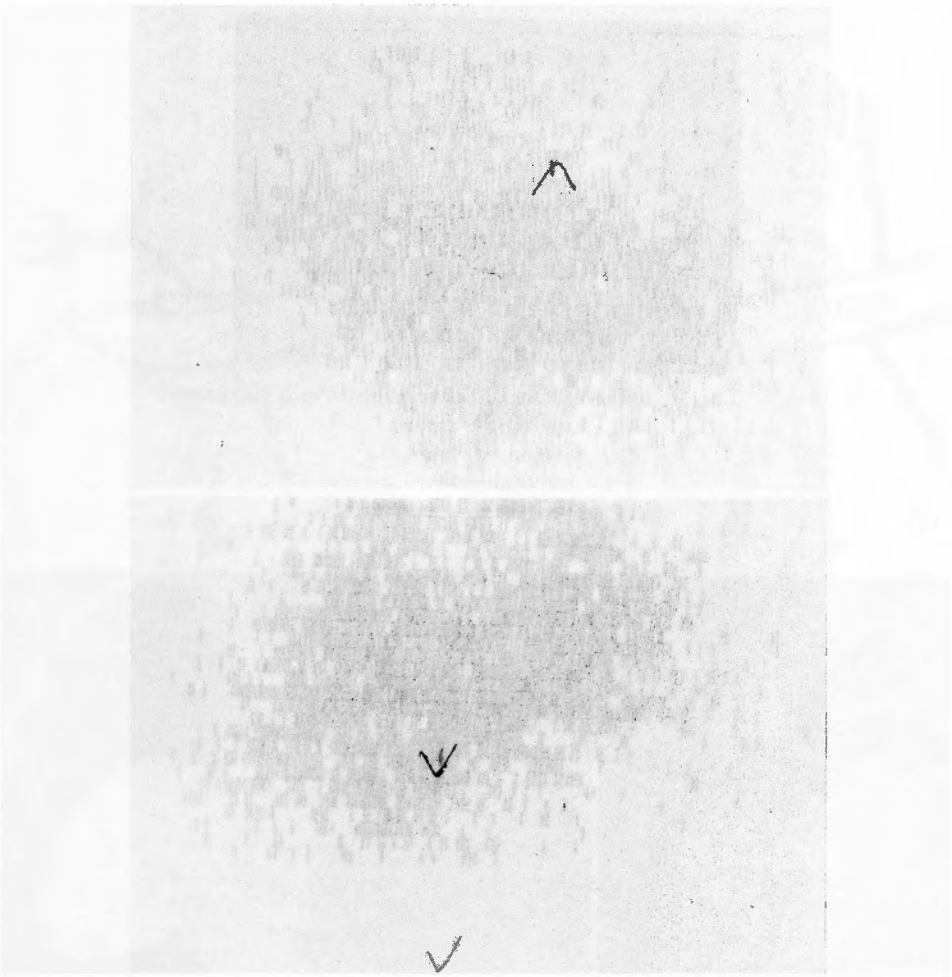

Fig 3 Scintigram Using Technesium (^{99m}Tc) A: postero-anterior B: antero-posterior Presence of the liver (rather symmetrical) and absence of the spleen are proved.

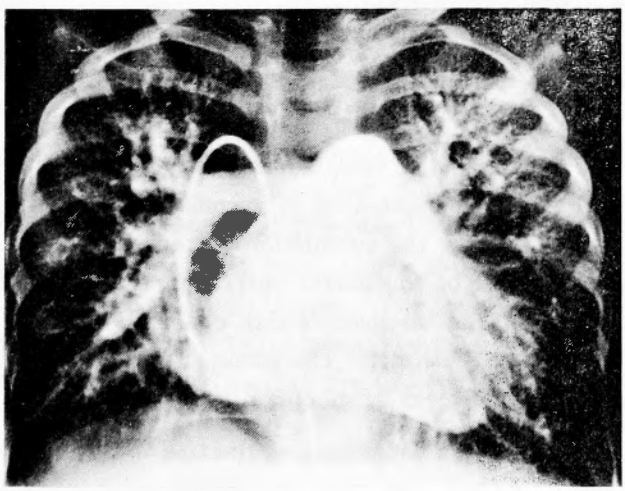

Fig 4 Selective Right Ventriulography The couse of catheter which is introduced via saphenous vein shows absence of inferior vena cava and azigos connection. Branching of the pulmonary arteries strongly suggests bilateral trilobulated symmetrical lungs (bilateral right- sideness).

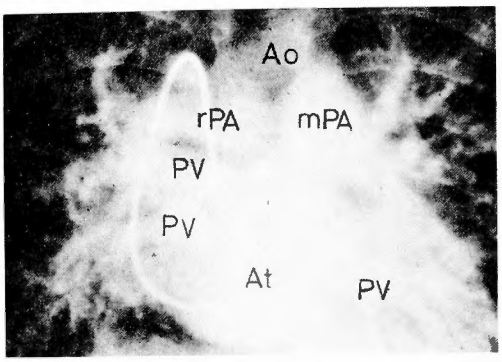

Fig 5 The levogram of the Selective Right Ventriculography
At : large single atrium,
PV : pulmonary veins,
rPA : right pulmonarv arterv,
mPA main pulmonary artery,
Ao : aorta

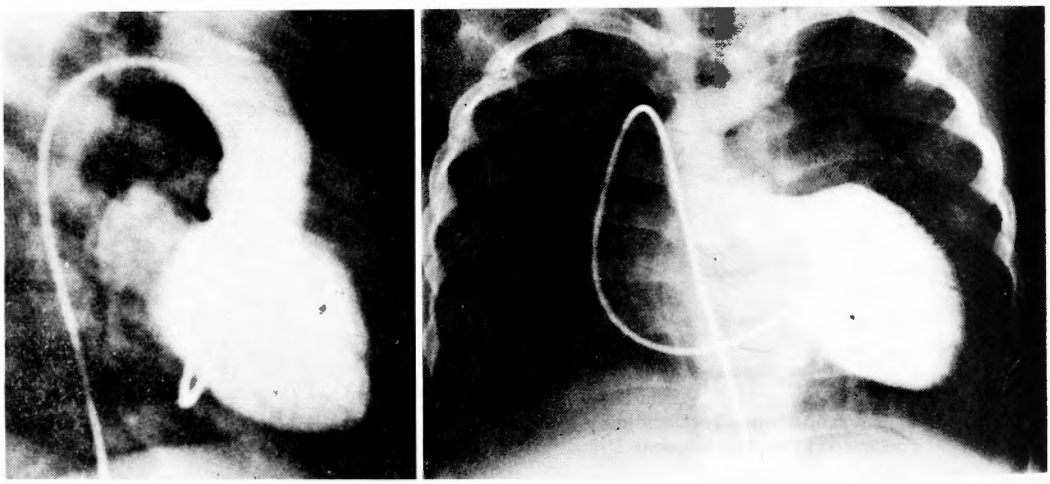

Fig 6 Selective Left Ventriculography
Typical goose neck appearance and left aortic arch are shown in the antero-posterior projection. Slight mitral regurgitation is seen in the lateral projection.

bundle of His ran from here to the ventricular crest via the right atrial-hepatic vein junction. Therefore, it appeared impossible to reconstruct the atrial septum in the usual manner without damaging the conduction system (Fig 7).

We first sutured the cleft of the mitral valve and created an atrial septum with a dacron patch partitioning the common atrial chamber so that the hepatic vein drained into the left side of the heart. The postoperative course was uneventful, and the patient was discharged 3 weeks after operation.

Postoperative evaluation

He was readmitted 3 months after surgery for postoperative evaluation including

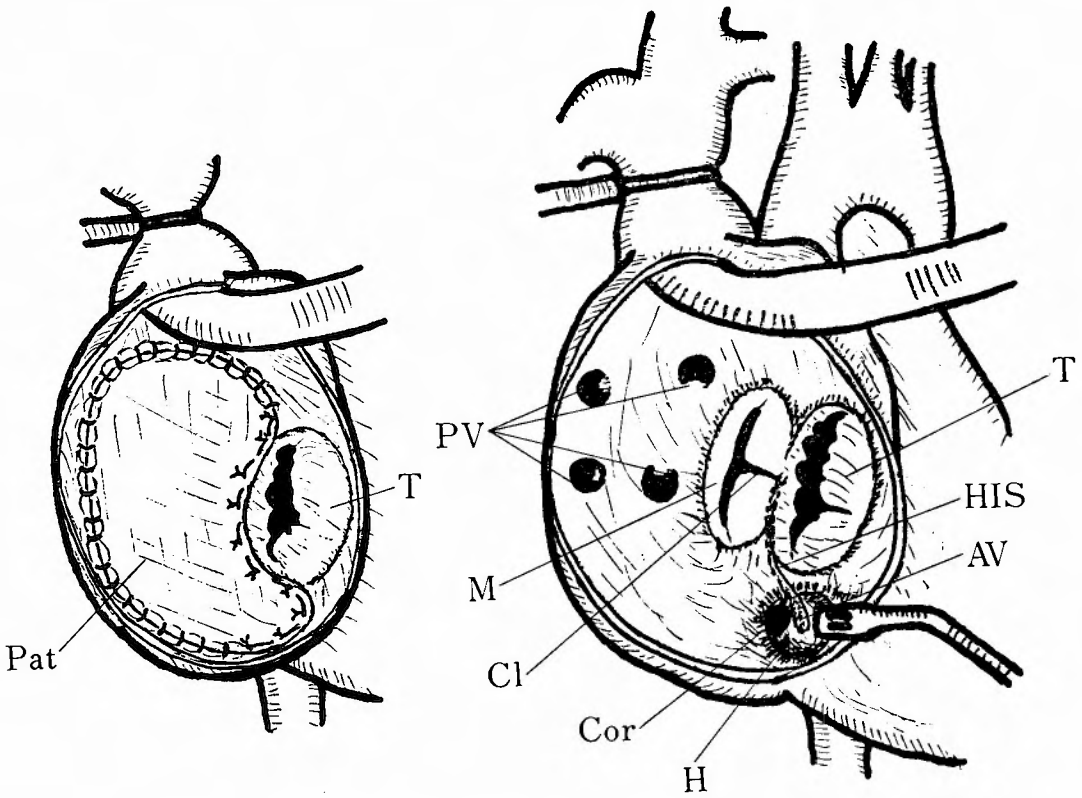

Fig 7 Operative Method and Finding
M : mitral valve, Cl : cleft, T : tricuspid valve PV : ostium of pulmonary vein, H : ostium of hepatic vein Cor : ostium of coronary vein, AV : atrioventricular node. HIS : bundle of His, Pat : A dacron patch by which the common atrial chamber was separated so that the hepatic vein and coronary vein drained into the left side of the heart avoiding surgical AV block.

cardiac catheterization and selective angiocardiography. No cyanosis was present. The chest X-ray showed that pulmonary congestion had disappeared and the cardiothoracic ratio had decreased to 0.60. Cardiac catheterization and angiocardiography showed slight tricuspid regurgitation and slight aortic regurgitation both of which might have been caused by the surgical procedure, but the pulmonary artery pressure was decreased ($P_p/P_s=0.35$). Arterial blood oxgen saturation was 88% despite drainage of the hepatic vein in to the left heart. The postoperative electrocardiogram showed atrioventricular junctional beats with right bundle branch block (Fig 8). He is now very active and leads a normal life.

Discussion

Asplenia is usually associated with severe and complex cardiac anomalies such

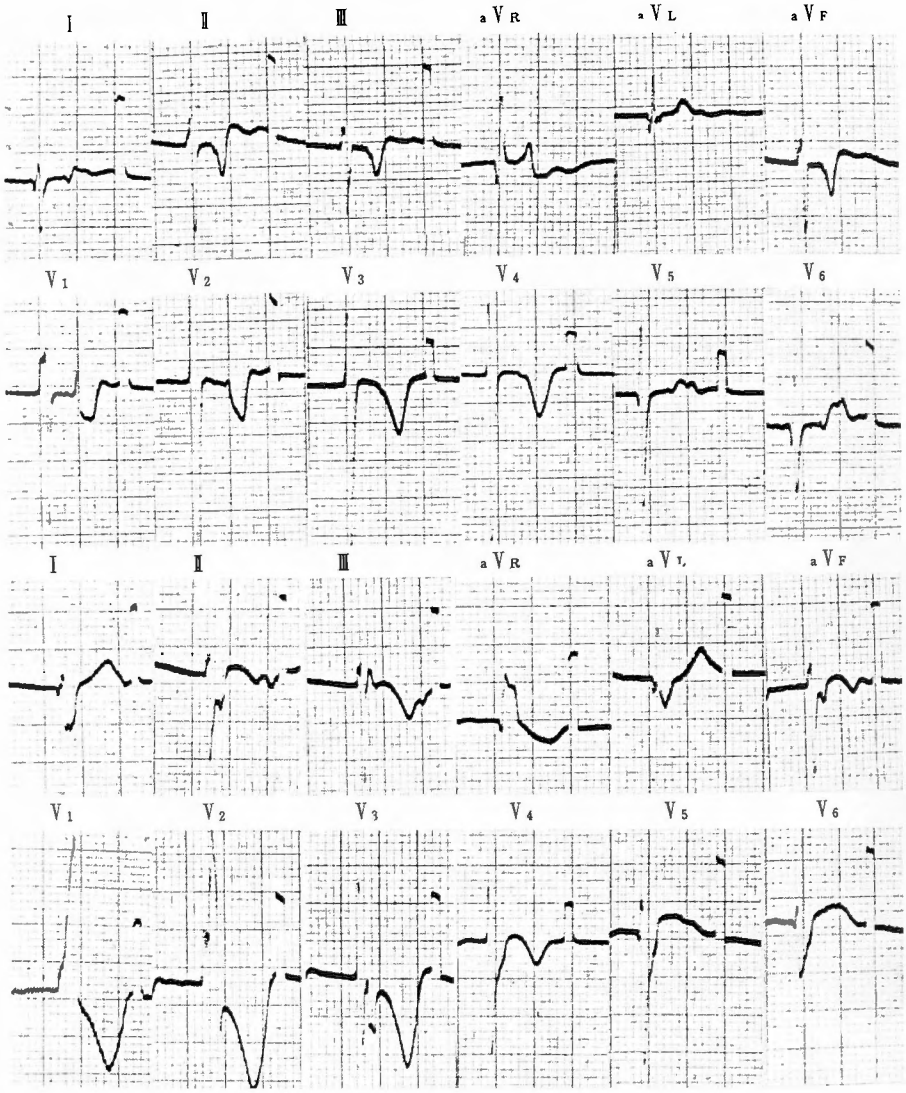

Fig 8 Pre- and Post-operative Electrocardiograms
A : Preoperative electrocardiograms show arrhythmia with AV junctional bearts. The electrical axis of ORS is -110 degrees.
B : Postoperative electrocardiograms show arrhythmia with A-V junctional beats and right bundle branch block. The surgical AV block could be avoided.

as D-transposition, L-transposition, double outlet right ventricle, pulmonary stenosis, single ventricle, common AV canal, total anomalous venous connection, etc[8,11,12,13]. There are few survivors beyond one year of age. Palliative surgical procedures are chosen in most cases.

On the other hand, polysplenia is associated with much milder cardiac anomalies, such as anomalous pulmonary venous connection, atrial septal defect, AV canal defect,

absence of the inferior vena cava, ventricular septal defect, etc[9,12]. Corrective surgery is usually possible.

Between these contrasting entities, there is a wide spectrum of intermediate forms. In this case, there was evidence of asplenia : HOWELL-JOLLY bodies[10] in the peripheral blood smear, negative spleen scintigram[3,5] and suspected bilateral trilobulated lungs[7]. Absence of the inferior vena cava, is unusual in asplenia, but rather common in polysplenia. Moreover, the intracardiac anomalies in this case were less severe than those in typical asplenia[2,3,14].

There is an entity consisting of isolated levocardia with viscero-atrial heterotaxia; in 94% of these cases splenic dysplasia is present[4,6]. It is reasonable to assume that this case represents an intermediate form from in the group of isolated levocardia with viscero-atrial heterotaxia, since there was clinical evidence of asplenia with less severe intracardiac anomalies than usual (Fig 9).

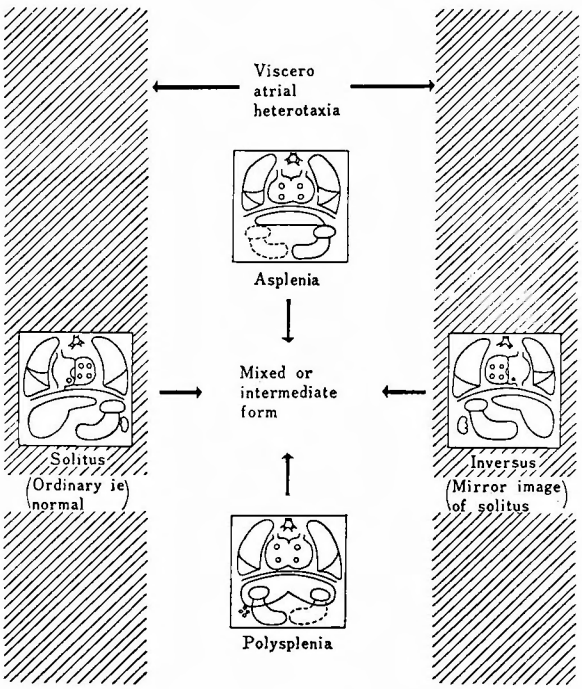

Fig 9 Situs of viscero and atria
Our present case may be classified as the intermediate form.

The most unusual finding in our case was abnormal position of the atrioventricular node. Many reports have noted the absence of the coronary sinus in asplenia[2,8]. As far as we know, no report has described the hepatic vein connection with the coronary vein and the unusual location of the atrioventricular node in asplenia.

We are now routinely performing intraoperative mapping to identify the course

of the bundle of His in corrected transposition of the great vessels, single ventricle and some cases of common atrioventricular canal defects. With this technique, we could find the exact course of the bundle of His and the position of the atrioventricular node in the present case and thus avoid surgical AV block by converting the hepatic venous flow into the left side of the heart. Practically, there is no adverse effect of this procedure at the present time. Intracardiac His bundle mapping is a useful tecnique in the treatment of complex cardiac malformations.

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和文抄録

無脾症を伴なった単心房の手術

——特に異常刺激伝導路の処置について——

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無脾症を伴なう先天性心疾患は非常に重症でかつ複雑であるため多くは1才未満に死亡し手術対象となることは少ない．しかも姑息的手術が殆んどであり解剖学的根治手術は極めて稀である．

われわれは最近3才の男児の無脾症を伴なう心奇型の開心術に成功した．本症例の心奇型は比較的軽症であって単心房，下大静脈欠損・奇静脈還流および房室結節性不整脈があった．

手術中に行なった電気生理学的ヒス束探索により房室結節は肝静脈開口部内に異常還流する冠静脈附近にあり，ヒス束は肝静脈右心房連結部を通り心室中隔に入ることが確認された．このため肝静脈および冠静脈の還流血を右心房に流入させるような心房中隔作製法では外科的房室ブロックの発生が必至と考えられた．そこでわれわれはこれらの静脈血を左心房に還流させる心房中隔作製法を行ない房室ブロックの発生を廻避することができた．この方法による不利益は現在の所認められていない．このような異常ヒス束走行と外科治療に関する報告はわれわれの知る限り皆無と思われるので詳細を報告する．