Arch Jpn Chir 50(5), 689～698, Sept., 1981

臨　　床

Surgical Treatment of Type B Complete Atrioventricular Canal

Kazuaki Minami, Norikazu Tatsuta, Yutaka Konishi, Katsuhiko Matsuda, Kazuo Hirata, Ario Yamasato, Yukio Chiba, Hiroshi Ishihara, Yoshisada Shiraishi, Tomohiko Muraguchi, Shinji Murata and Yorinori Hikasa

The 2nd Department of Surgery, Faculty of Medicine, Kyoto University
(Director: Prof. Dr. Yorinori Hikasa)

Tadashi Hayashidera and Tadashi Ueda

Department of Pediatrics, Faculty of Medicine, Kyoto University
(Director: Prof. Dr. Haruki Mikawa)
Received for Publication, July 1, 1981.

Introduction

The repair for complete atrioventricular (A–V) canal has improved after greater understanding of anatomic features and the operation mortality rate was reduced to a low level. Rastelli et al.[7] made a classification of complete A–V canal deformities into three subgroups (A, B and C) according to the configuration of the anterior leaflet of the common A–V valve. McMullan et al.[5] had achieved good results from their technique for repairing these deformities according to three types, especially type A and C. Type B of this complex anomalies is very rare, therefore there have been few reports describing in detail surgical procedures.

A 4-year-old boy with Down's syndrome was operated upon for common A–V canal and infundibular pulmonary stenosis. He survived the operation and was doing well 4 months postoperatively. The surgical procedure used to this case will be described herein.

Case report

A male baby was born after a uneventful full-term pregnancy. Delivery was normal and body weight at birth was 2790 gm. The patient had Down's syndrome. He was prone to respiratory infection and a cardiac murmur was detected at the age of 20 days. In July, 1976, he was admitted for 11 days in a hospital in Kyoto city during which time examinations revealed com-

Key words: Type B complete atrioventricular canal, abnormal papillary muscle, Cross-patch technique, Rastelli's classification, Ultrasonic cardiotomography.
索引語：完全型心内膜床欠損症B型，異常乳頭筋，十字型パッチ法，Rastelli 分類，超音波断層法.
Present address: The 2nd Department of Surgery, Faculty of Medicine, Kyoto University, Sakyo-ku, Kyoto, 606, Japan.

plete A-V canal. As congestive heart failure and cyanosis frequently occurred, Digosin was administered. He was subsequently referred to the Department of Pediatrics, Kyoto University Hospital, in August, 1976. In July, 1977, he suffered from high fever and general cramp, in August, 1977: measles, heart failure and dehydration, and in January 1979: Microplasma pneumonia and congestive failure.

Physical examination on admission revealed an acyanotic, well-developed, 4-year-old child, weighing 12 kg. The lungs were clear. A systolic thrill was present along the left sternal border, and the heart was mildly to moderately overactive, with a normal S1. S2 was fixedly split and P2 was slightly accentuated. A Grade 3-4/6 systolic murmur and a prominent diastolic rumble were present along the left sternal border and at the apex respectively. The liver was palpable 2 cm below the right costal margin. The spleen was not palpable. No palpebral or pedal edema was found.

Chest roentgenogram (CTR=0.65) and electrocardiogram were characteristic of endocardial cushion defect (Fig. 1, 2). Cardiac catheterization was performed (Table I). A left ventricular angiogram showed the typical goose neck deformity of the complete A-V canal, and mitral and tricuspid regurgitation were estimated to be severe (Fig. 3). Because of pulmonary hypertension, elevated pulmonary arterial resistance and infundibular pulmonary stenosis, corrective repair was deemed advisable.

On November 10, 1980, during standard cardiopulmonary bypass with cooling to 30°C, a type B of complete A-V canal was found. The repair was performed through a right atriotomy. Ideal exposure was gained by insertion of inferior cannulation as shown in Fig. 4.

The ventricular septal defect (VSD) extended beneath the common posterior leaflet, which was attached to the septum. The anterior common leaflet was divided but not attached to the crest of the ventricular septum. However the mitral and tricuspid portion of the anterior common

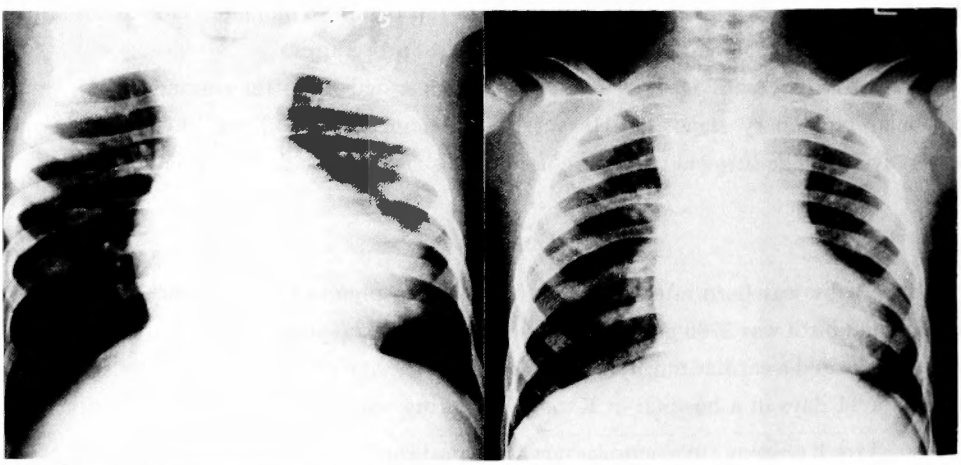

Fig. 1. Preoperative and postoperative chest roentgenograms
Preoperative chest roentgenogram (left) shows cardiac dilatation with a cardiothoracic rate of 0.65 and an increase in pulmonary vasculatures.
Postoperative chest roentgenogram (right) shows remarkable decrease in cardiac size

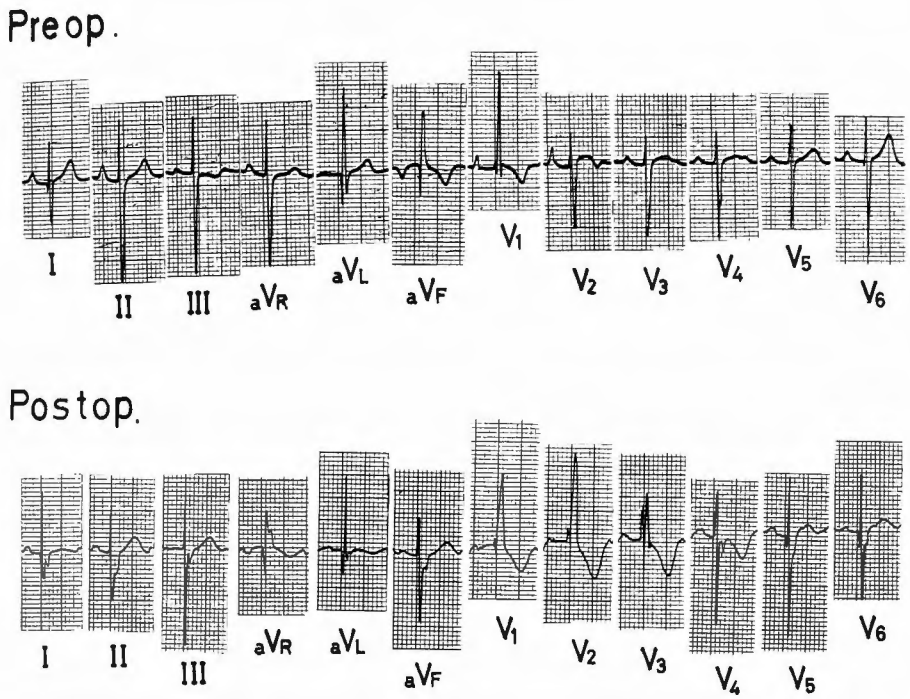

Fig. 2. Preoperative electrocardiogram shows incomplete right bundle branch block (R-BBB), with electrical axis of −90°
Postoperative electrocardiogram shows regular sinus rhythm with complete R-BBB.

leaflet were attached by chordae tendineae to an abnormal paillary muscle which originated from the right ventricular septum (Fig. 5). Very few cases of this type have been encountered and repaired[4,7]. There were two atrial septal defects (ASD); primum ostium approximately 32×22 mm and secundum ostium 10×10 mm. The crest of the ventricular septum was appro-

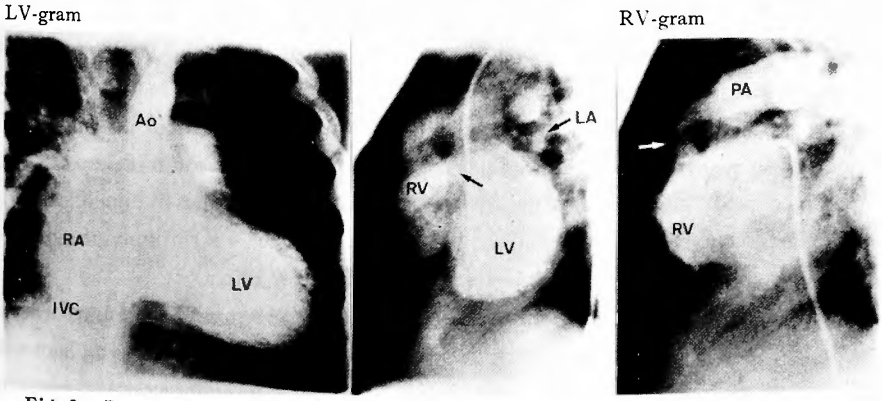

Fig. 3. Preoperative angiocardiography
A. Left ventriculography (A-P projection) shows severe mitral and tricuspid regurgitation. Left ventriculography (Lateral projection) shows a VSD and severe mitral regurgitation.
B. Right ventriculography shows infundibular pulmonary stenosis (arrow).

Table I. Preoperative cardiac catheterization data

	Pressure	O_2 (sat.)
1-PA	70/35 (45)	14.45
m-PA	70/40 (48)	13.97
RVinf	90/12	13.13
RAm	a 7 v 7 (4)	13.16
SVC	(4)	11.86
IVC	(4)	11.06
Ao(asc)	90/50 (64)	14.30 (79.2%)
LV	90/20	
LA	a 7 v 7 (4)	
PV		16.29 (90.2%)

CO 3.38 CI 5.83
PAR 612 Shunt L→R 3.35 (62.5%), R→L 1.37 (40.5%)

ximately 10 mm beneath the plane of the common A–V valves. After the anatomy of the anterior and posterior common leaflets was determined, the initial step in the repair was the division of the abnormal papillary muscle. With the ventricular wall remaining intact, the papillary muscle was divided into two groups, one group attached to the mitral valve and the other group to the tricuspid valve. The incision at the midline of the anterior and posterior common leaflet extended to the valvular ring. And then their mitral components were sutured together with nine interrupted sutures of 5–0 Prolene.

In order to close the VSD, 14 U-stay sutures were utilized to suture a patch to the right side of interventricular septum, thus avoiding the bundle of His. Therefore the suture-line was placed between the two divided halves of the papillary muscle which were adjacent to the septum. A cross patch made from two sheets of Dacron was designed so as to close the ASD and VSD and to compensate for any deficiencies of the valves. After the VSD was closed, the mitral rim of the patch was sutured to the reconstructed anterior mitral leaflet with mixing mattress suture and continuous running suture in some places using 5–0 Prolene. And then the anterior and posterior common leaflets were sutured together so as to make a tricuspid septal leaflet. The tricuspid rim of the patch was sutured to the septal tricuspid leaflet using the same method as on the mitral side. As the rim of the patch was 6 mm long, no deficiency of the tricuspid valve was found. Finally patch closure of the two ASD was performed, including secundum ostium ASD and the placing of the coronary sinus to the side of left atrium. Infundibular pulmonary stenosis was alleviated by the resection of the abnormal hypertrophic cardiac muscle through infundibulectomy. Right ventricular outflow tract was reconstructed with a Dacron patch.

After bypass, pressures (mmHg) were: right ventricle 59/13, main pulmonary artery 54/16, left ventricle 102/8, left atrium 24/12 and right atrium 19/17. After the operation, hemodynamics were readily maintained with minimal support by Dopamine. The endotacheal tube was removed on the night of the operative day. The patient did not regained full consciousness until the seventh postoperative day. Tachycardia with A–V dissociation on electrocardiogram lasted from the second postoperative day until the tenth postoperative day. Digitalis and diuretica were

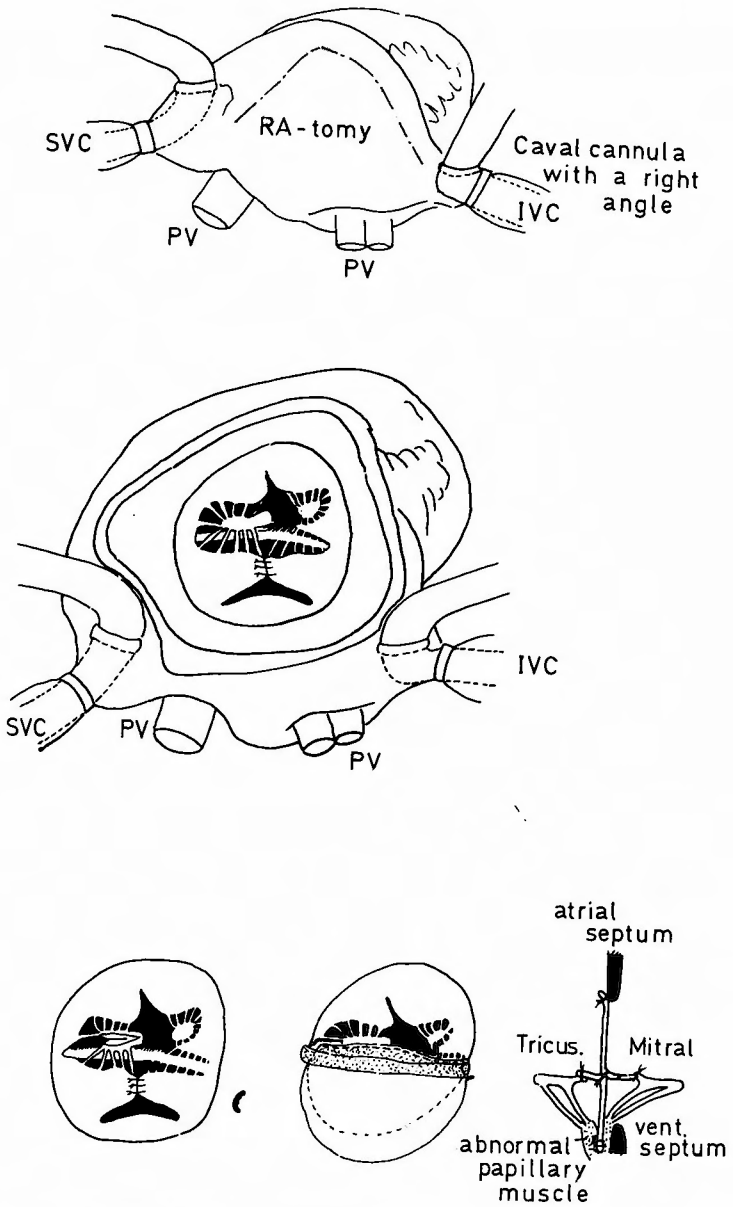

Fig. 4. Surgical procedures for type B complete A-V canal

administered and the rhythm gradually returned to normal. Until nine days after the operation, the patient had signs of fluid retention, as indicated by hepatomegaly and palpebral edema, with a liver palpable 4 to 5 cm below the right costal margin. Severe diuretic therapy was initiated, and a good response was gradually obtained.

Upon discharge 50 days after the operation, regular sinus rhythm was present with complete right bundle branch block (Fig. 2). Chest roentgenogram revealed a decrease in the size of

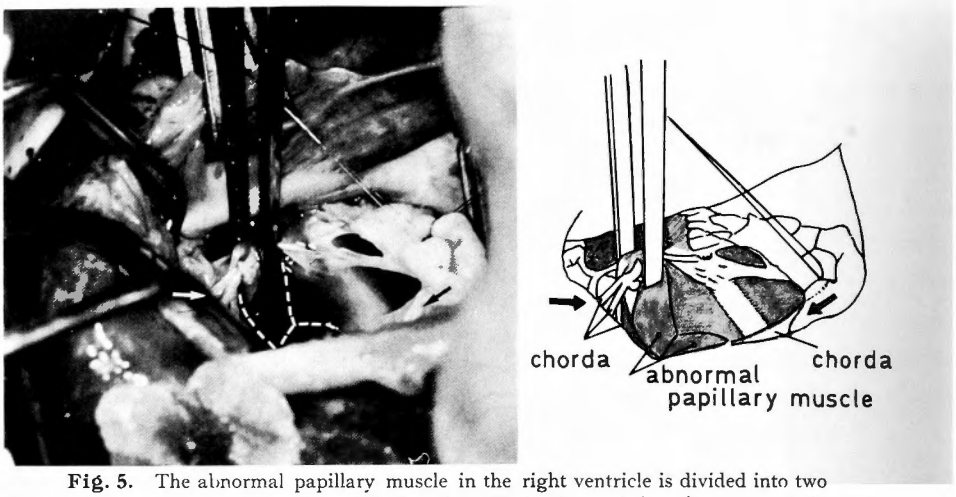

Fig. 5. The abnormal papillary muscle in the right ventricle is divided into two groups, one group attached to the mitral valve and the other group (held with tweezers) to the tricuspid valve.

Table II. Postoperative cardiac catheterization data

	Pressure	O_2 (sat.)
r-PC	14/5 (8)	
r-PA	46/16 (31)	9.76
m-PA	46/17 (30)	
RVout	45/0, 8	9.76
RVinf	48/0, 8	9.67
RAm	(13)	9.79
SVC	(13)	9.91
IVC	(13)	
LV	95/−5, 5	15.47 (90.9%)

cardiac shadow (Fig. 1). Physical examination revealed a Grade 2/6, blowing systolic murmur heard loudest along the left sternal border at the apex. The liver edge was 1 cm below the costal margin. Both lungs were clear. He was doing well and active at last follow-up, 4 months postoperatively.

On February 25, 1981, postoperative cardiac catheterization and angiocardiography were performed. The pressures (mmHg) were: right atrium (13), right ventricle 45/0, 8, main pulmonary artery 46/17 (30) and left ventricle 95/−5,5 (Table II). Mitral and tricuspid regurgitation were not found in agniograms of left or right ventricle (Fig. 6).

Discussion

RASTELLI et al.[7] made a classification of complete A–V canal into three groups (A, B and C) according to the configuration of the anterior leaflet of the common A–V valve. In type A, the anterior common A–V leaflet is divided into two portions, one mitral and tricuspid, both attached

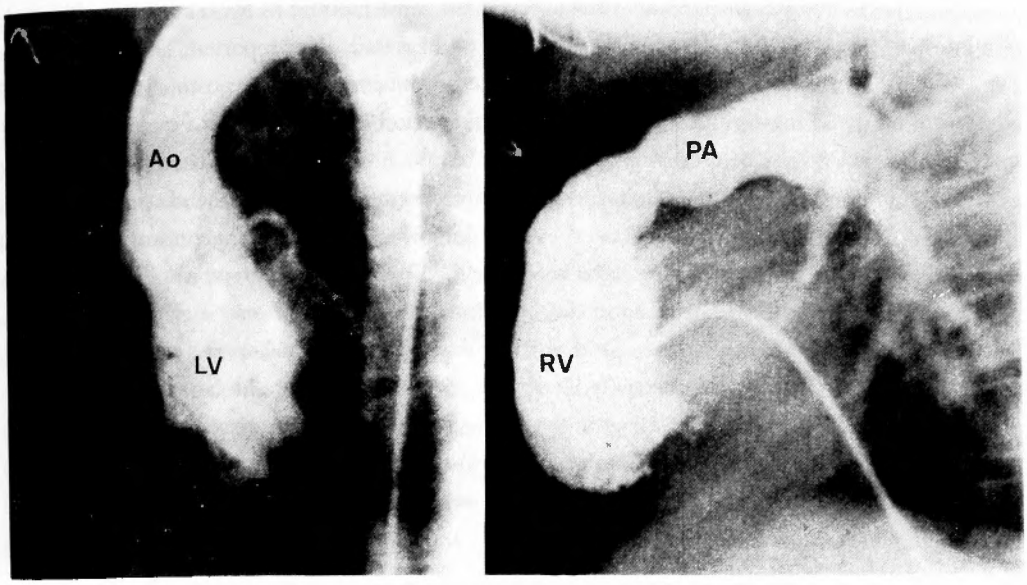

Fig. 6. Postoperative angiography shows minimum tricuspid and mitral regurgitation, alleviating infundibular pulmonary stenosis.

medially to the muscular septum. In type B, the anterior common leaflet is divided but not attached to the crest of the ventricular septum. The mitral and tricuspid portions of the anterior common leaflet are both attached by chordae to an abnormal papillary muscle that arises in the right ventricle near the septum. In type C, the anterior common A–V leaflet is undivided and unattched to the septum but freely floats above it. The posterior common leaflet in all three types is usually rudimentary and shows various anatomic arrangements similar to those described for anterior common leaflet. RASTELLI et al.[7] operated upon 38 patients; 25 of type A, 3 of type B and 10 of type C.

Type A is the most frequent and is usually found independent of other anomalies. Type B and type C have been frequently associated with other major cardiac anomalies, such as pulmonary stenosis, double outlet right ventricle and transposition of the great arteries etc. This present case was associated with infundibular pulmonary stenosis, and Down's syndrome. In addition to this description of the anatomy of the valve leaflets, there are frequently other abnormalities present. These can be seen as scanty valve tissue, shortened chordae tendineae, hypoplasia of the mitral valve tissue, abnormalities of the left ventricular and right ventricular papillary muscles, and double orifices in the mitral valve tissue. Although Rastelli's classification has been criticized on morphogenetic grounds[2], for the surgeon it is a convenient classification for describing surgical repair. In the repair of type B complete A–V canal as suggested by RASTELLI [7], good results were not achieved. This may be due to mitral and tricuspid regurgitation resulting from poor support by chordae tendineae. PACIFICO et al.[7] reported a successful case of type B complete A–V canal in which the chordae tendineae from the mitral portion of the anterior leaflet was divided to allow complete closure of the atrioventricular defect and the mitral portion

of the anterior leaflet was suspended to the patch in the same method as RASTELLI's[7]. Although the postoperative hemodynamics were not described in detail, they reported, however, a Grade 3/6 systolic murmur: this may be due to mitral regurgitation. As the postoperative degree of mitral and tricuspid regurgitation depends on whether chordal support can be maintained or not, the postoperative results of type A complete A–V canal have been better than those in type B and C. Therefore in order to maintain support by chordae tendineae, the abnormal papillary muscle should be divided in half. BEPPU et al.[1] demonstrated by using echocardiography that the movement of the mitral anterior leaflet was distrubed by the chordae tendineae postoperatively in some cases with endocardial cushion defect. But in this present case such a disturbance in the movement of the mitral and tricuspid valve was not found in postoperative echocardiogram (Fig. 7). In addition, the reconstructed tricuspid septal leaflet should be supplimented by a patch to compensate for any deficiencies in the valve areas. In the closure of the VSD ,the suture-line is always kept on the right ventricular side of the septum, running between the divided abnormal papillary muscle which is adjacent to the septum. In the repair of the complete A–V canal, the greatest problem lies in preventing mitral and tricuspid regurgitation.

There are various surgical procedures for this region. RASTELLI et al.[7] divided the anterior and posterior common leaflets in the right ventricle and sutured the divided leaflets into a single

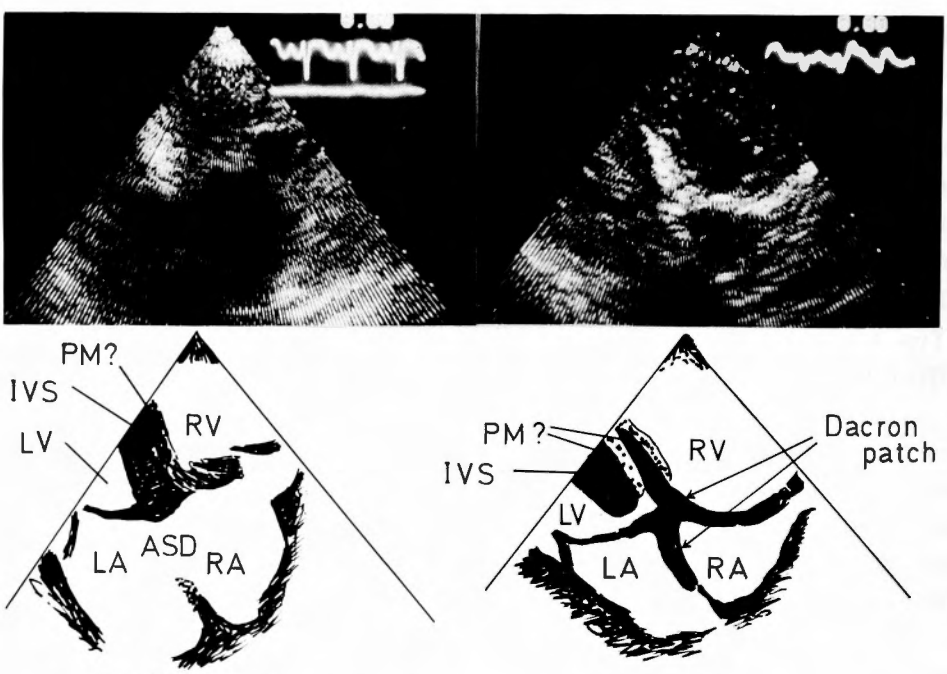

Fig. 7. Ultrasonic cardiotomography —Apical four-chamber view—
Preoperative UCG shows a primum ASD and slightly hypertrophied IVS.
Postoperative UCG shows four cardiac chambers separated by a Dacron Patch.
The primum ASD is closed.
RV: right ventricle, LV: left ventricle, RA: right atrium, LA: left atrium, ASD: atrial septal defect, PM: papillary muscle

patch. The disadvantage of this method is that the valve area is decreased by the suture, especially the tricuspid valve. A two-patch technique as suggested by SHIROTANI[8] and LINCOLN[4], has the advantage of avoiding the incision of the valve tissue and thus the decrease in valve area. This method is advisable when the valve tissue is extremely thin as in infants and small children. But when the advancement of mitral and tricuspid valve is needed in scanty valve leaflet, this method may be contraindicated. Therefore in complete A–V canal with severe scanty valve leaflet a cross-patch technique is advisable when a possibility exists that mitral and tricuspid regurgitation may occur postoperatively. There are various methods to make a cross-patch, such as KAWASHIMA's[3] method and ours. The cross-patch used in this present case was constructed from two "L" shaped sheets of Dacron as shown in Fig. 4. It is convenient to be able to change the size of this cross-patch which is readily made prior to the operation. The cross-patch technique may have greater application than the two-patch technique or the single patch technique. Good operative results in complete A–V canal depend on cold cardioplegic myocardial protection during the operation thus making the complicated procedures easier by permitting a clear operative field.

Complications of this complex surgery are:

1) Damage to the atrioventricular conduction system
2) Residual mitral and/or tricuspid regurgitation due to an inadequate repair of the common A–V leaflet
3) Residual VSD and ASD due to inadequacy of the adherence between the patch and rim of the atrioventricular canal defect
4) Transient anemia due to hemolysis

In this present case using cold cardioplegic myocardial protection, when placing the stitches in the area of the atrioventricular node during cardiac arrest, there are various possibility for making a complete A–V block. Thus the method in which the coronary sinus is placed on the side of the left atrium was adopted to increase safety. Residual shunt and regurgitation cause difficulties in postoperative management. Transient hemolysis in the postoperative period may be considerable due to residual shunt and regurgitation.

Summary

A 4-year-old boy with type B complete A–V canal, complicated pulmonary stenosis and Down's syndrome, was operated upon, using cold cardioplegic myocardial protection. The abnormal papillary muscle was divided into half, making a sutureline between the divided papillary muscle. The anterior and posterior common leaflets were incised in the midline and reconstructed with a cross-patch technique which was specially designed. The patient survived the operation. The postoperative cardiac catheterization revealed no residual shunt at atrial or ventricular levels and minimum regurgitation of either atrioventricular valves. The method in which the abnormal papillary muscle is divided and a cross-patch is used is advisable for type B complete A–V canal.

References

1) Beppu S, Nimura Y, et al: Minral cleft in ostium primum atrial septal defect assessed by cross-sectional echocardiography. Circulation **62**: 1099–1107, 1980.
2) Berger TJ, Kirklin JW, et al.: Primary repair of complete atrioventricular canal in patient less than 2 years old. Amer J Cardiol **41**: 906–913, 1978.
3) Kawashima Y, Tomokuni T, et al.. Endocardial cushion defect. Jap J Thorac Surg **28**: 773–780, 1975.
4) Lincoln C: Atrioventricular canal: Surgical anatomy and repair. Modern Cardiac Surgery edited by Longmore DB. Lancaster, MTP Press Limited 1978. p. 125–130
5) McMullan MH, Wallace RB, et al.. Surgical treatment of complete atrioventricular canal. Surgery **72**: 905–912, 1972.
6) Pacifico AD, Kirklin JW: Surgical repair of complete atrioventricular canal with anterior common leaflet attached to an anomalous right ventricular papillary muscle. J Thorac Cardiovasc Surg **65**: 727–730, 1973.
7) Rastelli GC, Ongley PA, et al.. Surgical repair of the complete form of persistent common atrioventricular canal. J Thorac Cardiovasc Surg **55**: 299–308, 1968.
8) Shirotani H: Total repair for complete atrioventricular canal. J Jap Assoc Thorac Surg **26**: 310–313, 1978.

和文抄録

B型完全型心内膜床欠損症の手術

京都大学医学部外科学教室第2講座（主任：日笠頼則教授）

南　一明，龍田　憲和，小西　裕，松田　捷彦，平田　和男
山里　有男，千葉　幸夫，石原　浩，白石　義定
村口　和彦，村田　真司，日笠　頼則

京都大学医学部小児科学教室（主任：三河春樹教授）

林寺　忠，上田　忠

4才男児，完全型心内膜床欠損症B型で，漏斗部狭窄およびダウン症候群を合併していた．手術は，体外循環，cardioplegia, topical cooling 併用下に cardiac arrest として，右房切開にて行なった．完全型心内膜床欠損症は，Rastelli 分類のB型で，右室にある異常乳頭筋が共通前尖の僧帽弁側および三尖弁側に腱索を送っていた．僧帽弁側の腱常索を切断すれば，中心性僧帽弁閉鎖不全を来たすと判断したので，異常乳頭筋を，腱索の方向から二分し，パッチによる閉鎖線は，切開した乳頭筋間を通した．パッチは，2枚の"L"型ダクロンを中央で縫合し十字型パッチとした．僧帽弁および三尖弁の弁腹を補うための庇は6mmとした．冠静脈洞は左房側へ行くようパッチをあてた．術後経過はおおむね良好で，術後心カテ検査で，遺残短絡はなくRV/LV 圧比は0.5と減少，PS は全く消失．僧帽弁・三尖弁での逆流もごく軽度であった．術後7ヶ月の現在，元気である．