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ive Surgery for Complicated Cardiac Anomaly and its Problems

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The results of total correction for complicated cardiac anomalies have been improving but there are several problems in each type of surgery of them. It is very difficult to represent in details on the various aspects on the surgery for all of complecated cardiac anomalies. Therefore, in this paper, the operative results and the main problems in total correction of major complicated cardiac anomalies are reported, based on our recent achievement.

1. Tetralogy of Fallot

Our latest operative result is shown in Fig. 1. One hundred sixteen patients with tetralogy of Fallot underwent total correction during the period of 1978 to 1982 in our hospital. The age distribution was from one year to 51 years old (the average was 9.6 years old). Only one operative death occurred, the mortality rate was accounted as 0.9%. This patient had hypoplastic left ventricle as only 9 ml of left ventricular end-diastolic volume (LVEDV), which was 21 ml/m^2 as end-diastolic volume index (LVEDVI). He died of severe left heart failure postoperatively. Based on this experience, the study on post-operative hemodynamics and left ventricular volume of this anomaly was performed and it revealed that the critical size of LVEDVI which permits safe application of total correction is over 30 ml/m^2.[1] After this criteria was established, there was no operative death in latest 105 patients.

Our operative procedure is as follows: Total correction is performed using cardiopulmonary bypass and cardioplegia. Infundibular portion of right ventricle is vertically incised just beneath the pulmonary valve ring as shown in Fig. 2. The length of incision is limited not exceeding 40% of the distance from pulmonary ring to apex of right ventricle in order to prevent right ventricular dysfunction. Ventricular septal defect is closed with Teflon patch using interrupted sutures. At closing ventricular septal defect, we have made effort to prevent complete right

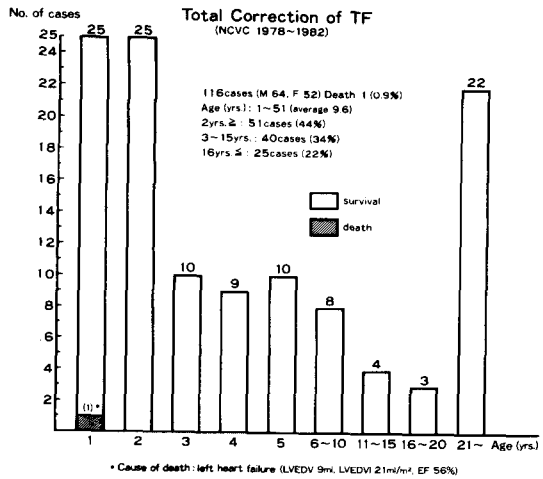

Fig. 1. Age distribution and operative results in patients who underwent total correction of tetralogy of Fallot between 1978 and 1982 at National Cardiovascular Center, Japan. M; male, F; female, yrs; years.

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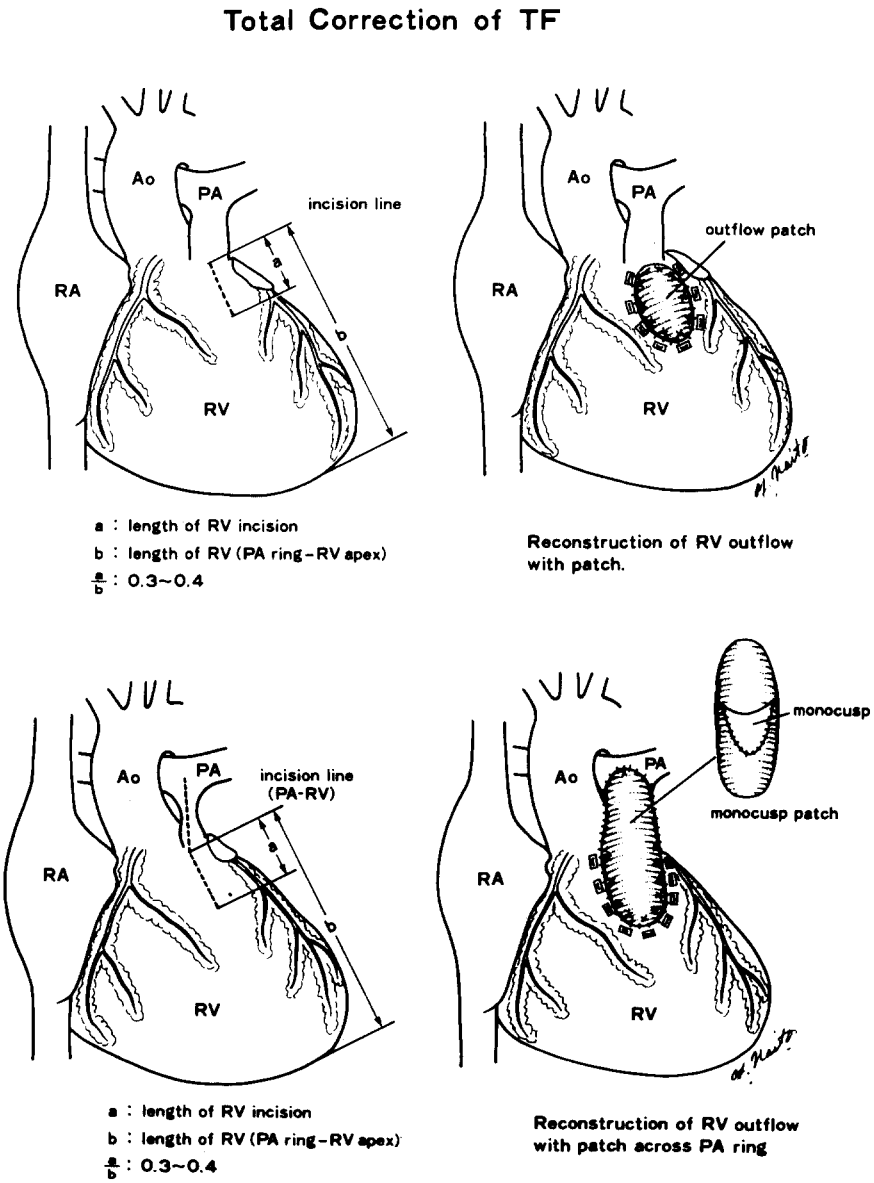

Fig. 2. The incision of right ventricle and reconstruction of the right ventricular outflow tract. Ao; aorta, PA; pulmonary artery, RA; right atrium RV; right ventricle.

bundle branch block (CRBBB) since 1979, because CRBBB may reduce right ventricular function and may be one cause for postoperative ventricular dysrhythmia which may lead to sudden death postoperatively. Our procedure to prevent CRBBB is shown in Fig. 3. We histologically confirmed that right bundle run along "fusion line" named by our colleague, Isobe[2], which lyes the groove between sinus septum and trabecula septomarginalis. In order to prevent CRBBB, this fusion line has to be kept away from suture, giving thread interval of 3 to 4mm. By this technique, incidence of postoperative CRBBB decreased and as incidence occurred only 32% in last year (Fig. 4).

Reconstruction of right ventricular outflow

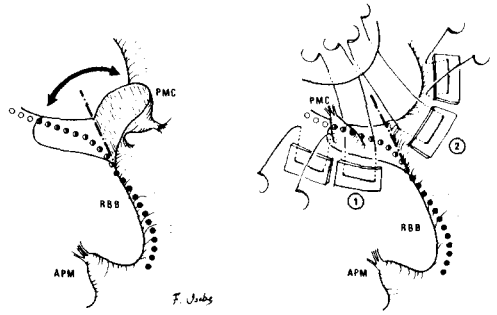

Fig. 3. Mode of travelling of the central right bundle branch (RBB) and method to prevent its injury in tetralogy of Fallot (malalignment type VSD).

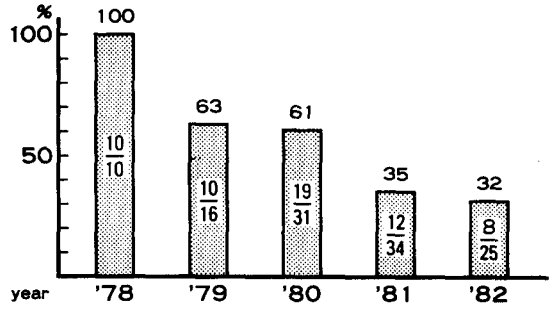

Fig. 4. Incidence of postoperative complete right bundle branch block (CRBBB).

tract is performed in accordance to my criteria[3-5] on each body surface area as shown in Fig. 5. After vertical incision on right ventricular infundibulum, the parietal band is sufficiently incised but as a rule, myocardium on septal band, parietal band and free wall of right ventricle is not removed in order to prevent reduction of right ventricular function postoperatively. Right ventricular infundibulum is enlarged by small patch at the incised portion (Fig. 2). When the patch enlargement has to be undertaken beyond the pulmonary ring up to the pulmonary trunk, the patch with pericardial monocusp is applied in order to prevent severe pulmonary valve insufficiency (Fig. 2), because it is one of the main factors influencing poor operative results[3-5].

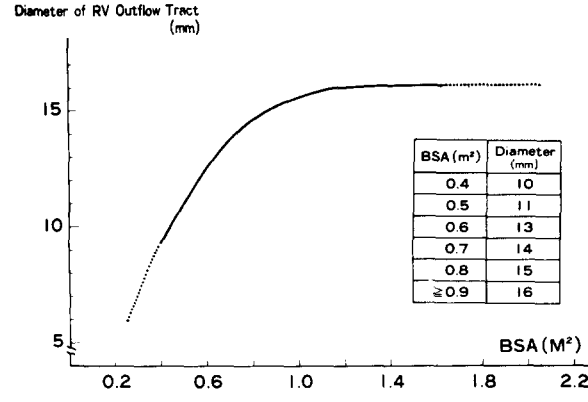

Fig. 5. Criteria for reconstruction of the right ventricular outflow tract in tetralogy of Fallot. The solid line shows the minimal diameter for the right ventricular outflow in each body surface area (BSA) to be enlarged in total correction of this anomaly. The table shows the value of this diameter in each body surface area derived from this curve.

2. Double outlet right ventricle

Twenty four patients with this anomaly underwent total correction and 8 deaths occurred, the operative mortality rate of 33% was accounted (Table 1). The most difficult part in surgery for this disease is Taussig-Bing anomaly. The operative procedures for Taussing-Bing anomaly

Table 1. Operative results of total correction of double outlet right ventricle.

	location of VSD	PS (−)	PS (+)
(S,D,D,)	subaortic	1 (0)	10 (5)
21 (8)	subpulmonary	7 (2)	0
38%	complete A-V canal	2 (1)	0
	noncommitted	0	1 (0)
(S,L,L,)	subaortic	0	2 (0)
3 (0)	subpulmonic	1 (0)	0
		11 (3)	13 (5)
		27%	38%

( ) : op. death (1983. 9. NCVC)
% : op. mortality

and their operative results are shown in Table 2. The Hightower procedure[6], which combines Mustard procedure[7] for transposition of the great arteries and closure of ventricular septal defect, has a problem on postoperative dysfunction of right ventricle as a systemic ventricle. The operative procedure which uses left ventricle as systemic ventricle is preferable. However, McGoon procedure[8] or Kawashima procedure[9] which creates intra-ventricular rerouting is technically quite difficult, if you want to minimize residual stenosis of both right and left ventricular outflow tract. As a result, recently, we prefer to choose Imai procedure[10] resembles to Rastelli procedure[11] or Jatene procedure[12] for the transposition of the great arteries.

Table 2. The operative procedure and results in Taussig-Bing anomaly.

Case	Age	Sex	Procedure	result
1. S.M.	2 yr.	M	Mc Goon	survived
2. K.Y.	1 yr.	F	Mc Goon	died
3. T.F.	2 yr.	M	Imai	survived
4. A.T.	1 tr.	M	Imai	survived
5. S.Y.	4 mo.	M	Hightower	survived
6. R.M.	7 mo.	F	Hightower	died
7. K.U.	7 mo.	M	Jatene	survived

(1983. 9. NCVC)

3. Transposition of the great arteries

Operative procedure in each type of this disease and their operative results are showed in Table 3. For the group I, which is intact ventricular septum group, the result of Mustard procedure[7] is good. In group 2, However, which has ventricular septal defect and pulmonary hypertension, their operative results are not satisfactory as you see 50% of mortality rate. Recently, right ventricular dysfunction following Mustard procedure[7] has became a topics. Therefore, operative method which uses left ventricle as a systemic ventricle such as Jatene[12] or Rastelli procedure[11] has been mainly applying at our service. The operative results on Rastelli procedure is good, we have not encountered any death, but on the other hand, on Jatene procedure the results is not satisfactory, the mortality rate of 60% was accounted.

As one of the postoperative complications in Mustard procedure, high incidence of dysrhythmia has been reported. To prevent this complication, we have modified Mustard procedure and have performed this procedure as shown in Fig. 6.[2,13] It is our concept that inter nodal conduction is maintained if the continuity of undamaged atrial muscle between S-A node and A-V node could be remained completely intact from incision and suture. In Mustard procedure, this limited path-

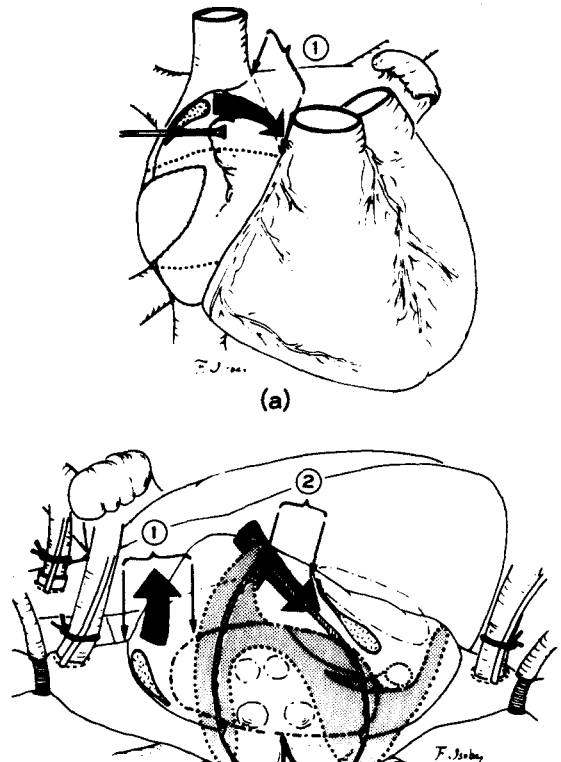

Fig. 6. Our modified Mustard procedure to prevent postoperative dysrhythmia.
– – – – – ; baffle suture line.
Arrow shows "noble zone".

Table 3. Operative procedure and results in complete transposition of the great arteries.

Type	Procedure	Age (Average)	No. of cases	No. of op death	motality (%)
I	Mustard	22D-14M (4.1M)	15	1	6.7
	Jatene	3M-11M (4.8M)	5	3	60.0
II	Mustard	1M-6M (3.6M)	8	4	50.0
	Rastelli	15M-18M(16M)	3	0	0
	Jatene	1M-31M (8.6M)	5	3	60.0
	Palli. Mustard	9Y-10Y	2	1	50.0
III	Rastelli	5Y-6Y (5.3Y)	3	0	0
IV	Mustard	2M	1	0	0
	Total		42	12	28,8

(1983. 9. NCVC)

Table 4. The incidence of postoperative arrhythmia following our Mustard procedure.

	preop.	early after op.	late after op.
SR	19	19 (2 0p. deaths)	16 (94.1 %)
NR	0	0	1*(5.9 %)

*sick sinus syndrome (1983. 9. NCVC)

way is confined to only the area which is pointed by arrow in Fig. 6. This pathway originates from S-A node to superior portion of the right atrium, enters to left atrium via atrial septum and runs toward A-V node through the narrow are a which lies between ventricular site of the superior limbic septum and suture line of the baffle. We call this limited area as "noble none". We are performing Mustard procedure to completely protect S-A node, its blood supply, A-V node and this noble zone. Table 4 shows the incidence of postoperative arrhythmia by our operative technique. In follow-up period, 94% of patients have maintained sinus rhythm.

4. Total anomalous pulmonary venous return

This anomaly could be divided into three types according to pathophysiological mainfestations.

Group A represents are patients with s.vere pulmonary venous obstruction (PVO), severe

Table 5. Operative results in total anomalous pulmonary venous return.

Physiological Group	Anatomical Group	Age <1M	<3M	<6M	<12M	12M≤	Total	
A	I	4(3)	2(2)					6(5)
	II	1(1)					1(1)	12(11)
								92%
	III	5(5)					5(5)	
B	I	2(0)	6(3)	1(0)	2(1)		11(4)	
	II	2(0)	3(0)		1(0)		6(0)	19(4)
	IV			1(0)	1(0)		2(0)	21%
C	I					1(0)	1(0)	
	II							3(0)
	III					2(0)	2(0)	0%
		14(9)	11(5)	2(0)	4(1)	3(0)		34(15)
		64%	45%	0%	25%	0%		44%

( ): op death, % : op. mortality (1983. 9. NCVC)

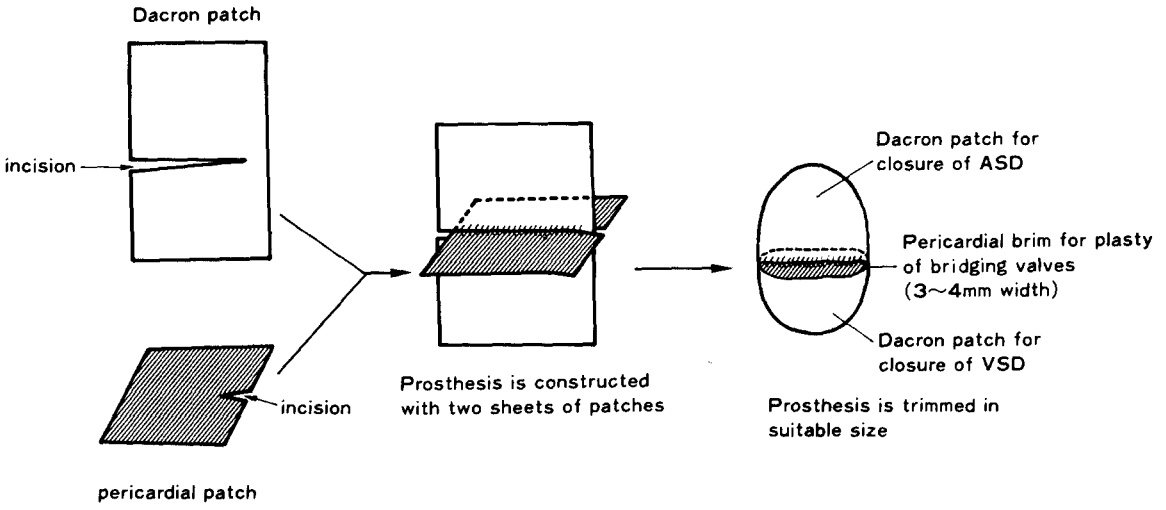

Fig. 7. Construction of prosthesis for intracardiac repair of complete atrioventricular canal.

pulmonary hypertension (PH) and mildly increased or decreased pulmonary blood flow (PF). Group B has moderate or mild PVO, severe PH and markedly increased PF. Group C represents absence of PVO, mild or absence of PH and markedly increased PF. The operative results by the combination of anatomical and physiological classification is shown in Table 5. The operative result was poor as operative mortality rate of 92% in Group A. But in Group B and C, their results were satisfactory. The surgical result according to the age showed that the mortality rate was 64% in newborn age group and 45% in less than 3 months of age and 11% in over the age of three months. In group A and group B, their clinical symptoms were grave and early attempt of surgery as soon as the diagnosis is is established is preferable.

5. Complete atrioventricular canal

Considering the natural history and the development of obstructive pulmonary vascular disease of this anomaly, surgical treatment for this disease should be performed under 2 years of age. However, operative results of intracardiac repair in such an infancy have been generally unsatisfactory. We have performed intracardiac repair using our own designed prosthesis[14]. This prosthesis consists of single Dacron patch with pericardial brim of 3 to 4 mm width on each side for plasty of atrioventricular valves (Fig. 7). Our operative procedure is shown in Fig. 8. By transatrial approac, anterior and posterior bridging leaflet are incised into two components of mitral and tricuspid valves. All of accessory chordae attached to the crest of ventricular septum are removed. Afterthen, ventricular septal defect, atrioventricular valves and atrial septal defect are repaired using our prosthesis.

The operative results in each type of Rastelli classification[15] of this anomaly are shown in Table 6. Applying this surgical procedure, 12 infants underwent intracardiac repair and 2 operative death occurred. This operative mortality rate of 16.7% could be acceptable as for operative results in infancy. In other five patients of more than 2 years old, there was no operative death.

6. Single ventricle

As a final category, the problem on intraventricular septation for single ventricle is presented. Total of 8 patients were operated (Table 7).

Table 6. Operative results of intracardial repair for complete atrioventricular canal in infancy.

type	Pts. No.	Op. death
A	9	2
B	1	0
C	2	0

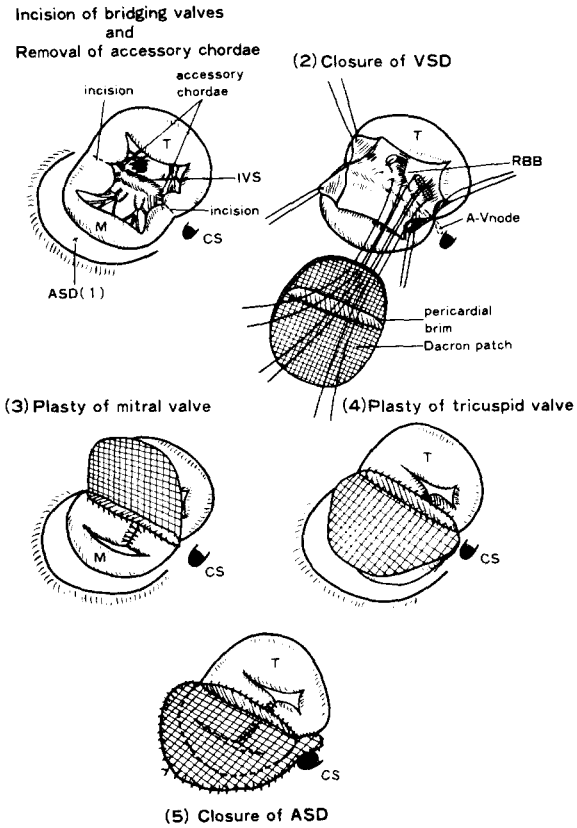

Fig. 8. Operative procedure for complete atrioventricular canal.

Five of them survived. Four of 5 patient without pulmonary stenosis were performed septation with success. On the attempt of septation, the prevention of A-V block is quite important. In Fig. 9, the pathway of conduction system in single ventricle with left anterior rudimentary chamber without pulmonary stenosis is demonstrated. The conventional suture line of septation is shown in the left side picture of Fig. 9. In this procedure, the suture line crosses non-branching bundle and results in high incidence of A-V block. We performed this operative technique on four patient and obtained A-V block on all of them. After this discouraging result, we have modified our procedure as shown in the right side picture of Fig. 9. The suture line is parallel to non-bran-

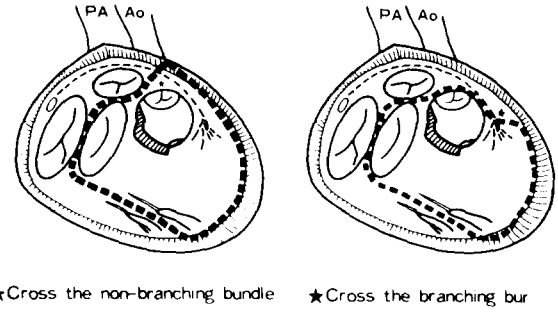

Fig. 9. Operative procedure of interventricular septation for single ventrice.
The left picture shows conventional suture line of septation and the right is our new suture line.
The fine dotted line represents non-branching bundle. The bald dotted line shows suture line of septation.

Table 7. Operative results of total correction for single ventricle.

Case	Age (yrs) Sex	Associated Anomaly	Procedure	Approach	Post-op A-V Block	Result
1. N.K.	4 M	Septation	RV	+	+	survived
2. H.N.	8 M	1-TGA, PH	Septation	RV	+	died
3. K.g.	1 M	1-TGA, PH	Septation	RA	+	survived
4. Y.H.	4 F	1-TGA, PH	Septation	RA	+	survived
5. S.I.	4 F	1-TGA, PDA, PH	Septation PDA ligation	RA	–	survived
6. E.M.	4 M	1-TGA, PS	Septation Enalrgement of PS	RA	+	died
7. S.M.	5 F	1-TGA, PS	Septation RV-PA conduit	RV	+	died
8. K.N.	3 M	CAVC, PA atresia	Fontan procedure		–	survived

ching bundle and is directed toward free wall after passing through its bundle. Latest two patients who were applied this modified technique have maintained without A-V block.

Conclusion

The results and problems on corrective surgery for main complicated cardiac anomalies were reported based on our latest experiences. The operative results in this field have recently much improved. It is our pleasure if this presentation will contribute to your knowledge concerning this particular type of surgery on congenital heart disease.

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