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Surgical Treatment of Ebstein's Anomaly

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Introduction

Ebstein's anomaly[7)28)] is a relatively rare congenital heart disease. The indication for surgical treatment, however, is still controvertial. While the diagnosis of the disease has been established today[9)24)37)], natural and surgical prognosis of each patient can not be decided on the definite basis irrespective of much concerning report[2)3)5)12)13)15)17)18)19)25)27)32)33)]. One of its main reasons is probably the small number of experiences in each clinic.

In this report, we present our experience of surgically treated 7 patients. This report is also of a small number of patients, however, we bleieve that it will add some new informations in this complicated field.

Materials and methods

Between 1958 and 1978, seven patients of Ebstein's anomaly were surgically treated at the Kyoto University Hospital and Tenri Hospital by the same surgeon (NT). All patients except for one were diagnosed preoperatively by cardiac catheterization and selective angiocardiography (Table 1). As associated lesions, all cases had atrial septal defect, 1 had pulmonary stenosis, and 1 had ventricular septal defect. As the tricuspid valve lesion, definite regurgitation was confirmed in 5 and stenosis was a dominate lesion in 2. Three patients had evidence of Wolff-Parkinson-White (WPW) syndrome[35)] All cases were surgically treated by corrective procedures including 4 valve replacements (TVR) and 3 valvuloplasties (TVR). Follow up study from 2 to 10 years was done. Functional disability was assessed according to the New York Heart Association Classification. Visible cyanosis was classified as severe, moderate, mild and none at rest.

Key words Ebstein's anomaly, tricuspid valve replacement, tricuspid valvuloplasty, atrialized right ventricle, WPW syndrome.
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Clinical and laboratory profile

Of these 7 patients, 4 were female and 3 were male. Age ranged from 24 days to 24 years at operation with a mean of 12 years. Functional classifications were two in class IV, 3 in III, 1 in II, and 1 was in class I, without considering frequent tachycardial attacks (Table 1).

One patient aged 24 days (case 5) showed severe congestive heart failure (functional class IV) and emergency operation was performed without cardiac catheterization under clinical diagnosis of severe pulmonary stenosis. One patient aged 19 years (case 6) with atrial fibrillation showed progressive deterioration, completely bedridden with chronic liver dysfunction (functional class IV). Three patients had histories of congestive heart failure (class III). All patients showed visible cyanosis at rest. Cyanosis was especially severe in a 5-year-old boy (case 3) with pulmonary stenosis and in a 19-year-old boy (case 6) with severe congestive heart failure and atrial fibrillation. All patients except for the 14-year-old girl (case 3) with tricuspid stenosis showed cardiomegaly. Cardiothoracic ratio of these 6 patients ranged from 59% to 76% (mean 68%). ECG examination revealed complete right bundle branch block in 4, WPW syndrome in 3, atrial fibrillation in 1, and histories of tachycardial attacks in 4 patients (Table 1).

Surgiral methods and anatomical findings

Heart-lung bypass with moderate hypothermia was employed in 6 patients, and surface cooling hypothermia (lowest rectal temperature 25°C) was employed in 1 infant (case 5, body weight 2,800 gm.). Tricuspid valve replacement (TVR) was done in our earlier 4 cases. The tricuspid valve was excised from the false valve ring and a caged disc valve (Kay-Shiley valve or Starr-Edwards valve) was sewn onto the true valve annulus except in the region of the coronary sinus ostium after Barnard's technique[3]. Elimination of the atrialized ventricle was never done. Valvuloplasty was done in our later 3 cases. True and false annuli were sewn together using separate U-stay sutures after Hardy's technique[12][13] in 2 patients. In one patient (case 5) preoperative clinical diagnosis was decompensated severe pulmonary stenosis. Under surface cooling hypothermia, the tricuspid valve was excised about 3/4 around at its insertion to the ventricular wall and was resutured to the true annulus. This infant had atrial septal defect, ventricular septal defect and tricuspid valvular stenosis as well, and all these lesions were corrected simultaneously. Atrial septal defects were completely closed in all patients. In one patient (case 4) the associated pulmonary valvular stenosis was relieved from the pulmonary artery. Displacement and deformity of the tricuspid valve were severe in all patients. In 5 patients dowmward displacement of the septal and the posterior portion of a large sail-like anteroposterior leaflet was observed. In 2 patients almost all valve ring except for a small portion around the central fibrous body was displaced. Various degrees of dystrophy of the valve were observed in all patients. A large sail-like anteroposterior leaflet was connected to the

Table 1

Case no.	Sex	Age	NYHA class preop.	NYHA class postop.	Cyanosis	Hb (g/dl)	CTR (%)	Histories of HF	Histories of PAT	ECG WPW	ECG RBBB	ECG PQ (sec)	ECG AF	Cardiac pressure RA a	Cardiac pressure RA m	Cardiac pressure RV peak
1	f	24y	III	I	moderate	16.1	59	+	+	B		0.16	−		6	15
2	m	20y	III	I	moderate	16.9	72	+	−	−	+	0.21	−		3	16
3	f	14y	II	I	mild	13.4	46	+	+	B		0.10	−	14	8	32
4	m	5y	III	I	sereve	20.1	76	−	−	−	+	0.16	−		7	92
5	f	24d	IV		mild	11.4	65	++	−	−	+	0.14	−			
6	m	19y	IV	II	sereve	19.7	70	++	−	−	+		+		14	40
7	f	7y	I	I	mild	13.5	64	−	+++	B		0.11	−		3	32

Legend : NYHA ; New York Heart Association
Hb ; hemoglobin
CTR ; cardio-thracic ratio
HF ; heart failure
PAT ; paroxysmal atrial tachycardia
ECG ; electrocardiogram
WPW ; Wolf-Parkinson-White syndrome
RBBB ; right bundle branch block
PQ ; PQ interval
AF ; atrial fibrillation
RV ; right ventricle
CI ; cardiac index

ventricular wall directly by short choradae in 1 patient. The anterior papillary muscle was located in the right ventricular outflow tract (conus portion) in 2 patients. Fenestrations were observed in the anteroposterior leaflet in 3 patients, and partial absence of the septal leaflet was observed in 1 patient. A seven-year-old girl (case 7) who had electrocardiographic findings of WPW syndrome had suffered from frequent tachycardial attacks. She was treated by division of the bundle of Kent as well as by valvuloplasty. Epicardial surface mapping was done and abnormal pre-exciting region was found at the inferior margin of the right ventricle. At this point, about 4 cm long incision of the atrial wall just parallel to the true tricuspid annulus was placed according to Sealy's method[30].

catheterization (mmHg)		Surgical findings			Surgical treatment	Results
CI	SAO_2	TR	TS	other anomalies		
l/min/m	%					
2.3	87	++	−	ASD	ASD closure TVR with K-S disc valve (Barnard's technic)	10 years after surgery uneventful 2 natural labors (3 years and 6 years after surgery) PAT(+), WPW(+), CTR 55% no physical limitation
2.0	90	++	−	ASD	ASD closure TVR with S-E disc valve (Barnard's technic)	9 years after surgery SVA(+), RBBB(+), CTR 62% no physical limitation
5.1	84	+	++	ASD	ASD closure TVR with K-S disc valve (Barnard's technic)	9 years after surgery PAT(+), WPW(+), CTR 45% no physical limitation
5.6	71	++	−	ASD PS	ASD closure Pulmonary valvotomy TVR with S-E disc valve (Barnard's technic)	late death 1 1/2 years after surgery (sepsis and malfunction of prosthetic valve)
		+	++	ASD VSD	ASD closure VSD closure TVP (partial excision and transposition of tricuspid valve)	operative death
1.4	80	++	−	ASD	ASD closure TVP (Hardy's technic)	late death 1 1/2 years after surgery (sudden death) TR(+)
4.5	94	+	−	ASD	ASD closure TVP (Hardy's technic) Division of His bundle	2 years after surgery PAT(−), WPW(−), RBBB(+) CTR : 58%, PQ : 0.18 sec. no physical limitation

RA ; right atrium
SAO_2 ; saturation of arterial blood oxgen
TR ; tricuspid regurgitation
TS ; tricuspid stenosis
PS ; pulmonary stenosis
ASD ; atrial septal defect
K-S ; Key-Shiley
S-E ; Starr-Edwards
TVR ; tricuspid valve replacement
TVP ; tricuspid valve plasty
SVA ; supraventricular arhythmia

Details of this case is described in another report by co-authors[38].

Results

There were one operative death and and 2 late deaths, with four patients surviving from 2 to 10 years after operation (Fig. 1). An infant (case 5) who was in severe congestive cardiac failure was operated upon at the age of 24 days without correct clinical diagnosis and died at operation from low cardiac output. One patient (case 4) who was treated by TVR showed decreasing cavity of the right atrium and the atrialized right ventricle postoperatively evidenced by angiocardiography (Fig. 2, 3), however, he died 1½

year after operation from severe infection. Clinical signs of sepsis, tachycardia and severe right heart failure were evident when he was readmitted. On fluoloscopy the valve poppet movement was markedly inhibited though he was on a continuous anticoagulant regimen. He died the next day of admission. Autopsy was denied. The direct cause of death was presumably thrombosed valve. A nineteen-year-old male died 1½ year after operation in sleep. He tolerated valvuloplastic procedure well though his preoperative situation was desperately ill. Liver enlargement and cyanosis dramatically disappeared, atrial fibrillation returned to normal sinus rhythm and physical activity increased. However, liver enlargement reappeared gradually and atrial fibrillation returned. Postoperative cardiac catheterization done at 1 month after operation revealed high atrial pressure. Evidence of moderate tricuspid regurgita-

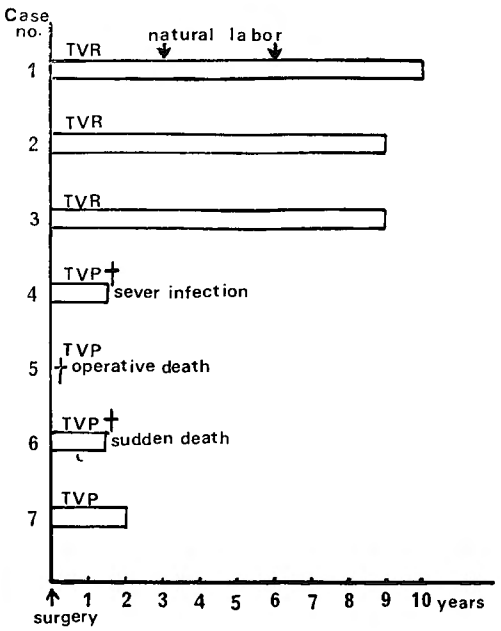

Fig. 1. follow up study of 7 cases of surgically treated Ebstein's anomaly.
TVR : tricuspid valve replacement
TVR : tricupid valvuloplasty
† : death

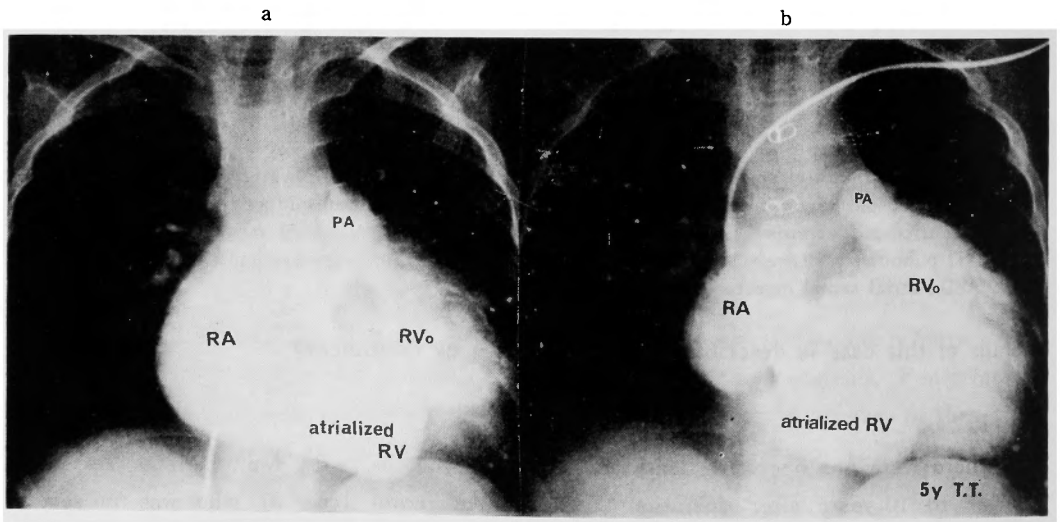

Fig. 2. This figure shows pre-and post-operative selective right atriography of case 4.
a : Large right atrium (RA) connecting to atrialized right ventricle, functional right ventricle (RV_0) and pulmonary artery, and pulmnary valvular stenosis are demonstrated.
b : Postoperative right atriography shows decreased volume of right atrium and atrialized right ventricle and increased volume of functional right ventricle.

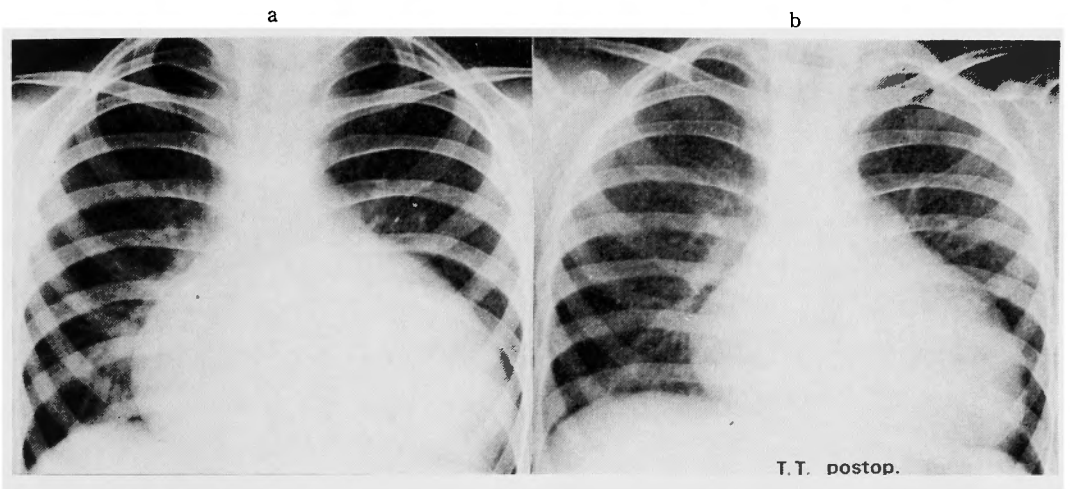

Fig. 3. This figure shows decrease of cardio thoracic ratio (CTR) after surgery of case 4.
a : preoperative x-ray (CTR . 76%) b : 1 year after surgery (CTR : 69%)

tion and and reopened communication between the atrialized and true right ventricle were obtained by angiocardiography. Conservative treatment by digitalis and diuretics were continued after his discharge, however, his physical activity gradually decreased and ventricular premature beat sometimes appeared. Necessity of reoperation was discussed, when he died suddenly probably due to dysrhythmia. A female patient (case 1, TVR) married after operation, experienced 2 natural labors 3 and 6 years after operation each, and is doing well after 10 years despite tachycardial attacks due to WPW syndrome (Fig. 4). A girl (case 2, TVR) is also doing well despite tachycardial attacks (WPW syndrome) nine years after operation. A male patient (case 2, TVR) is also doing well 9 years after surgery without physical limitation despite palpitation due to sinus premature beat (Fig. 5). A girl (case 7, TVP) is completely free from tachycardial attacks after division of Kent's bundle and doing well. In all four surviving patients cardio-thoracic ratio decreased, cyanosis disappeared and physical activity increased, however, arrythmias are continuing in 3 patients except for one whose bundle of Kent was divided (Table 1).

Discussion

Surgical treatment of Ebstein's anomaly consists of palliative and corrective surgery. As palliative procedure, superior vena cava to pulmonary artery anastomosis[26)29)34)], and closure of atrial septal defect[36)] have been performed, though their results have been uniformly unsatisfactory[8)20)22)26)33)34)]. Corrective procedures of Ebstein s anomaly are not simple because of its wide raging anatomical findings[2)4)5)11)19)21)23)].

Barnard et al[3)] performed tricuspid valve replacement without plicating the atrialized right ventricle placing the coronary sinus ostium on the right ventricular side. Lillehei et al[22)] recommended delayed closure of atrial septal defect after replacement of the tricuspid valve. Timmis et al[32)] emphasized the combined use of tricuspid valve replace-

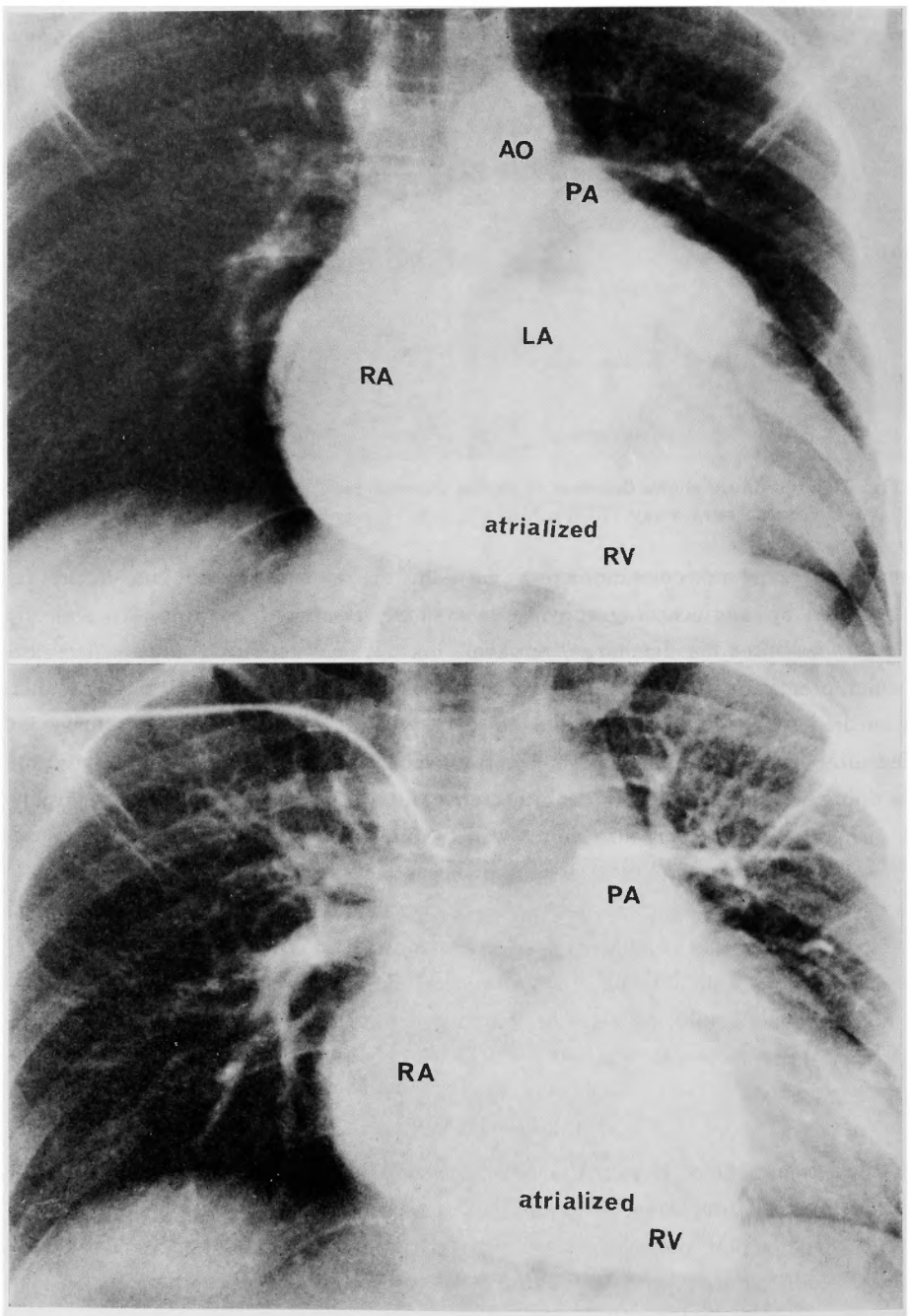

Fig. 4. This figure shows pre-and post-operative selective right atriography of case 1.

a : Preoperative right atriography shows enlarged right atrium (RA), large atrialized right ventricle, small functional right ventricle and pulmonary artery (PA). Left atrium (LA) and aorta (AO) are demonstrated simultaneously.

b : Postoparative right atriography shows decreased volume of right atrium and atrialized right ventricle. Pulmonary vasculature is filled sufficiently.

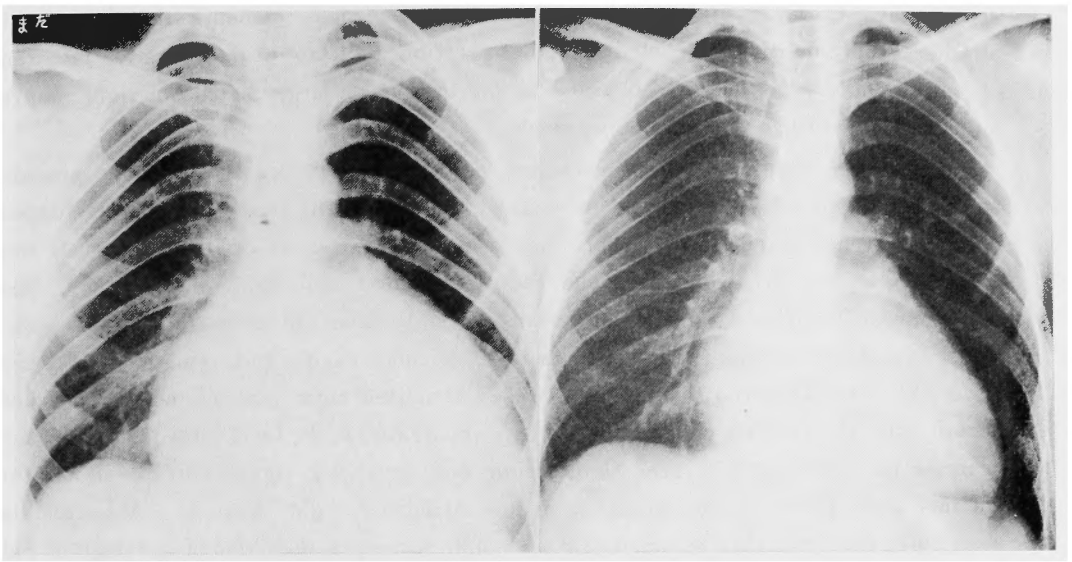

Fig. 5. Chest x-rays taken preoparatively and 9 years after tricuspid vulve replacement without obliterating atrialized right ventricle (case 2).

a : preoparative chest x-ray (CTR . 72%) b 10 years after surgery (CTR : 62%)
This patient has normal activity and no complaints except supraventricular arrhythmia.

ment, atrioventricular plication and atrioplasty. On the other hand, Hardy et al[12] first succeeded in valvuloplasty combined with plication of the atrialized right ventricle which was suggested by Hunter[14]. Hardy et al[13] reported further experience of valvuloplasty and emphasized superiority of his method. Bahson et al[2] discussed wide range of variety of valve deformity in this anomaly and proposed individualized choice of procedure. Recently McFaul et al[25] reported good results by obliteration of the atrialized right ventricle combined with tricuspid valve replacement.

Since 1968 we have practiced correcting this anomaly, and have obtained 4 long-term survivals out of 7 ; 3 of 4 TVR patients and 1 of 3 TVP patients. Better results were obtained in TVR group, however, we do not intend to indicate the superiority of TVR to TVP. Actually the plasty was performed in too severly ill patients (case 5 and 6), and moreover, we now feel that valvuloplasty was possible at least our earlier TVR cases (case 1 and 2).

However, we do not agree with Hady's opinion[13] that valvuloplasty is the choice of operation except for rare cases. Now we believe that it is necessary to do TVR considerably in many patients who have wide displacement of the tricuspid ring and severe dystrophy of the valve tissue and chordae tendineae as in our cases 5 and 6. As for the obliteration of the atrialized ventricle, we feel it not indispensable procedure because our long surviving 3 patients who had TVR without plication of the atrialized right ventricle showed decrease in cardio-thoracic ratio as well as improvement in physical activity. Furthermore, decreased cavity of right atrium and atrialized right ventricle were evidenced by selective right

atriography. However, it seems probable that reopened communication between the atrialized and true right ventricle behaved as contributing factor to postoperative recurrence of cardiac failure in our case 6 who had an excremely large atrialized right ventricle and in severe cardiac failure preoperatively.

From above mentioned experiences, we feel that the choice of operative procedure should be individualized in each case. Patient's disability with this disease largely depends upon right ventricular output. When the true right ventricular cavity is extremely small, obliteration of the atrialized ventricular cavity will cause right heart insufficiency. Space occupying artificial valves further aggravate the right heart function. In these cases, it may be reasonable to reserve the atrialized ventricular cavity and replace the tricuspid valve. We have observed that the thin walled atrialized right ventricle contraced simultaneously with the true right ventricle, though apparently weak. Long-term follow up showed decrease in cardio-thoracic ratio, right atrium and atrialized right ventricle in all three patients with TVR without plication of the atrialized right ventricle. Although right ventricular aneurysm that is sometimes seen after corrective operation of tetralogy of Fallot and a large right ventricular outflow patch actually impair ventrucular function to some extent, they do not induce right heart failure in many of these cases.

Furthermore, it is known that experimental destruction of free wall of the right ventricle in animals as long as the tricuspid valve function is maintained[1)6)16)31)]. However, improvement in ejection fraction by plicating the atrialized ventricle may benefit patients with extremely large immobile atrialized ventricular cavity.

It is also important to realize the conduction anomalies intrinsic to this unique anomaly[5)8)11)24)33)]. From our experience dysrhythmias including supraventricular arrhythmia, WPW syndrome associated with paroxysmal tachycardia, and complete right bundle branch block are not improved by so-called corrective surgery, and possibility of new dysrhythmia including postoperative A-V block is added. Danger of sudden deaths is not decreased after operation. In the natural histories of Ebstein's anomaly, occurrence of dysrhythmias increases with patient's age[5)] If the corrective surgery is done early in life, some of these dysrhythmias could be prevented. Division of Kent's bundle can be done which is reported to be localized at inferior margin of right ventricle[11)]. By these procedures, sudden deaths by dysrhythmia might be decreased in future. It was feared in earlier reports that dysrhythmias lead to patient's death during anesthesia and operation. In our experience, dysrhythmia including ventricular tachycardia and fibrillation occurred in all operated patients during anesthesia and operation, however, these were all controlled by means of antiarhythmic drugs, DC counter shock, or simple cardiac massage.

We are now considering to extend the operative indication of Ebstein's anomaly to patient with WPW syndrome with parorysmal tachycardia attacks, with moderate cyanosis, with moderate tricuspid regurgitation even in the absence of cardiac failure.

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和文抄録

エプスタイン奇型の外科治療

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エプスタイン奇型は先天性心疾患の中でも比較的稀な疾患であり，その手術適応および手術術式に関し尚多くの問題が残されている．われわれが1958年から1978年の間に京都大学第2外科および天理病院心臓血管外科において経験した7例の手術例を検討し，これに文献的考察を加えて報告する．

手術症例は男3例，女4例，手術時年令は24日から24才であった．新生児例をのぞき，全例術前に確定診断が行われた．全例にチアノーゼがみられ，NYHA臨床的重症度分類でIV度が2例，III度が3例，II度およびI度が各1例であった．心電図上WPW症候群を認めるもの3例，術前に発作性頻脈の病症のあるもの4例で特に第7例は臨床的重症度はI度であったが発作性頻脈が頻発することが手術適応とされた．

手術法では姑息的吻合手術はなく，全例いわゆる根治手術であったが，転位変形せる三尖弁に対する処置は，三尖弁置換術4例，三尖弁形成術3例であった．心房中隔欠損，心室中隔欠損，肺動脈狭窄症などの合併疾患に対しても同時に根治手術を行なった．

手術死は術前に心不全が強く確定診断を下し得なかった新生児例の1例のみであった．三尖弁置換および三尖弁形成術後のそれぞれ1例が術後約1年半で死亡し，現在生存中のものは弁置換3例，弁形成術1例である．何れも術後2年から10年を経過しているが，術前から存在した不整脈を除けば正常生活を営んでいる．弁形成術とKent氏束切断術を行なった症例は頻脈発作は全く消失した．また弁置換術の1例は2度の出産を無事に行なった．われわれの三尖弁置換術例は心房化右心室の縫縮を行なっていないが，術後心血管造影によって右心房および心房化右心室の縮小，胸部レ線像で心胸廓比の減少が認められている．

術前から存在するWPW症候群その他の上室性不整脈は，いわゆる根治術を行なっても治癒しないが，Kent氏束の検出切断を行なうことによりWPW症候群による頻脈発作を消失させることが可能であること，症例によっては弁形成術が有効であるが弁変形の著しい症例には弁置換術が必要であること，心房化右心室の縫縮は弁置換術の際必ずしも必須条件ではないことなどを確認し得た．

以上の結果われわれはエプスタイン奇型に対する手術適応は，合併する不整脈を含め，従来よりも拡大し得るものと考える．