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Central Palliation for the Tetralogy of Fallot*

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Fourteen cases of TOF were performed on the RVOT construction without closure of the VSD which was called central palliation. The cases consisted of an equal number of males and females. These patients ranged in age from six to forty-five years.

Before central palliation, initial palliative shunt was carried out in five cases. The remaining nine cases were provided with the central palliation as the initial surgery.

Twelve cases showed the ratio of PA/AO below one fourth or the ratio of RPA/AO below 0.3. Two cases showed an abnormal right coronary traversing the right ventricle. Resection of the abnormal right ventricle muscle band and pulmonary valvuloplasty were performed in nine cases (group A) and the placement of a patch graft was done in five cases (group B). Arterial oxygen saturation was increased in both groups postoperatively. The postoperative hematocrit was decreased in both groups. Postoperative PA/AO ratio became 1/3 in group A and 1/2.3 in group B, respectively.

Symptomatic improvement was good, especially in group B. The preoperative state of group B patients was classified as NYHA functional class IV. After central palliation, four cases changed to NYHA functional class II and one case could not be located during the follow up period.

Determination of the proper time for central palliation correction remains a problem that must be solved in the future. The time necessary for total correction will be about three or four years with occasional observation and follow up. (Received August 22, 1985 and accepted September 30, 1985)

Key words: Tetralogy of Fallot, Diminutive pulmonary tree, Central palliation, Patch graft

1 Introduction

Recent advances in preoperative evaluation allow us to gain good operative results for complex congenital heart diseases. Myocardial protection using potassium cardioplegia and postoperative management are also effective methods.

The total correction of TOF has been standardized with good results being obtained. Experience has shown us that one of the factor which is successful for total correction is the degree of development of the pulmonary arteries[1].

For TOF with diminutive pulmonary tree the RVOT construction without closure of VSD called central palliation, was performed.

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Abbreviations:

TOF	: Tetralogy of Fallot	RPA/AO	: right pulmonary artery and ascending aorta
RVOT	: right ventricular outflow tract	NYHA	: New York Heart Association
VSD	: Venricular Septal Defect	EPTFE	: Expanded polytetrafluoroethylene
PA/AO	: main pulmonary artery and ascending aorta		

The purpose of this communication is to discuss about the indications, postoperative results and prognosis of central palliation.

2 Patients and methods

Fourteen cases of TOF received the central palliation. The cases included an equal number of males and females. Age distribution was six to forty-five years (mean 21.5 years) at the time of central palliation administration. Before central palliation an initial shunt operation, such as Blalock-Taussig or Waterston operation, was performed in five cases. The remaining nine cases were provided with the central palliation as the initial surgery.

Results were expressed as mean±standard deviation. Student's t-test was used for statistical analysis. A P-value of less than 0.05 was considered significant.

3 Results

Twelve cases showed the ratio of PA/AO below one fourth or the ratio RPA/AO below 0.3. Two cases showed an abnormal right coronary artery traversing the right ventricle.

A resection of the right ventricle abnormal muscle band and pulmonary valvuloplasty were performed in nine cases (group A). Two cases of group A died due to low cardiac output syndrome at the time of total correction and brain abscess, respectively (Table 1).

A patch graft was used in five cases (group B), three cases of autopericardium, one case of autopericardium with EPTFE patch and one case of monocusp porcine pericardium stabilized with glutaraldehyde (Rygg) (Table 2).

Pre and postoperative arterial oxygen saturation was increased from 67.95±11.42% to 90.63±5.44% ($P<0.005$) in group A, from 63.72±6.81% to 86.38±8.25% in group B ($P<0.01$) respectively. Hematocrit was decreased from 55.75±6.84% to 43.81±5.93% ($P<0.005$) in group A, and from 58.6±5.5% to 47.52±9.23% in group B ($0.025<P<0.05$) respectively. Hemoglobin was not decreased significantly in group A, but decreased from 17.14±2.92 mg/dl to 15.34±1.95 mg/dl ($P<0.05$) in group B. Pre and postoperative red

Table 1 *Resection of the RVOT abnormal muscle band and pulmonary valvuloplasty (group A). F : Female M : Male*

Case	Age	Sex	Indication	O_2 Saturation		Hematocrit		Op.	Result
				Pre	Post	Pre	Post		
1	12Y	F	PA/AO (1/4)	73	–	55	46	Infundibulectomy & Pulmonary valvuloplasty	PO8Y (NYHA II)
2	18	M	PA/AO (1/5)	57	–	63	57	〃	cannot be located
3	10	M	PA/AO (1/4)	56.6	–	64	–	〃	〃
4	11	M	abnormal Rt. coronary A.	82	89	48	43	〃	PO3Y, Brain Abscess (Died)
5	28	M	PA/AO (1/4)	68	86	57	44	〃	PO7Y, Radical Surg (Died)
6	20	F	PA/AO (1/5)	59	–	58	42	〃	PO10Y (NYHA II)
7	6	F	PA/AO (1/5)	62	–	57	38	〃	PO11Y (NYHA II)
8	31	M	abnormal Rt. coronary A.	86	89	44	39	〃	cannot be located
9	39	F	–	–	98.5	–	41.5	〃	PO11Y, VSD closure, Af (NYHA II)

Table 2 *Patch enlargement of RVOT (group B).*

Case	Age	Sex	PA/AO	O_2 Saturation		Hematocrit		Outflow patch	Result (NYHA)
				Pre	Post	Pre	Post		
1	10	M	1 : 15	64.4	81.2	58	44.5	Pericardium	PO1Y6M (II)
2	13	F	1 : 5	57.7	77.5	54	52.0	Pericard. +EPTFE	PO1Y (II)
3	29	F	1 : 4	55.5	93.4	53	43.1	Rygg	PO1Y2M (II)
4	30	F	1 : 5	77.3	93.4	62	37.0	Pericardium	PO2Y (II)
5	45	M	1 : 5	—	—	66	61.0	Pericardium	cannot be located

blood cell count were not decreased significantly, in either of the groups.

The average follow-up period was 9.7 years in group A and 3.3 years in group B. One case in group A developed pulmonary hypertension (65/30 mmHg) which was demonstrated eleven years postoperatively by cardiac catheterization.

Two total corrections were performed in group A after seven and eleven years, respectively. While the former case resulted in death due to low cardiac output syndrome, the latter case ended in success by the transatrial patch closure of VSD and pulmonary valvuloplasty. She remains in NYHA functional class II with atrial fibrillation.

Preoperative states of group A patients were classified as NYHA class II in two cases and III in seven cases. Now four cases including one total correction changed to NYHA functional class II and the remaining three cases could not be located during the follow up period.

Preoperative states of group B patients were classified as NYHA functional class II and one case could not be located during the follow up period.

Postoperative PA/AO ratio became 1/3 in group A and 1/2.3 in group B respectively. Cardiothoracic ratio was not changed significantly in either of the groups. A decrease of right ventricular pressure and right-to-left shunt, increase of left ventricular end-diastolic volume were also noted.

4 Representative cases

Two representative cases were as follows.

Case 1: A 10 year old boy had undergone a left Blalock-Taussig shunt in another institution. The patient was admitted to our clinic for increased cyanosis. Angiogram showed an occlusion of the left Blalock-Taussig shunt and pulmonary atresia (Fig. 1).

PA/AO ratio at the time of operation was 1/15 and the diameter of the main pulmonfmy artery was 3 mm. Central palliation was provided using the autopericardium and it enlarged to the size of Hegar number 10. Postoperative angiogram disclosed the main and right pulmonary arteries (Fig. 2). One year and six months postoperatively the patient remains at NYHA functional class II.

Case 2: A 13.8 year old girl had undergone the Waterston shunt at the age of eight months and the left Blalock-Taussig shunt at 3.8 years respectively. Postoperative angiogram showed the opening of the left Blalock-Taussig shunt and a closing of the Waterston shunt. PA/AO ratio was 1/5. RVOT obstruction and right pulmonary artery stenosis were also noted (Fig. 3).

Central palliation by autopericardium with an EPTFE patch was selected. The patient remains at NYHA functional class II one year later (Fig. 4).

5 Discussion

Many patients with TOF can now be offered total surgical correction with a high probability of success.

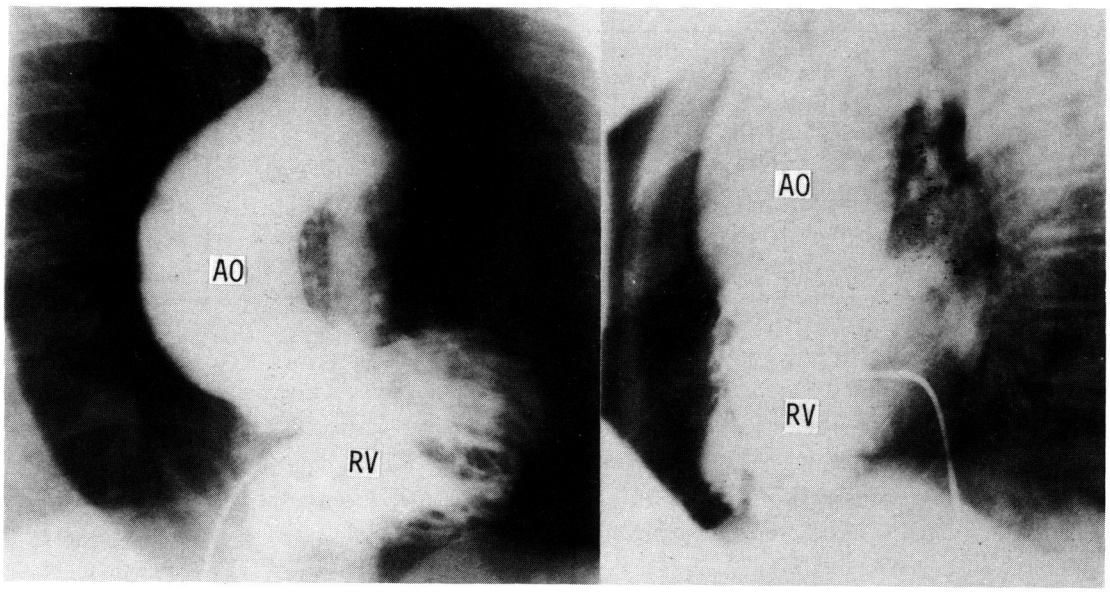

Fig. 1 10 year old boy. Preoperative angiogram showed the occlusion of the left Blalock-Taussig shunt and a poor development of the main pulmonary artery.

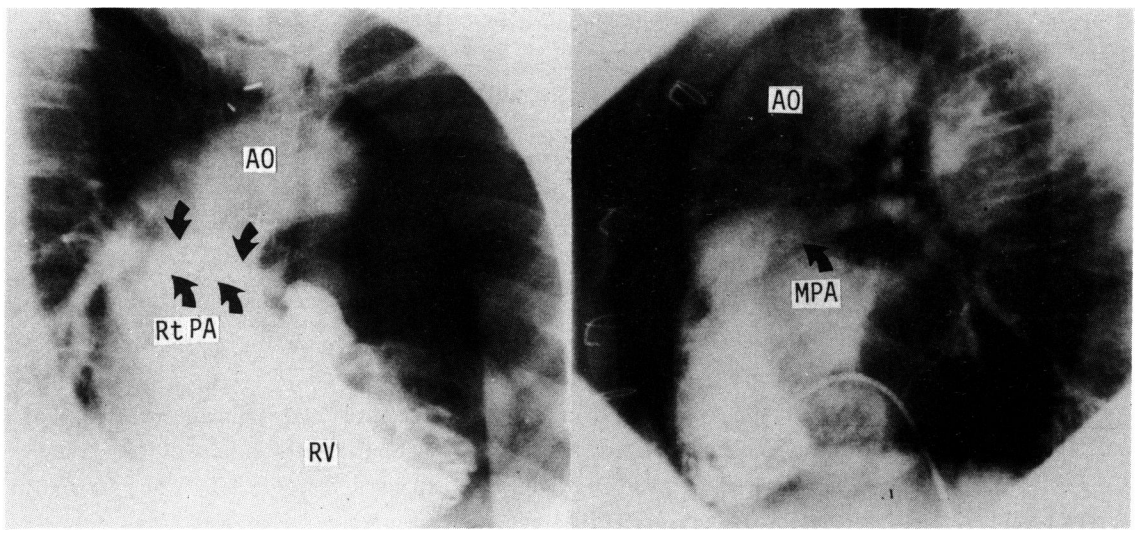

Fig. 2 Postoperative angiogram disclosed the main and right pulmonary arteries.

In the last four years, a total of 83 cases had surgical correction, including 66 cases of total correction, 13 cases of systemic-to-pulmonary shunt (2 cases of Blalock-Taussig shunt, 6 cases of modified Blalock-Taussig shunt, 5 cases of central shunt) and 4 cases of central palliation. Operative mortality of total surgical correction was 10.6%. The last 42 consecutive cases of total correction have ended in success in these past three years.

The first method of choice in palliative surgery for TOF is Blalock-Taussig shunt at our institution. If the pulmonary artery is too small to provide the Blalock-Taussig shunt, a modified Blalock-Taussig shunt

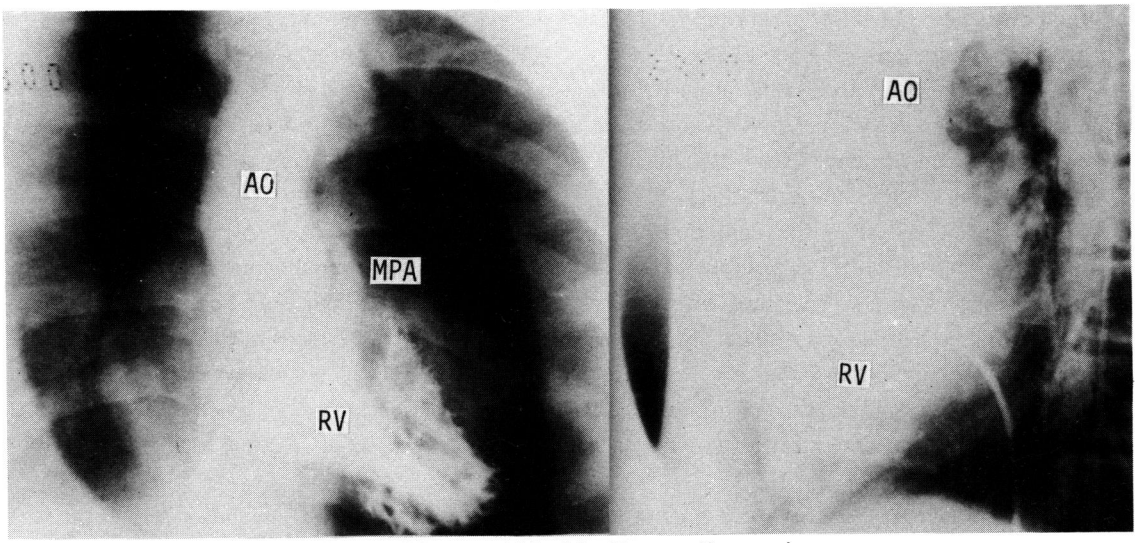

Fig. 3 13.8 years old girl. Preoperative angiogram showed that PA/AO ratio was 1/5.

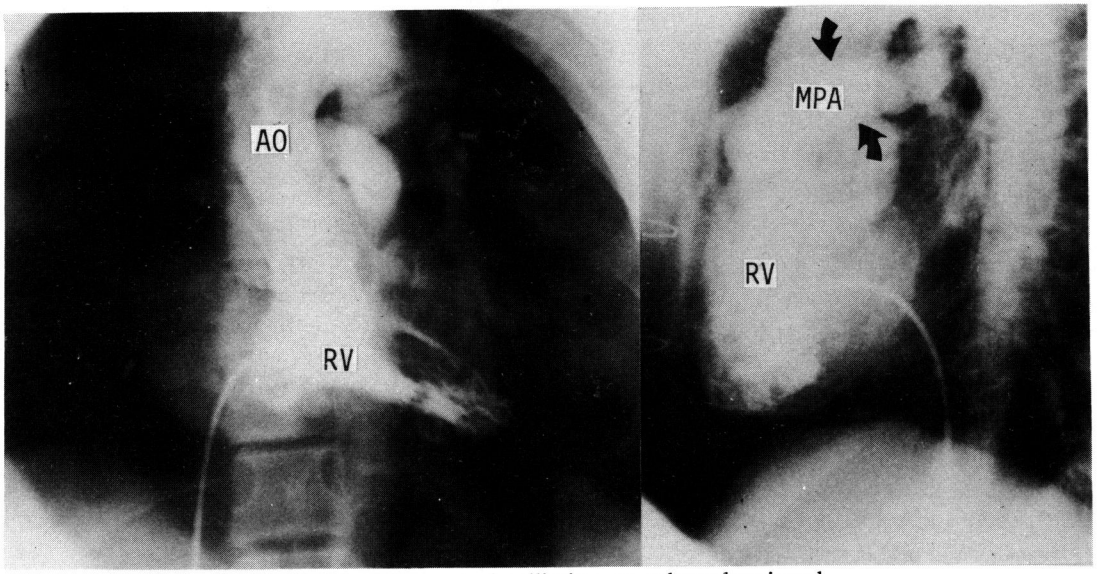

Fig. 4 After central palliation, an enlarged main pulmonary artery was noted.

or central shunt by the EPTFE graft are preferred. Various shunt operations have the problem of kinking the pulmonary artery, especially in Waterston shunt operation which sometimes require pulmonfry arterioplasty at the time of total correction of TOF. Brock's operation can obtain the physiological increase of pulmonary blood flow. But this operation cannot obtain enough pulmonary flow if severe RVOT obstruction exists. Damage to the aortic valve due to the blind procedure may occur. Recent advances in cardiopulmonary bypass operation allow safe RVOT construction by direct vision.

The indication of central palliation is found in the diminutive pulmonary tree whose ratio of PA/AO is below one fourth or the ratio of RPA/AO is below 0.3[2]. Poor pulmonary flow or unimproved symptoms after shunt operation and peripheral pulmonary artery stenosis are also added to the indication. In addition

abnormal distribution of the coronary artery, hypoplastic left ventricle and hypofunction of the kidney or liver must be considered. The age distribution of central palliation was over six years old.

The advantage of central palliation is the physiological pulmonary blood flow which is expected to develop in both pulmonary arteries[3]. Cyanosis and activity of the patient improves due to the reduction of the right-to-left shunt volume. Central palliation can avoid the kinking of the pulmonary artery which is sometimes seen after the shunt operation.

The disadvantages of central palliation are the postoperative pulmonary regurgitation and cardiac failure which occurrs if RVOT enlargement is set larger. The possibility of pulmonary hypertension exists if total correction is not performed over a prolonged period of time. The chance of brain abscess occurring at the time of infection through residual shunt may exist if total correction is not performed. So one of the problems of central palliation is the specific time when the total correction should be performed. The other problem is the adhesion at the time of total correction. An EPTFE sheet was used by Harada *et al.*[4] as an artificial pericardium after central palliation which made the total correction possible in a short operative time.

Akasaka[5] recommended that the degree of RVOT enlargement should be set smaller than at the time of total correction. Because of postoperative pulmonary regurgitation, cardiac failure arose due to the rapid increase of the left-to right shunt. He performed nine cases of central palliation and obtained good results in the cases which had a less than 40% left-to-right shunt after surgery.

Harada *et al,*[4] reported on six cases of central palliation for TOF, in which total correction seemed difficult to perform because of additional problems, including extreme hypoplasia of the pulmonary artery in two cases, anomalous of coronary arteries in three cases and a hypoplastic right ventricle in one case. Secondary total correction was successfully performed in all patients eleven to forty-four months after central palliation.

In our experience one case died due to brain abscess three years postoperatively and another case had pulmonary hypertension eleven years postoperatively. Group B had a better prognosis by use of a patch graft when compaired to group A. The time necessary for total correction will be about three or four years with occassional observation and follow up.

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ファロー四徴症に対する直視下右室流出路拡大術の検討

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ファロー四徴症の中で根治手術が困難と考えられた14例に対して，人工心肺使用下に右室流出路拡大術（直視下 Central Palliation）を行った．男女比は7：7で，年齢は6歳より45歳までであった．適応は①肺動脈発育不全例（PA/AO<$\frac{1}{4}$～$\frac{1}{5}$，RPA/AO<0.3），②各種姑息手術後の肺血流減少例，③前回短路手術後，なお有症状例，④左右肺動脈末梢部狭窄ないし発育不全例，⑤冠動脈走向異常例，⑥主要臓器に種々の二次的要因が加わった年長例であった．手術は右室流出路異常筋束切除と肺動脈弁形成施行（A群）が9例，流出路パッチ拡大術（B群）が5例であった．術後動脈血酸素飽和度は両群とも有意に上昇し，ヘマトクリット値も両群とも有意に低下した．術後A群のPA/AO比は1/3，B群は1/2.3に改善した．

本法は順向性に両側肺動脈へ生理的な血液供給が得られ，末梢肺動脈，左室の成長が期待でき，症状の改善が著明であった．特にパッチ使用群は全例，術前のNYHA重症度分類がIV度であったが，術後4例がII度（1例は不明）へ改善した．本法の根治手術（心室中隔欠損孔閉鎖）の時機は確立されていないが，3～4年後に予定している．