Title	Limited Capability of the Systemic-Pulmonary Shunt Alone to Prepare the Left Ventricle for Anatomic Correction in Simple Complete Transposition
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Limited Capability of the Systemic-Pulmonary Shunt Alone to Prepare the Left Ventricle for Anatomic Correction in Simple Complete Transposition

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Summary

To investigate the "preparatory" effect of systemic-pulmonary artery shunt on the left ventricle of simple complete transposition for future anatomic correction, seven infants underwent a modified BLALOCK-TAUSSIG (BT) shunt operation using a 4 mm Gore-Tex graft, and one infant long-term prostaglandin E1 (PGE1) administration. Ages at BT shunt ranged from 11 days to 8 months, with a median of 35 days. The BT shunt showed stable, improvable effects on re-hypoxia after BAS in all infants ($p<0.01$). However, only four infants had a good left ventricular preparation with a LV/RV pressure ratio of more than 0.80 at the expense of the occurrence of severe respiratory failure and pulmonary congestion due to a small interatrial defect. Three of the other four infants were not involved with respiratory failure, but their ventricular pressure ratio remained under 0.60. In seven infants, including two who later underwent additional pulmonary banding, anatomic correction was performed. One patient with the ratio under 0.60 died. Although six infants survived the anatomic correction after BT shunt despite prolonged aortic cross-clamping (mean: 134 min.), we would not avoid the conclusion that the BT shunt lacks the reliable ability to prepare the left ventricle for anatomic correction in all simple TGA. This is because severe respiratory failure developed in the cases having a small interatrial defect of 3 mm or less in diam. and it was a necessary evil to achieve an adequate LV/RV pressure ratio of more than 0.80, and because in some infants without respiratory failure the ratio remained less than 0.60.

Introduction

In recent years, there have been some reports on one-stage anatomic correction for complete transposition of the great arteries with intact ventricular septum (simple TGA) in the neonatal period,[5,14,17,18] but the primary procedure in neonates is not yet universally acceptable because of the still inadequate information on late surgical results. When two-stage correction is planned

Key words: Complete transposition of the great arteries with intact ventricular septum, Arterial switch correction, Blalock-Taussig shunt, Left ventricular preparation, Left to right ventricular pressure ratio.
索引語：完全大血管転位 I 型，解剖学的根治術，ブラロック・トウシッヒ短絡術，左室準備，左室右室圧比.
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for simple TGA, pulmonary artery banding (PAB) is now done in some institutes to prepare the left ventricle.[22] However, we believe that PAB would be disadvantageous both for severely hypoxic infants and in respect of postoperative adhesion and deformity of the banded pulmonary trunk.

Recently, we performed in seven infants the modified BLALOCK-TAUSSIG (BT) shunt operations[24] only, and long-term prostaglandin E1 (PGE1) infusion in one infant with simple TGA in an attempt not only to improve re-hypoxia after BAS but with the expectation of preparing the left ventricle. Thereafter, we performed anatomic correction[8] secondarily in six and primarily in one of the eight infants. In this report, we discuss the usefulness and dangers regarding the capabilities and limitations of BT shunt preparatory effects on the left ventricle in simple TGA.

Materials and Methods

1) Patients and clinical evaluation

Between May, 1984 and Jan., 1986, eight infants with simple TGA underwent two-stage anatomic correction after preliminary systemic-pulmonary artery shunt. Seven infants underwent modified left BT shunt using a 4 mm Gore-Tex graft. One other infant received long-term administration of PGE1 instead of a BT shunt. Balloon atrioseptostomy (BAS) was performed soon after admission in all patients and PGE1 was infused to neonates at a PaO2 level lower than 25 torr in room air[4,12]. A BT shunt was then performed for the following purposes: 1) in order to discontinue PGE1 administration when the infant had good intra-atrial mixing due to the obligatory shunt through the PGE1-dependent ductus, 2) to achieve the same obligatory shunt effect as the patent ductus when PGE1 was of no effect in neonates, or when the older infants showed a PaO2 level lower than 25 torr in room air, and 3) in expectation of an increase in blood flow through the left ventricle after BT shunt, that is, training the ventricle with much greater volume loading than during the preoperative state. Patients' ages at BT shunt ranged from 11 days to 8 months, with a median of 35 days.

The interatrial defect size was measured in each infant before the BT shunt was performed by two-dimensional echocardiography (2-DE). The presence or the degree of heart failure after the BT shunt was expressed as mild, moderate or severe, depending on clinical signs and symptoms. That is, the term "mild" was used when patients showed neither tachypnea nor pulmonary congestion on chest x-ray film and they were associated with only slight cardiac enlargement. The term "moderate" was used when patients showed both tachypnea and pulmonary congestion but did not require mechanical respiratory support. The term "severe" was used in those infants who needed mechanical support due to respiratory distress after the shunt. From the experiences we gained in patients with "severe" heart failure after BT shunt (patients #2 and #6), the long-term infusion of PGE1, by which the room air PaO2 level was maintained over 25 torr, was continued in one patient in place of BT shunt until the time of anatomic correction.

The left ventricular to right ventricular (LV/RV) pressure ratio and the left ventricular

Table 1. Pre-operative (before BT shunt) data in infants with simple TGA undergoing BT shunt or long-term PGE1 infusion

Patients	PGE1 administration	PaO2 (torr)	ASD size (mm in diam.)	LV/RV pressure ratio	(age)	age at BT shunt
1	−	25	5	···		2 m
2	+	23-25	3	···		17 d
3	−	38	***	0.41	7 m	8 m
4	+	···	4-6	0.90	1 d	35 d
5	+*	18	3	0.90	1 d	11 d
6	+*	22	2.5	···		11 d
7	+	44	4-5	0.90	27 d	35 d
8	+**	44	***	1.0	35 d	****

Abbreviations: *=ineffective to hypoxia;
**=infused until the time of correctlon;
***=large defect was confirmed at the time of correction;
****=primary correction after a long-term PGE1 infusion;
BT=Blalock-Taussig; TGA=complete transposition of the great arteries;
PGE1=prostaglandin E1; ASD=interatrial septal defect.

posterior wall thickness (LVPWT) in endsystole were measured after the shunt by catheterization and 2-DE. These data were utilized as important reference material with which to determine the indication for later anatomic correction.

2) Provisional settlement concerning indications for anatomic correction

Anatomic repair was performed without hesitation in patients with an LV/RV pressure ratio of 0.75 and over. For those patients in whom the ratio ranged from 0.5 to 0.74, the operation was performed under the condition that repeated observations of LVPWT by 2-DE revealed no diminutive tendency from birth onward. For those with the ratio less than 0.5, an atrial switch operation was performed. Anatomic correction was performed in seven patients in total, but only four patients had had prior BT shunt alone without any additional procedures.

3) Supplementary procedures at correction

All eight patients underwent rapid core cooling to below 20°C without surface cooling.

Table 2. Composition of blood-containing cardioplegic solution (employed for Case #2, #4, #5, #6, #7, #8.)

	Mean +/− SD		Range	
pH at 37°C	7.737	0.047	7,686	−7.809
PO2 (mmHg)	401	62	290	−466
Pco2 mmHg)	6.0	1.6	3.8	−7.3
B.E. (mmol/L)	−5.9	−3.5	−11.7	−2.9
Sodium (mEq/L)	81	5.8	73	−91
Potassium (mEq/L)	14.8	1.4	13.4	−16.8
Ionized Calcium (mmol/L)	0.17	0.08	0.11	−0.33
Hematocrit (%)	12.2	1.5	10.9	−14.2
Osmolarity (mOsm/L)	337	4.4	334	−343
Glucose (g/dl)	2.7	0.2	2.9	−2.5

Total circulatory arrest was introduced in all young infants to transect the great arteries at proper levels and to discreetly excise coronary artery ostia, including the surrounding aotic wall. The cardioplegic solution employed in all six infants, except Cases #1 and #3, was a sanguinous one that was carefully prepared so as to contain a constant concentration of each component at all operation times as listed in Table 2[3]. This solution was infused every half hour in 2–3 min with a one-time volume of 25–30 ml/kg. The temperature at the ventricular septum was maintained at 15–20°C. Aortic cross-clamp time at anatomic repair ranged from 108 to 184 min (mean: 141 min).

Results

1) Improvement of hypoxia

The PaO2 level in room air after the shunt increased in each patient to a range of 38–46 torr (mean: 40 torr) which was significantly higher than the preoperative level ($p<0.01$).

2) Occurrence of heart failure following systemic-pulmonary shunt

Six of the eight patients developed heart failure of a varing degree following BT shunt or PGE1 infusion as described in Table 3.

Four patients undergoing BT shunt whose interatrial defect was not less than 4 mm in diameter (#1, 3, 4, 7 in Table 1) showed no signs or only mild signs of heart failure with slight cardiac enlargement that were controlled easily with conventional management. None of these patients developed respiratory distress caused by pulmonary congestion.

The other four patients whose interatrial defect was 3 mm or less in diameter before the shunt or who had PGE1-dependent PDA developed pulmonary congestion and respiratory distress with tachypnea ranging up to 100/min (#2, 5, 6, 8 in Table 1). Patients #2 and #6 needed mechanical respiratory support. Especially for the latter, the interatrial defect could not be detected by 2-DE at the height of severe dyspnea. In infant #2, repeated palliative procedures such as the surgical creation of ASD, PAB, banding of the Gore-Tex graft, and finally the takedown of the BT shunt were done to relieve heart failure, but no alleviation could be achieved, compelling us to perform the JATENE operation[2]. Also in Case #6, judging from the complexities experienced during the postoperative course in the former case, a semi-emergency JATENE operation was performed three days after respirator connection.

3) LV/RV pressure ratio and LVPWT

In six post-BT shunt infants except Case #2, the LV/RV pressure ratio measured one to six months postoperatively ranged from 0.20 to 0.83. In Case 2, pulmonary to systemic (radial) artery pressure ratio was measured before PAB and its value was 0.98. Three patients had a ratio which was less than 0.60 and four had a ratio over 0.75, The patient undergoing long-term PGE1 infusion without the shunt had a ratio of 1.0.

All three patients with respiratory distress and pulmonary congestion had a ratio of over 0.80. The other four patients who did not experience respiratory failure had ratios ranging from 0.20 to 0.75, with three having a ratio less than 0.60. For Case #4 who had a ratio of 0.58, additional PA banding was performed in order to elevate the left ventricular pressure to a more

Table 3. Post-operative (after BT shunt) data and results of correction in the same infants as in Table 1

Patients	heart failure	PaO2 (torr)	LV/RV p.r.	(age)	LVPWT (ES) (mm)	(age)	Correction	(age)	Aortic clamp t. (min)	outcome
1	none	46	0.75	7 m	7.0	7 m	Jatene	10 m	108	alive
2	severe	36	0.98*	3 m	5.5	48 d	Jatene	3 m	117	alive
3	mild	40	0.20	15 m	7.0	11 m	Senning	15 m	0	alive
4	mild	39	0.58**	4 m	5.0	3 m	Jatene	12 m	129	alive
5	moderate	40	0.81	64 d	5.2	69 d	Jatene	2 m	144	alive
6	severe	38	0.83	40 d	5.3	39 d	Jatene	41 d	143	alive
7	none	40	0.54	78 d	4.8	3 m	Jatene	3 m	184	dead
8	moderate	···	1.0	35 d	4.5	35 d	Jatene	39 d	160	alive

Abbreviations: LV/RV p.r.=left ventricular to right ventricular pressure ratio;
LVPWT(ES)=left ventricular posterior wall thickness (end-systole);
*=pulmonary to systemic (radial) artery pressure ratio before PAB;
**=pressure ratio before PAB. The ratio after PAB was 1.0.

secure level due to the death of Case #7 after undergoing JATENE operation.

With regard to LVPWT, infants aged from one to three months at measurement had LVPWT value of 4.5 to 5.5 mm and two infants aged seven and eleven months each had values of 7.0 mm.

4) Switch operation

A venous or arterial switch operation was performed according to the provisional criteria described in Method 2). Seven of the eight patients underwent anatomic correction and the LECOMPTE maneuver[13] was accomplished in all. The remaining patient underwent the SENNING operation due to the extremely low left ventricular pressure. The type of coronary arteries was found to be JACOUB's type A[23] in all but two infants. The latter two types included a common origin for the right and left arteries from the posterior sinus in one (Case #7) and JACOUB's type D[23] in the other (Case #8). The left coronary artery in these two cases was distinctly smaller than in the other cases, having diameters of less than 1 mm. However, satisfactory transference of coronary arteries was accomplished in all cases, including the above two.

There was one operative death and no late deaths. The 3-month-old patient (#7) who died on the operating table developed severe left ventricular pumping failure and pulmonary congestion with an extremely elevated left atrial pressure at the time of weaning from bypass. Autopsy revealed no coronary artery abnormalities such as kinking. All six patients with the ratio of over 0.75 survived anatomic repair and their hearts came off bypass without difficulties including the case with the long aortic clamp time of 160 min.

The following complications occurred after anatomic repair: 1) transient left diaphragmatic nerve palsy (patient #1 and #5) 2) mediastinitis (#1). Spontaneous recovery was obtained in Cases #1 and #5 2 to 3 months after the operation. The latter was cured by continuous drainage and antibiotics.

Discussion

It has been suggested that the anatomic left ventricle has to be trained or prepared in order to tolerate the systemic work after anatomic correction in simple TGA[9,19,22]. From this viewpoint, YACOUB performed aggressively prior PA banding followed later by anatomic repair, obtaining favorable results[20,22]. More recently, from the viewpoint that primary repair can be accomplished without left ventricular preparation if in the neonatal period, CASTANEDA reported primary correction in 17 early infants including 12 neonates[5], YACOUB the same in 12 neonates[18], KANTER in 8 young infants associated with large PDA[10], and QUAEGEBEUR in 23 neonates[17], with survival rates of 82%, 92%, 88%, and 96%, respectively. With regard to PA banding for simple TGA in our hospital, however, we have been hesitant to perform it due to fears about the progression of postoperative hypoxia in preoperatively severe hypoxic infants, weakness and thinning of the banded pulmonary artery and the possibility of bleeding at re-anastomotic sites of the great vessels, and the lack of clarity regarding the proper transection level of the great vessels due to adhesion. From our recent first experience of marked improvement in hypoxia after BT shunt and the later successful anatomic correction in an infant[2], we began to study the ability of BT shunt to prepare the left ventricle for the tolerance of postoperative systemic work, as described in this report. However, in previous literatures we could not find any studies which indicated what kind of left ventriclar condition represented completion of the preparation. CASTANEDA et al. performed anatomic repair in patients with a pressure ratio over 0.61 and YACOUB et al. in patients with values over 0.58, however, in both reports there were no descriptions of the LV pressure or wall thickness for patients who died[5,18]. All of the patients with simple TGA in KANTER's report had associated PDA and he emphasized that the presence of PDA was important for maintaining the left ventricular capability[10]. QUAEGEBEUR reported that primary repair should be carried out within the first 2 weeks of life in patients without a large ductus[17]. One-stage successful correction for infants more than 1 month old with no ductus and with a pressure ratio under 0.50 was reported by GOOR in 2 cases (the ratio: 0.30, and 0.38)[7], by ABE in 1 case (0.38)[1], and by MAUK in 1 case (associated with small PDA, no pressure data were described after the neonatal period)[16]. With regard to LV wall thickness in TGA, there have been some available reports, but they do not offer practical considerations which include operative results[6,15]. Judging from the status quo as stated above, we decided to perform the JATENE operation in patients with a pressure ratio over 0.50 under the strict condition that they had never shown any diminutive tendencies with respect to the LVPWT. However, one pateint who had a ratio under 0.60 did not survive. When weaning from bypass, the patient's left ventricle showed severe pump failure in comparison with active contractility of the right ventricle. There were no differences in the cardioplegic composites and the method of infusion between the dead and surviving cases, nor could any explicable cause of death be found upon the transferred coronary arteries in the autoptic case. Cross-clamp time in the patient who died was 184 min, while the longest in the survivals was 160 min. However, the pressure ratio in the case with 160 min was 1.0. The permissible time of aortic clamping may be considerably shorter

for patients with a low pressure ratio. In this regard, GOOR et al. reported two successful corrections for simple TGA (6- & 9-month-old infants) with pressure ratios of less than 0.4 in which both patients had short aortic clamping times under 60 min[7].

We believe that there are two major factors which elevate left ventricular pressure and preserve the muscle mass, that is, fixed or dynamic left ventricular outflow obstruction (LVOTO)[21] and such a high volume loading of the left ventricle that elevates pulmonary pressure, as in the cases associated with PDA (including PGE1-dependent ductus in this paper) or large VSD. In our patients, there were no signs which suggested dynamic LVOTO by 2-DE during the period from BT shunt to total correction. We consider that factors which regulate BT shunt flow include the following; systemic arterial pressure, pulmonary vascular resistance, left atrial pressure, flow resistance at the site of the interatrial communication, right atrial pressure. If resistance at the interatrial defect plays a major part among the other regulating factors, there is the possibility that pulmonary-left ventricular circulation will have considerably higher blood flow postoperatively. In fact, each of our patients who developed respiratory failure after BT shunt had an extremely increased pulmonary blood flow status, although the evaluation was only qualitatively determined by 2-DE and PDE. With regard to wall thickness, KEANE et al. reported a significant correlation between left ventricular mass and pulmonary blood flow in simple TGA[11], and MARATO et al. reported a mean LVPWT value of 4.3 mm in systole by M-mode echocardiography in 53 patients with simple TGA[15]. Considering these reports, the mean LVPWT value of 5.7+/−1.0 mm observed in systole in our patients after the BT shunt may have relevance to the effects of the BT shunt itself. However, because an adequate left ventricular preparation after the BT shunt was only attainable in patients with a small interatrial defect by risking severe respiratory failure as described above, we cannot help but conclude that the BT shunt for simple TGA lacks universal acceptability from the standpoint of a safe and reliable "preparation", although it has the stable ability to improve hypoxia.

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和文抄録

完全大血管転位Ⅰ型における体肺動脈短絡術の“左室準備能力”に関する検討

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新生児3例を含む8例の，完全大血管転位Ⅰ型乳児例に対し，GoreTex 人工血管を用いた Modified Blalock-Taussig 手術又は prostaglandin E1 長期持続投与による動脈管開存により，体肺動脈短絡状態を作成し，BAS 後の rehypoxia の著明な改善を維持しつつ，将来の解剖学的根治術に備えての左室準備能力を検討した．8例中4例は，左室右室圧比0.8以上の左室準備状態を得たがいずれも肺うっ血を伴う呼吸不全を発症し，内1例は緊急根治術を余儀なくされた．呼吸不全を発症しなかった4例はいずれも左室壁厚に減衰傾向は認めなかったものの，内3例の左室右室圧比は0.6以下に留まった．呼吸不全の管理に難渋した症例は何れも心房間交通孔径が 3 mm 又はそれ以下であり，軽度呼吸不全例及び非発症例のそれは4 mm 以上であった．左室右室圧比0.5以上で且つ左室壁厚値の経時的観察で減衰傾向を認めない7症例に対し，同一心筋保護条件下に Jatene 手術を行った．左室右室圧比0.54，大動脈遮断184分の1例を除く他の6例は Jatene 手術を成功裡に行い得た．

心房間交通孔の小さい症例に短絡術を行い，術後慎重な呼吸循環管理を行いつつ Jatene 手術を行うという方針をとれば，短絡術単独でⅠ型症例を新生児期以後に Jatene 手術に導く事は本報告例のごとく充分可能ではあるが，高度呼吸不全が発症する可能性がある事，又心房間交通孔の大きい例では左室圧が低値に留まる例もある事から，短絡術単独の術後例全例に Jatene 手術を行う事は困難であり，左室準備状況に合わせた根治術式を選択すべきである．