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# Surgical Options in Congenital Heart Diseases in Infants Less than Three Months of Age; A Review of Seven Years' Experience

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**Abstract**: During a 7-year period from March 1984 to June 1991, 32 consecutive infants less than 3 months of age underwent 39 surgical procedures for congenital heart disease (both palliative procedures and one-stage repair) at the Yamanashi Medical College Hospital. Their ages ranged from 5 days to about 2 months.

There were 12 hospital deaths, with an overall mortality rate of 37%. Death rates in the one-stage repair and palliative surgery groups were 33% and 40%, respectively. Since 1989, the number of infants treated at less than one month of age (the neonatal period) has increased and the overall results have improved. Our recent operative mortality rate in infants less than 3 months of age was 24%.

We believe that the sooner such patients have an operation, even before the onset of symptoms, the better will be the outcome. This emphasizes the importance of early accurate enchocardiographic diagnosis and a short program of aggresive preoperative medical therapy. Finally, precise surgical treatment provides the best outcome in early infancy.

**Key words**: Congenital heart disease, Early infancy, Palliative, Corrective surgery

## INTRODUCTION

The advantages of early total correction of congenital cardiac defects now appear to be well accepted, and with the continuing progress in cardiac surgery more and more of these defects are being definitively treated in young infants. However, those infants who have either intractable congestive heart failure or severe cyanosis in very early infancy, especially at less than 3 months of age, still represent a formidable therapeutic challenge. In this group of patients, the results of either palliative or definitive surgery have been less than ideal.

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We felt it timely to review our operative experience at the Yamanashi Medical College Hospital in the 7-year period from March 1984 to June 1991, during which time a total of 41 consecutive infants less than one year of age were referred for the surgical treatment of congenital heart disease.

## PATIENTS AND METHODS

Between March 1984 and June 1991, we treated 41 patients under one year of age, including 32 consecutive infants less than 3 months of age (78% of all the infants), who underwent 39 surgical procedures (palliative surgery or one-stage repairs). There were 22 boys and 10 girls, and their ages ranged from 5 days to about 2 months. The major diagnosis were as follows: 5 cases of complete transposi-

tion of the great arteries (TGA), 6 cases of total anomalous pulmonary venous return (TAPVR), 1 case of congenital aortic valve stenosis (AS), 5 cases of isolated patent ductus arteriosus (PDA), 2 cases of pulmonary atresia with an intact ventricular septum (PA with IVS), 2 cases of pulmonary atresia with a single ventricle (SV), 2 cases of pulmonary stenosis (PS) with tricuspid atresia (TA), 2 cases of coarctation (COA) or interrupted aortic arch (IAA) with a ventricular septal defect (VSD), 3 cases of COA with TA or TGA, 1 case of complete A-V canal, 1 case of Taussig-Bing complex, 1 case of congenital mitral stenosis (MS) with tetralogy of Fallot (TOF), and 1 case of congenital A-V block. All patients underwent echocardiographic diagnosis prior to the operation, but cardiac catheterization was not performed in the recent of TAPVR, nor in the patients with PDA and AS.

*Preoperative preparation and anesthesia*

Many infants in this series were severely ill at the time of admission, and they required intensive medical therapy. Digitalis and diuretics were used in some infants, and prostaglandin $E_1$ was given intravenously to maintain ductal patency in many patients with severe hypoxemia or metabolic acidosis resulting from closure of the ductus Botallo. Some required intubation and assisted ventilation for respiratory distress.

Preanesthetic medication consisted of atropine, while anesthesia was typically inducted and maintained with intravenous fentanyl. Uncuffed endotracheal tubes were used routinely and were maintained for postoperative respiratory support. An arterial line was placed in a radial artery, and the arterial pressure and blood gases were determined before, during, and after surgery.

*Operative techniques*

Our operative indications for young infants have altered during this 7-year period, especially between the time before 1988 and that after 1989. As for corrective surgery, TGA has been corrected by the Jatene arterial switch procedure with Lecompte's maneuver scince 1990. TAPVR was corrected by a common pulmonary vein trunk-left atrial anastomosis with monofilament interrupted sutures has been improved. Aortic valve stenosis was managed by open direct valvotomy. PDA was simply ligated via an extrapleural approach. Among the palliative operations for complex heart disease, the classic Blalock-Taussig shunt was preferred at the contralateral site of the aortic arch. In failed cases, we used a modified Blalock-Taussig shunt with an expanded polytetra-fluoro-ethelene (EPTFE) graft that was 5 mm in diameter. Brock pulmonary valvotomy was done via the right ventricular outflow tract, but was not adequate in its effects. Coarctation repair was performed by the subclavian flap or EPTEE patch methods. Pulmonary artery banding was carried out according to the operative criteria of Shimada.

*Cardiopulmonary bypass technique*

All 12 patients with TGA, TAPVR, and AS underwent one-stage open-heart correction. During the first three years of the series (1984–1986), we had only two open-heart patients for TAPVR, who were managed by perfusion-induced deep hypothermia and total circulatory arrest. Since 1987, the remaining 10 infants have been operated on under conventional cardiopulmonary bypass with moderate hypothermia. An infant membrane oxygenator (Capiox-II, Terumo Co.) and a priming volume of 1.080 m*l* were employed. The priming solution consisted of ACD blood, acetate Ringer's solution, mannitol, sodium bicarbonate, calium chloride, insulin, CDP choline (Nicoline), and poloxamer 188 (Exocorpol). Myocardial protection during aortic clamping was performed by local myocardial cooling with ice water and crystalloid cardioplegia. However, blood cardioplegia was employed in the Jatene operation for TGA since it required prolonged cardiac arrest. The perioperative variables during cardiopulmonary bypass for the correction of TAPVR and TGA are summarized in Table 1. With our

Table 1. Perioperative variables during cardiopulmonary bypass (CPB) in the total correction of TAPVR and TGA

| Variables during CPB | | TAPVR (n = 6) | TGA (n = 5) | P value |
|---|---|---|---|---|
| Age | (days) | 19 ± 14 | 20 ± 11 | NS |
| Body weight | (Kg) | 3.0 ±0.7 | 3.0 ±0.3 | NSF |
| CPB time | (min) | 99 ± 24 | 250 ± 38 | P<0.01 |
| Anoxic arrest time | (min) | 70 ± 9 | 167 ± 28 | P<0.01 |
| Perfusion flow | (ml/kg) | 87 ± 15 | 111 ± 7 | P<0.01 |
| Mean perfusion pressure | (mmHg) | 53 ± 21 | 65 ± 17 | NS |
| Rectal temperature | (℃) | 21 ± 4 | 21 ± 3 | NS |
| Urine volume | (ml/h) | 10 ± 13 | 14 ± 18 | NS |

Abbreviations: CPB: cardiopulmonary bypass; TAPVR: total anomalous pulmonary venous return; TGA: transposition of the great arteries

present bypass method, a high perfusion flow (87–111 ml/kg/min) and mean perfusion pressure (53–65 mmHg) were maintained and a urine output of 3–4 ml/kg/h was obtained throughout the bypass procedure. The lowest body weight infant placed on cardiopulmonary bypass in this series was 53 days old, had a VSD and a previous coarctation repair, and weighted 2.1 kg.

## Results

Table 2 present the operative results in three age groups under one year and in the three periods from 1984 to 1991. Among the 32 consecutive infants less than 3 months of age in this series, there were 12 hospital deaths and an overall mortality rate of 37%. Since 1989, the number of patients less than one month old (so-called "newborn" babies) has increased and the operative results have also improved in general.

The operative results following one-stage correction or palliative surgery are shown in Table 3. There were 18 one-stage corrections and 15 palliative operations performed for operative mortality rates of 33% and 40%, respectively. The most common cause of early death was the low cardiac output syndrome (LOS) which occurred in 6 cases of TGA or TAPVR undergoing one-stage corrections. Of the 6 patients who died after palliative operations, congestive heart failure was the cause of early death in one patient with interrupted aortic arch complex and one with complete A-V canal. A lack of improvement of hypoxemia was encountered in one infant with PA plus IVS after the Brock operation and in another with MS and TOF after the Shuster operation. One sudden death occured in a 22-day-old boy immediately after epicardial pacemaker implantation for congenital A-V block.

The operative results for TGA were summarized in Table 4. There were 5 patients aged 10 to 38 days (average; 20 days) in the TGA group. All had TGA with an intact ventricular septum and no associated anomalies apart

Table 2. Operative results with mortality rates in infants less than one year of age from 1984 to 1991 at Yamanashi Medical College Hospital

| Period | Age | | | total mortality |
|---|---|---|---|---|
| | 0~28 days | 1~2 mon | 3~12 mon | |
| 1984~85 | 2 (2)<br>100% | | 3 (2)<br>67% | 5 (4)<br>80% |
| 1986~88 | 2 (2)<br>100% | 7 (3)<br>43% | 5 (1)<br>20% | 14 (6)<br>43% |
| 1989~91 | 14 (4)<br>29% | 7 (1)<br>14% | 1 (0)<br>0% | 22 (5)<br>23% |

( ) operative deaths

Table 3. Operative results for various congenital cardiac lesions after total correction or palliative procedures in infants less than three months of age

| | No of patients | Operative death | Mortality |
|---|---|---|---|
| A. Correction (18 cases) | | | |
| TGA | 5 | 3 | |
| TAPVR | 6 | 3 | 6/18 |
| VSD* | 1 | 0 | 33% |
| AS | 1 | 0 | |
| PDA | 5 | 0 | |
| B. Palliation (15 cases) | | | |
| CoA repair | 5 | 1 | |
| B-T shunt op | 6 | 2 | 6/15 |
| PA banding | 2 | 1 | 40% |
| Miscellaneous | 2 | 2 | |

VSD*: A patient with VSD had previous coarctation repair.
Abbreviations: PDA: patent ductus arteriosus; COA: coarctation of the aorta; B-T: Blalock-Taussig; PA: pulmonary artery.

Table 4. Clinical details of infants undergoing Jatene's correction for complete transposition of the great arteries (TGA)

| | Case 1. | Case 2. | Case 3. | Case 4. | Case 5. |
|---|---|---|---|---|---|
| Age at operation | 38 days | 20 days | 15 days | 17 days | 10 days |
| Sex | male | male | male | male | male |
| Body weight (Kg) | 2.6 | 2.7 | 3.0 | 3.3 | 3.2 |
| Associated anomalies | PDA<br>ASD | closed PDA<br>PFO | closed PDA<br>ASD | PDA<br>PFO | PDA<br>PFO |
| $PGE_1$ infusion | fair effect | no effect | no effect | strong-effect | good effect |
| BAS | no | yes | no | yes | (no) |
| LVSP/RVSP (mmHg) & ratio | 62/73<br>0.85 | 42/58<br>0.70 | 45/70<br>0.63 | 72/78<br>0.92 | no cathe |
| Type of op. | semi-emergency | elective | elective | semi-emergency | semi-emergency |
| Anoxic arrest time (min) | 146 | 170 | 214 | 151 | 156 |
| Operative result | alive | DOT (LOS) | DOT (AMI) | early death (CHF) | alive |

Abbreviations: PGE: Prostaglandin E; ASD: atrial septal defect; PFO: patent foramen ovale; BAS: balloon atrioseptostomy; LVSP/RVSP: left and right ventricular systolic pressure; DOT: death on table; LOS: low cardiac output syndrome; AMI: acute myocardial infarction; CHF: congestive heart failure.

from PDA. Left ventricular systolic pressure just before the Jatene operation was maintained well because of ductal patency with or without prostaglandin $E_1$ therapy in Cases 1, 4 and 5. However, in two patients (Cases 2 and 3), the ventricular systolic pressure ratios were lower (0.70 and 0.63, respectively). Three of these patients died less than 12 hours after the operation. Case 2 was a 20-day-old boy with a preoperative left ventricular pressure below 42 mmHg at the time of the Jatene operation–he died of LOS. We thought that an earlier one-stage operation or a two-stage operation following left ventricular pressure training (PA banding) should be recommended. In Case 3, a technical error of coronary artery switching resulted in acute myocardial infarction and he died on the operating table. Case 4 was a 17-day-old infant with severe congestive heart failure who developed anuria preoperatively because of a high flow rate PDA and the unnecessary use of $PGE_1$. An urgent Jatene operation was done consequently, but the low cardiac output failure could not be improved. Preoperatively, this infant was thought to be unsuitable for one-stage open heart repair. The two other patients had uneventful postoperative courses.

The operative results for TAPVR are shown in Table 5. There were 6 infants aged 5 to 45 days (average; 19 days) in the group with TAPVR. In 5 of the 6 cases (except for Case 3 with type IIa TAPVR), the clinical features of pulmonary venous obstruction were obvious. They had type Ib or cor triatriatum in two cases, and type IIIa in three cases. All the patients underwent echocardiography prior to the operation, but Cardiac Catheterization was performed only in the earliest two cases (Case 1 and Case 2), where the operations were managed under perfusion-induced deep hypothermia combined with total circulatory arrest. There were three operative death due to the low cardiac output syndrome, probably resulting from a PDA remnant which was not ligated (Cases 1 and 2), and from aspiration on the 15th postoperative day in Case 3. The more recent 3 patients have survived this operation.

Table 6 showed the details of PDA surgery

Table 5. Clinical details of infants undergoing correction of total anomalous pulmonary venous return (TAPVR)

| | Case 1. | Case 2. | Case 3. | Case 4. | Case 5. | Case 6. |
|---|---|---|---|---|---|---|
| Age at operation | 5 days | 8 days | 17 days | 45 days | 25 days | 16 days |
| Sex | male | female | female | male | male | male |
| Body weight (Kg) | | | | | | |
| at birth (g) | 3,120 | 2,670 | 3,455 | 3,854 | 3,290 | 2,334 |
| at operation (g) | 3,200 | 2,500 | 3,210 | 4,120 | 2,734 | 2,300 |
| Type of TAPVR | I b | Cor triatriatum | II a | III a | III a | III a |
| | ASD | PFO | PFO | ASD | ASD | PFO |
| Diagnosis by | echo | echo | echo | echo | echo | echo |
| | cathe | cathe | | | | |
| Emergency op. | yes | yes | semi- | yes | semi- | semi- |
| PDA ligation | no touch | no touch | yes | yes | yes | yes |
| Anoxic arrest | 70 | 60 | 67 | 70 | 68 | 86 |
| time (min) | c̄ total arrest | c̄ total arrest | | | | |
| Operative | LOS death | LOS death | Respiratory | alive | alive | alive |
| results | 2nd day | 2nd day | trouble death | | | |
| | | | 15th day | | | |

Table 6. Clinical details of infants operated on for patent ductus arteriosus (PDA)

| | Case 1. | Case 2. | Case 3. | Case 4. | Case 5. |
|---|---|---|---|---|---|
| Age at operation | 13 days | 26 days | 54 days | 57 days | 78 days |
| Sex | male | female | female | female | female |
| Gestational age | 40 wk | 29 wk | 27 wk | 37 wk | 38 wk |
| Birth weight (Kg) | 3,656 | 935 | 915 | 3,156 | 2,660 |
| Weight at operation | 3,280 | 795 | 1,460 | 3,470 | 2,000 |
| CTR (%) | 64 | 62 | 53 | 66 | 69 |
| Associated anomaly | | | BPD | ASD cleft lip | 46XX, $3^{q(+)}$ |
| Mefenamic acid | —— | no effect | no effect | —— | —— |
| Operative results | alive | alive | alive | alive | alive |
| Weight at discharge | 5,170 | 2,830 | 3,230 | 4,170 | 3,970 |
| (months postop) | (1.5 mon) | (3 mon) | (4 mon) | (4 mon) | (3.5 mon) |

in infants less than 3 months of age. There were 5 patients aged 13 to 78 days in this group. Their weights at simple ligation of the ductus ranged from 795 to 3,470 grams. Operations for premature infants were required in two cases (Cases 2 and 3), after pharmacological closure with mefenamic acid had failed. The operative results were satisfactory.

## Discussion

Of every 1,000 babies born alive, eight to ten have a congenital heart defect. At least one third of these children become critically ill during the first year of life and either die of heart disease or require surgical treatment. According to an earlier study[1], death from congenital heart disease mostly occurs within the first few months of life and in fact mainly within the first weeks after birth.

Because of the perceived high risk of open

heart surgery early in life, critically ill neonates have traditionally been treated with a first-stage palliative procedure[2]. However, recent advances in pediatric cardiac surgery have allowed early primary complete repair to be performed in many infants with congenital heart malformations. In fact, the advantages of early total correction of congenital cardiac defects now appear to be well accepted, with the continuing progress in cardiac surgery, more and more such congenital defects are being corrected in early infancy[3]. On the other hand, a substantial number of infants still require a palliative procedure to increase or decrease pulmonary blood flow during early infancy, particularly in the under three month of age group[4)–8)].

Although only small number of our patients are infants less than 3 months of age, our operative results have improved cosiderably for both palliative and corrective surgery for congenital heart disease in the neonatal period. But they are still less than ideal. The foregoing analysis of our operative deaths indicated that 4 out of 12 were due to technical errors, 4 were due to the improper selection of operative methods, 3 were due to a moribund preoperative state, and the other one was due to an error in postoperative respiratory care. The deaths in 6 patients with open-heart surgery were not believed to have been related primarily to the use of standard cardiopulmonary bypass. However, we now feel that it is not necessary to use total circulatory arrest for any corrective procedure, even in neonates. Two cardiac lesions (TGA and TAPVR) have the highest incidence of requiring early surgery in neonates, and primary repair is generally accepted as being desirable. Therefore, it is very important to reduce the operative mortality rate in patients with TGA and TAPVR.

In summary, Castaneda *et al.* of the Boston Children's Hospital group[3] have strongly emphasized that if the preoperative hemodynamic and metabolic states are reasonable and if the cardiac lesions lend themselves to repair, the neonate is in fact a resilient patient capable of overcoming serious adversity. They have been encouraged by the continously decreasing hospital mortality rate for arterial switch operations in neonates with TGA with IVS[9)10)]. An important objective of reparative cardiac surgery for any age group is obviously survival, but equally important (particularly in children) is the achievement of optimal lifelong function. In this respect, it is true that either the one- or two-stage operation has achieved a considerable improvement.

On the basis of his clinical experience to date, Castaneda[3] continues to favor the repair of congenital heart disease in symptomatic neonates rather than the use of palliative surgery. In addition to the already outlined anatomic and physiologic factors in favor of early repair, he stated that there are also important family and societal considerations. Primary repair reduces parental anxiety and decreases the burden of caring for a chronically ill child. There are also economic advantages to early primary repair, since further hospitalization is theoretically avoided or minimized. We expect that the elective repair of neonates with complex congenital heart disease will become an achivable therapeutic goal in the not-too-distant future.

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