

## Surgical Management of Tetralogy of Fallot at NICVD\*

KALIM UDDIN AZIZ, MRCP (E),  
M. REHMAN, FRCS,  
F. REHMAN FCPS and  
A. D. MEMON, M.B.B.S.

Tetralogy of Fallot is a complex malformation which is characterized clinically by Central cyanosis and varying degree of exercise intolerance. There are two main anatomic components of this malformation, a) Infundibular chamber hypoplasia and stenosis due to anterior deviation of crista supraventricularis (Parietal Band or Infundibular Septum with its abnormal fusion to the superior limb of Trabecula septomarginalis 1-3) b) Ventricular septal defect, which is usually large and subcrystal and is created by the septal malalignment of the crista and is therefore usually of para membranous and infundibular type (2). The right ventricular Hypertrophy is a consequence of infundibular stenosis and overriding of the aortic root across the ventricular septum occurs to a varying degree.

The degree of exercise intolerance is proportionate to the severity of infundibular stenosis and cyanotic spells are due to intermittent spasm of the infundibulum. The life expectancy without surgical correction is greatly reduced in patients with Tetralogy of Fallot (4,5). We present our experience with surgical management of patients with Tetralogy of Fallot from January, 1980 to October, 1983 at the National Institute of Cardiovascular Diseases, Karachi.

### MATERIAL AND METHODS

One Hundred and fifty patients age 0.1 to 26 years were included in the Study. Ninety Two patients had primary correction which was performed using cardiopulmonary bypass, moderate hypothermia and cardioplegia. The repair involved closure of ventricular septal defect with a dacron patch and resection of infundibular stenosis through a vertical ventriculotomy. The right ventricular outflow tract was roofed with a transpulmonary annular pericardial patch in all

but three patients. In these three patients a conus coronary artery was found to arise from the right coronary and crossed the infundibulum anteriorly to continue as anterior descending artery (6). In these cases resection of the infundibular stenosis was performed from across the pulmonic valve and ventricular septal defect was closed through right atrial approach.

Pericardial patch in the right ventricular outflow tract was extended into the main pulmonary artery when required. Two patients were noted to have a supracristal or doubly committed ventricular septal defect. (Fig. 1) (7-8). This defect was

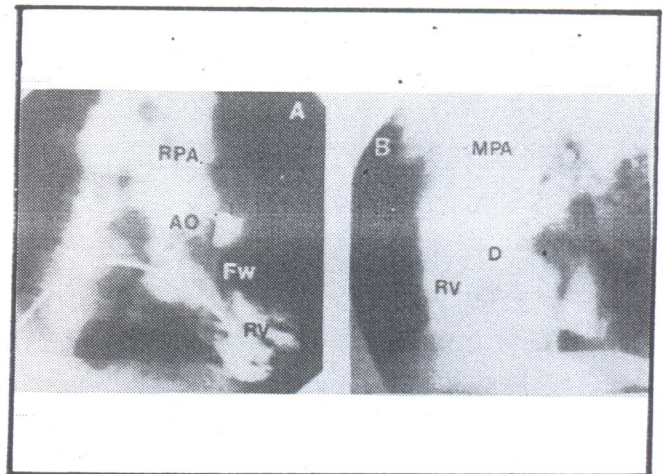


Figure - 1.

A. Right ventricular (RV) cine angiogram, right anterior oblique axial view, in a patient with Tetralogy of Fallot and Supracristal ventricular septal defect. Simultaneous opacification of Aorta (Ao) and pulmonary artery is seen. The main and right pulmonary arteries are enlarged and a jet of contrast through a stenotic pulmonic valve.

The usual cristal filling defect between the pulmonary artery and aortic root is absent suggesting the presence of supra cristal ventricular septal defect.

FW = Free Wall of the Right Ventricle.

B. Lateral view right ventricular (RV) cine angiogram of Patient in Pannel. Moderately large supra Cristal ventricular septal defect (D) is present at the subpulmonary area.

\*The National Institute of Cardiovascular Diseases, Karachi.



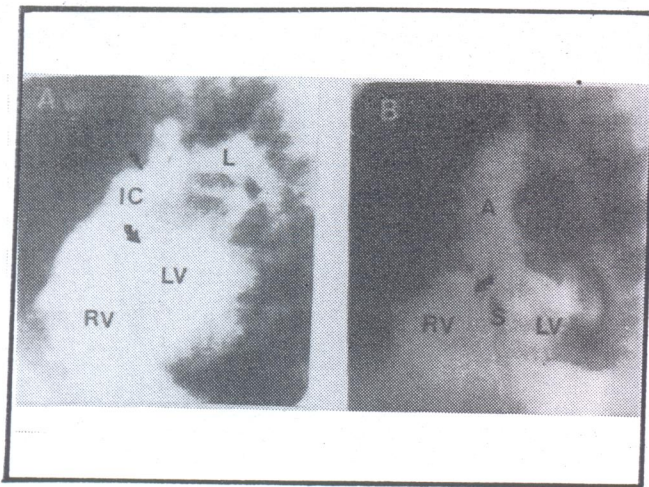


Figure - 2.

A. LATERAL VIEW : Right ventricular cine angiogram in a patient with typical features of Tetralogy of Fallot. A well developed Infundibular chamber (IC) is seen with domed stenotic pulmonic valve (Triangular arrow). A subcrystal or malalignment ventricular septal defect is well delineated (curved arrow). Slight supra valve stenosis of the main pulmonary artery is present (Triangular arrow). LV = left ventricle.

B. Lateral view left ventricular (LV) cine angiogram in a patient with tetralogy of Fallot showing the position of subaortic ventricular septal (arrow). Mild aortic (A) over ride is seen. S = Septum, RV = Right Ventricle.

closed with a dacron patch and resection of the infundibular stenosis was performed through a vertical ventriculotomy and a pericardial patch was inserted to enlarge the valve annulus.

Fifty eight patients age 29 days to 19 years underwent shunt operations. Blalock - Taussig and waterston-cooley shunts were performed during earlier period and recently Gore-Tex graft was instituted between the subclavian and pulmonary arteries (Modified Blalock Taussig shunt).

The indications for shunt operation were a) age less than 3-4 years b) weight less than 13 Kg and c) Hypoplastic pulmonary arteries or a small left ventricle. During early period shunt surgery was performed on older children who pre-operatively were having cyanotic spells or had Hematocrit (PCV) value greater than 66 per cent.

The clinical diagnosis of Tetralogy of Fallot was confirmed by two dimensional sector scan echocardiography and cardiac catheterization in all patients. Sector scan echocardiograms were obtained in an unselected patients in supine position. Various planer projections of sector beam

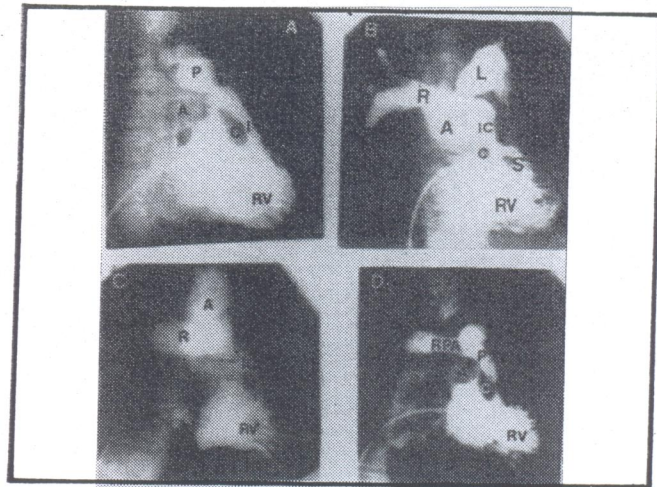


Figure - 3.

Right ventricular Cine angiogram (AP view) of 4 patients with tetralogy of Fallot (A-D) showing variable degree of hypoplasia of the infundibular chamber.

A. Long Infundibular chamber (1) with domed pulmonic valve and a well developed cristal (C) is seen.

Compared to the patient in Pannel A.

B. Relatively Shorter infundibular chamber length (IC) is shown. R = right pulmonary artery - L = Left pulmonary artery, S = Septum.

C. Still shorter IC and short main pulmonary artery and moderate supra valve narrowing (dotted). Crista (C) separates the pulmonary artery from aorta.

D. Greater severity of infundibular stenosis. Note the Infundibular chamber is narrowed to a string. Thickened domed pulmonic valve suggest concomittent pulmonic valve stenosis. A = aorta. P = Main pulmonary artery. RPA = Right pulmonary artery.

from various locations were employed for cardiac imaging (19).

Cardiac Catheterization was performed after pre-medication with chlorpromazine - phenergan - pethidine mixture. Axial angiography (10) was employed to image the heart. (Fig. 2, 3, 4). Angiographic Quantitation of the relative size of the main pulmonary Artery (MPA), right pulmonary artery (RPA), Ascending sortia (AA) and descending aorta (DA) was made by tracing the systolic frame angiographic images on a plain paper. The main pulmonary artery diameter was measured above the level of sinuses and in cases of discrete supra valve narrowing of the main pulmonary artery the measurements were obtained distal to the narrowing. Ascending aorta diameter was measured above the sinuses and

TABLE - I.

## ANGIOGRAPHIC QUANTITATIVE DATA OF 51 PATIENTS WITH T.O.F.

|                                                           | RPA/AA      | RPA/DA      | MPA/AA      | MPA/DA      | DA/AA       |
|-----------------------------------------------------------|-------------|-------------|-------------|-------------|-------------|
| SHUNTING GROUP :                                          |             |             |             |             |             |
| n = 15                                                    |             |             |             |             |             |
|                                                           | 0.44 ± 0.16 | 0.78 ± 0.34 | 0.42 ± 0.12 | 0.73 ± 0.19 | 0.58 ± 0.08 |
|                                                           | n = 15      | n = 15      | n = 14      | n = 14      | n = 15      |
| PRIMARY CORRECTION GROUP :                                |             |             |             |             |             |
| n = 14                                                    |             |             |             |             |             |
|                                                           | 0.51 ± 0.13 | 0.93 ± 0.19 | 0.56 ± 0.21 | 0.98 ± 0.28 | 0.52 ± 0.08 |
|                                                           | n = 39      | n = 40      | n = 30      | n = 29      | n = 36      |
|                                                           | P < 0.1     | P < 0.05    | P < 0.05    | P < 0.001   | P < 0.02    |
| ALL PATIENTS :                                            |             |             |             |             |             |
| n = 56                                                    |             |             |             |             |             |
|                                                           | 0.48 ± 0.14 | 0.89 ± 0.24 | 0.51 ± 0.2  | 0.89 ± 0.28 | 0.54 ± 0.09 |
|                                                           | n = 53      | n = 55      | n = 44      | n = 50      | n = 51      |
| P = Comparison of Shunting and Primary correction groups. |             |             |             |             |             |

TABLE - II

## ANGIOGRAPHIC QUANTITATIVE DATA OF 41 PATIENTS WITH PRIMARY CORRECTION

|                 | RPA/AA      | RPA/DA      | MPA/AA      | MPA/DA      | DA/AA       |
|-----------------|-------------|-------------|-------------|-------------|-------------|
| NON SURVIVORS : |             |             |             |             |             |
| n = 14          |             |             |             |             |             |
|                 | 0.48 ± 0.09 | 0.88 ± 0.22 | 0.53 ± 0.35 | 0.80 ± 0.32 | 0.52 ± 0.06 |
|                 | n = 13      | n = 14      | n = 8       | n = 8       | n = 12      |
| SURVIVORS :     |             |             |             |             |             |
| n = 27          |             |             |             |             |             |
|                 | 0.52 ± 0.14 | 0.95 ± 0.17 | 0.59 ± 0.18 | 1.0 ± 0.2   | 0.5 ± 0.1   |
|                 | n = 26      | n = 26      | n = 24      | n = 22      | n = 25      |
|                 | NS          | NS          | P < 0.1     | P < 0.1     | NS          |



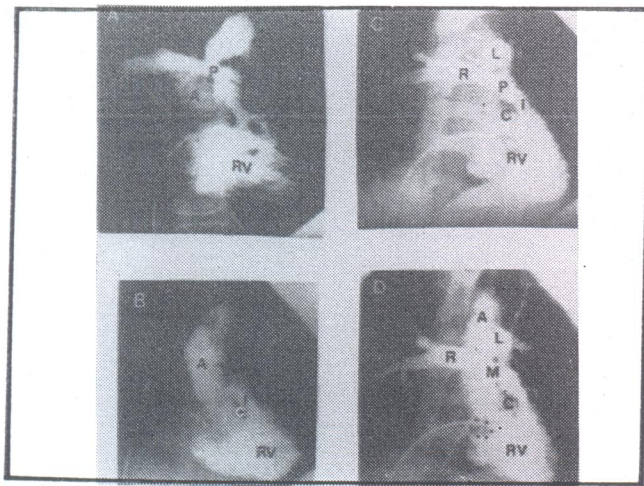


Figure - 4.

Right ventricular Cine angiograms of four patients with Tetralogy of Fallot, showing variable anatomy of the pulmonary vasculature.

A. Typical appearance of pulmonary vasculature. Main pulmonary artery is short and moderately Hypoplastic. The Hypoplasia extends to Proximal right and left branches. Distally right and left pulmonary arteries show dilatation. A = Aorta. P = Pulmonary Artery. C = Crista.

B. Markedly Hypoplastic main pulmonary artery is shown with Moderately Hypoplastic right and left pulmonary arteries ( R & L). A = Aorta. I = Infundibular chamber.

C-D. Mild ostial stenosis of left pulmonary artery (L) is shown (arrow) in pannel C and Dotts show the position of ventricular septal defect in pannel D. M = Main Pulmonary Artery. R = Right pulmonary artery.

descending aorta diameter was determined at the level of the diaphragm. The right pulmonary artery diameter was measured at a point mid way between its origin and first branching at the Hilum. Actual measurements were possible in 17 patients in whom catheters image was used to correct for X-Ray magnification. In 39 patients catheter size was not recorded therefore ratios of RPA/AA, RPA/DA, MPA/AA, MPA/AA and  $\Gamma$ A/AA diameters were calculated in all 56 patients. Forty one of these 56 patients underwent primary correction and 15 underwent shunting operations. Angiographic length of the infundibular chamber was measured between the lower edge of the cristal defect and the pulmonary valve annulus during diastole in an extended right anterior oblique angiograms in 17 patients in whom catheter image magnification factor could be determined.

Early mortality was defined as intra-operative

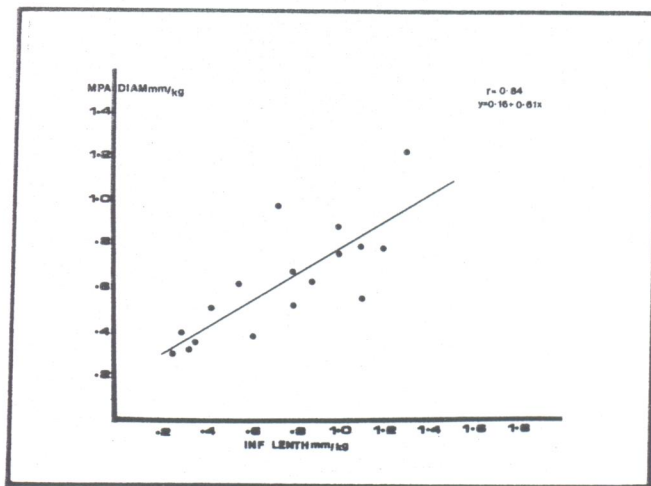


Figure - 5.

Angiographically measured normalised diastolic infundibular chamber length (mm/Kg) plot against normalised Main pulmonary artery (MPA) diameter (mm/Kg.) A statistically significant linear relationship is present suggesting that shorter infundibular chamber length is associated with proportionately shorter main pulmonary artery diameter.

or within the hospital stay. Average hospital stay period for an uncomplicated patient was 14 days.

The mortality data was available in all 150 patients but complete analysis of data was available in 121 patient; 74/92 patients underwent primary correction and in 47/58 patients had shunting operations.

## RESULTS :

### ANGIOGRAPHIC QUANTITATIVE ANALYSIS

In 56/150 patients with Tetralogy of Fallot quantitative analysis of the angiograms showed that right pulmonary artery (RPA) was approximately one half the diameter of the ascending Aorta (AA); (RPA/AA ratio =  $0.49 \pm 0.19$ ); and mean RPA diameter was slightly smaller than the mean descending aorta diameter, (mean RPA/descending Aorta (DA) ratio =  $0.9 \pm 0.24$ ). The mean diameter of the main pulmonary artery (MPA) was the same as RPA, MPA/AA and MPA/DA ratios were  $0.5 \pm 0.16$  and  $0.89 \pm 0.28$  respectively. Ascending aorta diameter was approximately twice the diameter of the descending aorta; DA/AA ratio was  $0.54 \pm 0.09$  (Table - I).

In 41 of the primary correction group the RPA and MPA diameters were similar, RPA/AA and MPA/AA diameter ratios were  $0.57 (\pm 0.13)$  and  $0.56 \pm 0.21$  respectively. Ascending aorta



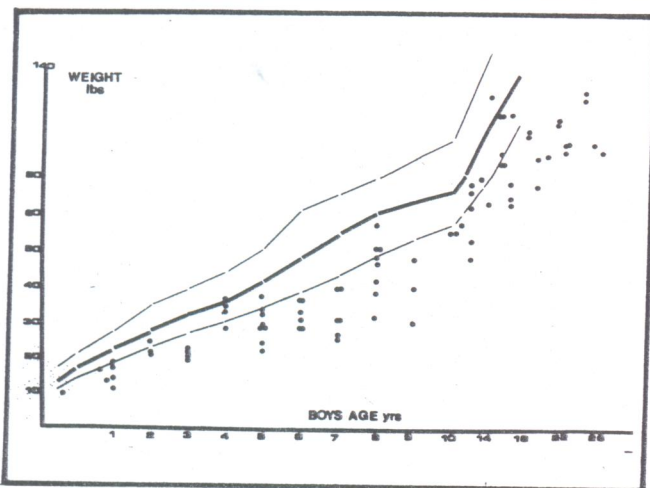


Figure - 6.

Body weight scattergram of 83 male patients with Tetralogy of Fallot against the normal 3rd-50th and 97th per centile limits (Harvard and Iowa Public health Standards). Majority of patients are below the 3rd per centile in all age groups.

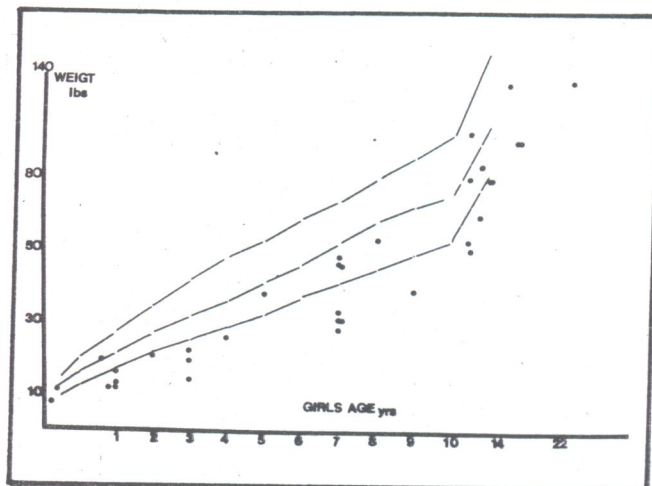


Figure - 7.

Body weight scattergram of 34 Female patients with Tetralogy of Fallot against the 3rd-50th and 97th normal percentiles. Like the Males in Fig. 6, majority of Female patients are well below the 3rd per cents.

diameter was approximately twice the diameter of the descending Aorta. Table-I. The diameters of the right and main pulmonary arteries was similar in 27 survivors of primary correction group compared to 14 non survivors (Table-II).

In 21 shunting group patients the RPA/AA and MPA/AA ratios were similar, ( $0.44 \pm 0.16$  and  $0.42 \pm 0.32$  respectively) and compared to patients who underwent primary correction the RPA and MPA diameters were significantly smaller  $P < 0.05$  and  $P < 0.001$  respectively. Table-I. The AA in shunting group was significantly larger ( $P < 0.02$ ) compared to the primary repair group patients suggesting greater infundibular stenosis and larger right to left shunt resulting in larger AA. Table-I.

In 17 patients actual measurements of the angiographic diastolic length of the infundibular chamber were available, Table-III.

The mean diameter of the MPA was  $11.1 \pm 4.1$  mm and mean diastolic infundibular chamber length was  $12.7 \pm 4.4$  mm. In these 17 patients normalised MPA diameter (mm/Kg body weight) was correlated with normalised diastolic infundibular chamber length (mm/Kg body weight). Linear correlation was noted indicating that short infundibular chamber length was associated with proportionally smaller MPA diameter ( $Y = 0.163 \pm 0.61x$ ;  $r = 0.84$ ;  $P < 0.001$ ). Fig.5.

The weights of these 150 patients with Tetralogy of Fallot were compared to normal standard weights and showed that majority of infants

and children were below the third percentile. The lower weight was more pronounced in younger patients but similar trend was noted in all age groups (Fig. 6 and 7).

#### SHUNTING OPERATION GROUP :

The mean age of 47/58 patients who underwent shunting operations was  $4.6 \pm 4.3$  years (range 29 days to 19 years). Detailed analysis of aged showed that three infants were less than 3 months old and one baby was 29 days old (mean  $0.11 \pm 0.2$  years). Five infants were between 3 - 12 months age (mean  $0.88 \pm$  years) and 39 children were greater than 1 year age (mean  $5.4 \pm 4.3$  years). Table-IV.

In 43/47 patients mean weight was  $8.7 \pm 3.2$  Kg and 4 patients had weight greater than 22 Kg. In 47/58 patients the mean Haemoglobin value was  $20.7 \pm 3.5$  G per cent.

All of the 58 patients undergoing shunting operation had cyanotic spells and were taking oral propranolol (Inderal) till the morning of operation in the mean dose of  $2.2 \pm 0.9$  mg/Kg body weight per day.

#### RESULT OF SHUNTING OPERATIONS :

During 1980 period six patients underwent shunting operation with one death, a survival rate of 83 per cent. In 1981 the number of shunt operations increased to 16 with the survival rate of 56 per cent. In 1982, 25 shunting operations

TABLE - III

**ANGIOGRAPHIC QUANTITATIVE DATA OF THE DIASTOLIC LENGTH OF THE INFUNDIBULAR CHAMBER**

| No. | Age           | Infundibular Length |                 | Main Pulmonary Artery diameter |                 |
|-----|---------------|---------------------|-----------------|--------------------------------|-----------------|
|     |               | mm                  | mm/Kg           | mm                             | mm/Kg           |
| 1.  | 1.5           | 6.0                 | 0.8             | 5.0                            | 0.67            |
| 2.  | 19.0          | 14.5                | 0.33            | 14.5                           | 0.33            |
| 3.  | 3.7           | 12.9                | 1.1             | 6.6                            | 0.55            |
| 4.  | 11.0          | 8.7                 | 0.26            | 10.4                           | 0.31            |
| 5.  | 2.2           | 8.4                 | 0.88            | 5.9                            | 0.62            |
| 6.  | 7.0           | 10.0                | 1.0             | 8.7                            | 0.87            |
| 7.  | 6.5           | 11.4                | 0.8             | 7.4                            | 0.53            |
| 8.  | 4.0           | 15.2                | 1.3             | 14.4                           | 1.20            |
| 9.  | 9.0           | 20.0                | 1.1             | 14.0                           | 0.78            |
| 10. | 15.0          | 15.4                | 0.30            | 20.8                           | 0.40            |
| 11. | 13.0          | 9.6                 | 0.36            | 9.6                            | 0.36            |
| 12. | 8.0           | 22.0                | 1.0             | 16.0                           | 0.73            |
| 13. | 2.0           | 8.0                 | 0.73            | 10.7                           | 0.97            |
| 14. | 7.0           | 12.0                | 0.55            | 13.6                           | 0.62            |
| 15. | 8.0           | 14.3                | 0.62            | 8.8                            | 0.38            |
| 16. | 9.0           | 9.2                 | 0.43            | 11.0                           | 0.51            |
| 17. | 6.0           | 17.6                | 1.20            | 11.2                           | 0.77            |
|     | $7.8 \pm 4.7$ | $12.7 \pm 4.4$      | $0.75 \pm 0.34$ | $11.1 \pm 4.1$                 | $0.62 \pm 0.25$ |

were performed and the survival rate improved to 72%. Further improvement in surgical survival from shunting operations was seen in 1983 when only one of 11 patients died, a survival rate of 92 per cent. The over-all survival for the entire 1980 - 1983 period was 72 per cent. (Table-V).

#### FACTORS AFFECTING SURVIVAL FROM SHUNT SURGERY :

Type of shunting operation had significant effect on survival. Eighteen patients who had Blalock-Taussig shunt had survival of 61 per cent and a similar survival rate (63%) was noted for sixteen patients undergoing waterston-cooley shunt. The survival rate however was significantly better (87%) for 24 patients who had Gore-Tex graft shunt Table-VI.

Age of the patient was not a significant factor for survival. The survival rate for 9 infants less than one year age was 75% comparable to 69% survival for 39 children greater than 1 year of age. It may be noted that infants under three

months were very few.

The mean age of 14 non survivors for shunt operation was  $5.2 \pm 4.2$  years and was not significantly different when compared to the mean age of  $4.2 \pm 4.4$  years of 34 survivors. Analysis of Hemoglobin data showed that it was not statistically significantly different for 33 survivors ( $20.3 \pm 3.6\%$ ) compared to 12 non survivors ( $22.1 \pm 2.89\%$ ).

#### PRIMARY CORRECTION GROUP :

Primary correction was performed on Ninety two patients but complete data was available in 74 of these. The mean age was  $11.9 \pm 5.8$  years (range 4 - 26 years). The mean weight was  $38.8 \pm 13.3$  Kg (range 13 - 59 Kg) and mean hemoglobin value was  $18.8 \pm 3.8$  G%.

Pre operatively eighteen of the 74 patients undergoing complete correction had hypercyanotic spells. The mean age of these patients was  $5.5 \pm 4.9$  year. All of the 74 patients had varying degree of exercise intolerance. Mild degree of exercise intolerance defined as inability to parti-



TABLE - IV

SHUNT SURGERY FOR TETRALOGY OF FALLOT

1980 - 1983

| Age (Months)       | Total No. | Died No. | Survival % |
|--------------------|-----------|----------|------------|
| < 3 (0.1 ± 0.19)   | 3         | 0        | 100        |
| 3-12 (0.88 ± 0.08) | 5         | 2        | 60         |
| > 12 (53 ± 43)     | 39        | 12       | 69         |
| Total :            | 47        | 14       |            |

TABLE - VI

SHUNT SURGERY FOR TETRALOGY OF FALLOT

1980 - 1983

| Type             | Total No. | Died No. | Survival % |
|------------------|-----------|----------|------------|
| Blalock-Taussig  | 18        | 7        | 61         |
| Waterston-Cooley | 16        | 6        | 63         |
| Gore-Tex Graft   | 24        | 3        | 87         |
| Total :          | 58        | 16       | 72         |

icipate in school or Mohalla games was present in six patients. Moderate intolerance i.e. in ability to attend school or take short walks outside of home was present in 27 and severe exercise intolerance involving inability to walk even short steps at home was noted in 41 patients.

RESULTS OF PRIMARY INTRACARDIAC CORRECTIVE OPERATION

In 1980 thirteen patients had primary correction operation and six died, a survival rate of 54 per cent. Since then the survival rate had steadily increased from 75% to 88% in 1983. The over-all survival rate for the 1980 to 1983 period was 72%. Table-VII.

TABLE - V

SHUNT SURGERY FOR TETRALOGY OF FALLOT

1980 - 1983

| Period  | Total No. | Died No. | Alive No. | Survival % |
|---------|-----------|----------|-----------|------------|
| 1980    | 6         | 1        | 5         | 83         |
| 1981    | 16        | 7        | 9         | 56         |
| 1982    | 25        | 7        | 18        | 72         |
| 1983    | 11        | 1        | 10        | 92         |
| Total : | 58        | 16       | 42        | 72         |

TABLE - VII

PRIMARY CORRECTION FOR TETRALOGY OF FALLOT

1980 - 1983

| Period  | Total No. | Died No. | Alive No. | Survival % |
|---------|-----------|----------|-----------|------------|
| 1980    | 13        | 6        | 7         | 54         |
| 1981    | 28        | 7        | 21        | 75         |
| 1982    | 33        | 9        | 24        | 72         |
| 1983    | 18        | 2        | 16        | 88         |
| Total : | 92        | 24       | 68        | 72         |

FACTOR AFFECTING SURVIVAL FOLLOWING PRIMARY CORRECTION :

In 26 patients greater than 12 year age (mean 18.7 ± 4.1 year) survival rate of 81% was not statistically greater compared to 68% survival rate in 44 patients less than 12 year age (mean 8.5 ± 2.4 year) P < 0.1.

There was also no age difference amongst 19 non survivors (mean age 10.2 ± 5.8 year) compared 52 survivors (mean age 12.8 ± 5.7 year) P < 0.1. The weight of the patients undergoing primary correction was analysed with the view to



determine its impact on surgical survival. Fifty five survivors had greater weight ( $31.8 \pm 13.8$  Kg) compared to 16 non survivors (mean weight  $25.8 \pm 12.9$  Kg)  $P < 0.05$ ). The smallest patient who had total correction was 13 Kg in weight at 4 years age. Hemoglobin value was not significantly different for 16 non survivors (mean Hemoglobin  $19.8 \pm 2.6$  G) compared to 52 survivors in whom mean Hemoglobin value was  $18.6 \pm 4.0$  G%).

Twelve of 72 patients were taking propranolol till the day of operation. The mean dose was  $2.2 \pm 0.94$  mg/Kg/day. Three of these died a statistically insignificant difference from 14 deaths from amongst 60 patients who were not taking propranolol ( $P < 0.5$ ).

The immediate cause of death could be analysed in 17 of the 24 deaths. Nine patients developed a low cardiac output syndrome, three died of post operative bleeding, one developed disseminated intravascular Coagulopathy due to generalised sepsis, one died due to fluid imbalance and one developed complete heart block and low cardiac output. Two patients died due to difficulties in weaning from the ventilator.

#### POST-OPERATIVE FOLLOW-UP :

Fourteen of the 42 survivors (33 per cent) from the shunting operation group returned at the follow-up out patient clinic.

The mean follow-up period was  $10.6 \pm 8.3$  months. The shunt was functioning in all of these with marked improvement in cyanosis. One additional patient was seen in the clinic and recatheterised due to a clinically non functioning shunt. However angiographically it was patent but small. Sixteen of the 68 survivors (23 per cent) who had primary repair returned for follow-up. The mean follow-up period was  $10.8 \pm 10.4$  months. Fourteen of these sixteen patients had result which were classified as good when assessed by mild residual murmur of Pulmonary insufficiency, near normal cardiac size on chest X-Ray and Sinus Rythm on Electrocardiogram. One patient developed congestive cardiac failure due to residual ASD and moderately severe pulmonary insufficiency. The remaining one patient developed bacterial endocarditis on the tricuspid valve.

Both responded well to medical therapy. Two of these 16 patients developed transient complete heart block during hospital stay but resumed sinus rythm at 2 - 4 weeks post-operatively.

#### DISCUSSION

Pediatric cardiac surgery was started on regular basis at NICVD in 1980. In the earlier period of our management of patients with Tetralogy of Fallot we deliberately selected older children for shunt surgery, who were severely symptomatic and severely cyanosed. However in the later period criteria for shunt operation and primary correction were developed. Primary repair was under-taken if the body weight was greater than 13 Kg and pulmonary arteries were not hypoplastic and the left ventricle was of adequate size. Our surgical experience shows that shunt operations were performed on patients who were severely cyanosed with recurrent cyanotic spells. The number of shunt operations had increased steadily during each succeeding year as has the survival rate. The mortality from shunting operations under three months of age specially during new born period is reported to be high (12). In this study infants under 3 months of age were few in number and consequently survival in infants less than 1 year age was not significantly different compared to greater than 1 year age patients. The data further suggests that age, weight and level of Haematocrit did not adversely affect survival following shunt operations. The most important factor for surgical survival was the type of shunting operation. The survival was best achieved with Gore-Tex Graft shunt between the subclavian and pulmonary arteries and the later mortality was comparable to the reported experience (12). The reason seems to be largely technical since compared to classical Blalock-Taussig or waterston-cooley shunt Gore-Tex shunt is easier to institute and can be performed on either side in the chest. It would be easier to take down at the time of primary repair. Presently we undertake Gore-Tex shunt operation in all patients beyond the new born period i.e. greater than 1 month age.

Our angiographic quantitative data showed that the patients who were selected for primary correction had comparatively larger pulmonary arteries than those who underwent shunting operation, although the selection for primary correction was not based on the quantitative data.

A post-mortem morphometric study by Becker et al (3) showed that the main pulmonary artery was approximately one half the size of the ascending aorta in 14 hearts with Tetralogy of Fallot. This data is similar to ours however they



reported that the length of the infundibulum of hearts with Fallot's Tetralogy was larger compared to normal hearts. They normalised the infundibular chamber length with right ventricular dimension which in tetralogy hearts has been reported to be smaller compared to the normal hearts (13). Furthermore normal control hearts were not properly matched for weight and age with Tetralogy of Fallot hearts.

There is evidence to suggest that growth of the cardiac structures is proportionate to the body weight (13). Consequently we normalised the infundibular diastolic length with the weight and showed that the normalised length of the infundibular chamber in patients with Tetralogy of Fallot was directly proportionate to the diameter of the main pulmonary artery. This we believe lends credence to the hypothesis that short infundibular chamber may be the primary anatomic abnormality in the morphogenesis of Tetralogy of Fallot malformation.

Over the past two and half year period of our surgical experience the survival rate for primary correction for Tetralogy of Fallot has steadily improved. This is in conformity with early experience with primary correction operation in most major centres (15-22). The mortality from primary correction performed during 1955 to 1968 at the Hospital for Sick Children at Toronto, Canada was 35 per cent and from 1965 onwards declined to 12 per cent (14). The early experience with primary correction operation at Mayo Clinic was reported by Fuster et al (15) and showed that during 1955-1964 period the mortality was 17%. The over all mortality at Houston Texas reported by Garson et al was 11 per cent (16).

Recently however greatly improved survival from primary correction operation has been reported at these and other centres even in the young infants (17-20) and adolescents (21).

Our initial experience with primary correction operation compares favourably with these reports. The survival rate for the first six months period in 1983 was 88 per cent and the over all survival for 2.5 years period was 72 per cent.

As with shunting operation greater number and smaller age patients are being operated upon each year. Age does not seem to be a factor affecting survival from primary correction in patients greater than 4 years including adolescents and adults (19). This data is consistent with reported experience. We did not operate on patients less than 4 year age when age has been shown to be an incremental risk factor.

Tetralogy of Fallot is the second most common lesion amongst children and infants referred to the N.I.C.V.D. and majority of patients present with cyanotic spells. All of these patients are treated with Propranolol (Inderal) so that surgery can be undertaken electively. Our data shows that administration of propranolol till the morning of surgery did not significantly affect mortality. Consequently we of necessity continue to use propranolol for medical treatment of cyanotic spells.

We have used transpulmonary annular pericardial patch to enlarge the right ventricular outflow tract in majority of our patients without significant problems with post operative pulmonary insufficiency. It is reported that pericardial patch may not be required if the pulmonary diameter is near half the diameter of Ascending aorta and further more pericardial patches in the right ventricular outflow tract may indeed be an adverse incremental risk factor in smaller babies and children less than 4 years of age. (19) We are now more selective in the use of pericardial patch for enlargement of the right ventricular outflow tract. Three of our patients were found to have conus Coronary Artery crossing the outflow tract to continue as anterior descending artery. In these patients satisfactory resection of the infundibular stenosis was achieved through transpulmonary and trans tricuspid valve approaches. The ventricular septal defect was closed through the right atrium. The standard practice of repair of these cases is to insert a conduit between the right ventricle and pulmonary artery thus by passing the right ventricular outflow obstruction (6). The use of Hypothermia and Cardioplegia now allows for a great relaxation of the heart and thus this approach of resecting the right ventricular outflow tract without ventriculotomy need to be retried since the integrity of conduit over succeeding years can not be assured. It is concluded that over the 2.5 year of our surgical experience greater and younger patients are undergoing palliative and corrective operations with improved survival.

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