Surgical Experience with Congenital Heart Disease at National Institute of Cardiovascular Diseases, Karachi

By

M. REHMAN, KALIMUDDIN AZIZ, A. SAMAD, AZHAR FARUQUI, A. HAQUE, FAIZ-UR-RAHMAN AND S.A. SYED.

Because of the unavailability of diagnostic and surgical expertise Congenital Heart Disease (C.H.D.) in Pakistan has essentially been left untreated until recently. One is likely not only to encounter patients in whom altered hemodynamics have caused irreversible damage to the Cardiopulmonary system but also patient in whom other body organs are affected resulting in varying degrees of functional incapacity. Furthermore, one is expected to find survivors with congenital heart disease in the adult population. The natural history studies show that the incidence of congenital cardiac malformalions is 8 per thousand live births[1] and approximately 50% of these die in the first years of life if no surgical treatment is offered[2]. The mortality continues steadily till adulthood. Specifically, for instance, most patients with transposition of the great arteries would die if no medical or surgical palliation is available.

The availability of Cardiopulmonary bypass technique has now made it possible to perform intra-cardiac corrective procedures for most cardiac malformations and has radically altered the natural history of survival of children born with congenital cardiac malformations[3].

Intra-cardiac repair using the technique of Cardiopulmonary Bypass has now become posisible at the NICVD.

The diagnostic work up of a child or an infant with C.H.D. is also possible at the National Institute of Cardiovascular Diseases, Karachi. This includes clinical, electrocardiographic, M-mode and two dimentional echocardiographic evaluation and hemodynamic and angiocardiographic work up. The purpose of this report is to review our crecent experience with surgical correction of Congenital Cardiac malformations during the period between October 1980 to November 1981.

Material and Methods:

The study includes patients operated during the period October, 1980 to November, 1981. The diagnosis was made on clinical, non-invasive echocardiographic and Cardiac Catheterization studies. Haemodynamic and angiographic studies were obtained in all patient undergoing Cardiopulmonary Bypass Surgery. Patients with patent ductus arterious were not studied by Cardiac Catheterization unless the diagnosis was in doubt or signs of pulmonary artery hypertension were present. Intra-Cardiac surgical repoair was performed with the use of Cardio pulmonary Bypass technique. Moderate hypothermia and cardioplegia was employed for myocardial preservation during the period of cardiac stand still.

The mortality was defined as intra-operative when the patient died in the operating room, early if death occurred during the period of hospitalization and late, when death occurred 3 months post-operatively following discharge from the hospital.

Results:

A total of 507 patients were operated. One hundred and fifty eight of these had congenital cardiac malformations. The age ranged between 0.3-60 years. One hundred and three patients were less than 12 years of age at the time of operation and fifty five were (Taole I) greater than 12 years. Eighty four patients underwent close heart operations. Fifty eight of 84 patients undergoing close surgical procedures were less than 12 years age and twenty six were greater than 12 years. Mortality was somewhat greater for less than 12 year group (Table II)

Table I: Age Distribution in C.H.D.

	<12 Years	*>12 Years*	*Total*
A.S.D.	11	17	28
T.O.F.	42	8	50
V.S.D.	5	11	16
P.D.A.	39	13	52
C.O.A.O.	2	6	8
Misc.	4	0	4
	103(66%)	55(34%)	158

Abbreviation

A.S.D. =Atrial Septal Defect
T.O.F. =Tetralogy of Fallot
V.S.D. =Ventricular Septal Defect
P.D.A. =Patent ductus arteriosus
C.O.A.O. =Coarctation of Aorta
Misc =Miscellaneus

Table II: Closed Heart Surgical Mortality Related to Age.

	<12 Years			*>12 Years*		
	No.	*Age (Years ±SD)*	*Mortality*	*No.*	*Age (Years ±SD)*	*Mortality*
P.D.A.	39	5±3	0 (0%)	13	22±7	0 (0%)
Shunts	13	4±3	4 (31%)	7	16±4	1 (14%)
COAO	2	10±3	0 (0%)	6	26±17	0 (0%)
P.A. Band	4	5±2	0 (0%)	0	±	0 (0%)
	58		6.0%	26		3.8%

(6 percent) compared to patients more than 12 years age (3.8 percent). Ligation of the patent ductus arteriosus was performed on 52 patients with mean age of 14±5 years (range 0.3-33 years). There was no mortality in this group. Aorto pulmonary shunt surgery was performed on 20 patients mean age 8±7 years. (Range 0.2-19 years). Sixteen of these had Tetrology of Fallot three had Transposition of the Great Arteries with ventricular septal defect and left ventricular outflow stenosis and one patient had double outlet right ventricle, V.S.D. and pulmonary stenosis (Table III). Blalock—Taussig

Table III: Anatomic Lesions in Patients Undergoing Shunt Surgery

Lesion	*No.*
TOF.	16
T.G.A. with V.S.D. and L.V.O.S.	3
D.O.R.V. with P.S.	1
Total	20

Abbreviations

T.O.F. =Tetralogy of Fallot
T.G.A. =Transposition of The Great Arteries
V.S.D. =Ventricular Septal Defect
L.V.O.S. =Left Ventricular Outflow Stenosis
P.S. =Pulmonary Stenosis
D.O.R.V. =Double Outlet Right Ventricle

shunt was performed in 7 patients, 2 died due to non-functioning shunt, Waterston Cooley shunt was performed in 8 with two deaths due to non-functioning shunt. Gortex graft was inserted between the left subclavian and left pulmonary artery in five with one death due to Graft thrombosis (Table IV). Repair of coarctation of aorta was performed in eight patients mean age 22±16 with no death. Pulmonary arterial banding was performed in four patients mean age 4.5±1.7 years (range 3.0-6.0 year). All four survived. The over all mortality for the close heart surgery was six percent (5/84), all of these deaths occurred in the shunt group (Table V).

Table IV: Types of Aorto-Pulmonary Shunts

	Total No.	*Died No.*	*Alive No.*
Balalock Taussig Shunt	7	2	5
Waterston Cooley Shunt	8	2	6
Gortex Graft	5	1	4
Total	20	5	15

Table V: Closed Heart Surgery C.H.D. (September 1980-November 1981)

Lesion	*Age (Yrs.±ISD)*	*No.*	*Mortality*	*SEX Male*	*Female*
P.D.A.	14±5	52	0 (0%)	19	33
Shunts	8±7	20	5 (25%)	15	5
COAO	22±16	8	0 (0%)	7	1
PAB	5±2	4	0 (0%)	2	2
Total		84	5 (6%)	43	41

Intra Cardiac repair of defects was performed using the technique of Cardio Pulmonary Bypass in 74 patients. Twenty Eight had atrial septal defect, mean age 16.2±8.1 (range 12.5-32 years). Twenty seven patients had atrial septal defect of secundum variety and one had primum atrial septal defect. Also one of 27 had repair of associated anomolous right pulmonary veins. There was no death in this group. Thirty patients, mean age 11±6 years, underwent total correction of T.O.F. Closure of ventricular septal defect was done using a Dacron patch, resection of infundibular stenosis and transannular pericardial patch enlargement of the outflow tract was done in all patients. 10 patients died (Mortality 30%). One patient died intra-operatively, due to low cardiac output syndrome, 9 patients died during early post-operative period (Table VI). The cause of death was respiratory insufficiency in three, post operative fluid overload in one,

Table VI: Total Correction of C. H. D. (September, 1980 - November, 1981)

Lesion	*Age (Yrs.±SD)*	*No.*	*Mortality*	*SEX Male*	*Female*
A.S.D.	16.2±8.1	28	0 (0%)	8	20
T.O.F.	11.0±6	30	10 (30%)	13	17
V.S.D.	14.0±7	16	4 (25%)	6	10
		74	14	27	47

low Cardiac output syndrome in 4, bleeding in one and sepsis in one. The mortality was significantly higher for the patients who were less than 12 years of age at time of repair 9/41 (41%) compare to those greater than 12 years 1/8 (12.5%) Table VII. Another significant factor contributing to higher mortality was the level of hemoglobin and hematocrit. The mortality was significantly greater ($P < 0.005$) for patients with Hb 19.9±2.8 Gm and PCV—59±10% compared to Hb 14.4±1.9 Gm and

Table VII: Surgical Mortality Related to Age (CPB)

	< 12 YRS.			> 12 YRS.		
		Mean ± SD			Mean ± SD	
Lesion	No.		Mortality	No.		Mortality
A.S.D.	11	8.4±2.7	0	17	21.1±6.3	0
V.S.D.	5	1.0±2.1	2(40%)	11	19.8±6.0	2(18%)
T.O.F.	22	8.5±2.4	9(41%)	8	19.6±3.6	1(14%)
	38		10(28%)	36		3(8%)

PCV 41±5% Fig. 1. Intra-cardiac repair of the ventricular septal defect was performed in sixteen patients, mean age 14±7 years. Three patients had associated aortic valve insufficiency, 2 had associated infundibular stenosis of the right ventricle and one had mitral regurgitation. Ten patients had only ventricular septal defect, one of whom had moderate degree of obstructive pulmonary vascular disease. There were four early deaths. Table VIII.

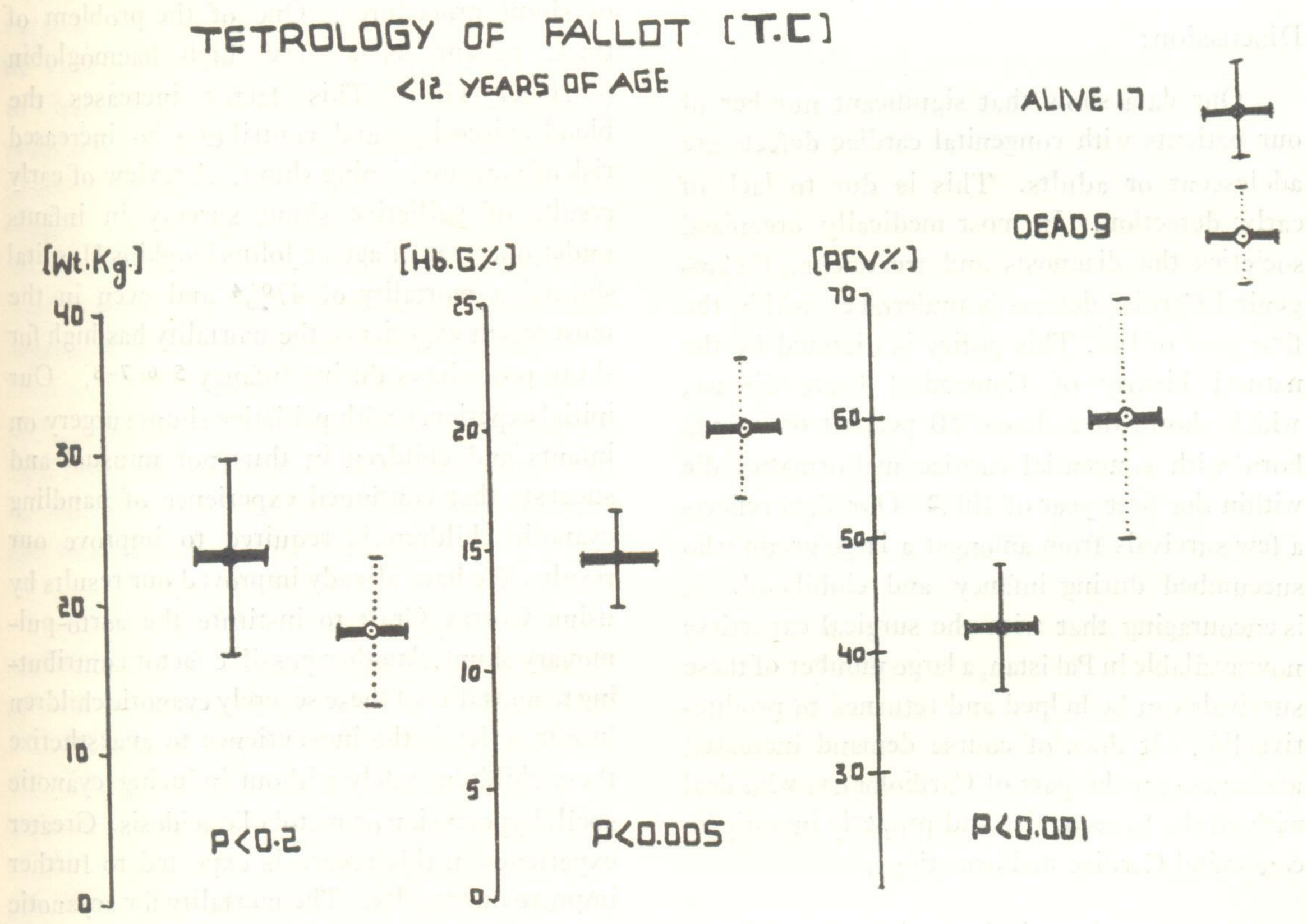

Fig. 1.

The effect of increased hemoglobin (Hb) and PCV is shown in Twenty Six patients with Tetralogy of Fallot who had total correction (TC). Note that patients with hemoglobin of 19G% and PCV of 59% had significantly greater mortality compared to those with lower Hb and PCV values.

Table VIII: Data on V.S.D. Patients Undergoing Primary Closure

	No.	*Dead*	*Alive*
V.S.D. Simple*	10	2	8
V.S.D. with A.I.	3	0	3
V.S.D. with P.S.	2	1	1
V S D with M I	1	1	0
	16	4	12

*One patient had pulmonary artery hypertension due to obstructive plumonary vascular disease.

Discussion:

Our data show that significant number of our patients with congenital cardiac defects are adolescent or adults. This is due to lack of early detection. In most medically organized societies the diagnosis and treatment of Congenital Cardiac defects is undertaken within the first year of life. This policy is dictated by the natural history of Congenital heart disease, which shows that almost 50 percent of infants born with congential cardiac malformaties die within the first year of life.[2] Our data reflects a few survivals from amongst a large group who succumbed during infancy and childhood. It is encouraging that with the surgical expertiese now available in Pakistan, a large number of these survivals can be helped and returned to productive life. It does of course demand increased awareness on the part of Cardiologists who deal with adults to recognise and properly investigate congenital Cardiac malformations.

Our present surgical experience is not large, however it does provide a frame work for analysis of various disciplines concerned with the surgical management. The surgical mortality in our series was significantly greater for patients less than 12 years age compared to patients greater than 12 years. This of course does not mean that we should delay correction of Cardiac defects because many of the infants and children would die and not attain adolescence. The data empasizes that greater expertise is needed for successful surgical correction in the younger patient.

The surgical mortality for close heart operations for acyanotic lesions was zero. All of the deaths for close heart procedures occured in cyanotic children undergoing various types of shunt procedures. One of the problem of these patients is a very high haemoglobin ($>$20 G%Hb). This factor increases the blood viscosity and contributes to increased risk of non functioning shunt. A review of early results of palliative shunt surgery in infants under one year of age at Johns Hopkins Hospital showed a mortality of 47%[4] and even in the most recent experience the mortality has high for shunt procedures during infancy [5-6-7-8]. Our initial experience with palliative shunt surgery on infants and children in thus not unusual and suggests that continued experience of handling cyanotic children is required to improve our results. We have already improved our results by using Gortex Graft to institute the aorto-pulmonary shunt. Another possible factor contributing to mortality of these severely cyanotic children in our series is the inexperience to anaesthetize these children safely without inducing cyanotic spell, hypotension or metobolic acidosis. Greater experience in this regard is expected to further improve our results. The mortality for acyanotic patients undergoing Intra-Cardiac repair for Atrial Septal Defect was nil. There were four deaths in patients in whom parimary closure of the ventricular septal defect was undertaken 25% mortality. One of the deaths was in a 7

years old patient with a large muscular VSD and high pulmonary arterial pressure due to increased pulmonary vascular resistence due to pulmonary vascular obstruction (P.V.O.). It has been shown that operative mortality is proportionately increased with increasing degree of P.V.O. in patients with hypertensive ventricular septal defect.[9] The 3 deaths remaining were not related to the associated lesions but can ony be attributed to post operative management difficulties. As with other intra-cardiac reparative surgery, the earlier mortality of surgical closure of V.S.D. was higher (10%) in Toronto series between 1959-1961 compared to later mortality whioh was less than 3 percent[10]. Our present results show no death amongst six children with V.S.D. less than 6 years age. The mortality, of total correction of patients with TOF was 38%. The present day data from Western Sources suggest a mortality of less than 5% in older children[11]. However, review of early experience with total correction in Toronto series during 1955-1968 period showed a mortality of 35% in the first half and was reduced to 12% in the second half of this period[11].

Amongst the factors contributing to increased mortality in our series is inexperience with pre-operative anaesthesia management of cyanotic children, with markedly elevated haemoglobin (>17G). Our present policy is to perform pre-operative exchange of the whole blood of the patient with 5% salt free. Albumin till the PCV is <60 percent[12]. Hemodilution technique on the Cardio pulmonary Bypass are now a routine for the cyanotic patient. Post operative management has a significant effect on the survival rate of these children. Our data shows that mortality for children with TOF less than 12 years age was mach greater than those greater than 12 years age. This is in part due to greater efficiency required to manage cyanotic children during the post-operative period.

The post-operative management can be broadly divided into a management of cardiovascular and ventilatory status. The Cardiovascular system monitoring includes maintenance of sinus rythm, treatment of post-operative arrhythmias, maintenance of blood volume and cardiac output by monitoring the right and left atrial pressure and systemic pressure. Younger the patient greater is the accuracy required for a successful outcome. A review of these factors suggests that greater expertise and awareness would be achieved with the passage of time. Large number of factors such as sepsis, fluid overload, respiratory insufficiency contributing to mortality in our series, are avoidable by adherence to a disciplined and systematic post-operative management. These factors can be largely removed. The improvement in the operative results can only be obtained by systematic organisation of all the disciplines required for the management of these children. We have begun to train nurses in the intensive care management. Ventilatory management requires constant vigilence, absolute aseptic techniques of patient handling, accurate monitoring of blood Gases and above all psychologic awareness of minute to minute care these children need. Modern anaesthesia has made great contribution to the improved surgical results.

Intra-Cardiac repairs of Congenital defects in the newborn and infants is not yet done at our Institute. Even in Western Countries only a few centres are aquipped to deal with this group

of patients[13]. We however perform shunt surgery for Cyanotic infants who have pulmonary stenosis and reduced Pulmonary blood flow. Our present experience gives us a frame work to develop increased expertise and high-lights areas needing improvement.

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