Surgical Management of Heart Valve Disease in Children*

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SUMMARY

From 1-1-1984 to 31-12-1988, 1022 patients had heart valve operations at our institute. 17% (170) out of these were 15 years or younger in age. 59 patients had mitral stenosis (MS), 50 had mitral regurgitation (MR), 22 had both mitral stenosis and regurgitation (MS, MR), 22 had mitral & aortic regurgitation (MR, AR) and 17 had aortic valve stenosis or regurgitation (AS or AR). 47 patients had closed mitral valvotomy (CMV), 35 and 15 & 19 had mitral, aortic and double valve replacement (MVR, AVR, DVR) respectively. 42 patients underwent mitral valve repair (MV repair) procedure. Open Mitral Valvotomy (OMV), double valve repair (DV Repair). Aortic valve repair and aortic valvotomy was carried out in another 12 patients. Overall operative mortality (30 days) was 6.4%. Assessment of disability after 3 months showed 80% of the patients were either asymptomatic, or in NYHA class I & II. 65% of the patients had a valve conserving operations (Group A) and 35% had the valve replaced (Group B). After an average follow up of 39 months, Group A patients (n 111) had 3 operative deaths, no minor or major embolic episodes, no endocarditis, no permanent anticoagulants, no bleeding and 3 re-operations. 10 had mild to moderate residual regurgitation, 7 were lost to follow up and 1 late mortality. Group B patients (n 57) had 8 operative deaths, 4 minor and one major embolic episodic, 2 endocarditis. 49 had permanent anticoagulations with 5 patients having bleeding, 2 peraprosthetic leaks, 3 late deaths and only 3 were lost to follow up. We conclude that 17% of our valve patients are 15 years or younger in age. The best valve conserving operation in appropriate patients in CMV. Majority of the valves, particularly mitrals, can be preserved by either CMV or open repairative procedures. Both early and late mortality, morbidity and economics are very favourable in those who have their valves conserved at surgery. It is strongly felt that in Developing Countries learning, practicing and improving valve conserving techniques for young patients should be a major consideration for cardiac surgeons.

INTRODUCTION

Surgery for heart valve disease in children has peculiarities, that are different from similar patients in older age group. This problem is compounded in developing countries like Pakistan with a population of 98 millions and per capita income of 394 US $ per annum and an incidence of admission for rheumatic fever and its effects 1.5 — 2.5 per thousand population[1]. Due to endemic nature of rheumatic fever, quite a sizable population of youngsters suffer from this disease. Lack of public education, scarcity of easily and timely available, suitable medical advice and socioeconomic factors add to the difficulties. This presentation deals with the analysis of the cardiac surgery performed for heart valve disease for patients of 15 years or below at Armed

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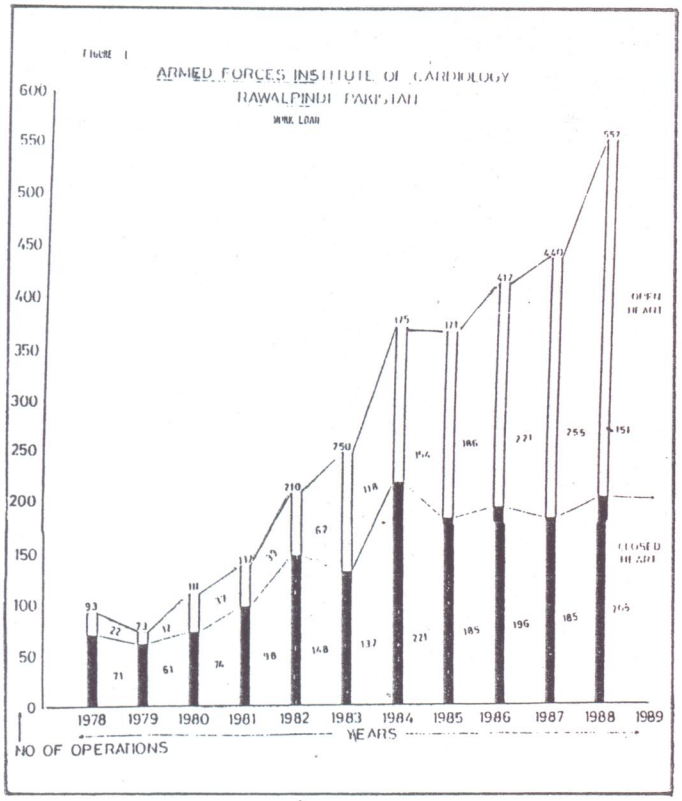


Forces Institute of Cardiology Rawalpindi, Pakistan which was established in 1978 (Figure —1).

PATIENT POPULATION

In 5 years, from Jan. 1984 to Dec. 1988 a total number of 1022 patients underwent operations for valvular heart disease at our institute. 170 (17%) of these were 15 years or younger in age. There were 83 males and 87 females, with an average age of 11 years (standard deviation ±2.6).

Palpitation was the commonest presenting symptom found in 120 (70%) patients. 104 (61%) complained of effort dyspnoea. 81 (48%) had one or more episodes of frank congestive cardiac failure. 28 (16%) complained of fleeting joint pains. 110 (65%) gave history suggestive of rheumatic fever in the past. 39 (23%) had established atrial fibrillation. 3 (2%) had cerebral embolic episodes. 31 (18%) had disability NYHA class II, 112 (66%) class III and 27 (16%) class IV.

Table - 1 shows the breakdown of the lesions. Commonest being isolated mitral stenosis and

TABLE 1	LESIONS		(n 170)
MS	59	34%	
MR	50	29%	MITRAL
MSMR	22	12%	153 (90%)
MR AR (MS)	22	12%	
AR (AS)	16	9%	
AS	1	0.5%	AORTIC
TR	11	7%	39 (23%)

Legend – MS: Mitral Stenosis, MR: Mitral Regurgitation, AR: Aortic Regurgitation, AS: Aortic Stenosis, TR: Tricupid Regurgitation.

rarest, isolated aortic stenosis. About 97% of these had direct, indirect or circumstancial evidence of being rheumatic in nature. Mitral valve was involved in 90% and aortic valve in 23% of the patients. 7% had tricuspid valve involvement (non organic). 22 patients (12%) had mild pulmonary hypertension (upto 50% of systemic pressure). 43 (25%) had moderate (51 to 75% of systemic pressure). 42 (22%) had severe pulmonary hypertension (more than 76% of systemic pressure). All these patients had involvement of the mitral valve. 12 patients (7%) belonged to well-to-do socio-economic class, 87 (51%) to the middle and 71 (42%) belonged to poor class.

OPERATIONS AND RESULTS

Table - 2 shows the breakdown of the operations performed. Closed mitral valvotomy being the commonest procedure. Valve repair procedures, mostly done for mitral valve comprised of mitral annuloplasty, releasing of the fused commissures, separation of the fused papillary muscles and shortening of cordae in appropriate patients, keeping in mind the principles described by Kay[2], Wooler[3], Duran[4], Carpentier[5], and Reed[6]. In the last 30 patients, Reed's measured mitral annuloplasty technique[7] was used with satisfactory results. We were able to restore a satisfactory competence of the aortic valve in six patients, three in conjunction with the mitral valve. In two patients the aortic cusps were extended to establish satisfactory co-optation,

TABLE 2 – OPERATIONS AND RESULTS n 170

CMV	47		MVR	35	
OMV	5		AVR	15	
MVRR	42		DVR	19	
DVRR	3				
AVRR	3				
OAV	1				
GP. A	101	(60%)	GP. B	69	(40%)
MOR	3	(3%)	MOR	8	(11%)

OVERALL MOR 11/170 6%

Legend – CMV: Closed Mitral Valvotomy, MVR: Mitral Valve Replacement, OMV: Open Mitral Valvotomy, AVR: Aortic Valve Replacement, MVRR: Mitral Valve Repair, DVR: Double Valve Replacement DVRR: Double Valve Repair, AVRR: Aortic Valve Repair, OAV: Open Aortic Valvotomy, GP.A: Group A, GP. B: Group B, MOR: Mortality.

using preserved bovine pericardium. In four patients, plication of one or more redundant aortic cusps at the commissure yielded good results. 11 patients had annuloplasty of tricuspid valve in addition to other procedures. Our youngest patient, aged 5 years had a satisfactory open aortic valvotomy. In 101 patients (60%) we were able to preserve the native valve (Group A), operative mortality[8] for this Group was 3%.

In 69 patients (40%) the valve/valves had to be replaced (Group B) either primarily for severe distortion due to rigidity, thickening or calcification etc. or when the attempts to repair did not bear satisfactory results. 35 patients had mitral, 15 had aortic and 19 had both aortic and mitral valve replaced. Group B, the valve replacement group, had an operative mortality of 11%. Operative mortality for the whole group (n 170) was 6%. Difference of operative mortality between Group A and B was significant ($P < 0.05$).

The follow up and late results are shown in table 3 and 4. We have a mean follow up of 43 months ranging from 4 to 66 months after operation. With information available for 94% of the patients (9 patients being lost to follow up). There are 87.6% known survivors (including operative mortality, late mortality and loss to follow up).

TABLE 3 FOLLOW UP 1

RANGE MONTHS		4	to	66
AVERAGE PER PATIENT				43
INFORMATION AVAILABLE FOR				94%
LOST TO FOLLOW UP		9		(5%)
KNOWN SURVIVORS		146		(86%)
NYHA	I	62		(42%)
NYHA	II	49		(34%)
NYHA	III	28		(19%)
NYHA	IV	7		(5%)

In Group A there is one late death, due to congestive failure after a year of operation. There were residual leaks in 12 patients, 3 severe enough to have re-operations (valve replacement). In Group B there were 3 late deaths, one 10 months post-op due to cerebral embolism in a patient who has double valve replacement, and two due to endocarditis, 6 months post double valve replacement and 15 months post mitral valve replacement. 4 patients had major embolic episodes (one fatal), 5 patients had significant anti-coagulant bleeding necessitating hospital admission, two had fatal endocarditis and two had to be re-operated (one for residual leak and haemolysis) and one for endocarditis.

CONCLUSIONS

In developing countries like Pakistan there is quite a sizable child population needing heart valve surgery, valve conserving operations are possible in majority of these patients with satisfactory short-term results, and are very cost effective. Valve replacement surgery has a higher mortality and complication rate. Mitral and double valve replacement are bad news. Prosthetic valve endocarditis, embolisation and anticoagulant related bleeding pose major threat t the children after valve replacement.

TABLE 4 RESULTS OF OPERATIONS AND FOLLOW UP II FOR 170 PATIENTS

	CMV	OMV	MVRR	DVRR	AVRR	OAV	GP.A	MVR	AVR	DVR	GP.B
NO.	47	5	42	3	3	1	101%	35	15	19	69
OPERATIVE DEATHS	0	0	2	1	0	0	3%*	3	1	4	11%
LATE DEATHS	0	0	0	1	0	0	1%	2	0	1	4%
LOST OF FOLLOW UP	4	0	3	0	0	0	7%	2	0	0	3%
RESIDUAL LEAKS	4	1	6	1	0	0	12%	1	0	0	0.5%
EMBOLISM											
MINOR	0	0	0	0	0	0	0	2	0	2	6%
MAJOR	0	0	0	0	0	0	0	0	0	1	0.5%
ANTICOAGULANT RELATED BLEEDING	0	0	0	0	0	0	0	2	1	2	7%
ENDOCARDITIS	0	0	0	0	0	0	0	1	0	1	3%
HAEMOLYSIS	0	0	1	0	0	0	1%	0	0	1	0.5%
REOPERATIONS	1	0	2	0	0	0	3%	1	0	1	3%

Legend — CMV: Closed Mitral Valvotomy, OMV Open Mitral Valvotomy, MVRR: Mitral Valve Repair, AVRR: Aotric Valve Repair, OAV: Open Aortic Valvotomy, GP.A: Group A (Valve Conserving Operations) MVR: Mitral Valve Replacement AVR: Aortic Valve Replacement, DVR: Double Valve Replacement, GP.B: Group B (Valve Replacement Group) *Difference from GP. B Significant ($P < 0.05$).

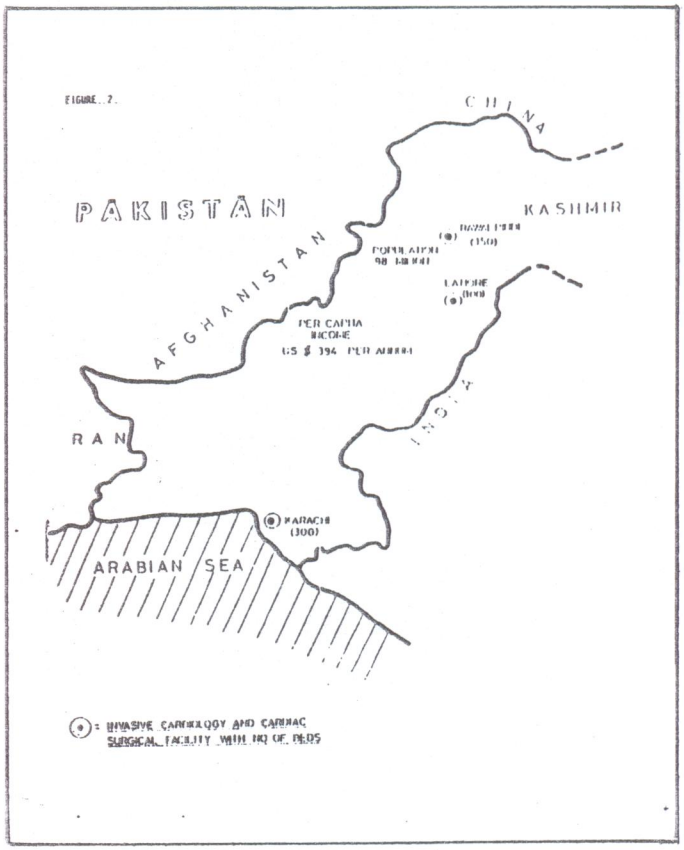
DISCUSSION

Pakistan is a developing country with a very low per capita income (Fig. 2). 70% of the population lives in rural areas, where basic social, educational and health facilities are limited. Only 25% of the people are educated. 40% of the population consists of children below 15 years. Cardiac surgery is a relatively a recent development. There are under 600 beds available for modern invasive cardiology and cardiac surgery. Facilities for proper management of complicated cardiac ailments in neonatal and infantile population are almost non-existent. Patients with severe congenital heart valve disease, usually perish in early infancy and childhood. Patients who come to surgery are usually older children and mostly are of rheumatic nature. Surgery for heart valve disease in young age group in aforementioned setting poses many peculiar problems, which are different from those in the adults and those in the developed world. The expected somatic growth[9], use of anticoagulation in youngsters and that too where wide spread knowledge and facilities of its precise control is not easily available, problem of availability of medi-

TABLE 5 VALVE USED AND PRICE (n 170)

	No. of Patients	Price Each
STARR EDWARDS	39	Rs. 11000
ST. JUDES	9	42000
DUROMEDICS	7	32000
CARBOMEDICS	5	26000
MEDTRONIC	5	20000
CARPENTIER EDWARDS	4	16000

(ONE £ EQUALS Rs. 33)



cines and laboratory facilities, non-compliance of the children to medical regimens, problems of child bearing in females[10], all bear importantly on the outcome of this type of surgery. Initially introduction of tissue valves substitutes seemed promising but sooner we learned about excellerated tissue failure and calcification in children who had these valves[11,12]. Earlier hopefulness of the use of mechanical prostheses in children with less aggressive approach to the use of anti-coagulants[13,14,15]. Specially after St. Judes valve[16] is being dispelled by recent reports of significant thromboembolic episodes[17]. In comparison to the valve replacement, good results of mitral valve repair procedures are being realised[18].

The above and many other factors, and excellent work done by the pioneers in the field of valve repair procedures[2,3,7,19,20,21] lead us to persue a valve conserving policy. We now believe that a slightly damaged but reasonably functional genuine God made valve, may not be perfect but is undoubtedly better than the man made or man assembled heart valve substitute. Most of our patients come to operation quite late, after having had congestive failure, pulmonary hypertension, atrial fibrillation and huge cardiomegaly. The young age and rheumatic disease provides us with a variety of different lesions posing a technical challenge to the surgeon. One should take this on and we are sure one would be able to preserve many a rheumatic valve and thus make the child's and his family's life more comfortable and safe as compared to the life with an artificial valve. It is essential to have a clear conception of functional anatomy and pathology of the valve and knowledge of various techniques available for valve preserving surgery and use appropriate methods suitable for individual patients. Perseverance, patience, skill and consideration for safety are the greatest assets in these situations. Costs and logestics for procurement and availability of equipment used for heart valve surgery also bares importantly for extending this facility to the population. On average the disposable equipment used for one open heart procedure, excluding the price of the valve, costs about Rupees thirty-two thousand (one thousand UK £). Table - 5 shows the comparative cost of various makes of the valves used in this series. This does effect the selection of the valve substitute by the Surgeon.

It is strongly felt that in developing countries learning, practicing and improving valve conserving techniques for young patients should be a major consideration for cardiac surgeons.

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