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CERTIFICATE

I, Dr. PRAMOD KUMAR JAISWAL .....hereby declare that I have actually performed all the procedures listed / carried out the project under report.

Signature.....

Place : TRIVANDRUM

Name in PRAMOD KUMAR JAISWAL.....

Date : 18-11-92 capital letters

Forwarded. He has carried out the minimum requirement of procedures / etc.

Signature

Head of the department

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LIST OF PROCEDURES DONE
PROJECT REPORT

TITLE OF THE PROJECT: **CLINICAL PROFILE AND NATURAL HISTORY OF ENDOMYOCARDIAL FIBROSIS**

NAME....PRAMOD KUMAR JAISWAL.....

PROGRAMME :..D.M. CARDIOLOGY.....

MONTH & YEAR
OF SUBMISSION :....NOVEMBER, 1992.....

Note:— (i) In the case compilation of procedures done, the contents and the subsequent pages should be made into different sections (a) Procedures done (b) Procedures assisted (c) Procedures participated (d) Procedures attended / participated etc. in Other Centres. Each section should be preceded by a leaf carrying the name of the section that is succeeding.

(ii) The Contents page will carry information as per model given under

PROCEDURES DONE

Closed Mitral valvotomy ---------- 124 (say)

Patent ductus arteriosus-ligation ---------- 10

Atrial septal defects ---------- 20

PROCEDURES ASSISTED

Closed Mitral valvotomy ---------- 100 (say)

(iii) In the subsequent pages details of each procedure done/assisted should be given in the format given below :—

Heading : Closed mitral valvotomy

Date	Name of the patient	Age	Sex	Patient No.

(iv) In the case of Project Report in the page immediately following the Certificate page the under-mentioned details should be given :—

(a) Title

(b) Duration

(c) Aim and scope

(d) 50 word summary of work done

ABSTRACT

The clinical course and natural history of 206 patients of endomyocardial fibrosis who were followed up between November 1975 and October 1991 were studied. The diagnosis was established by angiography, echocardiography or necropsy. Survival at the end of one year, five years and ten years were 82.5%, 53.5% and 37.3% respectively. Significant determinants of mortality were studied by univariate analysis and log rank test of Kaplan-Meier estimates and Cox proportional hazards multiple regression analysis. The factors influencing mortality by univariate analysis were younger age, lower hemoglobin level, rightward QRS axis in ECG more than $+90^{\circ}$, cardiomegaly in chest X-ray, lower systolic blood pressure, NYHA functional class 3 and 4, presence of left ventricular failure, presence of embolic episodes, right atrial mean pressure more than 20 mm Hg, right ventricular enddiastolic pressure more than 20 mm Hg, pulmonary artery mean pressure more than 40 mm Hg, arterial mean pressure less than 70 mm Hg, arterial oxygen saturation less than 85% and pulmonary artery oxygen saturation less than 70%. Multivariate analysis predictors of survival were NYHA functional class, rightward QRS axis in ECG, embolic episodes, cyanosis, severity of tricuspid regurgitation, elevated right atrial mean pressure, low aortic mean pressure and low arterial oxygen saturation. Analysis of larger number of patients over

longer period of follow up has clearly shown that survival was 53.5% at the end of five years and the prognosis was not as poor as originally reported from the African continent.

INTRODUCTION

Endomyocardial fibrosis is a restrictive cardiomyopathy of tropics. Ever since its recognition as a clinical entity by Bedford and Konstam[1] in 1946 followed by a detailed description of its clinical features by Davies[2] in 1948, the clinical and laboratory features of this condition have been studied fairly extensively by various workers. Yet, there is paucity of information regarding its clinical course and natural history[3,4]. We reported earlier the clinical course of endomyocardial fibrosis in the south western state of Kerala in India where the disease is prevalent[5]. In this study, we report our analysis of data on more number of patients followed for longer duration.

MATERIAL AND METHODS

295 patients of endomyocardial fibrosis were registered in our Institute during sixteen year period from November 1975 to October 1991. Eightynine patients were subjected to surgical intervention and were excluded from this study. Remaining 206 patients formed the material for this study. The diagnosis was confirmed by angiography in 179 patients,

by echocardiogram alone in 20 patients and by autopsy alone in 7 patients. There were 99 males and 107 females. Their age ranged from 2 years to 65 years, the mean age was 25.3 ± 13.5 years. Fiftyone had right ventricular endomyocardial fibrosis, 14 had left ventricular disease while 141 had biventricular disease.

After a detailed work up, all pertinent clinical and laboratory features were noted and these patients were followed up every six months by prior appointment. Patients who did not come for regular follow up were sent at least three reminder letters. Information regarding the death of patients at home was sought from close relatives whenever possible. Maximum followup was upto 136 months, the mean follow up being 35.5 ± 31.9 months. Seventy-seven patients were lost to follow up during varying time periods after one year of initial presentation.

STATISTICAL ANALYSIS

Twentyone patient variables for survivors and those who died were compared by 't' test for quantitative data and chi square test for qualitative data. The variables which were statistically significant were chosen for log rank test[6]. Eight significant variables were analysed for their impact on survival by the log rank test. Cox proportional hazards multiple regression analysis[7] was also applied to the variables

studied by the log rank test. We used a BIOSTAT program.

RESULTS

Clinical features - Salient clinical features of 206 patients at first presentation are shown in Table I. Duration of symptoms was less than 24 months in 46.1%, and 39.3% of patients were in NYHA functional class 3 and 4. Twentyeight percent of patients were below 15 years of age. Features of right heart failure were present in 71.8%, while those of left heart failure were seen in only 25.7%. Ascitis and edema of feet were present in about 25% of the patients. Central cyanosis was noted in 15.5% while associated clubbing was present in 9.7%. Despite the presence of atrioventricular valve regurgitation, one fourth of the patients did not have any murmur.

Laboratory features (Table 2) Significant eosinophilia was a feature in only 8.7% of patients. Cardiomegaly in X-ray of the chest was present in 64%. Atrial fibrillation was the rhythm in one third of patients while QRS axis of more than $+90^{\circ}$ was seen in 29.1%. Hemodynamic data are given in Table 3. More than half had moderate to severe elevation of right heart filling pressure. Over twentyeight percent of patients had moderate to severe elevation of pulmonary artery mean pressure while half had moderate to severe elevation of left heart filling pressure.

Seventyfour patients died during follow up period, the cause of death being shown in Table 4. Univariate analysis of qualitative data showed that factors which influenced survival were, NYHA functional class 2 or 1, presence of right ventricular disease more than left ventricular disease, absence of left heart failure, absence of embolic episodes, and normal intensity of pulmonary second sound.

Univariate analysis of quantitative data showed that mortality was determined by younger age, lower hemoglobin level, more rightward QRS axis, larger cardiothoracic ratio in X-ray chest, lower systolic blood pressure, higher right atrial and right ventricular enddiastolic pressures, higher pulmonary arterial mean pressure, lower aortic mean pressure, lower arterial oxygen saturation, and lower pulmonary artery oxygen saturation.

By log rank test of Kaplan-Meier estimates (Table 5) determinants of mortality were mitral regurgitation more than moderate, right atrial mean and right ventricular enddiastolic pressures over 20 mm Hg, pulmonary artery mean pressure exceeding 40 mm Hg, left ventricular enddiastolic pressure more than 15 mm Hg, aortic mean pressure less than 70 mm Hg, arterial oxygen saturation less than 85% and pulmonary artery oxygen saturation less than 70%.

Kaplan-Meier survivor curve for the 206 patients (fig I) showed 82.5% survival at one year, 74.3% at two years and 37.3% survivors at ten years after first presentation. Multivariate analysis by Cox regression method showed the factors which influenced the survival were NYHA functional class, rightward QRS axis in ECG, embolic episodes, cyanosis, right ventricular third heart sound, cardiac rhythm, severity of tricuspid regurgitation, right atrial mean pressure, aortic mean pressure and arterial oxygen saturation (Table 6).

DISCUSSION

The present study is one of the few with such a large number of patients with endomyocardial fibrosis followed up for a fairly long period after their first presentation to the hospital. Previous reports on the natural history of endomyocardial fibrosis consisted of smaller number of patients[3]. D'Arbela et al reported a 43.5% mortality at the end of one year and death in the majority of the remaining patients in the next 3 to 4 years[4]. The present study does not indicate such a poor prognosis, 82.5% survival at one year and 53.5% at five years. This even differs from our previous report on 145 patients in the survival trend[5] 53.5% vs 35.6% at five years and 37.3% vs 26.4% at ten years. Thus, larger number of patients followed up for a longer period gives a more accurate picture about the natural history. Moreover, patients with

minimal symptoms could also be identified by high resolution two dimensional echocardiography and such patients have good prognosis while those with very obvious physical findings are likely to have far too advanced disease which will naturally have a poorer prognosis.

Shorter duration of symptoms at first presentation apparently reflects a more severe form of the disease and probably its acute phase[3]. This is probably the reason for the high mortality in the first year as reported by D'Arbela et al[4]. However, this study differs from the other two studies as well as our previous report because, despite 46.1% patients having had shorter duration of symptoms, univariate analysis failed to show this to be a significant determinant of mortality.

Cyanosis is a less common feature in severe diastolic dysfunction of right ventricle with resultant high right atrial pressure causing stretching of inter atrial septum and right to left shunt across patent foramen ovale. This indirectly implies the severity of right ventricular disease and is one of the determinants of survival in multivariate analysis.

Cardiomegaly in chest x-ray is the most striking feature of right ventricular involvement and is mainly contributed by a large right atrium and to some extent by pericardial effusion. This again implies severe disease of the right ventricle and

is a predictor of mortality in univariate analysis.

Rightward axis of QRS in frontal plane of ECG can occur as a result of right ventricular outflow dilatation which is a feature of severe right ventricular involvement or it could be due to right ventricular hypertrophy secondary to severe pulmonary arterial hypertension due to left ventricular endomyocardial fibrosis. Hence, it influences survival in both univariate and multivariate analysis.

Right heart failure was present at first presentation in 71.8% of patients while left heart failure was a feature in only 25.7%, yet left heart failure significantly influenced survival thereby implying that right heart failure is tolerated better for longer period. However, more severe right ventricular involvement as evidenced by high right heart filling pressure carried a much worse prognosis as compared to patients with elevated left ventricular enddiastolic pressure and pulmonary artery mean pressure due to left ventricular disease. The two year survival was 32.7% for those with elevated right heart filling pressure as compared to 65% for patients with elevated left ventricular filling pressure. This degree of divergence is likely to be due to the disparity in the severity of elevation of right and left heart filling pressure taken into consideration for analysis in this study, viz. 20 mm Hg or more of right heart filling more of left heart filling

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pressure includes both moderate as well as severe degree of elevation.

Presence or absence of embolic episodes was a significant determinant in both univariate and multivariate analysis while cardiac rhythm was a determinant in multivariate analysis. Ventricular mural thrombus being less common in tropical endomyocardial fibrosis as compared to Loeffler's eosinophilic endocardial fibrosis[8], the most probable source of systemic embolisation is likely to be from the atrium. Thus atrial fibrillation influences the survival.

Arterial mean pressure less than 70 mm Hg, arterial oxygen saturation less than 85% and pulmonary arterial oxygen saturation less than 70%, all reflect low cardiac output and have significant influence on survival.

It can be concluded from this study that tropical endomyocardial fibrosis is by and large a biventricular disease and an analysis of data from 206 patients over a sixteen year period on medical management with digoxin, diuretics and vasodilators carries not too good a prognosis with only a little more than half of the patients surviving at the end of five years since their first presentation. However, this study shows that the prognosis is not as bad as it had been reported in previous studies from African continent with smaller number of patients followed up for shorter duration.

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Table 1 Clinical profile in 206 patients of endomyocardial fibrosis at first presentation.

Variables	Percentage
Fever at onset	7.2
Chest pain	17.9
NYHA Class 3 and 4	39.3
Duration of symptoms less than 24 months	46.1
Age less than 15 years	28.1
Right heart failure	71.8
Left heart failure	25.7
Cyanosis	15.5
Clubbing	9.7
Ascites	28.1
Edema feet	24.3
No murmur	25.7

Table 2 Laboratory features in 206 patients at first presentation.

Variables	Percentage
Absolute eosinophil count more than 2000/cmm	8.7
Hemoglobin less than 10 g%	9.2
Cardiothoracic ratio more than 60%	64.1
Atrial fibrillation	33.9
Ventricular extrasystole	2.4
Junctional rhythm or Heart block	4.4
Right axis deviation more than $+90^{\circ}$	29.1
Intraventricular conduction delay more than 120 msec	16.0
Atrial tachycardia or flutter	3.4

Table 3 Hemodynamic features in 179 patients of endomyocardial fibrosis

Variables (mm Hg)	Percentage
Right atrial mean pressure	
less than 7	18.4
7 - 11	29.0
12 - 15	16.7
more than 16	35.8
Pulmonary artery mean pressure	
less than 20	41.3
20 - 30	30.2
31 - 49	14.5
more than 50	14
Left ventricular enddiastolic pressure	
less than 12	29.0
12 - 17	20.0
18 - 24	21.8
more than 25	29.0

Table 4 Cause of death in 74 patients with endomyocardial fibrosis who died during follow up.

	No.	Percentage
Congestive heart failure	17	22.9
Severe right ventricular failure, heaptic coma.	9	12.1
Embolic phenomena	11	14.8
Terminal arrhythmias	6	8.1
Unknown (death at home informed by letter)	27	36.5
Others*	4	5.4

* Meningitis (1), Drug allergy (1), Chronic obstructive airway disease (2).

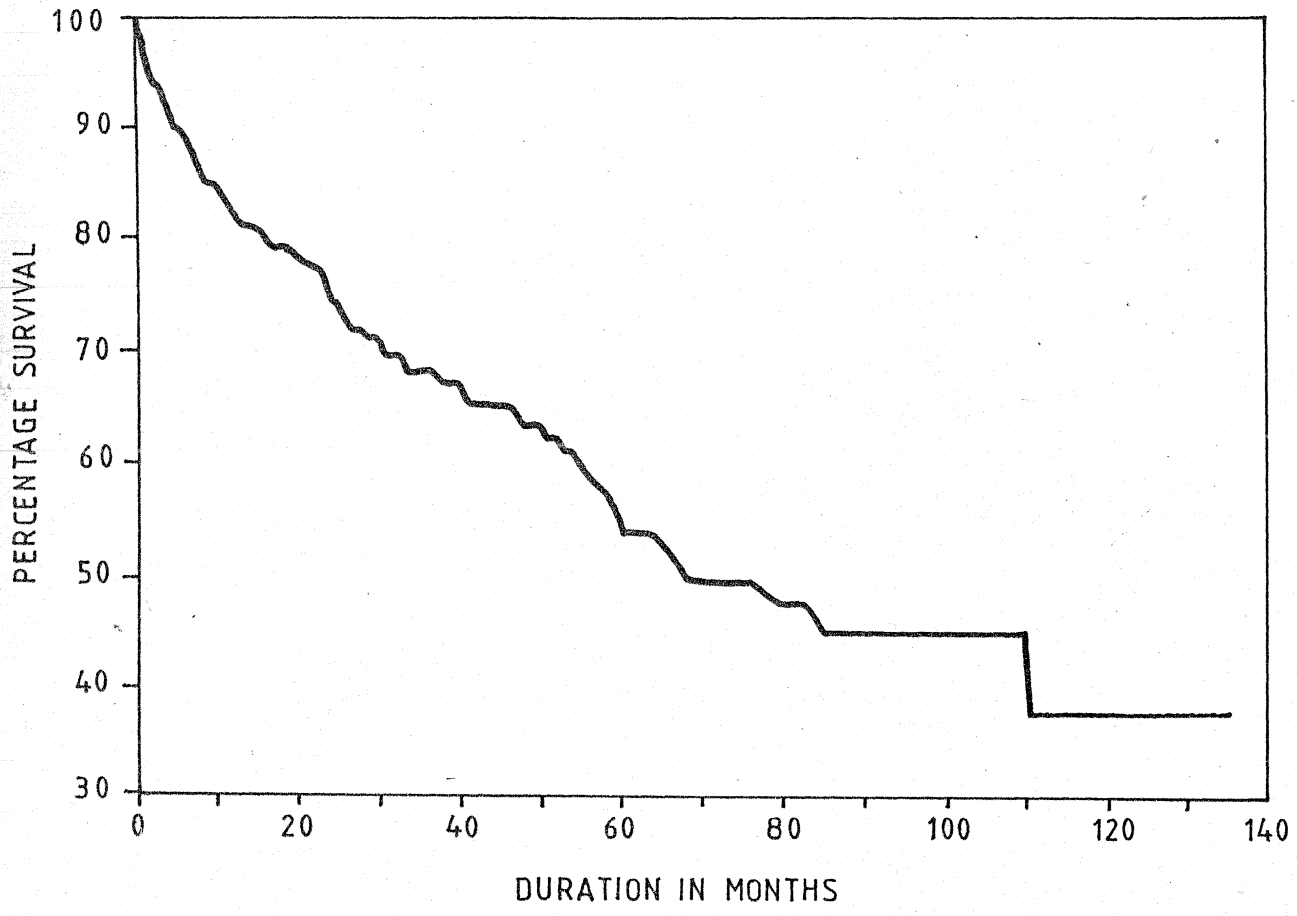
Table 5 Selected variables that were significant in univariate analysis - Log rank test of Kaplan Meier estimates

Variable	No. of patients	% Survival at 1 yr.	2 yrs.	5 yrs.	10 yrs.	P value
Grade of Mitral Regurgitation						
less than 2	136	83.6	74.7	62.7	33.4	.04
more than 2	43	78.8	58.4	28.5	None	
Right atrial mean pressure						
less than 20 mm Hg	145	87.2	76.1	63.7	39.2	.001
more than 20 mm Hg	34	60.1	43.7	11.9	None	
Right ventricular End diastolic pressure						
less than 20 mm Hg	145	87.2	78.3	61.7	40.8	.001
more than 20 mm Hg	34	59.9	32.7	21.8	None	
Pulmonary artery mean pressure						
less than 40 mm Hg	146	84.5	74.2	60.5	36.6	.006
more than 40 mm Hg	33	73.2	56.7	33.6	None	
Left Ventricular End diastolic pressure						
less than 15 mm Hg	89	86.6	76.6	64.2	50.5	.047
more than 15 mm Hg	90	78.2	65.0	44.8	None	
Aortic Mean Pressure						
less than 70 mm Hg	21	66.8	45.1	28.2	None	.019
more than 70 mm Hg	158	84.4	74.1	58.9	36.4	
Aortic Oxygen Saturation						
less than 85%	10	38.5	25.7	12.8	None	.001
more than 85%	169	84.7	73.2	60.4	37.9	
Pulmonary artery Oxygen saturation						
less than 70%	138	78.2	64.3	42.9	16.7	.001
more than 70%	41	94.8	88.6	88.6	88.6	

Table 6 Cox proportional hazards multivariate analysis of clinical and hemodynamic determinants of mortality

Variable	P value
NYHA class 1 & 2 vs 3 & 4	.01
QRS axis & $> +90^{o}$	.03
Embolic episodes present vs absent	.01
Cyanosis present vs absent	.03
Right ventricle S3 present vs absent	.005
Rhythm Atrial fibrillation vs Sinus	.03
Grade of tricuspid regurgitation > 2	.01
Right atrial mean pressure > 20 mm Hg	.01
Aortic mean pressure < 70 mm Hg	.04
Aortic oxygen saturation $< 85\%$	.001

SURVIVAL CURVE (KAPLAN-MEIER)
OF 206 PATIENTS



LIST OF PROCEDURES DONE
PROJECT REPORT

TITLE OF THE PROJECT: **CLINICAL PROFILE AND NATURAL HISTORY OF EBSTEIN'S ANOMALY OF TRICUSPID VALVE**

NAME: DR. PRAMOD KUMAR JAISWAL

PROGRAMME: D.M. CARDIOLOGY

MONTH & YEAR OF SUBMISSION: NOVEMBER, 1992

ABSTRACT

63 patients of Ebstein's anomaly of tricuspid valve were encountered from 1976 to 1991. 28 (44.4%) were male and 35 (55.6%) female. Their age at presentation ranged from 3 months to 51 years. 5 (7.9%) patients were asymptomatic, 48 (76.2%) had class II-III exertional dyspnoea, palpitation or both. 30 (47.6%) patients had cyanosis, 26 (41.3%) had clubbing, 26 (41.3%) had split S_1, 39 (61.9%) had widely split S_2, 35 (55.5%) had only systolic murmur, 25 (39.7%) had both systolic and diastolic murmur, while 3 (4.8%) had no murmur.

Electrocardiogram showed paroxysmal atrial fibrillation in 2, chronic atrial fibrillation in 4, (6.3%), paroxysmal supraventricular tachycardia in 7, atrial or ventricular ectopic beats in 5 (7.9%), 2 : 1 atrio-ventricular block in 1 (1.6%) complete atrioventricular block in 2 (3.2%) and type B WPW syndrome in 9 (14.3%) patients. X-ray chest showed no cardiomegaly in 7 (11.1%) and diminished vascularity in 22 (34.9%).

Diagnosis was established by both cardiac catheterization and echocardiography in 50 (79.4%) only cardiac catheterisation in 7 (11.1%) and only echocardiography in 6 (9.5%). Atrialized right ventricular chamber was demonstrated in 51 (80.9%) by angiography and in 40 (63.5%) by electrophysiology. Patients were followed up for 1 month to 172 months. 17 (26.9%) patients were operated. 3 (4.8%) patients died during medical follow up, and 5 (7.9%) died following surgery. Survival probability for 46 medical patients was 86.9% at 172 months. Factors affecting survival were pulmonary blood flow, cyanosis, clubbing and systemic arterial oxygen saturation.

INTRODUCTION: Ebstein's anomaly of Tricuspid valve was first described by Wilhelm Ebstein in 1866 in a 19 year old labourer who died due to tricuspid regurgitation caused by severe congenital malformation of tricuspid valve.[1] Yater and Shapiro reported the first case of Ebstein's anomaly with radiologic and electrocardiographic data in 1937, and Tourniaire et al diagnosed the anomaly in a living subject in 1949[2].

Its incidence is reported to be approximately 1 in 20,000 live births[3], and its prevalence is 0.5 percent or less among patients with congenital heart disease[4].

The natural history ranges from neonatal death to asymptomatic survival into late adulthood[5], and survival upto 85 years has been reported.[6]

Presentation during infancy, functional class 3 or 4, marked Cardiomegaly, cyanosis and recurrent paroxysmal tachycardias are reported to limit survival.

The present retrospective analysis was undertaken to assess the clinical and haemodynamic profile of Ebstein's anomaly, its natural history and factors influencing the survival.

Material and Methods

63 cases of Ebstein's anomaly of tricuspid valve without any major associated anomaly (eg. corrected transposition of great arteries or significant pulmonary stenosis) were encountered during a 16 year period from 1976 to 1991.

The diagnosis was confirmed by Cardiac Catheterization, and echocardiography in 50, only catheterization in 7 and only echocardiography in 6 patients.

28 patients (44.4%) were male and 35 (55.6%) were female. Their age at presentation ranged from 3 months to 51 years (Mean age 19.5 $\pm$ 10.95 years).

After a detailed workup, all the relevant clinical and laboratory findings were noted. The patients were followed up every six months by prior appointment. Those who did not come for follow up for more than six months were sent at least three reminder letters to know their state of health. Those who did not respond to letters were considered lost to follow up. In case of death at home or at local hospital, information regarding time and cause of death was obtained from family members whenever possible.

Follow-up duration ranged from 1 month to 172 months. Complete follow up was available in 52 patients (82.5%) while 11 patients were lost to follow up after 1 to 94 months. 17 patients (27%) underwent surgery and 46 (73%) were in medical follow-up.

Statistical Analysis

Student's T test and Fisher's exact test were used for analysis of quantitative and qualitative data respectively. Survival probability was calculated by Life-Table Method. All the data were entered into a computer data-base system and statistical analysis was done using SPSS/PC + Version 4 Advanced Statistics Module.

RESULTS

CLINICAL FEATURES

Salient clinical features of 63 patients at first presentation are shown in Table I and II. Main presenting symptoms were exertional dyspnoea (71.5%), exertional palpitation (36.5%) and history of cyanosis (30.2%). 4 patients had history of neonatal cyanosis which remitted in 2 of them and persisted in the other 2 patients. Paroxysmal palpitations were present in 10 patients (15.9%). 48 patients were in NYHA Class II and 10 in NYHA Class III. 5 patients were asymptomatic. Age at onset of symptoms was 6 months to 51 years (Mean 12.59 $\pm$ 10.63 years). 48.0% had cyanosis and 41.0% had clubbing. Other common physical signs were loud and split first heart sound, wide split second heart sound, third heart sound, grade 1/6 to 3/6 early systolic murmur and scratchy diastolic murmur at lower left sternal edge.

Radiological Features

Chest skiagram showed no cardiomegaly in 9 (14.3%), cardiothoracic ratio of 56-70% in 39 (61.9%) and more than 70% in 15 (23.8%). Diminished vascularity of lung fields was present in 22 (34.9%).

Electrocardiographic Features

Table III shows the electrocardiographic findings. Evidence of right atrial enlargement, complete right bundle branch block and polyphasic QRS complex each were present in about 50% of patients.

Type B W.P.W. Syndrome was found in 9 patients (14.3%). 11 patients (17.5%) had incomplete right bundle branch block. Supra ventricular tachycardia, paroxysmal or chronic atrial fibrillation, 2:1 atrioventricular block and complete atrioventricular block were present only in a minority.

Echocardiographic Features

Echocardiogram showed paradoxical interventricular septal motion in 82% and septal tricuspid leaflet displacement of 15 mm or more in 74.5% patients. The diagnostic criteria used were mitral to tricuspid closure delay of more than 80 msec, and displacement of septal tricuspid leaflet of more than 8 mm/m^2 body surface area[7].

Haemodynamic Features

Cardiac catheterization (Table IV) revealed normal left sided pressures in all patients and normal right sided pressures in the majority. Elevated right atrial mean pressure, right ventricular systolic pressure and pulmonary artery mean pressure were present in 14, 8 and 4 patients respectively. Oximetry showed right to left shunt in 19, bidirectional shunt in 14, no calculable shunt in 17 and left to right shunt in 4 patients. Atrialised chamber was documented by angiography in 98% patients and electrode catheter withdrawal in 89% patients. Right ventricular angiogram (Table V) showed grade 3 or 4 tricuspid regurgitation in 65% patients.

FOLLOW-UP AND TREATMENT

17 patients underwent surgical treatment during follow up. Usual indications for surgery were cyanosis, severe symptoms, severe tricuspid regurgitation or significant cardiomegaly. As shown in Table VI, the surgical patients had significantly lower systemic and mixed venous oxygen saturation, significantly higher incidence of Class III exertional dyspnoea, greater cardiothoracic ratio and higher incidence of grade 3 or 4 tricuspid regurgitation.

5 patients died following surgery and 3 patients died during medical follow-up.

Univariate analysis for factors influencing survival (Table VII) showed that lower mixed venous or pulmonary arterial oxygen saturation, lower systemic arterial saturation, lower pulmonary blood flow, a lower pulmonary to systemic blood flow ratio indicating pure or predominant right to left shunt, and presence of cyanosis or clubbing adversely affected the survival.

Life Table analysis for 46 patients on medical follow up showed a survival probability of 86.9% at 168 months with 95% confidence interval of 70.3% to 100% (Fig. 1).

Comparison of patients without and with cyanosis (Fig 2) showed no mortality in acyanotic patients as compared to a survival probability of 64.6% at 156 months in patients having cyanosis, (P value = 0.03).

Comparison of patients according to aortic oxygen saturation (Fig 3) showed no mortality in patients with more than 85% saturation while in those with 85% or less saturation the calculated survival probability was 66.7% at 72 months (P value = 0.0006). There was only one patient with less than 85% saturation during 72 to 84 months follow up period who also died.

DISCUSSION

The clinical profile of Ebstein's anomaly includes a spectrum ranging from severely symptomatic infant to relatively asymptomatic elderly.[2,4,5,8]

Watson[5] in an international Co-operative study of 505 cases reported an equal predilection for either sex, with 35 (6.9%) presenting during infancy and 279 (55.2%) cases presenting between 1 to 15 years of age. Our series also showed equal incidence in both sexes but a much lower percentage of patients presenting during infancy (3 patients 4.8%). 18 patients (28.6%) presented between 1 to 15 years of age which is similar to Bialostozky's series[9]. The paucity of neonates in this series may be attributable to the fact that ours is a tertiary referral centre and many of the sick infants do not survive long enough to reach this hospital. Furthermore, missed diagnosis may also contribute to a lower number of referrals in this age group.

The common presenting features were dyspnoea, palpitation and cyanosis which is similar to that reported by others.[8,9,10,11] Our series had higher number of patients (84%) in functional Class I

and II compared to 50% to 70% reported by others.[5,12] However, this is similar to the post neonatal subgroup of Gentle's Series[13].

Incidence of cyanosis varies in different series depending upon the proportion of cases presenting in neonatal period. In our series 48% had cyanosis which is similar to Bialostozky's report of 46%[9], and 41% in post neonatal subgroup of Gentles, series[13]. In the latter series[13] 16 out of 17 neonates had cyanosis. 95% of our patients had only systolic or systolic as well as diastolic murmurs which is similar to other reports[9]. Clinical evidence of heart failure was present in 15.8% which is similar to reported incidence of 14%[9]. Cardiomegaly was found on chest skiagram in the majority. Although the patients who died had somewhat greater Cardiothoracic ratio compared to survivors, this was not statistically significant. This is in contrast to other reports[8,11,13]. 22 of the 30 cyanosed patients had diminished vascularity of lung fields.

Electrocardiogram showed sinus rhythm and incomplete or complete right bundle branch block in the majority of cases, similar to other reports[9]. Type B W.P.W.syndrome was found in 14.3%, concordant with the reported incidence of 5 to 25%. None of the patients had normal electrocardiogram.

2 dimensional echocardiography was found to be more sensitive for diagnosis, compared to M-mode echocardiography. Atrialised right ventricular chamber, and morphology and mobility of tricuspid valve could be accurately assessed by 2-D. echocardiography. Doppler echocardiography reliably assessed severity of tricuspid regurgitation

nd helped in excluding tricuspid stenosis and other major associated nomalies.

57 patients underwent catheterization studies. Although many previous reports have stressed the high incidence of fatalities and rrhythmias (5.3% and 27.5% in Watson's study[5]), with greater experience, improved techniques, better equipment and safer contrast materials, the fatality rate has decreased markedly, and at present the ndications for cardiac catheterization in patients with Ebstein's anomaly are no different from other patients with congenital heart disease[15]. In our series there were only 5 complications in 4 patients including supraventricular tachycardia in 2, ventricular tachycardia in femoral arterial embolism in 1 and myocardial injection in 1. There were no fatalities related to catheterization. This low incidence of complications is similar to that reported by Kumar et al [11].

Majority of our patients had normal right heart pressures which is similar to other reports[11]. Finding of right to left shunt or bidirectional shunt in 33 patients, is similar to 19 out of 33 in Kumar's series[11].

Electrode withdrawal was done in 45 patients and showed characteristic dissociation between electrical and pressure phenomenon in 40 patients (88.9%). The false negative recordings may be explained by the withdrawal path, the catheter being at the mid atrial level or withdrawn very rapidly as suggested by Watson[5].

Right ventricular angiography was diagnostic of atrialized right ventricular chamber in 51 out of 52 patients (98.1%). The

angiogram in the remaining one patient was of poor quality. Moderate to severe tricuspid incompetence was found in 65.3% similar to 56.5% reported by others[11].

The survival probability in Ebsteins anomaly reported by various workers varies according to the proportion of cases presenting in neonatal period, with lower survival reported by those with a higher proportion of neonates.[4,11,13,14] Higher mortality is also reported in series or subgroups with significant associated lesions.[11,14]

Watson[5] reported an overall mortality of 15.2% by natural causes including an initial high mortality from congestive heart failure during the first few months of life, followed by an average of 12.4% which was scattered fairly uniformly throughout childhood and adolescence. Our finding of 13.1% mortality over a 172 months follow up is in concordance with Watson's report.

Various workers have reported that the factors adversely affecting survival are, presentation during early infancy[11,14] presence of significant associated lesions[5,11], more marked cyanosis[4,11] extreme cardiomegaly[9,11,13] N.Y.H.A. Class III or IV[9,13], paroxysmal tachycardias[4,9], and a higher ratio of combined area of right atrial and atrialized right ventricle to that of the combined area of functional right ventricle, left atrium and left ventricle in four chamber view at end diastole on echocardiography.[14] The latter information was not available in this retrospective analysis.

We found that a lower mixed venous, and systemic arterial oxygen saturation adversely affected survival. Lower pulmonary blood

flow, a pure or predominant right to left shunt on oximetry, and clinically appreciable cyanosis or clubbing also adversely influenced survival. This emphasizes the need for surgical intervention in the cyanotic patients. Determination of arterial oxygen saturation can clarify the issue in cases with doubtful cyanosis. Since haemodynamic variables were not found to influence survival, catheterization studies may be reserved for cases needing concurrent electrophysiological studies. However it is well recognised that echocardiographic delineation is usually better and catheterization is rarely necessary.[16,17]

We did not find cardiomegaly to be a significant predictor of survival, which is similar to the large series of Watson[5]. Patients who died had more severe dyspnoea compared to survivors, but this did not reach statistical significance. These two negative findings may possibly be due to comparatively smaller number of total patients and deaths.

Better survival probability in the present study compared to some other reports[4,11,13,14] is explained by a lower proportion of neonates and infants in the present series. This indicates possible referral bias in other series as the age range in the present study is more similar to the large series of Watson[5].

Surgery for Ebstein's anomaly was associated with a high mortality in the past[5]. With improvements in surgical techniques, the operative mortality is considerably lower at present.[18] In a recent review Mair[16] recommended operation for all patients with Ebstein's anomaly whose condition, despite medical therapy has deteriorated to

unctional Class III or IV, and also for patients with less severe symtoms who show progressive cardiac enlargement.

ONCLUSION

Ebstein's anomaly is by and large a benign congenital heart isease in the majority of patients. Natural attrition is about 1% er year. The determinants of survival were pulmonary blood flow, cyaosis, clubbing, and systemic arterial oxygen saturation. Surgery is ndicated only in severe forms of Ebstein's indicated by severity of yanosis or severity of tricuspid regurgitation.

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TABLE - I

	SYMPTOMS	No.	(%)
1.	D.O.E	45	(71.5)
	CLASS II	35	(55.6)
	CLASS III	10	(15.9)
2.	PALPITATION	33	(52.4)
	EXERTIONAL	23	(36.5)
	PAROXYSMAL	10	(15.9)
3.	CHEST PAIN	13	(20.6)
4.	H/o CYANOSIS	19	(30.2)
	NEONATAL	4	(6.4)
	LATE ONSET	15	(23.8)
5.	H/o EMBOLISM	2	(3.0)
6.	H/o BRAIN ABSCESS	3	(4.8)
7.	H/o SYNCOPE	4	(6.4)

5 PATIENTS WERE ASYMPTOMATIC

Abbreviations Used

D.O.E = Dyspnoea on exertion

H/o = History of

TABLE - II

	PHYSICAL SIGNS	No.	(%)
1.	CYANOSIS	30	(48.0)
2.	CLUBBING	26	(41.0)
3.	ELEVATED J.V.P.	10	(15.8)
4.	LOUD S_1	21	(33.3)
5.	SPLIT S_1	26	(41.3)
6.	WIDE SPLIT S_2	39	(61.9)
7.	S_3	46	(73.0)
8.	S_4	9	(14.3)
9.	SYSTOLIC MURMUR	43	(68.3)
10.	DIASTOLIC MURMUR	25	(39.7)

TABLE - III

ELECTROCARDIOGRAPHIC FEATURES

		No.	(%)
1.	RA ENLARGEMENT	32	(51.0)
2.	INCOMPLETE RBBB	11	(17.5)
3.	COMPLETE RBBB	33	(52.4)
4.	POLYPHASIC QRS	30	(47.6)
5.	WPW TYPE B	9	(14.3)
6.	S.V.T.	7	(11.1)
7.	CHRONIC A.F.	4	(6.4)
8.	PAROXYSMAL A.F.	2	(3.2)
9.	2 : 1 A.V. BLOCK	1	(1.6)
10.	COMPLETE A.V. BLOCK	2	(3.2)

Abbreviations used

RA = Right atrial
RBBB = Right Bundle Branch Block
WPW = Wolf Parkinson White Syndrome
S.V.T. = Supraventricular tachycardia
A.F. = Atrial fibrillation
A.V. = Atrioventricular

TABLE - IV

HAEMODYNAMIC FEATURES

	No. OF CASES
RIGHT ATRIAL MEAN PRESSURE >7 mmHg	14
RIGHT VENTRICULAR SYSTOLIC PRESSURE >30 mmHg	8
PULMONARY ARTERIAL MEAN PRESSURE >18 mmHg	4
LEFT VENTRICULAR OR AORTIC OXYGEN SATURATION<85%	13
RIGHT TO LEFT SHUNT	19
LEFT TO RIGHT SHUNT	4
BIDIRECTIONAL SHUNT	14
NO SHUNT	17

TABLE - V

SEVERITY OF TRICUSPID REGURGITATION BY ANGIOGRAM

GRADE	No.	(%)
0	2	(3.8)
1	3	(5.8)
2	13	(25.0)
3	19	(36.5)
4	15	(28.8)

TABLE - VI

COMPARISON OF MEDICAL AND SURGICAL PATIENTS

VARIABLE	MEDICAL (n = 46) MEAN (S.D)	SURGICAL (n = 17) MEAN (S.D.)	P VALUE
C.T.R. (%)	63.4 (9.1)	67.5 (7.5)	N.S.
M.V. O_2 (%)	67.8 (8.1)	59.9 (9.9)	0.005
Ao O_2 (%)	92.4 (5.8)	83.6 (11.5)	0.015
	No. (%)	No. (%)	
DOE GR. 3	4 (8.7)	6 (35.3)	0.01
T.R. GR. 3-4	20 (43.5)	12 (70.1)	N.S.

Abbreviations Used

C.T.R.= Cardio Thoracic Ratio ; O_2 (%) = Oxygen Saturation
M.V. = Mixed Venous ; Ao = Aortic
D.O.E.= Dyspnoea on exertion ; T.R. = Tricuspid Regurgitation

TABLE - VII

DETERMINANTS OF SURVIVAL BY UNIVARIATE ANALYSIS

A: QUANTITATIVE VARIABLES

S. No.	VARIABLE	ALIVE (n = 43) MEAN ± S.D.	DEAD (n = 3) MEAN ± S.D	P. VALUE
1.	MVO_2 (%)	69.1 ± 6.6	53.3 ± 7.7	.001
2.	RAO_2 (%)	73.6 ± 6.7	58.5 ± 6.4	.004
3.	RVO_2 (%)	74.7 ± 6.1	56.0 ± 6.1	<.001
4.	PAO_2 (%)	74.3 ± 7.8	53.0 ± 1.4	.001
5.	LAO_2 (%)	92.9 ± 5.1	76.0 ± 2.8	<.001
6.	LVO_2 (%)	90.8 ± 4.1	74.0 ± 3.6	<.001
7.	Ao O_2 (%)	93.6 ± 3.7	78.5 ± 0.2	<.001
8.	Q.P. (l/min)	4.6 ± 1.6	1.2 ± 0.5	.001
9.	QP : QS ratio	1.22± 0.40	0.56± 0.28	.01

B.: QUALITATIVE VARIABLES

S. No.	VARIABLE	ALIVE (n = 43) No.	%	DEAD (n = 3) No.	%	P. VALUE
1.	CYANOSIS	13	(30.2%)	3	(100%)	.037
2.	CLUBBING	10	(23.3%)	3	(100%)	.019

Note: Quantitative variables were analysed using Student's T test, and Qualitative variables were analysed using Fisher's exact test.

Abbreviations: O_2 (%) = Oxygen Saturation (%) ; M.V. = Mixed venous ; RA = Right atrial ; RV = Right Ventricular ; PA = Pulmonary arterial;; LA = Left atrial ; LV = Left Ventricular ; Ao = Aortic
QP = Pulmonary Blood Flow ; QP : QS = Pulmonary to systemic blood flow

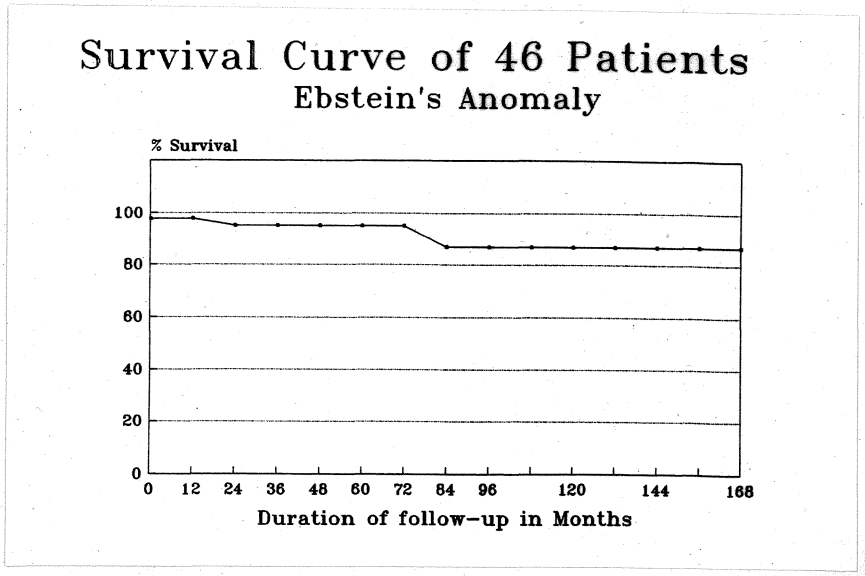

Fig 1: Survival curve for 46 patients on medical follow up

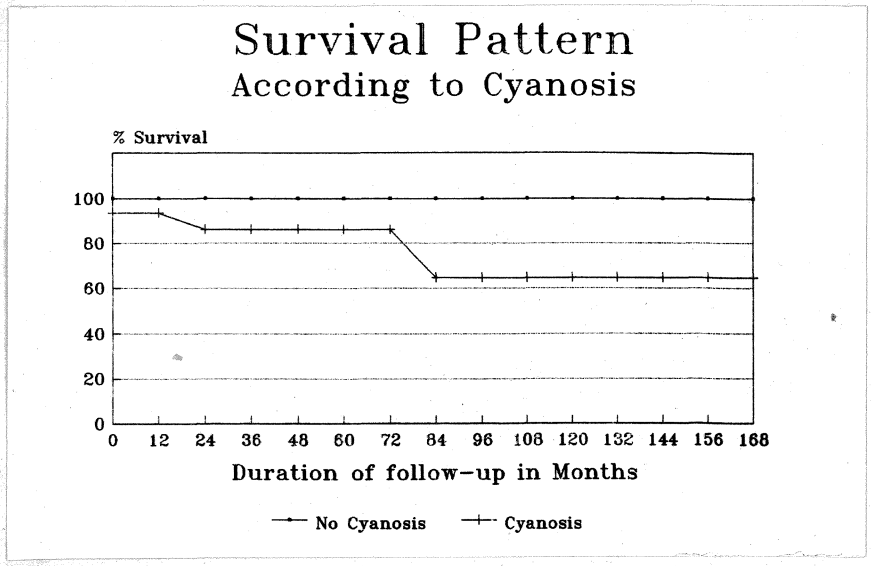

Fig 2: Survival curve according to cyanosis

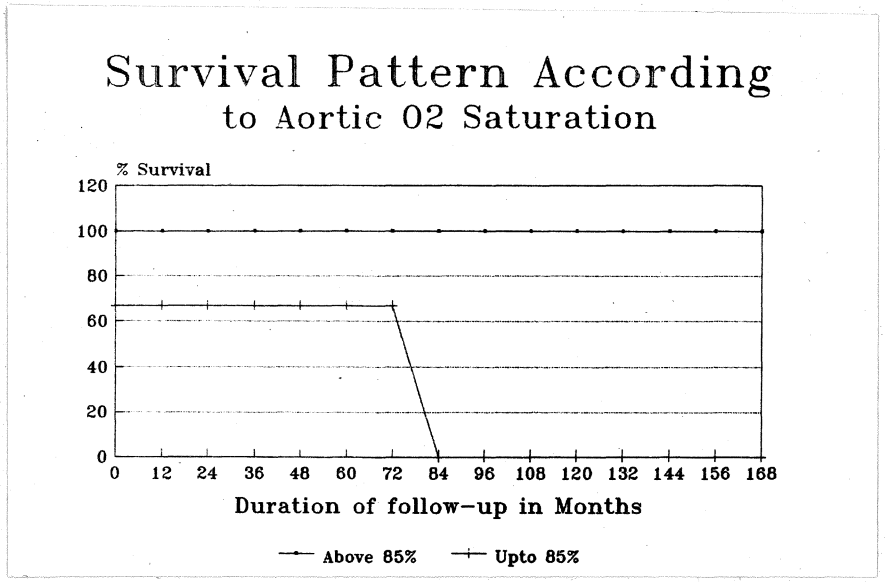

Fig.3: Survival curve according to aortic oxygen saturation

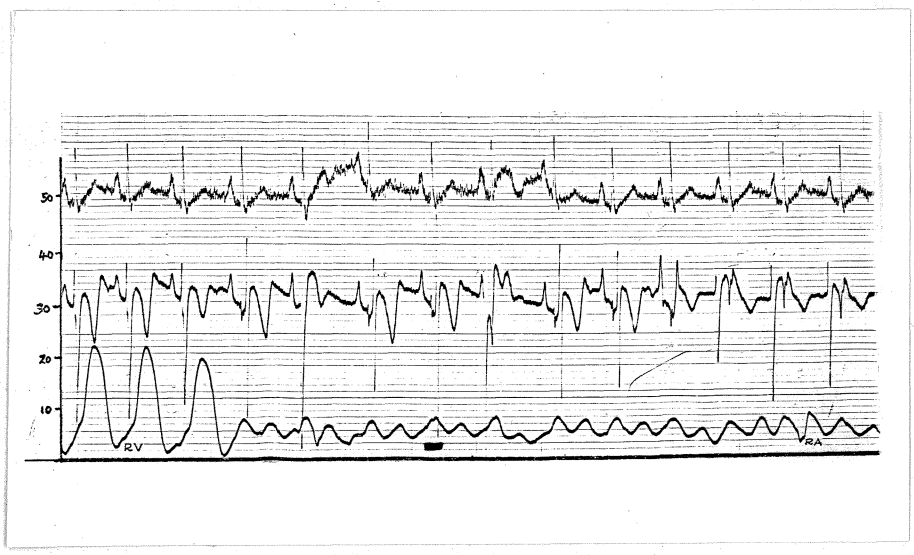

Fig 4: Electrode withdrawal tracing showing dissociation between electrical and pressure phenomenon

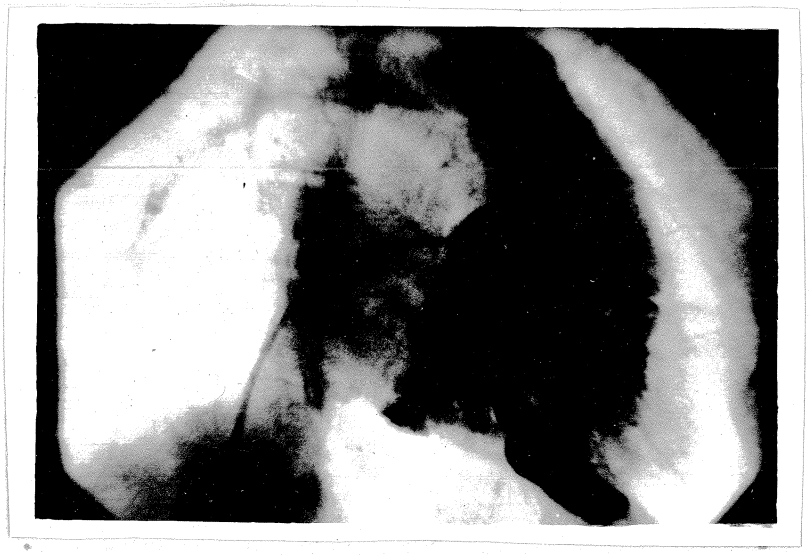

Fig.5: Right ventricular angiogram showing (1) displaced tricuspid leaflet, (2) atrialized right ventricular chamber and (3) tricuspid regurgitation.