P70

C O N T E N T S

CERTIFICATE

I, Dr. ARABINDA SAHA .....hereby declare that I have actually ~~performed all the procedures listed~~ carried out the projects under report.

Signature..... Arabinda Saha

Place : Trivandrum

Name in ARABINDA SAHA .....
capital letters

Date : 17.11.92

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

Signature

Head of the department

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

SUMMARY

Twenty patients (7 males and 13 females) of established endomyocardial fibrosis (EMF) with age ranging from 10 to 56 years (Mean 26.7±11.78 yrs) underwent detailed electrophysiological study from October 1991 to October 1992. 10 patients had right ventricular and remaining biventricular EMF.

Mean atrial pacing threshold was 0.72±0.46 mA, mean atrial injury potential was 0.99±0.73mV, mean atrial refractory period (ERP) was 27.6 ± 41.44 msec.

Sinoatrial conduction time and corrected sinus node recovery time were prolonged in 4 patients each. 1 patient had prolonged AH interval while 3 patients had prolonged HV interval. Mean pacing threshold at right ventricular apex (RVA) and right ventricular outflow tract (RVOT) were 1.87 ± 1.36 mA and 0.79 ± 0.38 mA respectively ($p < 0.01$). Injury potential at RVA and RVOT were 1.39 ± 1.25 mvolt and 2.89 ± 2.26 mvolt respectively ($p < 0.05$). Mean slew rate at RVA was 0.43 ± 0.28 volt/sec. while same value in control subjects at RVA was 0.735 ± 0.1 volt/sec. ($p < 0.01$). Thus EMF significantly alters ventricular electrophysiological parameters at apical region while atrial electrophysiological parameters are not significantly altered.

Key words: Myocardial Disease, Restrictive Cardiomyopathies. Endomyocardial fibrosis. Electrophysiology. Pacemakers, Artififial.

INTRODUCTION

Endomyocardial firbosis (EMF) is a severe restrictive cardiomyopathy characterised by fibrosis of the endocardium and to a lesser extent in the subendocardial myocardium of one or both vetricles. This results in decrease in ventricular compliance and atrioventricular valve dysfunction when it involves the atrioventricular valve apparatus. Bedford and Konstan[1] first drew attention to an unexplained type of heart failure amongst West African troops-the nature of the disease was subsequently found to be probably endomyocardial fibrosis. Davis[2] first defined endomyocardial fibrosis in 1948 from Uganda. Subsequently the clinical and laboratory features of Endomyocardial fibrosis has been described well from Africa, South America[3,4] and from India[5,6]. Electrocardiographic feature of this disease have been well described[7,8]. Incidence of atrial fibrillation is variable from 16 to 75% in various series[5,7,8]. Other abnormal rythm seen are paroxymal atrial tachycardia, junctional rythm, heart block and rarely sick sinus syndrome[5,7,8]. Other features like right atrial enlargement, left atrial enlargement, QRS axis shifts and relatively low voltage in ECG are described[5,7]. There is a paucity of data regarding the intracardiac electrophysiology in endomyocardial fibrosis. Emslie-Smith et al[8] recorded

intracardiac ECG and endocardial contact potential in 21 patients with right ventricular EMF (RVEMF) and had shown increased amplitude of p wave in 9 patients while right ventricular intracavitary potentials were similar to those noted in patients with congenital and rheumatic heart disease. The endocardial contact potential recordings in all patients showed normal ST segment elevation when obtained from right ventricular outflow tract whereas the potential from right ventricular apex showed ST segment elevation only in 14 to 21 patients. They concluded that loss of contact potential was due to fibrosis and mural thrombosis and hence did not regard this finding as a diagnostic test for endomyocardial fibrosis. Other investigators noticed higher pacing threshold in right ventricular apex in patients with RVEMF and used this feature to distinguish them from Eustein's anomaly of the tricuspid valve[10]. We observed difficulty in obtaining a low stimulation threshold during permanent pacemaker implantation in our patients with right ventricular EMF. Interestingly we also noted that the ventricular response in atrial fibrillation was slow even in those patients of EMF who were not receiving drugs like digitialis. This study was undertaken to examine the electrophysiological properties of the heart in patients with proven endomyocardial fibrosis.

MATERIALS AND METHODS

20 patients with established endomyocardial fibrosis diagnosed by clinical, roentgenographic, electrocardiographic, haemodynamic and angiographic features,[11] underwent electrophysiologic study in a post-absorptive state under local anaesthesia.

The protocol of electrophysiological study was as follows:-

A. In patients with sinus rhythm the following parameters were measured.

i) Right atrial electrogram through the pacing catheter connected to the chest lead of electrocardiograph at a frequency range of 0 to 50 Hertz. The electrode tip was pressed gently against the right atrial wall to record the Ta segment (to detect injury potential).

ii) Sinus node function study in the form of sinus node recovery time[12] and sinoatrial conduction time[13] and determination of atrial pacing threshold using standard techniques.

iii) Single atrial extrastimulation with decremental coupling interval to document AV nodal refractioness and atrial effective refractory period.[14]

iv) Unipolar ventricular electrogram recarded from

right ventricular apex and outflow tract by connecting the pacing lead terminal to the precordial lead of electrocardiograph, at a frequency range of 0 to 50 Hertz - the tip of pacing lead was pressed gently against the endocardium to record the ST segment elevation (injury potential). The intrinsic deflection (ID) was calculated from ventricular electrocardiogram and the slew rate (dv/dt). The last two were calculated from the right ventricular apical electrogram only.

v) Ventricular pacing threshold in milliamperes was determined in right ventricular apex and outflow region, using impulse with pulse width of 1.9 msec.

vi) Ventricular effective refractory period was determined using single extrastimulus at a decremental coupling interval from right ventricular apex and outflow.

B. Patients in Atrial fibrillation:

i) Atrial electrogram was obtained as per the previously described method to detect type of atrial fibrillation. 4 patterns are described.[15]

Type I-discrete atrial activity with clear baseline in between.

Type II -discrete atrial activity with irregular (non-isoelectric) baseline.

Type III-No discrete atrial activity with irregular baseline.

Type IV -Combinations of above 3.

ii) Ventricular pacing threshold, injury potential intrinsic deflection and its slew rate and effective refractory period were determined as in patients with sinus rythm.

C. 4 patients (undergoing electrophysiological evaluation for conduction system disease or for various tachyarrhythmias - who did not have endo- myocardial disease also underwent ventricular injury potential and slew rate recordings from right ventricular apex served as controls.

Statistical Analysis

All values, expressed as continuous variables were calculated as mean $\pm$ standard deviation. Discretes variables were expressed as percentages. Comparison between continuous variables were done by using unpaired 't' test.

OBSERVATION AND RESULTS

20 patients (7 males and 13 females) with age ranging of from 10 to 56 yrs. (Mean $\pm$ SD 26.7 $\pm$ 11.78 yrs) underwent electrophysiological study. 10 patients had only right

ventricular endomyocardial fibrosis (RVEMF) while 10 patients had biventricular endocmyocardial fibrosis (BVEMF) out of which 2 patients had predominant left ventricular endomyocardial fibrosis (LVEMF) with minimal RVEMF.

The pacing threshold at right atrium at pulse width of 1.9 msec. was determined in 7 patients with sinus rythm - The value was 0.72 ± 0.46 milliampere (range 0.4 to 1.7 MA). The atrial injury potential peak was 0.99 ± 0.73 millivolt (mv) with a range from 0.23 to 2.2 mv. Mean atrial effective refractory period was 276 ± 41.44 msec. (range 225 to 320 msec.) Sinoatrial conduction time was determined by Narula's method[13] in 9 patients with sinus rythm - it was more than 120 msec in 4 patients (44.44%). Corrected sinus node recovery time was estimated in 11 patients - in 4 patients it was prolonged beyond 525 msec. (Table 1). During incremental atrial pacing the pacing cycle length at which AV nodal Wenckebach phenomenon was observed varied from 300 to 500 msec. (Mean ± SD 383.75 ± 84.00 msec). None developed AV nodal Wenekebach Phenomenon at a Cycle length greater than 500 msec. (Table 2).

The basal His bundle recordings showed a mean AH interval of 108 ± 24 msec, while the mean HV interval was 43.89 ± 12.73 msec. AH interval was greater than 130 msec. in only 1 out of 13 patient while HV interval was beyond 55

msec in 3 patients (16%). (Table 2)

The mean pacing threshold at right ventricular apex was 1.87 ± 1.36 mA (at pulse width of 1.9 mesec) while at right ventricular outflow tract it was 0.79 ± 0.38 mA. This difference was statistically significant ($p < 0.01$). The injury potential (elevation of ST-segment from the baseline) at right ventricular apex and outflow tract were 1.39 ± 1.29 mv. and 2.89 ± 2.26 mv. respectively. This difference was also significant statistically ($p < 0.05$). Table 3). The injury potential at RV apex in control patients (3.27 ± 0.72 mvolts) was significantly higher ($p < 0.01$) than in patients (Table 4).

The intrinsic deflection of the ventricular electrogram at right ventricle was determined in 15 patients. It was purely negative in 10 patients and biphasic in remaining 5 patients. The mean value was 5.03 ± 2.07 mvolts with a range 1.58 to 7.6 mvolts (Table 3). The slew rate (dv/dt) (mean ± SD 0.43 ± 0.28 volt/sec.) in EMF patients was significnatly lower than in controls (0.735 ± 0.1 volt/sec.) (p 0.01) Table 4.

The effective refractary period at apex (266.28 ± 37.38 msec). was not significantly ($p>0.1$) different from the value noted at out flow tract (254.14 ± 43.00 msec)(Table 3).

DISCUSSION

Endomyocardial fibrosis (EMF), a severe form of obliterative restrictive cardiomyopathy has been shown to have various rhythm disturbances. Atrial fibrillation is seen in 16 to 75% of patients[5,7,8] other abnormal rythms being paroxysmal atrial tachycardia, atrial flutter, junctional rythm, varying degree of heart block and rarely sick sinus syndrome[5,7,8]. A slow ventricular response to atrial fibrillation is a common feature in endomyocardial fibrosis even in the absence of drug therapy to prolong AV nodal conduction. Difficulty in obtaining a low stimulation threshold from right ventricular cavity is a well documented phenomenon[10]. Absence of injury potential in intracardiac electrocardiogram recorded from right ventricular apex with preserved injury potential in electrogram obtained from right ventricular outflow tract was reported earlier by Emslie-Smith et al[9].

In the present study the stimulation threshold at right atrium was 0.2 $\pm$ 0.46 mA. which is well within the acceptable range at 1.5 mA at a pulse width of 0.5 msec[16]. The atrial injury potential (elevation of Ta segment) cannot be compared with normal because of absence of such data in normal persons. Mean effective refractory period of atria was on the higher side in the patients with sinus rythm.

The one-way sinoatrial conduction time (SACT) was prolonged in 44.44%. Normal values reported vary from 82 to 120 msec[12,17]. Only 4 out of 11 patients had prolonged corrected sinus node recovery time - (normal value 525 msec. or less)[18]. AV nodal effective refractory period was within normal limits. AH interval was normal in most patients while 3 patients (16%), had a prolonged HV interval. Thus a definite abnormality in atrial electrophysiology or in AV nodal conduction is not very apparent. In 5 patients with atrial fibrillation atrial activity could be recorded with rate varying from 420 to 500 per second. Thus atrial rate during AF also does not provide any definite reason for slow ventricular response.

The dense scar tissues may be several millimeters thick at the apical region and may extend towards the tricuspid ring encasing the papillary muscles. The infundibulum is spared and dilated[9]. Hence a difference in stimulation threshold between right ventricular apex and right ventricular outflow is expected. In our study this difference was striking (1.87 $\pm$ 1.46 mA at apex and 0.79 $\pm$ 0.38 mA at outflow; $p < 0.01$). The reported normal value of stimulation threshold being 0.8 mA at 0.5 msec. pulse width[16]. This findning is in concurrence with earlier report from Brazilian investigators who even used it as a differentiating point between right ventricular

endomyocardial fibrosis and Ebstein's anomaly of the tricuspid valves[10]. Another consequence of the apical/inflow fibrosis is the very low to unrecordable injury potential from electrogram recorded from RV inflow/apical regions. In our series we found a significantly higher ($p < 0.05$) injury potential at the right ventricular outflow tract as compared to right ventricular apex. Compared to controls the value from RV apex was significantly lower ($p < 0.01$) Our results corroborate that reported by Emslie - smith et al[9] who found that injury potential was not recordable from right ventricular apex in 7 out of 21 patient with RVEMF In all of them injury potential was recorded from right ventricular outflow tract. However they have not reported the exact values and there was no statistical comparison between values obtained from right ventricular apex and outflow tract. They attributed this loss of injury potential (contact potential) to endocardial fibrosis and overlying mural thrombus.

Injury potential correlates closely with the stimulation threshold during pacing. In fact an electrode position that offers negligible or absent current of injury, frequently represents malposition or contact with nonviable or fibrotic cardiac tissue and is often associated with a

higher than acceptable stimulation threshold[19]. Average ST segment elevation during implantation is 2.6 millivolt(mv)[20]. An injury potential of at least 2mv suggests satisfactory electrode tissue interface[19].

The ventricular electrogram obtained from normal myocardium during pacemaker implantation is usually biphasic. The mot important bioelectric event within the depolarisation signal is represented by a vertical, nearly straight line deflection with a rapid voltage transition- described by Lewis and Wilson as the Intrinsic deflection(ID)[21]. Usually endowed with the highest peak-to peak amplitude and Steepest slope the ID is well suited as the ideal cardiac signal since it most consistently meets the essential requisites for pacemaker sensing[19]. The mean amplitude for the endocardial ID signal is approximately 12 mv with a range of 4 to 20 mv[20]. In our series the mean ID was 5.03 $\pm$ 2.07 volts with a range of 1.58 to 7.6 mv which is well below the acceptable normal values. The slew rate, another physical characteristic of ID, is essential for proper sensing function[19] defined as the rate of change in signal voltage (dv/dt). The slew rate is expressed as volts per second. Most normal ID signals have a measured slew rate between 0.5 and 7 volts/sec. In our series the mean slew rate from right ventricular apex was 0.43 $\pm$ 0.28 volt/sec. and was significantly lower than the slew rate

obtained from 4 control patients (0.735 ± 0.11 volt/sec.) ($p < 0.01$). Hence this low ID and slew rate predicts a poor sensing in the right ventricular apex in patients with endomyocardial fibrosis.

Effective refractory period was not different significantly ($p > 0.1$) between RV apex (266.28 ± 37.38) and RV outflow (254.14 ± 43.00), probably because the fibrotic process does not interfere with the inherent electrophysiological property of the underlying myocardium.

In conclusion endomyocardial fibrosis significantly altered the electrophysiological parameters in the ventricles by elevating pacing threshold, decreasing intrinsic deflection and slew rate and decreasing injury potential in right ventricular apex compared to right ventricular outflow tract. This has an important bearing if the patient requires endocardial pacing. Atrial electrophysiological properties are not significantly altered although there was a trend toward higher HV interval, prolonged sinoatrial conduction time and corrected sinus node recovery time in a minority of patients. Further studies are necessary to look for other atrial or atrioventricular nodal electrophysiological abnormalities.

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TABLE No.1

ATRIAL ELECTROPHYSIOLOGICAL PARAMETERS IN PATIENTS WITH ENDOMYOCARDIAL FIBROSIS

Parameter	Mean	Standard deviation	Range
* Pacing threshold (Milliampere) (7)	0.72	± 0.46	0.4 to 1.7
* Injury potential (Millivolts) (7)	0.99	± 0.73	0.25 to 2.2
* Effective refractory period (Milliseconds)(5)	276.00	± 41.44	225 to 320
* Sinoatrial conduction time (one-way) (milliseconds)(9)	131.11	± 39.5	70 to 195
* Corrected sinus node recovery time (milliseconds) (11)	451.82	±367.65	180 to 1470

* Number in parenthesis indicate the number of patients in whom the parameter was measured.

TABLE No. 2

ATRIOVENTRICULAR NODAL CONDUCTION PROPERTIES IN PATIENTS WITH ENDOMYOCARDIAL FIBROSIS

Parameter	Mean	Standard deviation	Range
* A1l interval (Milli seconds) (13)	108 ±	24	75 to 160
* HV interval (milliseconds) (10)	42.89±	12.73	30 to 80
** S1-S1 at which AV nodal wenckebach occurs (milliseconds)(5)	383.75 ±	84.00	300 to 500

* Number of parenthesis indicate the number of patients in whom the parameter was measured.

**S1-S1 = Pacing cycle length.

Table No.3

VENTRICULAR ELECTROPHYSIOLOGICAL PARAMETERS IN PATIENTS WITH ENDOMYOCARDIAL FIBROSIS

	Right Ventricular apex	Right Ventricular outflow	Differnce
Pacing threshold (milliampere)	1.87 ± 1.36 (n=17)	0.79 ± 0.38 (n=18)	p <0.01
Injury potential (millivolts)	1.39 ± 1.25 (n=18)	2.89 ± 2.26 (n=17)	p <0.05
Effective refractory period(milliseconds)	266.28 ± 36.38 (n=14)	254.15 ± 43.00 (n=14)	p >0.1
Intrinsic deflection ID (millivolt)	5.03 ± 2.07 (N=15)	-	-

* Values expressed as mean ± standared deviation

* n = number of patients in whom the parameter was measured

TABLE No. 4

COMPARISON OF ELECTROPHYSIOLOGICAL PARAMETERS

BETWEEN PATIENTS WITH ENDOMYOCARDIAL FIBROSIS AND CONTROLS *

Parameter	Patients (determined at right) ventricular apex)	Control	Differences
Slew rate (Volt/sec)	0.43 ± 0.28 (n=16)	0.735±0.11 (n=4)	p<0.01
Injury potential (millivolts)	1.39 ± 1.25 (n=18)	3.27 ±0.72 (n=4)	p<0.01

* Values expressed as mean ± standard deviation
n = number of patients in whom the parameter was clucked

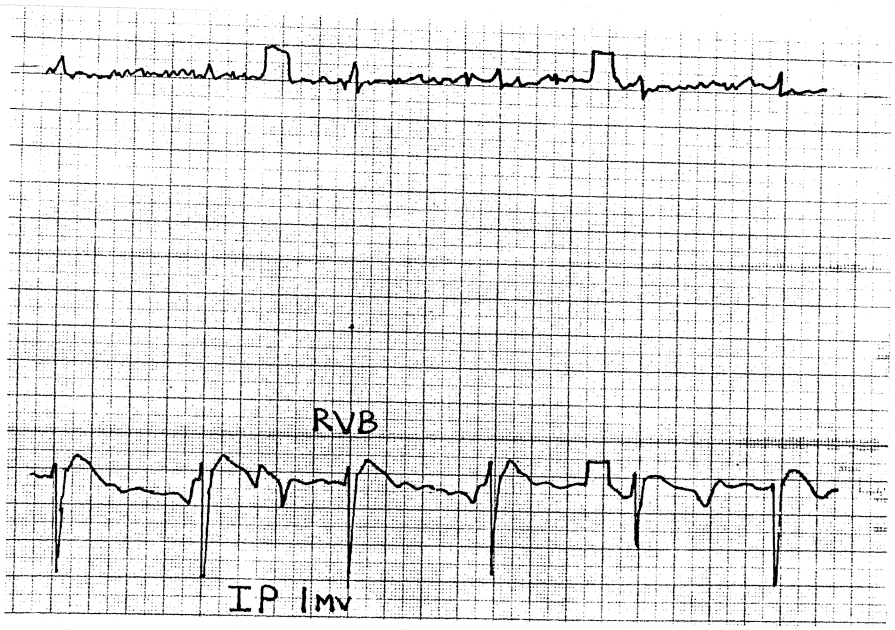

Fig. 1. Intracardiac electrogram in right ventricular body (RVB) showing an injury potential (IP) of 1 mvolt.

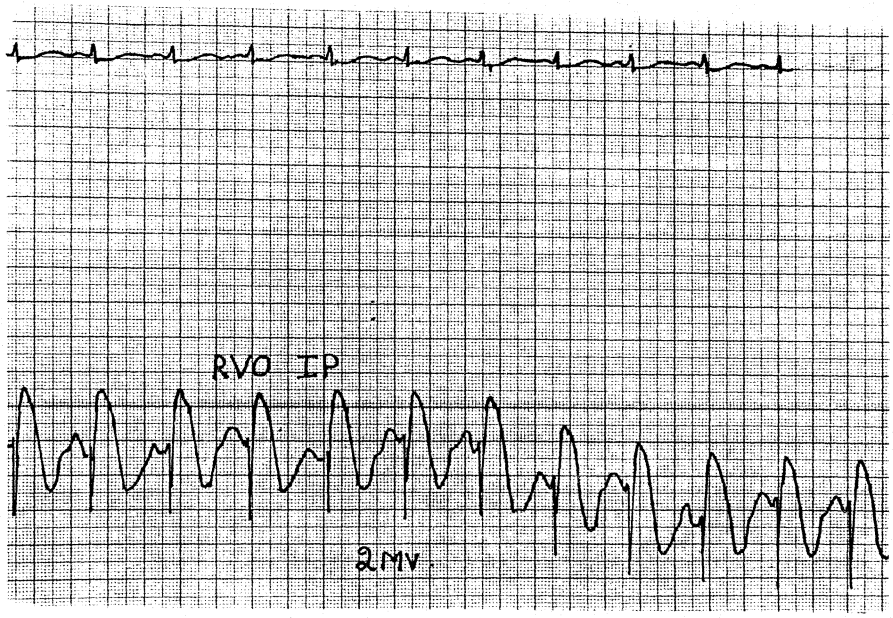

Fig. 2. Intracardiac electrogram (from the same patient, as in figure 1) recorded at right ventricular outflow (RVO) showing an injury potential at 2 mvolts.

~~LIST OF PROCEDURES DONE~~

PROJECT REPORT

TITLE OF THE PROJECT: **PROGNOSIS FOR PATIENTS WITH EISENMENGER SYNDROME OF VARIOUS AETIOLOGY**

NAME..... **ARABINDA SAHA**

PROGRAMME :.... **DM (CARDIOLOGY)**

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

ABSTRACT

Objective: To determine the long-term survival pattern, variables affecting long-term survival and complications occuring during follow-up of patients with Eisenmenger Syndrome.

Design: Retrospective study of patients with the diagnosis of Eisenmenger syndrome who have been followed up.

Setting: A tertiary care centre providing superspeciality services in various displines.

Subjects: 201 patients with Eisenmenger Syndrome - diagnosed by a combination echocardiography and peripheral arterial oxygen saturation study and/or cardiac catheterisation with or without angiocardiography - worked up and followed up for variable duration over a period of 16 yrs from 1976 to 1992.

Results: 109 patients were female and 92 were male - age of presentation varied from 3 months to 62 yrs (mean $\pm$ Standard Deviation 19.23 $\pm$ 12.62 yrs). A total of 12 different anatomic lesions were seen - commonest three being of ventricular septal defect (33.33%), atrial septal septal defect (29.85%), and patent ductus arteriosus (14.23%) History, Physical examination, chest skiagram and electrocardiogram established only the presence of pulmonary arterial hypertension except where differential cyanosis

indicating ductus was discernible or the degree of splitting of second heart sound provided some clue to the level of shunt. Contrast echocardiography done in 25.4% established the level of shunt in all. In others the diagnosis was confirmed by cardiac catheterisation. Twenty patients died during a mean follow-up period of 54.6 $\pm$ 54.47 month. Sudden cardiac deaths (30%), congestive heart failure (25%) and haemoptysis (15%) were the predominant causes of death. Only one patient died during puerperium. The acturial survival for the entire patient population at 5 yrs, 10 yrs and 15 yrs were 86.95%, 79.64% and 76.98% respectively. Level of shunt (atrial, ventricular or aortopulmonary) did not influence the survival ($p > 0.5$). Of all the variables tested in an univariate analysis, history of syncope at presentation ($p < 0.005$), elevated mean right atrial pressure (8mmHg or above) ($p < 0.05$) and systemic arterial desaturation below 85% ($P < 0.05$) were found to be important indicaters of a poor prognosis. Conclusion:- Eisenmenger Syndrome is compatibale with a fair intermediate term survival. History of syncope, elevated right sided filling pressure and systemic arterial oxygen saturation less than 85% indicated a poorer outcome.

Key Words: Eisenmenger Syndrome; Eisenmenger Complex; Heart Septal Defect Atrial; Patent Ductus Arteriosus; Hypertension, Pulmonary.

INTRODUCTION

In 1897, Victor Eisenmenger[1] described the clinical presentation and autopsy findings of a 32 year old man known to be cyanosed and short of breath for many years but working actively as a coachman till the onset of heart failure shortly before death. The autopsy findings showed a large ventricular septal defect with some aortic override, enlargement of right ventricle and atheroma of the pulmonary artery[1]. For the next 50 yrs aortic override was considered an essential part of Eisenmneger syndrome. Bing[2] drew the attention to the fact that pulmonary hypertension and pulmonary vascular obstruction were the determinants of the left to right shunt in ventricular septal defect[2]. Bond[3] corroboarted the same idea and noted a variety of Congenital Cardiac defects that had been reported in association with Eisenmenger Complex[3]. In 1958 Paul Wood[4] expanded the anotamical concept of Eisenmenger syndrome to include any large systemic - pulmonary communication at aortopulmonary, ventricular or atrial level. He defined the condition as pulmonary hypertension at or near systemic level with reversed or bidirectional shunt between the pulmonary and systemic circulation and pulmonary vascular resistance above 800 dynes. sec/cm-5 (10 Wood units). He added that at times the shunt may be balanced so delicately that there may be no net shunt in either direction. He

enlisted no less than 12 anatamical conditions which can result in such a physiology and called the condition Eisenmenger syndrome.

Since Paul Wood's publihsed series of 127 patients, six other large series[5-10] have been reported, each of them having more than 50 patients followed up for many years. In the present study, we have followed up 201 patients of Eisenmenger syndrome over the past 16 yrs in our Institute and anlysed their data to determine their survival pattern, impact of the disease on the life style and complications during long term follow up.

MATERIALS AND METHODS

Diagnosis of Eisenmenger syndrome was established on the basis of the presence of severe pulmonary arterial hypertension and cyanosis and was confirmed by cardiac catheterization or M-mode, 2 diamensional and Doppler echocardiography with or without contrast echocardiography. In 131 patients Cardiac Catheterization was done alongwith oxygen inhalation study with or without tolazoline infusion in a dose of 1mg/kg over 1 minute in the main pulmonary artery. Standard pressure recordings in pulmonary artery and aorta were obtained before and after oxygen inhalaltion and/or tolazoline infusion. 143 patients had echocardiographic study. In some of the patients during the

earlier period only cardiac catheterization study was performed while in the later period some patients had only echocardiographic study including contrast echocardiography along with arterial blood gas study to establish the diagnosis.

Follow-up

Follow up information was collected from patient charts and finally a questionnaire was sent to all patients or next of kin regarding the present survival status Patients who failed to respond to three consecutive questionnaires were considered lost to follow-up.

Statistical Analysis

Results were expressed as percentages for discrete variables, as mean $\pm$ standard deviation for continuous variables. Comparison between pulmonary and systemic blood flow, and systemic and pulmonary vascular resistance before and after O_2 inhalation and/or tolazoline infusion were compared using the students 't' test. The survival pattern was analysed by Life-Table Method. Variables considred important for survival were analysed using LEE-DESU Method. All data were entered into a computer data-base system and statistical analysis was done using SPSS/PC + version 4 software of Advanced Statistical Module.

OBSERVATION AND RESULTS

Out of the total number of 201 cases, 109 were females (54.22%) and 92 were males (44.77%). Age at presentation varied from 3 months to 62 years with a mean age of 19.23 ± 12.62 yrs. The most common aetiologic lesion was ventricular septal defect (VSD) accounting for 33.33% of all cases follwed by Atrial septal defect (ASD) in secundum location (29.85%), patent ductus arteriosus (PDA) (14.43%), double outlet right ventricle with VSD (3.98%), atrial septal defect in primum location (1.49%), complete endocardial cushion defect (1.49%), and aortopulmonary window (1.49%), in the order of frequency. Single atrium and sinus venous atrial septal defect accounted for 2 cases cach, while L-Transpostion of great arteries with VSD and total anomalous pulmonary venous drainage accounted for 1 case each Shunts at 2 levels (varying combination of VSD, ASD and PDA) were seen in 11 cases (5.47%) - out of them combination of VSD and PDA were commonest (2.98%). Aortic stenosis, mitral stenosis and coarctation of aorta complicated a shunt lesion in 3,1 and 3 cases respectively (Table No.1).

Clinical features

Dyspnoea on exertion, palpitation, oedema haemoptysis and syncope were the common presenting symptoms. Three patients (1.5%) presented with brain abscess. Only 1 patient

had a history suggestive of cerebral embolism(0.5%).(Table No.2)

Physical Findings common to all groups included central, cyanosis, clubbing of digits, prominence of jugular venous 'a' wave and occasional increase in mean venous pressure, right ventricular heave, accentuated pulmonic closure sound with a variable degree of splitting of second sound or a single second sound and a soft, short ejection systolic murmur at left sternal edge.

Characteristics of a specific defect were often identifiable. Differential cyanosis provided clue to the presence of ductus while wide and fixed spliting of second sound suggested atrial septal defect. A single second sound suggested ventricular septal defect while normal split of second sound favoured a diagnosis patent ductus arteriosus. Prominent 'a' wave in JVP was equally prevalent in all 3 groups (Table 3). Congestive heart failure was present in 17% of cases at presentation. Out of them 7 patients (20%) had either a combination of shunt lesions or a complicating lesion in the form of aortic as mitral valve disease.

Radiologic features

Cardiomegaly was seen in 67.2% and 98% had a prominant main pulmonary artery segment. Pulmonary vascularity was

diminished in the peripheral lung field in more than two thirds (69.7%) while it was increased in 7.5% and normal in 22.9%. 17.4% had Right atrial enlargement while 6 cases had left atrial enlargement.

Electrocardiographic Features

Almost all patients were in sinus rhythm. Atrial fibrillation was seen in one patient with atrial septal defect and complete heart block was seen in 2 patients, one in each of ventricular septal defect and atrial septal defect. Majority showed a rightward QRS axis. Patients with Ventricular septal defect had a higher incidence of leftward (3.9%) or right superior axis (5.3%) Biventricular enlargement was seen in 27.3% of patients with ventricular septal defect and 12.8% of patents with patent ductus arteriosus compared to 1.7% of patients with atrial septal defect. Both incomplete and complete right bundle branch block (RBBB) were seen more commonly amongst patients with atrial septal defect 23.33% and 18.66% respectively (Table No.4).

Echocardiographic Features

Echocardiography was done 143 cases (71.2%). A sizeable number of patent ductus arteriosus could not be picked up by echocariography. Contrast echocardiography with peripheral venous injection of agitated saline was done in 51 (25.4%)

patients. In all these patients right-to-left shunt at various levels were picked up by this technique. Doppler endocardiogram picked up significant mitral regurgitation in 10 patients, tricuspid regurgitation in 13 patients, pulmonary regurgitation in 13 patients and aortic stenosis in 3 patients and aortic regurgitation in 2 patients.

Haemodynamic Data

Cardiac catheterisation was done in 131 cases to obtain physiologic data. Atrial septal defects, both in secondum and primum location, were most commonly crossed by the catheter (94%). Patent ductus was crossed by catheter in 74% of cases. The defect could be negotialted in only 19% cases of ventricular septal defect.

Degree of arterial desaturation was similar in all patient groups except in cases with d-Transposition of great arteries (mean 61.5 $\pm$ 3.53). Left-to-right component of bidirectional shunt caused a mean oxygen step-up in the shunting chamber by 8% in atrial septal defect, 7% in ventricular septal defect and 5% in patent ductus arteriosus - total 76.74% of patients had this step-up.

Mean right atrial pressure was normal in all the three major groups while mean left heart filling pressure was higher in cases of patent ductus arteriosus only (14.00 $\pm$ 8.46 mmHg). The Mean of Pulmonary artery mean pressure for

the whole group was 68.4 ± 15.4 mmHg while mean aortic pressure was 80.63 ± 13.41 mmHg. Mean left to right shunt was 2.00 ± 1.83 litres per minute while mean right-to-left shunt was 1.34 ± 1.90 litres per minute. Mean pulmonary vascular resistance was 16.67 ± 10.96 units, while the mean pulmonary vascular resistance index (calculated only for children with body surface area less than 1 square metre) was 12.71 ± 6.026 units.

Oxygen inhalation and/or tolazoline infusion in pulmonary artery did not bring about any significant change in shunt in either direction or vascular resistance(Table 5).

Angiographic Features

Angiography was performed in 61 cases. Twenty-Eight had large single perimembraneous VSD. 5 patients had large single or multiple muscular VSDS, 1 patient had doubly commited VSD and 1 had inlet VSD. VSD was associated with a mildly obstructive subaortic membrane in 1 patient and with coarctation of aorta in 1 patient.

Patent ductus arteriosus was visualised on angio in 4 patients in whom the defect could not be crossed Aortopulmonary window (AOW) was visualised in 3 patients, Double-outlet right ventricule with VSD in 5 patients

(subaortic in 4 and subpulmonic in 1) Single ventricle in 3 patients, primum ASD with cleft anterior mitral leaflet (AML) and mitral regurgitation(MR) in 2 patients, single atrium with cleft AML and MR in 1 patient, d-transposition of great areries, VSD in one patient, Truncus Arterious and total anomalous pulmonary venous connection into Right Atrium in 1 patient each. One patient had proximal APW, PDA with interruption of aortic arch.

Follow-up Mortality Acturial Survival and Variable affecting Survival

The patients were followed up for a mean duration of 54.67 ± 54.47 months (range 1 to 193 months). 20 patients died during this follow-up period. Mean age at death was 25.25 ± 11.67 yrs (range, 4 to 55 yrs). 8 patients with atrial septal defect, 8 patients with ventricular septal defect, 2 patients with patent ductus arteriosus, one patient with proximal aortopulmonary window, patent ductus arteriosus and interruption of aortic arch (patient died after cardiac catheterization) and one patient with both patent ductus arteriosus and atrial septal defect died (Table 6). Mean age at death for VSD, ASD and PDA were 20.37 ± 5.95 yrs, 26.71 ± 6.65 yrs and 27 ± 21.49 yrs respectively - the mean age of death among three group was not significantly different ($p > 0.1$).

Survival patterns

For the entire patient population the acturial survival at the end of 1 yr, 5 yrs, 10 yrs and 15 yrs were 97.18%, 86.95%, 79.64% and 76.98% respectively (Figure No.1).

For the patients with pretricuspid level shunts the acturial survival at the end of 1 yr, 5 yrs, 10 yrs, and 15 yrs were 97.8%, 79.78%, 72.53% and 72.53% respectively. For patients with ventricular level shunt the values were 96.8%, 91.08%, 82.53% and 82.53% respectively. For patients with aortopulmonary level shunt, the values were 97.18%, 87.93%, 87.93% and 43.96% respectively (Figure No.2). However using the LEE-DESU survival analysis the survival patterns of three groups were not significantly different at any state ($p > 0.5$) (Figure No.3).

Several varaibles were tested for their impact on survival by LEE-DESU statistical survival curve analysis. The variables which were found to affect the prognosis adversely were syncope at presentation. ($P < 0.005$) elevated mean rigert atrial pressure (8 mmHg on above) ($p< 0.05$) and systemic arterial saturation below 85% ($p 0< 0.05$).

Other variables tested and found to be nonsignificnt were sex ($p > 0.5$) presence of multiple shunts or complication of lesions ($p>0.1$) angina ($p>0.05$) degree of effort intolerance ($p >0.1$) elevated mean jugular venous

pressure (p>0.05), presence of tricuspid regurgitation (p >0.1), presence of pulmonary regurgitation (p>0.1) cardiomegaly on chest X-ray (P > 0.1). Presence of cyanosis (p>0.1). QRS-Axis on electrocardiogram (p >0.1), severity of pulmonary arterial hypertension (P >0.1) left ventricular end-diastolic pressure (p > 0.1) degree of right-to-left shunt (p > 0.1) basal pulmonary vascular resistance (P > 0.1) pulmonary vascular resistence after oxygen inhalation or tolazoline infusion (P>0.1)

Out of the patients who were alive on follow-up, nearly 70% were in New York Heart Association Class II or above, 17% were in persistent congestive heart failure, 17% were having recurrent haemoptysis, 3% were having anginal chest pain and 2% experienced syncope, 5% had brain abscess during the period of follow-up and survived it. 1 patient had a history sudden monooccular blindness - probably due to retinal artery embolism. 26 pregnancies occured in 12 patients - 9 patients of ASD, 2 patients with VSD and 1 patient with PDA - 2 pregnancies were terminated medically, 1 resulted in still-birth. 22 pregnancies culminated unevenfully into live-births. Maternal death occured in 1 case of ventricular septal defect during puerperium.

DISCUSSION

In 1958, Paul Wood[4] expanded the anatomic concept of

Eisenmenger complex that is VSD with cyanosis and evidence of pulmonary vascular disease, to the physiologic concept of Eisenmenger Syndrome. This was defined as, pulmonary hypertension at systemic level due to high pulmonary vascular resistance (over 10 units) with a reversed or bidirectional shunt at aortopulmonary, ventricular or atrial level.

Our series showed in overall female preponderance (54.23% vs 45.77%) which is similar to the incidence reported by Brammell[6] et al but contrasts with an almost equal sex incidence reported by Wood[4] and a male dominance reported by Abraham et al[9].

Although Wood[4] reported that PDA with Eisenmenger reaction is most well tolerated amongst all causes of Eisenmenger Syndrome, our series showed in almost equal prevalance of dyspnoea and effort intolerance in all 3 major subgroups —96.1% in VSD, 98.3% in ASD and 97.4% in PDA. Patients with ASD presented at a later age compared to VSD or PDA (18.7 yrs vs 3.36 and 6.39 yrs),— similar to what has been reported from India[9,10] Incidence of Haemoptysis (16.9%) was similar to earlier reports[5,7,8,9], but at variance with the higher incidence observed Brammell et al[6] and Wood[4]. Squatting was found in 7% of patients in contrast to 16% of patients in Wood's series[4]. In our

series syncope was found in 7.5% of cases while its incidence varied from 1.5% to 18% in other reported series[6,7,10]. There was no major difference between Ventricular Septal Defect (VSD) patent ductus arterioses (PDA) and atrial septal defect in the occurance of syncope (9.1%; 5.0%, 5.0%) A history suggestive of congestive heart failure was found in 17.4% of cases which was no different from the other series. Thus except a later age of onset of symptoms in cases with pretricuspid level shunt, history was unable to provide any clue to the level of shunt.

Studies by Wood[4] have emphasized the physical signs useful in deciding the level of shunt reversal. In our series differential cyanosis was found in patients with patent ductus arteriosus in less than 50% of cases while it was about the same in the series reported by Wood et al[4]. Prominent 'a' wave was almost equally common in all 3 groups (31 to 41%) and was of no help indeciding the level of shunt. Splitting of the 2nd heart sound helped in this aspect, majority of patients with ASD had wide, fixed splitting, with VSD had single second sound while those with PDA had a normal inspiratory splitting. Tricuspid regurgitation was common in patients with atrial septal defect (ASD) as compared to VSD and PDA (25% vs, 10.3% and 11.7%) while pulmonary regurgitation, was less common in ASD

than the other 2 groups (13.3% vs 24.7% and 25.6%).

Chest Skiagrams also were not of much help in providing clue to the level of shunt except higher incidence of cardiomegaly in ASD patients (81.7% vs 57.1% in VSD and 53.8% in PDA) and pruning was seen in marginaly higher proportion in ASD patients (86.6% in ASD vs, 66.2% in VSD and 59.0% in PDA). Most patients did not have any atrial enlargement. No case with a right sided aortic arch was found amongst VSD patients, (compared to 16% in Wood's series)[4]

Amongst electrocardiographic features, presence of biventricular hypertrophy was rare in ASD patinets (1.7%) - that particularly patient had associated mitral regurgitation. incomplete or complete right bundle branch block was more common in ASD (43.66%) than VSD (10.3%) or PDA (5.3%) Rythm disturbances were conspicous by their near absence in the present series in contradistinction to a 20% incidence of various atrial tachyarrhythmias noted in Paul Wood's series[4].

One unique features of the present series is that 71.2% of patients underwent echocardiography and 25.4% had a documentation of right-to-left shunt by contrast echocardiography. We did not encounter any comparable series reporting on echocardiographic aspects of Eisenmenger

syndrome. In fact in a sizeable section of patients the diagnosis was based on echocardiographic demonstration of right-to-left shunt along with doppler estimation of right ventricular pressure, pulmonary artery diastolic pressure, and documentation of desturation in systemic arterial blood gas analysis, thus avoiding cardiac catheterization study which carries significant risk.

Cardiac catheterisation was done in 65% of cases while angiocardiography done in 30.5%. Angiography was avoided whenever the anatomy was delineated by 2D-echo and the defect was crossed by catheter. ASD was the lesion most easily crossed (94%) followed by PDA (71%) and VSD (19%)-the order was different in Wood's series - PDA (90%), ASD (66%) and VSD (28%)[4] There was a mean step-up of oxygen saturation around 8% in cases of ASD, 7% in case of VSD and 5% in case of PDA due to the left to right component of bidirectional shunt.

Reactivity of pulmonary vascular bed was assessed by oxygen inhalation and/or tolazoline infusion in the main pulmonary artery and there was no significant difference in pulmonary blood flow, systemic blood flow, shunt in either direction, pulmonary or systemic vascular resistance, before and after Oxygen inhalation/tolazoline infusion.

The commonest cause of death in the present series was sudden cardiac death accounting for nearly one-third (30%)- this is presumably due to ventricular arrythmia[7]. This accounted for nearly half of the deaths in Young's series[7] and 14% in Wood's series[4]. Other causes were similar to earlier reported series[4,6,7] - namely congestive heart failure (25%), haemoptysis (15%), brain absces (10%), pregnancy (5%) and cardiac catheterization (5%). No patient died of surgery - cardiac or non-cardiac, in contrast to earlier series[4,6,7]-probably becauses surgey as a preventable cause of death was well established by the time our patients were first seen and counselled. Similarly there were fewer pregnancy related deaths which is also attributable to the counselling they received against it.

The acturial survival curve showed 5 yrs., 10 yrs, and 15 yrs. survival as 86.95%, 79.64% and 76.98% respectively. No statistically significant difference was found between the 3 groups in the acturial survival pattern. Many of our patients were leading active lives - pursuing studies, attending gainful employments, doing household chores, etc. However, the mean age at death was 25.25 $\pm$ 4.67 yr. which is infact lower than the mean age observed by Wood[4]. Syncope was found to be an adverse prognostic variable. Probably syncope signifies more advanced pulmonary vascular obstructive disease with consequent inability to raise

cardiac output in the face of an increasing demand and more right-to-left shunting. Other two adverse prognostic variables were elevated mean right atrial pressure (above 8mmHg) signifying congestic heart failure and degree of arterial desaturation which is again dependent on pulmonary vascular obstructive disease. Hence the degree of pulmonary vascular obstructive disease was the main determinant of downhill course, although the evidence for this is indirect. Haemoptysis was not found to be associated with an adverse prognosis unlike its grave prognostic implication observed in Woods series. The grade 5 dilatation lesions of pulmonary vascular obstructive disease (Heath & Edward's classification)[11] has been shown to be the cause of repeated haemoptysis[12]. Another factor responsible for haemoptysis is pulmonary infarction. Therefore, haemoptysis may not always indicate severe pulmonary vascular obstructive disease, Clarkson et al[5] reported a 5 yrs. survival rate of 80% which is similar to our observations. They showed probability of survival was 95% for patients aged 10 through 19 yrs and 56% for those 20 yrs or older. No other long term follow-up study showing acturial survival has been published in English Language Literature.

The fact that Pregnancy was relatively well tolerated (26 pregnancies in 12 patients with 2 medical termination. 1

still birth and 1 maternal death during peurperium) may be ascribale to the fact many of them bore children before developing Eisenmenrger syndrome. The maternal mortality in other reported series is 27%[13]. The period of greatest hazard appears to be during parturition and the first 2 weeks. after delivery[8], the only patient in our series died in the 2nd post partum week.

CONCLUSION

Eisenmenger Syndrome, although implies a hopeless situation, is compatible with a fair intermediate term survival. History of syncope at presentation, elevated right sided filling pressure at presentation, and systemic arterial desaturation (below 85%) indicate poor prognosis.

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Table 1

DISTRIBUTION OF VARIOUS DEFECTS IN 201 PATIENTS OF EISENMENGER SYNDROME

Diagnosis	Frequency	Percentage	Male	Female
1. VSD	67	33.33	35	32
2. ASD	60	29.85	30	30
3. PDA	29	14.43	13	16
4. VSD+ASD	4	1.99	1	3
5. VSD+PDA	6	2.98	4	2
6. ASD+PDA	1	0.49	1	0
7. Shunt lesion & Aortic Stenosis.	3	1.49	0	3
8. Shunt lesion + Mitral Stenosis	1	0.49	0	1
9. Shunt lesion+coartation of aorta	3	1.49	0	3
10.DORV, VSD, PAH	8	3.98	1	7
11.d-TGA VSD	3	1.49	1	2
12.Primum ASD	4	1.99	2	2
13.Complete endocardial cushion defect.	3	1.49	1	2
14.Single atrium	2	0.98	1	1
15.Sinus Venous ASD	2	0.98	1	1
16.Aortopulmonary window	3	1.49	1	1
17.L-TGA.VSD	1	0.49	0	1
18.TAPVC	1	0.49	0	1
TOTAL	201	100.00	92	109

Abbreviation used:

[ASD = Atrial Septal Defect; DORV= Double outlet right ventricle; d-TGA = Dextro transposition of great arteries; L-TGA= Levo transposition of great arteries. PAH= Pulmonary arterial hypertension; PDA = Patent ductus arteriosus, TAPVC = Total anomalous pulmonary venous connection]. VSD = Ventricular Septal Defect;

Table 2.

FREQUENCY AND PERCENTAGE DISTRIBUTION OF SYMPTOM IN EISENMENGER SYNDROME

Symptoms	Frequence	Percentage(%)
1. Dyspnoe/Effort intolerance	196	97.5
2. Palpitation	160	79.6
3. Oedema	27	13.4
4. Haemoptysis	34	16.9
5. Syncope	15	7.5
6. Squatting	14	7.0
7. Angina	9	4.5

Table 3.

PERCENTAGE DISTRIBUTION OF PHYSICAL SIGNS IN EISENMENGER SYNDROME

Signs	Entire	VSD	ASD	PDA
1. Uniform Cyanosis	57.2	63.6	61.7	38.5
2. Differential Cyanosis	8.5	0	0	43.6
3. Clubbing	61.2	59.7	58.3	69.2
4. Prominent 'a' wave in JVP	38.0	31.23	40.00	41.00
5. Elevated mean jugular venous pressure	17.4	25.0	15.0	7.0
6. Right ventricular heave	78.7	70.1	81.6	72.3
7. Spliting of 2nd Heart sound				
a. Wide and fixed	26.4	5.2	68.3	7.7
b. Single	34.3	54.5	6.7	23.1
c. Close split	18.9	27.3	16.7	12.8
d. Normal split	20.4	13.6	8.3	56.4
8. Constant Ejection click in Pulmonary area	78.1	79.1	93.3	79.3
9. Tricuspid regurgitation	15.4	11.7	25.0	10.3
10. Pulmonary regurgitation	22.4	24.7	13.3	25.6

[Abbreviations used ASD = Arterial septal defect;
JVP = Jugular Venous Pulse]
PDA = Patient ductus arterioses
VSD = Ventricular septal defect;

Table 4.

PERCENTAGE DISTRIBUTION OF ELECTROCARDIOGRAPHIC FEATURES OF EISENMENGER SYNDROME

	Ventricular septal defect	Atrial sepetal defect	Patent ductus arteriosus
a) RYTHM			
Sinus	98.7	96.6	100
Atrial fibrillation	0	1.66	0
Complete heart block	1.29	1.66	0
b) QRS-axis			
Right axis	61.8	74.6	67.6
Left axis	3.9	1.7	0
Normal	21.1	18.6	18.9
Extreme right or left	5.3	3.4	5.4
Indeterminate	7.9	1.7	8.1
c) Ventricular enlargement			
Right ventricular hypertrophy	62.3	85.0	74.4
Left ventricular hypertrophy	3.9	0	2.6
Biventricular hyper-trophy	27.3	1.7	12.8
None	6.5	13.3	10.3
d) Right bundle branch block			
Incomplete Right Bundle Block	9.0	23.33	0
Complete	1.3	18.33	5.12

Table 5.

HAEMODYNAMIC DATA IN EISENMENGE SYNDROME
(For the Entire Patient Population)

. Ascending Aortic Saturation	85.4 ± 8.9%	
. Mean Right Atrial Pressure	6.95 ± 3.02 mmHg	
. Mean Pulmonary Arterial Pressure	68.04 ±15.8 mmHg	
. Mean Pulmonary artery wedge presence	11.33 ± 5.74 mmHg	
. Mean Aortic Pressure	80.63 ±13.41 mmHg	
. Left to Rigert Shunt		
Basal	1.14 ± 0.9 L/min	(p >0.1
After oxygen inhalation/tolazoline infusion	1.18 ± 0.93 L/min	Not Significant)
. Right to left shunt b		
Basal	0.78 ± 0.62 L/min	(p >0.1
After oxygen inhalation/tolazoline infusion	0.54 ± 0.62 L/min	Not Significant)
. Pulmonary vascular resistance		
Basal	28.98 ±11.24 wood units	(p >0.5
After oxygen inhalation/tolazoline infusion	17.09 ±12.05 wood units	Not Significant
Systemic vascular resistance		
Basal	22.6 ± 6.37 wood units	(p > 0.941
After oxygen inhalation/tolazoline infusion	22.8 ±11.99 wood units	Not Significant

Values expressed as mean ± standard deviation.

Table 6

CAUSE OF DEATH IN FISENMENGER SYNDROME

Cause	Frequency	Percentage
Sudden Cardiac death	6	30
Congestive heart failure	5	25
Haemoptysis	3	15
Brain abscess	2	10
Unspecified	2	10
Pregnancy related	1	5
Cardiac catheterization	1	5

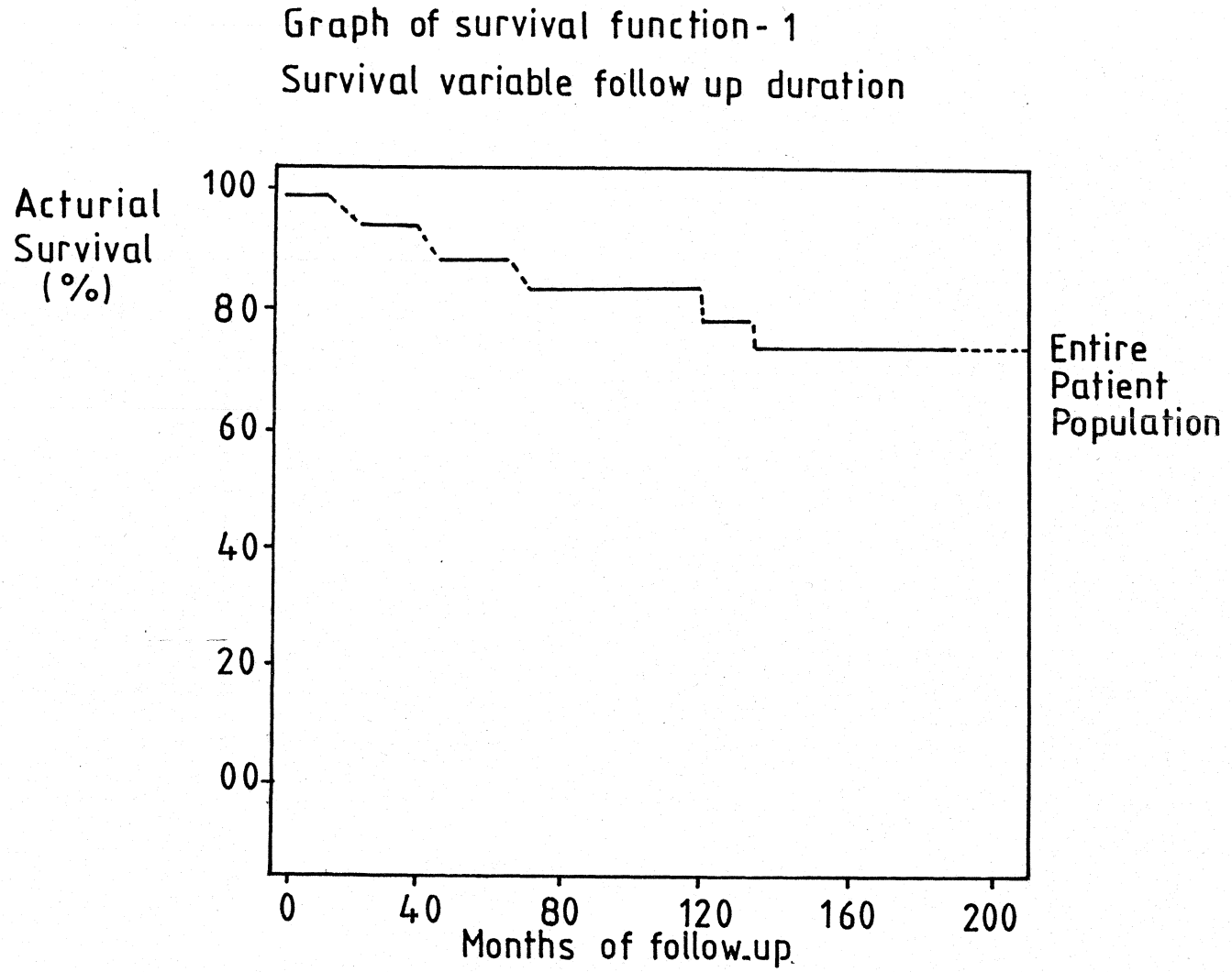

Fig. 1: Survival curve (constructed by to Life-Table Method) of the entire patient population with Eisenmenger Syndrome.

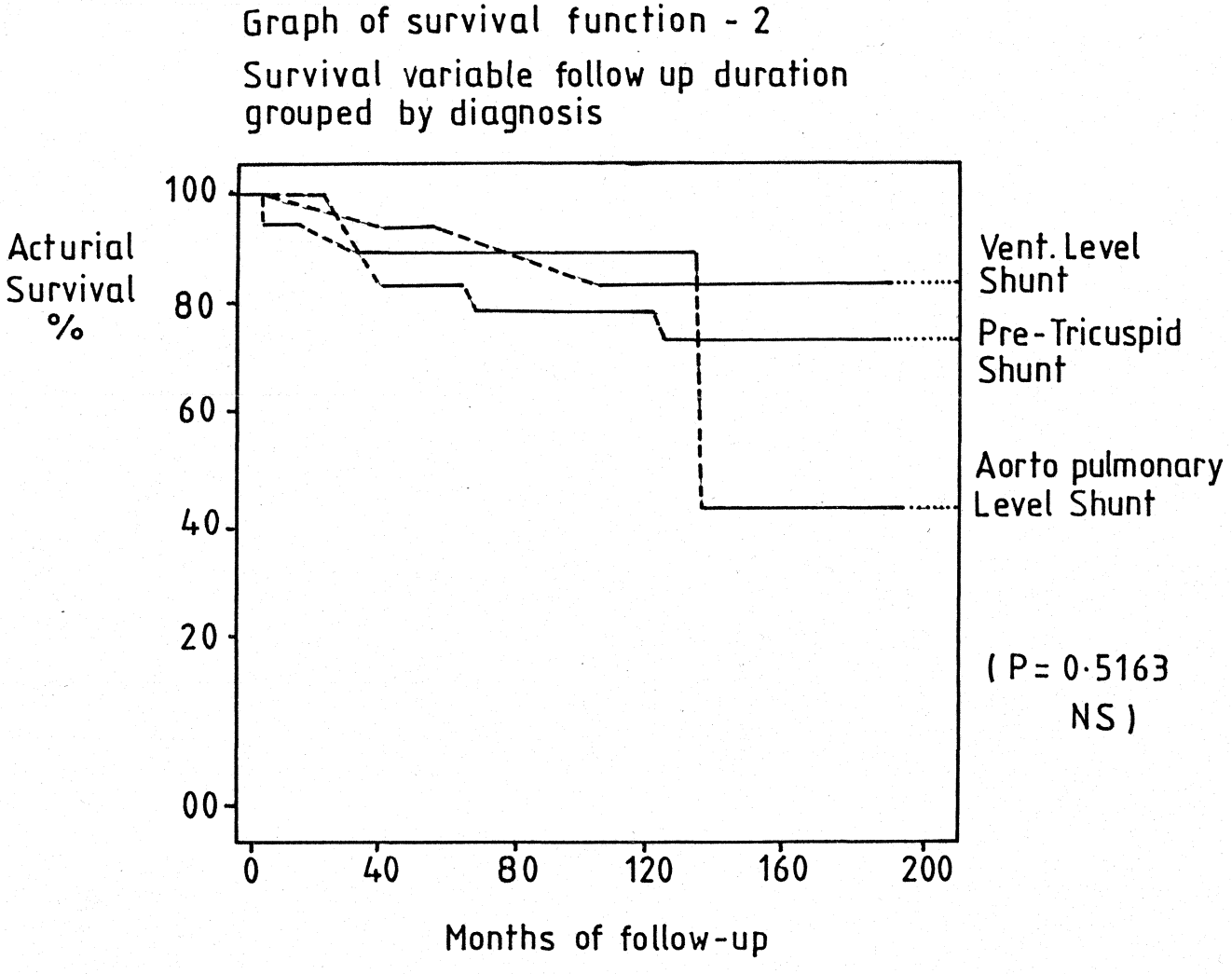

Fig.2: Survival curve (constructed by Life-Table method) of three main groups of patients with Eisenmenger Syndrome showing no difference in survival.
(NS - no significant; Vent. = Ventricular.)