Original Article

Eisenmenger Syndrome: A silent Killer in Adults born with Heart Septal defect: Experience from Cardiac Centre at Faisalabad

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ABSTRACT

Introduction: Eisenmenger syndrome is a term used to any large shunt between systemic and pulmonary circulation which results in high pulmonary arterial pressure and irreversible changes in pulmonary vascular bed with bidirectional shunt with physical limitation and shortness of breath.

Eisenmenger syndrome particularly creates problems to fetus and mother in pregnancy and there is a particularly risk during aneasthesia while performing general surgery.

Materials and Methods: We collected all consecutive patients above age 12 with atrial septal defect (ASD), ventricular septal defect (VSD) and patent ductus arteriosus (PDA) who attended echocardiography department between June 2008 to October 2010.we also analysed all pregnant females with eisenmenger complex during this period.

Results: Out of 309 patients diagnosis of one of three shunts was confirmed either by transthorasic echocardiography with intravenous saline infusion or transesophageal echocardiography

Eisenmenger syndrome was diagnosed in 39 patients (19 patients with ASD, 11 patients with VSD and 09 patients with PDA).

All 39 patients were followed till October 2010 and were alive.

Among 03 pregnant females, 02 completed pregnancy without any hazard to child and mother. However tubal ligation was opted at time of delivery. One lady opted abortion and ligation to prevent further pregnancy.

05 patients underwent non cardiac surgery under general anesthesia without any complication.

Conclusion: Eisenmenger syndrome a silent killer in a congenital treatable disease which is being neglected and diagnosis is being delayed. It seems eisenmenger syndrome is on rise in Pakistan.

We need to establish adult congenital heart disease department in each cardiac centre where trained persons should be appointed who had experience of congenital heart disease. Screening clinics need to be established at school and community level to diagnose this silent killer at a stage when pulmonary artery pressure is still reversible.

Key Words: Eisenmenger syndrome ES, Atrial septal defect (ASD), Ventricular septal defect (VSD) and patent ductus arteriosus (PDA).

INTRODUCTION

Eisenmenger syndrome (ES) is a term applied to any large communication between systemic and pulmonary circulation that results in irreversible changes in pulmonary vascular bed with reversal of shunt. There are growing number of adults with congenital heart disease presenting with progressive decline of cardio-respiratory function and other complications e.g, eisenmenger syndrome, hemoptysis and cyanotic heart disease[1].

A large group of patients can be diagnosed in a stage where cure is possible and ES can be prevented. This needs more congenital heart disease clinics and screening programmes at community level especially in schools.

In most of patients with ES cure is not possible and patients gradually deteriorate with functional limitation and finally death in most of these patients. This is reason we called it a silent killer in a treatable disease.

This disease may have particularly serious consequences when dealing pregnancy and non cardiac surgery[2].

In following lines we are presenting our experience of Eisenmenger complex at Faisalabad Institute of

Cardiology Pakistan .We are especially presenting our experience of 2 successful pregnancies with ES.

MATERIALS AND METHODS

From June 2008 to October 2010, 3680 consecutive patients who attended Department of Echocardiography at Faisalabad Institute of Cardiology were included in this study. All these patients were above the age of 12 years.

The 2-dimentional, M-mode and color Doppler images were obtained using VIVID 7 dimensional GE machine. Transesophageal echocardiography was performed by a consultant cardiologist on same machine with multiplane transesophageal probe.

Pulmonary artery systolic pressure (PASP) was estimated by measuring tricuspid valve regurgitant jet velocity by continuous wave Doppler ultrasound. PASP was also analysed by pulmonary regurgitant flow.

Patients were categorized to have mild pulmonary hypertension (PASP 31-40 mmHg), moderate PH (PASP 41-55 mmHg) or severe PH (PASP >55mmHG) and eisenmenger complex (PASP >100 mmHg and irreversible on vasodilator therapy) .In our study vasodilator therapy used was oxygen therapy and nitroglycerin therapy either sublingually or intravenously. Patients with clubbing and cyanosis with PASP >100 mmHg were declared eisenmenger syndrome without any further investigation.

Female patients in child bearing age were followed for pregnancy and were managed until delivery with help of obstetrician at Allied Hospital, Faisalabad.

Patients who required general anesthetist for non cardiac surgery were managed with help of consultant anesthetist and general surgeon.

RESULTS

Total patients above age 12 attended Echocardiography Department of Faisalabad Institute of Cardiology from June 2008 to October 2010 were 3630. Out of these patients with diagnosis of ASD, VSD and PDA were 309 (>9% of all patients in echocardiography Lab.). Patients with eisenmenger syndrome with PASP >100mmHG and non responsive to vasodilator therapy were 39 patients(11%).

Patient with diagnosis of ASD were 206 and ES were 19 (9%).

VSD patients were 62 and ES were 11(18%).

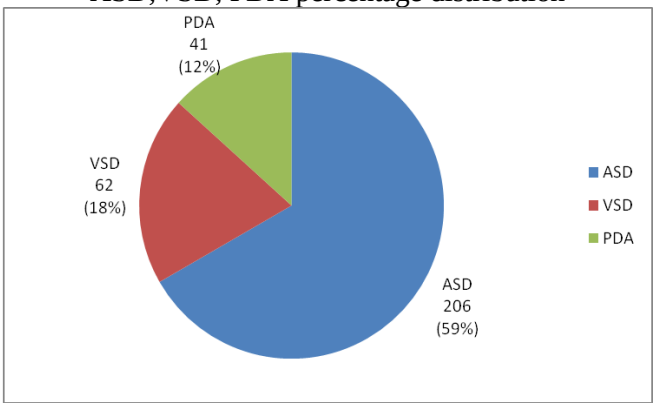
Patients of PDA were 41 and eisenmenger complex were 09(22%).

Female patients with Eisenmenger complex 23 and male were 16(1.2:1.0)

Mean age of first diagnosis of Eisenmenger complex was 24 years (range 12 -62 years).

n=309

ASD,VSD, PDA percentage distribution



Presentation at diagnosis of eisenmenger syndrome: Shortness of breath 39 patients (100%),Palpitation 39(100%),chest pain 24(61%),cyanosis and clubbing 09 (23%),evidence of severe pulmonary hypertension with loud P2 39(100%), life threatening haemoptysis in 2 patients and systolic murmur in all 39 patients with ES (Tab-1).

Tab-1:
Clinical Presentation in Eisenmenger Syndrome

n=39		
Shortness of Breath	39	100%
Palpitation	39	100%
Chest Pain	24	61%
Cyanosis & Clubbing	09	23%
Loud P2	39	100%
Haemoptysis	02	03%

Investigation results in patients with eisenmenger syndrome: Electrocardiogram (ECG): Right bundle branch block 11(28%) atrial fibrillation in 06 (15%) Right ventricular hypertrophy in 28(71%).

X-Ray chest showing evidence of pulmonary hypertension in form of prominent pulmonary artery and pruning of peripheral vessels in almost all patients.

Diagnosis was confirmed in all 39 patients with Eisenmenger complex either with transthorasic echocardiography alone with or without saline contrast infusion or with transesophageal echocardiography. In all 309 patients with ASD,VSD and PDA diagnosis

was established with transthorasic echocardiography. In 108 patients additional help of transesophageal echocardiography was required. Intravenous saline contrast was used in all 309 patients for confirmation of diagnosis.

Management in patients with Eisenmenger Syndrome: All 39 patients were treated medically without any effort to treat by device or surgery. 21 patients received sildenafil long term therapy with symptomatic treatment. It was not possible to prescribe oral Bosentan or intravenous prostaglandin in our setup. Cardiac transplantation was not possible in our circumstances.

Management of patients with eisenmenger syndrome and pregnancy: We managed 3 pregnant females during our study period. Two patients opted to continue pregnancy in-spite of full explanation of hazards. Management was performed by combined team including cardiologist, anaesthetist and obstetrician. Both these patients completed pregnancy without any complication to fetus and mother. One patients had full term normal delivery and other had c - section at 34th week without complication. One patient agreed for abortion and tubal ligation.

Management of patients with eisenmenger syndrome with non cardiac surgery under general anaesthesia: Five patients underwent surgical procedures (non cardiac) under general anesthesia. Medical team including cardiologist, anaesthetist helped the Surgical team with successful outcome of these procedures without any complication.

DISCUSSION

In 1897 Vicktor Eisenmenger described a patient with cyanosis and dyspnoea since infancy who died by massive hemoptysis at 32 years of age. Postmortem showed a large ventricular septal defect and severe pulmonary vascular disease[1].

In 1958 term Eisenmenger was used BY Paul Wood when he defined eisenmenger syndrome as a state in a septal defect when pulmonary pressure equalized systemic pressure with reversal of shunt[1].

Pulmonary vascular changes appear in childhood in normal population and these changes are progressive. Any communication between systemic and pulmonary circulation results in the development of pulmonary vascular disease. Release of vascular elastase in pulmonary vascular bed has been shown to induce other mediators resulting in smooth muscle migration, and hypertrophy with stimulation of elastin and collagen synthesis. These steps are critical in the formation of irreversible pulmonary vascular disease[3].

In Eurosurvey about congenital heart disease there were 1877 patients with ASD and VSD, only 15 patients had developed ES (0.88%). In our series 12% had eisenmenger syndrome which reflects delayed diagnosis in our population[4].

Natural history of patients with ES is quite different than with patients with primary pulmonary hypertension. Despite high pulmonary artery pressure in ES than primary pulmonary hypertension course is more favourable. In one study comparing 2 population groups acturial survival was 77% at 3 year in ES as compared 35% in patients with PPH. In our group follow up was 2 year without any mortality. In another paper out of 201 patients with ES 80% survived at 10 years and 77% at age 15 years after initial diagnosis. Risk factor for poor prognosis includes syncope, old age at presentation, poor functional class, complex underlying disease, supraventricular arrhythmias including artial fibrillation, oxygen saturation below 85%, increased serum creatinine, RV dysfunction, increased uric acid and presence of clubbing and cyanosis[2,5].

About diagnosis echocardiography is still gold standard. Both transthorasic and transerophageal echocardiography not only diagnose ASD, VSD and PDA but also measure pulmonary artery pressure.

Reversibility of pulmonary artery pressure can be measured by Doppler technique. In our series most of the pulmonary artery pressure was measured by tricuspid regurgitation Doppler technique[6].

Management of ES is descriptively simple and most of management is preventive in alleviating symptoms and treat systemic hypertension. Role of vasodilators like Bosentan (oral endothelin antagonist), Prostaglandins and Sildenafil is promising. There are few reports in which pulmonary pressure declined after oral sildenafil use and in one case report patient was operated for ASD closure. Results of reduction of pulmonary artery pressure by Bosentan are promising. Role of cardiac transplantation in ES is controversial as patients with ES had low mortality and should be considered a choice in those whose predicted mortality is less than 2 years[7,8,9,10].

Two most important situations with ES are pregnancy and non cardiac surgery.

During pregnancy high mortality figures for mother and child are reported literature some authorities recommends abortion and tubal ligation at early stage.

In our study two patients completed pregnancy without any risk. May be more research is required to decide about risk to infant and mother and to define clear indications for abortion and tubal ligations[11].

About non cardiac surgery and ES one needs particular care while performing non cardiac surgery under GA this includes team work between physician, anaesthetist and surgeon and surgery in intensive care unit setup[12].

CONCLUSION

Eisenmenger syndrome is quite high in Pakistan (about 12% in our study).This is alarming and needs good efforts to diagnose congenital heart disease at earlier stage before eisenmenger syndrome develops.

This may include school screening programs and development of adult congenital heart disease clinics in already existing cardiac centers.

REFERENCES

1. Woods P. Eisenmenger syndrome with pulmonary hypertension with central reversal shunt BMJ 1958:701-709.
2. Benello B, Renaod S, Dragnlesen et al. Eisenmenger complex epidemiology and natural history press Med 2009:21-23.
3. Levy M. Eisenmenger syndrome, Pathophysiology press Med 2009; 38: 193-199.
4. Daliento L, Somerville J et al. Eisenmenger syndrome, factors relating to deteriation and death. Euro Heart J 1998;19:1845-1855.
5. Hopkins WE ,Ochoea LL et al. Comparison of the hemodynamics and survival of adults with severe primary pulmonary hypertension or eisenmenger syndrome, Heart lung transplantation 1996 ;15:1000-1055.
6. Lim ZS, Vett JJ et al. Role of echocardiography in the assessment of adults with congenital heart disease. Minerva Cardio angiol 2009; 57:389-413.
7. Duluc Y. Eisenmenger syndrome conventional treatment and general management press Med 2009; 38: 147.
8. Apostopoulou SC, Magninas A, Cockkinos DV et al. Effect of oral endothelin antagonist bosentan on the clinical, exercise and hemodynamic status of patients with pulmonary arterial hypertension related to congenital heart disease. Heart 2005; 91:1447-1452.
9. Norgurd G, Berg A, Gat Zoulis MA et al. Diagnosis and Management of adult congenital heart disease Edinburg: Chur chill livingesto 2003: 413-421.
10. Gatzoulis MA, Rogers P et al. Safety and tolerability of Bosantan in adults with eisenmenger syndrome Int J Cardiol 2005;98:147-151.
11. Avila W, Bringerg M et al. Maternal and fetal outcome in pregnant women with eisenmenger syndrome. Eur Heart J 1995; 16: 460-464.
12. Ammash NM, Connolly HM, Abad MD et al. Non-cardiac surgery in eisenmenger syndrome J Am Coll Cardiol 1999; 33:222-227.

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