



## A Case Report Heart Block in Patient in a Patient with Eisenmenger Syndrome

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### Abstract

*Eisenmenger syndrome is a condition in which the left to right shunt lesions of the heart gets reversed to the right to left due to development of severe pulmonary hypertension. Arrhythmias occur in Eisenmenger syndrome and often these arrhythmias leads to further deterioration of these patients and sudden cardiac death. There are some case reports of Atrio-Ventricular (AV) block occurring in Eisenmenger syndrome and here we present a 57 year old male patient with Patent ductus arteriosus with reversal of shunt with second degree (Mobitz type I) AV block with transient complete AV block.*

### Introduction

Rhythm abnormalities have been of concern in patients with congenital heart disease (CHD) in the adulthood. Though the proportion of patients with arrhythmias in CHD is less when compared to coronary artery disease, often children and young adults with CHD and arrhythmias have poor quality of life and sudden cardiac death which has a psychological impact on the family. In developing countries still the people with congenital heart diseases are diagnosed late with complications and inoperability resulting in poor quality of life. Rhythm abnormalities may be intrinsic as in Ebstein's anomaly or acquired due to increase volume and pressure overloads for longer duration and surgical scars after repair had

lead to arrhythmias and conduction blocks. Treatment of such rhythm abnormalities is also technically difficult.

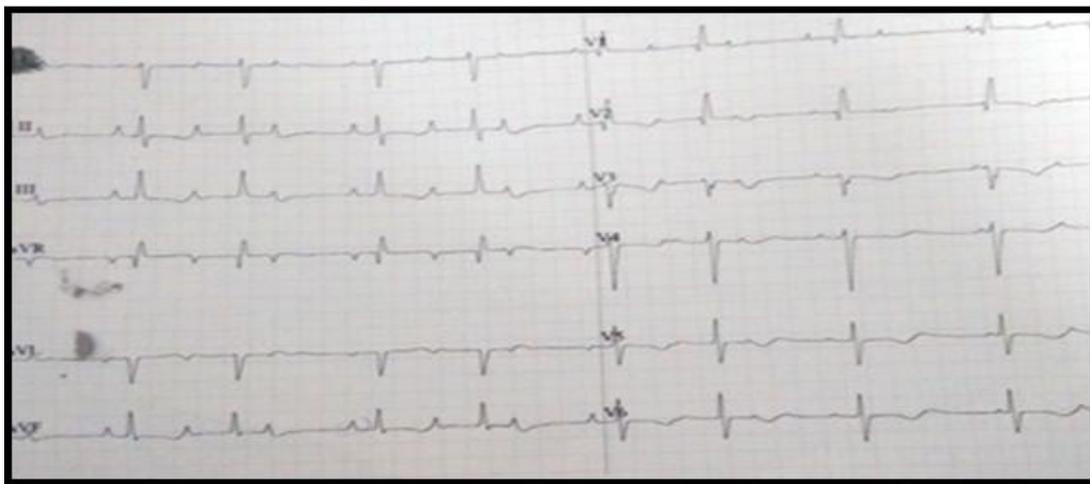
### Case Report

A 57 year old male patient presented with complaints of breathlessness even at rest for 15 days duration. He had breathlessness on exertion since childhood increased for the past 15 days. He also noticed swelling of feet and scrotum and abdominal distension. Patient was diagnosed to have a congenital heart disease ten years back and magnetic resonance imaging of the chest was done and diagnosed to have Patent ductus arteriosus with pulmonary hypertension and reversal of shunt and started on sildenafil.

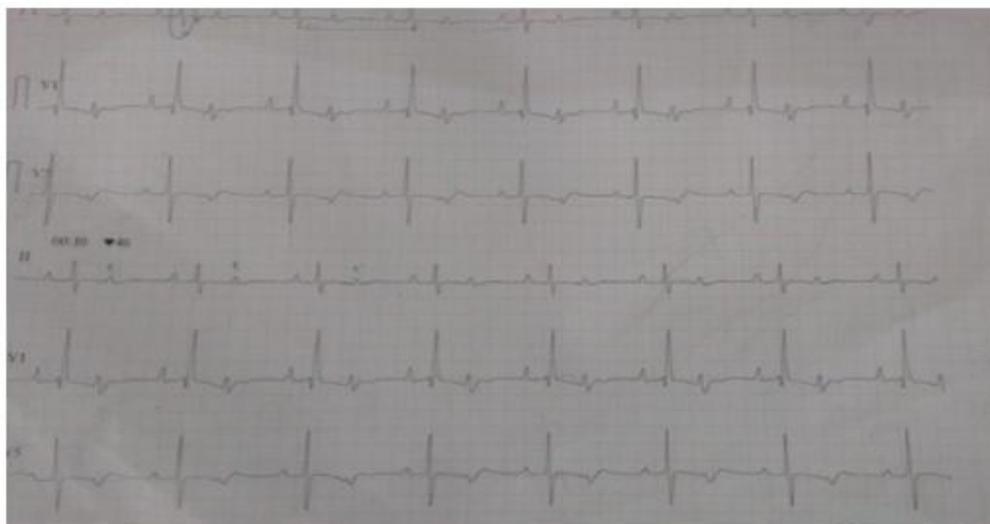
On examination patient conscious and oriented, pulse –56/ min regular, blood pressure – 130/70 mm Hg and jugular venous pressure elevated 11 cm. Patient had central cyanosis with clubbing present in the lower limbs and pedal edema. Systemic examination cardiac – parasternal heave with palpable P2 present; first and second heart sounds heard with a loud P2 and a early diastolic murmur of pulmonary regurgitation was present in the pulmonary area radiating to all over the chest. Bilateral fine basal crepitations were present in the lung fields and abdomen distended and shifting dullness was appreciated. Central nervous system examination was normal.

His complete haemogram as expected showed polycythemia with haemoglobin 14.7 g%,

haematocrit – 50.6% and RBC count –  $7.1 \times 10^6$  / cu mm. His renal function tests and serum electrolytes were normal. Electrocardiogram of the patient showed second degree Mobitz type I atrio ventricular (AV) block (wenckebach's phenomenon) with transient complete AV block spontaneously reverting to  $2^0$  degree AV block. Chest X ray of the patient showed cardiomegaly with fluffy opacities in the left mid and lower zones. 2 Dimensional echocardiography showed dilated right atrium and right ventricle with moderate tricuspid regurgitation and severe pulmonary artery hypertension. Patient was advised for pacemaker insertion.



**Figure 1:** ECG of the patient - second degree atrioventricular block (2:1 wenckebach's phenomenon) with complete Right bundle branch block with poor 'R' wave progression and marked right axis deviation



**Figure 2:** ECG of the patient showing complete AV block with regular PP interval and regular RR interval with no relationship between the P wave and QRS complex.

**Table 1:** Arrhythmias and associated congenital cardiac abnormalities in adults:

ARRHYTHMIA	CONGENITAL HEART DISEASE
<b>BRADYARRHYTHMIA</b>	
Congenital sinus node dysfunction	Heterotaxy syndrome
Acquired sinus node dysfunction	Postoperative Mustard; postoperative Senning; postoperative Fontan, others
Congenital AV block	Endocardial cushion defects, L-TGA
Acquired AV block	VSD closure, subaortic stenosis, AV valve replacement
<b>TACHYARRHYTHMIA</b>	
Accessory pathways	Ebstein's anomaly, L-TGA
Intra atrial reentrant tachycardia (Atrial flutter)	Postoperative Mustard; postoperative Senning; postoperative Fontan, others
Atrial fibrillation	Mitral valve disease, congenital aortic stenosis, unrepaired single ventricle
Ventricular Tachycardia	Tetralogy of fallot, congenital aortic stenosis, others

## Discussion

Eisenmenger syndrome is a reversal of the shunt from (left to right) to (right to left) with the development of pulmonary hypertension. It occurs early in life in VSD (ventricular septal defect) and late in life in Patent ductus arteriosus and Atrial septal defect. Patients present with cyanosis and palpable P2 due to pulmonary hypertension<sup>1</sup>. In early stages of Eisenmenger syndrome surgical repair of the shunt lesion is possible but if not then it may end up in needing a heart lung transplantation for definitive cure. The patients with Eisenmenger syndrome are conservatively managed with phosphodiesterase inhibitors and endothelin receptor antagonists and studies have shown symptomatic improvement in these patients<sup>1</sup>. The complications of Eisenmenger syndrome are fatal and include cardiac failure, pulmonary infections, pulmonary thrombosis, brain abscess, infective endocarditis, massive haemoptysis and ventricular arrhythmias<sup>1,2,3</sup>.

Arrhythmias are common in congenital heart diseases (CHD) and often lead to poor quality of life and sudden cardiac death in these patients. Some cardiac rhythm abnormalities are intrinsic to the structural malformation as Wolf-Parkinson-White syndrome in Ebstein anomaly and atrioventricular blocks in corrected transposition of great arteries. But in most patients with CHD arrhythmia is acquired due to surgical scars and abnormal pressure and volume overload for prolonged duration. Most of these abnormalities can develop in almost all forms of CHD<sup>2,3</sup>.

Atrioventricular (AV) block is seen commonly in patients with endocardial cushion defects and corrected transposition of great arteries (TGA). It may be due to abnormal location and function of the atrioventricular node in them. In TGA as much as 20% develop AV block in adulthood. Its likelihood increases with surgical correction<sup>2</sup>. But this patient had Patent ductus arteriosus with reversal and presented with 2<sup>nd</sup> degree AV block with transient complete AV block. Surgical correction of any CHD can lead to AV block as a result of direct trauma but now with more precise anatomical knowledge its incidence is less. But in our patient he was not operated and probably the long standing volume overload could result in AV block. There are reports of complete heart block occurring in patients with Eisenmenger's syndrome. Also studies have shown that prolonged PR and QT interval are risk factors for developing arrhythmias in patient with Eisenmenger syndrome<sup>4</sup>. This patient deteriorated with the occurrence of AV block resulting in worsening cardiac failure. These patients are more prone for sudden cardiac death.

Post surgical AV block is transient and recovers over a period of 10 days. It is due to myocardial stretch or edema post surgery. Patients with post surgical AV block not recovering within 10 days come under class II recommendation for pacemaker implantation<sup>2,5</sup>.

Sinus bradycardia is seen commonly in adults due to surgical trauma to the node or its arterial supply. Sinoatrial (SA) node dysfunction is seen

in patients with defects in the caval atrial junction. This is seen in patients with heterotaxy syndrome with patients with a single ventricle a very rare disease<sup>5</sup>.

Treatment of heart block in patients with CHDs with pacemaker is often difficult. The standard transvenous approach is difficult because of complex anatomy and increased risk of thromboembolism. Epicardial implantation is possible in these conditions but requires high skills because of the anatomy. Also these patients require repeated surgeries for lead replacement and battery changing. The indication for ICD (Intra cardiac defibrillators) in CHD is still evolving but the guidelines are similar to that of other cardiac conditions. Tetralogy of fallot and TGA are condition requiring ICDs. Also resynchronization for right bundle branch block seen in CHDSs is under trial<sup>6</sup>.

To conclude, the main aim of presenting this case report is highlight the arrhythmias in congenital heart disease. Arrhythmias are a serious issue in patients with CHD and there are no specific guidelines for management of these patients and often treatment of these patients is technically demanding. Further researches are required to do the best for these patients.

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