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A case study on acyanotic congenital heart disease due to ostium secundum-Atrial septal defect and Mitral Regurgitation

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Abstract- An Acyanotic Congenital Heart Disease is a type of congenital heart disorder in which the blood is shunted from the left part of the heart to the right part of the Heart. An Ostium Secundum ASD is a hole in the middle of the atrial septum. This results in the mixing of the blood from both sides leading to the inefficient working of the heart. A 23-year-old female patient came to the hospital with chief complaints of breathlessness for a year and palpitations on and off. The vitals of the patient seem to be normal. An echocardiogram reveals that the right atrium is dilated and the interatrial septum has an ostium secundum defect. Doppler Ultrasound shows the presence of mild mitral valve regurgitation. paCO₂ is higher than the normal range. Leucocyte count also shows increased levels. From the above clinical investigations, the patient was diagnosed with acyanotic congenital heart disease due to Ostium Secundum atrial septal defect and mild mitral regurgitation. OS-ASD closure was done using the autologous patch made from the pericardium and the mitral valve was repaired. Drugs are given to reduce the signs and symptoms of ASD and to reduce the risk of postoperative complications includes: T. Metolar 100 (Metoprolol), Neb. Levolin (Levosalbutamol), Inj. Heparin and T. Ecospirin (Aspirin). Postoperative treatment includes antibiotic prophylaxis to prevent Infective Endocarditis: Inj. Cefuroxime.

Keywords- Congenital heart disease, Ostium Secundum, Atrial septal defect, Mitral regurgitation, Autologous patch, Prophylaxis.

Abbreviations and Acronyms

CHD- congenital heart disease ASD- Atrial septal defect CT- Computed Tomography MRI- Magnetic resonance imaging MR- Mitral Regurgitation TTE- Transthoracic echocardiogram DA- ductus arteriosus PDA- Patent ductus arteriosus VSD- Ventricular septal defect OS- Ostium Secundum iv- intravenous po- per os OD- omne in die

I. INTRODUCTION

Congenital heart defects can be divided into two broad types: acyanotic and cyanotic defects. The most common types of acyanotic defects are atrial septal defect, ventricular septal defect, pulmonary stenosis, patent ductus arteriosus, atrioventricular canal, aortic stenosis, and coarctation of the aorta. An Atrial Septal Defect (ASD) is the third most common type of CHD and represents 30% to 40% of congenital heart defects (CHDs). Atrial septal defect (ASD) is characterized by the formation of a persistent hole in the interatrial septum after birth that allows direct communication between the left and right atria. It occurs anywhere along the atrial septum, but the most common site is in the region of the foramen ovale, termed an ostium secundum ASD. Diagnosis of ASD is made based on: a transthoracic echocardiogram, cardiac CT, and MRI. Both MRI and CT examine the structures that surround the heart and the thoracic cavity. Cardiac catheterization is contraindicated in patients who are young and present with small, uncomplicated ASDs. Conventionally Atrial septal defects are treated by surgical closure. It can also be closed using a pericardial or synthetic patch. Transcatheter device closure has increasingly been used with good results.^{[11][2]}

Mitral regurgitation (MR) is a common type of valvular heart disease. There are two types of mitral regurgitation; primary degenerative MR, and secondary MR. Mitral valve regurgitation is a type of heart valve disease in which the valve between the left heart chambers does not close completely, so there is a reflux of blood during systole from the left ventricle into the left atrium. Mitral regurgitation usually arises as a complication of mitral valve prolapse; other causes include LV dilatation (e.g., left-sided cardiac failure), infective endocarditis, acute rheumatic heart disease, and papillary muscle rupture after myocardial infarction.^{[4][5]}

Diagnosis of Mitral Regurgitation is made based on: a Transthoracic echocardiogram (TTE) which provides information about the size and function of the left and right ventricles. ^{[6][7]} Surgery is the treatment of choice for severe mitral regurgitation. For patients who are at high risk from surgery, and particularly those with severe heart failure, mitral clip implantation is preferred.

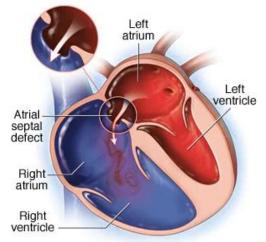


Figure 1 This image depicts the abnormal structure of Right atrium and mixing of blood between the atrium and ventricle.

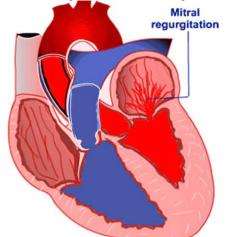


Figure 2 This image depicts the mitral regurgitation and its effect

II. HISTORY

The standardized history of acyanotic congenital heart disease started in 1930 with the establishment of Dr. Helen Taussig's pediatric cardiology clinic at Johns Hopkins Hospital in Baltimore and the Atlas of Congenital Heart Disease in 1936 published by Dr. Maude Abbott. Many experts from all over the world have contributed towards advances in Surgery, diagnosis, and interventions till the 1930s.

The initial surgical approach was ligation of patent ductus arteriosus accomplished by Dr. Robert Gross in 1938 at the Children's Hospital in Boston. The possibility of intracardiac repair came up with the development of Cardiopulmonary bypass technology in the 1950s, followed in the 1970s by the evolution of circulatory arrest along with deep hyperthermia, which made prolonged surgeries possible. Interventional techniques also went mutual with surgical advances. Balloon dilatation of the pulmonary valve was expressed in 1953 by Rubio-Alvarez and colleagues, which was not commonly used until the development of static balloon dilatation in 1982 by Kan and Colleagues. In 1966 Balloon Atrial septostomy was evolved to encourage mixing at the atrial level and greatly enhanced the result for newborns with complete transposition of the great arteries. Later the innovation began with the introduction of X-ray imaging, a diagnostic method that helped both surgical and nonsurgical interventions. Right Heart Catheterization was developed in the late 1940s and Left Heart Catheterization became available in the 1950s. The approach of two-dimensional echocardiography granted a vital step advancing in the treatment of congenital heart disease in the 1970s. Dr. Harold Rice created the first Cardiopulmonary bypass machine used at St. Paul's Hospital.

In 1953, Dr. Wilfred Bigelow from Canada determined the application of total body hypothermia for open Heart surgery, and the initial open-heart procedure was carried out by Dr. John Callaghan in Edmonton in 1954.

Due to the advances made since the 1930s, Children born with Acyanotic Congenital Heart disease now is much more expected to grow to middle age, but they are also able to undergo several operations for constriction in arteries or veins and replacement or insertion of valves.

III. BRIEF PATHOGENESIS

Pathogenesis of atrial septal defect (ASD)

An atrial septal defect is a type of congenital heart defect. The atrial septum develops between the fourth and sixth weeks of embryonic life. The initial phase is marked by the growth of a primary septum (septum primum) from the dorsal wall of the common atrial chamber toward the developing endocardial cushions. A gap initially separates the developing septum primum from the endocardial cushion. This gap is termed Ostium primum. Later a second opening called Ostium secundum appears in the central area of the primary septum. As the Ostium Secundum enlarges, a secondary septum (septum secundum) makes its appearance just to the right of the Septum primum. This septum secundum covers the ostium secundum leaving a small opening called Foramen Ovale. This is essential for the baby during its embryonic stage.^{[1][2]} At birth, the septum primum and septum secundum slap shut and then fuse. An atrial septal defect occurs when the septum that separates the right and left atrium does not close and remains open even after birth. An ASD allows blood from both atria to mix, causing the heart to work less efficiently. In addition, it causes a left-to-right shunt and volume overload of the right atrium and right ventricle. As a result, there is a gradual enlargement of the right side of the heart and the pulmonary arteries.^[5]

Pathogenesis of mitral regurgitation (MR)

Mitral valve regurgitation is a type of valvular heart disease in which the valve between the left chambers of the heart doesn't close completely, allowing blood from the left ventricle to leak backward across the valve to the right atrium. Mitral regurgitation results in left ventricular volume overload due to increased stroke volume, caused by increased blood volume within the left atrium and an increased preload delivered to the left ventricle during diastole.^{[3][4]} Later volume overload from MR causes ventricular dilatation, widening of the mitral annulus, and diminished coaptation of leaflets, leading to further worsening of MR. Alternatively, it can be associated with a cleft of the mitral valve, as occurs in persons with Down syndrome, or an ostium primum ASD.^[5]

IV. TYPES OF ACYANOTIC CONGENITAL HEART DISEASES

There are several types of Acyanotic congenital heart diseases. They are

Aortic Stenosis

Aorta is the major artery that carries blood from the heart and supplies it to other parts of the body. The aortic valve is located between the heart and the aorta. The function of the valve is to open and close during each cardiac cycle to prevent the backflow of blood into the heart. This allows a constant supply of blood to different organs of the body. When the aortic valve abnormality takes place, the valve will not close properly and this condition is called Aortic stenosis.^[8]

Atrial septal defect

Atrial septal defect (ASD) is characterized by the formation of a persistent hole in the interatrial septum after birth that allows direct communication between the left and right atria. Tiny Atrial Septal Defects can often be left alone, but larger Atrial Septal Defects may require a surgical procedure o close them.^[8]

Atrioventricular septal defect

It is the hole in the wall that separates the upper and lower chambers. This condition is also called an atrioventricular canal defect or an endocardial cushion defect.^[8]

Bicuspid aortic valve abnormality

The bicuspid aortic valve has only two flaps in contrast to the normal aortic valve which normally has three flaps that open and close to regulate the blood flow that results in the valve function abnormality.^[8]

Coarctation of aorta

The aorta will be narrowed or pinched in aortic coarctation which results in limited blood flow to the rest of the body.^[8]

Patent ductus arteriosus

A PDA, defined as failure of the ductus arteriosus (DA) to close within 72 hours after birth, may result in significant infant morbidity and mortality rates that approach 30%.^{[8][9]}

Pulmonary stenosis

The pulmonary artery carries deoxygenated blood from the heart to the lungs for oxygenation. The pulmonary valve is the valve that is present between the heart and the pulmonary artery. Narrowing of this valve leads to pulmonary stenosis.^[8]

Ventricular septal defect

This condition is like an Atrial septal defect, but it's a hole in the wall that separates both ventricles. Usually, closure of this hole takes place on its own, but some may need a surgical procedure.^[8]

V. CASE REPORT:

Chief complaints

A 23-year-old female patient came to the hospital with chief complaints of breathlessness for a year and palpitations on and off. The patient was conscious-oriented and afebrile. The vital signs of the patient are normal

Medical and medication history

The patient was diagnosed with Ostium Secundum ASD with mild Mitral regurgitation at 17 years of age during a regular health check-up. No other medical and medication history was observed.

INVESTIGATIONS

PARAMETERS	OBSERVED VALUE	NORMAL VALUE
Hemoglobin	12 g/dl	12-16 g/dl
Leucocyte	13,200 cells/µl	5000-11000 cells/µl
Eosinophils	4.8%	0-5%
Neutrophils	47%	40-60%

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Table 2 Arterial blood gas				
PARAMETERS	OBSERVED VALUE	NORMAL VALUE		
spO ₂	92%	90-100%		
paCo ₂	152 mmHg	75-100 mmHg		

Table 3 Electrolytes				
PARAMETERS	OBSERVED VALUE	NORMAL VALUE		
Potassium	3.1 mmol/L	3.5-5 mmol/L		

Echocardiogram

- An Echocardiogram is an ultrasound scan used to produce an outline of the heart's working action. This is the most widely used gold standard test to diagnose Atrial Septal defects. The movement of the heart is produced in pictures by sound waves.
- Both ventricles perform their functions normally.
- Right atrium is dilated due to excess shunting of blood from left to right atrium due to the presence of ostium secundum ASD.
- Interatrial septum was observed and a moderate-sized hole was reported leading to Ostium Secundum Atrial Septal Defect.
- Interventricular septum was observed to be intact.

Doppler Ultrasound

- Doppler Ultrasound is a test performed to analyze the blood flow through the blood vessels using high-frequency sound waves.
- Mitral valve shows mild mitral regurgitation which means the valves between the left chambers of the heart do not close completely, allowing blood to leak backward across the valve.
- No regurgitation was observed in Pulmonary and Tricuspid valves.

CLINICAL DIAGNOSIS

The clinical diagnosis was made based on the evidence of

- Breathlessness
- Palpitations
- Dilated Right Atrium is seen in Echo
- A moderate-sized hole at the interatrial septum was observed on Echo
- Mild Mitral Regurgitation is seen in the Doppler ultrasound
- > Increased $paCo_2$ levels of arterial blood gas.

The patient came to the hospital with complaints of breathlessness for a year and palpitations on and off. An echocardiogram test reveals that the right atrium of the patient was dilated due to an Atrial septal defect. It also shows that a moderate-sized hole present in the interatrial septum results in ostium secundum ASD.

On Doppler ultrasound, Mild mitral valve regurgitation was observed. On arterial blood gas analysis, partial pressure of Co_2 is highly increased due to the excessive flow of blood from the right atrium to the lungs. Based on the above evidence the patient was diagnosed with Acyanotic Congenital Heart Disease due to Ostium Secundum Atrial Septal Defect and Mild mitral regurgitation.

Table / Drugs prescribed

BRAND NAME	GENERIC NAME	ROUTE	DOSE	FREQUENCY	DAYS
Inj. M-CEFURO	Cefuroxime	IV	1.5 g	1-1-1	2 days
Inj. Heparin	Heparin	IV	2500 U	1-1-1	2 days
T. ECOSPIRIN	Aspirin	РО	75 mg	0-1-0	2 days
Neb. MUCOMIX	Acetyl Cysteine	Inhalation	2 ml	1-0-1	1 day
Neb. BUDECORT	Budesonide	Inhalation	1 mg	Q12hr	3 days
Neb. LEVOLIN	Levosalbutamol	Inhalation	0.63 mg	1-0-1	2 days
Inj. PUREMOL	Paracetamol	IV	1000 mg	1-0-1	2 days

T. LASILACTONE	Furosemide+ Spironolactone	PO	20/50 mg	OD	3 days
Inj. Fentanyl	Fentanyl citrate	IV	250 mcg	OD	1 day
Inj. PAN IV	Pantoprazole	IV	40 mg	1-0-0	3 days

TREATMENT OUTCOMES:

After the Ostium Secundum atrial septal defect closure using the autologous pericardial patch and the mitral valve repair procedures, the drugs were administered to prevent postoperative bacterial infections and platelet aggregation. The levosalbutamol and acetylcysteine were administered to reduce the shortness of breath. Medicines were also prescribed to relieve pain. After treatment, the patient was stable and all the vitals were found to be normal. The chest X-Ray revealed that the Cardiothoracic Ratio (CTR) has been slightly increased. On discharge, the patient was advised to have a restriction on the fluid intake (1500 ml/day) in order the reduce the cardiac workload. Diuretics and carvedilol were administered to reduce the workload of the heart. The patient must avoid spicy and saturated fatty foods. The patient was monitored closely for any signs of shortness of breath and chest tightness.

VI. DISCUSSION:

An acyanotic congenital heart disease is a class of congenital heart defects. In this condition, the blood is shunted from the left side of the heart. This is due to an atrial septal defect. The patient is also having mitral regurgitation, a condition where the mitral valve is damaged and cannot close properly. This results in the backflow of blood from the left ventricle to the left atrium. The patient started to experience symptoms such as shortness of breath and palpitation at 23 years of age. After proper diagnosis, the patient has undergone a surgical procedure to close the ASD using an autologous pericardial patch and a mitral repair was done. After surgery, the patient was administered antibiotics and antiplatelet drugs as prophylaxis. The patient was stable after surgery and treatment. She was discharged from the hospital with medical and lifestyle modification advice. An acyanotic congenital heart disease can be treated in most cases by a successful surgical procedure. The closure of ASD using an autologous pericardial patch has a higher success rate now. The drugs which are prescribed after surgical procedures can prevent complications and reduce the symptoms. There are more treatment options available now which are less invasive and have higher success rates.

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