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Atrial Septal Defect- A Review

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Abstract:- Among the most prominent congenital heart disease (CHD) in humans includes Aortic Stenosis (AO), Patent Ductus Arteriosus (PDA), Tetralogy of Fallot (TOF), Atrial Septal Defect (ASD), Ventricle Septal Defect (VSD) etc. Of these, Atrial septal defect (ASD) is the most prevalent, appearing in adulthood and being identified from an early age. Although the exact etiology of CHD has not been determined by numerous researches, variables like consanguinity marriages among first cousins and alcoholism, smoking, medication usage during pregnancy have been linked to ASD. Anatomically, several gene mutations cause this to happen. Open cardiac surgery with CPB and transcatheter closure are among the surgical technique used. If necessary, long term care will be provided after the procedure. Thus, the content of this article provides a summary of the know-how regarding atrial septal defect.

Keywords:- *Congenital Heart Disease (CHD); Atrial Septal Defect; Pimum Defect; Secundum Defect; Patent Foramen Ovale (PFO); Coronary Sinus Defect; Cardiac Catheterization; Open Cardiac CPB (Cardiopulmonary Bypass) Surgery.*

I. INTRODUCTION

A. Congenital Heart Disease

Childhood mortality from congenital heart disease (CHD), the most prevalent birth defect in humans, is still a major concern (1). Worldwide, 0.8% of newborns are born with congenital cardiac disease. Over 85% of these infants have survived to adulthood owing to advancements in medical and surgical therapies over the past few decades (2). Cardiovascular and non-cardiac comorbidities are widespread among adult survivors of congestive heart failure (CHD) despite improvements in therapeutic care improving the prognosis and quality of life for these patients (1). Grown-ups with congenital heart disease (GUCH) are a diverse category of people who are prone to age-related cardiac disorders such as coronary heart disease and heart failure (3). None of the following conditions were found to be correlated with consanguinity: Aortic Stenosis (AS), Tricuspid Atresia (TA), Co-Arction of the aorta (Coa) and Patent Ductus Or Tetralogy of Fallot (TOF). Consanguinity may thereby worsen underlying genetic risk factors in population with a high degree of inbreeding, especially in first cousin kids. Certain heart abnormalities may be caused by a recessive component (4).

B. Anatomy of Atrial Septum

The fibrous part of the heart's septal structures is known as the "membranous septum". Though it has somewhat understated appearance, it has a significant impact on our knowledge of heart illness, both acquired and congenital (5). In humans, there is a lot of variation in the interatrial septum, which is situated between the right and left atrium. Frequently described variations include the smooth septum, Patent Foramen Ovale (PFO) channel, and atrial septal pouches (SPs) (6). The passage between a non-adherent septum primum (SP) and secundum (SS) across the interatrial septum is identified as a Patent Foramen Ovale (PFO) (7). The formation of the fossa ovalis, which has two anatomical components, is the result of normal development of the atrial septum. The first is the muscular boundaries, which are contributed by the septum secundum; the second is the valve of the fossa ovalis, which attaches on the left atrial aspect of the septum secundum. The atrial component of the atrio-ventricular canal septum-from the fossa ovalis by lying anteriorly and inferiorly. The sinus venosus is the tissue that divides the superior vena cava, the posterior and inferior parts of the right atrial free wall, and the right pulmonary veins (8).

Upon dissecting a normal heart within the vicinity of the pre-suspected atrial septum, it becomes evident that the in folded atrial walls situated between the caval and pulmonary veins are the superior elements of the septum secundum. This was previously noted by His the Elder (1885), who called the superior border of the oval foramen a "Muskuloseleiste" (a muscular infoldings) (9).

C. Development of Atrial Septal Defect in Pregnancy

The confines of bilateral cardiogenic fields, the heart is made up of early progenitor cells from two or more sources (10). ASDs have been linked to mutations in a number of genes, including TBX5, GATA4, NKX2-5, and other cardiac transcription factor genes (11).

The right atrial roof folds during the sixth week of pregnancy, forming the initial stage of the dorsocranial margin of the septum secundum and progressively

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overlapping the foramen secundum in the septum primum. This folding occurs between the primary septum and the ostium of the superior vena cava. Fetal growth happens very quickly during the last 20 weeks of pregnancy, but the foramen ovale, which now has a gap between the primary and secondary septa, still corresponds to the size of the right atrium (12). The septum primum divides in two, with the lower portion functioning as the foramen ovale valve and the upper portion degenerating. Indeed, during approximately 20% of the cardiac cycle, this flap shuts the foramen ovale, this flap shuts the foramen ovale, as evidenced by echocardiography (13).

Similar to a flap valve, the septum primum directs blood flow from the right atrium to the left atrium via the ostium secundum, which continues to be an essential conduit for fetal blood flow. Due to increased pressure in the left atrium, the flap valve; or floor of the oval foramen, is forced against the muscular rim after birth, closing this opening. In cases where adhesion is not complete throughout the full rim boundary, a probing patent foramen ovale (PFO) will persist. The Eustachian ridge (sinus septum) and the vestibule that leads to the tricuspid valve are continuous with the antero-inferior rim of the oval fossa. The right atrial wall, the wall of the left ventricle, the fatty tissue of the inferior atrio-ventricular groove, and the artery supplying the atrioventricular node of the conduction system form a “sandwich” in this region, which was once known as the muscular atrio-ventricular septum. The tricuspid valve attachment is more apical than the mitral valve, and this difference in atrio-ventricular valve hinge line levels at the cardiac septum is what divides the right atrium from the left ventricle (12).

On rare occasions during intra uterine life, the ovale foramen, or FO, shuts. This could result in ascites, pericardial effusion, pleural effusion, significant hypertrophy of the right atrium and ventricle with arrhythmia, right heart failure and congestion, non-immune hydrops, and under development of the left side of the heart. Death frequently happens short after birth (14). While an actual ASD is a hole inside the atrial septum, most commonly used terminology classifies any hole that allows blood to be shunted between atrial chambers as an ASD, especially if the hole is larger than 3-4mm. The most typical are “secundum defects,” or holes, found in the oval fossa. “Ostium primum” defects, faults in the superior and inferior sinus venosus, coronary sinus defects, and confluent or common atrium are examples of inter atrial connections that occur outside the actual septum (15).

II. ATRIAL SEPTAL DEFECT

Three basic groups include inter atrial communications, or ASDs: Ostium secundum, Ostium primum, and sinus venosus abnormalities. An actual atrial septal defect called the ostium secundum affects the region surrounding the fossa ovalis (16). The inter atrial communications include several distinct defects in the cardiac terminations of the systemic and pulmonary veins (sinus venosus and coronary sinus defects) and in the inter atrial septum (atrial septal defects) (17). Three groups of defects result from variations in the atrial rims: muscular, doubly committed, juxta arterial (sub arterial) and peri membranous (18).

A. Patent Foramen Ovale

Patent foramen ovale is the space between a well-developed (valve-competent) septum primum and a normally formed septum secundum (19). It is an embryonic defect in the inter atrial septum that permits oxygenated blood to move from the right to the left atrium (20). After birth, left atrial pressure normally exceeds right atrial pressure and septum primum apposes septum secundum and the foramen ovale narrows. Nearly every infant has a patent foramen ovale, yet as people age, this condition becomes less common. In 70-75 % of adults, the foramen ovale closes completely antomically(19).

B. Secundum Atrial Septal Defect

Individuals who have an ASDII **(**Secundum Atrial Septal Defect) that results in enlarged right heart chambers are at significant risk for age-related morbidity and shortened life expectancy. Although they can be identified and treated in their early years, they often go unrecognized and symptomatic far into adulthood. As a result, these patients represent a diverse adult population in terms of the severity of their illnesses, the likelihood of complications, and the need for ongoing medical monitoring and care (21). The dimensions of secundum imperfections range from a few millimetres to 2-3cm. Significant deficiency, or possibly the loss of septum primum altogether, is typically linked to large malformation (8).

C. Primum Atrial Septal Defect

An inter atrial communication is situated between the anterio-inferior border of the fossa ovalis and the atrio-ventricular valves. Primum atrial septal defect is one of several variations of common atrio ventricular canal defects, also known as atrio ventricular septal defect. Anterior mitral leaflet clefts and other anomalies in the atrio ventricular valves are virtually invariably presents in this condition along with the septal defect. In contrast to other kinds of atrial septal defects, the full atrio ventricular canal defect is characterized by an abnormal conduction axis position (8). Complete repairs can be made with the least amount of mortality possible on patients in stable health before the age of two or three. Even though there is a higher risk of morbidity, early correction should be considered in newborns with severe congestive heart failure (22).

D. Coronary Sinus Defect

A shunt via the defect and the coronary sinus orifice is made possible by the partial or whole unroofing of the tissue dividing the left atrium from the coronary sinus. This unusual atrial connection is the consequence. Raghib syndrome is the phrase used to describe the combination of a persistent left superior vena cava and a coronary sinus septal defect (23).

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III. ETIOLOGY

Since there is no known explanation for the majority of ASDs, they can also occur in hereditary syndromes such as Down, Holt-Oram, Ellis van Creveld, and Noonan syndrome, where the prevalence is roughly 80%, 65%, and 20%, respectively. Furthermore a number of behaviours and illnesses have also been linked to a higher chance of producing children with ASD, such as maternal alcohol use, smoking, antidepressant medication use, and diabetes (11). ASD can appear as a single lesion or in combination with more complicated congenital heart disease forms, such as tricuspid valve abnormalities or Tetralogy of Fallot. Clinical presentation varies widely, from people presenting with severe pulmonary vascular disease and right heart failure (Eisenmenger syndrome) to asymptomatic subjects who live to adulthood without diagnosis (24).

IV. DIAGNOSIS

An essential tool for determining the presence of an ASD in both adults and children is the plain chest radiography. Most children are asymptomatic, therefore a considerable proportion of cases will be found on routine chest radiographs, even though the diagnosis is easily made by physical examination. However, the physical indicators could be unclear even though the majority of individuals have symptoms (25).

Significant atrial shunt lesions must be suspected in the event of any indescribable dilation of the right heart chambers. The diagnosis can be assists by careful TTE (Transthoracic Echocardiogram) imaging of the inter atrial septum using 2D echocardiography from unusual angles, the use of colour Doppler imaging and intravenous contrast agents. The anatomy of the interatrial septum may not be decisive in TTE situations with low imaging quality. Furthermore, colour Doppler imaging may be uncertain due to large shunt regions and consequently slow blood flow through the lesion. A substantial left-to-right blood flow in these patients may prevent the right heart's contrast agent from reaching the left heart. While the typical wash-out phenomena can be easily seen with TTE, also it can be challenging to find in TTE (26).

Preoperative imaging is utilized to evaluate the following: the size, confluence, and origin of the main pulmonary artery; the aortopulmonary collaterals; the presence of patent ductus arteiosus; other sources of collaterals, such as the bronchial and coronary heart disease as well as related aortic, pulmonary venous, and coronary defects. Following unifocalization and stent implantation, postoperative imaging is performed to evaluate patency, stenosis, and occlusion of the stent or perivascular lesions like seroma (27).

V. PROCEDURES

A. Cardiac Catheterizations

A procedure used by physicians to help identify and treat heart issues is cardiac catheterization. This examination can detect congenital cardiac anomalies, asses blood pumping capacity, measure oxygen and pressure in each chamber of the heart, and assist in the diagnosis of heart valve issues (28)

B. Transcatheter Closure

These days, transcatheter closure of secundum type atrial septal defect (ASD) and patent foramen ovale (PFO) is carried done in numerous sites worldwide, even in non-paediatric/congenital interventional laboratories. When there is a substantial left-to-right shunt across the atrial connection, right ventricular volume overload is evident, and pulmonary vascular resistance is normal, ASD closure is recommended (29). The ASO and the Gore Cardiform device are the two devices that are currently generally accessible in the US. After proving safety and effectiveness in a non-randomized IDE (Investigational Device Exempion) trial the ASO was the first TC-ASD (Transcatheter Atrial Septal Device) device to be approved by the FDA. This was further supported by a later multicentre registry research (30).

Since 1974, per-catheter devices have been developed for the closure of atrial septal defects (ASDs). In early 1997, the four major devices are ready for limited use. These consist of the Angel Wing device, the ASDOS device, the Buttoned device, and the Cardio Seal device (which is the replacement for the Clamshell device) in alphabetical order (31). Outside of the US, , Nit-Occlud® ASD –R (Atrial Septal Defect) and PDR-R (Patent Ductus Arteriosus) devices are used for percutaneous closure of atrial septal defects and patent ductus arteriosus. In the catheterization lab, ASD-R and PDA-R devices can be successfully retrieved. It is essential to employ appropriately sized sheathes and to grab the central portion of the right atrial disc of the ASD-R device or the pulmonary component of the PDA-R device (32).

The benefits of ASD interventional occlusion include less trauma and a speedy post-operative recovery; however, X-ray might cause harm to human tissue. In addition, there is a chance of allergies and renal failure while using contrast chemicals intra operatively.

C. Heart Surgery

Under CPB (Cardiopulmonary bypass), the ASDR surgical method is very advanced. Under direct gaze, the curative impact is precise and has a high success rate. With minimal mortality, nearly all forms of ASD can be treated in this manner. Nevertheless, the drawbacks include prolonged hospital stays, patient trauma, the requirement for significant blood expenses, a long-term dependency on antibiotics, the requirement for CPB, excessive expenses, and the potential for the pericardium to adhere to the heart's surface, which could necessitate difficult follow-up procedures. In addition, the patient is unwilling to accept this operation because of

the apparent and noticeable scars (33).

VI. FOLLOW-UP CARE

Adult CHD specialists may be necessary and beneficial for ASD patients at any point in their adult lives. This is especially true for patients with primum ASDs who are susceptible to left atrioventricular valve issues, as well as those who have late ASD closure and partial reverse right ventricular remodelling (11). The transition from paediatric to adult CHD services is necessary to guarantee effective lifelong care; skip over may be that contributed to higher late morbidity and mortality (34).

VI. CONCLUSION

The most prevalent birth anomaly in humans, ASD is still a major contributor to childhood death rates. Percutaneous closure is both efficient and safe. More hospitalization may result from CPB open surgery. However, the likelihood of CPB open surgery can be decreased with early identification of ASD. Following surgery, many patients will benefit from on-going monitoring so that any complications can be identified and treated early. Additionally, all patients should adopt healthy lifestyle choices and modifications to reduce their overall risk of cardiovascular disease and to optimize their recovery from ASD closure.

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