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Chronic Pulmonary Hypertension and Congenital Heart Disease

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Abstract

A mean pulmonary arterial pressure (MAP) of less than 25 mmHg is regarded as pulmonary hypertension. We examine pathogenesis, diagnosis, and treatment with a focus on their applicability to congenital heart disease. The prevalence of pulmonary hypertension in adults ranges from 5 to 10%, making it a rather common consequence of congenital heart disease. The size and kind of the heart abnormality, as well as hereditary and environmental variables, all play a part in the multifactorial cause that is acknowledged. Instead of the pure Eisenmenger complex, more complex disease is becoming more widely acknowledged. Increased pulmonary vascular resistance, which is brought on by remodelling of the pulmonary vascular bed, can be identified using a range of tests, including echocardiography, exercise testing, cardiac catheterization, MRI, and CT scanning. Medications that treat disease are employed in management, and their effectiveness is rising.

Keywords: Pulmonary hypertension • Eisenmenger • Pulmonary vascular resistance • Congenital heart disease

Introduction

About 10% of adult cases of congenital heart disease (CHD) have a complication known as pulmonary hypertension (PH). It is characterised by a mean pulmonary arterial pressure (PAPm) of less than 25 mmHg, as measured at rest during right cardiac catheterization. Sufferers with precapillary PH are classified as having pulmonary arterial hypertension (PAH), a subgroup of PH patients. A pulmonary artery wedge pressure 15 mmHg and a pulmonary vascular resistance (PVR) >3 Wood Units in patients without any other precapillary PH causes, such as lung disease and persistent thromboembolic pulmonary hypertension, are indicative of this.

Literature Review

According to reports, there are 97 instances of PH per million people in the UK, with a female to male ratio of 1.8. Additionally, in the USA, the age-standardized death rate from PH ranges from 4.5 to 12.3 per 100,000 people. In adults with CHD, PAH is present in 5–10% of cases, and this has a significant influence on mortality and morbidity, increasing the need for lifetime care. A Dutch study that examined the epidemiology of people with septal abnormalities found that 6.1% of them had CHD-APAH, with a median age of 38 and a female predominance of 60 percent. The population's estimated incidence of CHD-APAH is 2.2 per million, with a prevalence of 15.6 per million, with Eisenmenger syndrome accounting for 58% of cases. In addition, the most common underlying defect (42%) was ventricular septal defect (VSD). This study also emphasises the risk of PH following surgery to seal defects, with 3% of these individuals continuing to experience PH. The dynamic nature of the syndrome and its complex genesis are highlighted by the fact that the severity of CHD-APAH can vary significantly even in the presence of similar underlying cardiac abnormalities [1]. The underlying abnormality that causes CHD-APAH determines the cause, although it is also believed that environmental factors, genetics, and/or epigenetics may also be involved. This is demonstrated by a study that discovered BMPR2 mutations in 6% of individuals with CHD-APAH, which are also but less strongly (75 and 25%, respectively) related with familial and idiopathic PAH. PAH is well recognised to be caused by this as well as mutations in the gene encoding the protein ALK-1. Postcapillary hypertension and left heart obstructive disease are the two most frequent secondary causes of pulmonary hypertension in CHD patients. Atrial septal defect (ASD), persistent ductus arteriosus, and VSD are examples of common maladies. According to studies, the extent of the abnormality affects whether patients experience PAH [2].

For instance, the natural course of VSD patients reveals that 3% of individuals with mild or moderate abnormalities (1.5 cm in diameter) will go on to develop Eisenmenger syndrome. However, if there is no surgery, the condition will eventually occur in all major abnormalities (>1.5 cm). A recent article for ASDs supports the same conclusion, showing that the greatest defects (31.84 8.21 mm) were those most likely to have severe PAH. Early PAH is frequently associated with more complicated abnormalities, such as atrioventricular septal defects or truncus arteriosus. Furthermore, once the underlying heart defect has been fixed, people may still be diagnosed with PAH [3]. It's unclear whether this is because the pulmonary vascular disease has progressed despite surgical correction, although research suggests that early correction works to delay the onset of PAH in the future. Particularly, after the fontal procedure, PH has attracted a lot of interest. This procedure is the last resort for children born with single-ventricle CHD who are receiving stepwise palliation. Over the past 20 years, there have been significant advancements in early outcomes due to early surgical success, but pulmonary hypertension is still associated with late mortality and morbidity. Reduced ventricular filling and subsequently reduced cardiac output are caused by the absence of a subpulmonary ventricle, which helps to pump blood through the pulmonary vasculature. Long-term PVR to blood flow via the pulmonary system increases, although pressures are not elevated over a mean of 25 mmHg [4].

The pathogenesis is distinct and is the result of a complex web of interactions, including a lack of pulsatile flow, poor respiratory mechanics, and diminished diastolic, long axis, and systolic function of the systemic ventricles. As a result, the pulmonary circulation becomes volume-overloaded, although the PAP does not immediately rise as a result of the low-pressure system. The size of the ASD, as well as the right ventricle's compliance, is the primary determinants of the risk of developing PAH. However, left cardiac lesions and malfunction of the left ventricle can contribute. Eisenmenger rarely develops in pretricuspid lesions as evidenced by the fact that just 2% of ASD patients do.

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The left ventricle and pulmonary circulation are overloaded with volume as a result of high-pressure left-to-right shunts caused by post-tricuspid lesions. The onset of PAH happens within the first few years of life in certain abnormalities. The Eisenmenger complex, a kind of super systemic PVR, will develop in virtually all cases if this is not addressed, leading to shunt reversal [5].

Discussion

Vasoconstriction, medial wall hypertrophy, and pulmonary vascular bed remodelling are all factors in Eisenmenger's pathophysiology of PAH. The pulmonary arteries' histology in CHD-APAH reveals plexiform lesions, medial hypertrophy, intimal proliferative fibrosis, an expansion of smooth muscle cells into the peripheral pulmonary arteries, and rarefication of the pulmonary artery tree. High flow and pressure are thought to be the culprits behind the breakdown of endothelial barrier function, which in turn damages the pulmonary vascular endothelial cells. As a result, the extracellular matrix is broken down and FGF and TGF-1 are released, which activates vascular elastase and matrix metalloproteinase and causes them to work more efficiently. This release triggers the growth and proliferation of smooth muscle cells as well as the development of neo-intima. It is believed that inflammation and thrombosis result from the adhesion and activation of platelets and leukocytes following endothelial injury, with subsequent activation of the coagulation pathways. Overall, the endothelial dysfunction and subsequent pulmonary artery vascular remodelling cause elevated PVR and ultimately right ventricular failure.

Initial symptoms are frequently brought on by exertion and are connected to advancing right ventricular failure. There may be weariness, angina, syncope, and shortness of breath. With studies demonstrating that >90% of patients are in WHO class II or worse and 50% express severe limits, persons with Eisenmenger complex will typically have exercise intolerance. Symptoms can be felt when you're at rest in more severe cases. Furthermore, if right heart failure worsens, traditional symptoms such abdominal distension and ankle oedema may appear. Less frequently occurring symptoms include hoarseness caused by constriction of the left recurrent laryngeal nerve linked to dilated pulmonary artery and haemoptysis related to rupture of hypertrophied bronchial arteries. Angina occurs when the left major coronary artery is compressed between the dilated pulmonary artery and the aorta, whereas wheezing occurs when the big airways are compressed. Left parasternal lift, a loud second heart sound, a third heart sound connected to the right ventricle, pan systolic murmur (tricuspid regurgitation), and diastolic murmur are all clinical indicators of pulmonary hypertension (pulmonary regurgitation). Hepatomegaly, ascites, peripheral edoema, and cold peripheries may be present if PAH has progressed, as well as elevated jugular venous pressure. The appearance of clubbing, hepatic and renal failure, ischemic consequences, and the signs of endocarditis should all be noted because patients who are cyanosed frequently exhibit these symptoms.

Electrocardiography (ECG), chest radiography, peripheral oxygen saturations, objective assessment of exercise tolerance, and echocardiography are a few of the fundamental tests that should be performed first. The right ventricle and pulmonary vascular bed can then be studied further using chest computed tomography (CT) and magnetic resonance imaging (MRI). Electrocardiography it should be made clear that while an electrocardiogram can show signs of PH, a normal ECG does not rule out the diagnosis. Right ventricular strain and hypertrophy, right bundle branch block, right axis deviation, and right atrial hypertrophy are all possible CHD-APAH findings. The highest amplitude of the S wave in V5 or V6 and the R-wave amplitude in V1 can be added to determine the latter. There is evidence that this could offer specific predictive information for Eisenmenger complex patients. RV strain is more sensitive than RV hypertrophy in demonstrating PH, according to studies. The mean frontal QRS axis may be rightward even under normal circumstances in patients with complicated CHD, which is especially true. X-rays of the chest Pulmonary artery dilatation, aneurysms, or calcification are chest radiographic findings in CHD-APAH patients. The cardiothoracic ratio can be determined, and there may also be right atrial and/or right ventricular hypertrophy. Pulmonary venous congestion is a symptom of left heart disease. Additionally, consolidation brought on by infiltrates or pulmonary bleeding may occur. Despite this, many radiographs taken of these patients may be normal. testing of exercise This can be accomplished through either a 6-minute walk test or cardiopulmonary exercise testing with peak oxygen consumption monitoring. Both are frequently used to measure CHD-APAH, and both a decrease in distance in the former and a decrease in peak oxygen in the latter have been linked to a worse prognosis in these patients. Additionally, evidence of desaturation during exercise suggests that the shunt may be reversing, and evidence of probable right-to-left shunting in ASDs suggests a likely increased PVR and aids in the prediction of survival.

ICHD-APAH should be treated in specialised centres that see CHD and PH patients on a regular basis. A number of reviews in the UK and other countries are helping to clarify the standards that apply to CHD-PAH centres. Furthermore, patient education, awareness of potential risks and complications, and behavioural changes are critical in the care of these patients. Strenuous exercise is discouraged, but light activities are beneficial. Patients can experience clinical deterioration at any time during their treatment. These include dehydration, lung infections, high altitudes, and noncardiac surgery requiring general anaesthesia. Since pregnancy carries a high risk for both the mother and the foetus, effective contraception is crucial. Endothelia receptor antagonist users should use dual contraception due to the interaction of contraceptives with progesterone-based substances.

Diuretics are typically used for any fluid build-up during CHD-APAH therapy. Treatment for hepatic congestion, ascites, and peripheral oedema is symptomatic because this is a late manifestation. In real life, arrhythmias are the only situation where digoxin is employed. There is relatively little research supporting the use of oxygen treatment, which is basically only indicated for patients who have low night-time oxygen saturation levels, particularly in the presence of airway obstruction or coexisting long illness. Anticoagulants and calcium channel blockers are not recommended in CHD. Anticoagulation treatment specifically increases haemoptysis-related mortality in CHD-APAH patients. Calcium channel blockers that do not contain nondihydropyrodine are not advised since they are negatively inotropic.

Conclusion

Through ongoing research and teaching, it is possible to expect earlier detection and intervention of CHD-APAH given improvements in morbidity and mortality from early surgical intervention. Uncertainty exists on whether this will lessen the condition's prevalence. Future improvements in survival may result from on-going studies into the treatment of PH, particularly in patients with CHD [3,4,6]. For instance, trials for the novel dual receptor antagonist macitentan are now being conducted to evaluate its effectiveness in treating basic Eisenmenger syndrome. Most people tolerate it well, and there are positive signs about oxygen saturations and effectiveness. Future study can serve to provide greater understandings of disease processes in PH linked with CHD and hence lead prospective treatments. In addition, the pathophysiology of the impacts on lung function is currently unclear.

Conflict of Interest

None.

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