MINI REVIEW

Hypertension of the pulmonary arteries in children with sickle cell anemia

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ABSTRACT

Pulmonary Arterial Hypertension (PAH) causes the right ventricle to be strained, increasing the risk of heart failure in those who are affected. Its clinical appearance resembles that of many other diseases, delaying identification until the condition has progressed further. It is still one of the most common causes of death in individuals with Sickle Cell Anemia (SCA) all over the world. Even with a moderate rise of pulmonary artery pressure, it confers a considerable risk of death, with two-year mortality rates as high as 40%-50%. The median age of survival after being diagnosed with the condition is 25.6 months. Early detection of high pulmonary artery pressure in children and appropriate action, such as anti-hemolytic drug optimization, may help to avoid this problem from progressing.

Key Words: *Sickle cell anemia; Pulmonary artery hypertension; Children*

INTRODUCTION

Previously thought to be a rare consequence of Sickle Cell Disease (SCD), Pulmonary Artery Hypertension (PAH) affects a-out one-third of individuals with SCD. Much less is known about the frequency and natural history of the disease in children all across the world. Scientists conducted the first prospective study, which included 75 children aged six and up (mean age range of 9.41 years), and discovered a 30% prevalence. This is comparable to what was discovered in adults [1]. In contrast to another prospective study conducted by other scientists, a multicenter study conducted on 310 SCD patients aged three to twenty years found a prevalence of 11% (one case seen in a three-year-old patient with SCD), which was quite similar to that found in another prospective longitudinal study of 160 subjects aged three to twenty years where a prevalence of 14.1% was found. They both came to the conclusion that PAH in individuals with SCD starts early in life (as early as the third year of life) and increases with age. However, the results of a few small screening studies, a combination of prospective and retrospective investigations, and an aggregation of screening results from over 600 children with SCD revealed a prevalence of 35%, roughly identical to 32% in adults with SCD [2]. A study did a subgroup analysis on 208 consecutive SCD patients, aged ten to 52 years in steady-state to evaluate the prevalence of pulmonary artery hypertension and found a prevalence of 25% in West Africa, where the burden of sickle cell disease is highest. Despite acceptable laboratory data demonstrating an increase in hemolysis in patients, this finding is minimal when compared to the first prospective trial. According to the researchers, the lower prevalence identified in the study could be attributable to a fall in the older age group of patients used in the Nigerian study (7%) compared to the US-based study (46%). They linked this to the fact that SCD patients in West Africa have a lower life expectancy than those in the United States [3]. The prevalence of pulmonary hypertension was determined to be 26.2% in 44 adolescents who had outpatient echocardiography. The prevalence of sickle cell disease was determined to be 16% in a case comparison of echocardiograms on sickle cell disease patients and healthy controls. However, because these investigations were retrospective and only eligible patients were enrolled, they were prone to bias. As can be seen from the above, socioeconomic considerations may have a significant impact on the differing prevalence rates of pulmonary artery hypertension around the world. Even though the prevalence of PAH appears to rise with age in industrialized countries, as evidenced by studies from the United States, the opposite may be true in our setting, where patients with SCD have a shorter life expectancy.

Physiology of Pulmonary Artery Pressure

Each stroke volume delivers cardiac output to the lungs. The pulmonary circulation, which transports blood into the pulmonary microcirculation, is generally a high-flow, low-resistance system. The blood pressure in the pulmonary artery is measured by pulmonary artery pressure [4]. The typical range for mean pulmonary artery pressure is 8 mm-20 mm of mercury. When the pulmonary artery pressure exceeds 25 mmHg at rest and 30 mmHg during activity, pulmonary arterial hypertension is considered to have started. Except during infancy, this term applies to both adults and children [5]. There are a variety of other reasons for high pulmonary artery pressure, including the following:

- SCD Thalassemia, Hereditary Spherocytosis, and Paroxysmal nocturnal hemoglobinuria are examples of hereditary hemolytic anemia.
- Cirrhosis and portal hypertension are two hepatic disorders.
- Systemic Lupus Erythematous (SLE) and rheumatoid arthritis are both collagen vascular disorders.

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- Those with left to right shunting of blood at the atrial, ventricular, or great vessel level, such as ventricular septal defect, atrial septal defect, atrioventricular septal defect, and patent ductus arteriosus, have higher pulmonary blood flow.

Clinical Presentation of Pulmonary Artery Hypertension

Pulmonary artery hypertension frequently goes unnoticed for a long time. PAH symptoms in children vary, with symptomatology varying depending on the etiology and severity of the disease, as well as the patient's age. In babies, symptoms may include a lack of appetite and slow growth, while older children may have nausea, vomiting, lethargy, or excessive syncope. The most prevalent sign of pulmonary hypertension in people with SCD is increasing dyspnea when they exercise [6-8]. Clinicians may have mistakenly attributed this to anemia, and cardiovascular examination may have been delayed as a result [9]. Digital clubbing is one of the symptoms produced in affected patients. On cardiac examination, a loud P2 (ejection systolic murmur loudest in the pulmonary area, also known as the Graham Steell murmur) may be heard, and as the disease progresses, signs of right-sided heart failure such as an elevated Jugular Venous Pressure (JVP), hepatomegaly, ascites, pedal and peripheral oedema may be seen.

Diagnosis of Pulmonary Artery Hypertension

Cardiac catheterization is the gold standard for assessing Pulmonary Artery Pressure (PAP). However, because this method is so invasive, it is not appropriate for screening purposes [10]. Doppler echocardiography, on the other hand, is extremely sensitive and painless. In patients with SCD, the use of echocardiography to determine pulmonary artery systolic pressures has been thoroughly validated, and non-invasive assessment corresponds well with right heart catheterization measurements of pulmonary artery pressures.

CONCLUSION

During echocardiography, numerous measurements of TRV on multiple perspectives are taken to achieve an ideal image of TRV (apical 4 chambers, parasternal short axis, and parasternal long axis). To ensure accuracy, the predicted TRV is calculated by taking the average of these values.

The modified Bernoulli equation is then used to calculate the pulmonary artery systolic pressure.

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