

Prevalence of Pulmonary Arterial Hypertension among Children with Rheumatic Heart Disease Attending Follow up at the Cardiac Clinic at Teaching Hospital in Ethiopia

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Keywords: Rheumatic heart disease, Pulmonary hypertension

Background: There is general scarcity of published data related to childhood RHD in the African countries that are most affected by the disease. This study aimed to describe prevalence, associated comorbidity and treatment of PAH among children with RHD and to explore the relationship between the severity of valvular dysfunction, age and heart failure at presentation with PAH.

Method: A cross sectional study was conducted by including patients who were 18 years and less and had been diagnosed with rheumatic heart disease and have Echocardiography report from January 2014 to August 2017 at Jimma University Medical Center, Jimma, Ethiopia.

Results: A total of 247, (41.7% male and 58.3% female) medical record of children with rheumatic heart disease were reviewed. The average age of children at first follow up was 10.48 ± 2.98 years. 168 (68%) of the children came from rural, the rest were from urban areas. Mitral valve involvement was seen in almost all cases of the children; accounting 99.5%, while combination of mitral and aortic valve involvement seen in 150 children (60.7%).

Pulmonary hypertension associated with rheumatic heart disease occurred in 149 (60.3%) of the children on follow up. Among those who developed PAH 86 (57.7%) had mild PH and the rest 63 (42.3%) was found to had severe PH. 88 (59.1%) of the children who had PH were females and 99 (66.4%) were from rural areas. The average TR pressure gradient among those who developed PH was 51.17 ± 18.67 mmHg. The average age at the diagnosis of PH was 10.73 ± 2.837 years. The prevalence of PAH is affected by the age of the children at presentation when corrective treatment is not done, and the study showed this association with p value of 0.007.

Most of the children, 103 (69.1%) had heart failure at presentation. Among those children with severe PAH, 45 (71.4%) had heart failure at time of presentation. However, this was seen in only 23 children (23.5%) who had not PAH. And there is a statically significant association between PAH and HF at first time of presentation to the hospital ($p < 0.001$).

All patients were taking Byzantine penicillin G as a secondary prophylaxis, on monthly base. Despite this there were a total of 45(18.2%) rheumatic

recurrence cases from the study population and among this 32 (71.1% of all the rheumatic recurrences) was seen in those children having PAH. All associated comorbidities were more prevalent in children with PAH than those without PAH. Malnutrition 39 (26.2%) being the commonest associated illness, followed by pneumonia 22 (4.8%) and rheumatic recurrence 14 (9.4%) as a single entity. 34 (22.8%) of children had more than one comorbidity at a time or in separate times. And only 24 (16.1%) of the children with PAH - RHD were found to have no associated illness.

Mitral regurgitation was the most common valve lesion associated with PAH as an isolated or mixed involvement. Mixed Mitral and Aortic valve involvement was seen in 93 (62.4%) of children, there were 137 (91.9%) cases of MR among those who developed PAH. 32.8% of children with mild MR developed PAH, while 80.6% of children with severe MR developed PAH, and this was found to be statistically significant ($p \leq 0.001$). Aortic regurgitation was evident in 96 (64.4%) of children with PAH. 52.4% among Mild AR, 80% among Moderate AR and 78% among severe AR had PAH and there was significant association between the severity of AR and the prevalence of PAH ($p=0.001$). Mitral stenosis was found in 59 (39.5%) of children with PAH. Of those children with Mild MS 56.2%, Moderate MS 90.9 and all with Severe MS has developed PAH and this was also statistically significant ($p \leq 0.001$).

Medications were almost the only treatment modality, given to 147 (98.6%) children and a single child got a surgical repair. According to the standard treatment guidelines for patients with RHD, which is surgical management of the involved heart valve either by repair or prosthetic replacement, the practice in the study population was found to be almost absent.

Generally, the study has identified the higher prevalence of PAH among children with RHD, which make them more susceptible to acquire heart failure and associated comorbidities. The involvement of heart valves is similar with what is known from studies done previously and severity can greatly affect the presence of PAH. However, the practice of the definitive management for children with RHD is poor and needs a better attention to prevent the occurrence of PAH.

Limitation of the Study: The biggest limitation of this study was the retrospective design - this is inevitably complicated by some incomplete data and lost records.