"Comment on: Pulmonary hypertension screening in Children with sickle cell disease"

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To The Editor ,

We have read the article "Pulmonary hypertension screening in Children with sickle cell disease" by Kok Hoe Chan et al.1 with great interest and the work done by the authors is highly commendable. The article's concluding point is that in children with SCD, PHT symptoms are inconsistent with ECHO, NT-proBNP, or BNP outcomes. Children using hydroxyurea have a low incidence of PHT based on TRV, hence screening may not be recommended for this demographic. However, it would be an honor to draw your attention to a few concerns that we possess.

Firstly, the author refrained from mentioning the impact of certain genes on pulmonary hypertension in sickle cell disease as mentioned in a study conducted in 2016 2 which shows a decreased expression of the MAPK8 gene contributes to the pathogenic progression of precapillary pulmonary hypertension in SCD patients, and single nucleotide polymorphisms in ADRB1, ACVRL1, and BMP6 associated with PHT in patients with SCD.[3]

Second, the essential elements of the pathophysiology underlying vaso-oclusive crises and pulmonary hypertension in SCD patients are hemolysis and nitric oxide deficiency, both of which are significant in disrupting endothelium, endothelial proliferation, and platelet activation, they have not been highlighted in this study.[3]

Moreover, considering the fact that the author has highlighted the crucial PHT biomarkers NT-proBNP, BNP, and ECHO, a study conducted in 2020 reveals additional potential biomarkers, such as Factor-15 (GDF-15), a cytokine overexpressed in hypoxic conditions including PHT, and superoxide dismutase, a mitochondrial enzyme also upregulated in PHT, can also be utilized for diagnosing PHT in patients with SCD.[4]

Lastly, the administration of vasodilators, oxygen therapy, iron chelation, and stem cell transplant are among the treatments for PHT that the author could have discussed for a better understanding of the topic.[5]

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