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Lung function tests in children with SCD

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Abstract-*Background: Hemoglobin S(HbS)occurs due to one base pair change which encodes valine instead of glutamine in the 6th position in the beta-globin molecule. HbS cells change from a normal biconcave disc to a sickled form with resultant decreased deformability in deoxygenated conditions, which leads to occlusion of microvasculature followed with infarction, dysfunction and pain. The sickling phenomenon is exacerbated by hypoxia, acidosis, increased or decreased temperature and dehydration. Pulmonary function tests help in analysing the status in both physiological and pathological conditions.*

Objectives: To study the incidence of lung abnormality in sickle cell children & the type of lung disease- obstructive or restrictive. Methodology: The study is being conducted bedside with the help of MIR spirometer at AVBRH hospital, Sawangi on children with SS/AS pattern sickle cell disease/anemia from age 4-16 years.

Results: the results would be undertaken with SPSS. Conclusion- will be based on the findings of our study.

Keywords: *sickling, vaso-occlusive crisis, lung disease*

I. Introduction

Background/rationale: Hemoglobin S(HbS) occurs as base pair change encodes valine instead of glutamine in the 6th position in the beta-globin molecule. HbS cells undergo a transition from a biconcave disc to a sickled RBC which has diminished ability to get deformed in deoxygenated conditions, followed by an obstruction of vessels followed by infarction, poor functionability and tenderness. This is triggered by hypoxia, pH<7, fever and fluid loss [1].

Respiratory events lead to morbidity and mortality in sicklers . Clinically,this occurs in forms: Acute Chest Syndrome(ACS) and sickle cell Chronic Lung disease(CLD) [2].

Manifestations of ACS include: rise in body temperature,pain over the chest and abnormal chest xray. Sickle cell CLD, leads to radiographic interstitial pathologies, impaired functionability of lungs and in its most severe form, rise in pulmonary artery pressures. Attempts are being made in studying the pathophysiology and treatment of these events. During steady state sickle disease, the major change in lung function is restriction to ventilation , leading to a fall in Total pulmonary capacity (TLC) and decreased diffusion capacity for CO [3].

Though the development of chronic pulmonary disease in SCD has not been explained well, persistent microvascular blockage leading to pulmonary hypertension, endothelial changes and parenchymal fibrosis are the supposed mechanisms.

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Objectives: To study the incidence of lung abnormality in sickle cell children and the type of lung disease(if present)-obstructive or restrictive.

II. Methods

Study design: Prospective observational study

Setting: The study is being conducted at a super speciality hospital in Maharashtra, India which is a sickle belt from August 2018 to September 2020.

Participants: Inclusion criteria- children with sickle cell disease(SS/AS) pattern) age group 4-16 years

Exclusion criteria- Any child who has an e/o URTI 2 weeks back, children with a primary lung pathology like bronchial asthma, chronic bronchitis, and COPD

Variables: Cases of sickle cell anemia/disease aged 4 years to 16 years will be included in this study. A written informed consent will be taken by the parents and then enrolment will be confirmed. The children will be undergoing spirometry (FEV1,FVC, FEV1/FVC,PEFR).

Based on their values the disorders will be classified as restrictive or obstructive-

1.Physiological : Forced expiratory volume in one second, Forced Vital Capacity within the normal range (minimum 80% of expected) with a ratio of minimum 70%.

2. Obstruction : A ratio less than 70%, associated with low individual parameters (less than 80% expected).

3. Restrictive: (1) Decreased individual values (not more than 80% of expected) with a normal ratio (at least 70%), suggestive of low lung volumes

4. Mixed: the ratio is reduced.

Bias: children less than 4 years old were not included as they are not fit to undergo pulmonary function tests.

Variables: the normal values of the lung function parameters were compared to the standard values for given age and sex. Children were investigated for a primary lung pathology also with chest xray

Study size: Calculated sample size for sample comparison if mean to hypothesised value

Test ho m=79.4, where m is the mean in the population

Assumptions: alpha 0.0500 (2 sided)

Power 0.9000

Alternative m=74 sd=13.9

(PEF,% of predicted value :79.4 +/-13.9)

Estimated sample size needed n=70

Expected Outcomes/Results

Participants: Inclusion criteria- children with sickle cell disease/anemia(SS/AS) pattern) age group 4-16 years

Exclusion criteria- children with a primary lung pathology like bronchial asthma, chronic bronchitis, and COPD

Non-participation was due to development of cough and deterioration at each stage.

Descriptive data: participants were residents of Maharashtra; aged between 4 to 16 years. They were diagnosed cases of sickle cell anemia/disease (AS/SS) on folic acid,zinc and having frequent admissions(SS) for crisis .

Expected outcomes

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Functional ability of lung is impaired in children with SCD. Restrictive changes may become more evident with age.

III. Discussion

There may be no abnormal PFT's when the crisis incidence was less than 2 episodes in a year. There are few cases of obstruction observed. Airway obstruction in 0.05 to 0.35 % of population with sickle cell disease and restrictive function in 0.08 to 0.22 children. Rest of the researchers have claimed that restrictive changes become pronounced with advancing age in sickle cell disease [1] and its prevalence is as high as 74% among adults [2]. Initially, obstructive changes take place followed by of restrictive changes [3]. Our findings will help us in saying that lung function testing should be done in children with hemoglobin SS. Studies in the future should understand the cause of obstruction and restriction , the morbidities linked with these changes and the risk of these changes is soaring with age.[4] A number of related article son different aspects of this study were reviewed [5-55].

Limitations: We had used a bedside spirometer to evaluate the children before and after nebulisation with an inhaled beta agonist. We did not assess the DLCO and TLC. All parameters were not assessed, prime importance was given to FEV1/FVC ratio.

Interpretation: Lung abnormalities in sickle children increases with episodes of vaso-occlusive crisis. Monitor their lung function tests who have frequent hospital admissions for crisis and transfusion.

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