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Hypothetical Article

Rapidly Progressive Acute Chest Syndrome in Sickle Cell Anemia Patients in King Abdulaziz Hospital, Jeddah

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Introduction

Based on existing literature. State hypotheses:

One of the most common genetic disease globally is Sickle cell disease [1]. It is characterized by abnormal Hb production i.e HbS which cause RBCs to sickle and occlude small vessels [2]. The prevelance of the disease in Saudi Arabia differs by region being the highest in the Eastern provinces followed by southwestern provinces [2]. In Saudi Arabia the disease is one of the leading causes of morbidity [3] complications of sickle cell disease include Recurrent episodes of vaso-occlusion and inflammation result in progressive damage to most organs, including the brain, kidneys, lungs, bones, and cardiovascular system, which becomes apparent with increasing age [4]. The acute chest syndrome (ACS) is one of the most frequent complications of sickle cell disease [5]. ACS It can present as a complication of acute VOC or arise de novo on its own upon admission [6]. The etiology of ACS is unknown but it is frequently precipitated by a vaso-occlusive crisis, infection, fat embolism, hypoventilation and atelectasis (from any cause) [6,7]. Several risk factors that have associated with increased incidence of ACS including age less than 5 years, living in rural area, history of previous hospital admission; white blood cell count greater than 10,000/dL; hemoglobin concentration less than 7 g/dL and oxygen saturation ≤ 95% on admission. After controlling for confounding in multivariate logistic regression, only a history of previous admission

remained as an independent predictor of ACS (relative risk=4.20; 95% confidence interval: 1.79-9.87; P=0.001) [8]. There are multiple definitions of ACS one of which is Vichinsky *et al* that defines ACS as : A new pulmonary infiltrate involving at least one complete lung segment that is consistent with the presence of alveolar consolidation but excluding atelectasis, Chest pain, tachypnea, wheezing or cough, temperature higher than 38.5C [9,10]. But these definitions have not described the temporal relationship onset of ACS and the sudden decline in respiratory status [11]. However a recent study published in American Journal of hematology introduced a new term of Rapidly progressive ACS as a subtyphenotype of ACS. They tested the hypothesis that in patients with SCA, rapidly progressive ACS characterized by respiratory failure (requiring at least 3L of oxygen or intubation and mechanical ventilation to maintain oxygen saturation ≥ 90%) within 24 hours of onset of respiratory symptoms exists as a distinct phenotype and is associated with multi-organ failure.

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