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POSTER ABSTRACTS

114.SICKLE CELL DISEASE, SICKLE CELL TRAIT AND OTHER HEMOGLOBINOPATHIES, EXCLUDING THALASSEMIAS: CLINICAL AND EPIDEMIOLOGICAL

Provider Perspectives on Diagnosis and Management of Acute Chest Syndrome

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Introduction: Acute chest syndrome (ACS) is the leading cause of mortality, accounting for 25% of all deaths among individuals with sickle cell disease (SCD). The etiologies and clinical manifestations of ACS are variable, with a lack of clear risk stratification guidelines for the practicing clinician. Management of ACS is based on limited evidence and is currently guided primarily by expert opinion. Based on this gap in the diagnosis and management of ACS, we created a multicenter ACS work group in March 2022 that included pediatric and adult hematologists, and transfusion medicine physicians. We hypothesized that the risk stratification and management of ACS is significantly variable among providers involved in the care of SCD individuals.

Methods: The ACS work group created a survey to determine how providers from different subspecialty groups diagnose and manage ACS. The survey was anonymous. Five ACS diagnostic features had to be rated from 1 to 5 in addition to 3 case scenarios to reflect mild, moderate, or severe ACS events as determined by the work group. The study was IRB approved and distributed through RedCap to providers from adult and pediatric hematology/oncology, transfusion medicine, emergency medicine, critical care and pulmonology during a one-month period in June to July, 2023.

Results: Out of 465 providers who have received the survey to date, 161 providers have responded (34.6%). Of these, 103 (64%) reported caring for over 50 SCD individuals while 38 (24%) reported caring for >200 SCD individuals in the year prior. Most providers (68%) were from pediatric specialties (110/161) while 20% (32/161) were from solely adult specialties. One hundred and twenty-four (77%) of respondents were hematologists. Sixty-four percent (n= 103) of the providers rated a new pulmonary infiltrate as one of the top 2 criteria to diagnose ACS. However no other criteria for ACS such as hypoxia, lobar pneumonia, fevers, drop in hemoglobin of more than 2 g/dL from baseline, were rated as the top 2 criteria by more than 40% of the providers.

For the case scenarios, the case deemed moderate ACS by the work group received the most discordant results about its stratification and management. Although most respondents considered it ACS (93%), 42% graded it as mild ACS (63/150) and 31% graded it as moderate ACS (46/150). For the management of this case, transfusion practices varied with 28 providers choosing 10-15 mL/kg packed red blood cells (pRBCs), 67 choosing pRBCs to goal hemoglobin of 9-10 g/dL and 10 providers choosing RBC exchange transfusion (RBCX). For all the case scenarios, a mean of 37 providers (24%) selected that grading for ACS is not important as the patient had ACS regardless of the grading. In the case deemed mild ACS, transfusions were selected by 4% (n = 6) of the providers choosing 10-15 mL/kg of pRBCs, and 7.5% (n =12) providers choosing pRBCs to goal hemoglobin of 9-10 g/dL.

Conclusion: We conclude that diagnosis and management of ACS is highly variable, even among experienced specialty providers. Further research is needed to improve standardization of ACS risk stratification and management to optimize care for individuals with SCD.

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