ID - 2368

PROGNOSTIC MARKERS IN STROKE IN SICKLE CELL ANEMIA PATIENTS

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Introduction: Sickle cell anemia (SCA) is an autosomal recessive condition that presents with a wide variety of clinical manifestations. Among them, stroke is one of the most debilitating pathologies. **Objectives:** To analyze the potential associations between laboratory markers and the incidence of stroke in patients with sickle cell anemia (SCA) treated at the Hematology and Hemotherapy Foundation of Amazonas (HEMOAM). **Material and methods:** The study employed a cross-sectional observational design, encompassing a cohort of patients of both sexes, with diagnoses substantiated by the HPLC technique. The technique employed for the diagnosis of stroke was transcranial Doppler (TCD). The laboratory data were obtained through access to the patient's medical records. **Results:** A total of 274 patients participated in the study. In patients who developed stroke, there was a significant increase in direct bilirubin (DB) ($p < 0.001$), HDL ($p = 0.03$), iron ($p < 0.001$), ferritin ($p < 0.001$), and HBA1 ($p < 0.001$) when compared to those who did not develop stroke. **Discussion and conclusion:** Studies have shown that about 10% of SS patients have experienced a stroke. Studies show that up to 20% of patients over 19 years old and up to 50% of patients over 45 years old have strokes. The study found higher levels of BD, iron, ferritin, and HBA1 in people with stroke. This is similar to what other studies have found. These studies suggest that higher levels of these substances are linked to severe hemolysis. Hemolysis is when there is too much red blood cell destruction. This can lead to more frequent blood transfusions. But this is the first time that really high HDL levels have been linked to stroke. Studies usually show that SS patients have lower levels of HDL. It is important to know that doctors are using a treatment called hydroxyurea together with red blood cell transfusions at least once a month. This treatment is currently being used in blood centers to help patients keep a hemoglobin concentration of at least 9 g/dl and an HbS fraction of less than 30%. This helps reduce the number of stroke cases in patients. Stroke has been shown to have a devastating impact on patients with atrial fibrillation (AF), often resulting in increased mortality, disability, and loss of neurocognitive function. The implementation of effective screening tests and continuous transfusion therapy has led to the prevention of several clinical complications. The multifaceted nature of stroke risk, encompassing a constellation of risk factors and potential complications, is well-documented. Although stroke is more prevalent among adults, the risk is particularly elevated in children and in individuals who do not undergo comprehensive monitoring. Appropriate management, with an emphasis on early diagnosis and rigorous monitoring, has been demonstrated to significantly improve quality of life and reduce mortality among sickle cell disease patients.

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REAL-WORLD SAFETY PROFILE OF TWICE-DAILY DEFERIPRONE FOR IRON OVERLOAD IN US PATIENTS WITH THALASSEMIA, SICKLE CELL DISEASE OR OTHER ANEMIAS

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Introduction: Iron overload is a significant cause of transfusion-related morbidity and mortality. Deferiprone (DFP) is an oral iron chelator with > 28 years of safety and efficacy data. Initially approved for administration three times a day (TID), a twice-a-day (BID) formulation was developed to improve patient adherence. The United States (US) Food and Drug Administration (FDA) approved DFP BID for transfusional iron overload in patients with thalassemia in 2020, and DFP TID and BID for sickle cell disease (SCD) or other anemias in 2021. **Objectives:** To evaluate the real-world safety profile of DFP, the Ferriprox® Total Care Registry was established in the US. Here, we assess the safety of DFP BID in patients with thalassemia, SCD, or other anemias in real-world clinical practice. **Material and methods:** Data were obtained from US patients receiving DFP BID between July 1st 2020–August 31st 2023. DFP exposure: time active in the registry. Frequency of adverse events (AEs) and serious AEs (SAEs) by Medical Dictionary for Regulatory Activities (MedDRA) Preferred Term was assessed. **Discussion and conclusion:** A total of 425 patients were referred to the Ferriprox® Total Care Registry. Primary

diagnoses included “thalassemia” (n = 133, 31.3%), “SCD” (n = 197, 46.4%), “other anemias” (n = 30, 7.1%), and “other indications” (n = 65, 15.3%). Overall, 364 patients (85.6%) were ≥ 18 years of age. An estimated total exposure of 839 patient-years was observed in 412 patients who received ≥ 1 month DFP BID treatment. In total, 1250 AEs and 437 SAEs were reported. All reports of agranulocytosis (2 events) and neutropenia (7 events) recovered. None of the 33 fatal outcomes reported were assessed as possibly or probably related to DFP treatment. DFP BID is well-tolerated in real-world clinical practice in patients with thalassemia, SCD, or other anemias, with no new safety concerns identified compared to DFP TID. Limitations associated with real-world data collection must be acknowledged. Disclosures and financial support This project was funded by Chiesi USA, Inc. Medical writing support was provided by Cactus Life Sciences®, and funded by Chiesi Ltd.

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REFRACTORY AUTOIMUNE HEMOLYTIC ANEMIA AND RECURRENT THROMBOEMBOLIC EVENTS: A CASE REPORT

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Introduction: Autoimmune hemolytic anemia (AIHA) is a rare disease with an incidence of 0.2 per 1,000,000 individuals under 20 years of age. Patients with AIHA have a higher propensity for thrombotic events and other autoimmune disorders. Studies demonstrate that 11-20% of that population may develop thromboembolic events during anemic episodes. **Case description:** Male, 71 years old, diagnosed with primary AIHA in August/2023 after ruling out secondary causes. In December/2023, he presented spontaneous pulmonary thromboembolism (PTE) as the first thrombotic event. Anticoagulation was started with rivaroxaban for 6 months until July/2024, when it was discontinued due to gastrointestinal (GI) bleeding without identifiable lesions on colonoscopy. The patient has maintained AIHA treatment with prednisone since diagnosis. In February/2025, after 6 months without anticoagulation, he complained of exertional dyspnea and fatigue for 3 months, presenting markedly elevated D-dimer (5,873 ng/ml; NV < 500 ng/ml). Due to clinical suspicion of PTE, he was referred to the emergency department, where full anticoagulation with enoxaparin was initiated. Chest CT angiography confirmed filling defect at pulmonary artery bifurcation. During hospitalization, there was progressive hemoglobin (Hb) decline requiring increased corticotherapy from 15 mg/day to 100 mg/day. On the 10th day, pulsotherapy was performed as a result of his low level of Hb (5.8 g/dl) despite optimized medications. After 2 days, he developed hematochezia episodes maintained even after enoxaparin discontinuation. On the 17th day, due to pulse therapy-refractory AIHA, rituximab 375 mg/SC was prescribed, prednisone returned to 100 mg/day, and an inferior vena cava filter was placed. After 26 days, Hb stabilized at 9.5 g/dl, colonoscopy showed no abnormalities, and he was discharged with prednisone 60 mg/day and apixaban. During outpatient follow-up, he remains stable and without new bleeding episodes, with Hb at 12.3 g/dl. DISCUSSÃO: Several mechanisms justify thrombotic predisposition in AIHA: erythrocyte membrane alteration by autoantibodies with coagulation enzymatic complex formation; red blood cell surface destruction; free Hb from hemolysis increasing adhesion molecule expression; pro-inflammatory effects of free heme, among others. Additionally, glucocorticoid therapy also contributes to prothrombotic state in these patients. Anticoagulant prophylaxis, although not recommended by guidelines and lacking randomized clinical trials for validation, has proven useful in studies with hospitalized patients. In this case, the patient had a thrombosis history, but anticoagulation discontinuation became necessary due to GI bleeding without apparent cause on colonoscopy, thus attributed to rivaroxaban. Meta-analyses indicate greater association of rivaroxaban with GI bleeding compared to apixaban, which has a more favorable safety profile, justifying the medication switch at discharge. **Conclusion:** Despite being rare, AIHA is a severe disease with considerable morbimortality, deserving attention for early diagnosis and treatment. In the future, randomized clinical trials should be proposed to validate the benefit of thrombotic event prophylaxis in this patient profile, ensuring better quality of life and reduced morbimortality.

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RELAÇÕES ENTRE CINESIOFOBIA E PARÂMETROS CLÍNICOS EM PACIENTES COM DOENÇA FALCIFORME

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Introdução: Uma proporção significativa de pacientes vivendo com Doença Falciforme (DF) apresenta dor em mais de 50% dos dias. Uma abordagem biopsicossocial reconhece a influência de fatores sociais, psicológicos e comportamentais na experiência da dor, assim como os desafios no seu manejo considerando sua natureza multifatorial e subjetiva. Componentes físicos e psicológicos, como a cinesiofobia – conceituada como medo excessivo, irracional e debilitante de