hemorrágicas; 36 pacientes foram transplantados, sendo que 24 atingiram a remissão completa, demonstrando que o transplante é a primeira linha de tratamento da AA severa nesta faixa etária. MJ Hossain et al. avaliaram, de 2004 a 2014, 1.140 crianças com diagnóstico de AA, apontando que alguns dados clínicos estão associados à alta mortalidade na AA, como pancitopenia grave e contagem de plaquetas menor que 50.000. Apesar de Kojima S. et al. descreverem uma alta incidência de GVHD em 29% dos pacientes que sobreviveram 2 anos após o transplante, o mais difícil no tratamento de anemia aplástica foi o grupo de pacientes que não respondeu à imunossupressão, com evolução de sangramentos e sepses, sendo o transplante a terapia salvadora. Em outro estudo, Holmqvist et al. demonstraram uma taxa de mortalidade alta nas crianças que foram submetidas ao transplante e que permaneceu elevada até 15 anos após o transplante, associada ao alto risco de desenvolver GVHD. **Conclusão:** A taxa de óbito na anemia aplástica na infância, no Hemorio, no período analisado, foi de 18,7%, um pouco superior que a do Hospital da Criança da Universidade de Toronto que foi de 13%.

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## FUNÇÃO SEXUAL EM PORTADORES DE ANEMIA FALCIFORME DO SEXO MASCULINO - ESTUDO TRANSVERSAL

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**Introdução:** A Doença Falciforme (DF) é um termo que abrange um grupo de anemias hemolíticas hereditárias que se caracterizam pela alteração estrutural na cadeia da beta hemoglobina. É uma enfermidade genética com grande prevalência no Brasil e que gera impacto considerável na morbidade e mortalidade da população afetada. O fenômeno de vaso-oclusão que ocorre principalmente em pequenos vasos, representa a principal causa dos sinais e sintomas dos pacientes com doença falciforme. Uma das consequências desse fenômeno é o priapismo, condição que envolve a ereção prolongada não relacionada ao interesse sexual. Quando se estende além de vinte e quatro horas ou quando ocorre recorrentemente, este evento está associado a necrose tecidual e subsequente fibrose, deixando como sequela graus variados de disfunção erétil. Esta é uma complicação comum na anemia falciforme e que traz prejuízo à atividade sexual, comprometendo a qualidade de vida. Uma importante ferramenta para avaliar esse acometimento consiste no Índice Internacional de Função Erétil (IIEF), o qual analisa aspectos relacionados à ereção, penetração, orgasmo e confiança do paciente relativa à relação sexual. **Objetivo:** Avaliar a função sexual masculina dos pacientes com diagnóstico de Doença falciforme. **Métodos:** Trata-se de um estudo observacional e transversal com pacientes do sexo masculino, diagnóstico de Doença Falciforme e em acompanhamento no ambulatório do serviço de hematologia da Escola Pauista de Medicina/UNIFESP. Os participantes foram selecionados aleatoriamente, no período de novembro de 2022 a janeiro de 2023. Foi aplicado o questionário específico e validado para avaliação da função sexual, o IIEF. A partir do IIEF foram abordados 5 domínios da função sexual do homem, a função erétil, a função orgásmica, o desejo sexual, a satisfação individual e a satisfação geral. **Resultados:** Participaram do estudo 10 homens com vida sexual ativa, com mediana de: idade - 25 anos, Hb/Ht – 9 g/dL e 25,6%, HbS – 48,5% DHL - 507,5. Destes, 57% estavam em regime transfusional crônico, 28,5% em uso de Hidroxiureia em monoterapia, e 14,5% em terapia combinada. Ademais, 90% possuíam alguma complicação crônica relacionada a DF, sendo a mais comum Úlcera de membro inferior (50%) seguido de Priapismo (30%). Após aplicação de questionário, 90% não apresentavam disfunção erétil (mediana: 28 pts), com 1 paciente apresentando disfunção erétil moderada (13 pts). Acerca dos demais domínios avaliados, foi encontrado uma mediana de 8 para função orgásmica (0-10), de 7 para desejo sexual (2-10), de 13 para satisfação individual (0-15) e de 8 para satisfação geral (2-10). **Discussão:** No presente estudo verificamos um percentual de disfunção erétil inferior ao das demais evidências (10% vs 30-40%), com possível impacto do tamanho e prevalência de priapismo na casuística avaliada. Entretanto, também identificamos uma manutenção da funcionalidade sexual nos 5 domínios em um percentual significativo de pacientes, a despeito de valores elevados de HbS (maior – 81,9%), presença de complicações ou níveis hematimétricos, o que reforça a possibilidade de promoção de uma vida sexual funcional para esse perfil de pacientes. **Conclusão:** Pelo presente estudo pode-se concluir que, a despeito da Doença Falciforme, um percentual importante de pacientes aparenta manter uma vida sexual funcional.

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## HEMATOLOGIC RECOVERY INDUCED BY ELTROMBOPAG IN PATIENT WITH APLASTIC ANEMIA AFTER TEMOZOLOMIDE

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**Objective:** Presenting a rare clinical case of a patient with Aplastic Anemia (AA) developed after treatment with Temozolomide who achieved complete hematologic recovery induced by eltrombopag. **Methods:** Retrospective descriptive study of a clinical case carried out through data collection from HSPE medical records. **Results:** A 32-year-old woman who underwent surgical resection of a low-grade glioma associated with radiotherapy (56 Gy) and chemotherapy with temozolomide (15 mg/day for 30 days) presents with pancytopenia 15 days after the end of chemotherapy. As a

consequence of the cytopenias, which were at first attributed to the toxicity of the glioma treatment, the patient had retinal bleeding, metrorrhagia and several hospitalizations due to infection. There was no improvement in cytopenias one year after the onset of the condition, with hemoglobin (Hb) 5,8 g/dL leukocytes (Lt) 1770/mm$^3$ (neutrophils (N) 549, lymphocytes (Ly) 1044) Platelets 8000/mm$^3$, bone marrow biopsy (BMB) revealed 20% hematopoietic tissue, hypocellular in the three sectors, with better representation of the erythroblastic sector, erythroblasts in cohesive clusters, granulocytes with preserved maturation, scarce small and hypolobulated megakaryocytes, and 1% lymphocytosis. Diagnosis of AA was confirmed and eltrombopag 150 mg/day was started as a single therapy. After 8 months the patient presented: Hb 13.8 g/dL Lt 3200/mm$^3$ (N1152 Ly1504) Platelets 154000/mm$^3$, with complete improvement of symptoms. BMB performed 16 months after the introduction of eltrombopag showed 40% of representative hematopoietic tissue in the 3 sectors, compatible with regeneration after bone marrow aplasia. Corticosteroid weaning was performed and eltrombopag was withdrawn with maintenance of hematological response, with no further cytopenias in the blood count. Discussion: It is known that Temozolomide therapy can lead to hematological toxicity, with thrombocytopenia, leukopenia and, more commonly, neutropenia. Myelosuppression, however, is considered dose-dependent and it is usually reversible after drug's suspension in approximately 15 days, it occur in less than 5% of patients. A current study showed that 0.6% of patients who used Temozolomide developed AA, therefore a rare complication. The causality between the therapy used and the development of AA could not be definitively established, however discontinuing treatment after one year did not lead to bone marrow recovery. It is also known that the most frequent mechanism related to acquired AA is an immune-mediated destruction of hematopoietic stem cells (HSCs) and bone marrow progenitor cells. At first, thrombopoietin (TPO) agonists were associated with megakaryocyte stimulation and platelet production. However, in vitro studies also suggest that TPO plays a key role in the proliferation and maintenance of HSCs. In the case presented here, there was complete hematological recovery with the use of eltrombopag, a thrombopoietin agonist. **Conclusion:** Despite the safety of Temozolomide and its effectiveness, there are a few cases of myelosuppression and aplastic anemia resulting from its use in the literature, such as the report presented here. The treatment of AA induced by this medication is not well defined, but it was successfully in this report using eltrombopag.

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## SEPTIC ARTHRITIS IN SICKLE CELL DISEASE PATIENTS WITH OSTEONECROSIS: A COMPLICATION TO BE REMEMBERED

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**Introduction:** Several studies have reported an association between osteonecrosis (ON) and septic arthritis, mainly in the pediatric population with sickle cell disease (SCD). However, it is unclear whether the association applies to adults. **Objective:** To report a case of femoral head ON (ONFH) in an adult with sickle cell anemia (SCA) complicated by septic arthritis. **Case report:** Male patient, 20 years-old, with SCA, on Hydroxyurea 28 mg/kg/day and folic acid 5 mg/day, baseline hemoglobin (Hb) of 8 g/dL, Hb S 80.7%, Hb A2 2.6% and fetal Hb 16.7%. He underwent attendance due to pain in the right hip and was diagnosed with ONFH, affecting over 50% of the joint, associated with joint effusion, with no orthopedic surgical indication. After 20 days of discharge, he returned due to refractory pain and he had no fever. On the 10th day of hospitalization, he underwent mechanical debridement and osteotomy. He was diagnosed with septic arthritis caused by S. aureus and S. epidermidis and received antibiotic therapy, with progressive pain improvement. Currently, he is scheduled for total hip arthroplasty. **Discussion:** ON is a common complication of SCD due to episodes of ischemia and reperfusion in susceptible joints. Clinical includes hip pain, reduced joint range, and gait alterations. The femoral region is highly affected due to its poor blood supply, susceptibility to high mechanical load, and increased joint wear propension. In patients with SCD, bacterial opsonization is dysfunctional, leading to systemic inflammation and oxidative stress, which can increase the susceptibility to infections, justifying the association of ONFH with osteomyelitis and septic arthritis, a situation more commonly described in the pediatric population and rare in adults like the presented patient. The most common microorganism found in septic arthritis of patients with SCD is Salmonella, followed by enteric gram-negatives, but in the reported case, *S. aureus* and *S. epidermidis* were isolated. Bone decompression to stimulate revascularization of the femoral head may alleviate symptoms in the early stages of ONFH. Historically, poor operative outcomes have discouraged many individuals with SCD and ONFH undergoing hip arthroplasty. If surgical management is chosen, perioperative treatment should be done with adequate hydration, oxygenation, and pain control, along with advanced surgical techniques, so it may improve quality of life. Regarding septic arthritis, surgical debridement and joint irrigation are indicated. **Conclusion:** The overlapping of septic arthritis and osteonecrosis has a clinical picture that will not always be typical in adult individuals and may result in late diagnosis and higher morbidity and mortality. The complaint of pain should not be dismissed as just another painful crisis; hematologists must take into consideration other complications associated with avascular necrosis, such as septic arthritis and osteomyelitis.

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