descompressão tradicional e/ou combinado com algum tipo de tratamento adjuvante a artroplastia de quadril ou osteotomias femorais. A utilização de terapia celular em fases iniciais do acometimento evita deformidades definitivas e uso de próteses. **Conclusão:** A NAO apresentou prevalência significativa nos pacientes adultos, sendo a média de diagnóstico aos 26,3 anos. Os exames de imagem são imprescindíveis para o diagnóstico de NAO e para a indicação do manejo adequado. O diagnóstico precoce é de suma importância para a preservação articular e para a prevenção de desfechos com grandes impactos na qualidade de vida dos pacientes com DF e NAO.

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DIAGNOSTIC ACCURACY COMPARISON OF THE QUANTITATIVE POINT-OF-CARE ECO G6PD AND BREWERS TEST FOR ASSESSMENT OF G6PD DEFICIENCY IN INFECTIOUS DISEASES

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Introduction: Glucose-6-phosphate dehydrogenase (G6PD) deficiency is an X-linked, hereditary condition caused by mutations in the G6PD gene, that results in protein variants with different enzyme activity levels. G6PD provides reducing power to NADPH (reduced form of nicotinamide adenine dinucleotide phosphate). NADPH effect is critical in red blood cells (RBC) due to the absence of a nucleus. Clinical manifestations are usually mild and are induced by an exogenous agent, such as drugs, infections, or dietary (fava beans). The G6PD deficiency diagnosis is crucial in infectious diseases because both infections and treatment can induce acute hemolysis. Rarely, life-threatening cases can require transfusions of RBC and hemodialysis. The gold standard for the measurement of G6PD activity is a quantitative UV spectrophotometric assay, however, it is inaccessible in most cases. Methemoglobin reduction test (Brewer's test) is a qualitative spot test most used for G6PD deficiency screening, but is imprecise, requires a cold chain, some laboratory infrastructure, trained personnel, and is time-costing. Point-of-care (POC) is a simple and fast semi-quantitative test that is becoming increasingly important when a prompt diagnosis is required and in health care of out-of-reach communities. **Objective:** To compare the diagnostic accuracy of the quantitative POC ECO and Brewers test for assessment of G6PD deficiency. **Material and methods:** A cross-sectional diagnostic accuracy study was enrolled between May 2020 and December 2021 at the Instituto Nacional de Infectologia Evandro Chagas, Fundação Oswaldo Cruz. Individuals older than 17 years old with a suspicious diagnosis of an infectious disease were eligible. 125 convenience blood samples were tested to compare concordance analysis between Brewers test and POC STANDARD G6PD. The exclusion criteria were hemoglobin concentration < 7.0 g/dL due to technical device limitations. **Results:** A total of 125 subjects (38 females and 87 males) were recruited. The median age was 42.0 years (range 18–76) in females and 45.0 years (range 19–78) in males. The median of G6PD activity was 7.1 U/g Hb (0.2-17.5). We observed a trend of concordance between the G6PD status (deficient or normal), with low frequencies of discordances in the screenings. The strength of agreement between G6PD tests was classified as almost perfect in all participants (k = 0.82, IC95% = 0.66,0.97) and in subgroups female (k = 0.83, IC95% = 0.59,1.00) and male (k = 0.81, IC95% = 0.59,1.00). The total concordance percentage (number of concordances/total) was 95% in females, 97% in males, and 96% in all participants. **Discussion:** Here, we demonstrate POC ECO G6PD test could be performed during medical consultation, guiding the most appropriate therapy almost immediately. In addition to saving time and having comparable results to Brewer's test, it could be performed even in remote areas and with scarce resources, dispensing minimum laboratory requirements. Our findings will be of value to diagnose conditions that could have life-threatening consequences if not timely recognized, such as before initiating primaquine for the treatment of malaria. Therefore, it can improve healthcare in vulnerable isolated communities. **Conclusion:** We observed a high concordance result between Brewers test and POC STANDARD G6PD, which is becoming the reference test to diagnose G6PD deficiency worldwide.

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RITUXIMABE COMO TRATAMENTO DA SÍNDROME HIPERHEMOLÍTICA NA ANEMIA FALCIFORME: RELATO DE CASO

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Introdução: Anemia falciforme (AF) afeta mais de 250 milhões de pessoas e clinicamente caracteriza-se pela por crises dolorosas vaso-oclusivas e fenômenos hemolíticos. Várias complicações, tanto agudas quanto crônicas requerem transfusão de sangue (complementar ou de troca). Assim, esses pacientes são propensos a desenvolver complicações transfusionais, incluindo aloimunização e, mais raramente, síndrome hiper-hemolítica (SHH). **Objetivos:** Relatar um caso bem sucedido de tratamento com Rituximabe em paciente com SHH na AF. **Relato de caso:** Homem, 35 anos, com AF interna devido a úlceras de membros inferiores infectadas. Apresentava Hb 6,2 g/dL, Ht 18,7%, VCM 95,4 fL, Leucócitos 19074/mm^3 (Neutrófilos 14687/mm^3), plaquetas 453 mil/mm^3, reticulocitos 391700/mm^3, DHL 552 U/L, Bilirrubinas totais 2,35 /indireta 1,31 U/L. Possuía antecedentes de acidente vascular encefálico isquêmico na infância, priapismo de repetição, úlceras de membros inferiores e múltiplas transfusões sanguíneas. Optado por manter Hb; 9,0 g/dL e antibioticoterapia ampla recebendo alta após 8 dias com melhora do quadro. Após 5 dias é reinternado com priapismo e queda de Hb (Hb 6,4g/dL, Ht 19%, Reticulócitos 512000/mm^3, leucócitos: 18600/

mm^3, plaquetas 538 mil/mm^3, DHL 518 U/L, TAD negativo e HbS de 24,8%. Optado por observação e IGIV sob suspeita de SHH visto queda rápida de Hb. Após 5 dias de IGIV ele recebeu nova transfusão com Hb pré de 6,8 g/L e após transfusão 5,9 g/L e HbS 52,1% confirmando o diagnóstico de SHH. Paciente recebeu rituximabe 1000 mg, 2 infusões com intervalo de 2 semanas entre as mesmas com resolução de quadro hemolítico e independência transfusional (HbA1 65,6% e HbS de 29,8%). **Discussão:** SHH é diagnosticada quando ocorre anemia grave paradoxalmente após uma transfusão de sangue (Hb pós < Hb pré-transfusão). Resulta em hemólise concomitante das hemácias transfundidas e autólogas. Acredita-se que o mecanismo subjacente, a esta rara e geralmente fatal complicação, seja secundário a resposta imune contra os fosfolipídios expostos da membrana eritrocitária. A predisposição para SHH na AF também é variada e a busca por um padrão ou valor de predição tem sido evasiva. Pode ser dividida em 2 formas baseadas no tempo de transfusão até o desenvolvimento dos sintomas e formação de aloanticorpos. Aguda: sintomas clínicos são observados dentro de 7 dias pós transfusão. Os aloanticorpos geralmente não são formados nesse período e, portanto, um teste direto de antiglobulina (TAD) para detectar aloanticorpos provavelmente seria negativo. Tardia: sintomas clínicos são observados além de 7 dias após transfusão, e formação de aloanticorpos é mais provável. O TAD neste caso provavelmente seria positivo. Neste relato, a forma aguda foi observada porque os sintomas foram observados 5 dias após a transfusão e o TAD foi negativo. O tratamento visa suporte e evitar transfusões. IGIV e esteroides em altas doses mostram bons resultados em casos leves por suprimir a ativação macrofágica, encurtando a duração da hemólise. O Rituximabe também pode ser usado nestes casos de SHH e AF, como neste relato, com bons resultados. **Conclusão:** SHH é rara e necessita de pesquisas mais extensas para determinar indivíduos com maior predisposição a essa complicação de alta mortalidade. O alto índice de suspeição pode ser fundamental no manejo bem como monitorar pacientes com múltiplas transfusões.

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TWO CASE REPORT OF PARVOVIRUS B19 INFECTION IN HIV-INFECTED PATIENTS: DIFFERENT CLINICAL PRESENTATIONS FOLLOWING ACUTE AND PERSISTENT B19V INFECTION

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Introduction: *Parvovirus B19* (B19V) infection could cause clinical manifestations depending on the host's immunologic and hematologic status. In immunocompetents individuals, clinical manifestations range from asymptomatic, or flu-like symptoms in childhood including a viral exanthem (fifth disease) to a fatal hydrops fetalis in pregnant women. B19V infect red blood cell (RBC) precursors and in immunocompromised hosts, such as HIV-infected patients, it could be manifested as pure red cell aplasia and chronic anemia. B19V may cause transient aplastic crisis (TAC) in patients with increased RBC destruction, including hereditary spherocytosis (HS). **Objective:** To report cases of B19V infection in HIV-positive adults in follow-up at Instituto Nacional de Infectologia Evandro Chagas. **Results:** Case 1: A 27-year-old black male with HIV since childhood on irregular use of antiretroviral therapy (ART) complained of vertigo, headache, vomiting, fever. In April 2021 he had Hb 1,7 g/dL, Ht 6.5%, reticulocytes 0%, HIV viral load 40,305 cps/mL and $CD4^+$ count 6 cells/μL. Up to October 2021 he required multiple RBC transfusions. Myelogram showed giants proerythroblasts with nuclear inclusions, B19V real time polymerase chain reaction (qPCR) detected 4.3×10^{10} IU/mL, and B19V anti-IgM was negative. He received intravenous immunoglobulin (IVIG) and stabilized RBC counts. Nine months after treatment, he remained asymptomatic but had 11 cells/ μL CD4+ count and 27,642 HIV cps/mL due to poor adherence to ART. B19V was still detectable in qPCR (1.8×10^6 IU/mL), but he did not receive any additional treatment because blood counts were normal. Case 2: A 61-year-old white male with HIV since 2009 on regular use of ART, undetectable HIV viral load, but historically low $CD4^+$ count, presented four weeks of lethargy, and an acute incident of dizziness, diarrhea, and syncope. He exhibited pancytopenia (Hb 5.6 g/dL, Ht 15.2%, WBC 1.48×10^9/L, and platelets 41,000 per mm^3), non-immune hemolytic anemic features, jaundice, and palpable spleen. Reticulocyte counts were unavailable. He received transfusions of RBC, the HIV viral load at admission was undetectable, and $CD4^+$ count 127 cells/ μL. The myelogram presented giant proerythroblasts with nuclear inclusions, and bone marrow had 95% cellularity and mild erythroid hyperplasia. B19V anti-IgM was indeterminate, while B19V qPCR detected 8.9×10^3 IU/mL, in August 2021. The patient received IVIG and nine months later, showed resolution of pancytopenia, but maintained elevated indirect bilirubin, indicative of chronic hemolysis. B19-DNA was no longer detected by qPCR, hematoscopy showed spherocytes, and the osmotic fragility test suggested RBC membranopathy, revealing an undiagnosed HS. **Discussion:** Anemia is a common finding in the HIV setting, is often multifactorial, and represents a significant risk factor for mortality in AIDS. Our study highlights the importance of B19V infection be considered as a part of the differential diagnosis of anemia in this group. Clinicians should search for typical bone marrow and virologic findings. Identification of B19V DNA by qPCR was essential herein because of low sensitivity of serologic tests. **Conclusion:** Our findings showed that adherence to ART and the status of immunosuppression were crucial to the parvovirus clearance after IVIG. These cases highlight the challenging management of refractory parvovirus disease in immunosuppressed patient.

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CARCINOMA CELULAR SECUNDÁRIO A ÚLCERA CRÔNICA NA DOENÇA FALCIFORME

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