

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 $453\text{ mil}/mm^3$, reticulócitos $391700/mm^3$, 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: $18600/$