esquerdo, ocorrido havia 6 meses. Paciente apresentava anemia na admissão, com hemoglobina de 9,7 g/dL evoluindo com queda para 8,1 g/dL após 12 horas. Notava-se prolongamento do TTPa, com relação de 2,81, e TAP dentro da normalidade. Doppler arterial detectou pseudoaneurisma no sítio de punção da artéria radial, com controle do sangramento e estabilização de níveis hematimétricos após arteriorrafia. Apesar disso, o paciente manteve TTPa prolongado não justificado. Em investigação, foram solicitados anticorpos antifosfolípides (anticardiolipina, anti-beta2glicoproteína I e anticoagulante lúpico), com resultados dentro da normalidade, atividade do fator VIII, com valor de 1% e inibidor do FVIII de 10,72 UB/mL, confirmando o diagnóstico de hemofilia A adquirida. Paciente tratado com prednisona 1 mg/Kg/dia, sem necessidade de agente hemostático de bypass devido resolução completa do sangramento. Rastreio para neoplasias sem alterações. Após 6 semanas de terapia imunossupressora, paciente apresentava atividade do FVIII de 68% e inibidor de 0,8 UB/mL, sendo iniciada a retirada gradual do corticoide. Na 17ª semana, paciente sem corticoide, com atividade de FVIII de 144% é inibidor de 0,23 UB/mL. **Discussão:** Em contexto de TTPa isoladamente prolongado, o teste da mistura na abordagem inicial seria relevante. Na ausência de correção do TTPa, deve-se considerar a presença de inibidor de fator, de anticoagulante lúpico ou da atividade de anticoagulantes (heparina e anticoagulantes orais diretos). **Conclusão:** Ressaltamos, neste relato de sangramento grave adquirido em idoso, a importância da análise cuidadosa dos testes laboratoriais de triagem. No caso de TTPa isoladamente prolongado, a hipótese de hemofilia A adquirida deve ser considerada e investigada detalhadamente.

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LABORATORY DIAGNOSIS AND MONITORING OF FVIII AND FIX INHIBITORS IN BRAZIL: RESULTS OF THE EXTERNAL QUALITY ASSESSMENT PROGRAM (PAEQ-HEMOSTASIS)

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Introduction: The major complication of therapy in patients with hemophilia is the development of factor VIII (FVIII) and factor IX (FIX) inhibitors. The control of acute bleeding in patients with inhibitors depends on the titer of it, which also influences the choosing the most appropriate product to be used for treatment. Despite their clinical relevance, the laboratory diagnosis is a challenge. In 2022, data from the Ministry of Health showed that only 10.27% of hemophilia A, and 4.3% of hemophilia B patients were evaluated to inhibitor according to the *International Society on Thrombosis and Haemostasis* (ISTH) recommendations. The aim of this study was to assess the capacity of Brazilian laboratories to identify FVIII and FIX inhibitors. **Materials and methods:** Reference centers for the diagnosis and treatment of bleeding diseases in Brazil were contacted by PAEQ-Hemostasis, an international external quality assessment program, specializing in hemostasis tests. Three points were addressed: (1) Access to diagnosis; (2) Level of technical alignment of the laboratory according to ISTH and International Council for Standardization in Haematology (ICSH) recommendations; (3) Barriers to the correct interpretation of inhibitor data. **Results:** The preliminary result shows the participation of 29/41 (70.7%) of the laboratories. When evaluating geographical distribution, regional participation was 4/7 (57%)-North, 6/10 (60%)-Northeast, 3/4 (75%)-Midwest, 5/5 (100%)-South and 11/15 (73%)-Southeast. The southeast contributed 38% of the data. As for diagnostic access, 1/29 (3.4%) of the centers do not perform inhibitors tests. In 11/28 (39.29%) of the laboratories, the start of the inhibitor investigation is done with the APTT-based inhibitor test (APTT-Inib), of which 45.5% do not continue the inhibitor investigation if the tests shows a negative results. The inhibitor titration test recommended by the ISTH, Bethesda-Modified assay (BM), is available in 25/29 (86.21%) centers. Most laboratories are technically aligned with the ICSH guidelines. However, only 9/25 (36%) of the laboratories use chromogenic methodology, and a limited number of laboratories (< 30%) have the technical knowledge to change the protocol in special cases, such as patients on concomitant use of FVIII concentrate and emicizumab. Only 6/27 (22%) had problems with interruptions in the supply of reagents. In 17/25 (68%) of the cases, the nurse was responsible for reporting the data to the Ministry of Health (web system), and 13/25 (52%) reported updating the web system on a weekly basis. **Discussion and conclusions:** The Ministry of Health registry shows that the APTTInib is the first-line test used to evaluate the inhibitor in 80% of the HA patients tested, and only 10.27% of these patients were evaluated by BM. The data obtained in this study show that the BM is available in almost all Brazilian states, and that 54.5% of laboratories use it as the first-line test in the investigation of inhibitors, rather than the APTTInib. Currently, < 50% of reference laboratories in Brazil are technically prepared to support the new treatment therapies available to patients with hemophilia, and there is limited technical knowledge to identify the necessary changes in special cases. The next steps in this study will be to consolidate the results from the 41 treatment centers, and to send samples of patients pre-diagnosed with inhibitors to assess the quality of the results generated by the participants.

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SÍNDROME DO ANTICORPO ANTIFOSFOLÍPIDE – HIPOPROTROMBINEMIA: RELATO DE CASO

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Objetivo: Descrever o caso de uma paciente jovem com trombose provocada com síndrome do anticorpo antifosfolipide triplo positiva que teve múltiplos episódios de sangramento, sendo diagnosticada hipoprotrombinemia após investigação etiológica. **Materiais e métodos:** Dados foram extraídos do prontuário médico de consultas ambulatoriais. **Relato de caso:** Paciente feminina, de 45 anos, com quadro de Trombose venosa profunda em membro inferior direito em 2000 após 2 meses de uso de anticoncepcional hormonal combinado oral. Fez uso de varfarina por 2 anos. Após isso teve dois partos prematuros: o primeiro com 28 semanas concomitante a quadro de pré-eclâmpsia e o segundo com 32 semanas. Em investigação etiológica, teve diagnóstico de síndrome do anticorpo antifosfolipide por critérios clínicos somados com critérios laboratoriais, positivando anticorpo antifosfolípide, anticorpo anti-B2 microglobulina e anticorpo anti-cardiolipina. Desde então manteve quadro de sangramento uterino anormal, com miomectomia videolaparoscópica em 2015 com choque hemorrágico e conversão para laparotomia. Seguido a esse episódio, foi desencadeada investigação de histórico de sangramento, com paciente tendo 10 pontos em *bleeding assessment test*. Paciente inicialmente com testes de triagens de coagulação alterados com pesquisa de deficiências de fatores e inibidores, com perda de paralelismo dos fatores VII, IX e VIII, e paralelismo de curvas de fator II. Os achados caracterizaram a síndrome anticorpo antifosfolípide hipoprotrombinemia. Desde então paciente é manejada com observação clínica e uso de hidroxicloroquina pela reumatologia com objetivo de redução de risco de evento trombótico. **Discussão:** A entidade anticoagulante lúpico hipoprotrombinemia é uma entidade que predispõe a sangramentos pela presença do anticoagulante lúpico. A deficiência de fator II deve ser suspeitada quando há prolongação simultânea de tempo de protrombina e tempo de tromboplastina ativada. Não há recomendações do tratamento dessa síndrome atualmente. O achado dessa síndrome é incomum, tendo raros registros em periódicos. **Conclusão:** A síndrome antifosfolípide associada com hipoprotrombinemia é uma entidade rara, sem condutas bem definidas, cujo relato de casos similares devem ser descritos para tentativas de condutas comuns. O uso de hidroxicloroquina e a manutenção da paciente sem anticoagulação parecem ser condutas possíveis com certo sucesso clínico.

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MONITORING FIBRINOLYSIS WITH GLOBAL ASSAYS TO DETECT HYPER AND HYPOFIBRINOLYSIS PHENOTYPES

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Introduction: The fibrinolytic system is an important component of hemostasis that acts as a balance of blood coagulation to protect the vasculature from damaging thrombus formation. Monitoring fibrinolysis has been considered an emerging science, and recent studies show that fibrinolysis aids in the assessment of subgroups of critically ill patients at risk of bleeding and thrombotic complications. In this context, global tests to identify fibrinolytic potential have been developed, and knowledge about the performance of these tests is still limited. The aim of this study was to evaluate the performance of global fibrinolysis assays to detect hypo- and hyperfibrinolysis phenotypes. Method: In this study, samples from Healthy Individuals (HI), patients with hematologic Cancer (CA) and Venous Thrombosis (VTE), were obtained from the UNICAMP Hemocentro biobank. The methods assessed were: (1) Turbidimetric tPA-induced clot lysis time assay (tPA-CLT), (2) Turbidimetric tPA-induced clot lysis time assay with thrombomolulin (tPA-CLT-TM), (3) Plasmin generation test (PG). Demographic data, biochemical and coagulation tests, and clinical outcomes were collected. The study was part of the ISTH Reach the World Fellowship Program, in collaboration with the UNC Blood Research Center, Chapel Hill NC USA. **Results:** Plasmas from 107 individuals were evaluated in parallel: 30 CA, 25 VTE, and 52 HI matched by sex and age. The median age was 53 (21–71), 48 (25–72), 43 (19–66), the male to female ratio was 15:15, 17:8, 34:18 for CA, VTE and HI, respectively. For tPA-CLT, only the median Onset-Time (min) was found to be higher for VTE when compared to HI, 6.0 and 5.3 ($p < 0.05$). Interestingly, the tPA-CLT presented 4/6 parameters with higher median for CA compared to HI: (1) Time to peak (min) 10.67 and 9.23 ($p > 0.005$); (2) VMax (mOD/Min) 526.80 and 386.40 ($p < 0.0001$); (3) Turbidity change 0.6851 and 0.5000 ($p < 0.001$); (4) AUC 4.085 and 3.060 ($p < 0.05$). The tPA-CLT-TM test showed the same pattern of results as the tPA-CLT test for VTE and CA. However, for CA the results showed even more significant differences in the same parameters, with the addition of Lysis Time (min) 26.84 and 19.33 ($p < 0.001$). PG presented 2/5 parameters with lower median for VTE compared to HI: (1) Onset-Time (min) 4.465 and 4.760 ($p < 0.05$) and (2) VMax (mOD/Min) 22.25 and 23.26 ($p < 0.05$). To PG, CA presented 3/5 parameters with lower median compared to HI: (1) VMax (mOD/Min) 20.58 and 23.26 ($p < 0.001$); (2) Peak (nM) 54.40 and 60.26 ($p < 0.05$); EPP (nM.min) 337.1 and 479.5 ($p < 0.001$). CA patients showed an association between tPA-CLT parameters and creatinine ($p < 0.001$) and eGRF was assessed. VTE patients showed an association of tPA-CLT parameters with LDL, Triglycerides ($p < 0.05$). Thrombin generation test and D-dimer were evaluated. **Discussions and conclusions:** The performance of global fibrinolysis tests was different between VTE, CA and HI. The tPA-CLT and tPA-CLT-TM assays proved to be sensitive for CA patients, suggesting faster, larger and heavier fibrin formation. In a more specific evaluation, CA showed lower plasmin generation. The tPA-CLT with the addition of thrombomodulin, as