

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. Rastreo 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)

AP Francisco, JM Annichino-Bizzacchi, MRG Andrade, SC Huber, H Aguiari, BM Martinelli, EV Paula, MC Ozelo, SAL Montalvão

Laboratório de Ensaio de Proficiência, Hemocentro, Universidade Estadual de Campinas (UNICAMP), Campinas, SP, Brazil

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

G Cecchetti, LCB Peruffo, ACB Edir, CC Miranda, SHV Freiras, SB Lopes, FB Patricio, MDC Gonçalves, IS Barbosa, GL Macedo

Universidade Federal do Paraná (UFPR), Curitiba, PR, Brazil