recombinante ativado ou complexo protrombínico parcialmente ativado para controle de sangramento e imunossupressão para erradicação do inibidor. Atrasos nesse diagnóstico, como no caso do paciente, podem acarretar em risco de sangramentos que aumentam a morbi-mortalidade desses pacientes. **Conclusão:** Apesar de ser uma doença relativamente infrequente, é extremamente importante o reconhecimento clínico precoce desta entidade para assim iniciar o tratamento oportuno, e evitar complicações graves.

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EFFICACY, EFFECTIVENESS, AND SAFETY OF PLASMA-DERIVED BLOOD COAGULATION FACTOR X CONCENTRATE FOR THE TREATMENT OF HEREDITARY FACTOR X DEFICIENCY: A SYSTEMATIC REVIEW

MM Barbosa [a,b], ACPN Pinto [b,c,d], A Livinalli [b], CF Oliveira [b,e], J Machado-Rugolo [b,f], MS Lima [b,g], MO Almeida [b]

[a] *Universidade Federal de Minas Gerais (UFMG), Belo Horizonte, MG, Brazil*
[b] *Unidade de Avaliação de Tecnologias em Saúde (UATS), Hospital Alemão Oswaldo Cruz, São Paulo, SP, Brazil*
[c] *Universidade Federal do Amapá (UNIFAP), Macapá, AP, Brazil*
[d] *Cochrane Brazil, São Paulo, SP, Brazil*
[e] *Instituto de Saúde (SES), São Paulo, SP, Brazil*
[f] *Hospital das Clínicas, Faculdade de Medicina de Botucatu, Universidade Estadual Paulista (UNESP), Botucatu, SP, Brazil*
[g] *Hospital Universitário Professor Edgard Santos (HUPES), Universidade Federal da Bahia (UFBA), Salvador, BA, Brazil*

Introduction: Hereditary factor X deficiency (FXD) leads to bleeding episodes that vary in intensity depending on the level of factor X in the blood. A novel plasma-derived blood coagulation factor X concentrate (Coagadex®; pdFX) was developed for use in patients with this disease and it is available in Brazil. **Aim:** To analyze the efficacy, effectiveness, and safety of pdFX in patients with hereditary FXD. **Methods:** A systematic search (PROSPERO approval: CRD42022303062) was conducted in accordance with the Cochrane search methods, using the following electronic databases: Embase, MEDLINE via PubMed, Cochrane Central Register of Controlled Trials, and Latin American and Caribbean Health Sciences Literature, as well as clinical trials databases such as ClinicalTrials.gov and the EU Clinical Trials Register, to identify studies that included patients of any age, both sexes, with any type of severity of FXD. Two reviewers independently screened references, selected the studies, extracted the data, and assessed the risk of bias using the ROBINS-I tool. This study followed the Preferred Reporting Items for Systematic Reviews and Meta-analyses (PRISMA) reporting guideline. **Results:** A total of four studies involving 30 patients were included. Overall, the risk of bias was judged as serious. Across studies, pdFX was used to prevent bleeds (Ten01 and Ten02 studies), for perioperative bleeding management (Ten03), or for routine prophylaxis and on-demand treatment (Ten05). All studies included patients with severe or moderate severity. Only two participants with mild severity were included (Ten03). The efficacy of pdFX was rated "excellent"by the majority of investigators. No patients developed thrombotic events or factor X inhibitors, and no deaths or other serious adverse events were reported in any of the studies. **Discussion:** The best available evidence was provided by three single-arm trials and one retrospective study with compassionate use, with an overall serious risk of bias. None of the included studies compared pdFX with the current treatment: prothrombin-complex concentrate or fresh-frozen plasma. No validated tool was used to evaluate the effectiveness of the intervention regarding the bleeding episodes, instead, the investigators rated them subjectively. The included studies were performed in different populations, using pdFX for different purposes. Overall, the results of this systematic review reveals several weaknesses in the currently available evidence. **Conclusion:** This systematic review showed that the currently available data regarding pdFX for the treatment of hereditary FXD is very uncertain, mainly due to the lack of comparator arms and the overall serious risk of bias in the included studies. Therefore, we cannot draw any conclusions as to whether factor X concentrate is associated with a higher incidence of AEs or whether it can impact on the incidence and severity of bleeding events compared to other treatments.

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HEMOFILIA ADQUIRIDA A, UMA CAUSA RARA DE HEMATÚRIA

JO Martins [a], GM Santos [b], LM Corso [a], LL Perruso [a], RN Vita [a], TRF Rocha [b], Y Nukui [a], MG Cliquet [a]

[a] *Hospital IGESP, São Paulo, SP, Brasil*
[b] *Hospital das Clínicas, Faculdade de Medicina, Universidade de São Paulo (HCFMUSP), São Paulo, SP, Brasil*

Objetivos: Relatar caso de hemofilia adquirida A com apresentação de hematúria macroscópica isolada, bem como descrever a abordagem terapêutica utilizada. **Relato de caso:** Paciente masculino de 64 anos admitido com quadro de hematúria macroscópica persistente em pós operatório de ressecção transuretral de próstata, sem melhora com irrigação vesical contínua. Não apresentava outras manifestações hemorrágicas. Previamente hígido, sem antecedentes hemorrágicos, sem história familiar de coagulopatia. Em exames admissionais, apresentava hemograma com anemia (Hb 7,1g/dL Leuco 6.840/mm^3 Plaq 149.000/mm^3), coagulograma com TAP e TT normais e TTPa alargado (82- 97 seg), visto que em exames prévios apresentava TTPa dentro da normalidade (28 seg). Teste da mistura a 50% sem correção do TTPa, indicando a presença de inibidor. Dosagem de anticoagulante lúpico negativo. Dosagem de FVIII por método