

estratégias de tratamento e profilaxia a depender da magnitude das intercorrências. É importante que esses pacientes recebam atenção multidisciplinar e sejam referenciados para serviços que ofertem esta modalidade.

<https://doi.org/10.1016/j.htct.2021.10.388>

PERFIL EPIDEMIOLÓGICO DOS HEMOFÍLICOS EM TRATAMENTO EM UM AMBULATÓRIO DO NORDESTE BRASILEIRO



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Introdução: Hemofilia é uma doença crônica do sistema hemostático resultante de deficiência quanti ou qualitativa de uma ou mais proteínas plasmáticas, e pode se manifestar através de quadros hemorrágicos ou trombóticos. Etiologicamente, podem ser classificadas de acordo com as deficiências de fator em: hemofilia A (Fator VIII) ou hemofilia B (Fator IX). O Programa de Dose Domiciliar (DD), implementado pelo Ministério da Saúde em 1999, tem como objetivo permitir que o paciente com hemofilia possua em sua residência pelo menos uma dose suficiente de pró-coagulante, para uso em caso de hemorragia, até que ele possa buscar atendimento médico em seu centro de tratamento, sem interromper, de imediato, suas atividades diárias. **Objetivo:** Descrever o perfil dos pacientes portadores de hemofilia em um ambulatório do nordeste brasileiro que participam do programa oficial do Ministério de Saúde de DD. **Material e métodos:** Trata-se de um estudo descritivo, transversal, de abordagem quantitativa, com utilização de dados de prontuários dos pacientes cadastrados no programa de DD no estado de Sergipe. A coleta de dados ocorreu de janeiro de 2013 a dezembro de 2019. Foram coletadas informações sobre as seguintes variáveis: idade, sexo, estado civil, município, estado de origem e diagnóstico. Após a seleção de dados válidos, os mesmos foram organizados e analisados em planilhas do programa Excel. **Resultados:** Havia 66 pacientes cadastrados na unidade sendo todos do sexo masculino. A idade dos pacientes variou entre 4 a 60 anos com a média de 26,3 (DP+14,1) e a maioria tinha entre 11 e 30 anos (54,6%). Sobre o local de procedência, a maior parte residia no estado onde a pesquisa foi realizada (92,4%) sendo 36,1% da capital e 63,9% do interior. Em relação ao estado civil, os solteiros corresponderam a 77,3% dos pacientes, seguidos dos casados (15,2%); os outros 7,6% não foi informado no prontuário. No que se refere ao tipo de

hemofilia (A ou B), a hemofilia A foi o diagnóstico mais frequente e representou 92,4% dos pacientes cadastrados. **Discussão:** Hemofilia é uma doença genético-hereditária ligada ao cromossomo X e transmitida quase exclusivamente a indivíduos do sexo masculino, o que justifica o resultado desse estudo. Os dados encontrados em relação à idade dos pacientes se assemelham aos da literatura, tanto no que se referiu à média quanto à variação das idades. Quanto ao município de origem, verificou-se que a maior parte pertencia a municípios distantes da cidade sede, o que poderia dificultar um tratamento em caso de emergência. A DD permite o tratamento precoce do sangramento, reduzindo assim, suas complicações. Em relação ao estado civil, a quantidade de pacientes solteiros pode estar relacionada à média das idades encontradas. No que se refere ao tipo de hemofilia, foi encontrada uma quantidade maior de casos de hemofilia A e menor de B, quando comparado aos dados da Federação Mundial de Hemofilia, com relatos de 80 a 85% de hemofilia A e 10 a 20% hemofilia B. **Conclusão:** Este estudo atingiu seus objetivos, destacando a importância da atenção à saúde e do monitoramento constante dos dados, o que pode gerar subsídios para a elaboração de outros estudos relacionados às coagulopatias, com o intuito de contribuir para a qualidade de vida desses pacientes.

<https://doi.org/10.1016/j.htct.2021.10.389>

PHARMACOKINETIC-GUIDED PROPHYLAXIS FOR PEOPLE WITH HEMOPHILIA A



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Pharmacokinetic (PK)-guided prophylaxis for people with hemophilia A (PwHA) treatment has been related to fewer bleeding and cost savings have been reported. myPKFiT® is a population PK tool available for this purpose, developed for α -octocog (Advate®, Takeda) analysis. We evaluated the impact of myPKFiT®-guided prophylaxis in PwHA on replacement costs and bleeding episodes. Men with severe (FVIII < 1%) or moderate (FVIII 1%-5%) HA without inhibitors and treated with Advate® were invited at two Southern Brazilian Hemophilia Treatment Centers. Inclusion criteria involved ≥ 50 exposure days, age 1-65 years, weight 12-120 kg, bleeding-free for ≥ 2 weeks, and last surgical procedure ≥ 6 months. PwHA who developed inhibitor (> 0.6 BU/mL at two time-points) during the study period were excluded. Anthropometric and hemophilia-related data were obtained using a standardized form. PK analysis was performed according to myPKFiT® recommendations, using one-step test. After PK analysis, PwHA were approached to adjust their regimen according to bleeding phenotype, arthropathy, and physical exercise. Replacement regimen and FVIII disposal (from now called consumption) were assessed before and after treatment adjustment. PwHA younger than 15 years were evaluated for 6 months, while those 15 years or older were evaluated for 12 months. Annualized bleeding rate (ABR) was calculated from the reported treated bleeding episodes. FVIII disposal (from now reported as consumption) was adjusted by weight (kg) and monthly. The cost of 1 IU of Advate® was based on the median date of the evaluated periods, which was July/2019. We converted the price in Real into American Dollar, considering the currency at the date that the Brazilian Ministry of Health purchased the product. A total of 37 PwHA were included. In the younger group (n = 20), 75% were severe, and 65% had no hemophilic arthropathy. Half of these patients were on primary prophylaxis. Two PwHA had a previous history of successful immune tolerance induction. Among the older group (n = 17), 76% were severe, 1 PwHA was treated exclusively on-demand before adjustment, none were on primary prophylaxis, and 12% had no hemophilic arthropathy. One participant developed inhibitor during the study. Median [interquartile range, IQR] ranged from 9.5 [8.0-10.0] h, among younger individuals, to 10.5 [9.0-11.0] h, among the older ones. Three PwHA from the older group were excluded from the cost analyses: 1 developed inhibitor during follow-up, 1 was treated exclusively on-demand before PK-analysis, and 1 was prescribed plasma-derived FVIII few months after adjustment. From the remaining 34 PwHA, all individuals were advised about behavior and time of infusion, according to their bleeding phenotype, arthropathies, and physical exercise. Among younger PwHA, monthly total FVIII replacement cost increased after PK-based adjustment (p < 0.0001), while reducing costs of episodic therapy (p = 0.05) and increasing costs of prophylaxis (p < 0.0001). Median [IQR] ABR reduced from 3.0 [0.5-10.0] to 1.0 [0.0-2.0]. Among older PwHA, although monthly total and prophylactic costs did not change between the periods, costs of episodic treatment reduced (p = 0.039). ABR did not change either. PK-adjusted prophylaxis was related to increased treatment costs among PwHA < 15 years, but not among individuals ≥ 15 years. Although ABR reduced after adjustment among younger PwHA, no ABR change was evidenced among older participants.

<https://doi.org/10.1016/j.htct.2021.10.390>

PREVALENCE OF HEMATURIA IN PATIENTS WITH HEMOPHILIA A AND B AND ITS ASSOCIATION WITH COMORBIDITIES: EXPERIENCE FROM A BRAZILIAN CENTER

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Background: Hemophilia A and B are X-linked inherited bleeding disorders. Hematuria is a common manifestation among patients, resulting in a worsening of quality of life. Results of previous studies regarding the association between hematuria and hemophilia have been inconsistent, leading to the need for further analysis. **Aims:** Determine the prevalence of macroscopic or microscopic hematuria in patients with hemophilia, as well as its association with comorbidities. **Methods:** Transversal unicenter study with adult patients (≥ 18 years old) bearing hemophilia A or B under active outpatient clinic in a reference center over the year of 2019. Hematuria was defined as any macroscopic episode or ≥ 3 red blood cells per high power field in urine analysis. Using simple and multivariable logistic analysis, age, diagnosis type, disease severity, comorbidities and creatinine clearance have been studied as potential characteristics associated with the prevalence of hematuria. **Results:** In total, 179 male patients were included. Mean age was 26 years (sd = 15), the majority of them were diagnosed with hemophilia A (76%) and 66% presented a severe disease. Despite our young population, the prevalence of comorbidities was high, mainly hypertension (10%), dyslipidemia (10%), diabetes (5%) and obesity (10%). The prevalence of hematuria was 39% (95% CI 32% - 47%), of these a half presented at least one macroscopic episode. Age (OR = 1.03[1.01-1.05]), hemophilia B (OR = 0.47[0.21-0.99]), hypertension (OR = 3.88[1.45-11.55]), obesity (OR = 2.92[1.11-8.24]) and creatinine clearance (OR = 0.99[0.98-1.00]) were associated with hematuria in univariable analysis. No independent factors were found in multivariable analysis. **Conclusions:** Hematuria is common among patients with hemophilia and further studies are needed to elucidate the relationship between hematuria and increased morbidity.

<https://doi.org/10.1016/j.htct.2021.10.391>

PREVALÊNCIA DE AFECÇÕES HEMORRÁGICAS E OUTRAS DOENÇAS DO SANGUE E DOS ÓRGÃOS HEMATOPOIÉTICOS

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Objetivos: Descrever as internações hospitalares por afecções hemorrágicas e outras doenças do sangue e dos órgãos hematopoéticos no estado da Bahia, através da lista de morbidade do CID-10 (D65-D77) no período de 2010 a 2020, quanto aos

