

dos exames e sem ou pouco embasamento clínico). Isso demonstra uma necessidade de aprimoramento referencial e necessita de uma análise mais aprofundada para revelar a real motivação situacional na elaboração, seja ela uma sobrecarga na demanda, dificuldades no manejo por falta de recursos, insegurança/limitação de conhecimentos. Desse modo, se houver a resolutividade e aprimoramento do sistema referencial, as diretrizes do SUS serão contempladas de maneira mais integrativa, equitativa e com menos sobrecargas no sistema e, assim, a demanda terá uma necessidade real de seguimento na especialidade, não sendo ocupadas as vagas por casos de baixa complexidade.

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POLYCYTHEMIA VERA: AN EPIDEMIOLOGIC ANALYSIS AND PATIENT FOLLOW UP



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Objectives: The objectives were to identify the demographic characteristics and the method of treatment of the patients diagnosed with Polycythemia Vera (PV, CID-10 D45.0) in the service, and to help the institution on the creation of its own protocol for diagnosis and treatment of such patients.

Material and Methods: This retrospective and observational study was made with an analysis of data from 38 patients diagnosed with PV. The data range from 1998 to 2019 and was collected using a protocol established by the researchers.

Results: The sample features 60,5% of male and medium age of 57 years, 17 of those were classified as high risk patients. Observing the cardiovascular risk factors, the most prevalent was Arterial Hypertension (55,3%). The most related clinical finding was plethora (68,4%) followed by splenomegaly (42,1%). Five patients had thromboembolic events before the treatment and four had after it. As a treatment, almost all patients used hydroxyurea, with some using phlebotomy as prophylactic and/or maintenance option. Three of the patients evolved to myelofibrosis. Only six patients died during years studied and the survival rate was of 19,8 years after diagnosis. **Discussion:** Polycythemia Rubra Vera is the most common myeloproliferative disease, occurring in 1,9/100.000 inhabitants (USA). The disease has a discrete predominance over men, grows in incidence after the 6th decade and occurs earlier in women. The disease manifests with symptoms occurring from blood hyperviscosity or vasomotor factors, such as fatigue, headache, visual disturbance and pruritus. Splenomegaly is the most common sign. Clinical findings can persist with laboratory control of the disease and can aggravate with its evolution. As such, clinical parameters are unreliable factors for patient's control. Given the unspecific clinical findings, the diagnosis is established following a set of criteria anchored in laboratory findings and guided by WHO's guideline of 2016. Risk stratification is fundamental to establish the correct treatment, in an attempt of thrombosis prevention. The patient is considered as "high risk" if its age is over 60 years or it has

a history of previous thrombosis. Modern protocols for "low risk" patients recommend phlebotomy until hematocrit target level is reached and daily low dose aspirin. For "high risk" patients the recommendation is hydroxyurea and to evaluate the response, preceded by phlebotomy until target hematocrit levels. Antiplatelets agents, anticoagulants, phlebotomy and other drugs can be used in special cases. The disease's natural history is not well defined, mainly due to the established treatments that change its progression. Yet, life expectancy is lower compared to the overall population. **Conclusion:** The patient's array was epidemiologically consistent with the literature. However, symptomatology data was shown to be lower than reported in other studies. Phlebotomy was used in agreement with literature (when the hematocrit was over 45%). However, the usage of hydroxyurea in lower-risk patients disagrees with international treatment protocols. This may have been due to the patients' socioeconomic conditions which may make it difficult for the patient to receive phlebotomy only treatment. Despite this divergency in treatment, the study showed a lower thrombosis rate than what has been reported in the literature.

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QUALIDADE DE VIDA RELACIONADA À SAÚDE EM CRIANÇAS E ADOLESCENTES COM DOENÇA FALCIFORME



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Objetivo: Analisar a qualidade de vida relacionada à saúde de crianças e adolescentes com doença falciforme (DF). **Materiais e métodos:** Estudo transversal realizado com 97 pacientes na faixa etária entre cinco e 18 anos com diagnóstico clínico e laboratorial de DF, de ambos os sexos, atendidos no Hospital de Hematologia e Hemoterapia de Pernambuco. Foram aplicados os questionários Pediatric Quality of Life Inventory, versão 4.0 – relato da criança/adolescente. **Resultados:** A maioria dos pacientes era do sexo feminino (76,3%), apresentava anemia falciforme (89,7%) e de etnia parda (59,8%). Houve associação significativa na dimensão psicossocial entre a faixa etária 5 a 7 (72,10) e de 8 a 12 anos ($p \leq 0,05$), esta última com um pior score de qualidade de vida (65,36). Em relação ao sexo, as únicas diferenças significativas ocorreram na faixa etária de 13 a 18 anos na variável de dimensão física e na média das dimensões, sendo as médias nos dois casos mais elevadas no sexo feminino do que no masculino (70,89 x 56,88 na dimensão física e 71,72 x 59,77 na média das dimensões). **Discussão:** Nesse estudo houve diferença significativa na variável dimensão psicossocial entre as faixas etárias de 5 a 7 anos e 8 a 12 anos, sendo esta a que apresentou a média menos elevada (65,36). Em contrapartida, um estudo identificou que não houve associação significativa entre as faixas etárias em nenhuma dimensão (Menezes et al., 2013). Porém, todas as