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Demographics and clinical data of hemophilia patients presented in a tertiary care hospital of Pakistan

Arifa Khalid, Syed Nasir Abbas Bukhari, Safia Khan, Saadia Anwar, Mahwish Faizan, Usman Wasti The Children's Hospital & The Institute of Child Health, Pakistan

Background: Hemophilia is a rare X-linked recessive congenital bleeding disorder and has been classified as Hemophilia A (deficiency of Factor VIII), and Hemophilia B (Deficiency of factor IX). The overall incidence is 1 per 10,000 male live births with 80% cases of Hemophilia A. The characteristic phenotype is the bleeding tendency and most of the patients present with easy bruising, prolonged post traumatic bleeds and hemarthrosis. The severity of bleeding in Hemophilia correlates with the level of factor deficiency. Late diagnosis and improper management can lead to lifelong complication of chronic arthropathy. Therefore timely and proper factor replacement therapy can prevent complications and improve quality of life.

Objective: The study was conducted to analyze the clinical data of disease in a developing country with limited resources so that we can predict how we can prevent morbidity and improve the quality of life in our set up.

Methods: The population of the research was patients of Hemophilia, who were reported, in last one year, in Children's Hospital & Institute of Child Health, Lahore. A sample of 55 patients of Hemophilia A and B were reviewed retrospectively. The data was collected irrespective of any discrimination based on demographic factors and following factors were recorded: Clinical features, age at first presentation, age at diagnosis, severity of disease, frequency of admission, factor replacement, and life threatening bleed. Blood complete picture, bleeding time, PT/APTT, mixing studies and factor assays were noted.

Results: Among these 55 patients all were male. Around 93% of the patients were diagnosed as Hemophilia A and approximately 7% as Hemophilia B. Most of the patients diagnosed after 1 year of age (96.4%) although approximately 62% were symptomatic before 1 year of age. About 49% of them were having moderate disease, mild disease was noted in 18%, and rest were with severe disease. Family history was positive in 51% and in all the patients who presented before 6 months of age had severe hemophilia. APTT was prolonged in all the patients with the mean value of >1 minute. Severity of disease and family history has significant relation at p<0.005, while relation between severity of disease and the age at diagnosis is significant but compromised at p=0.05, i.e., critical line. In this research, post circumcision bleed (45.5%) was observed prominently as primary symptom followed by bruises and epistaxis, respectively. However, life threatening bleed was reported in 9% and about 66% developed hemarthrosis. Only 2 patients had APTT done before circumcision. Most of the patients were on demand factor replacement therapy i.e.74.6% and only 5.4% of them were on primary prophylaxis.

Conclusion: On the basis of early presentation and family history, we can predict the severity of the disease. Since family history has strong correlation with severity of disease, therefore performing PT/APTT before circumcision will be helpful and cost effective as screening and early diagnosis of Hemophilia. Early diagnosis, adequate and timely prophylaxis can prevent complications like hemarthrosis leading to chronic arthropathy and severe life threatening bleeds. It can help to improve the quality of life and decrease the morbidity and mortality in a developing country.

arifakhalid_15@yahoo.com