1/ Management of congenital in children at the Sétif pediatric center

M. Messasset, B;Bioud

Introduction:

Hemophilia is the most common and best-reported rare hemorrhagic disorder in the world. In children, the management is multidisciplinary, based mainly on prophylaxis and therapeutic and therapeutic education. **The aim** of this study is to describe epidemiological, clinical, therapeutic, evolutionary and economic aspects of the series of the series and to propose a national management model.

Material and methods:

Our study was ambispective, longitudinal and descriptive study a series of 84 children with hemophilia followed in the pediatric department of Sétif during the period from 1 January 2016 to December 31, 2021.

Results:

Hemophilia A was the most common frequent (78.57%) followed by hemophilia B (21.42%) with a ratio of 4.7:1. The severe form accounted for 76%. Consanguinity is found in (28.6%) The hemophilia prevalence rate in Sétif between 2016-2021 is 11.25/100,000 boys under 18 years of age rate is 23.98/100,000 male births. The average years, the birth prevalence at diagnosis was 21 months. The circumstances of discovery were bleeding (70.2%) during circumcision (35%). Personalized prophylaxis was adopted in 60% of cases, and the median prophylaxis median annualized bleeding rate was 2.01/patient /year, with a hematoma rate of 0.76 haematomas /illness/year and 0.55 mucosal bleeds/illness/year. bleeding/ patient /year, the median annualized annualized joint bleeding rate was 0.70 hemarthroses/patient/year (0.64 for HA and 0.06 for HB), no patient developed a target joint A mean HJSH (Hemophilia Joint Health Score) score of (Hemophilia Joint Health Score) of 12.4,a mean pain-doctor visual analog scale (VAS) of 3.6,the mean FISH (Functional Independence Score in Hemophilia) score of 22 and a mean Petersson radiology score of 1.1. osteoarticular complications are target joints target joints in (4.76%). patient had hepatitis C and (8.33%) of cases developed strong inhibitors in 100% of cases. (57%) of inhibitor-positive cases showed microinversion of intron 22 in half of them, and a missense mutation in 25%. Has average initial quality of life was moderately impaired at (54.87%) and final at (35%) as assessed by the HEMO QUAL questionnaire.

Conclusion: Our work has the expected progress in the management of hemophilia in children, from the role of the multidisciplinary team, the personalization of prophylaxis and its prophylaxis and its significant economic impact, from home treatment to therapeutic to therapeutic education and the management of complications. However, under-diagnosis of the disease radiological assessment and the limited use of physiotherapy.

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