Hemophilia-A Review

Brinda Khanna[1*], Archika[2], SumitBhateja[3], Geetika Arora[4]

[1,2]Dental Surgeon, [3]HOD, [4]Reader, [3]Dept. of Oral Medicine & Radiology, [4]Dept. of Public Health Dentistry, [1-3]Manav Rachna Dental College, Faridabad, Haryana, [4]Inderprastha Dental College & Hospital, Ghaziabad, Uttar Pradesh, India

***Corresponding Author: Brinda Khanna**
Email: brindakhanna.khanna09@gmail.com

Abstract
Abstract Hemophilia is a disease which is prevalent around the globe. The people across the world are more familiar with this when compared to other bleeding disorders owing to the fact that this is run in the family of Queen Elizabeth II. In this disorder, the females are the carriers of the disease while the males are the ones who are affected by the disease.

Keywords: Hemophilia A genetic, Management.

Introduction
Hemophilia A is a chromosomal bleeding disorder. It is an X linked recessive disorder.[1]

It is caused due to deficiency of plasma clotting factor viii. This deficiency can be inherited or can also arise from DNA mutation.[2]

Symptoms
There are three types which include, mild, moderate or severe. The severe type of disorder are usually detected in first few months after birth itself. The moderate and milder forms of the diseases are however usually detected in the later years or in adolscence where there is prolonged bleeding seen after minimal trauma. This disorder is seen in 1 out of 5000 males. The females are the carrier and the disease is manifested in the male off springs of the carriers.[3]

The most common symptoms include bleeding and brising after minimal trauma. Bleeding in such patients can occur internally or externally. The degree of severity of bleeding depends upon the stage of disease and the amount of factor viii deficiency. Bleeding can also occur in the form of nose bleeding, bleeding in joints that is haemarthrosis, blood in urine, stool etcetera.[4]

Types of Hemophilia
Hemophilia occurs due to two major causes:

1. When there is presence of non functioning factor viii and
2. When there is complete absence of factor viii in the blood[9]

"Mild - when the level of factor viii in blood is - >5 - <40%

Moderate - when the level of factor viii in blood is - 1-5%

Severe - when the level of factor viii in blood is < 1%".[5]

Diagnosis
Diagnosis is made with the conjugation of a detailed family history and blood tests. Family and Medical History of patients aids in tracing the origins of the disease and also confirms if similar disease is present in the siblings of the patient. Usually females are not affected by the disease but sometimes they also manifest symptoms. This usually occcurs in females who also have Turner's Syndrome etcetera. The blood tests undertaken to detect its presence include, complete blood cell count, activated partial thromboplastin time, partial thromboplastin time, thrombin time, prothrombin time. The blood cell picture of individuals who suffer from this is usually analogous and in most of the cases depicts a prolonged activated partial thromboplastin time or partial

thromboplastin time with a normal bleeding time and normal prothrombin time.[6]

Hemophilia A and liver diseases

Since Hemohphilia A patients recieve blood transfusions as a measure to control the disease and minimise its manifestations, often due to ingestion of contaminated blood they are at a risk of developing fatal viral infections like Hepatitis C and HIV infections. If an individual gets infected by Hepatitis C virus the chronic disease of liver acclearates leading to cirrhosis of liver which then leads to liver failure and ultimately leads to onset of Hepatocellular Carcinoma.[7]

In a detailed research where clinical trials were done to screen the "End-stage liver disease in persons with hemophilia and transfusion-associated infections" it was oberserved that more than 50% patients were coinfected with HIV along with Hepatitis C. Also, more than 50% of patients presented with bleeding esophageal varices, hepatic encephalopathy, persistent ascites, or death.[8]

Source of Funding

None.

Conflict of Interest

None.

References

1. https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3502605/
2. https://emedicine.medscape.com/article/779322-overview.
3. https://www.ncbi.nlm.nih.gov/books/NBK470265/
4. https://www.medicalnewstoday.com/articles/154880.php
5. https://www.ihtc.org/severity-of-hemophilia/.
6. https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4430880/
7. https://www.ncbi.nlm.nih.gov/m/pubmed/16427378/
8. https://ashpublications.org/blood/article/100/5/1584/106303/End-stage-liver-disease-in-persons-with-hemophilia
9. https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4304051/

How to cite this article: Khanna B, Archika, Bhateja S, Arora G. Hemophilia-A Review. *J Urol, Nephrol Hepatol Sci* 2019;2(3):41-2.