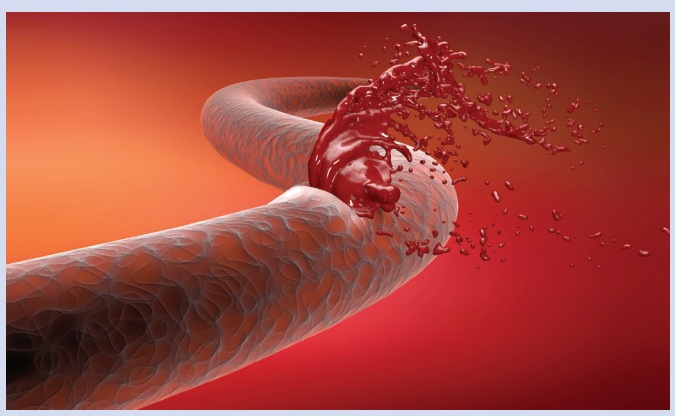
hope

Healing Opportunity
with Patient Education

HEMOPHILIA – AN OVERVIEW

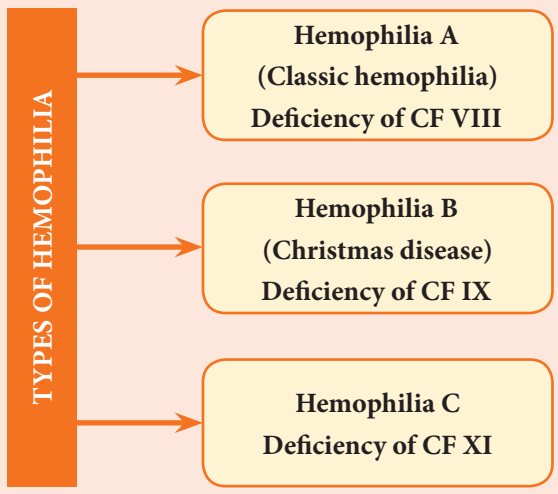
Hemophilia is an X-linked hereditary disorder of coagulation process.[1] This is due to deficiency of clotting factor(s), based on which it is categorized as; hemophilia A (deficiency of factor VIII) or hemophilia B (deficiency of factor IX). Hemophilia manifests as spontaneous bleeding, prolonged oozing after injuries or tooth extractions, excessive blood-loss following surgery, and recurrent bleeding prior to complete wound healing.[1,2] It has been suggested that the bleeding tendency is less severe in individuals with factor IX deficiency and may even be associated with better long-term outcomes.[3]



Sources: 1. Loomans JI, Lock J, Peters M, Leebeek FW, Cnossen MH, Fijnvandraat K. Haemophilia. *Ned Tijdschr Geneeskd.* 2014;158:A7357. **2.** Konkle BA, Josephson NC, Nakaya Fletcher S. Hemophilia A. 2000 Sep 21 [Updated 2014 Jun 5]. In: Pagon RA, Adam MP, Ardinger HH, et al., editors. GeneReviews® [Internet]. Seattle (WA): University of Washington, Seattle; 1993-2016. Available at: https://www.ncbi.nlm.nih.gov/books/NBK1404/. Accessed on: 12.10.16. **3.** Santagostino E, Fasulo MR. Hemophilia A and hemophilia B: different types of diseases? *Semin Thromb Hemost.* 2013 Oct;39(7): 697-701.

IN PATIENTS WITH SEVERE DISEASE, PROPHYLACTIC INFUSIONS OF FACTOR VIII CONCENTRATE THREE TIMES A WEEK OR EVERY OTHER DAY IS SUGGESTED TO MAINTAIN FACTOR VIII CLOTTING ACTIVITY MORE THAN 1% , WHICH NEARLY ELIMINATES SPONTANEOUS BLEEDING AND MAY PREVENT CHRONIC JOINT DISEASE

Types of hemophilia



Abbreviation: CF: Clotting factor.

Sources: 1. Guidelines for the management of hemophilia. Available at: https://www1.wfh.org/publication/files/pdf-1472.pdf. Accessed on: 4.11.2016. **2.** Hemophilia. Available at: https://www.ncbi.nlm.nih.gov/pubmedhealth/PMH0063001/. Accessed on: 4.11.2016. **3.** Hemophilia A. Available at: https://www.ncbi.nlm.nih.gov/pubmed/20301578. Accessed on: 4.11.2016.

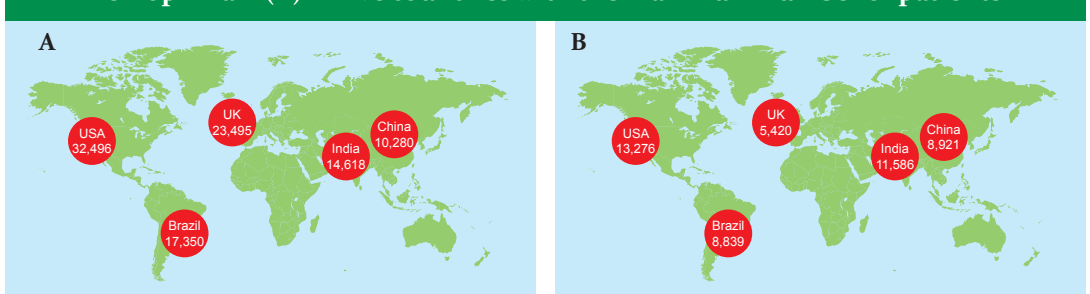
Epidemiological burden and incidence of hemophilia

Estimates indicate that 80% individuals with severe hemophilia belong to developing countries like India, with an incidence

of 1 per 10,000 births. This can be attributed to poor health resources and less than adequate treatment for hemophilic patients in these countries.[1-3]

HEMOPHILIA A IS COMMONER THAN HEMOPHILIA B, ACCOUNTING FOR 80-85% OF THE TOTAL HEMOPHILIA POPULATION

Global distribution of total reported cases with bleeding disorders (A) and hemophilia A (B) in five countries with the maximum number of patients



Source: Kar A, Phadnis S, Dharmarajan S, Nakade J. Epidemiology & social costs of haemophilia in India. *Indian J Med Res.* 2014;140(1):19-31.

Sources: 1. Kar A, Phadnis S, Dharmarajan S, Nakade J. Epidemiology & social costs of haemophilia in India. *The Indian Journal of Medical Research.* 2014;140(1):19-31. **2.** De Kleijn P, Odent T, Berntorp E, Hilliard P, Pasta G, Srivastava A, Iliescu A, Mohanty S. Differences between developed and developing countries in paediatric care in haemophilia. *Haemophilia.* 2012 Jul;18 Suppl 4:94-100. **3.** Coppola A, Di Capua M, Di Minno MND, et al. Treatment of hemophilia: a review of current advances and ongoing issues. *Journal of blood medicine.* 2010;1:183-195.

Emotional costs of hemophilia

Hemophilia has a strong impact on parents and families of the affected patients because of the chronic and progressive disability of the disease, high treatment costs, emotional turmoil, social issues, and the premature mortality.

It has been speculated that the inability to achieve educational goals and difficulty with employment suffered by many patients contribute to a feeling of low self-esteem, anxiety, and guilt. Approximately, one-third of individuals with a serious medical condition have symptoms of depression.

A constant underlying presence of such feelings can hinder one from leading a normal life.[1,2]

Sources: 1. Kar A, Phadnis S, Dharmarajan S, Nakade J. Epidemiology & social costs of haemophilia in India. *Indian J Med Res.*2014;140(1):19-31. **2.** duTreil S. Physical and psychosocial challenges in adult hemophilia patients with inhibitors. *J Blood Med.* 2014;5:115-122.

Hemophilia: A social stigma

In a country like India, having a son born with an incurable disease is a stigma the mother has to live with.

Apart from the guilt feelings associated with the birth of a child with hemophilia, the mother suffers a lot at the hands of relatives and society. Bringing up the child with hemophilia takes a heavy toll on parents physical and mental health. The emotional strains associated with a possibly affected newborn and his care throughout his life may have a grievous impact on couple dynamics and marital relationship.[1,2]

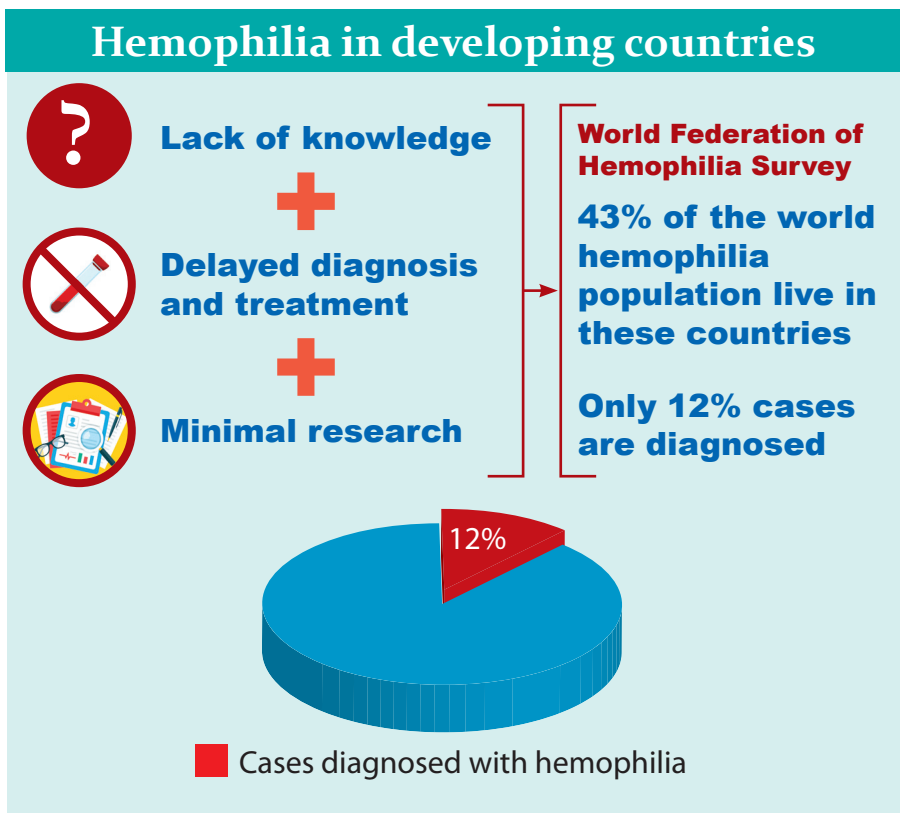
Sources: 1. Coppola A, Cerbone AM, Mancuso G, Mansueto MF, Mazzini C, Zanon E. Confronting the psychological burden of haemophilia. *Haemophilia.* 2011 Jan;17(1):21-7. **2.** Psychosocial dimension of hemophilia. Available at: http://hemophiliabangalore.org/Pdf%20%20File(H)/Psychosocial%20dimension%20of%20hemophilia.pdf. Accessed on: 25.10.2016.

Burning issue in hemophilia treatment

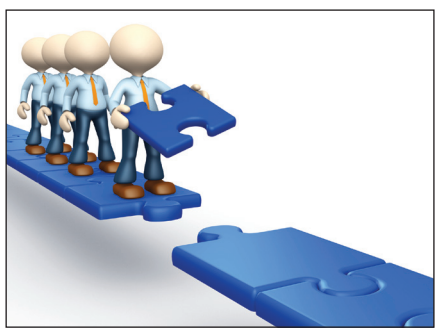
In developed countries, use of clotting factor concentrate has resulted in increased longevity of patients with hemophilia. However in India, lack of access to treatment remains a major factor that compromises the quality of life of patients. This is because of the:

- Lack of knowledge about the genetic and clinical implications of the disorder
- Delayed diagnosis and treatment
- Lack of safe and effective therapies and
- Minimal research activity to improve standards of care.[1]

As per the World Federation of Hemophilia (WFH) Survey, 43% of the world hemophilia population live in countries like India, Bangladesh, Indonesia, and China; sadly out of which, only 12% are diagnosed. However, these countries merely use 2% of the Worlds total factor usage.[2]



The extremely low per capita use of treatment product despite having the second largest number of global patients with hemophilia A illustrates the large treatment gap that exists in the country.



To address this problem, Hemophilia Federation India has taken initiatives to provide diagnostic facilities, supporting the cost of diagnosis, networking private laboratory facilities, and setting up coagulation laboratories.[1,2]

Sources: 1. Shapiro AD, Soucie JM, Peyvandi F, Aschman DJ, DiMichele DM; UDC Rare Bleeding and Clotting Disorders Working Group; European Network Rare Bleeding Disorders Database. Knowledge and therapeutic gaps: a public health problem in the rare coagulation disorders population. *Am J Prev Med.* 2011 Dec;41(6 Suppl 4):S324-31. **2.** Shetty S. Haemophilia - diagnosis and management challenges. *Molecular Cytogenetics.* 2014;7(Suppl 1):I44.

Expert Opinion - Hemophilia

Q: IS THE DIAGNOSIS OF HEMOPHILIA CHALLENGING?

A: Hemophilia is an X-linked congenital bleeding disorder. It is caused by mutations of the clotting factor genes, leading to the deficiency of clotting factors. Deficiency of factor VIII (FVIII) is seen in hemophilia A and deficiency of factor IX (FIX) causes hemophilia B. Since diagnosis of hemophilia is challenging due to lack of knowledge, therefore, most of the cases remain undiagnosed until adulthood. Platelet count, bleeding time, prothrombin time, and activated partial thromboplastin time may be used to screen a patient suspected of having a bleeding disorder. Tests of platelet function such as platelet aggregometry are preferred. In addition factor assays can also be of help in diagnosing and monitoring a case of hemophilia.

Q: WHAT IS THE MANAGEMENT OF HEMOPHILIA?

A: A general rule of emergency management of hemophilia is 'factor first', that is, when in doubt, factor replacement is done immediately before any further investigations. Whenever possible, the patient is treated with the specific factor concentrate. Because of the safety and quality issues of fresh frozen plasma and cryoprecipitate, their use in the treatment of congenital bleeding disorders is not recommended and is justified only when clotting factor concentrates are unavailable. Pharmacological agents such as desmopressin, tranexamic acid, and epsilon aminocaproic acid can also be used in mild to moderate cases of hemophilia.

Q: WHAT ARE THE PRECAUTIONS WHILE INJECTING OR VACCINATING A PATIENT OF HEMOPHILIA?

A: While injecting a patient of hemophilia, subcutaneous route is preferred over intramuscular route, unless covered by infusion of clotting factor concentrates, because of the increased chances of painful intramuscular bleeds via intramuscular route.

Sources: 1. Guidelines for the management of hemophilia. Available at: https://www1.wfh.org/publication/files/pdf-1472.pdf. Accessed on: 4.11.2016. **2.** Price VE, Hawes SA, Chan AK. A practical approach to hemophilia care in children. *Paediatr Child Health.* 2007;12(5):381-383. **3.** Bauer KA. Current challenges in the management of hemophilia. *Am J Manag Care.* 2015 Mar;21(6 Suppl):S112-22.

Psychosocial dimension of hemophilia

Psychosocial dimension of hemophilia is of utmost importance as psychological factors play a crucial role in managing a patient of hemophilia. It acts as a major psychological trauma not only for the individual who is diagnosed with hemophila, but for the entire family as well.

The road to acceptance of hemophilia is laid by turmoil of emotions such as guilt, depression, anger, and low self-esteem, thereby, emphasizing the importance of a caring and supportive family to rise above hemophilia and lead normal life.

Feelings experienced by an individual with hemophilia:

- Feeling of low self-esteem
- Feeling of guilt
- Displaced anger
- Fear and anxiety

FEELING OF LOW SELF-ESTEEM

Due to the fear imposed by this disorder, the individual with hemophilia feels isolated as he cannot engage actively in all the physical activities. Therefore, he suffers from low self-esteem and thinks himself to be a burden on the family. He feels like committing suicide because of the financial burden of the disease the family is facing.

FEELING OF GUILT

A person with hemophilia considers himself responsible for the burden of all family problems as he believes his problem to be the root cause for everything. This ensues a feeling of guilt and leads to self-rejection. Therefore, a supporting environment that acknowledges and nourishes the innate gifts of affected individuals is required to help them to rise above the disease.

THE ROAD TO ACCEPTANCE OF HEMOPHILIA IS LAID BY TURMOIL OF EMOTIONS SUCH AS GUILT, DEPRESSION, ANGER, AND LOW SELF-ESTEEM, THEREBY, EMPHASIZING THE IMPORTANCE OF A CARING AND SUPPORTIVE FAMILY TO RISE ABOVE HEMOPHILIA AND LEAD NORMAL LIFE

DISPLACED ANGER

The frustration, discomfort, or difficulty in making adjustments, in some cases, may lead to displaced anger of the individual on his mother, as he blames his mother for putting him in such a condition. Such pathological display of anger further aggravates the emotional stress of the mother who is already disturbed.

FEAR AND ANXIETY

On account of the unpredictable nature of the disease and the severe consequences, the individual constantly experiences fear or anxiety. The underlying presence of such feelings can hinder one from leading a normal life. As the person grows older, fear of rejection by others in the personal and professional life arises.

FEAR OF DISAPPROVAL, WORRIES OF BEING LEFT BEHIND BY A FAMILY, GUILT, AND ANGER ARE THE PSYCHOSOCIAL ISSUES THAT NEED TO BE DEALT WITH, WHICH REQUIRE A SUPPORTIVE AND CARING FAMILY

Source: Psychosocial dimension of hemophilia. Available at: http://hemophiliabangalore.org/Pdf%20%20File(H)/Psychosocial%20dimension%20of%20hemophilia.pdf. Accessed on: 4.11.2016.

Hemophilia Federation India and its current projects

Hemophilia Federation India (HFI) is a self-help non-governmental organization (NGO) which was established in 1983. Individuals with hemophilia successfully run this organization with help from medical fraternity. It is the only organization in India that conducts structured training for the medical and paramedical practitioners. Through this initiative, India is now represented as National Member Organization at the World Federation of Hemophilia (WFH) based in Canada. It also works collaboratively with World Health Organization (WHO) and National Aids Control Organization (NACO). Some of its ongoing projects include:

Heal A Soul: A BHEL project

Bharat Heavy Electricals Limited (BHEL) has extended its continuous support to HFI for past 3 years. Yet again in partnership with HFI, BHEL is currently assisting a project-Heal a Soul, which includes support of 120 women for Carrier Detection and Pre-natal Diagnosis.

The Hans Foundation projects

At present, the Hans Foundation is supporting 2 main projects in partnership with HFI. These include: 1) Treatment Project (The Healing Touch), which benefits 40 individuals with hemophilia (P/CwH) of factor VIII, 8 P/CwH of factor IX and 2 P/CwH of Feiba deficiency, being supported with 5000 IUs, and 2) Education Project (Educate a Child with Hemophilia) supporting 300 children with hemophilia (CwH) with Rs. 6000/- per year per child.

Give India project

Donations received via this project aid in saving lives by providing free/subsidized treatment to P/CwH in emergency.

Source: Hemophilia Federation (India)- Ongoing projects. Available at: http://www.hemophilia.in/index.php/projects/ongoing-projects. Accessed on: 12.10.16.

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This information is brought to you by Marketers and Makers of Antihemophilic Factors (Factocel VIII and Factocel IX)

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