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Editorial

Editorial on Blood Disorders

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There are many blood component disorders in human beings like Sickle Cell Disease, Thalassemia and Hemophilia which are prominently observed in India amongst all the bleeding disorders. Sickle Cell Disease (SCD) and Thalassemia got lot of attention in recent past from Govt. and Non-Govt. agencies for its diagnosis, management and rehabilitation etc. But the important third bleeding disorder i.e. Hemophilia is not getting proper attention from various Govt. and Non-Government agencies even our medical fraternity (doctors of various specialities) is not fully aware about the implications and sufferings of Hemophilic patients and hence we need to give more importance to the diagnosis, treatment, hereditary aspects and long term management of these patients with making Anti-hemophilia factors available in plenty at cheaper rates throughout our country. Some NGOs like Hemophilia Federation of India (H.F.I.) is doing a good job in mitigating all these aspects of Hemophilics to some extent, but their efforts are not sufficient to tackle the good number of Hemophilics, spread through nook and corners of our vast country with lot of illiteracy, poverty and lack of infrastructural facilities. We need further direct help from our Central Govt. and various State Governments to tackle the day-to-day problems faced by hemophilia patients.

Following is the brief description about hemophilia

Hemophilia is a disease of genetic origin, a sex-linked disorder occurring predominantly in males. The genetical aspect is such that, being carried through the X-chromosome, females are carrier and males are sufferers. This is better understood with the help of the diagram given later. The blood that flows in our body which is normally a liquid has the capacity to solidify (clot) under certain situations. Normally the flowing and clotting capacity are in equilibrium. Hemophilia is a condition where the bleeding from a site fails to stop due to lack of certain clotting factors. These factors are labelled as I, II … etc Hemophilia commonly is due to deficiency of factors VIII and IX. Thus, for a hemophiliac even minor injuries can be serious.

Mode of inheritance

Hemophilia is he classical example of a sex-linked recessive disorder. The genes which control E-VIII and E-IX production are both located on the X-chromosome. As males have only one X-chromosome, the Synthesis of F-VIII of FIX will be deficient if the relevant gene is defective. A hemophilic passes his abnormal X-gene on to all of his daughters and his Y chromosome which is normal, to his sons. Thus, all of his sons will be normal and cannot pass on the defective gene, and all his daughters will be carriers of defective x-gene. A woman who is a carrier has four possible outcomes with each pregnancy a normal girl, a normal boy, a girl who is carrier or a boy with hemophilia. Thus, the risk of having a baby with hemophilia is 1 in 4 for each pregnancy. There are rare instances of females with hemophilia. They are descendants of a hemophilia father and a carrier mother.

Awareness amongst the general public and doctors too is lacking. Mothers fail to give importance to the blue-black marks which occur on the child's body. Sometimes doctors tend to ignore them when parents bring it to their notice. Some children in rural areas

die before they reach teens, sometimes for as small a reason a non-stop bleeding from a lost tooth. Thus, the hemophiliac is forced to live a life of consistent risk. Living a life without even a minor injury is very difficult. A hemophiliac cannot play like other children. They need to be in constant surveillance. Sharp objects have to be kept out of reach. Not only this in case they bleed, but they also have to deal with pain. Bleeding in the joints is eventually crippling. Muscles get wasted.

The treatment of this malady is giving the required deficient factor. So that the blood clots and the bleeding stop in a short while. Besides the Anti-Hemophilia factor VIII and factor IX are imported from abroad. The cost of the factors and other medicines is prohibitive. An adult may need more than 1000 units per shot. The subsidized tae of Rs.5/- unit also becomes expensive for the poorer sections of the society. The subsidized rates are available through Hemophilia Society of India. The cost is nearly 4 times more than this at the retail outlet.

Hemophilia is a congenital lifelong disorder. The goals of therapy are to minimize disability and prolong life, to facilitate general social and physical wellbeing and to help each patient achieve full potential. The cornerstone of hemophilia treatment is an adequate supply of safe effective blood products which must be administered early in the course of haemorrhagic event.

Modern management includes administration of virally safe, effective concentrates of the deficient clotting factor, self-treatment, a professional team approach to the total care of the person with hemophilia and access to genetic technology for carrier detection and prenatal diagnosis. Early and adequate replacement of the deficient coagulation factor is therapy for the majority of bleeding episodes.

Hopefully Government and N.G.O.s will concentrate on this inherited disease at the earliest.