EDITORIAL

HEMOPHILIA CARE IN PAKISTAN

Bleeding disorders are known to mankind since early times. Hemophilia is often called the Royal Disease. It was prevalent in the royal families of Britain, Spain and Russia. Hemophilia is the most common inherited bleeding disorder affecting males predominantly because of X-linked inheritance. Rarely females who are carriers are also affected. Majority of patients are of hemophilia A (FVIII deficiency) and only a handful cases of hemophilia B (FIX deficiency) are seen. There are 178,500 patients of hemophilia in 106 countries as per Annual Global Survey 2014 report of World Federation of Hemophilia (WFH). In this report 549 patients were reported from Pakistan. This figure is much less than the actual burden as there is no national data based registry.

The severity of disease depends on the level of factor deficiency and categorised as mild, moderate and severe. The gold standard for diagnosis is factor assay of the deficient factor. The main stay of treatment in replacement of deficient factor by plasma derived factor or recombinant factor. This is done either on demand or following a prophylactic regimen. World Federation of Hemophilia provides regular management guidelines suitable for source constraint countries and wealthy countries. Hemophiliacs, if inadequately treated develop crippling painful arthopathy, muscle atrophy and intracranial hemorrhages, as they grow. A close cooperation is essential between hematologist, pediatrician, orthopedic surgeon, rehab specialist, psychologist and social welfare service. In developed countries the outlook of hemophiliacs has changed with extended pain free life.

In a developing country like Pakistan, the lifelong management of hemophiliacs is quite challenging and far from being satisfactory. Diagnostic facilities for bleeding disorders are rudimentary and there are only a few centers which extend complete diagnostic and follow up profile for effective management. The availability of expensive replacement factors is very difficult. No hemophilic family is able to bear the cost of factor VIII replacement therapy unless supported by some charity organization. In Pakistan only 0.003% patients are using FVIII replacement therapy and rest of the patients are inadequately treated either with cryoprecipitate or fresh frozen plasma. This inadequate treatment results in development of chronic and fatal complications and disabilities.

Improving access and quality of hemophilia care is a vital yet ignored challenge in this country. There are very few charity organizations like Pakistan hemophilia welfare society (HPWS), who are trying to fight this disease with their limited resources. I think their contribution is immense but the society, NGOs and the government has to come forward to join the fight. It is need of the hour to first develop a National Hemophilia Registry and to register all these patients. Next is the provision of free factor V111/1X to these patients. The government and NGOs have to work very closely in this respect to ensure free supply of these expensive factors. Close linkage with WFH and other international donor agencies is very important.

A dedicated hemophilia care center in each provincial capital with all diagnostic and consultative services under one roof is essential. Besides treatment, patient engagement, education and psychological support is essential for effective care. Improving patient care and quality of life requires an integrated approach among the patient's family, society and the government.

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