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# Editorial

**The Burden of Thalassaemia and Its Prevention: A Neglected Issue in Bangladesh**

Thalassaemias are a heterogeneous group of disorders of hereditary anaemia due to diminished or absent globin chain production. In alpha thalassaemia, alpha globin production is lowered or defective, and in patients with beta thalassaemia, beta globin chain production is lowered or defective. When one class of polypeptide chain is diminished, this leads to a relative excess of other chain. The results are precipitation of unstable haemoglobins, intramedullary RBC destruction, and ineffective erythropoiesis. When there is reduction or absence of alpha chain, it is called alpha thalassaemia and reduction of beta chain leads to beta thalassaemia. Often beta thalassaemia presents in combination with haemoglobinopathies like Haemoglobin E or Haemoglobin S.

Thalassaemia is the most common autosomal recessive disease with a high prevalance in the Mediterranean area, Middle East, Tropical Africa, and Caribbean populations[1]. According to recent data, approximately 300,000 to 500,000 children are born each year with the severe form of these diseases[2].

The World Health Organization (WHO) estimates that at least 6.5% of the world population are carriers of different types of inherited haemoglobin disorders. Thalassaemia is one of the most common inherited diseases in Bangladesh. In a study of Bangladeshi school children, the prevalence of beta thalassaemia trait was 4.1% and the Hb-E trait was 6.1% in Bengali children. Among tribal school children in Chittagong, beta thalassaemia trait was 4.2% and the Hb-E trait was 41.7%. It is presumed that approximately six thousand thalassaemic children are born each year[2].

Treatment of thalassaemia is very cumbersome, hazardous, and costly. Bone marrow transplantation is only a curative treatment, but it is risky and too costly for us. Furthermore, bone marrow transplantation in children was done only in CMH, Dhaka on 26.11.2019[3]. Other treatment modalities include frequent blood transfusions, iron chelating therapy, gamma chain induction, and splenectomy. Blood transfusion itself carries many complications, like infections with HIV or HBV. Patients also require treatment for endocrinopathies, growth failure, heart failure, etc. The cost of supportive treatment for a thalassaemic child in our country requires 70,000 to 90,000 taka per year[4]. So, management of thalassaemic patients puts considerable strain not only on the child but also on the family, community, and nation at large.

Each year, May 8 is observed as World Thalassaemia Day. The day reminds us of the struggles of patients suffering from the disease and helps us to raise awareness about its impact on personal lives as well as national levels. The World Thalassaemia Day theme for 2021 was "Addressing Health Inequalities Across the Global Thalassaemia Community". No doubt, the current theme describes the situation of our parents who cannot bear the expenses, do not have access to available facilities, and do not know how to prevent this calamity. In our country, ninety percent of thalassaemic patients cannot afford adequate treatment. Despite this dire situation, sixty percent of our donated blood is used to help thalassaemic patients[5]. Unfortunately, it is only the tip of the iceberg- 'the patients who require treatment'. The submerged portion of the iceberg (The undetected carriers) goes unnoticed and they are at high risk of giving birth to thalassaemic babies.

Over the past decades, there has been remarkable improvement in the survival of patients with thalassaemia in developed countries. This, however, may not be true for patients who are born in countries where resources are limited. Considering the lifelong sufferings of the patients along with their families and the limited resources of our country, the prime strategies should be directed to controlling the disease burden by raising public awareness, political commitment, improving screening programs, and giving emphasis on marital counseling.

Prevention is the most cost-effective measure for controlling the uprise of thalassaemia. The following strategies may be adopted:

1. Creating awareness: Raising awareness about thalassaemia among the public, policymakers, and health professionals. The media can play a vital role in circulating information about thalassaemia.
2. Mass screening: Mass screening is provided to the general population before and at childbearing age. NESTROF (Naked Eye Single Tube Red Cell Osmotic Fragility) method is nowadays used for mass screening.
3. Target screening: It is restricted to a particular group, such as couples preparing for marriage or before conception or early pregnancy.
4. Genetic counselling: Genetic counseling is provided to carriers and parents of thalassaemic children. It provides full information to the people at risk, gives them time for consideration and supports them in making a decision.
5. Prenatal diagnosis: Prenatal diagnosis can be done around 8-9 weeks of gestation by analysis of amniotic fluid cells or chorionic villi by means of DNA analysis or by the polymerase chain reaction (PCR) method. Selective termination of the affected foetus may prevent the birth of a thalassaemic child. Prenatal diagnosis with the option of abortion of the affected foetus has been very successful in limiting thalassaemic child births in Cyprus, Italy, Greece, and the United Kingdom, leading to the development of control programs in many other countries[1]. Iran has been very successful in decreasing the births of thalassaemic children after the introduction of prenatal diagnosis and a legally approved option for abortion of an affected child[6].
6. Marital counseling: Marriage between two carriers should be discouraged. If necessary, a prohibitory law can be passed in the parliament.

Now it is time for Bangladesh to build up strategies to control disease burden by augmenting public education, screening programmes, and appropriate counselling along with improvements in clinical care.

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