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Thalassemia in Bangladesh:Current Situation and Prevention Strategies

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Abstract: *Thalassemia is a common form of hemoglobin disorders which has been emerged as a global public health concern. It is more common in developing countries than the developed countries.There is a lack of adequate data on the population-based estimation of thalassemia in the country. A scoping study was performed according to York methodology. The study aimed to find out the current situation and prevention strategies of thalassemia in Bangladesh. World Health Organization (WHO) estimates beta-thalassemia carrier and hemoglobin E (HbE) carrier as 3% and 4% respectively in Bangladesh. A study among school children shows that the prevalence of beta-thalassemia trait and HbE trait in Bangladesh are 4.1% and 6.1% respectively. Although WHO has advocated and promoted thalassemia prevention programs from the early 1970s and several countries have already set up comprehensive national thalassemia prevention programs, there is no such program in Bangladesh still now. Considering the ethical issues, government policy, local structures, social values, religion, and cultural tradition of our country some comprehensive thalassemia prevention strategies can be suggested. Health education, awareness, premarital screening and thalassemia carrier identification, vulnerable couple counseling, fetal DNA analysis and family screening program can be taken by the government of Bangladesh. These preventative strategies could be effective for other developing countries as well.*

Keywords: *Thalassemia, Situation, Prevention, Strategies, Bangladesh.*

Introduction

Thalassemia is the commonest form of autosomal recessive disorder worldwide which is characterized by reduction or absent production of one or more of the globin chain that make up the hemoglobin (Hb) tetramers[1]. Two fundamental forms of thalassemia are α-thalassemia and β-thalassemia. Impaired synthesis of α globin chain is classified as α-thalassemia; on the other hand, impaired synthesis of β globinchain is classified as β thalassemia which is the most common type of thalassemia[2].

Now a day, inherited hemoglobinopathies have emerged as a global public health concern. It has been estimated that every year 320,000 babies are born with any type of clinically significant hemoglobin disorder[3]. It's a matter of big concern that about 80.0% of these births occur in low resource countries of the world. A number of studies determined that at least 5.2% of the world population carries hemoglobin variants[3] and more than

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100 million beta-thalassemia carriers with a global frequency of 1.5%[4]. Clinical appearances of hemoglobinopathies are due to the homozygous or compound heterozygous states between certain variants.

South Asia where around 23% (approximately 1.7 billion) of the world's population reside[5] in this region and it is been considered as a hotspot of hemoglobinopathies[3]. Several studies have been conducted in India on thalassemia; as a result, most of the research findings on thalassemia in South Asia come from India. Beta thalassemia carriers have been estimated to be between 2.78% to 4% in India[6,7] that comprises of approximately 30–48 million beta-thalassemia carriers in India. In Pakistan, there are around 5–7% thalassemia carriers which is approximately 5–12 million[3,8]. Although many studies have been conducted in India and Pakistan on thalassemia, there is a lack of researches and adequate data in Bangladesh. This study aims to find out the current situation and prevention strategies of thalassemia in Bangladesh.

Materials and methods

The search for this review was conducted throughout the study period (January to July 2020) to track new developments and published reports as well as articles. The review was carried out according to York methodology, outlined by Arksey and O'Malley, 2005 from the University of York, United Kingdom[9]. A comprehensive search for the published articles, reports and web page documents were carried out. Related articles, reports and web page documents that met the inclusion criteria for this study were selected and reviewed. Papers were included if the study was related to thalassemia issue and if the study was not based on thalassemia they were excluded from the study. Finally, all results were summarized and reported.

Results and discussion

Current situation of thalassemia in Bangladesh

Bangladesh is one of the most densely populated countries in the world having over 160 million population where approximately 6000 thalassemic children are born each year[10, 11].Although Bangladesh lies in the world's thalassemia belt, the information on different aspects of thalassemia like epidemiology, clinical course, mortality, complications and treatment outcomes is not available[12].

In 2005 there were approximately 0.1 million of present thalassemia affected children in Bangladesh[13]. It was an estimation of a research of Khal et al, whereas WHO estimates beta-thalassemia carrier and HbE carrier as 3% and 4% respectively in the country in 2007[7]. However, this estimation has several limitations as was conducted in 1980 and a small number of samples obtained from treatment centers [14]. The only published study on the prevalence of thalassemia (n=735) among the school children in Bangladesh showed the prevalence of beta-thalassemia trait is 4.1%, and a 6.1% prevalence for the HbE traitin 20128. In 2017, there was 6–12% of the population (about 10–19 million people) are thalassemia carrier in Bangladesh[12].

Study also estimated that the regional variation of beta-thalassemia carriers varies from 2.9% to 8.1% and HbE carriers ranging from 2.4% to 16.5% in Bangladesh[8]. The prevalence of HbE was found much higher (41.7%) among tribal children[15]. Another study was conducted by Hasan et al, in 2013 at Dinajpur Medical college hospital where they determined that among 60 patients, HbE disease was 30%, HbE Trait was 41.67% and HbE beta-thalassemia was 23.33%[16]. This situation clearly shows the increasing trend of thalassemia in Bangladesh. It is anticipated that if the situation continued, thalassemia will be a significant burden for Bangladesh.

Thalassemia prevention strategies

Thalassemia is a genetic disorder it can be managed only by preventing it. Considering this issue, some pilot programs on thalassemia carrier screening, counseling and prenatal diagnosis werestarted in several at-risk populations in the Mediterranean area in the late 1970s[17]. Currently, different countries such as UK, France, Iran, Thailand, Italy, Greece, Cyprus, Australia, Singapore, Taiwan, Netherlands, Belgium and Germany, Cuba and HongKong have setup comprehensive national thalassemia prevention program[18]. Despite of having a considerable number of thalassemia patients and carriers in Bangladesh, there is no national thalassemia prevention program.

Considering the context of the country following strategies can be used to prevent thalassemia:

Health education and awareness

Health education will be one of the best approaches to prevent thalassemia. Although, Srilanka provides free thalassemia screening facilities to the general population, due to lack of awareness and proper health education its uptake was very poor [19]. From this situation, it is evident that without proper health education and awareness thalassemia screening program will not be a successful one. Mass health education and awareness should be provided among the general population, health personnel, at-risk people, and also among policymakers. The prevention program should be directed considering the social values, religious laws and cultural characteristics of the country. Intensive monitoring of the program is also required to make the program successful.

Premarital screening and thalassemia carrier identification

Thalassemia carrier detection program should be designed to prevent marriage between two thalassemia carriers. Couples at risk should be determined and they should be monitored closely. Now, premarital thalassemia carrier screening is being performed on a voluntary basis in many countries of the world[17]. The Orthodox Church in Cyprus, requires a certificate of thalassemia carrier before marriage[17, 20]. The national premarital program is mandatory in many Muslim countries like Saudi Arabia, Iran, United Arab Emirates, Bahrain, Tunisia, Lebanon, Qatar and Gaza Strip[21, 22]. The objective of this obligation is to limit carrier marriage. The study observed that there is no national thalassemia screening or carrier identification program in Bangladesh[10]. So, premarital screening should be made compulsory for every citizen of Bangladesh. It can be obtained by making the availability of hemoglobin electrophoresisor high-pressure liquid chromatography (HPLC).

Counseling of at risk couple

Thalassemia prevention program can be directed for genetic counseling of the vulnerable couple. This program may include providing detailed information such as cause, symptom, prevention and prognosis of the disease. Additionally, genetic risk estimation of the parents could be effective to prevent thalassemia.

Fetal DNA analysis

Direct analysis of amplified DNA collected from fetal trophoblast or amniotic fluid cells is now available in some countries like Cyprus, Sardinia, several regions of Continental Italy, Netherland, Belgium and Germany, Northern and South America, Hong Kong, Taiwan, China, Indonesia, Malaysia, Jordan[17, 23]. Chorionic villus-sampling (CVS) at 10 weeks of pregnancy and amniocentesis at 16 weeks of pregnancy will help to detect thalassemia affected child before their birth[24]. The study found that, in 2014, a DNA lab for prenatal DNA analysis by chorionic villus sampling (CVS) and amniotic sampling has been set up in Dhaka Shishu (Children) Hospital, Bangladesh[10]. The lab has been established to detect the status of fetus where the parents are thalassemia carriers and the DNA lab is ongoing[10]. Fetal DNA analysis for prenatal thalassemia detection was installed in Bangabandhu Sheikh Mujib Medical University (BSMMU) for a very short period of time[10].

Recently, some private organization of Bangladesh has started prenatal DNA analysis but it is very expensive and unaffordable for the poor population [10]. So, nationwide parental fetal DNA analysis and fetal cell analysis in maternal plasma along with its technical advancements must be made available and affordable to prevent thalassemia in Bangladesh.

Family Screening

Relatives of thalassemia patients are also susceptible to thalassemia [20]. To prevent this issue comprehensive family screening for detecting thalassemia carriers will be effective.

Conclusion

From this literature review, we found a lack of population-based thalassemia research data in Bangladesh. Based on the available information it is evident that the burden of thalassemia in Bangladesh is rapidly increasing. We should not delay taking proper steps to prevent this important unavoidable public health problem. Thalassemia management and treatment facilities in Bangladesh are not adequate. Besides, its treatment is also very much costly that may fall a thalassemia affected family into financial hardship. Comprehensive thalassemia prevention programs should be taken including health education and awareness, premarital screening and thalassemia carrier identification, vulnerable couple counseling, fetal DNA analysis and family screening.

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