ORIGINAL ARTICLE

THALASSEMIA MAJOR - ADVANTAGES OF SEPARATE THALASSEMIA UNIT WITH REGARDS TO AWARENESS AND COMPLIANCE

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Abstract

Objective: To compare awareness amongst caregivers about thalassemia and the socioeconomic difficulties in routine management of these children in two tertiary care hospitals.

Methods: A cross-sectional study was conducted at two tertiary care government hospitals in Mumbai; one of which has a separate thalassemia unit for the management of thalassemics while the other hospital is treating thalassemic children along with the other routine paediatric cases. A total of 51 patients between 2 to 12 years were enrolled; 25 of which were from the hospital having thalassemia unit and 26 from hospital without a separate unit. Their caregivers were compared for the level of awareness and difficulties in their routine management using a pre-designed questionnaire.

Results: All caregivers were aware about the basic concept of thalassemia; however only 15.4% from hospital without a thalassemia unit were aware about the major complications of the disease like short stature, skeletal abnormalities, cardiac and endocrine problems as compared to 36% from thalassemia unit. Only 37.9% from non-thalassemia unit had sent their immediate relatives for screening as opposed to 62.1% from thalassemia unit, 81.5% caregivers from non-thalassemic unit were irregular in giving iron chelation therapy as compared to only 18.6% from thalassemic unit. The average expense on each visit from a thalassemia unit was Rs.328 as compared to Rs.627 from non-thalassemia unit and 68.4% caregivers lost their daily wages from non-thalassemia unit hospital as compared to 31.6% from those in thalassemia unit.

Conclusion: Overall awareness and compliance to treatment of thalassemia is definitely better amongst caregivers in a separate thalassemia unit. Thus there is a need for more of such units where comprehensive management, economic support and educational awareness for caregivers and patients with thalassemia is provided.

Key words: awareness, compliance, thalassemia unit.

Introduction

Thalassemia is a genetically transmitted disorder characterised by decreased synthesis of either α/β globin chain of haemoglobin causing ineffective erythropoiesis and increased haemolysis with progressive hypochromic microcytic anaemia and its ill effects. Over thirty million people are carriers of thalassemia gene in our country. (1) Every year 10,000 children with thalassemia are born in India, which constitutes 10% of the total number in the world. (2) The requirement of regular blood transfusions every month, expensive treatment of iron overload by chelators, lack of adequate thalassemia care centres, inadequate financial resources for various treatment modalities and above all the lack of knowledge regarding thalassemia itself, affect the child and his family economically and socially leading to irregularities in treatment and consequently adverse outcomes for the child.

Methods and Materials

A cross-sectional study was conducted at two tertiary care government hospitals in Mumbai. One hospital was having a separate thalassemia unit for the management of thalassemics (Hospital B) while the other hospital was treating thalassemic children along with the other routine paediatric cases (Hospital A). ß thalassemia major patients, confirmed by hemoglobin electrophoresis, between 2 and 12 years of age were included whereas those with mixed hemoglobinopathies, non- ß thalassemia and thalassemia trait were excluded. Their caregivers were interviewed using a pre-designed questionnaire after taking written informed consent. The questions were explained and asked to the participants in the language they knew well. Some questions were open ended and the closed ended questions were followed by the most likely options. The questionnaire had following broad categories: 1) Knowledge of thalassemia and its related features. 2) Concepts regarding regular follow up, investigations and chelation therapy. 3) Socioeconomic difficulties in the treatment and awareness about help from Non-Government Organisations (NGOs). 4) Reasons for non-compliance to treatment and follow up.

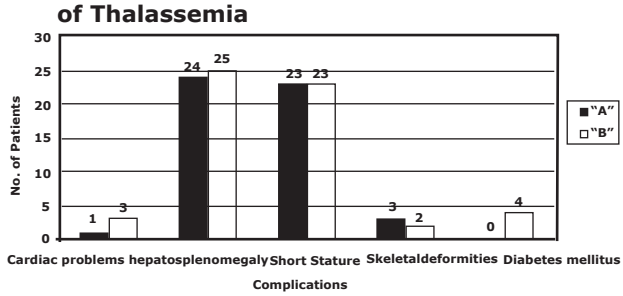
Statistical Methods

Descriptive statistical analysis has been carried out. Data is compared by standard error of difference between two proportions. Significance is assessed at 5% level of significance (p value < 0.05 was considered to be significant).

Results

A total of 51 children were enrolled in the study, of which 26 were from Hospital A and 25 from Hospital B. All of them were aware about the basic concept of thalassemia, that it is a blood disorder in which the formation of blood (red cells) is defective. However a total of only 4 (15.4%) from Hospital A knew about complications of thalassemia like short stature, skeletal deformities, cardiac and endocrine problems as compared to 9 (36%) from B (p<0.001). (Figure 1)

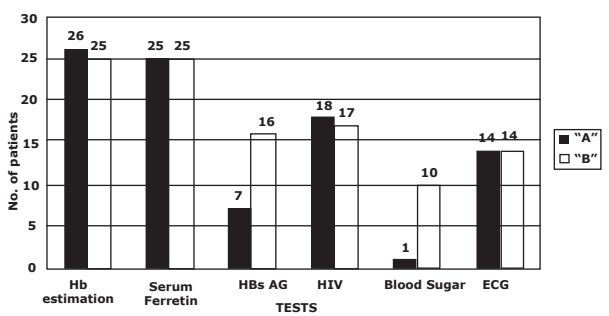
Figure 1 : Awareness about complications of Thalassemia



Twenty (54%) caregivers from Hospital B were aware about antenatal diagnosis of thalassemia as compared to 17 (46%) from Hospital A. Out of all those aware about screening of other family members or their partners before marriage, only 8 (26.7%) were from Hospital A and 22(73.3%) were from Hospital B ($p<0.01$). Only 11(37.9%) from Hospital A had sent their immediate relatives for screening as opposed to 18(62.1%) from Hospital B ($p<0.05$).Commonest reasons for non-screening of relatives were: 1) Relatives thought they can't be affected (50%). 2) Caregivers didn't know they had to send relatives for screening (45.5%). 3) Did not tell relatives since they were already married (4.5%). Only 11 (34.4%) caregivers from Hospital A were aware about bone marrow transplant (BMT) as a permanent cure for thalassemia in comparison to 21 (65.6%) from Hospital B ($p<0.01$).

Awareness about the regular tests to be done for monitoring of complications was as shown in Figure 2.

Figure 2 : Awareness about important tests



Out of all the caregivers who were regularly getting the investigations done as advised by their doctor, 11(30.6%) were from Hospital A and all 25 (69.4%) from Hospital B ($p<0.01$)

Twenty two (81.5%) caregivers from Hospital A were irregular in giving iron chelation therapy as compared to only 5 (18.6%) from Hospital B ($p<0.01$). The reasons for irregularity were high cost of iron chelators (47%), side-effects of drugs (31%), lengthy formalities to get medicines sanctioned from social workers (16%), negligent attitude of caregivers (6%). The common side-effects for which chelators were stopped by caregivers were joint pain (24%), nausea (4%), rashes (2%), burning micturation (1%). None of the caregivers from Hospital A were aware about the concession in chelation therapy offered by the Mumbai Thalassemia Society (MTS) as opposed to 22 from Hospital B ($p<0.01$). Thirteen (68.4%) caregivers from Hospital A had to suffer loss of daily wages when they accompanied their children to hospital as compared to 6 (31.6%) from Hospital B ($p<0.05$).The average expenses on each visit were Rs.627 for Hospital A and Rs.328 for Hospital B ($p<0.001$).

When the caregivers were asked, what more they expected from their treating doctors and the institute in which they were being managed, 68% said they wanted more knowledge about thalassemia and its related features, 27% wanted to speed up the entire process of being admitted and receiving blood transfusions every month, 3% said that the health care systems should publicise more about thalassemia so that more organisations would put efforts to help thalassemics and their families and 2% expected that their physicians spend more time with them and talk courteously.

Discussion

It is very well seen from our study that the overall awareness about thalassemia and its associated conditions, regularity with follow up and investigations, and the compliance to therapy was much better in the hospital with a separate thalassemia unit as compared to the hospital without a unit. A study conducted by Bandyopadhyay et al showed that compared to the thalassemic children attending paediatric ward at R.G. Kar Medical College, Kolkata, those attending thalassemia society of India underwent screening tests for carrier state detection in more numbers. (3)

The possible advantages of a separate thalassemia unit which we found were:- 1) Separate counselling sessions are held for patients when they are first registered in the thalassemia unit and books provided by the thalassemia society are readily available. 2) Patients come together regularly and organize lectures by doctors to increase the knowledge about thalassemia as well as workshops to discuss their problems. 3) Thalassemics of all age groups are given blood transfusions together in a thalassemia unit. Older thalassemics, some of whom are well educated and have access to internet and other information sources, share their knowledge about the disease with younger patients and their caregivers thus building a very healthy atmosphere for increasing awareness. 4) Older thalassemics through compliance with blood transfusions, iron chelation therapy- serve as an ideal example for the younger patients. 5) People from both the higher and lower socio-economic strata come to seek help here so it again increases the flow of information from the more privileged people to the less privileged ones- bridging the gap between them. 6) Forms an emotional support for the patient and his caregivers. 7) In the thalassemia unit, transfusions are given on an outpatient department (OPD) basis and the patient is discharged in the afternoon. So, the expenses on each visit are markedly less when compared to a hospital without a unit. 8) Financial help from the social workers and NGOs for chelation therapy was more readily made available in the thalassemia unit. A similar study carried out by Weinreich et al in Netherlands revealed that the "infotainment" programmes may have a positive effect on people from high-risk group. (4) A study conducted by Dehkordi et al in Iran said that the education whether through booklet or combined method has a significant positive effect on increasing knowledge of the investigated groups. This increased knowledge levels of parents can decrease the burden of thalassemia. (5) One study which closely resembles our research work, is the association between education and thalassemia carried

out by Zaman et al at the University of Peshawar, Pakistan. This research highlights that general health education must be provided to the thalassemics and the registration system must be improved and for this purpose properly trained persons should be appointed. (6) The highlight of this study is that establishment of a separate "thalassemia unit" in hospitals has a powerful impact on the awareness regarding various aspects of thalassemia and on the compliance with therapy, thereby significantly affecting the life expectancy and quality of life of thalassemics. It also prevents the further increase in thalassemia burden by spreading awareness about carrier testing prior to marriage and antenatal diagnosis of thalassemia during early pregnancy. The increase in awareness should be carried out by various infotainment programmes like seminars, workshops and pamphlet distribution in local languages.

Conclusion

Overall awareness and compliance to treatment of thalassemia is definitely better amongst caregivers in a separate thalassemia unit. Thus there is a need for more of such units where comprehensive management, economic support and educational awareness for caregivers and patients with thalassemia is provided.

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