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Knowledge of the Relatives of Thalassemia Patients with Special Regards to Splenectomy as a Treatment Modality: A Cross-Sectional Survey

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ABSTRACT

Background: Thalassemia is a hereditary autosomal recessive haemoglobinopathy that remains a major health problem throughout the world.

Aims and Objectives: The aim of this study was to evaluate knowledge of the relatives of Thalassemia patients regarding different aspects of Thalassemia with special emphasis on the role of splenectomy.

Material and Methods: This was a cross-sectional study conducted at a Government Tertiary Thalassemia day care centre. Initially, 131 relatives of Thalassemia patients were interviewed. Excluding 10 relatives, 87 (66.4 %) relatives having knowledge of splenectomy were further interviewed regarding various aspects of splenectomy.

Result: In our survey, 67.2 % relatives had knowledge about the congenital origin of the disease. The knowledge about premarriage counselling, risk of consanguineous marriage, antenatal screening and medical termination of pregnancy was 55%, 29.8%, 70.2% and 64.1% respectively.

Although all (100%) relatives knew about regular blood transfusion and iron chelation, only 74% and 63.4% of the relatives had the knowledge about splenectomy and bone marrow transplant as a treatment modality, respectively.

In 87 relatives interviewed further, 37.9 % came to know about splenectomy from other relatives. The indications of splenectomy known were: splenomegaly (98.8%), increased blood transfusion (68.9%), low platelet count (20.6%) and low TLC (4.6%). Only 47.1% were aware about the optimal age of splenectomy.

Awareness about serious complication of OPSI, its prevention by preoperative vaccinations and postoperative chemoprophylaxis was only 48.3%, 62% and 46% respectively.

Around 59% knew about role of blood transfusion and 74.7% were aware about the need of regular follow up after splenectomy.

Conclusion: This survey revealed that there is still a lot of lacuna existing in the knowledge of various aspects of Thalassemia and about splenectomy as a treatment modality among relative of Thalassemia patients. Relatives of thalassemia patient having knowledge about splenectomy can be good educators for other relatives and they should be actively recruited as health educators.

Keywords: Thalassemia, Splenectomy, Knowledge, Survey, Prevention and Control

Introduction

Thalassemia is a hereditary autosomal recessive haemoglobinopathy that originated in the Mediterranean region, resulting from the absence or reduced synthesis of either alpha or beta globin chain, categorized into Alpha-thalassemia and Beta-thalassemia respectively. Estimates of expected global annual births with Beta-thalassemia are around 60,000[1,2].

High prevalence is present in populations in the Mediterranean, Middle-East, Transcaucasia, Central Asia, Indian subcontinent and Far East. India is a home to 30–40 million carriers of the disease, with nearly 12,000 infants with Beta-thalassemia being born every year with the major form of the disease[3].

The severe imbalance of globin chain synthesis results in ineffective erythropoiesis. These abnormal shaped red blood cells (RBC) are rapidly destroyed by the reticuloendothelial system, particularly the spleen, leading to severe microcytic hypochromic anemia and early need of transfusion therapy. Patients are managed by repeated blood transfusion to keep hemoglobin (Hb) levels above 9-10gm/dl and iron chelation therapy initially[4].Allogeneic hematopoietic transplantation is the definitive curative treatment available till date[4,5].

Enlargement of spleen in Thalassemia is due to both extra medullary hematopoiesis and entrapment of abnormal shaped RBC.Splenectomy is indicated when blood transfusion exceed 250ml/ kg/year. Hypersplenism, abdominal discomfort due to massive splenomegaly, some degree of neutropenia or th rombocytopenia and splenic injury are other indications of splenectomy in thalassemia patients[6]. Splenectomy has its own complications such as overwhelming post splenectomy sepsis (OPSI), which can be prevented with pre-operative vaccination and post-operative chemoprophylaxis[7].

Patient's quality of life improved after splenectomy because of improved Hb levels. Decreasing blood transfusion requirement helps them to reduce the hospital visits with reduced cost of iron chelation therapy.

As the war against COVID-19 has shown, knowledge and public awareness can greatly help in controlling and reducing the burden of a disease in the society. Creating parental awareness,population screening, genetic counseling, and prenatal diagnosiscan prove to be cost-effective in the prevention of Thalassemia.

Previous studies has been done on relatives of Thalassemia patient regarding their knowledge and awareness of Thalassemia by Saxena et al[8], Biswas et al[9], Maheen et al[10], Ishaq et al[11], Ali et al[12] and Goyal et al[13] and about stress level among caregivers of Thalassemia patient by Inamdar et al[14].

Although, splenectomy has a vital role as a treatment modality for patient with Thalassemia, it was observed that knowledge regarding splenectomy was not evaluated in detail in the previous studies[8,9,10,11,12,13,14].

This survey was conducted with the aim of evaluating the knowledge regarding different aspects of Thalassemia in relatives of Thalassemia patients in our population with special emphasis on the role of splenectomy in management of Thalassemia, and to find out the knowledge vacuity. The results of this survey may help in making strategy for strengthening the existing programmes and formulation of new policies in the direction of reducing the burden of disease.

Material and Methods:

This was a cross-sectional surveyconducted at a Government Tertiary Thalassemia day care centre over a period of three months. Relatives of Thalassemia patients were interviewed in the vernacular language using a structured pretested questionnaire. Only a single relative, preferably the father, of each Thalassemia patient was interviewed. Those relatives having the knowledge of splenectomy as a treatment modality and whose patients had not undergone splenectomy were further interviewed regarding different aspects of splenectomy. Since it was only a survey of relatives, institutional ethical committee approval was not found to be necessary. Data was entered in computerised database and was analysed using pertinent statistical tests. The study variables were presented as percentage and means and standard deviation (+ S.D.)

Result:

In this study, relatives of 131 Thalassemia patients were interviewed. There were 81(61.8%) male patients and 50(38.2%) female patients.The male to female ratio was 1.62:1.The mean (+ S.D.) age of Thalassemia patients was 9.66 (+5.34)years(range: 1-30 years). There were 109(83.2%) male relatives and 22(16.8%) female relatives of Thalassemia patients who were interviewed.The mean (+ S.D.) age of relatives was 36.17(+7.34) years(range: 21-61years).

The socio-demographic profile is summarised in Table-1. Sixty-nine (52.7%) were from rural area. Their level of education ranged from illiterate to postgraduate of which 86 (65.6%) were educated only up to primary level. Ninety two (70.2%) belonged to lower class as per Modified BG Prasad Socio-economic Classification, Update-2019[15].

Table 1: Socio-demographic profile of Thalassemia patients and their relatives (n=131)

Parameter	Number (n= 131)	Percentage (%)
Gender of Thalassemia patient		
Male	81	61.8
Female	50	38.2
Gender of relatives		
Male	109	83.2
Female	22	16.8
Relationship with patient		
Father	105	80.2
Mother	21	16.0
Grand Mother	1	00.8
Uncle	4	03.1
Religion		
Hindu	106	80.9
Muslim	25	19.1
Place of residence		
Urban	62	47.3
Rural	69	52.7

Education level of relatives		
Primary	86	65.6
Secondary	23	17.6
Graduate	12	09.2
PostGraduate	7	05.3
Illiterate	3	2.3
Socioeconomic status;Average Per Capita Income Per Month (INR)as per Modified BG Prasad Socio-economic Classification, Update-2019[15]		
Upper class (7008 and Above)	-	-
Upper middle class (3504-7007)	1	00.8
Middle class (2102-3503)	9	06.9
Lower middle class (1051-2101)	29	22.1
Lower class (Below 1050)	92	70.2

The finding of knowledge of relatives of Thalassemia patients regarding the disease and its various treatment modalities is presented in Table 2.

Table 2: Knowledge of Relatives of Thalassemia patients regarding the disease and its various treatment modalities (n=131)

Knowledge regarding Thalassemia in general	Response (n=131)	Percentage (%)
Cause of Thalassemia		
Congenital	88	67.2
Destiny/Bad Karma	15	11.5
Contact with other Thalassemia patients	2	01.5
Any other	3	02.3
Don't know	23	17.6
Knowledge about premarriage counselling		
Yes	72	55.0
Don't know	59	45.0
Knowledge about rrisk of consanguineous marriage		
Yes	39	29.8
Don't know	92	70.2
Knowledge about aantenatal screening		
Yes	92	70.2
Don't know	39	29.8
MTP, if prenatal diagnosis revealed thalassemia		
Yes	84	64.1
Don't know	47	35.9
Different treatment modalities of thalassemia*		
Regular blood transfusion	131	100
Iron-chelation therapy	131	100
Splenectomy	97	74.0
Bone-marrow transplantation	83	63.4

Knowledge about complications of regular blood transfusion		
Yes	88	67.2
Don't know	43	32.8

*Multiple responses

In our study, only 67.2 % relatives had knowledge about the congenital origin of the diseaseand 55% and 29.8% knew about the importance of premarriage counselling and risk of consanguineous marriage, respectively. Although, 70.2 % had knowledge regarding antenatal screening, only 64.1 % knew about the option of medical termination of pregnancy (MTP) if prenatal diagnosis revealed Thalassemia.

As treatment modality, all (100%) had knowledge about both regular blood transfusion and iron chelation therapy and 74% knew about splenectomy, while 63.4 % were aware about bone-marrow transplantation. 67.2 % relatives were aware about complications of regular blood transfusions.

In our study, 97(74%) caregivers of Thalassemia patients knew about splenectomy as a treatment modality of Thalassemia; out of these,splenectomy has been already done in 10 (7.6%) patients. Excluding them, we further interviewed the remaining 87 (66.4 %)relativesof Thalassemia patientswho had not undergone splenectomy, about the various aspects of splenectomy in Thalassemia.

The findings of knowledge of relatives of Thalassemia patients with special regards to Splenectomy as a treatment modality are given in Table 3.

Table 3: Knowledge of relatives of Thalassemia patients with special regards to Splenectomy as a treatment modality (n=87)

Knowledge regarding Splenectomy	Response (n=87)	Percentage (%)
Source of knowledge about splenectomy*		
From other relatives (All)	33	37.9
(A. whose patient had underwent splenectomy)	(31)	(35.6)
(B. whose patient had not underwent splenectomy)	(2)	(02.3)
From treating doctor	31	35.6
From books/media	25	28.7
From nursing staff	4	04.6
Indications of splenectomy*		
Increased blood transfusion	60	68.9
Large/huge spleen/splenomegaly	86	98.8
Decreased TLC	4	04.6
Decreased Platelet count	18	20.6
Optimal Age for splenectomy, in completed years		
<5	-	-
>5	41	47.1
Don't know	46	52.9
Complications of splenectomy*		`
Injury to other abdominal organs	31	35.6
Chest infection	15	17.2
Wound infection	53	60.9
Incisional hernia	3	03.4
Malaria	27	31.0
OPSI	42	48.3
Mortality	7	08.0
Don't know	28	32.2

Institutions where splenectomy facility is available*		
Government hospitals in Same city	75	86.2
Government hospitals outside the city	19	21.8
Private hospitals in the same city	16	18.4
Private hospitals outside the city	3	03.4
Don't know	12	13.8
Preoperative vaccinations		
Pneumococcal	19	21.8
Meningococcal	-	-
Haemophillus influenzae	-	-
All	35	40.2
Don't know	33	37.9
Postoperative chemoprophylaxis		
Antibacterial	1	01.2
Antimalarial	9	10.3
Both	30	34.5
Don't know	47	54.0
Role of blood transfusion after splenectomy		
Yes	51	58.6
No	36	41.4
Need of follow-up after splenectomy		
Yes	65	74.7
No	22	25.3
Follow up speciality after splenectomy		
Only in Surgery	6	06.9
Only in Paediatric medicine	3	03.4
Both	64	73.6
Don't know	14	16.1

*Multiple responses

In this survey,relatives of other Thalassemia patients 33(37.9%) were the foremost source of knowledge regarding splenectomy (Table 3). Out of these 33, relatives of patients who had underwent splenectomy in the past were the main (n = 31, 93.9%) source of knowledge regarding splenectomy.

Splenomegaly was the most common (98.8%) known indication of splenectomy with low TLC (4.6%) as the least known.Less than half (47.1%) were aware about the optimal age of splenectomy.

Fifty nine (67.8%) relatives were aware about at least one complication of splenectomy, like wound infection (60.9 %), OPSI (48.3 %), injury to other abdominal organs (35.6%), malaria (31.6 %), etc., but still a large number of relatives (32.2%) had lack of such knowledge

Most of the relatives (86.2%) knew that splenectomy facility available in government institutions in the same city, while 13.8% were uninformed about any types of available facility.

Fifty four (62.1%) participants had knowledge about some preoperative vaccinations but only 40.2 % were aware about all the three types (Pneumococcal, Meningococcal, Haemophillus influenzae)of vaccinations, while 37.9% had no knowledge.

In the present survey, only 46% relatives knew about postoperative chemoprophylaxis while 54% participants had no knowledge regarding this. Only 34.5% were informed about both antibacterial and antimalarialagents as chemoprophylaxis.

Only 58.6% were aware about role of regular blood transfusion even after splenectomy. Majority (74.7%) had knowledge about need of post splenectomy follow up and73.6 % had knowledge about attending both Paediatrics Medicine and Surgery department.

Discussion:

In the present survey,we had male predominance with 61.8 % male and 38.2 % femaleThalassemia patients, which had similarities in male predominance with findings of Goyal et al[13], Bandyopadhyay et al[16] and Saxena et al[8].

In our study, 83% of relatives were males; this finding wasmuch more as compared to Biswas et al[9] (24.1%).

Only 67.2 % of the relatives knew that Thalassemia is a congenital disease, which had similarities with findings of Inamdar et al[14] (68.5%) and Aggarwal et al[17] (60.2%).The finding was better compared to Saxena et al[8](47.5%), Ishaq et al[11] (44.6%) and Biswas et al[9] (47.6%) but less compared to Ali et al[12] (82.0%).

Only 29.8 % knew about consanguineous marriage as a risk factor, which was poor as compared to the study reported by Basu et al[18] (53.27%). Fifty five percentage knew about premarriage counselling; the finding was more as compared to Inamdar et al[14] (31.4%) and similar to Biswas et al[9] (52.4%) but less compared to Ishaq et al[11] (84.3%).

Around 70% were aware regarding antenatal screening, which was similar with the study by Ali et al[12] (74%), but in the study by Saxena et al[8] only 45% were aware of antenatal screening of Thalassemia.Around 64% of relative knew about MTP, which was less compared the study by Basu et al[18] (81.31%).

In our survey,all relatives knew about regular blood transfusion and iron chelation are the treatment of Thalassemia which was better compared with study of Biswas et al[9] (75.9%) and Inamdar et al[14] (77.1%).

Majority of the survey participants (74%) knew splenectomy as a treatment modality of thalassemia which was far better than findings of Biswas et al[9] (19.2%) and better than Goyal et al[13] (66.0%). Knowledge of bone marrow transplantation was also present in 63.4 % which was more than Inamdar et al[14] (45.7%) and much more compared to findings of Biswas et al[9] (2.7%).

In our survey, out of 131, only 87(66.4%) participants were interviewed further, who had the knowledge about splenectomy, and their patients had not underwent splenectomy.

Relatives of the other thalassemia patient (37.9 %), treating doctors (35.6%) and books/ media (29%) played a crucial role as a source of knowledge about splenectomy. Relatives of the thalassemia patient who had undergone splenectomy (35.6 %) were a good source of knowledge for other relatives and they should be actively recruited as health educators. Surprisingly the role of nursing staff (5%) was very less.The nursing staff should be trained and motivated to play the role of health educators also among relatives of Thalassemia patients.(Table-3).

Findings of our survey revealedthe commonly known indications of splenectomy in relatives were presence of a large/ huge spleen (98.8%) and increased requirement of blood transfusion (68.9%). There is still a need of creating awareness among the relativesabout less recognized indications like the low platelet count(20.6 %) and low TLC (4.6 %).

In our survey, around 47% relatives knew that splenectomy should be done after 5 years of age, but still around 53% relatives were uninformed about this. Around 40 % of the relatives were under misconception that there will not be any requirement of blood transfusions after splenectomy, which should be removed, although around ¾ were aware about the need for regular follow up after the operation.

Conclusion:

This survey revealed that there is still a lot of lacuna existing in the knowledge of various aspects of Thalassemia and about splenectomy as a treatment modality among relative of Thalassemia patients.We should strengthen the health education activities among them as there should be 100% knowledge about all aspects of Thalassemia, if we are to think ofbetter prevention and treatment of this disease.Relatives of thalassemia patient having knowledge about splenectomy can be good educators for other relatives and they should be actively recruited as health educators.

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