

Thalassaemia: A Study on The Perception of Patients and Family Members

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SUMMARY

Marked improvement in the management of thalassaemia has not been matched by progress in psychosocial rehabilitation as thalassaemia continues to pose challenges to patients and their family members. Few studies have been carried out in Malaysia to look at such issues. This study is therefore to explore the concerns, beliefs and feelings about thalassaemia. It was conducted in the year 2009 over 7 months on "focus groups", in patients aged 8-22 years and parents attending Paediatric Clinic of Tengku Ampuan Afzan Hospital, Kuantan, Pahang. Results showed that concerns and adverse impact were related to lower grades in education, poor self-image, less chance of employment, marriage, financial burden and social integration. Compliance to subcutaneous iron chelator was poor. There were various concerns related to blood transfusion therapy. It is evident that thalassaemia greatly affects the psychosocial dimensions and a more structured long term psychosocial support is needed to improve quality of life of patients.

KEY WORDS:

Thalassaemia, Perception, Patients, Families

INTRODUCTION

Significant advances have been made over the years on thalassaemia leading to better understanding of genetic control of haemoglobin, the abnormalities in the different forms, the pathophysiology of the disease and hence improvement in treatment¹. This marked development in the diagnosis and management has not been matched by progress in psychosocial rehabilitation of thalassaemia patients². Only limited progress has been made in the psychosocial developmental aspects of care in families with afflicted members^{3,4,5}. Thalassaemia poses challenges to patients and family members at the physical, emotional and cognitive levels leading to disruption of their normal psychosocial life. As such more attention is needed in issues related to knowledge and understanding of the illness, lifelong compliance and adherence to chronic treatment regime and attitudes of patients and family members as well as quality of interpersonal relationships.

In Malaysia thalassaemias occur mainly among the Malay and Chinese community. Nearly 150-350 babies are born with

thalassaemia each year⁶. There are a total of 4,541 thalassaemia patients registered in the Malaysian Thalassaemia Registry till August, 2009. Of these 3,310 patients are transfusion dependent β thalassaemia major and Haemoglobin E (HbE) β thalassaemia patients, 455 patients with thalassaemia intermedias and 410 Haemoglobin H (HbH) disease⁷. It is therefore a health burden that needs to be addressed. As some degree of progress has been achieved in the clinical management of patients affected with thalassaemia in Malaysia morbidity and mortality related to thalassaemia is expected to reduce significantly. The quality of life can therefore now be considered an important index of effective health care.

Very few studies have been carried out in Malaysia thus far in relation to psychosocial aspects of thalassaemia patients and their immediate family members^{6,8}. Findings of studies conducted elsewhere may not reflect the true picture in Malaysia. Therefore this study aims to explore the concerns, beliefs and feelings of patients and their immediate family members.

MATERIALS AND METHODS

This study involved thalassaemia patients and their parents attending the Paediatric Clinic of the Department of Paediatrics, Tengku Ampuan Afzan Hospital (HTAA), Kuantan, Pahang, Malaysia. It was conducted following research and ethical committees' approval. A qualitative approach focus groups was used as it allowed participants the opportunity to provide their personal perceptions and to share commonalities and differences in experience about thalassaemia.

Patients and parents were invited to participate in the study via phone calls by a research assistant. Subjects who agreed to participate then provided their contact details and the focus group discussions was arranged. Written consent was obtained on the day of the scheduled focus group discussion. Travel reimbursement and refreshments was provided. The enrolled patients were divided into 5 focus groups according to their age groups; Group 1 - male and female patients aged 7-12; Group 2 - male patients aged 13-17; Group 3 - female patients aged 13-17; Group 4 - female patients aged 18-22 and Group 5 - male patients aged 18-22. There were also 5 parallel groups of parents.

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A question line was developed based on the objectives of the study (Table I). Pilot interviews were conducted with pre-identified thalassaemia patients in order to refine the question line and manner of questioning, by the facilitators.

In all 10 sessions of focus group discussions involving different groups were carried out from February 2009 to August 2009. The number of participants for each group ranged from 3-9. Two researchers acted as facilitators for each focus group. The sessions were conducted in Malay. The duration of each interview session lasted for one hour and it was video-taped.

Digitally recorded focus group discussions were transcribed and then rechecked for accuracy. Patients and family members in the study were given the opportunity to read or comment on the transcripts.

Two researchers independently coded the transcripts using a pre-developed code list derived from the literature and the pilot interviews. To ensure consistency in interpretation, all researchers discussed the coding and the emergent themes until consensus was reached. NVivo software (QSR International) was used to organize data. Illustrative quotes are reported verbatim (translated to English) to support the discussion. All data is handled and kept by the principal researcher.

RESULTS

In all 21 patients and 24 parents/guardians participated in this study. The demographic characteristics and backgrounds of these participants are shown in Table II. There were a total of 70 thalassaemic patients attending the Paediatric Clinic of HTAA, of which 21 were below the age of 7 years and therefore were not eligible to be included.

Six clear themes emerged in relation to concerns raised by thalassaemia patients and their parents. The themes and representative quotes illustrating the themes are summarised in Table III and discussed below. Quotes are identified by source (e.g. Group 1- patients, Group 1-parents).

1. Concerns

Education

Pursuing further studies

Majority of the parents were concerned about their thalassaemic children leaving home to further their studies in colleges or higher institutions. Issues raised were related to the well-being of their children and compliance with medication. The patients themselves if given the opportunity would have liked to be able to leave home to pursue education, but were discouraged by parents due to concerns over health. Although thalassaemic patients wanted to be away from home and reside in hostels, they were rather apprehensive about the perception of other students towards them. A specific point was related to the use of needles for subcutaneous administration of iron chelator. Patients feared of being accused of being drug abusers.

Academic performance and achievement

Both patients and parents believed that the disease did influence the academic performance. The patients had

performed less well academically, mostly as a result of having to be away from school for follow-up visits, blood transfusion in the day-care centre or admission to the ward. Such absence seemed to have an impact on the academic achievement. Periods of absence varied from one day to more than a week per month. Some of the patients also expressed their frustration regarding their poor achievement.

In addition many parents attributed the poor academic performance of patients to tiredness and lethargy. These symptoms were more prominent nearer to the time for the subsequent transfusion. There was also general perception among the parents that thalassaemia itself directly caused their children to be slow learners.

School support

It was generally agreed that the schools were supportive of thalassaemic children but the teachers could do more to help in improving the academic standard of patients.

Involvement in sport and co-curriculum activities

As a result of tiredness and lethargy most of the patients were not fully involved in sports or co-curriculum activities.

Self / Body image

Short stature/Growth failure

Not attaining the average height compared to their peers was perceived as a major problem by both parents and patients. This point was frequently raised by both parties during discussion. Patients were being called by 'nick names' and teased by their peers for their shorter stature. They were also constantly being compared to other siblings at home about their height and thus not respected by younger siblings. As a result of these perceived issues they felt anxious, stressed-up and admitted that it lowered their self-esteem. In addition due to this 'child-like' appearance, patients above 17 years of age refused to be followed-up in the adult medical clinic and preferred to remain under the care of paediatricians.

Thalassaemic facies and skin colour

Patients felt a degree of stigmatisation as a result of their "dark" skin colour. The thalassaemic facies also contributed significantly to the negative body image.

Delayed menses

The parents of female adolescent patients were very concerned about their children not attaining or delayed menarche as this may influence their ability to have a family of their own in future.

Employment

The difficulty of obtaining a job was expressed mainly by patients who had studied up to only secondary school. Patients who were working admitted to having difficult relationship with their employers as they had to take time off work frequently for transfusion and other treatment.

The parents of young children also had problems with their own employers mainly related to getting time off to accompany their children for treatment. Parents felt that employers did not understand or refused to understand their predicament. As such they had to plan their annual leave well ahead. Most of their leave days would have to be reserved for

their children except for other urgent matters. This was a more serious issue for parents working in private companies. As a result some parents preferred to be self-employed.

Marriage and starting a family

Future spouse

Even though the patients were interested in looking for a life partner, they could not help but express their worry about being rejected by partners because of their illness. All patients agreed however that they should be truthful to their potential spouse about their illness. The parents of adolescence female and male patients also voiced similar concern.

Effect of thalassaemia screening results

Six parents indicated during the discussion that if they had known that they were carriers they would have changed their decision about marrying their present spouse. However some (4 parents) said they would have proceeded with the marriage but refrained from having children. Parents voiced the pain of having to witness the suffering of their thalassaemic children as a result of them not knowing about their thalassaemic carrier status prior to marriage. They wished that the public awareness campaign of thalassaemia by the appropriate authorities had started very much earlier.

Financial issues

All parents were of the opinion that financial burden was an important issue. For some parents financial difficulty was the expenditure incurred when going to hospital for follow up visits. Also for some them any loss of working days meant financial loss as well. Being admitted to the ward posed further financial constraints due to additional hospital bills.

Adolescence patients felt the need to be financially independent as an important issue. They preferred to be independent and not cause burden to their parents.

Relationship and social integration

Family members

Although patients had good relationship with siblings and parents, a few problems were highlighted. Two examples were being teased by family members about their dark-skin colour and short stature. Some patients viewed that because of their low academic achievement they were not treated fairly by parents.

Peers

About one third of the parents (9 of 24) claimed that their ill children did not have many friends and were virtually housebound. They believed that generally it was the normal children that did not want to befriend the patients. Patients attributed the illness as an obstacle for them to integrate socially.

Problems of communication

Parents of adolescent/young adult patients (9 of 15) commented that most of their thalassaemic children were not keen on discussing about their disease. The patients themselves felt that discussing about the illness would not change their predicament. Four of the parents suggested that it would be extremely helpful if professional motivation and counseling service for the patients are provided by health

authorities.

Self esteem

Most patients (13 of 21 patients) had poor self-esteem and were emotionally affected. They had mixed feelings namely sadness and anger. Two of group 5 parents reported that their children preferred not to disclose their illness to others as they were embarrassed.

2. Treatment

The emerging themes related to beliefs and feelings about the long-term treatment of thalassaemia patients are categorised into those related to blood transfusion, subcutaneous iron chelation therapy and oral medication.

Blood transfusion

Fear related to complications of blood transfusion

Parents constantly worried about the adverse effects related to transfusion. Some of their concerns were:

'I am afraid if something happens during the blood transfusion. I am scared because the risk is high isn't it? Afraid that the blood is no good. Scared of AIDS or anything else.' (Group 3 – parent)

There was increased anxiety each time the children received transfusion:

'...I feel better if he is not transfused for that visit. What I meant is that we do not know the source of the blood, whether it comes from a good source or otherwise. That's why I feel better when a transfusion is not done. But when it is, I am anxious. I think about all sorts of bad things. I am calmer when he does not receive the blood transfusion.' (Group 3 – parent)

Weary of blood transfusion

Adolescence patients indicated that they were bored with having to adhere to the long-term treatment and in particular the innumerable needle pricks:

'Endless needle pricks! Endless needle pricks! We want to be free too, right? Others are. But we are not.' (Group 5 – patient)

Changes of behavior post-transfusion

Parents reported changes of behaviour in their children following transfusion. They either became more aggressive or quieter:

'Sometimes she became irritated after a blood transfusion. Other days, she would be mellowed. Is it due to the blood itself? Sometimes she sulks all day.' (Group 1 – parent)

'Sometimes he became angry after the transfusion. Sometimes, he became quiet. That is how it is.' (Group 1 – parent)

They believed that it was related to the temperament of donors of the blood units:

'There's blood which made him go berserk and there's blood which made him good. A lot of characters.' (Group 1 – parent)

Subcutaneous iron chelation therapy

Compliance

Generally compliance to subcutaneous iron chelation therapy was poor. This was due to several factors such as the side-effects, simply not wanting to administer, feeling of difficulty

and burdensome and no one to perform the subcutaneous injection:

'Sometimes, she does not want the injections. She developed rashes.' (Group 1 – parent)

'At times, he would allow us to inject him, but not all the time. When he does not want it, we don't give it to him. I think it easier if he can take the oral medication. Sometimes, these injections become a reason for quarrel.' (Group 1 – parent)

'Yes, he doesn't want to do it. He said it's easy for me to say. Try doing it yourself, he said.' (Group 3 – parent)

'He doesn't take the injections. He simply keeps the medications at home. Don't want the injections, painful. At the hospital, he will collect the medication every month, but administer only once during the month.' (Group 2 – parent)
'Previously her auntie use to do the injection. But when the auntie got married, she doesn't want to do it. Me...I'm afraid. I pity her.' (Group 2 – parent)

'Yes, I used to take injections. But not now. Not since I enrolled in the university. I don't know, it feels awkward and cumbersome.' (Group 5 – patient)

'Since I stay in the hostel, I do not bring them. It's not that I'm embarrassed. Not like that. But sometimes I might be busy. Don't know how to say it...' (Group 5 – patient)

'As for me, my mother will do the injections. But only when she is in the mood. If she does not, then she would ask me to do it myself.' (Group 4 – patient)

Oral medication

During interviews generally both parents and patients were referring to folic acid and oral penicillin for patients who had had splenectomy. Only 2 patients were on Deferasirox. Seven other patients were on Deferiprone. Compliance with oral medication was variable though generally better than that of subcutaneous iron chelator. The patients felt having to be on medications posed some hindrance to their social activities.

Alternative and complementary medicine

Parents of 12 thalassaemic children opted for alternative and complementary medicine in addition to the treatment provided in the hospitals at the initial stage of the diagnosis. These included Chinese traditional medicine, herbal medicine as drinks or as topical application in the splenic area, goat's milk, 'spirulina' and a type of fresh water oyster. Treatment from Malay traditional healer was also sought.

Parents and patients generally were able to cope with the disease through strong faith in religion. All patients were Muslims. They strongly upheld the concept of *Qada' and Qadar* in dealing with the disease. The parents and the patients themselves were hopeful that one day the patients would be cured of their disease although they were aware that with the modes of treatment received cure is not possible.

DISCUSSION

This study used focus groups because they provided the opportunity to reinforce cultural concepts of sharing and learning about thalassaemia from others. This method allowed the researchers to interact directly with the patients and family members and hence it gave opportunities for the clarification of responses, for follow-up questions and for the probing of responses⁹.

Although the small numbers of participants in focus groups limited the generalization of outcome to larger population, our study revealed several important findings. It is essential to note that participants in these focus groups came from diverse groups i.e. a mixture of thalassaemia intermedia and thalassaemia major patients. Only 2 patients had good iron chelation treatment compliance while others generally had poor compliance. In addition patients below 7 years of age were excluded because the method used was deemed not suitable for this age group.

The most significant finding was about the issue of psychosocial problems of the thalassaemics patients and their family members. It illustrated how 'clinical burden' of a disease like thalassaemia immensely affected the psychosocial aspects of families.

Few similar studies have been conducted in Malaysia and as such only limited comparison could be made. Ratip *et al.* found similar problems among thalassaemia intermedia patients¹⁰. Khurana *et al.* and Gharaibeh H *et al.* also showed similar areas of concern in transfusion dependent beta-thalassaemia major patients^{11,12}.

In our study poor academic standing among the patients was believed to be related mainly to absence from school as also reported by Canatan *et al.*¹³. Another study in Malaysia also showed that thalassaemia affected patients' academic ability and indicated that further studies needed to be undertaken to see if absenteeism does affect academic achievement in schools⁶.

Many parents perceived the anaemic symptoms as another factor that contributed significantly to the poor academic performance in their thalassaemic children. This therefore raises the question about the pre-transfusion haemoglobin level of these children and hence compliance with follow up visits.

It is also interesting to note that parents believed that thalassaemia itself causes their children to be slow learners. Thalassaemic children, more so those who were iron overloaded, have been demonstrated to have impaired abstract reasoning, deficits of language, attention, memory, constructional/visual spatial skills, and executive functions¹⁴. It is therefore essential for us to look more closely at this issue as optimal treatment should be able to improve their academic performance. Amstrong has suggested screening children with thalassaemia for specific neurophysiological impairments and that early intervention and special education access under the 'Individuals with Disabilities Education Act' are provided¹⁵.

Table I: Question line utilized to guide the focus group discussion

Purpose	Question <i>(The questions served as the agenda for the group discussion and were elaborated accordingly during the discussion)</i>
<p>To assess patients and parents knowledge and understanding of thalassaemia:</p> <ul style="list-style-type: none"> • Hereditary factor or other causes as perceived by patients or parents. • What is basically wrong? What does it lead to? • Clinical features/Complications/Treatment options. 	<p>What do you (parents /patients) understand about thalassaemia? <i>(Apa yang tuan/puan/adik-adik faham mengenai thalassaemia?)</i></p>
<p>To examine concerns and problems expressed by patients and parents of thalassaemia:</p> <ul style="list-style-type: none"> • To address both current and future concerns. 	<p>What are your (parents /patients) concerns? <i>(Tentu ada perkara yang merunsingkan tuan/puan/adik-adik penyakit ini?)</i></p>
<p>To explore the psychosocial problems experienced by patients and families with thalassaemia:</p> <ul style="list-style-type: none"> • To address the following areas and other areas as raised by parents and patients in relation to the disease: Education, anxiety, body image, secondary sexual characteristics, self-esteem, communication, support, relationships, social interaction, marriage, employment and employer-employee problems 	<p>Could you elaborate on problems you (parents /patients) have faced or are going through presently in relation to thalassaemia? <i>(Apakah masalah-masalah yang tuan/puan/adik-adik telah dilalui dan sedang hadapi?)</i></p> <p>How do you see yourself (patients) as compared to others? <i>(Bagaimana anda melihat diri anda berbanding dengan orang lain?)</i></p>
<p>To explore beliefs and feelings about adherent to the long term-treatment:</p> <ul style="list-style-type: none"> • Blood transfusion • Subcutaneous/oral iron chelators • Alternative and complementary medicine • Perception of the disease itself 	<p>What are your problems (parents/patients) in relation to the treatment? <i>(Apa masalah yang tuan/puan/adik-adik hadapi berkaitan dengan rawatan penyakit ini?)</i></p> <p>What are your feelings (parents/patients) about adhering to the long-term treatment? <i>(Apakah perasaan tuan/puan/adik-adik memandangkan rawatan penyakit ini adalah sepanjang hayat?)</i></p> <p>How do you cope with the illness? <i>(Bagaimana tuan/puan/adik-adik menghadapi keadaan ini?)</i></p>

Table II: Socio-demographic characteristics and backgrounds of the parents/guardian and patients

	Group 1	Group 2	Group 3	Group 4	Group 5
Patients					
Number of patients	6	5	4	3	3
Age	8-12	14-18	15-17	19-22	18-20
Gender	Male (3) Female (3)	Male (5)	Female (4)	Female (3)	Male (3)
Designation	Primary school student (6)	Secondary school student (5)	Secondary school student (4)	Unemployed (2) Waitress (1)	
Diagnosis	β Thalassaemia major (1) β thalassaemia intermedia (2) HbE-β thalassaemia (3)	β Thalassaemia major (1) HbE-β thalassaemia (4)	HbE-β thalassaemia intermedia (1) Hb H disease (1) HbE-β thalassaemia major (1) β Thalassaemia major (1)	β Thalassaemia major (3)	HbE-β thalassaemia intermedia (2) HbE-β thalassaemia major (1)
Parents					
Number of parents	9 (5 fathers, 4 mothers)	5 (4 fathers, 1 guardian)	4 (mothers)	3 (2 mothers and 1 guardian)	3 (fathers)
Number of children affected with thalassaemia	1-2	1-2	1-3	1-2	1-3
Occupation	Housewife (2) Self-employed (1) Clerk (2) Factory –operator (1) Laundry –assistant (1) Supervisor (1) Labourer (1)	Housewife (1) Cargo investigator (1) Fisherman (1) Supervisor (1) Clerk (1)	Housewife (4)	Housewife (1) Unemployed (1) Babysitter (1)	Assistant manager (1) Manager (1) Army officer (1)

Table III: Representative quotes from patients and parents illustrating the emerged themes related to concerns from focus group discussion

I Education:

Pursuing further studies

'Because I didn't like the idea of sending my child to a better school. I want to be close to her. I want her to stay at home. Told her that if I send her to a boarding school, it would be difficult to do the injections and to take the medications. I had to make a choice so that is why she is still at home.' (Group 5 – parent)

'Have been offered a place to study in KL the other day but mother said that it was too far away...to enter into a college is also not possible. Mother is undecided. She's afraid that there will be no one to help me if I fall sick. Also thinking of the injections...who would send me to the hospital.' (Group 4 – patient)

'I feel embarrassed to bring the injection needle everywhere. Afraid that people might mistaken me for a drug addict. Have overheard people saying that I do drugs. They do not know that I suffer from thalassaemia.' (Group 4 – patient)

Academic performance and achievement

'I get E for everything, nothing can get into this head of mine. If I missed school or assignments, there's nothing I can do. When I ask from the teacher about the school work that I missed, he does not want to assist. It's my fault that I missed them he said.' (Group 4 – patient)
Others can go study abroad...everyone who has finished school can further their studies together. We are not able to go anywhere. Not only to continue our studies, but to work is also difficult. (Group 4 – patient)

'Maybe...I also think that ...but to me, this is probably due to the thalassaemia because she understands what she has learned but then she is slow compared to others.' (Group 1 – parent)

'Like my child, he is weak in his studies. He goes to school but still he is a bit weak. He does study but he is slow in grasping all the information.' (Group 3 – parent)

School support

'The teachers in the secondary school, some of them do know about my condition but some do not. For those who do not know, they don't really care as long as you give them the medical leave certificate.' (Group 5 – patient)

'...like her, she went to school up until Form 3 only. At school, she is not even able to climb up to the upper floors as she does not have the energy.' (Group 4 – parent)

Involvement in sport and co curriculum activities

'All kinds of activities and sports cannot participate, for example, police cadet society also camping.' (Group 5 – patient)

II Body image:

Short stature/Growth failure

'And then, there is that issue where upon going into Form 1 of secondary school, his friends are much taller. But he is still at that height. Sometimes it's even difficult for him to ride the motorbike although he still tries. He doesn't want to be left out or even feel left out because he thinks that if the others could do it, he could also...at one point, I have even asked the doctor whether my son can grow any taller.' (Group 3 – parent)

'My daughter is 22 but she is small for her age. Like a year 1 or year 2 student. She is uncomfortable when she's out with her younger sisters, as she is the smallest. People will always think that she is the younger sister so that's why she's uncomfortable going out in public. On occasions such as 'Eid', she feels embarrassed receiving money from the adults as they thought that she is young.' (Group 4 – parent)

'Looking at the others, they are all bigger. I feel a bit of stress due to my small size.' (Group 5 – patient)

'I am like the same age as my younger sisters. But in actual fact, she is much younger than me....they don't seem to give me the respect I deserve being the older sister.' (Group 3 – patient)

'The average person is normal. But we are like primary school children. Feel different. Not the same. It is as if we are too heinous (or dreadful)..."' (Group 4 – patient)

'If the doctor wants to refer us to the clinic for adults.....eeee, I don't think I will want to go. It will be weird. People will question.' (Group 4 – patient)

'Like when I was admitted to the surgical ward for splenectomy, I heard people exclaiming surprise over why a child is admitted into an adult ward!' (Group 4 – patient)

Thalassaemic facies

'People say that thalassaemia patients can be identified by their facial features. That's what I think. If we look at it, yes we would feel that way (feel sensitive about it)...' (Group 5 – patient)

'If while walking with our siblings, people will make remarks ...this is whose child? People probably will not believe that we are related simply because we look different. This looks like the mother, that one like the father. But we do not look like anyone.' (Group 4 – patient)

Skin colour

'....they say that we are so black...' (Group 4 – patient)

'...His friends say that his skin is so black. That's why he feels embarrassed. I know he does.' (Group 3 – parent)]

Delayed menses

'Her friends have all attained menarche, even during primary school. Including those in Form 1, Form 2 and Form 3 of secondary school. Why have they had their menses but my daughter has not? Sometimes she questioned. Maybe she has discussed this with her friends. She asked why she has not gotten her period. Why not? She also asked me about this.' (Group 3 – parent)

III Employment

Being employed/Obtaining a job

'He cannot do heavy work. Because he has not the energy.' (Group 5 – parent)

'Not that we are lazy to find work. We have tried. Shops, supermarkets everywhere, but people will not take us in. What can we do?' (Group 4 – patient)

'They do not want to employ us as we sometimes have to take leave up to two or three days. What more if we have to be warded, sometimes up to two weeks...that's why people would not want to employ us for long..' (Group 4 – patient)

Relationship with employer

'Our employers cannot accept a long leave application. Some may understand our situation but some may not and there is nothing we could do.' (Group 4 – patient)

'...there's no establishment that would understand our situation. It's not our intention to simply miss work to go to the hospital twice a month. But we don't have a choice. Who's to assist except for the father..? Sacrifices have to be made. Twice a month is absolutely necessary...how much annual leave do we have? 30 days? Definitely not.' (Group 2 – parent)

'My boss does not understand about this. But I don't think he does not really understand. He just doesn't want to know.' (Group 1 – parent)

'Yes, private. When we work for a private organization, the risks are higher than when working with the government. When we have to send our child for treatment, we have to apply for leave...but they sometimes think that we simply want to be absent from work. They can reject us, sack us anytime. They would find ways or reasons so that we need not be compensated. So, I am always in a difficult situation. With a child that is sick. Only those in the same situation would understand.' (Group 2 – parent)

'Yes, more or less. They are more comfortable working on their own. It is easier as there is no need to apply for leave (or medical leave).'

(Group 2 – parent)

IV Marriage and starting a family

Future spouse

'I want her to know what I am going through, I want her to know what she should expect in the future, our children...' (Group 5 – patient)

'Not that we don't want to find a companion, but we are just afraid that we would not be accepted.' (Group 4 – patient)

'...I am thinking of my 25-year-old daughter, thinking about her prospects of finding a life companion with her condition. Also whether she would be able to find a job.' (Group 4 – parent)

Offspring

'...but my daughter is now 24 years old, I advised her to get married. She said that she does not want to do so. She's afraid, she said. She's afraid that her children will be just like her. I told her that it is all Allah's will. Yes, it's true. But then again, it is all up to her.' (Group 2 – parent)

Effect of screening results

'If the situation a long time ago is like today, I would still get married but I would practise family planning...' (Group 1 – parent)

'As for me, I would still go ahead with my wedding plans. Because I love her.' (Group 1 – parent)

'I felt a bit dismayed. If only I could have known earlier. If I had known, perhaps I could have avoided this situation....find somebody else.' (Group 1 – parent)

'...we are responsible for our child's condition, tears are always flowing...' (Group 2 – parent)]

'The relevant authorities should have informed us earlier...' (Group 1 – parent)

'There was no exposure at that time. Awareness was limited.' (Group 1 – parent)

V Financial issues

Financial burden

'A cab would cost me RM 15, and another RM 15 to return. I did not have a single cent of money in my pocket. But because my child had an appointment I had to take her to hospital. When she was on transfusion she told me she was hungry. I cried and was feeling very sad as I had no money in my pocket. All because of the follow up visit appointment. I was not ashamed to admit by Allah I actually cried.' (Group 2 – parent)

'I work nights, so there is not much problem. Still, finances are not good. Since I am paid on a daily basis, one day of absence will mean that I will not be paid for that day.' (Group 1 – parent)

'...if we need the transfusion before the clinic appointment date then we have to be admitted to the ward. Then we had to pay for the hospital stay.' (Group 4 – patient)

Financial independence

'If possible, we do not want to use our parents' money. We want to have our own money when we are older. We want to work and not depend on our parents anymore to buy this and that. Should we ask our mother to buy us our clothes still?' (Group 4 – patient)

VI Relationship and social interactions

Family members/siblings

'Sometimes they make a passing remark...they say that we are like vampires sucking blood every month.' (Group 4 – patient)

'When we are in a gathering with friends or cousins, when we ask them for their phone numbers, they would ask why they should give their numbers to little children. That's what they said.' (Group 4 – patient)

'Sometimes it feels like we are loved less. If our younger sister passes an exam, she gets a bicycle. Mother has also mentioned that I will never achieve anything even if I studied.' (Group 4 – patient)

Peers

'I see that in the afternoons, he is seldom out with his friends. He prefers to watch television (Astro channel) at home. He spends most of his time at home. He does not go out much. If he goes to the shops, he will come directly home after.' (Group 2 – parent)

'Hmm, she said a lot do not want to be friends with her. Her own friends do not come to the house.' (Group 4 – parent)

'I don't let her be involved in sports. She does not mix with friends much, I suppose that's why she keeps to herself. Not that she does not make friends at all but when she goes out, she will want to come home early. Spends her time at home.' (Group 2 – parent)

'It feels intimidating to make friends or to sit with those who are older than you.' (Group 4 – parent)

VII Communication

'I am the same. We talked about it more or less. But then sometimes he doesn't want to talk about it.' (Group 5 – parent)

'I don't want to talk about this with my mother or my father (while shaking his head side to side) because I simply don't want to. I don't know what to say, I feel it's useless to talk about it. It will not change things. I still have to live this way.' (Group 5 – patient)

'It's better if other people ask first rather than we tell them....at least, when they started asking and we tell them we would feel a bit relief...'. (Group 5 – parent)

'I think we need advice or perhaps professional advice. Err, and motivation from experts. We need counseling sessions by an expert. But we as parents he is so used to us and up to one extent, we would be angry because he would not listen to us.' (Group 5 – parent)

VIII Self-esteem

'A variety of emotions...sadness, anger...but what can we do, we have to accept our fate.' (Group 4 – patient)

'I feel inferior to others. As if I am not complete. But then, if we feel that way, that would not take us anywhere would it?' (Group 4 – patient)

'Personality-wise, he seems a bit arrogant. Maybe because he does not want people to know that he is sick. He wants.....to the extent he does not want to take his medication. He says he's fine.' (Group 5 – parent)]

Employment issue was not restricted to only difficulty in getting a job due to illness and marginal academic records. It was also about employer-employee relationship involving both patients who were working and parents of thalassaemic children. Some employers were not supportive of their employees (parents of young children) who had to take time off work to accompany their children to hospital for treatment. This problem was more prominent in the private sectors. We believe that since thalassaemia is a chronic disease these workers need their employers' full support in order to cope with what is already a difficult situation. Unemployment was raised as one of the psychosocial burden in several studies although it was not discussed in great details^{10,13}.

Financial burden seems to be a universal problem when discussing about management of chronic disease especially those of childhood and are hereditary in nature. This means more than one child in a family could be affected. Chronic diseases require prolonged follow-up visits, treatment and frequent admissions. As such it is a major problem among the lower socio-economics group¹¹. The burden is probably less if the parents were government servants as has been clearly observed in our study. We suggest that a mechanism be put in place where parents of thalassaemic patients be exempted from paying hospital bills. A suggestion was also made during the focus groups to designate thalassaemic patients under the "Orang Kurang Upaya" ('Disabled') group so that they are entitled to some form of financial and other assistance.

Physical deformities due to thalassaemia appeared to cause a certain degree of stigmatisation leading to low self-esteem and the feeling of insufficiency. It is worsened by the fact that retarded sexual development may hinder the possibility of marriage and parenthood¹⁶. Poor self-esteem and negative self-image have been documented by several other investigators. They highlighted the fear of the patients related to being rejected and ridiculed by others because of their physical appearance and feelings of insufficiency^{11,17,18}. Authors had suggested that medical treatment of thalassaemic patients should be accompanied by psychological support to cope with this long-term chronic illness. This is indeed crucial.

In our study the relationship of the patients with their siblings was not generally adversely affected although they felt different from the siblings as also documented by others^{10,11}.

In the Thai children it has been shown that there was no difference documented in terms of psychosocial problems among siblings of thalassaemics as compared to children without chronic illness¹⁹. On the contrary in Greece, siblings of thalassaemia patients in comparison with the control group were found to have significantly higher psychopathology after the age of 1020. We however did not include the siblings in our study. It would be useful to look at this matter in greater detail.

There was however poor social integration and generally patients felt marginalized. Children with beta-thalassaemia were shown to have significantly higher rate of social dysfunction than their unaffected siblings. It was believed to be related to negative experience associated with their medical problems²¹. Mazzone and his group therefore suggested a psychosocial support be provided aimed at reducing emotional distress in these patients¹⁴. In our study we found that patients who were higher academic achiever and those from higher socio-economic backgrounds tended to integrate better socially. Family members however did not experience social marginalization or stigmatisation.

With regard to treatment of thalassaemia there was endless fear among parents in relation to adverse effects of transfusion. Patients however did not raise this point, rather they were more concerned with the sense of being tied down with the treatment. Educating parents on matters relating to adverse effects of transfusion would indeed be helpful in relieving their anxiety. The routine process of obtaining informed consent prior to transfusion perhaps is not a sufficient platform to educate the parents.

Our study also highlighted one important misconception about transfusion. Parents appeared to believe that following transfusion there were changes in the patients' behaviour. They perceived this to be related to the personality of donors of the units of blood transfused. Thus far this issue has not been previously raised. More study is needed about this perception.

The compliance to subcutaneous desferrioxamine was obviously poor in most of the patients for various reasons. This finding concurred with the findings of others^{22,23}. A cross sectional study conducted in Kuala Lumpur on 112 transfusion dependent thalassaemias indicated that 48% of the patients were on sub-optimum desferrioxamine and that these patients had lower quality-adjusted life-years than those who were optimally chelated⁸. Although there is evidence

that adherence is related to regimen factors, there has been less emphasis on the relationship with psychosocial factors and adherence²⁴. The correlation needs further study in order to improve compliance. For as long as the compliance is poor, the long-term survival can never be improved since inadequate compliance results in substantial morbidity and mortality as well as increased costs^{22, 25}.

Different approaches were looked into and used to improve the compliance, one of which is the Cognitive-Behavioural Family Therapy (CBFT). Mazzone et al. indicated that CBFT can be a valid tool to increase compliance with chelation therapy in thalassaemic children¹⁴.

In this study the reasons given for the non-compliance apart from the local side-effects, were mainly related to the feelings of patients and family members about treatment. We therefore believe that we can approach this problem through cognitive-behavioural therapy in addition to creating a team of health personnel that could provide long-term psychosocial support. Switching totally to oral forms of iron chelator perhaps is more cost effective in the long run. Also since the strong faith in Islam was the main coping strategy of the parents and the patients in this study it could perhaps be the basis in the development of interventions related to enhancement of compliance and psychosocial support.

CONCLUSION

It is evident from our study that thalassaemia affects immensely the psychosocial aspects of the patients and family members. It is possible that not enough is being done for them in terms of the long term psychosocial support. Psychosocial support is crucial as it helps reduce emotional distress and strengthen coping strategy. These will then lead to better integration of treatment in daily life and hence a better quality of life for patients and their families. A more structured strategy is needed, one of which is a multidisciplinary approach to the management of thalassaemia.

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