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## Pakistani Children's Experience of Growing Up With Beta-Thalassemia Major

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## **Abstract**

In this study, we explored the lived experience of children with  $\beta$ -thalassemia major ( $\beta$ -TM). We considered children as experts on their experiences in contrast to the prevalent approach of asking parents or other adults about children's perspective. The sample consisted of 12 children, aged 8 to 12 years. There were two stages to data collection. In stage one we employed two focus group discussions and two role-plays and analyzed the data thematically. This directly informed stage two, consisting of 12 in-depth interviews subjected to interpretative phenomenological analysis (IPA). From our findings we show that living with  $\beta$ -TM involves a continuous struggle between feelings of being different and strategies to minimize these differences to strive for normalcy. We suggest that understanding the experiences of living with  $\beta$ -TM from children's perspective can provide unique insights into their experiences which can fill the gap in the existing, predominantly adult-oriented, research on chronic illness.

## **Keywords**

Asia, South / Southeast; children, illness and disease; coping and adaptation; developing countries; interpretative phenomenological analysis (IPA); illness and disease, experiences; lived experience

Beta-thalassemias are a group of inherited hematological disorders caused by a mutation in the beta ( $\beta$ )-globin chain of hemoglobin.  $\beta$ -thalassemia major ( $\beta$ -TM) is the most severe type, which involves either a complete lack of  $\beta$ -globulin in the hemoglobin or abnormally low  $\beta$ -globulin production causing life-threatening anemia. It occurs worldwide but is more common in Mediterranean populations, the Middle East, Indian subcontinent, and Southeast Asia (Higgs, Thein, & Wood, 2001) and is usually diagnosed within the first year of life. If untreated or under-treated, it can lead to an enlarged spleen and/or liver, bone deformities, and spontaneous fractures. Managing  $\beta$ -TM requires patients to have lifelong access to regular blood transfusions, every two to four weeks. However, because  $\beta$ -TM is associated with increased iron absorption by the gastro-intestinal system, repeated transfusions cause iron overload, which can damage the heart, liver, and other organs (Higgs et al., 2001). Blood transfusions are therefore supported with daily use of chelating agents such as deferoxamine and deferiprone.

Existing psychosocial research in  $\beta$ -TM suggests that lifelong blood transfusions and chelation pose major demands on children and their families. Previous studies have reported a higher incidence of depressive and anxiety disorders and an impaired general functioning in  $\beta$ -TM patients (e.g., Sadowski et al., 2002). Clinical complications have also been found to impact negatively on children's self-concept (e.g., Yalçan et al., 2007). Similarly, other researchers have reported a low health-related quality of life (HRQOL) in children with  $\beta$ -TM, with schooling identified as one of the most affected aspects of living with  $\beta$ -TM (Gharaibeh & Gharaibeh, 2012). Nevertheless, other researchers have reported normal psychosocial and family functioning, not only in adolescents with  $\beta$ -TM and their families (Di Palma, Vullo, Zani & Facchini, 1998) but also in adults with  $\beta$ -TM (Zani & Prati, 2013).

In these conflicting previous studies, the researchers have mainly employed standardized measures of psychosocial functioning which might not be sufficiently sensitive to capture the meanings that children give to their experiences (Woodgate, 1998). Furthermore, most researchers have relied on parental reports of children's functioning, a trend prevalent in the literature of children with chronic illness. However, parents tend to underestimate their children's HRQOL, particularly their emotional functioning and psychosocial health as compared to children's self-reports (e.g., Caocci et al., 2012). This neglect of the child's perspective is in contrast to the increasing emphasis on patient-centered models of health care. Recognizing the importance of children's experiences has resulted in the emergence of a newer model of childhood which highlights children as autonomous social actors who actively construct and act on their own lives and those of others around them (Christensen & Prout, 2005).

Indeed, there has been a rise in the number of child-centered qualitative studies exploring children's perspectives of chronic illnesses (e.g., Elliott, Lach, & Smith, 2005). However, compared to illnesses such as cancer and epilepsy, there is a scarcity of child-centered research in relation to  $\beta$ -TM. Within the context of hemoglobinopathies, psychosocial research with children with sickle cell disease has taken precedence over  $\beta$ -TM. Given the potential impact of  $\beta$ -TM on children's lives and their lifelong involvement with health care services, there is a clear need for a better understanding of their experiences. Surprisingly, only a handful of qualitative studies have explored young people's accounts of living with  $\beta$ -TM, in the UK, India, and Thailand (Atkin & Ahmad, 2001; Roy & Chatterjee, 2007; Yunak, Chontawan, Sripichayakan, Klunklin, & Jordan, 2009). Common to these studies are adolescents' accounts of struggling to achieve normalcy and manage differences brought about by the condition.

According to estimates, 5,000-9,000 children are born in Pakistan with  $\beta$ -TM annually. Despite the increase in life expectancy to third, fourth, or even fifth decades elsewhere (Borgna-Pignatti & Bertelli, 2011), the average life expectancy of these children is only 10 to 12 years in Pakistan (Ansari & Shamsi, 2010). Given the adoption of treatment protocols for  $\beta$ -TM in Pakistan health care, other issues such as treatment availability are implicated in such high mortality rates. The high rate of consanguineous marriages and low literacy rates have been associated with increases in the number of children born with  $\beta$ -TM in Pakistan (Alwan & Modell, 1997). Despite this rather dismal picture, no studies to date have explored children's experiences of living with  $\beta$ -TM in Pakistan. An understanding of such experiences, including those unique to Pakistani culture and those shared with other children/young people with  $\beta$ -TM, is particularly important to inform the care of these children and augment quality of life.

Our present study is the first attempt to explore children's lived experience of  $\beta$ -TM in Pakistan using a child-centered approach. We aimed to provide a rich account of children's experiences of living with  $\beta$ -TM, its management and the meanings ascribed to these experiences.

## **Method**

### *Recruitment and Participants*

IPA guidelines, namely purposive sampling and a small sample size, guided the recruitment process. This enabled us to make a detailed and rich interpretation of the subjective experiences of each case (Smith, Flowers, & Larkin, 2009). We therefore aimed to recruit 12-16 participants with a medical diagnosis of  $\beta$ -TM, registered with a pioneer center providing free treatment, receiving blood transfusions and chelation therapy, and living within one of the major districts of South Punjab, Pakistan. The center's managers identified 14 families which fulfilled the

inclusion criteria. We provided an invitation letter and information sheet about the project to these families. Two families declined to participate; thus 12 children, six girls (three urban and three rural) and six boys (three urban and three rural), aged 8 to 12 years participated in the study. All children gave their assent while their parents signed consent forms to give permission for their child to participate in the study. Ethical approval was obtained from the University of Westminster Ethics Committee.

### *Data Collection and Analysis*

We conducted our data collection in Urdu, within the foundation building. We also performed our analysis in Urdu and later translated it into English for academic purposes. An independent, bilingual individual checked the English translation against the original Urdu version for accuracy.

In order to understand the lived experiences of particular phenomena, it is important to begin without predetermined notions about the experiences to be explored (Shaw, 2001). In addition, keeping in view the scarcity of child-centered psychosocial research in relation to  $\beta$ -TM and the predominance of quantitative research, we wished to begin the project with participants' concerns in their own words in contrast to using existing research. Another important issue, particularly when working with children, is the power differential between researcher and participants (Greene & Hill, 2005). To avoid a hierarchy of power from the outset, we deemed it important "to allow participants to generate the relevant questions and thereby guide the research process" (Bibace, Young, Herrenkohl, & Wiley, 1999, p.10). Thus, considering these issues, we employed an innovative research design with data collected in two phases. Phase one consisted of focus group discussions (FGDs) and role-plays to develop themes used as the basis for further exploration in phase two, which consisted of individual interviews.

*Phase one.* FGDs are recommended tools for generating ideas, and in the case of children they might help address the power differential between the adult researcher and child participants (Hennessy & Heary, 2005). We used FGDs and role-plays both to build rapport with children and to ensure a child-centered approach.

We conducted single-sex FGDs (each with six girls/boys) in which children were asked to talk about their lives and  $\beta$ -TM in general. We avoided a direct question about the impact of  $\beta$ -TM on their lives because we deemed it important to avoid assuming that  $\beta$ -TM dominated participants' experiences. FGDs lasted for 85-90 minutes. We also conducted two role-plays, one with five girls and one with four boys (one girl and two boys were unavailable to participate) which lasted for 20-30 minutes. The scenario consisted of a mother/father and child's visit to the center and their discussion with the medical staff about the child's illness. The role-plays' setting allowed children to garner their concerns in relation to  $\beta$ -TM in their own words without interference from the researchers, thus helping us to bring out the main issues from the children's perspective. Furthermore, the role-plays helped to confirm and add to the insights obtained from the FGDs.

We subjected the data from the FGDs and role-play to semantic thematic analysis following the guidelines proposed by Braun and Clarke (2006). On the basis of this analysis, we identified three areas for further exploration in phase two: impact of thalassemia ( $\beta$ -TM); coping efforts; and relationship with health care professionals. The detailed steps and findings of the thematic analysis are reported elsewhere (Mufti, 2011).

*Phase two.* The first author conducted semi-structured interviews with all 12 children which lasted 45-65 minutes. We analyzed the interview data using interpretative phenomenological analysis (IPA). We felt this phenomenological approach was particularly



relevant to the current study, given the aims of IPA in helping us understand the lived experience of individuals and grasp how they make sense of their experiences based on their personal and social worlds (Smith et al., 2009).

In phase two, our individual interviews allowed children to discuss the issues identified in phase one in greater depth in accordance with their personal circumstances, generating rich data for IPA analysis. However, children were free to talk about new issues during their individual interviews. As per Smith et al.'s (2009) recommendations, we minimized the number of questions to allow participants to talk about their experiences and to ensure that their responses were not dictated by the interview schedule. We then invited children to make drawings of themselves in order to get an insight into their experiences and to give them an active occupation. We used this exercise to shift power to the children because they were the interpreters of their own drawings and described them to the first author. Children's detailed descriptions of their drawings were included as part of their interview transcripts.

We transcribed and analyzed the interview data (including the descriptions of the drawings) in accordance with IPA guidelines (Smith et al., 2009). The analytic steps were cyclical and initiated by the reading and re-reading of the first transcript, followed by noting down the summary descriptions and initial observations and interpretations in the right-hand margin. We then re-read the transcript and noted emerging themes in the left-hand column. We examined these themes for connections and then grouped them together to form clusters of themes. We reviewed these clusters again and combined some of the themes to produce subordinate themes. We combined these subordinate themes to create superordinate themes for each case.

We repeated the same process with every transcript; however, the list of themes from the first interview informed the analysis of subsequent interviews. Any new themes found in subsequent interviews were then compared with previous transcripts. After the analysis of individual interviews, we compared each case analysis with each other to produce a collective master list of themes. A theme was considered recurrent when it was reported by more than half the participants.

We took several steps to maximize the credibility and trustworthiness of the research. The combination of FGDs, role-plays, and interviews served as triangulation of data collection methods. The first author conducted the primary analysis in close collaboration with the third author, an experienced qualitative researcher, who independently analyzed two of the transcripts (analyst triangulation). We discussed interpretations and hierarchical structuring of themes at several stages of the analytic process to ensure trustworthiness. Our use of excerpts throughout the results supports the presentation of themes to facilitate transparency.

Our first author is a Pakistani woman with prior experience of working with children in a research context. This insider status might have facilitated rapport and sensitivity toward participants' perspectives and sociocultural context. Conversely it might have led to cultural assumptions, although this was mediated by analyst triangulation.

## **Findings**

Our analysis resulted in three superordinate themes (see Table 1) which reflect the burden and stigma of living with thalassemia (“being different”: loss of normal childhood), coping strategies to normalize the self (“there is nothing wrong with us”: minimizing differences), and the fragile process of redefining self (“we are half normal”: self-reformulation).

INSERT TABLE 1 HERE

Children continually vacillated between feelings of being different and self-reformulation, based on their vulnerable efforts to minimize differences (see Figure 1).

INSERT FIGURE 1 HERE

### **“Being Different”: Loss of Normal Childhood**

The core narrative running through this theme is the range of challenges faced by children, incorporating physical, emotional, cognitive, and social difficulties. Children reported feelings of loss while making upward social comparisons with idealized images of the normal lives of healthy children.

#### *“It Hurts!” Aches and Pains*

Children encountered pain as a direct result of  $\beta$ -TM and its treatment, and indirectly in the form of a stigmatized identity. Treatment (transfusion, and particularly chelation) was paradoxically both agonizing and empowering. The painful nature of treatment sometimes had a negative impact on adherence for all children, yet was also considered essential for survival.

Boy 1(B1): During every visit to the center for transfusion I have to have needles stuck in my skin twice or thrice. At home, I have to have at least one needle for injection. Most of the time nurse cannot insert the needle properly on the first attempt so she sticks it again and again. It hurts and it's annoying! My life has a lot of needles [pause] and my life depends on them.

To resolve this tension, all children at some point during the course of their illness unsuccessfully used alternative treatments such as *hikmat* (a form of traditional medicine, based on Hippocratic Humoral theory, commonly practiced in Pakistan), homeopathy, and spiritual healing. These unsuccessful attempts led children to reframe their thinking about the importance

of medical treatment. However, for most children pain remained one of the key differentiating features from their peers.

Girl 1(G1): They [healthy children] do not have to get these injections or transfusions; they do not need it because they are normal [healthy]. It is us who need transfusion and who have to have needle sticks in their bodies. They are always happy and free while we have to tolerate this pain.

Rural children had more difficulties in treatment adherence relative to urban children, mainly due to greater expense, time, and energy consumed by travelling. Rural girls faced additional difficulties due to needing a male companion when visiting the center.

Transfusions required timely availability of blood at the center; however, children frequently raised the problem of blood shortages. This resulted in deterioration in health and also necessitated additional physical, emotional, and financial resources, increasing the burden of treatment.

B2: I want my blood on time but sometimes we have to wait really long for my blood. It is very difficult to come every day to see if my blood is available! It is frustrating and annoying! I want my blood on time.

Although healthy children rarely think about death, these children discussed regularly facing their fears about death. Development of characteristic thalassemic features (protruding teeth and enlarged abdomen) reinforced such fears and was also associated with anxieties relating to the visibility of their illness.

G2: When your belly gets big then people can see it; they can see that there is something wrong [nervous]. I do not want to get one [big belly], it will make things worse. People will make fun of me, they can see it.

Witnessing a peer developing such symptoms or receiving news about their death further reinforced their own fears. This highlighted their powerlessness, yet it also acted as a strong motivator to adhere to treatment, particularly chelation.

G3: I used to think that if I follow my treatment regularly I will be fine but when my friend died I have now realized that it does not matter how hard I try [pause]. I can die just like my friend, no warnings!

G4: I hate my injections; I remember I used to refuse them but then I heard about death of a few children who also refused to have their injections and transfusions. I am now regular in my transfusions and chelation, although I don't like these.

#### *"It's Incarcerating": Restrictions and Isolation*

This subordinate theme underscores the constraints of living a cautious life of limitations and discrimination which negatively impacted on children's sense of self. This resulted in perceptions of an incarcerated and isolated childhood, in contrast to children's view of normal childhood.

G3: I do not have any friends. I cannot go out of home; often people make fun of me and no one comes to my home. I see other children they have so many friends, having fun together [umm] playing, laughing [pause], I am alone. I cannot go out even if I want to!

Living with  $\beta$ -TM required children to live more cautiously. Children's foremost concern was to maximize and conserve hemoglobin and energy levels, because they also acknowledged its importance in combatting physical limitations. Children perceived blood as precious and recognized the need to be disciplined and follow precautions to preserve it. This included adhering to the treatment regimen and avoiding strenuous activities. These precautions, however, confined children to their homes and reduced their social network considerably.

B3: Sometimes I really want to go out when I see my brothers playing outside but I know I should not, I cannot. I have to preserve my hemoglobin; I cannot afford to lose blood in case I get injured. I cannot buy blood, I have to protect it.

Thus, a cautious lifestyle provided a sense of control; however, it also led to isolation and frustration. One of the boys (B1) described this tension as "walking a rope".

They [healthy children] do not care about it; they do not have to be cautious all the time. I feel like walking a rope because I have to be cautious all the time [pause]. Do not do this, this will happen, [umm] do not do that you will get sick, it's annoying!

The social impact of  $\beta$ -TM was a major contributor to feelings of isolation. Indeed it was described as more debilitating than the disease itself and children found it particularly distressing to see their illness identity taking precedence over other aspects of self, primarily because of the profound negative social consequences. Humiliation and discrimination were graphically described by all children and included their extended family networks and wider community.

G5: I had a friend in my neighborhood but her mother did not let her play or eat with me. She thought that her daughter will also become ill like me; but how is it possible? [Pause] Is it possible? No, it's not, because my siblings live with me and they are just fine.

The label of "sick child" was consequently strongly rejected and children exhibited a strong desire to confront bullies aggressively, either verbally or physically. However, this desire was in conflict with their body image as fragile and frail, increasing their vulnerability in social settings. This conflict gave rise to helplessness and repressed aggression.

B1: I do not like it when people tease me but sometimes it happens. It makes me angry but usually I let it go. I do not like to confront them; I do not want to lose my self-control. Sometimes the other person is well built and can beat me very easily. I keep my control but [excited] when I lose control I lose it! I can at least give one blow to his face! I do not want to be known as a sick child [pause]. It does not make any difference but I feel [pause] insulted.

In addition to the lack of energy and frequent visits to the center for transfusion, bullying by peers also significantly contributed toward a high drop-out rate from schools. The support of teachers placed children under the limelight and augmented problems for them. Although a few children reported attending school, most rejected formal study and others chose other sources of education, such as home tuition.

B3: My class fellows used to bully me a lot. I always returned home with a bloody nose, they called me names [mmm] like blood child or sick child. They did not let me study! I

remember whenever I tried to finish my assignment during break time they used to snatch my notebook. I couldn't run after them . . . they run very fast . . . so I left school.

Such physical and social limitations led children to develop a negative body image. They described their body as weak, dependent, and faulty because it was unable to function normally. This interplay of physical, emotional, and social factors impacted on the self-concept of these children. Their devalued sense of self contrasted considerably with the perceived idealized self attributed to healthy children.

G3: I am sick, why would anyone be friends with me! I mean I cannot play with them the way they do. I do not play a lot with my cousins; I do not compete with them. I cannot, it is of no use. You see at the end I will lose eventually!

Discriminatory attitudes and behaviors magnified these negative feelings. For instance, in addition to parental overprotection, which reinforced feelings of vulnerability, discriminatory experiences outside their homes led to transient feelings of hopelessness. These often left children questioning their existence, resulting in frequent use of the term “children like us”, suggesting inferior status:

G3: Do you call this a living!! A never-ending humiliating experience! I prefer to die than to face this harsh world. People do not want us [pause] they do not want us. They do not want children like us in their homes, in their world [crying].

### **“There is Nothing Wrong With Us”: Minimizing Differences**



The core account of this superordinate theme is children's struggle to minimize the perceived differences between themselves and normal children using a variety of avoidance- and acceptance-based coping strategies, which led to redefining of self.

*Avoidance: Self-Preservation*

Avoidance often provided an effective defense against the adverse personal, emotional, and social consequences of illness and was primarily achieved through concealing and ignoring illness. Children demonstrated an aversion toward  $\beta$ -TM disclosure, particularly to strangers. The visible components of treatment such as the infusion pump were kept hidden from public view. The choice between disclosure and concealment provided children with a sense of control and largely depended on the type of relationship and potential consequences. Prior negative experiences and discriminatory experiences acted as a deterrent, whereas potential gains led to disclosure in a safe social setting.

G4: My teacher is very strict. I remember [smiling] she used to punish me and made me work hard. Then I told her about my condition, I told her that I have thalassemia, and now I do not have to face the strict behavior any more.

Children exhibited constant attempts to distract their attention from  $\beta$ -TM and related thoughts, to surmount the overwhelming helplessness of having an incurable illness. They strove to ignore their worries and focus instead on enjoyable activities and positive aspects of their lives.

B4: What will I get out of thinking about it [thalassemia]? I know that I will get nothing out of it, rather it will make it worse. So, I do not think about it [pause]. Will it go away if I think about it? No, of course not, then why should I think!

Another form of distraction was to reframe more positively their thinking about their condition. For instance, some children viewed death as a fundamental part of life, and thereby accepted its more overt presence in their own lives. Some children viewed aspects of  $\beta$ -TM as privileges not available to other children, such as extra love and attention by parents or teachers. Similarly, the majority of children viewed tolerating pain as a sign of their bravery which conferred superiority over healthy children. Others reported receiving preference over healthy children, as mentioned by one of the girls (G4): “My classmates get punished if they do not do their homework. I usually escape punishment partly and end up with just a little scolding [chuckled].”

Avoidance was, however, paradoxical, as it had potentially negative consequences in the form of repressed emotions. Similarly, concealment contributed toward isolation and fewer relationships, as efforts to conceal distanced them from close friends and increased feelings of vulnerability. Nevertheless, avoidance protected children from the negative consequences of thalassemia when other strategies failed, as described by one of the boys (B5): “When I feel that I cannot take it any more I watch my cartoons and everything just disappears. All I see is Tom chasing after Jerry . . .”

#### *Acknowledging Thalassemia: Sustaining Self*

Children considered it important to acknowledge the role of  $\beta$ -TM in their lives because it helped them to learn to live with it. One of the boys (B1) described it as follows: “Thalassemia is part of my life, a big part. . . . Half of my life is being spent with thalassemia so I have accepted it as it is.” However, this required constant effort, mainly consisting of understanding and integrating  $\beta$ -TM in their lives.

Their process of acceptance came from finding meaning in their suffering and making external attributions, primarily using medical and religious frameworks. This helped them to talk about  $\beta$ -TM objectively without guilt, thus making it easier to incorporate it into their lives. Children gathered information from educational leaflets as well as personal experience to create a bricolage.

B2: It damages my liver and spleen. It has a larger effect on liver. My liver cannot work properly so my spleen gets affected too and it starts drinking blood. This is why I lack blood: my spleen drinks it all.

Their belief in an omniscient Allah emerged as the primary defense against blaming self or others, helping children to put their suffering into perspective.

B4: Allah knows everything, everything depends on his will. If he wants it then my body will start producing its blood; otherwise it will not. He gives health and illness. He has a plan for everyone, so if thalassemia is written in my destiny then it's okay.

At times, children questioned "why me?" but their disenchantment was short-lived. The concept of an eternal happy afterlife led children, particularly girls, to embrace  $\beta$ -TM, viewing it as a test of their patience and devotion and also compelling them to adhere to treatment. One of the girls (G3) described it as follows: "Suicide is prohibited in Islam . . . if I do not get my treatment it is another type of suicide and Allah will not be happy with me."

Children attempted to view their treatment regimen as a routine process by incorporating it into daily life. Making it a routine helped them raise their threshold for bearing the burden. Children learned ways to work around their limitations, for example focusing on their

capabilities and replacing physical activities with less intense ones to overcome fatigue and weakness.

B2: I can play like other children, so why cannot I play? They play, then why cannot we play [angrily]! I do not play games which require running or fighting. I take care, [pause] I play all the games which I can while sitting.

### *Support Systems*

This subordinate theme illustrates the importance of external resources, such as health professionals, family, and friends. Interactions with health care professionals could either empower or frustrate children; professionals were seen as experts and acceptance of their advice was considered necessary. However, interactions (particularly with scolding nurses) were not always perceived favorably, impacting on children's hospital experiences as well as treatment adherence.

B1: See, if I am welcome here and doctor talks to me nicely, I mean, if they are nice and friendly then I will think about it at home and I will be happy. And if I am happy it will enhance my health, you see. I will come to the center happily and you know that happiness is very important for health.

Parents emerged as providing unconditional support for children's personal, emotional, and social concerns. This made them feel accepted and loved regardless of their condition. Children described parents as an immense support during difficult times, as explained by one of the girls (G6): "My mother is my sanctuary." Parents provided children with renewed vigor and motivation to follow treatment.

G2: My parents are taking so much pain to get me my treatment [pause]. I try my best to follow it regularly, but sometimes I do not want to [pause]. Then I think of my parents and I think that I should get my treatment and I do it.

Children, however, also acknowledged negative outcomes of parental support because it also gave rise to overprotection and overindulgence, which further reinforced children's sense of being different. Children both enjoyed and felt frustrated by such special treatment. Although they acknowledged actively manipulating their parents to maximize short-term benefits as compensation for their suffering, this could also be restrictive:

B3: I see my brothers and sisters playing around me, cycling, running. I play with them but then after some time my mom stops me [pause]. She does not stop my brothers and sisters, she does not stop them but she just stops me [pause]. It makes me angry sometimes.

### **“We Are Half Normal”: Self-Reformulation**

Children attempted to reformulate their wounded self by comparing their illness-self with other selves, thereby enhancing their self-concept and placing them in a relatively favorable position within the external world.

The fortnightly cyclical nature of transfusions provided children with an opportunity to reconstruct their sense of self according to treatment phase. The pre-transfusion self was associated with a stigmatized identity (being faulty), whereas the post-transfusion self was described enthusiastically because it was associated with completeness and normality. It led

some children to use the expression “half normal” to describe themselves, whereby half indicated the transitory feelings of normalcy immediately following transfusion.

B1: You can call us half normal. We are half normal because we become normal after transfusion for a few days [pause]. Then I get weaker and weaker till my transfusion date and my life moves on like this.

Similarly, one of the boys (B2) described himself as “a little normal” in relation to his abilities; the ability to do “everything but to a lesser extent” allowed him to define himself in a positive way.

Contextual connotations associated with the center and the home also affected children’s self-perception. The environment of the center and transfusion room exacerbated feelings of being ill and vulnerable, as described by one of the boys (B6): “When I visit the center I feel ill. The transfusion room is there to remind me of my lack of blood; at home I do not feel ill.” In contrast, at home children described feeling relatively normal and safe, which provided a sense of wellbeing. The ill self was replaced with the identity of son or daughter, sister or brother. Strong parental support and protection played an important role in the creation of a relatively normal home environment which was instrumental in enhancing self-esteem and feelings of health.

G2: I feel safe at home; no one is here to make fun of me, to tease me [pause]. When I am at home I sometimes even forget that I am sick and I have thalassemia. I know that I am safe at home, away from people’s harsh comments.

Although comparisons with healthy peers highlighted differences, other sick children were perceived as having the same caliber, which provided protection from feelings of inferiority. They were regarded as more understanding, supportive, and accepting than their healthy friends because they shared similar experiences. The realization that they were not alone lessened the feelings of burden, at least to some extent.

B2: I am better than many children. I see children, they are deaf or they do not have an arm. I see them every day; even today I saw a child who was begging on the street and he had no legs. I thank Allah that I am in a better position.

Some girls and boys used downward social comparison in relation to each other's lives. Both considered that the other gender faced harsher ramifications, which made their own condition relatively less burdensome.

G1: I have a cousin who also has thalassemia, poor guy! He cannot go outside often. I feel bad for him, I mean I usually stay at home so it does not make a big difference in my life; but he is a boy and he is locked off.

## **Discussion**

Using a phenomenological and a child-centered approach, we addressed the gap in the existing literature by providing a unique and detailed insight into the experiences of Pakistani children living with  $\beta$ -TM. We identified three core themes: the burden and stigma of living with  $\beta$ -TM (loss of normal childhood); mechanisms used to protect the self (minimizing differences); and the fragile process of self-reformulation. These findings highlighted the dynamic nature of children's experiences as they vacillate between feelings of being overwhelmed by their

condition and self-reformulation using a variety of coping mechanisms. We also elucidated the contribution of personal and contextual factors, such as deterioration of health, discrimination, and experiences of medical care, which can potentially disrupt coping efforts and influence the dynamic adaptational process.

Consistent with previous studies, we confirmed that living with thalassemia was a painful and isolating experience (Roy & Chatterjee, 2007; Yunak et al., 2009), which was managed by employing a range of effective yet vulnerable coping strategies (Atkin & Ahmad, 2001; Yunak et al., 2009). In the current study we shed light on several paradoxes in children's experiences. The lifelong and invasive treatment was the most painful and disruptive aspect of  $\beta$ -TM, yet was highly valued for survival and re-establishing normalcy. Similarly, children adopted a cautious lifestyle to manage  $\beta$ -TM; however, this exacerbated their isolation and frustration (Atkin & Ahmad, 2000; Yunak et al., 2009). Additionally, characteristic thalassemic features and deformities were viewed as intensifying differences (Yunak et al., 2009) and contributing toward marginalization. The identity of being a sick child was both loathed and desired; children rejected this label because of its negative connotations but also desired special treatment as compensation for their suffering.

Children's experiences of  $\beta$ -TM shared some similarities with those of other chronic illnesses such as epilepsy (Elliot et al., 2005) and asthma (Protudjer, Kozyrskyj, Becker, & Marchessault, 2009), particularly the difficulties associated with managing difference. This provides some support for the non-categorical approach which suggests that experience across chronic illnesses shares key features (Stein & Jessop, 1982). However, children's narratives also illustrated  $\beta$ -TM-specific issues, such as a shifting sense of self in relation to transfusion phase and location; and problems and stigma associated with blood arrangement, transfusion, and



chelation, which highlight the need to explore the experience of living with  $\beta$ -TM in its own right.

Through our findings we support the negative impact of  $\beta$ -TM on social relationships, including the widespread stigma and discrimination in South Asian communities (Atkin & Ahmad, 2001); however, we found an alarming level of discrimination and bullying in our research which might be indicative of wider societal attitudes toward chronic illness in Pakistan. Teachers react negatively to pupils' frequent absence for transfusions and their lack of energy (Gharaibeh & Gharaibeh, 2012). However, in the present study we additionally identified the contribution of peer bullying and discrimination to the high drop-out rate and poor educational prospects for these children. Children's recurring contact with bullying also contributed toward the gradual internalization of negative self-concept and social withdrawal. Given the importance of peer acceptance in psychosocial adjustment, school functioning, and self-formation, the findings are of concern and should be given greater consideration both in clinical practice and in future research.

A novel finding of our study was the way in which children's views of their bodies and identity varied according to transfusion phase, location (home vs. center), and social context. We suggest that using child-friendly methods, in particular role-plays and drawings, might have enabled children to reflect on their bodies and changing identity more fully than conventional interviews alone. Given the universal status of transfusion for hemoglobinopathies, future research can build on this finding by exploring its applicability and relevance to other hemoglobinopathies.

Religion has been identified as an important coping resource, particularly among Muslim participants (Atkin & Ahmad, 2000). In the present study, considering Allah as the ultimate

authority on health and illness reflected an external locus of control. In contrast to previous studies which have reported a negative impact of external locus of control on self-concept (e.g., Burkhart & Rayens, 2005), in this context religion helped children to put their helplessness in perspective and encouraged active coping (e.g., treatment adherence).

Supporting Telford, Kralik, & Koch's (2006) argument, we highlight the pitfalls of using the terms "acceptance" and "avoidance" in the way that is traditionally done in texts on coping strategies. We question the label of "avoidance-based coping" as inevitably maladaptive, because in our study it protected children from becoming overwhelmed by their worries and sense of helplessness. Similarly, although acceptance is generally adaptive, complete acceptance might lead to hopelessness and passivity and thereby disrupt the coping process.

Children's coping efforts led them to reformulate their identity to incorporate their illness into their lives. People with an acquired chronic illness typically make negative comparisons with their pre-illness selves, gradually developing a new identity compatible with their illness (Charmaz, 1983). However, the children in our study did not have a former normal self for such comparison, because diagnosis is typically within the first year of life. Negative self-image arose among these children when their faulty bodies clashed with culturally assigned ideal bodies. However, they acknowledged the negative impact of upward comparison, and in their pursuit of a positive self-image they resorted to comparisons pre- and post-transfusion, at home and at the center, and with other chronically ill children. This comparison with other chronically ill children gave rise to the thought that things could be worse, signaling the process of self-redefining (Morse & Carter, 1996).

An interesting finding was the process of normalization employed by children to manage the impact of  $\beta$ -TM. To address the limited studies exploring the process of normalization in

children, Elliot et al. (2005) reason that the concept of normalization has originated mainly from studies with chronically ill adults and the families of chronically ill children. However, in line with studies of children and adolescents with thalassemia (Atkin & Ahmad, 2001; Yunak et al., 2009), asthma (Protudjer et al., 2009), and juvenile idiopathic arthritis (Guell, 2007), we suggest through our findings a successful use of normalization by chronically ill children.

We found that despite using a number of coping strategies these children remained vulnerable, not only because of changes in health but also as a result of discrimination and lack of social support. These impacts made children constantly shift between feelings of being different and a reformulated self. Our findings are similar to those of Atkin and Ahmad (2001, p.624), who described adolescents' efforts to live normally as living in "constant tension"; they highlighted fluctuations in illness status, personal and social circumstances as the reasons underlying vulnerable coping.

The vacillating nature of children's experiences challenges the unidirectional view of adjustment proposed by models of childhood chronic illness (e.g., Thompson & Gustafson, 1996). Despite recognizing regressive steps in the course of adaptation, adjustment is implied to be the end goal in these models (Paterson, 2001). IPA supports the use of models and narratives to illuminate the developing elucidations and accounts from the data, which "can be used by other researchers to help to illuminate the phenomenon to which they apply, but they should be continuously checked out against incoming data and evolved where necessary" (Fade, 2004, p. 650). We recommend the development of a pediatric chronic illness model which can be applied to these dynamic circular experiences. In the adult literature, Paterson's (2001) shifting perspective model explains the experience of living with chronic illness as a process of

constantly shifting between two phases: illness in the foreground and wellbeing in the foreground. We suggest that children undergo similar processes in managing long-term illness.

Given the considerable challenges faced by these children, we call for interventions to help support their own coping efforts and reduce the impact of stigma and discrimination. Leaflets consisting of children's own experiences can be an accessible method to increase understanding of  $\beta$ -TM and its self-management (Grime & Pollock, 2004). Information about treatment phase-specific issues (e.g., increased vulnerability pre-transfusion) and intervening factors (e.g., social difficulties) can help parents and professionals provide appropriate support in both home and medical settings. We emphasize the need to develop educational programs for health care professionals to provide support to children and their families and increase awareness of the importance of treatment adherence in managing thalassemia.

Given the extensive discrimination in the daily lives of these children, social skills programs which include assertiveness training and problem-solving could be valuable in mitigating the negative social consequences of  $\beta$ -TM. In particular, disruption to their education requires stronger connections between thalassemia centers and schools in Pakistan to ensure both that teachers and other children have sufficient information about  $\beta$ -TM and that children with  $\beta$ -TM do not feel isolated and stigmatized.

In this study, we employed an innovative multi-method design to ensure the research was child-centered by actively engaging children in the research process, thereby retaining a phenomenological perspective. The sample, however, was small and we conducted the study at a single charity center primarily visited by economically disadvantaged families from Punjabi/Sariaki ethnic background. Therefore, further research is warranted with children from

different socioeconomic, ethnic, and cultural groups, as these contextual factors might impact on experiences of discrimination and the impact of illness.

In conclusion, in this study we highlight the multifaceted experiences of children living with  $\beta$ -TM in Pakistan. We shed light on the importance of understanding the process of self-reformulation as well as factors such as discrimination and lack of support which undermine the coping strategies of children.

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Table I Master List of Themes

<i>Superordinate themes</i>	<i>Subordinate themes</i>
"Being Different": Loss of Normal Childhood	"It Hurts!" Aches and Pains "It's Incarcerating": Restrictions and Isolation
"There is Nothing Wrong With Us": Minimizing Differences	Avoidance: Self-Preservation Acknowledging Thalassemia: Sustaining Self Support Systems
"We Are Half Normal": Self-Reformulation	