THALASSEMIA AND THE ADOLESCENT:
An Investigation of Chronic Illness, Individuals, & Systems.

Eugenia T. Georganda, Psy.D .

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> ... Serious physical illness may contribute to mastery of life. Thus, inner processes can erase or magnify the effects of external illness. Health and the "ego" must be considered together.
>
> George Vaillant, 1977, p. 14

Abstract:

In this phenomenological study of the impact of thalassemia, a serious hereditary blood disease, on the life of the affected teenager and his/her parent's five families participated. They were interviewed separately and for a maximum of one hour each. They shared their views regarding the impact of the illness on their everyday lives; on their relationship with each other; their relationship with their friends; their perception of themselves; and their perception of the future. Ways they described using in order to cope with, and adapt to, the illness were identified.

Introduction:

A review of the literature suggests that the presence of a chronic illness intensifies the difficulties of adolescence for both the adolescent and his/her parents. The diagnosis of a chronic illness, especially hereditary, creates for the parents feelings of guilt, shame and self-blame. In addition, families who have a member with a chronic illness invariably go through a phase of grieving of the loss of the healthy child they were hoping for (2, 3, 5, 10, 14).

For the adolescent a chronic illness represents being different and inferior with a consequent loss of self-esteem and increased ambivalence with respect to dependence and independence (11). Also it leads to an alteration of self concept with

unsatisfactory body-image, fear of rejection, problems in peer relationships, fear of independence and doubts about self-sufficiency (1, 4, 6, 8, 9).

This study focused on thalassemia, a chronic hereditary blood disease that is characterized by early onset of anemia and life-long dependency on blood transfusions. The basic defect in thalassemia, which is also known as Cooley's Anemia and/or Mediterranean anemia, is a decrease or absence of the synthesis of one or more of the globin chains of hemoglobin made by the body. Thalassemia may be inherited if both parents carry the gene for thalassemia. If each parent has the thalassemia trait, there is a one-in-four chances with each pregnancy that the child will have thalassemia. If only one parent has thalassemia trait, each child born will have a one-in-two chance of having the trait.

The earliest signs of the disease include paleness, weakness, irritability, and failure to thrive. Fever, feeding problems, diarrhea, and other gastrointestinal symptoms may also be present. An infant with thalassemia appears healthy at birth. Signs of the disease first develop a few months after birth and become progressively more severe. Early identification and treatment, important in treating thalassemia, now include prenatal diagnosis of the disorder. Although the test for the identification of the thalassemia trait is relatively simple and easy parents from high risk groups, primarily Greek and Italian immigrants, are resistant to being tested.

Thalassemia patients face complicated clinical problems. If inadequately treated thalassemia patients develop a characteristic appearance, with prominent cheekbones, slanting eyes, overgrowth of the upper jaw, jumbled upper teeth, and an overbite due to expansion of the bone marrow cavities of the skull and face.

As red blood cells break down, their components become waste products and are eliminated through a process occurring mainly in the spleen and liver. The accelerated breakdown of red blood cells in thalassemia produces more work for those organs. Consequently, they become oversized and often do not function properly. The iron released from the accelerated destruction of red blood cells remains in the body and is deposited in all the tissues, especially the heart, liver, endocrine glands, and skin. Excess iron, or iron overload, in the heart and liver interferes with the function of these organs. Excess iron in the endocrine glands can cause diabetes mellitus, calcium deficiency, poor thyroid function, and impairment of sexual function. Growth tends to be slower than normal. With early diagnosis and newer forms of treatment, however,-children with thalassemia are able to grow more normally, and do not have the typical faces of inadequately treated children.

Without treatment, thalassemia patients die in early childhood, usually as a result of overwhelming infection. Although blood transfusions may be used to treat the symptoms of anemia, patients who receive only blood transfusions eventually die from complications caused by deposits of excess iron in the heart muscle. However, a combination of blood transfusions and chelation therapy has extended life expectancy into the late twenties (a chelator is an agent that binds and removes iron from the blood stream, preferably through overnight subcutaneous infusion via a syringe pump, done at home).

Treatment is stared at the time of diagnosis. The quality of life has greatly improved since the introduction of modern therapy, but life expectancy of patients who are treated from early infancy is at present unclear, since such therapy represents a recent medical advance.

As a thalassemia patient has described it herself, the realities and implications of the disease are:

"Every thalassemia patient must remind herself to:

1. put on her pump every night. The pump infuses overnight, for 10 to 12 hours and, for some patients, for 24 hours per day, depending on individual needs, an agent that binds the excess iron in the blood and excretes it through the urine.
2. go to the hospital every month, sometimes twice a month, for her blood transfusion, which involves:
   (a) a visit to have blood drawn two days before the actual transfusion so that the blood bank can cross match the blood of the donor with that of the patient,
   (b) a visit to have the transfusion, which implies sitting for eight to ten hours with a needle in one's arm pushing the time to go by,
   (c) possibly having a reaction to the new blood, which means having chills and extreme shivering with a rise in the temperature, increased palpitations and difficulty breathing. These reactions do not occur as often with new techniques of processing the blood of the donor,
   (d) feeling a little miserable the same day and possibly the next day, and
   (e) possibly catching viruses and other infections, such as hepatitis, A.I.D.S., and so forth.
3. the patient must remind herself to make and keep regular appointments with doctors.
4. she has to schedule a yearly check-up which involves spending at least three or four days at the hospital every year.
5. She has to live with the high likelihood of developing other serious conditions, such as heart disease, diabetes, liver and hormonal problems, and so forth.
6. Finally, she has to live with the increased likelihood of dying, especially if not following all the necessary treatments. Forgetting, or pushing out of awareness, these important aspects of the patient's life, although tempting, is also life threatening."

In this project, adolescents who suffer from thalassemia and their parents were asked to share their experience of how the illness has affected their everyday lives and how this is manifested in their day-to-day life. Particular attention has been paid to the following topics: How the illness has affected the adolescent/ parent relationship;

a. How the illness has affected their perception of themselves;
b. How the illness has affected their relationship with friends; and
c. How the illness has affected their perception of the future.

Participants:

Five families participated that had an adolescent between the ages of 13 and19 years old who had the disease. Two of the families were of Greek origin (one from Cyprus), two of Italian, and one from India. The adolescents were six-two boys and four girls. The youngest was fourteen years old and the oldest was 19 years old. Four families had two children, one of whom was healthy and one had the disease, and one family had four daughters two of whom were healthy and two had the disease (for demographic information see Table 1).

TABLE 1
Demographic Data

	Members of the Family[1]						Religion and Ethnic Identity		Education and Occupation of Parents			
									Education		Occupation	
	Age	Sex	Relationship	Member with the Disease	Disease Diagnosed	Treatment Started	Religion[2]	Ethnic Identity	Mother	Father	Mother	Father
Papas	39	F	Mother									
	43	M	Father				Greek Orthodox	Greek	Masters	Masters	Teacher	Teacher
	14	F	Daughter	*	1972	1972						
	7	F	Daughter									
Bianco	39	F	Mother									
	50	M	Father				Catholic	Italian	High School	College	Teacher's Aide	Carpenter
	19	M	Son	*	1968	1968						
	18	M	Son									
Sifneos	42	F	Mother									
	50	M	Father				Greek Orthodox	Greek (from Cyprus)	High School	High School	Secretary	Store Manager
	19	M	Son									
	15	M	Son	*	1971	1972						
Ranjit	37	F	Mother									
	44	M	Father				Protestant Christian	Indian	R.N.	M.Sc.	Nurse	Scientist
	18	M	Son									
	16	F	Daughter	*	1973	1974						
Boscolo	36	F	Mother									
	45	M	Father				Catholic	Italian	R.N.	High School	Nurse	Foreman
	16	F	Daughter	*	1970	1970						
	16	F	Daughter	*	1970	1970						
	14	F	Daughter									
	10	F	Daughter									

[1]No families included grandparents as members of their family.
[2]Religious background and current religious affiliation were the same for all families.

Procedures:

Interviews were open ended and consisted of an introduction and open ended question at the beginning of the interview and probe questions if necessary to clarify participant's ideas and to explore similar subtopics with all of the interviewees. The same introduction and set of questions was used for both the adolescents and the parents who were interviewed separately and for a maximum of one hour each. General introduction and open ended question were stated as follows:

The purpose of this study is to explore the effect that thalassemia has on your life. I am interested in anything you have to say about what having the disease means to you and how this is manifested in your day-to-day life.

Areas that were probed if they were not addressed during the first half hour of the interview were:

1. What is the impact of the illness on your relationship with your parents/adolescent?
2. What is the impact of the illness on your perception of yourself?

3. What is the impact of the illness on your relationship with your friends?
   and
4. What is the impact of the illness on your perception of the future?

Interviews were audiotaped and transcribed verbatim by an outside transcriber. They were analyzed thematically and the themes that emerged spontaneously and in response to the probe questions were summarized separately for the adolescents and the parents. Finally the themes that emerged from the adolescents were compared and contrasted with the themes that emerged from the parents.

Results:

For a detailed synopsis of the themes that emerged separately from the adolescents and the parents both spontaneously and in response to the probe questions see Table 2.

Parents and their adolescents expressed similar feelings and thoughts about how their life has remained normal despite the presence of the illness. They defined normal as the ability to do everything everybody else does. They claimed that they live a normal life and have learned how to live with the disease because: (a) their teenagers can do what other "normal" teenagers do and (b) because they are treated as normal by both their families and their friends.

In terms of the relationship between the parents and their adolescents we see that the parents worry more about, and in a few cases are closer to, their ill teenager than their healthy siblings. In addition, the teenagers reported that they are aware that their parents worry more about them. For some families it is clear that a special bond develops between the parents and the teenager. In one family parents mentioned that, this is also true for their relationship.

The parents related that they try not to treat their ill adolescent differently than their healthy sibling. The teenagers emphasized that they do not like it when they are treated differently. They both complained however that other adults do not understand what the illness is all about and tend to treat them differently. The friends of the teenagers seem to be better at this although when physical differences are obvious peers oftentimes tease and make fun of the ill teenagers.

In terms of the impact of the illness on their perception of themselves we observe that there are no common themes between the parents and the adolescents. The teenagers seem to have more to say about this topic than their parents. They reported feelings of sadness, depression, and the presence of self-deprecatory remarks. At least some of these feelings seem to be related to the extent of physical differences and the teasing they have experienced. Although these teenagers belong to a well treated group of thalassemia patients with few and minimal physical differences we still see that such differences are intimately related to issues of self-esteem and -self-worth. It is interesting that both the adolescents and their parents reported that they look at worse diseases as a consolation for their pain.

Differences are also observed in the experience of parents and their teenagers in response to the impact of the illness on their relationship with their friends. It is interesting that parents tended to be more secretive -except in one family- about the illness than their teenagers. The parents reported that they do not talk to their friends about it because: (a) they do not want their teenager to be treated differently (b) they do not feel comfortable talking about it and (c) they see it as a personal problem that

they deal with by themselves.

It is also interesting that the parents related that their friends and relatives do not understand what the illness is and do not know how to deal with it, whereas the teenagers said that although their friends may not fully understand the implications of the illness, they cope with it well and do not treat them differently. On the contrary they often serve a supportive function by being fascinated and interested in what the disease is all about and by seeing their ill friends as brave and courageous. Despite their positive experience with friends most teenagers relate that they are afraid their friends may run away and/or not like them because they are sick.

In terms of the impact of the illness on their perception of the future parents and their adolescents expressed similar concerns and fears. They both related that they are concerned and scared they might die earlier or that they may have further medical complications. They also shared that they are concerned and ambivalent about whether they can be independent and self sufficient; about whether they can have a career and/or a family and kids of their own. They are worried that they may not be successful in the working world and/or that they might get rejected and hurt by significant others because they are sick.

For the parents as a group we see that they all mentioned the time of the diagnosis as the most difficult and traumatic. Parents invariably described a long process of grieving and coming to terms with what has happened. We see the initial denial and disbelief of what happened and how true the diagnosis is. The “why us” stage of anger and bargaining -maybe this will be the last time we are coming to the hospital. The feelings of sadness and depression and the slow resolution and acceptance of the diagnosis. In this process of grieving and accepting the implications of the disease understanding seems to play an important role. Having information about the disease and doctors that can explain and help parents understand what is going on seems important for their better adjustment.

In some families the parents shared their feelings of guilt and self blame for what has happened. They shared that they feel responsible and in some way try to make up for it by providing as much as they can to their ill teenager. In two cases we see that the teenagers are aware of those feelings. The one teenager shared that he tries to “undo" these feelings by proving that he is okay --by making his life better and better. The other teenager expressed her discontent about her situation and her anger towards her parents for what they have put her through.

For the adolescents as a group we see that they started by saying how the illness does not interfere with their lives and ended by talking about how the illness often disrupts their plans for fun and for success in the future. Although they have learned how to live with the disease and they described living a normal life -have friends, are involved in sports, do well in school, have plans for the future- they also related how difficult it is to adjust to the illness because it affects every part of one's life. The pump in specific is viewed as the most difficult part of their treatment because it is an everyday pain and reminder of what they have. The adolescents also reported that their life would not have been much better if they did not have the disease but it would have been easier because they would have had less things to worry about.

Thus, in the current study we can see that despite the fact that parents did not deny how devastating and traumatic the initial diagnosis was for them and that although both the parents and the teenagers did not deny their worries and their fears about a premature death and further medical complications, and the extra difficulties the illness has brought to their lives, they both mentioned that their life has remained

TABLE 2
Themes That Emerged from Parents and Adolescents Regarding the Impact of Thalassemia on Their Everyday Life

PARENTS	ADOLESCENTS
Introduction and Open-Ended Question	
Diagnosis—Devastating, shocking, heartbreaking, difficult to accept, unexpected, and traumatic	**Normal Life**—Illness does not interfere with their lives; they can do everything everybody else does
Normal Life—Teenagers live a normal life; can do everything everybody else does	**Worries and Difficulties**—Adjusting to the illness is difficult; hard to accept; worry about what else might happen
Important to Understand the implications of the illness and what it is all about	**Friends**—Their friends know and do not treat them differently
Worries and Difficulties—What else may happen; financial burden; complications and physical differences	**Doctors**—Give them lectures; do not know everything; should listen and respect their personal experience
Probe Questions	
I. Impact of the Illness on Their Relationship with Their Parents/Adolescent	
No Special Treatment—Special treatment implies they are worse off; do not spoil them and pamper them but encourage them to be like other kids	**Feeling Special**—Know that their parents worry more about them than their healthy siblings; afraid they might get hurt; closer to their parents, who are more understanding
Special Feelings—They worry more, they feel closer, feel sorry but do not let it interfere with how they treat them	**Upset When Treated Differently**—They do not like it when they are treated differently
Consolation—Look at worse things around them	**Leaving Home**—Half have plans to leave home, one is concerned she may not be able to, one does not want to leave home
Trust and Responsibility—Trust that their teenagers can take care of themselves	
II. Impact of the Illness on Their Perception of Themselves	
Decision to Have More Children—Three of the five immediately decided to not have more kids	**Self-Worth**—Put themselves down; feel unsure, sad, discouraged and sorry for themselves
Diagnosis—Disappointment of their life; reacted with sadness and disbelief	**Self-Image**—Feel conscious of how they look; become target of teasing because of how they look
Guilt and Responsibility—Feelings of guilt, self-blame, responsibility	**Consolation**—Looking at worse pain helps their pain
	Fear of Rejection—Afraid of how others will react; worried they may not like them and reject because they are ill
III. Impact of the Illness on Their Relationship with Their Friends	
Whom to Talk to—Do not tell their friends because they do not want their teenagers to be treated differently and because they consider it their personal problem	**Friends Know**—Has not affected their relationship
	Fear of Rejection—Worried that when they hear about the illness will run away and not like them anymore
How Their Friends React—Variety of responses is mentioned from supportive and sensitive to critical and noncompassionate	**How Friends Respond**—Do not treat them differently; see them as courageous and support them; fascinated and want to know all the details
How the Friends of Their Teenagers React—Do not treat them differently	**"Forgetting"**—Do not want to be reminded of the illness; forget what they have and do not dwell in it
IV. Impact of the Illness on Their Perception of the Future	
Fears, Worries, and Concerns—Worry about the prospect of an early death and further medical complications; feel discouraged about the future; are concerned about whether their teenager will find someone to marry and be self-sufficient	**Fears, Worries, and Concerns**—Scared and concerned about the future; their life may be shorter, they may have other medical complications, they may have to settle for less and not be able to be independent
Positive Thinking—They think positively; look at medical advancements	**Positive Thinking**—Hopeful about progress in the medical field
	Plans for the Future—Want to offer to others; want to study; want to make money and have their own house

normal and that they have learned how to live with the disease. Furthermore, the teenagers mentioned that despite feelings of discouragement and depression, self deprecatory remarks, fear of rejection, awareness of physical differences, etc. they have friends, are "one of the gang", get along with others fine, do well in school, have hobbies and interests, and have plans for the future.

Discussion:

George Vaillant in his book *Adaptation to Life* (1977) has described certain coping mechanisms that some of the "healthiest and most promising" undergraduates of one of "America's leading universities" used in order to adapt to life. We see these same mechanisms of suppression, anticipation, sublimation, altruism, humor, and hope used by these families. They suppress the impact of the illness on their lives by not thinking and talking about it all the time. Both the parents and the teenagers suggest that talking about it all the time makes it seem like a big issue and interferes more with their everyday lives. They prefer to forget about it and to not dwell in it. This is why it is important for mental health professionals to be sensitive to such coping mechanisms and to not necessarily view them as "denial", "secrecy", or "pathology".

The parents emphasized that it is important for the well being of their teenagers to create a normal environment in which they can live as normal teenagers. Special treatment implies that they are worse off and this is the wrong thing to do for these kids. On the contrary it is important to stop themselves from spoiling them and overprotecting them. However, as some teenagers pointed out, often the medical system takes over the overprotective role and poses unnecessary restrictions on the ill teenagers.

The parents also manifest the ability to foresee and pre-plan for future difficulties. They are not denying in a way that inhibits them from seeing reality. They see reality and they make plans in anticipation of future problems but they do not let the illness interfere with their everyday life. Furthermore, they try to put the illness in perspective and they are able to see how much worse it could have been instead of how much better. The parents mentioned that accentuating the positive while "keeping the window to hope always open" is also an important mechanism for coping with the stresses associated with the illness. They keep their hopes up for progress in the medical field and for a better future.

The adolescents related that they do not dwell in their feelings and do not let the illness preoccupy their mind. Instead they are active and have the courage to go after what they want. They have plans for the future and they want to offer to other people. They emphasized that the attitude one takes towards one's self and one's illness is important for successful coping. Of course parents' attitude and adjustment to the illness plays an important role in determining the teenagers' attitude and adjustment (7). The parents agreed that in this respect it is important to not feel sorry for their ill teenager and to transmit to them a positive and hopeful attitude.

Furthermore, the teenagers mentioned that having their parents there to talk to, to understand them, to support them, to build their confidence, and to give them extra tender loving care has been very important for coping with the illness. The closeness and the special feelings that develop between the parents and the ill teenagers serve as a coping mechanism rather than as a coalition that is dysfunctional. Mental health professionals may view such structural changes (12, 13, 16, 17) and bonds that develop in response to the illness (2, 8) as pathological and may try to change them,

but this may not be in the best interest of their clients. It is important for professionals working with families that have a chronically ill individual to be careful not to jump to their own conclusions about what should or shouldn't be but to rather respect their clients' experience and learn from them.

It is important to mention certain factors that may have contributed to these teenagers ability to successfully adjust to the trauma of the illness. These factors are: a) recent medical advancements, b) socioeconomic background, c) educational background, and d) "holding environment". As George Vaillant (1977) has mentioned, altruism and sublimation may be "luxuries that are derived from internalized human resources" that not all people can afford, and that "it is not the isolated traumas of childhood that shape our future, but the quality of sustained relationships with important people" (p. 29).

The teenagers that participated in the current project are lucky to be born in an era during which more is known about thalassemia and where there are more treatments available. Although new and improved treatments are available not all families can afford to provide for all that is needed. However, the families that participated in this project are able to provide for all that is necessary, although some with great struggle. In addition, it is important to remember that these families are educated enough to understand the evolutionary, genetic aspects of the disease. They can thus view the individual with less prejudice and misunderstanding, whereas in the countries where the disease is most prominent thalassemia is often viewed as a stigma for the whole family. Both the individual and his/her parents are faced with prejudice about what "they have" and often are isolated and excluded from their social network, even their extended families.

Finally, it is important to remember that the teenagers that participated in this study are privileged because they are being brought up in families that not only can afford treatment, can understand the implications of the illness and can treat them as normal individuals but also cared enough to struggle and provide them with the best they could. The parents in these families are not abusive, are not neglectful, are well adjusted individuals with no obvious evidence of major psychiatric illnesses or other serious personality disturbances. These teenagers described their parents as always there to help them, to comfort and support them. These teenagers are unfortunate to have been born with a chronic illness, but fortunate to have been born into families that are "privileged".

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