SPECIAL ARTICLE

THALASSAEMIC TEENAGERS' AND YOUNG ADULTS' OUTLOOK ON LIFE

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Abstract

Introduction: Thalassaemia major (TM) is an inherited disease that can affect the psycho-emotional sphere of people with the disease and shape their expectations for life.**Aim:** The aim of this study was to investigate in the literature the expectations of people suffering from thalassaemia major across the age range.**Material-Methods:** Recent articles and studies in English language were searched in the electronic databases PubMed, Cinahl, Google Scholar, Scopus. The keywords used were «expectations for life, thalassaemia, children, teenagers, young adult and adult».**Results:** The literature on the expectations experienced by people with thalassaemia major is very limited, without a specific measuring tool. The researchers concluded that when thalassaemics receive their treatment, they are able to cope with the difficulties of life that arise in adolescence or adulthood. In the last five years, a measurement tool has been developed for all age groups that can assess the level of expectations for all age groups and fulfillment for young adults and adults.**Conclusion:** Appropriate treatment of the positive and negative expectations felt by patients with thalassaemia major a helps to improve the standard of living of pediatric patients and their smooth development into psycho-emotionally healthy adults.

Keywords: expectations for life, thalassaemia, children, teenagers, young adult, adult

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Introduction

Hemoglobin diseases are hereditary hematological disorders that affect globally more than 300.000 infants every year.[1] Thalassaemia which is located in more than 60 countries, is recognized as the widest spread blood disorder universally from WHO.[2] Thalassaemia is caused by certain mutations in groups of hemoglobin genes that result in weakening the composition of one or more of the chains of the quadripartite hemoglobin. [3] Depending on the type of the involved hemoglobin chain, there are two distinct thalassaemic types: "A" and "B". [4] Beta-thalassaemia is the most important type as it is the most widespread and it results in severe anemia in homozygous and composite heterozygous form.[5]

Beta-thalassaemia has a variety of clinical symptoms. In mild cases it doesn't require treatment. In serious cases there is severe anemia with important alterations in the bones and it can be deadly without proper treatment in childhood and in the Pacific Islands.[6,7] WHO's statistics show that in those areas' thalassaemia frequency is between 0,1-4,9/1.000 births. In the recent years due to immigration hemoglobin diseases are not limited anymore to those high frequency areas. [8] Studies show that 3-10% of the global population caries the gene of thalassemia.[9] There are about 200.000 verified patients of thalassaemia with a tendency of raise of 60.000 every year.[10] In Greece thalassaemia appears almost everywhere in the country. Owing to prevention measures, only 319 affected births were reported from 2000 to 2015 instead of expected 2250 to 3000 and right now 8% of Greek population are caring the gene.[11,12]

The purpose of this paper is to research through the literature the expectations of thalassaemic patients in all the age spectrum. Today the patients of thalassaemia major can survive to mature early adult life.

Materials/methods

A literature research was carried out for articles in PubMed, Cinahl, Google Scholar and Scopus databases to identify articles that are concerned with expectations of the entirety of the age spectrum of thalassaemic patients. The key words used to carry this research were "expectations for life, thalassaemia, children, teenagers, young adult, adult". We sought descriptive and research articles, published after 1995 until 2021 and were focused on expectations of thalassaemic patients, written in the English and Greek language.

Results

In the paper of Zani et al., 1995,[13] an evaluation of the effects of thalassemia in the psychology and social behavior of Italian teenagers was made. The results showed that thalassaemic teenagers had a normal psychological development with self-esteem and coping mechanisms in place. They were thinking positively about their future, were placing high in their priorities the finding of employment and were searching for the right partner in life.

The same team from Ferarra Italy [14] researched in 1998 the effects of thalassaemia in psychosocial adjustment of teenagers and young adult patients. They were asked to fill a questionnaire (specifically tailored), and married thalassaemic patients were

interviewed. Some of the married thalassaemic patients had married and they had children. They were asked about marriage and social life. The researchers found that thalassaemic teenagers had normal psychosocial development with developed self-esteem and were well socially adjusted while they had developed strong bonds with their families. Married thalassaemics did not differ in behavior from healthy married couples. The research team concluded that when thalassaemics were receiving the appropriate treatment, can manage life's difficulties that are arising during puberty or in their adult life.

In the research with thalassaemic patients with ancestry from Greece and Italy, performed in the USA in 1998 [15] evaluated the orientation towards the future and the way thalassaemic patients were functioning on a psychical level regarding among other concerns, their future expectations and the way they were understanding social support. Through this, study the importance of coping strategies in the context of the chronic nature of an illness like thalassaemia, which is thought to lead to disability. It was also found that thalassaemic patients had just like their peers, expectations for their future, their social acceptance and image.

In the last research that included among others the parameter of expectations and was performed a few years ago in Crete, the results showed that thalassaaemic expectations were influenced by sex and admittance to hospital. Their attitudes and expectations in relation with the experiences they had from their therapeutic treatment showed the need chronic patients from thalassemia had into receiving better treatment and psychological support. [16]

Nevertheless, until recently there was no published tool to assess the content structure of expectations among thalassaemic patients. There was a study that affirmed the usefulness of a new multidimensional questionnaire of expectations that is appropriate for thalassaemia patients (multidimensional expectation questionnaire thalassaemia major patients MEQ-TMP) who reach their 4th decade of life and there were established four sub scales: Social support, Family orientation, Career development, Daily life. Cronbach's alpha for the whole scale was 0.87. This particular tool of assessing expectations MEQ-TMP was developed with 15 questions for each age group of patients with thalassaemia is a useful tool for the clinical staff that offers care to thalassaemic patients so that they can understand their perspective about life as they age, to have better psychosocial adjustment to the chronic nature of the disease, to be able to live as closer to normal as possible and to fulfill their ambitions, improving thus their satisfaction levels about their quality of life.[17]

This particular measuring instrument of expectations (MEQ-TMP) was adjusted for younger ages. The above questionnaire of expectations was adjusted to children and teenagers with thalassaemia to include 14 questions and three sub-scales: Family and daily life perspective, Social support and work perspective with Cronbach's alpha for the whole scale 0,81.[18]

From the results of the study, it was proved that all 303 of the young adults with thalassaemia fulfilled/ met the totality of their expectations. They were able to achieve more than they expected on the area of their carriers. Their high educational

level and their unobstructed access to blood transfusions, were helping them to overcome obstacles and to pursue career advancement. Thalassaemic young adults who participated in the study were lacking in comparison with their expectations in the areas of family. They were concerned about the possibility of complications and problems and were avoiding to plan for starting a family. It was also proved that the more they were informed about their health condition the stronger they became and they were facing better the challenges of their illness. On the other hand, the functionality of young thalassaemic patients was influenced by problems in blood transfusion. Finding the blood, they needed allowed them to have the social life and the social bonds they were expecting. In their majority (71,95%) they had normal depression levels resulting in positively affecting the fulfillment of their expectations, to be able to continue to expect support from their social network as it was already offered, to cope with their everyday activities as they desired and to hope to create a family.[18]

The results of this study with 74 children and teenagers with thalassaemia were that they had high expectations. Thalassaemic teenagers had a total score of expectations [3,10] and the score of expecting to create family/expectations of everyday life [3,15] was positively related with better knowledge of advanced therapies. Similarly their expectations about friends networking were equally high fact that was interpreted by the providing of oral chelation agents. The mean score of their expectations for carrier advancement was 2,68 and was positively related with the close relationships they were developing with nursing staff. Their close relationship with nursing staff helped them to discover what they wanted to do in their future lives. Better knowledge of new therapies helped them to continue having expectations and to project into the future how they wished their family life and their everyday life to be. In their majority (87,84%) had depression levels that were normal and their school absenteeism was related with the appearance of depression symptoms. The desire of the 74 children and teenagers with thalassaemia to have a circle of friends was affected by their good mood and their lessened strain during the chelation process. [19]

Discussion

For the first time a questionnaire that was multidimensional was created and adapted from the corresponding questionnaire for young thalassaemic patients and was used to research for the first time the expectations of children and teenagers with thalassemia in Greece. [17,18,19] The only similar effort about the expectations of teenage thalassaemic patients was one 25 years ago by the research group in Ferrarra, Italy [14] That research team had created an ad hoc questionnaire that wasn't validated about its validity, reliability and usefullness, if it was covering needs associated with their age like family relationships, social adjustment, heterosexual relationships, self-consciousness and coping mechanisms. [14] Nevertheless, it could along with the questionnaire of young adults to be studied further and to be more thoroughly edited in the context of timeless comparison of expectations thalassaemic patients have as the age.

Young adults with thalassaemia in Greece have achieved to take up their corresponding adult responsibilities. Although they live in a country with well-organized thallassaemic treatment facilities, they still fear the possibility of lack of blood transfusion and they seek support from their social circle. In thalassaemic children and teenagers, the knowledge of new treatments helps them to have elevated expectations and their friendly associations are affected by their good mood and their lessened strain during the chelation process. [18] Without doubt, people with thalassaemia need support with the goal of detection and treatment of their psychological problems, the improvement of their inner conflicts, the development of coping strategies, the adoption of appropriate behavior, of accepting their illness and the compliance to their therapeutic treatment along with a more realistic attitude towards illness.[18,19,21]

Equally important is considered the raising of awareness of the medical and nursing staff in matters of diagnosis and treatment of emotional disorders related to the illness. The appearance of psychosomatic disorders is frequent in people with thalassaemia and often is not treated sufficiently either because health professionals believe they are inevitable or even a natural consequence of the disease or because children are not in a position to ask for help. The treatment of psychological disorders from health professionals contributes to the amelioration of the life status of pediatric patients and to their smooth development into psychoemotionally healthy adults.[18-21]

Conclusion

An efficient prevention strategy for thalassaemia in Greece demands the contribution of all experts from the field of health to organize programs of prevention and education of high risk.

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