RESEARCH ARTICLE

Depression and Thalassemia in children, adolescents and adults

John Koutelekos [1] Nikolaos Haliasos [2]

1. Laboratory collaborator, Nursing Department A, TEI, Athens, Greece
2. Associate Professor of Pediatrics-Hematology, Medical School, University of Ioannina

Abstract

Thalassaemia consists one of the most common chronic and genetic hematological disorder, globally. Moreover, is a serious life-limiting and potentially life-threatening disease that causes substantial disruption in all dimensions of life. As more effective management and even cure of thalassemia become possible, attention is drawn to the evaluation of depression that frequently ollows suffering individuals.

Aim : The aim of this study was to review knowledge about depression in thalassemia detailing in children, adolescents, adults.

Method : The method of this study included bibliographic review of the literature, mainly in the PubMed data base, which referred to depression in thalassemic children, adolescents, adults. The search of the literature was conducted from 2001 to 2012 using the following key search terms: “depression”, “thalassaemia in children-adolescents-adults".

Results : Improvement of therapeutic regimen has significantly increased life expectancy of thalassemic patients but at the same time has created significant ongoing health care needs. According to the literature, depression involved in surviving thalassaemia is increasingly recognized, globally. The main factors triggering the onset of depression are personal, family, social or hereditary. Depressed thalassemic children, adolescents and adults face many difficulties in all facets of life. More in detail, depression in thalassemia is mainly related with low school or job performance, social isolation, failure of independence, maladaptive coping strategies, compromised physical ability and limited life opportunities. Provision of accurate information to thalasssemic individuals about the therapeutic regimen by expertise health professionals is a key-factor that plays a vital role on the outcome of depression.

Conclusions : Regular screening for depressive symptoms is essential to identify at-risk individuals so as to provide appropriate psychological support with ultimate goal to improve both emotional and physical health.

Keywords:Thalassemia- depression-children-adolescents-adults

Corresponding author : Koutelekos John, Agiou Nectariou 14, Glyfada Athens. PC 165-62, Tel : 6974875766.

Introduction

Thalassaemia consists one of the most common chronic and genetic hematological disorder, globally. There are approximately 240 million people worldwide who are heterozygous for β-thalessemia while approximately 200,000 affected homozygotes are born annually.[1] Greece, is a country with a mean frequency of thalassaemia carriers at 7 perent plus 1 percent of carriers of haemoglobin S and number of births surpassing 100.000 yearly.[2]

Thalassemia is a major health problem not only for the patients and their families but also for the Public Health System of each country due to the cost of treatment involving regular transfusions, iron chelation, frequent hospitalizati on and general medical follow up. [1,2]

As more effective management and even cure of thalassemia become possible, attention is drawn to the evaluation of psychiatric disorders

such as depression that frequently accompany these individuals.[3,4] Depression is a chronic disease which needs medical treatment and poses several limitations to patients' life given the disabilities that provokes, thus exerting a negative influence on their quality of life.[5]

Though the etiology of depression in children and adolescents is not well understood compared to adults, however it is considered as a result of interaction between different factors. The main factors involved in the onset of depression are stressful events (chronic illness), family-related factors (death in the family, divorce, physical abuse, intra-family conflicts or economic difficulties that undermine the quality of the relation within family) or social factors (peer group or school when children experience it as "pressure"). Relatively, hereditary factors seem to be responsible for the onset of depression, such as mental illness in the family that significantly increases the risk for the child to develop depression. However, it is not fully clear whether inheritance is responsible for the development of depression.[3,4,6] Furthermore, depression in children has demonstrated disturbances in the metabolism of neurotransmitters and endocrine disorders mainly cortisol as in adults. Finally and most strikingly, the way a person perceives the negative experiences seems to be responsible for depression onset, according to cognitive theory. [6-8]

Children and adolescents with major depressive disorder are at increased risk for suicidal behavior, substance abuse, physical illness, early pregnancy, exposure to negative life events and low educational or psychosocial functioning. The main factors associated with suicidal behavior are serious family difficulties, parents with mental illness and a high level of antisocial behavior in the family.[6-8]

Because mood disorders of children and adolescent are likely to continue into adulthood, identification, assessment and treatment at earlier stages are warranted for achievement a better prognosis in adulthood. What is more intriguing is that there is a scarcity of data about the effects of implementing psychological intervention on the outcome of depression.[6-8]

Nowadays, thalassemia consist a significant issue for health professionals since they don't only have to accommodate the demanding care including blood transfusions and iron chelation therapy but also to screen and treat psychiatric disorders and enhance psychosocial adaptation towards the disease.

Depression and thalassemia in children

The concept of depression during childhood has been a matter of controversy since this period is widely accepted as time full of joy without symptoms of despair. Following this line of view it becomes apparent that depression in children remains undiagnosed and consequently untreated. Additionally, failure of diagnosis is attributed to differences in clinical presentations across developmental stages that often raise the question of whether childhood-onset depressive disorder is a different illness than adults. Moreover, an equally important issue is whether depressive disorders in childhood consist an increased risk for psychiatric diagnoses in adulthood.[6-8]

However during the last 20 years, it has been slowly acknowledged that children may develop depressive disorders similar to those in adults. Children more frequently develop social withdrawal, complain of psychosomatic symptoms, such as headache, abdominal pain or show irritability, poor school performance, social isolation and inability to handle frustration.[6-8]

During the first years of life, children are unable to understand the real extent of the disease, however, they feel the general climate of disorganization that follows the disease. As they mature, they become more aware of the nature of thalassemia thus developing denial and

psychological distress. [9-11]

The impact of the disease in many dimensions of life becomes increasingly evident during the preschool and school age when children seek for independence. Another significant area at this period is related with comparisons between children and their peer group. Thalassemic children are forced to acknowledge differences between themselves and others which are associated either to the physical dimension (facial appearance, stunted growth, bone deformities) or their inability including lack of energy to accomplish daily tasks and prior physical activities that used to enjoy.[9-14]

Given the often unpredictable and fluctuating course of thalassaemia, it is apparent that the disease is related with frequent re-hospitalizations and repeated absences from school and therefore low school performance. Numerous other factors are responsible for low school performance, such as impaired abstract reasoning, deficits of language, attention, memory or visual spatial skills and executive functions which are more prominent in hemosiderotic patients.[9-14] Moreover, silent brain infarcts may be responsible for neurological and cognitive impairment mainly attributed to the chronicity of the disease.[15] Children should be evaluated so as to have early intervention, special education and psychological support.[16]

Controversy exists over the possibility of quitting school. For example, according to a survey conducted by Khodaie et al.,[11] in Iran, 9.6% of patients quit school due to the disease whereas a study conducted in Greece by Vardaki et al.,[13] showed that the education of thalassemic individuals was not affected by the disease. Quitting school is mainly attributed to the deficit of knowledge regarding the disease or wrong reactions and perceptions of parents.[17]

The chronicity of the disease affects negatively childrens' social life because it reveals the disease, triggers comments or questions in their environment, mainly the school. Usually, thalassemic children refuse to discuss their health problem with friends and dependent on parents. It is worth noting that each child has different ways of facing stressfull experiences including chronic illness. The different responses depend on their personal characteristics, age, stage of cognitive development, adaptability, previous experiences of illness, etc. [10,18,19]

Children at school and pre-school age need to experience confidence by their caregivers. However children feel psychological distress when caregivers put less emphasis on the importance of providing information or explanations to children about the nature of the disease.[6]

All factors mentioned above, have deleterious effects on children's self-esteem, personality development and emotional health. Usually children develop negative thoughts about their lives, experience feelings of loneliness, isolation and psychiatric morbidity that make harder their integration into social environment. [20-23]

Indeed, the ultimate goal of caring children with chronic disease is equal treatment and integration into society. More in detail, smooth social integration prerequisites avoidance of any privilege or overprotection, promotion of independence and autonomy, grow of interests within the limits that the disease imposes, avoidance of feeling "being different from the peer group" and alleviation of psychological distress. [20-23]

Teachers' role has a beneficial impact on minimizing the emotional burden of thalassemia disease since they are often able to change negative attitudes or perceptions of classmates thus offering precious help for their integration into society. It is widely accepted that enhancing awareness of teachers, strengthening their cooperation with health professionals and creating a strong bond between them and

parents and children significantly contribute to treatment of both chronic disease, thalassemia and depression.[20-23]

Additionally, schools are considered as the most suitable environment for the prevention, detection and early treatment of depression so as to help children to become responsible individuals free from mental disorders in adulthood. [20-23]

Depression and thalassemia in hospitalized children

Admission to hospital is a traumatic experience for both children and family as well as a significant destabilizing factor of life. In contrast with adults and adolescents, children are unable to understand the necessity of treatment and hospitalization.[24,25]

Numerous factors may trigger psychiatric disorders during hospitalization. However, separation from parents is the main stressful factor in

infants, school and preschool children. Regarding infants interruption of "mother-child" bond" is related with negative implications for the further psychological development. Similarly, preschool children are vulnerable to stress since they are more dependent on their parents, have greater needs and experience fear for frequent blood transfusions or administration of chelation. Older hospitalized children also experience lack of their personal space or lack of friends and anxiety for the success of therapeutic intervention. The existence of prior separation experience from their parents or other stressful and traumatic experiences related with emotional deprivation are also factors that trigger psychological distress in children.[23-29]

Another commonly held view is that psychological distress of hospitalized children is mainly attributed to the unfamiliar environment of clinical settings. For example, the conditions of hospitalization, the intense rhythms in clinical environment, the children's relationship with other hospitalized children and even sounds and smells are responsible to trigger some fears. Moreover, health professionals under the pressure of work, adopt an unfriendly behavior or even require full compliance of children. Finally, factors associated with the disease itself, such as the severity of the disease and the therapeutic treatment may arouse psychological distress.[23-29]

Individualized care according to the developmental stage of each child maximizes the benefits of hospitalization. For example, creative and therapeutic interventions that highlight personal skills, ensure smooth transmission to previous life, maintain daily activities are considered to minimize the loss of control and reduce fears arising from changes in physical dimension, thus preventing the onset of depression.[23-29]

Additional factors that markedly minimize the negative impact of hospitalization are early discharge from hospital and return to normal life, reintegration into school life, attitude of parents and teachers, the guidance for parents about access to health services and development of effective communication between parents and health professionals. It is not rare that children develop after discharge maladaptive coping strategies and experience emotional or behavioral disorders, such as regression, attaching to parents, sleep or nutrition disorders, behavioral problems at home or at school.[23-29]

Therefore, before discharge, it is necessary to assess both the needs of pediatric patients and levels of depression. Given that each child with thalassemia is a separate entity with its own personality, needs and defense mechanisms, detection of depression should be an integral part of the therapeutic approach. Interestingly, thalassemic children may require long-term psychological support. Cognitive-behavioral therapy can be an effective psychological approach because it contributes to treatment

compliance, reduces emotional burden of disease and improves quality of life. [23-29]

Also of importance is the acknowledgement of parents' stay by hospitalized children that contributes to adaptation to clinical settings and maintenance of emotional health. Health professionals should be aware of the needs of parents and encourage their participation in the therapeutic regimen. The concept of holistic approach enhances family stay in the hospital while the effective cooperation of parents and health professionals contributes significantly to the successful long-term management. [23-29]

Depression and thalassemia in adolescents

Adolescence, is an intermediate phase between childhood and adulthood, where takes place a permanent change in the body. Furthermore, adolescence is accompanied by many challenges, such as social, personal and career. At the same time, it is a period when significant changes in mood take place for various reasons. For example, teenagers wish to go though new circles in their life, experience grief for the prior joys of childhood and face emotional conflicts due to the release from their parents and the uncertainty of finding other sources of support. [30-35]

On the contrary, thalassemic adolescents, being already affected by the chronic illness and having realized the impact of its' chronicity, are more vigilant of their illness progression and potential health hazards. Accordingly, they face significant problems in all facets of life that contribute to the onset of depression. The main factors involved in psychiatric disorders are family (overprotective, negligent, or hostile parents), social (uncompassionate peers) and the burden of disease (complications, blood transfusions, iron chelation). Furthermore, thalassemic adolescents experience feelings of shame or denial, uncertainty about the outcome of the disease and the fear of stigmatization or the imminent death that impose restrictions on social life. [30-35]

The part of life that is mainly affected by the disease is the relation with peers or the social environment. At this age, thalassemic individuals are more self-conscious of their adversely affected physical appearance and consider that the illness has negatively affected their academic performance and their heterosexual relationships. Regarding the opposite sex, they feel different due to their delayed sexual development, awkward physical appearance including changes in body image. [30-35]

It is widely known that psychological separation and growth of self throughout adolescence are essential for the normal process of maturation. The period when normal teenager is trying to become independent, the thalassemic strives to live normally making hard efforts to handle the daily disabling nature of the disease which now requires more attention and planning, that previously might have been managed by their parents.[17] At the same time, the independence that the adolescents seeks for, is limited by the disease and its' complications leading them to "passive" state. For example, adolescents can not exert control of their body because they dependent on blood transfusions, which despite they relieve the symptoms of anemia however they imply increase iron load. Indeed, chronic therapy is a constantly reminder of vulnerability and dependency on others, that fosters personal growth, self identity and mental health. As a result, the period of adolescence may be expanded or receive an abnormal frame, such as complete dependency on parents, which is a limiting factor in the process of maturation.[30-35]

However, it is crucial to take into account the way individuals perceive their disease. Acceptance of the disease is the key-element for normal transition to adulthood. [30-35]

Health professionals and supportive net of adolescents (family, social,) play a crucial role in making them feel confident, build self-esteem and eventually become a self-sufficient individual.

Specifically, provision of care based on deep understanding both adolescents' development and their needs in conjunction with appropriate support contribute significantly to the development of adaptive strategies and defense mechanisms, which are a prerequisite for satisfactory social adjustment and prevention of psychiatric morbidity. [30-35]

One aspect in need of closer notice is recognizing clinical presentation of depression in adolescence which includes loss of appetite or excessive eating, sleeping difficulties, restriction of activities, somatic complaints replacing the subjective complaints and poor concern for physical appearance and mental health. Frequent manifestation is antisocial behavior and suicidal tendencies.[30-35]

Depression and thalassemia in adults

Improvement of thalassemia treatment including blood transfusion and chelation therapy has significantly increased life expectancy of patients. Initially the disease was less common in adults and in the early days of treatment, thalassemia was regarded as a condition of the young mainly treated by pediatric teams. Since 1960s, with advances in haemato-oncology, median survival in thalassaemia major has increased from 16 to 30 years. [36-39]

As an increasing number of young people reach adulthood, thalassemia has become a chronic disease having significant ongoing health care needs that must be delivered in an adult rather than pediatric setting, a process known as "transition". Indeed, adults being at risk for many complications, such as osteoporosis, cardiac disease, organ failure, require treatment in a not pediatric setting. This problem has two options, from the one hand health care professionals are less prepared for this transition or are unfamiliar with adult-specific issues on the other hand many centers remain focused on the care of pediatric patients including no availability of necessary equipment. Interestingly, adult patients moving from thalassemia centers to adult health care facilities, face the risk of not receiving proper treatment mainly attributed to the lack of specialists and centers. Management of any chronic illness, need ways to ease this important transition and prerequisite effective communication between health care professionals in children and adult settings. [36-39]

Nowadays, that patients move from the pediatric age group to adulthood and maturing adults experience different needs and challenges the distress has come to the forefront of clinical practice. It is noteworthy that causes of distress disorders appear to vary over the years and the improvement of treatment. For example, in previous decades high prevalence of psychosocial health disorders derived from their accommodation with hard life conditions and expecting death. However, in contemporary time, psychosocial health disorders are attributed to poor socio-economic state, uncertainty about the future or to concern of being a burden to the family.[17] Other factors held responsible for the onset of psychosocial distress are long-term complications of the disease, fertility, limited family, educational and carrier opportunities. Significant concerns are raised about its' treatment as it has been shown to exert a negatively influence on the therapeutic regimen. More specially, is considered to be a significant impediment to compliance with ongoing therapy.[36-39]

Routine hospitalization also exposes individuals to great risk of having to reveal their disease to colleagues and employers thus facing potential bias and discrimination. Enhancing public awareness after taking into account socio-cultural and religious factors is important to dispel any misconception and avoid being stigmatized by society. [36-39]

Regarding sex, there are observed differences with women to assess the

psychosocial aspects of the disease as important factors affecting quality of life. A possible explanation is that women live longer, withstand chelation therapy and have lower rates of cardiovascular event. [37]

In developing countries where is observed lack of public awareness or the cost of therapy is either too expensive or not available, helpless thalassemic adults at their effort to cope with the disease may experience severe emotional difficulties, such as frustration, sadness, hostility, depression, anxiety, fear of death, lack of confidence, isolation and anger.

Conclusions

Providing information to patients in understandable and accurate terms about the nature of the disease, the need for treatment, the new medical protocols, has beneficial effects both on the outcome of thalassemia and depression. Knowledge deficits may result in unnecessary depression. Provision of information should be individualized and appropriate to the age, developmental stage, psychological maturity, personality and family environment.

Thalassaemia patients require lifelong psychological support for prevention of mental health issues. The ultimate goal of implementation psychological programs for thalassemia patients is to combat depression by enhancing their integration into the social mainstream, minimizing knowledge deficits and providing help to plan and actualize their educational, personal and career goals thus leading fulfilling lives.

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