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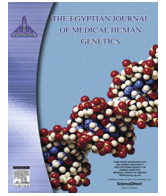


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Original article

Evaluation of health-related quality of life and muscular strength in children with beta thalassemia major

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ABSTRACT

Background: Thalassemia is an inherited blood disorder that requires repeated blood transfusions and chelation regimes. This may lead to restrictions in physical activities, social participation as well as decreased muscle strength.

Aim: The aim of this study was to evaluate the health-related quality of life (HRQoL), muscular strength and pain in children with β -thalassemia major.

Patients and method: One hundred and twenty children (60 with β -thalassemia major and 60 age-matched healthy) were participated in a cross-sectional study from both sexes (57 girls and 63 boys) with ages ranging from two to twelve years. HRQoL (physical, emotional, social and school functioning), muscular strength and pain were evaluated for all children by using the pediatric quality of life inventory™ (PedsQL™) 4.0 generic core scale, hand-held dynamometer and visual analogue scale (VAS) respectively.

Results: Children with β -thalassemia major showed a significant decrease in all domains of health-related quality of life and handgrip strength with a significant increase in VAS score ($p \leq 0.0001$).

Conclusions: The study concluded that thalassemia as a chronic disease has a negative impact on HRQoL and muscle strength of children in different age group.

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1. Introduction

Thalassemia is a genetic blood disorder, which is characterized by decreased synthesis or absence of globin [1]. This synthetic defect leads to the formation of fragile abnormal red blood cells (RBC), which can be easily hemolyzed, leading to chronic anemia [2]. According to clinical and hematological conditions, the severity of the disease is classified into three categories; beta thalassemia carrier state, thalassemia intermedia and thalassemia major [3].

The World Health Organization (WHO) recognized thalassemia as the most prevalent genetic blood disorder in the world, found in more than 60 countries with a carrier population of up to 150 million [4]. This disorder is highly prevalent among children in the Middle East, Mediterranean region, and South Asia [5].

Thalassemia is a chronic disease and presents with a wide range of serious clinical and psychological challenges. The effects of

thalassemia on physical health can lead to physical deformity, growth retardation, and delayed puberty [5–7]. Its impact on physical appearance, e.g., bone deformities and short stature, also contributes to a poor self-image [5,7]. Severe complications such as heart failure, cardiac arrhythmia, liver disease, endocrine complications, and infections are common among thalassemia patients [8]. These problems do not only affect patients' physical functioning but also their emotional functioning, social functioning and school functioning, leading to impaired health-related quality of life (HRQoL) of the patients [5,7,9,10].

Children with thalassemia are less active than their healthy peers and generally have decreased muscle strength and flexibility [11]. Pain has become increasingly common and an emergent complication of thalassemia. The exact mechanism of pain in thalassemia has not yet been clarified; however, iron overload, low hemoglobin level, and low bone mass have been suggested as possible causes [12]. Nevertheless, the current evidence does not confirm these suggestions. The most frequent sites of pain were the lower back (82%), the leg (56%), head (48%), and mid-back (47%). [13].

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Health Related Quality of Life studies on children with chronic illness such as thalassemia are limited. The assessment of HRQoL in children, especially in children with chronic illness such as thalassemia is particularly important [14].

1.1. Aim of the study

The aim of this study was to evaluate health related quality of life, muscle strength, and pain in children with β -thalassemia major.

1.2. Methods

1.2.1. Subjects

For the purpose of the study, 120 children ranged in age from 2 to 12 years were enrolled in a cross-sectional study from October 2017 to April 2018. 60 children with β -thalassemia major (32 girls and 28 boys) were selected by a blind physician from the outpatient clinic of “Abo El-Rish Pediatric Hospital – Cairo University Hospitals”. In addition, 60 healthy age-matched children with normal hemoglobin level (25 girls and 35 boys) were selected from Abo-Bakr El-Sedik Primary school in Sheikh Zayed City. All selected children were free from musculoskeletal deformities, visual and auditory impairments. However, children who had associated disease with β -thalassemia major as leukemia or marked cognitive or cardiac impairments were excluded from the study. The children either thalassaemic or normal were stratified into three age subgroups (I (2–4 years), II (5–7 years) and III (8–12 years)). Before the study, interviews were conducted with the children and their parents to explain the purpose, procedures as well as potential benefits of the study and signed a consent form of the ethical committee, Faculty of Physical therapy, Cairo University prior to participation. The study design, protocol and consent forms were carried out in accordance with the code of ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans.

1.2.2. Instrumentation for evaluation

1.2.2.1. Handgrip strength. The handheld dynamometer has been shown to be reliable in measuring hand-grip strength in children [15]. Hand-grip strength of the dominant side was measured in pounds using a hand-held dynamometer (Nicholas Manual Muscle Test system, Model 01163; Lafayette Instrument Company, Lafayette, IN).

1.2.2.2. Health related quality of life. Assessment of the HRQoL of children was performed using the pediatric quality of life inventory™ (PedsQL™) 4.0 generic core scale which was developed to measure HRQoL in healthy children and adolescent and those with acute and chronic health problems. PedsQL™ 4.0 is reliable, feasible, and valid to measure the health outcome of the pediatric population, as it facilitates risk assessment and tracking of community health [16]. The Arabic version of PedsQL™ 4.0 was considered to have satisfactory psychometric properties according to a Jordanian study conducted by Arabiat et al. [17]. A user agreement was signed with the MAPI Research institute, Lyon, France, prior to the use of the questionnaire. The PedsQL™ 4.0 generic core scale was designed to be appropriate for ages of the children. In children older than 4 years, it includes parallel child self-reports and parent proxy reports (age ranges 5–7, 8–12 and 13–18 years) and consists of 23 items. Moreover, in children aged between 2 and 4 years, parent proxy report was used and consists of 21 items. The scale was applied, and analyzed according to the PedsQL™ administration guidelinesSM. The children were asked to report problems regarding physical functioning (8 items), emotional functioning (5 items), social functioning (5 items) and school performance (5 items) that

have arisen for the past one month. The items' responses were measured on a five-point rating scale which consists of 0 (never a problem), 1 (almost never a problem), 2 (sometimes a problem), 3 (often a problem), and 4 (almost always a problem), yielding a total score with corresponding scores of 100, 75, 50, 25, and 0. The higher score indicating higher QoL, better health or a higher level of function. The PedsQL has demonstrated good internal consistency and validity in large samples of children with acute and chronic health conditions, as well as in healthy children and adolescents [14].

1.2.2.3. Pain. Pain was measured by using the Visual Analogue Scale (VAS), which represents quantitative measurement in terms of a straight line placed horizontally. Both ends of the line are labeled with descriptive terms to anchor the extremes of the scale and to give reference to any point in the continuum between them. The line was 10 cm in length and each cm has its own number that reflected the amount of pain. The VAS is the most common and reliable type of pain scale [18].

1.2.3. Procedures

1.2.3.1. Anthropometric measurements. Weight (kg) and height (m) were recorded using a calibrated floor scale (ZT-120 model), Hangzhou Tianheng Technology Co., Ltd. Hangzhou, China. Body mass index (BMI) was calculated as weight (kg)/height² (m).

1.2.3.2. Handgrip strength. The purpose and steps of the procedure were explained to the children and their caregivers. Each child was asked to indicate the dominant hand, any pain at one hand or both, or any kind of injury or surgery that could affect the result of the test. The children were seated in front of a table in an appropriately adjusted chair during measurements. The children sit with the shoulder adducted, the elbow flexed in a 90° angle, and the forearm and wrist in a neutral position. Before the start of each measurement, the child was asked to handle the dynamometer and when ready the child was told, “Squeeze your hand as hard as you can” then release. Three maximum voluntary contractions were recorded for the dominant hand with a rest period of 5 min. The first attempt is used for familiarization, and the score will be obtained by averaging the second and third attempts.

1.2.3.3. Health related quality of life. The PedsQL™ 4.0 generic core scale was explained to all children and their caregivers who were ensured of confidentiality of their information. In the present study, the questionnaires were distributed among the parents of children who filled out the questionnaires in the presence of the investigator. One questionnaire was given to each participant and the investigator completed the questionnaire with respect to ethical principles for those patients who were illiterate.

1.2.4. Statistical analysis

Descriptive statistics in the form of mean, standard deviation and frequencies were conducted for data presentation. *T*-test were conducted for comparison of the subject characteristics between groups. Chi-square test was conducted for comparison of sex distribution and school attendance between both groups. QoL, hand grip strength and VAS were compared between groups using *t* test. The level of significance for all statistical tests was set at $p < 0.05$. All statistical measures were performed through the statistical package for social studies (SPSS) version 19 for windows.

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