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Pamela Kohlbry,[1] Bashar al-Karmi[2] and Robert Yamashita[3]

[1]School of Nursing, California State University San Marcos, San Marcos, CA, United States of America. [2]Thalassemia Patients Friends Society, Ramalla, West Bank, Palestine. [3]California State University San Marcos, San Marcos, CA, United States of America. (Correspondence to Pamela Kohlbry: pkohlbry@gmail.com)

Abstract

Background: In countries with low resources, the health and quality-of-life of people living with thalassaemia can be severely affected.

Aims: This study examined the health-related quality-of-life of people living with thalassaemia in the West Bank and Gaza, Palestine.

Methods: This was a cross-sectional study of a convenience sample of 104 patients (71 adults and 33 children) who lived with thalassaemia and their families in 2015 in the West Bank and Gaza. Participants were surveyed using the 36-item Short Form Health Survey, version 2 (SF36v2), Pediatric Quality of Life InventoryTM (PedsQL) and PedsQL Family Impact Module to assess their quality-of-life. With the SF36v2, we used normed-based scoring and for the PedsQL and Family Impact Module, we used the 0–100 scoring. Scores are reported as means and standard deviations and P

Results: Quality-of-life scores were low across all domains, indicating poor quality-of-life. For bodily pain in the SF36v2, a significant difference was observed between the West Bank and Gaza. No significant differences were found between males and females. Data from the PedsQL showed no significant differences between the West Bank and Gaza. With the Family Impact Module, the summary score was higher among adults than among paediatric patients. Compared with other countries, thalassaemia patients in Palestine generally had lower quality-of-life scores in most domains.

Conclusion: The lack of access to healthcare and blood transfusions, and the geopolitical challenges may be responsible for the low quality-of-life scores of patients living with thalassaemia in Palestine.

Keywords: thalassaemia, quality-of-life, cross-sectional study, West Bank, Gaza.

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Introduction

Worldwide, genetic haemoglobinopathies such as thalassaemia are considered serious health burdens. In the United States of America, thalassaemia is rare and considered a disease of migration and it is managed as a chronic illness (1,2). However, in countries with less resources, the health burden of thalassaemia patients, which will also be a family burden, affects health-related quality-of-life (3,4). Thalassaemia is common in the Mediterranean, Middle East and sub-Saharan Africa (5). The prevalence of thalassaemia in Palestine which includes the Occupied West Bank and Gaza, is high; about 4% of the population are carriers (4). The political conflict and often violent conditions in Palestine complicate medical care of patients and their families who need to make regular clinic visits. Blood supply for transfusions is limited and chelation medication treatments are expensive or unavailable. Continuity of care is lacking, social and psychological support services are limited, and there are few healthcare workers with specialized training in the management of thalassaemia. The World Health Organization (WHO) lists Palestine as having the second-lowest health expenditure as a percentage of the Gross National Product among the Middle Eastern countries (6).

The quality-of-life and outcomes of people with thalassaemia vary depending on clinical care and other circumstances. In Palestine, thalassaemia patient-reported health outcomes are

poorly understood, and clinical research data are limited (7–10).

Research that measures quality-of-life can help identify the effect of chronic illness on patients' lives and allows comparison of different groups (11).

This study aimed to identify the quality-of-life of people with thalassaemia in the West Bank and Gaza. In 2009, we conducted a pilot study on the quality-of-life of patients with thalassaemia in Palestine (12). The study was designed to answer the question: could behavioural and social science research data be systematically collected in contested areas classified by WHO as a low-income country? Our study in 2015 is a follow-up to that pilot. During the time of this study, regional conflict was a challenge to conducting this research (13,14). This study also compares Palestinian data with other populations such as Turkey, Thailand, Italy, Greece and Middle Eastern countries.

Methods

Sample, instruments and data collection

Both the 2009 and 2015 studies were cross-sectional and used convenience samples recruited from regional clinics in Palestine. In 2009, only adults were part of the sample. In 2015 we expanded the sample to include children and their guardians. Data were collected on demographic characteristics of the sample. Three standardized quality-of-life tools were used: the 36-item Short Form Health Survey, version 2 (SF36v2), Pediatric Quality of Life InventoryTM (PedsQL) and PedsQL Family Impact Module (FIM).

Patients with thalassaemia and guardians of patients with thalassaemia who attended a transfusion clinic were approached to participate in the study. If they were interested they were provided with information and an informed consent form to sign. If they chose to participate and signed the consent form, they were given the surveys to complete.

The instruments used in the study measured a patient's quality-of-life and the impact the patient had on their family. The widely used SF36v2 instrument measures quality-of-life for young adults (ages ≥ 16 years) and older patients (15). The 36-item questionnaire measures 8 dimensions of general health-related quality-of-life: physical functioning, role physical (limitation because of physical health problems), bodily pain, general health, vitality, social functioning, role emotional (limitations due to emotional problems), and general mental health. Two summary scores assess physical and mental dimensions of health and well-being: physical component summary score and the mental component summary score. A high score indicates a more

favourable health state. The instrument has been used extensively in clinical trials and academic studies, across disease areas, including thalassaemia. Its validity and reliability to measure health-related functional status and well-being have been established (15). It has a high level of consistency across countries and the Arabic translation has been validated (16).

The PedsQL 4.0 generic core scales contain 23 items grouped into four scales: physical functioning, emotional functioning, social functioning and school functioning. It also includes composite scales for a total scale score (23 items) and a psychosocial health summary score which is the sum of emotional, social, and study/work functioning. A high score indicates a more favourable health state (17–19). All paediatric patients and their caregivers were surveyed using the PedsQL.

The FIM survey has 36 items and measures the effect of chronic paediatric health conditions on parents and the family. The survey measured six scales of parent self-reported functioning: physical functioning, emotional functioning, social functioning, cognitive functioning, communication and worry. It also measures two scales for parent-reported family functioning: daily activities and family relationships (20).

In 2009 we used the SF36v2. In 2015, we assessed paediatric health-related quality-of-life using the PedsQL. Because of the chronic nature of transfusion-dependent thalassaemia, we used the PedsQL FIM to assess the impact the disease had on caregivers.

Statistical analysis

The SF36v2 manual provides a standardized approach for statistically analysing the data. It allows a systematic approach for converting the individual data points into a meaningful component for a scale. The initial step converts the score into a 0–100 range with 0 (worst health) to 100 (best health) results. The SF36v2 designers recognized that the component nature of the scales and what they intended to measure were widely variable; the range between top and bottom scores varied substantially across health domains. Without understanding this, the data could be easily misinterpreted. Therefore, in order to simplify interpretation and make direct comparisons, a norm-based scoring approach can be used with the SF36v2, where each scale has the same average (50) and same standard deviation (SD) (11). This approach makes the health impact of a domain norm-based scoring score clearer and can more accurately reflect the impact of the disease. An individual score of