



Letter to the Editor

## Psychosocial burden in transfusion dependent *beta-thalassemia* patients and its impact on the quality of life and the problem of dignity



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### Dear Editor:

*Beta-thalassemia major* introduces a set of complex bioethical problems concerning patient dignity, including how it is related to psychosocial burden and impacts quality of life that have been inadequately addressed worldwide. This letter aims to prompt discussion about those issues and provides both concerns and recommendations.

*Beta-thalassemia major* is a blood disorder inherited by children from their parents. A genetic mutation inherited through the *Mendelian rule* is believed to be a factor that directly causes the disease. This condition causes severe chronic hemolytic anemia and patients require regular blood transfusions beginning early in life. In addition, this disorder

impacts patients through iron overload, which could impair their organs and bodily systems.

Unfortunately, *beta-thalassemia* epidemiological data are limited, but most patients with *beta-thalassemia* live in geographic “belt” areas that stretch from sub-Saharan Africa, the Middle East, South-East Asia, Northern Europe, the Mediterranean, and North America.<sup>1,2</sup> As a chronic disease, *beta-thalassemia* does not provide patients with many opportunities to get involved in any activity to develop themselves and shape their future fully. The prevalence of psychosocial maladjustment is 80% in *beta-thalassemia* patients, so they may experience many emotional, anxiety-related, and behavioral problems in their lifetime.<sup>3</sup> Burdens related to psychosocial maladjustment structure their lives on multiple levels.

For instance, at the social/environmental level, *beta-thalassemia major* is sometimes not easily disclosed by a person diagnosed because of the bad stigma. Kumar & colleagues (2019) found that the stigma felt by patients with *thalassemia* causes them to hold back their decisions in disclosing their disease status to people.<sup>4</sup> At the behavioral/psychological level, Naderi & colleagues (2012) found that patients with *thalassemia* experienced psychiatric disorders, anxiety disorders, depression disorders, and somatic symptoms.<sup>5</sup>

One cause of psychosocial burden for patients with *beta-thalassemia major* is the frequency of coming to the hospital for blood transfusions every 2–4 weeks, with the main burden being the chronicity of the disease.<sup>6</sup> Moreover, the biggest problem concerning patients with *beta-thalassemia major* is life expectancy. Patients with *beta-thalassemia major* often don't live past age 30 due to cardiac complications and iron overload.<sup>7,8</sup> Naturally, this

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contributes to the patient's psychological distress because of the future uncertainty prognosis of the disease. Similarly, 70.3% of dependent transfusion patients were worried about their life.<sup>9,10</sup>

Patients with chronic or terminal illnesses experience high psychological distress and symptoms that might include hopelessness or a reduced sense of dignity.<sup>11,12</sup> Consequently, a higher level of respect for dignity is required due to the nature and duration of the disease and is supported by the fact that it has a positive effect on medical outcomes.<sup>13</sup>

Human dignity is one of the essential concepts in chronic disease. Based on the literature evidence, this concept is currently being studied more often in chronic diseases with limited time between diagnosis and death due to known disease progression, such as cancer. In those cases, palliative and end-of-life care can be more easily planned. However, when referring to the sensitivity of the time, in the context of long-term chronic disease, patients with *beta-thalassemia major* also have the potential to experience problems with human dignity, its related psychosocial problems, and their quality of life.

Developing the theoretical framework for such a response, Nordenfelt distinguished dignity through four ideas that must be respected. These are that dignity 1) cannot be lost as long as the patient is alive; 2) is related to one's status in society; 3) is a moral stature linked to one's moral values and self-respect; and 4) is related to one's identity as a person and the integrity of body and mind.<sup>14,15</sup> It is essential to understand the psychosocial burdens and the concept of dignity because increased hopelessness, depression, and desire to hasten death are associated with the loss of dignity.<sup>16</sup>

Patients with *beta-thalassemia major* may have problems maintaining dignity while knowing that their quality of life will gradually decrease. Furthermore, the expectation of death will contribute to mental stress and negatively affect psychological quality of life. Disrespecting dignity also affects how patients see themselves from others' perspectives, particularly given the healthcare context in which patients receive transfusion care throughout their lives and thus enter permanently into the medical system. We see profound problems concerning patient dignity in their personal and social lives, as well as within the healthcare system.

In conclusion, patients with *beta-thalassemia major* might or might not be aware of their prognosis in terms of estimated life expectancy. Regardless of whether the patient's psychological state is affected by such awareness, overall, patients with *beta-thalassemia major* have burdens related their disease and it impacts their quality of life.<sup>17</sup> In the contexts of nursing and palliative or end-of-life care, we see that patients expecting death could be well-prepared and must be given a maximal chance to experience happiness, even while facing death. Despite the inevitable decrease in quality of life, all aspects of patient basic needs must be mapped and assessed to consider which parts could be supported to re-enforce human dignity. This would help patients to live their lives meaningfully and allow them to act in accord with their beliefs in their last precious moments.

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