EXPLORING PSYCHOLOGICAL RESPONSES TO THE STRESSOR OF MUSCULAR DYSTROPHY PATIENTS IN MALAYSIA: A QUALITATIVE STUDY

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Abstract

Muscular Dystrophy is a diverse group of acquired and inherited neuromuscular conditions that cause progressive muscle wasting and weakness, with varying degrees of pain and fatigue. In some cases, cardiac and respiratory complications are involved. As a result, there is a decline in mobility which leads to slowed walking, tripping and falls with some cases requiring walking aids or wheelchairs. The aim of this study is to explore the psychological responses to the stressor of Muscular Dystrophy patients. This study employs qualitative approach which used interviews for its data collection method. The participants involved in this study are one individual with Muscular Dystrophy disease, two parents with a Muscular Dystrophy child, one Muscular Dystrophy medical specialist, two physiotherapists with Muscular Dystrophy patients and one occupational therapist with Muscular Dystrophy patients. The responses to stressor brought up by the participants are shock and denial, anxiety, anger and guilt, and depression, demotivation, and low fighting spirit. Individuals with Muscular Dystrophy and their parents respond negatively to Muscular Dystrophy disease.

Keywords: Muscle disease, Muscular dystrophy, Psychological management

Introduction

Muscular Dystrophy is a general term for a number of hereditary, progressive degenerative diseases affecting skeletal muscles, and often other organ systems. This term includes many conditions associated to the muscle wasting and weakness where all are still genetic but having different types due to different genes and differ in severity (1). Muscular Dystrophy is one of the examples of neuromuscular disease.

Muscular Dystrophy is not one type of disease. Emery (2008) stated there are currently over 30 different types of Muscular Dystrophy have been recognized where some of these are very rare or only occur in certain parts of the world. Generally, there are nine main types of Muscular Dystrophy; Duchenne Muscular Dystrophy, Becker Muscular Dystrophy, Emery-Dreifuss Muscular Dystrophy, Limb-girdle Muscular Dystrophy, Facioscapulohumeral Muscular Dystrophy, Distal Muscular Dystrophy, Oculopharyngeal Muscular Dystrophy, Congenital Muscular Dystrophy, and Myotonic Dystrophy (1). The different muscular dystrophies are varied in who they affect and the symptoms of the specific type of Muscular Dystrophy (2).

Some of muscular dystrophies can appear at birth or during childhood, while others can only be detected later in life. In all types of Muscular Dystrophy, the disease will become worse as the patient's muscles grow weaker. According to the U.S. National Library of Medicine, National Institutes of Health, most patients with Muscular Dystrophy will eventually lose the ability to walk, either in a very early age or later in their life. Some individuals with Muscular Dystrophy have mild cases which worsen slowly, while other cases are more severe and disabling.

Currently, there is no cure for Muscular Dystrophy and the treatments for Muscular Dystrophy normally include physical therapy, speech therapy, orthopaedic devices,

surgery and medications (1, 2). Due to many disabling limitations and pain faced by the individuals with Muscular Dystrophy, this disease can cause medical and social problems among the individuals with Muscular Dystrophy. In addition, because it is a hereditary disease, it may recur in some families (3).

Besides having to face physical limitations, the individuals with Muscular Dystrophy also have to face some psychological setbacks (2-4). These setbacks are often left unattended by many medical practitioners and even the immediate family members of the patients (3-4). This psychological area of the individuals with Muscular Dystrophy will be the focal point of this study.

There are not many studies focussing on Muscular Dystrophy in Malaysia. The few available Muscular Dystrophy research in Malaysia mostly revolved around the medical (5) and genetic scienceaspects of it (6-9), very often neglecting the psychological and psychosocial sides of it. A literature search on psychological studies of Muscular Dystrophy disease in Malaysia only produce articles written by the researcher of this research (10, 11).

The availability of detailed or comprehensive or even concise studies on psychological management for the individuals with Muscular Dystrophy are very few globally, resulting lack of suitable references for this area of research. When the studies available, they were in general revolving around the quality-of-life research (12, 13) or psychosocial research (13, 14). Psychosocial research, if available, usually involving the studies on social adjustment, communication and peer relationship (13). The very few psychosocial studies involving psychological studies, if available, were usually only focussing on either anxiety or depression (13, 14). Due to this, in order to start psychological research on to Muscular Dystrophy, referencing will be quite a challenge.

Just like many other parts in the world (12, 13). There are many psychological issues need to be tackled when it comes to the management of Muscular Dystrophy disease in Malaysia, and various psychology studies related to Muscular Dystrophy need to be done in order to achieve this. Individuals with Muscular Dystrophy and their family members are at increased risk of depression and anxiety, particularly at major care transition points in the progression of the disease (3). Hence, there is a need of psychological intervention and emotional support for the individuals with Muscular Dystrophy, not just the physical and medical supports. Therefore, the aim of this study is to explore the psychological responses to the stressor caused by the disease.

Materials and Methods

The qualitative approach (narrative research) was used to explore experience and views of various stakeholders. Research questions and literatures contributed to the theoretical and conceptual frameworks, which helped to identify the general areas to probe in the interviews. These frameworks were further used as the basis of framing and building the interview guide. Ways of framing the questions have the effect of either opening up a discussion or closing down a discussion.

The aim of the interview was to understand the reality of Muscular Dystrophy in Malaysia and to explore the potential of psychological intervention in managing individuals with Muscular Dystrophy in Malaysia. Thus, a semi-structured interview by Mathieson and Stam (1995) was used and adapted as the basis of general structure of the interview. This interview has been adapted in the interviews of various individuals with cancer, individuals with AIDS or positive HIV, individuals with brain injury and many more. The questions were useful in capturing the sorts of stories which are essential to the disease experience. In order to allow the exploration of the participants' view, open-ended questions were included in every area.

The interview guide was categorized in four categories which were interview guide for the individuals with Muscular Dystrophy (13 open-ended questions), interview guide for the parents with Muscular Dystrophy children (13 open-ended questions), interview guide for the medical practitioners with Muscular Dystrophy patients (eight open-ended questions) and interview guide for the therapists with Muscular Dystrophy patients (seven open-ended questions). Even though the semi-structured interview guide by Mathieson and Stam (1995) was a well-known and established interview guide in Health Psychology field, further content validation was done by an external methodology expert from the International Islamic University of Malaysia (IIUM) (23).

Interviews were conducted face-to-face with various stakeholders which were an above 18 years old individual with Muscular Dystrophy, two parents to individuals with Muscular Dystrophy, a medical specialist (from Hospital Universiti Sains Malaysia, HUSM) who has the experience in handling Muscular Dystrophy cases, and two physiotherapists and one occupational therapist (also from HUSM) who have the experience in handling Muscular Dystrophy cases. For the record, the individual with Muscular Dystrophy chosen for this study did not have any cognitive impairment. There were five categories of the participants participated in this study which are individual(s) with Muscular Dystrophy, parent(s) with a Muscular Dystrophy child, Muscular Dystrophy medical specialist(s), physiotherapist(s) and occupational therapist(s) with Muscular Dystrophy patients.

Data saturation was already achieved after the interviews of the first participant from each category when there was no new theme appeared in the subsequent interviews. However, the purposive and snow-ball samplings were continued with all the available participants in order to support and strengthen the existing themes.

Interviews were transcribed and text-recorded verbatim, and anonymised before imported into NVivo which was used for storage and analysis of the interviews. In view of

the research questions, Narrative Analysis Approach as content analysis is regarded as the most fitting approach to analysis as narratives are powerful forms of giving meaning to experience. Thematic analysis was also used as the method of data analysis for the transcribed data.

Trustworthiness of the data was achieved through triangulation method which involve multiple different sources of data assembled from the research stakeholders. Comparison of interviews among the participants was made. Similar results arise from the analysis which prove the data validity.

Results

From the analysis, four responses to stressor were identified:

i. Shock and denial (including sadness and avoidance)
ii. Anxiety (including fear and helplessness as the basis of anxiety)
iii. Anger and guilt (including blaming others)
iv. Depression, demotivation, and low fighting spirit

The abbreviations are as follow:

i. In01: Individual with Muscular Dystrophy
ii. Pa01: First parent with a Muscular Dystrophy child.
iii. Pa01: Second parent with a Muscular Dystrophy child.
iv. Sp01: Muscular Dystrophy medical specialist.
v. PT01: First physiotherapist with Muscular Dystrophy patients.
vi. PT02: Second physiotherapist with Muscular Dystrophy patients.
vii. OT01: Occupational therapist with Muscular Dystrophy patients.

Response 1: Shock and denial (including sadness and avoidance)

Shock and denial were ones of the earliest responses by the participant with Muscular Dystrophy disease and parents with Muscular Dystrophy children when they were first introduced to Muscular Dystrophy disease.

The shock response by the individual with Muscular Dystrophy was due to unfamiliarity with the disease and its term. According to the individual with Muscular Dystrophy who participated in this study, it was the first time she heard about the disease, and the name of the disease was a totally unknown term to her. The shock response was shown through her confused and numb feeling. She could not focus on the explanation even though her doctor tried to explain about the disease. Therefore, she could not understand her condition properly when the diagnosis was first delivered.

"I was really young, a teenager. It was the first time in my life to hear the name of the disease. It was totally an alien term to me. I was confused, I felt numb. The doctor did explain a bit about the disease, but my mind couldn't focus on the explanation. So, I did not understand the condition properly when the diagnosis was first delivered".

(In01)

Avoidance is one of the signs of denial (14). The participant with Muscular Dystrophy disease further responded with avoidance. According to her, she ended up avoiding the matter. It was a gloomy period for her.

"Honestly, it was a gloomy period for me... so I pretty much avoided it".

(In01)

One of the parents interviewed in this study felt disappointed and was surrounded by sadness after receiving her child's diagnosis of Muscular Dystrophy disease. Shock response is normally accompanied by sadness (13)(14). This parent has a history of having a child with Muscular Dystrophy who has now deceased. When she first knew that her first child was born with Muscular Dystrophy disease, she had to accept it despite being surrounded by the feeling of sadness. When she knew she was pregnant with another boy, she kept praying that the same thing would not happen again but her second son was also born with Muscular Dystrophy disease. She felt sad and disappointed.

"When the first time I knew my first born had this disease, I had to accept… Only God knew how sad I was. When I was pregnant with another boy, I prayed night and day for the same thing that had happened to my first child wouldn't happen again, but God tested me again when he also was born with the same disease. Sad, disappointed, there were no words to express, but as time went by, I accepted it in the end".

(Pa01)

The family also could not accept the reality of having a family member with Muscular Dystrophy disease. The family of one parent with a Muscular Dystrophy child could not accept it because there was no history of Muscular Dystrophy in the family before. Due to this reason, they tried many healing methods, but nothing worked into their favour.

"At first, my family couldn't accept it because it never happened in the family before. Due to that, we tried so many healing methods, but nothing worked".

(Pa01)

Response 2: Anxiety (including fear and helplessness)

The participant with Muscular Dystrophy responded strongly with anxiety, fear and helplessness when dealing with Muscular Dystrophy disease. The individual with Muscular Dystrophy talked about how it was a distressing phase for her. She felt helpless and clueless for weeks after the diagnosis was delivered. This participant also expressed how she could not face the fear she was feeling.

"It was a pretty distressing time for me, I felt helpless and clueless for weeks after the diagnosis was delivered…. There was this feeling of fear I could not face…"

(In01)

Anxiety is a cognitive processing of fear (Porcelli, 2020). The individual with Muscular Dystrophy talked about how her fear and anxiety could soar every time she went outside. She would try to hide her disease as best as possible. She would try to act normal and pretend she was all right. She would avoid bringing up her disease as much as possible when she was in the public.

"I would try as hard as possible to hide my disease. The fear and anxiety were really high when I went outside. I tried to act normal, to pretend I was ok, I avoided any mention of my disease as much as possible when I was in public".

(In01)

Anxiety and fear can trigger the feelings of despair, hopelessness and helplessness (Porcelli, 2020). The participant with Muscular Dystrophy pointed out how she feels helpless from time to time and feels worried of the uncertainty of her future. She is always in fear, particularly when there is a progress in the disease.

"I'm always in fear, especially when there's a progress in the disease. I do feel helpless from time to time, worry of the uncertainty of my future".

(In01)

One parent who participated in this study said she always worried about the future of her child. She also felt helpless and did not know what to do because Muscular Dystrophy is unlike other curable diseases where if it were the case, then she would know how to try to heal the disease.

"I… always worried about my child's future".

"If it was like other diseases that could be cured, we would know what to try to heal it, but this… I felt helpless, didn't know what to do".

(Pa02)

Meanwhile, other parents (Pa01 talked on behalf of both of her husband and herself), who already experienced the death of a son due to Muscular Dystrophy disease, were afraid and worried that the same thing would happen again to their other son or daughters. The only thing they could do was constantly praying as they were informed that the disease has no cure.

"At first we were really afraid… We were worried that it would happen to our other son, but as Muslims we have to accept no matter what. I was also afraid that it might happen to our daughters. But what else could we do… We could only continue to pray, especially when we were informed that this disease has no cure".

(Pa01)

The individual with Muscular Dystrophy who participated this study also brought up how her parents were overwhelmed by the feeling of anxiety.

"That time… my parents were overwhelmed by their own feelings, the… anxiety".

(In01)

The parent who was interviewed in this study worried about the child's future also became helpless and did not know what action needed to be taken. She said she was probably confused during that time. She tried to continue her life as usual because she did not know what to do. She said she still had to continue with her life even if her heart was feeling restless and uneasy.

"I don't know how to say. Maybe I was confused at that time. I tried to continue life as usual because I didn't know what to do. My heart was restless, uneasy, but I still had to continue with my life".

(Pa02)

The sign of helplessness can be seen from the over-dependency of the individuals with Muscular Dystrophy on their parents, as stated by the occupational therapist participated in this study. According to the occupational therapist who participated in this study, there were individuals with Muscular Dystrophy who depended too much on the care of their parents. They did not follow the suggestion given by the therapists such as energy conservation.

"The patients depended too much on their parents' care. They didn't follow what was suggested even though we already taught them about energy conservation".

(OT01)

Response 3: Anger and guilt (including blaming others)

One of the parents participated in this study and the parent of the participant with Muscular Dystrophy disease responded strongly with guilt and helplessness when dealing with Muscular Dystrophy issues. According to the individual with Muscular Dystrophy, her parents were overwhelmed by the feeling of guilt.

"...my parents were overwhelmed by their own feelings, the guilt..."

(In01)

Meanwhile, the above mentioned participated parent opened up how she felt very guilty and always wondered if the disease came from her as the doctor informed her that Muscular Dystrophy is a type of genetic disease.

"The doctor said this is a genetic disease. I always wondered if it came from me. I felt so guilty..."

(Pa02)

The same parent said it was the hardest when there was nothing that could be done to help her Muscular Dystrophy child. She did not know what to do. The only thing she could do was to follow the doctor's and other medical practitioners' suggestions.

"The hardest would be when I could do nothing to heal her. When it was hard for her, or when she fell, or when she was sick, I couldn't replace her. I just didn't know what to do, only followed the doctor's and the hospital staff's

suggestions".

(Pa02)

Muscular Dystrophy patients seemed to show the sign of anger and helplessness when they had to keep explaining about the disease to other people or when there was no noticeable positive progress from their therapies. The participant with Muscular Dystrophy expressed that she has the tendency to miss consultation and therapy appointments from early on until now because there was no improvement in her physical condition.

"At the early stage, I did miss appointments. Still do, here and there along the way, too. The therapies as well. Because there was no change in term of physical improvement, I missed some therapy sessions".

(In01)

She also mentioned how hard it is for her to conform to the norm and to keep explaining to everyone around. She needs to be mentally and emotionally strong in order to go through that. According to her as well, immersing with the normal people requires hard work because it is not as easy as it has always been said.

"To keep explaining to the people around. It causes this tug in my heart every time I have to do it. And… To conform to the norm. It requires a very hard work in order to do that. I need to be strong mentally and emotionally in order to go through that. To immerse with normal people is not as easy as said".

(In01)

Response 4: Depression, demotivation, and low fighting spirit

The participant with Muscular Dystrophy disease highlighted the recurring depression and the consequent low fighting spirit. She revealed how she feels depressed from time to time. Although it is not serious with little frequency, it is a recurring occurrence. Due to the depression and her tendency to keep thinking about the uncertainty of her future, her will to fight has also continuously weakened.

"I can feel depressed from time to time. Well, it's not always but it's recurring, even though it's not serious. Because of the depression and I keep thinking about my unpromising future, my will to fight also can be low at times… it's on going".

(In01)

The participated occupational therapist and one of the participated physiotherapists also put emphasis on their patients' low fighting spirit and de-motivation due to the lack of positive progress. As described by a physiotherapist who participated in this study, there were patients who would just give up and would stop coming because their condition kept deteriorating even though they already followed through with the therapies.

"There were those who just gave up, because of the repeated blood tests every time they came. And the deterioration of their disease despite doing therapies as there's no cure for this disease. So, they ended up not coming at all. Therefore, nothing much can be talked about them".

(PT02)

According to the occupational therapist, when the patients became dispirited, they would not be present at the therapy sessions regularly and after a while, they would stop going to the therapy sessions altogether.

"When they were dispirited, sometimes they came, sometimes they didn't come. After that, they didn't come at all".

(OT01)

Low fighting spirit and de-motivation could become worse when the Muscular Dystrophy patients became too dependent on their parents. The occupational therapist highlighted how the patients will be demotivated when they feel tired. They will refuse to do the planned activities. It will become harder when the parents pamper the patients and ask their children to take the time out, especially if the patient is their only child.

Sometimes, the patients will be demotivated when they're tired, refuse to do the activities, added by the parents who always pamper them, especially if the patient is their only child. Sometimes, it's the parents who ask their child to take a break.

(OT01)

The occupational therapist also mentioned the visible stress and low fighting spirit in the parents. The parents were in stress and looked tired with no fighting spirit. Despite the fact that they did care for their children, they still could become dispirited.

There were parents who were in stress. They looked tired, with no fighting spirit. They do care for their children, but they were also dispirited.

(OT01)

The stressors are summarised as in Figure 1.

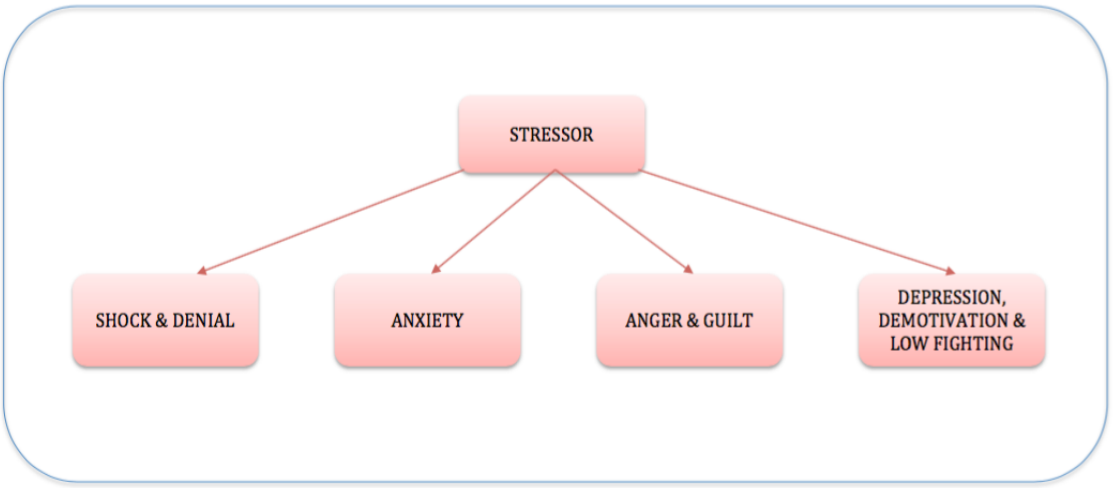

Figure 1: Responses to stressor (with the stressor being Muscular Dystrophy disease)

Discussion

Shock and denial were the earliest responses in general from the participants who have to directly deal with Muscular Dystrophy disease. Meanwhile, anxiety was the response more frequently brought up by the participants compared to the other responses. Anger and guilt, as well as depression, demotivation and low fighting spirit, while expressed a bit less by the participants are still crucial responses as they could affect the course of the therapies.

The participant with Muscular Dystrophy displayed both shock and denial responses through the exhibition of numb feeling and avoidance reaction. One of the parents with a Muscular Dystrophy child also had problems with acceptance despite before this already having a history of raising a child with Muscular Dystrophy who has now deceased. On top of having history with two Muscular Dystrophy children, it was more unfortunate when the family of this parent also could not accept the reality of having a family member with Muscular Dystrophy as this occurrence never happened in their family before.

Throughout the whole interviewing process with the participants personally dealing with Muscular Dystrophy disease, namely the individual with Muscular Dystrophy and the parents to children with Muscular Dystrophy, these participants kept repeating the theme of anxiety as the response to Muscular Dystrophy disease. This finding supports the study by Mori-Yoshimura et al. (2019) which stated that the participants who are individuals with Becker Muscular Dystrophy exhibited a high level of anxiety (24).

The individual with Muscular Dystrophy brought up fear and helplessness multiple times all through the interview, and also mentioned distress, anxiety and worry. All these feelings can be clustered together under the umbrella of anxiety response. According to the systematic review by von der Lippe et al. (2017), anxiety is normally triggered by uncertainty and lack of knowledge about the rare condition (15-18). Birnkrant et al. (2018) recommended Generalized Anxiety Disorder 7-item scale as a tool to screen anxiety at each neuromuscular clinic visit (3, 19, 20).

Both parents with Muscular Dystrophy children clearly presented their anxiety response through the mention of worries, with one parent exhibited anxiety response further with the expression of fear and the other parent exhibited anxiety further through the mentions of helplessness, restlessness and uneasiness. The individual with Muscular Dystrophy also mentioned how her parents were overwhelmed with anxiety. One of the participants who is an occupational therapist stated how her patients were having over-dependency on their parents. This subtle sign of helplessness seemed to affect the progress of their therapies when they refused to follow through the suggested course of therapy such as energy conservation.

Substantially, the parents with Muscular Dystrophy children responded with guilt meanwhile the individual with Muscular Dystrophy responded with anger. One of the participated parents had guilty thinking about how the disease could come from her as Muscular Dystrophy is a genetic disease. The other participating parent showed the sign of guilt when she expressed how she could do nothing to help her Muscular Dystrophy child. The individual with Muscular Dystrophy who participated in this study also mentioned how her parents were overwhelmed by the feeling of guilt. On a personal level, this participant with Muscular Dystrophy showed the sign of anger as a result of having no physical condition improvement, having to keep explaining the condition to everyone around and having to immerse with the normal people. According to this participant, she ended up missing consultation and therapy sessions. The almost same observations were found in the systematic review by von der Lippe et al. (2017), where ones of the aspects of emotional distress were feelings of fear, anger, blame and loss and guilt feeling was normally related to the risk of passing the condition on to children (15, 20).

Depression, demotivation and low fighting spirit could affect not only the mental and emotional health of the people who have to deal with Muscular Dystrophy disease, it could also affect the course and progress of the therapies of individuals with Muscular Dystrophy. This is in line with the study done by O' Dowd et al. (2021) which explained that depressive symptoms have the greatest association with quality of life in the mental health domain (21). Birnkrant et al. (2018) recommended Patient Health Questionnaire 9-item depression scale as a tool to screen depression at each neuromuscular clinic visit (3).

The participant with Muscular Dystrophy felt depressed from time to time. Even though with low frequency, the feeling is recurring. She also mentioned having a low fighting spirit. One of the physiotherapists and the occupational therapist who participated in this study talked about the demotivation shown by their Muscular Dystrophy patients. According to the physiotherapist, repeated blood tests and deterioration of the disease despite doing therapies caused the patients to become demotivated and eventually stop coming for the therapies altogether. The occupational therapist talked about how the patients refused to do activities when they became tired. Sharing the same experience as the physiotherapist, the occupational therapist also stated how the patients became dispirited and would miss their therapies regularly, and then eventually stop going to the therapies altogether. This occupational therapist also brought up how even the parents could become dispirited and look tired with low fighting spirit.

Conclusion

This research has identified how the individuals with Muscular Dystrophy and their parents respond negatively to this disease, and how these responses could affect the condition of cognitive and affective domains of the individuals with Muscular Dystrophy. The responses to Muscular Dystrophy disease can be categorized as shock and denial, anxiety, anger and guilt, and depression, demotivation and low fighting spirit. This psychological impact of Muscular Dystrophy disease denotes the potential of psychological management in the area of Muscular Dystrophy disease.

This study will benefit not just the Muscular Dystrophy community in Malaysia but also other Muscular Dystrophy communities around the world. Before this research, there was no proper psychological research in Malaysia, either on Muscular Dystrophy disease or other rare diseases, as mentioned in Rare Disease in Malaysia whitepaper (4). According to this whitepaper, there are no published reports of the psychosocial impact of rare diseases in Malaysia, and this needs to be addressed urgently by the local healthcare professionals and public health experts in the country.

This psychological study related to Muscular Dystrophy disease was novel research in Malaysia. It was a needed study (5) which is only a start for hopefully many more studies by many other researchers in the future. The data from this study can be improved by repeating the same study at other research sites. A systematic review hopefully can be produced many years in the future when more studies at various research sites are done.

Conflict of Interest

All authors declare no conflicts of interest.

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