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The Psychological Condition of the Parents of Children Suffering from *Spinal Muscular Atrophy*

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Abstract -- *Fatigue during the treatment period results in a negative response when taking care of their disabled child. A change in the behavior of the parents has an impact on their physical and mental health. This study aims to explore the experience and psychological condition of the parents with children suffering from Spinal Muscular Atrophy. This study was qualitative research with a phenomenological study approach. The participants totaled as many as 23 families selected through purposive sampling. Collecting the data was done through in-depth interviews analyzed thematically using Colaizzi's steps. The results show that the experience of the parents of children suffering from Spinal Muscular Atrophy consists of 17 themes formed of 5 categories and 33 sub-themes. The parents' experience related to their children suffering from Spinal Muscular Atrophy can be described referencing the following: 1) The parent's understanding of the childhood illness, 2) Parental burden, 3) The influence of illness on the family, 4) Self-obstacles, 5) Infrastructure barriers, 6) Grieving, 7) Sources of support, 8) Type of support, 9) Division of roles, 10) Coping mechanisms, 11) Skill improvements, 12) Efforts when looking for help, 13) The development of the children suffering from Spinal Muscular Atrophy, 14) Co-morbidities in the children suffering from Spinal Muscular Atrophy, 15) Substitute caregivers, 16) Acceptance and 17) Policy. The experience of the parents with children suffering from Spinal Muscular Atrophy in Indonesia is one that requires support from within themselves, from within their families and socially to reduce the burden and obstacles felt by the parents. It is expected that health care agencies and the parents can provide appropriate care for the children suffering from Spinal Muscular Atrophy.*

Keywords-- *Spinal Muscular Atrophy; Parent Experience; Child; Qualitative Research*

I. INTRODUCTION

There are five interactions between parents and their children, namely symbolic interactions, physical interactions, psychological interactions, ideological intellectual interactions and ethical moral interactions. Parents who are not experienced at applying these interactions will influence the development of their children's health, their adherence to care and they will not understand how to respond to the effects of the disease. When there is a long-term feeling of grief, the family members feel physically and mentally tired. They may reduce their affection for their child suffering from Spinal Muscular Atrophy[1],[2].

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Every change in emotional response and behavior shown by the parents will affect the treatment of the parents when taking care of their children. Fatigue during the treatment period results in a negative response when taking care of their child. Changes in the parents' behavior will have an impact on their physical and mental health. Parental burden is a multidimensional response to physical stressors (fatigue and sleep disorders), psychological stressors (anxious, worried, pessimistic and depressed), social stressors (limitations related to society) and financial-related stressors concerning the parents' experience when caring for their children [3].

The parents will feel stressed when taking care of their child. They will find it difficult to understand the feelings and condition that the child is experiencing. The ignorance of the needs and social life of the child also affects the psychological and physical aspects of the parents, especially the mothers. Most of the stressful events occurred among those aged 25 to 35 years old at a percentage of 48.4%[4]. Furthermore, the parents of children suffering from *Spinal Muscular Atrophy* experience grief and many aspects of this type of care affect the lives of their children and their self-identification. The parents experience internal conflicts centered on the stress of caring for their critically-ill children[5].

It was found that the parents experienced distress when their children became disabled. The parents felt helpless, depressed and in a critical situation. When the death of their child suffering from *Spinal Muscular Atrophy* happened, the parents felt that there was a loss of resources, poor social support, and decreased quality of life[6]. The parents seek psychological support, family harmony, and supportive care to make it easier when taking care of their children suffering from *Spinal Muscular Atrophy*[7, 8]. After parents were told about the diagnosis of *Spinal Muscular Atrophy*, they had to live with the deteriorating health condition of their child. They experience feelings of grief that affect family interactions and family functions[1].This study aimed to explore the experience and psychological condition of parents with the children suffering *Spinal Muscular Atrophy.*

II. METHODS

A qualitative method with a phenomenology study approach was used. The period of research was November 2017 to January 2018. The population consisted of families taking care of children suffering from *Spinal Muscular Atrophy* types 1 to 3. This population involved the *Spinal Muscular Atrophy* Indonesian community. The sample was obtained through purposive sampling. The sample size totaled 23 families.

The interview guidelines were developed by the researcher alongside the topic of the research so then the questions given did not came solely from of the research objectives [9]. Prior to the data collection, the researchers carried out interview guidance trial tests on two participants to validate the questions. The interviews could be carried out via telephone (telephone interviews), through social media or through CAPI (Computer-Assisted Personal Interviewing). The interview process was conducted until no new themes appear. The researchers used stationery and a recorder as the data collection tools.

The data analysis was done using the Colaizzi[10] method, namely (1) making verbatim notes; (2) all verbatim-transcripts read repeatedly to identify any particular meanings in the words or sentences and providing a code based on the similarities and differences; (3) explaining the meaning of significant sentences when collecting and organizing the formulation of the categories; (4) constructing the descriptions into themes; (5) creating the validation themes; (6) the categories and themes being extracted from the main idea of the statement and the sentence and (7) conducting a simultaneous data analysis process until data saturation occurred. This study received a certificate of ethical clearance from Ethical Commission No: 1185-KEPK on 26th November 2018.

III. RESULTS

In this study, there were 23 participants. They were mostly 26-35 years old totaling 12 participants (52%) and 22 of the participants were female (96%). There were 12 Javanese participants (52%). Those living in West Java totaled 15 participants (66%) while there were 12 Muslims (79%). For education, 11 had been to high school (47%), 18 were married (79%) and 13 were housewives (57%) (Table 1). This study produced 5 categories, 17 themes and 33 sub-themes (Table 2) which consist of the following:

Table 1. Participant frequency distribution (n = 23)

Characteristic	∑	%
Age (year)		
17 – 25	1	4
26 – 35	12	52
36 – 45	9	40
46 – 55	0	0
56 – 65	1	4
Sex		
Male	1	4
Female	22	96
Ethnicity		
Javanese	12	52
Suda	7	31
Padang	1	4
Chinese	3	13
Areas		
East Java	3	13
Middle Java	4	17
West Java	15	66
Bali	1	4
Religion		
Islam	18	79
Christian	4	17
Buddhist	1	4
Education		
High School	11	47
Vocational	3	13
Bachelor‘s	9	40
Marital Status		
Married	18	79
Divorced	3	13
Widowed	2	8
Occupation		
Housewife	13	57
Entrepreneur	5	22
Private	1	4
Teacher	1	4
Civil Servant	3	13

Table 2: Analysis of the Overall Themes

Categories	Theme	Sub-Theme
Understanding of the parents	Parents' understanding of the childhood illness	
Psychological experience: physical stressors, psychological stressors, social stressors and financial stressors	Load of parents	Physical stressor
		Psychological stressors
		Social stressors
		Financial stressors
	Influence in the family	
	Self-obstacles	Emotional obstacles
		Communication obstacles
		Obstacles to meeting ADL
	Infrastructure barriers	Barriers to getting to the health facilities
		Educational barriers
	Grieving	
Support system	Source of Support	Self support
		Family
		Social
		Health facility officer
	Type of support	Emotional
		Instrumental
		Friendship
		Information
The act of overcoming obstacles	The division of roles	The role of domestic work

		The role of looking for finances
		Caregiver's role
	Coping mechanism	Adaptive
	Skill improvement	Increased compliance with ADL
		Improved communication
	Efforts when looking for help	Looking for information
Hope	Development	Skills
		Health Status
	Co-morbid disease	
	Substitute caregiver	Circle Family
		Big Family
		Stranger or Social
	Acceptance	Family
		Social
	Policy	Health facility officer
		Government

Category 1: Understanding of the parents with children suffering from *Spinal Muscular Atrophy*
Theme 1: The parents' understanding of the childhood illness

Category 2: Psychological experience refers to the physical stressors, psychological stressors, social stressors and parental financial stressors when taking care of a child suffering from *Spinal Muscular Atrophy*. Five themes were found.
Theme 2: Parental Burden

1. Physical stress

Some of the participants experience fatigue and sleep disorders because they have to care for and look after the child 24 hours a day.

2. Psychological stressors

Some of the participants feel the psychological stressors all of the time because they are worried, pessimistic about their inability to care for their child properly and because they worry about the suffering of the children.

3. Time load

Some of the participants must always be around the child to care for and accompany the children's activities because of their limited movements.

4. Social Stressor
5. Financial stressors

Some of the participants felt helpless when it came to getting money for their living expenses and medical expenses. The child care costs were not small and sustainable.

Theme 3: Influence in the Family

The psychological experience of the participants included changes in behavior, resigning from work, or trying to find another job nearer to home.

Theme 4: Self-obstacles

1. Emotional Barriers

All of the participants were angry and felt exhausted while caring for the child suffering from Spinal Muscular Atrophy.

2. Communication Barriers

Some of the participants experienced barriers in their verbal and non-verbal communication with the children suffering from *Spinal Muscular Atrophy.*

3. Obstacles to Fulfilling ADL

Some of the participants experienced obstacles related to the fulfillment of ADL for their children such as their bathing, bowel movements, urinary movements, eating, moving and walking activities.

Theme 5: Barriers in the Infrastructure

1. Barriers to the Health Facilities

Medical devices and special therapies are less accessible to some of the participants, including medicine for Spinal Muscular Atrophy. Meanwhile, the participants were disappointed when the health facility officials did not know about the initial symptoms and when they did not enforce the initial diagnosis of the disease.

2. Educational Obstacles

The participants experienced educational barriers in the form of their children suffering from *Spinal Muscular Atrophy* not being accepted into public schools, in addition to their distance from the school and the facilities not being supportive.

Theme 6: Grieving

The grieving stage was experienced by all of the participants before their children were diagnosed. The stage of denial and depression is the first thing that the participants feel. Some of the participants were still unable to accept the illness experienced by their child. The bidding stage was described by several participants.

Category 3: The support systems are divided into two themes, namely the origin of support and the type of support.

Themes 7: Sources of Support

1. Self-support

The participants motivate themselves to remain patient, strong, and sincere when caring for their child suffering from *Spinal Muscular Atrophy* in Indonesia.

2. Family

The participants got support and motivation from their nuclear family and extended family.

3. Social

The participants get support from their social circle such as their friends, the community and people who are not known to Paris.

4. Health Facility Officer

Some of the participants received support from the health facility staff.

Theme 8. Type of Support

1. Emotional support

The participants get emotional support in the form of empathy, caring, and attention from their family, health care workers, friends, the community and strangers.

2. Instrumental support

The participants get financial support, medical devices and assignments from various sources by working hard on their own and from their husband, family, friends, community and strangers.

3. Friendship support

The participants get support for their feelings of solidarity from various sources such as their husband, family, friends, community and strangers.

4. Information support

The participants receive information-based support through communication networks including in the provision of advice and direction. The information comes from various sources such as their spouse, family, friends, communities, health care workers and strangers.

Category 4: The act of overcoming obstacles is carried out by the participants as an effort to cure their children of *Spinal Muscular Atrophy*. The act of overcoming obstacles is divided into 4 themes, namely:

Theme 9: Distribution of roles

1. The role of domestic work

Some of the domestic work is done by the participants themselves or they are assisted by their husbands, children who are healthy participants, other family members or ART.

2. The role of seeking financial help

Some of the participants tried to find their own finances with their husbands' help or assisted by their family, friends, communities and unknown people.

3. Caregiver role

The participants are either a single caregiver or they are assisted by their husband, family or ART, in addition to health care workers.

Theme 10: Coping mechanism

Coping mechanisms are one of the methods used by the participants when dealing with problems while caring for their children. Coping mechanisms can be adaptive in the form of sincerity and resignation. All of the participants are sincere and resigned as a carer.

Theme 11: Skill Enhancement

1. Increased Fulfillment of ADL

Some of the participants increased the fulfillment of the ADL of the children suffering from Spinal Muscular Atrophy by teaching the children to move continuously so then their muscles do not get stiff. This is so as to not accelerate the decrease in muscle strength.

2. Improved Communication

Improved communication as part of a participant's effort to communicate with their children makes it easier to do verbally and through signaling

Theme 12: Efforts when looking for help

Some of the participants seek information through the health facilities and social media.

Category 5: Expectations of the parents of the children suffering from *Spinal Muscular Atrophy*

Theme 13: Development

1. Skills

Some of the participants hope to be more skilled when caring, communicative and patient when confronting their children.

2. Health status

The health status of the children has the possibility of decline. All of the participants expect an increase in immunity and for the psychological health of the children suffering from *Spinal Muscular Atrophy* to always be good. All of the participants hope that their children can be gradually more independent in the future.

Theme 14: Co-morbidities

Some of the participants hope that their children will not experience respiratory problems, metabolic disorders, gastrointestinal disorders and impaired mobilization. If they already are experiencing the aforementioned due to the disease, then they hope that they can be treated appropriately.

Theme 15: Substitute Caregiver

1. Core Family

Some of the participants hope that their other children can help them when caring for the suffering children.

2. Big family

Some of the participants hope that if they have a large family, then there are others willing to care for their children.

3. Other or Social People

Some of the participants hope that their friends and other strangers are willing to help them to take care of the suffering children.

Theme 16: Acceptance

1. Family

The participants hope that the core and extended family can accept the condition of the child while providing assistance and support.

2. Social

The participants hope that their friends, neighbors and the social environment (everyone) can accept their child warmly.

Theme 17: Policy

1. Health Facility Officer

The participants hope that the health facility staff can provide appropriate treatment and care according to the condition of the child. The participants expect assistance in the form of information and finances to support the child’s optimal maintenance and they also expect easy access to medical treatment at any time.

2. Government

The participants expect attention and assistance from the government for the children suffering from *Spinal Muscular Atrophy* in the form of Spinranza’s legalization to facilitate ease of access.

IV. DISCUSSION

Most of the participants understand how their child has been diagnosed with *Spinal Muscular Atrophy*. The parents who have gained knowledge about the disease and how to handle it will take good care of their child. They will be aware of the preventive and/or treatment measures that will prevent the children from suffering from any negative impacts [11, 12].

Every change in the emotional response and behavior shown by the parents influences the treatment and care of their child. Fatigue during the treatment period has a negative impact during child care. Changes in the parents' behavior will have an impact on their physical and mental health [13, 14]. In addition, families reflect on distress because they have to make time-sensitive decisions to extend the life of the medical interventions without knowing what the long-term outcomes are, including the cost borne by the child in terms of the pain from the repeated care [15, 16].

The participants experienced emotional obstacles in the form of anger and fatigue when taking care of a child with *Spinal Muscular Atrophy*. Other participants experienced communication barriers with the children due to muscle weakness in the child's oral cavity and if a tracheotomy had been done. The muscle weakness in *Spinal Muscular Atrophy* sufferers is a barrier to ADL fulfillment. Most of their activities were assisted such as eating, bathing, bowel movements, urinary movements and mobilization. The considerable psychosocial impact of this condition begins with the difficulty of the diagnostic process and the view of how newborn screening can improve the process and show the important unmet needs in the presence of early diagnosis and effective treatment [13].

The participants feel disappointed and desperate when the health care officers do not know about the initial symptoms and do not enforce the early diagnosis of the disease. The parents whose children have been diagnosed with Spinal Muscular Atrophy Type 1 expressed concern and dissatisfaction about the lack of coordination in the health care setting and they suggested that there should be a care coordinator to provide family support [15, 17]. Previous research has shown that professionals who lack knowledge about rare diseases can withdraw from situations that they do not directly know about [18]. The lack of knowledge about this disorder has been highlighted in this study and it has also been shown previously[5, 13, 17, 19, 20]. Thus, the parents develop a level of expertise when managing their child's condition [21]. They wanted the health facility staff to increase their special knowledge of *Spinal Muscular Atrophy* disease in order to be more proactive.

Children suffering from *Spinal Muscular Atrophy* experience obstacles related to their education. As some of the participants revealed, children suffering *Spinal Muscular Atrophy* were not accepted in public schools near to their homes. Inclusive schools are still not comprehensive. Inclusive education itself is an education service system that provides opportunities for all children to learn together in public schools by paying attention to diversity and individual needs. This is so then the potential of children can develop optimally[22].

The grieving stage experienced by all of the participants began before the child was diagnosed and ran up until after the child was diagnosed with *Spinal Muscular Atrophy*. Some of the participants were not able to accept the pediatric disease diagnosis. The participants closed themselves off to finding out about their child's disease due to a lack of information from the doctor. Most of the participants understood the child's illness with feelings of acceptance and sincerity, as if it was God's plan. Once the families have accepted the fact that their child is inevitably going to die, the family grief must be recognized and information related to the palliative care process must be provided[5]. Some of the parents expressed guilt when they reflected on how the disease destroyed their hopes and aspirations for their children[13].

Most of the participants got support that was both external and internal in nature. Support from their extended family and/or the external environment results in a strength that results in confidence when providing care for children with disabilities. This is as well as the social support provided. Social support from the families and professional institutions has made it easy for the caregivers to care for both the patients and themselves, to deal with stress and to minimize their burden. Thus more social support from the health care professionals is needed to help minimize the caregivers' burden, such as by providing appropriate information and support from the medical staff and strengthening the home care services available for the caregivers to utilize [23]. The social support provided to the children with disabilities includes emotional support, information or materials provided by the tools used[24]. The role of the family is needed when there is the emergence of health problems in other family members [25].

Family-centered care could be an ideal approach for children with type 1 *Spinal Muscular Atrophy*. The family might make medical decisions regarding palliative care. The management of symptoms determines the direction of the children’s life. Important decisions regarding ventilation, airway cleaning and nutrition [26] affect the treatment course that extends life or that allows for a natural disease end to develop with palliative care following. The families involved in managing rare or long-term chronic diseases become experts in this field when it comes to understanding and managing their child's condition. They reported that it was important for them to feel that they were working with the medical health team regarding their child and any decisions that needed to be made [28].

Coping mechanisms are one of the methods used by the participants when dealing with the problems encountered while caring for the children suffering from *Spinal Muscular Atrophy*. All of the participants were sincere and surrendered to God. This is because the participants were confident and believed that God would always enable the participants to care for their children. When the participants took care of their children from suffering *Spinal Muscular Atrophy*, they assumed that it was a rewarding field. This expression was in line with the research that stated that caregivers respond in various ways to their disabled child. Some saw the presence of their child as a blessing or gift from God[14].

Our results show that the families developed complex coping mechanisms including multilevel medical care. Further family characteristics need attention in an era of potential future treatment choices [29]. Parents need an adaptive coping mechanism as a support when they are caring for their children. The more that the parents surrendered and were sincere towards following God's destiny, the better they were when caring for their child. The children suffering from *Spinal Muscular Atrophy* felt that their parents cared about them and this motivated their children's health.

Most of the participants taught their children to move continuously so then their muscles did not stiffen. The participants put on a brace and AFO (Ankle Foot Orthosis) as a movement aid. The mobilization aids used include both manual and electric (specifically) wheelchairs. Some of the participants were taught different pronunciations of certain letters due to the weakness of their oral cavity muscles. There was tension between wanting the children to finally become independent and the amount of support needed to make it happen[13, 30, 31].

Some of the participants hoped that the children did not experience respiratory problems, metabolic disorders, gastrointestinal disorders and impaired mobilization. Gastrointestinal complications often occur in individuals with Spinal Muscular Atrophy and it is not clear whether this is due to their immobility and nutritional deficiencies or whether there is another primary defect (gastrointestinal mobility) [32]. Moreover, secondary malnutrition due to the decreased oral intake could also be a dangerous problem for some of the children and adolescents suffering from type 2 *Spinal Muscular Atrophy*[33].

The policy result was that all of the participants were expected to gain the ability to support the health and intelligence of their children. Most of the parents reported believing that the health care professionals did everything possible for their children[34]. Maintaining good quality care for the children suffering from *Spinal Muscular Atrophy* required continuity in care and a coordination of resources [35]. This research implies that this can be achieved by obtaining a balance between the parents and the health care professionals when it comes to being proactive and taking the initiative to manage the special care of children suffering from *Spinal Muscular Atrophy* [34].

V. CONCLUSION

The parents of the children suffering from *Spinal Muscular Atrophy* in Indonesia require support from within themselves, within their family and socially to reduce the burdens and obstacles. It is expected that the health care agencies and parents together can provide appropriate care for the children suffering from *Spinal Muscular Atrophy*.

CONFLICT OF INTEREST

The authors would like to declare that there are no conflicts of interest.

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