

The invisible part of the iceberg: qualitative aspects of childhood vasculitis

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ABSTRACT

Objective. The aim of this study was to develop a multidimensional assessment instrument named "Juvenile Vasculitis Multidimensional Assessment Report" (J-VAMAR) to measure all the domains of the vasculitis. In this qualitative study, it is primarily aimed to enrich the item generation for the J-VAMAR.

Methods. Twelve children with vasculitis and their mothers (n=12) were enrolled in this study. The data were collected using both a demographic data form and a semi-structured interview form. The study was performed on individual patient face-to-face interview. Data were analysed by grounded theory and the N Vivo 9 software program.

Results. Four categories were obtained. These categories were (i) physical effects of the illness, (ii) emotional effects of the illness, (iii) social effects of the illness and (iv) experienced challenges related to treatment process. In the physical effect category severe pain, physical limitations, weakness and fatigue; in emotional effect category thought of death, hopelessness and dissatisfaction about body image; in the social effects category decrease in academic performance, absenteeism to school and concealing the sickness from friends were the most common features. In the fourth category, subjects complained of lifelong drug use and frequency of daily drug consumptions.

Conclusion. These results provide evidence-based data for the assessment of children with vasculitis by several domains including physical, emotional and social aspects as well as treatment protocols. The study provides the basis and/or justification for selecting the

domains that the developing multidimensional instrument should include.

Introduction

Primary systemic vasculitis in childhood is relatively rare, but can have a significant morbidity and mortality (1-3). Vasculitis creates some challenges physically (e.g. abdominal pain, fever, skin ulcers or infarctions, myalgia), psychologically and socially (e.g. school absenteeism, physical appearance) (4). Biological, psychological and social contexts are equally important dimensions and all dimensions play equally important roles in determining and affecting patient's behaviour in health and disease (5, 6). Patients' subjective experiences may represent key domains of illness that differ from clinicians' views (7). Outcome assessments in vasculitis should include both evaluations (8), while evaluating a patient it is important to consider all dimensions of the disease in addition to clinical aspects.

Fayers and Hays explained the steps of developing a questionnaire as literature review, qualitative interviews, expert review and pre-test of the instrument (9). The qualitative interviews are so important that they can bring out the domains of the illness from the patients' perspectives. They also promote information different from the biomedical perspective.

The qualitative study design and grounded theory approach was used for the study. Grounded theory was chosen as analytical methodology as it is particularly suited to develop concepts that represent the participants' experiences. Depending on the research tradition and field, qualitative researches aims

Table I. The number of interviews conducted with children and their mothers.

Children with vasculitis (n=10) >7 years	Mothers (n=10) with a child >7 years	Mothers (n=2) with a child <7 years	Total interviews (n=22) Total cases (n=12)
5 HSP	5 HSP	1 HSP	6 HSP
3 PAN	3 PAN	1 Takayasu	3 PAN
2 Behçet	2 Behçet		2 Behçet 1 Takayasu

HSP: Henoch-Schönlein purpura; PAN: polyarteritis nodosa.

to understand the world from the point of view of the research participants, interpret the experiences and meanings of people in complex settings, discover themes, knowledge, and relationships and explicate the ways people in particular settings come to understand, take action, and otherwise manage their day-to-day situations. Qualitative interviews prevent to remain disease-related data limited to the assessment of health professionals and provides to access patient-centered data in-depth (10, 11). The grounded theory emerges from the data and from the informants. The first step is data collection, through individual in-depth interviews. From the collected data, the key points are marked with a series of *codes*, which are extracted from the text. The codes are grouped into similar *concepts* in order to make them more workable. From these concepts, *categories* are formed, which are the basis for the creation of a *theory* (10, 11).

According to Herlyn (8), there are many manifestations of vasculitis (fatigue, reduced energy level) that were quite important to patients but not measured with the outcome instruments currently included in clinical trials of vasculitis. In this regard, the aim of the present study is to gain insight into needs of children with vasculitis from different domains of illness such as physical, psychological and social and to determine the affected areas by the disease that will provide a basis for the multidimensional instrument named "Juvenile Vasculitis Multidimensional Assessment Report" (J-VAMAR) in the lights of these results.

Methods

Methodology

Data were collected using both a demo-

graphic data form and a semi-structured interview form. Interview form was developed by the authors (experts), and included open-ended items to gather data on the experience of the participants. These items provided the participants with an opportunity to express their feelings and thoughts. The items in the interview form were based on the literature, theoretical framework and expert opinion (9). The questions of the form includes the difficulties due to the illness, physical, emotional and social impact of the disease, the school problems, the problems about relationships and the problems about medicines.

Participants

The research was conducted at the Paediatric Rheumatology Department of Gulhane Military Medical Academy and Hacettepe University in Ankara. Between August 2011 and April 2012, 12 children with childhood systemic vasculitis between the ages of 2–18 and their mothers (n=12) were enrolled in this study (Table I). Patients included in the study were between the ages of 2–18, could speak and understand Turkish.

In qualitative approaches, improved understanding of complex human issues is more important than generalisability of results (12). So in our study we preferred to use the "Judgement sample technique". Also known as purposeful sample, this is the most common sampling technique in qualitative researches. The researcher actively selects the most productive sample to answer the research question. This can involve developing a framework of the variables that might influence an individual's contribution and will be based on the researcher's practical knowledge of the research area, the available literature

and evidence from the study itself. It may be advantageous to study subjects who have specific experiences or subjects with special expertise (12).

The judgement sample technique tries to select patients with different vasculitis subtypes and with high severity to get more details about their diseases and the affected areas. So the developed multidimensional assessment instrument may be capable of detecting all the problems for all types of vasculitis.

Procedure

The study was made on individual patient face-to-face interviews. Firstly, the aims of the study were explained to the patients who volunteered to participate. They were informed that the interviews would be audio-taped.

It is important to use the questionnaires according to children's age and cognitive status. If the child is under 7, parent-proxy reports should be used (13). So, the interviews were conducted with both the children (whom over 7 years of age, n=10) and their mothers (n=10). If the child was younger than 7, the interview was conducted only with the mother (n=2).

The data of the study were collected through individual in-depth interviews using a specific methodology. The interviews were carried out in the clinics where the participants were being treated. The reason for choosing individual in-depth interviews was due to the idea that the individual interviews were regarded as the most appropriate for revealing experiences more freely. Interviews were carried out in a silent room. The answers given by the participants were recorded, and the author (BEF) took notes for further clarification of the answers. The interviews went on until data saturation was reached.

Analysis

Each interview was transcribed as soon as possible after being completed. The transcriptions were copied and were handed to the seven authors who are experts on outcomes. The expert group consisted of two paediatric rheumatologists (ED, SO), a paediatric psychiatrist (DK), an epidemiologist (CA) and three paediatric nurses (BEF, DK and DY). Each author scrutinised the texts

and took notes. Then, the authors got together to sustain consensus for the themes. Based on the collected data through the individual in-depth interviews, common themes were developed by merging similar statements for every category.

The study was approved by the local ethics committee of the research hospital. The data were analysed by the grounded theory and the N-Vivo 9 software program. This software program helps to find patterns and put things into groups and categories and see everything at once (14, 15). Under the leadership of this program, formulated meaning was organised into categories and clusters of themes as described in grounded theory. When differences arose among the investigators regarding the appropriate categories and themes (Fig. 1), they were discussed until a consensus was reached.

Results

Patients

Twelve children with systemic vasculitis were enrolled in this study. Six of the children were diagnosed with Henoch-Schönlein purpura (HSP), 3 of them with poliarteritis nodosa (PAN), 2 of them with Behçet’s disease and 1 of them with Takayasu arteritis (Table I). Patients with HSP in our study were complicated cases with renal and joint involvement and three of them were hospitalised during the study period for immunosuppressive treatment. PAN may be characterised by severe clinical presentations (16). In our study, patients with PAN were too complicated such as the one with cardiologic involvement, one with gastrointestinal tract involvement, and the other patient with renal involvement. One of the patient with Behçet’s disease had central nervous system involvement and the other one had severe ulcerations both genital and oral area, and skin involvement.

The median age of patients was 11.25±4.86 (min 5, max 17) years. Disease duration was 2.33±1.77 years (min 1, max 4 years).

Four categories and twelve themes of responses describing the experiences of the patients with vasculitis (Fig. 1) were determined. These categories and

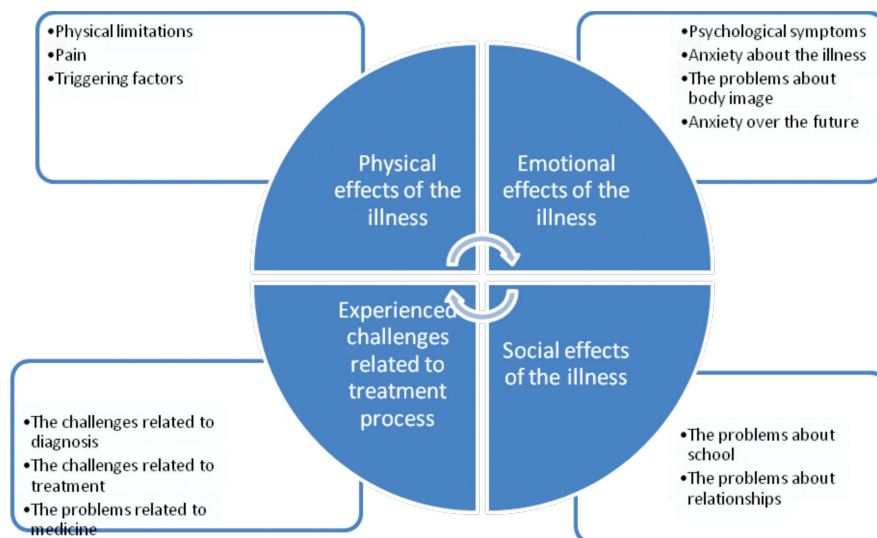


Fig. 1. Categories and themes resulted from interviews (All the categories and themes are generated with the N-Vivo 9 Software Program).

themes were emerged from the explanations, feelings and thoughts of the participants and they reflect the participants’ experiences about their illness.

Category 1:

Physical effects of the illness

Theme 1.1: Physical limitations

The most frequently stated physical symptoms by the participants were muscle weakness, fever, nausea, inability to run and/or stand on feet, oral and genital ulcers; numbness, bruising fingers swelling, exhaustion. Such disease related symptoms patients had to refrain from their hobbies, had difficulty in playing, described difficulty in writing, difficulty in eating due to oral ulcers, even in using bathroom due to inability to walk, as well as tiredness and muscle weakness. One child expressed herself as: “I was always watching my friends playing. I am not able to walk, run and play with them. I even could not go to bathroom without the help of my mother” (RA, 15 years old, male, PAN).

Theme 1.2. Pain

All children stated pain in different locations such as legs, abdomen, stomach, heart, muscle and joints. Furthermore, restrictions related to pain included inability to walk, insomnia, not willing to play, and study. In this regard, one of the mothers provided the following explanation: “Mostly he suffers from pain in

arms and knees. When the pain comes he tells that he wants to die. Even he wants me to stab him to death”.(Mother of N.O., 7 years old, female, Takayasu).

Theme 1.3. Triggering factors

Triggerring factors were variable and included cold and rainy weather, recurrent illnesses, and emotional stress.

Category 2:

Emotional effects of the illness

Theme 2.1: Psychological symptoms

When the psychological symptoms are assessed, children mostly expressed being upset, aggressive, stressed and even hiding and crying.

Parents stated changes in their children’s mood. They told that the child used to be very cheerful and started to turn into an introvert person. Families stated that they were having psychological support and treatment due to depression. Parents also complained that the disease affected the child negative. The children became so pessimistic, depressed and hopeless.

In this regard, one of the participants provided the following explanation: “... my child is sick. Why her? It does not fit to her. She is always pessimistic and thinking of dying. Does a five year old child know about death? Yes, she does. She asks me “Mom will I die?” She does not have any hope for the future (Mother of N.O., 7 years old, female, Takayasu).

Theme 2.2: Anxiety about the illness

The children and their parents stated that they really feel anxiety about the illness and expressed the major cause of anxiety as uncertainty. Patients stated anxiety about the uncertainty of disease outcomes, possibly having disease all life long and being dependent on someone. In this regard, one of the children provided the following explanation: "I am always under treatment and I don't think it is getting any better. I feel that it will always be like this all my life and it really puts me off" (E.A., 16 years old, female, Behçet's disease)

Parents expressed that they worried about recurrence of the disease, involvement of other organs and the fear that something bad would happen to their kids. "I am worried so much about his illness and I am afraid that he will not get better. Will that cause any problem in the future? Will it affect my kid's growth? I mean it seems that there is no cure for this. Doctors say that he has to use the drug all his life. What will happen in the end?" (Mother of N.O., 7 years old, female, Takayasu).

Theme 2.3: Problems about body image

Patients with vasculitis were dissatisfied with their body image. Some of them were complaining about changes (oral or facial ulcers) due to disease itself and some were complaining about the effects related to drugs (weight gain due to steroids).

Patients with Behçet's disease were bothered by their friends questioning about their facial lesions and feeling anxiety that it would leave scars on their faces. One of them stated that "At first I didn't have any ulcers on my face. When I started having them I really disgusted myself. I am very upset when my friends talk about this. They are really annoying at these ages." (E.A., 16 years old, female, Behçet's disease).

Some children using steroids were complaining about weight gain. They say that they look very awful because they look fat and feel embarrassed when they are with their friends. Questions such as "why are you getting this fat?" upset them a great deal.

Patients stated that they do not want to get together with their friends because of these changes in their physi-

cal appearance. One of the participants stated that "After the summer holiday my friends could not believe their eyes when they first saw me. They asked me 'What is that? Why did you get this fat?' I did not want to look in the mirror for a long time." (E.K., 16 years old, female, PAN).

Theme 2.4. Anxiety over the future

This category describes the patients' anxiety with regard to future. The reason for such emotions is that vasculitis has an unpredictable nature and it leads to difficulties for making plans for the future. One of the mothers of the participants stated that "We are worried, afraid and have future concerns. What will he do when we get old and not able to help him? We do have future concerns".

The majority of the participants are not sure about how they will feel tomorrow or another day. Their major anxiety is whether limitations due to their illness will increase or not.

The main reasons for their anxiety are getting worse and increasing in deformations. With regard to this anxiety, one of the subjects explained her situation as follows: "as I go to routine controls at the hospital I see a lot of Behçet's patients there. I see young people in wheelchairs. I am worried a lot. I ask what will happen to me in the future." (E.A., 16 years old, female, BD).

Category 3: Social effects of the illnessTheme 3.1. Problems about school.

Patients with vasculitis stated that they were bothered with not being able to attend school because of having to go to routine controls and because of disease symptoms. They said that they fall behind their school schedule and their classmates. Moreover, they complained that their academic performance is getting worse as they can not study properly because of disease manifestations and can not listen to lessons effectively. "It is very painful; I can not go to school anymore and always stay home. Of course it is affecting me psychologically because I can not go to school, I fall behind schedule. I was also getting prepared for my university exam, I feel behind and unable to catch up my friends. That is really bad" (E.K., 16 years old, female, PAN).

"He will have an exam tomorrow but we will receive laboratory test results two days later. We were expecting to go back today, now he won't be able to attend school for 3 days. He had the same experience while he was in secondary school. His teachers would send him home because he had severe headache due to hypertension. He could not understand his lessons because of absenteeism. (Mother of I.S., 12 years old, male, HSP).

Theme 3.2. Problems about relationships

Children state a lot of restrictions in their social lives, like being in the hospital on their birthdays and also state having problems with their relationships with their friends and relatives because of their physical appearance. One child stated that "when I go to school, my friends ask me 'What happened to you?' Some even couldn't recognise me. I sometimes ask myself 'Am I ugly?' I want no questions anymore and I want no one to see me." (S.T., 15 years old, female, PAN).

Some parents state that their children are sometimes frustrated because of their illness and the things they have been through.

Category 4: Experienced challenges related to treatment processTheme 4.1. The challenges related to diagnosis

Parents described some difficulties in diagnosis process. Symptoms being attributed to different diseases caused misdiagnosis and delay in diagnosis. Parents also believe that they were not informed well enough about their child's illness.

Theme 4.2. The challenges related to treatment process

What parents complained about treatment process were frequent routine controls, difficulty in getting to hospital due to heavy disease symptoms, appointments for a forward date and not being informed enough about the treatment procedure. A mother expresses her feelings about her child in pain "He is weary of life. He doesn't want anything. He says that he is tired of everything. He rebels 'If I am gonna die, let me, If I am gonna live, let me please'

Can you imagine taking a kid in such pain to a hospital? (Mother of R.A., 15 years old, male, PAN).

When it comes to patients, they complain about following challenges: absence of enough information about their treatment, adjustment to a salt free diet, confinement to bed in hospital and being concerned about having an infection.

"I can not eat salty food because I am afraid my blood pressure will increase as I have renal involvement. When I eat toasts and chips even in small amounts I really feel weird." (B.O., 14 years old, male, HSP)

Theme 4.3. The challenges related to medicine

Children with vasculitis mostly complain about using medications for a long time. Besides, they state that they do not want to take medicine at school because they hide their health problems from their friends. Children also say that they don't feel like using their medicine due to their side effects such as nausea, fatigue, loss of appetite, weight gain, susceptibility to infections, headaches, excessive hair growth or loss of hair but that they are afraid of having severe symptoms of the disease back if they discontinue to the treatment.

Discussion

This study represents the first step with a qualitative approach to children with systemic vasculitis that displays the effects of illness from different domains. The main findings of our qualitative study can be summarized as 1) Interviews with children with vasculitis and their families showed us the disease cause some effects such as physical, psychological and social other than organic manifestations 2) Proxy-reported outcomes provide deeper data than the assessment of the patients by only biomedical perspective. 3) It is necessary to develop a multidimensional assessment report that includes the affected areas due to childhood systemic vasculitis reflected from the patient perspective.

It seems difficult to evaluate the different domains of illness using current instruments. Considering the assessment of these aspects as well as other clinical manifestations would be effective

on treatment and outcome of the disease. Birmingham Vasculitis Activity Score (BVAS) and its modified version for children called Pediatric Vasculitis Activity Score (PVAS) assess the organic effects of childhood systemic vasculitis. In this study we showed the differences between adult and pediatric patients would encompass not only the previously mentioned features but also psychological and social factors (3).

Our results showed that the patients were dealing with the difficulty of performing daily life practices due to the pain and negative effects of their illness on their physical capacity. They also experienced emotional and social problems due to these effects. They mostly emphasised the changes in their physical appearance occurring as a result of their illness, and the relationship problems with friends. The most frequently stated problems by the participants were those related to daily life activities, and anxiety about the illness and about the future. Other frequently reported problems were related to diagnosis, treatment and medications.

Herlyn *et al.* reported fatigue, reduced energy level, and musculoskeletal symptoms were considered to be the most important burdens of disease by adult patients with vasculitis (8). Similarly, in our study physical limitations and pain were the major symptoms in the physical category. These problems are newly identified key domains in children with vasculitis and are extremely common with systemic rheumatic diseases and other chronic illnesses (17-21), but are often not directly measured in clinical trials or are considered to be manifestations of the underlying disease. In our study the physical symptoms were so dramatic that the child could not go to toilet by himself. Physical limitations and pain caused the school absenteeism and limitation of social activities. The children could not manage the activities appropriate for their age.

Additionally, several aspects of disease not covered by the list of physical and mental symptoms were commonly cited by patients, such as psychological symptoms (crying, depression, getting nervous) and anxiety about the illness because of uncertainty. There are some studies that evaluate these domains in

paediatric rheumatic diseases but there is no study focused on childhood vasculitis (21, 22). Some of the most important results of our study were that patients were highly concerned about uncertainty about progress of the illness and about their future. Our findings about substantial negative impact on patients' quality of life due to vasculitis are consistent with prior studies conducted with adults (23-27). Our study is the first one that evaluates these impacts on patients with childhood vasculitis.

One of the major sources of stress for children with vasculitis was the changes in body image related to the treatment or disease related damages. Jones *et al.* identified a correlation between negative reactions and visible symptoms (28). Our patients were so unhappy with gaining weight because of steroids that they did not want to show their new body images to their friends. Extreme changes in the physical appearance quite negatively influenced interpersonal relationships. Crandell and Moriarity identified a correlation between extreme physical difference and social rejection (29). In chronically ill children, the change in body image is frequent and it is especially important for adolescent ages (30).

According to Hockenberry (30), chronically ill children may conceal the illness from their friends, they may shy of their illness. The chronic process may affect child's academic success and social well-being. Our participants expressed that they did not want to go to school because of the illness. They could not attend school regularly and be successful academically. They stated that they concealed their illness from friends.

The diagnosis, the nature of symptoms and the extent of organ involvement affect the management of vasculitis (31). The decision of using steroids alone or in combination with immunosuppressive agents is largely dependent on vessel size and on the vascular bed that is involved (32). The patients must adjust to a salt-free diet because of the steroids. Also, the immunosuppressive agents may cause frequent infections. Our patients also frequently mentioned treatment-related problems that caused limitations such as infections and adjustment difficulties to a salt-free diet.

Current questionnaires can miss many of the patient-reported outcomes measurements (PROM) assessed in this study. It therefore seems essential to implement a disease-specific patient-reported questionnaire for patients with vasculitis to fill in this gap. Adding standardised validated PROM assessments to clinical studies in childhood vasculitis would provide greater insight into the impact of the disease. Adding patient outcome assessment to the current measurements in vasculitis would improve outcome assessment and might optimise clinical decisions and the detection of treatment effects. Finally, it is important to emphasise that multidimensional assessment is important as it is hard to obtain this information from routine clinic controls.

The areas that were significantly affected by the disease are determined based on the relevant data collected from the children and their mothers. This is a crucial step for item generation of "Juvenile Vasculitis Multidimensional Assessment Report" (J-VAMAR). It is important to note that this assessment report is original as this is the first time it is applied to children with vasculitis, and it is not adopted from an adult form.

Individual interviews provide an efficient way to collect data from users' perspectives. However, there is a limitation that should be kept in mind. The data relies upon participants' verbal reports and as such, may not reflect their actual behaviour or true feelings. Another potential concern is the constitution of the samples, which were small and may have been unrepresentative of their target populations on several dimensions. First, because the participants were volunteers and had no means of comparing their characteristics with those of non-responders, it was impossible to ascertain whether the samples exhibited characteristics that may have biased the results.

We conclude that currently used assessment tools for systemic childhood vasculitis are able to evaluate only the impact of biomedical effect of the disease. Our qualitative methodological approach showed this aspect of the disease that can not be evaluated from the biomedical perspective is very important in the follow-up. It is important

to develop a multidimensional assessment form to explore the areas which are significantly affected by the disease and this report should be appropriate for the children age and qualified to assess the impact of disease.

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