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Factors influencing quality of life in patients with inherited ichthyosis: a qualitative study in adults using focus groups

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Summary

Background There is limited information regarding quality of life in patients with inherited ichthyosis.
Objectives To identify factors influencing quality of life in patients with inherited ichthyosis.
Methods The study used focus groups and involved adult patients suffering from inherited ichthyosis from three French hospital centres. Group discussions were conducted by two facilitators and were continued until data saturation was reached. The verbatim transcripts were analysed independently by two investigators. Categories considered as key factors in the modulation of quality of life were negotiated until agreement was obtained.
Results Data saturation was reached after the fifth group. A total of 25 patients affected by various forms of ichthyosis attended these focus groups. The identified factors influencing quality of life were related to physical health, daily life, relations with others or oneself. However, together with difficulties related to ichthyosis, patients also underlined some positive aspects of the disease and described specific measures used to improve their quality of life.
Conclusions This is the first study investigating the different factors that could impact quality of life in patients with ichthyosis. This provides an essential framework from which physicians can develop strategies to improve patient care and quality of life and to develop a specific quality of life questionnaire.

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Inherited ichthyoses are disorders of cornification[1] with significant impact on quality of life (QOL).[2,3] QOL can be defined as an individual perception of one's position in life and is essential to assess in chronic diseases.[4] Factors influencing QOL have never been investigated in the setting of ichthyosis. The aim of our study was to identify these factors.

Methods

The protocol was approved by the local institutional ethics committee. Written consent was obtained from patients recruited from three departments with experts in the field of ichthyosis. The inclusion criterion was to be an adult suffering from inherited ichthyosis. A qualitative approach using focus groups was used. Discussions were conducted by two facilitators [a medical doctor (A.L.R.) and a psychologist (L.S.)] who had experience in leading focus groups and did not know the patients. The focus groups were continued until data saturation was reached. The transcripts were retranscribed verbatim and analysed independently by two researchers in the field of chronic diseases (A.L.R., I.A.) using a thematic content analysis. Categories considered as key factors in the modulation of QOL were negotiated until agreement was obtained.[5] The resulting analysis was sent to the participants for validation.

Results

Focus groups were conducted between October 2009 and September 2010. Saturation was reached after the fifth group (total of 25 patients) (Table 1).

Table 1 Characteristics of the 25 patients attending the focus groups

Characteristics	n = 25
Sex (female/male)	16/9
Age (years), mean ± SED (range)	41 ± 13·4 (21–67)
Forms of ichthyosis	
Lamellar ichthyosis	15
Erythroderma	4
Keratinopathic ichthyosis	2
Syndromic ichthyosis (Netherton syndrome)	3
X-linked ichthyosis	1
Systemic treatment (acitretin)	10
Employed	19
Marital status (couple)	13

Physical health

Patients suffered from an altered skin and eye appearance associated with significant symptoms (pain, pruritus and smelly skin). Worsening was reported in relation to environmental or psychological changes ('When I am stressed out, my skin changes'). Episodes of infections and heat intolerance were mentioned ('In a hot atmosphere I feel as if I were in a plastic bag'). Skin pain could be responsible for an impaired mobility. Sleeping could be disturbed by pruritus.

Daily life

Daily cream application was considered to be time consuming and nasty. Systemic retinoids were considered to be effective but side-effects were reported as tricky. Ichthyosis affected clothing (dark clothes were avoided, long sleeves were preferred). Patients complained about additional housework due to the scales. Some had to avoid professions with public contact or related to food; others reported being unselected due to ichthyosis. Nevertheless, some participants were content with their fight for a successful professional route. In relation to leisure, most had to avoid activities in hot atmospheres or risk inducing a worsening of their condition. Some also avoided activities in which they cannot hide the skin (e.g. swimming pool). Travel required adapted facilities and organization (shower availability, high quantities of creams). A majority of emollient costs were not reimbursed and this represented a significant cost.

Relationships with others

Patients reported various reactions from people including staring, tactlessness ('Why don't you see a doctor?') and inquisitiveness ('What happened to you?'). Nevertheless, some emphasized that this gave the opportunity of selecting people for more sincere relations. Acceptance of the disease and support from families or friends were considered as positively influencing their QOL. An important issue was related to intimate relations. Patients were afraid of reactions of repulsion and the relationship might not last long because of the burden of the disease. With regard to relations with the medical profession, patients considered that they had been treated insufficiently (lack of knowledge or interest). Conversely, the competence and listening skills of some doctors were considered as having a high input on QOL.

Relationship to oneself

The patients do not consider themselves to be handicapped even if they realize they have limitations. They reported fears in relation to pregnancy, treatment and ageing. Patients reported a number of negative feelings including sadness, discouragement and feelings of loneliness or anger. An important statement was the extreme variability in mood and feelings over time ('1 day I feel great, the following I feel awful'). Some positive feelings were also mentioned by some patients ('I am proud of the fact that I managed to face the disease'). The patients considered ichthyosis to have had a strong influence on the building of their personality ('without the disease I would be someone else'). Some patients hid themselves and had difficulties in approaching others. In contrast, others developed aggressiveness. Some considered that the disease made them become stronger, more mature and sensitive.

Specific procedures to improve quality of life

Some patients tried to find measures to improve their skin condition on specific occasions (intensification of treatment). They insisted on the importance of having an image of ichthyosis ('my skin is like a cracked desert'). They avoid mentioning the disease and meeting people. They also try to think positive, and push themselves, aiming for higher goals and increasing achievement ('we were obliged to do more and better than others'). Some patients gave a lot of themselves in patient support groups or research.

Discussion

Focus groups have been used widely in the social sciences and previously to study atopic dermatitis.[6,7] Nevertheless, this approach has some limitations as it cannot cover all topics and cannot be generalized. The two investigators were not experts in ichthyosis and therefore were not influenced in their analysis by their own knowledge.

This is the first study investigating factors influencing QOL in ichthyosis. We reported influencing factors related to physical or mental health, daily life and relations with others or oneself. One of the identified factors was the impaired appearance of the skin. Symptoms and impaired mobility were also highlighted by the patients. Another significant factor was the consequence of the disease on daily life, including the cost of the disease.[8] With regard to relations with others, patients suffer from the negative reactions of others, with intimate relations emerging as a major concern. Patients also expressed a significant dissatisfaction in relation to medical care.

To improve patient care, therapeutic educational programmes should be offered. These data are crucial in building specific programmes that would take into account the reported feelings, together with the necessity of improving social care, patient information and the training of clinicians. Control of pruritus and pain should also be a therapeutic goal.

Although we identified factors negatively influencing the QOL, the originality of this study is based on the identification of factors with a positive influence together with specific strategies of coping. It is important to take into account these positive aspects and not only the disabilities. Chronic disease can be considered as an opportunity for important changes rather that an ineluctable phenomenon. The perspective is to help the patient to 'become the same, yet different person', focused on patient empowerment.[9]

These data will also be used to build a specific QOL questionnaire that will be useful for further studies. This study identified factors that impact QOL in ichthyosis and provides an essential framework for improving patient care.

What's already known about this topic?

- Only two papers in the literature reported an altered quality of life in adult patients suffering from inherited ichthyosis.

What does this study add?

- This study identified factors influencing quality of life in patients suffering from inherited ichthyosis and specific measures used to improve their quality of life.
- This will help in developing strategies to improve patient care and a specific quality of life questionnaire.

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