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Recovering autonomy is a key advantage of home-based immunoglobulin therapy in patients with myositis

A qualitative research study

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Abstract

Immunoglobulins are 2nd or 3rd-line treatments in dermatomyositis (DM) or polymyositis (PM) refractory to high-dose corticosteroids and immunosuppressants. Immunoglobulins (2 g/kg/mo) are usually administered intravenously (IVIg) once a month and the patients stay at hospital for a few days. Recently, subcutaneous injections (SCIg) were proposed 2 to 3 times per week, in some dysimmune diseases. SCIg are administered at home preferably by the patient or by a nurse. We investigated the needs and attitudes of DM and PM patients with experience of IVIg and SCIg.

Seven patients (6 PM and 1 DM) from a single center participated in a focus group (N=6) or underwent in-depth interview (N=1). Six had the experience of both IVIg at hospital and SCIg at home; 1 has received only IVIg at hospital. Verbatim was recorded and transcribed for further content analysis and computer-aided textual analysis.

Clinical profiles and stories were heterogeneous. At diagnosis, muscle weakness, severe pain, and fatigue were at the forefront of patients' complaints impairing daily life. Patients reported considerable improvement with immunoglobulins. SCIg were described as easy, less disruptive for daily life, well tolerated, and less time-consuming. SCIg self-administration at home restored the feeling of autonomy and control.

Interviews of DM and PM patients revealed that recovering autonomy and control was a central advantage of home-based SCIg that were efficient, well tolerated, and perceived as a good compromise between treatment burden and efficacy.

Abbreviations: DM = dermatomyositis, Ig = immunoglobulin, ILD = interstitial lung disease, IV = intravenous, PID = primary immunodeficiency, PM = polymyositis, RT = replacement therapy, SC = subcutaneous.

Keywords: in-depth interviews, focus group, textual analysis, patients' expectations, preference, autonomy

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According to the French Public Health Code, approval of local ethic committee is not required for surveys related to patient satisfaction. Informed consent was obtained from all individual participants included in the study.

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1. Introduction

Dermatomyositis (DM) and polymyositis (PM) are idiopathic acquired inflammatory myopathies, a group of diseases encompassing DM, PM, autoimmune necrotizing myopathy, inclusion body myositis, cancer-associated myositis, and connective tissue disease-related myositis with an annual incidence of one in 100,000.^[1] DM, unlike PM which is not seen in children, has a bimodal age distribution, with peaks at 5 to 24 years and 45 to 64 years of age.

The DM and PM are two distinct autoimmune myopathies with specific histopathologic features. DM is a humoral-mediated vasculopathy, while PM is characterized by endomyosial T-cell infiltrates.^[1] They share proximal more than distal muscle weakness, electrophysiologic abnormalities, elevated serum creatine kinase levels, and inflammation on nuclear magnetic resonance (NMR) and muscle biopsy.^[2] Symmetric muscle weakness occurs subacutely or progressively, associated or not with muscle pain or tenderness. Inflammation of the skeletal and smooth muscles of the gastrointestinal tract can lead to dysphagia and delayed gastric emptying. Myositis is associated with characteristic skin manifestations like a heliotrope rash around the eyes or a macular rash over the face, trunk, and limbs. DM patients are at higher risk of interstitial lung disease (ILD),^[3] inflammation of the pericardium and cardiac muscle,^[4] and malignancies.^[5] Most myositis patients have autoantibodies

against nonorgan-specific antigens. More than 20 different autoantibodies are known. They are associated with different clinical phenotypes of myositis and different genetic markers. Analyzing survival of 192 Taiwanese PM and DM patients, Yu et al reported an overall cumulative survival rate of 79.3% at 1 year, 75.7% at 2 years, 69.9% at 5 years, and 66.2% at 10 years. Survival time was significantly reduced in patients with thrombocytopenia, diabetes mellitus, ILD, and cancer patients.^[6]

The inflammatory and autoimmune nature of DM and PM provide a rationale for immunosuppressive or immunomodulating therapy. Intravenous (IV) or subcutaneous (SC) immunoglobulins (Igs) could be used as 2nd or 3rd-line therapy after lack of adequate response with high-dose oral corticosteroids, combined or not with methotrexate, azathioprine, mycophenolate, or ciclosporine.^[7–9] Mechanisms of action of Ig are not clearly defined and multiple hypotheses have been proposed: interference with membrane attack complex deposition,^[10] downregulation of adhesion molecule,^[11] downregulation of Transforming growth factor Beta 1,^[12] inhibition of dendritic cells,^[13] competition with autoantibody binding to Fc receptors on antigen-presenting cells, neutralization and accelerated catabolism of pathogenic autoantibodies, downregulation of B cells receptors, interference with antigen recognition by T cells, and interaction with cytokines.

Accumulating evidence points out the efficacy of Ig in refractory, relapsed, or steroid-dependent DM/PM.^[14] Data are supported by 2 randomized placebo-controlled trials,^[15,16] 7 prospective uncontrolled studies,^[17–23] and 1 retrospective case-note review.^[24] Short-term benefit has been confirmed on long-term follow-up.^[19,25] Recent studies highlighted that SCIg were a cost-effective alternative to IVIg.^[26–28] However, to date, data regarding myositis patient's experience about Ig treatment remain scarce. We report here the qualitative analysis of the interviews of 6 PM and one DM patients having had the experience of IVIg and among whom all but 1, were receiving SCIg at home.

2. Methods

The PM or DM patients followed in a single center (Pitié-Salpêtrière Hospital, Paris, France), previously treated with IVIg or having the experience of both IVIg and SCIg (Gammanorm, Octapharma Lachen, Switzerland) were invited by their physician (PCh) to participate in a focus group or to undergo in-depth interviews. They were individually orally informed about the study and gave their oral consent. According to the French Public Health Code (Article R1121-1 amended with decree 2017-884 of May 9, 2017), approval of the local ethics committee is not required for surveys related to patient satisfaction or study in human and social sciences in the field of health. The interviews within focus group were semidirective thanks to a structured guide of interview (see Supplemental Data, <http://links.lww.com/MD/D701>). The latter was developed and designed on expert opinions (PCh). Patients were asked to talk about their inaugural symptoms, the treatment with Ig, and their needs of information. The focus group lasted 3 hours and the interview lasted 1 hour. They were audio recorded and verbatim was transcribed for further analysis. French transcripts were not translated before textual analysis.

A content analysis (AD) and a computer-aided textual analysis (PCI) were separately conducted. Content analysis was performed by examining major themes. Conclusions of both analyses were reconciled (PCI) and are reported here. Acquisition

of textual information was based on text mining techniques, methods of processing, and representation of textual data. Computerized textual analysis used an R interface for multidimensional analysis of texts and questionnaires (IraMuteQ version 0.7 alpha 2; <http://www.iramuteq.org/>). Textual analysis has initially been developed for the analysis of large texts but is now used for smaller corpus as a complement of other types of analysis such as content analysis. The use of a computer program provides the advantage of coding, organizing, and separating information, which allowed for rapidly locating the entire text segment used in the qualitative writing.

Verbatim transcript was divided a priori into 3 subcorpus exploring disease, Ig treatment, and the need for information. This division followed the schedule of the semistructured interviews. Inflected and derived words were reduced to their word stem. It means that conjugated verbs were replaced by the infinitive form and that plurals or feminine words were reduced to the masculine singular form. The analysis was performed separately for each subcorpus. Texts were cut in segments of 40 words. Active forms were counted separately from tool-words. The frequency of each form was analyzed in the global corpus and each subcorpus with iterative returns to the transcription to assess the relevance of the form relatively to the studied concept. Indeed, some words may be homonymous having different meanings and covering different themes. A short list of 54 active forms was determined and used for the analysis of each subcorpus. The analysis of similarities was graphically displayed using the Fruchterman–Reingold algorithm based on co-occurrences. A co-occurrence is the concomitant appearance of 2 or several forms in the same segment. Graphs are reproduced here with the French verbatim to respect the language structure. In addition, a correspondence analysis was performed to allow the analysis of specificities. Analyses outputs were sorted according to the main topics of the discussion. Analyses were reviewed by the different authors to ensure consistency regarding the source data.

3. Results

3.1. Population

Six patients participated in the focus group (interviewees #1 to #6) and one patient had an in-depth interview by phone (interviewee #7). Characteristics of interviewees are summarized in Table 1. All but 1 (#7) were female aged 31 to 83 years. All but 1 suffered from PM and 1 (#6) had a DM. One patient (#5) had an antisynthetase-associated PM. Two patients were working (#5 and #7), while one 36-year-old patient (#6) and one 59-year-old patient (#2) ceased working. All had a history of high-dose prednisone and immunosuppressants before hospital-based IVIg and all but 1 (#4) were currently receiving SCIg at home using pumps.

3.2. Corpus

The global corpus was composed of 7 texts (1 per interviewee), 24,117 occurrences (for a mean number of 3460 occurrences by text) and 1882 active forms. Verbatim was richer for the perceptions of the disease and its treatment than for the need of information. Subcorpus included 8110 occurrences and 1057 active forms for the perception of the disease, 10,462 occurrences and 1057 active forms for the treatment, 4925 occurrences and 701 active forms for the need of information.

Table 1
Characteristics of interviewees.

	Gender	Age	Working status	Diagnosis	Current Ig regimen
#1	Female	83	Retired	PM	Home-based SCIg
#2	Female	59	Stopped working	PM	Home-based SCIg
#3	Female	61	Retired	PM	Home-based SCIg
#4	Female	70	Retired	PM	None
#5	Female	31	Currently working	Antisynthetase-associated PM	Home-based SCIg
#6	Female	36	Disabled	DM	Home-based SCIg
#7	Male	45	Stopped working Currently working	PM	Home-based SCIg

DM = dermatomyositis, PM = polymyositis.

3.3. Perception of the disease

Interviewees often began sentences by “in my case . . .” suggesting that they did not perfectly recognize themselves in the words of others. Patients have understood that PM or DM were diseases affecting the muscle (25 occurrences), causing 2 major symptoms that were intimately linked: pain (24 occurrences) and muscular weakness (11 occurrences) (Fig. 1). Pain was described as “unbearable” (#1), “infernal” (#1), “constant” (#2), “burning” (#5), involving legs (#1), fingers (#5, #7), muscles and joints (#7).

- “My muscles became inflamed. It was like you catch fire.” (#6)

Predominantly proximal muscular weakness had consequences on daily life such as falls (6 occurrences), or difficulties in climbing stairs (9 occurrences), holding an object firmly (5 occurrences) or more rarely swallowing (4 occurrences). Daily activities were restrained by the disease. Loss of strength has forced some patients to ask for “help” (10 occurrences).

- “I had no strength; I could not climb on my bed, stand up when I was sitting on a chair, stand up from the toilets. When I was eating, it was difficult to swallow and it compelled me to drink a glass of water after each bite.” (#3)

Physical limitations negatively impacted their physical and leisure activities: “Muscular weakness imposed to me to stop sport.” (#6)

Impairment of muscular strength was associated with a severe and deep fatigue (18 occurrences)

- “Muscular weakness is beyond fatigue.” (#6)
- “I am tired when I wake up, when I go to bed; I cannot do very much. My head is willing but my body refuses.” (#2)

Most patients felt unable to work (15 occurrences), and some were forced to quit working.

Even if they do not use words as “anxiety” or “fear,” most interviewees reported they had not understood what had happened to them and the time they waited for a proper diagnosis was clearly a tense moment. Putting a name on their troubles was a relief and all patients were still grateful to their doctor for it.

Symptoms were associated with difficulty in talking to relatives about the disease and most patients have given up the will to speak about it.

- “It is difficult to talk about it, because it is an invisible disease.” (#2)
- “Words are missing: fatigue cannot render how much I feel tired.” (#6)

- “People did not guess I was sick. It was something inside me.” (#6)
- “Other people are not interested.” (#3)
- “I do not want to speak about my disease at work.” (#5)

3.4. Perception of Ig treatment

The IVIg were perceived as efficacious, but time-consuming and constraining. Uncomfortable side effects such as fever, thrills or headache have been experienced with IVIg. Some patients worried about their venous bed. One patient reported peak and through effect, feeling better during the 2 weeks following the IVIg injections and worse until the next administration. Hospital was perceived as reassuring but at the cost of a loss of freedom and control.

- “At hospital, we enter in a circuit and we do not control things anymore.” (#5)

All patients but 1 (#4) had an experience with both hospital-based IVIg and home-based SCIg. All of them expressed their preference for home-based SCIg. Surprisingly, most patients reported better efficacy with SCIg than with IVIg. Analysis of similarities revealed four dimensions in patient’s discourse about home-based SCIg: autonomy/management/ritualization; easiness; repetition/frequency; material and procedures (Fig. 2).

With SCIg, patients said they have recovered “autonomy” (12 occurrences) and this was clearly stated as an “advantage” over IVIg (5 occurrences). Autonomy was placed in the foreground especially by young active people (#5, #6, #7).

- “The first advantage of subcutaneous injections is autonomy, flexibility, choice of the day and time.” (#5)
- “Autonomy means liberty.” (#6)
- “We can do it when we want; we are free.” (#1)
- “I do it by myself, in autonomy. I manage myself successfully.” (#7)
- “I recovered the control of my life.” (#5)

Patients insisted on terms like “to manage” or “management” (15 occurrences), “to organize,” “organization” (10 occurrences), putting themselves forwards as actors of their own treatment. Only 1 patient (#2) relied on a nurse for preparing and doing the infusions since she felt unable to self-administer the treatment. Performing infusion at home was also associated with the protection of “intimacy” (#6). One participant talked about a “me-time” moment (#2).

The SCIg have to be repeated more frequently than IVIg and 1 patient (#6) remembered she felt freed between monthly

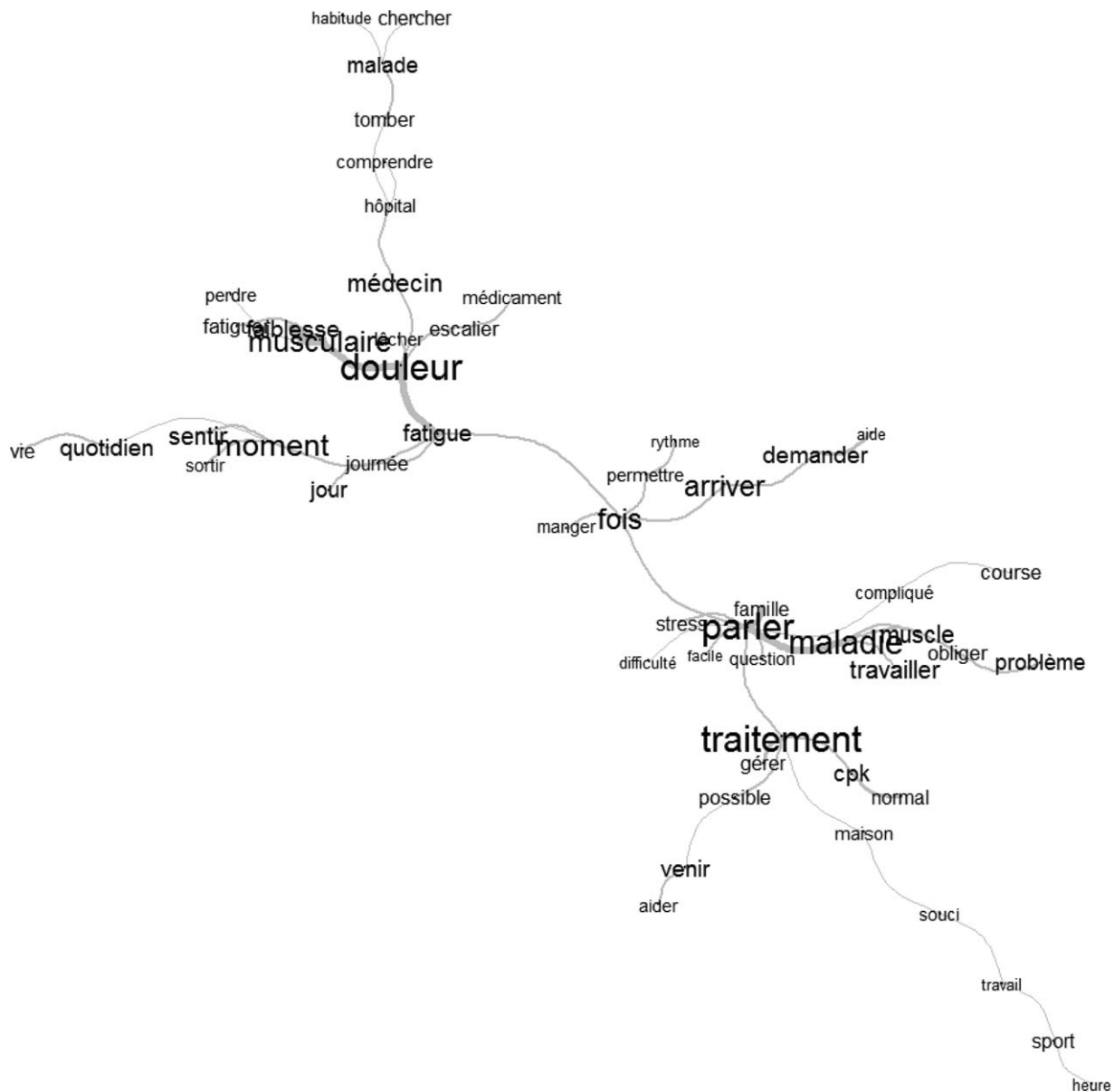


Figure 1. The size of words is proportional to their frequency. As an example, the word “Douleur” (pain) was more frequent than the term “Faiblesse musculaire” (muscular weakness). The word “Parler” (speak about) was frequently encountered and was close to the word “maladie” (disease), meaning that the patients had difficulties to speak about their disease with their relatives. The lines joining the words represent the co-occurrence of terms in the same segment and the line thickness is proportional to number of co-occurrences.

intravenous injections. Using words like “time” (61 occurrences), “week” (27 occurrences), “day” (24 occurrences), and “hours” (40 occurrences), the patients highlighted the frequency of injections, the necessary ritualization, and the shorter duration of SCIG. Choosing the day and time of infusion has conferred a feeling of liberty with the sensation of leading a normal life and to save time. “Ritualization” was opposed to the necessary “planning” when treated at hospital. “Rituals” (3 occurrences) helped the patient to integrate their treatment in their daily life; on the contrary, planning was necessary to deal with disruption of family and professional life when receiving IVIg. Indeed, due to the big volumes to be injected, hospitalization could last 2 to 5 days and this was clearly identified as disruptive.

- “Usually I have no problem of organization since I perform injections on fixed days.” (#5)
- “I always do it on Tuesday and Friday, at 2 pm, regularly.” (#3)
- “I prepare the material always at the same place.” (#6)
- “It is a ritual. On the morning between 8 am and 10 am when my children are departed to school. After that, I can rest for a few hours.” (#6).
- “I managed a kind of ritual.” (#5)
- “When the injection has started, I go about my everyday activities.” (#6)

Patients described the use of pump, syringe and needle as somewhat easy and felt successful in handling material. Nonetheless, the role of the nurse for the 1st infusions and of

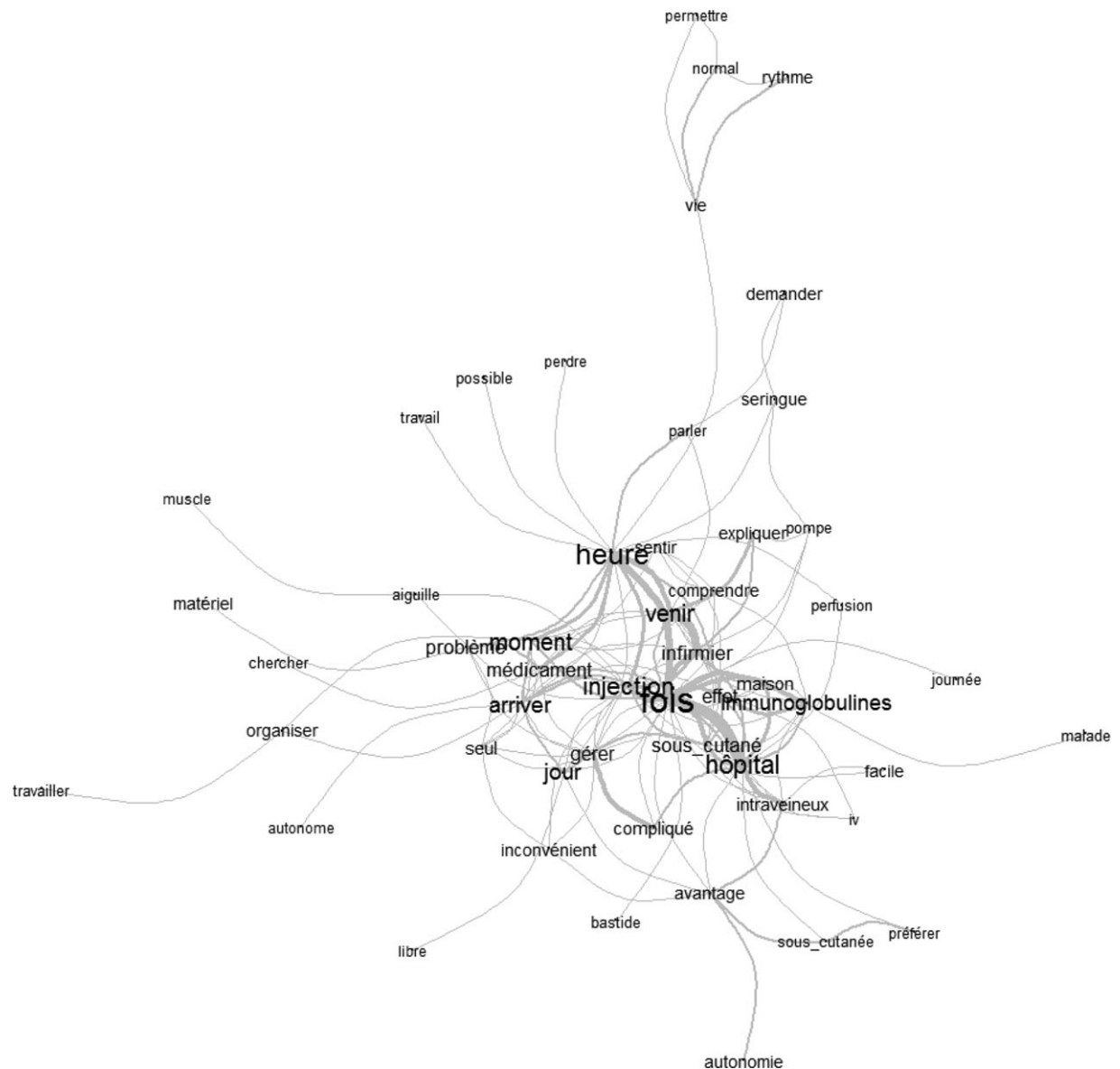


Figure 2. The size of words is proportional to their frequency. As an example, the word “Fois” (time) was very frequent emphasizing the frequency of the treatment. This word was close to “Immunoglobulines” (immunoglobulins) and “Hôpital” (hospital) meaning that the burden of repeated injections was heavier when the patient was treated at hospital than when treated at home. The lines joining the words represent the co-occurrence of terms in the same segment and the line thickness is proportional to number of co-occurrences.

the local provider of material was emphasized. Only 1 participant reported that 1st injections may be “painful” (1 occurrence).

Overall, patients stroke a balance between the time lost to administer the treatment and the expected benefit.

- “I know I will lose 3 hours but it will allow me to have an almost normal life. Putting these two things together, I do not complain of losing a few hours.” (#6).

3.5. Need of information

The patients admitted to understand neither the mechanisms of the disease nor why it happened to them (15 occurrences). Despite their resignation, they still expressed their surprise and incomprehension. “Stress” was suspected to be a trigger or an

aggravating factor (8 occurrences). Only 1 patient (#5) correctly explained that she was suffering from an autoimmune disease.

Participants expressed a need for “information” (28 occurrences) but above all for individual support (6 occurrences). Patients said they have been interested in getting more information about the disease (41 occurrences) and the potential treatments (15 occurrences) mostly when they were diagnosed with myositis; this need was less marked today.

- “I needed to understand why it happened to me.” (#1)

They had questions (12 occurrences) and referred to their doctor (36 occurrences) who was said to play a key role in providing relevant and personalized information. In case of questions, they would preferably wait until the next visit to the doctor.

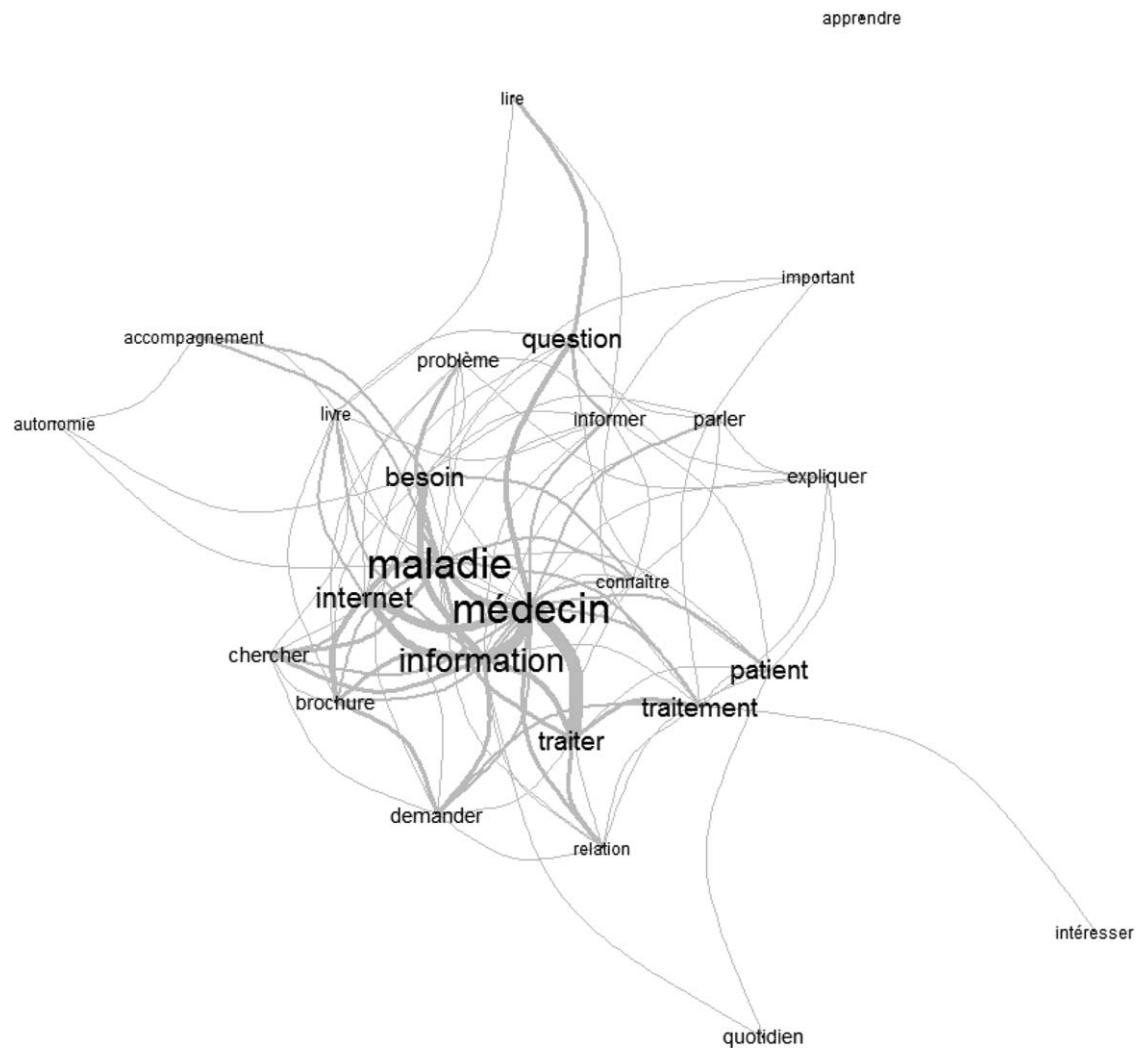


Figure 3. The size of words is proportional to their frequency. As an example, the word “Maladie” (disease) was very frequent and close to “Médecin” (physician), more cited than “Internet” as a source of information. The word “Apprendre” (to learn) is rarely used and patients preferred words as “Informer” (to inform), “Parler” (to speak), “Expliquer” (to explain), “Connaître” (to know). The lines joining the words represent the co-occurrence of terms in the same segment and the line thickness is proportional to number of co-occurrences. See the close relationship between “Médecin” (physician) and “Information.”

Internet was perceived as delivering potentially distressing, nonverified, irrelevant, and nonpersonalized information. Most participants had, however, spent a few hours on the internet to better know and understand their disease but all stated that they did not find a clear and adapted response to their questions. “Doctor” and “internet” were frequently opposed in a same sentence (Fig. 3). “Leaflets” (7 occurrences) and “books” (4 occurrences) were less used to get information but the participants would be happy to have a well-written brochure which could provide useful information between visits to the doctor. This would have been especially useful at the time of diagnosis.

- “Internet frightens me.” (#1)
- “I went on the internet. My doctor always told me not to do it.” (#1)
- “There is not a great deal about the disease on internet.” (#7)
- “I completely avoid the internet for medical questions. There are a lot of false things.” (#5)

4. Discussion

Our 1st finding was that clinical profiles and stories were heterogeneous. When discussing together, the interviewees were surprised by their so diverse experiences. Some complained from pain and others did not; some placed fatigue in the forefront while others pursued their work. All patients reported muscular weakness affecting proximal and to a lesser extent distal segments with sometimes dramatic consequences on autonomy.

According to our interviewees, treatment with Ig allowed the restoration of an almost normal life. This was obviously related to a selection bias since our patients had a successful past or current treatment with Ig after disappointing results with high-dose prednisolone and immunosuppressants.

All patients with experiences of IVIg and SCIg expressed a clear preference for SCIg which was described to be easy, less disruptive for daily life, well tolerated and less time-consuming. Preference was mainly related to a restoration of autonomy. Home-based self-administration reinforced the feeling of

independence as compared to hospital-based infusions and interviewees appreciated to be considered as autonomous actors of their treatment. Need for autonomy has been described as a common theme characterizing illness experience in patients affected by rare disease. This thematic domain was likely to be found in women patients as reported by Caputo.^[29] In their study, patients refused a dependency relationship with health services; on the contrary, they looked for a care provision that considers their emotional needs, priorities, and dignity, to be recognized as real persons, not just patients. It is likely that patients with DM and PM share this trait, and found in the SCIG mode of administration a way to less rely on medical healthcare professionals for their disease management.

4.1. Patient's attitudes regarding the chronicity of treatment and the role of rituals

The SCIG were preferred by patients even at the expense of more frequent injections. Patients clearly accepted it as the counterpart for having an almost normal life. IVIG or SCIG could be a very long-term treatment requiring frequent infusions. The optimal duration of therapy after complete remission is unclear, and there is no maximum timeline for use for those who continue to respond. The current guidelines recommend annual review for patients stable on long-term therapy.^[24] Consequently, the integration of the treatment in patient's daily life is of paramount importance. SCIG were well integrated in patient's routine. Infusions were ritualized, prepared, and made at the same place, on fixed days and hours. Home-based self-administration was said to require good organizational skills since the administration schedule has to be respected. This did not deter the patients who appreciated being involved in their treatment rather than having to undergo it. This positive attitude was clearly related to the feeling of having recovered autonomy and even the control of their life. Usually, ritualization improves adherence to treatment.^[30] Interviewing patients with primary immunodeficiency (PID) receiving a lifelong Ig replacement therapy, Cozon et al reported that patients who started Ig replacement therapy with hospital-based IVIG also appreciated the autonomy when switching to home-based SCIG.^[31] IgRT at home then allowed to save time, travel expenses, and days off work. Interestingly when PID patients talked about the frequency of SCIG they suggested that each infusion was perceived as a reminder they were sick.^[31] For this reason, most preferred once-a-week SCIG with pump rather than rapid push infusions with a syringe done 2 to 3 times a week.^[31] This could be due to the fact that PID patients are treated to decrease the risk of infections and that in everyday life they feel healthy. In DM and PM patients, increase in infusion frequency did not bear this negative aspect. As they have had severe long-lasting symptoms, SCIG was perceived as a curative rather than as a preventive therapy.

4.2. Strength and limitations of the study

As far as we know, this is the 1st report of in-depth interviews of DM and PM patients treated with SCIG at home for several years. The 2nd originality of our work is the dual approach of patients' discourses. Computer-aided textual analysis and content analysis contributed to a better understanding of patients' needs and attitudes. The main limitations of our study were the small sample of patients, an obvious selection bias, and the recruitment in a single center. Given the selected population, satisfaction

regarding SCIG may have been overestimated. Qualitative by nature, generalization of the results is limited and must be cautious.

5. Conclusion

Interviews of PM and DM refractory patients treated with subcutaneous high-dose immunoglobulin infusions revealed the symbolic meaning of managing their own treatment at home. In practical terms, the treatment was well integrated in daily routines.

Author contributions

Conceptualization: Patrick Chérin, Annaik Dokhan, Pierre Clerson

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Validation: Pierre Clerson

Visualization: Pierre Clerson

Writing – original draft: Pierre Clerson, Yann Fardini

Writing – review & editing: Pierre Clerson, Yann Fardini, Patrick Chérin, Taylor Pindi Sala, Annaik Dokhan, Martin Duracinsky, Jean-Charles Crave, Olivier Chassany

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