Daily life, needs and expectations of patients with acromegaly in France: An on-line survey

Frédérique Albarel, Fatine Elaraki, Brigitte Delemer

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Vie quotidienne, besoins et attentes des patients atteints d'acromégalie en France : le point de vue des « e-patients »

Dr Frédérique Albarel [a], Dr Fatine Elaraki[b], Pr Brigitte Delemer [c],

[a] Aix-Marseille Université, Institut National de la Santé et de la Recherche Médicale (INSERM), U1251, Marseille Medical Genetics (MMG), Marseille, France and Assistance Publique-Hôpitaux de Marseille (AP-HM), Department of Endocrinology, Hôpital de la Conception, Centre de Référence des Maladies Rares d'Origine Hypophysaire HYPO, 13005, Marseille, France.

[b] Ipsen pharma , Boulogne-Billancourt

[c] Service d'Endocrinologie, Diabète-Nutrition, CHU de Reims, 51092, Reims Cedex.

bdelemer@chu-reims.fr

Corresponding author:

Dr Frédérique ALBAREL

CRMR d'Origine Hypophysaire HYPO,

13385 Marseille Cedex 15 – France

Tel: 04 91 38 34 79

Fax: 04 91 38 41 31

E-mail: frederique.albarel@ap-hm.fr

Abstract
Acromegaly can impair quality of life, but impact on patients' daily life, needs and expectations been little explored.
Objectives: To better understand the impact of acromegaly on patients' daily life, and evaluate their needs and expectations.
Patients and methods: An on-line questionnaire survey of acromegaly patient and relative community members was conducted on the Carenity website.
Results: 25 patients and 3 relatives, with a mean age of 48.9 years, responded. Diagnosis of acromegaly was recent (60% within 10 years). Signs at diagnosis were mainly clinical (fatigue, headache) and physical changes (acral enlargement). Reported complications comprised morphological changes (93%), bone and joint symptoms (75%) and metabolic disorders (75%). Pain and fatigue impacted the daily life of 61% and 54% patients, respectively. Acromegaly had strong impact on mood (79%), leisure (75%) and social life (71%). Patients mostly got information from specialized websites (71%), their endocrinologist (61%) and patient associations (54%). The information sought was patient testimony (82%), practical advice (64%), and information on clinical trials (61%) and treatments (50%). Patients wished to have patient-physician discussion groups (25%), and better knowledge of acromegaly on the part of those they were in contact with (25%).
Conclusion: Acromegaly has a major impact on patients' daily life and mood. Patients wished their disease to be better known, and advocated setting up discussion groups. This study should improve acromegaly education programs and help adapt the services and information provided.

Keywords: acromegaly; daily life; impact; e-survey; needs; expectations

Résumé
L'acromégalie peut entraîner une altération de la qualité de vie. Ses conséquences sur la vie quotidienne des patients, leurs besoins et attentes sont peu connus.
Objectifs : Il s'agit de mieux comprendre l'impact de l'acromégalie sur la vie quotidienne et évaluer les besoins ainsi que les attentes des patients.
Patients et méthodes : Des patients acromégales et leurs proches, membres des associations de patients, ont répondu à un questionnaire sur internet sur la plateforme Carenity.
Résultats : 25 patients et 3 proches, 48,9 ans d'âge moyen, ont répondu au questionnaire. Le diagnostic d'acromégalie était récent (60 % < 10 ans avant). Les premiers signes au diagnostic étaient majoritairement des symptômes cliniques (fatigue, céphalées) et des modifications physiques (infiltrations des extrémités). Les complications rapportées étaient des modifications morphologiques (93 %), des symptômes osseux/articulaires (75 %) et des anomalies métaboliques (75 %). La douleur (61 %) et la fatigue (54 %) retentissaient sur la vie quotidienne. L'acromégalie avait un impact important sur le moral (79 %), les loisirs (75 %) et la vie sociale (71 %). Les principales sources d'information étaient les sites internet spécialisés (71 %), l'endocrinologue (61 %) et les associations de patients (54 %), avec des recherches sur des témoignages de patients acromégales (82 %), des conseils pratiques (64 %), des informations sur les essais cliniques (61 %) et les traitements (50 %). Ils souhaitaient des groupes de parole patients/médecins (25 %) et une meilleure connaissance de l'acromégalie autour d'eux (25 %).
Conclusion : L'acromégalie a un impact important sur la vie et le moral au quotidien des patients. Ces derniers voudraient que l'on connaisse mieux leur maladie et la

création de groupes de parole. Cette étude va permettre d'améliorer les programmes d'éducation dans l'acromégalie, et d'adapter les services et informations délivrées.

Mots clés : acromégalie ; vie quotidienne ; retentissement ; enquête internet ; besoins ; attentes

INTRODUCTION

Acromegaly is a chronic acquired disorder characterized by excessive secretion of growth hormone (GH) and increased insulin-like growth factor 1 (IGF-1) levels, caused in most cases by a benign pituitary adenoma. Nowadays, its prevalence may vary between 28 to 137 per million people, while its annual incidence ranges between 2 and 11 cases per million inhabitants [1].

Acromegaly is characterized by the development of a dysmorphic syndrome with progressive acral and cranial enlargement [2], accompanied by a number of comorbidities as hypertension, glucose intolerance and diabetes mellitus, sleep apnea and respiratory diseases, and heart failure. Chronic rheumatologic disorders, with joint deformations associated with pain and impotence [3] are also frequent and usually persist even after control of the disease. Acromegaly can have physical, psychological and social manifestations, which may impact quality of life (QoL), especially in patients with active disease [4]. The great majority of patients reported musculoskeletal and joint pain ; depressive and anxiety symptoms are commonly observed in those who have been diagnosed with an extended delay [5].

To better understand the impact of acromegaly on patients' daily life and evaluate the needs and expectations of patients in terms of services and information, a survey was carried out online in patients or relatives registered in a social network of individuals with acromegaly.

PATIENTS AND METHODS

Design of the study

The survey was carried out by Else Care company on the Internet platform Carenity (https://www.carenity.com/), the French social network of patients suffering from chronic diseases from March 2, 2016 to April 24, 2016.

An online questionnaire was sent to the members of acromegaly communities on Carenity website. Carenity patients' community was solicited to participate in the survey by means of invitations and reminders sent by email. The members of the French patients' association "Acromégales, Pas Seulement…" [APS; Patients with Acromegaly, but not only] were also recruited through the Facebook page and invitations and reminders sent by email.

Participants

All patients with acromegaly members of Carenity community and living in France (including overseas territories), as well as their relatives, could be included in the study. Relatives were to answer in place of the patient with acromegaly.

Questionnaire

The online questionnaire was designed by Carenity and validated by Dr F. Albarel and the association APS as an expert of the disease. The questionnaire comprised 4 parts: main stages and circumstances of acromegaly diagnosis, management of treatment and acromegaly impact on DL, search of information on acromegaly, and needs and expectations of acromegalic patients (Questionnaire available as supplementary information).

RESULTS

Demographics and clinical characteristics of respondents

Of 47 patients recruited by email, 36 responded to the questionnaire, 28 responses to the questionnaire were valid.

Demographics and clinical characteristics of respondents are shown in Table 1. Most respondents were female (79%) and 89% were patients, while 11% were relatives responding for the patient. Patients were aged 48.9 years on average and more than half was 41 to 60 years old (57%). Patients reported having been diagnosed recently (≤1 year earlier) or quite recently (2-5 years earlier) for 14% and 32% of respondents, respectively, while 18% of patients were diagnosed over 20 years earlier. To the question "which kind of medication for acromegaly do you have?" 25% answered having no treatment, while the others declared taking one to 3 medications.

First signs of acromegaly and reported complications

Respondents reported in average 3 warning signs that led to diagnosis. Physical changes (n=30 responses), with frequent foot and hand (12 et 11 respectively) deformation were reported (Figure 1). Patients also noticed clinical symptoms (n=41 responses), mainly fatigue (n=7 patients), headache (n=6) but also increased sweating and weight gain (5 patients each). Pain was also reported as a warning symptom (n=8 responses).

Furthermore, respondents declared undergoing a mean of 8 symptoms they considered as "complications" related to acromegaly. In 32% of cases, they reported more than 10 symptoms, among which morphological changes, bone and joint symptoms and metabolic disorders were the most frequently reported (93%, 75% and 75%, respectively) (Figure 2). Only one patient (4%) had no "complication".

Impact of acromegaly and its treatment on patients' daily life

Of the 21 respondents who declared being on treatment at the time of the survey, 19 (90%) had their treatment administered at home, and 2 (10%) at the nurse's office. Besides helping with the injection of the treatment, the nurse played also a role in the management of

acromegaly by providing psychological support for 28% of patients (n=5/18), giving information on the prescribed treatment (n=3/18) or providing care for acromegaly-related complications (n=3/18).

To get greater autonomy in the management of their treatment, patients most often expect the availability of a new oral treatment (n=6/19), more information on their disease (n=2/19) and more attentive listening from their physician (n=2/19). However, a third of them (n=6/19) report that they do not miss anything to be more autonomous.

Regarding the main acromegaly-related difficulties cited by the patients as impacting their daily life (Figure 3), pain and fatigue were the most commonly reported (n=17/28 and n=15/28, respectively). Other difficulties impacting quality of life were reported (Table 2), such as reduced mobility (n=5/28) and impact on professional life (n=4/18). Difficulties concerning management (burden of treatment, surgery…), or of psychological nature were less frequently cited. Regarding the impact of acromegaly-related difficulties on daily life, it was considered quite important to very important on their daily mood for 79% of patients, on their hobbies (75%) and on their social life (71%). The impact of the disease on their professional and sexual life was also quite to very important in 68% and 67% of cases, respectively.

Seeking information on acromegaly

The most common source of information on their disease for patients with acromegaly was the Internet with websites specialized in orphan diseases and acromegaly (71%), but also the endocrinologist at the hospital (61%) and patients' associations (54%), while they solicit their general practitioner in only 21% of cases (Figure 4). Patients get information through several ways (mean: 3.6 sources) and in about half the cases (46%), they have 4 or more sources. The vast majority (82%) seeks testimonies from patients with acromegaly, and practical advices

for a better daily life (64%), information on ongoing clinical trials (61%), treatments (50%), acromegaly-related complications (36%) and types of social assistances available (36%). Patients were also looking for information on specialists or reference centers (29%), administrative problems related to acromegaly in general (29%), information on acromegaly in general (18%), or practical advices for medical follow-up (14%).

Among the most expected services and information, patients cited the implementation of discussion groups including patients and physicians (n=7/28), a better knowledge of their disease (from the lay audience and health professionals in particular) (n=7/28), information tools on acromegaly (n=6/28), and a multidisciplinary management of their disease (n=4/28).

DISCUSSION

This survey carried out in a population of patients with acromegaly (n=28; 3 relatives) shows that this rare disease has an important impact on patients' daily life. Furthermore, although they already have several sources of information the patients express their need to be better informed, to have dedicated services, and their wish to share about their disease in discussion groups with patients and physicians.

In this survey, patients reported having experienced a mean of 3 warning signs suggestive of acromegaly, which vary depending on the patients. The most striking signs are non-specific clinical symptoms such as headache and fatigue, but also excessive sweating and weight gain, which are also observed along with a large range of symptoms in other studies [2, 3, 6].

Presence of concomitant physical changes, especially acral enlargement and facial changes, as reported in many series [2, 3, 6], was very frequently noticed by the patient themselves in our study. In contrast, joint and musculoskeletal pain are less common features noticed at diagnosis, in accordance with previous studies [2, 7], although arthralgia was reported in 50% of patients (n=26/52) at diagnosis in another study [3]. Symptoms considered by patients as

"complications" of acromegaly are multiple (8 in average per patient) and very frequent, especially morphological changes which are experienced by almost all patients (93%), but also bone and joint symptoms and metabolic disorders (75% of cases), which is in line with literature data [2, 3, 6].

Results of our survey also show that acromegaly have a notable impact on patients' daily life, especially pain and fatigue which are by far the most often cited as being a daily burden (60% and 54% of patients). Joint and musculoskeletal pain is described in up to 90% of acromegalic patients [8] and this condition, which may persist even after long-term remission of the disease [9, 10], has been found to impair patients' QoL in several studies [4, 8-10]. In our survey, acromegaly could also have a negative impact on QoL, as it restricts patients' access to hobbies and impact on social life of most patients. High pain scores of the spine, knee, and hip have been reported in long-term cured acromegaly patients which limit physical functioning and psychological well-being [8, 10]. This effect on psychological well-being was also observed in our survey, as 79% of the respondents estimated that acromegaly, besides impairing their professional, personal, sexual and social life, has an important to very important impact on their daily mood. Some patients also reported that their body image is impaired, some have difficulties to face scrutinizing looks and feel lack of understanding from their relatives, others experience depression. According to the literature, psychological status, and especially body image impairment, is one of the most relevant factors affecting QoL [4], even after long-term biochemical remission of acromegaly [11].

Management of treatment that needs to be injected could also be a burden for the patient. In most cases, in this survey, the patient has his treatment administered at home by a nurse. According to some patients on treatment (n=6/19), the best way to improve their autonomy in treatment management and what they expect would be a new oral treatment to be marketed. Patients also expect more information on acromegaly and listening from health professionals.

Although they have become experts in their own disease by getting information mostly on specialized websites and from their endocrinologist, acromegalic patients expect their disease to be better known from the lay audience and health professionals. But what they mostly await is getting information on existing and developing treatments or ongoing clinical trials, and practical advices and tools to improve their daily life. Due to their daily relationship with treated patients, nurses should have a key role to play by providing such information, but in fact, their major role in acromegaly management is the psychological support they give to the patient. Most patients (80%) seek testimonies from their "peers" who could help them to better understand their own problems and to feel less isolated with their rare disease. They also often search for information about their disease in patients' associations, forums, blogs and patients' networks. Hence, services patients mostly expect to better manage their disease daily is the implementation of discussion and meeting groups including patients and physicians, probably to get better understanding of their condition, psychological support, practical advices, information tools and accurate information on acromegaly and treatments that might finally improve their QoL.

This study has several limits, first because of its design. Sometimes, questionnaire has been sent through an association of patients with acromegaly and therefore 75% of patients knew its existence, which implies a recruiting bias. *Indeed, patient with the worse quality of life is more prone to look for solution and contact patient's association or website, so the result of this e-survey does represent the opinion of a subgroup of "connected" patient.* Furthermore, as it is a rare disease, the sample size of the study is small; it is thus difficult to generalize these results.

However, the main value of this study is that this is the first time acromegalic patients talk about their disease through such a "e-survey". This study therefore provides the direct reflection of how the patients really live with acromegaly, the impact it has on their daily life and the burden of the disease on their mood, their social, personal and professional life. These results will be a very useful basis to further implement patient education programs, and to

propose new services and information adapted to the needs of patients with acromegaly. Indeed, several pituitary education programs have been developed in France for the last few years, and this study, considering specifically acromegalic patients' needs will allow to improve and complete their contents.

In conclusion, here is the first "e-survey" evaluating daily life, needs and expectations of acromegalic patients. Symptoms and complications of acromegaly seem to have a major impact on patients' daily life, on their mood and their personal and professional activities. The main expectations of acromegalic patients are a better knowledge and better recognition of their disease, more information on current and developing treatments for acromegaly, practical advices and information tools, and finally, the possibility to talk about their disease with their peers and physicians within discussion and meeting groups. Probably evolution o existing education programs based on these data could partly meet their needs.

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Table 1. Demographics and clinical characteristics of respondents (N=28)

Tableau 1. Caractéristiques démographiques et cliniques des répondeurs (N=28)

Table 2. Impact of acromegaly on patients' daily life – Other difficulties (N=28 respondents, several possible answers, total number of responses: 36)

Tableau 2. Retentissement de l'acromégalie sur la vie quotidienne des patients - Autres difficultés (N=28 répondeurs, plusieurs réponses possibles, nombre total de réponses: 36)

Figure 1. Warning signs leading to diagnosis (N=28 respondents. Several possible answers, total number of responses = 79). Results are expressed in number of patients having cited the sign.

Figure 1. Signes d'alerte menant au diagnostic (N=28 répondeurs. Plusieurs réponses possibles, nombre total de réponse=79). Les résultats sont exprimés en nombre de patient ayant cité le signe

Figure 2. Rate of acromegaly-related manifestations reported as "complications" by the respondents at the time of the survey (N=28). Several possible answers, total number of responses: n=223.

Figure 2. Taux de manifestation en lien avec l'acromégalie rapportée comme des « complications » par les répondeurs (N=28). Plusieurs réponses possibles, nombre de réponses n=223

Figure 3. Impact of acromegaly on patients' daily life: main experienced difficulties. (N=28 respondents. Several possible answers, total number of responses: 47).

Figure 3. Retentissement de l'acromégalie sur la vie quotidienne des patients: principales difficultés rencontrées. (N=28 répondeurs. Plusieurs réponses possibles, nombre total de réponse : 47)

Figure 4: Sources of information for patients with acromegaly (N=28 respondents). Several possible answers, total number of responses: 100.

Figure 4: Sources d'informations pour les patients acromégales (N=28 répondeurs). Plusieurs réponses possibles, nombre total de réponse : 100

Figure 1. Warning signs leading to diagnosis (N=28 respondents. Several possible answers, total number of responses = 79)**.** Results are expressed in number of patients having cited the sign.

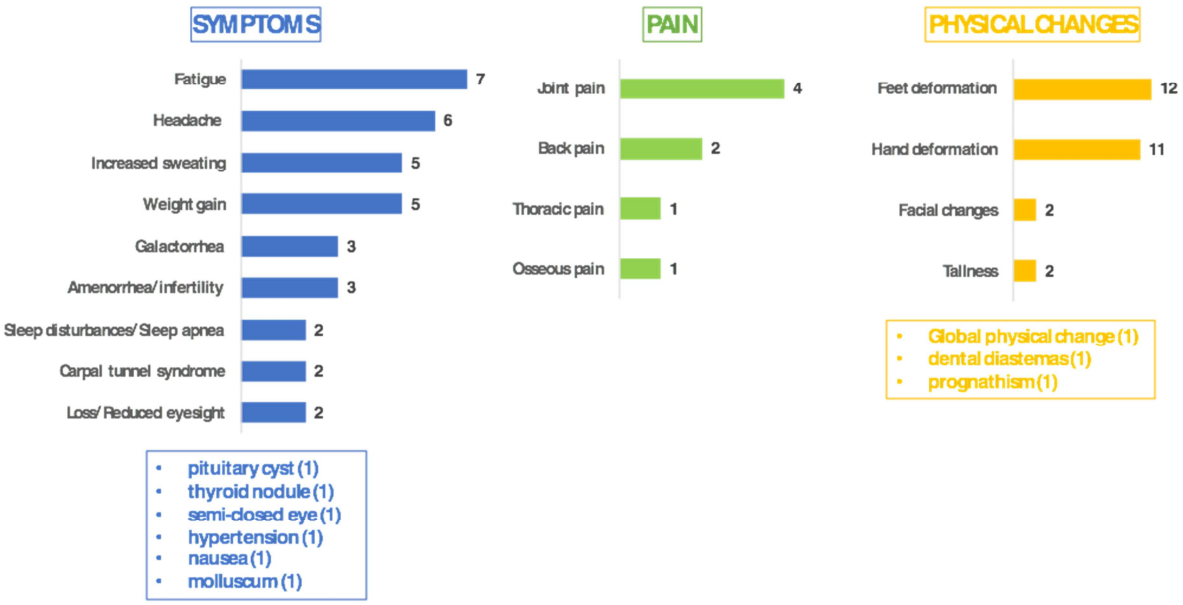

Figure 2. Rate of acromegaly-related manifestations reported as "complications" by the respondents at the time of the survey (N=28). Several possible answers, total number of responses: n=223.

Other: menopause (n=1/28), urinary incontinence (n=1/28), fatigue (n=1/28). ENT: Ear Nose and Throat

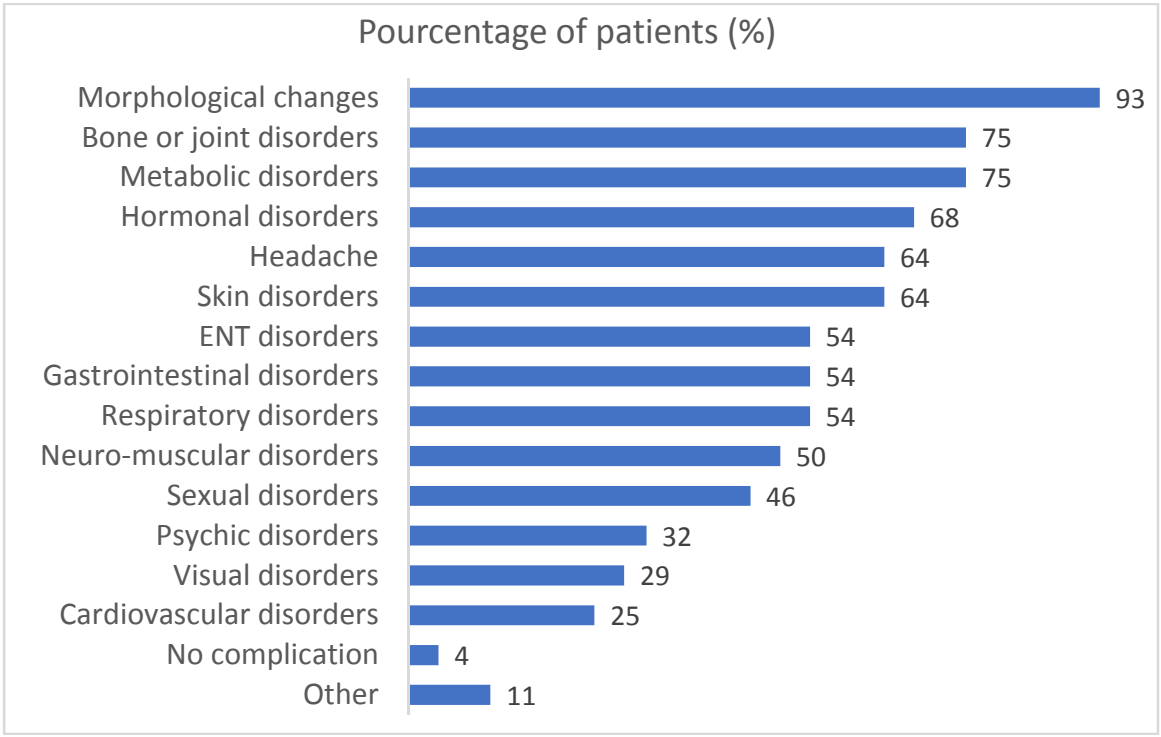

Figure 3. Impact of acromegaly on patients' daily life: main experienced difficulties. N=28 respondents. Several possible answers, total number of responses: 47.

Responses to the question "how would you describe today the impact of acromegaly on your daily life? What are the main difficulties experienced?"

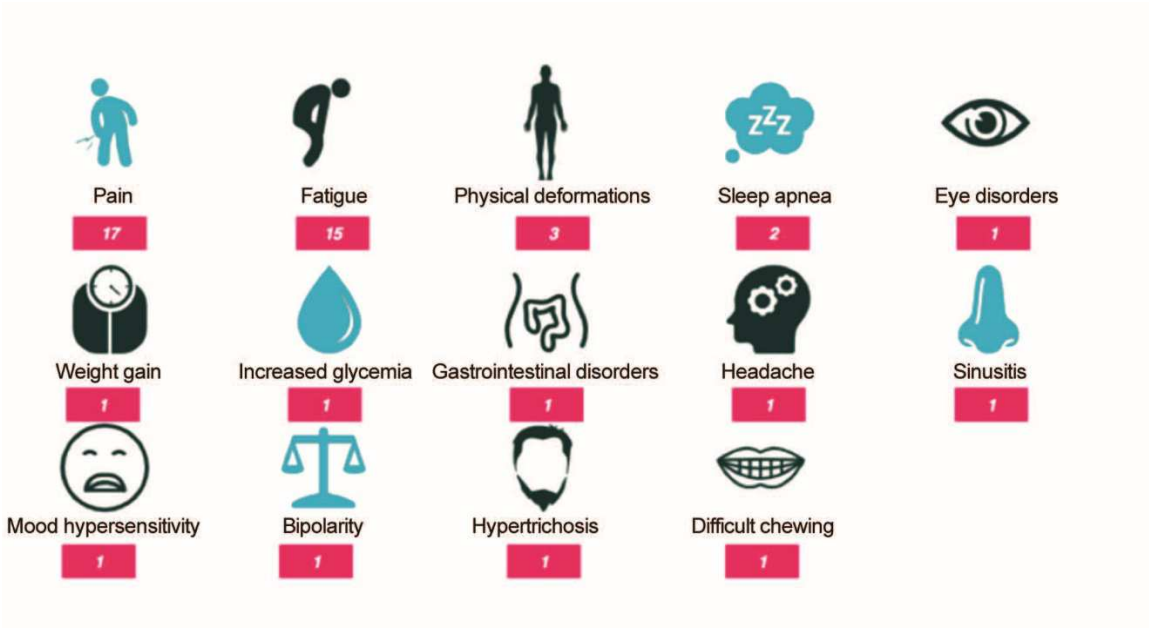

Figure 4: Sources of information for patients with acromegaly (N=28 respondents). Several possible answers, total number of responses: 100.

Other: Amrap (Association régionale des Acromégales de Midi-Pyrénées) n=1; Websites: n=1

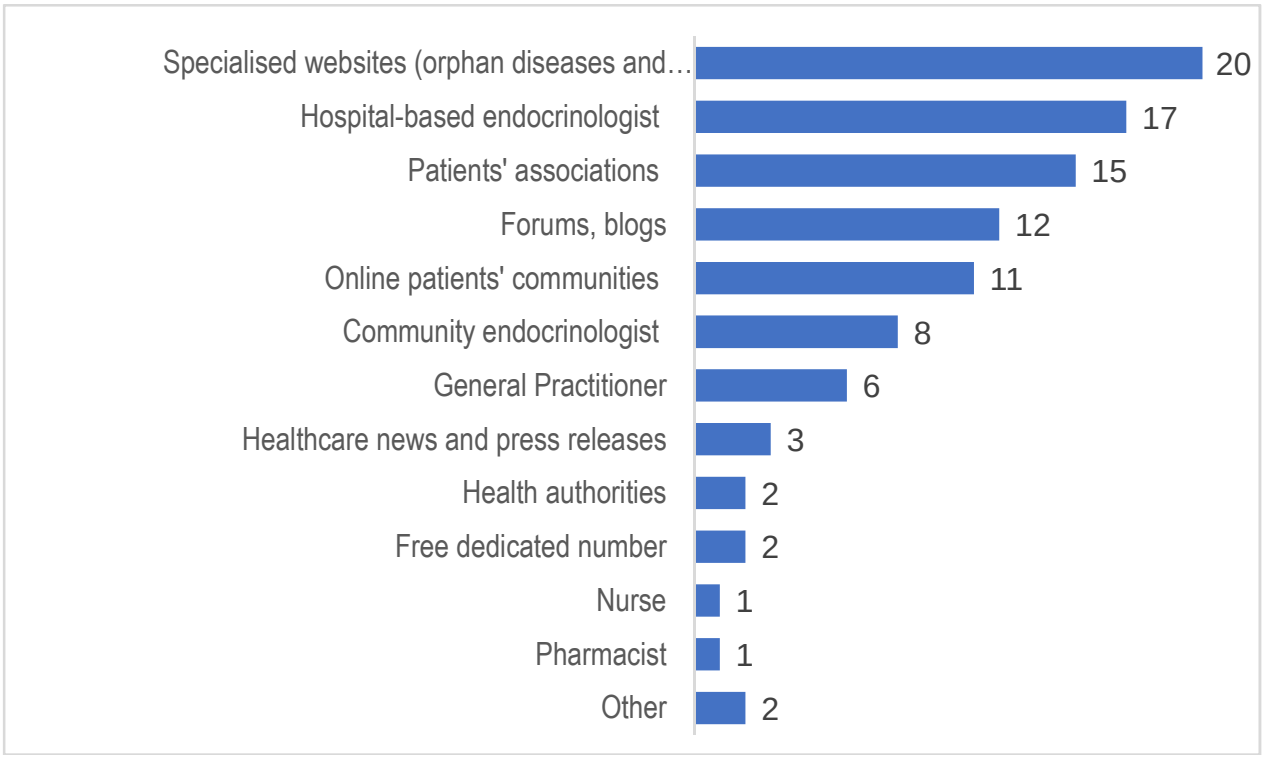

Table 1. Demographics and clinical characteristics of respondents (N=28)

Characteristics		**Respondents (N=28)**
Gender, %	Male	21
	Female	79
Age, y	Mean	48.9
Age category, %	18-30 years	7
	31-40 years	21
	41-50 years	32
	51-60 years	25
	61-70 years	11
	>70 years	4
Diagnosis anteriority, %	≤ 1 year	14
	2-5 years	32
	6-10 years	14
	10-20 years	22
	≥ 20 years	18
Treatment		
Number of drugs, n (%)	None	7 (25)
	1	14 (50)
	2	6 (21)
	3	1 (4)
Type of drug, n (%)		*N=36* [a]
	Lanreotide	12 (43)
	Octreotide	9 (32)
	Pegvisomant	6 (21)
	Cabergoline	2 (7)

[a] N= 36 responses, as some patients could take more than 1 treatment

Table 2. Impact of acromegaly on patients' daily life – Other difficulties (N=28 respondents, several possible answers, total number of responses: 36)

Type of Difficulty	Number of patients
Quality of life impairment (20 responses)	
Reduced mobility	5
impact on professional life	4
Rhythm of life disease-dependent	4
Financial difficulties	2
Reduced familial and social life	2
Daily fight against the disease	2
Difficulty to accept body image	1
Treatment management (8 responses)	
Burden of treatment / follow-up	2
Surgery	2
Lack of understanding from health professionals	1
Lack of information on disease progression	1
Medical "wandering"	1
Fear to develop other diseases	1
Psychological Difficulties (8 responses)	
Difficulty to face scrutinizing looks	3
Depression	2
Lack of understanding from relatives	2
Solitude	1