RMD Open
Rheumatic & Musculoskeletal Diseases

Original research

Clinical practice guidelines adherence, knowledge and awareness in rare and complex connective tissue diseases across Europe: results from the first ERN ReCONNET survey

Rosaria Talarico,[1] Diana Marinello,[1] Stefano Bombardieri,[2] Gerd Burmester,[3] Joao Fonseca,[4,5] Charissa Frank,[6] Ilaria Galetti,[7] Eric Hachulla,[8] Frederic Houssiau,[9] Ulf Mueller-Ladner,[10,11] Matthias Schneider,[12] Vanessa Smith,[13,14] Giuseppe Turchetti,[15] Jacob M van Laar,[16] Ana Vieira,[17] Maurizio Cutolo,[18,19] Marta Mosca[1,20]

To cite: Talarico R, Marinello D, Bombardieri S, *et al*. Clinical practice guidelines adherence, knowledge and awareness in rare and complex connective tissue diseases across Europe: results from the first ERN ReCONNET survey. *RMD Open* 2020;**6**:e001344. doi:10.1136/rmdopen-2020-001344

Received 28 May 2020
Revised 26 June 2020
Accepted 10 August 2020

For numbered affiliations see end of article.

Correspondence to
Rosaria Talarico; sara.talaricco76@gmail.com

ABSTRACT

Introduction The European Reference Network (ERN) ReCONNET is the ERN aimed at improving the management of rare and complex connective tissue and musculoskeletal diseases (rCTDs) across the European Union (EU). In the mission of ERN ReCONNET, clinical practice guidelines (CPGs) play a crucial role, representing a valid tool towards the harmonisation of the management of rCTDs while improving effectiveness and quality of care delivered to patients.

Methods ERN ReCONNET developed two surveys to map the adherence to rCTDs CPGs among healthcare providers and to assess the knowledge and awareness of CPGs for their diseases among patients, family members and caregivers.

Results The results of the surveys highlighted that healthcare professionals find it useful to apply CPGs in clinical practice (93%), while 62% of them experience difficulties and barriers in the application in their centres. Healthcare professionals also highlighted the need to develop CPGs for all rCTDs and to implement the use of the existing CPGs in clinical practice. On the other hand, patients, families and caregivers are relatively aware of the purpose of CPGs (51%) and 62% of them were aware of the existence of CPGs for their disease. Patient-friendly versions of CPGs and patients' lifestyle guidelines should be systematically developed contributing to the empowerment of patients in the disease management.

Conclusion ERN ReCONNET is addressing the main issues identified in the results of the survey, promoting practical actions for the local adaptation of CPGs across Europe, improving their routine clinical use and increasing the awareness on CPGs among rCTDs patients, family members and caregivers.

Key messages

What is already known about this subject?

- Clinical practice guidelines (CPGs) have an important role in guiding and supporting the decision-making processes. CPGs cover at present only some rare and complex connective tissue diseases and more efforts should be dedicated to the creation of the evidence needed to support the development of new CPGs.

What does this study add?

- This work provides the perspectives of European healthcare professionals and patients, family members and caregivers regarding their awareness, knowledge and adherence to CPGs.

How might this impact on clinical practice?

- The results of this work highlighted different unmet needs of the rCTDs communities, including the adaptation of existing CPGs and the implementation and development of the evidence needed to produce new CPGs. Considering that ERN ReCONNET represents a European infrastructure that can increase the knowledge and the awareness of existing CPGs, one of the major added value of the ERN ReCONNET is to address these unmet needs, providing a framework for the harmonisation of care in rare and complex connective tissue diseases in Europe.

INTRODUCTION

Around 5000–8000 rare diseases affect the daily lives of approximately 30 million people in Europe. Many of those affected by a rare condition have limited access to diagnosis and high-quality treatment. Unfortunately, expertise and specialist knowledge may be scarce because patient numbers are low and improving the evidence by pooling data can be a challenge.

In order to address these challenges, the European Commission launched the European Reference Networks (ERNs), virtual networks involving healthcare providers

(HCPs) across Europe. The aim of the ERNs is to tackle complex or rare diseases and conditions that require highly specialised treatment and a concentration of knowledge and resources. It is well known that no country alone has the knowledge and capacity to treat all rare and complex diseases, and exactly for this reason, ERNs were established as European infrastructures. ERNs offer, in fact, the potential to give patients and clinicians across the EU access to the best expertise and timely exchange of life-saving knowledge, making knowledge accessible to all patients, even when living in remote areas.[1]

Since their launch in 2017 in Vilnius,[2] 24 ERNs are currently working on a range of thematic issues, including rare connective tissue diseases, bone disorders, childhood cancer, metabolic disorders immunodeficiency and many others.

The ERN ReCONNET[3] is 1 of the 24 approved ERNs aiming at improving the management of rare and complex connective tissue and musculoskeletal diseases (rCTDs) across the EU. The ERN ReCONNET currently involves 26 full member HCPs from 8 different EU countries: Belgium, France, Germany, Italy, Netherlands, Portugal, Romania and Slovenia and 14 affiliated partners (APs) from additional 11 countries (figure 1). The network, coordinated by the Azienda Ospedaliero Universitaria Pisana in Italy, covers the following 10 rCTDs: antiphospholipid syndrome (APS), Ehlers-Danlos syndrome (EDS), idiopathic inflammatory myopathies (IIM), IgG4-related disease (IgG4), mixed connective tissue disease (MCTD), relapsing polychondritis (RP),€'s syndrome (SS), systemic lupus erythematosus (SLE), systemic sclerosis (SSc) and undifferentiated connective tissue disease (UCTD).

Full members HCPs are HCPs that have been identified as members of the ERN following the process of membership regulated by the Commission Delegated Decision of 10 March 2014 and the Commission Implementing

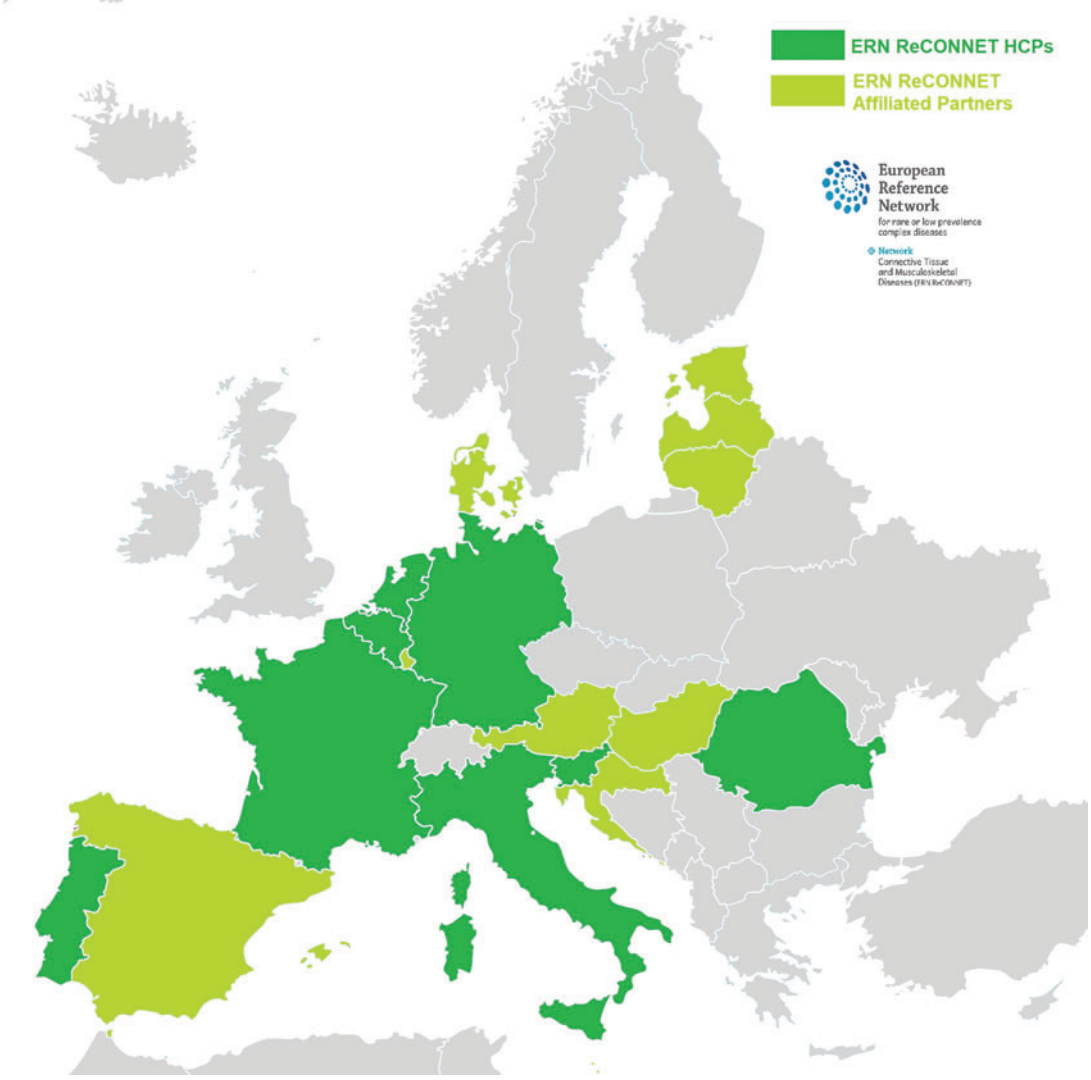

Figure 1 EU countries involved in the European Reference Network (ERN) ReCONNET.
ERN ReCONNET covers the following countries: Belgium, France, Germany, Italy, The Netherlands, Portugal, Romania and Slovenia with the HCPs full members, Austria, Croatia, Denmark, Estonia, Hungary, Latvia, Lithuania, Luxembourg, Malta and Spain with the affiliated partners.

Decision of 10 March 2014. APs) are HCPs belonging to member states that do not have representation from a full member within an ERN; APs are designated by their member states and can actively participate and contribute to the ERN activities. Other important components of the ERN ReCONNET are the external experts (individual clinician or healthcare professional with an expertise in the field of rCTDs that are approved by the ERN ReCONNET Steering Committee to participate in specific activities of the ERN) and the members of the ERN ReCONNET External Scientific Advisory Board (ESAB—experts considered relevant to the work and mission of the ERN, such as health economists, methodologists, policy-makers, etc). In addition, the ERN ReCONNET involves and engages with patients' organisations, thanks to ERN ReCONNET European Patients Advocacy Group (ePAG). The role of ePAGs is particularly important since they represent the voice of patients in all the activities of the ERN. All the activities and the stakeholders involved in the network are co-ordinated by the ERN ReCONNET Co-ordination Team, which involves the co-ordinator, project managers and expert rheumatologist acting also as methodologist.

Clinical practice guidelines (CPGs) serve as an equaliser in the field of rare diseases. Their application can have a high impact on the care of the patient and their usage highlights the difference between substandard care and patients living longer, healthier lives with fewer complications.[4–9] Guidelines, whether designed to support correct and early diagnosis or to guide appropriate care, can serve as a blueprint of excellence, bringing clinicians and healthcare professionals closer to the patients on how to treat them, reflecting the best possible knowledge and generating the best achievable outcomes. As mentioned in the Commission Delegated Decision of 10 March 2014, ERNs should (among other important activities) develop and implement clinical guidelines supporting the existing scientific societies in the process. For these reasons, ERN ReCONNET can play a crucial role in the implementation of CPGs for rCTDs, providing valid tools towards the harmonisation of the management of rCTDs throughout Europe while improving effectiveness and quality of care delivered to patients.

In this setting, during the first year of activity of the ERN ReCONNET, two anonymous surveys were created to map the adherence to rCTDs CPGs among HCPs members and to assess the knowledge and awareness of CPGs for their diseases among patients/family members/caregivers. Therefore, we will provide the results of the two surveys.

METHODS

Of the two surveys developed, the first 'ERN-ReCONNET survey on the use of clinical practice guidelines for HCPs' was dedicated to healthcare professionals and the second 'ERN ReCONNET survey on clinical practice guidelines knowledge and awareness in rare and complex connective tissue disorder patients, families and caregivers' was dedicated to patients, caregivers and family members.

ERN-ReCONNET survey on the use of clinical practice guidelines for HCPs

The survey was developed in English and it was aimed at assessing the adherence to rCTDs CPGs in European HCPs who are either members or collaborating with ERN ReCONNET and at identifying eventual barriers or limitations that can affect the specialist's clinical decisions in adopting CPG.

The survey consisted of 19-item questions in English, subdivided into three sections:

- General sections (demographics, professional characteristics, country of the respondent, etc).
- CPG section.
- Questions concerning potential difficulties/barriers to the application of CPGs.

The questions of the survey were designed by a group of methodologists, clinical experts in rCTDs and by the ERN ReCONNET Co-ordination Team and comprehended different types of answers (single or multiple-choice, Likert scale, open answer, etc).The survey was sent to healthcare professionals from one of the ERN ReCONNET full members HCPs or to those who were collaborating with the ERN as external experts or members of the ERN ReCONNET ESAB. In total, 110 healthcare professionals from 38 different European HCPs received the questionnaire. All the healthcare professionals who received the questionnaire are involved in reference centres for rCTDs, specifically, for APS, EDS, IIM, IgG4, MCTD, RP, SS, SLE, SSc and UCTD.

ERN ReCONNET survey on clinical practice guidelines knowledge and awareness in rare and complex connective tissue disorder patients, families and caregivers

The main purpose was to assess the knowledge and awareness of CPGs. The survey was codesigned in English with the active participation of the ERN ReCONNET ePAGs Representatives. Their involvement in the creation of the questions was particularly important in order to ensure that the language used was understandable and clear for most patients, caregivers and family members. The questions comprehended different types of answers (single or multiple-choice, Likert scale, open answer, etc).

In order to reach a wider community of patients, caregivers and family members, a comprehensive list of the existing European patients' organisations covering one or more rCTDs covered by ERN ReCONNET was created together with the ERN ReCONNET ePAGs representatives. The survey was submitted to a total of 98 rCTDs patients' organisations, which disseminated the survey among their members together with the support of EURORDIS, the federation of European rare diseases patients' organisations, that also contributed to spreading the survey in their communication channels.

The survey was developed to explore not only the awareness of CPGs, but also to collect the views of rCTDs patients, caregivers and family members on their knowledge of the purpose of CPGs, knowledge of the process of developing CPGs and to gather their perceptions and expectations. Specifically, the survey was designed to collect the feedback of patients, caregivers and family members living with one (or more) of the following diseases: APS, EDS, IIM, IgG4, MCTD, RP, SS, SLE, SSc and UCTD.

The survey consisted of 21-item questions in English, subdivided into three sections (general: demographics, level of education, disease, etc; CPGs knowledge and awareness; subjective perspective of CPGs).

RESULTS

ERN-ReCONNET survey on the use of CPGs for HCPs

Of the 110 healthcare professionals who received the survey, 56 completed the questionnaire (response rate 51%) mostly from Italy (36% of the respondents), France (16%) and the Netherlands (10%). Ninety-one percent of the respondents belonged to University Hospital and 77% see more than 450 patients per year.

As summarised in table 1, the majority of the respondents (96%) considered that using well-constructed CPGs would improve patient care and 93% found it helpful to apply CPGs in clinical practice. With regard to the existence of CPGs in the area of rCTDs (specifically, on APS, EDS, IIM, IgG4, MCTD, RP, SS, SLE, SSc and UCTD), the respondents provided irregular answers. Some respondents indicated that CPGs in the area of rCTDs are very few or lacking, while others reported that CPGs are well constructed and valid. The variability of the answers for these questions is related to the fact that for some rCTDs (as in the case of SLE, SS, SSc and IIM), high-quality CPGs are already in place, while for other rCTDs (such as RP and IgG4), CPGs are not yet available since the evidence on these diseases is still limited. Regarding the use of CPGs in routine clinical practice, 82% of the respondents use CPGs always or more than once a week, 16% less than once a week, while less than 2% never use CPGs when assessing patients. CPGs are more frequently used in the diagnosis (82%) and treatment (88%) and slightly less (67%) in monitoring patients with rCTDs. Sixty-two percent of the respondents experienced difficulties and/or barriers to the application of CPG in their centres and the main reasons were procedures and drug reimbursement (57%), time limit (49%), local legislative restrictions (34%), awareness/knowledge (20%) and poor dissemination of guidelines (17%).

When an open question was asked for a proposal to increase the adherence to CPGs, the main suggestions from the respondents were related to the need of a higher feasibility in clinical practice, in the development of actions to modify local legislative restrictions, and in the creation of easily accessible versions and education and dissemination activities.

ERN ReCONNET survey on CPGs knowledge and awareness in rare and complex connective tissue disorder patients, caregivers and family members

Four hundred and ninety-three anonymous responses were received from patients, caregivers and family members. The majority of respondents were suffering or taking care of patients with SLE (51%), SSc (19%) and SS (16%). The other diseases were represented as shown in table 2 and it is important to mention that patients' organisations exist only for some rCTDs in Europe; therefore, it is more difficult to reach the rarest rCTDs communities (such as IgG4). The question 'What condition/disease do you live with?' was specifically designed to enable the respondents to select more than one disease, in order to include eventual comorbidities. Most respondents were aware of the purpose of CPGs and how they are developed. However, only 62% of responders were aware of the existence of CPGs for their disease and 61% had actually read CPGs produced for their disease. Being aware of the CPGs developed for their disease made patients feel more empowered in the healthcare decisions (93%). Moreover, 95% of the respondents thought that the creation of a patient-friendly version of CPGs would be useful, especially considering that CPGs are generally developed in a complex medical language and therefore not accessible to patients, caregivers and family members. The creation of empowering tools was perceived to have a high impact, in particular, while discussing treatment options and on the general management of the disease.

The development of patients' lifestyle guidelines has collected great interest in 95% of the respondents. Many comments suggested that patients' lifestyle guidelines should be included in the CPGs to inform both patients and clinicians on how to better cope with the disease and on practical tips for daily life. Suggestions of topics to be included in the patients' lifestyle guidelines are guidance for caregivers, recommendations on nutrition and sport, how to maintain a good quality life, how to deal with pain and side effects, and practical everyday do's and don'ts.

In addition, many comments reported the need for CPGs to be fully applicable in clinical practice. This is perceived as achievable only if each CPGs are subject to local adaptations in each European country. According to the respondents, the local adaptations would enable clinicians to better apply those guidelines in their own country and improve the quality of care provided to their patients.

CPGs were also perceived as a potential tool to harmonise the clinical approach to the management of rCTDs and to provide all patients with a uniformed access to medicinal products and therapeutic interventions in all European countries.

It is also important to mention that the majority of respondents recur to healthcare professionals both for healthcare information and for CPGs. On the other hand, patients' organisations and internet/social media are also used while looking for healthcare information. Different comments have, in fact, highlighted the difficulty to

Table 1 Results of the ERN ReCONNET survey on clinical practice guidelines adherence in rare and complex connective tissue disorders: the perspective of healthcare professionals

ERN-ReCONNET survey on the use of clinical practice guidelines for HCPs

Instructions

The survey should take less than 10 minutes, and your responses are completely anonymous.
You can only take the survey once and all questions are required.
The survey is designed to be compled by experts of the following diseases:
We really appreciate your input!

- **Antiphospholipid syndrome (APS)**
- **Ehlers-Danlos syndrome (EDS)**
- **Idiopathic inflammatory myopathies (IIM)**
- **IgG4-related disease (IgG4)**
- **Mixed connective tissue diseases (MCTD)**
- **Relapsing polychondritis (RP)**
- **Sjögren's syndrome (SS)**
- **Systemic lupus erythematosus (SLE)**
- **Systemic sclerosis (SS)**
- **Undifferentiated connective tissue disease (UCTD)**

Results	
Your age	%
<25 years	–
25–34 years	21.4
35–44 years	33.9
45–54 years	26.8
55–64 years	17.9
>65 years	–
Where do you work?	%
Austria	–
Belgium	6
Bulgaria	–
Croatia	–
Cyprus	–
Czech Republic	–
Denmark	–
Estonia	–
Finland	–
France	16
Germany	8
Greece	–
Hungary	–
Ireland	–
Italy	36
Latvia	–
Lithuania	–
Luxembourg	–
Malta	–
Netherlands	10
Norway	–
Poland	2
Portugal	12
Romania	4
Slovakia	–
Slovenia	4
Spain	2
Sweden	–

Continued

Table 1 Continued

United Kingdom	–
Please select the option that best suits your healthcare provider (HCP)	%
General hospital	3.6
University hospital	91.1
Private hospital/healthcare service	1.8
Academic unit affiliated with general hospital	1.8
Mix of general and university hospital	1.8
What is the volume of your centre? (the average number of rCTDs patients seen per year)	%
Fewer than 150 yearly	3.6
150–250 yearly	3.6
250–350 yearly	10.7
350–450 yearly	5.4
More than 450 yearly	76.8
How many years have you been practising your specialty?	
<5	19.6
5–10	21.4
10–15	16.1
>15	42.9
How many physicians in your centre are actually working on rCTDs patients?	
<3	3.5
3–5	16.1
5–10	41.1
>10	39.3
What is/are your main area of expertise? (more than one option possible)	%
APS	46.4
EDS	3.6
IIM	42.9
IgG4	14.3
MCTD	44.6
RP	16.1
SS	44.6
SLE	75.0
SSc	62.5
UCTD	39.3
Using well-constructed CPGs will improve patient care *From 1 (strongly disagree) to 5 (strongly agree)*	%
1	–
2	1.8
3	1.8
4	33.9
5	62.5
CPGs would not improve the care I give to patients *From 1 (strongly disagree) to 5 (strongly agree)*	%
1	41.1
2	42.9
3	7.1
4	8.9
5	0.0

Continued

Table 1 Continued

I find it helpful to apply CPGs in clinical practice *From 1 (strongly disagree) to 5 (strongly agree)*	%
1	0
2	1.8
3	5.4
4	50.0
5	42.8
Existing CPG in my area of expertise are very few or lacking *From 1 (strongly disagree) to 5 (strongly agree)*	%
1	3.6
2	17.9
3	33.9
4	37.5
5	7.1
Existing CPG in my area of expertise are well constructed and valid *From 1 (strongly disagree) to 5 (strongly agree)*	%
1	1.8
2	30.4
3	33.9
4	32.1
5	1.8
How often do you use CPG when *assessing* patients?	%
Never	1.8
Less than once a week	16.1
More than once a week	37.5
Always	44.6
How often do you use CPG when *diagnosing* patients	%
Regularly	82.1
Occasionally	10.8
Never	7.1
How often do you use CPG when considering *treatment* options?	%
Regularly	87.5
Occasionally	12.5
Never	–
How often do you use CPGs when *monitoring* patients?	%
Regularly	67.9
Occasionally	30.4
Never	1.7
Do you experience any difficulties/barriers to the application of CPG in your HCP?	%
Yes	62.5
No	37.5
If yes, please indicate which difficulties/barriers you experience to the application of clinical practice guidelines in your healthcare provider (more than one option possible)	%
Procedures and drug reimbursement	57.1
Time limit	48.6
Local legislative restrictions	34.3
Awareness/knowledge	20.0
Poor dissemination of guidelines	17.0
Do you have any proposal to increase the adherence to the clinical practice guidelines? (open question)	

CPGs, clinical practice guidelines; rCTDs, rare and complex connective tissue and musculoskeletal diseases.

Table 2 Results of the ERN ReCONNET survey on clinical practice guidelines knowledge and awareness in rare and complex connective tissue disorders: patients, families and caregivers

ERN ReCONNET survey on clinical practice guidelines knowledge and awareness in rare and complex connective tissue disorder patients, families and caregivers

Introduction

The European Reference Network for rare and complex connective tissue and musculoskeletal diseases (ERN ReCONNET) is a virtual network involving 26 centres of expertise across Europe that aims at developing a comprehensive and harmonised approach to rare and complex autoimmune and hereditary connective and musculoskeletal diseases (rCTDs).

The ERN ReCONNET is conducting a series of surveys to investigate how much patients, families and caregivers know about clinical practice guidelines on rare and complex autoimmune and hereditary connective and musculoskeletal diseases in Europe.

In the next questions, we are going to ask you an honest opinion on clinical practice guidelines. Please do not worry if you do not understand certain questions or terms used, that is exactly what we are investigating in, it is really important that your answers are sincere and genuine.

Instructions

The survey should only take less than 10 minutes, and your responses are completely anonymous. You can only take the survey once and all questions are required, please make sure you donot complete the questionnaire twice.

The survey is designed to be completed by patients affected, or by family member and caregivers that live/take care of a patient affected by one or more of the following diseases:

Please note that this survey has been developed for patients, caregiver and family members who live in EU countries (Austria, Belgium, Bulgaria, Croatia, Cyprus, Czech Republic, Denmark, Estonia, Finland, France, Germany, Greece, Hungary, Ireland, Italy, Latvia, Lithuania, Luxembourg, Malta, Netherlands, Poland, Portugal, Romania, Slovakia, Slovenia, Spain, Sweden, United Kingdom) and Norway.

In case you are not living in one of the countries listed above, we kindly ask you not to answer the survey. Please note also that the results of this survey may be used in a brief scientific report/abstract to conferences.

If you have any questions about the survey, or about the ERN ReCONNET, please email us: ern.reconnet@ao-pisa.toscana.it

We really appreciate your input!

- **Antiphospholipid syndrome (APS)**
- **Ehlers-Danlos syndrome (all except vEDS)**
- **Idiopathic inflammatory myopathies (including PM and/or DM)**
- **IgG4-related disease**
- **Mixed connective tissue diseases (MCTD)**
- **Relapsing polychondritis (RP)**
- **Sjögren's syndrome**
- **Systemic lupus erythematosus (lupus/SLE)**
- **Systemic sclerosis (scleroderma, SS)**
- **Undifferentiated connective tissue disease (UCTD)**

Results	
Sex	%
Males	5
Females	95
How old are you?	%
Under 25 years	5.8
25–34 years	15.9
35–44 years	24.7
45–54 years	28.7
55–64 years	17.3
Over 65 years	7.6
What is the highest level of school that you have finished?	%
Below high school diploma	9.5
High school diploma	27.6
Bachelor's degree	29.5
Higher	26.3
Other	7.1
Where do you live?	%
Austria	0.2

Continued

Table 2 Continued

Belgium	11.6
Croatia	0.4
Cyprus	0.2
Denmark	4.5
Finland	2.6
France	20.5
Germany	7.8
Greece	0.9
Hungary	0.4
Ireland	4.7
Italy	12.1
Latvia	0.2
Lithuania	0.6
Luxembourg	0.2
Netherlands	5.2
Norway	0.4
Poland	0.2
Portugal	8.0
Spain	0.4
Sweden	0.9
United Kingdom	16.6
Other	1.4
What condition/disease do you live with?*	%
APS	10.4
EDS	15.6
IIM	1.2
IgG4	1.0
MCTD	7.0
RP	1.7
SS	15.8
SLE	50.8
SSc	19.3
UCTD	3.3
Other diseases	21.6
Are you involved in or a member of any patients' organisation/charity?	%
Yes	52.5
No	47.5
Do you know the purpose of clinical practice guidelines?	%
Yes	51.7
No	48.3
Do you know how a clinical practice guideline is developed?	%
Yes	71.3
No	28.7
Are you aware of any clinical practice guideline developed for your disease?	%
Yes	38.2
No	61.8
Have you ever read a clinical practice guideline for your disease?	%
Yes	39.5
No	60.5
If a clinical practice guideline for the treatment of your disease exists, would you use it to discuss with your doctor, friends and family?*	%

Continued

Table 2 Continued

Doctor	90.9
Friends/family	62.4
Other	5.4
What do you expect from a clinical practice guideline? (open question)	
What do you think clinical practice guidelines are developed for? (open question)	
Where do you go for healthcare information?*	%
Healthcare professionals	82.4
Internet/social media	65.4
Patients' organisations	54.5
Friends and family	8.5
Other	6.0
Do you perceive clinical practice guidelines as something positive or negative?	%
Positive	82.7
Negative	0.4
Not sure/donot know	16.9
Do you think that patients' perspective is properly included in the clinical practice guidelines development?	%
Yes	42.9
No	39.1
Other	18.0
Do you think that specific clinical practice guidelines on patients' lifestyle could be useful for the management of your disease?	%
Yes	89.6
No	5.6
Other	4.8
Do you think that developing a patient-friendly version of the clinical practice guidelines would be useful?	%
Yes	95.4
No	2.1
Other	2.2
In a few words, what kind of content would you like to see in the patient-friendly version of the clinical practice guideline? (open question)	
In your opinion, would you feel more empowered in the healthcare decisions if you were properly aware of the clinical practice guidelines developed for your disease?	%
Yes	93.3
No	4.0
Other	2.7
Who would you like to receive information on clinical practice guidelines from?*	%
European Reference Networks (ERNs)	45.6
Patients' organisations	71.4
Healthcare professionals	80.1
Scientific societies	33.0
Internet/social media	40.5

*Multiple-choice question.

identify reliable sources of information, especially on the internet.

DISCUSSION

The results of these surveys provide a first picture of the level of awareness as well as a first overview of the level of adherence to CPGs in rare and complex musculoskeletal and connective tissue diseases. Considering the critical impact that guidelines have in the care provided to patients, it is crucial that a network of expertise on rare diseases takes a snapshot of what clinicians and patients think about CPGs. The main message from the HCPs is that using well-constructed CPGs surely improves patient care. Well-constructed and valid CPGs exist for some rCTDs, but others have very few or no CPGs available, and in fact, this was also reflected in the variability of the responses. A large part of the enquired HCPs uses CPGs in routine clinical practice; however, some difficulties and barriers still exist, and they are mainly caused

by local legislative restrictions and time constraints during the assessment of patients. Many proposals have been suggested to implement the application of CPGs, mainly related to (i) having easier and more practical versions of existing CPGs (such as phone and/or computer applications or an online platform), (ii) increasing knowledge and education for both patients and professionals on CPGs, (iii) having more dedicated time during the routine clinical assessment and (iv) having government support and reduce any local legislative restrictions.

The voices of patients, family members and caregivers underlined that CPGs should be more widely disseminated and adapted to the different health systems in Europe in order to improve the level of care provided to patients. CPGs are generally perceived positively, but it is evident that more effort should be dedicated to the inclusion of patients in the development of CPGs. The expectations of respondents on CPGs were focused on the great benefit that CPGs scan induce on the management of diseases, especially if the patient is affected by a rare disease. Respondents also emphasised that patient-friendly versions of CPGs and patients' lifestyle guidelines could play a relevant role in the process of empowering patients in the management of their disease and to support shared decision with healthcare professionals. The results obtained in this survey will enable ERN ReCONNET to plan initiatives related to patients' empowerment and CPGs, such as the creation of patient-friendly version of CPGs and patients' lifestyle guidelines.

Since ERN ReCONNET aims at improving the level of adherence to CPGs across European HCPs, by providing practical tools and training to healthcare professionals, many of the initiatives emerging from this survey are being realised under the scope of the ERN ReCONNET, following the suggestions, expectations and points of view of the member HCPs and ePAGs. The ERN ReCONNET has already reviewed the landscape of the existing CPGs in rCTDs[10–19] and the existence of partially sufficient CPGs for some rCTDs was highlighted, while demonstrating the absence of evidence-based CPGs for other rCTDs.[10–19] Among the different unmet needs on CPGs, the results raised the issue that more attention should be devoted to the assessment of CPGs adherence in routine clinical practice. In accordance, the ERN ReCONNET aims at identifying strategies to improve the dissemination and implementation of CPGs throughout Europe. Other activities are also ongoing under the scope of the ERN ReCONNET, both to produce the evidence needed to create new CPGs, and to adapt existing CPGs to the different geographical and cultural European contexts and to fulfil the unmet needs and gaps emerged from the survey.

CONCLUSIONS

Well-constructed CPGs improve patient care, but some difficulties and barriers still exist in their implementation in daily clinical activities. Thanks to the engagement of the most relevant stakeholders involved in rCTDs, the ERN ReCONNET is promoting practical actions for the local adaptation of CPGs across Europe, improving their routine clinical use and contributing to the increase of the awareness on CPG across rCTDs patients, family members and caregivers.

Author affiliations
[1]Rheumatology Unit, Azienda Ospedaliero Universitaria Pisana, Pisa, Italy
[2]University of Pisa, Pisa, Italy
[3]Department of Rheumatology and Clinical Immunology, Charité Universitätsmedizin Berlin, Berlin, Germany
[4]Serviço De Reumatologia E Doenças Ósseas Metabólicas, Centro Hospitalar Universitário Lisboa Norte E.P.E, Lisboa, Portugal
[5]Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal
[6]Flemish Association for Hereditary Connective Tissue Disorders in Belgium, Koersel, Belgium
[7]FESCA, Federation of European Scleroderma Associations, Milan, Italy
[8]Département de Médecine Interne et Immunologie Clinique, Centre de Référence des Maladies Auto-Immunes Systémiques Rares du Nord et Nord-Ouest de France, Centre Hospitalier Universitaire de Lille, Lille, France
[9]Department of Rheumatology, Cliniques Universitaires Saint-Luc, Université Catholique De Louvain, Louvain-la-Neuve, Belgium
[10]Department of Rheumatology and Clinical Immunology, Kerckhoff-Klinik GmbH, Bad Nauheim, Germany
[11]Justus Liebig Universitat Giessen, Giessen, Germany
[12]Department of Rheumatology, Universitätsklinikum Düsseldorf, Düsseldorf, Germany
[13]Department of Rheumatology, Ghent University Hospital, Ghent, Belgium
[14]Department of Internal Medicine, Ghent University, Ghent, Belgium
[15]Institute of Management, Scuola Superiore Sant'Anna, Pisa, Italy
[16]Department of Rheumatology and Clinical Immunology, University Medical Center Utrecht, Utrecht, Netherlands
[17]Núcleo Síndrome De Sjögren, Liga Portuguesa Contra as Doenças Reumáticas, Lisbon, Portugal
[18]Department of Internal Medicine, University of Genoa, Genova, Italy
[19]Research Laboratory and Academic Division of Clinical Rheumatology, IRCCS Polyclinic Hospital San Martino, Genova, Italy
[20]Rheumatology Unit, University of Pisa, Pisa, Italy

Twitter Charissa Frank @charissafrank.

Acknowledgements The authors want to thank all the members of ERN ReCONNET, including all ePAGs and all the rCTDs patients' organisations that contributed to the dissemination of the survey, all HCPs and the ERN ReCONNET team for their precious contribution and commitment in the realisation of this work.

Contributors All the authors contributed to the realisation of the work.

Funding This publication was funded by the European Union's Health Programme (2014–2020).

Competing interests None declared.

Patient consent for publication Not required.

Provenance and peer review Not commissioned; externally peer reviewed.

Data availability statement Data sharing not applicable as no datasets generated and/or analysed for this study.

ORCID iDs

Gerd Burmester http://orcid.org/0000-0001-7518-1131

Eric Hachulla http://orcid.org/0000-0001-7432-847X

REFERENCES

1 Available https://ec.europa.eu/health/ern_en
2 Available https://ec.europa.eu/health/ern/events/ev_2010309_en
3 Available http://reconnet.ern-net.eu/
4 Consensus report, Institute of Medicine. Clinical practice guidelines we can trust (accessed 23 Mar 2011)
5 Dixon-Woods M, Agarwal S, Young B, *et al. Integrative approaches to qualitative and quantitative evidence*. London: Health Development Agency, 2004.
6 Graham H, Kelly MP. *Health inequalities: concepts, frameworks and policy*. London: Health Development Agency, 2004.
7 Kelly MP, Stewart E, Morgan A, *et al.* A conceptual framework for public health: NICE's emerging approach. *Public Health* 2009;123:e14–20.
8 Kelly MP, Morgan A, Ellis S, *et al.* Evidence based public health: a review of the experience of the National Institute of Health and Clinical Excellence (NICE) of developing public health guidance in England. *Social Sci Med (1982)* 2010;71:1056–62.
9 Lomas J, Culyer T, McCutcheon C, *et al. Conceptualizing and combining evidence for health system guidance: final report*. Ottawa: Canadian Health Services Research Foundation, 2005.
10 Limper M, Scirè CA, Talarico R, *et al.* Antiphospholipid syndrome: state of the art on clinical practice guidelines. *RMD Open* 2018;4:e000785.
11 Sulli A, Talarico R, Scirè CA, *et al.* Ehlers-Danlos syndromes: state of the art on clinical practice guidelines. *RMD Open* 2018;4:e000790.
12 Iaccarino L, Talarico R, Scirè CA, *et al.* IgG4-related diseases: state of the art on clinical practice guidelines. *RMD Open* 2019;4:e000787.
13 Meyer A, Scirè CA, Talarico R, *et al.* Idiopathic inflammatory myopathies: state of the art on clinical practice guidelines. *RMD Open* 2019;4:e000784.
14 Chaigne B, Scirè CA, Talarico R, *et al.* Mixed connective tissue disease: state of the art on clinical practice guidelines. *RMD Open* 2019;4:e000783.
15 Rednic S, Damian L, Talarico R, *et al.* Relapsing polychondritis: state of the art on clinical practice guidelines. *RMD Open* 2018;4:e000788.
16 Tamirou F, Arnaud L, Talarico R, *et al.* Systemic lupus erythematosus: state of the art on clinical practice guidelines. *RMD Open* 2019;4:e000793.
17 Romão VC, Talarico R, Scirè CA, *et al.* Sjögren's syndrome: state of the art on clinical practice guidelines. *RMD Open* 2018;4:e000789.
18 Smith V, Scirè CA, Talarico R, *et al.* Systemic sclerosis: state of the art on clinical practice guidelines. *RMD Open* 2019;4:e000782.
19 Antunes M, Scirè CA, Talarico R, *et al.* Undifferentiated connective tissue disease: state of the art on clinical practice guidelines. *RMD Open* 2019;4:e000786.