

The Fifth International Symposium on IgG4-Related Disease
4-6 April 2024
Milan, Italy

Title: Understanding the Needs of the IgG4-Related Disease Community by Taking a Patient-Centric Approach

Authors: Maria Picone¹, Francesca Milano², Christopher DeFelice¹, Valentina Di Mattei^{3,4}, Martina Mazzetti³, Kelly Gavigan⁵, Giorgia Casasco⁴, Esteban Rivera⁵, Antonio Catarinella^{3,4}, Erik Stone⁵, Marco Lanzillotta^{3,6}, Alessandra Santini⁷, John H. Stone⁸, Ilaria Galetti⁹, Emanuel Della Torre^{3,6}

Affiliations:

¹ TREND Community, Philadelphia, Pennsylvania, USA

² Department of Psychology, University of Milano-Bicocca, Milan, Italy

³ Clinical and Health Psychology Unit, IRCCS San Raffaele Scientific Institute, Milan, Italy

⁴ School of Psychology, Vita-Salute San Raffaele University, Milan, Italy

⁵ Global Healthy Living Foundation, Upper Nyack, New York, USA

⁶ Unit of Immunology, Rheumatology, Allergy, and Rare Diseases (UnIRAR), IRCCS San Raffaele Scientific Institute, Milan, Italy

⁷ Dynamicom Education, Milan, Italy

⁸ Division of Rheumatology, Allergy and Immunology, Massachusetts General Hospital, Harvard Medical School, Boston, Massachusetts, USA

⁹ Federation of European Scleroderma Associations (FESCA), Belgium, Italy

SCOPE OF WORK:

An inaugural congress was held in Milan, Italy, in October 2023 for members of the IgG4-related disease (IgG4-RD) community. Patients and caregivers participated in a series of roundtable discussions in Italian focusing on quality of life (QoL) and provided a valuable lens into the IgG4-RD experience. Additionally, 146 patients affected by IgG4-RD participated in a structured survey with questions designed to interrogate specific aspects of their experience. From these separate approaches emerges a series of challenges which were described both qualitatively and quantitatively.

MATERIALS AND METHODS:

At the congress, 50 patients split into groups to discuss and identify sources of concern within 5 predetermined QoL areas: subjective experience, limitations in activities, emotional difficulties, socioeconomic impact, and unmet needs. After each of 5 groups identified 4 subthemes within their QoL area, the groups reunited and voted on which of the 4 was the most impactful to them in each area. This same process was replicated for the 40 caregivers who attended. In a separate

but aligned effort, an online survey in Italian was developed and disseminated to patients with IgG4-RD who attend a clinic in Italy to better understand the patient journey and assess unmet needs.

RESULTS:

Within the emotional difficulties area of QoL, 51% of congress patients selected “new reactivations and worsening of QoL” as their top concern. In parallel, caregivers highlighted emotional burdens related to the “uncertainty of the future,” “the burden of caretaking and caretaking in the best way,” “a sense of helplessness,” and “loneliness and confusion in the path to care.” In other QoL areas, they also mentioned the “limited ability to plan future events” and the “need for increased psychological/emotional support.”

Proper access to information in the context of an illness is associated with a greater sense of agency and improved stress management. However, 64% of the 137 survey respondents said it took more than 1 month to find information about IgG4-RD after the first symptoms or after diagnosis, and 34% said it took more than 6 months. In the roundtable, the need for improved access to and quality of information arose multiple times.

CONCLUSIONS:

Using these 2 separate tools, several themes and unmet needs were identified that remain of interest to this community. Building more connections and offering more meeting opportunities would be impactful ways to support the community. Increasing the degree of clinical coordination across multiple disciplines would also benefit this population, especially for those traveling long distances for care. In the roundtables, both patients and caregivers described the need to take off work, and 38% of surveyed patients live more than 50 km from the specialist(s) they visit.

	Greatest Interest or Concern in Each QoL Area			
	Patients	%	Caregivers	%
Subjective experience	Understanding the cause of the disease	30	Lack of information for proper support in the disease journey	48
	Uncertainty about the future	28	Need for continuity of care	28
	Impact of incorrect diagnoses on symptoms and treatment	26	Opportunity for sharing and support in the disease journey	20
	Loss of autonomy	16	Limited involvement in emotional dynamics	5
Limitations in activities	Decrease in energy in work activities	47	Anxiety and fatigue	38
	Limitation of social activities	28	Limitation in planning future activities	38
	Limitation/interruption of leisure activities	13	Limitation in travel and social activities	15
	Reduction in sexual desire	13	Limitation in food preparation/consumption	8
Emotional difficulties	Concerns about new reactivations and worsening of quality of life	51	Uncertainty about the future	28
	Work-related concerns related to a decrease in physical fitness, professional dissatisfaction, and a reduction in self-esteem	30	Burden of caring for the patient and caring in the best way	28
	No emotional difficulties related to the disease	11	Sense of helplessness	25
	Sharing concerns with family members	9	Confusion and loneliness in the path leading to diagnosis	20
Socioeconomic impact	Economic cost to access a reference center	35	Need for dissemination of information about the disease	50
	Cost of diagnostic tests	26	Nonrecognition of disability	30
	Cost of medications not covered by the national health system	20	Economic problem due to the distance from the reference center	10
	Loss of workdays	20	Involvement of family members (need to take time off work)	10
Unmet needs	Multidisciplinary coordination to organize instrumental tests	47	More events like this (conferences/meetings)	69
	Recognition of the disease for 104 recognition (disability certificate)	28	Psychologist/emotional support	21
	Creation of a protocol	11	Sexual problems	5
	Awareness of the disease	15	Existence as a family member	5

Understanding the Needs of the IgG4-Related Disease Community by Taking a Patient-Centric Approach

Maria Picone¹, Francesca Milano², Christopher DeFelice¹, Valentina Di Mattei^{3,4}, Martina Mazzetti³, Kelly Gavigan⁵, Giorgia Casasco⁴, Esteban Rivera⁵, Antonio Catarinella^{3,4}, Erik Stone⁵, Marco Lanzillotta^{3,6}, Alessandra Santini⁷, John H. Stone⁸, Ilaria Galetti⁹, Emanuel Della Torre^{3,6}

¹ TREND Community, Philadelphia, Pennsylvania, USA

² Department of Psychology, University of Milano-Bicocca, Milan, Italy

³ Clinical Health Psychology Unit, IRCCS San Raffaele Hospital, Milan, Italy

⁴ Clinical and Health Psychology Unit, IRCCS San Raffaele Scientific Institute, Milan, Italy

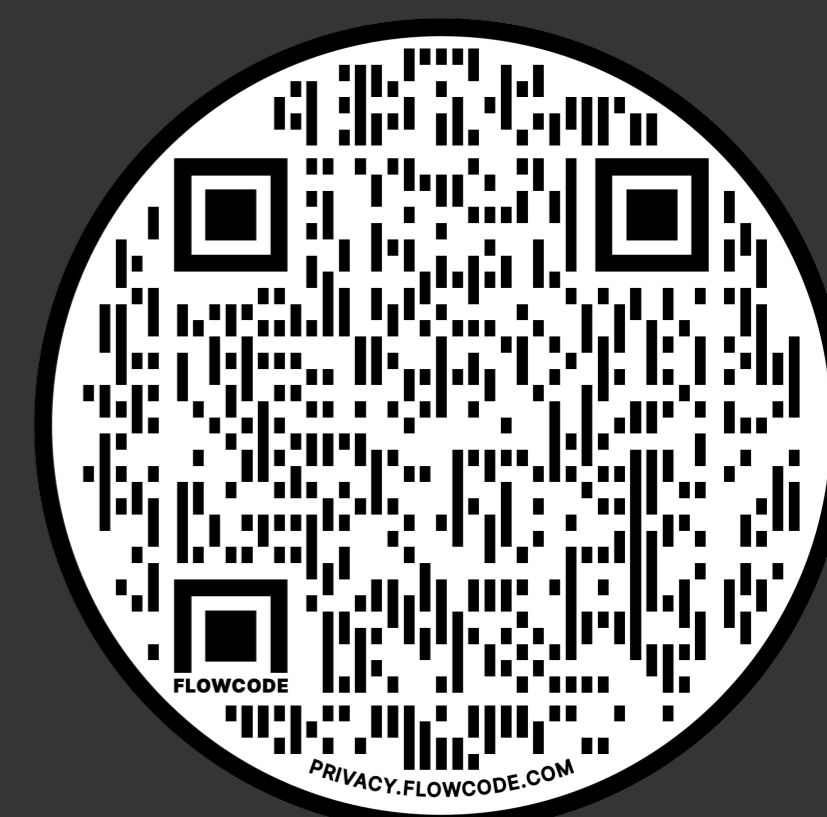
⁵ Global Healthy Living Foundation, Upper Nyack, New York, USA

⁶ Unit of Immunology, Rheumatology, Allergy, and Rare Diseases (UnIRAR), IRCCS San Raffaele Scientific Institute, Milan, Italy

⁷ Dynamicom Education, Milan, Italy

⁸ Division of Rheumatology, Allergy and Immunology, Massachusetts General Hospital, Harvard Medical School, Boston, Massachusetts, USA

⁹ Federation of European Scleroderma Associations (FESCA), Belgium, Italy

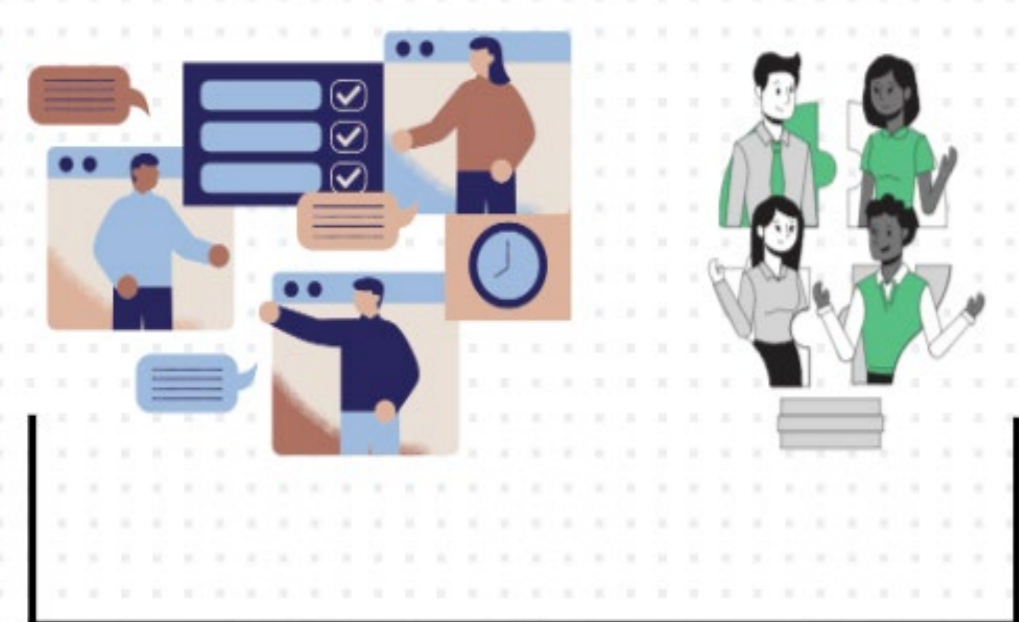


BACKGROUND

Immunoglobulin G4-related disease (IgG4-RD) is a fibroinflammatory disease in which dense accumulations of IgG4 positive plasma cells can lead to organ swelling and end-organ damage. Clinical presentation is diverse but has been shown to fall within four groups: Pancreato-Hepato-Biliary, Retroperitoneum and Aorta, Head and Neck - Limited, and Mikulicz Syndrome and Systemic.^{1,2}

METHODS

Figure 1. The Dual Approach - Survey and Roundtables



An inaugural congress was held in Milan, Italy, in October 2023 for members of the IgG4-related disease (IgG4-RD) community. Patients and caregivers participated in a series of roundtable discussions on the following quality of life (QoL) topics: subjective experience, limitations in activities, emotional difficulties, socioeconomic impact, and unmet needs.

At the congress, 50 patients split into groups to discuss and identify sources of concern within 5 predetermined QoL areas. After each of 5 groups had identified 4 sub themes within their QoL area, the groups reunited and voted on which of the 4 was the most impactful to them in each area. This same process was replicated for the 40 caregivers who attended.

In a separate but aligned effort, 146 Italian IgG4-RD patients participated in a structured online survey designed to better understand the patient journey. From these separate approaches emerged a series of challenges described both qualitatively and quantitatively.

RESULTS

Table 1. Roundtable Results

	Greatest Interest or Concern in Each QoL Area			
	Patients		Caregivers	
		%		%
Subjective experience	Understanding the cause of the disease	30	Lack of information for proper support in the disease journey	48
	Uncertainty about the future	28	Need for continuity of care	28
	Impact of incorrect diagnoses on symptoms and treatment	26	Opportunity for sharing and support in the disease journey	20
	Loss of autonomy	16	Limited involvement in emotional dynamics	5
Limitations in activities	Decrease in energy in work activities	47	Anxiety and fatigue	38
	Limitation of social activities	28	Limitation in planning future activities	38
	Limitation/interruption of leisure activities	13	Limitation in travel and social activities	15
	Reduction in sexual desire	13	Limitation in food preparation/consumption	8
Emotional difficulties	Concerns about new reactivations and worsening of quality of life	51	Uncertainty about the future	28
	Work-related concerns related to a decrease in physical fitness, professional dissatisfaction, and a reduction in self-esteem	30	Burden of caring for the patient and caring in the best way	28
	No emotional difficulties related to the disease	11	Sense of helplessness	25
	Sharing concerns with family members	9	Confusion and loneliness in the path leading to diagnosis	20
Socio Economic impact	Economic cost to access a reference center	35	Need for dissemination of information about the disease	50
	Cost of diagnostic tests	26	Nonrecognition of disability	30
	Cost of medications not covered by the national health system	20	Economic problem due to the distance from the reference center	10
	Loss of workdays	20	Involvement of family members (need to take time off work)	10
Unmet needs	Multidisciplinary coordination to organize instrumental tests	47	More events like this (conferences/meetings)	69
	Recognition of the disease for 104 recognition (disability certificate)	28	Psychologist/emotional support	21
	Creation of a protocol	11	Sexual problems	5
	Awareness of the disease	15	Existence as a family member	5

Table 2. Impact of IgG4-RD on Economic Situation

IMPACT	% (N = 146)
Majorly affected	44
Moderately affected	30
Not at all affected	2
It doesn't apply to my situation	24

Table 3. Impact of IgG4-RD on Social Life

IMPACT	% (N = 146)
Majorly affected	40
Moderately affected	25
Not at all affected	1
It doesn't apply to my situation	34

Figure 2. Distance from Home to Specialist

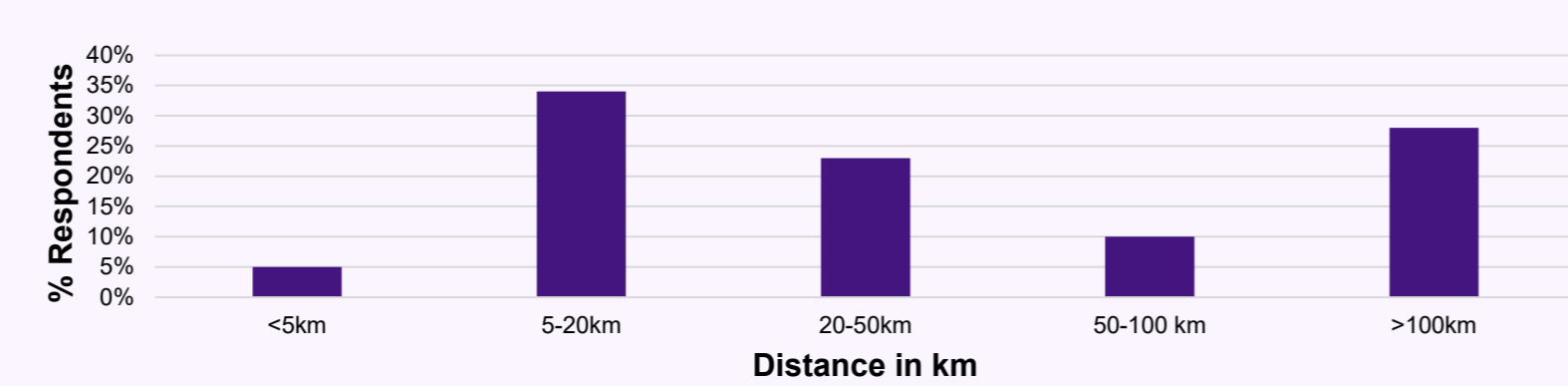
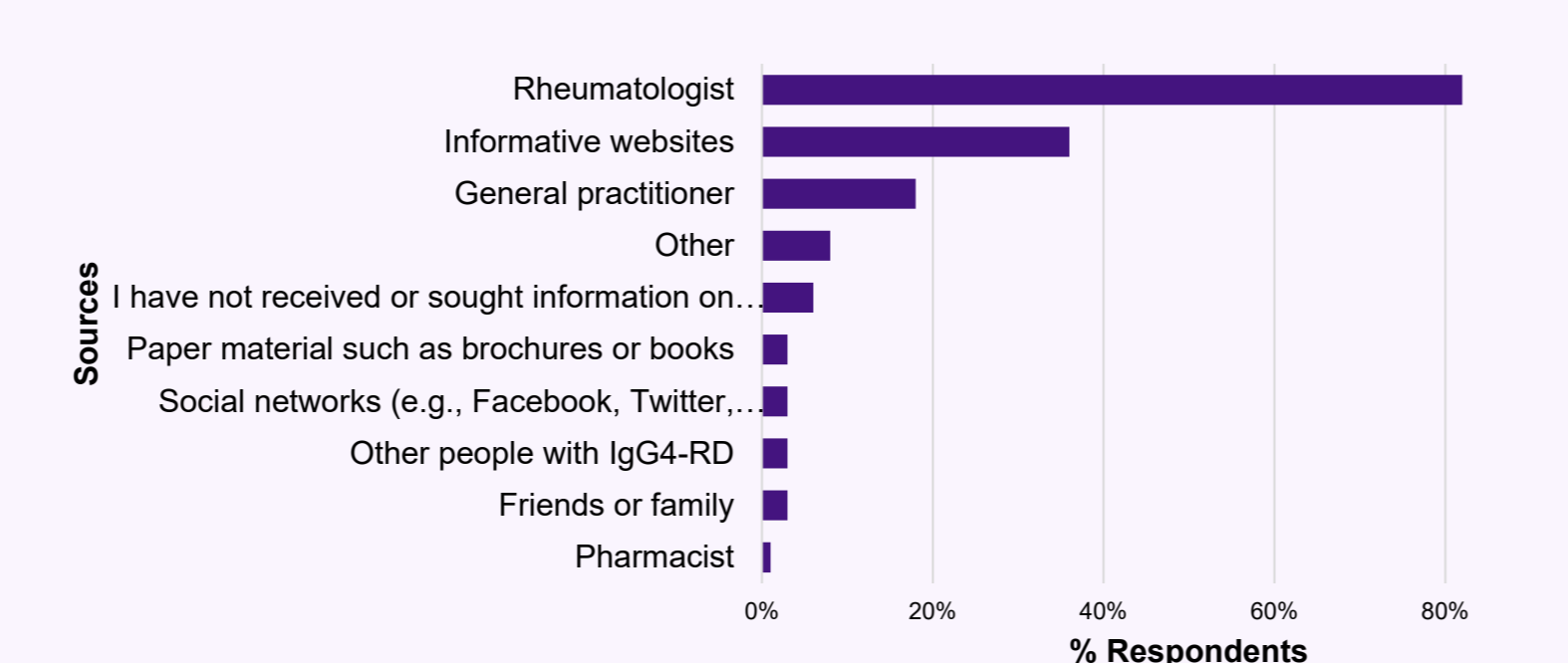


Figure 3. Sources of IgG4 Information



CONCLUSION



Using these 2 separate tools, several themes and unmet needs central to the IgG4-RD community were identified. Building more connections and offering more meeting opportunities would be impactful ways to support the community. Increasing the degree of clinical coordination across multiple disciplines would also benefit this population, especially for those traveling long distances for care. In the roundtables, both patients and caregivers described the need to take off work, and 38% of surveyed patients live more than 50 km from the specialist(s) they visit. Results also suggest opportunities for education via clinician engagement, as the vast majority (82%) rely on their Rheumatologist for information about IgG4-RD.

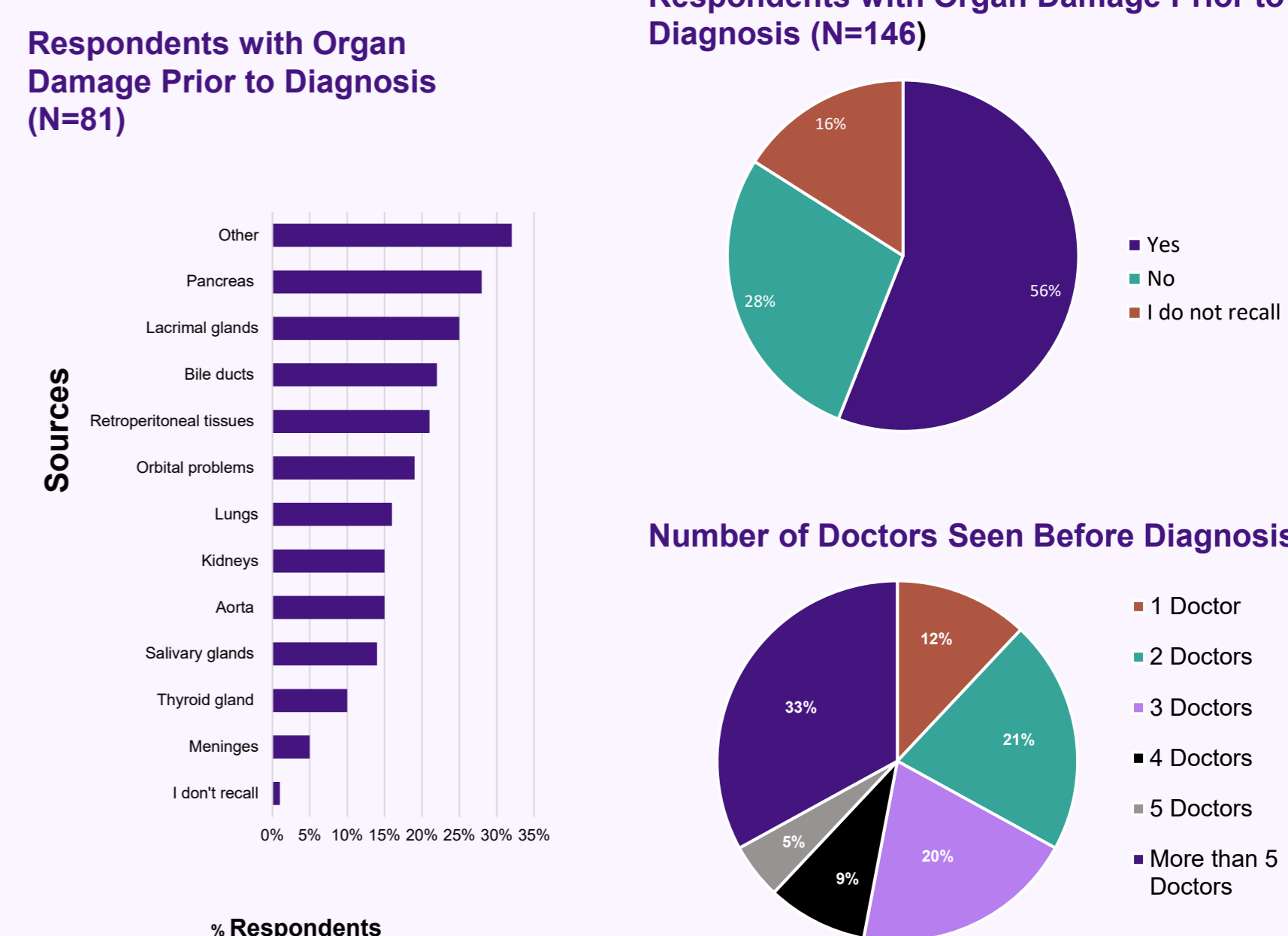
References:

- Wallace ZS, Zhang Y, Perugino CA, et al. Clinical phenotypes of IgG4-related disease: an analysis of two international cross-sectional cohorts. *Ann Rheum Dis.* 2019;78(3):406-412.
- Lanzillotta M, Mancuso G, Della-Torre E. Advances in the diagnosis and management of IgG4 related disease. *BMJ.* 2020;369:m1067.

Within the emotional difficulties area of QoL, 51% of congress patients selected "new reactivations and worsening of QoL" as their top concern. In parallel, caregivers highlighted emotional burdens related to the "uncertainty of the future," "the burden of caretaking and caretaking in the best way," "a sense of helplessness," and "loneliness and confusion in the path to care." In other QoL areas, they also mentioned the "limited ability to plan future events" and the "need for increased psychological/emotional support."

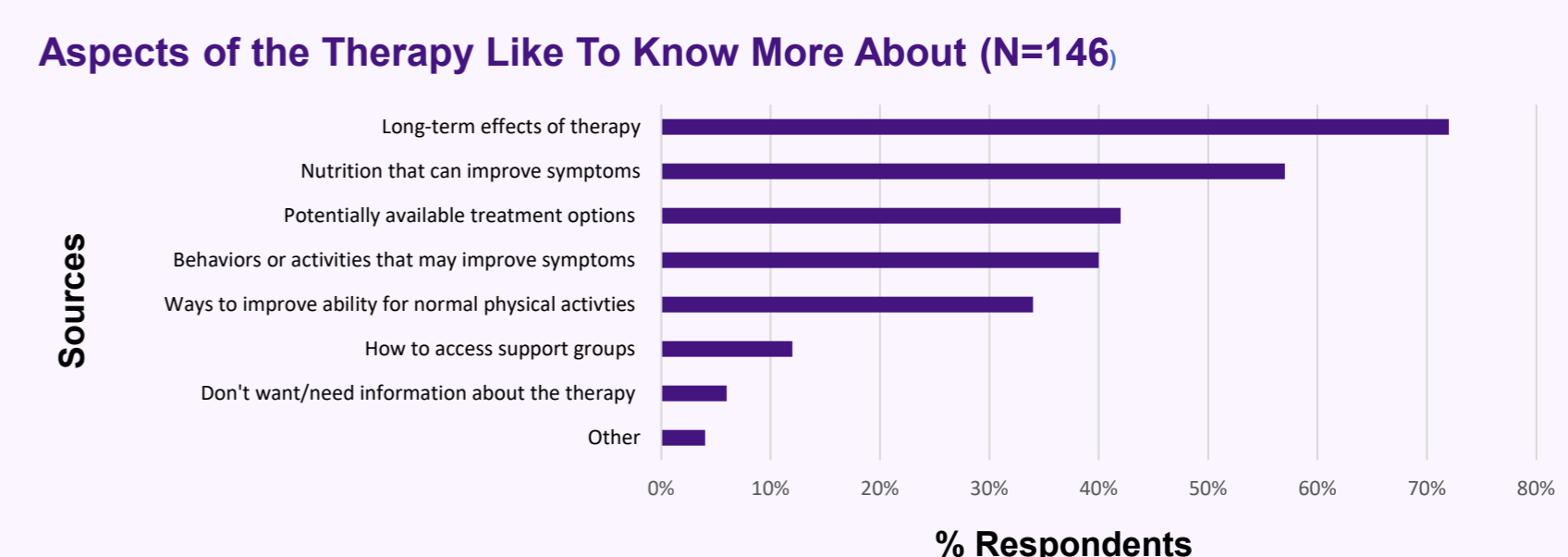
Proper access to information in the context of an illness is associated with a greater sense of agency and improved stress management. However, 64% of the 137 survey respondents said it took more than 1 month to find information about IgG4-RD after the first symptoms or after diagnosis, and 34% said it took more than 6 months. In the roundtable, the need for improved access to and quality of information which arose multiple times.

Figure 4. Before Diagnosis



Prior to diagnosis patients typically saw several doctors with a third of respondents seeing more than 5 doctors before receiving their diagnosis. During this diagnostic delay, the disease can progress as evidenced by over half of respondents having organ damage prior to diagnosis.

Figure 5. Knowledge Gaps



Knowledge gaps were evident from both the survey and the roundtable results. From the survey, figure 5 shows the aspects of therapy that respondents would like to know more about. In the roundtable, caregivers brought up information gaps twice. Once in response to the "subjective experience" prompt, and the other in regards to the need for information dissemination arose in the "socio-economic impact" theme. Online survey results echoed knowledge gaps at the congress, with only 43% of respondents reporting feeling "certain" about possible risks if IgG4-RD is not adequately treated.