

QUALITY OF LIFE IN DESMOID-TYPE FIBROMATOSIS

Development of a disease specific
quality of life questionnaire

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Anne-Rose W. Schut, MD, PhD candidate
Department of Surgical Oncology and
Gastrointestinal Surgery; Department of
Medical Oncology

Erasmus MC
University Medical Center Rotterdam



Cancer Institute

Desmoid-type Fibromatosis (DTF)

Desmoid-type fibromatosis (desmoid tumour / aggressive fibromatosis)

- Rare, soft tissue tumour
- Classified as “intermediate tumour”
- Unpredictable and variable clinical course
- Symptoms vary
- Active surveillance first line treatment
- Overall response to active treatment options remains modest
- Quality of life to guide treatment decision-making and to evaluate treatment efficacy and its consequences

Health-related Quality of Life (HRQoL)

EORTC QLQ-C30:

- *Were you tired?*
- *Do you have any trouble walking al long walk?*

Generic questionnaires:

- Contain items that might be irrelevant for DTF patients
- Do not cover all DTF-specific HRQoL problems

Gounder/DTRF Desmoid Symptom / Impact Scale (GODDESS)



QUALIFIED study

The evaluation of health-related quality of life issues experienced by patients with desmoid-type fibromatosis

Study design:

- International multicentre cross-sectional observational cohort study

Aim:

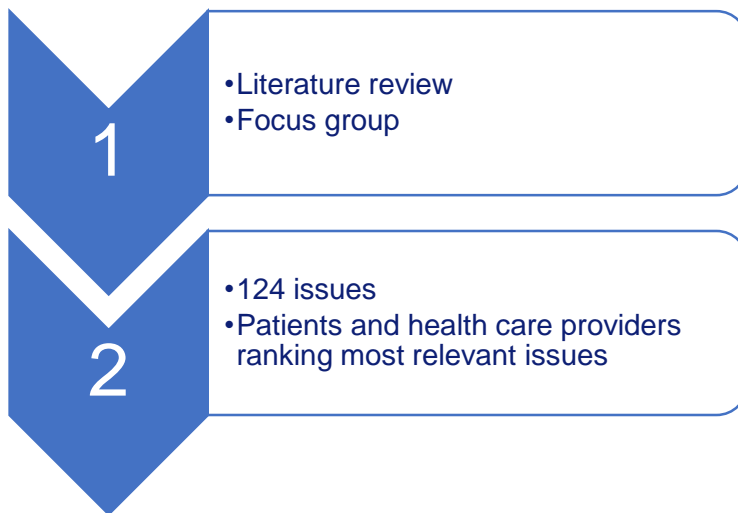
- To develop and pretest a DTF specific HRQoL tool, which can be used alongside the EORTC QLQ-C30

DTF-QoL development

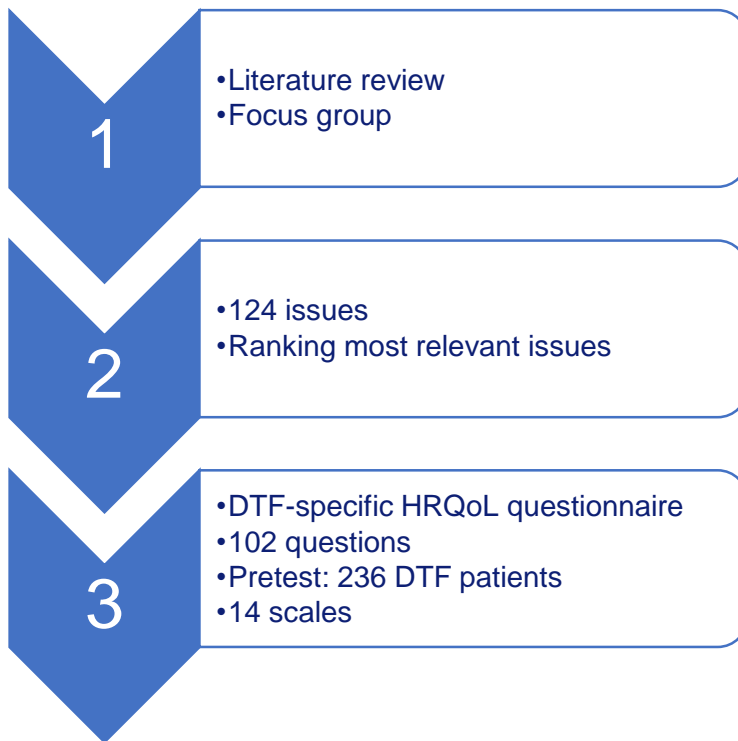
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- Literature review
- Focus groups in UK and NL

DTF-QoL development



DTF-QoL development



DTF-QoL symptom related scales

Emotional and psychological
consequences

Physical consequences

Pain and discomfort

*Have you been
unable to lean on
the tumour sites?
(e.g. due to local
pressure)*

Timeframe: *“During the Past week....”*

DTF-QoL disease impact scales

Worries and concerns

Job and education

Doctor-patient relationship, communication and information

Understanding and support by others

Physical burden and related limitations

Diagnostic and treatment trajectory of DTF

Parents and fertility

Body image and sensation

Supportive care

Concerns around treatment and its consequences

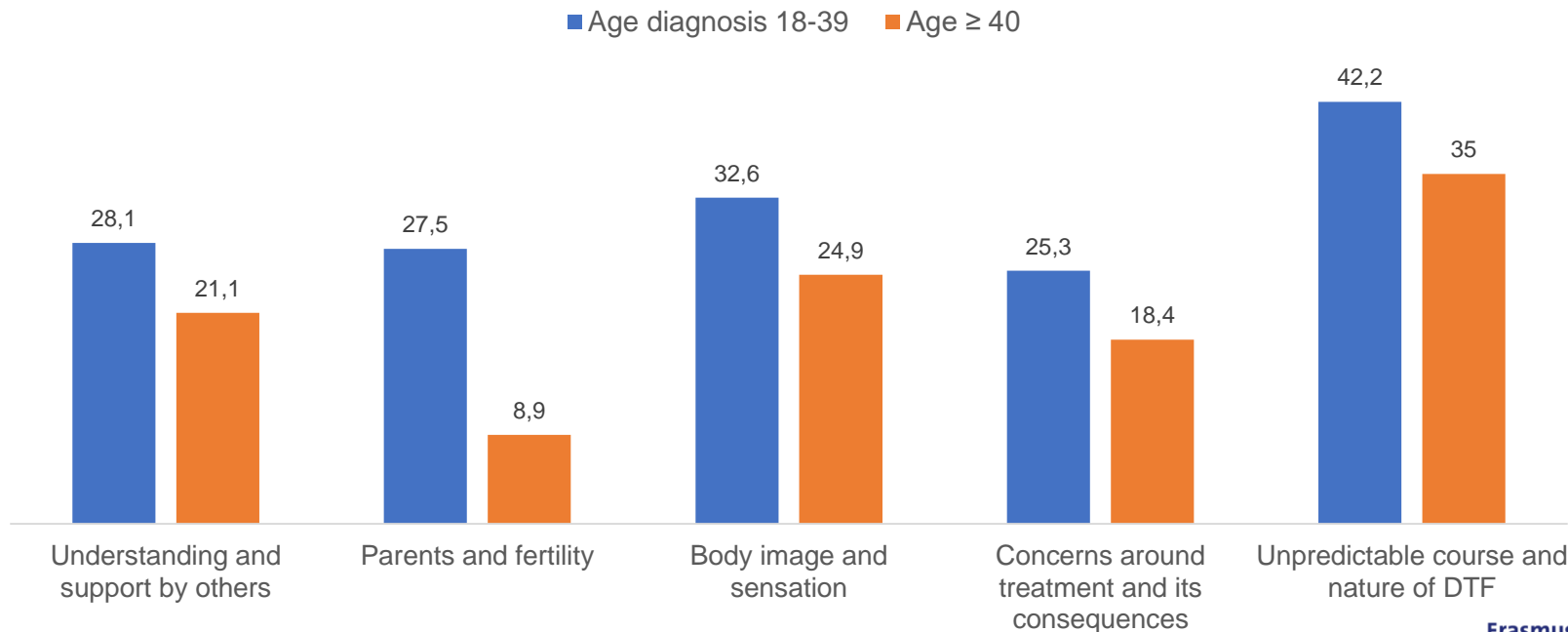
Unpredictable clinical course and nature of DTF

*Have you felt
frustrated
about the
“benign”
diagnosis with
cancerous
features?*

Timeframe: “Since your diagnosis.....”

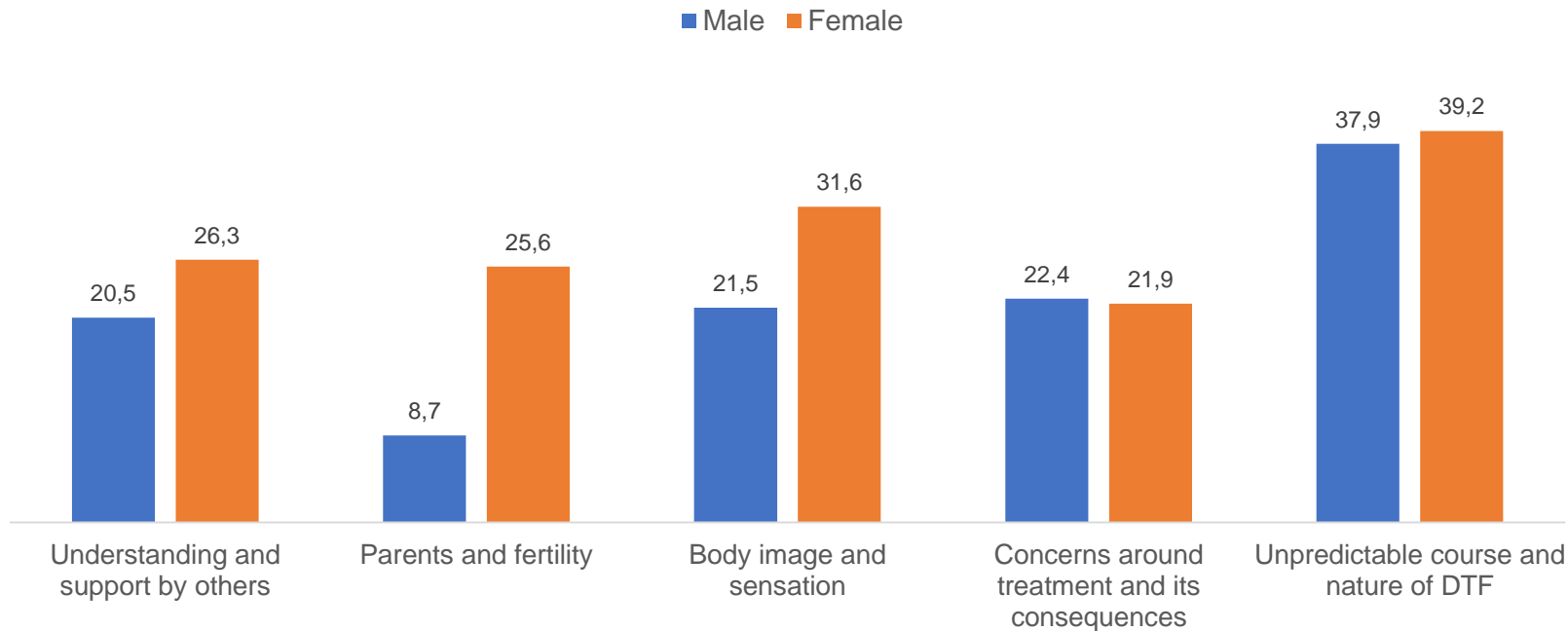
Age

- Scale 0 – 100
- Higher score indicates more symptoms / problems



Gender

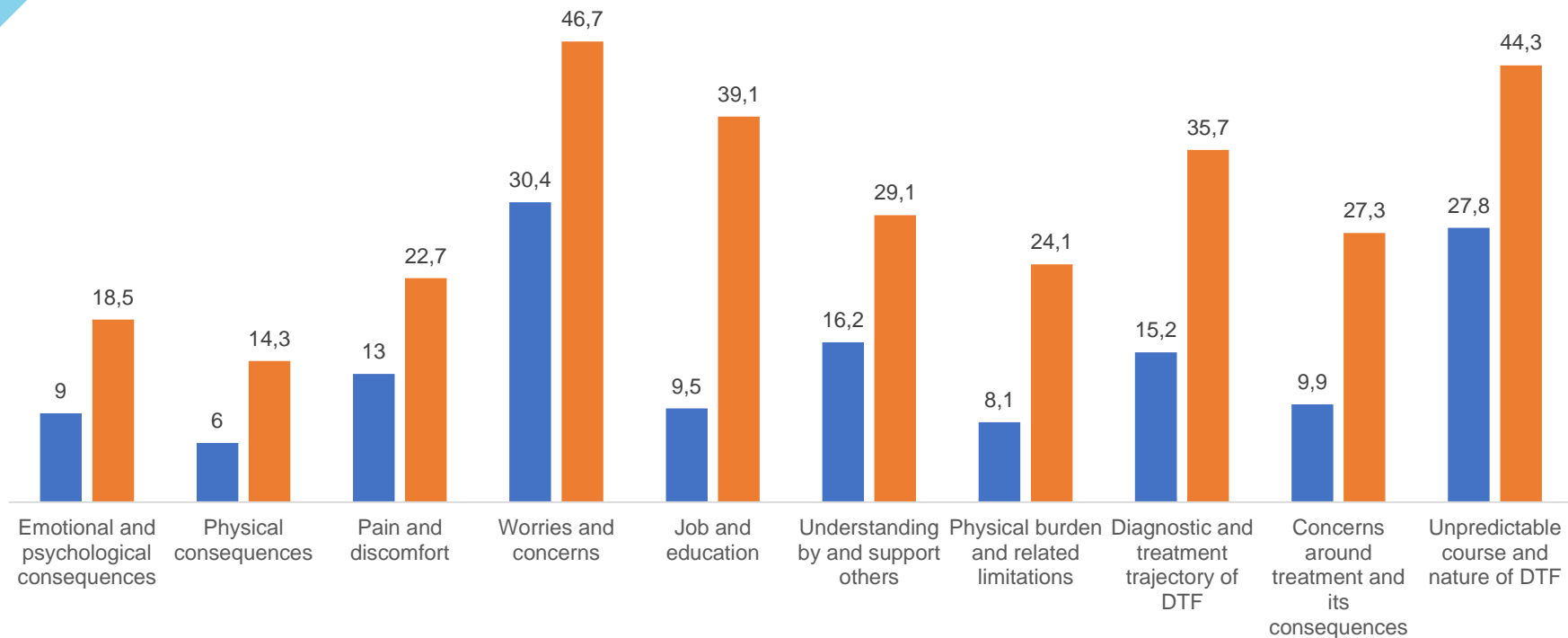
- Scale 0 – 100
- Higher score indicates more symptoms / problems



Treatment

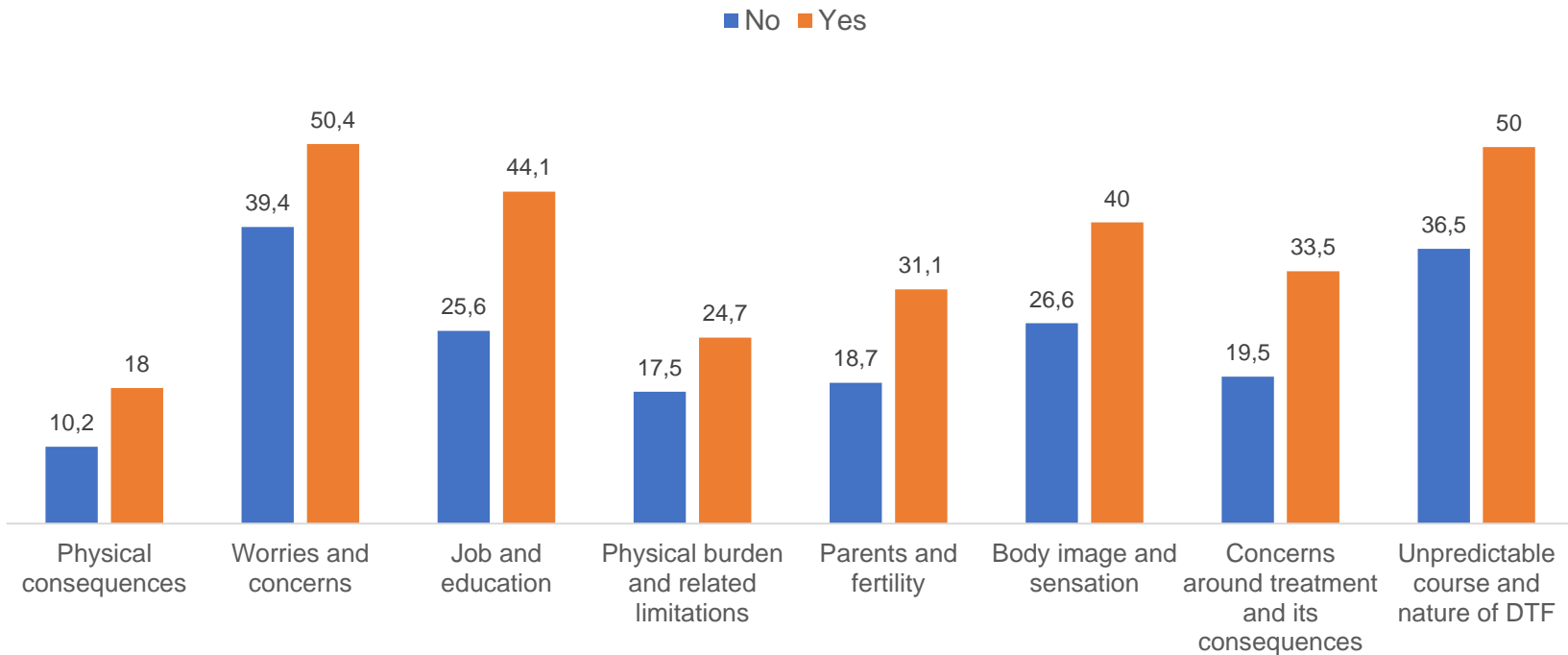
■ Only Wait and See ■ Active treatment

- Scale 0 – 100
- Higher score indicates more symptoms / problems



Recurrent disease

- Scale 0 – 100
- Higher score indicates more symptoms / problems



Implementation results

Identify patients at risk for poor HRQoL

Use subscales for specific groups or in specific research (e.g. health service research)

Use symptom scales in clinical trials



Conclusion

Development of a DTF-specific HRQoL questionnaire

- 95 items
- 14 subscales

DTF-QoL:

- Can be used alongside the EORTC QLQ-C30
- Answers help to gain more insight in HRQoL-issues DTF-patients to improve personalized care



Contributors

Erasmus MC Cancer Institute, Rotterdam, the Netherlands

Dirk Grünhagen

Stefan Sleijfer

Milea Timbergen

Cornelis Verhoef

Netherlands Cancer Institute, Amsterdam, the Netherlands

Winan van Houdt

Olga Husson

Winette van der Graaf

Royal Marsden NHS Foundation Trust, London, UK

Alison Dunlop

Spyridon Gennatas

Robin Jones

Emma Lidington

Eugenie Younger

Radboud University Medical Center, Nijmegen, the Netherlands

Johannes Bonenkamp





Contact

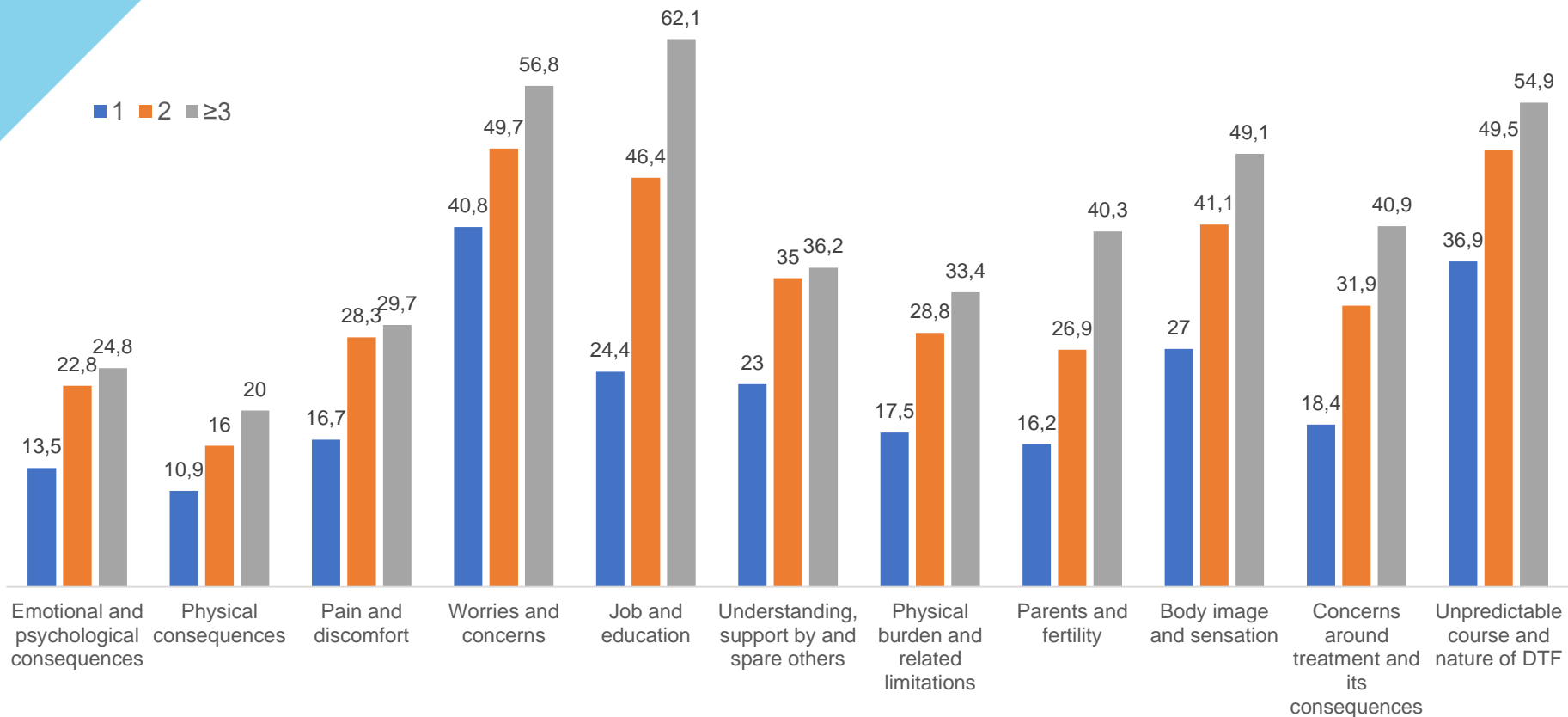
Anne-Rose Schut

a.schut@erasmusmc.nl

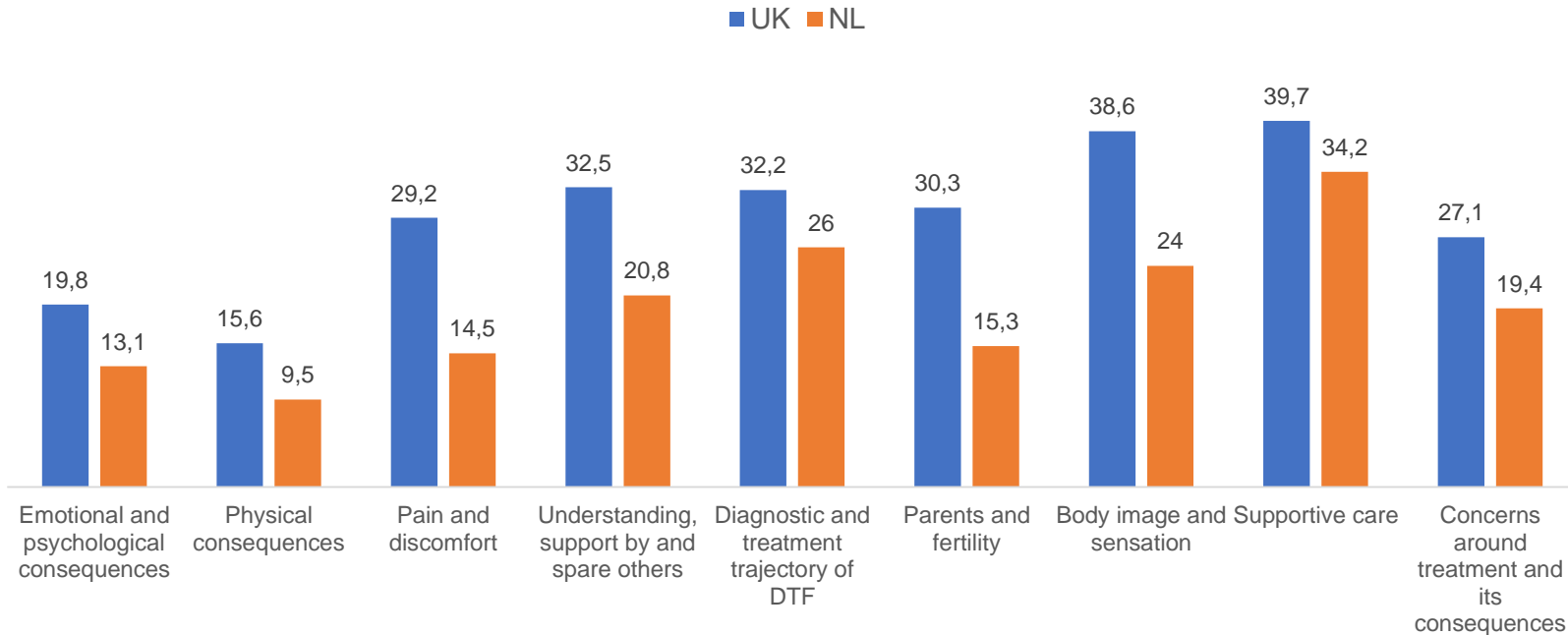




Number of treatments



Nationality



During the past week:

W1. Emotional and psychological consequences

- Have you been unsatisfied with your body?
- Have you had problems with your appearance?
- Have you felt isolated?
- Have you felt lonely?
- Have you worried about dying?
- Have you felt uncertain?
- Have you worried about the disease being aggressive?
- Have you had a bad temper because of the condition?

W2. Physical consequences

- Have you had any trouble walking?
- Have you had swelling in your legs or ankles (edema)?
- Have you felt disabled?
- Have you had stiffness in your limbs?
- Have you had problems getting dressed?

W3. Pain and discomfort

- Has pain interfered with your sleep at night?
- Have you had pain while sitting?
- Have you been unable to lean on the tumour sites? (e.g. due to local pressure)

In the last four weeks:

- Have you had a decreased libido?

Since your diagnosis as a result of your desmoid fibromatosis and/or treatment(s):

1. Worries and concerns

- Have you been afraid of tumour growth?
- Have you been afraid of getting another tumour?
- Have you been worried or concerned about the future?
- Have you felt worried constantly?
- Have you felt stressed about the diagnosis?
- Have you worried about your treatment?
- Has desmoid fibromatosis or its treatment caused those close to you to feel distressed?
- Have you felt stressed around check-ups?

2. Job and education

- 1. Have you had to take sick leave?
- 1. Have you had problems with your job or your education?
- 1. Have you worried about not being able to continue working or studying?
- 1. Have your career ambitions changed because of the diagnosis?
- 1. Have you had to change jobs as a result of your condition?

3. Doctor-patient relationship, communication and information

- Has there been mutual trust between you and your professional(s)?
- Have you been satisfied with your communication with your professional(s)?
- Have you received enough information about the possible treatment side effects?
- Have you had problems receiving enough information about your desmoid fibromatosis and its treatment?
- Have you lacked information about your desmoid fibromatosis and/or its treatment?
- Have you experienced a lack of continuity (seeing the same doctors / specialised nurse) in the care for your desmoid fibromatosis?

4. Understanding, support by and spare others

Have you felt that you have received less attention from family and friends because the condition is benign?

Do you think your condition is not well understood by people close to you?

Have you had difficulties explaining your condition to others?

Have you had problems with confidence?

Have you found it frustrating having to explain your condition to others?

Have you felt supported by your family members and/or friends?

Have you felt a need to keep your fears, concerns and/or symptoms from family members or friends?

Has the desmoid fibromatosis had a negative impact on your family life?

Have you worried about being treated unfairly because of your desmoid fibromatosis (i.e. at work, by insurance companies)?

Have you had to explain your circumstances to others?

Have you worried that you are a burden to other people?

Has your physical condition or medical treatment interfered with your marriage or intimate relationships?

Have you felt that you are standing still in life?

5. Physical burden and related limitations

Have you felt less independent and/or more dependent on others?

Have you had extra expenses due to your physical condition or medical treatment (e.g. for medication, transport and/or aids)?

Has your physical condition or medical treatment interfered with your social activities?

Has your physical fitness level reduced?

Have you had problems driving a car?

Have you lost friendships?

Have you had problems eating?

Have you felt embarrassed using mobility aids (e.g. wheelchair, scooter, electric bike)?

6. Diagnostic and treatment trajectory of DTF

Have you been bothered by long travel to the hospital?

Have you had a problem receiving treatments in a cancer hospital?

Were you passed from one hospital to another before the final desmoid fibromatosis diagnosis?

Did you think it took a long time to get a definite diagnosis?

Have you felt that doctors are unfamiliar with desmoid-type fibromatosis?

Have you received different contradictory recommendations about treatment options?

Were you frightened by the referral to the cancer hospital?

7. Parents and fertility

Have you been worried about tumour growth during pregnancy?

Have you had problems with your ability to have children because of your desmoid fibromatosis?

Has your parental role been affected because of your desmoid fibromatosis?

Have you worried about your ability to have children?

Have you worried about passing the condition to your children?

8. Body image and sensations

Have you wanted to cover-up the tumour area and /or scar(s)?

Have you felt a change in sensation in the area around the tumour?

Have you felt less feminine/masculine?

Have you felt asymmetrical and/or misshapen due to the desmoid fibromatosis or the treatment?

9. Supportive care

Have you lacked online support (forum and/or chat group)?

Have you wanted to meet others with desmoid fibromatosis?

Have you lacked psychological support?

10. Concerns around treatment and its consequences

Have you been afraid of needing a limb amputation?

Have you felt addicted to pain medication?

Have you had doubts about the effectiveness of your treatments?

Have you worried about a decrease in muscle strength after treatment?

11. Unpredictable course and nature of DTF

Have you felt there is no optimal treatment for you?

Do you think your prognosis (the expected improvement or worsening of your condition) is clear?

Have you been disappointed by the course of your condition?

Do you feel like you have a chronic disease?

Have you felt you had to fight this condition?

Have you felt there is something in your body that does not belong there?

Have you felt reassured by the benign nature of your disease?

Have you worried about being able to access treatments in the future?

Have you felt frustrated about the "benign" diagnosis with cancerous features?

Have you been afraid of your desmoid fibromatosis coming back?

Have you felt like a cancer patient?

Single items.

Has the colour of your hair changed?

Have you felt your desmoid fibromatosis has changed your life in a negative way?

Have you lost your hair?

Have you had a rash as a result of the treatment?

Have you felt you were wasting the time of cancer specialists?