

Appendix S1

# Questionnaire about pruritus in neurofibromatosis 1

## 1. Chronology

When did pruritus begin?

weeks  months  years

Is pruritus :

continuous  episodic

Do you feel pruritus :

Every day.....   
 Almost every day.....   
 Every week.....   
 Every month.....   
 Seldom.....

Please indicate the frequency of appearance of pruritus for each part of the day : (put a cross on each line)

	Never	Occasionnal	Often	Always present
Morning	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Afternoon	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Evening	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Night	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>

## 2. Location

Is pruritus localized on neurofibromas?

yes  no

## 3. Characteristics of pruritus

Does these sensations accompanying your itch?

	Yes	No
Sweating	<input type="checkbox"/>	<input type="checkbox"/>
Headache	<input type="checkbox"/>	<input type="checkbox"/>
Pain	<input type="checkbox"/>	<input type="checkbox"/>
Heat sensation	<input type="checkbox"/>	<input type="checkbox"/>
Cold sensation	<input type="checkbox"/>	<input type="checkbox"/>

Do you also feel these sensations?

	Yes	No
Stinging	<input type="checkbox"/>	<input type="checkbox"/>
Tickling	<input type="checkbox"/>	<input type="checkbox"/>
Crawling	<input type="checkbox"/>	<input type="checkbox"/>
Stabbing	<input type="checkbox"/>	<input type="checkbox"/>
Pinching	<input type="checkbox"/>	<input type="checkbox"/>
Burning	<input type="checkbox"/>	<input type="checkbox"/>
Biting	<input type="checkbox"/>	<input type="checkbox"/>
Stroking	<input type="checkbox"/>	<input type="checkbox"/>

### 4. Intensity

What is the intensity of pruritus on a scale from 0 to 10 ?

(0 : no pruritus ; 10 : the worst pruritus imaginable)

Put a cross on each line

	No pruritus	Worst pruritus									
	0	1	2	3	4	5	6	7	8	9	10
Now											
In worst moment											
In best moment											
In mean											

### 5. Treatment

Please indicate treatment (medication, cream...) you take for your pruritus and if no effect, short effect, long effect.

Treatment	No effect	Short effect (less than 24hours)	Long effect (more than 24hours)

### 6. Disruption of daily activities

Please indicate how each item affects your pruritus :

Put a cross on each line.

	Increases	Does not affect	Relieves
Sleep			
Rest			
Activity			
Stress			
Fatigue			
Physical effort			
Skin dryness			
Hot water			
Cold water			
Sweat			
Cold			
Heat			
Clothes			

### 7. Characteristics of scratching

Do you scratch ?

- No.....
- Rarely.....
- Often.....
- Very often.....

If yes, is scratching :

- Highly pleasurable.....
- Moderately pleasurable.....
- Neutral.....
- Moderately unpleasurable.....
- Highly unpleasurable.....

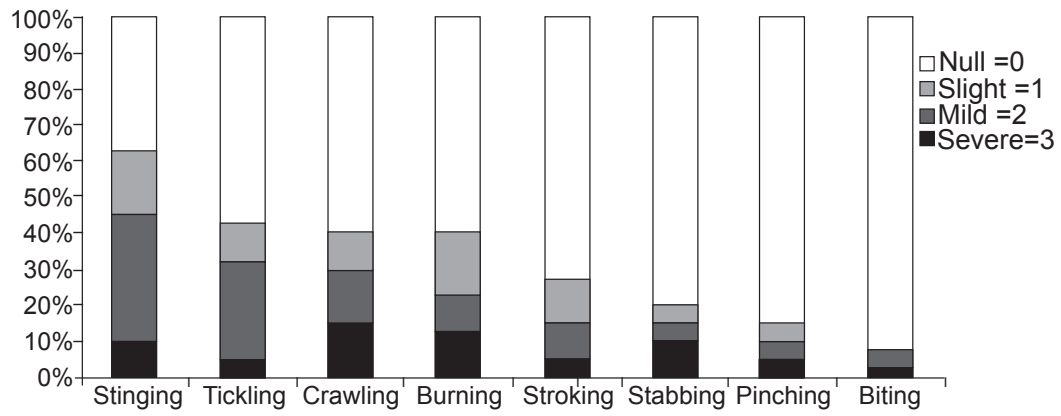


Fig. S1. Frequency of sensory symptoms in patients with neurofibromatosis 1, evaluated from 0 (nonexistent) to 3 (severe).

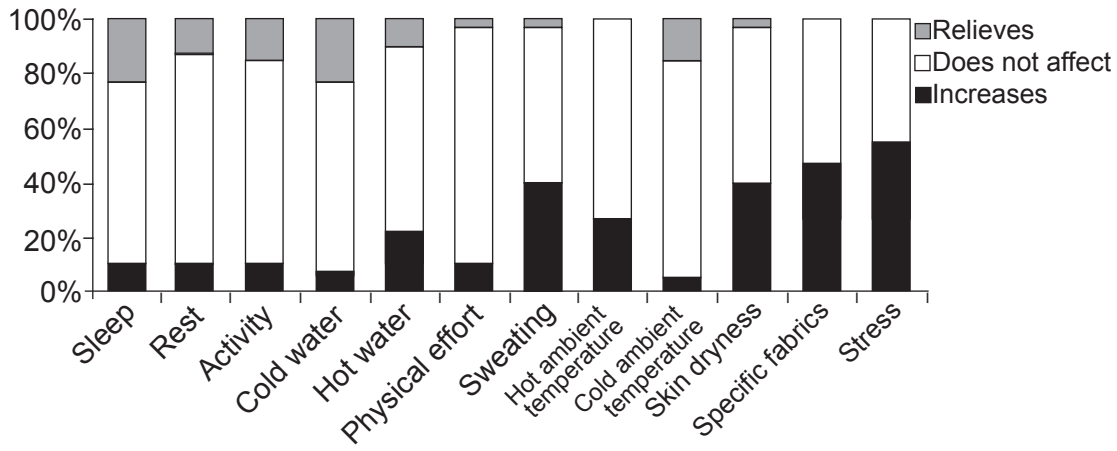


Fig. S2. Effects of daily activities on pruritus.

Table SI. Results from the 5-D itch scale

Duration	During the last 2 weeks, how many hours a day have you been itching?
<6 h/day	37 (92.5%)
6–12 h/day	1 (2.5%)
12–18 h/day	1 (2.5%)
18–23 h/day	0
All day	1 (2.5%)
Degree	Please rate the intensity of your itching over the past 2 weeks
Not present	1 (2.5%)
Mild	12 (30.0%)
Moderate	21 (52.5%)
Severe	1 (2.5%)
Unbearable	5 (12.5%)
Direction	Over the past 2 weeks has your itching gotten better or worse compared to the previous month?
Completely resolved	0
Much better but still present	3 (7.5%)
Little bit better, but still present	5 (12.5%)
Unchanged	28 (70.0%)
Getting worse	4 (10.0%)
Disability	Highest score on any of the four items (sleep, leisure/social, housework/errands, work/school)
Never affects this activity	10 (25.0%)
Rarely affects this activity	5 (12.5%)
Occasionally affects this activity	14 (35.0%)
Frequently affects this activity	7 (17.5%)
Always affects this activity	4 (10.0%)
Distribution	Number of parts of the body where itching has been present over the last 2 weeks, among: head/scalp, face, chest, abdomen, back, buttocks, thighs, lower legs, soles, palms, tops of hands/fingers, forearms, upper arms, points of contact w/ clothing, groin, tops of feet/toes
0–2 parts	12 (30.0%)
3–5 parts	12 (30.0%)
6–10 parts	14 (35.0%)
11–13 parts	0
14–16 parts	2 (5.0%)