BarTH Syndrome Symptom Assessment (BTHS-SA) – Adolescent version

Instructions: The following questions ask about Barth Syndrome. Please select the response that best describes your experience with Barth Syndrome over the past 24 hours. Please select only one answer for each question. Please answer all of the questions and do not skip any. There are no right or wrong answers to any of the questions.

Please indicate (with a check mark ☑️) responses to the questions below.

| Question                                                                 | No feeling of early fullness at all | Mild feeling of early fullness | Moderate feeling of early fullness | Severe feeling of early fullness | Very severe feeling of early fullness |
|-------------------------------------------------------------------------|-------------------------------------|--------------------------------|-----------------------------------|---------------------------------|--------------------------------------|
| 1. Please rate your worst feeling of tiredness at rest in the past 24 hours. | No tiredness at all                | Mild tiredness                 | Moderate tiredness                | Severe tiredness                 | Very severe tiredness                |
| 2. Please rate your worst feeling of tiredness during activities in the past 24 hours. | No tiredness at all                | Mild tiredness                 | Moderate tiredness                | Severe tiredness                 | Very severe tiredness                |
| 3. Please rate your worst feeling of muscle weakness at rest in the past 24 hours. | No muscle weakness at all           | Mild muscle weakness            | Moderate muscle weakness           | Severe muscle weakness           | Very severe muscle weakness           |
| 4. Please rate your worst feeling of muscle weakness during activities in the past 24 hours. | No muscle weakness at all           | Mild muscle weakness            | Moderate muscle weakness           | Severe muscle weakness           | Very severe muscle weakness           |
| 5. Please rate your worst feeling of muscle pain at rest in the past 24 hours. | No muscle pain at all                | Mild muscle pain                | Moderate muscle pain                | Severe muscle pain                | Very severe muscle pain                |
| 6. Please rate your worst feeling of muscle pain due to activities in the past 24 hours. | No muscle pain at all                | Mild muscle pain                | Moderate muscle pain                | Severe muscle pain                | Very severe muscle pain                |
| 7. Please rate your worst feeling of early fullness when eating in the past 24 hours. | No feeling of early fullness at all | Mild feeling of early fullness  | Moderate feeling of early fullness | Severe feeling of early fullness | Very severe feeling of early fullness |
| 8. Please rate your worst difficulty eating (for example, chewing and/or swallowing) in the past 24 hours. | No difficulty eating at all          | Mild difficulty eating          | Moderate difficulty eating          | Severe difficulty eating          | Very severe difficulty eating          |
| 9. Please rate your worst feeling of headache in the past 24 hours. | No headache at all                        | Mild headache                      | Moderate headache                      | Severe headache                      | Very severe headache                      |
BarTH Syndrome Symptom Assessment (BTHS-SA) – Adult version

Instructions: The following questions ask about Barth Syndrome. Please select the response that best describes your experience with Barth Syndrome over the past 24 hours. Please select only one answer for each question. Please answer all of the questions and do not skip any. There are no right or wrong answers to any of the questions.

Please indicate (with a check mark ☑) responses to the questions below.

| Question                                                                 | No tasting at all | Mild tasting | Moderate tasting | Severe tasting | Very severe tasting |
|--------------------------------------------------------------------------|-------------------|--------------|------------------|----------------|---------------------|
| 1. Please rate your worst feeling of tiredness at rest in the past 24 hours. | ☑                 | ☐            | ☐                | ☐              | ☐                   |
| 2. Please rate your worst feeling of tiredness during activities in the past 24 hours. | ☑                 | ☐            | ☐                | ☐              | ☐                   |
| 3. Please rate your worst feeling of muscle weakness at rest in the past 24 hours. | ☑                 | ☐            | ☐                | ☐              | ☐                   |
| 4. Please rate your worst feeling of muscle weakness during activities in the past 24 hours. | ☑                 | ☐            | ☐                | ☐              | ☐                   |
| 5. Please rate your worst feeling of muscle pain at rest in the past 24 hours. | ☑                 | ☐            | ☐                | ☐              | ☐                   |
| 6. Please rate your worst feeling of muscle pain due to activities in the past 24 hours. | ☑                 | ☐            | ☐                | ☐              | ☐                   |
| 7. Please rate your worst feeling of dizziness/lightheadedness in the past 24 hours. | ☑                 | ☐            | ☐                | ☐              | ☐                   |
| 8. Please rate your worst feeling of shortness of breath in the past 24 hours. | ☑                 | ☐            | ☐                | ☐              | ☐                   |
| Statement                                                                 | Never | Almost Never | Sometimes | Often | Almost Always |
|---------------------------------------------------------------------------|-------|--------------|-----------|-------|---------------|
| Being tired made it hard for me to play or go out with my friends as much as I'd like. |       |              |           |       |               |
| I felt weak.                                                              |       |              |           |       |               |
| I got tired easily.                                                        |       |              |           |       |               |
| Being tired made it hard for me to keep up with my schoolwork.            |       |              |           |       |               |
| I had trouble finishing things because I was too tired.                   |       |              |           |       |               |
| I had trouble starting things because I was too tired.                    |       |              |           |       |               |
| I was so tired it was hard for me to pay attention.                       |       |              |           |       |               |
| I was too tired to do sports or exercise.                                 |       |              |           |       |               |
| I was too tired to do things outside.                                     |       |              |           |       |               |
| I was too tired to enjoy the things I like to do.                         |       |              |           |       |               |
PROMIS Item Bank v1.0 – Fatigue – Short Form 8a

Fatigue – Short Form 8a

Please respond to each question or statement by marking one box per row.

| Question                                                                 | Not at all | A little bit | Somewhat | Quite a bit | Very much |
|--------------------------------------------------------------------------|------------|--------------|----------|-------------|-----------|
| **During the past 7 days...**                                            |            |              |          |             |           |
| I feel fatigued                                                         |            |              |          |             |           |
| during the past 7 days                                                   | □          | □            | □        | □           | □         |
| I have trouble starting things because I am tired                       | □          | □            | □        | □           | □         |
| **In the past 7 days...**                                                |            |              |          |             |           |
| How run-down did you feel on average?                                   | □          | □            | □        | □           | □         |
| How fatigued were you on average?                                        | □          | □            | □        | □           | □         |
| How much were you bothered by your fatigue on average?                   | □          | □            | □        | □           | □         |
| How often did you have to push yourself to get things done              | □          | □            | □        | □           | □         |
| **In the past 7 days...**                                                | Never      | Rarely       | Sometimes| Often       | Always    |
| How often did you have trouble finishing things                          | □          | □            | □        | □           | □         |

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Describing your health TODAY

Please check the ONE box that best describes your health TODAY.

Mobility (walking around)

I have no problems walking around
I have some problems walking around
I have a lot of problems walking around

Taking care of myself

I have no problems taking a bath or shower by myself or getting dressed by myself
I have some problems taking a bath or shower by myself or getting dressed by myself
I have a lot of problems taking a bath or shower by myself or getting dressed by myself

Doing usual activities (for example, going to school, hobbies, sports, playing, doing things with family or friends)

I have no problems doing my usual activities
I have some problems doing my usual activities
I have a lot of problems doing my usual activities

Having pain or discomfort

I have no pain or discomfort
I have some pain or discomfort
I have a lot of pain or discomfort

Feeling worried, sad, or unhappy

I am not worried, sad, or unhappy
I am a little worried, sad, or unhappy
I am very worried, sad, or unhappy
How good is your health TODAY

- We would like to know how good or bad your health is TODAY.
- This line is numbered from 0 to 100.
- 100 means the best health you can imagine.
  0 means the worst health you can imagine.
- Please mark an X on the line to show how good or bad your health is TODAY.
Patient Global Impression of Symptoms (Patient age 12-15)

Date Completed: _______________

The following questions ask you about your Barth Syndrome symptoms OVER THE PAST WEEK.

1. How bad have your Barth Syndrome symptoms been over the past week?
   - No Symptoms
   - Mild Symptoms
   - Moderate Symptoms
   - Severe Symptoms
   - Very Severe Symptoms

2. How bad has your feeling of tiredness at rest been over the past week?
   - No Tiredness
   - Mild Tiredness
   - Moderate Tiredness
   - Severe Tiredness
   - Very Severe Tiredness

3. How bad has your feeling of tiredness during activities been over the past week?
   - No Tiredness
   - Mild Tiredness
   - Moderate Tiredness
   - Severe Tiredness
   - Very Severe Tiredness

4. How bad has your feeling of muscle weakness at rest been over the past week?
   - No Muscle Weakness
   - Mild Muscle Weakness
   - Moderate Muscle Weakness
   - Severe Muscle Weakness
   - Very Severe Muscle Weakness

5. How bad has your feeling of muscle weakness during activities been over the past week?
   - No Muscle Weakness
   - Mild Muscle Weakness
   - Moderate Muscle Weakness
   - Severe Muscle Weakness
   - Very Severe Muscle Weakness
6. How bad has your feeling of muscle pain at rest been over the past week?

☐ No Muscle Pain
☐ Mild Muscle Pain
☐ Moderate Muscle Pain
☐ Severe Muscle Pain
☐ Very Severe Muscle Pain

7. How bad has your feeling of muscle pain due to activities been over the past week?

☐ No Muscle Pain
☐ Mild Muscle Pain
☐ Moderate Muscle Pain
☐ Severe Muscle Pain
☐ Very Severe Muscle Pain

8. How bad has your feeling of early fullness when eating been over the past week?

☐ No Early Fullness
☐ Mild Early Fullness
☐ Moderate Early Fullness
☐ Severe Early Fullness
☐ Very Severe Early Fullness

9. How bad has your feeling of difficulty eating (for example, chewing and/or swallowing) been over the past week?

☐ No Difficulty Eating
☐ Mild Difficulty Eating
☐ Moderate Difficulty Eating
☐ Severe Difficulty Eating
☐ Very Severe Difficulty Eating

10. How bad has your feeling of headache been over the past week?

☐ No Headache
☐ Mild Headache
☐ Moderate Headache
☐ Severe Headache
☐ Very Severe Headache
Patient Global Impression of Symptoms (Patient age >= 16)

Date Completed: ________________

The following questions ask you about your Barth Syndrome symptoms OVER THE PAST WEEK.

1. How bad have your Barth Syndrome symptoms been over the past week?
   - No Symptoms
   - Mild Symptoms
   - Moderate Symptoms
   - Severe Symptoms
   - Very Severe Symptoms

2. How bad has your feeling of tiredness at rest been over the past week?
   - No Tiredness
   - Mild Tiredness
   - Moderate Tiredness
   - Severe Tiredness
   - Very Severe Tiredness

3. How bad has your feeling of tiredness during activities been over the past week?
   - No Tiredness
   - Mild Tiredness
   - Moderate Tiredness
   - Severe Tiredness
   - Very Severe Tiredness

4. How bad has your feeling of muscle weakness at rest been over the past week?
   - No Muscle Weakness
   - Mild Muscle Weakness
   - Moderate Muscle Weakness
   - Severe Muscle Weakness
   - Very Severe Muscle Weakness

5. How bad has your feeling of muscle weakness during activities been over the past week?
   - No Muscle Weakness
   - Mild Muscle Weakness
   - Moderate Muscle Weakness
   - Severe Muscle Weakness
   - Very Severe Muscle Weakness
6. How bad has your feeling of muscle pain at rest been over the past week?
   □ No Muscle Pain
   □ Mild Muscle Pain
   □ Moderate Muscle Pain
   □ Severe Muscle Pain
   □ Very Severe Muscle Pain

7. How bad has your feeling of muscle pain due to activities been over the past week?
   □ No Muscle Pain
   □ Mild Muscle Pain
   □ Moderate Muscle Pain
   □ Severe Muscle Pain
   □ Very Severe Muscle Pain

8. How bad has your feeling of dizziness/lightheadedness been over the past week?
   □ No Dizziness/Lightheadedness
   □ Mild Dizziness/Lightheadedness
   □ Moderate Dizziness/Lightheadedness
   □ Severe Dizziness/Lightheadedness
   □ Very Severe Dizziness/Lightheadedness

9. How bad has your feeling of shortness of breath been over the past week?
   □ No Shortness of Breath
   □ Mild Shortness of Breath
   □ Moderate Shortness of Breath
   □ Severe Shortness of Breath
   □ Very Severe Shortness of Breath
Caregiver Global Impression of Symptoms (Patient age 12-15)

Date Completed: ________________

The following questions ask you about your child’s Barth Syndrome symptoms OVER THE PAST WEEK. Please base your responses on what you have observed or what your child has told you about their symptoms in the past week.

1. How bad have your child’s Barth Syndrome symptoms been over the past week?
   - [ ] No Symptoms
   - [ ] Mild Symptoms
   - [ ] Moderate Symptoms
   - [ ] Severe Symptoms
   - [ ] Very Severe Symptoms

2. How bad has your child’s feeling of tiredness at rest been over the past week?
   - [ ] No Tiredness
   - [ ] Mild Tiredness
   - [ ] Moderate Tiredness
   - [ ] Severe Tiredness
   - [ ] Very Severe Tiredness

3. How bad has your child’s feeling of tiredness during activities been over the past week?
   - [ ] No Tiredness
   - [ ] Mild Tiredness
   - [ ] Moderate Tiredness
   - [ ] Severe Tiredness
   - [ ] Very Severe Tiredness

4. How bad has your child’s feeling of muscle weakness at rest been over the past week?
   - [ ] No Muscle Weakness
   - [ ] Mild Muscle Weakness
   - [ ] Moderate Muscle Weakness
   - [ ] Severe Muscle Weakness
   - [ ] Very Severe Muscle Weakness

5. How bad has your child’s feeling of muscle weakness during activities been over the past week?
   - [ ] No Muscle Weakness
   - [ ] Mild Muscle Weakness
   - [ ] Moderate Muscle Weakness
   - [ ] Severe Muscle Weakness
☐ Very Severe Muscle Weakness

6. How bad has your child’s feeling of muscle pain at rest been over the past week?
   - No Muscle Pain
   - Mild Muscle Pain
   - Moderate Muscle Pain
   - Severe Muscle Pain
   - Very Severe Muscle Pain

7. How bad has your child’s feeling of muscle pain due to activities been over the past week?
   - No Muscle Pain
   - Mild Muscle Pain
   - Moderate Muscle Pain
   - Severe Muscle Pain
   - Very Severe Muscle Pain

8. How bad has your child’s feeling of early fullness when eating been over the past week?
   - No Early Fullness
   - Mild Early Fullness
   - Moderate Early Fullness
   - Severe Early Fullness
   - Very Severe Early Fullness

9. How bad has your child’s feeling of difficulty eating (for example, chewing and/or swallowing) been over the past week?
   - No Difficulty Eating
   - Mild Difficulty Eating
   - Moderate Difficulty Eating
   - Severe Difficulty Eating
   - Very Severe Difficulty Eating

10. How bad has your child’s feeling of headache been over the past week?
    - No Headache
    - Mild Headache
    - Moderate Headache
    - Severe Headache
    - Very Severe Headache
Caregiver Global Impression of Symptoms (Patient age >= 16)

Date Completed: ______________

The following questions ask you about the individual with Barth Syndrome that you care for and the symptoms that he/she has had OVER THE PAST WEEK. Please base your responses on what you have observed or what the individual with Barth Syndrome has told you about his/her symptoms in the past week.

1. How bad have his/her **Barth Syndrome symptoms** been over the past week?
   - No Symptoms
   - Mild Symptoms
   - Moderate Symptoms
   - Severe Symptoms
   - Very Severe Symptoms

2. How bad has his/her feeling of **tiredness at rest** been over the past week?
   - No Tiredness
   - Mild Tiredness
   - Moderate Tiredness
   - Severe Tiredness
   - Very Severe Tiredness

3. How bad has his/her feeling of **tiredness during activities** been over the past week?
   - No Tiredness
   - Mild Tiredness
   - Moderate Tiredness
   - Severe Tiredness
   - Very Severe Tiredness

4. How bad has his/her feeling of **muscle weakness at rest** been over the past week?
   - No Muscle Weakness
   - Mild Muscle Weakness
   - Moderate Muscle Weakness
   - Severe Muscle Weakness
   - Very Severe Muscle Weakness

5. How bad has his/her feeling of **muscle weakness during activities** been over the past week?
   - No Muscle Weakness
   - Mild Muscle Weakness
   - Moderate Muscle Weakness
   - Severe Muscle Weakness
☐ Very Severe Muscle Weakness

6. How bad has his/her feeling of muscle pain at rest been over the past week?
   ☐ No Muscle Pain
   ☐ Mild Muscle Pain
   ☐ Moderate Muscle Pain
   ☐ Severe Muscle Pain
   ☐ Very Severe Muscle Pain

7. How bad has his/her feeling of muscle pain due to activities been over the past week?
   ☐ No Muscle Pain
   ☐ Mild Muscle Pain
   ☐ Moderate Muscle Pain
   ☐ Severe Muscle Pain
   ☐ Very Severe Muscle Pain

8. How bad has his/her feeling of dizziness/lightheadedness been over the past week?
   ☐ No Dizziness/Lightheadedness
   ☐ Mild Dizziness/Lightheadedness
   ☐ Moderate Dizziness/Lightheadedness
   ☐ Severe Dizziness/Lightheadedness
   ☐ Very Severe Dizziness/Lightheadedness

9. How bad has his/her feeling of shortness of breath been over the past week?
   ☐ No Shortness of Breath
   ☐ Mild Shortness of Breath
   ☐ Moderate Shortness of Breath
   ☐ Severe Shortness of Breath
   ☐ Very Severe Shortness of Breath
Clinician Global Impression of Symptoms (all ages)

Date Completed: ________________

1. Overall, how severe are the patient’s Barth Syndrome symptoms today?
   - [ ] No Symptoms
   - [ ] Mild Symptoms
   - [ ] Moderate Symptoms
   - [ ] Severe Symptoms
   - [ ] Very Severe Symptoms