Manual for the Use of Hypertrophic Cardiomyopathy Symptoms Questionnaire (HCMSQ) V2.0 24h-recall Version

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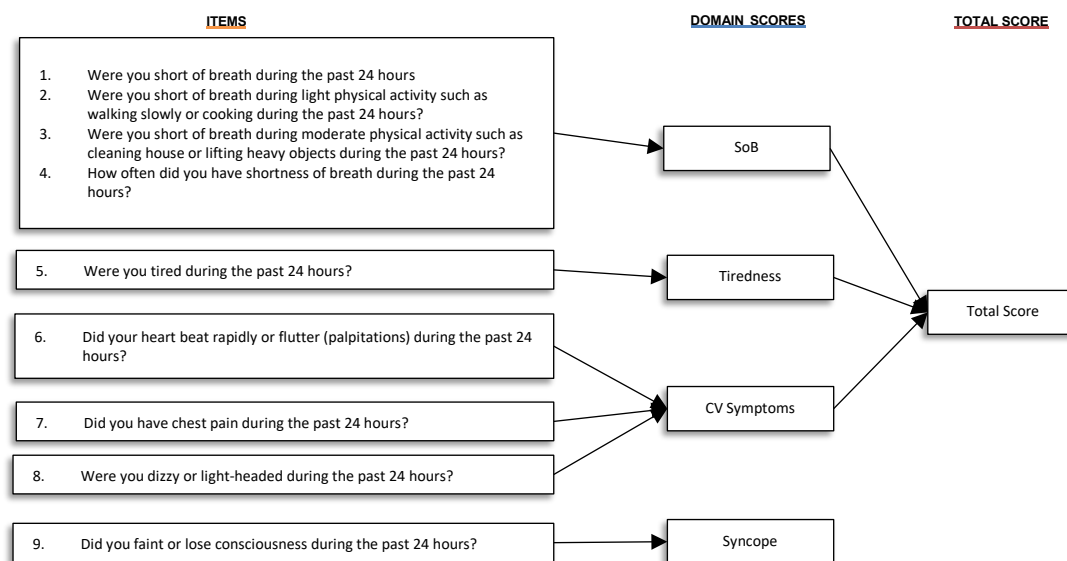
English for USA

Introduction

The HCMSQv2.0 24h-recall version contains nine patient-reported symptom items that patients with hypertrophic cardiomyopathy (HCM) answer daily to report their symptom status (Figure 1). These nine HCMSQv2.0 24h-recall items form four domains: shortness of breath (SoB), tiredness, cardiovascular (CV) symptoms, and syncope. Of these four HCMSQ domains, two are multi-item (SoB and CV symptoms), while HCMSQ tiredness and syncope are single item domains. Three of the four domain scores (SoB, Tiredness and CV symptoms) are used to create the HCMSQ Total Score.

The procedure to calculate each of the domain scores will be described in the Scoring Guide included in this user manual. Briefly, domain scores are computed from daily HCMSQ data; patients would have to complete a daily entry on at least 4 out of 7 consecutive days. The 7 consecutive days can start on any day of the week. For all domain and total scores, lower scores indicate a lower symptom burden.

It is important to note that the HCMSQv2.0 24h-recall version uses the weekly scores. While daily scores are collected to calculate the weekly score, they are not validated for use on their own. Therefore, caution should be exercised when interpreting daily scores in patients with hypertrophic cardiomyopathy.

Figure 1: Conceptual Framework of the HCMSQv2.0

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Participant Training

Please walk through the two examples of questions and responses below with your participant before administering the HCMSQ v2.0 24h-recall, and ask the participant to select **ONE** response that most accurately reflects their experience.

Question 1: Select the option below that best describes the weather today.

o Sunny
o Cloudy
o Raining
o Snowing

Some questions may contain response options as seen below. It is important that participants understand all response options to select **ONE** response that most accurately reflects their experience. Consider the following example.

Question 2: How short of breath were you when you washed your hands today?

o I did not attempt to do the activity
o Not at all
o Mildly
o Moderately
o Severely
o Very Severely
o Too short of breath to do the activity

- For "I did not attempt to do the activity" consider:
    o You did not attempt the activity today because the opportunity to do the activity did not occur.
    o For example, you were staying somewhere without soap and water

- For "Too short of breath to do the activity" consider:
    o You <u>did</u> attempt to do the activity today but you were unable due to shortness of breath
    o Or you <u>did not</u> attempt to do the activity today because you were unable due to shortness of breath

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Scoring Guide

Calculating the HCMSQv2.0 SoB domain score:
First, an HCMSQ SoB daily score should be calculated from items 1-4 of the HCMSQv2.0 for each day. The weekly HCMSQ SoB domain score can then be calculated from the daily scores if the participant has an HCMSQ SoB daily score on at least 4 out of 7 consecutive days.

Calculating an HCMSQ SoB **daily** score:
1. Code responses to each of **Items 1-4** according to Table 1
   - If "I did not attempt to do the activity" was selected for **Item 3**, the **Item 3** should be imputed as the mean of Items 1, 2, and 4 for that day, if available.
2. Calculate a daily score as the sum of **Items 1-4.**
   - A daily score cannot be calculated, and the HCMSQ SoB score is treated as missing for that day, if:
     - If there is no response selected to any one of the four items (i.e. at least one item response is missing)
     - The 'I did not attempt to do the activity' response choice is selected for **Item 2**

Calculating an HCMSQ SoB **domain** score:
Average the HCMSQ SoB daily scores collected over 7 consecutive days (a week).
1. If a patient has 4 to 7 daily non-missing HCMSQ SoB daily scores for a week, an HCMSQ SoB domain score can be generated.
   - The HCMSQ SoB domain score is calculated as the average of the HCMSQ SoB daily scores with completed entries. For example, when calculating the average score with four SoB daily scores, it is important to note that the denominator should be 4 and not 7, as the day with missing SoB daily score should not be counted as 0.
2. If a patient has only **0 to 3** daily non-missing HCMSQ SoB daily scores for a week, an HCMSQ SoB domain score **cannot** be generated and should be treated as missing.

The potential HCMSQ SoB daily score and the weekly HCMSQ SoB domain score ranges 0-18, where lower scores indicate less shortness of breath.

Table 1: Codes for responses to items 1-4 of the HCMSQv2.0

Core Symptom	Item	Question	Response Options
Shortness of Breath	1	Were you short of breath during the past 24 hours?	0 = Not at all 1 = Mildly 2 = Moderately 3 = Severely 4 = Very Severely
	2	Were you short of breath during light physical activity such as walking slowly or cooking during the past 24 hours?	missing = I did not attempt to do the activity 0 = Not at all 1 = Mildly 2 = Moderately 3 = Severely 4 = Very Severely 5 = Too short of breath to do the activity
	3	Were you short of breath during moderate physical activity such as cleaning house or lifting heavy objects?	missing = I did not attempt to do the activity 0 = Not at all 1 = Mildly 2 = Moderately 3 = Severely 4 = Very Severely 5 = Too short of breath to do the activity
	4	How often did you have shortness of breath during past 24 hours?	0 = Never 1 = Seldom 2 = Sometimes 3 = Often 4 = Almost Always

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Calculating the HCMSQv2.0 Tiredness domain score:

First, an HCMSQ Tiredness daily score should be calculated from item 5 of the HCMSQv2.0 for each of 7 consecutive days (a week). Then the weekly HCMSQ Tiredness domain score can be calculated from the 7 consecutive daily scores.

Calculating an HCMSQ Tiredness **daily** score:

Code responses to **Item 5** according to Table 2.

Calculating an HCMSQ Tiredness **domain** score:

Average the HCMSQ Tiredness daily scores collected over 7 consecutive days (a week).

1. If a patient has **4 to 7** daily non-missing HCMSQ Tiredness daily scores for a week, an HCMSQ tiredness domain score **can** be generated.
   - The HCMSQ Tiredness domain score is calculated as the average of the HCMSQ Tiredness daily scores with completed entries.
2. If a patient has only **0 to 3** daily non-missing HCMSQ Tiredness daily scores for a week, an HCMSQ Tiredness domain score **cannot** be generated and should be treated as missing.

The potential HCMSQ Tiredness daily score and the weekly HCMSQ Tiredness domain score ranges 0-4, where lower scores indicate less tiredness.

Table 2: Codes for responses to item 5 of the HCMSQv2.0

Core Symptom	Item	Question	Response Options
Tiredness	5	Were you tired during the past 24 hours?	0 = Not at all 1 = Mildly 2 = Moderately 3 = Severely 4 = Very Severely

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Calculating the HCMSQv2.0 CV Symptoms domain score:

First, an HCMSQ CV Symptoms daily score should be calculated from items 6-8 of the HCMSQv2.0 for each of 7 consecutive days (a week). Then the weekly HCMSQ CV Symptoms domain score can be calculated from the 7 consecutive daily scores.

Calculating an HCMSQ CV Symptoms **daily** score:

1. Code responses to each of **Items 6-8** according to Table 3
2. Calculate a HCMSQ CV Symptoms daily score as the sum of **Items 6-8.**
   - A daily score cannot be calculated, and the HCMSQ CV Symptoms score is treated as missing for that day, if there is no response selected to any one of the three items (i.e. at least one item response is missing)

Calculating an HCMSQ CV Symptoms **domain** score:

Average the HCMSQ CV Symptoms daily scores collected over 7 consecutive days (a week).

1. If a patient has 4 to 7 daily non-missing HCMSQ CV Symptoms daily scores for a week, an HCMSQ CV Symptoms domain score can be generated.
   - The HCMSQ CV Symptoms domain score is calculated as the average of the HCMSQ CV Symptoms daily scores with completed entries.
2. If a patient has only 0 to 3 daily non-missing HCMSQ CV Symptoms daily scores for a week, an HCMSQ CV Symptoms domain score cannot be generated and should be treated as missing.

The potential HCMSQ CV Symptoms daily score and the HCMSQ CV Symptoms domain score ranges 0-12, where lower scores indicate less shortness of breath.

Table 3: Codes for responses to items 6-8 of the HCMSQv2.0

Core Symptom	Item	Question	Response Options
Heart palpitations	6	Did your heart beat rapidly or flutter (palpitations) during past 24 hours?	0 = Not at all 1 = Mildly 2 = Moderately 3 = Severely 4 = Very Severely
Chest pain	7	Did you have chest pain during the past 24 hours?	0 = Not at all 1 = Mildly 2 = Moderately 3 = Severely 4 = Very Severely
Dizziness	8	Were you dizzy or lightheaded during the past 24 hours?	0 = Not at all 1 = Mildly 2 = Moderately 3 = Severely 4 = Very Severely

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Calculating the HCMSQv2.0 Total Score:

Calculating an HCMSQ Total **daily** score:

1. First, an HCMSQ Total daily score should be calculated from the HCMSQ SoB daily score, HCMSQ Tiredness daily score and HCMSQ CV Symptoms daily score for each of 7 consecutive days (a week).
2. The HCMSQ Total daily score is calculated as the equally weighted sum of the three daily scores, calculated as follows:

$$\frac{\text{HCMSQ SoB daily score}}{4 \text{ (items)}} + \frac{\text{HCMSQ Tiredness daily score}}{1 \text{ (item)}} + \frac{\text{HCMSQ CV Symptoms daily score}}{3 \text{ (items)}}$$

3. An HCMSQ Total daily score **cannot** be calculated, and the HCMSQ Total daily score is treated as **missing** for that day if there is no score for any one of the three daily scores required for the calculation in 2. above (i.e., at least one daily score response is missing).

Calculating an HCMSQ Total Score:

Average the HCMSQv2.0 Total daily scores collected over 7 consecutive days (a week).

1. If a patient has **4 to 7** non-missing HCMSQ Total daily scores for a week, an HCMSQ Total Score **can** be generated.
    a. The HCMSQ Total Score is calculated as the average of the HCMSQ Total daily scores with completed entries.
2. If a patient has only **0 to 3** daily non-missing HCMSQ Total daily scores for a week, an HCMSQ Total Score **cannot** be generated and should be treated as missing.

The potential HCMSQ Total daily score and the weekly HCMSQ Total Score ranges 0-12.5, where lower scores indicate less symptoms.

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Calculating the HCMSQv2.0 Syncope domain score:

First, an HCMSQ Syncope daily score should be calculated from item 9 of the HCMSQv2.0 for each of 7 consecutive days (a week). Then the weekly HCMSQ Syncope domain score can be calculated from the 7 consecutive daily scores.

Calculating an HCMSQ Syncope **daily** score:

Code responses to **Item 9** according to Table 5. The potential HCMSQ Syncope daily score is 0 or 1, where 0 represents no syncope in the past 24 hours and 1 represents at least one episode of syncope in the past 24 hours.

Calculating an HCMSQ Syncope **domain** score:

Sum the HCMSQ Syncope daily scores collected over 7 consecutive days (a week). The weekly HCMSQ Syncope domain score ranges 0-7, where lower scores indicate fewer days with syncope.

Table 5: Codes for responses to item 9 of the HCMSQv2.0

Core Symptom	Item	Question	Response Options
Syncope	9	Did you faint or lose consciousness during the past 24 hours?	0 = No 1 = Yes

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