

[Study Name/ID pre-filled]

Site Name: \_\_\_\_\_

Date Form Completed: \_\_\_/\_\_\_/\_\_\_ (mm/dd/yyyy)

Subject ID: \_\_\_\_\_

Visit: [pre-populated by database]

1. Was pulmonary function testing done?  Yes  No  
 If No, reason why not done: \_\_\_\_\_ (see below key)

### Reasons why assessment not done

1	Fatigue
2	Significant upper respiratory infection
3	Behavioral issues
4	Equipment failure
5	Child unable to follow directions
6	Low oral motor tone, unable to use mouthpiece
7	Unable to get subject into supine position: scoliosis, contractures, cannot move to bed (for supine only)
8	Other, specify _____

2. What type of pulmonary function testing is being performed? (Check all that apply. Record results in appropriate tables below):
- Slow Vital Capacity (sVC)
  - Maximum Inspiratory Pressure (MIP)
  - Maximum Expiratory Pressure (MEP)
  - Unassisted Peak Cough Flow (PCF)
  - Forced Vital Capacity (FVC)
  - Inspiratory Capacity (IC)

Note: Prior to beginning, subject needs to have a timed 10 minute rest period where they are sitting/laying comfortably. Position should remain consistent for all trials. Noseclips are required for all testing.

3. Position for the assessment:  
 Sitting  Supine (FVC only)
4. If assessment performed sitting, what was the subject's seated position?  
 Semi-erect  Erect  Leaning forward  N/A – assessment done supine
5. What type of mouthpiece was used (if applicable)?  
 Scuba  Cardboard
6. Ulna length: [pre-populated field] cm
7. Ulna length measured with:  
 Harrington clippers  Rosscraft segmometer  Other, specify \_\_\_\_\_
8. Type of Pulmonary Function Testing Equipment Used:  
 Manufacturer: \_\_\_\_\_  
 Model: \_\_\_\_\_  
 Software Program: \_\_\_\_\_

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Visit: [pre-populated by database]

### sVC Results

	Results
Trial 1	_____ L
Trial 2	_____ L
Trial 3	_____ L

### MIP Results

	Results
Trial 1	_____ cm H <sub>2</sub> O
Trial 2	_____ cm H <sub>2</sub> O
Trial 3	_____ cm H <sub>2</sub> O

### MEP Results

	Results
Trial 1	_____ cm H <sub>2</sub> O
Trial 2	_____ cm H <sub>2</sub> O
Trial 3	_____ cm H <sub>2</sub> O

### PCF Results

	Results
Trial 1	_____ L/min
Trial 2	_____ L/min
Trial 3	_____ L/min

### IC Results

	Results
Trial 1	_____ L
Trial 2	_____ L
Trial 3	_____ L

### FVC Results

	Results	Flow volume crossed X-axis?
Trial 1	_____ L	<input type="checkbox"/> Yes <input type="checkbox"/> No
Trial 2	_____ L	<input type="checkbox"/> Yes <input type="checkbox"/> No
Trial 3	_____ L	<input type="checkbox"/> Yes <input type="checkbox"/> No

#### Additional questions for MIP and MEP

1. Was there a difference of greater than 3cm of H<sub>2</sub>O in between trial efforts?    Yes    No

#### Additional questions for Forced Vital Capacity (FVC)

1. Was there a cough during the first second of exhalation?    Yes    No
2. Was there a leak during exhalation?    Yes    No
3. Was there early termination with steep cut off?    Yes    No
4. Are the 2 largest values for FVC within 0.100L of each other?    Yes    No

#### Missing Value Codes:

A = Lab or equipment failure  
M = Ran out of time  
N = No data  
X = Unknown  
E = Examiner error

O = Obsolete CRF  
D = Not applicable  
P = Unable to test due to permanent disability  
T = Unable to test due to temporary condition  
S = Scheduling problem

## GENERAL INSTRUCTIONS

This CRF contains data that would be collected when a pulmonary study is performed studying lung function.

Please note that the questions on Page 2 are for specific pulmonary function tests.

Important note: None of the data elements included on this CRF Module are classified as Core (i.e., strongly recommended for congenital muscular dystrophy clinical studies to collect if pulmonary studies are performed). All data elements are classified as supplemental (i.e., non Core) and should only be collected if the research team considers them appropriate for their study. Please see the Data Dictionary for element classifications.

## SPECIFIC INSTRUCTIONS

*Please see the Data Dictionary for definitions for each of the data elements included in this CRF Module.*

The CRF includes all instructions available for the data elements at this time. More detailed instructions will be added in Version 1.0 of this CRF Module.

- **Position for the Assessment** – Please note that “Supine” is for FVC only.
- **Sitting/Supine: Flow volume crossed X-axis** – Please note that this is for FVC only.
- **Ulna length** –If the Date performed on this form is the same as the Date performed on the Vital Signs form, then this field will be pre-populated from the value recorded on the Vital Signs form.
- **Missing Value Codes** – Please use these codes for all questions where a value is requested but no value is available. The purpose of these codes is to prevent any blanks on the CRF.