1. Date of Pulmonary Function Testing:

[ ] Not Done

Reason not done:

[ ]  Fatigue

[ ]  URI/LRI/ severe coughing

[ ]  Behavioral issues

[ ]  Equipment failure

[ ]  Unable to follow directions

[ ]  Low oral motor tone/unable to hold mouthpiece

[ ]  Unable to get subject into supine position due to scloiosis, contractures, cannot move to bed (for supine only)

[ ]  Other, specify:

1. 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)

[ ]  Helium lung volumes

[ ]  Sniff Nasal Inspiratory Pressure (SNIP)

[ ]  Maximal Voluntary Ventilation (MVV)

[ ]  Maximum Expiratory Pressure (MEP)

[ ]  Unassisted Peak Cough Flow (PCF)

[ ]  Forced Vital Capacity (FVC)

[ ]  Inspiratory Capacity (IC)

[ ]  Other, specify

Note: Position should remain consistent for all trials.

1. Position for the assessment:

[ ]  Sitting

[ ]  Supine (FVC only)

[ ]  Both (sitting and supine)

1. If assessment performed sitting, what was the subject’s seated position?

[ ]  Semi-erect

[ ]  Erect

[ ]  Leaning forward

[ ]  N/A – assessment done supine

1. What type of mouthpiece was used?:

[ ]  Scuba

[ ]  Cardboard

[ ]  Mask

1. Type of Pulmonary Function Testing Equipment Used:

Manufacturer:

Model:

Software program:

## Additional Pediatric-specific Elements

These elements are recommended for pediatric studies.

1. Ulna length: [pre-populated field] cm

Ulna length measured with:

[ ] Harpenden Anthropometer

[ ]  Rosscraft segmometer

[ ]  Other, Specify

1. Was patient taking brochodilator at time of testing?

[ ]  Yes

[ ]  No

Index of Lung Function Table

| Index of Lung Function | Trial 1Complete Exhalation[ ]  Yes [ ] No | Trial 2Complete Exhalation[ ]  Yes [ ] No | Trial 3Complete Exhalation[ ]  Yes [ ] No | BEST TRIAL |
| --- | --- | --- | --- | --- |
| FVC (liters) | [ ]  Yes [ ] No | [ ]  Yes [ ] No | [ ]  Yes [ ] No | [ ]  Yes [ ] No |
| FEV1 (liters) | [ ]  Yes [ ] No | [ ]  Yes [ ] No | [ ]  Yes [ ] No | [ ]  Yes [ ] No |
| FEV1/FVC (ratio/ no units) | [ ]  Yes [ ] No | [ ]  Yes [ ] No | [ ]  Yes [ ] No | [ ]  Yes [ ] No |
| FEV0.5 (liters) | [ ]  Yes [ ] No | [ ]  Yes [ ] No | [ ]  Yes [ ] No | [ ]  Yes [ ] No |
| FEV0.5/FVC (ratio/ no units) | [ ]  Yes [ ] No | [ ]  Yes [ ] No | [ ]  Yes [ ] No | [ ]  Yes [ ] No |
| FEF25-75 (liters/ second) | [ ]  Yes [ ] No | [ ]  Yes [ ] No | [ ]  Yes [ ] No | [ ]  Yes [ ] No |
| FEFmax (liters/ second) | [ ]  Yes [ ] No | [ ]  Yes [ ] No | [ ]  Yes [ ] No | [ ]  Yes [ ] No |
| PCF (liters/ second) | [ ]  Yes [ ] No | [ ]  Yes [ ] No | [ ]  Yes [ ] No | [ ]  Yes [ ] No |
| BEST TRIAL | [ ]  Yes [ ] No | [ ]  Yes [ ] No | [ ]  Yes [ ] No | [ ]  Yes [ ] No |
| TLC (liters) | [ ]  Yes [ ] No | [ ]  Yes [ ] No | [ ]  Yes [ ] No | [ ]  Yes [ ] No |
| SVC (liters) | [ ]  Yes [ ] No | [ ]  Yes [ ] No | [ ]  Yes [ ] No | [ ]  Yes [ ] No |
| IC (liters) | [ ]  Yes [ ] No | [ ]  Yes [ ] No | [ ]  Yes [ ] No | [ ]  Yes [ ] No |
| FRC (liters) | [ ]  Yes [ ] No | [ ]  Yes [ ] No | [ ]  Yes [ ] No | [ ]  Yes [ ] No |
| RV (liters) | [ ]  Yes [ ] No | [ ]  Yes [ ] No | [ ]  Yes [ ] No | [ ]  Yes [ ] No |
| FRC/TLC (ratio/ no unit) | [ ]  Yes [ ] No | [ ]  Yes [ ] No | [ ]  Yes [ ] No | [ ]  Yes [ ] No |
| RV/TLC(ratio/ no unit) | [ ]  Yes [ ] No | [ ]  Yes [ ] No | [ ]  Yes [ ] No | [ ]  Yes [ ] No |

## Additional questions for MIP/MEP

1. Was there a difference of greater than 3cm of H20 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

Was there a leak during exhalation?

[ ]  Yes

[ ] No

1. Are the 2 largest values for FVC within 10% of each other?

[ ]  Yes

[ ] No

1. Was there early termination with steep cut off?

[ ]  Yes

[ ] No

1. Was there a clearly defined peak flow?

[ ]  Yes

[ ] No

## 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 all CMD, DM and FSHD clinical studies to collect). All data elements are classified as Supplemental – Highly Recommended (i.e., non Core) and should be collected if the research team considers them appropriate for their study. Please see the Data Dictionary for element classifications.

FSHD General Instructions: The most useful measure of pulmonary function for FSHD is sitting and supine FVC. PFT is likely to have the greatest yield in clinical studies in which subjects are nonambulatory, have severe weakness or early (“infantile”) onset. Pulmonary function testing should be performed with a face mask in FSHD due to inability to form a tight seal on the mouthpiece. It is easy and inexpensive to perform.

## Specific Instructions

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

* Position for the Assessment – Please note that “Supine” 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.