1. Date of Pulmonary Function Testing (PFT)\* (yyyy-mm-dd):
2. What type of pulmonary function testing is being performed?\* (Check all that apply. Record results in appropriate tables on next page):

[ ]  Slow Vital Capacity (sVC)

[ ]  Maximum Inspiratory Pressure (MIP) – specify type below:

[ ]  Sniff Nasal Inspiratory Pressure (SNIP)

[ ]  Maximal Voluntary Ventilation (MVV)

[ ]  Maximum Expiratory Pressure (MEP)

[ ]  Peak Expiratory Flow (PEF)

[ ]  Forced Vital Capacity (FVC)

[ ]  Forced Expiratory Volume in 1 second (FEV1)

Note: A study should write detailed standard operating procedures (SOPs) for the PFT to help ensure consistent measurements. The SOP should cover details like the equipment used, the position of the participant/subject for the measurements, etc.

1. Position for the assessment:

[ ]  Sitting [ ]  Supine (FVC only) [ ]  Sitting and Supine

1. What type of mouthpiece was used (if applicable)?

[ ]  Scuba [ ]  Tube [ ]  Mask [ ]  Other

sVC Results\* [ ]  N/A MIP Results [ ]  N/A

Table 1 sVC Results

| Trial | Results\* (liters) | % of Predicted |
| --- | --- | --- |
| 1 | L  | % |
| 2 | L | % |
| 3 | L | % |

Table 2 MIP Results

| Trial | Results (cm H20) |
| --- | --- |
| 1 | cm H20ata to be entered by site |
| 2 | cm H20ta to be entered by site |
| 3 | cm H20a to be entered by site |

MEP Results **[ ]**  N/A PEF Results **[ ]**  N/A

Table 3 MEP Results

| Trail | Results (cm H20) |
| --- | --- |
| 1 | cm H20 |
| 2 | cm H20 |
| 3 | cm H20 |

Table 4 PEF Results

| Trial | Results (liters/min) |
| --- | --- |
| 1 | L/min |
| 2 | L/min |
| 3 | L/min |

FEV1 Results\* **[ ]**  N/A FVC Results\* **[ ]**  N/A

Table 5 FEV1 Results\*

| Trial | Results (liters) |
| --- | --- |
| 1 | L |
| 2 | L  |
| 3 | L |

Table 6 FVC Results\*

| Trial | Results\* (liters) | Flow volume crossed X-axis? | % Predicted Normal |
| --- | --- | --- | --- |
| 1 | Data to be entered by site | [ ]  Yes [ ]  No | TBD |
| 2 | Data to be entered by site | [ ]  Yes [ ]  No | TBD |
| 3 | Data to be entered by site | [ ]  Yes [ ]  No | TBD |

SNIP Results **[ ]**  N/A MVV Results **[ ]**  N/A

Table 7 SNIP Results

| Trial | Results (cm H20) |
| --- | --- |
| 1 | cm H20 |
| 2 | cm H20a to be e |
| 3 | cm H20  |
| Peak Pressure Value: cm H2O |

Table 8 MVV Results

| Trial | Results(liters/min) |
| --- | --- |
| 1 | L/min |
| 2 | L/min |
| 3 | L/min |

Nare: **[ ]**  Left **[ ]**  Right

Nasal Congestion: **[ ]**  Yes **[ ]**  No

## General Instructions

This CRF is good indicator of diaphragm strength because of its sitting vs. supine testing of FVC, which is of use in mitochondrial disease testing. In addition, the ratio of FEV1/FVC allows distinction between restrictive and obstructive ventilator defects.

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

Important note: The data elements noted with an asterisk on this CRF Module are classified as Supplemental – Highly Recommended (i.e., most recommended in relevant studies) for Mitochondrial Disease and Core (i.e., strongly recommended for all ALS studies). 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.

* Nare – This question is answered for SNIP
* Nasal Congestion – This question is answered for SNIP

\* Element is classified as Core for ALS and Supplemental – Highly Recommended for Mitochondrial Disease.