**Vision**

Right eye:  Normal  Abnormal, explain:

Left eye:  Normal  Abnormal, explain:

**Videonystagmography (VNG)** - Requires VNG system

1. \*\*Ocular Motor Assessment
   1. Saccades:
      1. Horizontal:  Normal  Abnormal, explain:
      2. Vertical:  Normal  Abnormal, explain:
   2. Smooth Pursuit:
      1. Horizontal:  Normal  Abnormal, explain:
      2. Vertical:  Normal  Abnormal, explain:
   3. Optokinetic:  Normal  Abnormal, explain:
   4. Gaze:
      1. Horizontal:  Normal  Abnormal, explain:
      2. Vertical:  Normal  Abnormal, explain:
   5. Spontaneous testing:
      1. Eyes open:  Normal  Abnormal, explain:
      2. Eyes closed:  Normal  Abnormal, explain:
2. \*\*\*Positional Testing
   1. Static Positions
      1. Supine:  Normal  Abnormal, explain:
      2. Head right:  Normal  Abnormal, explain:
      3. Head left:  Normal  Abnormal, explain:
      4. Lateral right:  Normal  Abnormal, explain:
      5. Lateral left:  Normal  Abnormal, explain:
   2. Dynamic (Dix-Hallpike) Positions
      1. Right:  Normal  Abnormal, explain:
      2. Left:  Normal  Abnormal, explain:
3. \*\*\*Caloric Irrigations - Warm and Cool Irrigations (Alternating Binaural Bithermal Irrigations)
   1. Peak Slow-phase Eye Velocity (PSEV)
      1. Right warm:
      2. Left warm:
      3. Right cool:
      4. Left cool:
   2. % Asymmetry:
   3. % Preponderance:

**\*\*\*Vestibular Evoked Myogenic Potential (VEMP) Testing** - Requires VEMP (Evoked Potential) System

|  |  |  |
| --- | --- | --- |
| **Cervical VEMP** | **Right** | **Left** |
| **P1 Latency** | Data to be filled in by site | Data to be filled in by site |
| **N1 Latency** | Data to be filled in by site | Data to be filled in by site |
| **P1 - N1 Amplitude** | Data to be filled in by site | Data to be filled in by site |
| **P1 - N1 Amplitude Ratio** | Data to be filled in by site | |

Sternocleidomastoid (SCM) muscle activity was monitored during testing?  Yes  No

| **Ocular VEMP** | **Right** | **Left** |
| --- | --- | --- |
| **P1 Latency** | Data to be filled in by site | Data to be filled in by site |
| **N1 Latency** | Data to be filled in by site | Data to be filled in by site |
| **P1 - N1 Amplitude** | Data to be filled in by site | Data to be filled in by site |
| **P1 - N1 Amplitude Ratio** | Data to be filled in by site | |

30-degree upward gaze (or other degree) was performed by the participant during testing?  Yes  No

**\*\*\*Rotational Vestibular Testing (RVT)** - Requires Rotational Chair

1. Sinusoidal Harmonic Acceleration (SHA)

|  |  |  |  |  |
| --- | --- | --- | --- | --- |
| **Angular Rotations**  **(All or every other starting with 0.01 Hz)** | **Velocity** | **VOR Gain** | **VOR Phase** | **VOR Symmetry** |
| **0.01 Hz** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **0.02 Hz** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **0.04 Hz** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **0.08 Hz** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **0.16 Hz** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **0.32 Hz** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **0.64 Hz** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |

1. \*\*\*Velocity Step/Impulse Test

|  |  |  |
| --- | --- | --- |
| **Step Accelerations 60 d/s** | **PSEV** | **Time Constant** |
| **Acceleration to the right** | Data to be filled in by site | Data to be filled in by site |
| **Deceleration from the right** | Data to be filled in by site | Data to be filled in by site |
| **Acceleration to the left** | Data to be filled in by site | Data to be filled in by site |
| **Deceleration from the left** | Data to be filled in by site | Data to be filled in by site |

|  |  |  |
| --- | --- | --- |
| **Step Accelerations 240 d/s** | **PSEV** | **Asymmetry** |
| **Acceleration to the right** | Data to be filled in by site | Data to be filled in by site |
| **Deceleration from the right** | Data to be filled in by site | N/A |
| **Acceleration to the left** | Data to be filled in by site | Data to be filled in by site |
| **Deceleration from the left** | Data to be filled in by site | N/A |

1. **\*\*\*Limits of Stability (LOS)** - Assess when possible
   1. Maximum sway excursion %
      1. Forward:
      2. Forward right:
      3. Right:
      4. Backward right:
      5. Backward:
      6. Backward left:
      7. Left:
      8. Forward left:

**\*\*\*Vestibular Head Impulse Testing (vHIT)** - Requires vHIT System

1. Lateral Canals
   1. VOR gain:
   2. #Overt/covert saccades:
2. Vertical Canals
   1. LARP Planes:
      1. VOR gain:
      2. #Overt/covert saccades:
   2. RALP Planes
      1. VOR gain:
      2. #Overt/covert saccades:

Recorder Signature: Date:

## General Instructions

Vestibular manifestations of mitochondrial disease are relatively rare. This case report form (CRF) contains data elements that collect information related to the characterization of vestibular function and that assess its severity in the affected individual. Questions on this CRF are well validated and easily applied to both pediatric and adult populations. Data may be collected at multiple time points to show progression over time. Normative reference ranges may vary depending upon methods of recording and/or stimuli used and consideration should be given to these variables.

Important note: None of the data elements included on this CRF Module are classified as Core (i.e., strongly recommended for all mitochondrial disease clinical studies to collect). Data elements are classified as Supplemental – Highly Recommended (i.e., essential information for specified conditions, study types, or designs) or Exploratory, as indicated by asterisks below

\*\*Element is classified as Supplemental – Highly Recommended

\*\*\*Element is classified as Exploratory

The remaining 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.

* Sinusoidal harmonic acceleration (SHA) velocity – SHA testing is usually performed at either 50 or 60 d/s.