The Pediatric FSHD Pulmonary Assessment Master Outline is intended to provide a comprehensive overview of data elements comprising a complete pulmonary assessment for a study patient. Such an assessment includes data from a variety of clinical areas, including a patient’s Demographics, History, Physical Exam, Diagnostic Test Results and Therapeutic Interventions, some of which may be found under the NINDS Neuromuscular Diseases CDE recommendations. Details regarding most of the specific data elements in the Pulmonary Assessment guidelines can be found in various CRFs, which are referenced in the document below. If an item or group of items are not relevant to your hypothesis or study design, then you do not need to include them in your study.

1. Demographics
2. Diagnosis – study-specific variable, not included in recommendations
3. Age/DOB – see Demographics CRF
4. Race/Ethnicity – see Demographics CRF
5. Gender – see Demographics CRF
6. Education Level (Patient and Parents/Guardians) – see Demographics CRF for patient education; Parents/ Guardians education not included in the recommendations
7. Social history (living situation, caregivers, home nursing, socioeconomic, medical insurance status) – see Social Status CRF
8. History
9. Date of diagnosis/age at diagnosis – see Medical History CRF
10. Family History (including non-neuromuscular pulmonary diseases, like asthma) – see Family History CRF – Pulmonary diseases would fall under “other” category
11. Method of diagnosis
12. Presenting signs and symptoms – study-specific variables, not included in the recommendations
13. Blood tests (like CK) – see Laboratory Tests CRF
14. Biopsy results (like muscle biopsy, including method of biopsy and testing) – see Biopsy CRFs (Muscle, Nerve, Skin, Fat Aspirate)
15. Genetic results (and details of genetic test used) – see Mutation Analysis CRF
16. “Sentinel Events” (Note: include date/age for all Sentinel Events)
17. Date/age at initiation of corticosteroids. – study-specific variables, not included in the recommendations
18. Date/age of loss of ambulation – study-specific variables, not included in the recommendations
19. Date/age of spinal fusion or chest wall surgery (including pre-op evaluation method, anesthesia and intra and periop resp support method, surgical method) – see Medical History Form
20. Date/age of initiation of assisted ventilation (and method – non invasive vs. trach/vent) – see Respiratory Interventions CRF
21. Date/age of tracheostomy, if applicable – see Respiratory Interventions CRF
22. Date/age of major cardiac intervention (e.g. pacemaker, defibrillator) – see Medical History Form
23. Date/age of G tube of other feeding tube (G-J, NG, etc.) – see GI CRF
24. Date/age of death – see Death Report CRF
25. Patient history at time of each clinic visit
26. Respiratory symptoms, such as dyspnea, cough or wheezing – study-specific variables, not included in the recommendations
27. Interval course, including hospitalizations (respiratory or non-respiratory), sick outpatient visits (respiratory or non respiratory) with dates and causes – study-specific variables, not included in recommendations
28. Therapies in use, including pharmacologic and mechanical – and compliance – see Concomitant Medications Form
29. Therapies started or stopped since last visit and dates –see Concomitant Medications CRF
30. Check list of symptoms of hypoventilation (dyspnea, morning headaches, new fatigue, increased napping, restless sleep, enuresis, deifficulty concentrating, change in school performance, dysthymia/depression, etc.) – study-specific variables, not included in recommendations; individual events may be included on the AE CRF
31. History indicating ineffective cough (persistent chest congestion with colds (>2 weeks), recurring bronchitis, etc.) – see Medical History CRF
32. Resuscitation status and End of Life decisions/documents – study-specific variables, not included in recommendations
33. Environmental history (especially personal and passive cigarette smoking) – see Behavioral History CRF (General Form)
34. Physical Exam Findings
35. General appearance (sitting vs. wheelchair; thin vs. overweight; dyspneic vs. comfortable) – may be documented on the Physical Exam CRF
36. VS including weight, height (specify standing or recumbent “heel to crown,” BMI, arm span, ulnar length, temp, BP, respiratory and heart rate, Oxygen Saturation, End-Tidal CO2) – see Vital Signs CRF
37. Breathing pattern (shallow, abdominal, presence or lack of paradoxical breathing, grunting, flaring, subcostal and/or supasternal retractions; wet or dry cough; throat clearing, etc.) – see Vital Signs CRF
38. Chest wall shape and character (excessive stiffness or pliability, bell-shaped, pectus excavatum or carinatum, Harrison’s grooves, asymmetry, effects of scoliosis – see G below). – study-specific variables, not included in the recommendations
39. Lung exam (symmetry of aeration, focal decrease in aeration, crackles, wheeze, rhonchi) – see Physical Exam CRF
40. Heart exam (S1, S2, rhythm, gallop, thrill, murmur) – study-specific variables, not included in the recommendations; could refer to cardiac CRFs 24 holter, MRI, Echocardiogram, and ECG CRFs
41. Scoliosis and/or kyphosis –see Clinical Outcomes recommendations
42. Respiratory and related diagnostics (with dates and results for each)
43. Pulmonary Function Tests: Supplemental – Highly Recommended CDEs with detailed real-time methods/flow diagrams and post-collection criteria for validation – see Pulmonary Function Test CRF
44. Measure of gas exchange with date and results and location sleep lab, home, clinic – See Gas Exchange and Polysomnography CRF
45. Radiographs (chest X-ray, chest CT; spine series, swallow/dysphagia studies, reflux studies, other) – study-specific variables, not included in the recommendations
46. Other relevant lab data (e.g. serum bicarbonate; nutritional parameters like serum Albumin/pre albumin, BNP. – See Laboratory Tests CRF
47. Therapies and Interventions. Include all start and stop dates, correlated to (II E 4) above
48. Pharmacologic therapies, including steroids with formulation, start date/age; and including homeopathic and complementary medicine therapies – see Concomitant Medications CRF
49. Respiratory therapies – See Respiratory Therapies CRF
50. Airway clearance and secretion mobilization devices (with start and stop dates)
51. Type (including brand and model)
52. Settings, if applicable
53. Frequency/daily hours/schedule of use
54. Compliance (objective, such as hour-meter; subjective, and source – pt, parent)
55. Respiratory support/assisted ventilation devices (start and stop dates)
56. Type (including brand and model)
57. Modality (non-invasive, trach)
58. Settings
59. Frequency/daily hours/schedule of use
60. Compliance (objective, such as hour-meter; subjective, and source – pt, parent)
61. Oxygen (start and stop dates)
62. Method of administration (trach, bled into NIV/vent, nasal cannulae)
63. Flow rate
64. Schedule and frequency/hours per day
65. Respiratory medications (start and stop dates)
66. Bronchodilators (name, dose, frequency)
67. Anti cholinergic and secretion management (Robinul, patch, ipratropium, other, etc.)
68. Inhaled Corticosteriods
69. Cardiac therapies (start and stop dates) – study-specific variables not included in the recommendations; drug therapies may be re-coded on the Concomitant Medications CRF
70. Pharmacologic, including medication, date of onset, and dose
71. “Mechanical’ including defibrillators, pacemakers
72. GI therapies (start and stop dates) – see GI CRF
73. Feeding including estimated intake if oral; and feeding schedule if via tube, with brand/caloric density of formula
74. Volumes
75. Oral
76. Tube
77. Nasogastric
78. Nasojejunal
79. Gastronomy
80. Gastrojejunal
81. Antireflux
82. Nissen fundoplicatoin and date/age (method: laparoscopic, laparotomy)
83. Medication
84. PPI
85. H2 blocker
86. Gastric motility
87. Other therapies relevant for the respiratory system (start and stop dates) – see Respiratory Interventions CRF
88. Physical therapy related to respiration and type (frequency)
89. Aquatic therapy, other
90. Adverse Events (see Adverse Events CRF)
91. Include inpatient and outpatient complications of respiratory and related Interventions such as noninvasive ventilation, Spinal fusion, G tube surgery, cardiac device placement and adverse events related to both mechanical and pharmacologic therapies.