

NEUROMUSCULAR WEAKNESS (PULMONARY)

REFERRAL GUIDELINE

MAINE MEDICAL PARTNERS - PEDIATRIC SPECIALTY CARE (DIV. PULMONARY MEDICINE) • 887 CONGRESS ST, SUITE 320, PORTLAND, ME • (207) 662-5522

HIGH RISK

SUGGESTED EMERGENT CONSULTATION

SYMPTOMS AND LABS

Repeated hospital admissions for pulmonary infections or a single admission to the intensive care unit

Change in feeding tolerance

Loss of mobility

Infants with weakness and feeding difficulties

Infants with thoracic dystrophy

Weakness with reliance on oxygen

SUGGESTED PREVISIT WORKUP

RECOMMEND:

Consult pulmonary medicine

Urgent consults are to expedite assessment of underlying anatomic or medical conditions that could be compromise airway patency

The intention is to prevent a life threatening complication and initiate therapies to reduce morbidity

MODERATE RISK

SUGGESTED CONSULTATION OR CO-MANAGEMENT

SYMPTOMS AND LABS

SYMPTOMS AND EXAM:

Progressive Thoracic Weakness

Onset of dysphagia

Recurrent pneumonia (two or more in a year)

Prolonged respiratory infections (>2 weeks)

Disrupted sleep

Headache

Labs/imaging

Elevated bicarbonate

Elevated hemoglobin/hematocrit

Onset or progression of thoracic scoliosis

SUGGESTED WORKUP

Swallow study with speech therapy

Chest x-ray to assess scoliosis

Follow up x-ray for pneumonias to ensure that consolidations are secondary to infection and not anatomic changes related to neuromuscular scoliosis

Speech and swallow center referral

Pulmonary referral is indicated

Consider GI referral

LOW RISK

SUGGESTED ROUTINE CARE

SYMPTOMS AND LABS

SYMPTOMS AND EXAM:

Thoracic weakness (cerebral palsy, muscular dystrophy, etc.)

Oral Motor Weakness (drooling and speech impediments)

Non-progressive neuromuscular disease

Labs:

Normal bicarbonate

Normal hemoglobin/hematocrit

Normal chest x-ray

SUGGESTED MANAGEMENT

MONITOR FOR:

Onset of progressive weakness

Difficulty with respiratory infections

Safety of swallow

Onset and progression of scoliosis

Symptoms of disrupted sleep

Snoring

Daytime irritability

Increasing daytime sleepiness

Concern for apnea

Consider one time early pulmonary consult to help determining timing and need for future studies and interventions

CLINICAL PEARLS

- Early pulmonary intervention in neuromuscular respiratory failure has been demonstrated to reduce morbidity and mortality and improve quality of life.
- Children with neuromuscular disease who snore and stop snoring may be developing worsening airway obstruction.

British Thoracic Society guideline for respiratory management of children with neuromuscular weakness. Thorax. 2012;67:i1e140

Respiratory Management of Pediatric Patients with Neuromuscular Disease. Pediatric Annals. December 2010 - Volume 39 · Issue 12: 769-776

Maine Medical
PARTNERS

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