

NEUROMUSCULAR DISORDERS

Muscular dystrophies, spinal muscular atrophy, congenital myopathies, and congenital myasthenic syndromes ([ICD10 codes](#))

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TARGET POPULATION

Inclusion Criteria

- Patients followed in the neuromuscular clinic (aka 'muscle clinic') or who have the following ICD10 diagnosis codes; G71, G11, G12, and G60 (see [Neuromuscular ICD10 Codes](#) for full listing)
- Patients who have a neuromuscular consult ordered or who have a diagnosis of Duchenne muscular dystrophy, spinal muscular atrophy, myotonic dystrophy, congenital myopathy, congenital muscular dystrophy, Friedreich's ataxia, and congenital myasthenic syndrome
- Patients newly diagnosed with a neuromuscular disorder

Exclusion Criteria

- Patients with cerebral palsy, spina bifida, dystonia, or other causes of muscle weakness

CLINICAL ASSESSMENT AND MANAGEMENT

Emergency Department

Initial Assessment

- Vital signs, SpO2, CR monitor
- Immediate clinical evaluation

See [Pre-Operative Evaluation](#) for any patient requiring sedated procedures.

Respiratory: High risk for pneumonia and acute respiratory failure.

- For respiratory distress or hypoxemia, use Cough Assist and start biphasic non-invasive ventilation (BiPAP or AVAPS). Use patient's own device with their "sick settings" if they brought it
- Hypoxemia should not be treated with simple oxygen
- Obtain chest X-ray, ECG, blood gas, renal function panel
- Call pulmonary service

Antimicrobials for respiratory infections ([Table 1.](#) and [Table 2.](#))

Cardiac: Directed at underlying cardiomyopathy and/or rhythm disturbance management.

- Obtain chest X-ray, ECG, blood gas, renal function panel.
- Check INR if on warfarin, digoxin level if on digoxin.
- Contact the cardiomyopathy service.

Endocrine

- Patients who are on chronic oral steroids may need stress dosing for illnesses. [Visit this website](#) or [click this link](#) for more information.

GI/Nutrition

- In SMA and other conditions, consider hypoglycemia if NPO for more than 4-6 hours or if expected to be NPO.
- Patients may need D10-containing fluids. There is no evidence for protein or lipid infusion in acute illnesses.
- Abdominal pain from constipation and/or dysmotility is common. Obtain abdominal film. Treat constipation with MiraLAX, milk of magnesia, or lactulose.

Admission Criteria

- Respiratory needs exceed what family can provide at home, use of non-invasive ventilation around the clock if not doing so at baseline, FiO₂ over 50%, frequent cough assist, concern for worsening status, new hypercapnia.
- Cardiac: new or worsening heart failure or dysrhythmia.
- Endocrine: need for stress steroids in the setting of acute illness.
- GI/nutrition: dehydration requiring initiation of NG feeding or IV fluids, persistent or unexplained hypoglycemia, worsening failure to thrive, metabolic acidosis.
- Discuss appropriate location with pulmonary, cardiology, and ICU.

Discharge from ED Criteria

- None of the admission criteria are met. Signs and symptoms to watch for include: worsened work of breathing, new oxygen requirement, dyspnea, chest pain, edema, dizziness, or syncope. Prompt follow up with PCP and neuromuscular clinic is recommended. Please notify the Neuromuscular Program Coordinator (Alison Ballard) via EMR of the patient's evaluation in the ED.

Inpatient Setting

See [Pre-Operative Evaluation](#) for any patient requiring sedated procedures.

Respiratory

Principles of respiratory support in neuromuscular disease

- Must be treated with cough assist in those with weak cough and non-invasive ventilation. This should be done regardless of respiratory rate, oxygen saturation, or blood gas values.
- Simple oxygen alone should not be used for hypoxemia as this can cause worsened hypercapnia in patients with chronic hypercapnic respiratory failure.
- CPAP and HHFNC are not appropriate, as bi-level support is needed to provide sufficient tidal volumes and optimal rest.
- Patients with SMA type I should lay supine. They are dependent on the diaphragm to breathe. They are at a mechanical disadvantage in the upright position. Supine positioning should not change in the setting of gastric feeding with or without Nissen fundoplication.

Airway Clearance

- Goal is to address weak cough. This should be done with mechanical in-exsufflation (commonly known as Cough Assist). Cough Assist pressures should exceed NIV inspiratory pressures by at least 5-10 cm H₂O. If possible, the patient's own device should be used. Cough Assist should not be withheld due to concerns for frequent removal of non-invasive ventilation.
- Initial airway clearance need not include vibratory therapies (e.g. vest, IPV, or chest physiotherapy), mucolytics (e.g. hypertonic saline, dornase alfa, N-acetylcysteine) or bronchodilators (albuterol, levalbuterol, ipratropium). There is no evidence that these are effective. Furthermore, mucus properties are normal and do not need to be thinned if the patient is well-hydrated.

Antimicrobials for Respiratory Infections ([Table 1.](#) and [Table 2.](#))

Cardiac

Cardiomyopathy

- Strict monitoring of intake and output.
- Contact cardiomyopathy service if not already done in ED or if concerned about new or worsened disease.
- Medications should not be adjusted or initiated without cardiomyopathy service involvement.

- ACE inhibitors should be held in cases of dehydration, renal insufficiency or injury, or hypotension.
- Avoid using NSAIDs while on ACE inhibitors or if volume depleted due to risk of renal injury. Tylenol can be used for pain or fever.
- Diuretics are only given to patients with symptomatic heart failure related to volume overload.

Electrical Disturbances

- Please consult cardiology for all patients with possible electrical disturbances. Patients with myotonic muscular dystrophy are at high risk for rhythm disturbances and should be on a CR monitor. An ECG should be obtained for any acute cardiac or respiratory complaint.
- Patients with implantable cardioverter defibrillator devices (ICD) or pacemakers should have the device interrogated for any acute cardiac or respiratory complaint.

Endocrine

- Patients with Duchenne muscular dystrophy and other neuromuscular disorders are often on chronic oral steroids. Long-term corticosteroid use may cause hypothalamic-pituitary-adrenal (HPA) axis suppression. Stress doses of corticosteroids may be needed in the perioperative period and during acute illness to prevent acute adrenal insufficiency or adrenal crisis. Click below for more information.
<https://www.parentprojectmd.org/wp-content/uploads/2018/03/PJ-Nicholoff-Steroid-Protocol.pdf>

GI/Nutrition

Key Points

- Children with neuromuscular disease are at risk for malnutrition (both over- and undernutrition, depending on disease status), swallowing difficulties, constipation, gastroesophageal reflux, and gastrointestinal dysmotility.
- Because of the high incidence of nutritional challenges, a Registered Dietitian (RD) should be involved in the care of children with neuromuscular disease from the time of diagnosis through the life course.
- A length measurement or height estimate is critical for the assessment of nutritional status. At each clinic visit, a standing height measurement should be obtained for ambulatory patients. For non-ambulatory patients, both segmental length and ulnar length should be obtained.
- Swallowing dysfunction is extremely common. When signs or symptoms of dysphagia are detected, or if patients are not maintaining their weight or malnourished, a Speech therapist should be consulted and Video Fluoroscopic Swallowing Study (VFSS or Modified Barium Swallow) obtained.
- Optimization of nutritional status is particularly important before any surgical procedures to improve wound healing and prevent pressure sores.
- A gastrostomy tube should be strongly considered for children with malnutrition, aspiration, or moderate/severe dysphagia.
- As noted above, children with severely reduced muscle mass are potentially at risk of hypoglycemia during prolonged fasting. This fact should be considered in the peri-operative environment, particularly if there is a prolonged period without enteral nutrition in the post-operative state. Children can be given glucose-containing clear liquids enterally up to 2 hours prior to general anesthesia.

Common nutritional concerns to be addressed by the team, including the RD

- Inadequate hydration
- Tube feeding management
- Feeding and swallowing difficulties, including aspiration into respiratory tract
- Suboptimal weight gain in infancy
- Underweight due to low muscle mass or malnutrition

- Overweight
- Vitamin D insufficiency/deficiency and/or insufficient calcium intake

Specific Nutrition Goals by Disease Type

- **Congenital Muscular Dystrophies and Congenital Myopathies**
 - Individualize energy needs based on history and growth (can be at, below or above RDA)
 - Protein needs = RDA
 - Fluid needs = maintenance
 - Adequate calcium intake to meet RDA, vitamin D intake as indicated to maintain normal 25-OH vitamin D levels
- **Duchenne Muscular Dystrophy (DMD)**
 - At diagnosis/up to age 7/10 years old: energy and protein at RDA for age
 - Ages 11-14 years old: Schofield weight equation for energy needs – decreased for lower muscle mass and loss of ambulation - steroid treated ambulatory boys with DMD $17.7 \times \text{wt (kg)} + 657 = \text{REE} \times \text{AF}$
 - Ages 15-18 years old: $\text{REE} \times 0.8$ for decreased muscle mass/immobility $\times \text{AF}$
 - Fluid needs = maintenance
 - Adequate calcium intake to meet RDA, vitamin D intake as indicated to maintain normal 25-OH vitamin D levels
- **Spinal Muscular Atrophy (SMA)**
 - Energy: 6-11 kcal/cm or 60-90% of DRI for Type I; 9-14 kcal/cm or 90-100% DRI for Type II
 - Protein: 1.6-2.4 g/kg infants; 1-2 g/kg children and adults
 - Fat: 45% of calories for infants; 30% of calories or less age 1 and above
 - Fluids: maintenance or increased to as much as 115 ml/kg if indicated by medical team
 - Adequate calcium intake to meet RDA, vitamin D intake as indicated to maintain normal 25-OH vitamin D levels

Common Gastrointestinal Problems in Neuromuscular Disease Patients

- **Constipation**
 - Risk Factors include dehydration, immobility, gastrointestinal dysmotility, use of narcotic analgesics, and scoliosis.
 - Frequently underdiagnosed and undertreated in neuromuscular disease patients.
 - Nutritional goals include ensuring patient is meeting hydration goals and fiber intake goals
 - Mainstay of treatment is osmotic laxative therapy using agents such as Polyethylene Glycol 3350 (Miralax), Milk of Magnesia, and lactulose.
- **Gastroesophageal Reflux**
 - Risk Factors include scoliosis, delayed gastric emptying.
 - Treatment options include proton pump inhibitors (omeprazole, eg) or Histamine 2 receptor antagonists (ranitidine, eg). Use of proton pump inhibitors must be balanced against recognized increase in risk with use for community acquired pneumonia, *Clostridium difficile* associated diarrhea, and bone fracture.
- **Gastrointestinal Dysmotility**
 - Includes delays in gastric emptying and small/large bowel transit contributing to feeding intolerance and constipation.
 - More likely seen with older children/adults with advanced disease.

- Treatment includes adjustment of feeding strategy, optimization of constipation management, and referral for gastroenterology consultation.

Rehabilitation | Physical Therapy

- **Rehabilitation:** Use the neuromuscular consult order to contact the rehabilitation service. The Neuromuscular Program Coordinator will evaluate for any rehabilitation MD/NP needs.
- **Physical/Occupational Therapy:** Consult with team and family for any new needs with mobility, transfers, range of motion (stretching) Blanket order for all patients is not needed
- **Equipment/DME:** Consult the Neuromuscular Program Coordinator and/or PT/OT can consult for any new outpatient/home equipment needs or equipment modifications because of change in function. Typically families will already have a vendor in place.
- **ADA rooms:** Overhead Guldman ceiling lift, shower (not tub) if not ambulatory (acutely or chronically) – rooms 601, 602, all of 6E, 648, 947, 948, PICU 14, 15, 18, 19). Guldman lifts require disposable slings. They can be obtained from Level 6 clinical practice specialist (7-6965 Level 6), Level 9 clinical practice specialist (7-1828 or charge can access on nights and weekends), PICU charge 7-6509, or sling from home. Will need to get appropriate size.
- **Hoyer lift:** Standard on levels 6 and 8 (not on 9 or in ICU) and can use disposable slings or encourage family to bring their own sling from home.
- **Bath/toilet equipment:** Adaptive reclining rolling shower/commode chair (some have padded drop down arms for lateral support and higher backs when needed). Call PT chart 7-8505 during daytime hours, availability is limited.
- **Appropriate bed:** Often will need alternating pressure mattress, may require bariatric equipment.
- **Chronic pain:** Patient or family should have management plan in place for chronic pain. Caution must be used to balance pain medications with complex cardiopulmonary disease and constipation risks.

Neurology

The neurologist provides:

- Knowledge about the disorder and standards of care
- Detailed examinations to document changes or progression of disease
- Any additional diagnostic work up that might be needed
- Management of steroids primarily for those with dystrophinopathies
- Interpretation of laboratory tests

Psychosocial Aspects of Chronic Disease

- Use the neuromuscular consult order or a social work order to involve the neuromuscular team social worker for psychosocial assessment.
- Note: A psychosocial assessment is completed by a clinical social worker and should include evaluation of the family support system, plan for family and in-home supports post- hospitalization, plan for transportation home in preparation for discharge, school issues/concerns, family mental health issues/concerns, discussions regarding advance directives, and provision of resources.

Discharge Criteria

- Discharge criteria should be determined on a case-by-case basis, as recommended criteria differ based on underlying disease state, level of baseline ventilator support, secretion clearance needs, family abilities, and psychosocial factors. Often nursing and respiratory therapy needs factor heavily into this decision. Detailed discussion between the family, outpatient providers (ie, neuromuscular team members), and home healthcare providers is mandatory.

- Return precautions: worsened work of breathing, new oxygen requirement, dyspnea, chest pain, edema, dizziness, or syncope. Prompt follow up with PCP and neuromuscular clinic is highly recommended. Please notify the Neuromuscular Program Coordinator (Alison Ballard) via EMR of the patient's evaluation in the ED.
- Discharge from hospital often requires significant coordination and new respiratory or PT equipment from DME companies. Obtaining new equipment may take 1-14 days, depending on equipment, patient location, and insurance.
- The DME coordinator x75881 can help with ordering, insurance verification and delivery. NOTE: this typically takes at least 24 hours and should be planned for in advance.
- Family may require additional help at home including home physical therapy and/or home nursing care. Please verify needs with physical therapist. The inpatient case manager can help set this up prior to discharge.

Intensive Care Unit

See [Pre-Operative Evaluation](#) for any patient requiring sedated procedures.

Respiratory

Principles of respiratory support in neuromuscular disease

- Initial treatment of hypoxemia or respiratory distress should include aggressive mechanical support with bi-level non-invasive or invasive devices and mucus clearance. While hypoxemia may respond to increases in FiO₂ or continuous positive airway pressure, these interventions can worsen hypercapnia, blunt respiratory drive, and lead to worsening acute respiratory failure.
- Must treat with Cough Assist in those with weak cough and non-invasive ventilation. This should be done regardless of respiratory rate, oxygen saturation, or blood gas values.
- Biphase non-invasive ventilation is the preferred mode of initial escalated respiratory support. For children on non-invasive support at baseline, use "sick settings" as a starting point. If sick settings are unavailable, titrate support with below goals in mind.
- CPAP and HHFNC are not appropriate initial therapies for escalation but may have a limited role during de-escalation of support.
- Typical Biphase NIV goals include an exhaled V_t of ~10 ml/kg, and an inspiratory to expiratory delta P of at least 10 cm H₂O.
- PC mode can be used to regulate inspiratory time and provide maximal rest.
- For intubated patients who do not meet ARDS criteria, the same tidal volume and provision of rest goals apply.
- Extubation readiness:
 - Goals for extubation should be to extubate to biphase non-invasive ventilation. For patients previously on NIV at baseline, would target ventilator settings that are similar to their sick settings.
 - Particular attention should be paid to ventilator-patient synchronization and trigger sensitivity as neuromuscular patients often have difficulty triggering ventilators.
- Patients with SMA type I should lay supine (rather than HOB at 30°). They are dependent on the diaphragm to breathe. They are at a mechanical disadvantage in the upright position. Supine positioning should not change in the setting of gastric feeding with or without Nissen fundoplication.

Airway clearance

- **Non-Invasive Mechanical Ventilation**
 - Goal is to address weak cough. This should be done with mechanical in-exsufflation (commonly known as Cough Assist). Cough Assist pressures should exceed NIV inspiratory pressures by at least 5-10 cm H₂O. If possible, the patient's own device should be used. Cough Assist should not be withheld due to concerns for frequent removal of non-invasive ventilation, as secretion clearance is vital to improvement.

- Initial airway clearance need not include vibratory therapies (e.g. vest, IPV, or chest physiotherapy), mucolytics (e.g. hypertonic saline, dornase alfa, N-acetylcysteine) or bronchodilators (albuterol, levalbuterol, ipratropium). There is no evidence that these are effective. Furthermore, mucus properties are normal and do not need to be thinned if the patient is well-hydrated.
- If vibratory therapies or mucolytics are used, they must be followed by Cough Assist and suction.
- Bronchoscopy should be avoided as sedation and elective intubation may lead to inability to extubate. Aggressive airway clearance must be used prior to consideration of elective intubation and bronchoscopy.
- **Invasive Mechanical Ventilation**
 - Goal is to address weak cough. This should be done with mechanical in-exsufflation (commonly known as Cough Assist). Cough Assist pressures should exceed PIP by at least 5-10 cm H₂O, up to suggested maximum of 35 cc H₂O. If possible, the patient's own device should be used. Cough Assist should not be withheld due to concerns for frequently disconnecting the ventilator circuit, as secretion clearance is vital to improvement.
 - Initial airway clearance need not include vibratory therapies (e.g. vest, IPV, or chest physiotherapy), mucolytics (e.g. hypertonic saline, dornase alfa, N-acetylcysteine) or bronchodilators (albuterol, levalbuterol, ipratropium). There is no evidence that these are effective. Furthermore, mucus properties are normal and do not need to be thinned if the patient is well-hydrated.
 - If vibratory therapies or mucolytics are used, they must be followed by Cough Assist and suction.
 - Bronchoscopy should be considered early in cases of focal lobar or segmental atelectasis or mucus plug.

Cardiac

Cardiomyopathy

- Strict monitoring of intake and output.
- Contact cardiomyopathy service if not already done in ED or if concerned about new or worsened disease.
- Chronic cardiomyopathy medications should not be adjusted or initiated without cardiomyopathy service involvement.
- ACE inhibitors should be held in cases of dehydration, renal insufficiency or injury, or hypotension.
- Avoid using NSAIDs while on ACE inhibitors or if volume depleted due to risk of renal injury. Tylenol can be used for pain or fever.
- Diuretics are only given to patients with symptomatic heart failure related to volume overload.
- Obtain digoxin levels and INR for patients on digoxin or warfarin, respectively.
- Inotropic therapy should be considered only in cases of symptomatic heart failure (severe ventricular systolic dysfunction, low blood pressure, or decreased perfusion AND evidence of decreased cardiac output, either in the presence or absence of volume overload). Consultation with cardiomyopathy team is recommended prior to initiation of therapy. Use of inotropic medications for asymptomatic heart failure in neuromuscular patients is potentially harmful.

Electrical disturbances

- Please consult cardiology for all patients with possible electrical disturbances.
- Patients with myotonic muscular dystrophy are at particularly high risk for a variety of rhythm disturbances. An ECG should be obtained for any acute cardiac or respiratory complaint.
- Patients with implantable cardioverter defibrillator devices (ICD) or pacemakers should have the device interrogated for any acute cardiac or respiratory complaint.

Endocrine

- Patients with Duchenne muscular dystrophy and other neuromuscular disorders are often on chronic oral steroids. Long-term corticosteroid use may cause hypothalamic-pituitary-adrenal (HPA) axis suppression. Stress doses of corticosteroids may be needed in the perioperative period and during acute illness to prevent acute adrenal insufficiency or adrenal crisis. [Click this link](#) for more information.

GI/Nutrition

- Goal is to provide as little interruption to enteral nutrition as possible, given vulnerability to catabolic state without constant protein delivery. Consider early enteral feeds with as rapid an advance to full feeds as tolerated.
- Consult RD to determine optimal feeding regimen.
- Continuous IV dextrose administration should be initiated to avoid hypoglycemia in any patient who is expected to be NPO for 4-6 hours. D10 fluids may be necessary regardless of age.

[Rehabilitation | Physical Therapy](#)

[Neurology](#)

[Psychosocial Aspects of Chronic Disease](#)

Criteria for Transfer out of ICU

- Transfer from ICU to floor should be determined on a case-by-case basis, as recommended criteria differ based on underlying disease state, level of baseline ventilator support and secretion clearance needs. Often nursing and respiratory therapy needs factor heavily into this decision.
- If considering discharging to home from the PICU, please see inpatient discharge criteria ([link to Inpatient Discharge Criteria](#))

ANTIMICROBIALS FOR RESPIRATORY TRACT INFECTIONS

- Patients with neuromuscular disorders are at an increased risk of respiratory tract infections, including pneumonia (PNA).
- Patients may present with community-acquired pneumonia (CAP), hospital-acquired pneumonia (HAP), or healthcare-associated pneumonia (HCAP), or aspiration pneumonia.
- See "[Uncomplicated Community Acquired Pneumonia](#)" Clinical Pathway for the inpatient management of CAP.
- Management of HAP, HCAP, and aspiration pneumonia includes antibiotic selection dependent on multiple factors including medical complexity, risk of aspiration, and risk factors for multi-drug resistant (MDR) pathogens.
 - Obtain diagnostic specimens (sputum culture, tracheal aspirate, bronchoalveolar lavage fluid) when able.
 - Considerations for broad spectrum antimicrobial coverage (including Methicillin-resistant *Staphylococcus aureus*, *Pseudomonas*, and MDR gram negatives):
 - Known history of resistant organisms in the last year
 - Current hospitalization of five days or more
 - Extended exposure to healthcare settings, including long term care facilities, hospitalizations in the last three months, chronic dialysis, home wound or infusion care
 - Exposure to broad spectrum antimicrobials in the preceding 90 days
 - Immunosuppressive disease and/or therapy

TABLE 1. SPECTRUM OF ACTIVITY OF ANTIMICROBIALS USED FOR PNEUMONIA

Antibiotic Choice	<i>Streptococcus pneumoniae</i> ^a	<i>Haemophilus influenzae</i> ^a	<i>Morexella catarrhalis</i> ^a	MSSA	MRSA ^b	Anaerobes ^c	<i>Pseudomonas aeruginosa</i> and/or Resistant Gram Negatives ^b
amoxicillin	+	+/- (72%)	-	-	-	-	-
amoxicillin-clavulanate	+	+	+	+	-	+	-
ampicillin	+	+/- (72%)	-	-	-	-	-
ampicillin-sulbactam	+	+	+	+	-	+	-
cefepime	+	+	+	+	-	-	+
ceftriaxone	+	+	+	+/-	-	-	-
ciprofloxacin ^e	-	+	+	- ^d	- ^d	-	+
clindamycin ^e	+ (86%)	-	-	+ (81%)	+ (77%)	+	-
levofloxacin ^e	+	+	+	+/- (93%) ^d	+/- (47%) ^d	-	+
linezolid ^e	+	-	-	+	+	-	-
metronidazole ^e	-	-	-	-	-	+	-
meropenem	+	+	+	+	-	+	+
piperacillin-tazobactam	+	+	+	+	-	+	+
sulfamethoxazole-trimethoprim ^e	+/- (72%)	+/-	+	+	+	-	-
vancomycin	+	-	-	+	+	-	-

^a *S. pneumoniae*, *H. influenzae*, and *M. catarrhalis* are all considered to be typical community-acquired pneumonia (CAP) pathogens. *S. pneumoniae* is the most commonly encountered bacterial pathogen in CAP and first line treatment in uncomplicated CAP at CHCO is ampicillin or amoxicillin (see Uncomplicated CAP CCG for details).

^b Considerations for broad spectrum antimicrobial coverage (including MRSA, *Pseudomonas*, MDR gram negatives): Known history of resistant organisms in the last year, current hospitalization of five days or more, extended exposure to healthcare settings (see text for details), exposure to broad spectrum antimicrobials in the preceding 90 days, immunosuppressive disease and/or therapy

^c Empiric coverage of anaerobic organisms may be considered if concerned for aspiration pneumonia; however this is controversial.

^d Fluoroquinolones, specifically levofloxacin, have moderate activity against *S. aureus* (both MSSA and MRSA), but resistance can develop while on therapy. Sensitivities are not automatically provided by the CHCO Microbiology Lab, but may be requested. Levofloxacin has better *S. aureus* coverage than ciprofloxacin.

^e Antibiotics with high oral bioavailability (IV=PO) and should be administered orally if clinically able.

TABLE 2. DOSING OF ANTIMICROBIALS USED FOR PNEUMONIA

Antibiotic Choice	Dose ^a
amoxicillin (PO)	<ul style="list-style-type: none"> 90mg/kg/day divided TID (max: 3,000mg/day)
amoxicillin-clavulanate (PO)	<ul style="list-style-type: none"> 90mg/kg/day divided TID (max: 3,000mg/day if using ES suspension and 2,625mg/day if using tablet)
ampicillin (IV)	<ul style="list-style-type: none"> 200mg/kg/day divided q6h (max: 8,000mg/day)
ampicillin-sulbactam (IV)	<ul style="list-style-type: none"> 200mg/kg/day (ampicillin component) divided q6h (max: 8,000mg/day)
cefepime (IV)	<ul style="list-style-type: none"> 100-150mg/kg/day divided q8h-12h (max: 2,000mg/dose and 6,000mg/day) Consider q8h dosing over q12h dosing if concerned about resistant gram negative or difficult to penetrate infections
ceftriaxone (IV)	<ul style="list-style-type: none"> 50mg/kg/day q24h (max: 2,000mg/day)
ciprofloxacin (IV or PO)	<ul style="list-style-type: none"> 30 mg/kg/day divided BID or q12h (max PO: 1,500mg/day, max IV: 800mg/day)
clindamycin (IV or PO)	<ul style="list-style-type: none"> 30-40mg/kg/day divided TID or q8h (max PO: 1,800mg/day, max IV: 2,700 mg/day)
levofloxacin (IV or PO)	<ul style="list-style-type: none"> Age greater than or equal to 6 months and less than 5 years: 20mg/kg/day divided BID or q12h (max: 750mg/day) Age greater than or equal to 5 years, adolescents, adults: 10mg/kg/day q24h (max: 750mg/day)
linezolid (IV or PO)	<ul style="list-style-type: none"> Infants and children less than or equal to 11 years: 30mg/kg/day divided TID or q8h (max: 1,800mg/day) Age greater than or equal to 12 years, adolescents, adults: 600mg q12h
metronidazole (IV or PO)	<ul style="list-style-type: none"> 30mg/kg/day divided TID or q8h (max:1,500mg/day)
meropenem (IV)	<ul style="list-style-type: none"> 60mg/kg/day divided q8h (max: 3,000mg/day)
piperacillin-tazobactam (IV)	<ul style="list-style-type: none"> Age 2 to 9 months: 240mg/kg/day (piperacillin component) divided q8h Age greater than 9 months, children, adolescents: 300mg/kg/day (piperacillin component) divided q8h (max: 12,000mg/day) Adults: 4,000mg q6h (max 16,000mg/day)
sulfamethoxazole-trimethoprim (IV or PO)	<ul style="list-style-type: none"> 10mg TMP/kg/day divided BID or q12h (max: 160-320mg TMP/dose) Not to be used in infants less than 2 months of age
vancomycin (IV)	<ul style="list-style-type: none"> Dosing variable for age, renal function, and past requirements Contact pharmacy for appropriate dose Therapeutic drug monitoring: goal trough = 10-20 mcg/mL (some sources suggest 15-20)

^a Note: Dosages based on normal renal and hepatic function, initial antibiotic therapy should be adjusted based on microbiologic data and clinical response to therapy

PRE-OPERATIVE EVALUATION

- Identify exact diagnosis (some children may have persistent CK elevation with no diagnosis)
- Most recent EKG/echocardiogram as patients may have primary arrhythmias such as long QTc, cardiomyopathy, or cardiac disease secondary to pulmonary hypertension, which may necessitate cardiac anesthesia.
- Most recent pulmonary function tests and home respiratory support
- Need for stress dose steroids secondary to chronic steroid use
- History of difficult airway secondary to limited jaw or cervical spine mobility
- Anesthetic precautions should be made as patients with neuromuscular disorders are at risk for malignant hyperthermia or rhabdomyolysis – these precautions vary by patient diagnosis
- All patients should receive hydration with lactate free isotonic fluids with dextrose
- Temperature and glucose should be closely monitored - infants and children with SMA are at particularly high risk of hypoglycemia and should receive D10 fluids if prolonged NPO status is expected.
- In general, succinylcholine and inhaled anesthetics except for NO₂ should be avoided
- Patients with known mitochondrial disorders should not receive large doses of propofol but can receive halogenated anesthetics
- Where possible, use of local (e.g. peripheral or neuraxial regional) anesthesia should be considered given risk of respiratory failure. Family should be prepared for possibility of prolonged intubation.

- Careful consideration should be made with family about risk of general anesthesia versus impact on ambulation of non-surgical intervention
- In recovery period, providers should be careful not to mask hypoventilation with supplemental oxygen.
- Notify ICU of need for patient bed following surgery

FOLLOW-UP

- Contact the Neuromuscular Coordinator or appropriate subspecialty clinic

PARENT | CAREGIVER EDUCATION

- MDA.org
- curesma.org
- curecmd.org

NEUROMUSCULAR ICD10 CODES

G71 Primary disorders of muscles

- G71.0 Muscular dystrophy
- G71.1 Myotonic disorders
- G71.11 Myotonic muscular dystrophy
- G71.12 Myotonia congenita
- G71.13 Myotonic chondrodystrophy
- G71.14 Drug induced myotonia
- G71.19 Other specified myotonic disorders
- G71.2 Congenital myopathies
- G71.3 Mitochondrial myopathy, not elsewhere classified
- G71.8 Other primary disorders of muscles
- G71.9 Primary disorder of muscle, unspecified

G11 Hereditary ataxia

- G11.0 Congenital nonprogressive ataxia
- G11.1 Early-onset cerebellar ataxia
- G11.2 Late-onset cerebellar ataxia
- G11.3 Cerebellar ataxia with defective DNA repair
- G11.4 Hereditary spastic paraplegia
- G11.8 Other hereditary ataxias
- G11.9 Hereditary ataxia, unspecified

G12 Spinal muscular atrophy and related syndromes

- G12.0 Infantile spinal muscular atrophy, type I [Werdnig-Hoffman]

- G12.1 Other inherited spinal muscular atrophy
- G12.2 Motor neuron disease
- G12.20 unspecified
- G12.21 Amyotrophic lateral sclerosis
- G12.22 Progressive bulbar palsy
- G12.29 Other motor neuron disease
- G12.8 Other spinal muscular atrophies and related syndromes
- G12.9 Spinal muscular atrophy, unspecified

G60 Hereditary and idiopathic neuropathy

- G60.0 Hereditary motor and sensory neuropathy
- G60.1 Refsum's disease
- G60.2 Neuropathy in association with hereditary ataxia
- G60.3 Idiopathic progressive neuropathy
- G60.8 Other hereditary and idiopathic neuropathies
- G60.9 Hereditary and idiopathic neuropathy, unspecified

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 Pharmacy & Therapeutics Committee – February 10, 2016

MANUAL/DEPARTMENT	Clinical Care Guidelines/Quality/Clinical Effectiveness
ORINATION DATE	May 10, 2016
LAST DATE OF REVIEW OR REVISION	May 10, 2016
APPROVED BY	 Lalit Bajaj, MD, MPH Medical Director, Clinical Effectiveness

REVIEW/REVISION SCHEDULE

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