Acute disturbance of motor function

EPNS Training Course
Neuromuscular Disorders
Budapest, 6-7 April 2017

Thomas Sejersen, Karolinska University Hospital
thomas.sejersen@ki.se

”Today´s menu”:

- Diagnosis av acute disturbance of motor function
- ”Urgent” conditions
- A few other acute conditions


Symptom
Diagnosis
Treatment

Diagnosis av acute disturbance of motor function:

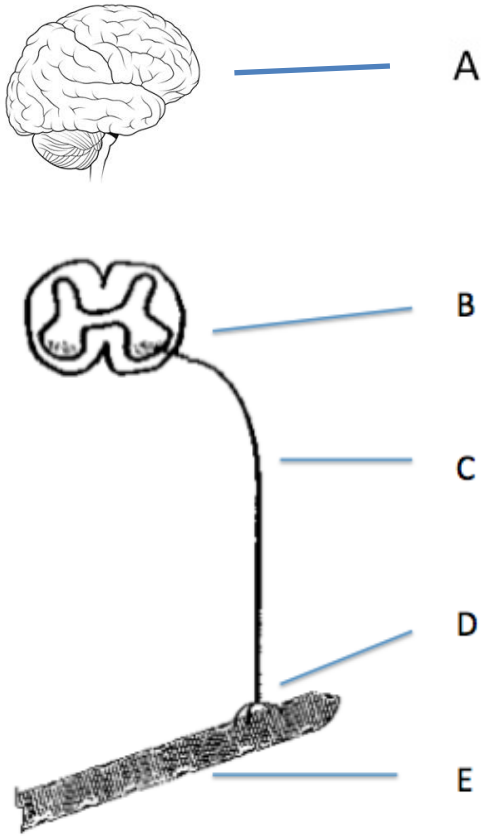
1. Type of abnormal motor function?
2. Anatomical level/extent?
3. Underlying cause?

1. Type of abnormal motor function?

- Muscle weakness/hypotonia
- Spasticity
- ”Movement disorder”
  - Ataxia
  - Dyskinesia
  - Dystonia
  - Atetosis
  - Myoclonus
  - Tics

2. Anatomical level/extent?

1. What muscle groups are engaged?
2. Level of lesion?



CNS (see Table 4.1)

Lower motor neuron (Table 4.2)

Axon (Table 4.3)

Neuromuscular junction (Table 4.4)

Muscle fiber (Table 4.5)

Physical exam findings each anatomic level:

Localization	Pattern of weakness	Sensory loss	Reflexes
Cerebral cortex, brainstem, or spinal cord	Distal > proximal, extensors > flexors, hemiparesis or single limb	May be present depending on whether sensory tracts or cortex are involved	Elevated however, reflexes may be decreased initially but later increase
Spinal cord	Distal > proximal, extensors > flexors, paraparesis, quadriparesis, rarely hemiparesis	May be present depending on whether sensory tracts are involved; loss of sensation below a certain spinal level is diagnostic	Elevated however, reflexes may be decreased initially but later increase
Anterior horn cell	Proximal and distal, fasciculations are prominent	Absent	Decreased if muscle bulk is severely decreased; increased in ALS
Peripheral nerve	In the distribution of the nerve, or diffusely present as stocking/glove weakness	Present	Decreased
Neuromuscular junction	First in eye muscles, neck extensors, pharynx, diaphragm, followed by more generalized weakness	Absent	Normal, decreased if muscle is paralyzed
Muscle	Proximal	Absent	Normal unless muscle severely weak

Neurocrit Care (2012) 17:S79–S95

3. Underlying cause?

Neuromuscular disorder (genetic)
Autoimmune cause
Infection
Toxic/metabolic cause
Trauma
Compression
Hypoxia
Vascular events
Epilepsy
Conversion syndrome
Myalgic encephalomyelitis

Systemic disorder
Fatigue
Pain
Catatonia

Supraspinal level

Infection/inflammation (meningitis, encephalitis, ADEM, MS) -> LP with CSF analysis, bacterial culture, virus isolation, brain MRI

Tetanus -> Clinical diagn, spatula test

Hypoxic/ischemic insult (near drowning, CO poisoning) -> Blood gas, EEG, neuroradiology, carboxyhemoglobin

Vascular events (stroke, TIA, vasculitis) ->Neuroradiology, coagulation panel, echocardiogram

Seizures (e.g. Todd´s paralysis) -> B-glu, electrolytes, EEG, neurorad, b-gas, toxic screen

Table 4.1 Acute disturbance of motor function at the supraspinal level

Condition	Typical symptom/finding	Initial diagnostic aid
Brain infection, postinfectious (meningitis, encephalitis, ADEM, multiple sclerosis)	Motor dysfunction typically accompanied by brain symptoms (See Chaps. 14 and 17). MS may initially present solely with motor disturbance (paralysis, ataxia, spasms)	LP with CSF analysis, bacterial culture, virus isolation, serology, brain MRI. See Chaps. 14 and 17
Toxic/metabolic	See Chap. 9	Blood gas. Toxicology screens. For acute neurometabolic crisis, see Chap. 9
Traumatic brain injury	Acute motor dysfunction typically accompanied by other CNS dysfunctions	Neuroradiology, ICP. See Chap. 12
Hypoxic/ischemic insult (near drowning, CO poisoning)	See Chap. 8	Blood gas, EEG, neuroradiology (MRI with spectroscopy). CO poisoning: carboxyhemoglobin. See Chap. 8
Tumor	Focal weakness/paralysis. May be accompanied by other brain symptoms. See Chap. 11	Neuroradiology. See also Chap. 11
Congenital brain malformation		
Chiari malformation: cerebellar tonsils displaced downward through foramen magnum	Headache, fatigue, muscle weakness in the head and face, difficulty swallowing, dizziness, nausea, impaired coordination, and, in severe cases, paralysis (of hands)	MRI of brain (and spinal cord)
Vascular events (stroke, transient ischemic attack, vasculitis) [6, 7, 16]	See Chap. 16	Neuroradiology, hypercoagulability panel, echocardiogram. See Chap. 16, Fig. 1
Seizures (e.g., Todd's paralysis, i.e., focal weakness in part of body after seizure)	Transient weakness in, e.g., hand, arm, or leg after (partial) seizure. May also affect speech, vision, or gaze. Usually subsides within 48 h	Blood glucose, electrolytes, EEG, neuroradiology, blood gas, toxic screens. See Chap. 2
Alternating hemiplegia [17, 18]	Diagnostic criteria: (1) onset before 18 months of age, (2) repeated episodes of hemiplegia, (3) episodes of bilateral hemiplegia/quadriplegia, (4) other paroxysmal attacks (tonic/dystonic attacks, nystagmus, strabismus, dyspnea), (5) immediate disappearance of symptoms upon sleep, (6) developmental delay, (7) not attributable to other known cause	Clinical diagnosis. brain MRI to exclude structural/vascular/metabolic Brain disorder. Metabolic screening to exclude mitochondrial disorders, CSF/blood glucose to exclude glucose transporter defects, thyroid panel to exclude periodic paralysis with thyrotoxicosis, video EEG to exclude epilepsy
Conversion disorder	Motor symptoms may present as, e.g., paralysis, impaired balance, gait problems, swallowing difficulties, dystonia, tremor, myoclonus, or other movement disorder	Clinical assessment, neurological findings inconsistent with symptom. See Chap. 7

Sejersen & Wang, Acute Pediatric Neurology, 2014, Springer

Spinal level:

Infection/inflammation (epidural abscess, osteomyelitis, transverse myelitis, neuromyelitis optica, poliomyelitis, West Nile virus)
->Spine MRI, LP (CSF analysis, bact culture, enterovirus PCR/immunidiagnostics, s-NMO IgG)

Trauma
->Spine X-ray and CT/MRI

Tumor
-> Spine CT/MRI

Table 4.2 Acute disturbance of motor function at the level of the spinal cord

Condition	Typical symptom/finding	Initial diagnostic aid
Infectious/postinfectious (epidural abscess, osteomyelitis, transverse myelitis [19], neuromyelitis optica [20], poliomyelitis, West Nile virus)	Progressive weakness in lower/all extremities, paresthesia, sensory loss, bladder/bowel incontinence, midline back pain, malaise. See Chaps. 14 and 17	Spine MRI, LP with CSF analysis, bacterial culture, enterovirus PCR/immunodiagnostics, s-NMO IgG elevated in neuromyelitis optica. See also Chaps. 14 and 17
Trauma	Acute weakness in lower/all extremeties, pareshesis, sensory loss, bladder/bowel incontinence, pain, malaise	Spine X-ray and CT or MRI
Tumor	See Chap. 12	Spine CT or MRI
Motor neuron disease (spinal muscular atrophy)	Generalized (symmetrical) muscle weakness with spared sensation. Fasciculations may occur (tongue). Decreased muscle tone and peripheral reflexes	Genetic testing smn1 gene (EMG)
Spinal cord malformation (dysraphism): spina bifida, diastematomyelia, spinal dermal sinus, spinal lipoma, tethered cord, syringomyelia	Chronic pain, abnormal sensation, paralysis, loss of urinary/bowel control, foot and spinal deformities. Signs of the disorder usually develop slowly, but sudden onset may occur	Spine CT or MRI
Foramen magnum stenosis (e.g., achondroplasia)	Apnea, paralysis (quadriplegia), muscle hypotonia	Brain and spine CT or MRI. Sleep study (polysomnography)

Sejersen & Wang, Acute Pediatric Neurology, 2014, Springer

Peripheral motor nerve:

Autoimmune (AIDP/Guillain-Barré**,** CIDP)
-> Neurophysiology (nerve conduction)

Trauma (brachial plexus injury, focal nerve compression)
->Neurophysiology (EMG, nerve conduction), MRI

Critical illness polyneuropathy
-> EMG, nerve conduction

Drug-induced neuropathy (e.g. Vincristine, cis-platinin, metronidazol, amiodarone)
-> EMG, nerve conduction

Table 4.3 Acute disturbance of motor function at the level of the peripheral motor nerve

Condition	Typical symptom/finding	Initial diagnostic aid
Autoimmune (AIDP, CIDP)	Frequently history of preceding infection. Initial abnormal sensation and motor function in feet/legs.	Neurophysiology (nerve conduction study): conduction slowing/block
Guillain–Barré syndrome [9–11]	Muscle weakness may progress to involve facial muscles and respiratory muscles resulting in respiratory failure. See Sect. 4.4.2.3	LP with CSF analysis: elevated CSF protein, normal/slightly elevated lymphocytes
Hereditary neuropathies	Distal symmetric muscle weakness with/without abnormal sensation. Rarely acute presentation. Pes cavus, family history common	Nerve conduction study of motor and sensory nerves demonstrates reduced nerve conduction speed (myelin damage) or reduced nerve conduction strength (axonal damage). Genetic testing
Facial palsy (congenital 8 %, Bell's palsy 42 %, infection (neuroborreliosis—Lyme disease) 13 %, trauma 21 %, leukemia/ tumor 2 %, hypertension, otitis media, mastoiditis)	Dysfunction of cranial nerve VII (facial nerve) resulting in paresis of muscles on affected side. Lyme disease frequent cause in areas endemic for Borrelia	Cranial nerve examination, eye closure (secure corneal lubrication), blood pressure, Borrelia serology spinal fluid + serum, serology herpes zoster, EBV, CMV, blood count, otoscopy, oral and parotid exam
Toxic/metabolic (chemotherapy, deficiency vitamin B12)	Initially paresthesia and sensory loss in a stocking distribution with later appearance of muscle weakness	Neurophysiology (nerve conduction study), vit B12 in blood, indirect markers homocysteine, methylmalonic acid, and holotranscobalamin
Trauma		
Brachial plexus injury	Obstetric brachial plexus palsy occurs in less than 1 % of live births and may result in pain, loss of sensation, or paralysis/weakness	Neurophysiology (EMG, nerve conduction study), MRI
Focal nerve compression (e.g., following awkward positioning in deep sleep)	Flaccid paralysis in muscle(s) supplied by affected nerve (e.g., radial nerve, common ulnar nerve). Sensory loss, decreased reflexes	Nerve conduction study
Critical illness polyneuropathy [21–23]	CIP is a frequent complication of critical illness, often together with CIM. It presents as flaccid weakness, usually symmetrical and sometimes severe, prolonged weaning from mechanical ventilation, muscle wasting	EMG, nerve conduction study

Sejersen & Wang, Acute Pediatric Neurology, 2014, Springer

Neuromuscular junction:

Infection: infant botulism
->Botulinum toxin in stool

Autoimmune: Myasthenia gravis
-> Ach rec a.b., MuSK a.b., LRP4 a.b.,rep nerve stim, sf-EMG

Snake venom (neurotoxins mostly in elapid snake species)
-> History, clin exam. BP, neurol assessm., ECG

Table 4.4 Acute disturbance of motor function at the level of the neuromuscular junction

Condition	Typical symptom/finding	Initial diagnostic aid
Infection: infant botulism [13]	Symptoms typically start 18–36 h after toxin ingestion. Constipation, muscle weakness, drooping eyelids, ophthalmoplegia, swallowing difficulty, drooling	Botulinum toxin in stool
	Hypotonia and weak reflexes	EMG with repetitive nerve stimulation (decrement), single-fiber EMG
Autoimmune: myasthenia gravis (MG) [12]	Transient neonatal MG: generalized muscle weakness, hypotonia, poor suck, respiratory difficulty	Anti-ACh receptor antibodies, anti-MuSK antibodies, EMG with repetitive nerve stimulation (decrement), single-fiber EMG
Hereditary congenital myasthenia	Generalized weakness, hypotonia, drooping eyelids, ophthalmoplegia, and delays in motor skills (crawling, sitting, and walking). Babies may have poor head control and difficulty feeding	EMG with repetitive nerve stimulation (decrement), single-fiber EMG, genetic testing
Snake venom (neurotoxins, mostly found in elapid snake species)	Presynaptic neurotoxins (e.g., Elapids, Viperids): progressive paralysis with onset >1 h after bite, postsynaptic neurotoxins (many elapids, e.g., cobra); flaccid paralysis reversible with antivenom therapy	History and clinical examination. Blood pressure, neurological assessment. ECG if general condition affected
	Dendrotoxins and fasciculins (e.g., mamba, rattlesnakes): spasms, fasciculations, tetany often in <1 h from bite	Hemoglobin, complete blood count, serum creatine kinase, blood gas, coagulation status (INT, aPTT, D-dimer), s-creatinine. Repeated tests over 24 h

Sejersen & Wang, Acute Pediatric Neurology, 2014, Springer

Skeleletal muscle:

Virus infection (coxsackie, infl, parainfl, EBV, adenovirus, Dengue fever, Lassa fever
->Serology, virus isolation, PCR

Brucellosis (zoonisis)
-> Brucella a.b., culture, PCR

Ricketsia (e.g. typhus, Q-fever, Mediterranean spotted fever)
->Typical clin presentation, PCR

Skeleletal muscle:

Malignant hyperthermia

Hereditary NMDs

Intensive care myopathy

Hypokalemic periodic paralysis
-> s-K, Exercise EMG, mutation *SCN4A, CACNA1S*

Hyperkalemic periodic paralysis
-> s-K/electrolytes, ECG, mutation *SCN4A*

Table 4.5 Acute disturbance of motor function at the level of the skeletal muscle

Condition	Typical symptom/finding	Initial diagnostic aid
Virus infection (coxsackie, influenza [24], parainfluenza, EBV, adenovirus, Dengue fever, Lassa fever)	Myalgia, muscle weakness. General malaise, fever, headache. Catarrhal symptoms	Serology tests, virus isolation, PCR
Brucellosis (zoonosis)	Brucellosis: history of exposure to infected animals or food. Fatigue, undulating fever, weakness, excessive sweating, myalgia, abdominal pain, arthralgia	Brucella antibodies/culture/PCR
Rickettsia (typhus, Q fever, Rocky Mountain spotted fever, Mediterranean spotted fever, African tick-bite fever)	Transmitted by mites. Skin lesions/rash, fever, headache, myalgia	Typical clinical presentation, immunoassays, PCR
Trichinosis (roundworm Trichinella spiralis)	Intake of undercooked pork, fish, or wild game. Diarrhea, facial edema, splinter hemorrhage under nails, myalgia	Exposure history, typical clinical presentation, lab: blood count (eosinophilia, serum creatine kinase elevation, immunoassays)
Tetanus (neurotoxin tetanospasmin from Clostridium tetani)	Insufficient protection tetanus vaccination? Muscle spasms of jaw, trismus, frequent 1st symptom. Swallowing difficulty, myalgia, and stiffness in neck, shoulders, back. Progressive spasms and convulsions potentially life threatening	Clinical diagnosis, spatula test (involuntary spasm of jaw upon touching posterior pharyngeal wall)
Dermatomyositis/polymyositis [25]	Symmetrical weakness, fatigue, malaise, weight loss, mild fever, myalgia, pain in chest/abdomen, palpitations. In dermatomyositis violet/dusky red rash most easily detected in face, eyelids, around nails	Serum creatine kinase elevated, autoantibodies, muscle biopsy, muscle MRI
Metabolic (hypothyroidism, vitamin D deficiency)	Vitamin D deficiency has been associated with muscle weakness and pain in both adults and children	25-OH-vitamin D in blood
	Hypothyroidism may present as constipation, muscle weakness, hypotonia, poor growth, poor mental development, delayed development	Thyroid panel

Sejersen & Wang, Acute Pediatric Neurology, 2014, Springer

Table 4.5 (continued)

Condition	Typical symptom/finding	Initial diagnostic aid
Hereditary muscle diseases: myopathies and muscular dystrophies [26, 27]	Delayed motor milestones. Muscle weakness with decreased distal reflexes. Contractures. Rarely presents acutely. See Chaps. 10 and 15	Serum creatine kinase (CK) elevated in muscular dystrophies (commonly >10×normal value), p-lactate (frequently elevated in mitochondrial myopathy, repeated tests recommended), genetic testing, neurophysiology (EMG), muscle biopsy. See Chaps. 10 and 15
Malignant hyperthermia [14, 15]	Masseter contraction early warning sign. Other early signs: unexplained tachycardia, elevated end-tidal CO2 concentration, and muscle rigidity. Hyperthermia is late sign. Urgent condition, see Sect. 4.4.2.5	Typical clinical presentation. Raised CK, myoglobin, K, and P levels in blood. Metabolic/respiratory acidosis
Hypokalemic periodic paralysis	Hereditary autosomal dominant hypokalemic periodic paralysis: family history common. Attacks of weakness, lasting hours to days, provoked by, e.g., exercise, carbohydrate-rich meal, or sudden temperature changes	s-K low during attack of paralysis. CMAP (exercise EMG) Hereditary hypokalemic periodic paralysis: Mutation analysis of *SCN4A* and *CACNA1S* genes
	Acquired hypokalemic paralysis may be caused by frequent diarrheas	s-K, s-electrolytes, blood gas, ECG
Hyperkalemic periodic paralysis	Hereditary autosomal dominant hyperkalemic periodic paralysis: family history common. Exacerbate by K intake or cold. Weakness and myotonia	Serum potassium, electrolytes, blood gas, ECG. Mutation analysis of *SCN4A*
Critical illness myopathy (CIM) [21–23]	CIM is a frequent complication of critical illness, often together with CIP. It presents as flaccid weakness, usually symmetrical and sometimes severe; prolonged weaning from mechanical ventilation; muscle wasting	EMG, nerve conduction study, muscle biopsy (loss of myosin)
Drug-induced myopathy [28]: Corticosteroid-induced myopathy Colchicine, chloroquine, lipid-lowering drugs (e.g., statins), diuretics, D-penicillamine (myasthenic syndrome)	Symmetrical weakness, often severe. Myalgia and muscle atrophy	Muscle strength reduced, often markedly. Tenderness on palpation

Sejersen & Wang, Acute Pediatric Neurology, 2014, Springer

Other musculoskeletal causes:

Table 4.6 Acute disturbance of motor function, other musculoskeletal causes

Condition	Typical symptom/finding	Initial diagnostic aid
Infection/postinfectious		
Septic joints Autoimmune arthritis Osteomyelitis	Local pain, malaise, weakness in muscles in affected area	Ultrasound/X-ray/MRI/scint, arthrocentesis, bacterial culture, CRP, SR, blood count, autoantibody tests
Trauma/sports injury		
Fracture	History of trauma. Pain, swelling, obvious deformity	X-ray, ultrasound, MRI, clinical exam
Elbow dislocation	Common dislocation in children under age 5 years (Nursemaid's elbow) pain, reduced movement of affected arm	Clinical examination (inability to rotate arm and flexing elbow fully)
Compartment syndrome	Abnormal sensation, (severe) pain, swelling, reduced strength, pallor Muscle compartment tense on palpation	Compartment syndrome: serum creatine kinase, urine-myoglobin, compartment pressure measurement
Deep vein thrombosis	Commonly in leg. Pain, swelling, tenderness. Red, warm skin	Ultrasound, venography, MRI
Snake venom (hemotoxic procoagulant or myotoxic venom)	Hemotoxins (e.g., most vipers) cause hemolysis and disrupted blood clotting. Commonly severe pain in wound site, followed by swelling and discoloration. General symptoms with vomiting, dyspnea, dizziness, affected circulation	Hemotoxins: D-dimer elevated, fibrinogen very low, prolonged prothrombin time, APTT, and clotting time. Assess renal function
	Myotoxins (e.g., rattlesnakes): rhabdomyolysis with muscle pain, tenderness, and paralysis	Myotoxins: serum creatine kinase, s-myoglobin, and u-myoglobin highly elevated. Assess renal function
Tumor		
Osteosarcoma, Rhabdomyosarcoma	Palpable tumor mass, pain, abnormal sensitivity, and muscle weakness if nerve compression. Osteosarcomas may cause bone pain, limping, and fracture	X-ray, MRI, CT scan, biopsy

Sejersen & Wang, Acute Pediatric Neurology, 2014, Springer

Muscle weakness, ”urgent conditions”:

1. Stroke/Cerebrovascular insult
2. Spinal cord compression
3. AIDP / Guillain-Barré syndrome
4. Myasthenic crisis and botulism
5. Malignant hyperthermia

AIDP/Guillain-Barrés syndrom

Acute inflammatory demyelinating polyneuropathy (AIDP)

Incidense: 0.5-1/100 000/yr

Symptom:

Preceding infection common. Pareasthesias in feet and hands followed by weakness (distal ->proximal). Pain may occur.

CAVE potential respiratory failure and dysautonomia!

Miller Fischer syndrome: svaghet proximalt -> distalt, ataxia, ophtalmoplegia

Diagnosis AIDP/Guillain-Barré:

Liquor: Protein elevated, with or w.o. pleocytosis (10-50/ml, lymphocyte predominance). Max appr day 10!

***Serology*:** Anti-ganglioside a.b. occur

***Neurophys*:** ENeG –slowed conduction velocities, F-response latency/lack of F-wave (sign demyelinating process).

Neuroradiology *(MR with contrast)*: Enhancement of nerve roots.

Treatment AIDP/Guillain-Barré:

IVIG:
0.4 mg/kg/d for 5 days, alternatively
2 g/kg over2-4 days.

Plasmapheresis

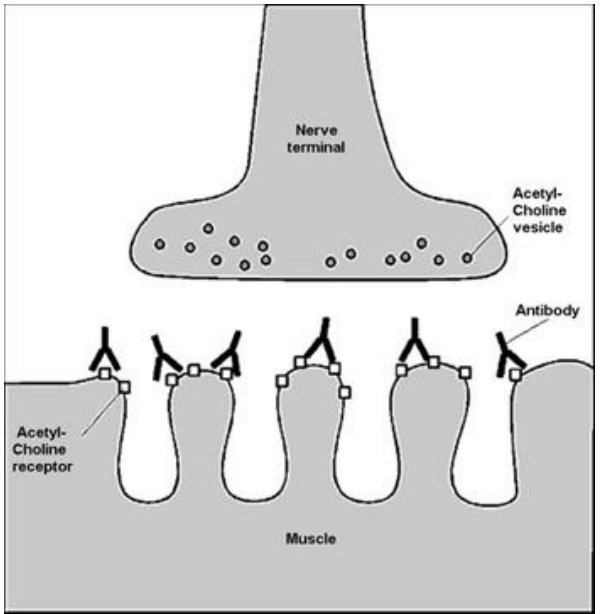
Profylaxis DVT: Low-molecular heparin
Profylaxis stress caused gastritis: H2-antagosist
GI funtion: Lactulosis, enema

Myasthenic crisis:

Myastenia gravis: Autoimmune defect of neuromuscular junction (Ach rec)

Myasthenic crisis: Acute worsening of MG. Provoked by e.g. fever, infection, aspiration, medicines (high dose cortisone, aminoglycosides, erythromycin, clindamycin)

Cholinergic crisis: Acute worsening from overdose of Cholinesterase inhibitors



Myasthenic crisis:

Symptoms: Ptosis, diplopia, general uscle weakness – fatigability, dysphonia, dysphagia

RESPIRATORY FAILURE!

Diagnosis: Neurophysiology (rep nerve stim, single fiber-EMG, Anti-AChR, Anti-MuSK). Clinical tests endurance

Edrophonium /”Tensilon” test (film response of chose muscle groups)

Myasthenic crisis:

Treatment: Cholinesterase inhibitor (Pyridostigmin, Infants: 0.5-1 mg/kg/dose each 4-6 h, older children 7 mg/kg/d distributed on 3-6 doses, adults 60 -180 mg x 3-6.

0.1 mg/kg per dose if i.v. route)

Surveillance respiratory function! Readiness for ventilator care!

Possibly IVIG, cortisone, plasmaferesis

Acute infantile botulism:

Symptoms: Remind of severe MG**.** NOTE! Often preceded by constipation. Ptosis, diplopia, general muscle weakness – fatigability, dysphonia, dysphagia

RESPIRATORY FAILURE!

Diagnosis: Neurophysiology (rep nerve stim, single fiber-EMG). Detection Cl difficile in faeeces, botulinum toxin in serum.

Treatment: Supportive care, surveillance respiration and readiness ventilator care. Botulinum IG.

Malignant hyperthermia, symptoms:

Specific

- Muscle rigidity*
- Increased CO_2 production
- Marked temperature elevation
- Rhabdomyolysis

Non-Specific

- Tachycardia
- Tachypnea
- Acidosis (respiratory; metabolic)
- Hyperkalemia

Malignant hyperthermia, diagnosis:

Muscle Contracture Test

- Caffeine Halothane Contracture Test(CHCT)
- Gold Standard
- MH Muscle Biopsy Center

Genetic Testing

- Ryanodine receptor (RYR1 Gene)
- Primary genetic focus

Malignant hyperthermia, triggers:

MH Triggers

- Potent volatile anesthetics
  - Sevoflurane
  - Desflurane
  - Isoflurane
- Depolarizing muscle relaxants
  - Succinylcholine

NOT MH Triggers

- Nitrous oxide
- IV induction agents
- Non-depolarizing muscle relaxants
- Opioids

Malignant hyperthermia, physiology:

- Inherited disorder of skeletal muscle
- Problem w/ reuptake of intracellular Ca^{2+}
- Exact cause uncertain
  - Ryanodine receptor / central core disease
- Disease inheritance is autosomal dominant
- MH-like reactions in other myopathies / MDs

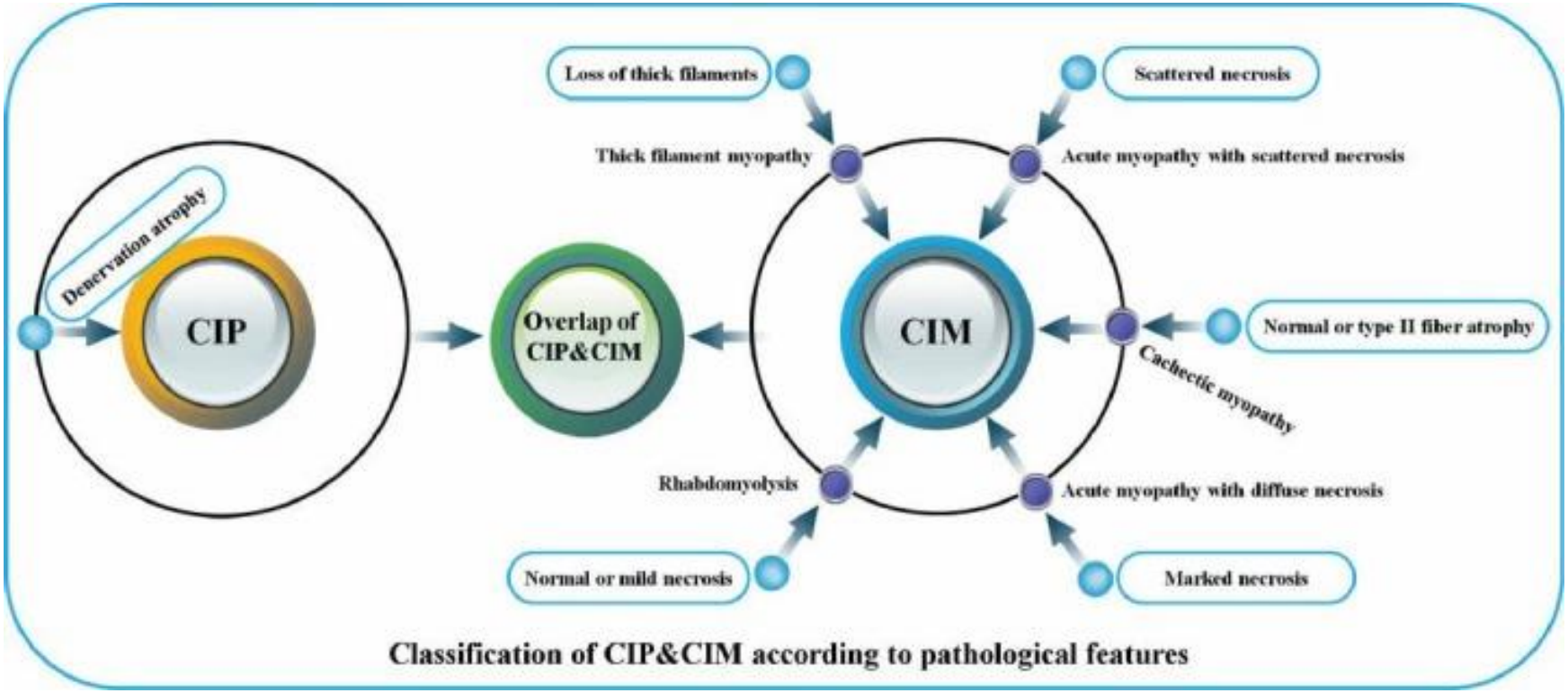
Malignant hyperthermia, treatment:

- Stop triggering inhalation agents/succinylcholine
- Hyperventilate high flow 100% O_2
- Dantrolene 2.5mg/kg push, repeat prn
- Continue monitoring & interventions
- Treat hyperthermia, acidosis, and arrhythmias

A few words on:

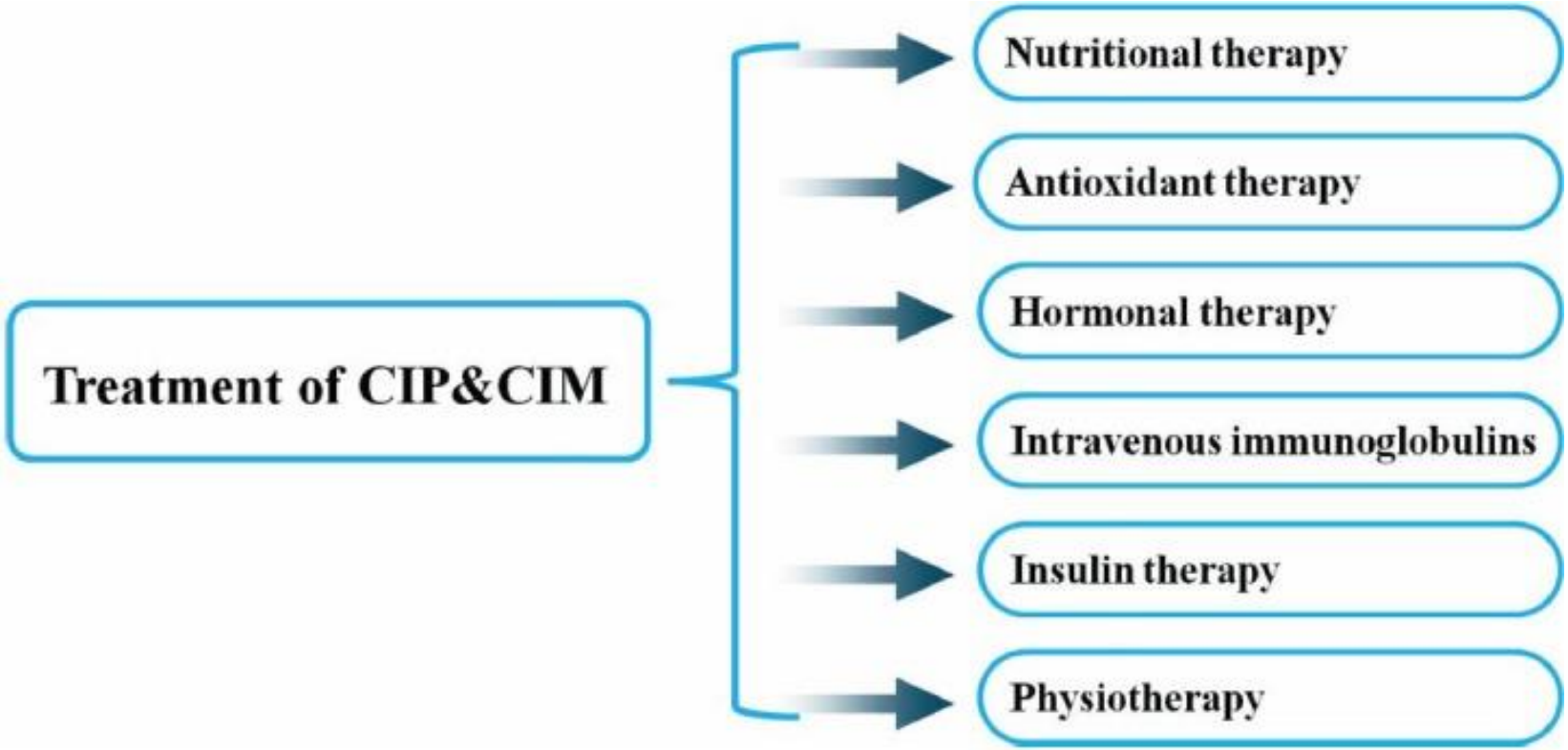
1. Intensive care neuropathy/myopathy
2. ”Floppy infant”
3. Status dystonicus

Intensive care neuropathy/myopathy



Neural Regen Res. 2014 Jan 1; 9(1): 101–110.

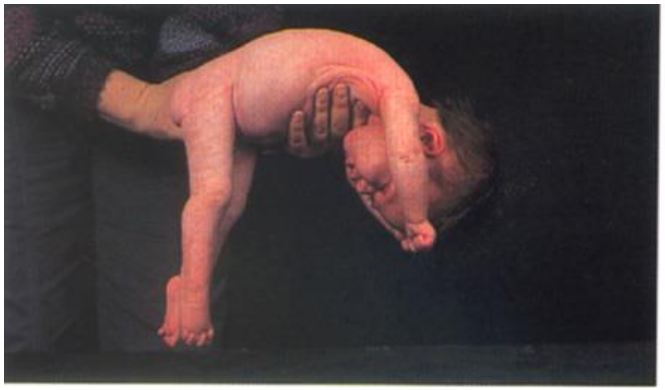
Intensive care neuropathy/myopathy



Neural Regen Res. 2014 Jan 1; 9(1): 101–110.

”Floppy infant syndrome”:

A condition of marked muscle relaxation in a baby so that when supported face down the baby droops over the hand like an inverted "U"



Central or peripheral cause?

Central cause

- Reduced conscience
- (w.o. weakness)
- Peripheral reflexes often increased
- Primitive reflexes decreased

Peripheral cause

- Normal conscience
- Weakness
- Absence of peripheral reflexes

Anatomical level - cause:

Table 1 Localization in the Floppy Infant

Origin of Hypotonia	Structural Localization	Clinical Pathological Conditions
Supraspinal/suprasegmental hypotonia (preserved DTR)	Brain	Systemic illness (sepsis, CHF, HIE)
	Brainstem	Syndromic hypotonia
		Cerebral dysgenesis
		Grossly normal brain
	Craniovertebral junction	Spinal cord injury
Segmental or motor unit hypotonia (DTR depressed or lost)	Anterior horn cell	Spinal muscular atrophy
	Peripheral nerve	HMSN
	Neuromuscular junction	Myasthenia gravis, congenital myasthenic syndromes, botulism
	Muscle	Congenital myopathies, metabolic myopathies, neonatal presentation of muscular dystrophy

DTR = myotatic reflexes (deep tendon reflexes); CHF = congestive heart failure; HIE = hypoxic ischemic encephalopathy; HMSN = hereditary motor sensory neuropathy.

Bodensteiner, Semin Pediatr Neurol 15:10-20, 2008

Investigations central cause:

- B-glucose, electrolytes, blood gas, Blood count, s-bilirubin, screen for sepsis incl LP
- EEG/aEEG
- Ultrasound/CT/MR CNS
- Metabolic workup, TORCH
- Karyotype/Array CGH if dysmorphic traits
- Genetic analysis Prader-Willi syndrome or other suspected hereditary disorder/syndrome

Investigations peripheral cause:

- s-CK (-> genetic analysis cong mu.dys)
- EMG, EnEG, repetitive nerve stim.
- muscle biopsy (-> genetic analysis cong myopathy, mitochondrial disease, metabolic myopathy)
- specific genetic tests: e.g. congenital DM1, SMA (deletion *SMN)*
- Short acting cholinesterase inhibitors (Edrophonium/Tensilon test) and/or attempt pyridostigmin therapy

Status dystonicus

‘Increasingly frequent and severe episodes of generalized dystonia (sustained involuntary muscle contraction leading to abnormal postures and movement) which requires urgent hospital admission’

Acute cause (e.g. near-drowning, asphyxia, encephalopathy)

or secondary to known dystonia

Triggering factors: Infection, stress, trauma, surgery, fever, onset/withdrawal of drug (e.g. Li, clonazepam).

Status dystonicus

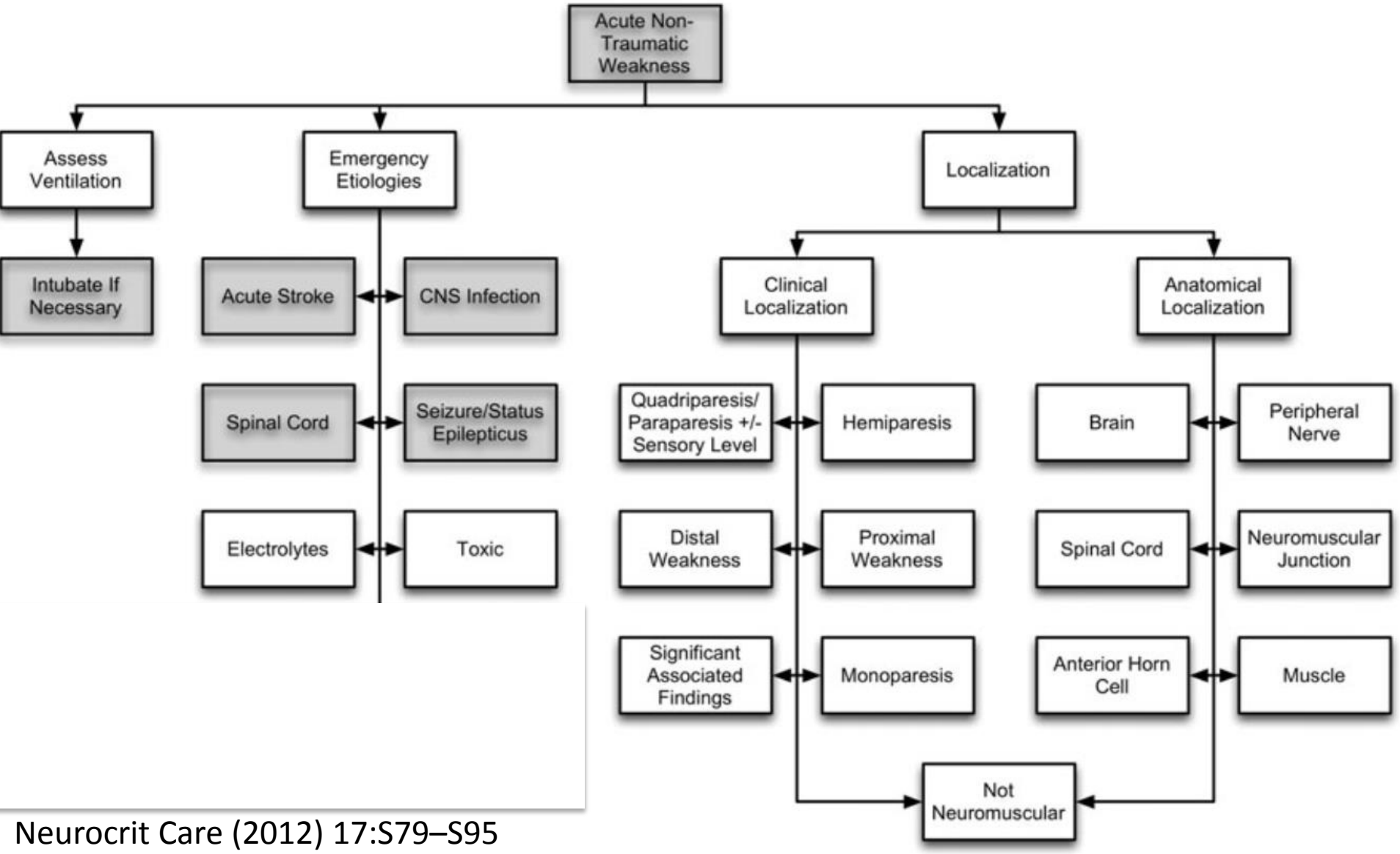
Treatment/management: Fluid and electrolyte balance, control vital parameters, analgetics.

ATT.! risk rhabdomyolysis -> myoglobinuria

->kidney failure!

Indication deep sedation (midazolam, propofol) with repirator?

ENLS: Initial management of acute weakness



Neurocrit Care (2012) 17:S79–S95


Thank you!