

THE
2ND DECADE



MYOTONIC
DYSTROPHY
FOUNDATION



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MYOTONIC
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Care and a Cure

BEYOND GENERAL ANESTHESIA: OPIATES AND PAIN MEDICATIONS



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Declarations

- This presentation will include:
 - ▣ off-label use of medications
 - ▣ commercial names of medications
- The author:
 - ▣ has no conflicts of interest with any of manufacturer medications discussed
 - ▣ is a co-investigator on clinical trials of neuromuscular disorders, including myotonic dystrophy type 1

Goals of anesthesia

- To make a procedure or surgery tolerable, by managing:
 - ▣ Consciousness
 - ▣ muscle tone
 - ▣ Pain
- And to monitor health status:
 - ▣ Cardiac
 - ▣ Respiratory
 - ▣ Metabolic
- These are all taken into consideration in the myotonic patient
 - ▣ Prolonged monitoring

Myotonic Dystrophies:

Multi-system vulnerability to medication side effects

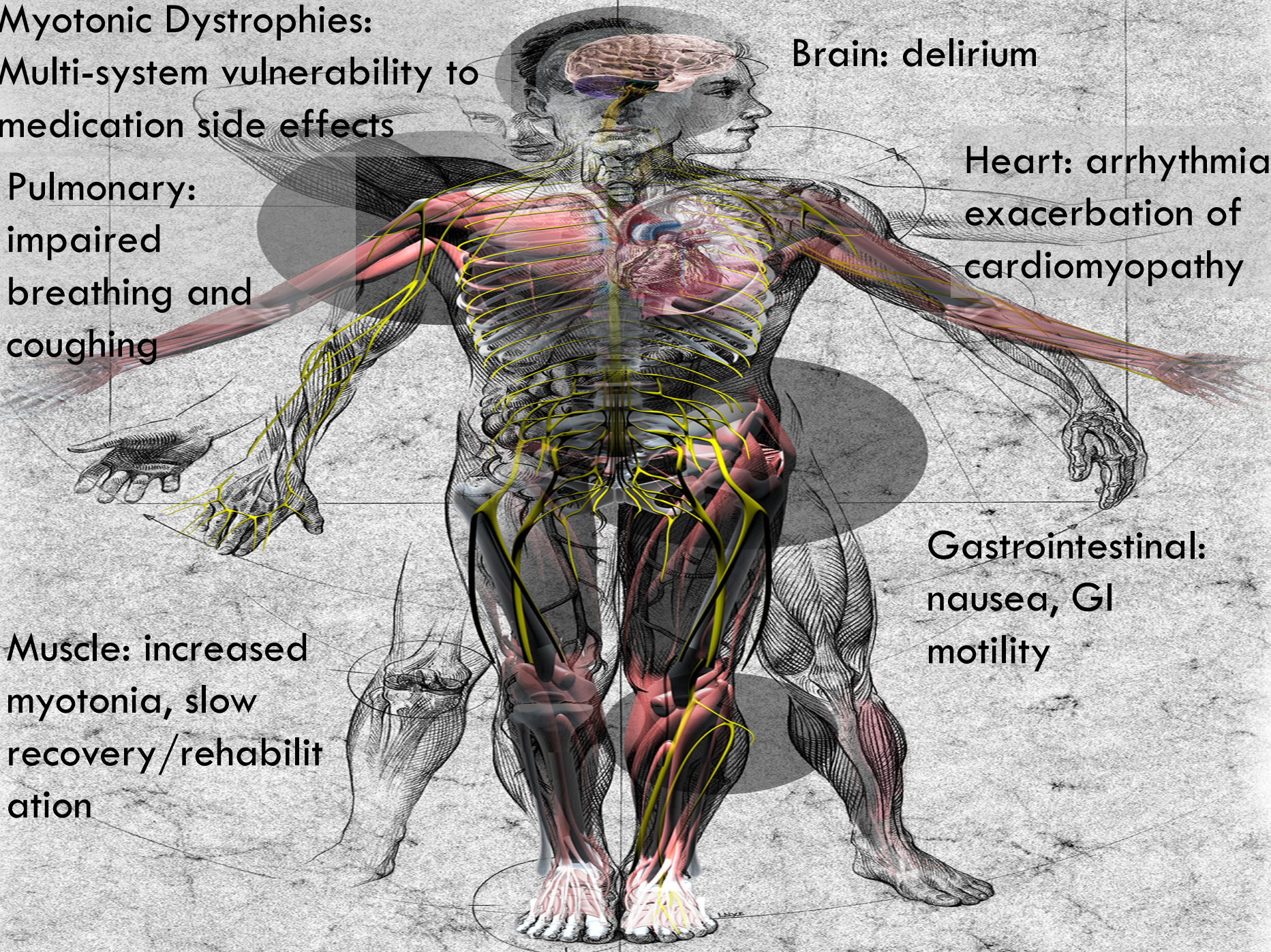
Pulmonary: impaired breathing and coughing

Muscle: increased myotonia, slow recovery/rehabilitation

Brain: delirium

Heart: arrhythmia, exacerbation of cardiomyopathy

Gastrointestinal: nausea, GI motility



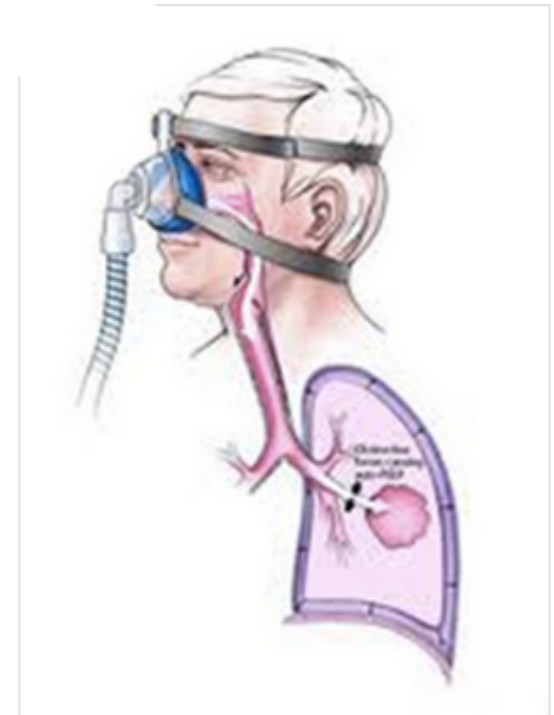
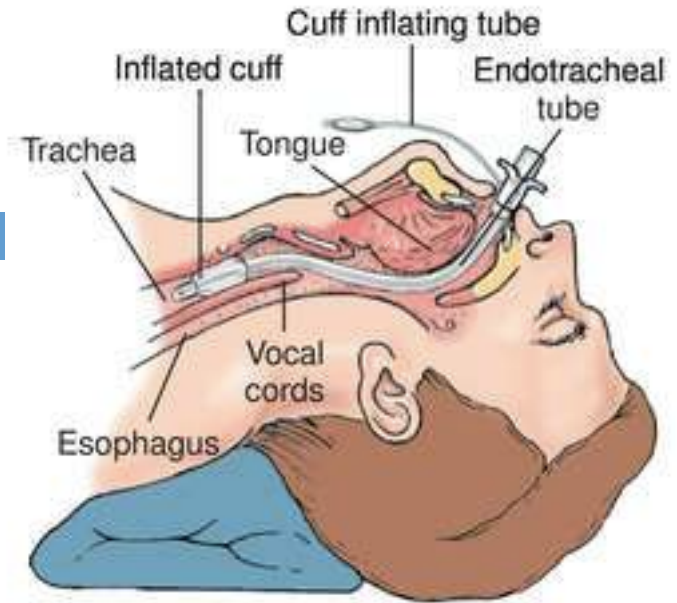
Cardiac concerns

- Preoperative
 - If you haven't had one recently, your doctor may want:
 - EKG or Holter (longer duration) EKG monitoring
 - echocardiogram
- Intraoperative monitoring
 - Cardiac monitoring
 - Blood pressure monitoring



Pulmonary concerns

- Pre-operative
 - Condition of your teeth, jaw, airway
 - Pulmonary function testing
- Intra-operative
 - Monitor oxygen (O_2), carbon dioxide (CO_2)
 - Manage ventilation
 - Intubation
 - Non-invasive ventilation



Pulmonary concerns

- Post-operative
 - Monitor for hypoventilation
 - (shallow breathing)
 - Resume or use non-invasive ventilation
 - Prevent aspiration (saliva, food, or fluids going into lungs)
 - Can cause pneumonia, resulting in re-intubation, prolonged hospitalization
 - Use incentive spirometry, cough assist



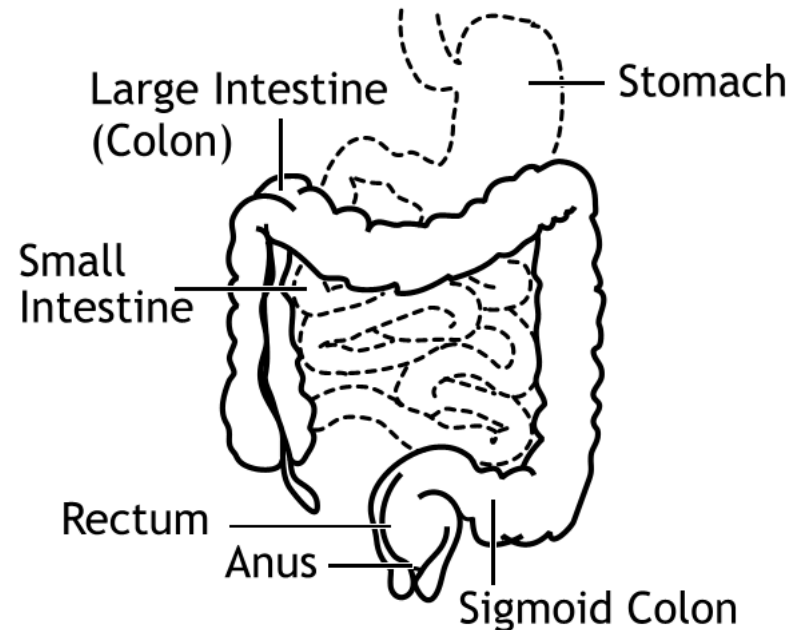
Incentive spirometry



Respironics, Inc.

Gastrointestinal concerns

- Decreased gut motility leads to:
 - Nausea
 - Vomiting (and risk of aspiration)
 - Delayed gastric emptying
 - Feeling full quickly
 - decreased appetite
 - Constipation
 - pseudo-obstruction or Ilius
 - Symptoms same as a bowel blockage
 - Unable to have a bowel movement or pass gas

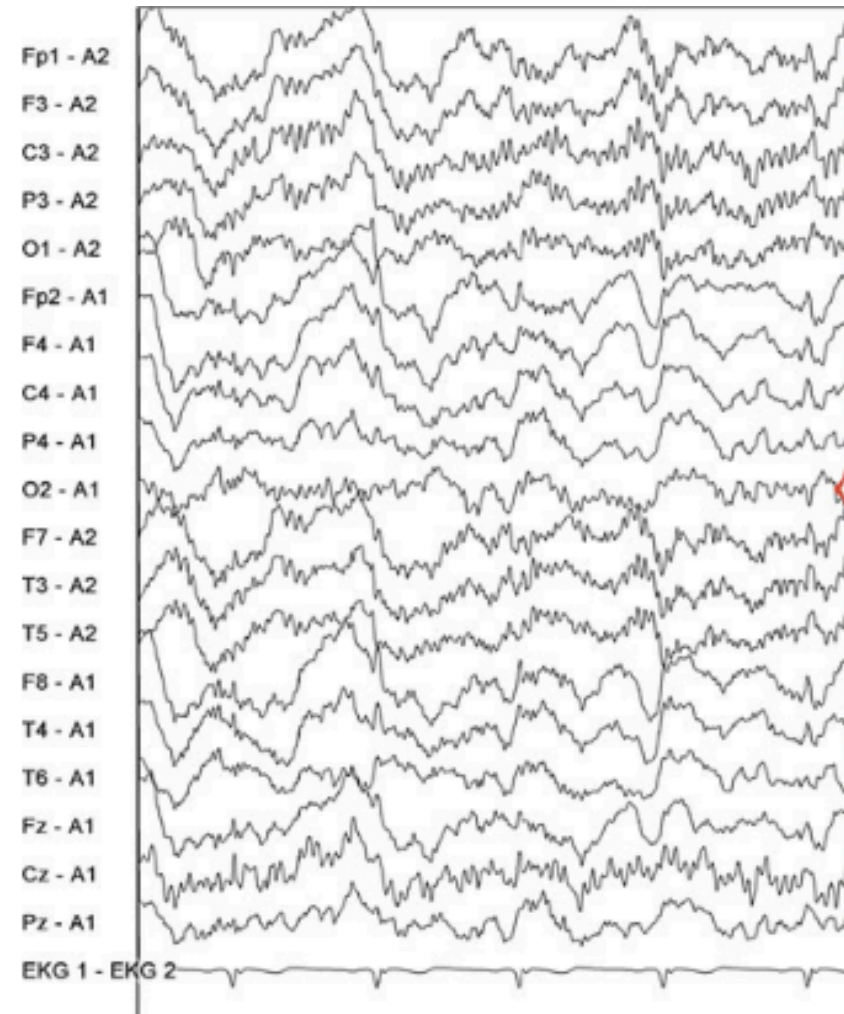


Muscle concerns

- Intra-operative
 - Myotonia exacerbation
 - Potassium imbalance
 - Cautery
 - Electrical stimulation
 - Shivering/low temperature
- Post-operative
 - Prolonged bedrest can lead to muscle atrophy
 - Physical therapy/rehabilitation

Brain concerns

- Delirium, also called:
 - Confusion
 - Encephalopathy
- Causes include:
 - Medication side effect
 - Infection
 - Metabolic imbalance
- Increase susceptibility if:
- Cognitive involvement

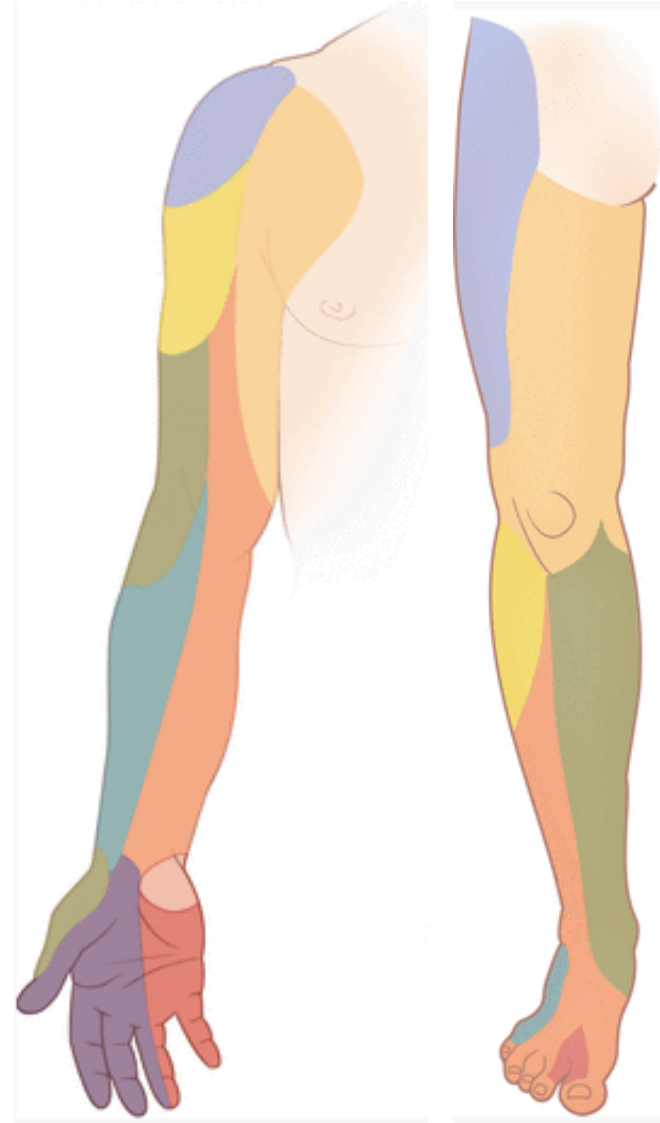


General anesthesia

- Inhaled agents:
 - halothane
 - flurothane
- Intravenous agents:
 - propofol
 - ketamine
- Muscle relaxants:
 - Succinylcholine (depolarizing)
 - Etomidate (non-depolarizing)

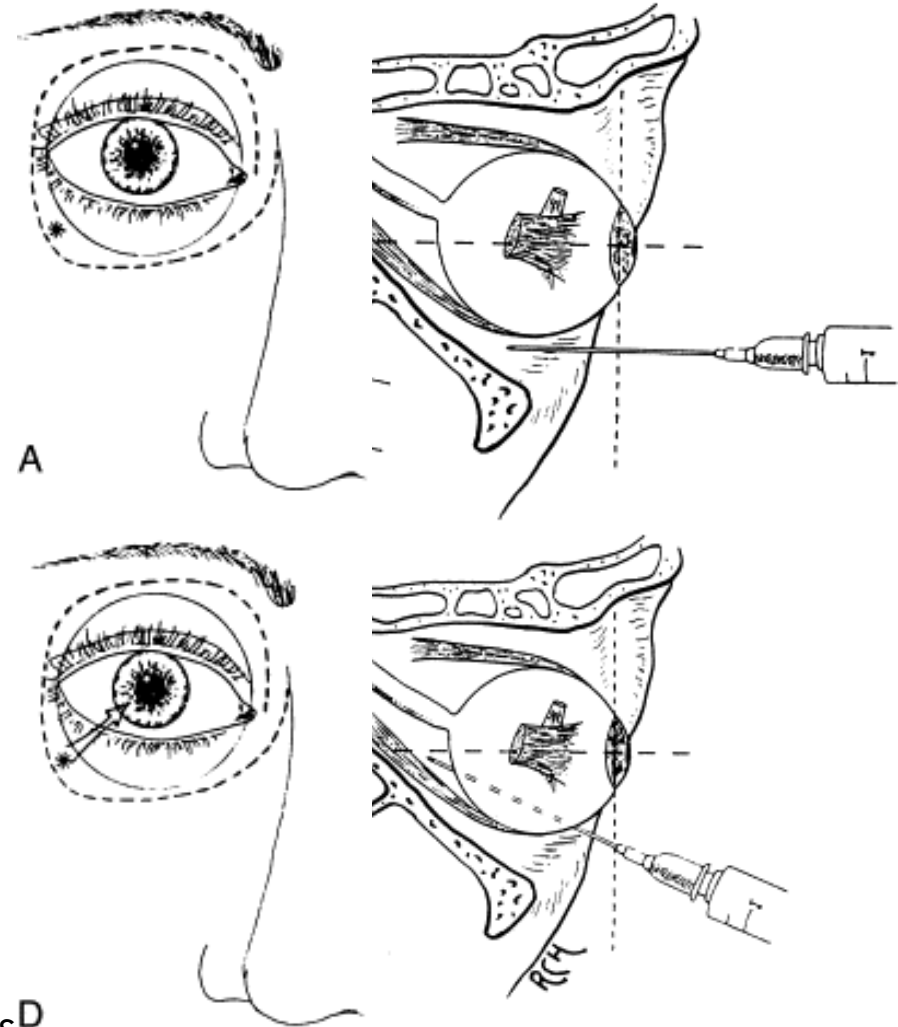
Local or regional anesthesia

- Nerve block
 - ▣ Intra-operative
 - ▣ Post-operative



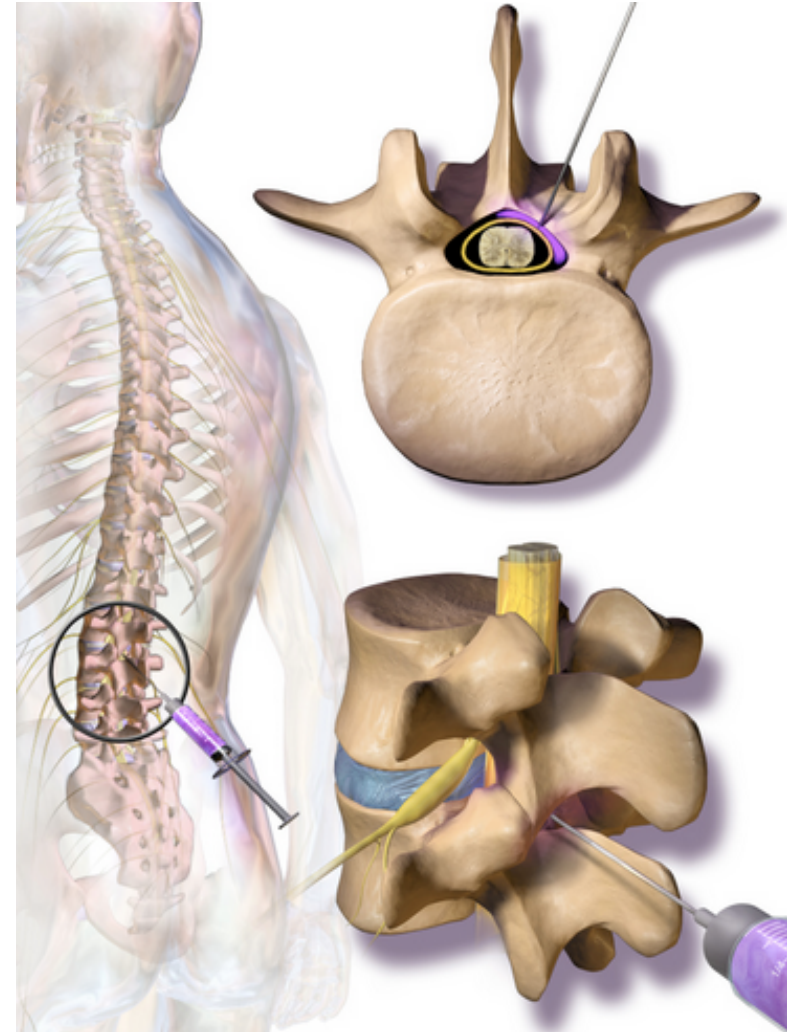
Local or regional anesthesia

- Cataract surgery
 - ▣ Anesthesia of orbit (eye and eye socket)



Spinal or epidural anesthesia

- Spinal
 - ▣ Into spinal fluid
- Epidural
 - ▣ Around nerve roots
- childbirth
- gynecologic surgery
- lower extremities



How do we feel pain?

signals

- Inflammatory molecules
 - ▣ Arachidonic acid
 - ▣ Cyclooxygenase
 - ▣ cytokines
- Receptors
 - ▣ Volt-gated sodium channels
 - ▣ TRPV and TRPA channels
 - ▣ Mu-receptors

circuits

- Peripheral nerve
 - ▣ Un- and thinly myelinated small nerves
- Spinal cord ↔ brain
 - ▣ spinothalamic tract
 - ▣ norepinephrine pathways

Anesthetics-lidocaine family

- ▣ Lidocaine
- ▣ marcaine
- ▣ bupivacaine
- Used in spinal, epidural, and local anesthesia
- Topical patches, ointments and creams

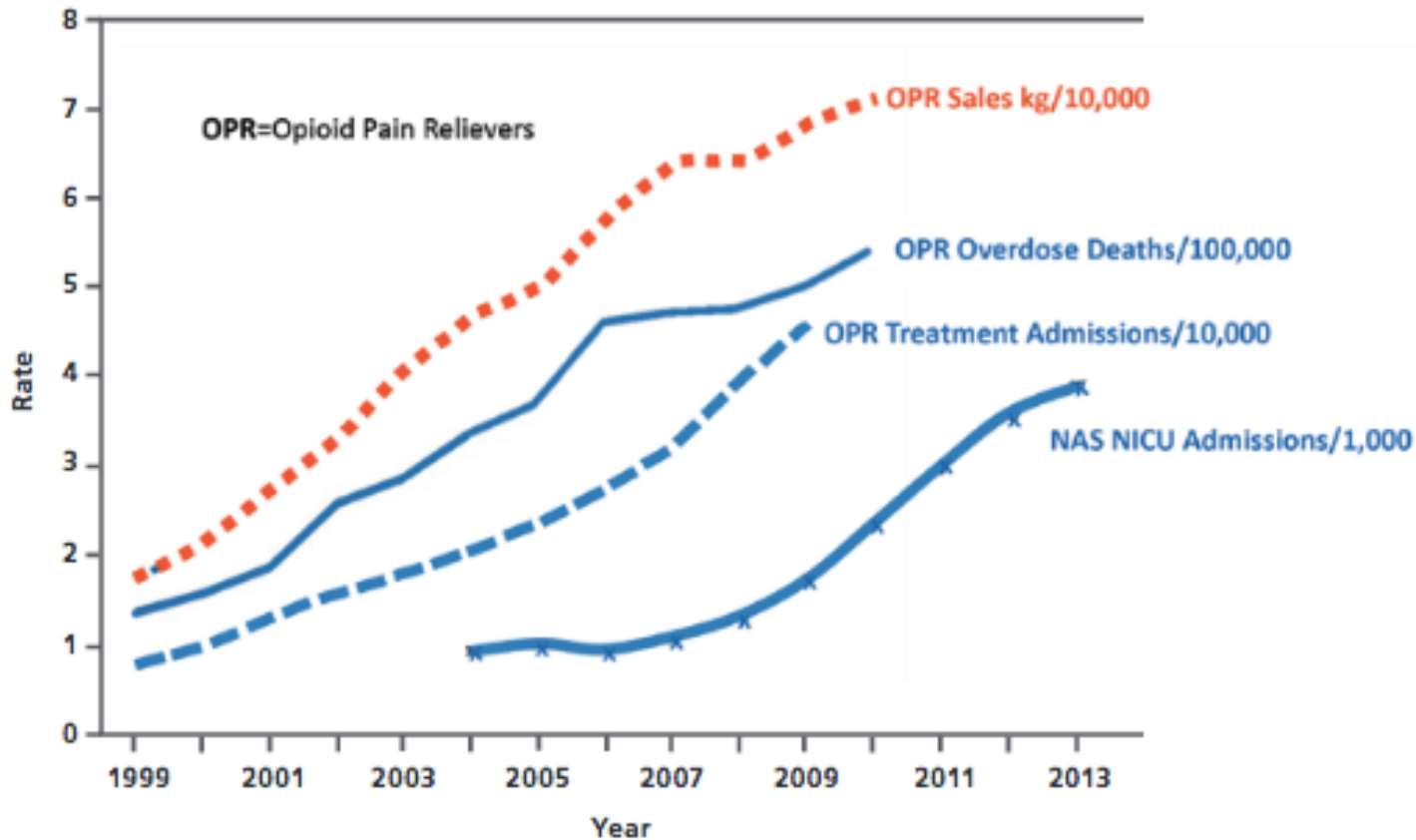
Mexiletine

- Oral medication
- Anti-arrhythmic medication (1 b)
- Sodium channel blocker (voltage-gated)
- Myotonia treatment

- Studied in post-operative pain management
- Chronic diabetic neuropathic pain

Opiates and opioids

□ Opioid epidemic



Opiates and opioids

- Natural (opiate)
 - ▣ morphine
 - ▣ heroin
- Synthetic
 - ▣ fentanyl
 - ▣ hydromorphone
 - ▣ hydrocodone
 - ▣ oxycontin
 - ▣ methadone
- in combination with other drugs
 - ▣ Vicodin
 - ▣ Norco
 - ▣ Lortab



Opiates and opioids

- Effects and side effects
 - ▣ Mu receptors
- Sedation, delirium
- Respiratory depression and failure
 - ▣ Cause of overdose deaths
- Itching
- Nausea
- Slows GI motility
 - ▣ Constipation or pseudo-obstruction
- pain sensitization
- Withdrawal



Non-steroidal anti-inflammatories (NSAIDs)

- Acetylsalicylic acid (Aspirin)
- Ibuprofen (Advil, Motrin, Nurofen)
- Naproxen (Aleve, Naprosyn)

- Selective COX-2 inhibitors
- Celecoxib (Celebrex)
- Inhibit prostoglandins, cyclo-oxygenases

- Excreted by kidneys
- Side effects:
 - Causes bruising
 - Can compete with aspirin cardioprotective effect

Acetaminophen

- paracetamol (Tylenol)
- Mechanism: Weak cyclo-oxygenase inhibitor
 - ▣ Not fully understood
- Metabolized by liver

Neuropathic pain medications

- Best for chronic pain
- Modulate pain perception and sensitization
- Delayed onset of action
 - ▣ Can take weeks to a month to see effect
 - ▣ Increase dose slowly to avoid side effects

Antidepressants for chronic pain

- Tricyclic antidepressants
 - ▣ Amitriptylene (Elavil)
 - ▣ Imipramine (Tofranil)
 - ▣ Nortriptylene (Pamelor)
- Serotonin-norepinephrine reuptake inhibitors (SNRIs)
 - ▣ Duloxetine (Cymbalta)
 - ▣ Venlafaxine (Effexor)
 - ▣ Desvenlafaxine (Pristiq)
 - ▣ Milnacipran (Savella)

Antiepileptics for chronic pain

- gabapentin (Neurontin, Gralise)
- gabapentin enacarbil (Horizant)
- pregabalin (Lyrica)

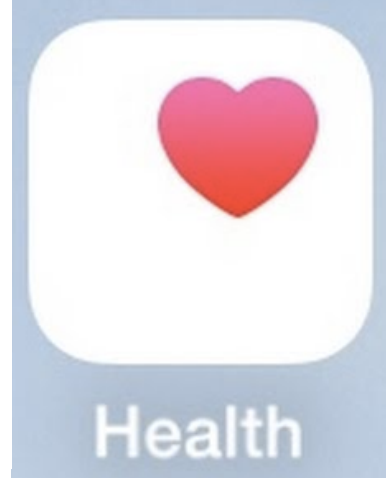
Non-medication pain management

- Heat/ Ice
- Stretching
- Massage
- Physical therapy, exercise
- Electrical nerve stimulation
- Biofeedback
- Mindfulness/meditation
- Adequate sleep
- Integrative/multidisciplinary pain management clinic

General recommendations

- When having surgery
 - Close monitoring not only during, but after
 - For sedating medications,
 - Start low, go slow
- If something doesn't seem right, say so
- Communication between care team
 - Neurologist
 - Anesthesiologist
 - Surgeon
 - Patient
 - caregivers

For your wallet or Smartphone



RECOMMENDATIONS FOR SURGERY AND ANESTHESIA

Patients with myotonic dystrophy often exhibit adverse reactions to sedatives, anesthetics, and neuromuscular blocking agents. Serious complications and fatalities can be avoided by careful preoperative assessment, avoidance of certain drugs, careful monitoring and good postoperative patient care throughout their hospitalization. It is especially important during post-op to monitor the heart and the respiratory system for ventilatory function and airway protection. Complications are not proportional to the severity of the disease; they often arise in seemingly mildly affected patients and it is worth considering whether regional anesthesia is a viable alternative or even if the surgical procedure is really necessary.

Preoperative:

- 1) Cardiological assessment: ECG essential, 24 hour Holter monitor if any indication of arrhythmia from ECG or history
- 2) Respiratory assessment: a) FEV1 and FVC both lying and standing b) chest x-ray, noting elevation of diaphragm or areas of atelectasis c) arterial blood gases
- 3) Premedication: avoidance of opiates, and caution with benzodiazepines

Intraoperative:

- 1) **Induction:** preferably a) avoid hypnotic agents with slow metabolism such as Thiopentone. Adverse reactions have also been reported with Propofol; lower doses are likely to be required. Careful titration of intravenous induction agents to avoid hypotension
- 2) **Relaxation:** a) avoid Succinylcholine b) short-acting, non-depolarizing muscle relaxants are best used and may be needed in smaller doses; recovery from these may be prolonged
- 3) **Reversal:** neostigmine may produce ACH-induced depolarization blockade
- 4) Protection of airway to minimize risk of aspiration; tendency to temporomandibular dislocation - care needed in manipulating jaw
- 5) Neuromuscular and capnograph monitoring
- 6) ECG monitoring essential due to risk of arrhythmias
- 7) Monitor core temperature; to avoid postoperative shivering, maintain normothermia by using warming pads
- 8) Avoid Potassium (K+) containing fluids

Postoperative: (first 24 to 48 hours)

- 1) Ensure respiration is fully re-established
- 2) Cardiac monitoring*
- 3) Respiratory monitoring: pulse oximetry, arterial blood gas analysis*
- 4) Use of a high dependency bed is preferable*
- 5) Early chest physiotherapy: these patients are especially prone to postoperative chest infections and atelectasis
- 6) Minimal use of opiates for analgesia; instead explore other methods, e.g. local anesthetic blocks or non-steroidal anti-inflammatory agents and paracetamol

*The extent to which these precautions are taken will depend on the length and nature of the procedure.

PERSONAL DATA

Name _____
 Address _____
 City _____
 State _____
 Zip _____
 Phone _____
 Cell Phone _____
 DOB _____
 Blood Type _____

EMERGENCY CONTACT

Name _____
 Relationship _____
 Cell Phone _____
 Home Phone _____
 Email _____

INSURANCE

Primary Company _____
 Policy # _____
 Secondary Company _____
 Policy # _____

PRIMARY PHYSICIAN

Name _____
 Address _____
 City _____
 State _____
 Zip _____
 Phone _____
 Cell Phone _____
 Email _____
 Relationship _____
 For additional information about myotonic dystrophy contact: _____
 Myotonic Dystrophy Foundation
 3031 Stanford Ranch Road
 Suite 2332
 Rocklin, California 95765
 info@myotonic.com
 www.myotonic.com

MEDICAL ALERT

The bearer of this card has **MYOTONIC DYSTROPHY**, a neuromuscular condition that may cause the following symptoms:


- muscle weakness, stiffness and balance difficulties
- extreme fatigue and sleepiness
- speech difficulties
- swallowing difficulties
- abnormal heart rhythm

PROBLEMATIC MEDICATIONS

- General anesthesia
- Benzodiazepines
- Quinins, procainamide, tocainide
- Liquid paraffin
- Neuroleptics
- Opiates

This list is not exhaustive and caution is recommended in the use of any medication which increases the metabolism in the cardiovascular or muscular systems. Consult with your physician prior to taking any medication.

MYOTONIC DYSTROPHY



MEDICAL ALERT AND HISTORY

Recommendations:

- MDF website
 - Short and long form guidelines

Anesthetic Management of Patients with Myotonic Dystrophy – Risks & Recommendations



Myotonic dystrophy (DM) is a genetic disorder that affects CNS, cardiac, respiratory, gastrointestinal, endocrine and muscular systems in ways that increase the risk of anesthesia.

Anesthesia Guidelines for pre-operative, intra-operative and post-operative care of DM patients, summarized below, are in the “Resources” section at www.myotonic.org.

Anesthetic Risks, as detailed in the Guidelines, result from DM effects that include:

- Cardiac conduction defects and potentially fatal arrhythmias
- Ventilatory insufficiency and poor airway protection
- Gastrointestinal dysmotility that frequently results in pseudo-obstruction
- Erratic responses to succinylcholine - though DM does not increase true malignant hyperthermia reactions, this drug should not be used in DM patients
- Prolonged and heightened sensitivity to sedatives and analgesics so that serious complications, including heightened risk of aspiration, are most common in the post-anesthesia period due to drug induced:
 - Reduction in level of consciousness
 - Exaggerated ventilatory weakness
 - Pharyngeal dysfunction with reduced airway protection
 - Gastrointestinal dysmotility and potential pseudo-obstruction

Methods to mitigate risk, detailed in the Guidelines, are summarized below:

- Preoperatively evaluate pulmonary, cardiac and gastrointestinal DM features in addition to its neurological and neuromuscular effects
- Use regional anesthesia when possible, to reduce or eliminate the need for general anesthesia
- Avoid pre-medications (e.g. sedatives and opioids) to the extent possible
- Keep the patient warm
- Consider precautionary application of defibrillator/pacer pads
- On induction, anticipate aspiration, and avoid the use of succinylcholine
- Adhere to strict extubation criteria. Given DM effects on CNS, GI, ventilatory and pharyngeal function, prepare the patient for prolonged post-anesthesia mechanical ventilation, commonly after having fully regained consciousness
- Prepare the patient for prolonged ventilatory assistance, for example by prior initiation of BiPAP with a mask that is immediately available post-anesthesia
- Plan for continuous SpO2 and ECG monitoring post-anesthesia until the patient fully regains pre-operative status, or longer if analgesics or sedatives are used in the post-anesthesia period
- Manage postoperative pain without narcotics when possible
- Encourage aggressive pulmonary toilet after anesthesia, including by use of a mechanical cough-assistance device if necessary

A shout out for research!

- Biobanking
 - ▣ If you are having a surgery (which involves removing any tissue)
 - We are interested!