Myotonic Dystrophy Clinical Care

Establishing Care for a Complex Condition

Advocating in the Clinic: Educating & Supporting Your Doctors

2014 Myotonic Dystrophy Foundation Annual Conference
Washington, D. C.
September 13, 2014
What to do when the going gets tough

• Complexities of Sleep
• Acute GI pain
• When you need surgery – what about anesthesia?
• How can you improve the situation?
Combatting Complex Conditions in DM

• Don’t give in to sleepiness
• Don’t rush to surgery for belly pain
• Respect but don’t overly fear anesthesia
• Take control – organize your community
Complexities of Sleep in DM

• Many factors reduce sleep quality in DM
  • Reduced sleep quality increases need for more sleep
  • Shallow breathing means shallow sleeping
    • Throat weakness increases snoring/obstruction – impairs breathing
    • Breathing muscle weakness makes breathing weak
    • Any cause of poor oxygenation reduces sleep quality
  • Abnormal movements in sleep also reduce sleep quality
• In addition to effects on breathing DM appears to directly increase need for sleep – like narcolepsy
• Many of these changes occur in the same person
Finding a Sleep Lab for DM

• They must have experience with DM
• They must keep looking – a single explanation (e.g., “sleep apnea”) may not explain the whole problem
• “Poor sleep hygiene” may be a consequence of DM sleep issues, not the cause
• Any one person may require several treatments
  • one for breathing issues
  • one for abnormal movements
  • one for narcolepsy-like generator demanding extra sleep
What to do about Acute GI pain?

• Go to ER, but tell them that DM frequently causes acute abdominal pain

• Acute GI pain in DM is often a non-surgical issue
  • It can cause “pseudo-obstruction”
  • Like everyone, individuals with DM get appendicitis, . . .

• Abdominal surgery in DM can make things worse

• Ask Emergency MDs to check MDF website and treat conservatively unless absolutely necessary
What to do about Anesthesia? – We need to communicate better

• To anesthesiologists “anesthetic risk in muscular dystrophy” implies acute deterioration of muscle
  • “Malignant Hyperthermia” type of reaction
  • They are rightly confident that they can prevent this
  • Even without specific prevention this is RARE in DM

• Biggest anesthetic risk in DM is drowsiness after anesthesia
  • Not particularly risky if well monitored
  • Exacerbated by using sedatives for sleep or pain

• Anesthesia CAN BE SAFE in DM if simple monitoring principles are followed
  • Monitor Heart and Breathing until fully awake
  • If pain or sedative medication is needed, continue monitoring
What Can you do to Improve Care?

• The primary goal is a doctor, team, hospital eager to learn with you

• Not all centers or physicians have the same goals
  • Some are solely focused on diagnosis, not management
  • Some are focused on general but not specialized care
  • Some incorporate many subspecialists

• There is power in numbers
  • 100 regional patients can improve 1 hospital more than on 10 hospitals
  • Even the most dedicated physicians require large patient volume to secure, strengthen and broaden hospital support
  • Get your DM community to consolidate at a chosen center
  • Keep working with that center to improve care - Create your own Center of Excellence
Combatting Complex Conditions in DM

• Don’t give in to sleepiness – keep demanding more thorough evaluation

• Don’t rush to surgery for belly pain - go to the Emergency Department but treat conservatively

• Respect but don’t overly fear anesthesia – it can be used safely and successfully with ongoing monitoring of heart and breathing function

• Take control – organize your community to help supportive physicians advocate for all necessary care