Myotonic Dystrophy Clinical Care

Establishing Care for a Complex Condition

Advocating in the Clinic: Educating & Supporting Your Doctors

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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
  - Exaccerbated 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