MYOTONIC DYSTROPHY AND ANAESTHESIA

Myotonic Dystrophy
SUPPORT GROUP


CRITICAL CARE TEAM
OV14 BXL
SN

Myotonic Dystrophy is not ***only*** a muscle condition

It is not ***only*** a neuromuscular condition

It is not ***only*** a genetic neuromuscular condition

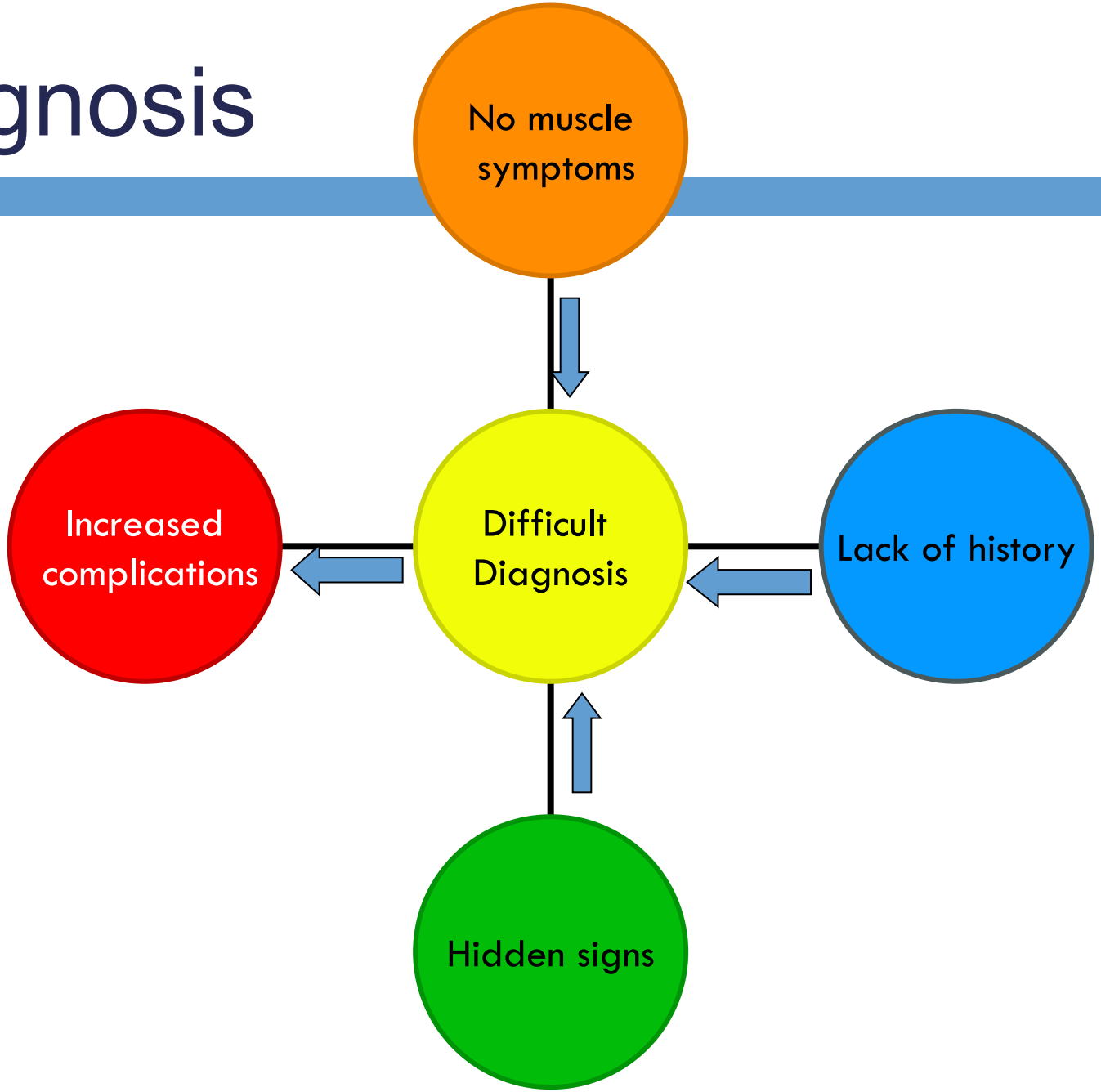
It is the most variable medical condition known

Prof. Peter Harper

Problems for your anaesthetist?

- Knowing the diagnosis
- Knowing the problems
- Knowing the solutions

Diagnosis

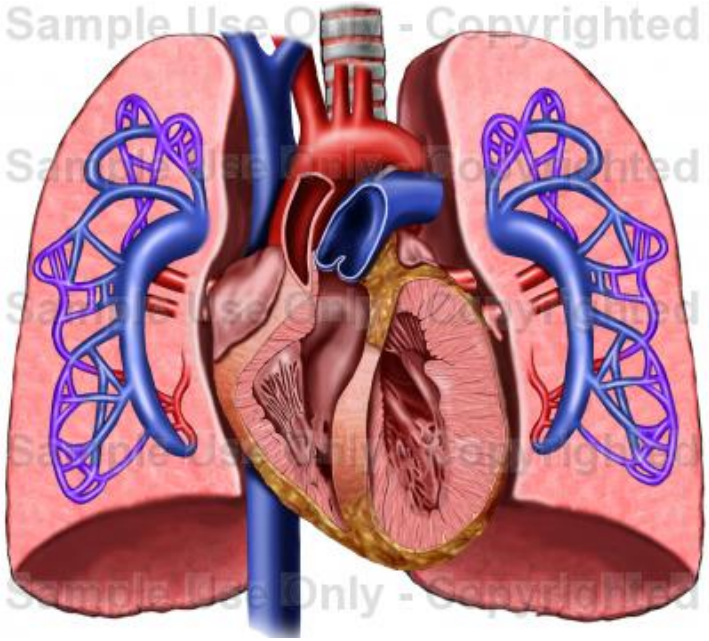

No muscle symptoms
Increased complications
Difficult Diagnosis
Lack of history
Hidden signs

Problem 1 – get the story right

- Difficulties with
  - Communication
  - Sleepiness
  - Lack of notes / information

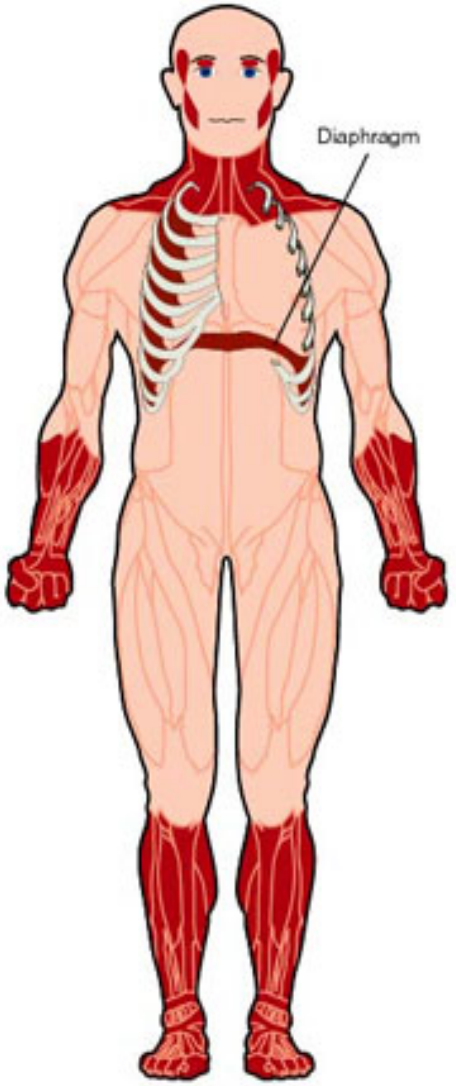
More problems…

- The muscles
- Reactions to drugs
- The heart and lungs



Muscle problems

- Things that bring on myotonia:
  - IV line
  - Pain from drug injection
  - Drugs
  - Cold, Shivering
  - Diathermy, Surgical manipulation
  - Postoperative pain



Drugs and Myotonia

- Suxamethonium (Scoline)
  - 1951
  - Used in emergencies only (!!)
  - Prolonged myotonic response possible
  - This is gradually now being replaced

Drug Sensitivity

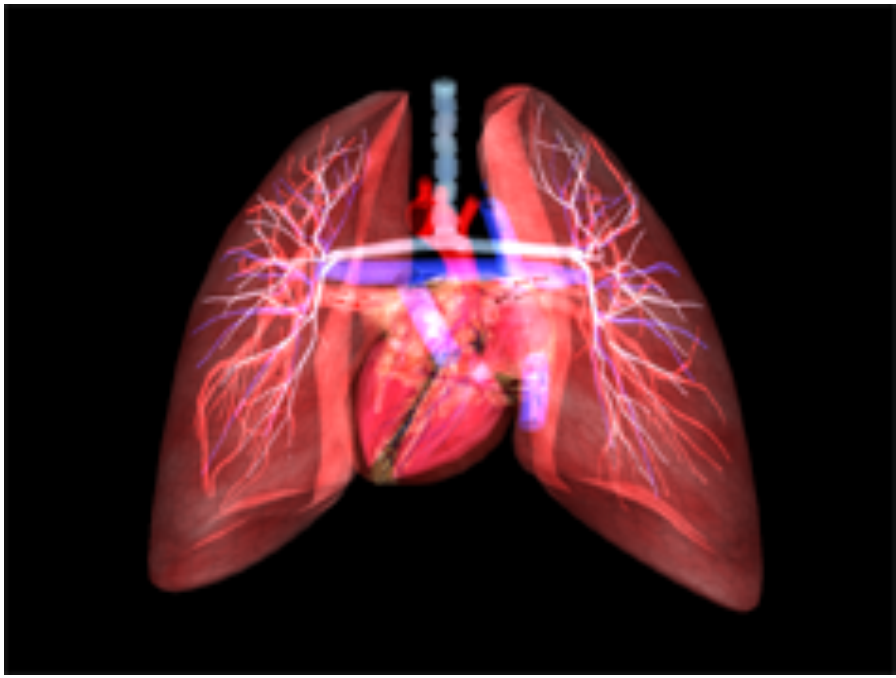
Almost all sedative, analgesic and anaesthetic drugs have been implicated and exonerated in the literature

(sometimes in the same edition of the same journal)

Little relationship between the severity of the muscle disease and how sensitive you are to sedatives

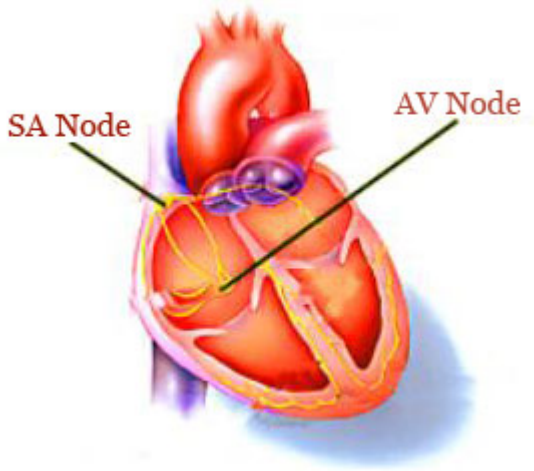
Heart and lung problems

- Complication rates very variable
  - 35 – 52% Aldridge 1985
  - 8.2% Mathieu 1997
  - 3.8 – 5.5% Imison 2001
  - 10% Sinclair 2010
- Commonest complication inability to breathe strongly after operation
- Good evidence that DM2 has less – 0.6% (Kirzinger 2010)

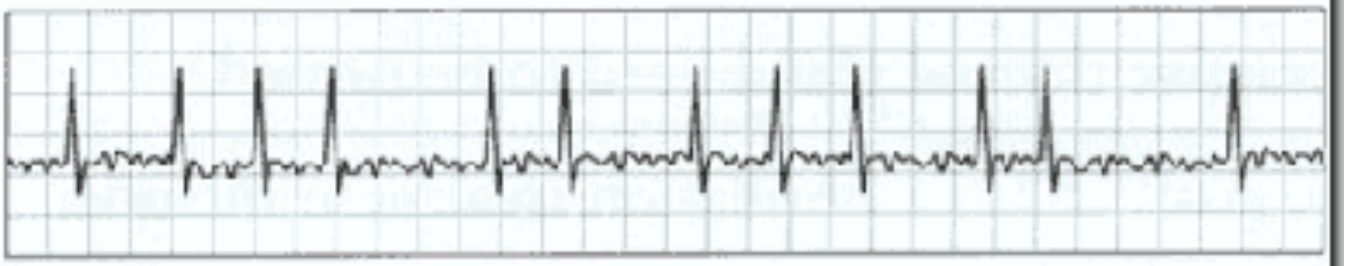


The heart

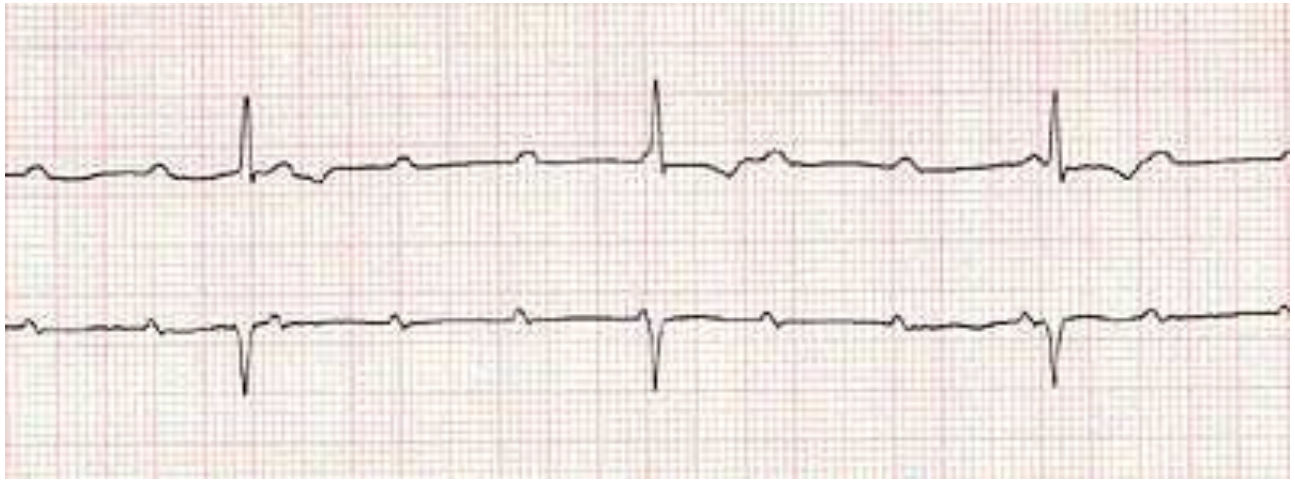
- There are often heart problems before muscle problems are obvious
- The stress of an operation can bring to the surface a heart problem
  - Irregular heart rhythmns - fast or slow
  - Failure of the “wiring” – needs pacemaker
  - Heart muscle problems
  - ICD vs Pacemaker



ECG tracing of a normal heart rhythm.



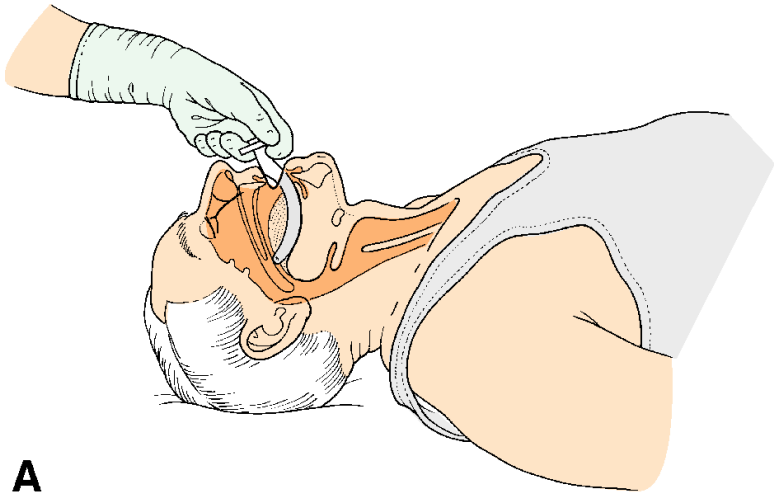
In atrial fibrillation, the tracing shows tiny, irregular "fibrillation" waves between heartbeats. The rhythm is irregular and erratic.



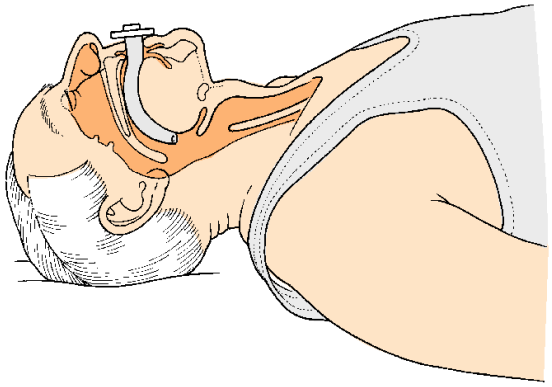
Lung problems

- Muscle weakness
- Lack of breathing coordination in brain
- Sleep apnoea
- Poor pharynx muscles - allows food into lungs
- Long recovery from anaesthetic
- Difficulty with airways?

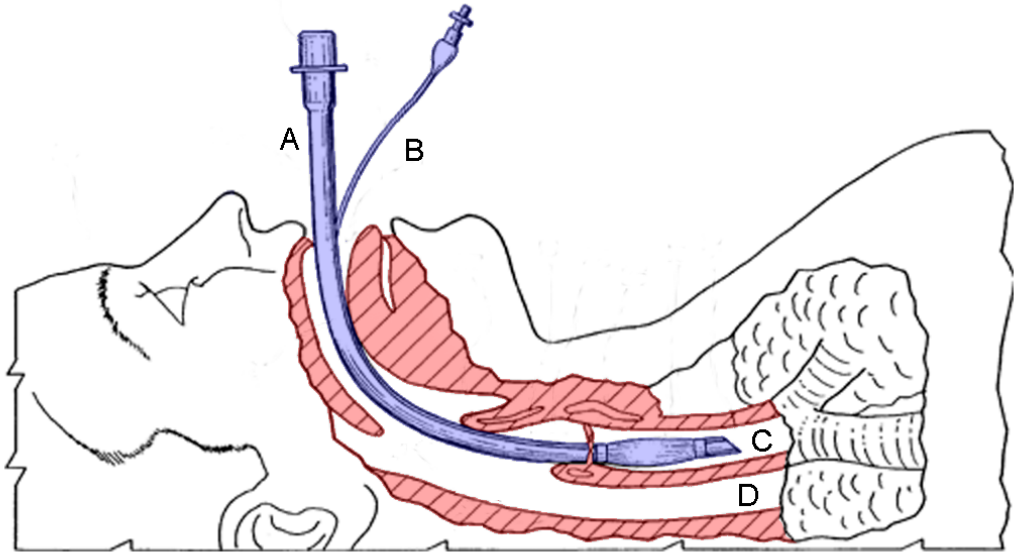
Airways



A



B



DM1 airway problems

- Little evidence
- Some reports in scientific journals
- Not enough information to prove it yet.

Problems that can occur

- Difficulty in seeing where to put the tube

and

Difficulty putting the tube there when you can see where you want to go

Things that make airways difficult:

- Short neck
- Receding lower jaw
- Small mouth
- Protruding incisors (buck teeth)
- Obesity

Simple tests

- Look at person's face and neck
  - From front
  - From side
- Neck movement
  - From side, is there good (>90°) movement?
- Mouth opening
  - Should be > 4 cm or 2 fingers
- Check teeth & tongue

Predictions

Predictions

- You can’t (predict correctly) for all of the people all of the time
- Tests only increase your level of suspicion of difficulty
- Needs detective work!

Ways of improving prediction

- Scoring systems
  - Mouth opening
  - Neck movements
  - Combinations

So we tried to do a bit of predicting……

- MDSG Conference 2011
- 54 “Willing” volunteers
- 2 anaesthetists

What we looked at…

- View of throat
- Mouth opening
- Neck movements
- Weight
- Receding chin?
- Buck teeth?
- Jaw movements

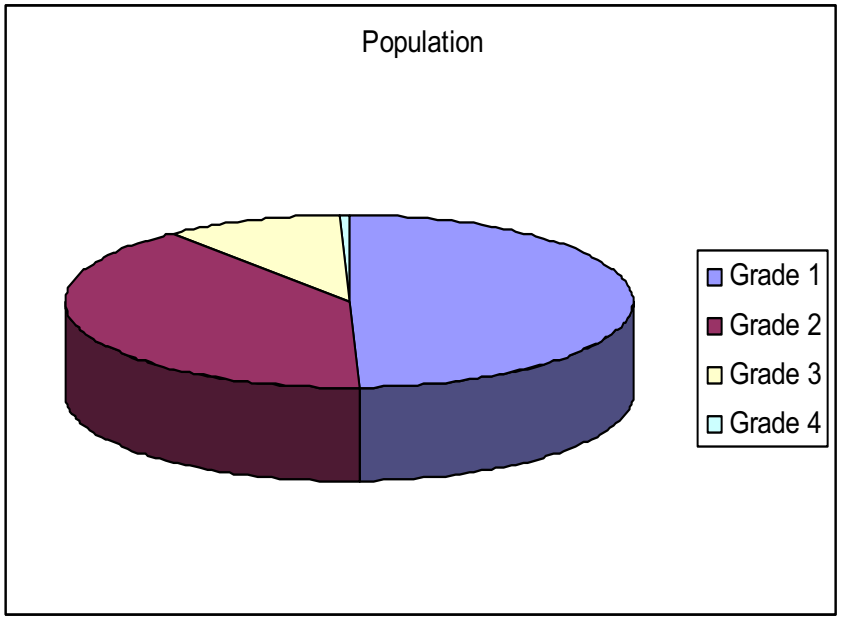
And then..

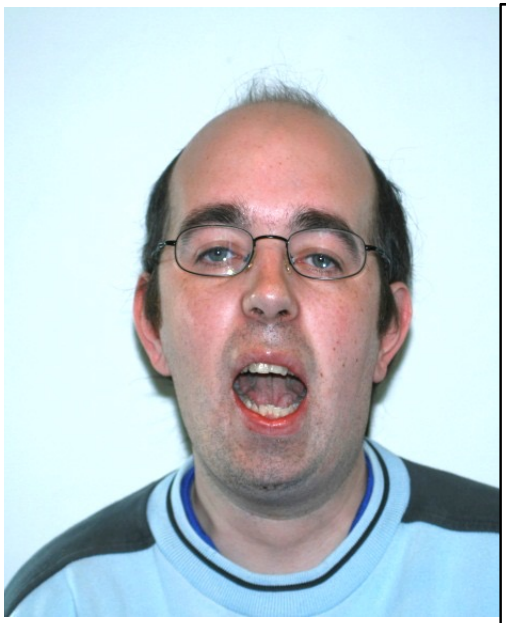
- Compare to internationally accepted population standards.
- And see if there were any differences

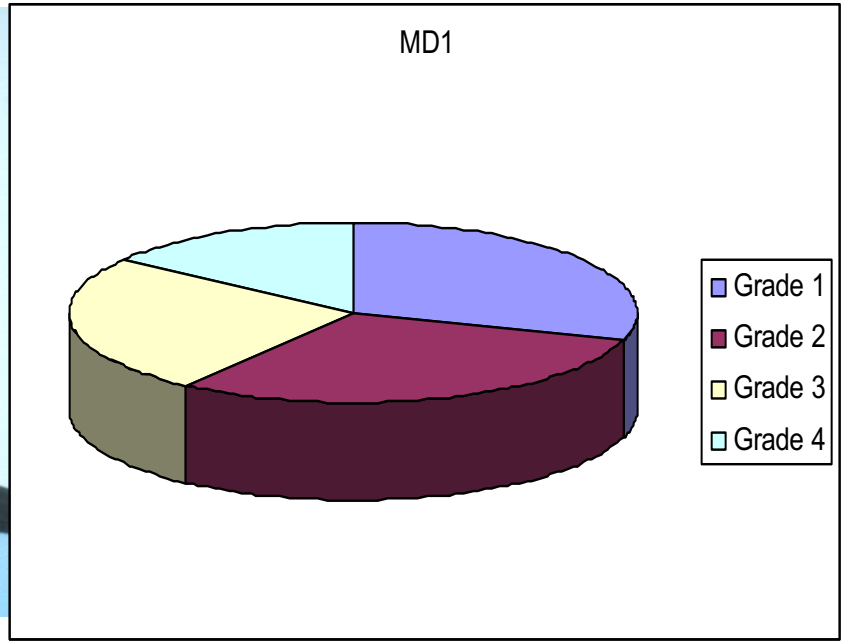
View of throat

1 2 3 4

Population	49.5%	40%	10%	0.5%
MD1	30%	30%	25%	15%

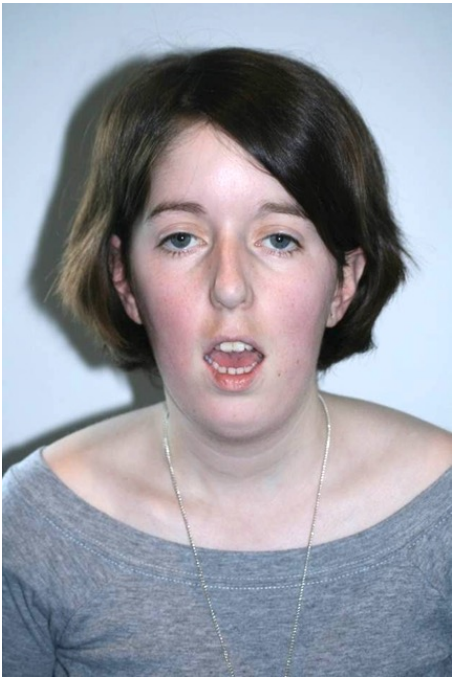

Population
Grade 1
Grade 2
Grade 3
Grade 4

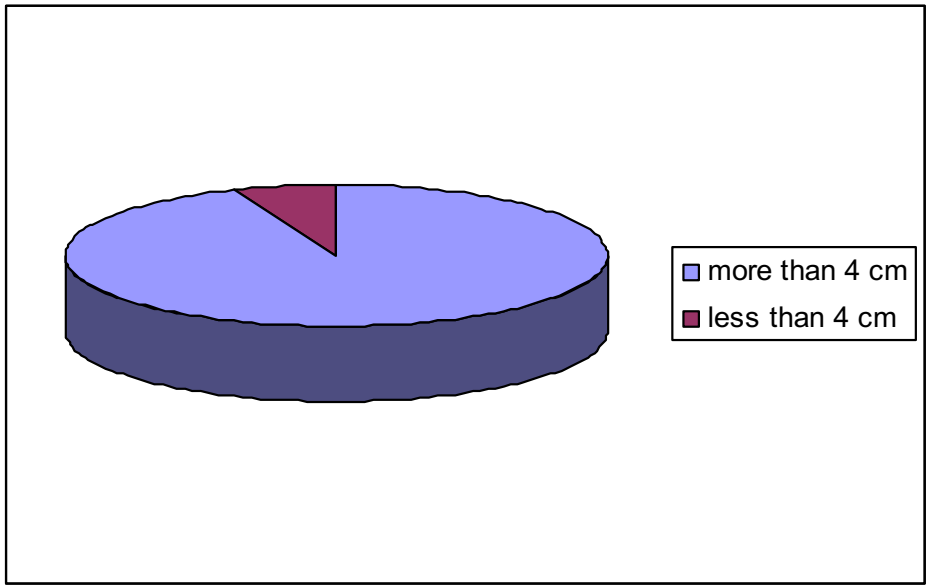


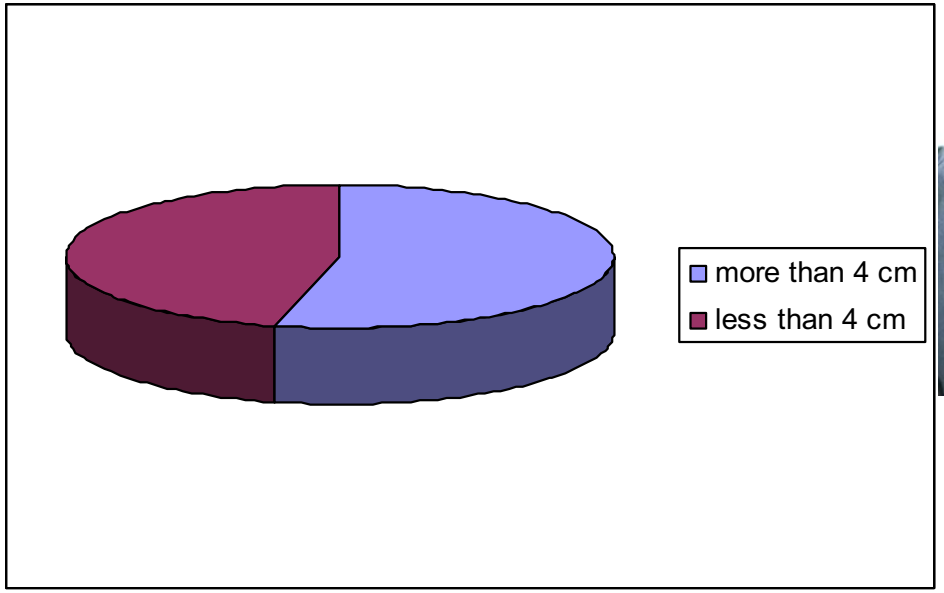

MD1
Grade 1
Grade 2
Grade 3
Grade 4

Mouth opening

	More than 4 cm	Less than 4 cm
Population	94%	6%
MD1	54%	46%




more than 4 cm
less than 4 cm


more than 4 cm
less than 4 cm

Other results…

- Weight – 78% of our group weighed more than 90kg (22% in population)

- Neck movement)
- Jaw movement)All the same
- Receding chin / buck teeth) as population standard

Problems with this information

- Very small number of people – 54
  - Means you can't presume it's correct for everyone
- All this predicting does not mean it will be a problem if you have an operation

What next?

- We need more information
- If you have an anaesthetic, let us know

The solution

- Planning
- Anaesthetic Assessment
- Investigations
- Appropriate techniques
- Good post-op facilities

Planning


MYOTONIC DYSTROPHY
ALERT
NAME
FURTHER MEDICAL DETAILS FROM
SEE REVERSE OF CARD

Anaesthetic Assessment

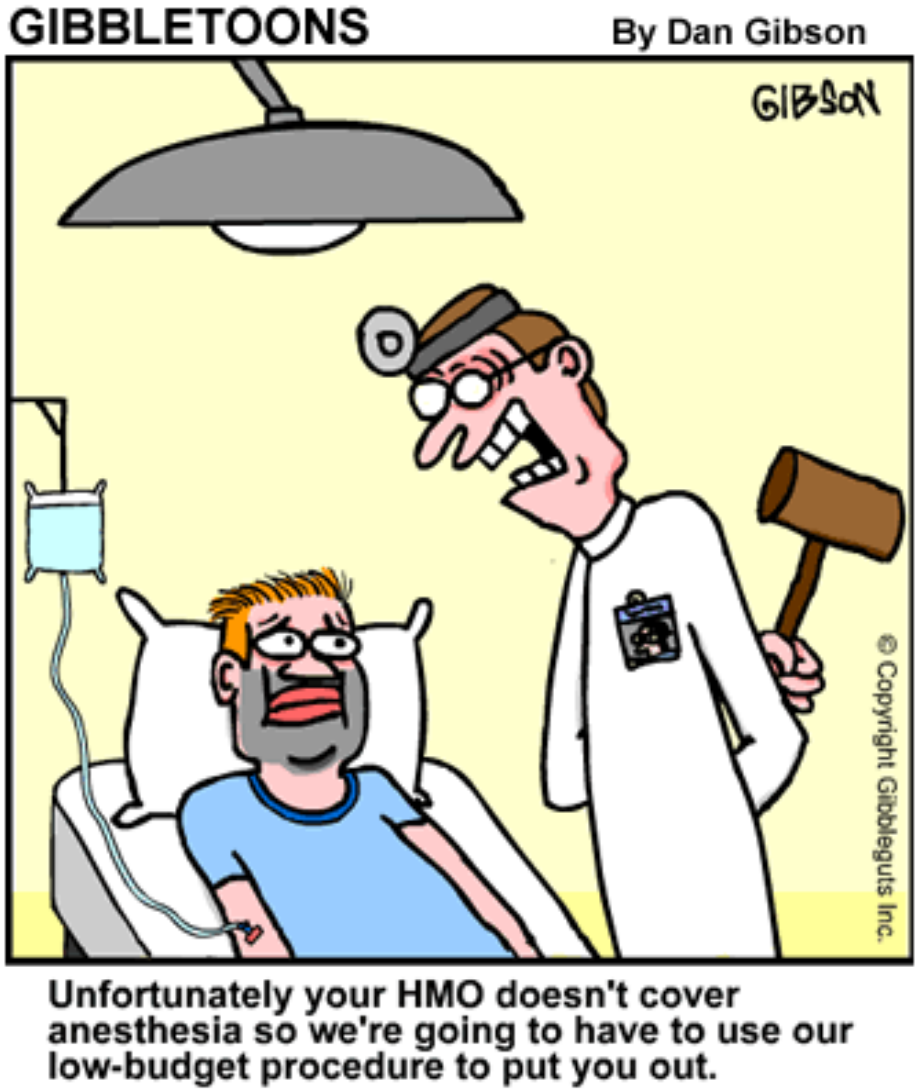
- Well in advance
- History and examination by doctor
- Investigations organised
- Discuss
  - Your concerns
  - Their concerns
  - Types of anaesthetic
  - Risks / benefits

Investigations

- ECG
  - Normality doesn't rule out problems
- Echocardiogram
  - If heart valve lesion suspected
  - May be of value for heart function assessment
- Chest Xray
  - ? Pre-existing problems
- Lung Function Tests
  - Volumes ? Restricted lung function
  - Blood ? Gas transfer problems

Types of anaesthetic

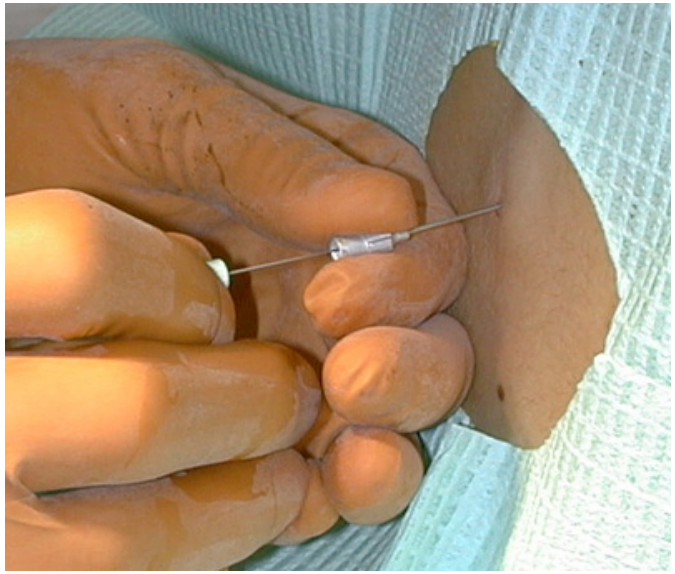
- General
- Regional
  - Spinal / epidural
  - Eye anaesthesia
- Local
  - Nerve blocks
  - Infiltration



General Anaesthetic

- Sometimes unavoidable
  - E.g. Heart / Brain operations
- MyD patients may be (or not) sensitive to all aspects of the anaesthetic:
  - Painkillers e.g. morphine
  - Sedatives e.g. valium
  - Muscle relaxants
- Still safe for majority if necessary

Avoid General Anaesthetic?



Spinal, epidural and nerve blocks all used successfully, but myotonia may still occur.

Spinal / Epidural Anaesthesia

- Suitable for:
  - Hip and knee replacements
  - Most hernias
  - Most gynaecology operations
  - Some lower bowel operations
  - Caesarian section

Nerve blocks

- Suitable for procedures on limbs and for eye surgery
- Needs care with sedation

Post-operative management

- Pain management
- Keeping warm & avoiding shivering
- Physiotherapy
- High index of suspicion for respiratory complications
- High Dependency (level 2) or Intensive (level 3) Critical Care

Sedation

- Endoscopy / Colonoscopy
- Bronchoscopy
- Dentistry
- Minor gynae procedures
- Accident and Emergency

Problems with sedation

- Use of sedative drugs that MyD patients are sensitive to
- Usually given by doctor doing procedure
- Rarely an anaesthetist involved
- Rarely in a high care area

- This can be a *lethal* combination

Summary

- Ensure your surgeon / physician knows the problems
- Talk to the anaesthetist early, before the day of operation
- Get and give as much information as you can
- Most procedures are safe with proper planning and work up

And…

- If you are having an anaesthetic, let me know..

Ian.Bowler@doctors.org.uk

Myotonic Dystrophy
SUPPORT GROUP