LETTER TO THE EDITOR

HOW GOOD AT NEUROLOGY ARE YOU?

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Every time I get the wrong answer in the 'Test yourself' section, my belief in subspecialization, for the benefit of our patients, is reinforced. I have hesitated writing, but eventually decided that the recent question on central core disease (*Practical Neurology*, 2003, 3, 251–253) merited further comment. The take-home message is simple and important – central core disease may

TEST YOURSELF

How good at neurology are you?

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Questions

1. Please consider the following:
A 50 year-old man presented with a two year history of behavioural changes and slowness. His MRI is shown below. His mother had died from 'paralysis'.
What is the diagnosis ?

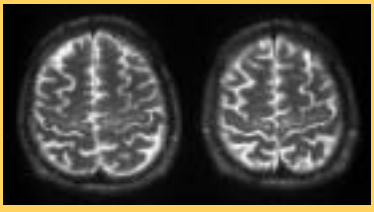


Image courtesy of Dr John Thorpe

2. Please read the following passage.
You receive a telephone call regarding a 30 year-old man about to undergo incision and drainage of a pilonidal abscess under general anaesthetic. His father had died unexpectedly following an appendicectomy. This led the anaesthetist to ask him some questions about his muscles. He said he had never been great at sport and he was the one who always dropped the ball in his family. The anaesthetist asks you whether this is relevant:
(a) What would be the best way to confirm the diagnosis ?
  (i) EMG
  (ii) Muscle biopsy
  (iii) Genetic test
  (iv) Plasma creatine kinase level
  (v) Other
(b) What would you advise the anaesthetist?
  (i) Reassure
  (ii) Proceed with operation, but be prepared for prolonged post-operative recovery
  (iii) Proceed with operation but do not administer curare-like agents
  (iv) Proceed with operation but do not administer non-depolarizing blockers
  (v) Cancel operation

3. Please study this plain skull x-ray.
This 84 year-old man presented with deafness. What is the diagnosis?

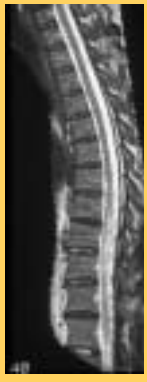
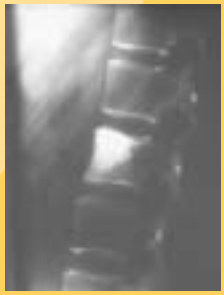


Image courtesy of Dr John Thorpe.

4. Please consider the following case:
A 66 year old man presented to his family doctor with back pain. His doctor thought the neurological examination was normal and arranged some Xrays. He sends the films through to you and asks whether any further investigation is necessary.
If you could only choose one, which of the following would you recommend ?
(a) measure blood calcium level
(b) measure parathormone level
(c) reassure
(d) blood cultures
(e) bone scan



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5. Please read the following:
You are asked, by his new family doctor, to review a 45-year-old man of low intelligence at a local institution on account of immobility and incontinence. It is unclear whether this is long-standing but, on the basis of your clinical findings, you arrange an MR scan.
On the basis of this scan, which of the following would you arrange?
(a) CSF analysis, including oligoclonal bands
(b) Very long chain fatty acids
(c) Organic and aminoacids
(d) White cell enzymes assay
(e) None of the above

be associated with malignant hyperthermia, which is most likely to develop in association with general anaesthesia and which can be fatal. A patient is described whose neuromuscular symptoms were limited to 'never been great at sport and he was the one who always dropped the ball in his family', and whose father died unexpectedly during anaesthesia. I appreciate the constraint of space available to the question setters, but three issues require further comment.

Firstly, it is suggested that the 'best way to confirm the diagnosis' would be to review the father's notes, the implication being that if central core disease was confirmed in the father, then it could be assumed that the son had the same condition. Experience with many genetic disorders shows that such an approach is fundamentally flawed. Particularly when a parent has an autosomal dominant disorder, with 50 : 50 risk of transmission, time and again we see anxiety and introspection leading to unfounded concern that a child is developing symptoms of the disease. I frequently see families with conditions such as Charcot-Marie-Tooth disease and facioscapulohumeral muscular dystrophy, in which there is anxiety on behalf of either the parents or the child that the child is developing problems, such as clumsiness of gait or winging of the scapulae, when subsequent testing proves that the child has not inherited the disorder. Even experienced examiners may 'over-interpret' signs in such offspring. The point, quite simply, is that the diagnosis in the offspring must be proved by appropriate assessment, not by implication. Not everybody who is poor at games and can't catch a ball has a neuromuscular disorder.

Secondly, the advice given to the anaesthetist was to 'Proceed with the operation but do not administer curare-like agents'. That is far from adequate. Although succinylcholine can trigger malignant hyperthermia, volatile anaesthetics, particularly halothane, are a much more potent trigger.

Finally, there was an unfortunate choice of histology. The H&E section shows entirely non-specific 'myopathic' changes without a core in sight. The characteristic cores are best seen with oxidative enzyme reactions, such as NADH.

Few general neurologists will have experience of central core disease or malignant hyperthermia but in this increasingly litigious age lack of personal experience is not a defence against incorrect advice.

RESPONSE

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Dr Hilton-Jones's comments are gratefully received. He highlights an important limitation of multiple choice/short answer questions where it is often difficult to strike the right balance between providing enough information to make the reader select the desired diagnosis, whilst not making the question too easy. The questions, although largely based on cases personally seen, are inevitably artificial. Like crosswords, one needs to get into a particular mindset. In this particular question we were really just trying to make a pragmatic point that if his father had died from a primary surgical mishap the level of concern would be different. The (sub) specialist advice and perspective is, however, welcomed and encouraged.