TEST YOURSELF

How good at neurology are you?

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QUESTIONS

1. Consider the following case.

A 67-year-old man presents with at least 2 years of painful numb feet. His body mass index is 26. A 10 m walk takes 8 s, unaided. Routine inquiry and investigations for common causes of neuropathy, including random blood glucose are all negative. Electrophysiological examination reveals a chronic axonal polyneuropathy. Which of the following is false?

- (a) An oral glucose load should be given to detect impaired glucose tolerance.
- (b) Hypertriglyceridaemia, not linked to diabetes mellitus, is associated with chronic axonal polyneuropathy.
- (c) Recent reports support a role for anti-ganglioside antibodies in chronic axonal polyneuropathy.
- (d) Taking a detailed occupational history may reveal a relevant history of exposure to solvents or lead.
- (e) Quality of life is significantly affected.

2. Concerning pergolide – which of the following are true?

- (a) Nausea and hypotension are the most common adverse effects.
- (b) Pergolide associated valvular heart disease is an idiosyncratic reaction.
- (c) Valve regurgitation is a rare adverse effect.
- (d) Pergolide associated valvular heart disease may be reversible.
- (e) The natural history of pergolide associated valvular heart disease is benign.

Neurology are you?

Figure 1

3. Look at Figure 1.

There are two pictures of an Asian youth illustrating the appearance of the right neck (a) and shoulder (b).

- (a) Where is the lesion?
- (a) What is the cause?

4. Name the disorder(s) featured in each of the following popular films.

- (a) *Vertigo*, 1958
- (b) *Rain Man*, 1988
- (c) *My Left Foot*, 1989
- (d) *Awakenings*, 1990
- (e) *Lorenzo's Oil*, 1992
- (f) *Hilary and Jackie*, 1998
- (g) *Iris*, 2001
- (h) *A Beautiful Mind*, 2001

Answers on page 254.

ANSWERS

1. (c) is false

Overweight patients like this man with pain, are more likely than matched, non-neuropathy controls, to show impaired glucose tolerance (see Hughes *et al.* in *Brain* 2004, 127, 1723–30). This is less likely when the neuropathy is painless. Impaired glucose tolerance is usually defined by glucose levels of 7.8–11 mmol/L 2 hours after a 75g oral glucose load. The fasting insulin concentration is also likely to be elevated. After adjusting for body mass index, hypertriglyceridaemia not linked to diabetes mellitus, is more likely in patients than controls, suggesting a 'metabolic' basis to chronic axonal polyneuropathy, which is not a direct consequence of hyperglycaemia *per se*.

2. (a) and (d) are true

Information on pergolide associated valvular heart disease is evolving. A high prevalence (> 40%) of 'serious' valve disease has recently been described in an echocardiographic case-control study (*Neurology* 2004, 63, 301–4). It seems highly likely that the ergot base, perhaps via serotonin excess mediated reactions, at 5-HT_{2B} receptors, is responsible for the rare cases of retroperitoneal, pericardial and pleural fibrosis. Pergolide associated *valvular* heart disease is presumably part of this spectrum, but may be much more common. Duration of exposure to the drug increases the risk of valvulopathy but the natural history is poorly understood. Some but not all lesions improve with drug withdrawal, but valvular insufficiency may necessitate replacement. The need to detect and monitor this adverse effect, fuelled by marketing of newer, non-ergot based dopamine agonists, spells a decline in pergolide use.

3.

- (a) Spinal accessory nerve, branch to the upper fibres of trapezius.
- (b) Iatrogenic - lymph node biopsy in the neck (see scar) for suspected TB.

4. Where the film is based on true events, the name of the sufferer portrayed is included in parenthesis.

- (a) Agoraphobia, acute melancholia, psychotic depression? Not a vestibulopathy!
- (b) Autism/idiot savant.
- (c) Cerebral palsy (Christy Brown).
- (d) Post encephalitic parkinsonism.
- (e) Adrenoleukodystrophy (Lorenzo Odone).
- (f) Multiple sclerosis (Jaqueline du Pré).
- (g) Alzheimer's disease (Iris Murdoch).
- (h) Schizophrenia (John Forbes Nash Jr.)