

Core curriculum questions 2002/3. Authors' comments and suggested "best answers".

Here are the TSC questions for 2002/3 with the authors' comments and suggested "best answers". A contact email is provided for each author. The format of some of the questions differs from that on the website as some questions had to be adapted to fit the online format. It should not be too difficult to match your answer to each scenario and question despite this.

1. Headache		Question setter: Dr R. Davenport rijd@skull.dcn.ed.ac.uk
Scenario 1		
An 18 yr old female presents with a 6 week history of severe, progressive incapacitating headaches. She now spends much of her time lying flat, as this is the only position in which she is headache free. There are no other neurological symptoms, and she is on no medication other than simple analgesia (up to 6 aspirin/day). She suffered from migraine when she was a child. Examination is unrewarding, although you are uncertain about whether her optic discs are normal as you only got a poor glimpse.		
Question 1		
The most likely diagnosis is:		
Chronic daily headache syndrome secondary to analgesic overuse		
Idiopathic intracranial hypertension		
Hypnic headaches		
SUNCT		
Spontaneous intracranial hypotension		Best answer 5
Status migrainosus		
Unclassifiable		
Question 2		
She undergoes the investigations shown in figures 1.1, 1.2, 1.3, and 1.4. The most appropriate interpretation is:		
Confirmation of the diagnosis of idiopathic hypertension, with evidence of decreased CSF absorption on the radionuclide scan, and dural ectasia as a result of the increased pressure		Best answer 1
No diagnosis may be reached on the basis of these tests		
A low pressure headache secondary to a CSF leak		
Froin's syndrome, with CSF block secondary to a structural thoracic spinal cord lesion, and secondary intracranial hypertension		
Evidence of seeding of the CSF pathways, with enhancement of the basal meninges, cause uncertain		

Answers and comments

Question 1:

Spontaneous intracranial hypotension. The crucial aspect is the postural nature of her headache, and is precisely the opposite of intracranial hypertension, and sounds like a post lumbar puncture headache, without the LP. SIH is a rare, but recognised cause of headache (1); the cause of this syndrome cannot always be identified, but many are related to a CSF leak.

Question 2:

CSF absorption on the radionuclide scan, and dural ectasia as a result of the increased pressure. Figure 1.1 shows intracranial air, sucked in during a failed lumbar puncture (before the diagnosis was appreciated) – In SIH, the opening pressure is typically less than 6 cm water. In this case, clearly it was negative, allowing air to enter the CSF space during the "dry" tap, confirming the diagnosis of SIH. Figure 1.2 is a radionuclide

cisternogram, and shows evidence of tracer pooling in the high thoracic area (the lower accumulation is early renal excretion). Figure 1.3 shows marked dural ectasia, and figure 1.4 is a CT myelogram showing leakage of contrast from the previously identified leak in the high thoracic region, probably due to incidental trauma to an area of dural ectasia. She was successfully treated with blood patching.

References

1. Rando TA, Fishman RA. Spontaneous intracranial hypotension: report of 2 cases and review of the literature. *Neurology* 1992;**42**:481-487.

Figures

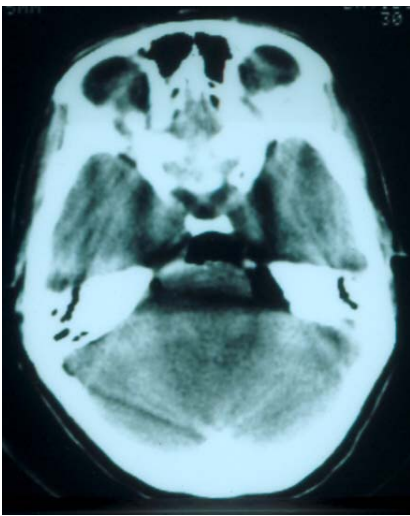


fig 1.1

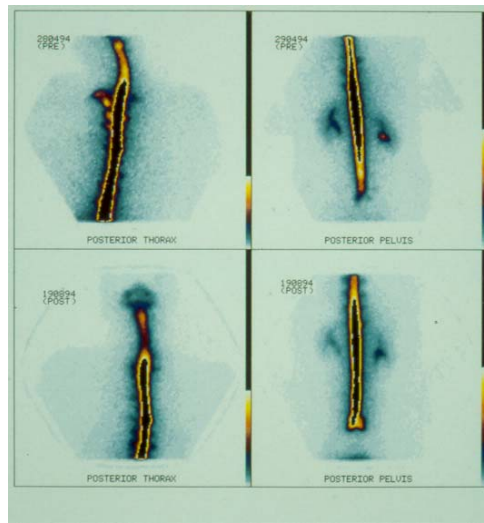


fig 1.2



fig 1.3



fig 1.4

2. Management of carotid disease		Question setter: Dr TP Enevoldson (peter@enevoldson.freeserve.co.uk)
Scenario 1		
A patient is referred to you for your opinion on the best management to prevent future stroke. Hypertension has been successfully treated (130/74) with perindopril and indapamide, and atenolol also has prevented recurrence of exertional angina for the last two years. Simvastatin 40 mg has recently been added. Previously aspirin 75mg daily was used but this was recently changed to clopidogrel 75 mg (rightly or wrongly) by the referring doctor (except in scenario 7 below).		
Select appropriate management options for each of the following scenarios (one or more may apply). Assume the carotid findings at ultrasound have been confirmed by another method, such as DSA or validated MRA, which did not reveal any other lesions. The European method has been used in quoting % stenoses.		
Question 1		
Male, age 68, middle cerebral TIA two weeks earlier. 90% ipsilateral carotid stenosis, 30% contralateral disease. No other significant diseases.		
Add aspirin to clopidogrel		
Change to aspirin & dipyridamole instead of clopidogrel		
Referral to experienced vascular surgeon for consideration of carotid endarterectomy		Best answer 3
Referral to interventional radiologist experienced in carotid stenting surgeon		
Anticoagulation with heparin, then warfarin vascular		
Anticoagulation with warfarin immediately		
Referral to experienced vascular surgeon for consideration of carotid endarterectomy, with delayed post-operative warfarin.		
Start prednisolone 60mg and arrange temporal artery biopsy		
Continue present medication, without change or addition		
Anticoagulation with heparin, before referral to vascular surgeon		
Question 2		
Female, age 65, two episodes of amaurosis fugax 12 weeks before. 75% ipsilateral carotid stenosis, 30% contralateral disease. No other illnesses.		
Add aspirin to clopidogrel		
Change to aspirin & dipyridamole instead of clopidogrel		
Referral to experienced vascular surgeon for consideration of carotid endarterectomy		
Referral to interventional radiologist experienced in carotid stenting surgeon		
Anticoagulation with heparin, then warfarin vascular		
Anticoagulation with warfarin immediately		
Referral to experienced vascular surgeon for consideration of carotid endarterectomy, with delayed post-operative warfarin.		
Start prednisolone 60mg and arrange temporal artery biopsy		
Continue present medication, without change or addition		Best answer 9
Anticoagulation with heparin, before referral to vascular surgeon		
Question 3		
Female, 60 years old, small branch retinal artery occlusion 12 weeks before. 70% ipsilateral carotid stenosis, 20% contralateral disease. Other eye severely amblyopic.		
Add aspirin to clopidogrel		
Change to aspirin & dipyridamole instead of clopidogrel		
Referral to experienced vascular surgeon for consideration of carotid endarterectomy		
Referral to interventional radiologist experienced in carotid stenting surgeon		
Anticoagulation with heparin, then warfarin vascular		
Anticoagulation with warfarin immediately		
Referral to experienced vascular surgeon for consideration of carotid endarterectomy, with delayed post-operative warfarin.		
Start prednisolone 60mg and arrange temporal artery biopsy		
Continue present medication, without change or addition		Best answer 9
Anticoagulation with heparin, before referral to vascular surgeon		

Question 4	
Male, 68 years old, middle cerebral artery TIA 18 months earlier. 80% stenosis of ipsilateral carotid artery, 30% contralateral disease. No other illnesses.	
Add aspirin to clopidogrel	
Change to aspirin & dipyridamole instead of clopidogrel	
Referral to experienced vascular surgeon for consideration of carotid endarterectomy	
Referral to interventional radiologist experienced in carotid stenting surgeon	
Anticoagulation with heparin, then warfarin vascular	
Anticoagulation with warfarin immediately	
Referral to experienced vascular surgeon for consideration of carotid endarterectomy, with delayed post-operative warfarin.	
Start prednisolone 60mg and arrange temporal artery biopsy	
Continue present medication, without change or addition	Best answer 9
Anticoagulation with heparin, before referral to vascular surgeon	
Question 5	
Female, 75 years old. Multiple amaurosis fugax over 3 days, followed by a dense monocular visual loss due to anterior ischaemic optic neuropathy 3 days before. 90% ipsilateral carotid stenosis, 60% contralateral disease. Some ache above the eyes in the last 2 weeks.	
Add aspirin to clopidogrel	
Change to aspirin & dipyridamole instead of clopidogrel	
Referral to experienced vascular surgeon for consideration of carotid endarterectomy	
Referral to interventional radiologist experienced in carotid stenting surgeon	
Anticoagulation with heparin, then warfarin vascular	
Anticoagulation with warfarin immediately	
Referral to experienced vascular surgeon for consideration of carotid endarterectomy, with delayed post-operative warfarin.	
Start prednisolone 60mg and arrange temporal artery biopsy	Best answer 8
Continue present medication, without change or addition	
Anticoagulation with heparin, before referral to vascular surgeon	
Question 6	
Male, age 75 years. Multiple unilateral amaurosis fugax 2 weeks before. 90% ipsilateral carotid stenosis, 40% contralateral disease. Also non rheumatic atrial fibrillation, apical rate 80, normal transthoracic echo.	
Add aspirin to clopidogrel	
Change to aspirin & dipyridamole instead of clopidogrel	
Referral to experienced vascular surgeon for consideration of carotid endarterectomy	
Referral to interventional radiologist experienced in carotid stenting surgeon	
Anticoagulation with heparin, then warfarin vascular	
Anticoagulation with warfarin immediately	
Referral to experienced vascular surgeon for consideration of carotid endarterectomy, with delayed post-operative warfarin.	Best answer 7
Start prednisolone 60mg and arrange temporal artery biopsy	
Continue present medication, without change or addition	
Anticoagulation with heparin, before referral to vascular surgeon	
Question 7	
Male, age 70 years. Middle cerebral artery TIA 4 weeks before. 90% ipsilateral carotid artery stenosis, 40% contralateral disease. Prosthetic aortic valve, requiring anticoagulation.	
Add aspirin to clopidogrel	
Change to aspirin & dipyridamole instead of clopidogrel	
Referral to experienced vascular surgeon for consideration of carotid endarterectomy	
Referral to interventional radiologist experienced in carotid stenting surgeon	Best answer 4
Anticoagulation with heparin, then warfarin vascular	
Anticoagulation with warfarin immediately	
Referral to experienced vascular surgeon for consideration of carotid endarterectomy,	

with delayed post-operative warfarin.	
Start prednisolone 60mg and arrange temporal artery biopsy	
Continue present medication, without change or addition	
Anticoagulation with heparin, before referral to vascular surgeon	
Question 8	
Male, 70 years old. Isolated mild hemiparetic stroke, clearing after 2 weeks, occurring 6 weeks before. 50% carotid stenosis on both sides. Two previous mild hemiparetic strokes leaving no deficit over the last year. Normal rhythm and echo. MRI shows 3 appropriate small infarcts.	
Add aspirin to clopidogrel	
Change to aspirin & dipyridamole instead of clopidogrel	
Referral to experienced vascular surgeon for consideration of carotid endarterectomy	
Referral to interventional radiologist experienced in carotid stenting surgeon	
Anticoagulation with heparin, then warfarin vascular	
Anticoagulation with warfarin immediately	
Referral to experienced vascular surgeon for consideration of carotid endarterectomy, with delayed post-operative warfarin.	
Start prednisolone 60mg and arrange temporal artery biopsy	
Continue present medication, without change or addition	Best answer 9
Anticoagulation with heparin, before referral to vascular surgeon	
Question 9	
Male, 83 years old. Middle cerebral TIA 1 week before. 90% bilateral carotid stenoses. Otherwise in good nick	
Add aspirin to clopidogrel	
Change to aspirin & dipyridamole instead of clopidogrel	
Referral to experienced vascular surgeon for consideration of carotid endarterectomy	Best answer 3
Referral to interventional radiologist experienced in carotid stenting surgeon	
Anticoagulation with heparin, then warfarin vascular	
Anticoagulation with warfarin immediately	
Referral to experienced vascular surgeon for consideration of carotid endarterectomy, with delayed post-operative warfarin.	
Start prednisolone 60mg and arrange temporal artery biopsy	
Continue present medication, without change or addition	
Anticoagulation with heparin, before referral to vascular surgeon	
Question 10	
Female, 72 years old. Cholesterol retinal emboli noted at routine optician's check 6 weeks earlier. 90% ipsilateral carotid artery stenosis, 30% contralateral disease. Otherwise well.	
Add aspirin to clopidogrel	
Change to aspirin & dipyridamole instead of clopidogrel	
Referral to experienced vascular surgeon for consideration of carotid endarterectomy	
Referral to interventional radiologist experienced in carotid stenting surgeon	
Anticoagulation with heparin, then warfarin vascular	
Anticoagulation with warfarin immediately	
Referral to experienced vascular surgeon for consideration of carotid endarterectomy, with delayed post-operative warfarin.	
Start prednisolone 60mg and arrange temporal artery biopsy	
Continue present medication, without change or addition	Best answer 9
Anticoagulation with heparin, before referral to vascular surgeon	
Question 11	
Male age 73 years. Middle cerebral artery TIA 8 weeks before. 90% ipsilateral carotid artery stenosis, 95% contralateral disease. Previous radiotherapy for carcinoma of the larynx.	
Add aspirin to clopidogrel	
Change to aspirin & dipyridamole instead of clopidogrel	
Referral to experienced vascular surgeon for consideration of carotid endarterectomy	
Referral to interventional radiologist experienced in carotid stenting surgeon	Best answer 4

Anticoagulation with heparin, then warfarin vascular	
Anticoagulation with warfarin immediately	
Referral to experienced vascular surgeon for consideration of carotid endarterectomy, with delayed post-operative warfarin.	
Start prednisolone 60mg and arrange temporal artery biopsy	
Continue present medication, without change or addition	
Anticoagulation with heparin, before referral to vascular surgeon	
Question 12	
<i>Female, 75 years old. Isolated mild hemiparetic stroke, clearing after 2 weeks, occurred 6 weeks before. 40% bilateral carotid artery disease. Two previous mild hemiparetic strokes (one each side) leaving no deficit over the last year. Normal rhythm and echo. MRI scan shows periventricular confluent high signal on T2 and 2 lacunes and 2 peripheral infarcts</i>	
Add aspirin to clopidogrel	
Change to aspirin & dipyridamole instead of clopidogrel	
Referral to experienced vascular surgeon for consideration of carotid endarterectomy	
Referral to interventional radiologist experienced in carotid stenting surgeon	
Anticoagulation with heparin, then warfarin vascular	
Anticoagulation with warfarin immediately	
Referral to experienced vascular surgeon for consideration of carotid endarterectomy, with delayed post-operative warfarin.	
Start prednisolone 60mg and arrange temporal artery biopsy	
Continue present medication, without change or addition	Best answer 9
Anticoagulation with heparin, before referral to vascular surgeon	

Answers and comments

1. Answer 3. With a clear-cut recent TIA and a 90% ipsilateral stenosis, this man is a clear-cut candidate for carotid endarterectomy according to both ECST and NASCET trials (1,2,3, 14).
2. Answer 9. Retinal events carry a much better prognosis than cerebral ones (1,4,5). The natural history is also better the younger the patient, whilst mortality from surgery is actually greater! The morbidity of carotid endarterectomy is greater in women than men (1). At 75%, the degree of stenosis is "borderline". Putting these factors together, carotid endarterectomy is not indicated in this lady. The graph in the final ECST paper (fig. 5) tries to summarise the effects of age, sex and carotid stenosis on this decision. See also ref. 14
3. Answer 9. The advisability of carotid endarterectomy in patients who have suffered just retinal events is debatable. The ECST figures suggest that it is rarely justified in trying to prevent stroke, whilst the NASCET figures would suggest that it is of value (4,5). However, the same ECST results do show that it is effective in reducing retinal stroke in these patients (5).
4. Answer 9. The natural history risk of a symptomatic carotid stenosis falls with time, especially for tight stenoses. Most of the risk in the first 6 to 12 months from the last (symptomatic) event. This is summarised in Fig. 3 of ECST (1). With "only" an 80% stenosis and no symptoms for 18 months, this patient probably has little to gain from surgery.
5. Answer 8. Anterior ischaemic optic neuropathy is almost always due to occlusion of multiple small arteries in the network supplying the optic nerve head. Such small vessel disease is either due to hypertension/diabetes or to arteritis. The former group will often have coincidental atheroma in carotid, coronary or peripheral arteries. Very seldom is AION demonstrated to be due to embolism. On the other hand a flurry of episodes of transient monocular blindness as occurred in this lady before the AION is quite often noted in cranial arteritis. This patient should be considered to have this disease until "proved" otherwise (very difficult in itself). Headache may or, sometimes, may not be present.

6. Answer 7. This patient has two potential embolic sources. It is impossible to know for sure which has caused her symptoms. However, transient monocular blindness tends to be associated with small emboli, probably platelet rich, which rapidly break up. On the other hand, emboli from fibrillating hearts tend to be larger and to cause large strokes. Therefore, it is far more likely that the carotid stenosis is the source of the emboli. The question then comes down to weighing up the relative risks of the natural history of a 75 year old man with a symptomatic 90% stenosis, giving rise to multiple events over 2 weeks, even if just retinal, against the risk from surgery. Assuming the cardiac problems are controlled and stable, carotid surgery would be indicated here. Following that, anticoagulation could be commenced for primary prophylaxis in a man of this age with previous but now controlled hypertension (if no contra-indications).

7. Answer 4. The carotid needs attention, but the patient's prosthetic aortic valve would make stopping anticoagulation risky. Some surgeons are willing to operate on anticoagulated patients, and perhaps even at the levels of anticoagulation used in such AVR patients. However, perhaps safer would be to treat the carotid by primary stenting. The CAVATAS trial showed that this can be achieved with reasonable safety. Indeed the procedural morbidity was about the same as endarterectomy, though both figures were considerably higher in that trial than in ECST, NASCET, and the every day practice of experienced surgeons.

8. Answer 9, or perhaps even better randomise to the ESPRIT trial. Carotid surgery is not appropriate here (1,2,3). In the absence of a strong indication of embolism from the heart, there is no indication for warfarin. This is definitely contra-indicated at high levels of anti-coagulation (6) and is of no additional benefit at lowish levels (7). The ESPRIT trial continues, and hopefully should answer this question once and for all. It also compares the aspirin & dipyridamole combination with aspirin alone, re-investigating this question which was also addressed by the ESPS2 trial (8), but not to everyone's satisfaction (9). There are no comparative studies between clopidogrel and the combination of aspirin & dipyridamole (option 2), and the MATCH trial concerning the combination of clopidogrel & aspirin in cerebrovascular disease has yet to report.

9. Answer C. Whilst the operative morbidity is greater in the elderly, the natural history risk increases even more so with age, and analysis of the NASCET data (11) shows that the elderly have more to gain from carotid endarterectomy than the younger patients.

10. Answer 9. We have no idea when these asymptomatic emboli arose and this carotid stenosis should be regarded as asymptomatic. In the UK, asymptomatic stenoses are usually not operated on because the natural history risk of stroke is low (5.7 % in 3 years in severe stenosis, ref.12). There is an on-going trial (ACST) which is addressing this issue, visited initially in the USA in the ACAS trial (12).

11. Answer 4. Previous radiotherapy may accelerate atherogenesis, and one often sees severe bilateral disease in these symptomatic patients. Furthermore surgeons do not like operating on these, because the tissue planes are often obliterated and surgical manipulation of the artery is far more difficult (and therefore thought to predispose to throwing off emboli before any clamp can be applied). Probably an indication to suggest stenting instead.

12. Answer 9. This sort of patient with confluent white matter ischaemic changes had a particularly high intracerebral haemorrhage rate in the SPIRIT trial (6): not for anticoagulation!

References:

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2. North American Symptomatic Carotid Endarterectomy Trial Collaborators. Beneficial effect of carotid endarterectomy in symptomatic patients with high grade stenosis. *NEJM* 1991; 325: 445-53
3. Barnett et al for the North American Symptomatic Carotid Endarterectomy Trial Collaborators. Benefit carotid endarterectomy in patients with symptomatic moderate or severe stenosis. *NEJM* 1998; 339: 1415-25

4. Streifler et al for the North American Symptomatic Carotid Endarterectomy Trial Collaborators. The risk of stroke in patients with first-ever retinal vs hemispheric transient ischemic attacks and high grade stenosis. *Arch. Neurol* 1995; 52: 246-249
5. Rothwell et al on behalf of European Carotid Surgery Trialists' Collaborative Group. Ocular versus cerebral ischaemic events: different disorders or a matter of chance. *JNNP* 1998; 64: 700-1
6. Gorter et al. Major bleeding during anticoagulation after cerebral ischemia: patterns and risk factors: Stroke Prevention in Reversible Ischemia Trial (SPIRIT). *Neurology* 1999; 53: 1319-27
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8. Diener et al. European Stroke Prevention Study. Dipyridamole and acetylsalicylic acid in the secondary prevention of stroke. *J. Neurol. Sci.* 1996; 143: 1-13
9. Antithrombotic Trialists' Collaboration. Collaborative meta-analysis of randomised trials of antiplatelet therapy for prevention of death, myocardial infarction and stroke in high risk patients. *BMJ* 2002; 324: 71-86
10. Eliazasziw et al for the North American Symptomatic Carotid Endarterectomy Trial. Significance of plaque ulceration in symptomatic patients with high-grade stenosis. *Stroke* 1994; 25: 304-08.
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12. European Carotid Surgery Trialists' Collaborative Group. Risk of stroke in the distribution of an asymptomatic carotid artery. *Lancet* 1995; 345: 209-12
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3 Headache and acute neurological deficits		Question setter: Dr TP Enevoldson (peter@enevoldson.freereserve.co.uk)
Below there are five brief case histories, and a list of possible diagnoses. Each may be used on more than one occasion.		
Question 1		
A previously well 45 year old right handed man abruptly developed the worst ever headache he had ever experienced. He was taken to hospital where examination revealed a distressed and ill patient with no abnormal signs. Unenhanced and then contrast enhanced CT brain scans were normal (7 hours after onset). Lumbar puncture revealed a pressure of 26 cm of water and 300 red cells but no other abnormalities. Over the next 24 hours his conscious level slowly fell (GCS 11). He had a seizure which commenced in his right arm before becoming generalised. This was repeated twice in the next hour. On recovery from these, he was noted to be dysphasic and to have a right hemiparesis. Repeat CT scanning revealed a generally swollen brain, with particular sulcal effacement over the left parietal region. Here there was an obvious superficial haemorrhage, surrounded by a much larger area of low attenuation. In addition, there was probably a tiny sliver of haemorrhage very superficially in the right parietal region. Administration of contrast did not reveal any other abnormalities.		
Pontine haemorrhage		
Vertebral artery dissection		
Carotid artery dissection		
Cerebral venous sinus thrombosis		Best answer 4
Subarachnoid haemorrhage		
Occipital lobe haemorrhage		
Pituitary apoplexy		
Cerebellar haemorrhage		
Brainstem infarct		
Chronic non-bacterial meningitis		
Acute bacterial meningitis		
Question 2		
A 58 year old man, with a history of incompletely treated hypertension, developed a severe occipital headache. This worsened further over the next 15 minutes and he became unable to stand. He developed rotatory vertigo and started to vomit. Workmates took him to A & E, where between the patient's retching, the Casualty Officer noted marked nystagmus to the right, but no cranial nerve problems, or obvious limb weakness, numbness or reflex abnormalities. BP was 170/100. A diagnosis of viral labyrinthitis was entertained and a Stemetil injection given. Over the next hour or so, the student nurse noted that he seemed to be settling since was not complaining so much, though he seemed to be sleepy and his speech was now slurred. On checking the patient an hour later, the Casualty Officer was surprised to find that the patient was now unrousable, and that his eyes were deviated to the left and his pupils were tiny. His plantar responses were now extensor. BP was now 200/130, and the respiration rate was irregular. He secured the patient's airway, and bleeped the on-call neurology registrar.		
Pontine haemorrhage		
Vertebral artery dissection		
Carotid artery dissection		
Cerebral venous sinus thrombosis		
Subarachnoid haemorrhage		
Occipital lobe haemorrhage		
Pituitary apoplexy		
Cerebellar haemorrhage		Best answer 8
Brainstem infarct		
Chronic non-bacterial meningitis		
Acute bacterial meningitis		
Question 3		
Your 45 year old consultant decided to re-live his glory days as a prop forward and played in the annual rugby match against the medical students. He realised his mistake at the first scrum, and was painfully reminded of		

<p>this at each one thereafter. The next morning he was uncertain whether to blame his headache and neck pain on the game or the evening after. The former settled over the next day, despite his out-patient clinic, but his neck seemed to become progressively worse. Two days later on his ward round, he was like the proverbial bear with a sore head (or neck), when he abruptly suffered severe vertigo and started to vomit and stagger to the right. As he was trying to make his excuses to Sister, he was aware his speech was slurred. The right side of his face began to tingle, and then felt numb. The left sided limbs and trunk felt cold and numb. Your respectful examination in addition reveals rotatory nystagmus, a palatal deviation to the left (testing palatal sensation is impossible because he politely suggests somewhere else to put your tongue depressor), no limb weakness or reflex abnormality, but a loss of pin prick appreciation in the right side of his face and left hand and foot. His right arm seems clumsy.</p>	
Pontine haemorrhage	
Vertebral artery dissection	Best answer 2
Carotid artery dissection	
Cerebral venous sinus thrombosis	
Subarachnoid haemorrhage	
Occipital lobe haemorrhage	
Pituitary apoplexy	
Cerebellar haemorrhage	
Brainstem infarct	
Chronic non-bacterial meningitis	
Acute bacterial meningitis	
Question 4	
<p>An 18 year old girl had suffered classical migraine with visual aura for 4 years when she developed her worst ever headache . The left sided headache was above the left eye and more intense than ever before. It persisted like this the next 2 days, most uncharacteristically. Her mother noticed her left eyelid was drooping slightly. She then abruptly developed a right hemiparesis and expressive dysphasia, and at this stage was taken to A & E. You find a left partial ptosis, that the left pupil is smaller than the right (especially in dim light), and confirm the right flaccid hemiparesis with brisk reflexes and an extensor plantar..</p>	
Pontine haemorrhage	
Vertebral artery dissection	
Carotid artery dissection	Best answer 3
Cerebral venous sinus thrombosis	
Subarachnoid haemorrhage	
Occipital lobe haemorrhage	
Pituitary apoplexy	
Cerebellar haemorrhage	
Brainstem infarct	
Chronic non-bacterial meningitis	
Acute bacterial meningitis	
Question 5	
<p>A 51 year old hypertensive man developed pain in the right temple and behind the ear. He had never suffered from headaches in the past. His voice had changed. On examination, right IXth and Xth nerve lesions were evident, and also a right Horner's. A CT scan was normal. Two days later, he developed a transient loss of vision 'in the right eye' for ten minutes, followed an hour later by a left hemiparesis and hemihypaesthesia, including the face.</p>	
Pontine haemorrhage	
Vertebral artery dissection	
Carotid artery dissection	Best answer 3
Cerebral venous sinus thrombosis	
Subarachnoid haemorrhage	
Occipital lobe haemorrhage	
Pituitary apoplexy	
Cerebellar haemorrhage	
Brainstem infarct	

Chronic non-bacterial meningitis	
Acute bacterial meningitis	
Question 6	
A sixty year old portly diabetic hypertensive man developed a severe pain around his left eye, and became aware something the matter with his vision. He was unable to read, apparently with either eye. He presented to A & E, c forehead and fully alert. On examination, his optic discs are normal, and he has a congruous right homonymous h not sparing the macula. He can count fingers easily using each eye but testing visual acuity with a Snellen chart is He can describe the details of the equipment on the opposite wall of the department quite well, both verbally and There are no other neurological signs.	
Pontine haemorrhage	
Vertebral artery dissection	
Carotid artery dissection	
Cerebral venous sinus thrombosis	
Subarachnoid haemorrhage	
Occipital lobe haemorrhage	Best answer 6
Pituitary apoplexy	
Cerebellar haemorrhage	
Brainstem infarct	
Chronic non-bacterial meningitis	
Acute bacterial meningitis	

Answers and comments

Question 1: 4 A sub-arachnoid like onset to the headache is well recognised in CVST, though not common. The normal initial imaging, and the LP findings are quite typical. The subsequent seizures with focal onset followed by a focal deficit announce the involvement of cortical veins and venous infarction. The CT appearances are classical. The empty delta sign on the enhanced CT is specific but only seen in about 30%.

Question 2: 8 Rapid onset cerebellar features with subsequent pontine signs. The initial story should always suggest cerebellar haemorrhage until proven otherwise, if not least because urgent surgery may be required.

Question 3: 2 A classical dorsolateral medullary syndrome, probably with cerebellar involvement, due to a posterior inferior cerebellar artery occlusion (according to classical teaching). At this age with the preceding neck stiffness, after the preceding trauma, a vertebral artery dissection is virtually certain.

Question 4: 3 Classical spontaneous carotid artery dissection, with delayed secondary embolism. The pain and the Horner's give the diagnosis.

Question 5: 3 The cranial nerve signs are 'false-localising', and are caused by compression of the nerves just outside the skull base by the expanded carotid (Mokri et al Arch., Otolaryngol. Haed Surg 1992; 118:431-435). (Higher cranial nerves may also be affected, e.g. in the cavernous sinus, perhaps by interference with their blood supply Scievink et al Neurology 1993; 43: 1938-1941)

Question 6: 6 The pain from occipital lesions are frequently referred to the ipsilateral eye. This man has a right homonymous hemianopia, and alexia without agraphia, due to extension to the splenium. His acuity is quite good apparently and he is fully alert, arguing against a major pituitary apoplexy compressing his left optic tract.

4 Movement disorders	Question setter: Dr S Fox (shfox@hotmail.com)
<p>A 59 year old lady with Parkinson's disease for 8 years is referred to the movement disorders clinic for consideration of surgery. She was initially treated with Sinemet. After an initial good response she developed wearing off symptoms, with stiffness and freezing. She was then commenced on Pergolide, which settled her symptoms. The dose was increased up to 5 mg but she developed poorly-formed visual hallucinations. The dose was reduced to 3 mg and the hallucinations settled with no recurrence. She had no other cognitive problems. The dose of sinemet was increased further to a total 800mg/day with entacapone 200mg with each dose. Over the past few months she has started to notice right sided involuntary movements occurring 2-3 h after each dose of sinemet, which at times prevented her from eating and interfere with walking. She has also developed increasing stiffness in her legs and freezing. She often switches off suddenly and unpredictably. She has difficulty getting out of bed at night with pain in her feet in the morning. She lives with her husband who is finding it increasingly difficult to look after her.</p>	
Question 1	
Choose the most appropriate of the following options for this patient:-	
Advise bilateral subthalamic nucleus stimulation deep brain stimulation (STN-DBS)	
Advise bilateral Pallidotomy	
Refer for neuropsychometry and review with results for consideration of STN-DBS	Best answer 3
Refer for neuropsychometry and review with results for consideration of Pallidotomy	
Advise thalamotomy	
Question 2	
Choose the most appropriate of the following options for this patient:-	
Add Sinemet CR for nocturnal symptoms and reduce daytime sinemet dose to reduce dyskinesia.	
Add Cabergoline for nocturnal symptoms and reduce daytime sinemet dose to reduce dyskinesia	
Use Apomorphine	Best answer 3
Add benzhexol to improve parkinsonism and tremor	

Answers and comments

Answers 3 and 3

She has marked levodopa-induced motor complications, including on-off fluctuations and dyskinesia. Continuing with oral medication will probably be unsuccessful and she previously developed hallucination on higher doses of dopamine agonists. The two options therefore include apomorphine or surgery. Apomorphine is an effective treatment for motor complications, either intermittent sc or infusion pump.

Surgical targets for PD include thalamus, medial globus pallidus (pallidotomy) or subthalamic nucleus. Thalamic Lesioning or DBS will reduce tremor but with no effect on other parkinsonian symptoms. Pallidotomy is particularly effective for severe unilateral dyskinesias. There is also a good effect on tremor, bradykinesia and rigidity but bilateral surgery can cause dysarthria and cognitive problems. The current preferred target is the STN as bilateral surgery can be performed with good effects on all parkinsonian symptoms. Antiparkinsonian medication can be cut by 50%, with a reduction in dyskinesia. Patient selection for surgery is important. Patients who do best are young, have a good response to standard levodopa challenge and have no evidence of cognitive impairment. Cognitive decline in elderly patients (> 70years) has been reported, following surgery. This patient had hallucinations induced by high dose Pergolide, and although there are no reported cognitive problems, she is at risk of further hallucinations and neuropsychometry is required to ensure there is no evidence of cognitive impairment.

At present it is not known if bilateral STNDBS is any more effective than apomorphine infusion for long-term management of PD. The ongoing PDSURG trial will hopefully answer this question.

5 Epilepsy	Question setter: Dr G Fuller (Geraint@fullerg.demon.co.uk)
<p>A 22 year old woman is referred urgently to out-patients by her GP with seizures which are increasing in frequency. She attends your clinic in early May. She had a seizure on New Years day which had been witnessed by her boyfriend. She recalls feeling slightly dizzy beforehand and then nothing till she arrived in hospital. Her boyfriend gave a good description on a tonic-clonic seizure. She had then had a second seizure in March; this occurred in the early morning (soon after getting up) having been out to a nightclub and drinking alcohol the night before. She had again attended A&E, was advised not to drive, started on carbamazepine 100mg twice a day and was referred to the neurology department. In mid April she had two further daytime seizures with tongue biting; her dose of carbamazepine was increased to 200 mg twice a day. In the last week in April she had 2 further tonic-clonic seizures preceded by a brief feeling of dizziness. She had felt very tired and headachy for the following day.</p> <p>She has no other symptoms. There is no family history of epilepsy. She has never had any other blackouts and did not have any peri-natal problems or a febrile convulsion. She denies having any funny feelings such as déjà vu, tastes or smells; there have been no nocturnal fits. Her boyfriend has not noticed anything else. She is on the combined oral contraceptive.</p> <p>Neurological examination is normal.</p>	
Question 1	
What is the most appropriate diagnosis?	
"Primary generalised epilepsy" – nature unspecified	
Juvenile myoclonic epilepsy	Best answer 2
Focal onset epilepsy	
Symptomatic focal onset epilepsy	
Cryptogenic focal onset epilepsy	
Cardiogenic reflex anoxic seizures	
Non epileptic attack disorder	
Question 2	
How could you confirm the diagnosis?	
MRI	
EEG	Best answer 2
72 hour ambulatory EEG monitoring	
ECG	
ECHO	
24 hour ECG monitoring	
Blood tests	
Clinical psychology assessment	
None of the above	
Question 3	
How would you treat her? Select the most appropriate measure	
Increase dose of carbamazepine	
Start sodium valproate	
Start lamotrigine	Best answer 3
Start phenytoin	
Start levetiracetam	
Start clonazepam	
Start topiramate	
Start clobazam	
Stop carbamazepine and try oxcarbazepine	
Advice about contraceptive pill interaction	

Answers and comments

This woman has had a series of tonic clonic seizures with non-specific warning. These probably represent the onset of a generalised seizure disorder. She has been started on carbamazepine and the frequency of the seizures has increased. This is very suggestive of a diagnosis of juvenile myoclonic epilepsy (JME). This diagnosis accounts for about 5-10% of patients with epilepsy and the main reasons for failing to make the diagnosis is failure to elicit a history of myoclonic jerks (Montalenti et al 2001), something which is often not thought of as being related by the patient. The diagnosis is usually made on the basis of the history; in the untreated EEG often demonstrates 3-6 Hz spike/polyspike and wave but in about third of patients demonstrates asymmetries that could be interpreted as indicating a focal onset disorder. Patients with JME usually respond well to sodium valproate or lamotrigine but their seizures can be aggravated by carbamazepine or phenytoin. In one study 68% of patients prescribed carbamazepine and 38% of patients prescribed phenytoin experience aggravation of seizures (Genton et al 2000).

There is a high risk of recurrence if treatment is stopped so therapy needs to be life long. In a woman of this age, lamotrigine is a better initial option than valproate because of the higher teratogenic potential of the latter.

6 Multiple Sclerosis		Question setter: Dr J Palace
You see a woman of 45yr old with clinically definite MS, who has a recent history of relapses. She asks if she can have interferon-beta. You have funding to prescribe according to the ABN guidelines and for the purpose of answering this question you need to do the following. Please answer true or false to each of the following statements/questions.		
Question 1		
Establish the exact nature of all her symptoms in the last two years.		False
Question 2		
Ensure that at least two relapses were severely disabling.		False
Question 3		
Ensure she is not planning to become pregnant while taking the drug.		True
Question 4		
Ensure that in the presence of severe relapses there is no mild 'hidden' background progression.		False
Question 5		
Establish if she suffers from significant depression and if so explain that it is contraindicated in her case.		False
Question 6		
As long as she can walk 10m with aid and fulfils the relapse criteria she can still have the drug.		True
Question 7		
Ensure that she is able to self-inject.		False
Question 8		
You have decided she fulfils the criteria and now need to run through other details about this treatment which include:		
Treatment will on average prevent about one third of relapses.		True
Question 9		
The drug will need to be stopped if she loses the ability to walk 100m unaided (it having been shown to be of no benefit beyond this level of disability).		False

Answers and comments

Question 1: **False**. just for two significant relapses within this time.

Question 2: **False**. significant is not generally thought to mean severe, but to describe troublesome relapses that are worth preventing.

Question 3: **True** according to current advice and information.

Question 4: **False** in secondary progressive MS beta-interferon appear to reduce relapses in the same way as in relapsing remitting MS and thus is useful if the relapses are a significant contributor to the disability suffered.

Question 5: **False** In one study there were more suicides in the treated group but this was not borne out in the many subsequent studies. However severe depression is a caution and patients should be warned about it.

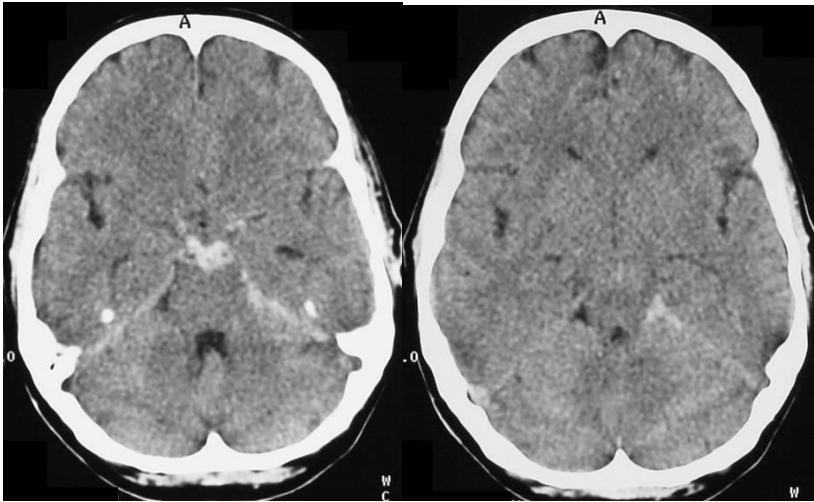
Question 6: **True** as this was the disability cut off used in the secondary progressive trials in which an effect was seen on relapses.

Question 7: **False** Avonex is given once weekly and can be given by a district nurse. Additionally other members of the family can give treatment.

Question 8: **True**

Question 9: **False**: stopping criteria depends on the frequency of relapses and the presence of significant progressive disease or until unable to walk. Not the loss of ability to walk 100m unaided.

7 Headache	Question setter Dr CJ Lueck (cl@skull.dcn.ed.ac.uk)
A 37 year old, right handed woman without previous medical history was gardening when she experienced the sudden onset of severe occipital and nuchal headache, and collapsed with the pain. On arrival in hospital some three hours later, she was found to be drowsy, with a Glasgow coma score of 14/15. She was afebrile and had a markedly stiff neck and a positive Kernig's sign. Neurological examination was otherwise normal.	
A CT scan was organised as an emergency and performed within the hour. The findings of the uncontrasted study are shown in the figure below.	
Question 1	
Which of the following is the most likely diagnosis?	
acute cerebellar haemorrhage	
acute hydrocephalus	
acute meningococcal meningitis	
herpes simplex encephalitis	
perimesencephalic haemorrhage	Best answer 5
ruptured basilar artery aneurysm	
ruptured middle cerebral artery aneurysm	
subdural haematoma	
pituitary apoplexy	
Question 2	
Which is the next most appropriate investigation?	
4-vessel catheter angiogram	Best answer 1
blood culture	
CT angiogram	
Doppler ultrasound of carotids	
lumbar puncture	
MRI scan	
MR angiogram	
no further investigation	
herpes simplex PCR on CSF	



Answers and comments

Answer: 5. Perimesencephalic haemorrhage. This CT scan is uncontrasted, and the increased density seen in the prepontine cistern and along the tentorium is a typical finding, representing subarachnoid blood in the perimesencephalic CSF space. Aneurysmal subarachnoid haemorrhage is not impossible, simply somewhat less likely (see below). The other possible answers are incorrect.

Answer: 1. 4-vessel angiogram. In a perimesencephalic haemorrhage, it is likely that there will not be an associated aneurysm. However, it is possible for aneurysmal rupture to give an appearance such as this, and there is always the possibility that there may be multiple aneurysms. The important point is that if the angiogram is negative, a further angiogram is unnecessary (in other types of subarachnoid haemorrhage a normal 4-vessel angiogram is usually followed by a repeat angiogram several weeks later in case local spasm has prevented detection of an aneurysm). CT and MR angiography are as yet not sensitive enough to detect small aneurysms which might have been responsible for this bleed. Lumbar puncture is obviously unnecessary as the diagnosis of haemorrhage has been made by the scan. The other tests are unlikely to be helpful. The prognosis of perimesencephalic subarachnoid haemorrhage is much better than that of aneurysmal subarachnoid haemorrhage. CT angiography (C) may be reasonable in some selected centres.

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8 Epilepsy again	Question setter Dr M Manford
<p>A sixty-two year-old female inpatient is referred to you from the general physicians in your DGH. She has a history of epilepsy of uncertain type dating back to adolescence, which has been well-controlled with phenobarbitone 90mg daily. She was admitted after a seizure; she has remained confused over the following four days. She has been unable to eat or drink and has been on IV fluids. During this time she suffered a further tonic clonic seizure, following which she was drowsy but then briefly was more lucid before returning to her previous state of confusion. When you go to see her she is confused with a glazed expression and at times her eyelids appear to flutter. She is afebrile with no neck stiffness and no focal neurological signs. The physicians arranged a CT scan, which was normal as were routine blood tests including blood count, U&E, glucose, calcium and LFTs.</p>	
Question1	
<p>What is the likely cause of this clinical picture?</p>	

A superadded metabolic encephalopathy	
Superadded cerebrovascular event	
Drug overdose	
Drug withdrawal	Best answer 4
Encephalitis	
Question 2	
What is the likely epilepsy syndrome?	
Focal epilepsy	
Generalized epilepsy	Best answer 2
Question 3	
What further investigation would you consider?	
Urgent AED blood levels	Best answer 1
CSF analysis	
Serum ammonia	
Transfer for EEG	
Question 4	
How would you treat this deterioration?	
IV benzodiazepine then titrate carbamazepine	
IV benzodiazepine then titrate lamotrigine	
IV benzodiazepine then loading dose valproate	
IV phenytoin	
IV phenobarbitone	Best answer 5

Answers and comments

This lady presents with non-convulsive status epilepticus. The eyelid fluttering and the improvement after a convulsive seizure suggest that this is due to generalized epilepsy.

The likely cause is drug withdrawal. Urgent phenobarbitone levels should be requested. An EEG would be helpful but treatment should not be delayed until it is available. The right treatment is to give a loading dose of phenobarbitone. Giving a benzodiazepine may transiently stop status but it is likely to recur in the context of antiepileptic drug withdrawal. Giving an alternative maintenance antiepileptic drug is unlikely to maintain remission from drug withdrawal from phenobarbitone. Further investigations would only be needed if there was not a good response to initial treatment.

9 Movement Disorders again		Question setter Dr NA Fletcher (nick.fletcher@thewaltoncentre.nhs.uk)
A 65 year old man presents at a DGH satellite neurology clinic with a 12 month history of a mild tremor in the right hand. There has been some reduction of dexterity in the hand for coins and buttons and he has become slightly stooped according to his wife. There is no relevant past medical or family history. On examination there is slight reduction of facial expression, a resting tremor in the right hand, mild cogwheel rigidity at the right wrist (detectable only with contralateral synkinesis) and slowing of finger movements in the right hand. When he walks there is a mild reduction of arm swing on the right. He is not disabled by any of his symptoms and is faintly reluctant to take medication.		
Question 1		
DIAGNOSIS: which of the following is most correct?		
The asymmetry of the signs is atypical and a CT scan is indicated to exclude a structural lesion.		
The probability of Parkinson's disease is at least 90%.		Best answer 2
Multiple system atrophy is excluded by the presence of the asymmetrical resting tremor.		
No diagnosis is possible until his response to medication has been assessed.		
It is important to exclude Wilson's disease with blood tests and a slit-lamp examination		
Question 2		
TREATMENT AND PROGNOSIS: which of the following is most correct?		
It is advisable to start treatment at this stage to provide neuroprotection and retard		

further disease progression.	
It is not necessary to take any medication at this stage.	Best answer 2
Dopamine agonist monotherapy and levodopa are equally effective symptomatic treatments in the first 5 years of Parkinson's disease.	
If the patient starts ropinirole, he will probably be able to avoid levodopa for up to 5 years.	
The risk of dementia in Parkinson's disease is over 50%.	
A good response to levodopa excludes multiple system atrophy	
A poor response to levodopa excludes Parkinson's disease	
A poor response to ropinirole monotherapy suggests multiple system atrophy	
Question 3	
PATHOLOGY: The patient dies after a fatal road traffic accident three years later. It is believed that he fell asleep at the wheel. He had been taking ropinirole and sinemet. He had donated his brain for neuropathological study in an advance directive. His family wish to discuss the findings with reference to the diagnosis of his parkinsonism. The salient findings were cell loss and gliosis in the substantia nigra, striatum, dentate and some brainstem nuclei. There are associated glial and neuronal cytoplasmic inclusion bodies which stain positively for alpha synuclein.	
Which of the following is most correct?	
The patient did not have idiopathic Parkinson's disease	Best answer 1
His children require referral for genetic counselling as they are at risk for autosomal dominant Parkinson's disease	
The correct diagnosis was dentatorubropallidoluysian atrophy (DRPLA)	
It is surprising that there were no cerebellar signs in life	
His fatal accident was unlikely to have been due to his medication.	

Answers and comments

1: answer = 2

- False.** Asymmetry of signs is typical of Parkinson's disease
- True.** The most recent estimate of diagnostic accuracy in a non-subspecialist setting is 90% ¹.
- False.** Resting tremor and asymmetry of onset are seen in MSA ².
- False.** It is reasonable to give a diagnosis of probable Parkinson's disease. To withhold any diagnosis until a response to treatment is observed (as occasionally occurs) is overstating the small diagnostic uncertainty. In non-subspecialist clinics, at least 90% of patients with this sort of presentation have idiopathic Parkinson's disease. The situation is different with atypical cases seen in tertiary movement disorders clinics ³ but this is not the case here. Some patients with straightforward Parkinson's disease are greatly unsettled by excessive diagnostic equivocation. It is appropriate to mention the small chance of an alternative parkinsonian disorder.
- False.** Wilson's disease is unlikely to present at this age.

2: answer = 2

- False.** Neuroprotective effects of selegiline and dopamine agonists in Parkinson's disease are unproven and speculative. While there is certainly a possibility of such effects, it cannot be argued that the evidence makes the use of such agents "advisable" at this stage of the illness – see ⁴.
- True.** The patient is not keen and there is no need to start symptomatic treatment until the patient's condition requires this ⁵

3. **False.** Levodopa is a superior symptomatic treatment – see ^{5 6}. Patients must be made aware of this if they are to make an informed choice about early therapy.
4. **False.** A key study is the ten year follow up of bromocriptine monotherapy versus levodopa ⁶. This showed a small reduction in dyskinesia at 10 years but not for moderate or severe dyskinesia. Other studies have not followed patients for 10 years and do not all distinguish between “motor complications”, “fluctuations” and “dyskinesia”.
5. **False.** The ropinirole monotherapy 056 study is helpful when advising patients in the clinic. At 5 years, only 47% of patients were still taking ropinirole; of these, only 34% did not need supplemental levodopa - see ⁷.
6. **False.** Dementia is seen in 25-30% of patients. The pathology is usually Lewy body dementia, Alzheimer’s disease or a mixed pathology ^{2 5}.
7. **False.** Although a poor response to levodopa should raise doubts about a diagnosis of Parkinson’s disease, and is more suggestive of MSA, some MSA patients have a good initial levodopa response ⁸. Similarly, some Parkinson’s disease patients have a poor response to levodopa. In an important recent study, a poor initial levodopa response was more common in MSA (58%), but it also occurred in some (23%) patients with PD ⁸.
8. **False.** See above.
9. **False.** Many patients with Parkinson’s disease will not respond well to dopamine agonist monotherapy and will therefore need rescue levodopa ^{4 7}.

3: answer = 1

1. **True.** These are the pathological features of MSA.
2. **False.** MSA is not hereditary and the presence of alpha synuclein in the cellular inclusions does not indicate autosomal dominant parkinson’s disease due to mutations of the alpha synuclein gene on chromosome 4q21.
3. **False.** Clinically and pathologically this is not DRPLA
4. **False.** Only about 50% of MSA patients develop clinically evident cerebellar involvement and this is an early feature in only about 20% ^{4 5 7 9}.
5. **False.** Ropinirole and pramipexole (as well as other anti-parkinsonism drugs) have been associated with sedation and sudden onset of sleep ¹⁰

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10 Yet more movement disorders	
A 70 year old man attends for a second opinion. He presents with falling and unsteadiness. Usually he falls backwards and this has been developing for nine months. He feels that his vision is not quite right. He is a smoker and there has been mild hypertension treated with bendrofluazide. There is no history of alcohol abuse, dietary deficiency or other medication. There is no relevant family history. On examination there is no cognitive impairment. His gait is slightly slow, on a narrow base. He has axial rigidity; the neck is stiff and there is a staring facial expression with frontalis overactivity. You think the voluntary and pursuit eye movements are normal, as are visual acuities, fields and fundi. He is slightly dysarthric; there is no limb rigidity or tremor; reflexes are normal and the plantar responses are flexor. BP is 130/80. The first neurologist has arranged a MRI brain scan, which is normal. The patient has been told he probably has progressive supranuclear palsy (Steele-Richardson-Olzewski disease).	
Question 1	
Which of the following is most correct?	
Progressive supranuclear palsy (Steele-Richardson-Olzewski disease) is unlikely with normal eye movements	
A cerebellar degeneration is a more likely diagnosis	
A paraneoplastic aetiology should be considered and investigations for an underlying malignancy are indicated	
The lack of cognitive impairment is consistent with PSP	Best answer 4
Parkinson's disease cannot be excluded here and a trial of levodopa should be completed before deciding on the likely diagnosis.	
If this is PSP, the plantar responses should be extensor.	
Question 2	
Two years later the patient is worse. He is very unsteady and unable to walk without assistance; there have been many falls. There is marked urinary frequency and urgency. The voluntary eye movements are slow and 50% restricted in all directions but pursuit movements are full, including downgaze. The consultant demonstrates a curious physical sign; if the patient is asked to look at an optokinetic nystagmus (OKN) drum, there is a sustained deviation of the eyes but no OKN. The neck is very rigid and extended.	
Which of the following is most correct?	
The urinary symptoms indicate that the patient probably has multiple system atrophy	
PSP is unlikely with preserved downgaze during testing of pursuit eye movements	
The loss of OKN indicates that the patient probably has a cerebellar degeneration rather than PSP	
The eye movement disorder described could occur in Parkinson's disease	
The neck rigidity suggests multiple system atrophy as the likely diagnosis	
The clinical picture is entirely consistent with PSP	Best answer 6

Answers and comments

Question 1

1. **False.** The eye movements may be normal in the early stages of the disease; indeed the characteristic eye movement disorder may not appear until very late or sometimes not at all¹⁻³.
2. **False.** A cerebellar degeneration is unlikely without other cerebellar signs; the gait is narrow based which is unlikely to be a cerebellar deficit. It is certainly true that parkinsonism, ophthalmoparesis and pyramidal signs may occur in a cerebellar degeneration (sometimes causing confusion with early PSP) but the combination of backwards falling, axial rigidity and staring appearance in a man of this age is more consistent with PSP.
3. **False.** Paraneoplastic syndromes are usually severe and subacute, especially paraneoplastic ataxia.
4. **True.** Cognitive impairment, although common in PSP often occurs late in the illness³.
5. **False.** Parkinson's disease is very unlikely with falling as a presenting feature; the patient does not have two out of the three cardinal features of bradykinesia, rigidity, tremor and postural instability. See PD brain bank criteria for the diagnosis of Parkinson's disease⁴.
6. **False.** Extensor plantars are a common finding in PSP, usually with brisk reflexes although severe spasticity is unusual. However, the plantars may be flexor in an early case. The point is that the "full house" of signs need not be present at this stage and the diagnosis cannot be rejected on this basis.

Question 2

1. **False.** Urinary symptoms are common in advanced PSP^{3,5}. It is the presence of severe and early autonomic dysfunction that is suggestive of multiple system atrophy.
2. **False.** Loss of pursuit eye movements is late and may occur after the loss of voluntary eye movements, which are impaired earlier. It is an error only to test pursuit in the clinic, as a loss of voluntary downgaze will be missed.
3. **False.** The OKN abnormality is typical of PSP³.
4. **False.** These eye movements are not consistent with Parkinson's disease.
5. **False.** Cervical rigidity is a key feature of multiple system atrophy⁶ but this is typically severe antecollis (flexion) not extension which is more typical of PSP.
6. **True.** See review in the recent JNNP supplement already referred to³.

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