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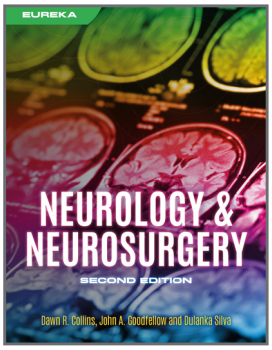

Self-assessment questions and answers

to accompany *Eureka: Neurology & Neurosurgery*, second edition

www.scionpublishing.com/eureka-neurology

SBA questions

Raised intracranial pressure and head injury

1. A 24-year-old man is hit by a car during a road traffic accident. He is unconscious with bleeding from an open scalp laceration. He has a compound open fracture in his left leg.
What is the single most appropriate next action?

A Call the on-call neurosurgeon for a review
B Immediate neurological examination, including GCS
C Perform primary assessment of the patient's ABCDE as per ATLS protocols
D Prescribe a dose of mannitol (0.5 g/kg)
E Urgent CT brain scan

2. A 40-year-old woman is assaulted while walking home from work. She has a GCS score of 8 (E1, V2, M5). Her right pupil is dilated and unresponsive to light. She is resuscitated, stabilised and transferred for an emergency CT brain scan. Her CT shows a right-sided, crescent-shaped hyper-dense collection.
What is the single most appropriate statement regarding her condition?

A Mild extradural haematoma
B Mild chronic subdural haematoma
C Severe acute subdural haematoma
D Severe chronic subdural haematoma
E Severe extradural haematoma

3. A 30-year-old woman has hit her head on the pavement following a fall. She has a short period of loss of consciousness after which she is well with a GCS score of 15. Two hours later she starts vomiting and collapses.
What is the single most likely cause of these symptoms?

A Acute subdural haematoma
B Chronic subdural haematoma
C Intracerebral haematoma
D Extradural haematoma
E Hydrocephalus

4. A 56-year-old man is in the intensive care unit following a road traffic accident. His brain CT scan shows some contusions in both frontal lobes. The on-call neurosurgeon is concerned about raised intracranial pressure.
What is the single next best step?

A Decrease CO_2 level by increasing ventilation rate
B Increase CO_2 level by decreasing ventilation rate
C Increase core body temperature with a warming device
D Place him in the 'head down' position
E Start corticosteroids infusion

5. A 78-year-old man is drowsy. His GCS score is 10 (E3, V2, M5). He had a fall 3 weeks ago. He is on warfarin for atrial fibrillation. His brain CT scan shows a significant chronic subdural haematoma (CSDH). The neurosurgeon decides to operate.
What is the single best next step?

A Observe the patient
B Operate on the patient immediately
C Stop warfarin and observe the patient
D Stop warfarin, give 2 units of platelets and operate
E Stop warfarin, give vitamin K and clotting factors, and operate

Headache and pain syndromes

1. A 40-year-old man has developed unilateral retro-orbital stabbing pain. He has ipsilateral eye watering, nasal stuffiness and ptosis. The pain is very severe and he is restless and pacing around the ward.
What is the single best initial treatment plan?

A High-flow oxygen and oral triptan
B High-flow oxygen and subcutaneous triptan
C Paracetamol and NSAID
D Paracetamol, NSAID and antiemetic
E Paracetamol, NSAID and morphine

2. A 29-year-old woman has unilateral throbbing headaches associated with nausea and mild photophobia. They occur once per month around her period and she sleeps them off in a dark room. She has not tried any medications. She wants to get pregnant in the future.
Which is the single best initial treatment plan?

A 900 mg aspirin, 10 mg domperidone during acute attack, no prophylactic treatment
B 900 mg aspirin, 10 mg domperidone during acute attack, propranolol
C 900 mg aspirin, 10 mg domperidone during acute attack, topiramate
D 1000 mg paracetamol, 30 mg dihydrocodeine during acute attack, no prophylactic treatment
E 1000 mg paracetamol, 30 mg dihydrocodeine during acute attack, propranolol

3. A 32-year-old woman has a severe left-sided headache that came on suddenly and reached its peak within 10 seconds. She is apyrexial, vomiting and photophobic.
What is the single best initial investigation?

A CT brain with contrast
B Lumbar puncture
C MRI brain
D MRI brain with contrast
E Plain CT brain

4. A 63-year-old woman has new persistent dull headaches, worse in the morning and on coughing, over the last 2 months. She has a history of breast cancer.
Which is the single best management option?

A Reassure her
B Refer to breast surgeon
C Routine referral to neurology outpatient clinic
D Urgent referral to neurology headache clinic
E Start treatment with high-dose NSAIDs and domperidone

5. A 21-year-old overweight woman has 2 months of increasing frontal throbbing headaches. They are worse on lying down and coughing. She has markedly reduced peripheral vision. A CT head and CT venogram are normal. Lumbar puncture shows a very high opening pressure of 42 cm of water.
What is the single best initial action?

A Analgesia and outpatient review
B Insert lumbar-peritoneal (LP) shunt
C MRI to identify any malignancy
D Repeat lumbar punctures to reduce intracranial pressure
E Start acetazolamide

6. A 34-year-old pregnant woman with a recent diagnosis of sinusitis has a fever and severe right-sided headache which is worse on straining. She is vomiting and has had a generalised tonic–clonic seizure.
What is the single most appropriate initial management plan?

A Check for proteinuria and hypertension
B CT venogram
C Lumbar puncture and antibiotics
D MRI head
E Observe and refer to first seizure clinic

Seizures

1. A 26-year-old woman is out shopping with her family when they notice her left arm and leg shaking. This continues for 2 minutes after which it stops spontaneously. She has no recollection of the event.
Which is the single best description of her seizure type?

A Complex partial
B Complex partial with secondary generalisation
C Generalised
D Simple partial
E Status epilepticus

2. A 44-year-old man is referred for investigation of his seizures. He describes suddenly waking up in the night with a sense of fear. He then has a funny 'metallic' taste in his mouth and hears unknown voices talking to him. The symptoms stop within a few minutes.
What is the single most likely site of origin for this seizure?

A Frontal lobe
B Occipital lobe
C Parietal lobe
D Pituitary gland
E Temporal lobe

3. A 19-year-old man has a recent diagnosis of epilepsy. He has several episodes during the day where he seems to 'lose focus and forget where he is', stopping talking and staring into space. He promptly recovers in 10–15 seconds. His EEG shows 3 Hz symmetrical spike-and-wave complexes in all leads.
What is the single best description of his seizure type?

A Complex partial
B Complex partial with secondary generalisation
C Generalised
D Simple partial
E Status epilepticus

4. A 30-year-old woman is newly diagnosed with epilepsy and her neurologist wants to start her on an antiepileptic drug. She suffers from complex partial seizures. She is planning on becoming pregnant in the near future.
What is the single most appropriate antiepileptic medication?

A Carbamazepine
B Clobazam
C Lamotrigine
D Lamotrigine and levetiracetam
E Sodium valproate

5. A 21-year-old man is newly diagnosed with epilepsy. His neurologist is planning on starting an antiepileptic drug. He suffers from sudden shock-like involuntary jerking movements of his arms and legs.
What single drug is most likely to exacerbate this type of jerking and should be avoided?

A Carbamazepine
B Lamotrigine
C Levetiracetam
D Sodium valproate
E Topiramate

6. A 29-year-old woman has been in seizure persistently for 10 minutes without regaining consciousness. She has no history of seizures. She is resuscitated and placed in the recovery position. No drugs have been administered. The seizures continue.
What is the single most appropriate next step?

A Give carbamazepine
B Give IV lorazepam
C Start magnesium sulphate infusion
D Start phenytoin infusion
E Transfer to the intensive care unit for intubation and ventilation

Neurovascular disease

1. A 58-year-old woman develops acute-onset weakness in the right side of her face and right arm with slurring of speech. She is assessed within 90 minutes of symptom onset and deemed a candidate for thrombolysis.
What is the single most appropriate next action?

A Alteplase infusion (0.9 mg/kg dose)
B Aspirin 300 mg immediately
C CT brain
D MRI brain
E Therapeutic dose LMWH infusion (1.5 mg/kg dose)

2. A 65-year-old man has left arm and face weakness with left-sided homonymous hemianopia. There are no other neurological deficits. The stroke physician diagnoses a stroke.
What is the single best description of his stroke according to the Oxford classification?

A Lacunar stroke
B Partial anterior circulation stroke (PACS)
C Posterior circulation stroke
D Total anterior circulation stroke (TACS)
E Weber's syndrome

3. A 49-year-old man has vertigo and difficulty swallowing. He is ataxic on the right side with right-sided Horner's syndrome. He has loss of pain and temperature sensation on the right side of his face and the left side of his body.
What is the single most likely vessel affected?

A Anterior cerebral artery
B Anterior communicating artery
C Middle cerebral artery
D Posterior cerebral artery
E Posterior inferior cerebellar artery

4. A 30-year-old woman had a sudden onset headache while having sexual intercourse 1 hour ago. She describes it as the worst headache of her life. She is GCS 15 with neck stiffness and photophobia. Her CT brain is normal.
What is the single next most appropriate course of action?

A Give analgesia, discharge and advise to return if symptoms worsen
B Perform cerebral angiography (digital subtraction angiogram)
C Perform lumbar puncture 6–12 hours after onset of symptoms
D Prescribe immediate IV antibiotics
E Repeat CT brain in 2 hours and discharge if normal

5. A 21-year-old man has a subarachnoid haemorrhage. His GCS score is 13 (E3 V4 M6) with weakness in his left arm. A CT angiogram confirms the presence of a middle cerebral artery aneurysm with normal-sized ventricles and no haemorrhage within them. He suddenly becomes comatose (GCS 3, E1 V1 M1) with a fixed dilated right pupil.
What is the single most likely cause of his symptoms?

A Hydrocephalus
B Hyponatraemia
C Repeat rupture of aneurysm with repeat haemorrhage
D Seizures
E Vasospasm and delayed ischaemic neurologic deficit (DIND)

6. A 29-year-old woman has developed a sudden onset seizure while at work. She is now clinically well with a GCS score of 15 and no focal neurological deficit. A CT brain reveals haemorrhage suggesting an underlying arteriovenous malformation. What is the single most appropriate investigation to perform?

A CT angiogram
B Delayed MR angiogram
C Formal cerebral (catheter) angiography with digital subtraction (DSA)
D MR spectroscopy
E MR venogram

7. A 75-year-old man's wife suddenly noticed his slurring of speech and right arm weakness which completely resolved within 30 minutes. He has a history of diabetes. His blood pressure is normal.
Which single answer best represents his ABCD2 score?

A 2
B 3
C 4
D 5
E 7

8. A 30-year-old woman has recently undergone cardiac surgery for rheumatic valve disease. She has a metallic mitral valve inserted and is started on warfarin for primary prevention of stroke.
What is the single best INR range that should be aimed for?

A <1.2
B 1.0–2.0
C 2.0–3.0
D 3.5–4.5
E 5.0–6.0

Neurological tumours

1. A 20-year-old man has noticed a progressive decline in his hearing bilaterally over the past 2 years. He has numbness and weakness affecting his face, worse on the right. His MRI scan demonstrates bilateral vestibular schwannomas.
What is the single most likely diagnosis?

A Cowden syndrome
B Neurofibromatosis type 1
C Neurofibromatosis type 2
D Tuberous sclerosis
E Von Hippel–Lindau disease

2. A 50-year-old man has sudden onset headache, vomiting and impaired eye movements. Over the past 6 months he has noticed a progressive decline in his peripheral vision and his shoes, hat and wedding ring have become too small for him. An MRI confirms he has a brain tumour.
What is the single most likely location of his lesion?

A Brainstem
B Cerebellum
C Frontal lobe
D Pituitary gland
E Thalamus

3. A 68-year-old woman has worsening back pain and weakness in both legs that has worsened over the past 3 days. She has difficulty walking but her bowel and bladder function is normal. She has a past history of breast cancer.
Which is the single most appropriate next step?

A CT spine
B Dexamethasone infusion
C MRI of the lumbar spine
D MRI of the whole spine
E Refer to oncology and neurosurgery departments

Neurological infections

1. A 58-year-old woman has a fever and meningism. She has no focal neurological deficits. Meningitis is suspected and a lumbar puncture is performed which reveals a high white cell count (mainly polymorphs), low glucose in comparison to serum and turbidity. Gram-negative bacilli are seen.
What is the single most likely causative pathogen?

A *Haemophilus influenzae*
B Herpes simplex virus
C *Neisseria meningitidis*
D *Staphylococcus aureus*
E *Streptococcus pneumoniae*

2. A 30-year-old man has a history of fever, memory impairment and behavioural change. There is no meningism. Shortly before an MRI scan of his brain he develops a seizure. His MRI demonstrates increased signal intensity and oedema affecting the temporal lobes, with some haemorrhagic regions.
What is the single most likely cause?

A Cytomegalovirus
B Herpes simplex virus type 1
C Herpes simplex virus type 2
D Human immunodeficiency virus
E Varicella zoster virus

3. A 21-year-old man is a university student who has recently been unwell with flu-like symptoms. Since yesterday he has had nausea and vomiting, aversion to light and neck stiffness. He has a rash that is non-blanching on his arms and legs.
What is the single most appropriate next action?

A Immediate broad-spectrum IM/IV antibiotics
B Perform lumbar puncture
C Prescribe oral antibiotics and discharge
D Transfer immediately to emergency department
E Urgent CT brain scan

4. A 34-year-old man has progressively deteriorated with increasing confusion and agitation with a recent history of weight loss and night sweats in the last 6 weeks. He recently arrived in the UK from abroad. A CT scan excludes a space-occupying lesion. CSF analysis reveals elevated white cell count (predominantly mononuclear), decreased glucose and elevated protein.
What is the single most likely causative pathogen?

A Echovirus
B HIV
C *Mycobacterium tuberculosis*
D *Streptococcus pneumoniae*
E *Toxoplasma gondii*

5. A 68-year-old man has seizures. He has become increasingly confused over the past week and now complains of headache and fever. It is difficult to confirm the presence of meningism. He has no other focal neurological deficits. Emergency CT brain is negative. A decision is made to treat him while awaiting CSF analysis.
What is the single most appropriate treatment?

A Ceftriaxone
B Ceftriaxone + vancomycin + ampicillin
C Ceftriaxone + vancomycin + ampicillin + aciclovir
D Ethambutol + isoniazid + rifampicin + pyrazinamide
E Ganciclovir + pyrimethamine

6. A 51-year-old man has lower lumbar back pain and fever. He had an operation 1 week ago to remove an intervertebral disc prolapse causing nerve root compression. He has no neurological deficit. An MRI scan confirms vertebral osteomyelitis following surgery.
What is the single most likely causative organism?

A *Aspergillus*
B *Escherichia coli*
C *Salmonella*
D *Staphylococcus aureus*
E *Mycobacterium tuberculosis*

Movement disorders

1. A 31-year-old woman's partner reveals that there has been a recent change in her personality. She has also started to develop irregular, rapid, jerking movements especially affecting her arms. She doesn't take any regular medication.
What single mutation is she most likely to have?

A Defect in *hamartin* gene on chromosome 9
B Defect in *merlin* gene on chromosome 22
C Defect in *tuberin* gene on chromosome 16
D Expansion in CAG trinucleotide repeats on chromosome 4
E Expansion in CTG trinucleotide repeats on chromosome 19

2. A 25-year-old woman has been increasingly forgetful and complains of hearing voices. She appears to have bradykinesia and bilateral tremor, and is jaundiced. Her liver function tests are abnormal. She is referred for slit lamp examination of her eyes.
What is the single most likely diagnosis?

A Dementia with Lewy body formation
B Huntington's disease
C Idiopathic Parkinson's disease
D Progressive supranuclear palsy
E Wilson's disease

3. A 40-year-old man has worsening tremor in his right hand and a generalised slowing in motor function. He has noticed that his arm is becoming increasingly stiff. He also has difficulty sleeping due to bad dreams.
What is the single most appropriate treatment?

A Apomorphine
B Carbidopa
C Levodopa (L-dopa)
D Ropinirole
E Subthalamic nucleus stimulation

4. A 27-year-old man has developed a bilateral tremor and generalised rigidity in his arms and legs. He has recently been diagnosed with schizophrenia and takes chlorpromazine.
What is the single most likely diagnosis?

A Antipsychotic medication non-compliance
B Dementia with Lewy bodies
C Drug-induced parkinsonism
D HSV encephalitis
E Idiopathic Parkinson's disease

5. A 50-year-old woman has a history of bilateral progressive tremor and recent falls. She has difficulty swallowing and is very tearful. On examination she has severe stiffness in her neck and trunk. She has difficulty moving her eyes in a vertical direction.
What is the single most likely diagnosis?

A Dementia with Lewy bodies
B Idiopathic Parkinson's disease
C Multiple systems atrophy
D Progressive supranuclear palsy
E Pseudobulbar palsy

6. A 69-year-old man has increasing stiffness and heaviness in his right arm and leg over the past 2 years with a general slowing in his movements. He has no tremor but has increased reflexes in his right upper and lower limbs. He has a history of ischaemic heart and peripheral vascular disease, hypertension and smoking. A trial of L-dopa hasn't improved his symptoms.
What is the single most likely diagnosis?

A Corticobasal degeneration
B Idiopathic Parkinson's disease
C Multiple systems atrophy
D Vascular parkinsonism
E Wilson's disease

Multiple sclerosis

1. A 40-year-old female has an MRI brain scan for episodic headaches that sound like migraines, which shows several lesions in the corpus callosum and cerebellar peduncles that have features of demyelination. She has no history of focal neurological symptoms.
What is the single most likely diagnosis?

A Clinically isolated syndrome (CIS)
B Devic's disease
C Primary progressive multiple sclerosis (PPMS)
D Radiologically-isolated syndrome (RIS)
E Relapsing–remitting multiple sclerosis (RRMS)

2. A 34-year-old woman with a history of CIS 4 months ago develops new left leg weakness, ataxia and diplopia. MRI demonstrates new demyelinating lesions and she is prescribed steroids. After discussion of the risks and benefits she is keen to avoid further relapses.
What is the single most appropriate management?

A Alemtuzumab therapy
B Further steroids
C Interferon injections
D Regular natalizumab infusions
E Vitamin D supplementation

3. A 42-year-old man has relapsing–remitting multiple sclerosis and has developed slowly progressive left arm weakness and numbness over 3 weeks. He has monthly natalizumab infusions.
What is the single most appropriate management?

A Inform patient they have entered the progressive phase
B MRI brain with contrast and lumbar puncture
C Prescribe steroids
D Prescribe steroids then give next natalizumab infusion
E Switch to an alternative drug due to failure of natalizumab

Spinal disorders

1. A 21-year-old man has sudden onset back and leg pain. He was lifting weights in the gym when he developed acute pain radiating down his right leg. He has difficulty passing urine and cannot feel the toilet paper while using the bathroom.
What is the single most appropriate next step?

A Analgesia and discharge with MRI scan in 6 weeks
B Analgesia and discharge with outpatient referral to neurosurgery
C CT spine
D Dexamethasone infusion
E MRI spine

2. A 40-year-old woman has neck pain and burning pain in her right arm radiating to her middle finger. She has weakness in elbow extension and absent triceps reflex. MRI spine demonstrates a cervical disc prolapse.
What is the single most likely combination of nerve root and disc to be affected?

A C4 nerve root with C3–4 disc prolapse
B C6 nerve root with C4–5 disc prolapse
C C6 nerve root with C5–6 disc prolapse
D C7 nerve root with C6–7 disc prolapse
E C7 nerve root with C7–T1 disc prolapse

3. A 56-year-old man has a history of leg weakness. He has no previous medical history. He has weakness in his left leg with loss of light touch and proprioception on the left side. He has loss of pain and temperature sensation on the right side. His MRI confirms a tumour affecting his spinal cord.
What is the single most likely location of the tumour?

A Anterior part of the spinal cord
B Central aspect of the spinal cord
C Left side of the spinal cord
D Posterior part of the spinal cord
E Right side of the spinal cord

4. A 78-year-old man has neck pain following a fall. On examination he has weakness in both hands with loss of reflexes. Pain and temperature sensation is reduced in both arms. He has increased tone in his legs with hyperreflexia and a positive Babinski sign. MRI demonstrates a C6 segmental cervical cord injury.
What is the single most likely diagnosis?

A Anterior cord syndrome
B Brown-Séquard syndrome
C Cauda equina syndrome
D Central cord syndrome
E Posterior cord syndrome

Systemic disease affecting the nervous system

1. A 21-year-old woman has a history of seizures and mood disturbance. On examination she has an erythematous rash over her nose and cheeks, arthritis within her finger and knee joints and oral ulceration. Urine analysis demonstrates a nephritic syndrome.
What single antibody is most likely to be positive in this woman?

A Anti acetylcholine receptor (ACh-R) antibody
B Anti double-stranded DNA (dsDNA) antibody
C Anti GQ-1B antibody
D Anti Hu antibody
E Anti mitochondrial antibody (AMA)

2. A 42-year-old woman has features of painful subacute sensory polyneuropathy affecting her lower limbs. She has a history of persistent nosebleeds with nasal crusting and on examination has a saddle-nose deformity. She develops haemoptysis and her renal function is deranged. She is ANCA +ve.
Which is the single most likely diagnosis?

A Churg–Strauss syndrome
B Dermatomyositis
C Giant cell (temporal) arteritis
D Lambert–Eaton myasthenic syndrome
E Granulomatosis with polyangiitis

3. A 50-year-old woman has subacute development of short-term memory deficits. She also complains of sleep dysfunction and generalised irritability, and her family think she may be having visual hallucinations. Paraneoplastic limbic encephalitis is suspected and she is positive for anti Hu antibodies.
What is the single most likely associated malignancy?

A Breast
B Lymphoma
C Ovary
D Small-cell lung cancer
E Thymoma

Neurodegenerative disease

1. A 68-year-old man has had dysarthria and dysphagia for 4 months and left arm weakness for 1 month but no sensory signs. He has a wasted left hand, brisk limb reflexes, brisk jaw jerk and a stiff, slow tongue.
What is the single most likely diagnosis?

A Bulbar onset motor neurone disease
B Cervical myelopathy
C Multifocal motor neuropathy
D Multiple sclerosis
E Primary lateral sclerosis

2. A 21-year-old woman has progressive dysarthria, dysphagia, ataxia and diplopia. She has poor smooth pursuits, nystagmus, a slow, stiff tongue, slurred speech, wide-based gait, reduced tone in her legs, reduced knee and ankle reflexes, and reduced pinprick and joint position sense to the shins.
What is the single most likely diagnosis?

A Bulbar onset motor neurone disease
B Friedreich's ataxia
C Multiple sclerosis
D Spinocerebellar ataxia
E Spinal muscular atrophy

3. A 30-year-old man has had 6 months of proximal leg weakness, 3 months of bilateral hand intention tremor and now tongue fasciculations. He has no leg reflexes, flaccid weakness in his leg which is worse proximally, and some past-pointing with both hands. He has no upper motor neurone signs.
What is the single most likely diagnosis?

A Friedreich's ataxia
B Limb onset motor neurone disease
C Multiple sclerosis
D Spinal muscular atrophy
E Spinocerebellar ataxia

4. A 20-year-old diabetic man has a history of difficulty walking since childhood. He has worsening stiffness, balance and coordination in his legs. His grandfather had a similar undiagnosed illness. He is kyphotic, has increased tone in his legs, no knee or ankle reflexes and loss of joint position sense in his legs.
What is the single most likely diagnosis?

A Friedreich's ataxia
B Hereditary motor neurone disease
C Multiple sclerosis
D Spinal muscular atrophy
E Spinocerebellar ataxia

Dementia

1. An 84-year-old man with a history of ischaemic heart disease, hypertension and a previous minor stroke develops progressive memory problems and disinhibition over 8 months. He is diagnosed with dementia.
What is the single most likely underlying aetiology?

A Alzheimer's disease
B Delirium
C Dementia with Lewy bodies
D Parkinson's disease
E Vascular dementia

2. A 68-year-old man with a 2-year history of dementia develops frequent daytime visual hallucinations and increasing stiffness and slowness in his movements.
What is the single most likely diagnosis?

A Alzheimer's disease
B Delirium
C Dementia with Lewy bodies
D Parkinson's disease
E Vascular dementia

3. A 45-year-old man has an 8-month history of progressive change in personality, sexual disinhibition, aggression and lack of insight into these problems. An MRI of his brain shows significant frontal lobe atrophy, but nothing else.
What is the single most likely diagnosis?

A Depression
B Creutzfeldt–Jakob disease
C Frontotemporal dementia
D Vascular dementia
E Young-onset Alzheimer's disease

Congenital and hereditary conditions

1. A 10-year-old boy has developmental delay in motor function. There is evidence of myotonia and muscle weakness in facial, ocular and shoulder girdle. He is to be referred for genetic testing.
What single genetic abnormality is most likely to be found in this case?

A Defect in *hamartin* gene on chromosome 9
B Defect in *merlin* gene on chromosome 22
C Defect in *tuberin* gene on chromosome 16
D Expansion in CAG trinucleotide repeats on chromosome 4
E Expansion in CTG trinucleotide repeats on chromosome 19

2. A 1-day-old baby girl has a cystic membrane-covered swelling over the lower part of her back that was noted immediately after birth. It does not transilluminate with light. She has a defect in the lower spine posteriorly and obvious deformities of her hips and ankles.
What is the single most likely diagnosis?

A Meningocele
B Myelomeningocele
C Spina bifida occulta
D Sturge–Weber syndrome
E Von Hippel–Lindau syndrome

3. A 12-year-old girl with 10 café-au-lait spots and several Lisch nodules develops resistant seizures.
What is the most likely diagnosis?

A Cerebral palsy
B Neurofibromatosis type 1
C Sturge–Weber syndrome
D Tuberous sclerosis complex
E Von Hippel–Lindau syndrome

4. A 9-year-old boy has a history of delay in speech and language development, and seizures. On examination there are depigmented areas of skin on his back and patches of 'leathery' skin. An MRI brain demonstrates a subependymal tumour. ECHO demonstrates a cardiac muscle rhabdomyoma.
What is the single most likely diagnosis?

A Cerebral palsy
B Neurofibromatosis type 1
C Sturge–Weber syndrome
D Tuberous sclerosis complex
E Von Hippel–Lindau syndrome

Peripheral neurological disease

1. A 19-year-old man has fallen from his motorcycle in a road traffic incident. His wrist and finger extension are impaired, as is the sensation on the dorsal aspect of the arm. Elbow extension is preserved.
Which is the single most likely diagnosis?

A Median nerve injury at carpal tunnel
B Radial nerve injury at axilla
C Radial nerve injury at elbow
D Radial nerve injury at wrist
E Ulnar nerve injury at elbow

2. A 50-year-old woman with diabetes has noticed weakness in her left leg in the last few weeks. She has a foot drop on the left side with weakness of great toe dorsiflexion and eversion. Inversion is preserved. There are no other clinical signs.
What single nerve is most likely to be causing her symptoms?

A Common peroneal nerve
B Deep branch of peroneal nerve
C Femoral nerve
D Sciatic nerve
E Tibial nerve

3. A 36-year-old woman has bilateral motor weakness affecting both legs that started distally and has spread proximally over a few days. She had a recent diarrhoeal illness 3 weeks previously but is otherwise well.
What is the single most appropriate treatment?

A Cyclophosphamide
B Intravenous immunoglobulin
C Intubation and ventilation
D High-dose steroids
E Plasma exchange

4. A 46-year-old man has a past medical history of alcohol use. He is unkempt and looks emaciated. On examination he is confused, has an ataxic gait and complains of diplopia especially on lateral gaze.
What is the single most likely diagnosis?

A Cobalamin deficiency
B Nicotinic acid deficiency
C Pyridoxine deficiency
D Riboflavin deficiency
E Thiamine deficiency

5. A 53-year-old woman has aching muscles. Both arms and legs are affected, with the proximal muscles affected more than the distal. She has a violet discoloration around her eyelids and purple patches over her knuckles. Her EMG is myopathic.
What is the single most likely diagnosis?

A Becker muscular dystrophy
B Dermatomyositis
C Fascio-scapulo-humeral dystrophy
D Inclusion body myositis
E Mitochondrial disease

6. A 29-year-old woman has bilateral weakness and drooping of both eyelids. She also has fatiguable weakness in both her arms and legs. She has a past medical history of pernicious anaemia.
What single treatment will cause a catastrophic deterioration in her condition if given?

A Anticholinergic
B Anticholinesterase
C Cyclophosphamide
D Intravenous immunoglobulin
E Steroids

7. A 68-year-old woman has weakness affecting her lower limbs initially. She doesn't have any ocular or bulbar symptoms. Her symptoms seem to improve following exercise. She is a lifelong smoker and has been newly diagnosed with small-cell lung cancer.
What is the single most appropriate investigation to perform?

A Anti AChR-antibody
B Anti dsDNA-antibody
C Anti Hu-antibody
D Anti Ri-antibody
E Anti VGCC-antibody

SBA answers

Raised intracranial pressure and head injury

1. **C**
   Management via ATLS (Advanced Trauma and Life Support) protocols requires immediate assessment, resuscitation and treatment of life-threatening injuries before embarking on specialist intervention. (B) would happen as part of the 'D' in the ABCDE with an urgent CT brain scan (E) once the patient is initially stabilised. Alerting the on-call neurosurgeon (A) should follow the CT brain scan and (D) (mannitol) is indicated only if there are clinical signs of brain herniation (e.g. dilated pupil unreactive to light) after discussion with a neurosurgeon.
2. **C**
   The GCS of 8 confirms a severe traumatic brain injury (TBI). Mild TBI is GCS 14–15 and moderate GCS 9–13. Crescent-shaped collections are subdural in origin. Extradural collections are bi-convex as they are restricted by skull sutures. The hyperdense nature confirms an acute collection. Chronic collections are hypodense.
3. **D**
   The history is typical of a lucid interval following traumatic injury, followed by sudden deterioration that is seen with an acute extradural haematoma. Acute subdural haematomas (B) can present like this, but are more commonly associated with impaired conscious level from outset, as are intracerebral haematomas (C). Chronic subdural haematomas present with progressive neurological deficits over a longer time period. Hydrocephalus (E) is unlikely given the clear history of trauma.
4. **A**
   Decreasing CO_2 levels will vasoconstrict cerebral blood vessels, decrease cerebral blood flow and volume, and decrease intracranial pressure. Increasing CO_2 levels (B) will cause vasodilatation, raising intracranial pressure. Hyperthermia (C) and placing the patient's head down (D) both increase intracranial pressure, the latter by impairing jugular venous outflow from the brain. Corticosteroids (E) are contraindicated in traumatic brain injury, as they are associated with poor outcomes and increased mortality.
5. **E**
   The patient has a significant impairment in conscious level with a haemorrhage. Therefore he cannot be observed (D). He requires an operation (B). He is coagulopathic and needs this to be corrected before any surgical intervention (A). The warfarin must be stopped. Platelet transfusions do not reverse warfarin-induced coagulopathy (C) and are used to reverse aspirin-induced coagulopathy. Vitamin K and clotting factors help rapidly reverse warfarin-induced coagulopathy.

Headache and pain syndromes

1. **B**
   The restlessness, ptosis, eye watering and nasal stuffiness are consistent with cluster headache. First-line treatment is with high-flow oxygen and subcutaneous or intranasal triptans.
2. **A**
   High-dose aspirin (or an NSAID) with a prokinetic antiemetic to speed up gastric emptying is first-line treatment for a migraine. She has infrequent attacks and wants to become pregnant, so prophylactic treatment is best avoided at present. Opioid medications should be avoided in migraine as they are ineffective.
3. **E**
   The very short time from onset to peak makes this likely to be a thunderclap headache; migraines usually take longer to peak. The photophobia and vomiting are potential red flag signs. Subarachnoid haemorrhage is the most serious cause to exclude initially and is best shown on plain CT.
4. **D**
   New persistent headaches in this age group are potentially secondary to serious underlying causes. The persistence of symptoms and their postural nature are also warning signs for a mass lesion. The history of breast cancer raises suspicion of metastatic intracranial masses. An urgent outpatient neurology clinic is required to assess the headache, and investigations are required.
5. **D**
   This headache has postural features and the high opening pressure on lumbar puncture confirms raised intracranial pressure. Scans excluded mass lesions and venous thrombosis. Idiopathic intracranial hypertension is the most likely diagnosis and is most common in young obese women. Immediate treatment with repeat lumbar punctures to reduce the intracranial pressure is the only option which will address the immediate risk of visual loss, proceeding to LP shunt (B) if this is unsuccessful.
6. **B**
   A septic venous sinus thrombosis is likely with the history of sinus infection, fever, severe headache and a new seizure. There is no confusion or meningism to suggest meningitis. An immediate CT venogram is required to confirm the diagnosis and exclude complications such as intracranial haemorrhage that would prohibit anticoagulation.

Seizures

1. **A**
   This is a seizure with a focal onset: a partial seizure. She has lost awareness during the seizure so it is a complex partial seizure not a simple partial seizure (D). It terminated after 2 minutes, excluding (E). It is not (C) because the seizure focus is clearly localised to the right frontal lobe with no secondary generalisation (B).
2. **E**
   Temporal lobe seizures produce a sense of fear and gustatory and auditory hallucinations. Frontal lobe seizures, (A) usually have motor features, whilst occipital lobe seizures (B) produce visual hallucinations. Parietal lobe seizures (C) usually produce sensory features (e.g. tingling). The pituitary gland (D) is not an anatomical site for seizures.
3. **C**
   This seizure semiology and the neurophysiology findings are classical for absence seizure, which is a type of generalised seizure. Status epilepticus (E) is when seizures continue uninterrupted for at least 30 minutes or when there is no regaining of conscious level in between seizures. Absence seizures are not simple partial (D), where there is a focal seizure onset with preservation of awareness. Nor are they complex partial with loss of awareness (B) or complex partial with secondary generalisation (C).
4. **C**
   Lamotrigine is very useful as first-line monotherapy for seizures especially if pregnancy is being considered, as it is the least associated with teratogenic side-effects. (D) is not indicated, as monotherapy with a range of medications must be used first. Valproate (E) and carbamazepine (A) are both teratogenic. Clobazam (B) is usually second-line and an add-on therapy.
5. **A**
   The clinical features are suggestive of myoclonic epilepsy. Carbamazepine is not recommended for myoclonic epilepsy as it may paradoxically worsen myoclonic seizures.
6. **B**
   This patient is in status epilepticus and needs prompt intervention to stop her seizures. Intravenous lorazepam or rectal diazepam are first-line treatments for status epilepticus. Phenytoin infusion (D) is a second-line treatment for status epilepticus and (E) with barbiturate infusion for deep sedation is third-line. Carbamazepine (A) is not used in status epilepticus. Magnesium sulphate (C) is used in eclampsia-related seizures.

Neurovascular disease

1. C
A CT of the brain is needed to exclude haemorrhagic stroke first, which would contraindicate thrombolysis. (A) and (B) would usually follow if this is proven to be ischaemic not haemorrhagic stroke. Therapeutic dose heparin is not indicated in the treatment of acute ischaemic stroke (E).
(D) is not performed to exclude haemorrhagic stroke acutely but has a role with diffusion-weighted imaging (DWI) sequences in confirming ischaemic stroke.

2. B
This is a PACS with motor weakness and homonymous hemianopia without higher cognitive dysfunction. With higher cognitive dysfunction, it would be a TACS (D). Posterior circulation strokes (C) usually affect the brainstem or medial temporal or occipital lobes, and usually have isolated visual dysfunction, cranial nerve or extensive long tract signs. Lacunar strokes (A) have only isolated features (e.g. pure motor, pure sensory, ataxic hemiparesis). Weber's syndrome (E) is due to midbrain stroke following occlusion of posterior cerebral or basilar arteries with ipsilateral 3rd cranial nerve palsy and contralateral hemiparesis.

3. E
These are clinical features of the lateral medullary syndrome of Wallenberg (stroke affecting right lateral medulla) with dysfunction of the vestibular nuclei (vertigo), vagal nerve nuclei (swallowing), cerebellum and inferior peduncle (ataxia), sympathetic tracts (Horner's), trigeminal nuclei (loss of pain and temperature on face) and crossed spinothalamic tract (loss of pain and temperature contralateral body). The vessel involved is usually the posterior inferior cerebellar artery (PICA).

4. C
This is an acute subarachnoid haemorrhage (SAH). The initial CT of the brain may be normal but xanthochromia following lumbar puncture can confirm the presence of acute SAH. (A) is unsafe and (E) will not alter management if repeat CT is normal. (B) is performed once SAH is confirmed and an aneurysmal cause is suspected. There is no indication for antibiotics with this clinical history, which is consistent with acute SAH and not infectious meningitis.

5. C
All of the options are known complications following a subarachnoid haemorrhage (SAH) and the patient will require close monitoring for them. Any drop in conscious level or neurological deterioration should prompt a search for and exclusion of all of them. However, the most important life-threatening complication following acute aneurysmal subarachnoid haemorrhage (especially in the first few days) is a repeat rupture of the aneurysm and recurrent haemorrhage. The presence of normal ventricles without haemorrhage makes hydrocephalus unlikely (A). Hyponatraemia does not occur in the acute phase in SAH or cause a fixed dilated pupil (B). Vasospasm and delayed ischaemic neurological deficits occur between days 4 and 9 (E). Seizures are unlikely to be the cause in the presence of features of uncal herniation (D).

6. C
DSA is the gold standard investigation to plan treatment for a vascular malformation (aneurysm or arteriovenous malformation). MR spectroscopy (D) evaluates the biometabolic parameters of brain tissue and so is more useful for neurological tumours. (A), (B) and (E) are non-invasive tests, which may be useful in diagnosis, but only formal angiography can measure exact inflow and outflow to an AVM and plan treatment.

7. D
This patient scores 1 for his age, 2 for the presence of weakness, 1 for diabetes and 1 for resolution of symptoms within 60 minutes. The ABCD2 score assesses risk of subsequent stroke. A score of 6 confers an 8.1% risk of developing a stroke within 2 days and a 35.5% risk of stroke within 7 days. Patients scoring 4 or more should be seen by a specialist stroke physician within 24 hours.

8. D
Metallic valves (especially mitral) have a high risk of thrombus development so require anticoagulation with warfarin at high therapeutic ranges to prevent systemic and cranial thrombotic complications, and stroke. (A) is normal coagulation function. (B) is mildly deranged coagulation function. (C) is the range employed in patients with atrial fibrillation or following deep vein thrombosis or pulmonary embolism. (E) is excessively over-anticoagulated.

Neurological tumours

1. C
The presence of bilateral vestibular schwannomas is diagnostic of NF2. There are no features to suggest NF1 (B) as a diagnosis, which requires at least 2 out of 7 features. TS (D) is classically associated with subependymal giant cell astrocytomas. VHL (E) is classically associated with cerebellar haemangioblastomas. Cowden syndrome (A) is an inherited syndrome predisposing to development of meningiomas.

2. D
The patient is likely to be presenting with pituitary apoplexy, which is a sudden haemorrhagic infarction of usually a pituitary tumour. The history suggests the presence of bitemporal hemianopia (due to optic chiasm compression). The finding that the patient's hat, shoes and wedding ring have all become too small is typical of patients with pituitary adenomas with excessive secretion of growth hormone because it results in increased size of head, hands and feet. Given that the history suggests a pituitary adenoma likely hyper-secreting growth hormone, the presentation is most likely due to a pituitary lesion.

3. D
The clinical features and history of malignancy suggest an acute spinal cord compression, so metastatic spinal cord compression must be excluded. (C) will not evaluate the whole spine. (A) is good for bone detail and fractures, but MRI is gold standard for evaluating spinal cord compression and evaluating soft tissue detail. (B) is only started once the diagnosis is confirmed radiologically and (E) follows imaging and confirmation of diagnosis.

Neurological infections

1. A
The CSF findings are consistent with bacterial meningitis. *Haemophilus influenzae* is the only Gram-negative bacillus listed. B is a virus, (D) and (E) are both Gram-positive cocci and (C) are Gram-negative cocci.

2. B
The history suggests encephalitis. HSV-1 is the commonest sporadic cause of viral encephalitis. The MRI changes affecting the temporal lobes are usually seen in HSV-1 encephalitis. (C) (HSV-2) can cause viral encephalitis in neonates especially. (A), (D) and (E) can all cause encephalitis but they are not the commonest causes of sporadic encephalitis. CMV in particular affects patients who are immunocompromised, such as patients with HIV infection and AIDS.

3. A
The most likely diagnosis is bacterial meningitis as the patient has photophobia and neck stiffness, and a non-blanching rash suggestive of meningococcal sepsis. Immediate treatment with broad-spectrum antibiotics is indicated by administering a dose of intramuscular antibiotics before admission. (D) should happen next followed by (E) to exclude any mass lesions and then lumbar puncture to obtain CSF. (C) would be dangerous, given the suspicion of bacterial meningitis.

4. C
The CSF picture and history are consistent with infectious meningitis of tuberculous origin. (A) and (B) are unlikely although with tuberculosis, a diagnosis of underlying immunosuppression with HIV should be considered. Bacterial meningitis is unlikely without a raised polymorphic white cell count. (E) is usually seen in immunosuppression and causes multiple space-occupying lesions in the brain.

5. **C**
Both bacterial meningitis and viral encephalitis must be considered, especially viral encephalitis given the seizures. Empirical antibiotics in an elderly patient must cover Gram-positive cocci (e.g. ceftriaxone and vancomycin), *Listeria monocytogenes* (ampicillin) and aciclovir to cover viral encephalitis. (A) and (B) are insufficient and (D) is treatment for tuberculosis. (E) is cover for CMV and toxoplasmosis (commonly used in patients with HIV).

6. **D**
The commonest cause of osteomyelitis due to iatrogenic surgery is *Staphylococcus aureus*. Empirical antimicrobials need to cover for both methicillin-sensitive (MSSA) and methicillin-resistant (MRSA) strains. There are no features to suggest (E) (e.g. foreign travel, immunosuppression, long clinical history). (C) is common in patients with sickle cell anaemia and immunosuppression. (A) is a rare cause of osteomyelitis and is common in immunosuppression. (B) can cause osteomyelitis but (D) is far more likely following recent surgery.

Movement disorders

1. **D**
The patient's young age, new psychiatric symptoms (change in personality), new abnormal movements (chorea) and lack of medicinal causes make Huntington's disease the likely diagnosis. This is caused by a CAG trinucleotide expansion on chromosome 4. (E) is the genetic abnormality in myotonic dystrophy. (A) and (C) are associated with tuberous sclerosis. (D) is the gene defect in neurofibromatosis type 2.

2. **E**
Neuropsychiatric manifestations together with features of parkinsonism and abnormal liver function in a young patient strongly raise the possibility of Wilson's disease, and slit lamp examination may reveal Kayser–Fleischer rings, a sign of Wilson's disease. (B) is an autosomal dominant condition with neuropsychiatric manifestations plus severe choreiform movements. (A) and (C) are unlikely in a young patient and (D) would include a vertical gaze palsy.

3. **D**
This patient, although young, has classical features of idiopathic Parkinson's disease (unilateral onset of tremor, bradykinesia and rigidity); non-REM sleep behaviour disorder may also be an early manifestation. Dopamine agonists are used first-line in young patients to delay the use of L-dopa (C). (A) is also used at a later stage. (E) is reserved for selected patients after failure of medical therapy. (B) is used in combination with L-dopa, not alone.

4. **C**
The patient's young age suggests a degenerative cause. His use of the dopamine-blocking agent chlorpromazine and the symmetrical nature of the symptoms make a drug-induced parkinsonism most likely.

5. **D**
Bilateral progressive tremor suggests atypical parkinsonian symptoms. Along with the difficulty swallowing, emotional lability, truncal axial rigidity and vertical gaze palsy, progressive supranuclear palsy is most likely. (A) causes atypical parkinsonian symptoms but presents with prominent visual hallucinations and no gaze paresis. (C) may present with predominantly cerebellar or autonomic symptoms. (B) presents with classical parkinsonism and no gaze paresis. (E) presents with difficulty swallowing and emotional lability, but usually no gaze restriction.

6. **D**
Stepwise decline in function and multiple risk factors for atherosclerotic vascular disease suggest vascular parkinsonism. Upper motor neurone signs (hyperreflexia and lack of improvement with L-dopa) also point to this. (B) would have parkinsonian symptoms and good initial response to L-dopa. (C) is an atypical cause of parkinsonism with prominent cerebellar or autonomic symptoms. (E) presents with neuropsychiatric manifestations and features of liver disease. (A) usually presents with myoclonus, dementia and abnormal arm movements.

Multiple sclerosis

1. **D**
RIS is the incidental finding of MRI evidence of CNS demyelination in the absence of neurological symptoms. Around 50% will progress to clinical symptoms and signs (CIS or RRMS). (A) is a single episode of clinical symptoms potentially caused by CNS demyelination with insufficient radiological or clinical evidence of previous episodes. (C) is slow, progressive accumulation of neurological disability with radiological and CSF evidence of demyelination. (B) (neuromyelitis optica) is an antibody-mediated CNS demyelinating disorder characterised by bilateral optic neuritis and transverse myelitis. (E) requires multiple clinical episodes of neurological dysfunction with resolution between episodes and demyelination on MRI/CSF.

2. **D**
This patient has an aggressive form of multiple sclerosis (two disabling episodes in 12 months). She is eligible for natalizumab to significantly reduce the frequency of relapse. Alemtuzumab (A) is used for failed or contraindicated therapy with natalizumab. Steroids (B) do not reduce relapse frequency. Interferon (C) is used as first-line disease-modifying therapy but with aggressive multiple sclerosis she is eligible for more potent therapies. Vitamin D (E) should be offered but has unknown effect on relapses or progression.

3. **B**
Suspected relapse or slowly progressive symptoms on natalizumab need investigating to exclude progressive multifocal leucoencephalopathy (PML) which is caused by the JC virus. An MRI with contrast will show new lesions and if the patient has PML there will be JC viral DNA in the CSF. The progression over 3 weeks is quite long for a relapse and needs investigating rather than treating (C, D and E). (A) cannot be accepted without excluding PML in a patient taking natalizumab, since this also causes progressive symptoms.

Spinal disorders

1. **E**
This is acute cauda equina syndrome (perianal numbness and saddle anaesthesia, urinary sphincter dysfunction), a medical emergency. The clinical history suggests acute lumbar disc prolapse. Urgent MRI of the spine will establish diagnosis. (A) and (B) are inappropriate, as surgical decompression within 48 hours is essential to preserve neurological function. A CT spine (C) is useful for bony visualisation but will not identify compression of the cauda equina and disc prolapse. (D) is not indicated in cauda equina syndrome.

2. **D**
The patient has clinical features of a radiculopathy. The dermatomal and myotomal distributions of sensory and motor symptoms respectively determine the spinal nerve root affected and the level of disc prolapse. Elbow extension is controlled by triceps and C7 nerve root in particular, which includes sensory distribution of the middle finger.

3. **C**
The clinical features suggest Brown-Séquard syndrome. Therefore the tumour is compressing the left spinal cord. (D) would cause a right-sided Brown-Séquard syndrome with motor weakness and dorsal column impairment on the right side. (E) would cause features of central cord syndrome. (A) would cause anterior cord syndrome with preserved dorsal column function but loss of bilateral motor function and pain and temperature sensation. (B) would cause bilateral loss of dorsal column function but preserved motor function and pain and temperature sensation.

4. **D**
This is classical central cord syndrome (upper limbs affected more than lower, distal muscles affected more than proximal, loss of pain and temperature in a cape-like distribution, lower motor neurone signs in arms with upper motor neurone signs in lower limbs). Anterior cord syndrome (A) is bilateral motor weakness and loss of pain and temperature distribution with preservation of dorsal column function. (B) is ipsilateral loss of motor and dorsal column function with contralateral loss of pain and temperature. (C) is sphincter dysfunction, perianal numbness and cauda equina syndrome. (E) is isolated dorsal column dysfunction.

Systemic disease affecting the nervous system

1. B
This patient has systemic lupus erythematosus, confirmed by the presence of classical malar rash, arthropathy, oral ulceration, and renal and neurological complications. The classic autoantibody is anti-dsDNA antibody. (A) is associated with myasthenia gravis. (C) is associated with the Miller Fisher variant of Guillain–Barré syndrome. (D) is associated with neurological paraneoplastic syndromes and (E) is associated with primary biliary cirrhosis.

2. E
Nasal discharge with saddle-nose deformity, renal complications (likely nephritic syndrome) and haemoptysis (as a result of pulmonary haemorrhage) suggests granulomatosis with polyangiitis. ANCA positivity confirms vasculitis. (D) is a paraneoplastic syndrome presenting with weakness, improving with exercise and anti-voltage-gated calcium channel antibodies. (C) presents with headache, jaw claudication and visual dysfunction. (B) is associated with arthropathy, characteristic rashes (Gottron's papules, shawl rash) and anti-Jo/Mi2 antibodies. (A) presents with asthma, eosinophilia and small-to-medium vessel vasculitis.

3. D
Anti-Hu antibodies are typically associated with small-cell lung cancer. (E) is usually associated with anti-CV2 antibodies, (C) with anti-Yo antibodies, (B) with anti-Tr antibodies and (A) may be associated with anti-Yo, anti-Ri and anti-amphiphysin antibodies.

Neurodegenerative disease

1. A
There are mixed upper and lower motor neurone signs, with upper motor neurone signs at a more rostral level of the nervous system than lower motor neurone signs (e.g. brisk jaw jerk, wasted hand muscles). Multifocal motor neuropathy (C) and cervical myelopathy (B) do not cause bulbar dysfunction. His age and progressive symptoms argue against multiple sclerosis. The lower motor neurone signs exclude primary lateral sclerosis.

2. D
Her sensory signs exclude motor neurone disease. Friedreich's ataxia could account for loss of reflexes and altered joint position sense but would not cause eye movement abnormality, nystagmus or slurred speech. Multiple sclerosis is possible but the progressive nature argues against the common relapsing–remitting form and it would not cause reduced tone and reflexes in the legs.

3. D
Spinal muscular atrophy causes slowly progressive lower motor neurone symptoms and signs in this manner. The lack of both upper and lower motor neurone signs at the same level argues against motor neurone disease. Spinocerebellar ataxia (E), Friedreich's ataxia (A) and multiple sclerosis (C) would not cause tongue fasciculation.

4. A
The long history of motor problems and physical deformity argue against motor neurone disease (B) and multiple sclerosis (C). Spinal muscular atrophy (D) does not cause increased muscle tone. The family history and diabetes are consistent with Friedreich's ataxia, as are the other symptoms and signs. There are no other cerebellar signs suggestive of spinocerebellar ataxia.

Dementia

1. E
The history of hypertension, ischaemic heart disease and previous stroke make vascular dementia the most likely cause. Alzheimer's disease is possible but these other risk factors make vascular dementia the most likely. There are no signs of parkinsonism to suggest Parkinson's disease or dementia with Lewy bodies.

2. C
The early visual hallucinations and symptoms of parkinsonism make dementia with Lewy bodies most likely. Alzheimer's doesn't usually cause the stiffness and slowness at such an early stage. Parkinson's disease can cause dementia but this usually occurs after the movement disorder and much later in the disease.

3. C
The symptoms point to frontal lobe dysfunction and the lengthy time course suggests a slowly progressive degenerative condition, making frontotemporal dementia most likely. Young-onset Alzheimer's usually presents with memory problems and Creutzfeldt–Jakob disease would cause dysfunction in other cognitive domains and have characteristic findings on MRI. There are no risk factors for vascular dementia. Depression can sometimes cause an apathetic or withdrawn state causing some impairment in cognition, but doesn't cause these types of personality change.

Congenital and hereditary conditions

1. D
The diagnosis is likely to be myotonic dystrophy. The genetic abnormality is in the *DMPK* gene with a trinucleotide repeat expansion of >37 repeats. (A) is associated with Huntington's disease. (C) and (E) are associated with tuberous sclerosis. (B) is associated with neurofibromatosis type 2.

2. B
The patient has a myelomeningocele as opposed to a pure meningocele (A). (D) is facial port wine stain with abnormal cranial vascular calcification. (E) is a syndrome associated with cerebellar haemangioblastomas. (C) is a 'closed' posterior spinal defect with normal skin overlying the defect and no herniation of spinal cord tissue.

3. B
The presence of more than 6 café-au-lait spots and multiple Lisch nodules suggests a diagnosis of neurofibromatosis type 1. The resistant seizures suggest an associated structural lesion within the CNS and that requires further investigation.

4. D
The clinical features meet the diagnostic criteria for tuberous sclerosis (TS) complex. The depigmented areas of skin are known as ash leaf macules. The patches of leathery skin describe shagreen patches. Shagreen patches, subependymal giant cell astrocytoma and cardiac rhabdomyoma are all major features. A definite diagnosis of TS occurs in the presence of two major features or one major and two minor (e.g. bone cysts, pitting of teeth) features.

Peripheral neurological disease

1. C
The extensors of the upper limb are supplied by the radial nerve. Preservation of elbow extension means the injury is at a lower level than the axilla, so it is not (B). Loss of wrist and finger extension with altered sensorium means the injury is at a higher level than the wrist, so it is not (A) or (D). The ulnar nerve supplies the flexor compartment with no involvement in the extensors, so it is not (E).

2. A
The patient has weakness in ankle dorsiflexion, great toe dorsiflexion and eversion indicating involvement of both branches of the peroneal nerve, so it is not (B). Preservation of inversion suggests that the tibial nerve is preserved, so it is not (E). The sciatic nerve divides into common peroneal and tibial nerves, so it is not the sciatic nerve (D). The femoral nerve (C) supplies knee extensors, which are unaffected.

3. B
The patient has clinical features consistent with Guillain–Barré syndrome (GBS). Intravenous immunoglobulin and plasma exchange are equally effective at improving symptoms, but intravenous immunoglobulin is easier to administer and so is the first-line treatment. Steroids and cyclophosphamide are not treatments for GBS. The patient has no signs of imminent respiratory failure requiring intubation and ventilation, but needs to be observed for this.

4. E
The clinical features are consistent with Wernicke's encephalopathy. (A) (B12) deficiency causes subacute combined degeneration of the spinal cord, (C) is vitamin B6, deficiency of which causes dermatitis, confusion and neuropathy; vitamin B3 deficiency causes pellagra, which is characterised by dementia, dermatitis and diarrhoea. (D) is vitamin B2 deficiency, which leads to anaemia, stomatitis and other skin changes.

5. B
The clinical features are consistent with a myopathy: the heliotrope rash (violet discoloration around the eyelids) and Gottron's papules (patches over the knuckles) suggest dermatomyositis, an inflammatory myositis. (D) commonly affects more elderly people, particularly males, with the distal muscles affected initially. (A) is an inherited muscular dystrophy affecting much younger patients. (C) affects specific muscle groups of the face, scapula and limb girdle. Mitochondrial disorders (E) frequently have slowly progressive myopathies but do not cause the skin changes described here.

6. A
The patient has myasthenia gravis, a disorder of cholinergic neurotransmission at the neuromuscular junction. Anticholinergic medication can acutely exacerbate the impaired neurotransmission in these patients and cause life-threatening respiratory and bulbar weakness. (C), (D) and (E) are all used in the treatment of myasthenia gravis.

7. E
This patient has features consistent with Lambert–Eaton myasthenic syndrome (LEMS), a paraneoplastic syndrome commonly associated with small-cell lung cancer in particular. The autoantibody associated is the anti-voltage-gated calcium channel antibody. (C) and (D) are paraneoplastic autoantibodies but not associated with LEMS. (A) is associated with myasthenia gravis in which the symptoms of weakness do not paradoxically improve with exercise. (B) is associated with systemic lupus erythematosus.