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Chapter 4

Notes & Notes for MRCP
By Dr. Yousif Abdallah Hamad Chapter 4

Neurology Prognosis • BPPV has a good prognosis and usually resolves spontaneously after a few weeks to months. MRCPUK-part-1-May 2017 exam: H/O vertigo and dizziness precipitated by a change in head position. What is the most appropriate next step to confirm the diagnosis? Dix-Hallpike manoeuvre

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Meniere's disease Definition • Meniere's disease is a disorder of the inner ear of unknown cause. • characterised by excessive pressure and progressive dilation of the endolymphatic system. Epidemiology • more common in middle-aged adults but may be seen at any age. • similar prevalence in both men and women. Features • Recurrent episodes of vertigo, (the prominent symptom) • Tinnitus and hearing loss (sensorineural). • Sensation of aural fullness or pressure • Nystagmus • Positive Romberg test • Episodes last minutes to hours • Typically, symptoms are unilateral but bilateral symptoms may develop after a number of years Natural history • symptoms resolve in the majority of patients after 5-10 years • the majority of patients will be left with a degree of hearing loss • psychological distress is common Management • patients should inform the DVLA. The current advice is to cease driving until satisfactory control of symptoms is achieved • Acute attacks: buccal or intramuscular prochlorperazine. • Restriction of salt and fluid may hasten resolution. MRCPUK-part-1-May 2013 exam: H/O recurrent attacks of 'dizziness' + 'roaring' sensation in the left ear. Weber's test localises to the right ear. What is the most likely diagnosis? Meniere's disease

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_Vestibular neuronitis Definition • Vestibular neuronitis is a cause of vertigo that often develops following a viral infection. Features • recurrent vertigo attacks lasting hours or days • nausea and vomiting may be present • horizontal nystagmus is usually present • no hearing loss or tinnitus

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Management • vestibular rehabilitation exercises are the preferred treatment for patients who experience chronic symptoms • betahistine is often used although the evidence base suggests it is less effective than vestibular rehabilitation

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Tinnitus Causes of tinnitus Meniere's disease Associated with hearing loss, vertigo, tinnitus and sensation of fullness or pressure in one or both ears Otosclerosis Onset is usually at 20-40 years Conductive deafness Tinnitus Normal tympanic membrane (10% of patients may have a 'flamingo tinge', caused by hyperaemia) Positive family history Acoustic neuroma Hearing loss, vertigo, tinnitus Absent corneal reflex is important sign Associated with neurofibromatosis type 2 Hearing loss Causes include excessive loud noise and presbycusis Drugs Aspirin Aminoglycosides Loop diuretics Quinine The combination of sensorineural deafness, facial nerve palsy and cranial nerve V involvement suggests a cerebellopontine angle tumour, for example, acoustic neuroma. Other causes include • impacted ear wax • chronic suppurative otitis media 0BClinical physiology of the ear • The scala media contains the organ of Corti, which produces nerve impulses in response to sound vibrations. • High-frequency waves are detected in the scala vestibuli. • Low-frequency waves are detected in the scala tympani. • Normal hearing frequency ranges from 20 to 20 000 Hz.

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Neurology

Hearing loss Conductive hearing loss Sensorineural hearing loss Age of Onset • commonly in childhood or young adulthood Aetiology • Otosclerosis • Otitis media • Ear barotrauma • Cerumen Impaction • External auditory canal atresia Pathophysiology • External or middle ear pathology that disrupts conduction of sound into the inner ear Clinical Features • Hearing improves in noisy environments • Volume of voice remains normal because inner ear and auditory nerve are intact • Sound normally is not distorted • Features of external auditory canal pathology (e.g., cerumen impaction) Weber Test(unilateral hearing loss) • Lateralization to impaired ear (cannot hear ambient room noise well, so detection of vibration is greater) Rinne Test(unilateral hearing loss) • Bone conduction > air conduction (vibrations bypass blockage to reach the cochlea) Down syndrome : Hearing loss • 60%-70 develop conductive deafness due to glue ear • 10%-15% develop sensorineural deafness

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• commonly in middle or late age • Ménière's disease • Acoustic neuroma • Noise-induced hearing loss • Internal ear infections • Presbycusis • Inner ear, cochlear, or auditory nerve pathology that impairs neuronal transmission to the brain • Hearing worsens in noisy environments • Volume of voice may be loud because nerve transmissions are impaired • Tend to lose higher frequencies preferentially, such that sounds may be distorted • Absent features of external auditory canal pathology • Lateralization to good ear (sound is not transmitted by damage inner ear or auditory nerve) • Air conduction > bone conduction (the inner ear or auditory nerve cannot transmit sound information well regardless of how vibrations reach the cochlea)

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Rinne's and Weber's test • Performing both Rinne's and Weber's test allows differentiation of conductive and sensorineural deafness. • Rinne's test tuning fork is placed over the mastoid process until the sound is no longer heard, followed by repositioning just over external acoustic meatus air conduction (AC) is normally better than bone conduction (BC) if BC > AC then conductive deafness • Weber's test tuning fork is placed in the middle of the forehead equidistant from the patient's ears the patient is then asked which side is loudest in unilateral sensorineural deafness, sound is localised to the unaffected side in unilateral conductive deafness, sound is localised to the affected side

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Motion sickness Overview • Motion sickness describes the nausea and vomiting which occurs when an apparent discrepancy exists between visually perceived movement and the vestibular systems sense of movement Management • the BNF recommends hyoscine (e.g. transdermal patch) as being the most effective treatment. Use is limited due to side-effects • non-sedating antihistamines such as cyclizine or cinnarizine are recommended in preference to sedating preparation such as promethazine

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Peripheral neuropathy Definitions • Allodynia: pain caused by a stimulus that does not normally cause pain (e.g. light touch, contact with clothing) • Dysesthesia: abnormal spontaneous sensations (burning, stinging, stabbing) from activities that do not normally cause pain) • Paresthesia: an abnormal skin sensation in the absence of a stimulus (described as burning, prickling, itching, tingling) • Hyperesthesia: increased sensitivity to sensory stimuli • Hypoesthesia: decreased sensitivity to sensory stimuli Classifications • neuropathy is classified into: mononeuropathy commonly due to entrapment or trauma; mononeuropathy multiplex commonly due to leprosy and vasculitis; and polyneuropathy due to systemic, metabolic or toxic etiology. • Peripheral neuropathy may be divided into conditions which predominately cause a motor or sensory loss

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Neurology Predominately motor loss Predominately sensory loss • Guillain-Barre syndrome • porphyria • lead poisoning • hereditary sensorimotor neuropathies (HSMN) - Charcot-Marie-Tooth • chronic inflammatory demyelinating polyneuropathy (CIDP) • diphtheria Types • Large-fibre neuropathy the earliest clinically identifiable feature of peripheral sensory motor neuropathy. Reduced light pressure sensation and vibration sensation are the earliest clinically identifiable manifestations of large fibre neuropathy. Features paraesthesia glove and stocking sensory loss increased risk of charcot arthropathy, particularly in association with autonomic nerve dysfunction. reduced vibration and proprioception sensation, loss of reflexes (diminished ankle jerks), muscle wasting increased blood flow. • Small-fibre neuropathy typically presents with pain and loss of temperature sensation, with relative preservation of other sensory modalities and muscle strength. General neurological examination and reflexes are usually normal not detectable on conventional nerve conduction studies, which can only investigate large fibres. Causes of small fiber neuropathy Diabetes is a common cause and should be excluded in any patient with a painful peripheral neuropathy. Conditions in which the small fibres are preferentially affected in the early stages include diabetes and amyloidosis. In the later stages however the neuropathy in these conditions also affects large fibres. Amyloidosis Fabry's

disease X-linked lysosomal storage disorder causes a painful peripheral neuropathy, due to deposition of glycosphingolipids within small sensory fibres. Nerve conduction studies are typically normal as large fibres are unaffected. Tangier's disease Hereditary sensory and autonomic neuropathy Sjogren's syndrome: pure sensory neuropathy (ganglionopathy). Chronic idiopathic small fiber sensory neuropathy Notes & Notes for MRCP
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• diabetes • uraemia • leprosy • alcoholism • vitamin B12 deficiency • amyloidosis • Sjogren's syndrome

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Small-fibre neuropathy Large-fibre neuropathy Loss of pain and temperature Loss of touch, vibration and position sense Sensory ataxia Preservation of reflexes and motor function Reflexes lost early and motor functions impaired Electrophysiological test is silent Skin biopsy are used Impaired nerve conduction velocity
Biopsy in diagnosis of neuropathy Skin punch biopsy can be done if a small-fiber neuropathy is suspected; loss of nerve endings supports that diagnosis. Nerve biopsy is occasionally done to help differentiate demyelinating from vasculitic largefiber neuropathies. If vasculitis is a consideration, the biopsy specimen should include skin and muscle to increase the likelihood of a definitive diagnosis. If all limbs are affected, MRI can be done to rule out cervical spinal cord compression. Lead neuropathy • purely motor neuropathy affecting mainly the upper limbs. Thalamic infarcts neuropathy • commonly cause late-onset of severe neuropathic pain weeks to months after the stroke. • The pain is intractable to analgesics. • The treatment of choice for neuropathic pain is amitriptyline/gabapentin.

Alcoholic neuropathy Epidemiology • Alcohol abuse and diabetes are the commonest causes of peripheral neuropathy in the United Kingdom. Pathophysiology • Typically, all fibre types are affected and it is seen with a higher alcohol consumption more than 30 units. • affects mainly the spinothalamic pathway. • secondary to both direct toxic effects and reduced absorption of B vitamins (thiamine deficiency) Features • slowly progressive • sensory symptoms typically present prior to motor symptoms • Pain is usually a more dominant feature Treatment • thiamine and cessation of alcohol use

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Peripheral neuropathy: axonal vs. demyelinating Peripheral neuropathy Causes Nerve conduction studies • alcohol • isoniazid • Simvastatin Axonal • Diabetes mellitus* • vasculitis • vitamin B12 deficiency* • Renal failure • hereditary sensorimotor neuropathies (HSMN) type II (*may also cause a demyelinating picture) • Guillain-Barre syndrome • chronic inflammatory demyelinating polyneuropathy (CIDP) • Paraproteinaemia • Amiodarone (Amiodarone can cause a mixed demyelinating and axonal picture) Demyelinating • Refsum's disease • hereditary sensorimotor

neuropathies (HSMN) type I (Charcot-Marie-Tooth disease) • Leukodystrophies. • Nerve conduction studies (NCS) are useful in determining between axonal and demyelinating pathology • Segmental demyelination is a feature seen in axons in the central nervous system with multiple sclerosis. Wallerian degeneration • Wallerian degeneration is degeneration of the portion of the nerve distal to the injury. • It occurs following axonal injury in both the peripheral and central nervous systems • usually begins within 24-36 hours of injury. Electromyogram (EMG) • A pattern of rapidly recruited low amplitude short duration motor units on the electromyogram (EMG) would be considered to represent myopathic changes rather than de-innervation. Notes & Notes for MRCP
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(NCS) • normal conduction velocity • reduced amplitude • reduced conduction velocity • normal amplitude

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Drugs causing peripheral neuropathy • Antibiotics: nitrofurantoin, metronidazole • Amiodarone • Isoniazid • Vincristine • Tricyclic antidepressants Critical illness polyneuropathy • Prolonged periods in the Intensive Therapy Unit, irrespective of the underlying pathology, are associated with a risk of developing critical illness polyneuropathy • It is an axonal neuropathy and thus muscle wasting may occur • May be predominantly sensory, predominantly motor or mixed

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Chronic inflammatory demyelinating polyneuropathy (CIDP) Chronic inflammatory demyelinating polyneuropathy is clinically similar to Guillain-Barre syndrome (hyporeflexia or areflexia, paraesthesia and mild sensory deficits in the upper and lower extremities, weakness) except that it follows a chronic progressive course. Overview • CIDP is characterised by progressive weakness and impaired sensory function in the upper and lower limbs. • subacute sensory and motor peripheral neuropathy • The cause of the demyelination is not understood, • More common in young adults and in men. • mainly causes motor impairment (distal and proximal). • CIDP causes a large fibre peripheral neuropathy (Joint position sense and vibration are carried through large fibres) Features • weakness of the limbs • areflexia • abnormal sensation (which typically begins distally) • fatigue. • Autoantibodies against GM1 gangliosides Differential diagnosis

1. CIDP is closely linked to Guillain-Barré syndrome (GBS), and is thought by some to be its chronic counterpart. Both CIDP and GBS can affect motor and sensory nerves (GBS) is an acute (which reaches its peak in severity within six weeks), postinfectious neuropathy Whereas CIDP is subacute (several months history)
2. Hereditary motor and sensory neuropathy (HMSN) is normally a very chronic neuropathy developing over many years and usually with a family history of the condition. Treatment • Corticosteroids • plasmapheresis • Intravenous immunoglobulin • Physiotherapy

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Diabetic neuropathy(see endocrinology system)

Neuropathic pain Definition • neuropathic pain may be defined as pain which arises following damage or disruption of the nervous system. • It is often difficult to treat and responds poorly to standard analgesia. Examples include: • diabetic neuropathy • post-herpetic neuralgia • trigeminal neuralgia • prolapsed intervertebral disc Management of neuropathic pain • first-line treatment: amitriptyline, duloxetine, gabapentin or pregabalin please note that for some specific conditions the guidance may vary. For example carbamazepine is used first-line for trigeminal neuralgia • if the first-line drug treatment does not work try one of the other 3 drugs • tramadol may be used as 'rescue therapy' for exacerbations of neuropathic pain • topical capsaicin may be used for localised neuropathic pain (e.g. post-herpetic neuralgia) • pain management clinics may be useful in patients with resistant problems January 2019 exam: severe 'shooting' pains after blistering rash. What is the most appropriate next step in management? Amitriptyline

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Autonomic neuropathy Features • impotence, inability to sweat, postural hypotension • postural hypotension e.g. drop of 30/15 mmHg • loss of decrease in heart rate following deep breathing • pupils: dilates following adrenaline instillation Causes • diabetes • Guillain-Barre syndrome • multisystem atrophy (MSA), Shy-Drager syndrome • Parkinson's • infections: HIV, Chagas' disease, neurosyphilis • drugs: antihypertensives, tricyclics • craniopharyngioma

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Hereditary sensorimotor neuropathy (HSMN) (Charcot-Marie-Tooth disease) Mixed motor and sensory symptoms, slowly progressing initially in the lower limbs and then to the upper limbs, together with a family history suggests a diagnosis of Hereditary sensorimotor neuropathy (HSMN) Definition • hereditary nerve disorders with defective production of peripheral myelin protein-22 which is involved in the structure and function of the myelin sheath. • Charcot-Marie-Tooth disease is the most commonly inherited neurological disorder, Genetics • autosomal dominant • caused by deletion in the PMP22 gene, the same gene mutation responsible for hereditary neuropathy with liability to pressure palsies. • Common peroneal nerve is the most commonly affected nerve (36%) followed by the ulnar nerve (28%). Types • HSMN type I the most common form primarily due to demyelinating pathology hence C fibres are not affected, as they are unmyelinated. Which nerve fibers are relatively preserved in this patient? C fibers due to defect in PMP-22 gene (which codes for myelin) loss of myelin in peripheral neurons features often start at puberty motor symptoms predominate distal muscle wasting, pes cavus, clawed toes foot drop, leg weakness often first features • HSMN type 2 primarily due to axonal pathology loss of peripheral neurone themselves Features • motor and sensory deficits. • early weakness of the distal muscles of the limbs. • scoliosis • pes cavus a deformity of the foot involving high arches, muscle wasting and clawed toes. Diagnosis • neurophysiology: Electromyography (EMG) and nerve conduction studies (NCS) may distinguish between the demyelinating (type 1) and axonal (type 2) forms. • Diagnosis confirmed by genetic testing. • Nerve biopsy, usually the sural nerve, will demonstrate "onion-bulb" formations due to continual remyelination and demyelination of peripheral nerves.

Neurology Management • The mainstay of the management is physical therapy. Prognosis • Life expectancy is normal. MRCPUK-part-1-September 2017 exam: A woman with Charcot-Marie-Tooth disease (type 1), how likely her children will get the disease? □ 50% (autosomal dominant)

Mononeuritis multiplex Definition : ≥ 2 isolated mononeuropathies Causes • Axonal injury caused by damage to vasa nervorum • Occurs in conditions characterized by the development of granulomas and/or microangiopathy (e.g., diabetes mellitus, rheumatoid arthritis, vasculitides, SLE, Lyme disease, amyloidosis, HIV, polyarteritis nodosa) Features: painful, asymmetrical sensory and motor symptoms Diagnosis: Nerve biopsy should be performed to confirm the diagnosis Treatment: includes prednisolone and cyclophosphamide

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Refsum's disease Overview • autosomal recessive disorder • caused by defective alpha oxidation of phytanic acid leading to its accumulation in tissues. • Phytanic acid is present in a wide variety of foods including dairy products, fish, beef and lamb. • The onset of the disease is normally in the late teens or 20s.
Features • sensorimotor peripheral neuropathy • sensorineural deafness, • anosmia, • cerebellar ataxia • pes cavus. • Night blindness and visual problems occur secondary to retinitis pigmentosa. • Cardiac conduction abnormalities and cardiomyopathies may also occur. • Epiphyseal dysplasia causes a characteristic shortening of the fourth toe. • Serum phytanic acid levels are elevated. Treatment • dietary restriction of foods containing phytanic acid.

Vasculitic neuropathy Overview • The presence of nail fold infarcts and the multifocal nature of the neuropathy indicate that a vasculitic cause is most likely • Hepatitis C infection may be associated with cryoglobulinaemia, which causes a vasculitic syndrome including neuropathy

Other conditions associated with vasculitic neuropathy include • Polyarteritis nodosa • Churg-Strauss syndrome • rheumatoid arthritis • systemic lupus erythematosus • systemic sclerosis • Wegener's granulomatosis Treatment include one or several of the following • high-dose intravenous steroids • plasma exchange • intravenous immunoglobulins

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Guillain-Barre syndrome FVC is used to monitor respiratory function in Guillain-Barre syndrome • also known as Post-infectious polyradiculopathy Definition • Guillain-Barre syndrome describes an immune mediated demyelination of the peripheral nervous system often triggered by an infection : □ classically Campylobacter jejuni □ cytomegalovirus Pathogenesis • cross reaction of antibodies with gangliosides in the peripheral nervous system • correlation between anti-ganglioside antibody (e.g. anti-GM1) and clinical features has been demonstrated • anti-GM1 antibodies in 25% of patients Features • characteristic features □ progressive weakness of all four limbs. The weakness

is classically ascending i.e. the lower extremities are affected first, however it tends to affect proximal muscles earlier than the distal ones. Sensory symptoms tend to be mild (e.g. distal paraesthesia) with very few sensory signs. However, a sensory level is NOT a feature and would suggest cervical myelopathy symmetrical involvement is typical, asymmetry present in only 9% of patients. Some patients experience back pain in the initial stages of the illness. • Other features areflexia cranial nerve involvement e.g. diplopia autonomic involvement: e.g. urinary retention Muscle wasting is typical with prolonged illness. Bulbar involvement occurs in 50%, with a risk of aspiration and respiratory insufficiency urinary incontinence or retention (in 20% of cases). • Less common findings papilloedema: thought to be secondary to reduced CSF resorption

Investigations • CSF analysis elevated protein, with normal glucose and no pleocytosis. a rise in CSF protein doesn't peak until the second or third week of the illness. CSF cell counts are usually within normal limits, • Nerve conduction studies (including F waves for the proximal spinal root, looking for widespread demyelination) • MRI may be indicated to rule out spinal cord lesions, peripheral neuropathies and neuromuscular junction disorders. Management • IV immunoglobulins (IVIG): First line therapy. as effective as plasma exchange. No benefit in combining both treatments. IVIG may be easier to administer and tends to have fewer side-effects • plasma exchange • steroids and immunosuppressants have not been shown to be beneficial • FVC regularly to monitor respiratory function . FVC of less than 1 litre would be an indication for immediate ventilation Forced vital capacity of 1.4 L is most likely to predict the need for invasive ventilation FVC of less than 15ml/kg (or less than 30% of FVC predicted) or a rising PaCO2 are indications for mechanical ventilation. Prognosis • 20% suffer permanent disability, 5% die • Poor prognostic features age > 40 years poor upper extremity muscle strength previous history of a diarrhoeal illness (specifically Campylobacter jejuni) high anti-GM1 antibody titre need for ventilatory support There is currently contradictory evidence as to whether a gradual or rapid onset of GBS is associated with a poor outcome MRCPUK-part-1-January 2008 exam: Regarding nerve conduction studies for suspected Guillain-Barre syndrome. Which finding would be most consistent with this diagnosis? Reduced conduction velocity MRCPUK-part-1-May 2019 exam: a patient developed weakness in his legs extended to his arms after viral illness. ↓↓ power, reflexes and sensation in his lower limbs. Developed SOB &↓↓ (FVC). Given the likely diagnosis, what is the treatment of choice? Intravenous immunoglobulin (Guillain-Barre syndrome (GBS) secondary to a viral illness, possibly the Epstein-Barr virus) MRCPUK-part-1-May 2020 exam: H/O double vision & ↓↓ eye movement + unsteadiness +
↓↓ reflexes + past-pointing. What is the most likely diagnosis? Miller Fisher syndrome

Miller Fisher syndrome • areflexia, ataxia, ophthalmoplegia • variant of Guillain-Barre syndrome • associated with ophthalmoplegia, areflexia and ataxia. The eye muscles are typically affected first

• usually presents as a descending paralysis rather than ascending as seen in other forms of Guillain-Barre syndrome • anti-GQ1b antibodies are present in 90% of cases

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DVLA: neurological disorders • The guidelines below relate to car/motorcycle use unless specifically stated. • For obvious reasons, the rules relating to drivers of heavy goods vehicles tend to be much stricter Specific rules • First seizure: 6 months off driving*. □ *previously rule was 12 months. It is now 6 months off driving if the licence holder has undergone assessment by an appropriate specialist and no relevant abnormality has been identified on investigation, for example EEG and brain scan where indicated • For patients with established epilepsy they must be fit free for 12 months before being able to drive • Stroke or TIA: 1 month off driving • Multiple TIAs over short period of times: 3 months off driving • Craniotomy e.g. For meningioma: 1 year off driving □ if the tumour is a benign meningioma and there is no seizure history, licence can be reconsidered 6 months after surgery if remains seizure free • Pituitary tumour: □ craniotomy: 6 months; □ trans-sphenoidal surgery 'can drive when there is no debarring residual impairment likely to affect safe driving' • Glioblastoma □ A patient with a high-grade glioma (that is, WHO grade 3 or 4) such as a glioblastoma will be unable to drive for at least two years following completion of treatment. □ After the two years have elapsed the DVLA will consult with the physicians involved in the patient's care and a decision is made regarding return of the licence. • Brain metastases □ solitary metastatic deposit that is fully excised would be considered for a licence one year after primary treatment if free from recurrence and no evidence of secondary spread elsewhere.

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Neurology □ multiple metastases would require at least two years off driving from time of completion of treatment. After the two years have elapsed the DVLA will consult with the physicians involved in the patient's care and a decision is made regarding return of the licence. • Narcolepsy/cataplexy: □ cease driving on diagnosis, □ can restart once 'satisfactory control of symptoms' • Chronic neurological disorders e.g. multiple sclerosis, motor neuron disease: □ DVLA should be informed, □ complete PK1 form (application for driving licence holders state of health) • Syncope □ simple faint: no restriction □ single episode explained and treated: 4 weeks off □ single episode, unexplained: 6 months off □ two or more episodes: 12 months off

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Susac syndrome • Susac syndrome presents with the triad of: □ Encephalopathy □ branch retinal artery occlusion □ and hearing loss • Due to involvement of the pre-capillary arterioles of the brain, retina and cochlea.

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Altitude related disorders Types • There are three main types of altitude related disorders: • All three conditions are due to the chronic hypobaric hypoxia which develops at high altitudes

1. acute mountain sickness (AMS), Features of AMS start to occur above 2,500 - 3,000m, developing gradually over 6-12 hours and potentially last a number of days headache nausea fatigue Prevention and treatment of AMS the risk of AMS may actually be positively correlated to physical fitness gain altitude at no more than 500 m per day acetazolamide (a carbonic anhydrase inhibitor) is widely used to prevent AMS and has a supporting evidence base Treatment: Descent generally a self-limiting condition.
2. high altitude pulmonary edema (HAPE) A minority of people above 4,000m go onto develop high altitude pulmonary oedema (HAPE) potentially fatal conditions HAPE presents with classical pulmonary oedema features Management of HAPE descent nifedipine, dexamethasone, acetazolamide, phosphodiesterase type V inhibitors All seem to work by reducing systolic pulmonary artery pressure oxygen if available
3. high altitude cerebral edema (HACE). A minority of people above 4,000m go onto develop high altitude cerebral oedema (HACE), potentially fatal conditions HACE presents with headache, ataxia, papilloedema Management of HACE descent dexamethasone

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Complex regional pain syndrome (CRPS) • (CRPS) is the modern, umbrella term for a number of conditions such as reflex sympathetic dystrophy and causalgia. • (CRPS) is a chronic pain condition that can affect any area of the body, but often affects an arm or a leg, and occurs after an injury or rarely after a sudden illness such as a heart attack or stroke. typically occur following surgery or a minor injury. • The condition can sometimes appear without obvious injury to the affected limb. • 3 times more common in women.

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Neurology • CRPS may have three stages (acute, dystrophic, and atrophic), with variable progression from one stage to another. There are two types of CRPS: • type I (most common): there is no demonstrable lesion to a major nerve • type II: there is a lesion to a major nerve Character of the pain • intense and burning • disproportionate to the original injury • worse over time • Spreads beyond the site of injury and • associated with hyperalgesia, hyperpathia or allodynia on examination. These features do not occur in DVT, osteomyelitis, or cellulitis. Features • progressive, disproportionate symptoms to the original injury/surgery • allodynia • temperature and skin colour changes • oedema and sweating • motor dysfunction • the Budapest Diagnostic Criteria are commonly used in the UK Diagnosis • clinical diagnosis • Plain radiographs may show soft tissue swelling, peri-articular osteoporosis, and rarely erosions • MRI may also show bone marrow oedema apart from these changes In the atrophic phase, imaging may show contractures. • 99mTc bone scan shows hypervascularity in the acute phase, and hypovascularity in the Management • early physiotherapy is important • neuropathic analgesia in-line with NICE guidelines • specialist management (e.g. Pain team) is required

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Dystonia Definition: • involuntary sustained or spasmodic muscle contractions Types • Focal dystonias Involves a single body part Cervical dystonia, or torticollis, is the most common focal dystonia. In 20-30% of patients, focal dystonias become segmental or multifocal. Blepharospasm is a type of dystonia described as a sustained eyelid twitch. It is associated with

stress, lack of sleep, nutrition, and strain. writer's cramp dystonia or musician's dystonia A common upper limb dystonia This task-specific dystonia, manifesting as hyperextension or hyperflexion of the wrist and fingers, unable to write may be triggered by repetitive activities such as writing and attempting to play the piano or other musical instruments.

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 often relieved by a geste antagoniste, in which palpation of another unaffected part of the body leads to relief of symptoms, thought to be a result of alternative sensory input to cortical networks with altered plasticity. • Segmental dystonia Affects 2 or more contiguous regions of the body • Multifocal dystonia Consists of abnormalities in noncontiguous body parts • Generalised dystonias, involve a greater number of muscle groups. involves the trunk and limbs. Treatment • Benztropine is an anti-cholinergic drug that is used in the treatment of Parkinson's disease, Parkinsonism, and acute dystonia.

Cervical dystonia (torticollis) • The term torticollis is derived from the Latin words tortus for twisted neck • Torticollis is a fixed or dynamic tilt, rotation, or flexion of the head and/or neck. • involuntary neck movements • commonly affects women • Secondary causes need to be excluded such as drugs (eg neuroleptics) and cervical spine abnormalities • Botulinum toxin injection is the first-line treatment for cervical dystonia (torticollis)

Botulism Descending weakness with autonomic dysfunction (fixed dilated pupils) is typical of botulism. Definition • Botulism is a neurological disorder caused by Clostridium botulinum and is characterized by flaccid paralysis due to inhibition of acetylcholine release at the neuromuscular junction. Features • The clinical presentation of descending weakness with autonomic dysfunction (fixed dilated pupils) is typical of botulism. • Typical initial features include: Diplopia Ptosis Facial weakness Dysarthria, and Dysphagia. • Later, respiratory difficulty and limb weakness occur. • impaired cholinergic transmission also involves autonomic synapses, causing poorly reactive dilated pupils, dry mouth, paralytic ileus and occasionally bradycardia. • Reflexes are depressed or absent,

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Neurology Investigations • It is a neuromuscular junction disorder and therefore nerve conduction studies and EMG are normal. • Cerebrospinal fluid analysis is usually normal. • Repetitive nerve stimulation shows incremental responses, which is diagnostic of botulism. • sensation is normal Treatment • Heptavalent antitoxin is the most appropriate therapy

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Botulinum toxin • Botulinum toxin is produced by Clostridium botulinum, a Gram-positive, sporeforming, obligate anaerobe • Botulinum toxin type A (or trade name Botox®) Action • block acetylcholine release at the neuromuscular junction and so to produce muscle weakness. • myasthenia gravis would be expected to worsen with this treatment Indications • Botulinum toxin

is the treatment of choice for focal dystonia (such as torticollis, and hemi-facial spasm) and focal dystonia. • Botulinum toxin injections are also used in patients with: hemifacial spasm blepharospasm spasticity spasticity associated with stroke spasticity associated with cerebral palsy Primary axillary hyperhidrosis Strabismus Cervical dystonia. Side effects • Occasionally systemic absorption of the toxin can affect distal muscles causing symptoms such as diplopia and dysphagia. • The main side-effect is excessive weakness in the treated muscle Contra-indications • myasthenia gravis • other generalised muscle conditions

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Paraneoplastic cerebellum syndrome The patient with progressive ataxia and dysarthria following malignancy Causes • Associated malignancies are lung cancer (usually with small cell lung carcinoma), breast cancer, ovarian cancer and lymphoma Features • include ataxia, dysarthria, vertigo, oscillopsia, nystagmus and dysmetria Investigations • Brain imaging and CSF analysis are either normal or show non-specific changes

• antibodies anti-Hu antibody (a type of antineuronal antibody) anti-Purkinje-cell antibodies • CT chest, abdomen and pelvis and mammogram are required to look for a primary neoplasm. • A whole body positron emission tomography (PET) scan is preferable but not widely available. Treatment • Occasionally patients respond to steroids, immunoglobulins or plasmapheresis.

Lumbosacral plexopathy The patient presents with generalised weakness of the right leg associated with pelvic pain, leg oedema and autonomic dysfunction. The most likely diagnosis is a lumbosacral plexopathy. Overview • Anatomically, the lumbosacral plexus consists of lumbar (L1-L4) and sacral (S1-S5) portions. • Upper: lumbar plexus lesion will cause weakness of hip flexion and adduction of the thigh and extension of the leg with anaesthesia over the anterior thigh and leg. • Lower: sacral plexus lesions will weaken the posterior thigh and foot muscles. • Lesions affecting the entire plexus will affect all muscle groups causing weakness or paralysis of the leg, areflexia and anaesthesia from the toes, to involve the perianal area. Causes • Trauma: Posterior hip dislocation, Sacral fracture • Metabolic, inflammatory, and autoimmune causes: DM (diabetic amyotrophy), Amyloidosis, Sarcoidosis • Infections and local abscess (e.g. vertebral osteomyelitis, tuberculosis, fungal infections, psoas abscess) • Radiation therapy of the abdominal and pelvic malignancies. • Pregnancy-related: Mostly occur in the third trimester and after delivery due to birth trauma. • Damage to the vasculature innervating the LS plexus: femoral vessel catheterization Epidemiology • More common in women due to the predisposing risk factors of pregnancy and gynecological cancers. Pathophysiology • Direct injury, compression or traction on the plexus (Trauma, tumor, hematoma • Microvascular injury and ischemic damage (Radiation) • Inflammatory or microvascular changes (Diabetic and non-diabetic) Features • Low back pain radiating to one side. Pain may be positional, worse in a supine position. Patients with diabetic LS plexopathy (diabetic amyotrophy) typically complain of unilateral pain in the proximal thigh. lumbosacral plexopathy secondary to radiotherapy is usually painless.

Neurology • Muscle weakness and atrophy may occur in severe cases. • A straight leg raise test is positive in more than half of the patients. • Knee jerk reflex is affected in lumbar plexopathy and ankle jerk is affected in sacral plexopathy. • Muscle weakness in hip flexion, knee extension, or adduction suggests a possible injury to the lumbar plexus. • Sensory changes ((numbness, paresthesias, dysesthesias (painful sensations elicited by nonpainful cutaneous stimuli, e.g., light touch)). Medial thigh, anterior thigh, and medial suggest lumbar plexus involvement Posterior thigh, dorsum of the foot, and perineum suggest sacral plexus involvement. A history of a road traffic accident, abdominopelvic neoplasm, radiotherapy, abdominal surgery, diabetes mellitus, bleeding disorders, or recent pregnancy hints towards lumbosacral plexopathy and narrows down the etiology. Radiation plexopathy can often present without pain, only weakness and sensory changes. Unlike other types of plexopathy, it is usually bilateral and can occur even years after radiation. Diagnosis • MRI with gadolinium contrast is the best test for the evaluation of the LS plexus. • When there are contraindications to MRI (e.g., a noncompatible pacemaker), a computed tomography (CT) scan with contrast can be utilized. • Electromyography (EMG) differentiate lumbosacral plexopathy from other types of neuropathy or radiculopathies. Denervation of the paraspinal muscles is commonly seen in radiculopathy and helps to differentiate from lumbosacral plexopathy. Treatment • Treatment of underline cause: e.g. relieve of compression • Symptomatic treatment with analgesics and muscle relaxants: Compression: NSAIDs, opioids Neuropathic pain: pregabalin, gabapentin, duloxetine, amitriptyline Diabetic amyotrophy is a transient condition that usually resolves with good glycemic control. • For radiation-induced plexopathy: there are no known treatments , physiotherapy and rehabilitation are the mainstays of treatment. Further radiotherapy sessions should be discontinued.

Notes & Notes for MRCP
By Dr. Yousif Abdallah Hamad

Cervical roots Root Dermatome distribution Myotome distribution Tendon reflex C2 Posterior half of the skull (cap)

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C3 High turtleneck shirt

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C4 Upper outer shoulder, Low-collar shirt Shoulder abduction Nil C5 Outer arm, forearm Shoulder abduction, elbow flexion Bicep C6 Index and thumb Wrist extension Supinator C7 Middle finger centre of palm Finger and elbow extension Triceps C8 Little and ring finger, ulnar border of hand Wrist/finger flexion Finger jerk Symptoms and signs of a C6 root lesion include • Paraesthesias in the thumb or lateral distal forearm • Weakness of brachioradialis, biceps, or triceps and • Diminished biceps and brachioradialis reflexes in conjunction with an increased triceps reflex. Spinal lesion at the level of C8: • Weakness of finger flexion + Loss of sensation over the medial aspect of the arm; forearm and hand (Lateral aspect of arm is C5) Erb-Duchenne palsy ('waiter's tip') • due to damage of the upper trunk of the brachial plexus (C5,C6) • may be secondary to shoulder dystocia during birth • the arm hangs by the side and is internally rotated, elbow extended Weakness of shoulder abduction • May be due to C5 or an axillary nerve lesion: C5 lesion weakness of biceps (C5, C6) loss of the sensation of the lateral aspect of the upper arm Axillary nerve lesion spinal root : C5/C6 C6: Make a 6 with your left hand by touching the tip of the thumb & index finger together Winging of the scapula is caused by paralysis of the long thoracic nerve to serratus anterior (C5, 6, 7).

Chapter 4

Neurology

motor function: innervate teres minor and deltoid muscles sensory function: give rise to superior lateral cutaneous nerve of arm which innervate the skin over the lower deltoid (regimental badge area) loss of sensation of the regimental badge area • Absence of sensory loss indicates

a lesion at the anterior horn cell. Thoracic roots Root Dermatome Myotome Reflex T 4 Nipples T 5 Inframammary fold T 7 Xiphoid process T 10 Umbilicus T1 nerve root injury: damage to both the median and ulnar nerves Global muscle wasting of the hand Lumber roots Root Dermatome Myotome Reflex L1 Inguinal ligament

-

L2 Upper anterior and medial thigh Psoas hip abductor

L3 Mid anterior and medial thigh Psoas quadriceps Patella (L3,L4) L4 Knee caps, medial aspect of leg, lower lateral thigh L5 Big toe, dorsum of foot (except lateral aspect), lateral aspect of leg L1: Inguinal ligament (L for ligament, 1 for 1nguinal) The L4 dermatome is located at the knee caps L5 = Largest of the 5 toes L5 lesion features: loss of foot/big toe dorsiflexion + sensory loss dorsum of foot Notes & Notes for MRCP

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Tibialis anterior, extensor halluces Patella (L3,L4) Extensor halluces, peroneal, gluteus medias, dorsiflexors, hamstrings L5 has no reflex. Therefore, an acute lumber disc prolapse resulting in L5 radiculopathy is commonly misdiagnosed as malingering.

Sacral roots Root Dermatome Myotome Reflex S1 Lateral foot, small toe. sole of the foot, Posterior, lateral thigh and calf S2 Popliteal fossa

Ankle (S1, S2) S3 -5 Medial buttock and perianal skin in a concentric manner with S3 most lateral and S5 closest to the anus Deep tendon reflexes: which test for which nerve root? C5 – Biceps C6 – Biceps, Brachioradialis C7 – Triceps

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L4 – Patellar (knee jerk) (femoral nerve mediated) S1 – Achilles (ankle jerk) (tibial nerve mediated) Notes & Notes for MRCP
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Peroneal planter flexor Ankle (S1, S2) Bladder, rectum S2-4 reflex is part of the anocutaneous reflex or anal wink.

Notes & Notes for MRCP
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Neurology

Inverted brachioradialis reflex (inverted supinator jerk): • An inverted supinator jerk, where the biceps jerk is absent but generates a supinator jerk with reflex flexion of the fingers, is indicative of cervical myelopathy with C5/6 nerve root damage. MRCPUK-part-1-September 2019 exam: H/O neck & arm pain like 'electric shocks', worse on turning head + decreased sensation on the dorsal aspect of the thumb and index finger. What is the most likely underlying diagnosis? C6 radiculopathy MRCPUK-part-1-septemer-2017: Which nerve (and its nerve root) are you tested in triceps reflex? Radial nerve C7 Which spinal dermatome is responsible for the initial vague periumbilical discomfort in appendicitis? T10 mrcpuk.org SCE sample question: H/O pain affecting buttock region and the lateral border and sole of his foot, in association with paraesthesiae of the sole on walking. What is the correct nomenclature for the nerve root from which these symptoms have arisen? S1 (The S1 nerve root is mapped to the sole of the foot)

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