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Notes & Notes for MRCP
By Dr. Yousif Abdallah Hamad

Disc prolapse Loss of sensation in the upper outer thigh is consistent with nerve root compression caused by a prolapsed vertebral disc. Pathophysiology • The intervertebral disk consists of a dense outer ring (annulus fibrosus) and a gelatinous core (nucleus pulposus). • disk protrusion or herniation through the annulus fibrosus into the central canal → adjacent nerve root impingement → sensorimotoric deficits in affected nerve root • The herniation of the nucleus pulposus is most commonly in the posterolateral direction as it is the weakest part of the surrounding annulus fibrosus. • The affected nerve root is typically the one below the level of disc herniation Common sites of prolapse • Most often occurs in the lumbar spine (95% of disc herniations occur at the L4-L5 and L5-S1 level). L5–S1 (most common site) L4–L5 (second most common site) • Cervical and thoracic disc herniations are rare Causes • Disc degeneration (the most common cause) • Trauma Features • Acute onset of severe neck or back pain Radicular pain: pain that radiates to the legs (sciatic pain) or arms The pain is either stabbing in nature or resembles an electric shock • Features of radiculopathy: lower motor neuron signs of the affected nerve root (typically unilateral) Paresthesia of the affected dermatome Muscle weakness Absent or diminished deep tendon reflexes • Character of pain Pain increases with pressure (e.g., from coughing or sneezing) Pain is typically better with rest: if it is unremitting or worse on resting you should consider other causes such as bony metastases or infection. Changing position reduces the pain Management • Gentle mobilisation and physiotherapy (the management of choice): most patients will make a spontaneous improvement within 4–6 weeks. • Surgery (Microdiscectomy or open discectomy) is a potential treatment options for patients with radiologically proven nerve root compression and severe symptoms or symptoms that do not resolve with conservative measures. • Local corticosteroid injection: symptomatic relief if not fit for surgery Intervertebral discs usually protrude/herniate posterolaterally, as the posterior longitudinal ligament is thinner than the anterior longitudinal ligament.

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Neurology This table demonstrates the expected features according to the level of compression Level of compression Features L3 nerve root compression Sensory loss from anterior thigh to medial aspect of lower leg Weak quadriceps ↓ knee reflex Positive femoral stretch test L4 nerve root compression Caused by L3/4 disc prolapse Sensory loss over the thigh and anterior

aspect of knee Weak quadriceps ↓ knee reflex Positive femoral stretch test L5 nerve root compression Caused by L4/5 disc prolapse Sensory loss dorsum of foot and lateral aspect of leg Weakness in foot and big toe dorsiflexion ('foot drop') Reflexes intact Positive sciatic nerve stretch test S1 nerve root compression Caused by L5/S1 disc prolapse Sensory loss posterolateral aspect of leg (posterior calf and the plantar surface of the foot) and lateral aspect of foot Weakness in plantar flexion of foot ↓ ankle reflex Positive sciatic nerve stretch test

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Prolapsed cervical disc (Cervical radiculopathy) Overview • Most commonly affects the C5/C6 and C6/C7 vertebrae. • Central protrusions can lead to symptoms of spinal cord compression. • Posterolateral protrusions can cause a stiff neck, pain radiating to the arm, weakness of the muscles affected by the nerve root and depressed reflexes. • X-ray may shows narrowing of the disc space between the C5 and C6 vertebrae. Deferential diagnosis • Cervical spondylosis occurs as a result of osteoarthritis. Muscle weakness is uncommon X-ray changes: Disc spaces can be narrowed Osteophytes seen in the central and posterior intervertebral joints. • Cervical rib can present with similar symptoms but the X-ray would be diagnostic (showing the presence of a cervical rib).

• Spasmodic torticollis sudden onset of a stiff painful neck with torticollis can occur in adults due to spasm of the trapezius and sternocleidomastoid muscles. X-ray of the cervical spine is usually normal. Common cervical radiculopathies Sensory deficits Motor deficits Reduction of C5 radiculopathy Anterior shoulder Biceps and deltoid Biceps From upper lateral elbow over radial forearm up to thumb and radial side of index finger C6 radiculopathy C7 radiculopathy Palmar: fingers II–IV (II ulnar half, III entirely, IV radial half) Dorsal: medial forearm up to fingers II–IV (II ulnar half, III entirely, IV radial half) C8 radiculopathy Dorsal forearm up to dorsal and palmar area of fingers IV (ulnar half)

Conus medularis syndrome

Conus medullaris syndrome is caused by compression of the T12-L2 cord and nerve roots, and therefore results in a mix of upper and lower motor neuron signs.

• Mixed upper and lower motor neurone signs. These include bilateral distal weakness with increased tone and hyper-reflexia, fasciculation, positive Babinski sign and clonus. Cauda equina would give just LMN signs, • Sensory loss is most marked in the perianal region. In Amyotrophic lateral sclerosis (the commonest form of motor neurone disease), There would be a mixture of UMN and LMN signs; however, they do not have any sensory signs or incontinence. • It is much rarer than cauda equina syndrome. Notes & Notes for MRCP
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reflexes Biceps and wrist extensors Biceps Brachioradialis Triceps and wrist flexors, finger extensors Triceps Finger flexors None

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Conus medullaris syndrome VS Cauda equina syndrome Conus medullaris syndrome Cauda equina syndrome Presentation Sudden and bilateral Gradual and may be unilateral leg sings initially Reflexes Knee jerk preserved Ankle jerk affected Both knee and ankle jerk affected Radicular pain Less severe More severe Sensory Numbness often localised peri-anal area (often bilateral) Numbness often localised saddle area (often unilateral) Motor Symmetrical Upper motor signs (hyperreflexic distal paresis, less than cauda equina, may be fasciculation) May be asymmetrical Lower motor signs (areflexic paraplegia, atrophy, fasciculations is rare) Impotence Frequent Often less marked Sphincter disfunction Urinary retention and atonic anal sphincter present early in disease (can cause overflow urinary incontinence) Urinary retention usually present later in course of disease Low back pain More marked Less marked Conus medullaris syndrome and cauda equina syndrome are medical emergencies requiring immediate surgical intervention.

Cauda equina syndrome Causes • herniation of a lumbar disc (at L4/L5 and L5/S1) • tumour (metastases, lymphoma, primary spinal tumours) • trauma • infection (epidural abscess). • Others: ankylosing spondylitis, Paget’s disease, and congenital spinal stenosis. Features • lower motor neuron signs: flaccid paraplegia, areflexia, flexor plantar reflexes • unilateral or bilateral lower limb motor and/or sensory abnormality • low back pain • Whilst classically patients present with a sensory level, this is variable in clinical practice. • bladder retention and overflow incontinence (bowel and/or bladder dysfunction with saddle and perineal anaesthesia) • Saddle anesthesia. Patients usually describe numbness and/or "pins-and-needles" sensations of the groin and inner thighs which would contact a saddle when riding a horse. This reflects involvement of the S3-S5 roots. Diagnosis • MRI is the investigation of choice Cauda equina syndrome is caused by compression of the lumbosacral roots, from L1 down to S5, and therefore results in only lower motor neuron signs.

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Autonomic dysreflexia Definition • A clinical syndrome occurs in patients who have had a spinal cord injury at, or above T6 spinal level (85% of patients). Mechanism • A strong sensory input (most commonly urine retention or constipation) travels up the spinal cord massive reflex sympathetic surge from the thoracolumbar sympathetic nerves widespread vasoconstriction, most significantly in the subdiaphragmatic (or splanchnic) vasculature hypertension crisis • The brain detects this hypertensive crisis through intact baroreceptors in the neck delivered to the brain through cranial nerves IX and X. • The brain attempts two maneuvers to decrease BP:

1. by sending descending inhibitory impulses of sympathetic surge which are unable to travel because of the spinal cord injury at T6 or above.
2. by slowing the heart rate through an intact vagus (parasympathetic) nerve; however, this compensatory bradycardia is inadequate, and hypertension continues. • In summary, the sympathetics prevail below the level of neurologic injury, and the parasympathetic nerves prevail above the level of injury. Triggers • urinary retention (cystitis, retention of urine or a blocked catheter): most common • constipation (faecal impaction) Features •

unbalanced physiological response, characterised by : extreme hypertension , may leads to complications flushing and sweating above the level of the cord lesion Agitation Bradycardia Treatment • recognition and removal of the triggers. • Vasodilators such as calcium antagonists may be used to treat the hypertension.

Spastic paraparesis Definition • Spastic paraparesis describes an upper motor neuron pattern of weakness in the lower limbs Causes • demyelination e.g. multiple sclerosis • cord compression: trauma, tumour • parasagittal meningioma (Spinal meningioma) progressive symptoms (not acute), well-defined sensory level MRI of the spine with gadolinium contrast is the investigation of choice • tropical spastic paraparesis classic presentation HTLV-1 positive patient presenting with paraparesis and urinary retention due to Adult T-cell lymphoma (ATL) caused by human Tlymphotropic virus type 1 (HTLV-I)

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Neurology • transverse myelitis e.g. HIV • syringomyelia • hereditary spastic paraplegia • osteoarthritis of the cervical spine Sudden onset Progressive onset Anterior spinal artery infarct Osteoporotic thoracic spine collapse Prolapsed thoracic disc demyelination e.g. multiple sclerosis Metastatic carcinoma Spinal meningioma Absent ankle jerks, extensor plantars Overview • Typically caused by lesion producing both upper motor neuron (extensor plantars) and lower motor neuron (absent ankle jerk) signs • Mixture of UMN and LMN signs Causes • subacute combined degeneration of the cord • motor neuron disease • Friedreich's ataxia (usually presents by age 30) • Syringomyelia • taboparesis (syphilis) • HIV • Spinal AVM • conus medullaris lesion Which neurological finding is most helpful in differentiating subacute combined degeneration of the cord from multiple sclerosis? Absent ankle jerk

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Subacute combined degeneration of spinal cord (SACDC) Basics • due to vitamin B12 deficiency • vitamin B12 deficiency increased levels of methylmalonic acid impairs spinal cord myelinization. • dorsal + lateral columns affected • if untreated stiffness and weakness persist Features • joint position and vibration sense lost first then distal paraesthesia • upper motor neuron signs typically develop in the legs, classically: extensor plantars, Plantars are initially flexor, and later extensor. brisk knee reflexes, (but may be increased, normal or absent) absent ankle jerks

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 On presentation, 50% of patients have absent ankle reflexes with hyperreflexia at the knees. • Spastic paresis • Gait abnormalities (spinal ataxia, positive Romberg's test) • Lhermitte's phenomenon is typically present in multiple sclerosis, but may also occur in subacute combined degeneration of the cord. Diagnosis • MRI typically shows increased signal on T2~weighted imaging in the dorsal columns

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Transverse myelitis Overview • inflammation across the entire width of one level, or segment of the spinal cord. • Characterised by acute or subacute motor, sensory and autonomic spinal cord dysfunction. • the thoracic region of the spinal cord is most commonly affected. Causes • Acute infection Viral: most commonly Bacterial infections: syphilis, Lyme disease • Post-infections or vaccination (immune mediated) • Autoimmune (SLE, MS) Features • Course of the disease: develop over hours to days, and are usually bilateral • Motor dysfunction (e.g., paresis, paraplegia) • Sensory dysfunction Sensory level is characteristic. Midline or dermatomal neuropathic pain can be present. • Autonomic dysfunction Sphincter dysfunction Urinary incontinence or retention Bowel incontinence or constipation Sexual dysfunction is common but vary in severity. Investigation • MRI to rule out the presence of structural lesions, to determine the presence of myelitis, which enhances with gadolinium in the acute phase. Evidence of inflammation can be confirmed via gadolinium-enhanced MRI. There may be more than one area of myelitis, and the lesions usually span at least two vertebral segments. there is variable enlargement of the spinal cord In the acute phase the MRI may be normal. • CSF analysis: pleocytosis and/or elevated IgG index Treatment • First-line: immediate high-dose IV corticosteroids • Plasma exchange can be given to those who fail to respond. • Patients with demyelinating disease can be started on long term immunosuppression.

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Neurology Prognosis • Predictors of poor prognosis rapidly progressive course severe weakness hypotonia areflexia • Improvement chances time frame: improvement can take three months and longer to develop percentage: 50 - 70% of patients have partial or complete recovery. One-third of patients recover with little or no sequelae One-third are left with a moderate degree of permanent disability One-third are left with severe disabilities • Risk of future MS: Depends on the pattern of transverse myelitis: complete transverse myelitis: only 5-10% will be diagnosed with MS incomplete transverse myelitis: 60-90% will be diagnosed with MS within 5 years.

Syringomyelia Syringomyelia typically causes loss of reflexes, spinothalamic sensory loss (pain and temperature), and weakness. It can be asymmetrical initially Definition • Syringomyelia is a degenerative disease of the spinal cord that is characterized by a fluidfilled cavity within the cervical spinal cord. Pathophysiology • development of cavity (syrinx) within the spinal cord • Syrinx (fluid-filled cavitation) in the central spinal cord, usually cervical. This can elongate and enlarge, causing compression of the corticospinal and spinothalamic tracts and anterior horn cells. • if extends into medulla then termed syringobulbia • Most of the cavities in syringomyelia lie between the second cervical and the ninth thoracic vertebrae. most commonly affecting the cervical region • collection of fluids within the central canal of the spinal cord enlargement spinal canal, leading to damage of the crossed fibers (anterior white commissure) of the spinothalamic tract loss of pain and temperature sensation in the upper extremities Epidemiology • more common in men than women • usually presents in the 20s and 30s although it can present later in life. Causes • Arnold-Chiari malformation type I impaired cerebrospinal fluid circulation The most common cause • arachnoiditis, • meningeal carcinomatosis, • space-occupying lesions • Post-traumatic syringomyelia

 complicate up to 4% of spinal cord injury often presents with pain, which spreads upwards from the initial injury site. • idiopathic. Features • maybe asymmetrical initially • slowly progressive sensory and motor symptoms, possibly over years • motor: wasting and weakness of arms • sensory: spinothalamic sensory loss (pain and temperature) bilateral loss of pain and temperature sensation in the upper extremities. fine touch sensation, vibration and proprioception are preserved • loss of reflexes, bilateral upgoing plantars • Horner's syndrome, seen in advanced syringomyelia due to disruption of sympathetic trunk neurons. • Bladder, bowel and sexual dysfunction can develop Investigations MRI is the investigation of choice • MRI of the spinal cord the diagnostic modality of choice. MRI enhanced with gadolinium has more sensitivity than regular MRI. • Myelography used to confirm the diagnosis but was associated with more deterioration Localization of the lesion • At syrinx (there is anterior horn cell involvement) → lower motor neuron pattern of weakness. • At central decussating fibres (spinothalamic tract) → dissociated sensory loss with late development of neuropathic arthropathy. • At corticospinal tracts below the level of the syrinx results in spastic paraparesis. Deferential diagnosis • Amyotrophic lateral sclerosis NO sensory deficits. • Anterior spinal artery thrombosis characterised by loss of motor function below the level of injury, loss of pain and temperature sensations, and preservation of proprioception, fine touch and vibration. • Post-traumatic spinal stenosis result in neurological changes below the level of stenosis. Management • The mainstay of the treatment of is surgery. MRCPUK-part-1-May 2010: feature of weakness & wasting of the small muscles of the hand. Which one of the following features would most support a diagnosis of syringomyelia? Loss of temperature sensation in the hands

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The presence of loss of pain and temperature sensation in a 'cape like' distribution is highly suggestive of syringomyelia. A fluid filled cavity within the cord develops, typically between C2 to T9, compressing the cord from inside to out.

Arnold-Chiari malformation (CM) Definition • Arnold-Chiari malformation describes the downward displacement, or herniation, of the cerebellar tonsils through the foramen magnum. Causes • may be congenital or acquired through trauma. Pathophysiology • Symptoms of Arnold-Chiari malformation, type I develop as a result of three pathophysiological consequences of the disordered anatomy:

1. compression of the medulla and upper spinal cord,
2. compression of the cerebellum,
3. disruption of cerebrospinal fluid flow through the foramen magnum. Classification • classified by extent with which parts of the brain protrude into the spinal canal. Chiari I malformation, the only type that can be acquired or can remain asymptomatic until late childhood or early adulthood. characterized by: the time of onset (late

childhood/early adulthood) and the downward herniation of cerebellar tonsils, without the involvement of brainstem tissue.

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 symptoms due to obstruction of cerebrospinal fluid flow. more severe types of Chiari malformations would involve additional herniation of brainstem tissue (Types II and III) or incomplete development of the cerebellum as a whole (Type IV). Features • non-communicating hydrocephalus may develop as a result of obstruction of cerebrospinal fluid (CSF) outflow • neck pain • Occipital headache exacerbated by cough, valsalva maneuver and exercise. • syncope due to intermittent obstructive hydrocephalus. • changes in balance, and poor hand coordination • Syringomyelia • Downbeat nystagmus is classically associated with lesions at the foramen magnum (Arnold-Chiari malformation) Investigations • MRI brain: Narrow posterior fossa Treatment • type II and III CM and in symptomatic type I CM Surgery • asymptomatic type I CM : Surveillance: annual MRI of the brain to look for development of syringomyelia and/or hydrocephalus

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MRI showing herniation of the cerebellar tonsils through the foramen magnum consistent with a Chiaria I malformation

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Anterior spinal artery thrombosis Anterior spinal artery thrombosis Sudden paralysis and loss of pain and temperature sensation and preservation of fine touch, vibration, and proprioception below the level of the lesion. Vibration and proprioception are typically spared because of an intact dorsal column Anterior spinal artery infarct occurs at the 'watershed' T4-T6 and would cause symptoms primarily in the lower limb and a sensory level. • it supplies, roughly the anterior 2/3 of the cord. • segments of the cord in the watershed area between the branches (around T2–T4) are vulnerable to ischaemia. Sequelae • Occlusion of the anterior spinal artery infarcts the ventral portion of the cord. • affects the structures found at the front of the spine corticospinal tracts (motor neurons) spinothalamic tracts (pain/temperature sensation). Feature • Acute (within hours) Back or chest pain Spinal shock Bilateral loss of temperature and pain sensation , motor function (flaccid paraparesis or quadriparesis) , and autonomic function (bladder, bowel, and sexual dysfunction, orthostatic hypotension) below the level of the lesion reflexes are diminished

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 Absent Bulbocavernosus reflex: squeezing the glans penis or pulling on a Foley catheter while digitally palpating the contraction of the anal sphincter • Late (after days or weeks) Continued sensory and autonomic dysfunction Power is reduced below the hips Pain and temperature sensation are lost to the waist. Spastic paraparesis or quadriparesis (increased muscle tone)

Hyperreflexia • Light-touch sensation, vibration and proprioception (joint-position sense) are normal because these are carried in the dorsal columns that are supplied by the posterior spinal artery. • Injury level Anterior spinal cord lesions above cervical vertebra 6 will result in tetraplegia with involvement of the upper and lower extremities. injuries from T1-T6 have normal upper extremity, although abdominal and chest muscles may be affected with diminished respiratory excursion. The region of thoracic vertebra 6 is the thoracic watershed zone; lesions below this level result in loss of bowel, bladder, and sexual functions. Anterior spinal arteries supply corticospinal and spinothalamic tracts, and anterior horns of the grey matter. What are the diagnostic possibilities of a lesion involve the anterior two thirds of the spinal cord which spares light touch, vibration and position sense, but causes loss of pain and temperature sensation distally? The diagnostic possibilities include : 1- anterior spinal artery occlusion sudden onset 2- intramedullary spinal cord metastasis

Chapter 4

Neurology Types of incomplete spinal cord syndromes • All types present with dissociated sensory loss: a pattern of selective sensory loss (“dissociation of modalities”); suggests a focal lesion of a single tract within the spinal cord Affected spinal tracts Etiology Clinical features Bilateral central corticospinal tracts and lateral spinothalamic tracts Central cord syndrome (most common) • Hyperextension injury (e.g., car crash) associated with chronic cervical spondylosis • Spinal cord compression Anterior cord syndrome Corticospinal and spinothalamic tracts • Trauma (e.g., penetrating injury, burst fracture of vertebra) • Bilateral motor paralysis, loss of pain and temperature sensation, and autonomic dysfunction below the level of the lesion Posterior cord syndrome • Occlusion of anterior spinal artery Bilateral posterior columns • Trauma (e.g., penetrating injury) • Ipsilateral loss of proprioception, vibration, and touch sensation below the level of the lesion Brown-Séquard syndrome (hemisection syndrome) • Occlusion of the posterior spinal artery • Multiple sclerosis Hemisection of the cord • Trauma (e.g., penetrating injury) • Spinal cord compression Diagnosis • Spinal MRI (best confirmatory test): excludes soft-tissue lesions (e.g., tumors, hematomas), bone lesions, and detects spinal cord parenchyma abnormalities (e.g., infarction) Notes & Notes for MRCP
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• Bilateral paresis: upper > lower extremities • Ipsilateral Loss of proprioception, vibration, and tactile discrimination below the level of the lesion Segmental flaccid paresis at the level of the lesion, spastic paralysis below the level of the lesion , and ipsilateral Babinski sign • Contralateral: loss of pain and temperature sensation one or two levels below lesion

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Brown-Séquard's syndrome Definition • Thoracic spinal cord lesion produced by a hemisection of the spinal cord. Causes • trauma, (most commonly) • tumours, and • multiple sclerosis. Features • Ipsilateral Weakness (paralysis) loss of position and vibration below the lesion (dorsal column dysfunction) Horner syndrome, If the lesion is above the spinal cord level T1, due to damage of the oculosympathetic pathway. • Contralateral loss of pain and temperature. Diagnosis • MRI is

the imaging of choice in spinal cord lesions Management • Steroids may decrease cord swelling.

Lower back pain Overview • Lower back pain (LBP) is one of the most common presentations seen in practice. • Whilst the majority of presentations will be of a non-specific muscular nature it is worth keeping in mind possible causes which may need specific treatment. Red flags for lower back pain • age < 20 years or > 50 years • history of previous malignancy • night pain • history of trauma • systemically unwell e.g. weight loss, fever

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Neurology The table below indicates some specific causes of LBP: Facet joint May be acute or chronic Pain worse in the morning and on standing On examination there may be pain over the facets. The pain is typically worse on extension of the back Spinal stenosis Usually gradual onset Unilateral or bilateral leg pain (with or without back pain), numbness, and weakness which is worse on walking. pain is worse with walking downhill and less with walking uphill. Resolves when sits down. Pain may be described as 'aching', 'crawling'. Relieved by sitting down, leaning forwards and crouching down Clinical examination is often normal Requires MRI to confirm diagnosis Ankylosing spondylitis Typically, a young man who presents with lower back pain and stiffness Stiffness is usually worse in morning and improves with activity Peripheral arthritis (25%, more common if female) Peripheral arterial disease Pain on walking, relieved by rest Absent or weak foot pulses and other signs of limb ischaemia Past history may include smoking and other vascular diseases

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Wernicke's encephalopathy Definition • Wernicke's encephalopathy is a neuropsychiatric disorder caused by thiamine deficiency, which is most commonly seen in alcoholics. Causes • Most common: alcohol • Rarer causes include: persistent vomiting, stomach cancer, dietary deficiency. Features • nystagmus (the most common ocular sign) • ophthalmoplegia • ataxia • confusion, altered GCS • peripheral sensory neuropathy • Sometimes bilateral wrist drop but more frequently bilateral foot drop with pain or pressure over the long nerves. Wernicke's encephalopathy: classic triad of:

1. nystagmus,
2. ophthalmoplegia
3. ataxia

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• petechial haemorrhages occur in a variety of structures in the brain including the mamillary bodies and ventricle walls Investigations • 1st investigations to order therapeutic trial of parenteral thiamine • decreased red cell transketolase • MRI Treatment is with urgent replacement of thiamine (Pabrinex (Intravenous))

Korsakoff syndrome Definition • a late neuropsychiatric manifestation of Wernicke encephalopathy. Pathophysiology • alcohol vitamin B1 (thiamine) deficiency damage to mammillary bodies (structures of the limbic system) Feature • Wernicke's encephalopathy + antero- and retrograde amnesia and confabulation. confabulation (false memories) is a disturbance of memory, defined as the production of fabricated memories without the conscious intention to deceive. dementia is typically not reversible. Investigations • MRI finding mammillary body degeneration Treatment • maintenance thiamine and rehabilitation

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Anti-NMDA receptor encephalitis (Autoimmune encephalitis) Definition • It is a type of brain inflammation due to antibodies. (a paraneoplastic syndrome), presenting as prominent psychiatric features including agitation, hallucinations, delusions and disordered thinking; seizures, insomnia, dyskinesias and autonomic instability. • might be misdiagnosed as a primary psychiatric illness. Mechanism • autoimmune with the primary target the N-methyl D-aspartate receptors (NMDAR) in the brain Epidemiology • 80% are female • particularly prevalent in Afro-Caribbean patients. Associations • Ovarian teratomas are detected in up to half of all female adult patients, Investigations • CSF can be normal initially. Korsakoff's syndrome: Inability to acquire new memories and confabulation

 may demonstrate pleocytosis antibodies against NMDA receptors CSF titers of anti-NMDA receptor antibodies correlate with clinical illness • MRI head can be normal in 50% abnormalities can be visualised on FLAIR sequences in the deep subcortical limbic structures. • evaluation for an ovarian teratoma by MRI, CT scan, or ultrasound Treatment • immunosuppression with intravenous steroids, immunoglobulins, rituximab, cyclophosphamide or plasma exchange, alone or in combination. • Resection of teratoma is also therapeutic.

CADASIL Overview • Cerebral Autosomal Dominant Arteriopathy with Subcortical Infarcts and Leukoencephalopathy (CADASIL) • A family history is almost always present, as it is an autosomal dominant condition, located to chromosome 19. • the most common genetic form of vascular dementia. Features • What is the pathophysiology of this condition?
 NOTCH3 mutation • strokes at a young age and • early vascular (subcortical) dementia (multi-infarct dementia) • patients often present with migraine • Recurrent ischaemic events (transient or permanent) & Severe mood disorders Diagnosis • Characteristic MRI changes include T2 weighted hyperintensity of the periventricular white matter. • DNA testing for the notch-3 gene mutation confirms the diagnosis. Treatment • the oral contraceptive pill should be stopped, given its association with stroke in migraine.

Myotonic dystrophy Definition • Myotonic dystrophy (also called dystrophia myotonica) is an inherited myopathy results in a selective atrophy of type I muscle fibers affects skeletal, cardiac and smooth muscle. Genetics • autosomal dominant, trinucleotide repeat disorder. Patients have

between 50 to 1,000 CTG trinucleotide repeats in the myotonin protein kinase gene (normal is less than 30 repeats).

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Types: There are two main types of myotonic dystrophy, DM1 and DM2. DM1 DM2 Genetics DMPK (Dystrophia Myotonica-Protein Kinase) gene on chromosome 19 ZNF9 gene on chromosome 3 Onset Congenital, juvenile, or adult-onset Adulthood Features Distal weakness more prominent Proximal weakness more prominent Severity Sever disease Mild disease Features • features developing at around 20-30 years old. • myotonic facies (long, 'haggard' appearance), frontal balding, atrophy of temporalis, masseters and facial muscle, bilateral ptosis • cataracts • myotonia (tonic spasm of muscle), slow-relaxing grip may be noticed on initial hand-shake with the patient and is typical of myotonic dystrophy. • weakness of arms and legs (distal initially) • mild mental impairment • diabetes mellitus (Insulin resistance) • testicular atrophy • Dysarthric speech secondary to myotonia of the tongue and pharynx • cardiac involvement: heart block, cardiomyopathy • dysphagia Diagnosis • Serology →increased serum CK • Electromyogram (EMG) → the most appropriate next step to confirm the diagnosis EMG changes → Waxing and waning of potentials , termed the “dive bomber effect” • Muscle biopsy • Genetic testing: the gold standard for confirming the diagnosis Treatment: mostly symptomatic • for weakness which is the main cause of disability →there is no treatment • for myotonia →phenytoin, quinine or procainamide may be useful , mexiletine is a sodium channel blocker often used for myotonic symptoms. • for cardiac abnormalities →pacemaker • for obstructive sleep apnea → CPAP • Foot drop can be managed with →ankle-foot orthosis and splints. • For ptosis : lid-lifting surgery has no place except in severe cases • may need cataract extraction. • Genetic counseling and testing Prognosis • The course is chronic progressive. • Cardiac complications reduce life expectancy.

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Neurology Top Tips

Dystrophinopathies Overview • X-linked recessive Affected father (Y, X): All sons will not be affected and not carriers (His sons will get the X chromosome from their mother) All his daughters will be carriers Carrier mother (X , X): 50% of sons will be affected (there is a 1 in 2 chance (50:50) of passing the gene on to their sons.) • due to mutation in the gene encoding dystrophin, dystrophin gene on Xp21

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• dystrophin is a protein in muscle which connects the muscle membrane to actin, part of the muscle cytoskeleton Diagnostic investigations • 1st investigations to order serum CK 50 to 100 times normal level consistent with Duchenne muscular dystrophy genetic testing DNA analysis Xp21 mutation may present in both Duchenne and Becker muscular dystrophies • Investigations to consider EMG EMG can distinguish between neuropathic and myopathic

pathology. myopathic reading with fast firing, short duration but polyphasic and decreased amplitude motor units with early recruitment in the affected muscles muscle biopsy absence of dystrophin Duchenne muscular dystrophy diminished quantity or quality of dystrophin Becker muscular dystrophy Duchenne muscular dystrophy (DMD) • most common and most rapidly progressive muscular dystrophy • there is a frameshift mutation resulting in one or both of the binding sites are lost leading to a severe form • progressive proximal muscle weakness from 5 years Usually, there is severe progression with wheelchair dependence by the age of 12 on average Death usually occurs as a teenager or in the early 20s from respiratory failure. • calf pseudohypertrophy • Gower's sign: child uses arms to stand up from a squatted position • intellectual impairment (30%) • urinary and bowel incontinence (common) • DMD patients tend to be hyperactive and have difficulty in focusing attention. Becker muscular dystrophy • there is a non-frameshift insertion in the dystrophin gene resulting in both binding sites being preserved leading to a milder form • develops after the age of 10 years • Similar type of disease to Duchenne's, with a later onset (average age at presentation 12 years), milder phenotype and longer life expectancy • intellectual impairment much less common • Occasionally, patients present with CHF and cardiac arrhythmias before complaining of muscle weakness and before diagnosis. Facio-scapulo-humeral muscular dystrophy (FSHMD) • autosomal dominant form of muscular dystrophy. • As the name suggests it is typically affects the face, scapula and upper arms first. • Symptoms typically presents by the age of 20 years. • may go unrecognised until later life

Neurology

• The presence of distal wasting and pes cavus (indicates a very chronic neuromuscular disorder with axonal loss) Oculopharyngeal muscular dystrophy • ptosis • weakness of the extraocular muscles • dysphagia • tongue atrophy

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Foster-Kennedy syndrome Foster Kennedy's syndrome is a combination of optic atrophy and central scotoma, contralateral papilloedema and anosmia. Overview • Foster-Kennedy syndrome describes a series of symptoms and signs associated with frontal lobe lesions. • It is caused by optic and olfactory nerve compression and raised intracranial pressure. • This is often secondary to a mass such as an olfactory groove meningioma. Features • optic atrophy in the ipsilateral eye • central scotoma in the ipsilateral eye • papilloedema in the contralateral eye • anosmia • symptoms of raised intracranial pressure such as nausea and vomiting, • frontal symptoms such as emotional lability and memory loss. The presence of optic atrophy on one side with contralateral papilloedema is characteristic of Foster Kennedy syndrome as it is usually due to frontal tumour or tumour within the olfactory bulb compressing the ipsilateral optic nerve and causing raised intracranial pressure.

Hypokalaemic periodic paralysis and thyrotoxic periodic paralysis Epidemiology • Most commonly seen in Asian men in their third to fifth decades • The prevalence is much higher in patients with thyrotoxicosis of Chinese origin versus Caucasians, (13-14% vs. 0.1-0.2%). • occurs in 10% of young Latin American or Asian men with thyrotoxicosis (of whatever aetiology).

Pathophysiology • autosomal dominant disorder • The underlying defect is a mutation in muscle voltage-gated calcium channels. • Increase Na+/K+-ATPase activity → shift of potassium into tissues

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Features • Episodes of paralysis: sudden onset of complete weakness with speedy recovery. Attacks of focal or generalized flaccid muscle weakness (periodic paralysis) Proximal muscles are more prominently affected; respiratory and facial muscles are generally spared Variable duration (hours to days) Concomitant fatigue, muscle pain, and/or altered state of consciousness during the attacks Neurological examination is usually normal between attacks. • Attacks may be precipitated by: Carbohydrate-rich meals Exercise Stress • May associate with thyrotoxicosis With thyrotoxicosis called →Thyrotoxic hypokalaemic periodic paralysis Without thyrotoxicosis called →hypokalaemic periodic paralysis. Diagnosis • ↓ K+, documentation of hypokalaemia during an attack • ↓ TSH, ↑ T3, T4 hormones (Thyrotoxic periodic paralysis) Management • Potassium infusion →provide immediate relief from symptoms • Continuous cardiac monitoring • Lifelong potassium supplementation • The periodic paralysis resolves when the thyrotoxicosis is treated. • Non-selective beta-blocker such as propranolol blunts the hyperadrenergic stimulation of Na+/K+-ATPase and thus prevents intracellular shift of potassium and phosphate.

Neuromyelitis optica (NMO) Definition • demyelinating disease involving the optic nerves and spinal cord but sparing the brain. Features • monophasic or relapsing-remitting • particularly prevalent in Asian populations • Vomiting is also a common presenting complaint. Diagnostic criteria: bilateral optic neuritis, transverse myelitis and 2 of the following 3 criteria:

1. Spinal cord lesion involving 3 or more spinal levels
2. Initially normal MRI brain
3. Aquaporin 4 positive serum antibody The classic antibody associated with neuromyelitis optica is NMO-IgG or antibodies against aquaporin-4.

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Neurology

Vertigo Overview • Vertigo is a sensation of spinning while you’re actually stationary. • Vertigo is caused most often by inner ear disease but can also be caused by disease of the vestibular nerve, brainstem, or cerebellum. • Inner ear causes of vertigo include benign paroxysmal positional vertigo (BPPV), labyrinthitis, and Ménière disease. • Horizontal-rotational nystagmus is associated with peripheral vertigo, whereas vertical nystagmus is associated with central vertigo. Common causes of vertigo Disorder Notes Labyrinthitis Recent viral infection or head trauma Sudden onset Nausea and vomiting Typically has associated tinnitus and a history of infection. Hearing may be affected Vestibular neuritis Recent viral infection Recurrent vertigo attacks lasting hours or days No hearing loss Benign paroxysmal positional vertigo Gradual onset

Triggered by change in head position Each episode lasts 10-20 seconds Meniere's disease Associated with hearing loss, tinnitus and sensation of fullness or pressure in one or both ears Vertebrobasilar ischaemia Elderly patient Dizziness on extension of neck Acoustic neuroma Hearing loss, vertigo, tinnitus Absent corneal reflex is important sign Associated with neurofibromatosis type 2 Distinguishing vertigo of brainstem and cerebellar ischemia from peripheral causes The HINTS exam is a three-part, rapid beside oculomotor test used to help differentiate central from peripheral vertigo. HINTS stands for Head Impulse, Nystagmus and Test of Skew. The test consists of three parts:

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1 – Patients with peripheral vertigo will have abnormal (positive) head impulse testing, while patients with central vertigo typically have a normal (negative) head impulse test.

2 – Patients with peripheral vertigo will have unidirectional, horizontal nystagmus, while patients with central vertigo can have rotatory or vertical nystagmus, or direction-changing horizontal nystagmus.

3 – Alternate eye cover testing may reveal skew deviation in patients with central vertigo, and should be absent in peripheral vertigo. Any of the following, whether present or untestable, suggest a brainstem or cerebellar lesion: • Normal head impulse test on both sides • Direction-changing nystagmus • Skew deviation The presence of all of the following suggests a peripheral lesion: • An abnormal head impulse test on one side • Unidirectional, horizontal, torsional nystagmus that increases in intensity with gaze toward the fast phase • Absent skew The importance of these oculomotor tests is that brain imaging with either CT or MRI may be normal during the acute phase of ischemic symptoms. In this regard, the HINTS test appears to be more sensitive for the diagnosis of acute stroke than even brain MRI within the first two days after symptom onset

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Benign paroxysmal positional vertigo (BPPV) Overview • vertigo triggered by change in head position (e.g. rolling over in bed or gazing upwards) • may be associated with nausea • each episode typically lasts 10-20 seconds Features Vertigo and nausea, with nystagmus, fit best with benign paroxysmal positional vertigo, which occurs due to otolith detachment into the semicircular canals of the inner ear. Diagnosis • Positive Dix-Hallpike manoeuvre First-line test for suspected BPPV Positive Dix-Hallpike test: positional vertigo and nystagmus triggered during the maneuver Further steps for positive test: Perform Epley repositioning maneuver. Treatment • Symptomatic relief may be gained by Epley manoeuvre (successful in around 80% of cases) • Medication is often prescribed (e.g. Betahistine) but it tends to be of limited value.

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