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Notes & Notes for MRCP
By Dr. Yousif Abdallah Hamad

Trigeminal neuralgia Overview • Sensation over the face is supplied by the trigeminal nerve • Trigeminal neuralgia is a pain syndrome characterised by severe unilateral pain. • Most often affecting women ♀> ♂(2:1) • Peak age incidence: 60–70 years Classification • Idiopathic trigeminal neuralgia Most common type no identifiable cause (unremarkable findings on MRI and electrophysiological tests) • Secondary trigeminal neuralgia Caused by a major underlying neurological disease, most frequently multiple sclerosis, a tumor at the cerebellopontine angle, or arteriovenous malformation. Red flag features suggesting a serious underlying cause Sensory changes Deafness or other ear problems History of skin or oral lesions that could spread perineurally Pain only in the ophthalmic division of the trigeminal nerve (eye socket, forehead, and nose), or bilaterally Optic neuritis A family history of multiple sclerosis Age of onset before 40 years Should be referred to neurology MRI of the brain is the next management step Features • The International Headache Society defines trigeminal neuralgia as: Unilateral disorder characterised by brief electric shock-like pains, abrupt in onset and termination, limited to one or more divisions of the trigeminal nerve Lasts several seconds (in rare cases, several minutes) and may occur up to 100 times per day Typically shoots from mouth to the angle of the jaw on the affected side Usually triggered by movements such as chewing, talking, or touch (e.g., brushing teeth, washing face); becomes worse with stimulation Management • Carbamazepine is first-line • Failure to respond to treatment or atypical features (e.g. < 50 years old) should prompt referral to neurology MRCPUK-part-1-January 2015 exam: History of electric shock like pains on the right side of the face. around 10-20 episodes a day which, each lasting for around 30-60 seconds. What is the most suitable first-line management? Carbamazepine Trigeminal neuralgia - carbamazepine is first-line

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Neurology

What is the nerve supply to the angle of the jaw? The angle of the jaw is supplied by nerve roots C2/C3 and not the trigeminal nerve. In patients with non-organic sensory loss, that loss usually extends to the edge of the jaw.

Abducens (VIth) nerve palsy Anatomy • Location: The abducens nucleus located in the caudal pons • Course: The nerve leaves the brainstem at the junction of the pons and medulla and runs upward into the subarachnoid space, travelling through the cavernous sinus alongside the internal carotid artery. It enters the orbit through the superior orbital fissure, like the other ocular cranial nerves, and innervates the lateral rectus, which serves to abduct the eye. • Function: The VIth nerve is motor to the lateral rectus muscle. It is responsible for abduction of the ipsilateral eye. Features • Horizontal diplopia that worsens when looking at distant objects • Inability to abduct the eye • In the neutral position the affected eye is deviated medially due to unopposed action of the medial rectus. • In patients with diplopia the 'cover test' can be used to determine the eye that has the problem. On covering the affected eye, the outermost image disappears. Eg : diplopia on right horizontal gaze , improved on covering the right eye the right abducens is affected Causes • Most common ocular cranial nerve palsy • Trauma • Pseudotumor cerebri • Cavernous sinus thrombosis • Due to the long course and anatomy of the VIth nerve it can be damaged in any condition causing raised intracranial pressure. It can therefore be a 'false localising sign'. Which nerve passes alongside the internal carotid artery within the cavernous sinus? • Cranial nerve VI, the abducens nerve.

Facial (VII) nerve Facial nerve branches (mnemonic) (superior to inferior) as they exit the anterior border of the parotid gland: To Zanzibar By Motor Car

1. T: temporal
2. Z: zygomatic
3. B: buccal
4. M: mandibular
5. C: cervical Facial Palsy + convergent squint ↓ lesion in Pons as VI th is encircled by VII th
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Abducens nerve palsy • The patient is unable to abduct the right eye. • Abducens nerve innervates the ipsilateral lateral rectus muscle that is necessary for lateral movement of the eye. Facial nerve paralysis is often accompanied by: • loss of taste, • hyperacusis, and • decreased salivation.

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Neurology Supply - 'face, ear, taste, tear' • face: muscles of facial expression • ear: nerve to stapedius (Hyperacusis is due to paralysis of stapedius) • taste: supplies anterior two-thirds of tongue • tear: parasympathetic fibres to lacrimal glands, also salivary glands • Orbicularis oculi is affected causing inability to blink/close eyelids. Causes of bilateral facial nerve palsy

1. Sarcoidosis
2. Guillain-Barre syndrome
3. polio,
4. Lyme disease Causes of unilateral facial nerve palsy - as above plus Lower motor neuron Upper motor neuron • Bell's palsy • Ramsay-Hunt syndrome (due to herpes zoster) •

Acoustic neuroma • Parotid tumours • HIV • Multiple sclerosis* may also cause an UMN palsy • Diabetes mellitus • Stroke LMN vs. UMN • upper motor neuron lesion 'spares' upper face i.e. forehead • lower motor neuron lesion affects all facial muscles Lesions • The majority of facial nerve palsy cases result from infranuclear lesions. • The most common cause of facial nerve paralysis is Bell's palsy.

Bell's palsy Definition • acute, unilateral, idiopathic, facial nerve paralysis. Causes • unknown • although the role of the herpes simplex virus has been investigated previously. Epidemiology • The peak incidence is 20-40 years • more common in pregnant women. Features • lower motor neuron facial nerve palsy - forehead affected • other features post-auricular pain (may precede paralysis),

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 altered taste, dry eyes, hyperacusis (seen in around a third of patients) Management • prednisolone 1mg/kg for 10 days should be prescribed for patients within 72 hours of onset of Bell's palsy. • Adding in aciclovir gives no additional benefit • eye care is important - prescription of artificial tears and eye lubricants should be considered Prognosis • if untreated around 15% of patients have permanent moderate to severe weakness MRCPUK-part-1-January 2012 exam: Which features would be most consistent with a diagnosis of Bell's palsy?
 Hyperacusis MRCPUK-part-1-May 2010 exam: What is the current evidenced base approach to the management of Bell's palsy? Prednisolone

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Ramsay Hunt syndrome Aetiology • Ramsay Hunt syndrome (herpes zoster oticus) is caused by the reactivation of the varicella zoster virus in the geniculate ganglion of the seventh cranial nerve. Features • auricular pain is often the first feature • facial nerve palsy • vesicular rash around the ear • tinnitus • vertigo Management • oral aciclovir and corticosteroids

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Acoustic neuroma Overview • Acoustic neuromas (more correctly called vestibular schwannomas) are benign tumors of the vestibular nerve (8th nerve). • account for 5% of intracranial tumours and 90 % of cerebellopontine angle • Bilateral acoustic neuromas are seen in neurofibromatosis type 2 vesicular rash around the ear (or anterior 2/3rds of the tongue and the soft palate): suggest a diagnosis of Ramsey Hunt syndrome. Loss of corneal reflex →think acoustic neuroma

Chapter 4

Neurology

Features • can be predicted by the affected cranial nerves cranial nerve V: absent corneal reflex cranial nerve VII: facial palsy cranial nerve VIII: hearing loss, vertigo, tinnitus, gait disturbances and imbalance. Investigation • MRI of the cerebellopontine angle is the investigation of choice mass Treatment • surgical removal remains the treatment of choice

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Abnormal gait Lesions of cerebellar vermis cause →truncal ataxia and tendency to fall backwards. Phenytoin toxicity →broad-based ataxic gait Abnormal gait Diagnosis Shuffling gait Parkinson’s disease Spastic hemi-paretic gait (circumducted) Stroke Waddling gait (with excessive hip swing) proximal myopathy Steppage gait (High stepping, Neuropathic gait) • Foot drop: peroneal nerve palsy and L5 radiculopathy. Choreiform Gait (Hyperkinetic Gait) Sydenham's chorea, Huntington's Disease Ataxic Gait Cerebellar disease, Phenytoin toxicity Sensory Gait (Sensory ataxia) • occurs when there is loss of this proprioceptive input • the patient will slam the foot hard onto the ground in order to sense it. Sensory ataxia is distinguished from cerebellar ataxia by positive Romberg's sign (normal coordination when the movement is visually observed by the patient, and worsened when the eyes are closed) Notes & Notes for MRCP
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If bilateral: • Charcot–Marie–Tooth disease • Polio • Multiple sclerosis • Syphilis • Guillain–Barré syndrome If unilateral: large fiber peripheral neuropathies • diabetic neuropathy disorders of the dorsal columns: • B12 deficiency, • tabes dorsalis

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Nystagmus Definition • involuntary oscillations of the eyes. Relation to directions of the gaze • constant direction regardless of the direction of gaze, suggests a labyrinthine or cerebellar lesion. • changes with the direction of gaze suggests widespread central involvement of vestibular nuclei. • presents only on lateral gaze lesion of the brain stem or cerebellum. • Nystagmus restricted to the abducting eye on lateral gaze (ataxic nystagmus) is due to a lesion of the medial longitudinal bundle between the pons and mid-brain as in multiple sclerosis (MS). Causes • Visual disturbances • Lesions of the labyrinth • Central vestibular connections, Brain stem or cerebellar lesions • Wernicke's encephalopathy • Nystagmus confined to one eye suggests: a peripheral lesion of the nerve or muscle, or a lesion of the medial longitudinal bundle. Vertical VS horizontal nystagmus • Vertical nystagmus: Upbeat nystagmus (occurring on upward gaze: due to a lesion in the mid-brain Downbeat nystagmus (fast phase downwards) suggests a lesion in the lower part of the medulla. It is therefore typical of the Arnold-Chiari malformation (Chiari type I malformation). • Horizontal nystagmus: occurs in unilateral disease of the cerebral hemisphere, with the fast phase directed to the side of the lesion. MRCPUK-part-1-May 2007 exam: Which disorder is most associated with downbeat nystagmus? Arnold-Chiari malformation

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Spinocerebellar ataxia (SCA) Upbeat nystagmus → cerebellar vermis lesions Downbeat nystagmus → foramen magnum lesions (Arnold-Chiari malformation) Spinocerebellar ataxia (SCA): • autosomal dominant • should be suspected in patients with progressive loss of coordination, unsteady gait and overall weakness.

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Hemiballism Hemiballism is caused by damage to the subthalamic nucleus The presence of severe flinging movements affecting proximal muscles and following no particular pattern is typical for hemiballism. Overview • damage to the subthalamic nucleus of the basal ganglia Hemiballism decreased suppression of involuntary movements. • Ballisic movements are involuntary, sudden, jerking movements which occur contralateral to the side of the lesion. • The ballisic movements primarily affect the proximal limb musculature whilst the distal muscles may display more choreiform-like movements • It is always unilateral, but it is common for arms and legs to move together. • Bilateral ballismus is rare and implicates a metabolic cause, usually non-ketotic hyperosmolar coma. • Symptoms may decrease whilst the patient is asleep. • The movements worsens with activity and decrease with relaxation. Causes • vascular events (stroke). infarction being the commonest cause. • traumatic brain activity • amyotrophic lateral sclerosis • hyperglycaemia • malignancy • vascular malformations • tuberculomas, and • demyelinating plaques. Treatment • tetrabenazine is the treatment of choice. • Anti-dopaminergic agents (e.g. Haloperidol) are the mainstay of treatment. • Topiramate can be used, as can intrathecal baclofen, botulinum toxin and tetrabenazine. • Functional neurosurgery can be used for cases which have failed to respond to other treatment. Prognosis • Usually the flinging movements stop spontaneously in the next 4-8 weeks MRCPUK-part-1-September 2012 exam: H/O involuntary, jerking movements of arms, resolved during asleep. Damage to which structure may lead to hemiballism? Subthalamic nucleus

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Epilepsy: Classification Basics • two main categories are generalised and partial seizures • partial seizures may progress to general seizures • other types: myoclonic, atypical absence, atonic and tonic seizures are usually seen in childhood Generalised - no focal features, consciousness lost immediately • Tonic-clonic (grand mal) • Absence seizures (petit mal) absences last a few seconds and are associated with a quick recovery mostly seen in children 1st line treatment→ ethosuximide good prognosis - 90-95% become seizure free in adolescence • myoclonic: brief, rapid muscle jerks • partial seizures progressing to generalised seizures Partial - focal features depending on location • Simple (no disturbance of consciousness or awareness) • Complex (consciousness is disturbed) • Jacksonian seizure also known as a focal (partial) motor seizure. In this condition an uncontrolled, spontaneous discharge of electricity from one motor cortex presents with contralateral motor signs. The patient has preserved consciousness as it is a partial seizure after the seizure it is common to have a Todd's paralysis where the limb is weak. • Temporal lobe epilepsy Focal seizure with impaired awareness (complex partial seizure) Can take the form of automatisms such as chewing and swallowing repeatedly, scratching the head or searching for an object.
 Most commonly arise in the temporal lobes. MRI is an appropriate investigation The commonest finding is hippocampal sclerosis • Gelastic seizures Gelastic seizures should be suspected in cases of erratic laughing or crying. typically arise from hypothalamic hamartomas Absence seizure (petit mal) • presents with a blank stare, 3 Hz brain waves and do not show postictal confusion. • Good prognosis: 90 -95% become seizure free in adolescence.

Neurology

Somatosensory seizures Spread of symptoms ('marching') in seconds The usual source is the parietal lobe. Example → tingling sensation starts in fingers and spreads in seconds to affect the whole arm and leg. Positive symptoms (jerking, tingling) usually signify epilepsy. Negative symptoms (weakness, numbness) are usually caused by transient focal ischaemia. Spread of symptoms ('marching') indicates migraine (in 5-20 minutes) or seizures (in seconds).

Epilepsy: investigations • Electroencephalogram (EEG) should be performed only to support a diagnosis An EEG should not be performed in the case of probable syncope because of the possibility of a false-positive result. should not be used in isolation to make a diagnosis of epilepsy. should not be used to exclude a diagnosis of epilepsy in whom the clinical presentation supports a diagnosis of a non-epileptic event. can be used to assess the risk of seizure recurrence in patient presenting with a first unprovoked seizure. When a standard EEG has not contributed to diagnosis, a sleep EEG should be performed. Long-term video or ambulatory EEG may be used in case of diagnostic difficulties after clinical assessment and standard EEG. • Neuroimaging: to identify underlying gross pathology MRI is the imaging investigation of choice. CT should be used if MRI is not available or is contraindicated. In an acute situation, CT may be used to determine whether a seizure has been caused by an acute neurological lesion or illness. Neuroimaging should not be routinely requested when a diagnosis of idiopathic generalised epilepsy has been made. Atonic seizure (also known as “drop seizure” or “drop attack”) • Sudden loss of muscle tone: sudden head drop or collapse (lasts < 15 seconds) • Frequently mistaken for syncope

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Epilepsy: treatment When to start antiepileptics? • Antiepileptics is generally recommended after a second epileptic seizure. • NICE guidelines suggest starting antiepileptics after the first seizure if any of the following are present: the patient has a neurological deficit brain imaging shows a structural abnormality the EEG shows unequivocal epileptic activity the patient or their family or carers consider the risk of having a further seizure unacceptable Which antiepileptics? • Focal seizures Female of childbearing potential: 1st line → lamotrigine 2nd line → levetiracetam 3rd line → oxcarbazepine (can impair the effectiveness of hormonal contraceptives) Male or female who are not of childbearing potential: 1st line → lamotrigine or carbamazepine 2nd line → levetiracetam, oxcarbazepine or sodium valproate • Generalised tonic-clonic (GTC) seizures Female of childbearing potential 1st line → lamotrigine 2nd line → levetiracetam, clobazam, or topiramate Male or female who are not of childbearing potential 1st line → sodium valproate 2nd line → lamotrigine, carbamazepine, oxcarbazepine • Absence seizures (Petit mal) Female of childbearing potential 1st line → ethosuximide 2nd line → lamotrigine 3rd line → combination of ethosuximide and lamotrigine Male or female who are not of childbearing potential 1st line → ethosuximide or sodium valproate 2nd line → lamotrigine 3rd line →

combination of two of these three AEDs: ethosuximide, lamotrigine or sodium valproate

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• Myoclonic seizures Female of childbearing potential 1st line → levetiracetam or topiramate (topiramate can impair the effectiveness of hormonal contraceptives.) 2nd line → add levetiracetam, or topiramate Male or female who are not of childbearing potential 1st line → sodium valproate 2nd line → levetiracetam or topiramate Indications for monitoring of AED blood levels • detection of non-adherence to the prescribed medication • suspected toxicity • adjustment of phenytoin dose • management of pharmacokinetic interactions (for example, changes in bioavailability, changes in elimination, and co-medication with interacting drugs) • specific clinical conditions, for example, status epilepticus, organ failure and certain situations in pregnancy Stopping of anti-epileptic drugs (AED) • Can be considered if seizure free for at least 2 years, with AEDs being stopped slowly over 2-3 months (withdrawing benzodiazepines and barbiturates may take up to 6 months or longer) • Benzodiazepines should be withdrawn over a longer period. Vagus nerve stimulation • indicated for use as an adjunctive therapy in reducing the frequency of seizures in adults who are refractory to antiepileptic medication AED cessation can be considered if seizure free for > 2 years – Stop AEDs over 2-3 months MRCPUK-part-1-January 2015 exam: Which one of the antiepileptic drugs is most associated with weight gain? Sodium valproate MRCPUK-part-1-September 2012 exam: What is the most appropriate first-line antiepileptic for myoclonic seizures? Sodium valproate

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Antiepileptic drugs (AED) Overview • Only start after a minimum of two fits. • Only use one drug at a time, and begin with a small dose, and gradually increase it, until control is achieved, toxic affects occur, or the maximum dose is reached. • The diagnosis of epilepsy needs to be critically evaluated if events continue despite an optimal dose of a first-line AED. • If the initial treatment is unsuccessful, then monotherapy using another drug can be tried. If there are absence or myoclonic seizures, or if juvenile myoclonic epilepsy (JME) is suspected, do not offer carbamazepine, gabapentin, oxcarbazepine, phenytoin, pregabalin, tiagabine or vigabatrin.

• If an AED has failed because of adverse effects or continued seizures, a second drug should be started (which may be an alternative first-line or second-line drug) and built up to an adequate or maximum tolerated dose and then the first drug should be tapered off slowly. Drug Mechanism Side effects Clinical uses Benzodiazepines ↑GABA action Sedation, tolerance, dependence, respiratory depression Phenobarbital ↑GABA action Sedation, impairment of motor and cognition systems after long term use, megaloblastic anaemia Rarely used due to sedation – been superseded by phenytoin Phenytoin Inhibits sodium channels Blocks Na+ channels ;zero-order kinetics Carbamazepine Inhibits sodium channels Fatigue, Headache, Itching, SJS Useful for absence seizures Lamotrigine Blocks voltage-gated Na+ channels, inhibits the release of glutamate Ethosuximide Blocks thalamic Ttype Ca2+ channels Sodium valproate ↑Na+ channel inactivation ↑GABA concentration by inhibiting GABA transaminase for partial seizures Levetiracetam SV2A receptor blocker; May modulate GABA and glutamate release, Inhibit voltagegated Ca2+

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1st line for acute Gum hypertrophy, arrythmias Cytochrome P-450 induction, Pseudo-lymphoma, Hirsutism, Nystagmus, Yellow-brown skin, Teratogenicity (fetal hydantoin syndrome), Osteopenia, Inhibited folate absorption, Neuropathy. Rare: SJS, DRESS syndrome, drug-induced lupus. Toxicity leads to diplopia, ataxia, sedation. Partial and generalised attacks, but not in absence. High doses my precipitate attacks 1st line for partial seizures. 2nd or 3rd line, when other drugs unsuccessful. Diplopia, ataxia, blood dyscrasias (agranulocytosis, aplastic anemia), liver toxicity, teratogenesis (cleft lip/palate, spina bifida), induction of cytochrome P-450, SIADH, SJS , skin rash Generalised seizures – 2nd line treatment SJS (must be titrated slowly), hemophagocytic lymphohistiocytosis (black box warning) 1st line for: Alopecia, Hepatotoxic, Pancreatitis, P-450 inhibition (reduces efficacy of contraceptive pill), Rash, Weight gain, Tremor, Teratogenesis (neural tube defects). Absence seizures & Generalised seizures 2nd line Neuropsychiatric symptoms (eg, personality change), fatigue, drowsiness, headache For partial and generalised

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Neurology Which antiepileptic drugs does not have interactions with warfarin? • Lamotrigine has no effect on liver enzymes and is the treatment of choice for patient taking warfarin • Phenytoin, carbamazepine, primidone and phenobarbital are liver enzyme inducers • Sodium valproate is a liver enzyme inhibitor Which antiepileptic drug is most likely to cause renal stones side-effects? Topiramate (The side effects of topiramate include: weight loss, renal stones and cognitive and behaviour changes). MRCPUK-part-1-September 2008 exam: H/O complex partial seizures, not able to tolerate either carbamazepine or sodium valproate. What is the most appropriate next line drug? Lamotrigine What is the likelihood of controlling seizures in a patient never previously on anti-epileptic medication? A study of patients with previously untreated epilepsy demonstrated that: • With a single first-line anti-convulsant agent →47% achieved control of seizures • 14% became seizure-free during treatment with a second or third drug. • An additional 3% became seizure-free with the use of two drugs simultaneously. If the person has myoclonic seizures or is suspected of having juvenile myoclonic epilepsy (JME), be aware that lamotrigine may exacerbate myoclonic seizures. Carbamazepine and oxcarbazepine but be aware of the risk of exacerbating myoclonic or absence seizures

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Juvenile myoclonic epilepsy Juvenile myoclonic epilepsy is the most common primary generalised epilepsy, but is underdiagnosed due to lack awareness of the condition by doctors Overview • is a common form of idiopathic generalised epilepsy, representing 10% of all patients with epilepsy. • typically, first manifests itself between the ages of 10 and 20 with brief episodes of involuntary muscle twitching occurring early in the morning. Genetic • The condition is genetically linked to the short arm of chromosome 6. Presentation • Bilateral symmetrical myoclonic jerks, primarily after awakening, without impaired consciousness • Generalized tonic-clonic seizures • Absence seizures

with impaired consciousness • Myoclonic jerks, especially of the upper limbs, which predominantly occur in the mornings shortly after waking (and may be so subtle as to be interpreted as 'clumsiness' when eating breakfast) • Triggers: sleep deprivation, alcohol consumption, flickering lights Investigations • Interictal EEG is diagnostic showing →generalised spike- and polyspike-wave activity; a photosensitive response may also be present Management • Female of childbearing potential 1st line → lamotrigine, levetiracetam or topiramate 2nd line → add lamotrigine, levetiracetam or topiramate • Male or female who are not of childbearing potential 1st line → sodium valproate 2nd line → add lamotrigine, levetiracetam, or topiramate Prognosis • Prognosis is extremely favourable if the condition is treated correctly, with many patients becoming seizure-free.

Status epilepticus Definition • ≥5 minutes of continuous seizure activity, or more than one seizure without recovery in between Treatment • Initial management: ABC. Maintain airway and circulation with intubation • 1st line anticonvulsant: two doses of benzodiazepines (Lorazepam is preferred). • 2nd line anticonvulsant: parenteral anti-epileptics (intravenous phenobarbital or phenytoin Fosphenytoin: (a pro-drug of phenytoin)

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Neurology advantages over phenytoin: it can be given IV or IM (phenytoin can only be given IV) can be given at infusion rates three times faster than phenytoin therapeutic levels are achieved within 10 minutes it has a lower incidence of adverse events than phenytoin. If the patient is already taking phenytoin, either IV phenytoin or fosphenytoin should still be given: it is likely that plasma levels are subtherapeutic. Phenytoin is not recommended in patients with underlying liver impairment therefore not used in status epilepticus secondary to alcohol withdrawal. • 3rd line: ICU for general anaesthesia (Midazolam or propofol) Monitoring: By EEG in unconscious patients to differentiate between sedation and nonconvulsive seizures → EEG pattern: Focal or focal with secondary generalization →nonconvulsive status epilepticus Generalized slowing, attenuation, lateralizing periodic discharges → postictal. If a patient in generalised status epilepticus does not respond to lorazepam and adequate doses of intravenous phenytoin, what is the next step in their management? Transfer to an Intensive Therapy Unit

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Epilepsy: pregnancy and breast feeding Overview • Epilepsy is not a contraindication to pregnancy. • the risks of uncontrolled epilepsy during pregnancy generally outweigh the risks of medication to the fetus Risk of congenital defects • Around 1-2% of newborns born to non-epileptic mothers have congenital defects. This rises to 3-4% if the mother takes antiepileptic medication. • All women thinking about becoming pregnant should be advised to take folic acid 5mg per day well before pregnancy until at least the end of the first trimester to minimise the risk of neural tube defects. What is the effect of pregnancy on epilepsy? • Two-thirds will not have seizure deterioration in pregnancy • The overall chance of postpartum seizures is relatively higher than during pregnancy. The use of phenytoin is not recommended in patients with underlying liver impairment therefore not used in status epilepticus secondary to alcohol withdrawal.

Management • Exposure to sodium valproate and other AED polytherapy should be minimised by changing the medication prior to conception. • We suggest NOT making changes to antiseizure drug regimen for the purpose of reducing teratogenic risk in established pregnancy • Aim for monotherapy. The lowest effective dose of the most appropriate AED should be used. • Some women who have been seizure free for a prolonged period may reasonably choose to discontinue antiseizure drug prior to conception. • Women with epilepsy taking AEDs who become unexpectedly pregnant: It is never recommended to stop or change AEDs abruptly without an informed discussion. • the levetiracetam has a favorable reproductive safety profile and has a broad spectrum of action across multiple seizure types. • If seizures are focal and begin after the first trimester, carbamazepine is another option. (carbamazepine often considered the least teratogenic of the older antiepileptics) • Sodium valproate should not be used during pregnancy and in women of childbearing age unless she is on a pregnancy prevention programme. Associated with neural tube defects and neurodevelopmental delay. • Phenytoin: associated with cleft palate It is advised that pregnant women taking phenytoin are given vitamin K in the last month of pregnancy to prevent clotting disorders in the newborn. • Lamotrigine: the rate of congenital malformations may be low. The dose of lamotrigine may need to be increased in pregnancy • Breast feeding is generally considered safe for mothers taking antiepileptics with the possible exception of the barbiturates

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Pseudoseizures Overview • Pseudoseizures are commonly misdiagnosed as true seizures and treated inappropriately with anti-epileptic drugs. • patients of any age can present with pseudoseizures. • features such as tongue biting and urinary incontinence are not absolute features of an organic seizure, they are often present in pseudoseizures. Factors favouring pseudoseizures • pelvic thrusting • family member with epilepsy • more common in females • accompanying underlying psychiatric concerns , e.g. crying after seizure, tearful around the time of the seizure. • attacks in public and absence of nocturnal events (don't occur when alone) Suspected psychogenic non-epileptic seizures →do Video-EEG recording

Neurology

• gradual onset • prolonged nature of the attacks (15-30 minutes) • Violent shaking • resistance to passive eye opening • very short post-ictal state • normal vital signs Factors favouring true epileptic seizures • tongue biting • raised serum prolactin Diagnosis • Video telemetry is useful for differentiating Treatment • Simple observation is the appropriate management.

Rett syndrome Overview • Rett syndrome is a neurodevelopmental disorder of the grey matter • inherited as an X-linked dominant disorder. • mostly affecting girls. Males affected by Rett syndrome die in utero or shortly after birth. • related to the MECP2 gene on the X chromosome Feature • Small hands and feet with deceleration of head growth. • Epileptic →repetitive hand

movements such as hand wringing. • loss of development, verbal abilities and cognition, ataxia • GI problems, such as constipation.

Tourette syndrome Definition • a chronic neurologic disorder that manifests with motor and vocal tics Epidemiology • Tourette syndrome presents before 18 years of age and many children grow out of it. • more common in males (4:1) Pathogenesis • due to genetic, environmental, and social factors resulting in an abnormality in the mesolimbic spinal system • the condition is familial in most cases Features • The motor tics often have a build-up that the patient is aware of, like an itch. • Commonly they involve blinking, throat clearing or shoulder shrugging. • Shouting of swear words is a typical vocal tic of Tourette's. Urinary incontinence can also occur in pseudoseizures, but tongue biting is rare

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Associated conditions • 90% of patients have a comorbid psychiatric disorder such as attention deficit hyperactivity disorder (~60% of cases) Management • first-line: Cognitive behavioural therapy • Second line: pharmacotherapy alpha-2 agonist (e.g., clonidine and guanfacine).

Huntington's disease (HD) Pathophysiology • Autosomal dominant →Increased number of CAG repeats (trinucleotide repeat disorder) in the huntingtin gene on chromosome 4 (coding for glutamine) → formation of abnormal proteins which have abnormal number of glutamine residues (huntingtin) → degeneration of GABAergic neurons (gamma-amino-butyric acid-ergic neurons in the striatum (particularly in the caudate nucleus) of the basal ganglia. • The striatum normally controls movement via inhibitory outputs to the globus pallidus internus. • Anticipation: increase in the number of CAG repeats in subsequent generations (The disease may develop earlier in life in each successive generation) Epidemiology • Symptom onset usually between 20 and 50 years of age Features • Personality changes (e.g. irritability, disinhibition, apathy, depression) and intellectual impairment (the earliest symptom) • Chorea Athetosis is a hyperkinetic movement symptom characterized by slow, involuntary, writhing movements. Huntington disease and cerebral palsy are the most common causes. • Lack of coordination and an unsteady gait • Dystonia • Saccadic eye movements • Dementia • Dopamine levels are increased • Gamma-aminobutyric acid levels are decreased • Acetylcholine levels in the central nervous system are decreased Diagnosis • DNA analysis is the most useful diagnostic test (e.g., via PCR) Trinucleotide CAG repeat expansion in the Huntington gene is diagnostic • MRI →caudate nucleus atrophy Atrophy of the caudate nucleus, putamen, and deep cerebral cortex are the hallmark features of Huntington's disease. Hydrocephalus ex vacuo Hydrocephalus ex vacuo is an expansion of the cerebral ventricles and surrounding subarachnoid space caused by atrophy of the underlying brain tissue, and not an expansion of CSF volume primarily.

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Neurology

 The role of neuroimaging is primarily to rule out other intracranial causes of a patient's symptoms, rather than to diagnose HD. Treatment • Tetrabenazine and reserpine works as a VMAT-inhibitor (vesicular monoamine transporter 2), involved in transportation of monoamines. It is indicated for Huntington's chorea to reduce hyperkinetic movements. • Haloperidol is a dopamine-2 antagonist used to treat movement disorders, hallucinations, and delusions in Huntington disease. Prognosis • progressive and incurable • Average life span after clinical onset is about 15 years (premature death). In Huntington disease, increased number of CAG repeats leads to the damage to the Caudate nucleus and results in decreased acetylcholine (Ach) and GABA. Cluster headache Epidemiology • More common in men (5:1) and smokers. • More common in younger males below the age of 40 Features • pain typical occurs once or twice a day, each episode lasting 15 mins - 2 hours • clusters typically last 4-12 weeks • intense pain around one eye (recurrent attacks 'always' affect same side) • The attacks are often nocturnal and are associated with parasympathetic overactivity. • patient is restless during an attack • accompanied by redness, lacrimation, lid swelling • nasal stuffiness • miosis and ptosis in a minority Management • Acute: 100% oxygen, subcutaneous or a nasal triptan the use of 100% oxygen at least 12 litres per minute via a non-rebreathable mask It is not recommended to offer paracetamol, NSAIDS, opioids, ergots or oral triptans for the acute treatment of a cluster headache. • prophylaxis: First line →verapamil, prednisolone , with other options including lithium, sodium valproate and gabapentin • NICE recommend seeking specialist advice from a neurologist if a patient develops cluster headaches with respect to neuroimaging

Notes & Notes for MRCP
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Differential diagnosis • The main differential is between cluster headaches and chronic paroxysmal hemicrania (CPH; which is treated with indomethacin). Distinguishing cluster headaches and Chronic Paroxysmal Hemicrania Cluster headache Chronic Paroxysmal Hemicrania more common in males more common in females frequency of attacks is 1 - 4 (maximum 8) in 24 hours. the frequency of attacks is higher, usually more than 15 in 24 hours The duration of headaches is (15-60 min). The duration of headaches is shorter (2-25 min) Not responds to indomethacin responds very well to indomethacin

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Neurology

Migraine Diagnostic criteria The International Headache Society has produced the following diagnostic criteria for migraine without aura: Point Criteria A At least 5 attacks fulfilling criteria B-D B Headache attacks lasting 4-72 hours* (untreated or unsuccessfully treated) C Headache has at least two of the following characteristics: •

1. unilateral location •
2. pulsating quality (i.e., varying with the heartbeat) •
3. moderate or severe pain intensity •

4. aggravation by or causing avoidance of routine physical activity (e.g., walking or climbing stairs) D During headache at least one of the following: •
5. nausea and/or vomiting •
6. photophobia and phonophobia E Not attributed to another disorder (history and examination do not suggest a secondary headache disorder or, if they do, it is ruled out by appropriate investigations or headache attacks do not occur for the first time in close temporal relation to the other disorder) • NICE suggests migraines may be unilateral or bilateral Migraine with aura • seen in around 25% of migraine patients • tends to be easier to diagnose with a typical aura being progressive in nature • may occur hours prior to the headache. • Typical aura include: transient hemianopic disturbance or a spreading scintillating scotoma ('jagged crescent'). Spreading (over minutes) sensory and motor symptoms Word-finding difficulties are also a common migraine aura symptom. autonomic symptoms such as a Horner syndrome negative auras of dark holes and tunnel vision Dizziness and fatigue are quite common prior to a migraine attack Patients may have mixed positive and negative auras. Positive auras include bright or shimmering light or shapes at the edge of their field of vision called scintillating scotoma. They can enlarge and fill the line of vision. Other positive aura experiences are zigzag lines or stars. • may occur with or without headache • NICE also give more detail about typical auras: are fully reversible develop over at least 5 minutes

 last 5-60 minutes • The following aura symptoms are atypical and may prompt further investigation/referral; motor weakness double vision visual symptoms affecting only one eye poor balance decreased level of consciousness. • Complicated migraine Complicated migraine is one which results in hemi sensory or hemi motor findings associated with a typical migraine presentation. • Confusional migraine involves alteration in sensorium rather than limb involvement. Other features: • family history of similar headaches is common • Bilateral fortification spectra Fortification spectra (jagged lines resembling battlements) and teichopsia (flashes) are common features of migraine. • Precipitation by oral contraceptives (contraindicated in migraine with aura) • Frequency reduced by tricyclic antidepressants (can be useful in the prophylaxis of migrain) • Third nerve palsy seen in ophthalmoplegic migraine ophthalmoplegic migraine was reclassified as a cranial neuralgia in the most recent International Headache Society classification. most commonly affects the third nerve, the deficits can be permanent. A subset of these patients will have gadolinium enhancement of the cisternal segment of the cranial nerve it is thought some of these patients actually have a demyelinating neuropathy. Migraine: management acute 5-HT agonists prophylaxis: β-blocker, 5-HT2 antagonist • 5-HT receptor agonists are used in the acute treatment of migraine • 5-HT receptor antagonists are used in prophylaxis. Acute treatment • first-line: combination of oral triptan and NSAID, OR oral triptan and paracetamol for young people aged 12-17 years: nasal triptan is preferred than oral triptan • if the above measures are not effective or not tolerated offer a non-oral preparation of metoclopramide* or prochlorperazine and consider adding a non-oral NSAID or triptan *caution should be exercised with young patients as acute dystonic reactions may develop with metoclopramide.

Neurology Prophylaxis • prophylaxis should be given if patients are experiencing 2 or more attacks per month. • Modern treatment is effective in about 60% of patients. • NICE advise either topiramate or propranolol or amitriptyline 'according to the person's preference, comorbidities and risk of adverse events'. Propranolol should be used in preference to topiramate in women of child bearing age as it may be teratogenic and it can reduce the effectiveness of hormonal contraceptives • if these measures fail NICE recommend 'a course of up to 10 sessions of acupuncture over 5-8 weeks' • gabapentin are not recommended now because evidence shows that it is not effective in preventing migraine. (NICE 2015) • NICE recommend: 'Advise people with migraine that riboflavin (400 mg once a day) may be effective in reducing migraine frequency and intensity for some people' riboflavin also known as vitamin B2 safe during pregnancy. • for women with predictable menstrual migraine treatment: NICE recommend either frovatriptan (2.5 mg twice a day) or zolmitriptan (2.5 mg twice or three times a day) as a type of 'mini-prophylaxis' • pizotifen is no longer recommend. Adverse effects such as weight gain & drowsiness are common Efficacy of Paracetamol in migraine • Migraine →↓gastric emptying →↓Paracetamol absorption →↓Paracetamol effects • Metoclopramide may be useful in accelerating gastric emptying. • paracetamol absorption technique is used to study gastric emptying. MRCPUK-part-1-January 2006 exam: Which type of medication would be most appropriate to reduce the frequency of migraine attacks?
 Beta-blocker (Topiramate is also recommended by NICE as first-line prophylaxis against migraine. However, a beta-blocker is a better choice in a female of child-bearing age)

Migraine: pregnancy, contraception and other hormonal factors Migraine during pregnancy • paracetamol 1g is first-line • aspirin 300mg or ibuprofen 400mg can be used second-line in the first and second trimester Migraine and the combined oral contraceptive (COC) pill • if patients have migraine with aura then the COC is absolutely contraindicated due to an increased risk of stroke (relative risk 8.72) Migraine and menstruation • many women find that the frequency and severity of migraines increase around the time of menstruation • SIGN recommends that women are treated with mefanamic acid or a combination of aspirin, paracetamol and caffeine. Triptans are also recommended in the acute situation Migraine and hormone replacement therapy (HRT) • safe to prescribe HRT for patients with a history of migraine but it may make migraines worse

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Triptans Action • Triptans are specific 5-HT1 agonists used in the acute treatment of migraine. • They are generally used first-line in combination therapy with an NSAID or paracetamol. Prescribing points • should be taken as soon as possible after the onset of headache, rather than at onset of aura • oral, orodispersible, nasal spray and subcutaneous injections are available Adverse effects • 'triptan sensations' - tingling, heat, tightness (e.g. throat and chest), heaviness, pressure Contraindications • patients with a history of, or significant risk factors for, ischaemic heart disease or cerebrovascular disease Epilepsy is not a contraindication to the use of triptans

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Idiopathic intracranial hypertension(IIH) Obese, young female with headaches / blurred vision : think idiopathic intracranial hypertension Postural headache but normal imaging →idiopathic intracranial hypertension Suspected Idiopathic intracranial hypertension →lumbar puncture to confirm the diagnosis is the next step Overview • also known as pseudotumour cerebri and formerly benign intracranial hypertension • classically seen in young, overweight females. Risk factors • obesity • female sex • pregnancy • drugs: oral contraceptive pill (eg: Dianette), Danazol (synthetic androgen used to treat endometriosis) steroids, tetracycline, vitamin A, Nalidixic acid *if intracranial hypertension is thought to occur secondary to a known causes (e.g. Medication) then it is of course not idiopathic

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