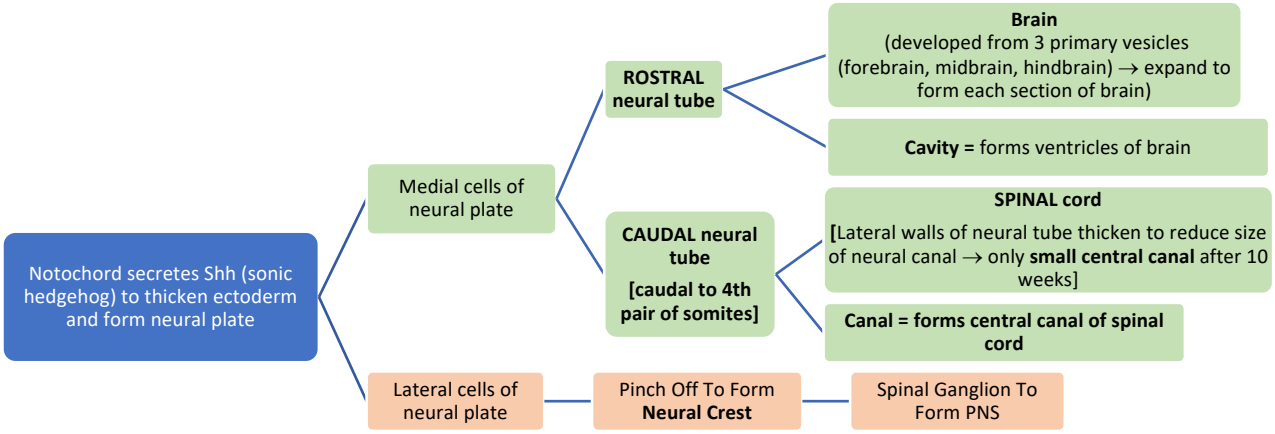
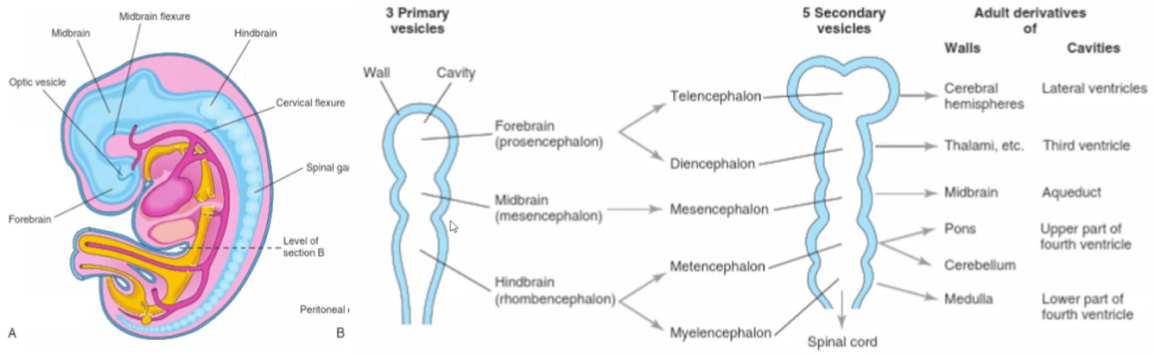
PAEDIATRIC NEUROLOGY

NEURO-EMBRYOLOGY

EMBRYOLOGICAL STEPS	EVENT
1. Gastrulation	forming the trilaminar layer of embryo
2. Neurulation	**Ectoderm** cells form brain, spinal cord and PNS
3. Somite development	Mesoderm cells form myotomes, dermatomes and sclerotomes
4. Cardiovascular system development	Heart tube develops from splanchnic mesoderm
5. Head and neck development	From 5 pairs of pharyngeal arches
6. Organogenetic period	From weeks 4-8



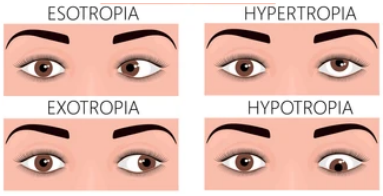


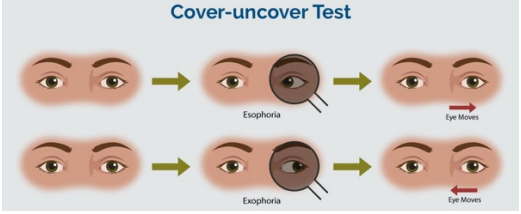
Neuroembryological defects:

CONDITION	Cause	Clinical observation	Symptoms
Spina bifida	When one or more neural arches fail to close during the 4th week of development = incomplete SC	Tuft of hair at back of spine	• Bowel obstruction • Faecal / urinary incontinence • Muscle weakness
Meroencephaly	Failure of rostral neuropore to close during the 4th week	Smaller head	• Forebrain, midbrain and hindbrain absent • Cognitive impairment • FTT – short stature

SQUINT (STRABISMUS)

PP	Types	Sx	Ix	Mx
• **Strabismus** / squint = misalignment of eyes • Young age allows for adaptation leading to one dominant eye vs one lazy eye *Causes* ➢ Hydrocephalus ➢ CP ➢ SoL (e.g. Rb) ➢ Trauma	• **Esotropia** (*affected eye towards nose)* • **Exotropia** (*affected eye towards ear)* • ***Hypertropia*** = *affected eye towards ear* (*upward moving affected eye)* • **Hypotropia** (*downward moving affected eye*)	• **Double vision** • **Amblyopia** (lazy eye becomes increasingly disconnected from brain)	Eye exam ➢ **Acuity** ➢ **Fields** ➢ **Reflexes** ➢ **Fundoscopy** (rule out Rb, cataracts + other retinal issues) **Hirschberg's test** = shine pen-torch 1m away – check if reflection is NOT central or symmetrical **Cover test** – see below	Treatment **BEFORE age of 8** as visual fields are still developing ➢ Pead opthal referral ➢ Occlusive patch to cover good eye ➢ Atropine drops on good eye ➢ Rx: cataracts and refractive errors

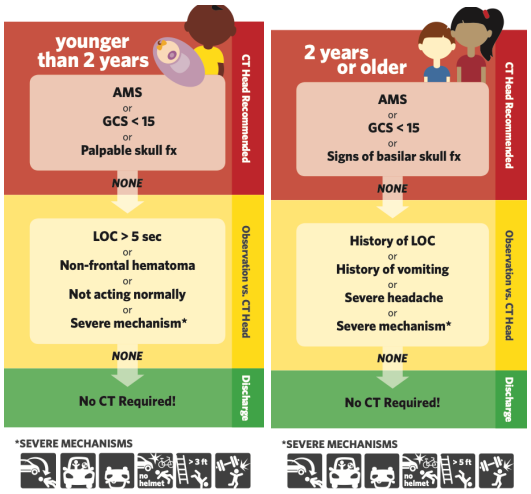




SYNCOPE WORK-UP – MAY CAUSES

	VASOVAGAL (Syncopal episodes)	BREATH-HOLDING SPELL	OTHER SYNCOPAL CAUSES
PP	ANS dysfn where strong stimulation of vagus nerve causes activation of PSNS ➢ Causing systemic vasodilatation, reduced cerebral circulation and hypoperfusion of brain tissue	Involuntary episodes when child holds breath usually after upsetting or scaring them ➢ Usu. between 6 and 18/12 old	
Cause	**Primary syncope (simple fainting)** ➢ dehydration ➢ hypoglycemia (missed meals) ➢ extended standing in warm environ. ➢ ++ emotions = pain, sudden surprise, sight of blood **Secondary causes** ➢ dehydration ➢ hypoglycaemia ➢ anaemia ➢ infection ➢ anaphylaxis ➢ arrhythmias ➢ valvular heart disease or HOCM	*Idiopathic* ➢ child upset, frightened or pain ***DDx:*** ➢ Fe def. (↑ risk of BHS) ➢ Seizure ➢ Cardiac?	
Sx	***Prodrome*** • Hot/clammy • Dizzy / lightheaded • Blurry or dark vision • Headache ***Event*** • Sudden LOC – fall to ground • Twitching/shaking/convulsion? ***Post-ictal*** • No WILD • Rapid recovery + memory of events	***2 types*** **1) Cyanotic breath holding spells** *upset or worked up child letting out long cry causing them to stop breathing AND LOC* - *recover within min but lethargy* **2) Reflex anoxic seizures (*pallid breath holding spells)*** *startled child causes vagus nerve to send strong signals to heart to stop beating* - *turns pale, LOC +/- twitching* - *resolves within 30s*	
Comp.	None - Falls risk? – bleeding disorder	None - Most outgrow by 4 or 5 yo	
Ix	➢ **Check for physical injuries** ➢ **Identify possible concurrent illnesses** o ECG (arrythmia, long QT) o 24 hr ECG (if paroxysmal arrhythmias suspected) o Bloods (FBC, EUC, BSL) o ECHO	**Exclude other pathologies** ➢ FBC ➢ EUC ➢ LFT ➢ BSL ➢ Fe studies ➢ ECG ➢ UA	
Mx	➢ Reassure – most resolve by adulthood (more common in teenager girls) *Lifestyle advice* ➢ Avoid dehydration ➢ Avoid Skipping meals ➢ Avoid standing for long periods of time If seizures or underlying pathology present refer to appropriate specialist	Breath holding spells linked with Fe def. anaemia ➢ Self-limiting and not harmful ➢ Rx for Fe deficiency ➢ **First aid** – protect head and limbs from injury	

Indications for Paediatric CT (PECARN score)



Under 2 Years Old	Between 2 and 18 Years Old
Altered mental status	Altered mental status
Scalp hematoma	LOC
Loss of Consciousness ≥ 5 seconds	History of vomiting
Severe mechanism of injury	Clinical signs of basilar skull fracture
Palpable skull fracture	Severe mechanism of injury
Abnormal behavior per parent	Severe headache

HEAD INJURIES

- **Raccoon eyes (peri-orbital) or battle sign (mastoid)**
- **CSF otorrhea, rhinorrhoea**
- **FND = RAPD, abnormal posture, GCS < 13**
- **Examine C-spine**

REVERSIBLE CAUSES

1) **HYPOglycaemia (< 3.5mM)**
   - **Jitteriness in babies (seen if BSL < 2mM)**
   - **Causes** = sepsis (for preceding illness), T1DM, congenital diseases, alcohol OD
   - **Rx: IV 10% dextroses (2mL/kg) or glucogel**
   - **AIM TO AVOID BRAIN DAMAGE**
2) **HypoNa (< 125mM)**
   - **Replace with 1-2mM 0.9% NS**
   - Replace 3mL / 3% NS (if severe)
   - *Avoid risk of ODS (pontine demyelination)
3) **Raised ICP**
   - Cushing's triad (irregular RR, widened PP, bradycardia)
   - **Rx: IV mannitol and HoB elevation**
   - **Ix:** CT brain + fundoscopy
4) **Infection**
   - **Locate source –** FBC, EUC, LFT, CRP, ABG, Blood culture, CXR, Urine MSU (M/C/S), swabs
   - **Rx: IV ceftriaxone or acyclovir**

HEADACHES IN PAEDIATRICS

	Assoc.	Treatment for migraines
Classical MIGRAINES	• **Unilateral severe throbbing headache** • Visual aura AND/OR photophobia/phonophobia • N/V +/- abdo pain • May have had **recurrent central abdo pain as child**	**Acute Mx:** ➢ Rest, fluids and low stimulus environment ➢ paracetamol ➢ Triptans → 50mg sumi ➢ NSAIDs ➢ Anti-emetics (maxolon) **Long-term Mx:** ➢ Avoid trigger (stress, lights, smells, dehydration, choc, critic acid, poor sleep) ➢ CBT ➢ Headache diary ➢ Relaxation (massage) ➢ **Vitamin B2 (riboflavin) –** reduce freq. + severity ➢ **Amitriptyline (TCA) but AE** = fatigue, dizzy, depression, insomnia ➢ **Prophylaxis (with Panadol + propranolol]**
Abdominal Migraines	• CENTRAL ABDOMINAL PAIN > 1 hr • Nausea/ vomiting • Anorexia + Pallor • Headache • **May develop into classical migraines during adulthood**	
Infections	Identify cause • Viral URTI, otitis media, sinusitis, tonsillitis	
Sinusitis	• Facial pain – behind nose, forehead and eyes (over respective sinuses – ethmoid, sphenoid, maxillary and frontal) • Coryza - nasal congestion, rhinorrhea, lacrimation • Rx: supportive and resolves within 2-3 weeks	

*Consider SoL – if suspicious findings (e.g. UWL, persistent headache, FND)

EPILEPSY

Def

- Umbrella term for condition where there is a tendency to have siezures
- Seizure = transient episodes of abnormal electrical activity in brain

Types

Age group	Description	Duration	1st line med	2nd line med
Simple febrile convulsions **(6/12 – 6 yo)**	➢ Generalized tonic-clonic seizures ➢ 1.8% of general population – no lasting damage on brain ➢ 1 in 3 will have another febrile convulsion	**< 15mins**	**Identify + manage cause** (usu. infection – tonsillitis, AOM) ➢ Regular analgesia ➢ NO further investigations ➢ Educate on acute Mx (see below)	
Complex febrile convulsions **(6/12 – 6 yo)**	➢ If partial or focal seizures ➢ OR multiple seizure in same febrile illness	**> 15 mins**	May require Ix	
Focal seizures	**Begin in temporal lobe:** ➢ Hallucinations, memory flash backs ➢ Déjà vu ➢ **Automatism** = *strange actions on autopilot*	**30-180s**	Lamotrigine Carbamazepine	**Sodium valproate** Levetiracetam
Generalised tonic-clonic "Grand mal"	➢ LOC + tonic-clonic jerks (tonic before clonic) ➢ WILD – post-ictal period	**3-5mins**	**Sodium valproate**	Lamotrigine Carbamazepine
Absence *"petit mal"*	**Typically children** ➢ **Typical** = Blank stare into space ONLY ➢ **Atypical** = plus automatisms	**< 10s**	**Sodium valproate** Ethosuximide	
Atonic **Lennox-Gestaut syndrome** (2-5yo)	**"drop attacks" = brief lapses in muscle tone**	**< 3mins**	**Sodium valproate**	Lamotrigine
Infantile spasms (west syndrome) **Infancy → 6/12 old**	➢ **Rare (1 in 4000)** ➢ **Full body spasms** ➢ **Bad prognosis – 1/3rd die by age 25**		**Prednisone** **Vigabatrin**	
Myoclonic seizures ***(juvenile myoclonic epilepsy)***	➢ **Sudden brief muscle contraction like a "sudden jump"** ➢ **Remains conscious** ➢ *DDx: CJD (mad cow disease)*		**Sodium valproate**	Lamotrigine Levetiracetam Topiramate
Status epilepticus (medical emergency)	**Defined as any seizure that:** ➢ Lasts longer than 5 mins OR ➢ More than 3 seizures in 1 hour **Stages:** ➢ **stage 1 = acidosis** ➢ **stage 2 = hypoglycaemia**	**>5mins**	**ABCDE** ➢ Secure airway ➢ High FIO2 ➢ Assess CV and Resp ➢ **Check BSL** ➢ IV lorazepam 4mg (repeat after 10 mins if seizure persists) ➢ Switch to IV phenytoin (if seizure persists) ***Can use buccal midazolam or PR diazepam (in community) → flumazenil (antidote to midazolam)**	
Non-epileptic seizures **(PNES)**	• Type of conversion disorder • May be caused by Factitious disorder and malingering where patient is purposely deceiving the physician • **NO AUTOMATISM, NO TONGUE BITING AND LASTS OVERLY LONG!**	5-10mins	About 9% to 15% of patients with psychogenic events have coexistent seizure disorders ➢ Psych counselling	

Ix

General Bloods

- **Postural BP**
- **Capillary glucose & Hba1C** (*exc. hypoglycaemia)*
- **EUC + VBG** (measure lactate + acidosis + BSL)
  - **?hypoBSL, hypoCa, hyperUrea**
- **CK & Prolactin** (both elevated in generalised tonic-clonic seizure**s**)
- **Cultures (?septic screen)**
  - **Blood M/C/S**
  - **Urine M/C/S**

Imaging

- **CT Brain →** *ICH, tumours, SoL*
- **CXR = aspiration pneumonia**
- **MRI →** visualise brain structure = SoL, *infarcts*
  - *First seizure in child < 2yo*
  - *Focal seizures*
  - *NO response to 1st line anti-epileptics*
- **EEG → DDx:** idiopathic generalized epilepsy from PNES,
  - **NB: low sensitivity/specificity →** abnormal EEG does NOT indicate brain pathology NOR does a normal EEG rule out epilepsy/seizure disorder

Other

- **ECG** (*prolonged QT interval)*
- **LP** (*if meningitis/encephalitis suspected*)
  - **Check CT / Papilloedema**

Neurology referral if:

- **1st seizure →** *most epilepsies treatable with 1st agent*
- **Refractory epilepsy**
- **Surgery**

A/E of meds

Sodium valproate	Carbamazepine	Ethosuximide	Lamotrigine	Phenytoin
↑ GABA activity and ↓ GABA transaminase	**Na channel blocker**	**Ca channel blocker**	**Na channel blocker**	**Na channel blocker**
➢ **Teratogenic** so patients need careful advice about contraception ➢ Liver damage and hepatitis ➢ Hair loss ➢ Tremor	➢ **Agranulocytosis** ➢ Aplastic anaemia ➢ Induces the P450 system so there are many drug interactions ➢ SJS (if mutant HLA B1502)	➢ Night terrors ➢ Rashes	➢ Stevens-Johnson syndrome or DRESS syndrome. These are life threatening skin rashes. ➢ Leukopenia Nb: hormonal agents reduce effectiveness	➢ Folate and vitamin D deficiency ➢ Megaloblastic anaemia (folate deficiency) ➢ Osteomalacia (vitamin D deficiency) ➢ Affect cerebellar function

Lifestyle impacts & legal considerations

Acute Mx for any seizure

1. Place patient in safe position (e.g. carpeted floor) in recovery position
2. Place soft pillow under head to prevent head injury
3. Time start and end of seizure
4. Call ambulance if:
   a. lasting > 5 mins or
   b. if this is 1st seizure

ABCDE approach for status epilepticus

See above

Long-term

- Take showers rather than baths
- Supervised when swimming
- Cautious with heights
- Cautious with traffic
- Cautious with any heavy, hot or electrical equipment

For teenagers

➢ Avoid driving unless they meet specific criteria to demonstrate their control of epilepsy

CEREBRAL PALSY

Def

- Permanent neurological problems due to brain damage around birth period
- **NOT progressive BUT symptoms do change over time**

Types

Spastic	Dyskinetic (athetoid OR extrapyramidal CP)	Ataxic	Mixed
• **HYPERTONIA** • **UMN damage**	• Issues w/ muscle tones (hyper + hypotonia) • Athetoid or oro-motor problems • Damaged basal ganglia	• Poor coordinated movement • Damage to cerebellum	Mix of spastic, dyskinetic and/or ataxic features

Causes

	Ante-natal	Peri-natal	Post-natal
Cause	➢ Maternal infections ➢ Trauma during pregnancy	➢ Birth asphyxia – HIE, IVH ➢ Pre-term birth	➢ Meningitis ➢ Severe neonatal jaundice ➢ Head injury

Sx

GENERAL Sx
- **Failure to meet milestones**
- **Increased/decreased tone**
- **Hand preference below 18 mths**
- **Coordination and speech issues**
- **Feeding or swallowing problems**
- **Learning difficulties**

Neurological issues
- **Hemiplegic gait** = UMN lesion
- **BROAD-based /ataxis gait** = cerebellar issue
- **High-stepping gait (foot drop)** = LMN lesion
- **Waddling gait** = pelvic muscle weakness = MD
- **Antalgic gait** → fracture, trauma, septic arthritis,

DDx for UMN lesion
- **Brain tumour**
- **Brain trauma (ICH)**

Ix

- **CLINICAL DIAGNOSIS**

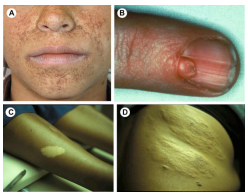
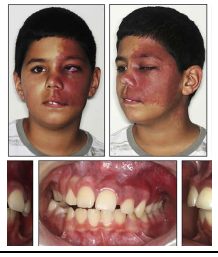
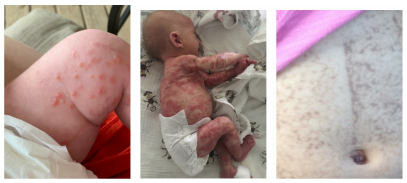
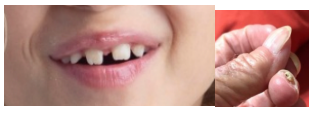
Comp.

- Learning disability
- Epilepsy
- Kyphoscoliosis
- Muscle contracture
- Hearing and visual impairment
- GORD

Lifestyle impacts & legal considerations

MDT management
- **PT** → strength and stretch muscles to prevent contractures
- **OT** → suggest strategies for ADL → e.g. getting dressed and using bathroom
- **Speech therapist** – *speech and swallowing (may need PEG or NGT)*
- **Social workers**
- **Dieticians**
- **Orthopaedic surgeons** – release contractures and lengthen tendons (tenotomy)
- **Paediatricians** – coordinate care and optimise meds
  - **Muscle relaxants** (e.g. baclofen) for muscle spasticity & contracture
  - **Anti-epileptics** for seizures
  - **Glycopyrronium bromide** for XS drooling

PAEDIATRIC NEUROCUTANEOUS SYNDROMES – CNS +skin issues (ectodermal derived)

	NEUROFIBROMATOSIS	TUBEROUS SCLEROSIS	STURGE-WEBER	Incontinentia Pigmenti
PP	Autosomal dominant ➢ NF1 – RAS mutant in Chr 17 ➢ NF2 - neurofibromin II in chr 22 (loss of cancer growth inhibition)	Autosomal dominant ➢ Variable expression ➢ TSC 1 = chr 8 ➢ TSC 2 = chr 16	Idiopathic / sporadic ➢ SOMATIC mutations ➢ Children and adults	X-linked dominant ➢ Females affected ONLY ➢ Males die early
Sx	**NF1** ➢ ≥2 benign SC neurofibromas -fleshy pedunculated growths ➢ ≥ 6x café au lait spots ➢ Axillary freckling (aged 3-5) ➢ Lisch nodules (in iris) **NF2 (rarer)** ➢ Schwannomas = acoustic neuroma – CN8 pathologies ○ Vertigo, tinnitus, ataxia, imbalance, headache ➢ Peripheral neuropathies – foot drop ➢ SC nodules ➢ Juvenile cataracts	• **Ungual fibromas** (≥2x and > 5mm) • **Ash leaf spots** (hypomelanotic) • **Shagreen patches** (usu. on back) • **Adenoma sebaceum** 	**Capillary malformations lead to:** ➢ Port-wine sign on face (1st and 2nd division of trigeminal) ➢ Episcleral haemangiomas ➢ leptomeningeal angiomas (calcifications in brain) 	***4 stages of skin issues*** ➢ **Stage 1** = linear red vesiculo-pustular rash ➢ **Stage 2** = wart-like skin papules on distal limbs and scalp ➢ **Stage 3**= hyperpigmented skin ➢ **Stage 4** = atrophic scarring and lowss of hair (mostly in lower legs) **Other minor criteria** ➢ **Hypotonia** (*low number of teeth*) ➢ **Nail dystrophy** (*damage or lose shape*) 
Comp.	➢ Long-bone dysplasia ➢ Short stature ➢ Seizures ➢ Macrocephaly	➢ Cardiac rhabdomyomas (HF, arrythmias) ➢ Renal angiomyolipoma (haematuria, CKD) ➢ Epilepsy (from cortical tubules)	GLAUCOMA Mental retardation	
Ix	NF1 – Refer to clinical criteria NF2 - CT brain – vestibular scwhannomas	MRI = calcified cortical tubes in periventricular spaces	BRAIN MRI – Tram track calcification	Molecular genetic testing
Mx	➢ Psych support, genetic counselling ➢ Optic glioma → chemo ➢ Schwannoma → surgical excision		NONE	**Skin lesions DDx** ➢ **Impetigo,** ➢ **Chicken pox** ➢ **Scabies**

PAEDIATRIC NEUROSURGERY

	HYDROCEPHALUS	CRANIOSYNOTOSIS	PLAGIACEPHALY & BRACHYCEPHALY
PP	CSF accumulation in cerebral ventricles ➢ **Communicating** = overproduction or decreased CSF reabsorption of arachnoid villi ➢ **Non-communicating** = blockage	Skull sutures fuse prematurely causing ABNORMAL head shapes ➢ **Sagittal synostosis** = long and narrow from front to back ➢ **Coronal synostosis** – bulging one side of forehead ➢ **Metopic synostosis** – pointy triangular forehead ➢ **Lamboid synostosis** – flattening on one side of occiput	**Positional** plagiocephaly → Abnormal head shapes ➢ Plagio = oblique / slanted ➢ Brachycephaly = flattened back of head
Sx	• **Bulging anterior fontanelle** *suture not fused until 2yo normally • Poor feeding and vomiting • Poor tone • sleepiness	• Anterior fontanelle closes before 1yo • Small head compared to rest of body DDx: ➢ Plagiocephaly ➢ **Congenital muscular torticollis (CMT) - shortened SCM on one side**	➢ 3-6/12 babies ➢ **Preferences to sleep on one side** DDx: ➢ Craniosynotsoses ➢ **Congenital muscular torticollis (CMT) - shortened SCM on one side**
Comp.	**Raised ICP** → vomit, visual impaired, cognitive impaired, dev. delay		➢ Sudden infant death syndrome (SIDS)
Ix	Clinical Dx ➢ CT brain	Skull XR CT head – exclude or confirm Dx if XR in doubt	Clinical Dx
Mx	VP shunt ➢ Connect SAS with peritoneal cavity ➢ Valve located subcutaneously connects SAS to peritoneal cavity to regulate amount of CSF drained **Main complications of VP shunting** ➢ Infection ➢ Blockage or XS drainage ➢ Intraventricular heamorrhage ➢ Outgrowing shunts	Surgical reconstruction – *endoscopic craniotomy* ➢ *Only offered if aged < 3 mths -bones not fused* ➢ Lifelong scar	***Reassurance – advise on simple lifestyle measures*** ➢ Positioning them on rounded side for sleep ➢ Supervised tummy time ➢ Rolled towels or props ➢ Minimising time in push chair or car seats ***If persisting:*** ➢ Physiotherapy ➢ Plagiocephaly helmets = risk of contact dermatitis and psychosocial problems

	SPINA BIFIDA	MUSCULAR DYSTROPHY	SPINAL MUSCULAR ATROPHY
PP	Neural tube defect causing incomplete neural tube fusion **Different types** ➢ Incomplete SB ➢ Failed cranial closure = anecephaly ➢ No closure = cranio-rachischisis (incompatible for life)	Umbrella term for genetic conditions causing gradual weakening and wasting of muscles ➢ **DMD** ➢ Becker's MD ➢ Myotonic Dystrophy ➢ Facioscapulohumeral MD ➢ Oculopharngeal MD ➢ Limb-girdle -MD ➢ Emery-Dreifuss MD	Autosomal recessive ➢ Loss of LMN **Onset** / **LE** **SMA Type 1**: 1st months of life / 2 years **SMA Type 2**: 1st 18 months [most never walk] / adulthood **SMA Type 3**: After 1st year of life / Normal **SMA type 4**: Onset in 20s [sig. fatigue] /
RF	• Assoc. with B12 and folate def. (e.g. meds = MTX, anti-epileptics, trimethoprim)	• FHx	➢ Consaguionous parents ➢ FHx
Sx	Spectrum of symptom severity ➢ **Asymptomatic** → spina bifida occulta (sacral dimple or tuft of hair are subtle sign) ➢ **Meningocele** → no neuro issues ➢ **Myelomeningocele** → exposed SC nerves and meninges (causing lower limb neuro issues)	**Gower's sign** = proximal muscle weakness – lying to stand up [**hands on legs to stand up**] **DMD**: 3-5yo [X-linked recessive] mostly males affected, females are carriers **Becker's**: 8-12yo; Dystrophin gene less affected – walk with assistance **Myotonic**: Adulthood; ➢ Prolonged tetanus [cannot let go after shaking one's hand]; ➢ Cataracts + arrythmias **FSH**: Childhood weakness around face, progressing to shoulder and arms; Cannot puff cheeks w/o air leaking + **OPMD**: Bilateral ptosis + ophthalmoplegia; Dysphagia **LGMD**: Teenager – weakness in shoulder and hip girdles **EDMD**: Childhood – contractures in elbows and ankles causing restricted ROM; ➢ Affects upper arms and lower limbs 1st	➢ **Progressive muscle weakness** ➢ **Hypotonia + reduced muscle bulk** ➢ **fasciculations** ➢ **Reduced /absent reflexes**
Comp.	➢ Neurogenic bladder ➢ Hydrocephalus ➢ Chiari malformation = speech, weakness, SOB ➢ Impaired gait/immobility (scoliosis)	➢ Respiratory failure ➢ Spinal scoliosis ➢ Heart Failure	Respiratory failure
Ix	**Ante-natal screening** ➢ Fetal USS at 15 or 20 weeks (morphology) ➢ CVS or amniocentesis **Newborn exam** ➢ Bulging fontanelle ➢ Sacral dimple or tuft of hair ➢ Confirm w/ head CT or USS	Muscle biopsy • Genetic Testing	Nerve conduction studies Genetic Testing
Mx	***Prevention*** ➢ 0.5mg folic acid (5mg for GDM, epilepsy etc.) ➢ Folate rich fluids = spinach, watermelon, banana ***Surgery + manage complications*** ➢ MDT ➢ VP shunt – hydrocephalus ➢ Intermittent catheter → neurogenic bladder	**MDT approach – supportive** ➢ OT, PT and medical appliances (e.g. wheelchairs, braces) ➢ PO steroids – slow progression of muscle weakness ➢ Creatinine supp. – slight improvement on muscle strength	**MDT approach – supportive** ➢ PT – maximise muscles and retain resp. function ➢ Resp. support (non-invasive ventilation) esp. during sleep o SMA type 1 children – may need tracheostomy + mech ventilation to extend life ➢ PEG – for weak swallow