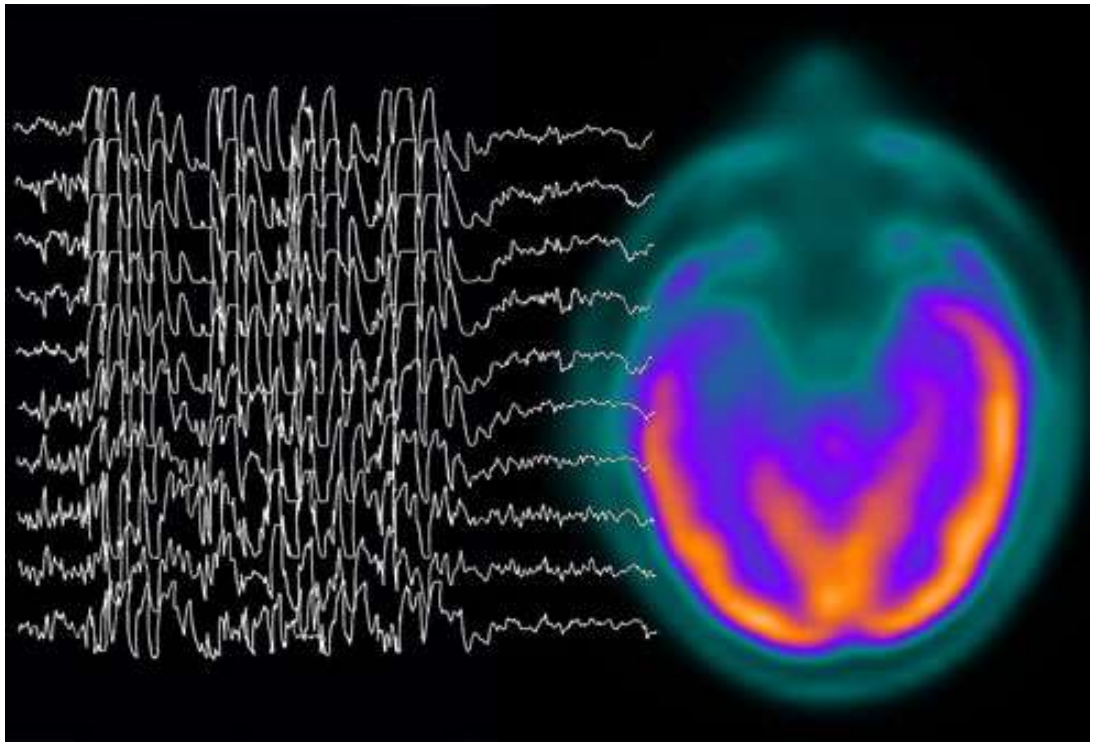
EPILEPSY AND EPILEPSY IMITATORS



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Outlines

- Epilepsy
  - definition and new classification
- Epilepsy imitators
  - Headache
  - Other imitators

EPILEPSY


EXAS

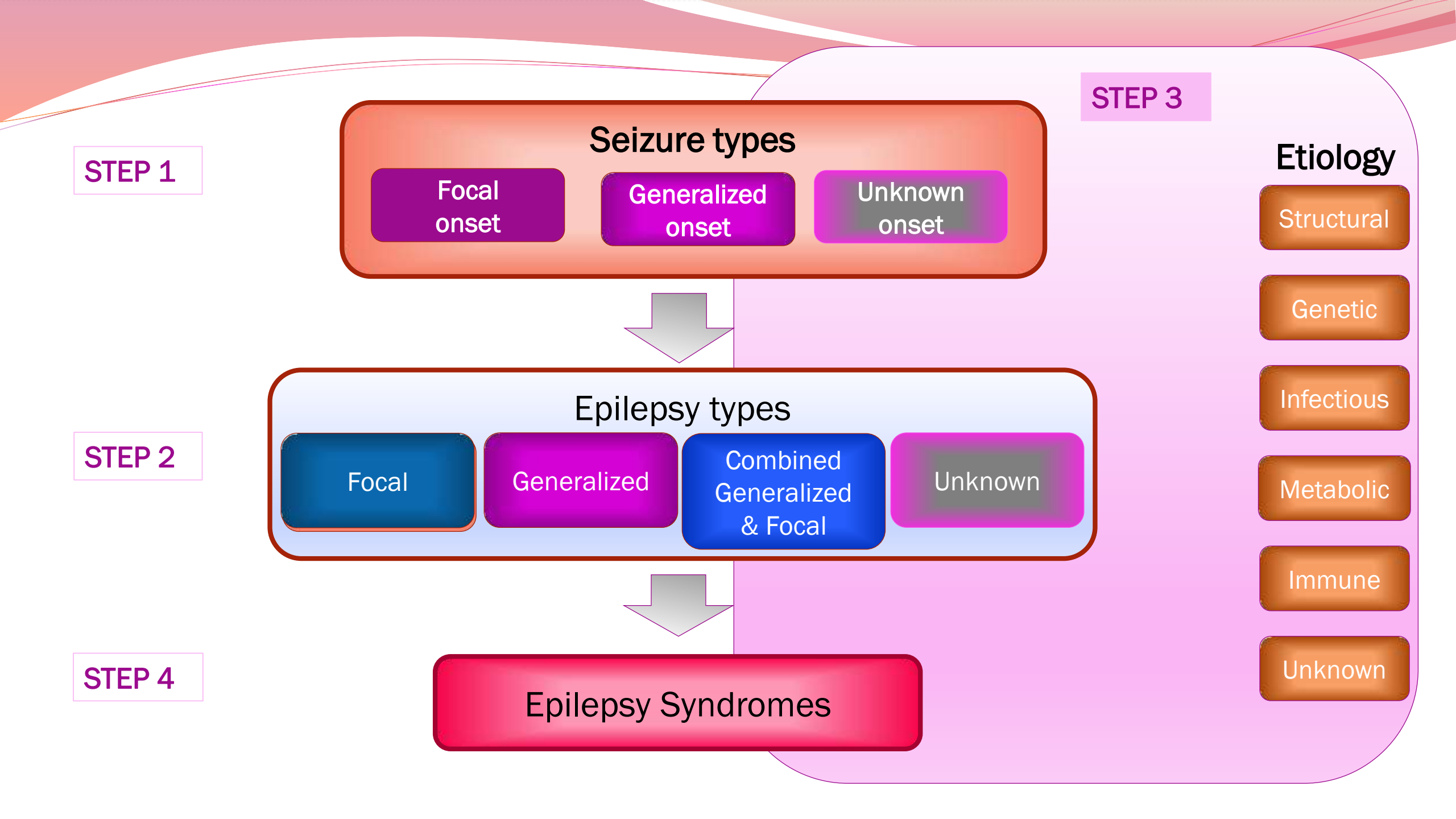
DEFINITION

Seizure:

Is a transient occurrence of signs and/or symptoms due to abnormal excessive or synchronous neuronal activity in the brain.

Epilepsy : is a disease of the brain defined by any of the following conditions:

- At least two unprovoked (or reflex) seizures occurring more than 24 hours apart
- One unprovoked (or reflex) seizure and a probability of further seizures similar to the general recurrence risk (at least 60%) after two unprovoked seizures, occurring over the next 10 years
- Diagnosis of an epilepsy syndrome


STEP 1
Seizure types
Focal onset
Generalized onset
Unknown onset
STEP 3
Etiology
Structural
Genetic
Infectious
Metabolic
Immune
Unknown
STEP 2
Epilepsy types
Focal
Generalized
Combined Generalized & Focal
Unknown
STEP 4
Epilepsy Syndromes

ILAE 2017 Classification of Seizure Types Expanded Version [1]

Focal Onset

Aware | Impaired Awareness

Motor Onset
- automatisms
- atonic [2]
- clonic
- epileptic spasms [2]
- hyperkinetic
- myoclonic
- tonic

Nonmotor Onset
- autonomic
- behavior arrest
- cognitive
- emotional
- sensory

focal to bilateral tonic–clonic

Generalized Onset

Motor
- tonic–clonic
- clonic
- tonic
- myoclonic
- myoclonic–tonic–clonic
- myoclonic–atonic
- atonic
- epileptic spasms

Nonmotor (absence)
- typical
- atypical
- myoclonic
- eyelid myoclonia

Unknown Onset

Motor
- tonic–clonic
- epileptic spasms

Nonmotor
- behavior arrest

Unclassified [3]

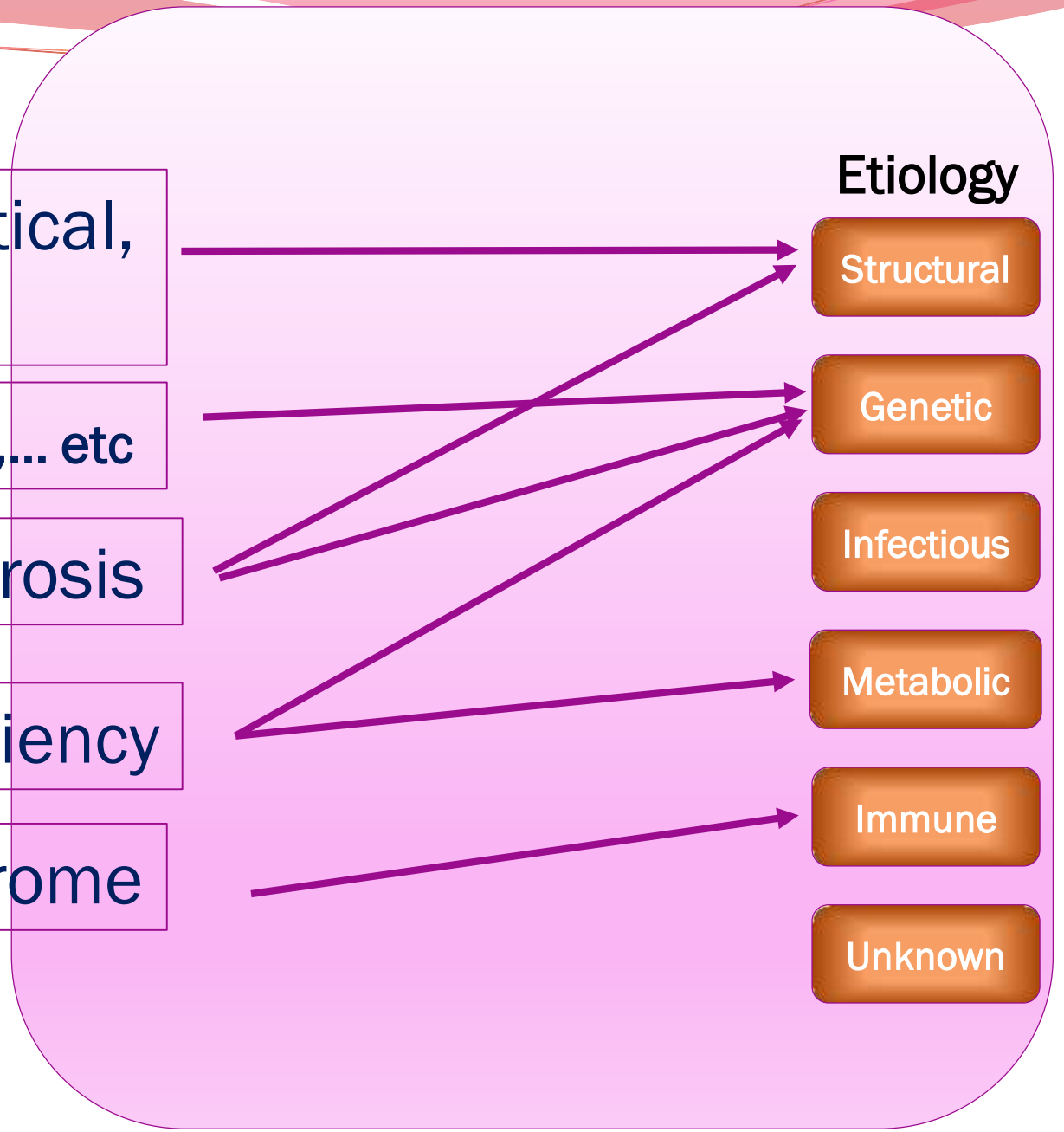
Vascular, trauma, tumor, cortical, hypoxic ischemic … etc

Trisomies, angelman,KLEINFELTERS,… etc

Tuberous Sclerosis

GLUT1 deficiency

Rasmussen syndrome



Epilepsy Syndromes

- Identified *based on age at onset*, *seizure type(s)*, *EEG characteristics*, *etiology*, and other associated factors

Neonatal/infantile

- *benign familial neonatal epilepsy,*
- *early myoclonic encephalopathy*
- *Ohtahara syndrome*
- *Dravet syndrome*
- *Myoclonic epilepsy of infancy*
- ----- *etc*

childhood

✓Febrile seizures plus
✓Panayiotopoulos syndrome
✓Epilepsy with myoclonic atonic (previously astatic) seizures
✓Benign epilepsy with centrotemporal spikes
✓Autosomal-dominant nocturnal frontal lobe epilepsy
✓Epilepsy with myoclonic absences
✓Lennox-Gastaut syndrome
✓------ etc

Adolescent/adult

✓juvenile absence epilepsy
✓juvenile myoclonic epilepsy
✓epilepsy with generalized tonic-clonic seizures alone
✓progressive myoclonus epilepsies
✓autosomal dominant epilepsy with auditory features
✓----- etc

Variable age

- Familial focal epilepsy with variable foci
- reflex epilepsies

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- **Developmental encephalopathy**: where there is just developmental impairment without frequent epileptic activity associated with regression or further slowing of development;
- **Epileptic encephalopathy:** where there is no preexisting developmental delay and the genetic mutation is not thought to cause slowing in its own right; and developmental and epileptic encephalopathy where both factors play a role.

Often it may not be possible to disentangle whether the epileptic or developmental component is more important in contributing to a patient's presentation.

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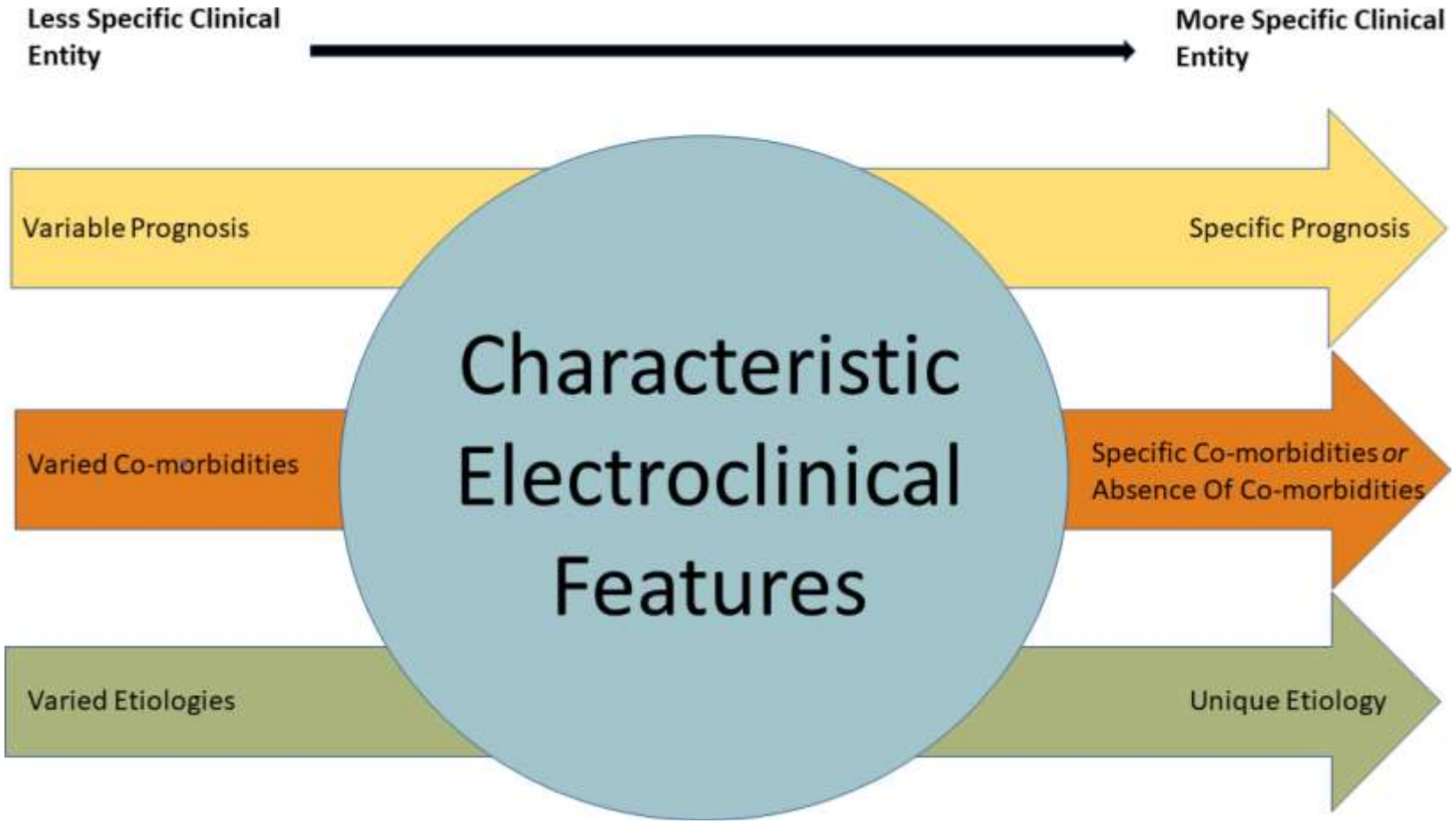
- **Thus "benign," as a descriptor for epilepsy, is replaced by both "self-limited" and "pharmacoresponsive,"**

- each replacing different components of the meaning of benign. "Self-limited" refers to the likely spontaneous resolution of a syndrome. "Pharmacoresponsive" means that the epilepsy syndrome will be likely to be controlled with appropriate antiepileptic therapy.
- It is important to acknowledge, however, that there will be individuals with these syndromes who are not pharmacoresponsive

ILAE 2022 epilepsy syndrome



ILAE 2022 epilepsy syndromes

- **We defined an epilepsy syndrome** as “a characteristic cluster of clinical and EEG features, often supported
- by specific etiological findings (structural, genetic,metabolic, immune, and infectious).”
- **The diagnosis of a syndrome** in an individual with epilepsy frequently carries prognostic and treatment implications. Syndromes often have age-dependent presentations, meaning that they typically
- start at specific ages, and in some cases may also remitat certain ages.
- **Many syndromes are strongly correlated** with a range of specific intellectual, psychiatric, and other comorbidities, whereas in other syndromes, the absence of such comorbidities is a characteristic feature (Figure).**Epilepsy syndromes have traditionally been grouped according to age at onset**. Accordingly, the ILAE position papers describe separately syndromes with onset in neonates and infants (up to age 2 years), syndromes with onset in childhood, and syndromes that may begin at variable ages (meaning in both pediatric and adult patients).

The syndromes are further subdivided into generalized,focal, or generalized and focal, based on
- seizuretype(s), with a separate category for syndromes with developmental and epileptic encephalopathy (DEE) or progressive neurological deterioration.

ILAE 2022 epilepsy syndromes in neonates and infants.

Self-limited epilepsies

- Self-limited neonatal epilepsy (SeLNE)
- Self-limited familial neonatal-infantile epilepsy (SeLFNIE)
- Self-limited infantile epilepsy (SeLIE)
- Genetic epilepsy with febrile seizures plus (GEFS+)
- Myoclonic epilepsy in infancy (MEI)

Developmental and epileptic encephalopathies (DEE)

- Ealy infantile developmental and epileptic encephalopathy (EIDEE)
- Epilepsy in infancy with migrating focal seizures (EIMFS)
- Infantile epileptic spasms syndrome (IESS)
- Dravet syndrome (DS)

Etiology-specific syndromes

- *KCNQ2*-DEE
- Pyridoxine-dependent (*ALDH7A1*)-DEE (PD-DEE)
- Pyridox(am)ine 5'-Phosphate Deficiency (PNPO)-DEE (P5PD-DEE)
- *CDKL5*-DEE
- *PCDH19* clustering epilepsy
- Glucose Transporter 1 Deficiency Syndrome (GLUT1DS)
- Sturge Weber syndrome (SWS)
- Gelastic seizures with hypothalamic hamartoma (GS-HH)

- Ac syndrome
- the childhood onset epilepsy syndromes, most of which have both mandatory seizure type(s) and interictal electroencephalographic (EEG) features. Based on the 2017

Classification of Seizures and Epilepsies, some syndrome names have been updated using terms directly describing the seizure semiology. Epilepsy syndromes beginnin

- in childhood have been divided into three categories: (1) self-limited focal epilepsiescomprising four syndromes: self-limi epilepsy with centrotemporal spikes, self-limitedepilepsy with autonomic seizures, childhood occipital visual epilepsy, and
- photosensitive occipital lobe epilepsy; (2) generalized epilepsies, comprising three syndromes:childhood absence epilepsy, epilepsy with myoclonic absence, and epilepsywith eyelid myoclonia; and (3) developmental and/or epileptic encephalopathies,comprising five syndromes: epilepsy with myoclonic–atoni

seizures, Lennox–Gastautsyndrome, developmental and/or epileptic encephalopathy with spike-and-waveactivationin sleep, hemiconvulsion–hemiplegia

- Epilepsy syndrome, and febrile infection-related epilepsy syndrome. We define each, highlighting the mandatory seizure(s),

EEG features, phenotypic variations, and findings from key investigations.

Diagnosis

- History and physical exam
- Work up depends on initial impression, genetic, metabolic, imaging (brain MRI) , CSF study ….etc
- EEG

Overview of selected lateralizing signs during seizures

Lateralizing sign	Frequency	Lateralizing value	Symptomatogenic zone
Aura			
Unilateral sensory aura [10]	6.1% epilepsy patients	89% contralateral	Brodmann areas 1, 2, and 3
Hemifield visual aura [26]	28.6% OLE[a]	100% contralateral	Brodmann areas 17–19 and adjacent areas
Motor			
Version [46,53]	22.2% FLE	100% contralateral	Brodmann areas 6 and 8
Clonic activity [53]	44.4% FLE	83% contralateral	Brodmann areas 4 and 6
Tonic activity [53]	48.1% FLE	89% contralateral	SMA, possibly also Brodmann area 6, the anterior cingulate gyrus, and subcortical structures
"Figure-of-4 sign" [60,64]	17.7% TLE; 15% ETLE	89% contralateral	SMA or prefrontal areas
Unilateral dystonic posturing [75,78]	43.9% TLE	100% contralateral; one exemption reported by Yen	Activation of basal ganglia
Automatisms and preserved consciousness [85,88]	5.7% TLE	100% non-dominant; one exemption reported by Janszky [88]	Unknown, possible impairment of consciousness with left or bilateral hippocampal impairment
Ictal spitting [97]	0.3% EMU patients	75% non-dominant	Possible asymmetry of the CAN
Ictal vomiting [102,105]	2% EMU patients	81% non-dominant	Medial, lateral superior and inferior structures of the nondominant temporal lobe and Papez circuit
Unilateral ictal eyeblinking [112]	1.5% EMU patients	83% ipsilateral	Unknown

Language			
Ictal speech [145]	34.2% EMU patients	83% non-dominant	Impairment of areas other than those involved in language production
Ictal dysphasia and aphasia [145]	34.2% EMU patients	100% dominant	Impairment of language areas
Postictal features			
Postictal palsy [157]	0.6% EMU patients	93% contralateral	Possible exhaustion or inhibition of Brodmann areas 4 and 6
Postictal nosewiping [170]	53.2% TLE	92% ipsilateral	Unknown

Demographic data
Epilepsy in Jordan Al-Qudah et al study 2017

		N (%)
Age groups	2-12 month	23 (3.5)
	>1 year- 6 years	239 (36.0)
	> 6 years-12 years	275 (41.5)
	> 12 years-18 years	126 (19.0)
Age at seizure onset	<1 month	27 (4.1)
	1-12 month	153 (23.0)
	>1-6 year	373 (56.3)
	>6-12 year	105 (15.8)
	>12-18 years	5 (0.8)
Gender	Male	377 (56.9)
	Female	286 (43.1)
Family History		225 (33.9)

Etiology of epilepsies 2017

	Frequency
A.Genetic	117 (17.7%)
Electro clinical syndromes (presumed genetic)	109
Others	8
B.Structural – metabolic	278 (41.9%)
Perinatal insults	89
Malformation of cortical development	26
Infection	23
Neurocutaneous syndromes	23
Trauma	16
Hydrocephalus	12
Metabolic disorders	9
Vascular Anomaly	7
Tumor	5
Stroke	3
Others	54
Corpus collosum	10
Brain atrophy	23
Microcephaly	8
Brain cysts	10
Delay Myelination	3
Distinctive constellations	11
Mesial temporal lobe epilepsy with hippocampal	10
Rasmussen syndrome	1.0
C.Unknown	268 (40.4%)
Electro clinical syndromes	82
Other types of epilepsy:	186
Focal temporal lobe	45
Focal frontal lobe	27
Focal parietal lobe	5
Focal occipital lobe	4
Focal (undetermined origin)	19
Generalized	70
Mixed seizure	16
Total	663 (100%)

type of zeizure

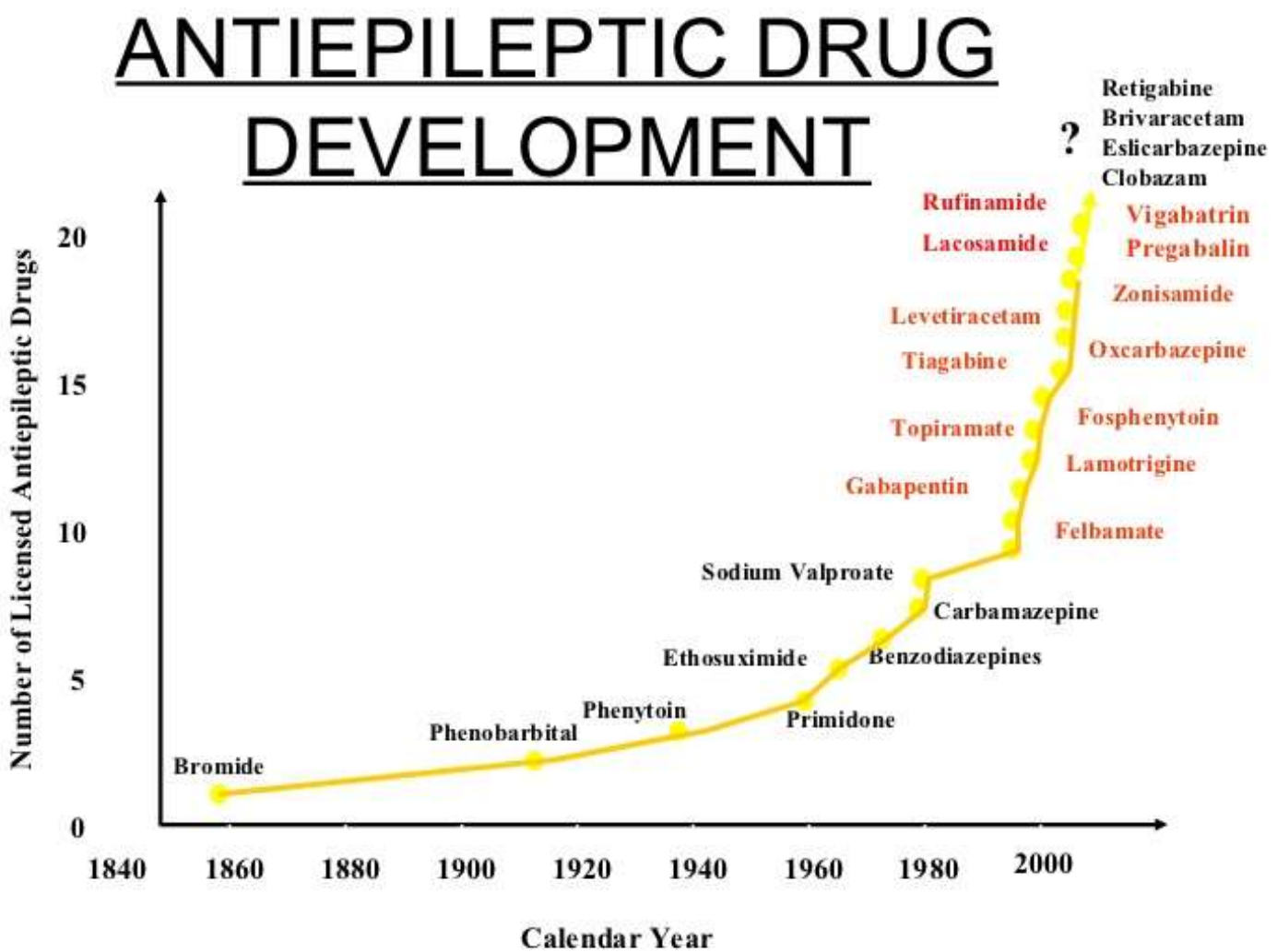
Number of * patients	One seizure type n=598 (90.2%)	Mixed seizures n=65 (9.8%)	Total number of patients=663
Number of seizures	One seizure type n=598 (81.1%)	Mixed seizures n=139 (18.9%)	Total number of seizures n=737
Generalized	248 (41.5)	103 (74.1)	351 (47.6)
• Tonic Clonic	139 (23.2)	29 (20.9)	168 (22.8)
• Absence	41 (6.9)	11 (7.9)	52 (7.1)
• Myoclonic	24 (4.0)	28 (20.1)	52 (7.1)
• Clonic	4 (0.7)	3 (2.2)	7 (0.9)
• Tonic	27 (4.5)	16 (11.5)	43 (5.8)
• Atonic	13 (2.2)	16 (11.5)	29 (3.9)
Focal	317 (53.0)	29 (20.9)	346 (46.9)
• without ICA*	59 (9.9)	9 (6.5)	68 (9.2)
• with ICA	178 (29.8)	18 (12.9)	196 (26.6)
• with ICA and evolving to BCS **	43 (7.2)	1 (0.7)	44 (6.0)
• without ICA* and evolving to BCS**	37 (6.2)	1 (0.7)	38 (5.2)
Spasms	33 (5.5)	7 (5.0)	40 (5.4)
Intractable			161/663 (24.3%)

TREATMENT

- Antiepileptic drug therapy
- Antiepileptic drugs new and traditional
  - Immune therapy (rasmussen ....)
  - Treatable metabolic disorders (pyridoxine, folonic acid .....)

- Epilepsy surgery
- Ketogenic diet
- Vagal nerve stimulation
- Others calosotomy,immune therapy,herbal...

Antiepileptic drug therapy



- *DRUGS THAT AFFECT VOLTAGE-DEPENDENT SODIUM CHANNELS*
  - Old : Carbamazepine Phenytoin
  - New : Lamotrigine Oxcarbazepine Zonisamide Lacosamide Rufinamide
- *DRUGS THAT AFFECT CALCIUM CURRENTS*
  - Ethosuximide
- *DRUGS THAT AFFECT GABA ACTIVITY*
  - *Benzodiazepines, barbiturates, gapabentine*
- *DRUGS THAT AFFECT GLUTAMATE RECEPTERS*
  - *topiramate*
- *DRUGS WITH MULTIPLE MECHANISMS OF ACTION*
  - *Topiramate, valproic acid*
- *Unknown*
  - Levetiracetam

Status epilepticus

- SE is a condition resulting either from the failure of the mechanisms responsible for seizure termination or from the initiation of mechanisms which lead to abnormally prolonged seizures (***after time point t_1***). It is a condition that can have long-term consequences (***after time point t_2***) , *including neuronal death, neuronal injury, and alteration of neuronal networks*, depending on the type and duration of seizures

Operational dimensions with t_1 indicating the time that emergency treatment of SE should be started and t_2 indicating the time at which long-term consequences may be expected

Type of SE	T 1	T 2
Tonic–clonic SE	5 min	30 min
Focal SE with impaired consciousness	10 min	>60 min
Absence status epilepticus	10–15 min	Unknown

ILAE.org 2017

Treatment of SE :

- Assessment (ABC)
- Supportive care
- Anticonvulsant :
  - 0 to 5 minutes : (Benzodiazepine : Diazepam. Medazolam …)
  - 5 to 10 minutes : (Benzodiazepine : second dose …)
  - 10 to 15 minutes ;(Fosphenytoin (second line), Phenobarbital)
  - 15 to 30 minutes : (phenobarbital, valproic acid, pyridoxine)
  - After 30 minutes : obtain anesthesiology consult

Febrile seizures

- " A seizure occurring in childhood after 1 month of age associated with a febrile illness not caused by an infection of the central nervous system (CNS), without previous neonatal seizures or a previous unprovoked seizure, and not meeting the criteria for other acute symptomatic seizures”

- Simple febrile seizures
- Complex febrile seizures

Neonatal Seizures

- Incidence is higher during this period than in any other period in life:
  - 57.5/1,000 in infants with birth weights <1,500 g
  - 2.8/1,000 in infants weighing between 2,500 and 3,999 g

There are 5 main neonatal seizure types :

1 . Subtle seizures:

include transient eye deviations, nystagmus, blinking, mouthing, abnormal extremity movements, fluctuations in heart rate, hypertension episodes, and apnea. Subtle seizures occur more commonly in premature than in full-term infants.

2. Clonic seizures can be focal or multifocal
3. Tonic seizures can be focal or generalized
4. Spasms are sudden generalized jerks lasting 1-2 sec
5. Myoclonic seizures are divided into focal, multifocal, and generalized

- Etiology

AGES 1-4 DAYS

- Hypoxic-ischemic encephalopathy
- Drug withdrawal, maternal drug use of narcotic or barbiturates
- Drug toxicity: lidocaine, penicillin
- Intraventricular hemorrhage
- Acute metabolic disorders
- Inborn errors of metabolism
- Pyridoxine deficiency (must be considered at any age)

AGES 4-14 DAYS

- Infection
- Metabolic disorders
- Drug withdrawal, maternal drug use of narcotics or barbiturates
- Benign neonatal convulsions, familial and nonfamilial
- Kernicterus, hyperbilirubinemia

AGES 2-8 WK

- Infection
- Head injury
- Inherited disorders of metabolism
- Malformations of cortical development
- Lissencephaly
- Tuberous sclerosis

Diagnosis :

- Taking the prenatal and postnatal history and performing an adequate physical examination.
- EEG is considered the main tool for diagnosis.
- Blood test : glucose, calcium, magnesium, electrolytes, and blood urea nitrogen
- Lumber puncture.
- Metabolic work up
- Imaging (brain MRI)

Prognosis:

- Prognosis of neonatal seizures has become better owing to improvement and advancement of obstetric care and intensive neonatal care.
- Mortality from neonatal seizures has decreased from 40 to 20%.
- EEG is highly associated with the outcome in premature and full-term infants. predictor of less-favorable later outcome
  - abnormal background.
  - prolonged electrographic seizures (>10 min/hour)
  - multifocal periodic electrographic discharges
  - spread of the electrographic seizures to the contralateral
- The underlying etiology of the seizures is the main determinant of outcome.

Treatment ：

- Treat underlying cause
- AED
  - ❑Phenobarbital
  - ❑Phenytoin and Fosphenytoin
  - ❑Lorazepam
  - ❑Diazepam, midazolam

Drug Resistant Epilepsy

- **Current practical definition:**
  - DRE is defined as failure of seizure control after adequate medical therapy with two or more appropriate anti-epileptic drugs.

- Epilepsy surgery can be indicated earlier when drug-resistance is highly expected such as in the mesial temporal lobe epilepsy with hippocampal sclerosis or when adverse effect of poor seizure control is expected on patient's development in young children.

1) K wan P, Arzimanoglou A, Berg AT , Brodie MJ, Allen Hauser W, Mathern G, Moshé SL, Perucca E, Wiebe S, French J: Definition of drug resistant epilepsy:consensus proposal by the ad hoc task force of the ILAE Commission on therapeutic strategies.Epilepsia 51: 1069–1077, 2010 2) K wan P, Schachter SC, Brodie MJ: Drug-resistant epilepsy. N Engl J Med 365: 919–926, 2011

Non-invasive pre-surgical evaluations for epilepsy

1. Detailed Clinical history and exam
2. Neuropsychological evaluation*
3. EEG and video EEG
4. MRI* Epileptongenic lesion
5. FDG-PET
6. Iomazenil-SPECT
   Ictal ECD-SPECT
7. Magnetoencephalography
   (MEG) and Functional MEG
8. Functional MRI

Masaki et al 2016

HEADACHES

INTRODUCTION

- Headache is the most common reason that children are referred to child neurology practices
- The prevalence of headache ranges from 37 to 51 percent in 7-year-olds, gradually rising to 57–82 percent by age 15
- The prevalence of migraine headache steadily increases through childhood and the male: female ratio shifts during adolescence.

- The prevalence of migraine rises from 3 percent at age 3–7 years to 4–11 percent by age 7–11, and up to 8–23 percent during adolescence.
- The mean age of onset of migraine is 7.2 years for boys and 10.9 years for girls
- New ICHD – 3 classification

ICHD – 3

Part ONE : Primary headaches

1. **Migraine**
   - 1.1 Migraine without aura
   - 1.2 Migraine with aura
   - 1.3 Chronic migraine
   - 1.4 Complications of migraine
   - 1.5 Probable migraine
   - 1.6 Episodic syndromes that may be associated with migraine

2. **Tension-type headache**
   - 2.1 Infrequent episodic tension-type headache
   - 2.2 Frequent episodic tension-type headache
   - 2.3 Chronic tension-type headache
   - 2.4 Probable tension-type headache

3. **Trigeminal autonomic cephalalgias**
   - 3.1 **Cluster** headache
   - 3.2 Paroxysmal hemicrania
   - 3.3 Short-lasting unilateral neuralgiform headache attacks
   - 3.4 Hemicrania continua
   - 3.5 Probable trigeminal autonomic cephalalgia

4. **Other primary headache disorders**
   - 4.1 Primary cough headache
   - 4.2 Primary exercise headache
   - 4.3 Primary headache associated with sexual activity
   - 4.4 Primary thunderclap headache
   - 4.5 Cold-stimulus headache
   - 4.6 External-pressure headache
   - 4.7 Primary stabbing headache
   - 4.8 Nummular headache
   - 4.9 Hypnic headache
   - 4.10 New daily persistent headache (NDPH)

Part two: the secondary headaches

5. Headache attributed to trauma or injury to the head and/or neck
6. Headache attributed to cranial or cervical vascular disorder
7. Headache attributed to non-vascular intracranial disorder
8. Headache attributed to a substance or its withdrawal
9. Headache attributed to infection
10. Headache attributed to disorder of homoeostasis
11. Headache or facial pain attributed to disorder of the cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cervical structure
12. Headache attributed to psychiatric disorder

Part three: painful cranial neuropathies, other facial pains and other headaches

13. Painful cranial neuropathies and other facial pains
14. Other headache disorders

Migraine :

- Recurrent headache disorder manifesting in attacks lasting 4-72 hours. Typical characteristics of the headache are unilateral location, pulsating quality, moderate or severe intensity, aggravation by routine physical activity and association with nausea and/or photophobia and phonophobia

- In children and adolescents (aged under 18 years), attacks may last 2-72 hours

Migraine

1.1 Migraine without aura

1.2 Migraine with aura

1.3 Chronic migraine

1.4 Complications of migraine

1.5 Probable migraine

1.6 Episodic syndromes that may be associated with migraine

1.2.1 Migraine with typical aura (visual, sensory, motor ....)

1.2.2 Migraine with brainstem aura (verigo, nausea, diplopia ....)

1.2.3 Hemiplegic migraine

1.2.4 Retinal migraine

Migraine

- 1.1 Migraine without aura
- 1.2 Migraine with aura
- 1.3 Chronic migraine
- 1.4 Complications of migraine
- 1.5 Probable migraine
- 1.6 Episodic syndromes that may be associated with migraine

→ Headache occurring on 15 or more days per month for more than three months, which, on at least 8 days per month, has the features of migraine headache

Migraine

- 1.1 Migraine without aura
- 1.2 Migraine with aura
- 1.3 Chronic migraine
- 1.4 Complications of migraine
  - 1.4.1 Status migrainosus (> 72 HR)
  - 1.4.2 Persistent aura without infarction
  - 1.4.3 Migrainous infarction
  - 1.4.4 Migraine aura-triggered seizure
- 1.5 Probable migraine
- 1.6 Episodic syndromes that may be associated with migraine

Management of migraine

- Medical treatment (abortive and preventive)
- Complementary and Alternative Treatments

The abortive (symptomatic) therapy

- Analgesics (**Acetaminophen, Ibuprofen …)**
- Triptans
- Ergotamine drugs

PREVENTIVE THERAPY :

- ANTIDEPRESSANT AGENTS (Amitriptyline .....)
- ANTIEPILEPTIC AGENTS (Topiramate, Valproic acid, Levetiracetam ....)
- ANTIHYPERTENSIVE AGENTS (Propranolol, Clonidine .....)
- CALCIUM CHANNEL BLOCKERS (Nimodipine ....)
- ANTIHISTAMINES (Cyproheptadine .....)

Tension-type headache

- Typically bilateral, pressing or tightening in quality and of mild to moderate intensity, lasting minutes to days.

- The pain does not worsen with routine physical activity and is not associated with nausea, but photophobia or phonophobia may be present.

2. Tension-type headache

- 2.1 Infrequent episodic tension-type headache → At least 10 episodes of headache occurring on <1 day per month
- 2.2 Frequent episodic tension-type headache
- 2.3 Chronic tension-type headache
- 2.4 Probable tension-type headache

2. Tension-type headache

2.1 Infrequent episodic tension-type headache

2.2 Frequent episodic tension-type headache → At least 10 episodes of headache occurring on 1-14 days per month

2.3 Chronic tension-type headache

2.4 Probable tension-type headache

2. Tension-type headache

- 2.1 Infrequent episodic tension-type headache
- 2.2 Frequent episodic tension-type headache
- 2.3 Chronic tension-type headache → ≥15 days per month
- 2.4 Probable tension-type headache

EPILEPSY IMITATORS

SYNCOPE AND ANOXIC SEIZURES

BEHAVIORAL, PSYCHOLOGICAL AND PSYCHIATRIC DISORDERS

SLEEP RELATED CONDITIONS

PAROXYSMAL MOVEMENT DISORDERS

MIGRAINE ASSOCIATED DISORDERs

MISCELLANEOUS EVENTS

SYNCOPE AND ANOXIC SEIZURES

- Vasovagal syncope
- Reflex anoxic seizures
- Breath-holding attacks
- Hyperventilation syncope
- Compulsive valsalva
- Neurological syncope
- Imposed upper airways obstruction
- Orthostatic intolerance
- Long QT and cardiac syncope
- Hyper-cyanotic spells

→

- ✓ affects all ages
- ✓ brief, lasting seconds
- ✓ Preceded by triggers
- ✓ Convulsive movements occur in 50 %
- ✓ Positive F. Hx is common

SYNCOPE AND ANOXIC SEIZURES

- Vasovagal syncope
- Reflex anoxic seizures
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- Hyperventilation syncope
- Compulsive valsalva
- Neurological syncope
- Imposed upper airways obstruction
- Orthostatic intolerance
- Long QT and cardiac syncope
- Hyper-cyanotic spells

- Occurs from early infancy onwards, either remit or evolve into vasovagal
- Preceded by sudden stimulus such as bump or knock, result in profound vagal stimulus, transient asystole
- Child become exceedingly pale and loss of consciousness, tonic posturing in possible

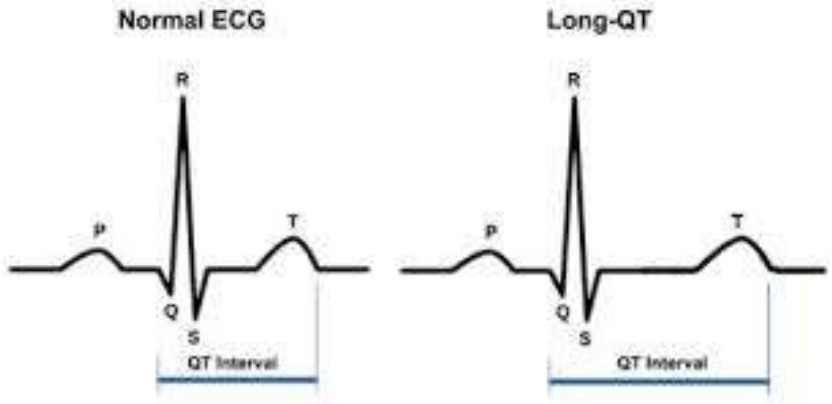
SYNCOPE AND ANOXIC SEIZURES

- Vasovagal syncope
- Reflex anoxic seizures
- Breath-holding attacks
- Hyperventilation syncope
- Compulsive valsalva
- Neurological syncope
- Imposed upper airways obstruction
- Orthostatic intolerance
- Long QT and cardiac syncope
- Hyper-cyanotic spells (in TOF)

- affect pre-school children
- Start with crying then stop breathing in expiration
- becomes blue with deep cyanosis
- They breath in or go to transient syncope, tonic posturing in possible
- more common if the child has iron deficiency anaemia

SYNCOPE AND ANOXIC SEIZURES

- Vasovagal syncope
- Reflex anoxic seizures
- Breath-holding attacks
- Hyperventilation syncope
- Compulsive valsalva
- Neurological syncope
- Imposed upper airways obstruction
- Orthostatic intolerance
- Long QT and cardiac syncope
- Hyper-cyanotic spells (in TOF)



- In long QT syndrome a ventricular tachyarrhythmia may be spontaneous or triggered by fright, exercise, surprise, and immersion in water.
- Syncope in sleep, a strong family history of syncope and a history of sudden death or drowning should raise suspicions of a cardiac syncope.
- Sensorineural deafness is associated with some types of long QT syndrome

BEHAVIORAL, PSYCHOLOGICAL AND PSYCHIATRIC DISORDERS

- Daydreaming /inattention
- Infantile gratification
- Eidetic imagery
- Tantrums and rage reactions
- Out of body experiences
- Panic attacks
- Dissociative states
- Non-epileptic seizures
- Hallucinations in psychiatric disorders
- Fabricated / factitious illness

- self-stimulation includes behavior which may be seen from infancy onwards, more so in pre-school girls.
- Rhythmic hip flexion and adduction may be accompanied by a distant expression, a flushed face and sometimes followed by sleepiness.

SLEEP RELATED CONDITIONS

- Sleep related rhythmic movement disorders
- Hypnogogic jerks
- Parasomnias
- REM sleep disorders
- Benign neonatal sleep myoclonus
- Periodic leg movements
- Narcolepsy-cataplexy

PAROXYSMAL MOVEMENT DISORDERS

- Tics
- Stereotypies
- Paroxysmal kinesigenic dyskinesia
- Paroxysmal nonkinesigenic dyskinesia
- Paroxysmal exercise induced dyskinesia
- Benign paroxysmal tonic upgaze
- Episodic ataxias
- Alternating hemiplegia
- Hyperekplexia
- Opsoclonus-myoclonus syndrome

- ✓ involuntary, sudden, rapid, repetitive, non-rhythmic, simple or complex movements or vocalizations
- ✓ common in childhood and have a tendency to wax and wane in frequency over time
- ✓ urge or compulsion to perform the tic, and an ability to suppress the tic (to some degree) are important features on history that support the diagnosis

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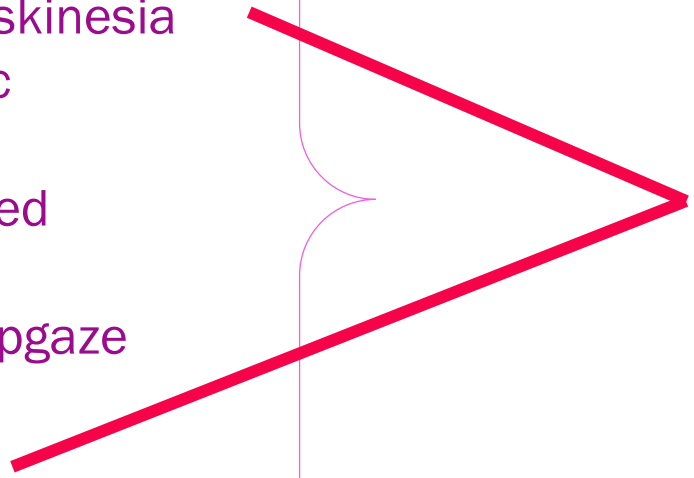

Genetically determined movement disorders

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- ✓ characterised by an exaggeration of the normal startle response and has several genetic linked to dysfunction of the inhibitory glycinergic pathway in the nervous system
- ✓ evident from the neonatal period or early infancy
- ✓ Die of apnea

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- ✓ autoimmune neurological disorder that may be seen in association with neuroblastoma, following viral infections .....
- ✓ The earliest feature is often ataxia followed by opsoclonus, followed by myoclonus

Episodic syndromes that may be associated with migraine

- Recurrent gastrointestinal disturbance
  - Cyclical vomiting syndrome
  - Abdominal migraine
- Benign paroxysmal vertigo
- Benign paroxysmal torticollis

- Recurrent episodic attacks of intense nausea and vomiting, usually stereotypical in the individual and with predictable timing of episodes.
- Attacks may be associated with pallor and lethargy.
- There is complete resolution of symptoms between attacks.
- Boys and girls are equally,
- The usual age of onset is 5 years
- Typically a self-limiting episodic condition occurring in childhood, children will “outgrow” these attacks by age 10

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- recurrent brief attacks of vertigo, occurring without warning and resolving spontaneously, after minutes to hours without loss of consciousness, in otherwise healthy children,
- Associated with at least one of the following: nystagmus, ataxia, vomiting, pallor, fearfulness

Episodic syndromes that may be associated with migraine

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- Recurrent episodes of head tilt to one side, perhaps with slight rotation, which remit spontaneously. The condition occurs in infants and small children, with onset in the first year.
- Associated with at least one of the following: pallor, irritability, malaise, vomiting, ataxia

MISCELLANEOUS EVENTS

- Benign myoclonus of infancy and shuddering attacks
- Jitteriness
- Sandifer syndrome
- Non-epileptic head drops
- Spasmus nutans
- Paroxysmal extreme pain disorder
- Spinal myoclonus

- This syndrome is seen in young children with gastro-oesophageal reflux (with or without vomiting).
- Events are often seen with or after feeding. Typically there is arching of the back, dystonic posturing of the limbs and turning/tilting of the head.

THANK YOU