Epilepsy Quiz:

Q.1 True about Aicardi Syndrome is (Dr Jean Dennis Aicardi 1926 – 2015)

A) Seen in female infants
B) No significant visual symptoms
C) Normal brain anatomically
D) Benign myoclonic jerks

Q.2 Which is seen in Panayiotopoulos syndrome?

A) Mainly tonic-clonic seizures
B) Onset age 10-20 years
C) Prolonged seizures of more than half an hour
D) Normal EEG

Q.3 What is characteristic of Landau kleffner syndrome?

A) Poor speech and hearing since birth.
B) Speech is normal throughout the course
C) Seizures are infrequent
D) Normal behaviour

Q.4 Sclerosis of which area of hippocampus occur in MTLE?

A). CA1
B). CA2
C). CA3
D). CA4

Q.5 Which drug causes Nephrolithiasis?

A) Oxcarbazepine
B) Lacosamide
C) Lamotrigine
D) Topiramte

Q.6 Which antiepileptic can produce parkinsonism

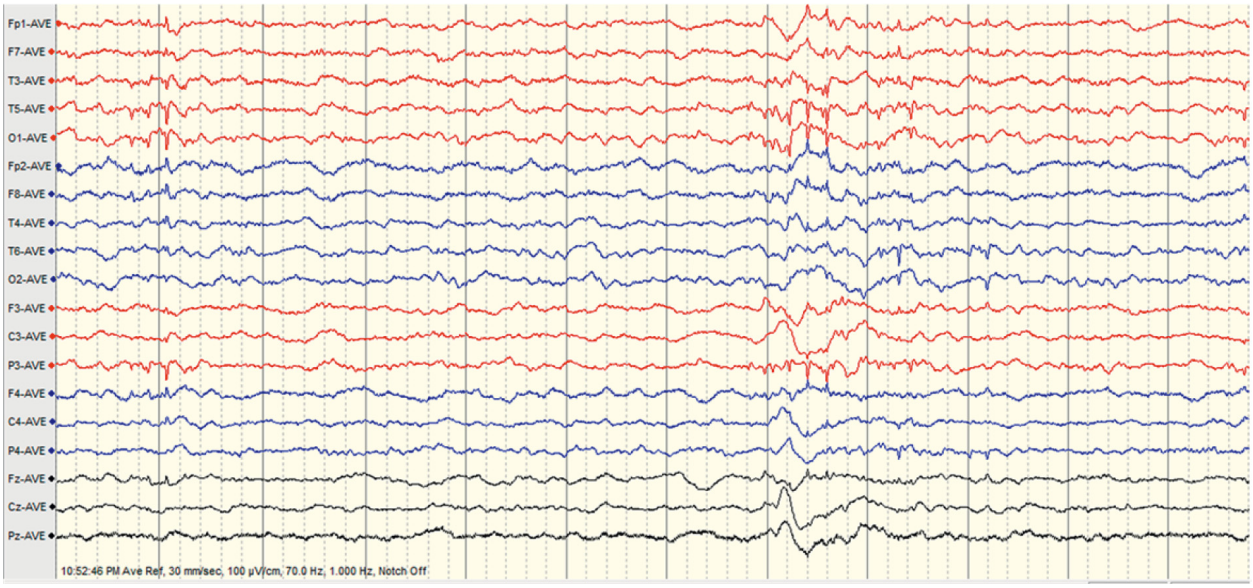
A) Phenytoin
B) Sodium Valproate
C) Levetiracetam
D) Oxcarbazapine

Q.7 Which antiepileptic can worsen seizures in benign rolandic epilepsy (BECTS)

A) Clobazam
B) Carbamezapine
C) Sodium valproate
D) Levetiracetam

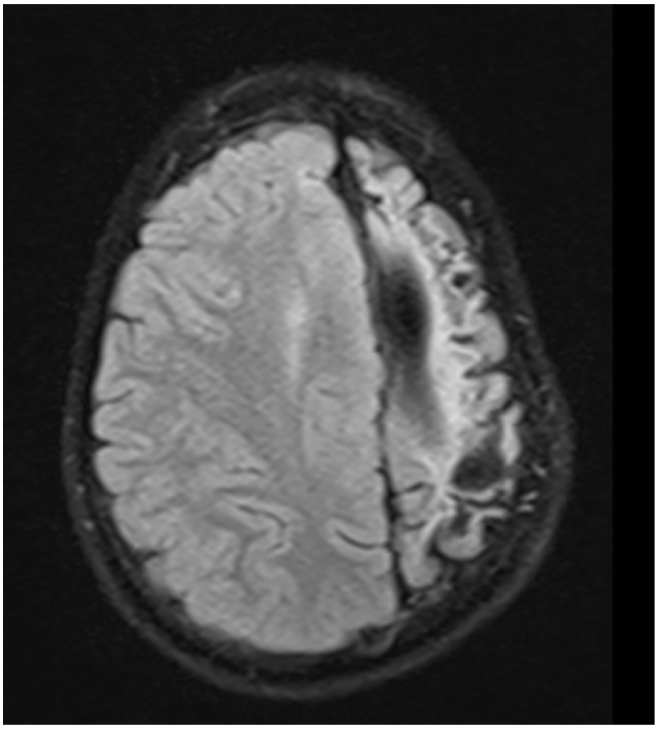
Q.8. What is the diagnosis in the current EEG

A) Left posterior spike and wave discharges
B) EKG artifacts
C) 6-Hz benign variants
D) None of the above



Q.9. A 35-year obese female is having migraine and primary generalised epilepsy. Which anti-epileptic you will prefer

A) Carbamezapine
B) Topiramate
C) Sodium Valproate
D) Lamotrigene



Q.10. This child is having recurrent daily seizures for last 2years with poor response to antiepileptics and right hemiaparesis. What is the best treatment option?

A) Multilobar resection
B) Focal resection of left frontal lobe
C) Vagus nerve stimulation
D) Hemispherotomy

Q.11. Hemiconvulsion-hemiparesis and epilepsy syndrome (HHE) is characterised by –

A) Progressive hemiparesis
B) Generalised seizures at onset
C) Permanent and static hemiparesis
D) Normal MRI brain

Epilepsy Quiz Answers:

1. Answer- A. **Seen in female infants.**

Explanation/Comment: Aicardi syndrome is thought to be an X-linked dominant disorder lethal to males. Except for 2 male children, all reported instances have been in females. Both males had XXY genotypes, which further supports an X-linked male lethal genetic substrate.

Aicardi syndrome is often complicated by severe mental retardation, intractable epilepsy-mainly infantile spasms, retinal abnormalities, absence of corpus callosum and a propensity to pulmonary complications. The condition often leads to death in the first decade.

References: Aicardi J, Lefebvre J, Lerique-Koechlin A. A new syndrome: spasm in flexion, callosal agenesis, ocular abnormalities. Electroencephalogr Clin Neurophysiol. 1965. 19:609-10

2. Answer -C. **Prolonged seizures of more than half an hour**

Explanation/Comment: Typically, the seizures are infrequent but long; 44% have seizures lasting 30 minutes or more, consisting of autonomic status epilepticus. One third of patients have a single seizure only. About half have 2-5 seizures. Only 5% have more than 10 seizures.

Panayiotopoulos syndrome is defined as “a benign age-related focal seizure disorder occurring in early and mid childhood. It is characterized by seizures, often prolonged, with predominantly autonomic symptoms, and by an EEG that shows shifting and/or multiple foci, often with occipital predominance.” In Panayiotopoulos syndrome, seizures make up an unusual constellation of autonomic, mainly emetic, symptoms, often with unilateral deviation of eyes and other, more conventional, symptoms. Seizures are nocturnal in about two thirds of patients. The full emetic triad (ie, nausea, retching, vomiting) culminates in vomiting in 74% of seizures.

References: Ferrie C, Caraballo R, Covanis A, Demirbilek V, Dervent A, Kivity S, et al. Panayiotopoulos syndrome: a consensus view. Dev Med Child Neurol. 2006 Mar. 48(3):236-40

3. Answer -C. **Seizures are infrequent**

Explanation/Comment: The prevalence of clinical seizures in Landau kleffner syndrome is 70-85%. In one third of patients, only a single seizure (or episode of status epilepticus) is recorded. In about one half of affected children, a seizure is the initial manifestation of acquired epileptic aphasia. In some, a few years may pass between the first seizure and the onset of any speech problems, whereas the opposite is true in others. Seizures usually appear between ages 4 and 10 years, and many series show that remission of the seizures before adulthood (often before age 15 y) is the rule. Clinical seizures are often easy to treat, but normalization of EEG discharges can be challenging.

References: Rossi PG, Parmeggiani A, Posar A, et al. Landau-Kleffner syndrome (LKS): long-term follow-up and links with electrical status epilepticus during sleep (ESES). Brain Dev. 1999 Mar. 21(2):90-8.

4. Answer -A. **CA1.**

Explanation/Comment: Hippocampal sclerosis (HS) describes a condition of severe neuron loss with gliosis, in the absence of cystic cavitation, usually in the CA1 sector and subiculum of the hippocampus. It is commonly recognized as medial temporal sclerosis in young individuals with temporal lobe epilepsy.

According to the International League Against Epilepsy (ILAE) Commission Report, HS is defined as neuronal loss and gliosis in hippocampal area CA1 (Sommer sector) and area CA4 (endplate/ hilus/end folium). Histologically, the segmental loss of pyramidal neurons in area CA1 is severe, with less prominent neuronal loss in areas CA3 and CA4.

References: Blumcke I, Thom M, Aronica E, Armstrong DD et al . International consensus classification of hippocampal sclerosis in temporal lobe epilepsy: a Task Force report from the ILAE Commission on Diagnostic Methods. Epilepsia 2013; 54: 1315–29

5. Answer -D. **Topiramte.**

Explanation/Comment: Albeit a rare occurrence, certain medications can induce urinary stone disease. Medication-induced calculi can be composed of the drug or one of its metabolites, and their formation may be promoted by the urinary supersaturation of these substances. Alternatively, the drug may induce physiologic changes that facilitate the formation of "metabolic stones'. Topiramate selectively inhibits cytosolic (type II) and membrane associated (type IV) forms of carbonic anhydrase. The action on carbonic anhydrase isoenzymes may contribute to the drug's side-effects, including its propensity to cause metabolic acidosis and calcium phosphate kidney stones. Preventive measures include high fluid intake, limited sodium intake, and consumption of citrate-containing fluids. Furthermore, patients receiving long-term topiramate therapy may require bone densitometry testing to detect early calcium loss secondary to acid buffering by bone.

References: Kuo RL, Moran ME, Kim DH, Abrahams HM, White MD, Lingeman JE. Topiramate-induced nephrolithiasis. J Endourol 2002; 16: 229–231

6. Answer B. **Sodium Valproate**

Explanation/Comment: The long-term use of valproic acid was found to induce parkinsonism in 5% of patients; while the exact underlying pathophysiology has not been clarified, one suggestion is that it is due to oxidative stress and mitochondrial dysfunction. VPA induced parkinsonism is not so rare and has been under-reported and under-recognized. When we are confronted with the patients who develop parkinsonism after VPA administration, the possibility of VPA induced parkinsonism should be considered in the differential diagnosis. Old age, long duration of treatment, and VPA dose maintaining serum therapeutic levels might be predisposing factors for this syndrome.

References: Jamora D, Lim SH, Pan A, Tan L, Tan EK. Valproate-induced Parkinsonism in epilepsy patients. Mov Disord. 2007;22:130–133

7. Answer-B.. **Carbamezapine**

Explanation/Comment: In patients with BECTS, CBZ has been reported to cause aggravation of epilepsy in some well-documented cases, with increase in seizure frequency, appearance of atypical absences with myoclonic and atonic components, speech disturbances and decrease in school performance, appearance on the EEG of diffuse slow spike–slow wave discharges, and continuous diffuse spike–wave discharges during slow sleep. here is also evidence that carbamazepine can worsen speech production

References: Lerman P. Seizures induced or aggravated by anticonvulsants. Epilepsia 1986;27:706–10

Park JI, Kim SJ, Kim HG. Acoustic effects of carbamazepine in benign rolandic epilepsy. Epilepsy Behav 2005;7:468–71.

8. Answer C. **6Hz Benign variants.**

Explanation/Comment: Sharp positive component, rounded negative component,predominantly in light sleep,runs usually less than 2 seconds,posterior temporal, unilaterally or bilaterally,medium amplitude: 20-60 uV. seen predominantly in adolescents, especially age 13-14 years, although it can be seen at any age. 14-and-6 Hz positive bursts were first described by Gibbs and Gibbs (1951). In normal subjects the bursts are typically a mixture of 14-and-6 Hz surface positive wave forms lasting from 500 to 1000 msec, occurring in drowsiness and sleep of children and young adults, with a maximum incidence at 13-15 years and an amplitude emphasis in the posterior temporal region.

References: Gibbs, E.L. and Gibbs, F.A. Electroencephalographic evidence of thalamic and hypothalamic epilepsy. Neurology, 1951, 1:136-144.Klass, D.W. and Westmoreland, B.F. Nonepileptogenic epileptiform electroencephalographic activity. Ann. Neurol., 1985, 18: 627-635.

9. Answer- B. **Topiramte**

Explanation/Comment: Topiramate and sodium valproate are known to have good response in migraine and well as primary generalised epilepsy. However in view of possible side effect of weight gain, sodium valproate will not be a good choice in this patient. Topiramate will be preferred in view of its known side effect of weight loss. There are various reports of increased prevalence of migraine in patients with juvenile myoclonic epilepsy. The high prevalence of MA in JME might be related to theory of cortical spreading depression (CSD) starting more often in patients with JME than without JME.

References: Schankin CJ, Remi J, Klaus I, Sostak P, Reinisch VM, Noachtar S, Straube A (2011) Headache in juvenile myoclonic epilepsy. J Headache Pain 12(2):227–233

10. Answer-D **Hemispherotomy**

Explanation: Hemispherotomy is the most effective epilepsy surgery in a patient with severe hemispheric atrophy. MRI in this patient is showing severe left hemispheric atrophy. As the patient is having already established hemiparesis, doing hemispherotomy is not going to worsen any major deficits and the expected results are excellent in terms of seizure control.

11. Answer-C **Permanent and static hemiparesis**

Explanation/Comments: Hemiconvulsion-hemiplegia syndrome' is a rare dramatic sequence comprising a sudden and prolonged unilateral clonic seizure in a febrile child of less than 4years of age, followed by permanent ipsilateral hemiplegia. Subsequent to hemiconvulsion and hemiplegia, nearly 80% of patients develop focal epilepsy of the complete hemiconvulsion-hemiplegia-epilepsy syndrome. MRI Brain will show extensive atrophy of the involved hemisphere.

References: Freeman JL, Coleman LT, Smith LJ, Shield LK. Hemiconvulsion-hemiplegia-epilepsy syndrome: characteristic early magnetic resonance imaging findings. J Child Neurol. 2002;17:10–6

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