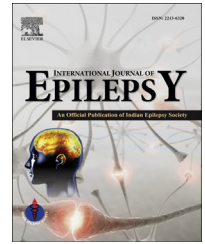


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## Quiz

# MCQs: International Journal of Epilepsy

## Questions

- Which of the following mutations has not been associated with generalized epilepsy with febrile seizures plus (GEFS+)?
  - SCN1D
  - SCN1A
  - SCN1B
  - SCN 2A
  - GABGR2
- Which of the following statements about epilepsy surgery is false?
  - Multiple subpial transections are performed for epileptogenic lesions in the eloquent cortex
  - Corpus callosotomy usually splits the anterior one third of the corpus callosum
  - Anterior temporal lobectomy results in seizure freedom in 70% of mesial temporal epilepsy
  - Vagal nerve stimulation is always done on the right vagus nerve
- Neonatal seizures include the following types except
  - Tonic
  - Clonic
  - Tonic clonic
  - Myoclonic
  - Subtle seizures
- Which of the following statements best describes the role of Electroencephalography (EEG) in a typical febrile seizure?
  - Predicts the recurrence risk of febrile seizure
  - Useful in both simple and complex types
  - A set point to diagnose febrile convulsion
  - Predicts the future risk of epilepsy
  - Not recommended in febrile seizures
- Which of the following statements about hypothalamic hamartomas is false?
  - They are associated with gelastic seizures
  - These tumors strongly enhance on contrast imaging
  - Sessile or intrahypothalamic hamartomas are the ones most strongly associated with epilepsy

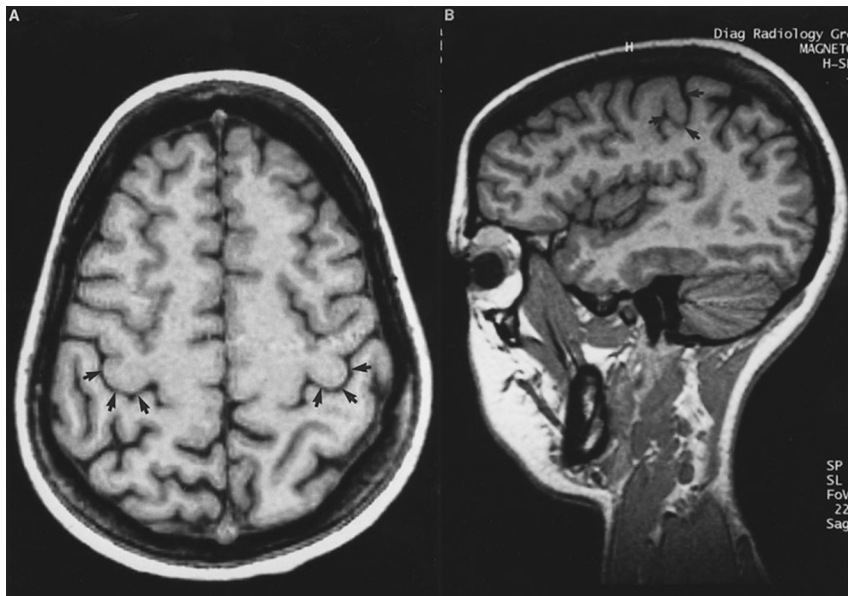


- They are associated with gelastic seizures
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- d. Routine scalp video EEG monitoring of seizures may be misleading
  - e. Patients with medically refractory seizures may need surgical resection or radiosurgery
6. Regarding NMDA receptor antibody encephalitis, which of the statements is incorrect?
- a. It typically affects girls and young women
  - b. Adult women may have associated teratomas and such patients have better outcome
  - c. EEG shows frequent epileptiform discharges
  - d. NMDA-R antibodies are found in the serum and CSF
  - e. Dyskinesias, Seizures and dysautonomia are common
7. Which of the statements about High frequency oscillations is incorrect?
- a. Higher frequency Oscillations or HFOs are cerebral potentials between 80 and 500 Hz
  - b. HFOs are best recorded using scalp electrodes
  - c. Fast HFOs between 250 and 500 Hz (also known as fast ripples) are specifically associated with epileptogenic cortex
  - d. Complete surgical resection of these HFOs is more likely to result in a seizure free outcome after epilepsy surgery
8. The EEG pattern in the picture represents.



- a. An epileptic ictal pattern
  - b. Rhythmic temporal theta of drowsiness
  - c. Wicket rhythm
  - d. Subclinical rhythmic EEG discharge of adults
  - e. An electrical artifact
9. The region marked by arrows on the axial T1 W image of the brain corresponds to.



- a. The face motor area
  - b. The Broca's area
  - c. The leg sensory area
  - d. The leg motor area
  - e. The hand motor area
10. Antiepileptic drug exposure especially to valproate in the mother is documented to affect cognitive outcomes in the child. The most crucial period for this effect is during
- a. First trimester of pregnancy
  - b. Last trimester of pregnancy
  - c. Throughout pregnancy
  - d. During lactation
  - e. Pregnancy and lactation
11. In seizure semiology, which of the following is a strongly lateralizing sign?
- a. Unilateral hand automatisms
  - b. Early ill sustained head version
  - c. Post ictal aphasia
  - d. None of the above

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## Answer key

1. Answer – e

**Explanation:** GEFS plus is a syndromic autosomal dominant disorder where afflicted individuals can exhibit numerous epilepsy phenotypes. GEFS+ can persist beyond early childhood (i.e., 6 years of age). GEFS+ is also now believed to encompass three other epilepsy disorders: severe myoclonic epilepsy of infancy (SMEI), which is also known as Dravet syndrome, borderline SMEI (SMEB), and intractable epilepsy of childhood (IEC). There are at least six types of GEFS+, delineated by their causative gene. Known causative genes are the sodium channel  $\alpha$  subunit genes SCN1A, SCN2A, an associated  $\beta$  subunit SCN1B, and a GABAA receptor  $\gamma$  subunit gene GABGR2.

**Ref:** M. Ito, K. Yamakawa, T. Sugawara, S. Hirose, G. Fukuma, S. Kaneko. Phenotypes and genotypes in epilepsy with febrile seizures plus. *Epilepsy Research* 2006 August; 70:199–205.

2. Answer – b

**Explanation:** Corpus callosotomy is offered as a palliative procedure in patients with refractory secondary generalized seizures, especially with frequent falls. The procedure involves the splitting of the anterior two-thirds or the entire corpus callosum. In 70–80% control of generalized seizures is good.

**Ref:** Spencer DS & Spencer SS. Corpus callosotomy in the treatment of medically intractable secondarily generalized seizures of children. *Cleveland Clinic Journal of Medicine* 1989; 56(Suppl. Pt 1): 69–78.

3. Answer – c

**Explanation:** Subtle, focal clonic and myoclonic are the most common seizure types in the neonate. Generalized tonic clonic seizures are exceptional. This is probably due to the anatomical and physiological immaturity of the neonatal nervous system, such as the incomplete myelination pattern of neonates which tends to prevent highly-organized, synchronized, generalized seizure activation.

**Ref:** Volpe, JJ. Neonatal seizures. In: *Neurology of the newborn*. 5th edition. WB Saunders Elsevier, Philadelphia, PA; 2008: 203–244.

4. Answer – e

**Explanation:** The yield of routine EEG is low in neurologically normal children with febrile seizures, even if the seizure is complex. Abnormal posterior slowing may occur shortly after the seizure and may be detected for as long as 10 days afterwards. This finding can serve to confirm the clinical impression that a seizure has occurred. However, EEG abnormalities are not predictive of recurrence or development of future epilepsy. This led to the conclusion that the routine practice of obtaining EEG in neurologically normal children with febrile seizures is not justified.

**Ref:** Jan M.M. Assessment of the utility of pediatric electroencephalography. *Seizure* 2002; 11: 99–103.

5. Answer – b

**Explanation:** Sessile hypothalamic hamartomas are more frequently associated with epilepsy, and are usually non enhancing lesions within the tuber cinereum. Pedunculated hamartomas are commonly present with isosexual precocious puberty.

While there is evidence of the hamartoma being primarily epileptogenic, but secondary epileptogenesis from surrounding neocortex is a well established observation. Scalp video EEG recordings are therefore often not useful to localize seizure onset, and may at times be misleading.

**Ref:** Pati S, Sollman M, Fife T.D., Ng Y.T. Diagnosis and management of epilepsy associated with hypothalamic hamartoma: an evidence based systematic review. *J Child Neurol* 2013 July; 28(7):909–916.

6. Answer – c

**Explanation:** NMDA Receptor encephalitis is an autoimmune disorder predominantly affects children and young adults, especially girls. It may occur in association with a systemic tumor. The most commonly associated tumor in women is an ovarian teratoma. 77% of patients in a series of 100 patients had generalized or predominantly frontotemporal slow or disorganized activity (delta theta) without epileptic discharges. MRI does not reveal any abnormality in over 50% of patients. Patients with a systemic tumor do better with treatment (with resection and immunotherapy) than do patients without a tumor (immunotherapy alone).

**Ref:** Dalmau J, Lancaster E, Martinez-Hernandez E, Rosenfeld M.R., Balice Gord R. Clinical experience and laboratory investigations in patients with anti-NMDAR encephalitis. *Lancet Neurol* 2011; 10:63–74.

7. Answer – b

**Explanation:** High-frequency oscillations (HFOs) between 80 and 500 Hz which can be recorded with EEG. While mostly recorded on intracranial EEGs, new studies suggest that identification of HFOs on scalp EEG or magnetoencephalography (MEG) is also possible. HFOs may be novel markers of the epileptogenic zone. HFOs are clearly linked to the seizure onset zone. The surgical removal of regions generating those correlates with a seizure free post-surgical outcome, and HFOs become more frequent after reduction of antiepileptic drugs.

**Ref:** Jacobs J, Staba R, Asano E. et al. High-frequency oscillations (HFOs) in clinical epilepsy. *Prog Neurobiol* 2012 Sept; 98 (3): 302–315.

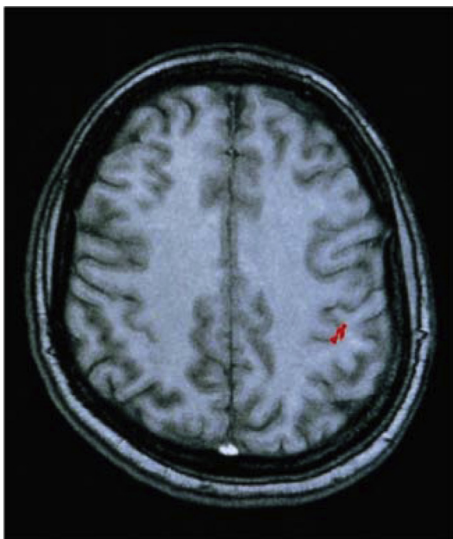
8. Answer – d

**Explanation:** SREDA is a burst of rhythmic predominantly theta activity with a bisynchronous, widespread distribution, but often more prominent over the parietal and temporal head region. It can last several minutes and is a “non evolving pattern”. It is usually seen in older adults, and may occur in both, sleep and wakefulness. It is a benign variant and its non epileptic nature and can be confirmed by clinical testing during the discharge.

**Ref:** Westmoreland B.F., Klass D.W. Unusual variants of subclinical rhythmic electrographic discharge of adults (SREDA). *Electroencephalogr Clin Neurophysiol* 1997 Jan; 102(1):1–4.

9. Answer – e

**Explanation:** Axially (A) the precentral knob is omega-shaped in the left and epsilon-shaped in the right hemisphere and correspond to the hand motor area. The knobs are posterior to the intersection of the superior frontal sulcus with the precentral sulcus. Sagittally, (B) the posteriorly directed hook is identified at the level of the posterior part of the insula. In the image below, fMRI in a healthy volunteer is showing an intraparenchymal area of activation (red) in the pre-(knob) and postcentral gyrus.



Ref: T.A. Yousry, U.D. Schmidt, H. Alkadhi, D. Schmidt, A. Peraud, A. Buettner, P. Winkler. Localization of the motor hand area to a knob on the precentral gyrus: A new landmark. *Brain*.1997 Jan; 120 (Pt 1), 141–157.

10. Answer – c

**Explanation:** Although the crucial period for all structural abnormalities caused by drug exposure or other reasons is limited to the first trimester, drug exposure throughout pregnancy might affect the cognitive development of the fetus and the child. After controlling for maternal IQ, patients with valproate exposure had average IQ scores lower than after exposure to lamotrigine, carbamazepine, or phenytoin.

Ref: Meador K.J., Baker G.A., Browning N, et al. Cognitive function at 3 years of age after fetal exposure to antiepileptic drugs. *N Engl J Med* 2009; 360:1597–605.

11. Answer – c

**Explanation:** Postictal aphasia, virtually always localized to the dominant temporal lobe. Non-verse head turning is an unforced head movement that is seen in both frontal and temporal epilepsies and may be ipsilateral, but it does not have the strong localizing value of the sustained late verse head movement. Unilateral hand automatisms are usually a manifestation of contralateral limb dystonia that prevents bimanual automatisms, and therefore may not be, in itself, strongly lateralizing.

Ref: Krikor Tufenkjian, Hans O. Lüders. Seizure semiology: its value and limitations in localizing the epileptogenic zone. *J Clin Neurol* 2012; 8:243–250.

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