Juvenile Absence Epilepsy (JAE)

Juvenile Absence Epilepsy (JAE) is one of the commonest forms of epilepsy in adolescence.

The seizures can typically include absences, infrequent generalised tonic clonic seizures (GTCS) and myoclonic seizures. Absence seizures are the predominant seizure type.

The EEG has generalized spike or polyspike slow-wave complexes.

JAE is a lifelong disorder, although seizures can be controlled in 70% to 80% of patients.

Treatment is with sodium valproate, lamotrigine, clobazam, or ethosuximide. Ethosuximide does not control GTCS.

DEMOGRAPHICS

Prevalence: ~2% to 3% of all epilepsies – more common in young adults.
Age at Onset: Peak at 9 to 13 years (70% of the patients).
Genetics: Genetically determined.

SYMPTOMS AND SIGNS

Seizure Semiology: JAE manifests with severe typical absence seizures; many patients (~80%) also suffer from infrequent GTCS and ~1/5 have sporadic myoclonic jerks.

- Absence seizures are severe. The absences occur less frequently than in Childhood Absence Epilepsy (approximately ≤ 5 per day). Duration varies from 4 to 30 sec (~16 sec).
- GTCS can occur, usually infrequently, and are usually controlled with medication.
- Myoclonic jerks are infrequent, mild and of random distribution.

Neurological and Mental State: Normal

Seizure-Precipitating Factors

- Typical absence seizures can be provoked by hyperventilation in the untreated or poorly controlled patient.
- Sleep deprivation is the major precipitant of GTCS.

INVESTIGATIONS

EEG:

- Normal background. Generalized 3-4 Hz spike or polyspike slow-wave complexes.

Neuro Imaging:

- Not usually required.
DIFFERENTIAL DIAGNOSIS

- Mainly from other IGEs such as CAE or Juvenile Myoclonic Epilepsy (JME).
- At onset, both JAE and JME may present with GTCS. JME has prominent early morning myoclonic seizures (may occur at other times) in addition to GTCS and absence seizures.
- CAE generally occurs in a younger child but age is not an absolute discriminant. In CAE, there are many daily absences.

PROGNOSIS

- JAE is thought to be a lifelong disorder, although seizures can be very well controlled in ~70% to 80% of patients.

MANAGEMENT OPTIONS

- Sodium valproate is often very effective. However, for women of child bearing age there are concerns of teratogenicity. Other alternatives are lamotrigine, clobazam or ethosuximide in combination with one of the former drugs (ethosuximide does not control GTCS).
- Sodium valproate has been associated with significant concerns of teratogenicity (i.e. malformations, cognitive impairment, and Autistic Spectrum Disorder). This is particularly true at higher dosages. The risk of teratogenicity increases with increasing dosage. It is important clinicians and women of child bearing age are aware of this risk. Ideally, pregnancies in women with epilepsy should be planned and managed by a neurologist. Medication choices should be selected and discussed keeping in mind the safety of mother and foetus.
- Patients should be warned with regard to precipitating factors of GTCS.
- Avoidance of sleep deprivation, fatigue, and alcohol.

DISCUSSION WITH FAMILY

- Safety
- Epilepsy Medical Record
- Potential of performing baseline educational assessment (through school counsellor)
- Drug Handout
- Driving resources

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REFERENCES:
