IGE with phantom absences

- Prognosis:
  - Most of the patients were seizure free with treatment
  - Some patients refused to take medication

Panayiotopoulos CP, et al. JNNP 1997;63:622

Absence status epilepsy

- 11 cases
- Main type of seizure is recurrent, unprovoked absence status
- Rare typical absences
- Infrequent GTC seizures
- Onset of absence status is usually after puberty or early adult
- No family history


Absence status epilepsy

- All patients were given wrong diagnosis of complex partial seizures with secondarily GTC or just “grandmal seizure”
- Prognosis is quite good with adequate treatment (VPA, LTG)


Late onset absence status

- Absence status in the elderly
- Presence of absence status epilepticus at the age of 60-80
- Most occurs “de novo”
- A portion of patients had prior history of IGE when they were young in some series
- Almost half of patients had precipitating factors
  eg. Benzodiazepine withdrawal

Panayiotopoulos CP, et al. JNNP 1997;63:622
Late onset absence status

• Thomas P, et al.
• Reported 11 cases of “de novo” absence status at the age onset >40 years
• Mean age of onset 58.9 yrs (48-81 yrs)
• One pt had one prior GTC seizure due to benzodiazepine withdrawal


Late onset absence status

• 9 pts received psychotropic drugs include benzodiazepines and in 8 of 9 pts benzodiazepine was discontinued 2-3 days prior to AS
• AS were stopped with benzodiazepine or phenobarbital in all cases


Review of 64 cases in the literature

Middle age or elderly pts
• No prior history of epilepsy
• Female predominance
• 72% had precipitating factors


Generalized epilepsy with febrile seizure plus (GEFS+)

• Phenotypic spectrum are extremely variable
• Multiple febrile seizures in early childhood and persists beyond 6 years old
• History of febrile seizures in the family

**Generalized epilepsy with febrile seizure plus (GEFS+)**

- Generalized seizure or focal seizures
- Seizures remitted in early teenage although in some cases can persist until late adolescent or older
- Seizure types
  - GTCs, absence, myoclonic, atonic, tonic seizures
  - Unilateral clonic seizures, visual, psychic aura

**Is IGE in adult different from childhood IGE?**

- Nicolson, et al
- Study clinical manifestation of 72 cases late onset IGE compared to 844 cases of early onset IGE

### Table 1: Prognosis in adult onset IGE

<table>
<thead>
<tr>
<th>Variable</th>
<th>Age of onset &lt; 20 years (n = 772)</th>
<th>Age of onset &gt; 20 years (n = 72)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Seizure type</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Absence</td>
<td>338 (44.4)</td>
<td>11 (15.3)</td>
</tr>
<tr>
<td>Myoclonic</td>
<td>363 (47.0)</td>
<td>30 (41.7)</td>
</tr>
<tr>
<td>Tonic-clonic</td>
<td>716 (93.0)</td>
<td>69 (95.8)</td>
</tr>
<tr>
<td>Epilepsy type</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Absence epilepsy</td>
<td>216 (28.0)</td>
<td>7 (9.7)</td>
</tr>
<tr>
<td>Myoclonic epilepsy</td>
<td>363 (47.0)</td>
<td>30 (41.7)</td>
</tr>
<tr>
<td>TC seizures only</td>
<td>193 (25.0)</td>
<td>35 (48.6)</td>
</tr>
<tr>
<td>Remission rate</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Absence epilepsy</td>
<td>112 (54.1)</td>
<td>36 (55.6)</td>
</tr>
<tr>
<td>Myoclonic epilepsy</td>
<td>173 (49.4)</td>
<td>15 (53.6)</td>
</tr>
<tr>
<td>TC seizures only</td>
<td>124 (70.5)</td>
<td>18 (62.1)</td>
</tr>
<tr>
<td>Overall</td>
<td>409 (55.8)</td>
<td>68 (56.7)</td>
</tr>
</tbody>
</table>

Nicolson A, et al JNNP 2004

**Generalized epilepsy with febrile seizure plus (GEFS+)**

- Genetics
  - AD with incomplete penetrance
  - Different mutations
    - SCN1B gene (19q13.1) µ1 subunit of sodium channel
    - SCN1A gene (20q21-33) α1 subunit of sodium channel
    - GABRG2 gene (5q31-33) θ2 subunit at the benzodiazepine binding site of GABA_A receptor

**Prognosis in adult onset IGE**

<table>
<thead>
<tr>
<th>Syndrome</th>
<th>Remission (%)</th>
<th>Relapse (%)</th>
<th>Uncorrected (%)</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Juvenile myoclonic epilepsy</td>
<td>40 (73)</td>
<td>4 (7)</td>
<td>11 (20)</td>
<td>55</td>
</tr>
<tr>
<td>Epilepsy with generalized tonic-clonic seizures</td>
<td>15 (54)</td>
<td>5 (16)</td>
<td>8 (28)</td>
<td>26</td>
</tr>
<tr>
<td>Epilepsy with generalized tonic-clonic seizures or awakening</td>
<td>5 (71)</td>
<td>2 (28)</td>
<td>2 (28)</td>
<td>7</td>
</tr>
<tr>
<td>Juvenile absence epilepsy</td>
<td>3 (75)</td>
<td>1 (25)</td>
<td>0</td>
<td>4</td>
</tr>
<tr>
<td>Late-onset myoclonic epilepsy</td>
<td>3 (33)</td>
<td>1 (33)</td>
<td>2 (33)</td>
<td>3</td>
</tr>
<tr>
<td>Epilepsy with generalized tonic-clonic seizures during sleep</td>
<td>2 (67)</td>
<td>1 (33)</td>
<td>0</td>
<td>3</td>
</tr>
<tr>
<td>Childhood absence epilepsy</td>
<td>1 (50)</td>
<td>1 (50)</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td>Epilepsy with myoclonic absences</td>
<td>1 (100)</td>
<td>0</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Total</td>
<td>66 (84)</td>
<td>10 (13)</td>
<td>27 (36)</td>
<td>103</td>
</tr>
</tbody>
</table>

Implication to our practice

• IGE in adult does exist although uncommon
• Clinical syndromes of IGE in adult are still not well defined
• Through history of seizure characteristics and EEG (sleep deprived, hyperventilation and photic stimulation) may be helpful for diagnosis of IGE in this age group
• Beware of frontal lobe epilepsy that may mimic IGE

Implication to our practice

• Importance of diagnosis of IGE in adult
  – Select appropriate medication
  – Reduced cost of neuroimaging
  – Importance of avoiding precipitants