

**Primary generalized epilepsy:  
clinical variability from childhood  
to adult**

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**Outline**

- IGE in adult: Is this common?
- Clinical spectrum of IGE in adult
- Is IGE in adult different from childhood IGE?
- Clinical implications

**IGE in adult: Is it common?**

- Idiopathic generalized epilepsy is common and accounts for 20-40% of all epilepsies
- Before 1989 classification, Gastaut reported that in his series seizures began after adolescent in 35% of IGE patients

**IGE in adult: Is it common?**

- Loiseau J, et al 1998
- Review in literatures and 4 large epilepsy database in France for IGE of very late onset (>60 yr old)
- Found 5 case reports in literature
  - All female
  - 4 presented with GTCs and one with myoclonic seizure
- Found no pts in the databases with very late onset IGE

Loiseau J, et al. Seizure 1998;7:485

**IGE in adult: Is it common?**

	Cutting 2001	Marini 2003	Nicolson 2004
No. of pts with IGE	313	121	844
Adult onset IGE	42 (13%)	34 (28%)	72 (8.5%)
Mean age of onset	23.8	33	23.8

Nicolson A, et al JNNP 2004;75:72  
Marini C, et al JNNP 2003;74:192

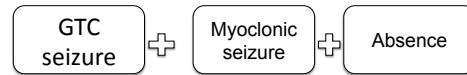
**Classic syndromes of IGE that can be  
found in adult**

- Childhood absence epilepsy
- Juvenile absence epilepsy
- Juvenile myoclonic epilepsy
- Epilepsy with grand mal seizures (generalized tonic-clonic seizures) on awakening

### Clinical spectrum of IGE in adult

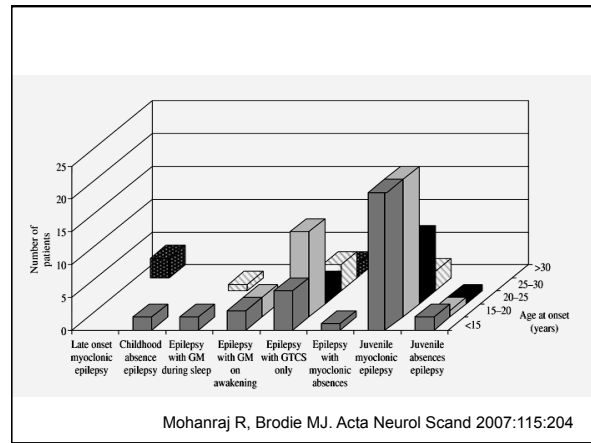
- GTC seizure alone
- GTC seizure +/- myoclonic seizure +/- absence seizure
- Absence status
- Absence status in the elderly

### Clinical spectrum of IGE in adult



	Cutting 2001	Marini 2003	Nicolson 2004
Adult onset IGE	42 (13%)	34 (28%)	72 (8.5%)
Mean age of onset	23.8	33	23.8
Seizure types			
GTC seizure	41 (97%)	32 (94.1%)	69 (95.8%)
Myoclonic	21 (50%)	6 (17.6%)	30 (41.7%)
Absence	16? (38%)	3 (8.8%)	11 (15.3%)
Epilepsy type			
AME	21 (50%)	6 (17.6%)	30 (41.7%)
GTS-A/ GTC	3 (7%)	25 (73.5%)	35 (48.6%)
Absence E	-	3 (8.8%)	7 (9.7%)
Unclassified	11 (43%)	-	-

Nicolson A. et al JNNP 2004. Marini C. et al JNNP 2003



### Generalized tonic clonic seizure



### Epilepsy with GTC seizures only

- Appeared in recent proposed classification scheme in 2001
- This appeared to include the classically described "epilepsy with GTC seizures on awakening"
- Peak age of onset at the end of second decade (range 5-50 year)

Engel J Jr. Epilepsia 2001;42:796

### Epilepsy with GTC seizures only

- Seizure types: GTC seizures, few also have myoclonic and absence seizures
- Seizures are mainly provoked by alcohol or sleep deprivation
- In GTC seizures on awakening, 90% of all seizures occur within 1-2 hrs after awakening or at the end of the day

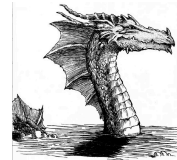
### Epilepsy with GTC seizures only

- EEG
  - Interictal: generalized 4-5 Hz swc, generalized polyspikes
- Genetics
  - Frequently have positive family history
  - Different mutations of chloride channel 2 gene (CLCN2) on chromosome 3q26 are associated with GTC on awakening

### Epilepsy with GTC seizures only

- Treatment
  - Valproate, lamotrigine
  - Topiramate, levetiracetam, zonisamide
- Prognosis
  - Good chances of seizure control and 2-5 yrs seizure freedom: 70-90%

### Myoclonic seizure



### Adult onset myoclonic epilepsy

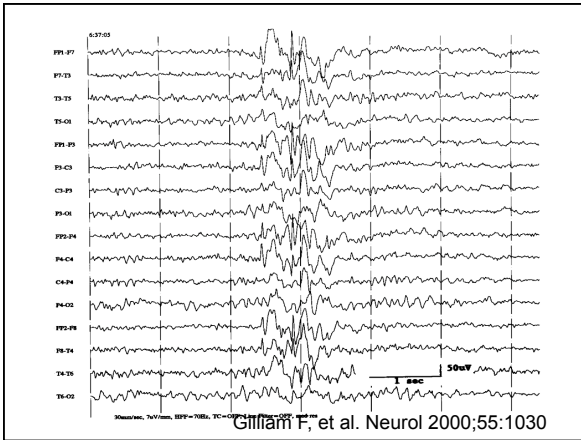
- Gilliam, et al 2000
- 11 cases of adult onset myoclonic epilepsy
- Mean age of onset 39 yrs (range 28-53 yrs)
- Seizure types
  - Myoclonic seizures
  - Absence seizures
  - GTC seizures (less than 1 per year)

Gilliam F, et al. Neurol 2000;55:1030

### Adult onset myoclonic epilepsy

Patient no.	Age at onset of myoclonic seizures, y	Age at onset of absence seizures, y	Age at onset of GTC seizures, y	EEG	Family history
1	37	37	—	poly SW	+
2	34	34	—	poly SW	—
3	53	53	53	S/W	—
4	38	38	38	S/W	+
5	28	—	28	PPR	—
6	41	—	41	PPR	+
7	32	—	32	S/W	Sister with febrile seizures
8	35	—	51	S/W	+
9	25	27	27	S/W	—
10	29	—	29	S/W	—
11	41	—	38	poly SW	Sister with febrile seizures

Gilliam F, et al. Neurol 2000;55:1030

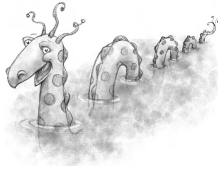


### Adult onset myoclonic epilepsy

- Seizures were controlled in all pts with VPA or LTG
- Authors' conclusion
- Except for the age of onset, the clinical and EEG features were similar to classic IGE

Gilliam F, et al. Neurol 2000;55:1030

### Absence



### IGE with phantom absences

- Found in 3% (13pts) of 410 consecutive epilepsy pts over 16 yrs old in one series
- Clinical manifestations
  - Phantom absences that are difficult to recognize and never appreciate prior to GTC onset
  - Infrequent GTC seizures, usually starting in adulthood
  - Absence status epilepticus occurs in 50% of patients

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

### IGE with phantom absences

- 3 of 13 pts have family history
- Absence status were wrongly diagnosed as complex partial seizures in 4 pts
- EEG: generalized 3-4 Hz spike/polyspikes and wave complexes, no photoparoxysmal response

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

### IGE with phantom absences

