DEFINITION AND CLASSIFICATION OF EPILEPSY

KAMORNWAN KATANYUWONG MD.

6th epilepsy camp 2015 : Nakornprathom

OUTLINE

- **Definition** of epilepsy

  Definition of seizure

  Definition of epilepsy

- Epilepsy **classification**

การให้คำนิยาม ให้การวินิจฉัยอาการชัก หรือ วินิจฉัยโรคลมชัก ≠ การให้การรักษาคนไข้

DEFINITION OF EPILEPSY

Glossary of Descriptive Terminology for Ictal Semiology:
Report of the ILAE Task Force on Classification and Terminology

Epilepsia 2001

Special Article

Epileptic Seizures and Epilepsy: Definitions Proposed by the International League Against Epilepsy (ILAE) and the International Bureau for Epilepsy (IBE)

Epilepsia 2005

An Operational Clinical Definition of Epilepsy

ILAE website 2013

A practical clinical definition of epilepsy

Epilepsia 2014

SEIZURE

- **Greek** : meaning ***to take hold***
- **Modern** : sudden and severe event
- **Seizure = epileptic seizure**
- *Cardiology : heart seizure*

แพทย์ VS ประชาชนทั่วไป

2001

I GENERAL TERMS

1.0 SEMIOLOGY

That branch of linguistics concerned with signs and symptoms.

2.0 EPILEPTIC SEIZURE

Manifestation(s) of epileptic (excessive and/or hypersynchronous), usually self-limited activity of neurons in the brain.

Neuron excitation + Hypersynchrony

3.0 ICTUS

A sudden neurologic occurrence such as a stroke or an epileptic seizure.

4.0 EPILEPSY

a) Epileptic Disorder: A chronic neurologic condition characterized by recurrent epileptic seizures.
b) Epilepsies: Those conditions involving chronic recurrent epileptic seizures that can be considered epileptic disorders.

2005

Table 1: Conceptual Definition of Seizure and Epilepsy – 2005 Report

An **epileptic seizure** is a transient occurrence of signs and/or symptoms due to abnormal excessive or synchronous neuronal activity in the brain.

Epilepsy is a disorder of the brain characterized by an enduring predisposition to generate epileptic seizures, and by the neurobiological, cognitive, psychological, and social consequences of this condition. The definition of epilepsy requires the occurrence of at least one epileptic seizure.

2005

Elements of a definition of seizure	Elements of a definition of epilepsy
• Mode of onset and termination • Clinical manifestations • Abnormal enhanced synchrony	• History of at least one seizure • Enduring alteration in the brain that increases the likelihood or future seizures • Associated neurobiologic, cognitive, psychological and social disturbances

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2013: DEFINITION OF EPILEPSY

A **disease** of the brain defined by any of the following conditions

1. At least 2 unprovoked seizures occurring more than 24 hours apart
2. One unprovoked seizure and a probability of further seizures similar to the general recurrence risk after two unprovoked seizures (approx 75% or more)

2013: WHY 2 ?

- After 2 unprovoked non-febrile seizure, the chance of having another is 73% (Hauser et al 1998) at 4 years

- After a single unprovoked seizure, the chance of having another is 40-52% (Berg&Shinnar 1991)

2013: WHY 24 HOURS APART?

Answer:

if seizures clustering within 24 hours

⇨ risk factor for later seizures

⇨ = risk after a single seizure

2013

- Stroke, CNS infection and trauma is important

- If the patient has a single unprovoked seizure after a remote brain insult ⇨ risk of a second unprovoked seizure = risk for further seizures after two unprovoked seizures

2013

- Some patient with a single unprovoked seizure in a circumstance of an epilepsy syndrome
  - ⇨ high risk of recurrence
  - ⇨ epilepsy

2013

- *Definition/ Diagnosis*

- **Prognosis/outcome**

No longer present ≠ cure

2013: EPILEPSY

Epilepsy is considered **to be no longer present** for

- individuals who had an age-dependent epilepsy syndrome but are now past the applicable age or

- those who have remained seizure-free for at least 10 years off AEDs

- No known risk factors associated with a high probability (≥ 75%) of future seizure

2013

- Cure = disappearance
- Remission = abeyance of a disease

- “ no longer present” = the person no longer has epilepsy, although it does not guarantee that it will not return

2014: DEFINITION OF EPILEPSY

1. At least 2 unprovoked **(or reflex)** seizures occurring > 24 hours apart
2. One unprovoked (or reflex) seizure and a probability of further seizures similar to the general recurrence risk **(>60%)** after 2 unprovoked sz, occurring over the next 10 years
3. Diagnosis of a epilepsy syndrome

Epilepsia 2014

2013 = no longer present

2014

Resolved

- Had age-dependent epilepsy syndrome but are now past the applicable age
- Seizure free for at least 10 years and off AEDs for at least the last 5 years

2013 sz free 10 yrs off AED

Resolved is not identical to remission or cure

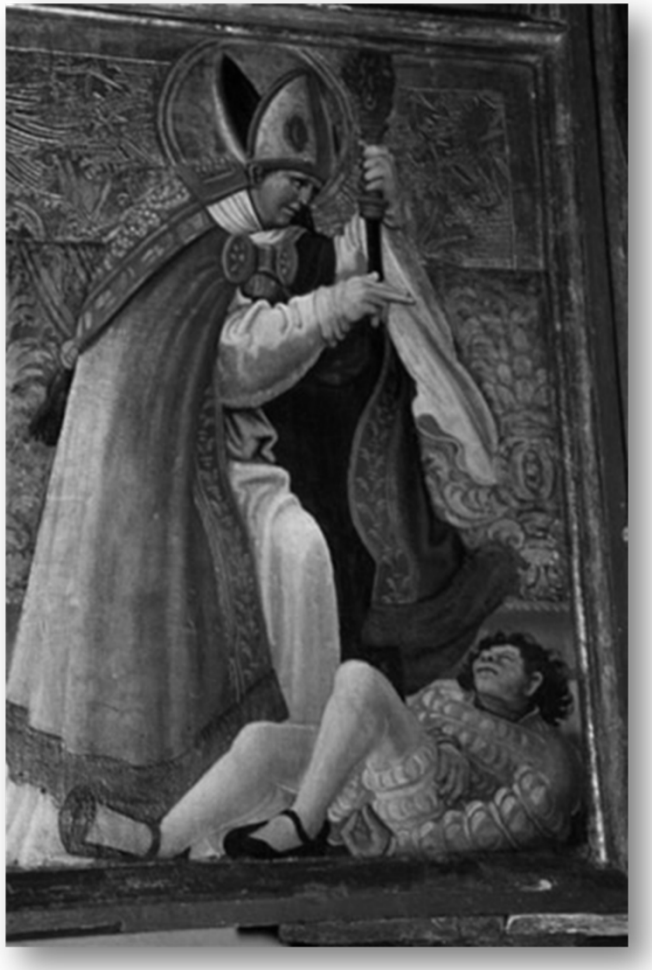
2014

- A decision for treatment does not equate to a diagnosis of epilepsy
- A diagnosis of epilepsy does not require treatment

NEW DEFINITION

- ? Affect prevalence of epilepsy
- Making the clinicians more comfortable in initiating treatment after some unprovoked seizures
- Required specialized diagnostic and interpretative skills- esp in ***assessing recurrence risks*** or in ***diagnosing syndromes***

CLASSIFICATION



WHY CLASSIFICATION IS NEEDED ?

- A universal vocabulary that facilitated communication among clinicians

- Also established a taxonomy foundation for the research on epilepsy

In general

TYPES OF CLASSIFICATION

- Biology:
- Etiology: 1^0 (idiopathic) or 2^0 (symptomatic)
- Pathology: Cancer
- Imaging: Cortical dysplasia
- Clinical criteria e.g. age onset, disease course, distribution of symptoms: HA

- Mixed:

COMMISSION ON CLASSIFICATION AND TERMINOLOGY OF ILAE

- Classification of Epileptic Seizures in 1981
- Classification of Epilepsies and Epileptic syndromes in 1989

- A proposed diagnostic scheme for people with epileptic seizures and with epilepsy : Report of the ILAE Task Force on Classification and Terminology in 2001

- ….2006,……2010……2013……2015

2005-2009

ILAE CLASSIFICATION WORKING GROUP

Epilepsia, 51(4):676–685, 2010
doi: 10.1111/j.1528-1167.2010.02522.x

SPECIAL REPORT

Revised terminology and concepts for organization of seizures and epilepsies: Report of the ILAE Commission on Classification and Terminology, 2005–2009

*†Anne T. Berg, ‡Samuel F. Berkovic, §Martin J. Brodie, ¶Jeffrey Buchhalter, #**J. Helen Cross, ††Walter van Emde Boas, ‡‡Jerome Engel, §§Jacqueline French, ¶¶Tracy A. Glauser, ##Gary W. Mathern, ***Solomon L. Moshé, †Douglas Nordli, †††Perrine Plouin, and ‡Ingrid E. Scheffer

THE CLASSIFICATION CRITERIA OF

Epilepsies (Merlis 1970)

- Clinical criteria
  - Seizures
  - Neurologic status
  - Age of onset
  - Etiology
- EEG criteria
  - Interictal EEG
  - Ictal EEG

Epileptic seizures (Gastuat 1970)

- Clinical seizure type
- EEG seizure type
- EEG interictal expression
- Anatomical substrate
- Etiology
- Age

ILAE 1981

Clinical seizure type | EEG sz type | EEG interictal expression

1. Partial (focal, local) seizures

Simple partial sz
- with motor signs
- with somatosensory symptoms
- with autonomic symptoms and signs
- with psychic symptoms

Complex partial sz
- start with SPS followed by impairment of consciousness
- with impairment of consciousness at onset

Partial sz evolving to 2^0 gen sz
- SPS → GTC
- CPS → GTC
- SPS → CPS → GTC

2. Generalized sz (convulsive and non-convulsive)

Absence, Myoclonic, Clonic, Tonic, Tonic-clonic, Atonic

3. Unclassified epileptic sz

4. Prolonged or repetitive seizure (status epilepticus)

ILAE 1989

1. **Localization-related epilepsies and syndromes**
   - 1.1 Idiopathic
     - benign childhood epilepsy with centro-temporal spike
     - childhood epilepsy with occipital paroxysms
     - primary reading epilepsy
   - 1.2 Symptomatic e.g. TLE, FLE, PLE, OLE
   - 1.3 Cryptogenic

ILAE 1989

2. Generalized epilepsies and syndromes

2.1 Idiopathic (with age-related onset, listed in order to age)
- Benign neonatal familial convulsions
- Benign neonatal convulsions
- Benign myoclonic epilepsy of infancy
- Childhood absence epilepsy (pyknolepsy)
- Juvenile absence epilepsy
- Juvenile myoclonic epilepsy
- Epilepsy w grand mal (GTCS) sz on awakening
- etc.

2.2 Cryptogenic or symptomatic (in order to age)
- West syndrome
- Lennox-Gastaut syndrome
- Epilepsy w myoclonic-astatic sz
- Epilepsy w myoclonic absences

ILAE 1989

2. Generalized epilepsies and syndromes

2.3 Symptomatic

2.3.1 Non specific etiology
- EME
- EIEE w supression burst
- other symptomatic generalised epilepsies not defined above

2.3.2 Specific syndromes/etiologies
- Cerebral malformation
- IBEM

3. **Epilepsies and syndromes undetermined** whether focal or generalized e.g. SMEI, LKS, CSWS, neonatal sz
4. **Special syndromes** e.g. FC, reflex epilepsy, isolated sz

TWO DICHOTOMIES, A 4-PART CLASSIFICATION

Cryptogenic
Special syndromes

	Localization-related	**Generalized**
Idiopathic	Localization-related Idiopathic **IPE**	Generalized Idiopathic **IGE**
Symptomatic	Localization-related Symptomatic **SPE**	Generalized Symptomatic **SGE**

2001

Axis 1: Ictal phenomenology

Axis 2: Seizure type: Partial Sz or Gen Sz

Axis 3: Epileptic syndromes:

Axis 4: Etiology: Idiopathic, Cryptogenic, Symptomatic

Axis 5: Impairment

2010

2005-2009

ILAE CLASSIFICATION WORKING GROUP

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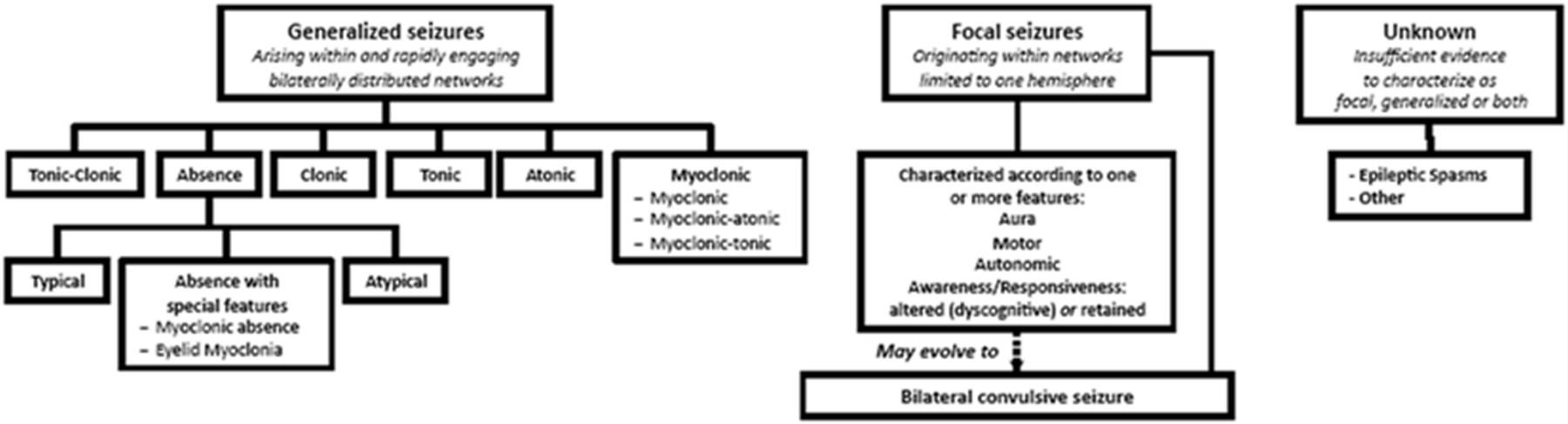
ILAE Proposal for Revised Terminology for Organization of Seizures and Epilepsies 2010

Classification of Seizures



Changes in terminology and concepts

New Term and Concept	Examples	Old Term and Concept
Etiology		
Genetic: *genetic defect directly contributes to the epilepsy and seizures are the core symptom of the disorder*	*Channelopathies, Glut1 deficiency, etc.*	**Idiopathic:** *presumed genetic*
Structural-metabolic: *caused by a structural or metabolic disorder of the brain*	*Tuberous sclerosis, cortical malformations, etc.*	**Symptomatic:** *secondary to a known or presumed disorder of the brain*
Unknown: *the cause is unknown and might be genetic, structural or metabolic*		**Cryptogenic:** *presumed symptomatic*
Terminology		**Terms no longer recommended**
Self-limited: *tendency to resolve spontaneously with time* **Pharmacoresponsive**: *highly likely to be controlled with medication*		**Benign** **Catastrophic**
Focal seizures: *seizure semiology described according to specific subjective (auras), motor, autonomic, and dyscognitive features*		**Complex partial** **Simple partial**
Evolving to a bilateral convulsive seizure: *eg. tonic, clonic, tonic-clonic*		**Secondarily generalized**

old

New

Table 1. Comparison of major changes between the 1989 and 1981 Classification and Terminology and the newly proposed Terminology and Concepts (Commission 1981, 1989; Berg et al., 2010)

Old terminology and concepts	Recommended new terminology and concepts
Focal and generalized	
For seizures	
Focal (previously "partial"): the first clinical and electroencephalographic changes indicate initial activation of a system of neurons limited to a part of one cerebral hemisphere	Focal seizures are conceptualized as originating at some point within networks limited to one hemisphere
Generalized: the first clinical changes indicate initial involvement of both hemispheres	Generalized seizures are conceptualized as originating at some point within and rapidly engaging bilaterally distributed networks
For epilepsies	
Localization-related (focal, partial): epilepsies with focal seizures *Generalized*: epilepsies with generalized seizures	These terms were abandoned as overarching categories for classifying epilepsies per se, as many syndromes include both seizure types; they may still apply in some but not all instances

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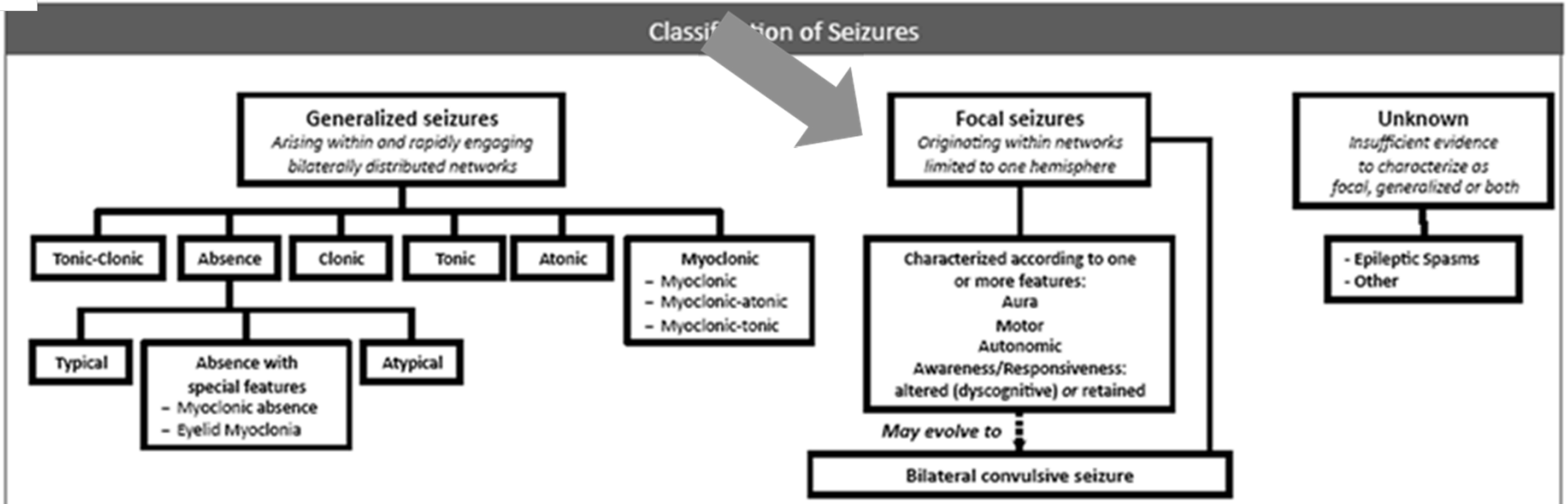

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TABLE 1-4 Seizure Types and Terminology Used in the 1981 Classification of Seizures and Recommended in the 2010 Report[a,b]

Mode of Onset	1981 Seizure Types[c]	2010 Seizure Descriptions[d]
Focal	Simple partial With motor signs With sensory symptoms With autonomic symptoms With psychic symptoms (but no impaired consciousness)	Without impairment of consciousness or awareness: With observable motor or autonomic components Involving subjective sensory or psychic phenomena only, corresponding to the concept of an aura
	Complex partial Consciousness impaired at onset Simple partial onset followed by impairment of consciousness	With impairment of consciousness or awareness. *Dyscognitive* is a term that has been proposed for this concept.[21]
	Partial evolving to secondarily generalized seizure (tonic, clonic, or tonic-clonic) Simple evolving to generalized tonic-clonic Complex evolving to generalized tonic-clonic (including those with simple partial onset)	Evolving to a bilateral, convulsive seizure (involving tonic, clonic, or tonic and clonic components).
Generalized onset	Tonic-clonic Myoclonic	Tonic-clonic (in any combination) Myoclonic Myoclonic Myoclonic-atonic Myoclonic-tonic
	Absence With various accompanying manifestations Atypical	Absence Typical absence Atypical absence With special features Eyelid myoclonia[e] Myoclonic absence
	Clonic Tonic Atonic (astatic)	Clonic Tonic Atonic
Not clear	Anything that does not fit in above, eg, rhythmic eye movements, chewing, swimming movements	Epileptic spasms

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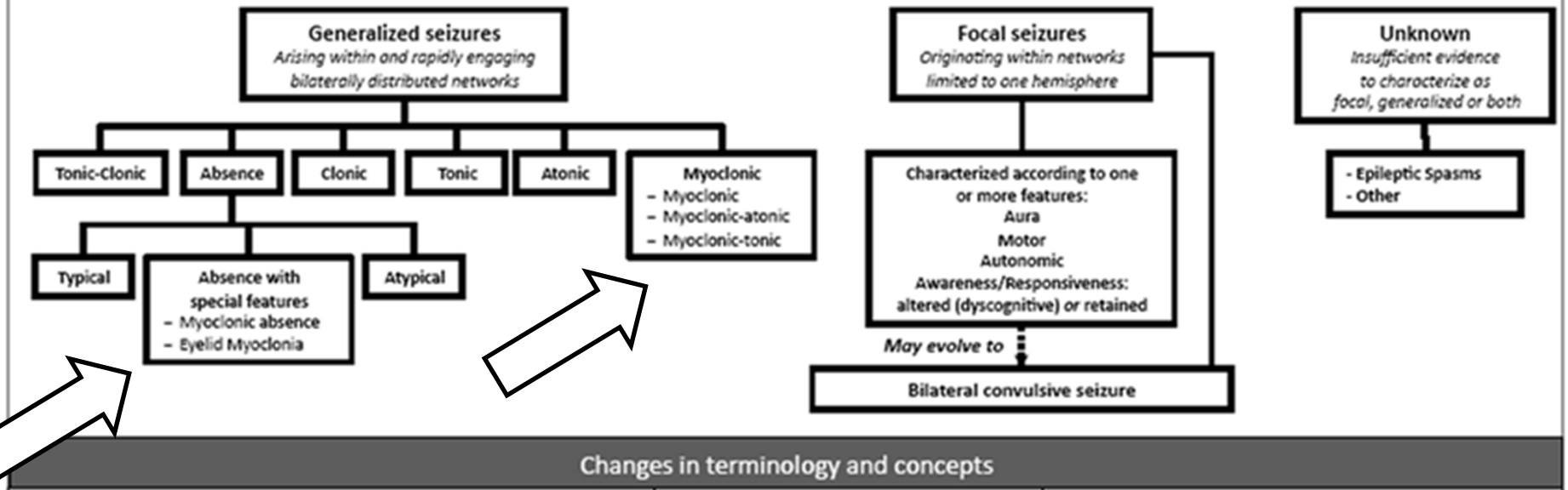

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focal seizure should be described according to their manifestation

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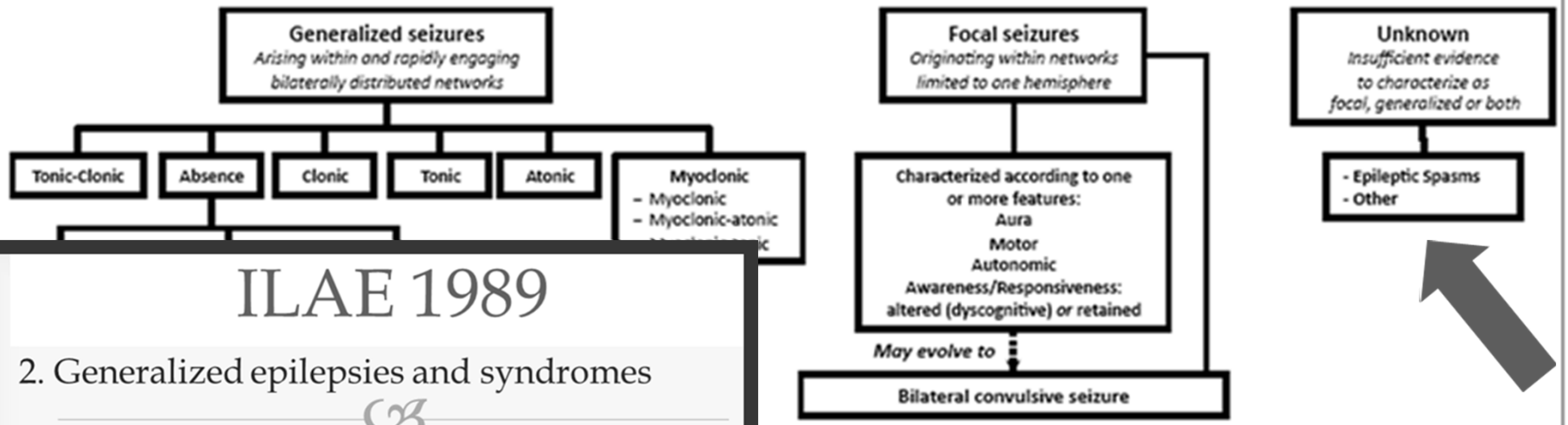

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Symptomatic: the epilepsy is the consequence of a known or suspected disorder of the central nervous system	Structural/metabolic: there is a distinct other structural or metabolic condition or disease that has been demonstrated to be associated with a substantially increased risk of developing epilepsy. These disorders may be of acquired or genetic origin. When of genetic origin, there is a separate disorder interposed between the gene defect and the epilepsy
Cryptogenic: this refers to a disorder whose cause is hidden or occult. Cryptogenic epilepsies are presumed to be symptomatic	Unknown: the nature of the underlying cause is unknown; it may have a fundamental genetic basis (e.g., a previously unrecognized channelopathy) or it may be the consequence of an unrecognized structural or metabolic disorder not yet identified

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2015: Structural/metabolic……
include immune and infectious cause

old

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II

ILAE Proposal for Revised Terminology for Organization of Seizures and Epilepsies 2010

Electroclinical Syndromes and Other Epilepsies Grouped by Specificity of Diagnosis

Electroclinical syndromes

One example of how syndromes can be organized: Arranged by typical age at onset*

Neonatal period
- Benign neonatal seizures^
- Benign familial neonatal epilepsy (BFNE)
- Ohtahara syndrome
- Early Myoclonic encephalopathy (EME)

Infancy
- Febrile seizures^, Febrile seizures plus (FS+)
- Benign infantile epilepsy
- Benign familial infantile epilepsy (BFIE)
- West syndrome
- Dravet syndrome
- Myoclonic epilepsy in infancy (MEI)
- Myoclonic encephalopathy in nonprogressive disorders
- Epilepsy of infancy with migrating focal seizures

Childhood
- Febrile seizures^, Febrile seizures plus (FS+)
- Early onset childhood occipital epilepsy (Panayiotopoulos syndrome)
- Epilepsy with myoclonic atonic (previously astatic) seizures
- Childhood absence epilepsy (CAE)
- Benign epilepsy with centrotemporal spikes (BECTS)
- Autosomal dominant nocturnal frontal lobe epilepsy (ADNFLE)
- Late onset childhood occipital epilepsy (Gastaut type)
- Epilepsy with myoclonic absences
- Lennox-Gastaut syndrome (LGS)
- Epileptic encephalopathy with continuous spike-and-wave during sleep (CSWS)+
- Landau-Kleffner syndrome (LKS)

Adolescence – Adult
- Juvenile absence epilepsy (JAE)
- Juvenile myoclonic epilepsy (JME)
- Epilepsy with generalized tonic-clonic seizures alone
- Autosomal dominant epilepsy with auditory features (ADEAF)
- Other familial temporal lobe epilepsies

Variable age at onset
- Familial focal epilepsy with variable foci (childhood to adult)
- Progressive myoclonus epilepsies (PME)
- Reflex epilepsies

Distinctive constellations/surgical syndromes

Distinctive constellations/Surgical syndromes
- Mesial temporal lobe epilepsy with hippocampal sclerosis (MTLE with HS)
- Rasmussen syndrome
- Gelastic seizures with hypothalamic hamartoma
- Hemiconvulsion-hemiplegia-epilepsy

Nonsyndromic epilepsies**

Epilepsies attributed to and organized by structural-metabolic causes
- Malformations of cortical development (hemimegalencephaly, heterotopias, etc.)
- Neurocutaneous syndromes (tuberous sclerosis complex, Sturge-Weber, etc.)
- Tumor, infection, trauma, angioma, antenatal and perinatal insults, stroke, etc

Epilepsies of unknown cause

* The arrangement of eletroclinical syndromes does not reflect etiology.
^ Not traditionally diagnosed as epilepsy
+ Sometimes referred to as Electrical Status Epilepticus during Slow Sleep (ESES)
** Forms of epilepsies not meeting criteria for specific syndromes or constellations

This Proposal is a work in progress.....
We welcome your thoughts on this proposal. Please visit our Classification & Terminology Discussion Group at: http://community.ilae-epilepsy.org/home/ to login and register your comments.

2010

- Classification system versus diagnosis

- 2010 entail little or no change in what health care providers do in daily practice
  (diagnose and treat individual patients)

- CAE : IGE (old)
  : generalized epilepsy, absence, genetic cause (new)

Thank you for your time