Epilepsy Classification: Hot Controversies in 2012
December 4\textsuperscript{th}, 2012

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Symposium Co-Chairs
North American Commission of the International League Against Epilepsy Symposium 2012
Disclosure

Dr. Haut
Acorda
Vivus
Upsher Smith
Neuronex

Consultant
Consultant
Consultant

Dr. Scheffer
Nothing to disclose
Learning Objectives

1. Participants will become familiar with the revised classification system for epilepsy, which will lead to greater diagnostic specificity for epilepsy treatment and research
2. Participants will become more aware of the role of genetic and immunologic testing in epilepsy
3. Improvement in coding accuracy for clinical epilepsy practice.
Initial controversies

Elimination of the “focal” and “generalized “ epilepsy categories

Revision of the etiologic subgroups from “symptomatic; cryptogenic; and idiopathic” to “genetic; structural-metabolic; and unknown”

Use of the term “constellations”

Addressing these controversies

Ongoing feedback has been welcome

Development of the new organization continues to be an evolving process

The most recent revisions will be presented during this symposium
Schedule

• Introduction–Sheryl Haut, M.D.

• Update on the new Organization: Where Have the Modifications Taken Us? Ingrid E. Scheffer, M.B.B.S., Ph.D.

• Diagnostic Specificity: Applying This Concept to Every Patient - J. Helen Cross, M.B.Ch.B., Ph.D.

Controversies

• Genetic: How Do I Tell the Patient? Sameer Zuberi, M.B.Ch.B, M.D.

• Structural: Genetic or Acquired? James Barkovich MD

• Immune: Which Patients Should Be Tested? Christian Bien, M.D.

• Coding: Will This Make a Difference to My Practice? Donna C. Bergen, M.D.

• Conclusions - Ingrid E. Scheffer, M.B.B.S., Ph.D.

• Discussion
Update on the new Organization: Where have the modifications taken us?

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Disclosure

Name of Commercial Funding:
NINDS, CURE, US DOD, NHMRC, ARC
SAB: Dravet.org, PCDH19 Alliance

Type of Financial Relationship
UCB, Janssen-Cilag, Athena Diagnostics, Biocodex, GlaxoSmithKline
Impact on Clinical Care and Practice

• Primary clinical tool in daily practice
• Affects every patient we see
• Updated terminology for seizures
• Approaches to epilepsy diagnosis
• New subgroups for etiology of epilepsy
Learning Objectives

• To learn about the new Organization
• To understand how to use the new Organization
Purpose of the International Classification of Seizures and Epilepsies

- To provide a common international terminology and classification
- Largely for clinical (treatment) purposes
- Purpose of classification: to organize items according to their fundamental relationships

SPECIAL REPORT


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Refinements to the Organization

• We have listened to your valuable feedback!
• “Reinstated” focal and generalized epilepsies as useful diagnostic entities where they work
• Different approaches to epilepsy diagnosis
• Modified the organization to reflect emerging etiological subgroups
• Aim to reflect current understanding
Concepts

- Seizures
- Epilepsies
- Diagnostic domains
- Syndromes
- Etiologies
Focal seizures

• Originate within networks limited to one hemisphere
• May be discretely localized or more widely distributed....
Focal seizures

Blume et al Epilepsia 2001

• Previous term: simple partial
  • No impairment of awareness or consciousness
  • Motor or autonomic components eg. focal clonic
  • Subjective sensory or psychic features -&gt; Aura

• Previous term: complex partial
  • Impairment of awareness or consciousness
    ➔ Dyscognitive

• Previous term: secondarily generalized
  • Evolving to bilateral, convulsive seizure
  • With tonic, clonic or tonic and clonic components
Generalized seizures

- Originate at some point within and rapidly engage bilaterally distributed networks
- Can include cortical and subcortical structures but not necessarily the entire cortex
Generalized seizures

Tonic-clonic (in any combination)

Absence
  - Typical
  - Atypical
  - Absence with special features
    Myoclonic absence
    Eyelid myoclonia

Myoclonic
  - Myoclonic
  - Myoclonic atonic
  - Myoclonic tonic

Clonic
Tonic
Atonic

Seizure types thought to occur within and result from rapid engagement of bilaterally distributed systems
Epilepsies

- Generalized epilepsies
- Focal epilepsies

Use where they work!

- Not every patient can be classified as either focal or generalized
  - Overlap not unusual
  - Especially many epileptic encephalopathies
    - e.g. Dravet syndrome
Electroclinical epilepsy syndromes

- Unchanged!
- A diagnosis can be made as previously e.g.
  - Lennox-Gastaut syndrome
  - Childhood Absence Epilepsy
- A diagnosis is *not* the same as a classification
Clinicoradiological entities

- Replace “constellation” as does not translate
- Denote associated findings with treatment implications such as surgery
  - Mesial temporal lobe seizures and hippocampal sclerosis
  - Gelastic seizures and hypothalamic hamartoma
Etiology

• Genetic
• Structural
• Metabolic
• Immune
• Infectious
• Unknown

• Use terms that mean what they say!
• Replace old fashioned terms: idiopathic, symptomatic, cryptogenic
Genetic

• Concept:
  – Epilepsy is the direct result of a known or inferred genetic defect
  – Seizures are the core symptom of the disorder

• Evidence
  – appropriately designed family studies or
  – replicated molecular genetic studies

• Genetic does not exclude the contribution of environmental factors
Concept: epilepsy is the result of a distinct other structural condition or disease – eg. tuberous sclerosis

Evidence: Must have a substantially increased risk of developing epilepsy with the condition

Can have two etiologies: eg. Structural-genetic
Metabolic

- **Concept:** epilepsy is the result of a metabolic condition or disease with widespread manifestations
  - eg. aminoacidopathies
  - pyridoxine-dependent seizures

- **Evidence:** Must have a substantially increased risk of developing epilepsy with the metabolic condition
• **Concept**: epilepsy is the result of autoimmune mediated central nervous system inflammation 
e.g. autoimmune encephalitides  
– anti-NMDA encephalitis  
– limbic encephalitis

• **Evidence**: Must have a substantially increased risk of developing epilepsy with the immune condition
Infectious

• **Concept:** epilepsy is the result of an infectious cause eg. Tuberculosis, HIV, neurocysticercosis, malaria

• **Evidence:** Must have a substantially increased risk of developing epilepsy with infection
Unknown

• Concept: The underlying cause is unknown
Modifications to the Organization of the Epilepsies

- Changes in response to feedback - more “user friendly”
- Adoption of new seizure terminology occurring; concepts well accepted
- Diagnostic domains
  - Electroclinical syndromes unchanged
  - Clinicoradiological entities
  - Etiological subgroups now separated & updated
Modifications to the Organization of the Epilepsies

• Multiple ways to approach epilepsy diagnosis
• Flexible – you can organize it how you wish
• Must remain a dynamic and evolving classification
• Future – scientifically based classification founded on biological mechanisms
ILAE Commission for Classification and Terminology

- Anne Berg
- Edouard Hirsch
- Sameer Zuberi
- Pippo Capovilla
- Mary Connolly
- Laura Guilhoto
- Yue-Hua Zhang
- Sam Berkovic
- Doug Nordli
- Ingrid Scheffer

Classification Taskforce

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- Andrew Lux
- Lynette Sadleir
- Stephan Schuele
- Yoshimi Sogawa
- Elaine Wirrell
- Jeffrey Buchhalter