Received: 11 August 2022 | Accepted: 20 October 2022

DOI: 10.1002/epi4.12665

Epilepsia Open™ Open Access

CONCEPTS AND HYPOTHESES

3D figure of epilepsy syndromes

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Abstract

We propose an instructive figure that summarized the classification of epilepsy syndromes according to the 2022 report of the ILAE Task Force on Nosology and Definitions. Our aim is to present on the same figure different concepts such as the names of epilepsy syndromes, their extreme and classical ages of onset, their epilepsy types (generalized, focal, or generalized and focal) but also their membership in groups of epilepsy syndromes as for self-limited or developmental and epileptic encephalopathies. With this figure, we provide an interactive tool, as supplementary data, helping to present this classification and link it to electroclinical mandatory, alerts, and exclusionary criteria of each syndrome, in accordance with the ILAE position papers on syndromes classification and nosology. This report may be used as an illustrative tool for teaching epilepsy syndromes and as a practical and comprehensive aid for the classification of epilepsy individuals' syndromes.

KEYWORDS

developmental and epileptic encephalopathy, epilepsy syndromes, interactive classification, self-limited epilepsies, teaching tool

The production of instructive figures that summarize complex concepts, such as classification of epilepsy syndromes for the education of epileptologists, general practitioners, and patients and their families can be very challenging. We propose the following figure (Figure 1) to illustrate in a single comprehensive graphic representation the various epilepsy syndromes, maintaining consistency with the 2017 ILAE Classification of the Epilepsies paper[1] and of the ILAE Task Force on Nosology and Definitions position papers in 2022.[2–6]

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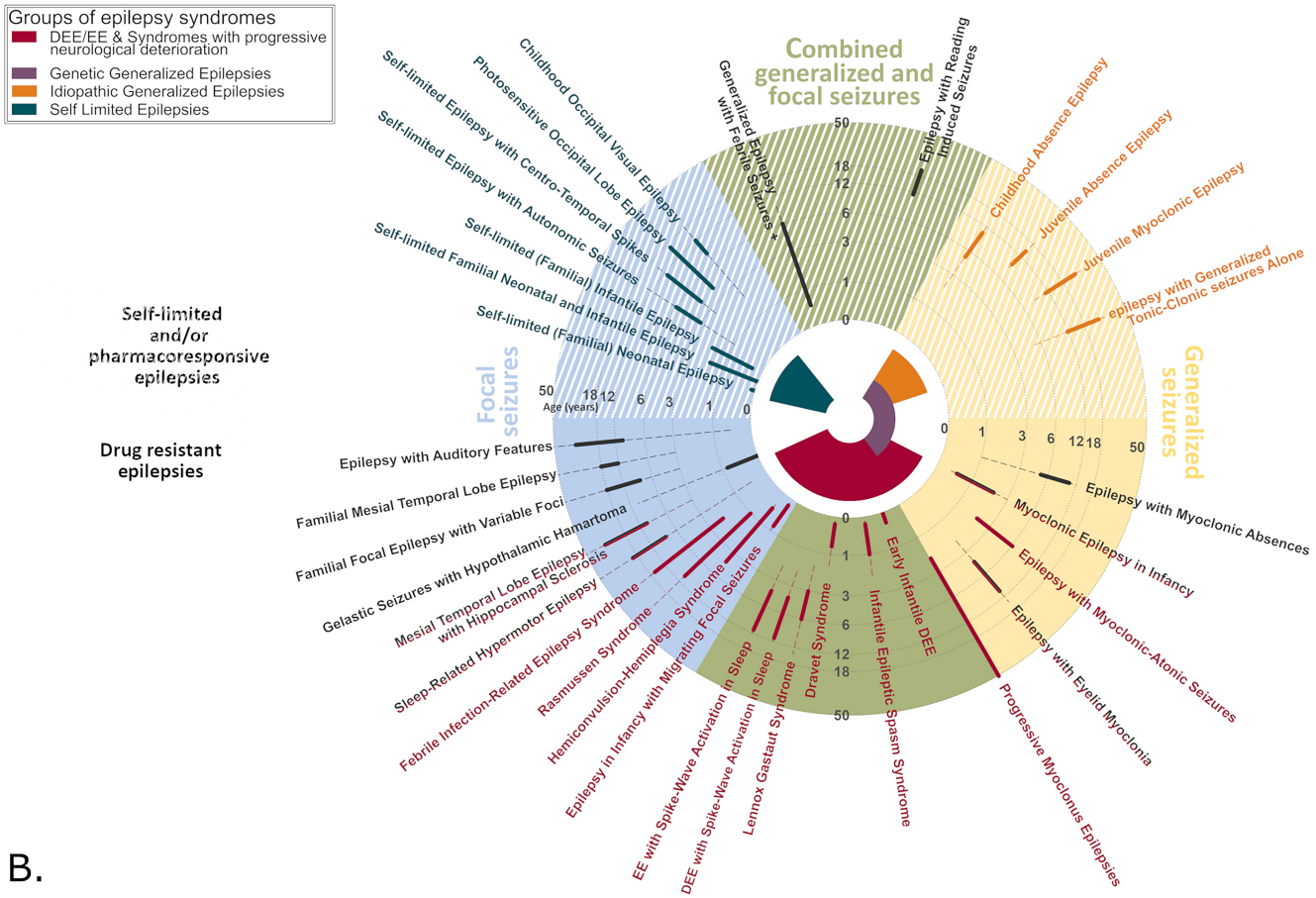
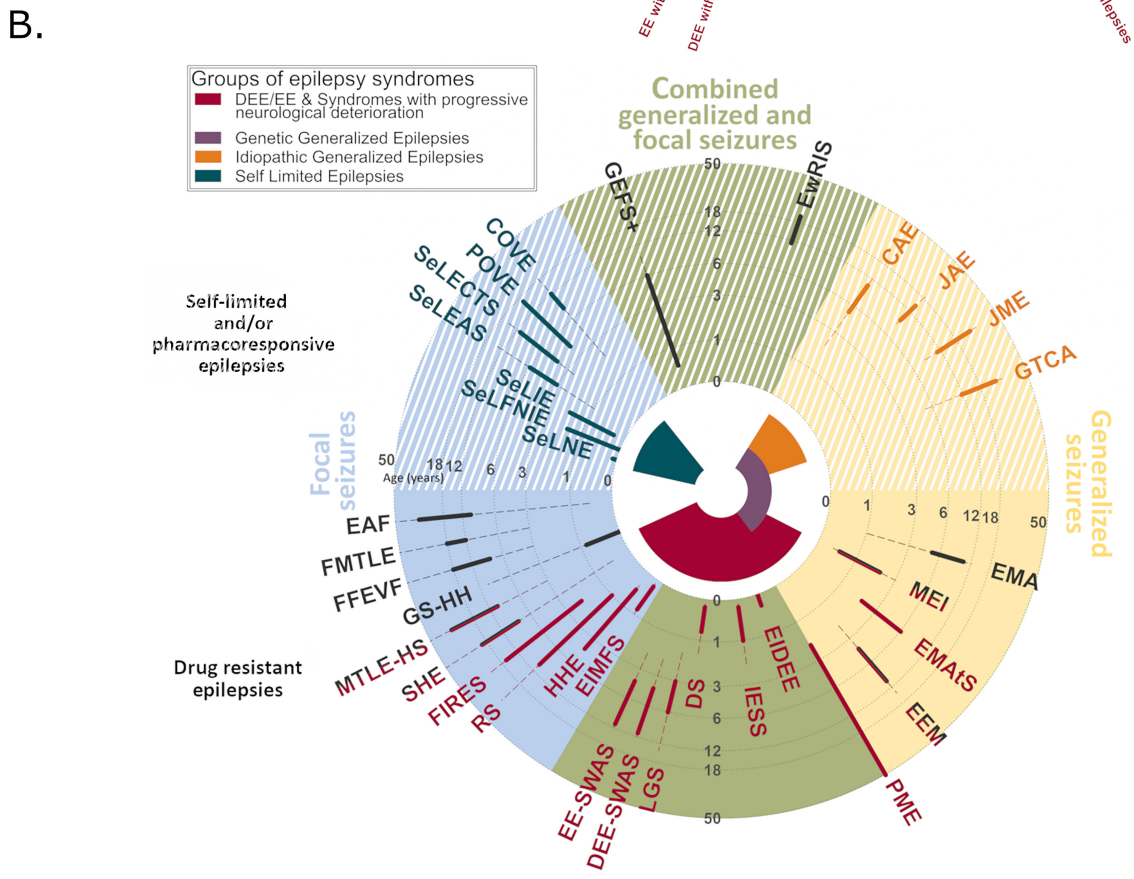
A.


Groups of epilepsy syndromes
DEE/EE & Syndromes with progressive neurological deterioration
Genetic Generalized Epilepsies
Idiopathic Generalized Epilepsies
Self Limited Epilepsies
Combined generalized and focal seizures
Self-limited and/or pharmacoresponsive epilepsies
Drug resistant epilepsies
Focal seizures
Generalized seizures
Age (years)
Childhood Occipital Visual Epilepsy
Photosensitive Occipital Lobe Epilepsy
Self-limited Epilepsy with Centro-Temporal Spikes
Self-limited Epilepsy with Autonomic Seizures
Self-limited (Familial) Infantile Epilepsy
Self-limited Familial Neonatal and Infantile Epilepsy
Self-limited (Familial) Neonatal Epilepsy
Generalized Epilepsy with Febrile Seizures +
Epilepsy with Reading Induced Seizures
Childhood Absence Epilepsy
Juvenile Absence Epilepsy
Juvenile Myoclonic Epilepsy
epilepsy with Generalized Tonic-Clonic seizures Alone
Epilepsy with Auditory Features
Familial Mesial Temporal Lobe Epilepsy
Familial Focal Epilepsy with Variable Foci
Gelastic Seizures with Hypothalamic Hamartoma
Mesial Temporal Lobe Epilepsy with Hippocampal Sclerosis
Sleep-Related Hypermotor Epilepsy
Febrile Infection-Related Epilepsy Syndrome
Rasmussen Syndrome
Hemiconvulsion-Hemiplegia Syndrome
Epilepsy in Infancy with Migrating Focal Seizures
EE with Spike-Wave Activation in Sleep
DEE with Spike-Wave Activation in Sleep
Lennox Gastaut Syndrome
Dravet Syndrome
Infantile Epileptic Spasm Syndrome
Early Infantile DEE
Epilepsy with Myoclonic Absences
Myoclonic Epilepsy in Infancy
Epilepsy with Myoclonic-Atonic Seizures
Epilepsy with Eyelid Myoclonia
Progressive Myoclonus Epilepsies

B.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Self Limited Epilepsies
Combined generalized and focal seizures
Self-limited and/or pharmacoresponsive epilepsies
Drug resistant epilepsies
Focal seizures
Generalized seizures
Age (years)
GEFS+
EmRIS
COVE
POVE
SeLECTS
SeLEAS
SeLIE
SeLFNIE
SeLNE
CAE
JAE
JME
GTCA
EAF
FMTLE
FFEVF
GS-HH
MTLE-HS
SHE
FIRES
RS
HHE
EIMFS
EE-SWAS
DEE-SWAS
LGS
DS
IESS
EIDEE
PME
EEM
EMAtS
MEI
EMA

FIGURE 1 The epilepsy syndromes according to the 2017 epilepsy syndrome classification[1] and the 2022 report of the ILAE Task Force on Nosology and Definitions,[2–6] using the full names of the different epilepsy syndromes (A) or their specific abbreviations (B). The different epilepsy syndromes are divided into different colored areas according to their type of seizure: generalized in yellow, focal in blue, or combined focal and generalized in green. The concentric dotted circles indicated the age of onset of the syndrome. For each epilepsy syndrome, the dotted line represented the extreme age of onset and the thick line the classical age. The main groups of epilepsy syndromes are depicted in specific colors: self-limiting epilepsies in turquoise, idiopathic generalized epilepsies in orange, developmental and/or epileptic encephalopathies and syndromes with progressive neurological deterioration in red and genetic generalized epilepsies in purple. Note that idiopathic generalized epilepsies are a subcategory of genetic generalized epilepsies. Developmental and Epileptic Encephalopathy (DEEs) are a group of epilepsy syndromes where impairments (encephalopathy) are due to both the underlying etiology of the epilepsy and frequent seizures or epileptiform discharges, whereas in epileptic encephalopathy (EE), the encephalopathy is due to frequent seizures or epileptiform discharges. A horizontal axis was used to separate self-limited and/or pharmacoresponsive epilepsies (white diagonal hatching) from commonly drug-resistant epilepsies. The letters used for the abbreviations of the different epilepsy syndromes have been bolded. It is important to bear in mind that this figure reflects the current organization of epilepsy syndromes as defined in the 2022 report of the ILAE Working Group on Nosology and Definitions, but it gives scant information on the underlying etiologies.

This proposed multilevel figure considers the new terminology used in the position papers and encompasses several dimensions.

First, we classified the epilepsy syndromes according to epilepsy type as reported in the ILAE classification, namely focal, generalized, and combined generalized and focal.[1] Second, we used a concentric scale to illustrate the age of onset of the different epilepsy syndromes, an important criterion reported in the definition of the syndromes. By using a continuous logarithmic scale and not only a qualitative one, we were also able to go beyond childhood including variable age syndromes and syndromes with adult onset. We detailed the extreme range of onset age using dotted lines and the classical age range using a solid line. Third, to identify and visualize the different groups of epilepsy syndromes as per the position papers,[2–6] we filled the groups with specific color (red for developmental and/or epileptic encephalopathies and syndromes with progressive neurological deterioration, turquoise for self-limited epilepsies, orange for idiopathic generalized epilepsies, and pink for genetic generalized epilepsies). Finally, we added the self-limited dimension separating the figure into two blocks, self-limited often drug-responsive epilepsies with white diagonal hatching versus drug-resistant epilepsies and acknowledge that the separation might not be always very clear.

In conclusion, this figure can be used to succinctly illustrate the entire classification but might be also parcellated according to the user's needs. It can be an illustrative tool for teaching epilepsy syndromes and can also be used as a practical aid for syndrome classification of patients.

ACKNOWLEDGMENT

RN acknowledges GEEN-DS chair position institut lmagine is supported by FAMA Fund hosted by Swiss Philanthropy Foundation.

CONFLICT OF INTEREST

None of the authors has any conflict of interest to disclose related to this study. We confirm that we have read the Journal's position on issues involved in ethical publication and affirm that this report is consistent with those guidelines.

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SUPPORTING INFORMATION

Additional supporting information can be found online in the Supporting Information section at the end of this article.

How to cite this article: Nabbout R, Kuchenbuch M, Tinuper P, Cross JH, Wirrell E. 3D figure of epilepsy syndromes. Epilepsia Open. 2023;8:217–220. https://doi.org/10.1002/epi4.12665