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► **To cite this version:**

Rima Nabbout, Mathieu Kuchenbuch, Paolo Tinuper, J. Helen Cross, Elaine Wirrell. 3D figure of epilepsy syndromes. *Epilepsia Open*, 2023, 8 (1), pp.217-220. 10.1002/epi4.12665 . hal-04095763

HAL Id: hal-04095763

<https://u-paris.hal.science/hal-04095763v1>

Submitted on 12 May 2023

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CONCEPTS AND HYPOTHESES

3D figure of epilepsy syndromes

Rima Nabbout^{1,2}  | Mathieu Kuchenbuch^{3,4}  | Paolo Tinuper^{5,6}  |
J. Helen Cross⁷  | Elaine Wirrell⁸ 

¹Reference Center for Rare Epilepsies, Department of Pediatric Neurology, Necker-Enfants Malades Hospital, APHP, member of EpiCARE, University Paris cité, Paris, France

²Imagine Institute, National Institute of Health and Medical Research, Mixed Unit of Research, INSERM, Paris, France

³Department of Pediatric Neurology, University Hospital of Nancy, member of EpiCARE, Lorraine University, Nancy, France

⁴Research Center for Automatic Control of Nancy (CRAN), Lorraine University, CNRS, UMR, Vandoeuvre, France

⁵Department of Biomedical and Neuromotor Sciences, University of Bologna, Bologna, Italy

⁶Institute of Neurological Sciences, member of EpiCARE, Scientific Institute for Research and Health Care, Bologna, Italy

⁷Programme of Developmental Neurosciences, University College London National Institute for Health Research Biomedical Research Centre Great Ormond Street Institute of Child Health, Great Ormond Street Hospital for Children, and Young Epilepsy Lingfield, London, UK

⁸Divisions of Child and Adolescent Neurology and Epilepsy, Department of Neurology, Mayo Clinic, Rochester, Minnesota, USA

Correspondence

Rima Nabbout, Reference centre for rare epilepsies, Department of Pediatric Neurology, Necker Enfants Malades University Hospital, APHP, 149 rue de Sèvres, Paris 75015, France.

Email: rima.nabbout@aphp.fr and rimanabbout@yahoo.com

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 electro-clinical 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 paper¹ and of the ILAE Task Force on Nosology and Definitions position papers in 2022.²⁻⁶

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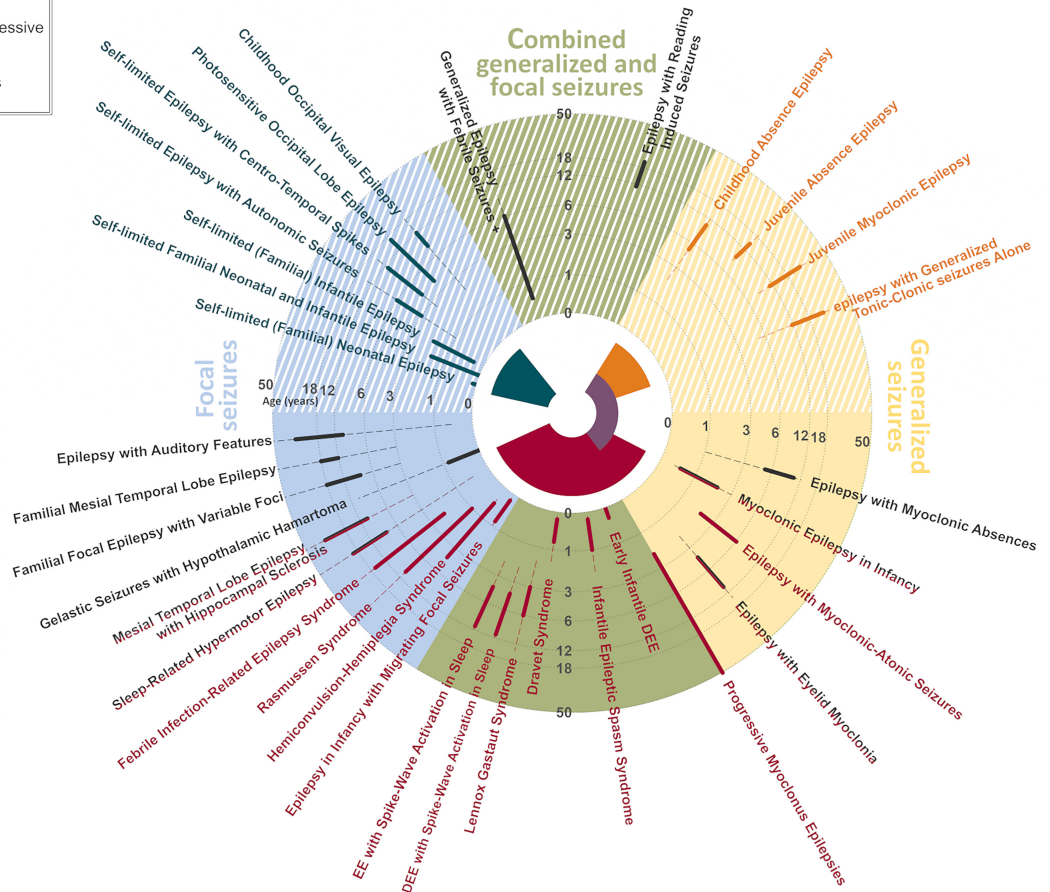
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A.

- Groups of epilepsy syndromes
- DEE/EE & Syndromes with progressive neurological deterioration
 - Genetic Generalized Epilepsies
 - Idiopathic Generalized Epilepsies
 - Self Limited Epilepsies

Self-limited and/or pharmacoresponsive epilepsies

Drug resistant epilepsies



B.

- Groups of epilepsy syndromes
- DEE/EE & Syndromes with progressive neurological deterioration
 - Genetic Generalized Epilepsies
 - Idiopathic Generalized Epilepsies
 - Self Limited Epilepsies

Self-limited and/or pharmacoresponsive epilepsies

Drug resistant epilepsies

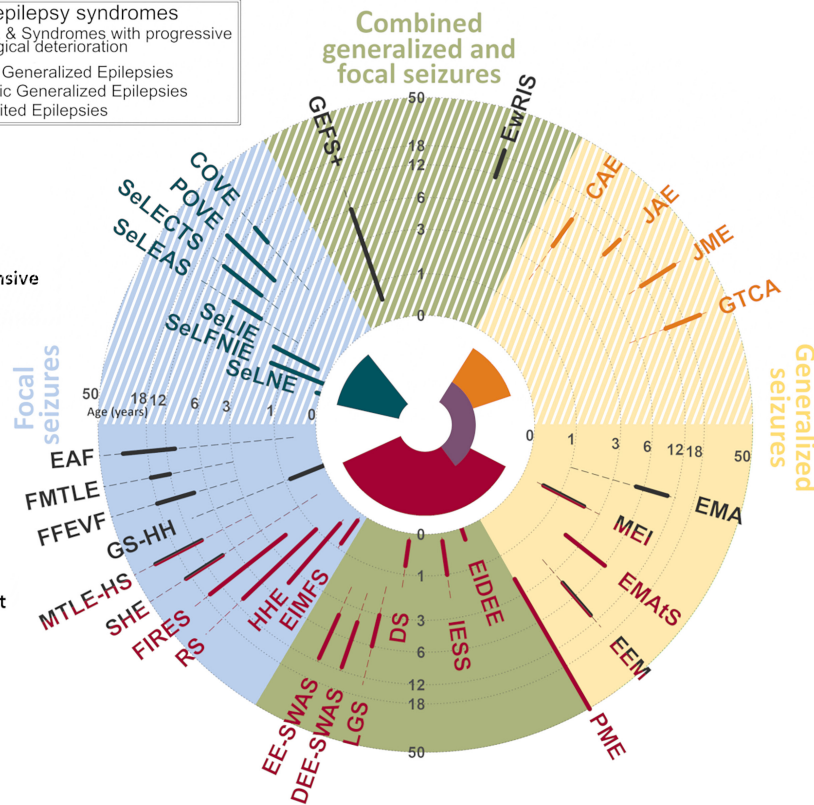


FIGURE 1 The epilepsy syndromes according to the 2017 epilepsy syndrome classification¹ and the 2022 report of the ILAE Task Force on Nosology and Definitions,²⁻⁶ using the full names of the different epilepsy syndromes (A) or their specific abbreviations (B). The different epilepsy syndromes are divided 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 pharmaco-responsive 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 parcelated 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 Imagine 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.

ORCID

Rima Nababout  <https://orcid.org/0000-0001-5877-4074>

Mathieu Kuchenbuch  <https://orcid.org/0000-0002-5944-2204>

Paolo Tinuper  <https://orcid.org/0000-0002-0588-3063>

J. Helen Cross  <https://orcid.org/0000-0001-7345-4829>

Elaine Wirrell  <https://orcid.org/0000-0003-3015-8282>

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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*. 2022;00:1–4. <https://doi.org/10.1002/epi4.12665>