



# تَشخيص وتصنيف التهاب العصب البصري

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Samedi 09-DEC-2023, 10:00-11:00  
Marrakesh, 35<sup>th</sup> International Neuroscience Days  
axel petzold



# Divulgations

NIHR UK, UCSF  
Stichting MS Research NL  
Novartis, Heidelberg Academy



# Contenu

- Contexte
- Procédure Delphi
- Comité
- Classification
- Critère diagnostique
- Résumé



# Hunain Ibn Is-Haq (808-873)

• *“Therefore we see that the vision has ceased or diminished without our finding any change in the pupil and there is a heaviness in the head and particularly its deep part and the parts surrounding the orbit. We know that the affection is caused by an abundant moisture which has run to the optic nerve of the eye and has compressed or swelled it . . . the argument for the obstruction of the nerve is if you shut one eye [the unaffected eye] and observe whether the pupil of the other is dilated.”*



# The Lancet

## THE LANCET

### Series and commissions



SERIES

### Optic Neuropathies

*The Lancet Neurology*

Published: September 22, 2022

## THE LANCET Neurology

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### Optic Neuropathies

Published: September 22, 2022

#### Executive Summary

Optic neuropathies can reflect a wide range of pathophysiologies, both acquired and inherited. This Series provides an update on the clinical, imaging, and laboratory findings that differentiate these disorders, allowing clinicians to focus their diagnostic studies and optimise treatments. Multimodality optic nerve imaging—including fundus photography, optical coherence tomography, and MRI—has greatly advanced the diagnosis and follow-up of patients with optic neuropathies. Also reviewed in this Series, new evidence shows that optic neuritis can frequently indicate autoimmune neurological disorders, including multiple sclerosis and the recently recognised disease categories of aquaporin-4 antibody-associated neuromyelitis optica spectrum disorder and myelin-oligodendrocyte glycoprotein antibody-associated disease. Early clinical recognition of optic neuritis is, therefore, important for prognosis and treatment. Also reviewed in the Series, a unifying feature in the pathophysiology of hereditary disorders of the optic nerve is mitochondrial dysfunction. Treatments are emerging for optic neuropathies, including immunotherapies and genetic therapies.

#### Series

##### Imaging of the optic nerve: technological advances and future prospects

Valérie Biousse, Helen V Danesh-Meyer, Amit M Salindane, Cédric Lamirel, Nancy J Newman  
*The Lancet Neurology*  
Published: September 22, 2022  
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##### Optic neuritis and autoimmune optic neuropathies: advances in diagnosis and treatment

Jeffrey L Bennett, Fiona Costello, John J Chen, Axel Petzold, Valérie Biousse, Nancy J Newman, Steven L Galetta  
*The Lancet Neurology*  
Published: September 22, 2022  
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##### Understanding the molecular basis and pathogenesis of hereditary optic neuropathies: towards improved diagnosis and management

Nancy J Newman, Patrick Yu-Wai-Man, Valérie Biousse, Valerio Carelli  
*The Lancet Neurology*  
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Axel Petzold, Clare L Fraser, Mathias Abegg, Raed Alroughani, Daniah Alshwaeir, Regina Alvarenga, and others  
*The Lancet Neurology*  
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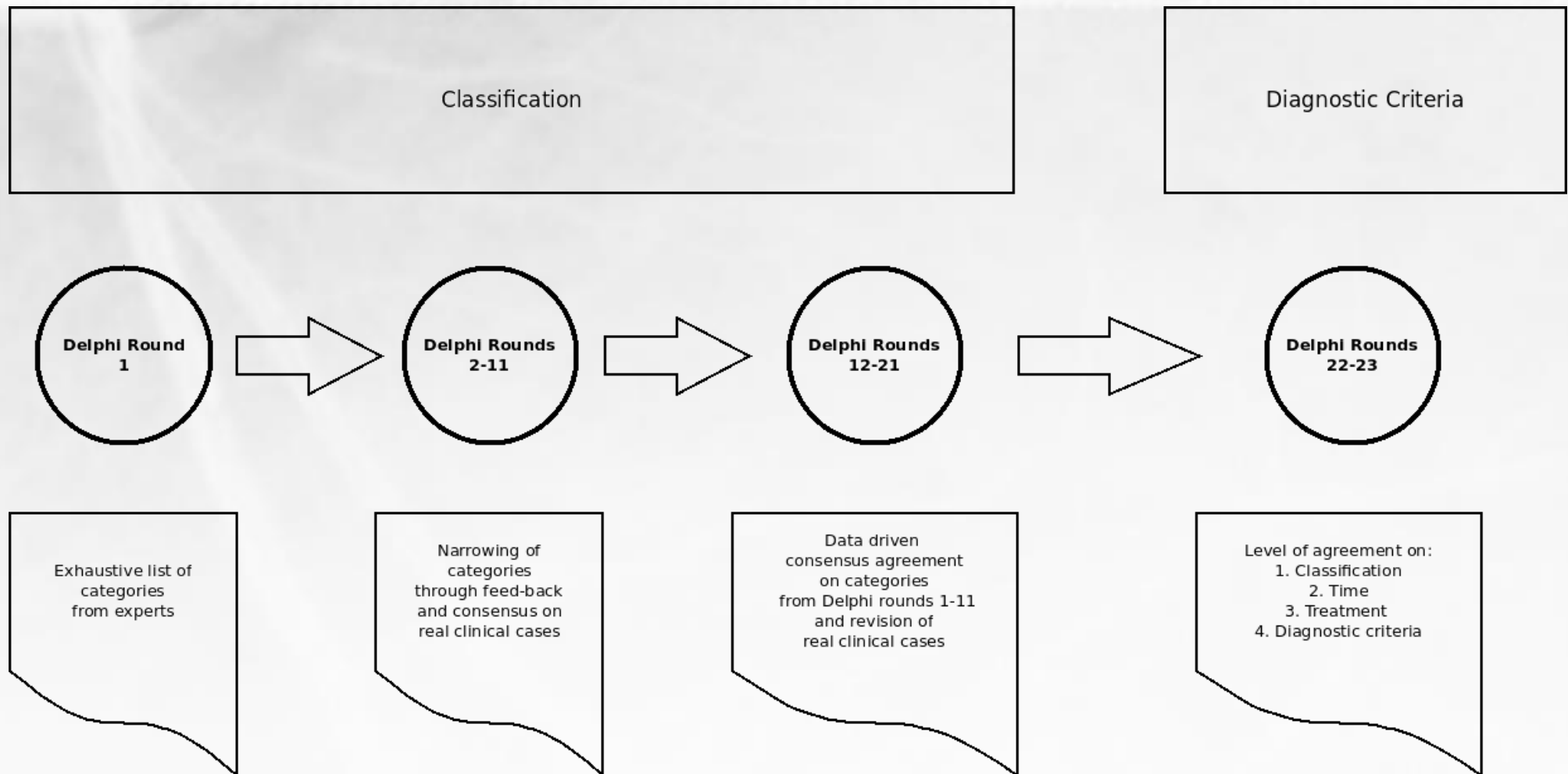


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# Procédure Delphi

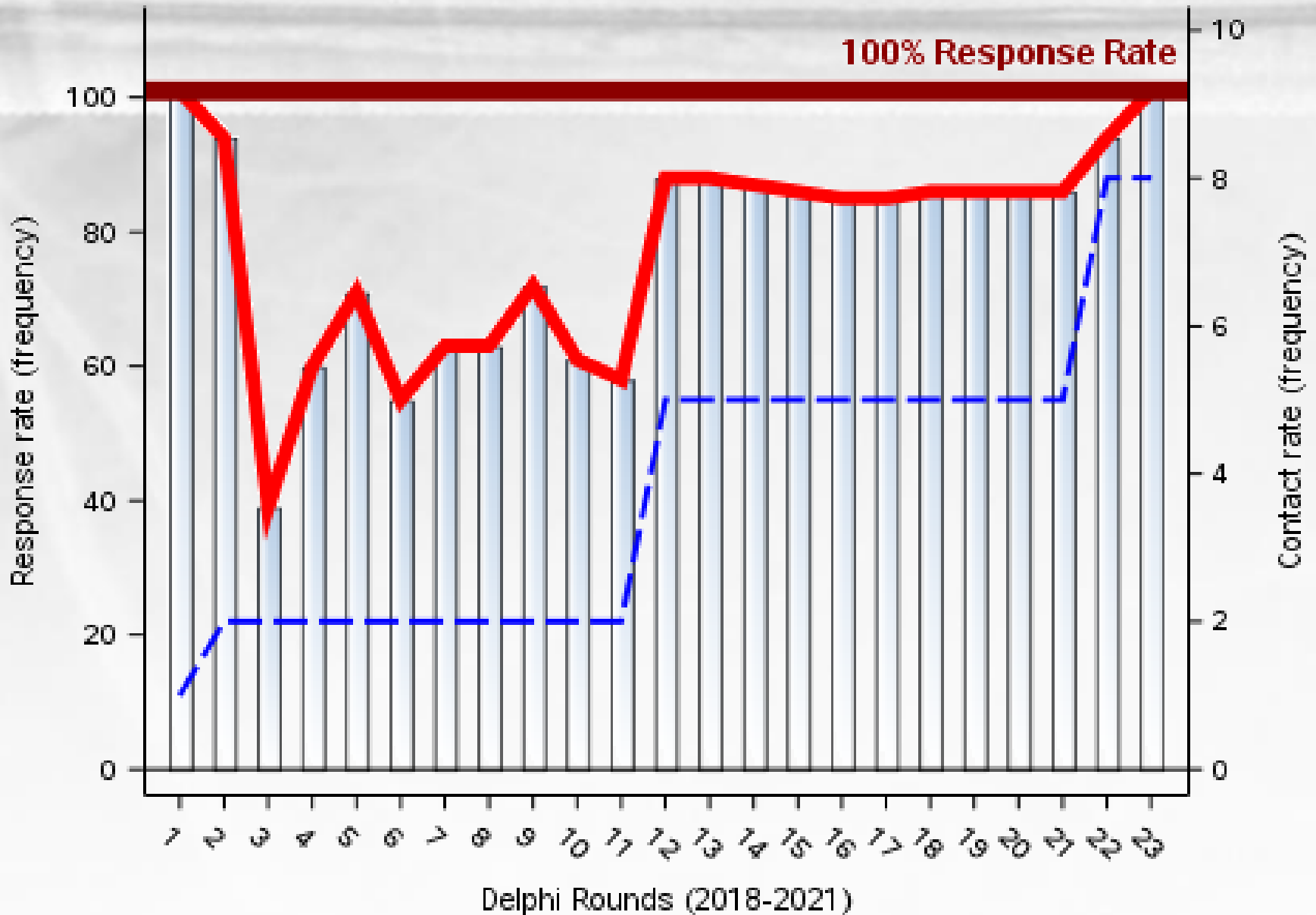


Définition du consensus

Expertisecentrum Neurologie >80% Amsterdam UMC



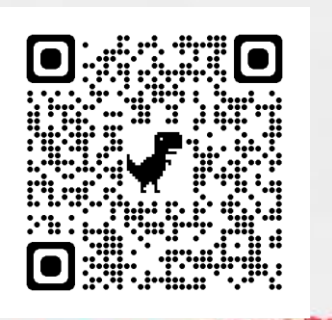
# Delphi (2018-2021)







# Cas clinique exemplaire



- A 50 year old man had developed a febrile illness with fatigue, anosmia and loss of taste. About 3 weeks later he experienced bilateral retrobulbar pain, worsening on eye movements with visual loss. Visual loss progressed over 7 days before he presented to Moorfields Eye Hospital. Visual acuities were reduced to RE hand movement and LE count finger. There was a right RAPD and bilateral swollen optic discs. He was treated with high dose steroids (1g/day i.v. for 5 days). Within 4 days his vision recovered to a VA of RE 6/9 and LE 6/5.

I have seen similar case(s) before / This is a new case for me

- His MRI (coronal T1 with contrast, Figure 29) showed contrast enhancement of both optic nerves and nerve sheets. There were no lesions typical for multiple sclerosis elsewhere in the brain.

This information changed my clinical working diagnosis /

This information confirmed my clinical working diagnosis

- A nasopharyngeal swab was positive for COVID (PCR). Routine blood tests were normal. He was seronegative for AQP4 and MOG. The CSF showed matched oligoclonal bands.

This makes a definite diagnosis /

This is non-specific

- I think this patient has a diagnosis of (please type your own answer here) [...]



Figure 29: Case 5 MRI.

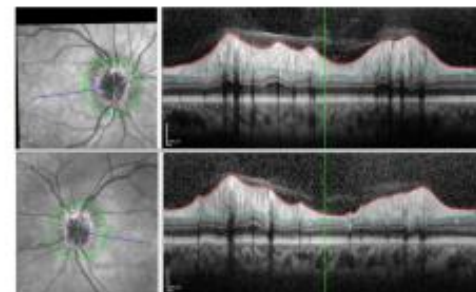


Figure 30: Case 5 OCT.

- Do you think the three week interval between onset of COVID and his bilateral optic neuritis is acceptable to strongly consider this to be a post-infectious optic neuritis?  yes/ no
- The bilateral optic disc swelling was documented by OCT (Figure 30).

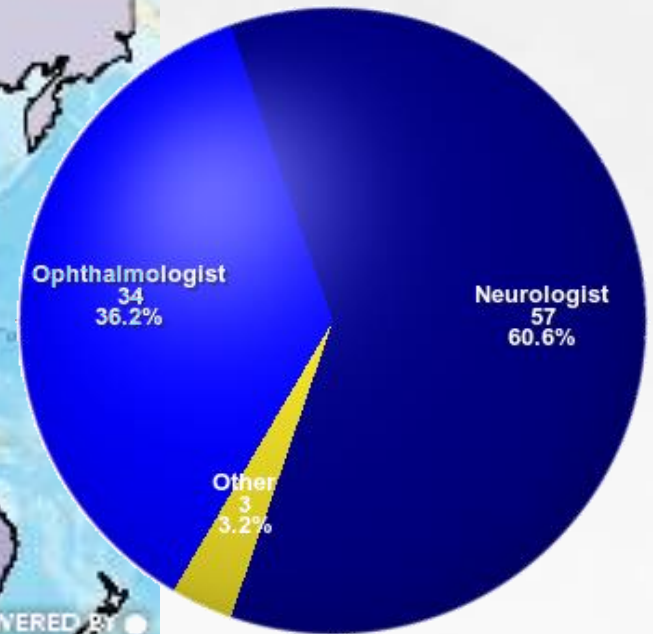
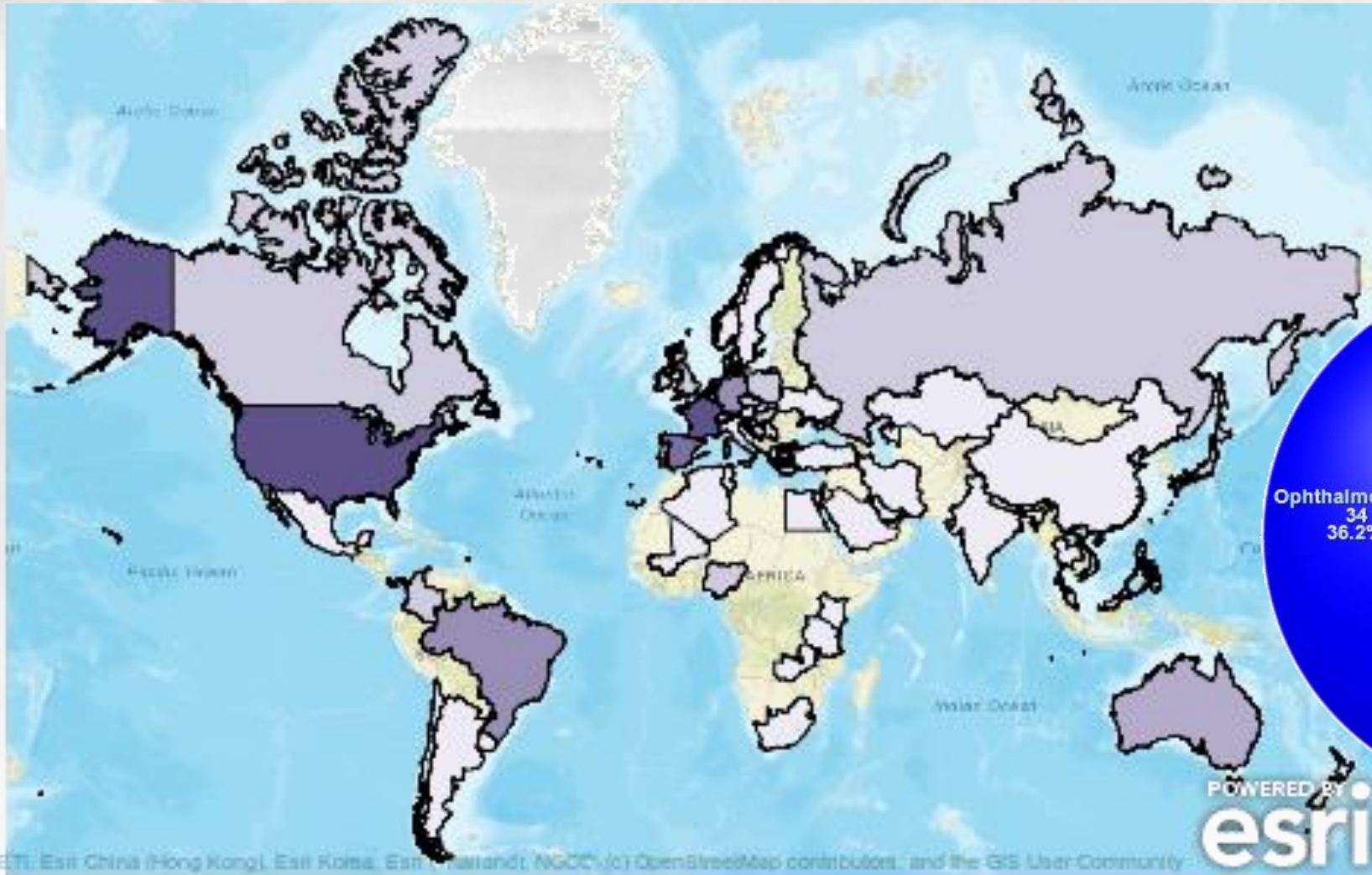


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# Commite





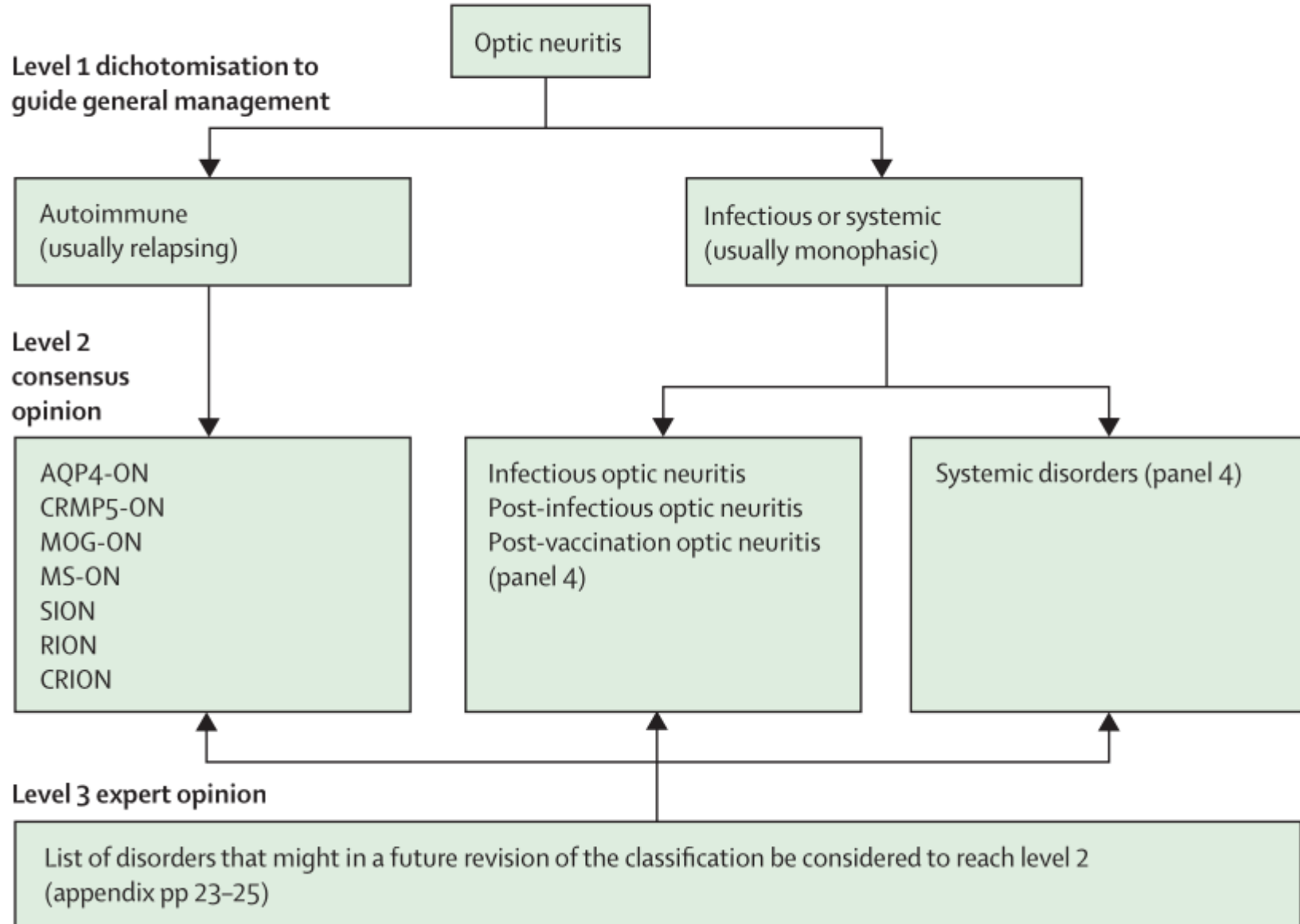
# Contenu

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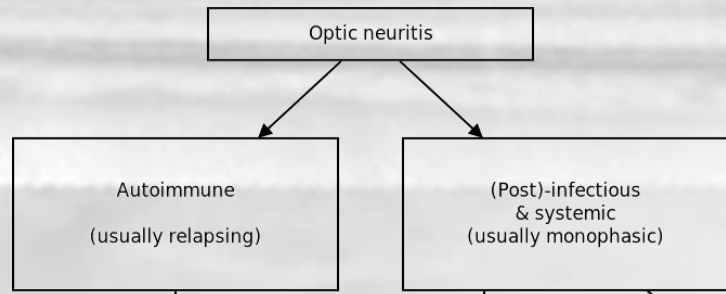
# Classification

## B Classification of optic neuritis





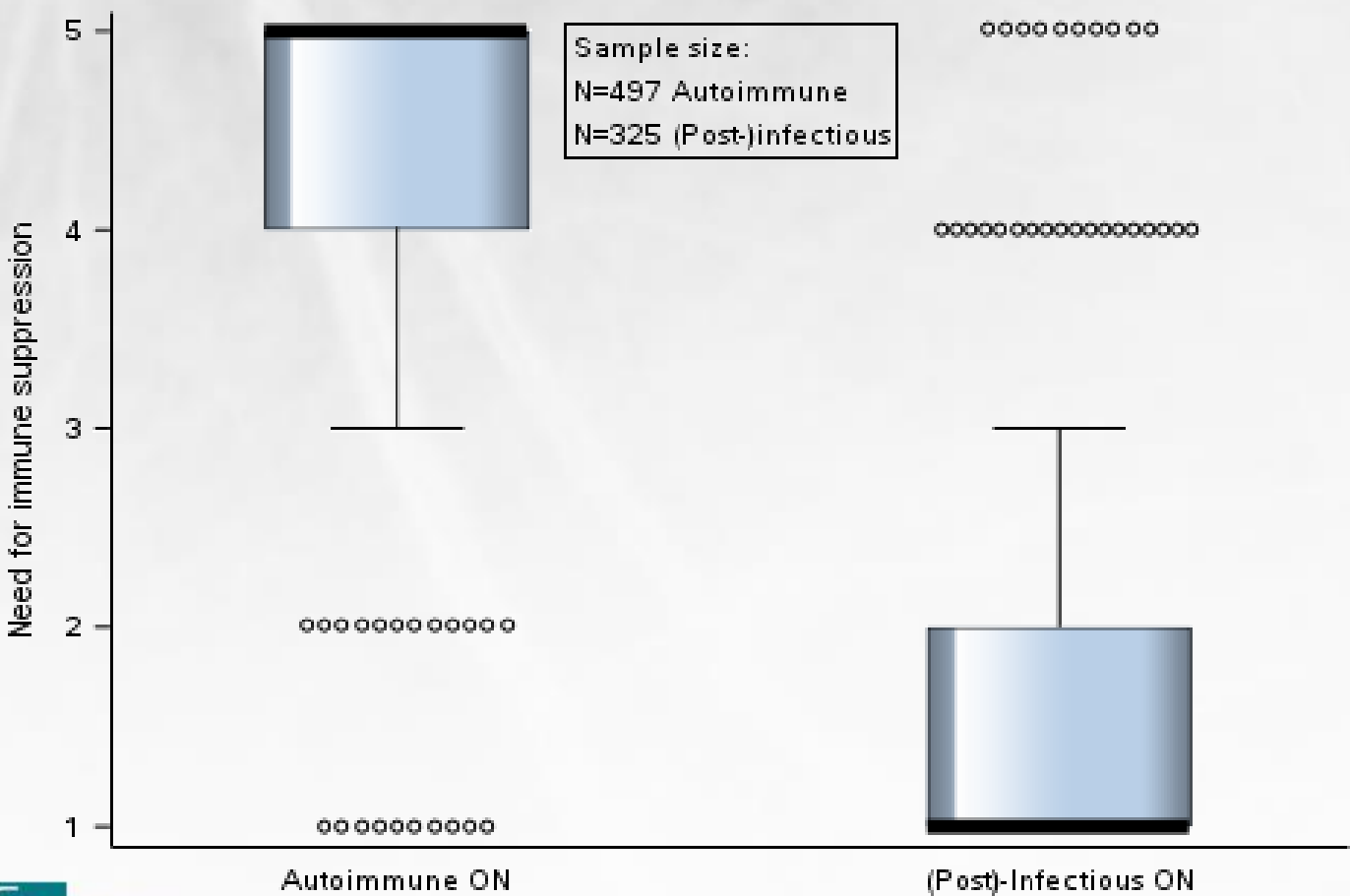
# Niveau 1 de la classification



Niveau 1: 95% consensus

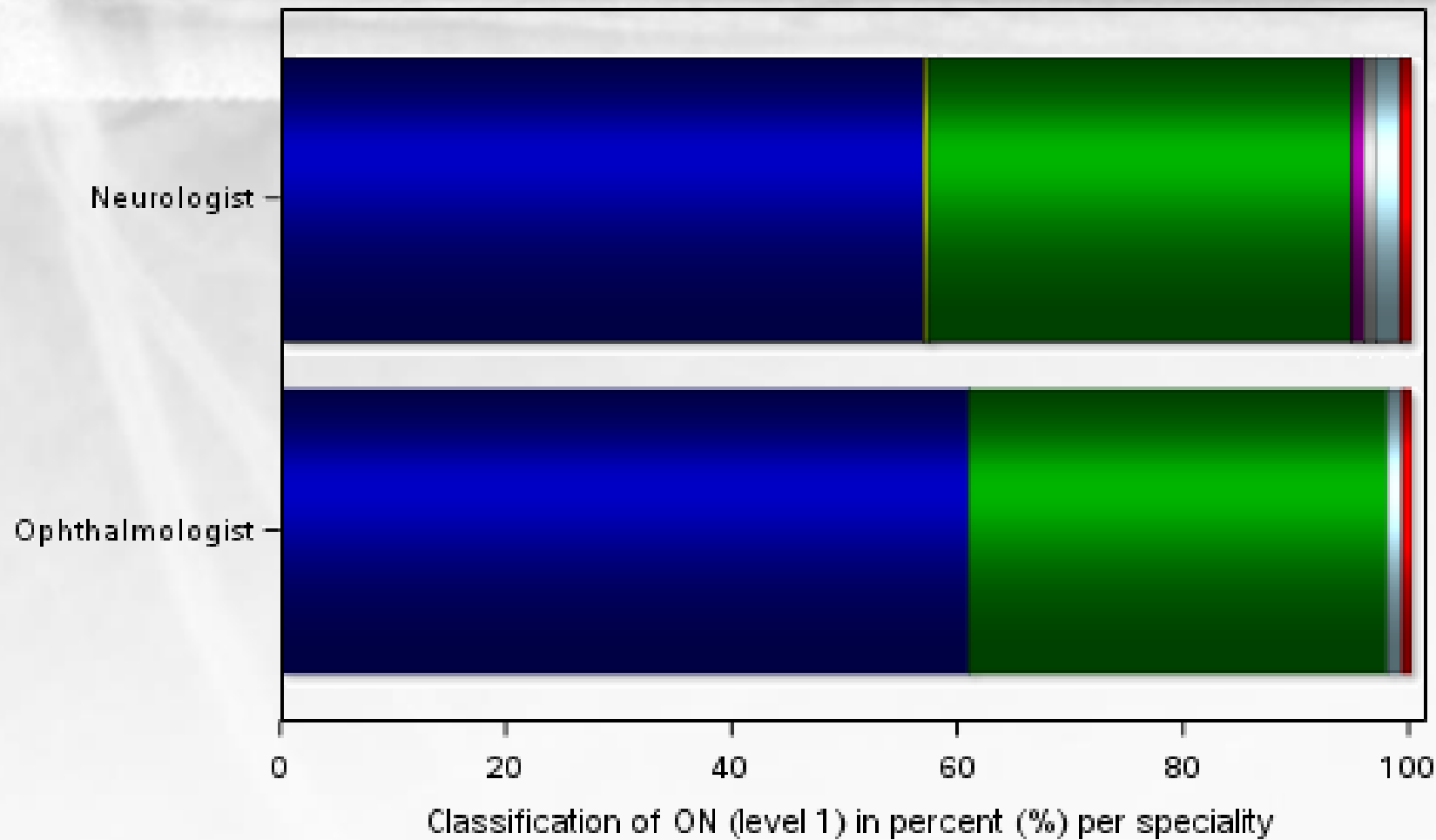
Les résultats de la procédure itérative Delphi ronde 2-21

Relevant pour la prise en charge des malades



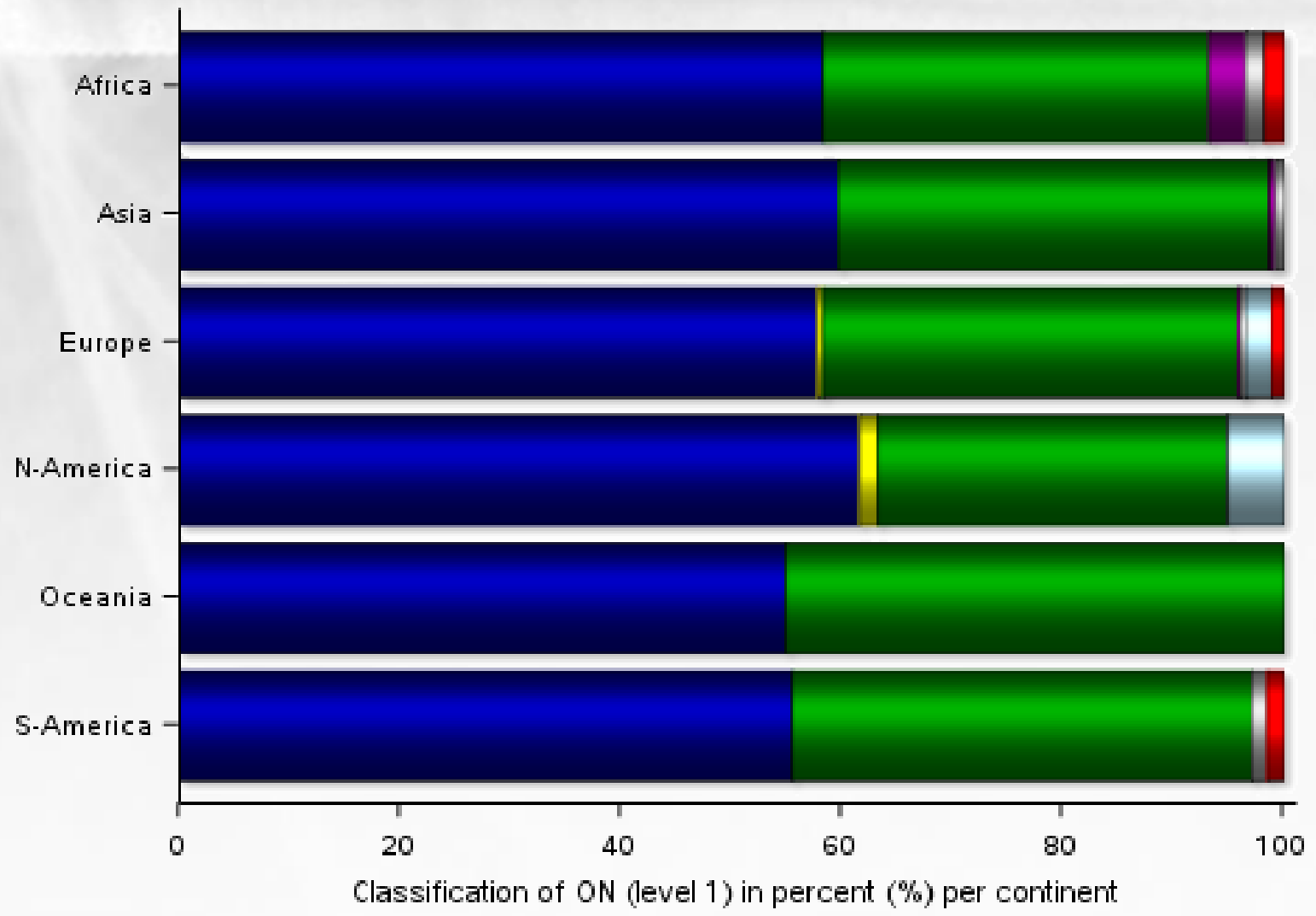


# Consensus: spécialiste





# Consensus: continental







# Clinique

## Panel 3: Signs and symptoms aiding the clinical classification of optic neuritis and exclusion of alternative pathologies

### Clinical presentation\*:

- Sequence of symptoms over time
- Preceding infection or vaccination
- Ethnic background or location
- Progression of pain or visual loss (>2 weeks)
- Absence of pain
- Associated epilepsy†
- Simultaneous bilateral ON
- Evidence of retinitis or retinal dysfunction from OCT or electrophysiology
- Presence of severe optic disc oedema
- Absence of optic disc oedema
- Unexplained optic atrophy in either eye at onset
- Fever or other systemic symptoms and signs‡
- Other focal neurological signs

### Disease course§:

- Progressive loss of vision
- Progressive retinal layer atrophy for more than 12 months
- Sequential bilateral optic neuritis
- Absence of spontaneous recovery (>3 months)
- Corticosteroid dependence

### Medical history:

- Medical history of cancer or diseases listed in panel 4
- Family history of a suspected hereditary optic neuropathy
- Family history of other mitochondrial cytopathy



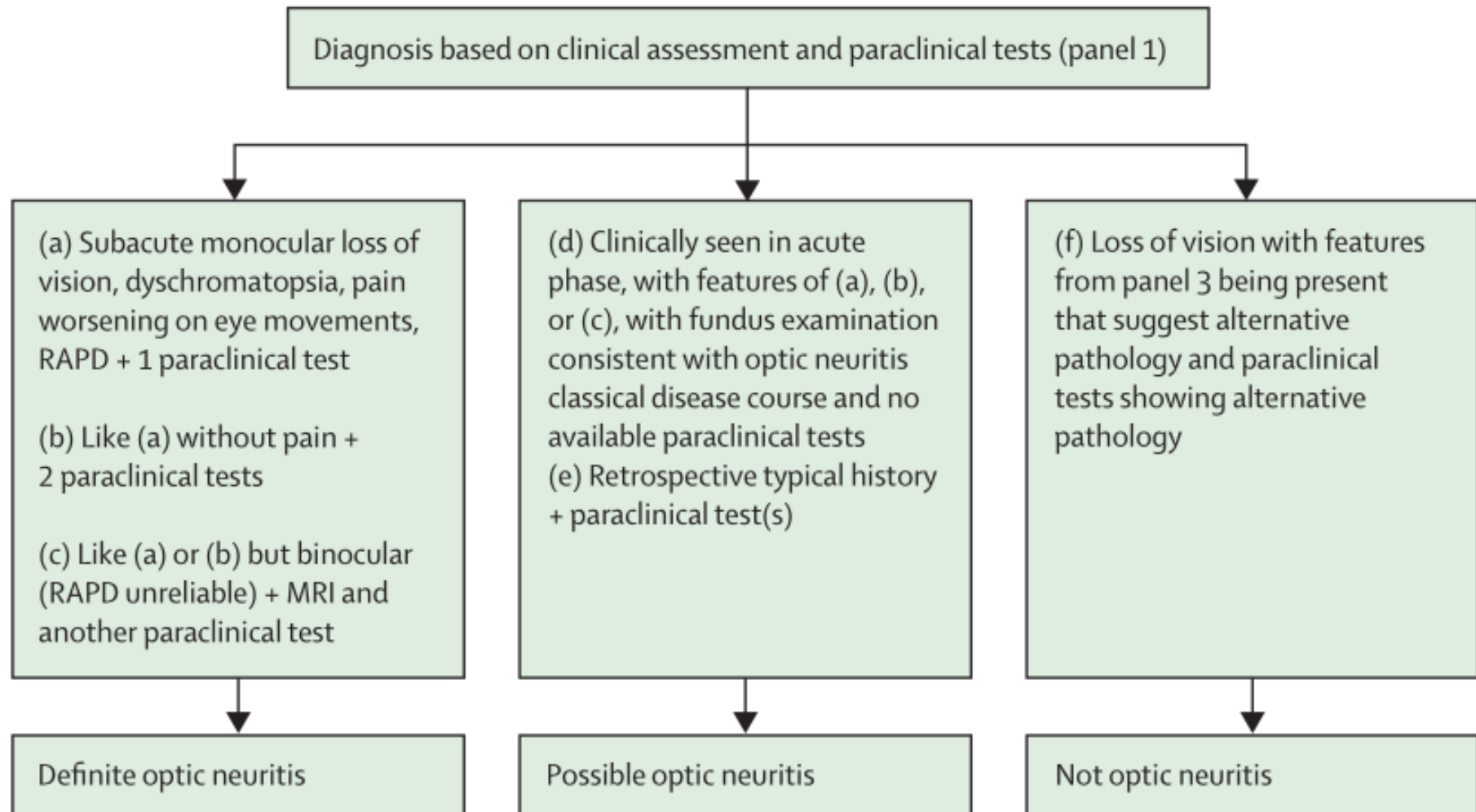
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# Diagnostique

## A Diagnosis of optic neuritis





## Panel 1: Diagnostic criteria for optic neuritis

### Clinical criteria

- A: Monocular, subacute loss of vision associated with orbital pain worsening on eye movements, reduced contrast and colour vision, and relative afferent pupillary deficit
- B: Painless with all other features of (A).
- C: Binocular loss of vision with all features of (A) or (B).

### Paraclinical criteria

- OCT: Corresponding optic disc swelling acutely or an inter-eye difference in the mGCIPL of  $>4\%$  or  $>4\mu\text{m}$  or in the pRNFL of  $>5\%$  or  $>5\mu\text{m}$  within 3 months after onset.
- MRI: Contrast enhancement of the symptomatic optic nerve and sheaths acutely or an intrinsic signal (looking brighter) increase within 3 months.
- Biomarker: AQP4, MOG, or CRMP5 antibody seropositive, or intrathecal CSF IgG (oligoclonal bands).

### Application of the clinical and paraclinical criteria

#### *Definite optic neuritis*

- (A) and one paraclinical test
- (B) and two paraclinical tests of different modality
- (C) and two different paraclinical tests of which one is MRI

#### *Possible optic neuritis*

- (A), (B), or (C) if seen acutely but in absence of paraclinical tests, with fundus examination typical for optic neuritis and consistent with the natural history during follow-up
- Positive paraclinical test or tests, with a medical history suggestive of optic neuritis



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# Résumé

- Névrite optique: développement d'une perspective internationale, après Hunain Ibn Is-Haq
- Critère nouvelles avec tomographie par cohérence optique (sensitivité 61-100%), IRM (sensitivité 22-44%), biomarqueur (spécificité >95%)
- Classification nouvelle que priorise la pratique clinique
- Nous avisons des révision future





Merci