SLCTRIMS
Sri Lanka Committee for Treatment and Research in Multiple Sclerosis and Related Disorders

ICON

European Charcot Foundation

The ICON 2022 Diagnostic Criteria for Optic Neuritis

SLCTRIMS 21-JAN-2024, 7:00-7:30, Symposium 4

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Disclosures

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

The ICON 2022 story

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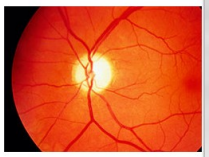
Optic Neuropathies

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 Saindane, 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
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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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Related Content

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Axel Petzold, Clare L Fraser, Mathias Abegg, Raed Alroughani, Daniah Alshowaeir, Regina Alvarenga, and others
The Lancet Neurology
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Romain Marignier, Yael Hacohen, Alvaro Cobo-Calvo, Anne-Katrin Pröbstel, Orhan Aktas, Harry Alexopoulos, and others
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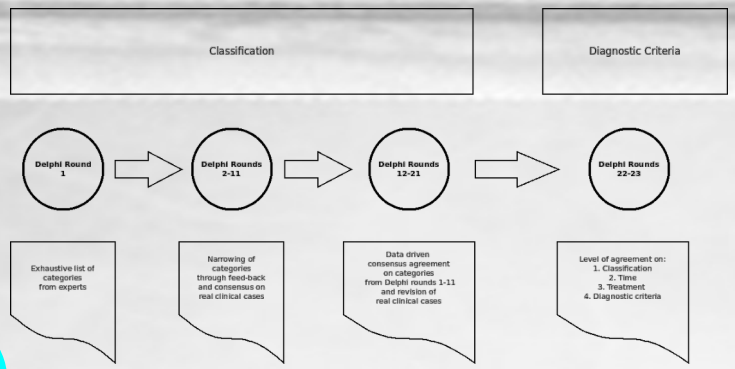
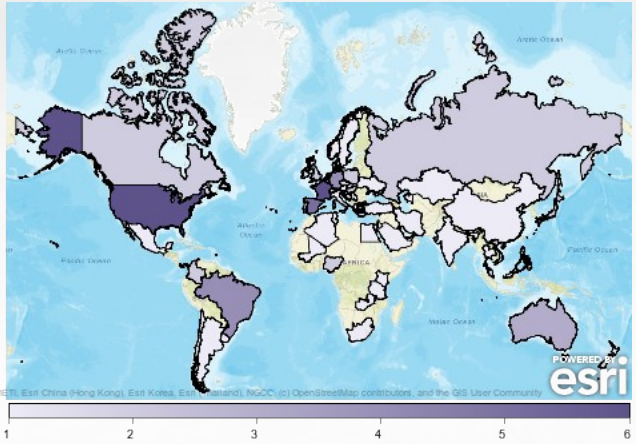
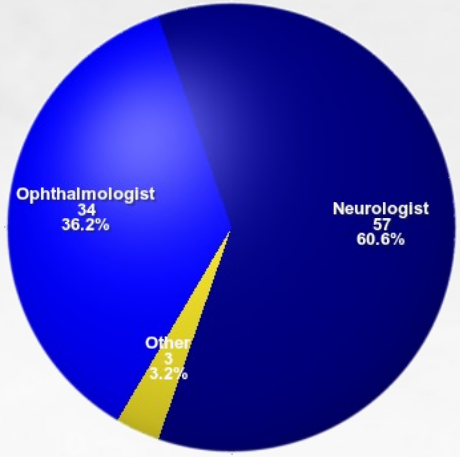
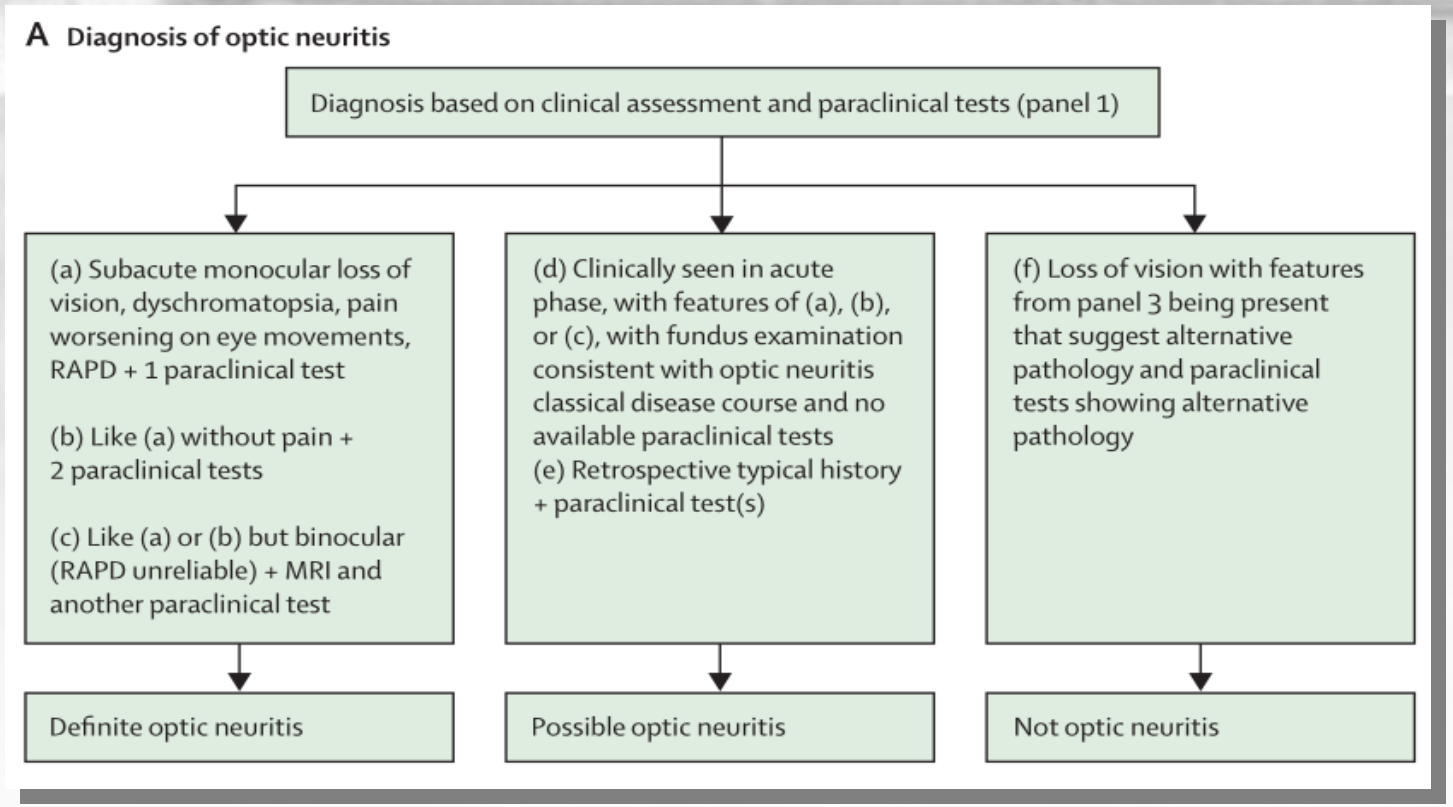




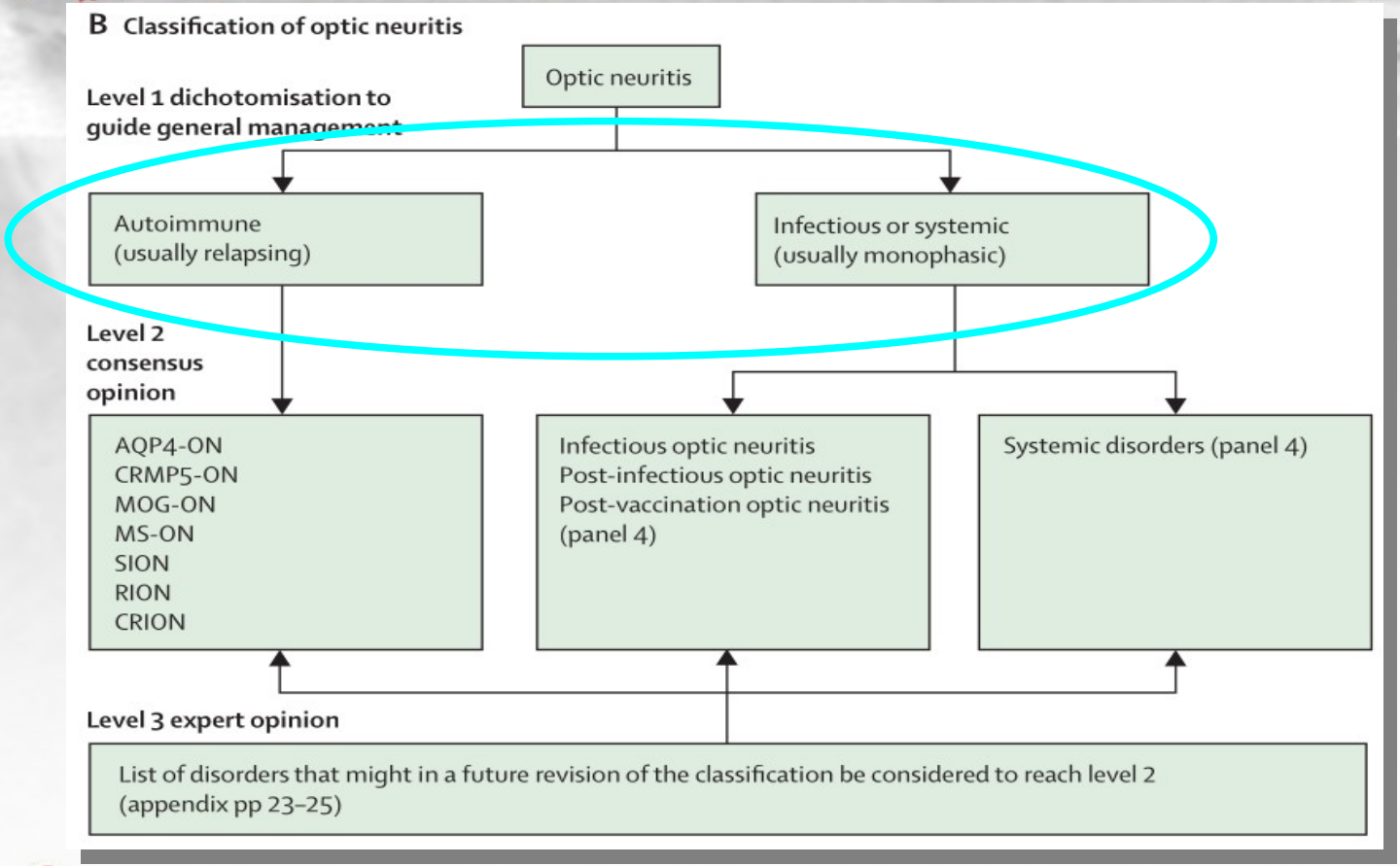


Definition of consensus
>80% expert agreement

ICON 2022 Diagnostic Criteria



ICON 2022 Classification



1st Case

- 34 year old Caucasian female patient
- 7 day history of pain in the right eye which worsens on eye movements
- Reduced colour vision
- VA RE: 6/9, left eye LE: 6/5
- Right RAPD
- Reports: fatigue, cognitive problems, urinary incontinence, depression
- PmHx: right sided numbness lasting 1m, 3y ago

1st Case

- Bloods all normal except for low Vitamin D at 22 nmol/L (normal 50-200 nmol/L)
- MRI: DIS & DIT
  three Gd+ non-symptomatic lesions
- CSF not done

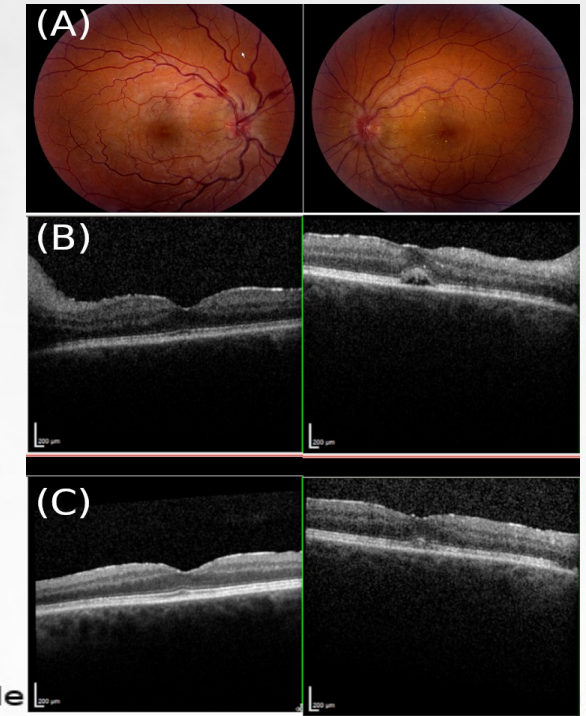
What is the most likely diagnosis?

2nd Case

- 28 year old, Afrocaribbean male
- Painless loss of vision LE (6/38)
- Dyschromatopsia
- L RAPD
- Several steroid responsive episodes over ~20 y fup
- OCT: pRNFL atrophy LE (IEPD >5%)
- MRI a swollen, Gd+, left optic nerve. No lesions elsewhere
- AQP4 seropositive

3rd Case

- 72 year old male develops febrile illness in Vietnam
- 2-3 weeks later bilateral, sequential, painless loss of vision (PL)
- no RAPD (but both pupils constrict with accomodation)
- Fundus (next slide):
  - Bilateral disc edema
  - RE hemorrhages
  - LE macular scar, CMO
- No recovery @ 6m fup (IVMP given ~6w after onset)



3 clinical scenarios of increasing complexity

- Case 1: is this MS ?
  Scenario A: painful, monocular, subacute LOV, dyschromatopsia, RAPD
- Case 2: is this NMO ?
  Scenario B: no pain, monocular, subacute LOV, dyschromatopsia, RAPD
- Case 3: what is this ?
  Scenario C: binocular, subacute LOV, dyschromatopsia, no pain, no RAPD

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 µm or in the pRNFL of >5% or >5 µm 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).

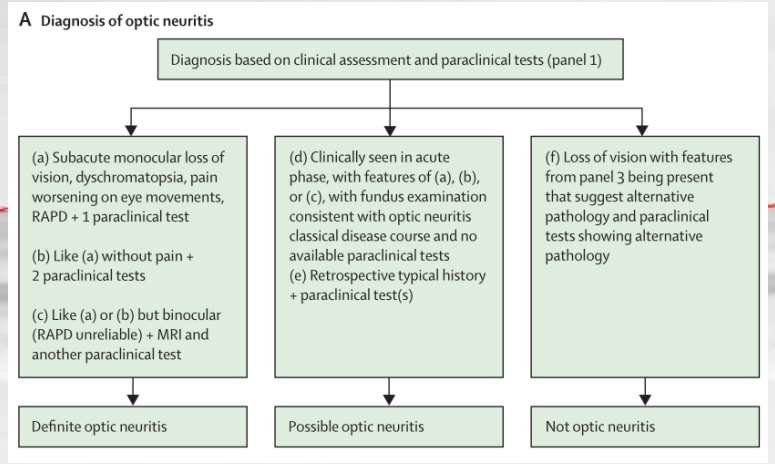
Application of the clinical and paraclinical criteria

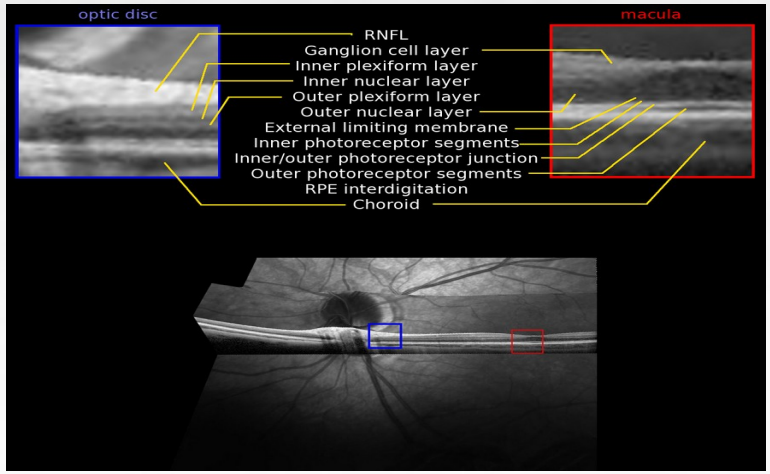
Definite optic neuritis

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

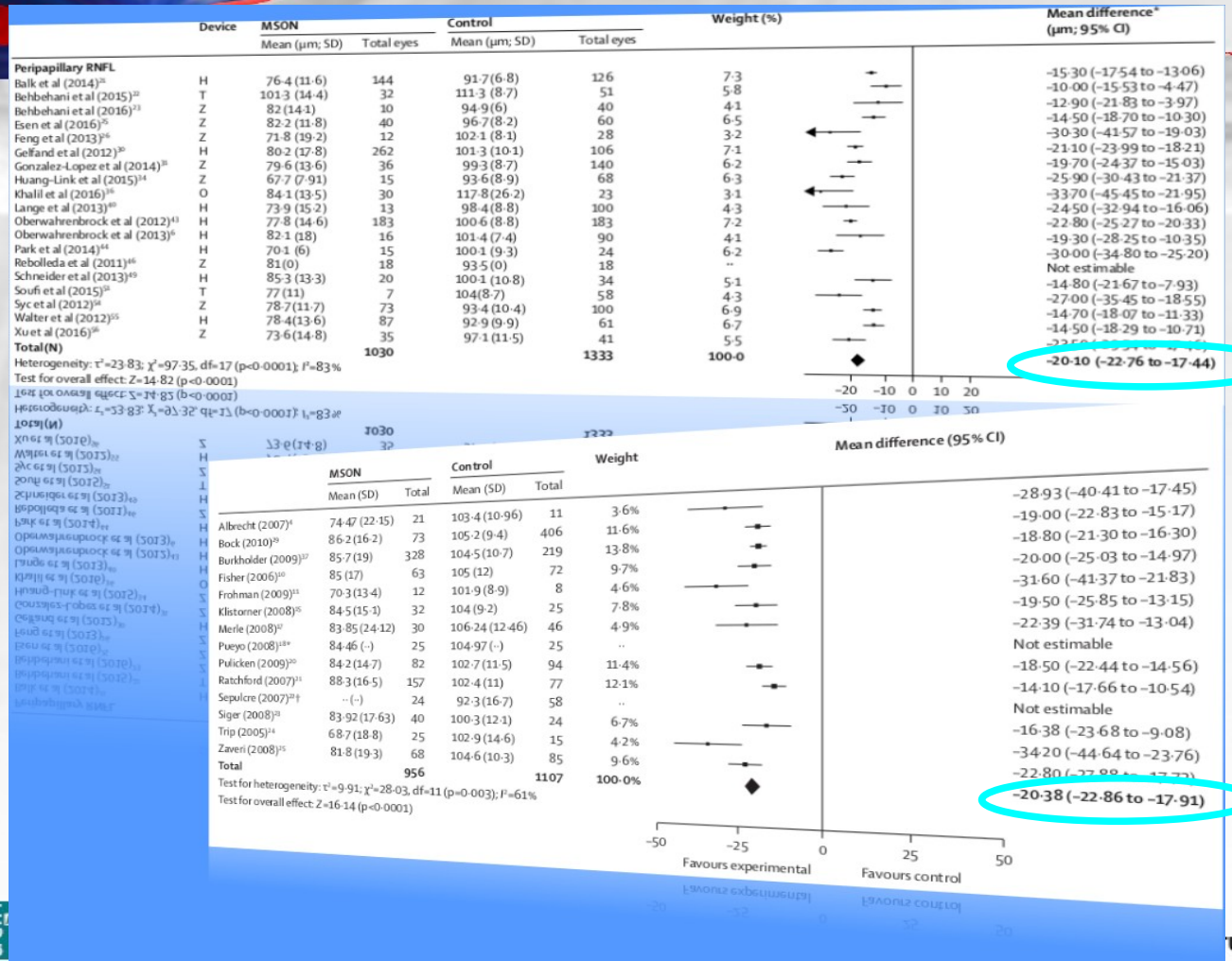
Possible optic neuritis

- (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





OCT in MS-ON



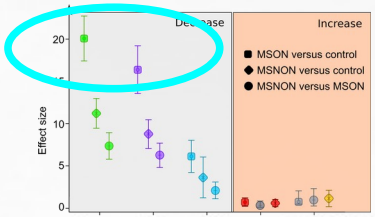
pRNFL atrophy

TLN 2010

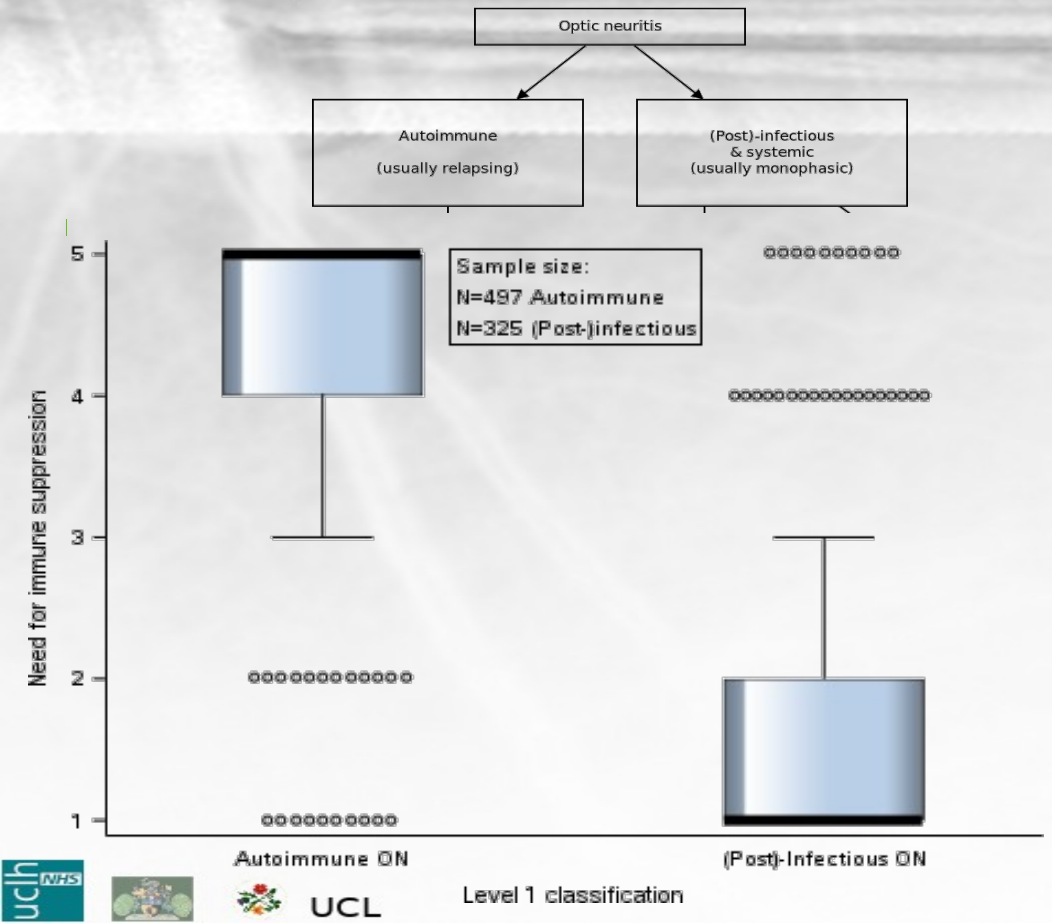
20.10 (17.44-22.76) µm

TLN 2017

20.38 (17.91-22.86) µm



How did we get there?

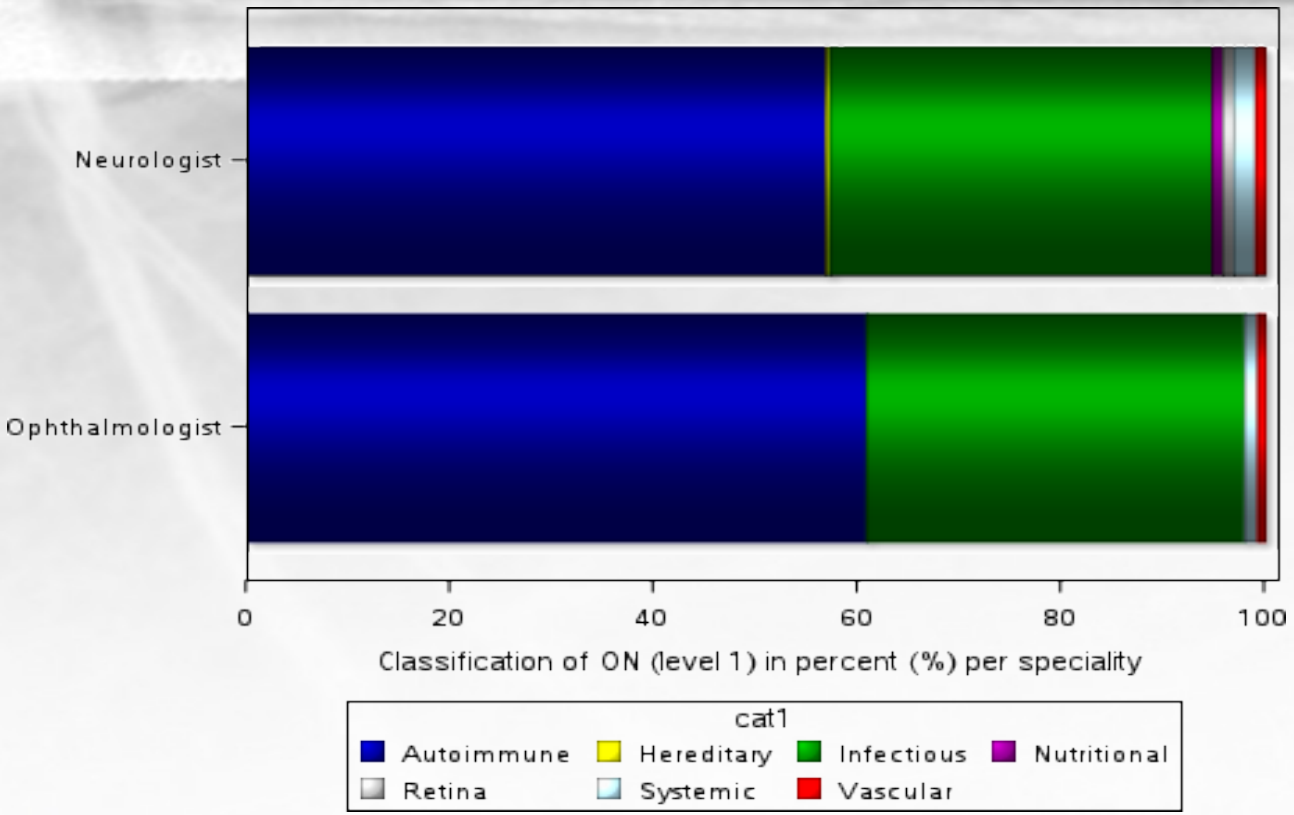


Level 1: 95% agreement

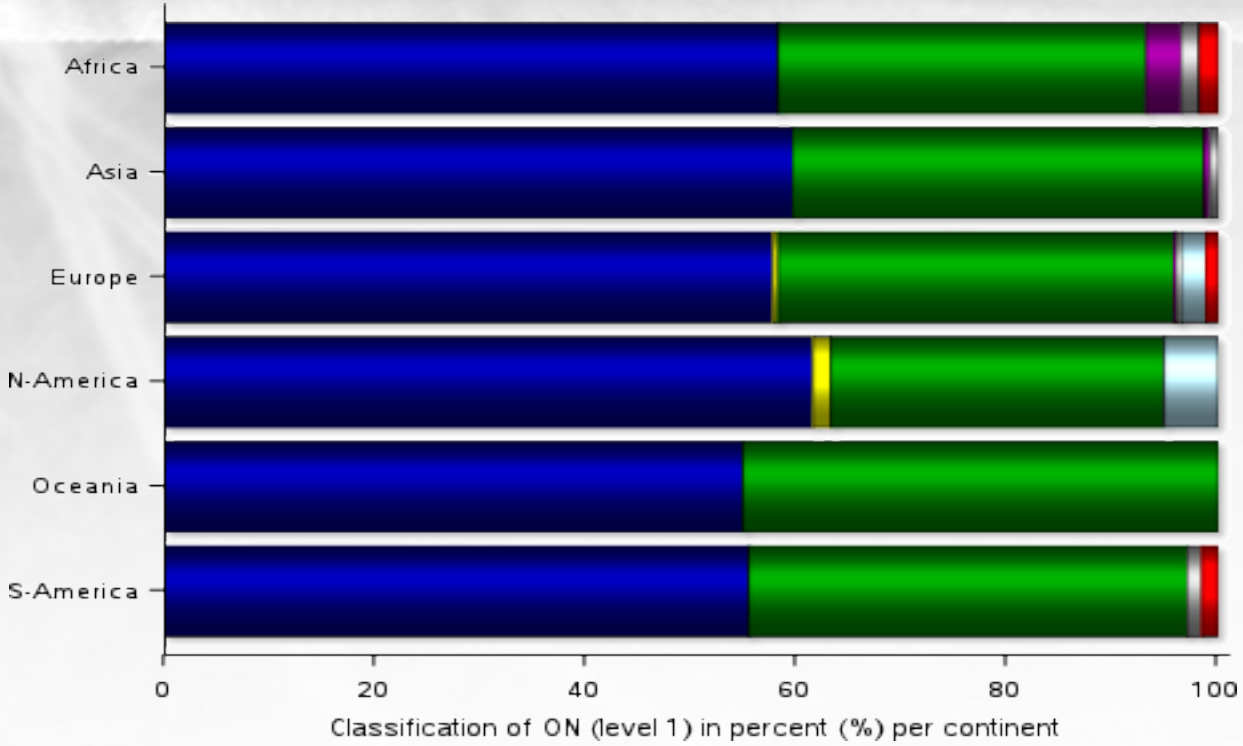
Based on iterative assessments from Delphi rounds 2-21

Relevant for patient management

Agreement: Speciality



Agreement: Continent



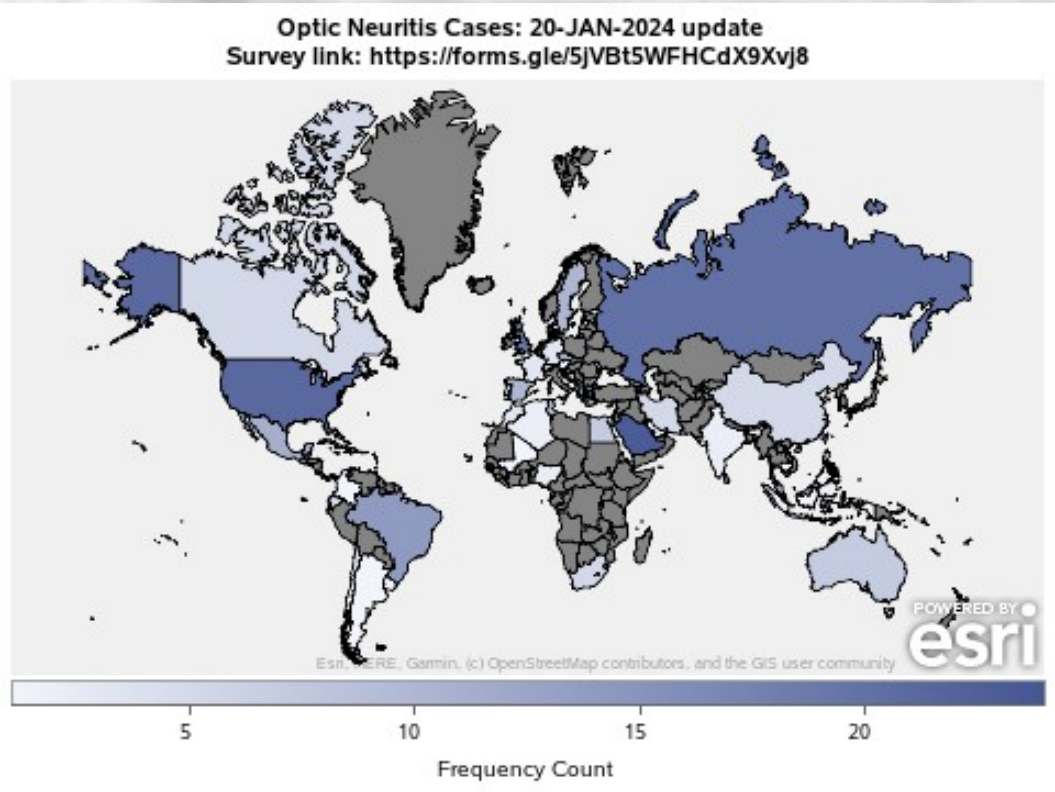
Cases summary

- Case 1: **MS-ON**
  Scenario A: painful, monocular, subacute LOV, dyschromatopsia, RAPD
- Case 2: **NMO-ON**
  Scenario B: no pain, monocular, subacute LOV, dyschromatopsia, RAPD
- Case 3: **post-infectious ON (Dengue)**
  Scenario C: binocular, subacute LOV, dyschromatopsia, no pain, no RAPD

Overall summary

- Optic Neuritis: Clinical approach
- ICON 2022 Diagnostic criteria
- ICON 2022 Classification
- Future revisions planned to optimise diagnostic sensitivity and specificity

Thank you - Q&A


Optic Neuritis Cases: 20-JAN-2024 update
Survey link: https://forms.gle/5jVBt5WFHCdX9Xvj8
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5
10
15
20
Frequency Count