Overview and classification of Optic Neuritis

axel petzold

Session: TC09 Diagnostic approach to optic neuritis
30-JUN-2024 15:35-16:15

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Disclosures

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Novartis, Roche, Heidelberg Academy

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DOI:https://doi.org/10.1016/S1474-4422(22)00200-9

Optic Neuritis

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
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The Lancet Neurology
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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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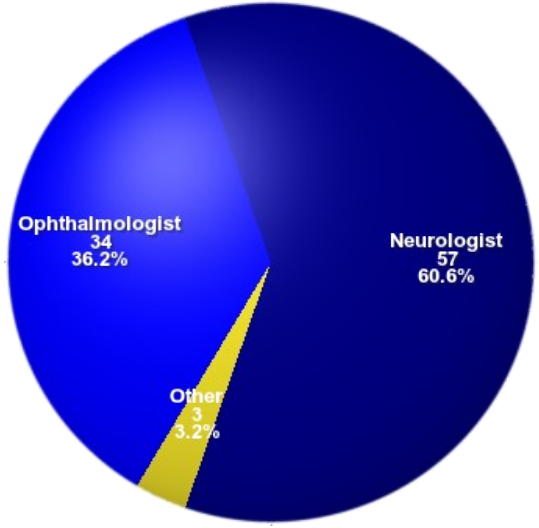
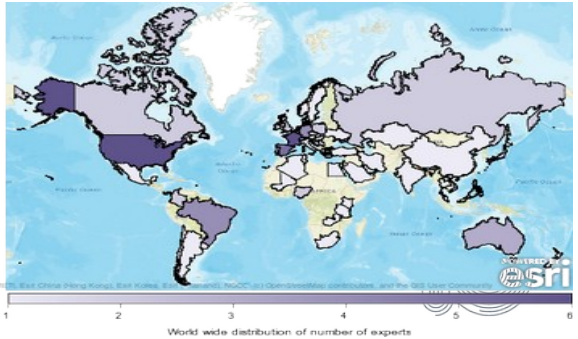
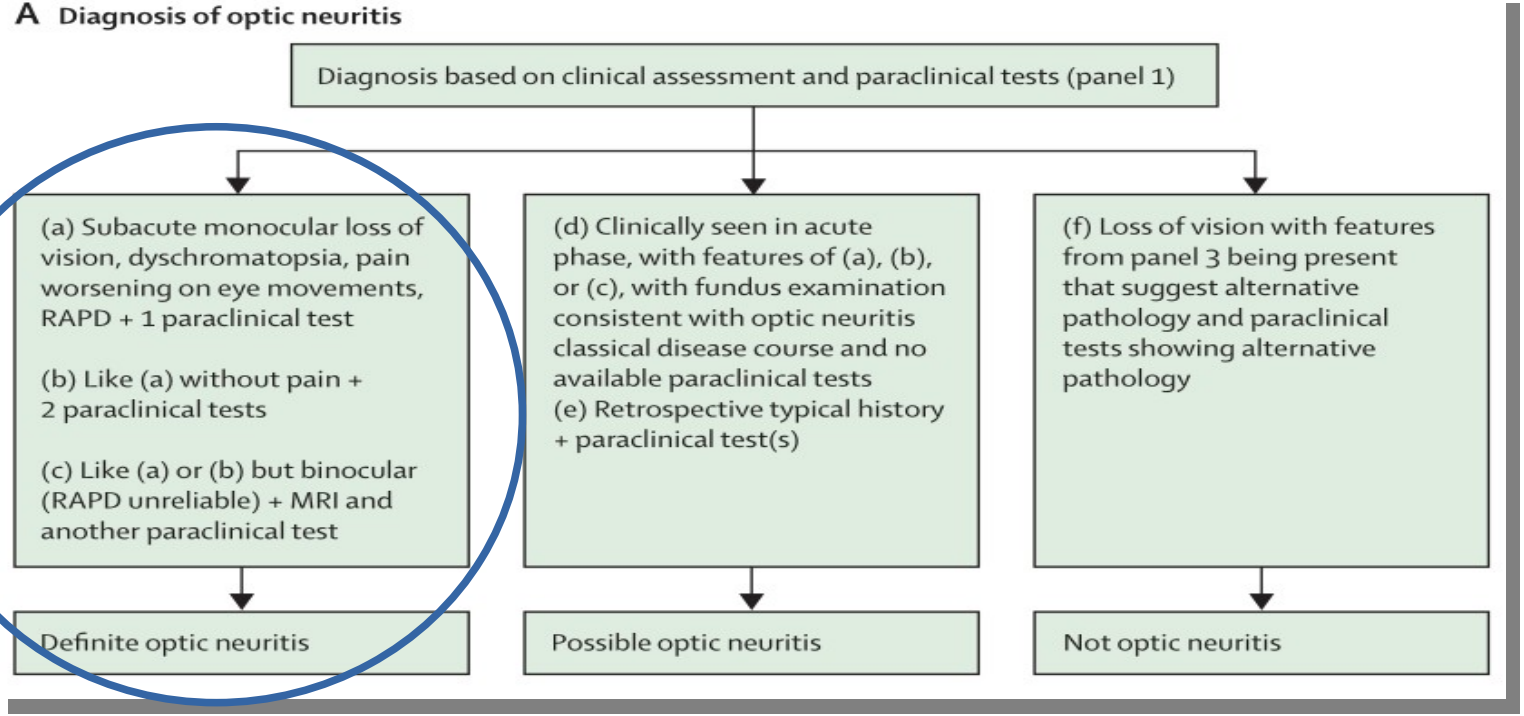


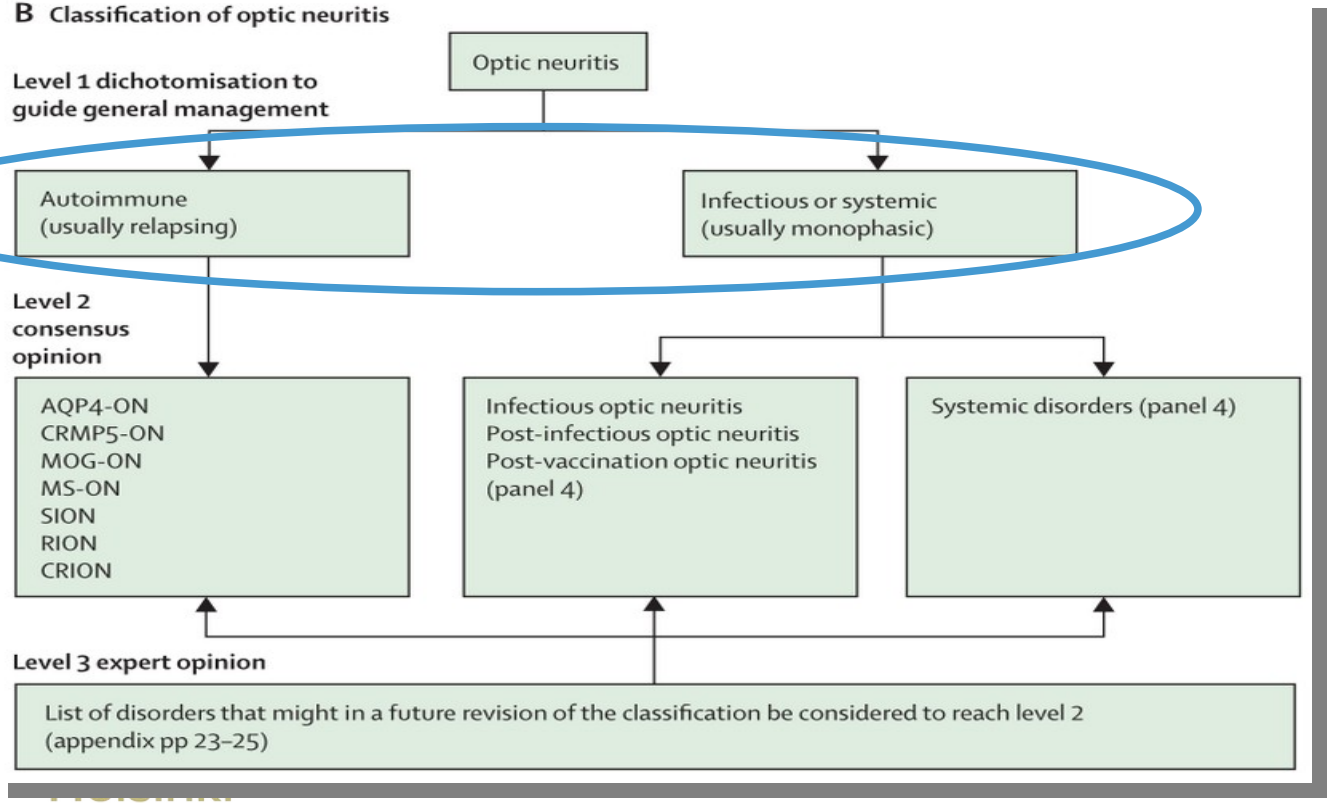


World wide distribution of number of experts

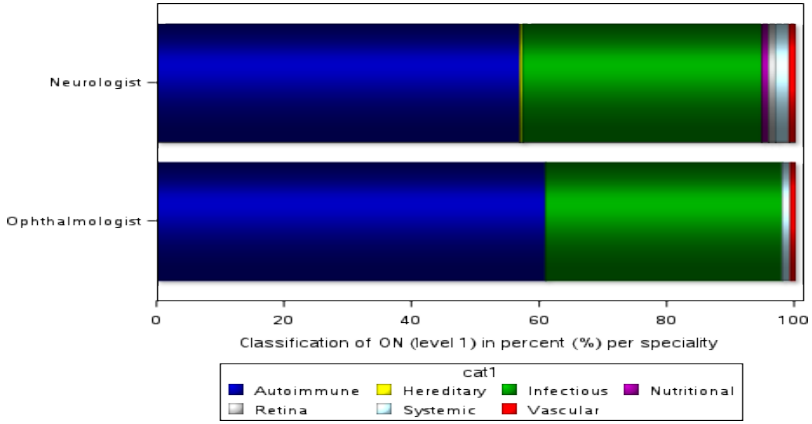
Diagnostic Criteria

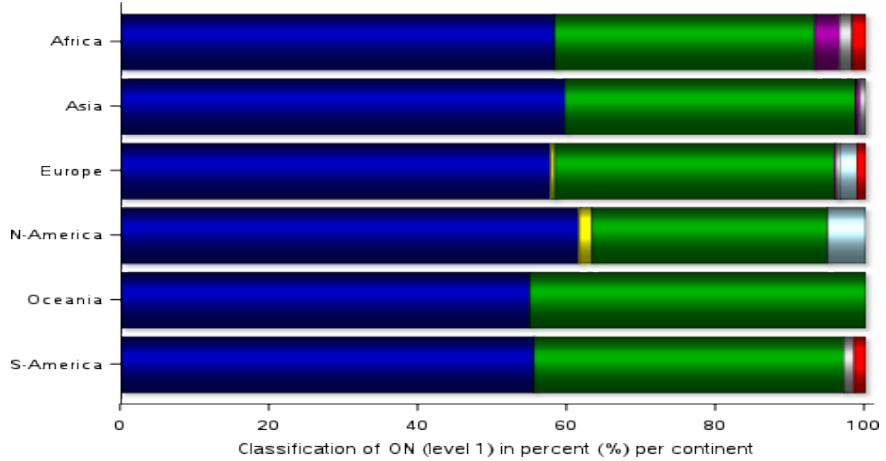


Classification



Consensus


Neurologist
Ophthalmologist
0
20
40
60
80
100
Classification of ON (level 1) in percent (%) per speciality
cat1
Autoimmune
Hereditary
Infectious
Nutritional
Retina
Systemic
Vascular


Africa
Asia
Europe
N-America
Oceania
S-America
0
20
40
60
80
100
Classification of ON (level 1) in percent (%) per continent

Please scan QR

for 3 teaching cases

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1st Case

- 34y old Caucasian female patient
- 7d RE pain, worsening on eye movements
- Dyschromatopsia & VA RE 6/9, LE 6/5, Right RAPD
- Reports: fatigue, cognitive problems, urinary incontinence, depression
- PmHx: right sided numbness lasting 1m, 3y ago
- MRI: DIS & DIT & 3 Gd+ non-symptomatic lesions

10 ICON cases

This survey aims to gather data on optic neuritis cases globally, tracking over three months from their initial presentation. We request participating experts to review 10 consecutive cases, ideally 3 months after onset, adhering to the ICON 2022 diagnostic criteria, detailed in the subsequent flowchart. Experts should respond to 10 specific questions for each reviewed case. The survey is conveniently accessible and can be completed using a smartphone. The survey will remain open until December 31, 2024, and we prefer data from **10 prospectively collected cases** using the ICON 2022 Criteria.

By participating, you agree to allow the secure storage of your email address as per GDPR guidelines. We will contact you for a collaborative publication using the study name from The Lancet Neurology "ICON".

You can access the ICON 2022 diagnostic criteria in various languages and there is a teaching video.

* Indicates required question

Email *

Your email address

Diagnosis based on clinical assessment & paraclinical tests (OCT, MRI or biomarker)

Definite ON:
(a) Subacute monocular loss of vision, dyschromatopsia, pain worsening on eye movements, RAPD + 1 paraclinical test

(b) Like (a) without pain, + 2 paraclinical tests

(c) Like (a) or (b) but binocular (RAPD unreliable) + MRI and another test

Possible ON:
(d) Clinically seen in acute phase, with features of (a)-(c) fundus examination consistent with ON classical disease course no paraclinical test(s)

(e) Retrospective typical history + paraclinical test(s)

- ◯ Definite ON
- ◯ Possible ON

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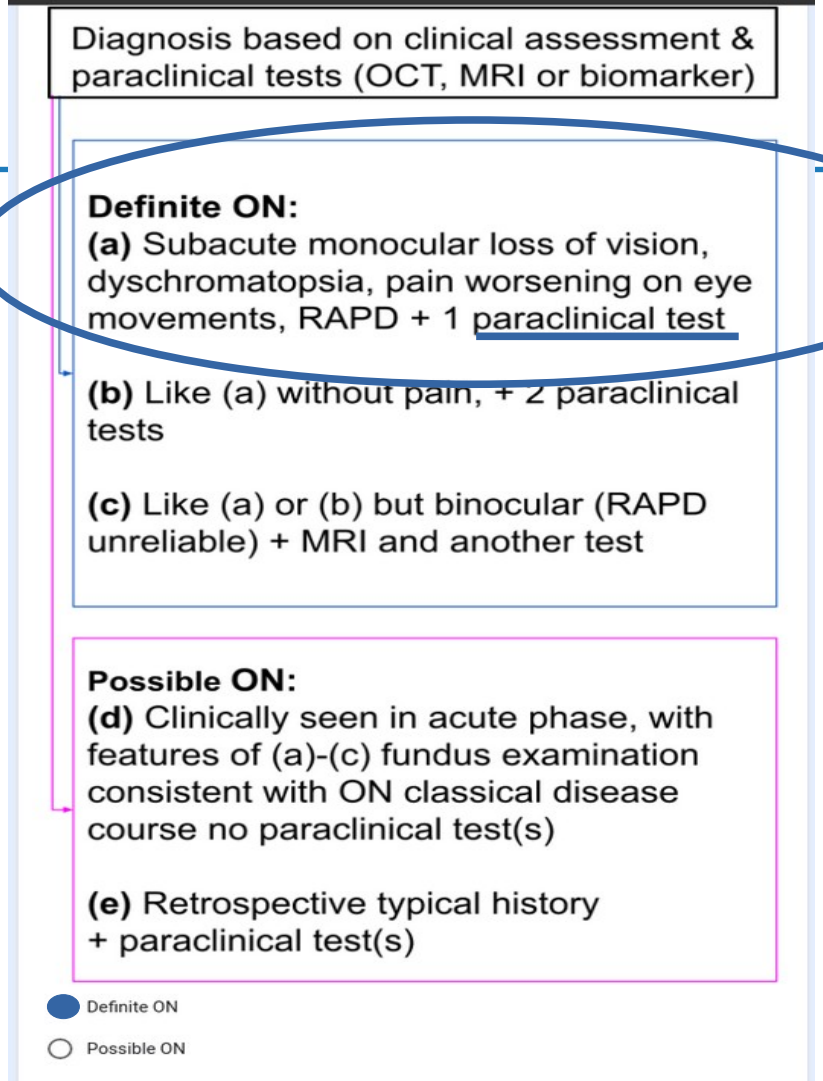


1st Case

Definite ON

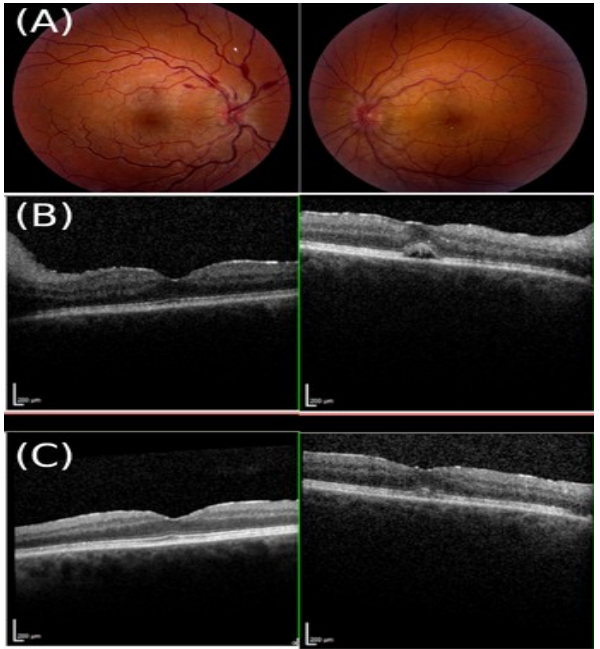
MRI: DIS & DIT

2nd Case

- 28y old, Afro Caribbean male
- Painless loss of vision LE (6/38), dyschromatopsia
- L RAPD
- Several steroid responsive episodes over ~20y fup
- OCT: pRNFL atrophy LE (IEPD >5%)
- MRI a swollen, Gd+, left optic nerve, brain & cord normal
- AQP4 seropositive

3rd Case

- 72 year old male, 2-3 weeks after febrile illness
- Bilateral, painless loss of vision (PL)
- No RAPD
- Fundus & OCT
- MRI shows bilateral ON, nil else
- IVMP given ~6weeks later
- No recovery ov vision @ 6m fup



3 Clinical scenarios

- 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, RAPD unreliable

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).

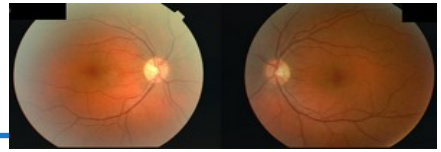
Application of the clinical and paraclinical criteria

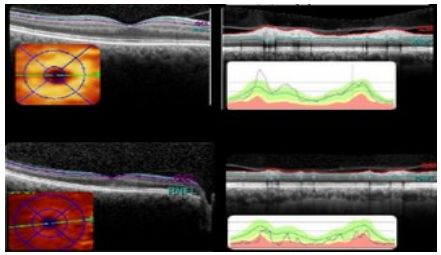
Definite optic neuritis

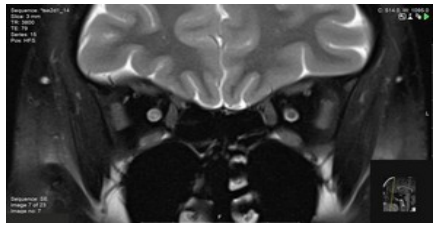
- (A) and one paraclinical test
- (B) and two paraclinical tests of different modality
- (C) and two different paraclinical tests of which one is MRI

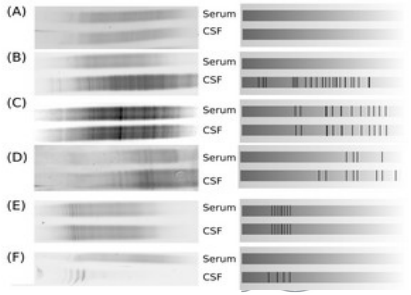
Possible optic neuritis

- (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

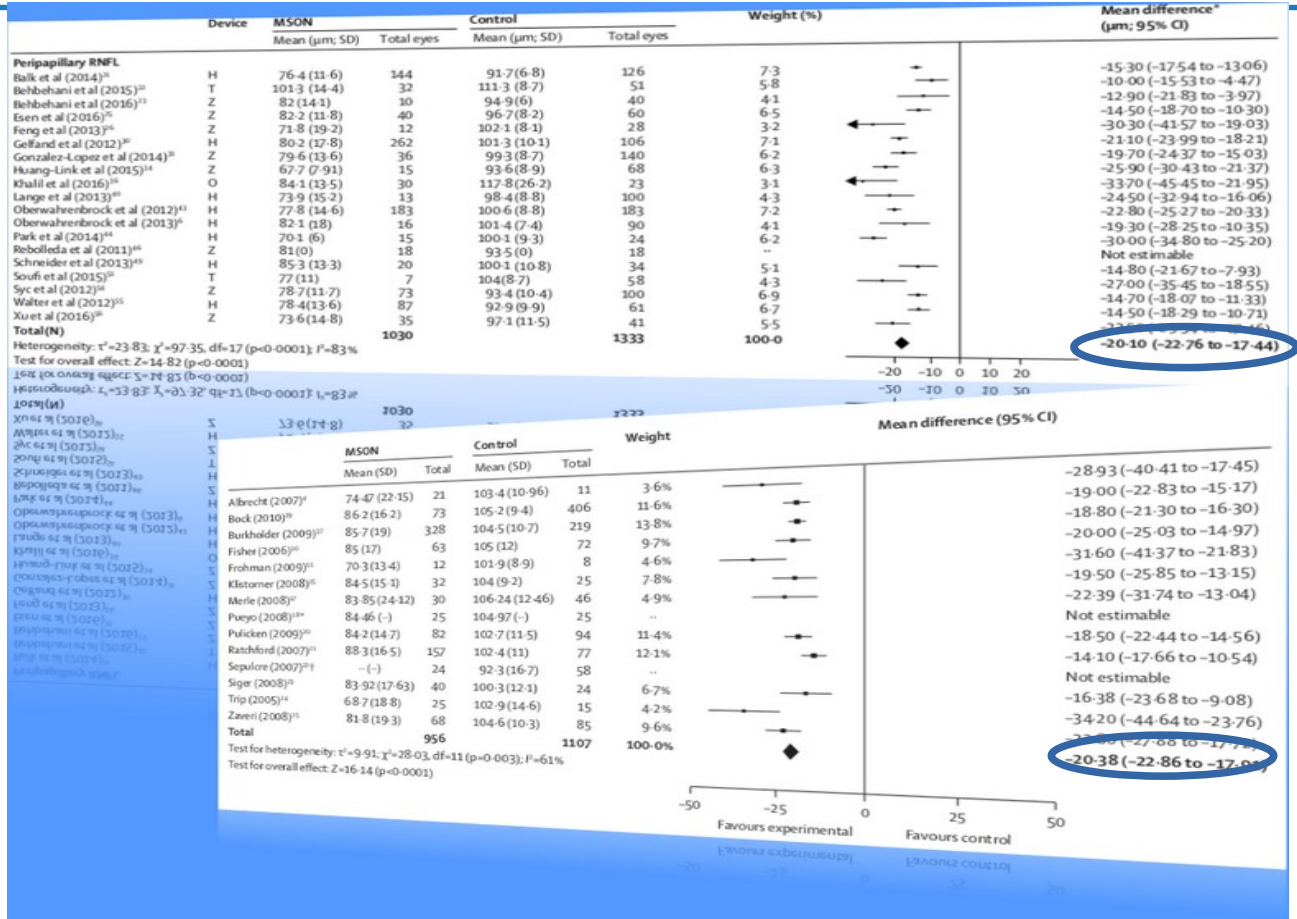








OCT in MS-ON



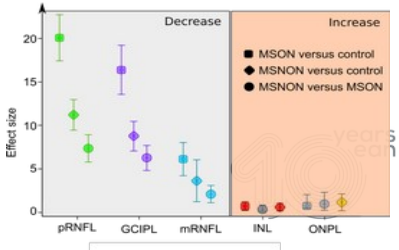
pRNFL atrophy

TLN 2010

20.10 (17.44-22.76) µm

TLN 2017

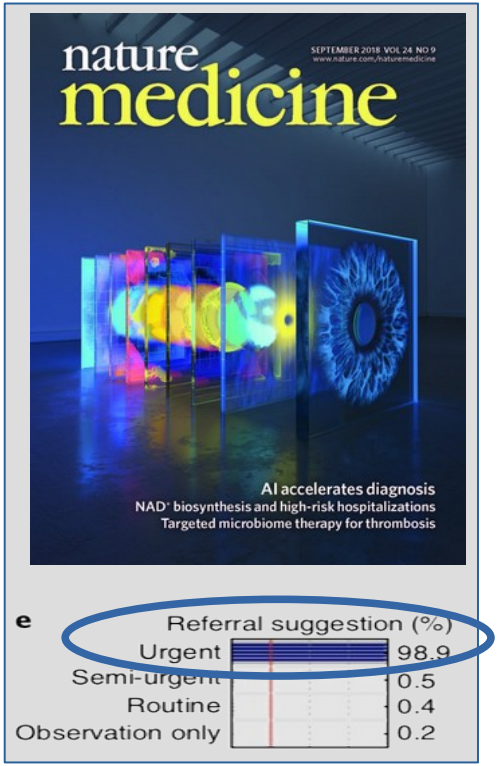
20.38 (17.91-22.86) µm

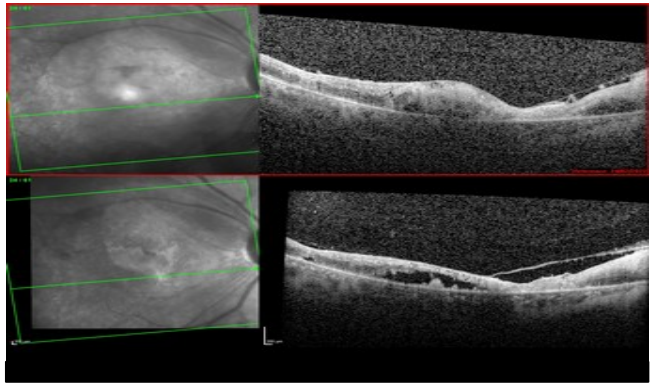


Last case

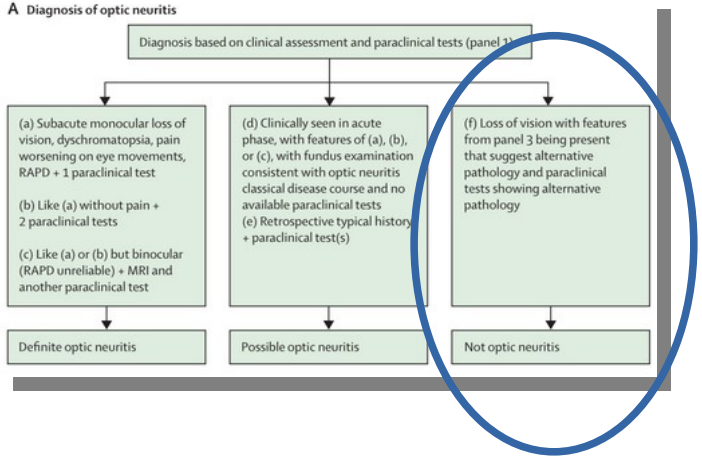
- 32 year old woman with RRMS
- Natalizumab for > 10 years
- Develops progressive cloudy vision in right eye
- Started on corticosteroids for suspected MS-ON
- MRI: no enhancement of right optic nerve, no new lesions
- Vision continues to worsen (HM)
- 22 days after onset seen @MEH
- OCT: …

OCT & AI





- **<u>VZV vitritis</u>**
  AI results:
  - Observation: 1.71%
  - Routine: 24.09%
  - Semi-Urgent: 46.39%
  - Urgent: 27.80%



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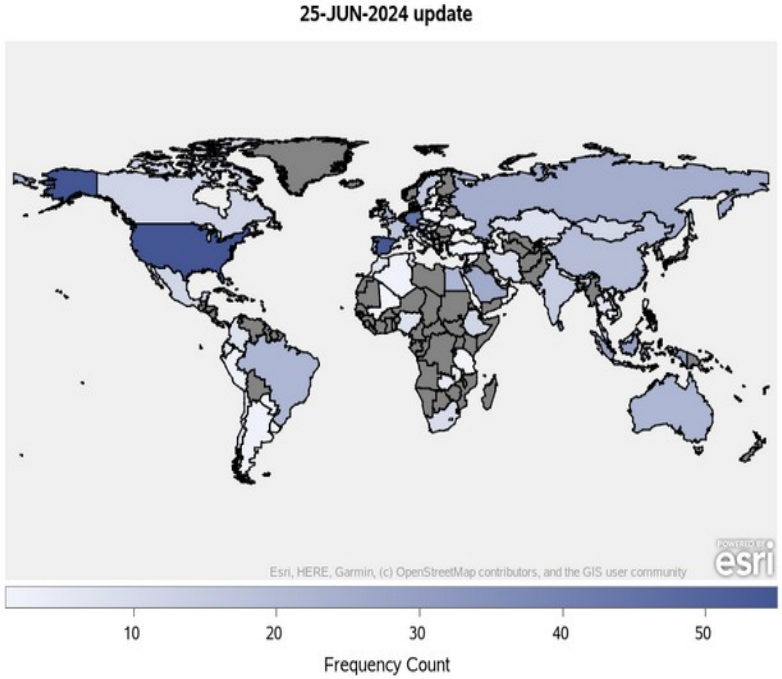
ICON survey



- Teaching
- Overview on global distribution of ON subgroups
- Identification of lack of resources as a powerful instrument to advocate resource mobilisation
- Co-authorship in joint publication under study group name “ICON”

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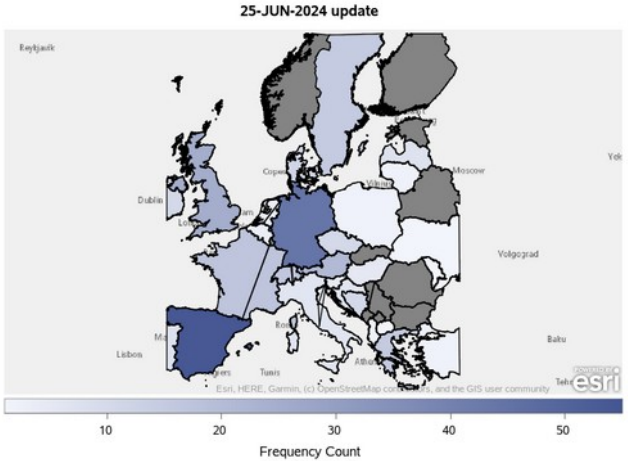
Summary

- Overview on Optic neuritis
- ICON 2022 Diagnostic Criteria
- Classification of Optic neuritis
- 3 Clinical Scenarios
- Invitation to participate in ICON survey
- Outlook: AI

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Thank you
Q&A