Diagnose der Neuritis nervi optici gemäß den ICON 2022 diagnostischen Kriterien

Wiener Ophthalmologische Gesellschaft
Wissenschaftliche Sitzung
14.10.2024 19:30 Uhr
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Disclosures

Stichting MS Research NL
NIHR UK, UCSF, Amsterdam UMC
Novartis, Roche, Heidelberg Academy

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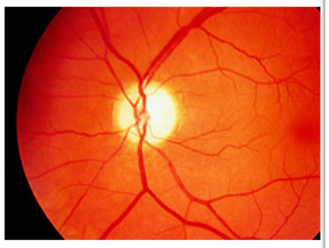
Published: September 22, 2022

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.



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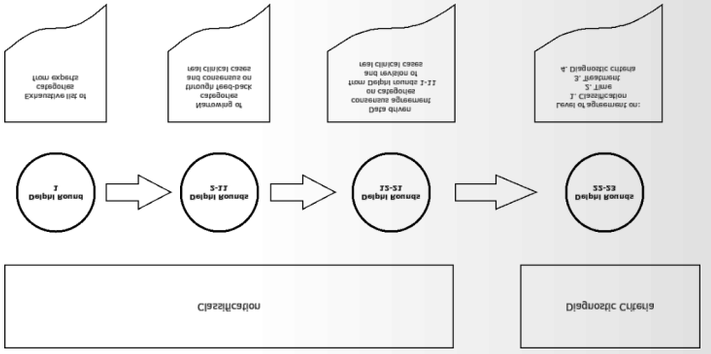
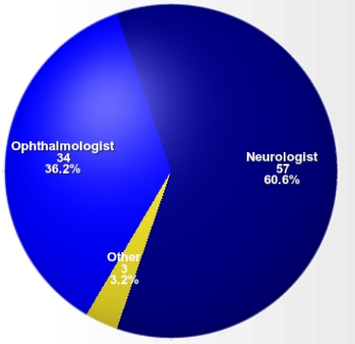
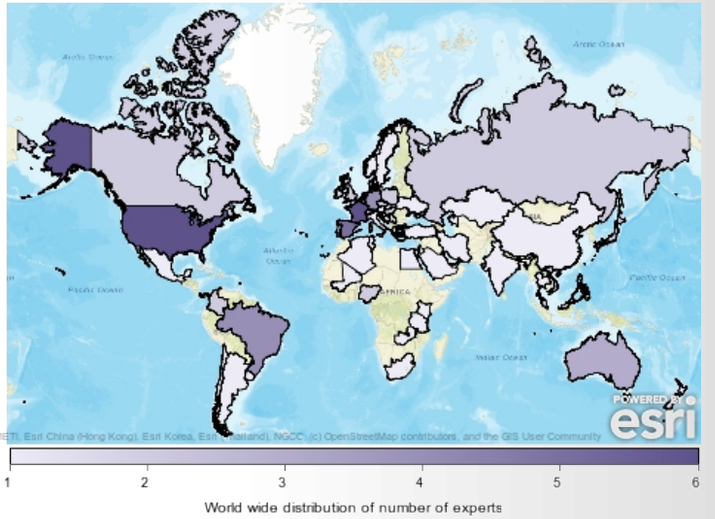
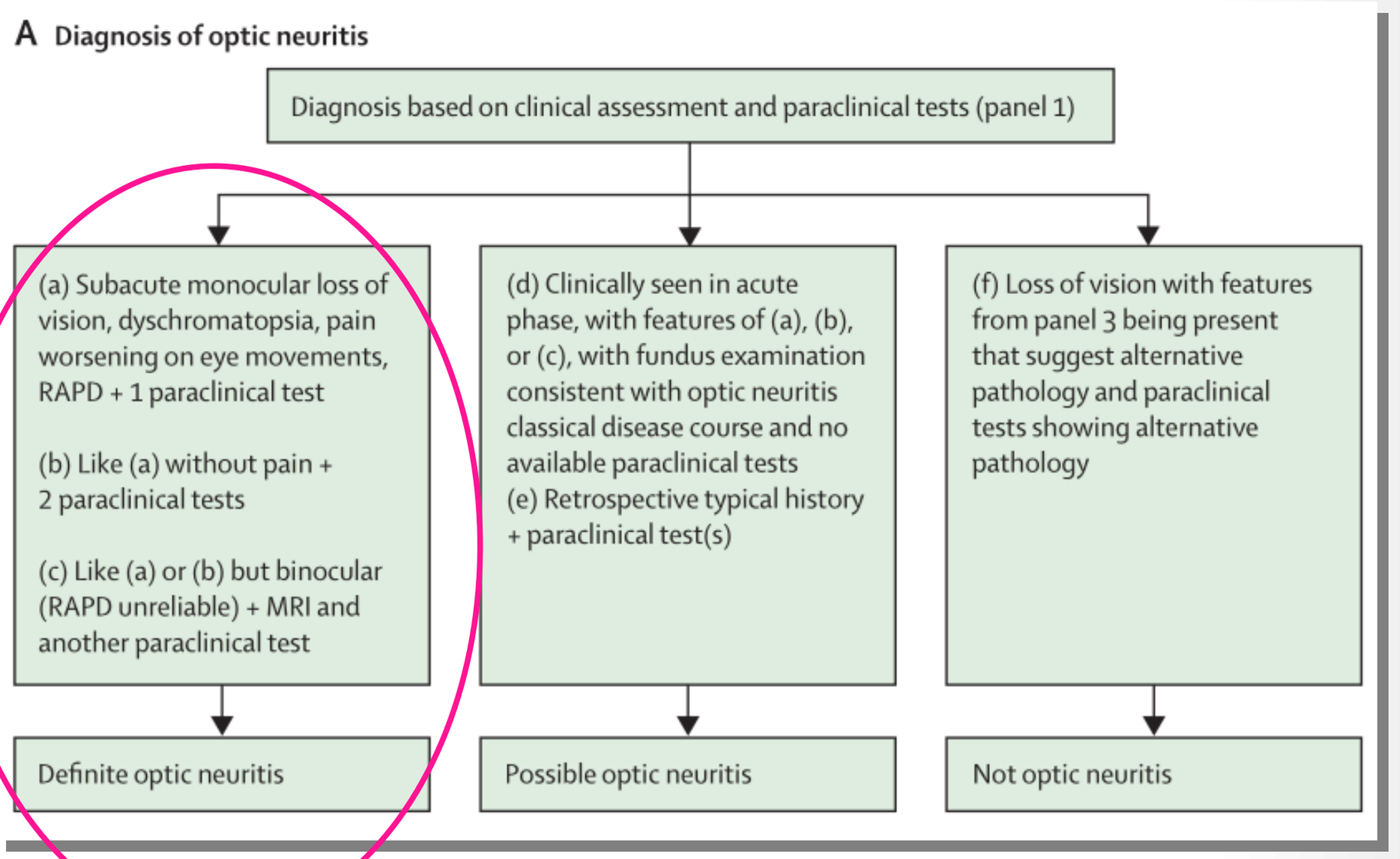






Diagnostische Kriterien



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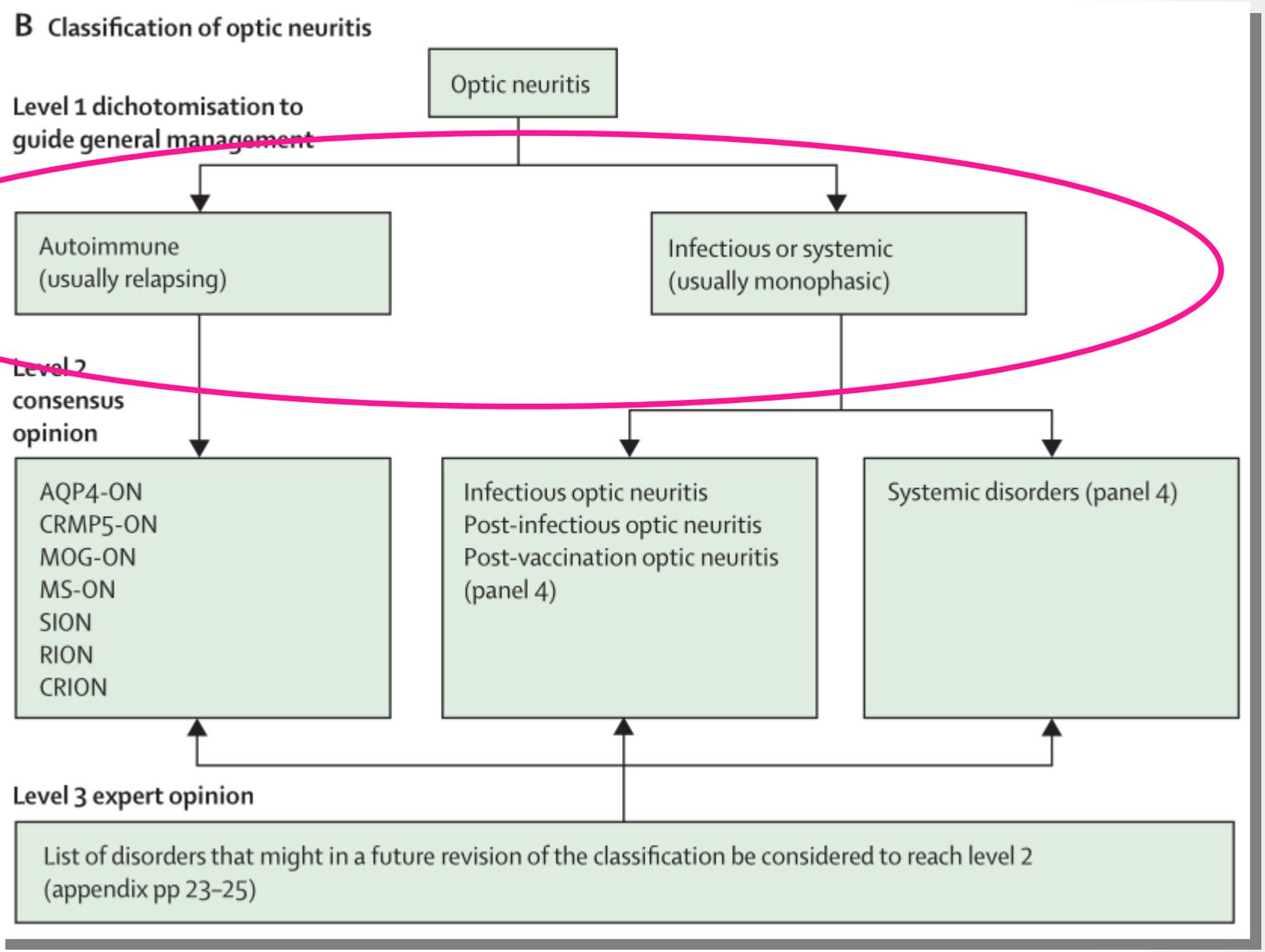
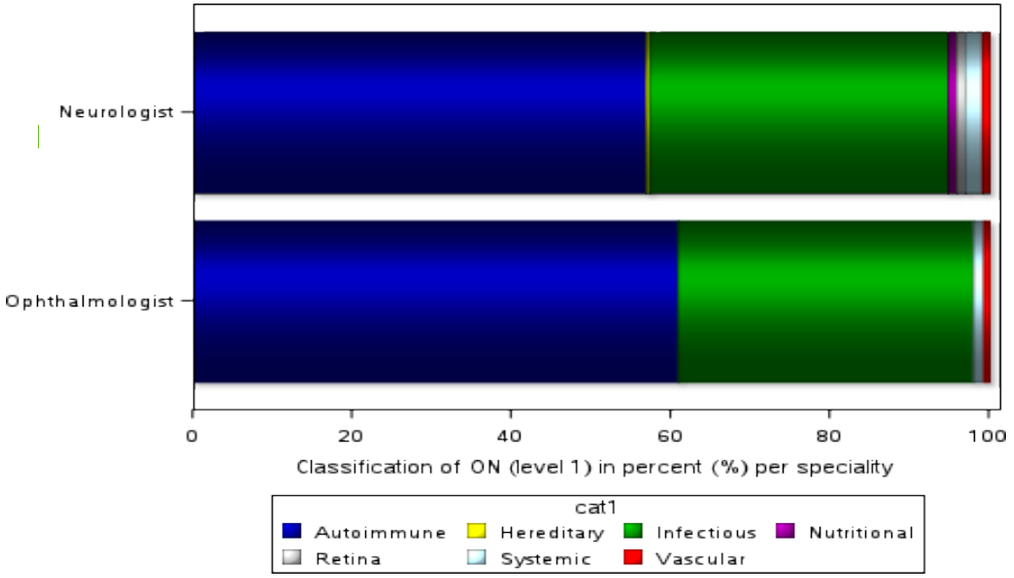


Konensus kann erreicht werden



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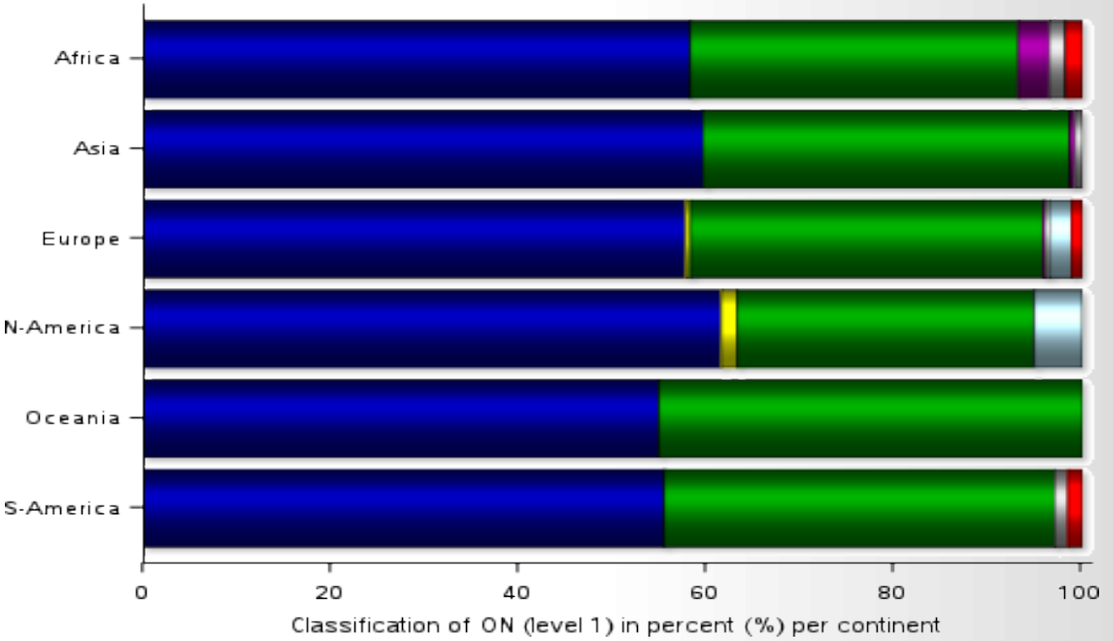

Geographisch

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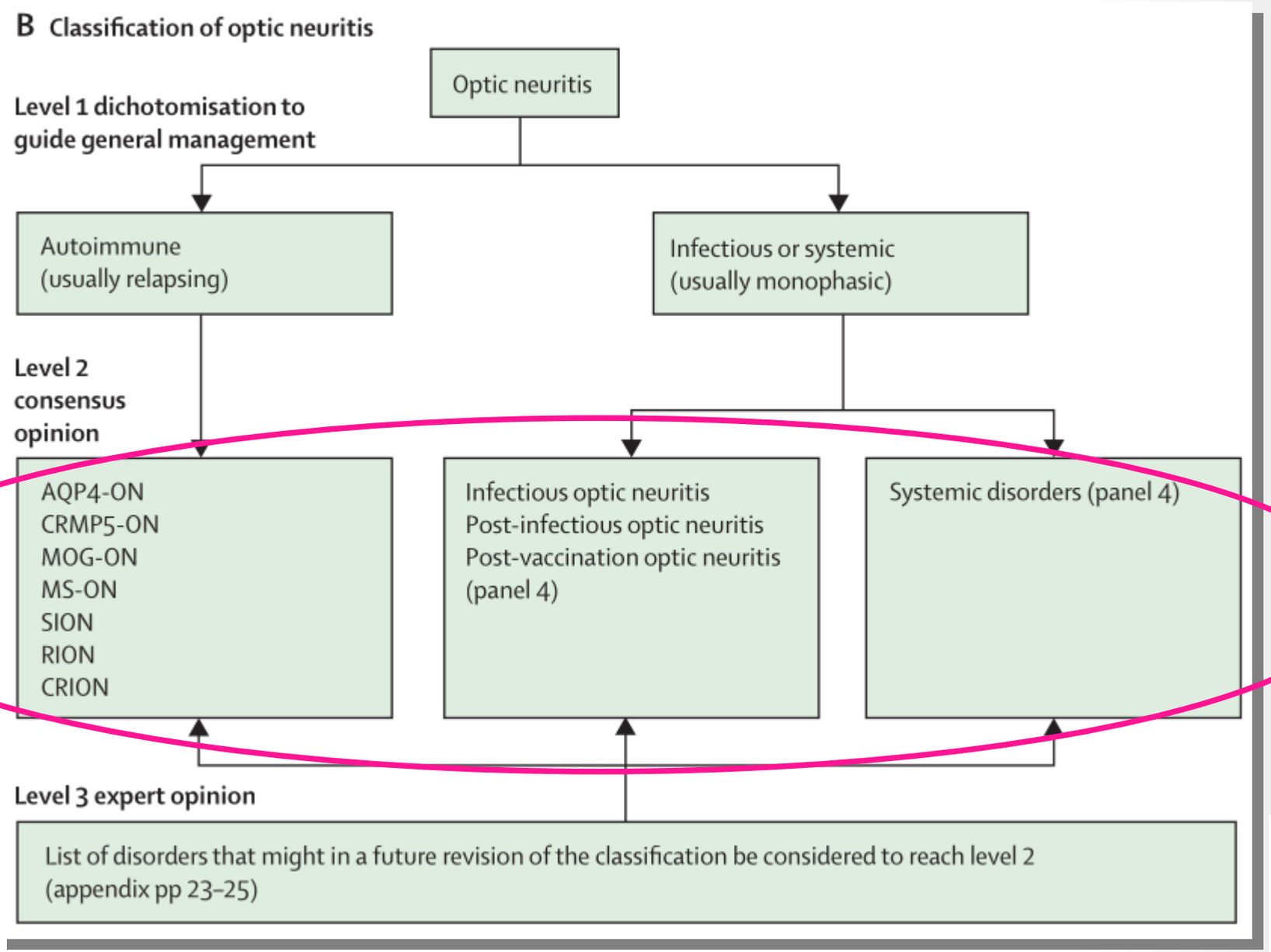


1. Fall

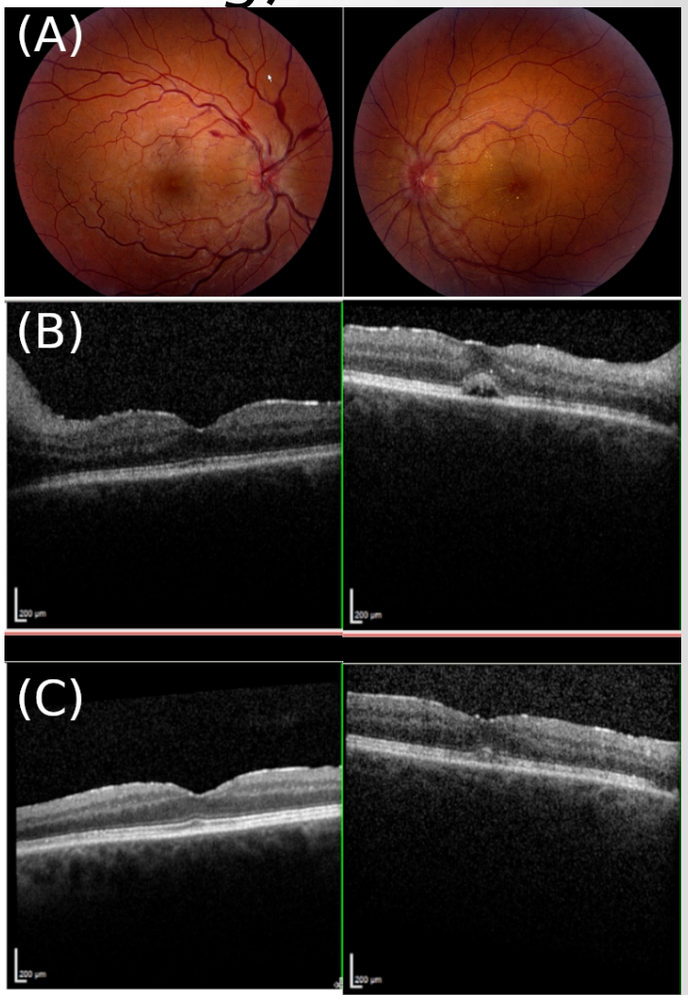
- 34-jährige kaukasische Patientin
- 7 Tage Schmerzen OD die verschlimmern mit Augenbewegungen
- Dyschromatopsie und VA OD 6/9, OS 6/5
- RAPD OD
- Müdigkeit, kognitive Probleme, Harninkontinenz, Depression
- vor 3 Jahren rechtsseitiges Taubheitsgefühl
- MRT: DIS & DIT & 3 Gd+ nicht-symptomatische Läsionen

2. Fall

- 28 Jahre alt, afrokaribischer Mann
- Schmerzloser Sehverlust OS (6/38)
- Dyschromatopsie
- RAPD OS
- Mehrere Episoden die auf Behandelung mit Corticosteroiden ansprechen über ~20 Jahre
- OCT: pRNFL-Atrophie OS (IEPD >5%)
- MRT geschwollener, kontrastaufnehmender Sehnerf Gehirn und Rückenmark normal
- AQP4 Antikörper seropositiv

3. Fall

- 72-jähriger Mann erkrankt in Vietnam an Fieber
- 2-3 Wochen später beidseitiger, sequentieller, schmerzloser Sehverlust (Lichtwahrnehmung)
- kein RAPD (aber beide Pupillen verengen sich durch Akkommodation)
- Fundus und OCT:
- Keine Verbesserung nach 6 Monaten (Corticoide ca. 6 Wochen nach Beginn des Visusverlustes verabreicht)



3 Szenarien

- Fall 1: Ist das MS?

  Szenario A : schmerzhaft, monokulär, subakuter Visus Verlust, Dyschromatopsie, RAPD

- Fall 2: Ist das NMO?

  Szenario B : keine Schmerzen, monokulär, subakuter Visusverlust, Dyschromatopsie, RAPD

- Fall 3: Was ist das?

  Szenario C : binokulär, subakuter Visusverlust, Dyschromatopsie, keine Schmerzen, RAPD unzuverlässig

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).

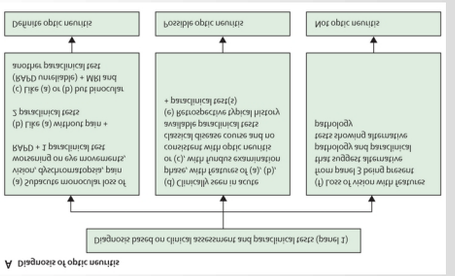
Application of the clinical and paraclinical criteria

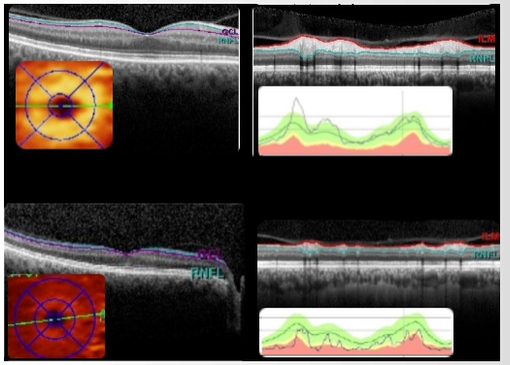
Definite optic neuritis

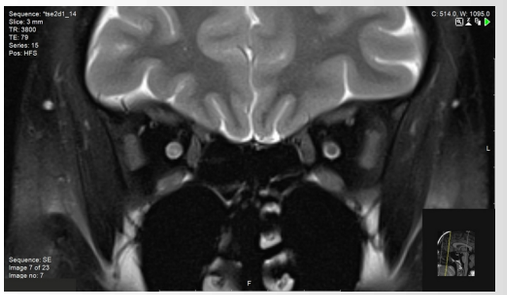
- (A) and one paraclinical test
- (B) and two paraclinical tests of different modality
- (C) and two different paraclinical tests of which one is MRI

Possible optic neuritis

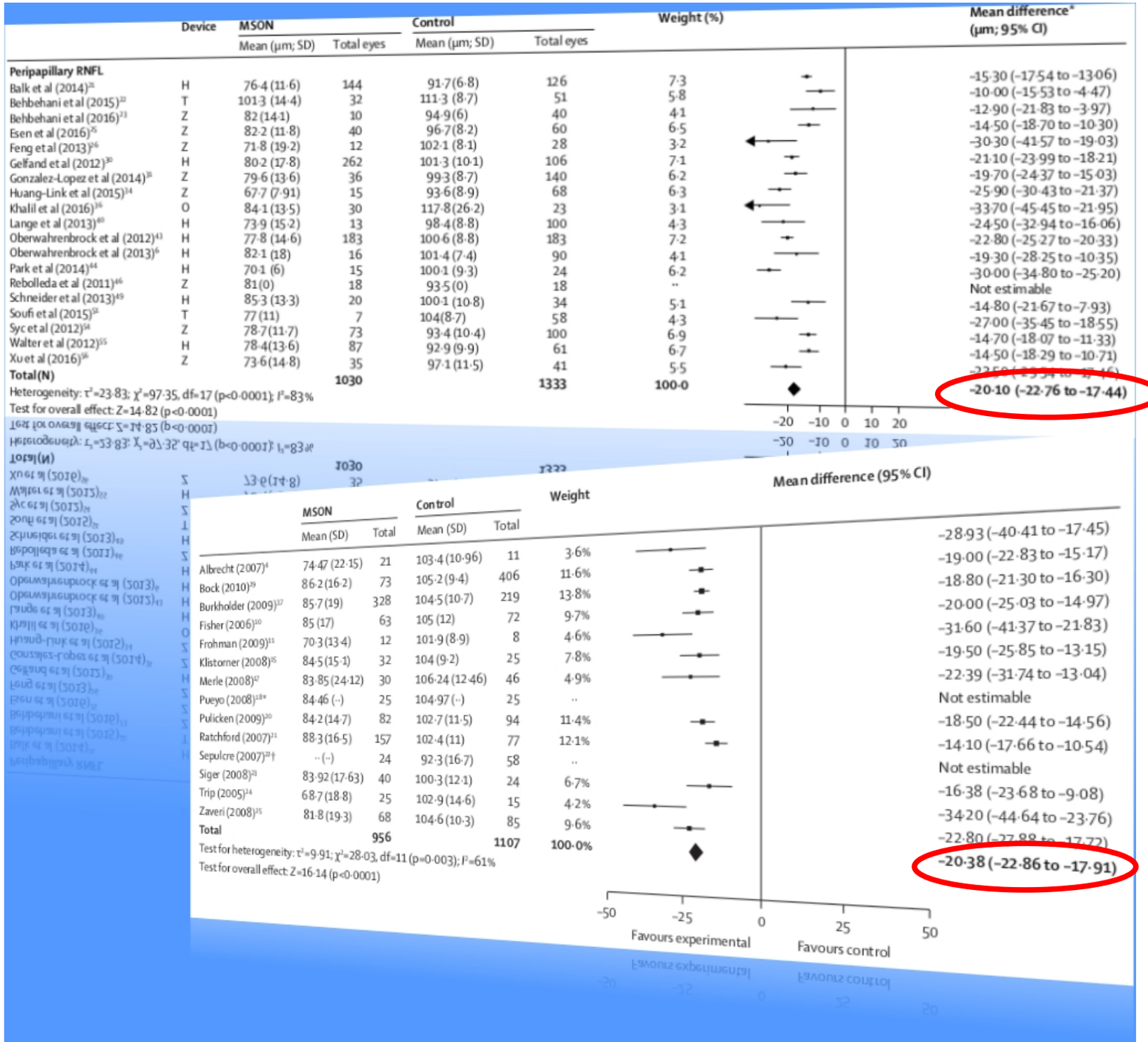
- (A), (B), or (C) if seen acutely but in absence of paraclinical tests, with fundus examination typical for optic neuritis and consistent with the natural history during follow-up
- Positive paraclinical test or tests, with a medical history suggestive of optic neuritis







OCT in MS-ON



pRNFL-Atrophie

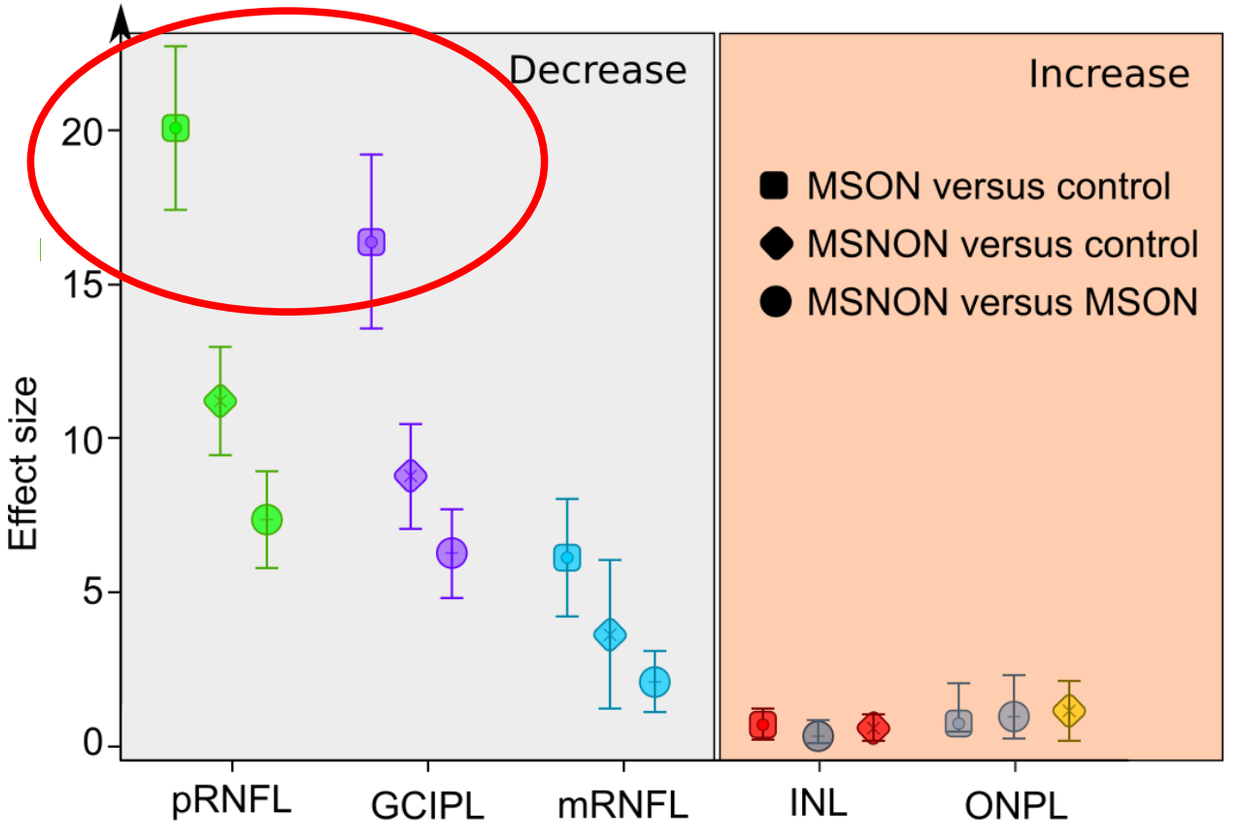
TLN 2010

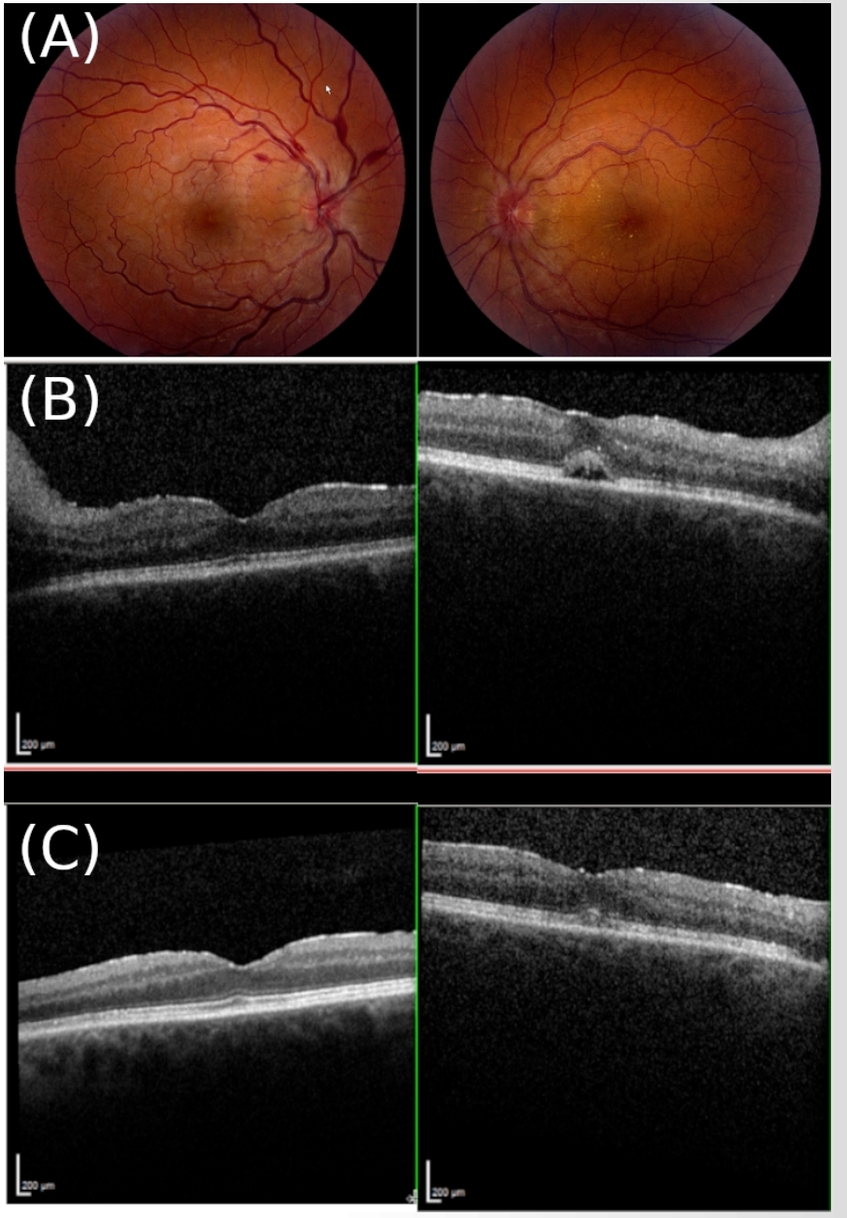
20,10 (17,44 – 22,76) µm

TLN 2017

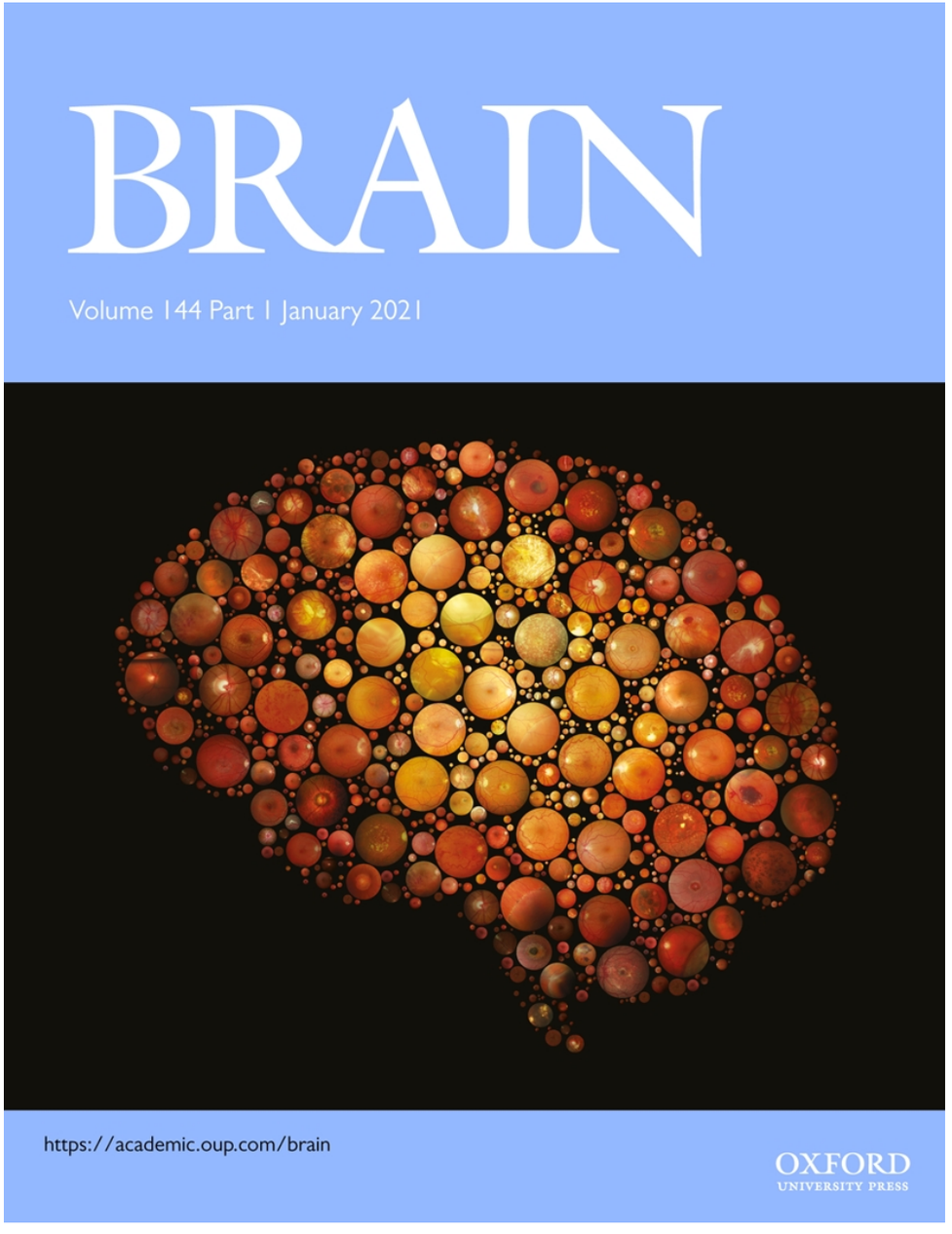
20,38 (17,91 – 22,86) µm

Was ist relevant?





Retinal Asymmetrie



Inter-eye difference:

Prozentual (IEPD): %

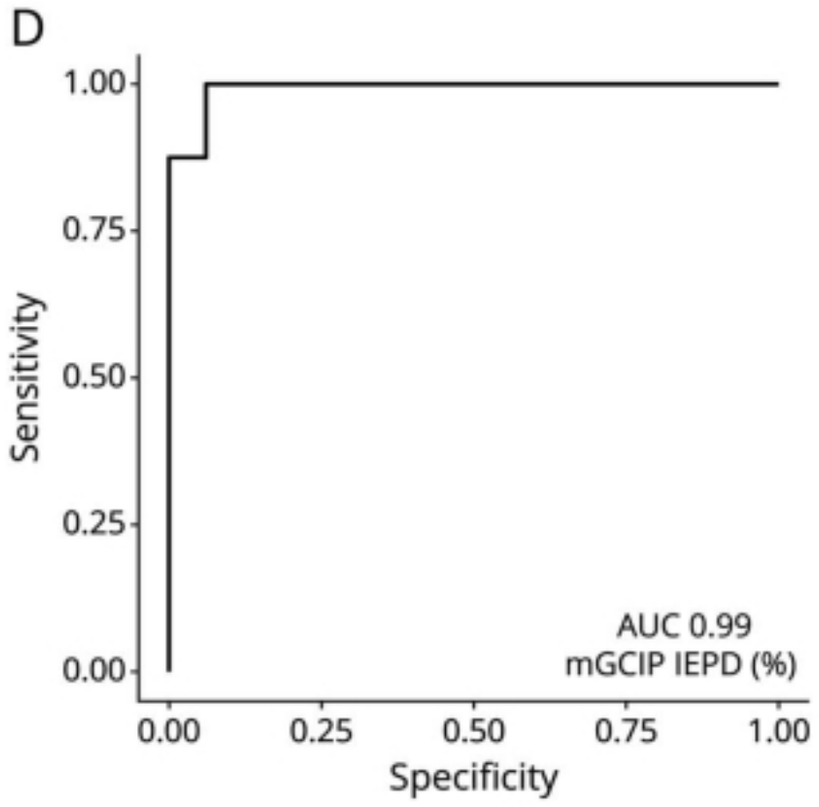
Absolute (IEAD): µm

OCT als diagnostisches Kriterium

Retinal Asymmetrie	Grenzwert	Literatur	Patientengruppe	Spezifität	Sensifität
IEPD mGCIPL	6 %	Coric et al. 2017	Symptomatic unilateral MSON vs. healthy controls	97	70
IEAD mGCIPL	4.0 µm	Nolan-Kenney 2019	Symptomatic unilateral MSON vs. non-MSON	77	68
IEAD mGCIPL	3.5 µm	Behbehani 2020	Unilateral optic neuritis vs. healthy controls	98	100
IEAD mGCIPL	2.83 µm	Davion 2020	Symptomatic unilateral or bilateral MSON vs. non-MSON[a]	67	67
IEPD/IEAD mGCIPL	4% / 4 µm	Petzold 2020	MS without MSON vs controls (n=72,120)	83 / 87	52 / 44
IEAD mGCIPL	1.42 µm	Outteryck 2020	CIS patients with vs. without an asymptomatic optic nerve lesion on 3D-DIR MRI	73	89
IEPD mGCIPL	2 %	Outteryck 2020	CIS patients with vs. without an asymptomatic optic nerve lesion on 3D-DIR MRI	70	89
IEPD/IEAD mGCIPL	4% / 4 µm	Oertel 2023	NMO-ON vs controls	96 / 98	75 / 82
IEPD/IEAD mGCIPL	4% / 4 µm	Volpe 2024	MOG-ON vs controls	82 / 82	>99 / >99

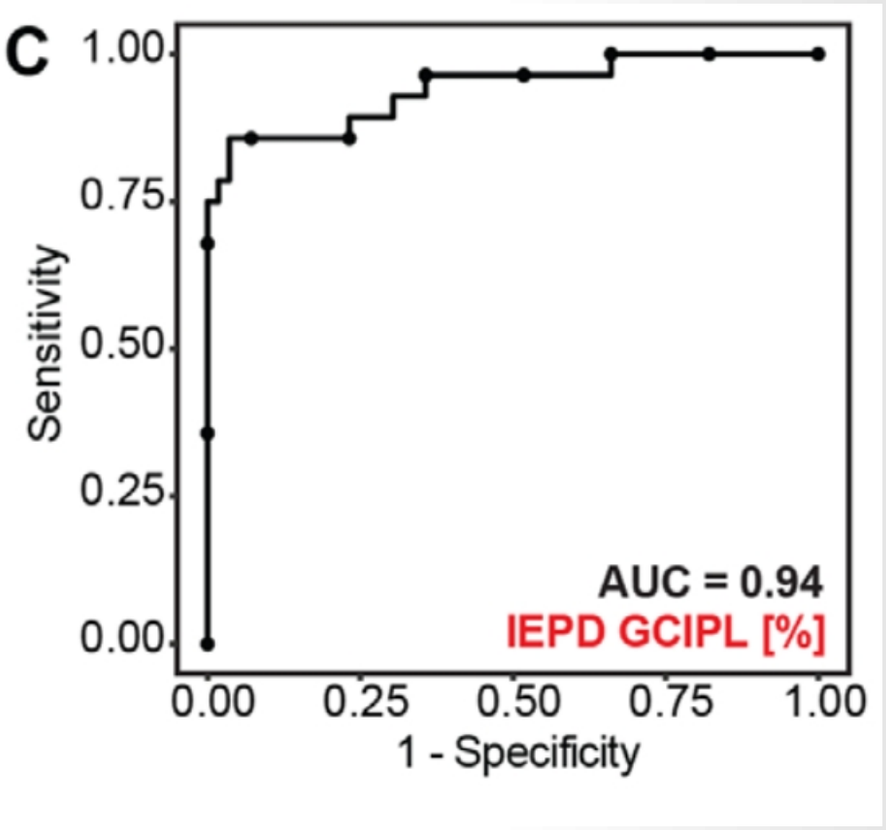
Retinale Asymmetrie im OCT

MOG-ON



Volpe, G. et al. Neurol Neuroimmunol Neuroinflamm 2024

NMO-ON



Oertel, F. et al. JNPP 2023

Was ist neu für die Augenheilkunde?

- Anatomisch: Kompartimente
- Phenotypisch:
  - Pre-läminare neuritis nervi optici
  - Primär progressive neuritis nervi optici (PPON)

Kompartimente

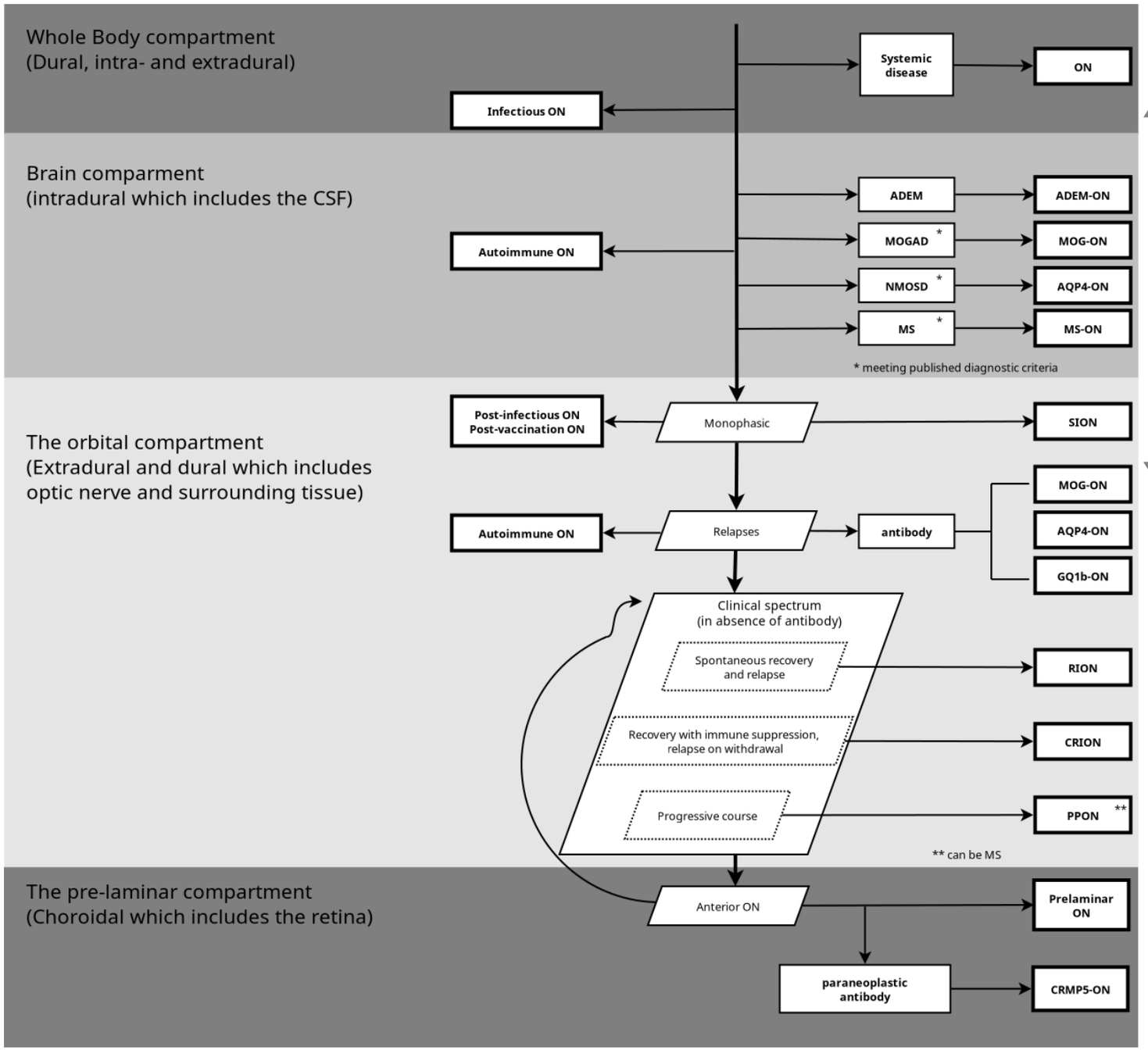
Compartment

An anatomically and immunologically defined space as relevant to pathogenesis of optic neuritis:

- dural, intradural, and extradural (whole body)
- intradural (including CSF compartment)
- extradural and dural (includes optic nerve sheath and surrounding tissue)
- choroidal (includes retina and uveal tract)

Antibody production can be intradural, extradural, or choroidal, and the target antigen can be limited to one or more compartments.

Kompartimente



Neurologen

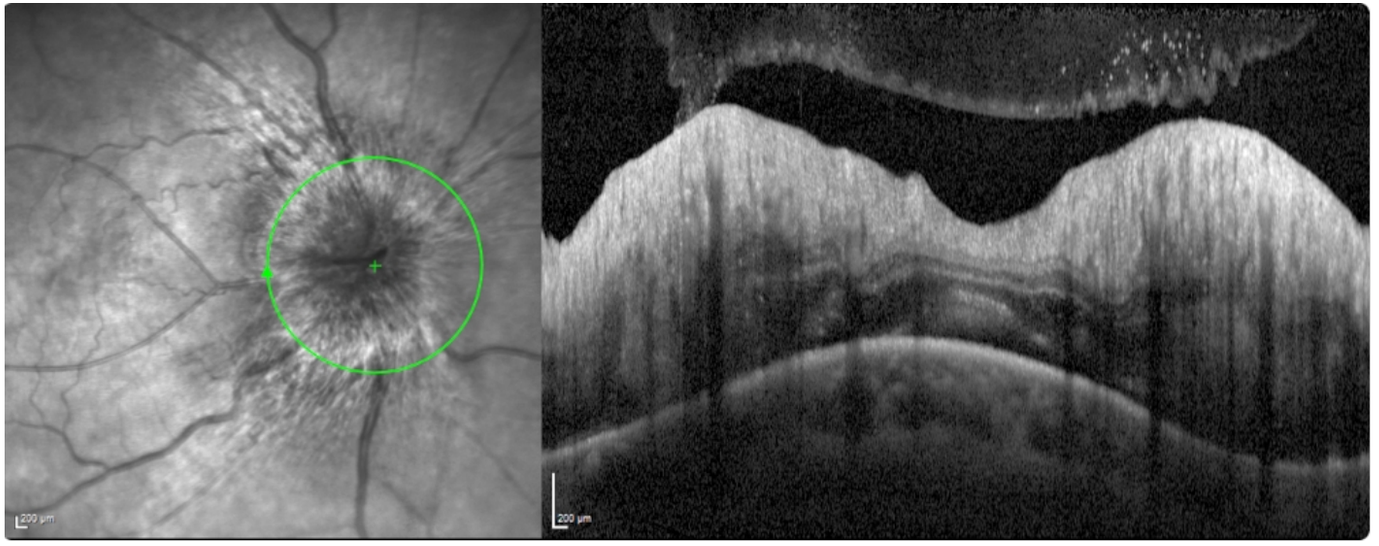
Augenärzte

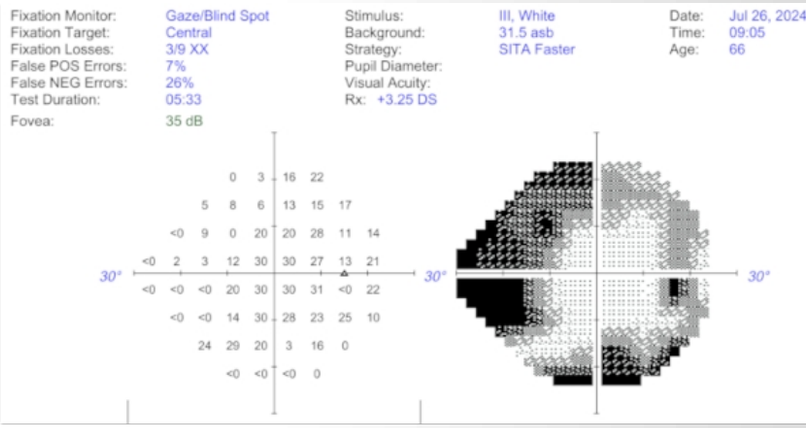
Pre-läminare neuritis nervi optici

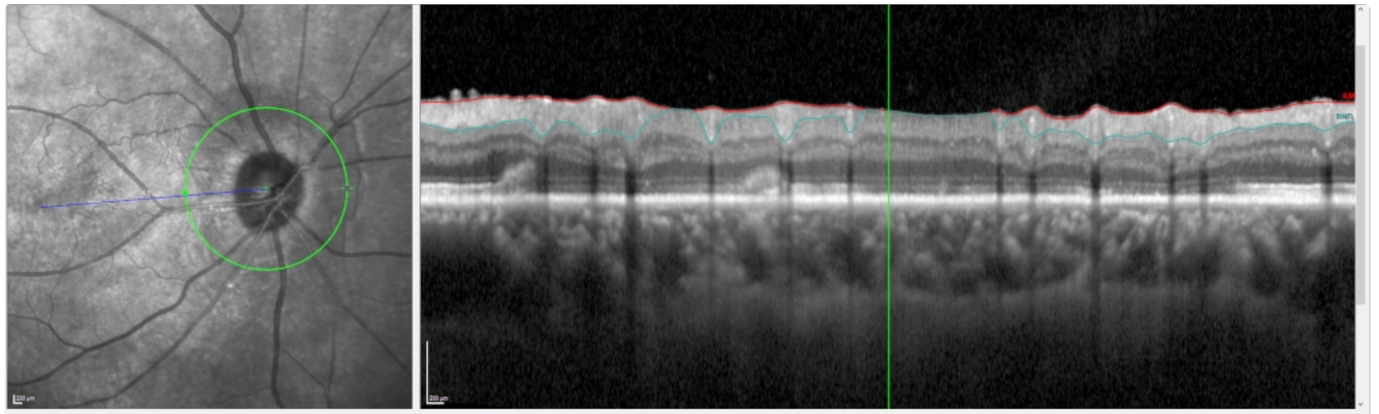
Prelaminar optic neuritis

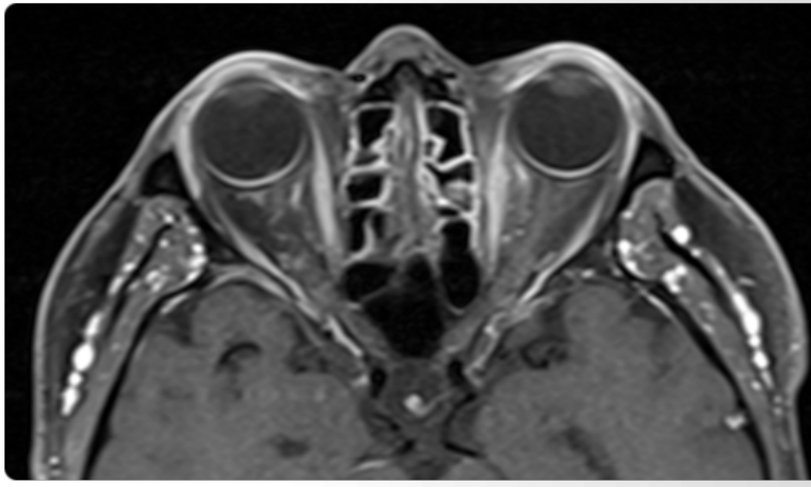
The most anterior manifestation of optic neuritis, which involves the non-myelinated retinal axons and ganglion cell layer and which remains restricted to the prelaminar optic nerve. Acutely, the MRI of the retrobulbar optic nerve does not show an abnormality. Prelaminar optic neuritis is an anatomically based description that applies to all subforms of optic neuritis.

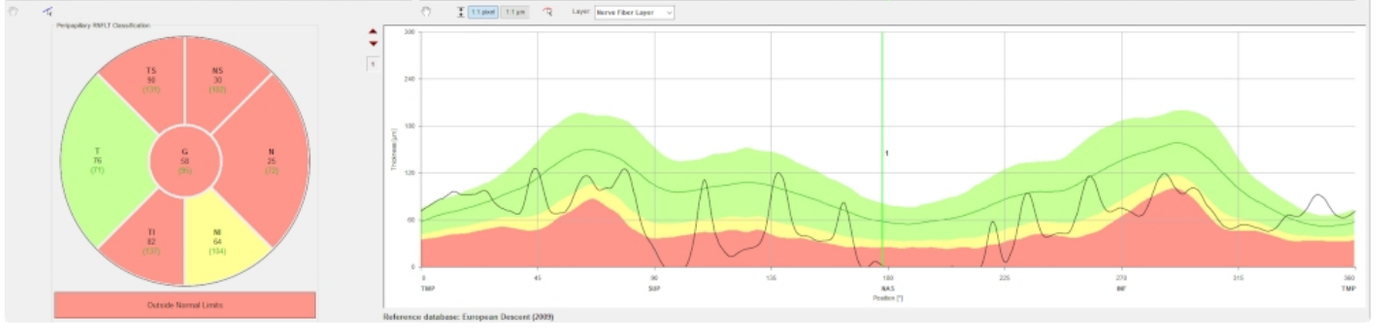
Pre-läminare neuritis nervi optici

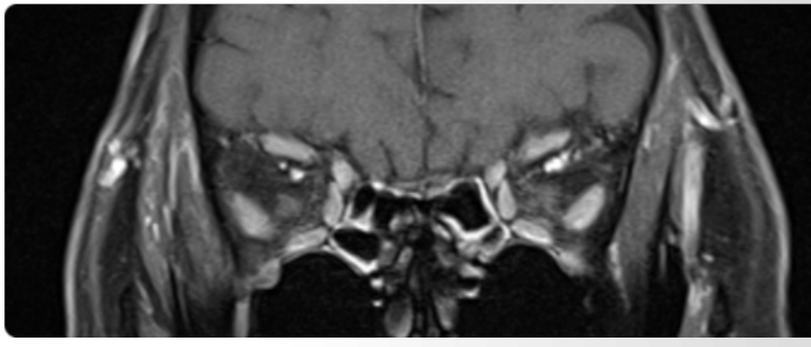












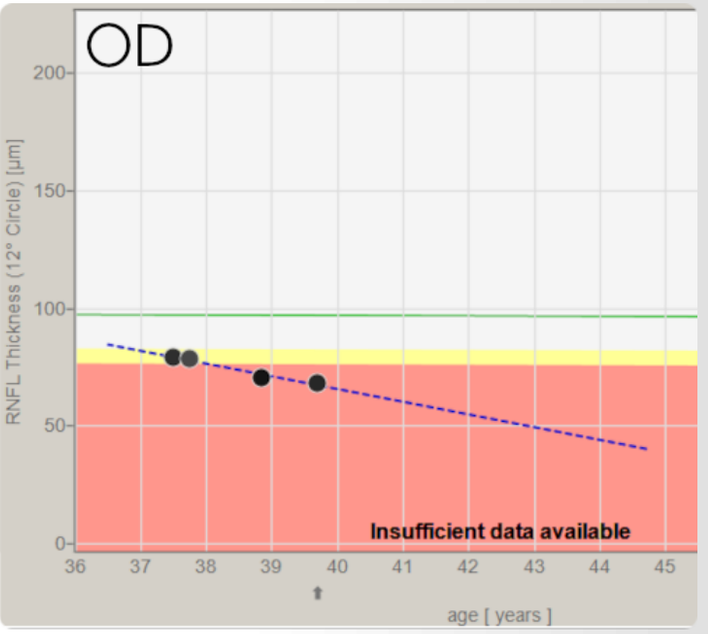
Primär progressive neuritis nervi optici

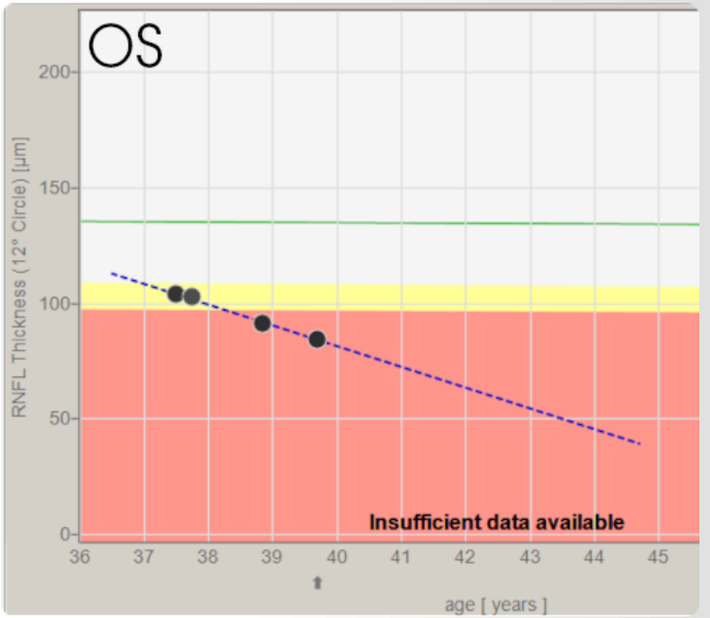
PPON

Primary progressive optic neuritis. Diagnosis requires progressive atrophy or progressive visual loss, or both for >12 months. Diagnosis of PPON is based on time and applies to all subforms of ON that present with a progressive rather then a relapsing disease course.

Primär progressive neuritis nervi optici

- MS im 27. Lebensahr, nimmt Dimethyl Fumarat
- Visus:
  - 2022 OD 6/9 OS 6/24
  - 2023 OD 6/18 OS 6/24
  - 2024 OD 6/48 OS 6/60
- Pattern VEP langsam und klein. Pattern ERG N95:P50 klein. Full field ERG normal.
- MRI: DIS/DIT stabiel





Lösungen zu den 3 Fällen

- Fall 1: **MS-ON**
  Szenario A: schmerzhaft, monokulär, subakuter Visusverlust, Dyschromatopsie, RAPD
- Fall 2: **NMO-ON**
  Szenario B: keine Schmerzen, monokulär, subakutes Visusverlust, Dyschromatopsie, RAPD
- Fall 3: **postinfektiöse ON (Dengue)**
  Szenario C: binokulär, subakuter Visusverlust, Dyschromatopsie, keine Schmerzen, RAPD unzuverlässig

Zusammenfassung

- Klinischer Ansatz zur neuritis nervi optici
- ICON 2022 Diagnostische Kriterien
- ICON 2022 Klassifizierung
- 3 Klinische Szenarien
- Neu: primär progressiv & pre-laminär
- Zukünftige ICON Revisionen sind geplant

Herzlichen Dank

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