

Diagnose der akuten und späten Neuritis nervi optici in der Augenheilkunde

Universitätsspital Basel
Augenklinik 05.11.2024
07:30-08:00 Uhr
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Disclosures

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

Hintergrund

Series from the Lancet journals

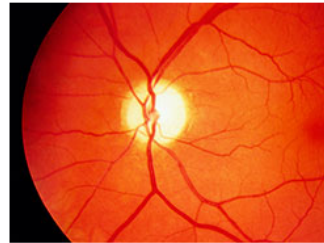
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Optic Neuropathies

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.



Related Content

POSITION PAPER Diagnosis and classification of optic neuritis

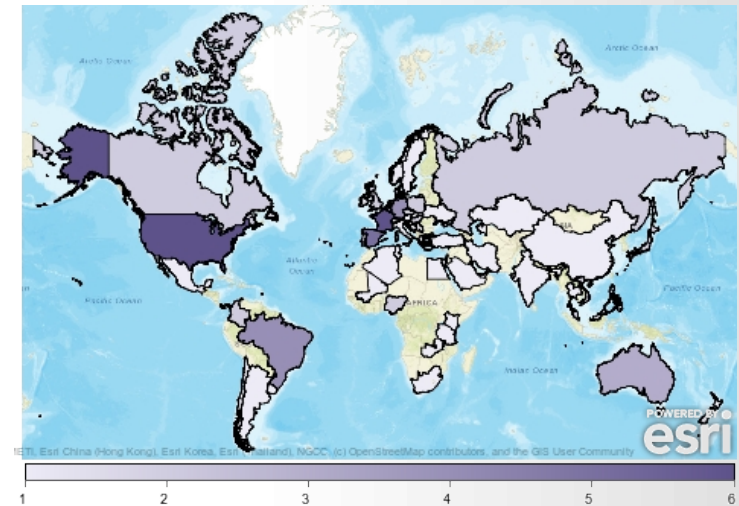
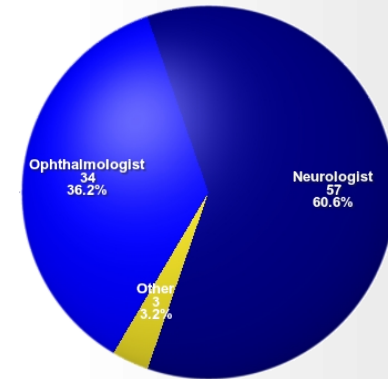
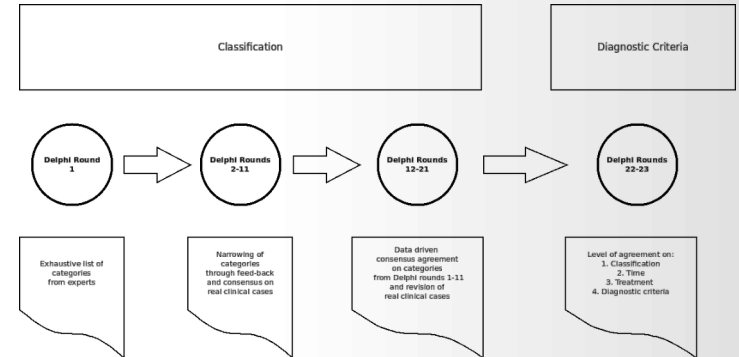
Axel Petzold, Clare L Fraser, Mathias Abegí Raed Alroughani, Daniah Alshowaier, Regina Alvarenga, and others
The Lancet Neurology
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PERSONAL VIEW Myelin-oligodendrocyte glycoprotein antibody-associated disease

Romain Marignier, Yael Hachohen, Alvaro Cobo-Calvo, Anne-Katrin Pröbstel, Orhan Aktas, Harry Alexopoulos, and others
The Lancet Neurology, Vol. 20, No. 9
Published: September, 2021
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REVIEW Mitochondrial disease in adults: recent advances and future promise

Yí Shiau Ng, Laurence A Bindoff, Gráinne S Gorman, Thomas Klopstock, Cornelia Kornblum, Michelangelo Mancuso, and others
The Lancet Neurology, Vol. 20, No. 7
Published: July, 2021



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
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Optic neuritis and autoimmune optic neuropathies: advances in diagnosis and treatment

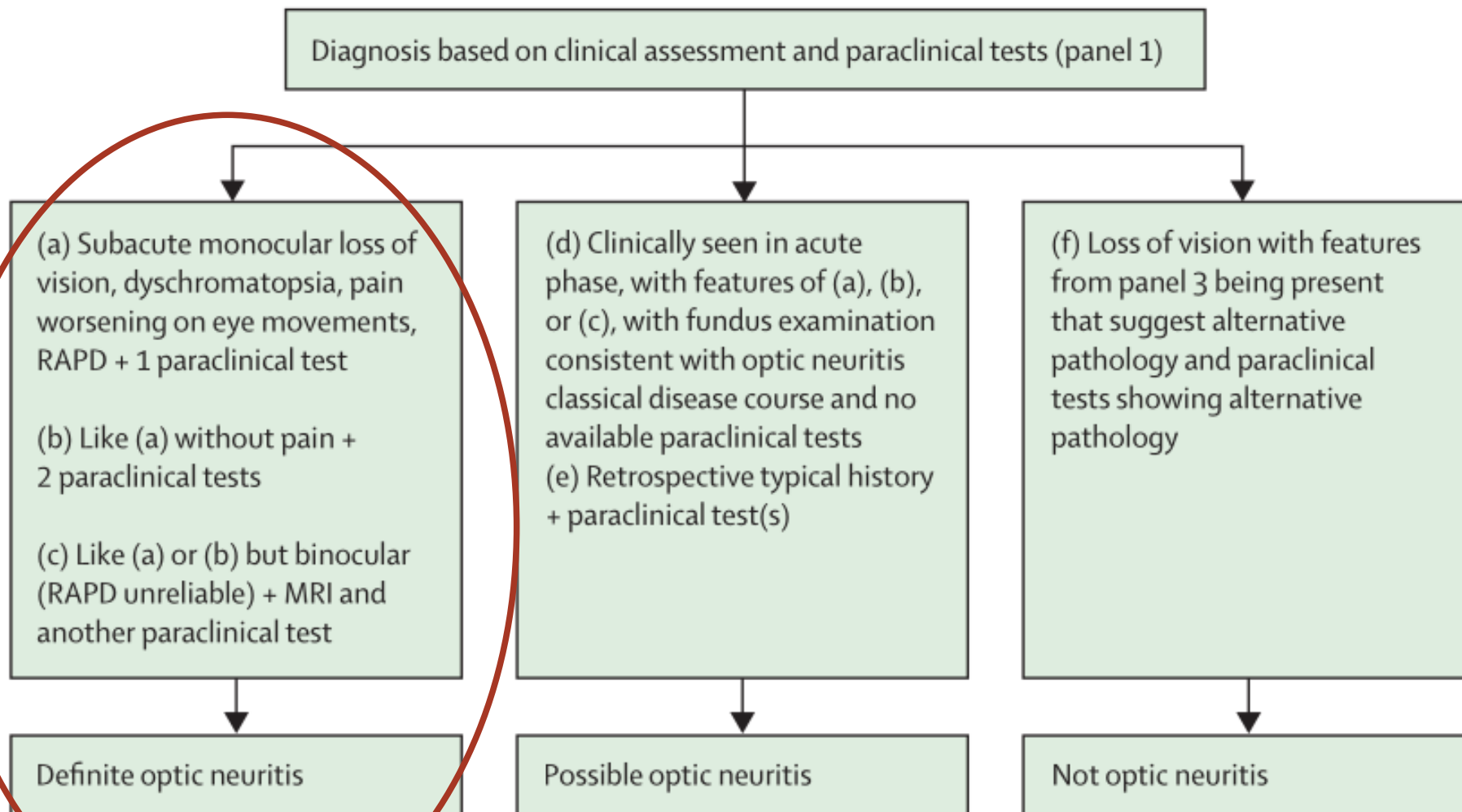
Jeffrey L Bennett, Fiona Costello, John J Chen, Axel Petzold, Valérie Biousse, Nancy J Newman, Steven L Galetta
The Lancet Neurology
Published: September 22, 2022
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Understanding the molecular basis and pathogenesis of hereditary optic neuropathies: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

Nancy J Newman, Patrick Yu-Wai-Man, Valérie Biousse, Valerio Carelli
The Lancet Neurology
Published: September 22, 2022
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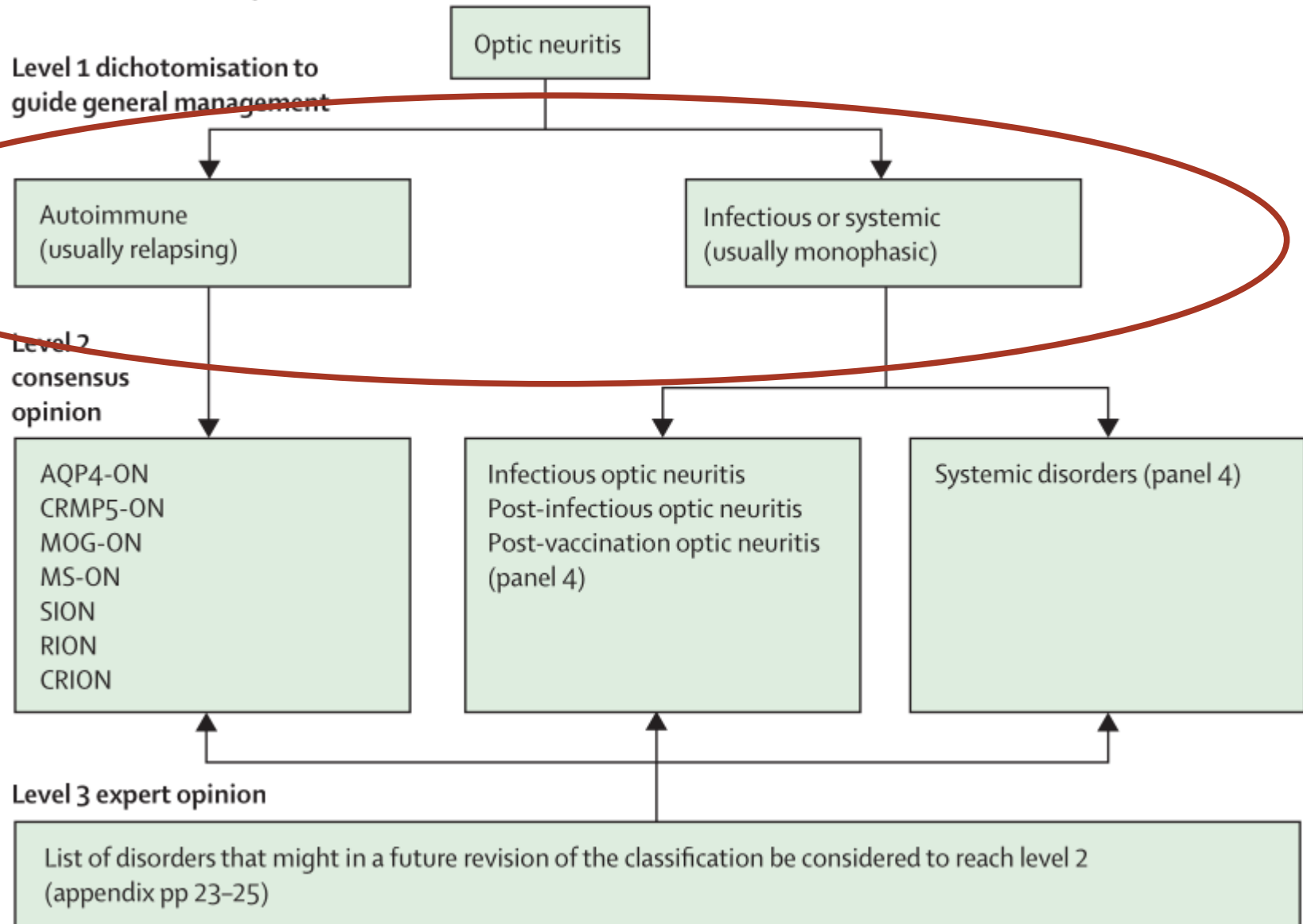
Diagnostische Kriterien

A Diagnosis of optic neuritis

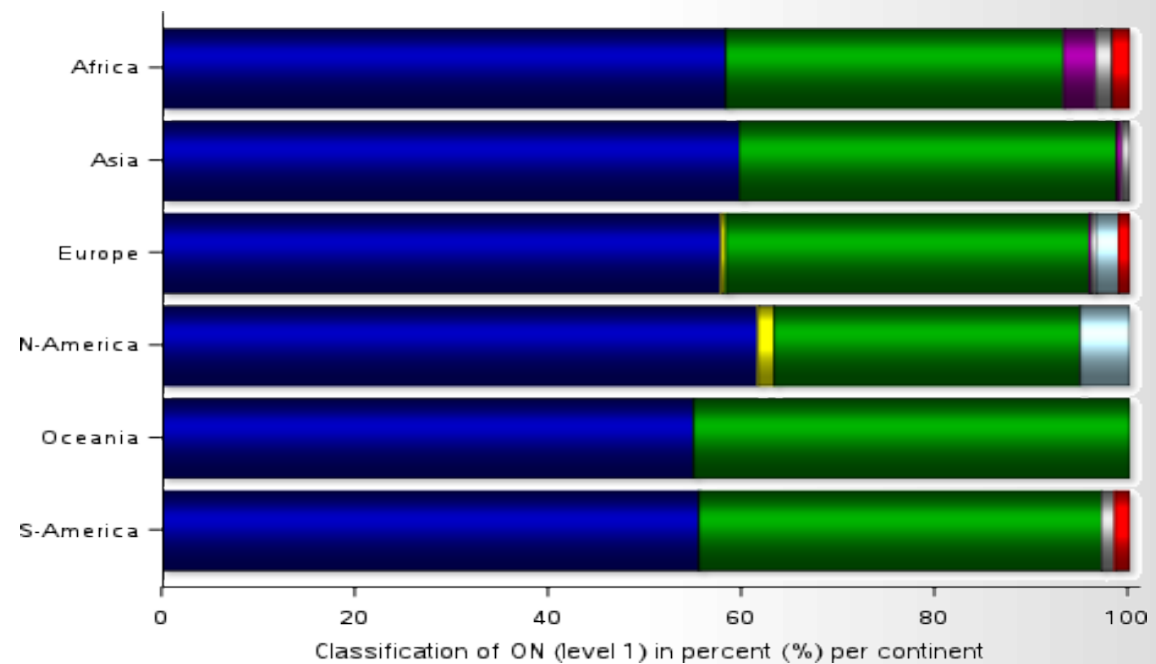
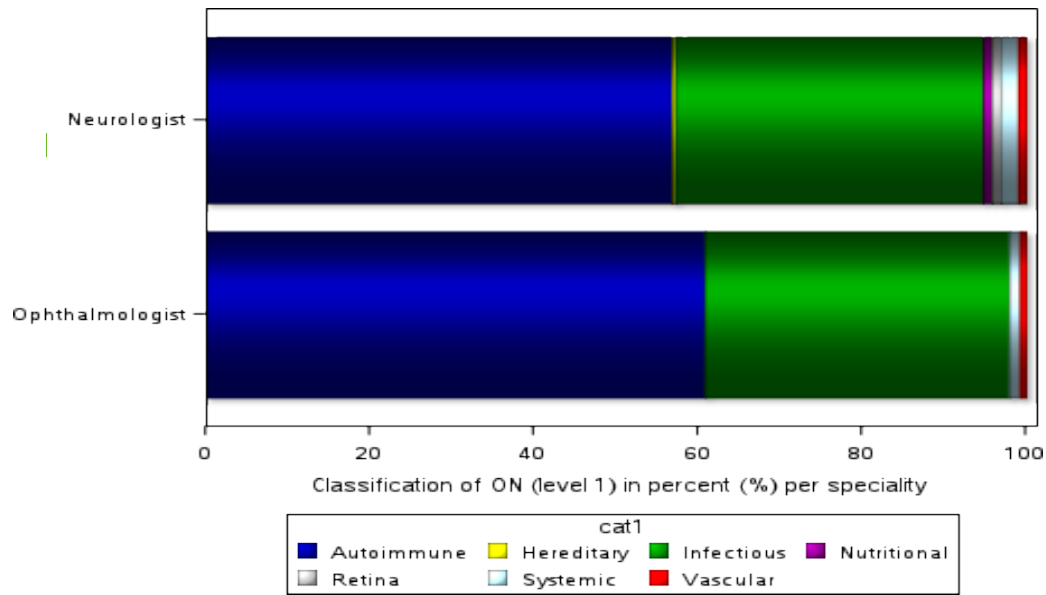


Klassifizierung

B Classification of optic neuritis



Konensus ist möglich

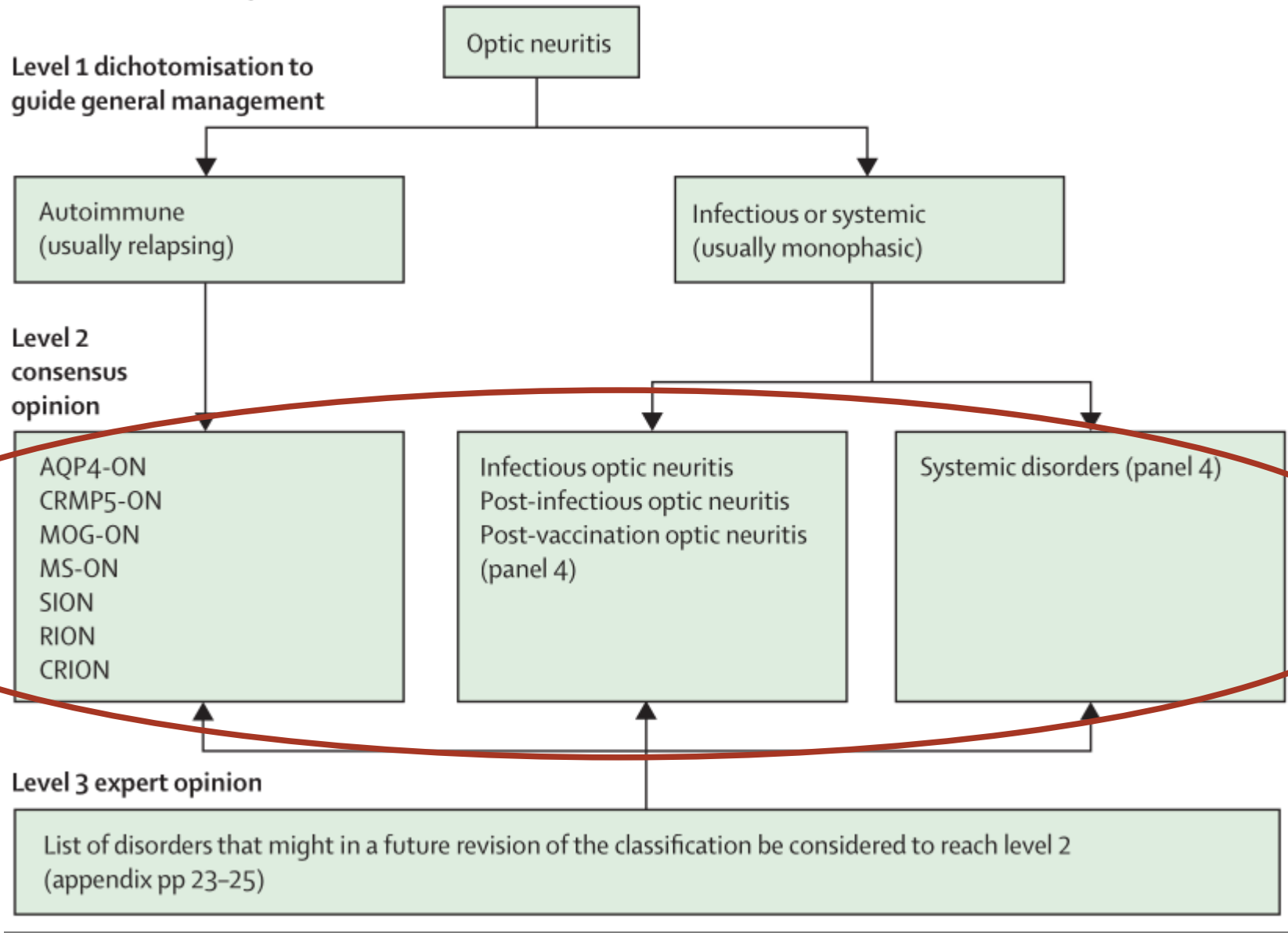


Intrerdisciplinär

Geographisch

Klassifizierung

B Classification of optic neuritis



1. Fall

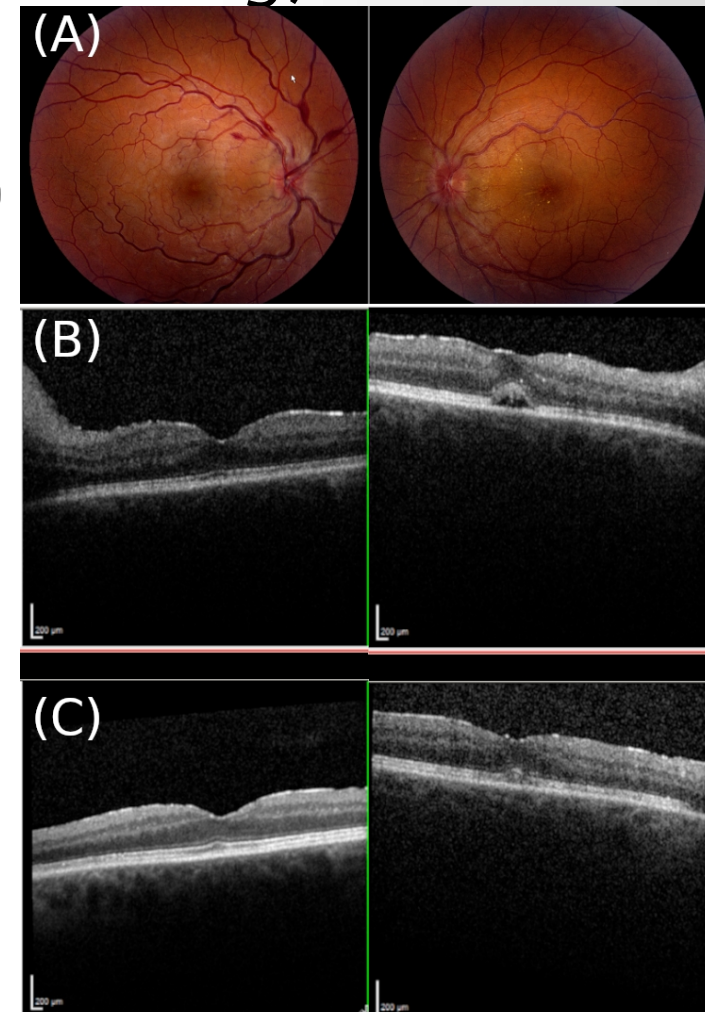
- 34-jährige kaukasische Patientin
- 7 Tage Schmerzen OD die verschlimmern mit Augenbewegungen
- Dyschromatopsie und VA OD 6/9, OS 6/5
- RAPD OD
- Müdigkeit, kognitive Probleme, Harninkontinenz, Depression
- vor 3 Jahren rechtsseitiges Taubheitsgefühl
- MRT: DIS & DIT & 3 Gd+ nicht-symptomatische Läsionen

2. Fall

- 28 Jahre alt, afrokaribischer Mann
- Schmerzloser Sehverlust OS (6/38)
- Dyschromatopsie
- RAPD OS
- Mehrere Episoden die auf Behandlung mit Corticosteroiden ansprechen über ~20 Jahre
- OCT: pRNFL-Atrophie OS (IEPD >5%)
- MRT geschwollener, kontrastaufnehmender Sehnerv
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 $>4\mu\text{m}$ 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).

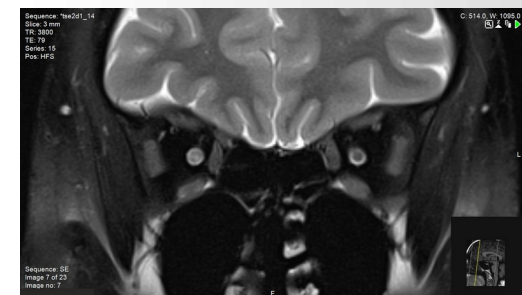
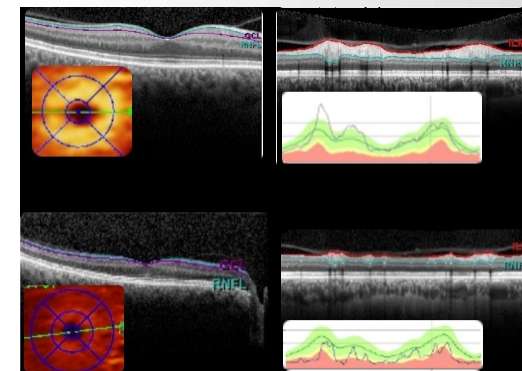
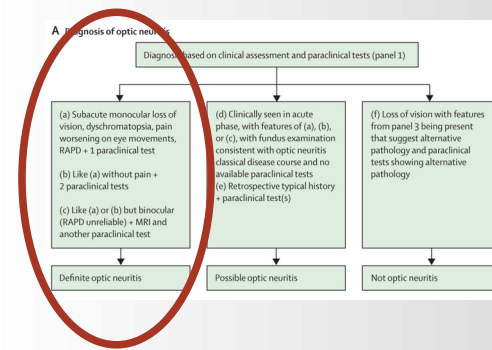
Application of the clinical and paraclinical criteria

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



Konsensus Zeitintervalle

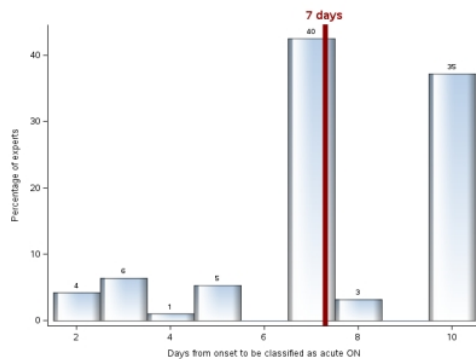
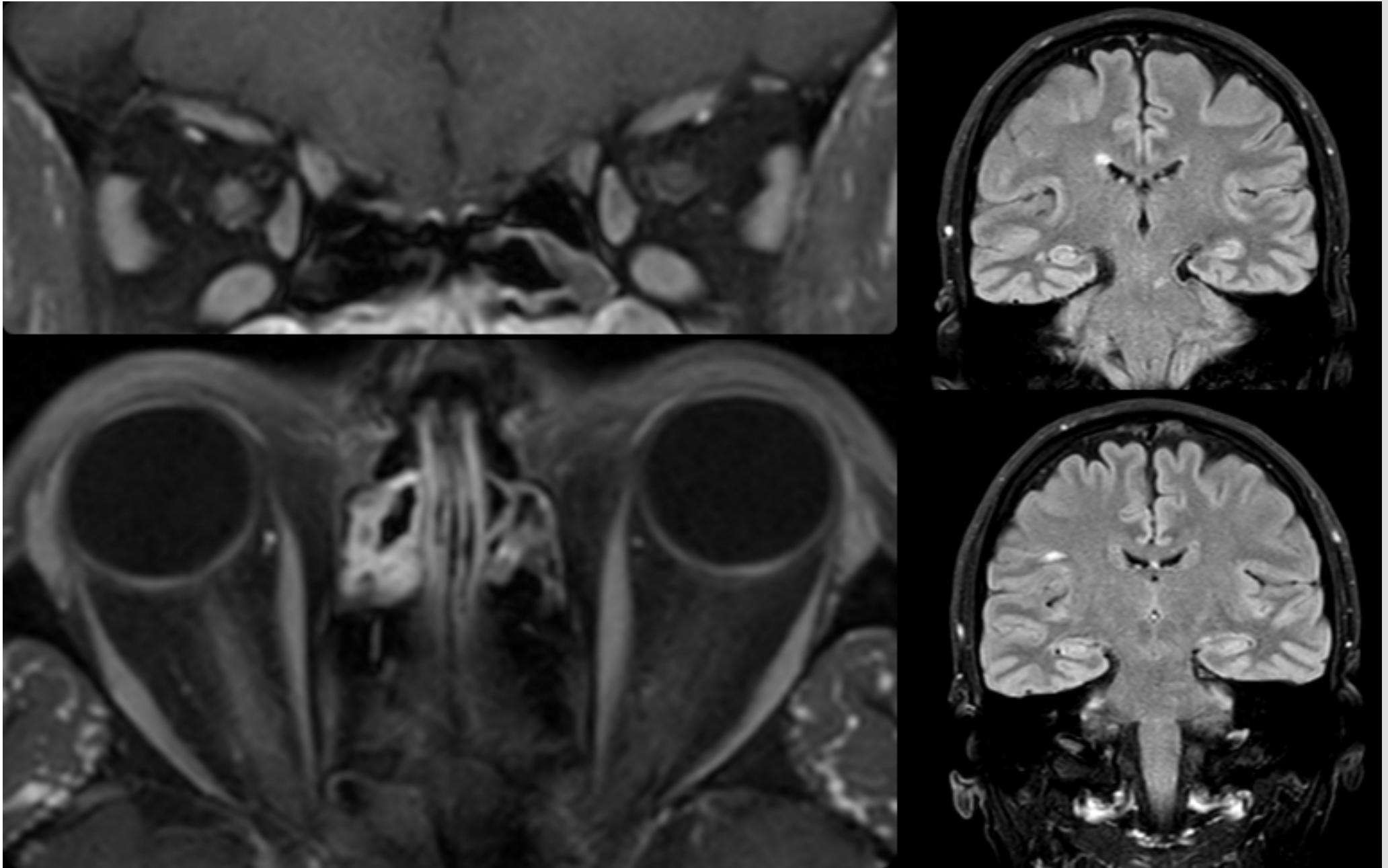


Figure 20: The definition of acute optic neuritis was considered. A 96% consensus was reached for the majority choice of a 7 day interval in the next Delphi round (#23).

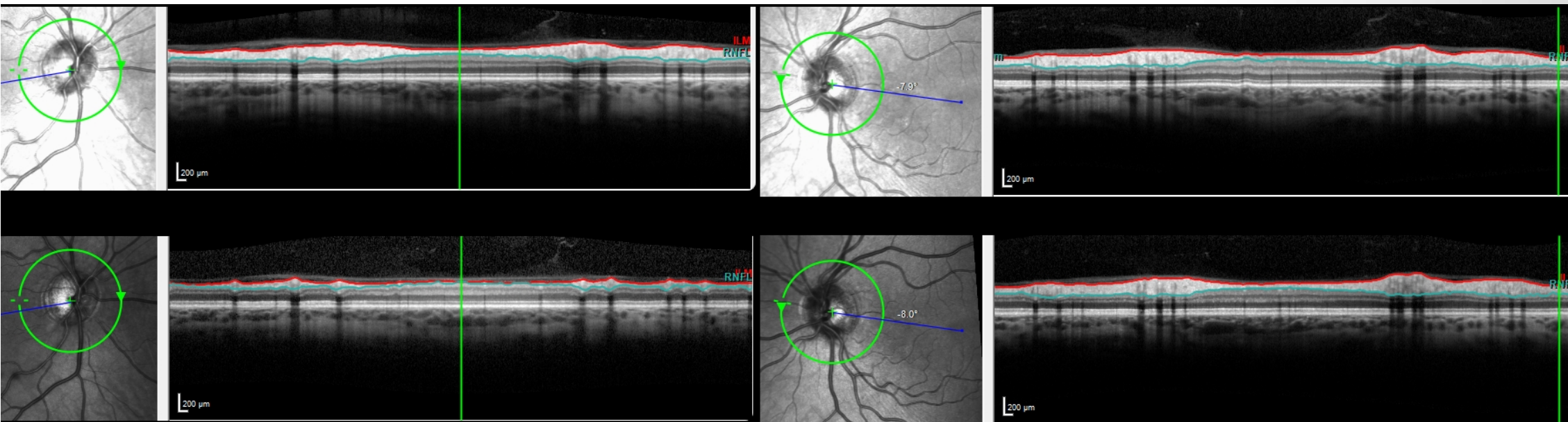
- Akut: < 7 Tage
- Subakut: 1-12 Wochen
- Spät: > 3 Monate

- OCT: Corresponding optic disc swelling acutely or an inter-eye difference of mGCIPL of $> 4\%$ or $> 4\mu\text{m}$ or pRNFL $> 5\%$ or $> 5\mu\text{m}$ within ≥ 3 months after onset
- MRI: Contrast enhancement of the symptomatic optic nerve and sheaths acutely or intrinsic signal increase (looking brighter) within ≥ 3 months

Neuritis nervi optici: MRT akut

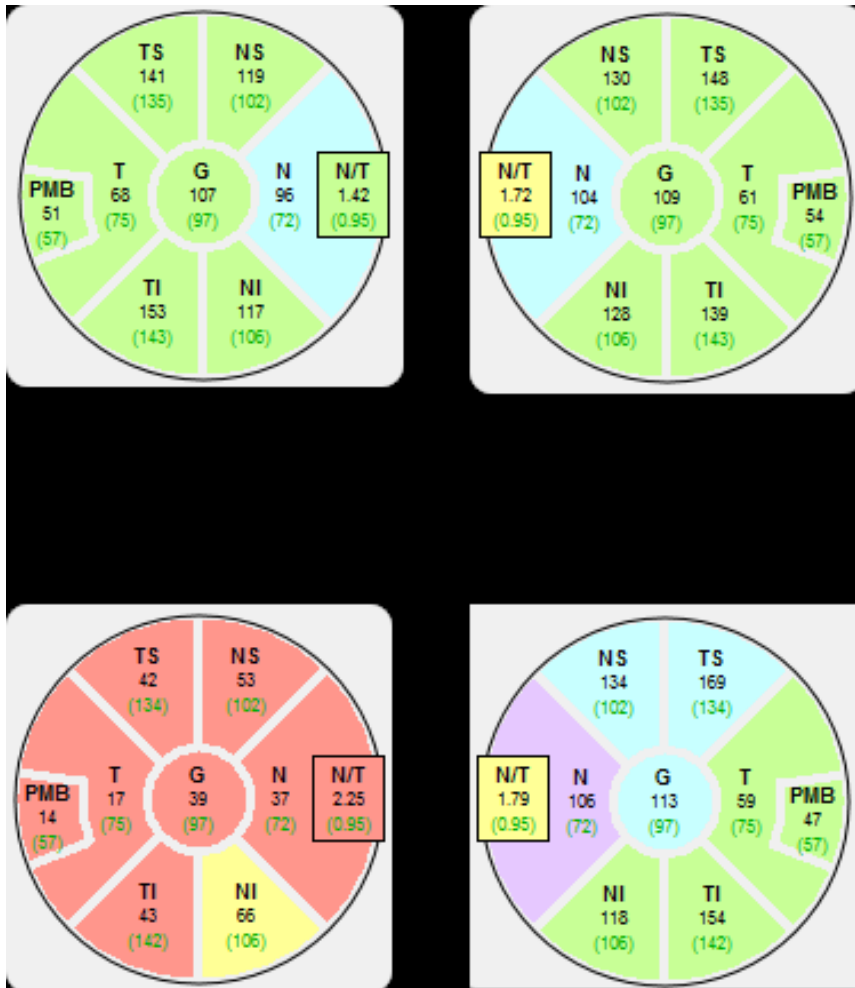


OCT Papille akut & spät



41 Jahre alter Mann, 3 Tage nach simultanem Beginn von Schmerzen bei Augenbewegungen rechts mit Visusverlust. VA (logMAR) OD 0.16, OS 0.63, Ishihara OD 10/15, OS 15/15, RAPD OD, pVEP OD absent, OS 118 ms

Neuritis nervi optici: spät



Retinal Asymmetrie
im akutem Stadium:

IEPD 2 %

IEAD 2 μm

-
- im spätem Stadium:
- IEPD 65 %
- IEAD 74 μm

OCT macula akut & spät

Akut:

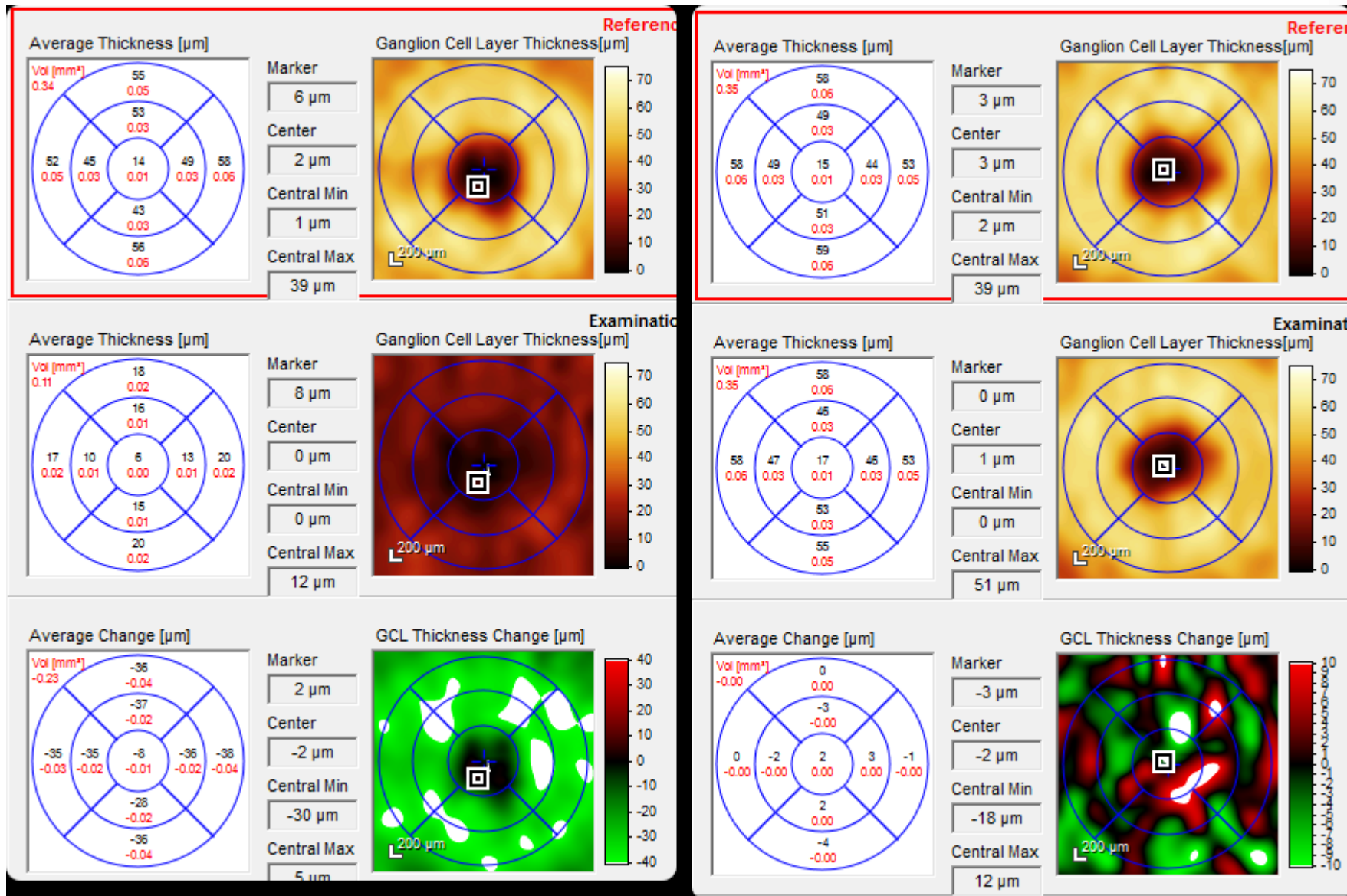
3 %

0.1 mm³

Spät:

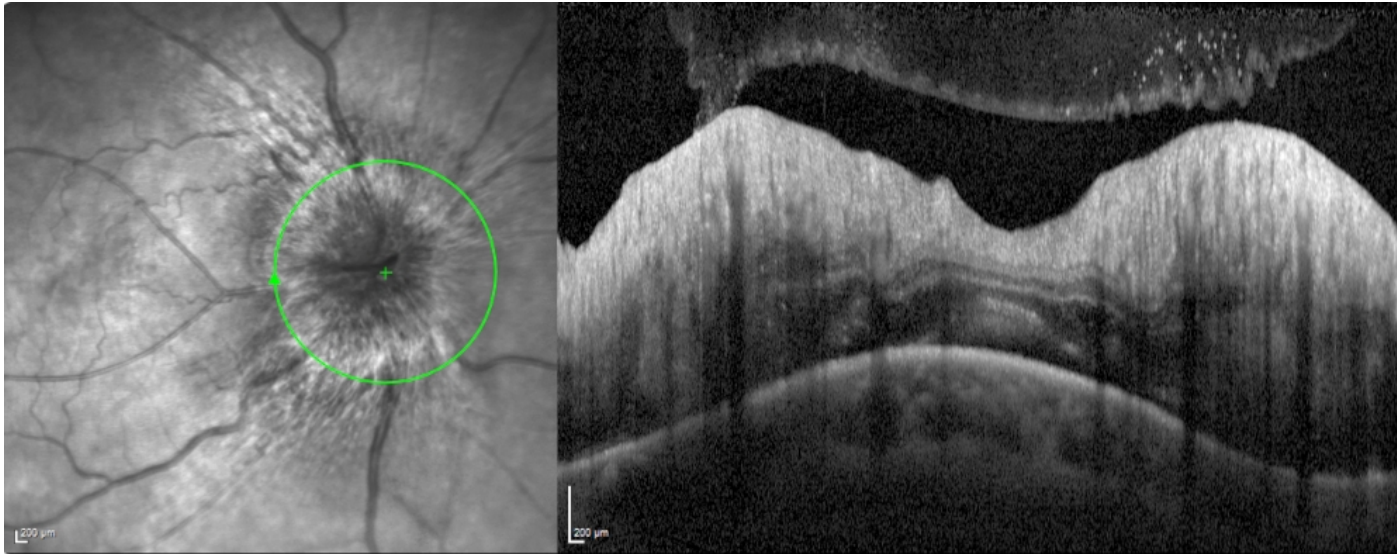
69 %

0.24 mm³



Tip: Prozente (%) sind einfacher, schneller, geräteunabhängig und präziser als absolute Werte.

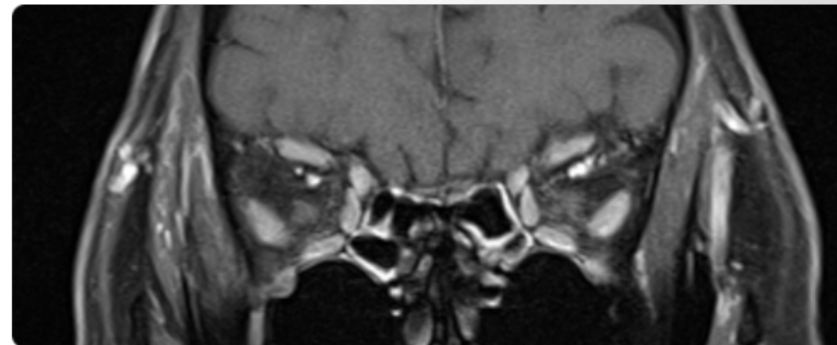
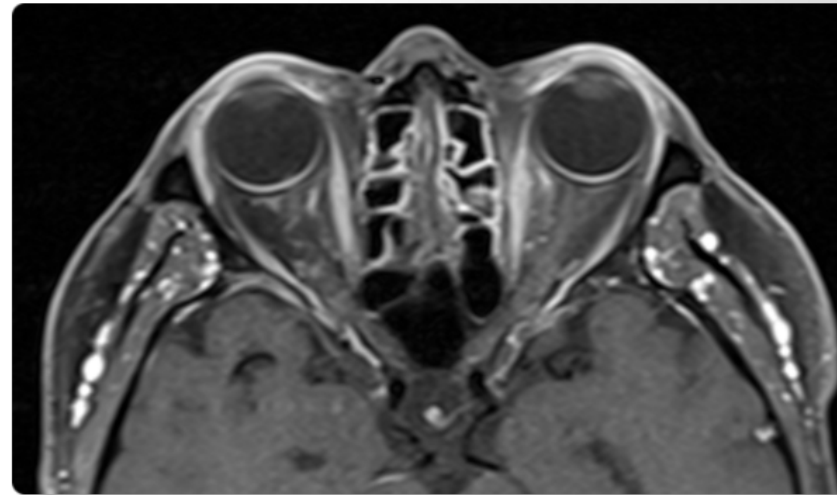
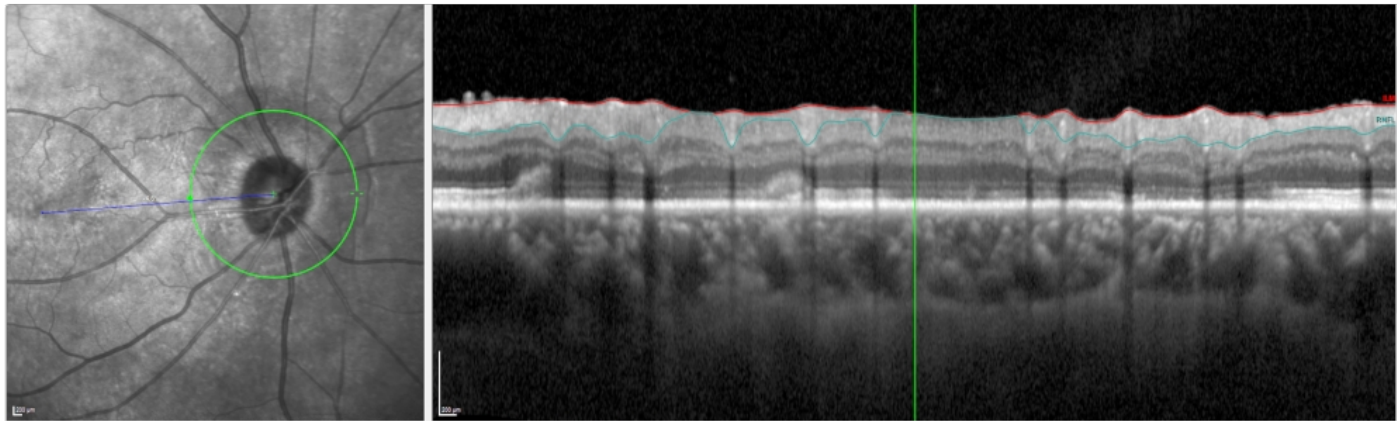
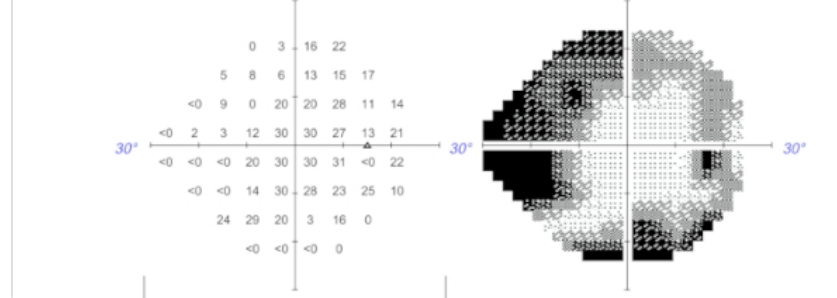
Der Sonderfall



Fixation Monitor: Gaze/Blind Spot
 Fixation Target: Central
 Fixation Losses: 3/9 XX
 False POS Errors: 7%
 False NEG Errors: 26%
 Test Duration: 05:33
 Fovea: 35 dB

Stimulus: III, White
 Background: 31.5 asb
 Strategy: SITA FASTER
 Pupil Diameter:
 Visual Acuity:
 Rx: +3.25 DS

Date: Jul 26, 2024
 Time: 09:05
 Age: 66

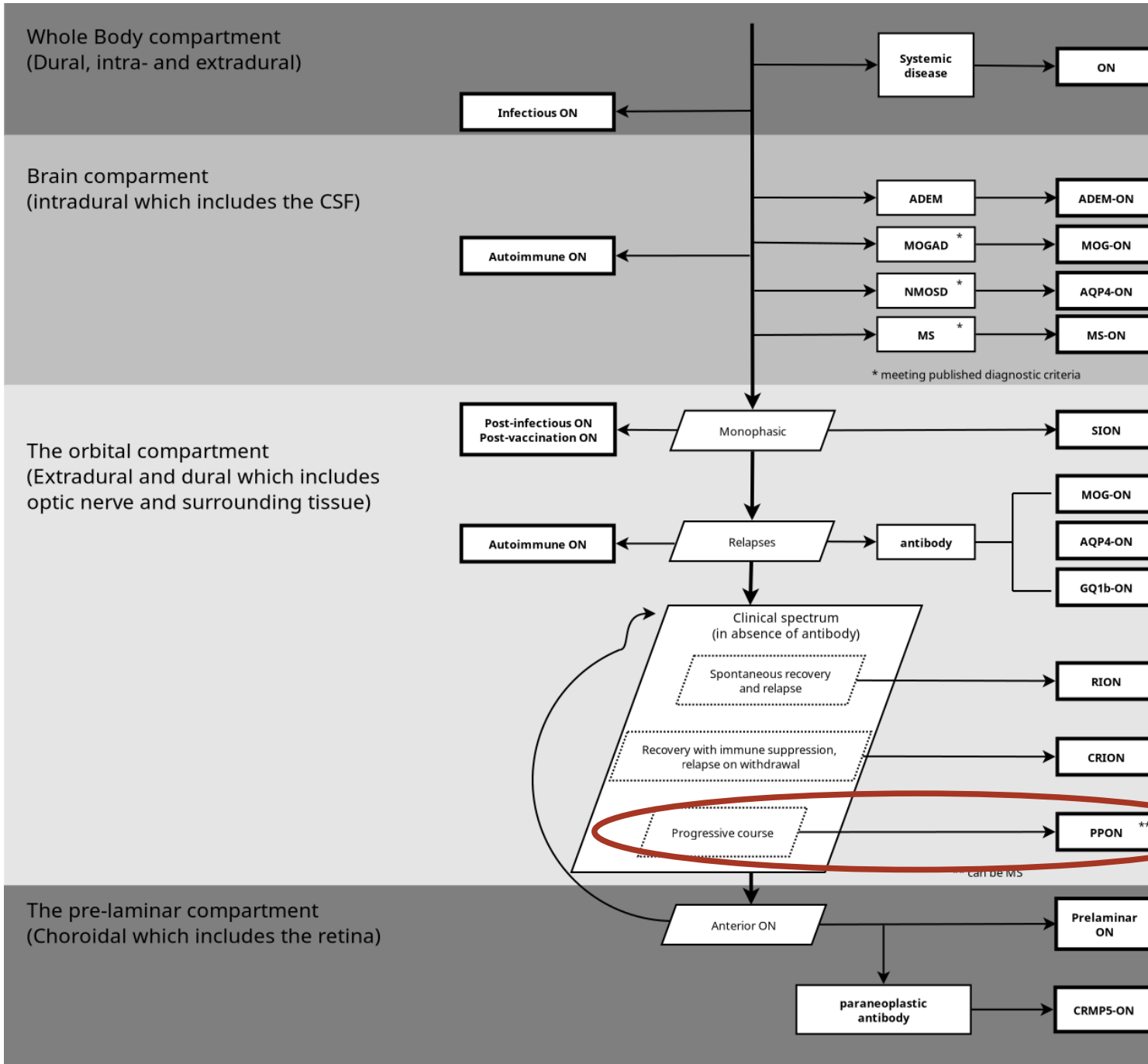


Pre-läminare neuritis nervi optici

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.

Anatomie



Neurologen



Augenärzte

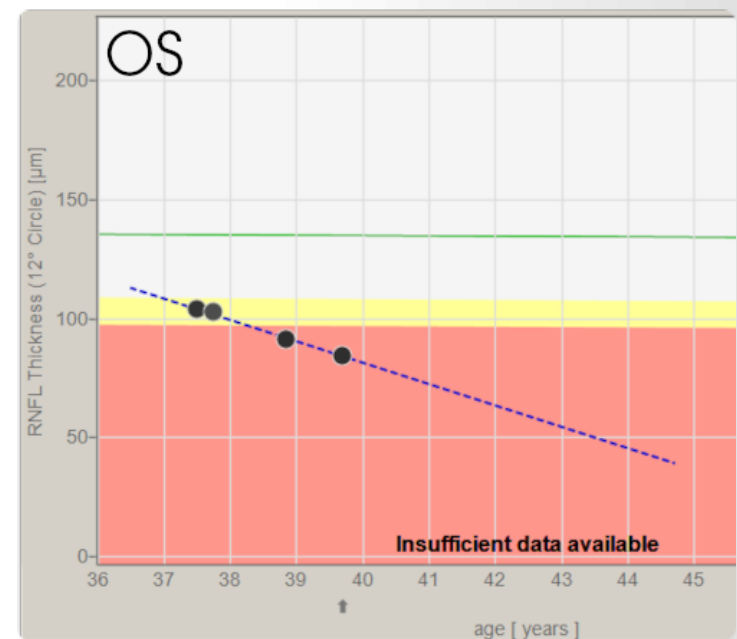
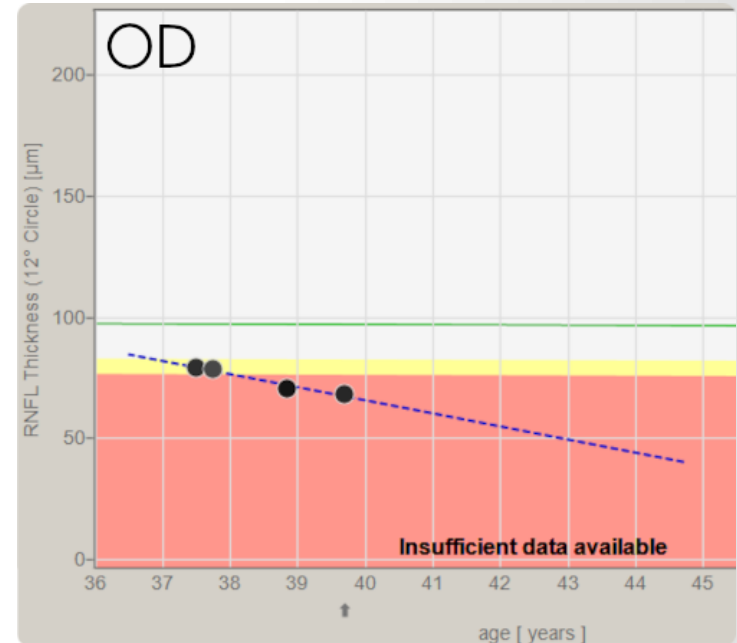
Primär progressive neuritis nervi optici

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 than a relapsing disease course.

Primär progressive neuritis nervi optici

- MS im 27. Lebensjahr, 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 stabil



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
- Akut/spät: Zeitpunkte / Retinale asymmetrie (%)
- Primär progressive neuritis nervi optici
- Zukünftige ICON Revisionen sind geplant

Herzlichen Dank

