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39	A	B	C	D	E

PRACTICE QUESTIONS

Consortium of Multiple Sclerosis Centers (CMSC)

CRND

Certification Examination in Rare Neuroimmunologic Disorders

EDITION

Demo

QUESTIONS

9

FORMAT

Limited Edition PDF

Before you begin

What this is

9 practice questions for Consortium of Multiple Sclerosis Centers (CMSC) CRND , each with its answer key and an explanation. Answer a question before you read its key — the explanation is worth more once you have committed.

If something looks wrong

Send us three things and we will check it:

- the exam code, CRND
- the question number
- the email address on your order

Write to **support@mometrix.net**. We reply within one business day.

QUESTION 1

MULTIPLE CHOICE

Which of the following clinical and serologic features support a diagnosis of aquaporin-4 (AQP4) antibody-positive NMOSD? (Choose two.)

- ☒ **A** Area postrema syndrome with intractable hiccups or nausea and vomiting
- ☐ **B** Brain MRI showing diffuse confluent periventricular white matter lesions consistent with multiple sclerosis
- ☒ **C** Longitudinally extensive transverse myelitis (LETM) spanning three or more vertebral segments
- ☐ **D** Oligoclonal bands restricted to the cerebrospinal fluid
- ☐ **E** Isolated optic neuritis with normal brain and spinal cord MRI and negative AQP4-IgG

ANSWER A - C

WHY Area postrema syndrome (intractable nausea/vomiting or hiccups) and longitudinally extensive transverse myelitis are core clinical characteristics of NMOSD in AQP4-IgG-positive disease. Diffuse periventricular white matter lesions are more typical of multiple sclerosis, and CSF-restricted oligoclonal bands are also characteristic of MS, not NMOSD. Isolated optic neuritis with normal MRI and negative AQP4-IgG does not meet current diagnostic criteria for NMOSD.

QUESTION 2

MULTIPLE CHOICE

Which of the following characteristics are most consistent with MOG-IgG–associated disease (MOGAD)? (Choose two.)

- ☒ **A** Optic neuritis that is often bilateral or recurrent, with optic nerve MRI showing long-segment enhancement
- ☐ **B** Positive AQP4-IgG on cell-based assay
- ☒ **C** ADEM-like presentations, particularly in children, with poorly demarcated deep gray matter and juxtacortical lesions
- ☐ **D** Brain MRI demonstrating central vein sign on susceptibility-weighted imaging
- ☐ **E** Spinal cord lesions that are typically short-segment (<3 vertebral segments) and laterally located

ANSWER **A - C**

WHY MOGAD commonly presents with bilateral or recurrent optic neuritis with longitudinally extensive optic nerve enhancement, and in children with ADEM-like syndromes including poorly demarcated deep gray and juxtacortical lesions. AQP4-IgG positivity defines AQP4-positive NMOSD, not MOGAD. The central vein sign is a marker of multiple sclerosis. Short-segment, laterally placed cord lesions are more typical of MS.

QUESTION 3

SINGLE CHOICE

In a patient with a single clinical episode of optic neuritis and a positive AQP4-IgG cell-based assay, what is the minimum additional requirement to diagnose NMOSD under the 2015 International Panel for NMO Diagnosis (IPND) criteria?

- ☐ A At least one additional core clinical characteristic
- ☐ B MRI evidence of a longitudinally extensive spinal cord lesion
- ☐ C CSF oligoclonal bands
- ☒ D No additional requirement

ANSWER D

WHY Under the 2015 IPND criteria, a positive AQP4-IgG cell-based assay plus at least one core clinical characteristic (optic neuritis, acute myelitis, area postrema syndrome, acute brainstem syndrome, symptomatic narcolepsy/diencephalic syndrome, or symptomatic cerebral syndrome) is required. A patient with a single clinical episode (e.g., optic neuritis) and positive AQP4-IgG meets criteria with just that one core characteristic — no additional core clinical event, spinal cord lesion, or CSF finding is required.

QUESTION 4

SINGLE CHOICE

Which MRI finding is most characteristic of MOG-IgG–associated optic neuritis?

- ☐ A Short-segment perioptic nerve enhancement limited to the optic nerve head
- ☒ B Longitudinally extensive optic nerve enhancement involving more than half the optic nerve length, often bilateral
- ☐ C Chiasmal enhancement with extension into the hypothalamus
- ☐ D Isolated optic tract involvement without nerve enhancement

ANSWER B

WHY MOGAD optic neuritis classically shows longitudinally extensive enhancement of the optic nerve, frequently involving more than half the nerve length, and is commonly bilateral or sequentially recurrent. Chiasmal/hypothalamic extension is more typical of AQP4-positive NMOSD. Isolated optic tract involvement is uncommon.

QUESTION 5

SINGLE CHOICE

Which serologic assay methodology is currently preferred for detecting AQP4-IgG in suspected NMOSD?

- ☐ A Indirect immunofluorescence on rodent tissue substrate
- ☐ B Enzyme-linked immunosorbent assay (ELISA) using linearized AQP4 peptides
- ☒ C Cell-based assay (CBA) expressing full-length human AQP4
- ☐ D Western blot using M23-isoform recombinant protein

ANSWER C

WHY The cell-based assay (CBA) using full-length human AQP4 (M1 or M23 isoform) is the most sensitive and specific method for detecting AQP4-IgG and is recommended by the IPND as the preferred assay. Tissue-based immunofluorescence and ELISA have lower sensitivity; Western blot is not recommended.

QUESTION 6

SINGLE CHOICE

Which of the following is a recognized core clinical characteristic of NMOSD?

- ☒ A Area postrema syndrome
- ☐ B Progressive cognitive decline
- ☐ C Persistent hemiplegic migraine with aura
- ☐ D Subacute combined degeneration of the spinal cord

ANSWER A

WHY Area postrema syndrome (intractable nausea, vomiting, or hiccups lasting >48 hours) is one of the six core clinical characteristics of NMOSD defined in the 2015 IPND criteria. Progressive cognitive decline, hemiplegic migraine, and subacute combined degeneration are not core features of NMOSD.

QUESTION 7 SINGLE CHOICE

Which therapy is FDA-approved for aquaporin-4 antibody–positive NMOSD as a long-term relapse-prevention treatment?

- ☐ A Dimethyl fumarate
- ☒ B Eculizumab (anti-complement C5)
- ☐ C Interferon beta-1a
- ☐ D Glatiramer acetate

ANSWER B

WHY Eculizumab, a monoclonal antibody against complement C5, is FDA-approved for AQP4-IgG–positive NMOSD based on the PREVENT trial demonstrating significant relapse reduction. The other agents listed are disease-modifying therapies for multiple sclerosis and have not shown efficacy — and may be harmful — in NMOSD.

QUESTION 8 SINGLE CHOICE

Which feature most reliably distinguishes an MOGAD transverse myelitis lesion from an NMOSD longitudinally extensive transverse myelitis (LETM) lesion?

- ☐ A Cord swelling and T2 hyperintensity are greater in NMOSD than in MOGAD
- ☒ B MOGAD lesions preferentially involve the conus medullaris and often have central (H-shaped) gray matter involvement
- ☐ C MOGAD lesions are always less than one vertebral segment in length
- ☐ D MOGAD lesions characteristically enhance for more than 6 months

ANSWER B

WHY MOGAD myelitis characteristically involves the conus medullaris and demonstrates central gray matter (H-shaped) involvement on axial imaging, whereas AQP4-positive NMOSD LETM typically involves central gray matter across long segments of the cervicothoracic cord. MOGAD lesions can be LETM as well, so segment length is not the defining distinction. Persistent enhancement for months is atypical.

QUESTION 9

SINGLE CHOICE

Which clinical syndrome is most strongly associated with antibodies against the N-methyl-D-aspartate (NMDA) receptor?

- ☐ A Limbic encephalitis with prominent memory loss and seizures
- ☒ B Encephalitis with prominent psychiatric symptoms, dyskinesias, autonomic instability, and decreased level of consciousness
- ☐ C Subacute cerebellar degeneration with ataxia and nystagmus
- ☐ D Brainstem encephalitis with intractable nausea and hiccups

ANSWER B

WHY Anti-NMDA receptor encephalitis classically presents with a multistage syndrome beginning with psychiatric symptoms, followed by memory dysfunction, seizures, dyskinesias (especially orofacial), autonomic instability, and decreased consciousness. Limbic predominance is more characteristic of LGI1 or GAD65 encephalitis; subacute cerebellar degeneration is associated with anti-mGluR1 or anti-Yo antibodies; intractable nausea/hiccups (area postrema syndrome) is a feature of AQP4-positive NMOSD.