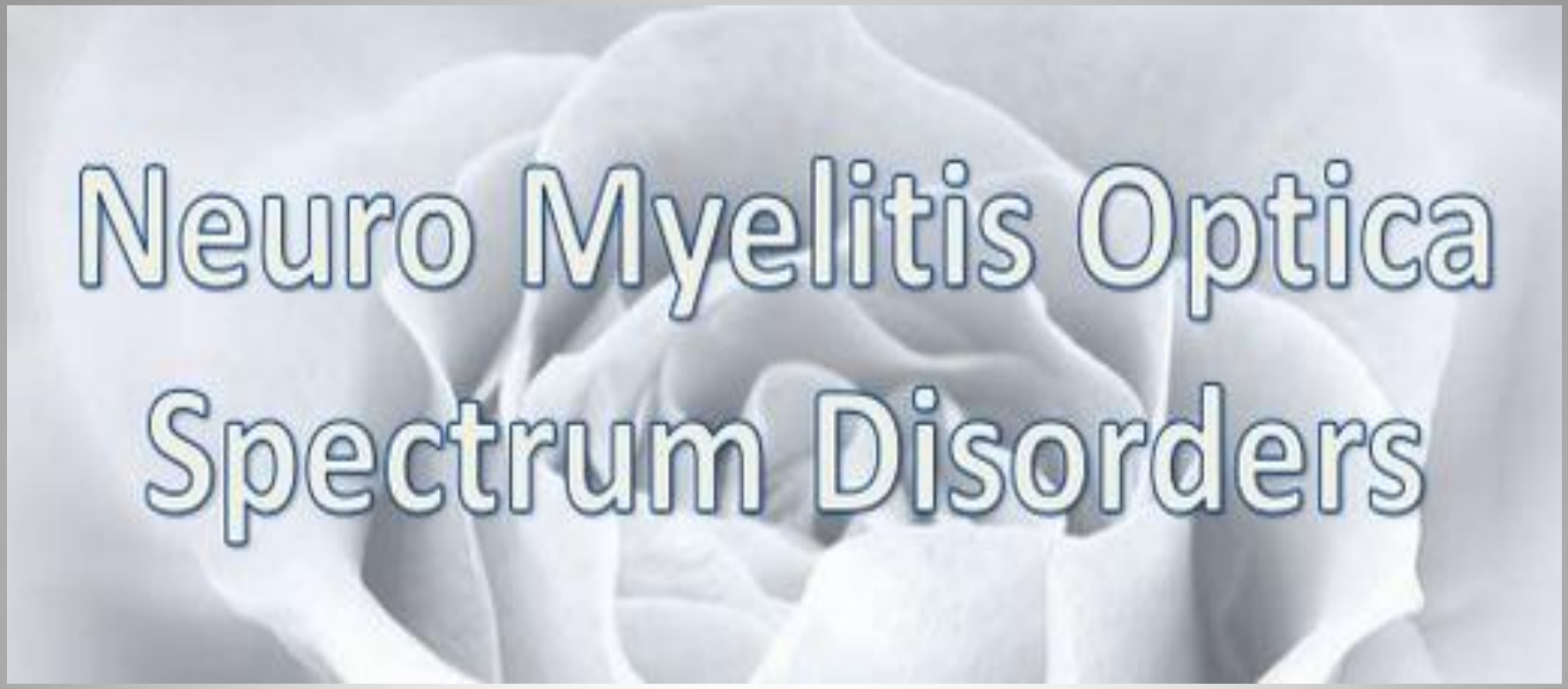


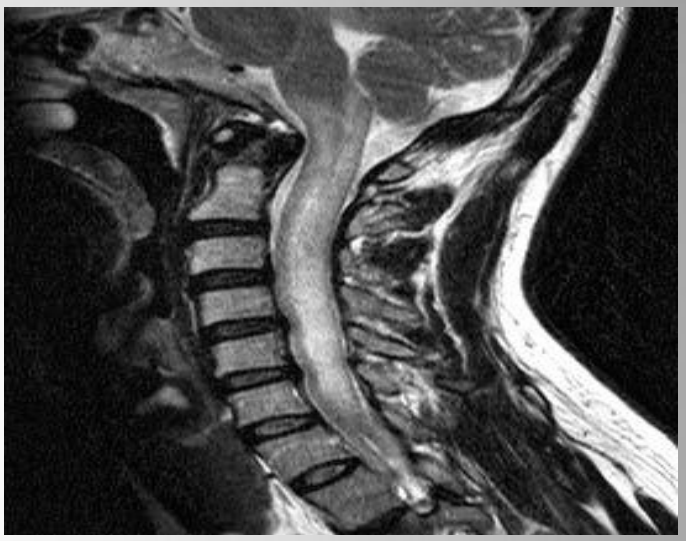
TIÊU CHUẨN CHẨN ĐOÁN

PGS.TS Cao Phi Phong

Cập nhật -2018

NMO Criteria (2006)

Tiêu chuẩn chẩn đoán NMO
(Wingerchuk et al. Neurology. 2006)



Định nghĩa NMO

Two Absolute criteria:

1) Optic Neuritis

2) Transverse Myelitis

≥3 vertebral segments (LETM)

And at least 2 of 3 supportive criteria

1) Presence of continuous spinal cord MRI lesion extending over three or more vertebral segments.

2) MRI brain not satisfying diagnostic criteria for Multiple Sclerosis.

3) Aquaporin-4 antibody present in serum.

NMO Spectrum Disorder

Optic neuritis OR Transverse myelitis

AND presence of aquaporin- 4 antibodies in blood

Triệu chứng, test chẩn đoán và điều trị NMO và NMOSD rất giống nhau hay tương tự và thường gọi chung là NMOSD

Tại sao tiêu chuẩn chẩn đoán 2006 không đầy đủ trong năm 2014 ?

- ❑ Discovery of NMO-IgG
- NMO can be recognized reliably at an earlier point
- ❑ Limited versions of NMO
- recurrent myelitis or recurrent optic neuritis
- ❑ Brain lesions may occur
- may be the presenting manifestations
- may be highly suggestive or diagnostic
- ❑ Co-association of other autoimmune conditions
- Do they exclude NMO

International Panel for NMO Diagnosis (IPND)

Convened October, 2011

Co-chairs: Dean Wingerchuk

Brian Weinshenker

REVISED DIAGNOSTIC CRITERIA:

NMOSD with AQP4-IgG

Nguyên tắc đồng thuận ban đầu IPND

Chẩn đoán lâm sàng

AQP4 antibodies

Không đủ (Not sufficient)

False positives

Population-based study of MS-like illness: clinical NMO 0.2%; CBA+ 0.3%; ELISA+ 0.7% *(Pittock et al. 2014 JAMA Neurol.doi:10.1001/jamaneurol.2014.1581)*

Không đòi hỏi (Not required)

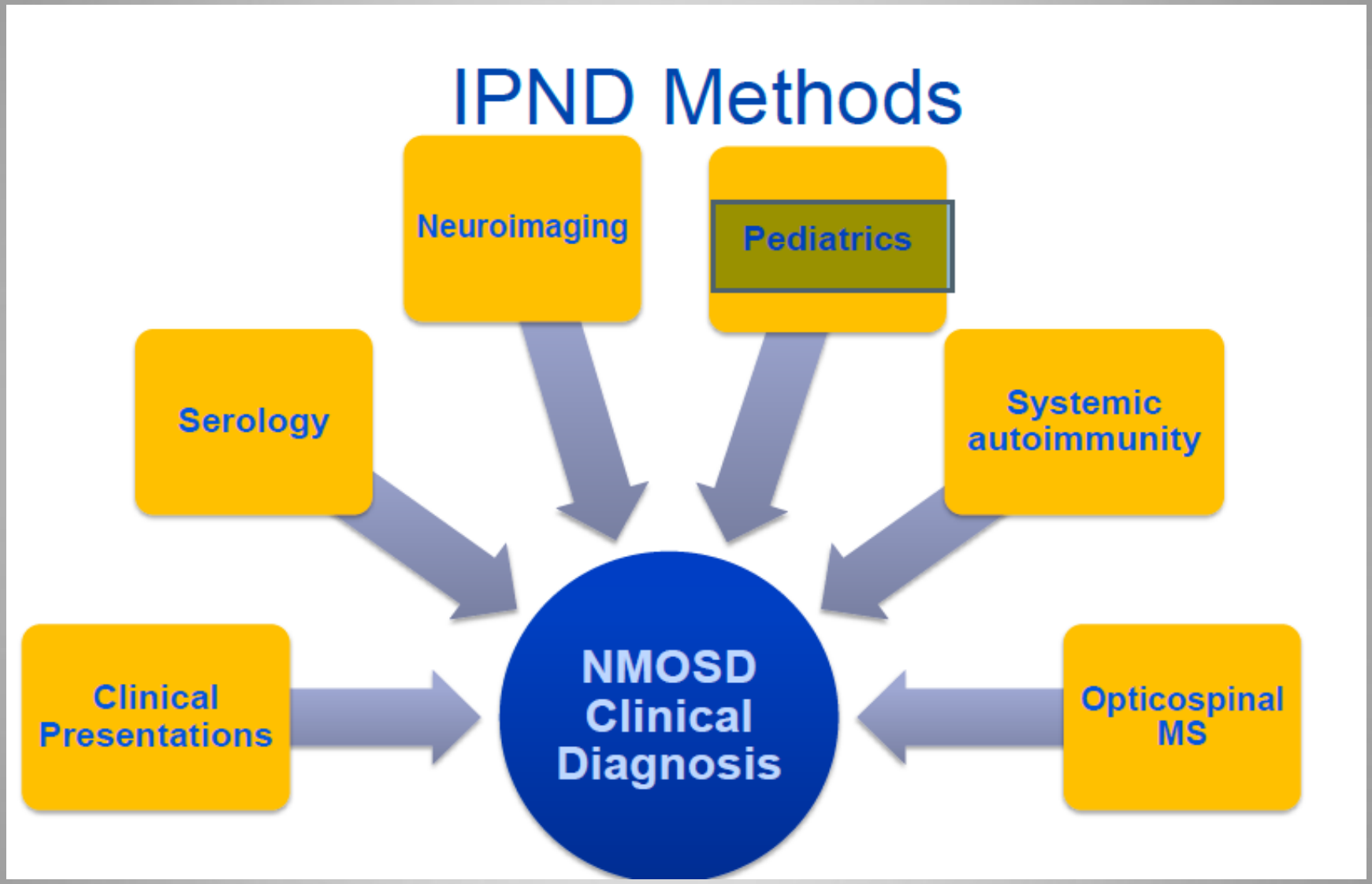
Sự khác biệt giữa seronegative và seropositive ranh giới rất thấp

Sử dụng bằng chứng có thể dùng được tốt nhất và đồng thuận của hội thảo

Phương pháp

18 members from 9 countries

6 Working Groups Specific charges relevant to NMO diagnosis


IPND Methods
Neuroimaging
Pediatrics
Serology
Systemic autoimmunity
Clinical Presentations
NMOSD Clinical Diagnosis
Opticospinal MS

Kết quả:

Nomenclature (thuật ngữ)

NMOSD: thuật ngữ thống nhất (unified term)

Phân tầng tình trạng huyết thanh

- *NMOSD with AQP4-IgG*
- *NMOSD without AQP4-IgG (or testing unavailable)*

- *Allows for future revisions*

e.g. discovery and validation of other antibodies associated with NMOSD clinical phenotype

Diagnostic Criteria for Neuromyelitis Optica 2014

Tiêu chuẩn sửa đổi: NMOSD AQP4-IgG (+)

Đòi hỏi (Requirements):

1. *At least 1 core clinical characteristic (ít nhất một triệu chứng lâm sàng đặc trưng)*
2. *Positive test for AQP4-IgG*
3. *No better explanation ((không thể giải thích tốt hơn)*

   - Clinical and MRI red flags (dấu hiệu báo hiệu lâm sàng và MRI)

Core Clinical Characteristics

1. *Optic neuritis*
2. *Acute myelitis*
3. *Area postrema syndrome: nausea/vomiting/hiccups*
4. *Other brain stem syndrome*
5. *Symptomatic narcolepsy or acute diencephalic syndrome with MRI lesion(s)*
6. *Symptomatic cerebral syndrome with MRI lesion(s)*

Revised Diagnostic Criteria:
NMOSD with AQP4-IgG

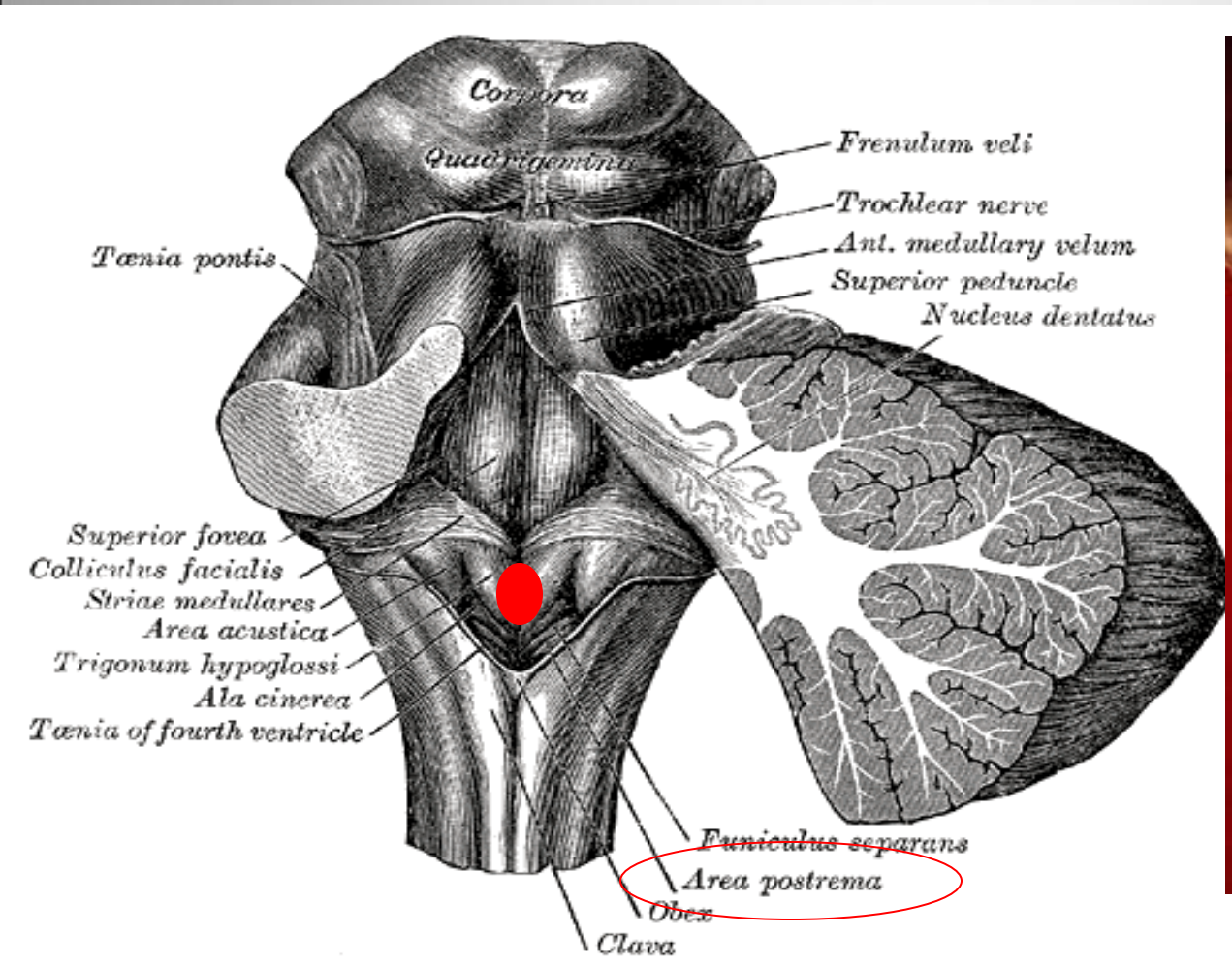
Requirements

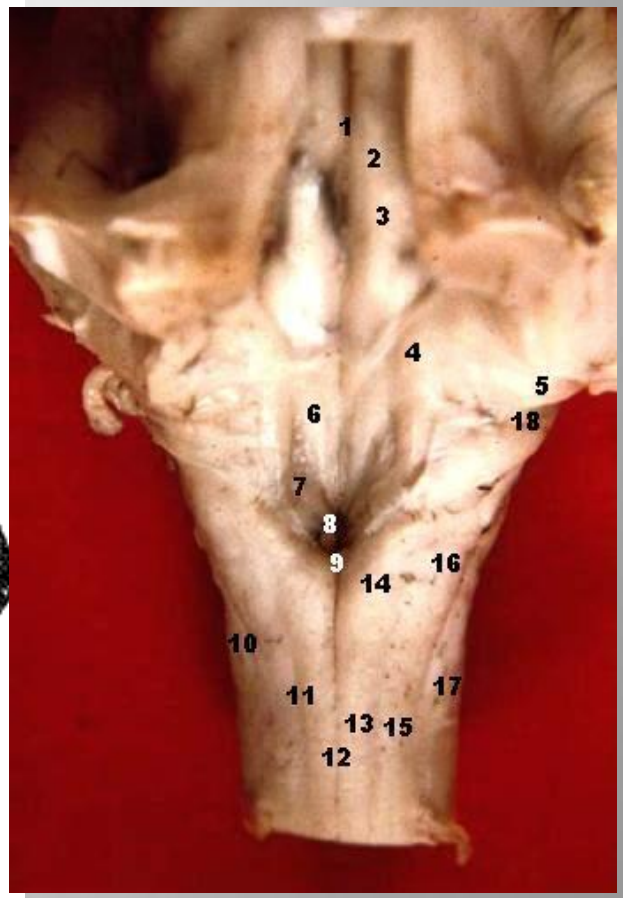
- At least 1 core clinical characteristic
- Positive test for AQP4-IgG
- No better explanation
  - Clinical and MRI red flags

Core Clinical Characteristics

- Optic neuritis
- Acute myelitis
- Area postrema syndrome:
  - nausea/vomiting/hiccups
- Other brain stem syndrome
- Symptomatic narcolepsy or acute diencephalic syndrome with MRI lesion(s)
- Symptomatic cerebral syndrome with MRI lesion(s)

*The **area postrema** is a medullary structure in the brain that controls vomiting. Its privileged location in the brain also allows the area postrema to play a vital role in the control of autonomic functions by the central nervous system (vị trí đặc quyền trong não)*





Tiêu chuẩn sửa đổi:

NMOSD AQP4-Ig (-) hay không có xét nghiệm

***At least 2 core clinical characteristics all satisfying**:*

- *1 of ON, myelitis, or area postrema syndrome*
- *Dissemination in space (lan tỏa theo không gian)*

Isolated recurrent ON or recurrent TM do not qualify

- *Additional MRI requirements (đòi hỏi MRI)*

AP syndrome: dorsal medulla lesion (area postrema)

Myelitis: LETM

ON: normal brain MRI **OR >1/2 ON OR chiasm lesion**

- *Negative test(s) for AQP4-IgG using best available assay, or testing unavailable*

No better explanation for the clinical syndrome

(không giải thích phù hợp hơn cho hội chứng lâm sàng)

Revised Diagnostic Criteria:
NMOSD without AQP4-Ig (or unavailable)

- At least 2 core clinical characteristics all satisfying:
  - 1 of ON, myelitis, or area postrema syndrome
  - Dissemination in space
    - Isolated recurrent ON or recurrent TM do not qualify
  - Additional MRI requirements
    - AP syndrome: dorsal medulla lesion
    - Myelitis: LETM
    - ON: normal brain MRI ***OR*** >1/2 ON ***OR*** chiasm lesion
  - Negative test(s) for AQP4-IgG using best available assay, or testing unavailable
- No better explanation for the clinical syndrome

Red Flags: Clinical and Laboratory

Clinical course/lab more typical of MS or other pathology

Progressive course

Rapid nadir (infarction)

Continual worsening more than 4 weeks from onset

Partial TM without LETM

CSF oligoclonal bands

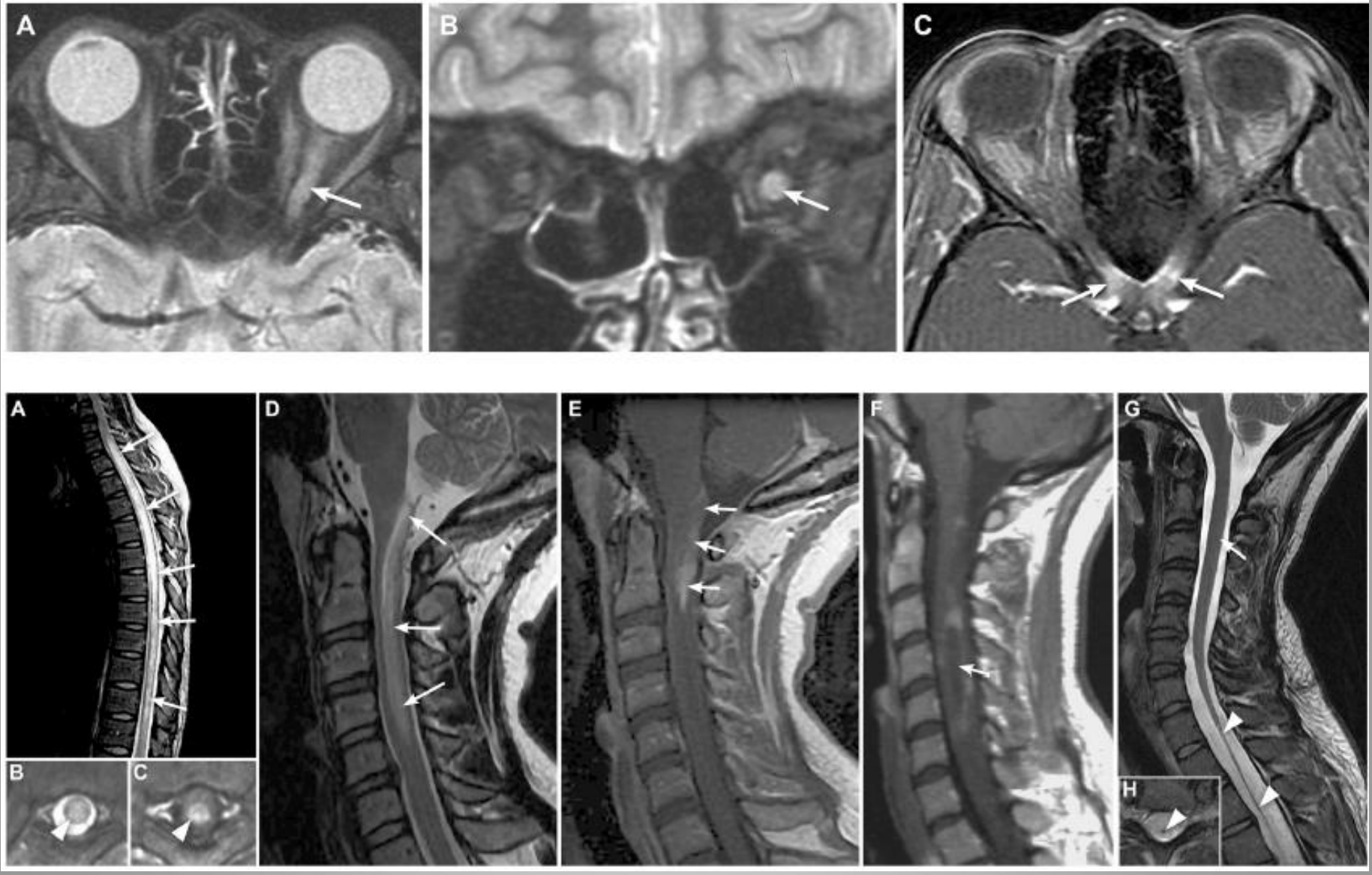
Comorbidity, established or suspected, that mimics NMOSD

Sarcoidosis

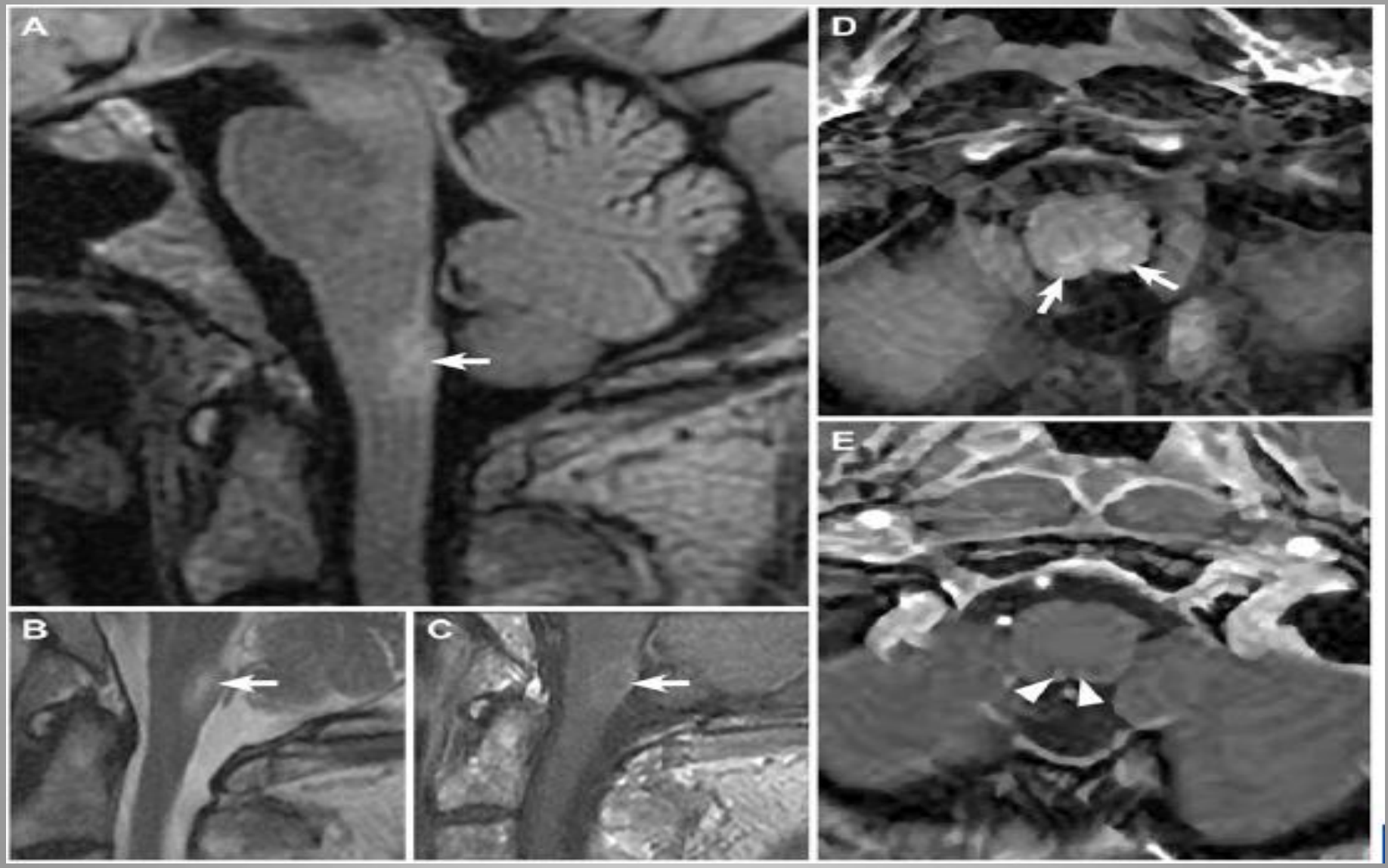
Cancer (lymphoma or CRMP-5 associated ON/myelopathy)

Infection with potential neurologic involvement (e.g., HIV, syphilis)

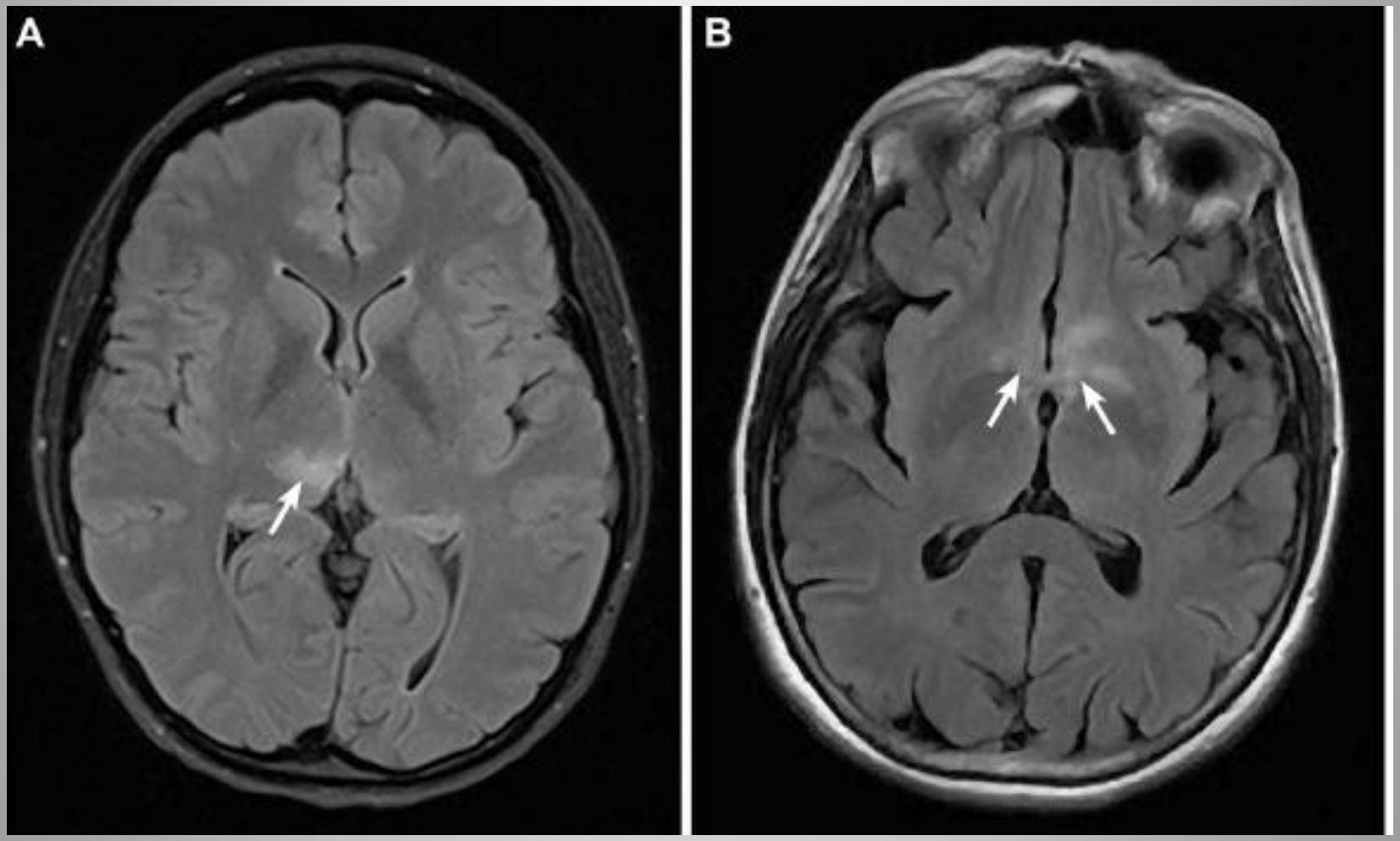
ON and Spinal Cord MRI Lesions



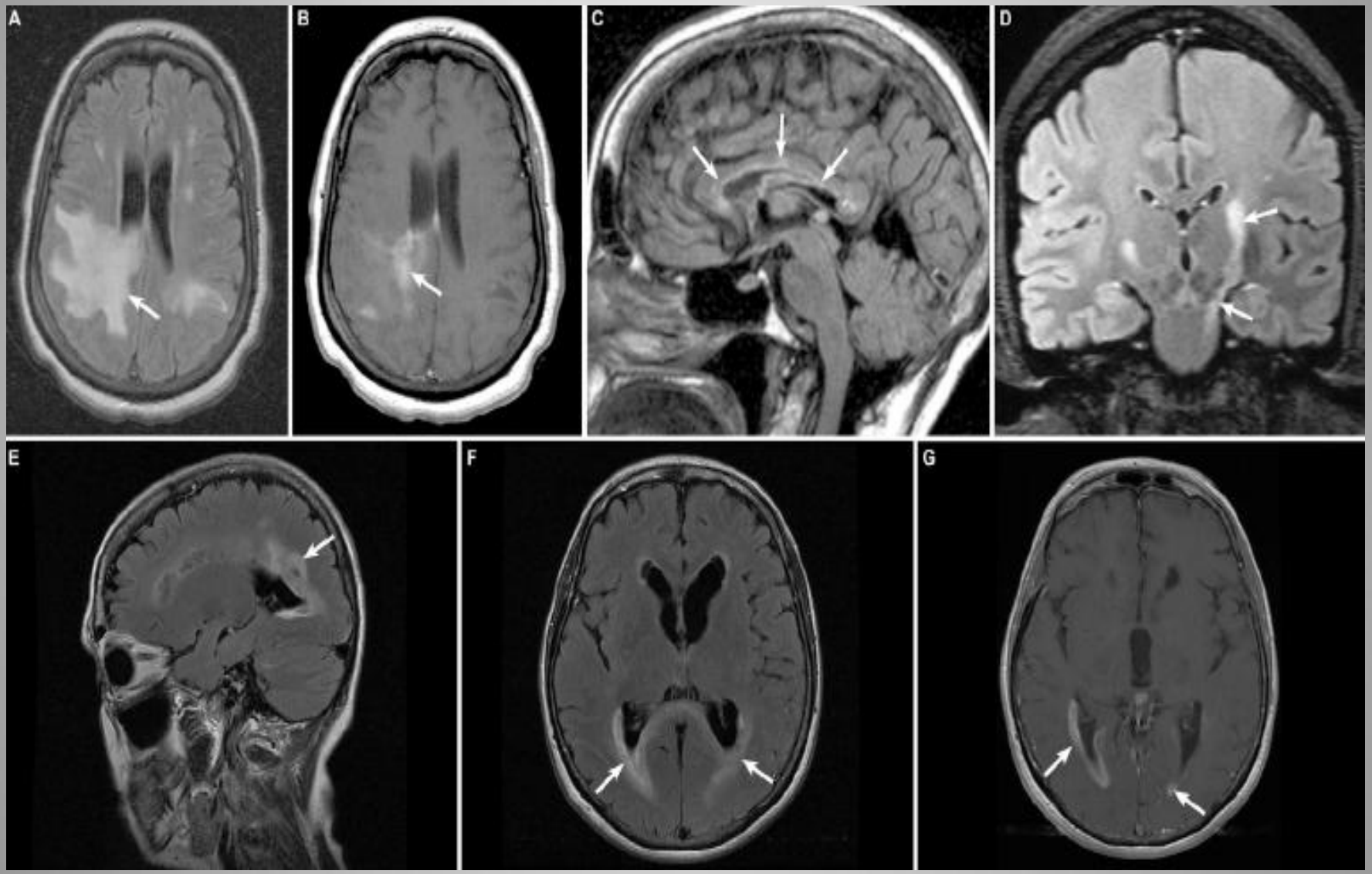
Area Postrema/Dorsal Medulla MRI Lesions



Diencephalic MRI Lesions



Cerebral MRI Lesions



Red Flags:

Radiology

Brain

- ***"MS-typical" lesions***

  "Dawson's fingers"

  adjacent to lateral ventricle temporal lobe

  Juxtacortical lesion(s)

  Cortical lesion(s)

- ***Suspicious of other pathology***

  Lesion(s) with persistent (>3 months) gadolinium enhancemen

Spinal Cord

- ***MS-typical***

  Short cord lesion(s)

  Predominantly (>70%) peripheral cord on axial T2

  Asymptomatic cord lesion(s)

  Diffuse, indistinct T2 signal change (longstanding or progressive MS) (lan tỏa, tín hiệu thay đổi trên T2 mờ)

- ***Suspicious of other pathology***

  Persistent (>3 months) gadolinium enhancement

"Tractopathy" (e.g., paraneoplastic disorder)

Pediatric NMOSD

Tiêu chuẩn giống NMOSD người lớn

Cẩn thận: LETM trong MS trẻ em

Ảnh hưởng trên não nhiều hơn

(Greater incidence of cerebral presentations)

Opticospinal MS

Quan trọng về mặt lịch sử

Confusing terminology a form of MS versus NMO versus something unique?

Similarly defined in Asia, patients have the same disease

"Superseded" terminology (thuật ngữ "thay thế")

Tóm tắt

1. NMOSD thuật ngữ thống nhất cho NMO/NMOSD
2. AQP4-IgG Seropositive: đòi hỏi ít nhất 1 trong 6 triệu chứng trung tâm (core clinical characteristics)
3. AQP4-IgG Seronegative hay không thực hiện được (Unavailable):

- ít nhất 2 triệu chứng trung tâm

  + ON, acute myelitis hay area postrema syndrome

  + MRI

  + lan tỏa theo không gian (Dissemination in space)

KẾT LUẬN

- Neuromyelitis optica bệnh mất myelin riêng biệt có tiêu chuẩn chẩn đoán chính xác

- Nhân khẩu học (Demographic) và bệnh sử tiên đoán tái phát (predict relapses)

- Phòng ngừa tái phát(Relapse prevention) đòi hỏi **broad-spectrum or B-cell-specific immunosuppression**
- **(ức chế miễn dịch hàng loạt hay chuyên biệt tế bào B)**

Cảm ơn sự theo dõi