

- 1) 視力低下が固定した時点で、視力低下が強い方の眼の視力が(0.1)未満。
- 2) 脊髄 MRI で 3 錐体以上の脊髄病変を認めるもの。

**別表 2 : 改訂 NMO 診断基準 (Wingerchuk2006) より改変引用)**

1. 視神経炎
2. 急性脊髄炎
3. 以下の 3 項目のうち 2 つを満たす
  - a) 3 脊椎体以上の長さを有する 脊髄 MRI 病変
  - b) 発症時に脳 MRI 病変が MS 基準を満たさない
  - c) 抗 AQ-4 抗体が末梢血で陽性

**別表 3 : IVIG の免疫作用機序 (中尾 2012) より転載)**

1. 病原的な自己抗体を不活性化する抗 idiotypic 抗体の自己免疫修正
2. 抗体 Fragment c (FC) 受容体のブロックによるリンパ球活性化の抑制と炎症反応抑制
3. B 細胞の抑制による自己抗体産生の抑制
4. 遊離自己抗体の中和
5. 補体活性化や補体の組織・血管への沈着抑制
6. 炎症性サイトカインの産生抑制
7. 中枢神経の髄鞘再形成の促進

**別表 1 : 抗アクアポリン 4 抗体陽性視神経炎の診断基準**

主要項目

- ⑦ 突然発症する片眼または両眼の重度の視力障害
- ⑧ 眼球運動痛, 眼痛, 眼窩痛, 頭痛
- ⑨ 中心暗点, 水平半盲, 耳側半盲, 同名半盲などの重度の視野障害
- ⑩ 急性期には頭部 MRI 冠状断 STIR 法および T2 強調像で罹患視神経に高信号
- ⑪ ステロイド治療に抵抗性

必須項目

- ⑫ 血清抗アクアポリン 4 抗体陽性

副次項目

- ⑤ 他の血清自己抗体が陽性である (抗核抗体, リウマチ因子, 甲状腺関連自己抗体 (抗 TSH 受容体抗体, 抗サイログロブリン抗体, 抗ペルオキシダーゼ抗体), 抗 SS-A 抗体,

抗 SS-B 抗体など)

- ⑥ 脊髄 MRI で 3 椎体以上の脊髄病変
- ⑦ 発症時に脳 MRI 病変が MS 基準を満たさない
- ⑧ 10 代から 70 代までの女性に幅広く分布してみられる

鑑別診断

- ⑰ 脱髄性視神経炎（多発性硬化症に伴う視神経炎）
- ⑱ 特発性視神経炎
- ⑲ 慢性再発性炎症性視神経症（chronic relapsing inflammatory optic neuropathy : CRION）
- ⑳ 圧迫性視神経症
- 21 Leber 遺伝性視神経症
- 22 後部虚血性視神経症
- 23 傍腫瘍性視神経症
- 24 中毒性視神経症

主要項目 5 項目のうち 3 項目と必須項目をみたしたものを抗アクアポリン 4（aquaporin 4 : AQP4）抗体陽性視神経炎とする。

重症度分類

軽症：視力低下が固定した時点で、視力低下が強い方の眼の視力が(0.1)以上。

重症：

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**別表 3：IVIG の免疫作用機序**（中尾 2012）より転載）

- 8. 病原的な自己抗体を不活性化する抗 idiotypic 抗体の自己免疫修正

9. 抗体 Fragment c (FC) 受容体のブロックによるリンパ球活性化の抑制と炎症反応抑制

- 10. B細胞の抑制による自己抗体産生の抑制
- 11. 遊離自己抗体の中和
- 12. 補体活性化や補体の組織・血管への沈着抑制
- 13. 炎症性サイトカインの産生抑制
- 14. 中枢神経の髄鞘再形成の促進

## IV. 関連業績一覧

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