Epidemiology and Current Treatment of Neuromyelitis Optica: A Systematic Review

INTRODUCTION

- Neuromyelitis Optica (NMO; also known as Devic’s disease) is a rare autoimmune condition characterised by:
  - Acute relapsing optic neuritis
  - Extensive transverse myelitis
- Historically, NMO was viewed as a subtype of Multiple Sclerosis
- Anti-Aquaporin 4 antibody (against aquaporin-4 antigen) is specific, and present in approximately 70% of people with NMO
- Many MS treatments (such as beta-interferon) may actually increase relapse rates in NMO
- The epidemiology of NMO is poorly described worldwide
- A curative treatment for NMO does not exist to date
- Most treatment recommendations are mainly based on case reports and retrospective case series

OBJECTIVES

- To determine the epidemiology of NMO
- To provide an algorithm of treatment for NMO

MATERIALS AND METHODS

- A systematic search was conducted of the relevant published evidence from Embase, MEDLINE, and Cochrane
- Search limits were articles in English and human
- Retrieved citations were screened by two independent reviewers according to inclusion criteria:
  - Population: NMO patients with any age
  - Interventions: Any interventions for treatment for NMO
  - Outcomes: Incidence and prevalence
- The analyses of comparable outcomes were carried out as per appropriate statistics along with critical appraisal of the studies

RESULTS

- Total records identified through database searching: N=849
- Records excluded after abstract screening: N=672
- Records after duplicates removed: N=713
- Articles assessed for full-text eligibility: N=41
- Full Text Articles excluded: N=25
- Articles included for final review: N=16
  - Studies about NMO epidemiology: N=6
  - Studies about NMO treatment algorithm: N=10

EPIDEMIOLOGY

- Incidence:
  - 0.05 per 1,00,000 (United Kingdom)
  - 0.4 per 1,00,000 (Southern Denmark)
- Prevalence:
  - 0.44 per 1,00,000 (United Kingdom)
  - 4.4 per 1,00,000 (Southern Denmark)
- Peak prevalence of NMO occurs among the people at 40-49 years of age

CURRENT TREATMENT

- Low level evidence recommended methylprednisolone 1g/day for 3 to 5 days or 2 to 3 sessions of plasmapheresis per week, up to 7 sessions for acute attacks of NMO
- Nine studies observed the improvements in the reduction of mean annualized relapse rate

DISCUSSION

- NMO is an unpredictable, often disabling disease of the central nervous system and resulting in permanent disability
- It is more prevalent in female than males
- The worldwide incidence and prevalence of NMO remains poorly characterized
- NMO represents less than 1.5% of individuals with demyelinating disorders
- The highest reported incidence is in Denmark: 4 new cases per 1,00,000 people per year
- There is currently no cure for NMO
- NMO is managed with a variety of medications:
  - Acute NMO attacks: High dose intravenous corticosteroid and plasmapheresis
  - Maintenance therapy: Low-dose oral corticosteroids and non-specific immunosuppressant drugs
- Most treatment recommendations are mainly based on case reports, case series, and a few prospective studies, all of which only meet evidence class III-IV
- Several areas of uncertainty still persist:
  - Whether treatments of seronegative NMO and seropositive NMO are similar?
  - What is the appropriate treatment for atypical forms of APQ4-Ab-positive NMO?
  - What is the relative efficacy of different treatment strategies for different forms of NMO?

CONCLUSIONS

- There is limited evidence on current available treatment therapies for NMO
- The available low level evidence found that high dose intravenous corticosteroid pulse and plasmapheresis may help in acute attacks of NMO
- Further well designed, adequately powered studies are required in this context

REFERENCE


Poster presented at ISPOR 18th Annual European Congress, 7-11 November, 2015, MiCo-Milano Congressi, Milan, Italy