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Epidemiology and Current Treatment of Neuromyelitis Optica: A Systematic Review

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INTRODUCTION

- Neuromyelitis Optica (NMO; also known as Devic's disease) is a rare autoimmune condition characterised by:
  - Acute relapsing optic neuritis
  - Extensive transverse myelitis [1]
- Historically, NMO was viewed as a subtype of Multiple Sclerosis[2]
- Anti-Aquaporin 4 antibody (against aquaporin-4 antigen) is specific, and present in approximately 70% of people with NMO [3]
- Many MS treatments (such as beta-interferon) may actually increase relapse rates in NMO [4]
- The epidemiology of NMO is poorly described worldwide [5]
- A curative treatment for NMO does not exist to date[6]
- Because NMO is rare and frequently severe, adequate prospective, randomized controlled trials are not available to evaluate treatment efficacy [6]
- Most treatment recommendations are mainly based on case reports and retrospective case series [6]

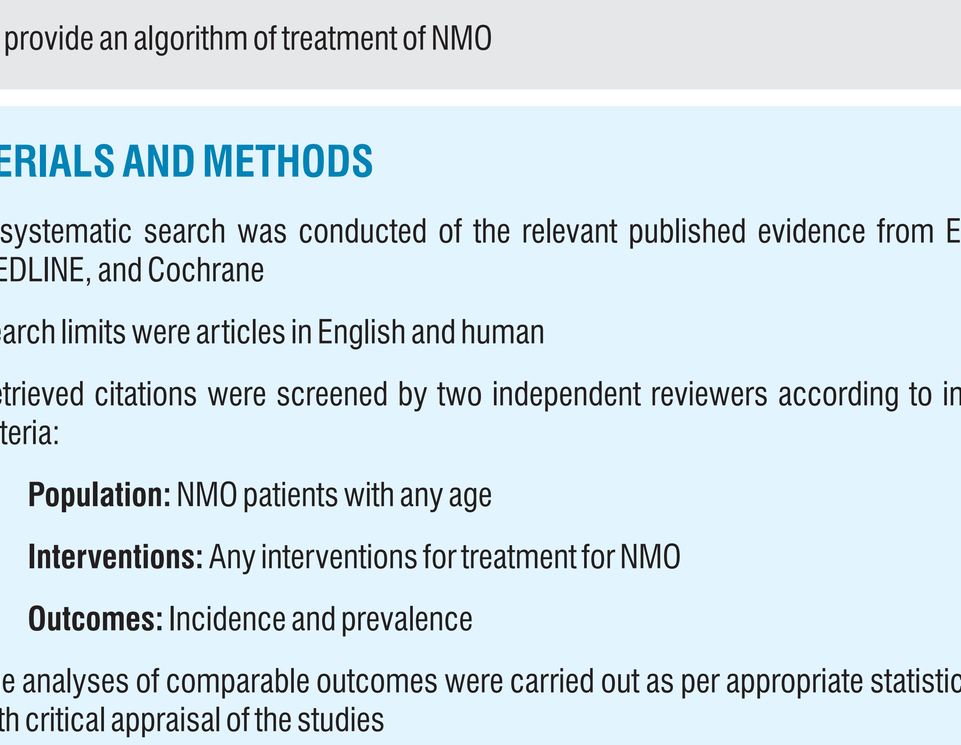
OBJECTIVES

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

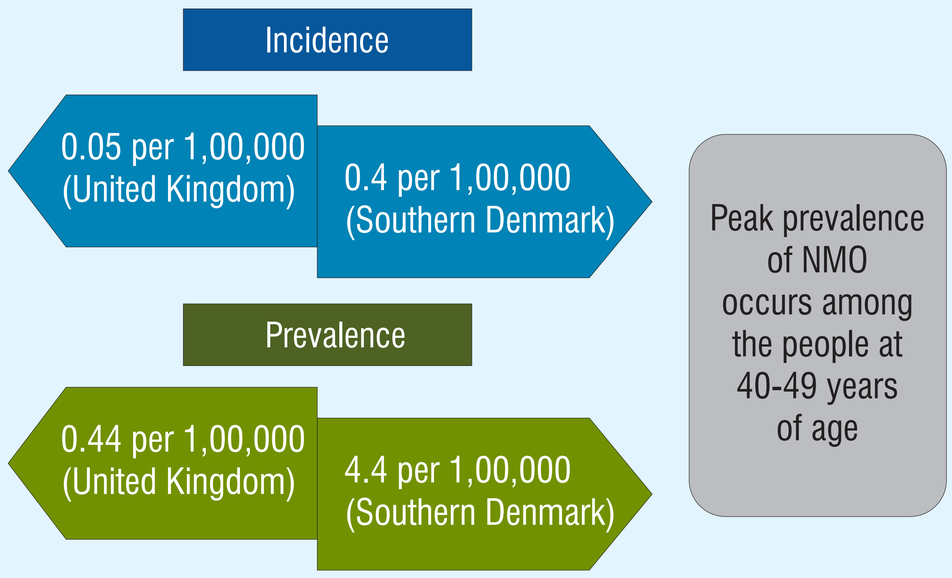
MATERIALS AND METHODS

- 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



EPIDEMIOLOGY



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[5]
- The worldwide incidence and prevalence of NMO remains poorly characterized [7]
- NMO represents less than 1.5% of individuals with demyelinating disorders [5]
- The highest reported incidence is in Denmark: 4 new cases per 1,000,000 people per year [8]
- There is currently no cure for NMO[6]
- NMO is managed with a variety of medications:[6]
  - 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[6]
- 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

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