**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 (MS).

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.

There is currently no cure for NMO.

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 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**

- Total records identified through database searching: N=849
- Records after duplicates removed: N=713
- Articles assessed for full-text eligibility: N=61
- Articles included for final review: N=16
  - Studies about NMO epidemiology: N=6
  - Studies about NMO treatment algorithm: N=10

**EPIDEMIOLOGY**

<table>
<thead>
<tr>
<th>Incidence</th>
<th>Prevalence</th>
<th>NMO Prevalence</th>
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<tbody>
<tr>
<td>0.05 per 1,00,000 (United Kingdom)</td>
<td>0.4 per 1,00,000 (Southern Denmark)</td>
<td>Peak prevalence of NMO occurs among the people at 40–49 years of age</td>
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<tr>
<td>0.44 per 1,00,000 (United Kingdom)</td>
<td>4.4 per 1,00,000 (Southern Denmark)</td>
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**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 females 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 in Denmark: 4 new cases per 1,000,000 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 immunosuppressive 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?
  - Whether treatments of seronegative NMO and seropositive NMO are similar?
- 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**