



# Quality of Life in 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<sup>1</sup>
- Historically, NMO was viewed as a subtype of Multiple Sclerosis<sup>2</sup>
- Anti-Aquaporin 4 antibody (against aquaporin-4 antigen) is specific, and present in approximately 70% of people with NMO<sup>3</sup>
- Many multiple sclerosis (MS) treatments (such as beta-interferon) may actually increase relapse rates in NMO<sup>4</sup>

### NMO is associated with significant reduction in quality of life (QoL)

- NMO is a relapsing condition: every relapse causes further disability, requiring a period of rehabilitation
- Optic neuritis is associated with blindness, loss of colour vision, central scotoma and pain on eye movement<sup>1</sup>
- Longitudinal extensive transverse myelitis is associated with bilateral motor weakness, sensory loss including numbness, intense paraesthesia, paroxysmal tonic spasms, neuropathic pain, itching and bladder and bowel dysfunction<sup>2</sup>
- Brain stem involvement can cause prolonged hiccoughs, nausea, vomiting, vertigo and respiratory failure<sup>2</sup>
- MS is also associated with transverse myelitis
- Transverse myelitis from MS is typically not as fulminant compared with NMO:
  - Less motor involvement
  - Greater chance for recovery<sup>5</sup>
- At autopsy, transverse myelitis from MS is:
  - Partial
  - Associated with fewer spinal levels
  - Demonstrates relative axonal preservation<sup>6</sup>
- Even though pathophysiology of NMO and MS are similar, there may be differences in the QoL of patients with these diseases

## OBJECTIVE

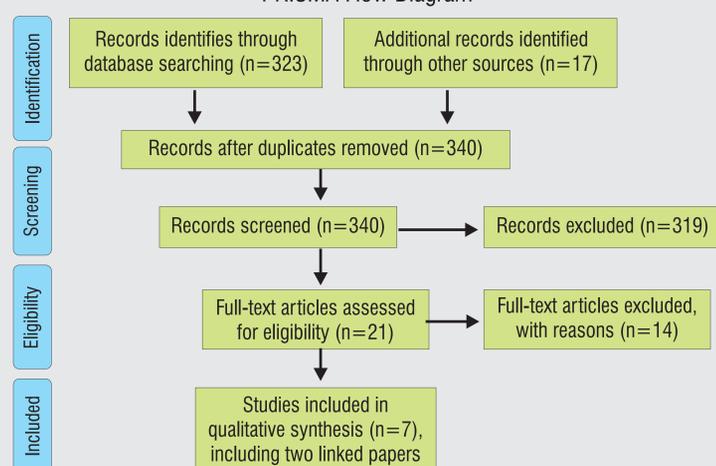
To evaluate the QoL in patients with NMO by conducting a systematic review of published peer-reviewed studies

## METHODS

- Literature Search: performed in "MEDLINE" and "EMBASE"
- Studies that included patients with NMO and reported use of validated QoL instrument were included
- Key Words:
  - Quality of life
  - Neuromyelitis optica
  - Devic's
- Population: Patients with Neuromyelitis Optica
- Outcome: Pain level and QoL
- Language of studies: English only
- Time of publication: All studies published before November 2014
- Study design: Observational studies
- Two independent authors screened the titles and abstracts and extract the data in standardized format of the studies included. All disagreements were resolved through discussion

## RESULTS

PRISMA Flow Diagram



- Number of studies included for analysis: 7, out of which two were linked
- Country from where studies were reported: USA (1), UK (1), France (1), Japan (1), Argentina (1)
- Total number of patients: (174)
- QoL instruments used:
  - Short form-36
  - Various pain severity scores (Short Form of the Brief Pain Inventory, McGill Pain Questionnaire)
  - Fatigue (EMIF-SEP)
  - Depression (EHD)
  - Extended Disability Status Scale
- Comparator:
  - Patients with MS (three studies) patients
  - Normal subjects (two studies)

Study (year)	Country	No of patients	Comparator	QoL instrument	Findings
Kanamori 2011 (1 linked abstract)	Japan	37	MS patients	• HRQoL: SF-36 • Pain: SF-BPI	• Pain was far more common in NMO (83.8%) than in MS (47.1%). • Pain Severity Index score (BPI) was significantly higher in NMO. • SF-36 scores in NMO were lower than MS, especially in bodily pain.
Qian 2012	USA	29	MS patients	• HRQoL: SF-36 • Pain: McGill Pain Questionnaire	• Pain was more common in subjects with NMO (86.2% vs 40.9%; P < 0.001) • Pain was more severe in NMO (5.38 vs 1.85 on a 10-point scale; P < 0.001). • Pain remained more common after controlling for disability and number of spinal cord segments (P=0.03)
Zhao 2014 (1 linked abstract)	UK	50	None	• Douleur Neuropathique 4 • BPI • Extended Disability Status Scale • SF36	• Neuropathic pain was identified in 62% patients. • Pain was constant in 68% affecting most Activities of Daily Living • Pain was associated with significant reduction of the SF36 Mental Composite Score.
Chanson 2012	France	40	Normal subjects and MS groups	• HRQoL: SEP-59 • Fatigue (EMIF-SEP) • Depression (EHD)	• HRQoL scores were lower in NMO patients (p < 0.01) when compared to normal subjects • HRQoL in term of Cognitive function, Sphincter dysfunction, and Psychological dimension of fatigue was better in NMO when compared to MS patients
Vanetti 2012	Argentina	18	Controls	• SF36 • Brief Repeatable Neuro-psychological Battery (BRNB) • Beck Depression Inventory II (BDI II).	• QoL was significantly lower in NMO patients with respect to physical functioning (p=0.007), pain (p=0.038) and general health (p=0.005). • NMO patients when had significant impairment in various domains of the BRNB • Depression was found in NMO patients: mild: 13.9%; moderate: 5.6%; severe: 11.1% • SF-36: NMO patients showed significant decline in memory and energy/fatigue (p=0.012), visual memory and emotional well-being (p=0.027) • BDI II: NMO patients showed decline in energy/fatigue (p=0.047) and social function (p=0.043).

- Most studies reported that QoL in NMO is:
  - Lower than QoL in MS patients
  - Much lower than QoL in normal subjects
- The lower QoL score in NMO patients corresponded with higher pain scores

## DISCUSSION

Summary of major differences between NMO and MS<sup>7</sup>

	NMO	MS
Distribution of symptoms and signs	Usually restricted to optic nerves and spinal cord	Any white -matter track
Attack severity	Usually severe	Usually mild
Head MRI	Usually normal/ non specific changes	Multiple periventricular white matter lesions
Spinal cord MRI	Longitudinally extensive central necrotic lesions	Multiple small peripheral lesions
CSF cells	Pleocytosis during attacks	Rarely > 25 white cells
Oligoclonal bands	Usually absent	Usually present
Permanent disability	Usually attack -related	Usually in late progressive phase
Coexisting autoimmunity	Frequent (31 -40%)	Less common
Serum anti -NMO antibody	Present	Absent

### In a study reported in 2012:<sup>6</sup>

- Pain affects patients with NMO more frequently and severely
  - Frequency of current pain: NMO vs MS: 86.2% vs 40.9% (p < 0.001)
  - Severity (10-point scale): NMO vs MS: 5.38 vs 1.85 (p < 0.001)
- This resulted in higher amount of prescription medication requirement (NMO vs MS: 75.9% vs 37.8%; p < 0.001)

## CONCLUSIONS

- NMO patients are associated with higher levels of pain and lower QoL scores than MS patients
- Further research is required

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