The frequency of Devic's NMO syndrome in a cohort of NMOSD patients

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Introduction:

The term neuromyelitis optica (NMO) was introduced by Eugène Devic and Fernand Gault in 1894, who described the association of bilateral optic neuritis (ON) and myelitis as a new clinical entity. The original concept of NMO has been dramatically changed since the last year of the 20th century.

Objective:

To evaluate the frequency of Devic's NMO, according to its original description in a reference center for demyelinating diseases of the central nervous system.

Methods:

We reviewed the medical records of a group of patients referred to our Center with a possible diagnosis of neuromyelitis spectrum disorder (NMOSD). Patients who met the 2015 Wingerchuk diagnostic criteria and had optic neuritis were selected. Criteria for Devic's original diagnosis of NMO included (1) acute bilateral optic neuritis simultaneously associated with acute myelitis; (2) absence of evidences of brain involvement; and (3) monophasic course.

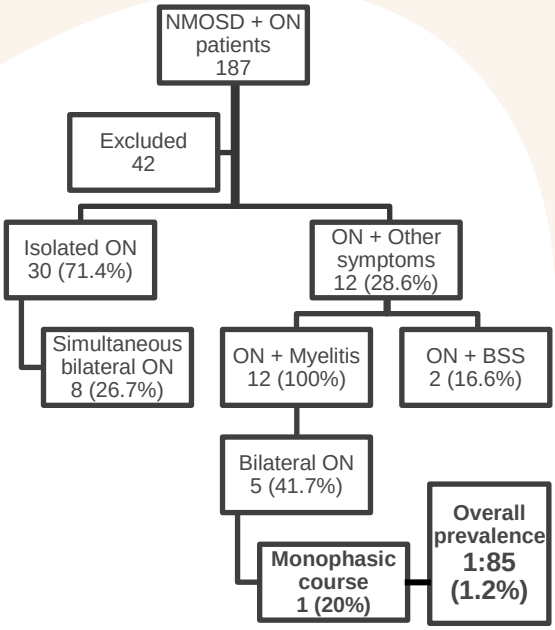

Figure. Flowchart demosntrating the criteria for Devic's Syndrome and the study patient that meets the criteria (highlighted in bold)

Results:

Out of 187 patients, 85 were included and 42 (49.4%) of them presented ON at onset. Optic neuritis occurred as an isolated symptom in 30 (71.4%) patients. Simultaneous bilateral ON occurred in 8 (26.7%) patients. Twelve (28.6%) patients presented ON associated with other symptoms (myelitis in 12 (100%); and brainstem symptom in 2 (16.6%). In 5 (41.7%) of the 12 patients with ON and myelitis, ON was bilateral. One (20%) out of these 5 patients had a monophasic course. Only a single (1/85; 1.2%) patient had Devic's NMO.

Conclusions:

Optic neuritis and myelitis are frequent manifestations of NMOSD either at its onset or during its course. However, the association of simultaneous bilateral ON and myelitis at disease onset is very infrequent. The additional requirement of a monophasic course turns Devic's NMO syndrome a remarkable rare disease. Therefore, Devic's NMO syndrome is an extremely rare condition in the phenotypical variability of NMOSD.

References:

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