# Cranial Neuropathy in CIDP

*January 1, 2021*

*By **Michael Rubin**, MD*

Professor of Clinical Neurology, Weill Cornell Medical College

*Dr. Rubin reports he is a consultant for Merck Sharp & Dohme Corp.*

**SYNOPSIS:** Cranial nerves are infrequently involved in typical chronic inflammatory demyelinating polyneuropathy, but involvement is more common in multifocal acquired demyelinating sensory and motor neuropathy (about 50%). The facial nerve is the most commonly affected cranial nerve, often bilateral.

**SOURCE:** Shibuya K, Tsuneyama A, Misawa S, et al. Cranial nerve involvement in typical and atypical chronic inflammatory demyelinating polyneuropathies. *Eur J Neurol* 2020;27:2658-2661.

Typically, chronic inflammatory demyelinating polyneuropathy (CIDP) presents over the course of several months, with gradually progressive symptoms of a symmetric sensorimotor polyneuropathy, with proximal and distal motor involvement exceeding sensory impairment.

Several studies have reported tremor as a common symptom, with cranial nerve and bulbar involvement in 10% to 20% of patients, but the prevalence, characteristics, prognosis, and relations of cranial nerve palsy with CIDP subtypes have yet to be defined. A retrospective data review of 132 consecutive patients with CIDP, seen between 1984 and 2019 at Chiba University Hospital, Chiba, Japan, was undertaken to address these questions.

All patients had been followed for at least one year and fulfilled the European Federation of Neurological Societies and Peripheral Nerve Society criteria for definite or probable CIDP. Patients with multifocal motor neuropathy, and those positive for antimyelin-associated glycoprotein (anti-MAG) or sulfated glucuronyl paragloboside antibodies were excluded.

CIDP patients were grouped into subtypes encompassing typical CIDP (n = 89), multifocal acquired demyelinating sensory and motor neuropathy (MADSAM) (n = 31), distal acquired demyelinating symmetric neuropathy (DADS) (n = 9), and other (n = 3). Cranial nerve palsy that developed during the progressive phase of CIDP was deemed related to CIDP, but spinal accessory nerve (cranial nerve XI) involvement was excluded from evaluation, since the muscles (trapezius and sternocleidomastoid) also are innervated by upper cervical spinal nerves. Disability was assessed using the Hughes functional grading scale. Statistical analysis encompassed Fisher's exact test and the unpaired t test, with $P < 0.05$ established as the level of statistical significance.

Cranial nerve palsy was found in only 11% of both typical CIDP and DADS but was present in 48% of MADSAM patients. None of the three patients with other forms of CIDP had cranial neuropathy. Among 89 patients with typical CIDP, seven had VII nerve palsy which was bilateral in all affected, six had bulbar palsy with bilateral involvement of cranial nerves IX and X, two had bilateral trigeminal neuropathy, and one had bilateral III nerve palsy.

Of 31 patients with MADSAM, five had bulbar palsy with involvement of cranial nerves IX and X, of which four were bilateral; four had facial palsy, which was bilateral in one; three each had a III or VI nerve palsy; two each had a V or VIII nerve palsy; and one had optic neuropathy. Only a single patient with DADS had cranial neuropathy expressed by bulbar palsy with bilateral involvement of cranial nerves IX and X.

Cranial neuropathy recovered well in typical CIDP, despite being bilateral, whereas in MADSAM, despite being unilateral, cranial neuropathy was relatively refractory. Facial and bulbar palsy are the most common cranial nerve involvement seen in demyelinating inflammatory neuropathy, regardless of subtype.

**COMMENTARY** Immune modulation, using intravenous immune globulin (IVIG), glucocorticoids, or plasma exchange, is the recommended initial therapy for CIDP, the choice between equals being influenced by cost, availability, venous access, comorbidities, and side effects.

Often, those treated with IVIG require maintenance therapy. Is a higher dose of IVIG, given less frequently, better than a lower dose delivered more frequently? Among 25 CIDP patients randomized in a placebo-controlled crossover trial, receiving 20 g to 80 g of IVIG in intervals ranging from 14 to 135 days, no benefit regarding efficacy or side effect profile was appreciated with one regimen over the other.[1]

## REFERENCE


1. Kuitwaard K, Brusse E, Jacobs BC, et al. Randomized trial of intravenous immunoglobulin maintenance treatment regimens in chronic inflammatory demyelinating polyradiculoneuropathy. *Eur J Neurol* 2020; Sep 2. doi:10.1111/ene.14501. [Online ahead of print].



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**Issue Date:** January 1, 2021