**Supplementary Table 1: Clinical and serological characterization of the MOG PNS cohort**

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| **Patient Details** (age of onset, ethnicity, gender, disease course) | **PNS syndrome** | **Chronology and clinical characterization of CNS and PNS episodes, and treatment of CNS events** | **MRI Spine characterization** | **Neurophysio-logical characterization** | **CSF analyses** | **Additional investigations** | **Response of PNS symptoms to immunotherapy (change in SFSS)\*** | **Results of PNS antibody characterization** | **Follow up duration(months)** |
| Patient 1  9 yo Caucasian F  Monophasic | AIDP | Concurrent CNS and PNS involvement  E1 LETM + AIDP | Markedly swollen T3-T10, L1/conus with gadolinium enhancement, swollen nerve roots and exiting nerves | Normal motor conduction and poorly elicited F waves in upper limbs consistent with proximal demyelination, absent CMAPS in lower limbs and distal denervation consistent with severe AIDP with axonal injury | 5 mononuclear cells, 1 PMN, protein 3.95 g/L, OCBs negative | AQP4 ab negative | IV MP and dexamethasone for six weeks, plus IVIG induction – improvement in upper limbs but not lower limb power – ongoing paraplegia thought to be secondary to spinal cord ischaemia/necrosis; SFSS 5 to 4 | Serum from onset not available for testing | 104 |
| Patient 2  31yo Caucasian F  Monophasic | Myeloradiculitis | Concurrent CNS and PNS involvement  E1 Three months post-partum LETM plus diffuse lumbar spinal nerve root involvement | Markedly swollen thoracolumbar cord (T8-conus) with diffusely enhancing lumbar spinal nerve roots; resolution of imaging changes 10 months post onset | Normal upper limb SSEPs, LL SSEPs consistent with a myelopathy below the cervical cord | 134 white cells (95% monocytes), protein 0.68 g/dL, negative for intrathecal OCBs | Negative for AQP4 ab, ACE, ANA, ENA, ANCA, Lyme, HTLV1/2, HIV, VDRL | Complete recovery with IV MP followed by an oral prednisone wean of nine months; SFSS 4 to 0 | Serum from onset not available for testing | 21 |
| Patient 3  34 yo Caucasian M  Monophasic | Myeloradiculitis | Concurrent CNS and PNS involvement  E1 - Bilateral optic disc swelling + acute meningoencephalitis + cauda equina nerve root enhancement | MRI brain at onset demonstrated leptomeningeal enhancement and cauda equina nerve root enhancement  The patient received treatment and had complete clinical resolution with no new symptoms. Progress MRI was performed two months post disease onset and demonstrated a new posterior C2 lesion and enhancing right intradural S1 nerve root, although the patient was clinically asymptomatic from this. | NCS revealed subtle changes consistent with proximal nerve/ nerve root involvement (lower limb SNAP lower limit of normal, F wave latencies upper limit of normal for height and abnormal, EMG of right tibialis anterior abnormal) | 178 mononuclear cells, protein 2g/L, negative for intrathecal OCBs | Mildly elevated ACE 49-79u/L, negative autoimmune & paraneoplastic panel, AQP4 and GQ1b abs. Whole body CT-PET negative for malignancy | Complete resolution within one month of onset after treatment with two doses of dexamethasone and IVIG x5; SFSS 3 to 0 | Negative | 12 |
| Patient 4  26 yo Middle Eastern F  Relapsing | Multifocal motor neuropathy | PNS involvement first  E1 right median nerve multifocal motor neuropathy (IVIg)  E2 R UON 30 months post onset (regular IVIg for one year with complete resolution) | Normal spinal imaging | NCS consistent with right median neuropathy with conduction block at the elbow, normal SSEPs | Not done | Negative GM1 and GQ1b abs | Complete recovery following E1 with induction and monthly IVIg for one year, with symptom recurrence after cessation. Symptom resolution following restarting IVIg for a further one year, with complete resolution after this; SFSS 2 to 0. | Negative | 69 |
| Patient 5  29 yo Caucasian F  Relapsing | Bilateral distal lower limb pain and paresthesia and NCS consistent with an axonal sensorimotor neuropathy | CNS involvement first  E1 BON  E2-5 over the subsequent thirty years at least three recurrent episodes of unilateral ON, as well as a brainstem relapse (ataxia/ hemisensory and motor loss)  Previous diagnosis thought to be MS and trialled on various immunomodulatory therapies over the years with last treatment dimethyl fumarate – ceased approximately 29 years post disease onset.  Starting 30 years post disease onset – ascending paresthesia in left foot progressing to right foot. She was re-evaluated at this stage by a new neurological service and due to atypical radiology for MS and her history of recurrent ON, she was investigated and diagnosed as positive for MOGAD. By 32 years post onset, she was experiencing pain and paresthesia to bilateral calves. Due to new symptoms of peripheral pain and a revised diagnosis of MOGAD, she was commenced on rituximab six monthly (32 years post disease onset). | Normal spinal imaging | NCS consistent with length dependent axonal sensorimotor neuropathy | Not available | AQP4 ab negative | Partial improvement in bilateral lower limb sensory symptoms following initiation of rituximab after first treatment, however symptoms have progressed and unresponsive following repeated rituximab treatments; SFSS 3 to 2 | NF155 antibody positive (end point dilution 1:6400), positive with total IgG, IgG1, IgG2, and IgG4 secondary antibodies | 384 |
| Patient 6  54 yo Caucasian F  Relapsing | Brachial neuritis | CNS involvement first  E1 BON (IV MP) E2 left brachial neuritis 23 months post onset | Normal spinal imaging | NCS consistent with left brachial neuritis | 1 mononuclear cell, protein 0.45 g/L, OCB present in both serum and CSF | Negative for AQP4 abs, ANA, ENA, dsDNA, ANCA, C3/C4, EPG/IEPG | No specific treatment for brachial neuritis with spontaneous recovery over a three month period; SFSS 3 to 0 | Negative | 57 |
| Patient 7  58 yo Caucasian M  Relapsing | Brachial neuritis | PNS involvement first  E1 left brachial neuritis two weeks after H1N1 vaccine (two weeks of oral prednisone)  E2 LETM 72 months post onset (IV MP) | Normal spinal imaging with E1, MRI spine with E2 consistent with LETM T6-T10 | NCS consistent with left brachial neuritis | 2 mononuclear cells, negative for intrathecal OCBs | IgM kappa paraprotein 4g/L, negative for AQP4 abs, ANA, ENA, ANCA | E1 improvement with two weeks of oral prednisone but patient had persisting residual shoulder weakness with wasting of the left deltoid, supraspinatus, infraspinatus and weakness of left forearm pronation and pincer grip. Three months post onset, the patient developed increasing pain – retreatment of IV MP and weaning prednisone resulted in significant improvement in pain. Has ongoing paresthesia and motor deficits following E1; SFSS 3 to 2. | 3 serial samples tested from E1 (n=1), and E2 (n=2 – one at E2, one three months after E2).  GM1 IgM antibody positive in first sample only (at dilution of 1:100) | 126 |
| Patient 8  30 yo Caucasian F  Relapsing | Bilateral distal upper and lower limb pain and paresthesia, facial pain and paresthesia | CNS involvement first  E1 R UON (IVMP and briefly on fingolimod before diagnosis revised to MOGAD)  E2 R UON 30 months post onset (IV MP)  E3 Sensory symptoms commencing 48 months post onset - pain and paresthesia bilateral soles of feet intermittent over months, persisting paresthesia left toe; persisting right facial paresthesia and pain not consistent with trigeminal neuralgia  E4 worsening bilateral distal lower limb paresthesia 61 months post onset (started on 50 mg od prednisone with improvement of symptoms by day three)  E5 right lower limb radicular pain/paresthesia 65 months post onset (resolved spontaneously over a few weeks)  E6 progressive bilateral (R>L) LL pain/paresthesia in feet 70 months post onset (symptom resolution within three days of commencing high dose oral prednisone, commenced on MMF)  E7 right facial pain and paresthesia 74 months post onset (spontaneously resolved)  E8 worsening pain and paresthesia bilateral hands 81 months post onset while on MMF (resolved within one week of recommencing oral prednisone)  Examination findings - reduced pinprick sensation bilateral hands/feet consistent with a glove and stocking pattern | Normal spinal imaging at onset and subsequent follow up. MRI 54 months post onset demonstrated asymptomatic T1/T2 lesion which subsequently resolved on follow up imaging. | Normal NCS and SSEPs | Not available | Negative for AQP4, abs ACE, EPG/IEPG, ANA, ENA, dsDNA, ANCA, cardiolipin antibodies | Not responsive to pregabalin.  Noted resolution of pain and paresthesia by day three of 50 mg od prednisone during E4, E6, E7;  SFSS 3 to 1 | 3 serial samples available from E3, E4, and E5  CASPR2 antibody positive in all samples to end point dilutions of 1:3200, 1:3200, and 1:12,800 respectively | 86 |
| Patient 9  13 year old Caucasian F  Relapsing | Bilateral distal upper and lower limb pain and paresthesia | CNS involvement first  E1 L UON (IV MP plus two weeks oral prednisone)  E2 L UON 48 months post onset (IV MP)  E3 bilateral intermittent hand and feet pain and paresthesia plus headaches 60 months post onset – persisted throughout remainder of clinical course  E4 R UON 70 months post onset (IV MP + commencement of MMF)  E5 R UON 95 months post onset while on MMF (IV MP + increased oral prednisone + RTX)  Normal examination apart from depressed ankle jerks bilaterally | Normal spinal imaging | Normal NCS | 5 mononuclear cells, normal protein, negative for intrathecal OCBs | Positive SS-B (with sicca symptoms of dry eyes and dry mouth), negative AQP4 abs, ANA, ANCA, dsDNA, cardiolipin antibodies, beta 2 glycoprotein, lupus anticoagulant, ACE, VDRL | No response to pregabalin or amitryptiline.  Distal upper and lower limb pain and paresthesia responded to rituximab commencement with E5 with return of symptoms six months post initial RTX treatment; SFSS 3 to 2. | Negative | 108 |
| Patient 10  26 yo Caucasian F  Relapsing | Bilateral distal upper and lower limb pain and paresthesia | CNS involvement first  E1 BON (IV MP and oral prednisone two week course)  E2 L UON two months post onset (oral prednisone two week course)  E3 L UON four months post onset (oral prednisone two month course)  E4 bilateral hand pain and paraesthesia eight months post onset following an episode of gastroenteritis (?Lhermitte’s) (resolved spontaneously over one month)  E5 L UON 41 months post onset  E6 L UON 46 months post onset  E7 R UON 49 months post onset  E8 BON 56 months post onset following respiratory tract infection (oral prednisone one month course)  E9 BON 70 months post onset (oral prednisone two week course)  Between 70 months to 107 months post onset – five discrete episodes (E10-14) of bilateral hand and feet pain and paresthesia - on two episodes following lower respiratory tract infection (E10 – E13 resolved spontaneously over one month each)  E14 treated with oral prednisone two month course with symptom resolution in first week. | Normal spinal imaging | Normal NCS and SSEPs | Normal cell count and biochemistry, negative for intrathecal OCBs | Negative for AQP4 abs, dsDNA, beta 2 glycoprotein, lupus anticoagulant, ANA, ENA, C3/C4, ACE | E4, E10-E13 each resolved spontaneously over one month each.  E14 treated with oral prednisone with symptom resolution in the first week; SFSS 3 to 1. | Negative  4 serial samples tested from E1, E7, E10, E14 | 128 |
| Patient 11  36 yo Caucasian F  Relapsing | Bilateral distal upper and lower limb pain and paresthesia | PNS involvement first  E1 pain and paresthesia bilateral hands and feet progressing over six months, L>R  E2 L UON six months post onset  E3 recurrent pain and paresthesia right hand eight months post onset  Anticipated to commence RTX at time of latest follow up | Normal spinal imaging | Not available | Not done | Negative for AQP4 abs | No immunotherapy trialled for PNS symptoms; SFSS 2. | Negative | 9 |
| Patient 12  45 yo Caucasian F  Relapsing | Bilateral lower limb radicular pain and paresthsia | CNS involvement first  E1 BON plus anterior and intermediate uveitis (oral prednisone one month wean and topical prednisone eye drops)  E2 BON and bilateral uveitis 12 months post onset (MMF)  E3 R UON and uveitis 34 months post onset  Since E2 has been having recurrent bilateral lower limb radicular pain and paresthesia | Normal spinal imaging | Normal SSEPs, NCS not available | Intrathecal OCBs present | Negative for ANA, RF | Symptoms present despite being on MMF, not trialled on further immunotherapy; SFSS 2. | Negative | 68 |
| Patient 13  51 yo Caucasian F  Relapsing | Left lower limb radicular pain and paresthesia | CNS involvement first  E1 BON  E2 Left lower limb shooting pain and paresthesia 24 months post symptom onset, with each episode lasting up to one week – recurrent symptoms occurring over one year. Responded on two occasions to oral prednisone. | Normal spinal imaging | Not done | Not done | Negative dsDNA, ANA, ENA, ANCA, lupus anticoagulant, beta 2 glycoprotein, C3/C4, cardiolipin, ACE; mildly elevated anticardiolipin IgM 6 MPL (normal range <5) and IgG 16 GPL (normal range <5) | Symptoms resolved within 24 hours of oral prednisone use on two occasions. Otherwise spontaneously resolved over weeks. SFSS 2 to 1. | Negative | 84 |
| Patient 14  68 yo Vietnamese F  Relapsing | Left upper and lower limb radicular pain and paresthesia | CNS involvement first  E1 L UON following hepatitis B immunization (IV MP followed by 11 month wean of oral prednisone) E2 two week episode of left leg radicular pain and paresthesia seven months post onset  E3 intermittent left hand and leg radicular pain and paresthesia 11 months post onset E4 four weeks of left leg radicular pain and paresthesia 20 months post onset (spontaneously resolved) | Normal spinal imaging | Not done | Not done | ANA 1:650, negative AQP4 abs, HLA-B27, RF, anti CCP | No immunotherapy trialled for PNS symptoms; SFSS 1. | Serum from onset not available for testing | 35 |
| Patient 15  19 yo Caucasian M  Relapsing | Bilateral lower limb radicular pain plus bilateral distal lower limb pain and paresthesia | CNS involvement first  E1 R UON (IV MP with near complete recovery)  E2 R UON 12 months post onset (some recovery with IV MP)  E3 R UON 48 months post onset (IV MP but minimal recovery after this episode, started Avonex  E4 L UON + paresthesia and numbness of both thighs 54 months post onset (IV MP plus oral prednisone two year wean + RTX + commenced on MMF)  E5 right > left leg intermittent paroxysmal shooting pain in addition to persisting right thigh paresthesia 58 months post onset (added on induction PLEX and monthly PLEX)  E6 L UON 78 months post onset (treated with IVMP and restarting oral prednisone, restarting six monthly RTX, continuing MMF)  E7 L UON 100 months post onset (IVMP plus ongoing RTX)  E8 105 months post onset started developing bilateral foot pain and paresthesia gradually extending up to mid-calf bilaterally over two year period  Examination findings - reduced pinprick sensation bilateral hands/feet consistent with a glove and stocking pattern | Normal spinal imaging | Normal upper limb SSEPs. Lower limb SSEPs consistent with a lesion involving large fibre sensory pathway from left tibial nerve central to the lumbar root entry zone | 6 mononuclear cells, 6 red cells, protein 0.31 g/L, intrathecal OCBs present | Negative AQP4 abs, ANA, ENA, ANCA, ACE, VDRL, HIV, LHON genetic testing | Minimal response to gabapentin/ pregabalin  No significant response to any immunotherapy trialled including prednisone, MMF, RTX, PLEX; SFSS 3. | Negative | 134 |
| Patient 16  36 yo Caucasian F  Relapsing | Left lower limb radicular pain plus bilateral distal lower limb pain and paresthesia | CNS involvement first  E1 BON four months post-partum (IV MP x 3/7)  E2 BON three weeks post onset (IV MP x 3/7)  E3 BON five weeks post onset (PLEX, oral prednisone over 2.5 years; MMF)  E4 Left lower limb radicular pain and paresthesia over two weeks 18 months post onset, spontaneously resolved over two months  Over subsequent seven years, has had intermittent tingling, paresthesia and heat sensations, itch, and pain over bilateral feet and lower legs progressing up to calves (trial of PLEX 96 months post onset resulted in symptom resolution for a three week period with recurrence of symptoms in the week prior to next monthly PLEX due) | Normal spinal imaging | Normal NCS | No cells, protein 0.16 g/L, negative for intrathecal OCBs | ANA 1:160 speckled, negative ENA, dsDNA, RF, ANCA, ACE | Not responsive to gabapentin or amitryptiline  Trial of PLEX 96 months post onset results in three weeks of symptom freedom with recurrence of PNS symptoms in the week prior to the next plasma exchange; SFSS 2 to 1. | Negative  2 serial samples available from E1 and after E4 | 100 |
| Patient 17  31 yo Caucasian F  Relapsing | Migratory sensory neuritis (Wartenburg’s pattern) | PNS involvement first  E1 left facial burning pain diagnosed as L occipital neuralgia  E2 left UL weakness and paresthesia 37 months post onset    E3 R ear + R LL burning pain and paresthesia 40 months post onset (on glatiramer acetate)  E4 R face, left upper and lower limb pain and paresthesia (associated with right sensory cortex lesion) 49 months post onset (changed to teriflunomide)  E5 burning pain and paresthesia both arms 53 months post onset  E6 burning pain and paresthesia both arms 57 months post onset which recovered spontaneously  E7 burning pain both arms and left side of face following a lower respiratory tract infection 60 months post onset  E8 burning pain both arms following a viral upper respiratory tract infection 63 months post onset  Teriflunomide ceased at this stage with plans for prednisone +/- RTX if symptoms progress | Normal spinal imaging.  MRI brain during E4 identified a large right sensory cortical lesion consistent with left hemiparesthesia, however repeated MRI brains did not reveal other white matter lesions which could account for other sensory episodes | Normal UL and LL SSEPs and NCS | 3 white cells, 5 red cells, protein 0.51 g/L, intrathecal OCBs present | Negative ANA, ENA | Mild symptom improvement with trials of amitryptiline, gabapentin, carbamazepine, local lignocaine injection/ nerve block  No immunotherapy trialled for PNS symptoms; SFSS 1. | 2 serial samples available from E2 and E7  GM1 IgM antibody positive in first sample only (at dilution of 1:200) | 75 |
| Patient 18  39 yo Caucasian M  Relapsing | Migratory sensory neuritis (Wartenburg’s pattern) | PNS involvement first  E1 Hyperesthesia and pain over right C5 dermatome (spontaneously resolved)  E2 right facial paresthesia two months post onset (IV MP and two weeks of oral prednisone)  E3 recurrent burning/ bruised feeling on left buttocks, either thigh, or calf bilaterally 12 months post onset - each episode lasting approximately one week (MMF)  E4 - right occipital hyperesthesia and pain 33 months post onset thought to be short TM (planned to start on oral prednisone at last follow up – lost to follow up) | Normal spinal imaging up until E4 (increased signal C1) | Normal NCS | Normal protein, OCB negative, negative for AQP4 antibodies | High serum ACE on two occasions – negative chest CT and whole body PET scan with no evidence of systemic sarcoidosis | No significant response to any immunotherapy trialled (including prednisone, MMF); SFSS 1. | Negative  2 serial samples tested from E1 and E4 | 35 |
| Patient 19  49 yo Caucasian F  Relapsing | Migratory sensory neuritis (Wartenburg’s pattern) | CNS involvement first  E1 R UON; a few weeks later also had abdominal allodynia and paresthesia lasting a few weeks (spontaneously resolved)  E2 L UON 12 months post onset  E3 L UON 24 months post onset (IV MP and one month prednisone wean)  E4 right leg/ left arm/ truncal pain and paresthesia with profound allodynia/ hypersensitivity 27 months post onset (one week of oral prednisone)  E5 R UON 37 months post onset (IV MP and one month oral prednisone)  E6 left arm paresthesia 48 months post onset (IV MP and one month oral prednisone taper)  Between E4 to E7 – recurrent episodes every few months lasting up to two weeks - of pain followed by paresthesia in discrete locations – upper limbs/ lower limbs.  E7 82 months post onset short TM C5 (oral prednisone 2 months) | Normal spinal imaging up until E7 (increased signal C5) | Normal NCS and SSEPs | 21 white cells in the presence of a bloody tap (7450 red cells), negative for intrathecal OCBs | Negative AQP4 abas, dsDNA, ANA, ENA, ACE, VDRL, Lyme, HIV | E4 treated with one week of oral prednisone with complete symptom resolution in 24 hours after starting prednisone.    Recurrent episodes between E2 and E7 frequently treated with short course of oral prednisone with symptom resolution. SFSS 2 to 1. | Negative  2 serial samples available from E4 and E7 | 180 |

\* The SFSS scores are documented to represent the changes from pre immunotherapy to post immunotherapy.

Abbreviations: AIDP acute inflammatory demyelinating polyneuropathy; BON bilateral optic neuritis; CASPR2 contactin-associated protein-like 2; CNS central nervous system; CSF cerebrospinal fluid; E episode; L left; GM1 – ganglioside epitope; H1N1 an influenza A virus subtype; LETM longitudinally extensive transverse myelitis; MOGAD myelin oligodendrocyte glycoprotein antibody-associated disorder; monos mononuclear cells; MRI magnetic resonance imaging; NCS nerve conduction studies; NF155 neurofascin 155; OCB oligoclonal bands; PMN polymorphonuclear cells; PNS peripheral nervous system; R right, IVIg intravenous immunoglobulin; SFSS sensory functional system score (a component of the Expanded Disability Status Scale); SSEP somatosensory evoked potentials; TM transverse myelitis; UON unilateral optic neuritis