

# GUIDE

## *Axonal GBS*

This series of guides is produced by the Guillain-Barré Syndrome Support Group. We are a registered charity that supports those affected by the Guillain-Barré syndrome (GBS) and related conditions in the United Kingdom and the Republic of Ireland. The related conditions include chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) and Miller Fisher syndrome (MFS).

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It had been known for some time that in severe cases of GBS, a ‘bystander’ effect of the demyelination of the nerve could be damage to the nerve core or axon. In 1986, Feasby et al [Brain 1986 Dec;109 (Pt 6):1115-26] reported autopsy studies on a patient with a clinical diagnosis of GBS and who had died that showed severe axonal degeneration in nerve roots and distal nerves without evidence of demyelination. It was suggested that this might represent a variant of GBS characterised by an acute axonal neuropathy.

In 1995, Griffin, Ho et al reported on their findings after investigating the yearly epidemic of GBS amongst children in northern China [Brain 1995 Jun;118 (Pt 3):577-95, 597-605]. Twelve autopsied cases were studied. Three of the twelve cases showed the same characteristics of classic demyelinating GBS (AIDP). Six cases showed predominantly axonal damage with only minimum demyelination. (Paradoxically, the other three cases showed only mild changes to the nerve roots and sciatic nerves.) Within the group of six that showed axonal damage, three showed damage to both motor and sensory nerves and three had damage almost exclusively to the motor nerves. The patterns were described as acute motor-sensory axonal neuropathy (AMSAN) and acute motor axonal neuropathy (AMAN).

Of 129 Chinese patients who were studied, 65% had the axonal form, 24% the demyelinating form and 11% could not be classified. One batch of 38 patients (55% axonal, 32% demyelinating, 13% unclassified) was tested for antibodies to the bacterium *Campylobacter jejuni*. Sixty-six

percent of the 38 showed evidence of recent *Campylobacter jejuni* infection compared with 16% in the control).

It did not take long for the axonal neuropathy as described by Feasby et al and the 'Chinese paralytic syndrome' to be regarded as one and the same and it was quickly recognised that *Campylobacter jejuni* was probably the most common trigger for GBS in the West as it seemed to be in China. [Hughes RA, Rees JH, *Infect Dis* 1997 Dec;176 Suppl 2:S92-8]

In 1997, Ho et al reported [*Neurology* 1997 Mar;48(3):717-24] on the mechanisms of paralysis and recovery during AMAN. The most severe cases showed degradation of motor axons affecting the ventral roots as well as the peripheral nerves. In contrast, a patient with the characteristic findings of AMAN recovered quickly after plasmapheresis. A sural nerve biopsy proved normal but a biopsy at a neuromuscular junction showed denervation (possibly explaining the Chinese paradox). Antibodies have also been found to be binding to the nodes of Ranvier (between the myelin segments) preventing transmission. There are clearly different mechanisms at work here: one resulting in a slow and incomplete recovery and another resulting in a rapid recovery. Note: Chinese AMAN patients had been found to recover at an identical rate as Chinese AIDP patients suggesting they fell into the latter category.

So while some patients with 'axonal GBS' may recover quickly, others have considerable axonal damage. They will be joined by those who have bystander axonal damage as a result of AIDP (and indeed CIDP). The only proven treatments are plasma exchange and IVIg (AIDP) plus corticosteroids (CIDP). A problem arises because while demyelination appears to be effectively and promptly repaired by remyelination, axonal degeneration can cause severe persistent disability. [Hughes et al, *Mult Scler* 1997 Apr;3(2):88-92].

For the possible consequences of Severe GBS, see our guide *After GBS – Severe GBS Supplement*.

If after reading this guide you still have anxieties and unanswered questions, telephone our helpline on 0800 374803 (UK) or 0033 1529 415278 (RoI). Alternatively, you can e-mail us or register for support on-line

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GBS Support Group, LCC Offices, Eastgate, Sleaford, Lincs, NG34 7EB

Tel: 01529 304615 E-mail: [admin@gbs.org.uk](mailto:admin@gbs.org.uk) Web site: [www.gbs.org.uk](http://www.gbs.org.uk)

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