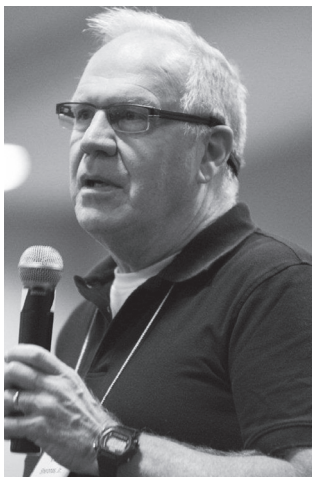

It was fantastic and eye opening and full of love! C.S. OK

I liked the variety of the sessions. J.R. CA

The experience cannot be duplicated. The knowledge I was able to acquire was vast. I hope to help more people due to this. All attendees were of utmost character. The hospitality of the Foundation was impeccable! I really had a wonderful time. T.F.Z. LA

Wonderful time, wonderful people, great education, unforgettable stories of pain, frustration, recovery and now somewhat normal living for most. The most incredible people that I have ever met. New friends forever. 2014 cannot come too soon. J.Y. NJ

Your staff and presenters, overall, did a great job. We thank you for putting this together. PA

Meeting folks with GBS/CIDP and knowing there is life after is spiritually uplifting. A.E. KS

Ask the Experts was a great way to wrap up Saturday session. H.B. NY

Chronic Inflammatory Demyelinating Polyneuropathy

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The primary function of the immune system is to differentiate between self and non-self, keep self healthy and destroy or neutralize non-self. When the immune system malfunctions and attacks self, it is known as an autoimmune disease.

Chronic inflammatory demyelinating polyneuropathy (CIDP,) is considered an autoimmune disease. The myelin sheath, which covers the nerves and assists with impulse transmission, is attacked. This is known as demyelination. Due to the nature of the immune attack, there is usually inflammation. The result is an interruption in nerve signals between the peripheral nerves and the muscles they control. CIDP presents slowly, usually over several months, unlike the acute form of demyelinating neuropathy, Guillian-Barré Syndrome (GBS.) GBS presents rapidly, usually over days, but sometimes even more quickly. Frequently GBS occurs following some sort of infection or illness. Unlike GBS, CIDP is usually a chronically progressive neuropathy and is rarely associated with antecedent illnesses or respiratory failure.[1]

CIDP usually presents as a motor predominant neuropathy with prominent proximal weakness, meaning the muscles responsible for movement closest to the torsoare affected first. The weakness is typically symmetrical, affecting both sides of the body equally. Occasionally CIDP can present in the pattern of a mononeuropathy multiplex, large-fiber neuropathy with sensory ataxia, pure motor neuropathy, or small-fiber neuropathy.[1]

CIDP may go undiagnosed for a while. This can be due to many factors. The symptoms may be vague and brushed off for a while until they become more profound and/or interfere with everyday functioning. Once someone does go to a physician, a definitive diagnosis still may not follow. Neuropathy has many causes, and CIDP has several variants.[2] Therefore, it is important that a thorough health history and physical and neurological examination be performed in order to determine the cause of the neuropathy. CIDP is rare, but the incidence ranges greatly due to the potential of over- or underdiagnosis, again partially a resultof the many causes of neuropathy. Someone may be thought to have CIDP when it is actually another form of neuropathy, and the reverse can happen as well. Many physicians and patient groups have worked on a standard way to identify CIDP more quickly and accurately. Appropriate diagnosis remains a challenge.

TABLE 1.

CIDP AND VARIANTS[2]
A. Symmetric proximal and distal motor predominant CIDP
B. Lewis-Sumner syndrome (LSS) (or multifocal acquired demyelinating sensory and motor neuropathy)
C. Demyelinating neuropathy with IgG or IgA paraprotein
D. Sensory predominant demyelinating neuropathy
E. CIDP neuropathy with central nervous system (CNS) demyelination
F. Demyelinating neuropathy associated with systemic disorders 1. Hepatitis B or C 2. HIV 3. Lymphoma 4. Diabetes mellitus 5. Systemic lupus erythematosus or other collagen vascular disorders 6. Thyrotoxicosis 7. Organ or bone marrow transplants 8. Nephrotic syndrome 9. Inflammatory bowel disease
G. CIDP in patients who have inherited neuropathy

[1]*Lewis, RA Chronic Inflammatory Demyelinating Polyneuropathy Neurol Clin 25 (2007) 71–87*

Symptoms are first noticed as numbness, tingling, pain and weakness, which are vague and can be the initial symptoms of many conditions. This usually occurs first in the toes and feet, eventually resulting in foot drop or drag and increased difficulty in walking. The weakness and numbness is typically symmetrical – equal on both sides of the body. Sensory loss is often in a stocking and glove distribution.

The diagnosis is based on an electrophysiologic pattern of multifocal demyelination identified through an EMG/ nerve conduction study, elevated CSF (cerebral spinal fluid) protein, and, when necessary, nerve biopsy. These

TABLE 2.

TEST	What is it?
EMG – Electromyography*	A procedure to measure and record muscle activity to show which muscles and nerves are affected.
NCS – Nerve Conduction Study*	A procedure to measure the speed and efficiency of electrical signals of the nerves.
Lumbar puncture	A spinal tap to look at the cerebral spinal fluid for abnormalities. Protein in the CSF is usually indicative of an immune response and can be present in CIDP.
Nerve biopsy	A section of the nerve is taken and examined to look for cause of damage. Only done if diagnosis is unclear.

**An EMG and NCS are almost always both done in order to appropriately diagnose CIDP.*

tests, combined with a thorough health history and neurological exam will help guide the physician to a correct diagnosis.

Once CIDP is diagnosed, treatment options should be considered and discussed. The treatment of CIDP is based on immunomodulating therapies which are summarized in Table 3. Immunomodulation refers to suppression or alteration of the immune response so attack on the self subsides and symptoms improve. CIDP does respond to corticosteroids, however, long term use of high-dose steroids comes with its own set of issues. Side effects can be severe and affect multiple organ systems. Plasmapheresis is generally reserved for refractory patients – those who have tried all the standard therapies and the condition is still not controlled.2 The only treatment that has received FDA approval for the management of CIDP is intravenous immunoglobulin (IVIG).

Summary

1. CIDP is an acquired, typically motor-predominant demyelinating sensorimotor neuropathy that is classified as an autoimmune disorder.
2. CIDP typically causes progressive, symmetrical weakness of the proximal and distal musculature, impairing walking and other activities of daily living.
3. CIDP can be treated with a variety of immunomodulatory therapies. The only treatment that has received FDA labeling for the treatment of CIDP is IVIG.

TABLE 3.

Standard Immunotherapy For Immune-mediated Neuropathies[2]

Therapy	Neuropathy Types	Route
Prednisone*	CIDP, VN	PO
Methylprednisolone*	CIDP, VN	IV
Azathioprine (Imuran)*	CIDP	PO
Cyclophosphamide (Cytoxan)*	CIDP, VN, MMN	PO
Cyclophosphamide	CIDP, VN, MMN	IV
Cyclosporine (Neoral, Sandimmune)*	CIDP	PO
IVIG*	GBS, CIDP, MMN	IV
Plasmapheresis	GBS, CIDP, MMN	IV
Rituximab (Rituxan)*	MMN, IgM - associated neuropathy	IV

Abbreviations: CIDP=chronic inflammatory demyelinating polyneuropathy; GBS=Guillain-Barré syndrome; IgM=immunoglobulin M; IVIG=intravenous immunoglobulin; MMN=multifocal motor neuropathy; VN=vasculitic neuropathy.

** Not FDA approved for this indication.*

References:

[1]*Wolfe, G. Do you need the reference for the chapter. It is as follows: Trivedi J.R. and Wolfe G.I. Peripheral neuropathies. In: Rakel R.E. and Bope E.T., eds. Conn's Current Therapy. Philadelphia: W.B. Saunders, 2005; pp 1086-1096.*

[2]*Lewis, RA Chronic Inflammatory Demyelinating Polyneuropathy Neurol Clin 25 (2007) 71–87*

GBS | CIDP
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CHANGE SERVICE REQUESTED

DIRECTORY

Check the enclosed chapter directory and contact the chapter nearest you. In addition, our "subgroups" are listed below.

- **"CIDP" Group**
  For those with a diagnosis of chronic inflammatory demyelinating poly-neuropathy. Please identify yourself to the National Office in order to be put in contact with others around the country.
- **Children with GBS**
  Call Lisa Butler, 215-628-2771
  670 Penllyn Blue Bell Pike
  Blue Bell, PA 19422
  Son, Stuart had GBS at 5 1/2 years old
- **Children with "CIDP"**
  For children diagnosed with chronic inflammatory demyelinating polyneuropathy. A separate registry has been created. Please contact the National Office for details.
- **Group for Having GBS Two Separate Times**
  Please call the National Office for contact with others.
- **Miller Fisher Variant Group**
  Please call the National Office for contact with others.
- **Wheelchair Limited Group**
  Please call the National Office for contact with others.
- **AMSAN Group**
  Please call the National Office for contact with others.
- **A Teenage Pen Pal Group**
  Arielle Challander, 231-946-7256
  413 Shawn Drive
  Traverse City, MI 49684
  E-mail: GBSTeenPenPal@hotmail.com
  Arielle had GBS in 2006 at age 13. She is willing to share experiences that others might not understand. To have a teenage GBS'er pen pal, write, call or e-mail to Arielle.
- **Pregnant Women with GBS**
  Robin Busch, 203-972-2744
  264 Oenoke Ridge,
  New Canaan, CT 06840
  Robin has offered to share her experience with GBS which came about during her pregnancy. We have many such cases and reassurance from someone who has gone through this is needed support.
- **Bereavement Group**
  A group for anyone who has lost a loved one due to GBS/complications. Please contact: Bereavement Group at the National Office.
- **The "Campy" Group**
  Those whose GBS onset was identified as a result of the campylobacter bacteria. Numbers to be used for research purposes.