Accordant
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Clinical Practice Guidelines

Chronic Inflammatory Demyelinating Polyradiculoneuropathy

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Approved by:
Andrew Krueger, MD
Mark B. Bromberg, MD, PhD
Donald Sanders, MD

Accordant Clinical Practice Guidelines:
Chronic Inflammatory Demyelinating Polyradiculoneuropathy (CIDP)

Table of Contents

INTRODUCTION

The AccordantCare™ program works with health plans to assess, monitor, and support those with certain complex, chronic conditions. The focus of the program is to improve health outcomes and prevent or limit disease-related complications. AccordantCare offers unique services at no additional charge to the patient, putting them in a strong position to adhere to their treatment plan.

There are several ways AccordantCare augments physicians' efforts. Through regular telephone contact, AccordantCare nurses:

- Keep patients informed about the disease process
- Coach patients in self-motivation and self-care skills
- Encourage patients to alert their physician when new symptoms arise
- Direct patients to resources that help pay for medication, transportation, home modifications, etc.
- Ensure preventive and screening measures are accomplished
- Provide emotional support to patients and caregivers
- Screen for depression
- Find local support groups

We invite physicians to make use of the services offered by AccordantCare and to suggest ways we can further patients' treatment goals. To offer feedback, get more information, ask questions, or voice concerns, call toll-free 1-800-948-2497 to speak with a program representative from 8 a.m. to 9 p.m., Monday through Thursday, and from 8 a.m. to 5 p.m. on Friday, Eastern Time. Messages left after hours will be returned the next business day.

Intent of Guidelines

The purpose of this Clinical Practice Guideline is to describe current patterns of practice where there is no fully established national guideline for diagnosis and management. It is not meant to dictate care of patients. Decisions about care are made by the physician and the patient based on the individual needs of that patient.

A patient's health plan may or may not pay for the all medicines, tests, equipment, or services mentioned in this document. Benefits should be checked with the individual's health plan to assure payment.

DISEASE OVERVIEW

CIDP, or chronic inflammatory demyelinating polyradiculoneuropathy, is an aquired demyelinating neuropathy that is likely autoimmune in nature. It is a rare nerve disorder marked by progressive weakness and impaired sensory function

in the legs and arms.[1] Also known as chronic inflammatory demyelinating polyneuropathy (the "radiculo" meaning "root" is often omitted), CIDP is considered to be a disease syndrome with several variant disorders that are based primarily on differences in clinical presentation. The exact causes of these disorders are still unknown.

While doctors and researchers are still considering how best to define CIDP and how best to classify the variants of the disease, most neurologists agree that typical or classical CIDP presents with symmetric motor weakness and sensory dysfunction of all the extremities.[2]

CIDP can be considered the chronic equivalent of acute inflammatory demyelinating polyneuropathy, the most common form of Guillain-Barré syndrome (GBS).[1] Note, that despite the similarities it is a different disorder.

Types of the Disease

No consensus exists on the best approach to the naming and classifying of these CIDP-related disorders.[3] The following classification scheme shows disorders that some doctors currently consider to be variants of CIDP, or in the same family as CIDP.[4]

- Typical CIDP[5]
  - symmetric, proximal, and distal muscles weakness
  - sensory loss
  - decreased or absent deep tendon reflexes
  - predominantly motor nerves affected, but distal sensory nerves affected also
- Variants (in approximate order of frequency or importance):
  - distal acquired inflammatory neuropathy
  - IgM (immunoglobulin M) paraprotein–related neuropathies with anti-MAG antibodies (myelin-associated glycoprotein)
  - demyelinating neuropathy with IgG (immunoglobulin G) or IgA (immunoglobulin A) paraprotein (monoclonal gammopathies)
  - sensory predominant demyelinating neuropathy (with dystaxia)
  - multifocal motor neuropathy with conduction block
  - Lewis-Sumner syndrome—multifocal, acquired, demyelinating sensory and motor neuropathy with focal sites of conduction block
  - POEMS (polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin changes syndrome)
  - Demyelinating neuropathy associated with systemic disorders, for example, with
    - hepatitis B or C
    - HIV
    - lymphoma
    - diabetes mellitus
    - systemic lupus erythematosus or other collagen vascular disorders

    - overactive thyroid gland and the resulting conditions
    - organ or bone marrow transplants
    - inflammatory bowel disease
  - CIDP neuropathy with central nervous system demyelination
  - CIDP in patients who have inherited neuropathy
  - CANOMAD (chronic ataxic neuropathy ophthalmoplegia M-protein agglutination disialosyl antibodies)

Prevalence/Incidence
CIDP has a prevalence estimated as ranging from 0.8 to 1.9 per 100,000 population.[4] The crude annual incidence of CIDP is about 0.15 per 100,000 population.[6]

DIAGNOSIS OF DISEASE

The most frequently used diagnostic criteria are those proposed by task forces of the American Academy of Neurology in 1991 and those by the European Federation of Neurological Societies/Peripheral Nerve Society (EFNS/PNS) in 2005 and revised in 2010.[7,8] Koski and coworkers developed another set of diagnostic criteria that relied on clinical evaluation alone when electrodiagnostic criteria are not fulfilled.[8,9]

For definite CIDP, one must have a typical or atypical clinical picture with clear demyelinating electrodiagnostic changes in two nerves, or probable demyelinating features in two nerves plus at least one supportive feature.

Electrodiagnostic testing is recommended for all patients with suspected CIDP:
- nerve conduction studies
- needle EMG studies

Supportive studies may be indicated in select patients:
- cerebrospinal fluid analysis
- nerve biopsy
- MRI of spinal roots, brachial plexus, and lumbosacral plexus
- evaluation for inherited neuropathies

Studies suggested to exclude other disorders:

- serum and urine paraprotein detection by immunofixation (consider repeating this study in patients who are or who become unresponsive to treatment)
- fasting blood glucose
- complete blood count
- renal function test
- liver function test
- thyroid function test
- antinuclear factor test
- to be performed if clinically indicated:
  - oral glucose tolerance test
  - HIV antibody test
  - Borrelia burgdorferi serology test
  - C reactive protein test
  - extractable nuclear antigen antibodies test
  - angiotensin-converting enzyme test
  - chest radiograph
  - skeletal survey (consider repeating this survey in patients who are or who become unresponsive to treatment)

APPROACH TO MANAGEMENT OF PRIMARY CONDITION

Treatment Goals

The main goals of treating CIDP are to

- control the patient's symptoms—for example, weakness, sensory loss, and pain[10];
- improve the patient's functional ability and quality of life[10]; and
- prevent or lessen disease worsening in people with progressive disease, or reduce the frequency of relapses in people with relapsing CIDP.[11]

To attain these treatment goals, it is necessary to modulate the autoimmune process, thereby stopping or slowing inflammation and preventing secondary axonal damage, the main cause of disability. To be successful, doctors must initiate therapy as early as possible in the disease course.[12]

Principal Treatments[8]

- corticosteroids (usually prednisone)
- high-dose IVIG (intravenous immunoglobulin)[13]
- plasma exchange (if both corticosteroids and IVIG are ineffective)

Immunosuppressants/Immunomodulators[8]

These agents are often used together with corticosteroids to reduce the need for IVIg or plasma exchange or when there is an inadequate response to any of the principal treatments. The Immunomodulating and immunosuppressant agents sometimes used to treat CIDP include

- alemtuzumab (Campath®);
- azathioprine (Imuran®);
- cyclophosphamide (Cytoxan®, Neosar®);
- cyclosporine A (Sandimmune®);
- etanercept, (Enbrel®);
- interferon alpha-2a (Roferon-A®) & pegylated interferon alpha-2a (Pegasys®);
- interferon beta-1a (Avonex®, Rebif®);
- methotrexate;
- mycophenolate mofetil (CellCept®);
- rituximab (Rituxan®);
- tacrolimus (Prograf®); and
- hematopoietic stem cell transplant.

Physiotherapy

Only very limited research examines the value of exercise and physiotherapy for patients with CIDP.[7,14] However, some doctors believe that physiotherapy and occupational therapy both play an important role in the assessment and management of the disease.[15] Planned exercise may help to maximize a patient's physical potential, particularly where weakness is the main problem.[15]

Other

Offer advice about foot care, exercise, diet, lifestyle management, and patient support groups to CIDP patients.[8]

PREVENTION AND MANAGEMENT OF COMPLICATIONS

Accordant helps patients prevent and manage complications by teaching early warning signs, encouraging adherence to treatment plans, offering supportive care, and recommending physician contact where needed. The list of goals and cooperative interventions below does not represent a comprehensive list of complications but reflects some of the more common clinical situations specific to CIDP. General health topics (e.g., age-appropriate cancer screening) are beyond the scope of this document.

Goal: Control the symptoms and prevent progression of childhood CIDP; improve overall functional ability and quality of life.
Cooperative interventions: Teach parents to:

- Recognize that common symptoms in children include gait disturbance and falling[16];
- Understand the importance of being careful to avoid falling caused by weakness or unsteadiness[17];

- Know that, in some cases, weakness, hand tremor, and an inability to coordinate voluntary muscle movements in the upper limbs may be present also[16];
- Understand that children tend to have a very favorable response rate to first-line therapy with corticosteroids, IVIG, or plasma exchange—with 80% to 100% of children responding to the initial treatment[18];
- Realize that although the unpredictability of CIDP can be very upsetting, the disease is not usually life-threatening, and the majority of people with CIDP can expect a good quality of life[18];
- Understand the advantages and disadvantages of each potential therapy for CIDP and discuss with their child's doctor the best individualized choice[7];
- Understand the importance of taking all medications as prescribed;
- Learn the medical and cosmetic complications of long-term steroid therapy (slowed growth, osteoporosis)[16];
- Understand the possible side effects of treatment (e.g., risk of thrombosis with IVIG[19]) and how to monitor and treat those side effects;
- Recognize opportunities to improve their child's mobility and quality of life by modifying the home, car, and other environments based on the child's individualized needs and capabilities (e.g., using a portable step to help climb into the car[3,7], setting hot water heater temperature low); and
- Understand the importance of communicating the child's condition to his or her teachers each year.

Goal: Minimize the patient's pain and allow for an optimal quality of life
Cooperative interventions: Teach patients to:

- Understand the symptoms of nerve pain[20]:
  - Numbness, tingling, and pricking sensations (paresthesia)
  - Sensitivity to touch
  - Painful cramps and uncontrolled muscle twitching
  - Burning pain
  - Loss of pain sensations
- Understand that nerve pain can be difficult to control and may be worse at night;
- Use mechanical aids such as hand or foot braces to help lessen pain[20];
- Realize the benefit of wearing orthopedic shoes (if prescribed) to improve gait disturbances[20];
- Avoid driving if feet are numb;
- Avoid climbing, lifting, or carrying heavy objects;
- Try applying moist heat or a cold compress for mild nerve pain[21];
- Educate about the pharmaceutical options for mild pain and their associated side effects/risks (e.g., capsaicin [Zostrix®], NSAIDs [Advil]);
- Educate about the pharmaceutical options for severe pain and their associated side effects/risks (e.g., antiepileptic medications[3], tricyclic antidepressants[3], pregabalin [Lyrica®], or duloxetine [Cymbalta®]); and
- Understand the importance of taking all medicines exactly as prescribed.

Goal: Avoid hospitalization, surgery, and disability associated with osteoporotic fractures

Cooperative interventions include teaching patients to[22]:

- Know the importance of risk assessment;
- Know the importance of BMD testing;
- Recognize the importance of calcium and vitamin D supplements;
- Discuss treatment options to prevent and/or repair bone loss with a physician, especially if taking corticosteroids;
- Learn fall prevention strategies (e.g., correct vision problems)[23];
- Improve home safety (e.g., install grab bars in bathroom, keep walkways well lit and free of clutter)[24];
- Participate in exercise that improves balance, agility, strength, and posture (e.g., Tai Chi[23]). Encourage members to get physician approval of new exercise programs;
- Understand the importance of smoking cessation; and
- Learn the importance of reducing alcohol consumption.

PATIENT EDUCATION

The Accordant Health Communities website at:
https://www.accordant.com offers resources for patients with CIDP.

The GBS/CIDP Foundation International website at:
www.gbs-cidp.org

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