

VISIT 2

Goal: Use a standardized instrument to confirm (or refute) the diagnosis of ME/CFS; then provide the patient with resources and an initial treatment program.

This section contains:

- 1994 CDC or International Criteria (Chart 2)
- 2003 Canadian Clinical Working Case Definition (Chart 3)
- Patient Handout
- Links of Interest to Persons with ME/CFS
- Initial Management of ME/CFS -- An Outline

First, use CHART 2 (based on the 1994 CDC or International Criteria*) or CHART 3 (based on the 2003 Canadian Clinical Working Case Definition **) below to formally confirm or refute the diagnosis of ME/CFS.

If the patient DOES NOT meet either one of these criteria, treat her/him appropriately and re-evaluate periodically.

If the patient's diagnosis is CONFOUNDED, treat the concurrent or confounding problems and re-evaluate periodically.

If the patient DOES meet either one of these criteria, provide her/him with the patient HANDOUT below and develop an initial treatment program.

For further information on the case definition criteria:

* CDC or International Criteria
Fukuda K, et al., *Annals of IM*, 121:953-959, 1994
<http://www.cdc.gov/cfs/cfsfullcasedefinition.htm>

** Canadian Clinical Case Criteria
Carruthers BM, et al., *J CFS*, 11(1):7-115, 2003
<http://www.cfids-cab.org/MESA/ccpc.html>

Chart 2 INTERNATIONAL / CDC CFS CASE DEFINITION*

NAME: _____ DATE: _____

Step 1. **Exclude** alternative diagnoses and other causes of chronic fatigue.

- Alternative diagnoses and other likely causes of chronic fatigue have been excluded by history, physical examination, exclusionary laboratory tests, and mental status examination.
- There is no evidence of melancholic depression, major psychiatric illness, current eating disorder or substance abuse.

Step 2. Does the patient have **chronic fatigue** that is both:

- A perception of diminished and finite energy that with usual or normal activity demands a substantial change in work or school and the usual lifestyle?
- New in onset (not lifelong) and has been persisting or relapsing for at least 6 months?

Step 3. Does the patient show **at least four classic symptoms**, which have been chronically or intermittently present for at least 6 months, make up a significant component of the illness, but have not predated the fatigue?

- The new onset of cognitive dysfunction characterized by short term memory loss; word searching or poor recall; diminished comprehension or oral or written information; new difficulty in processing, maintaining, or expressing thoughts; possibly difficulty with sequencing of events or numbers, or difficulty with simple math (making change, keeping up finances).
- Fatigue associated with post-exertional malaise (exhaustion or exacerbation of symptoms for a day or more following mental or physical exertion).
- Pharyngitis. A non-exudative, "scratchy" or sore throat, frequent or relapsing in nature.
- Lymphodynia or lymphatic soreness in at least two sites: anterior cervical, posterior cervical, axillary, or inguinal.
- Muscle discomfort of a generalized nature. Flu-like myalgias or tenderness to touch (allodynia or "touch me not").
- Joint discomfort (arthralgias) usually migratory and involving large joints more than small joints.
- Headaches of new onset or increased intensity, frequently retro-orbital or occipital and worsening with stress or exertion.
- Sleep is disturbed and/or non-restorative.

It is helpful to subcategorize your patient.

Onset of illness

- Abrupt onset over hours or days
- Gradual or insidious onset

Severity of symptoms (clinician's global impression)

- Minimal – some symptoms especially with effort. Usually able to work.
- Mild – mild symptoms and limitations, even at rest. May be able to work.
- Moderate – moderate symptoms at rest, worse with effort. Unable to work.
- Severe – often housebound or bedbound.

FINAL CLINICAL IMPRESSION

_____ Patient meets criteria for Chronic Fatigue Syndrome (Step 1 + Step 2 + \geq 4 symptoms)

_____ CFS is probable or possible but confounded by a concurrent medical or psychiatric condition, or lacks sufficient criteria

_____ Idiopathic chronic fatigue. CFS unlikely or excluded by _____.

* Based on the international case definition criteria (Fukuda, et al. *Annals of IM*, 1994) and "A case definition for practitioners" (Lapp C, *Annals of IM*, 1995)

Chart 3 CANADIAN CLINICAL CASE DEFINITION OF ME/CFS

CLINICAL WORKING CASE DEFINITION OF ME/CFS	
<p>In order to be diagnosed with ME/CFS a patient must meet criteria for fatigue and post-exertional malaise; and/or fatigue, sleep disruption, and pain; <i>plus</i> have two or more neurological/cognitive manifestations; <i>plus</i> one or more symptoms from two of the categories of autonomic, neuroendocrine, and immune manifestations; <i>plus</i> adhere to Item 7</p>	
	<p>1. Fatigue: the patient must have a significant degree of new onset, unexplained, persistent, or recurrent physical or mental fatigue that substantially reduces activity level.</p>
	<p>2. Post-exertional malaise and/or fatigue: there is an inappropriate loss of physical or mental stamina, rapid muscular and cognitive fatigability, post-exertional malaise and/or fatigue and/or pain and a tendency for other symptoms within the patient's cluster of symptoms to worsen. There is a pathologically slow recovery period – usually 24 hours or more.</p>
	<p>3. Sleep disruption: there is unrefreshed sleep or sleep quantity or rhythm disturbances such as reversed or chaotic diurnal sleep pattern</p>
	<p>4. Pain: there is a significant degree of myalgia. Pain can be experienced in muscles and/or joint, and is often widespread and migratory in nature. Often there are significant headaches of new type, pattern, or severity.</p>
	<p>5. Neurological / cognitive manifestations. <u>Two or more</u> of the following manifestations should be present: confusion, impairment of concentration and short term memory consolidation, disorientation, difficulty with information processing, categorizing and word retrieval, and perceptual and sensory disturbances, e.g. spatial instability and disorientation and inability to focus vision. Ataxia, muscle weakness, and fasciculations are common. There may be overload phenomena: cognitive, sensory -- e.g. photophobia, hypersensitivity to noise – and/or emotional overload, which may lead to “crash” periods or anxiety.</p>
	<p>6. At least <u>one</u> symptom from <u>two</u> of the following categories:</p> <ul style="list-style-type: none"> ❑ Autonomic manifestations: orthostatic intolerance -- Neurally Mediated Hypotension (NMH), Postural Orthostatic Tachycardia Syndrome (POTS), delayed postural hypotension; light-headedness; extreme pallor; nausea and irritable bowel syndrome; urinary frequency and bladder dysfunction; palpitations with or without cardiac arrhythmias; exertional dyspnea. ❑ Neuroendocrine Manifestations: loss of thermostatic stability – subnormal body temperature and abnormal diurnal fluctuation, sweating episodes, recurrent feelings of feverishness and cold extremities; intolerance of extremes of heat and cold; marked weight change – anorexia or abnormal appetite; loss of adaptability and worsening of symptoms with stress. ❑ Immune Manifestations: tender lymph nodes, recurrent sore throat, recurrent flu-like symptoms, general malaise, newer sensitivities to food, medications, and/or chemicals.
	<p>7. The illness persists for at least six months: usually it has a distinct onset, although it may be gradual. Preliminary diagnosis may be considered earlier. Three months is appropriate for children.</p>

	<p>To be included, the symptoms must have begun or have been significantly altered after the onset of this illness. It is unlikely that patient will suffer from all symptoms in criteria 5 & 6. The disturbances tend to form symptom clusters that may fluctuate and change over time. Children often have numerous prominent symptoms but their order of severity tends to vary from day to day. There are a small number of patients who have no pain or sleep dysfunction, but no other diagnosis fits but ME/CFS. A diagnosis of ME/CFS can be entertained when this group has an infectious disease type onset. Some patients have been unhealthy for other reasons prior to the onset of ME/CFS and lack detectable triggers at onset, or have more gradual or insidious onset.</p>
	<p>Exclude active disease processes that explain most of the major symptoms of fatigue, sleep disturbance, pain, and cognitive dysfunction.</p>

FINAL CLINICAL IMPRESSION

_____ Patient meets criteria for ME/CFS (Item 2 or Items 1,3,4 + Items 5,6 and 7)

_____ Patient does NOT meet criteria for ME/CFS

PATIENT HANDOUT

Chronic Fatigue Syndrome / Myalgic Encephalopathy (ME/CFS)

Chronic fatigue syndrome, or CFS, is a debilitating and complex disorder characterized by profound fatigue that is not improved by bed rest and that may be worsened by physical or mental activity. Persons with CFS most often function at a substantially lower level of activity than they were capable of before the onset of illness. In addition to these key defining characteristics, patients report various nonspecific symptoms, including weakness, muscle pain, impaired memory and/or mental concentration, insomnia, and post-exertional fatigue lasting more than 24 hours. In some cases, CFS can persist for years. The cause or causes of CFS have not been identified and no specific diagnostic tests are available. Moreover, since many illnesses have incapacitating fatigue as a symptom, care must be taken to exclude other known and often treatable conditions before a diagnosis of CFS is made.

Similar Medical Conditions

A number of illnesses have been described that have a similar spectrum of symptoms to CFS. These include fibromyalgia syndrome, myalgic encephalomyelitis, neurasthenia, multiple chemical sensitivities, and chronic mononucleosis. Although these illnesses may present with a primary symptom other than fatigue, chronic fatigue is commonly associated with all of them.

Other Conditions That May Cause Similar Symptoms

In addition, there are a large number of clinically defined, frequently treatable illnesses that can result in fatigue. Diagnosis of any of these conditions would exclude a definition of CFS unless the condition has been treated sufficiently and no longer explains the fatigue and other symptoms. These include hypothyroidism, sleep apnea and narcolepsy, major depressive disorders, chronic mononucleosis, bipolar affective disorders, schizophrenia, eating disorders, cancer, autoimmune disease, hormonal disorders, subacute infections, obesity, alcohol or substance abuse, and reactions to prescribed medications.

Other Commonly Observed Symptoms in CFS

In addition to the eight primary defining symptoms of CFS, a number of other symptoms have been reported by some CFS patients. The frequencies of occurrence of these symptoms vary from 20% to 50% among CFS patients. They include abdominal pain, alcohol intolerance, bloating, chest pain, chronic cough, diarrhea, dizziness, dry eyes or mouth, earaches, irregular heartbeat, jaw pain, morning stiffness, nausea, night sweats, psychological problems (depression, irritability, anxiety, panic attacks), shortness of breath, skin sensations, tingling sensations, and weight loss.

Prevalence of CFS

Chronic fatigue syndrome (CFS) affects more than one million people in the United States. There are tens of millions of people with similar fatiguing illnesses who do not fully meet the strict research definition of CFS.

Risk Factors for CFS

People of every age, gender, ethnicity and socioeconomic group can have CFS.

CFS affects women at four times the rate of men.

Research indicates that CFS is most common in people in their 40s and 50s.

Although CFS is much less common in children than in adults, children can develop the illness, particularly during the teen years.

Defining CFS Symptoms

CFS is marked by extreme fatigue that has lasted at least six months; is not the result of ongoing effort; is not substantially relieved by rest; and causes a substantial reduction in daily activities.

In addition to fatigue, CFS includes eight characteristic symptoms:

postexertional malaise (relapse of symptoms after physical or mental exertion);

unrefreshing sleep;

substantial impairment in memory/concentration;

muscle pain;

pain in multiple joints;
headaches of a new type, pattern or severity;
sore throat; and
tender neck or armpit lymph nodes.

Symptoms and their consequences can be severe. CFS can be as disabling as multiple sclerosis, lupus, rheumatoid arthritis, congestive heart failure and similar chronic conditions. Symptom severity varies from patient to patient and may vary over time for an individual patient.

Diagnosis of CFS

There are no physical signs that identify CFS.

There are no diagnostic laboratory tests for CFS.

People who suffer the symptoms of CFS must be carefully evaluated by a physician because many treatable medical and psychiatric conditions are hard to distinguish from CFS. Common conditions that should be ruled out through a careful medical history and appropriate testing include mononucleosis, Lyme disease, thyroid conditions, diabetes, multiple sclerosis, various cancers, depression and bipolar disorder.

Research conducted by the Centers for Disease Control and Prevention (CDC) indicates that less than 20% of CFS patients in this country have been diagnosed.

Treatment of CFS

Since there is no known cure for CFS, treatment is aimed at symptom relief and improved function. A combination of drug and nondrug therapies is usually recommended.

No single therapy exists that helps all CFS patients.

Lifestyle changes, including prevention of overexertion, reduced stress, dietary restrictions, gentle stretching and nutritional supplementation, are frequently recommended in addition to drug therapies used to treat sleep, pain and other specific symptoms.

Carefully supervised physical therapy may also be part of treatment for CFS. However, symptoms can be exacerbated by overly ambitious physical activity. A very moderate approach to exercise and activity management is recommended to avoid overactivity and to prevent deconditioning.

Although health care professionals may hesitate to give patients a diagnosis of CFS for various reasons, it's important to receive an appropriate and accurate diagnosis to guide treatment and further evaluation.

Delays in diagnosis and treatment are thought to be associated with poorer long-term outcomes. For example, CDC's research has shown that those who have CFS for two years or less were more likely to improve. It's not known if early intervention is responsible for this more favorable outcome; however, the longer a person is ill before diagnosis, the more complicated the course of the illness appears to be.

Recovery from CFS

CFS affects each individual differently. Some people with CFS remain homebound and others improve to the point that they can resume work and other activities, even though they continue to experience symptoms.

Recovery rates for CFS are unclear. Improvement rates varied from 8% to 63% in a 2005 review of published studies, with a median of 40% of patients improving during follow-up. However, full recovery from CFS may be rare, with an average of only 5% to 10% sustaining total remission.

Possible Causes of CFS

Despite an intensive, nearly 20-year search, the cause of CFS remains unknown. Many different infectious agents and physiologic and psychological causes have been considered, and the search continues.

Much of the ongoing research into a cause has centered on the roles of the immune, endocrine and nervous systems may play in CFS. More recently, interactions among these factors are under evaluation.

Genetic and environmental factors may play a role in developing and/or prolonging the illness, although more research is needed to confirm this. CDC is applying cutting-edge genomic and proteomic tools to understand the origins and pathogenesis of CFS.

CFS is not caused by depression, although the two illnesses often coexist, and many patients with CFS have no psychiatric disorder.

LINKS of Interest to Persons with ME/CFS

CFS Facts (CDC)

This site is an excellent source of general information on CFS as well as programs supported by the Center for Disease Control and Prevention. The CDC monitors and studies diseases that occur in the USA.

<http://www.cdc.gov/ncidod/diseases/cfs/index.htm>

CFS Facts (NLM)

This site is a compilation of articles on CFS, summaries, and interactive tutorials, provided by the National Library of Medicine in Bethesda, MD.

<http://www.nlm.nih.gov/medlineplus/chronicfatiguesyndrome.html>

FM Facts (NLM)

This site is a compilation of articles on FM, summaries, and interactive tutorials, provided by the National Library of Medicine in Bethesda, MD.

<http://www.nlm.nih.gov/medlineplus/fibromyalgia.html>

CFIDS Association of America

The CAA is America's largest and most reliable patient advocacy organization. The website provides current information, links, and a access to articles previously published by the organization.

<http://www.cfids.org>

American Fibromyalgia Syndrome Association

AFSA is a major provider of patient advocacy and research funds with regard to FM.

<http://www.afsafund.org>

FM Network

This site provides information and brochures on FM, as well as access to "Fibromyalgia Network" newsletter, which summarizes the latest FM information and research.

<http://www.fmnetnews.com/>

CFIDS / FM Self Help

Dr. Bruce Campbell was an educator with the Stanford chronic illness project when he was struck with ME/CFS in 1997. By following a Stepwise Approach, journaling, and slowly but steadily increasing his activity level, Campbell was able to recover from his illness within 5 years. He has now produced a book (with a forward written by Dr. Lapp) and supervises a 6 week internet-based self improvement course. For details go to www.cfidselfhelp.org.

Co-Cure

Co-cure is a patient-run information exchange, most notable for the "Good Doctors List" that provides names and addresses of local doctors who are willing to see persons with ME/CFS/FM.

<http://www.co-cure.org/>

Parents of Sick and Worn-Out Children

This privately maintained site is devoted to providing direction and resources for parents of children with ME/CFS/FM.

<http://home.bluecrab.org/~health/sickids.html>

The Pediatric Network

A website network for parents, youth, and professionals concerned with chronic fatigue syndrome, fibromyalgia, and orthostatic intolerance.

<http://www.pediatricnetwork.org/>

WebMD

This AMA-sponsored commercial site provides trustworthy, credible, and timely health information.

<http://webMD.com>

INITIAL MANAGEMENT OF ME/CFS FOR THE PRACTITIONER -- AN OUTLINE

Keep these goals in mind...

- Balance rest with low levels of activity
 - Recommend 2-3 periods of supine, quiet rest each day
- Stay active but not too active
 - Advise that energy is no longer boundless, but finite
 - Recommend a daily schedule or regimen
 - Avoid "pushing and crashing," that is exceeding limits of energy
- Practice good sleep hygiene, namely:
 - Use the bed for sleeping only
 - Avoid stimulant foods and beverages at night
 - Get up every morning at the same time
 - Try not to take daytime naps (although short rest periods are fine)
 - Wind down at night
 - Aim for 8-10 hours of sleep nightly
- Recommend an easy, low level, aerobic exercise program
 - Walking, stationary cycling, and swimming are best
 - Start with 3-5 minutes of exercise at a time, followed by 5 minute rest
 - Increase exercise time slowly, do as many repetitions as is comfortable
 - Try to exercise daily or every other day
 - Consider referral to a physical therapist with experience in ME/CFS, or a pool therapy program specifically for arthritis, FM, or ME/CFS

Recognize that there are no cures for ME/CFS, but like many chronic illnesses the symptoms can be managed effectively.

Persons with ME/CFS (or PWCs) are frequently sensitive to medications, especially sedatives. Therefore, start with low doses and advance slowly ("start low, go slow").

Treat the major symptoms first. These are sleep, pain, and fatigue:

Sleep.

Sleep initiation therapies might include:

- Melatonin, valerian
- Benadryl/ diphenhydramine, Tylenol PM
- Sonata / zaleplon or Lunesta / escopiclone
- Ambien / zolpidem
- Klonopin / clonazepam 0.5-1mg or other benzodiazepine

Sleep maintenance might include:

- Desyrel / trazadone in low doses of 25-50 mg nightly
- Doxepin, nortriptyline, or amitriptyline in low doses of 10-25 mg nightly

Obtain a sleep consultation if simple therapies are inadequate

Pain

Follow the WHO stepladder approach

Tramadol (as Ultracet or Ultram) has been shown to reduce both pain and tenderpoints when used regularly.

Obtain a pain consultation if simple analgesics are not satisfactory

Fatigue

SSRIs, NSRIs, and other antidepressants may reduce irritability, sleeplessness, mood disorders, and pain caused by a deficiency of neurotransmitters in ME/CFS

Excessive daytime sleepiness may respond to sleep management and/or stimulants such as modafanil / Provigil, amphetamine, or methylphenidate

On subsequent visits you may wish to address secondary problems such as dyscognition, **orthostatic intolerance** (if present), hormonal imbalances (estrogen, testosterone, thyroid), and allergies

Also, concurrent syndromes occur frequently in PWCs including irritable bowel syndrome, overactive bladder, TMJ syndrome, restless leg syndrome, chemical sensitivities, vasomotor (autonomic or non-allergic) rhinitis, gut motility disorder with dysphagia, early satiety, nausea, and/or constipation, sprue-like disorders with sensitivity to wheat, grains, or gluten, autonomic dysfunction with low blood pressure, orthostatic symptoms or syncope, sicca complex or a Sjogren's-like syndrome, vulvodynia or vulvar vestibulitis, joint hyperlaxity, milk protein intolerance, costochondritis, and metabolic syndrome.

Topics in **purple** are further explained in the GLOSSARY.