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## *THEORY USED IN CLINICAL PRACTICE*

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# Guidelines for the Diagnosis of Pediatric Chronic Fatigue Syndrome: Things Parents Need to Know

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**SUMMARY.** In this volume, chronic fatigue syndrome (CFS) in children and adolescents is specifically addressed. It is a topic long overdue. It is my sincere hope that the criteria presented here will begin a process of rigorous clinical testing and refinement so that pediatricians and other

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medical providers will come to have a reliable and accepted way of making the diagnosis of ME/CFS in a person under 18 years of age.

This short review is meant for parents and other caregivers as a brief summary of the guidelines that may be of value. The primary role of these guidelines is to present a strict and rigorous definition that can be tried and tested. This summary is to make the process of diagnosis somewhat easier for parents and caregivers alike until the testing process is completed. Therefore, for more detailed symptom description and exclusionary illness description, I would refer the reader to the primary article.

Professional caregivers and clinicians may make this article available to inform parents with a child/adolescent suffering from CFS. doi:10.1300/J092v13n02\_05 [Article copies available for a fee from The Haworth Document Delivery Service: 1-800-HAWORTH. E-mail address: <docdelivery@haworthpress.com> Website: <<http://www.HaworthPress.com>> © 2006 by The Haworth Press, Inc. All rights reserved.]

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## **BACKGROUND**

### ***Need for an Accurate and Specific Diagnosis of Chronic Fatigue Syndrome (ME/CFS)***

The disability of CFS in children and adolescents can be severe and last for years. Yet the illness is only rarely recognized and/or diagnosed by pediatricians, family care physicians or other medical care providers. Treatment recommendations for those diagnosed are usually uneducated guesses as there are almost no treatment trials in children and adolescents. The only way this situation can be corrected is to establish clear, accurate, and 'usable' diagnostic criteria for the illness in children and adolescents. The criteria presented in this journal are not meant to be a final answer to this problem, but a starting point that can be tested and improved.

Fortunately, the majority of children and adolescents with ME/CFS do relatively well and may resume normal or near-normal activity within a few years. However, even in the mildest of cases currently recognized, discomfort and disruption of educational and social life at a time of identity formation may have lifelong effects. Lack of diagnosis in any chronic condition increases anxiety and medical expenditures, and creates an undue hardship on patients and their families. Therefore

an accurate clinical diagnosis is essential for appropriate clinical management as well as symptom and educational management.

### ***Need to Address Medical Profession Skepticism***

In the United States, articles about CFS have been published since 1983, and in the United Kingdom and other countries since 1950. There have been descriptions of illnesses that probably represent clusters of ME/CFS going back to 1938 in the modern literature. However, every report seemed to generate its own name for the condition, and because of the difficulty inherent in describing symptoms, ME/CFS has not stood out as a specific clinical entity until recently. The confusion over names as well as the difficulty in “proving” the reality of the illness has led to widespread skepticism among the medical profession.

This skepticism has been particularly prominent when children and adolescents develop the illness. The central paradox of CFS is debilitating fatigue and exhaustion in a person who looks well. Pediatricians, despite their commitment to the well-being of children, quickly assume that fatigue and its impact upon daily activity is factitious, imaginary, exaggerated, hysterical or ‘benign.’ As a result, after routine exam and laboratory screen, the child or adolescent with debilitating and unexplained fatigue is frequently dismissed both without diagnosis and with medical provider disinterest, or at least is interpreted as such. The routine referral to a child psychiatrist without establishing the presence of depression or anxiety reinforces the perception of disinterest. This skepticism has spread to educators, and the pediatric ME/CFS patient and his/her family must fend for themselves arranging school accommodations and fighting allegations of child abuse and neglect for truency from school. The lifelong potential for harm in this scenario is enormous as it occurs during an important period of identity formation.

The fact that CFS is “real” is no longer at issue. The Centers for Disease Control have worked on this illness for years, and helped organize the effort to develop diagnostic criteria for adults. The National Institutes for Health have issued calls for grant applications on CFS. The science on CFS has made tremendous strides with over 1,000 papers published in the medical literature. Yet these advances have not translated into better care for the child or adolescent with ME/CFS. It is hoped that the development of pediatric CFS diagnostic criteria will begin to have an impact upon skeptical medical and educational providers.

### ***PROPOSED DIAGNOSTIC CRITERIA FOR CFS***

It is important to keep in mind is that CFS is a syndrome. We are defining it by a process that includes both defining a characteristic symptom pattern and the exclusion of other illnesses. The fact that we do not know the cause of this syndrome does not cast doubt on its existence. It is likely that there is more than one cause. We know, for instance, that infectious mononucleosis as a clinical illness can be caused by more than one infectious agent. But despite not knowing which virus may be the cause, the diagnosis of infectious mononucleosis, made on clinical grounds, is of great value to patients and their families.

The length of time for symptom presence has been suggested to be three months. In the process of testing these criteria we will attempt to establish the wisdom of this. If it appears that the adult criteria of six months is more accurate, changes can be made. The presence of anxiety and depression do not invalidate the diagnosis of ME/CFS. They may represent targets for treatment if present, just as sleep and pain represent targets for treatment. The presence of fibromyalgia tenderpoints or chemical sensitivities also do not invalidate the diagnosis.

#### ***Conditions Which Exclude the Diagnosis of ME/CFS***

As with all other criteria proposed for ME/CFS, the proposed pediatric criteria do not allow the presence of other medical or psychiatric conditions that can explain the activity limiting fatigue. From a medical perspective, this is relatively easy. If a child or adolescent has untreated hypothyroidism for example, the diagnosis of CFS/ME cannot be made. If, however, hypothyroidism was present but after appropriate treatment (usually defined by a normal thyroid stimulating hormone) the fatigue remains, then hypothyroidism was not the cause of the fatigue and the diagnosis of CFS/ME can be considered.

Similarly, children and adolescents with severe fatigue can be depressed. The mere presence of depression does not mean that it is causing the fatigue. If, in the clinician's view the depression is not causing activity limitation from severe fatigue, the diagnosis can again be considered. Factors such as the desire to go to school and participate in sports, the type of exhaustion experienced, the history of emotional symptoms before the illness began, all can help the clinician distinguish between the two. Conditions that do not permit CFS/ME to be diagnosed would include schizophrenia or psychotic disorders, bipolar

disorder, active drug or substance abuse (except nicotine) or active (untreated) eating disorders. This latter should not be confused with nausea and eating pattern changes that may be symptoms of the illness.

### ***Clinical Symptoms Necessary for Diagnosis of ME/CFS***

First and foremost, there must be clinically evaluated but unexplained fatigue that has been present for at least 3 months, is not the result of ongoing exertion, is not relieved by appropriate rest, and substantially reduces activity.

There is a symptom pattern necessary for the diagnosis of ME/CFS that needs to have been present for the past 3 months. Some symptoms may have been present prior to the reduction of activity or the severe fatigue. This pattern includes the following:

1. Post-exertional fatigue or malaise. This means that typically after exertion, and not necessarily strenuous, there is a worsening of exhaustion and/or other symptoms. This post-exertional worsening of the illness often takes more than 24 hours to resolve to the pre-exertion level.
2. Unrefreshing sleep, or disturbance of sleep quality or rhythm. This symptom can range from too much sleep (hypersomnolence) to light and broken sleep, and day/night (sleep phase) reversal, usually resulting in sleep that does not make the subject feel refreshed.
3. Widespread pain. This would include muscle and joint pain, widespread achiness, recurrent headache, eye pain, abdominal pain, chest pain.
4. Two or more Neurocognitive disturbances. The common complaints include impaired memory, difficulty in focusing, disturbed concentration, difficulty finding the right word, slowed thinking, need to focus on one thing at a time, and difficulty expressing thoughts. Neurocognitive difficulties could be present when academic difficulties are not explained by school absences. Neurocognitive problems could be present when the child/adolescent suddenly presents lower results than his pre-morbid achievements and this without any obvious reason.
5. At least one symptom from two of the following three categories:
  - a. Autonomic manifestations: Neurally mediated hypotension, postural orthostatic tachycardia, delayed postural hypotension,

palpitations with or without cardiac arrhythmias, dizziness, feeling unsteady on the feet, disturbed balance, shortness of breath.

- b. Neuroendocrine manifestations: Recurrent hot and cold spells, cold extremities, subnormal body temperature, unusual sweating, intolerance of extremes of heat and cold, marked weight change, worsening of symptoms with stress.
- c. Immune manifestations: Recurrent sore throat or flu-like symptoms, tender lymph nodes in neck or armpits, new sensitivities to food, odors, or chemicals.

The description presented here is altered slightly from the criteria presented in this volume for the convenience of parents and caregivers.

### ***PROGNOSIS IN PEDIATRIC PATIENTS***

Scientific reviews show that 54 to 94% of the pediatric CFS cases report full or significant partial recovery. Only a minority of children and adolescents remain severely disabled. However, the research issues in pediatric CFS are more complex than in adult, and at present debilitated. Subsequently, no specific predictive factors are known. It is possible that the degree of severity in the first few years of pediatric CFS may predict likelihood of recovery. Once the diagnosis is made, alternative explanations for disabling fatigue, such as diabetes or malignancy, are rare. In general, it seems that the sooner a diagnosis is made, the sooner patients recover up to their pre-morbid level.

### ***HOW TO PUT THEORY INTO PRACTICE***

Profound disruptions in family life occur when parents are confronted with this pattern constellation of symptoms in their child. Concern about physical well-being is complicated by the difficulty in making the diagnosis. But even when the diagnosis is established, children and adolescents should have tasks and responsibilities within the family. Avoiding chores or homework (anything that the individual does not want to do) because of discomfort is called "secondary gain." It is essential that while the physical symptoms need to be respected, they do not necessarily exempt the individual from tasks. Some children may

use physical problems to influence household tasks, to avoid school attendance or to get away from difficult confrontations. In secondary gains, children and adolescents emotionally react to pedagogical rules or educational conditioning. Secondary gains means that children or adolescents should achieve something extra and not losing something they had already acquired. These secondary gains can not be associated to CFS.

When chronic pain is present, it is hard to determine when it is appropriate to ease a particular burden. The best answer to this is to establish the level of activity for pleasurable tasks and then expect the same for less pleasant tasks. For example an adolescent too ill to be present at school for six hours should not be able to play with friends and neighbors for six hours. Common sense is a good guide to work through this difficulty.

Secondly, everyone should have a good working relationship with their medical doctor or medical provider (MD). Some families are fortunate enough to have developed a trusting relationship with a medical provider (MD) who has been with them prior to the onset of the illness, and can thus see the changes the young person is experiencing. As discussed before, however, many providers are reluctant to make the diagnosis because standard laboratory testing is normal. It is helpful for the parents to draw up an outline of both symptoms and activity prior to visiting the provider. A one-week activity log, detailing activity, including leisure time, can be very helpful. An understanding of the relationship between reduced activity (fatigue) and other symptoms of CFS is difficult but important to communicate to medical providers. A diagnosis of CFS should not be made if alternative explanations of reduced activity or altered behavior is present.

Finally, children might suffer from social or school phobia, depression, trauma, mobbing/teasing or a lack of communication skills. The latter means that some children do not know how to express their needs to their environment. The language of physical symptoms is universal and therefore an easy way for children to get their needs met. This situation is not the same as secondary gain because the language of bodily symptoms is only used to acquire basic needs such as a feeling of security.

All above-mentioned conditions should be properly addressed before any diagnosis is provided. School or educational personnel will inquire about these conditions. Subsequently, the more efficient approach is to screen for these conditions.

***MANAGEMENT OF PEDIATRIC CFS***

Although the prognosis for the vast majority of children and adolescents with CFS is good, a prolonged course is common, and functional disability may occur. Cognitive behavioral therapy (CBT) can be useful in the long term management of these young persons as it will facilitate understanding and coping with the chronic problems that evolve. Issues of symptom control, coping strategies and educational planning will help in identity formation at this critical stage of life. Cognitive behavioral therapy (CBT) can efficiently protect further identity formation because it will introduce feelings of control, success and future perspective; all essential in identity formation. Furthermore, it will prevent unintentional parental reinforcement. In addition, individual symptoms can be targeted with specific medical therapy and introducing relaxation techniques can positively influence potentially stress-producing situations. Emphasis on the importance of acquiring normal coping skills and social competence may promote acceptance of recommendations for regular school attendance, which could begin with a partial-day attendance plan.

Institution of a well-integrated, multidisciplinary program is important to address the many areas imposed by pediatric CFS. These areas include physical pain, family interactions, peer interactions, short and long term education, management of emotions, and access to appropriate referral sources to meet the development of independence and autonomy. Every community will vary as to the particular skills available, and the parents need to steer through the maze to obtain those services that are of greatest need for their child.

Parents and children/adolescents should receive the most efficient treatment plan, according to the needs of the patient. Therefore, treatment plans should be adapted to the illness processing or the adaptation to a chronic and debilitating condition. Patients in crisis, for instance, should not be exposed to an active treatment plan because they are in shock, lack perspective, lack disease-insight due to a lack of knowledge and, more importantly, are passive. After stabilization, active therapeutic intervention such as CBT can successfully be introduced.



## APPENDIX

### Definition of ME/CFS for Children: Checklist for Parents or Caregivers

- I.  Clinically evaluated, unexplained, persistent or relapsing chronic fatigue over the past 3 months that:
- Is not the result of ongoing exertion
  - Is not substantially alleviated by rest
  - Results in substantial reduction in previous levels of educational, social and personal activities
- II.  The concurrent occurrence of the following classic ME/CFS symptoms, which must have persisted or recurred during the past three months of illness. Symptoms may predate the reported onset of fatigue.
- Post-exertional malaise and/or post-exertional fatigue.  
With activity (it need not be strenuous and may include walking up a flight of stairs, using a computer, or reading a book), there must be a loss of physical or mental stamina, rapid/sudden muscle or cognitive fatigability, post-exertional malaise and/or fatigue and a tendency for other associated symptoms within the patient's cluster of symptoms to worsen. The recovery is slow, often taking 24 hours or longer.
  - Unrefreshing sleep or disturbance of sleep quantity or rhythm disturbance.  
May include prolonged sleep (including frequent naps), disturbed sleep (e.g., inability to fall asleep or early awakening), and/or day/night reversal.
  - Pain (or discomfort) that is often widespread and migratory in nature. At least one symptom from any of the following:
    - Muscle and/or joint pain
    - Abdominal pain, nausea, vomiting
    - Head pain/headache
    - Chest pain
    - Pain on sensory stimuli (light, noise or odors)
  - Two or more neurocognitive manifestations:
    - Impaired memory
    - Difficulty focusing and concentrating
    - Difficulty finding the right word
    - Slowness of thought

- Need to focus on one thing at a time
- Trouble expressing thought
- Difficulty comprehending information
- Frequently lose train of thought
- New trouble with math or other educational subjects

At least one symptom from two of the following three categories:

- Autonomic manifestations:
  - Neurally mediated hypotension
  - postural orthostatic tachycardia
  - delayed postural hypotension
  - palpitations
  - dizziness
  - shortness of breath
- Neuroendocrine manifestations
  - Recurrent feelings of fever and chills
  - sweating episodes
  - cold extremities
  - worsening of symptoms with stress
- Immune manifestations:
  - Recurrent flu-like symptoms
  - Sore or scratchy throat
  - Tender lymph
  - New sensitivities to foods or allergens

III.  Exclusionary conditions: Evaluation by a physician to exclude severe medical (e.g., lupus erythematosus, HIV, or multiple sclerosis) or psychiatric condition (drug abuse, psychosis, untreated severe eating disorder) that could cause the symptom pattern.

Note: Diagnosis of pediatric ME/CFS may co-exist with fibromyalgia, multiple chemical sensitivities, treated medical disorders, mild to moderate anxiety disorder or depression.