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# New Pediatric Case Definition Helps Pediatricians Diagnose Chronic Fatigue Syndrome (ME/CFS)

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**T**here is a new, pediatric case definition for Chronic Fatigue Syndrome (CFS) developed by an international working group of the International Association for Chronic Fatigue Syndrome (IACFS/ME). The definition appears in print in the Journal of Chronic Fatigue Syndrome (Jason et al., 2006) and is available on the IACFS/ME website ([www.iacfs.net](http://www.iacfs.net)). Chronic Fatigue Syndrome (CFS) is an illness that affects more adult women than men, but also occurs in children and adolescents. Diagnosing CFS is difficult because there is no clinical test indicating a positive diagnosis. The diagnosis of CFS in adults depends upon the patient meeting the criteria of the generally accepted, international case definition (Fukuda et al. 1994). Diagnosing children using the adult case definition has been very problematic. The Fukuda case definition does not capture all adult patients with CFS, and captures even fewer children and adolescents with CFS. Hence the development of the new pediatric case definition containing diagnostic criteria specific for children and adolescents is a big step forward. Pediatricians reading about CFS need to be aware that CFS is the name used in the United States, whereas, Myalgic Encephalomyelitis (ME) is the name used in Europe and elsewhere. To be inclusive, the acronym ME/CFS is gaining popularity worldwide and is also used in the new pediatric case definition (Carruthers et al. 2003, and Jason et al. 2006). For a child or adolescent to be diagnosed as having ME/CFS under the new criteria the following conditions need to be present:

- Unexplained, persistent, or relapsing fatigue that has lasted for at least three months. The fatigue is not the result of ongoing exertion, nor is it relieved by rest. The fatigue must result in a substantial reduction in previous activities
- The concurrent persistence or recurrence of symptoms, from each of the following five groups, for at least three months:
  1. Post-exertional malaise, fatigue, or worsening of other symptoms, with loss of mental and/or physical stamina, and delayed recovery of more than 24 hours
  2. Un-refreshing sleep, disturbance of sleep quantity or rhythm, daytime hypersomnia, nighttime insomnia and/or day/night reversal
  3. Widespread or migratory pain, which can be located in the muscles, the joints (without signs of inflammation), the abdomen, the chest, the eyes (or sensitivity to light), or an increase in severity of headaches, or nausea or vomiting
  4. Two or more neuro-cognitive manifestations, including impaired short term memory, difficulty in concentration or focusing, difficulty finding words or numbers, absent mindedness, slowness of thought, difficulty understanding information and expressing thoughts, educational difficulties
  5. At least one symptom from two of the following three subcategories:
    - a. Autonomic manifestations, including neurally mediated hypotension, postural hypotension, postural orthostatic tachycardia, palpitations, dizziness, shortness of breath, disturbed balance

b. Neuro-endocrine manifestations, including feeling of feverishness, cold extremities, low body temperature, sweating, intolerance to heat or cold, change of appetite or weight. Symptoms worsen with stress

c. Immune manifestations, including recurrent flu-like symptoms, sore throats, fevers and sweats, tender lymph nodes, new sensitivities to food, medicines, odors, or chemicals.

ME/CFS is a diagnosis of exclusion. Many illnesses with symptoms that mimic ME/CFS symptoms preclude the diagnosis of ME/CFS: untreated hypothyroidism, sleep apnea, narcolepsy, malignancies, leukemia, active hepatitis, multiple sclerosis, juvenile rheumatoid arthritis, lupus erythematosus, HIV/AIDS, severe obesity, untreated celiac disease, Lyme disease, mononucleosis, juvenile fibromyalgia, childhood schizophrenia or psychotic disorder, bipolar disorder, active alcohol abuse, active eating disorder, major depressive disorder. Other medical and psychiatric illnesses are not necessarily exclusionary: school phobia, separation anxiety, anxiety disorders, somatoform disorders, depressive disorders, fibromyalgia, multiple chemical sensitivities, any other medical condition that has been adequately treated, before or after the onset of the ME/CFS symptoms, any isolated unexplained physical abnormality or laboratory test which is insufficient to explain the presence of an exclusionary condition.

In summary, the case definition diagnoses ME/CFS primarily on the basis of pathological fatigue, the pattern of other symptoms, and the exclusion of other fatiguing illnesses by medical history, physical examination, and appropriate tests. It is important to make the diagnosis as early as possible. While routine testing may yield normal results, specialized testing may show various abnormalities in immune, nervous and/or cardiovascular systems. Changes in cellular energy production are detectable in some patients. Adequate rest and other supportive treatment may lessen the impact of this illness. Dramatic improvement is more likely to occur in the first four years when treatment is provided. Recovery rates of up to 40% have been reported.

Not covered in the case definition but information helpful for diagnosing pediatric ME/CFS is knowing that adolescents 12 to 17 years of age are more likely to develop ME/CFS than younger children. Be aware that children as young as 4 years of age have been diagnosed with ME/CFS. In adolescents, the onset usually starts suddenly with a fever and flu-like symptoms.

Diagnosis at this point may be difficult because although the symptoms are severe, the routine blood tests are normal. Moreover, the patient must be ill for three months before a diagnosis of ME/CFS can be officially made. A gradual onset of ME/CFS occurs less frequently and is more common in younger children. In younger children, the onset may take months or several years.

## New Pediatric Case Definition

Diagnosing ME/CFS in younger children is further complicated by the inability of younger children to appreciate that their fatigue and other symptoms are abnormal. In such cases, the diagnosis of ME/CFS may be made retrospectively when the child is older. Children and adolescents with ME/CFS often look well. A lack of obvious external physical signs of illness may mean that the first sign of illness will be a marked limitation in either physical or mental activity, which is usually first noticed by a parent or teacher. The external, well appearance of a ME/CFS child may lead to the accusation of school avoidance behavior (school phobia) or the citing of a parent for Munchausen's syndrome by proxy. ME/CFS usually occurs as sporadic cases of the illness. Interestingly, for 20% of patients, more than one family member has the illness suggesting either an environmental or genetic link. Clusters of cases, or outbreaks of the illness have been found worldwide. In many of these outbreaks the illness has been prominent in

school-age children. The prevalence of ME/CFS in children and adolescents is uncertain. Different prevalence studies have used different criteria to diagnose ME/CFS and some have not distinguished children with chronic fatigue symptoms from children with possible ME/CFS. The severity of ME/CFS varies. Some children are severely disabled and bedridden, while others can go to school and a few are capable of playing sports. Most children are between these two extremes. The pattern and the severity of symptoms experienced by a child may change markedly from day to day or during the day. It is important to listen to what the child has to say about the severity of his/her symptoms. Remissions and relapses are common. Relapses may be caused by overexertion or by other infectious illnesses. Over time, slow improvement is likely. Children whose health improves to near pre-illness levels are likely to find that they need more rest than their contemporaries.

### **References:**

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3. Carruthers BM, et al. ME/CFS: Clinical working case definition, diagnostic and treatment protocols. *J. CFS.* 2003; 11(1):7-115.

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## **Diagnosing Pediatric/Adolescent Chronic Fatigue Syndrome**

For a child or adolescent to be diagnosed with Chronic Fatigue Syndrome (ME/CFS) using the new Pediatric case definition the following symptoms need to be present:

- Pathological fatigue and at least seven other symptoms (see below)
- The fatigue and the symptoms are unexplained and persist, or are relapsing for at least three months. They result in a substantial reduction in previous activities
- Other fatiguing illnesses must be excluded by history of the illness, by physical examination or by medical tests.

Symptoms:

Details of symptoms:

Pathological fatigue: The fatigue is not the result of ongoing exertion. It is not relieved by rest.

Post-exertional malaise: Mild or moderate exertion is followed by malaise, fatigue, or worsening of other symptoms, with loss of mental and/or physical stamina, and delayed recovery of more than 24 hours

Sleep problems: Sleep is un-refreshing, with a disturbance of quantity or rhythm, including daytime hypersomnia, nighttime insomnia and/or, day/night reversal

Pain, (at least one symptom): Pain can be widespread or migratory. It can be located in the muscles, the joints (without signs of inflammation), the chest, the abdomen (or nausea or vomiting), the eyes (or a sensitivity to light), or a new type, or an increase in severity of headaches

Two or more neurocognitive manifestations

These include impaired short term memory, difficulty in concentration or focusing, difficulty finding words or numbers, absent mindedness, slowness of thought, difficulty understanding information and expressing thoughts, educational difficulties

At least one symptom from two out of these three subcategories:

1. Autonomic manifestations: including neurally mediated hypotension (NMH), postural hypotension, postural orthostatic tachycardia (POTS), palpitations, dizziness, shortness of breath, disturbed balance
2. Neuro-endocrine manifestations: including feeling of feverishness, cold extremities, low body temperature, sweating, intolerance to heat or cold, change of appetite or weight. Symptoms worsen with stress
3. Immune manifestations: including recurrent flu-like symptoms, sore throats, fevers and sweats, tender lymph nodes, new sensitivities to food, medicines, odors, or chemicals.