HDSA

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Jo Anne B. Barnhart
Commissioner of Social Security
Social Security Administration
P.O. Box 17703
Baltimore, Maryland 21235-7703

Revised Medical Criteria for Evaluating Neurological Impairments
Advance Notice of Proposed Rulemaking

Dear Ms. Barnhart:

Thank you for the opportunity to provide input on the Social Security Administration's plan to update and revise the rules to evaluate neurological impairments of adults and children who apply for, or receive, disability payments from the Social Security Administration, in particular as that criteria applies to individuals with Huntington's Disease.

The Huntington's Disease Society of America (HDSA) is the national organization representing our HD families across the United States. HDSA funds research, provides services and referrals to families, and educates both the public and healthcare professionals about this fatal genetic disorder.

Individuals, family member, social workers and doctors at our HDSA Centers of Excellence report a variety of difficulties and delays in obtaining social security disability benefits for those affected by this disease. The difficulties in obtaining benefits can be categorized as follows:

- The absence of readily apparent dysfunction especially if the chorea or movement disorder has not yet appeared or is the more subtle symptoms such as loss of fine motor coordination and the slowing of eye movements;
- The belief on the part of the social security caseworker that the applicant and his or her symptoms will improve with treatment;
- The relative youth of the applicant, especially for individuals applying in their thirties and forties;
- The patient's lack of insight (which is a symptom of the disease itself) about the progression of the disease with the result that the affected individual is not able to give an accurate assessment of his or her condition to the caseworker;
- Difficulty, given the often subtle onset of symptoms, in pinpointing exactly when a person with HD first became disabled;
- Lack of familiarity with Huntington's Disease on the part of many caseworkers.

We believe that these issues could, and should, be addressed in a revised Listing of Impairments in the so-called "Blue Book". An improved listing would save time, money, resources and emotional energy on the part of the Social Security Administration and the individuals and families it serves who suffer with the effects of Huntington's Disease.

NEUROLOGICAL-ADULT

The Neurological overview provides expansive explanations of epilepsy, brain tumors, persistent disorganization of motor function, multiple sclerosis and traumatic brain injury. Blue Book at §11.00. Huntington's Disease does not fit within any of those categories.

The more specific "Category of Impairments, Neurological", provides as follows:

11.17 Degenerative disease not listed elsewhere, such as Huntington's chorea, Friedreich's ataxia, and spino-cerebellar degeneration. With:

A. Disorganization of motor function as described in 11.04B; or
B. Chronic brain syndrome. Evaluate under 12.02.

The disorganization of motor function found in 11.04B provides as follows:

11.04 Central nervous system vascular accident. With one of the following more than 3 months post-vascular accident:

A. ….
B. Significant and persistent disorganization of motor function in two extremities, resulting in sustained disturbance of gross and dexterous movements, or gait and station (see 11.00C).

This definition is accurate as far it goes. However, the definition is not extensive enough to explain the complexities of Huntington's Disease ("HD"), its age of onset, or the fact that while physical therapy and psychiatric treatment may temporarily ameliorate, and briefly stabilize, the symptoms of the disease, there is no treatment to reverse the relentless course of HD.

HD has a triad of clinical features, including motor abnormalities, dementia, and disorders of mood and perception. The early symptoms of HD are usually much more subtle than recognized by the Blue Book's emphasis on the movement disorder component of HD. This emphasis is highlighted by the fact that the Blue Book refers to Huntington's Disease as Huntington's chorea.

The first symptoms of HD in many HD patients are emotional and psychiatric – personality changes, irritability, mood swings, depression, obsessive-compulsive behavior, inability to concentrate and decreased motivation. Individuals with HD, even in the relatively early stages, have particular difficulty with decision-making, multi-tasking and performing under time pressure or with the stress of inter-personal interactions, especially in technical or complex jobs. Obviously such difficulties make it progressively difficult to perform satisfactorily in the work place.

For many individuals struggling with HD, it is the behavioral and mood changes much more than the “disorganization of motor function” that interferes with their ability to work.

The description of Huntington’s chorea found at §11.17 includes “Chronic brain syndrome. Evaluate under 12.02.” The emotional, psychiatric and cognitive problems caused by HD are indeed abnormalities associated with dysfunction of the brain. However, the psychological and behavioral abnormalities associated with HD encompass more than the disorders listed in §12.02. Individuals with HD commonly suffer from affective disorders such as severe depression, sleep disturbance, feelings of worthlessness, difficulty concentrating or thinking, paranoia, thoughts of suicide, and, in some cases, hallucinations. “Affective disorders” are covered in §12.04. In addition, generalized persistent anxiety, obsessive compulsive behavior and panic attacks resulting in marked restriction of activities of daily living, difficulties in social function and concentration commonly disable individuals with HD. These “anxiety-related” disorders are covered in §12.06.

As a medical definition, these “affective” and “anxiety” disorders in individuals with HD are caused by the ongoing neurological damage, and are part of the “organic brain disorder” caused by HD. However, an individual caseworker with no context in which to review these symptoms may not understand that HD is relentlessly progressive and these seemingly behavioral based problems will not improve over time.

The term Huntington’s chorea and the limited information in the Blue Book does not accurately reflect the complexity of the disease and results in needless delays, denials and difficulties for individuals with HD and their families. The relative rareness of the disease makes it especially important that the Blue Book give clear guidance as to the complex nature of this disease and its many emotional and cognitive manifestations beyond the “chorea” with which HD has historically been associated.

Huntington’s Disease needs its own explanatory section under section 11.00 putting the complexity of the disease, its early age of onset, its irreversibility and the subtle nature of the early symptoms in context. Without this context, individuals with HD and their families will continue to struggle to obtain SSDI benefits.

NEUROLOGICAL-CHILDHOOD
JUVENILE HD

The diagnosis of HD in an adult is usually made in a person who has a family history of HD, motor abnormalities, cognitive changes, behavioral or psychiatric problems. The presenting symptoms may be a little different in a child, particularly a child under 10 years. While there is no symptom or group of symptoms that are absolutely required for the diagnosis of juvenile HD, most affected children have several of these features at the time that the diagnosis is made. Chorea is uncommon in children developing HD within the first decade, but may be one of the first symptoms in a teenager. Severe behavioral disturbance may be the first symptom in an adolescent.

The Neurological Listing for evaluating impairments of children under age 18 includes convulsive epilepsy, nonconvulsive epilepsy, motor dysfunction and impairment of communication. Blue Book at §111.00. The specific categories of neurological

impairment includes: major motor seizure disorder, nonconvulsive epilepsy, brain tumors, motor dysfunction, cerebral palsy, meningomyelocele and communication impairment associated with documented neurological disorder.

While seizures are said to occur in about 25% of children with juvenile HD, HD is not a form of epilepsy. Younger children are likely to have motor dysfunction, most commonly rigidity and dystonia. However, the typical initial symptoms of juvenile HD are a positive family history of HD, usually in the father, stiffness of the legs, clumsiness of arms and legs, decline in cognitive function, changes in behavior, seizures and changes in oral motor function. As with adult onset HD, these problems cut across the neurological listing and the mental disorders listing.

To minimize the difficulty faced by families attempting to obtain social security disability benefits for their children, we request a separate listing for the juvenile form of HD under the general neurological listing and a specific listing under the specific categories of neurological impairments.

CONCLUSION

The Huntington's Disease Society of America respectfully requests that revisions to the Listing of Neurological Impairments include:

1. References to Huntington's Disease, not Huntington's chorea;

2. A separate heading under § 11.00 describing the pathology of HD. A suggested format is attached as Exhibit A.

3. A separate listing for HD, revised to include a cross-reference to the suggested revision in 2 above and to include Affective Disorders, Anxiety Related Disorders, in addition to Organic Mental Disorders for evaluating HD. A suggested format is attached as Exhibit B.

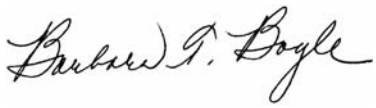
4. A separate heading under § 111.00 describing the pathology of Juvenile Huntington's Disease. A suggested format is attached as Exhibit C.

5. A separate listing for Juvenile HD to include cross-references to the applicable sections under Mental Disorders. A suggested format is attached as Exhibit D.

The Huntington's Disease Society of America will provide any medical research, documentation or testimony that may be necessary or helpful in your review process upon request.

Thank you for the opportunity to participate in this important work.

Sincerely,



Barbara T. Boyle.
National Executive Director/CEO

SUGGESTED FORMAT FOR REQUESTED CHANGES TO LISTING OF NEUROLOGICAL IMPAIRMENTS – ADULT
HUNTINGTON'S DISEASE

EXHIBIT A

Listing under § 11.00. Proposed new paragraph

G. *Huntington's Disease*. HD is an inherited neuropsychiatric disorder that is progressive and terminates in death of the affected person. Recovery or remission never occurs. Treatment is ineffective in terms of halting or slowing the progression of the disease. The usual age of adult onset is between the ages of 30 and 50, although the age of adult onset may be younger or older. Incapacitation occurs relatively early in the course of this debilitating illness with progression to total disability and dependency for all activities of daily living. There are three characteristic clinical features: (1) loss of ability to control bodily movements; (2) loss of ability to think and act quickly, to learn new material and to remember, and (3) apathy, personality changes, irritability, mood swings, depression, anxiety, inability to concentrate, decreased motivation, obsessive-compulsive disorder and severe depression. Individuals with HD also exhibit poor social judgment and may be irritable and aggressive. Inability to work is due to a combination of cognitive disturbance, behavioral or mood changes, poor coordination of voluntary movements, the presence of involuntary movements. Individuals with HD, even in the relatively early stages, have particular difficulty with decision-making, multi-tasking and performing under time pressure or with the stress of interpersonal interactions. The course of the disease varies among individuals and families. The cognitive and behavioral problems may become debilitating before disorganization of motor functions. For other individuals, the motor dysfunction may appear first.

EXHIBIT B

Proposed revised categorical listing.

11.15 ***Huntington's Disease***. With
A. Disorganization of motor function as described in 11.04B; or
B. Chronic brain syndrome. Evaluate under 12.02, 12.04, and 12.06.

SUGGESTED FORMAT FOR REQUESTED CHANGES TO LISTING OF NEUROLOGICAL IMPAIRMENTS - CHILDHOOD
JUVENILE HUNTINGTON'S DISEASE

EXHIBIT C

Listing under § 111.00. Proposed new paragraph:

E. *Juvenile Huntington's Disease*. While there is no symptom or group of symptoms that are absolutely required for the diagnosis of juvenile HD, most affected children offer several of the following features at the time that the diagnosis is made: motor dysfunction, characterized by rigidity and dystonia, seizures, declining cognitive function, behavioral or psychiatric problems such as depression, aggressiveness and impulsiveness, irritability, mood swings, and obsessions. Huntington's Disease is a hereditary disorder and individuals with very early onset of HD are far more likely to have an affected father than an affected mother.

EXHIBIT D

Proposed revised categorical listing

110.10. *Juvenile onset of Huntington's Disease.*

A. Motor dysfunction. Evaluate under 111.06; or
B. Behavioral or psychiatric problems. Evaluate under 112.02, 112.06 and 112.08