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Post-polio syndrome

Proposal ID : 1116 - **Proposal State :** Accepted **Proposal for Update**

Implementation Date : 1/2010

Originator : *Lori Moskal* - **Last Update made by :** *Lori Moskal*

Creation Date : *22-Aug-2006 23:15 CET* - **Last Update :** *09-Jan-2009 19:31 CET*

Previously Discussed in the group(s): MBRG

Primary Code Affected : G10-G14

Secondary Codes Affected : None

Volumes Affected : 1,3

Proposal Type : Addition of new code

Change Reason : Need to reflect a change in clinical knowledge

Detailed Description

Add code to block level:

List of three-character categories

Diseases of the nervous system

(G00-G99)

Systemic atrophies primarily affecting the central nervous system (**G10- ~~G13~~G14**)

G10 Huntington's disease

G11 Hereditary ataxia

G12 Spinal muscular atrophy and related syndromes

G13* Systemic atrophies primarily affecting central nervous system in diseases classified elsewhere

G14 Postpolio syndrome

Add excludes note:

B91 Sequelae of poliomyelitis

Excludes: postpolio syndrome (**G14**)

Add category:

Systemic atrophies primarily affecting the central nervous system (**G10-G13G14**)

G14 Postpolio syndrome

Postpolio myelitic syndrome

Excludes: sequelae of poliomyelitis (B91)

Add excludes note:

M89.6 Osteopathy after poliomyelitis

Use additional code (B91), if desired, to identify previous poliomyelitis.

Excludes: postpolio syndrome (G14)

Special tabulation lists for mortality and morbidity

Tabulation list for morbidity (298 causes)

Note: These lists were adopted by the World Health Assembly in 1990 for the tabulation of data. They are described, and their use is explained, in Volume 2, the Instruction Manual.

Tabulation list for morbidity

129 Other diseases of the nervous system

**G10-G13 G14, G21-G26, G31-G32,
G36-G37, G46-G47, G60-G73, G90
-G99**

Volume 3

Add subterms and lead term:

Postpoliomyelitic - see also condition

- osteopathy **M89.6**

- syndrome **G14**

Postpolio syndrome G14

Syndrome - see also Disease

....

- postphlebitic **I87.0**

- postpolio (postpoliomyelitic) G14

- postvagotomy **K91.1**

Rationale

1. Justification:

1.1 Definition

PPS is a neurological disorder characterized by a new or reoccurring muscular weakness and/or muscular fatigue abnormal in individuals who had acute poliomyelitis, many years before (DALAKAS, 1995).

Although this aggravation is a late effect of poliomyelitis, it is about a defined nosological entity, that we believe cannot be identified as a sequelae, since it characterizes itself as an evolving disease of diverse etiology and physiopathology compared to basic disease.

PPS is classified as motor neurone disease due to its histological and clinical chart being intimately related with the functions of inferior motor neurons. (DALAKAS, 1995).

1.2 Risk Factors

The risk factors to the development of PPS have not, until now, been elucidated accordingly. Based on repeated observations and epidemiological reports, however, the following factors seem to be associated with a precarious beginning of PPS: (1) New symptoms appear first, or could be firstly in the limbs previously damaged and in patients with more aggravated paralysis; (2) Precarious bulbar or respiratory difficulties occur in residual patients with loss of muscular strength of bulbar intervention and respiratory musculature; and (3) Symptoms occur precarious in patients who have acute poliomyelitis in a later age.

1.3 Physiopathology

Jubelt et al in 1999 conducted a study demonstrating the existence of 9 theories that explain the physiopathology of PPS, however the most welcome is of the "Super-training" formulated initially by Charcot, in 1875, and complimented and proven through electrophysiological studies by Dalakas et al since 1995.

In the acute infection of poliomyelitis, the virus damages the cells of the anterior medullary horn partially or totally, with denervation of some motor units through the neuronal plasticity, blossoming occurs renovating the denervated fibers although this last one depends on the number of preserved neurons after this period of recovery, we go to a period of latency, also called stability plateau. According to the theory of "super-training", thirty or forty years after the acute disease through the metabolic solicitation request from the giant motor units formed due to the neuronal recovery, there is the beginning of motor neuronal failure, mainly in the distal portions of the *axonius*, bringing it to a new denervation known as new weakness and muscular atrophy.

1.4 Clinical Aspects

The symptoms and signals of PPS include a combination of muscular skeletal symptoms of the progressive muscular atrophy, post-poliomyelitis (AMPP) (DALAKAS, 1995).

These symptoms classified as more frequent include: new muscular weakness, new muscular atrophy with or without the presence of muscular and articular pain. There is still other symptoms although less frequent in evidence: muscular weakness of bulbar innervations, intolerance to cold, myalgia, fasciculations, new breathing difficulties, and sleep apnea.

In the research developed in UNIFESP-EPM, we have verified the presence of other forms of clinical occurrences including sleep disorders, increase of corporal weight, memory disorders, dizziness, syncope, and morning headache related to sleep disorder. Studies conducted in diverse populations show the same characteristics having percentual as the only variation. Another important

factor is the duration of the interval between the acute disease and the beginning of PPS symptoms is a determining factor with the highest rate of incidence of thirty to forty years.

1.5 Incidence and Prevalence

This may vary according to the definition of PPS, from diagnosed criteria used and also of population research undertaken.

Brazil: In depictive studies made in the Neuromuscular disease ward of Escola Paulista de Medicina UNIFESP, were found respectively 68% and 77.2% of incidents among the patients that were consulted (OLIVEIRA, 2002; QUADROS, 2004).

Japan: In a study conducted in the RECOVERY WARD of Cosmos Hospital, Usuki city, County of Ohita, found 85% of incidents and a prevalence of PPS in Kitakyushu was of 18 cases/100,000 inhabitants (TAKEMURA, 2004).

Norway: The study in 1994 was made through national medical reports, and of social situations, and in a total of 2392 cases of poliomyelitis victims, many of whom registered in "the national association of polio victims" in Norway with a prevalence of 85% of PPS (GRELH, 1996).

Germany: A study conducted found evidence of 68.07% of PPS in patients. (TIENVOLL, 1997).

New Zealand: A loss of muscular strength was related in 47%, and it is estimated that there maybe a number of 3000-5000 polio survivors in New Zealand, that may be suffering from PPS (CHETWYND, 1993).

Australia: It is estimated that at least 20,000 – 40,000 individuals developed paralyzing poliomyelitis from 1930-1988.

Scotland: A study conducted in Edinburgh found 67% of PPS (PENTLAND, 1999).

Denmark: 54% of PPS was found (LONNBERG, 1992).

Sweden: 18.6% of PPS was found in a research (AHLSTROM, 1993).

Canada: It is estimated that 66,000 survivors exist and that approximately two thirds have or will develop PPS (URIADKA, 1997).

United Kingdom: 74,280 cases were reported and the incidence of PPS found was 77% (BRUNO, 1997).

Europe: There is nearly 250,000 patients of PPS in Europe and 20,000, 000 around the world (BOSCH, 2004).

2. Final Considerations

Considering that PPS is an evolving clinical situation irreversible and incurable, related to the progressive dysfunction of the motor units, it cannot and should not be classified as a polio sequelae. Sequelae means (see thesaurus/dictionary). In ICD-10 a poliomyelitis situation is contemplated with the codes: A-80, A-80.0, A-80.1, A-80.2, A-80.3, A-80.4, A-80.9 and B-91, being in this way not possible to characterize PPS as a nosological entity defined in these categories.

In November of 2003, a group of associations of PPS in Europe, and 20 members of the European Parliament met and agreed to create the European Union of Polio having as a goal to obtain recognition and funds of the European Parliament (BOSCH, 2004).

It becomes fundamental for post-polio syndrome to be recognized as a nosological entity and it should have a specific characterization in ICD-10 due to the relevance of the legal and social assistance.

3. References:

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Uriadka C. Physiotherapy management of the late effects of polio. Post-polio clinic, West Park Hospital, Toronto, Ontario, Canada, 1997.

Proposal held over in 2007.
URC secretariat

This proposal was accepted in 2008.
URC

Supporting Publications (Uploaded Files)



Proposal Summary

This item was discussed at the URC session of WHOFIC in Trieste, 2007 and it was decided to refer it to the MBRG for further work on the best fit within the classification for a new code and possible subcategories.

10.4.08: Revised proposal attached for comments.

Comments

23-May-2007 14:50 CET by **Michael Schopen**

Comment attached to the vote of the user for Round 1 of year 2007. Voted:Can't Decide

We are seeking expert advice.

28-Jun-2007 05:31 CET by **Julle Rust**

Comment attached to the vote of the user for Round 1 of year 2007. Voted:Can't Decide

Seeking clinical advice

21-Aug-2007 10:06 CET by **Robert Jakob**

Comment attached to the vote of the user for Round 2 of year 2007. Voted:Can't Decide

seeking advice

30-Aug-2007 11:56 CET by **Olafr Steinum**

Comment attached to the vote of the user for Round 2 of year 2007. Voted:Yes

We support this proposal. Our neurologist suggests, however, that subcategories of G14 might be useful.

06-Sep-2007 09:23 CET by **Michael Schopen**

Comment attached to the vote of the user for Round 2 of year 2007. Voted:Can't Decide

We hope to have expert opinion for Trieste.

07-Sep-2007 09:37 CET by **Julie Rust**

Comment attached to the vote of the user for Round 2 of year 2007. Voted:Can't Decide

Still following up clinical advice - hopefully for the meeting in Trieste.

10-Sep-2007 19:40 CET by **Donna Pickett**

Comment attached to the vote of the user for Round 2 of year 2007. Voted:Can't Decide

Still obtaining clinical advice

10-Sep-2007 22:02 CET by **Lori Moskal**

Comment attached to the vote of the user for Round 2 of year 2007. Voted:Can't Decide

Still trying to get clinical input.

18-Feb-2008 03:32 CET by **Julie Rust**

Comments from Australia

There are a number of options for the inclusion of this syndrome in ICD-10;

1. Add 'post-polio syndrome' as an inclusion term at G12.2 Motor neuron disease (attached rationale and other papers indicated that it is a motor neuron disease) and also code the B91 Sequelae of poliomyelitis
2. Create a new code at G14 and expand category G10 - G13 (to G14) to include this - also code out B91.

However, we think that this proposal raises the bigger issue of who decides when a condition is a recognised disease (and what criteria are applied?). There could be similar arguments for inclusion in ICD-10 of a number of other conditions and we're not sure that the responsibility for inclusion rests with the URC.

18-Feb-2008 03:36 CET by **Julie Rust**

ICD Neurological application

Just for information, ICD-NA (1997) has expanded the category B91 in the following manner:

- B91 Sequelae of poliomyelitis
 - B91.-0 Progressive postpolio muscular atrophy
 - B91.-1 Postpolio pain syndrome due to joint deformity
 - B91.-2 Postpolio pain syndrome, idiopathic
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28-Mar-2008 20:00 CET by **Donna Pickett**

Post Polio Syndrome

The U.S. has received a response from the Neuromuscular Disease Section of the American Academy of Neurology (AAN) regarding Post-Polio Syndrome.

They support the creation of a new code and note that post polio syndrome is probably not a late effect. Based on this information the U.S. agrees with the creation of a new category G14 which could be titled to allow for further expansion in the future, and create a new code under the new category (G14.x).

Below is a discussion thread from Medlink Neurology that accompanied the AAN response.

Etiology

It is now recognized that normal aging alone cannot explain the development of post-polio syndrome because the normal loss of anterior horn cells and motor units does not become prominent until after age 60 (Jubelt and Cashman 1987). More important than a patient's chronological age is the interval from their acute polio to the onset of post-polio syndrome, an interval that averages between 30 and 40 years (Jubelt and Cashman 1987). The presently accepted most likely etiologic possibilities are degeneration of enlarged motor units, a chronic persistent poliovirus infection or an immune-mediated disease.

Degeneration of enlarged motor units. The enlarged motor units that develop via sprouting after the acute polio may never fully stabilize (Wiechers 1985). Findings from single fiber electromyographic (SFEMG) studies reveal that the largest motor units are more likely to become unstable later in life (Cashman et al 1987a; Emeryk et al 1990), and with increasing time from the acute polio, neuromuscular transmission becomes more unstable, as increased jitter and blocking occur (Wiechers and Hubbell 1981). Spontaneous denervation activity, jitter, and blocking occur more frequently in symptomatic muscles (Ryniewicz et al 1990; Maselli et al 1992). These findings are supported by muscle biopsy studies that describe an increasing number of angulated fibers accumulating over time (Dalakas and Illa 1991). This is followed by degeneration of axonal branches as demonstrated by the appearance of small group atrophy (Drachman et al 1967; Cashman et al 1987a). This can be followed by large group atrophy suggesting neuronal degeneration (Dalakas and Illa 1991). It has been frequently hypothesized that the increased metabolic demand of an increased motor unit territory results in premature exhaustion and death of the motor neuron (Jubelt and Cashman 1987). Even though there are no definitive studies examining the cell soma to prove this theory; electrophysiologic and muscle biopsy data appear to be supportive. The overuse of weakened muscles results in excessive muscular fatigue (Sharma et al 1994; Grimby et al 1996; Agre et al 1998; Sunnerhagen et al 2000; Thomas and Zijdwind 2006), which appears to contribute to the excessive metabolic demand on motor neurons and the premature exhaustion.

Chronic persistent poliovirus infection. Poliovirus and other picornaviruses can persist in the CNS of animals and cause delayed or chronic disease (Jubelt and Cashman 1987; Destombes et al 1997). Poliovirus and other enteroviruses can also persist in the CNS and systemically in immunodeficient children (Jubelt and Cashman 1987). Studies in tissue culture have found that poliovirus mutants can persist without killing the host cell (Colbere-Garapin et al 1989; Borzakian et al 1992) and can also persist in neurons (Pavio et al 1996). Support for the persistent poliovirus hypothesis was enhanced by the findings of Sharief and colleagues (Sharief et al 1991), who demonstrated poliovirus antibodies and poliovirus-sensitized cells in the CSF of post-polio patients. Leon-Monzon and Dalakas (Leon-Monzon and Dalakas 1995) found elevated IgG poliovirus antibodies in the sera of post-polio syndrome patients as compared to controls; however, ALS patients had similar elevated levels. Other investigators have been unable to find poliovirus antibodies in the CSF of post-polio patients (Kurent et al 1979; Dalakas et al 1986; Salazar-Gruesso et al 1989; Melchers et al 1992; Jubelt et al 1995). CSF specimens have also been examined for the presence of poliovirus RNA by polymerase chain reaction, and the majority of studies have been negative or inconclusive (Melchers et al 1992; Leon-Monzon and Dalakas 1995; Leparç-Goffart et al 1996; Muir et al 1996). The most positive study was that of Julien and colleagues (Julien et al 1999) who detected poliovirus genome sequences in the CSF of 11 of 20 post-polio syndrome patients but in none of the 20 control patients. These same authors had reported similar findings, post-polio syndrome 5 of 10 positive, controls 0 of 23 positive, in an earlier study (Leparç-Goffart et al 1996). Conclusive viral isolation and histochemical or hybridization studies have not as yet been reported using spinal cord tissues and will be required to resolve this possibility. An immune-mediated disease. The strongest support for an inflammatory or immune-mediated mechanism for post-polio syndrome stems from the study of Pezeshkpour and Dalakas (Pezeshkpour and Dalakas 1988) in which inflammation in the spinal cords of seven post-polio patients was found. It consisted of both perivascular and parenchymal lymphocytic infiltrates, neuronal degeneration, and active gliosis. All changes were more prominent in three patients with new weakness. Other findings that support this hypothesis are the finding of oligoclonal bands in the CSF (Dalakas et al 1986) and activated T-cells in the peripheral blood (Ginsberg et al 1989). Others have not found oligoclonal bands in post-polio syndrome patients (Cashman et al 1987a; Salazar-Gruesso et al 1989); however, other histologic studies suggest an immune-mediated or viral-induced pathogenesis or at least an inflammatory mechanism. Miller (Miller 1995) examined the spinal cord from one post-polio patient and found perivascular intraparenchymal chronic inflammatory infiltrates primarily composed of B lymphocytes with rare macrophages and no T-cells. Kaminski and colleagues (Kaminski et al 1995) found inflammation in the spinal cords of 8 of 9 post-polio syndrome patients. More recent studies supporting an immune-mediated process is the finding of inflammatory cytokines (TNF-alpha, IFN-gamma, IL-4, IL-10) in the CSF of post-polio syndrome patients (Gonzalez et al 2002; 2004).

21-Apr-2008 23:54 CET by **Olafr Steinum**

Let's go!

Let us go for G14 and move the proposal to URC.

22-Apr-2008 10:05 CET by **Marion Mendelsohn**

proposal to URC

I agree with the comment of Olafr

29-Apr-2008 15:52 CET by **Ulrich Vogel**

Let's move it to URC

We agree with Olafr's comment.

18-Jun-2008 20:02 CET by **Robert Jakob**

Comment attached to the vote of the user for Round 1 of year 2008. Voted:Yes

clear evidence for this change

28-Aug-2008 20:10 CET by **Lori Moskal**

Comment attached to the vote of the user for Round 2 of year 2008. Voted:Yes

Should there be an excludes note added at M89.6 Osteopathy after poliomyelitis
Excludes: Post-polio Syndrome (G14)

30-Aug-2008 11:14 CET by **Glen Thorsen**

Comment attached to the vote of the user for Round 2 of year 2008. Voted:Yes

Support Lori's suggestion of exclusion note

