Abnormal Movements in Sleep as a Post-Polio Sequelae

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Dr. Richard L. Bruno

The Post-Polio Institute
International Centre for Polio Education

ABSTRACT

Nearly two-thirds of polio survivors report abnormal movements in sleep (AMS), with 52% reporting that their sleep is disturbed by AMS. Sleep studies were performed in seven polio survivors to objectively document AMS. Two patients demonstrated Generalized Random Myoclonus (GRM), brief contractions and even ballistic movements of the arms and legs, slow repeated grasping movements of the hands, slow flexion of the arms and contraction of the shoulder and pectoral muscles. Two other patients demonstrated Periodic Movements in Sleep (PMS) with muscle contractions and ballistic movements of the legs, two had PMS plus Restless Leg Syndrome (RLS) and one had sleep starts involving only contraction of the arm muscles. AMS occurred in Stage II sleep in all patients, in Stage I in some, and could significantly disturb sleep architecture even though patients were totally unaware of muscle contractions. Poliovirus-induced damage to the spinal cord and brain is presented as a possible cause of AMS. The diagnosis of post-polio fatigue, evaluation AMS and management of AMS using benzodiazepines or dopamimetic agents is described.

Abnormal Movements in Sleep as a Post-Polio Sequelae

Despite numerous late-onset symptoms reported by polio survivors -- fatigue, muscle weakness, pain, cold intolerance, swallowing and breathing difficulties -- one symptom was totally unexpected: abnormal movements in sleep (AMS). As early as 1984 our post-polio patients were reporting muscle contractions as they fell asleep. The 1985 National Post Polio Survey included two questions about AMS: "Do your muscles twitch or jump as you fall asleep" and "Is you sleep disturbed by muscle twitching?" (1) It was surprising that 63% of the 676 respondents reported that their muscles did twitch and jump during sleep and that 52% -- a third of the entire sample -- said that their sleep was disturbed by twitching.

These percentages are markedly elevated as compared to the incidence of AMS in the general population. In one survey only 29% of those without neurological disease who were at least 50 years old reported AMS, versus 63% of surveyed polio survivors who were 52 years old on average. (2) In another survey only 34% of those older than 64 reported AMS, slightly more than half the incidence of AMS in the younger post-polio sample. (3)

Given the apparent increased prevalence of AMS in polio survivors, and with daytime fatigue the most commonly reported Post-Polio Sequelae (PPS), we were

interested in objectively documenting AMS, relating them to possible disturbances in sleep architecture and identifying an effective treatment for AMS. (1)

METHODS

Subjects. Seven polio survivors were referred for sleep studies to a sleep disorders center. This was a sample of convenience, in that the subjects were patients presenting with PPS who themselves knew (three patients) or whose bed mates knew (four patients) that AMS were occurring. Patients were on average 54 years old and 44 years post acute polio, which occurred at age 10. The patients had had AMS for a mean of eight years, which was on average 35 years post acute polio. Patients reported moderate-to-severe difficulty sleeping at night and moderate-to-severe daytime fatigue that did not respond to the treatments of choice for post-polio fatigue (i.e., pacing of activities, daytime rest periods, energy conservation and use of appropriate assistive devices. (4) In addition to fatigue, patients reported an average of two limbs having late-onset muscle weakness.

Procedure. Patients underwent a standard polysomnographic evaluation with EEG and facial EMG recorded for sleep staging. (5) Blood oxygen saturation, measured using a finger pulse oxymeter, chest and abdominal wall excursion and nasal air temperature were also recorded; video monitoring of sleep was also performed. Surface EMG was recorded from patients' legs as well as from limbs in which AMS were reported.

RESULTS

Four types of AMS were seen: Two patients presented with what has been called Generalized Random Myoclonus (GRM) (1,6), two patients had Periodic Movements in Sleep (PMS), two had PMS plus Restless Leg Syndrome (RLS) and one had Sleep Starts (also called "hypnogogic massive myoclonic jerks").

Generalized Random Myoclonus. GRM was seen in two patients. One had had bulbar polio with little or no arm or leg involvement acutely, while the other has been diplegic since polio but had no bulbar or respiratory symptoms acutely. These patients had random contractions of muscles throughout their bodies. One had such violent contractions of the trunk muscles that she was pulled into the fetal position during the night. This patient had been very aware of GRM for about 10 years.

However, the other patient had been completely unaware of GRM until they were noticed by her husband (Figure 1). Random, rapid muscle contractions were noted in all four limbs, jaw and pectoral muscles, in addition to slow repeated grasping movements of the hands, slow flexion of the arms and movement of the shoulders. The presence of bilateral toe flexion was notable since the patient's right leg has always been totally paralyzed except for a minimal ability to flex her toes; the toes of her right foot contracted numerous times the during
the night.

In both patients, GRM occurred during Stage II sleep; the latter patient also had GRM in Stage I. The patient with violent trunk flexion had muscle contractions, causing a severe disturbance of sleep architecture, only during the first third of the night. She also had a few episodes of obstructive apnea that were not related to the muscle

contractions but did disturb her sleep. She was prescribed clonazepam, 0.5 mg B.I.D., which eliminated her GRM. The other patient had GRM throughout the night but had no disturbance of sleep architecture and was not treated pharmacologically.

Periodic Movements in Sleep. Two patients demonstrated PMS with contractions only of the leg muscles of which neither patient was aware. Both had limb and respiratory involvement with the acute polio and had PMS during Stage II sleep with one patient also having muscle contractions during Stage I. The former patient had nearly continuous EMG activity in his legs throughout the night and had a severe disturbance of sleep architecture (Figure 2). He also had some central episodes of apnea early in the night as he was falling asleep that did not disturb his sleep. The latter patient had PMS occurring only during the first half of the night that caused no disturbance of sleep architecture. However, he had frequent hypopneas that did severely disturb his sleep. Both patients were prescribed lorazepam, 1.0 mg H.S., which eliminated the PMS.

PMS plus Restless Leg Syndrome. Two patients had PMS plus Restless Legs Syndrome. RLS is characterized by the subjective feeling that the legs must be moved. This feeling increases during the evening, often preventing sleep onset because patients feel as if they must get up and walk. The patients with PMS plus RLS had been very little affected by the acute polio, one having no polio residual and the other having one leg weakened. PMS were seen in both legs and occurred during Stage II in both patients and during Stage I in one patient. One patient's leg muscle contractions were so violent that she was propelled one to two inches off the surface of the bed. Although her PMS occurred only during the first half of the night, her sleep was severely disturbed and she was very aware that she had had PMS for about 5 years. She was prescribed L-dopa/carbidopa (Sinemet) 200/50 mg, 1/2 tablet B.I.D., and clonazepam, 0.5 mg H.S. and at 3 A.M., which reduced the RLS and PMS by about 80% and allowed her to have a restful nights sleep.

The other patient did not know he had PMS which were continuous throughout the night and did moderately disturb his sleep architecture. He was prescribed L-dopa/carbidopa, 200/50 mg H.S., which eliminated his RLS and PMS.

Sleep Start. One patient was diagnosed as having a Sleep Start, her arms ballistically abducting as she began to fall asleep. She was very mildly affected by the acute polio and had no AMS in the legs, even in the leg in which she reported new muscle weakness. The patient's sleep was markedly disturbed since her arms would move as she started to fall asleep and prevent sleep onset. She was prescribed alprazolam, 0.125 mg H.S., which eliminated her AMS.

DISCUSSION

Sleep studies in this sampling of post-polio patients objectively documented three different types of AMS. Whether other types or combinations of AMS occur in polio survivors cannot be determined from this study, nor can this study or the 1985 National Post-Polio Survey state the actual incidence of AMS in polio survivors, since neither sample was random or population-based. However, the objective documentation of AMS in these post-polio patients, and the Post-Polio Survey finding that 63% of polio survivors reported muscle "twitching or jumping"as they fell asleep, suggest that AMS may in some way be related to the pathophysiology of the original poliovirus infection.

In 1964, Loeb coined the phrase “hypnic myoclonus” to describe muscle contractions during sleep onset in healthy individuals without neurological disease. (7) Loeb described the contractions as rapid (less 0.5 seconds long), arrhythmic (occurring without a pattern) and causing a variety of movements - finger flexion, thumb adduction, forearm and foot flexion and extension, shoulder elevation and facial twitching during Stages I and II sleep - identical to those seen in the post-polio patients. However, in contrast to the post-polio patients, none of Loeb’s subjects demonstrated contractions in more than one muscle group and in none had sleep disturbed by hypnic myoclonus.

Loeb thought that hypnic myoclonus resulted from an abnormality at the level of the brain stem reticular formation causing decreased descending inhibition of anterior horn motor neurons during sleep. Martinelli (8) thought PMS also resulted from an increase in anterior horn cell excitability, with Walters (9) finding that PMS decreased with the administration of an opiate receptor agonist.

Loeb and Askenasy suggested that AMS were also related to abnormal discharges from the thalamus, cerebellum and basal ganglia. (7,10) The implication of the basal ganglia in the generation of AMS is interesting since PMS are common in patients with Parkinson's disease, whose decreased dopamine production impairs basal ganglia functioning, in patients with narcolepsy, who have an increased nu
mber and sensitivity of dopamine receptors in the basal ganglia, and have been found to decrease with the administration of dopamine receptor agonists. (9,11,12)

AMS, Polioencephalitis and Poliomyelitis. All of the CNS regions implicated in the pathogenesis of AMS are known to have been lesioned by the poliovirus. The anterior horn motor neurons, cerebellar nuclei and reticular formation were frequently and severely damaged by the poliovirus. (13) The periaquiductal gray, paraventricular hypothalamus and lamina II dorsal horn neurons were all lesioned by the poliovirus (cf. 9;13); damage to these opioid peptide-secreting neurons may be evidenced not only by AMS but also by polio survivor's doubled sensitivity to pain. (14-16) Finally, the thalamus and basal ganglia (the substantia nigra, putamen and globus pallidus) were also damaged by the poliovirus, damage that has been implicated in the pathogenesis of post-polio fatigue. (16-18) Given the distribution and extent of poliovirus lesions in all of the CNS areas implicated in the pathogenesis of AMS, we should not have been surprised in 1985 that a majority of polio survivors reported muscles that twitch and jump during sleep.

Clinical Implications. PPS remains a diagnosis of exclusion. All possible causes for new symptoms in polio survivors, especially causes for late-onset fatigue, must be ruled out before them diagnosis of PPS is made. Therefore, it is important to rule out a sleep disorder as a cause of late-onset fatigue. Clinicians need to take a thorough sleep history from their post-polio patients, asking not only about symptoms of sleep apnea, which occurs frequently in polio survivors, but also about AMS. (20) The patient’s bed partner must also be asked about AMS since the majority of polio survivors will not know that they have AMS.

Patients are referred for a sleep study if sleep apnea or AMS is suspected. The lowest dose of a short acting benzodiazepine will be prescribed before sleep by the Post-Polio Institute physiatrist if a patient has AMS, since these medications seem to virtually eliminate GRM and PMS in our post-polio patients. Treatment of sleep apnea is deferred to the sleep disorders center as is treatment for RLS, since a dopamimetic agent in

combination with a benzodiazepine may be required. However, there is a caveat to prescribing dopamimetics for polio survivors. One of our PPS patients developed vasovagal syncope with cardiac asystole during the administration of a dopamine receptor agonist. (21) We consider a history of vasovagal syncope or unexplained faints a contraindication to prescribing dopamimetics for polio survivors with AMS or RLS. (22)

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