

## Case Report

# Benign neonatal sleep myoclonus in a child

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### ABSTRACT

Benign neonatal sleep myoclonus (BNSM), is a disorder generally mistaken for seizures during the newborn period. Benign neonatal sleep myoclonus is featured by myoclonic "lightninglike" jerks of the extremities that exclusively occur during sleep; it is not associated with epilepsy that occur only during sleep and cease abruptly when the child is agitated. This case reported was a 50 days-old baby boy with a history suggestive of abnormal movements for limbs noted over the preceding 7 days. Diagnosis of BNSM in infant based on history taking, clinical manifestation, and with no electroencephalographic changes. BNSM is usually not associated with any other neurologic impairment and spontaneously subsides within the first year of life. Its importance lies in the differential diagnosis with the epileptic, especially myoclonic, seizures of infancy. BNSM can be misinterpreting for neonatal seizures or even neonatal status epilepticus, the recognition of benign sleep myoclonus of infancy is imperative to elude unnecessary diagnostic studies and treatments.

**Keywords:** Benign neonatal sleep myoclonus, Clinical criteria, Diagnostic criteria, Infant, Newborn, Non-epileptic events

### INTRODUCTION

Benign neonatal sleep myoclonus (BNSM) is a non-epileptic movement disorder and self-limiting disorder that develops in the neonatal period.<sup>1</sup> This phenomenon of the first weeks of life characterized by neonatal onset myoclonic jerks during non-rapid eye movement (NREM) sleep, and consistent cessation with arousal with no electroencephalographic changes.<sup>1</sup> Myoclonus is a clinical sign characterized by sudden, brief, jerky, shock-like involuntary movements.<sup>2</sup> Other have suggested that a transient disorder of the serotonergic system might play a role in the pathogenesis of this disorder.<sup>3</sup> Siblings have been seen with BNSM, suggesting a genetic link, but this also remains speculative.<sup>4</sup> The prevalence of most reports stated that the BNSM is unknown but assumed that the condition is common and likely underrecognized. Two reports roughly estimated an incidence that varies

between 0.8 and 3.0 cases per 1000 births.<sup>5</sup> Neurologically normal term infants sometimes present with repetitive, rhythmic myoclonic jerks that occur during quiet sleep but stop with awakening. This common condition may be induced by rocking the infant or by repetitive sounds. The recognition of BNSM will avoid unnecessary diagnostic test and antiepilepsy treatment also for accurate family counseling regarding the prognosis.<sup>6</sup>

### CASE REPORT

A 50 days old baby boy who was admitted to the outpatient clinic at Dr. Soetomo General hospital. The subject were having no complaints were reported until he started to present multiple abnormal movement at a 9 days old, with a frequency of one to three per second, affecting all limbs, especially the upper extremities,

occurring in clusters and only during sleep. There were no changes in skin color, eye deviation, face movements, heart rate during any of the events. He remained sleepy with the jerks worsening. The jerks started shortly after the initiation of rocking and became more noticeable with increasing frequency of the rocking. The repetitive jerks would continue during the maneuver and for a short time after its conclusion. The jerks did not stop by holding the limb. The medical history revealed that he was delivered spontaneously at term, by healthy mother and visit to the health care regularly. There was no history of illness, drugs or traditional herbs taken during the pregnancy. There were spontaneously crying, no cyanosis, no convulsion and no icteric. The birth weight was 2800 gram with head circumference 36 cm. He was breastfed until now. He already got Hepatitis B, Polio and BCG vaccinations. He could keep eye contact with the parents and respond with smiling. There were no history of seizure and vomiting before. Family history revealed there were no history of seizure, hypertension nor diabetes mellitus.

In addition, on the physical examination the baby boy was an alert, active and well-nourished boy, the body weight was 4000 gram, the height was 55 cm and head circumference 37 cm the pulse was 140 beat / minute, respiratory rate 30 breath/minute and the temperature 36.60C. There were no neurological impairment. There were no lymph node enlargements in the coli region. The chest movement was symmetric, no signs of respiratory distress. The heart was normal without murmur and gallop. The breath sound in the both lungs was vesicular without rales and wheezing. The abdomen was normal, ascites and meteorismus were not found and the bowel sound was normal. There were no organomegaly. The extremities were red, dry, warm with capillary refill time less than 2 seconds and no edema was found.

Neurological examination revealed, a boy with composmentis consciousness. Pupil was symmetric, 3 mm/3 mm with strong light reflex. Meningeal signs were negative. Pathological reflexes babinski, chaddock reflexes and clonus were negative. Motoric evaluation impressed no lateralization. Physiological reflexes were normal.

From the (Figure 1) and (Figure 2) the growth chart of the patient revealed well-nourished and normocephaly, and with this condition have no problems with growth development.

Laboratory and chest X-ray's result was normal. Electroencephalography (EEG) planned to perform. EEG initially on January 15th, 2015 revealed within normal limit on the (Figure 3). The ictal EEG shows there isn't brief bursts of generalized spikes-and-waves or polyspikes and waves. Conventional sleep and awake EEG's were normal waves. Based on the history taking, clinical manifestations and laboratory data the patient was diagnosed with benign neonatal sleep myoclonus.

The patient didn't given any medication. Treatment of this patient is an advice for both parents to report and monitoring in case such as repetitive jerks get worse, should evaluated the duration of the jerks. The parents suggested to continue the neurological evaluation on the Neurology pediatrics outpatient clinic for the follow up.

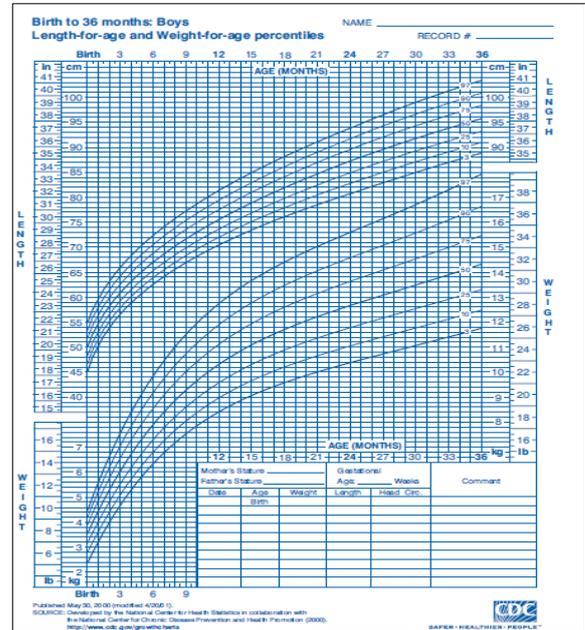


Figure 1: Growth chart of the patient revealed normal.

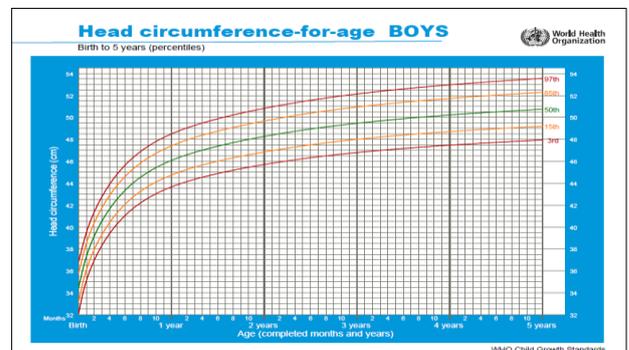


Figure 2: Head circumference of the patient revealed normocephaly.

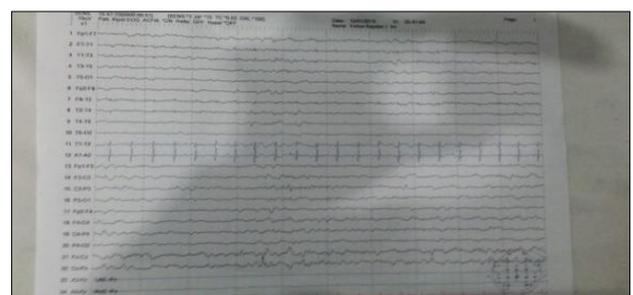


Figure 3: Electroencephalography revealed normal limit, no epileptiform waves are seen.

## DISCUSSION

Benign neonatal sleep myoclonus (BNSM) defined as a self-limiting disorder characterized by neonatal onset myoclonic jerks during non-rapid eye movement (NREM) sleep, and consistent cessation with arousal with absence of concomitant electroencephalographic findings.<sup>7</sup> A transient serotonin imbalance apart from genetic factors may be involved in its etiology.<sup>8</sup> The myoclonic jerks in BNSM can be focal, multifocal or generalized. They characteristically appear only during NREM sleep and stop consistently when the child is aroused. This feature of BNSM helps in its recognition as well as its differentiation from neonatal seizures.<sup>1</sup> The majority of the neonates have generalized myoclonia, mainly in the distal part of limbs, jerking is of short duration, lasting 10 to 20 seconds though they may persist for more than 30 minutes.<sup>1,8,9</sup> The general condition of the child is unaffected and there are no associated abnormalities on neurodevelopmental examination. The jerks of BNSM may be precipitated by stimuli such as sound or touch or even by benzodiazepines.<sup>10</sup> Manoeuvres such as rocking in a head-to-toe direction and repetitive sound stimuli have been described as activating procedures for myoclonic jerks in BNSM. These methods of provocation were not routinely tested in group of infants, but were observed by the parents of three children, thus confirming the fact that simple manoeuvres may help to differentiate this benign condition from other paroxysmal motor events.<sup>11</sup>

Benign neonatal sleep myoclonus is disorder characterized by myoclonic jerks in an otherwise healthy infant. The myoclonic jerks have been described as being bilaterally symmetric, with involvement of both upper and lower extremities. The myoclonus may be synchronous or asynchronous. Notably, the jerks are seen during sleep and are never present during the awake state. Benign neonatal sleep myoclonus is a nonepileptic condition and is not associated with epileptiform discharges on electroencephalography.<sup>3</sup>

It also defined BNSM as a rare condition characterized by nonepileptic spasms. It begin before age one year and self-limited disease. The electroencephalogram is invariably normal, and neurologic development is not affected.<sup>12</sup> Benign myoclonus of early infancy is characterized by its onset between 3 and 15 months and by clusters of tonic or myoclonic seizures with frequent involvement of the head. Although the clinical features and natural history of benign myoclonus of early infancy have been well characterized, the etiology is not clear. It may represent an exaggeration of physiologic myoclonus of early infancy.

Myoclonus is a sudden, brief, involuntary muscle contraction. Cortical myoclonus involves a brief (20-75 ms) contraction of agonist and antagonist muscles.<sup>7</sup> The tonic contractions in children with benign myoclonus of early infancy, however, may last up to 2 to 4 seconds.<sup>12</sup>

Benign neonatal sleep myoclonus can be differential diagnosed from benign familial neonatal seizures by the following criteria:

- The movements only occur during sleep,
- They stop abruptly and consistently when the child is aroused,
- Electrographic seizures do not occur during events.<sup>1-3</sup>

By definition, the involuntary movements only occur in sleep but may be severe and prolonged enough to mimic convulsive status epilepticus. They may be worsened by touch, simple restraint, and anticonvulsant treatment, but cease promptly on arousal. An EEG is not essential for diagnosis if attention is paid to clinical information, but it may be required to distinguish benign neonatal sleep myoclonus from benign neonatal seizures; the absence of concomitant electrographic change on an EEG done with surface electrodes would likely exclude an epileptic basis for generalized myoclonus but not for focal myoclonus.

EEG was a little assistance in making the diagnosis of benign familial neonatal seizures, although they also pointed out that EEGs had either not been performed in many of the cases reported or the results were insufficiently described. Many of the patients diagnosed as having benign neonatal seizures without information on concomitant EEG changes may well have had benign neonatal sleep myoclonus. The precise characterization of paroxysmal events requires concurrent EEG data.

In this case conventional asleep and awake EEGs were normal. According to Daoust-Roy et al, there are no associated EEG abnormalities in BNSM, which has been confirmed by long term video-electroencephalographic monitoring. An EEG may be required at times to distinguish BNSM from neonatal convulsion.<sup>1</sup> The jerky movements should be differentiated from jitteriness/clonus, which occur due to poor myelination of corticospinal tract in neonates. The closest alternative diagnosis is benign myoclonus of early infancy, which may rarely have a neonatal onset and have a normal EEG but these typically occur when the infant is awake.<sup>1</sup>

In this case, the jerk was getting improved day by day. BNSM is a self-limiting disorder with spontaneous resolution between 2 to 6 months of age and a normal neurodevelopmental outcome on follow up.<sup>1</sup> Both the cases described fit into the above description of BNSM with typical myoclonic jerks occurring only during sleep and an entirely normal neurological status and outcome. The course is self-limited and not affected by treatment. Spells normally decrease considerably within 3 months of onset and usually stop before age 2 years. Neurologic development is always normal. Recognition as an innocuous and non-epileptic entity is important so that needless treatment, unnecessary investigations and undue hospitalization can be avoided, and parental anxiety minimized.<sup>12</sup>

## CONCLUSION

It has been reported a rare case of benign neonatal sleep myoclonus in a 50 days-old baby boy. The diagnosis based on history taking, physical examination and electroencephalography findings. The benign neonatal sleep myoclonus, is a self-limiting disorder. The prognosis is good and the patient didn't given any medication and long term follow up were needed.

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