



SLEEPING BEAUTY SYNDROME

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ABSTRACT Kleine-Levin syndrome (KLS), also known as Sleeping Beauty syndrome, generally experience recurrent episodes of the condition for more than a decade and may return at a later age. Individual episodes generally last more than a week. Some times lasting for months. The condition greatly affects the personal, professional, and social lives of sufferers. The severity of symptoms and the course of the syndrome vary between sufferers. Patients commonly have about 20 episodes over about a decade. Several months generally elapse between episodes. The onset of the condition usually follows a viral infection; several different viruses have been observed to trigger KLS. It is generally only diagnosed after similar conditions have been excluded; MRI, CT scans, lumbar puncture, and toxicology tests are used to rule out other possibilities. The syndrome's mechanism is not known, but the thalamus is thought to possibly play a role. Tomography has shown hypoperfusion in the brains of patients during episodes.

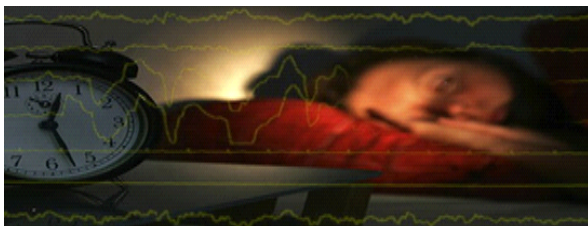
KEYWORDS :

INTRODUCTION

Kleine-Levin syndrome is a rare disorder that primarily affects adolescent males (approximately 70 percent of those with Kleine-Levin syndrome are male). MacDonald Critchley, who first wrote about the condition in 1942, described 11 cases he had examined and reviewed 15 other published cases in a 1962 publication. In the report, which included patients he had examined in the Royal Navy World War II, observed that irritability and depersonalization often occurred while patients were awake. He named the condition Kleine-Levin syndrome and noted four common traits: hypersexuality, adolescent onset, spontaneous resolution, and compulsive eating. He believed that the condition only affected males, but later studies showed some female patients. In the 1970s, several psychoanalytic psychodynamic for the condition were proposed. In 1980, a Hawaiian-Caucasian family was found in which nine family members suffered from the condition.

DEFINITION

Kleine-Levin syndrome (KLS), also known as Sleeping Beauty syndrome, is a rare sleep disorder characterized by persistent episodic hypersomnia and cognitive or mood changes. Many patients also experience hyperphagia, hypersexuality and other symptoms.



EPIDEMIOLOGY

Population-based studies of KLS have not been performed. Its prevalence is about 1 case per million people. In France, KLS has a prevalence of 1.5 per million people. It occurs most frequently among Jews in the US and Israel. First-degree relatives of people who have suffered from the syndrome are much more likely than the general population to suffer from it, although only in about one percent of cases do family members contract it. About 70 to 90 percent of patients are male. Patients with the syndrome are more likely than the general population to have genetic disorders.

CAUSES

- The thalamus probably plays a role in the out-of-control sleeping.
- Patients with diencephalic-hypothalamic dysfunction caused by tumors experience symptoms similar to those of KLS patients.
- The involvement of the thalamus, temporal lobe, and frontal lobe of the brain suggests that there is a multifocal, localized encephalopathy. There are also persistent subclinical abnormalities in KLS sufferers.
- Another possible explanation concerns the metabolism of

serotonin and dopamine. An imbalance in the neurotransmitter pathways of these chemicals could play a role.

- Viral infections
- Androgen might (indirectly) block melatonin receptors, possibly by means of vasodilation, and cause cholinergic abnormalities in some cases of Kleine-Levin syndrome.

SYMPTOMS

It is characterized by recurring but reversible periods of excessive sleep (up to 20 hours per day). Symptoms occur as "episodes," typically lasting a few days to a few weeks.

- Episode onset is often abrupt, and may be associated with flu-like symptoms.
- Excessive food intake,
- Irritability,
- Childishness,
- Disorientation,
- Hallucinations,
- An abnormally uninhibited sex drive may be observed during episodes.
- Mood can be depressed as a consequence, but not a cause, of the disorder.

Affected individuals are completely normal between episodes, although they may not be able to remember afterwards everything that happened during the episode. It may be weeks or more before symptoms reappear.

DIAGNOSIS

KLS can be diagnosed when there is confusion, apathy, or derealisation in addition to frequent bouts of extreme tiredness and prolonged sleep.^[5] earliest it can be diagnosed is the second episode, KLS is rare, and other conditions with similar symptoms are usually considered first.

MRIs can determine if the symptoms are caused by certain brain disorders, stroke, and Multiple Sclerosis. Lumbar puncture determine if there is Encephalitis. KLS must be differentiated from substance abuse by toxicology tests.^[14] use of Electroencephalography (EEG) can exclude temporal status epilepticus consideration. EEGs are normal in about 70% of KLS patients, but background slowing may sometimes be detected. In addition, low-frequency high-amplitude waves can be observed during waking hours.

Initially, KLS appears similar to bipolar depression.

with frontal-lobe syndromes Klüver-Bucy syndrome display similar symptoms, but these conditions can be differentiated by the presence of brain lesions should also be distinguished from very rare cases of menstruation-caused hypersomnia.^[3]

TREATMENT

Several drug therapies have been used on patients with KLS. A 2016 Cochrane Review concluded that "No evidence indicates that pharmacological treatment for Kleine-Levin syndrome is effective and safe". Stimulants, including modafinil reported to have a limited effect on patients, often alleviating sleepiness.

They can cause behavioral problems, lithium decrease the length of episodes and the severity of their symptoms and to increase the time between episodes it is effective in about 25 to 60 percent. Its use carries the risk of side effects in the thyroid or kidneys. Anti-psychotics and benzodiazepines can help alleviate psychotic and anxiety related symptoms, Carbamazepine less effective than lithium but more effective than some drugs in its class

KLS patients generally do not need to be admitted to hospitals. It is recommended that caregivers reassure them and encourage them to maintain sleep hygiene. It may also be necessary for patients to be prevented from putting themselves in dangerous situations, such as driving

PREVENTION

Lithium is the only drug that appears to have a preventive effect. Anti-depressants do not prevent recurrence.

PROGNOSIS

The frequency of KLS episodes can vary from attacks one week in length occurring twice a year to dozens of episodes that follow each other in close succession

Unusually young or old patients and those who experience hypersexuality tend to have a more severe course. Patients who initially have frequent attacks generally see the disease cease earlier than others. The condition spontaneously resolves, the patient is considered to be cured if there have been no symptoms for six years.

CONCLUSION

There is no definitive treatment for Kleine-Levin syndrome and watchful waiting at home, rather than pharmacotherapy, is most often advised. Stimulant pills, including amphetamines, methylphenidate, and modafinil, are used to treat sleepiness but may increase irritability and will not improve cognitive abnormalities. Because of similarities between Kleine-Levin syndrome and certain mood disorders, lithium carbamazepine is prescribed and, in some cases, have been shown to prevent further episodes. This disorder should be differentiated from cyclic re-occurrence of sleepiness during the premenstrual in teen-aged girls, which may be controlled with birth control pills. It also should be differentiated from encephalopathy, recurrent depression, or psychosis.

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