Kleine-Levin Hibernation Syndrome

Alternative Names
Kleine-Levin Syndrome

Record Category
Disease phenotype

WHO-ICD
Diseases of the nervous system > Episodic and paroxysmal disorders

Incidence per 100,000 Live Births
0-1

OMIM Number
148840

Mode of Inheritance
Autosomal dominant

Gene Map Locus
N/A

Description
The Kleine-Levin hibernation syndrome (KLS) is a rare autosomal dominant disorder characterized by episodes of behavior and cognitive disturbance, and hypersomnia. The condition may also be associated with episodes of increased feeding, hypersexuality, and/or mood disorders and depression. These episodes may occur from 2 to 12 times every year, with each episode lasting for a few days, or sometimes even up to a month. Hypersexuality is more commonly seen in male patients, while mood disorders are more frequent in females. On the whole, the condition affects more males than females, and age of onset is usually around 15 years.

KLS is an extremely rare disease, affecting one in a million people worldwide. Diagnosis can be made through EEG and brain scintigraphy in addition to a detailed medical history. Since the symptoms of the condition mimic these of severe depression, and each episode may be followed by short periods of high energy, patients may be incorrectly diagnosed with bipolar disorder. Treatment is supportive.

Molecular Genetics
KLS is not typically an inherited condition. However, in a small percentage of cases, familial clustering has been noted. In addition, the fact that the Jewish population is more susceptible to the condition has also suggested a genetic component to the syndrome. In the familial cases, KLS has been shown to follow an autosomal dominant pattern of inheritance. An association of KLS with HLA-DQ2 has been described in some studies. However, there is no known gene to cause KLS. Some familial studies have also indicated autosomal recessive inheritance.

Epidemiology in the Arab World

Saudi Arabia

BaHammam et al. (2008) described a Saudi family with multiple affected individuals with KLS. The parents were close relatives. Six out of 12 individuals in this family were found to be affected; the father, three sons and two daughters. The age of onset of KLS varied among affected family members, and ranged from 15 to 21 years. The signs of KLS was present in two out of six affected individuals. Detailed neurological and psychiatric assessments were unremarkable for all family members. Human leukocyte antigen (HLA) typing was identical in the father and two children. Four out of the six affected members were found to be homozygous at DQB1∗02 loci.

References

BaHammam AS, GadElRab MO, Owais SM, Alswat K, Hamam KD. Clinical characteristics and HLA typing of a family with Kleine-Levin syndrome. Sleep Med. 2008; 9(5):575-8. PMID: 17761456.

Related CTGA Records

External Links
http://klsfoundation.org/what-is-kleine-levin-syndrome/

http://rarediseases.org/rare-diseases/kleine-levin-syndrome/
http://www.orpha.net/consor/cgi-bin/OC_Exp.php?lng=en&Expert=33543

Contributors

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