History of KLS

• First observed in 1925 by Frankfurt Neurologist Willi Kleine. This report was followed four years later by details of similar cases by New York-based psychiatrist Max Levin who published his findings in 1935.

• Foundation started by the Maier and Farber families in 1999.

• Data collection shows cases all over the world with greater incidence on the East and West Coasts of the US and Israel where there are greater concentrations of Jewish people.

• Major clinical research ongoing at Stanford University, Emory University and Sorbonne University in France.
Incidence

- 1:1 million
- Median age at onset is 14 +/- 2 years
- Can occur at any age
- First episode often follows a viral infection
- Clusters of cases noted in Israel, East and West coasts where there is larger Jewish population, suggesting a genetic component
- Median duration of illness is 8 years from the first episode although some patients suffer for much longer.
- Episodes can recur after a long period of wellness and/or continue for decades
• Marked hypersomnia in otherwise healthy individual
• Derealization and cognitive impairment with:
  • Flat affect
  • Childlike behavior and speech
  • Agitation when not allowed to sleep
  • Hyperphagia
  • Hypersexuality
  • Hallucination
  • Delusion
  • Pupils appear dilated and eye are glossy
  • Pallor is common
Episodes

- Patients usually have prodromal symptoms
  - Heavy feeling behind the eyes
  - Headache
  - Feeling sleepy
- Median episode length is one week to 10 days; can last for months or longer
- Patient will sleep 18 to 21 hours per day waking to drink, eat and toilet
- Limited memory of events during episode after emerging to wellness
- Relative normalcy between episodes; some patients report cognitive decline between without full return to baseline
Diagnosis

- Rule Out diagnosis after negative:
  - Tox screen
  - EEG
  - MRI/PET scan – may show hypoperfusion of the hypothalamus
  - Labs
  - Lumbar puncture for CSF
  - Neuro exam

Second episode is highly suggestive of KLS pattern
KLS patient Alanna Wong on Anderson Cooper

There is nothing pretty about kleine-levin syndrome
KLS caregiver Diane Wong on why awareness is so critical
KLS patient Michael Hamper
Treatment

- There is no proven treatment for KLS.
- Lithium has been shown to reduce the frequency and severity of episodes in some patients. Also of some limited value:
  - Antibiotics, Antivirals, Steroids, Xyrem and Romazicon.
- Supportive care AT HOME
- Supervision is necessary while cognitive function is impaired
- Allow patient to sleep and protect from hazardous behavior such as:
  - Driving
  - Operating machinery
  - Going out alone
  - Cooking
  - Bathing
KLS is thought to be a self-limiting disease course although some patients report episodes continuing into adulthood without remitting.

Since KLS frequently occurs during adolescence, assistance to manage absences from school and work will be necessary.

Known triggers for episodes to avoid:
- Illness
- Alcohol consumption
- Drug use
- Stress
- Emotional upset
- Travel
Some patients mistakenly treated with psychiatric protocol are exposed to mental health units where they are not allowed the supporting environment and sleep. This can result in a downward spiral of inappropriate medication and a life blown off course with irreparable socialization to in patient psych environments.

Patients can suffer psychosocial consequences for abnormal behavior during an episode if not diagnosed and kept at home with supervision.

Early identification of KLS is the ideal clinical outcome so that the patient can be given treatment options and the supportive care that is needed.
Please Consider KLS

- Providers have the visibility to patients who present with symptoms suggestive of KLS; please consider what you have seen and heard here; you can save a life from irreparable damage.

- Consultation with a provider familiar with KLS is available:
  - Dr. David Rye – Emory University Sleep Center
    404-712-7533
  - Dr. Chris Hamper – Clinician in Florida
    561-743-9077

- For more information go to www.klsfoundation.org