Idiopathic Hypersomnia (IH) is a rare sleep disorder. The primary symptom is excessive daytime sleepiness (EDS) that is not improved, regardless of how much time is spent asleep. IH tends to be a progressive disorder that responds poorly to traditional treatments. It often impacts upon the patient's life to such an extent that working, socializing and even driving eventually become impossible due to their inability to be awake.

Symptoms
The most common symptoms shared by sufferers of IH are:
- Greater than 10 hours sleep per 24 hour period – often as much as 16+ hours per 24 hour period
- Long unrefreshing naps that normally last several hours
- Awaking from sleep feeling unrefreshed, often with significant sleep inertia (commonly known as ‘sleep drunkenness’)
- An inability to be woken from sleep – even multiple alarm clocks or physical attempts made by family/friends are largely unsuccessful
- Cognitive problems caused by the overwhelming desire to sleep (commonly known as ‘brain fog’)

As the condition progresses less common symptoms can include:
- Anxiety and depression – often as a result of the limits this disorder creates on what is able to done with their time awake
- Raynaud’s type phenomena – freezing cold hands and feet
- Loss of impulse control – especially in regard to food
- Impotence

Those with IH often describe themselves as experiencing two types of sleepiness:
1) A physical exhaustion that ‘normal’ people might experience after missing several nights sleep in a row
2) A cognitive exhaustion similar to Executive Dysfunction that can make even simple tasks like reading, conversation with friends or watching a movie beyond their reach

Diagnosis
IH is a diagnosis of exclusion and often takes many years after the initial symptoms appear. Other more common causes of excessive daytime sleepiness must all be ruled out and diagnosis is normally only made following a sleep study.

Unlike Narcolepsy an overnight sleep study will often show a perfectly normal nights sleep for someone with IH. The Mean Sleep Latency Test (MSLT) is used to rule out Narcolepsy, with IH patients falling asleep quickly but not entering REM in their naps. For those with IH the urge to sleep can be fought to some extent before becoming totally overwhelming and they are unlikely to fall asleep without warning.
Traditional Treatments
Currently there are no treatments available specifically for Idiopathic Hypersomnia. The traditional treatments used are normally prescribed either “off-label” or through purposeful misdiagnosis of the patient with conditions such as Narcolepsy, ADHD, or Depression. The most common treatments prescribed are:
- Amphetamines (such as Dextroamphetamine or Adderall)
- Methylphenidate (such as Ritalin or Concerta)
- Modafinil (such as Provigil or Modavigil)
- Armodafinil (such as Nuvigil or Waklert)
- Sodium Oxybate (such as Xyrem)
- Antidepressants

Unfortunately patients with IH commonly respond poorly to the traditional treatments. Often increasingly large doses are needed and it commonly reaches the point where negative side effects outweigh the benefits.

Recent Research
Since the classification of IH in the mid-1900's very little progress has been made towards understanding the mechanism of action in the patient that might create these symptoms. This all changed late in 2012. A recent paper from researchers at Emory University suggests that the sleepiness of those with IH might be caused by hypersensitivity in their brain to the amino acid known as GABA.

The GABA system is well understood as the primary neurotransmitter responsible for wakefulness. Many medications exist to manipulate this system. For example common sleeping pills, such as benzodiazepines, and general anesthetics, such as propofol, are effective because they work as agonists on the GABA System.

The researchers suggested that the brains of patients with Idiopathic Hypersomnia are hypersensitive to GABA and they are experiencing the same effects a ‘normal’ person might experience if they were put on a continuous dose of sleeping pills or a mild general anesthetic. It’s possible for patients to fight this tiredness/fatigue for a while, but in the medium to long term it becomes impossible for them to function.

Rather than being a ‘sleep disorder’ it might be more useful to consider it as a ‘wakefulness disorder’ where the patient is chemically unable to stay awake for long periods or think clearly during their time awake.

Interestingly, the researchers at Emory University found a similar hypersensitivity to GABA in patients diagnosed with other sleep disorders including Narcolepsy without Cataplexy. It is postulated that IH and Narcolepsy without Cataplexy might turn out to be the same disorder, just at different extremes of a spectrum.

Future Treatments
This discovery opens up several new possibilities for treatments for patients with IH. Rather than using treatments to try and stimulate the brain to be ‘awake’ it is possible to use medicine to counter the hypersensitivity to GABA. The primary contender is a drug known as Flumazenil. Flumazenil was discovered in the 1980's and FDA approved in 1991 for reversing general anesthetic and reversing sleeping pill over-doses in ER. Another possible treatment is an antibiotic called Clarithromycinc that reduces the hypersensitivity to GABA too.

The researchers at Emory University did a small trial with 7 patients suffering from a mixture of IH and Narcolepsy without Cataplexy. They administered flumazenil and recorded a significant improvement in wakefulness in the patients. They have since conducted a clinical trial with 20 patients using Clarithromycin and once again, recorded a significant improvement in wakefulness.

While it is too early to suggest that Flumazenil or Clarithryomycin might become to treatments of choice for IH it is promising to see new research uncovering fresh options. As further trials are conducted and more patients with IH are found to respond, or not respond, this information can serve to help future patients in returning to a more normal life.

Further Information
http://www.livingwithhypersomnia.com
http://clinicaltrials.gov/show/NCT01183312
http://clinicaltrials.gov/show/NCT01146600

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