Observation of a nervous disease attended by disturbed sleep, at times lethargic and at times convulsive. Edmé Chauvot de Beauchêne (1786)

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HISTORICAL NOTE

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Kleine–Levin syndrome (KLS) is a rare disease characterised by recurrent episodes of hypersomnia and, to varying degrees, behavioural/cognitive disturbances, compulsive eating and hypersexuality. Kleine, a neurologist, was the “first” to describe a series of nine cases of recurrent hypersomnia in a 1925 publication; one case involved a young woman. In 1936, Levin, a psychiatrist, added a series of five other cases, focusing on the relationship between hypersomnia and disordered eating. In 1962, Critchley, who gave the disorder its name, added to the literature 11 cases he had personally observed as a physician for the British Royal Navy. In a recent review of the literature, Arnulf compiled 186 cases dating from 1962 to 2004.

Huang and Arnulf report on a description of KLS written by A Brière de Boismont in 1862. In this article, I present a case history of episodic hypersomnia associated with disordered eating and neurological disturbances. The report was written by a French physician, Edmé Chauvot de Beauchêne (1749–1824), and was published in 1786.

Beauchêne completed his studies at the renowned medical school in Montpellier, its reputation dating back to the middle ages. His professors included F Boissier de Sauvages, author of an esteemed nosology. Beauchêne embarked on his career towards the end of the enlightenment and, like his contemporaries P Pomme and J Raulin, he became interested in “vapours”. In his book entitled De l’influence des affections de l’âme dans les maladies nerveuses des femmes avec le traitement qui convient a ces maladies (1781) (How affections of the soul influence nervous diseases in women, and the appropriate treatments for these diseases), he offers a precursor to descriptions of what late 19th century doctors termed hysteria and their 20th century counterparts, psychosomatic illness: “At the age when passions begin to take root in a woman’s heart, she has no vapours; but as these passions develop and intensify, nervous diseases exert the most devastating influences upon her temperament. The violent transports agitating the senses transmit an impetus which accelerates movement, in turn destroying equilibrium in the physical constitution”. After fleeing to his estate in Burgundy to save his neck during the French Revolution, Beauchêne returned to the court of King Louis XVIII during the restoration where he was accorded the illustrious title of “Physician to the Ladies of the Court”.

Given his specialisation in women’s diseases, it is not surprising that Beauchêne took particular interest in the unusual case of a 26-year old woman. Between the ages of 7 and 11, she had experienced episodes of extensive erysipelas, accompanied by fever, digestive problems, headaches and convulsions. “In her fourteenth year, she was overcome with a lethargic sleep which lasted several days; and it was so profound that she was believed dead. From that point forward, the affection of sleep recurred at irregular intervals; it usually lasted eight to ten days, continuing at times for fifteen; and upon one sole occasion, it persisted into the seventeenth day.”

Beauchêne first examined the patient when she came to Paris at the age of 24 years. He personally observed four episodes of hypersomnia, lasting between 24 h and 8 days. He reports: “During the first four years of her disease, this poor girl had appetites as bizarre as they were dangerous, causing her to eat lime, plaster, soil, and vinegar. Thereafter, these appetites subsided, and she nourished herself indiscriminately with all sorts of aliment, excepting bread, for which she maintained an insuperable loathing till she was perfectly cured. This fear always occasioned vomiting”. Beauchêne does not mention megaphagia per se, but his descriptions of recurrent episodes of hypersomnia, accompanied by peculiar eating habits, in some ways resemble KLS.

In contrast, he describes a process of waking which differs from that associated with KLS: “The patient awakened only by degrees, and her reason cleared in due proportion, such that the veil hanging over it was progressively lifted by each convolution, and its development, which reached completion in four or five hours, offered a succession of analogies with the earliest years of life. First the patient smiled in a childlike manner, and as these passions develop and intensify, nervous diseases exert the most devastating influences upon her temperament. The violent transports agitating the senses transmit an impetus which accelerates movement, in turn destroying equilibrium in the physical constitution”.

Given his specialisation in women’s diseases, it is not surprising that Beauchêne took particular interest in the unusual case of a 26-year old woman. Between the ages of 7 and 11, she had experienced episodes of extensive erysipelas, accompanied by fever, digestive problems, headaches and convulsions. “In her fourteenth year, she was overcome with a lethargic sleep which lasted several days; and it was so profound that she was believed dead. From that point forward, the affection of sleep recurred at irregular intervals; it usually lasted eight to ten days, continuing at times for fifteen; and upon one sole occasion, it persisted into the seventeenth day.”

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Figure 1 Title page.
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**OBSERVATION**

Sur une Maladie nerveuse avec complications d’un sommeil lent, avec halleux lithargique, sauvage et hystérique.

Dans tous les temps les Maladies nerveuses ont présenté de grandes difficultés à la Médecine, et il semble que l’Art de guérir n’a pas encore acquis suffisamment les canons qui les enveloppent, pour que l’on puisse savoir avec une certaine de certitude, leur traitement.

La maladie incitait de haute et ses maladies; mais malgré leurs fureurs, ils étaient dépouillés de toutes leurs qualités, et les phénomènes furent nombreux qu’elles développèrent, conduisant ainsi à des conclusions précises que l’on n’arrive pas à cacher même aux plus éclairés médecins, pour ainsi dire même à ceux qui les détruisent. Ainsi le patient passa plusieurs semaines, et imposant à chacun d’eux un calme irréprochable, il l’aida dans l’espoir de les reconnaître, afin de leur parler avec facilité.

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Figure 2 First page of the text.

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References


observed that the initial expression of this sentiment was never a matter of chance, but was in fact always directed towards the person to whose services she was most indebted. Once fully awake and in complete possession of her reason, the patient was overcome by a torrent of tears, which we took to indicate her awareness of her pitiful state.

Such unusual symptoms upon waking—convulsions (but were these what we currently call convulsions?), hiccups, trouble breathing—do not match the classical profile of primary KLS. Arnulf et al reported 18 cases of secondary KLS, involving various neurological disturbances (frontal, bilateral pyramidal and pseudobulbar syndromes) but no cases of epilepsy. KLS may involve cognitive disturbances such as derealisation, hallucinations, irritability and depression. Beauchêne describes pathological behaviour on waking—logorrhoea, dysthymia, and childish behaviour—which may be in keeping with KLS.

Beauchêne never mentions any unusual sexual behaviour. Perhaps there simply were no such symptoms, but the rules of decorum may have prevented him from making any observations on this subject.

Beauchêne notes that the patient’s behaviour returned to normal between the episodes of hypersomnia, as occurs in KLS: “We noted that in the intervals between these periods of sleep, the patient had preserved much of her strength; her colour was normal between the episodes of hypersomnia, as occurs in KLS: The physiopathology of KLS remains unclear. A specific genetic predisposition (HLA DQB1*201) may favour the development of autoimmune lesions in the hypothalamus towards the end of an infectious episode. The hypothalamus regulates homeostatic control of sleep, hunger and sexual activity. In relation to this hypothesis, the erysipelatous humour described by Beauchêne might be seen as the initial infectious process, generating the autoimmune response which in turn provoked the patient’s pathology.

Beauchêne notes learning that the “patient had passed a worm, still alive, of the lumbricoid species”. Ancylostomiasis or hookworm disease is an intestinal parasitosis. Ascariasis, caused by Ascaris lumbricoides, occurs at all latitudes. While this condition triggers respiratory and digestive symptoms, no cutaneous or neuropsychological manifestations have ever been described.

In conclusion, Beauchêne offers the oldest observation currently identified which is consistent with KLS.

in 2000, Kesler et al reported nine female cases of the disease. Therefore, the fact that Beauchêne’s patient was a woman does not exclude the possibility of KLS.

Beauchêne offers an interpretation of his patient’s pathology, attributing her condition to contaminated humours in accordance with the precepts of Hippocratic medicine. He describes the initial cause as follows: “We proceeded from the idea that the recurring erysipelatous humour, described above, was the source of all subsequent disorders in the stomach, bowels, and even the head. It was our impression that this humour, whose existence we had no reason to doubt, had chiefly affected the digestive organs; it had tainted the gastric juices, then denatured the patient’s sense of taste, giving rise to the most bizarre appetites, to which she yielded by eating soil, lime, and plaster over a period of four years”.

The physiopathology of KLS remains unclear. A specific genetic predisposition (HLA DQB1*201) may favour the development of autoimmune lesions in the hypothalamus towards the end of an infectious episode. The hypothalamus regulates homeostatic control of sleep, hunger and sexual activity. In relation to this hypothesis, the erysipelatous humour described by Beauchêne might be seen as the initial infectious process, generating the autoimmune response which in turn provoked the patient’s pathology.

Beauchêne notes learning that the “patient had passed a worm, still alive, of the lumbricoid species”. Ancylostomiasis or hookworm disease is an intestinal parasitosis causing infected individuals to break out in a rash (could this explain the erysipelas described by Beauchêne?) followed by digestive pain associated with fever, but without accompanying megaphagia or other abnormal eating behaviours. Hookworm disease is currently considered a tropical parasitosis. Ascariasis, caused by Ascaris lumbricoides, occurs at all latitudes. While this condition triggers respiratory and digestive symptoms, no cutaneous or neuropsychological manifestations have ever been described.

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