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Les crises d'épilepsie diencépahlique
Excessive yawning and sleepy attacks

extrait du site http://www.baillement.com

Les crises d'épilepsie diencépahlique (épilepsie partielle), rares, se manifestent par des accès de bradychardie, variations brutales de la pression artérielle, du rythme respiratoire, du diamètre pupillaire, d'élévation de la température, de sueurs profuses, de salivation extrême, de troubles des postures, de troubles de la vigilance. Les crises gélastiques sont caractérisées par des accès de rire sans cause, (Pathological laughter and crying) et témoignent le plus souvent d'un hamartome de l'hypothalamus, parfois d'autres tumeurs suprasellaires, de séquelles de traumatismes craniens. Les causes de ces épilepsies sont : tumorale congénitale, des anomalies de la gyration et/ou de la neurostructure d'un noyau, une séquelle dégénérative localisée après virose herptique ou à CMV, ou à une neurosarcoïdose. L'EEG est souvent normal et l'IRM fonctionnelle peut-être la seule manière de visualiser la zone pathologique. Les variations de la circulation du LCR, des concentrations en neuromédiateurs Hypocrétine, GABA (déplétion post-ictale) commencent seulement à être étudiées.

Voici un cas exceptionnel, évoquant des crises épileptiques générées par l'hypothalamus et irradiant vers les noyaux thalamiques, sous forme d'une baisse de la vigilance avec salves irrépressibles de bâillements, évoluant par crises de 20 à 30 minutes jusqu'à dix fois par jour, se raréfiant sous antiépileptiques, sans cause tumorale retrouvée.

« I am a thirty two year old female. In my mid-twenties I began to develop an 'intolerance' to alcohol. Half a glass of wine would make me extremely and uncontrollably sleepy. It is only in retrospect that I realise this problem started years ago - at the time it just seemed normal to me.

Two years ago, I started getting these 'sleepy attacks' during the day, without alcohol. I got one every couple of days or so, and seemed to be related to meals or food. I still assumed it was some sort of allergic reaction, although I couldn't pinpoint it at all. Then these attacks became more frequent, about one a day. This was the pattern for about a month. Then suddenly, almost over night, I was getting up to eight or ten attacks a day, not related to meals specifically. They always follow the same pattern - no warning, then extreme drowsiness, accompanied by excessive yawning and a feeling of being out of it, for up to half an hour. Then I feel normal again. It happens while driving, eating, talking, walking, working etc. and is totally uncontrollable. This has been the pattern for the last two years. Some days are better than others are, but generally I have about four to six attacks a day. I do not actually fall asleep, unless close to normal sleep time.

Generally I sleep in excess of eight hours a day, and fall asleep in a normal length of time. My husband says that I am a fairly restful bed partner.

My father, who is a GP, watched my symptoms getting worse and started reading up a little. He sent me to Wentworth Hospital for tests. The neurologist suspected seizures and tried to get me to take some anti-seizure medication for a month, which I refused to do ‹ not wanting to be a guinea pig. He also sent me for an MRI and EEG and MSLT (without a polysomnogramme), all of which where normal.

Since I have had a consultation and EEG with Dr M G, Meulmed, Pretoria. She came recommended by a sleep disorders support group. She diagnosed me with pathological sleepiness and recommended ritalin(cf méthylphénidate = amphétamine). I was dissapointed with the consultation, because as she herself admitted, the term 'pathological sleepiness' is a dumping ground or term for unexplained sleepiness. I have used reactivan for a month or two, and it initially helped,

but I got to the point where I would have to up the dose to get the same effect. So I stopped using it rather. I did not try Ritalin.

When I experience an attack, it feels something like this:

I will be normally alert and engrossed in my work, social activity and will become aware of an impending attack sometimes by a feeling of wanting to yawn ‹ sometimes just a sense ‹ like a weight has been attached to my mind and is slowing me down. I also get irritable, sensitive to noise and light and other stimuli. The attack, with constitutes a feeling of diminshed awareness and extreme drowisiness, lasts about 20 minutes. I yawn uncontrollably and cannot surpress a yawn. The yawns are frequent and huge, with a sense of urgency which is relieved temporarily by the yawn. I can tell immediately almost to the second when it is over. The 'fog' seeems to lift in my mind. During the attack my body temperature seems to drop and it is often accompanied by goose bumps and cold. My eyes tear and nose runs a bit. I seldom fall asleep during an attack. I have never been a sleepy person during the day, and I fight the attacks like crazy. Its like being switched off against your will. I feel unwell during an attack, and they can be quite severe, although I never lose consciousness or awareness. Sometimes my whole day can be a write-off ‹ since the between periods are also foggy, with an impending sense of an attack all the time. I get short stabbing pains in the head sometimes during the attack and sometimes in isolation. I get sensations on my scalp, like a ticklish feeling on my forehead, which comes and goes intermittently.

I cannot control the attacks, although I can sometimes with extreme effort postpone it for half an hour or so. Chewing some gum can postpone an attack a while.

Driving has become a real problem, since I will almost certainly have an attack, even on short trips. I will be driving in an alert state, will experience an attack, and will resume driving in alert fashion once over. My work as an architect is severly affected, especially on a bad day.

I have had a 24 hour EEG at Entabeni Hospital under Dr H S, my current neurologist. He has been wonderful. His best attempts at diagnosis have been possibly epilepsy or a form of migraine aura. He had me on Epilim,(cf valproate sodique = dépakine) which had very good results. I am trying to conceive now, and am off the Epilim, which means that my symptoms have got worse again. They never go away entirely, with my best day having one attack, and my worst as many as fifteen, almost head to head, and lasting up to forty minutes.

I have had HLA typing for narcolepsy, which was negative.

I am average weight, physically active, insulin resistant, asthmatic and otherwise healthy. There is a family history of strokes. (My mothers father) A concern for me is that the Epilim, which controlled my symptoms so well, has recently been linked to a worsening in PCOS and insulin resistance, although this does not seem to be an accepted fact. »

Référence :

1. **Benarroch, E. E. (1993). "The central autonomic network: functional organization, dysfunction, and perspective." Mayo Clin Proc 68(10): 988-1001.** The central autonomic network (CAN) is an integral component of an internal regulation system through which the brain controls visceromotor, neuroendocrine, pain, and behavioral responses essential for survival. It includes the insular cortex, amygdala, hypothalamus, periaqueductal gray matter, parabrachial complex, nucleus of the tractus solitarius, and ventrolateral medulla. Inputs to the CAN are multiple, including viscerosensory inputs relayed on the nucleus of the tractus solitarius and humoral inputs relayed through the circumventricular organs. The CAN controls preganglionic sympathetic and parasympathetic, neuroendocrine, respiratory, and sphincter motoneurons. The CAN is characterized by reciprocal interconnections, parallel organization, state-dependent activity, and neurochemical complexity. The

insular cortex and amygdala mediate high-order autonomic control, and their involvement in seizures or stroke may produce severe cardiac arrhythmias and other autonomic manifestations. The paraventricular and other hypothalamic nuclei contain mixed neuronal populations that control specific subsets of preganglionic sympathetic and parasympathetic neurons. Hypothalamic autonomic disorders commonly produce hypothermia or hyperthermia. Hyperthermia and autonomic hyperactivity occur in patients with head trauma, hydrocephalus, neuroleptic malignant syndrome, and fatal familial insomnia. In the medulla, the nucleus of the tractus solitarius and ventrolateral medulla contain a network of respiratory, cardiovagal, and vasomotor neurons. Medullary autonomic disorders may cause orthostatic hypotension, paroxysmal hypertension, and sleep apnea. Neurologic catastrophes, such as subarachnoid hemorrhage, may produce cardiac arrhythmias, myocardial injury, hypertension, and pulmonary edema. Multiple system atrophy affects preganglionic autonomic, respiratory, and neuroendocrine outputs. The CAN may be critically involved in panic disorders, essential hypertension, obesity, and other medical conditions.

2. **John S. Imaging and epilepsy Duncan Brain (1997), 120, 339–377**
3. **J. Parvizi, Pathological laughter and crying: a link to the cerebellum Brain 2001, 124 (Pt 9): 1708-19**
4. **Khadilkar, S., K. Menezes, et al. (2001). "Gelastic epilepsy--a case report with SPECT studies." J Assoc Physicians India 49: 581-3.** A 24 years male presented with daily episodes of uncontrollable laughter followed by urinary incontinence since the age of nine years. Some of these attacks progressed to generalized tonic-clonic seizures. General and neurological examination did not reveal any abnormality. Ictal and interictal video EEGs were normal. MRI showed a hypothalamic hamartoma. Interictal SPECT scan showed normal perfusion in the hamartoma. SPECT scan obtained four minutes after beginning of seizure showed that the perfusion increased in right cingulate gyrus but not in the hamartoma, suggesting the involvement of the cingulate gyrus in the seizure origin or pathway.
5. **Akman, C. I., R. Schubert, et al. (2002). "Gelastic seizure with tectal tumor, lobar holoprosencephaly, and subependymal nodules: clinical report." J Child Neurol 17(2): 152-4.** Gelastic seizures are characterized by inappropriate, stereotyped laughter and are often first recognized when other epileptic manifestations occur. They are frequently associated with hypothalamic hamartomas. Central nervous system developmental abnormalities are rarely reported with gelastic seizures. There is only one case report of gelastic seizure caused by holoprosencephaly. We report a 2-year-old girl with multiple brain structural abnormalities including tectal tumor (possibly hamartoma), multiple subependymal nodules, and holoprosencephaly. She developed seizures during the newborn period and presented with gelastic seizure and simple partial seizure at 3 months of age.
6. **Sturm, J. W., F. Andermann, et al. (2000). ""Pressure to laugh": an unusual epileptic symptom associated with small hypothalamic hamartomas." Neurology 54(4): 971-3**. Gelastic seizures are the hallmark of the epilepsy syndrome associated with hypothalamic hamartomas. Patients typically develop cognitive deterioration and refractory seizures. The authors describe three patients with small hypothalamic hamartomas without these features and thus identify a mild end to the clinical spectrum. All had the unusual symptom of "pressure to

laugh," often without actual laughter. This symptom could be dismissed as psychogenic but should be recognized as a clue to the presence of this unusual lesion.

7. **Boeve, B. F., E. F. Wijdicks, et al. (1998). "Paroxysmal sympathetic storms ("diencephalic seizures") after severe diffuse axonal head injury." Mayo Clin Proc 73(2): 148-52.** We describe a patient with a severe traumatic head injury who exhibited paroxysmal sympathetic storms, similar to those described in "diencephalic seizures." No epileptiform activity was evident on electroencephalography, and therapeutic levels of anticonvulsants failed to alter the spells; however, use of morphine sulfate abolished them. The features of this and several previously reported cases refute the primary roles of the diencephalon and seizures in this syndrome. Rather, in the setting of already compromised autonomic neuronal integrity, subtle fluctuations in intraventricular pressure or activation of reflexes triggered from muscle mechanoreceptors or chemoreceptors during episodes of hypertonia are more likely. "Paroxysmal sympathetic storms," a more appropriate descriptive term for these phenomena, should be recognized; thus, unnecessary diagnostic evaluations can be minimized, and appropriate therapy can be initiated.
8. **Morris, H. H., 3rd (1988). "Sudden diencephalic events." Ann Neurol 23(2): 208.**