**KLUVER BUCY SYNDROME IN WOMEN AND MEN-AUG 2016 FROM INTERNET**

**Kluver-Bucy syndrome -- an experience with six cases.**

[Jha S](http://www.ncbi.nlm.nih.gov/pubmed/?term=Jha%20S%5BAuthor%5D&cauthor=true&cauthor_uid=15472430)1, [Patel R](http://www.ncbi.nlm.nih.gov/pubmed/?term=Patel%20R%5BAuthor%5D&cauthor=true&cauthor_uid=15472430).

[**Author information**](http://www.ncbi.nlm.nih.gov/pubmed/15472430)

**Abstract**

The Kluver-Bucy syndrome (KBS) is a neurobehavioral syndrome and can be seen in association with a variety of neurological disorders. Case records of 6 patients with KBS seen during a period of 5 years in a university hospital were reviewed. During the study period 6 patients with KBS, aged between 4 and 14 years, were seen. Hyperorality, hypersexuality, and abnormal behavior were the most common manifestations. Of the 6 patients, 5 had recurrent unprovoked seizures. The associated neurological disorders included anoxia-ischemic encephalopthy (2), herpes simplex encephalitis (1), neurocysticercosis (NCC) (1), traumatic brain injury with gliosis (1 case) and tuberculous meningitis (1 case). Prognosis was poor in all the patients except in the patient with NCC.

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Eastern Journal of Medicine 16 (2011)215-217 M. Okur et al / Kluver-Bucy syndrome and convulsion

Case Report

 215

Kluver-Bucy syndrome developed after convulsion: A case report

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Abstract. Kluver-Bucy syndrome is characterized by increased appetite, hypersexuality, hypermetamorphosis, memory disorders, visual agnosia, stagnancy, aphasia, bulimia, polyuria, and polydipsia. A 14 year old girl had generalized tonic-clonic convulsions at admission, and an incomplete Kluver-Bucy syndrome with hypersexuality, recent memory disturbance, hypermetamorphosis, speech disturbance, hyperactivity, agitation, aggressiveness, and hallucinations, developed the following day. Here in, we report a case of KBS in a child with epilepsy.

Key words: Child, hypersexuality, hallucinations, agitation, epilepsy 1. Introduction Kluver-Bucy syndrome (KBS) is characterized by increased appetite, hypersexuality, hypermetamorphosis, memory disorders, visual agnosia, stagnancy, aphasia, bulimia, polyuria, and polydipsia (1). Kluver and Bucy was first described KBS, in 1939, as a neurobehavioral syndrome in rhesus monkeys in whom rhinencephalon and bilateral lobes were rejected (2). Terzian reported a case of KBS after bilateral temporal lobectomy as the first human case (3). The potential causes of KBS include Herpes simplex encephalitis, head trauma, subarachnoid hemorrhage, epilepsy, bilaterally thalamic infarct, bilaterally temporal lobe resection, glioblastoma, shigellosis, Pick disease, adrenoleukodystrophy, hypoglycemia, Reye syndrome, Alzheimer disease, porphyrinuria, anoxic-ischemic encephalopathy, neurocystocercosis, tuberculosis meningitis, arachnoid cyst, Huntington chorea, Parkinson disease, Systemic lupus erythematosis, carbon monoxide intoxication, leukoencephalopathy associated with methotrexate, radiation, and stroke (4). \*Correspondence: Dr. Avni Kaya Department of Pediatrics, Women and Children's Hospital, Van, Turkey E-mail: avnikaya@gmail.com. Received: 23.03.2010 Accepted: 03.11.2010 In this report, we describe a case of KBS in a 14 year old girl with epilepsy who presented with incomplete Kluver-Bucy syndrome with hypersexuality, recent memory disturbance, hypermetamorphosis, increased appetite, speech disturbance, hyperactivity, agitation, aggressiveness, and hallucinations. 2. Case reports A 14 year old girl with epilepsy was admitted to our emergency department. She had periodic convulsions once every 15 days in the last year. On the day before admission, she had had at least three generalized tonic clonic convulsions and each had continued for 15 minutes. She had no fever, headache, vomiting, personality change, head trauma or intoxication in her history. She had stopped taking her antiepileptic drugs (valproic acid and carbamazepine) without any reason two weeks before admission. There were no pathological features in her family history. On physical examination, the patient’s general condition was moderate and vital signs were normal. On neurological examination, she was lethargic, the pupils were myotic, pupil reflexes to penlight were positive bilaterally, deep tendon reflexes were normal bilaterally, and plantar response was flexor bilaterally. There were no signs of meningeal irritation. Laboratory tests revealed the following: hemoglobin 12,8 g/dL, hemotocrit 36%, white blood cell 18230/mm3 and platelet count was 348000/mm3. Cerebrospinal fluid protein was 14

 Eastern Journal of Medicine M. Okur et al / Kluver-Bucy syndrome and convulsion

216

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Wong at al. reported a case of KBS that occurred after encephalitis with poliuria, polidipsia, narcolepsy, personality changes, amnesia, and hypersexuality (11). Yilmaz et al. has reported a child with incomplete KBS developed during acute encephalitis (12). Neuroleptics, carbamazepine, dopamine blockers, benzodiazepines, and fluvoxamine can be used in treatment of KBS (2,3-13). Our patient was started on haloperidol therapy for behavioral changes, and valproic acid and carbamazepine were continued. A month after discharge, the patient recovered completely and convulsions did not recur. Haloperidol therapy was stopped and the anticonvulsive therapy was continued without any change. In conclusion, we would like to emphasize that patients with epilepsy should be closely followed for KBS especially in patients with hypersexuality, hallucinations, hypermetamorphosis, increased appetite, memory disturbance, speech disturbance, hyperactivity, agitation, and aggressiveness. Overall the prognosis of the syndrome depends largely on the underlying pathology.

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217

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Eastern Journal of Medicine 16 (2011)215-217 M. Okur et al / Kluver-Bucy syndrome and convulsion

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 215

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 Eastern Journal of Medicine M. Okur et al / Kluver-Bucy syndrome and convulsion

216

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217

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# [The Strange Lives of Women With No Fear](http://jeffwise.net/2010/12/21/the-strange-lives-of-women-with-no-fear/)

– DECEMBER 21, 2010**POSTED IN:**[PSYCHOLOGY](http://jeffwise.net/category/psychology/)

For a woman with profound brain damage, SM seems rather unremarkable. Her IQ tests normal; she speaks like an average person, and her memory and perception show no sign of dysfunction. But the 44-year-old woman does have one very specific, very unusual, and for neuroscientists, a very interesting impairment: she has no amygdala, the part of the brain that’s the central switching box for analyzing external threats. SM has no fear.

SM’s story received a great deal of attention lately thanks to a paper describing her condition that was published in the journal [Current Biology](http://www.cell.com/current-biology/abstract/S0960-9822%2810%2901508-3). ([Neurophilosopy](http://scienceblogs.com/neurophilosophy/2010/12/the_woman_who_knows_no_fear.php?utm_source=feedburner&utm_medium=feed&utm_campaign=Feed%3A+ScienceblogsChannelBrain+%28ScienceBlogs+Channel+%3A+Brain+%26+Behavior%29" \t "_blank) did a particularly incisive and digestible rundown of the paper’s findings.) The authors introduced SM, whose amygdalae were destroyed by a genetic condition called Urbach-Wiethe disease, to a variety of situations that a normal person might well find fear-inducing. They took her to an exotic animal shop where she handled snakes and looked at tarantulas; they took her to a “haunted house” attraction; showed her clips of movies like “The Blair Witch Project”; and told her that Sarah Palin had been appointed to the Supreme Court. (OK, not the last one). In each case, she showed no signs of fear, and reported feeling no anxiety. In fact, while scampering through the haunted house she was so delighted and curious that she scared one of the “monsters” by trying to poke its mask.

For most of us, fear seems like a negative emotion, one that stresses us out and inhibits us from trying things that might make our life more rewarding. But as the Current Biology paper makes clear, SM’s fearlessness has cost her a great deal. On the most obvious level, it has left her vulnerable to all kinds of dangers. She lives in a dangerous part of a big city, and several times she has walked obliviously into potentially violent encounters. One time, she was held up at gunpoint; another time, a drug addict accosted her and held a knife to her throat. Intriguingly, though she did not feel scared during those encounters, she did report feeling angry and upset afterward. Her emotional deficit is quite specific.

But in a sense SM’s fearlessness is not the worst part of losing her amygdalae. Because, as it turns out, the amygdala does much more than regulate the fear response. Monkeys who have that part of the brain removed exhibit what’s known as Kluver-Bucy syndrome, a condition that’s characterized not only by fearlessness but also by docility, hypersexuality, and a tendency to explore the world by touching things with the mouth.

These monkeys share with similarly afflicted humans a striking lack of self-control. In SM’s case, one of the most pronounced effects of her brain damage was a lack of social restraint. She often behaved inappropriately, acting excessively friendly and making crude sexual remarks.

Recently, Psychology Today blogger [Kelly McGonigal](http://www.psychologytoday.com/blog/the-science-willpower/201012/why-we-need-little-fear) wrote about another woman who also lost the functioning of her amygdala. The 24-year-old, who McGonigal calls Lucy, is an epileptic who suffered such severe temporal-lobe seizures that her doctors decided to remove part of her brain, including one of her two amygdalae. In the wake of the operation, her seizures were dramatically reduced. But then, writes McGonigal:

*Five years later, Lucy showed up in the emergency room again. She has suffered another generalized seizure. The attending physicians described her as “lethargic and unresponsive, but medically stable.” That’s when things get weird. According to the original case report in the Journal of Neurology, Neurosurgery, and Psychiatry, “She was left unattended in an examination room. About 30 minutes later, she was found in an adjacent room performing fellatio on an elderly male cardiac patient.”*

*It turns out Lucy had continued to have seizures after the surgery, usually when she forgot to take her medication. For one to two hours after each seizure, she would engage in a wide variety of unusual sexual behaviors, such as masturbating in public and trying to seduce family members and neighbors. She also lost control around food in the post-seizure period. Her family reported extreme binge eating episodes that disappeared between seizures.*

What had happened was that the recurring seizures effectively turned off her sole remaining amygdala, leaving her with no function in that region at all.

Together, the cases of SM and Lucy show that the amygdala, while indeed being a crucial juncti0n in the brain’s fear circuitry, also plays a surprisingly crucial role in an even more important brain function: self-control. This has traditionally been seen as falling under the purview of the brain’s “higher” cortical regions, especially the prefrontal cortex. And indeed, patients with prefrontal lesions often show inappropriate behavior and hypersexuality just like SM and Lucy. But what’s clear from studying these amygdala-less women is that the “lower” parts of the brain — specifically, the limbic region that generates our emotional responses — is just as key. When it comes to keeping ourselves in check, it seems, emotion is as powerful as thought.

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## **What should we think of people whose addled brains are driving them to nymphomania?**

*By*[*Jesse Bering*](http://www.slate.com/authors.jesse_bering.html)

What if Hank Moody had Klüver-Bucy Syndrome?

If you are a materialist holding the logical belief that the human brain, with all of its buzzing neural intricacies, its pulpy, electrified, arabesque chambers and labyrinthine coves, has been carved out over countless eons by the slow-and-steady hand of natural selection, then you will grant that specific brain regions evolved because they generated behaviors that were beneficial to our ancestors. When one part of the brain is compromised—through injury, disease, or some other unfortunate event—the constellation of symptoms that result are often remarkably specific. "The brain is the physical manifestation of the personality and sense of self,"[**writes**](http://onlinelibrary.wiley.com/doi/10.1002/bsl.857/abstract) University of Michigan neuroscientist Shelley Batts in a 2009 issue of Behavioral Sciences and the Law, "and focal damage to brain areas can result in focal changes in behavior and personality while leaving other aspects of the self unchanged."

Not to get too technical, but if you're unlucky enough to develop a lesion that interferes with the functioning of your dorsolateral prefrontal cortex, a specialized patch of neural tissue that's intricately braided into your anterior cingulate cortex, then your working memory, strategy-formation, and planning skills are going to take a major nosedive. Something as simple as coming up with a list of groceries that you'll need for the next few days becomes a major achievement.

Most of us—materialist and dualist alike—have sympathy aplenty for those patients whose brain disturbances have interfered with their everyday cognitive abilities. We're perfectly willing to accommodate their intellectual disabilities by, say, helping them create a new mnemonic strategy or giving them a pat on the back or a word of encouragement when they're trying to remember someone's name. Yet when chunks of gray matter that have evolved to control and inhibit, say, our sexual appetites and other Bacchanalian drives experience a similar catastrophic blowout, are we so understanding? What if those impairments lead their victims to display … oh, I don't know, let's call them moral disabilities? Cases of libidinal brain systems going haywire have our kind-hearted, humanistic materialism rubbing elbows—or butting heads—with our belief in free will and moral culpability.

Although **[Klüver-Bucy Syndrome](http://www.ninds.nih.gov/disorders/kluver_bucy/kluver_bucy.htm%22%20%5Ct%20%22_blank)**  is relatively rare, it's one of the most notorious neurological causes of a complete breakdown in one's ability to control sexual urges. In 1939, neuroanatomists Heinrich Klüver and Paul Bucy removed the greater portions of both temporal lobes and the rhinencephalon from the brains of rhesus monkeys. Among a host of other peculiar effects of this rather cruel vivisection, the monkeys became incredibly randy, displaying a prominent and indiscriminate desire to copulate. The first documented case of full-blown Klüver-Bucy in humans arrived in 1955, when an epilepsy patient underwent a bilateral temporal lobectomy (a surgical excision of the lobes) and subsequently developed a ravenous sexual appetite, among other things. More often, the syndrome appears in lesser degrees, precipitated by a nasty insult to the medial temporal lobe. That might result from a case of [**herpes encephalitis**](http://en.wikipedia.org/wiki/Herpesviral_encephalitis) or [**Pick's disease**](http://www.ncbi.nlm.nih.gov/pubmedhealth/PMH0001752), or from trauma and oxygen deprivation. Not all such patients experience hypersexuality, mind you, but some do. Other symptoms aren't terribly appealing, either, however; they include hyperorality (a compulsive desire to put things in one's mouth), apathy, emotional unresponsiveness, and various sensory disorders.



Dramatic case studies illustrating the devastating effects of Klüver-Bucy Syndrome abound in the clinical literature, and they raise intriguing philosophical questions for us to consider. That some patients so stricken are overcome with excessive [**carnal urges**](http://www.scientificamerican.com/blog/post.cfm?id=my-lust-2011-02-14) and are not simply using the disorder as a convenient excuse to become freely promiscuous, lewd, and lascivious is perhaps best demonstrated by a 1998 Clinical Neurology and Neurosurgery[**study**](http://www.ncbi.nlm.nih.gov/pubmed/9879850) by Indian neurologist Sunil Pradhan and his colleagues. In this report, a group of boys between the ages of 2.5 and 6 began to exhibit hypersexualized behaviors after partially recovering from comas induced by herpes encephalitis. One to three months after emerging from the comatose state, "all seven children," note the authors, "demonstrated abnormal sexual behavior in the form of rhythmic hip movements (two patients), rubbing genitals over the bed (two patients) and excessive manipulation of genitals (all seven patients)." Were these children just helpless, hapless puppets of their ancient, pleasure-driven brains? The authors believe so. "As all patients [at the time of study], except one, were 4 years of age, with no possibility of environmental learning of sex, these movements most probably represented phylogenetically primitive reflex activities."

It may be awkward enough telling other parents why your preschooler is humping everything in sight—just try rehashing the foregoing description of Klüver-Bucy Syndrome to your friends at the day care—but we do tend, as adults, to be mostly forgiving of a child's improprieties. When this sort of hypersexuality strikes a post-pubescent individual whose sexuality is driven by [**orgasm-propelled**](http://www.scientificamerican.com/article.cfm?id=reopening-the-case-of-the-female-or-2009-12-01) desires, things become more interesting—at least, in a philosophical sense. Although it would be entirely inaccurate to portray Klüver-Bucy patients as sex-crazed lunatics, they very often display behaviors that would be considered inappropriate by conventional standards. One gentleman in his early 70s, for instance, hugged a female parishioner at his church and repeatedly kissed her. According to the [**clinical case report**](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2868923/?tool=pubmed), he then asked the shocked woman, "Why don't we do it again?" Over the ensuing years, his sexual fantasies skyrocketed and his hyperorality became unmanageable. The report notes that, according to his wife, "he would put any object in his mouth, including dog food, candles, adhesive bandages, and his wedding ring. His appetite seemed insatiable. … He died at age 77 years of asphyxiation on several adhesive bandages."

In a 2005 [**letter to the editor**](http://www.ncbi.nlm.nih.gov/pubmed/16018931) of European Psychiatry, two physicians describe the case of a 14-year-old schoolgirl ("Ms. A"), who, prior to developing Klüver-Bucy Syndrome after being in an encephalitis-caused coma, "was an intelligent and social girl with a good academic record." This quiet, well-behaved teenager became somewhat challenging, to say the least, after recovering from her illness. You think you're raising a difficult teen? Consider what these parents were dealing with:

[T]he patient started … disrobing in front of others, manipulating her genitals, and making sexual advances toward her father. She would lick any object lying on the ground and whenever she got an opportunity, she would rush to the toilet and try to put urine and feces into her mouth (urophagia and coprophagia, respectively).

In another case, an epileptic woman underwent an unsuccessful left temporal lobectomy to help stop debilitating seizures. Klüver-Bucy symptoms, including hypersexuality, emerged following the surgery. She began [**masturbating**](http://www.scientificamerican.com/article.cfm?id=one-reason-why-humans-are-special-a-2010-06-22) in public and aggressively soliciting her family members and neighbors for sex. After having another seizure, she was brought to the emergency room, where, after a half hour in the waiting area, she began [**performing fellatio on an elderly cardiac patient**](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC1014870/pdf/jnnpsyc00476-0083.pdf?tool=pmcentrez). (This may or may not be one of the few examples where one person's syndrome is another's lucky day; it's also unclear if this was a display of hypersexuality or hyperorality, but it's inevitable, perhaps, that the twain should occasionally meet.)

Other temporal-lobe epileptics have also exhibited hypersexuality in the "postictal" state, which is the period of recovery time following a seizure. In a recent issue ofEpilepsy and Behavior, New York University neurologist Vanessa Arnedo and her colleagues [**present the case**](http://www.ncbi.nlm.nih.gov/pubmed/19793682) of a 39-year-old man who began having semi-frequent seizures during the middle of the night. After nocturnal convulsions, he'd sleep for another 10 minutes, wake up, and then rape his wife. (In the authors' more delicate wording, he was described as "becoming sexually aggressive toward his wife by forcing intercourse.") Importantly, however, "the tremendous remorse and abhorrence for what he had done when he learned of his actions led him to pursue possible surgery mainly to eliminate this postictal behavior." Other people with similar epileptic profiles also become hypersexualized in the postictal state. To his later horror, one man motioned for his 12-year-old daughter to [**join him and his wife**](http://www.ncbi.nlm.nih.gov/pubmed/5411364) in the bedroom following a nighttime seizure.

It is these last few examples, where Klüver-Bucy Syndrome manifests in criminal behavior, such as [**rape**](http://www.slate.com/id/2281138/) or child molestation, that our materialistic convictions are really put to the test. In 2003, University of Virginia neurologists Jeffrey Burns and Russell Swerdlow described how an otherwise well-behaved, 40-year-old man developed a case of "new-onset pedophilia" after suffering the appearance of a right orbitofrontal tumor. The man denied any pre-existing interest in children; he did have a predilection for pornography before the tumor, say Burns and Swerdlow, but now he was downloading child porn and making subtle sexual advances to his prepubescent stepdaughter. His hypersexuality applied to full-grown women, too—so much so, in fact, that he couldn't keep himself from fondling female nurses and staff during a neurologic examination. Long story short, when the man's tumor was removed, his prurient interests and behaviors all but disappeared, and since he was no longer deemed a threat to his stepdaughter, he returned home. But his headaches returned, his tumor regrew, and so did the criminal impulse. A "re-resection" of the tumor was accomplished, the man became a good citizen again, and, as far as we know, that remains true today. (In a [**more recent case**](http://www.informaworld.com/smpp/content~content%3Da916970996~db%3Dall~jumptype%3Drss) co-published by famed neuroscientist [**Oliver Sacks**](http://www.oliversacks.com/), and neatly summarized by the **[Neuroskeptic](http://neuroskeptic.blogspot.com/2009/11/brain-damage-pedophilia-and-law.html%22%20%5Ct%20%22_blank)** blog, a 51-year-old man without any criminal history had a portion of his right temporal lobe removed to prevent seizures. Following this, he developed telltale signs of Klüver-Bucy, including hypersexuality. His was another case of "new-onset [**pedophilia**](http://www.scientificamerican.com/article.cfm?id=pedophiles-erotic-age-orientation)" but as Sacks laments, in spite of this he was nevertheless sentenced to several years in prison for downloading child porn.)

What's the take-away message? I'll let you do the hard work of thinking through the implications for our belief in free will and how it might or might not apply. But another intriguing question emerges, too: If a "good" person's brain can be rendered morally disabled by an invasive tumor or an epileptic fuse-shortage, subsequently causing them to do very bad deeds, then isn't it rather hypocritical to assume that a "bad" person without brain injury—whose brain is anatomically organized by [**epigenetics**](http://www.nature.com/nature/supplements/insights/epigenetics/index.html) (the complex interplay between genes and experiences)—has any more free will than the neuroclinical case? After all, perhaps it's just a matter of timing: The "good" are born with brains that can "go bad," whereas the "bad" are hogtied by a morally disabled neural architecture from the very start. And although it may be less common, if a "bad" person behaves in an upstanding manner, could that be the result of fortuitous brain damage or epilepsy, too?

It's all brain-based in the end, including the parameters by which one can contemplate and, especially, execute their free will. Perhaps we're only as free as our genes are pliable in the slosh of our developmental milieus.

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