

Kluver-Bucy Syndrome in Children

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Kluver-Bucy syndrome is an uncommon syndrome of behavioral abnormalities following bilateral temporal lobe injury. Only four children have been reported previously with this syndrome. We report three additional pediatric patients who developed Kluver-Bucy syndrome following hypoxic insults. In two patients, features of the syndrome were transient. Problems in intermediate memory were present in each patient. Behavioral abnormalities did not respond to the medications administered. Our experience suggests that Kluver-Bucy syndrome may occur more commonly in children than was suspected previously, especially following hypoxia.

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Introduction

Kluver-Bucy syndrome (KBS) is an unusual group of behavioral abnormalities which was described initially in 1938 following bilateral anterior temporal lobectomy in monkeys [1]. Incomplete forms of KBS have been observed subsequently in humans following bilateral temporal lobe damage. Although there are over 100 case reports of adults [2-15], there are only 4 reports of children with KBS [16-19].

The abnormalities described in monkeys include:

- (1) "Psychic blindness" or visual agnosia (i.e., the monkeys could see but did not appear to be able to rely on visual characteristics alone to identify objects);
- (2) Compulsion to examine objects orally;
- (3) Repetitive movements;
- (4) Hypermetamorphosis or irresistible impulse to notice and react to everything within sight;
- (5) Placidity;
- (6) Increased appetite; and,
- (7) Increased sexual activity with animals of either sex.

Memory also may have been affected because Kluver and Bucy observed that the monkeys had difficulty learning form-board tasks.

We believe that KBS, accompanied by memory defects, occurs in children more frequently than suspected previously, especially following hypoxic insults. The behavioral abnormalities do not respond to medication; however, spontaneous improvement or recovery generally occurs.

Case Reports

Patient 1 is a male, 4 years and 11 months of age. At 26 months of age he suffered an anoxic episode following a prolonged seizure with fever. He was admitted to a local hospital and treated with phenobarbital. No source of the fever was found and he was discharged. Two weeks later he was admitted to Wyler Children's Hospital because his parents became concerned about a persistent change in his mental and physical state. On examination the child was alert and rather placid but easily distractible by any stimulus within the room. His vision was intact, but he appeared more attentive to auditory than to visual stimuli. He had difficulty identifying body parts or pictures of animals. When offered toys, he mouthed or bit them consistently before playing with them. He also frequently attempted to stroke the breasts and genitals of the examiners and staff. Pupils were equal, reactive, and responsive to light. Fundi were normal and visual fields were intact. His neck was supple. Cranial nerve, motor, and deep tendon reflex examinations were normal. He had a prominent rooting reflex. The gait was somewhat wide-based and unsteady. Laboratory examinations included a lumbar puncture and electroencephalogram (EEG) which were normal. Convalescent viral titers were unremarkable. Visual evoked response demonstrated intact primary visual pathways. Computed tomography (CT) revealed prominent cortical sulci and mildly enlarged lateral ventricles but otherwise was within normal limits.

The child was discharged and returned two months later for examination. At that time he was alert and responsive, but easily distractible. He had good visual fixation and following, but he still tended to place small objects which were offered to him in his mouth. The previously observed sexual behavior had ceased. Subsequent examinations failed to reveal any visual agnosia or propensity to mouth objects.

Psychometric testing at 4 years and 11 months of age indicated an IQ equivalent of 72 on the Stanford-Binet Intelligence Scale. His performance on individual items showed that he had better developed verbal than visual-motor abilities. Short attention span also was observed. Immediate auditory sequential memory as tested by digit span was normal (i.e., the ability to repeat 4 digits in a forward order); however, he could not remember three unrelated object names after three minutes, a task which most normal 3-year-olds can perform successfully. On the comprehension section of the Northwestern Syntax Screening Test he scored at the 4-year-old level, although he had difficulty comprehending reversible active and passive sentences for which the order of the words was important. This difficulty may be secondary to a problem in intermediate auditory memory.

Patient 2 is a 14-year-old female with a seizure disorder who was well until 10 years of age when she had a generalized seizure in a swimming pool and was found face down, cyanotic, and without pulse. She was taken to a local hospital where she was intubated and resuscitated. After discharge, her parents complained that she had a voracious appetite, that she masturbated frequently in public, and that she had recurrent generalized seizures.

At 14 years of age the patient was referred to Wyler Children's Hospital for seizure disorder evaluation. On examination she was an

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obese white female. She was alert, but easily distractible, reacting to any auditory or visual stimuli in the room. She tended to grab the examiner's hand and kiss it. When presented with objects to identify, she immediately brought them to her mouth as if her ability to identify objects depended on their oral characteristics. Pupillary and fundoscopic examinations were normal. Visual fields and cranial nerves were intact. Motor examination, gait, and coordination were normal. Laboratory studies included an EEG which demonstrated epileptiform discharges in the right temporal area. CT depicted prominent cortical sulci and bilateral temporal lobe atrophy. The patient was gradually withdrawn from phenytoin treatment and begun on carbamazepine therapy. Seizure control improved but the behavior remained unchanged. The patient did not appear for subsequent evaluation.

No formal intellectual testing was performed. She was able to repeat 5 digits forward and 3 backward and could remember her address and her birthday but could not recall three objects after 5 minutes. She was able to draw a picture of a man at the 5-year-old level and was able to read a first grade reader, although she had no informational recall.

Patient 3 is an 11½-year-old male with asthma. At 6½ years of age he was transferred to Wyler Children's Hospital because of increasing respiratory distress and carbon dioxide retention. His course was complicated by prolonged episodes of profound acidosis and hypoxia. On the fourth hospital day he failed to demonstrate any movement between doses of pancuronium. A few hours after pancuronium was discontinued he had a sudden episode of increased tone followed by tonic-clonic movements of all extremities lasting for several minutes. Neurologic examination revealed dilated asymmetric pupils, the right greater than the left, limited abduction of the right eye, downward bobbing of the eyes, and absent corneal reflexes bilaterally. There was flaccid paralysis of all extremities; deep tendon reflexes were very brisk and symmetric and plantar responses were extensor bilaterally. The following day the patient's respiratory status improved and he was extubated. During the first 2 weeks he was extremely restless, constantly walking about the halls. He had a flat affect and rarely showed any emotion, except for sudden episodes of rage-like screams associated with hyperventilation and flexion of his extremities. He was able to see, but he appeared to have trouble identifying objects or places. On several occasions he was found wandering in the halls, unable to locate his room. On one occasion when his mother visited him, he did not recognize her. At one point he asked to see his mother, even though she was sitting in the room with him. He frequently mouthed his bed rail and other inedible objects. His speech was disorganized with much perseveration and echolalia. An EEG prior to discharge demonstrated diffuse slowing in the 6-7 Hz range with no epileptiform discharges. Ophthalmology examination revealed an acuity of 20/25 bilaterally.

Over the next 3 years he continued to have behavioral problems. He was expelled from school because he engaged in sexual play with another boy. He also made sexual advances to adults of both sexes. No problems with vision or mouthing of inedible objects were observed.

He was readmitted to La Rabida Hospital at 9½ years of age for asthma and to evaluate whether some of his sudden changes in behavior represented seizures. On admission he appeared undernourished and apathetic. He ate voraciously sometimes and hoarded food from other patients' trays, but after eating complained of hunger.

Two EEGs and a CT were normal. Prior to discharge he had a generalized tonic-clonic seizure and was placed on phenytoin. On neurologic examination he was unusually placid and seemed apathetic; however, towards the end of the examination, during the Babinski maneuver, he became hysterical suddenly. As quickly as this outburst began, it ceased and he was placid again.

On mental status examination he was able to write his name and to spell several words. He read a third grade reader perfectly but was not able to answer any questions about what he had read. He was able im-

Table 1. Features of Kluver-Bucy syndrome

Syndrome Features	Patients		
	1	2	3
Visual agnosia	Yes	Yes	Yes
Oral examination of objects	Yes	Yes	Yes
Repetitive movements	No	No	No
Hypermetamorphosis	Yes	Yes	Yes
Placidity	Yes	No	Yes
Increased appetite	No	Yes	No
Altered sexual activity	Yes	Yes	Yes
Impaired intermediate memory	Yes	Yes	Yes

mediately to repeat six digits forward and four digits backward, but he could not recall any of three objects that he had been asked to remember after 5 minutes. He was able to perform simple addition and subtraction but his drawing of a person was extremely simplistic and more appropriate for a 5- or 6-year-old. Neurologic examination otherwise was normal. EEG demonstrated left anterior temporal epileptiform discharges. Over the next 18 months his behavior improved slowly. His preoccupation with sexuality became less prominent and he has made some progress in a class for the severely learning disabled.

On the Wechsler Intelligence Scale for Children-Revised (WISC-R) administered at 6 years and 10 months of age, the patient achieved a verbal score of 79, a performance score of 78, and a full scale IQ equivalent of 77. The patient received subsequent psychometric testing at 9½ years of age. At this time his WISC-R verbal score was 69 and his performance score was 70, yielding a full scale of 68. On individual subtests of the WISC-R he received age-equivalent scores ranging from 6 years and 2 months to 7 years and 10 months. On the Wide Range Achievement Test he received grade equivalents of 2.9 on the arithmetic subtest, 4.1 on the spelling subtest, and 4.0 on the reading subtest which tests the ability to read single words. On the Gilmore Oral Reading Test, which involves paragraph reading, he received a grade score equivalent of 3.5, but his comprehension was only at the 1.0 grade level. Difficulty encoding or retrieving new information was observed: the patient was unable to recall three objects that he had been asked to remember after 5 minutes; after several sessions he was unable to recall the examiner's name or to remember how to return to his hospital room.

Discussion

Complete KBS rarely has been observed in humans [15]. The symptoms of KBS in humans are somewhat different than in animals. Psychic blindness in humans includes the inability to recognize significant persons (i.e., discriminate family members from strangers). Hypermetamorphosis consists of frequent exploration of the environment and extreme distractibility. Changes in sexuality often are more restricted to verbal behaviors. Hyperorality often takes the form of bulimia

or eating inedible objects. Because isolated features of the syndrome occur commonly with lesions outside the temporal lobes, three or more characteristics are essential to establish the diagnosis; visual agnosia and hypermetamorphosis are essential features [6]. Most patients have had additional problems, including aphasias, dementia, amnesia, and seizures.

Our three patients had many, but not all, of the associated KBS features (Table 1), a finding similar to other reported patients. They all lacked repetitive movements; however, each patient's behavior was characterized by an inability to identify objects, places, or people, a compulsion to examine objects orally, extreme distractibility, and by increased and inappropriate sexual behavior. In two patients some of the behavioral abnormalities persisted for only 2 weeks to 3 months, but in both patients the unusual behavior was documented by multiple observers, including two neurologists. Although each patient demonstrated a tendency to examine inedible objects orally, only Patient 2 exhibited sufficient change in appetite to produce obesity. Placidity or apathy was observed in two patients. In Patient 3, placidity alternated with sudden and brief outbursts provoked by trivial stimuli — so-called "sham rage." This finding is in contrast to the inability to induce fear or rage reactions in monkeys following temporal lobectomy. Such swings in behavior have been observed in some adult patients with KBS [18].

The significance of KBS in monkeys is the localization of certain behavior to specific areas of the temporal lobe. The oral tendencies, hypersexuality, and placidity can be induced by lesions confined to the amygdala [20], whereas impairment of visual discrimination and learning can be produced by lesions of white matter tracts of the temporal lobe and hippocampus [21].

Although extensive lesions of the temporal lobes underly the symptoms of this complex in animals, the precise correlation between behavioral abnormalities and anatomical lesions in humans is not clear. Patients with KBS, including the present ones, all suffered from disease processes that had generalized effects on the central nervous system, in addition to localized effects on the temporal lobes. Pathologic studies of KBS in humans have demonstrated involvement of the amygdala as well as more generalized cortical lesions in all patients. In contrast to animals, involvement of the temporal lobe, aside from the amygdala, is not uniform [4,5].

These findings led Pilleri to conclude that medio-basal lesions of the temporal lobe as well as a generalized cerebral disturbance are essential for KBS in humans [5]. Although bilateral temporal lobectomy in monkeys produces KBS consistently, Scoville and Milner reported that patients subjected to bilateral anterior temporal lobe resection suffer primarily from defects in recent memory and are remarkably free of gross behavioral changes [22]. There are only five

reports of partial expression of KBS following bilateral temporal lobectomy and most of these lack convincing visual agnosias or have only two of the characteristics of the syndrome [2,18,23-25]. Case reports of KBS have been confined to patients with encephalitis, degenerative brain diseases, or severe head trauma followed by temporal lobe resection.

Our patients do not help to clarify the anatomic basis of the behavioral abnormalities. Two of the three patients had evidence of abnormalities in electrical activity localized to the temporal lobe. One patient had evidence of bilateral atrophy of the temporal lobes which was associated with generalized cerebral atrophy. The lack of specificity of CT findings or EEG pattern was reported in a large series of patients by Aichner [3]. Nevertheless, our patients do underscore several points about KBS. Our experience suggests that KBS occurs more commonly in children than was suspected previously and it frequently occurs following hypoxia. The features of our patients are distinct from the symptoms observed commonly in hypoxic encephalopathy. Seizures and disturbances in memory and vision are frequent sequelae of hypoxia. Our patients, however, demonstrated visual agnosia which is distinct from cortical blindness commonly observed following hypoxia, in addition to other features of KBS not observed in hypoxic encephalopathy. Association of the KBS with hypoxia has not been reported previously. The prominence of sexual behavior in our patients indicates that alterations in sexual behavior may occur before puberty. This finding is in contrast to the speculation by Chutorian and Antunes that hypersexuality was not a feature of prepubertal children [16]. Our patients illustrate the transient nature of KBS symptoms, a feature which has not been recognized widely. The symptoms in two of our patients were transient; Patient 1 had an almost complete recovery, while Patient 3 had a rapid resolution of some symptoms with gradual improvement of others. Improvement or recovery also appears to occur in most adult patients who survive or in whom the underlying disease process is not degenerative or progressive [6]. Similarly, recovery from some KBS features has been observed in monkeys [26].

Finally, each of our patients appeared to have difficulty with auditory and visual memory, in addition to global effects on intelligence. The difficulty each patient had in recalling three objects after a 3-5 minute delay suggests a defect in intermediate memory when compared to their ability to repeat digits immediately. Recognition of intellectual difficulties has been helpful in adjusting the type of classroom and expectations for these children. The anticipation that behavioral and learning difficulties will improve gradually or will resolve also has been helpful for long-term planning.

It has been suggested that carbamazepine, haloperidol, and anti-cholinergic medication may be useful in improving KBS behavioral abnormalities [8,19,27,28]. We found carbamazepine useful in con-

trolling seizures but not useful in controlling behavioral abnormalities. Haloperidol and thioridazine also were not helpful beyond patient sedation. Considering the diversity of the pathologic processes and anatomical lesions responsible for this syndrome, it appears unlikely that a single medication will alter behavior substantially. Reports of improvement with medication fail to consider spontaneous recovery or improvement.

In summary, our experience suggests that KBS occurs more commonly in children than was suspected previously. Hypersexuality is a feature even in prepubertal children. It is important to identify the defect in intermediate memory in order to make appropriate educational plans for these children. Although there is no response to medication, behavioral and intellectual deficits do improve over time.

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