

Neuropsychological characteristic of post-traumatic Klüver-Bucy Syndrome

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Summary

Aim. Klüver-Bucy syndrome was described in the 50's of the 20th century as a group of neuropsychological symptoms such as changes in behaviour and emotions, hypersexuality (homo-, hetero-, autosexuality), changes in dietary habits (hyperorality), visual agnosia, "oral" tendency, hypermetamorphosis, which can develop in human beings, after bilateral damage or dysfunction of the medial part of temporal lobes. The main course of Klüver-Bucy syndrome may be central nervous system infection, especially herpetic, Pick disease, temporal epilepsy, paraneoplastic encephalopathy or bilateral brain injury. The disease is very rare in children and its occurrence in adolescents was described only in few cases. The intensity and the mode of its manifestation depend on many different factors.

Material and methods. The first comparative presentation of two clinical cases of children at the same age (16 years), following head injury (the same mechanism of trauma), with the description of computed tomography scans.

Results. The diagnosis of the syndrome does not require all the symptoms to be manifested simultaneously. Both patients were treated with carbamazepine. Regression of neuropsychological symptoms was different in both children despite similar brain damage.

Conclusions. Fully symptomatic Klüver-Bucy syndrome is very rare. Psychological status in Klüver-Bucy syndrome depends not only on the extent of the lesion, but also on emotional and intellectual development before the injury and social stimulation after brain damage.

Klüver-Bucy Syndrome / neuropsychological symptoms / neuropsychiatric disease

INTRODUCTION

In the 30's of the 20th century, two scientists - Heinrich Klüver and Paul Bucy - described the role of temporal lobe in regulation of emotional states. They did bilateral temporal lobectomies (which also involved the amygdalae, situated deep within the lobes) in Rhesus monkeys. The procedure resulted in a dramatic change in their behaviour, described by Klüver himself as the

most striking and apparent alteration ever observed in consequence of surgical experiments performed on animal brains. Klüver and Bucy divided the symptoms into six major groups: 1) "Mental blindness", today referred to as visual agnosia - inability to visually recognise objects in spite of normal visual acuity, 2) Hypermetamorphosis - the urge to react to every visual stimulus, 3) Change of dietary habits, e.g. eating large amounts of meat (the type of behavior was rarely observed in monkeys), 4) "Oral tendency" - the so-called oral perception - examination of objects by mouth through licking, sucking and chewing, 5) Emotional changes (one of the most distinct symptoms) - complete obliteration of fear responses to threatening painful stimuli, docility and loss of the so-called defen-

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sive aggression, diminished emotional reactivity (at times even completely obliterated, particularly apparent in case of aversive and painful stimuli), loss of social behaviours, 6) Changes in sexual behaviours, increased sex drive, hetero and homosexual behaviours, autosexuality [1, 2, 3]. Shortly after the discovery of Klüver and Bucy, similar neurobehavioural disturbances were observed also in humans following bilateral temporal lobe damage; neurology named the cluster of symptoms Klüver-Bucy syndrome (KBS) [4]. The first description of a patient with the syndrome originates from 1955; this was a patient after bilateral temporal lobectomy due to drug-resistant epilepsy [5]. In humans, KBS syndrome is a constellation of the following symptoms: reversal of emotional states (loss of fear and defense responses to pain or general disinhibition combined with euphoria), hypermetamorphosis, hyperorality (hyperphagia, bulimia, the so-called "raging hunger"), sex drive disturbances (leading towards masturbation). In some cases there also concomitant such symptoms as emotional lability, aggressive behaviours, loss of emotional ties with the loved ones, changes in dietary preferences. The diagnosis of Klüver-Bucy syndrome does not require concomitant occurrence of all the symptoms, since the fully symptomatic form of the disease is extremely rare [1, 2, 5, 6, 7, 8, 9, 10].

As it has been established, the key role in the genesis of the above symptoms is fulfilled by changes in neurotransmission resulting from destruction of the amygdale and its connections with other structures of the limbic system. When the efferent fibres extending from the temporal lobe to the amygdale are disrupted, the level of dopamine and serotonin drops, while the level of norepinephrine increases in the amygdale [11].

The most common cause of bilateral damage of the amygdalae and in consequence of Klüver-Bucy syndrome is herpetic encephalitis, frontotemporal dementia, late-stage Alzheimer disease, paraneoplastic limbic encephalitis, bilateral temporal lobe stroke, as well as disturbances associated with epileptic seizures. The syndrome may also develop in consequence of temporal lobe damage following resection of brain tumours situated in the medial line, as a result of transtentorial herniation and adrenoleukodystrophy. Cases of KBS syndrome have been de-

scribed following excessive restoration of sodium levels in hyponatremia and after chemotherapy (interferon alpha) [6, 7, 8, 12, 13, 9].

Klüver-Bucy syndrome develops relatively less frequently after craniocerebral injuries. Moreover, inasmuch as it is well documented in adult patients, descriptions of the syndrome in children are rare [4, 9, 10,]. In addition, reports addressing neuropsychological characteristics of patients with Klüver-Bucy syndrome focusing on their cognitive and intellectual functioning are also scant. As it follows from isolated studies (mostly case reports), in almost all cases, the patient with the syndrome also suffers from aphasia (predominantly of sensory character) [14] and amnesia (loss of episodic and declarative memory, anterograde amnesia) [6, 15]. In some patients, KBS syndrome is concomitant with semantic dementia, described as a form of frontotemporal lobar degeneration characterised by progressive fluent aphasia, anomia, reduced vocabulary, problems in understanding isolated words and their meaning [16]. Additionally, the affected individuals present with signs of visual agnosia and prosopagnosia (impairment of familiar face recognition leading in consequence to inability to recognise friends, loved ones and strangers) [10]. In single cases, problems in recognising facial expressions and discerning between positive and negative emotions have been also described [11].

The present article shall provide a description of two 16-year old patients with post-traumatic Klüver-Bucy syndrome. Thanks to a follow-up examination performed four years after the injury, it was possible to perform a long-term evaluation of cognitive and social functioning of the patients, as well as to compare the degree of their neuropsychological adaptation to their post-accident situation.

MATERIAL

The investigation included two patients – a girl and a boy aged 16 years – in whom Klüver-Bucy syndrome as a consequence of craniocerebral injuries had been diagnosed. The patients were treated at Neurosurgery Ward, University Children's Hospital of Kraków, in 2004. Both the girl and the boy suffered from craniocerebral injuries inflicted in road traffic accidents, although the

injury mechanisms were slightly different. The boy was a passenger of a car involved in a car crash (MVA – Moving Vehicle Accident), during which he was thrown through the windshield while traveling without his seat-belt fastened. The girl was hit by a car on a pedestrian crossing (RA – Road Accident). The victims were admitted to University Children's Hospital of Kraków in severe condition with Glasgow Coma Scale score assessed as 3; they were unconscious, with respiratory and circulatory failure. Their condition indicated a necessity of placing the patients in Intensive Care Unit, employ respiratory therapy and infusions of catecholamines. Neurologically, the female patient did not present with meningeal syndrome, her pupils were narrow, poorly reacting to light, she manifested anisocoria (R>L) and in response to a painful stimulus, she demonstrated extension reaction. The boy responded to painful stimuli with flexion of all four extremities, he was deeply unconscious, his pupils were of equal size but narrow, poorly reacting to light stimuli, he presented with bilateral extensor plantar response. Stat imaging (computer tomography in both cases) demonstrated extensive intracranial pathologies. The girl presented with a subdural hematoma in the left temporal region, a supradural hematoma in the right temporal region with hemorrhagic brain contusion involving the right hemisphere and intensified cerebral edema. Head CT of the boy showed bilateral contusion in the temporal and parietal lobes, hemorrhage into the subcortical nuclei and a paracerebral hematoma. The girl required surgical treatment to evacuate the supradural and subdural hematomas and she was subjected to left frontotemporoparietal craniotomy. The postoperative course was uneventful and within a few days her condition improved. Neurologically she presented with mixed aphasia, increased muscle tone in four extremities, intensification of deep reflexes, left extensor plantar response and involuntary movements. The boy was treated conservatively. In the initial days of hospitalisation his general condition deteriorated, he did not regain consciousness, developed right hemiplegia and occasionally flexion responses. On day 12 following the admission, he regained consciousness, was capable of logically interact with others, yet continued to present with right hemiparesis and right

positive extensor plantar reflex. Both patients received antiedematous therapy and therapeutic rehabilitation. The boy required psychiatric treatment with haloperidol due to his intensive positive productive symptoms.

Based on the imaging studies and clinical observations, post-traumatic Klüver–Bucy syndrome was diagnosed in both patients. Among neuropsychological disturbances, a tendency towards oral perception predominated (the female patient reached for objects and placed them in her mouth), hypermetamorphosis and coprolalia. Her dietary habits changed and she exhibited a tendency towards hyperoral behaviours. She had excessive appetite. Additionally, she manifested auto and allopsychic disorientation, was periodically confused and agitated. The patient was visibly emotionally labile, demonstrating behaviours ranging from uncontrolled bouts of aggression and expressing her anger by physical force to complete passivity, emotional dullness, indifference and stupor.

Among symptoms of Klüver–Bucy syndrome observed in the boy, a tendency towards oral perception was also the most apparent; on many occasions, the patient attempted eating inedible objects (coins, building blocks). He manifested psychomotor sluggishness, emotional passivity and periodically agitated. He showed tendencies towards hypersexual, especially autosexual behaviors (masturbation).

Intensified symptoms of Klüver–Bucy syndrome were successfully controlled employing the commonly accepted therapy, namely administration of carbamazepine 3 x 200 mg p. o., with the serum therapeutic level of 6µg/ml. The management allowed for improving the condition of both patients. Following hospitalisation, the boy demonstrated resolving right hemiparesis (grade 4/5 according to the Lovett's scoring scale), while coprolalia persisted, although to a lesser degree, similarly as a tendency towards masturbation and hypermetamorphosis. Neurological examination of the female patient on the day of discharge showed signs of residual sensorimotor aphasia and less intense selected symptoms of KBS syndrome described above.

METHOD

Data from patient history collected from the family members and observation of the patients' behaviour:

Four years after completion of inpatient treatment, both patients were subjected to catamnestic examinations. Neurologically, neither the boy nor the girl did manifest signs of focal central nervous system damage. An alarming observation, however, were neuropsychological disturbances that predominated in the clinical picture, especially in the female patient.

The objective of the neuropsychological examination was assessment of cognitive and intellectual efficiency, adaptation to fulfilling social roles and adaptation to the situation.

Education

As it followed for history taken from the father, the social functioning of the patient – currently 20 year old – approximated the pre-accident level. The boy continued his education (vocational training for a car mechanic). He completed his practical education in a car shop, yet was unemployed at the time of the study. The girl, his peer, completed education in a vocational special school. As it followed from her mother's relation, the patient's school performance was unsatisfactory. Prior to the accident, her grades had been much better; currently, she experienced problems with learning new information and remembering facts. After the accident, the patient clearly lost most of her previous interests, she stopped reading, watching television and being interested in current events happening in Poland and abroad. No similar memory deficits or problems with assimilation of simple information were observed in the boy, as reported by his family members. He continued pursuing his former passions and interests. He completed a driving course and drove a car on his own. None of the patients developed epilepsy.

Social functioning

The boy's relations with his peers were satisfactory, he had a group of close friends; in the

opinion of the father, his relations with siblings were good. The girl's relations with peers were not so satisfactory. She had a small group of male acquaintances, mostly from school. For some time now, she was dating a schoolmate. However, problems were encountered in her relations with younger siblings; she tended to be irritable and aggressive towards her sibs and other family members, even resorting to physical violence. Her character changed drastically, she found it difficult to control anger episodes, which were in contrast with her tendency towards apathy and taciturnity. Her behaviours tended to be excessively spontaneous, unforeseeable and – in the opinion of her mother – even irrational and dangerous. On many occasions, they posed a threat not only to the patient herself, but also to others (attempts at exiting the house through a window situated at a high altitude). The boy's father also signalled a slight tendency of his son to have problems with controlling emotions, but such disturbances were sporadic and did not cause major anxiety.

In the mother's assessment, in the post-accident period, the girl became much less independent and sporadically agreed to stay home alone. She was reluctant to participate in family life; her contact with family members also deteriorated. She had a tendency towards deluding others into the belief in facts or events that in reality had not occurred. She also had problems with remembering and tended to fill memory gaps with confabulations. She found it difficult to understand her behaviour, did not accept criticism or information indicating errors she had made. Her nutritional habits also changed and she lost the distinction between foods she liked and did not like. A significant problem was posed by her tendency to uncurbed appetite and eating binges, what forced the family to impose some external control and limit her access to food.

Tools employed in the neuropsychological examination:

Wechsler Adult Intelligence Scale WAIS – R (PL).

Selected experiments from the Łucki's Set for examinations of patients with organic brain damage.

RESULTS

In the course of the neuropsychological examination, the female patient seemed to be withdrawn and taciturn. Her statements were poor in contents; many times she lost the drift of conversation. She manifested visible difficulties in word finding, decreased verbal readiness characterised by aphasic difficulties – *residua amnestica*.

On the contrary, the patient was talkative; he behaved and conversed adequately to the situation. His statements, though of low complexity, were logical and coherent.

Neuropsychological examination of the female patient demonstrated normal auto and allopsychic orientation.

Speech examination demonstrated decreased verbal fluency, both semantic and phonemic. There was a pronounced tendency towards perseveration. Analysis of spontaneous and propositional speech confirmed *residua amnestica* accompanied by a distinct difficulty in word finding.

Memory testing showed pronounced deficits of short-term auditory memory of material arranged into a logical unity. The patient again manifested numerous perseverations. Moreover, she showed disturbed ability to learn new material as demonstrated by Luria's Learning List. The investigators observed a low durability of the memory trace. The patient did not display a critical approach to her memory deficits. No visual gnosis defects were observed.

Intellectual functioning of the female patient was tested employing the Wechsler Adult Intelligence Scale WAIS- R (PL). Her full-scale IQ score was 83, what placed her below average. A significant difference was observed between the verbal (IQ 65) and performance (IQ 105) score, what indicated an organic central nervous system lesion. While analysing the profile of results, it was apparent that the girl's linguistic aptitude was clearly decreased, what might have resulted from persistent residual aphasia. The patient was characterised by a limited vocabulary of notions and ability to define them. She also scored low in general knowledge, ability to form abstract concepts and understanding of social situations. She received relatively higher scores, within the average range, in performance tests. The patient

was observant and capable of organising observable data. Her ability to visually analyse and synthesise was average. Also her abilities to practice causative reasoning and anticipate consequences of an initial event were within the average range. On the other hand, the analysis of the results achieved in the performance scale showed her to score lowest in the "digit symbol" subscale, what indicated her learning problems, especially in involuntary visual-motor learning.

Neuropsychological examination of the male patient demonstrated normal auto and allopsychic orientation. The patient showed normal attention focus. While he performed particular experimental tasks, he demonstrated a coherent plan of action (the serial subtraction task was performed fast and without any mistakes). His verbal fluency was decreased, especially his letter verbal fluency. Yet testing his linguistic processes did not show any signs of aphasia.

Examination of mnemonic processes demonstrated normally functioning short-term visual memory. His results were slightly poorer in short-term auditory verbal memory testing involving material arranged in a logical unity, what indicated decreased durability of memory trace.

Intellectual functioning of the boy was tested employing the Wechsler Adult Intelligence Scale WAIS- R (PL). His full-scale IQ score was 84, what placed him below average. No significant difference was noted between the verbal Scale (IQ 84) and performance scale (IQ 85) score, although the profile of results was markedly disharmonious. He achieved the lowest scores in the following subscales: "information", "vocabulary", "similarities", "digit symbols", what indicated a low level of general knowledge, vocabulary of and ability to define notions, abstract reasoning and ability in the field of visual-motor learning. His understanding of social situations and the so-called social intelligence were below average. On the other hand, his ability to perform mathematical operations, perceptiveness, ability to perform visual analysis and synthesis and causative reasoning were within the average range. It should be emphasised, however, that compared to his WAIS-R (PL) score achieved immediately after the accident in 2004 (full-scale IQ score of 67, verbal scale - IQ 79, performance scale - IQ 61), an improvement was noted in the patient's intellectual functioning.

DISCUSSION OF RESULTS

Four years following the accident, the female patient continued to present with some symptoms of Klüver-Bucy syndrome, mostly as manifestations of an organic personality change. These included hyperorality and change in dietary habits, emotional dullness and the so-called decrease of high emotionality interchanged with impetuosity and impulsiveness, as well as a tendency towards reacting with physical violence. She showed decreased criticism of her behaviours and disinhibition. Her interests dwindled.

The history of the patient obtained from her mother did not confirm hypersexuality, but it should be borne in mind that the mother might have neglected reporting intimate information on sexual behaviours of her daughter. A detailed neuropsychological examination revealed additional problems in cognitive functioning of the patient, which are also listed in the literature as neuropsychological components of KBS syndrome, being a consequence of bilateral permanent frontal lobe damage. What is meant here are residual speech problems in the form of amnesic aphasia and amnesic disturbances involving episodic memory, auditory-verbal memory and ability to learn new material. Decreased verbal fluency, flexibility of thinking and fluency of speech were also noted. The above personality-associated and cognitive problems are most likely permanent. Such symptoms of KBS syndrome as for example hypermetamorphosis, apparent in the immediate post-injury period, subsided; they are probably characteristic of the early, acute stage of the disease and central nervous system damage that bilaterally involved the temporal lobes.

In the case of the male patient, a catamnestic examination performed four years after the injury showed resolution of the majority of symptoms characteristic of Klüver-Bucy syndrome. No hyperorality was noted, nor hypersexuality, visual agnosia, excessive impulsiveness or change in dietary habits. Nevertheless, the patient demonstrated a significant decrease of his intellectual abilities, with IQ score below average, decreased verbal fluency, deficits of short-term auditory memory of material arranged into a logical unity. There was a continued tendency

towards concrete thinking (loss of abstract attitude) and a decreased ability in the field of visual-motor learning.

It should be emphasised that – despite the above listed cognitive problems – the patient managed to effectively adopt himself to his post-accident situation, both from the professional and social viewpoint.

CONCLUSIONS

Klüver-Bucy syndrome is a neuropsychiatric disease, where the intensity of neurological deficits does not correspond with personality disturbances.

Oftentimes, the consequences of a head injury and symptoms of the syndrome are not observable “at a first glance”, and yet a more detailed neuropsychological examination and closer contacts with the patient reveal personality-associated issues as being in the foreground. Unfortunately, these problems are reflected in every day functioning of the patient, resulting in social maladjustment, lack of independence and a need for being cared for.

While analysing the results of follow-up neuropsychological examinations of the two patients, one notes they markedly differ in the present level of functioning and intellectual capacity, with the male presenting better performance. The difference seems to be interesting inasmuch as there is a similarity in the age of the patients, location and extent of their brain injuries. We should probe into possible causes of such a difference. Possibly, it is ingrained in environmental determinants and associated with stimulation of cognitive processes by individuals closest to the patient. The girl came from a large family (6 children). Perhaps the attention of close individuals was to a lesser degree concentrated on the patient (interestingly, during history taking, the mother was unable to report the medications the patient was taking, claiming she did not remember the names). After the accident, the girl was placed in a special school where, as reported by the mother herself, the educational level was exceptionally low; conceivably, the patient was not confronted with sufficient demands that might stimulate her cognitive functioning.

On the other hand, the boy immediately began learning a practical job. He stayed among healthy individuals, what might have had a positive effect on his adjustment to life and intellectual functioning.

Finally, an interesting fact is that none of the patients sought help of a neuropsychologist in association with injury negative consequences of a psychological nature they experienced after the accident. In such patients, early initiation of specialist therapeutic assistance directed towards rehabilitation of selected cognitive processes and modification of pathological behaviours is highly recommended. As to pharmacotherapy, when Klüver-Bucy syndrome is suspected, carbamazepine therapy must be early introduced, with the dose corresponding to serum level of 6-10 µg/ml.

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