

BRIEF COMMUNICATION

Partial Klüver–Bucy syndrome in a Paediatric patient: A post-neurosurgical and neuropsychological cases

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Abstract

A variety of cognitive, behavioural, and emotional impairments have been reported in the literature that are associated with the resection of the temporal cortex. Klüver–Bucy syndrome is one infrequently reported disorder in the paediatric population. This paper describes the neuropsychological findings of a female paediatric patient at 7 and 10 years of age with a diagnosis of partial Klüver–Bucy syndrome (pKBS) following total resection of the amygdala and right hippocampus to resect a glioma. The patient presented emotional problems, aggressiveness, hypermetamorphosis, social indifference, and behavioural dysexecutive syndrome, which was found at both 7 and 10 years, but with a decrease in the severity of alterations in attention, impulsivity, hyperactivity, and aggressive behaviour in a second evaluation after she had a neuropsychological intervention. These findings describe the neuropsychological profile of paediatric case with resection of the amygdala and right temporal lobe.

KEYWORDS

case study, dysexecutive syndrome, partial Klüver–Bucy syndrome; neuropsychology, right amygdala

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INTRODUCTION

Klüver–Bucy syndrome (KBS) is a rare neurobehavioural disorder caused by an alteration in the functioning of the medial temporal lobes (amygdala, temporal, and hippocampal networks). The presentation of this syndrome is not homogeneous but typically includes hyperorality, hyperphagia, indiscriminate dietary behaviour, hypersexuality, apathy, visual agnosia, distractibility, emotional and memory impairment, and excessive visual attentiveness, known as hypermetamorphosis (Bertoux et al., 2018; Jha & Ansari, 2010; Juliá-Palacios et al., 2018; Lippe et al., 2013). Latchminarine et al. (2022) define classic KBS as including all of the cardinal symptoms, and partial Klüver–Bucy syndrome (pKBS) as including three cardinal symptoms or other symptoms, could be diagnosed if the temporal neocortex or amygdala is affected in the absence of the bilateral classic temporal lesion (Bhat et al., 2009; Carroll et al., 2001), and it can occur at any age (Lippe et al., 2013). The major symptoms reported in children (Table 1) are the presence of emotional impairment (lability, irritability, mood swings, and excessive joy), aggressiveness, attentional difficulties due to hypermetamorphosis, and indifference towards peers and parents; the variability of the symptoms in children could be associated with the immaturity of some neural networks and lack of learning of some behaviours (Lippe et al., 2013).

The objective of this clinical case study was to describe the neuropsychological findings in a patient at the ages of 7 and 10 years after complete resection of the amygdala and right temporal lobe for a temporal glioma. The patient was diagnosed with pKBS based on the presentation of hypermetamorphosis, emotional flattening, mood swings (irritability and mood lability), memory deficits as cardinal and other symptoms, and right temporal and amygdala damage.

CLINICAL CASE

Case description

The patient is a 10-year-old daughter of healthy Mexican parents. This was the mother's second pregnancy; it was a twin gestation. The mother had prenatal care from the second month of pregnancy with hematinic and folic acid. The baby was delivered by caesarean section at 37.5 weeks' gestation; she cried and breathed immediately at birth, with an APGAR score of 8–9, weighed 2350 g, and measured 43 cm. At birth, she presented hypoglycaemia and inflammatory stomach reaction to formula feeding and required an incubator for 5 days. From 7 to 8 months of age, she was treated for epilepsy with valproate, with a seizure frequency of two to three per day. She required medical attention from age 5 to 6 because she presented bronchial aspiration while eating.

TABLE 1 Symptoms and diagnostic criteria for complete and Partial Klüver–Bucy Syndrome in Children.

Cardinal symptoms	Other symptoms	Diagnostic criteria	
		Klüver–Bucy syndrome	Partial Klüver–Bucy syndrome
Hyperorality	Polydipsia	All cardinal symptoms and bilateral temporal brain damage	Three cardinal or other symptoms and unilateral temporal cortex damage
Hypersexuality	Sleep disorders		
Hypermetamorphosis	Seizures		
Visual Agnosia			
Emotional flattening			
Memory problems			
Expressive language disorders			
Increased appetite			
Apathy			
Mood swings between irritability and emotional attachment			

According to the parents, before the age of 7, she was socially isolated, with difficulty relating to her peers, uninhibited behaviour, expression of inappropriate ideas for the social context, and difficulty in social interaction. She attended 2 years of public preschool beginning at age 4, during which time her teachers reported behavioural problems and recommended that her parents take her to a specialist to be evaluated for attention deficit hyperactivity disorder (ADHD). An ADHD diagnosis was confirmed the same year by medical personnel, but the parents indicate that no psychometric or neuropsychological assessments were performed, and she received no therapeutic or pharmacological treatment. At the age of 6, she began primary school, where she presented difficulties in reading, writing, and mathematics.

When she was 7, a first MRI found a right temporal glioma that compromised the hippocampus, amygdala, parahippocampal gyrus, and lateral ventricle at the level of the temporal horn (Figure 1.1). A complete resection of the amygdala and right temporal lobe was performed (Figure 1.2).

In 2019, 1 month after surgery, she received a neuropsychological evaluation to explore her cognitive functions, behaviour, and emotions, and 3 years later, a follow-up assessment was performed. Currently, at age 10, she is in fourth grade and takes remedial and special education classes in addition to the regular curriculum. From August 2021 to January 2022, before the second assessment, she received an intervention using TeleNeuropsychology (TeleNP) due to the COVID-19 pandemic. This intervention, which focused on attention, working memory, and learning abilities, used the Zoom® platform, with three weekly sessions lasting 45–60 minutes, for 18 weeks.

Instruments

Because the evaluation was carried out 1 month after neurosurgery, the patient presented behavioural and cognitive difficulties with the application of a broad neuropsychological battery; so it was resorted to using behavioural inventories; however, it is important to emphasize that our country has a limited number of standardized instruments; therefore, the description of the results was carried out with inventories that have revealed high reliability and validity even for other ages of our population, such as the BRIEF-P (García-Anacleto & Salvador-Cruz, 2017).

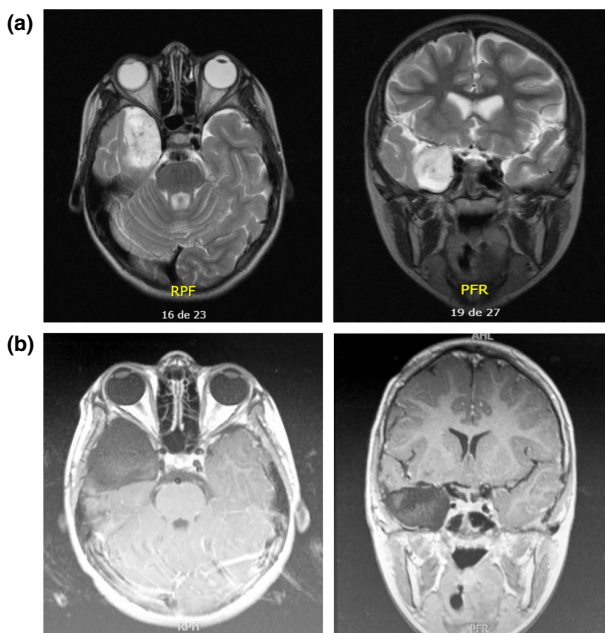


FIGURE 1 Horizontal (left) and Coronal (right) Images of the Right Temporal Glioma. (a) before neurosurgery (above); (b) after neurosurgery (down).

Wechsler intelligence scale for children (WISC-IV)

This instrument assesses total IQ and verbal comprehension index, perceptual reasoning, working memory, and processing speed. It is standardized for the Mexican population and was administered in the first and second assessments (Wechsler, 2008). Although each WISC-IV subtest is part of the intelligence construct, some of these could be associated with the assessment of certain cognitive domains, for example, the comprehension subtest with social judgement (Hernández Galván & Yáñez Téllez, 2013). To have a major descriptive of the cognitive function, we made an interpretation of each task according to Flanagan and Kaufman (2009), Brenlla (2013), Cantú (2016), and Lezak et al. (2012) criteria; because the patient's cognitive limitations and behaviour dysfunction made it difficult to apply another neuropsychological instrument.

Behavioural assessment of executive function-2 (BRIEF-2)

The BRIEF-2 inventory ecologically evaluates behaviours associated with alterations of executive functions in children (Maldonado et al., 2017). It was administered in the first and second assessments. Both questionnaires (at 7 years and 10 years) were answered by the mother.

ADHD-5 assessment scale for children and adolescents (DuPaul et al., 2018)

This scale is used to detect or diagnose ADHD according to the criteria of the DSM-5. It was administered in the first and second assessments. Both questionnaires (at 7 years and 10 years) were answered by the mother.

SENA child and adolescent evaluation system (Fernández-Pinto et al., 2015)

This instrument evaluates emotional and behavioural problems in patients aged 3–18 years. It was administered in the first and second assessments. Both questionnaires (at 7 years and 10 years) were answered by the mother.

Neuropsi attention and memory (Ostrosky et al., 2019)

This instrument was administered in the second evaluation to assess attention skills, memory, and executive functions. It has normative scores for the Mexican population. It could not be applied in the first evaluation because of the patient's problems with attention, understanding, and emotional lability.

Ekman face test (Ekman et al., 1976)

This test assesses the ability to identify emotions in faces. The Mexican normative data and version used includes 30 faces (six each of joy, sadness, anger, and fear, and six neutral faces) and has been used to assess and compares Mexican people with Williams syndrome, Down syndrome, and neurotypical development (Serrano-Juárez, 2020; Serrano-Juárez et al., 2021) It could not be administered in the first session because of the patient's problems with attention, understanding, and emotional lability.

To obtain a better description of the patient's profile and to unify all the scores into a single measurement scale, the standardized scores were converted into z -scores ($z = \left(\frac{\text{Patient Score} - \text{Mean score}}{\text{Standard Deviation}} \right) - 1$). The z -scores of the BRIEF-2 and SENA scales were multiplied by -1 because high standardized scores

on these scales indicate poor functioning; with this transformation, low z -scores (< -1) indicate poor functioning or a higher prevalence of problematic behaviours. The criteria used for interpretation for all scores were: severe impairment < -2 ; below average < -1 ; average -1 to 1 , and above average > 1 .

The reliable change index (RCI; Jacobson & Truax, 1991) was used to compare the results between the two evaluations. The data for the normative samples were used for both analyses as there is no clinical sample for pKBS. For the comparison of the scores of the standardized instruments and to control the assessment learning effect, the RCI with the Iverson (2018) modification was used. A score greater than 1.96 on the RCI implies a significant clinical change with $p < .05$ (Duff, 2012; Iverson, 2018; Jacobson & Truax, 1991).

RESULTS

During the post-surgical evaluation in 2019, the patient was uncooperative, easily distracted, and required constant motivation, and breaks were taken so that she could carry out the requested tasks. The results of the first and the 2022 follow-up evaluation are shown in Table 2 (standardized instruments) and Table 3 (behavioural questionnaires).

In the first assessment, the total IQ (TIQ) score of 60 was not interpretable due to the variability of the component scores; however, a borderline index of general capacity (IGC) of 74 was obtained. It was observed that the patient had an adequate level of awareness, perception, language, and personal orientation, and obtained average scores on standardized instruments measuring perceptual reasoning and visuoconstructive praxis. However, the assessment also found severe impairment in digit span, working memory, verbal and visual abstract thinking, social judgement, vocabulary amplitude, processing speed, and semantic long-term memory. Behavioural data assessment with questionnaires showed externalized problems, such as anger control, aggression, defiant behaviour, and poor functioning of personal-social resources, such as problems with emotional regulation, integration and social competence, emotional intelligence, and isolation. In daily life behaviour executive functions, there were severe impairments of working memory, inhibition, flexibility, self-monitoring, emotional control, inhibition, and planning and organization. On the ADHD-5, the patient showed a high prevalence of hyperactive-impulsivity behaviour but not inattention.

The second evaluation found minor problems of initiative and apathy at the beginning of the test administration; however, as the assessment progressed, the patient's attitude improved. She obtained a non-interpretable TIQ of 60 and a borderline IGC of 70. She showed adequate language and personal orientation and had average scores on standardized instruments measuring working memory, motor functions, visuoconstructive praxia, Stroop inhibition, and face emotion recognition, but severe impairment was found in vocabulary amplitude, social judgement, semantic long-term memory, visual abstract thinking, logical reasoning, digit and block span, sustained and selective attention, categorization, and verbal fluency. Behavioural questionnaires showed that externalized and emotional problems had decreased significantly since the first evaluation ($p < .05$) to the point of obtaining average scores in anger control, aggression, and defiant behaviour. Some personal–social resources, such as isolation and integration and social competence, also showed significant improvement ($p < .05$), but were still in the below-average and severe impairment range. Daily life behaviour executive functions of flexibility, initiative, working memory, and organization of materials were still poor; average scores and reliable change ($p < .05$) were obtained in inhibition, emotional control, task monitoring, and planning and organization and self-monitoring have a negative reliable change and minor score. On the ADHD-5, the patient showed minor hyperactivity-impulsivity and inattention behaviours ($p < .05$). She showed better cooperation, group integration, and learning, with no errors in the articulation of phonemes except for /rr/. She could write short sentences, albeit slowly, and sometimes with spelling mistakes and excessive space between words. She could perform simple addition and subtraction with supporting material but confused the plus and minus signs and made errors.

TABLE 2 Results of Neuropsychological Instruments at 7 and 10 Years Old.

Scale	7 years		10 years	
	Z-score	Interpretation	Z-score	Interpretation
WISC-IV				
Verbal Comprehension Index	-2.2	Severe impairment	-2.6	Severe impairment
Similarities (Verbal abstract thinking)	-1.3	Below average		n/a
Vocabulary (Vocabulary amplitude)	-2	Below average	-2.3	Severe impairment
Comprehension (Social judgement)	-2.3	Severe impairment	-3	Severe impairment
Information (Semantic long-term memory)		n/a	-1.7	Below average
Perceptual Reasoning Index	-0.9	Average	-1.1	Below average
Block design (Visuoconstructive praxia)	0.3	Average	0	Average
Picture Concepts (Visual abstract thinking)	-1.7	Below average	-1.3	Below average
Matrix reasoning (Logical reasoning)	-1	Average	-1.3	Below average
Working Memory Index	-2.5	Severe impairment	-2.3	Severe impairment
Digit span	-1.3	Below average	-1.7	average
Number and letter sequencing	-3	Severe impairment		n/a
Arithmetic		n/a	-2.3	Severe impairment
Processing Speed Index	-3.3	Severe impairment	-2.9	Severe impairment
Coding	-3	Severe impairment	-2.3	Severe impairment
Symbol search	-3	Severe impairment	-2.3	Severe impairment
Total IQ	-2.7	Severe impairment	-2.7	Severe impairment
General Capacity Index	-1.7	Below average	-2	Below average
Cognitive Proficiency Index	-3.3	Severe impairment	-2.1	Severe impairment
Neuropsi Attention and Memory				
Orientation				
Time orientation	n/a		-3	Severe impairment
Space orientation	n/a		-3	Severe impairment
Personal orientation	n/a		0	Average
Attention				
Digit span	n/a		-2.7	Severe impairment
Block span	n/a		-1.3	Below average
Visual detection (correct)	n/a		-2	Below average
Digit regression	n/a		-0.7	Average
Block regression	n/a		-1	Average
Executive function				
Category Formation	n/a		-2.3	Severe impairment
Semantic fluency	n/a		-2.3	Severe impairment
Phonological fluency	n/a		-3	Severe impairment
Motor functions	n/a		0.3	Average
Stroop (time)	n/a		0	Average
Stroop (correct)	n/a		1	Average

TABLE 2 (Continued)

Scale	7 years		10 years	
	Z-score	Interpretation	Z-score	Interpretation
Ekman faces				
Face emotion recognition				
Joy	n/a		0	Average
Sadness	n/a		3	Above average
Anger	n/a		1.6	Above average
Fear	n/a		1.4	Average
Neutral	n/a		1	Average
Total	n/a		3	Above average

Note: For Z-scores, M = 0 and SD = 1; n/a = not administered; severe impairment > -2; below average > -1SD; average \pm 1.5 SD; above average > 1.5 SD.

DISCUSSION

The objective of this clinical case study was to describe the neuropsychological findings in a patient at the ages of 7 and 10 years after complete resection of the amygdala and right temporal lobe for a temporal glioma. The results show that the patient met the diagnostic criteria for pKBS 1 month after the neurosurgical intervention and again 3 years later, with signs and symptoms of hypermetamorphosis, apathy, emotional lability and memory impairments as cardinal symptoms, and social indifference and problems of social integration with peers and parents as secondary symptoms. She also met the Godefroy et al. (2018) criteria for a behavioural dysexecutive syndrome. The patient already presented some dysexecutive symptoms (inattention and disinhibition) and poor learning that were probably associated with the presence and localization of the glioma; after the neurosurgery, the symptoms increased and new signs appeared in the assessment, including hypermetamorphosis, apathy, social indifference, and emotional lability and unilateral temporal and amygdala damage, that were sufficient for a diagnosis of pKBS with a behavioural dysexecutive syndrome. These neuropsychological findings describe the behavioural and cognitive profile of a girl with right amygdala and temporal cortex that could be related to social behaviour and emotion. According to these results, we propose three possible hypotheses about these findings:

The first hypothesis: the social and right amygdala

The amygdala is believed to function as an emotional processor, organizing sensory input and emotional responses through the activation of different subcortical centres (Janak & Tye, 2015; Sánchez-Navarro & Román, 2004). Its connections with the hippocampus and the cingulate, orbito-frontal, and ventromedial cortices play a role in working memory, emotional decision-making, and the organization of action based on emotion. In addition, the network of the medial and basolateral amygdala and the hippocampus has been described as responsible for the system of social reward (Felix-Ortiz & Tye, 2014; Hu et al., 2021), and the right amygdala seems to have a major influence on social behaviour: damage to it has been associated with isolation behaviours (Adolphs, 2003; Baeken et al., 2014) and multiple changes on the social behavioural and emotional levels (Venta et al., 2018; Watanabe et al., 2019), and usually causes reactive aggressiveness, alexithymia, antisocial behaviour, and other externalized behavioural problems (Farah et al., 2018; Felix-Ortiz & Tye, 2014; Hu et al., 2021; Sánchez-Navarro & Román, 2004). We suppose that minor social interactions with peers in our patient could be associated with right amygdala damage and affect the social responses in multiple social contexts.

TABLE 3 Results of behavioural questionnaires at 7 and 10 Years Old.

Scale	7 years		10 years	
	Z-score	Interpretation	Z-score	Interpretation
BRIEF-2				
Behavioural Regulation Index	-2.5	Severe impairment	-2	Below average
Inhibition ^a	-2.4	Severe impairment	-0.8	Average
Self-monitoring ^a	-2	Below average	-3.6	Severe impairment
Emotional regulation index	-3	Severe impairment	-2.2	Severe impairment
Flexibility	-3	Severe impairment	-3.4	Severe impairment
Emotional control ^a	-2.5	Severe impairment	-1	Average
Cognitive regulation index	-1.1	Below average	-1.2	Below average
Initiative	-1	Average	-2	Below average
Working memory	-1.1	Below average	-1.4	Below average
Planning and organization ^a	-1.9	Below average	-0.7	Average
Task monitoring ^a	1.3	Above average	-0.2	Average
Organization of materials	-1.2	Below average	-1.1	Below average
General Index of Executive Function	-2.3	Severe impairment	-1.9	Below average
SENA				
Index of emotional problems	1.1	Above average	0.3	Average
Depression	0.6	Average	0.3	Average
Anxiety	0.6	Average	-0.1	Average
Social anxiety ^a	1.2	Above average	-0.3	Average
Somatic complaints	1	Average	1.1	Below average
Index of behavioural problems ^a	-3.1	Severe impairment	0	Average
Attention problems	-0.2	Average	-0.1	Average
Hyperactivity-impulsivity	-0.2	Average	0.5	Average
Anger management problems ^a	-2.3	Severe impairment	0.9	Average
Aggression ^a	-3.6	Severe impairment	-0.5	Average
Defiant behaviour ^a	-2.2	Severe impairment	-0.6	Average
Unusual behaviour	-0.9	Average	-0.9	Average
Personal Resource Index ^a	-3.4	Severe impairment	-2	Below average
Emotional regulation problems	-1.4	Below average	-0.2	Average
Rigidity	-1.1	Below average	-0.3	Average
Isolation ^a	-3.9	Severe impairment	-1.4	Below average
Integration and social competence ^a	-4.5	Severe impairment	-3.1	Severe impairment
Emotional intelligence	-1.5	Below average	-0.4	Average
Willingness to study	-1.7	Below average	-1.2	Below average
Index of problems in executive functions	-1.0	Average	-0.3	Average
Global Problem Index	-0.7	Below average	-0.2	Average

TABLE 3 (Continued)

Scale	7 years		10 years	
	Z-score	Interpretation	Z-score	Interpretation
ADHD-5				
Inattention ^a	-1	Average	0	Average
Hyperactivity-impulsivity ^a	-1.22	Below Average	0	Average
Inattention + Hyperactivity-impulsivity ^a	-1.17	Below Average	0	Average

Note: For Z-scores, M=0 and SD=1; n/a=not administered; severe impairment > -2; below average > -1SD; average \pm 1.5 SD; above average > 1.5 SD.

^aRCI>1.96 ($p<.05$).

The second hypothesis: The social brain hypothesis (Adolphs, 1999, 2001, 2002; Sánchez-Cubillo et al., 2012)

Adolphs social model posits three levels, with different domains and brain structures for social behaviour, where damage to these structures causes problems in social behaviour (Adolphs, 2003). In the case study presented here, the patient seems to retain the ability to identify and recognize emotions in faces (social perception) but also seems to have alterations in social judgement and social approach behaviour (social cognition and behaviour). In the social brain hypothesis, the right temporal sulcus has been identified as the basis for identification or prediction of movement and behaviour, and the temporal pole is responsible for social motivation, so access to past experiences and the ability to identify the behaviour of other people could influence social approach behaviour (Sánchez-Cubillo et al., 2012). The social deficit and emotional lability in this case could be related to failures in the social judgement and emotional intelligence domains of social cognition; both were associated with temporal and right amygdala in the social brain hypothesis (Adolphs, 2001, 2002).

The third hypothesis: the behavioural dysexecutive syndrome

Although the alteration of executive functions is not considered a symptom in pKBS patients, both our findings and those of previous case studies (Bertoux et al., 2018; Juliá-Palacios et al., 2018; Lippe et al., 2013) report symptoms of an orbito-ventro-prefrontal dysexecutive syndrome (abulia, disinhibition, aggressiveness, and emotion lability). According to Godefroy et al. (2018), the diagnosis of dysexecutive disorder can be used for disorders of behavioural and cognitive executive function, regardless of the location of brain damage, and could show one of three profiles: a behavioural dysexecutive disorder, a cognitive dysexecutive disorder, or both (Godefroy et al., 2010, 2018). For a behavioural dysexecutive disorder, the patient must show: (1) a behavioural disorder assessed by direct observation or an inventory; (2) highly suggestive deficiencies, such as hypoactivity with apathy, hyperactivity with distraction or disinhibition, persevering behaviour, and/or social behaviour disorders; and (3) supporting disorders, such as anosognosia, confabulation, and/or paramnesia. For cognitive dysexecutive disorder, the patient must have (1) cognitive disorder assessed with a test; (2) highly suggestive impairments, such as initiation and sustained alertness, arrogance, planning, inhibition, and/or change-of-scene disorder; and (3) supporting disorders, such as those in theory of mind, emotion identification, metacognition, strategic processing in episodic memory, working memory, and/or coordination in dual tasks (Godefroy et al., 2010, 2018). The patient described here seems to show behavioural problems measured with an inventory, hypoactivity with apathy, and social behaviour problems as highly suggestive deficiencies, and memory problems with confabulation errors and intrusions as supporting disorders and met the Godefroy et al. (2010) and Godefroy et al. (2018) criteria for behavioural dysexecutive syndrome. Godefroy et al. (2010, 2018), Baeken et al. (2014), and Watanabe et al. (2019) found that the

fronto-temporo-amygdala networks have an important function in decision-making, behavioural anticipation, self-monitoring, and cognitive flexibility, and suppose that the damage in this network could reveal dysexecutive symptoms.

Finally, the findings of this case also describe TeleNP intervention on the behaviour of children with pKBS. This intervention included restorative and compensatory strategies for the stimulation of attention and working memory and behavioural techniques provided to parents for executive function. It could have influenced the scores on the second assessment and possible improvement in inattentive, hyperactive, impulsive, and aggressive behaviour, as well as working memory scores. Her social behaviour and apathy remained below average, however. Other authors (Domínguez-García et al., 2022; Jiménez-Jiménez & Marques, 2018; Serrano-Juárez et al., 2018) have also found improvements in cognitive and behavioural variables after face-to-face and on-line intervention programs for children with neurodevelopmental disorders and poor executive functions.

The limitations we faced included not having behaviour inventories and standardized instruments that would allow us to measure all the domains of social cognition and executive functions for 6–18 years. However, other reviewed studies with pKBS in children (Jha & Ansari, 2010; Lippe et al., 2013) and adults (Cho et al., 2016; Jha et al., 2016) in the last 10 years, have not reported quantitative measures, the magnitude of cognitive and behavioural symptoms in patients with pKBS. Despite the standardized instruments' limitation, these results describe the neuropsychological profile of a girl with right temporal and amygdala damage and neurosurgery resection.

A second limitation, it was not possible to carry out follow-up evaluations to document the evolution of the patient's symptoms in greater detail because of the COVID-19 pandemic; hence, another assessment after 1 year could be helping to determine if the intervention has a long-lasting positive and test-training effect. Another limitation is the lack of a behaviour inventory in Mexico specifically for this population; however, this study describes the cognitive and behavioural profile of a girl with a resection of the entire right temporal lobe, since the other studies with a paediatric sample are associated with encephalitis (Jha & Ansari, 2010).

A final limitation was that there was no formal assessment before neurosurgery; only the clinical data reported by the parents and the ADHD diagnosis were available. It could be that some symptoms of dysexecutive disorder and a borderline index of general capacity were probably already present and possibly secondary to the glioma. However, the postoperative evaluation revealed major social, emotional, externalizing, and dysexecutive problems.

In conclusion, this case seems to be one of the first to describe quantitative results of the neuropsychological profile of a girl who meets the criteria for pKBS derived from a glioma and complete resection of the right temporal lobe. Despite the measurement limitations that this case presents, it was possible to find symptoms of hypermetamorphosis, emotional flattening, mood swings, memory problems, and aggressiveness, which had already been described in paediatric population with pKBS by other authors (Jha & Ansari, 2010; Lippe et al., 2013); however, social interaction problems and a dysexecutive behavioural syndrome were also observed and identified in this case with right temporal damage. Finally, we suggested three possible hypotheses of these findings that could be studied in future research.

AUTHOR CONTRIBUTIONS

Alejandra Estefanía Hernández-Martínez: Conceptualization; data curation; methodology; project administration. **Carlos Alberto Serrano-Juárez:** Conceptualization; data curation; formal analysis; methodology; supervision; writing – original draft; writing – review and editing. **Karen Griselda Barrera-Medellín:** Data curation; methodology; project administration. **Cecilia Inés Ramírez-Quiroga:** Investigation; project administration. **Alma Griselda Ramírez-Reyes:** Conceptualization; supervision; writing – review and editing. **Roberto Casarrubias-Islas:** Project administration; supervision; writing – review and editing. **Belén Prieto-Corona:** Methodology; supervision; writing – review and editing.

CONFLICT OF INTEREST STATEMENT

All authors declare that they have no conflict of interest.

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REFERENCES

- Adolphs, R. (1999). Social cognition and the human brain. *Trends in Cognitive Sciences*, 3(12), 469–479. [https://doi.org/10.1016/S1364-6613\(99\)01399-6](https://doi.org/10.1016/S1364-6613(99)01399-6)
- Adolphs, R. (2001). The neurobiology of social cognition. *Current Opinion in Neurobiology*, 11(2), 231–239. [https://doi.org/10.1016/S0959-4388\(00\)00202-6](https://doi.org/10.1016/S0959-4388(00)00202-6)
- Adolphs, R. (2002). Neural systems for recognizing emotion. *Current Opinion in Neurobiology*, 12(2), 169–177. [https://doi.org/10.1016/S0959-4388\(02\)00301-X](https://doi.org/10.1016/S0959-4388(02)00301-X)
- Adolphs, R. (2003). Cognitive neuroscience of human social behaviour. *Nature Reviews Neuroscience*, 4(3), 165–178. <https://doi.org/10.1038/nrn1056>
- Baeken, C., Marinazzo, D., Van Schuerbeek, P., Wu, G.-R., De Mey, J., Luyckaert, R., & De Raedt, R. (2014). Left and right amygdala—Mediofrontal cortical functional connectivity is differentially modulated by harm avoidance. *PLoS One*, 9(4), e95740. <https://doi.org/10.1371/journal.pone.0095740>
- Bertoux, M., de Paula França Resende, E., & de Souza, L. C. (2018). Klüver & Bucy syndrome: An investigation of social and affective cognition. *Neurocase*, 24(4), 180–187. <https://doi.org/10.1080/13554794.2018.1524497>
- Bhat, P. S., Pardal, P. K., & Das, R. C. (2009). Partial Klüver-Bucy syndrome as a delayed manifestation of head injury. *Industrial Psychiatry Journal*, 18(2), 117–118.
- Brenlla, M. E. (2013). Interpretación del WISC-iv: puntuaciones compuestas Y modelos CHC. *Ciencias Psicológicas*, 7, 183–197.
- Cantú, G. (2016). *Notas para una interpretación clínica de los subtests de comprensión verbal en el WISC-IV. VIII Congreso Internacional de Investigación y Práctica Profesional en Psicología XXIII Jornadas de Investigación XII Encuentro de Investigadores en Psicología del MERCOSUR*.
- Carroll, B. T., Goforth, H. W., & Raimonde, L. A. (2001). Partial Klüver-Bucy syndrome: Two cases. *CNS spectrum*, 6(4), 329–332. <https://doi.org/10.1017/S1092852900022033>
- Cho, A.-R., Lim, Y.-H., Chung, S.-H., Choi, E.-H., & Lim, J. Y. (2016). Bilateral anterior opercular syndrome with partial Klüver-Bucy syndrome in a stroke patient: A case report. *Annals of Rehabilitation Medicine*, 40(3), 540–544. <https://doi.org/10.5535/arm.2016.40.3.540>
- Domínguez-García, C. M., Serrano-Juárez, C. A., Rodríguez-Camacho, M., Moreno-Villagómez, J., Araujo Solís, M. A., & Prieto-Corona, B. (2022). Neuropsychological intervention in attention and visuospatial skills in two patients with Williams syndrome with different types of genetic deletion. *Applied Neuropsychology: Child*, 1–10, 177–186. <https://doi.org/10.1080/21622965.2022.2063723>
- Duff, K. (2012). Evidence-based indicators of neuropsychological change in the individual patient: Relevant concepts and methods. *Archives of Clinical Neuropsychology*, 27(3), 248–261. <https://doi.org/10.1093/arclin/acr120>
- DuPaul, G. J., Power, T. J., & Anastopoulos, A. D. (2018). *Escala de evaluación TDAH-5 para niños y adolescentes*. Manual Moderno.
- Ekman, P., Friesen, W., & Press, C. (1976). *Pictures of facial affect* (Vol. 21). Consulting Psychologists Press.
- Farah, T., Ling, S., Raine, A., Yang, Y., & Schug, R. (2018). Alexithymia and reactive aggression: The role of the amygdala. *Psychiatry Research: Neuroimaging*, 281, 85–91. <https://doi.org/10.1016/j.psychres.2018.09.003>
- Felix-Ortiz, A. C., & Tye, K. M. (2014). Amygdala inputs to the ventral hippocampus bidirectionally modulate social behavior. *The Journal of Neuroscience: The Official Journal of the Society for Neuroscience*, 34(2), 586–595. <https://doi.org/10.1523/JNEUROSCI.4257-13.2014>
- Fernández-Pinto, I., Santamaría, P., Sánchez-Sánchez, F., Carrasco, M., & Del Barrio, V. (2015). *Sistema de Evaluación de Niños y Adolescentes*. SENEA. TEA Ediciones.
- Flanagan, D. P., & Kaufman, A. S. (2009). *Claves para la evaluación con WISC-IV*. Editorial El Manual Moderno.
- García-Anacleto, A., & Salvador-Cruz, J. (2017). Adaptación y validación en población mexicana de la escala BRIEF-P. *Cuadernos de Neuropsicología / Panamerican Journal of Neuropsychology*, 11(3), 42–60.
- Godefroy, O., Azouvi, P., Robert, P., Roussel, M., LeGall, D., Meulemans, T., & Behalf of the Groupe de Réflexion sur l'Évaluation des Fonctions Exécutives Study Group. (2010). Dysexecutive syndrome: Diagnostic criteria and validation study. *Annals of Neurology*, 68(6), 855–864. <https://doi.org/10.1002/ana.22117>
- Godefroy, O., Martinaud, O., Narme, P., Joseph, P.-A., Mosca, C., Lhommée, E., Meulemans, T., Czernecki, V., Bertola, C., Labauge, P., Verny, M., Bellmann, A., Azouvi, P., Bindschaedler, C., Bretault, E., Boutoleau-Bretonniere, C., Robert, P., Lenoir, H., Krier, M., & Roussel, M. (2018). Dysexecutive disorders and their diagnosis: A position paper. *Cortex*, 109, 322–335. <https://doi.org/10.1016/j.cortex.2018.09.026>

- Hernández Galván, A., & Yáñez Téllez, M. G. (2013). Evaluación de la Cognición Social en Adultos Mayores: Presentación de la batería COGSOCA-AM. *Revista Argentina de Clínica Psicológica*, XXII(3), 269–278.
- Hu, R. K., Zuo, Y., Ly, T., Wang, J., Meera, P., Wu, Y. E., & Hong, W. (2021). An amygdala-to-hypothalamus circuit for social reward. *Nature Neuroscience*, 24(6), 831–842. <https://doi.org/10.1038/s41593-021-00828-2>
- Iverson, G. L. (2018). Reliable change index. In J. Kreutzer, J. DeLuca, & B. Caplan (Eds.), *Encyclopedia of clinical neuropsychology* (pp. 1–4). Springer International Publishing. https://doi.org/10.1007/978-3-319-56782-2_1242-3
- Jacobson, N. S., & Truax, P. (1991). Clinical significance: A statistical approach to defining meaningful change in psychotherapy research. *Journal of Consulting and Clinical Psychology*, 59, 12–19. <https://doi.org/10.1037/0022-006X.59.1.12>
- Janak, P. H., & Tye, K. M. (2015). From circuits to behaviour in the amygdala. *Nature*, 517(7534), 284–292. <https://doi.org/10.1038/nature14188>
- Jha, K. K., Singh, S. K., Kumar, P., & Arora, C. D. (2016). Partial Kluver-Bucy syndrome secondary to tubercular meningitis. *BMJ Case Reports*, bcr2016215926. <https://doi.org/10.1136/bcr-2016-215926>
- Jha, S., & Ansari, M. K. (2010). Partial Klüver–Bucy syndrome in a patient with acute disseminated encephalomyelitis. *Journal of Clinical Neuroscience*, 17(11), 1436–1438. <https://doi.org/10.1016/j.jocn.2010.03.015>
- Jiménez-Jiménez, S., & Marques, D. F. (2018). Impacto de la intervención neuropsicológica infantil en el desarrollo del sistema ejecutivo. Análisis de un caso. *Avances en Psicología Latinoamericana*, 36, 11–28.
- Juliá-Palacios, N., Boronat, S., Delgado, I., Felipe, A., & Macaya, A. (2018). Pediatric Klüver–Bucy syndrome: Report of two cases and review of the literature. *Neuropediatrics*, 49(2), 104–111. <https://doi.org/10.1055/s-0037-1609036>
- Latchminarine, N., Wahashi, E. A., Amalraj, B., & Abubakr, A. (2022). Transient Kluver-Bucy syndrome as a manifestation of post-temporal lobe seizure: A rare case entity. *Cureus*, 14(11), e31696. <https://doi.org/10.7759/cureus.31696>
- Lezak, M. D., Howieson, D. B., Bigler, E. D., & Tranel, D. (2012). *Neuropsychological assessment* (5a edición ed.). Oxford University Press.
- Lippe, S., Gonin-Flambois, C., & Jambaqué, I. (2013). Chapter 135—The neuropsychology of the Klüver–Bucy syndrome in children. In O. Dulac, M. Lassonde, & H. B. Sarnat (Eds.), *Handbook of clinical neurology* (Vol. 112, pp. 1285–1288). Elsevier. <https://doi.org/10.1016/B978-0-444-52910-7.00051-9>
- Maldonado, M., Fournier del Castillo, M., Martínez Arias, R., González Marqués, J., Espejo-Saavedra Roca, J., & Santamaría, P. (2017). Evaluación Conductual de la Función Ejecutiva.
- Ostrosky, F., Gómez, M. E., Matute, E., Rosselli, M., Ardilla, A., & Pineda, D. (2019). *Neuropsi atención y memoria (3º)*. Manual Moderno.
- Sánchez-Cubillo, I., Tirapu-Ustárrroz, J., & Adrover-Roig, D. (2012). Neuropsicología de la cognición social y la autoconciencia. In J. Tirapu-Ustárrroz, M. Ríos-Lago, A. García Molina, & A. Ardila (Eds.), *Neuropsicología del córtex prefrontal y las funciones ejecutivas*. (pp. 353–390).
- Sánchez-Navarro, J. P., & Román, F. (2004). Amígdala, corteza prefrontal y especialización hemisférica en la experiencia y expresión emocional. *Anales de Psicología/Annals of Psychology*, 20(2), 223–240.
- Serrano-Juárez, C. A. (2020). *Diferencias entre genotipos y fenotipos neuropsicológicos en personas con síndrome de Williams*. Universidad Nacional Autónoma de México.
- Serrano-Juárez, C. A., Prieto-Corona, B., Rodríguez-Camacho, M., Venegas-Vega, C. A., Yáñez-Téllez, M. G., Silva-Pereyra, J., Salgado-Ceballos, H., Arias-Trejo, N., & De León Miranda, M. A. (2021). An exploration of social cognition in children with different degrees of genetic deletion in Williams syndrome. *Journal of Autism and Developmental Disorders*, 51(5), 1695–1704. <https://doi.org/10.1007/s10803-020-04656-4>
- Serrano-Juárez, C. A., Prieto-Corona, D. M. B., & Yáñez-Téllez, M. G. (2018). Intervención Neuropsicológica en un caso de una niña con Síndrome de Williams. *Cuadernos de Neuropsicología/Panamerican Journal of Neuropsychology*, 12(2), 1–13.
- Venta, A., Sharp, C., Patriquin, M., Salas, R., Newlin, E., Curtis, K., Baldwin, P., Fowler, C., & Frueh, B. C. (2018). Amygdala-frontal connectivity predicts internalizing symptom recovery among inpatient adolescents. *Journal of Affective Disorders*, 225, 453–459. <https://doi.org/10.1016/j.jad.2017.08.064>
- Watanabe, N., Bhanji, J. P., Tanabe, H. C., & Delgado, M. R. (2019). Ventromedial prefrontal cortex contributes to performance success by controlling reward-driven arousal representation in amygdala. *NeuroImage*, 202, 116136. <https://doi.org/10.1016/j.neuroimage.2019.116136>
- Wechsler, D. (2008). *Escala de Inteligencia Wechsler para adultos-WAIS IV- (Moreno, G.)*. Manual Moderno.

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