Chapter 68
The Cerebellar Cognitive Affective Syndrome and the Neuropsychiatry of the Cerebellum

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Abstract Cognitive and limbic functions are represented in the cerebellar posterior lobe and vermis. Lesions of these areas produce dysmetria of thought, manifesting as the cerebellar cognitive affective syndrome (CCAS; Schmahmann's syndrome). This is the counterpart to the cerebellar motor syndrome which results from lesions of the motor representation in the cerebellar anterior lobe and lobule VIII. The CCAS is characterized by impairments in executive function, visual spatial processing, linguistic deficits, and regulation of affect. The affective component of the CCAS, conceptualized as the neuropsychiatry of the cerebellum, is grouped according to five major domains: attentional control, emotional control, autism spectrum disorders, psychosis spectrum disorders, and social skill set. Within each of these domains, behaviors may reflect cognitive overshoot or undershoot, akin to the disorder of motor control seen in the cerebellar motor syndrome. This chapter focuses on the behavioral neurology and neuropsychiatry of the cerebellum and emphasizes the clinically relevant manifestations for adults and children with a wide range of cerebellar disorders. Recognition of the CCAS throughout the age spectrum is important for patient care, and it highlights the promise that new insights into cerebellar function hold for novel interventions in patients with neurobehavioral and psychiatric diseases linked to the cerebellum.

Keywords Cognition • Emotion • Limbic • Cerebellar cognitive affective syndrome • Neuropsychiatry • Mutism

The cerebellar cognitive affective syndrome (CCAS; Schmahmann's syndrome) represents a disruption of the cerebellar contribution to distributed neural circuits linking different regions within the cerebellar posterior lobe with cerebral cortical

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association and paralimbic areas that subserve higher order perceptual processing, intellectual function, and emotion (see Chap. 11 for anatomical tract tracing studies in nonhuman primates) (Schmahmann et al. 1997; Schmahmann 2010). Clinical observations of changes in behavior and intellect following cerebellar injury are further supported by resting state functional connectivity MRI showing topographically precise arrangement of cerebellar connections with motor and nonmotor areas of human cerebral cortex, (Buckner et al. 2011) and task based functional MRI of cerebellar engagement in executive functions, spatial tasks, language, and emotional processing (see also Chap. 51 for details) (Stoodley and Schmahmann 2009b; Stoodley et al. 2012).

68.1 The Initial Description

The CCAS was described by Schmahmann and Sherman (1998) in 20 patients with lesions confined to cerebellum. The neurobehavioral observations identified arose from lesions involving predominantly the cerebellar posterior lobe, characterized by clinically relevant deficits in executive function, visual spatial performance, linguistic processing and dysregulation of affect (Table 68.1). The CCAS has since been named eponymously as Schmahmann's syndrome (Manto and Mariën 2015).

Neurobehavioral tests performed as part of the neurological examination showed that 18 patients had problems with executive functions, including poor working memory (in 11 of 16 tested), motor or ideational set shifting (in 16 of 19), and perseveration of actions or drawings (in 16 of 20). Verbal fluency was impaired in 18 patients, presenting as telegraphic speech, occasionally so limited as to resemble mutism.

Table 68.1 Deficits that characterize the cerebellar cognitive affective syndrome

Executive function
Deficient planning, mental flexibility, multi-tasking, abstract reasoning, working memory. Decreased verbal fluency, to the point of telegraphic speech or mutism. Perseverative ideation
Spatial cognition
Visuospatial disintegration with impaired attempts to draw or copy a diagram. Disorganized conceptualization of figures. Impaired visual-spatial memory. Simultanagnosia in some
Linguistic difficulties
Anomia, agrammatic speech, abnormal syntactic structure, abnormal prosody including high pitched, hypophonic whining
Personality change
Aberrant modulation of behavior and personality when posterior lobe lesions involve midline structures. Flattening or blunting of affect alternating or coexistent with disinhibited behaviors such as over-familiarity, flamboyant and impulsive actions, and humorous but inappropriate and flippant comments. Regressive, childlike behaviors and obsessive-compulsive traits. (See neuropsychiatry of the cerebellum, Table 68.2)
The net effect of these disturbances in cognitive functioning is lowering of overall intellectual function.

Decreased verbal fluency was unrelated to dysarthria. Visuospatial disintegration was found in 19 patients, who were disorganized in their sequential approach to drawing and conceptualization of figures (Fig. 68.1, left panel). Four patients had simultanagnosia. Naming was impaired in 13 patients, usually being spared in those with smaller lesions. Six with bilateral acute disease had agrammatic speech, and elements of abnormal syntactic structure were noted in others. Prosody was abnormal in eight patients, with tone of voice characterized by a high pitched, whining, childish and hypophonic quality. Mental arithmetic was deficient in 14 patients. Verbal learning and recall were slightly abnormal in 11, and visual learning and recall were impaired in 4 (of 13 patients tested). Ideational apraxia was evident in two individuals.

Difficulty modulating behavior and personality style was a prominent feature in 15 patients, particularly those with large or bilateral infarcts in the territory of the posterior inferior cerebellar artery, and in a patient with surgical excision of the vermis and paravermian structures. Flattening of affect or disinhibition manifested as overfamiliarity, flamboyant and impulsive actions, and humorous but inappropriate and flippant comments. Behavior was regressive and childlike, and obsessive-compulsive traits were occasionally observed.

Autonomic changes occurred in a patient with stroke involving the fastigial nucleus and paravermian cortex, with spells of hiccupping and coughing precipitating bradycardia and syncope.

Neuropsychological testing confirmed the neurological observations, demonstrating impaired executive function (planning, set-shifting, abstract reasoning, verbal fluency, working memory), often with perseveration, distractibility or inattention; visual-spatial disorganization and impaired visual-spatial memory; personality change with blunting of affect or disinhibited and inappropriate behavior; and difficulties with language production including dysprosodia, agrammatism and mild anomia. The net effect of these disturbances in cognitive abilities was a general lowering of intellectual function. Findings were more pronounced in patients with bilateral and acute disease. Posterior lobe lesions were particularly important in the generation of the syndrome and the vermis was consistently involved in patients with pronounced affective presentations. Patients with stroke improved over time, although executive function remained abnormal. The CCAS was hypothesized to reflect dysmetria of thought, analogous to dysmetria of movements resulting from damage to the motor cerebellum in the anterior lobe.

68.2 Subsequent Reports

The principal features and clinical relevance of the CCAS were confirmed in patients with cerebellar stroke or hemorrhage. Findings include problems with frontal/executive function such as impaired cognitive control, multitasking, mental flexibility, and working memory (reverse digit span and n-back task); deficits in visuospatial planning, visuomotor tasks and visual memory; anomia, irregularity of speech, agrammatism, dysprosodia, acquired dyslexia, and impaired verbal fluency (phonemic > semantic), as well as metalinguistic deficits – problems with metaphor,

inference, ambiguity, and verbal expression of complex thoughts; (Güell et al. 2015) and apathy, disinhibition, and impaired emotional intelligence. Impaired focused and sustained attention, delayed recall of verbal or visual information, facial agnosia, amusia, and temporal disorientation, and limb kinetic apraxia are also reported (Schmahmann and Sherman 1998; Malm et al. 1998; Leggio et al. 2000; Neau et al. 2000;Exner et al. 2004; Gottwald et al. 2004; Hoffmann and Schmitt 2004; Kalashnikova et al. 2005; Hokkanen et al. 2006; Ravizza et al. 2006; Richter et al. 2007; Ziemus et al. 2007;Hoffmann and Cases 2008;Manes et al. 2009; Stoodley and Schmahmann 2009a; Schweizer et al. 2010;Alexander et al. 2011; Tedesco et al. 2011).

68.3 The Cerebellar Cognitive Affective Syndrome in Children

Levisohn et al. (2000) first studied cognition in children who underwent resection of cerebellar tumors without receiving radiation therapy or methotrexate which can lead to poor cognitive outcome. The cohort consisted of 19 children, ages 3–14 with medulloblastoma, astrocytoma, and ependymoma. Problems were noted with attention and executive function as evidenced by deficits in digit span (57 % of the 14 tested), sequencing, and planning. Establishing and maintaining set was hampered by perseveration. Deficits in expressive language, present in 58 % of the cohort, included brief responses, lack of elaboration, reluctance to engage in conversation, long response latencies, and word finding difficulties. Language initiation and word finding difficulties occurred in the context of average scores on verbal tests. Confrontation naming deficits were ameliorated with phonemic cues. Many demonstrated difficulty with initiation of responses and problem-solving strategies. Visual spatial difficulties were present in 37 %, characterized by impaired planning and organizational aspects of tasks (Fig. 68.1, right panel). Verbal memory was impaired in 33 % of 15 children tested, particularly for unstructured recall of information. Story retrieval improved with multiple-choice prompts. There was failure to organize verbal or visual-spatial material for encoding, which impacted retrieval of information. Impaired regulation of affect was particularly evident when cerebellar damage included the vermis, manifesting as irritability, impulsivity, disinhibition, and lability of affect with poor attentional and behavioral modulation.

The CCAS in children has also been confirmed by others. Deficits include executive dysfunction with impaired planning, sequencing, mental flexibility and hypothesis generation and testing, visual-spatial function, expressive language, and verbal memory (Karatekin et al. 2000; Grill et al. 2004; Turkel et al. 2004; Berger et al. 2005; Ronning et al. 2005; Vaquero et al. 2008). Impairments in verbal intelligence, auditory sequential memory, and language follow right-sided tumors; deficient nonverbal tasks including spatial and visual sequential memory and impaired prosody after left cerebellar hemisphere tumors (Riva and Giorgi 2000).

The affective component of the CCAS in children includes mood disturbances, pathologic laughing and crying (Kossorotoff et al. 2010), disinhibition, impulsivity

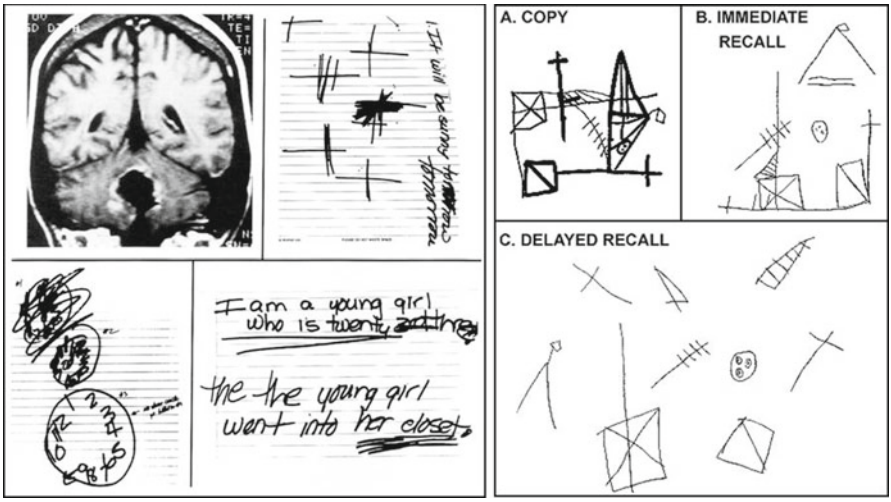

Fig. 68.1 *Left panel*: T1-weighted coronal MRI of the brain of a patient showing the site of excision of a ganglioglioma, and her responses when asked to bisect a line, draw a clock and write a sentence (From Schmahmann and Sherman 1998) *Right panel*: drawings of the Rey-Osterrieth figure by a 6-year old boy 11 months following resection of a left cerebellar cystic astrocytoma (From Levisohn et al. 2000)

and irritability (Maryniak and Roszkowski 2005). dysphoria, inattention and irritability (Turkel et al. 2004), anxiety and aggression (Richter et al. 2005), hyperspontaneous and disinhibited behavior, and flattened affect with apathy and poverty of spontaneous movement (Aarsen et al. 2004). Emotional lability may be marked, with rapid fluctuation of emotional expression gravitating between irritability with inconsolable crying and agitation, to giggling and easy distractibility. Surgically induced vermal lesions may result in autistic features such as avoidance of physical and eye contact, complex repetitive and rhythmic rocking movements, stereotyped linguistic utterances, and lack of empathy (Riva and Giorgi 2000). Attention deficit disorder, addiction, anorexia, uncontrolled temper tantrums and phobias are also reported (Steinlin et al. 2003).

68.4 Postoperative Pediatric Cerebellar Mutism Syndrome (CMS)

More than half the children in Levisohn et al. (2000) with surgical damage to the vermis developed mutism or markedly reduced speech 1–2 days after the surgery, often with hypotonia and oropharyngeal dysfunction/dysphagia. The CMS is frequently accompanied by the cerebellar motor syndrome, cerebellar cognitive affective syndrome and brain stem dysfunction including long tract signs and cranial neuropathies. The mutism is transient, but recovery from CMS may be prolonged,

speech and language may not return to normal, and the motor dysfunction and CCAS often persist (Gudrunardottir 2016). CMS has had other names, including posterior fossa syndrome (Daly and Love 1958; Wisoff and Epstein 1984;Rekate et al. 1985; Dietze and Mickle 1990; Catsman-Berrevoets et al. 1992; van Dongen et al. 1994; Kingma et al. 1994; Kirk et al. 1995; Pollack et al. 1995; Schmahmann and Sherman 1997, 1998; Schmahmann 1998; Levisohn et al. 2000; Sadeh and Cohen 2001).

68.5 Neuropsychiatry of the Cerebellum; the Affective Component of the CCAS

Emotional dysregulation occurs when lesions involve the limbic cerebellum – the vermis and fastigial nucleus (Schmahmann 1991; Schmahmann and Sherman 1998; Richter et al. 2007), with altered regulation of mood and personality, psychotic thinking, and behaviors that meet criteria for diagnoses of attention deficit hyperactivity disorder, obsessive compulsive disorder, depression, bipolar disorder, disorders on the autism spectrum, atypical psychosis, anxiety and panic disorder (Schmahmann et al. 2007). These manifestations segregate into five neuropsychiatric domains – attentional control, emotional control, social skill set, autism spectrum disorders, and psychosis spectrum disorders (Table 68.2) (Schmahmann et al. 2007). The behaviors are conceptualized as either excessive (hypermetric) or reduced (hypometric) responses to the external or internal environment. Deficits in social skill set likely reflect altered social cognition, defined as the set of mental processes required to understand, generate and regulate social behavior (Garrard et al. 2008; Sokolovsky et al. 2010; D'Agata et al. 2011; Hoche et al. 2015). Prominent affective changes are seen, for example in opsoclonus myoclonus syndrome which produces mood changes and inconsolable irritability with lability, aggression and night terrors, dysphoric mood, disinhibition and poor affect regulation, disruptive behaviors and temper tantrums as well as cognitive and language impairments (Turkel et al. 2006; Gorman 2010). Pathological laughing and crying occurs after stroke in the pontocerebellar circuit (Tei and Sakamoto 1997; Gondim et al. 2001; Parvizi et al. 2001; Schmahmann et al. 2004a;Jawaid et al. 2008), in post-infectious cerebellitis (Dimova et al. 2009), and in the cerebellar form of multiple system atrophy (Parvizi et al. 2007).

68.6 Cognition in Ataxic Disorders

Cognitive changes occur in most of the spinocerebellar ataxias (SCAs) (Geschwind 1999). Neuropathology in many SCAs involves brainstem, basal ganglia, thalamus, and sometimes the cerebral cortex, so the cerebellar lesion may not be solely

Table 68.2 Neuropsychiatric manifestations in patients with cerebellar disorders, arranged according to major domains, each with positive and negative symptoms

	Positive (exaggerated) symptoms	Negative (diminished) symptoms
Attentional control	Inattentiveness	Ruminativeness
	Distractibility	Perseveration
	Hyperactivity	Difficulty shifting focus of attention
	Compulsive and ritualistic behaviors	Obsessional thoughts
Emotional control	Impulsiveness, disinhibition	Anergy, anhedonia
	Lability, unpredictability	Sadness, hopelessness
	Incongruous feelings, pathological laughing/crying	Dysphoria
	Anxiety, agitation, panic	Depression
Autism spectrum	Stereotypical behaviors	Avoidant behaviors, tactile defensiveness
	Self stimulation behaviors	Easy sensory overload
Psychosis spectrum	Illogical thought	Lack of empathy
	Paranoia	Muted affect, emotional blunting
	Hallucinations	Apathy
Social skill set	Anger, aggression	Passivity, immaturity, childishness
	Irritability	Difficulty with social cues and interactions
	Overly territorial	Unawareness of social boundaries
	Oppositional behavior	Overly gullible and trusting

From Schmahmann et al. 2007

responsible for the cognitive deficits. CCAS features include mild generalized cognitive impairment, impaired executive functions, deficits in verbal short-term memory (Kish et al. 1988; Burk et al. 2001; Tedesco et al. 2011), visual and verbal attention, verbal fluency, planning and strategy (Zawacki et al. 2002; Braga-Neto et al. 2011), concentration problems, impaired conceptual reasoning, and emotional instability and impulsivity (Gambardella et al. 1998; Storey et al. 1999; Lilja et al. 2005; Suenaga et al. 2008; Cooper et al. 2010; Sokolovsky et al. 2010; Horton et al. 2011). Developmental cognitive impairment occurs in SCA 13 (Herman-Bert et al. 2000), and dementia in SCA 17 (Koide et al. 1999; Nakamura et al. 2001) and SCA 21 (White et al. 2000). The cognitive profile in Friedreich's Ataxia is variable – some report normal cognition (Botez-Marquard and Botez 1993) whereas others describe impaired visual-perceptual and visual-constructive abilities, slowed information processing, decreased attention, reduced verbal span, deficits in letter fluency, and impaired acquisition and consolidation of verbal information (Devos et al. 2001; Wollmann et al. 2002), as well as irritability, poor impulsive control, or blunting of affect (Mantovan et al. 2006).

68.7 Cerebellar Lesions Impair Cognition in the Developing Brain

The cerebellum has a protracted developmental trajectory, and is vulnerable to environmental influences (Limperopoulos et al. 2005a). Adolescents born very pre-term (<33 weeks gestation) have reduced cerebellar volumes, and deficits in executive, visual-spatial and language skills including impaired reading (Allin et al. 2001). Malformations, agenesis and hypoplasia of the cerebellum are associated with motor, linguistic, intellectual and emotional manifestations (Chheda et al. 2002; Richter et al. 2005; Gross-Tsur et al. 2006; Tavano et al. 2007) including delayed milestones, mild motor impairments, and intellectual handicap (Gardner et al. 2001). Children with cerebellar hypoplasia and non-progressive cerebellar ataxia may have the developmental CCAS with decreased general intelligence scores, alertness and sustained attention, and difficulties with visuoconstructive tasks and visual perception (Steinlin et al. 1998). Autistic features and speech delay, together with ataxia, hypotonia, and ocular signs correctly predict 86 % of children with cerebellar hypoplasia (Wassmer et al. 2003), and autistic features are seen in more than 40 % of preterm children who suffered prenatal cerebellar hemorrhage (Limperopoulos et al. 2007).

Children with complex malformations of the cerebellum may have cognitive and emotional deficits in addition to the motor disorders, as seen in rhombencephalosynapsis, Joubert syndrome, some cases of Dandy Walker syndrome, and Chiari malformations, among others.

68.8 Clinical Implications

Clinicians should recognize the CCAS/Schmahmann syndrome because it can be assessed with mental state tests of cognitive and emotional domains, and patients and families can be informed about nonmotor aspects of cerebellar function which may have meaningful long-term impact. Current therapeutic interventions for CCAS include behavioral measures, psychopharmacology, and emerging therapies such as brain modulation, to improve the lives of patients with cerebellar disorders.

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