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Topical Review

Working Memory Impairments in Cerebellar Disorders of Childhood

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ABSTRACT

The cerebellum is a crucial center for motor control and integration. Increasing evidence supports the notion that the cerebellum is also involved in nonmotor functions. Along these lines, multiple cerebellar disorders of childhood and adulthood are associated with behavioral and cognitive symptoms, including impairments in memory. One form of memory commonly affected in cerebellar disorders is working memory, which uses attention to manipulate information that is immediately available to execute cognitive tasks. This article reviews the literature illustrating that working memory impairments are frequently observed in acquired, congenital, and genetic/developmental cerebellar disorders of childhood. Functional neuroimaging studies demonstrate that working memory tasks engage many posterior regions of the cerebellar hemispheres and vermis. Thus, the cerebellum acts as one important node in the working memory circuit, and when the cerebellum is involved in childhood disorders, deficits in working memory commonly occur.

Introduction

Despite only occupying 10% of the intracranial space, over half of the brain's neurons are derived from the cerebellum.[1] Not surprisingly, central nervous system (CNS) pathology localized to the cerebellum is extensive. To aid in their classification, pediatric cerebellar disorders can be considered acquired, congenital, or genetic/developmental (Fig 1). Acquired conditions include those that are oncologic, vascular (e.g., hemorrhage or infarct limited to the posterior fossa), infectious (e.g., acute cerebellitis, acute necrotizing encephalopathy of childhood), or parainfectious (e.g., postinfectious acute cerebellar ataxia, acute disseminated encephalomyelitis). Congenital conditions are often associated with malformations (e.g., Joubert syndrome, rhombencephalosynapsis, vermian hypoplasia, mega cisterna magna, Chiari I malformation). Genetic/developmental disorders can cause conditions that primarily involve the cerebellum (e.g., ataxia telangiectasia [AT], spinocerebellar ataxias [SCAs]) or conditions with a complex phenotype that also includes cerebellar abnormalities (e.g., fragile X syndrome [FXS], Williams syndrome, chromosome 22q11.2 deletion syndrome, autism spectrum disorder, attention-deficit/hyperactivity disorder). Fitting with the classic view that the cerebellum is crucial for the control of movements, many of these conditions are associated with significant motor disability. Increasing evidence, however, has demonstrated that the cerebellum also contributes to nonmotor functions.[2,3]

Many nonmotor phenotypes are associated with cerebellar disorders. In adults, deficits in executive functioning, language, and affect are often present when the cerebellum is involved. These impairments comprise Schmahmann and Sherman's cerebellar cognitive affective syndrome (CCAS).[4] The literature extending CCAS to children is limited despite evidence that numerous nonmotor symptoms are present in pediatric cerebellar disorders.[5,6] For example, emotional dysregulation and difficulty with maintaining attention are some of the most commonly reported symptoms.[7] Language impairments are also often observed,[8] most dramatically as transient "cerebellar mutism" after surgical removal of posterior fossa masses.[9] Sociability deficits have also been reported.[10] Along these lines, the most common neuropathological findings in autism spectrum disorder are decreased volumes of the cerebellar vermis and decreased numbers of cerebellar Purkinje cells.[11]

Perhaps most surprising are the deficits in memory. Working memory is one form of memory commonly and reliably affected in adults with CCAS.[12] Working memory uses attention to manipulate immediately accessible information to execute cognitive tasks and form associations that promote mnemonic processing.[13,14] That

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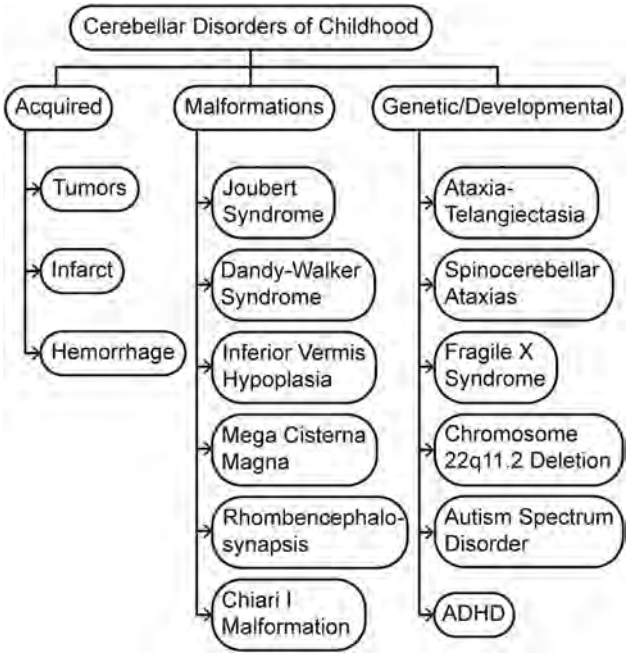

FIGURE 1. Pediatric cerebellar disorders are associated with nonmotor deficits, including working memory impairments, and can be acquired, congenital, or genetic/developmental. Although in no way comprehensive, this chart highlights some of the cerebellar disorders of childhood that are associated with difficulties in encoding memories. Acquired conditions are restricted in space and time, but congenital malformations of the posterior fossa can be associated with malformations of other brain regions and the effects of genetic/developmental changes are not necessarily restricted to the cerebellum, thereby complicating the ability to infer functional implications of the cerebellar structural differences. ADHD, attention-deficit/hyperactivity disorder.

working memory may be affected in cerebellar disease aligns with multiple functional imaging studies confirming that the cerebellum plays a prominent role in the brain networks engaged during working memory. For example, in multiple meta-analyses of functional magnetic resonance imaging (fMRI) studies, verbal working memory activates the posterior cerebellar hemispheres.[15,16] Moreover, transcranial magnetic stimulation of the cerebellum interferes with proper working memory function.[17,18] Diffusion tensor imaging studies confirmed that the performance on working memory tasks is related to the integrity of the cerebellar peduncles as well as the tract connecting the cerebellum to the pons.[19,20] Although working memory deficits are reported in children with confirmed cerebellar diseases during neurodevelopmental and neuropsychological evaluations, a review of the pediatric literature is lacking. As such, this report aims to present a comprehensive overview of working memory impairments appreciated in cerebellar disorders of childhood while emphasizing the involved cerebellar regions (e.g., hemispheres, vermis, or deep nuclei).

Methods

Data for this review were identified through searches of MEDLINE and PubMed and of references from relevant articles completed between October 1, 2019, and January 8, 2020, using the terms cerebellum, cerebellar, and working memory. Included articles were systematic reviews, meta-analyses, topical reviews, original research reports, case reports, and case series. Relevant clinical articles included those that were done in children. Adult studies were included only if the reported conditions were expected to have cerebellar effects during childhood (e.g., malformations and genetic variants). Clinical studies that did not meet these criteria were excluded.

Working memory in acquired cerebellar disorders of childhood

One advantage of studying acquired conditions of the cerebellum is that the pathology and symptoms are spatially and temporally restricted, respectively. Tumors and vascular insults of the cerebellum comprise the two most studied acquired conditions associated with working memory impairments (Table 1). Regarding the former, nearly half of all brain tumors in children occur in the cerebellum. In one of the first large studies assessing working memory in children with cerebellar tumors, over half had impairments specifically in digit span testing.[21] Multiple studies have confirmed this robust working memory impairment in separate cohorts of children with cerebellar tumors.[22,23] Kirschen and colleagues found that verbal, but not visual, digit span was impaired in children following resection of pilocystic astrocytomas and that the degree of this deficit was greatest when lobule VIII of the left cerebellar hemisphere was involved.[24] Children with astrocytomas localized to the right cerebellar hemisphere as well as children with medulloblastomas of the inferior cerebellar vermis exhibited impairments in another form of verbal working memory, sequential memory.[9] In line with these observations that verbal working memory is impaired, many studies reported deficits in story comprehension.[9,21,23] Thus, tests in patients with cerebellar tumors have uncovered many impairments when patients are asked to use working memory to recall presented sequences.

Focal cerebellar strokes are also helpful to assess how the cerebellum may contribute to working memory. Working memory impairments were evident in a large prospective series of young adults who suffered ischemic infarcts of the cerebellum,[26] whereas working memory impairments were not appreciated in another study in older adults.[27] Unfortunately, studies in younger children are limited. Steinlin described one child with a pure left cerebellar infarct with short-term memory difficulties,[25] although formal neuropsychological results were not reported and thus it is not clear what forms of working memory were affected. Cerebellar hemorrhage associated with prematurity is associated with impairments in "cognitive development,"[28-30] as assessed by neurodevelopmental tests such as the Batelle Developmental Inventory and Bayley Scales of Infant and Toddler Development, second edition. Prematurity is associated with impairments in multiple measures of both verbal and spatial working memory when tested in children or adolescents.[31-33] Greater working memory impairments were present when tested by reverse digit span in school-aged children born prematurely with lower birth weights.[34] Using diffuse tensor imaging, Shany and colleagues demonstrated that both the volume and fractional anisotropy of the right middle cerebellar peduncle correlated with working memory scores in children with a history of prematurity.[34] Moreover, in school-aged children with a history of prematurity, working memory scores on the Weschler Intelligence Scale for Children, fourth edition, correlated with volumes of crus I, crus II, and lobule VIIB of the cerebellar hemispheres.[35] However, no study has specifically tested if cerebellar intraparenchymal hemorrhage associated with prematurity is associated with working memory impairments later in life.

Working memory in cerebellar malformations

In contrast to acquired cerebellar disorders, congenital malformations are present at birth and thus are also present throughout development (see Fig 2 for examples of some of the malformations

TABLE 1
Working Memory Impairments in Acquired Cerebellar Disorders of Childhood

Diagnosis	Study	N	Cerebellar Abnormality Associated with Memory Impairment	Working Memory Impairment
Tumors	Levisohn et al[21]	19	Medulloblastoma, astrocytoma, ependymoma Localization not specified	Digit span Story comprehension
	Scott et al[22]	7	Medulloblastoma, astrocytoma, glioma, ependymoma Bilateral hemispheres	Digit span
	Steinlin et al[23]	24	Astrocytoma, choroid plexus papilloma, gangliocytoma, hemangioblastoma Vermis and left hemisphere	Digit span RVDLT
	Kirschen et al[24]	12	Astrocytoma Left hemisphere lobule VIII and vermis lobules VII and VIII	Digit span Immediate and delayed recall of letters and words
	Riva & Giorgi[9]	26	Medulloblastoma, astrocytoma Vermis and bilateral hemispheres	Sequential memory
Infarct	Steinlin[25]	1	Pure cerebellar infarction Localization not specified	Working memory test not specified

Abbreviation:
RVDLT = Rey Visual Design Learning Test

discussed here). Given that many cerebellar malformations are associated with genetic mutations and/or abnormal neuroembryology, potential extracerebellar CNS effects make it difficult to draw conclusions about function. Nonetheless, in conjunction with those clinical observations appreciated in acquired conditions, impairments in working memory are also present in multiple cerebellar malformations (Table 2). Distinctive hypoplasia of the cerebellum and brainstem characterizes Joubert syndrome, a posterior fossa malformation syndrome noted by the "molar tooth sign" on brain magnetic resonance imaging.[46] In a recent large, multicenter prospective study, Bulgheroni and colleagues reported weaknesses in arithmetic skills and coding subtests, and these

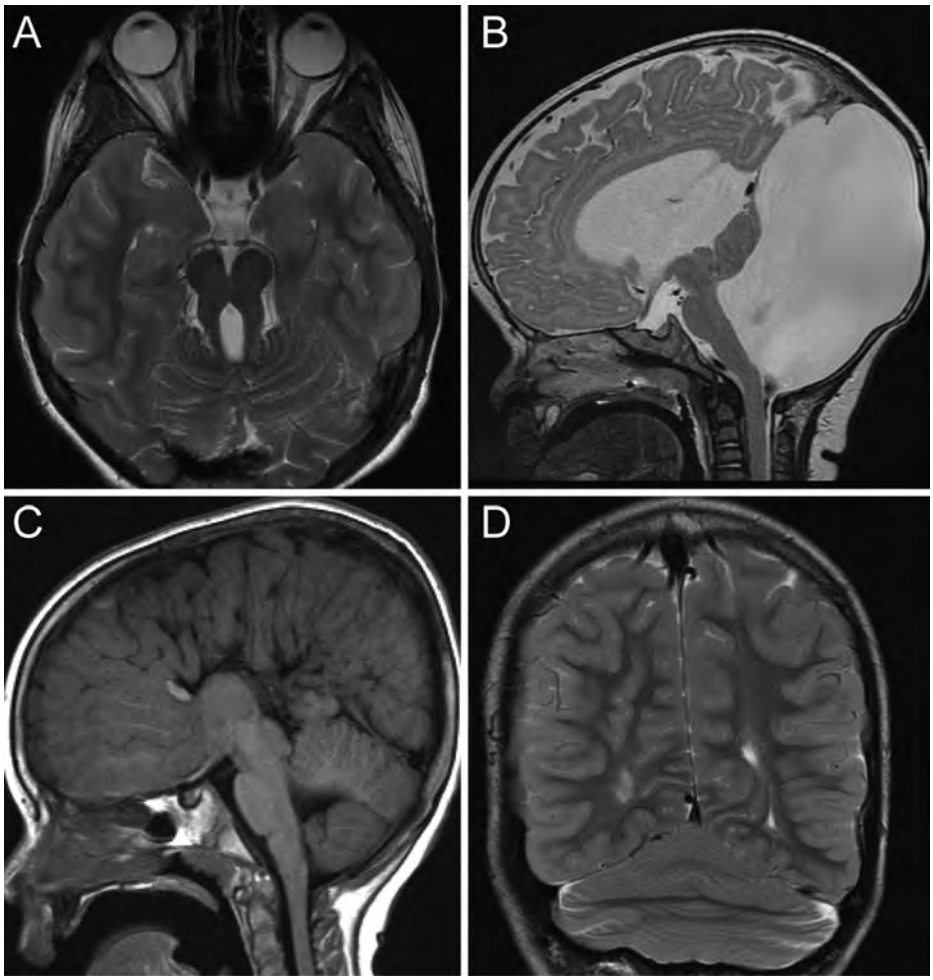

FIGURE 2. Impairments in working memory are often present in malformations involving the cerebellar vermis. (A) Joubert syndrome is a ciliopathy characterized by the "molar tooth sign" on brain magnetic resonance imaging that involves midbrain and cerebellar vermis hypoplasia or agenesis. (B) The Dandy-Walker syndrome is the most common malformation of the posterior fossa that consists of hypoplasia of the cerebellar vermis, dilatation of the fourth ventricle, and enlargement of the posterior fossa. (C) As its name suggests, hypoplasia of the inferior vermis involves partial absence of the inferior lobules of the cerebellar vermis. (D) Rhombencephalosynapsis is a rare malformation that involves fusion of both cerebellar hemispheres and dentate nuclei with absence of the midline vermis.

scores correlated significantly with those of digit span, consistent with impairments in working memory.[36] Impairments in digit span and delayed recall of stories were also present in another subset of patients.[37] A separate study compared neuropsychological testing results from patients with isolated vermian hypoplasia with those with Joubert syndrome. Visuospatial working memory and arithmetic deficits were present in both groups, whereas verbal working memory (measured by digit span and memory of sentences) was significantly more impaired in the Joubert syndrome group.[38] Another prominent malformation of the posterior fossa is evident in the Dandy-Walker syndrome, which involves enlargement of the fourth ventricle, absence or hypoplasia of the cerebellar vermis, and sometimes hypoplasia of the cerebellar hemispheres. Although large clinical studies are lacking, working memory impairments have been reported in case reports of patients with Dandy-Walker syndrome.[39,40] For example, Prigatano and colleagues reported a working memory impairment in the first trial of the Rey Auditory Verbal Learning Test that improved significantly after third ventriculostomy.[39] Working memory impairments are thus appreciated in multiple conditions in which the cerebellum is malformed.

Together, the above-mentioned results raise an interesting question: does malformation of the cerebellar vermis contribute to working memory impairments in children? In a prospective study of 20 children with isolated hypoplasia of the inferior cerebellar vermis that was radiologically confirmed with prenatal ultrasound, performance on the Weschler Intelligence Scale for Children Working Memory Index was comparable to those of control subjects.[47] Mega cisterna magna is another malformation defined by a large (>10 mm) midline infracerebellar cerebrospinal fluid space. After reviewing over 19,000 consecutive cases, Zimmer et al.[41] confirmed that isolated mega cisterna magna was present in 49 patients. Patients who completed a neuropsychological evaluation performed significantly worse than controls on a test of verbal working memory (Rey Auditory Verbal Learning Test). In addition, patients performed worse on digit span, and this effect closely approached statistical significance ($P = 0.06$). Agenesis of the cerebellar vermis with resultant fusion of the cerebellar hemispheres and dentate nuclei characterizes rhombencephalosynapsis. Case reports of adults and children with rhombencephalosynapsis have shown inconsistent results, with working memory impairments appreciated in one child[42] but no working memory impairments present in two other patients.[42,48] Unfortunately, larger clinical studies of children with rhombencephalosynapsis have not yet been performed. Finally, Chiari I malformation consists of downward herniation of the cerebellar tonsils, which are continuous with the uvula of the cerebellar vermis, through the foramen magnum. In a large study of over 600 adults with Chiari I malformation, Allen and colleagues reported working memory impairments during the immediate recall test of the Rey Auditory Verbal Learning Task.[43] However, in their systematic review, Rogers et al.[49] noted that deficits in working memory are inconsistently appreciated in both adults and children with Chiari I malformation. Given the varying results appreciated across these studies, the precise contributions of vermian involvement to working memory performance in childhood cerebellar disorders remain unclear.

Working memory in genetic/developmental conditions associated with cerebellar abnormalities

Many genetic syndromes are associated with both cerebellar abnormalities and deficits in working memory (Table 2). AT, for example, is an autosomal recessive condition secondary to mutations in the *ATM* gene that is associated with neurodegeneration of the cerebellar vermis and hemispheres, immunodeficiency, and telangiectasias.[50] To help address the potential confounding issue that genetic conditions like AT can involve extracerebellar CNS structures, Hoche and colleagues divided their cohort into two subgroups: one with only cerebellar signs and another with both cerebellar and extracerebellar signs on neurological examination. Multiple measures of working memory were impaired in both groups, and the impairments in the group with extracerebellar signs were significantly more robust than those in the cerebellum-only group.[44] Like AT, the SCAs are a group of progressive genetic conditions involving the cerebellum. In SCA 6, the degree of the

TABLE 2
Working Memory Impairments in Congenital and Genetic Cerebellar Conditions

Diagnosis	Study	N	Working Memory Impairment
Joubert syndrome	Bulgheroni et al[36]	49	Digit span Verbal comprehension Arithmetic Coding
	Fennell et al[37]	6	Digit span Story recall
	Tavano & Borgatti[38]	8	Digit span Story recall Arithmetic Visuospatial memory
Cerebellar vermis hypoplasia	Tavano & Borgatti[38]	16	Arithmetic Visuospatial memory
Dandy-Walker syndrome	Prigatano et al[39]	1	RAVLT, BVMT
	Graf et al[40]	1	Working memory test not specified
Mega cisterna magna	Zimmer et al[41]	18	Digit span RAVLT
Rhombencephalosynapsis	Poretti et al[42]	1	Digit span
Chiari I malformation	Allen et al[43]	638	RAVLT
Ataxia telangiectasia	Hoche et al[44]	22	Digit span Verbal comprehension Working memory IQ
Spinocerebellar ataxias	Ma et al[45]	18	Digit span Immediate and delayed recall of words

Abbreviations:
BVMT = Brief Visuospatial Memory Test
IQ = Intelligence quotient
RAVLT = Rey Auditory Verbal Learning Test

impairment in working memory correlated with the duration of disease.[51] Moreover, using voxel-based morphometry, Cooper et al.[52] demonstrated significant correlations between working memory performance and gray matter density in multiple lobules of the inferior cerebellar hemispheres. In SCAs 1, 2, and 3, immediate and delayed recall of words are both impaired, and as expected from the SCA 6 studies, the degree of working memory deficits in these patients correlated significantly with the patients' degree of ataxia.[45] Thus, even though extracerebellar brain regions are affected in both AT and SCA, cerebellar abnormalities are associated with the working memory impairments observed in both conditions.

The cerebellum is also affected in many neurogenetic disorders associated with working memory impairments (Table 3). Most notably, extensive clinical and preclinical research has been done in FXS, the most common form of inherited intellectual disability. In the *Fmr1* knockout (KO) mouse model of FXS, the shape, number, and synaptic signaling of hippocampal dendritic spines are all abnormal.[76-80] Like in the hippocampus, the shape of Purkinje cell dendritic spines and the plasticity of the parallel fiber synapses that innervate these spines are both abnormal in *Fmr1* KO cerebellar vermis.[53] Cerebellar learning is also affected: classical delayed eyeblink conditioning, a task that requires the cerebellum, is impaired in both *Fmr1* KOs and patients with FXS.[53] Notably, working memory is impaired in patients with FXS through many different tests, including forward and reverse digit span, visuospatial tasks asking subjects to recall the location of objects, and nonword repetition tasks.[55,56] Finally, in patients with FXS, the volume of the inferior vermis is decreased compared with controls, and the size of the inferior cerebellar vermis is also correlated with performance on a measure of working memory.[54] Although further preclinical work is needed to test the contributions of the cerebellum to working memory impairments in the *Fmr1* KOs, such as through selective knockdown of *Fmr1* in the cerebellum, as has been done in the mouse model of tuberous sclerosis,[81] data from patients suggest that the cerebellar vermis contributes to the condition's cognitive phenotype.

FXS is a common single-gene cause of working memory deficits, but impairments in working memory are present in larger genetic changes (e.g., duplications or deletions) as well. In Williams syndrome, a rare genetic disorder involving deletions of genes on chromosome 7, the volume of the inferior vermis is enlarged.[57] Multiple groups demonstrated impairments in both verbal and spatial working memory tasks in Williams syndrome.[58-60] Chromosome 22q11.2 deletion syndrome is a neurodevelopmental disability that includes DiGeorge and velocardiofacial syndromes. The neuropsychological profile of chromosome 22q11.2 deletion syndrome is complex, but both verbal and visuospatial working memory impairments are present.[68-72] Moreover, some of the most consistently reported brain abnormalities in chromosome 22q11.2 deletion syndrome are reductions in the volumes of the cerebellar hemispheres and vermis.[61-67] Although suggestive, drawing conclusions about the functional contributions of these cerebellar findings is challenging given how heterogeneous and complex these genetic conditions are.

Additional neurodevelopmental disabilities are also associated with cerebellar abnormalities and working memory impairments. For example, decreased volumes of the cerebellar vermis, decreased density of cerebellar Purkinje cells, and robust deficits in working memory are frequently reported in autism spectrum disorder.[11,82] Using fMRI, Rahko and colleagues showed that children with autism spectrum disorder, compared with controls, exhibit decreased activation of the bilateral posterior lobes of the cerebellar hemispheres and lobules VIII and IX of the cerebellar vermis when asked to recall each time an object appeared in the same place as the predetermined preferred location. However, when the working memory task was made slightly more complex—namely, subjects were asked to recall the object's location from two trials beforehand—there were no group differences in the activation of the cerebellum.[73] A similar study was done in children with attention-deficit/hyperactivity disorder, where volumes of the posterior cerebellar hemispheres and inferior vermis are decreased.[74] Massat and colleagues showed decreased activation of lobule VIIA and crus I of the bilateral cerebellar hemispheres during both the immediate and delayed recall tasks.[82] Although contributing to the body of literature implicating the cerebellum in working memory impairments, the above-mentioned studies highlight the growing need for future studies to further parcellate the cerebellum's contributions to attention and working memory, given these processes can undoubtedly influence one another.[83]

Cerebellar correlates of working memory

Impairments in working memory have been extensively described in adults with cerebellar disease and are part of CCAS.[4] As discussed in this review, working memory is a commonly affected component of pediatric CCAS and one of the most common forms of memory that is frequently abnormal across multiple cerebellar conditions of childhood. Working memory employs attention to temporarily store and manipulate both verbal and nonverbal information to achieve a task; as such, working memory contributes to learning.[13,14] Thus, impairments in working memory in cerebellar conditions of childhood may have important implications during developmental demands that normally rely on the

TABLE 3
Cerebellar Abnormalities and Working Memory Impairments in Other Neurodevelopmental Disabilities

Diagnosis	Cerebellar Findings	Working Memory Impairments
Fragile X syndrome	Abnormal Purkinje cell dendritic spines[53,*] Abnormal parallel fiber synaptic plasticity[53,*] Decreased volume of inferior vermis[54] Impaired cerebellar learning[53]	Digit span[55] Nonword repetition[55] Visuospatial working memory[56]
Williams syndrome	Increased volume of vermis[57]	Immediate and delayed recall of objects,[58] letters,[59] locations,[59] and matrices[60]
22q11.2 deletion syndrome	Decreased volume of cerebellum[61-64] Decreased volume of vermis[65-67]	Arithmetic[68] Visuospatial working memory[68-70] Immediate and delayed recall of location[70] Verbal working memory[71,72]
Autism spectrum disorder	Vermian hypoplasia and decreased Purkinje cell density in cerebellar hemispheres[11]	N-back task performance correlates with activation of posterior hemispheres and inferior vermis[73]
Attention-deficit/ hyperactivity disorder	Decreased volumes of cerebellar hemispheres and inferior vermis[74]	N-back task performance correlates with activation of posterior hemispheres[75]

[*] Demonstrated in Fmr1 knockout mice.

manipulation of sensory stimuli and experiences to encode long-term memories and perform executive functions.

Many neuroanatomical substrates have been implicated in working memory, including the cerebellum, but it is not yet clear how the entire system works together. Functional imaging and lesion studies have uncovered roles of the dorsolateral prefrontal cortex, parietal lobe, temporal lobe, and hippocampus in working memory.[84-86] In addition to the aforementioned pediatric studies, the cerebellum's involvement in this network is supported by evidence that damage to the cerebellum in adulthood impairs working memory performance.[87-89] Moreover, with fMRI, multiple studies have demonstrated that the cerebellum is activated during working memory tasks. Hayter and colleagues used the Paced Serial Addition Task, where subjects added a number (n) to the number presented before it (n − 1), and as numbers were serially presented, subjects needed to disregard their recent response as well as the n − 2 stimulus. Functional MRI results yielded significant activations of bilateral paravermal lobules VI and VII.[90] Kuper and colleagues studied the cerebellar correlates of working memory tasks that were increasingly more difficult. To do so, they utilized the n-back working memory task in which subjects were asked to recall what was presented to them with distractors presented between the original and probe stimuli. Subtraction analysis yielded multiple areas of activation following increased verbal and nonverbal working memory demands, including regions of the posterior cerebellar hemispheres (lobules VI, VII, VIII, and IX, crus I, and crus II), posterior paravermal cortex (lobule VI, crus I, and crus II), and the inferior vermis (lobules VI, VII, VIII, and IX).[91] This involvement of the posterior cerebellar hemisphere in working memory aligns with animal work demonstrating that this region neuroanatomically connects with the prefrontal cortex.[92]

To determine if the cerebellar correlates of working memory varied when verbal or nonverbal working memory tasks were performed, Ng and colleagues employed two different tasks during neuroimaging, including recall of a previously presented letter or a previously presented object. In the verbal working memory blocks, right lobules VI and VIIB were more predominantly activated, whereas nonverbal working memory tasks engaged bilateral lobules VI and VIIB of the posterior cerebellar hemispheres as well as lobule IX of the cerebellar vermis.[93] It is interesting to note that Chen and Desmond also reported this lateralization of verbal working memory within the posterior cerebellar hemispheres as well as cerebellar vermis lobule VI. Moreover, right cerebellar activation was even more prominent as the difficulty of the working memory task increased.[94] In a meta-analysis performed by Stoodley and Schmahmann, different verbal working memory tasks activated bilateral medial and lateral lobule VI as well as lateral crus I; larger activation clusters, including one in lobule VIII, were present in the right hemisphere.[15] In a more recent meta-analysis of fMRI studies done to assess the neural correlates of working memory, Emch and colleagues confirmed the presence of a robust, right-lateralized activation of crus I with verbal working memory tasks.[16] This lateralization, however, contrasts with some of the data obtained in pediatric cerebellar tumors discussed above, where lobule VIII of the left cerebellar hemisphere predicted the degree of verbal working memory impairment in children with pilocystic astrocytomas.[24] Thus, further work is needed to understand if the cerebellar correlates of verbal working memory change throughout development.

The above-mentioned clinical and neuroimaging data confirm that the cerebellum plays a critical role in working memory. The working memory profiles of many cerebellar disorders of childhood, however, remain understudied. As mentioned previously, the working memory abilities of children who suffered cerebellar hemorrhage as neonates are not known. Moreover, postinfectious acute cerebellar ataxia is a relatively common and often self-limiting neurological condition in children. Although its neuropsychological outcome is generally considered to be favorable,[95,96] focused studies of working memory are needed. In addition, further preclinical work is needed to understand how the cerebellum contributes to the existing working memory neural circuit and how, if at all, these cerebellar interactions are developmentally regulated.

Conclusions

Working memory impairments are a common nonmotor presentation of cerebellar disorders of childhood. That the cerebellum contributes to the working memory circuit is further supported by functional neuroimaging studies. Working memory should be assessed in children with cerebellar disorders throughout their development to ensure their educational plans are individually optimized.

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