Verbal Adynamia and Conceptualization in Partial Rhombencephalosynapsis and Corpus Callosum Dysgenesis

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Abstract: Verbal adynamia is characterized by markedly reduced spontaneous speech that is not attributable to a core language deficit such as impaired naming, reading, repetition, or comprehension. In some cases, verbal adynamia is severe enough to be considered dynamic aphasia. We report the case of a 40-year-old, left-handed, male native English speaker who presented with partial rhombencephalosynapsis, corpus callosum dysgenesis, and a language profile that is consistent with verbal adynamia, or subclinical dynamic aphasia, possibly underpinned by difficulties selecting and generating ideas for expression. This case is only the second investigation of dynamic aphasia in an individual with a congenital brain malformation. It is also the first detailed neuropsychological report of an adult with partial rhombencephalosynapsis and corpus callosum dysgenesis, and the only known case of superior intellectual abilities in this context.

Key Words: dynamic aphasia, adynamia, rhombencephalosynapsis, corpus callosum dysgenesis, spoken language

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ASD = autism spectrum disorder. **CCD** = corpus callosum dysgenesis. **RS** = rhombencephalosynapsis. **RT** = response time.

Spoken language is central to everyday life. Propositional language, also known as *spontaneous speech*, is spoken language that is spontaneous and voluntary and that includes a novel idea. Spontaneous speech is distinct from core language skills such as naming, reading, repetition, and comprehension, although all of these components are crucial for successful communication. In some individuals, spontaneous speech output becomes profoundly reduced but cannot be attributed to an impairment in any of the core language skills. This is the hallmark profile of *verbal adynamia*, which in some cases is severe enough to be considered dynamic aphasia.

DYNAMIC APHASIA

Dynamic aphasia (or frontal dynamic aphasia) is a rare language output disorder. Individuals with dynamic aphasia exhibit severely reduced spontaneous speech output in the context of generally intact core language skills. These individuals have been described as "completely deprived of spontaneous speech and seldom using it for the purpose of communication" (Luria, 1970, p. 199). All previous cases of dynamic aphasia, apart from one, have been documented in individuals with focal lesions (eg, Robinson et al, 1998, 2005) or neurodegenerative diseases (eg, Esmonde et al, 1996; Robinson et al, 2006, 2015; Snowden et al, 1996). A recent study by Berthier et al (2020) reported a single case of what they referred to as *developmental dynamic dysphasia* in an adolescent boy with cortical malformations, atypical placement of white matter tracts, and abnormal callosal fibers, which was the first case of dynamic aphasia to be reported in an individual with a congenital brain malformation.

Luria (1970) provided the first theoretical explanation for dynamic aphasia, suggesting that a breakdown in the translation of a conceptual plan into a sentence causes the impairment. Because individuals with dynamic aphasia have the requisite linguistic skills for spoken language yet show a severe paucity of spontaneous speech, detailed investigations into cases of dynamic aphasia have provided a unique insight into the cognitive processes that are not language per se but are critical for spoken language production and communication. Such investigations have identified three cognitive mechanisms as being involved in the successful production of spontaneous speech: idea selection, idea generation, and idea sequencing. More than one mechanism might be disrupted in an individual with dynamic aphasia (Robinson, 2013).

Idea Selection

Individuals with a deficit in idea selection have difficulty selecting a verbal response when many competing response options are available. That is, the individual's performance is reduced when a stimulus activates many semantic representations (eg, "room"—bed*room*, bath*room*, to make *room*) compared to when a stimulus activates a single dominant representation (eg, "Paris"—the city) (Robinson et al, 1998, 2005). This deficit has been documented in individuals with focal lesions of the left inferior frontal gyrus (Robinson et al, 2010).

Idea Generation

Individuals with a deficit in idea generation exhibit severely reduced performance on verbal and nonverbal fluency tasks (Bormann et al, 2008; Robinson et al, 2006, 2015) in addition to markedly reduced spontaneous speech, even though their core language skills are intact. This deficit has been documented in individuals with generalized bilateral frontal and subcortical damage (eg, progressive supranuclear palsy [Esmonde et al, 1996; Robinson et al, 2006, 2015] and bilateral striatocapsular infarctions [Gold et al, 1997]). A deficit in idea generation is thought to be a domain-general impairment.

Idea Sequencing

Individuals with a deficit in idea sequencing have problems shifting between ideas or linking ideas together (Robinson, 2013; Robinson et al, 2006). Thus, repeated ideas are considered indicative of sequencing difficulties. This deficit may be evident on fluency tasks; that is, reduced novel responses with a high number of perseverations indicate generation and sequencing deficits. A sequencing deficit can also cause poor coherence of spontaneous speech (Barker et al, 2017). For example, the narratives of individuals with impaired sequencing are generally sparse and include multiple repetitions of the same idea (Robinson et al, 2006), or there are problems conceptually linking ideas in a sequence across a narrative (Barker et al, 2017).

RHOMBENCEPHALOSYNAPSIS

Rhombencephalosynapsis (RS) is a rare malformation of the cerebellum. It is characterized by agenesis or dysgenesis of the cerebellar vermis and apparent fusion of the cerebellar hemispheres (Aldinger et al, 2018; Razek and Castillo, 2016). RS was first described in 1916 by Obersteiner, and the first case of partial RS (ie, partial fusion of the cerebellar hemispheres) was described ~16 years ago (Demaerel et al, 2004). Additional brain abnormalities such as septo-optic dysplasia, holoprosencephaly, fused thalami, hydrocephalus, and aqueductal stenosis (see also Gómez-López-Hernández syndrome [Razek and Castillo, 2016; Rush et al, 2013]) are often observed in individuals with RS.

There is no typical clinical presentation of RS, partly due to the range of associated supratentorial abnormalities (Tully et al, 2012). However, RS is commonly associated with intellectual impairments as assessed by intelligence scales (Pasquier et al, 2009; Poretti et al, 2009). To our knowledge, there are fewer than a handful of adult cases of RS with average intellectual functioning (Bell et al, 2005; Guyot et al, 2000; Obersteiner, 1916). Overall, descriptions of adult cases are largely absent from the literature (Pasquier et al, 2009), but with the use of MRI becoming more commonplace, it is likely that diagnoses in adulthood will become more frequent (Aldinger et al, 2018).

The cerebellum plays a key role in language, emotional processing, and even executive cognition (eg, Schmahmann, 1991; Schmahmann and Sherman, 1998; Veretennikoff et al, 2018). Cerebellar cognitive affective syndrome describes a set of neuropsychological symptoms that are associated with cerebellar damage and are characterized by deficits in executive function, impaired spatial cognition, personality changes (eg, flattened or blunted affect, disinhibited behavior), and language problems (eg, mild anomia, dysprosodia, agrammatism) (eg, Levisohn et al, 2000; Paulus et al, 2004; Schmahmann, 2004; Schmahmann and Sherman, 1997, 1998; Wolf et al, 2009). Cognitive and/or behavioral profiles consistent with cerebellar cognitive affective syndrome have been documented in individuals with RS (Poretti et al, 2009; Verri et al, 2000). Language problems have also been documented in individuals with cerebellar lesions, including mutism and subsequent dysarthria (van Dongen et al, 1994) as well as difficulties with complex language, such as sentence formation (Riva and Giorgi, 2000).

CORPUS CALLOSUM DYSGENESIS

Corpus callosum dysgenesis (CCD) is commonly observed in individuals with RS (Ishak et al, 2012), which may be because both CCD and RS potentially result from the failed induction of midline structures (Gobius et al, 2016; Razek and Castillo, 2016). CCD comes in various forms, ranging from complete and partial absence of the corpus callosum to a thin and thick corpus callosum (Paul et al, 2007). Neuropsychological investigations of individuals with CCD have revealed that their general cognitive ability tends to be within the average range (Paul et al, 2007). However, individuals with CCD have been shown to exhibit poor social cognition, emotional processing, theory of mind, processing speed, problem solving, and abstract reasoning skills (see Brown and Paul [2019] and Paul et al [2007] for reviews).

With regard to language, core language skills such as naming and reading generally remain intact in individuals with CCD (eg, Moutard et al, 2003; Siffredi et al, 2013), whereas complex language processes may be impaired. A longitudinal case study of a man with CCD reported a marked reduction of spontaneous speech and a lack of topic coherence that persisted into adulthood (Stickles et al, 2002). A reduction of spontaneous speech with spared core language skills is the hallmark of verbal adynamia, but adynamic speech has not yet been systematically investigated in individuals with CCD.

We report the case of an individual who presented with verbal adynamia, or a subclinical form of dynamic aphasia, in the context of partial RS and CCD.

CASE REPORT

Our participant is a high-functioning, 40-year-old, left-handed, male native English speaker with partial RS, CCD, and a language profile that is characteristic of dynamic aphasia, albeit milder or subclinical. He exhibited markedly delayed speech initiation and a significant reduction in spontaneous output in the context of a superior IQ. To our knowledge, we are the first group to document (a) an adynamic language profile that is consistent with a subclinical presentation of dynamic aphasia in the context of a high-functioning adult with a congenital brain malformation, and (b) a detailed neuropsychological investigation of an adult with partial RS and a superior IQ.

Our participant was recruited for a larger study (Principal Investigator L.J.R.) that had been approved by The University of Queensland's Human Research Ethics Committee. He contacted us to participate in the study via the AusDoCC Support Group, which is for individuals and families with CCD, and he provided informed and written consent. As a part of this larger study, our participant had an MRI scan at the University of Queensland Centre for Advanced Imaging in Brisbane, Australia. He also completed an interview and a baseline neuropsychological assessment at the University of Queensland School of Psychology at a visit that spanned 2 days in July 2017 and 1 day in May 2018.

INITIAL SCREENING

Brain Imaging

Our participant underwent an MRI at the University of Queensland Centre for Advanced Imaging (Figure 1).

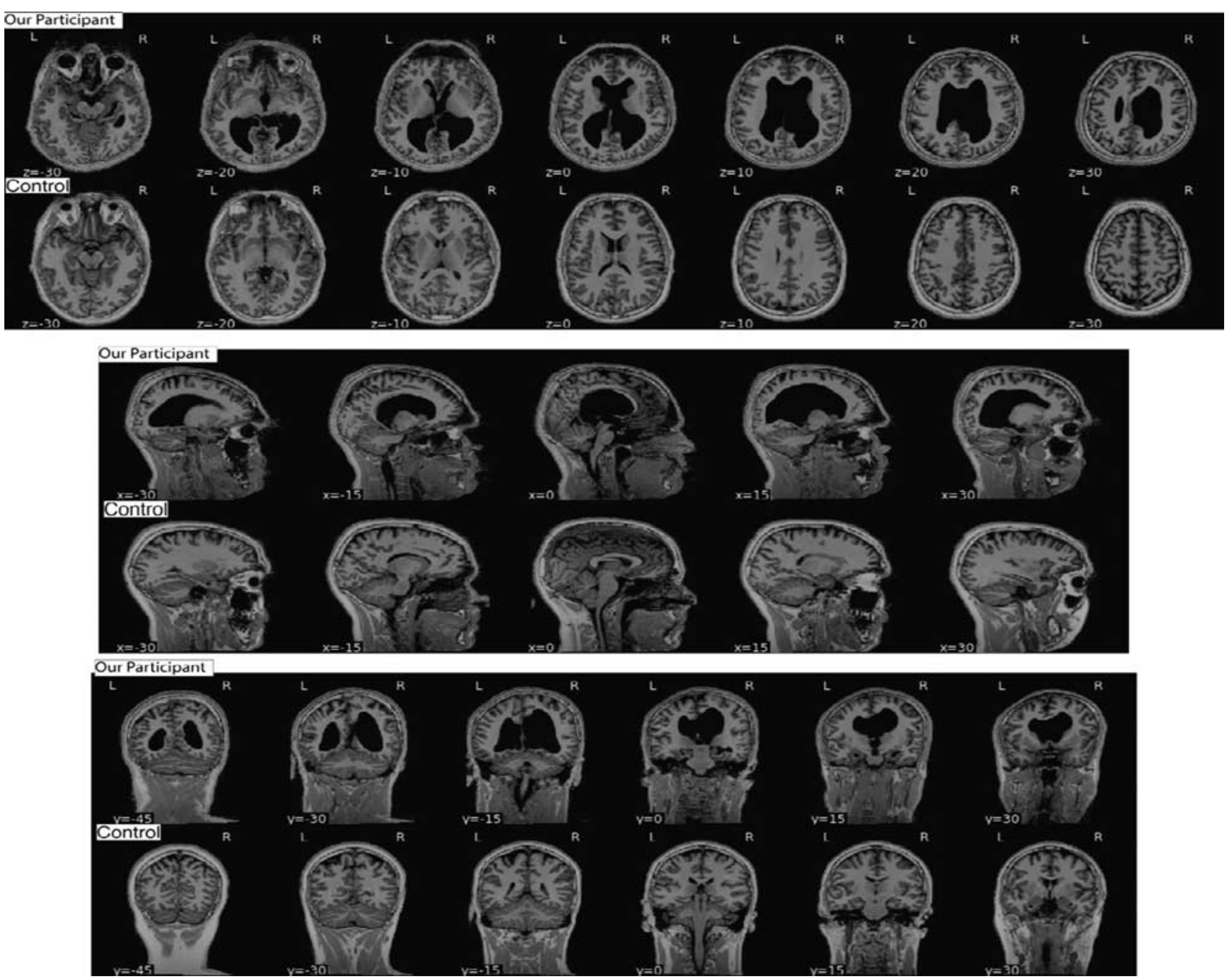

FIGURE 1. MRI of our participant and of an age-matched control. **L** = left. **R** = right.

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The structural MRI performed at 7T was evaluated by a neuroradiologist who specializes in brain malformations (S.M.). The MRI revealed partial RS and a dysmorphic brainstem. There was moderate to severe chronic supratentorial ventriculomegaly due to a high web obstructing the aqueduct of Sylvius. There were no features of acutely raised pressure.

As a result of prior arrested hydrocephalus, our participant's septum pellucidum was thinned, and the corpus callosum was stretched. The corpus callosum was also foreshortened and dysplastic, with a dysmorphic rostrum and genu, thin anterior body, very thin posterior body, and absent splenium (ie, diagnosis: partial absence or partial agenesis of the corpus callosum). There was a small anterior commissure, resulting in little interhemispheric forebrain connectivity overall. The posterior cingulum was disordered, and both hippocampi were hypoplastic, dysplastic, and malrotated. White matter volume and myelination were normal, but there was unusual sulcation of the left sylvian fissure, with unusual left lateral temporal radiating folds and sulcal conglomerates.

Interview

Our participant presented as a neatly and casually attired individual. He had a bachelor's degree in science and mathematics and worked as a medical administrator. He reported no history of a learning disorder, and he had not repeated any grades nor received any tutoring in school. He reported some gross motor (coordination) difficulties when he was younger. He also reported a history of anxiety and depression when he was younger, but he had never required psychotropic medications. There was no evidence that he was experiencing mood symptoms at the time that we saw him (Hospital Anxiety and Depression Scale [Snaith and Zigmond, 1994]). His levels of apathy were normal but slightly elevated (Apathy Evaluation Scale [Marin et al, 1991]).

Our participant's vision was corrected with spectacles, and he denied any hearing difficulties. He exhibited some clumsiness when navigating his spatial environment, and he bumped into objects when walking. His affect was somewhat restricted (ie, reduced range/intensity of facial expressions of emotion), and he frequently fidgeted, tapped, rocked in his chair, and produced verbal stereotypies (eg, *sss*). In response to questions, he was very slow to initiate spoken language, with several seconds of silence between the question and his answer. He did not initiate conversation with the interviewer. His thought processes appeared logical. We did not observe figure-of-eight head shaking.

Our participant informed us that he had received a diagnosis of autism spectrum disorder (ASD) from his health care team when he was younger, although this was unable to be confirmed. We administered the Autism-Spectrum Quotient (Baron-Cohen et al, 2001b) and he scored 36, which is above the cutoff of 32, suggesting self-reported ASD traits.

Baseline Neuropsychological Assessment

During the baseline neuropsychological assessment, our participant was enthusiastic and engaged. He was offered frequent breaks, and his fatigue levels were monitored closely, although he often declined breaks and requested that we extend the session beyond the scheduled finishing time because he enjoyed the tasks.

The baseline neuropsychological assessment included a range of standardized neuropsychological cognitive and language tests that assessed the following domains: general intelligence, verbal and visual memory, language, literacy, praxis, perception, attention, executive function, social cognition, and processing speed. The tests were administered by doctoral candidates (M.S.B. and J.L.K.) in accordance with published manuals and were reviewed by a senior clinical neuropsychologist (G.A.R.). The majority of the neuropsychological assessment took place in a quiet room at the University of Queensland School of Psychology in July 2017. The May 2018 testing was administered by M.S.B. and was completed in a quiet room at the University of Queensland Neuropsychology Research Clinic.

At the July 2017 visit, our participant completed all cognitive and language tests except for the following, which were completed at the May 2018 visit: Test for Reception of Grammar (Bishop, 2003), Kissing and Dancing Test (Bak and Hodges, 2003), Pyramids and Palm Trees Test (Howard and Patterson, 1992), and Verbal Microplanning Tasks (Bormann et al 2008; Costello and Warrington, 1989) (see below for details). These additional tests were selected based on our participant's pattern of performance at the July 2017 visit and were designed to further investigate his language skills and adynamic profile.

An age-, gender- and education-matched control group (n = 10: age: M [SD] = 36.4 [5.7] years; education: M [SD] = 17.7 [3.3] years) of native English speakers, drawn from the larger study, also completed the neuropsychological assessment and all of the experimental tasks, as well as an MRI scan. The controls were recruited via the University of Queensland research participation scheme, university newsletters, and word-of-mouth. All neuropsychological testing of the control group was administered by M.S.B. and J.L.K. in a quiet room at the University of Queensland School of Psychology over two testing sessions during 2017. One control participant reported a history of anxiety but was currently not medicated; no other neurologic or psychiatric histories were reported.

Cognitive Tests

We administered the following standard neuropsychological tests to both our participant and the control group: Wechsler Abbreviated Scale of Intelligence (Wechsler, 1999); Advanced Progressive Matrices (Raven, 1976); Oral Graded Difficulty Arithmetic Test (Jackson and Warrington, 1986); Incomplete Letters subtest from the Visual Object and Space Perception Battery (Warrington and James, 1991); Benton Facial Recognition Test (Benton et al, 1983); Recognition Memory Test for Words (Warrington, 1984), Faces (Warrington, 1984) and Topography (Warrington, 1996); Rey Complex Figure Test (Osterrieth, 1944; Rey, 1941); Rey Auditory Verbal Learning Test (Schmidt, 1996); Hayling Sentence Completion Test (Burgess and Shallice, 1997); Stroop Test (Trenerry et al, 1989); Brixton Test (Burgess and Shallice, 1997); Proverb Test (Murphy et al, 2013); Trail Making Test A and B (Reitan, 1956); Symbol

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Digit Modalities Test (Smith, 1982); Digit Span subtest from the Wechsler Adult Intelligence Scale—Third Edition (Wechsler, 1997); Test of Everyday Attention (Robertson et al, 1996); Reading the Mind in the Eyes Test (Baron-Cohen et al, 2001a); The Awareness of Social Inference Test (A) Part 1 (McDonald et al, 2003); and tests of praxic function (eg, imitation of meaningless gestures, execution of transitive and intransitive gestures) and orofacial praxic function (eg, puffing out cheeks).

Language Tests

We administered the following standard language tasks to both our participant and the control group: Graded Naming Test (McKenna and Warrington, 1980), Sentence Repetition (3–6 words in length; McCarthy and Warrington, 1984), Pyramids and Palm Trees Test, Kissing and Dancing Test, Synonym Test (Warrington et al, 1998), Test for Reception of Grammar, National Adult Reading Test (Nelson and Willison, 1991), Oral Graded Difficulty Spelling Test (Baxter and Warrington, 1994), and the Nonword Reading subtest from the Psycholinguistic Assessments of Language Processing in Aphasia (Kay et al, 1992).

Baseline Neuropsychological Assessment Results

All cognitive and language test scores for our participant and the controls are reported in Table 1.

Cognitive Tests

Our participant's intellectual functioning at the time of testing was in the high average range for nonverbal abilities and in the superior range for verbal abilities (Wechsler Abbreviated Scale of Intelligence, Advanced Progressive Matrices; Table 1). His numeracy skills were superior (Oral Graded Difficulty Arithmetic Test): Our participant produced no errors and completed each arithmetic problem exceptionally fast within the 10-second time limit. His visual perceptual and face perception skills were intact (Incomplete Letters subtest, Benton Facial Recognition Test). His verbal learning and recall memory were average (Rey Auditory Verbal Learning Test), with high average to superior recognition (Rey Auditory Verbal Learning Test, Recognition Memory Test Words). By contrast, his recognition memory for faces and topography were below average (Recognition Memory Test Faces, Recognition Memory Test Topography). Our participant recalled only 5% of the Rey Complex Figure after no delay, despite an accurate copy (Figure 2).

Our participant's executive functioning was mostly intact, although it was mildly reduced in the context of his superior IQ. His overall performance on the Hayling Sentence Completion Test was low average. In this test, the experimenter reads aloud a sentence with the last word omitted (eg, "*The old house will be torn …*"), and the participant orally generates either a sensible word (eg, "*down*": Section 1—initiation) or an unconnected word (eg, "*banana*": Section 2—suppression) as quickly as possible. Our participant had excessively prolonged response times (RTs), particularly on the initiation section (Table 1). However, the initiation and suppression condition total RTs were almost identical (total RT = 62 vs 63 seconds), indicating no difficulty with verbal inhibition, only initiation. Our participant produced no suppression errors. Corroborating this, our participant scored >99th percentile on the Stroop task. His spatial reasoning (Brixton Test), cognitive flexibility, and switching (Trail Making Test B) were average, and his interpretation of nonliteral language was good (Proverb task). His auditory–verbal attention and working memory were superior (Digit Span), as were his sustained and selective auditory attention and divided auditory/visual attention, although his visual selective attention was average (Test of Everyday Attention subtests Elevator Counting, Elevator Counting With Distraction, Dual Task Decrement, Telephone Search time/target, respectively). His psychomotor speed was average to slow (Symbol Digit Modalities Test, Trail Making Test A). His theory of mind was average (Reading the Mind in the Eyes Test); however, his performance on an emotion evaluation task (The Awareness of Social Inference Test) was very poor.

Our participant's ability to imitate meaningless gestures was slow and effortful, particularly for the left hand. His execution of transitive and intransitive gestures was similarly effortful, with poorer performance for transitive gestures, which involved gestures with objects such as a comb or hammer. His orofacial praxic function was satisfactory, with no difficulty in executing movements with his mouth.

Language Tests

Our participant's naming performance was within the average range (Graded Naming Test), and his repetition of sentences was flawless. His semantic knowledge was intact (Pyramids and Palm Trees Test, Kissing and Dancing Test), his single word comprehension was average to high average (Synonym Test), and his reception of grammar was excellent (Test for Reception of Grammar). His literacy skills were in the superior range (National Adult Reading Test, Oral Graded Difficulty Spelling Test), and his reading of irregular words (National Adult Reading Test) and nonwords was good. In summary, our participant's performance on the language tests indicated that his core language skills were intact. Given our participant's lack of conversation during the interview, but intact core language skills on testing, we suspected an adynamic language profile and decided to conduct a formal dynamic aphasia assessment.

DYNAMIC APHASIA INVESTIGATION

Based on the results of our participant's baseline neuropsychological assessment (ie, evidence of an adynamic language profile), we decided to complete a dynamic aphasia investigation. Thus, we administered additional experimental tasks that were specifically designed to characterize our participant's spontaneous speech and investigate the cognitive mechanisms that might underpin adynamia (ie, idea selection, generation, and sequencing as well as verbal microplanning). In this section, we first describe the formal assessment of our participant's spontaneous speech and then proceed with the experimental series, which systematically investigated the cognitive mechanisms that might be contributing to his reduced

TABLE 1. Summary of Demographic Information and Baseline Neuropsychological Assessment Scores for our Participant and the Age- and Education-matched Control Group (n = 10)

Characteristic	**Controls M (SD)**		**Our Participant**
Age (years)	36.4 (5.7)		40
Sex†	10 M: 0 F		M
Handedness†	10 R: 0 L		L
Education (years)	17.7 (3.3)		17
Cognitive Tests	**M (SD)**	**Raw Score**	**SS/Percentile**
Intellectual functions/numeracy			
Intellectual Functioning (WASI FSIQ)	119.6 (8.3)	—	IQ = 123
Verbal IQ	116.1 (10.0)		IQ = 122
Performance IQ	119.4 (10.7)		IQ = 118
Advanced Progressive Matrices (/12)	9.3 (2.2)	10	SS = 12; 79th percentile
Oral Graded Difficulty Arithmetic Test (/24)	14.6 (5.6)	24	SS >17
Perceptual functions			
Incomplete Letters subtest (/20)	19.7 (0/5)	19	WNL
Benton Facial Recognition Test (/54)	48.1 (1.9)	47	WNL
Memory			
Recognition Memory Test			
Words (/50)	48.0 (2.6)	50	>90th percentile
Faces (/50)	41.9 (5.6)	38	5–10th percentile
Topography (/30)	28.4 (1.4)	21	10–25th percentile
Rey Complex Figure Test			
Copy	35.6 (0.8)	34	80th percentile
Immediate Recall	25.4 (6.5)	2	<10th percentile
Rey Auditory Verbal Learning Test			
T1–5 total (/75)	55.5 (11.1)	44	35th percentile
T6 immediate recall (/15)	12.2 (2.6)	8	30th percentile
T7 delayed recall (/15)	11.9 (3.0)	9	45th percentile
Recognition hits (/15)	14.4 (0.8)	14	75th percentile
Executive functions			
Hayling Sentence Completion Test‡			
Initiation (RT) SS	5.8 (0.6)	—	1 (impaired)
Suppression (RT) SS	5.8 (0.6)		4 (low average)
Suppression error SS	5.5 (1.9)		8 (good)
Overall SS	5.5 (0.9)		4 (low average)
Stroop Test (c–w)§	132.3 (32.4)	128	>99th percentile
Brixton Test	8.7 (4.8)	Errors = 17	SS = 6 (average)
Proverb Test (/8)	5.9 (1.7)	7	Pass
Trail Making Test B	51.8 (11.1)	69 seconds	20–30th percentile
Speed and attention			
Trail Making Test A	22.4 (3.7)	42 seconds	<10th percentile
Symbol Digit Modalities Test (90 seconds)	56.3 (5.3)	46	54th percentile
Digit Span			
Forwards (/16)	11.6 (2.2)	16	SS = 19
Backwards (/14)	7.8 (1.9)	14	
Total (/30)	19.4 (3.6)	30	
Test of Everyday Attention			
Elevator Counting (/7)	6.9 (0.3)	7	WNL
Elevator Counting w/Distraction (/10)	8.6 (2.2)	10	SS = 13; >75th percentile
Telephone Search time/target	2.4 (0.4)	3.3 seconds	SS = 9; 25–50th percentile
Dual Task Decrement	1.1 (1.3)	–0.7	SS = 19; >99th percentile

TABLE 1. (*continued*)

Characteristic	Controls M (SD)	Our Participant	
Social cognition			
Reading the Mind in the Eyes Test (/36)	26.4 (4.6)	25	38th percentile
The Awareness of Social Inference Test–A (Part 1) (/28)	25.3 (1.9)	22	8th percentile
Praxic function			
Gesture production			
Meaningless L/R (/10 each side)	9.6 (0.7)/9.9 (0.3)	8/10	Abnormal/WNL
Transitive (/10)	9.9 (0.3)	8	Abnormal
Intransitive (/10)	9.8 (0.4)	9	WNL
Orofacial praxis (/15)	Not tested	15	WNL
Language tests			
Graded Naming Test (/30)	20.5 (2.9)	17	25th percentile
Sentence Repetition (/10)	10.0 (0.0)	10	WNL
Pyramids and Palm Trees Test (/52)	Not tested	51	WNL
Kissing and Dancing Test (/52)	Not tested	48	WNL
Synonym Test			
Abstract (/25)	20.3 (2.9)	22	50–75th percentile
Concrete (/25)	19.8 (2.0)	22	50–75th percentile
Total (/50)	40.1 (4.7)	44	50–75th percentile
Test for Reception Of Grammar (/80)	Not tested	78	WNL
Reading (NART–estimated IQ)	106.3 (8.1)	7 errors	IQ = 122
Oral Graded Difficulty Spelling Test (/30)	19.1 (3.9)	28	>96th percentile
Nonword Reading subtest (/24)	22.3 (2.5)	24	WNL
Mood symptoms			
HADS			
Anxiety (/21)	7.2 (2.9)	7	WNL
Depression (/21)	2.4 (1.6)	2	WNL

†Control group scores for sex and handedness represent ratio (not mean) scores.
‡Hayling overall SS range = 1–10, initiation SS range = 1–7, suppression/suppression error SS range = 1–8, 6 = average.
§Stroop c–w = number of colors correctly named in the color-word condition of the Stroop Test in 2 minutes.
F = female. **HADS** = Hospital Anxiety and Depression Scale. **L** = left. **M** = male. **NART** = National Adult Reading Test. **RT** = response time. **SS** = scaled score, range 1–19, 10 = average. **WASI FSIQ** = Wechsler Abbreviated Intelligence Scale Full Scale IQ. **WNL** = within normal limits.

spontaneous speech. These experimental tests were completed at the time of the neuropsychological assessment and MRI scan (July 2017 and May 2018).

Spontaneous Speech

Our participant's spontaneous speech was characterized by long pauses of several seconds between utterances, which were then produced in quick bursts of words. His speech was softly spoken, had abnormal prosody, and was of an atypical rhythm; for instance, he would often draw out particular phonemes (eg, *there'sssss a guy*). However, his content was appropriate and rich. He rarely, if ever, initiated conversation. He had no articulatory or grammatical difficulties, no speech sound distortions, and no semantic or phonological paraphasias during general conversation or testing.

Spontaneous Speech Assessment

We decided to use two picture-elicited narrative tasks and a self-generated narrative topic task to assess the spontaneous speech of both our participant and the control group. The pictorial scenes were the Beach Scene from the Queen Square Screening Test for Cognitive Deficits (Warrington, 1989) and the Cookie Theft Scene from the Boston Diagnostic Aphasia Examination (Goodglass et al, 2001). Both are complex scenes in which various actions are taking place, and the task is to "describe what is happening in this picture using full sentences." Both picture description tasks were 1 minute in duration. For the self-generated narrative Best Holiday task, participants were asked to "talk about your favorite holiday" for 2 minutes.

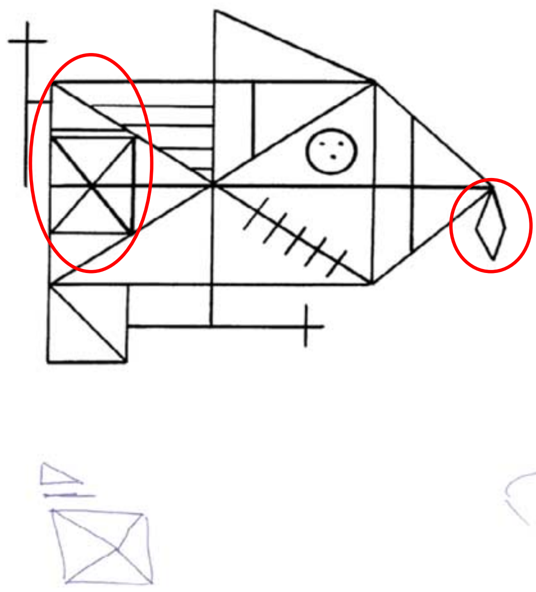

FIGURE 2. Our participant's Rey Complex Figure Recall alongside the original figure (from *Le Test de Copie d'Une Figure Complexe*, Osterrieth, 1944).

An Olympus WS-813 digital voice recorder was used to record all speech samples. Samples were transcribed in English orthography, and transcriptions included all words, sounds, and repeats. Contractions were counted as two words (eg, won't = will not). Nonwords (eg, "umm"), habitual starters (eg, "okay" "alright then"), questions and comments directed at the experimenter (eg, "how much longer?"), and words that were subsequently repaired (eg, direct repetitions) were not counted in the total number of words.

Spontaneous Speech Results

Table 2 provides the results of the spontaneous speech tasks for both our participant and the control group. Our participant produced 53 wpm on the Beach Scene task and 44 wpm on the Cookie Theft Scene task. By contrast, the controls produced a mean of 174.4 (SD = 46.9) wpm on the Beach Scene task and a mean of 124.5 (SD = 45.7) wpm on the Cookie Theft Scene task. Thus, on both tasks, our participant produced ~35% of the words that the controls did, and his wpm score was ~2 SDs below the mean of the controls.

Similarly, on the Best Holiday task, our participant produced 98 words in 2 minutes, and the controls produced a mean of 317.3 (SD = 103.9) words in 2 minutes. Again, our participant produced ~30% of the spontaneous speech that the controls did, and his wpm score was ~2 SDs below the mean of the controls. Given our participant's severely slowed verbal initiation, one might speculate that perhaps he was simply slower to begin speaking and therefore had less time remaining in the task. However, his pattern of performance across the 2 minutes of the Best Holiday task shows that he did not produce a normal amount of spontaneous speech after an initial delay (1st minute = 42 words, 2nd minute = 56 words).

TABLE 2. Words per Minute Produced on the Spontaneous Speech Tasks for Our Participant and the Control Group (n = 10)

Task	Controls M (SD)	Our Participant
Beach Scene—1 minute	174.4 (46.9)	53*
Cookie Theft Scene—1 minute	124.5 (45.7)	44
Best Holiday—2 minutes	317.7 (103.9)	98
Mean (SD) wpm	152.6 (25.5)	48.7 (4.5)**

Our participant was compared to the healthy control group using a modified *t* test (Crawford and Garthwaite, 2002).
*Significant at $P < 0.05$.
**Significant at $P < 0.01$.

Our participant's wpm scores were not as reduced as those of the individuals with dynamic aphasia reported by Robinson et al (2015; M = 23.4). However, our participant's spontaneous speech output was disproportionately and markedly reduced relative to his intact core language skills and compared to the control group (participant M = 48.7 wpm; controls M = 152.6 wpm, P = 0.003). This language profile is the hallmark of dynamic aphasia.

Experimental Series

The experimental series of tasks aimed to investigate possible factors underpinning our participant's reduced spontaneous speech. Previous investigations of individuals with dynamic aphasia have offered various explanations for the reduction in spontaneous speech output. We decided to investigate idea selection, idea generation, and the fluent sequencing of novel ideas (Bormann et al, 2008; Robinson et al, 1998, 2005, 2006, 2013, 2015). We also investigated verbal microplanning (ie, constructing sentences from discrete words) because this skill may also be impaired in individuals with dynamic aphasia (Costello and Warrington, 1989; Snowden et al, 1996; Warren et al, 2003).

Idea (Proposition) Selection

Difficulty with idea selection, that is, selecting an idea from competing alternatives, was the core impairment in a subset of individuals with dynamic aphasia (Crescentini et al, 2008; Robinson 2013; Robinson et al, 1998, 2005). Our participant completed a series of experimental sentence generation and sentence completion tasks that are sensitive to idea-level selection deficits (Barker et al, 2020; Robinson et al, 1998, 2005, 2010). Each task included high- and low-constraint stimuli: High-constraint stimuli activate a single dominant proposition and therefore place low demands on selection mechanisms, whereas low-constraint stimuli activate many competing representations and place high demands on selection mechanisms. Poorer performance in low-constraint conditions relative to high-constraint conditions is illustrative of an idea-level selection deficit in individuals with dynamic aphasia (Robinson et al, 1998, 2005).

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For all sentence generation and completion tasks, a stopwatch was used to record the RT— the time between the end of stimulus presentation and the beginning of the response. An error was recorded if an individual did not produce a grammatical or meaningful response, or, in the case of the latter two tasks, if the target word/s were omitted.

Idea Selection Assessment. We administered three tasks that are sensitive to idea-level selection: (a) sentence completion, (b) sentence generation from a single word, and (c) sentence generation from a word pair. The *sentence completion task* requires oral generation of a single word to complete a sentence frame with the final word omitted. High-constraint sentences are designed to activate a dominant response, for example "*He scraped the cold food from his* plate" (Hayling Sentence Completion Test). Low-constraint sentences are designed to activate many competing response options, for example, appropriate responses to "*They went to see the famous* ____" may include "actress," "museum," "singer," or "building" (Bloom and Fischler, 1980). The stimuli included 15 high- and 15 low-constraint sentence stems.

The *sentence generation from a single word task* and the *sentence generation from a word pair task* require oral generation of a meaningful sentence that includes either one or two target words. The sentence from a single word stimuli were high-frequency (common) words and proper nouns. High-frequency words are low constraint because they activate many competing possible responses (eg, "chair"—dining chair, lounge chair, or even committee chair). By contrast, proper nouns are high constraint because there is only one definition of the word (eg, "Sweden"—the country of Sweden); thus, they activate a dominant proposition. The stimuli included 15 high-frequency words and 15 proper nouns. The sentence generation from a word pair stimuli were high-association and low-association pairs: High-association pairs (eg, crater–moon) are high in constraint because they activate a dominant proposition, whereas low-association pairs (eg, crater–cave) are low in constraint because they activate many propositions. The stimuli included 22 high-association pairs and 22 low-association pairs.

Idea Selection Results and Conclusion. We compared our participant's performance on the selection tasks to that of our control group using a modified *t* test (Crawford and Garthwaite, 2002). A summary of results is presented in Table 3. Unlike previously reported individuals with dynamic aphasia (eg, Robinson et al, 1998, 2005), our participant did not produce any errors in either the high- or low-constraint conditions on any of the selection tasks. The controls also made virtually no errors on the tasks.

Overall, our participant's RTs across all three tasks, and in both selection conditions, were significantly slower than those of the controls, all $P < 0.001$. This finding is unsurprising given that impaired verbal initiation was also evident on our participant's baseline neuropsychological assessment (see Hayling Sentence Completion Test, Table 1). However, the magnitude of this impairment is vast: On all three sentence tasks, our participant's mean RTs were at least five times (up to 11 times) slower than those of the controls (Table 3).

In order to investigate potential deficits in idea selection, we compared the performance between the low- and high-selection conditions of each task. Previously, we have used difference scores (ie, low-constraint RT − high-constraint RT) to compare the mechanism of selection between groups (Barker and Robinson, 2019). However, given our participant's extremely slow RTs, this approach was not considered appropriate for the current data. Instead, a selection contrast ratio was calculated for each individual on each task by dividing the low-constraint condition mean RT by the high-constraint condition mean RT (selection contrast ratio = low-constraint mean RT/high-constraint mean RT) (Barker et al, 2020).

Selection contrast ratios provide a measure of *the degree* to which performance in the low-constraint condition is slower than performance in the high-constraint condition, such that the higher the number, the greater the selection deficit. This approach allowed a direct comparison between our participant and the control group. The results revealed that our

TABLE 3. Mean Response Times and Errors on Selection Tasks for our Participant and the Control Group (n = 10)

Selection Demands (*Constraint*)	**Controls M (SD)**	**Our Participant**
Sentence completion		
High constraint		
Number correct (/15)	15.0 (0.0)	15
Mean RT (seconds)	0.9 (0.3)	4.5***
Low constraint		
Number correct (/15)	15.0 (0.0)	15
Mean RT (seconds)	1.5 (0.5)	16.9***
Selection contrast ratio (*low/high*)	1.6 (0.4)	3.8***
Sentence generation from a single word		
High-frequency words (*low*)		
Number correct (/15)	15.0 (0.0)	15
Mean RT (seconds)	2.4 (1.1)	19.5***
Proper nouns (*high*)		
Number correct (/15)	15.0 (0.0)	15
Mean RT (seconds)	2.2 (1.3)	12.3***
Selection contrast ratio (*low/high*)	1.3 (0.6)	1.6
Sentence generation from a word pair		
High association (*high*)		
Number correct (/22)	21.7 (0.9)	22
Mean RT (seconds)	2.2 (0.9)	11.0***
Low association (*low*)		
Number correct (/22)	22.0 (0.0)	22
Mean RT (seconds)	2.6 (1.2)	13.9***
Selection contrast ratio (*low/high*)	1.2 (0.2)	1.3

Our participant was compared to the healthy control group using a modified *t* test (Crawford and Garthwaite, 2002).
***Significant at $P < 0.001$.
RT = response time.

participant's selection contrast ratio for the sentence completion task was significantly higher than that of the controls, $P < 0.001$. In fact, our participant was almost four times slower in generating a response in the low-constraint condition (compared to the high-constraint condition), whereas the controls were 1.6 times slower. Our participant's selection contrast ratio was not significantly higher than that of the controls on the other two tasks (both $P > 0.05$).

Although our participant was generally slow in generating responses, he was markedly slower in the low-constraint condition of the sentence completion task when the demands on the selection mechanisms were high. By contrast, the controls were not proportionally slowed to the same degree in the low-constraint condition of the sentence completion task. Close inspection of the raw RTs for the initiation section of the Hayling Selection Completion Test revealed that our participant was slowest to respond to the item with the lowest probability of a dominant response (ie, "George could not believe that his son had stolen a _____." Our participant responded "toy" after 19 seconds.). Taken together, these findings suggest that an idea selection deficit may be an underlying factor driving our participant's reduced spontaneous speech; however, in contrast to previously reported cases of dynamic aphasia, it is manifest in RTs rather than in errors.

Idea Generation and Sequencing

The two mechanisms of idea generation and sequencing have been impaired in several previous cases of dynamic aphasia (Robinson, 2013; Robinson et al, 2006, 2015). Fluency tasks require generation of responses across a defined time period in response to a single cue, and a deficit in idea generation manifests as a reduction in the total number of items generated on verbal and/or nonverbal fluency tasks. A deficit in idea sequencing may manifest as perseverative or repetitive responses on fluency tasks (Robinson et al, 2006, 2015) or as spontaneous speech that lacks coherence (Barker et al, 2017).

Fluency Tasks Assessment. Our participant and the control group completed two verbal and two nonverbal fluency tasks. The verbal fluency tasks included phonemic and semantic word fluency (Benton, 1968), where participants are aurally presented with a phonemic cue (ie, letter of the alphabet: F, A, S) or a semantic category (eg, animals) and are asked to orally generate in 60 seconds as many words as they can that begin with that letter or belong to that category. Participants are instructed not to use proper nouns, numbers, or the same word with different endings for phonemic fluency (Strauss et al, 2006).

The nonverbal fluency tasks included design fluency (free and fixed conditions; Jones-Gotman and Milner, 1977) and gesture fluency (meaningless and meaningful conditions; Jason, 1985). The design fluency free condition requires participants to draw as many abstract designs as possible that do not represent identifiable objects in 5 minutes. In the fixed condition, there is an additional constraint that each design consist of only four straight or four curved lines (4 minutes allowed). The gesture fluency task requires individuals to produce as many discrete movements with their hands (ie, gestures) that are either meaningful (ie, waving) or meaningless in 2 minutes. Responses are coded as correct, repetitions, or rule-break errors. Total correct unique responses and repetitions for both the participant and the control group are reported in Table 4.

Coherence Assessment. To assess coherence, we inspected the spontaneous speech samples of our participant and the control group for coherence errors: conceptual incongruence (ie, ideas not directly related to the stimulus), tangential sentences (ie, ideas that are related to the stimulus but irrelevant to the task, eg, "this is a beach / I went to the beach last week"), and propositional repetitions (ie, repeated ideas) (Marini et al, 2011; Robinson et al, 2006).

Idea Generation and Sequencing Results and Conclusion. We compared our participant's performance with that of the control group using a modified t test (Crawford and Garthwaite, 2002). In terms of total responses generated, our participant did not perform significantly below the controls on any of the fluency tasks, all $P > 0.05$.

TABLE 4. Total Number of Unique Responses and Repetition Errors on Verbal and Nonverbal Fluency Tasks for our Participant and the Control Group (n = 10)

Task	Controls M (SD)	Our Participant
Verbal fluency		
Phonemic fluency (FAS: each 1 minute)		
Total	42.7 (8.5)	30†
Repetitions	1.4 (1.8)	0
Semantic fluency (animals: 1 minute)		
Total	22.8 (4.6)	18
Repetitions	0.5 (0.9)	1
Nonverbal fluency		
Design fluency		
Free (5 minute)		
Total	28.8 (26.2)	15
Repetitions	0.3 (0.5)	0
Fixed (4 minute)		
Total	26.0 (17.4)	10‡
Repetitions	0.3 (0.7)	0
Gesture fluency		
Meaningless (2 minute)		
Total	15.0 (8.8)	7‡
Repetitions	1.7 (1.8)	0
Meaningful (2 minute)		
Total	18.7 (7.2)	8‡
Repetitions	0.7 (0.9)	0

† <10th percentile based on normative data from Tombaugh et al (1999).

‡ <10th percentile based on normative data from Robinson et al (2012).

FAS = F, A, and S form of the Controlled Oral Word Association Test.

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However, comparison with normative data (Tombaugh et al, 1999) showed that he performed at the 10th percentile for his age and education level on the phonemic fluency task, which is low in the context of his superior IQ. Similarly, he performed at the 10th percentile or below on fixed design fluency and gesture fluency compared with normative data (Robinson et al, 2012).

Slow and effortful praxis, particularly with the left hand, may have impacted our participant's nonverbal fluency performance. Nevertheless, these scores tentatively suggest that there may be a mild deficit in our participant's generation of ideas. His reduced totals cannot be attributed to delayed initiation because he was consistent in his rate of responding across the tasks, including gesture fluency. With regard to sequencing, he produced only one perseverative response across all of the fluency tasks. Furthermore, there were no coherence errors in his spontaneous speech. (All of his speech transcripts are provided in the supplemental digital content at http://links.lww.com/CBN/A82.) Thus, we considered a sequencing difficulty unlikely to be an underlying factor driving his reduced spontaneous speech.

Verbal Microplanning

We gave our participant a set of tasks that were designed to investigate microplanning (Costello and Warrington, 1989; Snowden et al, 1996). The sentence construction task, which was originally reported by Costello and Warrington (1989), requires rearrangement of words printed on cards to construct a meaningful and grammatical sentence. A microplanning task, first described by Bormann et al (2008), requires the rephrasing of sentences without altering their meaning (eg, The car drives behind the bus → The bus drives in front of the car). Each task contains 10 trials. On both tasks, our participant performed flawlessly, producing zero errors. This finding excludes an impairment of verbal microplanning as an underlying factor driving his reduced spontaneous speech.

Experimental Series: Conclusion

Our participant's reduced spontaneous speech cannot be explained by a core language deficit (eg, in naming, comprehension, reading, or articulation). We suggest that his language profile represents mild or subclinical dynamic aphasia, or verbal adynamia, because his spontaneous speech was not as reduced as that of other individuals with dynamic aphasia. Our experimental investigation revealed that he had an idea selection deficit and a potential mild problem with the generation of ideas.

DISCUSSION

Our participant is the first documented case of an adult with superior intellectual abilities in the context of partial RS and CCD. Indeed, one of the most striking aspects of our participant's cognitive profile is his superior IQ. The few descriptions of adults with RS in the literature usually report intellectual impairments (Pasquier et al, 2009; Poretti et al, 2009). We are aware of one individual with RS and an average IQ (Bell et al, 2005) and two others whose IQs were estimated to be in the average range based on their occupations (Guyot et al, 2000; Obersteiner, 1916). However, we are not aware of any neuropsychological reports of an adult with partial RS.

Our participant's cognitive profile was similar to that of the case described by Bell et al (2005): a 55-year-old man with RS, and the only other detailed neuropsychological investigation of an adult with RS to date. Both our participant and the case reported by Bell and colleagues (2005) had reduced recognition memory for faces, poor motor dexterity, and naming skills that were slightly below what would be expected based on education level. However, our participant had a significantly higher full-scale IQ than the case reported by Bell and colleagues (2005) (our participant's IQ = 123; Bell et al case's IQ = 90), and Bell and colleagues did not provide any data on their case's spontaneous speech. Bell and colleagues (2005) attributed their case's largely intact intellectual functioning to limited supratentorial abnormalities, in line with Guyot et al (2000), who reported a case whose IQ was estimated to be in the average range based on her employment as a secretary (formal IQ testing was not conducted). However, our report of our participant suggests that intellectual functioning may remain intact even when multiple supratentorial abnormalities are evident, indicating vast heterogeneity in cognitive outcomes.

Consistent with known deficits in CCD (Paul et al, 2007; Symington et al, 2010), our participant's social cognition was poor (The Awareness of Social Inference Test). It is likely that poor social cognition contributed to his diagnosis of ASD; indeed, CCD is considered a major risk factor for an ASD diagnosis (Brown and Paul, 2019; Lau et al, 2013; Paul et al, 2014). Similarly, there is evidence in the literature that the cerebellum is implicated in ASD (eg, Becker and Stoodley, 2013; Fatemi et al, 2012).

Clinically, there is clear overlap between ASD and cerebellar cognitive affective syndrome. Our participant exhibited aspects of cerebellar cognitive affective syndrome in his poor visual/spatial memory, flattened affect, and dysprosodic speech—the latter two of which also may have contributed to his ASD diagnosis. By contrast, our participant's proverb interpretation was excellent, and his narratives were logical and coherent, which is not typical of individuals with CCD or ASD (Paul et al, 2004; Rehmel et al, 2016). Neither did he display any signs of disinhibition.

Finally, slow processing speed is considered a core cognitive deficit in individuals with CCD (Brown and Paul, 2019), but whereas our participant was slow on Trail Making Test A, he was superior on other timed tasks such as block design, mental arithmetic, and the Stroop task, which is atypical for individuals with CCD. For this reason, we do not consider slowed processing speed to be a significant factor contributing to our participant's language profile. He had difficulty with praxic function in his left hand (his writing hand), which may have slowed his performance on Trail Making Test A.

Spontaneous Speech: Subclinical Dynamic Aphasia?

In contrast to our participant's semantic knowledge and his reading, repetition, comprehension, spelling, and grammar skills, which were good, our participant's spontaneous speech was markedly reduced, and his verbal initiation was extremely slow. In this way, our participant bears some resemblance to the case reported by Stickles et al (2002) of an individual with CCD whose vocabulary was spared but whose spontaneous speech was reduced and "halting," with unusually long pauses and a lack of conversational initiation.

Reduced spontaneous speech that is not attributable to impaired core language skills or articulation problems best fits the profile of verbal adynamia or dynamic aphasia. In many ways, our participant's profile was similar to that of a case of dynamic aphasia in the context of frontotemporal degeneration and nonfluent progressive aphasia (Robinson et al, 2005). Like our participant, the Robinson et al (2005) case had a high premorbid and nonverbal IQ and good executive functioning (eg, inhibition). For example, on the Hayling Sentence Completion Test inhibition section, both our participant and the other case implemented a strategy and produced few errors. However, our participant's spontaneous speech was less profoundly reduced than that of the other case and was devoid of the articulation difficulties that the other case displayed. Robinson and colleagues (2005) attributed the other case of dynamic aphasia to a deficit in selecting an idea from an array of competing alternatives (see also Robinson et al, 1998).

Idea Selection

Both our participant and the case reported by Robinson and colleagues (2005) performed more poorly on the initiation section of the Hayling Sentence Completion Test than on the inhibition section, indicating slowed verbal initiation. Close inspection of our participant's Sentence Completion Test scores revealed that he, like the other case, had exceedingly long RTs for the sentences with the lowest probability of a dominant response (eg, "George could not believe his son had stolen a …"). In line with this, on the experimental idea selection sentence completion task, our participant was 3.8 times slower to produce a word to complete a low-constraint sentence than he was to complete a high-constraint sentence, whereas the controls were only 1.6 times slower. On the other experimental idea selection tasks, our participant was slow, and although his *proportional* slowness in the low-constraint conditions relative to the high-constraint conditions was not statistically significantly different from that of the controls, the raw RTs showed a trend in that direction. Taken together, this pattern of performance suggests some degree of impairment in idea selection, similar to that of the case reported by Robinson and colleagues (2005), albeit not as severe. Whereas the case reported by Robinson and colleagues (2005) was unable to produce responses in the low-constraint conditions, our participant was able to produce responses, but they were extremely slow. RT data, as opposed to "no response" errors, may be sensitive to idea selection deficits at the subclinical level (Barker et al, 2020; Barker and Robinson, 2019).

Idea Generation

A deficit in the ability to generate novel ideas for expression has been offered as one explanation for dynamic aphasia (Robinson, 2013; Robinson et al, 2006, 2015). In the context of dynamic aphasia, reduced production on verbal and nonverbal fluency tasks has been interpreted as a problem in generating ideas. Our participant's verbal fluency totals were not as low as those previously reported (eg, Robinson et al, 2006, 2015), and his semantic fluency was actually within normal limits, but his phonemic fluency was reduced compared to normative data (Table 4) and what would be expected based on his superior IQ. It is not uncommon for individuals with dynamic aphasia to perform better on semantic fluency tasks than phonemic fluency tasks, possibly because semantic skills are intact in dynamic aphasia (Esmonde et al, 1996; Robinson et al, 2005).

Similar to phonemic fluency, our participant's nonverbal fluency was not impaired but was reduced, although his slow psychomotor speed and praxis difficulties may have contributed to a lowering of these scores. Overall, our participant's mild reductions in fluency are consistent with a very mild problem generating ideas for expression. A deficit in idea generation may reflect a disorder of *energization*, or an inability to maintain responding over time in the absence of external cues; problems with energization are characterized by a "drop-off" pattern of responding (Barker et al, 2018; Robinson et al, 2012). However, our participant's pattern of responding does not fit an energization account because his language output did not decline over time; rather, it was characterized by lengthy pauses throughout and an overall reduction in quantity.

Neuroanatomical Correlates

Brain imaging studies purport a widely distributed network of brain regions that are involved in spontaneous speech (propositional language) production (Awad et al, 2007; Blank et al, 2002; Braun et al, 2001; Brownsett and Wise, 2010; Grande et al, 2012; Tremblay and Small, 2011; Troiani et al, 2008). More specifically, there is some suggestion that initiation processes and organizational aspects of language are associated with the frontoparietal brain regions (Blank et al, 2002; Braun et al, 2001; Brownsett and Wise, 2010; Grande et al, 2012; Troiani et al, 2008). It is possible that in our participant, these networks were disrupted due to poor interhemispheric connectivity as a result of his CCD or other developmental miswiring. Indeed, abnormal frontoparietal connectivity has been documented in individuals with CCD (Hearne et al, 2018). This explanation is consistent with the only other known case of dynamic aphasia in the context of congenital brain malformation, whereby profoundly reduced spontaneous speech was partially attributed to abnormal white matter tracts and callosal fibers disrupting language networks (Berthier et al, 2020). Interhemispheric transfer may play a key role in spontaneous speech production, which is complex and draws on higher order cognitive skills.

Based on lesion studies and individuals with neurodegenerative disease, there is evidence that the left inferior frontal gyrus supports idea selection (Robinson et al, 2005, 2010), whereas bilateral frontal regions and frontostriatal circuits support idea generation (Barker et al, 2018; Robinson et al, 2006, 2015). Berthier and colleagues (2020) proposed that, in addition to interhemispheric connectivity problems, malformed frontal regions, including the left inferior frontal gyrus, may have contributed to the paucity of spontaneous speech in their neurodevelopmental dynamic aphasia case. Likewise, in our participant, it is possible that idea selection and generation problems were related to frontal disruption as a result of cortical malformations, miswiring, or disturbed connections between the cerebellum and frontal cortices. At least with regard to idea generation, poor phonemic fluency has been reported in cerebellar pathology (Veretennikoff et al, 2018). However, neuroanatomical correlates of idea selection and generation have been proposed in the context of brain lesions and/or neurodegeneration, whereby a previously functional system has been disrupted. Therefore, caution is required when generalizing to a neurodevelopmental context because the degree of cortical reorganization or rewiring in the case of our participant is unknown.

Finally, it is unlikely that our participant's language profile was due to hydrocephalus because his overall cognitive pattern was not consistent with the global flattening that is generally seen in individuals with hydrocephalus. Despite this finding, the presence of hydrocephalus should incite caution when evaluating this case.

Theoretical Implications

Models of spoken language production posit a preverbal stage of conceptualization, or idea formulation, that occurs before linguistic formulation (the lexical or word level) and articulation (eg, Levelt, 1989, 1993, 1999; Sherratt, 2007). During conceptualization, a communicative intention or message is generated. Message generation is a process that requires the selection, generation, and sequencing of ideas. It represents the interface between broader cognitive skills and language. Our participant is the first demonstration of co-occurring deficits in idea selection and generation with no evidence of a sequencing problem. Together with the existing dynamic aphasia literature, our case sheds light on the architecture and relationship between these mechanisms, suggesting that they are organized in an interconnected manner and that disruptions to any combination of these mechanisms may result in reduced spontaneous speech.

CONCLUSION

Spontaneous speech, or propositional language, is key for successful social communication. We reported a case of a high-functioning adult with markedly reduced spontaneous speech in the context of a congenital brain malformation, and we characterized this as a subclinical presentation of dynamic aphasia, or verbal adynamia. Language profiles consistent with dynamic aphasia are generally not considered in developmental disorders. Except for one, all previous reports of dynamic aphasia have been in the context of brain lesions or neurodegenerative disease. Our investigation highlights that the same deficient mechanisms underpinning reduced spontaneous speech in acquired cases of dynamic aphasia (eg, problems with idea selection and/or generation) may be applicable in neurodevelopmental cases with similar language profiles. In addition, we have provided the first detailed neuropsychological profile of an adult with partial RS and CCD. To our knowledge, this is the first documented case of an adult with a superior IQ in partial RS. Our results speak to the heterogeneity of outcomes in individuals with partial RS and CCD.

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