ELECTROPHYSIOLOGICAL ASSESSMENT OF ASPECTS OF LANGUAGE IN A CHILD WITH CONGENITAL DISORDER OF GLYCOSYLATION: A CASE STUDY

by

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Table of Contents

List of Figures vi
Abstract vii
Introduction 1
Method - Experiment 1 13
Participants 13
Materials & Stimuli 15
Procedure 17
EEG Acquisition 18
ERP Analyses 19
Results - Experiment 1 20
Discussion - Experiment 1 24
Method - Experiment 2 26
Participants 26
Materials & Stimuli 26
Procedure 27
EEG Acquisition 28
ERP Analyses 29
Results - Experiment 2 30
Discussion – Experiment 2 33
General Discussion 35
References 38
Appendix A. Consent Form – Typically Developing Children 45
Appendix B. Consent Form – CDG Participant 48
Appendix C. Feedback & Debriefing Form – Typically Developing Children 51
Appendix D. Feedback & Debriefing Form – CDG Participant 53

List of Figures

Figure 1. ERP Responses for Typically Developing Children in Experiment 1 ... 21
Figure 2. ERP Responses for RS in Experiment 1 ... 23
Figure 3. ERP Responses for Typically Developing Children in Experiment 2 ... 31
Figure 4. ERP Responses for RS in Experiment 2 ... 32

Abstract

The current study examined the language abilities of a child, RS, who has Congenital Disorder of Glycosylation (CDG). Due to severe verbal and motor impairments, event-related potentials provide the only suitable means to assess her cognitive functioning. Typically developing children served as a comparison group in two experiments. In the first, participants heard their own names interspersed with seven others. Both RS and the comparison group displayed a larger negativity to their own names compared to other names. The second experiment had two conditions. In the word condition, a standard word was replaced on some trials by a second word, the deviant. In the pseudoword condition, a standard word was replaced by a pseudoword deviant. While the comparison group elicited a mismatch negativity (MMN) to word and pseudoword deviants, RS elicited an MMN-like response to pseudoword deviants only. Overall, RS appears to recognize her own name and distinguish words from pseudowords.

Keywords: language, children, Congenital Disorder of Glycosylation (CDG), event-related potentials (ERP), mismatch negativity (MMN), P300, name recognition, auditory.

Electrophysiological Assessment of Aspects of Language in a Child with Congenital Disorder of Glycosylation: A Case Study

Language is a very important and unique part of the communication system used by humans. Understanding and producing language in one form or another facilitates interactions with others and allows for an increase in knowledge and awareness. Much research in the field of cognitive neuroscience is concentrated on understanding how language develops and how it is represented in the brain. Such research may have clinical implications by providing a better understanding of a patient's disorder or, more importantly, the possibility to develop and deliver effective interventions (Connolly, Mate-Kole, & Joyce, 1999).

Congenital Disorders of Glycosylation (CDG) are a group of rare genetic disorders with approximately one thousand identified cases in the world to date (Sparks & Krasnewich, 2011). The disorder can have a multidimensional impact on development and may consequently inhibit the proper acquisition of language. Failure to thrive, developmental delay, hypotonia, and seizures are symptoms common to most subtypes, while other subtypes have more unique features such as liver disease, clotting disorders, or cystic kidneys (Sparks & Krasnewich, 2011). While the previous nomenclature consisted of roman numerals and letters, each subtype is now denoted by the name of the affected gene, which reflects the unique enzyme deficiency (e.g., PMM2-CDG; Jaeken, 2011).

At a metabolic level, CDG disrupts glycosylation, a cellular process responsible for the addition of glycans (i.e., sugar chains) to molecules such as proteins and lipids (Freeze, 2006). N-linked and O-linked glycosylation of proteins may be affected, but N-

linked glycosylation may be obstructed more readily as it is a longer pathway with an extra processing step (Jaeken & Matthijs, 2007). N-glycosylation occurs in the cytosol, the endoplasmic reticulum (ER), and the Golgi, while O-glycosylation only begins in the Golgi (Jaeken & Matthijs, 2007). The quality control system of the ER lumen ensures that glycoproteins are properly folded and those that are not will ultimately be degraded (Vleugels et al., 2009). Glycoproteins and glycolipids are essential for the normal growth and function of cells, tissues, and organs, and without them development can be greatly hindered (Varki, 1993).

There are currently 45 subtypes of CDG identified to date, but it is possible that many more will develop in the future as there are over 200 genes involved in the process of glycosylation (Jaeken, 2011; Jaeken, Hennet, Freeze, & Matthijs, 2008). Although some subtypes are more prevalent than others, it is thought that the disorder may be under-diagnosed in general. While clinical features of the disease can even vary within a subtype (Sparks & Krasnewich, 2011), continued research into CDG in a variety of fields will most certainly assist with gaining a better understanding of the disorder and facilitating the identification of the disorder in future patients.

Although most research on CDG has focused on the biochemical aspects of the disorder, the current case study examined certain cognitive features of one participant who has been diagnosed with ALG9-CDG (*CDG-1L*). As the individual is developmentally delayed and non-communicative, the current study assessed aspects of the participant's language abilities through electrophysiological measures. Event-related potentials (ERP) are extracted from electroencephalography (EEG) recordings by time-locking brain responses to external stimuli. Such measures allow for a high degree of

temporal accuracy in the responses detected and do not necessarily require a behavioral response (Connolly, Byrne, & Dywan, 1995). ERP measures can also be used to complement behavioral tests or provide information that is, at times, more precise and informative.

Using ERP measures to assess language in non-communicative individuals has been supported by research. For example, Byrne, Dywan, and Connolly (1995) were able to use ERP measures to show that a patient with Cerebral Palsy (CP) had an age-appropriate vocabulary even though he was unable to be assessed with traditional behavioral measures. The method used was a cross-modal, computerized version of the Peabody Picture Vocabulary Test – Revised (PPVT-R) that had been validated by Connolly et al. (1995). Additionally, Connolly et al. (1999) used ERP measures to show that a patient with a traumatic brain injury was able to process semantic anomalies in sentences similarly to non-brain injured controls. While the measurement of cognitive capacities in participants with motor and language impairments can be extremely difficult due to their inability to physically respond, it is indeed possible to make use of ERP measures for cognitive assessments (Byrne et al., 1995).

There are several ERP responses that are linked to various aspects of one's cognitive abilities, and thus may be used clinically. One highly studied ERP component is the P300, which is elicited to infrequent stimuli that are particularly relevant to a task or participant. The P300 is a broad, positive-going ERP component that is elicited approximately 300 ms after the presentation of a stimulus and has a centro-parietal distribution (Duncan et al., 2009). An oddball paradigm, in which some stimuli are presented more frequently than others, is typically used to elicit a P300. In an oddball

task, stimuli presented less frequently generally lead to a P300 response that is greater in amplitude than the frequently presented stimuli (Duncan et al., 2009).

Research in the past has shown that there can be variations in the latency of P300 responses depending on the nature of the task. A seminal study by Squires, Squires, and Hillyard (1975) showed that two different P300 responses could be elicited to infrequent auditory stimuli: the P3a, peaking between 220 and 280 ms and frontally distributed, and the P3b, peaking between 310 and 380 ms and parietally distributed. Only the P3a was elicited when participants did not attend to the stimuli being presented (i.e., they were instructed to ignore the stimuli and read a book), but the P3a response was more clearly defined for some participants than others (Squires et al., 1975). When stimuli were actively attended and participants counted stimuli as they were presented, the P3b component was elicited in all participants along with a P3a response in most participants (Squires et al., 1975). Those who had small P3a components in the "ignore" condition did not show P3a responses in the "attend" condition (Squires et al., 1975). As most studies require participants to attend and respond to stimuli, references to a P300 component most often refer to the P3b response.

P300 responses can be elicited even in the absence of a task during the presentation of stimuli (e.g., behavioral responses, mental operations). Bennington and Polich (1999) presented young adults with auditory and visual stimuli using an active task, in which participants responded to target stimuli, as well as a passive task, in which participants attended to the stimuli but did not respond in any way. However, they did not have their participants ignore stimuli in the passive task as in the study by Squires et al. (1975). While P300 amplitudes were greater for the active task in comparison to the

passive task, P300 responses were larger in the auditory modality for the passive task than in the visual modality (Bennington & Polich, 1999).

Particularly relevant to the current study is research conducted by Berlad and Pratt (1995) that showed the P300 could be used as a marker of familiarity in a name recognition task. Adult participants were tested in an active and passive task, the latter of which was broken down into two tasks. Berlad and Pratt's first passive task had the participant's name played 20% of the time, while another word matched for length and other characteristics was played more frequently at a probability of 80%. In the second task, Berlad and Pratt had the participant's name played 20% of the time, but there was another word played at a probability of 20% along with a word played 60% of the time. They found that the average P300 response was larger to the name of a participant than to the high probability word (i.e., word played 60% or 80% of time) or the other low probability word in the second task (i.e., word played 20% of time). Berlad and Pratt discussed that the larger P300 to the participants' own names in comparison to the other low probability word demonstrates that the relevance of a stimulus has an effect on the amplitude of the P300 beyond the probability of its presentation, as one's name is particularly relevant to him or her.

Related to Berlad and Pratt's (1995) study is a name recognition experiment by Folmer and Yingling (1997) that used a slightly different paradigm in adults. In the first experiment, each participant heard 30 repetitions of his or her own first name along with 80 other first names of the same gender matched for intensity, duration, and number of syllables. In the second experiment, which acted as a control for the first, participants heard 30 repetitions of a random name and 80 other first names, as in the first

experiment. After comparing the three conditions (i.e., name, other name, assorted names), Folmer and Yingling demonstrated that the strongest P300 responses were elicited to the participants' own names.

The P300 response to one's own name can even be detected in brain-injured patients with various states of consciousness. Perrin, Goldman, and Moonen (2006) conducted a study using a name recognition task in coma survivors with five patients who were in a vegetative state (VS), six who were in a minimally conscious state (MCS), and five who had locked-in syndrome (LIS). Perrin et al. presented eight names to each participant with one of the names being the patient's own name. With the exception of one LIS patient and two VS patients, all participants showed a significantly larger P300 to their own names than the other names presented (Perrin et al., 2006). A larger P300 to one's own name may reflect familiarity with the name, but it may also imply that some linguistic abilities are intact (Perrin et al., 2006).

While the majority of studies involving the P300 have used adult participants, there has been research showing that a P300 can be elicited in younger populations. Using simple auditory stimuli (i.e., tones), McIsaac and Polich (1992) demonstrated that infants as young as 5 months old are able to show P300 responses that are largest over centro-parietal sites. In another study using tones, Ladish and Polich (1989) showed that young children (i.e., 5 to 9 year olds) are able to show P300 responses as well. These two groups of researchers both showed that the P300 component increases in amplitude and decreases in latency with age. Parise, Friederici, and Striano (2010) conducted the only known study that has used a name recognition task on a younger population. In this task, the participants heard their own name in equal probability to either one or ten names of

strangers. Parise et al. found that participants' own names elicited a significantly more positive component over anterior sites between 100 and 380 ms in comparison to names of strangers when only one stranger's name was presented. In addition, between 200 and 600 ms, there was a significantly more negative response to participants' own names in comparison to strangers' names over parietal channels in the condition where ten were presented. This response was somewhat expected by Parise et al., as Thierry, Vihman, and Roberts (2003) had previously demonstrated that infants elicit a greater negativity, rather than a positivity, to familiar words in comparison to unfamiliar words.

Another ERP component known as the mismatch negativity (MMN) is particularly useful in the assessment of children and clinical populations as it is a response that can be detected in the absence of attention (Näätänen, 1992). The MMN is a negative-going ERP component with a fronto-central distribution that peaks between 100 and 250 ms (Näätänen, 1992). MMN paradigms have participants listen to many repetitions of one stimulus (i.e., the standard) interspersed with fewer repetitions of another stimulus (i.e., the deviant). The amplitude of the brain's response to deviant stimuli is generally greater than that to standard stimuli. The MMN response itself is a difference waveform reflecting this heightened response to deviant stimuli, and is obtained by subtracting the average response to standard stimuli from that of deviant stimuli (Morr, Shafer, Kreuzer, & Kurtzberg, 2002).

The MMN response can be elicited to very small acoustic changes in auditory stimuli, but the difference in sound must generally exceed a certain level (Näätänen, Paavilainen, Rinne, & Alho, 2007). Results from a study by Sams, Paavilainen, Alho, and Näätänen (1985) showed that MMN difference waveforms were clear only when the

frequencies were high enough to be behaviorally discriminable from the standard. The deviant that was on the threshold only yielded a very small MMN (Sams et al., 1985).

The maturation of the MMN is not well understood due to somewhat inconsistent results. It has been shown that an MMN response can be detected in infants, as in a study by Alho, Sainio, Sajaniemi, Reinikainen, and Näätänen (1990), which demonstrated that sleeping newborns elicited broad MMN responses to differences in pitch. However, what is considered to be an MMN response in participants this young may not always be the same as the MMN elicited in adults. For instance, some, but not all infants will show positive-going components instead of negative-going ones (Kushnerenko, Čeponiene, Balan, Fellman, & Näätänen, 2002; Morr et al., 2002; Rivera-Gaxiola, Silva-Pereyra, & Kuhl, 2005). A study conducted by Morr et al. (2002) with infants and preschoolers (i.e., those aged between 2 and 47 months) found that while a 1000 Hz difference between the standard and deviant elicited adult-like MMN responses in almost all participants, a smaller 200 Hz difference elicited a negativity in only some older participants, but a positivity in most younger infants. Additionally, the latency of the MMN was found to decrease at a rate around 1 ms per month.

Using slightly older participants, a study conducted by Bishop, Hardiman, and Barry (2011) showed that MMN responses could be detected in children (i.e., 7 to 12 years old) and adolescents (i.e., 13 to 16 years old), but that the mean amplitude of this component increases with age. Shafer, Morr, Kreuzer, and Kurtzberg (2000) also found that children aged four to 10 years old elicited MMN responses, although they were larger in amplitude than those in adults. Conversely, Maurer, Bucher, Brem, and Bradeis

(2003) demonstrated that school-aged children (i.e., 6 and 7 year olds) consistently showed a positive mismatch response rather than the typical MMN seen in adults.

Aside from simple paradigms such as those using tones, many researchers have examined MMN responses to basic speech sounds, such as phonemes (e.g., /*b*/ as the standard vs. /*p*/ as the deviant). Evidence for language-specific memory traces to phonemes can be detected beginning around 1 year of age (Cheour et al., 1998). Adult data further support the notion of language-specific representations in the brain, as MMN responses to phoneme deviants from a first language have been found to be enhanced in comparison to phoneme deviants from another language (Näätänen et al., 1997).

While MMN responses to phonemes reveal that there are basic language representations in the brain, many studies have gone even further to show that there are differences in MMN responses to words in comparison to non-words. Korpilahti, Krause, Holopainen, and Lang (2001) conducted a study with children using tones, words, and pseudowords. A late mismatch negativity (lMMN; 350-500 ms) could be detected in addition to the early mismatch negativity (eMMN; 150-350 ms), which is roughly the timeframe of a typical MMN response. Tones elicited a significantly larger eMMN than words, but words elicited a significantly larger lMMN in comparison to pseudowords. The lMMN to words tended to also be larger in amplitude than tones, but the difference was not significant.

In an adult study examining the difference between word and pseudoword deviants, Pulvermüller et al. (2001) presented native Finnish-speaking adults with disyllabic stimuli. All standard stimuli were pseudowords with one condition containing word deviants and the second consisting of pseudoword deviants. Due to the nature of the

Finnish language, Pulvermüller et al. were able to make the onsets of the stimuli identical within a condition and the endings (i.e., second syllables) the same across conditions. The endings were what differentiated a particular stimulus into a word or a pseudoword. The ERPs were measured to the onset of the last phoneme, which was considered to be the divergence point. It was found that word deviants elicited MMN components that were significantly greater in amplitude than pseudoword deviants. Because there was an enhancement of the MMN responses to words that was beyond any acoustic differences, these results imply that long-term memory traces for words exist (Pulvermüller et al., 2001).

Shtyrov and Pulvermüller (2002) later conducted a study using English monosyllabic stimuli in a group of healthy adults to see if the enhanced MMN response detected in the study by Pulvermüller et al. (2001) was simply due to lexical differences between the standard and deviant. Using three different conditions, Shtyrov and Pulvermüller employed English monosyllabic stimuli. In the first condition, the standard was a word, /*taɪp*/ (i.e., *type)*, while the deviant was also a word, /*taɪt*/ (i.e., *tight*). In the second condition, the standard was a pseudoword, /*baɪp*/, while the deviant was a word, /*baɪt*/ (i.e., *bite*). Finally, in the third condition, the standard was a word, /*paɪp*/ (i.e., *pipe*), and the deviant was a pseudoword, /*paɪt*/. All characteristics of the stimuli were carefully controlled, with onsets matched for formant frequencies, word duration kept as similar as possible, and endings cross-spliced onto each onset.

Results of the study by Shtyrov and Pulvermüller (2002) demonstrated that the findings of Pulvermüller et al. (2001) were not because of lexical differences. MMN responses were enhanced to words in comparison to pseudowords; however, Shtyrov and

Pulvermüller also found that there was no significant difference between the word deviants, which either had a pseudoword or a word standard. This study further supports the hypothesis that an enhanced MMN response is related to long-term memory traces for words (Shtyrov & Pulvermüller, 2002).

The current study used two experiments designed to elicit the P300 and MMN components, respectively, in order to assess aspects of language functioning in a child with CDG. In addition to the CDG participant, typically developing children aged 4 to 6 years old were included in the study for comparison. Although some of the participants in the comparison group were around the same age as the CDG participant, a range including younger participants was selected due to the developmental delays of the patient. According to the patient's neurologist, traditional forms of assessment have been unable to pinpoint her developmental age; therefore, a comparison group matched on cognitive abilities was not possible. The two experiments in the current study were designed to fit the CDG participant's apparent abilities. Specifically, due to the participant's poor visual acuity and limited motor skills, the tasks were auditory and did not require behavioral responses.

During the name recognition experiment, recordings of eight different names, including the participant's own, were played randomly to each participant. The MMN experiment was very similar to that of Shtyrov and Pulvermüller (2002); English monosyllabic words were used as stimuli along with one pseudoword. In the first condition, participants heard a word as the standard and a second word as the deviant. In the second condition, a different word was the standard stimulus while a pseudoword was

the deviant. The MMN difference waveforms were compared across conditions to see if there was a larger difference to word deviants.

Due to the early age at which a P300 response can be elicited to simple auditory stimuli (Ladish & Polich, 1989; McIsaac & Polich, 1992) and the fact that P300 responses can be elicited to participants' own names (Berlad & Pratt, 1995; Folmer & Yingling, 1997; Perrin, Goldman, & Moonen, 2006) it was hypothesized that typically developing children would show, on average, enhanced P300 responses to their own names. While infants showed negative-going ERP responses in the name recognition task conducted by Parise et al. (2010), it was still expected that older children in the current study would elicit enhanced P300 responses to their own names in comparison to other names.

For the MMN experiment, it was expected that there would be a difference in the MMN responses such that the amplitude of the MMN would be greater for the word deviant than the pseudoword deviant as seen in the study by Shtyrov and Pulvermüller (2002). However, the exact nature of the MMN component (negative or positive-going) was not predicted due to the variability of responses observed for young children, including those that are school-aged (Maurer et al., 2003).

Data collected from the typically developing children in the current study were used as a comparison for the ERP responses elicited by the CDG participant. The objective of the current study was to see if the CDG participant could recognize her own name and show evidence of long-term memory traces for words. To date, no one, including medical professionals, has been able to determine whether or not she is able to detect her name or distinguish between words and pseudowords. Therefore, while it was

not entirely comprehensive, the current study was the first true assessment of a fairly basic level of her language ability. Results could also be useful to those in the field as CDG is such a rare disease and case studies are important for better understanding its wide range of effects on physical and cognitive development.

Method - Experiment 1

Participants

Comparison Group. Eleven typically developing children participated in the study. Three of the participants were males and the age of participants ranged from 4 years 4 months to 6 years 7 months. The mean age of participants was 5.52 years. The aim was not to recruit children of the same chronological age, as it was known that the CDG participant had developmental delays. Although it was not possible to determine the patient's developmental age, the lower end of the range was selected based on the idea that the best data could be obtained from children aged 3 years old and up. All participants had normal or corrected-to-normal hearing, were right-handed, were native English speakers (i.e., English is the first language they learned at home), and had no history of learning disabilities, psychiatric illnesses, or neurological illnesses.

Recruitment of participants was made possible through the use of advertisements (e.g., posts in local bulletins) and word of mouth in the local community surrounding Acadia University, which is a region that is different from the community in which the CDG participant resides. Interested parents could contact the researchers via e-mail or phone in order to participate in the study. An experimenter then conducted a brief screening by e-mail or phone with those interested in the study to ensure that their

children met the criteria for participation. Monetary compensation of ten dollars per hour was given to the participants and they also had the opportunity to choose a small prize.

Patient RS. A single participant with Congenital Disorder of Glycosylation (CDG), RS, was the clinical case of the current study. RS was 5 years 10 months at the time of testing and is a young female who has been raised in an English-speaking household. At 3 months of age, RS presented with symptoms of failure to thrive, developmental delay, vomiting, and watery stools, which were followed by a generalized tonic seizure at 4 months. She had mild dysmorphic features, a bulbous tip to the nose, and slightly low set ears. Imaging of the brain and bowel revealed no structural abnormalities. She was found to have a small pericardial effusion, which subsequently resolved. At age 6 months she started having infantile spasms and was found to have hypsarrhythmia on EEG (i.e., random high-voltage slow waves and spikes that spread to all cortical areas).

CDG was suspected clinically and was confirmed at 2 years 3 months by serum transferrin isoelectric focusing, a blood test which determines whether or not there are abnormalities in the addition of glycans for the transferrin glycoprotein. Test results showed a pattern of CDG-1 on three specimens from the patient, but results were normal in the parents. RS's specific subtype was determined to be ALG9-CDG (*CDG-1L*), whereby ALG9 refers to a specific enzyme. This was due to an accumulation of the lipid-linked oligosaccharides $Man_6GlcNAc_2$-PP-dolichol and $Man_8GlcNAc_2$-PP-dolichol, which is a finding indicative of an ALG9 enzyme deficiency. Subsequent direct sequencing of the *ALG9* gene revealed a homozygous p.Y286C (c.860A>G) mutation that confirmed the diagnosis.

At the age of EEG testing, the patient continued to have severe developmental delay. She appeared alert, happy, and was somewhat socially responsive. She turned to voices but showed no shyness with strangers and no interest in herself in a mirror. She made different sounds, but no recognizable words, and did not point or communicate non-verbally. It was difficult to judge if she had any understanding of words. She readily followed large objects or people with her eyes and would reach out to grasp them; however, her handedness was not clear. She did not respond to small objects but was sensitive to sound. She was able to sit without support but was unable to stand.

According to RS's parents, she has continued to have focal seizures one to two times per month, and was on treatment with clobazam and levetiracetam at the time of testing. Her EEG showed some interictal discharges in the left centro-parietal and right fronto-central regions, and the background activity was slow (4-5 Hz) compared to typically developing children of her age. RS received ten dollars per hour for her participation and also received a small toy.

Materials & Stimuli

Comparison Group. Each participant had a set of eight different names prepared with his or her own name and seven other same-syllable names matched for duration and loudness. Three of the other names were considered to be of the same gender as the participant's own name and four were considered to be of the opposite gender. These names were ones that the participant would not hear on a regular basis as determined in a brief screening with a parent or guardian some time before the study. However, the parents may have forgotten some familiar names when they gave their lists, allowing those to be included by chance. One parent noted to the researchers that one name in

particular would have been familiar to her child, who had mentioned some of the names heard during the experiment. All names were recorded using Adobe Audition CS5.5 software digitized at 16-bit and 44.1 kHz. While each of the eight names in a list was coded separately for presentation, the participant's own name corresponded to the "self" condition and all other names were part of the "other" condition. Participants were tested in the Cognitive Neuroscience Laboratory at Acadia University and sat in a comfortable chair as they listened to sounds played to them through ER-1 insert earphones.

During testing, a 32-channel Waveguard electrode cap was positioned on the participant's scalp and was connected to an electroencephalogram (EEG) amplifier. The amplifier was connected to two laptop computers: one presenting the stimuli through E-Prime 2.0 software and another that was receiving the data using Advanced Source Analysis (ASA) 4.7.9 software. Electrogel was used to fill the electrodes that were embedded in the cap using a blunt needle and wooden sticks were also used to help lower the impedances. The mastoids were cleaned using an exfoliating gel, NuPrep, to lower impedances. Further details regarding the electrophysiological setup are given below.

Patient RS. Prior to conducting the study, the parents of RS also provided a list of names that were familiar to RS and those names were not included as one of the seven others played in the study. However, RS's parents told the researchers after the experiment that one of the names included, Name 3, might have been familiar to RS as it was the name of a girl who lives on their street. The seven other names that were chosen consisted of the same number of syllables as RS's and were matched closely for duration and loudness (i.e., amplitude). Three of the other names were female and four were male.

RS was tested in her home and remained in her wheelchair for the duration of the experiment. RS's parents indicated she would most likely find headphones uncomfortable. Therefore, to minimize any stress RS may have experienced, sounds were played to her over speakers.

Procedure

Comparison Group. When a participant entered the laboratory, he or she was greeted by the experimenters and was asked to sit in the comfortable chair. Their parents read and signed the consent form before beginning (see Appendix A) while the participants gave verbal assent and were told that they could stop at any point. Each participant watched part of a movie as experimenters prepared the EEG cap by properly fitting it onto the participant's head and inserting electrogel into the appropriate electrodes. Once all of the equipment was set-up and functional, the movie and the lights were turned off. The participants were then instructed to listen for their own name and the two lists of names prepared for the participant were played. There was an inter-trial interval of 1000 ms between each name. There were 40 repetitions of each name overall for a total of 320 stimuli. The trials were split into two lists, with 160 names in each list and a short break in between. Within each list there were 20 repetitions of each name and the order was pseudo-randomized so that no more than two repetitions of the same name would occur in a row.

Patient RS. When the experimenters arrived to RS's house, the laptop computers and equipment were first set up. Consent was also obtained at this point as RS's parents read and signed the consent form (see Appendix B). RS remained in her wheelchair and the appropriate-sized cap was chosen and prepared. Music was played to help keep RS

relaxed as the electrodes were filled and experimenters tried to get impedances as low as possible. Once the equipment was set up and the lights were turned off, RS was told to listen for her name and had the two lists of stimuli played to her with a short break between each. While it could not be determined if RS understood the instructions to listen for her name, Squires et al. (1975) demonstrated that a P3a response can be detected even if stimuli are ignored by participants.

EEG Acquisition

Comparison Group. EEG data were collected for typically developing children at 512 Hz using a 32-channel Waveguard cap containing Ag/AgCl electrodes. Only 16 of these electrodes were used: Fp1, Fpz, Fp2, F3, Fz, F4, C3, Cz, C4, T7, T8, P3, Pz, P4, M1, and M2. An electrode embedded in the cap, AFz, served as the ground. M1was used as the reference electrode during recording and data were filtered using a 60 Hz notch filter and a band-pass filter of 0.1-100 Hz. Impedances were kept below 10 kΩ, with the exception of one participant who had slightly higher impedances at some channels despite clean waveforms. This participant's data were still included.

Patient RS. As with the comparison group**,** EEG was recorded at 512 Hz using a 32-channel Waveguard cap. However, Fp1, Fpz, and Fp2 were not used for the CDG participant, leaving only 13 channels to be filled in addition to the ground electrode: F3, Fz, F4, C3, Cz, C4, T7, T8, P3, Pz, P4, M1, and M2. M1 was also the reference during recording and the same filters were used. The impedances at all channels were verified as being acceptable, but were not all below 10 kΩ.

ERP Analyses

Comparison Group. BrainVision Analyzer 2 software was used to analyze and edit the data to make the average waveforms as clear and reliable as possible. The average output from the mastoids (i.e., M1 and M2) was used for a reference in creating the waveforms to be analyzed. Data were not analyzed at the mastoids, Fp1, Fpz, or Fp2, which left 11 sites to be analyzed: F3, Fz, F4, C3, Cz, C4, T7, T8, P3, Pz, and P4. Data were epoched from 100 ms before stimulus onset to 800 ms after and the band-pass filter was reduced to 0.1-20 Hz (24 db/oct). A pre-stimulus period of -100 to 0 ms was used for the baseline correction.

All segments that were greater than ±100 μV were automatically rejected from all analyzed channels. The percentage of rejected trials ranged from 12.5% to 60% for each participant in each condition. One participant had 77.5% of trials rejected, which was considered to be overly high; therefore this participant was subsequently excluded from analyses for the first experiment. Data were averaged across participants for each condition and site. To reduce the signal to noise ratio (SNR), one of the seven names in the other condition was randomly selected for each participant to create a grand average of trials for that condition. It was ensured that the name familiar to one participant was not chosen. The mean amplitude for the ERP response to names in the self condition as well as the other condition was scored for each channel based on the average amplitude between 250 and 400 ms.

Patient RS. In general, the same parameters were used for analyses of RS's data as those used for typically developing children. However, RS's background EEG consisted of slower frequency activity, particularly in the range of 4-5 Hz. Therefore, the

band-pass filter was adjusted to 1.6-20 Hz (24 dB/oct) for RS. The same sites were analyzed as typically developing children.

If the amplitude of a particular ERP segment was greater than ±100 μV, it was considered to be an artifact and was rejected. Following artifact rejection, 24 trials remained for RS's own name and 175 for the other trials in total. There were a high percentage of rejections as RS tended to move her eyes and frequently touched and moved her head during testing. Trials were averaged for each condition, with the names in the other condition averaged separately as well as all together. Because the SNR was much higher for the grand average of all other names in comparison to the average for RS's own name, four trials from each of the seven names were randomly selected to better equate the SNR for the two conditions.

Results – Experiment 1

Comparison Group. To determine if the typically developing children who participated in the study showed P300 responses themselves, analyses were conducted separately for their data. Figure 1 shows a comparison of the grand average waveforms obtained from the self and other conditions across the various analyzed sites for control participants. It was clear from visual inspection of the waveforms that the expected trend of a P300 that was larger to the participants' own names was not present. Conversely, a negative component was elicited to participants' own names between 250 and 400 ms.

Due to the inconsistency between the elicited waveforms and what was expected, data were not analyzed solely over centro-parietal sites, where P300 responses are typically greatest according to the literature (Duncan et al., 2009). Instead, a 2x9

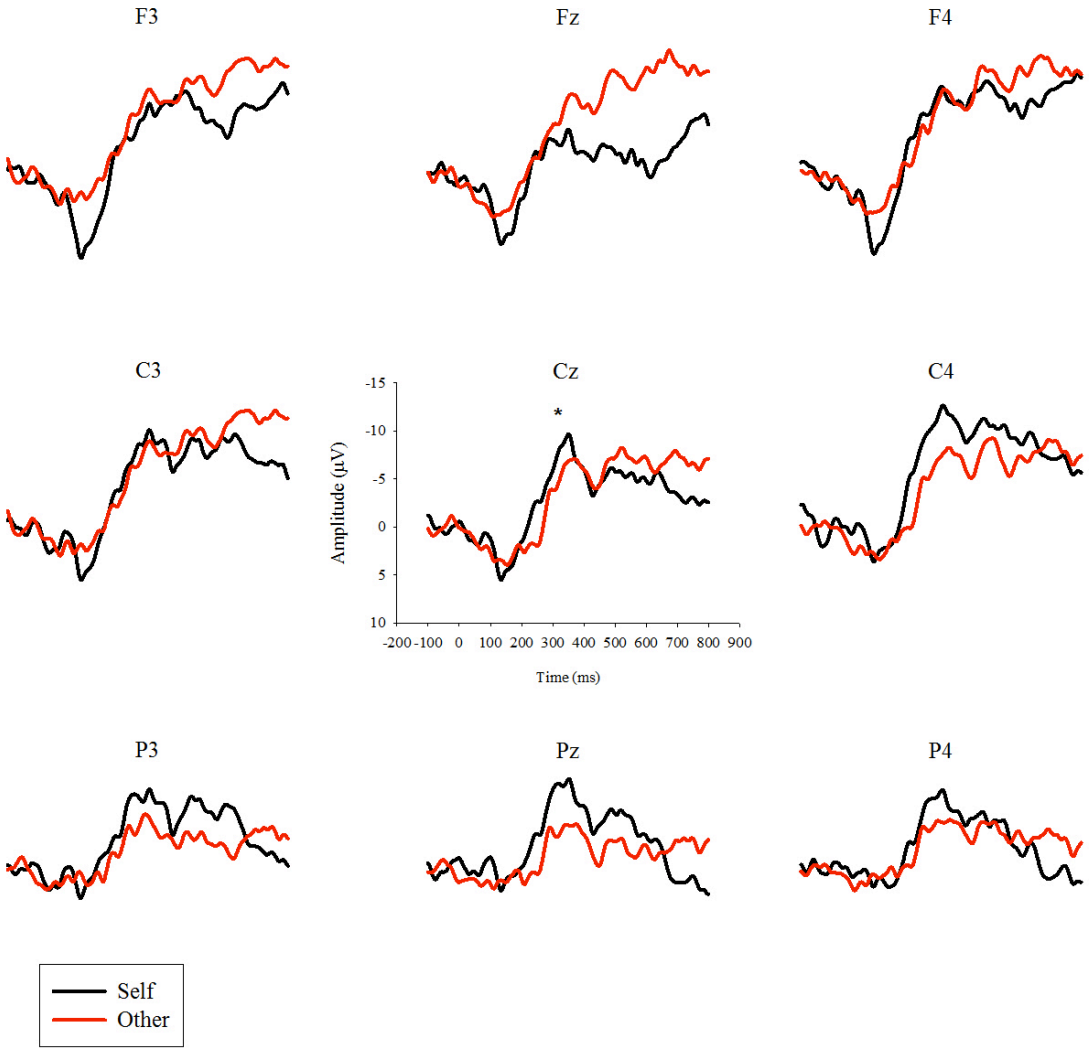

Figure 1. Grand average event-related potential waveforms ($N = 10$) elicited by the comparison group to the auditory stimuli in the self (black line) and other (red line) conditions. Electrodes are displayed in relation to how they were positioned on the scalps of participants with anterior sites plotted at the top of the figure. Asterisk indicates the point in time when the difference in ERP activity between conditions occurred.

repeated-measures factorial ANOVA was conducted including frontal sites as well in order to examine the main effects and interactions between the two within-subjects factors: condition (two levels: self and others) and site (nine levels: F3, Fz, F4, P3, Pz, P4, C3, Cz, and C4). There were no main effects of condition, $F(1, 9) = 0.73$, $p = 0.416$, or site, $F(8, 72) = 1.42$, $p = 0.203$. However, there was a significant Condition x Site interaction, $F(8, 72) = 3.61$, $p = 0.001$.

To further analyze the significant Condition x Site interaction, two paired-samples t-tests were conducted to compare the amplitude of the negative-going component between the self and other conditions at two electrode sites: C4 and Pz. These sites were chosen based on visual inspection of the grand averaged waveforms, which showed that the largest differences between conditions appeared to be at these two sites. Results revealed that while the negativity in the self condition at C4, $M_{self} = -9.89$, was not significantly different than that observed in the other condition at C4, $M_{other} = -5.51$, $t(9) = -1.94$, $p = 0.084$, the negativity at Pz for the self condition, $M_{self} = -7.22$, was significantly larger (i.e., more negative) than that observed in the other condition, $M_{other} = -3.43$, $t(9) = -2.30$, $p = 0.047$.

Patient RS. Statistical analyses were not conducted on the ERP data collected from RS and, instead, data were assessed qualitatively by means of visual inspection. Figure 2 compares the grand average waveform for RS's own name to each of the seven individual grand average waveforms for the names in the other condition as well as the grand average waveform for the other condition with a few trials from each name averaged together.

Figure 2. Grand average event-related potential waveforms elicited by RS to the auditory stimuli in the self (black line) and other (red line) conditions. Comparisons are made between each of the other names as well as the grand average for all names. Electrodes are displayed in relation to how they were positioned on RS's scalp with anterior sites plotted at the top of the figure. Asterisk indicates the occurrence of the early negativity; double asterisk indicates the occurrence of the later negativity.

RS appears to have been distinguishing between her name and the other names presented. Within the broad range of 300 to 600 ms RS's own name elicited a response that is more negative in amplitude than the responses to the names in the other condition for most comparisons. However, the difference was clearest for the comparisons between RS's own name and conditions Name 2, Name 3, Name 4, Name 6, and All Names. While the trend for RS's own name to elicit a greater negativity between 300 and 600 ms can be detected across sites, the difference is generally largest at Fz. It is important to note that this negative-going response is comparable to the response elicited by typically developing children between 250 and 400 ms, but the comparison group had a significant difference between conditions at Pz. Additionally, unlike the typically developing children, RS also showed an earlier negativity between 100 and 300 ms to the other condition in contrast to her own name for most comparisons. This negativity tends to be slightly more noticeable at Cz and Pz than at Fz.

Discussion – Experiment 1

The data from the name recognition task in Experiment 1 were not consistent with what was hypothesized. Instead of the typically developing children showing a P300 to their own names, they elicited a negative-going response between 250 and 400 ms that was larger in amplitude to their own name in comparison to the other names. The difference between own and other names was found to be largest at Pz. The negativity exhibited by the typically developing children was similar to a response detected over parietal sites in infants in a name recognition task conducted by Parise et al. (2010). The finding of a negativity for participants' own names also fits with the work of Thierry et al. (2003) on infants, which found a negativity elicited to familiar words around 200 ms,

but not to unfamiliar words. The participants' own names would certainly be familiar to them and the names included in the other condition should not have been nearly as familiar, as a pre-screening with parents ensured they did not hear such names on a regular basis. While there have not been any name recognition studies conducted to date that have included young children, it has been shown that infants and young children elicit a P300 to tones and that the P300 response matures with age (Ladish & Polich, 1989; McIsaac & Polich, 1992). Therefore, it was expected that the young children in the current study would show responses to names that were more like those of adults than infants. Perhaps it takes longer for the P300 to mature to names and words than it does to tones.

In terms of RS's performance on the name recognition task, there was a clear negativity between 200 and 350 ms that was larger for her own name in comparison to the other names for five of the eight comparisons. Although the earlier negativity she showed to the other names between 100 and 300 ms compared to her own name was not seen in the grand average data for the comparison group, the negativity she elicited to her own name was very similar to the responses of the typically developing children. Interestingly, the later negativity elicited by RS was larger over frontal sites and, although still apparent, was not as distinct over parietal channels as it was in the comparison group or in the study by Parise et al. (2010). CDG is a diffuse disorder that affects the entire brain, which may have led RS to use slightly different mechanisms, possibly supported by another part of the brain, compared to typically developing children. Despite these subtle differences, the results suggest that RS recognizes her own name and can distinguish it from others.

Method – Experiment 2

Participants

The same eleven participants from the comparison group in the first experiment took part in the second experiment along with RS.

Materials & Stimuli

Comparison Group. All stimuli used in the second experiment were monosyllables and all words were determined to be high frequency at the grade one level (i.e., above 20 occurrences of 584,693 total instances in grade appropriate books) using the Educator's Word Frequency Guide (Zeno, Ivens, Millard, & Duvvuri, 1995). There were three English words used, /*baɪt*/ (i.e., *bite*; frequency of 54), /*baɪk*/ (i.e., *bike*; frequency of 173), and /*naɪt*/ (i.e., *night*; frequency of 716), and one pseudoword, /*naɪk*/.

The stimuli fell into two conditions. In the word condition, /*baɪt*/ was the standard while /*baɪk*/ was the deviant. In the pseudoword condition, /*naɪt*/ was the standard whereas /*naɪk*/ was the deviant. Different lexical contrasts were created using the deviants; there was no lexical contrast for the word condition as the standard and deviant were both words, but a lexical contrast was present for the pseudoword condition as the standard was a word and the deviant was a pseudoword.

An adult female English speaker recorded the stimuli using Adobe Audition CS5.5 software. To keep stimuli identical within a condition until their divergence points (i.e., point where the two stimuli differentiate), the onsets of /*baɪk*/ and /*naɪk*/ were used after being separated from their terminal plosive consonant, /*k*/. The onset /*baɪ*/ had a duration of 294 ms while the onset /*naɪ*/ had a duration of 380 ms. The onsets were then each cross-spliced with the terminal plosive consonants /*t*/ and /*k*/ that were taken from

two recorded words not included in the study, /*taɪt*/ and /*taɪk*/ respectively. Divergence points within each condition occurred at the beginning of the terminal consonant (i.e., after 294 ms for the word condition and after 380 ms for the pseudoword condition). First and second formant frequencies were kept as similar as possible across conditions.

A well-known study conducted by Ganong (1980) indicates that participants will not identify nonwords as words unless the stimuli are ambiguous (i.e., the phonemes are not clear). Therefore, stimuli used in the MMN portion of the study were first piloted to ensure that the pseudoword included, /*naɪk*/, would be identified as such.

The sounds were played through the same ER-1 insert earphones that were used in the first experiment. The devices and computer software used remained the same as well. Furthermore, participants watched a muted movie on a laptop computer placed on a table in front of them during the presentation of auditory stimuli.

Patient RS. The stimuli that were presented to RS were identical to the stimuli presented to the comparison group. RS remained in her wheelchair for the second experiment and the sounds were played through speakers. During the presentation of the auditory stimuli, RS watched a muted show on a large television on the wall in front of her. The same devices and software were used to present the stimuli as in the first experiment.

Procedure

Comparison Group. The typically developing children received a short break before having the stimuli played to them. The stimuli were presented in two lists, with one list consisting of the stimuli from the word condition and the other consisting of the stimuli from the pseudoword condition. The presentation of lists was semi-

counterbalanced across participants, with six of the eleven hearing the list for the word condition first and the other five hearing the list for the pseudoword condition first. However, the participant who was excluded had heard the pseudoword list first, leaving only four participants who heard the pseudoword word list first. Each list consisted of 1000 stimuli with a 500 ms inter-trial interval between stimuli. Each list had 85% of the stimuli as standards and 15% of the stimuli as deviants. The stimuli in each list were presented in a pseudo-random order so that there were two or more standards between each deviant.

The participants watched a muted movie as the stimuli were presented; however, one participant did not want to watch a movie and instead sat quietly as the stimuli were presented. Once the experiment was finished, the cap was removed and participants and their parents/guardians were given a debriefing form (see Appendix C), compensated, and the participants were allowed to choose a prize.

Patient RS. The CDG participant received a short break before having the two lists of stimuli played to her as well. The stimuli from the word condition were presented first and then the list containing stimuli from the pseudoword condition was played second. RS watched a muted TV show her mother selected as the stimuli were presented. After the study was finished and the cap was removed, RS's parents were compensated, given a debriefing form (see Appendix D), and were allowed to ask any questions they had. RS also received her prize for participating at that time.

EEG Acquisition

The same settings were used for the recording of data in the second experiment as in the first experiment for RS and the comparison group.

ERP Analyses

Comparison Group. The same parameters were used to analyze the data as the first experiment with the exception that the waveforms were epoched from 100 ms before the divergence point (i.e., beginning of the terminal consonant) to 800 ms after. One participant had stopped halfway through the experiment after the first list of stimuli was presented. This participant's data were removed, as a complete dataset was required from each participant for analysis. Of the remaining participants, between 0.7% and 45% of trials were rejected for each participant in each condition. Data were averaged across remaining trials for each condition and site. MMN difference waveforms were obtained for each condition by subtracting the standard grand average waveforms from those of the deviants. The mean amplitude of the MMN for each condition between 100-150 ms was measured for each channel.

Patient RS. The criteria used to analyze the data were the same as those used for the comparison group. Trials were again rejected automatically, with up to 13% of trials rejected for the word condition and up to 33% of trials rejected for the pseudoword condition (i.e., only 132 trials remained for /*baɪk* /, 736 for /*baɪt* /, 100 for /*naɪk*/, and 649 for /*naɪt*/). Artifacts may have been due to factors such as movement, blinking, and muscle spasms, which were unable to be controlled, as RS wouldn't respond to requests to remain still. The list of stimuli in the pseudoword condition were also played last, which may have led to boredom, more movement, and consequently more rejected trials. MMN difference waveforms were also obtained in a similar fashion as for the typically developing children.

Results – Experiment 2

Comparison Group. Figure 3 demonstrates the grand average waveforms for the typically developing children from the word condition and the pseudoword condition plotted together across fronto-central sites. An MMN response can be seen between 100-150 ms for both conditions over frontal and central sites. A 2x6 repeated-measures factorial ANOVA was conducted to examine the main effects and interactions between the two within-subjects factors: condition (with two levels: word and pseudoword) and site (with six levels: F3, Fz, F4, P3, Pz, and P4). Only fronto-central sites were used for the analyses, as MMN responses are typically larger over those sites (Näätänen, 1992). Mauchly's test of sphericity was violated; therefore, Green-Geisser *F*-values and *p*-values are reported (Greenhouse & Geisser, 1959). There were no main effects of condition, $F(1, 9) = 0.01$, $p = 0.940$, or site, $F(5, 45) = 0.40$, $p = 0.649$. Additionally, the Condition x Site interaction was not significant, $F(5, 45) = 0.37$, $p = 0.665$. Therefore, even though MMN responses were elicited to deviant stimuli between 100 and 150 ms in each condition, the deviant in the word condition did not elicit a response that was greater in amplitude than the deviant in the pseudoword condition as hypothesized.

Patient RS. As in the first experiment, statistical analyses were not conducted for RS's data and visual inspection was used instead. Figure 4 shows a direct comparison of the waveforms elicited in the two conditions across fronto-central sites. The data were very noisy at Pz and P4 due to artifacts from RS moving and touching her head; therefore, parietal channels were not included. Unlike the typically developing children, RS did not show an MMN response to the deviant stimuli in the word condition. However, there was an MMN-like component elicited to the pseudoword deviant

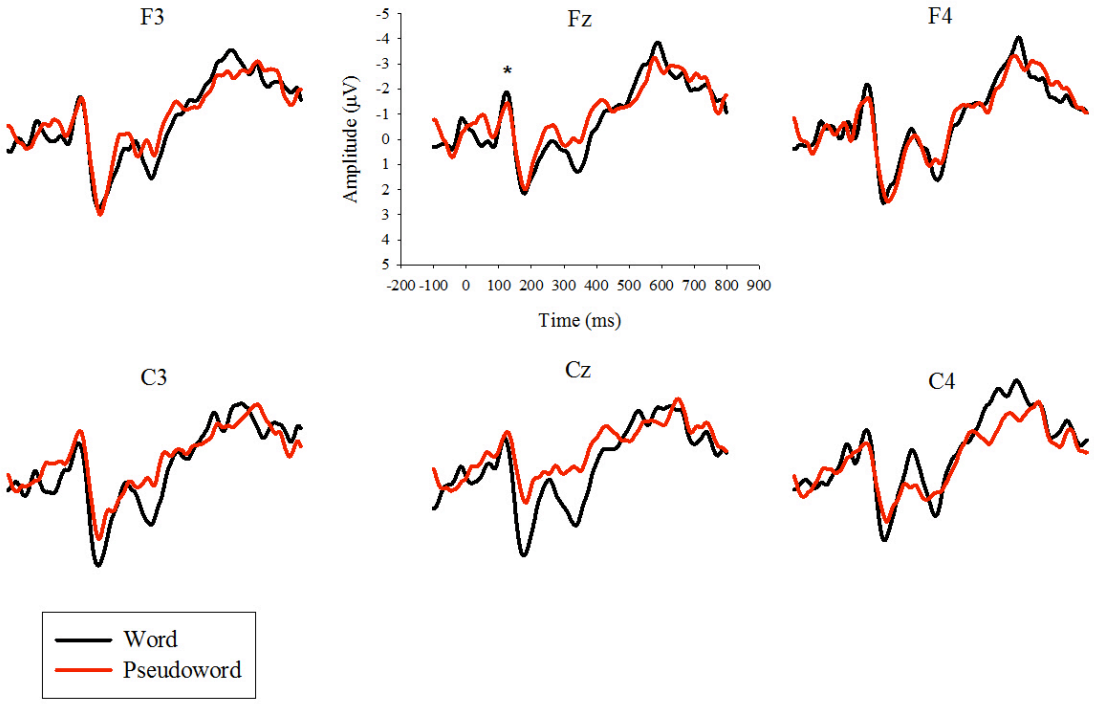

Figure 3. Grand average event-related potential waveforms (N = 10) elicited by the comparison group to the auditory stimuli in the word (black line) and pseudoword (red line) conditions. Electrodes are displayed in relation to how they were positioned on the scalps of participants with anterior sites plotted at the top of the figure. Asterisk indicates the occurrence of MMN responses in both conditions.

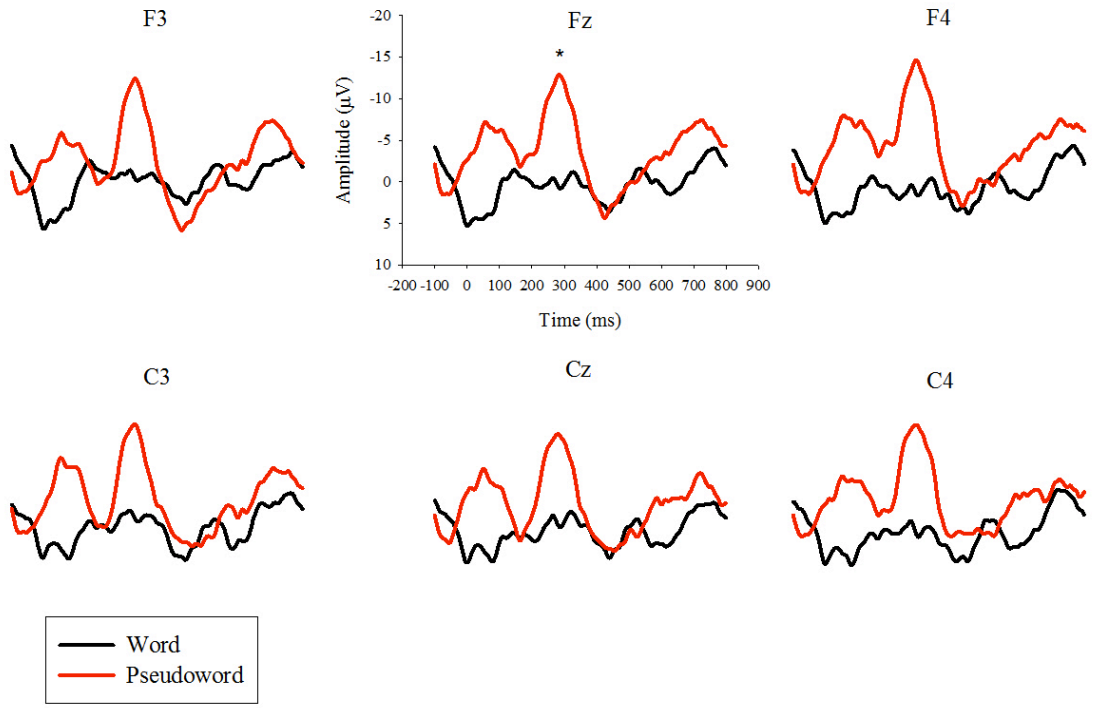

Figure 4. Grand average event-related potential waveforms elicited by RS to the auditory stimuli in the word (black line) and pseudoword (red line) conditions. Electrodes are displayed in relation to how they were positioned on RS's scalp with anterior sites plotted at the top of the figure. Asterisk indicates the occurrence of MMN-like response in the pseudoword condition.

between 200 and 350 ms at all frontal and central sites. This response is later than that of the comparison group, but still falls somewhat within the range of an MMN response, which is typically seen between 100 and 250 ms (Näätänen, 1992). Furthermore, the MMN-like component was notably greater in amplitude than the MMN responses seen in typically developing children, as waveforms were based on one individual compared to multiple participants in a grand average.

Discussion – Experiment 2

The data for the typically developing children in Experiment 2 were not consistent with the hypothesis. It was expected that there would be a difference between the MMN responses for the two conditions such that MMN for the word deviant would be greater in amplitude than the MMN for the pseudoword deviant, but there was no significant difference detected. However, both conditions elicited clear MMN responses between 100 and 150 ms that were largest over fronto-central sites. The timing and distribution of the MMN in the current study is consistent with previous studies (Näätänen, 1992). Such responses were expected as MMN components can be detected in infants to phoneme contrasts (Cheour et al., 1998), which were present at the divergent points of the stimuli in the current study.

The lack of difference in amplitude between the word and pseudoword conditions conflicts with previous research. Pulvermüller et al. (2001) and Shtyrov and Pulvermüller (2002) found enhanced MMN responses to word deviants in comparison to pseudoword deviants in adults. This difference was not based on a lexical difference and was instead thought to reflect long-term memory traces for words (Shtyrov & Pulvermüller, 2002). The results are also not consistent with the work of Korpilahti et al. (2001) who found

that children around the same age as the participants in the current study showed enhanced lMMN responses to word deviants in comparison to pseudoword deviants.

It seems unlikely that the typically developing children in the current study lacked long-term memory traces for the words presented, as they were relatively common, monosyllable words that were considered to be high frequency at a grade one level (Zeno et al., 1995). The corpus used only had frequency ratings for children as young as grade one. Although many participants were not yet in grade one, it is probable that they were familiar with the words used (i.e., *bite, bike, night*) as they are common in everyday language. It seems more likely the brains of these participants had not matured enough to show evidence of long-term memory traces for words.

An alternative explanation as to why differences in MMN amplitude were not observed between the conditions is that some of the participants may have elicited a positivity instead of a negativity. Previous studies have demonstrated that some infants show a positive-going response (Kushnerenko et al., 2002; Morr et al., 2002; Rivera-Gaxiola et al., 2005) and such responses have also been detected in school-aged children (i.e., 6 and 7 year olds) as well (Maurer et al., 2003). It was difficult to ascertain such differences through visual inspection of the individual data in the current study. However, it appeared as though at least one participant showed more of a positivity, which may have affected the amplitude of the grand average MMN in one or both conditions.

Unlike the comparison group, RS did not show an MMN response at all to the word deviant over fronto-central sites, but did seem to show what might be either a delayed MMN response between 200 and 350 ms to the pseudoword deviant or possibly a distinct ERP response. Given that the response was prominent over fronto-central sites, it

seems likely that the response she elicited is, at the very least, MMN-like in nature. Having said that, the data from parietal channels contained too many artifacts to clearly determine if the difference waveforms at parietal sites resembled those at fronto-central sites.

It is interesting that RS did not elicit any differential response between the standard and deviant in the word condition. As previously noted, electrophysiological evidence has shown that infants are able to distinguish between phonemes (Cheour et al., 1998) and the stimuli within each condition in the current study differed by a phoneme at the divergence point. The data would seem to suggest that RS was not even distinguishing between basic speech sounds for this condition; however, given that she did show a MMN-like response in the pseudoword condition, it seems more likely that she was judging the lexicality of the stimuli. In other words, it is possible that she was assessing the lexicality of each stimulus as it was presented instead of simply responding to a deviant in a train of standards. As there was a lexical difference in the pseudoword condition, perhaps RS processed deviant stimuli as lacking a lexical representation in her brain while standards indeed had a lexical representation. RS would have heard these stimuli after she heard those in the word condition, where both standard and deviant stimuli were words for which she could have had lexical representations.

General Discussion

The current study aimed to provide an assessment of aspects of language in a young child living with a rare disease known as CDG. Specifically, it was of interest to see if the patient, RS, who is non-motor and non-verbal, is able to recognize her own name and demonstrate long-term memory for words. To do so, two electrophysiological

experiments were conducted: one with a name recognition task and the other with an MMN task. The ERP responses elicited by RS were compared to a group of typically developing children and it was determined that she was recognizing her name and distinguishing between words and pseudowords.

The lack of prior research using similar paradigms on school-aged children makes it difficult to interpret the results of the current study. For instance, there have not been any known name recognition studies on younger populations with the exception of the work by Parise et al. (2010) on infants. Additionally, the work by Korpilahti et al. (2001) on children had a slightly different structure and made use of disyllabic Finnish stimuli, which have unique properties in comparison to English words. The study upon which the current study was based was that of Shtyrov and Pulvermüller (2002), who used adult participants. The words used by Shtyrov and Pulvermüller were not high frequency at a grade one level; therefore, different stimuli were used.

It may have been preferable to test the stimuli from the current study on adult participants as well to ensure that P300 responses and differences in the MMN responses across conditions could be detected to the stimuli chosen in a fully matured brain. More insight regarding the lack of difference between the MMN responses in the word condition and the pseudoword condition could be provided if adults were tested as well. In the future, it may be beneficial to test children of various ages, adolescents, and adults using the tasks from the current study to see how, or even if, their responses change with age. Additionally, follow-ups with the participants in the current study could also determine if their responses have changed over time.

It may have seemed ideal to conduct an experiment similar to Byrne et al. (1995) using a computerized version of the PPVT-R. A previous study by Connolly et al. (1995) validated this method using adult participants and the PPVT-R contains levels appropriate for younger populations as well. Furthermore, the CP patient tested in the study by Byrne et al. was non-verbal and non-motor like RS. However, RS's vision is too poor to rely on visual experiments.

While RS's data from the first experiment were more conclusive than the second, caution must be taken when interpreting her data in general. Movements, blinking, and muscle spasms may have lowered the quality of the data even though artifacts larger than 100 μV were excluded. It is indeed very encouraging, however, that ERP responses could be obtained from RS to provide the first true assessment of her language abilities. Traditional behavioral methods require the use of verbal and/or motor responses, which RS is unable to provide. While the current study does not represent a comprehensive assessment of RS's language abilities, it provides key insight for her family and the medical professionals who work with her in terms of her apparent ability to recognize her own name and possibly to distinguish words from non-words. Such knowledge may be helpful in tailoring activities for RS in school or in any rehabilitative therapy she may undergo in the future.

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Appendix A

Consent Form – Typically Developing Children

Electrophysiological Assessment of Aspects of Language in a Child with Congenital Disorder of Glycosylation: A Case Study

Dr. Randy Lynn Newman (Student Supervisor), Acadia Psychology Department, (902) 585-1405
Rebecca Webster (Honours Student), Acadia Psychology Department, 099621w@acadiau.ca

Purpose of the Research

The purpose of this study is to determine if a participant with a rare disease called Congenital Disorder of Glycosylation (CDG) has receptive language abilities. The participant is developmentally delayed and is unable to communicate, but it may be possible to detect signs of language comprehension in this individual by recording brain waves using electrophysiological measures. It is of interest to see how the CDG participant will respond in comparison to healthy controls. Therefore, typically developing children are required to act as control participants in the current study. In order to take part in this study, children must be right-handed, be three to six years old, have normal or corrected-to-normal hearing, have no history of neurological illness (e.g., epilepsy) or learning disabilities, and be a native English speaker (i.e., English is the first language that was taught at home).

Description of the Research

If you agree to your child's participation, and your child gives his or her consent as well, we will ask him or her to complete two tasks using electroencephalography (EEG). Both tasks will be passive, meaning your child will not need to respond. In the first task, your child will be played a recording of names through headphones. One of the names will be your child's mixed with seven different names. Your child will simply be asked to sit still and listen to the names. In the second task, a recording will be played to your child through headphones again as he or she sits still and watches a show (or movie). There will be two different recordings with the first including two different words and the second including one word and one nonword. We record brain activity using EEG by placing a cap with embedded electrodes over your child's head. A small amount of gel is inserted into each electrode in order to pick up the electrical activity generated by the brain. The gel will mess up your child's hair, but he or she will be able to wash it afterwards just outside the laboratory. The entire EEG session, including setup, will last between 1 and 1 ½ hours.

Potential Harms

We are not aware of any serious physical or psychological risks associated with this research. However, some participants may experience boredom during the testing. Gel from the cap will also be left in your child's hair after completion of the experiment. We will provide you with an opportunity to wash your child's hair before you leave.

Participating in this study is strictly voluntary and you have the right to withdraw your child from the study at any time without explanation and without loss of your promised compensation (i.e., money or prizes).

Potential Benefits
This study will not likely benefit you or your child directly. However, his or her participation will assist greatly in determining if the participant with CDG has any receptive language abilities. Such findings may have strong implications for the family and for understanding more about the disability in general.

Confidentiality
Your child's participation in this study will not be revealed to anyone that is not involved in the research team. All information will be kept confidential and will be used for research purposes only. No one other than members of the research team will have access to the data. Data will be stored on drives that are kept in a locked cabinet located in the laboratory of Dr. Newman.

Publication
This research is being conducted with the intention of having it published in a scientific journal. Neither your name nor your child's name (or any other identifying markers) will be included in the published results. Although you may withdraw your child's data after participating in the study, it is not possible to withdraw data once results have been published or disseminated.

Reimbursement
Your child will have the opportunity to choose a small prize after participating in the study. Even if you decide to withdraw from the study, your child will still receive a prize. You will also receive monetary compensation in the amount of $10 per hour (or part thereof) for the time associated with bringing your child to participate in this research. If you decide to withdraw from this research, you will still receive the reimbursement for which you are eligible.

Participation
All participants have the right to refuse to participate in this study. Even if you agree for your child to participate now you will be able to withdraw from the study at any point without negative consequences. Any agreement to participate is not legally binding and by consenting, participants have not waived any rights to legal recourse in the event of research-related harm. Participants will be asked throughout the study if they would like to continue participating. It is important to note that withdrawal of participation does not also include the withdrawal of data collected up until the point of withdrawal; if you decide to withdraw after we have already completed some of the testing, we will keep your child's data unless you ask us to delete it. As previously noted, your child's data may be removed from the study after participation but it is not possible to do so once results have been published or disseminated.

Identity of Sponsor
This research is funded by the Natural Sciences and Engineering Research Council of Canada (NSERC).

Consent
The objective of this experiment is to determine if a participant with CDG has receptive language abilities using healthy, typically developing children as controls. It has been explained that there are no known physical or psychological risks associated with your child's participation in this research but that there are no real personal benefits to participating in this research either.

If you have any questions, now or in the future, please contact Rebecca Webster (099621w@acadiau.ca) or Dr. Randy Lynn Newman, Student Supervisor, (randy.newman@acadiau.ca). If you have any ethical concerns about this study, please contact the Acadia Research Ethics Board at smaitzen@acadiau.ca or 902-585-1498.

I have read and understood the research consent form and I agree to participate.

……..
Participant Name

……..	……
Participant's Parent/Guardian Name	Signature
……..	……
Researcher's Name	Signature

Appendix B

Consent Form – CDG Participant

Electrophysiological Assessment of Aspects of Language in a Child with Congenital Disorder of Glycosylation: A Case Study

Dr. Randy Lynn Newman (Student Supervisor), Acadia Psychology Department, (902) 585-1405
Rebecca Webster (Honours Student), Acadia Psychology Department, 099621w@acadiau.ca

Purpose of the Research

The purpose of this study is to determine if your child, who has Congenital Disorder of Glycosylation (CDG), has receptive language abilities. Although your child is developmentally delayed and unable to communicate, it may be possible to detect signs of language comprehension by recording brain waves using electrophysiological measures. It is of interest to see how your child will respond to the experimental tasks in comparison to healthy controls. These healthy controls are typically developing children who are right-handed, are three to six years old, have normal or corrected-to-normal hearing, have no history of neurological illness (e.g., epilepsy) or learning disabilities, and are native English speakers (i.e., English is the first language that was taught at home).

Description of the Research

If you agree to your child's participation, and your child gives his or her consent as well, we will ask him or her to complete two tasks using electroencephalography (EEG). Both tasks will be passive, meaning your child will not need to respond. In the first task, your child will be played a recording of names through headphones. One of the names will be your child's mixed with seven different names. Your child will simply be asked to sit still and listen to the names. In the second task, a recording will be played to your child through headphones again as he or she sits still and watches a show (or movie). There will be two different recordings with the first including two different words and the second including one word and one non-word. We record brain activity using EEG by placing a cap with embedded electrodes over your child's head. A small amount of gel is inserted into each electrode in order to pick up the electrical activity generated by the brain. The gel will mess up your child's hair, but you will be able to easily wash it for her afterwards. The entire EEG session, including setup, will last between 1 and 1 ½ hours.

Potential Harms

We are not aware of any serious physical or psychological risks associated with this research. However, some participants may experience boredom during the testing. Gel from the cap will also be left in your child's hair after completion of the experiment. It can be easily washed out with normal shampoo. Participating in this study is strictly voluntary and you have the right to withdraw your child from the study at any time without explanation and without loss of your promised compensation (i.e., money or prizes).

Potential Benefits
This study is not a clinical determination of your child's abilities and may not be conclusive due to artifacts from movement, blinking, muscle spasms, etc. However, it may help to provide some insight into whether or not your child is able to understand spoken language.

Confidentiality
Your child's participation in this study will not be revealed to anyone that is not involved in the research team. All information will be kept confidential and will be used for research purposes only. No one other than members of the research team will have access to the data. Data will be stored on drives that are kept in a locked cabinet located in the laboratory of Dr. Newman. If requested, findings of the study can be provided directly to your family. Only with your permission will results also be given to your child's doctors. However, pediatric neurologist Dr. Meek will be made aware of the results due to his involvement as a researcher in the current study.

Publication
This research is being conducted with the intention of having it published in a scientific journal. Neither your name nor your child's name (or any other identifying markers) will be included in the published results. Although you may withdraw your child's data after participating in the study, it is not possible to withdraw data once results have been published or disseminated.

Reimbursement
Your child will have the opportunity to choose a small prize after participating in the study. Even if you decide to withdraw from the study, your child will still receive a prize. You will also receive monetary compensation in the amount of $10 per hour (or part thereof) for the time associated with participation in this research. If you decide to withdraw from this research, you will still receive the reimbursement for which you are eligible.

Participation
All participants have the right to refuse to participate in this study. Even if you agree for your child to participate now you will be able to withdraw from the study at any point without negative consequences. Any agreement to participate is not legally binding and by consenting, participants have not waived any rights to legal recourse in the event of research-related harm. Participants will be asked throughout the study if they would like to continue participating. Although this may be difficult for your child, the researchers will look for gestures as responses. Testing will not proceed if any signs of discomfort (e.g., facial expressions, tense body language) are observed. It is important to note that withdrawal of participation does not also include the withdrawal of data collected up until the point of withdrawal; if you decide to withdraw after we have already completed some of the testing, we will keep your child's data unless you ask us to delete it. As previously noted, your child's data may be removed from the study after participation but it is not possible to do so once results have been published or disseminated.

Identity of Sponsor
This research is funded by the Natural Sciences and Engineering Research Council of Canada (NSERC).

Consent
Although results may not be conclusive, the objective of this experiment is to determine if your child has receptive language abilities. Healthy, typically developing children will be used as controls and their data will be compared to your child's. It has been explained that there are no known physical or psychological risks associated with your child's participation in this research.

If you have any questions, now or in the future, please contact Rebecca Webster (099621w@acadiau.ca) or Dr. Randy Lynn Newman, Student Supervisor, (randy.newman@acadiau.ca). If you have any ethical concerns about this study, please contact the Acadia Research Ethics Board at smaitzen@acadiau.ca or 902-585-1498.

I have read and understood the research consent form and I agree to participate.

……..
Participant Name

……..	……
Participant's Parent/Guardian Name	Signature
……..	……
Researcher's Name	Signature

Appendix C

Feedback and Debriefing Form – Typically Developing Children

Electrophysiological Assessment of Aspects of Language in a Child with Congenital Disorder of Glycosylation: A Case Study

The purpose of this research is to determine whether or not a child born with a rare disease called Congenital Disorder of Glycosylation (CDG) is able to understand spoken language. This will be made possible by analyzing the event-related brain potentials (ERPs) that were measured during the experimental tasks. ERPs are signals obtained by recording the brain's electrical activity with electroencephalography (EEG). Specific ERP components reflect the brain's response to the presentation of an external stimulus and are used in the study of cognitive processes such as language, attention, and memory.

The first task was a name recognition task in which participants heard their own name along with some other names. Previous research has shown that when a participant's own name is presented at a relatively low probability, an ERP component called the P300 will be elicited. Because a participant's own name is particularly relevant to him or her, P300 responses are generally larger when participants hear their own name compared to when they hear another name (Berlad & Pratt, 1995; Folmer & Yingling, 1997). Therefore, it is expected that typically developing participants will show larger P300 responses for their own names in comparison to the other names presented. It is of interest to see if the CDG participant will show P300 responses similar to typically developing children.

In the second task, participants heard many repetitions of one stimulus (i.e., the standard) and fewer repetitions of another stimulus (i.e., the deviant). There were two conditions and your child experienced both. Each condition had an English word as the standard, but while the deviant in the first condition was another English word, the deviant in the second condition was a pseudoword (i.e., a combination of sounds that could be perceived as a word but do not actually exist together as a word in the English language). An ERP component called the Mismatch Negativity (MMN) is seen in tasks such as this and is generally largest for deviant stimuli. The MMN reflects the brain's automatic response to any slight change in auditory input even without attention. However, MMN responses can provide more information about the brain other than its basic recognition of acoustic differences. Some studies have found that MMN responses can be significantly stronger when the deviant is a word compared to a pseudoword, which is thought to reflect long-term memory for words (Korpilahti, Krause, Holopainen, & Lang, 2001; Pulvermüller, Shtyrov, Kujala, & Näätänen, 2004; Shtyrov & Pulvermüller, 2002). Therefore, we are expecting that typically developing participants will have MMN responses that are larger for word deviants than pseudoword deviants. But most importantly, it is of interest to see if the CDG participant will show responses similar to the typically developing participants in this portion of the study. A larger MMN to word deviants would suggest that that the child with CDG has some ability to understand language.

Thank you for your participation in this study! If you have any questions or concerns, now or in the future, please do not hesitate to contact Dr. Randy Newman (randy.newman@acadiau.ca). We also welcome you to contact us if you would like a copy of the results of the study. If you have any ethical concerns about this study, please contact the Acadia Research Ethics Board at smaitzen@acadiau.ca or 902-585-1498.

Appendix D

Feedback and Debriefing Form – CDG Participant

Electrophysiological Assessment of Aspects of Language in a Child with Congenital Disorder of Glycosylation: A Case Study

The purpose of this research is to determine whether or not your child, who was born with Congenital Disorder of Glycosylation (CDG), is able to understand spoken language to some degree. This will be made possible by analyzing the event-related brain potentials (ERPs) that were measured during the experimental tasks. ERPs are signals obtained by recording the brain's electrical activity with electroencephalography (EEG). Specific ERP components reflect the brain's response to the presentation of an external stimulus and are used in the study of cognitive processes such as language, attention, and memory.

The first task was a name recognition task in which participants heard their own name along with some other names. Previous research has shown that when a participant's own name is presented at a relatively low probability, an ERP component called the P300 will be elicited. Because a participant's own name is particularly relevant to him or her, P300 responses are generally larger when participants hear their own name compared to when they hear another name (Berlad & Pratt, 1995; Folmer & Yingling, 1997). Therefore, it is expected that typically developing participants will show larger P300 responses for their own names in comparison to the other names presented. It is of interest to see if your child will show P300 responses similar to typically developing children. If so, it would indicate that she does know her own name, or at the very least, this name is relevant to her in some way.

In the second task, participants heard many repetitions of one stimulus (i.e., the standard) and fewer repetitions of another stimulus (i.e., the deviant). There were two conditions and your child experienced both. Each condition had an English word as the standard, but while the deviant in the first condition was another English word, the deviant in the second condition was a pseudoword (i.e., a combination of sounds that could be perceived as a word but do not actually exist together as a word in the English language). An ERP component called the Mismatch Negativity (MMN) is seen in tasks such as this and is generally largest for deviant stimuli. The MMN reflects the brain's automatic response to any slight change in auditory input even without attention. However, MMN responses can provide more information about the brain other than its basic recognition of acoustic differences. Some studies have found that MMN responses can be significantly stronger when the deviant is a word compared to a pseudoword, which is thought to reflect long-term memory for words (Korpilahti, Krause, Holopainen, & Lang, 2001; Pulvermüller, Shtyrov, Kujala, & Näätänen, 2004; Shtyrov & Pulvermüller, 2002). Therefore, we are expecting that control participants will have MMN responses that are significantly larger for word deviants than pseudoword deviants. But most importantly, it is of interest to see if your child will show responses similar to the control participants in

this portion of the study as well. A larger MMN to word deviants would suggest that your child has long-term representations for words in the brain.

If your child does not show results that are similar to typically developing children, it does not necessarily mean that she does not have language comprehension abilities. Artifacts from movements, blinking, muscle spasms, etc. interfere with our ability to obtain clear data that are meaningful and can lead to inconclusive results. Although the study may indeed provide some insight into whether or not your child is able to understand spoken language, we are not completing a comprehensive assessment.

Thank you for your participation in this study! If you have any questions or concerns, now or in the future, please do not hesitate to contact Dr. Randy Newman (randy.newman@acadiau.ca). As previously noted, findings of the study can be provided directly to your family if requested. However, only with your permission will results also be given to any of your child's doctors other than Dr. Meek, a researcher in the current study. If you have any ethical concerns about this study, please contact the Acadia Research Ethics Board at smaitzen@acadiau.ca or 902-585-1498.