IMPLEMENTING A NATURALISTIC RECAST APPROACH IN ADULTS WITH DOWN SYNDROME DISINTEGRATIVE DISORDER: A TUTORIAL

A Thesis
by
COURTNEY HALE

Submitted to the School of Graduate Studies
at Appalachian State University
in partial fulfillment of the requirements for the degree of
Master of Science

December 2022
Department of Rehabilitation Sciences

IMPLEMENTING A NATURALISTIC RECAST APPROACH IN ADULTS WITH DOWN SYNDROME DISINTEGRATIVE DISORDER: A TUTORIAL

A Thesis
by
COURTNEY HALE
December 2022

APPROVED BY:

_____
Holly Hanley, Ph.D.
Chairperson, Thesis Committee

_____
Kimberly McCullough, Ph.D.
Member, Thesis Committee

_____
Bliss Hemric
Member, Thesis Committee

_____
Gail S. Donaldson, Ph.D.
Chairperson, Department of Rehabilitation Sciences

_____
Marie Hoepfl, Ed.D.
Interim Dean, Cratis D. Williams School of Graduate Studies

Abstract

IMPLEMENTING A NATURALISTIC RECAST APPROACH IN ADULTS WITH DOWN SYNDROME DISINTEGRATIVE DISORDER: A TUTORIAL

Courtney Hale
B.S., University of Rochester
M.S., Appalachian State University

Chairperson: Holly Hanley, Ph.D.

Researchers have reported unexplained regression in young adults with Down Syndrome (DS). This regression has been termed Down Syndrome disintegrative disorder (DSDD) (Worley et al., 2015), catatonia not otherwise specified (Ghaziuddin et al., 2015; Lyons et al., 2020; Miles et al., 2019), early-onset parkinsonism (Palat et al., 2018), new-onset autistic regression, dementia, and insomnia (Worley et al., 2015). The demographic profile of Down Syndrome disintegrative disorder (DSDD) includes a post-pubertal onset, a female to male ratio of 2:1, and a mean age of regression of 17.5 years (Rosso et al., 2019). These individuals exhibit seven times as many depressive symptoms and experience six times as many life stressors, such as a change in school or death of a family member (Santoro et al., 2020).

Symptoms typically associated with DSDD include impaired cognition, or cognitive decline to a dementia-like state, new-onset insomnia, new-onset autistic characteristics, and a decline in baseline skills including activities of daily living (Ghaziuddin et al., 2015; Worley et al., 2015). Research indicates that DSDD is accompanied by a significant decline in speech or language, as 94% of individuals with DSDD experience a speech impairment and mutism is observed in 57% of individuals with DSDD (Mircher et al., 2017). DSDD results in a decline in frequency of speech; however, many individuals with DSDD also experience a decrease in oral motor control, resulting in a significant decrease in speech intelligibility (Jacobs et al., 2016). Researchers have

identified possible medical treatment options for DSDD, including electroconvulsive therapy, immunotherapy treatment, and various pharmaceutical treatments, (Cardinale et al., 2019; Ghaziuddin et al., 2015; Miles et al., 2019; Rosso et al., 2019). Some of these treatment methods have resulted in a temporary remission of symptoms and/or an improvement in activities of daily living, however individuals being treated have not yet shown improvement in speech impairments, mutism, or speech intelligibility (Mircher et al., 2017; Rosso et al., 2019).

Although research has not yet focused on treating the decline in speech and language in individuals with DSDD, the effectiveness of treatments targeting speech intelligibility in individuals with DS have been evaluated. One therapeutic approach that has been used to target speech intelligibility in individuals with DS is naturalistic recasts, also termed Broad Target Recasts, Broad Target Speech Recasts, and conversational recasts (Yoder et al., 2016). The use of naturalistic recasts in individuals with DS has resulted in improvements in both speech intelligibility and language (Yoder et al., 2005; Yoder et al., 2016). This paper highlights the use of a naturalistic recast approach in individuals with DSDD as an appropriate therapeutic intervention strategy to improve their speech intelligibility as well as language, and therefore could result in significant improvements in the quality of life of said individuals.

Acknowledgements

I would like to express my deepest appreciation to my professor and committee chair, Dr. Holly Hanley. Thank you for helping me create this project from a single experience and for providing me with guidance and motivation at any and all times. I would also like to thank my committee members, Dr. Kimberly McCullough and Bliss Hemric, for their encouragement, enthusiasm, and feedback. Lastly, I would like to express gratitude to my amazing friends and family. I appreciate your never-ending supply of positivity and support.

Dedication

This thesis is dedicated to my sister Abbey, for inspiring this project. Thank you for being unapologetically yourself and for providing an unending number of smiles and knock-knock jokes.

Table of Contents

Down Syndrome Disintegrative Disorder

In recent years, researchers have reported unexplained regression in young adults with Down Syndrome (DS). This regression has been termed Down Syndrome disintegrative disorder (Worley et al., 2015), catatonia not otherwise specified (Ghaziuddin et al., 2015; Lyons et al., 2020; Miles et al., 2019), early-onset parkinsonism (Palat et al., 2018), new-onset autistic regression, dementia, and insomnia (Worley et al., 2015). Worley et al. (2015) assert that Down Syndrome disintegrative disorder is a disorder featuring autistic regression, cognitive decline resulting in a dementia-like state, occurrence at an older age than usual for autistic regression, and no other diagnosis established to explain the condition.

Affected Population

Rosso and colleagues (2019) reviewed and summarized current data on Down Syndrome disintegrative disorder (DSDD) and asserted that DSDD is a clinical syndrome that should be considered in adolescents and young adults with DS and subacute-onset behavioral changes. Additionally, Rosso et al. (2019) found that the demographic profile of this disorder includes a post-pubertal onset, as well as an elevated female-to-male ratio of 2:1. DSDD seems to take place when patients are older, compared to the typical age of onset of autistic regression in individuals with DS (Rosso et al., 2019). In reviewing 35 cases of individuals with regression in DS, Santoro et al. (2020) found a mean age of regression of 17.5 years. Researchers Mircher et al. (2017) reported on 30 cases of regression in individuals with DS, and found the mean age of regression to be 18 in females and 21 in males.

Potential Risk Factors

Some researchers have identified potential causes or triggers of regression in individuals with DS. Santoro et al. (2020), after reviewing 35 cases of individuals with "unexplained regression in DS," found that the individuals with regression had four times as many mental

health concerns compared to age- and sex-matched controls, “driven by higher prevalence of issues related to mood, sleep, appetite, incontinence, and transition.” Individuals with regression were also found to have six times as many life stressors compared to controls, such as a change in school/employment, loss of a friend/family member due to a move, and/or death of a family member. Additionally, these individuals with “unexplained regression in DS” were found to have seven times as many depressive symptoms, such as social withdrawal, poor concentration, loss of interest, and/or poor memory, compared to age- and sex-matched controls with DS (Santoro et al., 2020).

In describing seven children with DS who presented with developmental regression, Lyons and colleagues (2020) identified that for some of these individuals, symptom onset coincided with a potential psychosocial trigger. These possible triggers included an illness, family bereavement, a sibling moving away, and moving to a new school (Lyons et al., 2020). Similarly, after reporting on 30 cases of regression in patients with DS, Mircher et al. (2017) found that a stressful life event may have triggered regression in the majority of patients, such as a change of school, awareness of disability, separation from parents, wedding of a sibling, assault, illness or death of a relative, and/or overstimulation. Furthermore, in three of the four patients with DSDD reported on by Cardinale et al. (2019), regression occurred after significant life events or traumas, including influenza immunizations, being physically assaulted, having a flood in the home, and a change in job responsibilities. Additionally, researchers Jacobs et al. (2016) reported on one individual with DS and clinical deterioration, which began after the individual graduated from high school. Researchers Palat et al. (2018) also reported on one individual with DS and behavioral regression, and found that the individual’s symptoms became more noticeable when she was placed in a more demanding environment. Furthermore, in

reporting on seven individuals diagnosed with DS and acute clinical regression, Takeuchi et al. (2017) found that these individuals developed intellectual regression after a significant environmental change, such as family unhappiness or troubling interpersonal relationships.

Associated Symptoms

Research has indicated that individuals with DSDD experience a wide range of emotional, physical, and cognitive symptoms. One study found that four individuals with DSDD demonstrated catatonic regression, impaired cognition, increased stereotypies, unusual movements, and a decline in activities of daily living. It was also reported that these four individuals all experienced a decrease in speech or language, or changes in speech and language such as verbigeration (Ghaziuddin et al., 2015). Another study found that of 11 children and adolescents, with a mean age of 14.8 years and DSDD or new-onset autistic characteristics, 100% of participants experienced autistic deterioration, 91% experienced cognitive decline to a dementia-like state, and 82% experienced new-onset insomnia (Worley et al., 2015). One individual with DS was reported by Jacobs et al. (2016) to have had a loss of activities of daily living and loss of baseline skills, perceived decline in cognition, unexplained decompensation in behavioral functioning and mood, significant loss of speech and written expression, staring episodes, and probable psychotic symptoms.

Mircher and colleagues (2017) reported on 30 cases of regression in young patients with DS. Within these 30 individuals, the most frequent psychiatric symptoms included mood disorders, such as apathy, slowness, or catatonia, as well as stereotypies, such as repetitive or ritualistic movements, postures, or utterances. Additionally, auto- or hetero-aggressive behavior was reported in 40% of these individuals. 87% of these patients presented with a total loss of independence, 94% experienced a speech impairment, and mutism was observed in 57% of the

patients, with the first sign often being a feeble voice or barely audible whisper (Mircher et al., 2017). An additional individual with DS, reported by Palat et al. (2018), presented with a one-year history of slowing in her general movements as well as behavioral regression. This individual previously spoke in full sentences; however, her speech was soft and limited to one to two words at a time after her regression. She also experienced tremors, which impacted her ability to complete activities of daily living, occasional bowel or bladder accidents, and symptoms of depression. Palat et al. (2018) reported that this individual was still able to follow conversations and directions, but she did so more slowly after her regression. It is evident that DSDD results in a decline in frequency of speech, but many individuals with DSDD also experience a decrease in oral motor control, resulting in a significant decrease in speech intelligibility (Jacobs et al., 2016).

In researching possible treatment for DSDD, Cardinale et al. (2019) summarized the symptoms of four patients with DS and this regression. One patient was reported to have developed acute onset insomnia, followed by autistic characteristics, catatonic symptoms, cognitive decline, and hallucinations. Following regression, this individual was unable to read and perform activities of daily living. This same patient experienced a second episode of regression, in which she developed sudden-onset insomnia, cognitive decline, language decline, and catatonia. The patient also exhibited severe verbigeration, mild echolalia, and mild mutism. The second patient reported on by Cardinale et al. (2019) developed acute onset of severe psychosis, followed by insomnia, catatonia, and autistic features. This individual exhibited severe mutism and mild verbigeration. The third patient, according to Cardinale and colleagues (2019), experienced a subacute decline, beginning with disorganization and anxiety. She then exhibited a decline in cognition and language, speaking only in short phrases and eventually

being unable to follow one-step commands. This patient also developed impulsivity, emotional lability, aggression, stereotypies, and auditory hallucinations. The final patient reported on by Cardinale et al. (2019) developed acute psychosis characterized by hallucinations and catatonia. He experienced severe insomnia, increased autistic features, incontinence, and headaches, as well as mild echolalia, verbigeration, and mutism.

Researchers Miles et al. (2019) reported on seven young patients with DS, all of whom were diagnosed with catatonia. Among these seven individuals, the most common symptoms of regression consisted of mutism, immobility, getting "stuck" during motor activities, involuntary movements, perseveration or obsessive symptoms, negativism, and social withdrawal. Loss of independence in daily living skills, sleep disturbances, and emotional lability were also common symptoms among these patients (Miles et al., 2019). Lyons et al. (2020) described the developmental regression of seven additional children with DS. Each individual's speech was impacted by their regression; they experienced a delay in speech initiation, mutism or a loss of speech, and/or speech characterized by monosyllabic responses or whispering. The majority of these seven individuals also exhibited different types of posturing, as well as grimacing and teeth grinding. Additionally, some individuals developed hand stereotypies, arm waving, flicking objects, and interlocking fingers (Lyons et al., 2020).

Rosso et al. (2019) reviewed and summarized current data regarding the clinical phenotypes of DSDD. The defining feature of DSDD was identified as a regression of previously attained skills, most notably in the domains of language, communication, and social skills. Through summarizing data from previous studies, Rosso et al. (2019) revealed that up to 87% of patients with DSDD were diagnosed with speech and/or language regression, with symptoms ranging in severity from dysfluency to mutism. Among individuals in whom severity of speech

regression was quantified, 38% of individuals experienced partial speech regression and 52% experienced mutism. Additionally, mood symptoms were commonly reported in individuals with DSDD, including depression, social withdrawal, anxiety, catatonia, new-onset insomnia, and occasionally psychotic symptoms (Rosso et al., 2019).

DSDD Treatment Options

Some researchers have identified possible treatment options for DSDD, such as electroconvulsive therapy, immunotherapy treatment, and treatment using benzodiazepines, antipsychotic drugs, as well as drugs typically used to treat parkinsonism and/or dementia. Ghaziuddin et al. (2015) reported on and treated four patients with DS and catatonic regression. All four individuals were first treated with lorazepam, with mixed results of improved symptoms, partial improvement, and no response. Each individual was then treated using electroconvulsive therapy with bilateral electrode placement, resulting in remission of symptoms in three cases and improving symptoms in one case. This research does not specify whether improvements were made across some or all identified domains, including motor, speech, behavior, mood, and daily living skills (Ghaziuddin et al., 2015). Similarly, Miles and colleagues (2019) treated seven young patients with DS who were diagnosed with catatonia. After being treated with lorazepam and electroconvulsive therapy, the majority of these patients responded to treatment by walking faster, looking around at their environment more, having more relaxed posture, and showing less negativity. Following treatment, individuals continued to have symptoms of facial movements, shoulder shrugging, staring, and resisting previously enjoyed activities (Miles et al., 2019).

Researchers Lyons et al. (2020) treated seven children with DS who presented with developmental regression. These patients were treated with lorazepam, and responses to this

treatment varied, as some responses were excellent, some were partial, and some were limited responses (Lyons et al., 2020). One individual, reported on by Jacobs et al. (2016), was treated with clozapine, resulting in a return to 85% of his baseline level of functioning. Although this clozapine treatment allowed him to return to many community-based activities, it also resulted in significant weight gain, sedation, and articulation difficulties caused by poor motor control (Jacobs et al., 2016). Additionally, researchers Takeuchi et al. (2017) treated seven individuals with DS and clinical regression using donepezil, a drug typically used for individuals with dementia. Three of these seven patients exhibited clinical improvement after this treatment (Takeuchi et al., 2017). Furthermore, one individual with DS and behavioral regression, reported on by Palat et al. (2018), was treated using levodopa, a drug typically used to treat parkinsonism. This treatment resulted in a return to baseline motor function, however speech-related motor function and/or speech intelligibility are not addressed (Palat et al., 2018).

Researchers Mircher et al. (2017) reported on 30 cases of regression in young patients with DS, in which the majority of patients were treated with benzodiazepines, selective serotonin reuptake inhibitors, antipsychotic drugs, and/or electroconvulsive therapy. Of these 30 individuals, 10% experienced worsening symptoms, 37% experienced stabilization of symptoms without recovering, 43% experienced partial recovery, and 10% experienced total recovery. Furthermore, 60% of these individuals were able to regain their autonomy and improving their daily living skills, however these individuals never spoke again (Mircher et al., 2017).

Cardinale and colleagues (2019) studied possible immunotherapy treatment for four patients with DSDD. Within weeks following immunotherapy treatment, all four patients had improvement in overall function; improvements were noted in symptom domains of cognition, autism, catatonia, insomnia, psychosis, and neurologic conditions (Cardinale et al., 2019).

Researchers Worley et al. (2015) reported on 11 individuals with DS and regression, in which one individual returned to his pre-deterioration condition of autism and two individuals were treated with steroids with no improvement. The majority of individuals experienced gradual improvement, not due to any intervention, however these individuals were left with autism and intellectual disability worse than before their regression (Worley et al., 2015).

Rosso et al. (2019) state that antipsychotics, selective serotonin reuptake inhibitors, and anticholinergic drugs have been used to address neuropsychiatric disturbances observed in DSDD. Benzodiazepines and electroconvulsive therapy have also been used for patients with DSDD and catatonia, resulting in at least a partial response. Ultimately, Rosso et al. (2019) concluded that future, prospective studies are needed in order to identify effective therapies for each symptom of DSDD.

Through reviewing the literature on DSDD or regression in DS, it is evident that this disorder has a significant impact on the overall frequency of speech and could negatively impact speech intelligibility due to motor deficits resulting from DSDD. It is also evident that the medical treatment options for DSDD have varied outcomes and limitations. There does not appear to be previous or current research on the impact of behavioral speech therapy on the speech and language symptoms associated with DSDD, nor does there appear to be research focused on improving speech intelligibility of these individuals with DSDD.

Speech Intelligibility Treatment

Although research has not yet focused on treating the decline in speech and language in individuals with DSDD, research has been used to evaluate the effectiveness of certain treatment approaches on improving speech and language in individuals with DS. Furthermore, treatment

approaches that prove to be effective in improving speech intelligibility in individuals with DS may have similar effects on individuals with DSDD.

The majority of interventions for people with DS aim at improving language skills rather than speech sound production or speech intelligibility (Coppens-Hoffman et al., 2012). Despite this, some treatment methods that have been used to target speech sound production or speech intelligibility in individuals with DS, including electropalatographic (EPG) therapy and the Lee Silverman Voice Treatment (LSVT). Research indicates that EPG therapy, used to target speech sound productions in children with DS aged 8 to 18, results in improved speech sound production with no transfer to overall speech intelligibility (Page & Johnson, 2021). Researchers Mahler and Jones (2012) evaluated the effectiveness of LSVT on targeting speech intelligibility, loudness, and phonatory stability in two individuals, aged 33 and 34, with DS and dysarthria. In this case, LSVT resulted in an increase in speech intelligibility in one participant (Mahler & Jones, 2012).

One therapeutic approach that has been shown to increase speech intelligibility in individuals with and without DS is naturalistic recasts, also termed Broad Target Recasts, Broad Target Speech Recasts, and conversational recasts (Yoder et al., 2016). Using naturalistic recasts, clinicians repeat or recast a client's attempt to use target morphemes, correcting for articulation or grammatical errors when they occur (Hall & Plante, 2020).

Researchers Yoder et al. (2005) completed a randomized group experiment to determine the efficacy of Broad Target Recasts (BTR) on mean-length utterance and speech intelligibility. In this study, 52 preschoolers with specific speech and language impairments were randomly assigned to a control group or a BTR treatment group. Individuals in the BTR treatment group received three 30-minute treatment sessions per week for six months, whereas individuals in the

control group were not provided BTR. BTR was not effective immediately following treatment, however a treatment effect was seen on follow-up speech intelligibility. Additionally, BTR had the largest impact on speech intelligibility of children with poor pretreatment speech accuracy (Yoder et al., 2005). In a later study, Yoder and colleagues (2016) compared treatment effects in students with DS who were treated using Broad Target Speech Recasts and Easy Does It, an approach used for individuals with apraxia. This study revealed that individuals with Down Syndrome who were treated using Broad Target Speech Recasts experienced superior gains in their speech comprehensibility (Yoder et al., 2016).

A similar treatment approach to naturalistic recasts was also used by Hewitt and colleagues (2005), in which researchers used a hybrid naturalistic recasts treatment approach to target syntax in adults with DS. This hybrid treatment approach featured components of naturalistic recasts as well as drill-based therapy. By the end of this study, all three adults with DS showed an increase in the use of target syntactical structures in obligatory contexts (Hewitt et al., 2005).

Although a naturalistic recast treatment approach has been used most often in young children, research also suggests that this approach may be effective in improving speech intelligibility and comprehensibility, syntactic language development, and morphological development in adolescents and adults. Naturalistic recasts have also been implemented with clients with a variety of disorders, including Specific Language Impairment and Down Syndrome (Hewitt et al., 2005; Yoder et al., 2005, 2016).

Proposed Implementation of a Naturalistic Recast Approach

In implementing a naturalistic recast approach in adults with DSDD, Yoder and colleagues note that that a successful recast cannot be delivered unless the client talks or uses approximate verbalizations with the clinician, and unless the client's message is understood or can be inferred (Yoder et al., 2016). Yoder et al. (2016) utilized Broad Target Speech Recasts to treat speech comprehensibility in children with DS by incorporating verbal routines and topic-continuing questions. This increased the likelihood that the clinician was able to predict and understand client utterances, and therefore allowed the clinician to successfully implement a recast approach (Yoder et al., 2016). Similarly, Hewitt and colleagues (2005) utilized a hybrid naturalistic recast approach to target expressive grammar in adults with DS by incorporating both naturalistic and structured intervention procedures. In order to ensure that clinicians could anticipate and successfully recast client utterances, these researchers provided two types of conversational prompts, including asking clients about experiences, likes, and dislikes, as well as asking clients to comment on photographs (Hewitt et al., 2005).

Although there have been no studies using naturalistic recast with DSDD, similar procedures could be implemented when utilizing this treatment approach in adults with DSDD. For example, treatment procedures should allow clinicians to have a high likelihood of understanding client utterances and should be age-appropriate. In order to successfully implement a naturalistic recast treatment approach, clinicians should consider utilizing a variety of relevant, age-appropriate conversational prompts, including asking about personal experiences, likes, dislikes, and prompting clients to describe or comment on relevant photographs. Clinicians may also consider having clients re-enact scenes from books, shows, or movies of interest. These prompts will likely help to motivate the client and increase

generalizability of treatment effects, and they will also ensure that the clinician implementing a recast approach is able to understand and repeat client utterances, despite a possibility of decreased speech intelligibility (Hewitt et al., 2005; Yoder et al., 2016). Ultimately, clinical research must be done to determine if the use of naturalistic recasts can help to increase speech intelligibility in adults with DSDD.

When evaluating and treating individuals with DSDD, clinicians should take into consideration that DSDD is a disorder featuring speech-language regression, and the disorder also lacks research on clinical, non-pharmaceutical treatment options. Further research also must be done to determine if a naturalistic treatment method, such as naturalistic recasts, can be effective to target various speech and language goals for individuals with progressive disorders such as DSDD.

Discussion

Although research has not yet focused on treating the decline in speech and language in individuals with DSDD, research has been done to treat behavioral and/or psychological symptoms associated with DSDD. For instance, electroconvulsive therapy has been used to target symptoms associated with DSDD and has, in some cases, resulted in improved or remission of symptoms (Ghaziuddin et al., 2015). Although electroconvulsive therapy has the potential to be an effective treatment, it requires sedation and repeated sessions, and has the potential to result in memory impairment for the treated individual (Rosso et al., 2019). Various immunotherapy treatments and pharmaceutical drugs have also been used to target symptoms associated with DSDD, in which the majority of patients may respond with partial recovery. Some patients, however, may respond with worsening symptoms, significant weight gain, and additional articulation difficulties due to poor motor control (Jacobs et al., 2016; Mircher et al.,

2017). Furthermore, the vast majority of research conducted to treat individuals with DSDD does not address effects on speech intelligibility or speech-related motor skills.

Treatment methods that have been used to target speech sound production or speech intelligibility in individuals with DS include electropalatographic (EPG) therapy, the Lee Silverman Voice Treatment (LSVT), and naturalistic recasts. Research indicates that EPG therapy may improve speech sound production without any transfer to speech intelligibility (Page & Johnson, 2021). Additionally, EPG therapy may be difficult to implement in individuals with DSDD, as their history of regression may make them unable to consistently follow directions and speak frequently. In one study evaluating the effectiveness of LSVT on targeting speech intelligibility in individuals with DS and dysarthria, speech intelligibility increased in one of two patients (Mahler & Jones, 2012). Additionally, due to the highly structured nature of LSVT, this treatment protocol would likely be difficult to implement in individuals with DSDD due to the possibility of further regression.

Regarding naturalistic recasts, research indicates that this treatment approach is effective for improving speech intelligibility and comprehensibility, syntactic language development, and morphological development in adolescents and adults with and without DS (Hewitt et al., 2005; Yoder et al., 2005, 2016). Ultimately, a naturalistic recast approach could be easily implemented in individuals with speech regression, low intelligibility, and/or low speech frequency, such as those with DSDD. Implementing a naturalistic recast approach in individuals with DSDD could simultaneously improve their speech intelligibility and language skills, and therefore could result in significant improvements in the quality of life of said individuals.

Conclusion

Down Syndrome disintegrative disorder (DSDD) is a regressive disorder in individuals with Down Syndrome, characterized by impaired cognition, cognitive decline to a dementia-like state, new-onset insomnia, new-onset autistic characteristics, and a decline in baseline skills including activities of daily living (Ghaziuddin et al., 2015; Worley et al., 2015). Research indicates that DSDD is often accompanied by a significant decline in speech frequency as well as speech intelligibility (Jacobs et al., 2016; Mircher et al., 2017). A variety of medical treatment options have been used to treat symptoms associated with DSDD; many of these treatment methods bring with them significant risks, such as sedation, memory impairment, worsening symptoms, and further decline of oral motor skills (Jacobs et al., 2016; Mircher et al., 2017; Rosso et al., 2019).

There does not appear to be previous or current research on the impact of behavioral speech therapy on the decline in speech intelligibility and frequency in individuals with DSDD. Research has shown, however, that one approach which has been effective for improving speech intelligibility in individuals with and without DS is a naturalistic recast approach (Yoder et al., 2005, 2016). Implementing a naturalistic recast approach in individuals with DSDD, in hopes of improving speech intelligibility and expressive language skills, could significantly improve the quality of life of these individuals and their families.

When evaluating and treating adolescents and young adults with DS, clinicians should take regression due to DSDD into consideration. Further research should be done to continue to evaluate the potential risk factors of DSDD, in hopes that families and caregivers of individuals with DS can be made aware of said factors as well as possible preventative measures to take. Research must also be done to determine the impact of this disorder on the quality of life of

patients and their families. Additionally, detailed evidence must be gathered regarding the specific decline in motor-speech skills, speech frequency, and speech intelligibility, as the timeline of such regressions may have clinical and therapeutic significance.

References

Cardinale, K. M., Bocharnikov, A., Hart, S. J., Baker, J. A., Eckstein, C., Jasien, J. M., Gallentine, W., Worley, G., Kishnani, P. S., & Van Mater, H. (2019). Immunotherapy in selected patients with down syndrome disintegrative disorder. *Developmental Medicine & Child Neurology, 61*(1), 847-851. https://doi.org/10.1111/dmcn.14127.

Coppens-Hoffman, M. C., Maassen, B. A. M., van Schrojenstein Lantman-de Valk, H. M. J., & Snik, A. F. M. (2012). Speech difficulties and poor speech intelligibility in adults with down syndrome: A review of the literature. *Journal of Hearing Science, 2*(1), 9-16. https://doi.org/10.17430/882708.

Ghaziuddin, N., Nassiri, A., & Miles, J. H. (2015). Catatonia in down syndrome; A treatable cause of regression. *Neuropsychiatric Disease and Treatment, 11*(1), 941-949. https://doi.org/10.2147/NDT.S77307.

Hall, J., & Plante, E. (2020). Data-informed guideposts for decision making in enhanced conversational recast treatment. *American Journal of Speech-Language Pathology, 29*(1), 2068-2081. https://doi.org/10.1044/2020_AJSLP-20-00017.

Hewitt, L. E., Hinkle, A. S., & Miccio, A. W. (2005). Intervention to improve expressive grammar for adults with down syndrome. *Communication Disorders Quarterly, 26*(3), 144-155. https://doi.org/10.1177%2F15257401050260030301

Jacobs, J., Schwartz, A., McDougle, C. J., & Skotko, B. G. (2016). Rapid clinical deterioration in an individual with Down Syndrome. *American Journal of Medical Genetics, 170*(A), 1899-1902. https://doi.org/10.1002/ajmg.a.37674

Lyons, A., Allen, N. M., Flanagan, O., & Cahalan, D. (2020). Catatonia as a feature of down syndrome: An under-recognized entity? *European Journal of Pediatric Neurology,*

25(1), 187-190. https://doi.org/10.1016/j.e.jpn.2020.01.005.

Mahler, L. A., & Jones, H. N. (2012). Intensive treatment of dysarthria in two adults with down syndrome. *Developmental Neurorehabilitation, 15*(1), 44-53. https://doi.org/10.3109/17518423.2011.632784.

Miles, J. H., Takahashi, N., Muckerman, J., Nowell, K. P., & Ithman, M. (2019). Catatonia in down syndrome: Systematic approach to diagnosis, treatment and outcome assessment based on a case series of seven patients. *Neuropsychiatric Disease and Treatment, 15*(1), 2723-2741. https://doi.org/10.2147/NDT.S210613.

Mircher, C., Cieuta-Walti, C., Marey, I., Rebillat, A. S., Cretu, L., Milenko, E., Conte, M., Sturtz, F., Rethore, M. O., & Ravel, A. (2017). Acute regression in young people with down syndrome. *Brain Sciences, 7*(6), 57-66. https://doi.org/10.3390/brainsci7060057.

Page, C. G., & Johnson, K. (2021). Electropalatographic therapy and speech production for children with down syndrome. *Perspectives of the ASHA Special Interest Groups, 6*(485-493). https://doi.org/10.1044/2021_PERSP-20-00175.

Palat, P., Hickey, F., Patel, L., & Sannar, E. (2018). Levodopa-responsive early-onset parkinsonism in down syndrome. *Case Reports in Neurological Medicine, 2018*(2314791), 1-3. https://doi.org/10.1155/2018/2314791.

Rosso, M., Fremion E., Santoro, S. L., Oreskovic, N. M., Chitnis, T., Skotko, B. G., & Santoro, J. D. (2019). Down syndrome disintegrative disorder: A clinical regression syndrome of increasing importance. *American Academy of Pediatrics, 145*(6), 1-11. https://doi.org/10.1542/peds.2019-2939.

Santoro, S. L., Cannon, S., Capone, G., Franklin, C., Hart, S. J., Hobensack, V., Kishnani, P. S., Macklin, E. A., Manickam, K., McCormick, A., Nash, P., Oreskovic, N. M.,

Patsiogiannis, V., Steingass, K., Torres, A., Valentini, D., Vellody, K., & Skotko, B. G. (2020). Unexplained regression in down syndrome: 35 cases from an international down syndrome database. *Genetics in Medicine, 22*(1), 767-776. https://doi.org/10.1038/s41436-019-0706-8.

Takeuchi, C., Kanbara, Y., Minatogawa, M., Shibata, N., & Mochizuki, Y. (2017). Dementia and acute intellectual regression in down syndrome. *Journal of the Neurological Sciences, 381*(1), 1021. https://doi.org/10.1016/j.jns.2017.08.2882.

Worley, G., Crissman, B. G., Cadogan, E., Milleson, C., Adkins, D. W., & Kishnani, P. S. (2015). Down syndrome disintegrative disorder: New-onset autistic regression, dementia, and insomnia in older children and adolescents with down syndrome. *Journal of Child Neurology, 30*(9), 1147-1152. https://doi.org/10.1177/0883073814554654.

Yoder, P., Camarata, S., & Gardner, E. (2005). Treatment effects on speech intelligibility and length of utterance in children with specific language and intelligibility impairments. *Journal of Early Intervention, 28*(1), 34-39. https://doi.org/10.1177%2F105381510502800105.

Yoder, P. J., Camarata, S., & Woynaroski, T. (2016). Treating speech comprehensibility in students with down syndrome. *Journal of Speech, Language, and Hearing Research, 59*(1), 446-459. https://doi.org/10.1044/2015_JSLHR-S-15-0148.

Vita

Courtney Hale was born in St. Louis, Missouri. She graduated from Barat Academy in Missouri in May 2016. The following August, she began studying Neuroscience at the University of Rochester. She was awarded the Bachelor of Science degree in May 2020. Ms. Hale began her studies at Appalachian State University in June 2020. She was a member of Appalachian State's iSHINE program throughout the 2020-2021 academic year, and she worked as a graduate assistant throughout the 2021-2022 academic year. In December 2022, Ms. Hale received the degree of Master of Science in Speech-Language Pathology.