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Implications of Huntington's Disease on Daily Living: An Educational Tool and Guide for Occupational Therapists Treating Individuals with Huntington's Disease

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**Implications of Huntington's Disease on Daily Living: An Educational Tool and
Guide for Occupational Therapists Treating Individuals with Huntington's Disease**

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APPROVAL PAGE

This Scholarly Project Paper, submitted by Jacquelin Jones and Megan Thompson in partial fulfillment of the requirement for the Degree of Master's of Occupational Therapy from the University of North Dakota, has been read by the Faculty Advisor under whom the work has been done and is hereby approved.

Faculty Advisor

Date

PERMISSION

Title: Implications of Huntington’s Disease on Daily Living: An Educational Tool and Guide for Occupational Therapists Treating Individuals with Huntington’s Disease

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ABSTRACT

Huntington's disease (HD) is a neurodegenerative disease that can cause stress on the individual as well as their family members and caregivers. Due to the rareness of Huntington's disease (HD), it is common that professionals are often at a loss when treating patients who have it. Dawson et al. (2004) discussed the need for individualized and client-centered care for those individuals diagnosed with HD, as well as providing practical supports for those diagnosed and their families. Furthermore, a major challenge to assessment and intervention is the complexity of the symptoms present with HD. Etchgary (2011) found in her study that caregivers and those diagnosed with HD discovered many primary care providers lacked the foundational knowledge of HD and often were not able to find a correct diagnosis for the symptoms presented in the initial phase, thus contributing to the problem of lack of supportive care for this client population.

Implications of Huntington's Disease on Daily Living: An Educational Tool and Guide for Occupational Therapists Treating Individuals with Huntington's Disease, was developed to address the continuing need for supportive care in healthcare for those affected by Huntington's disease. A literature review was conducted to identify evidence based and best practices. The guide is designed to address various occupational components affected by Huntington's disease thus increasing quality of care, and decreasing lack of supportive care for individuals affected by Huntington's disease and their caregivers.

CHAPTER I

Introduction

Huntington's disease (HD) is a neurodegenerative disease that can cause stress on the individual as well as their family members and caregivers. It is a rare disease that affects 10-20,000 individuals (UCSF Memory and Aging Center, 2013). The formal definition of Huntington's disease as stated by the UCSF Memory and Aging Center (2013) is as follows: degeneration of neuronal structures in the brain and basal ganglia, that are responsible for movement and coordination. The disease also affects circuitry structures in the brain primarily responsible for thought, perception, emotions, and memory.

Due to the rareness of Huntington's disease (HD), it is common that professionals are often at a loss when treating patients who have it. Dawson et al. (2004) discussed the need for individualized and client-centered care for those individuals diagnosed with HD, as well as providing practical supports for those diagnosed and their families. Furthermore, a major challenge to assessment and intervention is the complexity of the symptoms present with HD. Etchgary (2011) found in her study that caregivers and those diagnosed with HD discovered many primary care providers lacked the foundational knowledge of HD and often were not able to find a correct diagnosis for the symptoms presented in the initial phase, thus contributing to the problem of lack of supportive care for this client population. The symptoms related to movement can include: clumsiness, rigid muscles, loss of balance, difficulty walking, uncontrolled movements of arms, legs and trunk (UCSF Memory and Aging Center, 2013). Symptoms related to personality, cognition and mental health function might include:

hallucinations, delusions, neglect of appearance and hygiene, neglect of responsibilities, depression, irritability, and cognitive difficulties as well as withdrawal (UCSF Memory and Aging Center, 2013).

Huntington's disease is delineated by three stages in its progression; early, middle, and late stage. The early stage is marked by the onset of a triad of symptoms including physical, behavioral and cognitive impairments (UCSF Memory and Aging Center, 2013). The middle stage according to HDSA (2010), is marked by an increase in motor control problems as well as cognitive deficits that begin to have a greater impact on an individual's abilities to carry out Activities of Daily Living (ADLs). Last, the late stage is characterized as the loss of muscle, the presence of contractures, increased involuntary control of movement, even more severe cognitive deficit, and the need for increased safety precautions (HDSA, 2010).

As the disease progresses, independence in activities of daily living decrease and coping with these changes becomes more challenging. Activities of Daily Living is defined as, "Activities oriented toward taking care of one's own body. (AOTA, 2014)" These tasks are fundamental to an individual's basic needs of survival and well being in the societal world (AOTA, 2014). Occupational therapists (OTs) can be an essential team member to aid in managing symptoms and providing education and resources to those affected by HD and their families. Occupational therapy as stated by the *Occupational Therapy Practice Framework: Domain and Process 3rd Edition* (AOTA, 2014), is:

The therapeutic use of everyday life activities (occupations) with individuals or groups for the purpose of enhancing or enabling participation in roles, habits, and routines in home, school, workplace, community, and other settings. Occupational therapy practitioners use their knowledge of the transactional relationship among the person, his

or her engagement in valuable occupations, and the context to design occupation-based intervention plans that facilitate change or growth in client factors (body functions, body structures, values, beliefs, and spirituality) and skills (motor, process, and social interaction) needed for successful participation (Pp. S1).

For the project, the primary concerns are the need for supportive care for those with HD and what treatments are established and up and coming for HD specifically relating to the practice of occupational therapy. In addition, a main focus will be on what an occupational therapist can do to treat an individual with HD in a client-centered and progressive approach, while helping the caregiver as well as based on evidence-based and best practice standards.

To be able to develop a well-rounded guide for OTs, an occupation-based model was chosen to guide the process of developing this project. The Canadian Model of Occupational performance and Engagement was chosen because of the key concepts outlined in the model such as Spirituality, occupational engagement, and a primary focus on client-centered practice. “Spirituality does not refer to religion, but the essence of self, the place where determination and meaning are drawn (Polatajko, Townsend, & Craik, 2007)” Engagement is one of the main concepts covered in this model because it offers a less restricted focus on occupational performance (Polatajko, Townsend, & Craik, 2007). Although a person may not be able to occupationally perform in their desired occupations, they can still engage in them through the help of modification, adaptation and caregiver role (Polatajko, Townsend, & Craik, 2007). Last, the model describes the therapeutic process from a client’s perspective which is essential because the client is the agent of change while the therapist enables the process of change through empowerment and enables the skills of the client, thus the use of client-centered practice (Polatajko, Townsend, & Craik, 2007).

By developing a comprehensive program that focuses on the implications of the disease, and treatment strategies for all stages and areas affected in the individual's life it is believed that the quality of care can be dramatically increased. Occupational therapy can play a vital role in delivering client-centered service for progressive care and modifications for the individual affected.

Key Terms and Concepts

1. Basal ganglia: is located deep within the cerebral hemispheres in the telencephalon region of the brain. Its function is to contribute to controlling cognition, movement coordination and voluntary movement.
2. Canadian Model of Occupational performance and Engagement (CMOP-E): occupational order has six perspectives – physical rehabilitative, psycho-emotional, socio-adaptive, neurointegrative, developmental or environmental – in relation to the arbitrary performance areas of self-care, leisure and productivity. Quality of function is assessed in terms of both performance and satisfaction, Disorder may occur in the dimensions of person, occupation or environment, or when the momentum of experience is lost due to unresolved issues. Intervention aims to improve transactions between person, occupation and environment, through the process of enablement rather than treatment. Enablement involves working with clients to facilitate autonomy, and does not focus primarily on performance components (Polatajko, Townsend, & Craik, 2007)
3. Charcot-Marie-Tooth (CMT) Disease: one of the most common inherited neurological disorders, also known as hereditary motor and sensory neuropathy (HMSN) or peroneal muscular atrophy. CMT is caused by mutations in genes that produce proteins involved in the structure and function of either the peripheral nerve axon or the myelin sheath.

Consequently, these nerves slowly degenerate and lose the ability to communicate with their distant targets. The degeneration of motor nerves results in muscle weakness and atrophy in the extremities (arms, legs, hands, or feet), and in some cases the degeneration of sensory nerves results in a reduced ability to feel heat, cold, and pain. (National Institutes of Health, 2-23-15)

4. Cortical Regions: Any of various regions of the cerebral cortex
5. Cortico-striatal Degeneration: A heterogeneous group of degenerative syndromes marked by progressive cerebellar dysfunction either in isolation or combined with other neurologic manifestations. Sporadic and inherited subtypes occur. Inheritance patterns include autosomal dominant, autosomal recessive, and X-linked. (Healthdictionary.info, 2015, ¶1)
6. Cytosine-Adenine-Guanine or CAG triple: Cytosine thymine guanine and adenine are the nitrogenous bases found in the nucleotides that form DNA. DNA is made up of 3 parts: (1) Phosphate group (2) Deoxyribose (3) Nitrogen containing base.
7. Deoxyribonucleic acid (DNA): It's the molecule that stores genetic information in an organism. That makes the nucleotide the most basic subunit of DNA, or, more generally, of any nucleic acid. (Chin, 2015)
8. Huntington's Disease (HD): degeneration of neurons of structures deep within the brain, the basal ganglia, which are responsible for movement and coordination. Structures and circuitry responsible for thought, perception, emotions and memory are also affected, likely due to connections from the basal ganglia to the frontal lobes (UCSF Memory and Aging Center, 2013, pg. 1).
9. Magnetic Resonance Imaging (MRI): a test that uses a magnetic field and pulses of radio wave energy to make pictures of organs and structures inside the body. (WebMD)

10. Multiple System Atrophy (MSA): a progressive neurodegenerative disorder characterized by symptoms of autonomic nervous system failure such as fainting spells and bladder control problems, combined with motor control symptoms such as tremor, rigidity, and loss of muscle coordination. Although what causes MSA is unknown, the disorder's symptoms reflect the loss of nerve cells in several different areas in the brain and spinal cord that control the autonomic nervous system and coordinate muscle movements. (National Institutes of Health 2015, ¶1)
11. Neuropsychiatric: The branch of medicine dealing with diseases involving the mind and nervous system. (Dictionary.com, 2015, ¶1)
12. Occupations: “daily life activities in which people engage (Framework, 2014, pg. S6)
13. Progressive Supranuclear Palsy (PSP): a rare brain disorder that causes serious and progressive problems with control of gait and balance, along with complex eye movement and thinking problems. One of the classic signs of the disease is an inability to aim the eyes properly, which occurs because of lesions in the area of the brain that coordinates eye movements. Some individuals describe this effect as a blurring. Affected individuals often show alterations of mood and behavior, including depression and apathy as well as progressive mild dementia.(National Institutes of Health, 2015, ¶1)
14. Post Polio Syndrome (PPS): refers to a cluster of potentially disabling signs and symptoms that appear decades — an average of 30 to 40 years — after the initial polio illness. (Mayo.org, 2015, ¶1)
15. Psychomotor: of or relating to a response involving both motor and psychological components. (Dictionary.com, 2015, ¶1)

16. Quality of Life: the degree of satisfaction an individual has regarding a particular style of life. Although assessment tools are available to evaluate physical and social dimensions, an individual's general sense of well being or satisfaction with the attributes of life is more difficult to evaluate (medical-dictionary.thefreedictionary.com/quality of life, 2015, ¶1)
17. Ribonucleic acid (RNA): a nucleic acid found in all living cells, constituting the genetic material in the RNA viruses, and playing a role in the flow of genetic information; it is a linear polymer which on hydrolysis yields adenine, guanine, cytosine, uracil, ribose, and phosphoric acid and which may contain extensive secondary structure (medical-dictionary.thefreedictionary.com/quality of life, 2015, ¶1)

Chapter II presents the results of a comprehensive literature review in addition to an overview of the product. A review of literature regarding the etiology and symptomatology of HD, what health care and caregiver challenges are present in research, how quality of life is affected for the individual with HD and caregiver, what the OT role is, the theory basis for this project, and what the proposed programming will look like and entail for the product will be discussed. Chapter III will present the methodology and the activities used to develop the product. The product in its entirety is available in Chapter IV. Finally, Chapter V is a summary of the project and includes recommendations and limitations of the product.

CHAPTER II

Introduction

Huntington's disease (HD) is a "rare" debilitating condition that affects 10-20,000 individuals (UCSF Memory and Aging Center, 2013). As the disease progresses, independence in activities of daily living decrease and coping with these changes becomes more challenging for both the individual and their caregivers. One challenge is that due to the rarity of Huntington's disease, it is common that professionals are often at a loss when treating patients who have it. A major challenge to assessment and intervention is the complexity of the symptoms.

Occupational therapists (OT's) can be an essential team member to aid in managing symptoms and providing education and resources to those affected by HD and their families. The primary concerns are the need for supportive care for those with HD and what treatments are established and current for HD. A thorough literature review was conducted to more fully understand the depth of the symptomatology assuring the needs of the individual were going to be addressed. The literature review focused on identifying evidence-based research on the efficacious treatments and therapies that can be used to preserve the quality of life of the individual as well as family and caregivers. This chapter presents information which has resulted in the development of an educational tool and guide for occupational therapy practitioners.

Definition

As stated prior, little is known in the OT literature. So an initial step is to establish a basic definition of what Huntington's Disease(HD) is. HD, is a rare neurodegenerative disease that affects one in every 10-20,000 individuals and has typical onset in mid-adulthood (30-55) in the United States (Memory and Aging Center at the University of California, San Francisco, 2013), It is estimated that approximately five to seven people in 100,000 are affected by HD in western countries (World Health Organization, 2014). The formal definition of HD is as follows:

Degeneration of neurons of structures deep within the brain, the basal ganglia, which are responsible for movement and coordination. Structures and circuitry responsible for thought, perception, emotions and memory are also affected, likely due to connections from the basal ganglia to the frontal lobes (UCSF Memory and Aging Center, 2013, pg. 1).

The disease is initially characterized by physical, cognitive, and emotional/behavioral changes. Physical changes include: involuntary movement, dysarthria, poor coordination, and loss of balance. Cognitive changes are present in memory loss, disorganization, and the loss of the ability to multitask. Emotional/behavioral changes encompass: depression, anger, withdrawal, and paranoia (UCSF Memory and Aging Center, 2013). The etiology is complex and its influence on the body is significant in its progression. The next section adds more detail on the etiology of the disease process.

Etiology

Genetic Factors

HD is caused by a cytosine-adenine-guanine or CAG triplet repeat expansion within the huntingtin gene, which encodes an expanded polyglutamine stretch in the huntingtin protein

(Pringsheim et. al., 2012). Guanine, cytosine, and adenine are three of the four nucleobases found in the nucleic acids of deoxyribonucleic acid or DNA and ribonucleic acid or RNA. The specific combination of cytosine-adenine-guanine can be found both DNA and RNA (Genetic Home Reference, 2014).

Huntingtin is a protein that is thought to play an important role in nerve cells in the brain (Genetic Home Reference, 2014). The huntingtin protein is found within various tissues throughout the body; however is most recognized within the brain. It is predicted that the protein plays an important role in chemical signaling, attaching to other proteins and structures, transporting materials, and protecting the cell from apoptosis or self-destruction (Genetic Home Reference, 2014).

One particular part of the huntingtin protein holds a DNA segment dedicated to the CAG repeat. This is the segment that is abnormally altered in Huntington's disease. (Genetic Home Reference, 2014) "The gene is inherited in an autosomal dominant manner with age-dependent penetrance (Pringsheim et. Al., 2012, p. 1083)." The average tract length of CAG repeats in the general population is 16 to 20, and with the diagnosis of HD this tract expands to 36 repeats or greater (Pringsheim et. al., 2012). According to Genetic home Reference (2012), signs and symptoms of HD may or may not evolve in an individuals with 36 to 39 CAG repeats, whereas people with 40 or more repeats almost always develop the disorder.

With abnormal CAG repeat, an altered and longer version of the huntingtin gene is produced. This protein is cut into small, abnormal pieces that bind together and multiply in neurons, which in turn affects the normal function of the neuronal cell. The regions where this is the most prevalent are in the brain, particularly in the striatum and the cerebral cortex. Disruption

and eventual cell death of neurons take place in these regions, thus outlining the typical symptoms of HD (Genetic Home Reference, 2014).

Delmaire et al. (2013), explain that many studies have been performed with Huntington's disease and identifying patterns of neuronal loss in the striatum, and other brain regions including the cortex and Brodmann areas eight, nine, and ten through magnetic resonance imaging (MRI). However, limited research has been conducted on the effects of HD specifically on the cortico-basal ganglia. The researchers in this study hypothesized that selective psychomotor, cognitive and neuropsychiatric deficits are implicated partly by lesions caused to not only the basal ganglia, but the cortical regions as well (Delmaire et al., 2013). The results showed that after using verbal based DTI analysis, that cognitive, behavioral, and motor deficits were associated with regionally specific cortico-striatal degeneration. With these results it can be determined that degeneration, associated with HD, is not only subcortical, but also cortical damage as well. These results add to the body of evidence used to determine what areas are affected with HD and how to better determine diagnosis and stages with clinical expression of symptoms (Delmaire et al., 2013). While the body systems are affected at the cellular level, specific characteristics and symptoms become present with the onset of HD.

Symptomatology

The presentation of symptoms for HD is complex. While there are markers in symptomatology that indicate stages of progression, there are also symptoms that can occur that are not well established under a certain stage of progression, such as symptoms related to movement and/or personality changes.

Symptoms related to movement that are associated with HD can occur at any stage of progression, and become more severe as progression occurs through the early, mid and late

stages of HD. These symptoms may include clumsiness, rigid muscles, and loss of balance, difficulty walking, uncontrolled movements of arms, legs and trunk. There may also be disruptions in the cerebral cortex, which in turn affects coordinated movement, thinking, and emotions (Genetic Home Reference, 2014).

Similarly, symptoms related to personality, cognition and mental health function might influence hallucinations, delusions, neglect of appearance and hygiene, neglect of responsibilities, depression, irritability, and cognitive difficulties as well as withdrawal (UCSF Memory and Aging Center, 2013). To better understand the clinical presentation of an individual affected by HD in each stage of progression, the early, mid, and late stages of HD will be further analyzed. This analysis will include a breakdown of what symptoms may be present at each stage, what inabilities start to occur, and what the caregiver's role will become in each stage.

Early Stage

The early stage of HD is defined as having the presence of a triad of symptoms including physical, behavioral and cognitive impairments (UCSF Memory and Aging Center, 2013). HDSA, 2010 state that early stage HD is most often characterized by, "difficulty starting, stopping and sequencing actions (pp. 27)." Changes in cognition are also present in the early stages of HD. These changes include diminished attention span and concentration, reduced short term memory and new learning capabilities, trouble beginning and ending activities, difficulty repeating the same thoughts or ideas, trouble controlling impulses, and increased irritability or outbursts (HDSA, 2010).

In the early stages of HD, movement disorder symptoms may or may not be present. If present the symptoms could include, impaired balance mobility, and/or motor control

impairment. Involuntary movements may become present in this stage, and can affect meal preparation skills, personal hygiene and other activities of daily living (ADLs).

In this stage, it is important to consider the safety of the individual affected with HD and their abilities to continue living with independence in their homes. It becomes the role of the caregiver to provide a safe environment for the individual to live in. Given the added safety implications that become present with this stage of progression, occupational therapy can serve as a vital resource for environmental modification and safety training and awareness.

Mid Stage

In the mid stages of HD, motor control problems as well as cognitive deficits begin to have a greater impact on an individual's abilities to carry out ADLs (HDSA, 2010). The individual's memory begins to deteriorate and they start to forget the things that they once knew, such as previously learned habits or tasks that they once completed on a daily basis (HDSA, 2010). The mid stage is also marked as the onset of increased severity with motor control problems in combination with cognitive deficits. This creates numerous challenges to activities of daily living and engagement in occupations for the individual (HDSA, 2010).

According to HDSA 2010, people who are in the mid stage of HD may have difficulties starting a task and once started, may fatigue quickly. Experiencing fatigue will hinder the person from completing a task or role that they particularly carry out every day. More specifically, fatigue will also affect the person's ability to eat, carry out hygiene tasks, and completing their daily dressing tasks.

Due to motor control problems associated with the mid stage, eating is a common area of difficulty. Impaired postural control affects the person while sitting at a table or in a chair and slouched sitting causes a great deal of food spillage (HDSA, 2010). Motor impersistence and

muscle atrophy can cause difficulty with holding eating utensils, coordinating the movement from the plate to the mouth as well as cutting or scooping up food (HDSA, 2010). Maintaining balance, and doing fine motor tasks, while performing ADL's, may be hard for the individual due to motor control issues.

Occupations such as functional mobility and community mobility may become more challenging due to these motor control issues. Functional mobility, as defined by the American Occupational Therapy Association is the ability to move from one place to another including transfers and functional ambulation (2014). Additionally, community mobility is defined as being able to move around in the community through private or public transportation such as driving, walking, bicycling and/or using transportation systems (AOTA, 2014). With the presence of motor impersistence and involuntary movement, it becomes increasingly harder to remain independent with these occupations. Due to the individual having increased reliance on a caregiver to carry out their occupations or taking them over fully, the individual affected by HD may start to experience greater bouts of depression.

At the mid stage of HD, depression is increased due to the inability to be independent and experience the feeling of accomplishment of completing one's own daily occupations. Impairments have increased which is associated with diminished ability for a person to grasp an object. Evidence of limited grasp is usually reported and signified by atrophied muscles in the person's hands. When a person is not able to be independent, it greatly affects their sense of self worth and satisfaction with life's roles. For example, hygiene may be limited due to the individual experiencing lack of motivation as well as having decreased cognition, which it becomes a chore for the person to sequence each task and associated movement (HDSA, 2010).

Sequencing as defined by as a process required to carry out a desired activity (AOTA, 2014). Within each activity, specific steps sequencing and timing requirements are required for successful completion. For example, the sequencing for brushing ones teeth include: picking up the toothbrush, turning on the faucet, getting the toothbrush wet, opening the cap of the toothpaste, applying the toothpaste onto the brush, bring toothbrush up to the mouth and brushing in a circular motion onto teeth for a timed two minutes. This activity can pose as a problem for individuals in the mid stage of HD, due to the several step task and sequencing the steps it takes to brush one's teeth being too complex because it is hard to comprehend with the presence of cognitive impairment. Due to the increasing cognitive and motor impairments present in the mid stage of HD, the caregiver takes on a larger role of care for the individual's care and safety.

Late Stage

Late-stage of HD is defined as the loss of muscle, the presence of contractures, increased involuntary control of movement, even more severe cognitive deficit, and the need for increased safety precautions (HDSA, 2010). It is discussed that care for those with HD is complex due to the array of symptoms that are present with the disease (Simpson, 2007).

At this stage of the disease, the utmost important task is to keep the person safe, adapt and modify their environments, and prevent injury due to continuous movements (HDSA, 2010). Environmental safety will be important as to not have any sharp objects in the room by padding hard furniture which will prevent injury from falls or choreic movements (HDSA, 2010). Contractures may or may not be present within this stage. Other common concerns in this stage are the person's skin breaking down due to the inability to voluntarily change positions (HDSA, 2010). With the progression of HD, care in the home becomes increasingly difficult to

accomplish due to the severity of the symptoms. Frequently, care is shifted to an institutional setting such as a nursing home or long term care unit (Simpson, 2007).

From the family or caregiver's view it has been noted that many families wish to have a better plan devised for the maintenance and care of their family member affected by HD (Simpson, 2007). This includes gathering resources in the community to utilize, and planning for individual care in the future. From the patient's view many themes have been outlined for how they would like things to be handled in their current and future care (Simpson, 2007). This includes determining what their future care plan will look like, and what they want and do not want in the end stages for life saving options such as the placement of feeding tube to prolong life (Simpson, 2007).

A common theme that has been identified is that many doctor do not encourage patients diagnosed with HD to devise a plan for long-term care while they are still competent (Simpson, 2007). This poses a problem in the late stages of the disease; because the family or caregiver is left to make decision that they are not sure the patient will be comfortable with or agree upon (Simpson, 2007). Therefore, with this care plan devised by the author, Simpson, it is encouraged for families and their family member with HD to develop a care plan or advanced directive in the early stages of the disease so that the patient is still able to have a say with their care, and the transition of care is easier for the family member or caregiver to complete in the late stages of progression with HD (Simpson, 2007). Due to the complexity of symptoms that are present with HD, there are a few challenges that are present for those afflicted by HD, their caregivers and healthcare professionals.

Challenges

There is a noticeable lack of knowledge about HD in health care as well as minimal supports for families and individuals affected by the HD (Etchegary, 2011). Not only is there a lack knowledge of the diagnosis itself due to its rarity, there is a noticeable lack in literature and education on how to effectively treat and refer for an individual affected by HD. This includes primary care physicians, therapy professions, social work and psychologists.

It is important to understand that there is a noticeable lack of care and knowledge for the occupational therapy profession as occupational therapists, currently depend on referral from a primary care physician to treat. Without proper referral, occupational therapy, in general, is not able to adequately treat this population, therefore; it is of benefit to the healthcare professional to research and educate themselves to effectively treat and refer patients with HD.

Health Care Professional Challenges

According to Dawson, Kristjanson, Toye, and Flett (2004), the need for supportive care is amplified in their study. Dawson et al. (2004) discussed the need for individualized and client-centered care for those individuals diagnosed with HD, as well as providing practical supports for those diagnosed and their families. Furthermore, in a study conducted by Etchegary (2011), it was found that there was a need for improved care in the initial phases of the disease, such as the diagnosis and early stages and well into the mid and late stages of the disease when institutionalization normally happens. Another trend found within the study was that the caregivers and those diagnosed with HD discovered many primary care providers lacked the foundational knowledge of the disease and often were not able to find a correct diagnosis for the symptoms presented in the initial phase (Etchegary, 2011).

One study conducted by Harding, Stewart, and Knight (2012), assessed quality of life in

health-care workers who worked on a specialist Huntington's disease (HD) unit and their perceptions of how their contributions and other service provider's contributions affected quality of life in the patients diagnosed with HD. Service staff that was involved in this study included: dietetics, occupational therapy, nursing, physiotherapy, psychology, and speech and language therapy staff. (Harding, Stewart & Knight, 2012)

A theme of a multidisciplinary approach with the care team was outlined and the importance it has on delivering the best services to patients with HD. Many of the participants professed that due to the complexity of the symptoms present with HD that it takes several professions working together to deliver the best care. (Harding, Stewart & Knight, 2012)

Another theme found was communication, in regards to patient to professional communication. Many of the participants stated that when communication becomes impaired in the patients they are working with, that they shift to a 'what is in their best interest' type of perception. Many of the participants described that one contributor to having to use the best interest policy was the lack of advance care directives being established before the patient with HD became too impaired to complete one. (Harding, Stewart & Knight, 2012)

This information previously presented opens up the doorway to a crucial conversation that needs to be happening among healthcare professionals. How can HD be effectively treated? Research provides a gateway into healthcare professionals understanding that there is a need for supportive care; this is the biggest challenge for healthcare professionals currently. Lack of knowledge and exposure to the disease, symptomology, and progression negatively affects a healthcare professional's abilities to effectively treat HD at any varying stage. With this being said, caregiver challenges are present as well with understanding the needs and supports of those diagnosed with HD.

Caregiver Challenges

This can be an increased burden if the caregiver is not sure of the supports they need to give and do not have any resources to utilize for help with the individual diagnosed with HD. In an article by Aubeeluck and Brewer (2008), the caregiver can experience a diminished quality of life when caring for a person with HD due to not being able to locate resources and supports for the person, dealing with healthcare professionals who do not understand the dynamics of a family with HD, and who have little knowledge of the disease as well. Even when the disease is appropriately classified, numerous individuals still vocalized issues with trying to receive various resources for their family member diagnosed with HD (Etchegary, 2011).

One participant in a study conducted by Etchegary (2011) voiced that:

It's been a really, really hard struggle. Everything I do, I have to fight, fight for support. Fight for a respite worker for my son, even a blender (pause) . . . there are things they won't give. They won't give him a cane. I couldn't afford it. These are things that they need. (Pg. 230)

The caregiver may feel an undue amount of stress while taking care of their loved ones affected by HD. The individuals with HD challenges, also needs to be taken into consideration as well.

Individuals Affected by HD Quality of Life

Calvert et al. (2013), conducted a study that assessed health-related quality of life in individuals diagnosed with rare long-term neurological conditions, and their access to supportive care for their disease. The conditions tested include Huntington's Disease (HD), Charcot-Marie-Tooth (CMT) disease, Multiple System Atrophy (MSA), Progressive Supranuclear Palsy (PSP), and Post Polio Syndrome (PPS).

The researchers found that health related quality of life was significantly lower compared to the general population. Reports of pain, depression, mobility impairments, and decreased ability in self-care and usual activities were also recorded. Also, results yielded that supportive and coordinated care given to these participants could be improved (Calvert et al., 2013)

The conclusion yielded that not only do rare long-term neurological conditions have an impact on health-related quality of life, but also that improved levels of care and social care services could potentially help increase health related quality of life in these patients (Calvert et al., 2013)

In a study conducted by Harding, Stewart, and Knight (2012), a major theme derived from their results was hopes, fears, and behaviors of patients affected by HD. The participants in this study, which were healthcare workers, explained that many of the patients they worked with had fears for what they would be like as the disease progressed and in a sense if their dignity would be lost after a certain stage of the disease. Service providers reported that they encouraged patients to stay positive and still set positive goals for themselves even in their condition, so that they would stay motivated (Harding, Stewart & Knight, 2012). With these results, this study provides a detailed analysis of the perceptions of healthcare workers working with individuals with HD and how they feel they contribute to their quality of life as well as perceptions of the patients themselves (Harding, Stewart & Knight, 2012). To better treat symptoms holistically that are present in each stage of HD, health care providers providing care to individuals and families have conducted numerous studies on implementing a multidisciplinary approach in treatment.

Impact of Multidisciplinary Approach on Clients

Thompson et al. (2013), conducted a study on a nine-month rehabilitation program that was implemented for patients in a multidisciplinary fashion. Motor function, cognition, depression, body composition, postural stability, and quality of life were measured for feasibility, safety and efficacy of the program. The multidisciplinary program, being implemented into an individual's life, was measured for its effect concerning efficacy, quality of life, and daily function. The study discussed how other diagnoses such as Alzheimer's and Parkinson's had been reviewed and treated using a multidisciplinary approach, but few had been conducted on Huntington's disease. (Thompson et al., 2013)

In Thompson et al. (2013), the study, a randomized control trial, had twenty participants who completed the entire study. The participants were recruited from the North Metropolitan Area Mental Health Service (NMAMHS) databases under the inclusion criterion of a positive genetic test for Huntington's disease, and a clinical diagnosis of the onset with the ability to follow verbal command and to perform submaximal exercise. The participants were split randomly into two groups and the intervention group was randomly assigned. The dual-energy X-ray absorptiometry, Sensory Organization Test, Activities-Specific Balance Confidence (ABC) Scale, Symbol Digit Modalities Test, Hopkins Verbal Learning Test-Revised, D-KEFS Colour Word Interference Test and Trail Making Trials, Beck Depression Inventory-II, Goal Attainment Scale, SF-36v2 Health Questionnaire, and Huntington's Disease Quality of Life Battery for Carers were all used to establish a preliminary outcome measure of all participants. (Thompson et al., 2013)

The interventions included exercise programs developed by physical therapy, and patient-focused programs developed by occupational therapy that focused on targeting deficits detected

by psychologists. The psychologists conducted a series of assessments targeted towards assessing cognitive deficits, and levels of depression, and in turn the occupational therapy assessed how these deficits hindered occupational performance. It was found that rehabilitation and programs implemented, using the multidisciplinary approach, decreased chorea, and slowed motor and postural deterioration. However, little effect was seen with cognitive strengthening. Overall, it is discussed that all Huntington's disease patients would benefit from a multidisciplinary program/rehabilitation approach, and that more research should be done in the future to investigate the implications and effects on cognitive performance. (Thompson et al., 2013)

Veenhuizen and Tibben (2009), also developed a plan of multidisciplinary care for HD patients within an outpatient clinic in their article. The multidisciplinary team consisted of a neurologist, psychologist, occupational therapist, speech and language therapist, social worker, social worker and nursing home doctor. The idea of addressing all factors and symptoms associated with the onset and progression of the disease prompted an urgent need for a program or protocol for adequately assessing, treating, and following up with patients and families. The team set up a project plan that addressed the process of referral, evaluation, treatment, referral, and further follow-up with patients. In the journal article two cases were provided for how the project plan was carried out, and the outcomes of the two cases. In both cases the participants reported higher satisfaction and quality of life with their care provided by the multidisciplinary team and other facilities involved with their care (Veenhuizen and Tibben, 2009). The team did not use an experimental design in this study, but rather report the implementation of a strategic project plan devised by the team of researchers, and how it was applied with two individuals in a case study fashion (Veenhuizen and Tibben, 2009). the focus of this scholarly project is on one

specific healthcare profession, occupational therapy. the next section will present the role of occupational therapy in relation to being a member of the multidisciplinary team.

The Role of Occupational Therapy

The primary concerns with the need for supportive care for those with HD are: what treatments are currently available for HD as well as what an occupational therapist can do to treat an individual with HD in a client-centered and progressive approach, while helping the caregiver as well. Occupational therapists can step in to help manage symptoms and provide education and resources to those affected by HD and their families.

Occupational therapy provides an array of services that would benefit those diagnosed with HD and their caregivers. According to the *Occupational Practice Framework: Domain and Process 3rd Edition* (2014), Occupational therapy is defined as, “The therapeutic use of everyday life activities (occupations) with individuals or groups for the purpose of enhancing or enabling participation in roles, habits, and routines in home, school, workplace, community, and other settings. (AOTA, pp. S1)” Furthermore, it is described that the occupational therapy practitioner,

Uses their knowledge of the transactional relationship among the person, his or her engagement in valuable occupations, and the context to design occupation-based intervention plans that facilitate change or growth in client factors (body functions, body structures, values, beliefs, and spirituality) and skills (motor, process, and social interaction) needed for successful participation. Occupational therapy practitioners are concerned with the end result of participation and thus enable engagement through adaptations and modifications to the environment or objects within the environment when needed. (AOTA, 2014, PP. S1)

Occupational therapy can play a vital role in assessing, treating, and providing adaptations for those affected by HD and their caregivers. Occupational therapists have foundational knowledge in cognitive, psychosocial, and physical assessments relevant to the symptomatology and presentation of HD. Furthermore, intervention is client-centered and directed by client needs and abilities in regards to cognitive, psychosocial and physical functional capacities.

Occupational therapist's can also be a key contributor on a multidisciplinary team by providing valuable insight into clinical presentation. Occupational therapist's specialize in activity analysis and in understanding the individual in terms of functional capacities in their lives. They can also provide and suggest adaptations and modifications for individuals affected by HD for a better quality of life. Overall, occupational therapy is a healthcare profession that can compliment the multidisciplinary team in terms of treating an individual with HD.

Occupational Therapy Assessment

There is a gap in literature for occupational therapists utilizing evidence-based treatments and interventions with people who have been diagnosed with HD. Furthermore, there is even less literature pertaining to effective assessments used to measure functional abilities environmental modifications, and safety needs for individuals afflicted by HD. Though it is a rare disease, occupational therapy professionals are in dire need to be more informed because HD is a result of motor control loss and decreased cognitive ability, which in turn diminishes the patient's ability to live independently (Bylsma, Rothlind, Hall, Folstein & Brandt, 1993). With the previous problems stated, this affords the profession of occupational therapy the chance to assert their involvement with developing evidence-based, and relevant assessments for measuring functional capabilities, environmental modification and safety needs for individuals with HD.

Although there is limited information on OT related assessments for individuals with HD there are definite focus areas that an OT can assess that can identify the needs of these individuals. Occupational therapists are trained to assess cognitive areas, psychosocial aspects, and physical abilities of a person. Cognitively, occupational therapists look at the person in a holistic view to adapt or grade activities specific to that person so that they are able to engage in occupations that are meaningful. Psychosocially, occupational therapists assess peoples mental capacities, emotional regulation skills, and activity demands needed to complete an occupation requiring psychosocial aspects. Physically, occupational therapists are trained to assess range of motion, strength and weaknesses, disabilities, and anything that may be preventing them from engaging in occupations.

Based upon the needs of the clients an occupational therapist can provide skilled intervention planning for the individual and caregivers. Various interventions and strategies have been identified for treating individuals with HD, and can be utilized by occupational therapy to help manage symptoms and maintain the highest possible quality of life.

Interventions and Strategies

Although there is no cure or way to stop the progression of HD from happening, Imbriglio, Tarapata, and Lovecky, 2010 imply that there is no current treatment for HD, but there are many ways for occupational therapists, physical therapists and speech therapists to help address the movement disorders found in HD as well as the psychosocial needs of the individual and caregiver. Occupational therapists are academically prepared to meet the needs of individuals' with HD. It has to begin by not looking at the diagnosis but looking at the occupational needs and challenges of the individual regardless of the disability. So it is essential to begin by using the OT Framework as a starting point for the assessment, intervention and

strategy identification. The next step is to use the strength of the activity analysis and occupational profile to ensure all aspects of OT intervention is client centered. All of this is pulled together by the use of an occupation-based model.

Summary

The etiology and presentation of HD is complex, making it a challenge to treat (UCSF Memory and Aging Center, 2013). Symptoms can be moderately delineated within each stage, however; due to the complexity in presentation of the symptoms HD is noted as a difficult disease to treat within the healthcare realm (Simpson, 2007). Research has outlined that there are noticeable gaps in treatment due to lack of knowledge and education about the disease, which leads to lack of supportive care for those affected by HD and their caregivers (Etchegary, 2011). Research has been conducted on the effectiveness of implementing a multidisciplinary approach to treating HD with promising results (Thompson et al., 2013).

The healthcare profession of occupational therapy can play a vital role in the life of a person diagnosed with HD and their family; however, the education of what occupational therapy can do is dismal (Aubeeluck and Brewer, 2008). With that being said occupational therapy has the opportunity to advocate for its scope of practice and involvement in effectively treating individuals with HD and their families and caregivers. Occupational therapy also has the chance to delineate their effective role of working with a multidisciplinary care team to successfully treat and serve this population. Based on the review of literature, an occupational guide has been developed to inform occupational therapists of assessments, interventions and strategies that can be used with this population through each of the stages.

Proposed Program

An educational guide: *Implications of Huntington's Disease on Daily Living:*

An Educational Tool and Guide for Occupational Therapists Treating Individuals with Huntington's Disease, was designed for occupational therapists to gain competence and confidence in working with this population. This guide provides best practice interventions and assessments that are applicable to this population to effectively treat individuals with HD in all stages. The guide is formatted by stages of diagnosis of HD and provides the occupational therapist with definitions of each stage as well as what it may look like on certain levels such as: occupational, personal, environmental, and, spiritual level.

Model

The Canadian Model of Occupational Performance and Engagement was chosen to guide the development of the guide and the assessment, and intervention process. A primary reason for choosing the Canadian Model of Occupational Performance and Engagement (CMOP-E) was due to the necessity of addressing spirituality that is needed for a client to stay motivated and engaged while having a disease such as Huntington's disease. Not only is it common for people to experience loss of self worth, but also direction without self worth present.

The CMOP-E also discusses a concept of enablement.

Occupational therapy is the art and science of enabling engagement in everyday living, through occupation; of enabling people to perform the occupations that foster health and well-being; and of enabling a just and inclusive society so that all people may participate to their potential in the daily occupations of life. (Townsend et al., 2007, pp. 89)

Enablement is a concept drawn upon by the developers of the CMOP-E to establish that the individual does not need to be physically capable to participate, but rather be motivated and able to engage in their desired occupations.

The model influenced the development of the guide due to the CMOP-E being based off of a client's perspective, which is essential when treating individuals with HD, because the client is the agent of change while the therapist enables the process of change through empowerment and enables the skills of the client.

Occupational Therapy Practice Framework also provided the organization of the model in terms of using areas of occupation, the client factors of the person and the elements of environment that can be adapted or modified to help enable and engage the individual with Huntington's disease.

Organization of Proposed Program

The proposed program was developed in such a way, that the occupational therapist begins to develop a holistic perspective into the treatment of HD according to the application of the CMOP-E. Initially, the program helps the reader develop an understanding for the CMOP-E and how it is specifically implemented in regards to assessment and intervention strategies pertaining to those affected by HD and their caregivers. Next, the program provides an in-depth look into differing assessments that can be implemented in differing stages and for differing symptomology present with HD. then, the program takes a step-by-step look at the breakdown of symptomatology between stages, clinical presentation, characteristics, and strategies for intervention. An overview of the product can be seen in the table of contents below:

Table of Contents

- ❖ ***The Canadian Model of Occupational Performance and Engagement - An Overview***
- ❖ **Assessment**
 - Table 1.1
- ❖ ***Introduction to Interventions***
- ❖ **Early Stage Huntington's Disease**
 - Research Literature

- Table 1.2
- ❖ ***Mid Stage Huntington's Disease***
 - Research Literature
 - Table 1.3
- ❖ ***Late Stage Huntington's Disease***
 - Research Literature
 - Table 1.4
- ❖ ***Resources***
 - Table 1.5
- ❖ ***References***

Conclusion

HD is a debilitating condition that can cause stress on not only the affected individual, but their families as well. With the progression of the disease, greater dependence on a caregiver becomes apparent and vital for maintained quality of life. Without having a team of healthcare professionals that not only understand the implications and course of the disease, but also provide quality client-centered care, life in general is diminished for the caregiver and person with HD.

As the disease progresses, independence in activities of daily living decrease and coping with these changes becomes more challenging. Occupational therapists can be an essential team member to aid in managing symptoms and providing education and resources to those affected by HD and their families. The primary concerns are the need for supportive care for those with HD and what treatments are established and up and coming for HD. In addition the focus will be on what an occupational therapist can do to treat an individual with HD in a client-centered and progressive approach, while providing the caregiver with the support he/she needs as well.

Occupational therapy can play a vital role in delivering client-centered service for progressive care and modifications for the individual affected, and supports and resources to the

families and caregivers. By developing a comprehensive program that focuses on the implications of the disease, treatment strategies for all stages and areas affected in the individual's life, and identifying resources that are helpful for the individual and caregivers, quality of care can be dramatically increased for families impacted by HD.

In conclusion, it is the hope of the authors that the occupational therapy practitioner will develop an understanding of HD, clinical treatment of HD, and more importantly how to deliver client-centered service to these individuals in a holistic and compassionate manner. The product, in its entirety is available in Chapter IV. Chapter II presents the methodology and activities used to develop the scholarly project process.

CHAPTER III

Methodology

Huntington's disease, due to its rare nature, is not a subject that has a large focus in the healthcare industry. The authors of this scholarly project have had similar and differing experiences with observing, treating, and evaluating treatment for those individuals that are affected by Huntington's Disease. The authors came to the conclusion that there was a noticeable gap in supportive care from all healthcare professionals for this population. This resulted in the formation of the focus of the project; to develop a tool for occupational therapists to utilize in practice for treating those affected by HD to increase quality of care, and hopefully quality of life for those affected by HD and their caregivers.

Initially, a broad literature review was conducted to more fully understand the depth of the symptomatology; challenges in healthcare as well as for caregivers and possible multidisciplinary types of care that have increased delivery of healthcare services for those affected by HD and their caregivers. The focus of the literature review also identified evidence-based research on the efficacious treatments and therapies that can be used to preserve the quality of life of the individual as well as family and caregivers. All research was gathered using the databases CINAHL, PubMed, PsycInfo, EbscoHost, and the American Journal of Occupational Therapy Online Database to collect peer-reviewed and evidence-based research to guide this project.

An occupational therapy frame of reference and/or model was explored and employed to guide the development of an educational tool and guide for occupational therapy practitioners.

The primary text by Polatajko, Townsend, and Craik (2007), concerning the Canadian Model of Occupational Performance and Engagement (CMOP-E) was selected to guide this project. A primary reason for choosing the Canadian Model of Occupational Performance and Engagement (CMOP-E) was due to the necessity of addressing spirituality that is needed for a client to stay motivated and engaged while having a disease such as Huntington's disease. Not only is it common for people to experience loss of self-worth, but also direction without self-worth present. Main concepts from the model were interwoven into the development of the guide to make the tool firmly tied to occupational therapy practice, and what is primarily addressed in occupational therapy.

Based on the literature review the guide was developed and formatted. A plan was devised on how to clearly delineate how to address each stage of the disease, and how the occupation-based model of CMOP-E would be incorporated within each stage. Assessments were researched, and a battery was implemented into the product along with intervention strategies for the person, environment and occupation within each stage of the disease. Lastly, resources were researched and listed for occupational therapists to use and to refer caregivers and those affected by HD to for further supports.

CHAPTER IV

Product

The product, *Implications of Huntington's Disease on Daily Living: An Educational Tool and Guide for Occupational Therapists Treating Individuals with Huntington's Disease* is presented in its entirety in following:

CHAPTER V

SUMMARY

The purpose of this scholarly project was to develop and format a guide for occupational therapists treating individuals with Huntington's disease. As discussed prior, Huntington's disease is a neurodegenerative disease that is rare in the United States and all around the world; therefore, occupational therapists are not well educated when treating individuals with Huntington's disease. Although occupational therapists are educated on the more common diagnosis, its the rare diagnosis that make it more difficult to treat.

The guide is organized in seven sections which include: the Canadian model of occupational performance and engagement, assessments, introduction to interventions, early, mid and late stages of Huntington's disease, resources and references. Specific sections have subsections that give introductory information to the assessments, early stage, mid stage and late stages of Huntington's disease. These specific sections are not to be used as a guide but more of a tool when treating those with Huntington's disease. The guide gives the occupational therapist resources and recommendations as well as specific characteristics of each stage and what each stage may look like.

The Guide was designed based upon the Canadian Model of Occupational Performance and Engagement. Using an overarching occupation-based model assisted the authors to establish a client-centered vision when analyzing the person, environment, occupation and spirituality of the individual with Huntington's disease. The main aspect that are important within the model is

that it is a client-centered model in which the client is the agent of change while the therapist enables the process of change through empowerment and enables the skills of the client. The Occupational Therapy Practice Framework also provided the organization of the model in terms of using areas of occupation, the client factors of the person and the elements of environment that can be adapted or modified to help enable and engage the individual with Huntington's disease.

Limitations

The authors did outline a few limitation of this product:

1. Caregiver involvement may be a limitation to implementing this guide if they are not involved in caring for the individual diagnosed with HD. Collaboration with the implementation of care is a key component to using this guide, and can hinder the purpose and/or effectiveness of implementing this guide if there is an absent caregiver role.
2. Knowledge about HD of the healthcare provider can also be a limitation if they do not understand the disease and the symptomology or the clinical presentation of HD. Due to the disease rareness; it is common to see healthcare provider lack of knowledge when treating this population. Therefore, it is the healthcare providers responsibility to educate themselves about the disease and what their role is in providing care to this client population.

Recommendations

From the limitations defined, the researchers made recommendations for future research that would benefit occupational therapy practice and the healthcare arena with treating those affected by Huntington's disease.

1. Expanded research on the implications of occupational therapy on HD treatment would add to the body of knowledge contributing to the positive implications that OT provides for those affected by HD.
2. Research on the impact of multidisciplinary approaches towards treating those affected by HD would also contribute to the increased applicability of successfully treating those diagnosed with HD.
3. Finally, additional research could be conducted on the available treatments for effectively treating those diagnosed with HD in turn decreasing symptomology present with this disease.

Conclusion

Occupational therapy can play a vital role in delivering client-centered service for progressive care and modifications for the individual affected, and supports and resources to the families and caregivers. By implementing a comprehensive program that focuses on the implications of the disease, treatment strategies for all stages and areas affected in the individual's life, and identifying resources that are helpful for the individual and caregivers, quality of care can be dramatically increased for families impacted by HD.

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