Social Skills and Executive Functioning in Children with PCDH-19

by

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ABSTRACT

Social skill impairments and executive dysfunctions caused by epilepsy adversely affect the social, psychological, and cognitive wellbeing of children and their families.

Studies show that children with epilepsy are exposed to social, emotional, academic, personality, and behavioral problems when compared to healthy peers. This study focused on identifying the gaps between social skills and executive functioning among children with PCDH-19.

The researcher relied on the responses from the sampled population to create reliable findings, discussions, conclusions, and recommendations for this project. The study used quantitative design and self-report approach whereby the participants completed survey that was comprised of various rating scales. The study sample consisted of 25 participants. Results demonstrated a close correlation between social skills and executive functions among the children with PCDH-19 epilepsy. SSIS assessment indicated that children exhibited problems in social skills, academic competence, and behaviors. BRIEF-2 rating showed planning, attention, problem-solving, cognitive and emotional problems. The findings exhibited that the significant challenges encountered by girls with PCDH-19 include low levels of social competence which affect decision making in friendships, communicating, and interaction.

Keywords: social skills, executive functioning, PCDH-19, epilepsy, seizures, social assessment, cognitive measurement

Dedication

This has been a 10-year process for me to undertake. In that time, there have been many people that have supported me through the process. I would like to thank my husband Trent, for believing and supporting me from the beginning to the end with this program and for always believing in me. I would like to thank my mom for also supporting me through the process and offering her assistance as needed. I would like to thank my in-laws for also offering support and kid coverage. To Dom and Mary, for their support and for always believing in me. I would like to thank Dr. Watkins, Dr. Caterino and Dr.Woodrich for believing in me and welcoming me into the program. I would like to thank Dr. DiGangi and Dr. Bryce for working with me in the end to make sure I completed my dissertation. I would like to thank Dr. Cohen, Dr. Asher-Drinen and Dr. Sheehan for supporting me during my internship and every day after! Dr. Shadeh- you kindly offered your help and motivation to also get me motivated to finish this project. A big thank you to my cohort- TJ, Erin, Ashley, and Crystal- I couldn't have asked for a better group of people to do this PhD thing with! A big thanks to my Tavan and Papago family for motivating me to continue and finish, especially Courtney and Bronwyn! Thank you to all the PCDH-19 families who contributed to my research. Also a big thank you to Tara Burke with ASU for always answering my questions and helping to get me through the process. And finally, I would like to thank my children, they were and always will be the reason I do anything in life. I love you Gracie, Charlie, Lulu, Landon, and Harrison!

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ACRONYMS

ADHD:	Attention Deficit/Hyper Activity Disorder
AEDs:	Antiepileptic drugs
ASD:	Autism Spectrum Disorder
BASC-2:	The Behavior Assessment System for Children, Second Edition
BASC-3:	The Behavior Assessment System for Children, Third Edition
BRI:	Behavioral Regulation Index
BRIEF-2:	The Behavior Rating Inventory of Executive Function, Second Edition
EEG:	Electroencephalogram
EFMR:	Epilepsy of Females with Associated Mental Retardation
IBE:	International Bureau for Epilepsy
ID:	Intellectual Disability
ILAE:	International League against Epilepsy
MI:	Metacognition Index
MRI:	Brain imaging with magnetic resonance
NES:	Non-epileptic Seizure
PCDH-19:	Protocadherin 19
PCDH19-FE:	Female Limited Epilepsy
PRS:	Parent Rating Scales
SCZ:	Schizophrenic Disorder
SDH:	Structured Developmental History
SOS:	Student Observation
SRP:	Self-Report of Personality

SSIS: The Social Skills Improvement Scale

SSRS: Social Skills Rating System

ToM: Theory of Mind

TRS: Teacher Rating Scales

VNS: Vagal nerve stimulation

VUS: Variant of Unknown Significance

CHAPTER ONE: INTRODUCTION

Introduction

Social skills and executive functioning are vital components of an individual's life and can impact their immediate and long-term quality of life. Impairments and dysfunction in these skills can adversely affect their social, psychological, and cognitive wellbeing. Kiley-Brabeck and Sobin (2006) agree with past studies that prosocial behavioral abilities resulting from social and executive skills lead to academic performance and development of admirable personalities. What do children need the most to develop these admirable personalities? The common social behaviors that assist children the most in their development include: ability to share, helping others, giving compliments, asking for assistance, engaging others in conversations, and solving problems. Self-awareness and consciousness are also listed as vital traits among teenagers for them to develop ideal social skills.

Executive functioning is an advancement of social skills that includes the use of cognitive processes to make decisions. Studies have indicated that the ideal period that these skills start developing is between the ages of 3 and 8 (Alduncin, Huffman, Feldman, and Loe, 2014). The basic processes in executive functioning include identification and manipulation of situations and information to solve problems, plan, organize, and strategize. Executive functioning skills rely on cognitive processes to help children make choices, retain memory, and reach specific goals. The need to understand the role and significance of social and executive functioning skills have led researchers to the identification of various disorders that impact the development of such abilities, such as autism.

PCDH-19 is a genetic epilepsy that is prevalent among young girls and is thought to adversely impact their social and cognitive progression. PCDH-19 is an intractable epilpesy, indicating that treatment and other interventions may not adequately bring the seizures under control. PCDH-19 results from the absence or presence of a non-working protocadherin 19 whose main function is to facilitate brain communication.

The mutation of the PCDH-19 gene leads to permanent modification of the DNA sequence and results in non-functioning and disruption of the protocadherin 19 protein. PCDH-19 is prevalent among girls because the condition involves alteration of the genes in the X-chromosomes (Jacobson, Williford, and Pianta, 2011). While there have been no studies that have investigated PCDH-19 and its effect on the development of social and executive functioning skills, the increasing cases of children diagnosed with the condition at a young age warrants studies that are investigating on how PCDH-19 is affecting other domains.

Background Information

Smith et al. (2018) studied cases of PCDH-19 in the United States to establish the trends and statistics associated with the condition. In the study outcomes supported by Hessen, Alfstad, Torgersen, and Lossius (2018), Smith et al. (2018) report that one in every 10 girls diagnosed with seizures before the age of five develop PCDH-19 epilepsy. Studies do also indicate an overlap of PCDH-19 and Dravet Disorder which exhibits similar symptoms. According to Kolc et al. (2019), over 50,000 people in the United States are living with PCDH-19 epilepsy. Kolc et al. (2019) further indicates that young girls constitute over 80% of this population.

The onset of PCDH-19 is prevalent between the ages of five months and five years when seizures are most common among children. This period is considered the risk age for young girls who may be diagnosed as having PCDH-19. Research suggests that there is a decline of seizure activity as the child moves to adolescence and early adulthood. There are a variety of types of seizures that are attributed to PCDH-19. The most common reported are tonic, generalized tonic-clonic, clonic, atypical absence, complex partial, atonic, and myoclonic (Yeager and Yeager, 2013).

According to Smith et al. (2018), the mutation of the PCDH-19 gene was discovered as the main cause of intellectual disability and epilepsy among children in 2008. The pathogen deviations in PCDH-19 causes medically refractory seizures that lead to common cognitive disorders such as behavioral dysfunction and dysregulation, autism spectrum disorder, and intellectual disability. The increased testing for genetic variation in children with epilepsy, have resulted in broad spectrum relating to the role of PCDH-19 in the onset of epilepsy and other cognitive disorders such as autism.

Problem Statement

The increase in number of children diagnosed with seizures in the last few decades has increased attention to understand the role of the PCDH-19 gene mutilation in causing brain inactivity and abnormality. Moriguchi (2014) considers the rising cases of social skill and executive functioning challenges among children as a significant issue of concern for parents and medical professionals. There are inadequate studies that investigate PCDH-19 and its impact on social skills and executive functioning development among girls. Tse, Hamiwka, Sherman, and Wirrell, (2007) note the increasing cases of misdiagnosis which can affect past research. Non-epileptic seizures (NES) that have no adverse impacts on the

social or cognitive development of children have been treated as cognitive disorders that result in deficiencies in executive functioning and slow social skill development. Until recently, video-EEG was the only tool available for diagnostic of epileptic seizures with only a small number of families being able to afford this option.

The separation of epileptic and non-epileptic seizures including the contribution of PCDH-19 continues to impact the understanding of the actual causes of deficiencies in social skills and executive functioning. Therefore, there is a need to identify and evaluate affordable screening tools that can separate the NES from epileptic seizures to inform correct interventions. The impacts of misdiagnosis have led to treatment of epilepsy conditions leading to adverse effects of the neural, behavioral, and cognitive development (Shorvon, 2010).

The existing gaps in understanding the effects of PCDH-19 on social skills development and executive functioning of children stems from variables in key aspects in identification and management of the condition. The inadequate and inconclusive investigation in this area indicate that there are still no standard procedures for diagnosis and treatment of cognitive and psychological disorders emanating from mutations of the PCDH-19 gene.

There is also a need to investigate the separation of NES and PCDH-19 to avoid misdiagnosis. Establishing better screening tools that identify differences in levels of social development and cognition is significant in ensuring that the causes of delays in social and cognitive development are accurately diagnosed. Determining the appropriate diagnostic tools and procedures will help determine the social skills and executive functioning levels and planning appropriate interventions.

The analysis of the impacts of behavioral, social, and mental development deficiencies is a critical component in establishing how PCDH-19 affects the social skills and executive functioning of children who have been diagnosed as having PCDH-19. This dissertation will investigate the impacts of PCDH-19 on the social skills and executive functioning among children who have been diagnosed as having PCDH-19. The study intends to fill gaps in the past studies through use of primary data to assess the prevalence of social and cognitive dysfunction among girls diagnosed with PCDH-19 epilepsy.

Purpose of the Study

The drive of many studies that seek to identify gaps in social skills and executive functioning among children include gaining new information of the causes and effective intervention methods. The increase in the number of cases among children and adults who have been diagnosed has led to the rise in the number of researchers delving into research to understand the influence of PCDH-19. Until recently, such studies that looked at pediatric patients with epilepsy had been minimal considering the unavailability of data relating to social, cognitive, and psychological development.

The identification of children as the most affected population by PCDH-19 has renewed interest among researchers and therapists on the causes, symptoms, risk factors, and treatment options. Levan (2015) conducted a study that looked at the social skills and executive functioning in children with epileptic and non-epileptic seizures. Levan hoped to predict diagnosis based on social skill measures using the BASC-2 and SSIS. However, she was not able to show social skills as a successful predictor of epilepsy or NES diagnosis.

Since the completion of this investigation, there has been no other extensive investigations to examine the social skills and executive functioning in the PCDH-19 population. Therefore, this study is necessary to contribute to the overall research on PCDH-19, including impacts to social skills and executive functioning, especially considering that more cases are being identified each year. Importantly, this project will establish the relationship between PCDH-19 and social skill development and executive functioning. The focus on children will help in filling literature gap considering that available study outcomes have mainly focused on the effects of epilepsy on the social and execution functioning among adults. This study will follow the process that Levan (2015) used, however, it will only be looking at children who are diagnosed with PCDH-19.

Research Objectives

The completion of this project will rely on the following objectives that will facilitate development of research questions and aid in the data collection process.

1. To determine impairments and dysfunction of social skills of children diagnosed with PCDH-19
2. To determine impairments and dysfunction of executive functioning of children who are diagnosed with PCDH-19

Research questions and hypothesis

1. Do children diagnosed with PCDH-19 have demonstrated impairment in social skills?
    a. Hypothesis- Children diagnosed with PCDH-19 will demonstrate deficits in their social skills.

2. Do children diagnosed with PCDH-19 have demonstrated impairment in their executive functioning?
    a. Hypothesis- Children diagnosed with PCDH-19 will demonstrate deficits in their executive functioning.

Definition of Terms

PCDH-19 (protocadherin 19): PCDH-19 is a vital gene in X-chromosomes which makes a protein that facilitates brain communication. The interruptions of this this gene leads to inefficiency in the work of protocadherin 19 protein

PCDH-19: An intractable epilepsy characterized by delays in cognitive and sensory functions, onset seizures, and problems in behavior. Results from the alteration of PCDH-19 in X-chromosomes

Executive Functioning: refers to a set of mental or cognitive processes that facilitate development and control of behaviors. Executive functioning aids in planning, paying attention, organizing, regulation of emotions, self-monitoring, appreciating diverse views, and self-awareness.

Social skills: Include abilities to interact and communicate with others verbally or non-verbally. Appropriate social skills should enlist positive evaluation and responses from other people.

CHAPTER 2: LITERATURE REVIEW

Introduction

Epilepsy is more than just seizures, often it is comorbid with social and psychological problems such as anxiety, autism, and stress among other related psychological syndromes. The challenges in social skill development and capabilities in executive functioning stem from diverse developmental deficiencies (Jones et al. 2014). Available literature identifies the ages of three and five as the age for development of psychological and social disorders that can adversely affect personality, emotional, social, and cognitive growth and expression.

Children suffering from epilepsy are believed to experience more social, emotional, academic, personality, and behavioral problems compared to healthy peers. (Jones et al. 2014). Many studies have demonstrated the prevalence of deficiencies in social skills and social competence is common among individuals diagnosed with epilepsy. The increase in chronic seizures has been a motive among research to study the changes in social and cognitive growth.

The similarity of different studies is that adults suffering from epilepsy demonstrate weaker social skills and slower cognitive development. Additionally, behavioral problems are also prevalent among children who are regularly affected by epilepsy. Holley et al. (2014) also noted the increasing cases of mental health problems among children living with epileptic conditions. The literature shows that the treatment of specific social and mental disorders improves the personality, interaction, and cognitive functioning of epilepsy patients. However, absence of treatment for the disorders can lead to permanent social and psychological disabilities including cognitive delay and brain dysfunction.

According to Schaffer et al. (2017), research exploring the issue of social skills and executive functioning of children diagnosed with epilepsy is gaining momentum in academic research. In the past, it appeared that researchers were only concerned with the social skills and executive functioning of adults with epilepsy. Hamiwka, Hamiwka, Sherman, and Wirrell (2011) researched social skills of children with epilepsy and those of healthy and chronic disease controls. Their results found that children with epilepsy were less cooperative, less assertive and displayed less self-control.

The researchers also concluded that having a chronic disease may have played a role in the social functioning of the children with epilepsy. It has also been discussed that adults, whose seizures started in childhood, will often have poor long-term social outcomes than adults whose seizures started after childhood. Yet, there is no research that looks at why seizures that started in childhood have much more effects on the social outcomes for those affected.

With the studies being limited on the social skills and executive functioning of children with epilepsy, research that focuses on specific type of epilepsies is almost non-existent now. The focus of this research is to add to the foundation of knowledge regarding social skills and executive functioning of children diagnosed as having Protocaderin 19 (PCDH-19). In the following sections, a comprehensive summary of epilepsy is presented, along with an introduction to PCDH-19, a genetic form of epilepsy that predominantly affects girls. A review of the literature on social impairments of children with epilepsy is also included. The theoretical and conceptual frameworks as foundations for this inquiry are also presented.

PCDH-19

Protocadherin 19 (PCDH19) was formerly known as Epilepsy of Females with Associated Mental Retardation (EFMR). PCDH19 is characterized by mutations in the PCDH19 gene which is located at Xq22.1 (Cappelletti et al., 2014) and is believed to primarily affect females although there have been a small number of males identified as being affected and not just carriers of the mutation. The mutation can be genetic, meaning that it is inherited from an unaffected father or an affected mother, or it can be considered de novo, meaning that the mutation occurred spontaneously in utero and there is no genetic link. The increased availability of genetic testing is helping to identify the mutation that causes PCDH-19 in children and adults.

With the increased ability to diagnose PCDH-19, there are now a growing number of newly identified cases of PCDH-19. Due to the increased prevalence, research has grown exponentially in the last five years on PCDH-19, yet is still very limited in nature and is still limited in the number of experimental studies. PCDH-19 has garnered much interest due to its unique expression pattern since the phenotype is almost solely restricted to females (Cappelletti et al., 2014). PCDH-19 can cause epilepsy that varies in severity. Seizures for those diagnosed typically begin before 12 months of age and often occur in a cluster (Dibbens et al., 2008).

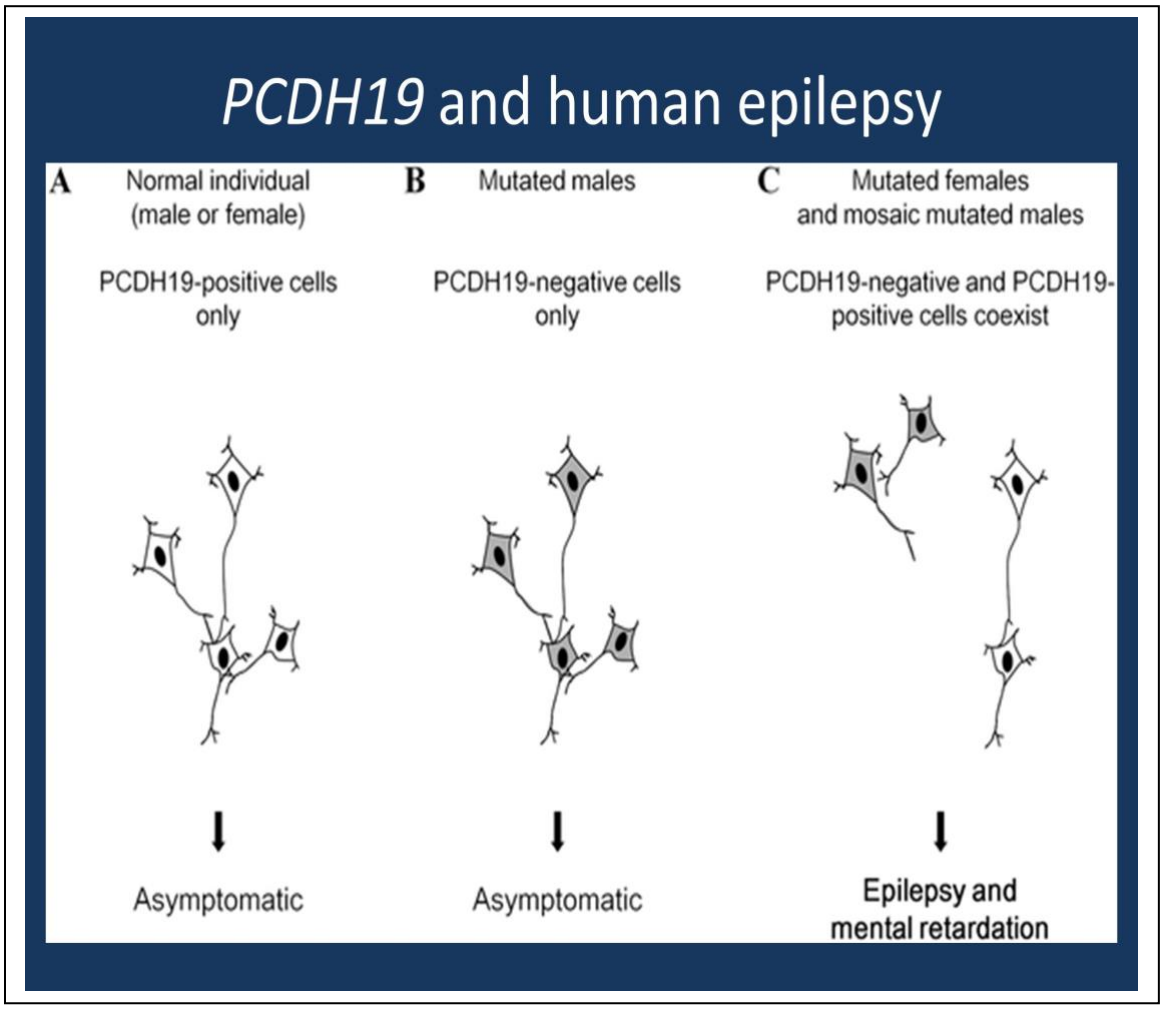
For some of the affected, the seizures can be triggered by illness such as fever. Others have reported that seizures were triggered by teething and others experience no observable trigger. The presentation of seizures varies from child to child and can change in presentation over time. It can be accompanied by intellectual delay, autism, autistic characteristics, anxiety, and other psychological impairments. Most diagnosed patients had

normal to borderline cognitive function before the seizures occurred but, suffered delays post seizure onset (Schefer, 2008). It is important to note that there is a subset of affected children who have limited to no cognitive delay even after the onset of seizures.

Breuillard et al. (2016) examined autism spectrum disorder phenotype and intellectual disability in females with PCDH-19. Their findings showed a high proportion of autism spectrum disorder (ASD) and intellectual disability as well as adaptive and social disturbance. They also noted that the literature from 2008 to 2013 indicated that 46 percent of the 125 students had some autistic features or behavioral disturbances and 78% of the 125 patients exhibited intellectual disability. It is important to note that the ASD profile the researchers found showed a specific phenotype where communication and social interaction domains were the most affected. Their results indicated more of a theory of mind deficit which they noted could add difficulty in establishing an ASD diagnosis based on clinical assessment and not utilizing standardized assessment.

Bassani et al. (2018) categorizes PCDH-19 as a syndromic, functional and rare single gene mutation genetic component, and that this can facilitate the severity of Autism, Intellectual Delay, Epilepsy, ADHD, and Schizophrenia among other psychological disorders. The relevance of PCDH-19 to autism, anxiety, and schizophrenia stems from rare mutations that have directly impacted the brain functioning and social skill development. The molecular function of PCDH-19 includes a calcium cell-adhesion protein expressed in the brain. The prevalence of PCDH-19 among females has led researchers to conduct experiments on blood levels and components of allopregnanolone. However, there are no conclusive study reports that point to low levels of allopregnanolone and pregnenolone as risk factors for PCDH-19.

Figure 2.1 Representation of PCDH19 Mutilation



PCDH-19 was initially thought by many to be rare in the population. However, with the availability of new genetic testing, diagnostic screenings have shown that the PCDH-19 mutation is being found more frequently than what was initially believed. There has not yet been a large epidemiologic study on PCDH-19 to determine incidence nor has there been a study published at the time of this writing looking specifically at the social skills and executive functioning of children diagnosed with PCDH-19. In their study on female epilepsy, Bassani et al. (2018) concluded that the mutation of PCDH-19 on the Xp22.1

chromosome causes female limited epilepsy that leads to intellectual and social challenges among children and adults.

Genetic Prevalence of PCDH-19

The prevalence of female limited epilepsy with or without autism or intellectual delay is attributed to genetic causes (Tan et al 2015). Until the identification of the PCDH-19 mutation as the cause of female limited epilepsy (PCDH19-FE), genetic mapping and sequencing had been done for years to determine the specific genes and chromosomes susceptible to epileptic seizures among women. The study by Tan et al. (2015) includes the consideration of seizure frequency, signs of autism, clinical aspects of schizophrenia, depression, and obsessive compulsive disorder. The study included two clusters of participants: an older group comprised of six females who have previously suffered from female limited epilepsy in their childhood but had not experienced seizures in their adult life. The second cohort included a young population with an average of eight years who experienced seizures.

The investigation also included identification of gene transmitting males and females to identify the genetic expression and sequencing. The study concluded that mutation in PCDH-19 is significant in causing epilepsy among females. The inheritance of PCDH19-FE by the X-chromosome including alter gene expression thus leading to the prevalence of seizures among women.

Epilepsy has often been referred to as a disorder or a family of disorders and not a disease (Fisher et al., 2014). This was due to the notion that epilepsy was comprised of not just one disease or condition. However, this may lead to misconceptions by the public. Fisher and colleagues (2014) note that disorders are often thought to not be permanent and

implies a "functional disturbance". They clarify this misconception by comparing epilepsy to cancer and diabetes; stating that these disorders, while they comprise numerous sub-disorders, are still considered diseases. The International League against Epilepsy (ILAE) and the International Bureau for Epilepsy (IBE) have recently presented that epilepsy is best considered a disease (Fisher et al., 2014). Therefore, for the remainder of this paper, epilepsy will be referred to as a disease.

Epilepsy is a disease that is multifaceted. It can affect those afflicted in a multitude of ways, including: their health, cognition, behavior, academic performance, psychological functioning, and their social functioning (Hamiwka, Hamiwka, Sherman, & Wirrell, 2011). According to the Institute of Medicine of the National Academies (2012) epilepsy is the fourth most common neurological disease in the United States. They also report that 1 in 26 people will develop epilepsy at some point in their lives. It is estimated that 2.2 million Americans have epilepsy and that each year there are 150,000 newly diagnosed cases in the United States.

The National Society of Epilepsy (2016) report that the onset of epilepsy is most common in early childhood and in later life, thus children and seniors are most at risk for developing epilepsy. Epilepsy is the most common neurological disease in children (Elliott, Lach, & Smith, 2005). The Centers for Disease Control and Prevention (2016) reports that 1% of children aged 0 to 17 years have had a diagnosis of a seizure disorder or epilepsy. It is the most frequent chronic neurologic condition in childhood, yet there is limited research that focuses on how epilepsy could affect social skills and/or executive functioning in the pediatric epilepsy population.

McHugh and Delanty (2008) explored the general prevalence of epilepsy among genders without consideration of social or cognitive effects. The authors found a pattern relating the gender differences in the epileptic cases reported in health care institutions. However, males report more cases of general epileptic cases compared to females. The trend has been attributed to the exposure to risk factors. That notwithstanding, the idiopathic epileptic seizures have prevalence among females representing over 20% of all reported cases in early 2000s (McHugh and Delanty, 2008). Epilepsy cases resulting from genetic mutations are more common among young women compared to older females and males.

Seizure Classification

Staley (2015) adds that the research on the causes of epilepsy point to the diversity of seizures that trigger neurological dysfunction. However, the understanding of seizures has provided new information on the specific brain attacks that cause epilepsy and eventual challenges in social skills development and executive functioning.

The International League against Epilepsy (ILAE) state that seizures are a transient occurrence of signs and symptoms due to abnormal or synchronous neuronal activity in the brain. Seizures are caused by an abnormal excitability of neurons, which are often regulated by ion pumps or control gated ion channels. The presence of seizures can be noted by observable behavior and/or an electroencephalogram, commonly known as EEG. A seizure can be behaviorally defined as an abrupt unconscious change in behavior, movement, autonomic function or sensation. There are many known causes for seizures, including: genetic mutations, medications, head trauma, strokes, infections, brain malformations, and other diseases. There are also cases where the cause is simply unknown. It is imperative to

note that the occurrence of a seizure does not necessarily mean a clinical diagnosis of epilepsy.

According to Scharfman (2007), the effective method to gain insights into the causes and prevalence of epilepsy is by understanding the classification of seizures and their recurrence among the susceptible populations. In her study on the neurology of epilepsy, Scharfman (2007) indicates that it's complex to understand the components and severity of seizures for individuals already affected by epilepsy compared to people in normal health. Scharfman (2007) defines seizures as abnormal periods of neural lapses thus needs to partial or total cognitive and behavioral blackout.

Most studies agree that seizures can last a few seconds, some minutes or can be long and continuous especially with status epilepticus. While seizures are characterized by physical dysfunctions including fainting, the clinical manifestations vary and may be devoid of any convulsions or muscular contractions. The confusion that exists in the relating epilepsy and seizures is that all seizures result in epileptic conditions. However, Scharfman (2007) clarifies that epilepsy is a state of spontaneous and recurrent seizures. Provoked seizures that only occur once do not point to an epileptic condition or neural disorder.

Social Skills

Social skills or social competence as often referred to in literature can broadly be defined as the ability to function age-appropriately to social contexts and demands (Masten & Coatsworth, 1998). Social skills are learned behaviors that enable a person to interact with others effectively and avoid socially unacceptable responses (Tse, Hamiwka, Sherman, and Wirrell, 2007). The presence of social impairments can lead to other

outcomes such as poor academic achievement, peer rejection, depression, and other forms of psychopathology (Bellini, Peters, Benner, and Hope, 2007). The most important outcome of having social impairments is the effect it has on establishing meaningful social relationships (Bellini et al. 2007).

Social skills are critical to successful cognitive, emotional and social development. Acquiring and developing social skills is considered one of the most important goals in childhood (Tse et al., 2007). If there are problems in acquiring appropriate social skills during the formative years, there could be lasting effects on psychosocial adjustment that last into adulthood. Children with chronic conditions such as epilepsy have been found to have an increased risk for poorer social competence. There are concerns regarding difficulties in social functioning reported in children with epilepsy and there is an increased prevalence of poor social competence in children with epilepsy from preschool to adolescence (Hamiwka, Hamiwka, Sherman, and Wirrell, 2011).

Rantanen et al. (2009) conducted a study that looked at the social competence of preschool children with epilepsy. Their findings showed children with complicated epilepsies had fewer age appropriate social skills as well as more behavior and attention problems than non-epileptic children. They also found that children with non-complicated epilepsies were within the normal range. Their results implicate that when a child is experiencing complicated epilepsy, there are more deficits in behaviors and attention.

When the social skills of 101 children and teens (aged 3 to 17 years old) that had epilepsy were compared with the social skills of their nearest-aged sibling who did not have epilepsy, results showed that the children and teens with epilepsy had poorer social skills and were less assertive than their siblings. The children with epilepsy had

significantly poorer ratings on the Social Skills Rating assessment. Results from this study showed that the may be a relationship between social function and internalizing behaviors for those that have epilepsy. The researchers also noted that the presence of a learning disability and abnormal family functioning were significant predictors of impairment in social skills (Tse et al., 2007).

Lew et al. (2015) examined whether children with generalized and focal epilepsy within a community sample from mainstream education, showed deficits in measures of social cognition. They administered the Strange Stories task and the Mind in the Eyes task as well as an IQ assessment to the study participants and comparison participants. The Strange Stories task consisted of 16 short passages followed by a test question. Eight of the stories were constructed to measure the ability to infer mental states from scenarios involving lies, double bluff, white lies, persuasion, an appearance/reality distinction, and misunderstanding (Lew et al., 2015).

The remaining eight stories were non-mental control stories measured the understanding of physical events (Lew et al., 2015). The Mind in the Eyes task comprises 28 photographs of the eye region of male and female adults. The child was to choose from four words or short phrases that are provided to assess the child's ability to attribute cognitive or affective mental states. Their results found that both groups of children with epilepsy performed worse than the control children on the Mental Stories component of the Strange stories task. They discussed that the performance on Mental Stories was related to pragmatic communication for the generalized epilepsy group. There were no differences between the epilepsy groups or control group in the Mind in the Eyes task.

Another study by Byars et al (2014) establishes that individuals suffering from epilepsy have increased propensity to different social functioning lapses. The researchers completed a study of 193 children diagnosed and living with epilepsy for at least 36 months. The objectives of the experiment were to understand the association between social functioning and language development. The outcomes of the study revealed that children with recurring and persistent epileptic seizures developed poor language and social functioning skills. The poor language and social integration was evident from the onset of seizures compared to the sibling with no recurrent seizures. The inadequate social and language skills were evident from the first of months of diagnosis to the last period of the experiment. The authors considered the poor language development as the main cause of inadequate social skill functioning among epileptic girls.

Further, Stafstrom and Carmant (2015) point out that social skill challenges includes different aspects that relate to social competences, interactions, and relationships. The inability to establish and sustain social relationships and interactions at the young age is attributed to the different psychological and cognitive dysfunction While other psychological and mental disorders have been attributed to the rise in the low development of social skills among girls, the increase in clinical diagnosis shows that the cases of social dysfunction rise in relation to the number of epileptic seizures (Stafstrom and Carmant, 2015).

Children with epilepsy are more likely to develop emotional, social, behavioral, and mental problems compared to their peers. The significant challenges that face girls with epilepsy is the low levels of social competence which affect decision making in friendships, communicating, and interaction. As adults, the individuals will endure

challenges in seeking and settling in jobs, sustaining relationships, and managing marriages. Individuals with frequent seizures may develop low self-esteem and self-confidence which are essential components in perfecting social and emotional development (Byars et al. 2014).

Lugo, Swann, and Anderson (2014) carried out a study on the early onset of seizures and its resulting deficits in social behaviors and learning. The outcomes of the investigation showed that children diagnosed with epilepsy and unprovoked seizures had a high co-morbidity with psychological syndromes such as autism. The study concluded that the presence of psychological disorders among children was increased by the prevalence of unprovoked and recurrent seizures (Lugo, Swann, and Anderson, 2014).

The conclusion from the selected studies shows that the development of social skills is very critical in childhood growth and development. Social skills facilitate the acquisition of specific traits such as responsibility, emotional control, personality, assertion, and compassion. Children living with epilepsy and other disorders that affect their cognitive functioning may find it challenging to navigate through normal social situations due to limited physical activity and inability to make social choices. Lugo, Swann, and Anderson (2014) also found out that the recurrence of seizures contributes to isolation of children with epilepsy when parents fear the occurrence of seizures during playtime or other social activities.

Stafstrom and Carmant (2015) assert that children with epilepsy have the same levels of social skills and capabilities compared to healthier kids. However, the condition leads to the slow mastery of social skills due to onset of learning challenges and other related factors such as cognitive and psychological disorders. Before the age of eight, girls

struggle to develop adequate social skills when the recurrence of seizures is at its peak. Byars et al. (2014) adds that the earlier epilepsy affects children, the higher the severity of adverse impacts on attainment of social skill milestones. The eventual impact is the lack of essential skills that affects engagement in routine activities that define the physical, emotional, psychological, and mental growth.

From the review of literature, there appears to be deficiencies in the social skills of children who have epilepsy. What remains unclear is the cause or the extent to what is contributing to the deficiencies. However, the early recognition and identification of the social difficulties that may accompany epilepsy, is critical for appropriate intervention (Byars et al. 2014) to improve outcomes for the children as they age into adulthood. This is important for children who are diagnosed with PCDH-19, as parents have noted concerns about their social skills and there is no current literature to support these concerns.

Social Communication

Language is the main component of communication, verbally and nonverbally. Verbal language is also the underlying foundation for much of emotional development. Social communication can be impaired when there is impaired use of language that is used to organize and process thoughts or responses (Caplan et al., 2002). Language disability in children is known to be comorbid or develop due to conditions that involve the central nervous system such as epilepsy (Parkinson, 2006). There have been studies that have found evidence of formal thought disorder in patients who have complex partial seizure disorder. Children acquire the ability to organize their thinking and provide the listener with the necessary links to follow who and what they are talking about from the toddler period to adulthood (Caplan et al. 2002).

The onset of epilepsy and repeated uncontrolled seizures during the acquisition period could impact the development of those skills. Children with focal epilepsy demonstrate deficits in various aspects of communication, including pragmatics (Lew et al., 2006). Pragmatics refers to the social language skills that are used daily in interaction with others. This can include what is said, how something is said, and even body language. Individuals who have poor pragmatic skills can often misinterpret others and can have difficulty responding appropriately to others. Some examples of pragmatic skills include asking questions, eye contact, using humor, turn taking, and using appropriate strategies for gaining attention.

Deficits in pragmatic language are present in a range of psychiatric disorders such as autism and attention deficit hyperactivity disorder (Broders, Geurts, and Jennekens-Schinkel, 2010). These deficits of pragmatics have been suggested to be features of many children with epilepsy. There have been studies that have looked at the relationship between pragmatics and epilepsy, yet many of the results have been inconsistent. Broders, Geurts, and Jennekens-Schinkel (2010) denoted four reasons that could explain the inconsistencies those four reasons are the confounding effect of comorbidity, general intellectual functioning, research tools adopted do not always capture pragmatics and age.

The researchers also explored pragmatic language deficits in children with epilepsy and found that children with epilepsy had more pragmatic difficulty than typically developing age-matched controls. Others examined the relationship between language ability and social functioning and found that children who had persistent seizures had poorer language function, even at the onset of their seizures (Byars et al., 2014). These findings brought into question the effects of language function and its role in the social

competence. Children who are diagnosed with PCDH-19 are believed to have deficits in their language impairment which could be affecting their social skills. However, to date there are no studies that have looked at the correlation.

Executive Functioning

Executive functioning is the term used to describe the set of behaviors that individuals need to complete tasks and solve problems. Executive function is believed to be the overseer of other cognitive functions such as memory and attention. Having deficits with executive functioning can impact daily life and impact how they interact with others, thus affecting their social skills. Children with epilepsy are at a higher risk of experiencing neurocognitive deficits, which are thought to be related to executive functioning (Holley, Whitney, Kirkham, Freeman, Nelson, Whitlingum, & Hill, 2014).

Chan, Smith, Greenberg, Hong, and Mailick (2017) assert that executive functioning is a collection of behaviors that enable individuals to achieve their goals and adapt to the dynamic environment. Neuropsychologists and neurologists prefer the use of executive function terms to refer to cognitive and mental capability that define social behavior and decision making (Chan et al. 2017). The term executive refers to the lead and supervisory role over other cognitive functions such as perception, memory, and attention.

The normal executive functioning among young people include the ability to organize and dictate the performance of all cognitive processes towards good decision making, behavior, awareness, and relations. Normal executive functioning represents the highest level of mental and cognitive activities that define the capabilities to make judgements, sustain consciousness, and free will. The connection of executive functioning

and epilepsy is explored through establishing the differences in performance and planning abilities between healthy and epileptic subjects.

Miller, Loya, and Hinshaw (2013) point out that individuals suffering from genetic-motivated seizures show deficiencies in their executive functions and stated that psychological, medical, and biological factors also contribute to the impairment in executive functioning. Notably, most cognitive and brain disorders result in deficiencies in cognitive functioning leading to effects on executive reasoning. Therefore, levels of executive functioning vary according to the underlying cases and the presence of brain and psychological disorders.

Nixon (2013) carried out a study to identify the external factors that contribute to the deficiency in executive functioning. The inquiry identified alcohol and drug abuse as risk factors to the impairments in cognitive functioning. However, permanent impacts in brain activities have been linked to the genetic mutations of brain cells or the prevalence of cognitive disorders. The genetic mutation of PCDH-19 directly impacts the brain system thus posing adverse effects on cognitive processes. Miller, Loya, and Hinshaw (2013) indicate that seizures arising from the brain system are more severe on the executive functions compared to unprovoked seizures arising from other body systems.

Albert, Abu-Ramadan, Kates, Fremont, and Antshel (2018) provide a different understanding of executive functioning through their definition that mentions mental skill weaknesses that impair learning processes. In their understanding of executive functioning, the authors identify different issues such as problems in flexible thinking, development of working memory, and self-control. Albert et al. (2018) consider executive functioning as the master of the brain because it allows planning, setting goals, and making decisions.

Children struggling with cognitive problems resulting from poor executive functioning report adversity in daily life activities including learning and socialization.

The increasing cases of attention skills and the prevalence of ADHD disorders contribute to the development of challenges in executive functioning. The presence of learning disabilities leads to weaknesses in executive skills. Elosúa, Del Olmo, and Contreras (2017) note that proper executive functioning skills are responsible for different skills such as sustaining attentive capabilities, planning, organizing, concentration, identifying different points of view, and regulating emotions.

Futher, Elosúa, Del Olmo, and Contreras (2017) assert that the development of executive skills occurs mainly between childhood and early adulthood. The differences in attainment of milestones in social and mental development may indicate presence of impairment in executive skill development. The presence of seizures may further indicate brain dysfunction and disorders that create barriers in optimal development of cognitive abilities.

Yamamoto and Imai-Matsumura (2019) completed a recent study in Japan to understand the gender differences in behavioral regulation and executive skill development among 5-year olds. The study assessed the working memory, self-control, and behavioral competence. The assessments were conducted inside classroom, playing environment, and at homes. The studies show minimal gender differences in all aspects measured in the experiment. However, girls showed fewer issues relating to behavioral, working memory, and attention issues. The authors concluded that many studies that seek to understand executive functioning are gender biased (Yamamoto and Imai-Matsumura, 2019).

Some research has found that often in new-onset cases of pediatric epilepsy, impaired executive functions are often observed even prior to the introduction of medication. It was also noted that children who have new-onset epilepsy demonstrated less impaired executive functions than children who have chronic epilepsies (Reuner, Kadish, Doering, Balke, & Schubert-Bast, 2016). Some research has noted that issues with executive functioning can be associated with issues with social functioning and affect peer interactions. To date there is no research that looks at executive functioning of children diagnosed with PCDH-19.

Diagnosis and Treatment

The diagnosis and treatment of epileptic seizures was not specialized until the identification of different causes of seizures. Brain imaging and scanning through MRI scans have been the traditional diagnostic tool for identifying the onset of provoked and unprovoked seizures. Electroencephalography or EEG includes testing that support MRI scans to provide comprehensive brain activity reports including the stability of the central nervous system. EEG diagnoses the development of cognitive and other brain functions and can indicate gaps or non-existence of mental abilities (Rinsky and Hinshaw, 2011).

The techniques also detect any discharges in upper part of the brain particularly the frontal and temporal lobes. The diagnosis of PCDH-19 relies on genetic testing methods that use blood or saliva samples in addition to medical history, EEG, physical examination, and family history.

Genetic testing includes extracting DNA from the samples and then analyzing it for gene mutations. At this level, testing is significant to inform selection of treatment method and therapy. Recommendation for genetic testing is ideal for children who experience

recurrent seizures. Early diagnosis is important to help patients and their families start immediate therapies and treatment.

A positive result from genetic testing means that the source of gene mutation is known. A negative outcome would be that no condition is identified as a cause to seizures at that time. The result does not necessarily point to absence of genetic epilepsy. Additional testing including large blood samples are used to rule out the presence of already identified genetic mutations. Other results may be inconclusive due to abnormalities in the DNA thus an inability to establish any genetic relation to the seizures. The outcomes are usually resolved through including other family members in the diagnosis to confirm the presence of variant of unknown significance (VUS).

The initial treatment for PCDH-19 constitutes direct interventions to prevent and stop seizures. Treatment is a balance between risk and benefit (Shorvon, 2010). Schachter, Shafer, & Sirven (2013) report that about 60% of people with epilepsy respond well to the first one or two anti-epileptic drugs used. They go on to note that if the first two drugs were unsuccessful, only 3% will have results if other drugs are tried. Medications are chosen based on seizure classification, type of epilepsy and the clinical presentations of the seizures. Although the aim of medication intervention is complete seizure control, this only occurs in some of the population, as stated above. Others suffer from epilepsies that are termed intractable or medically resistant.

Williams and Sharp (2000) state that 20% to 30% of children suffer from intractable or medically resistant epilepsy. As stated prior, PCDH-19 is an intractable epilepsy. Loring & Meador (2001) discuss that the side effects of medication may adversely affect a patient's quality of life. They discuss that antiepileptic drugs (AEDs) are designed to

reduce neuronal irritability and are often the first choice as treatment for new-onset epilepsy. Some of the most common AEDs include Carbamezephine (Tegretol), Keppra, Topiramate (Topamax), Lamotrigine (Lamictal), Phenobarbital, and Valproic Acid (Depakote). AEDs work on abnormal brain activity but also affect normal brain activity and this can affect a patient's cognitive function.

The neuropsychological functions that are most likely to be affected by AEDs are psychomotor speed, vigilance, memory, and mood (Loring & Meador, 2001). Social skills and executive functioning could theoretically be impacted by many of the AEDs. Children may be more at risk for experiencing cognitive effects of AEDs because of the effect on attention and memory over extended time. Other side effects of AEDs include loss of bone density, language impairment, drowsiness, tremors, weight loss or gain (Williams & Sharp, 2000). It is important to note that side effects can vary drastically from medication, dosing and individual.

Surgery can be an option when medical therapy has not been effective and the epilepsy is focal (Loring & Meador, 2001). There are several different types of surgeries that may be performed on the brain to alleviate seizures. There are several options to consider when determining surgical treatment and often neuropsychology evaluations are completed prior to get baseline information on neurological functioning. Fernandez, Gedela, Tamber, and Sogawa (2015) discuss that often surgery is limited in children with intractable epilepsy and that VNS has been used as an adjunctive therapy. VNS is a device that is sometimes referred to as a "pacemaker for the brain" (Schachter, Shafer, & Sirven, 2013). It intermittently applies electrical currents to the vagus nerve to prevent seizures (Loring & Meador, 2001). It is placed under the skin of the chest wall and has a wire that runs from

the device to the vagus nerve in the neck. The vagus nerve is part of the autonomic nervous system, which is responsible for controlling functions such as heart rate and other functions that are involuntary (Schachter et al., 2013).

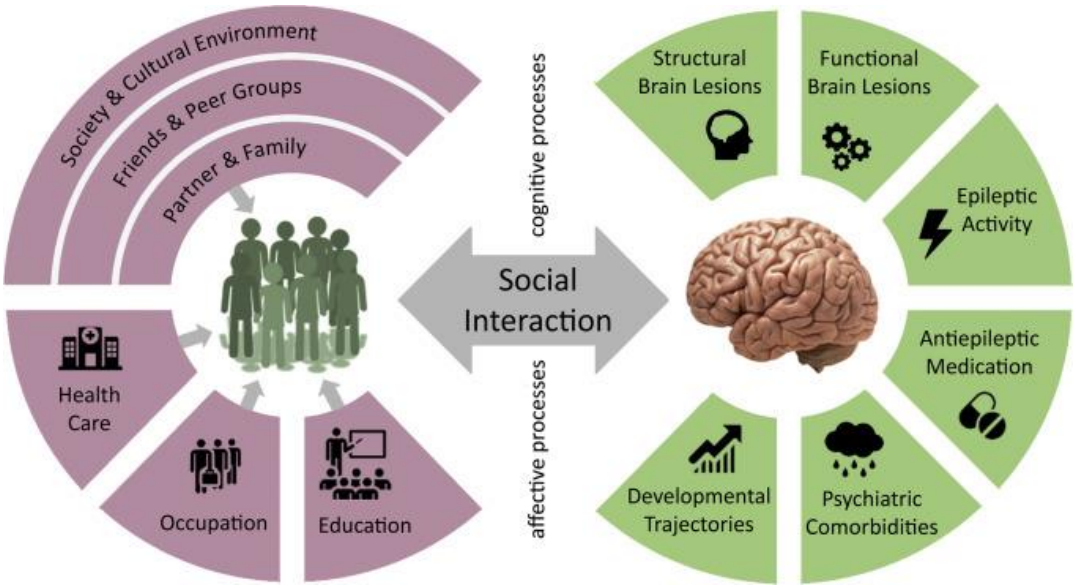
The treatment for PCDH-19 historically has relied on prescription drugs and VNS (Shorvon, 2010). The commonality of epileptic medicine stems from affordability and the least amount of side effects. The use of medicines is ideal due to its gradual use until the correct levels that enable control of seizures. The use of epileptic medicine varies on gender and age of the child, type and frequency of seizures, prevalence of psychological syndromes, and presence of other health and medical conditions that can influence the effectiveness of the medicine.

The other common method is the use of vagus nerve stimulation through therapy sessions where electrical devices are planted under chest skin to enable sending impulses to the brain. The devices reduce the frequency of seizures including combating their severity. The use of ketogenic diets is also getting popularity as a treatment for intractable epilepsy among children. The intervention includes restricting patients to meals that are high in facts and lows in carbohydrates. However, ketogenic is considered when the other methods are not responsive in stopping or reducing unprovoked seizures among children. Recently, some families have begun the use of medical marijuana to help control seizures and have noted a potential decrease of seizures and an increase of cognitive processes and a decrease in maladaptive behaviors.

Theoretical Framework

Different theories have been used to explain the relationship between PCDH-19 epilepsy and social skills and executive functioning. Steiger and Jokeit (2017) uses theory of mind (ToM) to show the influences of epilepsy on social and cognitive functioning. According to this theory, individuals diagnosed with epilepsy or seizures that eventually lead to epileptic conditions are exposed to risks of low social competencies and general decline quality of life. The deficits in social and cognitive development impaired social interactions and individual performance. The foundation of ToM is that social cognitive functions correlate in achieving balanced mental stability and desired behavior.

Figure 2.2: Representation of ToM



Source: Steiger and Jokeit (2017)

ToM explains the connection of social competence and executive functioning through understanding beliefs, emotions, desires and knowledge. Social interaction is considered the key aspect in establishing the social skill development and presence of a

stable mental state that can support executive functioning. The presence of the epileptic condition affects the social interaction through impacting the cognitive processes and affective functioning. Psychological disorders also affect the balance between social and mental processes leading to impacts on the quality of life.

CHAPTER 3: METHODOLOGY

Introduction

Collecting data to identify the relationship among PCDH-19, social skills, and executive functioning requires use of specific research approaches and design. The researcher will exclusively rely on the responses from the sampled population to create reliable findings, discussions, conclusions, and recommendations for this project. While the study requires quantitative data through collection of different measurements from the respondents, the self-report approach is considered appropriate for this investigation. In self-report studies, the participant's complete questionnaires, surveys, and forms without any interference of the researcher. The approach is recommended for studies that seek responses in experiments and observational inquiries.

Self-reports are extensively used in psychology to help therapists collect the symptoms of various disorders to develop appropriate interventions. Use of highly structured questionnaires is ideal for psychological inquiries due to its strength in meeting large number of participants. However, self-report studies have persistent validity issues due to high chances of provision of wrong or exaggerated data. The confidentiality and privacy associated with most disorders in psychology also impact the reliability of data collected through self-report forms. Despite such obstacles, the approach has been very

effective in studying behavioral, social, and cognitive abilities among children. The inclusion of parents, teachers, and guardians in the study process remains significant in achieving credibility of data.

Study Population

The target population refers to entire total number of subjects that the researcher intends to study. However, time and financial resources limits the number of participants that can be involved in a study. The researcher identified PCDH-19 parent groups across the world that have been created for parents of children diagnosed with PCDH-19. Parents were recruited through social media to complete the research study.

Recruitment of the Participants

Recruitment participants for the study included a structured process starting from description of the nature, objectives, and purpose of the study to the potential participants. The ability of the research to provide conclusive and compelling description of their projects including rational and significance can retain all the participants throughout the research process. The participant information sheet was used to provide details for the study including a brief background, problem statement, research aim and objectives, significance, and expected outcomes. The information sheet was used to identify and document the possibilities of the participants and the researcher.

Since the study includes minors, the researcher ensured that that parents or guardians, understood the requirements objectives and requirements of the study of behalf of the children. The informed consent statement was also used to seek agreement from the guardians for the participation of their children in the study. The survey provided brief details on the voluntarily participation of the participants in the study including the freedom

to withdraw from the research process without any reasons. The completion of the survey also served as the agreement that the respondents will participate in the study and provide information towards meeting the objectives and goals of the research project. Importantly, the guardians were notified of the measurement tools used to assess the behavioral, social, and executive functioning skills of the participants.

Sampling and Sample Size

The identified parent groups had over 100 cases of PCDH-19 reported in the last three years. However, the nature of study that includes genetic testing through use of different measurement scales and systems requires a smaller sample to allow for quality data analysis and comparison. Therefore, the researcher identified a study population of 36 families that met exclusion and inclusion criteria. Simple random sampling was used to pick families to participate in the study. However, only 25 parents completed the entire survey at the end of the recruitment process. Thus, the number of participants in the present study included 25 children and adolescents, between the ages of 6 and 17 who have previously been diagnosed with PCDH-19 through genetic testing (n=25).

Table 3.1 Study Population and Sample Size

Sample Size	
Target Population (N)	36
Sample Size (n) - 70% of N	25

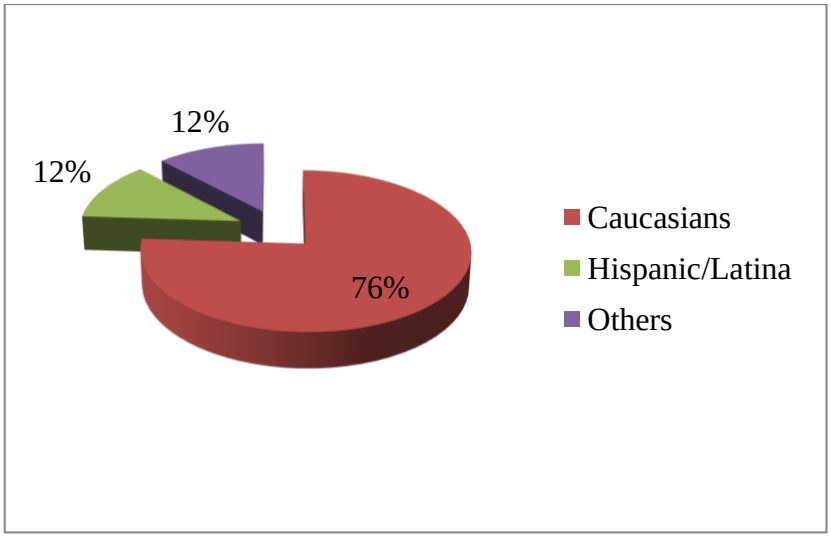
Source: Researcher (2019)

Demography of Participants

Age, social, behavioral, and cognitive development were key features for consideration in picking the participants. Participants ranged in age from 6 to 17, with the

mean age being 9.72 years, 100% were female participants. The ethnicity of the participants was reported. The sample consisted of 19 Caucasians representing 76% of the population, 3 Hispanic/Latina at 12% and 3 as other ethnicities which was 12%. There were no significant group differences between age, gender, ethnicity, socioeconomic status or other medical diagnoses.

Figure 3.1: Participant Demographics



Source: Smith (2019)

Exclusion and Inclusion Criteria

The criteria on inclusion and exclusion will set the boundaries for selection of participants. The researcher included the criteria in the survey to help potential participants evaluate their fitting into the study. Various attributes such as age, health condition, availability, parent or guardian consent, and existing disorders will be used to include or exclude participants. The researcher will replace any respondents included in the sample but do not meet the inclusion requirements.

Table 3.2 Inclusion and Exclusion Criteria

Inclusion Criteria	**Exclusion Criteria**
Aged between 6 and 17 years	Aged below 6 or above 17 years
Participant diagnosed as having PCDH-19 epilepsy	Participant does not have diagnosis of PCDH-19 epilepsy
Parent/guardian completed survey in its entirety	Parent/guardian did not complete survey in its entirety

Measures

Levan (2015) notes the existence of many tools and techniques that have been used to assess the social and executive functioning among young populations. The common tools include the Social Skills Improvement Scale (SSIS), the Behavior Assessment System for Children, Third Edition, (BASC-3), and the Behavior Rating Inventory of Executive Function, Second Edition (BRIEF-2).

Social Skills Improvement Scale (SSIS)

Crowe, Beauchamp, Catroppa, and Anderson (2011) point out that the SSIS is one of the most common measures for social skill development, behavioral changes, and academic capability. The assessment tool was developed in 1990 to help parents and teachers establish any abnormalities on the elementary, preschool, and secondary school learners. The techniques include 80 items and provides scores on social skills, academic competence, and problem behaviors. Percentages are used to rank the different scores with interventions to any abnormalities included as recommendations to improve social skill development and suppress identified problem behaviors.

The Social Skills Improvement Scale (SSIS) is a multi-rater assessment of student social behaviors that can affect student-teacher relations, peer interactions, and academic performance. The SSIS provides information on the domains of social skills, problem behavior, and academic competence. The parent and teacher forms are available for various age ranges which cover preschool, kindergarten through grade 6, and grades 7 through grade 12. The measure takes 10-25 minutes to complete (Crowe, Beauchamp, Catroppa, & Anderson, 2011). Prior to the SSIS, it was the Social Skills Rating System (SSRS) which was created to look at social skills and had been standardized for a sample of over 4000 children without disabilities (Levan, 2015).

The SSIS has become one of the most used measures in looking at social skills of school-aged children. The SSIS has not been used in the literature with children diagnosed with PCDH-19 yet. In this study, SSIS will be used to measure the levels of social skills among the participants as well as identify the existence of problem behaviors that can hinder the normal growth of interaction and social competencies. SSIS will be used to measure behavior, interaction, and emotional changes through the social skills scale. The social skill screen uses both student and parent forms to assess the changes in interactions, behaviors, and emotions among the subjects. The specific areas for assessment in social skill scale include communication, assertion, cooperation, empathy, responsibility, self-control, and engagement.

The Behavior Assessment System for Children, Third Edition, (BASC-3)

BASC-3 is a rating questionnaire designed to identify differential diagnosis and classification of emotional and behavior disorders in children. The BASC-3 is the latest revision of the BASC. The BASC-2 has been used to examine the emotional and behavioral

functioning of children diagnosed with epilepsy. Titus, Kanive, Sanders, and Blackburn (2008) used the BASC-2 on a pediatric population diagnosed with epilepsy and found that the children who had poor seizure control had higher parent and teacher ratings on the depression, somatization and withdrawal subscales. Levan (2015) used the BASC-2 in looking at the social skills and executive functioning in children with epileptic and non-epileptic seizures.

The BASC-3 parent rating form is comprised of 175 questions that a parent rates using a four-choice response format. There are no current studies in literature that have used the BASC-3 with children diagnosed with PCDH-19. BASC-3 consists of different rating scales to broaden the results and make appropriate recommendation before making conclusions. The typical assessment tools include parent rating scales (PRS), teacher rating scales (TRS), student observation system (SOS), Self-report of personality (SRP), and structured developmental history (SDH).

The combination of the different scales enables creation of a single result database that tests social skills, emotions, problem behaviors, psychological stability, and executive functioning abilities. BASC-3 is restricted to children and adolescents, thus is not considered appropriate for adult assessments.

Essentially, the screening tool provides diagnosis and classification of behavioral and emotional challenges that define the social skill capability among children. The scales provide measurements for the levels of anxiety, aggressions, learning challenges, attention, and conduct problems when compared to a normal sample. The researcher will use the guardian form to collect data on the social components to evaluate the social skills on the sampled population. Preliminary studies show that students with identifiable social,

behavioral, and emotional skills score high on the depression and anxiety scales and low on attention and learning measures.

The BASC-3 will assist in linking the social and behavioral outcomes of different measurement scales to cognitive and executive functioning of the participants. The BASC-3 scales that will be included in this study include attention, learning abilities, depression, and interpersonal relations scales. These measures are considered more impactful in measuring social skill functioning among children diagnosed with epilepsy.

The Behavior Rating Inventory of Executive Function, Second Edition (BRIEF-2)

The BRIEF-2 is a comprehensive and efficient assessment that looks at executive functioning. The BRIEF-2 is a 63-item questionnaire that assesses executive functioning such as planning/organization, initiation, attention, problem solving, and skills related to emotional regulation. The measurement tool is an update to a previous 86-item questionnaire that was used to measure executive functioning through different scales such as problem solving and attention. The specific measurements will include behavioral regulation, cognition index, self-management of tasks, problem solving, and self-evaluation and monitoring.

Data Screening and Cleaning

The common attributes of data collected from heterogeneous population include inconsistencies and missing responses. Therefore, the data was screened for missing responses and for overly negative or inconsistent responses. Of the 25 cases, all measures were complete and had no missing responses. A review of the responses also found that there were no measures that had an overly negative or inconsistent response pattern. It was

noted that because the literature is lacking in PCDH-19, parents were highly motivated to contribute to the research process. The researcher relied extensively on the responses from the parents/guardians to build concrete conclusion on the topic. The experiences among parents with children diagnosed with PCDH-19 provided valuable information that closed any data gaps and inconsistencies.

Normality tests are essential in research to ensure that random selection of variables are evenly distributed to guarantee validity of data. Tests of normality were run. Levan (2015) noted that due to a small sample size, the distribution shape via histograms should not be relied upon as it is often not a good approximation to normal in small samples. Because the sample size was small, normality was determined from the Shapiro-Wiik test. The results of the Shapiro-Wiik test showed that the assumption of validity was not violated in this study.

CHAPTER 4: RESULTS AND FINDINGS

The first aim of this study was to investigate whether children diagnosed with PCDH-19 have demonstrated impairment in social skills. In order to investigate this question, Social Skills Improvement Scale (SSIS) and The Behavior Assessment System for Children, Third Edition, (BASC-3) were used to measure the different social, behavioral, and emotional skills among the children diagnosed with PCDH-19. The SSIS was used to measure social skills, academic competence, and problem behaviors through specific areas including communication, assertion, cooperation, empathy, responsibility, self-control, and engagement. On the other hand, BASC-3 measured anxiety, aggression,

learning challenges, attention, and conduct problems through the parent rating scales (PRS).

Descriptive Statistics for SSIS

The descriptive statistics for social skill scales were obtained and are as shown in table 1.

Table 1: Descriptive Statistics

	N	Minimum	Maximum	Mean	Std. Deviation
Communication	25	5.00	17.00	11.2800	3.66879
Cooperation	25	4.00	16.00	10.8400	3.57864
Assertion	25	6.00	17.00	11.2800	3.40979
Responsibility	25	6.00	16.00	10.7200	3.82448
Empathy	25	4.00	14.00	8.7600	3.34515
Engagement	25	6.00	13.00	9.2800	2.40693
Social skills standard score	25	6.00	12.00	8.8400	2.21133

Results in table 1 show that the minimum scale for communication was 5, the maximum was 17, the mean was 11.28 while the standard deviation was 3.66. This shows that most of the children had poor communication skills. Furthermore, the minimum scale for cooperation was 4, the maximum was 1, the mean was 10.84 while the standard deviation was 3.57. This also shows that most of the children had below average corporation skills. The minimum social skill standard score was 6, the maximum was 12, the mean was 8.84 while the standard deviation was 2.21. this implies that most of the children with PCDH-19 had below average social skill standard scores.

Descriptive Statistics for BRIEF-2

Descriptive statistics were obtained for ratings on executive functioning variables and the results were as shown in table 2.

Table 2: Descriptive Statistics

	N	Minimum	Maximum	Mean	Std. Deviation
Planning	25	63.00	79.00	69.9200	4.36768
Attention problems	25	64.00	80.00	71.9200	4.83839
Problem Solving	25	51.00	73.00	63.6000	5.49242
Self-evaluation	25	66.00	73.00	69.3200	1.88680
Cognitive index	25	63.00	75.00	69.8000	3.71932
Emotional regulation	25	62.00	72.00	68.0800	2.82725
Organization	25	62.00	74.00	68.3200	2.73435

From table 2, it is evident that the executive functioning variable that had the highest mean rating (71.92) was attention problems while the one that had the lowest mean rating (68.08) was emotional regulation. Furthermore, problem solving had the smallest minimum rating (51) while attention problems had the highest maximum rating (80). All scores were noted to be elevated and would require follow-up. The mean for Attention Problems was noted to be the highest and was in the clinically significant range.

Descriptive statistics for BASC-3

Table 3: BASC-3 Descriptive Statistics

	N	Minimum	Maximum	Mean	Std. Deviation
Externalizing	25	54.00	81.00	68.0000	6.74537
Hyperactivity	25	45.00	77.00	71.4000	6.91014
Aggression	25	48.00	75.00	64.6000	6.06905
Conduct problems	25	51.00	73.00	63.6000	5.49242
Internalizing	25	49.00	70.00	64.1200	5.27036
Anxiety	25	64.00	73.00	67.5600	2.21886
Depression	25	59.00	67.00	63.8400	2.44404
Somatization	25	58.00	69.00	61.9600	2.92233
Behavior problems	25	62.00	78.00	70.0400	4.73005
Attention	25	64.00	80.00	71.9200	4.83839
Learning problems	25	61.00	74.00	68.4000	3.32916
School problems	25	66.00	72.00	69.2400	1.66533

Adaptive skills	25	16.00	31.00	23.4800	4.43584
Adaptability	25	16.00	29.00	22.4400	3.37984
Leadership	25	17.00	29.00	21.6000	2.64575
Study skills	25	13.00	28.00	21.7200	3.32315

From table 3, it is evident that the variable that had the highest mean rating (71.92) was attention problem while the one that had the lowest mean rating (21.6) was leadership. Furthermore, study skills had the smallest minimum rating (13) while externalizing had the highest maximum rating (81).

Correlation Analysis

Correlation analysis was carried out to determine the relationship between the variables.

Table 4: Correlation between academic competence and communication

		Academic competence	Communication
Academic competence	Pearson Correlation	1	.537**
	Sig. (2-tailed)		.006
	N	25	25
Communication	Pearson Correlation	.537**	1
	Sig. (2-tailed)	.006	
	N	25	25

**. Correlation is significant at the 0.01 level (2-tailed).

Pearson product correlation was run to determine the relationship that exists between academic competence and communication among children with PCDH-19. Results in table 4 show that there is a strong relationship between academic competence and communication which was statistically significant (r=0.537, n=25, p=0.001).

Table 5: Correlation between academic competence and Responsibility

		Academic competence	Responsibility
Academic competence	Pearson Correlation	1	.744**
	Sig. (2-tailed)		.000

	N	25	25
	Pearson Correlation	.744**	1
Responsibility	Sig. (2-tailed)	.000	
	N	25	25

**. Correlation is significant at the 0.01 level (2-tailed).

The Pearson product correlation was run to determine the relationship that exists between academic competence and responsibility among children with PCDH-19. Results in table 5 show that there is a strong relationship between academic competence and responsibility which is statistically significant (r=0.537, n=25, p=0.000).

Table 6: Correlation between academic competence and social skills standard score

		Academic competence	social skills standard score
	Pearson Correlation	1	.781**
Academic competence	Sig. (2-tailed)		.000
	N	25	25
	Pearson Correlation	.781**	1
social skills standard score	Sig. (2-tailed)	.000	
	N	25	25

**. Correlation is significant at the 0.01 level (2-tailed).

The Pearson product correlation was run to determine the relationship that exists between academic competence and social skills standard score among children with PCDH-19. Results in table 6 show that there is a strong relationship between academic competence and social skills standard score which is statistically significant (r=0.781, n=25, p=0.000).

Table 7: Correlation between academic competence and corporation

		Academic competence	Cooperation
	Pearson Correlation	1	.586**
Academic competence	Sig. (2-tailed)		.002
	N	25	25
Coorperation	Pearson Correlation	.586**	1

	Sig. (2-tailed)	.002	
	N	25	25

**. Correlation is significant at the 0.01 level (2-tailed).

The Pearson product correlation was run to determine the relationship that exists between academic competence and cooperation among children with PCDH-19. Results in table 7 show that there is a strong relationship between academic competence and cooperation which is statistically significant (r=0.586, n=25, p=0.002).

Table 8: Correlation between academic competence and engagement

		Academic competence	Engagement
Academic competence	Pearson Correlation	1	.565**
	Sig. (2-tailed)		.003
	N	25	25
Engagement	Pearson Correlation	.565**	1
	Sig. (2-tailed)	.003	
	N	25	25

Pearson product correlation was run to determine the relationship that exists between academic competence and engagement among children with PCDH-19. Results in table 8 show that there is a strong relationship between academic competence and engagement which is statistically significant (r=0.565, n=25, p=0.003).

Table 9: Correlation between planning and cognitive index

		Planning	Cognitive index
Planning	Pearson Correlation	1	.638**
	Sig. (2-tailed)		.001
	N	25	25
Cognitive index	Pearson Correlation	.638**	1
	Sig. (2-tailed)	.001	
	N	25	25

**. Correlation is significant at the 0.01 level (2-tailed).

Pearson product correlation was run to determine the relationship that exists between planning and cognitive index among children with PCDH-19. Results in table 9 show that there is a strong relationship between planning and cognitive index which is statistically significant (r=0.638, n=25, p=0.001).

Table 10: Correlation between planning and emotional regulation

		Planning	Emotional regulation
Planning	Pearson Correlation	1	.240
	Sig. (2-tailed)		.248
	N	25	25
Emotional regulation	Pearson Correlation	.240	1
	Sig. (2-tailed)	.248	
	N	25	25

The Pearson product correlation was run to determine the relationship that exists between planning and emotional regulation among children with PCDH-19. Results in table 10 show that there is a weak relationship between planning and cognitive index which is not statistically significant (r=0.240, n=25, p=0.248).

Table 11: Correlation between planning and problem solving

		Planning	Problem Solving
Planning	Pearson Correlation	1	.235
	Sig. (2-tailed)		.259
	N	25	25
Problem Solving	Pearson Correlation	.235	1
	Sig. (2-tailed)	.259	
	N	25	25

The Pearson product correlation was run to determine the relationship that exists between planning and problem solving among children with PCDH-19. Results in table 11 show

that there is a weak relationship between planning and cognitive index which is not statistically significant (r=0.235, n=25, p=0.259).

Table 12: Correlation between depression and social skills

		Depression	social skills
Depression	Pearson Correlation	1	.071
	Sig. (2-tailed)		.736
	N	25	25
social skills	Pearson Correlation	.071	1
	Sig. (2-tailed)	.736	
	N	25	25

The Pearson product correlation was run to determine the relationship that exists between depression and social skills among children with PCDH-19. Results in table 12 show that there is a weak relationship between depression and social skills which is not statistically significant (r=0.071, n=25, p=0.736).

Table 13: Correlation between communication and social skills

		Social skills	Communication
Social skills	Pearson Correlation	1	.499*
	Sig. (2-tailed)		.011
	N	25	25
Communication	Pearson Correlation	.499*	1
	Sig. (2-tailed)	.011	
	N	25	25

*. Correlation is significant at the 0.05 level (2-tailed).

The Pearson product correlation was run to determine the relationship that exists between communication and social skills among children with PCDH-19. Results in table 13 show that there is a weak relationship between communication and social skills which is not statistically significant (r=0.499, n=25, p=0.011).

CHAPTER FIVE: DISCUSSION

The number of people and children with social skills and executive functioning problems has increased creating a need to understand more about PCDH-19. The children affected with PCDH-19 demonstrate social and cognitive dysfunctions and the aspect establishes a requirement for therapists to understand the causes, symptoms and treatment measures of the disorder. Understanding the social, cognitive and psychological development of children remains vital in addressing the effects of PCDH-19. Numerous cases of PCDH-19 are identified each year, and the discussion of the results seeks to provide more understanding regarding the disorder, social skills and executive functioning of children. The discussion of the results shall close the existing gaps in understanding the effects of PCDH-19 on social skills development and executive functioning among those diagnosed.

The social and cognitive dysfunctions of children are attributed to PCDH-19, and the discussion provides an essential understanding of the aspect. The discussion covers the literature gap on the understanding of PCDH-19 and social, cognitive and behavioral dysfunctions among children. The change in cognitive development at an early age has led to the growth of both epileptic and non-epileptic seizures and people need to differentiate their causes, impacts, and treatment. Moreover, the chapter covers diagnostic and intervention mechanisms for PCDH-19 as well as the limitations.

The results demonstrated that children experience PCDH-19 epilepsy through the early onset of seizures, cognitive delays, and behavioral problems. Mutations in PCDH-19 have a high influence on the cause of epilepsy among girls specifically. The early seizure onset seems to contribute to the cognitive and behavioral problems among the girls. During

particular stages of cognitive development among girls, the brain undergoes some changes that lead to the development of various cognitive processes and specific behaviors. The results have demonstrated that during early development, the functional changes of the brain coincide with cognitive and behavioral alterations and increases the onset of seizures at an early stage.

Considering the data and results from the study, the children demonstrated impairment in social skills and deficits in executive functioning. Based on ratings provided by the parents, children with PCDH-19 exhibited poor and abnormal social skills. About the individual functioning domains, the children were found to have specific deficits in areas of communication, cooperation and self-control among the others evaluated. The findings concur with the clinical impressions that demonstrate that children with PCDH-19 have social difficulties. The needed interventions should precisely help the children communicate and cooperate with the other peers, parents, and teachers through initiating social interactions and being responsible for social and interpersonal solutions.

Examining the social skills and executive functioning of the children sometimes becomes critical on how to intervene. Children with PCDH-19 have challenges in inhibiting behaviors that undermine learning and social interactions. Children diagnosed with PCDH-19 experience decline in social skills that undermine the interactions, communication, problem-solving, and self-control (Austin, Dunn, Johnson and Perkins, 2004). Both parents and teachers working with children with PCDH-19 describe numerous challenges that affect them. The overall social skills and intellectual ability of children with epilepsy have deficits that undermine their social interaction and learning capabilities.

PCDH-19 can lead to multiple problems in different areas such as behavioral and emotional adjustment, social competence and academic achievement.

Self-control and assertion were the main domains highly scored indicating difficulties among the children in such attributes. The average scores in this SSIS assessment showed that the sampled children exhibited fewer behaviors, social skills, and academic competence compared to their peers. Most children diagnosed with PCDH-19 experience decline in social skills and other attributes that rely on interactions, communication, problem-solving, and self-control. Bara, Bosco, and Bucciarelli, (1999) indicate that the effects undermine learning, executive functioning and social interaction.

Children with PCDH-19 demonstrate challenges in self-control and assertion as well. Children with high and average scores of the SSIS assessment exhibit fewer behaviors, social, skills, and academic competence when compared to peers without the syndrome. The outcomes conformed to existing evidence that children diagnosed with PCDH-19 experience decline in social skills and other attributes that rely on interactions, communication, problem-solving, and self-control. The outcomes were analyzed to determine the impact on academic and executive functioning to create relationships needed in completing this investigation.

Social engagement and children participation in physical activities among peers remains a regular issue during early childhood. From the results, seizures and social skills impairment and exclusive functioning challenges among the children exclude them from the full participation in the social experiences, academic and recreational practices. Children with PCDH-19 raise concerns on their safety and social isolation from the parents and peers. Parents may advocate for extra support for their children from the schools to

promote social interaction through engagement in different activities. When children with PCDH-19 engage in social interaction activities, they have the opportunity to build their social and physical development as well as emotional wellbeing.

The stigma associated with epilepsy among children undermines the adaptive and emotional behavior responses. Children with epileptic disorders can get isolated and bullied by peers, and the issues alienate them from the social interactions among peers. Children may feel inadequate to interact with their peers, and the this can foster low self-esteem. These experiences can alienate epileptic children from the social learning experience that promotes social development and self-esteem. Although children with learning disabilities are attributed to extensive brain abnormalities, most of them have focal brain abnormalities such as seizures and mesial temporal sclerosis. Seizures and fatigue often lead to transitory learning problems that make the children have poor performances at school. Educational staff should consider proper teaching and learning strategies to improve the academic success of children who have epilepsy.

Children with PCDH-19 demonstrate emotional and behavioral challenges. As well. Increased anxiety, depression and irritability occurs among the children with PCDH-19. Aggression among epileptic children occurs in different ways depending on the brain and the anti-epileptic drug therapy. Parents in clinical settings indicate different scenarios in which they observe different levels of aggression among their children. Introduction of increased dosages of anti-epileptic medications coincides with behavioral changes such as physical aggression, irritability, hyperactivity, and depression.

The results demonstrated that children with epilepsy have more significant cognitive dysfunctions when compared to peers without the syndrome. BRIEF-2 examined

the cognitive dysfunction of children with PCDH-19. Some of the children diagnosed with the PCDH-19 have impairment in executive functioning. The children show poor planning, organization, attention, problem-solving, self-evaluation, cognitive index, and emotional regulation. Parents and others who work with children with the PCDH-19 may struggle due to low executive functioning regarding such aspects.

According to Austin et al. (2004), social skills facilitate communication and enables people to have acceptable behavior in society. Acquiring proper social skills begins from childhood through different development stages until an individual becomes an adult. However, social skills problems emanate through PCDH-19 and other epilepsy syndromes thus causing impairments and dysfunctions among children. The results of the research study demonstrate that children with PCDH-19 epilepsy have problems in social skills and executive functioning. The children exhibited severe challenges based on the subscales such as communication, cooperation, empathy, responsibility, and engagement.

The actual nature of epilepsy reduces social interaction among children which affects their academic affairs and cognitive development. Poor social skills lead to social isolation, poor social adaption, and low self-esteem. Children with PCDH-19 demonstrated low social competence when compared to peers without epilepsy. When children with PCDH-19 become adults, they may still exhibit social problems despite being intellectually and neurologically in the normal range. The SSIS demonstrated that children with PCDH-19 have social skill problems that affect their behavior, academic performance, and social interaction. Children with PCDH-19 have low social competence skills when compared to peers without the syndrome.

Results also showed that the children had poor assertion and indicates that they might be less able to ask for information when compared to their peers. The aspect was evident in the relationship to both the parents and teachers. Results also showed that the children were less responsible and less likely to have better communication with their peers, parents, and teachers. The children also demonstrated poor and abnormal self-control in a conflicting situation that needs critical decisions.

Results also indicated that the children had more internalizing problems such as depression and anxiety. The factors contributed to the low social competence among PCDH-19 children. This aspect could be attributed to various elements that affect the children physically and psychologically. Increased social issues, behavioral and emotional challenges can be an adaptive reaction to an ailment or reaction to the psychosocial and biological aspects.

Children with social skills and behavioral challenges are exposed to personal and learning problems (Bishop and Baird, 2001). The seizures interrupt and slow down many body processes that occur during growth and development. Most operations are essential for the development of the brain, and when interrupted, they damage the neural leading to accumulative neuropsychological disabilities. Early seizures induce increased epilepsy sensitivity and durable defects that are witnessed through social skills and behavioral measurement skills. Most people with epilepsy have attention problems, slow mental processing and memory deficits (Bishop and Baird, 2001). Based on data collected, appropriate knowledge among the parents and the teachers increases the probability of the children having less effects due to increased interventions.

Considering the increased prevalence of PCDH-19 among children, behavioral and psychological problems remain a challenge. Behavioral and emotional problems caused by neurological disorders have adverse effects on children. The intellectual capacity gets affected, and the ability to learn becomes challenging. PCDH-19 is not a mere medical disease but may have psychological impacts on the children (Austin et al., 2004). Social skills problems of children with epilepsy go hand in hand with the behavioral and emotional challenges. The issues affect all the activities of the children due to the psychological and social problems caused by the disorder.

Children with epilepsy often have increased emotional and behavioral impairments when compared to their peers without epilepsy. Geurts and Embrechts (2008) indicate that epilepsy affects behavioral development of children which undermines most of their activities. The aspects indicate that the clinicians should prioritize the diagnosis and management of epilepsy among children that have such symptoms. The average of the social skills scale exhibited that the children with PCDH-19 have abnormal behavior aspects that affect their lives. PCDH-19 causes or worsens the existing behavior challenges among the children. When seizures began at an early age, the children endure troublesome behavior problems that end up affecting their normal activities. Normal brain functions can be affected by brain damage, seizures, electrical discharge between seizures and the effects of seizure medicines.

Studies demonstrate that the diagnosis of children with epilepsy depends on how the parents and other people react to the behavioral problems of the children. In the beginning, many parents get overwhelmed when their children are diagnosed with epilepsy. Although it appears reasonable for the parents to have such experiences, the aspect affects

the children self-concept and may undermine the recovery process of the child. Most parents feel depressed when the children seizures do not stop, and the element makes the parents even withdraw the child from active medical care which becomes dangerous.

Children with PCDH-19 had poor communication and showed abnormal skills to the parents. Bishop and Baird, (2001) demonstrate that people with epilepsy face language and communication challenges that compromise their social interaction and learning ability. Communication impairment is among the aspects that determine epileptic syndrome among children. The children with PCDH-19 indicated delayed or lack of communication gestures and proper spoken language development. Children with epilepsy have challenges to maintain and sustain communications with their parents and teachers. Communication behavior of PCDH-19 children involves poor gestures and vocalization. The younger children with PCDH-19 have problems in using joint attention acts and gestures which undermines their ability to coordinate their communication.

The level of communicative challenges among children with epilepsy at an early age determines how the effects can be severe after growing up. Children with epilepsy often use rigid language that cannot be understood by the people. Parents recorded abnormal communication of their children with PCDH-19. The children did not maintain eye contact, exhibit politeness nor appreciated during communication. Under such circumstances, peers, parents, and teachers endure a challenging experience to handle the children. Communication behavior undermines their social interaction and learning. In most cases, the communication behavior communication problem among children leads to isolation and intensifies other adverse effects such as stress and depression.

The children exhibited abnormal cooperation behavior rating as indicated by their parent's ratings. Results showed that children with PCDH-19 have challenges in following rules, assisting and sharing with the other people. The BASC-3 results that measured the social functioning of the children show that the social skills aspects are influenced by external and internal factors, behavioral symptoms and adaptive skills. External factors such as conduct problems and aggression influence the behavior of an individual and undermine the cooperation. Anxiety and depression make people uncomfortable and determine how people associate themselves with others (Bara, Bosco, and Bucciarelli, 1999). The parents recorded high behavioral symptoms index on attention and learning problems. The components have a high influence on the children's behavior, and their prevalence undermines the cooperation.

The assertion rating recorded high levels. PCDH-19 children have challenges in introducing themselves, seeking information from their peers and providing responses to any issues concerning them. The negative assertion behavior is attributed to the external and internal problems that affect the children. Hyperactivity, aggression and conduct problems lower the assertion behavior of epileptic children. Disruptive behavior undermines the assertion virtue and makes it challenging for peers, parents and the teachers to handle the children. Psychological distress and attention problems also have a negative influence on the assertion behavior of children with PCDH-19.

Parents reported abnormal rating on the empathy aspect, thus indicating that the children have difficulty expressing empathy and this could be affecting their social skills. The high scores recorded in anxiety, issues of attention and school show that it's challenging for the children to have good behavior.

People with epilepsy often may have poor self-control due to behavior challenges that result from adverse effects of the disease. The results concur with the findings made in other studies that exhibit that the children with epilespy have poor and abnormal self-control. The BASC-3 rating shows that the attributes measured have a high influence on the social and emotional functioning of the epileptic children. Poor rating on the internal and external aspects contributes to poor self-control among the epileptic children (Bishop and Baird, 2001). Moreover, attention and learning problems affect the self-control aspect of PCDH-19 children and intensify the self-control woes.

The association between social skills, behavioral and emotional functions have demonstrated close links to each other. The challenges of the social abilities undermine the social tasks of children with epilepsy. When the children have high abnormal levels of aggression, hyperactivity, and aggression, it becomes difficult for them to have proper communication, self-control and other vital components that promote social life activities. The results support the findings that the social skills and behavior aspects of PCDH-19 correlate.

Social bonds exist at the center of daily living among the people and determine the quality of life people expect to live. Epilepsy among children impedes social functions and compromise social interactions. Neuropsychological deficits such as anxiety, memory loss, and attentional difficulties interfere with the reciprocity of the social interactions among the affected children (Parsch and Ellegren 2013). Children with PCDH-19 have exhibited lower social competence as indicated by parent reports. The deficits in social functioning contribute to challenges in learning, social interactions and engaging in other life aspects. When aiming to improve the life quality of children with epilepsy, social functioning

remains an essential consideration. The impairments witnessed in social functioning among the children, makes it difficult for them to be responsible, assertive and self-controlled (Norbury, Nash, Baird and Bishop, 2004). When the challenges are not controlled at an early age among the children, they become adverse when the children become adults.

The results demonstrated a close relationship between the social skills and executive functioning of the respondents. The findings supported the hypothesis that social skills and executive functioning correlate. The children with PCDH-19, BRIEF metacognition index measurements exhibited a correlation with the BASC-3 adaptive skills. Parent's reports of the children on the Brief behavioral regulation index significantly correlated with parental SSIS social skills for the PCDH-19 children. Previous studies indicate that poor peer relationship exists due to the impaired executive functioning among people with epilepsy. Children with the PCDH-19 have demonstrated poor social interaction among peers due to poor executive functioning. From the results, the children exhibited high levels of BRI an aspect which proved poor and abnormal social skills ratings.

Cognitive problems among children with epilepsy remain a critical issue in the contemporary world (Ketelaars, Cuperus, Van Daal, Jansonuis and Verhoeven, 2009). The cognitive function involves several skills that include attention, learning, reasoning, and planning among others. The skills influence other services such as behavior and emotions that remain critical in people's daily life. During development, the growth of cognitive processes is prolonged among children from infancy to adolescence. Also, other aspects such as the executive functions extend to adulthood.

Studies indicate that early diagnosis of children with epilepsy show that most of them have cognitive deficits that undermine their learning, attention, and concentration. The challenges are attributed to structural brain anomalies that endanger the proper functioning of the neurological system — children with the epilepsy record poor cognitive tasks rather than the peers without the syndrome.

Many people with epilepsy have executive functions damages. The adverse effects originate from biological, medical and psychosocial factors interact to produce executive functions problems. The effects of epilepsy affect specific cognitive activities and cause challenges that undermine the executive functions. Children with epilepsy exhibit learning and memory deficits that affect their daily activities. Through parent report, the findings of the study indicated that children with PCDH-19 had cognitive problems that affected their behavior and emotions. The cognitive effects of epilepsy among children remains a concern to parents and teachers since the impairments affect the learning capability and memory functioning of the children. Poor executive functions exist among children diagnosed with epilepsy. The executive function skills such as attention, organization and working memory play a vital role in memory development and improve in attaining goal-directed behavior, planning, and goal formation.

Children are prone to executive dysfunctions due to poor skills on performance-based and neuropsychological measures (Parsch & Ellegren 2013). Executive functions encompass daily activities such as adaptive social skills, social interaction, and learning. Disruption of the typical acquisition of social skills affects the executive functions which affects the children's quality of life. The study provided more evidence that executive dysfunction is a challenge associated with children with PCDH-19 that have poor and

abnormal social skills. The results were congruent with other previous studies on the BRIEF in epileptic children on the association of social skills and executive functions.

Centers for Disease Control and Prevention (2016) reveals that executive dysfunctions are associated with epileptic disorders. This study indicates that children with PCDH-19 have clinically significant problems with working memory and planning. The findings exhibited that executive function deficits were a major barrier for the children to achieve the high related quality of life. The BRI rating of inhibition, shift and emotion control were related to all the aspects of HRQOL. Also, the metacognition index rating indicated that the working memory, panning, and organizing were also associated with most dimensions of HRQOL.

The findings indicate that although difficulties in planning and organizing are common in PCDH-19 children, other executive function domains such as the working memory, shifting, and inhibition translated into the HRQOL. The aspects of behavioral regulation index namely, inhibition, shifting and emotional control recorded high executive function effects. The percentile rating demonstrated that a little difference in the EF aspects among other children of the same age group diagnosed with PCDH-19 epilepsy. The high T-scores for the BRI attributes indicate that the children in the study have challenges in controlling emotions, controlling their behavior and shifting between activities. Therefore, based on the findings of the study, executive function deficits contribute much to the HRQOL.

The executive functioning problems among children with PCDH-19 have become more evident as indicated in the findings from the study. Wolfe, Walsh, Reynolds, Mitchell, Reddy, Paltin, and Madan-Swain (2012) demonstrated that children with epilepsy

exhibit that they have poor executive functions when compared to non-affected peers. The executive function deficits have cognitive, emotional and learning challenges that affect daily life activities of the children with the syndrome. The identification of executive dysfunctions affecting children with PCDH-19 remains essential for the development of a preventive intervention to improve social and academic aspects. From this study, the BRIEF results have demonstrated the relationship between executive dysfunctions and the poor health-related quality of life.

Children with PCDH-19 demonstrated high executive functioning deficits as measured through the BRIEF scale. The MI domains that comprise of working memory initiating, planning and organizing remain more important for school going students. When challenges occur in the aspects among the children with PCDH-19, it becomes challenging for the parents, teachers, and clinicians to identify them when compared to the components within the behavior regulation such as inhibition and the emotion control. The metacognition aspects are essential for social function and parents should not overlook them. The findings of the study provide extra knowledge about the relationship between the various elements of social competence and executive functioning of the children with PCDH.

The relationship between the executive functions and the social function among children with epilepsy has been explored in different research studies. The findings generated by the study are in line with other studies that demonstrate that BRI and MI from BRIEF-2 play a role in the social function among the children with PCDH-19 epilepsy. The study did not show a vast difference between the BRI and MI components. The number of participants was enough to examine the BRI and MI aspects of the PCDH-19 children.

The findings determined how behavior and metacognition aspects affect the executive function of epileptic children.

The BRIEF scores for the children with PCDH-19 exhibited higher executive function deficits in behavioral and metacognition aspects. Executive functions encompass data collection of different abilities that enable the people to engage effectively in problem-solving situations and adapting to varying conditions in the real world. From the study findings, PCDH-19 children indicated low levels of inhibition aspect which exhibited that they cannot choose and resist interference when encountered in complicated situations. The inhibitory control of attention enables children to focus on what they have selected and avoided other stimulating events that can distract them (Breuillard, Leunen, Chemaly, Auclair, Pinard, Kaminska, Nabbout, 2016).

The executive functions include complex brain functions that control neurological functions that involve problem-solving abilities and control of the goal-oriented behavior among other factors (Wasserman, Hilliard, Schwartz and Anderson, 2015). The behavioral rating inventory of executive function has been constructive in examining the daily executive dysfunctions of people with epilepsy. Decreased attention and learning problems are identified among most people with epilepsy (Elliott, Lach, and Smith, 2005). Children with PCDH-19 have challenges in adaptability and social skills, an aspect that undermines their daily living activities. From the study findings, children with PCDH-19 epilepsy are perceived to have severe difficulties with metacognitive skills. The raw measurements from the clinical scales indicated that children with PCDH-19 have executive function deficits that undermine their social skills.

Executive functions play an essential role in the children's learning process as well. Children have the opportunity to organize their behavior and control their learning for them to record proper learning results. During the process, children require metacognition skills that help them on how to manage their thinking (Freeman, Locke, Rotheram-Fuller, and Mandell, 2017). Thinking meta-cognitively is essential for the children to hold goals in the working memory, inhibit behaviors that are not needed in the current task and shift attention when adjusting strategy (Depienne, Trouillard, Bouteiller, Gourfinkel-An, Poirier, Rivier, and LeGuern, 2011). When people suffer from epilepsy, executive function deficits occur as a result of the impairments in metacognitive skills. Executive functions among children mature at different times, and when epilepsy occurs early in children, the cognition aspects of the brain become affected.

The MIT T-scores of the participants indicates an imbalance in the executive functioning of the children. For the children, this aspect leads to language and learning problems that leads to a poor academic outcome. Children with epilepsy develop cognition impairments that disable them to process information correctly and also to adopt a proper adaptive behavior. The memory impairments, attention deficits and mental slowness form part of the cognitive problems that contribute to the developing of behavioral and executive function deficits. Elliott, Lach, and Smith (2005) demonstrate that children with epilepsy tend to be more affected by the epilepsy when compared to adults.

Children with PCDH-19 may have a greater risk of developing memory problems. The children experience impairments in executive functions which leads to slow the decision-making process and poor problem-solving abilities. Fisher et al. (2014) demonstrate that several behavioral and cognitive disorders are associated with epilepsy

and undermine the cognition aspects of the people. The neuropsychological studies show that seizures slow down, accelerate or modify the processes that take place during brain development which leads to executive function deficits. People with epilepsy endure challenges on self-regulated behavior, planning, and goal-oriented activity when engaging in different daily living activities. Children with such difficulties develop anxiety, depression and low self-esteem which affect their ability to learn, solve problems and even make the right decisions.

Limitations

The study involved a small sample of participants, and the aspect makes it cautious when interpreting the results. Despite the sample being relatively small, some previous studies regarding epilepsy have used similar samples and recorded proper findings. The sample of 25 participants was considered vital in the study as other studies examining PCDH-19 epilepsy among children have used a sample of 20 and 30 participants that has proved successful concerning the issues evaluated. Despite the aspect, it is essential for further studies to consider using a larger population sample to increase statistical power and eliminate any statistical errors that could undermine the research findings. Also, the larger statistical sample shall enable the researchers to understand the prevalence and effects of PCDH-19 among various groups.

The research study did not involve a comparison sample to determine the degree of variance between the aspects measured between the children with PCDH-19 and peers without the disease. However, the rating forms used were normed on a typical child population.

The use of self-reports in the study could bring some errors that could undermine the results and findings of the study. Self-reports have been associated with the provision of wrong and false information that could undermine the results of the investigation. The participants were exposed to different evaluations that sometimes could not yield the expected results. Inclusion of the participants from different ethnicities helped in providing a wide range of information and assisting in examining the condition of children with PCDH-19.

From the research results and findings, it is impossible to generalize the results since the collection of data was made on specified and identified participants. The study only had female children with PCDH-19 which might make it difficult to generalize the findings. The study also did not evaluate the treatment interventions of PCDH-19 epilepsy. The research study only focused on the challenges of social skills, cognitive and executive function deficits and did not cover the treatment interventions of the PCDH-19. The treatment interventions of PCDH-19 epilepsy remain vital, and its examination in the study helps in the understanding of the seizure control. Seizures are challenging to control even after the application of medicine, and it could be possible for the research study to evaluate the impact of treatment among the participants chosen.

CHAPTER SIX: CONCLUSION AND RECOMMENDATIONS

PCDH-19 contributes to severe cognitive and executive functions deficits that undermine behavior, emotional, communication and problem-solving abilities of the children. Social and executive functioning are essential aspects of a child's life and influence the daily living activities. Impairments and dysfunction in social skills and

executive functions adversely affect the social, psychological, and cognitive wellbeing of the children, peers, and families. The executive functions of the children depend on the cognitive processes, and when they are affected by the neurological syndromes, it becomes challenging for the children to make the right decisions and also provide solutions to the issues about different aspects of life (Higurashi et al., 2012). The study examined the impairments and the dysfunctions on social skills and executive functions regarding the syndrome.

The methodology incorporated twenty-five participants and indicted different results regarding the various aspects evaluated. The results showed that children with epilepsy develop emotional, social, behavioral, and mental problems compared to healthier peers. The findings exhibited that the significant challenges that face girls with PCDH-19 are the low levels of social competence which affect decision making in friendships, communicating, and interaction. The frequent seizures in children may affect their self-esteem and self-confidence which will in turn inevitably affect their social and emotional development (Bishop and Baird, 2001). Social skills aspects evaluated indicated that the children with PCDH-19 have self-control, responsibility, communication and empathy problems. The SSIS and (BASC-3) measurements exhibited that the children diagnosed with PCDH-19 had poor social skills, behavioral and emotional issues.

The study exhibited a correlation between social skills and executive functioning. The results and findings of the social abilities predicted the behavioral and emotional challenges presented by PCDH-19. Despite the findings of the study, the research experienced some limitations such as the small sample size and use of self-report that is

prone to errors and confidentiality issues. However, the study provides the right area for further research regarding PCDH-19 epilepsy among children.

6.1 Recommendations

The research study provides essential information regarding the correlation between social skills and executive functions among children with PCDH-19. Epilepsy incorporates several syndromes that affect children at an early age, and it is essential to conduct further investigations regarding epilepsy among children (Marini et al., 2010). Limited data exist regarding the PCDH-19 syndrome among young girls, and the study adds more to the existing literature about the disorder. However, PCDH-19 epilepsy is a rare disease, and information is needed for the public to understand more about the disease and how it affects cognitive and executive functions of the children. Most people do not differentiate the different types of seizures that lead to specific epilepsy, and through further research, the people must acquire more understanding about the disease.

However, future researchers should consider using a large sample size and incorporate different age groups to generate broad and adequate information about PCDH-19 epilepsy. The research study has shown the correlation between social skills, behavioral and executive functions but the investigation has not focused much on the treatment interventions of the PCDH-19.

Most countries lack proper information about the existing healthcare resources concerning the management of seizures among children and adults (Iwata, Wakita Shin, Fukuda and Akaike, 2013). The health practitioners in the health care settings have inadequate knowledge about the epilepsy syndromes, and it is essential for further studies to focus on training of the clinicians who should handle the patients. The management of

seizures is challenging, and the doctors require proper knowledge of the epilepsy diagnosis and the treatment interventions. The current gaps concerning epilepsy care, knowledge and education compromise the efforts made in controlling the disorder among the children and the adults. People living with epilepsy endure severe challenges from living with seizures and dealing with treatment side effects and increased chances of early mortality among the children affected with epilepsy.

For the people to provide proper healthcare services to patients with epilepsy, they must understand more about the epilepsy diagnosis, symptoms, treatment, and management. Given the range of effects encountered by the people with epilepsy, it is essential for the patients to get support from the families and the community. The support services offered to the children and adults with epilepsy should focus on reducing stigma, depression, and stress that affect the patients and their families. Data collection and surveillance efforts should be increased regarding epilepsy to increase knowledge of epilepsy to the public.

Moreover, the prevention efforts for PCDH-19 should be developed and evaluated to make them useful. Additionally, new studies should focus on understanding the influence of genetic factors in the rise of PCDH-19 and other epilepsy conditions among children. The existing inquiries do not provide specific findings on the role of genetics in the rising cases of PCDH-19 among the young female population. While this investigation has provided foundational information and results on the association of PCDH-19 with social skills and executive functioning, extensive studies including subjects from both genders is vital in understanding the genetic contribution in the expanding epileptic condition.

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