

Abstract

Prader-Willi Syndrome (PWS) is a rare genetic disorder with two notable manifestations: mild to moderate mental retardation and a constant, insatiable hunger. Many people in the United States and throughout the world have never heard of and do not understand this syndrome. Further, many countries lack the resources for the diagnosis of PWS and the provision of support services for those affected by the syndrome and their families. The purpose of this paper is to address the need for more knowledge on PWS by providing a theoretical framework, a review of the literature about PWS and two projects that focus on the value of inclusion for people with PWS. The projects stem from two different gaps, or needs, in the universal PWS system for inclusion. The first project focuses on the need for inclusion of children with PWS into mainstream extra-curricular activities. The proposed solution, “Putting the Pieces Together: Understanding Prader-Willi Syndrome,” is an interactive program designed to educate non-PWS children in the United States about the syndrome in order to promote acceptance, and thereby a higher probability of inclusion children with PWS. The second project, “Starting a chapter of the International Prader-Willi Syndrome Organization in Costa Rica,” focuses on the need to establish chapters of the International Prader-Willi Syndrome Organization in all countries in order to ensure inclusion into the PWS network for all people affected by the syndrome. This project specifically identifies a process for starting a chapter in Costa Rica, including an outline of tasks, and a cultural analysis of Costa Rica. These projects serve to spread knowledge of the syndrome, which will increase inclusion and acceptance of people with PWS into society and improve the quality of life for everyone involved.

Table of Contents

I.	Theoretical Framework	3
II.	Purpose	5
III.	Personal Motivation for Projects	7
IV.	Review of Literature: The Basics of Prader-Willi Syndrome	10
V.	Project #1: “Putting the Pieces Together: Understanding Prader-Willi Syndrome.”	23
VI.	Project #2: “Starting a Chapter of the International Prader-Willi Syndrome Organization in Costa Rica.”	35
VII.	Reference List	50
	Appendix A	53
	Appendix B	55
	Appendix C	56
	Appendix D	58
	Appendix E	66

This paper is dedicated to my cousin, Gabriella Eisen, and the hard work and dedication to the International Prader-Willi Syndrome Organization of my aunt, Pamela Eisen.

I. Theoretical Framework

To achieve the goal of increasing inclusion for people with Prader-Willi Syndrome (PWS) in all contexts, it is important to look at all of the factors that interact to both create a problem and facilitate a solution. A useful theoretical framework is the systems theory, as utilized in professional social work practice. According to this theory, “human needs and mastery of developmental tasks require adequate resources in the environments and positive transactions between people and their environments” (Hepworth, et. al., 2006, p.17). An environmental limitations, personal dysfunction, or an interaction effect can lead to impairment or stress, which requires a coping response, defined as achieving a more adaptive fit between the person and the environment. Helping individuals to enhance adaptation often necessitates improving access to adequate resources or increasing the availability of environmental support. The systems approach can be applied broadly across a range of diverse interactions, from the individual to the global level to analyze complex [problems and to propose viable solutions. The systems that will be focused on in this paper impact people with PWS, specifically involvement in extra-curricular activities in the United States, and the global PWS network, notably in Costa Rica.

The two over-arching, or macro systems, involved with respect to PWS are the International Prader-Willi Syndrome Organization, also called the IPWSO, and the individual organizations in each of the fifty-four participant countries, including the Prader-Willi Syndrome Association of the United States of America, also called the PWSA (USA). These organizations provide aid to people affected by PWS and their families through funding for research, links to research, medical aid, a wide array of

literature concerning the syndrome, belonging and understanding for people with the disorder, and numerous other benefits for all micro and mezzo level systems. The micro level includes the individuals with PWS, and the systems with which they have daily contact constitute the mezzo level.

The value base of inclusion underlies these projects. It is the belief inclusion is a human right and should be offered to all people, regardless of their genetic blueprints. Strides have been made in the United States and worldwide to diminish racism, and discrimination based on gender; however, trying to overcome ableism is historically overlooked. Ableism, as defined by Thomas Hehir (2006), who is currently the Director of the School of Leadership Program at the Harvard Graduate School of Education, and has formerly served as Director of the U.S. Department for Education's Office of Special Education Programs, is "societal prejudice against people with disabilities." Hehir believes that in order to promote inclusion, it is necessary to "minimize the impact of disability and maximize the opportunity to participate" (Hehir, public speech, 2006). Because "human behavior is more a result of surrounding conditions than individual characteristics and personality" (Ramanathan & Link, 2004, p. 30), all children greatly benefit from inclusive experiences in which people of all different levels of abilities are encouraged to participate and experience acceptance. Inclusion helps to diminish the negative effects of ableism, and thereby promotes the adaptive functioning for all individuals, regardless of their "differential ability."

II. Purpose

Looking at the IPWSO through an application of the systems theory, guided by with the value of increasing inclusion, two gaps in research and opportunities were apparent. First, focusing on the mezzo level of the IPWSO, and the PWSA (USA), a gap was found in materials for educating non-PWS children about PWS. This type of education leads to more acceptance and social inclusion of children with PWS into mainstream activities, which is beneficial for them in numerous aspects of their lives, and also helps to fulfill the basic human right of inclusion. There is a particular lack of research considering on inclusion of people with PWS in mainstream extra-curricular activities. Inclusion into mainstream extra-curricular activities is beneficial for all children involved in the larger PWS system, including the organization, the non-PWS children, the children with PWS, and the families of all of the children. The focus of the first project is on the need for inclusion of people with PWS into mainstream extra-curricular activities in the United States as well as the need for educating children without PWS about the syndrome. In order to fill this gap, an interactive program, “Putting the Pieces Together: Understanding Prader-Willi Syndrome” was designed to educate non-PWS children about the syndrome and to promote acceptance, thereby generating a higher probability of acceptance for the child with PWS into mainstream extra-curricular activities.

Second, when looking on a global, macro level, it is apparent that only fifty-four countries are currently included in the IPWSO network, through a chapter in their country. According to the IPWSO (2007), PWS is equally apparent in all races and/or nationalities. It is important to have the organization active in all countries in order to

provide support and inclusion into the global PWS network across micro and mezzo level systems. While there are many gaps in this global system based on the number of countries not yet included, this project focuses on the need in Costa Rica. A family in Costa Rica reached out to the IPWSO for help with their daughter who they suspected of having PWS during October, 2006. The author was studying in Costa Rica at the time, and was able to meet with the family and help organize the beginning of a chapter of the IPWSO in Costa Rica. The second project, "Starting a Chapter of the International Prader-Willi Syndrome Organization in Costa Rica," outlines the steps taken to start a chapter of the IPWSO in Costa Rica, and provides a cultural analysis of Costa Rica that will enable the IPWSO to better adapt the chapter to the specific needs of Costa Rica which, in turn, will enhance it's ability to aid people with PWS in Costa Rica.

This paper displays an attempt to fill the gaps in a global PWS network through creating inclusive environments for people with PWS. It is comprised of two projects that serve to spread knowledge of the syndrome, which will increase inclusion and acceptance of people with PWS into society and improve the quality of life for everyone involved. In the words of Socrates, "The only good is knowledge and the only evil is ignorance." Promoting the spread of knowledge pertaining to PWS will combat the evil of exclusion based on ableism both in the United States and other societies around the world.

This paper will begin with a personal story from the author is provided in order to explain her ultimate motivation for the projects. Next, a review of the literature on the basics of PWS that includes in-depth information on the history, cause, diagnosis and manifestations is included. Finally, descriptions of both projects are detailed.

III. Personal Motivation for Projects

Band – Aids of every kind, crates filled to the brim with colorful hair bows, and plastic jewelry boxes overflowing with a wide array of childlike bracelets, necklaces, earrings and rings fill my twenty-five year old cousin's niche in her group home. Growing up is a huge challenge in itself, but imagine growing older without maturing at a "normal" rate. Imagine never being able to satisfy a lurking hunger in your gut. This is the life that my cousin, Gabriella, leads.¹ She was born with Prader-Willi syndrome, a genetic eating disorder, and is also diagnosed with moderate to severe mental retardation.

When I was a child, Gabriella and I were inseparable. Despite our five-year separation in age, we were constant playmates and shared a deep, almost primal understanding of each other. We had the ability to communicate without any verbal communication; we could speak in a glance of the eye. I was slightly aware of her odd gait, speech impediments and inability to control her appetite, but I never understood why people would constantly stare and point at her. My aunt always told us that it was "because she [was] just so beautiful," but I remember having a slight notion that my aunt's explanation was not the complete truth.

My family moved to Texas when I was six; Gabriella would no longer be my faithful playmate. The next time I saw Gabriella, I was eight years old. I had grown up. She had too; but not in the same way that I did. She wanted to play the same games from two years ago and she looked the exact same; she had barely grown a ¼ inch. It finally dawned on me that I had surpassed my cousin, both mentally and physically. Gabriella

¹ Gabriella is on the lower end of the spectrum of development for people with PWS; this story depicts her specific situation and is not universal to all people with PWS.

was five years older, but far less mature than me. She was incapable of growing up and maturing as my siblings, friends and I were doing. My parents explained that Gabriella was mentally handicapped and had Prader-Willi syndrome. I began to understand and communicate with her again, but on a much different level. While Gabriella was older than me, I now realized that she would always be stuck at the maturity level of a young child. With time, I began to realize that even though she was different, I was capable of keeping an outstanding relationship with her and that she was an amazing individual with her own talents and personality. I now consider her a close friend and teacher.

People stared at her as if she was an alien. They avoided her as if she was contagious. Alas, it was not because they thought that “she [was] just so beautiful.” Other people did not seem to accept her as a competent human being. This bewildered and infuriated me. At that point in my life, I made a conscious decision to turn my frustration into an opportunity. As I grew older and began asking my aunt questions about the life that Gabby led as a child, I was astonished at how unaccepted she had been in schools, and extra-curricular activities. My aunt had to fight in order for “special” schools and activities to accept her and it was virtually impossible to convince “mainstream” activities/schools to accept her.

My awakening to this situation inspired me to focus on inclusion of people with disabilities into activities that promote their quality of life and acceptance in our society. I gained a true understanding of the hardships they dealt with; I knew that I wanted to do something to change the perceptions and negative attitude that people have towards children/people with disabilities – specifically PWS.

This inspired me to become extremely active with several volunteer organizations that help the special needs community. I love every second of the time that I spend volunteering and working with the people with special needs and their families. During my senior year of high school, I taught dance and creative movement classes to multi-handicapped children and their siblings. While the classes were incredibly challenging to teach, the emotional reward was indescribable when a child accomplished a task in the class, or when a parent smiled as their child performed at the recital. My hope is that more teachers of all kinds will have the opportunity to share this feeling with me, rather than shy away from it or be prevented from it.

*Gabriella, my cousin, was once my playmate; now she is one of my heroes, and close friend. As a young adult, the situation with Gabriella has motivated me to develop a strong compassion for people with special needs and their families. My volunteer work with the special needs community has changed my life in many positive ways. Gabriella taught me how to accept everybody for who they are, no matter how different they may be. She enabled me to develop my passion for working with the special needs community, which has made a significant impact on my life. And most importantly, Gabriella has taught me acceptance, compassion, and appreciation for others who are not like myself – specifically those who have different levels of abilities. Yes, **abilities**. This is perhaps the best “gift” I have received from anyone and this is a “gift” worth sharing.*

IV. Review of Literature

PWS currently affects numerous families, as it is one of the most prominent rare disorders. A syndrome is defined as a “a set of symptoms that that characterize a certain disease” (Morehead, 1995, p.664). There are a host of common symptoms within a PWS diagnosis; not all people with PWS share the same symptoms. However, two major manifestations are universal to PWS. These include mild to moderate mental retardation and a constant, insatiable hunger. Research has found that roughly one in twelve to twenty-thousand individuals in the United States of America are affected based on documented cases, although it is assumed that some cases are undiagnosed (Raab, 1986). According to the PWSA (USA) (2007), PWS is one of the five most common syndromes in birth defect clinics in the U.S.A and is the most common genetic cause of obesity that is identified at this time.

As a measure of reference, PWS is roughly one-fifth as prevalent as Downs Syndrome², the most common syndrome found in birth defect clinics (Aguilar, 1990). Out of one-hundred randomly selected college students, only one could accurately describe Prader-Willi Syndrome, 15 had heard of the syndrome and 84 had never heard of it before. After describing the symptoms, 45 said that they think that they may have met or seen someone with the syndrome. Ninety-five of the students had heard of and could accurately describe Downs Syndrome. Not only is knowledge of PWS sparse in college students, but according to Pamela Eisen, the current president of the IPWSO, it is also sparse in children, adults, and even in medical professionals (Wett, 1983) (Eisen, personal communication, March 2007).

² Downs Syndrome was used as a measure of reference because it is a more commonly known genetic disability.

History of Prader-Willi Syndrome

Three endocrinologists by the names of Prader, Labhart, and Willi published a report describing an unusual pattern of abnormalities in 1956 (Prader, Labhart, & Willi, 1956). This was the first record in medical literature about this syndrome, and it was named Prader-Labhart-Willi Syndrome, and then later shortened to Prader-Willi Syndrome (Bray, et al, 1983). Prader, Labhart and Willi recorded abnormalities including: diminished fetal activity, profound poor muscle tone, feeding problems in infancy, underdeveloped sex organs, short stature, retarded bone age, small hands and feet, delayed developmental milestones, characteristic faces, cognitive impairment, onset of gross obesity in early childhood due to insatiable hunger, and a tendency to develop diabetes in adolescence and adulthood when weight was not controlled (Prader, Labhart, & Willi, 1956).

Cases that presumably included people with PWS were found in earlier research, most notably in a case studied by Dr. Langdon Down, after whom Down Syndrome was named. He recorded the case of a twenty-five year old woman with multiple characteristics of PWS seventy years before Prader, Labhart and Willi identified the syndrome (Gelbart, 1999). Before the syndrome was established, it is assumed that the majority of affected individuals died young because of overeating and complications that arose from the condition. There are multiple accounts of doctors only thirty years ago who told parents of children with PWS to take their affected children home and that “they [were] going to die from overeating” (Nyhan, 2006). The concept that people with the symptoms of PWS had an insatiable hunger was established, but therapy, etc. was not customary until after the diagnosis was established in 1956. It is possible that historical

accounts and literature that condemn and speak of gluttonous people are referring to individuals affected by PWS and that they were strongly discriminated against and even killed as a result of their genetic abnormality.³

There was a substantial amount of research on PWS performed in the 1960's and orthopedic, dental and developmental characteristics that could be used as diagnostic tools were added to those described by Prader, Labhart and Willi. Two distinct and identifiable phases were defined to strengthen the accuracy of the clinical diagnosis of PWS. They were based on the drastic differences that incur as a child with PWS matures from infancy into early childhood and beyond (Donaldson, et al, 1994). These phases consist of Phase 1: the prenatal, neonatal, and early infancy period, and Phase 2, which develops around the age of 2 or 3 continues into adulthood. A third phase has been suggested, but is not yet established. If adopted, the third phase will most likely mark the differences in behavior and PWS characteristics that develop around adolescence.

The 1970's and 1980's brought studies and related literature describing the behavioral, personality and medical problems that are associated with PWS. There was a huge turning point in the understanding of PWS in 1981, when Dr. David Ledbetter and his colleagues discovered that many PWS patients had the same deletion on chromosome 15. This discovery enabled more research into the precise cause of PWS, and the engineering of a genetic method for accurately diagnosing PWS in a person (Butler, & Keder, 2004).

As time went on, studies became more optimistic and focused on ways to help people with PWS, rather than simply describe it. In 1987 there was a study published by Louise Greenswag (1987) that established a correspondence between appropriate

³ Research does not exist on this topic – it is probable; however, not found in research.

nutritional control and the life expectancy of the individuals. The majority of families describe behavioral and emotional abnormalities in their child/sibling with PWS as being a prominent manifestation of the syndrome; therefore, this stage of research about the psychological, emotional and social effects of PWS on the individual and family are of utmost importance. There is still a great need for more research in this area, and it is being continued in current studies (Dykens, Hodapp, & Masion, 1997).

Research today is continuing to focus on possible causes and medications/therapy in order to aid in the quality of life for people with PWS, and hopefully prevent it. According to the Prader-Willi Syndrome Association of the U.S.A. (2007), some of the most recent studies include one on the eating patterns of bears and the relationship to those of people with PWS, and the increased levels of ghrelin, a hunger hormone, in people with PWS. Both of these studies have shown correlations with people affected by PWS. More research is needed to establish significance in both of these contexts. PWS research is active in laboratories around the world with hopes to provide a better explanation of PWS, provide aid for people who are affected, and shed some light into understanding obesity and mental retardation as a whole (Pinyerd, 2002).

Causes of Prader-Willi Syndrome

There are three important genetic characteristics of PWS identified through research. First, more than one gene is involved; these genes are all located in the “long arm” of chromosome 15, in a region labeled 15q11-q13. Secondly, the critical genes must come from the paternal father in order to function properly. While this region does contain genes from the mother, they are “turned off” in a process referred to as genomic imprinting. Historically, genomic imprinting was thought to only be present in plants;

however, PWS research has provided important information on how this can affect humans. Third, there are at least three different chromosomal errors that can cause PWS. All of these are focused in the same region of chromosome 15, and include a paternal deletion, maternal or uniparental disomy, or an imprinting defect (Prader-Willi Syndrome Association (USA), 2007).

The most common cause of PWS, accounting for roughly seventy percent of the documented PWS cases is a paternal deletion of the PWS critical genes (11-13 on chromosome 15). Currently, there is no known cause for the deletion and no way to prevent it. Some studies have indicated “a causal relationship with exposure to concentrations of hydrocarbons at [the fathers] work,” as corresponding with the likelihood of a deletion occurring (Cassidy, 1989). However, more research is needed to prove significance so that precautionary measures can be implemented to minimize the possible risks.

Maternal or Uniparental disomy (UPD) accounts for twenty-five percent of documented PWS cases. In these cases, the fetus typically starts out with an extra fifteenth chromosome (trisomy 15), with two of them being from the mother. One of the chromosomes will be lost during development, and if it is the fathers, then the child will be born with PWS due to the genomic imprinting that causes the mothers chromosomes to be “turned off” in this region. In this case, the person does not have a normally functioning fifteenth chromosome since the mothers chromosome is turned off in this area and the fathers was lost instead of the second one from the mothers (Prader-Willi Syndrome Association – (USA), 2007).

An imprinting defect has only been found in five percent of documented PWS cases. This is the only cause associated with a couple's production of multiple children with PWS, both twins and siblings alike. In fact, with this imprinting defect, a sibling or twin has up to a fifty-percent chance of also having PWS (Kennerknecht, 1992). An imprinting center on chromosome 15 in the same area as the PWS critical genes and this center controls the genes of the chromosome. A microdeletion or other defect in the imprinting control center causes the chromosome from the father not to function normally. This defect can appear suddenly at any point in development. However, it is sometimes present on the father's maternal fifteenth chromosome and was "turned off," but then passed on to his offspring. This is the only cause of PWS that can be predicted, in some cases, through a genetic analysis of the fathers DNA. The defect in the fathers chromosome can be found before the child is conceived, but is so rare that it is not a common concern (Prader-Willi Syndrome Association – (USA), 2007).

Diagnosis of Prader-Willi Syndrome

There are currently two different methods that are typically used together in order to diagnose a child with PWS. These include clinical and genetic diagnosis. Both are effective and reliable. The use of both methods has a higher concordance rate with accurate diagnosis than either clinical or genetic diagnosis separately. Using both methods also helps determine which symptoms the individual has and can aid in planning therapy: medical, behavioral, and psychological.

Clinical diagnosis of PWS has been in practice since the discovery of PWS in 1956. The method that is currently used was developed in 1993 and has proven to be

accurate (Gunay-Aygun, et al., 2001). The diagnosis scale⁴ contains symptoms that are differentiated into major, and minor criteria and the diagnosis is based on a point system. Each major criteria counts for one point, and each minor criteria counts for ½ of a point. The scale is also differentiated by the age of the person; children under three need at least five points coming from any combination of the major and minor criteria, and those over three need eight points, with five of them coming from the major criteria list. The scale also includes a supportive findings section, which includes common symptoms that people with PWS exhibit, but do not contribute to the accumulation of points for diagnosis (Butler, Cassidy, Greenswag, Hanchett, Holm, Whitman, et al, 1993).

Genetic testing is almost uniformly necessary for a true diagnosis of PWS. The DNA-based methylation test that detects abnormal parent-specific imprinting within the PWS critical region is most commonly used, and accurate. It detects more than ninety-nine percent of documented affected individuals. Three other genetic testing procedures are also used. These procedures include: a FISH/Quantitative PCR that accurately identifies seventy percent of affected individuals (it only detects people affected by deletion), a uniparental disomy study that accurately identifies twenty-five percent of affected individuals (it only detects people affected by uniparental disomy), and a sequence analysis, which is the only accurate method for detecting a person with PWS due to an imprinting center defect.

The most important aspect of diagnosis for people with PWS is for it to be early. The earlier the diagnosis, the better off the child/person and family will be. Although there is not a cure for PWS, there are therapies such as growth hormone therapy that can greatly aid in healthy development (Myers, et al, 2006). Research has shown that

⁴ The diagnosis scale is located in Appendix A.

children diagnosed at a young age are significantly less obese, healthier, and happier than those diagnosed at an older age. A study of members in the Prader-Willi Syndrome Association of New Zealand found that all members who were diagnosed at age eleven or older were clinically obese, which was a substantially larger percentage than those who were diagnosed younger than age 11 (Dawson, & Thornton, 1990).

Early diagnosis is only possible when there is a doctor, parent, or other professional who is aware of the syndrome and knows where to look and how to obtain resources for diagnosis and care. Therefore, the most necessary initiative within the IPWSO right now is spreading the organization into all countries in order to provide inclusion into the program. This allows for people with PWS to have their basic medical needs handled, which is the first and foremost step in providing a better quality of life for people with PWS and their families (Eisen, interview, March 2007).

Manifestation of Prader-Willi Syndrome

PWS has many manifestations, or ways that it is revealed in a person. The genetic abnormality manifests in abnormal brain function, which in turn, manifest in cognitive, physiological, and behavioral abnormalities. This section is not exhaustive of the manifestations of PWS.

- *Brain function:* The most prominent symptoms of PWS are connected with dysfunction of the hypothalamus, which controls numerous aspects of human functioning. The hypothalamus is a “forebrain structure near the base of the brain just ventral to the thalamus” (Kalat, 2007, p.318). Hypothalamic dysfunction accounts for the majority of typical manifestations of PWS discussed in this section as well as many others. These include decreased growth hormone, altered reproductive hormones,

abnormal emotional expression, altered regulation of the autonomic nervous system, which impacts temperature, water balance, and sleep pattern regulation, and auditory short term memory (Dorn, & Goff, 2003). The rest of this section describes, in moderate detail, some of the most prominent manifestations.

- *Cognitive:* One of the most typifying characteristics of a person with PWS is mild-severe mental retardation. Mild mental retardation is the most common in people with PWS, and severe mental retardation is extremely rare (Borghraef, et al, 1990). Mental retardation is distinguished in four categories, including mild, moderate, severe and profound. The majority of people with PWS, and eighty-five percent of people with mental retardation in general have mild mental retardation, signified by IQ's between 55 and 70. Moderate mental retardation is the second most prominent level found in people with PWS and accounts for ten percent of people with mental retardation in general. It is signified by IQ levels between 40 and 55. Severe mental retardation is uncommon in people with PWS, unless there are other disabilities/diagnosis interacting with PWS. This level of mental retardation accounts for three to four percent of all people diagnosed with mental retardation and is indicted by IQ levels between 25 and 40. Profound mental retardation only accounts for 1-2 percent of the population of people with mental retardation and is signified by IQ levels below 25 (Woods, 1997).

Learning disabilities are prevalent in most PWS cases. The degree of ability varies depending on the individual, but the types of learning disabilities are typically similar across the board. Depending on the severity of the child's symptoms and school system, children with PWS are usually classified as receiving special education, although some are in self-contained classrooms, some are in resource rooms, and some spend the whole

day in mainstream classrooms and are fully included – typically with an aid. According to “The Student with Prader-Willi Syndrome: Information for Educators,” published by the PWSA (USA), and the PWSA of Wisconsin, learning strengths include long term memory skills, receptive language, savant-type skills with puzzles, basic math skills and visual learning. Their weaknesses typically include short-term memory, expressive language, fine and gross motor skills, and sequential processing deficits – difficulty understanding abstract concepts. Interestingly, what people with PWS lack in short-term memory, they tend to make up for with long-term memory. There have not been studies on this particular aspect, but it has been noted that people with PWS have abnormally extensive long-term memory capabilities.

- *Physiological:* The most telling sign of a person with PWS is their insatiable hunger. Although the exact cause for this is unknown – it is known that parts of the hypothalamus, specifically in the lateral and ventromedial areas, and multiple hormones have something to do with this effect. Research has shown a connection with ghrelin, which is a peptide that is related to food deprivation. Ghrelin levels are typically decreased when eating occurs. People with PWS continue to have high ghrelin levels even when they overeat. Therefore, it has been hypothesized that people with PWS may have an inability to turn off ghrelin release, which may contribute to the insatiable hunger that people with PWS feel (Kalat, 2007). Researchers, specifically in the weight-loss industry are continuing to study this aspect of the syndrome; there is currently no cure for the insatiable hunger (Funai, 2007).

It does not matter how much the individual eats, what time of day it is, or what else they are doing – they feel hungry. This manifestation of the syndrome is central to

many of the behaviors that are apparent in people with PWS, such as food seeking and stealing, as well as eating products that are not edible (Dorn, & Goff, 2003). Families and caregivers must make many adaptations in order to accommodate for these behaviors. Behavioral therapy does help to decrease the feelings of hunger in most cases; however, the hunger is not psychological and is impossible to “treat” until further research pinpoints the exact reason for the hunger and ways to suppress it.

Low muscle tone and short stature are apparent to varying degrees in people with PWS, and are both due to decreased growth hormone. Low muscle tone contributes to difficulties with gross and fine motor skills and also impacts the body’s caloric need. Muscle mass helps the body to burn calories; the less mass, the less calories are needed for sustenance and the less calories needed for weight gain (Dorn, & Goff, 2003). Beverly Ekaitis, a dietician technician at The Children’s Institute of Pittsburgh in order to help parents, engineered a Prader-Willi Syndrome-specific food pyramid and caregivers understand what to feed a person with PWS. While the food pyramid contains the same six food groups that are in the normal food pyramid, the positions on the pyramid and number of servings for the food groups are altered in order to provide a low calorie and nutritious diet. A typical diet for a person with PWS consists of eight hundred to twelve-hundred calories; their caloric needs are about sixty percent of those without PWS. In order to lose weight, which is often needed, caloric intake can be reduced as low as six-hundred calories per day (Ekaitis, 2007).

As a measure of reference, say you were to go to Chik-fil-a for lunch one day and ordered a char-grilled chicken sandwich with no condiments, a small order of waffle fries, and a medium lemonade. That meal constitutes seven-hundred and sixty calories.

After that meal, a person with PWS who is on a restricted diet of one-thousand calories per day could have one medium-sized apple that has around one-hundred calories, a bag of celery with low-fat dressing that has thirty calories, and one-hundred calorie pack of crackers. This comes to a total of nine-hundred and ninety calories for that day, so half of a strawberry could be added for that last ten calories.⁵ Counting calories and making sure to provide proper nutrition within the limited caloric intake is a life-or-death chore for people with PWS and their families and/or care-givers. The combination of insatiable hunger and a decreased caloric need can be a tough combination to handle.

- *Behavioral:* Behavioral manifestations of PWS are apparent in all people with the syndrome and are similar to cognitive manifestations in the respect that they are apparent on a wide spectrum depending on the individual. Some of the more prominent behavioral manifestations are skin picking, perseveration, and tenuous emotional control (Dorn, 2002).

People with PWS typically have a lower pain threshold and will pick at their skin without realizing how much they are hurting themselves. This behavior has the potential to lead to severe medical risks (Kellerman, 2002). The best way to prevent this from occurring is by watching the individual and re-directing their attention when they start to pick. Extra-curricular activities with positive social interactions are a great distraction from this behavior.

Perseveration is common in people with PWS. It is described by Barbara Dorn (2002) as a “tendency to get caught on one issue or thought to the point where it overshadows the main theme of the learning or social event.” A person with PWS may

⁵ This is just an example and not necessarily realistic as to what a person with PWS would eat in a day. Most people with PWS are on very strict diets that allow them to eat at least three meals a day and receive proper nutrition within their limited caloric intake.

repeatedly ask a question, and/or have trouble transitioning from one activity to another, or adapting to new or unexpected circumstance. There are many ways to manage this behavior. Ignoring, using reflection or going over the situation, having schedules in writing, and having specific time-lines are all helpful (Dorn, 2002).

Tenuous emotional control describes behaviors such as tantrums that signify that the person with PWS has lost emotional control for the moment. The important thing to remember with these emotional outbursts is that the person with PWS has little control over this behavior; it is part of their genetic make-up. Reasoning is typically lost during these episodes and there are numerous suggested strategies for dealing with this behavior (Dorn, 2002). Since these behaviors and the best way to deal with them varies greatly depending on the person, it is recommended to discuss this behavior manifestation carefully with the parent, care-giver or medical professional who is familiar with the person with PWS before deciding on the best strategy.

While some of these behaviors can be tough to handle, the most important aspect to remember is that these behaviors are not optional for the person with PWS. Therapy and other methods can be used to lessen the impact of these manifestations on their behavior; becoming angry at the person with PWS is not the solution. Positive, inclusive environments have been correlated with a decrease in negative behaviors within the PWS population, which makes it even more important for them to be included in positive social environments (Jackson, 2001).

V. Project #1: “Putting the Pieces Together: Understanding Prader-Willi Syndrome.”

Introduction

Knowledge is an integral part in promoting acceptance; ignorance is the key to segregation and rejection of those who are different. People are automatically inclined towards those with whom they share traits. Children shy away from others who they do not understand; it is natural to become scared. Studies have shown that “merely placing students with disabilities into mainstream environments will not guarantee that these students will be more accepted by their peers;” (Carter, & Kemp, 2002, 392) education is the key to providing acceptance. (Gresham, 1984; Sale & Carey, 1995) Educating children about the syndrome brings it out of the unknown, lifts fear, and increases the chances for acceptance. Without education, ignorance will breed exclusion and discrimination.

This section describes an innovative program called “Putting the Pieces Together, Understanding Prader-Willi Syndrome” that focuses on educating non-PWS children about the syndrome so that they will be more likely to accept children with PWS into mainstream extra-curricular activities. First, two case studies that provide a glimpse into the importance of this project are included, followed by an explanation of the need for this project, and finally the proposed solution is detailed. Additional information is provided in the Appendices.

This project proposal will be presented to both the IPWSO, as well as the PWSA (USA) with the intention of being adopted for publication by one or both of them and adapted for their specific needs. This would allow for widespread use of the program and

an avenue to market it through. If this program is adopted by the IPWSO and/or the PWSA (USA), it will be available on their on-line publication stores and available for the public. There will likely be a cost or suggested donation associated with the program in order to allow the IPWSO or PWSA(USA) to cover the cost for the materials. This program could also be made available on-line in order to cut the cost for all parties involved and increase its availability.

Case Study #1

The past two summers I worked as the Day Camp Director at Camp Krem, which is a residential camp for people with disabilities. The Day Camp program was an integration program in which children both with and without disabilities learned and grew together through camp activities. I was in charge of handling all of the logistics of the program. This included handling our budget, communicating with the parents, and supervising and working with the counselors. Most importantly, my job was to design and implement a program that would enhance all of the children's lives while promoting inclusion and acceptance for everyone involved.

On my first day as the Day Camp Director in June 2005, I spent my morning greeting my campers and memorizing names as an intense argument broke out between a typically developing camper and a camper with cerebral palsy because the one with cerebral palsy took too long at the water fountain. At that point, I knew that I had to do something to educate all of the children about their differences in order to have the possibility of acceptance. After talking to the parents, I found out that the majority of the typically developing children had very little or no experience/exposure to children with

disabilities. The school district kept them in self-contained special education classrooms; mainstreaming had not been adopted in the small town of Boulder Creek, California.

I began by using arts and crafts and games that promoted diversity by demonstrating how each child was unique, yet important to the group. Although the children understood and enjoyed the activities, the majority of them were still in the pre-operational stage as described by (first name) Piaget and had trouble transposing the activity into their relationships with each other. (Piaget) The hostility between the campers continued. About three weeks into camp, I decided that something else needed to be done, my method was slowly beginning to work – but the change in behavior was not significant enough to continue solely using that method. I decided to have a special meeting with the children who had disabilities and find out what they wanted their peers to know. They helped me come up with a way to explain their condition in “kid” terms. The next day we packed picnic lunches and set out on hike to a secluded spot surrounded by redwood trees where we could comfortably get away from the hustle-and-bustle of the residential camp. After eating lunch, each of the children who had a disability helped me educate their fellow campers on their disability, highlighting their abilities and the areas/things that they needed help with. The campers were allowed to ask questions and it provided for a great dynamic and learning experience for all involved. This beautiful experience in the Red Wood trees was a turning point for the lives of all of the children, and counselors involved.

We continued to have a policy of openness and education within the program and I witnessed the core group of children grow as caring people throughout the summer. The summer started with a lot of hostility between the children who had disabilities and

the children who were typically developing. However, through all of the integrative and educational activities that we did, the kids all grew to accept and love each other. The typically developing kids went from making fun of and avoiding the children with disabilities to accepting, helping and sticking up for them. It was an amazing transformation, and I know that our future generations will benefit greatly if all children are given this experience. (journal).

Case Study #2⁶

A young boy with PWS, Bill (not his real name) was in a mainstream class, where he was fully included into classroom activities with the help of an aid. The majority of the children in the classroom would not interact with him and he got made fun of a lot for his abnormal behaviors. A few weeks into the school year, Bill and his aid agreed that he would leave the room for a while and she would educate the other children about his syndrome. Bill helped his aid decide what to discuss and made sure that he was comfortable with everything that she would say. He was especially interested in knowing how many of the kids thought that he had a disability.

On the appointed day, the aid went into the class prepared to explain PWS to the children with hopes that understanding the syndrome would increase acceptance for Bill. She found out that less than forty per-cent of the students actually thought that Bill had a disability. They just thought that his abnormal behaviors (temper-tantrums, lack of certain social skills, skin-picking, etc.) were his fault and that he was just trying to be annoying and frustrating to them. The aid educated the children on PWS, and then Bill returned to class the following day.*

⁶ Provided by: Prader-Willi Syndrome Association (USA), & Prader-Willi California Foundation, 2003.

Bill noticed that he was much more accepted by his peers following the PWS education day! He told his mom that students who had never talked to him before actually stopped to say “Hi” in the hallway, and that the other students were much more friendly and willing to work with him and play with him throughout the day. School was now a much better experience for him, and the other children who were learning to accept those who are different.

Explanation of the Need in the United States

As these case studies have demonstrated, not only does education about PWS help the people directly affected by the syndrome, but also promotes acceptance and understanding of diversity in all children who participate in the inclusive environment. Promoting this acceptance of diversity in young children is a stepping-stone to a more accepting future for the general public, as well as people affected by PWS – both the individual and their families. Research has shown that people with PWS thrive in situations where positive interaction is practiced and where “their social environment is a positive one.” (Jackson, K., 2001) This concept is universal. Providing positive interactions in inclusive social environments are beneficial to all involved

Studies have shown that parents of children with PWS think that educating the public about PWS is an important endeavor in the struggle to obtain a better quality of life for their children. (Dawson, K., & Thornton, L., 1990) Knowledge sparks acceptance; acceptance allows for a positive inclusive environment. Most parents are deterred from enrolling their children in “mainstream” extracurricular activities due to negative attitudes that lead to the unwillingness of extracurricular programs to accept children with disabilities – especially a syndrome that is rare and has aspects that are very

different from the majority of developmental and physical disabilities. According to Pamela Eisen, president of the IPWSO, one of the scariest and most dissuading aspects of inclusion of children with PWS into mainstream activities for the families, and the affected child is the fear that the other children will not accept them and that a negative experience will result. There are wonderful pamphlets, articles and even videos about PWS that describe how to include children with PWS in the mainstream education system, but none of these aim specifically at educating other children about the syndrome.

The IPWSO has designated a need for spreading awareness of PWS in general. There is also a need for literature and programs designed to educate children and adults about the benefits for everyone involved of including children with PWS in extra-curricular activities. There have been some programs implemented through the PWSA-USA, however, they are limited. Using the research previously described, personal experiences, and IPWSO and PWSA-USA resources, I have designed a program for educating children about PWS.

“Putting the Pieces Together: Understanding Prader-Willi Syndrome”

“Putting the Pieces Together: Understanding Prader-Willi Syndrome,” is a program that I have designed to educate elementary-aged children in mainstream extra-curricular activities about PWS in order to promote acceptance of children with PWS into these much-needed activities and social interactions. It can also be used in a classroom or in other environments where children with PWS will be included with both typically developing children and/or children with different disabilities. The development of this

program has taken all of the literature about PWS, the authors personal experiences, interviews and research related to educating children into account.

This program acts to benefit all systems involved, including: the individual with PWS, the children without PWS, the families of all of the children involved, the instructor/teacher and the organization. A heightened awareness of PWS and an understanding of the syndrome will enable all of the children involved to include and befriend the child with PWS, which will aid in social development for all children involved. The child with PWS will be provided with a caring, understanding and open environment and the children without PWS will realize their potential for accepting diversity and the positive outcome that stems from that concept. The families will benefit from the positive experiences that their children are having and the instructor/teacher of the group being offered this program will benefit from their own increased awareness about PWS, but also from fundamental concept that this program promotes about including people of all abilities into activities. There is a need for inclusive extra-curricular programs and organizations that adopt the inclusion model for their activities will benefit from the extra business, as well as the overall higher quality of their services for the children with regards to developing them as a whole child.

The program is designed so that any responsible adult can read the background information previously displayed, and have enough of an understanding about PWS to educate children using the materials provided. The responsible adult who will be leading this program is referred to as “the leader” in the text. The leader of this program can be a parent, extra-curricular instructor, or teacher, a sibling of someone with PWS, or a school teacher, and the suggested audience is between five and thirty children. This program

will most likely be used when a child with PWS is about to join into a group through an extra-curricular activity or enter into a mainstream classroom. If there are more than thirty children, it is suggested to split the group up into sections and have multiple leaders if possible. The previous portions of this thesis, along with links to interesting articles about PWS and tips for public speaking and educating children will be included along with the presentation outline will be included in order to provide the presentation leader with moderately in-depth information, as well as the reasoning behind this program. The presentation is entitled “Putting the Pieces Together: Understanding Prader-Willi Syndrome,” with the theme stemming from the typical increased ability with puzzles of people with PWS. This theme allows for a specific ability of people with PWS to be focused on, rather than a disability. The focus of the program is geared towards *abilities* and the positive aspects of engaging in a friendship with a child with PWS. A letter to the leader is included to explain the basic intention of the program, as well as an outline for the leader, including an outline of what to discuss as well as all materials needed. A power-point that corresponds with the outline is also included and can be used if the technology is available; if not, the leader can use other materials, such as hand-outs, a chalkboard, butcher paper or any other means to provide the information from the power-point for the children. The previous portions of this paper will also be included, excluding the sections pertaining to the project in Costa Rica. This will provide the leader with knowledge of the basics of PWS, as well as the importance and ideology behind this program. A list of websites and books that can be used for more in-depth information will also be included.

Materials Included in the Program

Following are descriptions of the three main materials included in this program and their purpose. Brief descriptions are given for each and they can be found in the appendix in their entirety. All of these may be subject to alterations if the program is adopted by the IPWSO and/or the PWSA(USA) in order to meet their specific needs and criteria.

- *Letter to the Leader:*⁷ The letter to the leader is an introductory letter that will be included in the materials for this program. It is a basic introduction to the program that describes the intentions for the program, a few tips for being the leader, and what materials are included. Most importantly, it displays a sincere appreciation for their time and effort. It will be the first thing that the leader will read, and therefore needs to make them feel good about what they are doing, as well as explain the basic idea behind the program and interest them in reading further and going through with the program.

- *Presentation Materials:* The presentation materials consist of a power-point presentation⁸ that can be used in order to present the information to the children, as well as an outline for the presentation leader⁹ that corresponds with the slides on the power-point presentation. The power-point is colorful, and designed to help keep the children interested in the presentation as well as provide structure. The outline provides a rough script of what the presentation leader will say, as well as descriptions of all of the interactive activities involved in the presentation.

Description of the Presentation

The presentation is composed of six main components that incorporate audio, visual and tactile learning. Each of these sections correspond with the power-point

⁷ The Letter to the Leader is located in Appendix B.

⁸ The Power-Point Presentation is located in Appendix C.

⁹ The Outline for the Presentation Leader is located in Appendix D.

presentation and the outline for the presentation leader, which are both found in the appendix. Following is a brief description of each of the six sections. Please refer to the power-points (Appendix C) and the outline for the presentation leader (Appendix D) for more in-depth information on each of the six sections.

- *Visualization:* The presentation begins with a visualization exercise, in which the presentation leader reads a script of a day in the life of a child with PWS. The visualization script is provided for the presentation leader in the outline.
- *Prader-Willi What?:* This section includes an elementary description of PWS in order to provide the participating children with enough information to help them understand why some people have PWS and what it means to have PWS.
- *Puzzle Time:* This activity provides a visual representation of the moral of the presentation for the participating children. The presentation leader is instructed to take a poster board, or something of that nature and cut it up into enough jig-saw puzzles for everyone in the group to get one, prior to the presentation. Participating children are asked to decorate their puzzle piece in a way that describes them. After they are finished, the puzzle is put together to form a whole. Following this activity are discussion questions. These questions work to have the children discover, themselves, that everyone is different but equally important and necessary in a group.
- *Some Things to Remember...:* This section describes how people with PWS are definitely capable of being wonderful friends. It also gives the children some common scenarios that may happen when interacting with a friend who has PWS and ways to best handle those situations.
- *Rap-a-Long:* This section includes a rap, written by Tad Tomaseski who is a

youth minister in Frisco, Texas, along with his youth leaders, and teen group. They formed a musical group named “Xcel,” and wrote and recorded “My Name’s Not Willy©” with the purpose of boosting self-esteem in people with PWS at a PWSA (USA) convention in 2003. It is from the perspective of a person with PWS. Different activities are included for the presentation leader to do with this rap in order to further enhance the educational aspects of it. One of these activities is a freeze dance game in which everyone has to freeze when the rap is turned off and whoever freezes first or last (it is up to the presentation leader), gets to describe what the rap is talking about. This activity promotes careful listening and interpretation on the part of the child, which will help them to get the most out of the rap.

- *Questions?:* The presentation wraps up with a question section, in which the children are reminding that it is always alright to have questions and that their presentation leader will always answer the questions that they can, or find out the answer if they do not know. Multiple materials and references will be provided for the presentation leader so that they can find out answers that they do not know.

Future Work

Although this program is well developed, there is always room for future work and there is a lot that can be done to improve upon it. First, the program needs to be presented to the IPWS and the PWSA(USA) and adapted to fit their exact needs so that it can be made available to the public. Other possibilities for future work include adapting the program for different age groups and environments, and making the program into an interactive DVD that can include adapted presentations for different age groups and environments. Having a DVD would make the program more accessible for the public

and less time consuming for the presentation leader. Lastly, running a test to determine if the program positively correlates with an increase of acceptance among peers for children with PWS and a higher rate of acceptance into mainstream extra-curricular activities is necessary in order to determine the validity of the program.

IV. Project #2 “Starting a Chapter of the International Prader-Willi Syndrome Organization in Costa Rica”

Personal Story – Family in Costa Rica:

When I arrived in Costa Rica for a study abroad program in October of 2006, I expected several challenges: a new language, a new culture, and a new living arrangement. However, I could not foresee the intense challenge that I would embark upon three weeks into my experience. The challenge was to lay the groundwork for a new chapter of the International Prader-Willi Syndrome Organization (IPWSO) in Costa Rica at the request of my aunt, Pamela Eisen, the president of IPWSO. Shortly before I arrived in Costa Rica, a mother who suspected that her daughter had Prader-Willi Syndrome (PWS) contacted the IPWSO. The family was desperate for assistance from the organization.

I was excited about taking on this new challenge, but faced the obstacles of communicating proficiently in Spanish and having no idea where to begin with this daunting task. After performing extensive research, I learned that forming a chapter of IPWSO requires both a family affected by this syndrome and an interested doctor to sponsor the organization. My challenge was to establish a network of interested individuals while I was in Costa Rica so I could continue working on the project after returning to the United States.

I began corresponding with the family via e-mail while awaiting a chance to meet them in person. E-mail made communicating in Spanish much easier. I could look up unfamiliar words and have a Spanish professor proofread my messages before sending them to the family. For potentially interested doctors, I compiled a packet of literature

about PWS in Spanish, and wrote a cover letter explaining the goal of starting a chapter of IPWSO in Costa Rica.

During December 2006, I finally had the chance to meet with the family. I planned to have a friend who was fluent in Spanish go with me; however she became ill and was not able to attend. I was forced to step out of my comfort zone and really test everything that I had learned in three months of intensive Spanish classes. I successfully conducted an interview of questions provided by IPWSO, and spent the whole day interacting with their family solely in Spanish. They had never met anyone who knew someone with PWS before, and they were ecstatic and relieved to have the opportunity to talk to someone who understood their experiences.

Explanation of the need in Costa Rica

I am currently in contact with the Costa Rican family, prospective doctors, and IPWSO. We are expecting to have the Costa Rican chapter of IPWSO started before 2008. The girl that was suspected of having Prader-Willi Syndrome; however, the results came back negative. She is now being tested for Bardet-Biedl Syndrome, which is a rare disorder that has similar characteristics to and is sometimes mistaken for Prader-Willi Syndrome when relying solely on a clinical diagnosis. This obstacle and the difficulties with the diagnosis have lengthened the amount of time that it is taking to start an organization in Costa Rica. The IPWSO allows people who are affected by other rare disorders to join their organization and work together with other organizations to help the affected individuals and families. The family and the IPWSO are still interested in starting a chapter of the IPWSO in Costa Rica to help both this family, as well as other families who are affected by PWS in Costa Rica.

There are most likely between fifty and two-hundred families unknowingly living with a person who has PWS in Costa Rica based on their national population and the estimations of prevalence of PWS and the fact that PWS is spread across all races, nationalities, etc. None of the doctors that we have contacted know of a child with PWS, and the majority of them were not familiar enough with the syndrome to describe it accurately. Tools for diagnosis of PWS are not currently available in Costa Rica, and the family that I met had to send the information to Italy in order to receive the diagnosis. Currently, the IPWSO and myself are searching for a doctor that is familiar with, or willing to become familiar with both Bardet-Biedl, and PWS in order to help the family, as well as establish a chapter of the IPWSO. Establishing a chapter of the IPWSO in Costa Rica will provide resources for medical professionals, as well as all other systems involved, such as families and schools, in order to raise awareness of the syndrome so that affected individuals can be diagnosed and receive proper medical, educational and social opportunities.

Costa Rica: Cultural and Communication Analysis

Understanding the communication and cultural patterns/differences apparent in a specific culture is of utmost importance when striving for successful communication, both interpersonally and organizationally. Numerous factors go into the communication and cultural patterns of a culture and these patterns, in turn, have a large effect on the culture; it is a continuous cycle. In order to start an organization in Costa Rica, it is vital to understand these patterns and the mediums through which they are transferred. Although the IPWSO is an international organization and organized in order to include countries from around the world, it is still of utmost importance to truly understand the

culture of each individual country in order to adapt the process of implementing and running the organization to the countries needs and provide the most impact.

Jennifer Chandler, a Spanish-English translator from Georgia stated that knowledge of these communication patterns “are very important in [her] work,” and that she often refers to them when “deciphering and translating messages for people from different cultures so that the true meaning of the message is conveyed and noise from cultural barriers do not interfere” (Chandler, J., interview, November 21, 2006). Many people do not think of these non-linguistic differences and barriers and many businesses, and organizations have trouble expanding internationally due to problems with this part of translation and adaptation.

This section explores the specific communication and cultural patterns that are apparent in Costa Rica. The majority of cultural patterns in Costa Rica are generalizable for Latin American countries; however, there are intricacies specific to Costa Rica. Edward T. Hall’s (1977) dimension of high vs. low context culture and Geert Hofstede’s (1991) five dimensions of cultural diversity are the most universally accepted theories of communication/cultural patterns and taking both of their perspectives together provides an expansive understanding of Costa Rican culture. Both of these scales are also generalizations of an over-arching cultural opinion in a country, and not necessarily accurate of each individual in a culture.

- *High Context/Low Context Cultures:* Edward T. Hall devised the continuum of high vs. low context culture, which promotes his theory that every person is faced with so many perceptual stimuli that it is impossible for them to pay attention to them all and therefore, their culture provides a screen that helps people decide what to notice and how

to interpret their observations. On this continuum, Costa Rica is ranked as a high-context culture. High context cultures are defined as preferring to communicate via high-context messages which puts the emphasis of their communication in the realm of non-verbal messages. The majority of messages are interpreted through the listeners perceptions of the physical setting, and their own predetermined internal beliefs, values and norms. Low context cultures prefer to use low-context messages in which the majority of the information is coded explicitly in the messages that are presented. Interactions with computers are categorized as being very low-context due to the necessity that computers have for detailed, precise messages that do not leave information to be presumed, while intimate relationships in any culture are typically very high-context. Intimate partners can usually sense and understand each other without much verbal communication and non-verbal messages become increasingly more important, even in the most low-context of cultures (Hall, 1977).

- *Uncertainty Avoidance*: Geert Hofstede, a professor from Denmark, based his dimensions for explaining the differences between cultural patterns on the assumption that people carry mental programs that develop during childhood and are implemented and reinforced through culture. The uncertainty avoidance scale measures how cultures adapt to changes and cope with uncertainties and/or ambiguities. The degree of uncertainty avoidance is very important within a culture and their communication due to the wide variety of unknown aspects within daily life and thoughts of the future. Costa Rica is ranked high on the uncertainty avoidance scale, which is different than the majority of Latin American countries. Most Latin America countries are ranked very low

on this scale, which means that they are more accepting of uncertainty and risk (ITIM International, 2006).

- *Individualism/Collectivism:* The individualism vs. collectivism scale measures the degree to which individuals are integrated into groups. On the individualist side, societies have looser ties between individuals and everyone is expected to look after him/herself and his/her immediate family. On the collectivist side societies tend to have very strong ties with cohesive in-groups, which are primarily comprised of family, including extended family. However, these in-groups of individuals can also be comprised of groups of friends or co-workers. The individual is more important in an individualistic society and the group is more important in the collectivistic societies (Hofstede, 1991).

Overall, this scale is the one most scholars have turned to when analyzing the differences between Latin Americans and North Americans, specifically within their communication patterns within the workplace. Costa Rica, and all countries in Latin America are ranked as collectivistic cultures and the United States is ranked as the most individualistic culture on Hofstede's scale (ITIM International, 1987-2003).

- *Description of Power Distance:* Hofstede's dimension of power distance focuses on the common concern of human equality/inequality that all cultures deal with. Power distance describes the amount of distance between people in power and/or in the upper class and the other members of a society. The dimension of power distance also takes into account the way in which a culture values or believes in inequality – whether they consider inequality to be positive or negative, right or wrong, fair or unfair. Power distance reflects the degree to which the culture believes that institutional and

organizational power should be distributed equally and how much the decisions of the power holders should be challenged or accepted (Lustig and Koester, 2006).

Costa Rica is considerably low on the power distance scale. This means that Costa Ricans believe in the importance of minimizing social class inequalities, questioning or challenging authority figures, reducing hierarchical organizational structures and using power only for legitimate purposes. Hierarchy and inequality are not well tolerated in Costa Rica.

- *Description of Masculinity vs. Femininity:* Hofstede's scale of masculinity versus femininity uses these two words in a symbolic way to describe behaviors of a culture that are more stereotypically masculine, or feminine. Cultures that are ranked towards the masculine side are more achievement based, and those that are ranked as more feminine are more nurturance based. Masculine cultures tend to "believe in achievement, and ambition in judging people on the basis of their performance and in the right to display the material goods that they have acquired. Feminine cultures tend to focus more on the intrinsic aspects of the quality of life, such as service to others and empathy for the unfortunate. Costa Rica ranked as a feminine culture on Hofstede's scale (Lustig and Koester, 2006).

Understanding these cultural and communication patterns in Costa Rica allows for the adaptation of strategies in order to effectively and successfully communicate with Costa Ricans about starting a chapter of the IPWSO. Knowing that the culture is ranked as being high context, collectivistic, uncertainty avoidant, low in power distance, and feminine allows for many assumptions to be made concerning the best way to discuss the benefits of this organization with families, and medical professionals. For example,

stressing that the organization is designed for collaboration between all members in order to aid in the development of the individuals affected by PWS and their families so that they have a less uncertain future due to the knowledge of the syndrome that they will have access to will be very important. This idea stresses the in-group, nurturing and low power-distance nature of the program as well as its ability to limit uncertainty.

- *Overview of Communication Technology:* Not only is knowledge of cultural and communication patterns important, but so is knowledge of the preferred mediums through which communication is transmitted. The field of communication technology research is a relatively new area of communication studies that explores how different communication mediums are used, and how they affect interpersonal, group and public communication. Of particular interest in this field are computer-mediated-communication (CMC) and telecommunications. As our society advances in technology, we continue to have more options for communicating with others. Each medium for communication has different qualities that make them better for certain types of messages than others, as well as more or less preferred by different cultures. This phenomenon has generally been explained in the communication studies field through the media richness theory which defines the richness of a communication medium in terms of its ability to reduce uncertainty and equivocality. Within the three main mediums of communication, face-to-face communication is the most “rich” form of communication with telecommunications coming next, and CMC (computer-mediated-communication) coming last (Salem, P., personal communication, February 2005).

Communication technology is bringing the world closer; or is it? We can call, e-mail, or chat on-line with a person who is sixteen hours ahead of us in time, in a different

season, around the globe, or even just next door without leaving our homes, or office. Computers can even cross language barriers with online translating services. The basic assumption about communication technology, is that people from all over the world are being brought together by it, which is greatly supported by the recent giant influx of globalization. However, we often overlook how the communication/cultural patterns in different cultures socially construct people's perceptions and preferences for communication mediums. It has been researched and proven that part of being a professional translator is knowing the culture well enough to be able to translate a message correctly, while also translating it in a manner that is acceptable and understandable in the other culture. Likewise, part of using communication technology is remembering that it "is always understood in cultural contexts and that communication socially constructs perceptions of technology" (Leonardi, 2002). Therefore, it is important to not only understand the communication and cultural patterns that effect a cultures perceptions, but also their feelings and thoughts about different mediums of communication.

- *Media Richness Theory*: The media richness theory, as mentioned previously, is based on contingency theory and information processing theory, and the first proponents of this theory were configured by Draft and Lengel, two communication scholars. The two main assumptions of this theory are that people want to overcome equivocality and uncertainty in organizations and interpersonal relationships and that a variety of media commonly used work better for certain tasks than others. Using four criteria, Daft and Lengel presented a media richness hierarchy, arranged from high to low degrees of richness, to illustrate the capacity of media types to process ambiguous communication in

organizations. The criteria are (a) the availability of instant feedback; (b) the capacity of the medium to transmit multiple cues such as body language, voice tone, and inflection; (c) the use of natural language; and (d) the personal focus of the medium. Face-to-Face communication is the richest communication, followed by telephone and then electronic mail, with mediums such as fliers and bulletins holding rank as the least “rich.” (Daft and Lengel, 1984)

The “richness” of the medium directly affects how people from different cultures perceive and prefer different mediums of communication. There has been a small amount of research on how “rich” people from different cultures rate mediums of communication, and it was found that people from low-context cultures, such as the United States rank telephone communication as being very high on the “richness” scale, whereas people from Latin American backgrounds who are more high-context rank telephone communication much lower on the “richness scale.” (Rice, D’Ambra, More, 1998) Some nonverbal cues are transmitted over the phone, most specifically in the category of paralanguage with a small amount of cues in the categories of chronemics and sensorics, however, the majority of the nonverbal cues in a message are lost through this medium of communication. (Lizano, 2006 (2)). High-Context cultures rely more on nonverbal cues than low-context cultures, which explains why low-context cultures are more likely to rate telephone communication as being more “rich” than those from high-context cultures.

CASE STUDY #1

Paul M. Leonardi, a graduate student at the University of Boulder has done extensive research in how first generation Latin Americans socially constructed

perceptions of communication technology in the workplace. His research was qualitative rather than empirical, and therefore relied solely on focus group discussions that he organized with a group of first generation working class Latino Americans. The qualitative nature of this study allows us to view what the participants actually said and thought about the different communication mediums and, therefore, understand the reasoning behind their feelings towards these communication mediums. This aspect of the study was imperative in determining that their preferences and ideas about the different communication technology mediums had to do with cultural values, specifically collectivistic cultural values.

Leonardi hypothesized and found that the collectivistic culture of first generation United States Latinos affected their perceptions and uses of cell phones, the computer and the internet. The mainstream culture in the United States views cell phones, the computer and the internet as methods of enhancing communication. However, this study found that out of these three mediums, first generation United States Latinos only consider the cell phone as enhancing cultural communicative values and the computer and internet as inhibiting these values. Close interpersonal contact is very important in collectivistic cultures, especially with members of their in-groups (family, close friends, long-term work partners, etc.) and therefore, the cell phone was regarded as allowing them to keep this interpersonal contact with their family members at all times, especially as they were adjusting to the typical United States lifestyle in which family time is not as important, or easy to fit into the schedule when they were having to work all of the time to make enough money to live in the United States. The participants in this study almost unanimously agreed that computer-mediated-communication is not beneficial for

interpersonal relationships. They viewed computers and the internet as “promoting individuality and removing the user from social life,” and therefore felt that this medium for communication is putting their personal/cultural values into jeopardy. Leonardi found a direct correlation between the collectivist culture values that this group of Latin Americans had with their preferences for communication technology (Leonardi, 2003).

Case Study #2

In order to relate this topic specifically to Costa Rica and the United States, I devised a short survey¹⁰ and had ten Costa Rican University students and ten students from the United States, all studying at Veritas University in Zapote, Costa Rica. All of them stated that they have easy access to both phones, cell phones and the internet. However, the United States American students were asked to fill this survey out as if they were at home, in the United States because the majority of them do not have cellular phones, or as much access to computers while studying in Costa Rica as they do in the United States. This study is not definitive because of the small group of participants, the short length of the survey, the fact that the survey was given in English (the second language of the Costa Rican students), both groups of students are minorities (University students in an elite art and architecture school, and students who are studying abroad), the questionnaire was given in the cafeteria while the students were surrounded by friends and had a lot of distractions, and because the actual questions in the questionnaire have not been tested to see if the answers definitely signify the meanings that I have attributed to them. However, it is a good basic example of how Costa Rican and United States culture differ in their preferences for certain communication technology mediums in different situations.

¹⁰ The survey and results are located in Appendix E.

Due to the specific communication/cultural pattern differences between Costa Rica and the United States and the research that was just cited, I predicted that the Costa Rican students would prefer telephone communication over the internet. Based on my observations of the United States American college-level students, I predicted that they would prefer to use computer-mediated-communication or text messaging over the phone. I think this is a growing phenomenon in the United States and it has even gone so far that I have observed and spoken to people who experience social anxiety about making phone calls, both for business purposes and even with their friends.

Overall, this survey did show a small, but significant difference in the answers between the Costa Rican students and the United States American students. Overall, Costa Rican students preferred using the phone and the United States American students preferred using computer-mediated-communication, or text-messaging over the phone. The results, along with the survey questions are detailed in Appendix E.

The results of this small study significantly displayed differences in the communication technology preferences between Costa Rican university students and United States American university students. Overall, Costa Rican students chose using the phone more than United States American students; however the specific situations did have a large impact on their answers. I hypothesized that cell phone use would be more popular than e-mail with the Costa Rican students due to their upbringing in a culture that is high-context, collectivistic, and uncertainty avoidant, and vice-versa with the United States American students. E-mail does not allow for nonverbal messages and is a lot less “rich” than phone and especially face-to-face communication. It also contains more uncertainty since it is impossible to make sure that a person actually receives a message,

who reads the message, and/or how the message is interpreted. These cultural/communication pattern differences definitely contribute to the differences in preferences, although there are a lot of other factors, such as availability of computer resources, that need to be explored in order to make this study conclusive. In the future, a qualitative study over this topic would be beneficial in order to examine the exact reasoning behind the students' answers so that less is left up to assumption.

Understanding the communication medium preferences is beneficial when trying to communicate with people in Costa Rica from the United States. I was having a great deal of trouble using e-mail for correspondence and now know that telecommunications will be a better way, even though it is more expensive and less convenient. I've been contacting doctors via e-mail and have realized that I either do not receive a reply, the reply is significantly delayed, or the doctor does not seem to have any interest. I did try calling one doctor and had more success and will continue to try to use telephone communication whenever possible since face-to-face communication is not possible at this point in time.

Summary

Knowledge of the previous cultural analysis of Costa Rica, and the two case studies will aid in starting a chapter of the IPWSO in numerous ways. Having a full understanding of the culture will allow the IPWSO to better adapt the chapter in Costa Rica for the culture and their specific needs. One example is the type of resources that will be valuable in Costa Rica. Costa Rica is a collectivistic culture and, therefore, children typically live with their parents until they get married. In the case of people with disabilities, it is more common for them to live at home or with family members rather

than in a group home. Therefore, research related to taking care of an adult with PWS in the household will be more valuable than research on how to choose a group home, or group home dynamics for people with PWS. Also, understanding communication technology preferences in Costa Rica will allow the IPWSO to best use their resources when looking for an interested doctor, and/or other necessary components in the future Costa Rican chapter.

Future Work

Future work for this project is basically getting a chapter of the IPWSO up and running in Costa Rica. First, the chapter needs to make sure that the family described at the beginning of this section receives an official diagnosis. Meanwhile, the search for an interested doctor needs to continue until one is found since it is necessary to have a doctor be one of the founding members of a chapter of the IPWSO. We also need to continue corresponding with the family previously described in order to help them to the best of our ability, as well as determine if at least one member of the family is willing to work towards starting a chapter in Costa Rica and serve as the family representative. On a more global perspective, only fifty-four countries are currently represented in the IPWSO. PWS has been found to be apparent in all races, ethnicities and countries and therefore, working to establish a chapter in every country around the globe, one country at a time, is definitely the long-term goal of this project.

VI. References

- Aguilar, G. (1990). Jane: A victim of Prader-Willi syndrome and the system. *Journal of Social Work, 35*(3), 279-282.
- Bray, G. et al (1983). The Prader-Willi syndrome: A study of 40 patients and a review of the literature. *Medicine, 62*(2), 59-80.
- Borghgraef, J., Fryns, J., & Van Der Berghe, H., (1990). Psychological profile and behavioral characteristics in 12 patients with Prader-Willi syndrome. *Genetic Counseling, 38*(2), 141-150.
- Butler, M., Cassidy, S., Greenswag, L., Hanchett, J., Holm, V., & Whitman, B., (1993). Prader-Willi syndrome: consensus diagnostic criteria. *Pediatrics, 91*, 398-402.
- Butler, M., & Keder, L. (2004). *Genetics of Prader-Willi syndrome: An explanation for the rest of us*. PWSA (USA): Retrieved on February 13, 2007 from http://www.pwsausa.org/syndrome/Genetics_of_PWS.htm
- Cassidy, S.B. (1989). Prader-Willi syndrome. Characteristics, management and etiology. *The Alabama Journal of Medical Sciences, 24*(2), 169-175.
- Carter, M., & Kemp, C. (2002). The social skills and social status of mainstreamed students with intellectual disabilities. *Educational Psychology, 22*(4), 391-411.
- Dawson, K., & Thornton, L. (1990). Prader-Willi syndrome in New Zealand: A survey of 36 affected people. *New Zealand Medical Journal, 103*, 97-98.
- Donaldson, M., (add all names) (1994). The Prader-Willi syndrome. *Archives of Disease in Childhood, 70*, 58-63.
- Dorn, B. (2002). Information for school staff: supporting the student who has Prader-Willi Syndrome. *The Gathered View, September – October, 2002*, 13-14.
- Dorn, B. & Goff, B. (2003). *The student with Prader-Willi syndrome: Information for educators*. PWSA (USA) and PWSA of WI, Inc. pp. 1-65
- Dykens, E., Hodapp, R., Masino, L., & Masion, L. (1997). Families of children with Prader-Willi syndrome: Stress-support and relations to child characteristics. *Journal of Autism and Developmental Disorders, 27*(1), 11-24.
- Ekaitis, B. (2007). *A Prader-Willi food pyramid*. PWSA (USA): Retrieved, March 5, 2007 from <http://www.pwausa.org/syndrome/foodpyramid.htm>.
- Funai, E. (2007) *Ghrelin, hormone that stimulates appetite, found to be higher in PWS*. PWSA (USA): Retrieved, March 14, 2007 from

<http://www.pwsausa.org/research/ghrelin.htm>.

- Gelbart, M. (1999). In our parents' shadow: Prader-Willi syndrome. *Nursing Times*, 95(2), 36.
- Greensway, L. (1987) Adults with Prader-Willi syndrome: A survey of 232 cases. *Developmental Medicine and Child Neurology*, 29(2), 145-152.
- Gunay-Aygun, M., Heeger, S., O'Riordan, M., & Schwartz, S. (2001). The changing purpose of Prader-Willi syndrome clinical diagnosis criteria and proposed revised criteria. *Pediatrics*, 108(5), p. E92.
- Hall, E. (1977). *Beyond Culture*: New York: Bentam Doubleday Dell Publishing Group, Inc.
- Hopworth, D.H., Rooney, R.H., Rooney, G.D., Strom-Gottfried, K., & Larsen, J.A. (2006). Direct Social Work Practice: Theory and Skills. Belmont, CA: Brooks/Cole.
- Hofstede, G. (1991). *Cultures and Organizations: Software of the Mind*. London: McGraw-Hill.
- International Prader-Willi Syndrome Organization website. Retrieved February 2007. <http://www.ipwso.org/>
- ITIM International (1986-2003). Geert Hofstede™ Cultural Dimensions. Retrieved October 15, 2006 from <http://www.geert-hofstede.com/>
- Jackson, K. (2001). Characterizing and managing behavior in Prader-Willi syndrome. *The Gathered View, January – February 2001*, 8-11.
- Kalat, J. (2007). *Biological Psychology: Ninth Edition*. Canada: Thomson Learning, Inc.
- Kellerman, T. (2002). Picky, picky, picky: a parent's perspective. *The Gathered View, January-February 2001*, 5.
- Kennerknecht, I. (1992). Differentiated recurrence risk estimations in the Prader-Willi syndrome. *Clinical Genetics*, 41(6), 303-308.
- Lightfoot, J., Sloper, S., & Wright, S. (1998). Supporting pupils in mainstream school with an illness or disability: Young people's views. *Child: Care, Health and Development*, 25(4), 267-283.
- Lustig, M., & Koester, J. (2006). *Intercultural Competence in Interpersonal Communication Across Cultures*. Boston, MA: Pearson Education.

- Morehead, P. (2005). *The New American Webster Handy College Dictionary: Third Edition*. New York: Penguin Books Ltd.
- Nyhan, P. (2006, September 7). A School for Hannah: Rare genetic flaw made her parents' choice a crucial one. *The Seattle Post*. (need p #)
- Pineyerd, B. (2002). *Prader-Willi syndrome as a model for obesity*. Retrieved February 24, 2007 from <http://www.pwusa.org/research/ModelforObesity.htm>.
- Prader-Willi Syndrome Association (USA). (2007) *Genetics – Chromosome 15*. Retrieved, February 5, 2007 from <http://www.pwsausa.org/syndrome/genetics>.
- Prader, A., Labhart, A. & Willi H. (1956). Ein syndrome von adipositas, kleinwuchs, kryptochismus and oligophreni nach myatonieartigem zustand in neugeborenenalter. *Schweiz Med Wochensch*, 86, 1260-1261.
- Prader-Willi syndrome: physical and neurodevelopmental benefits. *American Journal of Medical Genetics*, 143(5), 443-448.
- Prader-Willi Syndrome Association (USA), & Prader-Willi California Foundation, 2003. *Understanding the Student with Prader-Willi Syndrome: Strategies for Success*. California Prader-Willi Syndrome Association (USA), & Prader-Willi California Foundation.
- Ranathamam, R. & Asamoah, Y. (2004). Understanding the human condition and human Behavior in a global era. In C.S. Ramanathan and R.J. Link, (Eds.). *All our futures: Principles and resources for social work practice in a global era*. Belmont, CA: Brooks/Cole. pp. 30-51.
- Raab, E.T. (1986). *Understanding Praer-Willi syndrome: A literature review for educators and families*. Edina, MN: Prader-Willi Syndrome Association.
- Wett, R. (1983). Prader-Willi syndrome: the disabled child. *North Carolina Medical Journal*, 44(8), 499.
- Woods, M. (1997). *Mental retardation*. Encarta Online Encyclopedia 2007. Retrieved on February 24, 2007 from http://encarta.msn.com/encyclopedia_761552226/Mental_Retardation.html

Appendix A

Major Criteria – 1 point each

Minor Criteria – ½ point each

Children under 3 – need 5 pts. Needed

Children 3 and older – need 8 pts. At least 5 must be major criteria.

MAJOR CRITERIA:

- *Neonatal and infantile central hypotonia with poor suck and improvement with age.*
- *Feeding problems and/or failure to thrive in infancy, with need for gavage or other special feeding techniques.*
- *Onset of rapid weight gain between 12 months, and six years of age, causing central obesity.*
- *Hyperphagia*
- *Characteristic facial features: narrow bifrontal diameter, almond-shaped palpebral fissures, down-turned mouth.*
 - *Hypogonadism: Genital hypoplasia: small labia minora and clitoris in females; hypoplastic scrotum and cryptorchidism in males.*
 - *Incomplete and delayed puberty.*
 - *Infertility*
- *Developmental delay/mild to moderate mental retardation/multiple learning disabilities.*

MINOR CRITERIA

- *Decreased fetal movement and infantile lethargy, improving with age.*
- *Typical behavior problems, including temper tantrums, obsessive-compulsive behavior, stubbornness, rigidity, stealing, and lying.*
- *Sleep disturbance/sleep apnea*
- *Short stature for the family by 15 years of age*
- *Hypopigmentation*
- *Small hands and feet for height age*
- *Narrow hands with straight ulnar border*
- *Estropia, myopia*
- *Thick, viscous saliva*
- *Speech articulation defects*
- *Skin picking*

SUPPORTIVE FINDINGS – not part of diagnosis

- *High pain threshold*
- *Decreased vomiting*
- *Scoliosis and/or kyphosis*

- *Early adrenarche*
- *Osteoporosis*
- *Unusual skill with jigsaw puzzles*
- *Normal neuromuscular studies (e.g. muscle biopsy, EMG, NVC)*

(Butler, Cassidy, Greenswag, Hanchett, Holm, Whitman, et al, 1993)

Dear Program Leader,

We are very excited that you have chosen to use “Putting the Pieces Together; Understanding Prader-Willi Syndrome” in order to educate the children in your organization about Prader-Willi Syndrome (PWS). Included in this packet is a compilation of moderately in-depth background information about PWS for you to read prior to leading the program for the children. This information will help you to fully understand PWS, as well as the necessity for programs such as this one. An outline of the presentation, including suggested materials and a power-point that can be used as a visual aid during the presentation have also been included. We suggest that you look over the outline and power-point first and then read the supplemental information so that you can make sure to focus on aspects that you may be more unfamiliar with, or suspect that you will be asked a lot of questions about based on the nature of the group that you will be educating. A list of web-sites with more in-depth information can be found has also been included.

Although it should only take about one or two hours to read the information on Prader-Willi Syndrome and take a look at the links provided, it is suggested that you prepare at least one week in advance so that time is left for questions to be answered. It is also a good idea to do a few run-throughs of the presentation to make sure that you are comfortable presenting it. Practicing on family members, or other adults is a great idea!

We sincerely appreciate your help in this effort to provide positive inclusive environments for children with PWS, and hope that this program provides a positive experience for everyone involved.

Sincerely,

_____?

Contact Info

Appendix C
Power-Point Presentation

Appendix D

“Putting the Pieces Together: Understanding Prader-Willi Syndrome”

Outline for the Presentation Leader:

Slide #1 (Title of Presentation)

V. Visualization:

Slide #2 (Visualization)

The visualization exercise is a great way to put the kids into the shoes of a person with Prader-Willi Syndrome. Depending on the maturity level and preferences of child or children with PWS who this program is intended to help, they may choose to write this exercise themselves so that it is more specific to them. The following is a script that can be used:

Please close your eyes and put yourself in the shoes of a child your same age with Prader-Willi Syndrome. You just came home from school, and you are hungry; you have been hungry all day. No matter how much you eat, you are still hungry. There are some things that you can do to take your mind off of it, but the feeling never stays away for very long. You had a pretty good day at school, but some of the kids made fun of you for the way that you acted when the schedule got changed at the last minute and lunch was twenty minutes late. You could not help the way you acted, it is just a part of who you are. How would they feel if they always felt hungry, could only eat very healthy foods and a portion of what the other kids could eat? How would they feel if they didn't always have control over their emotions? The other kids don't always understand why it is harder for you to do well in some subjects, but can do really well at some things like doing jig-saw puzzles. They just don't understand that you are the way that you are because of Prader-Willi

Syndrome, something that you were born with. They just don't always see the wonderful, fun, and nice person underneath who really wants to play with friends, join extra-curricular activities and just live the life that all kids want – a life full of fun, friendships and adventures. (Pause) Please open your eyes, can someone tell me how this it felt to be in the shoes of someone with Prader-Willi Syndrome? Now, you may be confused and wondering, what is this thing called Prader-Willi Syndrome, well, let's find out!

VI. Prader-Willi What???

This section is used to explain the basics of Prader-Willi syndrome to the kids.

It is OK to add in more information if the kids are at a more mature level, or if there is something that you previously read that you think will help the kids understand PWS. A key feature in this section is to focus on differences and abilities, rather than *disabilities*. *Slide #3 – The picture is of a person “building” DNA. It is a visual aid to show how the microscopic DNA, which are made out of genes can be symbolized as a person's building blocks that make them who they are.*

- A child gets Prader-Willi Syndrome before they are born, while they are in the mother's stomach! It is a genetic disorder, which means that one of the building blocks that make up a person (a gene) is formed differently than usual. There is no way to “catch” this syndrome from a person who has it – it is not contagious. Nobody can control whether a person is born with PWS or not, it is just a way that some people are made.

- **ACTIVITY:** A supplemental activity that can be done at this point is to have everyone with brown eyes (or another trait like asthma, etc.) stand up and ask them if they chose to be that way. It's the same with PWS, the child does not choose to be born with it, it is just a part of them and something that they have to live with.

Slide #4 (Prader-Willi What??? continued – manifestations of PWS)

- The genes, or building blocks that are different in a person with PWS cause a part of the brain called the hypothalamus to function differently than people without PWS. This causes many differences, including:
 - **Constant hunger:** Their brain does not get the message that they are full, so they continue to feel hungry even if their stomach is full! People with PWS also have a lower need for calories, which are energy from food. That means that they have to eat less than other people that are their same size in order to stay in good physical condition. They usually have a very strict diet.
 - **Different ways of learning:** People with PWS are usually extra good at activities like putting together jig-saw puzzles and remembering things for a LONG time – they have a great long-term memory. They do learn differently and it may take them longer to perform some academic and physical skills, like

learning how to read or remembering how to do something right after they are taught (short-term memory).

- Different body type: People with PWS are born with lower muscle tone, so they have to work extra hard to keep their body in good physical shape and it may be harder for them to keep up with other kids. Sometimes they will have distinct facial and body features that look different from other people.
- Different ways of handling emotions: People with PWS are usually very nice and love to help and be friends with others! They do, however, sometimes have a harder time handling their emotions. People of all ages with PWS are more likely to have temper-tantrums than other people their age. It is important to remember that the reason for their different emotional and behavioral responses is due to PWS and not because they are trying to be mean, annoying, or childish. It's just like when a person who has asthma has an asthma attack, they usually can't control it. When a person with PWS has temper-tantrum or is acting out, the best thing to do is leave them alone and try not to pay attention. An adult will handle the situation.

VII. Puzzle Time

This activity is intended to visually represent diversity among all of the

participants. The children will learn that all of the puzzle pieces are different and necessary to complete the puzzle, just like in real life everyone is different but important to the group. This activity can be short – five minutes, or a longer time can be spent on it depending on the nature of the group.

Materials Needed:

- One large sheet of butcher paper, poster board, etc. that is cut into enough jigsaw pieces for every child (adults can be included as well) to have a jig-saw piece.
- Markers, colored pencils, crayons, paint, etc. for the children to decorate their puzzle piece.
- Tape to keep the puzzle together. It also works to have another poster board with a piece of tape for each jig-saw piece already on it to make the activity faster and easier.

Slide #5 – Doing the Puzzle Activity

Instructions:

- Hand each child a jig-saw piece and instruct them to decorate their puzzle piece in a way that describes them. Some suggestions include pictures of their hobbies, friends, family, pets, favorite subjects, or a picture of themselves.
- Have the children one-by-one, or all together put the puzzle together.

Slide #6 – De-briefing the Puzzle Activity

The purpose of this section is to discuss the activity with the children, and

make sure that they understand the moral behind it. Here are the discussion questions and the intended ideas that are portrayed by them:

- Are any of the pieces the exact same? No, because everyone is different.
- What would happen if a piece were left out? (Visually removing a piece may be helpful) The puzzle is not complete. If someone is left out of a group, the group would not be whole.
- How would you feel if YOUR piece was taken out? Left out; not very good. That is how a person with PWS feels if they are left out of the group and not accepted for who they are.
- Are any of the pieces more important than another one? No. They are all equally important in completing the puzzle. Everyone in a group is equally important, no matter what they look or act like!

IV. Some things to Remember...

This section answers some questions that the children might have about being friends with a child with PWS. It also goes over what they should do in certain situations and how they can help the person with PWS to be included.

Slide #7

- Having a friend with Prader-Willi Syndrome opens up the door to a new and rewarding friendship! People with PWS have a lot to offer as a friend and there are a lot of ways that you can both help each other!
- What do I do in situations that involve...
 - *eating?* Try not to eat, or have snacks around a person with PWS unless you have talked to the instructor/teacher or their parent or care-

giver first. If they grab food from you, please let them have it and then notify the instructor/teacher or their parent or care-giver. However, people with PWS have very strict diets, so please never offer them food no matter what they say or do in order to get food from you. People with PWS eat very healthily, maybe you can learn about having a healthy diet from them!

- *different behaviors and emotions?* If a person with PWS is having a temper-tantrum or other disrupting behavior, the best way that you can help is to ignore them. Move away from them if you are close and allow an adult to handle the situation. Sometimes, a person with PWS will perseverate, or repeat a question, or phrase many times in a row. The best way to handle this is to stay calm and politely tell them that you will answer one more time. If it continues and you begin to feel annoyed, tell an adult and they will help handle the situation.
- *the child with PWS being made fun of?* A great way to handle this is to either tell the adult in charge and allow them to handle it, or politely explain to the other child that this person has PWS and that is just the way that they are made. The best way to help with this is to show the children who are making fun of the person with PWS that he/she is your good friend even though they may be a little different!

V. “My Name’s Not Willy!” Rap-Along

Slide #8

This rap was written by Tad Tomaseski who is a youth minister in Frisco,

Texas, along with his youth leaders, and teen group. They formed a musical group named “Xcel,” and wrote and recorded “My Name’s Not Willy©” with the purpose of boosting self-esteem in people with PWS at a PWSA-USA convention in 2003. It is from the perspective of a person with PWS. A copy of the words is included so that they can be handed out, made into a poster or taught to the kids so that they can rap-along.

Dancing-along is also a lot of fun!

- *Activity Idea:* A great activity idea for this song is to play the “freeze” game with it. First, let the children listen to the whole song one time through. Then, have them all stand up and dance and sing along until the music stops. Strategically stop the song at points that would be good for discussion. Then, have someone (it can be the first or last person who freezes, a volunteer, or just a random child) explain what the rap is talking about at that time. Then, turn the music on and do it all over again.

VI. Questions!!!

Slide #9

Remind the children that questions are always welcome, even after the presentation is over. If they have questions that you cannot answer, please write them down so that you can find out and let them know. Feel free to contact us with any questions that you do not know or feel uncomfortable answering and we can find out and help formulate an answer!

Appendix E

1. It is 6:00 p.m. and you are going to see a movie with a friend at 7:30, however, you just found out that the movie actually starts at 7:00 and you need to tell your friend. You:

- a. Use a phone to call them.
- b. Use a cell-phone to text message them.
- c. Send an e-mail.

The Costa Rican students unanimously, and five of the U.S. American students chose (a). The other five U.S. American chose (b). This situation displays an urgent situation with a person who the participant is close to. Text messaging was added into this question, however, text messaging is not as popular in Costa Rica due to the high prices for cellular phones, and therefore is not relevant. In this question it provided a text-based option for students to choose since e-mail would probably not be quick enough, or efficient for this particular situation.

2. You need to change a flight reservation with an airline company that allows you to do this online or over the phone. You:

- a. Use a phone to call them
- b. Use the internet.

Nine of the Costa Rican students, and all of the U.S. American students chose (b). One of the Costa Rican students chose (a). This question signifies a situation with a person/company that is not close to the person, or in their in-group. However, the factor that both household and cellular phones in Costa Rica are rather expensive could greatly contribute to this answer.

3. You are living away from your parents and need to talk to them about your plans for going home to visit in a month. You:

- a. Use a phone to call them.
- b. Send an e-mail

All of the Costa Rican students and 7 of the U.S. American students chose (a). The remaining three U.S. American students chose (b). This question signifies a situation with people whom the subject is very close to, with a time frame that is not urgent.

4. You have a question for a professor who accepts both phone calls and e-mails (with a quick response rate). You:
 - a. Use a phone – call him/her.
 - b. Use the internet – e-mail.

Seven of the Costa Rican students and three of the United States American students chose (a). Three of the Costa Rican students and seven of the U.S. American students chose (b). This question signifies a situation in which the subject has a business/educational relationship with the person.