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THE EDUCATIONAL IMPACTS OF TUBEROUS SCLEROSIS COMPLEX

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Abstract

Tuberous Sclerosis Complex (TSC) is a rare genetic disorder that causes tumours to grow throughout the body. This can have serious consequences for individuals affected throughout their lifetime. People with TSC are affected by this disorder in terms of their medical health, cognitive and behavioural functioning. There is currently no research in the literature addressing the educational profile, needs, or cognitive profile from an educational framework of children with TSC. As children with TSC move through the education system, a number of issues can potentially affect their educational success. This qualitative research study examined the educational impacts of TSC on children currently in the education system. The aim of the study was to gain an understanding of practices that are facilitating success in these children's educational experience and specific deficits commonly experienced. In depth, semi-structured interviews were carried out with 10 parents of children with TSC (in Grades 1 thru 8), and 6 of their teachers/educational assistants (EAs). Academic, social and medical findings and their implications are discussed.

Keywords: Tuberous Sclerosis Complex, TSC, education, accommodations, behavioural differences, Autism Spectrum Disorder

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Introduction

This thesis describes a qualitative research study that examined the educational impacts Tuberous Sclerosis Complex (TSC) has on children currently in the education system. The aim of this study was to gain an understanding of what types of practices are facilitating success in these children's educational experience, and an understanding of what specific deficits are commonly experienced. This thesis begins with a description of the research problem and rationale for the current study. A literature review follows, which describes the basic features of the disease as they manifest themselves physically and cognitively. This is followed by a look at research into other genetic diseases and a justification for the need for more research into TSC. Next, a methodology section outlines the specific research methods used in this project, as well as the theoretical underpinnings for these choices. This is followed by the results of the study and a discussion. A discussion of the limitations of the study follows.

TSC is a rare genetic disorder that causes tumours to grow throughout the body. This can have serious consequences for affected individuals throughout their lifetime. People with TSC are affected by this disorder in terms of their medical health, cognitive functioning, and behavioural functioning. As children with TSC move through the education system, a number of issues can potentially affect their educational success, including specific cognitive deficits that can range from mild to severe, behaviour problems, and complex medical needs.

The Research Problem

TSC is an under researched disease. It can present in a number of ways, and can have far reaching consequences for individuals affected (Humphrey, Williams, Pinto, & Bolton, 2004). This study proposes to examine what some of the educational impacts of TSC can be on children affected. Little research has examined what sorts of specific deficits children with TSC tend to have, and no studies could be found in the literature outlining what kinds of effects these deficits, as well as the behavioural and social components of this disease, might have on the schooling of such children (Carlisle, 2003; de Vries et al., 2005). Besides indicating that children with TSC often fall into the intellectual and developmental disability range of cognitive functioning, there are little data indicating specifically what they experience difficulty with, and what strategies and approaches that teachers, paraprofessionals and parents have found to be helpful to their educational success (Carlisle; Humphrey et al.).

Study into the aetiology of TSC from an educational perspective can potentially help individuals affected with this disease by assisting their parents and educators to understand what specific deficits they are likely to encounter. It could also help them to understand what sorts of strategies have been tried in the educational setting that have had some success, and what types of educational services children with TSC are currently receiving. With the current trend towards inclusion into mainstream education, this study might also help educators in Ontario, and elsewhere, as they are increasingly likely to have children in their classrooms with special needs. Other rare genetic disorders have been put forth as

genetic models of certain learning disorders, helping researchers to gain insights into the genetic bases of various disabilities (see for example Mazzocco, 2001). It is possible that in the future, with further research, disorders such as TSC could become models for specific learning difficulties, extending the understanding of the genetic mediation of such processes. With strong co-morbidity between autism spectrum disorders and TSC, it is also possible that further study could reveal some genes that may be linked to autism (Baker, Piven, & Sato, 1998). The large majority of autism cases, an estimated 94 percent, have no known genetic cause, while one percent of autism cases are caused by TSC (Goorden et al., 2007). Thus, the possibility of gaining a better understanding of the genetics behind autism through studying TSC is great. Napolioni, Moavero and Curatolo (2009) note that in studying TSC, we are provided with an excellent model of current issues in the developmental neurosciences. Thus there are many important reasons to study TSC. This study attempts to be a starting point for more research into this disorder and its impact on children in the education system.

This study employed a qualitative research methodology because qualitative methods are "useful in studying a limited number of cases in depth" (Johnson & Onwuegbuzie, 2004, p. 20). A case study approach was used, focused on gaining an understanding of the current educational experiences of a small number of school-aged children with TSC. The targets for this study were elementary school children, ranging from Grade 1 to Grade 8. The study consisted of interviews with teachers and educational assistants (EAs) involved

with the child to gain an understanding of what successes and difficulties these children had in school, and what services and accommodations were involved in the child's education. Interviews were also conduced with parents to try to gain an overall, global understanding of the child's progress, goals and difficulties.

Researching the Personal

In the course of preparing and carrying out this research, I had some ethical and methodological considerations to sort out. I became interested in Tuberous Sclerosis after my first child was diagnosed with this disorder. This has led me down the path of discovering what it meant to research something I was personally invested in, and what that meant for my research and for me.

Scholarly writing and researching that involves the personal is a controversial topic that has received more attention in recent years (Bleich & Holdstein, 2001). My methodological choices, their consequences, and my own investment in the study of TSC will be explored with reference to the current literature, primarily from education, on the use of narrative, researcher-participant relationships, writing from the self, issues of voice and representation, personal history research and power issues in research relationships.

I came to research TSC after my daughter was born in 2006 with this disorder. After dealing with the initial shock, denial and other bereavement symptoms, similar to what Holland (1996) describes in other parents dealing with a TSC diagnosis have gone through, I started to wonder how I would incorporate this new reality into my life. I went to a conference, hosted by TSC Canada, a non-profit organization supporting parents and people affected by TSC. Two doctors spoke. One, a neuroscientist named Dr. Whittemore, has a child with TSC and in fact herself has TSC, and has dedicated her

career to working with the TSC Alliance (Whittemore, 2007). Another doctor, Dr. Hulbert, a urologist, spoke about how he came to research TSC. He has a son, now a teenager, who was born with this disorder, and he wanted to do something to help children affected with it. He devoted his research career to researching kidney and urological issues in TSC. He suggested everyone at the conference had some sort of gift that they could use to help others with TSC (Hulbert, 2007). For some it might be ability in fundraising, or public speaking. It got me to thinking, what was I good at? What could I do to help? I realized that as a student pursuing a Master's degree in education, I was in a unique position to contribute to the field of study in TSC. So I decided to begin this study for my thesis about the educational impacts of TSC.

In the course of planning the study, I realized I would also need to find out about the children's medical difficulties and manifestations of the disease to date, age, method of diagnosis, challenges and behavioural differences, and a whole range of other related questions. This made me reflect on my own feelings towards those questions, and how I might feel if I were the participant answering those questions instead of the researcher. Holland (1996) and Whitehead and Gosling (2003) have both found that the initial diagnosis of TSC can bring up many negative emotions, similar to grieving. De Vries et al. (2005) found that parents and caregivers of people with TSC experienced considerable stress in caring for these individuals. Thus, in conducting this research, I may have been bringing up some negative emotions.

Researching from within a community or group, in which the researcher is already a member, is a process DeLyser (2001) calls "professionalizing the personal" (p. 446). Some researchers, including myself, "find topics close to home, or close to our

hearts-topics so compelling we can't leave them alone-and we try to find ways to use our "insider" status to help, not hinder, insights." (DeLyser, p. 446). In my choice to study the educational issues of children with TSC, I was very much studying from an "insider" position. Despite the possible complications of studying from within the community of parents of children with TSC that I have become a member, the hope that I could contribute something positive and help others affected by this disorder, of which I have so much personal knowledge and understanding, was quite compelling to me.

Although the most challenging aspect of the project was recruiting participants, most of the participants that I spoke to seemed happy and willing to share their experiences with me. I think TSC is a rare enough disorder that many of the parents relished the opportunity to have someone who understood the disorder listen to their experiences. Being an insider researcher helped to bridge the gap between my participants and myself. It helped my participants feel they could talk to me about their experiences, and not feel it was such an intrusion into their private world. This seemed especially true for the participants from Canada. In Canada, the organization to support families with TSC is very small, and there is only one TSC clinic, which has very recently opened. Thus, many parents do not have the opportunity to meet with and speak to other families affected by TSC. The situation is somewhat different in the United States, as their TSC support organization (the TSC Alliance) is very large and has 26 TSC clinics across the country. Many American parents have more opportunities to meet other families with TSC and have the opportunity to see specialists in TSC. In the education field, the TSC Alliance has a full time educational advocate that parents can contact for assistance in getting appropriate services and programming for the child in

school. Therefore, those parents may feel more supported in their efforts to navigate the education system.

In one case, the interview started off with the mother sounding a little weary of my questions, until a comment of mine about my daughter having the same type of seizure as her daughter made her realize I had a child with the same disorder as her. Her whole demeanour seemed to change, and she seemed to relax and share more openly. My insider status seemed to make this participant feel more at ease with my questions.

Lincoln (2005) suggests that in ethnographic research it is sometimes insiders, members of the community who become researchers, who are able to more accurately represent their community from the inside than an outside researcher coming in. She is referring to research about different cultures, but I think this same logic can be applied to a researcher studying any community from the inside. The affirming questions often asked by participants during an interview, questions like 'You know what I mean?', might be more meaningfully answered by a member of the community. I wondered if by entering the researcher-participant relationship as an "insider", a member of the community of parents with children who have TSC, I might be able to build a more equal relationship with participants. Although I would not be so naive as to suggest that power relationships can be equalled and that my participants and myself can be equal partners and collaborators, efforts to address the research relationship are valuable and worthwhile. The research relationship necessarily changes when the researcher is doing insider research, and having this insider status may lessen the divide between researcher and participant. By situating myself and acknowledging my own position in the research

as a mother of a child with TSC, I felt I could build a more open and honest relationship with my research participants.

The interviews with parents differed in several ways from the interviews conducted with teachers and EAs. The parent interviews were much longer than the teacher/EA interviews. This is somewhat surprising, as this was an educational study, one would have expected the teachers/EAs to have more information to contribute as the experts on education. The parent interviews averaged 50 minutes, with the longest being 200 minutes and the shortest being 23 minutes, while the teacher/EA interviews averaged 23 minutes in length with the longest being 44 minutes and the shortest being 12 minutes.

There was a difference as well in the amount of times the parents sought reciprocal information and asked personal questions as compared to the teachers. Owens (2006) argues that researchers must be prepared to be asked for reciprocity in sharing, and that this sharing on the part of the researcher may help to lessen the feelings of shame on the part of the participant by evening out the power differences inherent in the relationship. This is an important point to consider in the context of my own study, as many of the participants may have felt strong emotions about having a child with a disability, recounting painful stories about how they were diagnosed and what the education system has done (or not done) to accommodate their special needs. The possibility of shame and strong emotions was very real. By building a relationship with the participants from a shared space of having a child with the same diagnosis, it was my hope that the interview space could be expanded for sharing and reciprocity, and that my participants would feel more comfortable sharing their stories with me. Almost without exception, the parents I interviewed at some point asked me reciprocal questions about

my experiences with my daughter and her manifestations from TSC. By contrast, the teachers did not generally ask me many reciprocal questions. A couple of teachers asked me about my research, or about TSC, but no personal questions about me and my experiences.

I expected participants to express a need for reciprocity, but what I did not anticipate was the strength of my need for reciprocity. I felt whenever a participant was sharing their experiences, it was hard for me not to jump in with my own experiences and relate their story back to mine. Although the process was therapeutic, to have the opportunity to hear from other mothers who had endured many of the same difficulties and hardships that I had, I found it was somewhat difficult to pull myself back and restrain from letting the relationship between myself and my participants from getting too close. It was important for me to recognize and acknowledge that these experiences and stories I was hearing, although similar to mine, were not mine and were separate from me. Otherwise, I feared I would put too much of my own experience into the data and not let it speak for itself when it came time to analyze what I had collected.

There were many times in the parent interviews when my participants asked me reciprocal questions about my own daughter, her seizures, her other physical manifestations. This was expected, but what was unexpected was that I noticed several points in the interviews when I offered up information that was not solicited by participants. While I think part of the reason for this was my own need to reciprocate and feel validated by these other mothers with children having the same rare disease, I think part of it may have also been to ameliorate their possible sense of shame in discussing such a difficult thing as having a child with a disability. It brought me back to when my

daughter was first diagnosed and I felt it was somehow a secret I had to keep from the world. As if I somehow was not living up to the expected trajectory everyone's life was supposed to take; get married, have a career, have a perfect, normal child; as if I had somehow failed and needed to hide my shame. It was as though in some of my interviews I felt the need to tell my participants it was ok to let out these stories; that I had experienced similar things, almost as if I thought if I shared my story, it would lessen their shame and make it ok for them to tell their story.

In some ways I suppose part of my decision to research something in which I am personally involved and to include my own story in my research reflect my desire to create what Eakin (2004) calls a "counterstory". Eakin notes "telling counterstories enlists lifewriting in the service of social change, recasting the discourses of disability" (p. 12). Instead of the story of the child's helpless and devastated parent, I wanted in a way to create a counterstory of the parent of the child with a disability as powerful and able to affect change from within the community of people affected with this disorder. I had hoped that this research endeavourer might help other parents of children with TSC to create their own counterstories about their role as parents of children with disabilities. Clark/Keefe (2006) suggests a transformative process when working with participants with whom the researcher shares a background. By coming to my participants on a somewhat equal footing, as an insider, it was my hope that the researcher-participant relationship could transcend the traditional power differences and boundaries and become a true conversational space, where participant and researcher alike are free to share their stories and meanings in a deep and personal way. This was evident in my interviews

with parents. In several cases, when I finished the interview questions, the interview continued and turned into a conversation.

In my own research, I feel in order for the reader to truly appreciate the position from which I am writing, and to understand my conclusions, reporting, omissions, research relationships, and interview process, they must come to my work with the understanding that I am writing from within the community about which I speak. As a parent of a child with TSC, I am writing from a unique position within the field of research about TSC.

While it is my intention to represent the interviews and studies I conduct as accurately as possible, there is little doubt that my interpretations and even presentations of my data will be necessarily influenced by my own experiences and feelings about having a child with the disorder I am studying. For this reason, I have made the methodological choice to be upfront with both my participants and my readers about my own personal investment and involvement in the subject I am studying. Lincoln (2005) notes that there has been recent recognition of the limitations of so-called objective research. Even in positivist research, there is likely no research that is free from subjectivity. The illusive objectivity that many researchers subscribe to, although possibly a worthy goal in some circles, is likely a physical impossibility. As researchers, we play a large part in shaping our encounters with participants. "Consciously or not, we listen and make sense of what we hear according to particular theoretical, ontological, personal, and cultural frameworks and in the context of unequal power relations" (Luttrell, 2000, p. 499). Even a transcript of an interview is "already an interpretive and contextualized text; it is interpretive because it is shaped by the interpretive processes of

researcher and participant and their relationship, and it is contextualized because of the particular circumstances of the interview's origins and setting." (Clandinin & Connelly, 2000, p. 94). Thus, there is little doubt that any research project, such as this one, will be subjective. Luttrell suggests that researchers cannot "eliminate tensions, contradictions, or power imbalances, but I do believe we can (and should) name them" (Luttrell, p. 500). By "naming" my position upfront in my research project, I am acknowledging both its limitations, and possibly some of its strengths.

In deciding to research something that is personal to the researcher, either by researching from within a community in which the researcher is a member, or by researching something in which the researcher has personal experience, several key considerations come to the fore. Although it is unlikely that any qualitative research could really be "objective" and not influenced by many personal factors on the part of the researcher, the researcher must ask themselves, by acknowledging their subjectivities in an open way, what is lost and what is gained (see Luttrell, 2000,). I suggest that what can potentially be gained is a more open research relationship, in which power differences are decreased, that can ultimately lead to more open, honest sharing of participants narratives. Owens's (2006) conception of the "shaming" interview can be lessened when studying an emotionally laden topic when the researcher is coming to the interview experiencing many of the same feelings as the participants. This can help to form a more interactive approach to researching (as described by Chase, 2005). The researcher sharing similar narratives might satisfy the need for participants to have some form of validation and reciprocity in their experiences. Both the researcher and the participant can use the research process as a way to create a positive counterstory for themselves,

and to restory their own narrative into a positive story. It is my hope that my participants were able to accomplish this by feeling that they contributed proactively to the educational experience of their child by participating in a research project aimed at improving education for children with TSC.

Context and Background—the Relevant Literatures

Research in TSC has historically been focused on the medical concerns and medical aetiology of the disease. It has primarily been viewed as a medical and psychiatric disorder, and framed as such. Thus, no research on TSC from an educational framework exists.

It is important to gain some background understanding of the basic aetiology of TSC, as it manifests itself physically and cognitively. By gaining insight into the medical complexities of this disease, the reader can begin to appreciate the numerous issues a child with TSC may bring to the classroom.

Tuberous Sclerosis Complex is a multi-system, autosomal dominant, genetic disorder that can affect almost any organ system in the body (O'Callaghan & Osborne, 2000). Individuals affected develop benign tumour growths in major organ systems throughout the body (Crino, Nathanson, & Henske, 2006; O'Callaghan & Osborne). It is estimated that approximately 1 in 10, 000 live births will have this disease (Humphrey, Williams, Pinto, & Bolton, 2004). TSC can be inherited from a parent with the disease, but it is thought that approximately two thirds of the cases are the result of a random mutation (Humphrey et al.). The disease results when a mutation occurs on the TSC1 or TSC2 gene. These two genes play an important role in suppressing tumour

growth. They act together on a GTPase activating protein complex that, in turn, affects the mTOR pathway, which is thought to have a regulatory role in growth on a cellular level. Thus, a mutation in either gene affects this complex, resulting in the same disorder (Jansen et al., 2008a).

There has been some research to indicate that individuals with the TSC1 mutation tend to have a better outcome and less severe disease phenotype than those with the TSC2 mutation (Winterkorn, Pulsifer, & Thiele, 2007). Jansen et al. (2008a) recently examined this relationship and found that, as a whole, individuals with TSC2 tended to have an earlier age of seizure onset, lower cognitive abilities and more brain involvement as seen on MRI scans as tubers, than people with a TSC1 mutation. However, the within group variability was wide, and there was much overlap in these two groups of patients.

Occasionally, TSC can be diagnosed pre-nataly through ultrasound imaging of the fetus that identifies cardiac rhabdomyoma, benign tumour growths in the heart. These heart tumours are common in newborns and young children with TSC, occurring in as many as 47-60 percent (Schwartz, Fernandez, Kotulska, & Jozwiak, 2007). They develop during weeks 22 to 28 of prenatal development (Franz, 2004). This method of detection is becoming more common, as more women have ultrasounds in the later half of pregnancy (Jozwiak et al. 2008). If the disease is not caught in this way, it is often diagnosed during childhood through presentation of seizures or skin manifestations.

In order to be diagnosed with TSC, individuals are clinically evaluated for two or more major features of the disease or one major and two or more minor

features of the disease. Major features include cardiac rhabdomyoma (benign tumours in the heart), cortical tubers (tumours that develop during the prenatal period in the cortex of the brain), and facial angiofibroma (red bumps that appear on the face and continue to grow, sometimes large enough to be disfiguring). Minor features include dental pits, confetti skin lesions, and bone cysts (Crino, Nathanson, & Henske, 2006). Interestingly, the clinical findings in TSC vary with developmental stage. For example, cortical tubers and rhabdomyoma form early in gestation, and are thus often detected in infancy; whereas, skin manifestations, such as angiofibromas, and kidney findings often develop during later childhood or puberty, and lung tumours generally only affect women with TSC past the age of thirty (Crino et al.). Thus, depending on the age of the child, different issues may be at the forefront of parent's concern. There may be different issues the educator or others who work with children with TSC may need to be cognizant of during different developmental stages. For example, children often experience increased seizure activity and worsening of skin manifestations during puberty. Increased seizure activity is something that anyone working with a child with TSC should be aware of and carefully monitor for, and the skin lesions may cause self-esteem issues for the child. Both issues can present themselves within the classroom environment, and may impact the schooling of such children.

TSC can affect almost any organ system in the body. The tumours that develop have variable penetrance, and people with TSC may be affected by all or only a couple of these tumours (Crino et al., 2006). The disease presents with

congenital tumours in any organ system, including the heart, skin, eyes, kidneys and central nervous system. In some organ systems, such as the kidneys, these tumours can continue to grow, whereas in the heart, they tend to be present at birth and shrink as the child grows (Goodman, Lamm, Engal, Shepherd, Houser, & Gomez, 1997). In the central nervous system, cortical tubers are often present throughout the brain and subependymal nodules (SENS), calcified areas, develop in the ventricles of the brain. The SENS can occasionally grow into subependymal giant cell astrocytomas (SEGAS), giant tumours, which can lead to a blockage of spinal fluid and hydrocephalus. In these cases, brain surgery to remove the SEGA or implant a shunt may be necessary.

Recent research and trials with drugs such as Rapamycin, a drug commonly used in transplant patients, give hope to TSC patients about possible new treatments. Rapamycin has been shown to shrink kidney, lung and brain lesions in several pre-clinical trails (Crino, 2008). Interestingly, recent work with patients treated for subependymal giant cell astrocytomas (SEGA) has shown that treatment with these and similar drugs may prevent or lessen epilepsy and cognitive dysfunction (Krueger et al., 2008). Although early, these results are very exciting for both researchers and people living with TSC. It is possible that these recent advances may someday lead to more and improved treatment options for people living with TSC that may improve learning, social and behavioural interaction and physical manifestations (Crino). Until these treatments become available and are proven effective, however, it is important to research ways to

improve the current cognitive functioning and academic success of children with TSC.

With the complexity and variety of physical manifestations of TSC, it is little wonder that there are some severe cognitive deficits often associated with this disease. The relationship between the physical symptoms and the cognitive effects of these manifestations, however, is not well understood. Cortical tubers are thought to play a role in the severity of an intellectual handicap present in an individual, with some literature finding that children with seven or more often fall into the severely affected range (Goodman et al., 1997). However, the relationship is not linear, with some individuals being severely affected with only a small number of tubers and some individuals having many tubers and being only mildly impaired (Roach, 1997). More recently, Jansen et al. (2008b) have advanced a hypothesis that it is not tuber load per se, but instead what they call tuber-brain-proportion or TBP. That is, they found the amount of brain taken up by lesions to be a better predictor of cognitive functioning, epilepsy and age of onset of epilepsy, than simply the number of tubers. Still, the authors point out that a large TBP "is neither necessary nor sufficient for early seizure onset or cognitive impairment" (Jansen et al., 2008b, p. 921). Zaroff et al. (2006) found that the two best predictors of lower cognitive functioning were the age of seizure onset, and whether cortical tubers were found on both sides of the brain instead of just one side. Their work also pointed to a strong association between poor cognitive outcome and a particular type of epilepsy, infantile spasms, but this association failed to reach statistical significance. Recent work in mouse models

of TSC further complicates the picture of cognitive functioning in TSC. Goorden et al. (2007) found that TSC knockout mice were impaired both cognitively and socially even in the absence of epilepsy or brain lesions.

Epilepsy is one of the common complications of TSC, occurring in an estimated 78 to 96 percent of TSC patients (Kopp, Muzykewicz, Staley, Thiele, & Pulsifer, 2008). Uncontrolled epilepsy is known to cause intellectual impairment in many cases; however, the relationship in TSC is not completely predictive (Humphrey et al., 2004; Husain et al. 2000; Roach, 1997). Continued and uncontrolled seizure activity is associated with a poor cognitive outcome (Winterkorn, Pulsifer, & Thiele, 2007). Although any type of seizure can be seen in TSC, infants with TSC often develop a particularly devastating form of epilepsy called infantile spasms (IS), which, if unable to be controlled with medication, often have very severe developmental consequences (Goodman et al., 1997). Jansen et al. (2008a) found that early onset epilepsy, particularly IS, was related to poor cognitive outcome, and that normal or above average intelligence with these two factors in place was very unlikely. They reported that previous studies had found 85 to 100% of patients with IS fell into the severe developmental disability range. Interestingly, in a companion paper, these same authors found that epilepsy was not a necessary condition for low cognitive functioning (Jansen et al., 2008b). They found several cases of patients with no history of epilepsy who were severely cognitively impaired (Jansen et al, 2008b). This further complicates the causes of intellectual impairment in TSC.

Behavioural problems are common in TSC. Many families report that behaviour is their primary concern in dealing with their children's TSC (de Vries et al., 2005). A range of behavioural manifestations have been reported in TSC, including: aggression, social difficulties, self-injurious behaviours and sleep disorders (Curatolo, Verdecchia, & Bombardieri, 2002). Many children with TSC meet the diagnostic criteria for ADHD. ADHD has been reported in the literature to occur as often as 86% of cases and as infrequently as 26% of cases of TSC (Murzykewicz et al., 2007). Muzykewicz et al. found 30% of their sample of 241 patients with TSC had ADHD. However, there is growing evidence to suggest that even in cases where the diagnostic criteria for ADHD are not met, children with TSC "can show specific attention deficits and impaired goal directed behaviour associated with executive control processes" (de Vries et. al., p. 185). In terms of attention deficits in TSC, de Vries and Watson (2008) suggest that attention issues need to be viewed in a neurodevelopmental approach that indicates that if one stage in attention development is compromised, then other stages will be affected in a sequential manner, rather than attentional skills being viewed as separate deficits that develop independent of each other. In TSC, attention deficits may occur as a consequence of earlier attention skills not developing correctly (de Vries & Watson).

Murzykewicz et al. (2007) report that the four highest comorbid psychiatric disorders in TSC are anxiety, ADHD, aggression/disruptive behaviours, and depression. Staley et al. (2008) report up to ten percent of people with TSC engaged in self-injurious behaviour. These researchers noted that

patients with TSC who exhibited self-injurious behaviours more frequently had a history of epilepsy, developmental delay and autism. Thus, dealing with those affected by TSC can be much more complicated than just dealing with the medical issues of this disorder. TSC can include a variety of behaviour and psychological issues of which educators and other professions who work with these children should be aware. De Vries et al. (2005) suggest that children should be evaluated for such co morbid disorders as ADHD, autism and more subtle deficits such as memory and attention difficulties. These researchers suggest a comprehensive and regular schedule of monitoring of individuals with TSC to catch potential problems early to allow for appropriate interventions. These complex difficulties are likely to impact the educational success of children with TSC, although no studies have been done in this area (de Vries et. al.).

Communication difficulties are widely reported in TSC. In some studies, it has been found that approximately half of those with TSC have an autism spectrum disorder, a primarily communication related disorder, that can also cause behavioural differences (Weber, Egelhoff, McKellop, & Franz, 2000). Studies have varied, but it is estimated that between one and nine percent of children with autism have TSC (Ferguson, McKinlay, & Hunt, 2002; Goorden et al., 2007). However, there are some conflicting reports in the literature about the actual prevalence of autism in TSC. Smalley (1998) reported that estimates of the occurrence of autism in TSC ranged from 17 to 68 percent. An early study by Hunt and Shepherd (1993) found about a quarter of their study participants fit the diagnostic criteria for autism, and a further 19 % (all girls) fulfilled the criteria for

pervasive developmental disorder, without fulfilling all the criteria for autism. In that study, autism was associated with both intellectual delay and with epilepsy, but neither factor was sufficient to account for all cases of autism. Several children had both cognitive delay and epilepsy, but no behaviour consistent with an autism diagnosis. Interestingly, in the general population the ratio of boys to girls with autism is approximately 4:1, but in the Hunt and Shepherd study, the prevalence of autism was equally distributed in boys and girls. Baker, Piven and Sato (1998) estimated the prevalence of autism in their TSC sample to be significantly lower than the aforementioned studies, at twenty percent. They suggest that their methods for detecting autism may have been more strict than others previously reporting higher rates of autism, but also point out that they may have excluded some cases, particularly in the severely cognitively affected range, as they can be difficult to asses. Several children in this study did not fulfil the criteria for Autism Spectrum Disorder (ASD), but still exhibited some of the features of this disorder, further suggesting a possible connection between autism and TSC (Baker et al.).

In their study, Baker et al. (1998) found a correlation between autism and hypsarrythmia (an abnormal brain wave pattern present in infantile spasms). Smalley (1998) also found a correlation between seizure disorders and autism in TSC, and note that there are few cases in which children have autism without seizures. However, they point out that seizures are present in up to 90% of people with TSC, so there are few people with TSC who do not have a history of seizures. There has been debate as well if there is a certain brain region that, if

affected by tubers, might be correlated with autism in TSC. For example, in one study, the authors postulated that autism was positively correlated with tubers in the cerebellum (Weber et al., 2000).

Educational Issues

The developmental and intellectual delays associated with TSC are not well understood (Humphrey et al., 2004). Approximately 40 percent of children with TSC fall into the severely cognitively affected range, defined as having an IQ below 70 (Ferguson et al., 2002). Intellectually, there seems to be what is known as a bi-modal distribution in intelligence, with individuals being moderately to severely cognitively affected or functioning within the normal intelligence range (de Vries et al., 2005). However, recent research has indicated that even in those TSC children and adults that are within the normal intelligence range, there seems to be "specific cognitive deficits in attentional skills and executive skills" (de Vries et al., p. 185). Language delay is also common regardless of intellectual status, suggesting some specific feature of TSC affects language skills (de Vries et al.; Ferguson et al., 2002). Despite these recent findings, no research has investigated the educational or occupational outcomes for people affected with TSC (de Vries et al.).

Although a large number of TSC children have intellectual handicaps, there are no studies and no data in the literatures on how to support their development and as to what types of interventions should be attempted. In other genetic and congenital disorders, such as Down's syndrome and Fragile X syndrome, research has focused on various early intervention programs to combat the intellectual impairment associated with these disorders (de Vries et al., 2005;

Humphrey et al., 2004). Hodapp and Fidler (1999) suggest the importance of understanding behavioural and cognitive phenotypes for various genetic disorders to facilitate appropriate special education programs and interventions. No similar research yet exists for children with TSC (de Vries et al.).

Teachers in Ontario's classrooms, and elsewhere, have to deal with a wide variety of learning needs and issues. Since the passing of Ontario's Education Amendment Act, also known as Bill 82, the Ontario Government has mandated that all students with special needs are entitled to special education services and programs at no cost to parents (Ministry of Education, 2005). Initially, this meant students with exceptionalities were often placed in self-contained special classes, until "Regulation 181, enacted in 1998, legislated the requirement that the first consideration regarding placement for an 'exceptional pupil' be placement in a regular class with appropriate supports" (Ministry of Education, p. 2). This has meant that most students with exceptionalities spend at least 50% of their day within a regular classroom (Ministry of Education). The situation is similar in other provinces in Canada, and in the United States of America, with the passing of similar laws that have mandated special educational services in the least restrictive environment for exceptional students. For example, the Individuals with a Disability Education Act (IDEA) in the United States of America specifies that children with special needs are entitled to receive an appropriate education, that is provided, where possible, in the school the child would have attended, had they not had a disability (US Department of Education, 2008). Likewise, in the province of Alberta, schools are required to provide special education

programming to students with identified special education needs (Government of Alberta, 2003). "Alberta Education's directive is to place students identified with special education needs in typical classrooms in neighbourhood or local schools as the first option" (Government of Alberta, p. 1). The question for educators becomes how to best meet the needs of these diverse learners, while at the same time meeting the needs of mainstream learners. Research in the literature about effective educational practices for children with TSC is lacking, making it a challenge to meet their needs in an inclusive setting. Gaining an understanding of TSC from an educational perspective can help educators who teach these children.

Summary

Tuberous Sclerosis Complex is a genetic disorder that can have serious developmental and cognitive consequences, as well as a variety of medical concerns, for individuals affected. Despite this, little research currently exists in the literature about the specific cognitive profile and appropriate intervention strategies and educational programs that should be implemented for children with this disease. The current study attempts to be a first look into some of the educational issues facing children with TSC, as well as some anecdotal reporting of strategies and interventions currently in use that have seen some success. A semi-structured interview process for a small number of families and educational professionals was conducted to allow for in-depth, qualitative data collection about each child's current educational experience. Information was gained about

the types of manifestations of the disease and the educational interventions and programs experienced by children with TSC currently in the education system. This qualitative look at the services involved and the manifestations of TSC currently experienced by the children in the study is hoped to be a stimulus for further research.

Method

Participants

Participants for the research project were solicited through Tuberous Sclerosis Canada (www.tscanada.ca), an organization dedicated to raising the profile of this disease and supporting parents and individuals with TSC. TSC Canada advertised the study in their tri-annual newsletter "Connections" that is sent out to all the families that are part of their organization. They also posted information about the study on their web site. The study was also advertised on the TSC Canada Listserv, and a TSC Facebook page. The TSC Alliance in the United States advertised the study on their National List Serv as well. The Neurology Department at the Children's Hospital of Eastern Ontario (CHEO) posted a poster in their lobby explaining the current study and inviting participation. A neurologist at the hospital, Dr. Doja, acted as the site investigator to allow participants to be sought in this way. Participants reported finding out about the study from the American ListServe, the TSC Canada Website and the TSC Canada newsletter. The other methods of solicitation did not attract any participants. The advertisement used for the study can be seen in Appendix A.

The participants for this study were nine parents of children with TSC, one parent had identical twins with TSC. Thus there were ten children with TSC in the study. The children ranged from Grade 1 to Grade 8, with the youngest being not yet six and the oldest participant having finished Grade 8 in the previous June. For this participant, information was sought about his experiences while in Grade 8 and below, and his Grade 8 teacher from the previous year was interviewed.

Eight of the children in the study were female and two were male. TSC affects males and females equally, so it is unclear why there were so many more females whose parents decided to participate in the study than males. Of the parents who were interviewed, 9 were mothers and 1 was a father. Five of the children lived in Ontario, one in Alberta, two in Pennsylvania, one in Georgia, one in Oregon. For six of the ten children with TSC in the study an interview with an educational professional was possible. Four teachers and two EAs were interviewed. Informed consent to participate in this study was sought from each participant (see Appendices B and C). All interviews were completed by telephone.

Interview Process

The interview methodology was idiographic in approach, borrowing from ethnographic methods that seek to "know" from an insider's perspective (Wolcott, 1988). The idiographic approach to research posits "one can only understand the social world by obtaining first-hand knowledge of the subject under investigation...The idiographic approach emphasises the analysis of subjective accounts" (Burrell & Morgan, 1979, p. 6). The interviews sought subjective understanding of the educational experiences and difficulties of children with TSC. Polkinghorne (1988) states "knowledge of human experience requires the use of interpretive or hermeneutic approaches" (Polkinghorne, p. 159). This study attempted to gain a subjective understanding of the "human experience" of children with TSC in the educational system.

The interview process was semi-structured, allowing for participants to become engaged by having the freedom to elaborate on answers they were giving. The interview was semi-structured to allow for flexibility and responsiveness on the part of the

researcher and participant as appropriate. Goodson and Mangan (1996) note the importance of researchers attempting to gain "understanding of the life-world of social actors as they themselves understand it" (p. 43). In this research project, the interest was in trying to gain understanding of the children with TSC from the unique perspectives of the parents, teachers and EAs, and all attempts to respect their interpretation of their current educational reality was made.

Assumptions were made that the educational experiences of the research participants (including current practices and strategies that are proving effective, or not effective, current services that are in place, what is lacking in terms of services and supports for these students and a variety of other information about these participants and their current school situation) could be understood through interviewing the most relevant people in their school experience, namely their teachers, EAs, and parents. As Lincoln (2005) points out, there is a difference between reality as it is experienced, as it is understood, and as it is expressed in language through a narrative. Thus, the reality elicited from the participants of this study was understood as reality as the participants uniquely saw it, and as they uniquely were able to articulate it.

The reality that I sought to understand through this study was not the subjective reality directly experienced by the children that were at the centre of this research project, but rather reality as seen through the educational lens of those directly involved in these children's educational experience. Thus, reality is being understood through people who are in contact with these children, but not from the children themselves. This is because the focus of this study was the

educational practices currently in place for these children and on the educational services utilized by these children, as well as the types of difficulties experienced by children with TSC.

The semi-structured interview process was selected to allow participants the freedom to elaborate on questions asked and to give an opportunity to find out particular information about the education of these children. Weiss (2004) notes, "Good interviews are windows into people's lives" (p. 48). It was hoped that by using a semi-structured interview, a window into the educational lives of children with TSC could be gained. Notes were taken during the interview process, and each interview was recorded to ensure accurate transcription.

Some of the information sought during the interview process with the parents was: where the child receives the majority of his/her education (regular class, segregated special needs classroom, half day program, etc.); which services are involved in the child's education (e.g., occupational therapy, speech and language, EA, etc.); what the main clinical manifestations are to date (epilepsy, skin manifestations, kidney findings, etc.); and at what age and how the child was first diagnosed. This information was collected to try to gain an understanding of how individuals are affected with TSC, and how this complex disease affects their current education. See Appendix D for a full list of interview questions.

The interview of the teachers and Educational Assistants (EAs) included items such as: what special educational services are in place for the child; in which areas is the child having difficulties; what behavioural concerns have been noticed; what accommodations has the teacher put in place, and how successful have they been; and

what types of practices have added to the child's educational success. See Appendix E for a full list of interview questions.

After being collected by telephone, data was analysed for an emerging picture and understanding of possible common deficits experienced by children with TSC, and possible common effective strategies and programs in which these children are involved.

The completed interview transcripts were then analyzed multiple times for emerging themes. On the first analysis, data such as medical findings, history, current school situation, and greatest area of concern, as reported by parents, was recoded in a chart (see Appendix F). The teacher and EA interview transcripts were then analysed for basic data such as which services were involved, what the teacher or EA felt the child's greatest area of difficulty was, what type of program the child participated in, and what accommodations were felt to be effective with the child. These results were added into a chart (see Appendix G). Data from parents and teachers or EAs was then compared for consistency. Inconsistencies were analysed, and where teacher/EA and parent disagreed with a medical diagnosis (such as Autism), the parents were deemed to be likely to be correct. Where EAs/teachers and parents disagreed with an area of greatest academic difficulty, teachers/EAs were deemed to be correct. All interview transcripts were then re-visited to look for emerging themes. Themes were assigned a label, and labels were discussed between the researcher and the thesis supervisor to add strength to the data. Transcripts were then re-read to look for the occurrence of established themes.

Triangulation, in which various data sources and analyses are compared to validate qualitative research results (Oliver-Hoyo & Allen, 2006), was used to strengthen

the validity of the data. Data were triangulated between teacher/EA and parent in cases where both could be interviewed, and between participants to look for strong themes. Data were also analysed by the researcher and the thesis supervisor, and potential themes were discussed and agreed upon. This approach was intended to be a preliminary study of the research problem. Some important questions, such as: where most of these students receive their education; what sorts of deficits are commonly reported; how has the disease manifested itself to date and what sorts of supports are currently being used to accommodate them, were expected to be a valuable first look into this under-researched population.

Results and Discussion

The results of this study will be presented below together with the discussion section. These two sections were combined to allow the reader to gain an understanding of the results of the study within the context of the literatures and to gain an understanding of some possible interpretations of the data. Since no research currently exists from an educational framework, it was decided that merging these sections would facilitate the understanding of the educational impacts found in this study within the context of the complex medical literature on TSC.

Several themes emerged in the present study in the areas of: functioning, educational programming, advocacy, and discrepancies between parent and educator findings. These themes will be reported and discussed below, with particular emphasis on how they might relate to the school experience for children with TSC. The discussion will begin with an examination of functioning. Specifically, intellectual functioning will be discussed, including: estimated current intellectual level and possible contributing factors to intellectual impairments. Anxiety, behavioural differences, and the relation between TSC and ASD will also be analyzed within the theme of functioning. An analysis of educational programming will follow, with an emphasis on current difficulties experienced by the children in the present study, accommodations and programming for these children, and current level of support and intervention in place. Possible gaps and recommendations for the educational programming of children with TSC will also be discussed. The importance of advocacy will be discussed as a theme from the perspective of the parent and the educator. Finally, areas where parent reporting differed

from educational professional reporting will be discussed as a theme and possible reasons for these discrepancies will be suggested.

Functioning

Intellectual Functioning

A bimodal distribution of intellectual functioning has been suggested in TSC (Joinson et al., 2003). For the purposes of this study, overall cognitive functioning was estimated based on parental reports and, in cases where teacher/EA interviews were available, from a combination of parental and teacher/EA reports. Based on these reports, three children from the present study are estimated to fall into the severely cognitively impaired range. Two of these children are reported to be on a totally modified academic program that included living skills goals, and one of these children did not yet communicate verbally. Six children seemed to be functioning in the normal range, with some specific deficits. One child's intellectual status was difficult to estimate from information supplied by the mother, and her teacher was not available to interview. The lower rate of severe cognitive impairments in the present study is likely a result of the small sample size and selection bias. It is conceivable that fewer parents of children with severely impaired children would volunteer for a study of this nature due to the higher levels of stress often reported in dealing with a child with a serious developmental disability (Whitehead & Gosling, 2003).

There is much debate about the exact causes of intellectual impairment seen in TSC (Jansen et al., 2008b). However, many studies have reported epilepsy as one of the highest predictors of cognitive impairment (Zaroff et al., 2006). In the present study, all the significantly delayed children did have refractory epilepsy. One of these children had

significant cognitive impairment, autism, and was on a completely modified program in school. In this child's case, she was on her 11th medication, and her parents were exploring the possibility of surgery for her. Another child in the study who was cognitively impaired was currently on two medications. In this child's case, her diagnosis of TSC was not discovered until the age of eight. The other child in the study that presented with significant cognitive delays was currently on five different medications, still without good seizure control. One child in the study had surgery for refractory status seizures, and was not significantly cognitively impaired, but did have some learning issues. He currently participates in a regular classroom with a full time EA's support. His current learning needs were reported to be memory, retention and language skills, however as he was only in grade one, it is difficult to estimate the eventual cognitive effect on his schooling his current learning issues may have. He had not developed autism or current serious cognitive impairment as a result of his early and severe epilepsy.

Infantile Spasms (IS) were not evidenced with cognitive delay in this study, despite reports in the literature that IS lead to severe cognitive delay in up to 100% of cases (Jansen et al., 2008b). One child (see above description) with a history of IS participated in a regular program with EA support. This child did experience some learning difficulties, but this may have been due to the status seizures that led to his need for surgery rather than the IS in particular. One child with a history of IS was in a gifted program. In both of these cases, the IS were controlled relatively quickly, which could account for the favourable intellectual outcome, as some research has suggested that it is important to control IS quickly to prevent serious cognitive impairment (Zaroff et al.,

2006). The third child in the study with IS was in a regular classroom with EA support and resource pull out. This case seemed the most cognitively affected of the children with a reported history of IS, but it should be noted that it is unclear if this child actually had a history of IS or not. The parent thought the child had IS retrospectively after seeing a video of IS, but this was not confirmed by a doctor. If this child did experience IS, she was not treated for it, which could potentially explain some of the increased cognitive symptoms. In another case the mother did not know what types of seizures her child had, making it possible that her child also experienced IS. It is, in fact, likely, given that this child's epilepsy began very early in life, at two months of age. Children with early onset epilepsy with TSC often progress to IS (Thiele, 2004).

Age of onset of seizure disorder has also been cited as a contributing factor to cognitive impairment in TSC. Humphrey et al. (2008) conducted a longitudinal study of infants with TSC before the onset of seizure activity, and found a pattern of regression in some infants with seizure onset, while in other infants a regression occurred before clinical seizure activity was evident, raising the possibility that these children were experiencing sub-clinical seizure activity before clinical seizures were evident. Zaroff et al. (2006) found seizure onset before one year of age was associated with a poor cognitive outcome. In the current study, five of the children had seizure onset under a year of age, and an additional child possibly had undiagnosed seizures early in life. There is a close association between early seizure onset and poor cognitive outcome, but this was not seen in the current study. In the current study, of the six who possibly had seizure activity under a year of age, three were in the intellectually impaired range, and three were functioning in the average range with support or above average range.

Further, one child who was significantly cognitively impaired did not develop clinical seizure activity until the age of two and a half. There is wide variability in the affects of TSC, and the present results confirm the complex nature of cognitive impairment in TSC.

Joinson et al. (2003) found that people with TSC who fall into the average range of cognitive ability have a mean Intelligence Quotient (IQ) that, as a whole, is shifted slightly to the left. That is, their overall average IQ scores on standardized tests are slightly lower than the average for the general population, so that the curve of scores follows the same curve as for the rest of the population, but it is shifted down slightly. They compared the average IQ scores for a group of people with TSC to their unaffected siblings, and found that their mean IQ was significantly lower than for their unaffected siblings, even though they fell into the normal range of functioning. It is interesting to note that in the present study, there were no children who did not have some sort of deficit in functioning. Even the two children whose teachers felt were gifted had a specific impairment in at least one area. The current results, although from a small and voluntary sample, would seem to support previous research that cognitive deficits in TSC are common, even in those who fall into the average or above average range of cognitive functioning.

Anxiety

Anxiety was a common theme, but it seemed to manifest differently depending on the level of functioning of the child. For example, there were several cases of children who functioned in the average to above average range intellectually who had anxiety about their schoolwork and marks, while several children in the lower range of functioning experienced anxiety as well but more in relation to heightened noise levels

and other stimuli. For example, one teacher of a Grade 8 student noted, "I know she had a little bit of an issue getting worried about not finishing her work, and I think she did have more...and her marks, she really wanted to do well. If she does poorly, she really is hard on herself if she does poorly and she'll cry." In another example, a Grade 3 teacher explained, "when she starts to sense she's not understanding she gets very anxious...She can monitor herself very easily. She knows before I do sometimes when she doesn't understand it, and it does make her anxious." In contrast, a child who was more cognitively impaired was noted to experience great anxiety with certain environments, Her teacher described how going to a self-contained classroom for part of the day causes her anxiety: "the noise level, it's just screaming, it's constant. And you can see that it really bothers her and adds to her anxiety." Anxiety was common, but seemed to be mediated by cognitive functioning levels. This raises the possibility that anxiety itself is a central manifestation of the disorder, and it just manifests itself differently depending on factors such as intellectual functioning.

Muzykewicz et al. (2007) found a high rate of anxiety in the patients they examined in their clinic, particularly in the older, higher cognitively functioning group. They pointed out that many seizure medications can also act as mood stabilizers and could decrease anxiety in TSC patients who are taking them. One mother in the current study reported this in her child's case. She has identical twins with TSC, and the less developmentally impaired child has a severe anxiety disorder. The more impaired twin also has anxiety, but the mother felt her anxiety levels were ameliorated by her seizure medications. She explained: "she does have anxiety, we see similar behaviours between her and [our other child], but with [her] I think because of some of the medications she's

on help with that, that we don't see it as bad. With her she's always been on some type of medication that works with anxiety." Another possible reason offered by Muzykewicz et al. for the lower reported anxiety levels in the more cognitively impaired group was the possibility that this group might be less able to express their anxiety than the higher cognitively functioning group. In one case in the current study, the EA of a Grade 6 student who was interviewed was unable to recognize any anxiety in the student she worked with, even though the mother reported the results of testing by the school psychologist found her to be "very, very high on anxiety". The EA reported that the child was "spoiled" by parents and grandparents. When asked about the child's anxiety, she responded: "I think it's because she's really spoiled. You know, like she's her papa and nana's centre of attention and I know she spends a lot of time there and yeah...She likes to get her way...She doesn't get anxious, she gets mad and stubborn." This suggests that this child was experiencing anxiety that was unrecognized by those who worked with her, who may have just thought she was overreacting in certain situations. This child was quite cognitively impaired, and this raises the possibility that, because of this child's impairment, her anxiety was less recognizable, possibly because her language skills caused her to be less able to articulate her anxiety, so that it is being identified as externalized behavioural problems rather than anxiety per se.

Recent work by Kopp, Muzykewicz, Staley, Thiele, and Pulsifer (2008) found lower anxiety rates in children than studies with adults. It is noteworthy that anxiety levels in the present study were evaluated on the basis of parent and teacher/EA reporting, rather than based on any clinical test or direct observation. It is conceivable that some clinical tests are not as sensitive with this age group. Parents see their children

more often and in more situations that any other individual. It is possible that they may be better able to pick up on anxiety than an outside evaluator who may not see a child for as long. The role of parent reporting then in TSC may be especially important. This may be particularly important for the more cognitively impaired group, who may lack the language and expressive skills to articulate their feelings of anxiety. Kopp et al. suggest that anxiety in children with TSC may be underestimated. This is certainly an area where more research is warranted.

Anxiety is a common manifestation in TSC that parents, teachers and other professionals who work with these individuals should be aware of. Anxiety may impact a child's self-esteem, performance in school and quality of life and the high rates in TSC warrant looking into.

Teachers also experienced what they called "anxiety" about having a child with TSC in their classroom. Their feelings may be more accurately characterized as feelings of a lack of self-efficacy. For example, some felt unsupported in their efforts to accommodate these children, while most felt unprepared and felt there was a lack of knowledge and understanding about the disorder and how it might manifest itself in the child they were going to be teaching. A Grade 8 teacher reported: "When I first heard about it I was just beside myself, I thought, oh my goodness. I guess it was just, you know you kind of focus on the negative. You know, this poor child, and I hope I can service him and meet those needs, right? And this is of course before we even knew what [he] was like. So yeah, we both had a little anxiety. We wanted to make sure he was comfortable in the class, and that other students were educated and that we were accommodating him accordingly." This is consistent with findings of a study of children

with health needs in the classroom that found that many special education teachers felt that training in dealing with special health concerns was inadequate (DePaepe, Garrison-Kane, & Doelling, 2002). These authors found that both special and general education teachers "must have the background knowledge and skills necessary to plan for and provide appropriate educational services, school-related health services, and accommodations for students diagnosed with a wide variety of health conditions" (p. 2). They outline some specific skills that teachers should be trained in to deal with students who have health issues.

The teachers in the current study did not engage in any specialized training to deal with having a child in their classroom with a complex disorder. With one exception, all teachers and EAs reported getting their information from the parents directly, and a couple went on to do some further research on the Internet. No teachers reported receiving any information or preparation from their administration, school board or resource team at their school or board. One teacher reported not having been informed by the parents about the child he was teaching having TSC. When asked if he knew the child in his class had TSC, he said: "Yeah, I heard in passing from another teacher, from her last year teacher." A teacher of a Grade 8 student explained, "certainly, when it comes to learning difficulties, LDs, ADHD that kind of thing, we have lots of info, but when it comes to more rare disorders like TSC, we really don't have anything and really need to do our own research. I think in cases like this the parents are your best ally, your best bet; they know their child best." One teacher expressed serious concern over the lack of information and support provided to plan and accommodate for the student in her class's complex needs. She pointed out that "they never have anything in place for the

first day of school, or for the first marking period. So when we start with these special needs kids in our classrooms, even though the special ed department knows we are getting them, nothing is there for us." In another example, a teacher of a Grade 8 student felt he would have liked more information, "Just to know what to keep my eye on. Things to be aware of." This would point to a possible gap in the education system when it comes to teaching children with complex needs. Teachers and other professionals working with these children may need more information and support to adequately plan for and accommodate these children. Receiving this information before starting to teach the child may help to lessen the anxiety felt by these teachers and may contribute to a more successful experience for these children. Resource teams, administration, school board consultants and outside agencies such as hospitals and epilepsy organizations may have a role in educating those people who will be working with children who have TSC.

Autism Spectrum Disorder and Its Relation to TSC

ASD is primarily characterized by language and social difficulties. Children with ASD are delayed in speaking, and then continue to have difficulties in processing certain aspects of language. They have differences in relating to peers, and often experience difficulty in reciprocal social interactions. Their behaviour is often noted as being atypical, sometimes involving narrow interests and fixation on certain objects, subjects or sensations (Graetz, Mastropieri, Scruggs, & Agosta, 2004). In the current study, differences were seen in behaviour, language processing, and social interactions. These areas of difference may suggest a higher incidence of ASD in the current sample than was reported by participants. Alternatively, these differences may indicate some subtler autistic like tendencies, PDD-NOS (Pervasive Developmental Disorder Not Otherwise

Specified), or may be the result of some other aspect of TSC. In the following sections, behavioural differences, language processing differences and social issues seen in the present sample are presented, along with an analysis of the possible links between these differences and TSC, and between TSC and ASD in general.

Behavioural Differences

Behaviour differences were noted in most participants in the current study (n=8). Five parents reported their child currently or previously had tantrums, where the child would go into an uncontrollable rage. Two of these parents reported that these rages could involve some physical aggression towards family members. Hyperactivity was reported in four of the children, with a definitive diagnosis of ADHD having been made by a doctor in two of these cases. Several children (n=4) in the study exhibited some type of fixating behaviour that had indications of obsessive-compulsive disorder. Children also had behavioural differences such as: repeating what had been said, not understanding social cues, taking language too literally, and difficulty with routine changes.

De Vries et al. (2005) report that many parents feel the behavioural issues in TSC are the most challenging aspects of the disorder to deal with. This was the case in the current study with at least one participant. This mother of a child in Grade 1 described her son's behaviour this way: "he has an anger management problem. It's so bad, we just couldn't deal with it." The behavioural difficulties reported in the present study ranged from a severe anger management problem, in which the child showed physical aggression towards family members, to rages, to oppositional behaviour. The variety of behavioural differences did not vary based on degree of cognitive impairment, although behaviour difficulties in general did seem more common in the more cognitively impaired children

in the study. In one notable exception, a child with average cognitive functioning experienced uncontrollable rages and outbursts. Her mother reported behaviour services were very helpful in this regard in giving her child better ways to deal with her aggression. She states: "[she has] come a long way in a year and a half, since she's had services, prior to that she would go into rages that she couldn't stop."

Surprisingly, several parents and teachers didn't recognize common traits of TSC as being part of the disorder. Several passed off odd behaviours or disorganization as just a personality trait of the child rather than part of TSC. For example, one mother of a girl in Grade 3 said, "I have no idea what she would be like if she didn't have TS, but she's pretty quirky, but I think that's just her. And its funny, when she was on medication, there was all these things we attributed to the medication, but when we took her off it turns out it was just her. *Laughs*...And um, she's funny, funny, funny. But, um, she's moody, I mean, but who knows what's from what." One parent felt that their child's early "clumsiness" and slower to develop motor skills were nothing out of the ordinary. She explains, "She was definitely clumsy, but on the other hand neither my husband or I are super coordinated." One teacher of a Grade 8 student noted that the child he taught had difficulty organizing anything, but didn't recognize this difficulty as a possible manifestation of TSC.

On the one hand, it is conceivable that some parents may have a bit of denial about how seriously the disorder had affected their child, but there seemed to be no emotional reasons for educators to deny the existence of specific TSC related deficits. This again comes back to the lack of knowledge about TSC in general for both teachers and parents. Although the medical findings are certainly well publicized and seemed

better understood by both parents and educators (for example the possibility of seizures) the more subtle differences were not recognized as a feature of TSC. This would suggest the need for more research and education about these features of the disease so that parents and educators could recognize them and try to accommodate accordingly.

Language Processing Differences

Five children in the present study were reported to be late to speak. Two additional children in the study were developing speech fairly typically, and then regressed due to seizure activity. Only one child in the study currently couldn't communicate, and an additional child benefited from sign language. Although several studies have identified language difficulties as common in TSC, regardless of intellectual status (de Vries et al. 2005; Prather & de Vries, 2004), no studies could be found that characterize what these language difficulties looked like and how they manifest themselves.

The language difficulties seen in the current study varied, but several included a specific deficit in receptive language skills. For one Grade 1 child, the primary academic difficulty was reported to be a lack of receptive language skills. In another case, a psychological evaluation had been done that showed scattered language skills, that were low in receptive but, interestingly, high in expressive skills. Her mother explained her deficit was: "not so much with her verbal expression, a lot of people still will meet her and go gee, she does so well, she's such a good speaker, but I'm like: Not! When the SLP report came out, like she had such a scatter on all of her testing that everybody's like oh my god! What a mess! So she comes out really, really high in verbal expression, but then the language is like the reception of language and that kind of thing. And you know

so like understanding two sentence structures or even she starts talking but then she's off on a tangent about other things. So it's more the comprehension, the understanding of language." Thus in this child's case, there was a specific deficit in the understanding of language, but she was able to adequately express herself verbally. Early language difficulties were also seen in the present study. It would be interesting to do a larger scale study to try to further characterize the language deficits seen in TSC. Preliminary evidence from this study would seem to point to receptive skills in particular as being problematic in these children, as well as slower to develop language skills, as was seen in most of the children in the current study.

Children who have low receptive language skills and higher expressive language skills may present a language delay that is more difficult for educators to pick up on, as expressively, these children may be fairly typical, but they may have difficulty processing or understanding what has been said to them. One EA of a child in Grade 6 child noted this difficulty in the child she taught. She pointed out that this child often "talks like she gets it, but she does not. Which is amazing, you know." This child would appear as if she understood, but her receptive skills were sufficiently low, significantly impairing her understanding of social situations and instructions. This may be a specific area that educators need to be aware of, so that appropriate accommodations can be made. Specialized testing, such as a psychological evaluation or a speech and language evaluation may be useful here to pick up on these more subtle deficits that the educator may have difficulty identifying in the regular classroom.

Some of the behavioural differences noted earlier, such as echolalia, compulsions, fixating on certain things, not understanding social cues and taking language too literally,

combined with the aforementioned speech and language delays, are often cited as symptoms of ASD (Graetz et al., 2004), but only three parents reported their child had an ASD diagnosis. This brings up questions about the prevalence of ASD in TSC children. Studies have varied widely, with some reporting the occurrence of autism in TSC as high as 68 percent while other studies have reported it in as few as 17 percent of cases (Smalley 1998). Baker et al. (1998) report that earlier studies into ASD did not use a rigorous standard of ASD testing. This begs the question: what testing standard, if any, was used to evaluate the children in the present study? Might more have been diagnosed with an ASD under certain testing conditions, or would it be more accurate to say they do not have ASD but have a milder form of ASD such as PDD-NOS (pervasive developmental delay not otherwise specified)? Is ASD more common in TSC than has been thus far reported in the literature, or are behavioural differences that include components of ASD being seen? Alternatively, are some of the features seen in this study part of a non-verbal learning disorder? All of these questions are worthy of further investigation.

It would have been interesting to find out how, or if, all children in the current study were evaluated for the possibility of an ASD. Some of parents in the present study were unsure of their child's ASD diagnosis. One mother reported an early diagnosis of ASD, but thought that diagnosis was overturned in favour of the TSC diagnosis. Another mother in the study was unsure if her neurologist had confirmed ASD or not, but thought he may have given that label. Teachers were also not always informed about the possibility of ASD in the children they taught. One EA was unaware the child she worked with was autistic. This would seem to highlight the need for children with TSC

to be formally evaluated for ASD and for parents and educators to then be informed of the diagnosis. De Vries et al. (2004) recommend all children with TSC be evaluated in the toddler years and preschool years for developmental disorders such as ASD.

Educators and paraprofessionals may need to be aware of the possibility of children with TSC having an ASD and adjust their teaching methods and accommodations accordingly. Even if a child does not qualify for an ASD diagnosis, teachers might need to be aware of the strong possibility that these children might display some autistic tendencies and plan for accommodations to suit their behaviours. There are many good resources about accommodating and teaching children with ASD that might be appropriately adapted to fit into the complexities of the child presenting with TSC (see for example Ruble & Dalrymple, 2002).

Social Interactions

A more subtle behavioural difference was noted in several of the children in this study. In three of the children in this study who were reported to be cognitively average to above average (one of whom was in a gifted program), their teachers reported that they got along with other children well and had friendships but did not have one particularly close bond with one individual. Although there certainly is a range of normal when it comes to childhood friendships and appropriate bonding, it is, nevertheless, interesting that several children whose teachers reported to be socially normal, failed to have one close significant bonded friendship and rather chose to get along with everyone and have diverse friendships. One teacher of a Grade 8 student noted "you know how girls they have their own little groups, so I haven't really seen her fit into a specific group, I think she's kind of like friends with everyone…I've seen her, I would say there's three

different groups, I've seen her talk to all three groups, you know on several occasions." This theme was echoed in another teacher's description of the Grade 3 student she taught: "She does [have friends], but not super close. She gets along with everyone, but I wouldn't say she's formed an especially close bond with any one student." Could this be a subtle social deficit exhibited by those children with TSC who fall into the average range of cognitive ability? Is there some social awareness or skill lacking in these children that prevents them from forming a close friendship or from wanting a close friendship? Several of the teachers and parents described the children in the study as marching to the tune of their own drummer, or quirky. One teacher described her student's behaviour this way: "she definitely marches to the beat of her own drummer. Um, not in an anti-social way...Um, but she defiantly runs with her own ideas and doesn't necessarily need others to join her in order for her to enjoy her own ideas." Could it be that some aspect of social referencing is missing or not adequately developed? Again, this is purely speculation and could easily be argued as being part of regular individual differences that might be found in any sampling of a population, but it may be a topic worth further investigation.

One mother reported that her child, although reported by all accounts to be normal cognitively and socially average, seemed oblivious to the school bully's intentions and instead disarmed him by attempting to befriend him. It seemed as though this child was unable to recognize certain social cues. In this case it seemed as though this lack of understanding of subtle social cues had served this child well, allowing him to befriend a variety of children and avoid a potentially difficult bullying situation, but it is possible that in some other cases this might be a social deficit that may be a hindrance to these

children. In another example, a reportedly cognitively average Grade 3 child's mother reported that one of her main problems was social interaction. She reported that her child had difficulty understanding social situations, particularly those in which negative interactions were generalized to the child thinking that the other children no longer wanted to be friends. As she explains, the most difficult area for her child is: "you know, socialization, and that if a friend says something to you it doesn't necessarily mean that they don't like you, they might just be having a bad day, you know just that part of understanding what friends mean when they talk to you." It seemed that for this child, language was taken too literally, even in the absence of cognitive delay.

Feedman and Silverman (2008) argue that one of the biggest areas of concern in children with high functioning autism (HFA) is a poor understanding of other people's intentions and a too literal interpretation of language. Given that Baker and Piven (1998) found some children with TSC who do not fulfill the criteria of autism still display some of the characteristics, could there be a subset of children with TSC who are in the normal range of functioning, but who have a specific deficit in social behaviour? Could it be that there is some measurable deficit in these children's understanding and interpretation of subtle social cues and language? Kopp, Muzykewicz, Staley, Thiele, and Pulsifer (2008) recently reported that the behavioural differences found in their sample of children with TSC were often autistic-like and included a lack of awareness of others and socially withdrawn behaviour. Perhaps this lack of awareness of others manifested itself in a lack of social referencing in the present study.

It is possible that children with TSC, even the apparently cognitively average ones, might benefit from some type of social training similar to that in use for autistic

children, to develop their skills in reading social cues. Freedman and Silverman (2008) suggest intense social skills training can be beneficial to children with HFA. It is possible that children with TSC who appear to be cognitively average might benefit from this type of intervention as well. Social stories have been shown to be beneficial to children with ASD in helping them to recognize and interpret social situations (Graetz, Mastropieri, Scruggs, & Agosta, 2004). Perhaps social stories could be adapted and used with this population as well. Certainly it is something that educators and school psychologists should be made aware of to monitor. This would seem to be in line with what de Vries et al. (2007) were referring to in their examination of psychopathologies in TSC; they note that even in the most cognitively "able" children with TSC, clinicians should pay careful attention to whether they need to consider diagnosing ASD, mood, language or hyperactivity disorders. They note that regardless of intellectual status, there appears to be an increased risk that children with TSC will potentially have a variety of neurocognitive disabilities (de Vries et al.).

Freedman and Silverman (2008) note that children with HFA often have "subtle deficits, particularly in terms of self regulation (adapting to a difficult situation), changing routines, and organizational skills" (p. 64). These differences in organizational skills and difficulties with routine changes were certainly seen in the present study, again bringing up the possibility that some type of ASD may have been more prevalent in the present study than reported by parents and teachers. Prather and de Vries (2004) note that children with TSC often have difficulty with transitions and changes in routines, and propose that this may be due to differences in frontal lobe functioning in these patients. They reported that sometimes children with TSC might overact to something as simple as

a routine change, and that their reaction may be seen as out of proportion to the event. This "overreaction" may be a sign of undiagnosed ASD, or autistic tendencies.

Interestingly, recent work on mouse models of TSC has found that mice that have been bred to have a TSC mutation (known as TSC knockout mice) have difficulty engaging in certain social behaviours even in the absence of lesions in their brains and epilepsy (Goorden et al., 2007). In this study TSC knockout mice that didn't have epilepsy or overt brain lesions were compared with wild type mice in several experiments to test social behaviour and cognitive functioning. In one test, mice were placed in a cage and allowed to habituate, and then two novel mice were placed in the cage with them. This test evaluates social interaction. Normal mice will approach and interact with the novel mice. In the TSC knockout mice, this interaction and approach was significantly impaired. In another test of social functioning, mice were given nest-building materials. The TSC mice were again significantly impaired in this normal social behaviour (Goorden et al.). Although nest building and novel mouse tests are not completely relatable to the much more complex human interactions of making friends in children, it is interesting to note that something in the TSC mice is preventing them from behaving like the normal mice. Goorden et al. point to neuronal functioning as a possible culprit in these deficits, rather than overt brain pathology. This would echo recent work by de Vries and Howe (2007) that suggests we need to look beyond tubers for the cognitive deficits in TSC. De Vries and Howe hypothesize a molecular process that may inhibit the normal cognitive and social development of children with TSC. Likewise the children in the current study may have some genetic, molecular, neuronal or as yet unknown commonality of TSC, which makes their social interactions atypical.

Educational Programming

Academic Difficulties

Academic difficulties were reported in several areas. Fine and gross motor functioning (n=3), and difficulties in such areas as math, language (n=5) were common. Memory difficulties were reported in two children, while organization difficulties were reported in three. Accommodations currently implemented for the children in school were reported to be: IEPs (n=6), repetition (n=2), one-on-one instruction (n=2), and visuals (n=2). Four of the children had a full time EA, and one child also had a full time nurse. Two of the children participated in a totally modified program. One of these children was pulled out of the regular classroom to a resource teacher, as well as pull out to a living skills program, while the other child was pulled out for an hour every day to work with an autism teacher in a self-contained classroom. The other four children participated in regular classrooms. One of these children was pulled out to participate in a gifted program, and another child had been referred for gifted testing. One child participated in a regular program with a full time EA's support. One child participated in a regular program with accommodations only for motor difficulties and attention issues. Two of the children were still receiving OT and PT support at school, and two were receiving behaviour therapy in the school. One child participated in music, art and sensory room therapies.

Three teachers/EAs reported that a psychological or speech and language assessment had been done. One child had a psychological assessment done that highlighted recall difficulties, one child who had a speech and language assessment done had issues in both receptive and expressive language reported, and the other child with a

speech and language report had reported scattered testing results that were high in expressive language skills but low in receptive skills.

Six children in the present study had an IEP. For several of these children, the IEP outlined a modified program with goals in social domains, living skills and basic functional academics. Children with active epilepsy also often had medical components to their IEPs. One mother of a Grade 6 student reported that her child's IEP had to contain mostly medical information as her disability was classified as a medical disorder for the purposes of funding for EA support and other supports within the school. This child's IEP was reported to contain much less academic goals than it previously had, as the result of changes to the provincial funding system that necessitated the change to a medical classification. Her mother explains, "it's so political, like her IEP used, like it would have the normal like, will be able to write eight sentences making sense, you know, with a picture prompt, but because Alberta did this huge, huge audit on any special needs funding in schools, M's code is a medical disability code, so her IEP needs to match her code to get the money." This seemed somewhat counterproductive. Although the medical components of the disease are certainly important and should find a place on the IEP, the learning needs also need to be clearly set out.

One child had only fine motor and testing accommodations outlined on his IEP, while another child had such accommodations as repetition and use of visuals as important considerations on his IEP. Behaviour services, either from a consultant coming in and working with a child or from goals on the IEP were important considerations for several children in the study. Thus there were a variety of needs and issues highlighted on these documents, as might be expected when looking at any group of children with

special needs. However, it would have been interesting to include an examination of these documents into the present study, to further look for similarities and differences.

Accommodations

Several accommodations emerged as important for the children in the current study. Repetition and re-teaching were cited as important strategies by four teachers/EAs. Two of the children were reported to have memory issues, and had difficulty with new or unfamiliar material and routines. An EA of a child in Grade 1 explained it this way: "I think the biggest issue with him is memory and retaining, so it will take him a few repeats of a routine or something like that in order for him to solidify it. So, you know, it might take one kid one or two times to get something, it might take him five or ten times depending on the complexity of the task." She explained that when new concepts are being taught to the class, the child needed much more support. She explains: "basically the way we go about it is the teacher will teach the whole class, and then the students will probably get an assignment to work on, so I'll sit with him and basically re-teach the lesson in the context of the work....So whereas other students will get input once and then they'll apply it, he'll get input and then get input again." A teacher of a Grade 3 child in the study explained that things needed to be "constantly reinforced everyday. It needs to be re-taught everyday." In another example, a teacher of a Grade 3 student who was described as being very bright and excelling in all areas noted that "Sometimes a new or unfamiliar situation can take her a little more time to figure out...something that's unfamiliar can be a little bit frustrating for her in the beginning."

One child in Grade 8 needed repetition and re-teaching to fill in gaps that developed in his learning due to his attention difficulties. His Grade 8 teacher explained:

"he did well in every single subject, however, I think the thing with [him] is that often what would happen because of the focus problem, often there would be gaps that would need to be filled in. Otherwise he would have trouble with an assignment or evaluation. So what you would have to do, he would benefit from that extra time coming in and getting that extra support, just filling in that gap, you know? Maybe on one day, you know on a rougher day, he would miss maybe a good third of a lesson, so, you know, we would review the day after, on a better day to fill in that gap."

These findings are not surprising. De Vries et al. (2005) found memory issues were common in TSC, regardless of intellectual functioning. Teachers and others who work with children with TSC might benefit from being made aware of potential memory and retention difficulties in these children and may need to find ways to accommodate for this need through re-teaching and repetition of routines until they become familiar and comfortable.

The use of visuals, providing material in a visual format as well as orally when teaching, was also an important accommodation for this group. This may be due to subtle language processing difficulties, or it may be connected to an under diagnosis of ASD or non-verbal disability, as the use of visuals is often recommended as an effective strategy in these disorders. These children may benefit from receiving information in more than one way (verbally and visually) to help them to process messages. In one case, a child in Grade 3 also benefited from sign language to help reinforce verbal messages. Her teacher notes, "Mostly visual. It has to be visual with [her]. And we do use sign language, but mother has taught us. I guess when [she] was young, she wasn't able to speak for a number of years, and they found that using sign helped. And she does act

quicker with sign, instead of just verbal instruction." Again, educational professionals should be aware of possible gaps in receptive language skills in these children, and the possible need for visuals to be used as an accommodation.

Some of the children in the study were reported to do better with one on one instruction. A mother of a boy in Grade 1 felt that her child benefited more from individual teaching, but that the school did not use enough one-on-one instruction. She explains: "A one to one teach, like we do at home...I think the teachers don't do that. What they do is a group thing every time right?" This seemed especially true of the children in the present study who were more cognitively impaired. Perhaps this comes back to Harrison et al.'s (1999) observation that children with TSC often have difficulty with set-shifting, that is, shifting their attention from one stimulus to another. These children might benefit from individual attention and teaching because they then can focus on one stimulus, and not have to try to set shift between the teacher and other distractions such as other students. One teacher noted that when her student went to a self-contained classroom for part of the day, she experienced much anxiety because of the other children in the room and the noise level. This teacher felt that her student would benefit from more individual attention, rather than participating in a group situation in a self contained classroom. Allocation of personnel and resources to accommodate needs such as individual attention for children with TSC should be made on an individual basis, but educators and administrators might do well to be aware of the possibility that more individualized attention may be needed.

Note-taking and scribing were also important accommodations, which is not surprising given the number of gross and fine motor issues found in this study. One

parent felt that although her child currently didn't need any accommodations for fine motor skills, being in Grade 1, her fine motor skills were extremely low, given the amount of fine motor intervention the mother and other programs had given her. She felt that if she hadn't given her child as much practice with fine motor activities "they would be even lower. When you think about her overall IQ, we're going to probably have to help her later on. So I mean the computer will be huge for her." Another mother reported that note-taking and writing activities were really holding her child back academically, and that once that was recognized and accommodations made for it, his performance improved greatly. Assistive technologies such as personal computers and speech to text software may have a role for children with TSC, given the high proportion of fine motor issues, and teachers and parents should evaluate this possibility.

Organizational Skills

Organizational skills emerged as a theme in this study. This probably falls under what de Vries et al. (2005) call executive functioning skills. Several children were noted to have difficulty with organization, and needed assistance to remember sequences of tasks, such as packing a school bag, or steps in getting ready to go out. Following multi-step directions was challenging for several children in the study. One parent reported that her child, who was cognitively average, needed direct support in organization, even at the age of 13. She reported needing to have laminated cards prepared for him to remember even the simplest tasks such as bringing keys, packing his bag and other daily tasks. Simple organizational tools and supports may be needed by children with TSC in the classroom to facilitate their independence. Such things as visual schedules and cues to

remember certain common sequences of events may be beneficial to these children and might be considered by teachers and other professionals who work with these children.

Difficulties in organization were not dependant on intellectual status in this study. The cognitive functioning of the children in this study who reported difficulty with organization ranged from very impaired to gifted. Although most teachers were informed about the diagnosis of TSC and many were informed about the medical issues, not many teachers mentioned the child's organization as something they worked on or accommodated. In fact one teacher did not realize that organization difficulties were a possible result of TSC, and instead believed it was the result of the child's gifted status. When told that organizational difficulties were common in TSC, he said, "I never thought of it like that. I thought of it...I knew that she um, she was gifted, and over the past few years I made the correlation between gifted and organization. They have a...I guess their forte isn't really organization...I didn't even think about her condition, that's a good point." It would seem then, that teachers might benefit from being made aware of some of these subtler aspects of TSC, and that many children with TSC, regardless of cognitive status, may need some direct instruction, modeling, and scaffolding in organizational skills. This again points to a possible need for educators to be well informed about all the possible consequences of TSC, and being as well informed about possible appropriate interventions that may need to be tried regardless of intellectual status.

Interestingly, a couple of parents reported that their children were hyper organized instead of having difficulty in organization. They were bothered by things that were out of place, and had an almost obsessive need to have things organized. A mother of a child in Grade 1 reported: "we actually think he's a little OCD because he's so organized. Not

necessarily in a bad way, but if something's on the floor, he'll pick it up." Another mother noted of her child, who was also in Grade 1, "You know, her playroom, her toys, she puts them back in the right positions. She doesn't like a messy playroom, bedroom, things like that." Obsessive-compulsive disorder has been reported to be a common manifestation of TSC, so it is possible that this type of behaviour might be due to an obsessive disorder (Au et al. 2007). Teachers and other professionals should also be aware of this possibility, as anxiety may be caused by not allowing these children to complete certain organizational tasks.

Teacher Perceptions of Making Accommodations for Children with TSC

Some teachers reported they felt accommodating for the TSC child in their classroom was the same as accommodating other special needs children they had taught in the past. One EA of a Grade 1 student reported that: "I think any child that had, I guess, the language delay that he has, I would make the same accommodations. It's because of the language piece, not specifically because he has TSC, that I make accommodations. The language delay just happens to be the outcome." Some teachers, however, felt that accommodating for the child with TSC differed from other special needs they had encountered. For example, a teacher of a Grade 3 student pointed out that: "Her environment has to be different than most other special needs students that I've had...because of the health issues...With the blood pressure and the seizures, that's what we worry about the most... I'm saying that I know she's different than a regular kid. And she has to be treated a little bit differently...I think a lot of us are bitter with having all of these special needs kids placed in our classroom. So that a lot of the regular ed teachers are treating the special ed kids like they were regular ed kids. And in doing so,

it's hurting the special needs child." With the additional medical complications, it could be argued that although some of the accommodations for the cognitive manifestations of TSC might be similar to other children (for example one child in the study had a main cognitive manifestation of ADHD, and the teacher felt the accommodations she made for that child would be the same as they would be for any child she had in her classroom with ADHD) there are additional variations and complications that cannot be overlooked. That same child also had some complications due to a changing brain tumour that required additional vigilance and supervision, certainly not the average for a special needs child that may present in a typical classroom. While good teaching strategies and accommodations for specific cognitive impairments are likely good universally, regardless of the cause of the cognitive delay, the scope of accommodations and the additional complications from medical issues necessitates a different constellation of accommodations for the child with TSC.

Medical Issues in the Classroom

All educational personnel who are in contact with these children might benefit from being made aware of the possibilities of new or different seizure activity, heart difficulties, kidney failure, complications due to tumour changes and other possible physical complications of TSC. For example, almost all of the children in the current study had heart findings, although most were not symptomatic. Regardless of the current ramifications of cardiac involvement, this is an area where professionals working with these individuals may need to be vigilant and should be informed of the possibility of a heart lesion becoming symptomatic. In the current study, only one teacher reported any knowledge of vascular manifestations. In that particular case, the child had high blood

pressure and needed to be monitored frequently throughout the day to ensure her blood pressure remained in a tolerable range. Professionals working with these children might benefit from being aware of what the sudden onset of cardiac symptoms may look like so that they can watch for these potentially dangerous signs.

Vigilance in monitoring for seizure activity is consistent with DePaepe, Garrison-Kane and Doelling's (2002) recommendations regarding epilepsy in the classroom. They note that teachers must be aware of a child's potential seizure activity. Academically, students with absence seizures are at higher risk for academic difficulty as they may miss content during brief seizure episodes. Children experiencing other types of seizure activity such as partial or complex partial (particularly common in TSC) may be confused or disoriented after seizure activity and may need reassurance after a seizure. Another important reason for teachers and other school personnel to be vigilant when it comes to monitoring seizure activity is that continued and poorly controlled seizure activity is associated with poorer cognitive outcome (Winterkorn, Pulsifer, & Thiele, 2007). Thus it is important that educators and other personnel working with these children be educated and aware of what to look for with seizure activity so that this information can be documented and reported back to parents who can inform doctors managing these children's health. Increased seizure activity may point to the need to change or increase medications, and given that school personnel are likely to spend the better part of the day with these children, they may be in the best position to see seizure activity develop. Given the potentially negative cognitive consequences of uncontrolled seizures, teachers may have an important role to play.

In the current study, most children with active epilepsy had a medical component to their IEP. DePaepe, Garrison-Kane and Doelling's (2002) emphasize the importance of having a medical plan in place in the child's IEP for children with complex medical needs. In these documents, there were reported to be special procedures in the event of a seizure. Seizures lasting more than five minutes can cause permanent brain damage, so most plans included a special procedure in the event a seizure lasted this long. These plans included such things as evacuating the other children from the classroom, designated personnel to call emergency medical services, and designated personnel to administer emergency seizure medication to the child. Having a medical plan in place may be a necessary precaution in the case of a complex disorder such as TSC, so it was encouraging that many of the children had one. It was somewhat surprising that some of the children did not have a medical plan in place. It could be argued that even children without active seizure disorders should have a medical procedure in place in case a seizure disorder were to present itself. In the case of TSC, epilepsy can present at any stage of life, and being seizure free, even for a number of years, does not preclude the possibility of seizures re-appearing at a later time.

Advocacy

Advocacy emerged as a strong theme in this study. Six of the parents and one teacher noted the importance of strong parental advocacy in getting services for their child.

Parental advocacy is important when dealing with any medical condition (Whitehead & Gosling, 2003). It is possible that advocacy is even more important when dealing with a rare disorder such as TSC, in which educators and even medical

professionals may not know much about the disease and its possible ramifications. Whitehead and Gosling found that parents in their study only achieved a diagnosis, testing and access to services for their child with TSC through strong advocacy. They note that in the case of TSC, medical professionals often do not have expertise or knowledge of the disease, and parents must act as experts. Whitehead and Gosling note that a "collaborative relationship between parents and professionals has been thought to be important to disabled children, particularly for emotional support and joint teaching and learning" (p. 117).

The role of a collaborative relationship between parents and teachers in the case of TSC seemed very important in the present study. Given that none of the teachers/EAs in the present study had received any training or information from anyone outside of the parents, this relationship would seem to be the teacher/EA's only avenue of information and support. Ruble and Dalrymple (2002) also note the importance of a collaborative model in planning and implementing programming for children with ASD. They found that consultation from a school psychologist or educational consultant delivered in collaboration with parents and teachers was the most effective way to successfully plan IEPs. This collaborative framework might be appropriate for adaptation to TSC, even in cases without an ASD diagnosis. In one case in the current study, the teacher did not feel supported by any of the school consultants or specialists, and so collaboration was not effective. This teacher felt that she would be much more successful if given the opportunity for more collaboration.

One mother in the study reported having received a full time EA for her child, now in Grade 1, through advocacy to her board of education. As she put it, "you have to

be a very strong advocate...You know the squeaky wheel gets the oil, not that you want to be that, but you need to make it sound worse than what it is sometimes." This sentiment was echoed by a teacher of a Grade 3 student in the study who noted, "I think the louder the parents bark, the more they get." Another parent of third Grade twins reported: "I don't want to say it was just given to us. There was a lot of pushing and advocating on our side to get her to where she is now." This raises the issue of what happens in cases where the parent is unable to be a strong advocate? Some parents may be unfamiliar with the education system and possible services and accommodations that may be worth advocating for. A Grade 3 teacher echoed this sentiment, "I don't think they know everything available they could possibly get." Some parents may have a language, intellectual or other barrier to prevent them from acting as their child's advocate. In these cases, teachers may have a role to play. It would be valuable for educators and other professionals to have more information available to them about TSC and the possible cognitive and academic deficits that are associated with it, so that they can help to ensure that children with TSC in the education system are receiving appropriate supports and services. Educational professionals may need to be vigilant for any area of difficulty, and may need to be prepared to plan accommodations accordingly and to refer to other professionals such as OT/PT, speech, and school psychologists.

Parent Reporting and Teacher/EA Reporting

For most of the participants in the current study, the information provided by teachers and EAs confirmed and supported information provided by parents. Unfortunately, there were four cases in which information from a teacher or EA was not available.

There were, however, a couple of points in the current study in which parent reports and teacher reports did not match. As discussed earlier, in one case a child was reported by her mother to experience great anxiety, but her EA reported she did not experience anxiety. In another case, a boy in Grade 1's mother reported math to be his area of greatest academic concern, but his EA reported this area to be receptive and expressive language skills. This is an interesting, and unexpected finding in the present study. It might point to a need for better communication between school personal and parents, or it might represent a lack of understanding of the manifestations of psychological symptoms such as anxiety in the first case, and a lack of understanding of academic concerns in the second case.

For the purposes of this study, the child with the conflicting anxiety reports was counted as having significant anxiety, as it was felt that her mother likely understood her behaviours and feelings better than her EA, and because the mother reported psychological testing that confirmed anxiety. It is interesting that the school psychologist had done this evaluation, so the results would presumably be in the child's school records, and yet the EA was unaware of this child's difficulty in this area. This same EA was not aware of an earlier autism diagnosis. The mother reported her child had been diagnosed with ASD, but then thought that label had been removed as a result of her being diagnosed with TSC. This comes back to the earlier discussion about the actual prevalence of ASD in the present sample. Behaviour reports by the EA certainly seemed to support an ASD diagnosis, for example she reported that: "I have noticed once in awhile, the teacher will read a story about mustangs, and it could be the horse or it could be the car, but she'll neigh... And she gets stuck on things... and it's repetitive *laughs*

very repetitive!" These reports, taken together with the mother's report of an earlier diagnosis of ASD, and the language processing difficulties, would seem to point to ASD. It is interesting that the EA would not have picked up on this and accommodated as if there was ASD. As an EA, she probably has worked with many children with ASD in the past. In the second case, the child's greatest area of academic concern was recorded as language skills, as it was felt that the EA would probably be more familiar with what was academically difficult for the child, as she would see him in an educational setting, compared to his peers, and his mother may not see him in this setting. It would have been interesting in this case to have evaluated formal testing results, such as a psychological educational evaluation, to see which area came out as a greater area of need.

Limitations

As with any research, this study has several limitations to the validity of the results. Only six of the ten teachers or EAs who worked with the ten children in the present study could be contacted and interviewed. So for four of the cases, only the parent was interviewed. This lessened the strength of the data collected about these children, and in some cases, information about specific accommodations and programming was unavailable. Medical and seizure activity of all participants was reported by the parents from their memory or their own records. Thus information was not confirmed by any third party or medical professional. It would have been interesting to examine the medical records of the participants to get a more accurate understanding of seizure type, age of onset and other manifestations of the disease. For example, one participant reported that her child had many different seizure types, but she did not know

the exact types. This child was likely the most significantly cognitively affected of all the children in the study, and it would have been interesting to know if there had been a history of IS in this case. Much research has looked at the importance of infantile spasms in later cognitive functioning (Jansen et. al., 2008). It would have been beneficial to get a more accurate number of the participants who had experienced IS and to see if that related to cognitive functioning in their case.

There was an inherent selection bias in this study, as only those people who volunteered to participate were included. The invitation to participate was only mailed out to those families who are members of TSC Canada, and who are on their mailing list, and was posted on websites and a neurology clinic. Thus, it may not have reached a representative sample of people with TSC. It is possible that some common feature causes certain families to participate in such a study and certain families to decline participation. This may have skewed the results of the study. Sample size for the interview participants was small, limiting the generalizability of these results. However, that is why a qualitative understanding was selected for this study.

Due to the rare nature of the disorder, participants were sought in both the United States of America and in Canada. There was the possibility that the educational experience would differ from province to province, and between Canada and the United States of America. However, the educational legislation in the Ontario, Alberta, and the federal Education legislation in the United States of America are similar, in that they all mandate the provision of special education services and programming to exceptional students, and all legislate these services be provided in the local school where possible (Alberta Ministry of Education, 2003; Ministry of Education, 2005; & U.S. Department

of Education, 2006). Given these similarities, for the purposes of this study, the educational issues examined were felt to be similar enough to allow for meaningful data collection, despite the diverse geographical areas in which participants lived.

Another possible area for bias is the interview procedure itself. The interview questions may have been leading or may have elicited certain responses from the respondents. To ameliorate this possibility, a thesis supervisor and ethics review board reviewed all questions before conducting the study to identify possible question bias, and approved the semi-structured question list. Researcher bias may have played a role in the current study as I have a vested interest in the study of TSC, having a daughter with this disease. However, it is my belief that this strengthened the results of the current study, as participants may have felt more comfortable sharing their potentially painful and difficult stories with me since I was researching from an insider's position. As discussed in the introduction, participants tended to seem much more comfortable in their sharing, and in fact seemed to relish the opportunity to share, once they learned I also had a daughter with TSC. The thesis supervisor acted as a reliability checker to compensate for any possible parent bias that may have been present in the study.

Due to the rare nature of the disorder, all interviews were conducted by telephone, as all participants lived at a distance. This may have been a limitation to the study, as it may have limited the responsiveness on the part of the researcher during the interview process. It has been suggested that much of a communication occurs through body language, and this was not possible during

the telephone interview situation. It would have been interesting to conduct some of the interviews in person to see if there were any differences in the results.

All data collected in the course of this study was from the point of view of those closest to the children with TSC. Thus, their educational experiences, and effects of TSC, were understood from the perspective of their teachers, parents and EAs. An understanding from the point of view of the children and their perspective on their educational experience was beyond the scope of this study, but would be an interesting area for further research. In-depth interviews were only conducted with a small number of participants. Further detailed interviewing of more families and teachers of children with TSC would be a logical next step for future research.

Conclusions

Research in TSC has historically been done from a medical framework. This makes sense, as many of the physical symptoms are not only serious, they can be life threatening. Gaining control and understanding of the physical processes of this complex disease will benefit all people affected by TSC. However, de Vries et al. (2005) have noted that, despite the serious medical complications that arise from TSC, parents and caregivers often report their greatest concern in dealing with their child is the behavioural components of the disease. After the medical concerns have stabilized, parents' concerns may shift to the schooling of their children. Children spend a great deal of their time for many years in an educational institution, and it is thus important to make this experience as successful as possible for all children, including those with complex disorders such as TSC. Despite some of the limitations of the present study, the strength of it lies in the fact that this is the first study to examine the educational impacts of this disorder.

Several important themes emerged in this study, including: functional issues such as anxiety, regardless of intellectual status, and cognitive functioning; the relation between TSC and ASD, including behavioural, linguistic and social differences; educational programming issues, including specific academic difficulties, accommodations, and medical issues in the classroom and the importance of advocacy; all of which suggest possible areas for further inquiry. Including documentation such as formal testing and program specifics such as IEPs would add to the understanding of how TSC has affected children in school.

Clearly, more information may be needed for teachers, EAs and other professionals and paraprofessionals who work with children who have TSC. As these children are integrated into mainstream education classrooms, all professionals and paraprofessionals may benefit from information about common manifestations of the disease, how it may present itself in the classroom, medical concerns to be vigilant for, common accommodations that can be implemented to facilitate success and important indicators to watch out for that may signify the need for further testing and intervention. Since there is such a wide variation in the possible manifestations of TSC, understanding each child's individual case and situation within the context of the disorder may be valuable for educators.

In the current study, the people that worked with children with TSC were provided with no special training or information about the disease, manifestations or things they may need to watch out for. All teachers, except one, reported getting their information about TSC from parents directly, and a couple of teachers reported going on to look up information on the Internet. In several examples, a lack of understanding of

the manifestations of the disorder seemed to impair the successful integration of these children. For example, not recognizing the difficulty in organizational skills, and the possible need for assistance in this area; not recognizing fine motor issues and referring for OT/PT; and not identifying anxiety all may contribute to these children not being as successful as they could potentially be. Teachers and other professionals who work with children with TSC might benefit from being made aware of the potential manifestations of the disease and common difficulties and deficits, particularly the more subtle ones that can present in the higher cognitively functioning group. Teachers may also benefit from being aware of the high degree of variability of manifestations of TSC, so as not to stereotype, but rather to focus on the individual needs of the student. Any student with TSC who shows differences in motor functioning, social skills, anxiety, memory and retention may need to be referred for further testing and intervention to the appropriate professionals. Early interventions in other genetic disorders has been shown to be important to eventual cognitive outcome (Humphrey et al., 2004), and the same may be true for children with TSC. Identifying the specific area of difficulty experienced by the child with TSC through testing and evaluation by professionals may help teachers and other educational professionals to better program for these children and better accommodate their specific needs. It is reasonable that, in keeping with de Vries et al.'s (2005) recommendations, all children with TSC who reach school age may benefit from having a full psychological - educational evaluation done to identify potential problem areas and subtle differences early. This would seem reasonable given the results of this current study and others that have shown areas of academic need and cognitive deficits in particular areas. Since there is such wide variability in the manifestations of TSC, it may

be helpful to have testing such as psychological evaluations done to identify the specific learning profile of each individual child with TSC. Several parents reported their children had psychological educational reports done, and all showed interestingly scattered results. It would be interesting to conduct a larger scale study of children with TSC and conduct psychological educational testing as part of the research. This might enable more similarities to emerge in learning profile of these children.

This is an area that parental advocacy may play an important role. Whitehead and Gosling (2003) reported the importance advocacy played in getting a diagnosis and appropriate medical care for children with TSC, and the same may be true for children with TSC in the education system. One teacher in the current study spoke directly to this point, noting that in the past, parents who advocated more strongly were able to get funding for their children to get computers and other important accommodations. One parent noted that it was because of strong advocacy that she secured a full time EA for her child. Another parent named advocacy as the reason she was able to get a full psychological educational evaluation done on her child as she entered school. Specialized testing, such as psychological education reports, are done by psychologists who work with school boards, and they have notoriously long waiting lists. Testing is often done on the basis of need, in a triage model of service. Thus, children who may have only subtle cognitive impacts from TSC may be much lower of the waiting list and seen as a lower priority. Given the importance of early intervention, and the range of possible manifestations of TSC, parents may need to take on the role of strong advocate early and push for such specialized testing that might catch a subtle deficit that a teacher or EA may miss.

Differences in social behaviour may need to be examined, and coaching and intervention may be considered to facilitate social interaction. Strategies that have been found to be effective with ASD such as social stories and intensive social skills training may be of use here, regardless of whether or not the child has a definitive diagnosis of ASD. Providing training and information to teachers about the potential for seizure activity and other potentially serious physical manifestations such as heart rhythm difficulties, kidney failure and growing brain tumours may help educators feel more at ease having a child with TSC in their classroom, and may be important for the safety of the child with TSC. This information might be of value to educators regardless of whether a child has experienced any of these manifestations to date, given the nature of the disease, and the tendency for medical concerns to change throughout the lifespan of individuals affected (Curatolo, Verdecchia, & Bombardieri, 2002). A medical plan might be considered for inclusion into the IEP of affected children, in keeping with the suggestions of DePaepe et al. (2002). This would be useful given that serious complications, such as seizure disorders, can appear or worsen at any time. Having a medical plan in place may be valuable to ensuring the safety of these children and may provide valuable information for anyone working with the child.

Although most of the available literature and information about TSC is from a medical framework, the results of the current study show there are potentially challenging academic and social issues that can present in the classroom. With the current movement towards inclusion of students with a variety of special needs into mainstream classrooms, educators need to be prepared to best meet the needs of all students, including those with complex needs, such as those with TSC. Therefore, more research into this side of TSC

is needed. The current study was meant to be a first look into the educational issues facing children with TSC, and it raises several areas where more research should be pursued. The present study should be carried out on a larger sample. Information regarding the physical manifestations from medical records might add to the information gained in this type of study. A closer look at learning issues is warranted, and may be accomplished by conducting psychological educational testing and including information from documents such as IEPs into a study design. The ramifications of the information currently available, and not available, to educators about TSC and its' possible manifestations may be an important area to research in this population.

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Appendix A

The University of Western Ontario, Faculty of Education

Western

Volunteers wanted for a research study about the Educational Impacts of Tuberous Sclerosis Complex

Do you have a child in Grade One to Grade Eight who has Tuberous Sclerosis Complex (TSC)?

If so, you might be eligible to participate in a study about the educational impacts of TSC.
This study will involve an interview with you about your child and his/her educational successes and challenges, as well as an interview with your child's current teacher.

To learn more about this study, please contact:

Jennifer Flinn at jflinn@rogers.com or (613) 823-8998

This research will be conducted under the supervision of Dr. Jacqueline Specht, Associate Professor, Director, Centre for Inclusive Education, Faculty of Education at the University of Western Ontario

Appendix B

Letter of information and consent for parents

The Educational Impacts of Tuberous Sclerosis Complex (TSC)
Letter of Information

I am a Masters student in the Department of Education at The University of Western Ontario and the information I am collecting will be used in my thesis.

Introduction
You are invited to participate in a research study looking at the educational impacts of Tuberous Sclerosis Complex. The purpose of this letter is to inform you about the details of the research study, and to give you all of the necessary information to make an informed decision to participate in this study.

Purpose of this study
The purpose of this study is to investigate how Tuberous Sclerosis Complex affects individual children, and how this in turn affects their schooling.

Research tests or procedures for this study
If you take part in this study, you will be asked to participate in an interview with the researcher about your child, your child's current manifestations of TSC, any difficulties your child is having, or has had in school, and what sorts of accommodations/modifications in school have helped your child. The interview can take place in your home, in a mutually convenient place, or over the telephone. The interview will take approximately one to two hours to complete.
You will be asked to sign a consent form to allow the researcher to interview your child's teacher(s) and Educational Assistant(s). The purpose of this interview of your child's teacher(s) and Educational Assistant(s) is to get their professional perspective about how TSC affects education, and what they think is helpful in effectively teaching your child, and what accommodations/modifications they have put in place to help your child.

Risks and discomforts to you if you participate in this study
There are no known risks for participation in this study. We will be discussing how your child was diagnosed and what difficulties and medical challenges they have faced as a result of having TSC, which you may find sad to talk about. While answering some of the questions you may feel sad or upset. If this happens please tell the interviewer and she will discuss these feelings with you.
The benefits to you if you take part in this study
There are no direct benefits to you for participating in this study. However, your participation may help us get new knowledge that may benefit the education of children with TSC.

Participant Initials_____

What will happen to the information that is collected?
Interviews will be audio-recorded and then a transcript of the interview will be typed. Any identifying information will be removed during the transcription process. The researcher will analyze all the interviews from all the participants in the study to try to get a better understanding of the educational impacts of TSC, and what can be done in the classroom to help these children.

What to do if you want to withdraw from this study
Participation in this study is voluntary. You may refuse to participate, refuse to answer any questions or withdraw from the study at any time without penalty. If a question makes you uncomfortable, you may decide not to answer it.
Specific things you should know about confidentiality
Your research records will be stored in the following manner: locked in a cabinet in a secure office; audio tapes of interviews will be listened to only by members of the research team and they will be destroyed after 2 years.
If the results of the study are published, your name will not be used and no information that discloses your identity will be released or published without
your specific consent to the disclosure.

How long will this study last and how many people will be enrolled
This study will last approximately 6 months, and there will be approximately 30 people participating.
If you have any questions about this study please contact ...
Jennifer Flinn at (613) 823-8998 or jflinn@rogers.com
Or my research supervisor, Dr. Jacqueline Specht at (519) 661-2111 extension 88876 or jspecht@uwo.ca

If you have questions about your rights as a research subject you may contact:
Director of the Office of Research Ethics
The University of Western Ontario
519-661-3036

If you would like to receive a copy of the overall results of this study please let the interviewer know.

Participant Initials_____

The Educational Impacts of Tuberous Sclerosis Complex (TSC)
Consent Form

I have read the Letter of Information/Consent document, have had the nature of the study explained to me and I agree to participate. All questions have been answered to my satisfaction.

Name (please print)

_____

Signature

_____

Date

_____

Appendix C

Information and Consent Letter for Educators
The Educational Impacts of Tuberous Sclerosis Complex (TSC)
Letter of Information

I am a Masters student in the Department of Education at The University of Western Ontario and the information I am collecting will be used in my thesis.

Introduction
You are invited to participate in a research study looking at the educational impacts of Tuberous Sclerosis Complex. You have a child in your classroom that has TSC, and that child's parents have given their consent for you to discuss this child with the researcher. The purpose of this letter is to inform you about the details of the research study, and to give you all of the necessary information to make an informed decision to participate in this study.

Purpose of this study
The purpose of this study is to investigate how Tuberous Sclerosis Complex affects individual children, and how this in turn affects their schooling.

Research tests or procedures for this study
If you take part in this study, you will be asked to participate in an interview with the researcher about the child you teach who has TSC. Questions will concern: any difficulties the child is having, or has had in school, and what sorts of accommodations/modifications in school have helped the child. The purpose of this interview is to get your professional perspective about how TSC affects education, what you think is helpful in effectively teaching this child, and what accommodations/modifications you have put in place to help this child. The interview can take place in your school, home, in a mutually convenient place, or over the telephone. The interview will take approximately one hour to complete.

Risks and discomforts to you if you participate in this study
There are no known risks for participation in this study.

The benefits to you if you take part in this study
There are no direct benefits to you for participating in this study. However, your participation may help us get new knowledge that may benefit the education of children with TSC.
What will happen to the information that is collected?
Interviews will be audio-recorded and then a transcript of the interview will be typed. Any identifying information will be removed during the transcription process. The researcher will analyze all the interviews from all the participants in the study to try to get a better understanding of the educational impacts of TSC, and what can be done in the classroom to help these children.
Participant Initials_____

What to do if you want to withdraw from this study
Participation in this study is voluntary. You may refuse to participate, refuse to answer any questions or withdraw from the study at any time without penalty. If a question makes you uncomfortable, you may decide not to answer it.

Specific things you should know about confidentiality
Your research records will be stored in the following manner: locked in a cabinet in a secure office; audio tapes of interviews will be listened to only by members of the research team and they will be destroyed after 2 years.
If the results of the study are published, your name will not be used and no information that discloses your identity will be released or published without
your specific consent to the disclosure.

How long will this study last and how many people will be enrolled
This study will last approximately 6 months, and there will be approximately 30 people participating.

If you have any questions about this study please contact ...
Jennifer Flinn at (613) 823-8998 or jflinn@rogers.com
Or my research supervisor, Dr. Jacqueline Specht at (519) 661-2111 extension 88876 or jspecht@uwo.ca

If you have questions about your rights as a research subject you may contact:
Director of the Office of Research Ethics
The University of Western Ontario
519-661-3036

If you would like to receive a copy of the overall results of this study please let the interviewer know.

Participant Initials_____

The Educational Impacts of Tuberous Sclerosis Complex (TSC)
Consent Form

I have read the Letter of Information/Consent document, have had the nature of the study explained to me and I agree to participate. All questions have been answered to my satisfaction.

Name (please print)

_____

Signature

_____

Date

_____

Appendix D

Interview questions for Parents

How was your child first diagnosed (what symptoms brought you to be diagnosed?

How old were they?

Has you child had epilepsy?

Which types?

Has your child had to take medications for epilepsy, if so, which ones?

Does your child have any skin/heart/kidney/brain (from MRI)/or other manifestations?

Does your child have any behavioural differences?

Has your child been diagnosed with an autism spectrum disorder?

What type of school does your child attend?

What types of school skills does your child experience difficulty with?

What types of services through the school (or privately) are involved in your child's education?

What types of accommodations have the teachers and Educational Assistants made that have been helpful for your child?

What types of accommodations have been less helpful?

Are there any accommodations or services that your child does not receive that you wish they did?

What is the most challenging aspect of school for your child? What is the most challenging aspect of sending your child to school?

Appendix E
Interview Questions for Educators

What type of program does the child participate in?

What do you know about Tuberous Sclerosis?

Where did you learn about it?

Where do you look when you need additional information about this child's disorder or needs?

How would you characterize the impact this disorder has on this child's education?

Which subject areas are the most challenging for the child?

What types of specific delays have you observed in the child?

What types of behaviour differences have you observed in the child?

What types of services through the school (or privately) are involved in the child's education?

Is the child pulled out of the classroom for resource or remedial help? With who?

What types of accommodations have you made that have been helpful for the child?

What types of accommodations have been less helpful?

How do you decide on accommodations? Is there a team that you consult with?

How do your accommodations for this child differ from accommodations you might make for another child with a disability?

Are there any accommodations or services that this child does not receive that you wish they did?

Are there supports lacking for this child?

What is the most challenging aspect of school for this child? What is the most challenging aspect of teaching and accommodating this child?

Appendix F

	Age and Method of diagnosis	Seizures	Age of Seizure Onset	Skin	SENS	Tubers	Kidneys	Heart	Hyper Active	Autism	Behaviour	Late to Speak	Early Intervention Services	Current Service	Parent reported biggest need	Parent reported accommodations	Advocacy Mentioned
1A	skin findings, age 5	CP, IS*	unsure, retrospectively, mom thinks as a baby	Angiofibroma, Shagreen Patches		X	X	X	X		tantrums, rages	18 months, stuttered, hard time getting language out	no	IEP, Resource Pull out	academic	IEP, resource teacher, one on one, visual, EA, repetition	No
2A	seizure two months	Many, unsure which types	2 months	Ash Leaf Spots		X	X			X	Tantrums, anger mngmnt	still doesn't communicate	OT, PT, Speech	EA	behaviour, motor difficulties	behaviour goals on IEP, does better with one on one instruction	No
3A	seizure at 19 months	CP	19 months	Ash Leaf Spots, angifibromas	X	X	X	X			Quirky, needs routines, anxiety over performance, organization	only a couple of words at age two	speech, parents report clumsiness but didn't see OT	none	memory difficulties, needs routines, multi step directions		no
4A	seizure at 8 months	IS	8 months	angiofibroma	x	x	x-cystic	x				no	no	none	organization	none	no
5A	seizure at 6.5 months	CP, IS, P, Status	6.5 months		X	X	X	X	X-on Concerto for ADHD		Fixates, OCD?	No, but regression with each status seizure	OT, PT, speech,	IEP, full time EA,	ADHD, memory issues, math (EA says Lang)	repetition, visual	Yes
6A	unusual heart murmur at birth	Abnormal EEG, 1 Febrile Seizure				X	X	X			Anxiety, rages, Tantrums, organization	no	speech, IEP for preschool	Behaviour services,	math, socialization, takes things too literally	preferential seating for eye problem,	
7A	identical twin sister born with unusual murmur	CP, SP, full body	1 month	Angiofibroma	X	X	X	X		X	Anxiety, violence towards family, repeating, echolalia	yes, age 5, sign used before	OT, PT, Speech, infant massage	Behav. And ASD services, OT/PT, Speech	social, speech,	Completely modified program	Yes
8A	Dermatologist, age 5			angifibroma	X	X		X	ADHD		repetitive movements, like pen tapping	no	no	IEP	ADHD	Computer access, quiet testing, extra test time	yes
9A	ultrasound found Rahbdomyoma, at birth	Drop?	2 years	Ash Leaf Spots, Angiofibromas	X	X	Cystic kidneys	X				Didn't mumble	OT, SP, integration service, home dev. therapist	Nothing now	fine and gross motor organization on paper	Mom sometimes modifies hmwrk	Yes
10A	geneticist ordered tests, based on heart murmur, ash leaf, seizures, brain tumours age 8	P, CP, Gilastic	2.5 years	Ash Leaf Spots	X	X	?	X	X	X	anxiety, OCD, Fixates, difficulty with social cues, oppositional, ADHD, tantrums	regressed after seizures	no	IEP, Living Skills, OT, PT, EA,	math, fine, gross motor, social, behaviour, organization	scribe, read for her, living skills class, functional math, sensory room, planning for anxiety provoking situations	yes

Appendix G

	Teacher	Educational Assistant	Area of Difficulty	psych ed or Speech Lang done	IEP	Has EA Support	OT	PT	Speech Therapy	Program Type	Autism	Behaviours	Anxiety	Accommodations	Friendships	Anxiety about teaching child with TSC	Information from
3B	X		New or Unfamiliar information or routines	psych ed done age 4, recall issues highlighted						Regular, referred for gifted		Quirky, does own thing	When she doesn't understand work	Repetition, modelling, breaking down steps	social has friends, no one best friend		parents
4B	X		Organization							Regular, gifted pull out		Quirky, some difficulty understanding social cues, takes language too literally?	About marks and school work	organization	social has friends, no one best friend	would have liked more info before he got student	No information provided from parents or otherwise
5B		X	Language, memory, retention, new and unfamiliar material	Speech language reports gaps in receptive and expressive	X	Full time			probably on waiting list for SP	Regular with EA support		fixates, repeats things, ADHD		repetition, review		anxiety before she got to know child	Parents and internet
7B	X		anxiety, blood pressure goes up when she has to go to special classroom, totally modified program		X	full time, and full time nurse	X	X	X	Regular classroom with full time EA, all expectations modified, pull out to Autism support special self contained classroom	X	Repeats,	Yes, especially to loud noises	repetition, visuals, sign language, totally modified prog, touch math, backwards chaining	enjoys interacting with students, who have been made aware of her condition, and embrace her	anxiety and doesn't feel supported or prepared, talks about importance of parent advocacy	parents
8B	X		ADHD, difficulty focusing, difficulty with fine motor (taking notes), tired quickly		X					Regular program, with accommodations for writing, focusing, re-teaching		ADHD, difficulty paying attention, tapping	About achievement	Note-taking, scaffolding, computer for note taking, quit testing, re-teaching to fill in gaps	many, but no one group or particular best friend	yes, worried she wouldn't be able to teach him or make him comfortable in class, mentioned the importance information	parent and internet
10B		X	functioning at grade 2 level (student in gr. 6)	Speech language showed scattered testing, high in expression, but low in receptive	X	Full time	X	X	X	Regular classroom, all expectations modified, living skills pull out, resource pull out	Say no, but mom says yes	fixates, socially inappropriate	Says no, but mom says yes	consultant came in to educate class-mates, jobs given in the school,	difficulty understanding social situations		parents

Appendix H

THE UNIVERSITY OF WESTERN ONTARIO
FACULTY OF EDUCATION

USE OF HUMAN SUBJECTS - ETHICS APPROVAL NOTICE

Review Number: 0805-6
Applicant: Jennifer Flinn
Supervisor: Jacqueline Specht
Title: *The educational impacts of tuberous sclerosis complex*
Expiry Date: December 31, 2008
Type: M.Ed. Thesis
Ethics Approval Date: June 18, 2008
Revision #:
Documents Reviewed & Approved: UWO Protocol, Letters of Information & Consent, Advertisement

This is to notify you that the Faculty of Education Sub-Research Ethics Board (REB), which operates under the authority of The University of Western Ontario Research Ethics Board for Non-Medical Research Involving Human Subjects, according to the Tri-Council Policy Statement and the applicable laws and regulations of Ontario has granted approval to the above named research study on the date noted above. The approval shall remain valid until the expiry date noted above assuming timely and acceptable responses to the REB's periodic requests for surveillance and monitoring information.

No deviations from, or changes to, the research project as described in this protocol may be initiated without prior written approval, except for minor administrative aspects. Investigators must promptly report to the Chair of the Faculty Sub-REB any adverse or unexpected experiences or events that are both serious and unexpected, and any new information which may adversely affect the safety of the subjects or the conduct of the study. In the event that any changes require a change in the information and consent documentation, newly revised documents must be submitted to the Sub-REB for approval.

Dr. Jason Brown (Chair)

2007-2008 Faculty of Education Sub-Research Ethics Board

Dr. Jason Brown	**Faculty (Chair 2008)**
Dr. Elizabeth Nowicki	**Faculty**
Dr. Jacqueline Specht	**Faculty**
Dr. Wayne Martino	**Faculty**
Dr. J. Marshall Mangan	**Faculty**
Dr. Immaculate Namukasa	**Faculty**
Dr. Robert Macmillan	**Assoc Dean, Graduate Programs & Research (*ex officio*)**
Dr. Jerry Paquette	**UWO Non-Medical Research Ethics Board (*ex officio*)**

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Karen Kueneman, Research Officer
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Copy: Office of Research Ethics