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School Factors Related to the Social and Behavioral Success of Children and Adolescents with Tuberous Sclerosis: Special Education Placement, Services, and Parental Involvement

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School Factors Related to the Social and Behavioral Success
of Children and Adolescents with Tuberous Sclerosis:
Special Education Placement, Services, and Parental Involvement.

by

Kathleen Walker Carlisle

A dissertation submitted in partial fulfillment
of the requirements for the degree of
Doctor of Philosophy
Department of Psychological and Social Foundations
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For Cheryl, James, and Richard.

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ABSTRACT

The researcher examined the relationships between tuberous sclerosis, a multi-system genetic disorder, and school functioning through the use of a parent questionnaire and behavior rating scale. Information was gathered on the typical school experiences of children with tuberous sclerosis, including educational placement and services, behavioral functioning, parent involvement, and parent satisfaction. The results indicated that the majority of students with tuberous sclerosis are in special education and receiving related services. Three-quarters received one or more related services through the public school, and 30% received private related services paid for by their parents. Parent involvement was positively correlated with parent satisfaction, and negatively correlated with t-scores on the Withdrawn/Depressed subscale of the CBCL. Parents of children receiving Autism services were generally less satisfied with their children's school experiences than other parents. Parent satisfaction was negatively correlated with the Attention Problems scale of the CBCL. Student age was negatively correlated with time in inclusion and with related services. This information forms the basis for a discussion of school psychologists' roles in the educational success of students with tuberous sclerosis and the critical areas towards which interventions should be directed.

Chapter I

Introduction

School psychologists deal with an increasingly diverse population of students (Graden, Zins, Curtis, & Cobb, 1989). Among these students are those with medical and genetic disorders often associated with psychoeducational difficulties (Lail & Schroeder, 1990). With the passage of Public Law 94-142 in 1975, and subsequent reauthorization of IDEA in 1997, schools are now required to provide a free and appropriate education to all children. Children who previously were excluded because of mental retardation, behavior problems, or severe communication disorders must now be served in the least restrictive environment. In addition, better medical care has allowed children with chronic illnesses, medical complications, and physical handicaps to live longer, more normal lives (Lail & Schroeder, 1990). Together, these factors mean that school psychologists now work with a greater number of children with a broader range of needs than ever before.

Given the sheer number of psychiatric, medical, and genetic conditions affecting children, it is not surprising that many school psychologists are not prepared to address the range of psychoeducational needs of these children. However, as professionals who work with teachers, parents, students, and administrators, school psychologists are in a key position to coordinate and inform services for children with these conditions.

One genetic disorder often associated with a wide range of psychoeducational issues is tuberous sclerosis. Tuberous sclerosis (TSC) is a multi-system genetic disorder that causes benign tumors to form in many different organs, such as the brain, eyes, heart, kidney, skin, and lungs (Gomez, 1998a; Whittemore & Roach, 1999). The predominant manifestations of tuberous sclerosis are seizures, cognitive impairment, and skin marks. However, behavioral, communication, and psychiatric disorders also are common. The type and severity of symptoms associated with tuberous sclerosis vary widely, depending on the location, number, and type of tumors in the body (Hunt & Shepherd, 1993).

The prevalence of tuberous sclerosis in the general population is difficult to determine, as many individuals with milder symptoms remain undiagnosed until later in life (Hunt & Shepherd, 1993). General estimates, however, suggest that tuberous sclerosis occurs in 1 in 5,800 births (Osborne, Fryer, & Webb, 1991). No gender or ethnic variations have been found (Hynd & Willis, 1988; Smalley et al., 1994).

Tuberous sclerosis, or TSC, for tuberous sclerosis complex, is the leading genetic cause of seizure disorders (Smalley, et al, 1992). Less than one percent of the general population is estimated to have a seizure disorder, affecting approximately 500,000 children and adolescents in the United States (Sachs & Barrett, 1995). However, seizure disorders are estimated to occur in 85-95% of individuals with TSC (Curatolo et al., 1991). There appear to be strong links between the presence of seizures in TSC and intellectual disability (Shepherd & Stephenson, 1992; Baltaxe, 1998). Nearly all individuals with TSC and intellectual impairment also exhibit seizure disorders (Shepherd & Stephenson, 1992).

Tuberous sclerosis is associated with many physical characteristics, including skin marks. The most common skin marks associated with TSC, called facial angiofibromas, appear as small reddish spots or bumps across the cheeks and nose (Riccio & Harrison, 1998; Whittemore & Roach, 1999). TSC can affect virtually any part of the body. Commonly affected organs include the brain, kidneys, heart, and eyes (Webb, Fryer, & Osborne, 1996; Roach, 1992). When tumors form in these organs they adversely affect organ functioning, causing any number of medical complications.

Tuberous sclerosis also is associated with a variety of behavioral, communication, and psychiatric disorders. Common behavioral problems include hyperactive or impulsive behavior and aggressive or destructive behavior (Smalley, Tanguay, Smith, & Gutierrez, 1992). In addition, most children with TSC and normal intellectual functioning have speech, language, and communication deficits in one or more areas, including expressive vocabulary skills, auditory language processing, abstract language skills, and expressive semantic-grammatical skills (Baltaxe, 1998). Psychiatric disorders such as autism, anxiety disorders, and attention-deficits, may be found in up to 75% of individuals with TSC (Smalley et al., 1994). The most common psychiatric disorder co-occurring with TSC is autism. Approximately 50% of children with tuberous sclerosis demonstrate autistic behaviors by age 5 (Hunt & Dennis, 1987). Up to 14% of children already diagnosed with autism may also have tuberous sclerosis, with higher frequencies for children who also have a seizure disorder (Hunt & Shepherd, 1993).

Tuberous sclerosis is clearly associated with a number of health, learning, and behavior issues at many different levels of severity. The extent to which these problems impact school functioning is mediated in part by the response of the school environment

to the individual with TSC. Little research is available, however, about how schools typically respond to students with TSC, including special education placements and support services provided.

Best practices suggest that schools must respond with integrated health and educational services, and interventions which are tailored to the specific learning or social difficulties experienced by the child protected by IDEA or 504. (Allen & Graden, 1995). Interventions may focus on minimizing the social/emotional impact of seizures and skin marks, designing specific accommodations for learning and behavior problems, providing emotional support for families and siblings, and monitoring general progress (Grant, 1989; Thompson, 1995; Teeter & Samrud-Clikeman, 1998; Riccio & Harrison, 1998). Clearly, these interventions could involve school personnel from a wide sphere of disciplines. However, as professionals with expertise in learning, behavior, and consultation, school psychologists are in a prime position to take the lead in tailoring services to students with TSC. But in order for psychologists to make informed decisions supporting the educational success of students with TSC, they must first know the factors affecting the school experiences of children with TSC and the ways students resemble or differ from students with similar disabilities. It is suspected that the school experiences of children with TSC are dependent on a number of factors, including the child's cognitive functioning, the type and severity of specific learning, communication, and behavior problems, educational placement, and parent involvement.

The first area that may affect the school experiences of students with TSC is the level of cognitive functioning. Approximately 50% of individuals diagnosed with tuberous sclerosis have some form of cognitive impairment (Gomez, 1988; Roach, 1992).

These numbers are even higher for individuals with TSC who also have seizure disorders (Shepherd & Stephenson, 1992). This would suggest that a majority of students with TSC receive special education services. Very little research has been done, however, on the educational placements of children with TSC. One study of communication disorders in children with TSC found that 68% of the participants were in special education (Baltaxe, 1998). Of these, more than a quarter were in resource 'pull-out' programs and another quarter were receiving self-contained services for students with learning disabilities. Most of the remaining students were attending "special day classes", which were not further described. It is well known that students who do not fit into the mainstream of educational functioning have a more difficult time in school, and are more likely to drop out of school (Chesapeake Institute, 1994; Valdes et al., 1990). In addition, students with academic difficulties who do not qualify for special education services may receive little academic support from the school (Jackson, 1990).

The second area that may affect the school experiences of students with TSC is disruptive, non-compliant, and aggressive behavior. In the general school-aged population the prevalence of conduct disorders is estimated to be between 2% and 6%, with higher occurrences of attention-deficit behaviors, especially among African-American students (Kazdin, 1993; Nolan, Gadow, & Sprafkin, 2001). Children with TSC are much more likely to experience behavior problems, such as hyperactive or impulsive behavior (26%), or aggressive or destructive behavior (48%) (Smalley, Tanguay, Smith, & Gutierrez, 1992). Educational research has suggested that students with emotional or behavioral disorders are more likely to perform below grade level, have a higher rate of absenteeism, and a higher drop-out rate --48%, compared with 30% of all students with

disabilities, and 24% of all high school students (Chesapeake Institute, 1994; Valdes, et al., 1990). They also have greater difficulty making and keeping friends (Cartledge & Milburn, 1995). As a result, the educational or social-emotional needs of children with TSC and concomitant behavior problems are largely unmet. However, as stated previously, the extent to which these problems impact school functioning is mediated by the response of the school environment, including accommodations made and services provided.

The third and perhaps most important area which may affect the school experiences of students with TSC is parent involvement and advocacy. Parental involvement has been shown to benefit children's learning and school success (Eccles & Harold, 1993; U.S. Department of Education, 1994; Soodak, 1998). In addition, the more schools interact with parents, the more parents have positive attitudes towards communication with the schools (Arnold, Michael, Hosley, & Miller, 1994). Interactions that increase the parents' level of satisfaction include phone calls from the teacher, personal invitations and encouragement to attend school programs, and notes home (Arnold, Michael, Hosley, & Miller, 1994).

Although a great deal of research has been conducted on the education, behavioral, and family needs of students with significant disabilities, little research has been conducted to determine the relevancy of this research to students with TSC. More specifically, school-based research is lacking in areas that relate to educational placement, intervention programs that deal with the problems often associated with TSC, and parent involvement and perceptions of the school's response to their child with TSC.

This information is especially important for psychologists, as they are in a unique position to coordinate and inform services to students with TSC.

The purpose of this dissertation was to examine the relationship between tuberous sclerosis and school functioning by gathering information on the typical school experiences of children with TSC, including educational placement, academic, communication, and behavioral functioning, support services, and educational accommodations. This dissertation also examined the relationships between student functioning, educational placement, and levels of parental involvement and satisfaction with the school experience. The information gathered forms the basis for a discussion of school psychologists' roles in the educational success of students with TSC and the critical areas to which interventions should be directed.

Research Questions

1. What percentage of children with TSC is in special education, and in what setting (type of school) are they educated?
2. Under what special education labels or categories are children with TSC served?
3. What percentage of school time is spent in the special education setting?
4. What percentage of students receives additional or related services, and what are those services?
5. What percentage of children has cognitive functioning in the Above Average, Average, Low Average, or Low ranges?
6. What percentage of parents communicates with their child's teacher or school: Never, 1-3 times each year, 4 or more times each year.

7. What percentage of parents are Very Satisfied, Satisfied, Unsatisfied, or Very Unsatisfied with the school's response to their child's needs?
8. What is the relationship between parental involvement and:
 - a. special education label/category.
 - b. behavior problems.
 - c. type of school.
 - d. time spent in special education.
 - e. related services.
 - f. cognitive functioning.
9. What is the relationship between parental satisfaction and:
 - a. special education label/category.
 - b. behavior problems.
 - c. related services.
 - d. type of school.
 - e. time spent in special education.
 - f. cognitive functioning.
10. What is the relationship between special education labels or categories and:
 - a. behavior problems.
 - b. related services.
 - c. time spent in special education.
 - d. cognitive functioning.
11. What is the relationship between behavior problems and:
 - a. related services.

- b. type of school.
- c. time spent in special education.
- d. cognitive functioning.

Chapter II

Literature Review

This chapter will provide a basic overview of the history, prevalence, and major characteristics of tuberous sclerosis complex (TSC). This general information is included because TSC is a relatively rare disorder and it is strongly suspected that school psychologists are unfamiliar with its various manifestations. It will also serve to highlight the differences between TSC and other related disorders producing similar symptoms. Studies of TSC in children of interest to school psychologists fall into two categories. First, there are a number of population studies that seek to describe the prevalence and major characteristics of TSC. Second, there are studies examining particular problems related to TSC, including seizure disorders, delayed or impaired cognitive functioning, behavior and psychiatric problems, sleep disorders, and communication disorders. It is important to note that no studies were found dealing specifically with the educational needs and concerns of children with TSC and their parents. However, all of the studies reviewed contain information that may be used to inform the care and services provided for children with TSC in schools.

History

The tuberous sclerosis syndrome was first described in the late 1800's by Bourneville, who named the disease "tuberous sclerosis of the cerebral circumvolutions" (1880). His patient was a 15 year-old epileptic and mentally handicapped girl, whose

face bore the distinctive red bumps which were improperly called "adenoma sebaceum", but are now known as "facial angiofibromas". These three characteristics--seizures, mental retardation, and facial angiofibromas--came to be known as the central features of a diagnosis of TSC. However, it is now suspected that less than a third of all patients with TSC can be recognized by this triad (Whittemore & Roach, 1999).

Currently, TSC is known as "one of three neurocutaneous (involving the nerves and skin) syndromes that account for a significant number of referrals to pediatric neurology clinics" (Hynd & Willis, 1988). It is characterized visually by birthmarks of the skin known as facial angiofibromas that increase in number and size with age. These flesh-colored marks and bumps often resemble acne and are typically displayed across the nose and cheeks in a butterfly distribution (Riccio & Harrison, 1998). The name tuberous sclerosis is derived from the lesions found in the cerebral cortex of the brain. These lesions result in areas of hardened tissue, or tumors, in various locations throughout the body (Roach, 1992). The wide variation of expression of the disorder is dependent on the location, number, and type of tumors in the body (Hunt & Shepherd, 1993). Therefore, there is no typical profile of the child with tuberous sclerosis (Miller & Bigler, 1982). Seizures and intellectual impairment are considered core characteristics of more severe cases, however; and other common characteristics include behavior and psychiatric problems, sleep disorders, and communication disorders.

Prevalence

Today, estimates place the prevalence of TSC at approximately 1 in 6,000 births (Osborne, Fryer, & Webb, 1991; Whittemore & Roach, 1999). This means that at least one child born each day in the U.S. will be affected with TSC. However, the prevalence

of TSC among the general population is difficult to determine because of the number of undiagnosed cases at any point in time. Individuals with milder forms of TSC often are not diagnosed until their teens, with the average age of diagnosis ranging from 11-15 years old among children and adolescents (Webb, Osborne, & Fryer, 1991). In addition, a significant number of adults of average intelligence are not diagnosed until medical complications occur, or until a more severely affected family member is identified (Hunt & Shepherd, 1993; Medcalf, 1991).

It was once thought that TSC had a nearly 80% mutation rate, indicating that it was not inherited from either parent (Hunt & Shepherd, 1993). However, as more individuals with milder symptoms are diagnosed, including family members of individuals with TSC, the estimated mutation rate has decreased. Currently, one-third of TSC cases are known to be genetically transmitted. The other two-thirds are believed to be a result of spontaneous mutation. Historically, TSC was thought to be exceedingly rare, with population estimates as sparse as 1 in 150,000 (Stevenson & Fischer, 1956). But as methods for detecting tuberous sclerosis have grown in sophistication more cases have been identified. As recently as a decade ago, researchers estimated that TSC occurred in 1 in 12,000 children (Sampson, Scahill, Stephenson, Mann, & Connor, 1989). Now that number is thought to be considerably greater. (Whittemore & Roach, 1999).

Comorbidity of Seizures and Intellectual Impairment

Two of the primary characteristics of TSC are seizures and cognitive impairment. Research has shown the two are so closely tied that they will be dealt with together in this section. It is estimated that 50% of the individuals with TSC have mild to profound cognitive impairment (Hunt, 1995; Hunt & Dennis, 1987; Shepherd et al., 1991; Riccio

and Harrison, 1998; Gomez, 1988a; Roach, 1992). Of those, approximately 35% would be identified as mentally retarded, with the other 15% experiencing milder forms of cognitive impairment (Curatolo, et al., 1991). Across all age levels, however, approximately half of the individuals affected with tuberous sclerosis have normal cognitive abilities (Medcalf, 1991; Sampson et al., 1989).

Seizures occur in approximately 80% of individuals with TSC (Webb, Fryer, & Osborne, 1996). The co-occurrence of seizures and cognitive impairment in individuals with TSC is significant. Moreover, there appears to be a relationship between the type and severity of seizures and the presence of mental retardation. Several studies have sought to elucidate the relationship between seizures and intellectual disability. Gomez (1979) found that 88% of patients with TSC had seizures and only half of the patients had an intellectual disability. However, all patients with intellectual disabilities also had seizures. Another study found 85% of individuals with TSC had seizures, and of these, 68% presented with a type of seizure known as infantile spasms (Hunt, 1983). The relationship between infantile spasms and cognitive impairment is a critical one, but was not explored in-depth until years later.

One of the first in-depth studies of the relationship between seizures and intellectual disability was undertaken by Shepherd and Stephenson (1992). This study sought to examine the relationship between different types of seizures, age of onset, and level of intellectual disability in all cases of TSC in a defined population. Shepherd and Stephenson (1992) used the population of all ascertained cases in the west of Scotland as defined by Sampson et al. (1989) in a previous study. In addition, further cases were identified through medical examination and interview. The participants were 104

individuals with TSC. The researchers examined the hospital record of each participant and gathered information about their seizures. Seizure types were then classified using the 1981 classification system of epileptic seizures (Wolf, 1985). Intellectual impairment was classified as moderate (IQ 35-70) or severe (IQ <35) (Shepherd & Hosking, 1989). Individuals in regular education classes were considered to have normal intelligence and did not undergo intellectual assessments. The sample was divided into two age groups, based on the availability of medical records for the older group. For the older group, the only consistently reliable facts were the presence or absence of seizures and the presence or absence of intellectual disability. A third of those with seizures had normal intelligence, but all individuals with intellectual disability also had seizures.

The second group was comprised of younger individuals with TSC for whom more detailed medical records were available. These detailed medical records allowed for an examination of the relationship between seizure type and degree of intellectual disability. Of those whose presenting seizures were infantile spasms (a type of seizure occurring at less than 2 years of age), 83% had a severe intellectual disability (IQ <35). The age of onset of seizures was also predictive of intellectual impairment. Of individuals whose onset of seizures was before age 1, 64% had severe intellectual disabilities. Of individuals whose onset of seizures was between ages 1 and 5, only 25% had severe intellectual disabilities. None of the individuals whose seizures began after age 5 had any type of intellectual disabilities.

The number of different seizure types experienced by the individual was also predictive of cognitive impairment. Of individuals with only one type of seizure, 9% had a moderate intellectual disability (IQ 35-70), and 35% had a severe intellectual disability

(IQ <35). Of individuals with more than one type of seizure, 16% had moderate disabilities and 64% had severe disabilities.

These results support the strong relationship between seizures and intellectual disability in individuals with TSC. The best intellectual outcomes are associated with the late onset of seizures and only one type of seizure, or the complete absence of seizures. In most of the individuals with any type of intellectual impairment, the disability was severe, as has been found in other studies (Hunt & Lindenbaum, 1984; Webb, Fryer, & Osborne, 1996; Yamamoto et al., 1987). This, however, may reflect sampling bias, as those with moderate disability may be less likely to seek medical services.

Building upon the work of Shepherd and Stephenson, Hunt (1993) examined the relationships between seizures and varying degrees of learning difficulties in children with TSC. Hunt used a postal questionnaire to gather information on 300 families in which a member was diagnosed with tuberous sclerosis. The questions concerned seizures, development, and behavior of the person with TSC, and services received by the family. Behaviors reported were analyzed for psychiatric disorder, such as autism, and for behaviors that disrupted family life, such as sleep disturbance, unpredictable behavior, and inappropriate social behavior in public. Most of the questionnaires were completed by parents on behalf of their affected children.

Developmental delay or learning difficulties were reported by 80% of respondents. Of those with learning difficulties, all but one (99%) had a history of seizures. Those with learning difficulties also showed higher incidences of other disturbances. Sixty-six percent of those with learning difficulties also had sleep problems in general. Respondents indicated that 35% were overactive, 74% displayed

“unpredictable actions,” 55% displayed “uninhibited public behavior,” and 55% reported non-compliant behavior.

In addition, those with learning difficulties also had high incidences of obsessive or ritualistic behavior. Repetitive and odd play was reported by 44% of respondents, and unusual routines and gestures were reported by 50% of respondents. Between 31% and 46% of children with learning difficulties were obsessive about routines or objects.

The majority of aggressive behaviors were by people with learning difficulties. Ninety-six percent of temper tantrums, 95% of attacks on others, and 97% of self-injurious behavior occurred in this group.

Despite the challenges that families of persons with TSC and learning difficulties may face, less than half (45%) received respite care or professional assistance. Most of the specialized services received were by pre-school counselors, speech therapists, physical therapists, and psychologists. It seems most likely that these services were offered and received through public education.

Several years later, Webb, Fryer, and Osborne (1996) examined the intellectual functioning of both children and adults with TSC in the total population of southwest England. The researchers classified intellectual functioning into three grades: severe, for those without language and requiring complete care; moderate, for those able to hold reasonable conversation but requiring close supervision for daily living; and mild, for those with difficulties in reading and writing but largely living independently. No formal intellectual assessment procedures, such as IQ tests, were used. Seizures occurred in 78% of the participants. Learning difficulties occurred in half the total sample, all of whom had a history of seizures. Learning difficulties were also highly correlated with

early onset of seizures, infantile spasms, and poor seizure control. Of all subjects with varying learning disabilities, 85% required supervision for daily living. Sixty-five percent had little or no language. All of those with learning difficulties had a history of seizures.

A more recent study by Jozwiak, Goodman, and Lamm (1998), again examined the factors associated with decreased intellectual functioning in individuals with TSC, but focused exclusively on children. The researchers sought to identify clinical risk factors for poor mental development among individuals with TSC. The participants were 106 patients with TSC seen at the Child Neurology Clinic of the Children's Memorial Health Institute in Warsaw, Poland between January 1984 and December 1995. The variables examined were seizure type, age of seizure onset, sex, and history of diphtheria, tetanus, and pertussis (DPT) Immunization. Intellectual functioning was measured using the Psyche Cattell Intelligence Test for Small Children (for children under 30 months), the Termann-Merill Mental Development Test (for children 30 months to 5 years), and the Wechsler Intelligence Scale for Children (for children older than 5). Intellectual functioning was classified into Fair or Poor mental status, based upon the International Statistical Classification of Disease and Related Problems, Ninth Revision. The upper three strata--normal, low average, and mild delay (IQ of at least 70)--were classified as Fair mental status. The lower three strata--moderate, significant, and profound delay (IQ of 69 or less)--were classified as Poor mental status. The diagnosis of TSC was established on the basis of the criteria of Gomez (1991) and the 1992 criteria of the National Tuberos Sclerosis Association (now known as the TSAAlliance) (Roach, Smith, & Huttenlocher, 1992).

The results indicated that only one variable, seizure type, showed a consistent and independent relationship to Poor mental development. Age of onset was not significantly related after adjustment for seizure type. Infantile spasms, the authors concluded, are a significant risk factor for mental retardation in individuals with TSC.

Even in the non-TSC population, seizures disorders are associated with reduced intellectual functioning, though not to the extent that they are in individuals with TSC (Sachs & Barrett, 1995). Again, several factors appear to influence the degree of cognitive impairment associated with seizures, including age of onset, duration of disorder, type of seizure, frequency of seizures, and anti-epileptic drugs used (Dodrill, 1992). Seidenberg, O'Leary, Berent, and Boll (1981) studied the relationship between age of onset of seizures and IQ scores. The researchers conducted a study of 410 adults with epilepsy, using the Wechsler Adult Intelligence Scale. For individuals whose age of onset of seizures was before age 5, the average Full Scale IQ was 90.66. For individuals whose onset of seizures was 6 to 12 years old, the average Full Scale IQ score was 94.89. And for individuals whose onset of seizures was between 13 and 18 years, the average Full Scale IQ score was 96.74, differences that, though small, were statistically significant.

Duration of the seizure disorder was also correlated with degree of cognitive impairment, although the combination of longer duration and increased seizure frequency was a better predictor of cognitive impairment. The type of seizure experienced was also predictive of cognitive impairment. Generalized convulsive seizures, versus absence seizures, were associated with higher rates of intellectual impairment.

Test-retest IQ studies suggest that anti-epileptic drugs may also have a role in cognitive impairment. Certain anti-epileptic drugs, such as phenobarbital and Zaronin, seem to have a negative impact on IQ scores over time (Dodrill, 1992; Sachs & Barret, 1995). Other researchers have demonstrated improved IQ scores with anti-epileptic drugs, but this appears to be more closely related to decreases in seizure rates than to a direct drug effect (Seidenbert, et al., 1981). The use of multiple anti-epileptic drugs is more likely to have adverse effects on general cognitive functioning (Bourgeois et al., 1983). However, these findings are confounded by the fact that "individuals requiring multiple medications also are more likely to have underlying observable [Central Nervous System] abnormalities, a known risk for cognitive deficits" (Sachs & Barrett, 1995, p.140).

The research on seizures in individuals without TSC suggests that, although seizures themselves may be associated with mental retardation, the presence of TSC makes those rates substantially higher.

In summary, the relationship between TSC, intellectual impairment, and seizures is a complex one. Several assertions, however, may be made with reasonable certainty. First, individuals with TSC have significantly higher rates of seizures and intellectual impairment than the general population. Second, individuals with TSC and seizures have higher rates of intellectual impairment than the general population of those with seizure disorders. Third, infantile spasms, in particular, are related to decreased cognitive functioning. Finally, individuals with TSC who have normal cognitive functioning are also more likely to experience seizures than the general population, although the age of onset of seizures is typically later than for those with severe impairment.

Behavior and Psychiatric Problems

Although not considered a core characteristic, behavior and psychiatric disorders are another common characteristic of TSC in childhood. Physicians are often concerned with the control of seizures, but "for many families it is the behavior problems that cause major disruptions" (Hunt, 1999). In a review of 50 published case reports, Smalley, et al., (1992) found that most behavior/psychiatric disorders can be divided into three major categories: autism and autistic-like characteristics, hyperactive or impulsive behavior, and aggressive or destructive behavior. Approximately 36% of children with TSC display autism or autistic-like characteristics, 26% display hyperactivity or impulsive behaviors, and 48% display aggression or destructive behavior. The authors did not specify whether these behaviors covaried. Because the literature base on the link between TSC and autism is so expansive, it will be discussed more specifically in a separate section to follow.

Psychiatric and behavioral problems are not uncommon in persons with TSC. Smalley, et al., (1994) sought to identify physical and behavioral variation of TSC in a single extended family. Information was gathered using a semi-structured interview widely used in genetic epidemiological research (the SADS-LA and K-SADS-E) and the Autism Diagnostic Interview. The researchers found a significant clustering of psychiatric disorders among persons affected with TSC as compared to their unaffected relatives. Notably, 59% of individuals with TSC had an anxiety disorder, compared to 13% of their unaffected relatives ($p=.016$). Individuals with TSC also had greater percentages of mood disorders, Attention Deficit Hyperactivity Disorder (ADHD), eating disorders, and substance abuse. Taken together, 76.5% of individuals with TSC had at

least one of these disorders, compared with 25% for unaffected family members (p=.009).

Other studies have shown a clear association between intellectual impairment and the presence of psychiatric disorders. In a population-based study of 28 affected children, only 3 children were found to be free of psychiatric problems, behavioral problems, or both--all 3 were of average intelligence (Gillberg, Gillberg, & Ahlsen, 1994). All the studied individuals with below-average intelligence exhibited a psychiatric disorder. The most frequent disorder was autism (61%), although ADHD was comorbid in 39% of children with autism and in 21% of children with autism-like characteristics. Among children with average intelligence (IQ greater than or equal to 85), only 29% exhibited psychiatric or behavioral problems, including Asperger's Syndrome. There were also 4 "near average" children in the study, with IQ scores between 71 and 84, who all exhibited some type of psychiatric or behavioral problem.

In another study that focused primarily on communication disorders, Baltaxe (1998) also reported the incidence of psychiatric diagnoses among participants. Only 21% of subjects had specific diagnoses, including autism, ADHD, anxiety disorders, depression, and psychosis. However, Baltaxe noted that psychiatric symptomology was present in a much larger percentage of subjects, based upon the patient's behavior, and on the developmental, medical, and family information gathered. This symptomology included (in order of most frequent to least frequent) anxiety and fearfulness, distractibility, ritualistic or self-stimulatory behaviors, hyperactivity, anger and rage, opposition to authority figures, extreme mood swings, impulsivity, obsessive-compulsive

behaviors, and phobias. Specific percentages of each were not included by the author, and raw data was not provided.

Infantile spasms, common in persons with TSC, have themselves been associated with higher incidences of behavior/psychiatric problems (Riikonen & Amnell, 1981). The question remained whether the psychiatric disorders often seen in children with TSC were associated only with infantile spasms, or whether they were directly related to the presence of TSC. Hunt and Dennis (1987) sought to answer this question using a 321-item parent interview to gain information on the adaptive functioning, play, and social interactions of children with TSC. The researchers also reviewed participant's medical records to gather as much information as possible on a wide variety of outcomes and antecedents.

The participants were 89 children and 1 young adult all with a confirmed diagnosis of TSC and whose families were members of the Tuberous Sclerosis Association (TSA) of Great Britain. The ages ranged from 5 months to 18 years. Of the 69 children who had experienced infantile spasms, 70% (40) were diagnosed as having "psychoses" by the researchers (autism being the most common form), 59% (41) were hyperactive, and 46% (32) were both hyperactive and "psychotic" (autism being the form of psychoses most common). Of the 17 children with seizures other than infantile spasms, 73% (11) were hyperactive, only 40% (6) had psychoses, and 33% (5) were both hyperactive and psychotic. No behavior or psychiatric problems were reported for the 4 children without seizure disorders. Overall, 50% of the children in this study showed autistic behavior by 5 years of age. The authors point out that the incidence of autistic behavior among children with TSC is much higher than would be expected simply as a

sequel to infantile spasms or mental retardation. The authors point out that 80% of mentally retarded children with TSC display autistic behavior, compared to 13% of mentally retarded children without TSC, stating, "Although these children are mentally retarded, their behavior is not that of the majority of retarded children" (p.196).

This study suggests that, although the presence of seizures may be related, the severe behavior disorders experienced by children with TSC is most likely a result of TSC, rather than the result of seizures alone. Hunt & Dennis (1987) also noted that "the behavior of the majority of children in this study seriously disrupted family life and their parents needed continuing advice on management problems" (p. 196). This was also one of the first studies to emphasize the link between TSC and autism. The researchers point out "both autistic and hyperkinetic children are recognized to need very specialized educational techniques which differ from those for other children with severe learning difficulties (Rutter, 1985; Taylor, 1986)" (p. 196)

In summary, children with TSC experience a greater number of behavior and psychiatric problems than the general population. The three main problems are autistic or autistic-like behaviors, hyperactive or impulsive behavior, and aggressive or destructive behavior. Some of this appears to be related to the presence of seizures. However, when compared to non-TSC individual with seizures, the rates of behavioral problems are still higher than expected. Thus, the presence of these disorders cannot be explained solely by the presence or absence of seizures or intellectual impairment, but appear to be related to the presence of TSC.

TSC and Autism

The first description of autistic behavior in TSC was provided by Critchley and Earl (1932), who studied 29 patients with TSC in mental hospitals. The behavior was categorized by the researchers as a primitive form of catatonic schizophrenia, but the behaviors described, such as repetitive movements, obsessive thoughts, and social isolation, fell soundly within the Autism spectrum.

Riccio and Harrison (1998) cite autism as one of the major psychoeducational problems of children with TSC. They noted that 50% of children with TSC demonstrated autistic behavior by age 5. Other researchers estimate the prevalence of autism in TSC to be 1 in 4 for children with TSC in general, and 1 in 2 for those with TSC and mental retardation (Hunt & Shepherd, 1993). Conversely, the frequency of TSC found in studies of autistic children ranges from 1%-14% with higher frequencies (8-14%) for those who also have a seizure disorder (Hunt & Shepherd, 1993).

Autism-spectrum disorders, such as Pervasive Developmental Disorder (PDD) and Asperger's Syndrome also may be represented in this group, as well as individuals with some autistic characteristics. In one instance, parents of children with TSC who were asked to complete a questionnaire on learning difficulties also noted repetitive and odd play (44% of respondents), unusual routines and gestures (50% of respondents), and obsessive behavior concerning routines and objects (31%-46% of respondents) (Hunt, 1993).

Hunt and Dennis (1987) were among the first to note the relationship between TSC and autism. The researchers used a 321-item family interview which covered family information, clinical effects of TSC, adaptive behavior, play and social interactions, the

impact of professionals' advice and support, and some open-ended questions about family life. The purpose of this interview was to gain information about psychiatric disorders among children with TSC, and their relationship to a wide variety of outcomes and antecedents. The researchers hypothesized that if psychiatric disorders were produced only by infantile spasms, an incidence similar to that found by Riikonen and Amnell (1981) would be expected. If the incidence of psychiatric disorder were more specific to TSC in general, a higher incidence would be expected. The reader will recall that Riikonen and Amnell (1981) studied the relationship between infantile spasms and psychiatric disorders in children with TSC, finding that 70% of children with infantile spasms had psychiatric problems, compared to only 40% of children with other types of seizures, and the absence of psychiatric problems among children without any seizures.

Hunt and Dennis (1987) found a strong correlation between infantile spasms and psychiatric problems and hyperactivity. Autism was the most common psychiatric problem noted, with 50% of all children in the study displaying autistic behavior by the age of five.

Since the work of Hunt and Dennis, a number of researchers have examined the relationship between autism and TSC, using different diagnostic criteria for autism. Riikonen and Simell (1990) reported the frequency of autism in 24 patients with TSC who had also had infantile spasms. They found that 17% were autistic, 21% were hyperactive, and 4% had poor social contact or extreme shyness. Curatolo et al. (1991) found that, of 34 participants with TSC, 26% met the criteria for infantile autism, as defined as a score of 10 or greater on the 13-item questionnaire used by Hunt and Dennis (1987). Other researchers place the estimated prevalence of autism in children with TSC

between 17 - 58%--a considerably wide range (Smalley, Tanguay, Smith, & Gutierrez, 1992).

Conversely, some researchers sought to ascertain the frequency of TSC among the autistic population. Ritvo et al. (1990) found only 1 case of TSC in a sample of 233 participants with autism (0.4%). Gillberg, Steffenburg, and Schaumann (1991) found 1 participant (1.89%) with TSC of 55 individuals with autism. In their study of 24 autistic individuals with infantile spasms, Riikonen and Amnell (1981) found 2 had TSC for a frequency of 8%. Gillberg (1991) found 14% of 66 individuals with autism and seizures to have TSC. Thus, the frequency of TSC among the autistic population was found to range from 0.4 - 3%. However, in a subgroup of autistic persons with seizures, the prevalence of TSC was much higher--between 8 - 14%.

One of the first studies of TSC and autism to use thorough diagnostic criteria for autism was undertaken by Smalley, Tanguay, Smith, and Gutierrez (1992). The study included 14 autistic participants and 13 TSC participants. Autism was diagnosed using the Autism Diagnosis Interview (ADI; LeCouteur et al., 1989). The researchers found that autistic children--with and without TSC-- look very similar in their socialization and communication. However, autistic children with TSC showed fewer repetitive rituals, suggesting that a diagnosis of autism in children with TSC should consider the core deficits (social and language deficits) more than motoric behaviors and stereotypies. Thus, autism may have a slightly different expression in children with TSC than it does in other individuals.

In 1993, Hunt and Shepherd published another prevalence study of autism in tuberous sclerosis. This study sought to determine whether the prevalence of autism in

TSC could be explained by a history of mental retardation or seizures. The parents of 21 children previously identified with TSC were interviewed about the behavior of their children. Through these interviews, five of the children (24%) were diagnosed as autistic using the DSM-III-R criteria. Another four (19%)--all girls--had socially impaired behavior categorized as Pervasive Developmental Disorder (PDD), without meeting all of the criteria for autism in the DSM-III-R. Therefore, the estimated prevalence of autism in TSC from this study was 1 in 4. For those with TSC and mental retardation, however, the prevalence was 1 in 2. In addition, autism in TSC seems to affect males and females equally, which differs from the normal 4 to 1 ratio of males to females found in the general population of autistic persons. Hyperactivity was shown by both autistic and non-autistic children with TSC with differing levels of intelligence. Hunt and Shepherd (1993) suspect that hyperactivity itself may be related to the underlying neurological damage associated with TSC. This study suggested that mental retardation and seizures alone were not enough to explain the prevalence of autism in children with TSC. However, because of the relatively small population studied, caution should be used when extrapolating from the results.

The relationship between TSC, autism, and hyperactivity was further examined by Gillberg, Gillberg, and Ahlsen (1994). The purpose of their study was to report findings relating to autistic and hyperactive behavior problems in a total population of children and teens with TSC. The participants were 32 children and teens, representing all known cases of TSC in persons under the age of 20 in the western region of Sweden.

The researchers used a detailed structured interview with parents of children with TSC concerning physical and mental symptoms, an attention deficit score, and

developmental progress. In addition, a semi-structured interview was used to gather information concerning the types of problems considered most serious by the family. The mother of each child was then interviewed in accordance with the Handicaps, Behaviors, and Skills schedule (HBS) (Wing & Gould, 1979), Childhood Autism Rating Scale interview (Schopler et al., 1988), and the Autistic Behavior Checklist (Krug et al., 1980). Finally, data collection concluded with a detailed physical neurological/neurodevelopmental and neuropsychiatric examination of each child.

Of the 32 participants, 11 were “severely retarded”, with an IQ or Vineland social quotient of 50 or less. Seven children had “mild retardation”, with IQ's between 51 and 70. Four children were in the “near average” range (71-84), and six were average or above average (IQ>84).

Over half (17) of the children were diagnosed with Autism, according to the DSM-III-R criteria. Another six children met the criteria for autistic-like conditions. One girl met the requirements for Asperger's Syndrome. Of the 32 children, 11 met the criteria for ADHD and Autism. No child was diagnosed as only ADHD. Only three children were considered completely free of psychiatric or behavioral problems, and all these were of average intelligence. Of the 24 individuals without autistic behavior, 14 had a history of infantile spasms; thus, autistic behavior was not exclusively associated with infantile spasms. However, all but one child with infantile spasms later showed severely autistic behavior.

Gillberg, Gillberg, and Ahlsen concluded that TSC itself predisposes individuals to autism and infantile spasms, rather than infantile spasms being specifically associated with autism. The rate of ADHD found in this study was lower than that reported by Hunt

and Dennis (1987)--43% compared to 59%. In this study, the researchers identified only 1 child with TSC and ADHD that did not also exhibit autistic behavior.

The authors note that, "It would be unrealistic to assume that 86% of all TSC cases show autistic behavior. Even though our study was population-based, we are well aware that a number of mild or asymptomatic cases of TSC may have been missed by our screening" (p.54). In other words, a substantial number of people with normal intelligence and TSC might have been overlooked in the identification process. Nevertheless, autistic behavior is extremely common in children with TSC. In fact, the authors suggest that the triad of mental retardation, epilepsy, and autistic behavior under the age of 5 may have a stronger correlation with a diagnosis of TSC than the traditional triad of mental retardation, epilepsy, and skin marks.

This study also suggests that as many as 9% of all children with autism have TSC. And, given the high rate of females with TSC and autism, 20% of all autism in cases in females might be accounted for by TSC. This is higher than other estimates of 3-5% (Hunt and Shepherd, 1993).

Clearly, autism and TSC are closely linked. There also may be a relationship between hyperactivity/ADHD, autism and TSC. Yet, little is known about what, if any, risk factors predict or influence the development of autism in persons with TSC. This question was explored by Gutierrez, Smalley, & Tanguay (1998). The researchers sought to describe the prevalence and clinical presentation of autism and pervasive developmental disorder (PDD) in a group of persons diagnosed with TSC, and to test the significance of various risk factors which many influence the development of autism in persons with TSC. In addition, the researchers compared rates of psychiatric disorders

among first degree relatives of TSC patients with autism/PDD to rates of psychiatric disorders found among first degree relatives of TSC patients without autism/PDD.

The participants were 28 persons diagnosed with TSC, who were matched with non-TSC autistic participants on the basis of age and mental functioning. Through the study, 8 (28.6%) of the 28 persons with TSC met the criteria for autism. An additional four participants (14.3%) met the broad criteria for PDD. Both TSC autistic persons and non-TSC autistic persons scored similarly on the Autism Diagnostic Interview (ADI) on all domains (social, communication, and repetitive rituals).

Mental retardation was found to be a contributing risk factor for the development of autism in TSC. There were no differences between the TSC autistic/PDD group and the TSC-only group with respect to seizure presence, onset, or frequency. However, the presence of infantile spasms was significantly greater in the TSC autistic/PDD group (58.3%) than in the TSC-only group (16.7%).

Previous studies have suggested that autism is associated with increased incidences of social phobia, mood disorders, and substance abuse among first-degree relatives (Abramson et al., 1992; DeLong & Dwyer, 1988; Piven et al, 1991; Smalley et al., 1994). The current researchers further supported this notion. They found that relatives of the TSC autistic/PDD group had significantly higher rates (50%) of social phobia than did TSC-only group family members (8.3%). Sixty percent of families from the TSC autistic/PDD group had at least one family member with a substance abuse disorder, while none of the TSC-only group families did. In addition, 70% of families from the TSC autistic/PDD group had at least one family member with a major mood disorder, compared with 33.3% of families of the TSC-only group.

This finding--that family members of those with autism/PDD were more likely to exhibit psychiatric and substance-abuse disorders--was noteworthy. It suggests that the relationship between TSC and autism is more complex than previously thought. TSC itself is related to the development of autism, yet risk factors such as mental retardation, infantile spasms, and family history of psychiatric and substance abuse disorders also play a part.

The differences between autistic and non-autistic individuals with TSC were further examined by Baker, Piven, and Sato (1998). The purpose of their study was to estimate the prevalence of autism among a sample of individuals with TSC, and to report any health problems that seemed to be related to the presence or absence of autism. This study is also unique because participants were examined directly through the use of a screening questionnaire, standardized interview, and clinical observation.

The participants were 32 patients in a Pediatric Genetics Clinic, who were assessed using the Autism Behavior Checklist (ABC) (Krug, Arick & Almond, 1980). Parents of patients with ABC scores greater than or equal to 58 were then interviewed with the Autism Diagnostic Interview (ADI; Le Couteur et al., 1989), a semistructured diagnostic interview that includes probes and ratings for behaviors commonly seen in autistic individuals. All subjects meeting ADI criteria were also directly assessed by a psychiatrist experienced in the diagnosis of autism, using the DSM-IV autism criteria. Finally, medical records were reviewed for clinical features contributing to the diagnosis of TSC, family history of TSC, and EEG results. If available in the medical record, previous IQ results were also recorded.

The results of medical records examination showed several differences between autistic-TSC patients and nonautistic-TSC patients. However, as the sample was too small for meaningful statistical analysis, only descriptive statistics were calculated. A history of cardiac involvement was found in 75% of autistic-TSC patients, but only in 12.5% of nonautistic-TSC patients. In addition, 75% of autistic-TSC patients had a history of hypsarrhythmia (abnormal brain waves associated with seizures), while only 12.5% of nonautistic patients did. None of the autistic-TSC patients evidenced eye problems, while 37.5% of nonautistic-TSC patients did evidence eye problems.

Estimates of the prevalence of autism in the general population have ranged from 1 in 2,000 (Lotter, 1966) to 1 in 500 (Centers for Disease Control and Prevention, 1997). The prevalence of TSC in the general population is estimated to be 1 in 6,000 (Whittemore & Roach, 1999). The probability of finding four or more autistic individuals in a sample of 20 from the general population is extremely low. These results further indicate a significant relationship between TSC and autism. In this study, autism was found in 20% of individuals with TSC. This number is somewhat lower than the other estimates summarized in this section, most likely because of the more stringent criteria used for the diagnosis of autism.

In summary, the relationship between TSC and autism is a particularly complex one. The prevalence of autism among children with TSC varies from 25% among all children with TSC to 50% among children with TSC and mental retardation. Among individuals with TSC, mental retardation, infantile spasms, and family history appear to further dispose children towards autism. The rate of autism among children with TSC is evenly split between boys and girls, suggesting that up to 20% of autism in females may

be accounted for by TSC. Thus, TSC itself appears to be related to the development of autism, but other risk factors such as mental retardation, infantile spasms, and family history of psychiatric disorders also play a part.

Sleep Disorders

In addition to seizures, cognitive deficits, and behavioral/psychiatric disorders, another common characteristic of TSC is sleep disorders. Hunt (1993) used a postal questionnaire to gather information on 300 families in which a member was diagnosed with TSC, which indicated sleep disorders were common. (Other aspects of study have been reviewed in previous sections of this chapter.) In 1994, Hunt and Stores probed further into the relationship between epilepsy and sleep disorders in children with tuberous sclerosis. This study was undertaken to obtain a clinical description of sleep problems in a group of children with TSC, for comparison with non-disabled children and children with other learning problems. In addition, this study examined the relationship between sleep disorders and various psychological, social, and biological factors within the TSC groups.

Sleep disorders were investigated in 40 children with TSC and compared with sleep disorders in two groups of non-learning disabled children: siblings of the children with TSC, and a separate group of 37 regular education students matched to the TSC sample. Sleep disorders of the children with TSC were also compared to those in a group of children with various learning disabilities, to determine if sleep disorders were more closely associated with the TSC or with learning difficulties in general. Significantly higher levels of sleep disturbance were found in children with TSC than in any of the other groups of children.

Within the TSC group, sleep disturbance was associated with seizures and daytime behavior disturbances (see also Clements et al, 1986), but was not associated with Pervasive Developmental Delay or high parental stress levels. These results suggest that epilepsy, especially in more severe forms, plays a central role in sleep disturbance. To summarize, then, sleep disturbances are more common in children with TSC than in other children with or without learning disabilities; and are more common for TSC children with seizures and behavior problems.

Communication Disorders

Another common characteristic of TSC is communication problems. Communication problems have been reported in many studies of children with TSC, but not discussed in detail (Hunt & Dennis, 1987; Gomez, 1988). Baltaxe (1998) noted that there were no systematic studies that identify and describe the communication characteristics seen in TSC, and the extent to which they are related to cognitive functioning, seizure disorder, and autism. Therefore, Baltaxe (1998) began to identify areas of communication that are deficient in children and adolescents with TSC.

The purpose of this study was to examine the areas of communication in which children and adolescents with TSC show deficits, as well as the areas in which they function within and below IQ expectancy. In addition, this study examined the communication strengths and weaknesses of children and adolescents in the mentally retarded range, and the developmental, medical, and other characteristics that further differentiate the normal intelligence group in social communication when autistic features are present.

The participants were 34 people with TSC, with a median age of 13. The participants ranged in age from 6 to 23. They were recruited through the NTSA and Tuberous Sclerosis Association of Illinois. All participants were assessed through their medical, developmental, and educational history. Each was also given a complete language evaluation, examining language, speech, and social communication.

Three IQ groups were analyzed separately in terms of language behaviors. The high functioning group had IQ scores of 85 and above, the median functioning group had IQ scores between 70 and 84, and the low functioning group had IQ scores less than 70. Comparisons were made between communication behaviors, IQ levels, and aspects of developmental, medical, and family data.

Nearly 90% of participants had a history of seizures. More than half reported seizures in the first year of life (infantile spasms) and more than half also reported multiple types of seizures. Nearly a quarter of subjects in the normal IQ range had a history of infantile spasms, while 82% of subjects in the below average range reported infantile spasms. All participants had normal hearing; however, more than half had abnormalities of vision. Nearly half reported language delays only, with normal gross motor development. Twenty-three percent reported both gross motor and language milestones delayed.

Baltaxe's study is one of the few that report school placement of participants. In this study, 32% of participants were enrolled in regular classes and 68% were in special education. Of those in special education, more than half were identified as attending "special day classes" (possibly for the mentally handicapped), and more than a quarter

were attending a regular classroom, with pull-out services for special areas. Close to a quarter were enrolled in full-time programs for the learning disabled.

The main focus of Baltaxe's study was on communication behaviors of children with TSC. Most of those with TSC and normal intelligence still had speech, language, and communication deficits in one or more areas. Problems were seen in the areas of complex expressive vocabulary skills, auditory language processing, abstract language skills, and expressive semantic-grammatical skills. Subjects functioned below IQ expectancy in these areas. Areas less affected included receptive vocabulary skills and receptive semantic-grammatical skills.

When IQ was below average, participants still functioned below IQ expectancy in auditory processing and abstract language. A higher percentage of subjects in the below average range also showed speech deficits. The type of seizures, seizure history, developmental language history, and presence of autistic features also affected speech and language functioning. Most children in the study had social communication problems.

This study is important for several reasons. First, it highlights the communication problems children with TSC face in the context of their other characteristics. Second, it is one of very few studies which reports on the educational status of participants in a meaningful way.

TSC and Special Education

Thus far, a basic overview of TSC has been provided, followed by a discussion of the major characteristics associated with TSC. These characteristics included seizure disorders, delayed or impaired cognitive functioning, behavioral or psychiatric problems,

sleep disorders, and communication disorders. Each of these has the potential to greatly impact the educational experiences of children with TSC, and influence the services they receive from schools. Many of these characteristics may be addressed through classroom accommodation and behavior management plans. Yet it is likely that many symptoms are addressed through special education programs. Over the last ten years, the number of students in the United States receiving special education services has grown 30%--an increase that exceeded growth both in US population and in school enrollment (National Household Education Survey of 1999). The increase from 1988 to 1998 was especially evident in the categories of Learning Disability, with an increase of 36.6%, Emotional Disability, with an increase of 21%, Multiple Disability, with an increase of 22%, and Other Health Impairment, with an increase of 318%! No data was kept on Autism services in 1988, so rates of increase could not be calculated. However, the very fact that Autism is now a major category of eligibility suggests that its numbers have increased dramatically in the last ten years.

At the same time, the location and level of these services has changed. The percentage of students served outside the regular classroom for a quarter to a half of their day has decreased 10%. The number of students served outside the regular classroom for less than a quarter of their day has increased 15%. This move to less restrictive services comes as a result of increased reintegration efforts consistent with the Individuals with Disabilities Education Act (IDEA).

Based upon the research completed with children and adolescents with TSC, it is highly probable that many of them receive special education services for learning disabilities, varying levels of mental retardation, autism, speech and language

impairment, emotional or behavioral disabilities, other health impairments, or multiple disabilities. Yet many questions remain unanswered. For example, what percentage of children with TSC receives special education services and what services do they receive? In what setting do they receive services?

Perhaps more important than the category and amount of services received is whether or not these services address the unique range of needs evidenced by many children with TSC. Are parents satisfied that their children's needs have been fully addressed? Special education research in general would seem to indicate that most parents are satisfied with the services their children receive. For example, Lynch and Stein (1982) conducted interviews with 434 parents of students with wide-ranging disabilities and found that 76% were satisfied or very satisfied with their children's current special education placement. Abramson, Willson, Yoshida, and Hagerty (1983) found that 76% of parents whose children received Learning Disability services were moderately to very confident that their children's teachers were improving their children's academic and social abilities. Leyser (1988) surveyed 663 parents of students with disabilities, and found 85-90% were highly satisfied with their children's education. Other researchers found that 91% of parents were satisfied or very satisfied with their children's resource room experience (Lowenbraum, Madge, and Affleck, 1990). However, the reasoning behind the parents' satisfaction in these studies was ambiguous. No direct links had been drawn between parent satisfaction and any specific variable regarding special education services, such as academic gains or self-confidence.

Green and Shinn (1994) looked specifically at the factors that influence parents' satisfaction with special education and views towards reintegration. The participants

were 19 parents and 2 guardians of special education students in grades three through five. Each student had an IEP that included reading and received services in the resource room for less than half the school day. The study used interviews with small groups of parents to obtain information on parents' attitudes about special education services and the factors that form the basis for these attitudes, and parents' attitudes about potential reintegration into general education in light of their current level of satisfaction with special education. The parents overwhelmingly responded that they were very satisfied with the services their children received. In fact, the parents did not identify any areas in which they believed there needed to be improvement. The majority of parents explained their satisfaction with special education services in terms of their child's self-esteem, the caring attitude of the teacher, and the extra "one-on-one" help received. Only 10% rated skill acquisition as the most important factor in their children's reading success. In other words, academic achievement outcomes were not the basis for parental satisfaction.

Perhaps due to their great satisfaction with special education, over half of the parents held extremely negative views towards reintegration. The authors speculated that part of the problem was that exit criterion had not been specified when children entered special education, or, at least, were not remembered by the parents.

Most of the individuals surveyed in studies of parent satisfaction with special education are parents of learning disabled children. This is, in part, because learning disabilities make up fully half of all special education placements. Studies looking specifically at the satisfaction of parents of mentally retarded and emotionally disturbed students showed greater variability in satisfaction, and more reluctance towards mainstreaming (Mlynek, Hannah, & Hamlin, 1982). Other studies demonstrated parent

reluctance to place their children in the more socially stigmatizing categories of emotional disability and mental retardation (Cohen, 1986; Crowell, 1993). Among students who do have more severe disabilities, however, parental satisfaction with educational programs was highly correlated with objective measures of program effectiveness (Rivers, 1990). Thus, parents of students with severe disabilities may be more conscious of student outcomes than parents of students with learning disabilities.

Parent satisfaction can also be influenced by parent involvement in school, parent-school communication, and school climate (Griffith, 1996). In a study of 33,244 parents from 122 elementary schools, Griffith (1996) found that parent-school communication and school climate showed the strongest direct effects on parental satisfaction. Parental involvement contributed little variance to parental satisfaction. However, an interaction effect was found showing the relationship of parental involvement to satisfaction was moderated by how well parents were informed and empowered by the school, and their perceptions of a positive school climate. In other words, parent involvement contributed to parent satisfaction when schools informed and empowered parents, and created a positive atmosphere. In schools that did not empower or inform parents, or that had a less positive climate, parent involvement was not related to parental satisfaction.

Parent involvement, itself, has been linked with student achievement. Ann Henderson completed two extensive literature reviews in 1981 and 1987, concluding that “the evidence is now beyond dispute: parent involvement improves student achievement. When parents are involved, children do better in school, and they go to better schools” (1987; p.1). Important factors of parent involvement were: a variety of roles over a period of time in which parents can be involved, two-way communication, and

involvement which is well-planned, comprehensive, and long-lasting. Some of the major benefits of parent involvement demonstrated in the literature review include higher grades and test scores and long-term academic achievement (Henderson, 1987). Parent involvement has shown positive effects on student learning, but also on student self-esteem, behavior problems, and attendance (Sattes, 1985).

Summary

This chapter has reviewed the history, prevalence, and major characteristics of TSC. Comparisons were made between these characteristics and their occurrence in the general population. The relevance of special education in serving the needs of children with TSC was then discussed, as well as the role of parent satisfaction and involvement with special education.

TSC is a neurological disorder that manifests itself through a variety of symptoms, but is generally identified through a grouping of birthmarks on the skin, seizures, and intellectual impairment. Other common characteristics include behavioral and psychiatric problems, sleep disorders, and communication disorders. The tuberous sclerosis syndrome was described first by Bourneville in the late 1800's, whose 15-year old patient manifested the three core characteristics of TSC—seizures, mental retardation, and facial angiofibromas. It was once thought to be an exceedingly rare disorder that was not often inherited, but more often a spontaneous genetic mutation. Today, estimates place the prevalence of TSC at 1 in 5,800 births.

Approximately 50% of individuals with TSC have mild to profound cognitive impairment, and 80% have experienced seizures. Early onset of seizures is particularly correlated with decreased cognitive functioning. Compared to the general population of

those with seizure disorders, individuals with TSC and seizures have higher rates of intellectual impairment, indicating that the seizures, alone, are not responsible for the deficits.

Individuals with TSC also display higher rates of behavioral and psychiatric disorders, even when compared to other individuals with cognitive impairment and/or seizures who do not have TSC. The three main problems are autistic or autistic-like behaviors, hyperactive or impulsive behavior, and aggressive or destructive behavior. The prevalence of autism among children with TSC ranges from 25% (of all children with TSC) to 50% (among children with comorbid mental retardation).

Sleep disorders and communication disorders are also more common among children with TSC than in children with other types of learning disabilities or seizures. Children with TSC and seizures/behavior problems, in particular, had higher rates of sleep disorders.

Each of these characteristics of TSC has the potential to greatly impact the educational experiences of children with TSC. It is likely that many issues are addressed through special education services. Little is known of the types of services received by children with TSC, or whether these services address the range of needs seen in students. Even less is known of the level of parental satisfaction with the services received.

The purpose of the present study was to examine the impact of these characteristics on the school experiences of children with TSC, including their educational placements, academic and social/emotional functioning, support services received, and educational accommodations. This dissertation will also examine the relationships between students' cognitive, behavioral, and academic functioning, the

schools' responses to the challenges of TSC, and the level of parental satisfaction with the school experiences.

Chapter III

Method

Participants

The participants in this study were the parents of children with TSC, ages 6-17. Participants were obtained through collaboration with the Tuberous Sclerosis Alliance (TSAAlliance), previously known as the National Tuberous Sclerosis Association. The TSAAlliance is the only voluntary health organization in the United States for the genetic disorder known as tuberous sclerosis. Its mission is to find a cure for TSC while improving the lives of those affected. Its membership of over 1100 families includes parents, relatives, and friends of individuals with TSC, adults with TSC, physicians, educators, social workers, and health professionals. This was a random sample of 300 TSAAlliance members, self-identified as parents of children with TSC.

Research Design

This study was a survey design in which data were collected through responses to self-report questionnaires (Achenbach, 1991).

Measures

This study collected demographic information about each child with TSC, including age, gender, grade, school setting, services received, parental involvement, and parental satisfaction through the use of a parent questionnaire. Information about behavior problems was obtained through the use of a child behavior rating scale.

Parent Questionnaire. A parent questionnaire was used to obtain data on the child's age, level of cognitive functioning, school setting (public or private), special education and related services received, amount of time spent in special education per week, parent involvement and parental satisfaction with the school's response to the child's needs. (See Appendix A). The questionnaire was developed by the examiner for use in this study. Questions were modified from those used in similar questionnaires.

The parent involvement questions were modeled after those used by Arnold, Michael, Hosley, and Miller (1994) in their survey of parents of children with mild learning problems. The researchers used a two-stage sampling procedure in a statewide survey of parents of children with disabilities. One hundred fifty-nine school districts were randomly sampled from the 612 districts in Ohio. Of these, 89 districts responded (56%) with confidential lists of addresses for parents of children receiving special education services. From these lists, a simple random sample of 4,962 (15%) participants was selected. The survey was mailed to participants, and follow-up mail prompts were used 10 days, 20 days, and 27 days following the original mailings.

Participants were asked to respond to a 126-item forced-choice questionnaire concerning background factors, demographics, and factual issues. Parents' attitudes towards communication with their child's school were measured using a 5-point likert-type scale. Content validity for these items was demonstrated using a group of experts, including parents and professionals in special education who were members of the state's Developmental Disabilities Planning Council Education Subcommittee. The reliability of the attitudinal questions was assessed using Cronbach's alpha ($\alpha=.85$) from a pilot sample of 70 subjects. A factor analysis was conducted using 369 respondents to

determine the construct validity of the scale (no further information given in the article), with results indicating two items should be dropped. With those items dropped, the remaining items were used as the dependent variable.

The researchers found that parent attitudes towards school were most positively influenced by frequent interactivity between the school and family. Therefore, questions were adapted from the portions of their survey that measured family-school interaction (Arnold, Michael, Hosley, & Miller, 1994). Questions were reworded only slightly to separate out interactions with regular education teachers from interactions with special education teachers. The same response scale is used.

The parent satisfaction questions were modeled after those found in the National Household Education Survey of 1999 (NHES: 1999), completed by the Office of Educational Research and Improvement, part of the U.S. Department of Education's National Center for Educational Statistics. The NHES:1999 was a telephone survey of households in the United States developed by the National Center for Education Statistics. The NHES:1999 was a compilation of key items from previous NHES surveys, and included a parent interview, youth interview, and adult education interview. The study examined a variety of educational topics including early childhood program participation, emerging literacy and numeracy, parent satisfaction with and involvement in the child's education, school practices to involve and support larger families, learning activities with children outside of school, family involvement in learning outside of school, and parent reports about child's postsecondary education plans. Demographic characteristics, household characteristics, and information about the child's health and disability statuses were also collected. Interviews were completed for 24,600 children

ages birth through 20 years old and in the 12th grade or below. This examiner was unable to locate any information regarding reliability and validity of the NHES:1999 survey (NHES:1999 Methodology Report, 2000). Information was included regarding sampling procedures, correcting for sampling error, and weighting of survey data based on demographic information.

The parent satisfaction questions included in the NHES:1999 questionnaire included satisfaction with educational services received, school-home communication, and educational options. These formed the basis for the parent satisfaction questions included in this examiner's parent survey. The questions were reworded only slightly to include more aspects of school that parents may be satisfied or unsatisfied with, such as communication with the school and school discipline. The response scale is the same.

The questions regarding the special education and related services received were derived from the research previously reviewed on the major clinical manifestations of TSC, including cognitive impairment, seizures, autistic characteristics, emotional/behavioral problems, and communication difficulties. Based upon these typical manifestations of TSC, questions were generated which seek to assess the services schools might typically provide to address those issues.

The parent questionnaire used in this study (Appendix A) contained three sections: demographic information, services received, and parent involvement and satisfaction. Section I, Demographic Information, consisted of 6 questions regarding the child's age, gender, age of diagnosis, type of school attended, grade in school, and approximate level of cognitive functioning. Section II, Services Received, consisted of 5 questions regarding special education services received, amount of time in regular vs.

special education, and related services received either through the school or privately. Section III, Parent Involvement and Satisfaction, consisted of 7 questions regarding the number and type of interactions with the school, satisfaction with services received, and changes that parents would like to make. These questions were selected because of their use in previously described studies, and because of their relevancy to the research questions posited in this study.

The final question on the parent questionnaire was an open-ended question in which parents were asked to write-in changes they would like to see in their children's educational programming. These responses were grouped into categories according to topic for qualitative interpretation.

Child Behavior Check-list, Ages 6-18 (CBCL/6-18). The CBCL/6-18 is a revision of the CBCL/4-18. It is a general measure of child and adolescent emotional and behavior problems, designed to be completed by parents. (See Appendix B). It combines a 113-item behavior problems checklist with a seven-part social competency checklist, measuring Internalizing Problems, Externalizing Problems, and Competence. The CBCL/6-18 is perhaps the most researched scale of its kind. Over 1700 empirical studies have employed either the CBCL/6-18, its predecessor, or its preschool counterpart, and it has been used in national evaluation studies, such as the Fort Bragg and the Center for Mental Health Services system of care projects (Furlong & Wood, 2001). The CBCL has been translated into 50 languages and is accepted as the premiere instrument of parent-reported emotional and behavioral problems (Furlong & Wood, 2001).

Reliability for the CBCL/6-18 has been assessed through test-retest reliability, inter-rater agreement, and long-term stability. The internal consistencies of empirically

based problem scales were in the .78 to .97 range. For the DSM-oriented scales, alphas ranged from .72 to .91 (Achenbach & Rescorla, 2001).

Inter-interviewer reliability has also been tested by comparing results of the CBCL's completed with assistance of an interviewer and those filled out by the parents. The overall interclass correlation coefficient (ICC) was .96 for specific problem items (Achenbach & Rescorla, 2001).

According to the 13th Mental Measurements Yearbook, "the CBCL is the standard in the field of child psychopathology against which the validity of other instruments is often measured" (Furlong & Wood, 2001). For this reason, it is difficult to determine the validity of the CBCL itself. Achenbach provides multiple indices that show high concurrent correlations with other instruments purporting to measure similar constructs, such as the Conners' Parent Rating Scale and the Quay Problem Behavior Checklist (Achenbach, 1991). Strong discriminant validity has been demonstrated by the ability of the Total Problems and Social Competency scores, alone and in combination, to appropriately classify matched groups of referred and nonreferred youth (Achenbach, 1991).

Procedure

Pilot Study. Ten parents of children receiving special education services were asked to review the parent questionnaire and attempt to complete it with reference to their own children in special education. Feedback from those parents was gathered regarding the organization and content of the survey. Based upon that information, changes were made to the format and organization of the survey in order to make it more easily read and understood by parents.

Selection of Participants. Three hundred participants for this study were selected through a multi-step process. First, the computer membership database of the TSAlliance was used to select individuals who are self-identified as parents of children with TSC. From this list of approximately 800 parents, employees of the TSAlliance selected every 3rd name. The first name selected was determined through drawing a random number from 800 possibilities. When the end of the list was reached, employees started over at the beginning of the remaining names, until 300 participants were selected. This method of selection was chosen because of its simplicity and convenience for the TSAlliance volunteers who compiled the materials and put the mailing labels on the envelopes. For reasons of confidentiality, this researcher did not view or handle the address labels.

Materials Preparation. Materials were collected and envelope packets were assembled. Each packet included: a letter of support from the TSAlliance, a letter of introduction and instructions from this examiner (Appendix B), explaining the purpose of the study, confidentiality, and informed consent, a parent questionnaire, a CBCL-4-18, and a self-addressed, stamped envelope for the return of packets. To maintain confidentiality, all materials were mailed from the TSAlliance office.

Statistical Analysis

The analysis includes descriptive statistics to gain information regarding the representativeness of the sample, and for the purposes of calculating correlations. Inferential statistics was used to examine relationships between students' needs, special education services, and parental involvement and satisfaction.

Variables: Eight variables were used to complete the statistical analyses:

1. Type of School: This is a categorical variable derived from Question 4 on the parent questionnaire. Parents indicated whether their child with TSC attends public, private, or alternative school.
2. Cognitive Functioning: This is an ordered categorical variable derived from Question 6 on the parent questionnaire. Parents indicated their child's approximate level of cognitive functioning. It is assumed that nearly all students in special education will have undergone intellectual assessment, which is then reported to the parents. Based on this information, parents should be able to estimate their child's level of cognitive functioning. If no testing has been completed, it can be assumed that the child has average or near-average intelligence.
3. Special Education Category: This is a series of dichotomous responses derived from Question 7 on the parent questionnaire. Because parent permission is required before a student can be placed in special education, and because the federal government mandates that parents be involved in the development of an individual education plan, it is assumed that parents will be aware of their child's special education category (or categories).
4. Time Spent in Special Education: This is an ordered categorical variable derived from Question 8 on the parent

questionnaire. Responses were summed to form one variable. Because the federal government mandates that parents be involved in the development of an individual education plan, including decisions regarding the amount of time spent in special education, it is assumed that parents are aware of their child's approximate time spent in special education. To increase the accuracy of parent information, responses were grouped into broad categories: None of the day, less than half the day, about half the day, more than half the day, or all day.

5. Related Services: This is a series of dichotomous responses derived from Questions 10 and 11 on the parent questionnaire. Because parent permission is required before a student can be placed in special education or receive any related services, and because the federal government mandates that parents be involved in the development of an individual education plan, it was assumed that parents will be aware of the related services their child receives.

6. Parental Involvement: This variable is derived from Questions 12, 13, 14, 15 and 16 on the parent questionnaire. Responses were summed to form one variable.

7. Parental Satisfaction: This is a set of likert-type responses derived from Question 17 on the parent questionnaire. Responses were summed to form one variable.

8. Behavior Problems: After CBCL's were scored, T scores for Total Behavior Problems, Internalizing, and Externalizing Problems were used as three separate variables.

Research Questions :

1. What percentage of children with TSC is in special education, and in what setting (type of school) are they educated? To answer this question, percentages and confidence intervals were calculated.
2. Under what special education labels or categories are children with TSC served? To answer this question, percentages and confidence intervals were calculated.
3. What percentage of school time is spent in the special education setting? To answer this question, percentages and confidence intervals were calculated.
4. What percentage of students receives additional or related services, and what are those services? To answer this question, percentages and confidence intervals were calculated.
5. What percentage of children has cognitive functioning in the Above Average, Average, Low Average, or Low ranges? To answer this question, percentages and confidence intervals were calculated.
6. What percentage of parents communicates with their child's teacher or school: Never, 1-3 times each year, 4 or more times each year . To answer this question, percentages and confidence intervals were calculated.
7. What percentage of parents are Very Satisfied, Satisfied, Unsatisfied, or Very Unsatisfied with the school's response to their child's needs? To answer this question, percentages and confidence intervals were calculated.

8. What is the relationship between parental involvement and:
- a. special education label/category.
 - b. behavior problems.
 - c. type of school.
 - d. time spent in special education.
 - e. related services.
 - f. cognitive functioning.

To answer this question, correlations and multiple regressions were calculated.

9. What is the relationship between parental satisfaction and:
- a. special education category.
 - b. behavior problems.
 - c. related services.
 - d. type of school.
 - e. time spent in special education.
 - f. cognitive functioning.

To answer this question, correlations and multiple regressions were calculated.

10. What is the relationship between special education labels or categories and:
- a. behavior problems.
 - b. related services.
 - c. time spent in special education.
 - d. cognitive functioning.

To answer this question, correlations and multiple regressions were calculated.

11. What is the relationship between behavior problems and:

- a. related services.
- b. type of school.
- c. time spent in special education.
- d. cognitive functioning.

To answer this question, correlations were calculated. Product Moment correlations and Point Biserial correlations were used as appropriate, and multiple regressions were calculated.

12. What is the relationship between the age of the student, the level of services received, and the severity of tuberous sclerosis? To answer this question, correlations and multiple regressions will be calculated.

Chapter IV

Results

A total of 166 parent questionnaires and Child Behavior Check-lists were returned, for a total response rate of 55%. This response rate was consistent with previous postal questionnaires of parents of children with TSC (Ferguson, McKinlay, & Hunt, 2002; Hunt, 1993). Of those surveys returned, 88 were usable. The remaining 78 surveys were not in the correct age-range for this study. The response rate for usable surveys was therefore 29%.

The general demographics of the children described by their parents in this study are summarized below. Approximately 58% were male and 42% were female. They were predominantly White/non-Hispanic (See Table 1). The greatest percentage of students were in kindergarten through 5th grade (34.8%), 21.3% were in grades 6-8, and 19.1% were in grades 9-12. In nearly 25% of responses, no grade was given.

Table 1
Percentage of Students by Ethnicity

Ethnicity	Percent	95% C.I.
White/Non-Hispanic	87.5	79-93
Black/African-American	1.1	.2-6
Hispanic	5.7	3-13
Asian/Pacific Islander	5.7	3-13
Native American	0	-
Other	0	-

The majority of students displayed one or more psychological, psychiatric, or medical disorders (see Table 2). Over 80% of children had a seizure disorder, and nearly 40% had a diagnosis of Autism. Autism and ADHD were comorbid in 5% of respondents, ADHD and Oppositional Defiant Disorder were comorbid in 1% of respondents, and Anxiety and Depression were comorbid in 7% of respondents.

Table 2
Number of Psychiatric/Medical Disorders by Gender

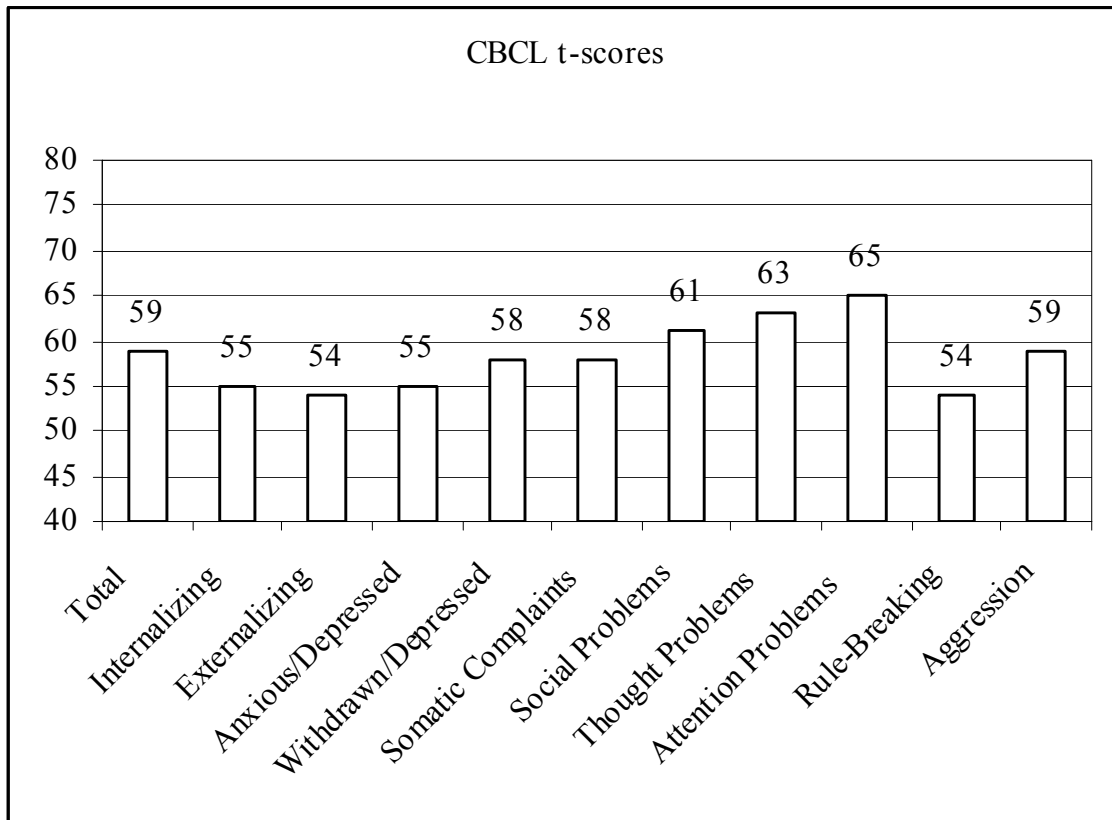
Disorder	Males	Females	% of Sample	% of Population
Anxiety	5	6	12%	59%
Depression	0	7	8%	N/A
Autism	24	11	39%	25%-50%
Seizure Disorder	45	29	83%	80%
Conduct Disorder or Oppositional Defiant Disorder	6	3	10%	N/A
Attention-Deficit Hyperactivity Disorder	8	4	14%	13%
Obsessive Compulsive Disorder	3	3	7%	N/A

Scores on the Child Behavior Check-list, completed by parents of children with TSC indicated a number of behavioral concerns. The average t-score for the Total Behavior Problems Index was 59, which is in the high end of the average range. T-scores

for subscales are reported below (see Figure 1). Data analysis of the Child Behavior Check-list indicated high internal consistency (Cronbach's alpha=.9532).

Figure 1

Mean CBCL t-scores



The results of this study are summarized below according to each research question.

Research Question 1: What percentage of children with TSC is in special education, and in what setting (type of school) are they educated? Ninety-one percent of students were in special education. Sixty-three percent attended regular public schools, 28% attended alternative schools, paid for by the school district, 4% attended regular

private schools, 2.5% attended alternative private schools, and 2.5% attended some other type of school (See Table 3.)

Table 3
Type of School Attended

Type of School	Frequency	Proportion	95% C.I.
Regular Public School	50	.63	.46-.67
Alternative School, paid for by district	22	.28	.17-.35
Regular Private School	3	.04	.01-.10
Alternative school, paid for by parent	2	.03	.01-.08
Other	2	.03	.01-.08

Research Question 2: Under what special education labels or categories are children with TSC served? Approximately 8% of students were classified as Learning Disabled, 6% as Mildly Mentally Retarded, 41% as Moderately or Severely Mentally Retarded, 16% as Autistic, 16% as Other Health Impaired, and 5% as “Other” (See Table 4).

Table 4
Frequency of Special Education Classifications

Classification	Male	Female	Total N	Proportion	95% C.I.
No special education	6	2	8	.09	.05-.17
Learning Disability	6	1	7	.08	.04-.16
Mild Mental Retardation	3	2	5	.06	.03-.13
Moderate or Severe Mental Retardation	15	21	36	.41	.31-.51
Autism	14	0	14	.16	.10-.25
Other Health Impairment	6	8	14	.16	.10-.25
Other	1	3	4	.05	.02-.11
TOTAL N	51	37	88		

Research Question 3: What percentage of school time is spent in the special education setting? Approximately 9% of students did not spend any time in a special education class or with inclusive special education services. Inclusion services are those in which the student remains in the regular classroom, while a special education teacher comes into the classroom and assists the student, and others who may need help. Of the remainder of students, 34% spent less than half the day, 6% spent about half the day,

29% spent more than half the day, and 50% spent all day with special education support (See Table 5).

Table 5
Amount of School Day Spent in Special Education
(Proportion of Students, with 95% Confidence Intervals)

Setting	None	Less than half	About half	More than half	All
Special education class	.21 (.13-.30)	.07 (.03-.14)	.05 (.02-.11)	.26 (.18-.36)	.42 (.32-.53)
Regular classroom with inclusion support	.60 (.50-.70)	.27 (.19-.37)	.01 (.002-.06)	.03 (.01-.10)	.08 (.04-.16)
Regular classroom with no extra support	.82 (.73-.89)	0	.02 (.01-.08)	.07 (.03-.14)	.09 (.05-.17)

Research Question 4: What percentage of students receives additional or related services, and what are those services? Approximately 24% of students did not receive any related services through the school, 24% received one related service, 27% received two related services, and 25% received 3 related services. Approximately 30% of students also received private related services, paid for by parents (See Table 6.) Private related services were defined as services which the parent directly pays for, and includes both students who attend public school and those who attend private school.

Table 6
Summary of Related Services

Group	Related Service	Frequency	Proportion	95% C.I.
PUBLIC	None	21	.24	.16-.34
	Speech/Language Therapy	59	.67	.57-.76
	Physical Therapy	20	.23	.15-.33
	Occupational Therapy	38	.43	.33-.54
	Individual Counseling	6	.07	.03-.14
	Group Counseling	1	.01	.002-.06
	Other	11	.13	.07-.21
PRIVATE	None	61	.69	.59-.78
	Speech/Language Therapy	10	.11	.06-.20
	Physical Therapy	5	.06	.03-.13
	Occupational Therapy	8	.09	.05-.17
	Individual Counseling	7	.08	.04-.16
	Group Counseling	1	.01	.002-.06
	Other	11	.13	.07-.21

Note. “Private” related services were defined as those paid for by parents, and include children who attend public school and those who attend private school.

Research Question 5: What percentage of children has cognitive functioning in the Above Average, Average, Low Average, or Low ranges? For parents who responded to this question, approximately 48% of students had cognitive functioning in the Low range (IQ score below 70), 8% had cognitive functioning in the Low Average range (IQ

score of 70-84), and 20% had Average cognitive functioning (IQ score of 85-115). About 21% of parents did not know their child’s approximate level of cognitive functioning (See Table 7).

Of those students who also had seizure disorders, 25% had Average cognitive functioning, 8% had Low Average cognitive functioning, and 68% had Low cognitive Functioning. Of those students without seizure disorders, 30% had Average cognitive functioning, 20% had Low Average, and 50% had Low cognitive functioning.

Table 7
Summary of Reported Cognitive Functioning

IQ	Frequency	Proportion of Sample	95% C.I.
Low	37	.42	.32-.53
Low Average	6	.07	.03-.14
Average	15	.17	.11-.26
Don’t Know	19	.21	.14-.31
Missing	12	.14	

Research Question 6: What percentage of parents communicates with their child’s teacher or school, through what means, and how often? Approximately 70% of parents communicate with their child’s regular education teacher at least 1-2 times a semester. Approximately 98% of parents communicated with their child’s special education teacher at least 1-2 times a semester (See Table 8).

Table 8
 Percentage of Parent-School Communication
 (With 95% Confidence Intervals)

Communication Type	Never	1-2 times a semester	Monthly	Weekly	Daily
General:					
Regular Education (95% CI)	22.8 (.14-.35)	31.6 (.21-.45)	24.6 (.15-.37)	8.8 (.04-.19)	12.3 (.06-.23)
Special Education (95% CI)	2.5 (.05-.45)	15.2 (.09-.25)	12.7 (.07-.22)	26.6 (.18-.37)	43.0 (.33-.54)
Related Services (95% CI)	7.7 (.03-.17)	56.9 (.45-.68)	15.4 (.09-.26)	12.3 (.06-.23)	7.7 (.03-.17)
Regarding Progress:					
Regular Education (95% CI)	40.0 (.28-.53)	36.4 (.25-.50)	9.1 (.04-.20)	9.1 (.04-.20)	5.5 (.02-.15)
Special Education (95% CI)	7.7 (.04-.16)	25.6 (.17-.36)	14.1 (.08-.24)	14.1 (.08-.24)	38.5 (.29-.50)
Regarding Problems:					
Regular Education (95% CI)	62.3 (.49-.74)	17.0 (.09-.29)	3.8 (.01-.13)	9.4 (.04-.20)	7.5 (.03-.18)
Special Education (95% CI)	19.7 (.12-.30)	14.5 (.08-.24)	10.5 (.05-.19)	18.3 (.11-.29)	36.8 (.27-.41)

Research Question 7: What percentage of parents is Very Satisfied, Satisfied, Unsatisfied, or Very Unsatisfied with the school’s response to their child’s needs? The average rating across all areas of satisfaction was between “Very Satisfied” and “Satisfied” (See Table 9).

Table 9

Parent Satisfaction Percentages
(With 95% Confidence Intervals)

Area	Very Satisfied	Somewhat Satisfied	Somewhat Dissatisfied	Very Dissatisfied
Instructional Program	53.6 (.43-.64)	27.4 (.19-.38)	11.9 (.07-.21)	7.1 (.03-.15)
Special Ed. Program	56.4 (.45-.67)	19.2 (.12-.29)	16.7 (.10-.27)	7.7 (.04-.16)
Related Services	36.2 (.26-.48)	33.3 (.23-.45)	20.3 (.16-.31)	10.1 (.05-.20)
Communication	57.8 (.47-.68)	24.1 (.16-.34)	9.6 (.05-.18)	8.4 (.04-.16)
Discipline	61.0 (.50-.71)	23.2 (.15-.33)	12.2 (.07-.21)	3.7 (.01-.10)
Social Skills	44.0 (.34-.55)	27.4 (.19-.38)	17.9 (.11-.27)	10.7 (.06-.19)

Research Question 8: What is the relationship between parental involvement and:

- a. special education label/category.
- b. behavior problems.
- c. type of school.
- d. time spent in special education.
- e. related services.
- f. cognitive functioning.

Parental involvement, as a single variable, was calculated by adding together the ordinal values assigned to various levels of communication between parent and school, from Section III on the Parent Questionnaire. Parental involvement was positively correlated with attendance at a private school ($r=.455$, $p=.005$), and with private related services ($r=.469$, $p=.004$). The entire correlation matrix is presented in Appendix C.

The data were also analyzed by multiple regression, using as regressors the variables listed in the research question above. Categorical variables (type of school and special education label/category) were dummy-coded to allow for linear regression. Dummy coding was done by making each type of school (regular public school, alternative public school, private school) and each special education label (No Special Education, Learning Disability, etc.) a separate variable. Each of these separate variables was then coded either a 0, if the parent did not mark the item, or 1, if the parent did mark the item. For example, if a parent indicated his child attended regular public school, then the variable “Regular public school” was coded as 1, and all the other type of school variables (private school, alternative public school, etc.) were coded as 0. For each original variable, one dummy coded variable was eliminated from the regression equation on the grounds of multicollinearity. This means that one of the dummy coded variables can be predicted perfectly from the others, and these are the standard grounds for eliminating a variable from a regression equation. This same dummy coding procedure also was used for the remainder of the research questions.

The regression model accounted for 63% of the variance in parent involvement, and the overall relationship was significant ($F=5.22$, $p=.002$). Several of the individual variables had significant effects, including cognitive functioning, total time in special

education, attendance at public schools, and a special education classification of Other Health Impairment (See Table 10.)

Table 10

Regression Model for Parent Involvement

Variable	B	SE	Beta	t	p	Squared Semi-partial Correlation
Total Behavior Problems	.12	.14	.14	.87	.40	.010
Total Time in Special Education	2.78	1.22	.45	2.27	.04	.070
Public Related Services	.69	1.13	.09	.61	.55	.005
Private Related Services	1.01	1.35	.11	.75	.47	.008
IQ	7.69	2.79	.78	2.75	.01	.103
Regular Public School	-15.05	4.63	-.84	-3.25	.01	.144
Alternative Public School	-17.50	4.82	-.91	-3.63	.002	.180
Learning Disability	-6.85	4.61	-.22	-1.49	.16	.030
Autism	-3.78	3.43	-.16	-1.10	.29	.016
Other Health Impairment	-11.86	4.14	-.50	-2.87	.01	.112

Note. R-Square=.782. Adjusted R-Square=.632. N=25. Based on an analysis of the residuals, there appeared to be no consequential departures from normality or homoscedasticity, although there were several influential outliers (all Cook's D < 1.34).

Research Question 9: What is the relationship between parental satisfaction and:

- a. special education category.
- b. behavior problems.
- c. related services.
- d. type of school.
- e. time spent in special education.
- f. cognitive functioning.

Parent satisfaction, as a single variable, was calculated by adding together the ordinal values assigned to various levels of parent satisfaction, from question 18 on the Parent Questionnaire. Responses to this variable were reflected so that higher numbers meant greater satisfaction. Parent satisfaction was negatively correlated with the special education category of autism ($r=-.332$, $p=.008$). Parent satisfaction was positively correlated with parental involvement ($r=.524$, $p=.001$). The entire correlation matrix is presented in Appendix C.

Parents of children receiving autism services were less satisfied in all areas than parents of other children. They were especially dissatisfied with Instruction, Related Services, Special Education Program, and Social Skills. The figures below compare the satisfaction of parents of children receiving Autism services to the satisfaction of all other groups in this study (See Figures 2 through 5).

Figure 2

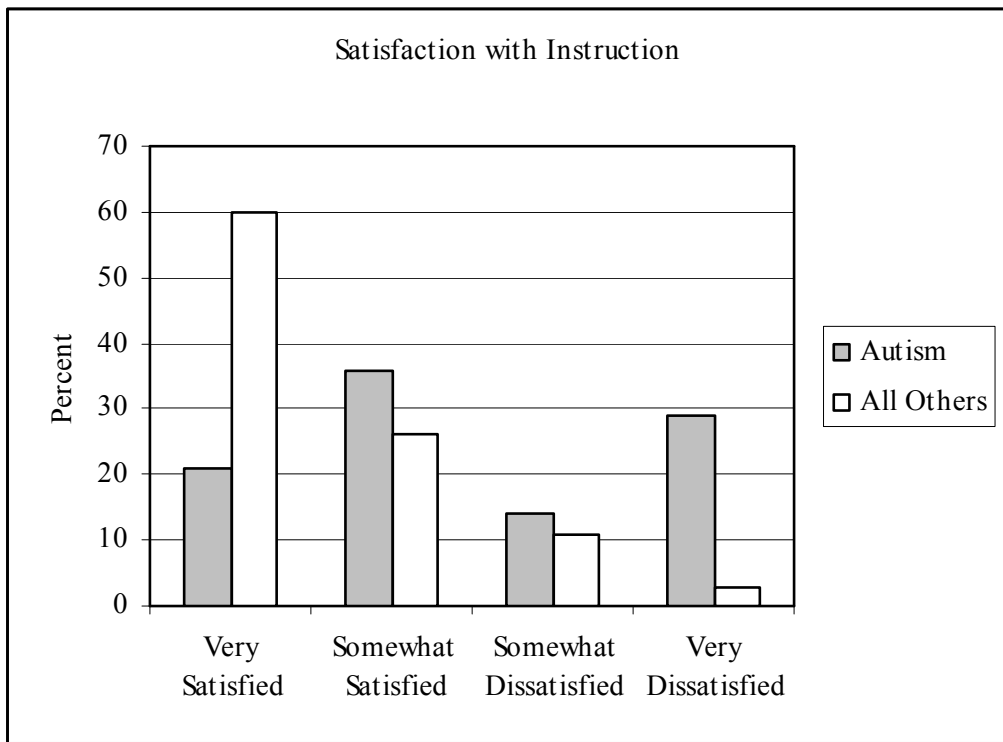


Figure 3

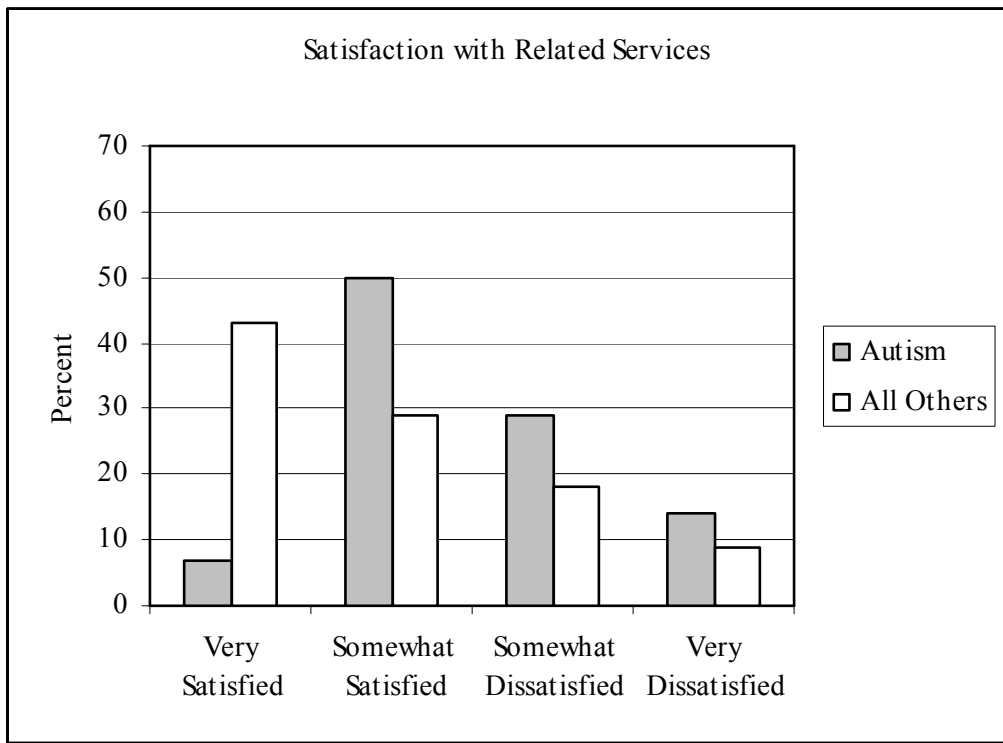


Figure 4

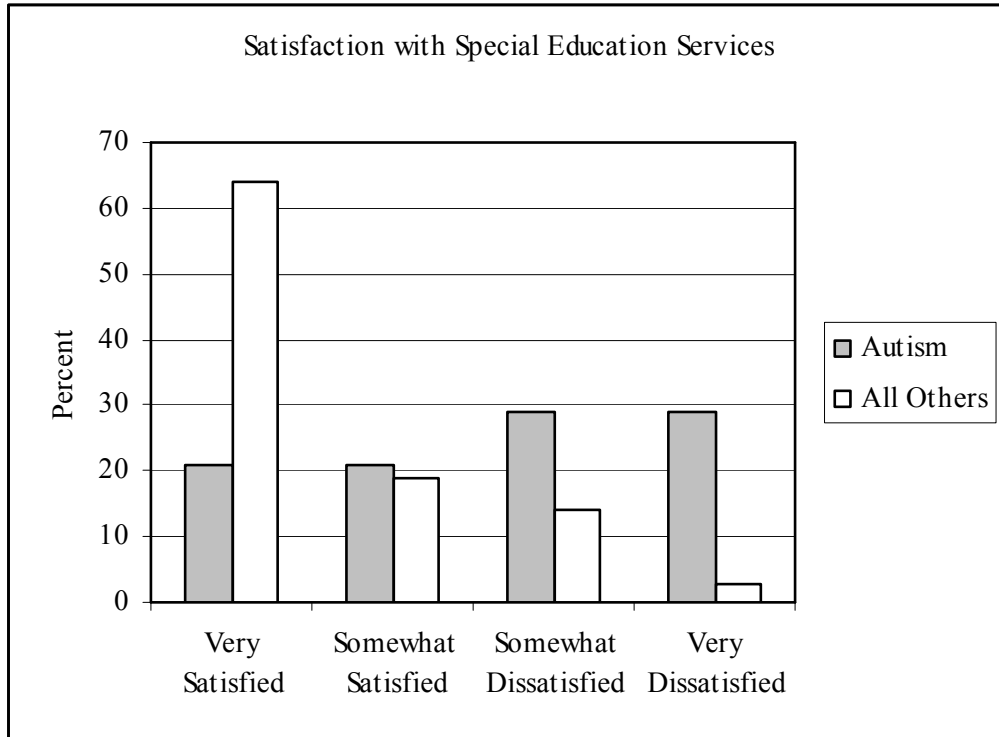
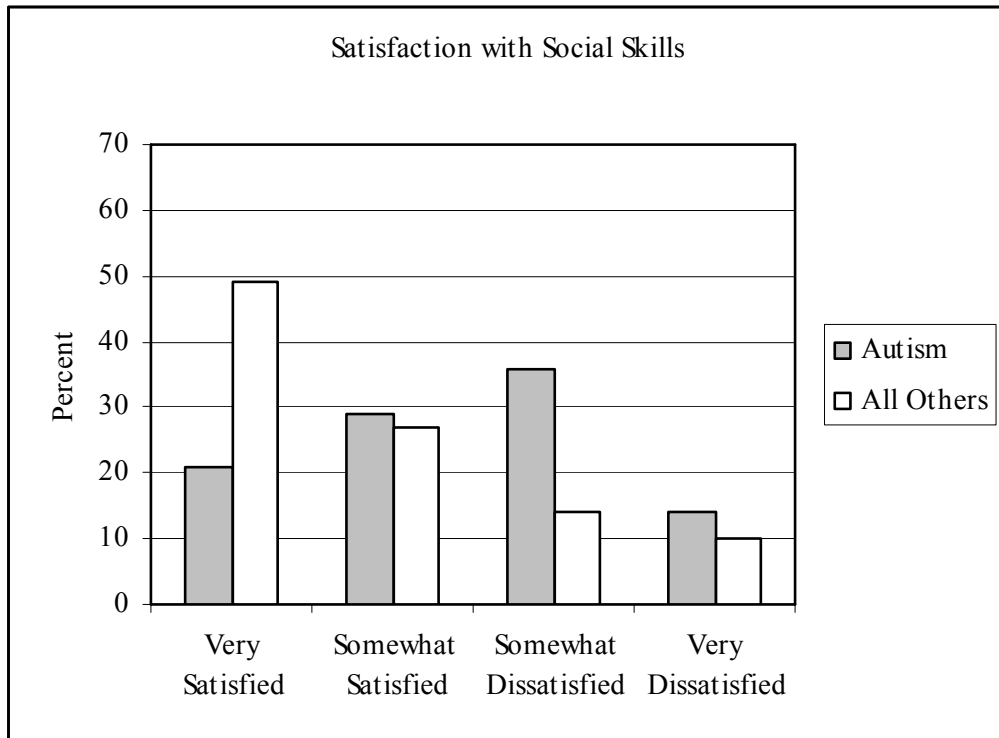


Figure 5



The data were also analyzed by multiple regression, using as regressors the variables listed in the research question above. Categorical variables (type of school and special education label/category) were dummy-coded to allow for linear regression. The regression model accounted for only 19% of the variance in parent satisfaction, and the overall relationship was not significant ($F=1.84$, $p=.084$). The Adjusted R-square value was actually slightly higher when the regression model did not include type of school variables. Only one of the individual variables, total behavior problems, had a significant effect (See Table 11.)

Table 11

Regression Model for Parent Satisfaction

Variable	B	SE	Beta	t	p	Squared Semi-partial Correlation
Total Behavior Problems	-.40	.15	-.65	-2.74	.01	.139
Total time in Special Education	.45	1.71	.09	.26	.79	.001
Public Related Services	1.41	1.05	.24	1.35	.19	.034
Private Related Services	1.16	1.46	.14	.80	.43	.012
IQ	-2.81	2.51	-.40	-1.12	.27	.111

Table 11 (Continued)

Variable	B	SE	Beta	t	p	Squared Semi-partial Correlation
Alternative Public School	-.940	1.73	-.09	-.54	.59	.035
Private School (Regular or Alternative)	-5.46	5.65	-.15	-.97	.34	.017
Learning Disability	.39	.73	.01	.05	.96	<.001
Autism	-4.84	4.88	-.40	-.99	.33	.018
Other Health Impairment	-.46	4.75	-.03	-.10	.92	<.001
Mental Retardation	-.56	4.76	-.05	-.12	.91	<.001
No Special Education	8.39	7.29	.24	1.15	.26	.025

Note. R-Square=.408. Adjusted R-Square=.186. N=88. Based on an analysis of the residuals, there appeared to be no consequential departures from normality or homoscedasticity, nor were there any influential outliers (all Cook's D < .42).

Research Question 10: What is the relationship between special education labels or categories and:

- a. behavior problems.
- b. related services.
- c. time spent in special education.
- d. cognitive functioning.

There were nine special education labels or categories for parents to choose from, including Learning Disability, Mild Mental Retardation, Moderate or Severe Mental Retardation, Behavior/Emotional Disorder, Autism, Other Health Impairment, Orthopedic Impairment, Other, and No Special Education Services. The two categories of Mental Retardation were combined into one category for purposes of correlation and regression analysis. This was done because the category of Mild Mental Retardation had only 5 responses, and because it was thought that parents may have been confused by the separate categories.

Learning Disability was positively correlated with attendance at a private school ($r=.291, p<.006$). A classification of mild, moderate, or severe mental retardation was positively correlated with time in special education ($r=.470, p<.001$) and with externalizing behavior problems ($r=.211, p<.049$). A label of Autism was positively correlated with public related services ($r=.323, <.002$). A label of Other Health Impairment was positively correlated with cognitive functioning ($r=.517, p<.001$), and negatively correlated with time in special education ($r=-.260, p<.014$). The entire correlation matrix is presented in Appendix C.

The data were also analyzed by multiple regression, using Learning Disability, Mental Retardation, Behavior/Emotional Disorder, Autism, Other Health Impairment, and No Special Education Services as predictor variables. The regression model using total behavior problems as the dependent variable, accounted for only 16% of the variance. However, the overall relationship was significant ($F=4.278, p=.002$). The special education categories of Autism ($t=2.707, p=.008$) and Mental Retardation ($t=2.455, p=.016$) had significant effects on the regression model (See Table 12).

Table 12
Regression Model for Total Behavior Problems

Variable	B	SE	Beta	t	p	Squared Semi- partial Correlation
No Special Education	-1.00	5.60	-.03	-.18	.86	<.001
Learning Disability	7.25	5.73	.20	1.27	.21	.016
Autism	14.04	5.16	.52	2.701	.01	.071
Other Health Impairment	8.54	5.19	.32	1.65	.10	.026
Mental Retardation	11.76	4.79	.59	2.46	.02	.058

Note. R-Square=.207. Adjusted R-Square=.159. N=88. Based on an analysis of the residuals, there appeared to be no consequential departures from normality or homoscedasticity, nor were there any influential outliers (all Cook's D < .21).

The regression model for the dependent variable of public related services accounted for 18% of the variance, and the overall relationship was significant ($F=4.694$, $p=.001$). Two variables had a significant effect, No Special Education ($t=-3.229$, $p=.002$), and Learning Disability ($t=-2.366$, $p=.020$) (See Table 13).

Table 13
Regression Model for Public Related Services

Variable	B	SE	Beta	t	p	Squared Semi- partial Correlation
No Special Education	-2.00	.62	-.52	-3.23	.002	.108
Learning Disability	-1.50	.63	-.37	-2.37	.02	.054
Autism	-.14	.57	-.05	-.25	.80	.001
Other Health Impairment	-1.07	.57	-.35	-1.87	.07	.034
Mental Retardation	-1.01	.53	-.46	-1.91	.06	.035

Note. R-Square=.223. Adjusted R-Square=.175. N=88. Based on an analysis of the residuals, there appeared to be no consequential departures from normality or homoscedasticity, nor were there any influential outliers (all Cook's D < .16).

The regression model for the dependent variable of private related services accounted for less than 1% of the variance, and the overall relationship was not significant (F=1.077, p=.379).

The regression model for the dependent variable of total time in special education was calculated without the predictor of No Special Education, since children not receiving special education services would not be spending any time in special education or inclusion classes. The model accounted for 54% of the variance, and the overall relationship was significant (F=26.674, p<.001) (See Table 14).

Table 14
Regression Model for Total Time in Special Education

Variables	B	SE	Beta	t	p	Squared Semi- partial Correlation
Learning Disability	2.35	.46	.45	5.15	<.001	.139
Autism	2.92	.38	.76	7.75	<.001	.316
Other Health Impairment	1.42	.38	.37	3.76	<.001	.074
Mental Retardation	2.97	.31	1.05	9.44	<.001	.469

Note. R-Square=.562. Adjusted R-Square=.541. N=88. Based on an analysis of the residuals, there appeared to be no consequential departures from normality or homoscedasticity, nor were there any influential outliers (all Cook's D < .64).

The regression model for time in a special education classroom or resource class was also calculated without the predictor variable of No Special Education. The model accounted for 40% of the variance, and the overall relationship was significant (F=15.561, p<.001) (See Table 15).

Table 15

Regression Model for Time in Special Education or Resource Class

Variable	B	SE	Beta	t	p	Squared Semi-partial Correlation
Learning Disability	1.33	.58	.23	2.31	.02	.037
Autism	2.41	.48	.56	5.03	<.001	.175
Other Health Impairment	1.05	.48	.25	2.19	.03	.033
Mental Retardation	2.80	.40	.89	7.01	<.001	.339

Note. R-Square=.429. Adjusted R-Square=.401. N=88. Based on an analysis of the residuals, there appeared to be no consequential departures from normality or homoscedasticity, nor were there any influential outliers (all Cook's D < .15).

The regression model for time in inclusion was again calculated without the predictor variable of No Special Education. The model accounted for less than 1% of the variance, and the overall relationship was not significant ($F=1.080$, $p=.372$).

The regression model for cognitive functioning accounted for 51% of the variance, and the overall relationship was significant ($F=16.10$, $p<.001$). Two variables, special education categories of Autism ($t=-4.10$, $p<.001$) and Mental Retardation ($t=-5.58$, $p<.001$), had significant effects.

The regression model for parent satisfaction accounted for less than 8% of the variance, and the overall relationship was not significant ($F=2.122$, $p=.075$).

Research Question 11: What is the relationship between behavior problems and:

- a. related services.
- b. type of school.
- c. time spent in special education.
- d. cognitive functioning.

The Total t-score on the CBCL was used as the single variable of behavior problems. In addition, correlations were calculated between the subscale t-scores and the other variables listed in the research question. Total t-score was positively correlated with public related services ($r=.360$, $p=.001$) and with total time in special education ($r=.358$, $p=.001$). Total t-score was negatively correlated with a category of No Special Education ($r=-.337$, $p=.001$). T-scores for Externalizing behaviors were correlated with the special education categories of Autism ($r=.250$, $p=.019$) and Mental Retardation ($r=.211$, $p=.049$). T-scores for Internalizing behaviors were not significantly correlated with other variables.

Specific subscales of the CBCL were correlated with the variables listed in the research question. T-scores on the Anxious/Depressed subscale were positively correlated with attendance at regular public school ($r=.334$, $p=.003$), and negatively correlated with attendance at an alternative public school ($r=-.307$, $p=.006$). T-scores on the Withdrawn/Depressed subscale were negatively correlated with parental involvement ($r=-.340$, $p=.042$). T-scores on the Somatic Complaints subscale were positively correlated with private related services ($r=.244$, $p=.022$). T-scores on the Social Problems subscale were positively correlated with public related services ($r=.286$,

$p=.007$), and total time in special education ($r=.306$, $p=.004$), and were negatively correlated with cognitive functioning ($r=-.412$, $p=.001$).

T-scores on the Thought Problems subscale were positively correlated with public related services ($r=.326$, $p=.002$), with the special education label of Mental Retardation ($r=.211$, $p=.048$), and with total time in special education ($r=.353$, $p=.001$). Thought Problems were negatively correlated with cognitive functioning ($r=-.580$, $p<.001$).

T-scores on the Attention Problems subscale were positively correlated with public related services ($r=.339$, $p=.001$), total time in special education ($r=.428$, $p<.001$), and a special education category of Mental Retardation ($r=.278$, $p=.009$). Attention Problems were negatively correlated with cognitive functioning ($r=-.549$, $p<.001$) and with parent satisfaction ($r=-.298$, $p=.014$). It should be noted that Attention Problems was the only subscale of the CBCL to be significantly correlated, either positively or negatively, with parent satisfaction.

T-scores on the Rule-Breaking Behavior subscale were positively correlated with public related services ($r=.229$, $p=.032$), and negatively correlated with cognitive functioning ($r=-.336$, $p=.010$). Finally, T-scores on the Aggressive Behavior subscale were positively correlated with public related services ($r=.323$, $p=.002$) and total time in special education ($r=.334$, $p=.001$), and were negatively correlated with cognitive functioning ($r=-.494$, $p<.001$). The entire correlation matrix is presented in Appendix D.

The data were also analyzed by multiple regression, using as regressors the variables listed in the research question above. The only categorical variable (type of school) was dummy-coded to allow for linear regression. The regression model for total behavior problems accounted for 38% of the variance, and the overall relationship was

significant ($F=5.91$, $p<.001$). One of the individual variables, cognitive functioning, had a significant effect ($t=-4.09$, $p<.001$) (See Table 16.)

Table 16
Regression Model for Behavior Problems

Variable	B	SE	Beta	t	p	Squared Semi-partial Correlation
Public Related Services	1.55	1.03	.19	1.51	.14	.025
Private Related Services	1.91	1.24	.18	1.54	.13	.026
IQ	-7.67	1.88	.72	-4.09	<.01	.191
Total time in Special Education	-1.56	1.11	-.24	-1.41	.17	.022
Regular Public School	1.40	7.61	.07	.18	.86	<.001
Alternative Public School	-.85	7.74	-.04	-.11	.91	<.001
Private School (Regular or Alternative)	-13.64	10.68	-.20	-1.28	.21	.018

Note. R-Square=.458. Adjusted R-Square=.381. N=57. Based on an analysis of the residuals, there appeared to be no consequential departures from normality or homoscedasticity, nor were there any influential outliers (all Cook's D < .13).

The subscales were also analyzed using regression, though most did not explain a significant portion of the variance. The regression model for the Anxious/Depressed subscale accounted for only 12% of the variance, and the overall relationship was not

significant ($F=2.125$, $p=.058$). The regression model for Withdrawn/Depressed accounted for less than 1% of the variance, and the overall relationship was not significant ($F=.569$, $p=.778$). The regression model for Somatic Complaints also accounted for less than 1% of the variance, and the overall relationship was not significant ($F=1.711$, $p=.128$). The regression model for Rule-Breaking Behavior accounted for less than 1% of the variance, and the overall relationship was not significant ($F=1.224$, $p=.308$).

Some of the regression models, however, did explain a significant portion of the variance. The regression model for Social Problems accounted for 15% of the variance, and the overall relationship was significant ($F=2.418$, $p=.033$). One variable, cognitive functioning, had a significant effect ($t=-2.907$, $p=.005$).

The regression model for Thought Problems accounted for 41% of the variance, and the overall relationship was significant ($F=6.599$, $p<.001$). One variable, cognitive functioning, had a significant effect ($t=-3.794$, $p<.001$).

The regression model for Attention Problems accounted for 33% of the variance, and the overall relationship was significant ($F=4.996$, $p<.001$). One variable, cognitive functioning, had a significant effect ($t=-2.734$, $p=.009$).

Finally, the regression model for Aggressive Behavior accounted for 32% of the variance, and the overall relationship was significant ($F=4.673$, $p<.001$). One variable, cognitive functioning, had a significant effect ($t=-3.285$, $p=.002$).

Research Question 12: What is the relationship between the age of the student, the level of services received, and the severity of tuberous sclerosis? The level of services was assessed using the variables of public and private related services, time in

inclusion, time in resource, total time in special education, and special education label. The cognitive functioning variable was used to assess the severity of tuberous sclerosis. Correlations were calculated between these variables. Age of the child was positively correlated with a special education category of Mental Retardation ($r=.255$, $p=.016$), and negatively correlated with time in inclusion ($r=-.257$, $p=.016$) and with public related services ($r=-.440$, $p=.000$). The entire correlation matrix is presented in Appendix E.

The data were also analyzed using multiple regression, with Age as the independent variable. The variables for level of services and cognitive functioning accounted for 40% of the variance in age, and the relationship was statistically significant ($F=4.72$, $p<.001$). Five variables in the regression model had a significant effect: public related services ($t=-3.697$, $p=.001$), private related services ($t=2.171$, $p=.035$), and the special education categories of Learning Disability ($t=2.879$, $p=.006$), Autism ($t=2.404$, $p=.020$), and Mental Retardation ($t=2.889$, $p=.006$) (see Table 17).

Table 17
Regression Model for Child's Age

Variable	B	SE	Beta	t	p	Squared Semi-partial Correlation
Time in Inclusion	-.67	.58	-.19	-1.16	.25	.014
Total Time in Special Education	-.97	.87	-.38	-1.12	.27	.013
Public Related Services	-1.69	.46	-.52	-3.70	<.01	.145

Table 17 (Continued)

Variable	B	SE	Beta	t	p	Squared Semi-partial Correlation
Private Related Services	1.12	.52	.27	2.17	.034	.050
No Special Education Services	.92	2.92	.07	.32	.75	.001
Learning Disability	9.92	3.45	.51	2.88	<.01	.088
Autism	5.80	2.41	.63	2.40	.02	.061
Other Health Impairment	4.88	2.45	.49	1.99	.05	.042
Mental Retardation	7.09	2.46	.99	2.89	<.01	.089
IQ	-.99	1.03	-.24	-.96	.34	.164

Note. R-Square=.501. Adjusted R-Square=.395. N=58. Based on an analysis of the residuals, there appeared to be no consequential departures from normality or homoscedasticity, nor were there any influential outliers (all Cook's D < .38).

Additional regression equations were calculated, with cognitive functioning, and with the variables indicating level of services as dependent variables (special education labels were not included as dependent variables, as they were dichotomous). The variables for age and level of services accounted for 80% of the variance in cognitive functioning, and the relationship was statistically significant ($F=23.397$, $p<.001$). Three variables in the regression model had significant effects: time in inclusion ($t=6.104$, $p,.001$), total time in special education ($t=-6.731$, $p<.001$), and the special education category of Learning Disability ($t=2.648$, $p=.011$).

Regressions were then calculated using the variables for level of services as dependent variables, with cognitive functioning and age as independent variables. The regression model using time in special education as the dependent variable, with age and cognitive functioning as independent variables, accounted for 70% of the variance in time in resource, and the overall relationship was significant ($F=68.851$, $p<.001$). The regression model for time in inclusion accounted for only 6% of the variance, and the overall relationship was not significant ($F=2.799$, $p=.070$). The regression model for total time in special education accounted for 54% of the variance, and the overall relationship was significant ($F=34.50$, $p<.001$). The regression model for public related services accounted for 32% of the variance, and the overall relationship was significant ($F=14.192$, $p<.001$). The regression model for private related services accounted for <1% of the variance ($F=.675$, $p=.513$).

Qualitative Information

Though not specifically addressed in the research questions, one item on the parent questionnaire was analyzed qualitatively. Question 19 on the Parent Questionnaire asked what changes parents would like to see in the areas of instruction, special education, related services, communication, discipline, and social skills. Parents' responses were grouped according to general categories. The most frequent change parents requested was additional related services. Parents either believed that their children needed additional time in therapy, or additional types of therapies. The second most frequent change parents requested was additional small-group or one-on-one instruction. Other common changes noted by parents included additional social skills instruction or behavior management, better training for teachers, greater flexibility and

patience among regular education teachers, greater compliance with Individual Education Programs (IEP's), additional instructional time, and Applied Behavior Analysis (ABA) programs for autistic students.

Chapter V

Discussion

This discussion will focus on four topics: the general characteristics of the sample, parent involvement, parent satisfaction, and the relationship between cognitive functioning, scores on the Child Behavior Check-list (CBCL), and seizures.

The first area for discussion is the degree to which the sample approximated characteristics of all students with TSC in the United States. The general characteristics of the sample closely resembled population characteristics in a number of areas, including gender, the presence of psychiatric and medical disorders, and the percentage of students with cognitive impairment. The sample differed somewhat from population characteristics in the area of ethnicity.

The sample included 58% males and 42% females, thus the sample was skewed slightly towards males. Respondents were primarily White/Non-Hispanic, with only one African-American respondent in the sample. This may be a reflection of the membership of the Tuberous Sclerosis Alliance. At the time of this discussion, there was no available ethnic breakdown of the Tuberous Sclerosis Alliance parent membership. However, research has not identified any gender or ethnic variations among individuals with TSC (Hynd & Willis, 1988; Smalley et al, 1994). This suggests that the sample was not representative of the full range of ethnicities represented in the population of individuals with TSC in the United States. The conclusions drawn from this study, therefore, may

primarily apply to White/Non-Hispanic children with TSC. The information may not be valid for children with TSC of other ethnicities.

The ages and grades of students represented in the sample were representative of school-age children. The majority of students were in elementary or middle school, with a smaller percentage in high school. There was no correlation, however, between cognitive functioning and age. Because more severely affected individuals would be more likely to be diagnosed earlier in life, a correlation between age and cognitive functioning might suggest that the younger children in the sample were generally more severely affected than the older children in the sample. However, this was not the case. This suggests that the sample represented the spectrum of affected individuals within each age-range.

Psychiatric and medical disorders, such as autism, anxiety disorders, and attention-deficit disorders may be found in up to 75% of all individuals with TSC (Smalley et al, 1994). The sample in the current study reflects this percentage. In addition, the percentage of children in the sample with a seizure disorder also reflects population statistics. Population research suggests that approximately 80% of all individuals with TSC have seizures (Webb, Fryer, & Osborne, 1996). The current study confirms this research, as 83% of individuals in the sample indicated the presence of a seizure disorder.

The cognitive functioning of the children in the sample differed somewhat from population parameters. About half of the children in this study had cognitive impairment, which closely approximates population parameters. Of those, the vast majority had more severe impairments. This also mirrors previous research on individuals with TSC (Hunt & Lindenbaum, 1984; Webb, Fryer, & Osborne, 1996; Yamamoto et al., 1987). In addition, a very recent study of children with TSC found that IQ was bimodally distributed, with 55.5% in the normal range, 14% with mild to severe impairments, and 30.5% with profound disability (Joinson, et al., 2003). However, only 17% of children in the current sample had Average cognitive functioning, compared with approximately half of the population of individuals with TSC. It should be noted that individuals with normal cognitive functioning may not be diagnosed until later in life, or even possibly not until they have a more severely affected child. Thus, the sample in the present study may be representative of the population of *children identified* with TSC, in the United States. It may also reflect the membership of the Tuberous Sclerosis Alliance. Parents of more severely affected children may be more likely to join an affinity group than parents of mildly affected children. This will be discussed further in the section on limitations.

It is difficult to determine whether the sample reflects the general population in the area of special education placement, because there are no population statistics with which to compare placement. In a study of communication disorders in children with TSC, Baltaxe (1998) reported that 68% of participants were enrolled in special education. Of those students, more than half were identified as attending “special day class” (possibly for the mentally handicapped), and more than a quarter were attending a regular classroom with pull-out services for special areas. Close to a quarter were enrolled in

full-time programs for the learning disabled. However, the actual categories of special education were not reported, so it is impossible to know what types of services students actually received. In addition, some of the participants were as old as 23, which may explain why the percentage of participants receiving special education was lower in the Baltaxe study than in the current sample.

Ninety-one percent of children in the current sample received some type of special education services. Based upon the incidence of cognitive impairments, seizures, communication disorders, behavior disorders, psychiatric disorders and autism reported in children with TSC, this percentage seems to approximate what might be expected among the general population of students with TSC. In addition, 91% of children in the sample attended public school.

Upon examination of specific categories of special education some interesting differences between males and females appeared. Twenty-one females received special education services for Mental Retardation, compared with 15 males. Considering there were more males than females in the sample, this discrepancy is notable. However, another difference may help explain the discrepancy. All 14 of the students receiving special education services for Autism were males. Prior research suggests the incidence of autism in TSC is approximately equal between males and females (Hunt & Shepherd, 1993), thus one would expect an approximately equal number of males and females to receive Autism services in school. Since more girls received Mental Retardation services, and more boys received Autism services, it may be that school personnel are more likely to recognize and properly identify signs of autism in boys than in girls. Certain autistic features are also seen in students with mental retardation, thus those

characteristics may have been attributed by school personnel to mental retardation in girls, and to autism in boys. If this is the case, school psychologists and other school personnel should pay special attention to the services required by girls with TSC, especially with respect to autism.

The time students spend in special education is another area where little population information is available for comparison. In the sample, approximately 50% of students received all-day special education support in either a special education class or an inclusion class, and 42% received all-day self-contained services. This seems congruent with the cognitive functioning of students in the sample. It also reflects estimates of the cognitive functioning of children with TSC in the population (Joinson, et al., 2003).

There is almost no population information with which to compare the types of related services received by students with TSC. In the current sample, 75% received one or more related services through the public school, the most common of which were Speech and Language Therapy (67%), Occupational Therapy (43%), and Physical Therapy (23%). Thirty percent received one or more private related services, the most common of which were Speech and Language Therapy (11%) and Occupational Therapy (9%). This seems to be a relatively high percentage of parents seeking private related services, given that public schools provide these services free of charge. Upon examination of the sample data, it is evident that the majority of children attending private schools received private related services. However, more than a quarter of children attending public schools also received private related services. This suggests that schools are not providing all the related services students require. The Office of

Civil Rights has leveled consistent criticism towards state departments of education, asserting that schools are not providing related services in required numbers to students in special education (G. Batsche, personal communication, October 20, 2003). Even if students received related services through the public schools, parents may think that their children could benefit from additional time or services that the school is unwilling to provide. Many school districts provide related services (only if those students are identified as special education students) for children attending private school, as well, if parents transport students to the public school for those services. Parents of children attending private school may not realize that those services are sometimes available to them. It would appear that IEP teams need to do a better job of communicating with parents regarding the perceived needs of their children, assessing those needs, and seeking to accommodate those needs through public related services.

In general, the high percentage of students receiving publicly funded Speech and Language Therapy was expected because of the communication disorders associated with TSC. However, the high percentage of students receiving Occupational and Physical Therapies was unexpected. This suggests that motor functioning and coordination is impaired in many students with TSC. Some of the impairment could be related to mental retardation or autism, which are sometimes associated with deficits in these areas. However, upon examination of the students receiving Physical Therapy through the public school, only 45% were also receiving services for Mental Retardation and 15% for Autism. The remaining students were divided between the special education categories of Other Health Impaired, and "Other". Of the students receiving Occupational Therapy through the public school, only 48% were students receiving services for Mental

Retardation, while 32% were in an Autism program. The remaining students were again divided between the special education categories of Other Health Impaired and “Other”. Therefore, the incidence of Occupational and Physical Therapy services was not related solely to cognitive impairment or autism. None of the research reviewed in Chapter III indicated such a high prevalence of motor delays requiring Occupational or Physical Therapies. School personnel, as well as physicians treating students with TSC, need to be aware that motor delays might be another central characteristic of children with TSC.

Overall, despite a lack of ethnic diversity, the sample appeared to reflect the general population of children with TSC in the areas of gender, psychiatric and medical disorders and cognitive impairment. The sample differed in the percentage of individuals with average cognitive functioning. Because no prior research was found on the specific types of special education and related services students received, no comparison can be made between the sample and population in these areas. However, the special education services received by the sample do seem to reflect the major characteristics of children with TSC.

The second area for discussion is parent involvement. Of the parents whose children received special education services, 43% communicated with teachers daily, 27% weekly, 13% monthly, and 15% communicated with teachers once or twice a semester. A very positive finding was that parents and teachers were more likely to communicate regarding a child’s progress than about a child’s problems.

There was a positive correlation between parent involvement and enrollment in private school or private related services. This may suggest one of two things. First, private schools and private related service providers may encourage and facilitate more

parent communication and involvement. Second, highly involved parents may be more likely to choose private schools and private related services than parents who are less involved. In the regression analysis, public school attendance accounted for a significant portion of the variability in parent involvement, indicating that public school attendance is predictive of lower levels of involvement. Cognitive functioning also accounted for a significant portion of the variability in parent involvement, with higher levels of cognitive functioning predictive of higher levels of involvement.

Parent involvement was also positively correlated with parent satisfaction. Involved parents were more likely to be satisfied with the services their children received in all areas. This finding is similar to that of Griffith (1996), who found that parent-school communication and school climate showed the strongest direct effects on parental satisfaction.

Parent involvement was negatively correlated with t-scores on the Withdrawn/Depressed scale on the CBCL. This was, in fact, the only scale of the CBCL that was significantly correlated with parent involvement. Parent involvement may have a hand in reducing withdrawn-depressed behaviors in students. The argument could be made that, since depression is sometimes hereditary, students showing symptoms of withdrawn-depressed behavior are more likely to have parents who are themselves depressed, making involvement difficult. However, involvement was not just measured in terms of parent effort, but also the school's efforts at communicating with and involving parents. Thus, it was not entirely dependent on parent response. In addition, parent involvement was not correlated with t-scores on the Anxious/Depressed scale on the CBCL. If parent depression accounted for the lack of involvement, parents of

children with symptoms of anxiety and depression might also be less involved, but this was not the case.

One explanation for this correlation could be that students who are withdrawn in school are less likely to be noticed by teachers than students with other types of behavioral concerns. Teachers may be less likely to seek out opportunities to communicate with parents of children who are quiet and unobtrusive, even if this behavior is part of a pattern of withdrawn-depressed behavior.

Overall, the information on parent involvement suggests several actions for school personnel. First, public school psychologists, teachers, administrators, and other school personnel should continue their efforts to involve parents in their children's education, as it has been linked to improved student achievement (Henderson, 1981, 1987), as well as improved self-esteem, behavior, and attendance (Sattes, 1985). Second, school personnel may need to make additional efforts to involve parents of lower functioning children with TSC, as they were less likely to be involved than parents of higher functioning children with TSC. Third, school personnel should make additional efforts to involve parents of children with TSC who exhibit withdrawn-depressed behavior. It is possible that parent involvement itself is a protective factor against withdrawn-depressed behavior.

Educators should be aware of several "red flags" which signal the need for specific intervention in the area of parent involvement. These include lower cognitive functioning and withdrawn/depressed behavior, together with low parent involvement. Together, these red flags may indicate high needs families. School personnel should

watch for these variables and actively become involved in increasing parent involvement when these variables are noted.

Finally, teachers and related service providers should continue to communicate with parents regarding their children's progress, and not just regarding problems encountered. This facilitates positive two-way communication between parents and school personnel.

The third topic for discussion is parent satisfaction. The majority of parents in this study were very satisfied or somewhat satisfied with schools' efforts on behalf of their children with TSC. The areas of greatest satisfaction for all participants were Discipline, Instruction, and Communication. The areas of least satisfaction were Social Skills and Related Services. However, even in these areas, the majority of parents were satisfied.

There was a negative correlation between parent satisfaction and the special education category of Autism. Parents of children receiving Autism services in school were less satisfied in all areas than parents of other children. Parents of autistic students were especially dissatisfied with Instruction, Related Services, Special Education Program, and Social Skills.

There are a number of possible explanations as to why parents of children receiving Autism services are generally less satisfied than other parents. First, this may be merely a reflection of the dissatisfaction that many parents of children receiving Autism services, with or without TSC, experience. Children with autism experience more deficits in the area of social skills than other children, making educational planning more difficult. Parents may be expressing their dissatisfaction with this area, regardless

of comorbid TSC. At least two recent studies have been undertaken to examine the parent satisfaction of children with autism or pervasive developmental disorders (PDD) in public school. The most recent study surveyed parents of children with autism or PDD who entered public school following intensive behavioral treatment in a private center-based program (DiPietro, et al., 2002). The researchers reported that parents were uniformly satisfied with the services their children were receiving in public schools. The second study surveyed parents of students with autism pervasive developmental disorders regarding their perceptions of, and satisfaction with, educational services (Starr, Foy, & Cramer, 2001). Overall, parents of nonverbal children, parents of younger children, and parents of more than one child with PDD rated classroom environment and education team variables more highly than did other parents of children with PDD. However, parents described themselves as generally satisfied with services. Based upon these studies, it would appear that the presence of autism, alone, does not explain lowered parent satisfaction levels.

An alternative explanation is that, because autism differs slightly in children with TSC (Smalley, Tanguay, Smith, & Gutierrez, 1992), the services they receive must also differ slightly from typical services for children with autism. If schools have not yet addressed these differences, parents may be left feeling that their children's needs have not been met. The area in which they differed, however, was that children with TSC had fewer stereotypical or repetitive behaviors than other autistic children. It seems unlikely that this would negatively influence parents' perceptions of the Autism services their children receive. Neither of these explanations seems satisfactory. Overall, it was

unclear why parent satisfaction with Autism services was so low. This is clearly an area for future research.

Traditionally, students with autism in public schools have not received the services they require, and have often been mis-categorized and placed into classrooms for emotionally disturbed students. As a result, autism advocacy groups have taken a lead in lobbying for expanded services for students with autism in the public schools. These advocacy groups have legitimate complaints regarding school services and hence may present a negative view of public school services. It is possible that parents of children with autism have also adopted this negative view, more so than parents of other children with special needs. This is another possible explanation as to why parents of children receiving autism services in this study were significantly less satisfied than other parents.

Parent satisfaction was also negatively correlated with t-scores on the Attention Problems scale of the CBCL. This was the only CBCL scale correlated with parent satisfaction. Again, this may not be unique to parents of children with TSC. Many parents of children with attention difficulties may feel dissatisfied with schools' ability to adapt instruction to meet their children's specific needs. Though not significantly correlated, t-scores on the Total Behavior Problems scale of the CBCL accounted for a significant portion of the variance in parental satisfaction. Not surprisingly, parents of children with lower t-scores on the Total Behavior Problems index were generally more satisfied with their children's education.

Overall, this study suggests that school personnel can increase parent satisfaction in several ways. First, psychologists, teachers, and other school personnel can increase parent satisfaction by decreasing the severity of behavior problems children with TSC

experience. As professionals with expertise in behavior management and consultation, school psychologists are in a unique position to assist in this area. Second, teachers, administrators, and school psychologists may increase satisfaction by closely examining their Autism services. By communicating with parents regarding their concerns, school personnel can work to provide improved services to children with TSC.

The fourth topic for discussion includes cognitive functioning, CBCL scores, and seizures. The majority of children in this study were in the Low range of cognitive functioning (IQ scores less than 70). Over 20% of parents indicated that they did not know their child's approximate level of cognitive functioning. This was somewhat surprising, given that nearly all of the children in the study received special education services. Intellectual assessment is generally a core component of special education decisions. None of the parents indicated a cognitive functioning range of Above Average for their children with TSC. A very recent study found that cognitive functioning tended to be bimodally distributed among individuals with TSC (Joinson, et al., 2003). Using a large, epidemiological sample in England, the researchers found that 55.5% had an IQ in the normal range, 14% had mild to severe impairments, and 30.5% had profound disability (IQ<21). In total, they found that 44% of individuals with TSC had an IQ score below 70. This statistic is very similar to the 42% of children with TSC with an IQ score below 70 found in the present study.

Predictably, Low cognitive functioning was associated with the special education category of Mental Retardation. Of children whose cognitive functioning was in the Low range, 70% received special education services for Mental Retardation, and 24% received services for Autism.

Children with Low Average intelligence seemed to be spread over several categories of special education. Of children whose cognitive functioning was in the Low Average range (IQ=70-84), 17% received no special education services, 33% received services for a Learning Disability, 33% received services for Mild Mental Retardation, and 17% received services for an Other Health Impairment.

Average cognitive functioning was closely associated with the special education category of Other Health Impairment. Of children whose cognitive functioning was in the Average range, (IQ=85-114), 27% received no special education services, 13% received services for Autism, and 47% received services for an Other Health Impairment. The special education category of Other Health Impairment (OHI) is sometimes used for diagnoses of attention-deficit hyperactivity disorder (ADHD). In this study, parents were not asked to specify the medical diagnosis associated with an OHI eligibility. However, TSC itself could qualify as the medical diagnosis associated with an eligibility of OHI.

Based on the responses in this study, cognitive functioning alone is not generally a good predictor of special education services. Even children with Low cognitive functioning were divided between services for Mental Retardation and services for Autism. It is notable that 17% of children with Low Average cognitive functioning were not receiving any special education services. These are likely to be the children that fall in the “Slow Learner” range, and do not qualify for Learning Disability services because they lack a discrepancy between their IQ and Achievement scores. It is often these children that remain unidentified by special education, who experience significant behavioral and academic difficulties in school.

The relationship between cognitive functioning and scores on the CBCL was also examined. Children with the Low Average intelligence had slightly higher t-scores on the Social Problems, Attention Problems, and Aggression scales. In most other areas, children with Low functioning had higher t-scores for behavior problems than the other two groups. There was a significant, negative correlation between cognitive functioning and t-scores on the Social Problems, Thought Problems, Rule-Breaking, and Aggression scales on the CBCL. Again, these problems are more likely to be associated with lower cognitive functioning.

In contrast, children with Average cognitive functioning had higher scores on the Anxious/Depressed scale than children with either Low Average or Low functioning. These results suggest that even children with TSC and average cognitive functioning may have significant emotional or behavioral concerns, especially in the areas of anxiety or depression. They also suggest possible areas of behavioral intervention for children with TSC. School psychologists can work with parents and teachers to address the specific behavior concerns of children with TSC in their schools.

Several recent studies have examined the relationship between intellectual disability and behavior problems, using the CBCL. These studies found significantly higher mean scores on the CBCL among children with intellectual disability, than among children randomly selected from the general population (Dekker, Koot, van der Ende, & Verhulst, 2002; Baker, Blacher, Crnic, & Edelbrock, 2002.) One study found that almost 50% of children with intellectual disability had a Total Problem score in the borderline or clinical range, compared to about 18% of children without intellectual disability (Dekker, Koot, van der Ende, & Verhulst, 2002). The researchers noted that the most prominent

problem behavior areas were Social Problems, Attention Problems, and Aggressive Behavior. In the present study, Social Problems, Attention Problems, and Thought Problems had the highest mean t-scores among children with TSC (61, 65, and 63, respectively.) None of the mean scores were in the clinical range of significance, and only one mean score, Attention Problems, was in the borderline range of significance. However, the highest percentages of students receiving scores in the borderline or clinically significant ranges were found in the areas of Attention Problems (49%), Thought Problems (43%), Aggression (27%), and Total Problems (32%).

At least one other study examined the relationship between autism and behavior problems, using the CBCL (Boelte, Dickhut, & Poustka, 1999.) The researchers found that autistic children could be identified by higher scores on the scales measuring attention problems, social problems, and thought problems, and low scores on the scale for somatic complaints. In the current sample, children receiving Autism services also had high scores for attention problems, social problems, and thought problems. In addition, they had higher scores for aggression.

Upon examination, most scales of the CBCL were not able to distinguish between different special education categories in the current sample. However, the Social Problems, Thought Problems, and Attention Problems subscales could significantly discriminate between children receiving Mental Retardation services and children receiving no special education. The Thought Problems and Attention Problems subscales could also significantly discriminate between children receiving Autism services and children receiving no special education.

The relationship between cognitive functioning and seizures was also examined. In the current study, seizures were present in 80% of children with Average intelligence, 67% of children with Low Average intelligence, and 87% of children with Low intelligence. Population research suggests that approximately 80% of all individuals with TSC have seizures (Webb, Fryer, & Osborne, 1996). Of those individuals, approximately a third has normal intelligence (Shepherd & Stephenson, 1992). The present study did not examine the age of onset of seizures or the types of seizures—both of which have been associated with cognitive deficits. But the sample did replicate the relationship between seizures and intellectual impairment demonstrated in previous studies.

The results from this study regarding cognitive functioning, CBCL scores, and seizures suggest several conclusions. First, children with TSC and Low Average intelligence may not be receiving needed special education services. Second, children with TSC and Average intelligence may still have significant emotional or behavioral concerns. Finally, teachers serving children receiving services for Autism and Mental Retardation, in particular, may need continued assistance with behavior management to address the behavior difficulties of children with TSC.

Contributions of this Research Study

This research is important to the study of children with TSC for several reasons. First, it provided basic data that was not previously available regarding the percentage of children with TSC receiving special education services, the eligibility category of those services (Learning Disability, Mental Retardation, Autism, etc.), the amount of time during the school day that children receive special education services, and the setting (regular classroom with inclusion assistance, or resource room) in which those services

are delivered. This information can form the foundation of future research into the specific needs of children with TSC and whether those needs are fully addressed through the school system. It also provides knowledge with which parents of children with TSC can equip themselves to advocate for their school-age children.

In addition, this study contributed basic information about the types of related services (speech and language therapy, occupational therapy, physical therapy, etc.) that children with TSC receive, and whether those services are provided by public schools, or if they are privately pursued by parents. Again, this information can enable schools to examine the extent of their related services offerings, and whether those are sufficient to meet the needs of children with TSC. Future research directions have also been suggested by the high incidence of students receiving occupational and physical therapies. At present few research studies were found to address motor skills deficits in children with TSC.

The present study will also contribute to the school services of children with TSC by providing information to school personnel regarding the behavior problems experienced by children with TSC. This information can be used to suggest areas for behavioral intervention for school psychologists and all school personnel who work with children with TSC.

This study brings to light a critical area in which public schools need to improve and expand services, the area of mental health services. The majority of children in this study had at least one type of psychiatric diagnosis, as well as behavioral concerns expressed by parents. The only school related services specifically designed to address those issues would be individual or group counseling. Yet, these were the related

services least utilized by schools. There are many ways schools could address the behavioral and psychiatric demands of children with TSC, including classroom behavior management support from behavior specialists, school psychologists, and other professionals with expertise in the area of behavior. Even with the smaller class sizes special education allows for, teachers are often overwhelmed by the number and severity of behavior problems they must address. In addition, training provided by teacher training programs in the area of behavior management is generally inadequate, especially as concerns more difficult behaviors and students with lower cognitive functioning. The challenge for today's teachers is no longer simply to teach the subject matter, but to do so while contending with increased violence in the schools, disruptive classroom behaviors, parent support that is often minimal or non-existent, and greater pressure due to high-stakes testing. Mental health services is the one area most needed and most ignored in public schools today.

The Tuberous Sclerosis Alliance can serve an important function in educating parents about their special education rights and the importance of parent advocacy in the special education process. This study suggested that children with TSC and low-average intelligence had significant behavior concerns (and most likely, academic concerns, as well) that were not currently addressed through special education. Parents who are informed about the types of special education categories and services available to their children are better able to ask for and are more likely to receive those services from schools. This is especially important for older students, who may be transitioning from school into vocational settings.

Limitations

All survey research has limitations. Surveys cannot probe more deeply into respondents' opinions and actions (Gall, Borg, & Gall, 1996). Surveys are also subject to sample bias. The sample that participated in this study appeared to be representative of the population in a number of important areas, including gender, psychiatric and medical disorders, and cognitive impairment. Because no prior research was found on the specific types of special education and related services students received, no comparison can be made between the sample and population in these areas. However, the sample was not representative of the ethnic diversity of children with TSC. In addition, it may not have fully represented the population of children with TSC who have normal intelligence.

A second limitation of this study is that the information gathered in the questionnaires is on *reported* clinical manifestations of TSC, services received, and parent involvement, rather than on *actual* medical records, special education records, and objective measures of parental involvement. Future studies could reduce or eliminate this consideration simply by gathering school records and teacher behavior rating scales as comparison information.

A third limitation is the potential for response sets, such as social desirability. Participants may have changed their responses to questionnaire items in an effort to present themselves in a favorable light ("faking good"), or they may have exaggerated the extent of problems ("faking bad"). The CBCL does not contain a "lie scale" because of the developers' desire to "restrict the CBCL to items that are meaningful in themselves...and because scores should never be used to make clinical judgments in

isolation from other information” (p.237). Though subject to possible bias, questionnaires are one of the most time-and cost-efficient methods of collecting data that otherwise would require observers and researchers months, or sometimes years, to gather (Gall, Borg, & Gall, 1996). Interpretations from this questionnaire must be made cautiously, as the information is primarily correlational in nature.

A fourth limitation is the difficulty associated with identifying TSC at early ages, and the difficulty of collecting a large sample of children with TSC. First, the prevalence of TSC itself has been difficult to establish because of the number of undiagnosed cases at any point in time. Individuals with milder forms of TSC may not be diagnosed until their teens, with the average age of diagnosis ranging from 11-15 years old among children and adolescents (Webb, Osborne, & Fryer, 1991). The sample completing this questionnaire may represent parents of more severely affected children, who were diagnosed earlier in life due to their more obvious characteristics of TSC. Although the cognitive functioning of the sample appears to reflect that of the population, the percentage of students receiving special education services is higher than in other studies, which may indicate that a more severely affected sample was obtained. However, previous studies did not gather information on specific special education services received, thus it is difficult to estimate the true population parameters for this variable.

The fifth limitation occurs as a result of using members of the TSAlliance as participants. The problems of using families from a patient’s association are significant, since often only those with a more severely affected family member join those groups. However, because of the relatively unknown status of TSC and the difficulty in obtaining information about it, the membership of the TSAlliance may represent a wider spectrum

of the disorder (Hunt & Dennis, 1987). Furthermore, it would be nearly impossible to obtain a large enough sample for this type of study without using a pre-defined population of individuals with TSC, such as the membership of the TSAAlliance.

The sixth limitation occurs as a result of the large number of analyses conducted in this study. With the increased number of analyses, the chance of a Type I error rate increases, and can no longer be stringently controlled.

The final, and perhaps most correctable limitation of this study was the sample size. Due to the difficulty in recruiting participants that fit the parameters of this study, the response size was smaller than hoped for. However, the sample did appear to be representative of the population in substantial ways.

Future Research

Future research should focus on gathering more objective information regarding special education categories, services received, and time. In addition, the information from parent's behavior rating scales can be combined with that of teachers, to elicit a more complete picture of children's behaviors.

Ethnic diversity may be increased by adding the membership of state and national TSC affiliation groups. The addition of state groups may increase the percentage of minorities participating in the study, so that the sample can approximate population parameters.

Future research should examine the incidence of motor impairment in children with TSC, given the high percentage of children receiving Occupational or Physical Therapies. In addition, research on related services should examine the reasons why

parents choose private related services, when public related services are available to their children.

Research should also continue into the relationship between Autism and parent satisfaction. Future research could compare the satisfaction of parents of children with Autism in the general population, versus parents of children diagnosed with TSC and Autism. If both groups were equally dissatisfied with their children's educational experiences, additional research could focus on ways for schools to improve in the areas of concern.

References

- Abramson, M., Willson, V., Yoshida, R.K., & Hagerty, G. (1983). Parents' perceptions of their learning disabled child's educational performance. Learning Disability Quarterly, 6(2), 184-194.
- Abramson, R.K., Wright, H.H., Cuccaro, M.L., & Lawrence, L.G. (1992). Biological liability in families with autism. Journal of the American Academy of Child & Adolescent Psychiatry, 31(2), 370-371.
- Achenbach, T.M. (1991). Manual for the Child Behavior Checklist/4-18 and 1991 Profile. Burlington, VT: University of Vermont, Department of Psychiatry.
- Achenbach, T.M. & Rescorla, L.A. (2001). Manual for the ASEBA School-Age Forms & Profiles. Burlington, VT: University of Vermont, Research Center for Children, Youth, & Families.
- Allen, S.J., & Graden, J.L. (1995). Best practices in collaborative problem solving for intervention design. In A. Thomas & J. Grimes (Eds.), Best Practices in School Psychology III (pp. 667-678). Washington, DC: National Association of School Psychologists.
- Arnold, K.D., Michael, M.G., Hosley, C.A., & Miller, S. (1994). Factors influencing attitudes about family-school communication for parents of children with mild learning problems: preliminary findings. Journal of Educational and Psychological Consultation, 5(3), 257-267.
- Baker, B.L., Blacher, J., Crnic, K.A., & Edelbrock, C. (2002). Behavior problems and parenting stress in families of three-year-old children with and without developmental delays. American Journal on Mental Retardation, 107 (6), 433-444.
- Baker, P., Piven, J., & Sato, Y. (1998). Autism and tuberous sclerosis complex: Prevalence and clinical features. Journal of Autism and Developmental Disorders, 28(4), 279-285
- Baltaxe, C.A.M. (1998). A systematic study of communication behaviors in children and adolescents with tuberous sclerosis: Progress report. Paper presented at the Tuberous Sclerosis Complex Consensus Conference, Annapolis, MD.

Baraitser, M., Paton, M.A. (1985). Reduced penetrance in tuberous sclerosis. Journal of Medical Genetics, 22, 29-31.

Bernstein, D.K. & Tiegerman, E. (1993). Language and communication disorders in children (3rd ed.). New York: Macmillan.

Boelte, S., Dickhut, H., & Poustka, F. (1999). Patterns of parent-reported problems indicative of autism. Psychopathology, 32 (2), 93-97.

Bourgeois, B.F.D., Prensky, A.L., Palkes, H.S., Talent, B.K., & Busch, S.G. (1983). Intelligence in epilepsy: A prospective study in children. Annals of Neurology, 14, 438-444.

Bourneville, D.M. (1880). Contribution a l'etude de l'idiote-sclerose tubereuse des dirconvolutions cerebrales: Idiotie et epilepsie hemiplegique. Archives de Neurologie, 1, 69-91, 670-671.

Brown, L.W. (1997). Seizure disorders. In M.L. Batshaw (Ed.), Children with disabilities, Fourth Edition (pp.553-594). Baltimore: Paul H. Brookes.

Cartledge, G., & Milburn, J.F. (1995). Teaching social skills to children and youth: Innovative approaches (3rd ed.). Boston: Allyn & Bacon.

Chalhub, E.G. (1976). Neurocutaneous syndromes in children. Pediatric Clinics of North America, 23, 499-516.

Chesapeake Institute (1994, September). National agenda for achieving better results for children and youth with serious emotional disturbance. Washington, DC: U.S. Department of Education.

Clements, J., Wing, L., & Dunn, G. (1986). Sleep problems in handicapped children: A preliminary study. Journal of Child Psychology and Psychiatry and Allied Disciplines, 27(3), 399-407.

Cohen, S.B. (1986). Parents' attributions of exceptionality: Social distancing effects in the mainstreamed classroom. RASE: Remedial & Special Education, 7(4), 48-53.

Critchley, M., & Earl, C.J.C. (1932). Tuberous sclerosis and allied conditions. Brain, 55, 311-346.

Crowell, A.R. (1993). Contrasting perspectives on programming for students with emotional disturbance and behavioral disorders in Mississippi. Behavioral Disorders, 18(3), 228-230.

Curatolo, P., Cusmai, R., Cortesis, F., Chiron, C., Jambaque, L., & Dulac, O. (1991). Neuropsychiatric aspects of tuberous sclerosis. Annals of the New York Academy of Sciences, 615, 8-16.

Dekker, M.C., Koot, H.M., van der Ende, J., Verhulst, F.C. (2002). Emotional and behavior problems in children and adolescents with and without intellectual disability. Journal of Child Psychology & Psychiatry & Allied Disciplines, 43 (8), 1087-1098.

DeLong, G.R. & Dwyer, J.T. (1988). Correlation of family history with specific autistic subgroups: Asperger's syndrome and bipolar affective disease. Journal of Autism & Developmental Disorders, 18(4), 593-600.

DiPietro, E., Luiselli, J.K., Campbell, S., Cannon, B.O., Ellis, J.T., Taras, M. (2002). A parent survey approach to evaluate public schools education of children with Autism/Pervasive Developmental Disorder following center-based behavioral treatment. Special Services in the Schools, 18 (1-2), 199-131.

Dodrill, C.B. (1992). Neuropsychological aspects of epilepsy. Psychiatry Clinics of North America, 15, 383-394.

Dreifuss, F.E. (1994). Partial seizures (focal and multifocal). In K. Swaiman (Ed.), Pediatric neurology (pp.509-530). St. Louis, MO: Mosby.

Eccles, J.S. & Harold, R.D. (1993). Parent-school involvement during the early adolescent years. Teachers College Record, 94, 568-587.

Ferguson, A.P., McKinlay, I.A., Hunt, A. (2002). Care of adolescents with severe learning disability from tuberous sclerosis. Developmental Medicine & Child Neurology, 44(4), 256-262.

Ferry, P.C., Banner, W., & Wolf, R.A. (1986). Seizure disorders in children. Philadelphia: J.B. Lippincott Company.

Finn, J.D. (1998). Class size and students at risk: What is known? What is next? Washington, DC: National Institute on the education of At-risk Students, Office of Educational Research and Improvement, U.S. Department of Education.

Foreman, P., & Arthur, M. (2002). Parental perspectives on educational programmes for students with high support needs. European Journal of Special Needs Education, 17(2), 175-184.

Gallagher, P.A., Floyd, J.H., Stafford, A.M., Taber, T.A., Brozovic, S.A., & Alberto, P.A. (2000). Inclusion of students with moderate or severe disabilities in educational and community settings: Perspectives from parents and siblings. Education & Training in Mental Retardation & Developmental Disabilities, 35(2), 135-147.

Gibson, P.A. (1993). Psychosocial interventions for adolescents with epilepsy. Epilepsia, 34, 31s.

Gillberg, C. (1991). The treatment of epilepsy in autism. Journal of Autism and Developmental Disorders, 21, 61-77.

Gillberg, C., & Coleman, M. (1992). The biology of autistic syndromes. 2nd Ed. Clinics in Developmental Medicine. No.126. London/New York: Mac Keith Press with Cambridge University Press.

Gillberg, I.C., Gillberg, C., & Ahlsen, G. (1994). Autistic behavior and attention deficits in tuberous sclerosis: A population based study. Developmental Medicine and Child Neurology, 36, 50-56

Gillberg, C. Steffenburg, S., & Shaumann, H. (1991). Is autism more common now than ten years ago? British Journal of Psychiatry, 158, 403-409.

Gimpel, G.A., & Huebner, E.S. (1994). Parental perceptions of school psychological services: One school district's survey method and findings. Special Services in the Schools, 9(1), 159-174.

Glass, G.V. & Smith, M.L. (1978). Meta-analysis of research on the relationship of class size and achievement. San Francisco: Far West Laboratory for Educational Research and Development.

Gomez, M.R. (1979). Clinical experience at the Mayo Clinic. In Gomez, M.R. (Ed.) Tuberous sclerosis. New York: Raven Press.

Gomez, M.R. (1988a). Tuberous sclerosis: Neurologic and psychiatric features (2nd ed.). New York: Raven Press.

Gomez, M. (1991). Phenotypes of the tuberous sclerosis complex with a revision of diagnostic criteria. In W.G. Johnson & M.R. Gomez (Eds.), Tuberous sclerosis and allied disorders. New York: New York Academy of Sciences.

Graden, J.L., Zins, J.E., Curtis, M.J., & Cobb, C.T. (1989). The need for alternatives in educational services. In J.L. Graden, J.E. Zins, and M.J. Curtis (Eds.), Alternative educational delivery systems: Enhancing instructional options for all children. Washington, DC: National Association of School Psychologists.

Grant, V. (1989). Tuberous sclerosis (Bourneville's disease). Journal of Ophthalmic Nursing and Technology, 8, 143-147.

Green, S., & Shinn, M. (1995). Parent attitudes about special education and reintegration: What is the role of student outcomes? Exceptional Children, 61(3), 269-281.

Griffith, J. (1996). Test of a model of the organizational antecedents of parent involvement and satisfaction with public education. Human Relations, 49 (12), 1549-1472.

Gutierrez, G.C., Smalley, S.L., & Tanguay, P.E. (1998). Autism in tuberous sclerosis complex. Journal of Autism & Developmental Disorders, 28(2), 97-103.

Henderson, A.T. (1981). Parent participation-student achievement: The evidence grows. NCCE Occasional Papers. (Educational Research Information Center No ED 209 754)

Henderson, A.T. (1987). The evidence continues to grow: Parent involvement improves student achievement. An annotated bibliography. National Committee for Citizens in Education Special Report. (Educational Research Information Center No ED 315 199)

Howe, N., Chambers, B., & Abrami, P.C. (1998). The effects of an academic restructuring program on parental attitudes and behaviors. Alberta Journal of Educational Research, 44(1), 106-110.

Hunt, A. (1983). Tuberous sclerosis: A survey of 97 cases. Developmental Medicine and Child Neurology, 25, 346-349.

Hunt, A. (1993). Development, behaviour and seizures in 300 cases of tuberous sclerosis. Journal of Intellectual Disability Research, 37(1), 41-51.

Hunt, A. (1995). Gaining new understanding of tuberous sclerosis. Nursing Times, 91(33), 31-33.

Hunt, A. (1998). A comparison of the abilities, health and behaviour of 23 people with tuberous sclerosis at Age 5 and as adults. Journal of Applied Research in Intellectual Disabilities, 11(3), 227-238.

Hunt, A. (1999). Psychiatric and Psychological Aspects. Tuberous Sclerosis Complex: Developmental Perspectives in Psychiatry (M.R. Gomez, J.R. Sampson, and V.H. Whittemore, Eds.)

Hunt, A., & Dennis, J. (1987). Psychiatric disorder among children with tuberous sclerosis. Developmental Medicine and Child Neurology, 25, 350-352.

Hunt, A. & Lindenbaum, R. (1984). Tuberous sclerosis: A new estimate of prevalence within the Oxford region. Journal of Medical Genetics, 21, 272-277.

Hunt, A., & Shepherd, C. (1993). A prevalence study of autism in tuberous sclerosis. Journal of Autism and Developmental Disorders, 23, 323-339.

Hunt, A., & Stores, G. (1994). Sleep disorder and epilepsy in children with tuberous sclerosis: A questionnaire-based study. Developmental Medicine & Child Neurology, 36(2), 108-115.

Hynd, G.W., & Willis, W.G. (1988). Pediatric neuropsychology. Orlando, FL: Grune & Stratton.

Jackson, J.H. (1990). School psychology after the 1980s: Envisioning a possible future. In T.B. Gutkin and C.R. Reynolds (Eds.), The handbook of school psychology (2nd edition). New York: John Wiley & Sons.

Jambaque, I., Cusmai, R., Curatolo, P., Cortesi, F., Perrot, C., & Dulac, O. (1991). Neuropsychological aspects of tuberous sclerosis in relation to epilepsy and MRI findings. Developmental Medicine and Child Neurology, 33, 698-705.

Joinson, C., O'Callaghan, F.J., Osborne, J.P., Martyn, C., Harris, T., & Bolton, P.F. (2003). Learning disability and epilepsy in an epidemiological sample of individuals with tuberous sclerosis complex. Psychological Medicine, 33 (2), 335-344.

Jozwiak, S., Goodman, M., & Lamm, S.H. (1998). Poor mental development in patients with tuberous sclerosis complex: Clinical risk factors. Archives of Neurology, 55, 379-384.

Kaufman, J.M. (1993b). Characteristics of emotional and behavioral disorders of children and youth (5th ed.). Englewood Cliffs, NJ: Merrill/Prentice Hall.

Kazdin, A. (1993). Treatment of conduct disorder: Progress and directions in psychotherapy research. Development and Psychopathology, 5 (1/2), 277-310.

Krug, D.A., Arick, J., & Almond, P. (1980a). Behavior checklist for identifying severely handicapped individuals with high levels of autistic behavior. Journal of Child Psychology and Psychiatry, 21, 221-229.

Krug, D.A., Arick, J., & Almond, P. (1980b). Autism screening instrument for educational planning. Portland, OR: ASIEP.

Lail, J.L. & Schroeder, C.S. (1990). Health-related problems in school-aged children. In T.B. Gutkin and C.R. Reynolds (Eds.), The handbook of school psychology (2nd edition). New York: John Wiley & Sons.

Lane, V., Samples, J. (1984). Tuberous sclerosis. Case study of early seizure control and subsequent normal development. Journal of Autism and Developmental Disorders, 14, 423-427.

Le Couteur, A., Rutter, M., Lord, C., Rios, Pl, Robertson, S., Holdgrafer, M., & McLennan, J., (1989). Autism diagnostic interview: A standardized investigator-based instrument. Journal of Autism and Developmental Disorders, 19, 363-386.

Leyser, Y. (1988). Let's listen to the consumer: The voice of parents of exceptional children. Schol Counselor , 35(5), 363-369.

Lotter, V. (1966). Epidemiology of autistic conditions in young children: I. Prevalence. Social Psychiatry, 1, 124-137.

Lowenbraun, S., Madge, S., & Affleck, J. (1990). Parental satisfaction with integrated class placements of special education and general education students. RASE: Remedial & Special Education, 11(4), 37-40.

Lynch, E.W. & Stein, R. (1982). Perspectives on parent participation in special education. Exceptional Education Quarterly, 3(2), 56-63.

Male, D.B. (1998). Parents' views about special provision for their child with severe or profound and multiple learning difficulties. Journal of Applied Research in Intellectual Disabilities, 11(2), 129-145.

McWilliam, R.A., Maxwell, K.L., & Sloper, K.M. (1999). Beyond "involvement": Are elementary schools ready to be family centered? School Psychology Review, 28(3), 378-394.

Medcalf, J. (1991). Tuberous sclerosis and its management. Nursing Standard, 5(30), 28-30.

Miller, V.S., & Bigler, E.D. (1982). Neuropsychological aspects of tuberous sclerosis. Clinical Neuropsychology, 4, 26-34.

Mlynek, S., Hannah, M.E., & Hamlin, M.A. (1982). Mainstreaming: Parental perceptions. Psychology in the Schools, 19(3), 354-359.

National Household Education Survey of 1999 [Electronic data file]. (2001). Washington, DC: National Center for Educational Statistics (<http://nces.ed.gov/pubs200/2000076.pdf>).

Nolan, E.E., Gadow, K.D., Sprafkin, J. (2001). Teacher reports of DSM-IV ADHD, ODD, and CD symptoms in schoolchildren. Journal of the American Academy of Child & Adolescent Psychiatry, 40(2), 241-249.

O'Callaghan, F., Shiell, A., Osborne, J.P., & Martyn, C.N. (1998). Prevalence of tuberous sclerosis estimated by capture-recapture analysis. The Lancet, 351(9114), p.1490-1491.

Osborne, J.P., Fryer, A., & Webb, D. (1991). Epidemiology of tuberous sclerosis. In W.G. Johnson & M.R. Gomez (Eds.), Tuberous sclerosis and allied disorders. New York: New York Academy of Sciences.

Piven, J., Chase, G.A., Landa, R., & Wzorek, M. (1991). Psychiatric disorders in the parents of autistic individuals. Journal of the American Academy of Child & Adolescent Psychiatry, 30(3), 471-478.

Piven, J., Gayle, J., Chase, G.A., & Fink, B. (1990). A family history study of neuropsychiatric disorders in the adult siblings of autistic individuals. Journal of the American Academy of Child & Adolescent Psychiatry, 29(2), 177-183.

Plunge, M.M., & Kratochwill, T.R. (1995). Parental knowledge, involvement and satisfaction with their child's special education services. Special Services in the Schools, 10(1), 113-138.

Public Law 94-142. (November 29, 1975). Education for All Handicapped Children Act.

Reber, M. & Borcharding, B.G. (1997). Dual diagnosis: Mental retardation and psychiatric disorders. In M.L. Batshaw (Ed.), Children with disabilities, Fourth Edition (pp.405-424). Baltimore: Paul H. Brookes.

Riccio, C.A., & Harrison, P.L. (1998). Tuberous sclerosis. In L. Phelps (Ed.), Health related disorders in children and adolescents (pp. 683-690). Washington, DC: American Psychological Association.

Riikonen, R., & Amnell, G. (1981). Psychiatric disorder in children with earlier infantile spasms. Developmental Medicine and Child Neurology, 23, 747-760.

Riikonen, R., & Simell, O. (1990). Tuberous sclerosis and infantile spasms. Developmental Medicine and Child Neurology, 32, 203-209.

Ritvo, E.R., Mason-Brothers, A., Freeman, B.J., Pingree, C., Jenson, W.R., McMahon, W.M., Petersen, P.B., Jorde, L.B., Mo, A., & Ritvo, A. (1990). UCLA-University of Utah Epidemiological survey of autism: Etiologic role of rare diseases. American Journal of Psychiatry, 147, 1614-1621.

Rivers, E. (1990). The relationship between cultural background and parental perceptions of functional educational programming for students with severe handicaps. (ERIC Document Reproduction Service No. ED 330 182)

Roach, E.S. (1992). Neurocutaneous syndromes. Pediatric Clinics of North America, 39, 591-620.

Roach, E.S., Smith, M., & Huttenlocher, P. (1992). Diagnostic criteria: Tuberous sclerosis complex: Report of the Diagnostic Criteria Committee of the National Tuberous Sclerosis Association. Journal of Child Neurology, 7, 221-224.

Rutter, M. (1985). The treatment of autistic children. Journal of Child Psychology and Psychiatry, 26, 193-214.

Sachs, H.T. & Barrett, R.P. (1995). Seizure disorders: A review for school psychologists. School Psychology Review, 24, 131-145.

Sampson, J.R., Scahill, S.J., Stephenson, J.B.P., Mann, L., & Connor, J.M. (1989). Genetic aspects of tuberous sclerosis in the west of Scotland. Journal of Medical Genetics, 26, 28-31.

Schopler, E., Reichler, R.J., & Renner, B.R. (1988). The childhood autism rating scales (CARS)-Revised. Los Angeles: Western Psychological Services.

Seidenberg, M., O'Leary, D.S., Berent, S., & Boll, T. (1981). Changes in seizure frequency and test-retest scores on the Wechsler Adult Intelligence Scale. Epilepsia, 22, 75-83.

Shepherd, C.W., Beard, M., Gomez, M.R., Kurland, L.T., Whisnant, J.P. (1991). Tuberous sclerosis complex in Olmsted County, Minnesota, 1950-1989. Archives of Neurology, 48, 400-401.

Shepherd, C.W., & Hosking, G. (1989). Epilepsy in schoolchildren with intellectual impairments in Sheffield. Journal of Mental Deficiency Research, 33, 511-514.

Shepherd, C.W., & Stephenson, J.B.P. (1992). Seizures and intellectual disability associated with tuberous sclerosis complex in the west of Scotland. Developmental Medicine and Child Neurology, 34, 766-744.

Smalley, S.L., Burger, F., & Smith, M. (1994). Phenotypic variation of tuberous sclerosis in a single extended kindred. Journal of Medical Genetics, 31, 761-765.

Smalley, S.L., Tanguay, P.E., Smith, M., & Gutierrez, G. (1992). Autism and tuberous sclerosis. Journal of Autism and Developmental Disorders, 22, 339-355.

Soodak, L.C. (1998). Parents and inclusive schooling: Advocating for and participating in the reform of special education. In S.J. Vitello & D.E. Mithaug (Eds.), Inclusive schooling: National and international perspectives (pp. 113-131). Mahwah, NJ: Lawrence Erlbaum.

Starr, E.M., Foy, J.B., & Cramer, K.M. (2001). Parental perceptions of the education of children with Pervasive Developmental Disorders. Education and Training in Mental Retardation and Developmental Disabilities, 36 (1), 55-68.

Stevenson, A.C., Fischer, O.D. (1956). Frequency of epiloia in Norther Ireland. British Journal of Preventive Social Medicine (Br J Prev Soc Med), 10, 134-135.

Taylor, E.A. (Ed.) (1986). The overactive child: Clinics in developmental medicine no. 95. London: S.I.M.P. with Blackwell Scientific; Philadelphia: Lippincott.

Teeter, P.A. & Semrud-Clikeman, M. (1998). Seizure disorders. In L. Phelps (Ed.), Health-related disorders in children and adolescents (pp. 583-595). Washington, DC: American Psychological Association.

Thompson, P. (1995). The impact of epilepsy on behavior and emotional development. In A.P. Aldenkamp, F. Dreifuss, W. Renier, & T.P.B.M. Suumeijer (Eds.), Epilepsy in children and adolescents (pp.239-250). Boca Raton, FL: CRC Press.

U.S. Department of Education, (1994). Strong families, strong schools: Building community partnerships for learning. Washington, DC: Author.

Valdes, K.A., Williamson, C.L., & Wagner, M. (1990). The National Longitudinal Transition Study of special education students Vol. 3: Youth categorized as emotionally disturbed. Palo Alto, CA: SRI International.

Wacschlag, L.S., Cook, E.H., Hammond, D.N., Leventhal, B.L., & Hopkins, J. (1991). Autism and tuberous sclerosis. Journal of Autism and Developmental Disorders, 21, 95-97.

Webb, D.W., Fryer, A.E., & Osborne, J.P. (1996). Morbidity associated with tuberous sclerosis: A population study. Developmental Medicine and Child Neurology, 38, 146-155.

Webb, D.W., Osborne, J.P., & Fryer, A.E. (1991). On the incidence of fits and mental retardation in tuberous sclerosis. Journal of Medical Genetics, 28, 385-388.

Webb, D.W., & Osborne, J.P. (1995). Tuberous sclerosis. Archives of Disease in Childhood, 72, 471-474.

Westling, D.L. (1997). What parents of young children with mental disabilities want: The views of one community. Focus on Autism & Other Developmental Disabilities, 12(2), 67-78.

Whittemore, V.H. & Roach, S. (1999). Clinical manifestations of tuberous sclerosis. [On-line]. Available: <http://www.nts.org/index.pl/research/physicia2>

Wing, L., & Gould, J. (1979). Severe impairments of social interaction and associated abnormalities in children: Epidemiology and classification. Journal of Autism and Developmental Disorders, 9, 11-29.

Wolf, P. (1985). The classification of seizures and the epilepsies. In R.J. Porter & P.L. Morselli, (Eds.) The epilepsies. London: Butterworths.

Yamamoto, N., Watanabe, K., Negoro, T., Matsumoto, A., Miyazaki, S., Kumagai, I.T., & Iwase, K. (1987). Long term prognosis of tuberous sclerosis with epilepsy in children. Brain and Development, 9, 292-295

Appendices

Appendix A: Parent Questionnaire

Parent Questionnaire

Please answer the following questions about your child with tuberous sclerosis:

Note: If you have more than one child with tuberous sclerosis, please complete this form for the child whose first name comes first in the alphabet.

I. Demographic Information

1. Today's Date: ____ / ____ / ____ (month/day/year)
2. Child's Date of Birth: ____ / ____ / ____ (month/day/year)
3. Child's gender: (circle one) M F
4. Child's age at diagnosis of tuberous sclerosis: (years-months) ____ - ____
5. Child's race/ethnicity:
 - ____ White, non-hispanic
 - ____ Black/African-American
 - ____ Hispanic
 - ____ Asian/Pacific Islander
 - ____ Native American
 - ____ Other: _____
6. Has your child been diagnosed with any of the following disorders by a physician, psychiatrist, or psychologist?
 - ____ Anxiety
 - ____ Depression
 - ____ Autism
 - ____ Attention-Deficit/Hyperactivity Disorder (ADHD)
 - ____ Obsessive-Compulsive Disorder (OCD)
 - ____ Conduct Disorder/ Oppositional Defiant Disorder (CD or ODD)
 - ____ Seizure Disorder
7. School your child attends:
 - ____ Regular Public School
 - ____ Alternative school, paid for by school district
 - ____ Regular Private School
 - ____ Alternative school, paid for by parents
 - ____ Other (please specify) _____
8. Grade in school: (K through 12) ____
9. What is your child's approximate level of cognitive functioning (in school)?
 - ____ Above Average (IQ score higher than 115)
 - ____ Average (IQ score of 85 - 115)
 - ____ Low Average (IQ score of 70 - 84)
 - ____ Low (IQ score below 70)
 - ____ Don't Know

II. Services Received

10. What is your child's PRIMARY category of special education in school?

(check only ONE)

- Receiving NO special education services (Skip to Question 12)
- Learning Disabilities (LD)
- Mild (educably) Mental handicaps or disabilities
- Moderately or Severely (trainably/profoundly) mentally handicapped
- Behavior Disorder / Emotional Disorder
- Autism
- Other Health Impairment
- Orthopedic Impairment
- Other (please specify) _____

11. Approximately how much of the school day does your child spend in:

a) a special education or resource classroom? (circle response)

- NONE
- LESS THAN HALF
- ABOUT HALF
- MORE THAN HALF
- ALL
- DON'T KNOW

b) a regular (general education) classroom with special education support? (For example, with an "inclusion" or "mainstreaming" teacher in the room, or with an aide for your child.) (circle response)

- NONE
- LESS THAN HALF
- ABOUT HALF
- MORE THAN HALF
- ALL
- DON'T KNOW

c) A general education classroom with no extra support? (circle response)

- NONE
- LESS THAN HALF
- ABOUT HALF
- MORE THAN HALF
- ALL
- DON'T KNOW

12. Check all Related Services your child receives:

THROUGH THE SCHOOL

(check all that apply)

- Speech/Language Therapy
- Physical Therapy
- Occupational Therapy
- Individual counseling
- Group counseling
- Other (please specify) _____
- Don't Know
- None

PRIVATELY

(check all that apply)

- Speech/Language Therapy
- Physical Therapy
- Occupational Therapy
- Individual counseling
- Group counseling
- Other (please specify) _____
- Don't Know
- None

III. Parent Involvement and Satisfaction

13. In an average school year, how often do you communicate with your child's Teacher(s)?

Regular education teacher(s)?

Never Daily Weekly Monthly 1-2 times a semester

Special education teacher(s)?

Never Daily Weekly Monthly 1-2 times a semester

Related service providers? (Example: Speech therapists, occupational therapists, etc.)

Never Daily Weekly Monthly 1-2 times a semester

14. In an average school year, how many times does your child's Teacher(s) call you on the phone to tell you about your child's progress?

Regular education teacher(s)?

Never Daily Weekly Monthly 1-2 times a semester

Special education teacher(s)?

Never Daily Weekly Monthly 1-2 times a semester

15. In an average school year, how many times does your child's Teacher(s) call you on the phone to tell you about your child's problems?

Regular education teacher(s)?

Never Daily Weekly Monthly 1-2 times a semester

Special education teacher(s)?

Never Daily Weekly Monthly 1-2 times a semester

16. In an average school year, how many times do you meet informally (other than the IEP meeting) and sit down with your child's:

Regular education teacher(s)?

Never 1 - 2 times 3 - 4 times 5 - 6 times More than 6 times

Special education teacher(s)?

Never 1 - 2 times 3 - 4 times 5 - 6 times More than 6 times

17. In an average school year, how many times does your child's Teacher(s) and/or school staff invite or encourage you to visit your child's program?

Regular education teacher(s)?

Never 1 - 2 times 3 - 4 times 5 - 6 times More than 6 times

Special education teacher(s)?

Never 1 - 2 times 3 - 4 times 5 - 6 times More than 6 times

18. Would you say that you are very satisfied, somewhat satisfied, somewhat dissatisfied, or very dissatisfied with your child's school's...

[1 = VERY SATISFIED; 2 = SOMEWHAT SATISFIED; 3 = SOMEWHAT DISSATISFIED; 4 = VERY DISSATISFIED]

	VS	SS	SD	VD
a. Instructional program? (instruction received in school)	1	2	3	4
b. Special Education program? (the program designed for your child's specific needs)	1	2	3	4
c. Related services? (speech therapy, counseling, etc.)	1	2	3	4
d. Communication? (with teachers, administrators, and staff)	1	2	3	4
e. Discipline? (the classroom management and discipline procedures)	1	2	3	4
f. Social Skills? (your child's social progress and behavior)	1	2	3	4

19. What changes would you like to see made in these areas? Be as specific as possible.

Appendix B: Parent Letter

Date

Dear Parent,

I am conducting a study on the school factors that increase the success of students with Tuberous Sclerosis, in conjunction with the TS Alliance and the University of South Florida. As a parent of a child with TS, you can provide valuable insight that can change the way TS is viewed and treated by educators. It is my hope that this information will help parents and teachers make educational decisions to improve the social, behavioral, and academic functioning of students with TS.

If you have a child with Tuberous Sclerosis between the ages of 6 and 17 years old, please complete the enclosed questionnaire. Participation is voluntary. If you choose to participate, you will be asked to provide some basic information about your child's education and to complete a rating scale about your child's behavior. It is estimated that the total time needed to complete these questionnaires is 15 minutes. If you choose not to participate, or do not have a child with TS between the ages of 6 and 17, please return the questionnaires in the enclosed self-addressed, stamped envelope.

There are no risks to participants. Participation is voluntary, and all of the information you provide is completely confidential. The information you provide will not be given out or used for any other purpose. The long-term benefits of this study include a better understanding of factors that influence and support the social and behavioral success of children with TS in school.

I sincerely hope that you will assist in this important research opportunity.
Thank you for your help.

Kathleen Carlisle, Principle Researcher
Licensed Specialist in School Psychology
Doctoral Candidate
University of South Florida

For questions, comments, or concerns, please contact me, Kathleen Carlisle, at:
(phone #).
To contact the TSAlliance regarding this research, please call:
Holly Knorr, title, phone #

Appendix C: Correlation Matrix for Research Questions 8, 9, and 10

Variable	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15
1. Parent Involvement	1	.52	--	.26	-.17	-.27	.16	-.08	-.08	-.30	.46	.04	.47	.19	.04
2. Parent Satisfaction		1	.02	.14	-.32	-.12	.21	-.24	-.18	.04	.24	.01	.14	.07	.05
3. No Special Education			1	-.09	-.14	-.14	-.30	-.34	.17	-.21	-.08	-.30	.14	.42	-.75
4. Learning Disability				1	-.13	-.13	-.28	-.07	-.18	-.14	.29	-.14	.12	.08	.02
5. Autism					1	-.19	-.41	.20	-.06	.16	-.11	.32	.08	-.14	.20
6. Other Health Impairment						1	-.41	-.05	.20	-.12	-.11	-.04	-.12	.52	-.26
7. Mental Retardation							1	.21	-.20	.23	.07	-.04	-.14	-.56	.47
8. Total Behavior Problems								1	-.03	.09	-.01	.36	.15	-.49	.36
9. Regular Public School									1	-.82	-.34	.04	-.07	.32	-.27
10. Alternative Public School										1	-.16	.03	-.11	-.36	.28
11. Private School											1	-.03	.30	.06	.12
12. Public Related Services												1	.21	-.29	.3
13. Private Related Services													1	.15	-.03
14. IQ														1	-.74
15. Time in Special Education															1

Appendix D: Correlation Matrix for Research Question 11

Variable	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15
1. CBCL-Anxiety	1	.36	.61	.30	.34	.12	.07	.33	.06	.09	.33	-.31	-.13	-.15	.18
2. CBCL-Withdrawn/Dep.		1	.31	.43	.34	.23	.11	.35	.07	.03	-.04	.12	-.17	-.09	-.06
3. CBCL-Somatic Complaints			1	.44	.37	.08	-.01	.31	.17	.24	.14	-.19	.19	-.05	-.06
4. CBCL-Social Problems				1	.63	.59	.46	.72	.29	.15	-.09	.06	.14	.31	-.41
5. CBCL-Thought Problems					1	.58	.41	.70	.33	.13	-.01	.10	-.06	.35	-.58
6. CBCL-Attention Problems						1	.53	.69	.34	.03	-.04	.09	.01	.43	-.55
7. CBCL-Rule-Breaking							1	.67	.23	-.03	.05	.02	-.07	.18	-.34
8. CBCL-Aggression								1	.32	.13	.05	.02	-.03	.33	-.49
9. Public Related Services									1	.21	.04	.03	-.03	.38	-.29
10. Private Related Services										1	-.07	-.11	.30	-.03	.15
11. Regular Public School											1	-.82	-.34	-.27	.32
12. Alternative Public School												1	-.16	.28	-.37
13. Private School													1	.12	.06
14. Time in Special Education														1	-.74
15. IQ															1

Appendix E: Correlation Matrix for Research Question 12

Variable	1	2	3	4	5	6	7	8	9	10	11	12
1. Age	1	-.44	-.17	.12	-.28	-.08	.01	.04	-.19	-.03	.26	-.12
2. Public Related Services		1	.21	.33	.02	.38	-.30	-.14	.32	-.04	-.04	-.29
3. Private Related Services			1	-.13	.13	-.03	.14	.12	.08	-.12	-.14	.15
4. Time in Special Ed. Class				1	-.50	.69	-.53	-.12	.12	-.25	.50	-.84
5. Time in Inclusion Class					1	.28	-.19	.18	.08	.03	-.10	.21
6. Time in Special Education						1	-.75	.02	.20	-.26	.47	-.74
7. No Special Education							1	-.09	-.14	-.14	-.30	.42
8. Learning Disability								1	-.13	-.13	-.28	.08
9. Autism									1	-.19	-.41	-.14
10. Other Health Impairment										1	-.41	.52
11. Mental Retardation											1	-.56
12. IQ												1

About the Author

Kathleen Walker Carlisle completed her undergraduate degree in psychology at Houghton College in western New York, and received her Masters and Doctoral degrees from the University of South Florida in Tampa, where she was a recipient of the Graduate Fellowship Award two years in a row. Kathleen has been a presenter at the Florida Association of School Psychologists and the National Family Conference for the Tuberos Sclerosis Alliance. She has also completed a number of writing projects, including the Learning Disabilities and Child Study Team chapters in the school handbook for the organization *National Consultants for Education*. Kathleen has lived and worked as a school psychologist in Texas, Virginia Beach, and the Washington DC area. She and her husband currently reside in Texas, awaiting the birth of their first child.