

ADOLESCENTS WITH TUBEROUS SCLEROSIS COMPLEX

Below is an overview of the "normal" developmental tasks of adolescence while concentrating on the particular health care needs and challenges as faced by the teen with tuberous sclerosis complex (TSC). There are many excellent books on parenting a teenager as well as raising a handicapped child. Please refer to additional Fact Sheets that have outlined the specific issues related to the child and adult with TSC and concurrent attention difficulties, autism, mental retardation and other psychiatric and behavioral difficulties.

Adolescent years are marked by changes in almost every aspect of the child's life. Adolescents are developing a strong sense of their identity and discovering their personal values, adjusting to dramatic body changes, developing abstract thought, acquiring interpersonal skills, establishing independence from their family while negotiating a new relationship with their family, setting goals for future achievement and choosing a vocation. It is important to recognize some of the normal behaviors and reactions of parents and teens during this period. Many parents may perceive certain teen attitudes and behaviors to be problems. In reality those attitudes and behaviors may reflect very typical adolescent patterns totally aside from the problems of an adolescent with a chronic illness such as TSC.

WHAT ARE NORMAL ADOLESCENT CHANGES?

Early adolescence begins with the teen asking themselves, "Am I normal?" and "Will I be okay?" Teens look to their parents and peers for solving these questions and for affirmation that they are okay.

Mid-adolescence is characterized by the teen asking more in-depth questions like "Who am I?", "Am I accepted?" and "Do I have power to make decisions about myself?" Peers become more important in helping to answer those questions, while parents may have less of an influence.

In late adolescence, the teen may ask, "Am I smart enough?", "Am I attractive?" and "What should I do with my life?" The peer group becomes extremely important in influencing behaviors, attitudes, dress and values.

For teens with a chronic lifelong illness, the developmental problems normally associated with adolescence seem magnified. The overwhelming concerns relate to social acceptance. Peer acceptance is extremely important to a teen, and a teen's perception of his or her physical appearance often determines psychological adjustment. Adolescence is a time of significant changes. Achieving independence from family, formulating values and self-concept, and planning for the future become important objectives.

How moody should an adolescent be? How talkative, rebellious, oppositional or resistant? What is normal teenage behavior? Why does my teen appear to be so angry all the time? These questions are difficult to answer specifically. In general, normal behavior will not interfere with a person's ability to cope with his/her environment or to get along with others.

But how do you determine what is a normal amount of moodiness in your teenager with TSC? Other Fact Sheets outline the specific issues that TSC may have in influencing the child with learning, hyperactive or impulsive behaviors, speech and communication. Many adults with TSC have reported feelings of anxiety, depression and anger. (See TSC Fact Sheet "Psychiatric and Behavioral Issues in Adults with Tuberous Sclerosis Complex.")

Some questions to ask yourself regarding your teen's behaviors may include:

1. How often does the behavior occur?
2. Does the behavior interfere with the teen's ability to function in the environment?
3. Does the behavior interfere with others, such as siblings or classmates, teachers, coaches, neighbors and others whom they may have daily contacts with?
4. Have you considered your teen's individual differences? How different is the behavior or attitude when compared with other children in their age group?

SPECIAL PROBLEMS THAT MAY OCCUR

Epilepsy

An estimated 80 percent to 90 percent of all patients with TSC experience a first seizure with approximately 27 percent occurring from birth to age 16. The kind of seizures that the child with TSC has been experiencing in childhood may change as

adolescence approaches. These changes may not indicate any significant problem but simply mean the child's nervous system is growing and developing into a mature adult's. Some adolescents may not experience a seizure until their teen years and may have sudden or unusual alterations in their behavior.

It is very important to clarify the episodic event with careful observation and documentation. Complex partial seizures may be presented as unusual changes in behavior and teens may experience a variety of "somatosensory" symptoms (unusual sensations) associated with these seizures. Complex partial seizures are those seizures that involve only part of the brain (usually the temporal or the frontal lobes), which alter consciousness or awareness. It may be accompanied by automatisms-movements that often consist of smacking of the lips, chewing, picking at clothes, or wandering about in a confused fashion. Teens may have difficulty in finding the correct word to say, or they may know the correct word yet are unable to say it, or say it clearly. They may have difficulty in comprehension of words or phrases.

Teens experiencing complex partial seizures may describe a variety of unusual sensations of the abdomen (cramps or nausea) or body sensations such as a certain smell, usually an unpleasant, strong odor such as the smell of burning rubber. Some seizures may begin with the sensation of fear, followed by changes in blood pressure, heart rate and the color of their skin (either paleness or facial flushing). Teens may describe flashbacks or strong memories or feelings, a déjà vu. Complex partial seizures may create or recreate one, all or any combination of these feelings or experiences.

Some questions to ask yourself regarding these episodes may include:

1. In what setting did the event occur?
2. What was the description of the event from beginning to end?
3. What happened first, second and third?
4. What kinds of movements were noticed? Was only one side of the body affected, or did the movements go to the other side as well?
5. How long did the event last? Were you able to time it?
6. During the episode, did the teen seem to lose the ability to communicate with you or understand you?
7. After the episode, was the return to their normal behavior immediate or seem delayed?
8. If the return to normal or "baseline" behavior was delayed, was the teen able to speak or function normally?
9. Was the speech pattern normal?

Between 10 percent to 20 percent of all children in the United States are affected by chronic illness, mostly due to a genetic disorder. For the adolescent with a chronic disease such as TSC and epilepsy, it can be more complex. The goal for our teens as they grow older will be for them to become responsible for more of their own care, while learning independence and responsibility for their health. Adolescence is the time when teens should begin the transition from childhood into independent adulthood.

The transition is a gradual process beginning in childhood so that by adolescence they will have the skills needed to care for their epilepsy and other health conditions associated with TSC. This may mean that they know the names of the medications they are taking, the doses and the times of administration. They should be responsible for keeping track of their seizure frequency if they have recurrent seizures. Teenaged patients should be fully informed about their epilepsy and should be allowed to make their own decisions, within the limits of reason and experience. Allowing a teen to make a choice may mean the teenager has the right to make choices about medication and therapeutic recommendations. At some point, the teen may refuse or avoid taking their medication on a regular basis. This may be expected and should be dealt with as a matter of fact, emphasizing the risk of seizures. All parents worry that something bad may happen to their child, but allowing your child the opportunities to take risks and become responsible for their own health needs ultimately will help the adolescent to achieve good emotional and social development.

Teenagers with epilepsy and TSC must have education about their condition and be advised about the possible implications of epilepsy and their treatments on adult issues, such as birth control and family planning. Teenaged girls must be aware of the effect of anticonvulsants on the unborn baby as well as the genetic risk involved with future pregnancy. Genetic counseling should be encouraged for all adolescents affected with TSC. Most adolescents want to be given specific information about their reproductive potential, and particularly the probability of passing on TSC to the next generation. Anxiety regarding the uncertainty of these life long decisions will often surface.

Adolescents with epilepsy may be at greater risk for depression and suicidal thoughts than their peers. The possibility of pregnancy in adolescence has major implications for the teenager with TSC. Young women should understand the interactions between anticonvulsants and contraceptive pills. Many anti-seizure medications change the effectiveness of birth control pills and these teens may need higher doses or stronger birth control. Young women with TSC must be informed about the potentially adverse effects of epilepsy and seizure medication on the developing fetus. It is always wise to consider questions they ask about making any changes in medication or reducing medication before conceiving a baby. In other words, plan ahead! If there were a concern that the adolescent female would become pregnant, Folate (1mg daily) is usually recommended for prevention of neural tube defects (spina bifida).

Teens need to understand the potentially adverse effects of alcohol and substance abuse on epilepsy and the potential they have in making their seizures worse. Seizures may increase in frequency or change during the rapid growth that occurs during adolescence. Seizure frequency, quality or duration of seizures may change as a reflection of changes in the size of subependymal nodules. These lesions rapidly grow between birth and 18 years of age. Girls may notice that their seizures increase prior to, or during their menstrual cycles. Sleep or lack thereof is already a documented problem in those with TSC and may also interfere with seizure

control. Promotion of good health habits, encouraging proper rest and exercise, as well as good nutrition are essential to any plan for managing epilepsy during the busy and active schedule that teenagers keep.

Compliance with medical issues and recommended therapies in adolescents with epilepsy is often a difficult issue. A recent Finnish study of 300 adolescents with epilepsy ages 13-17 revealed that 22 percent of adolescents with epilepsy followed suggested health regimens, while 44 percent placed themselves in the category of 'satisfactory compliance', and the remaining 34 percent reported poor compliance. Compliance with their recommended lifestyle was poorest, while the highest degree of compliance was recorded for medication. Compliance is especially important if the teen is driving a motor vehicle. Sometimes their only motivation to be compliant with medication hinges upon obtaining a driver's license. It is often helpful to gently remind young teens that driving will not be possible until their seizures are controlled for a specific length of time and that there is assurance that medication is taken as prescribed on a regular basis.

Kidney Problems

Because there are so many important changes to other body systems during adolescence, the teen should be educated regarding other manifestations of TSC and potential changes that would warrant seeking medical treatment. Other than central nervous system involvement, the kidneys are the next organ system most frequently affected. Renal (kidney) cysts often represent the earliest manifestation of TSC, often developed in infancy. If there is known renal involvement such as kidney cysts in association with TSC, teens are monitored for the development of hypertension (high blood pressure) that often precedes the development of more functional impairment of the kidneys. Controlling high blood pressure is very important in the care of teens with severe cystic involvement of the kidneys, as well as routine monitoring of kidney function by laboratory or diagnostic testing. Compliance with anti-hypertensive medications is very important.

Angiomyolipomas (AMLs) is the most common kidney lesion in patients with TSC. AMLs appear somewhat later in life (young adulthood) and generally do not cause any symptoms unless there is sudden hemorrhaging, which may lead to a potentially life-threatening situation. If the teen should experience blood in the urine or dull pain in the flank area, lower back or abdominal area, this may represent internal bleeding and may indicate a change that would need immediate medical attention. Teens with TSC should have their kidneys imaged on a regular basis every 1-2 years. The renal lesions may remain stable for long periods of time and may not require any specific treatment. Rarely, renal cell carcinoma or cancer in the kidney is seen. The tumors should be removed as soon as possible.

Skin Problems

Skin manifestations are an important part of TSC and are reported to occur in more than 80 percent of children over 5 years of age. Because body image is so important to a developing teenager, sensitivity to addressing their concerns regarding these facial features and appropriate treatments is critical. As a child enters puberty, the typical skin lesions may become even more apparent. The facial angiofibromas can be removed using dermabrasion or other laser techniques. Research has suggested that the facial angiofibromas should be removed using laser treatment when they are small and less fibrous.

LETTING GO

Of all the challenges facing the parent of a child with a disability or chronic illness, it is both most difficult and essential to encourage independence and making the transition to adulthood. While the grief of a parent or sense of parental responsibility cannot be underscored, it is vital that you encourage, support and help motivate your child's/teen's independence. Your goal should be to make your role much less important as years pass. Problem solving together while nurturing independence and treating or thinking of your teen "as if" they are capable of achieving their potential for living and working is important. Helping your teen learn more about their disease and how to manage the many aspects of it are crucial for understanding and living with TSC as well as accepting it.

Parents of children who have a problem of any type are prone to overprotect the child. If your teen has epilepsy, the problem is often worse because of the unpredictable nature of seizures. Parents of all teens must learn to gradually allow the teen to take more risks as they develop. Overprotecting your teen should be avoided as it may have more serious impact on emotional and social development.

ACCEPTING THE DIAGNOSIS

Teens will feel different and isolated and may experience poor self-image as angiofibromas become more apparent or as flair ups of seizures occur. The teen at some point will have to confront their own disability or health problem while adjusting to the changing demands of these difficult years. Teens may act out at school or withdraw. Compliance with medication or following suggested routines may be a struggle for a teen confronting their health problems or disability.

Issues of planning for the future and what to expect in adulthood may seem extremely overwhelming to a teenager who is unsure of themselves and of their diagnosis. Help your teen to build support systems outside of the family. As the adolescent discusses their illness with their friends it allows them to develop positive life skills necessary to manage their care (e.g., keeping scheduled appointments, when to take medicine or seek the advice of their physician, moderate physical activities, etc.). Encourage the adoption of behaviors that are favorable to their health and reinforce these behaviors (participating in extra-curricular activities, jobs, etc.).

CONCLUSION

Raising a teenager can be a very frustrating experience, however, understanding the unique perspective that your teen may

have is a first step in helping your teenager with the transition to adulthood and independence. Epilepsy can make parenting more difficult. TSC presents unique problems in parenting and difficult challenges. Studies involving chronically ill adolescents have identified how parents may need to remain very involved in their adolescent's health care but at the same time encourage growth and independence into adulthood.

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