The Tuberous Sclerosis Association

This document can be found at **www.tuberous-sclerosis.org**

It reports on a presentation at a meeting of the TS Alliance in San Diego in July 2001.

What Teachers Need to Know About Tuberous Sclerosis

Report of session given by Dr Cecilia McCarton, clinical professor of paediatrics, and Dr Candice Erikson, associate professor of clinical paediatrics.

Dr McCarton gave the first part of the presentation on learning difficulties in TS.

There are a number of brain lesions in TS, all of which are associated with learning disabilities – the cortical tubers, nodules, SEGAs or brain tumours, and cerebellum lesions.

The various types of learning disability in TS include mental retardation, attention deficit hyperactivity disorder, language disorder, problems with reading, writing, arithmetic, and problems with executive function and memory.

Mental retardation: in TS, the prevalence is 50-80%. If there is a cognitive impairment, this will affect all aspects of learning.

ADHD: this is quite common in TS, occurring in 20-45%. A child with ADHD may be fidgety, squirm in their seat, have their hands and head in constant motion, run round the room a lot. If such a child is in a classroom which has a number of distractions, it will be very difficult for them to learn in that kind of an environment. Inattention in TS has many forms. A child may be easily visually distracted, or be distracted by sounds (perhaps a chair scraping on the floor). A child may have limited attention, may move quickly from one task to another. They may loose things, such as books or pencils, or forget to bring their homework, or leave their jacket at school. They don't seem to listen to what is being said, are in another world. They don't have the ability to finish the work, it's difficult to engage them in a task and sustain it.

Impulsivity: It's difficult for these children to take turns. They jump right in, may talk excessively or make sounds excessively. They interrupt others, and don't respect anyone else's space. They blurt out answers and cannot wait their turn.

Language: This is one of the most difficult areas in TS. About 55% don't have language, and those that do have difficulties in expressing themselves and in understanding. They may find it difficult to put words together and so be restricted to single word expression. At home, visual prompts are often given with verbal directives,

such as pointing when asking them to “Go and pick that up”. Abstract questions, though, such as “How was school today?” are difficult to answer. These are usually the questions that begin with who, what, when, where, and why.

Temporal and spatial relationships are difficult, too. Telling the time, or understanding whether something is “in”, “on”, or “under” is also very difficult.

Short-term auditory memory: Some children can have a hard time remembering an instruction seconds after it was said. If is it coupled with a visual cue the retention of what was said will be much greater. So, if a child is a visual learner, show them or give them a picture to help them remember. You can often tell those children with an auditory processing disorder just by looking at them. You can see in slow motion that it’s taking them time to process what you said. They buy time by echoing it back to you, so you repeat it, or you then bring in a visual cue. After you’ve repeated it 2 or 3 times, the child will understand. There are complex tests which require the child’s co-operation (suitable for children from age 6 upwards) to test for this auditory processing disorder, and there is also a technique to help children with the problem (called auditory integration training).

Reading: Some children have a phonological impairment, a problem translating the written word to the equivalent speech word. For them, decoding the written word can be very hard. If a child has a short term memory impairment, the child may read something, understand it, then forget it within 5-10 minutes. This is a specific reading memory problem. And, of course, overall cognitive ability is important – you may or may not be able to read.

What does it take to read? There is a language and a visual processing area in the brain. Reading requires attention and a planning ability, the ability to read a line to the end, then to go down to the next line etc. Pathways are involved – information goes from the left eye to the left temporal area. Info from the right eye is transferred across the corpus callosum, also to the left side. The language centre is larger in the left than in the right. If there is a reading disability, PET scans have shown a decrease in activity in the left temporal lobe and abnormal functioning on the left side, dysfunctioning in visual systems (retina to visual cortex to temporal region). Also, abnormalities in the thalamus – important in motor planning, where the eye needs to drop to next line.

Writing: You may have problems with spatial disorientation and write everything up in one corner of the page, or put all the letters on top of each other. There are also memory and attention problems - you can’t write something down if you can’t remember it or if you haven’t paid attention. If you have a motor impairment, holding a pencil may be difficult, so it’s hard to write.

Maths: You may find it difficult to add, subtract, multiply or divide. Or you may not be able to read or write, you may not be able to make columns properly, so you put your tens and units in the wrong column, which then makes it impossible to add the figures up correctly.

Dysfunction of Executive Functioning: Executive functioning takes place in the frontal lobe. It is the ability to inhibit or defer a response, to formulate a sequential strategic plan of action, and to encode relevant information in your memory for future use. It is important in planning how to solve a problem. Without executive functioning, you have bad organisational skills, you are unable to lay out a logical plan. Executive functions are necessary for organisation, planning, problem solving, selective attention, vigilance, inhibition, creativity and impulse control. (Impulse control is when you take information in, pause, then take in other stimuli which might be important). People who lack executive function are very concrete, very literal, and inferences are very difficult for them.

Memory impairment can be auditory or visual. With a short term auditory memory problem you cannot retain information. With a short term visual memory , the teacher may have written something on the board and then erased it, but you can't remember what it is for long enough to it down. These problems can be tested on standardised testing.

Other conditions in TS such as autism or seizures impact on these learning disabilities, sometimes profoundly. Autism affects about 40% and involves problems with language, social interaction and repetitive behaviours and affects learning. Social isolation in children with autism leads to depression and secondary psychiatric problems. Seizures are prominent in TS and when someone is fitting they are not available to learn and take in stimuli.

Aggressive behaviours affect about 50%. They can be self-directed or directed at someone else, but they mean that you can't learn or access therapies that are going on. This means you may be isolated or even removed from school. Non-compliant behaviour in children with TS is associated with learning new tasks, where there is often great resistance.

Sleep problems affect many children with TS, and are associated with bi-frontal and temporal lesions:. We are not at our best the next day if we're deprived of sleep, and this makes it hard to learn. Sleep can also be affected by seizure disorders in TS.

How do you determine if someone really has a learning disability? There are tests to determine the degree of learning disability, but you need to be careful with these. If your child has a language problem, they won't do well on standardised testing because standardised testing assumes you have useful language - but to benefit from extra therapies, your child will need testing.

- Cognitive. There are various types of assessments for verbal and non-verbal IQ, but a child with an attention or motor problem may not be able to do some of these tests.
- There are also some academic achievement tests, for reading, writing and arithmetic.
- Speech and language evaluations look at expressive language, receptive language, word retrieval and auditory memory problems.

- Visual perceptual tests measure hand-eye co-ordination and will show if you have trouble with writing or visual perception.
- There are a variety of attention tests, including visual-auditory and school observation. To make a diagnosis of attention problems, you need to observe the child in school as well as at home. Many children can't be tested – they test you back in return and may not behave typically when you want them to!

Summary
There is an enormous range of learning disabilities in TS – language, reading, writing, attention. To deal with it, you must have an idea of what it is you're dealing with. For teachers to be able to help your child, you must be knowledgeable about what is wrong with your child, because your teacher often isn't.

Dr Erikson then addressed the problems of behaviour and behaviour management.

What your teacher needs to know about TS is really the same as what parents need to know. There are many behavioural issues in TS. The majority of people with TS have seizures, 50% have learning disabilities and many have stereotypic behaviours. Those with seizures have a greater risk of psychological and emotional problems, whether or not the seizures are controlled.

The unpredictability of seizures can affect a child, as can their lack of control over their seizures. The child fears being different from other children. As the child grows up, it is difficult for them to become more autonomous because of the seizures, and the care givers pass on their own anxiety about the child's seizures and the effect they have on his lifestyle. All of this has an effect on the child's emotions and behaviour. Seizures also have an effect on cognition and development, and the anti-epileptic medication has an effect, too. Different anti-epileptic medications cause various problems, such as cognitive dysfunction, concentration problems, lethargy, insomnia, mood disorders, disinhibition and sleepiness.

Children with mental delay or a language disorder get frustrated. Those with autism have sensory problems. Adults may have inappropriate expectations – they may expect too much, or indeed too little. And such children have a limited repertoire of coping strategies.

Behaviour impacts on school and cognitive functioning, on relationships at school and at home, on someone's own self-concept and on their leisure or play.

Behavioural disturbances impact at school, making it difficult for a child in the classroom, and making for poor relationships with other children and the teachers. They impact on the child's socialisation. They may be impulsively aggressive, have poor listening skills, be poor participants in team sports.

Treatment goals are to reduce major behavioural symptoms. There are some psycho-social interventions – behaviour management, social skills training, calming or relaxation

techniques – as well as sleeping medications, pharmacology, academic intervention, methods to improve the environment, and to improve language skills.

There are 3 categories of Behaviour.

1. Behaviour which is appropriate and desirable. This should always be reinforced.
2. Behaviour which is disruptive but not dangerous. Ignore it. (e.g. humming, fidgeting, just generally annoying)
3. Behaviour that is dangerous. You must block or punish this immediately. (e.g. aggressive behaviour).

Stereotypic behaviour such as rocking or hair twirling is very difficult to stop. You need to work at encouraging a more acceptable alternative but it you won't stop it altogether. Try to encourage a more acceptable alternative by reinforcing the alternative and ignoring the stereotypic behaviour.

Before you can do anything to change the behaviour, you need to find out the function of that behaviour, to find out why it is happening. If they have poor or no verbal skills, then TS children communicate by their behaviour. Just as a baby communicates by crying – with a hungry cry, or a tired cry, or a pick-me-up cry - so children with little or no speech resort to crying or demanding behaviour to communicate what they want.

Escape from demand is also a function of behaviour. A child who is resistant to learning a new task will do something to get out of it. You can use time-out in such a situation, as long as you make sure that after the period of time-out the child returns to the new task required of him! If by getting time-out the child escapes from demand successfully, he has achieved what he wanted so will try it again next time!

Attention is the single most powerful reinforcer of behaviour. A child always wants attention, especially parental attention, even if it's negative attention or scolding, which is actually an incredible amount of attention! Excusing yourself from a phone call to tell them off for being disruptive during that call actually gives them the attention they seek, thus reinforcing this behaviour. So, it is best to interrupt the call yourself when you see them playing nicely, just to say how much you like the fact that they're playing so well.

Self-stimulation – we all do it, whether it's chewing gum, smoking, tapping our feet etc. But our children may have self-stimulating behaviour which is very upsetting and which we'd like to replace with something more acceptable. Self-stimulation itself is such a positive reinforcer, but we can try to move away to something else.

Children with learning disabilities often have very little control over their lives and may adopt inappropriate behaviour to exert some control.

Illness or other medical problems may also drive their behaviour.

Behavioural Management involves Antecedent Management, which means changing the environment where the behaviour takes place.

Antecedent management:

1. Ensure the child has adequate sleep and nutrition, at home and at school. If the child is fatigued, let them rest at school until their sleep cycle is readjusted.
2. Physical energy – give them a time and a place to play and burn up some energy.
3. Minimise the effects of the medication. Be aware of the effects of your child's medication, and be aware if your child's behaviour is post ictal.
4. Attend to the physical arrangement of the room. Ensure that the classroom is uncluttered, that there are clear rules, that there is quiet and structure to the day.
5. Use calming techniques, give them ways they can calm themselves . Some children can use hypnotic techniques to calm them when they're anxious. Let them have familiar music at relaxing times of the day, which whenever they hear it will produce an automatic relaxing response which will help at times when they are anxious.
6. Use clear rules.
7. Have a reward system, using both praise and reasonable tangible rewards, too.
8. Follow formal behavioural programs.

Behaviour modification programmes are exhausting so you must take care of yourself. You'll need your own energy and resources to help your child. Recognise each small step along the way. Keep track of your child's behaviour in a journal and let the teacher know how things are going.

It's also hard work for the child, so when your child works well give him a reward, whether it's praise, a hug or whatever. Establish a routine, make things predictable for your child. Then you don't have to fight the battle every time since it's routine. Let the child be angry at a "thing" and not at you. So, if you are trying to get him to go to bed earlier, have a timer which rings out, so the child gets angry at the timer and not at you. Maintain consistency, that's very important, especially if the child is impulsive.

Adopt a few clear and consistent rules and avoid rules that the child can't possibly obey. Phrase things in the positive. How you use language is really important. If someone says "don't think of pink elephants", you immediately think of pink elephants! If they say "don't hit", the image that comes to mind is of someone hitting. If you say "I give you a hug because you didn't hit", you still create an image of hitting. So don't keep saying "don't do this" or "don't do that" since it creates over and over again the image of what you don't want the child to do! So, instead, say "Be gentle" or "be whatever….". Frame it positively.

Define what you want behaviourally. Don't say "be good" or "tidy your room". Say "Go upstairs. Pick up your toys and put them in the box before lunch". Reinforce the rules every time. With reinforcement, label what you're rewarding every time – "I like the way you're…."

Make the rules part of the routine. If your child says "Can I go out to play" you can answer "Yes, you can go out to play after you've changed your clothes", rather than "You can't go out to play until you've changed your clothes".

With rewards, avoid foods if possible. Praise or attention is a wonderful reward. Give appropriate rewards, and don't give one if they haven't deserved it.

Behaviours will always get worse before they get better. Bu don't abandon your attempt before they get better otherwise they'll scream longer and louder! Use planned ignoring along with positive reinforcement for something else they do that you approve of. Redirect the child to something else if possible, then praise the new behaviour while ignoring the old behaviour.

Consequence Management

- Use non-physical consequences. Hitting doesn't work and what you're teaching your child to do if you use it is to hit when you get angry.

- It's easier to earn privileges than to get rid of them. So, "every day, if you do this, you can do this …" . Let them earn their TV time by doing something you want, rather than have the TV taken away from them for doing what you don't want.

- And don't take away privileges you need! If you need them to sit quietly by a TV while you cook supper, don't punish them by taking away the TV!

- Don't make threats to remove privileges that you won't carry out.

- Time out can be very useful. It is a removal from a positive reinforcer, a removal from attention. It enables the child to cool down and is rather boring for them. Afterwards, the child must still do what he refused to do before the time out! Use a timer to help you and the child keep track of time.

- Psycho-social interventions in school.

- A daily report between school and home.

- Social skills training can be effective, especially if done in groups.

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