

Teachers and Staff Working in Education

This guide is written for teachers and staff working in education who may work with children and young people living with Tuberous sclerosis complex (TSC) now or in the future. Your knowledge, support and care can make an enormous difference in the educational outcomes for these students.

Children and young people living with TSC have a lot to offer their school or college community - they will bring skills such as attention to detail, special interests and a world view that can give their peers a different life approach.

About TSC

TSC is a multisystem genetic disorder with a range of physical features that require lifelong evaluation, surveillance and management. TSC can cause benign tumours to grow in one or more organs of the body. It can affect every organ system, with some more common during infancy and childhood and others more likely to affect individuals as adults. In the UK there are up to 10,000 people living with TSC.

Diagnosis is based on a careful physical examination, in combination with possible scans of the brain, lungs, liver, heart and kidneys. Genetic testing can be used to diagnose and/or confirm a diagnosis of TSC.

Getting the right care and management is crucial for improving the quality of life of children and young people living with TSC and those who care for them. An important part of this is the coordination of medical and therapeutic care from a range of professionals. This is particularly crucial during the transition from child to adult health and care services.

Some of the main features include:

- **Epilepsy is the most common neurological feature of TSC, occurring in 85% of individuals. Epilepsy is often difficult to control and at least 50% of people living with TSC develop refractory epilepsy (drug resistant epilepsy).^[1]**
- **About 35% of children living with TSC will have experienced infantile spasms.^[1]**
- **Kidney tumours (angiomyolipomas or AMLs) are common and may not cause any problems until adulthood. There is also an increased risk of cysts in one or both kidneys.^[1]**
- **TSC can affect the skin and lead to several conditions including reddish bumps or a rash on the face (angiofibromas) and small growths around the fingernails and toes (ungual fibromas).^[1]**
- **Lymphangiomyomatosis (LAM) is a lung complication of TSC which is most common in women between puberty and menopause.^[1]**
- **Heart tumours (cardiac rhabdomyomas) occur in about 50% of people living with TSC. These generally become smaller over time and only a few people will have long term problems.^[1]**

- People with TSC may have an overgrowth of the gums (fibromas) and dental pits (weakened enamel of the teeth).^[1]
- Cysts and AMLs may occur in other organs such as the pancreas, ovaries or liver.^[1]

International guidelines on the treatment and care of people living with TSC are available here - <https://bit.ly/2xTPQJc>. A set of UK guidelines is due to be published winter 2018.

TSC Associated Neuro-Psychiatric Disorders (TAND)

People living with TSC also have a range of Neuro-Psychiatric issues. Around 90% will have some of these difficulties during their lifetime. These often represent the greatest challenge of the condition and include:

- Behavioural - examples include repetitive or ritualistic behaviour, poor eye contact, shyness, hyperactivity, restlessness and impulsivity.
- Psychiatric - the most common psychiatric diagnoses in TSC are autism, ADHD, anxiety and depression.
- Intellectual – there is a wide range of intellectual ability within TSC ranging from high to low.
- Academic – there is a wide range of academic ability in TSC, and even those without intellectual impairment, may still need support with their learning needs.
- Neuropsychological – this covers brain skills, such as communication and language, memory, attention, planning and visuospatial skills.
- Psychosocial – covers quality of life elements such as self-esteem, relationship difficulties and family stress.

Monitoring any behavioural or psychological change is important as this may also indicate a change in the physical health of a child or young person living with TSC.

A TSC TAND Checklist has been developed to help families and people living with TSC have a conversation with their clinicians to identify difficulties early and to prioritise treatment. Items listed below can be found on the Tuberous Sclerosis Association (TSA) website- www.tuberous-sclerosis.org.

- The TAND checklist can be found here - <https://bit.ly/2ObP3wn>.
- Guidelines for the Assessment of Cognitive and Behavioural issues in TSC can be found here - <https://bit.ly/2Faz6yE>.
- The Neuropsychiatry of Tuberous Sclerosis - A presentation on YouTube by Prof Petrus de Vries can be found here- <https://bit.ly/1xhVTSX>.

Learning and TSC

For students whose TSC affects their brain, issues with learning and retaining information can develop. Children and young people who have no intellectual impairment, may still require support to manage learning needs within the classroom setting. An assessment by an educational psychologist can be helpful in determining a baseline and finding ways of supporting a student's learning style. No two students living with TSC are the same, and many children and young people living with TSC may have an uneven profile – having great skills in some areas, and low ability in others.

For some students their abilities may alter over the course of the academic year, depending on their seizures and impact of any tumours that may have grown, or any other health need they may have. More regular assessments may be needed each term to look at progress and support needs. Any changes in behaviour or development, may indicate that tumours have grown, it is important to flag any changes with other staff and carers.

Children and young people living with TSC often have additional learning needs and may need specific help with reading, maths, writing and literacy-based tasks within the classroom. Processing language may be difficult and written or visual cues may assist help keep them on target.

Autism and TSC

There is an increased incidence of autism in children and young people with TSC. Some of the management techniques that you may use with children and young people with autism will also work with children and young people living with TSC. See National Autistic Society website for more details (www.autism.org.uk).

There are many positives too – it may also mean they have a high level of skill in some areas – enjoying logical subjects such as maths or computer science and expertise in their chosen interest – anything from road maps to historical battlefields.

Language and Communication – what helps

- **Say the child's name first to get their attention before giving any further instruction.**
- **Break down instructions step by step.**
- **Repeating instructions where necessary.**
- **Use visual cues and picture timetables where possible.**
- **Sensory issues may interfere with a child's response – adjusting seating or position of desk may help.**

Absence from school

TSC is a complex condition which requires input from a range of medical and therapeutic professionals. This may mean frequent appointments and time away from the curriculum. For children who have seizures and are unwell because of their TSC, this may mean further absences from school or college. This may be difficult for the child or young person and their family. Finding ways to keep the child or young person involved in school life as much as possible can help minimize the effects.

Further Learning

BMJ Module–The TSA has worked with the British Medical Journal (BMJ) to develop a module on TSC. It is designed to help health professionals recognise TSC and offers more information about the treatments currently available. The module is available for anyone to access and is free of charge. Users who successfully complete the module will receive one Continuing Professional Development (CPD) point.

Register at <https://learning.bmj.com/learning/home.html>.

Epilepsy Action produce resources and training materials for staff and students.

See www.epilepsy.org.uk

Support from the Tuberous Sclerosis Association (TSA)

- The TSA is the only UK charity dedicated to supporting people affected by TSC.
- We have a team of advisers around the UK offering help and support to people living with and those affected by TSC- see www.tuberous-sclerosis.org for more details.
- Support is available to professionals who are working with people who are living with TSC.
- There are multidisciplinary TSC Clinics throughout the UK, contact the TSA for details of your nearest TSC Clinic.
- Research is ongoing to find new treatments and to make them available to all who need them.

For more information about the TSA and its work go to www.tuberous-sclerosis.org or contact admin@tuberous-sclerosis.org or 0207 922 7731 (main switchboard)

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We can provide this information in other formats such as large print or audio and in other languages.

Disclaimer

We have made every effort to ensure that information in this publication is correct at the time of going to print. We do not accept liability for any errors or omissions and policy and practice may change.

Reference

Website:

1. Tuberous Sclerosis Alliance (2018) *Signs and Symptoms of TSC*, 6 Sept 2018 [<https://www.tsalliance.org/about-tsc/signs-and-symptoms-of-tsc/>]

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