



## Self-injury and aggression in adults with tuberous sclerosis complex: Frequency, associated person characteristics, and implications for assessment



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### ABSTRACT

Even though self-injury and aggression are common in tuberous sclerosis complex (TSC), understanding of these behaviours in adults with TSC and intellectual disability (ID) is limited. Little is known about their frequency in comparison to other ID-related genetic disorders or their association with other TSC-Associated Neuropsychiatric Disorders (TAND). This study determined the caregiver-reported frequency of self-injury and aggression in adults with TSC plus ID in comparison to Down syndrome (DS) and Angelman syndrome (AS), and assessed demographic and behavioural characteristics associated with the occurrence of each behaviour in TSC. Rates of self-injury and aggression in adults with TSC plus ID were 31% and 37.9% respectively. The odds of self-injury for adults with TSC were nearly twice as high as the odds for adults with DS, and the odds of aggression were over 2.5 times higher for adults with TSC than for adults with DS. When compared to adults with AS, odds of self-injury in TSC were around half those of the AS group, and odds of aggression were less than a third of those for adults with AS. These differences were not statistically significant. In adults with TSC, poorer communication and socialisation skills, gastric health problems and impulsivity were associated with self-injury; compulsive behaviour and impulsivity were associated with aggression. Caregivers and professionals should be alert to the likelihood of these behaviours in adults with TSC plus ID, and to characteristics associated with increased risk for their occurrence. We suggest assessment strategies to identify those at elevated risk.

*What this paper adds:* This paper adds specific examination of behavioural difficulties in adults with tuberous sclerosis complex who also have intellectual disability, a population at heightened risk of adverse behavioural outcomes which has received limited focussed examination to date. Findings support existing suggestions that there is relatively high risk for both self-injury and aggression, and provide novel insight into characteristics that may be associated with the presence of these behaviours.

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## 1. Introduction

Tuberous sclerosis complex (TSC) is a genetic disorder characterised by abnormal growths in multiple organs, including the central nervous system, caused by mutation of either the *TSC1* gene on chromosome 9q34 (van Slegtenhorst et al., 1997) or *TSC2* gene on chromosome 16p13 (European Chromosome 16 Tuberous Sclerosis Consortium, 1993). Between 70 and 85% of those with TSC have seizure disorders (Chu-Shore, Major, Camposano, Muzykewicz, & Thiele, 2010), and it is acknowledged that seizure history and severity can be associated with poorer cognitive and neuropsychiatric outcomes (Bolton, Park, Higgins, Griffiths, & Pickles, 2002; Chu-Shore et al., 2010). TSC-Associated Neuropsychiatric Disorders (TAND) are seen in the majority of individuals with TSC (Curatolo, Moavero, & de Vries, 2015; de Vries et al., 2015; Leclezio & de Vries, 2015). Approximately 45% of individuals with TSC have intellectual disability (ID) (Joinson et al., 2003), around 40–50% meet criteria for autism spectrum disorder (ASD) (Bolton et al., 2002) and 50% for attention deficit hyperactivity disorder (ADHD) (de Vries, Hunt, & Bolton, 2007). Self-injury and aggression are reported frequently. A recent summary of TAND (Leclezio and de Vries, 2015) estimated rates of self-injury to vary from 17 to 69%, and aggression from 51 to 66%. Most of what we know about TAND is based on studies of children and adolescents, rather than adults.

In the wider population of people with ID, self-injury and aggression affect the physical health and quality of life of affected individuals and their families (Borthwick-Duffy, 1994; Konarski, Sutton, & Huffman, 1997; Nissen and Haveman, 1997; Spreat, Lipinski, Hill, & Halpin, 1986). For TSC specifically, clinically elevated levels of parenting stress are reported by over half of caregivers, and stress levels are associated with problem behaviours (Kopp, Muzykewicz, Staley, Thiele, & Pulsifer, 2008). In adulthood, difficult behaviours are perceived to impact the family negatively, and to affect caregivers' decision-making about potential out-of-home placements (McIntyre, Blacher, & Baker, 2002). The impact of self-injury and aggression may become more detrimental with age as these behaviours become more difficult to manage. Severe self-injury and aggression can affect access to services, result in exclusion from placements and require more costly services, issues particularly relevant to older individuals likely to live away from home (Emerson, 2001).

A substantial literature describes the prevalence and phenomenology of self-injurious and aggressive behaviours in individuals who have ID. In a total population study of individuals with ID, prevalence rates of 4% for self-injury and 7% for aggression have been found (Emerson et al., 2001). A range of person characteristics have been described in association with the development of self-injury and aggression in individuals with ID. Such characteristics may facilitate identification of those at elevated risk for developing these behaviours. These include specific genetic disorders, ASD, features of ADHD, low mood and communication deficits (Arron, Oliver, Moss, Berg, & Burbidge, 2011; McClintock, Hall, & Oliver, 2003; Oliver & Richards, 2015). Pain and illness also increase the likelihood of self-injury and aggression (Carr & Owen-DeSchryver, 2007). These observations are highly pertinent to TSC, given the high prevalence of ID, ASD and ADHD, as well as the complex health problems potentially associated with pain (e.g. renal angiomyolipomas that can cause flank pain and bleeding, subependymal giant cell astrocytomas that can cause increased intracranial pressure accompanied by headaches, and the discomfort associated with seizures, Roach & Sparagana, 2004).

To date only one study, conducted by Eden, de Vries, Moss, Richards, and Oliver (2014), examined whether rates of self-injury and aggression in TSC are comparable to those seen for other syndromes also associated with ID, and assessed which person characteristics may predict those most at risk for these adverse outcomes. Rates of self-injury and aggression, and a range of characteristics previously found to be associated with these behaviours, were examined in children and adolescents with TSC aged 4–15 years, using questionnaire methodology. Data for children and adolescents with TSC, as well as those with idiopathic ASD, fragile X syndrome, and Cornelia de Lange syndrome, were contrasted with data for a group with Down syndrome (DS). DS was chosen as the reference group due to its status as an ID-associated genetic syndrome with a relatively well-characterised behavioural phenotype. The prevalence of self-injury and aggression in children and adolescents with TSC was 27% and 50% respectively (Eden et al., 2014). Whilst these frequencies were considerably higher than estimates of these behaviours in people with ID in general, the odds of children and adolescents with TSC showing self-injury and aggression did not statistically significantly exceed those of the DS reference group (Eden et al., 2014). Presence of self-injury and aggression in children and adolescents with TSC was predicted by a number of anticipated person characteristics including stereotyped behaviours, low mood, overactivity, impulsivity, repetitive use of language, and, importantly, pain-related behaviours.

The developmental course of self-injury and aggression in TSC, including any age-related changes in the prevalence of these behaviours, and stability of their associations with person characteristics across the lifespan, is currently unknown. Such an understanding is important, however, as the behaviours are likely to be associated with negative outcomes both for adults with TSC and those who care for them. Given the absence of any literature examining the issue of self-injury and aggression in adults with TSC, the current study aimed to examine the frequency of these two behaviours in adults with TSC plus ID, to quantify the odds of these behaviours relative to two reference groups and to identify whether the person characteristics associated with self-injury and aggression found in children and adolescents with TSC in the Eden et al. study (2014) were similar in adults. In the current study a DS reference group was employed, in line with the Eden et al. study, and an Angelman syndrome (AS) reference group was added to enable comparison to a genetic syndrome also associated with high levels of epilepsy (around 90% of individuals are reported to experience seizures) and with increased risk of adverse behavioural outcomes, particularly aggression (Arron et al., 2011; Pelc, Boyd, Cheron, & Dan, 2008).

Given the existing literature the following hypotheses were proposed:

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