Sleep Disturbance in Tourette’s Syndrome: a review

Dr Bushra Hasnie
Senior House Officer
Bedfordshire and Luton Mental Health and Social Care Partnership NHS Trust

Introduction

Gille de la Tourette’s syndrome (TS) is a neuro-psychiatric disorder that is characterised by multiple motor (movement) and vocal tics. The clinical features usually begin in childhood and persist into adulthood, although symptoms may vary in severity and can wax and wane (Cohrs et al 2001). It occurs more commonly in males than females (Apter et al 1991) and is associated with disturbances of sleep. This paper describes some of the changes that occur in sleep patterns in people with TS (Romano et al 2004).

Symptoms in TS

A tic can be defined as a sudden, rapid, recurrent, non-rhythmic stereotyped movement or vocalisation. Vocal tics include grunting, snarling and similar noises. Complex motor tics are slower movements that appear as if they have a purpose, such as touching, biting, head banging and itching. Obsessive Compulsive Disorder (OCD) and Attention Deficit Hyperactivity Disorder (ADHD) occur frequently in people with TS. There are other associated symptoms such as the automatic and pointless imitation of another individual’s speech and movements, known as echolalia and echopraxia respectively. Additionally there may be over-activity and difficulties in learning.

Pattern of Sleep

Sleep can be considered a stage of extremely reduced or nearly absent voluntary control of cognition, emotion and movement (Cohrs et al 2001). Sleep is usually divided into REM (rapid eye movement) or non-REM sleep. REM sleep is also called dream sleep. Throughout the night the normal pattern of sleep involves 4-5 cycles of alternating REM and non-REM sleep with REM sleep becoming progressively more prominent. The total time spent in REM sleep is ninety minutes or twenty per cent of the total sleep period. Brief periods of wakefulness comprise five percent of sleep and the remaining three quarters of time is spent in non-REM sleep.

Sleep Disturbance in TS

Sleep problems can affect up to 60% of people with TS (Romano et al 2004) and complaints about difficulty falling asleep or early wakening
are the most often reported (Cohrs et al 2001). These alterations in sleep occur in people who have both TS and Attention Deficit Hyperactivity Disorder (ADHD), with around 40% of people in this group raising concerns that their sleep was affected (Allen et al 1992). Sleep disorders may be linked to disturbances in the build up or breakdown of chemical transmitters of the brain, particularly serotonin. Sleep disturbances are found more frequently among children with TS than among adults with TS and may be linked to a delay in the normal processes of maturing (Champion et al 1988).

Sleep quality, quantity and motor activity during night sleep in people with TS is also affected. The total time period spent asleep may be decreased in those with TS and a study looking at children with TS who were not on medication at the time, showed decreased sleep. The length of time taken to fall asleep is known as sleep latency and this is increased in patients with TS. That is to say, it takes longer for people with TS to fall asleep than those without TS (Cohrs et al 2001, Kostanecka-Endress et al 2003). An increased sleep latency is also true for people with a combination of both TS and obsessions and compulsions. These obsessions and compulsions may themselves be manifestations of chemical disturbances in the brain involving dream sleep (Drake et al 1992). Overall, the efficiency and therefore effectiveness of sleep in children with TS is also reduced and the more severe the TS in the day, the poorer the sleep efficiency (Cohrs et al 2001, Kostanecka-Endress et al 2003, Drake et al 1992).

There is prolonged wakefulness after the onset of sleep (Kostanecka-Endress et al 2003). This is supported by recordings of electro-encephalogram (EEG) brain wave patterns of people with TS, which show an increased number of awakenings (Drake et al 1992). In tic disorders alone, nocturnal awakenings and movements are greater. Chronic tics may persist in sleep and cause awakenings. There is a higher reported frequency of sleep walking and night terrors in children with TS. Disorders of initiating and maintaining sleep have also been recorded and there is an association between bedwetting at night (nocturnal enuresis) and TS (Romano et al 2004, Stores 2001). The severity of TS during the daytime is associated with a decreased sleep quality. Sleep with an increased number of awakenings as a sign of reduced sleep continuity is known to be less restorative and children with TS may be more irritable during their waking hours because of their reduced sleep quality (Cohrs et al 2001).

Non REM (or non-dream) sleep is divided into four stages of sleep. Stages 1 and 2 comprise lighter sleep. Collectively stages three and four are called slow wave sleep otherwise known as deep sleep. Generally, the number of sleep stage changes is unaffected (Kostanecka-Endress et al 2003) although a correlation with the severity of TS and sleep stage changes has been suggested (Cohrs et al 2001). Short bursts of brain electrical activity usually occur in stage 2 sleep. These bursts of brain activity are increased in those with TS and therefore sleep is lighter (Silvestri et al 1995). There may be less stage 2 sleep in children with TS (Kostanecka-Endress et al 2003). There is also a greater percentage of stage 1 sleep and therefore people with TS tend to have lighter sleep (Cohrs et al 2001). Deep sleep or slow wave has been shown to vary (Drake et al 1992, Silvestri et al 1995). In those patients with both TS and ADHD, the loss of slow wave or deep sleep is most marked as compared to those with TS, the TS and OCD group and tic disorders alone (Drake et al 1992). This means that children with TS may not feel as rested as those without. Movements that occur typically when awake are seen occasionally during
sleep and are most likely to occur after awakening from sleep (e.g. stage one sleep) and rarely during the deeper phases of sleep. Movements that occur during sleep without awakenings are usually preceded by bursts of brain activity (sleep spindles on the EEG) or slow waves (Fish et al 1991).

REM sleep in TS can vary. One study indicated that REM sleep was actually decreased (Silvestri et al 1995). In people with both TS and OCD less time is needed before REM sleep is reached. Another study showed that children with TS and ADHD groups had increases in REM sleep latency, so that more time was needed before dream sleep began. This group had a significantly higher rating of aggressive behaviour.

Tics occur in all sleep stages (Cohrs et al 2001). Tic frequency and regular movements are higher in REM than non-REM sleep. The increased rate of tics during REM sleep parallels the increased movements of patients during REM and non-REM sleep (Cohrs et al 2001). Some people with TS show increased partial arousals out of deep stage 4 sleep. These sudden intense, partial arousals can manifest as night terrors, sleep walking (or somnambulism), bedwetting (or enuresis) or behaviours like forced vocalisations which often take the form of obscene words, phrases (coprolalia) or 'bird-calls'. Both tics and partial arousals from sleep decrease with treatment of TS (e.g. clonidine) (Dhal & Puig-Antich 1990). The poor sleep quality in children with TS following on from increased arousal phenomena could be intrinsic to the disorder or might trigger behavioural problems such as irritability during the daytime. Tic Disorders and REM sleep behaviour disorders co-exist and there is overlap in the pathophysiological mechanisms of both (Trajanovic et al 2004).

Periodic limb and arm movements (PLMS) during sleep are a frequent finding in patients with TS. The presence of PLMS during sleep in TS may point towards evidence for a relationship between TS and restless leg syndrome, since they both share a disorder of one of the chemical pathways of the brain (dopaminergic system) but this has not been confirmed by the different responses to treatments (Voderholzer et al 1997).

Sleep disorders following treatment with medication have also been described. An irregular sleep-wake pattern can occur with anti-psychotic treatment (e.g. haloperidol) and be restored by using alternative medication (e.g. risperidone). It is possible that the described disruption of the sleep-wake schedule is medication rather than illness-related in TS. Therefore it is important to keep in mind that sleep disorders may be a side effect of medication rather than TS itself (Ayalon et al 2002). However, in one particular case of a boy with TS, an improvement was seen in sleep related symptoms, coupled with less daytime irritability and tiredness following a change in the scheduling of medication to include a bedtime dose.

Serotonin and noradrenaline are involved in sleep regulation as well as in TS. An unaltered serotonergic firing is a prerequisite for sleep maintenance. There is a positive correlation between TS severity and features of disturbed sleep quality, which is compatible with the inverse correlation between tic severity and cerebrospinal fluid changes of tryptophan, a precursor substance of serotonin (Cohrs et al 2001).

TS gene carriers are at increased risk of life threatening apnoeas of infancy. The cause of sudden infant death syndrome is unknown but an impairment of arousal and hyperdopaminergic and hyposerotonergic dysfunction, such as seen in TS, may contribute to the pathophysiology of these
sleep disorders. The prevalence of sudden infant death syndrome in the families of those with TS may be up to five times the prevalence in the general population (Sverd & Montero 1993).

Conclusion

In summary, there is an association between disturbances of sleep and Tourette’s syndrome (Cohrs et al 2001). These sleep disturbances are characterised by changes in the length of time spent asleep, which may be increased or decreased, the greater length of time taken to actually fall asleep, poor sleep quality and reduced sleep efficiency. Prolonged wakefulness after sleep onset is seen with a greater number of awakenings during sleep. There are alterations in light sleep so that there is a greater percentage of stage 1 sleep with less or the same amount of stage 2 sleep. REM sleep quality is reduced or unaffected and slow wave or deep sleep may be increased or decreased. Tics are greatest in REM sleep and partial arousals or movements occur in stage 4 sleep. General movements and periodic limb movements are increased and a REM sleep behaviour disorder has been described in patients with tics. Generally, the sleep wake cycle is disrupted. Further research is required to establish any consistent pattern of sleep changes in TS and the cause of sleep disorders in TS. Medication used in the treatment of TS may be implicated in disturbances of the sleep-wake cycle. The exact nature of sleep disturbance in children with TS remains to be elucidated.

References


