Sleep disorders in children with Down syndrome

Professor Gregory Stores
MD MA DPM FRCPsych FRCP

University of Oxford
Prevalence of childhood sleep problems

General population 25%

High risk (50-90%)
- neurodevelopmental disorders
- psychiatric disorders
- chronic paediatric conditions

Down syndrome 31-54% *(Tietze et al 2012)*
Neglect of children’s sleep problems

Neglect in public health education and professional training

Limited provision of help and advice

Particularly unfortunate regarding children with a developmental disorder
Sleep disturbance in neurodevelopmental disorders: general points

Same sleep disorders and consequences as other children
Severe, persistent if not treated
Causes
  Behavioural
  Physical
  Co-morbidity (Bax & Gillberg 2010)
Treatment possible
Potential benefits from improving sleep
Effects of poor sleep

A. Emotional state and behaviour

Sleepiness, fatigue
Irritability, aggression
Anxiety, depression
Effects on family well-being
Impaired quality of life
B. Cognitive function

Attention, vigilance
Memory
Motor skills
Abstract or creative thinking
Complex tasks
Poor educational progress
C. Physical health

*(Colton & Altevogt 2006)*

Gastrointestinal disorders
Cardiovascular disease inc hypertension
Menstrual, pregnancy problems
Obesity
Diabetes
Impaired immunity including infections
Growth effects
Types of sleep problem in children with Down syndrome

Bedtime settling and night waking problems (Richman et al 1975)

Obstructive sleep apnoea (Silverman 1988)

Sleeplessness (including sleep-wake cycle disorders)
Excessive daytime sleepiness
Parasomnias
Possible causes of sleep problems in children with Down syndrome

As other children

Difficulty learning good sleep habits (range of intellectual impairment)

Comorbidities (medical, psychiatric)

Medication effects
Examples of causes of sleep problems in children in general

Parenting practices
  lack of routine
  poor limit setting
  inadvertent reinforcement

Psychological problems including anxiety or depression

Adolescence: pubertal changes in sleep physiology; altered life-style
Down syndrome comorbidities likely to be associated with sleep problems: physical conditions
(Charleton et al 2011; Stores & Wiggs 2001)

Obstructive sleep apnoea (50%+)
Cardio-respiratory disorders
Visual and hearing defects
Pain eg musculo-skeletal disorders
Gastro-oesophageal problems
Skin disease eg atopic dermatitis
Thyroid disorder
Obesity (Murray & Ryan-Krause 2010)
Epilepsy (5-10%)
Sleep Disturbance in Children and Adolescents with Disorders of Development: its Significance and Management

Edited by Gregory Stores and Luci Wiggs
OSA in neurodevelopmental disorders

Down syndrome
Mucopolysaccharidoses
Prader-Willi (some)
Fragile X
Cerebral palsy
Causes of respiratory obstruction in children with Down syndrome

Midfacial and mandibular hypoplasia
Large posteriorly placed tongue
Congenital narrowing of the trachea
Hypotonia of the pharyngeal musculature
Larygomalacia
Infection of tonsils and adenoids
Obesity
Parasomnias associated with OSA
(Schenck & Mahowald 2008)

Children

Arousal disorders (sleepwalking, sleep terrors, confusional arousals)
Nocturnal enuresis
Bruxism

Adults

Arousal disorders
Sleep-related eating disorder
Nightmares
REM sleep behaviour disorder
Nocturnal epileptic seizures

Successful treatment of OSA reduces parasomnias
OSA and epilepsy

OSA can trigger seizures
  sleep fragmentation
  sleep deprivation
  hypoxia

Treatment of OSA can improve seizure control

Misdiagnosis
Down syndrome comorbidities likely to be associated with sleep disorders: psychiatric conditions

(Dykens 2007; Ivanenko & Johnson 2008; van Gameren-Oosterom et al 2011)

Anxiety states
Depression
Conduct disorder
Attention deficit hyperactivity disorder (ADHD)
(Ekstein et al 2011)
Autistic spectrum disorders (ASD) (Magyar et al 2012)
Medication as a possible cause of children’s sleep problems  
(Mindell & Owens 2010)

Sleeplessness
   - Stimulants for ADHD
   - Decongestants (pseudoephadrine)
   - Bronchodilators (theophylline)
   - Beta antagonist antihypertensive agents (propanalol)
   - Analgesics (NSAIDs)
   - Some antidepressants

Excessive sleepiness
   - Sedatives-hypnotics (benzodiazepines, antihistamines)
   - Major tranquillisers
   - Some antiepileptic drugs

Parasomnias (eg sleepwalking)
   - Some hypnotics eg zolpidem
   - Some antidepressants
Assessment

Sleep problem may not have come to light

Screening for symptoms and signs

Diagnosis of sleep disorder
Sleep problem may not have come to light

Parents lack basic knowledge about sleep

(Owens & Jones 2011)

Even severe sleep problems not reported to family doctors

(Blunden et al 2004; Robinson & Richdale 2004)

Overlooked by paediatricians

(Owens 2001; Chervin et al 2001)

Importance of screening including for OSA in children with Down syndrome (Rosen et al 2011)
Routine screening for sleep problems

History taking
- Bedtime difficulties or problems falling asleep
- Breathing difficulties while asleep
- Waking in the night
- Unusual behaviour, experiences or movements at night
- Difficulty waking up in the morning
- Sleepy or ‘overtired’ during the day
The Children’s Sleep Habits Questionnaire completed by parents

(Owens et al 2000; Goodlin-Jones et al 2008)

Versions for toddlers-preschool and school-age children

Measures

- Bedtime resistance problems
- Wakings during the night
- Difficulty getting to sleep
- Short duration sleep
- Anxiety about sleep
- Sleep disordered breathing
- Parasomnias
- Daytime sleepiness

Satisfactory psychometric properties

Used with typically developing and developmentally delayed children
Sleep disturbance

Three sleep problems (complaints)

Insomnia/sleeplessness
Excessive daytime sleepiness
Parasomnias

ICSD-2: over 80 sleep disorders ie possible underlying psychological and physical causes of the sleep problems
Precise diagnosis required
Diagnosis of sleep disorder

**History** (from parents and others)
- Precise description of sleep problem
- 24hour sleep–wake pattern
- Developmental history (medical, psychological)
- Family history
- Family circumstances

**Examination**
- Physical
- Behaviour

**Further assessments**
- Questionnaires
- Diaries
- Sleep studies (polysomnography, actigraphy)

**Referral to sleep specialist?**
Management of children’s sleep disorders

General principles

Education for parents and children
Sleep hygiene
Treat any underlying disorder

Behavioural methods

Pharmacological

Hypnotics
Melatonin
Stimulants

Other eg dopaminergic agents or iron supplements for RLS/PLMD

Other

Chronotherapy (sleep/wake cycle disorders)
Physical (adenotonsillectomy or CPAP for OSA)
Treatment of sleep disorders in children with a developmental disorder

Sleep education for parents including treatment prospects

Sleep hygiene  (Jan et al 2008)

Behavioural treatment  (Richdale & Wiggs 2005)

Medication  (Pelayo & Dubik 2008)
Treatment for sleep disorders in children with Down syndrome

Treatment for childhood sleep disorders *per se*
Mainly behavioural rather than pharmacological 
(Kuhn & Elliott 2003)
Group training in behavioural methods 
(Stores R et al 2004)
Improvements in children’s behaviour and parental wellbeing (Richdale & Wiggs 2005)

Management of comorbid conditions
OSA complicated (Rosen 2011)

Adjustment of medication
Main points

Sleep disturbance is a significant complication to the lives of children with Down syndrome and their families.

Multifactorial causes complicates treatment which is possible in principle.

Need for increased awareness of problem and possible help available.

Further more sophisticated research required with full diagnostic and treatment resources.