IRISH SLEEP SOCIETY
Cumann Codhladh na hÉireann

Practice Management Support Document

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Practice Management Support Document
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This is the second set of practice guidelines for the assessment and management of patients in Ireland.

This Practice Management Support Document has been written by local experts, synthesising the information and recommendations of International Societies but with the extra consensus expertise of local experts who have adapted the information to reflect the nature of Health Care provisions in Ireland.

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Humans spend up to one third of their lives asleep, yet surprisingly little is known about the biological role of sleep, and little attention has been paid until recent years to sleep disorders as an important cause of ill health. There is general agreement that sleep is beneficial. Shakespeare’s Macbeth described sleep as the “chief nourisher in life’s feast”, and the age-old remedy for the sick person is to get “plenty of sleep”. Some classical writers, however, have viewed sleep with deep suspicion. Sir Thomas Browne wrote “Sleep, in fine, is so like death, I dare not trust it with my prayers”, and Tennyson described sleep as “death’s twin brother”.

The modern practice of sleep medicine and the investigation of sleep-related disorders owe much to the discovery of electroencephalography in 1928, which demonstrated clear differences between wakefulness and sleep. In 1953 rapid-eye-movement (REM) sleep was described and it was proposed that dreaming was a function of this sleep stage, which led to the concept of two distinct sleep states, namely non-REM and REM. The physiological roles of non-REM and REM sleep appear fundamentally different in that there is evidence that non-REM sleep, especially slow-wave sleep, is primarily concerned with restorative functions, whereas REM sleep appears to be more concerned with synthetic functions in the brain.

Sleep-related disorders are remarkably common and insomnia affects most people at some stage in life, usually stress-related. Chronic insomnia is highly prevalent and can have major adverse effects on quality of life and functional status. Management is often difficult and requires careful and detailed assessment of the sufferer by an experienced practitioner. Sleep-related medical disorders are also highly prevalent, particularly sleep apnoea syndrome, which affects at least 4% of the adult population. Sleep apnoea is associated with significant morbidity and mortality, and has been clearly associated with an increased risk of heart attack and stroke, in addition to a significant risk of automobile accidents and injury in the workplace because of the associated daytime sleepiness. However, the condition is readily treatable, and affected patients can respond dramatically well to the continued home use of nocturnal continuous positive airway pressure (CPAP).
The growth and development of sleep medicine in Ireland has been a gradual process over the years. The first clinical sleep laboratory was established in St. Vincent’s Hospital, Dublin, in 1985, and over the intervening years several other laboratories have been established throughout the country. Sleep medicine is a multi-disciplinary specialty involving different medical specialists including Respiratory Physicians, Neurologists, ENT Surgeons, Psychiatrists and Dental specialists, in addition to Physiologists, Scientists and Nurse Specialists. The high prevalence of sleep disorders and the multi-disciplinary nature of patient care make the development and implementation of guidelines desirable in order to promote the highest standards of patient care.
Section 1. Obstructive Sleep Apnoea (OSA)

Definition:
Obstructive Sleep Apnoea Syndrome (OSAS) is a complex disorder characterised by brief interruptions of breathing during sleep. Airflow into and out of the lungs is reduced or diminished due to closure of the upper airway, despite continued respiratory effort. The most common presenting symptoms are excessive daytime sleepiness, loud snoring and witnessed apnoeas. The condition is diagnosed by an objective measure of abnormal nocturnal respiration coupled with a compatible clinical picture.

Pathophysiology of OSAS:
This condition is characterised by intermittent upper airway obstruction during sleep due to multiple factors, including physical narrowing of the upper airway, muscle fatigue of the upper airway muscles and/or a neurochemical imbalance of respiratory drive. These events are commonly associated with recurring episodes of arterial oxygen desaturation. Apnoeas are typically terminated by a brief micro-arousal resulting in sleep fragmentation and diminished amounts of slow-wave and REM sleep. These mechanisms have been reviewed in detail by White et al.

Epidemiology:
No data are available at present on the prevalence of OSAS in the Irish population, but international data indicate that at least 4% of the middle aged adult population have OSAS. These data imply that there may currently be over 50,000 sufferers in Ireland. ISS recommends that research be conducted in this area as soon as possible to more accurately determine the extent of this syndrome in the Irish population.

Risk Factors for development of OSAS:
- Obesity
- Upper airway abnormality – Retrognathia, Tonsillar Hypertrophy, Receding Jaw, Macroglossia, Nasal Obstruction
- Endocrine disorders including Acromegaly, Hypothyroidism
- Postmenopausal state (females)

Consequences for non-treated OSAS:
- Hypertension
- Ischemic heart disease
- Stroke
- Diabetes Mellitus Type 2, poor diabetic control
- Sudden death
- Impaired quality of life
- Road traffic accidents (RTA)

Why the need for diagnostic sleep studies?

Clinical probability of OSAS based on history and examination compared with AHI obtained from full overnight sleep studies in 250 consecutive patients referred to St Vincent’s University Hospital with suspected OSA. Deegan ERJ 1996.
Symptoms and Clinical Features of OSAS:
The following symptoms are associated with OSAS, but may not all be present in each patient.

Table 1 Major Symptoms and Clinical Features associated with OSAS

<table>
<thead>
<tr>
<th>Daytime Symptoms</th>
<th>Night-time Symptoms</th>
<th>Clinical Features</th>
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<tbody>
<tr>
<td>Excessive daytime sleepiness (EDS more likely to be reported by men)</td>
<td>Snoring (often loud) / Snorting Witnessed apnoea Fragmented/Restless sleep Nocturia/Enuresis Night time sweating Nightmares/Unpleasant dreams Recurrent arousals Morning headache Nocturnal choking/Gasping Impotence</td>
<td>Narrowed upper airway/ Craniofacial anatomy/retrognathia Systemic hypertension Obesity Cardiac arrythmias</td>
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<td>Fatigue (this is more likely to be reported by women)</td>
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<td>Neurocognitive dysfunction</td>
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<td>Personality changes</td>
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<td>Depression</td>
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ISS Priority screening for OSAS:
Since OSAS can contribute to road traffic accidents as well as contributing to long term poor health ISS recommends that the following categories of patients be considered for screening for OSAS:
Persons reporting road traffic accidents due to sleepiness
Commercial drivers
Systemic hypertension, particularly if uncontrolled or non-dipping at night
Clear-cut upper airway abnormalities (very large tonsils, significant retrognathia in whom upper airway surgery is being considered)

Neuromuscular disorders (at risk of nocturnal hypoventilation)
COPD
Unexplained Cardiomyopathy/Stroke/Cardiovascular disease
Such screening should initially be based on a detailed history and examination and, if supportive clinical features are present, an overnight sleep study is indicated.
Assessment of daytime sleepiness:
Excessive daytime sleepiness (EDS) is one of the most common symptoms of OSAS, but can also be associated with other medical conditions. In the context of OSAS, **ISS recommends** that the Epworth Sleepiness Scale (ESS) be routinely used to assess the patient’s subjective perception of daytime sleepiness.

The ESS is a subjective questionnaire, which scales the patient’s perception of daytime sleepiness between 0-24. It asks the patient to number appropriately the likelihood of falling asleep in certain daily situations. Some of these situations are very passive, (i.e. lying down, watching TV), others are very active (i.e. driving, conversation).

More objective measures, such as the Multiple Sleep Latency Test (MSLT) can be considered when OSAS has been eliminated. **ISS recommends** validation of the ESS within the Irish population and that further research be undertaken to develop alternative low intensity tools for assessing daytime sleepiness

Diagnosis of OSAS:
To make a diagnosis of OSAS a complete clinical assessment including both history and physical examination should be undertaken, looking in particular at daytime sleepiness and coping strategies for EDS, in addition to an objective recording of nocturnal respiration and other related variables. **ISS recommends** that the clinical assessment should be comprehensive and include the following:

**Clinical Assessment**
Investigations of the patient’s primary reason for presentation to the clinic (e.g. snoring, concern over apnoea or effect of excessive sleepiness

It is highly desirable to also interview the bed partner who can provide important additional information based on direct observation of the patient while asleep

In order to assess the severity of sleepiness, the lifestyle, sleep and work routine: occupation, sleep duration, exercise pattern as well as sleep hygiene factors (shift work/sleep patterns) should be investigated. Patient should fill in the ESS.

Factors such as caffeine/alcohol/cigarette consumption and drug use, all of which contribute to weight gain and poor sleep should be assessed.

Driving safety assessment to include coping strategies at presentation should be assessed. If there is a concern about the subject’s ability to drive, in particular if they drive professionally, then there is an obligation by the Health Care professional to advise the subject to avoid driving and if they must drive then to be aware of the need to pull over and sleep for 20 minutes or to take a caffeinated drink. **ISS recommends** that it is also the duty of the health care professional to ensure that they arrange for investigation and treatment as rapidly as possible.

Past medical history
Family history, especially of OSAS/other sleep disorders/endocrine/cardiac diseases

**Clinical Examination**
Measure of BMI (height and weight)
Neck circumference
Mandible size (qualitative assessment)
Nasal patency
Upper airway and oral examination, including jaw and tongue position when supine
Blood pressure
Cardiac and respiratory examinations

**Diagnostic Criteria for OSAS**
A Task Force of the American Academy of Sleep Physicians, which also involved representatives of other major international sleep and respiratory scientific societies, has developed criteria for the diagnosis of sleep apnoea. According to these criteria, the patient suspected of OSAS
Key Points

- Obstructive sleep apnoea (OSA) is characterised by recurring breathing pauses during sleep, usually due to obstruction at the oropharynx.
- OSA syndrome (OSAS), which combines OSA with relevant clinical features such as daytime sleepiness, has a prevalence of at least 4% in the general adult population, and is twice as common in males as females.
- Although most patients with sleep apnoea snore, only a small proportion of snorers have OSAS.
- OSAS is a major independent risk factor for cardiovascular diseases such as hypertension, ischaemic heart disease, and stroke.
- Driving accidents are up to 10 times more common in patients with OSAS.

Objective Recording of Nocturnal Respiration

The objective assessment of nocturnal respiration during an overnight study must include as a minimum, oro-nasal airflow, respiratory and abdominal effort, and continuous oxygen saturation by pulse oximetry. This type of study is referred to as a limited sleep study or a multi-channel respiratory recording with most proprietary systems available also including clinically relevant variables such as heart rate and rhythm, body position and snoring noise.

The gold standard overnight sleep assessment is Polysomnography (PSG), which incorporates the above signals in conjunction with EEG (electroencephalography), EOG (electrooculography), and EMG (electromyography) to relate the sleep/wake pattern with the respiration signals. PSG is recommended for patients with sleep related conditions such as narcolepsy, and periodic limb movement disorder (PLMD).

Unrefreshed sleep
Daytime fatigue
Impaired concentration

C. Overnight monitoring demonstrates five or more obstructed breathing events during sleep. These events may include any combination of obstructive apnoeas/hypopneas or respiratory effort-related arousals.

ISS recommends that attended in-hospital PSG study is the gold standard. However, PSG set up with manual analysis by adequately trained technical/clinical personnel without full overnight in-hospital supervision is also acceptable.
**Sensors for Airflow and Effort**
ISS supports the new international guidelines (AASM Manual for the Scoring of Sleep and Associated Events Version 2.0.2 August 2013), which recommend that both oronasal thermal sensor and nasal air pressure transducer be used together to assess airflow.

These new guidelines provide literature evidence that supports the sensor best suited to detect absence of airflow for identification of an apnoea is an oronasal thermal sensor and the sensor for identification of a hypopnoea is a nasal air pressure transducer.

In addition, the recent international guidelines and **ISS recommends** that the sensors for detection of respiratory effort should be either based on calibrated or uncalibrated inductance plethysmography, rather than strain gauges, piezo sensors or thoracic impedances devices.

**Home-Based Limited Studies**
Home-based limited sleep studies should be viewed as one option among a range of options for the investigation of a patient with a suspected sleep disorder, which also include full PSG studies in a dedicated sleep laboratory.

Home-based limited studies offer the potential to reduce the demands on sleep laboratories but require careful quality control in terms of patient set-up, quality and range of recordings, and expert analysis by adequately trained technical staff who apply accepted manual scoring rules. The over-arching principle, supported by ISS, is that home-based limited studies must be initiated by an established sleep disorders clinic and that such home-based studies remain under the direct supervision of the clinic concerned.

**ISS recommends** that a full PSG be performed in all patients when the limited study is negative for OSAS in the presence of a high clinical suspicion.

Since the assessment of sleep disorders requires the expert integration of clinical evaluation together with the findings from sleep studies, a physician with training and experience in sleep disorders must be involved in the clinical diagnosis of all patients.

**Analysis and Scoring of Overnight Sleep Studies**
**ISS recommends** that all studies, both PSG and limited, must be manually scored and automated reports should not be accepted for clinical interpretation. **ISS recommends** that The AASM Manual for the Scoring of Sleep and Associated Events along with the Rechtshaffen and Kales (R&K) guidelines be applied to the analysis of sleep staging in PSG studies. The AASM guidelines can also be applied to the manual analysis of all respiratory events recorded during both PSG and limited studies.

**Dual Channel Airflow System**
**ISS endorses** the following rules, based on the latest AASM scoring guidelines (Version 2.0.2 2013) Score an apnoea when all of the following criteria are met

- There is a drop in peak thermal sensor excursion by ≥90% of pre-event baseline using an oronasal thermal sensor (diagnostic study), PAP device flow (titration study) or an alternative apnea sensor (diagnostic study).
- The duration of the ≥90% drop in sensor signal is ≥10 seconds use one of the following (alternative apnoea sensors):
  a. nasal pressure transducer (with or without square root transformation) **RECOMMENDED**
  b. RIPsum (calibrated or uncalibrated) **RECOMMENDED**
  c. RIPflow (calibrated or uncalibrated) **RECOMMENDED**
  d. PVDFsum **ACCEPTABLE**
- Identification of an apnoea does not require a minimum desaturation criterion.
Score an apnoea as **obstructive** if it meets apnoea criteria and is associated with continued or increased inspiratory effort throughout the entire period of absent airflow. **RECOMMENDED**

Score an apnoea as **central** if it meets the apnoea criteria and is associated with absent inspiratory effort throughout the entire period of absent airflow. **RECOMMENDED**

Score an apnoea as **mixed** if it meets the apnoea criteria and is associated with absent inspiratory effort in the initial portion of the event, followed by resumption of inspiratory effort in the second portion of the event. **RECOMMENDED**

For the identification of a **hypopnea** during a diagnostic study, use a nasal pressure transducer (with or without square root transformation of the signal) to monitor airflow. **RECOMMENDED**

When the nasal pressure transducer is not functioning or the signal is not reliable, use one of the following (alternative hypopnea sensors)

a. oronasal thermal airflow **RECOMMENDED**

b. RIPsum (calibrated or uncalibrated) **RECOMMENDED**

c. RIPflow (calibrated or uncalibrated) **RECOMMENDED**

d. dual thoracoabdominal RIP belts (calibrated or uncalibrated) **RECOMMENDED**

e. PVDFsum **ACCEPTABLE**

The AASM continues to recommend scoring **hypopneas** in adults when there is a greater than or equal to 3% oxygen desaturation from pre-event baseline and/or the event is associated with an arousal. However, it is acceptable for accredited sleep centers to score hypopneas in adults when there is a greater than or equal to 4% oxygen desaturation from pre-event baseline.

You must specify in the PSG report whether hypopneas were scored using the recommended rule 1A or the acceptable rule 1B:

**Recommended**

1A. Score a respiratory event as a hypopnea if **ALL of the following criteria are met:**

a. The peak signal excursions drop by greater than or equal to 30% of pre-event baseline using nasal pressure (diagnostic study), PAP device flow (titration study), or an alternative hypopnea sensor (diagnostic study).

b. The duration of the greater than or equal to 30% drop in signal excursion is greater than or equal to 10 seconds.

c. There is a greater than or equal to 3% oxygen desaturation from pre-event baseline and/or the event is associated with an arousal.

**OR**

**Acceptable**

1B. Score a respiratory event as a hypopnea if **ALL of the following criteria are met:**

a. The peak signal excursions drop by greater than or equal to 30% of pre-event baseline using nasal pressure (diagnostic study), PAP device flow (titration study), or an alternative hypopnea sensor (diagnostic study).

b. The duration of the greater than or equal to 30% drop in signal excursion is greater than or equal to 10 seconds.

c. There is a greater than or equal to 4% oxygen desaturation from pre-event baseline.

Please note that the criterion involving arousals is included in 1A and excluded from 1B.

The AASM also reminds members that this clarification is applicable only to the adult scoring criteria for hypopneas. Criteria for scoring hypopneas in children, remain unaffected.

There is now a specific definition for Central vs Obstructive Hypopnoeas.
**ISS strongly recommends** that only one method of manual scoring of apnoeas and hypopnoeas be used and that the selected method is indicated on the final report used for the clinical interpretation.

The Apnoea/Hypopnoea Index (AHI) is defined as the number of apnoeas and hypopnoeas per hour of sleep (PSG) or per hour of study time (limited study).

The grading of severity of OSAS based on the frequency of abnormal respiratory events during sleep:

- **Mild** 5 - < 15
- **Moderate** 15 - 30
- **Severe** > 30

**ISS recommends** the above American Academy of Sleep Medicine (AASM) severity grading criteria for PSG studies, but advise caution when applying these to limited sleep study results.

**Definitions:**

- **PAP**- Positive Airway Pressure
- **RIP**- Respiratory Inductance Plethysmography
- **RIPsum** is the electrical sum of the signals from the thoracic and abdominal RIP sensors, excursions in the signal are an estimate of tidal volume
- **RIPflow** is the time derivative of the RIPsum signal, excursions in the signal are an estimate of airflow
- **PVDF**- Polyvinylidene fluoroide sensor is a fluropolymer substance that reacts to changes in temperature when used as a thermal airflow sensor and to impedance changes when used as an effort sensor
- **PVDFsum**- is the electrical sum of signals recorded from the thoracic and abdominal PVDF sensors

**Key Points**

- Even expert clinical assessment by history and physical examination alone has inadequate power to distinguish OSAS from non-OSAS patients
- Overnight polysomnography remains the gold standard for investigation of OSAS but is expensive and time consuming, however it should be considered in all patients when a limited sleep study is inconclusive
- Cardiorespiratory monitoring without sleep staging is accurate in identifying moderate to severe OSAS patients but may not detect some non-OSAS sleep disorders
- Future technological developments will permit many patients to be investigated at home by means of portable monitoring systems
Treatment of OSAS

OSAS should be considered a chronic disease requiring long-term multidisciplinary management by experienced and trained medical, nursing and scientific/technical specialists. Depending on the symptoms and the severity of OSAS during overnight sleep study, there are a number of treatment options available for OSAS. Positive airway pressure therapy is the mainstay treatment for adults with OSA, though in some patients, particularly those with mild disease, alternative therapies such as oral appliances or in selected cases upper airway surgery may be the preferred option. Please see Sections 2 and 3 for further information on these alternative options.

Behavioral modifications, including weight reduction, alcohol and sedative avoidance, not sleeping in the supine position as well as improving sleep hygiene are also recommended for all patients diagnosed with OSAS.

ISS recommends that the clinical circumstances and the patient’s own preference be involved in the major decision processes for choosing treatment options.

PAP Therapy – Modes of Operation

There are several modalities of PAP treatment available now, fixed CPAP, BPAP, APAP but CPAP remains generally the first-line treatment of choice for patients with OSAS. Research to date does not indicate that either APAP or BPAP are superior to CPAP in the treatment of non complex OSAS, but a trial of APAP and BPAP may be advised for patients who do not tolerate CPAP. Subgroups of patients with coexisting central sleep apnoea or significant hypoventilation may benefit from BPAP or adaptive servo-ventilation (ASV) instead of CPAP.

All PAP devices blow air continuously through either the nose or nose and mouth, via a well fitting mask, which is strapped to the patient’s nose.

When the optimal positive pressure is delivered, PAP will resolve the recurring upper airway collapse and resulting consequences. Therefore whatever modality option is chosen the goal of therapy aims to resolve signs and symptoms of OSAS, improve sleep quality, and normalize both the AHI and oxygen hemoglobin saturation levels.

A large number of patients can obtain substantial clinical improvement, particularly with daytime somnolence, using PAP therapy on a continuous nightly basis. The positive benefits continue for as long the patient adheres to therapy and the optimal positive pressure is maintained.
Optimal Pressure
The optimal pressure in CPAP therapy is defined as the lowest pressure that eliminates different respiratory events in all positions and sleep stages, resulting in normalized sleep architecture. Automatic titrating systems (APAP) are engineered to continuously adjust the pressure to the “optimal” level (4 – 20 cmH20). APAP devices mainly use the flow signals to detect apnoea, hypopnoea, or flow limitation.

Titration is the process of making a trade-off between eliminating all obstructive events by increasing the pressure and reducing side effects by using the lowest possible effective pressure. However, the pressure required to prevent upper airway collapse varies considerably between patients and cannot be predicted from clinical features and/or disease severity. Thus, the initiation of CPAP therapy is complex and requires one or more titration studies to determine the optimum pressure level for each individual patient.

CPAP Titration Study
The Gold standard for CPAP titration is manual titration during a PSG study. However, APAP devices can provide an automatic titration and are most frequently the device of choice when determining a fixed optimal treatment pressure for long term CPAP therapy. The titration study can be performed overnight supervised/unsupervised in-hospital or at the patient’s home. To ensure an optimum choice of pressure is prescribed based on the APAP titration study, ISS recommends that at the least measurement of pulse oximetry, but preferably limited sleep studies are performed at the same time. Most APAP devices are regulated by an internal algorithm that responds on a breath by breath basis to changes in the patient’s airflow. At the end of the study the sum of all the pressures recorded during the night are calculated and a pressure called the 95th centile is displayed on the report. This is usually the recommended fixed pressure for home treatment with CPAP. However, it is vital that the graphic summary of the study is reviewed as well, so that periods of artifact or mask leak are identified and eliminated as these may adversely affect the 95th centile value. Therefore, the prescribed fixed pressure may not always be the same as the 95th one.

APAP devices are not recommended for titration studies for:

• The diagnosis of OSAS
• Use on patients with Congestive Heart Failure, significant lung disease (e.g. COPD), abnormal arterial oxygen saturation due to conditions other than OSA (e.g. obesity hypoventilation syndrome), patients with central sleep apnoea
• Split night studies

CPAP Education and Treatment
ISS strongly recommends that initiation on PAP must include a detailed educational session outlining the risk factors, natural history and consequences of untreated OSA, in particular relating to driving risks. A mask fitting session is also essential during the initiation on PAP treatment and both these sessions should be provided by adequately trained clinical personnel, and ideally should take place in the hospital/sleep laboratory. Group education sessions have been shown to be effective and individual mask fitting sessions allow the patient time to select the best fitting mask and to adapt to the sensation of air pressure delivery from the PAP device. Patients should also be educated about the use, care and maintenance of their PAP device and potential side effects. Although very common side effects are mostly avoidable, especially in the early stages of treatment and patients must be given support and advice on how to manage these. Frequent evaluation during the first few weeks of PAP therapy by telephone or face to face consultations are advised to identify and manage side effects that will negatively affect long-term adherence to treatment. Therefore providing patients with written information and contact help phone
numbers is vital to ensure that patients will continue with treatment, especially during the first 6-8 weeks at home.

**ISS recommends** that PAP therapy is only available upon prescription from a clinician treating respiratory sleep disorders.

All PAP devices and consumables (mask interfaces etc) should be CE marked and compliant with international standards (eg IEC 60601-1 General Requirements for Safety of Medical Electrical Equipment)

**Follow up for Patients on PAP**

Once established on long term PAP therapy, patients require regular follow up to assess efficacy of treatment. Reduction in adherence to therapy will lessen the potential benefits of PAP therapy.

**ISS recommends** that where formal assessment has taken place in hospital with a supervised overnight titration on an APAP device then it is acceptable to perform clinical follow up as an outpatient. This follow up should include an assessment of symptom change, review of objective PAP compliance data, and discussion of side effects.

**ISS recommends** that where the objective diagnosis has been established with limited home-based sleep studies and where APAP has also been performed at home that patients should have a follow-up sleep study to ensure efficacy of treatment, in addition to the clinical assessment of ESS, objective PAP compliance, and discussion of side effects.

If PAP use is showed to be inadequate based on objective monitoring and clinical evaluation, intensive efforts should be made to improve PAP use or consider alterative therapies including possible alternate diagnoses

**PAP Suppliers/Manufacturers**

ISS recognises the important role of PAP manufacturers/suppliers in the provision of equipment and consumables but recommends that this role be limited to technical support. The choice of masks, determination and review of the prescribed pressures, and selection of the PAP modality should remain the primary responsibility of the sleep centre responsible for each individual patient.

**Funding for CPAP Therapy**

**ISS recommends** that the HSE cover the cost of the device rental and mask/headgear in full to patients in receipt of a medical card and that all other patients be covered as part of the drug refund scheme.
References
Clinical Guidelines for the Use of Unattended Portable Monitors in the Diagnosis of Obstructive Sleep Apnoea in Adult Patients: JCSM 2007; 3(7):737-747
ARTP Standards of Care for Sleep Apnoea Services V3.0 April 2010
Recommendations for the Management of Patients with Obstructive Sleep Apnoea and Hypertension. ERS/ESH Taskforce. Eur Respir J 2013;41:523-538

Key Points
• Nasal CPAP is the treatment of choice in most patients with OSAS particularly in moderate to severe OSAS
• CPAP frequently produces dramatic improvements in daytime alertness levels and is associated with major improvements in cardiovascular morbidity and mortality, in addition to reduced accident risk
• Other modalities of PAP including APAP, BPAP and adaptive servo-ventilation are available to treat more complex cases of OSAS or to provide alternative options when CPAP is not tolerated
• Behavioural modifications including weight loss is recommended for all OSAS patients
• A structured education session combining all the relevant components is delivered to patients at initiation of PAP therapy by experienced and trained personnel
• No pharmacological therapy currently available produces clinically relevant improvements in OSAS
• Regular follow-up is recommended by the ISS
Section 2. Dental Oral Appliance Therapy

Definition:
Oral appliance therapy (OAT) involves the fabrication and fitting of dental devices for suitable patients to improve airway patency. European Union legislation for dental devices requires compliance with Medical Devices Directive 93/42/ EC. OAT is a service, which should be prescribed only following medical assessment. Customized dental appliances may be provided for the treatment of mild to moderate sleep apnoea and primary snoring following medical diagnosis of these sleep disordered breathing conditions. Management of these conditions is overseen by the appropriate medical practitioner.

These types of devices adjust the position of the lower jaw to a more anterior and inferior position. This increases the diameter of the pharynx and upper airway reducing the potential for obstruction to develop. These appliances are commonly referred to as mandibular advancement devices (MAD).

Oral appliance therapy (OAT) should only be provided in the absence of dental pathology, oral pathology and jaw dysfunction. The design of the particular oral appliance utilised for an individual patient is determined by a number of anatomical, physiological and psychological variables.

Oral appliances, ideally customised, require a sufficient number of supporting teeth and or dental implants for adequate retention of the device.

Tongue retaining oral devices (TRD) for the edentulous patient have been reported in the literature.

Clinical Protocol
Provision of an oral appliance therapy (OAT) is recommended only after patient consultation with the relevant sleep physician who has responsibility for making the medical diagnosis.

The dentist must receive a referral from the medical practitioner outlining the medical diagnosis and the rationale for consideration of the prescription of an oral appliance to address the patient’s sleep disordered breathing complaint. This referral must be maintained in the patient record.

Literature has demonstrated that oral devices can be considered as first-line treatment for snoring and mild Obstructive Sleep Apnoea (OSA), but they are not recommended for moderate to severe OSA patients. They can however be prescribed in cases where patients do not respond to controlled positive airway pressure (CPAP) therapy or are CPAP intolerant or as an adjunctive parallel therapy in conjunction with CPAP to address sleep disordered breathing conditions.

On referral, a dental evaluation is carried out to determine an individual patient’s suitability for appliance therapy. Customisation of appliances is necessary to meet specific anatomical and treatment requirements.

The benefits and risks of oral appliance therapy (OAT) should be clearly explained to the patient and consented to prior to the initiation of therapy.

Dental Assessment
ISS recommends that a full patient history and clinical examination, including, complete extra-oral and intra-oral examination be required to determine overall patient suitability and the absence of pathology or significant anatomical or physiological anomalies in the stomatognathic system.

Patient Examination should involve review and documentation of the following parameters:

Oral or dental pathology
State of the dentition: Edentulous, Partially Edentulous or Fully Dentate
Jaw Function evaluation to include, line of movement, range of movement and muscle palpation for evidence of spasm or dyskinesia.

Temporomandibular joint anomalies

Gag reflex

Static and dynamic occlusal evaluation of the dental arches to include overbite and overjet, tooth position and tooth mobility.

Evidence of tooth wear due to parafunctional habits.

Evidence of erosion due to perimolysis.

Tongue size anomalies

Throat form including Mallampati score.

Angle Classification of skeletal jaw form.

Quality and quantity of saliva and any dry-mouth findings.

Psychological parameters that may make the patient unsuitable to commence OAT.

**Additional special tests:**

Trial advancement bite registration test.

Mounted casts of upper and lower arches.

Orthopantomograph (OPG) and standard periapical radiographs where necessary.

Referral for medical opinion and investigation in cases of perimolysis.

**Types of Appliances**

There are two main categories of oral appliances for the dentate or partially edentulous patient. Both categories aim to protrude and depress the mandible.

The “Monobloc” appliance category is a one-part bimaxillary appliance.

The “Titratable” category is a two-part adjustable bimaxillary appliance.

Depending on the clinical diagnosis an appliance may be prescribed on a provisional basis as an interim or diagnostic device, or alternately on a definitive long-term basis. Liaison with dental laboratory professionals to optimise the provision of mandibular advancement appliance services is essential whether provisional or definitive in nature.

**Follow-up:**

The dentist should communicate the proposed treatment plan for OAT with the referring physician and other appropriate healthcare providers and provide regular updates on the progress and follow-up as well as any other pertinent information.

**ISS recommends** that after the dental device has been fitted, the dentist must conduct a follow-up evaluation of treatment.
in terms of appliance efficacy and tolerance. This evaluation should include:

Providing the patient with knowledge of the most commonly experienced side effects and their appropriate management.

Providing the patient with information on oral appliance care and maintenance considerations.

Ongoing monitoring of appliance acceptance and wear compliance by the patient at specified interval of three months to a year.

Recall examinations at least annually, which should include a review of both occlusal stability and the structural integrity of the device and also screen for possible side effects.

Comparison with baseline parameters such as temporo-mandibular joint function, occlusion, gingival health, tooth mobility.

Subjective evaluation of efficacy as provided from bed-partners response where appropriate, and post treatment Epworth Sleepiness Scale.

Reinforcing of advice to the patient in written form.

Scheduling of the return visit to the referring physician so that objective efficacy testing and follow-up through either polysomnography or limited sleep studies as deemed appropriate by the relevant sleep physician can be completed.

Evaluation and documentation of the treatment planning decisions, outcomes, and any subsequent modifications, which may arise following the delivery of these appliances, should be communicated to the other medical professionals involved in patient management.

References
Pathogenesis of obstructive and central sleep apnoea. White et al.

Section 3. Surgery for Snoring / OSA

3a. Otolaryngological (ENT) Surgical Guidelines

The evidence to date in the literature does not support the widespread use of surgical interventions in the management of unselected patients with OSAS (Sundaram et al, 2005). However, a variety of surgical options are available for carefully selected patients including tonsillectomy, adenoidectomy, septoplasty, turbinate reduction and removal of nasal polyps. Tonsillectomy in patients with grossly enlarged tonsils frequently results in major improvements in OSAS severity but the degree of benefit varies depending on the presence of other contributing factors such as obesity. Surgical relief of nasal obstruction, such as correction of deviated nasal septum, provides inconsistent benefit in reducing OSAS severity but the resulting improvements in nasal airway patency can improve the efficacy and tolerance of CPAP.

Other surgical procedures include: uvulopalatopharyngoplasty (UPPP), genioglossus advancement, radiofrequency ablation, and mandibular osteotomy. However these surgical options are only of potential value in carefully selected patients and procedures such as maxillo/mandibular osteotomy are complex and only suited to highly specialised centres. UPPP procedures can be performed to different degrees ranging from conventional surgical resection of the posterior soft palate and surrounding redundant tissues (UP3) to limited resection of the posterior palate (UP2), often by a laser-assisted technique (LAUP). However, the evidence of efficacy for UPPP is inconsistent, particularly for LAUP, and at best indicates a less than 50% long-term success rate. Furthermore, LAUP is a very painful procedure and UPPP can be complicated by nasopharyngeal reflux. A recently introduced limited procedure to stiffen the soft palate, the Pillar procedure, is relatively simple and non-invasive, but efficacy is not yet proven.

ISS recommends that a multidisciplinary approach be adopted in the assessment of OSAS patients being considered for surgical intervention and that surgery be performed only in carefully selected patients. Upper airway surgery is rarely appropriate in patients with moderate or severe OSAS unless there is a clearly identifiable obstructing lesion such as enlarged tonsils. ISS also recommends that randomised and controlled studies be performed to accurately evaluate ENT surgical outcomes.

Role of the ENT specialist in Screening Patients for Snoring/OSAS/other sleep disorders:

ISS recommends that the ENT surgeon’s role in screening patients who present for ENT surgery for relief of snoring or suspected OSAS should include a thorough history and examination in addition to appropriate investigations:

Clinical Assessment:
Epworth Sleepiness Scale
Snoring Symptoms Inventory
Partner VAS / Partner Questionnaire
Body Mass Index & neck circumference
Nasal obstruction
Oral cavity & Oropharynx
Freidman tonsil score
Mallampati score of pharyngeal congestion

Sleep Nasendoscopy can help to identify the level of obstruction i.e. soft palate, tongue base or larynx. Croft and Pringle first described this procedure in 1991. The patient is sedated until snoring is achieved and the procedure allows visualisation of the vibrating structures and also the site & extent of upper airway collapse. A classification system was developed in 1993 and was further modified in 1995. However, it is unlikely that sedation-induced sleep correlates well with natural sleep and there is currently no standardised sedation protocol; thus the true efficacy of this procedure is still debated.
**Clinical Investigations, where available:**
- Sleep studies - may be limited / ambulatory in those suspected of non-apnoeic snoring
- Nasopharyngoscopy / Mueller Manoeuvre
- Acoustic Rhinomanometry
- Sleep Nasendoscopy
- Acoustic Analysis / Snore Sound Characteristics
- Radiological investigations – Cephalometry, Somnofluoroscopy, CT, MRI

OSAS patients undergoing surgery will require extra care during recovery and where possible they should be encouraged to use their CPAP device.

**Cephalometric Analysis**

Patients with OSA have smaller upper airways than normals (Rivlin 1984)

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### 3b. Bariatric Surgery

Obesity is a major risk factor for OSAS and weight loss is associated with considerable reduction in OSAS severity in obese patients. Voluntary weight reduction is often unsuccessful in obese OSAS patients and the reasons are complex and incompletely understood.

Bariatric surgery represents a potential management option for OSAS in severely obese patients who fail to lose sufficient weight by conservative means and who are intolerant or unwilling to comply with long-term CPAP therapy. A variety of surgical techniques are available including gastric banding, gastric bypass, and gastroplasty. Systematic review of the literature regarding efficacy of bariatric surgery in alleviating OSAS indicates that most such patients experience substantial weight reduction associated with either resolution or substantial improvement in OSAS as measured by AHI. However, bariatric surgery can be associated with complications and a post-operative mortality of up to 1% has been reported in a systematic review. Furthermore, such patients are at increased anaesthetic risk because of the co-existence of OSAS with obesity, particularly in the recovery period from anaesthesia.

**ISS recommends** that bariatric surgery in obese OSAS patients be performed only in specialized centres with experience and expertise in the surgical and anaesthetic management of such patients.

### References


**Section 4. Restless Leg Syndrome & Periodic Leg Movement Disorder**

**Definition**

Restless Leg Syndrome (RLS) is a complex lifelong sensorimotor disorder. Patients experience an intense, disagreeable creeping sensation in the lower extremities, especially in the evenings, which is relieved by moving the legs. There are both primary and secondary forms of RLS.

Periodic Leg Movement Disorder (PLMD) is characterised by repetitive leg movements during non-REM sleep, most commonly the extension of big toe, dorsiflexion of ankle, or knee, or hip every 20-40 seconds. PLMs may cause EEG arousals, which reduce sleep quality and results in excessive daytime sleepiness. PLMs are noted in at least 80% patients with RLS. As a result the occurrence of both waking and sleeping periodic leg movements PLMD is now recognized as opposed to pure sleep associated PLMs.

**Primary and secondary forms of RLS & PLMD**

The same constellation of symptoms may occur in the setting of several disorders and physiologic changes, notably iron deficient anaemia, pregnancy and polyneuropathy. This has led to the term “secondary” RLS. A positive family history is a defining, but not mandatory, feature of the primary form of RLS.

<table>
<thead>
<tr>
<th>Primary form</th>
<th>“Secondary” form</th>
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<tbody>
<tr>
<td>Positive family history</td>
<td>Peripheral neuropathy</td>
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<tr>
<td></td>
<td>Uraemia with anemia</td>
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<td>Iron deficiency anaemia</td>
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<td>Pregnancy</td>
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<td>Thyroid drugs, myelopathy, varicose veins</td>
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**Epidemiology and aetiology**

In population based studies the prevalence for RLS ranges from 0.6% to 15%. A recent study found a 27% prevalence of RLS in pregnant females. In 17% they were limited to the duration of pregnancy, reflecting the role of iron deficiency. Abnormal traffic of iron and related changes in dopamine receptor function are major aetiological factors in primary RLS. Iron deficiency should be treated. PLMD can occur at any age, more frequently in the elderly.

**Differential diagnosis**

- Peripheral neuropathy
- Lumbosacral radiculopathy
- Hypnogenic myoclonus (sleep starts)
- Nocturnal leg cramps
- Venous/arterial insufficiency
- “Painful legs and moving toes syndrome”
- “Growing pains”
- Akathisia

*PSG recording showing bursts of repetitive PLMs during NREM sleep*
Diagnosis of RLS & PLMD
The clinical history and otherwise normal examination are generally sufficient to make a diagnosis of RLS. But objective measures can be used also. Actigraphy measures the surrogate marker PLM and is extensively used in therapeutic research. Polysomnography can measure leg PLM activity overnight during a sleep study recording. ISS recommend that bilateral leg EMG should be measured if PSG is used to make a diagnosis of RLS, but objective measures may also be used.
Carry out FBC, creatinine, glucose, ferritin, folate, B12, to check for iron deficiency, uraemia.

Diagnostic criteria based on clinical assessment
The urge to move legs, with unpleasant sensations (legs)
An increase or onset of symptoms with rest or inactivity
Decreased symptoms on movement, eg, stretching
An increase in symptoms during the evening and night
A variable course of symptoms
A normal physical examination in idiopathic RLS
Complaint of sleep disturbance

Supportive clinical features
A response to dopaminergic therapy
Objective evidence of periodic leg movements during wakefulness or sleep

Therapeutic guidelines.
The available agents have been reviewed and rated by the RLS Task Force 2004 Standards of Practice Committee of the American Association of Sleep Medicine (Hening W, et al. RLS Task Force 2004 Standards of Practice AASM Sleep 2004;27:560-83). Newer agents have extended the armamentarium, most recently rotigotine.

Overall management
Provide education and support to patients diagnosed.
Limit therapy to patients with sleep disturbances and consider age, severity, and motivation for treatment.
Treat anaemia if present
Use dopamine agonist for any stage of RLS — mild, moderate, and severe

Problems with l-dopa
Loss of efficacy over time.
Rebound phenomenon: Recurrence of symptoms later in the night or in the early morning after the initial response.
Augmentation: Symptoms earlier in the day, with an increased intensity and extension to other regions
82% of patients on l-dopa develop augmentation
These problems may relate to the difficulty in achieving steady state plasma levels without the need for frequent dosages. The elimination half-life (EHL) for l-dopa is approximately one hour, increasing two – threefold with the addition of carbidopa and entacapone (Stalevo combines all three).

Dopamine agonists (DA)
Theoretical advantages: Reduced tendency to rebound and augmentation, with longer EHL. The availability of sustained release formulations may enhance this capacity of the DA to achieve steady state plasma levels, eg, the new transdermal preparation of rotigotine (Neupro). Several have obtained Irish Medicines Board (IMB+) approval of their indication for RLS, including pramipexole and rotigotine, but the full array are in general use and may have to be deployed when problems of diminished efficacy are successively encountered.
Pramipexole (Mirapexin) (IMB+)
0.75 - 1.5 mg (salt weight) 1 hr before bed; EHL 8 hr
98 % decrease in PLMS
84% decrease in RLS
Total sleep time, number of awakenings and sleep efficiency, unchanged
REM suppressed
Unwanted effects (UE): Nausea, fatigue, dose related
Augmentation 8.5 – 18%

Ropinirole (Adartrel) (IMB+)
1 – 4 mg single daily dose, building from 0.25 mg; EHL 6 hr.
This agent is more commonly marketed as Requip for Parkinson’s disease, also available in a once daily sustained release preparation (Requip Modutab). Multiple studies show improved RLS and associated sleep measures.
UE: Nausea, somnolence.

Rotigotine (Neupro) (IMB+)
Transdermal patch
1 - 3 mg/24 hr; EHL 7 hr after discontinuation
Clinical remission in 47.3% vs 22.8% placebo
UE: nausea, application site reactions, fatigue and headache.

Bromocriptine (Parlodel)
2.5 – 5.0 mg daily; EHL 12-14 hr
UE: Nausea, hallucination.

Cabergoline (Cabaser)
1 – 4 mg daily
Long EHL: 65 hr
UE: Pleural effusion / fibrosis, ergot side effects, including hallucination

Pergolide (Celance)
0.5 mg 2 hr before bed; EHL 27 hr
Reduced PLMS with arousal (2.3 / 8.9), RLS decreased
No change in total arousals, or REM, but sleep time increased
UE: Nausea, headache, rhinitis, vomiting, abdominal pain, dizziness
Augmentation 15 – 27%. Fibrosing endocardial valvulopathy has emerged as a significant risk and requires regular monitoring of inflammatory markers and echocardiography.

Pharmacological treatment: sequential trials
First-line pharmacological treatment
L-dopa
Combined with enzyme inhibitors
(+ benzerazide = Madopar; + carbidopa = Sinemet; carbidopa + entacapone = Stalevo).
Anticipate early augmentation and rebound
Typically recommended only for mild symptoms

Dopamine agonists
Use in ascending order of toxicity; pergolide requires special caution (see above)
Ergot derivatives generally more toxic (cabergoline, bromocriptine, pergolide)
Second-line pharmacological treatments
Benzodiazepines, opioids, anticonvulsants
Add clonazepam as needed for sleep - watch for exacerbation of OSAS
Empiric iron therapy not justified unless ferritin <50mcg/l
**Key Points**

- RLS is a complex lifelong sensorimotor disorder
- PLM are repetitive leg movements during Non-REM sleep
- PLM’s are noted in 80% patients with RLS
- Clinical history is usually sufficient to diagnose RLS
- Objective measurements (PLM during PSG sleep study) required to diagnose PLM
- Provide patient education and support upon diagnosis
- Treat anaemia if present
- Limited pharmacological treatment to patients with sleep disturbances, but consider age, severity and motivation to therapy as well
**Definition**

Insomnia is the most prevalent sleep disorder in the general population. Classification systems exist but no single system is agreed in common practice. The diagnosis of insomnia is based on subjective complaints of falling asleep, staying asleep, early morning awakenings or non-restorative sleep. These problems occur despite adequate opportunity and circumstances for sleep.

At least one of the following daytime impairments associated with insomnia should be reported.

1. Fatigue
2. Attention, concentration or memory impairment.
3. Social dysfunction or school performance.
4. Mood disturbance or irritability.
5. Daytime sleepiness.
6. Motivation, energy or initiative reduction
7. Prone to errors/accidents at work or while driving.
8. Tension, headaches or gastrointestinal symptoms in response to sleep loss.
9. Concerns or worries about sleep.

Identifying daytime dysfunction is essential to rule out “short sleepers” who have reduced sleep times in comparison to the general population but function well without daytime consequence.

Transient or Adjustment Insomnia is usually related to life circumstances and is ubiquitous. Adjustment insomnia can be broken down into Acute Insomnia (lasting up to one week) or Sub-acute Insomnia (lasting from one week to 3 months). This usually settles in response to reduction or adaptation to the stressor (e.g., moving house, short term illness, time zone shift). However, insomnia often becomes a chronic illness with up to 80% of insomnia cases, lasting greater than 3 months.

Sufferers develop heightened levels of anxiety about their sleep disturbance and daytime consequences, as well as conditioned negative anticipation of night time sleep; which leads to increasing frustration with sleep and sleep effort. This forms the basis of chronic/conditioned insomnia referred to as psycho-physiological insomnia, emphasizing the importance of early identification and intervention to address precipitating factors, and maladaptive responses to them.

**Epidemiology:**

Insomnia affects millions of individuals worldwide and is the most commonly reported sleep problem in industrialized nations around the world. It is reported that insomnia occurs in one third of the general population. While estimates vary, approximately 10% -15% of the general population with current complaints of insomnia, will be classified as suffering a moderate to severe disorder. 5% of primary care patients seek treatment for their insomnia. Recognition of the problem is poor and unfortunately to date, physician training in this area remains minimal. Insomnia is more common among women, middle-aged and older adults, shift workers, and patients with medical or psychiatric disorders and possibly unemployment and lower socio-economic status.

Women are consistently more likely to have insomnia than men; pregnancy and menopause are frequent triggers.
Increased age, marital separation and divorced or widowed status, are population risk factors. The higher prevalence rates in women, increases with age. The elderly pattern of early bedtimes, sleep fragmentation at night, and daytime napping is not a form of pathological insomnia but a common focus of complaint. Patients with co morbid medical and psychiatric conditions are at particularly increased risk, with psychiatric and chronic pain disorders having insomnia rates as high as 50% to 75%.

Consequences of Insomnia:
Insomnia is associated with an impaired quality of life, daytime psycho-motor impairment, heart disease, immune dysfunction, and several endocrine and metabolic derangements. Insomnia is also associated with difficulties and accidents at work as well as more absenteeism. Chronic insomnia confers an important three fold increased risk of suicide attempt, and sleep must be targeted in the treatment of acute depression. It is also a risk factor for alcohol relapse in patients recovering from alcohol abuse.

Assessment of Insomnia:
Screening for insomnia is indicated in routine health examinations. Assessing a patient with insomnia requires a detailed exploration of their medical, psychiatric and psychological history. ISS recommends that full physical and mental state examinations along with a full sleep history are required when there is a complaint of chronic insomnia. Detailed screening for associated co-morbid psychiatric conditions is of proven value. Insomnia may be co-morbid with a number of psychiatric conditions, mood disorders, anxiety disorders including nocturnal panic, or a history of drug or alcohol use. Psychiatric disorders are highly prevalent amongst patients with insomnia suggesting that underlying psychiatric illness plays a large role in the development and resistance of insomnia. There is also an increased risk of developing new psychiatric illness following the onset of insomnia. It is an independent risk factor for development of depression and an increased risk for its recurrence and chronic course.

More extended history taking includes history of substance abuse, social history and sexual history (such as sexual abuse). There are numerous sleep questionnaires to assess sleep quality, duration, etc. It is important to screen for nocturnal symptoms which can cause or exacerbate co-existing insomnia.

Snoring or apnoeas (to suggest Obstructive Sleep Apnoea), respiratory distress, leg jerks or kicks (Restless Leg Syndrome / Periodic Limb Movement Disorder), Nocturia (OSA or urological issues), gastro-oesophageal reflux disease, pain, headaches and so forth, all of which are relevant and perpetuate sleep disturbance. Other illnesses which may well pose relevant to the insomnia include Dementia, Hyperthyroidism, or Parkinson’s disease.

Certain prescription drugs which can cause insomnia including Theophylline, antidepressants, Attention Deficit Disorder drugs, Clonidine, Propranolol, Atenolol, Albuterol, Salmeterol, Methyldopa, Levodopa, Quinidine, Corticosteroids, oral contraceptives, Progesterone, Thyroxine, Phenytoin, amphetamines, decongestants, weight loss products, and pain relievers containing caffeine.

Recreational drugs / drugs of abuse and misuse can also trigger insomnia; common offenders include alcohol, caffeine, tobacco, ecstasy, cocaine and amphetamines. Alcohol and drug use are a leading cause of insomnia, especially in older patients.

It is well documented that poor sleepers tend to overestimate their degree of sleep disturbance. There may be large discrepancies between their perceived degree of tiredness / sleepiness and objective ratings through questionnaires as well as large discrepancies between reported sleep duration and objective findings during polysomnography or actigraphy.

Paradoxical Insomnia or Sleep State Misperception (a later addition to the International Classification of Sleep
Disorders ICSD) defines a discrepancy between perceived and actual sleep times. Although their sleep times may be normal, their complaints of daytime consequences match those of other insomnia patients, indicating the possible effect of hyper-arousal or psychological state on perception of sleep and daytime functioning.

**Diagnostic tools**

On reviewing the current diagnostic practices to clinically evaluate insomnia, sleep diaries are useful. Ask patients to record their bedtime, total sleep time, time it took to fall asleep, the number of times awakening at night, use of sleep medications, time out of bed in the morning, and a rating of subjective sleep quality and daytime symptoms, over a 2-week period. Various sample sleep diaries are available on the internet for printing. Collateral history from a bed partner is often valuable but not always confirmatory. Additional psycho-physiological measures (other than taking a history regarding constitutional heightened arousal levels) although useful, are mostly unavailable.

Polysomnographic data / sleep studies or Multiple Sleep Latency Tests, to define insomnia are not applicable in the routine outpatient diagnostic setting. Polysomnography is not routinely indicated in the diagnostic work-up and treatment of insomnia; however it is indicated in cases where sleep-disordered breathing, Narcolepsy, Nocturnal Movement Disorder (such as Periodic Limb Movement Disorder) or Parasomnia are suspected.

Actigraphy (a small wrist-watch device which tracks sleep patterns over time) is not routinely indicated for diagnosing insomnia but it can prove a useful tool for some patients specifically where suspected Paradoxical Insomnia is present (to provide objective evidence of sleep duration and sleep pattern) or to rule out an underlying circadian rhythm disorders such as the commonly occurring - Delayed Sleep Phase Syndrome.

Two major diagnostic systems for sleep disorders are currently in use. The International Classification of Sleep Disorders nosology includes approximately 42 diagnoses which may be associated with an insomnia complaint. The Diagnostic and Statistical Manual of Mental Disorders, Fourth Edition, although less detailed than ICSD, maintains the essential division between primary dyssomnias, including insomnia, and those disorders secondary to other psychiatric or medical conditions.

**Treatment for Insomnia:**

Treatment for all patients should include education on sleep hygiene. Sleep hygiene instruction alone however, is not evidenced as effective but it is essential in conjunction with other measure.

Correction or alleviation of identifiable underlying causes of insomnia is essential where possible, such as alleviating underlying pain, depression, anxiety disorders, and physical symptoms (such as wheeze, dyspnoea, nocturia etc).

Treatment for insomnia is largely divided into pharmaceutical treatment and cognitive behavioral therapy. Medication is most commonly used to treat insomnia and has a value in short term use. However it is both frequently inappropriately withheld or overused. Many patients will cite minimal relief from hypnotic agents yet they will continue to take them on a regular basis for years. Prescribing sedative hypnotics in older patients is of particular concern given the frequency of “poly-pharmacy” and drug interactions as well as delayed metabolism effects, fall risk and aggravation of cognitive difficulties. Rebound insomnia with the use of short acting hypnotics may perpetuate the sleep disturbance. It is also worth noting that hypnotics should not be used in patients with suspected (or untreated) sleep-disordered breathing as it can exacerbate their respiratory compromise. Patients and physicians alike are expressing increasing concern surrounding emerging data about the long-term risks associated with hypnotics (and benzodiazepines).
Behavioral therapies are often not adequately tried but are as, or more effective in chronic insomnia, and confer longer lasting improvement. In the intermediate term (i.e. 3-8 weeks), meta-analysis indicates that behavioral treatment for insomnia is just as effective as medication treatment. Over the long term (i.e. 6-24 months), patients receiving non-pharmacological therapies enjoy long lasting relief while many of those treated with medication return to their baseline insomnia levels. Unfortunately the availability of access to Behavioral Therapy for insomnia is very limited. There are educational books and online resources available which may prove helpful to patients in the absence of an expert provider.

Sleep hygiene:
Patients (and the general population alike) should be educated on healthy sleep behaviours and preferable environmental conditions such as the light, noise, temperature and bedroom environment. Lifestyle factors include exercise, diet and use of caffeine, alcohol and nicotine. Information about normal sleep patterns and individual differences in sleep needs and changes in sleep physiology throughout our lifetime should also be included. Sleep hygiene is included with other interventions as opposed to being used as a primary intervention.

Treatment – Behavioural therapies for primary insomnia

Cognitive behavioural therapy:
Insomnia often arises from psychological factors such as conditioned arousal, maladaptive sleep habits and sleep schedules, dysfunctional thinking about sleep and its consequences, and sleep preoccupation. (Randomized Clinical Effectiveness Trial of Nurse – Administered Small Group Cognitive Behavior Therapy for Persistent Insomnia in General Practice. Sleep, Vol.30, No 5. 2007). The main objective of CBT is to alter these factors that perpetuate or exacerbate sleep disturbances. Another objective of CBT is to teach individuals self-management skills to cope with their sleep disturbances.

Relaxation therapy:
These techniques are used to reduce somatic tension and intrusive thoughts interfering with sleep. They include progressive muscle relaxation which focuses on reducing physiological arousal, imagery training helps with the elimination of intrusive thoughts and reduces worry. Other techniques used include autogenic training and bio-feedback. Pre-bedtime wind-down routines are also an important tool in helping to eliminate anxiety at bedtime.

Stimulus Control Therapy:
Consists of a set of instructions designed to strengthen the connection/association between the bed and sleep, in order to re-establish a consistent sleep wake schedule.
These instructions include:
1. Go to bed only when sleepy.
2. No reading, watching TV, listening to radio or using phones or smart devices in bed.
3. Get up time should be consistent 7 days a week regardless of the amount of sleep the night before.
4. 15 Minute Rule: The individual should be instructed to get out of the bed if he/she is awake for 15 minutes and go to another part of the house (not another bedroom). He/she should only return to bed when sleepy. This should be repeated whenever the individual has been awake in bed for 15 minutes.
5. Napping should be avoided.

Sleep restriction therapy:
This reduces nocturnal sleep disturbances by restricting the time allotted for sleep each night so that eventually the time spent in bed closely matches the individuals presumed sleep requirement. Time In bed (TIB) prescription is calculated from sleep logs and actigraphy over a period of some weeks. Initial TIB prescription is
not set below 5 hours per night. Once sleep efficiency (SE) has reached 90% TIB can then be gradually increased (15 minutes per week) over a period of weeks until optimal sleep is achieved. Caution is needed when using sleep restriction and individuals should be warned on the dangers of driving and operating heavy equipment.

**Pharmacological Treatment for Insomnia:**
Let us review the general principles. Patients should be advised when treatment is started that it will be of limited duration (between 2 and 4 weeks) at the lowest effective dose. Prescribers should explain that when the dosage is progressively decreased or stopped, the likelihood of transient rebound or persisting insomnia exists; and is not an indication for continuance. If medication use is to be extended, it should be for documented with proportionate reasons. If use is extended, medication should be taken discontinuously (drug free intervals) wherever possible. Long term use of hypnotics is not well supported by trial data, and tolerance is commonly reported, so their continuous use offends good prescribing principles.

Patients who have difficulty in falling asleep can be treated effectively with short or intermediate-acting hypnotic. Patients experiencing periods of wakefulness during the night, or early awakening, need a hypnotic with an intermediate half-life. Patients should be discouraged from taking hypnotics during the night should they awaken (this reinforces awakenings). Long-acting hypnotics, in general should not be prescribed to ensure there is no sedative hangover during the day.

For patients dependent on hypnotics, especially the elderly, careful evaluation of risk benefit ratios to stoppage should be undertaken as well as careful assessment of withdrawal risk, before being stopped.

Abusive use of hypnotics warrants discontinuation with adjunctive addiction counseling. All stoppages should follow tapering regimens to circumvent withdrawal reactions.

Discussing each and every hypnotic agent’s application is beyond the scope of these guidelines. In general the newer non-benzodiazepines (so called “Z” drugs) are preferable to more traditional benzodiazepines. The three primary groups of Z-drugs are Imidazopyridines (zolpidem), Cyclopyrrolones (zopiclone) and Pyrazolopyrimidines (zaleplon) They boast advantages over benzodiazepines (which actually worsen sleep architecture), whereas the Z-drugs may have less or no disruption to sleep architecture and have fewer propensities to daytime sedation, cognitive impairment, dependence and rebound insomnia. They are however, occasionally abused.

**Prescribed agents also used but not licensed as hypnotics:**
Sedative antidepressants in low doses are in common use but with the qualified exception of Trazodone, are poorly supported by data and carry significant side effect burdens. There have been several studies reporting Trazodone as being effective in treating insomnia for patients on SSRIs or SNRIs. Antihistamines are not recommended due to daytime sedation despite widespread use. Sedative neuroleptics are only appropriate when secondary indications apply.

**Over the counter insomnia treatments:**
The rationale for choosing OTC sleep aids includes ready availability, low cost, and favorable perceptions of safety or naturalness. Antihistamine based compounds are commonly used, however because of changes in sleep architecture, notably a reduction in REM sleep caused by their anti-cholinergic effects; they can result in a reduction in cognitive function, day time sedation, increased risk of accidents, development of tolerance, and interference with other medications. As such they are not generally recommended beyond a limited term use by ISS. Certain anti-histamines due to their anticholinergic effect may also worsen Restless Leg symptoms.
As robust data does not exist to support the hypnotic effect and safety of acute treatment of herbal extracts of Valerian on patients suffering insomnia and as paradoxical reactions and hepatotoxicity have been described, ISS do not recommend this medication.

**A note on Melatonin:**
Melatonin was licensed for use in Ireland in 2009. It is available on prescription in prolonged-release form at a 2 mg dose for the short term treatment of primary insomnia in patients over 55 years old. Although its side effect profile is favorable, it has limited utility in patients with Psycho-physiological Insomnia, and is more efficacious when applied to treating patients with complaints of Circadian Rhythm Disorders, such as Delayed Sleep Phase Syndrome, Shift-work Sleep Disorder, and travelers experiencing jet lag. It can also prove beneficial in Hospitalized or institutionalized patients, where resultant change in environment and loss of traditional zeitgebers (natural daylight, schedules, alarm clock) can lead to dysregulation of the sleep-wake cycle and subsequent insomnia.

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### Key Points

- **Adjustment (short term) insomnia is common during life transitions and stress**
- **A full physical and mental state examination along with a full sleep history is required for longer term insomnia**
- **Detailed screening for associated co-morbid psychiatric conditions, mood disorders, anxiety disorders or a history of drug or alcohol use, are of proven value**
- **Chronic insomnia is most frequently secondary to or co-morbid with medical and psychiatric conditions**
- **Education on good practices around sleep is an essential part of all insomnia treatment. Correction or alleviation of underlying causes of secondary chronic insomnia is also necessary**
- **Medication has a value in treating short term insomnia. The choice of agent should be guided by the most effective agent at the lowest dose, for the least unbroken period. Prescribers should explain transient rebound is not an indication for continuance**
- **Long term use with these agents is not recommended but stoppage in persistent users should be carefully evaluated and managed**
- **Long term primary insomnia can be managed by Cognitive Behavioral Therapy with methods including sleep restriction, stimulus control and relaxation techniques.**
References


13. An American Academy of Sleep Medicine Review. Michael J. Sateia, MD1, Karl Doghramji, MD2, Peter J. Hauri, PhD3, Charles M. Morin, PhD. 1999

Parasomnias are undesirable physical events or experiences that occur during entry into sleep, within sleep or during arousal from sleep. Parasomnias may occur during non-rapid eye movement sleep (REM), or during transitions to and from sleep. They are encountered more frequently in children and typically resolve by puberty but may persist (or arise de novo) in adolescence or adulthood. Patients may experience more than one type of parasomnia.

According to the International Classification of Sleep Disorders (ICSD 3) they are divided into:

1. Arousal disorders - sleepwalking, sleep terrors, sleep related eating disorder and confusional arousals. Sleep related abnormal sexual behaviours are primarily classified as confusional arousal and are a subtype.
2. Parasomnias associated with REM sleep – including nightmare disorder, recurrent isolated sleep paralysis and REM sleep behaviour disorder (RBD)
3. Other parasomnias - including exploding head syndrome, sleep eneuresis, sleep related hallucinations, parasomnia due to a medical disorder and parasomnia due to a medication or substance.

**Background**

The non-REM sleep group of arousal disorders is characterized by a sudden and incomplete arousal from deep or slow wave sleep (SWS) either in the first or in subsequent sleep cycles. The arousal is associated with a dissociated reaction between cortical activity and an elaborated motor activity in sleep walking and an autonomic discharge in night terrors. The increase in the depth of sleep, assessed by EEG, during the preceding minutes may partially explain this dissociation. The deep sleep of patients who sleep walk and have night terrors is unusually fragmented by frequent brief arousals and this can be seen during polysomnography even in the absence of major events.

Disorders of arousal share the following characteristics:

1. Similar genetic and familial patterns
2. Similar pathophysiology of partial arousals from deep sleep
3. Similar priming by sleep deprivation and biopsychosocial stressors
4. Not secondary to psychiatric disorders
5. Not generally secondary to neuropathology or head injury
6. Associated with absent or minimal effect on cognitive functioning
7. Associated with amnesia for the event
8. May be triggered by sound, touch or other stimuli.

Animal experiments have revealed that in RBD, in addition to a lack of typical muscle atonia in REM sleep there is a disinhibition of motor activity in the cortex. In patients with this disorder, the EMG tone on
polysomnography remains elevated and there may be excessive jerking. Obstructive sleep apnea syndrome is increasingly seen as a precipitant of parasomnias in both adults and children. Travel, sleeping in unfamiliar surroundings febrile states in children, stress, premenstrual period in women and certain psychotropic drugs such as lithium and hypnotic / sedative agents have been implicated.

Sleep walking and sexsomnia as a defence are being raised more frequently in criminal cases.

**Epidemiology**

There are no data available estimating the prevalence of parasomnias in the Irish population and international data vary. Arousal parasomnias are common in childhood but normally fade out in adolescence.

**Prevalence in Adults**

<table>
<thead>
<tr>
<th>Condition</th>
<th>Prevalence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sleep walking (&gt; 1/month)</td>
<td>0.5 – 3%</td>
</tr>
<tr>
<td>Night terrors</td>
<td>2.2%</td>
</tr>
<tr>
<td>Confusional arousals</td>
<td>2.9 – 4.2%</td>
</tr>
<tr>
<td>Sleep talking (frequently)</td>
<td>3%</td>
</tr>
<tr>
<td>Nightmares</td>
<td>2-8%</td>
</tr>
<tr>
<td>Sleep paralysis</td>
<td>6%</td>
</tr>
<tr>
<td>REM sleep behaviour disorder</td>
<td>0.38 – 0.5%</td>
</tr>
</tbody>
</table>

**Symptoms of Main Conditions**

**Sleep walking**

Family history is frequent.
Involves partial arousal from deep sleep.
Usually occurs in the first 1/3 of the night.
Lasts 1 – 5 minutes but may be longer.
Communication is difficult or impossible but subject may sleep talk or shout.
Behaviour can be simple and non-goal directed or complex and may have symbolic meaning. Behavior may involve actions such as urinating in a waste basket, moving furniture or climbing out a window.
Agitated, violent or aggressive behaviours can take place.
Injuries may arise but are uncommon.

**Night Terrors**

Family history common.
Usually first 1/3 of the night, and may be more than one episode/ night.
Last between 30 seconds and 3 minutes.
Patient will sit up and scream and appear to be in a state of terror with tachycardia, tachypnoea, dilated pupils and impossible to reassure.
May evolve into sleep walking or running.
Can be very dangerous as patient tries to escape or inappropriately protect bed partner.
Episodes tend to ‘run themselves out’ and memory is of terror and a static image.
Underlying psychopathology is common.

**Confusional Arousals**

Episodes of marked confusion during and after arousal but without sleepwalking or sleep terrors.
The subject wakens only partially and exhibits marked confusion, disorientation and perceptual impairment.
Behaviour is often inappropriate.
The confusion lasts several minutes to half an hour.
Predisposing factors include anything that deepens sleep
or impairs ease of wakening – in adults recovering from
sleep deprivation, fever, and CNS depressant medications.
It can also occur in a variety of medical conditions such as
metabolic, toxic and other encephalopathies, sleep apnoea
syndrome and idiopathic hypersomnolence.
Where there is no obvious cause, a family history is
usually found.
Inappropriate sexual behaviour in sleep with oneself or
an individual in close proximity is now recognized as
a parasomnia. It is primarily classified as a confusional
arousal, as events typically occur without any behaviour
outside the bed, but also can be less commonly associated
with sleep waking.

**Sleep Related Eating Disorder (SRED)**
Consists of recurrent episodes of involuntary eating and
drinking during arousals from sleep associated with
diminished levels of consciousness and subsequent recall.
More frequent in females and those with eating disorders
but must be distinguished from conscious overeating
during the night.
More than half of the patients with SRED have a history
of another parasomnia – it appears that two basic drive are
intertwined – sleeping and eating.
Often associated with sedative hypnotic agents in
particular benzodiazepine receptor agonists. Recent
evidence suggests it may be a non motor manifestation of
restless legs syndrome.

**Nightmares**
Nightmares generally occur in the second half of the night
within REM sleep.
Succession of images with threatening content.
Wakes the patient but there is less terror than in night

**REM Sleep Behaviour Disorder (RBD)**
Characterized by vigorous movements related to
unpleasant and often combative dreams. Vigorous and
violent behaviours of RBD commonly result in injury
which can at times be severe and even life-threatening to
both patient and bed partner.
Another theme would be participation in an active sport
such as football.
The action may be associated also with shouting and
cursing and less commonly non violent behaviours such
as laughing. Subjects rarely leave the bed but may fall out
of it.
Typically the subject wakes easily and reports a dream
which corresponds with the behaviour.
Strong male preponderence – 85%.
It may be idiopathic or associated with neurodegenerative
diseases that involve the brain stem structures that regulate
REM sleep, such as Parkinson’s disease, dementia with
Lewy bodies and multiple system atrophy. In some studies
up to 80% of subjects will develop the above within 10
years.
In some patients it may precede the motor and cognitive
symptoms.
Parasomnias in Adults

Mean age at presentation in 3 large studies ranged from 53 – 62 years. Sometimes there is a prodromal period where sleep talking, yelling and body jerking but no complex behaviours take place.
Pseudo RBD can occur in severe obstructive sleep apnoea syndrome and responds to treatment with nasal CPAP. A close association has also been found between RBD and narcolepsy. It may be associated with antidepressants, some beta blockers and anticholinesterase inhibitors.

Sleep Paralysis (isolated)
Sleep paralysis consists of episodes of inability to perform voluntary movements either at sleep onset or on wakening. Although it is one of the classic symptoms of narcolepsy, it occurs much more frequently on its own. Characteristically, limb, trunk and head movements are impossible but patient is able to move the eyes and make some degree of respiratory effort. Episodes typically are brief and disappear spontaneously or are aborted by someone touching them. They appear more frequently when patient is ‘overtired’ or in shift workers and rarely require specific treatment.

Clinical Assessment of Patients
The aims of the clinical assessment are to;
1. Establish if possible the diagnosis through clinical details.
2. Estimate the severity of the condition – in terms of frequency and disruption to the patient and partner or family.
3. Estimate the danger posed by the parasomnia to the patient, bedpartner and possibly children. Document injuries, breakages and note if patient has left bedroom, gone downstairs, broken or tried to escape through a window or if he/she has ever left the house or apartment.
4. Ensure there are no other sleep or medical disorders/medications contributing to the situation – in particular OSA
5. Enquire re. family history.
6. See if there are general underlying factors such as stress or inadequate sleep.
7. Ascertain after particular events, the precipitating factors eg. Excess alcohol or sleep deprivation due to long haul flight the night before.
8. Enquire about sleep environment both at home and away and ensure it is as safe as possible. If night terrors are taking place, window area should be secure.
9. If diagnosis is unclear, the condition is frequent or causes upset or involves harm to patient or others, the patients should be referred for further investigations through a sleep centre or neurologist.

Differential Diagnosis
Different parasomnias
Epilepsy
Psychiatric disorder
Background medical problem such as narcolepsy or Parkinson’s disease.
Investigations

1. If suspicion of epilepsy - EEG or sleep deprived EEG
2. Nocturnal polysomnography and audiovisual recording of activity
   - difficult to capture events in strange environment
   - increase in arousals from deep sleep seen in arousal disorder even in absence of events but not specific from legal point of view
   - important to verify if obstructive sleep apnoea syndrome is present or not
   - In RBD muscle tone is maintained during all or some of REM and there is a general increase in phasic motor activity. Periodic limb movements also common
3. MRI of brain where RBD is suspected.

Management of Main Parasomnias

Arousal Disorders
The treatment of the arousal disorders depends on the frequency of the episodes and seriousness of what takes place.
In all cases;
Attention to adequate sleep duration and avoidance of excessive alcohol and caffeine.
The use of all technology types is associated with increased frequency of parasomnias especially sleep walking after TV viewing and should be curtailed.
Background issues in terms of stress should be addressed.
Safety of bedroom environment should be discussed.
Medical and psychiatric conditions and their medications should be evaluated.
If the events are mild and infrequent, it is a good idea to ask patient to keep a diary and see if any precipitating factors become obvious. The frequency of these events should diminish with time in young adults.
A cognitive behavioral approach may be useful for the behaviour itself and also to improve sleep duration if required.
In more severe cases, where the diagnosis is clear, treatment with clonazepam 0.5-2 mg taken 2 hours before sleep can be very useful for a period with the dose tapered and discontinued after a couple of months of episode free sleep. Clonazepam can exacerbate obstructive sleep apnoea syndrome so it is important to ensure this is not in the background. It is also quite sedative, particularly in the morning and patients should be warned about driving especially at this time.

REM Sleep Parasomnias
The management of nightmares depends on the clinical background.
Attention to lifestyle or medications may be all that is required.
A psychiatric assessment may be useful if a diagnosis such as post traumatic stress syndrome is being considered.
As in the arousal disorders, clonazepam in similar doses can help in chronic cases. However, as nightmares may be a symptom of severe obstructive sleep apnoea syndrome it is vital that this possibility is clarified with a sleep study as prescription of clonazepam in this type of patient could be quite dangerous.
RBD is also very amenable to treatment with clonazepam but again after a sleep study has ruled out significant sleep apnoea syndrome as this mainly male, middle aged to elderly group would be at high risk for the condition.
Recent studies have also shown Melatonin to be effective in RBD.
In narcolepsy, RBD may appear spontaneously or be induced by use of clomipramine prescribed for cataplexy.
**Key Points**

- The main parasomnias are arousal disorders arising from slow wave sleep and REM sleep
- Parasomnias are common in children but relatively uncommon in adults
- Nightmares and violent acting out of dreams can be seen in both severe obstructive sleep apnea syndrome and REM sleep behaviour disorder (RSBD)
- An assessment of danger to both patient and bed partner should always be carried out
- No practice guidelines exist in the literature for the management of parasomnias

**References**


Schenck CH, Bornemann MC. Alcohol-induced sleep walking or confusional arousal as a defence to criminal behaviour: a review of scientific evidence, methods and forensic considerations. J Sleep Res. 2007. 16, 198-212.

Section 7. Diagnosis and Management of Narcolepsy In Ireland

Introduction

Narcolepsy is a chronic neurological condition characterized by excessive daytime sleepiness and sometimes cataplexy. In the recent International Classification of Sleep Disorders 3 (ICSD-3), narcolepsy is divided into Narcolepsy Type 1 where cataplexy is present and Narcolepsy Type 2 where there is no cataplexy. These two symptoms are often associated with the intrusion into wakefulness of other elements of REM sleep, such as sleep paralysis and hypnogogic hallucinations.1

Narcolepsy was first described in the late 19th century and the tetrad of core symptoms outlined in the 1950s.2 The link to human leucocyte antigen (HLA) was discovered in the 1980s and the strong association between narcolepsy and the HLA type DQBI*O602 indicates a genetic predisposition to the disorder.3 In 2000, hypocretin deficiency was established as the cause of narcolepsy.4 Examination of postmortem brains showed that the decrease or absence of CSF hypocretin was due to the loss of 70,000 hypocretin cells in the hypothalamus.5 However, efforts at demonstrating immune targeting of these cells has failed. Many investigations have searched for autoantibodies and although associations have been found these have not been consistently reproduced.

Reports from Finland and Sweden in 2010 of an increased incidence of narcolepsy in children following the H1N1 pandemic vaccine were followed by similar findings in other northern European countries.6,7,8 In Ireland, a 13 fold increase in the incidence of narcolepsy in vaccinated children < 18 years was found in comparison to non-vaccinated children.9 Epidemiological observations from China also suggest a role for H1N1 virus infection as a trigger for onset of narcolepsy.10 The prevalence of narcolepsy in European communities has been estimated at around 0.05%. Onset is most frequently in the second decade, making it a lifelong disorder. Children as young as 3 years of age have been diagnosed and rarely patients can develop the condition as late as the 6th decade.

Symptoms

- Excessive daytime sleepiness.
- Cataplexy – loss of skeletal muscle tone on emotion especially laughter.
- Sleep paralysis.
- Hypnogogic hallucinations.
- Disturbed nocturnal / sleep.
- Parasomnias including nightmares, night terrors, sleep walking and talking are reported more frequently in this group of patients. There also appears to be a higher rate of REM sleep behaviour disorder.
- Cognitive symptoms such as poor short term memory and concentration are also reported.
- Psychosocial difficulties.
- Accidents and safety issues.
Work and home accidents are more common than in non-sufferers. Patients with narcolepsy are more likely to smoke and may do so when sleepy. People with narcolepsy are 4 times more likely than controls to report sleep-related road traffic accidents.

**Assessment**
History should be structured and detailed and has the following aims:
1. Identify symptoms that support diagnosis.
2. Assess severity of condition.
3. Highlight other possible causes of daytime sleepiness such as OSA, remembering that more than one condition may co-exist.
4. Clarify lifestyle issues that may be exacerbating the condition – sleep deprivation and circadian rhythm disorders can mimic narcolepsy without cataplexy.
5. Assess the impact of the condition on the patient’s life and effects on other family members.
6. Driving history.
7. Use of structured questionnaires such as Epworth Sleepiness Scale is useful but is NOT a substitute for good history taking.

**Paediatric narcolepsy**
Cataplexy presents differently in children in the early stages of the disorder. It often appears abruptly as both emotionally triggered episodes (watching cartoons, being tickled) or as spontaneous falls to the ground (walking, running) or with generalized hypotonia with prominent facial involvement resulting in ‘cataplectic facies’. This involves facial muscle weakness with jaw slackness, ptosis, tongue protrusion and slurred speech. Other findings include grimacing, body swaying and other purposeless movements.11
Sleep paralysis and hypnogogic hallucinations may be difficult to ascertain in young children. Very disturbed sleep with parasomnias and restlessness is common in this age group. Total sleep time is long but symptoms improve spontaneously after about 2 years giving way to more ‘adult’ type cataplexy and shorter more age appropriate sleep times.11
Irritability and significant weight gain are also prominent features of paediatric narcolepsy.

**Examination**
A full physical examination should be carried out looking for other underlying causes of sleepiness, e.g. obesity and obstructive sleep apnea syndrome, hypothyroidism or CNS signs of an intracranial lesion leading to a secondary narcolepsy. Mood should also be assessed and signs of psychological distress.

**Investigations**
1. **Sleep studies:** (Recommended by AASM in all possible narcoleptic patients to clarify diagnosis)12
   - Full polysomnography with leg EMG
   - Multiple Sleep Latency Test (MSLT)
As narcoleptic patients are more prone to other sleep disorders like OSA and restless legs syndrome, polysomnography may need to be repeated on several occasions throughout the patient’s life.

**Nocturnal polysomnography:** a REM sleep onset period (SOREM) is seen in 50% of patients with type 1 narcolepsy. Very fragmented sleep is the norm with frequent returns to wake and micro arousals. OSA and PLMs are seen more often than in normal populations. REM sleep behaviour disorder can present at an early age in narcolepsy with raised EMG tone in REM sleep but EMG tone is often raised even without symptoms of the condition.

**Multiple Sleep Latency Tests** are time consuming and require a lot of experience and skill to be accurate. A negative test does not mean that patient does not have narcolepsy and results must be interpreted with caution.
Only 71% of narcoleptics with cataplexy have a mean sleep latency of < 8 minutes and 2 Sleep Onset REM Periods on initial testing. Repeated testing fails to bring this figure above 80%. Prior to testing patients should be off all medication for at least 2 weeks and 3 weeks in the case of fluoxetine.

2. Actigraphy is recommended but at least a sleep diary should be kept for at least 2 weeks beforehand. This is to reduce the incidence of false positive tests due to sleep deprivation or circadian rhythm disorders.

3. HLA typing is an expensive test and does not need to be carried out routinely. The HLA type DQBI*O602 is present in 95% of narcoleptic patients (with cataplexy) but it is also present in 18 –30% of the normal population and therefore is of more help when it is negative and helps exclude narcolepsy in doubtful cases.

CSF hypocretin levels are low (<110pg/ml) in 87%-96% of patients with Type 1 narcolepsy. In Type 2 narcolepsy hypocretin estimation is less helpful: a recent study showed low levels in 24%, intermediate levels (110-200pg/ml) in 8% and normal levels in 68% of cases.13

5. Magnetic Resonance Imaging of Brain. To ensure no underlying secondary condition such as multiple sclerosis or hypothalamic tumour in children.

Multiple Sleep Latency Protocol
• The MSLT must be performed after a full overnight polysomnogram (nPSG).
• The MSLT should only be performed after at least 6 hours sleep (adults), 8-9 hours (children and adolescents depending on age).
• A sleep diary or preferably Actigraphy should be carried out for two weeks prior to testing. The MSLT should be cancelled if there is evidence of an irregular sleep wake cycle or other factors which may lead a false positive result.
• Testing should begin the earliest at 90 minutes after rise time and no later than 3 hours after rise time.
• 5 naps should be conducted at 2 hour intervals, unless there was 2 sleep onset REM periods (SOREMs) in the first 4 naps. The minimum number of naps required for a MSLT is 4 naps.
• Standardisation of testing conditions is important (every nap for every individual should be conducted under the same testing conditions).
• Rooms should be dark and quiet and at a comfortable temperature.
• Stimulant medication should be withdrawn for the appropriate time prior to testing.
• Drug screening to be performed on the morning of the test.
• Smoking should be stopped at least 30 minutes prior to each nap.
• Caffeine should be avoided from midnight until the end of the test.
• Bright light and LED screens should be avoided on test day.
• Sleep Technologists should be well experienced in conducting the test.
• The patient should be monitored in between naps to ensure they are following the rules of testing.
• Recording montage minimum requirements: frontal, central and occipital EEG, EOG and EMG.
• Patients should be dressed and changed into normal day wear prior to the first nap.
• Bio calibrations should be performed before every nap.
• The patient should be given the same instructions at the start of every nap: “Please lie quietly, assume a comfortable position, and try to fall asleep” the bedroom lights should then be turned off.
• The patient should not lie or sit on the bed between naps and they are to remain awake in between naps.
• Sleep onset is defined as the time from lights out to the first epoch (>50%) of any stage of sleep.
• If there is no sleep, the nap is ended after 20 minutes and sleep latency is 20 minutes.
• If there is sleep, the test continues for 15 minutes of clock time after the first epoch of sleep.
• REM latency is the time from the first epoch of sleep to the first epoch of REM sleep.
• The MSLT report should include the start and stop times of each nap, latency from lights out to first epoch of sleep, mean sleep latency and the number of sleep-onset REM periods.

Mean sleep latency findings: (please note these ranges are for >16 years of age and should be used as a guideline in conjunction with the patient history)

<table>
<thead>
<tr>
<th>&lt; 5 minutes</th>
<th>Pathologically sleepy</th>
</tr>
</thead>
<tbody>
<tr>
<td>5 – 10 minutes</td>
<td>Grey area</td>
</tr>
<tr>
<td>&gt; 10 minutes</td>
<td>Within normal limits</td>
</tr>
</tbody>
</table>

Differential Diagnosis
1. Obstructive sleep apnea syndrome
2. Idiopathic hypersomnia – long and short sleep type
3. Depression
4. Delayed sleep phase syndrome
5. Chronic sleep deprivation.

Management of Narcolepsy
ISS considers it very important to diagnose narcolepsy early and correctly to minimize educational and social impact. Management of narcolepsy involves not only the use of medications which are effective for both daytime sleepiness and cataplexy but also giving general information (oral and written) on the symptoms and specific information on education, work and contraception/pregnancy as required. Children may be entitled to extra tuition from the department of education to maintain their academic status.

Discussion on sleep hygiene and use of strategic napping is also important.

Driving ability needs to be assessed on every visit over the years. It is mandatory to report narcolepsy to motor insurance company and to Road Safety Authority at renewal of licence.

Management will vary over the course of a lifetime and follow up is recommended at yearly intervals to specialist involved in care. Associated conditions such as OSA, restless legs syndrome should be identified and treated. Obesity and other eating disorders may need to be addressed.

If Xyrem is being used, patient/parents must understand from the beginning that for safety reasons, it can never be associated with the intake of alcohol.
### Treatment of Excessive Daytime Sleepiness\(^\text{15}\)

<table>
<thead>
<tr>
<th>Drug</th>
<th>Side effects</th>
</tr>
</thead>
<tbody>
<tr>
<td>Modafinil 200-400 mg / day in single or divided doses. (not licenced for &lt; 18 years)</td>
<td>Headache, nervousness Contraceptive issues</td>
</tr>
<tr>
<td>Methylphenidate 20-80mg / day (Ritalin or Concerta)</td>
<td>Irritability, headache, insomnia</td>
</tr>
<tr>
<td>Dexamphetamine 10-60mg / day</td>
<td>As above, reduced appetite, palpitations</td>
</tr>
<tr>
<td>Gammahydroxybutyrate 3-9g / night in divided doses (14)</td>
<td>Nausea, dizziness, eneuresis. Cannot be associated with alcohol. Contraindicated with untreated OSA</td>
</tr>
</tbody>
</table>

### Treatment of Cataplexy\(^\text{16}\)
- Venlafaxine - 37.5 - 150 mg / day
- Tricyclics - clomipramine 20 - 150 mg / day (AASM guideline)
  - SSRIs (AASM guideline)
- Gammahydroxybutyrate - effective and well tolerated even in paediatric age group\(^\text{17}\)
  (but not licenced in < 18 age group)

Treat other co-existing sleep disorders appropriately eg. CPAP for OSA

### Key Points
- It is very important that narcolepsy is diagnosed close to onset to ensure good academic and social development
- Management includes behavioral modifications as well as medication
- In case of children, good parental and school support is vital
References
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Links

www.irishsleepsociety.org Irish Sleep Society, Ireland
www.isat.ie OSAS support group Ireland
www.sleepy-heads.org Narcolepsy support group Ireland

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