

NARCOLEPSY

University Hospital Aintree Sleep Service Pathway

GUIDELINE

Diagnosis of Narcolepsy made by Consultant Sleep Physician at University Hospital Aintree according to ICSD-3 criteria (*International Classification of Sleep Disorders. 3rd ed. (2014) American Academy of Sleep Medicine*)

1st Line agent A Ret

- Stimulant therapy: Modafinil 100-600mg daily for Excessive Daytime Sleepiness; Anti-depressant REM suppressant therapy for cataplexy: Clomipramine /imipramine/fluoxetine/venlafaxine which are titrated to response.
- If the patient's Narcolepsy is well controlled i.e. improvement in daytime sleepiness and decreased frequency of cataplexy episodes and REM intrusion phenomena: Continue 1st line treatment of modafinil and REM suppressant anti-depressants. If ineffective then stop and change.
- Before prescribing for patients they will be assessed for clinical suitability. Patients should have their blood pressure and heart rate monitored every at every clinic appointment following initiation which is usually every three to 6 months depending on clinical stability.

2nd Line agent RED

- If the patient's excessive daytime sleepiness is not controlled with modafinil therapy, a 2nd line option may be the initiation of stimulants such as methylphenidate or dexamphetamine titrated to response. If ineffective then consider adding another agent which should allow down titrating of stimulants and/or anti-cataplexy drugs.

Before prescribing for patients they will be assessed for clinical suitability. Patients should have their blood pressure and heart rate monitored every at every clinic appointment following initiation which is usually every 3 to 6 months depending on clinical stability.

If the subject's Narcolepsy symptoms (excessive daytime sleepiness/cataplexy/REM intrusion phenomena) is poorly controlled with or intolerant / contraindicated to the above, then an assessment should be made considering the following variables:

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| – Comorbidity | – BMI |
| – Psychiatric history | – Coexisting untreated OSA or hypoventilation (consider CPAP or NIV therapy) |
| – Frequency of cataplexy | – History of alcohol/sedative/illicit drug use |
| – Degree of REM intrusion phenomena | – History of coexisting insomnia |
| – History of nocturnal incontinence | |
| – Whether there has been a previous adverse reaction to sodium oxybate or pitolisant | |

Continued

Note: Patients who are not eligible for treatment under this statement may be considered on an individual basis where their GP or consultant believes exceptional circumstances exist that warrant deviation from the rule of this policy. In this situation, follow locally defined processes.

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3rd Line Agent **RED**

Indications to favour Pitolisant as 3rd line therapy

- Contraindicated or adverse reaction to the use of modafanil, dexamphetamine / methylphenidate
- Untreated obstructive sleep apnoea / hypoventilation
- Significant anxiety (psychiatric history)
- History of urinary incontinence at night
- Low BMI i.e. less than 18 / history of active eating disorder
- Previous patient who has had adverse reaction or intolerant to sodium oxybate
- Narcolepsy specific issues: Less severity of cataplexy (10-15 episodes a week), paucity of REM intrusion phenomena
- History of regular daily alcohol consumption and unwilling to reduce this
- Coexisting sedative use/misuse or potential for this to occur
- Unwilling to wake up after 2 hours to take 2nd dosage of medication/prefers once a day morning regime
- History of significant hypertension and cardiovascular comorbidity

Sodium oxybate

Indications to favour sodium oxybate as 4th line therapy or in favour before pitolisant **RED**

- Previous adverse reaction to or lack of clinical response to pitolisant
- Narcolepsy specific issues: Significant (i.e. multiple nightly) REM intrusion phenomena, greater frequency of cataplexy e.g. >15 episodes a week
- Coexisting insomnia
- Obesity

All patients commenced on pitolisant or sodium oxybate MUST be followed up every 3 months in the specific Narcolepsy clinic.