Idiopathic hypersomnia (IH) is a neurologic disorder characterized by excessive daytime sleepiness (often with associated cognitive dysfunction), despite getting a full night’s sleep or longer. Person(s) who have IH (PWIH) may experience other symptoms related to the autonomic nervous system, such as feeling lightheaded when standing up quickly or abnormal body temperature regulation.\(^2\)

PWIH may face unique challenges related to a hospitalization or a procedure (including dental) with anesthesia. It is crucial for both patients and healthcare providers to pay special attention to managing IH symptoms before, during, and after hospitalization or anesthesia.

This Guide will help with advance preparation, setting expectations, and understanding how the diagnosis of IH can impact care. Current research indicates significant overlap between IH and narcolepsy type 2 (NT2), so much of this Guide may also be applicable to people with NT2.

Special Consideration Needed for PWIH Because

1. Sedating medications (such as opioids and benzodiazepines) can worsen IH symptoms.

2. PWIH may respond differently to anesthesia and be slower to awaken from anesthesia—“delayed emergence” (this is also quite common in people without IH and does not indicate a poor outcome). Alternative approaches to anesthesia may reduce this risk. All anesthesia providers (including dentists) should be prepared to treat PWIH having difficulty emerging from anesthesia (e.g., with flumazenil, which is also used as a treatment for IH).

3. The medications used to treat IH may significantly interact with anesthetics or other medicines. When safe to do so, it is preferred to continue IH medicines.

4. To best manage their symptoms, PWIH often require a specific sleep and medication schedule, with as few interruptions to sleep as possible. A procedure or hospitalization may significantly disrupt that schedule and lead to severe sleepiness/cognitive dysfunction that could mimic delirium. By discussing in advance, scheduling changes and alternative treatments can be designed to minimize disruption of IH treatment.

5. PWIH may be very difficult to awaken from sleep, with severe/prolonged sleep inertia.

**BEFORE ANESTHESIA/HOSPITALIZATION**

Create a Care Plan Now

IH is uncommon, so most healthcare staff are unlikely to be familiar with IH and its sleep and medication needs. It is advisable to develop an Anesthesia/Hospital Care Plan (see our form\(^1\)). Because an emergency may arise unexpectedly, it is best to have a Care Plan ready at all times. When anesthesia or hospitalization is planned, it is advisable to revise the Care Plan for that event.

**Meet in Advance With IH Healthcare Provider**

It is crucial for the IH provider to directly communicate this Guide, the Care Plan and sleep study reports to the surgical and anesthesia teams. Well before anesthesia or hospitalization, PWIH should schedule time with their IH healthcare provider to review their symptoms and this Guide and to optimize care and revise the Care Plan. Discuss strategies for coping with IH symptoms/medicines before, during, and after anesthesia or hospitalization and communicating this information. Because IH is uncommon, PWIH need to advocate for themselves, and, whenever possible, bring a friend/family member to help with that advocacy.\(^3\)

**Plan for oxybate salts (e.g. Xyrem), given the potential severity of interactions (including with anesthesia/opioids), lack of hospital availability, variable and late-night dose timing, and importance of carefully conserving/dosing. Consider a med holiday/substitution for brief hospitalizations. Otherwise, consider advocating for patient self-mixing and administration, with nurse notification/monitoring, and storage in patient’s locker.**

**Meet in Advance With Surgeon/Hospital-Admitting Doctor AND Anesthesiologist**

Because IH can cause significant sleepiness and/or cognitive impairments, it is crucial that PWIH meet in advance with the providers who will be in charge of their care, e.g., if a PWIH is being admitted to the hospital for chemo, this will be their oncologist. If a PWIH is having a colonoscopy, they may be referred directly without first meeting the GI doctor. However, it is strongly advised that an appointment with the GI doctor be scheduled in advance of the procedure to discuss this Guide and the Care Plan.

Patients often meet with the anesthesia team just minutes before a procedure. It is crucial for the surgeon to help facilitate this meeting well in advance (it may be in-person or via telemedicine) and to provide this Guide, the Care Plan, and sleep studies to the anesthesia team prior to that meeting. Although it may not be possible to know in advance which anesthesiologist will be attending the PWIH’s procedure, an advance meeting is still essential. PWIH may then specifically request that the anesthesia provider with whom they met provide the anesthesia care (assuming the schedule permits) or communicate the Guide and Care Plan with their colleague who will be providing care. **Discuss the following (and revise the Care Plan as needed):**

1. **Anesthetic plan, including type of anesthetic(s).** This will be based on the risks and benefits of using general anesthesia, regional anesthesia, or sedation. If possible, the anesthesiologists should use regional anesthesia (also referred to as a “nerve block”) to temporarily anesthetize a particular area of the body to prevent sensation or pain during the procedure. Nerve blocks may last for a number of hours after the procedure, thereby decreasing the amount of opioid medications needed to control pain. This may be beneficial, as opioids can potentially interfere with sleepiness levels and recovery from anesthesia, but a regional anesthetic may not always be possible.
2. IH medicines may interact with anesthetics, and IH symptoms may cause a need for different amounts or types of anesthetics to maintain appropriate depth of anesthesia. Consider monitoring depth of anesthesia and using shorter-acting anesthetics whenever possible.

3. The pain management plan should attempt to use “multimodal analgesia” (multiple different techniques or types of medications for pain control) in order to minimize the use of opioids.

4. PWIH may need closer-than-usual monitoring for alertness or over-sedation during and after anesthesia.

**THE DAY OF ANESTHESIA/ DURING HOSPITALIZATION**

Many IH medications should be taken as usual for as long as possible before anesthesia or hospitalization. (A notable exception is oxybate salts.) This is important not only for controlling IH symptoms, but also for minimizing any side effects related to re-establishing the dose. The details should be part of the Care Plan.

**Bring the Following to Procedure/Hospitalization**

- **Care Plan, Guide and FAQ Cards.** Have several copies to keep with the chart and to share with the healthcare team and accompanying family members/friends.
- **Medical alert card,** or any other preferred medical alert (such as a bracelet or smartphone app).
- **A supply of prescribed IH medications, in case the pharmacy cannot get them. Note: PWIH should inform their team that they have these medications with them.**

**Review the Care Plan and Guide**

The entire healthcare team needs to be made aware of the IH diagnosis, this Guide, and the Care Plan.

- **Confirm details of any anesthesia plan,** which were ideally discussed in advance and added to the Care Plan.
- **Review the treatment plan,** including pain management and: 1) usual response to pain medicines, including opioids; 2) the need to avoid sedating medications as much as possible; 3) the need to use alternative medications, if available, if one can’t take oral medications; and 4) that sleep deprivation can significantly worsen IH symptoms.
- **Pay careful attention to the level of sleepiness.**
- **Optimize scheduling to avoid treatment interruptions that may quickly lead to severe sleepiness/cognitive dysfunction mimicking delirium.** Every time a PWIH leaves their room for a procedure (such as PT) or transfers to a different clinical area, their sleep and/or IH medication schedule may be delayed or interrupted. This should be discussed in advance with the entire healthcare team so that all staff knows to prioritize the PWIH’s medication and sleep needs.
- **Resume usual treatment as soon as safely possible,** after careful consideration of possible drug interactions, to avoid worsening of IH symptoms.

**AFTER ANESTHESIA/HOSPITALIZATION**

**Review the Discharge Plan**

- Discuss any potential medication side effects, such as drowsiness or slowing of breathing from opioids.
- Discuss any possible interactions between new and regular medications.
- It is important to balance the level of pain control with any possible sedating side effects or drug interactions.
- PWIH should be advised to keep track of their level of sleepiness after discharge home. If sleepiness worsens, caregivers and doctors should be informed, to determine if any medication changes are needed.

**Escort Home**

If discharged the day of anesthesia, an escort home will be needed.

**Avoid Driving or Operating Heavy Machinery**

After anesthesia, PWIH should avoid driving or operating heavy machinery until it is safe to do so. Usual recommendations for all patients are to avoid these activities for at least 24 hours, but PWIH may need to avoid these activities for even longer. Medications may need to be adjusted before PWIH resume these activities.

**Obtain Records**

It is a good idea for PWIH to obtain copies of all of their medical records. Some PWIH report that their IH symptoms seem better or worse for an extended time period after anesthesia. Especially in these cases, it is prudent to obtain anesthesia records, which can then be used to help guide future anesthesia and for research studies in which the PWIH wishes to participate.

**REFERENCES/RESOURCES**

1. See hypersomniafoundation.org/anesthesia-and-ih for full references/resources
2. hypersomniafoundation.org/ihsummary
3. hypersomniafoundation.org/document/self-advocacy
4. hypersomniafoundation.org/medical-alert

**DISCLAIMER:** This Guide is intended for educational purposes only; it is not meant to substitute for medical advice or to be taken as such. Every individual is different, and their medical conditions can vary widely. Please consult your own healthcare professionals before making any decisions regarding your healthcare or your medical management.

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