

How do you plan for emergencies, hospitalization & anesthesia if you have a hypersomnia?

1. Who can benefit from preparing for anesthesia and hospitalization?

- Everyone who has a hypersomnia, such as IH, narcolepsy, or KLS.
 - Emergencies are unpredictable.
 - Almost everyone in the U.S. has anesthesia at some point in their lives, whether for a dental procedure, a colonoscopy, or a surgery.
- Because hypersomnias are uncommon, healthcare professionals may not be familiar with them.
- Hospitalization and anesthesia can affect:
 - hypersomnia symptoms
 - medication needs
 - strategies for coping (such as daytime sleep needs)

2. Why do you need special consideration and preparation for anesthesia and hospitalization if you have a hypersomnia?

1. Sedating medications can worsen hypersomnia symptoms.
 - These meds include opioid pain medicines and benzodiazepines, which are often used in anesthesia and during hospitalization.
 - Alternatives should be considered, whenever possible.
2. You may be slower to awaken from anesthesia.
 - This is called “delayed emergence” and is also quite common in people without hypersomnias. This does not indicate a poor outcome.
 - Alternative approaches to anesthesia may reduce the risk.
 - All anesthesia providers (including dentists) should be prepared to treat people having difficulty emerging from anesthesia (for example, with flumazenil, which is also used as a treatment for IH).
3. The medications used to treat hypersomnias may significantly interact with anesthetics or other medicines.
 - Therefore, you may need to temporarily stop some of your hypersomnia meds (such as Xyrem).
 - When safe to do so, it is preferred to continue your hypersomnia meds.
4. To best manage your symptoms, you may need 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 and cognitive dysfunction that could mimic delirium.
 - By discussing this possibility in advance, scheduling changes and alternative treatments can be designed to minimize disruption of hypersomnia treatment.

5. If you may be very difficult to awaken from sleep or you experience severe sleep inertia (also called sleep drunkenness), it is important for your healthcare teams to know your specifics.
 - How long does your sleep inertia usually last?
 - Do you experience automatic behaviors (seeming normal but then forgetting what you said or did)?
6. If you have cataplexy, it is important for your healthcare teams to know your specifics.
 - What are your usual triggers?
 - How long are your typical attacks, and what do they look like?

3. When and how do you prepare an emergency Care Plan?

1. Prepare now!
2. Download our materials from www.hypersomniafoundation.org/anesthesia-and-ih.
3. Schedule an appointment with your hypersomnia care professional to discuss.
4. Prepare a rough draft of your written Care Plan.
5. Share HF's Guide and your draft Care Plan with your hypersomnia care professional in advance of your appointment if possible, so they'll have the opportunity to review it.
6. Plan to advocate for yourself.
 - Whenever possible, bring a friend or family member to help with that advocacy.
 - See our self-advocacy handout at www.hypersomniafoundation.org/document/self-advocacy.
7. Create or update your medical alert(s). See hypersomniafoundation.org/medical-alert for options.

4. How should you share your Care Plan to ensure it's accessible when you need it?

- Share the most current version in one or more of the following ways:
 1. Provide it to your emergency contact.
 2. Upload it to a designated folder in your personal secure cloud service, such as Google Drive or Dropbox.
 3. Ask to have it added to your medical record (or you may be able to upload it yourself to your electronic medical records).
 4. Provide it directly to your anesthesia, surgical, and/or hospital teams.
- What should you include with your Care Plan?
 1. Your sleep study reports
 2. HF's Anesthesia/Hospitalization Guide
 3. HF's IH FAQ card
 4. Any other medical records that might be important, like anesthesia records.
- Update your medical alert (card and/or smartphone app) to direct to your Care Plan
 - Via your emergency contact's phone number and/or
 - Via the web address for your cloud service folder. This will allow you to change the contents of your folder at any time without changing the web address. (You

may want to shorten the web address with a free url shortener, like Bitly or TinyURL.)

5. How do you complete your Care Plan?

1. Fill out as much as you can on your own.
2. Work with your hypersomnia professional to update your emergency Care Plan.
3. Discuss strategies for coping with your hypersomnia symptoms and medicines before, during, and after anesthesia or hospitalization. Discuss how to communicate this information to your emergency care teams.
4. When you plan to have anesthesia or be hospitalized, work with your hypersomnia professional and your anesthesia/hospital professionals to update your Care Plan for this specific event.
 - a. It is crucial for your hypersomnia professional to:
 - i. directly communicate the Guide and your Care Plan and sleep study reports to your surgical, anesthesia, and/or hospital teams
 - ii. help facilitate your meeting in advance with the anesthesia, surgical and/or hospital teams (consider telemedicine)
 - b. Examples of events needing Care Plan updates:
 - i. For any procedure with anesthesia, you should meet with the anesthesia professional in advance. This will often require self-advocacy, and it is critical for your hypersomnia professional and/or the professional doing your procedure to help facilitate your meeting in advance with the anesthesia professional.
 - ii. Colonoscopies are usually performed by a GI specialist or colorectal surgeon. You should have an appointment with this professional to discuss and update your Care Plan in advance of your procedure and then facilitate a meeting with the anesthesia professional.
 - iii. For dental procedures, you should discuss your Care Plan in advance with your dentist. Most do their own anesthesia.
 - iv. For hospitalization for chemotherapy, you should discuss and update your Care Plan with your oncologist before you're admitted to the hospital.
 - v. For other planned hospitalizations, you should usually discuss and update your Care Plan in advance with the professional who will be admitting you to the hospital.

6. Why is it so important to meet with the anesthesia, surgical, and/or hospital teams in advance?

1. You will have more time to discuss the issues on your Care Plan.
2. You can plan to have a supporter with you to help with your self-advocacy and/or note-taking.
3. This will allow you to better prepare for the anesthesia or hospitalization.

- For example, if you take an oxybate med, like Xyrem, you will be able to make a careful plan for that.
- 4. This will give your healthcare teams time to learn about hypersomnias, including how to provide the best care.
- 5. Patients often meet with the anesthesia team just minutes before a procedure.
 - Since hypersomnias can cause significant sleepiness and/or cognitive impairments, you may not be in a cognitive state to discuss the potential issues on your Care Plan in that small and often stressful time period.
 - You may also be less able to have a supporter with you to help you advocate for yourself.
- 6. Although it may not be possible to know in advance which anesthesiologist will be attending your procedure, an advance meeting is still essential. You may then specifically request that:
 - the anesthesia professional with whom you met provide the anesthesia care (assuming the schedule permits), **or**
 - communicate the Guide and Care Plan with their colleague who will be providing care.

7. What information goes on the Care Plan?

There is a sample completed care plan on our web page.

1. Your name and date of birth.
2. The date of the Care Plan.
 - Note: you should change the date of the Care Plan each time it is updated.
3. Your medical diagnoses.
 - Your hypersomnia disorder.
 - Other significant diagnoses, like diabetes, hypertension, anxiety disorder, etc.
4. Name and phone number of your hypersomnia professional (and other significant healthcare professionals).
5. Describe your usual hypersomnia symptoms and how well they are controlled.
 - Include symptoms such as sleepiness, nap/sleep needs, cognitive function, automatic behaviors, autonomic symptoms (such as lightheadedness on standing, urinary frequency, etc), cataplexy.
 - Include information about other medical issues, if needed.
6. Date of your most recent sleep studies.
 - Obtain copies to share with your Care Plan.
7. Details about your medications, both prescription and over-the-counter.
8. Describe your usual response to opioid pain medications.
 - In general, sedating medications may worsen hypersomnia symptoms and should be avoided when possible.
 - Although opioid pain medicines are usually sedating for most people, they may have the opposite effect on some people with hypersomnias.
9. List allergies or sensitivities to medications.

10. List any problems you've had in the past during anesthesia, surgery, or hospitalizations. Include records if possible.
11. Discuss with your healthcare professionals which, if any, of your medicines, you should avoid around the time of anesthesia and/or hospitalization. Are there any recommended substitute treatments?
 - For example, oxybates such as Xyrem and Xywav should be avoided around the time of anesthesia.
12. Discuss with your healthcare professionals if there are any substitutes for your medications if/when you can't take them orally (for example, if you are intubated or experiencing significant nausea/vomiting).
 - Most hypersomnia medications do not have non-oral substitutes, but some, such as clarithromycin, do.
 - Medications you take for other conditions may have non-oral substitutes (such as intravenous, transdermal, rectal, etc.).
13. List any anesthesia modifications you've had for your hypersomnia disorder in the past. If you have a planned procedure with anesthesia, discuss with your anesthesia professional whether any of the following might help speed your recovery (with a goal to avoid worsening of your hypersomnia symptoms):
 - use of shorter-acting anesthetics
 - limitation of opioid use
 - use of regional anesthesia, wherever applicable (such as a nerve block for shoulder or knee surgery; or an epidural for labor and delivery).
14. Discuss with your anesthesia professional if it will be helpful for you to have any extra care or monitoring planned for your procedure.
 - Example: depth of anesthesia monitoring.
15. Who will be your escort home if needed, such as if you go home the same day as anesthesia or are unable to get yourself safely home for another reason?

8. What should you do if you take an oxybate medication, such as Xywav?

- It's a good idea to discuss a careful plan with your prescribing doctor. There are several concerns:
 - potential severity of med interactions (including with anesthesia and opioids)
 - lack of hospital availability
 - variable and late-night dose timing
 - the importance of carefully conserving your medicine and carefully dosing
- For anesthesia and brief hospitalizations, consider a med holiday or substitution.
 - ESSDS pharmacists advise skipping oxybates the night before and the night after anesthesia.
 - Some prescribers also advise separating oxybate doses from opioid doses by at least 12 hours, although ESSDS pharmacists may advise a shorter time period.
- For longer hospitalizations, you and your doctor can consider advocating for you to self-mix and take your oxybate medication, with nurse notification/monitoring, and storage in your locker.

- Hospitals often have a policy that medications brought in by patients must be taken to the hospital pharmacy for storage and dosing and then administered by nursing staff. In this case, you and your prescribing doctor may need to strongly advocate for a policy exception. Why? We have heard numerous stories from people who've had their oxybates taken to the hospital pharmacy for storage and dosing, which has in some cases led to:
 - lost bottles of medicine;
 - lost volume of medicine, presumably wasted during dose measurements and not discovered until the patient ran out of medicine later at home
 - inaccurately measured doses
 - delayed dosing
- Hospital staff are for the most part unfamiliar with oxybates and often do not understand that it is extremely expensive and rigidly controlled, such that even losing a small amount of medicine can be a significant hardship for the patient.
- Some patients who have experienced these problems report having hidden their oxybates in their locker or purse during subsequent hospitalizations. However, this can be very dangerous. It is extremely important that your hospital team be aware that you are taking this medication because of potentially dangerous med interactions and also so they are aware that you are on it at night.

9. You've updated your Care Plan with your hypersomnia professional and your surgical or hospital admitting team. Now, what should you discuss with the anesthesia team in your advance meeting?

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.
 - Is a long-acting nerve block possible? If so, this may further reduce your need for sedating post-op pain meds over a period of a few days. However, it may cost more and also have other side effects (such as inability to move your arm for the duration of a shoulder nerve block).
2. Hypersomnia medicines may interact with anesthetics, and hypersomnia 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 medicines for pain control) in order to minimize the use of opioids.
4. You may need closer-than-usual monitoring for alertness or over-sedation during and after anesthesia.
5. If you need scheduled daytime sleep sessions (naps) to function, will your time under anesthesia count as sleep?
 - Maybe not to probably not. For example, conscious sedation, such as you often get for colonoscopies, usually does not count as sleep.

10. What should you bring on the day of anesthesia or admission to the hospital?

1. 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.
 - FAQ cards provide a very quick overview of IH and can be displayed on your chart, bed rail, hospital room door, etc.
2. Medical alert card and/or any other preferred medical alert (such as a bracelet or smartphone app).
3. A supply of prescribed hypersomnia medications, in case the pharmacy cannot get them.
Note: It is very important to inform your team that you have these meds with you.

11. What should you do while you're at the facility or hospital?

1. The entire healthcare team needs to be made aware of your hypersomnia diagnosis, HF's Guide, and your Care Plan.
2. Confirm the details of any anesthesia plan, which were ideally discussed in advance with the anesthesia team and added to the Care Plan.
3. Review the treatment plan, including pain management and:
 - Your usual response to pain medicines, including opioids;
 - the need to avoid sedating medications as much as possible
 - the need to use alternative medications, if available, if you might not be able to take oral medications
 - that sleep deprivation can significantly worsen hypersomnia symptoms.
4. Pay careful attention to your level of sleepiness.
5. Optimize scheduling to avoid treatment interruptions that may quickly lead to severe sleepiness/cognitive dysfunction mimicking delirium.
 - Every time you leave your room for a procedure (such as physical therapy or a CAT scan) or transfer to a different clinical area, your sleep and/or your 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 your medication and sleep needs.

6. Plan to resume your usual treatment as soon as safely possible, after careful consideration of possible med interactions, to avoid worsening of hypersomnia symptoms.

12. What should you do when it's time to go home?

1. Review your 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.
 - Keep track of your level of sleepiness after discharge home. If sleepiness worsens, you should inform caregivers and healthcare professionals and determine if any medication changes are needed.
2. Confirm if you'll need your planned escort to take you home.
 - Usually, if you're discharged the day of anesthesia, an escort home will be needed.
3. Confirm how long you need to avoid driving or operating heavy machinery
 - After anesthesia, you 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. However, people with hypersomnias may need to avoid these activities for even longer.
 - Your medicines may need to be adjusted before you can safely resume these activities.
4. Obtain medical records
 - It is a good idea to obtain copies of all of your medical records, or you may be able to access them through your electronic health record (see www.hypersomniafoundation.org/document/self-advocacy).
 - Some people with hypersomnias report that their hypersomnia symptoms seem better or worse for an extended time period after anesthesia. Especially in these cases, it is a good idea to obtain anesthesia records, which can then be used to help guide future anesthesia and for research studies in which you wish to participate.

13. Is there a quick overview of all these steps?

The "Before, During, and After" graphic on the Care Plan instruction page and the web page provides a summary.