

## Anesthesia recommendations for ROHHAD

**Disease name:** ROHHAD (acronym of Rapid-onset Obesity with Hypothalamic dysfunction, Hypoventilation and Autonomic Dysfunction)

**ICD 10:** Does not have a specific ICD code. It utilizes several codes, examples include E23.3 (Hypothalamic dysfunction, not elsewhere classified), E66.8 (other obesity), R06.89 (other abnormalities of breathing), and G90.9 (Autonomic Nervous System disorder, unspecified).

**Synonyms:** Rapid onset obesity, hypoventilation, hypothalamic dysfunction, and autonomic dysfunction

**Disease summary:** ROHHAD is a clinical entity with a median age of 3 years at onset characterized by sudden onset of dramatic weight gain, dysautonomia, and pulmonary complications. These include alveolar hypoventilation, obstructive sleep apnea, and decreased central responsiveness to hypercarbia. There are also several endocrinopathies associated with this disorder including hypernatremia, hyperprolactinemia, hypothyroidism, hypocortisolism and diabetes insipidus. Additional features of the disease include behavioral and mood disorders, as well as seizures and learning impairment. There is an associated entity known as ROHHADNET with a predisposition towards the development of neuroectodermal tumors such as neuroblastoma and ganglioneuroma [1]. As ROHHAD could be a neurological paraneoplastic syndrome, all patients with ROHHAD syndrome should be carefully screened for the presence of these tumors [2]. There is a recent increase in the number of cases reported from Asia, and their data are included in this update.

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Medicine is in progress



Perhaps new knowledge

Every patient is unique

Perhaps the diagnosis is wrong

Translations may not always reflect the most recent updates of the English version

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Find more information on the disease, its centers of reference and patient organizations on Orphanet: [www.orpha.net](http://www.orpha.net)

## Emergency information

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<b>A</b>	<b>AIRWAY / ANESTHETIC TECHNIQUE</b>	Ventilation more difficult due to soft tissue collapse, obesity. To date, there has not been literature suggesting difficult airway. Central and obstructive sleep apnea.
<b>B</b>	<b>BLOOD PRODUCTS (COAGULATION)</b>	There is no known bleeding diathesis associated with this disorder.
<b>C</b>	<b>CIRCULATION</b>	Obesity/alveolar hypoventilation affects lung function. Maybe right ventricular hypertrophy (RVH).
<b>D</b>	<b>DRUGS</b>	All medications with respiratory depressant effects should be administered judiciously. Caution is also advised with medications having long half-lives and respiratory depressant effects.
<b>E</b>	<b>EQUIPMENT</b>	Patients may have a tracheostomy tube in place, all necessary precautions for tracheostomy utilization should be employed. Postoperative cardiorespiratory monitoring.

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## Typical surgery and procedures

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Depends on the presentation of disease. May vary from GI procedures for associated Hirschsprung's disease, surgeries to palliate abnormal ventilation (diaphragmatic pacing [3], tracheostomy placement), or dysautonomia (permanent cardiac pacemaker placement). Surgery for removal of neural crest tumor (ganglioneuroma, neuroblastoma) [4,5]. May also be present for any surgery in the typical array of pediatric surgeries.

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## Type of anesthesia

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Either general or regional anesthesia can be used for these patients with the following caveats. General anesthesia: patients may have impaired thermoregulation, hypothalamic dysfunction resulting in electrolyte disturbances, cardiac dysrhythmias and hemodynamic fluctuations due to autonomic dysfunction.

In case of ganglioneuroma or neuroblastoma, preoperative systemic hypertension may be present as well as large swings in intraoperative blood pressure during surgery: these cases should be managed as a pheochromocytoma [1,4,5].

Regional anesthesia may be complicated by obesity and the effect of sympatholysis and dysautonomia on the consequent hemodynamic response (or lack of one).

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## Necessary additional pre-operative testing (beside standard care)

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Evaluation for all associated disorders. This includes an assessment of gastric emptying (gastroparesis: preinduction gastric ultrasound), cardiac abnormalities, (echocardiography to rule out pulmonary hypertension and right ventricular dysfunction) including the presence or absence of a pacemaker. Other concerns include electrolytic abnormalities, seizure disorders, endocrinopathies, behavioral disorders, and possible thermal dysregulation. At the very minimum, the anesthesiologist should perform a detailed history and physical examination to ensure adequate treatment of existing disorders and identify any other conditions which need optimization prior to surgery.

In addition, children should be evaluated for associated disorders that have not been diagnosed. Especially the endocrinopathies, the stress of surgery may uncover unknown and untreated disorders. These include:

- Abnormal hypothalamic-pituitary-adrenal axis
- Central hypothyroidism
- Impaired glucose tolerance or diabetes mellitus
- Diabetes insipidus

In all patients with ROHHAD preoperative pulmonary evaluation including sleep studies (evaluation of central and obstructive apnea and their response to treatment) is strongly recommended) in order to optimize the patient's respiratory status before surgery. When respiratory function measurements and/or sleep studies are abnormal, non-invasive ventilation (NIV) may be indicated and initiated before surgery [6]. Consequently, these patients should be trained in NIV before surgery and assisted with NIV during sedation, regional anesthesia and in the postoperative period.

If a tumor has been identified, the urine should be checked for the presence of catecholamine metabolites because they are a sign of a catecholamine-secreting tumor [1,4,5].

BiPAP has been used to manage nocturnal hypoventilation. Therefore, BiPAP should be available in the immediate postoperative period.

### **Particular preparation for airway management**

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There has not been an increased identified incidence of difficult airway in these patients. There is, however, an increased risk of postoperative apnea. Moreover, morbid obesity and the high incidence of OSA makes these patients high risk for difficult mask ventilation and at risk of hypoxemia during laryngoscopy and intubation. Apneic oxygenation with nasal prongs could be useful to prevent hypoxemia at induction.

Many of these patients already have pre-existing tracheostomies. Considerations for prolonged surgery include switching an uncuffed tracheostomy tube for a cuffed endotracheal tube.

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### **Particular preparation for transfusion or administration of blood products**

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There are no unique blood dyscrasias for this population that have been defined.

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### **Particular preparation for anticoagulation**

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Despite the presence of morbid obesity, there is no indications for anticoagulation in this patient population before adolescence.

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### **Particular precautions for positioning, transportation and mobilization**

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These patients may have tracheostomy tubes or PEG tubes. They are also quite obese, which could lead to compressive neuropathies from improper positioning.

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### **Interactions of chronic disease and anesthesia medications**

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In case of endocrine dysfunction, the usual treatment should be administered on the morning of surgery. A stress dose of corticosteroid may be considered in case of major surgery.

Many of these patients are on anticonvulsants and other psychotropic or mood enhancing medications.

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### **Anesthetic procedure**

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The judicious use of premedication with minimal respiratory effects (e.g., dexmedetomidine), and usual treatment in children with behavioral disorders may be of benefit. These have been used safely [7].

Intraoperatively, the use of inhalational agents is safe in these children. Additionally, the use of intravenous agents with short elimination half-lives and minimal respiratory effects is advisable. Intravenous agents that have been used safely intraoperatively include ketamine and dexmedetomidine. The use of remifentanyl, which is metabolized by plasma esterases, has significant benefit in this patient population due to short lived depressant effects coupled with reliable metabolism.

Non depolarizing muscle relaxants have been used safely. There is no known contraindication to the use of succinylcholine.

As for other obese patients, dosage of IV drugs is best based on calculated lean body weight than on total (actual) body weight [8].

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### **Particular or additional monitoring**

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Due to presence of dysautonomia, using invasive blood pressure monitoring should be considered for invasive or long-lasting procedures.

For the same reason, monitoring of body temperature is mandatory to avoid both hypo- and hyperthermia.

Postoperatively, these children are prone to prolonged apnea and carbon dioxide retention. Therefore, careful postoperative care includes respiratory monitoring with end tidal CO<sub>2</sub> monitoring, blood pressure monitoring for any lability due to dysautonomia, and the judicious use of pain medications to control pain to avoid respiratory depression. It has been suggested that opioids be avoided in order to avoid respiratory depression and that non-opioid pain adjuvant should be used in their entirety to control pain. While this seems intuitively correct, there is not enough data to support this view.

Several of these children are also on non-invasive ventilation such as BiPAP. If the children are on NIV preoperatively, this should be available postoperatively for ventilatory assistance.

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### **Possible complications**

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Most of the complications usually occur when the disorder has not been diagnosed yet in the differential diagnosis of prolonged postoperative apnea. Blood pressure lability from dysautonomia is also a possible complication. Careful attention to thermal dysregulation should also be monitored by use of temperature measurement devices.

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### **Post-operative care**

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Patients who are on non-invasive ventilation prior to surgery should continue their ventilation postoperatively. PICU admission should be strongly considered to closely monitor postoperative respiratory parameters.

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### **Disease-related acute problems and effect on anesthesia and recovery**

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Prolonged postoperative apnea needs to be distinguished from prolonged anesthetic medication effect, which is very difficult in practice. Hence the suggestion of the use of medications with minimal respiratory depressant effects (e.g., remifentanyl) and short half-lives to minimize the probability of the latter.

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### **Ambulatory anesthesia**

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Although there is no data to suggest that ambulatory anesthesia is intrinsically dangerous in these patients, the nature of the disease does suggest the patients should be hospitalized for at least 24 hours postoperatively to minimize any lingering medication related issues.

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### **Obstetrical anesthesia**

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No data exists currently on this subject.

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**This recommendation was prepared by:**

**Author**

**Arvind Chandrakantan**, MD, MBA, MS: Professor of Anesthesiology & Pediatrics, Texas Children's Hospital/Baylor College of Medicine, Houston, TX 77030  
[arvind.chandrakantan@stonybrookmedicine.edu](mailto:arvind.chandrakantan@stonybrookmedicine.edu)

*Disclosure The authors have no financial or other competing interest to disclose. This recommendation was unfunded.*

**This recommendation was reviewed by:**

**Reviewer 1**

**Fabrizio Racca**, Anaesthesiologist, Ospedale Pediatrico ASO Alessandria, Italy  
[fabrizio.racca@gmail.com](mailto:fabrizio.racca@gmail.com)

**Cesare Gregoretti**, Anaesthesiologist, ASO CTO-CRF-Maria Adelaide, Italy  
[c.gregoretti@gmail.com](mailto:c.gregoretti@gmail.com)

**Reviewer 2**

**Hui Ben Chew**, Clinical Geneticist, Kuala Lumpur Hospital, Malaysia  
[hbchew155@hotmail.com](mailto:hbchew155@hotmail.com)

**Editorial review 2019**

Tino Münster, Department of anaesthesiology and intensive care medicine, Hospital Barmherzige Brüder, Regensburg, Germany  
[Tino.Muenster@barmherzige-regensburg.de](mailto:Tino.Muenster@barmherzige-regensburg.de)

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**Author**

**Arvind Chandrakantan**, MD, MBA, MS: Professor of Anesthesiology & Pediatrics, Texas Children's Hospital/Baylor College of Medicine, Houston, TX 77030  
[arvind.chandrakantan@stonybrookmedicine.edu](mailto:arvind.chandrakantan@stonybrookmedicine.edu)

**Reviewer**

**Francis Veyckemans**, Retired pediatric anaesthesiologist and honorary professor UCLouvain Medical School, Ottignies-Louvain-la-Neuve, Belgium  
[veyckemansf@gmail.com](mailto:veyckemansf@gmail.com)

**Editorial Review**

**Christine Gaik**, Anesthesiologist, Department of Anesthesiology and Intensive Care Medicine, University Hospital Giessen and Marburg, Campus Marburg and Philipps University of Marburg, Germany  
[gaikc@med.uni-marburg.de](mailto:gaikc@med.uni-marburg.de)

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