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Congenital Central Hypoventilation Syndrome

orphanaesthesia

a project of the German Society
of Anaesthesiology and Intensive Care Medicine

SUPPLEMENT NR. 6 | 2026

OrphanAnesthesia –
ein krankheitsübergreifendes Projekt des Wissenschaftlichen Arbeitskreises Kinderanästhesie der Deutschen Gesellschaft für Anästhesiologie und Intensivmedizin e.V.

Ziel des Projektes ist die Veröffentlichung von Handlungsempfehlungen zur anästhesiologischen Betreuung von Patientinnen und Patienten mit seltenen Erkrankungen. Damit will OrphanAnesthesia einen wichtigen Beitrag zur Erhöhung der Patientensicherheit leisten.

Patientinnen und Patienten mit seltenen Erkrankungen benötigen für verschiedene diagnostische oder therapeutische Prozeduren eine anästhesiologische Betreuung, die mit einem erhöhten Risiko für anästhesieassoziierte Komplikationen einhergehen. Weil diese Erkrankungen selten auftreten, können Anästhesistinnen und Anästhesisten damit keine Erfahrungen gesammelt haben, sodass für die Planung der Narkose die Einholung weiterer Information unerlässlich ist. Durch vorhandene spezifische Informationen kann die Inzidenz von mit der Narkose assoziierten Komplikationen gesenkt werden. Zur Verfügung stehendes Wissen schafft Sicherheit im Prozess der Patientenversorgung.

Die Handlungsempfehlungen von OrphanAnesthesia sind standardisiert und durchlaufen nach ihrer Erstellung einen Peer-Review-Prozess, an dem eine Anästhesistin bzw. ein Anästhesist sowie eine weitere Krankheitsexpertin bzw. ein weiterer Krankheitsexperte (z. B. Pädiaterin bzw. Pädiater oder Neurologin bzw. Neurologe) beteiligt sind. Das Projekt ist international ausgerichtet, sodass die Handlungsempfehlungen grundsätzlich in englischer Sprache veröffentlicht werden.

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OrphanAnesthesia –
a project of the Scientific Working Group of Paediatric Anaesthesia of the German Society of Anaesthesiology and Intensive Care Medicine

The target of OrphanAnesthesia is the publication of anaesthesia recommendations for patients suffering from rare diseases in order to improve patients' safety. When it comes to the management of patients with rare diseases, there are only sparse evidence-based facts and even far less knowledge in the anaesthetic outcome. OrphanAnesthesia would like to merge this knowledge based on scientific publications and proven experience of specialists making it available for physicians worldwide free of charge.

All OrphanAnesthesia recommendations are standardized and need to pass a peer review process. They are being reviewed by at least one anaesthesiologist and another disease expert (e.g. paediatrician or neurologist) involved in the treatment of this group of patients.

The project OrphanAnesthesia is internationally oriented. Thus all recommendations will be published in English.

Starting with issue 5/2014, we'll publish the OrphanAnesthesia recommenations as a monthly supplement of A&I (Anästhesiologie & Intensivmedizin). Thus they can be accessed and downloaded via www.ai-online.info. As being part of the journal, the recommendations will be quotable. Reprints can be ordered for payment.

Find a survey of the recommendations published until now on:

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Anesthesia recommendations for

Congenital Central Hypoventilation Syndrome

Disease name: Congenital Central Hypoventilation Syndrome

ICD 10: G47.3

Synonyms: Undine Syndrome, Ondine's Curse, Congenital Central Alveolar Hypoventilation Syndrome, Haddad Syndrome (when coupled with Hirschsprung Disease)

Disease summary: Central hypoventilation syndrome (CHS) is a rare disorder, which can be both congenital [1] and acquired [2]. Congenital central hypoventilation syndrome (CCHS) is caused by chromosomal mutations in the PHOX2B gene on chromosome 4p12 [3]. The non-congenital or acquired form of CHS may be due to brain stem tumor, infarct, or edema [4]. As the acquired form of this disease is quite rare, the main focus of this article will be the congenital form of CHS. The main characteristic of CCHS is small tidal volumes and monotonous respiratory rates while awake and asleep, with more profound alveolar hypoventilation during sleep [5]. Due to hypoventilation, these patients develop hypercapnia and hypoxemia but lack the normal ventilatory responses to overcome these conditions while asleep [6]. However, while awake, they do have the ability to consciously alter the rate and depth of breathing. While sleeping, these children exhibit shallow respirations interspersed with periods of apnea most commonly during non-REM sleep [7]. Diagnosis is primarily made by excluding a primary lung, cardiovascular, neuromuscular or brainstem disease that accounts for the clinical picture [8]. CCHS is a lifelong condition and will require some form of ventilatory support throughout life, either positive pressure ventilation via tracheostomy or nasal mask. Other forms of long-term management include negative pressure ventilation and diaphragmatic pacing. CCHS usually manifests itself in the newborn period with episodes of cyanosis and apnea, and most infants will require mechanical ventilation immediately after birth [6]. CCHS can also present in later infancy, childhood and even adulthood and is then termed as Late Onset CCHS (LO-CCHS) [9]. The diagnosis of LO-CCHS should be considered if there is hypoventilation, cyanosis or seizures after administration of CNS depressants or anesthetics, after pulmonary infection, or during treatment of obstructive sleep apnea. LO-CCHS reflects the variable penetrance of PHOX2B mutations [10]. As the PHOX2B gene plays a role in neural crest cell migration, this disease is also linked with other neurocristopathies such as Hirschsprung disease caused by an absence of segmental colonic ganglia.

Hirschsprung disease is present in approximately 20% of patients with CCHS, a combination referred to as Haddad syndrome [10]. Additionally, CCHS may be associated with neural crest-derived tumors such as neuroblastoma, ganglioneuroblastoma, and ganglioneuroma, which occur in 5–10% of affected individuals [7].

Patients with CCHS also commonly experience a range of symptoms related to autonomic nervous system dysfunction (ANSD). These include bradycardia and transient abrupt asystoles, decreased pupillary light response, esophageal dysmotility, breath holding spells,

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reduced body temperature, sporadic profuse sweating and lack of physiological responses to the challenges of exercise and environmental stressors [11].

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

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Emergency information

A	**AIRWAY / ANESTHETIC TECHNIQUE**	Children with CCHS have been described as having box-shaped, short, and flat facial features; however, there are no reports indicating an increased incidence of difficult airways. Some patients may already have a tracheostomy tube in place, which may need to be replaced with a cuffed tracheostomy tube or an endotracheal tube for direct tracheal intubation. In such cases, careful attention must be paid to avoid right mainstem bronchial intubation, as this can exacerbate hypoxia and contribute to the development or worsening of pulmonary hypertension.
B	**BLOOD PRODUCTS (COAGULATION)**	No special considerations for blood products or coagulation.
C	**CIRCULATION**	Cardiac arrhythmias – particularly bradyarrhythmias and transient asystole – may occur intraoperatively. In severe cases, some patients may require the implantation of a pacemaker.
D	**DRUGS**	Preoperative sedatives such as benzodiazepines should be avoided due to their potential to cause respiratory depression. Both intravenous and volatile anesthetics have been used safely, with a preference for short-acting agents. Muscle relaxants may be administered, but neuromuscular monitoring must be employed to ensure complete reversal at the end of the procedure. Atropine and glycopyrrolate should be readily available to manage brady arrhythmias or asystole if they occur. Opioid analgesics should not be excluded from postoperative pain management, as patients typically receive continuous assisted ventilation. However, where feasible, the use of short-acting opioids, non-opioid analgesics (e.g., NSAIDs, acetaminophen), and regional anesthesia is recommended to optimize pain control.
E	**EQUIPMENT**	Ventilatory support may include devices capable of delivering positive pressure ventilation via tracheostomy or noninvasive methods. Additionally, diaphragm pacemakers and cardiac pacemakers – both of which require preoperative programming and postoperative interrogation – should be carefully managed in the perioperative period.

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Typical surgery and procedures

Dental surgery, tracheostomy, insertion of diaphragmatic pacers [4] insertion of cardiac pacemakers, gastrostomy tube insertion, anti-reflux surgical procedures and patients with Hirschsprung disease may require colostomy for distal intestinal obstruction. Due to pulmonary hypertension, some patients may have to undergo cardiac catheterization for right- and left-heart studies.

Type of anesthesia

No existing guidelines or randomized trials have been identified regarding the optimal anesthetic approach, whether general or regional anesthesia.

In the authors' opinion, regional anesthesia should be used whenever appropriate, as it helps to avoid respiratory depression and thereby reduces the risk of prolonged mechanical ventilation. Due to long-standing hypoxia and apnoic episodes, patients with CCHS have inevitably some degree of pulmonary hypertension and consequently right ventricular failure. This may not be the case in patients with LO-CCHS where the disease may have been present for a short time. In patients with severe pulmonary hypertension and right ventricular failure spinal anesthesia should be avoided as it may cause a profound sympathetic blockade, decrease in venous return and bradycardia that can lead to right heart failure. In these patients a carefully titrated lumbar epidural would seem to be the anesthetic of choice.

Peripheral nerve blockade for extremity surgery would have minimal effect on the patient's central respiratory drive and would also minimize the use of opioids for postoperative analgesia.

General anesthesia can be challenging. If required it is important to use short-acting agents e.g., remifentanil, propofol, and desflurane or sevoflurane [4]. Inhalation anesthetics may need to be avoided, as their clearance would depend on adequate postoperative ventilation, which may be impaired in patients with CCHS. Though short-acting muscle relaxants such as succinylcholine have been used for tracheal intubation [12], due to the hypotonia associated with this disorder it would be prudent to avoid succinylcholine until more data is available concerning its use in this patient population. If muscle relaxation is to be used, it is important to detect complete neuromuscular recovery prior to extubation of the trachea [8]. Since the availability of sugammadex, rocuronium can be used more confidently in this patient group due to the potential for rapid and predictable reversal. Due to long-standing disease and the need for mechanical ventilatory support most patients will have a definitive airway in place. In such cases a cuffed tracheostomy tube is required and if no definitive airway is present it is necessary to intubate the trachea, as these patients are at a higher risk of aspiration due to impaired gastric emptying secondary to autonomic dysfunction. Positive pressure ventilation is required as these patients will not breathe spontaneously when asleep.

Necessary additional pre-operative testing (beside standard care)

CCHS is a progressive respiratory disease characterised by hypoventilation and hypoxia. Due to this patients may present with cardiovascular symptoms secondary to pulmonary hypertension.

A chest x-ray should be obtained to rule out pulmonary infection.

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Lung function tests including lung volumes and arterial blood gas analysis should be performed to evaluate grade of pulmonary involvement.

Cardiac function tests, including electrocardiography and transthoracic echocardiography should be performed to evaluate the presence of cardiomyopathy. Invasive tests to determine degree of pulmonary hypertension would include right and left heart catheterization to determine pressures.

Patients may develop polycythemia due to their ongoing hypoxia and a preoperative hemoglobin and hematocrit should be obtained [8].

Seventy-two hour recordings of electrocardiography (Holter monitoring) are required to rule out aberrant rhythms, sinus pauses and the frequency of shorter pauses (< 3 seconds) [13].

Particular preparation for airway management

Due to autonomic dysfunction, patients with CCHS have defective swallowing and decreased gastric emptying. This makes gastroesophageal reflux common in these patients [14]. The laxity of the diaphragm also contributes to lower pressure in the oesophageal sphincter and this decreases the anti-reflux barrier. Therefore drugs are required to reduce gastric acid production and increase gastrointestinal motility.

Particular preparation for transfusion or administration of blood products

Not reported.

Particular preparation for anticoagulation

There is no evidence to support the need of particular anticoagulation. But the impaired mobility and longer bedrest of advanced stage disease patients may suggest a higher risk of postoperative thrombosis.

Particular precautions for positioning, transportation and mobilization

Keep normothermia, oxygenation and normocapnia during transportation. Avoid hypothermia, hypoxia and hypercarbia as these factors can worsen pulmonary hypertension and cause right ventricular failure. Another reason to provide special care to temperature is that patients with CCHS can have problems with the central control of body temperature. Some patients may be wheelchair-bound and rely on bulky ventilatory support devices integrated into their mobility equipment. As these devices are not permitted in the sterile operating room environment, equivalent ventilatory support must be provided using hospital-grade ventilators during transport and throughout the procedure. Patients with diaphragmatic pacemakers often have external transmitters, which should be handled with care to avoid damage or disruption of function [8].

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Interactions of chronic disease and anesthesia medications

If the patient has been receiving respiratory stimulants, i.e. aminophylline or other sympathomimetics, then the inhalation agent halothane should be avoided as it can increase the risk of cardiac arrhythmias.

Provide steroid substitution for patients who are on long-standing oral steroid therapy or have discontinued it within the last 6 months.

Anesthetic procedure

Anesthesia for patients with CCHS should be performed in centres equipped with an Intensive Care Unit.

There has been one reported case in which succinylcholine has been used in a patient with CCHS [12]. Succinylcholine should be avoided in cases where hypotonia is present due to the risk of hyperkalemia and rhabdomyolysis. Since the availabilty of sugammadex, rocuronium can be used more confidently in this patient group due to the potential for rapid and predictable reversal of neuromuscular blockade.

Inhalation agents should be used with caution as they may cause cardiac and respiratory depressant effects. Their clearance depends on adequate postoperative ventilation and a need for postoperative mechanical ventilatory support.

In case of cardiomyopathy, avoid nitrous oxide because of cardiodepressant effects.

Propofol should be used with caution and injected slowly with close observation of the patients electrocardiogram monitoring. This is based on one case report that describes complete atrio ventricular heart block in a child with CCHS who received a bolus of propofol for induction of anesthesia [15].

Opiates and local anaesthetics have been used without any complication. However, short-acting opiates such as remifentanil or fentanyl should be used to avoid prolonged effects.

Non-depolarizing neuromuscular blocking agents can be used safely in these patients, there are reports of the use of rocuronium without adverse effects. Careful monitoring of neuromuscular function should occur to detect complete recovery [8]. However, due to a diminished respiratory drive most patients can tolerate tracheal intubation and ventilation without the need for muscle relaxants.

Antagonisation of neuromuscular blockade with pyridostigmine or neostigmine seems to be possible. However, a case can be made to use sugammadex to completely reverse rocuronium when it is used [8].

Patients must be fully awake to be able to resume spontaneous ventilation, while asleep they will not have a respiratory drive and will be unable to breathe [3].

Due to the respiratory depressant effects of anesthetic agents and opioids it is likely that these patients will require a period of postoperative ventilatory support.

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Particular or additional monitoring

Monitoring of the neuromuscular blockade is strictly recommended if any neuromuscular blocking agent is used: it is useful to obtain baseline values before injection of the non-depolarizing neuromuscular blocking agent.

Monitor body temperature, oxygen saturation and end tidal CO_2 to avoid hypothermia, hypoxia and hypercarbia. These factors can cause pulmonary vasoconstriction worsening pulmonary hypertension and may cause right ventricular failure.

Due to autonomic dysfunction and cor pulmonale arterial cannulation for invasive blood pressure measurement and central venous line placement is recommended. In case of cardiomyopathy, intraoperative transesophageal echocardiography is very useful.

It is imperative that neuromuscular blockade is monitored carefully and its effects are completely reversed after surgery.

Possible complications

In the case where the patient suffers from a seizure disorder then the inhalation agent enflurane should be avoided as it has shown evidence of electroencephalographic spike and wave activity.

At present there is only one reported case concerning the use of succinylcholine in a patient with CCHS [12]. If hypotonia is present, succinylcholine should be used with caution as it may cause hyperkalemia and cardiac arrest [16].

Patients with CCHS are at risk for hyperkalemic cardiac arrest (succinylcholine) and rhabdomyolysis (volatile anesthetics).

Induction with propofol has been shown to cause complete atrioventricular heart block [15].

Sedative drugs and opioids can cause somnolence, which reduces the ventilator drive in patients with CCHS requiring postoperative ventilation.

Muscle relaxants such as rocuronium can be used but adequate monitoring and reversal with suggamadex is required. Residual muscle relaxation can result in prolonged postoperative ventilation and awareness.

CCHS patients are at risk for respiratory and cardiac insufficiency.

Post-operative care

After anesthesia, patients must be provided with rate-controlled ventilatory support with supplemental oxygen. Postoperative monitoring should include electrocardiography and continuous blood pressure. In addition close monitoring of oxygenation and end tidal CO_2 should commence in the postoperative period. Serial arterial blood gas analysis is required.

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Most patients may have an in situ tracheostomy and home ventilation, which can be restarted after surgery.

Disease-related acute problems and effect on anesthesia and recovery

Administration of opioids via the intravenous, intrathecal and epidural route can cause severe respiratory depression in patients with CCHS [8].

In cases where there is severe ANSD complete cardiovascular collapse may occur on the induction of anesthesia. Cardiac dysrhythmias, asystole, convulsions and respiratory arrest may occur.

Patients may require prolonged postoperative ventilation and respiratory monitoring with continuous pulse oximetry in a intensive or high dependency care unit [17].

Diaphragmatic pacing should be turned on after surgery but may not be well tolerated by patients who have had abdominal surgery [8].

Ambulatory anesthesia

Ambulatory anesthesia can be performed on patients with early disease with mild ANSD and in those cases having minimal and minor surgery, only if home ventilation is present.

Obstetrical anesthesia

There are no reported cases of obstetric anesthesia in patients with CCHS. However, the use of epidural anesthesia has shown to be safe with patients with CCHS. Spinal anesthesia may not be safe in patients with advanced CCHS and right ventricular failure in these cases the anesthetic of choice would be a carefully titrated lumbar epidural [18].

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