Aktiv Druck & Verlag GmbH | ISSN 0170 - 5334 | 02330

www.ai-online.info

60. Jahrgang | September 2019

A&I

ANÄSTHESIOLOGIE & INTENSIVMEDIZIN

Offizielles Organ: Deutsche Gesellschaft für Anästhesiologie und Intensivmedizin e.V. (DGAI)
Berufsverband Deutscher Anästhesisten e.V. (BDA)
Deutsche Akademie für Anästhesiologische Fortbildung e.V. (DAAF)
Organ: Deutsche Interdisziplinäre Vereinigung für Intensiv- und Notfallmedizin e.V. (DIVI)

Bronchopulmonary dysplasia

Cerebrotendinous xanthomatosis

orphanaesthesia

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

SUPPLEMENT NR. 11 | 2019

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 Patienten mit seltenen Erkrankungen. Damit will Orphan Anesthesia einen wichtigen Beitrag zur Erhöhung der Patientensicherheit leisten.

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 Anästhesisten damit keine Erfahrungen gesammelt haben, so dass 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 ein Anästhesist sowie ein weiterer Krankheitsexperte (z.B. Pädiater oder Neurologe) beteiligt sind. Das Projekt ist international ausgerichtet, so dass die Handlungsempfehlungen grundsätzlich in englischer Sprache veröffentlicht werden.

Ab Heft 5/2014 werden im monatlichen Rhythmus je zwei Handlungsempfehlungen als Supplement der A&I unter www.ai-online.info veröffentlicht. Als Bestandteil der A&I sind die Handlungsempfehlungen damit auch zitierfähig. Sonderdrucke können gegen Entgelt bestellt werden.

Bisher in A&I publizierte Handlungsempfehlungen finden Sie unter:

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OrphanAnesthesia –

a common 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.

A survey of until now in A&I published guidelines can be found on:

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Anaesthesia recommendations for

Bronchopulmonary dysplasia

Disease name: Bronchopulmonary dysplasia

ICD 10: P27.1

Synonyms: Chronic lung disease of prematurity

Disease summary: Bronchopulmonary dysplasia (BPD) is a chronic lung disease that remains one of the most prevalent long-term sequelae of premature birth. Northway and colleagues first described the condition in 1967, after 32 preterm infants (mean gestational age 34 weeks) with respiratory distress syndrome developed characteristic radiographic changes following the initiation of positive pressure mechanical ventilation [1]. Four stages of lung injury were described in "classic" BPD: exudative (age 1–3 days); necrosis and early repair (age 4–10 days); microcyst formation and pulmonary fibrosis (age 10–12 days); and severe cystic changes and cor pulmonale (after 30 days of age) [2].

The clinical definition of BPD has evolved with time and advancements in neonatal care including surfactant therapy, antenatal steroid administration, and improved ventilator strategies [3,4]. Most infants currently developing BPD are born between 24-28 weeks gestational age, during the time of canalicular and saccular development. "New" BPD is characterised by a uniform arrest of lung development, with simplified alveolar structures and dysmorphic capillaries. These infants are more likely to present with a mild respiratory distress syndrome and a continued need for supplemental oxygen [5]. Three levels of BPD severity (mild, moderate, or severe) are determined by the gestational age of the infant, oxygen dependency at 36 weeks post-conceptual age, total duration of oxygen supplementation, and positive pressure requirements [6].

Long-term BPD survivors may experience persistent airway obstruction and pulmonary hypertension, complicating anaesthetic management.

Medicine is in progress

⚠ Perhaps new knowledge

Every patient is unique

Perhaps the diagnosis is wrong

Find more information on the disease, its centres of reference and patient organisations on Orphanet: www.orpha.net

▶ **Citation:** Vadi M, Malkin M, Lauer R: Bronchopulmonary dysplasia. Anästh Intensivmed 2019;60:S448–S455. DOI: 10.19224/ai2019.S448

Typical surgery

Procedures addressing sequelae of prematurity includes: gastrostomy tube placement; inguinal hernia repair; ligation of patent ductus arteriosus; ophthalmologic procedures; ventriculoperitoneal shunt placement.

Type of anaesthesia

There is no definite recommendation for either general or regional anaesthesia.

Long-term BPD survivors experience airway obstruction and hyperreactivity. It is recommended that BPD patients do not undergo general anaesthesia for elective procedures during an acute respiratory infection; surgery should ideally be postponed for 4–6 weeks. Laryngeal mask airway may decrease bronchospasm risk, but does not protect the airway from aspiration [7]. Regional or local anaesthesia avoids airway manipulation, but may not be feasible for certain sites of surgery or in uncooperative paediatric patients. Avoid spinal anaesthesia in patients with severe pulmonary hypertension and right heart strain, as decreased venous return and bradycardia may precipitate right heart failure.

Necessary additional pre-operative testing (beside standard care)

A thorough medical history must be obtained. For BPD infants requiring surgery while on the neonatal unit, determine the anaesthetic history, current medications, allergies, cough or sputum production, supplemental oxygen requirements, and the extent of positive pressure ventilator support.

Additionally, question parents of BPD patients returning for follow-up procedures after neonatal unit discharge regarding interval hospitalisations (including the need for tracheal intubation), changes in supplemental oxygen requirements, non-invasive positive pressure requirements (ex. CPAP), and exercise tolerance. Poor exercise tolerance may present as diaphoresis or cyanosis during feeding or failure to thrive in patients not yet able to ambulate.

Physical examination should assess vital signs, presence of wheezing or cough, accessory muscle use, cyanosis, and hydration status.

Consider the following testing in patients with persistent disease:

- Electrolyte panel: in patients receiving chronic diuretic therapy
- Arterial blood gas and pulse oximetry: in patients with recent increases in oxygen requirements or in any infant requiring supplementary oxygen
- Chest radiograph
- Echocardiogram: in any infant remaining oxygen-dependent or in patients with clinical markers concerning for pulmonary hypertension such as a continued need for positive pressure ventilation, oxygen requirements out of proportion to the degree of lung disease, elevated $PaCO_2$, recurrent cyanotic episodes, failure to thrive, or recurrent hospitalisations [8].

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- Cardiac catheterisation: reserved for BPD patients with pulmonary hypertension by echocardiogram who (1) have clinical cardiorespiratory deterioration not related to airways disease; (2) are suspected of having significant pulmonary hypertension despite optimal management of lung disease; (3) are candidates for long-term pulmonary hypertension drug therapy; or (4) have unexplained, recurrent pulmonary oedema [9].

Particular preparation for airway management

BPD has no direct correlation with difficult tracheal intubation. However, BPD patients are at risk for subglottic stenosis, airway granulomas, and pseudopolyps if prolonged tracheal intubation occurred during infancy [9–11]. Smaller tracheal tubes may be required.

Particular preparation for transfusion or administration of blood products

Retrospective studies have shown correlations between the receipt of blood transfusions and the development of BPD in very low-birth weight infants [12–14], possibly due to iron-induced oxidative stress [15]. However, there is no clear evidence for a causal relationship.

Patients with known BPD may require diuretics administration in conjunction with transfusion to avoid pulmonary oedema and worsening hypoxaemia.

Particular preparation for anticoagulation

There is no evidence to support the need of particular anticoagulation.

Particular precautions for positioning, transportation and mobilisation

Avoid hypothermia, hypoxia, and hypercarbia as these factors may worsen pulmonary hypertension and precipitate right ventricular failure. Oxygen should be made available for operating theatre transport. Patients requiring positive pressure ventilation should be intubated and ventilated during transport. As ineffective bag ventilation may result in hypercapnoea, consider mechanical transport ventilator use to ensure consistent minute ventilation.

Interactions of chronic disease and anaesthesia medications

Post-natal systemic or inhaled corticosteroids are administered by some centres to reduce inflammation and improve lung function in infants with evolving or established BPD [16–19]. Long-term BPD survivors may receive oral, intravenous, or inhaled steroids as treatment for severe reactive airway disease in the outpatient setting.

All routes of glucocorticoid administration (oral, inhaled, intranasal, topical, intramuscular, and intravenous) have been associated with suppression of the hypothalamic-pituitary-adrenal axis [20]. Physiologic stress including injury, surgery, or severe infection may precipitate adrenal crisis. Consider adrenal suppression in patients with a history of exogenous

steroid use and unexplained hypotension or hypoglycaemia under anaesthesia. Administer stress-dose glucocorticoid supplementation in patients who are on long-standing oral steroid therapy (greater than 8–12 mg/m^2/day for greater than 2 weeks), or those who have discontinued steroid use within the last 6 months [21].

Anaesthetic procedure

Anaesthetic management goals include avoidance of bronchoconstriction, elevations in pulmonary vascular resistance, or decreases in cardiac contractility.

Propofol is a bronchodilator and is a useful induction agent in the haemodynamically stable patient with reactive airway disease. This agent should be used with caution in patients with pulmonary hypertension, as large boluses can significantly decrease systemic vascular resistance and impair biventricular function [22].

Ketamine maintains arterial pressure and systemic vascular resistance while simultaneously enhancing bronchodilatation. It is useful in the haemodynamically unstable and actively wheezing patient requiring emergency surgery. Ketamine administration in children with pulmonary hypertension remains controversial, as some studies suggest its use may increase pulmonary vascular resistance [23–25].

Volatile agents decrease hypoxic pulmonary vasoconstriction. Sevoflurane is preferable for inhalational inductions due to its bronchodilating effects as well as an association with a decreased incidence of laryngospasm and cardiac arrhythmias when compared to other volatile agents. Carefully titrate volatile agents in patients with pulmonary hypertension, as their use may result in a dose-dependent depression of cardiac contractility and systemic vascular resistance [22].

Tracheal intubation may precipitate bronchospasm. Consider avoiding tracheal intubation by using a mask or supraglottic airway device for appropriate cases [7]. Ensure a deep level of anaesthesia before airway manipulation. Deep extubation reduces the risk of bronchospasm from coughing on the tracheal tube but does not protect the patient from aspiration or laryngospasm [22]. Regional anaesthesia avoids airway manipulation but is not appropriate for all sites of surgery.

Avoiding histamine-releasing neuromuscular blocking agents in patients with BPD is advisable. Acetylcholinesterase inhibitors need to be used with caution during reversal of neuromuscular blockade due to the risk of bronchospasm. Sugammadex reverses neuromuscular blockade without muscarinic side effects but has not been extensively studied in the infant and neonatal populations [26].

Particular or additional monitoring

Monitor pulse oxygen saturation, end tidal carbon dioxide, and body temperature to avoid hypoxia, hypercarbia, and hypothermia. These factors may worsen pulmonary hypertension and lead to right heart failure.

Perform arterial blood gas sampling at regular intervals.

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Consider arterial cannulation for invasive blood pressure monitoring and central venous line placement for inotrope administration in patients with pulmonary hypertension for surgical cases of increased length or complexity.

Monitor neuromuscular blockade and ensure its effects are completely reversed after surgery.

Possible complications

Children with BPD frequently experience wheezing, but are less likely to respond to bronchodilators than children with asthma [27,28]. Still, metered dose inhaler administration of bronchodilators is safe and effective for many BPD patients during acute bronchospasm [29]. Refractory bronchospasm may require IV terbutaline or epinephrine administration as a last resort.

In patients with pulmonary hypertension, avoid increases in pulmonary vascular resistance to prevent a pulmonary hypertensive crisis. Moderate hyperventilation with 100% oxygen, correction of acidosis, improved analgesia, and the administration of pulmonary vasodilators are useful in this setting. Inhaled nitric oxide decreases pulmonary vascular resistance without significant systemic effects. Inotropic support is considered for persistent systemic hypotension despite pulmonary vasodilator therapy [30].

Post-operative care

The degree of post-operative monitoring is dependent on the surgical procedure and the physical condition of the patient. Intensive care unit admission may be required for patients experiencing pulmonary hypertensive crisis or requiring mechanical ventilation. Prolonged post-operative ventilation may promote ventilator-associated lung injury and should be avoided.

Disease-related acute problems and effect on anaesthesia and recovery

Not reported.

Ambulatory anaesthesia

Typically avoided. Ambulatory anaesthesia is only considered in BPD patients with mild disease (i.e. no baseline wheezing, cyanosis, home oxygen use, or pulmonary hypertension).

Obstetrical anaesthesia

Not reported.

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Date last modified: January 2015

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

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Disclosures: The reviewers have no financial or other competing interest to disclose.

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Verlag & Druckerei

Aktiv Druck & Verlag GmbH
An der Lohwiese 36 |
97500 Ebelsbach | Deutschland
www.aktiv-druck.de

Geschäftsführung
Wolfgang Schröder | Jan Schröder | Nadja Schwarz
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E-Mail: info@aktiv-druck.de

Anzeigen | Vertrieb
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Verlagsrepräsentanz
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Herstellung | Gestaltung
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Titelbild
Dipl.-Designerin Monique Minde, Nürnberg

Erscheinungsweise 2019
Der 60. Jahrgang erscheint jeweils zum Monatsanfang, Heft 7/8 als Doppelausgabe.

Bezugspreise (inkl. Versandkosten):

- **Einzelhefte** 30,- €
- **Jahresabonnement:**
  Europa (ohne Schweiz) (inkl. 7 % MwSt.) 258,- €
  Schweiz 266,- €
  Rest der Welt 241,- €

  Mitarbeiter aus Pflege, Labor, Studenten und Auszubildende (bei Vorlage eines entsprechenden Nachweises)
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