orphanaesthesia

Anesthesia 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 [3-5]. Most infants currently developing BPD are born at less than 29 weeks gestational age, during the time of canalicular and saccular development [5,6]. "New" BPD is characterized by 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 [5-7]. Three levels of BPD severity are determined at 36 weeks post-menstrual age (gestational age plus time elapsed from birth) based on supplemental oxygen and positive pressure requirements [8-10]. Long-term BPD survivors may experience persistent airway obstruction and pulmonary hypertension, complicating anaesthetic management.

Diagnosis may be incorrect; if uncertainty exists, the diagnosis should be re-evaluated.

Every patient is unique; individual circumstances must always guide clinical care.

Medicine is in progress; new clinical knowledge may not be yet reflected in this guideline Perhaps new knowledge.

Recommendations are not rules or laws; they provide a framework to support clinical decision-making. Although this recommendation has passed a structured review process, it does not meet the formal criteria of a guideline.

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

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

A	**AIRWAY / ANESTHETIC TECHNIQUE**	No direct correlation with difficult tracheal intubation. Risk of subglottic stenosis, airway granulomas, and pseudopolyps if prolonged intubation occurred during infancy. No definite recommendation for either general or regional anesthesia.
B	**BLOOD PRODUCTS (COAGULATION)**	Tailor preparation and administration of blood products to the specific surgical procedure. No associated coagulation disorders.
C	**CIRCULATION**	Anesthetic risk is increased in BPD patients with pulmonary hypertension. Avoid hypoxemia, hypercarbia, and acidosis. Inhaled nitric oxide and cardiac inotropes may be required during a pulmonary hypertensive crisis.
D	**DRUGS**	Glucocorticoid administration for reactive airway disease may cause adrenal suppression. Assess the need for stress-dose steroid supplementation. Diuretic use may lead to electrolyte imbalances.
E	**EQUIPMENT**	Oxygen should be available for operating theatre transport. Consider mechanical transport ventilator use for intubated patients requiring positive pressure ventilation to ensure consistent minute ventilation.

Typical surgery and procedures

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

Type of anesthesia

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

Long-term BPD survivors experience airway obstruction and hyperreactivity. It is recommended that BPD patients not undergo general anesthesia for elective procedures during an acute respiratory infection; patients should be re-evaluated at least two weeks after full symptom resolution [11]. Existing pulmonary morbidity and procedural needs will determine appropriate airway management (i.e., mask, laryngeal mask airway, or endotracheal tube) [13]. Laryngeal mask airway may decrease bronchospasm risk but does not protect the airway from aspiration [14]. In children with severe BPD and low lung compliance, endotracheal intubation and mechanical ventilation may be required regardless of procedure type [13].

Regional or local anesthesia avoids airway manipulation but may not be feasible for certain sites of surgery or in uncooperative pediatric patients. Avoid spinal anesthesia 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 anesthetic 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 hospitalizations (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: screening for pulmonary hypertension by transthoracic echocardiography is indicated 1) if severe respiratory compromise is present in any preterm infant less than 28 weeks' gestation; 2) in any infant with established BPD at 36

weeks post-menstrual age and before hospital discharge; 3) in any infant with prolonged oxygen requirement, poor growth, or unsatisfactory clinical improvement [15].

- Cardiac catheterization: reserved for BPD patients with pulmonary hypertension by echocardiogram who have (1) unexplained hypoxic respiratory failure or recurrent pulmonary edema; (2) pulmonary hypertension out of proportion to echocardiographic findings; (3) pulmonary hypertension requiring multiple or chronic pulmonary vasodilator therapy; or (4) the presence of persistent shunt lesions [16].

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 [17-19]. 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 [20-22], possibly due to iron-induced oxidative stress [23]. However, there is no clear evidence for a causal relationship based on a 2023 systematic review and meta-analysis [24].

Patients with known BPD may require diuretic administration in conjunction with transfusion to avoid pulmonary edema and worsening hypoxemia.

Particular preparation for anticoagulation

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

Particular precautions for positioning, transportation and mobilization

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. As ineffective bag ventilation may result in hypercapnia, consider mechanical transport ventilator use for intubated patients to ensure consistent minute ventilation.

Interactions of chronic disease and anesthesia medications

Post-natal systemic or inhaled corticosteroids are administered by some centers to reduce inflammation and improve lung function in infants with evolving or established BPD [5,6,23,25]. 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 [26,27]. 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 hypoglycemia under anesthesia. 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 [28].

Anesthetic procedure

Anesthetic 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 hemodynamically 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 [28].

Ketamine maintains arterial pressure and systemic vascular resistance while simultaneously enhancing bronchodilatation. It is useful in the hemodynamically unstable and actively wheezing patient requiring emergency surgery. Ketamine administration in children with pulmonary hypertension was previously controversial, as some studies suggested its use may increase pulmonary vascular resistance [29-31]. More recent studies show that ketamine has minimal impact on hemodynamics in children with congenital heart disease and/or pulmonary hypertension when used at usual clinical doses [32,33].

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 [28].

Tracheal intubation may precipitate bronchospasm. Consider avoiding tracheal intubation by using a mask or supraglottic airway device for appropriate cases [13]. Ensure a deep level of anesthesia 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 [28]. Regional anesthesia 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 rocuronium- and vecuronium-induced neuromuscular blockade rapidly without muscarinic side effects [34,35].

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. 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, particularly when exposed to stressful conditions such as exercise. Formerly preterm children with persistent obstructive airway disease have a significantly greater response to bronchodilators after exercising, when compared to preterm and term controls with normal lung function [36,37]. Metered dose inhaler or in-line nebulizer administration of bronchodilators is safe and effective for many BPD patients during acute bronchospasm [38]. 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 [28].

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 anesthesia and recovery

As previously mentioned, bronchospasm and pulmonary hypertensive crisis may complicate both intra- and post-operative care. Moderate- to high-risk patients should have anesthetics performed in facilities with experienced pediatric anesthesiologists and a pediatric intensive care unit capable of managing acute pulmonary hypertensive crises.

Ambulatory anesthesia

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

Obstetrical anesthesia

Not reported.

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Date last modified: **December 2025**

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