COHEN SYNDROME
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Cohen Syndrome: Anesthesia

Cohen syndrome presents with multiple clinical manifestations that can be a challenge to the anesthesiologist. These difficulties may include hypotonia, mental disability, difficult airway management, and susceptibility to respiratory infections (1). Common scenarios requiring some form of sedation or general anesthesia include dental and eye procedures. With this in mind, procedures should be overseen by an anesthesiologist with access to advanced airway equipment. Multiple case reports have highlighted specific issues associated with anesthesia (2-3). As with any other patient, a thorough pre-anesthetic evaluation should be undertaken to address these areas of concern prior to any procedure.

Newborns with Cohen syndrome usually will experience decreased muscle tone (hypotonia) that may lead to feeding and breathing difficulties (1).Unlike some other diseases associated with hypotonia, anesthetic agents are not known to be contraindicated in Cohen syndrome. The anesthesiologist may wish to avoid muscle relaxation, which may be unnecessary for tracheal intubation. General anesthesia for patients with hypotonia can be worrisome, as infants may be prone to respiratory failure secondary to weakness of the muscles of respiration. Parents should be alerted to the risk for prolonged intubation following procedures requiring general anesthesia (4).

Developmental delay creates numerous issues, and the anesthesiologist must depend on the caregiver for the entirety of the patient's medical history and consent, regardless of the patient's age. Obtaining diagnostic information, such as routine bloodwork, can be difficult. Assessing the patient's airway may be challenging due to a lack of cooperation (5). Anesthetic induction will often include some form of oral, intranasal, intravenous, or even intramuscular application of premedication. Some degree of parental presence may be helpful in certain situations, but should be left to the discretion of the anesthesiologist.

Patients with Cohen Syndrome present with micrognathia (small mouth), microcephaly (small head), prominent upper incisors and truncal obesity, which all complicate airway management. As mentioned earlier, the patient's mental disability may inhibit the anesthesiologist's ability to do a thorough airway exam. The practitioner should do their best to obtain an adequate airway exam and produce an airway management plan in accordance with the ASA difficult airway algorithm (6). There should be adequate access to advanced airway equipment, which may include fiberoptic intubation or video laryngoscopy.

Individuals may present with mild to moderate neutropenia leading to some patients experiencing repeated respiratory infections (1). Children who undergo general anesthesia with a current or recent upper respiratory infection are at an increased risk of perioperative adverse events. These are typically related to airway hyperreactivity and can include laryngospasm, bronchospasm and hypoxia (low blood oxygen levels). Anesthesiologists recommend waiting 2 to 4 weeks after onset of symptoms to proceed with general anesthesia (7).

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