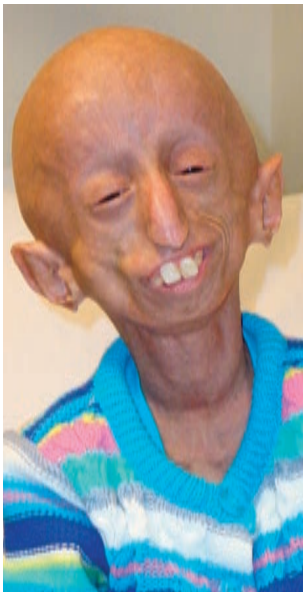
Anesthesia & Airway Management

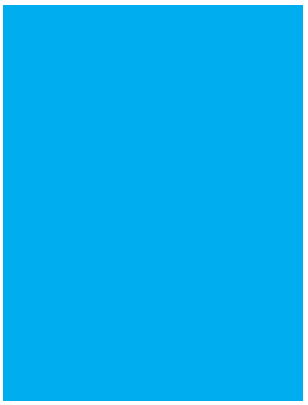


Improvements in the practice of pediatric anesthesia have enhanced the safety of sedation and general anesthesia for purposes of diagnostic, interventional, or surgical procedures in children. Children with Progeria, however, are at higher risk of complications during sedation or anesthesia, related to their challenging airway anatomy as well as to the potential for cardiovascular events. Even an experienced pediatric anesthesiologist may not have had the opportunity to care for a child with Progeria, so this section discusses the special considerations for anesthesia and airway management.

The typical airway features of children with Progeria include the following:

- Mandibular hypoplasia
- Micrognathia and/or retrognathia
- Small mouth opening
- Abnormal dentition (delayed eruption, crowding)
- High-arched palate
- Decreased flexibility of neck and temporo-mandibular joints
- Skeletal contractures and decreased neck mobility
- Decreased subcutaneous fat
- Narrowed nose and small nares

These features may cause difficulty with patient positioning, mask seal, and visualization of the larynx. As such, the clinician must be prepared to utilize techniques for the difficult airway, including laryngeal mask airways (LMAs) and fiberoptic intubation techniques. For children who cannot be intubated by direct visualization, fiberoptic intubation may be necessary. For non-oral procedures, if the procedure can be safely accomplished without endotracheal intubation, use of bag-mask ventilation or an LMA should be considered.



Retrognathia in Progeria: Be prepared to use smaller than expected equipment and endotracheal tube size

Nasal intubation may be challenging due to small nares and unusual glottic angle. Children with Progeria are proportionally smaller for age than their age-matched peers, thus selection of airway equipment sizes may be more accurate based on height than on age. Moreover, there is an increased risk of hypothermia due to alopecia and the paucity of subcutaneous fat.

During sedation or anesthesia, the provider must be aware of the cardiovascular and cerebrovascular disease that characterizes Progeria. Most young children with Progeria have normal ECGs and echocardiograms. As disease progresses, they may develop systemic hypertension, left ventricular hypertrophy, and mitral or aortic valve abnormalities. Unfortunately, studies such as stress tests may not be helpful to predict the risk of intra-operative events.

The coronary and cerebral vasculopathy associated with Progeria results in loss of vessel elasticity and increased risk of cardiac or cerebral ischemic events during states of hypovolemia or hypoperfusion. Children should remain well hydrated prior to and following planned procedures, and medications that may increase myocardial oxygen consumption or produce hypotension should be avoided. Many children are advised to take prophylactic aspirin; the risks and benefits of stopping aspirin therapy prior to planned surgery should be discussed with the surgeon, cardiologist, and/or neurologist involved in the patient's care.