

## 4. Emergency Care / Critical Care

First response

Other considerations



### First response

Children affected by Progeria are at increased risk of more typically adult emergencies such as angina, myocardial infarction, transient ischemic attacks, and strokes. The child with Progeria who presents with chest pain should be assumed to have ischemic heart disease until proven otherwise. Treatment is largely supportive, including supplemental oxygen and IV fluids. If the child is not taking prophylactic aspirin at baseline, he/she should be encouraged to chew a baby aspirin (81mg). In general, avoid medications such as nitrates that can acutely drop blood pressure. Treat pain and anxiety as needed to mitigate the effects of tachycardia on myocardial oxygen demands. If an arrhythmia develops, standard Pediatric Advanced Life Support algorithms are recommended.

The cerebrovascular disease in Progeria can be significant. A history of seizures, severe headaches, or weakness may signify a prior transient ischemic attack or small stroke. Many children who suffer a clinically-recognized stroke are found to have evidence of prior silent ischemic events by MRI. Management of suspected TIA or stroke is largely supportive, such as supplemental oxygen and IV fluids to improve hydration status. Seizures are treated according to usual guidelines for pediatric patients.

*Vascular access is deceiving in children with Progeria. A vein may appear prominent, but be inelastic and difficult to cannulate.*

## Other considerations

Other considerations for children with Progeria with emergency medical conditions include the following:

- **Vascular access:** Although peripheral veins may appear prominent due to the paucity of subcutaneous fat, the vessels are typically less elastic and more difficult to cannulate than they would appear.
- **Bruising:** Children with Progeria may experience significant bruises that are present for long periods of time, even with minor trauma. Large hematomas of the scalp are not uncommon.
- **Joint symptoms:** Joint pain is a common complaint in children with Progeria, especially in the hips and knees. Most joint symptoms can be treated with over-the-counter analgesics; more significant pain should prompt referral to an orthopedic specialist due to the increased risk of hip subluxation and avascular necrosis of the femoral head.