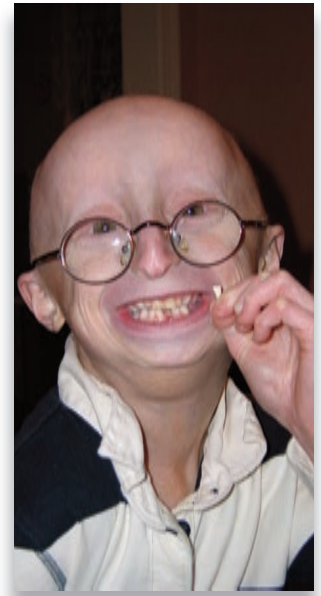


9. Dental Recommendations

For the families

For the medical and dental professional



For the families

There are many dental findings that are prevalent in children with Progeria:

- Crowding of the dentition
- Delayed eruption and/or failure of eruption of baby and adult teeth
- Insufficient space for permanent teeth
- Gum disease
- High rate of cavities
- Small, underdeveloped jaws
- Attrition (wear) of the primary teeth

One of the most important things you can do is establish a relationship with a dentist early in your child's life. By age 1, or by the time your child's first tooth erupts, your child should see a dentist – preferably a pediatric dentist. Due to your child's increased risk for dental disease, it is recommended that your child visit the dentist twice per year, for routine check ups, cleanings, and fluoride treatment, and more frequently if the dentist finds dental issues that need attention. This will not only enable frequent oral assessments, but also help your child feel comfortable in the dental setting.

Delayed eruption of baby teeth is extremely common in Progeria. Secondary teeth may eventually erupt behind primary teeth, but some may never erupt.

For the medical and dental professional

> Typical dental findings in Progeria

- Severe crowding
- Malocclusion
- Ectopic tooth eruption
- Delay and failure of eruption of primary and permanent dentition
- Insufficient space for secondary dentition
 - Tooth size/arch length discrepancies
 - Permanent molars often located in the ramus
- Gingivitis and periodontal disease
- Localized gingival recession
- High caries rate (cavities)
- Attrition of primary dentition
- Hypodontia
 - Agenesis of permanent teeth, especially second premolars
- Ankyloglossia
- Palatal pseudocleft

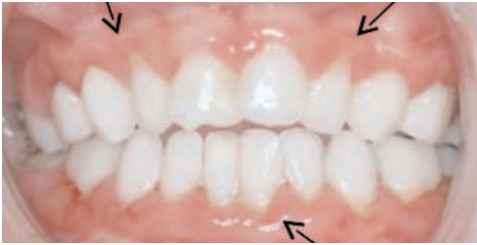
> Craniofacial findings in Progeria

- Alopecia
- Prominent scalp veins
- Perioral cyanosis
- Convex profile
- Limited range of motion
- Hypoplastic maxilla and mandible
- Micrognathia
- Retrognathic maxilla and mandible
- Class II skeletal malocclusion

› Things to consider

- Early visits to the dentist
- More frequent recalls; consider 6-month recalls for exam, prophylaxis, and fluoride treatment
- Establish a relationship with a dentist by age 1 or when first tooth erupts
- Importance of educating parents:
 - High cavities rate in this population
 - Education on oral hygiene instructions
 - Education on etiology of cavities
 - Education on ways to prevent cavities
 - Discourage use of sippy cup and bottle with cariogenic beverages; water only in the cup or bottle
 - Early implementation of fluoride toothpastes, rinses, and in-office application of fluoride
- Orthodontic consideration:
 - Severe crowding and eruption disturbances may necessitate extraction therapy
 - Susceptibility to periodontal disease and limited opening often contraindicates conventional orthodontic therapy

> Figures demonstrating typical dental findings:



Crowded teeth, with areas of gum recession (see arrows)



Cavities



Crowding with malposition of permanent teeth



Attrition (wear) of primary teeth



Attrition of primary central incisors and ectopic eruption of permanent mandibular incisors



Ankyloglossia



Palatal pseudocleft