

Dental Care Challenges in Children with Ataxia Telangiectasia

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Background

Ataxia Telangiectasia (AT) is a rare autosomal recessive neurodegenerative disorder with an estimated incidence of 1 in 88,000 live births in the United States. Caused by pathogenic variants in the ATM gene, AT results in defective DNA repair. Clinical features include progressive cerebellar ataxia, immunodeficiency, radiosensitivity, genomic instability, and a markedly increased risk of malignancy. AT is the only documented condition with a true contraindication to routine surveillance dental radiographs, necessitating strict radiation avoidance. Despite these significant dental implications, guidance in the dental literature remains limited. We describe two preschool-aged children with AT requiring comprehensive dental treatment under general anesthesia to highlight management considerations in this vulnerable population.

Discussion

- Defective DNA repair causes extreme radiosensitivity and increased malignancy risk.
- Routine dental radiographs are contraindicated; treatment-necessary imaging requires physician consultation and informed consent.
- Cumulative lifetime radiation exposure necessitates careful risk–benefit assessment.
- Multisystem disease (immunodeficiency, pulmonary disease) often necessitates hospital-based general anesthesia.
- Antibiotic prophylaxis is not routine and should be determined case-by-case with the managing physician.
- Undiagnosed odontogenic infections may lead to severe or life-threatening complications.
- Preventive care and frequent recall are essential due to limited radiographic monitoring and elevated caries risk.
- Long-term planning should consider future oncologic therapies and oral health implications.

Case 1

Patient: Preschool-aged male

Medical History: Ataxia-telangiectasia, epilepsy, moderate persistent asthma, eczema, recurrent pulmonary infections, and an immunodeficiency disorder

Presentation: Extensive caries across primary dentition

Management: Multidisciplinary consultation (infectious disease and pulmonology), antibiotic prophylaxis recommended

Treatment: Full-mouth rehabilitation under general anesthesia, eight radiographs, two stainless steel crowns, five extractions, two pulpotomies, one composite restoration, and three unilateral space maintainers

Outcome: Previously undetected periapical pathology identified radiographically; 8 year follow up with no clinical decay (patient now in permanent dentition)



Figure 1. Preoperative bitewing radiograph showing primary dentition and furcal spaces for Case 1



Figure 2. Preoperative bitewing radiograph showing primary dentition and succedaneous crowns for Case 2

Case 2

Patient: Preschool-aged male

Medical History: Ataxia-telangiectasia, immunodeficiency, mild persistent asthma

Presentation: Caries affecting primary anterior teeth and lower right first primary molar; referred due to radiograph refusal elsewhere

Management: Immunology consultation confirmed treatment-necessary radiographs acceptable; minimal imaging strategy employed; antibiotic prophylaxis not required

Treatment: Full-mouth rehabilitation under general anesthesia with two bitewing radiographs, anterior resin crowns, stainless steel crowns, extraction of lower right first primary molar, interproximal reduction for cleansibility, and unilateral space maintainer

Outcome: Uneventful recovery with successful completion of care



Figure 3. Pre-operative and Post operative intraoral photo from Case 2

Key Takeaways

1. Ataxia Telangiectasia confers extreme radiosensitivity, making routine dental radiographs contraindicated.
2. Dental care requires multidisciplinary planning, with selective imaging, physician collaboration, and frequent use of hospital-based general anesthesia.
3. Prevention and early clinical detection are essential, as immunodeficiency and limited radiographic surveillance increase the risk of severe infection and future treatment complications.