

# Dental considerations for a patient with Xeroderma Pigmentosum: A Case Report

Andrew Graves DMD

University of Nevada, Las Vegas - School of Dental Medicine

## Introduction:

Xeroderma Pigmentosum is an inherited condition characterized by an extreme sensitivity to ultraviolet (UV) light [1, 2]. This condition is named due to dry skin (xeroderma) and changes to skin color (pigmentation) from sun exposure. Within a few minutes of sun exposure, affected children can develop a severe sunburn [1]. They have an increased susceptibility to Squamous cell carcinomas (SCC) that most commonly occurs on the face, lips, and eyelids [1]. There is no cure for this condition and the main treatment goal is UV avoidance [2]. In Dentistry, light plays a very important role in adequate visualization for diagnosis and treatment as well as the setting and curing of different dental materials. It is recommended that light-cure resins be replaced with glass-ionomer or auto-cure materials [4]. Children with Xeroderma Pigmentosum may experience social and psychological barriers that make it challenging to adapt to their surroundings [3]. This condition may contribute to difficulties in patient management.



Figure 1: Xeroderma Pigmentosum patient viewed affected eyes, face, head, and lips

## Case Report:

A 5 year-old male patient presented to the University of Nevada, Las Vegas Pediatric dental clinic for a comprehensive exam. His chief complaint involved continuous generalized dental pain. Reported medical history includes Xeroderma pigmentosum that is visualized on his face and arms with chronic squamous cell carcinoma lesions on his scalp and forehead that are routinely removed by a dermatologist. There is no history of radiation or chemotherapy. He has no known food or drug allergies and reports taking no medications. Upon initial exam, the patient was unable to tolerate radiographs and clinical findings were documented for occlusal caries on all molar teeth. The comprehensive exam, including radiographs, were planned for the same day of treatment and referred for General Anesthesia at University Medical Center hospital. The patient was instructed to apply sunscreen to arms and face prior to treatment. Precautions were taken by turning off overhead lights and dimming operatory room lights for minimum light exposure. Exposed skin was isolated from light by covering with cloth wraps. On examination, caries was detected on eleven primary teeth. The treatment plan consisted of prophylactic cleaning, fluoride varnish application, therapeutic pulpotomies on #K and L, stainless steel crowns on #A, J, and K, and Sprig Zirconia crowns on #B, I, L, M, N, Q, R, and S. Treatment was influenced by poor hygiene and home care as well as light sensitivity using self-curing glass ionomer cements. After completion of dental treatment, plastic surgery residents removed the SCC lesion on the patient's forehead. The patient was scheduled for a two-week follow-up appointment.

## Discussion:

Long term follow-up on treatment is not available yet due to the recent completion of this treatment, and a six-month follow-up was scheduled. At the patient's two-week follow-up appointment, improvement in behavior was observed. This may be due to a resolved chief complaint considering patient reported no longer experiencing pain and discomfort. On examination, gingival tissue has shown adequate healing and observed good retention and intact full coverage crowns for both zirconia (Sprig) and stainless steel (SSC). No follow-up radiographs were taken due to the nature of a necessary happy visit for the patient.

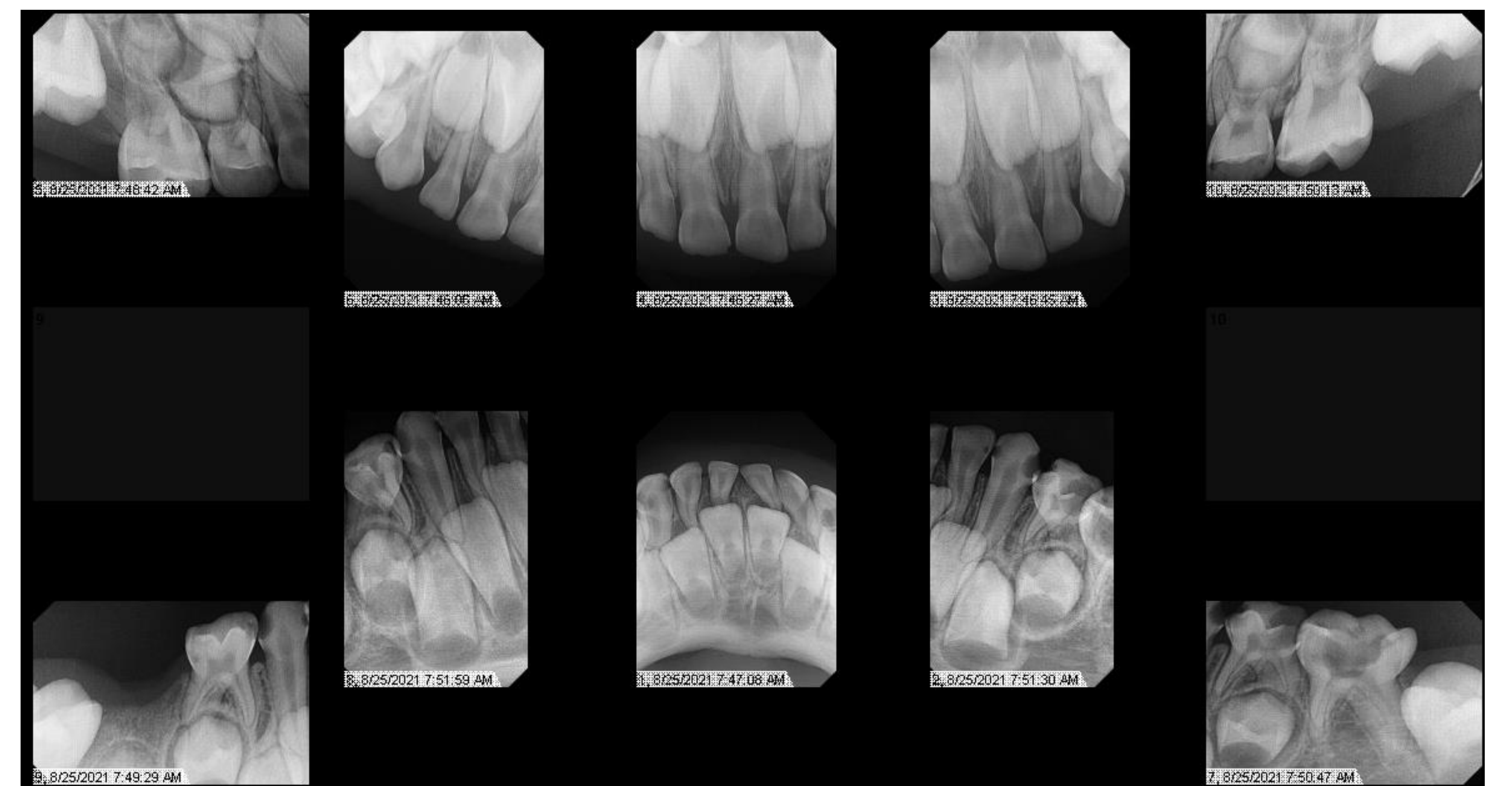


Figure 2: Full Mouth PA's

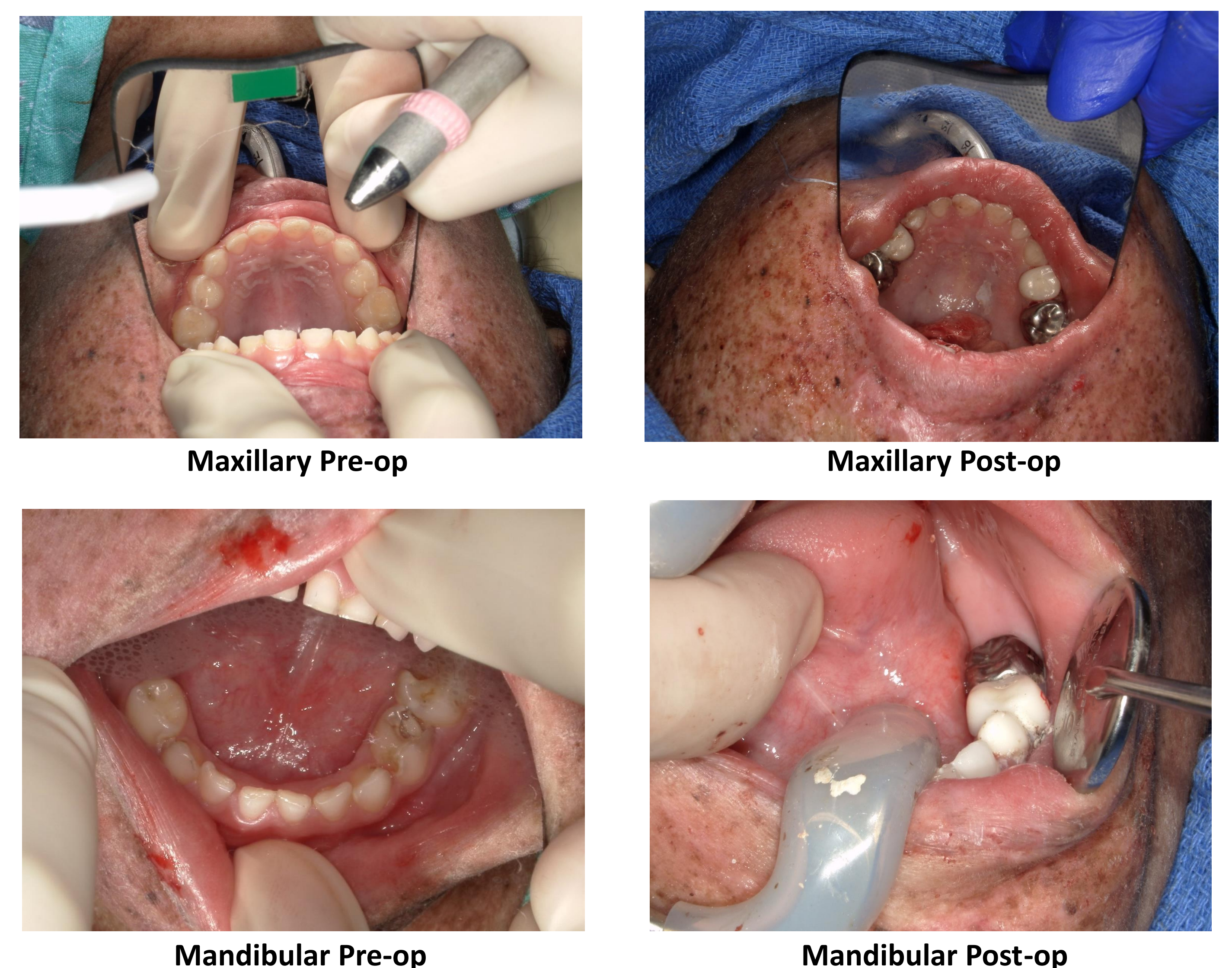


Figure 3: Pre-op and post-op photos

## Summary:

This case is a good example of creative dental care, full coverage restorative treatment, and management of challenging medical history. It also demonstrates a case of hospital dentistry and a circumstance of general anesthesia for a patient with physical and behavioral challenges in need of invasive treatment. It may be advantageous to treat these patients more aggressively with frequent recall appointments and consider the use of other non-invasive procedures including SDF and GI sealants.

## References:

1. Xeroderma pigmentosum. Genetics Home Reference. U.S. Library of Medicine. 26 June 2018. Retrieved 28 June, from, <https://medlineplus.gov/genetics/condition/xeroderma-pigmentosum/>
2. Ngan V. (2002). Xeroderma pigmentosum. [dermnetnz.org](https://dermnetnz.org/). Retrieved 25 February 2020, from, <https://dermnetnz.org/topics/xeroderma-pigmentosum>
3. Pérez-Chávez, D. E., Llópiz-Guerra, K., Núñez Lira, L. A., Garay, J. P. P., Vargas, I. M., & Hernández, R. M. (2020). The Xeroderma pigmentosum syndrome: an experience of integral attention to children with special needs in Cuba. *Revista Latinoamericana de Hipertension*, 15(2), 106-111.
4. Al Wayli H. (2015). Xeroderma pigmentosum and its dental implications. *European journal of dentistry*, 9(1), 145-148. <https://doi.org/10.4103/1305-7456.149664>