Amelogenesis Imperfecta: Genetic Diagnosis and Treatment at UCSF Pediatric Dental Clinic

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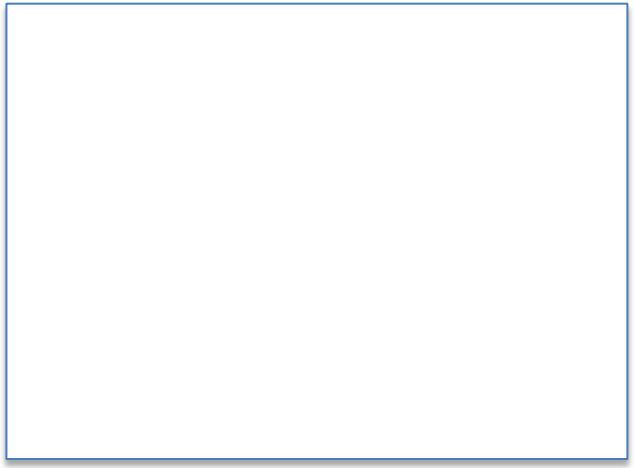
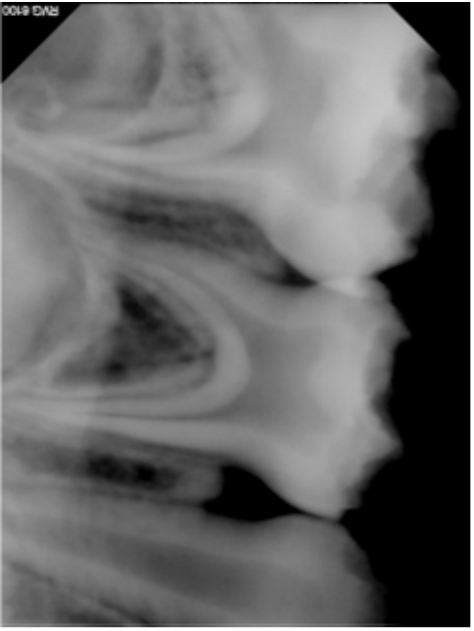
String of Pearls, CSPD 2015
Dana Point, CA

Inherited Enamel Defect in Primary Teeth (3 y.o. female patient)

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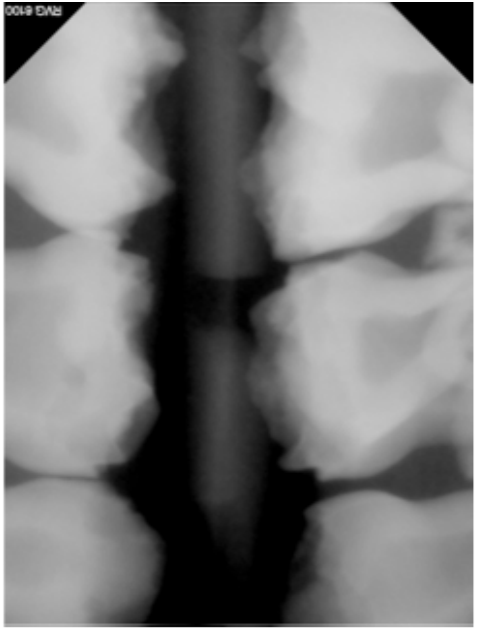
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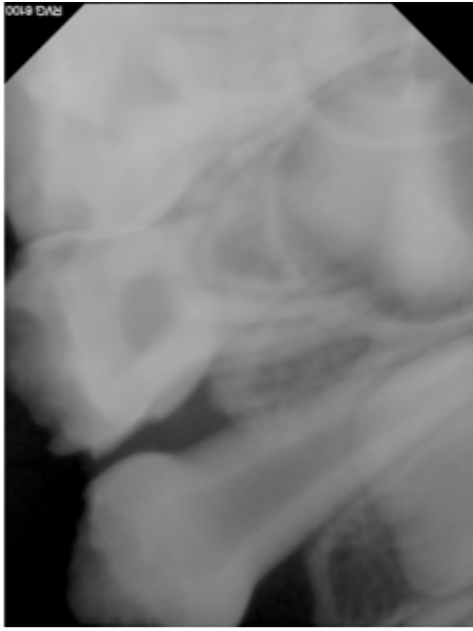
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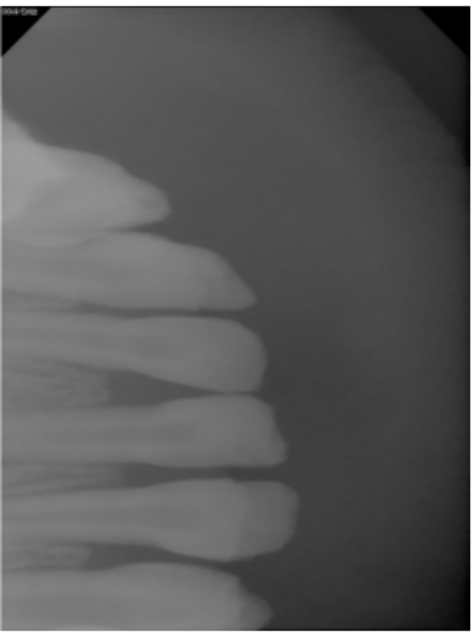
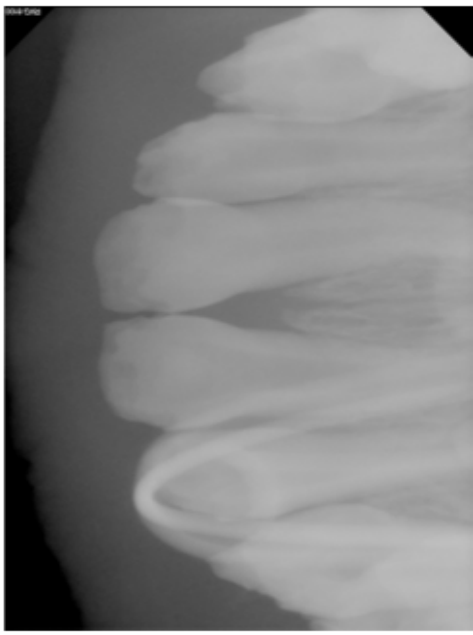
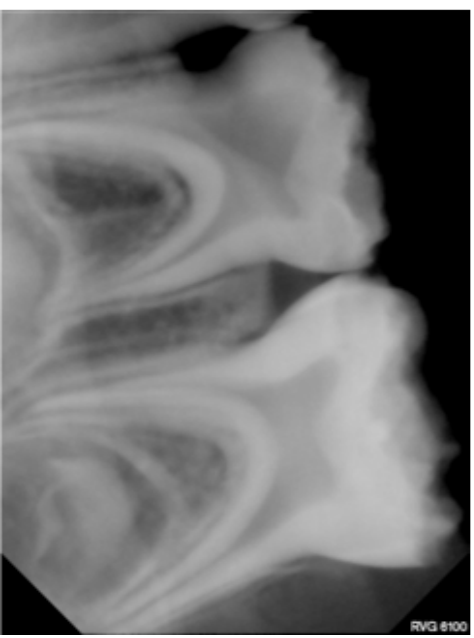
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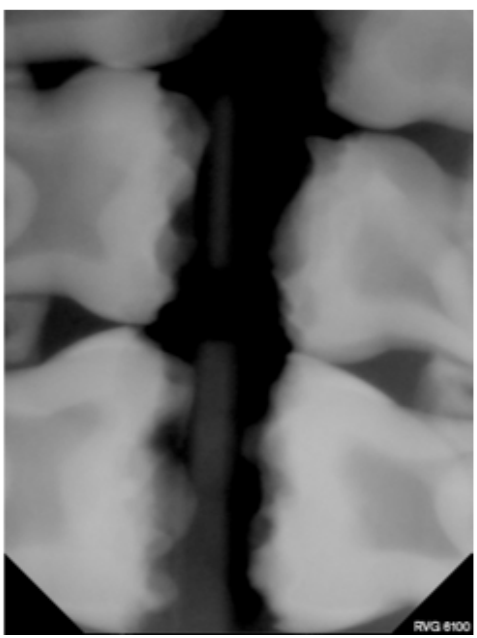
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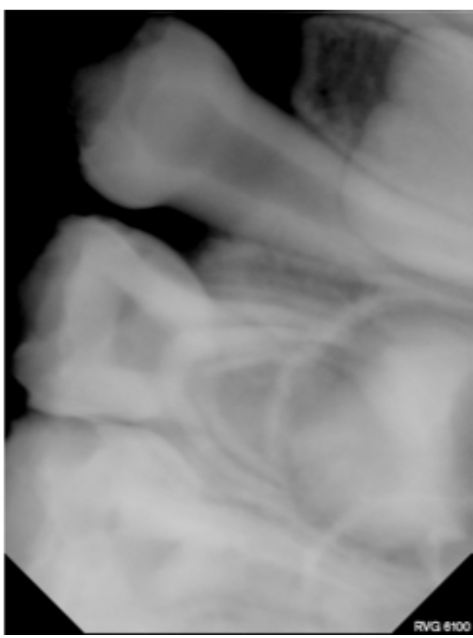
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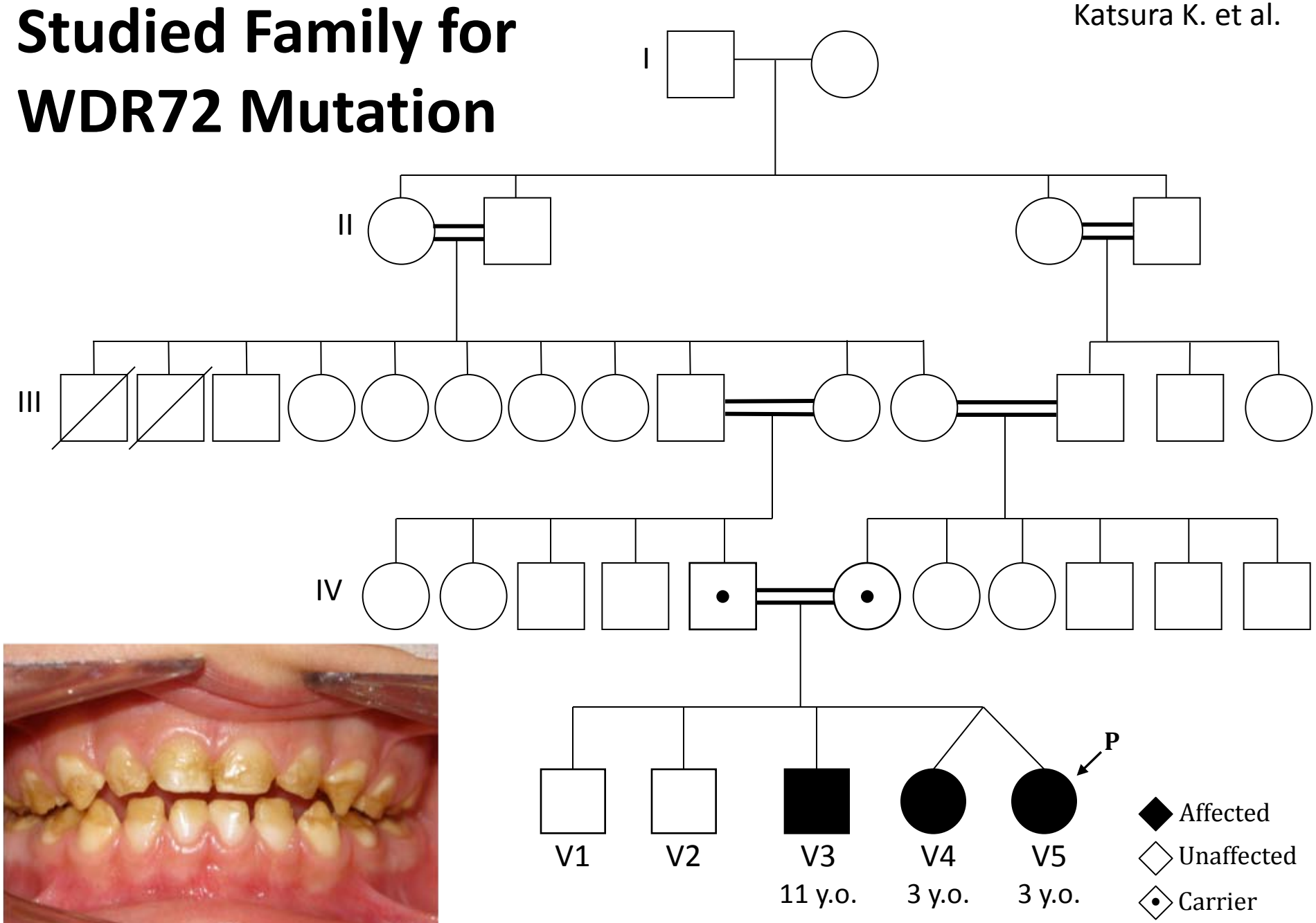
Studied Family for WDR72 Mutation

Katsura K. et al.



Studied Family for WDR72 Mutation

I

II

III

IV

V1 V2 P V3 V4 V5

11 y.o. 3 y.o. 3 y.o.

Affected

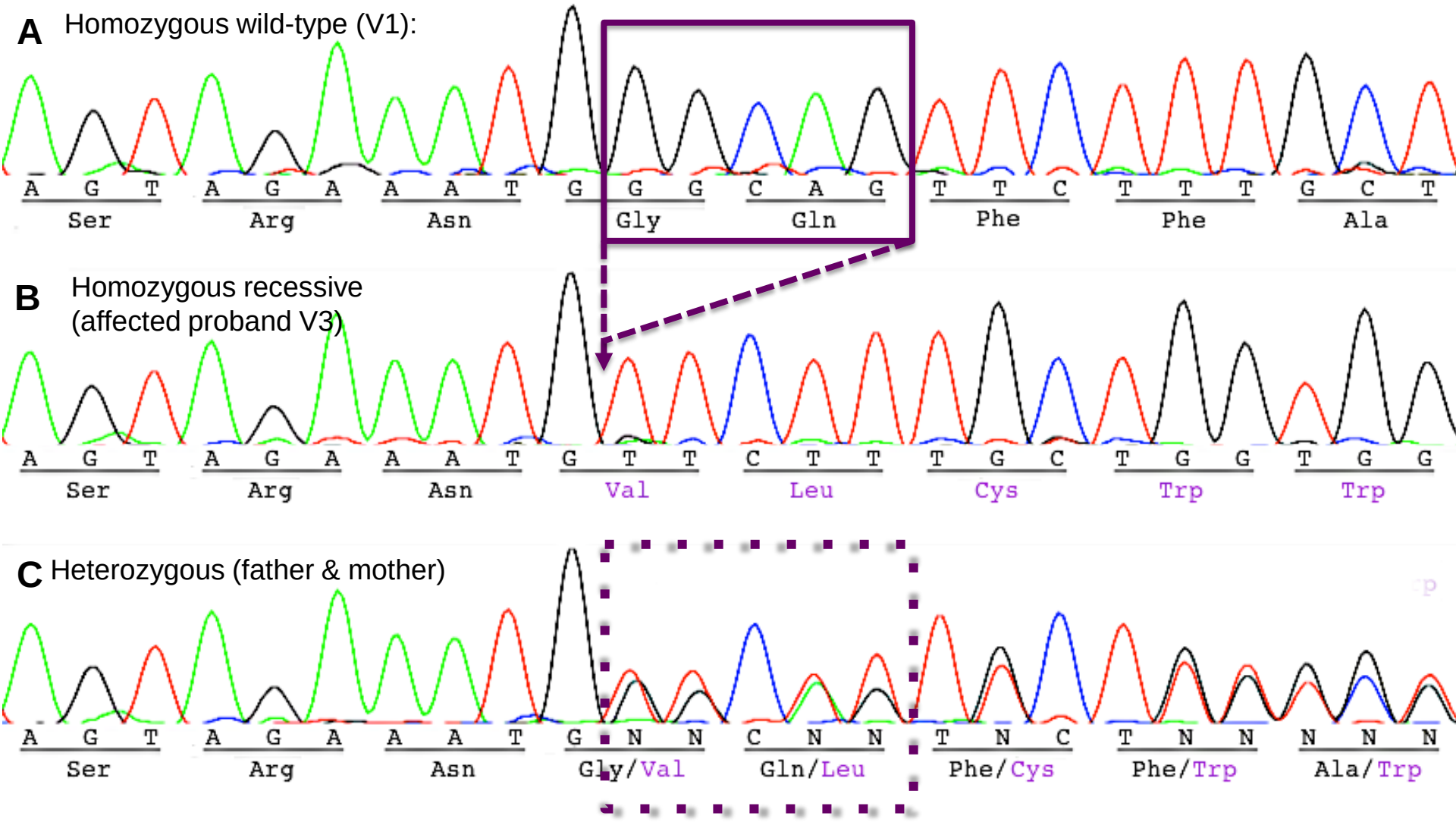
Unaffected

Carrier

Diagnose by Collecting Saliva Samples of Subjects.

ORAgene·DISCOVER
For collection of human DNA

Mutational Analysis of WDR72 Results in Frameshift Mutation (p.G255fsX294)



Post Treatments with Posterior SSCs and Anterior Esthetic Composite Crowns

At 6 y.o., Congential Enamel Defect of 1st Permanent Molars

GI Sealant Placement to Reduce Thermo-Sensitivity

Before GI placement

After GI placement

SSCs for 1st Permanent Molars Soon after Fully Erupted, ~ 7 y.o.

Pt's at 8 years old

Pictures obtained on 3/25/15

Temporary composite veneers were done for partially erupted incisors using self-etch bonding agent Clearfil S3, helping to reduce thermal sensitivity and improved esthetics.

Team of participants

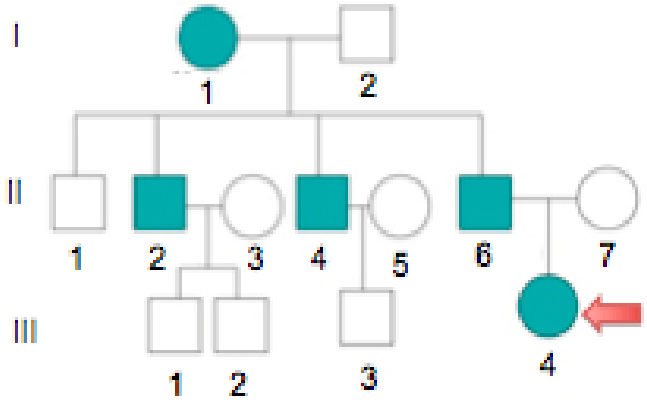
- Kei Katsura (genetic mutation work)
- Dr. Thuan Le
- Dr. Ling Zhan
- Dr. Rung Warotayanont

Family Pedigree and Patient's Clinical Presentation

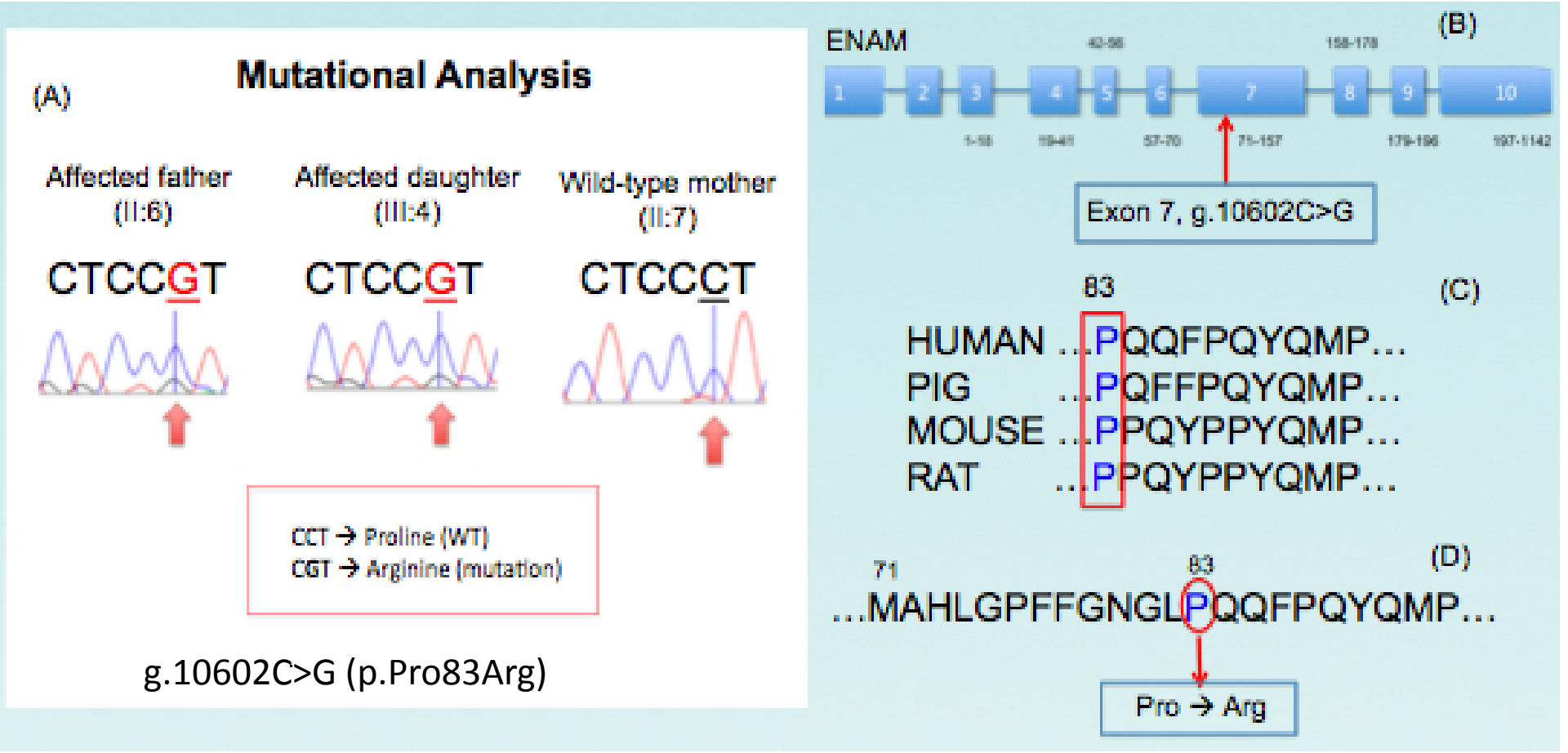
(A)

Family Pedigree

Autosomal Dominant or X-linked Dominant



Mutational Analysis



Clinical management for ENAM mutation, 14 y.o. female

- SSCs for posterior upper and lower premolars and molars
- Fiber-reinforced resin crowns for upper anterior teeth
- Ortho txt completed

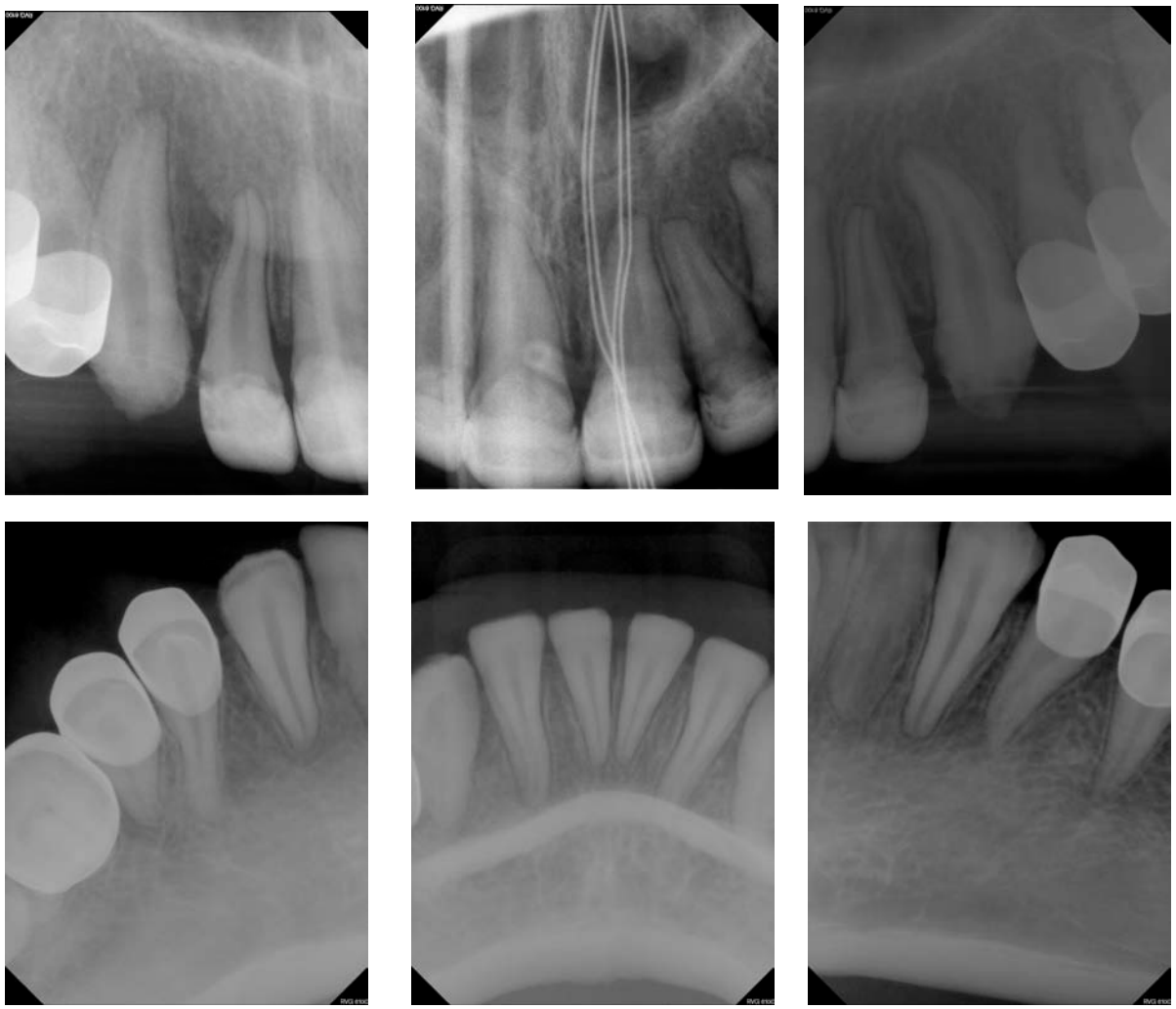
- Planning Cerec crowns using CAD/CAM system for her anterior permanent teeth

Diagnostic wax up model used for provisional crowns fabrication

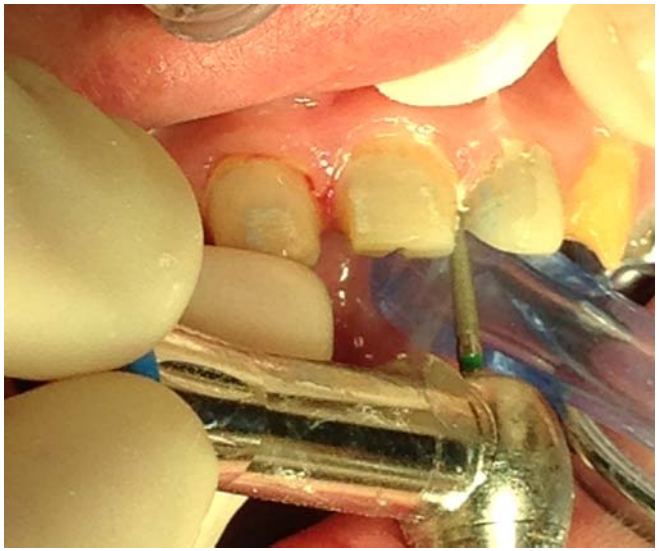
Provisional resin crowns were pre-fabricated

Pre-op photos in O.R.

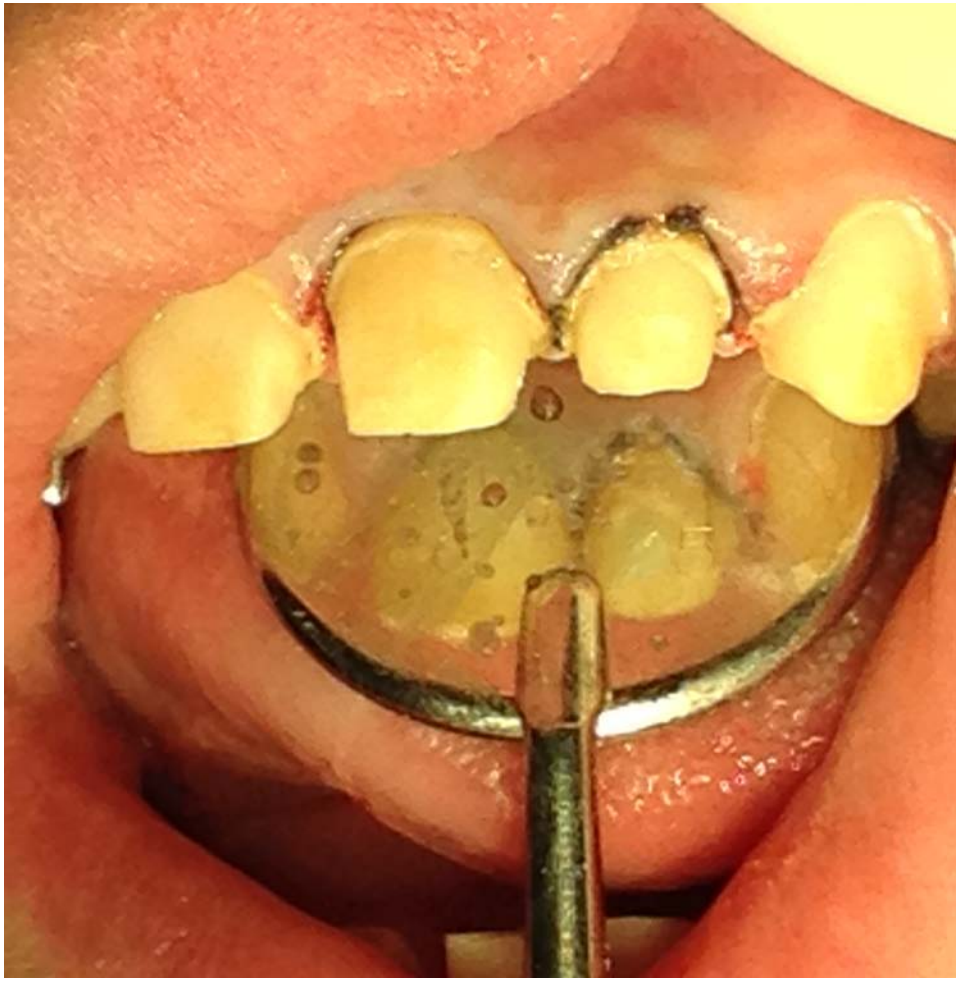
Pre-op PA X-rays for anterior teeth



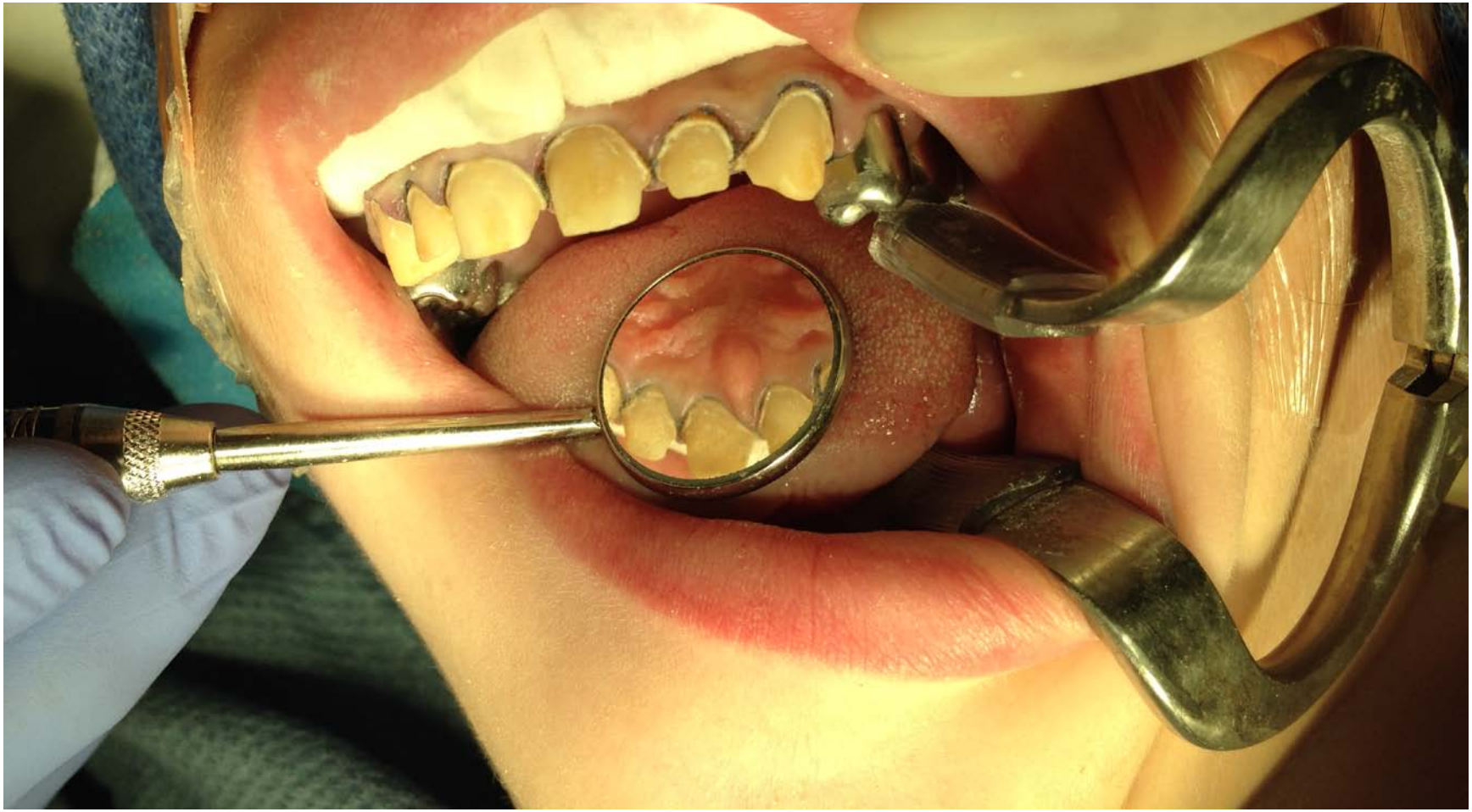
Crowns preps



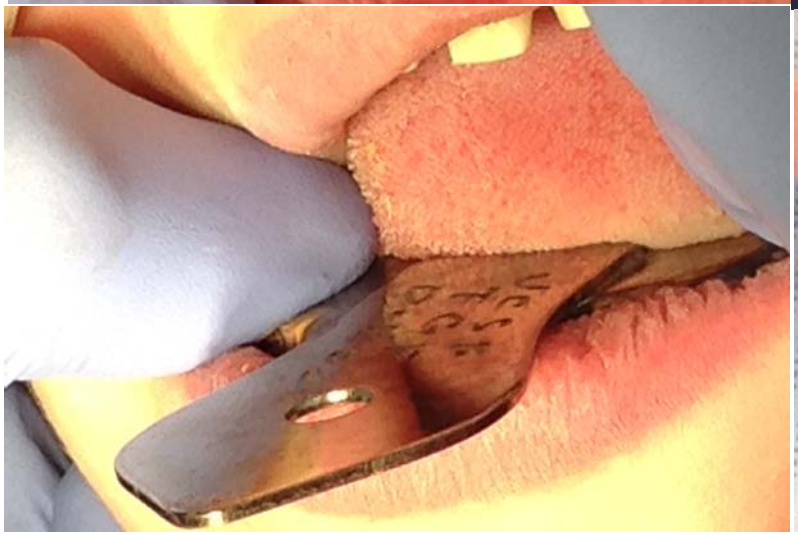
Upper anterior crowns preps and placement of retraction cords for impressions, showing crown shoulder margins



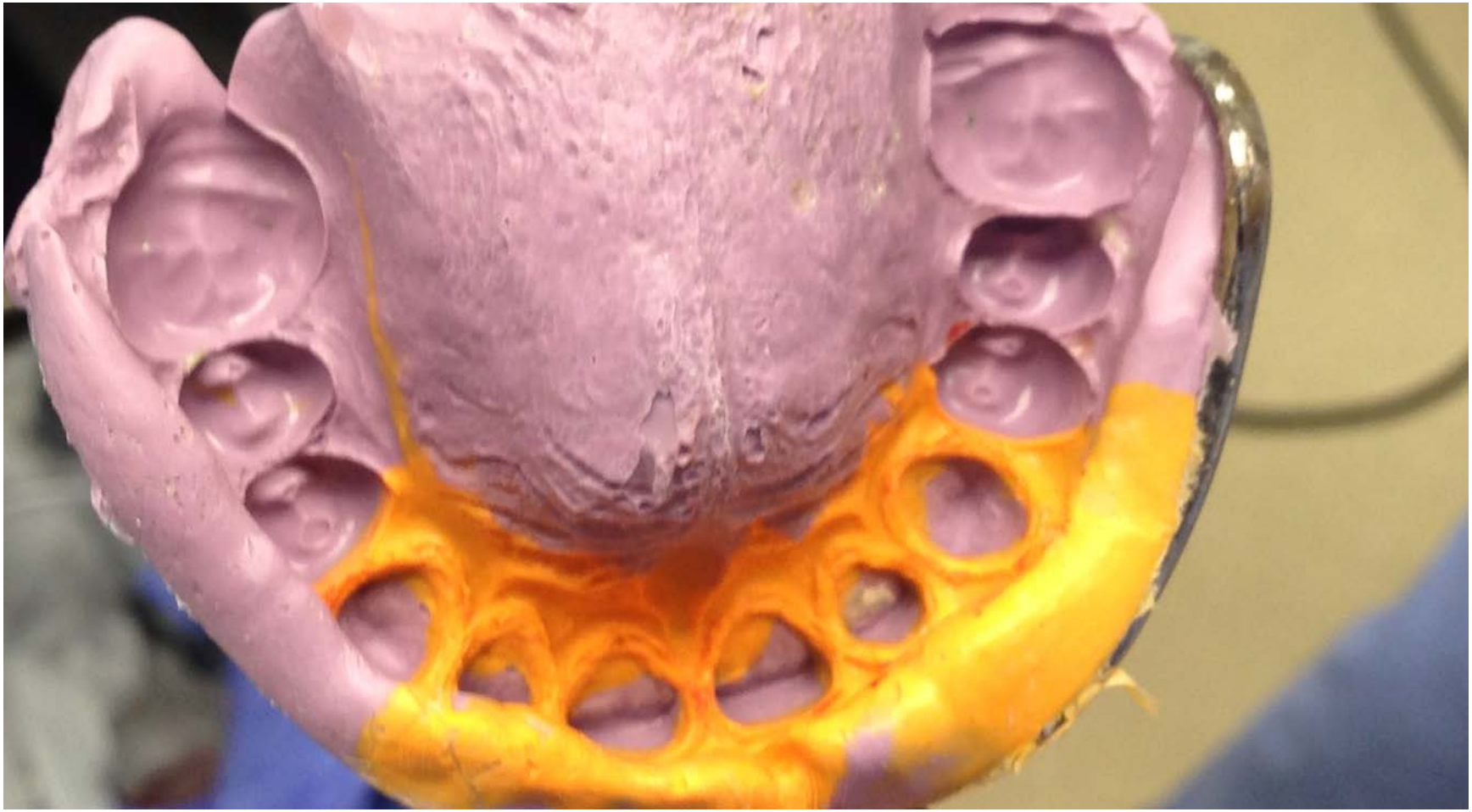
Retraction cords placement



Taking impressions techniques



Upper PVS impression



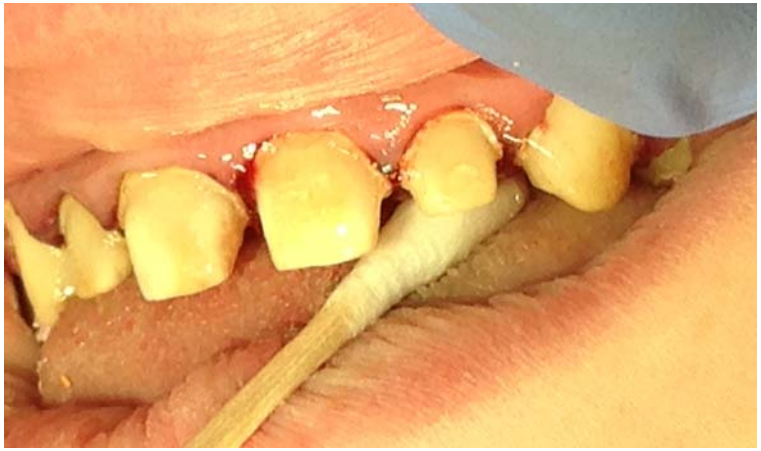
Lower anterior crowns preps

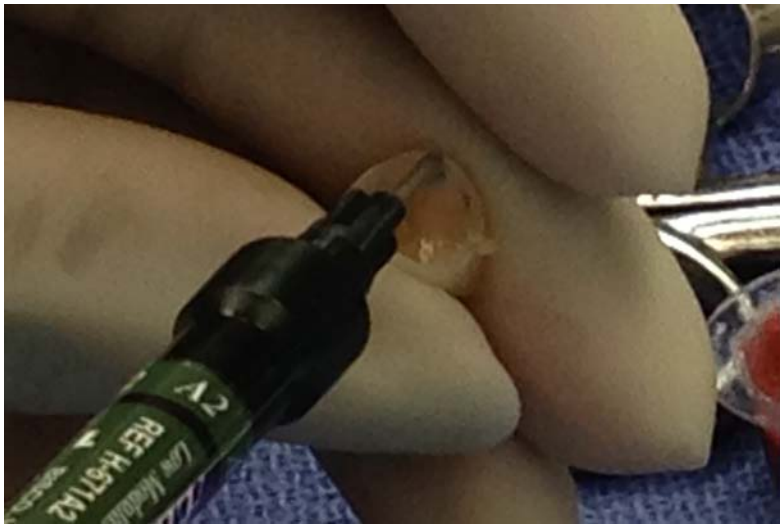
Retraction cords placement, showing shoulder margins for impression

Aluminum chloride used for retraction cords

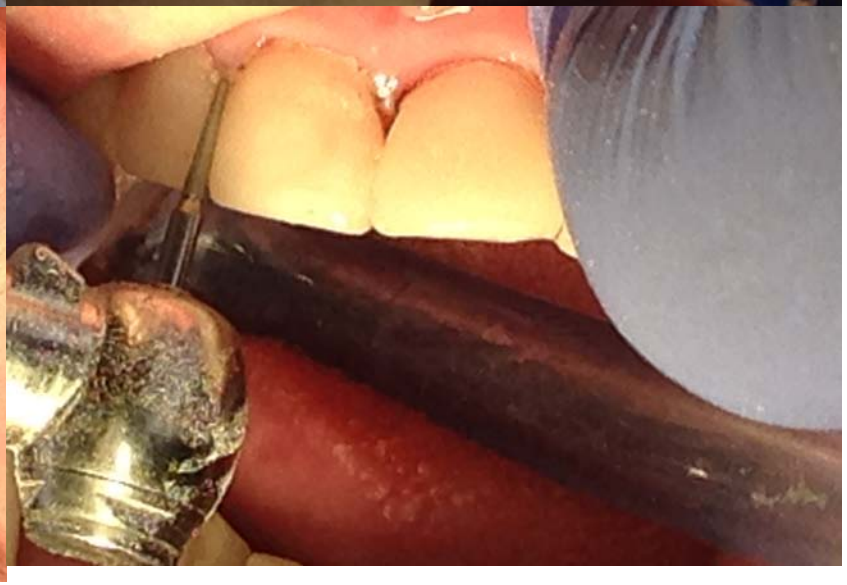
Lower PVS impression

Re-lining provisional crowns with flowable composite





Removing excess flowable composite, light curing, further re-shaping the contour of temp crowns



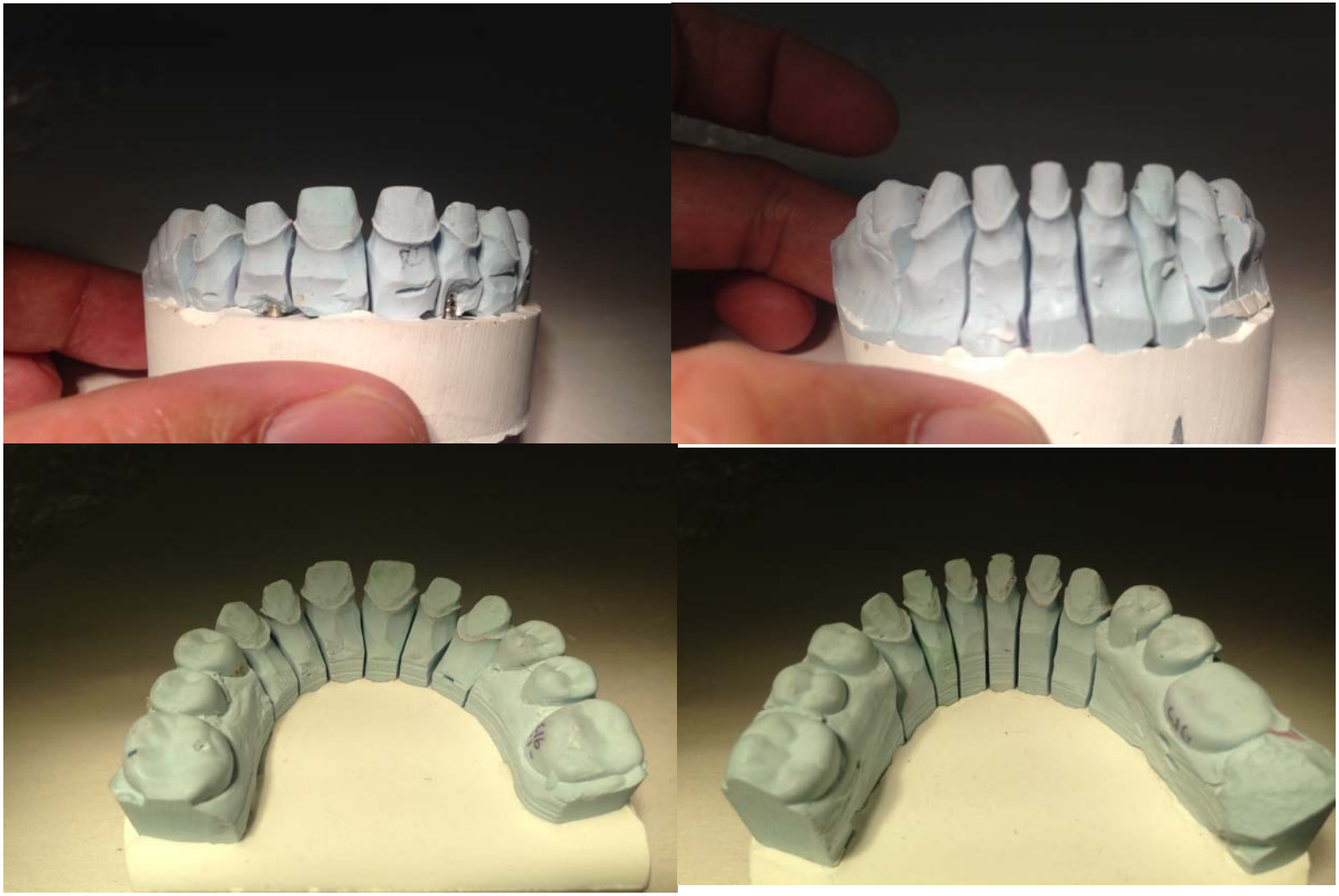
Cementation of temp crowns

Provisional crowns esthetics

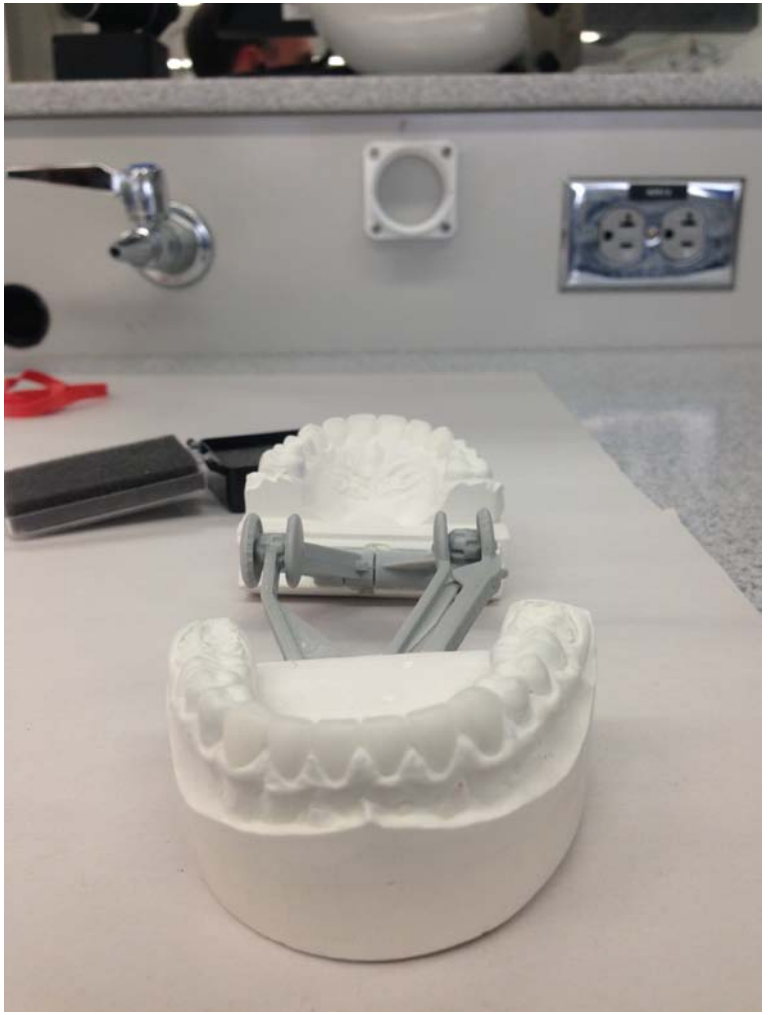
CAD/CAM

- After CAD/CAM milling of final crowns, which are comprised of ~ 85% ceramic and ~15% composite, the final esthetic crowns will be cemented for both upper and lower anterior teeth.
- If chipped or defect detected in the future, crowns can be restored with regular composite resins.

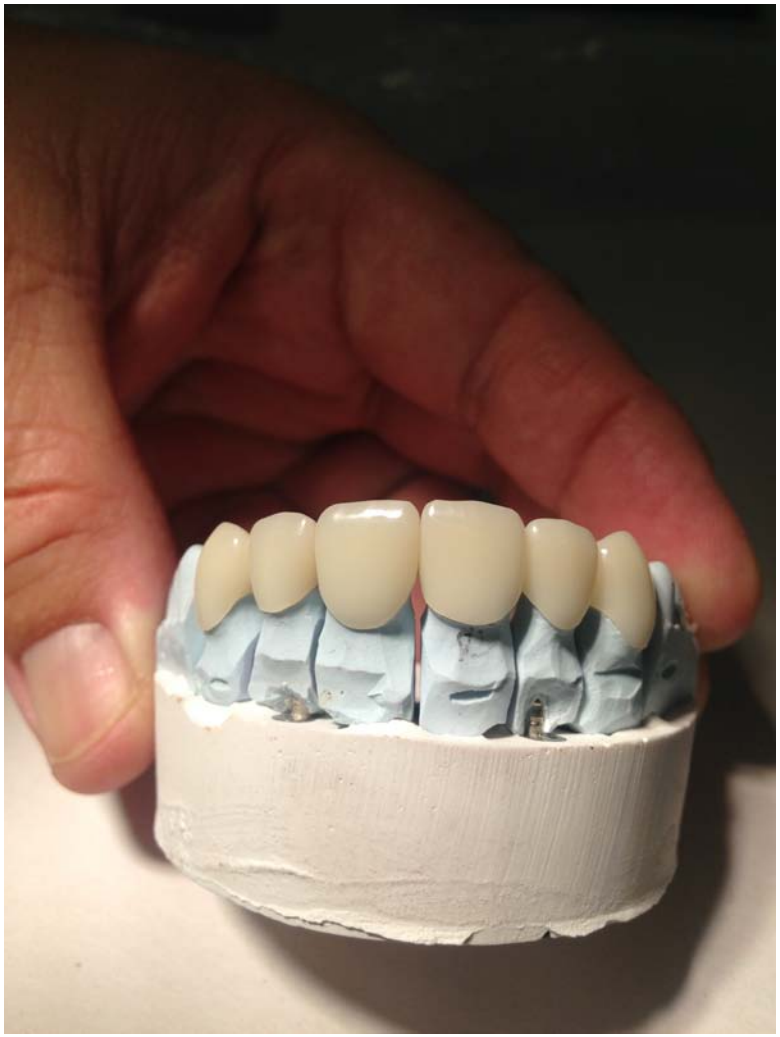
Completed die trimming for upper and lower anterior teeth, showing shoulder margins were captured.



Final provisional wax-ups for CAD/CAM milling procedures



Lab fabrication of CAD/CAM ceramic crowns (85% ceramic + 15% composite



Upper teeth

Lower teeth

Crowns with adequate thickness were evaluate on casts with proper occlusion, and clearance

Prepared teeth after removing temporary crowns

Patient was under minimum oral conscious sedation using 10 mg of Diazepam (2 tabs, 5 mg/tab), and 50% N2O/50% oxygen for anti-anxiety.

Crown cementing system (composite cement)


A
Composite mixing
silanate
Porcelain Etch Gel
9.6% HF
Mix
Multilink® Primer A+B (1:1)
cleaning paste
Flowable composite
B
ivoclar vivadent:

Mixing A + B = self-etched adhesive

Applied for 20 sec, then air dry gently, followed by light cure

Preparation of ceramic crowns for increased bond strength with composite cement

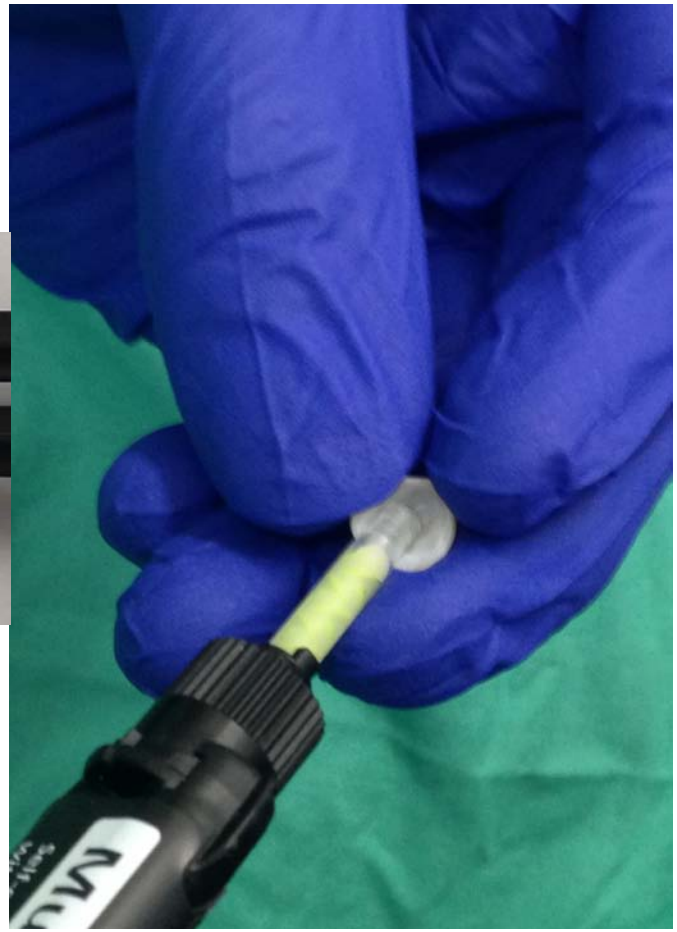
Etch ceramic crown with with 9.6% hydrofluoric acid (HF)

Remove debris post etching

Silanate ceramic crowns to chemically modify surfaces

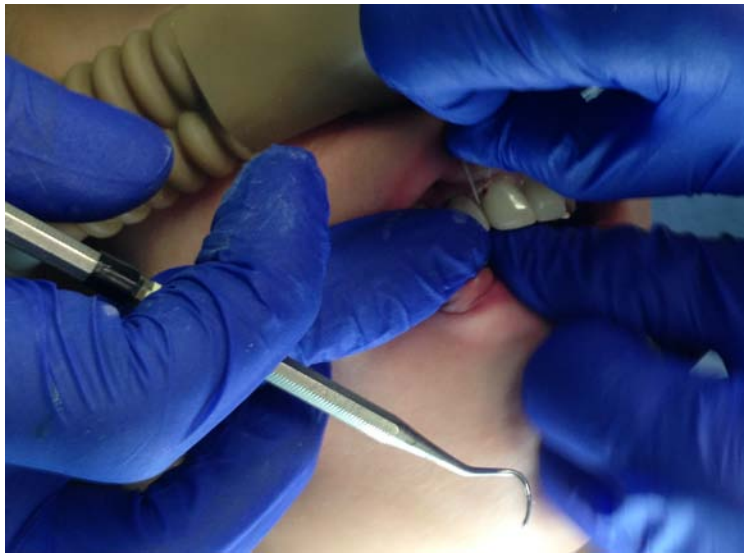
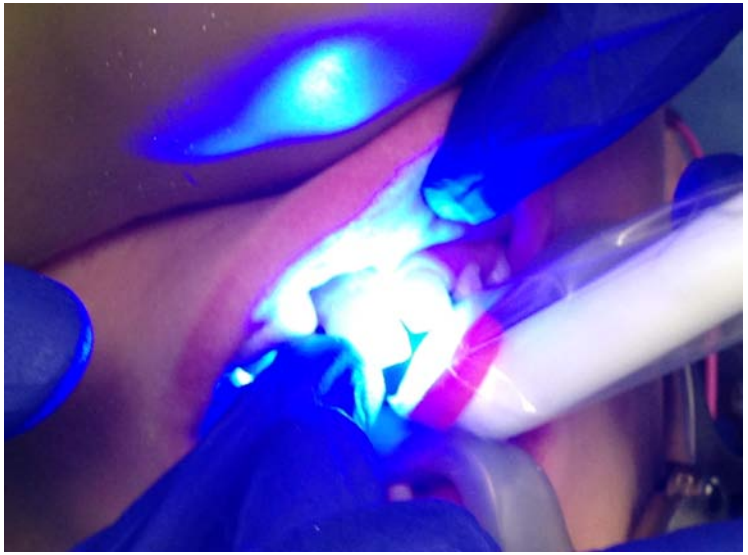
Composite cement was added to chemically-modified ceramic crowns

Composite cement



Crown placement, remove cement excess, and light curing



Flowable composite was added at any opened margins' areas, and removal of fine excess cement

Immediate post-op photo after crowns cementation

Final X-rays

Team of participants

- Dr. Pam Den Besten
- Dr. Sam Huang
- Dr. Thuan Le
- Dr. Jessica Massie (genetic mutation work)
- Dr. Ram Vaderhobli