

Williams Syndrome







European Reference Network

for rare or low prevalence complex diseases

Network

Craniofacial anomalies and ear, nose and throat disorders (ERN CRANIO)

Member

Hôpitaux Universitaires de Strasbourg — France

Clinical case

General dg.: Dental dg.: Williams-Beuren syndrome mixed dentition, with retention of deciduous teeth, microdontia, teeth agenesis

Characteristic features:

- "elfin" appearance, thick lips, saddle nose, anteverted nostrils (blue eyes)
- highly sociable, talkative

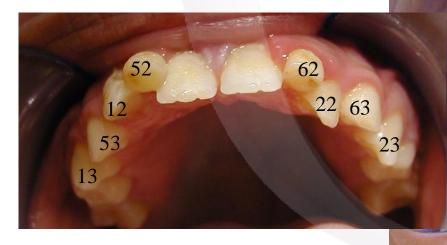
(Dr. Arina Vinereanu)

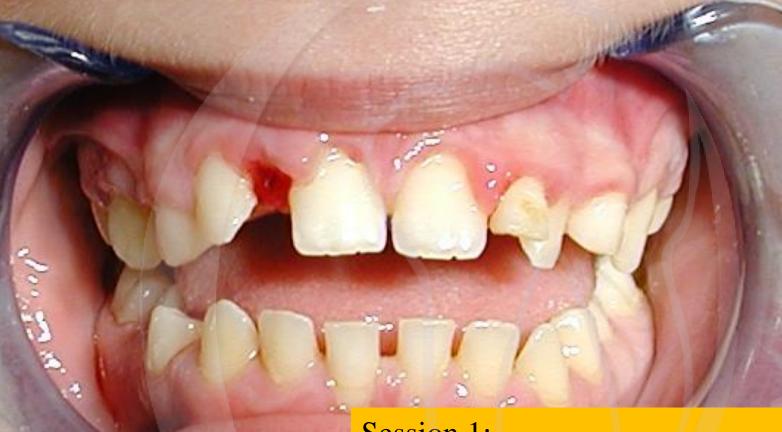
Clinical findings:

 Retention of 6 primary teeth – suspicion of multiple agenesis – Xray needed

(Dr. Arina Vinereanu)

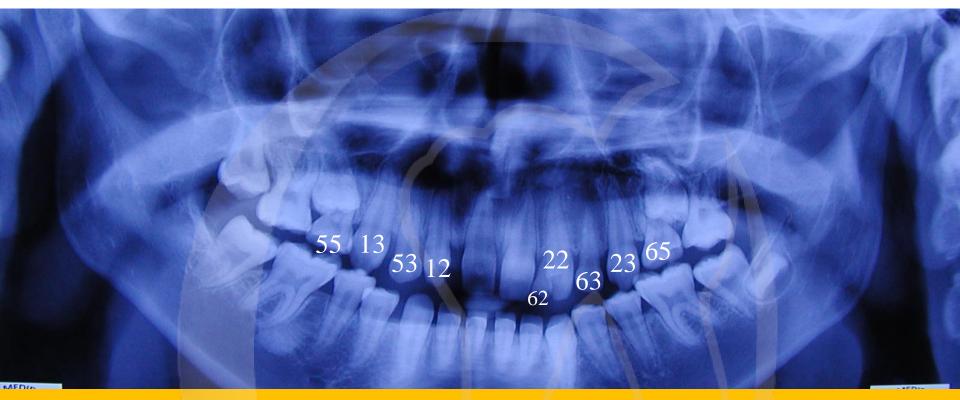
- Malocclusion
- Poor hygiene





Session 1:

- professional cleaning
- extraction of 52 (topical anaesthesia)



- Panoramic Xray (taken after extraction of 52) confirms agenesis (probably 14, 24).
- Hypodontia and microdontia permits the upper deciduous canines to remain on the arch, while one upper premolar is missing on boths sides (14/15?; 24/25?).
- Extraction of 62 and 55, 65 are planned in order to make place for the existing premolars to emerge. 53, 63 shall be kept.
- After extractions patient did not show for follow-ups.