Oral Manifestations of Noonan Syndrome

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Dental Problems in Noonan Syndrome

- Most issues are during childhood/adolescent years
- Tend to normalize in adulthood

Oral Manifestations in Noonan Syndrome

- Structural and Developmental Issues of the Jaws
- Structural and Developmental Issues of the Teeth
- One Jaw Tumor of Significance
- Eruption and Shedding of Teeth
- Malocclusions (teeth don't fit together as they should)
- Enamel Defects

Most Common Oral Findings in Noonan Syndrome

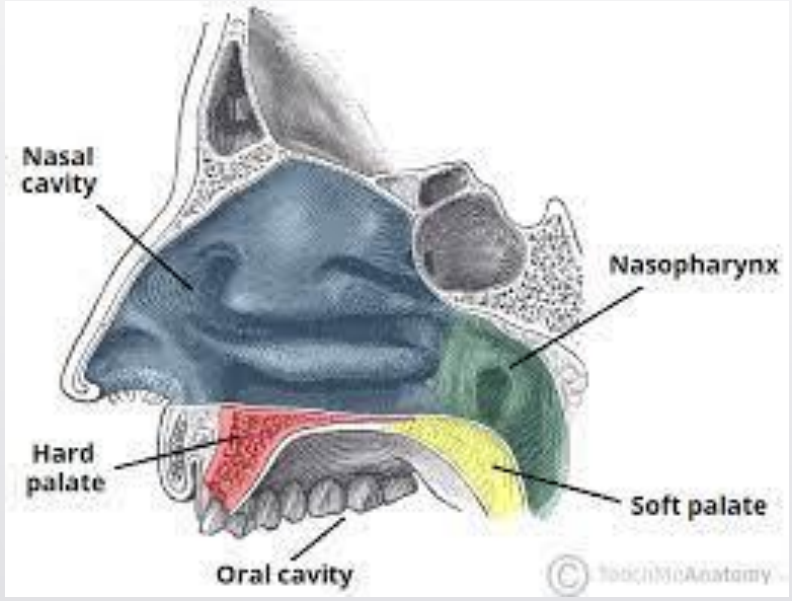
- Crowded Teeth

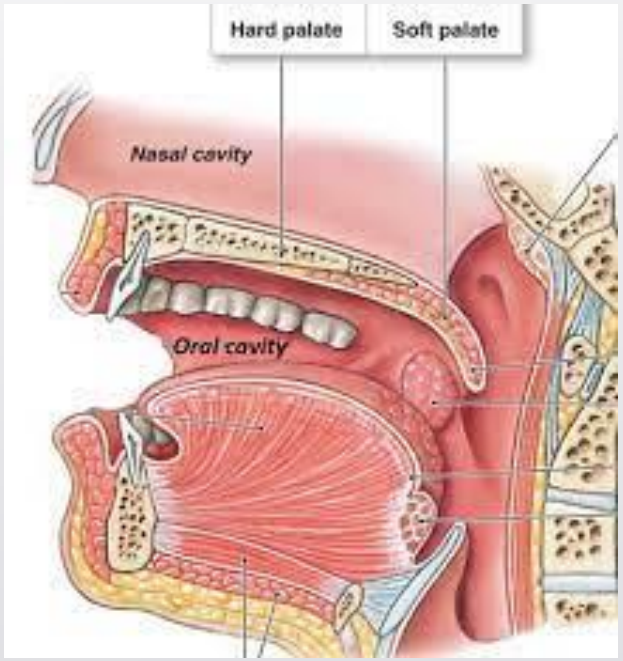
Tooth Crowding in Upper Jaw Versus Lower Jaw

Why do we tend to have crowded teeth in the upper jaw?

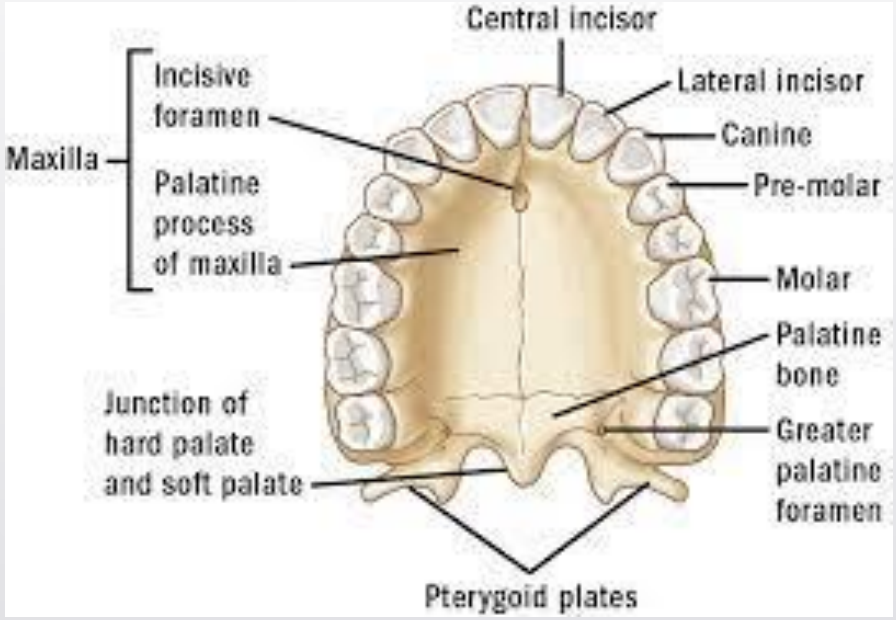
- High (Narrow) Palate

Hard Palate (Roof of Mouth)



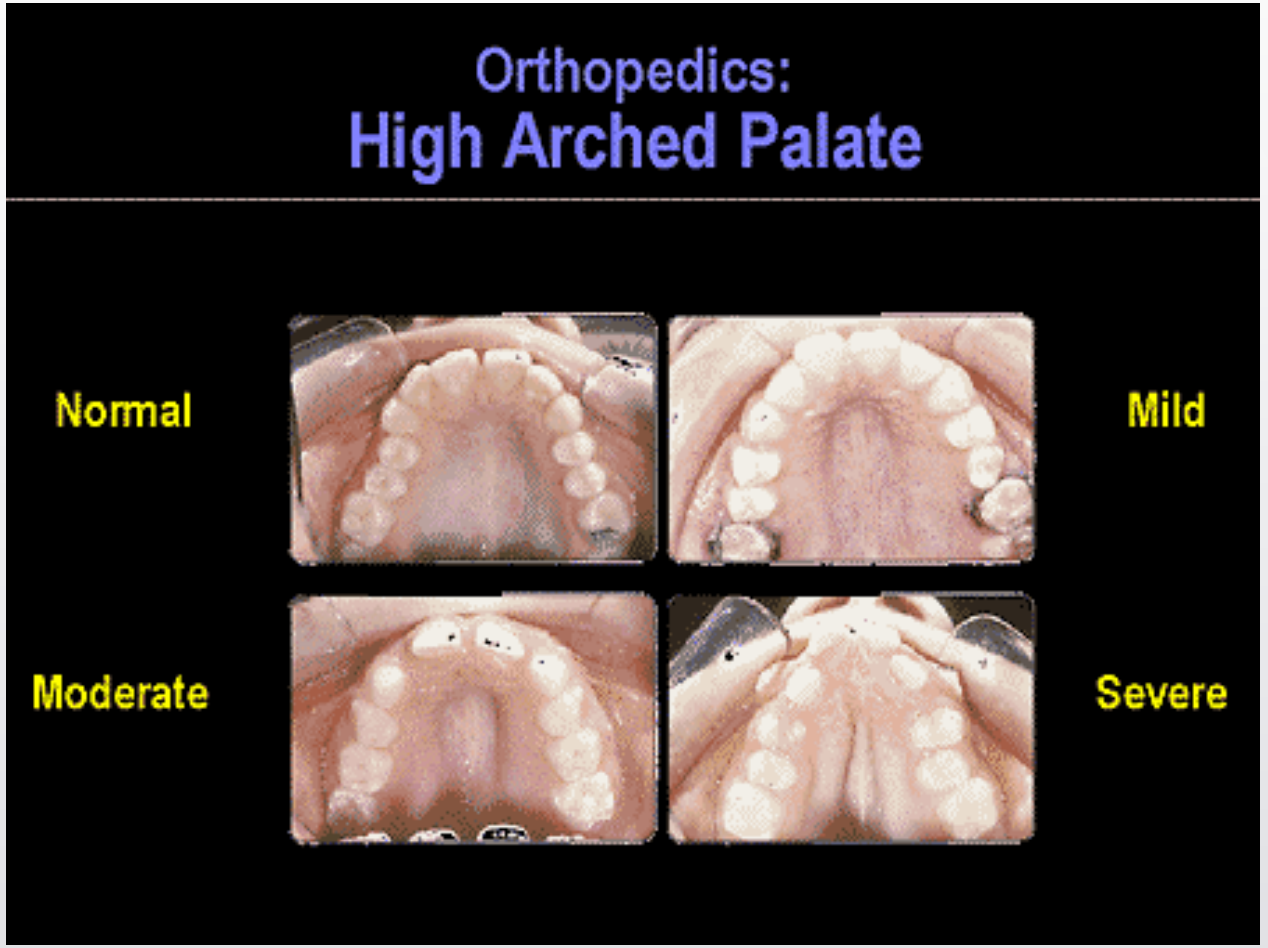


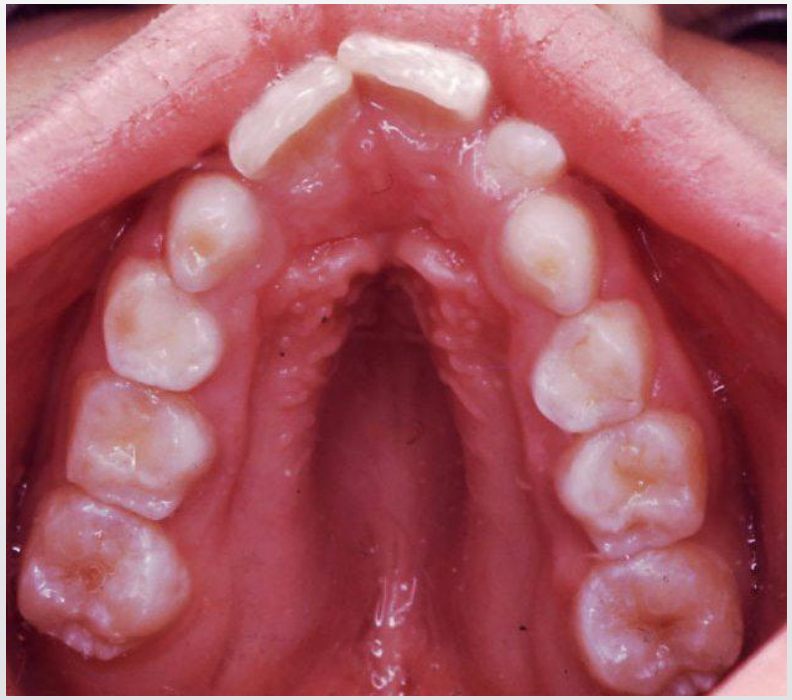
Hard Palate Anatomy



High Arched Palate

- Tooth Crowding due to a V shaped arch instead of a U shaped arch


Orthopedics:
High Arched Palate
Normal
Mild
Moderate
Severe



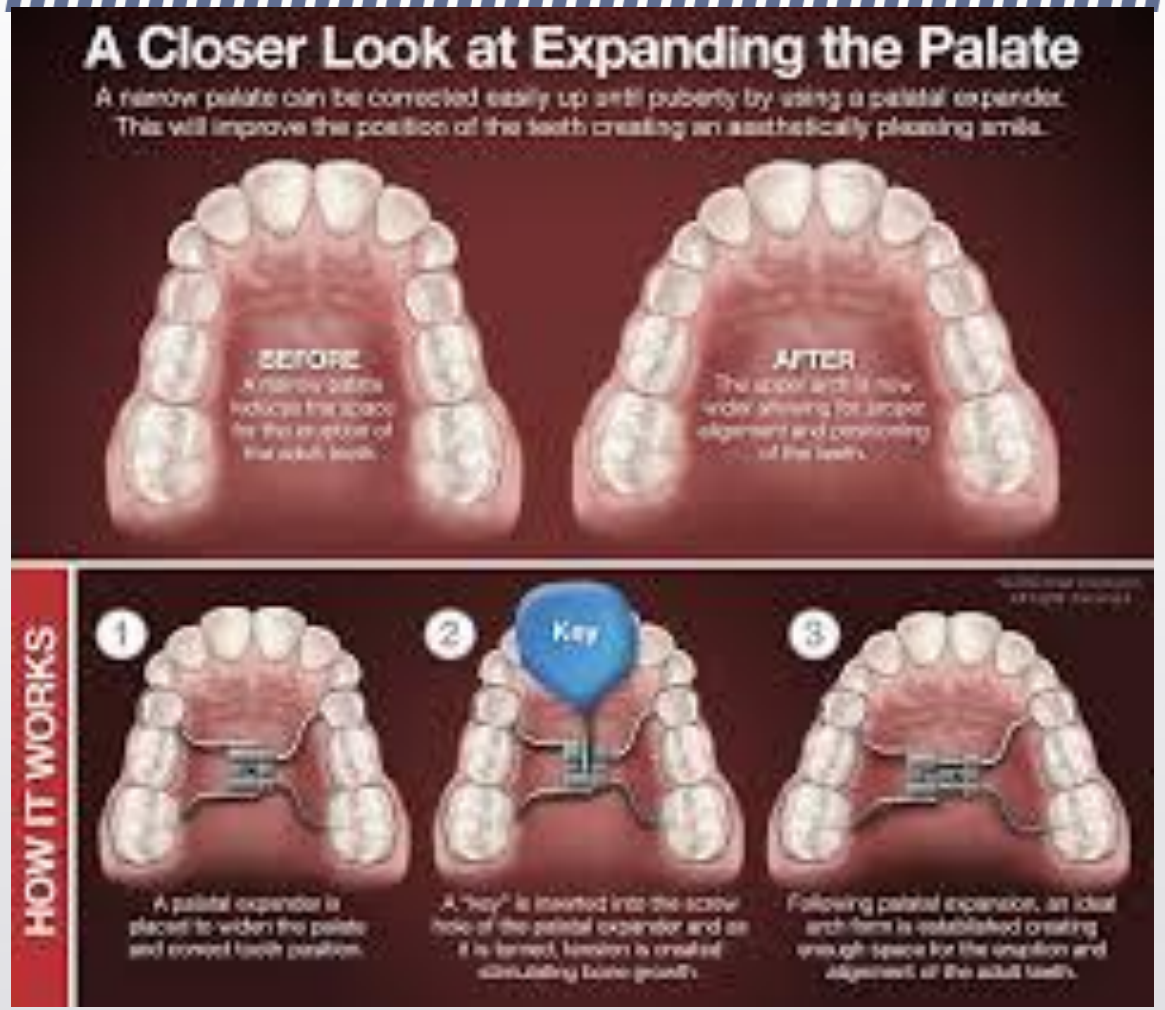
High Arched Palate

- Can lead to something called crossbite

Normal Bite (upper teeth overlap lower teeth)

Crossbite (upper teeth inside of lower teeth)


A
B
C
D
© orthoLemay.com


A Closer Look at Expanding the Palate
A narrow palate can be corrected easily up until puberty by using a palatal expander. This will improve the position of the teeth creating an aesthetically pleasing smile.
BEFORE
AFTER
HOW IT WORKS
1
2
Key
3

Palatal Expander


Amount of
expansion


Before
After
Before
After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Palate Expander Attached To Molars

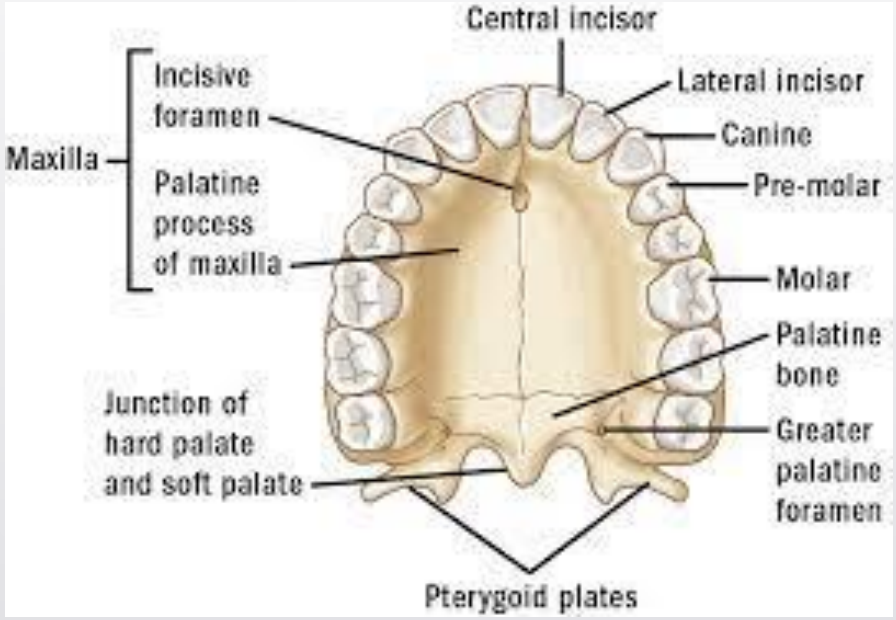
Before Placement

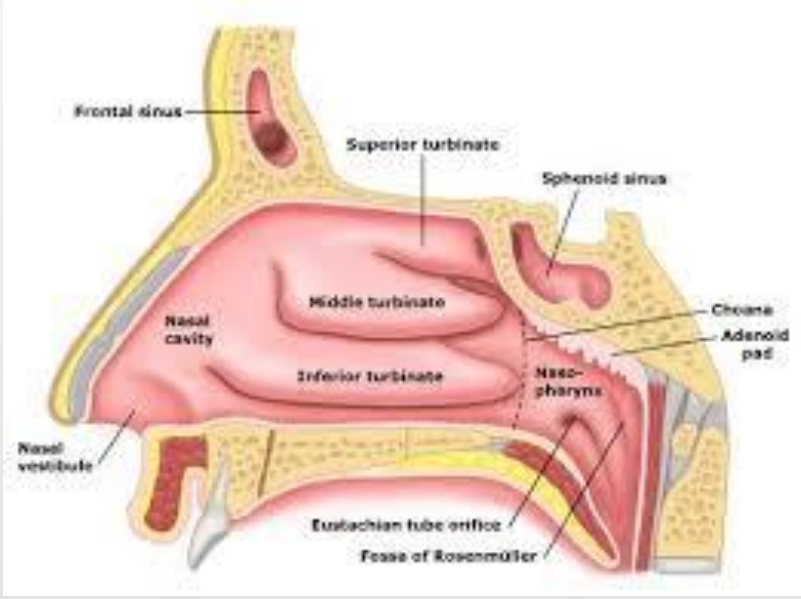
Narrow Palate

After Expansion

2-4 Weeks After Placement

Hard Palate Anatomy




Frontal sinus
Superior turbinate
Sphenoid sinus
Middle turbinate
Nasal cavity
Choana
Adenoid pad
Inferior turbinate
Naso-pharynx
Nasal vestibule
Eustachian tube orifice
Fossa of Rosenmüller

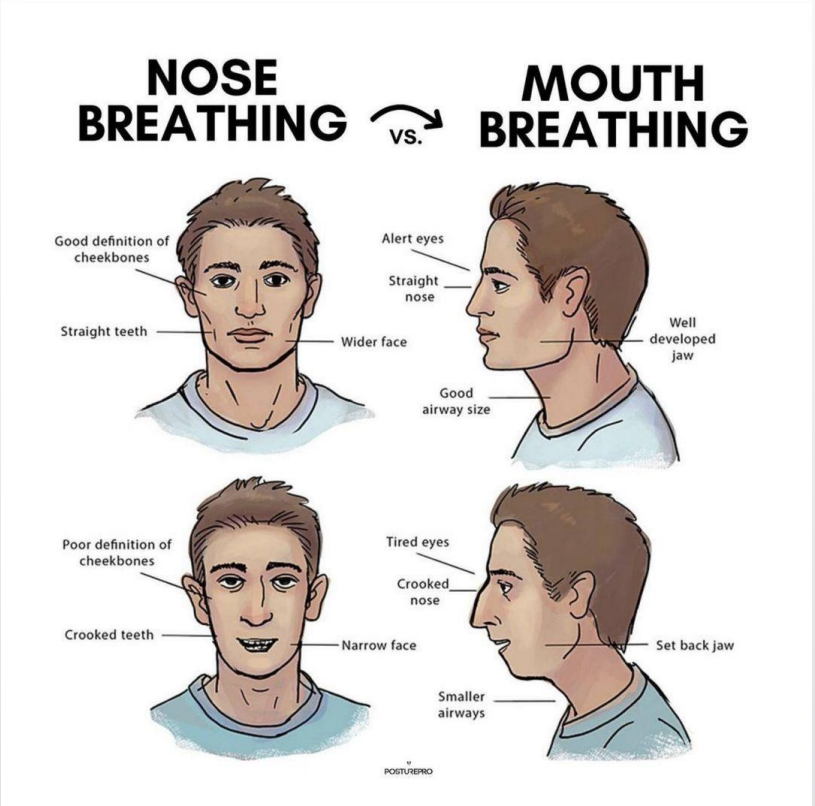
High Arched Palate

- Palate is roof of mouth, but also floor of the nose
- Decreases size of the nasal cavity
- Makes nasal breathing difficult
- Small nasal areas difficult to clear mucous can lead to congestion
- Increase in sleep disordered breathing

How Does Our Breathing Habit Change?

High Arched Palate

- Mouth breathers
- Humans did not evolve to breathe through the mouth
- Changes facial features (long face, down slanting eyes)
- Pressure from tongue on the roof of mouth contributes to midface development (try breathing through mouth with tongue on palate)
- Leads to inflammation of the gum tissue


NOSE BREATHING
vs.
MOUTH BREATHING
Good definition of cheekbones
Straight teeth
Wider face
Alert eyes
Straight nose
Well developed jaw
Good airway size
Poor definition of cheekbones
Crooked teeth
Narrow face
Tired eyes
Crooked nose
Set back jaw
Smaller airways
POSTUREPRO

Gingival Inflammation from Mouth Breathing

Enamel Demineralization from Mouth Breathing

Why Do We Have Crowded Teeth in the Lower Jaw?

Micrognathism (undersized jaw)

Micrognathia (small jaw) Symptoms in Children

- Because jaw is undersized, it leads to tooth crowding
- Infants tend to show signs of “failure to thrive”
- Apneic spells (temporary stop in breathing during sleep)
- Inability to feed
- Prolonged feeding
- Noisy breathing
- Poor ability to sleep
- Poor ability to gain weight

Micrognathia (small jaw)

- Most of these problems occur because the tongue is blocking the airway

Good News

Micrognathia (small jaw)in Noonan Syndrome

- Tends to correct itself to an extent as the child grows
- Most are treated without the need for surgery

Malocclusions

- Teeth do not align correctly
- Consequence of high palate and small lower jaw

Most Common Malocclusions in Noonan Syndrome

Overcrowding

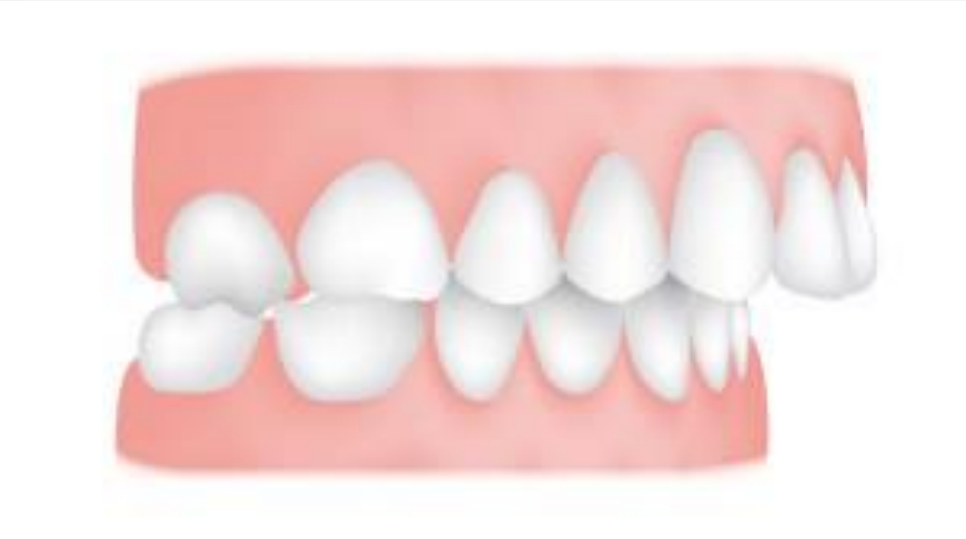
- Caused by lack of space
- Very common
- Results in teeth that are crooked and overlap

Overcrowding

Overjet

- Top teeth extend past your bottom teeth horizontally
- Due to small lower jaw

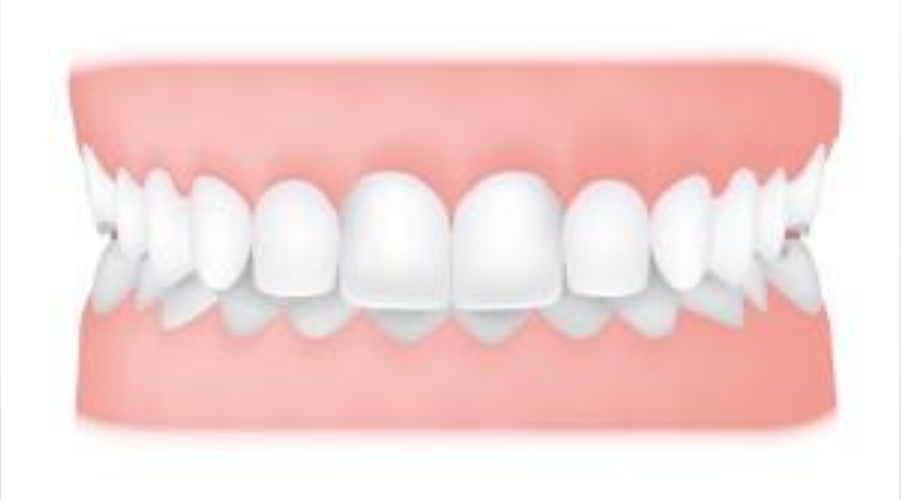
Overjet



Overbite

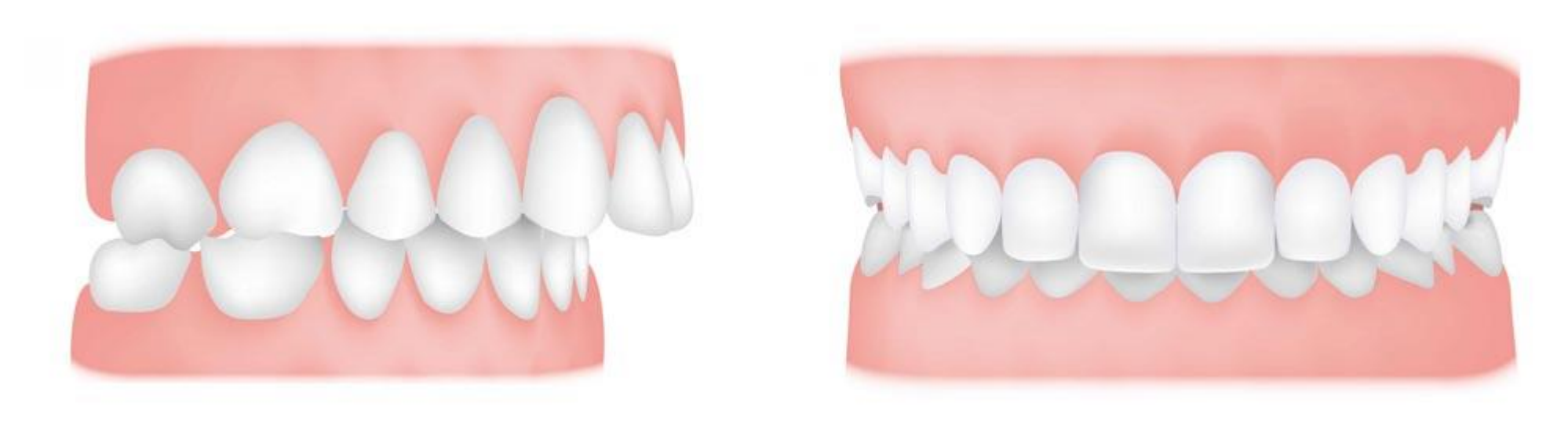
- Overlap of the front teeth over the bottom teeth
- Normal
- Excessive overbite can cause the front teeth to bite down on the lower gums

Excessive Overbite (Deep Bite)



Easy to Confuse Overjet vs Overbite

Overjet (protrusion) vs Overbite (curtain effect)



Crossbite

- Upper teeth bite inside of your lower teeth
- Consequence of a high palate

Normal Bite

Crossbite


Cross-bite

Open Bite

- Front teeth do not overlap the bottom teeth
- Frequently associated with tongue thrust habit
- Associated with mouth breathing (upper jaw doesn't develop normally)

Open Bite

Treatment of Malocclusions (teeth don't fit together as they should)

- Orthodontics
- Jaw surgery in a few select cases

Tooth Development Anomalies

Missing Tooth (Hypodontia)

- Certain teeth do not develop
- In Noonan Syndrome, seen with wisdom teeth, upper lateral incisors, lower premolars most commonly

Missing Tooth


a
b
c
d
e
f


A
B
C
D


A
B
C
D
E

Missing tooth (hypodontia) in Noonan Syndrome

- Not necessarily a bad thing
- Alleviates some of the crowding
- Many times, involves less work for the orthodontist
- Less time in braces for the patient

Other Dental Anomalies

- Supernumerary Teeth (extra teeth)
- Retained baby teeth
- Unerupted permanent teeth
- Abnormal root morphology

Supernumerary Teeth or Hyperdontia (Shark Teeth)

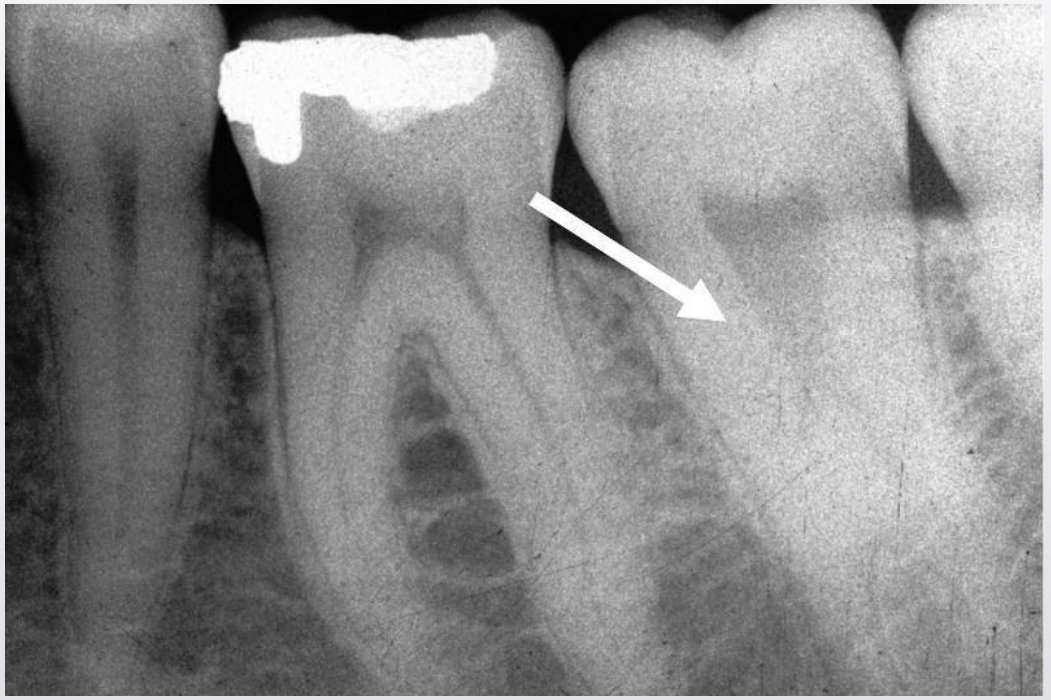
“Shark Teeth”
5 rows, 3000 teeth, lose 100 daily

Retained Primary Teeth

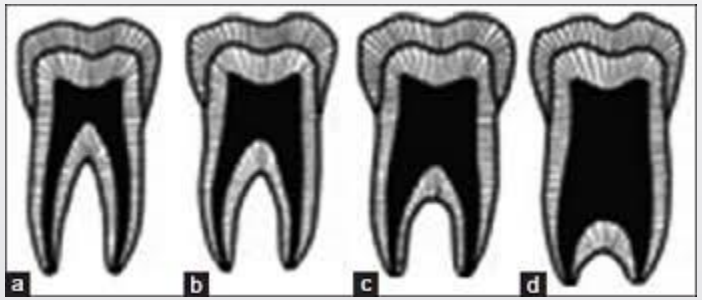
Unerupted Permanent Teeth (Impacted Teeth)

Unerupted Permanent Tooth

Abnormal Root Morphology (Taurodontism)



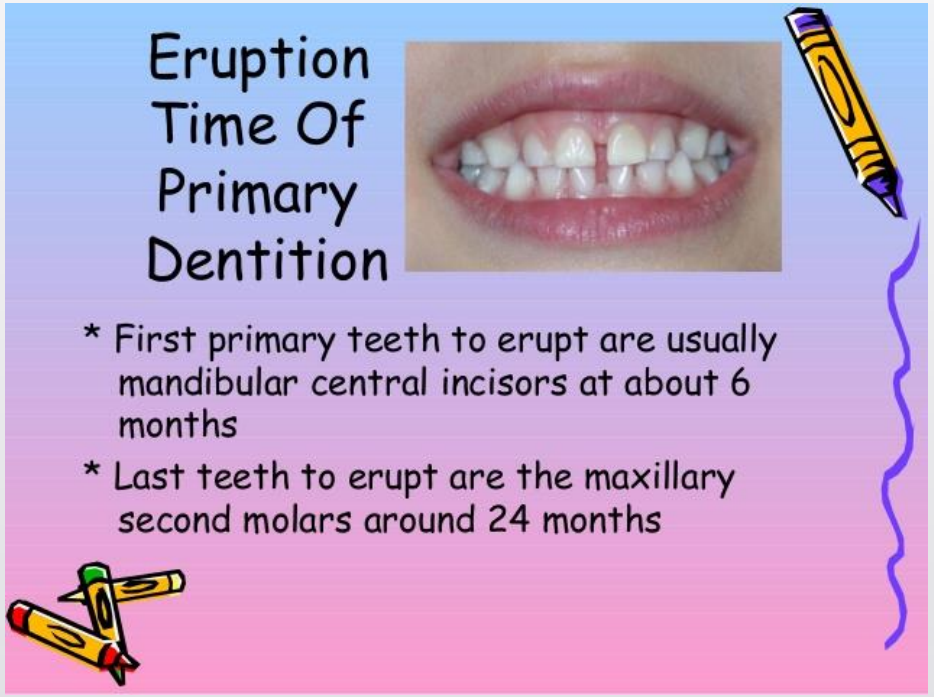
Taurodontism



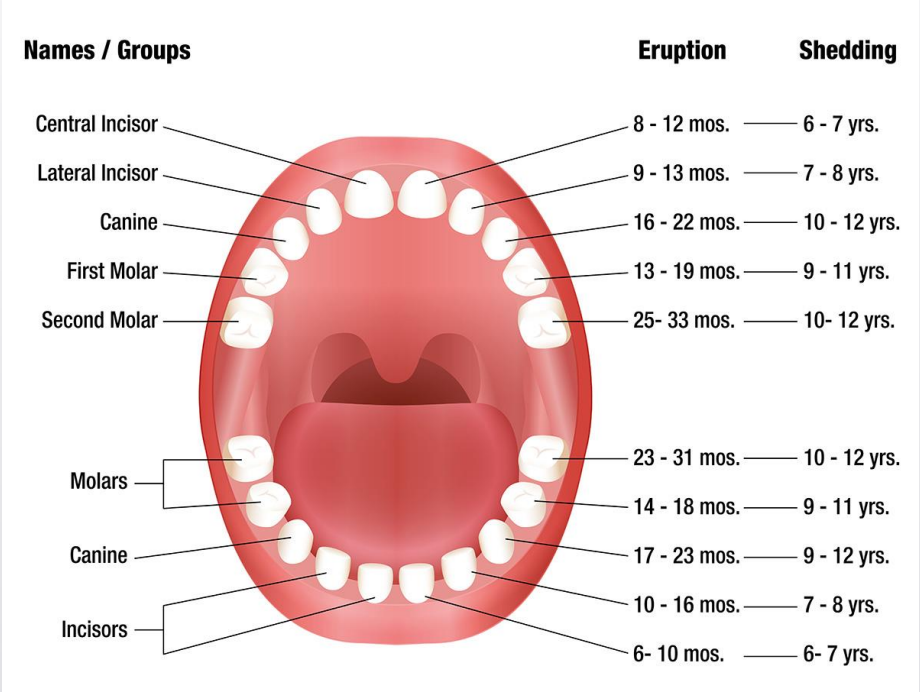
Double Tooth

Short Root Anomaly

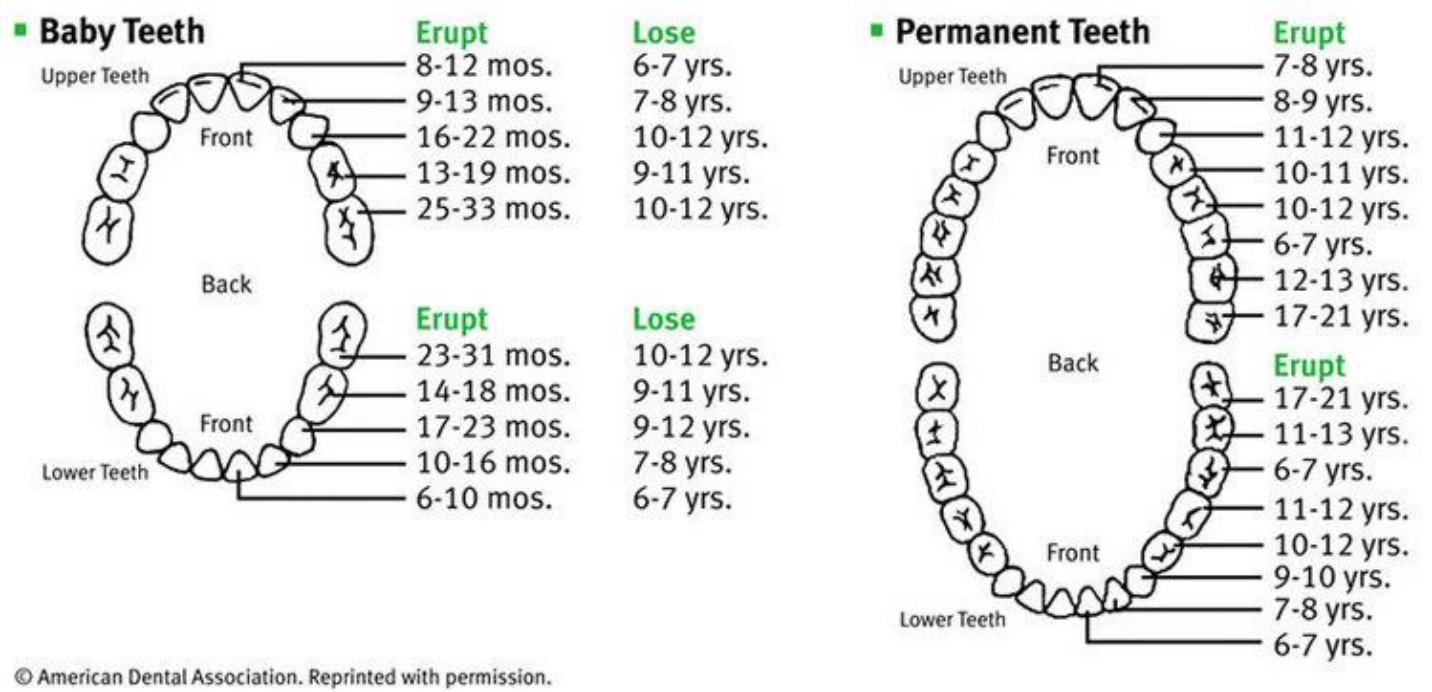
Eruption Times



Delays In Getting Primary Teeth/Losing Primary Teeth



Delays in Permanent Teeth Eruption

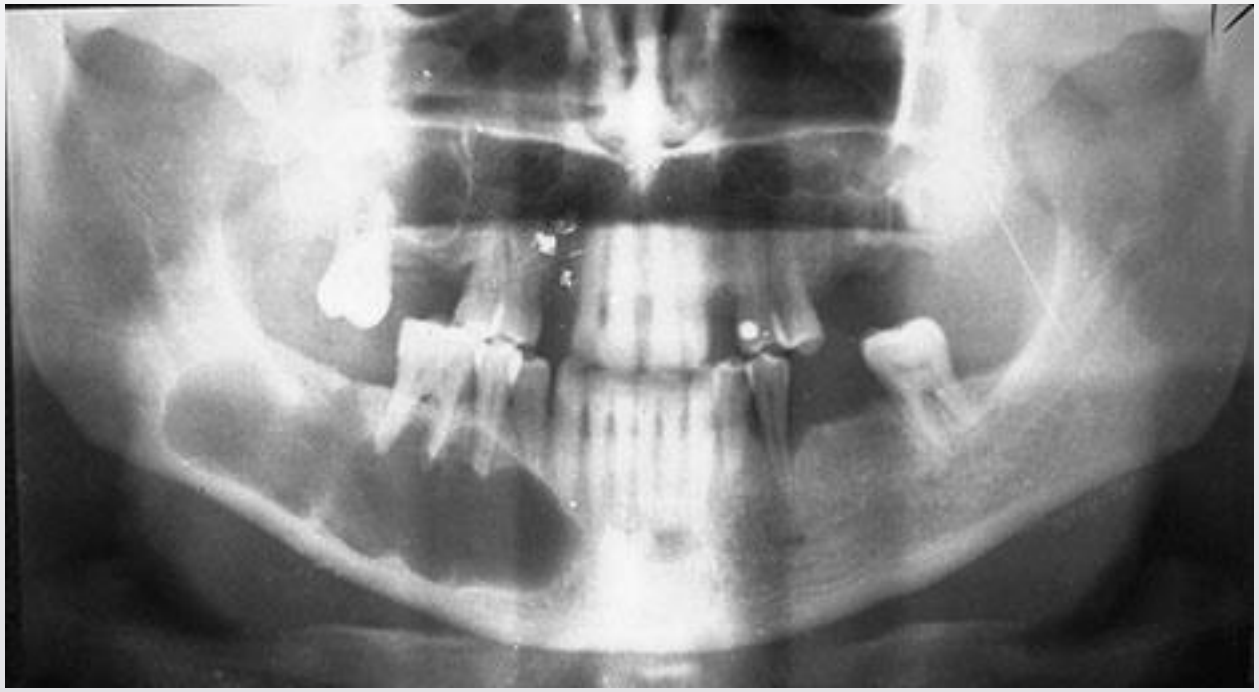


Tooth Fairy Time Is Delayed

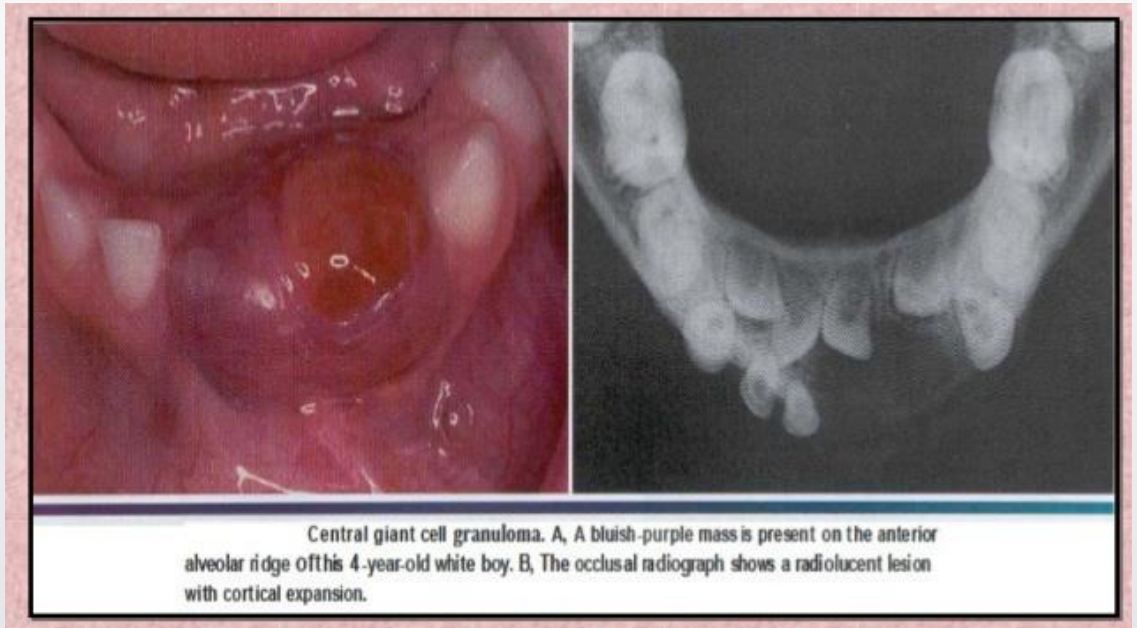
Central Giant Cell Granuloma

- Tumor of the jaw
- Not cancer
- It is potentially destructive

Central Giant Cell Granuloma (Dark Area)



Central Giant Cell Granuloma



Central giant cell granuloma. A, A bluish-purple mass is present on the anterior alveolar ridge of this 4-year-old white boy. B, The occlusal radiograph shows a radiolucent lesion with cortical expansion.

-Aggressive lesions may exhibit pain,rapid growth,cortical perforation and root resorptio

-Non aggressive exhibit no symptoms,slow growth,no perforation,no resorption

Central Giant Cell Granuloma

- Most are discovered on routine radiographic examination or as painless expansion of the affected bone

Central Giant Cell Granuloma

- Asymptomatic slow growing lesion to aggressive tumor causing pain and expansion of the jaw
- Anterior mandible/premolar area is most common area of occurrence
- Younger adults (30yo or under)
- Females>Males (2x female predilection)
- Aggressive type tends to recur
- Can displace teeth

Central Giant Cell Granuloma

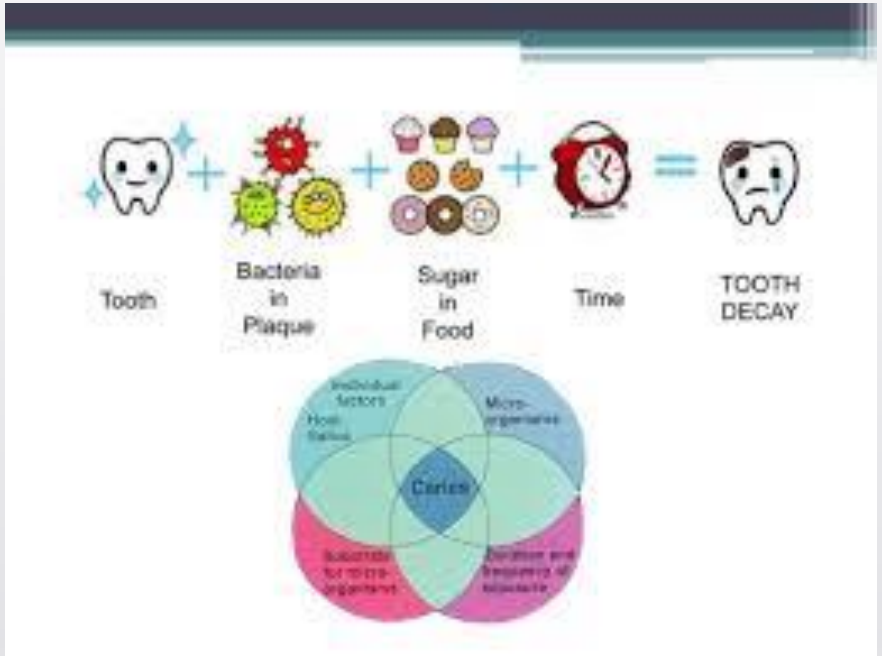
- 60% occur before age 30
- 70% arise in mandible

Central Giant Cell Granuloma

- Treatment is surgical curettage
- In patients with aggressive tumors, three alternatives to surgery
- 1) corticosteroids
- 2) calcitonin
- 3) interferon alfa-2a

Are those with Noonan Syndrome more prone to decay?

What exactly is a cavity?



Enamel Hypoplasia (poorly developed enamel)

Bleeding Disorders and Dental Treatment

- Are of variable clinical severity and stem from different defects in the coagulation and platelet systems in NS patients
- Patients with NS should have a thorough coagulation evaluation
- Dental complications related to coagulation are unlikely

Conclusion

- Oral health care for NS patients should commence in the first year of life, in order to prevent problems from becoming irreversible
- Proactive orthodontic treatment should be administered as early as possible to address tooth crowding/bite issues
- Regular dental visits are a must
- Just because it does not hurt is no excuse to skip your visit to the dentist

In Memory of Dr. Jackie Noonan
Oct 28, 1928 - July 23, 2020