What to do when **all** the teeth are impacted: the challenge of CCD

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What is Cleidocranial Dysplasia?

- Genetic disorder affecting development of bones and teeth
- Late physical development
- Short stature
- Incomplete or missing clavicles
- Wormian bones of the skull (multiple ossification centers)
- Frontal bossing and median furrow

What is Cleidocranial Dysplasia?

- Mutation of RUNX2 gene
- Autosomal dominant
- Many sporadic cases
- Males and females equally affected
- No mental retardation

Dental Disability in Cleidocranial Dysplasia

- Class III skeletal pattern
- Reduced lower face height
- Poor alveolar development
- Delayed dental development – 3 years late
- Normal eruption of deciduous teeth
- Non-eruption of permanent premolars and canines
- Normal eruption of permanent molars - key to tx success

Dental Disability in Cleidocranial Dysplasia

- Over-retained deciduous teeth
- Unerupted permanent teeth
- Multiple supernumerary teeth
- Lessened eruption potential
- Abnormal tooth form
- Enamel hypoplasia – pitted enamel

Aims of Treatment

- Extract over-retained deciduous teeth
- Extract supernumerary teeth
- Erupt the impacted normal permanent teeth
- Align the newly-erupted permanent teeth
- Correct the jaw relation
**Treatment Timing: Dental**

Teeth normally erupt when 2/3 of the root is developed

- Incisors: 1/2 molars erupt at dental age 7 years (approx)
- Canines/premolars erupt at dental age 8-9 years (approx)

Dental age in CCD patients is 3-4 years later than normal

- Incisors 1st molars reach 2/3 root development at chronologic age 10-11 yrs (approx)
- Canines/premolars reach 2/3 root development at chronologic age 13-14 yrs (approx)

**Working assumption:** The teeth will not erupt without biomechanical assistance

**Principle #1:** Do not extract deciduous/supernumerary teeth without applying extrusive forces to unerupted permanent teeth

**Principle #2:** Surgically expose permanent teeth when they have 1/2 - 2/3 of their expected root length

**Principle #3:** Ensure solid anchor base & control of arch form (soldered lingual arches)

**Principle #4:** Erupt upper teeth versus lower teeth with intermaxillary, up-and-down, elastic forces – reciprocal anchorage

**Phase 1 treatment (chronologic age 12 yrs - dental age 9 yrs)**

**Aim of Phase 1:** erupt & align incisors only

This means:

1. Extract anterior deciduous teeth
2. Extract supernumerary teeth
3. Expose 1st and permanent incisors
4. Apply traction to erupt incisors
5. Align incisors

**Chronologic age 12 years, Dental age 8-9 years**

*In general, teeth will not erupt following extraction of deciduous and supernumerary teeth!*

Neither does wholesale open exposure guarantee spontaneous eruption

Furthermore, we need anchor molars as a stable base from which to deliver directed forces – and these are expected to erupt autonomously at age 10-11

So, we must wait!!

______ but, is there anything useful we can do in the meantime?

Pre-phase 1 – maxillary protraction for the very young skeletal class 3 patient

**In a normal healthy child**

- Eruption occurs when the tooth has 2/3 developed root, i.e. incisors normally erupt at age 6-8 yrs
- A premolar or canine reaches this stage of development at age 10-11 yrs
- Skeletal growth largely completed 18-22 yrs

Therefore, a normal child with impacted incisors, canines and premolars requiring orthognathic surgery for a skeletal class III needs 3 phases of treatment

**In a CCD child**

- Incisors erupt at age 6-9, 9-11 yrs
- Premolars and canines erupt at age 10-11, 13-14 yrs
- Growth largely completed 18-22, 21-25 yrs

The Jerusalem Approach to treatment for CCD

- Phase 1 orthodontics (in orthodontic office):
  - Set up an orthodontic anchor base from which to apply extrusive forces
- Phase 1 surgery (in operating theater):
  - Extract deciduous incisors, canines and all accessible supernumeraries, expose permanent incisors for bonding
- Phase 1 orthodontics (in operating theater):
  - Bond attachments, tie-in traction device and activate immediately
Phase 2 treatment (age 15 yrs, dental age 12 yrs)

Phase 2 orthodontics:
Establish a new anchor base from which to apply extrusive forces

Phase 2 surgery (in operating theater):
Extract deciduous molars and all remaining supernumerary teeth, expose permanent teeth for bonding

Phase 2 orthodontics (in operating theater):
Bond attachments, tie-in traction device and activate immediately

Phase 3 treatment (age 21 yrs) Orthognathic Surgery

- correct skeletal a-p relations
- correct skeletal vertical relations
- correct skeletal coronal relations
- provide optimized bony support for soft tissue drape

The post-surgical outcome of all 3 phases aims to:

- increase height lower third of face
- normalize facial profile
- improve form of the chin
- improve nasal profile

The comprehensive orthodontic & surgical planning, timing & interdisciplinary cooperation is able to achieve improved health, function & appearance in Cleidocranial Dysplasia patients