

Cleidocranial Dysplasia: Surgical and Orthodontic Management of Multiple Impactions in the mandible

Abstract

A 24-year-old female presented with the distinctive dentofacial features of cleidocranial dysplasia (CCD) including multiple supernumerary and permanent impacted teeth. Cone beam computed tomography (CBCT) was essential to accurately identify and plan the surgical recovery of the compromised permanent teeth. All obstacles in the paths of eruption were surgically removed, and OrthoBoneScrews (OBSs) with 3D lever arms provided the traction mechanics. Patients with CCD are complex problems, requiring carefully coordinated interdisciplinary care. (Int J Orthod Implantol 2016;42:84-96)

Key words:

supernumerary teeth, impaction, CBCT, OrthoBoneScrews, 3D lever arm, cleidocranial dysplasia

Introduction

Frontal bossing of the forehead, antimongoloid palpebral fissures, and a large number of impacted permanent and supernumerary teeth are consistent with the developmental anomaly cleidocranial dysplasia (CCD)(Fig. 1).¹⁻⁴ The preferred treatment for achieving an optimal dentition is to uncover and bond an attachment on each impacted permanent tooth, clear the path of eruption, extrude the impaction(s) with



■ Fig. 1:

A pre-treatment facial photograph and panoramic radiograph reveal the common dentofacial anomalies of cleidocranial dysplasia (CCD).

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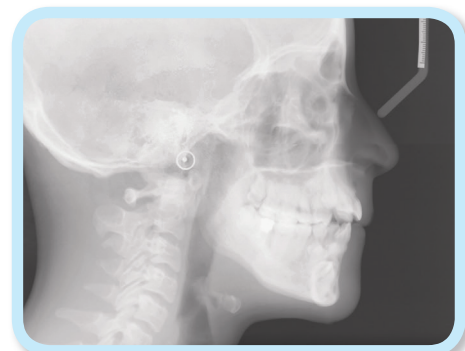
closed eruption technique, and optimally align each arch.⁵ Specific surgical and orthodontic approaches are customized to achieve efficient treatment.⁵⁻⁹ This case report is a step-by-step guide to the surgical and orthodontic procedures for managing patients with multiple impactions of permanent and supernumerary teeth. A checklist is provided to assist clinicians in the complex procedures.

Case Study

A 24y/o female presented for orthodontic consultation with a chief complaint of a malocclusion with many impacted teeth. Facial evaluation was a straight profile, frontal bossing of the forehead, and facial asymmetry due to maxillary and mandibular deviation to the right (*Fig. 1*). Intra-oral examination revealed an anterior open bite, 5mm overjet associated with severe crowding in the anterior maxilla (*Fig. 2*), numerous retained deciduous teeth (*Fig. 4*), and multiple impacted permanent, as well as supernumerary teeth (*Fig. 1*). Despite multiple dental eruption problems, the sagittal relationship of the malocclusion was Class I (*Fig. 5*). Panoramic radiography (*Fig. 1*) and cone beam computed tomography (CBCT) images (*Fig. 6*) were used to classify eight retained deciduous teeth, twelve impacted permanent teeth, and five supernumerary teeth in the symphysis area, and four supernumerary teeth in the maxilla. (*Figs. 1, 6 and 7*). The 3D CBCT images were used to define the precise morphology and location of the impacted teeth prior to their extraction or surgical uncovering and bonding in preparation for orthodontic traction.⁵ These specialized procedures are presented in the subsequent checklist.



■ **Fig. 2:**
An intraoral photograph shows anterior crowding and an open bite.



■ **Fig. 3:**
A lateral cephalometric radiograph is consistent with the facial form of CCD.

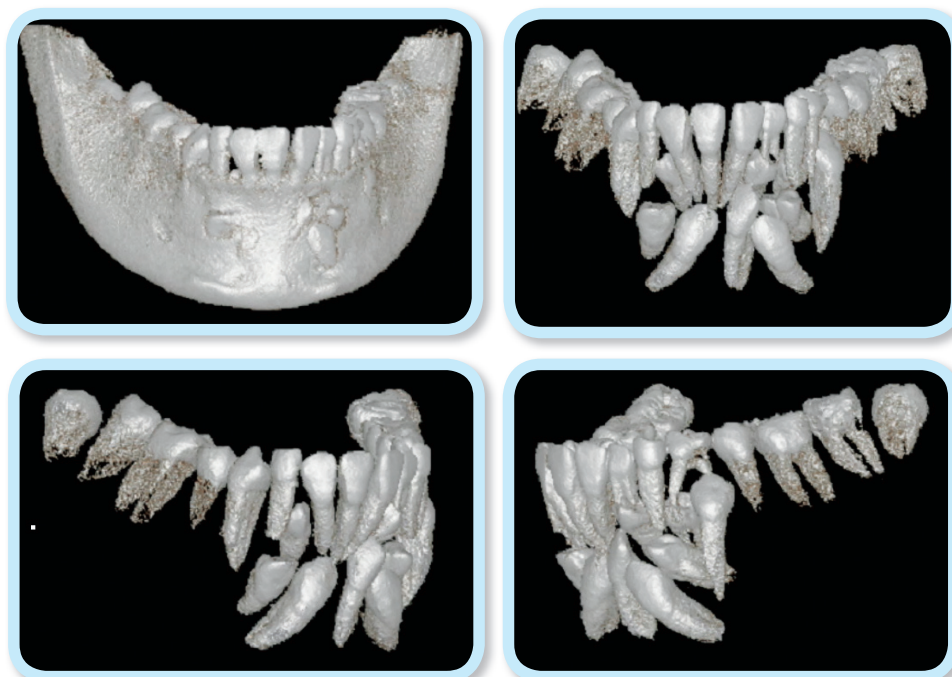


■ Fig. 4:

Intraoral occlusal photographs reveal a highly arched palate with severe crowding in the upper arch (left) and multiple retained deciduous teeth in the lower arch (right).

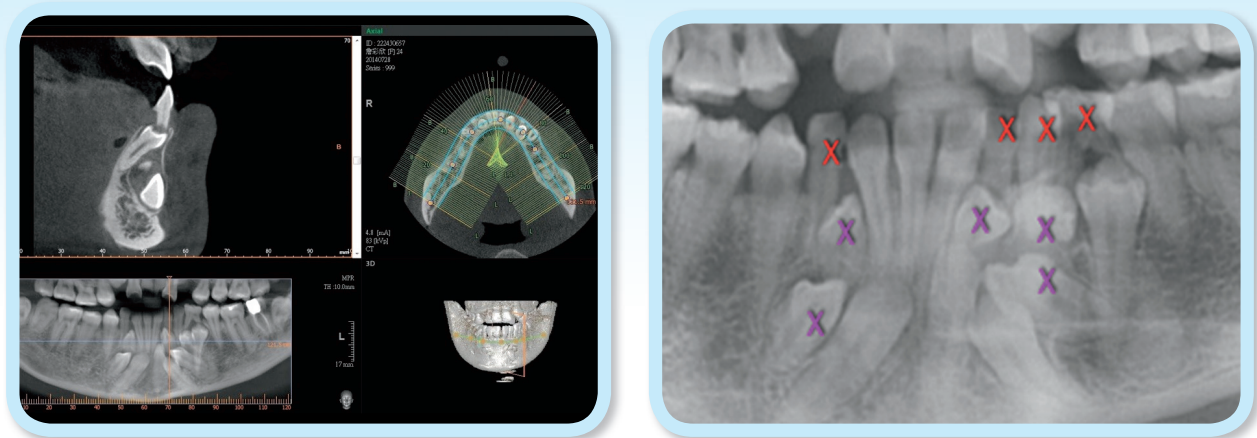


■ Fig. 5: Buccal-view intraoral photographs document a Class I malocclusion on both sides.



■ Fig. 6:

Cone beam computed tomography (CBCT) images of the lower arch reveal a complex array of impacted supernumerary and permanent teeth.



■ Fig. 7:

CBCT views (left) and a panoramic image (right) were utilized to identify and define the 3D positions of the impacted supernumerary and permanent teeth. The red Xs denote deciduous teeth that were extracted, and the purple Xs are supernumerary teeth that were removed to allow the impacted permanent teeth to erupt.

Surgery Procedures

Right side 1st surgery

1. Orientation: Begin with the end in mind.

The CBCT image is used as a digital map to design and sequence the clinical steps. Two dimensional (2D) plane radiographs (*panoramic and periapical films*) are inadequate for complex problems because they fail to reveal depth. CBCT provides the precise location of teeth in 3D.⁵ CBCT slices assess the thickness and density of the covering bone. In addition, 3D imaging is critical for differential diagnosis because it shows root morphology and other anomalies that may affect treatment decisions. It is a waste of time and money to try to recover an unserviceable tooth.⁶

2. Flap Reflection

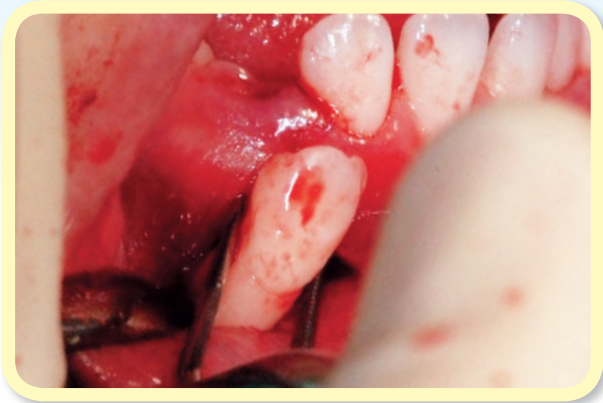
Apply local anesthesia to the surgical site, and then use the vertical incision subperiosteal tunnel access (VISTA)⁷ technique to access the impactions.

The vertical incision is made with a No. 15c surgical scalpel (Fig. 8), and the mucogingival flap is reflected with a surgical curette and periosteal elevator. The deciduous canine and adjacent supernumerary teeth are removed (Fig. 9). Use an explorer to penetrate overlying bone to locate the position of the permanent cuspid crown. There is a distinct, sharp recoil when an explorer engages enamel compared to bone. This is a simple and effective way to identify the crown of an impacted tooth.^{2,3}

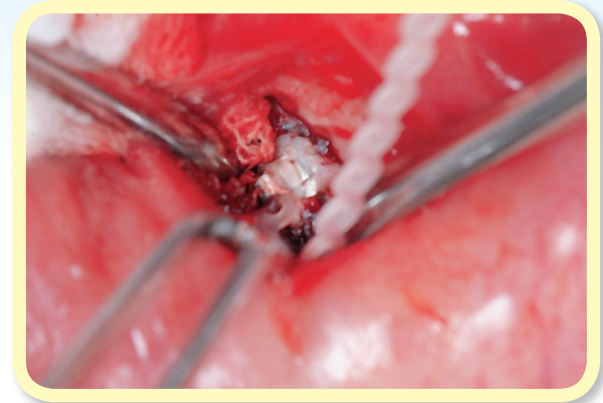


■ Fig. 8:

An intraoral photograph of the lower labial area shows a vertical incision for the minimally invasive VISTA surgical procedure.



■ **Fig. 9:**
The deciduous canine and adjacent supernumerary teeth were removed as atraumatically as possible.



■ **Fig. 10:**
To avoid excessive bleeding during the bonding procedure, an eyelet is bonded on the enamel of the impacted canine before removing the bone to clear the path of eruption.

3. Etching & Bonding

Moisture control is essential for successful bonding of attachments, but it is also the most difficult part of the procedure. Bleeding can be controlled with electrocautery or gauze pressure. Water, saliva and residual blood are removed with high power suction, and then a bondable eyelet is bonded on the crown portion of the impacted canine (Fig. 10). A well trained, experienced assistant is very important for reducing the bonding time, which increases the probability of success. The longer the duration of the bonding procedure, the more likely it will fail.

4. Bone Removal

A high speed handpiece with a #5 carbide round bur removes the overlying bone down to the cementoenamel junction (CEJ). Removing the

bone apical to the height of the crown curvature can facilitate tooth eruption. It is best to remove bone near the CEJ with a hand instrument to avoid injuring the tooth. Cervical damage to the tooth can subsequently result in external root resorption.⁸

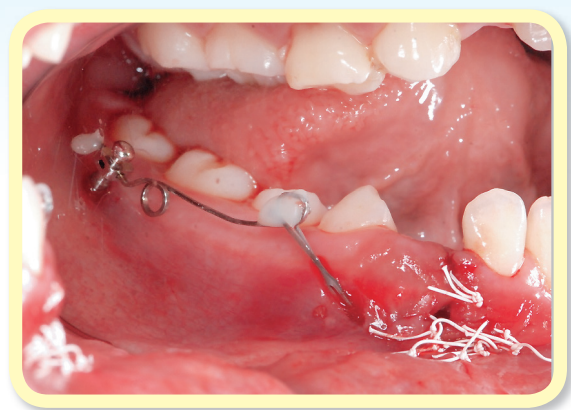
5. Anchorage Screw and 3D lever arm

Make an indentation with an explorer in the right mandibular buccal shelf and then insert a 2x14mm OrthoBoneScrew® (OBS) with a rectangular hole through the head of the screw (Newton's A Ltd, Hsinchu, Taiwan) (Fig. 11). Make a series of vertical incisions at the corner of dental arch, apical to the area of the lower right cuspid and lateral incisor. Connect a power chain to the impacted canine, thread it beneath the soft tissue (Fig. 12), and attach the proximal end to the distal portion of the 3D lever arm. The latter is a spring made of



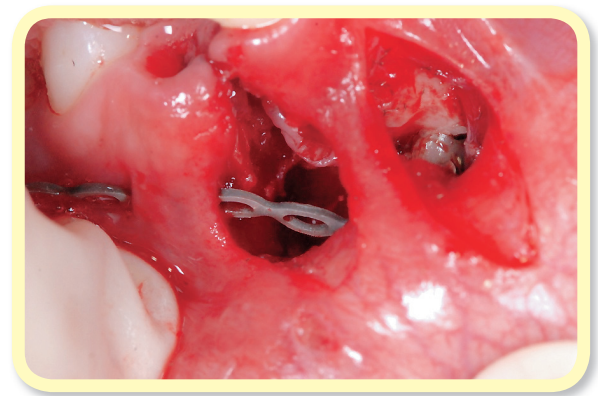
■ Fig. 11:

Marks (indentations) in the distal portion of the screw driver engaging the OBS are consistent with the screw holes for 3D lever arm insertion. This feature allows precise positioning of the OBS relative to the desired orientation of the 3D lever arm.



■ Fig. 13:

Traction is applied to the impaction by a power chain bonded to the distal end of the 3D lever arm.



■ Fig. 12:

A power chain coursing beneath the alveolar mucosa (VISTA procedure) delivers the traction force from the 3D lever arm to the eyelet bonded on the impaction. See text for details.

.019x.025" SS that is inserted into the rectangular hole in the head of the OBS (Fig. 13). Polymerize flowable resin on both ends of the 3D spring to achieve retention in the OBS and to stabilize the power chain attachment to the lever arm (Fig. 13), which is activated to upright the impacted canine. The force system is designed to provide

direct traction to upright the impaction without producing deleterious side effects for the rest of the dentition. The flap is repositioned and closed with interrupted 5-0 GORE-TEX® sutures (Flagstaff, AZ). A post-operative panoramic radiograph confirms the desired position of the mechanics and angulation of the traction force (Fig. 14). In recovering the impaction, beware of impinging



■ Fig. 14:

A post-operative panoramic radiograph was exposed to check the position of the traction mechanism.

on the roots of adjacent teeth or in creating occlusal interferences.⁴ It may be necessary to reposition the OBS as the impaction is extruded. The following is the specific series of surgical and traction procedures for the current patient:

Left side 1st surgery - 2 weeks later

1. Flap Reflection

Instead of the VISTA approach, an open flap surgical procedure was selected for the left side.⁹ Since the surgical field was much wider on the left side of the arch, the minimally invasive VISTA surgical approach was impractical (Fig. 15).



■ **Fig. 15:**
An intrasulcular flap is reflected to expose the impacted canine.

2. Bone and Supernumerary Teeth Removal

The treatment objective was to retain all of the deciduous teeth for as long as possible to provide

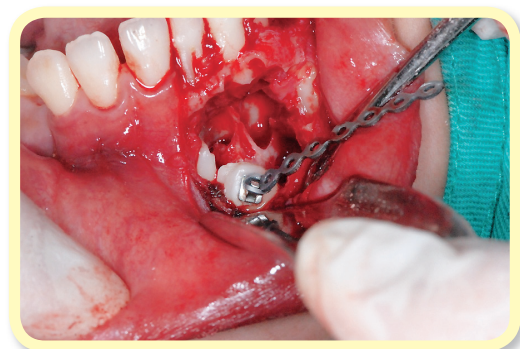
space maintenance and optimal masticatory capability. Considerable surgical care was required in removing the supernumerary teeth (Fig. 16).



■ **Fig. 16:**
Careful surgical technique is required for the relatively atraumatic extraction of the supernumerary teeth.

3. Etching & Bonding

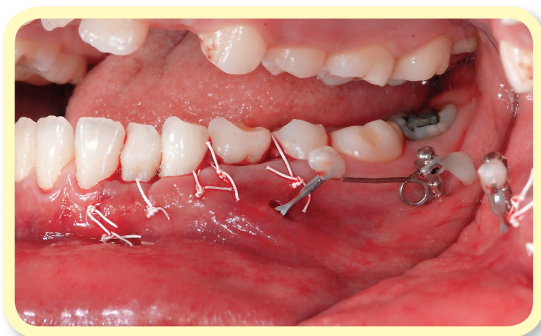
A eyelet was bonded only on the canine because the lateral incisor is expected to erupt spontaneously when all the obstacles in its path were removed. It is important to only apply traction when needed; otherwise allow natural forces to prevail (Fig. 17).



■ **Fig. 17:**
A power chain is attached to the eyelet on the impaction prior to sealing the connection with the polymerization of flowable resin.

4. Screws Insertion and 3D Lever Arm

Make a small incision along the intended line of force to allow the power chain to pass through the soft tissue. The flap is then repositioned and closed with interrupted 5-0 GORE-TEX® sutures (Fig. 18). A panoramic radiograph is taken every month post-operatively, to check the progress in extruding the impactions, and to determine when a change in the direction of traction is required.



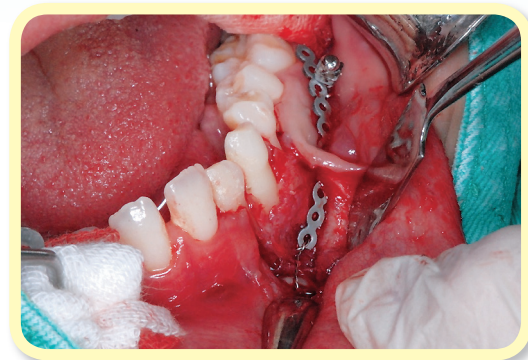
■ Fig. 18:

The 3D lever arm is activated by attaching a power chain (upper view). The activated position is shown in the lower view.

Left side 2nd surgery - 6 weeks later

The OBS was repositioned and the 3D lever arm

was removed to achieve a more direct direction of traction on the impaction (Fig. 19).



■ Fig. 19:

The position of the OBS is positioned according to the desired direction of traction.

Right side 2nd surgery - 1 month later

A steel ligature from the impaction was extended through the soft tissue incision, and was then attached to the lever arm to extrude the impaction (Fig. 20).



■ Fig. 20:

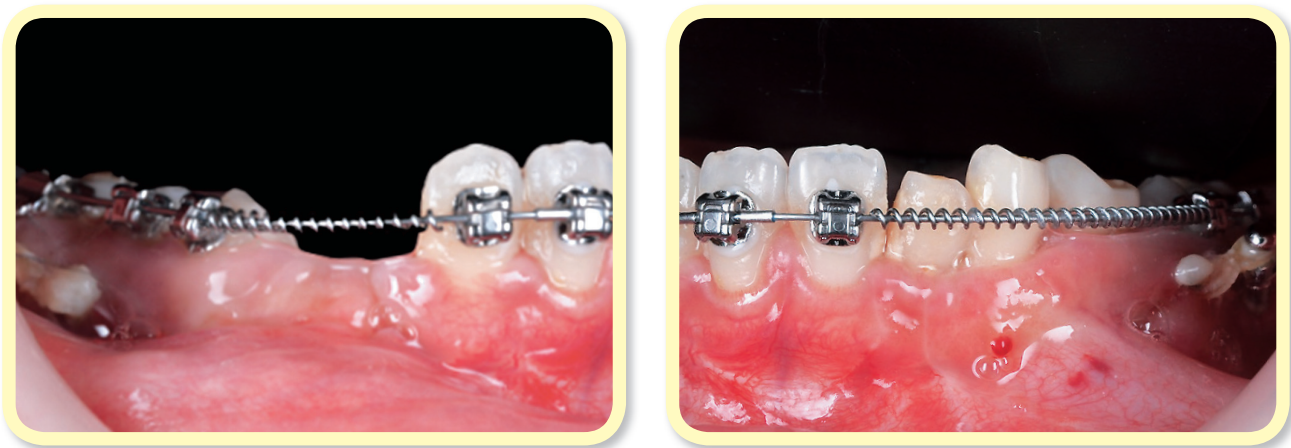
The activated section of power chain is superficial to the flap.

Bonding lower bracket - 3 months later

When the panoramic radiograph showed that the lower left lateral incisor and the canines were in the desired positions, the right side lever arm was removed, brackets were bonded on the lower teeth, and an .014" CuNiTi archwire was placed.

Create space for eruption - 6 weeks later

The archwire size was increased to .018" CuNiTi and an open coil spring was activated to create spaces for the erupting teeth (*Fig. 21*).



■ *Fig. 21:* NiTi open coil springs are used to create enough space to align the impactions when they erupt.

Change traction direction - 1 month later

The deciduous teeth in the path of eruption were extracted and the archwire was increased to .014x.025" CuNiTi. Elastic ligature was connected directly to the archwire to complete the extrusion of the impactions (*Fig. 22*).



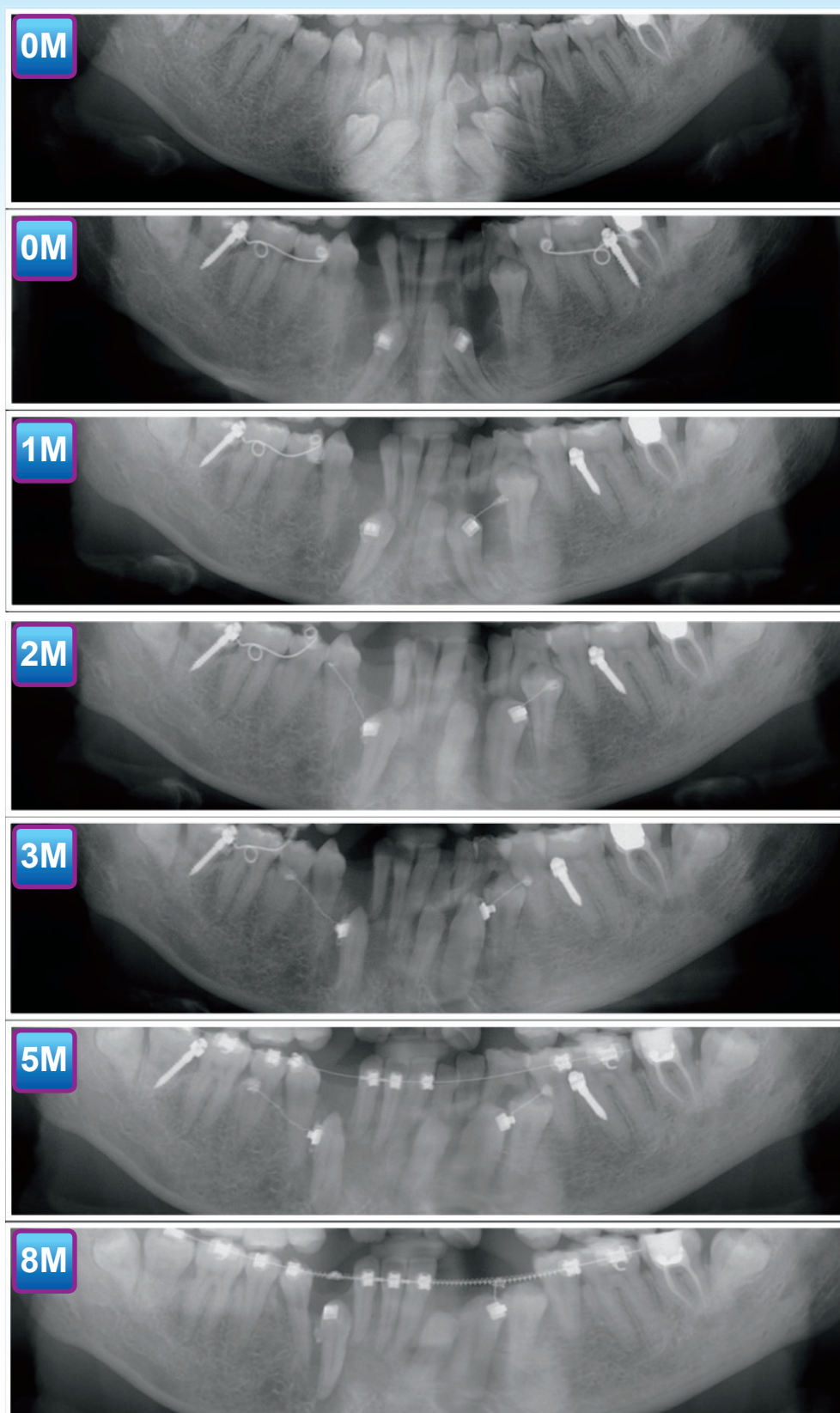
■ Fig. 22:

After the remaining deciduous teeth are extracted, both lower impacted canines are bonded with attachments and traction is applied bilaterally.

Discussion

Marie-Sainton syndrome, also known as cleidocranial dysplasia (CCD), is an autosomal dominant skeletal dysplasia affecting primarily the development of the bones and teeth.¹⁻³ Expression of CCD can vary widely, even within the same family, and its prevalence is approximately one in a million live births, worldwide.

The etiology of the disorder is a mutation of the RUNX2 gene, that been mapped to chromosome 6p21 within the same region as CBFA1. The latter gene controls the differentiation of precursor cells into osteoblasts and is essential for both membranous and endochondral bone formation.¹



■ **Fig. 23:**
A sequential series of panoramic radiographs (0-8M) shows the progress for the bilateral recovery of the impacted lower canines.

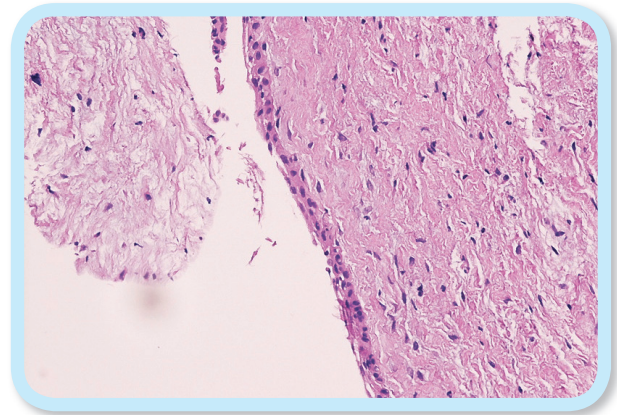
Distinctive facial features of CCD include:

- Wide, short skull (*brachycephaly*)
- Prominent forehead.
- Hypertelorism, i.e. excessive space between the eyes
- Flat nose
- Underdeveloped maxillary arch, i.e. small upper jaw

CCD may be associated with decreased bone density (*osteopenia*) that progresses to osteoporosis and low trauma bone fractures at a relatively early age. In addition, hearing may be impaired and infections are relatively common in the inner ear and facial sinuses. Affected children may be mildly delayed in the development of motor skills, but intelligence is usually normal.

Characteristic dentoalveolar anomalies are frequently associated with CCD.¹⁰ The palate is often highly arched, and clefts of the hard and soft palates are relatively common. Retention of the deciduous dentition in conjunction with impacted permanent teeth is a relatively common finding. If the primary teeth do exfoliate normally, the patients may be partially edentulous for a long period of time before the permanent teeth erupt, if they ever do. Permanent teeth usually show a delay of root development but still have the potential to erupt; however, the paths of eruption may be blocked by multiple supernumerary teeth. Surgical procedures to promote the eruption of the permanent teeth include extraction of deciduous teeth as well as removal of bone and supernumerary teeth

blocking the paths of eruption. The large number of supernumerary teeth (~30) accompanied by a dentigerous cyst (Fig. 24) is a severe expression of CCD. The radiographic picture for supernumerary teeth is usually similar to a normal tooth, but there may be a lack of cellular cementum on the root,¹¹ and abnormal bone morphology.¹² For CCD patients, coordinated orthodontic and surgical procedures are effective for achieving acceptable dental form and function.



■ Fig. 24:

A tissue biopsy from the area distal to the impacted mandibular (Fig. 23:1) shows fragments of the stratified squamous epithelium that lined a dentigerous cyst.

Conclusion

Keys to successful impaction recovery are:

- CBCT: Precise tooth identification and position are essential for surgical planning.
- Proficient Surgical Team: Decreases surgical time and increases reliability for bonding attachments.
- Clear the Eruption Pathway: Remove all the obstacles to eruption including deciduous teeth, supernumerary teeth, and bone.

- Bone near the CEJ: Carefully remove all osseous tissue impeding eruption with hand instruments.
- Well Designed Mechanics: An OBS with a 3D lever arm is recommended

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