Dental Management of an Arthrogryposis Multiplex Congenita Patient with Odontoma

AlSaeed M[1], AlMohareb R[1], AlKahtani A[1]

[1]Department of Pediatric Dentistry, Prince Sultan Military Medical City, Riyadh, Saudi Arabia

Introduction

Arthrogryposis Multiplex Congenita (AMC) is a rare congenital condition characterized by multiple non-progressive joint contractures and musculoskeletal anomalies. Though limb involvement is most common, craniofacial and orofacial manifestations—such as micrognathia, dolichofacial growth, and restricted mouth opening—can significantly impact oral function and access to care.

Odontomas are benign odontogenic tumors composed of disorganized dental tissues. Compound odontomas may prevent eruption or displace adjacent teeth. While both AMC and odontomas are associated with delayed dental eruption, their coexistence has not been previously reported. This case presents a rare overlap and emphasizes the importance of multidisciplinary management in syndromic pediatric dental care.

Case Description

A 10-year-old girl with a confirmed diagnosis of AMC was referred for dental management. Her medical history included bilateral developmental dysplasia of the hips and patellar dislocation, both surgically treated. She has a strong family history of AMC, with a younger, cousins, and aunts exhibiting similar musculoskeletal features.

She exhibits classic AMC features including short stature, camptodactyly, overlapping toes, pes cavus, and a limping gait. Despite these physical limitations, she had normal cognitive development and engaged well during clinical visits. Facial evaluation revealed asymmetry and vertical overgrowth, consistent with a dolichofacial pattern.

Figure 1. Extraoral Photographs

Dental & Craniofacial Manifestations

Craniofacial Features

- Facial asymmetry, long face
- Micrognathia and retrognathia
- Dolichofacial vertical growth pattern
- Cephalometric analysis: Class II skeletal pattern with mild mandibular retrusion & high mandibular plane angle

Functional Observations

- Limited mouth opening (22 mm)
- Pronounced gag reflex
- Tongue thrust and involuntary tongue movements

Intraoral Findings

- Enamel demineralization
- Gingivitis and plaque accumulation
- Multiple carious lesions
- High-arched, constricted maxilla
- Anterior open bite, bilateral crossbite

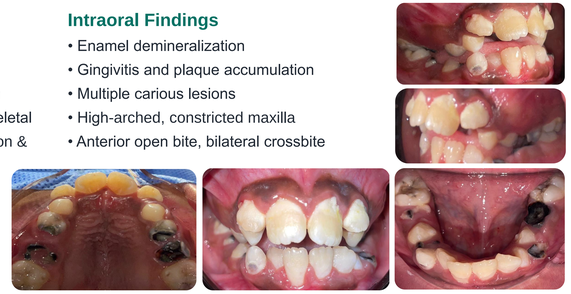

Figure 2. Pre-Treatment Intraoral Photographs

Radiographic Findings

- CBCT revealed a well-defined compound odontoma in the right mandibular body
- Displaced the unerupted permanent canine #43 inferiorly
- Caused minor perforation of the inferior border of the mandible
- The lesion was not clinically palpable and showed no swelling
- Tooth #43 had incomplete root formation but a favorable eruption path
- The lesion was not clinically palpable and showed no swelling

Figure 3. Pre-Treatment radiographs

Treatment

Comprehensive Treatment Under General Anesthesia

Restorative Management

- Composite restorations on upper first molars
- Indirect pulp capping and stainless-steel crowns on lower molars

Surgical Management

- Extraction of non-restorable primary teeth
- Full-thickness mucoperiosteal flap via envelope incision
- Extraction of retained primary canine (#83)
- Conservative bone windowing and enucleation of compound odontoma
- Preservation of permanent canine (#43) in situ
- Site irrigated and closed with 4-0 vicryl sutures

Figure 4. Odontoma surgical removal

Treatment (cont.)

Postoperative Functional Therapy

- Structured home program: lateral excursions, tongue mobility drills, and progressive tongue depressor stacking (30 seconds, 3–5 reps/session, 5x/day)
- Exercises were pain-free and caregiver-supervised
- Mouth opening improved from 22 mm to 34 mm within 3 months
- Gag reflex significantly reduced, improving procedural cooperation

Figure 5. Mouth opening before treatment - Mouth opening after treatment

Orthodontic Phase (Growth Modification)

- Initiated after improved oral access and hygiene
- Modified Hyrax appliance with tongue crib
- Lower lingual holding arch
- Removable 5mm posterior bite block to assist in mandibular autorotation and vertical control
- Radiographs showed improved positioning of #43 and new bone formation

Figure 6.

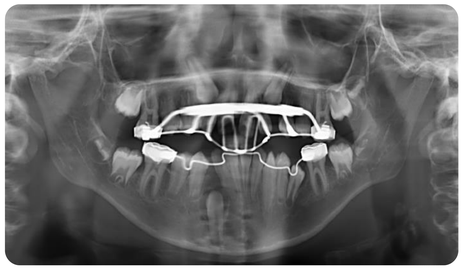

Figure 7. 3 months post surgery

Preventive Strategy

- Fluoride varnish applied every 3 months
- Tailored caregiver-led oral hygiene instruction
- Soft-bristled toothbrush and proxy brush
- Dietary counseling to minimize caries risk

Discussion

This case underscores the complexity of managing dental anomalies in AMC and presents the rare co-occurrence of AMC with compound odontoma.

Craniofacial Manifestations in AMC:

Craniofacial anomalies—such as facial asymmetry, restricted mandibular opening, and a high palatal vault—are often underrecognized in AMC, despite their significant impact on function and treatment planning.

Compound Odontoma in AMC:

Although not previously reported in AMC, the compound odontoma coincided with delayed eruption—a feature shared by both conditions. Conservative enucleation preserved the displaced canine and allowed for favorable repositioning.

Functional Challenges:

Trismus improved from 22 mm to 34 mm through structured home therapy, significantly reducing gag reflex and enhancing procedural tolerance.

Orthodontic and Preventive Care:

Timely orthodontic intervention in growing patients with AMC may reduce the severity of skeletal malocclusion over time and potentially minimize the need for future surgical correction. Preventive protocols emphasized fluoride, hygiene instruction, and caregiver engagement.

Multidisciplinary Planning:

Successful management relied on collaboration across specialties, staged care delivery, and home-based rehabilitation enabled positive outcomes despite physical and behavioral challenges.

Conclusion

This case underscores the importance of proactive, prevention-focused care and highlights the role of coordinated multidisciplinary teams in managing complex pediatric dental conditions. To our knowledge, this may represent the first documented co-occurrence of compound odontoma in an AMC patient.

References: