International Journal of Contemporary Pediatrics
Khatri A et al. Int J Contemp Pediatr. 2025 Aug;12(8):1438-1442
http://www.ijpediatrics.com
pISSN 2349-3283 | eISSN 2349-3291

DOI: https://dx.doi.org/10.18203/2349-3291.ijcp20252230

Case Report

Silver-modified atraumatic restorative technique approach to dental and craniofacial care in Apert syndrome: a case-based perspective

Amit Khatri, Rishi Tyagi, Deepak Khandelwal, Padma Yangdol, Shaikh Misbah*, Harshita Bisht, Aman Kumar

Department of Pediatric and Preventive Dentistry, University College of Medical Sciences, Guru Teg Bahadur Hospital (University of Delhi), Delhi, India

Received: 28 May 2025
Revised: 04 July 2025
Accepted: 09 July 2025

***Correspondence:**
Dr. Shaikh Misbah,
E-mail: misbah08shaikh1996@gmail.com

ABSTRACT

Apert syndrome is a rare congenital disorder first noted by Wheaton in 1894 and more precisely described by Eugene Apert in 1906. It is primarily characterized by craniosynostosis, midfacial hypoplasia, and complex syndactyly of the hands and feet, along with various systemic and oral abnormalities. The syndrome is estimated to occur in approximately 1 in 1,60,000 live births and typically follows an autosomal dominant inheritance pattern. Most cases are associated with spontaneous mutations in the fibroblast growth factor receptor 2 (FGFR2) gene, which plays a crucial role in bone development and tissue formation. This case report details the clinical and dental management of a 4-year-old female who was diagnosed with Apert syndrome at birth. The patient presented with hallmark craniofacial features such as a high, prominent forehead, flat midface, syndactyly affecting both hands and feet, and notably delayed developmental milestones. She had a history of undergoing multiple surgical interventions, including cranial vault reconstruction to alleviate intracranial pressure and syndactyly release procedures to improve hand functionality. Intraorally, the patient showed maxillary hypoplasia, a high-arched palate, delayed eruption of primary teeth, dental crowding, and multiple carious lesions. Dental care was tailored to her special healthcare needs, utilizing the silver-modified atraumatic restorative technique (SMART), which involves the use of silver diamine fluoride and glass ionomer cement to arrest caries and restore teeth in a minimally invasive manner. A preventive oral health plan, including dietary counselling, fluoride varnish application, and regularly scheduled recall visits, was instituted to maintain long-term dental and overall well-being.

Keywords: Apert syndrome, Craniosynostosis, Dental management

INTRODUCTION

Apert syndrome, initially reported by Wheaton in 1894, gained widespread recognition after Eugene Apert provided a comprehensive clinical description in 1906, leading to the eponymous naming of the disorder. Due to the historically high infant mortality associated with the condition, its overall incidence in the general population remains quite low, estimated at approximately 1 in 160,000 live births.[1] This rare congenital disorder, also referred to as acrocephalosyndactyly, is primarily characterized by craniosynostosis specifically of the coronal suture which results in premature fusion of skull bones and an abnormal head shape known as turribrachycephaly (short and tower-shaped skull).

The syndrome presents with a broad range of craniofacial, limb, and systemic abnormalities. Common craniofacial features include midface hypoplasia, ocular anomalies such as shallow orbits, exophthalmia, strabismus, hypertelorism, and down-slanting palpebral fissures. Additional manifestations may include chronic otitis media, conductive hearing loss, and obstructive sleep apnea, often due to upper airway constriction, resulting in

habitual mouth breathing. A hallmark feature is syndactyly of the hands and feet, involving partial or complete fusion of the skin and sometimes the bones of fingers and toes, often resulting in "mitten-like" hands.

Oral and dental manifestations are also diverse and include a skeletal anterior open bite, cleft palate, dental crowding, bilateral posterior crossbite, hypotonic lips, and impaired speech. Other features include supernumerary teeth, shovel-shaped incisors, enamel opacities or hypoplasia, and delayed eruption and maturation of both primary and permanent dentition.[2]

Apert syndrome follows an autosomal dominant inheritance pattern, although most cases arise from de novo mutations. Several environmental and genetic factors have been implicated, including advanced paternal age, maternal infections, drug exposure during pregnancy, and cranial inflammatory conditions. The genetic basis of Apert syndrome lies in a missense mutation in the fibroblast growth factor receptor 2 (FGFR2) gene, located at chromosome 10q26. Understanding both the genetic and environmental influences is essential for early diagnosis, multidisciplinary care, and long-term management.[3]

CASE REPORT

A 4-year-old female patient reported to the department of Pediatric and Preventive Dentistry, University College of Medical Sciences, Guru Teg Bahadur (GTB) hospital with a chief complaint of pain in his left lower front tooth region since 7 to 10 days. The diagnosis of Apert syndrome was established at birth. The patient's past medical history revealed that she underwent fronto-orbital advancement with cranial vault reconstruction, Bardach's palatoplasty for cleft palate and syndactyly release of second and fourth web space 2 years prior. Family history was negative for similar presentations or any other congenital abnormalities. Both the parents were normal and the patient is the second child of a non-consanguineous marriage. She has one sibling who is phenotypically normal. The antenatal period was uneventful, with no history of maternal trauma, infections, or drug exposure. The mother had a normal full-term delivery.

On general physical examination, the patient appeared moderately built with short stature and demonstrated a spastic gait pattern. Developmental milestones were noted to be delayed, and cognitive assessment indicated features consistent with intellectual disability. Craniofacial assessment revealed a turribrachycephalic (abnormally tall and broad) configuration accompanied by occipital flattening, anterior fontanelle within normal limits, prominent ocular proptosis, increased interorbital distance (hypertelorism), and a downward slant of the lateral palpebral fissures, consistent with dysmorphic facial features (Figure 1). She had midfacial deficiency with hypoplastic and retruded maxilla. Bilateral, symmetrical syndactyly was observed, involving complete fusion of all five digits on both hands, along with medially deviated thumbs. The second and fourth interdigital spaces of the left hand had been surgically separated during a procedure performed two years earlier. Also, syndactyly was present with both feet with deformation of the great toe. The fused fingers and toes had separate nails (Figure 2). The radiographic representation of digit syndactyly is also shown in Figure 2.

Figure 1: Extraoral frontal and lateral view showing a turribrachycephalic (abnormally tall and broad) head with occipital flattening, prominent ocular proptosis, increased interorbital distance (hypertelorism), and a downward slant of the lateral palpebral fissures.

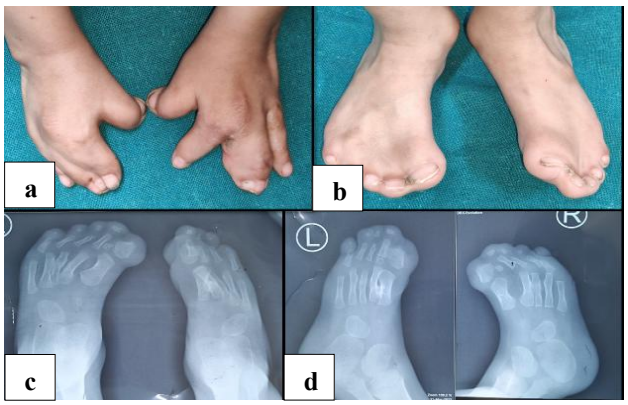

Figure 2: (a and b) Clinical photographs of bilateral, symmetrical syndactyly of all five digits of both hands and feet except the second and fourth interdigital spaces of the left hand (that had been surgically separated during a procedure performed two years earlier), and (c and d) anterio posterior radiograph showing bilateral, symmetrical syndactyly of both hands and feet.

Oral examination demonstrated maxillary hypoplasia, a high-arched palate, v-shaped maxillary arch, gingival swelling with respect to upper primary molars, open bite, significant crowding, delay in eruption of permanent dentition, and caries with respect to tooth 73 (Figure 3).

Management

Dental management for the carious tooth 73 (ICDAS 3) included the use of the SMART technique, a minimally invasive approach for managing dental caries in children,

especially in special healthcare needs children and children with behavioral or developmental challenges.

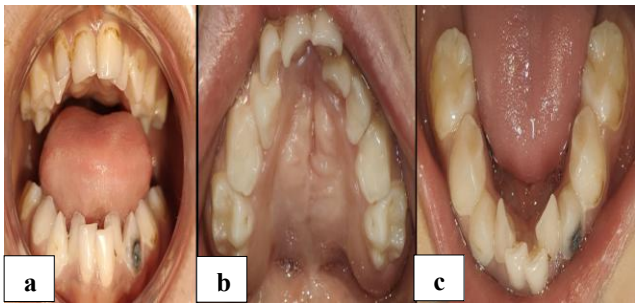

Figure 3: (a) Intraoral frontal view depicting open bite, (b) intraoral view of maxillary arch showing v-shaped arch, gingival swelling with respect to upper primary molars, and crowding, and (c) intraoral view of mandibular arch depicting significant crowding and caries with respect to tooth 73.

Table 1: Key characteristics depicting the clinical and radiographic manifestations of Apert syndrome seen in the present case.

System/feature	Clinical and radiographic findings
Craniofacial	Craniosynostosis, turribrachycephaly (tall, short skull), midface hypoplasia, flat forehead and occiput, depressed broad nose with bulbous tip and deviated septum
Opthalmologic	Shallow eye socket (ocular proptosis), hypertelorism (wide-spaced eyes), down-slanting palpebral fissures, exopthalmia
Limbs	Symmetric 2nd to 4th digit syndactyly in hands and feet (mitten hands and sock feet)
Neurologic	Mild mental/intellectual deficit, developmental delay
Respiratory	Mouth breathing
Skin	Excessive sweating
Dental	Skeletal anterior open bite, soft palate cleft, narrower dimensions of both dental arches (v-shaped) with crowding, gingivo/ periodontal alterations, hypotonic (upper) lip, impaired speech, delay in maturation/eruption in primary and permanent teeth

The technique combines two key materials: silver diamine fluoride and glass ionomer cement, designed to arrest decay and restore teeth without the need for drilling or anaesthesia. Firstly, silver diamine fluoride was applied directly to the carious (decayed) area of the tooth. It works by penetrating the decayed tooth structure, stopping bacterial growth and preventing further demineralization. It has the added benefit of remineralizing the enamel and dentin, making the tooth more resistant to future decay (Figure 3).

After the application of silver diamine fluoride**,** glass ionomer cement was used to fill the carious lesion and restore the function and appearance of the tooth (Figure 3). Glass ionomer cement was preferred because it adapts well to the cavity, is easy to apply, and is particularly effective in pediatric patients who may not tolerate more complex procedures. It is a material that bonds chemically to the tooth structure and releases fluoride over time. This fluoride release helps protect the tooth from further decay and encourages remineralization of the surrounding enamel. The material was placed directly over the decayed area after SDF treatment and is allowed to set. The procedure is intended to be minimally invasive, quicker, and more comfortable for pediatric patients. A comprehensive preventive and oral hygiene maintenance plan were implemented, accompanied by scheduled recall appointments to ensure continued dental health and follow-up care.[4,5]

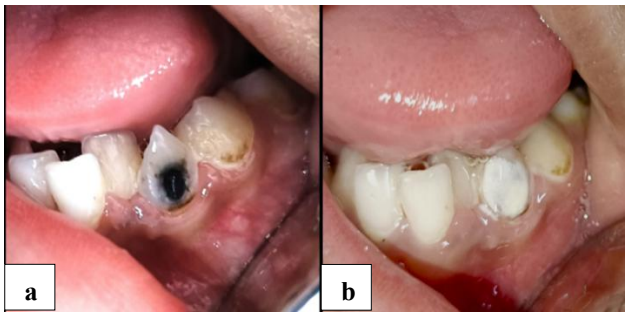

Figure 4: (a) After application of silver diamine fluoride on carious tooth 73, and (b) final postoperative clinical picture showing glass ionomer cement in place to restore the function and appearance of the tooth.

DISCUSSION

The present case underscores the classical presentation of Apert syndrome and emphasizing the importance of timely surgical, dental, and developmental interventions. Craniofacial features and systemic findings, including midfacial hypoplasia with a retruded maxilla, turribrachycephaly with occipital flattening, ocular proptosis, hypertelorism, downward-slanting palpebral fissures, and developmental delays, were characteristic of a syndromic presentation consistent with Apert syndrome.[6]

Understanding the phenotypic spectrum of Apert syndrome is essential for accurate diagnosis and differentiation from other craniofacial syndromes. Other differential diagnoses include syndromes with overlapping craniofacial characteristics, such as Crouzon, Carpenter, and Pfeiffer syndromes. Apert syndrome and Crouzon syndrome represent the two most common forms of syndromic craniosynostosis, collectively classified under

the term acrocephalosyndactyly, and together comprise approximately 70% of all reported cases. While all may involve craniosynostosis, Apert syndrome is uniquely distinguished by the presence of complex syndactyly of the hands and feet.[7,8]

Moreover, Apert and Crouzon syndromes both show reduced middle cranial fossa length and midface shortening, particularly in the upper segment. Apert syndrome features rotated zygomatic bones, while Crouzon syndrome shows shorter zygomatic bones without rotation. Differences in maxillary, zygomatic, and palatal deformities contribute to unique midface malformations with varying severity in each syndrome.[9] In contrast, Crouzon syndrome typically lacks limb anomalies, Carpenter syndrome often includes polydactyly and intellectual disability, and Pfeiffer syndrome is characterized by broad thumbs and toes.[7,8]

The majority of molecularly characterized cases of Apert syndrome are attributed to two specific point mutations in the fibroblast growth factor receptor 2 (FGFR2) gene, located at 10q26 on chromosome 10. These mutations are C-to-G transversions occurring at adjacent codons within exon IIIa of the gene. The first, a C934G transversion, alters the codon from TCG to TGG, resulting in a serine-to-tryptophan substitution at amino acid position 252 (S252W or Ser252Trp). The second, a C937G transversion, changes the codon from CCT to CGT, leading to a proline-to-arginine substitution at position 253 (P253R or Pro253Arg).[10,11]

Table 2: Mechanism of action of each component used in SMART technique.

Procedure	Mechanism of action
Silver diamine fluoride is applied to the decayed area	It works by arresting caries and preventing further progression through its antimicrobial properties
Glass ionomer cement is used as a restorative material	GIC releases fluoride, which helps in remineralization and provides additional protection against future caries.

Intraoral examination demonstrated maxillary hypoplasia with a constricted V-shaped arch. The patient has a documented history of cleft palate repair via Bardach palatoplasty, a two-flap technique involving mucoperiosteal flap elevation and repositioning to achieve functional and anatomical closure of the palatal defect. This technique represents a modification of the von Langenbeck palatoplasty, wherein incisions are placed along both the cleft and alveolar margins, meeting anteriorly to facilitate elevation of mucoperiosteal flaps. These flaps are pedicled on the greater palatine neurovascular bundles, preserving vascular integrity. The soft palate is closed in a straight-line fashion, and levator veli palatini muscle dissection with reconstruction of the muscular sling is performed akin to intravelar veloplasty. This approach is widely adopted in contemporary cleft palate repair practice.[12]

The patient also presented anterior open bite, significant dental crowding, delayed eruption of permanent teeth, and dental caries in the primary mandibular left canine (tooth 73), indicating the need for early preventive and restorative intervention. Given the patient's limited cooperation, multiple dental needs, and syndromic presentation, a minimally invasive approach was prioritized. The silver modified atraumatic restorative technique (SMART) was selected for managing cavitated carious lesions, particularly for the primary mandibular left canine (tooth 73), which exhibited active decay.[4,13] The SMART technique involves the application of 38% silver diamine fluoride to arrest caries, followed by restoration with glass ionomer cement to seal the lesion and restore function. This method avoids the use of rotary instruments and local anaesthesia, making it well-suited for pediatric patients with behavioral or developmental challenges.

CONCLUSION

This case report contributes to the field of pediatric dentistry and craniofacial anomaly management by illustrating how minimally invasive approaches, specifically the SMART can be effectively adapted for patients with complex syndromic conditions such as Apert syndrome. It highlights the flexibility of dental treatment strategies in addressing the distinct anatomical and developmental challenges these patients face. Additionally, the case reinforces the value of integrating preventive care and multidisciplinary collaboration as a foundation for long-term, comprehensive management. It further emphasizes the critical role of early dental intervention and personalized treatment planning in enhancing both oral health and overall quality of life in children with craniofacial disorders.

ACKNOWLEDGEMENTS

Authors would like to acknowledge all the study investigators for conducting this study, manuscript review and edit support.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Khatri A, Tyagi R, Khandelwal D, Yangdol P, Misbah S, Bisht H, et al. Silver-modified atraumatic restorative technique approach to dental and craniofacial care in Apert syndrome: a case-based perspective. Int J Contemp Pediatr 2025;12:1438-42.