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Case Report

# Macrodontic maxillary incisor in alagille syndrome

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## ABSTRACT

This case report describes the surgical-orthodontic guided-eruption of a deeply impacted macrodontic maxillary central incisor in a 10-year-old patient with Alagille syndrome (ALGS). In the first stage, orthodontic treatment with fixed appliance on deciduous teeth allowed to create enough space for the eruption of the maxillary right central incisor. The second stage included closed surgical exposure and vertical traction. After impacted tooth erupted in the proper position, accessory periodontal treatment and dental reshaping procedures may be indicated to camouflage macrodontic incisor with the adjacent teeth. This is the first report that presents a patient with ALGS undergoing orthodontic and surgical treatment.

**Key Words:** Alagille syndrome, impacted teeth, macrodontia

## INTRODUCTION

Alagille syndrome (ALGS) is an autosomal dominant disorder caused by heterozygous mutation in the JAG1 (Jagged 1) gene on chromosome 20p12. Its estimated prevalence is about 1 out of 70,000 births, when ascertained by the presence of neonatal jaundice.[1] It has been traditionally defined by a paucity of intrahepatic bile ducts, in association with five main clinical abnormalities: Cholestasis, cardiac disease, skeletal abnormalities, ocular abnormalities, and a characteristic facial phenotype.[2]

Abnormalities were reported in different systemic organs: In the eye, posterior embryotoxon and retinal pigmentary changes were observed; in the heart, pulmonic valvular stenosis as well as peripheral arterial stenosis; in the bones, abnormal vertebrae ("butterfly" vertebrae) and decrease in interpediculate distance in the lumbar spine; in the nervous system, absence of deep tendon reflexes and difficulty learning; in the faces, characteristic traits such as broad forehead, deep-set eyes, prominent nose, pointed mandible and chin, bulbous tip of the nose, and varying degrees of fingers foreshortening.[3] Unilateral coronal craniosynostosis was also reported in two patients with mutation-proven ALGS.[4] No gene mutations associated with craniosynostosis was found, therefore, JAG1gene was suggested to play a possible role in cranial suture formation. Furthermore, severe hypodontia was described in a 3-year-old Asian boy with ALGS.[5]

Several genetic studies have been carried out during the years.[6,7] Twenty-two percent of the patients with ALGS had spontaneous or post-procedure bleeding in various organs and were subjected to special risk for bleeding.[8] Therefore, it can be speculated that abnormalities in the JAG1-gene may impair hemostatic function. Nine percent had vascular anomalies or events, and vascular accidents accounted for 34% of mortality. Vascular anomalies including basilar and middle cerebral artery aneurysms, internal carotid artery anomalies, aortic aneurysms, and co-arctation of the aorta were documented in ALGS patients.[9] Furthermore, latex-hypersensitivity and severe susceptibility to dental decays or decalcifications on the posterior deciduous teeth were described.[10]

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This report describes a combined surgical-orthodontic procedure for guided-eruption of a deeply impacted macrodontic maxillary central incisor in young ALGS patient. Although an interdisciplinary approach was needed, surgical procedure may be complicated by the severe risk of increased bleeding and vascular events. None of the studies dealt with orthodontic surgical treatment performed in an ALGS patient.

## CASE REPORT

### Diagnosis and treatment plan

A 10-year-old male was presented to our dental office complaining of delayed eruption of a maxillary central incisor [Figure 1]. His medical history indicated a diagnosis of ALGS with no previous systemic treatment or complication. Clinical examination revealed characteristics ALGS traits: Broad forehead, deep-set eyes with eye-socket, pointed mandible and prominent chin; maxillary hypoplasia, maxillo-mandibular retrusion; and increased vertical facial dimension were also noted. He presented a Class I molar relationship on both side, a correct overjet and a deep overbite (90%). Absence of maxillary right central incisor with no adequate space in the maxillary arch was noted. Crowns of the adjacent teeth had slightly drifted into the unoccupied space. An intra-oral examination revealed the presence of several decalcifications on deciduous teeth with previous conservative treatment on first permanent molars and maxillary first deciduous molar on right side. Panoramic radiograph showed a macrodontic maxillary right central incisor vertically retained.

Cephalometric analysis indicated a Class I skeletal pattern with retrognathic maxilla and mandible, prominent chin, and slight hyperdivergent tendency.

Treatment objective was to create space in the maxillary arch and allow the eruption of the impacted central incisor. Surgical exposure and orthodontic traction with a fixed appliance was planned.[11]

Risk of increased bleeding, vascular events, and latex hypersensitivity were the major concern, requiring the use of latex-free materials, controlled hemostatic procedure, and antibiotic prophylaxis as recommended by the cardiologist.

### Treatment progress

Bands on deciduous second molars and Begg's brackets on permanent incisors and deciduous canines were placed. After alignment and levelling were completed using 0.014″ nickel–titanium archwire, a relatively rigid stabilizing wire (0.018″ round Australian wire) was placed, and a coil spring was used to create adequate space between the central incisor and the right lateral incisor. Periodontal surgery was performed to expose the maxillary right central incisor. An access flap was elevated to expose the impacted tooth and a lingual button connected to a ligature wire was bonded to the palatal surface. A closed surgical exposure technique was chosen. An elastic module for applying an apically directed force was applied to the stabilizing wire [Figure 2]. Vertical traction allowed the incisor eruption to be guided along the alveolar ridge. Once crown of the maxillary incisor adequately appeared in the maxillary arch, a Begg bracket was bonded to the vestibular surface and alignment and levelling were completed using 0.014″ nickel titanium wire, and later replaced by 0.018″ round Australian wire [Figure 3]. Active treatment took 9 months.

The impacted maxillary right central incisor was brought into proper alignment with the adjacent teeth. Once alignment was achieved, greater height and crown width of the macrodontic incisor caused gingival margin and incisal border alterations [Figure 3]. Conservative treatment and minimal restorative interventions,[12] accessory periodontal treatment, and dental reshaping procedures[13] may be indicated to re-establish the proper size and dimension of the macrodontic incisor.

## DISCUSSION

ALGS represents a genetic disorder involving complications in different organs. The major concern in treating this patient was the cardiological and vascular risk following surgical periodontal procedure, such as dental implant insertion,[14-17] since high impaction position of a retained tooth may often require a traumatic surgery. Increased bleeding, vascular events, and latex hypersensitivity were previously reported in ALGS patients,[8-10] therefore, the use of latex-free materials and controlled hemostatic procedure were mandatory, even if these patients reported no previous medical history of specific hypersensitivity or hemostatic dysfunction. In agreement with cardiological recommendations, it was decided that antibiotic prophylaxis was not required since surgical procedure was not considered a risk for bacterial endocarditis.

In accordance, closed surgical exposure technique was chosen for two reasons. First, closed-eruption technique may often avoid a second surgical procedure

**Figure 1:** Pretreatment photographs. A 10-year-old male with history of Alagille syndrome, presence of macrodontic central incisor vertically retained were shown

**Figure 2:** Closed surgical exposure and vertical traction of the central incisor

**Figure 3:** Post-treatment photographs 9 months later. Macrodontic incisor caused gingival margin and incisal border incongruence with the adjacent teeth

or re-intervention, second, it usually provides the most esthetically pleasing result since it preserves as much as possible gingival margin contour of the central incisor and adequate zone of attached gingival.[18] Open surgical exposure has the disadvantage of producing a non-keratinized vestibular gingival margin during vertical traction.[19] Using this technique, papillary apical positioned partial-thickness flap during the orthodontic traction may be needed to achieve an esthetic gingival margin contour over the central incisors and provide the teeth with an adequate zone of attached gingival labial to the crown.[18] Furthermore, because of the relatively high prevalence of gingival defects in some studies, adjunctive post-orthodontic periodontal surgery might be required in many patients treated with this method.[19]

An impacted maxillary central incisor in a child often represents, in addition to a disturbing esthetical problem, the parent's main concern according which a patient can be referred to the orthodontist to evaluate the possibility of a disturbance eruption. Orthodontic treatment has not been usually postponed until the complete eruption of the permanent dentition especially when the problem can be treated in the early mixed dentition stage.[18] However, it is important to properly inform the patient about the possibility of failure before undertaking extensive measures necessary for the management of a severely impacted tooth.[20] Factors suggesting clinical success may be considered on the basis of position and orientation of the impacted tooth, amount of root formation, and degree of root dilacerations.[21] Brand *et al*.[20] and Sabri[22] reported that movement of an impacted central incisor could be impossible because of ankylosis and external root resorption.

Ho and Liao[23] reported that unsuccessful predictors for impacted central incisors were older age, highimpacted tooth, and dilacerated incisors. In this case, high impaction in the maxillary bone was the only negative factor increasing the likelihood of unsuccessful treatment result. However, central incisor has been successfully brought into proper position after having provided sufficient space in the maxillary arch, but an unavoidable unesthetic higher gingival margin in respect to the adjacent teeth was present after alignment; this was due to the different crown size of the macrodontic central incisor.[19] In fact, problems related to macrodontic tooth can include difference in size and shape, space deficiency, interproximal contact points with the adjacent teeth, and incongruence in gingival margin or incisal border.

Nevertheless, this case cannot be considered as a true macrodontia; when true macrodontia occurs in the incisal region, it can often be confused with conjoined teeth or fusion of teeth, since the union of two or more teeth results in a single large tooth during the

odontogenesis.[24] The main difference is that size reduction of macrodontic central incisors by proximal grinding is not possible without jeopardizing tooth vitality. In this case, reshaping procedure or minimal restorative interventions such as indirect composite veneers may be indicated.[25] The esthetic outcome of these procedures can be excellent without unnecessary loss or teeth extraction. Knowledge of the use of indirect composite veneers can provide another treatment option for esthetic problems in the management of tooth anomalies, including macrodontia.[12]

## CONCLUSION

In conclusion this case report demonstrates that is possible, controlling carefully the procedure, to perform a combined surgical and orthodontic treatment in an ALGS patient.

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