Isolated Congenital Aglossia in a 20-Month Child: A Case Report

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ABSTRACT

The failure of the lateral lingual swellings to form between the fourth and eighth week of development results in aglossia. Other congenital abnormalities frequently coexist with it. A case of a 20-month infant girl with the complaint of not having a tongue since birth presented to the plastic surgery outpatient department. Following a thorough history-taking, examination, and laboratory studies, we came to the conclusion that isolated aglossia does not need special care because the surrounding tissues can compensate for the tongue's absence when swallowing and eating. However, managing such illnesses requires a multidisciplinary team approach in order to improve the patient's quality of life and manage any early concerns.

Key Words: *Aglossia, Congenital anomaly, Tongue development.*

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INTRODUCTION

Congenital aglossia is a rare condition that results from the failure of tongue development between the fourth and eighth weeks of gestation, and results in absent tongue at birth. The tongue is the most mobile organ in the body and is essential for coordinated functions including speaking, chewing, swallowing, and sucking as well as for the healthy growth of the jaws and teeth.[1] This deformity can occur in isolation or, more frequently, in conjunction with other syndromes or systemic abnormalities, such as limb hypoplasias and a variety of oromandibular disorders.[2] In the eighteenth century (1718/1719), de Jussieu wrote the first description of aglossia congenita.

A case of a 20-month infant girl with congenital aglossia since birth is presented in this case report.

CASE REPORT

The parents of a 20-month child brought her to the outpatient department, stating that she had an absent tongue since birth. There were no problems during the mother's pregnancy or the child's birth, and she was delivered at full term by a routine vaginal delivery. The absence of her tongue was noticed during crying and feeding. With the exception of a short mandible, her facial characteristics were normal (Figure 1).

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The measurements revealed that the distances were as follows: 6.5 cm on both sides from the angle of the mandible to the symphysis menti, 3 cm from the temporomandibular joint (TMJ) to the angle of the mandible, 5 cm on both sides from the lateral canthus to the tragus, and 7 cm from the oral commissure to the tragus. An acceptable oral opening was found during an intraoral examination, and the tongue was completely absent. Both the upper and lower jaws contained teeth, *albeit* some of them were carious (Figure 1). The remaining oral tissues, including the palate, uvula, and gums, seemed healthy. She had never experienced a feeding issue, even at birth. Her parents claimed that she was limited to speaking *baba* and *nana*. The systemic evaluation and general physical examination were unremarkable. The three other siblings were normal, and her parents did not have any unusual family history of congenital defects or consanguinity. The patient's radiography also showed mandibular hypoplasia (Figure 2).

DISCUSSION

Although the precise aetiology is still unknown, aglossia can be explained embryologically by the tuberculum impar not developing and the two lingual swellings failing to grow.[3] The literature does, however, provide a number of possible explanations, such as a lack of folic acid, infection, febrile illness in the mother, hypothyroidism, or vascular disturbances during tongue development.[4] This aberration has also been linked to chorionic villous sampling carried out prior to the 10th week of gestation.[5] In this case, the mother's medical history at the time of her pregnancy did not include any of these concerns. Aglossia has been found to be associated with different syndromes: Moebius syndrome, Goldenhar syndrome, Aglossia adactylia syndrome, *etc*. Few cases of mental retardation have also been reported.[5] These conditions were not present in this patient. Hall's classification of oromandibular limb hypogenesis syndrome (OLHS) divides the spectrum of manifestations into five types and is widely accepted.[3]

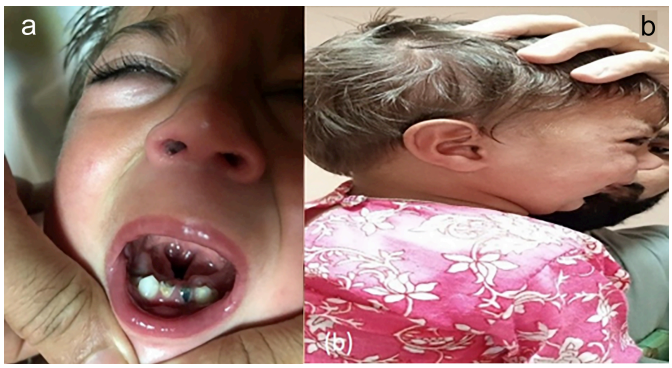

Figure 1: (A) Intraoral view shows an absence of tongue and carious teeth. (B) Lateral view of the patient showing micrognathia.

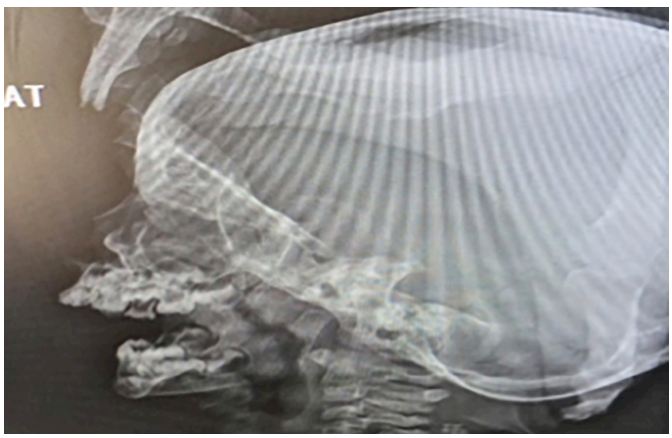

Figure 2: X-ray, lateral view of the face showing hypoplastic mandible.

Based on this classification, our patient was classified as type 1B. Sucking, speaking, mastication, swallowing, taste perception, jaw growth, and occlusion all depend heavily on the tongue. Although a few cases reported by Thorp *et al*. required tube feeding and even tracheostomy, which fortunately our patient did not need. Most individuals with aglossia do not have swallowing or breathing difficulties.[6] Speech and swallowing are two examples of activities that often get better with time because of the orofacial structures' adaptive mechanisms. These systems improve taste perception, speaking, swallowing, and feeding assistance. The adaptive capacity could potentially account for the hypoplastic mandible, absence of breastfeeding issues, and limited speech sound production, including "*baba*" and "*nana*".[7] This patient, who was a case of isolated aglossia, had only associated hypoplastic mandible. No other anomalies or the above- mentioned syndromes were present. She had normal facial structures, normal limbs, no feeding problems, and had normal response. However, multidisciplinary team involvement is necessary for dentofacial deformities such as developing malocclusion and maldeveloped mandible. The patient was then referred to the maxillofacial department for assessment of the hypoplastic mandible. As the short mandible affects facial aesthetics, so we will follow the patient for any surgical intervention if she needs it in the future.

Although only a few cases of aglossia have been reported, nevertheless, facial sequelae of aglossia compromise facial aesthetics and severely affect psychological well-being.
So, involvement of a multidisciplinary team approach is essential for improving patients' quality of life, and thus mitigating parents' psychological trauma.

PATIENT'S CONSENT:
Informed consent was obtained from the patient's parents to publish the data concerning this case.

COMPETING INTEREST:
The authors declared no conflict of interest.

AUTHORS' CONTRIBUTION:
JUH: Conception and design.
WK: Drafting of the manuscript.
SS: Acquisition of data.
MAS: Critical evaluation and final approval.
All authors approved the final version of the manuscript to be published.

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