

## Case Report



## Ankyloglossia superior syndrome: a challenging case report and literature review

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## Abstract

Ankyloglossia superior (palatoglossal adhesion) is an exceedingly uncommon congenital disorder, with just 20 cases recorded in the medical literature. This abnormality is considered to be a part of the ankyloglossia superior syndrome (ASS) when it occurs with other congenital abnormalities such as gastrointestinal anomalies, cleft palate, and limb deformities. We describe the uncommon occurrence of a cleft palate, limb deformity, and ASS with a synechial band extending from the incisive foramen to the tip of the tongue. In addition, surgical repair is detailed. Considering the origin of ASS, we reviewed the current research and explored several hypotheses. When a newborn is diagnosed with ankyloglossia superior, clinicians must have a high level of suspicion for additional congenital defects.

## Introduction

The fusion of the tongue and hard palate is a defining characteristic of ankyloglossia superior. Ankyloglossia superior was initially reported by Illera in 1887 and Kramer characterised it in 1911.<sup>1</sup> Only 20 cases of ankyloglossia superior, an extremely rare congenital illness, have been reported. In this article, we discuss a newborn with limb deformities, ankyloglossia superior syndrome (ASD), a cleft palate, and tongue bifida.

## Case Report

We report the case of an Iranian female neonate, born to a healthy 33 years mother G2P2 on March 2, 2022. She was born through cesarean section after 39 weeks of gestation, weighing 4300 g, and measuring 53.5 cm in length. The baby's Apgar score was 9 at 1 minute and 10 at 5 minutes. The obstetrician observed malformation of the limbs and a link between the tongue tip and palate. She had a feeding issue and was fed by nasogastric tube (NGT) despite no signs of respiratory distress were observed. She had superior ankyloglossia and was referred to our department on the second day of life. The mother had no previous medical conditions or drug use. The mother denied smoking and drinking during pregnancy. She received the right prenatal care and went to the physician's office before giving birth. Prenatal ultrasounds revealed no indication of congenital abnormalities. She had one healthy sibling and no prior family medical history.

The physical examination revealed superior ankyloglossia, which resulted in an anterior oral band and intraoral occlusion (Figure 1). There was a restriction on mouth opening and a wide midline anterior oral band that obstructed the view of the oropharynx. Laboratory testing and the CXR were both normal. Additionally, only two phalanges were visible in the left hand's second and fourth fingers (Figure 2). Additionally, echocardiography revealed ASD, but other body parts were normal and there were no other congenital defects.

Due to the presence of a large midline anterior mouth band, oral intubation was not possible. After that, we released the tissue linking the tongue to the hard palate under local anaesthetic (Figure 3). After removing fibrous bands, we discovered a bifid tongue and a wide cleft palate. Sutures were used to reconstruct the bifid tongue. After achieving hemostasis, 4-0 Vicryl sutures were used to enclose the wound edges. NGT was removed and mouth openness was improved. One day following the operation, the baby was spoon-fed. The procedure had no postoperative complications, and the patient was discharged from the hospital five days following the surgery with a satisfactory postoperative outcome. The patient had no trouble resuming oral intake at the postoperative checkup and showed no signs of misarticulation or dysarthria. The tongue mobility had changed by the time the patient underwent a second evaluation one month after the initial procedure. The cleft palate is corrected in

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**Figure 1.** Ankyloglossia superior: Tongue fused to the palate



**Figure 2.** The congenital lip deformity at the left hand

10 months after a monthly check-up with the patient.

### Discussion

A disorder known as ankyloglossia is characterized by a short, thick lingual frenulum that attaches the tongue to the floor of the mouth, whereas superior ankyloglossia attaches the tongue's tip to the palate. Superior ankyloglossia are related to aglossia-adactylia, hypodactylia, microglossia, Hanhart syndrome, syndactyly, Pierre Robin syndrome, ankyloglossia, Moebius syndrome, amniotic band syndrome, and Charlie M syndrome.<sup>2</sup> Mandible, tongue, maxilla, and limbs are all affected by the aforementioned defects. Similar defects were superseded by the oromandibular limb hypogenesis syndrome, which were classified by Hall into five groups in 1971. The third group was assigned to superior ankyloglossia.<sup>3</sup> Although superior ankyloglossia



**Figure 3.** After the tip of the tongue has been released

and aglossia adactylia share some characteristics, such as micrognathia hypoglossia or aglossia, cleft palates, or limb abnormalities, superior ankyloglossia can be distinguished from aglossia adactylia by the fibrotic band 135 years after Iller originally described the condition, the cause is still unclear. According to a number of theories about the pathogenesis of superior ankyloglossia, all anomalies occur during embryo development.<sup>2</sup> Between the eighth and eleventh weeks of pregnancy, ankyloglossia superior syndrome is thought to have started to develop inside the uterus.<sup>4</sup> At around the fourth week of pregnancy, the tongue starts to develop as two lateral lingual swellings and one medial swelling, tuberculum impar. The first pharyngeal arch is where the lingual swelling originates; in fact, the first pharyngeal arch is where the front two-thirds of the tongue is generated. The posterior of the tongue is made up of the second third, and a part of the fourth arch. Two shelf-like outgrowths from the maxillary protrusion that began growing in the seventh week make up the secondary palate. The secondary palate develops as the palatal shelves move from a vertical to a horizontal position above the tongue and fuse between weeks six and seven.<sup>5</sup> Knowing how the mouth cavity develops could help pinpoint when superior ankyloglossia malformations first appeared. The pathophysiology of superior ankyloglossia has been the subject of several theories. A cleft palate and tongue adhesion to the palate can result from sedative medicines depressing the fetal swallowing reflex, which can also delay palatal shelf elevation, extend palatopharyngeal and palatoglossal epithelial contact, and delay palatal shelf elevation.<sup>6</sup> According to the most widely accepted hypotheses, these bands are leftovers from the buccopharyngeal membrane. The thin membrane that divides the early pharynx from the mouth is called the buccopharyngeal membrane. The layer eventually disappears, but the tongue's motility was constrained by the buccopharyngeal membrane, which remained.<sup>2</sup> The placement of the tongue between the

palatal shelves prevents the fusion of the palatal shelves, which results in a cleft palate.<sup>7,8</sup> According to another hypothesis, the adhesion between the tongue's tip and the palate is caused by epithelial lesions of the tongue. The development of the tongue, palate, and limb was hampered by epithelial.<sup>9</sup> According to another hypothesis, amniotic bands syndrome may be brought on by placental injury. Numerous congenital malformations, including limb or digital amputations, can result from the fiber band of the amniotic sac.<sup>10</sup> Missense mutations in TX22 (the T.box transcription factor) can result in a cleft palate and ankyloglossia. The T.box gene is crucial for palatogenesis.<sup>11</sup>

Superior ankyloglossia may be caused by a variety of environmental and inherited variables.<sup>12</sup> Urinary tract infections, drugs, vaginal hemorrhage, hypoxia, radiation, hypoglycemia, rubella, and avitaminosis are the examples of environmental and inherited variables that may be connected to superior ankyloglossia.<sup>2,13,14</sup> In prior studies, 10 cases had cleft palates as a clinical characteristic, 13 had superior ankyloglossia with limb abnormalities, one had no further abnormalities, and two had congenital heart defects. In the majority of cases, general anesthesia was employed. Ours is the described example of superior ankyloglossia coupled with cleft palate, limb abnormalities, ASD, and tongue bifida (Table 1). One of the most difficult clinical scenarios is the problematic airway in the superior ankyloglossia because direct laryngoscopy is impossible due to the fibrous bands

and direct laryngoscopy via endotracheal intubation is difficult and debatable. Numerous techniques have been employed to induce anesthesia in patients with superior ankyloglossia, including oral intubation using a paraglossal route, direct oral intubation, nasal intubation using a flexible pediatric bronchoscope, blind nasal intubation, and local anesthetic (Table 1), among them, we used local anesthesia in our case. A laryngeal mask is not ideal because it is easily misaligned.<sup>15</sup>

Surgery for superior ankyloglossia is scheduled based on the patient's clinical status. In situations of elective surgery, the surgeon can hold off until the neonate has gained weight for a few weeks.<sup>16</sup> Neonates with breathing or feeding issues require emergency surgery. However, difficulties with the temporomandibular joint result in prolonged surgery delays.

### Conclusion

ASD is an uncommon congenital condition associated with craniofacial malformations, limb deformities, and gastrointestinal atresia. With initial multidisciplinary therapy, children with specific congenital abnormalities grow without substantial physical handicaps.

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**Table 1.** Summary of all reported cases of ankyloglossia superior syndrome

| Gender | Age at operation | Limb anomalies | Cleft palate/defect | Other anomalies                                         | Anesthesia   | Ref.     |
|--------|------------------|----------------|---------------------|---------------------------------------------------------|--------------|----------|
| Male   | 2 days           | ✓              | ✗                   | Tongue hypoplasia                                       | Not reported | 13       |
| Male   | Not reported     | ✓              | ✗                   | Facial nerve paralysis, micrognathia, microglossia      | Local        | 2        |
| Male   | Not reported     | ✓              | ✓                   | GI anomaly, microglossia                                | General      | 14       |
| Female | 2 days           | ✓              | ✗                   | ✗                                                       | General      | 16       |
| Male   | 14 days          | ✗              | ✓                   | ✗                                                       | Local        | 9        |
| Male   | 1 year           | ✓              | ✗                   | Microglossia, micrognathia                              | General      | 17       |
| Female | 5 months         | ✓              | ✗                   | Micrognathia, hypoglossia                               | General      | 10       |
| Female | 45 days          | ✗              | ✓                   | Strabismus, micrognathia                                | General      | 12       |
| Female | 2 days           | ✗              | ✓                   | Bifid tongue                                            | Local        | 18       |
| Male   | Not reported     | ✗              | ✗                   | ✗                                                       | Not reported | 19       |
| Female | 25 days          | ✓              | ✗                   | Facial paralysis, GI, abnormality, abducens nerve palsy | General      | 20       |
| Male   | 5 years          | ✓              | ✓                   | ✗                                                       | Local        | 21       |
| Female | Not reported     | ✓              | ✓                   | ✗                                                       | General      | 15       |
| Male   | 5 days           | ✓              | ✗                   | Micrognathia                                            | General      | 22       |
| Female | 0 days           | ✓              | ✗                   | ✗                                                       | Local        | 23       |
| Male   | 1 days           | ✗              | ✓                   | Micrognathia, ASD, VSD                                  | General      | 24       |
| Female | 5 months         | ✓              | ✓                   | Micrognathia, moebius, syndrome, strabismus             | General      | 25       |
| Female | 7 days           | ✗              | ✓                   | Abnormal, genitalia, PDA                                | General      | 26       |
| Female | 42 days          | ✓              | ✓                   | Microglossia, natal teeth, micrognathia                 | General      | 27       |
| Female | 2 days           | ✓              | ✓                   | ASD, bifid tongue                                       | Local        | Our case |

PDA: Patent Ductus Arteriosus; ASD: ankyloglossia superior syndrome

**Authors' Contribution****Conceptualization:** Shahin Abdollahifakhim.**Data curation:** Faezeh Ghaderyan.**Formal analysis:** Faezeh Ghaderyan.**Funding acquisition:** Shahin Abdollahifakhim, Faezeh Ghaderyan.**Investigation:** Shahin Abdollahifakhim, Hassan Soleimanpour.**Methodology:** Shahin Abdollahifakhim, Faezeh Ghaderyan.**Project administration:** Faezeh Ghaderyan.**Resources:** Shahin Abdollahifakhim.**Software:** Hassan Soleimanpour.**Supervision:** Shahin Abdollahifakhim.**Validation:** Hassan Soleimanpour.**Visualization:** Faezeh Ghaderyan.**Writing—original draft:** Faezeh Ghaderyan.**Writing—review & editing:** Hassan Soleimanpour.**Competing Interests**

The authors declare that they have no competing interests.

**Consent for Publication**

Written informed consent was obtained from the parents of patient to publish this case report and accompanying images.

**Data Availability Statement**

The datasets generated and analyzed during the current study are not publicly available due to legal reasons but are available from the corresponding author, H.S. on reasonable request.

**Ethical Approval**

Not Applicable.

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