

## Severe congenital hypoplasia of tongue and abnormal shape of soft palate and tonsillar pillars

A. Rachmiel<sup>1</sup>, I.T. Jackson<sup>1</sup>, S.R. Sabapathy<sup>2</sup> and R.A. Forté<sup>1</sup>

<sup>1</sup> Institute for Craniofacial and Reconstructive Surgery Affiliated with Providence Hospital, Southfield, Michigan, USA

<sup>2</sup> Raja Sabapathy, Ganga Hospital, Swarnambika Lay-out, Coimbatore-641009, India

**Summary.** This paper describes an 8-year-old boy who presented with the very rare condition of congenital right-sided aglossia. The left side of tongue was hypoplastic and tethered to the floor of the mouth. The soft palate was short and almost absent on the right side with abnormal positioning of the right anterior and posterior tonsillar pillars; this situation resulted in significant velopharyngeal incompetence (VPI). The left mandible was slightly hypoplastic with anodontia posterior to the first premolar tooth. There has been no previous report of a similar defect. The possible differential diagnoses related to the abnormality are discussed. The correction of the VPI is presented to demonstrate the versatility of the sphincter pharyngoplasty.

**Key words:** Congenital hypoglossia – Sphincter pharyngoplasty – Velopharyngeal incompetence – Soft palate agenesis

Congenital anomalies of the tongue are uncommon and mainly consist of congenital aglossia and hypoglossia. They may occur as isolated situations or as part of the oromandibular limb hypogenesis syndrome (OLHS) [8]. Abnormalities, of the soft palate, anterior and posterior pillars are very rare.

The purpose of this paper is to present the basic deformity and a variation of the sphincter pharyngoplasty used to correct this patient's VPI. A review of the literature on tongue and palate malformations is also presented.

### Case history

A 8-year-old boy from Madras, India was referred for evaluation of severe tongue hypoplasia, soft palate deformity and velopharyn-

geal incompetence (Fig. 1). He was accompanied by his plastic surgeon who supplied information and reported on speech assessment performed in India. This was necessary because the child did not speak or understand English. On initial examination and later under anesthesia, he was found to have multiple intraoral deformities (Table 1). The soft palate was short and almost totally absent on the right side with abnormal positioning of the right anterior and posterior tonsillar pillars. The posterior pillar appeared not to be connected to the palate and the anterior pillar was bound to the cheek (Fig. 2). The uvula was cleft and the tonsils were enlarged bilaterally. When asked to speak, good movement was observed in the palate and the muscles around the velopharyngeal area using the flexible endoscope placed intraorally. The tongue was virtually absent on the right side and the remaining hypoplastic part of tongue on the left side was tethered to the floor of the mouth (Fig. 3). What movement was present appeared to come from the left side. The left mandible was slightly hypoplastic with anodontia posterior to the first premolar tooth (Fig. 4); the incisal opening was normal. There was no cranial nerve involvement, no limb or other skeletal abnormalities, and no family history of congenital anomalies. The birth history was also normal.

Nasendoscopy and videofluoroscopy were unsuccessful because of poor patient cooperation. Therefore, an examination under anesthesia was performed. The operative plan was to obtain improved tongue mobility and correction of the VPI (Fig. 5a–f).

### Operative technique

The band stretching from the tonsillar area to the buccal aspect of the right cheek was incised in a V-shape manner back to the right tuberosity of the maxilla in order to create a hemi-soft palate (Fig. 5b). The donor site was closed primarily with resorbable sutures. The posterior and lateral pharyngeal wall was inspected and palpated to determine the position of the great vessels of the head and neck. Once it was determined to be safe, an incision was made down the right lateral wall of the oral pharynx and across the posterior pharyngeal wall. The flap of the newly created soft palate was then sutured into this position (Fig. 5c). At this point, the uvula was sutured on to the left side of the soft palate for exposure and attention was turned to the left lateral pharyngeal wall. An incision was made laterally and anterior along the posterior tonsillar pillar and a dissection was made under the pillar moving posteriorly and medially (Fig. 5d). The superiorly based flap that was created by this dissection was sutured into the incision in the posterior pharyngeal wall, the medial edge superiorly and the lateral edge inferiorly. It was also sutured onto the newly created right

*Correspondence to:* I.T. Jackson, MD, Institute for Craniofacial and Reconstructive Surgery, Third Floor Fisher Center, 16001 West Nine Mile Road, Southfield, Michigan, USA