Microglossia in a Newborn: A Case Report and Review of the Literature
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INTRODUCTION
Microglossia is a very rare condition with approximately 50 cases reported in literature to date. Frequently, this disorder presents in association with limb abnormalities and is further grouped as a hypoglossia-hypodactilia syndrome. In 1718, de Jussieu was the first to describe the condition. He reported on a 15-year-old man with what he termed “congenital lingual hypoplasia”. In 1932 the literature up to that date was reviewed by Rosenthal and he also documented the first report of the hypoglossia-hypodactilia syndrome. Subsequent reports have been released which confirming a relationship between hypoglossia and hypodactilia [1,3]. In this poster we discuss a case of isolated microglossia, its workup, and management.

CASE PRESENTATION
Patient AR was born to a 21 year old G1P1 female via Cesarean section secondary to fetal distress and polyhydramnios at 40 4/7 weeks at an outside hospital. Otolaryngology consultation was performed to evaluate the patient’s craniofacial abnormalities. Weight at birth was 3770 grams and Apgars were 4, 7, and 8 at 1, 5, and 10 minutes respectively. The pregnancy was otherwise uneventful. He was transferred to Tufts Medical Center for further workup of his microglossia and retrognathia at day of life 1. Initially he was kept n.p.o. and received tube feeds via a nasogastric tube until his swallowing function could be fully assessed. On physical exam he was noted to have marked retrognathia and a high arched palate, without associated limb abnormalities (figure 2). His tongue deflected posteriorly with a small anterior two-thirds of the tongue but normal appearing posterior tongue. The posterior third of the tongue effaced the valleculae and epiglottis. On imaging he was noted to have a sabertooth appearance to his trachea along with evidence of supraglottic macaria on laryngoscopy (figure 3). After evaluation by the feeding and swallowing team, he was made n.p.o. due to risk for aspiration, which is a common finding in the small number of documented microglossia cases. After a modified barium swallow study and functional endoscopic evaluation of swallow which confirmed aspiration, he underwent placement of a gastrostomy tube for feeding on DOL 24. He was discharged home on DOL 31 on G tube feeds in good condition with close follow up every 1 to 2 months. Genetic workup did not reveal any syndromic causes of his craniofacial abnormalities. At five months of age, he passed a subsequent modified barium swallow and was allowed to start soft solids with supplemental G tube feeds. His parents have been referred to audiology for sign language classes as his future expressive language may be delayed due to his microglossia. Finally a craniofacial consultation was performed to discuss potential intervention for his mild micrognathia.

A conclusive etiology for microglossia remains at this time unknown however several theories have been postulated. Certain drugs used during pregnancy such as diazepam, chlorpromazine, meclizine and tigan have been thought to possibly contribute to the condition. Another proposed risk factor is hyperthermia in utero especially in association with Moebius syndrome. There is no substantial evidence for heredity or sexual preference in the development of microglossial [1,3]. Interference of the stapedial blood supply has also been postulated by some as a potential cause leading to malformation of the 2nd branchial arch [6]. Congenital abnormalities associated with some cases of microglossia or aglossia are: Hanhart Syndrome, Charles M. syndrome, Robin sequence & Moebius syndrome [3]. Several cases of microglossia have also been associated with situs inversus and dextrocardia [6].

On physical exam, various oral anomalies may be present in association with microglossia or aglossia. These patients tend to have a narrowing of the face referred by many as “bird-like” along with retrognathia, a high palatal vault and an excessive overbite [2]. Cleft palate has also been reported in a small subset of cases. Intraoral bands may be present between the mandible and maxilla involving soft tissue or bone. These attachments may require surgery due to limitation of oral excursion or feeding. Uvular enlargement is present in many cases of hypoglossia. A consistent finding later during development are dental anomalies, notably absence of both the molars and midarchis [3]. Several developmental issues need to be followed as the patient matures. The tongue is involved in speech, mastication, swallowing, and dental development [3]. These issues have to be addressed through infancy and childhood. The treatment team should involve a nutrition, psychology, speech & hearing, general dentistry, and orthodontics if necessary [5]. Follow up should continue through adolescence to correct possible speech impediments, deal with self-esteem issues from her facial appearance and dentistry to address occlusal issues.

REFERENCES