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Case report

Speech characteristics in a Ugandan child with a rare paramedian craniofacial cleft: A case report

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ABSTRACT

The purpose of this study is to describe the speech characteristics in an English-speaking Ugandan boy of 4.5 years who has a rare paramedian craniofacial cleft (unilateral lip, alveolar, palatal, nasal and maxillary cleft, and associated hypertelorism). Closure of the lip together with the closure of the hard and soft palate (one-stage palatal closure) was performed at the age of 5 months. Objective as well as subjective speech assessment techniques were used. The speech samples were perceptually judged for articulation, intelligibility and nasality. The Nasometer was used for the objective measurement of the nasalance values. The most striking communication problems in this child with the rare craniofacial cleft are an incomplete phonetic inventory, a severely impaired speech intelligibility with the presence of very severe hypernasality, mild nasal emission, phonetic disorders (omission of several consonants, decreased intraoral pressure in explosives, insufficient frication of fricatives and the use of a middorsum palatal stop) and phonological disorders (deletion of initial and final consonants and consonant clusters). The increased objective nasalance values are in agreement with the presence of the audible nasality disorders. The results revealed that several phonetic and phonological articulation disorders together with a decreased speech intelligibility and resonance disorders are present in the child with a rare craniofacial cleft. To what extent a secondary surgery for velopharyngeal insufficiency, combined with speech therapy, will improve speech intelligibility, articulation and resonance characteristics is a subject for further research. The results of such analyses may ultimately serve as a starting point for specific surgical and logopedic treatment that addresses the specific needs of children with rare facial clefts.

1. Introduction

The overall incidence of clefts among newborn babies in Uganda over a period of 1 year is 0.73 of 1000 live births [1,2]. A database of 1304 patients (median age: 2.8 years, range: 1 month to 60 years) revealed a ratio of 37 for cleft lip, a ratio of 12 for cleft lip and palate and a ratio of 1 for cleft palate [3]. More severe facial clefting also presented at the cleft clinic of CoRSU, with patients coming from a wide geographic area: from as far as Sudan, DR Congo, and Rwanda. The typical classification system used to describe standard clefts of the lip and palate is based on careful anatomic description. More severe facial clefting is most commonly described using Tessier's orbitocentric system of numbering [4], a classification system that is very useful in routine clinical practice. The classification of Tessier assigns numbers to various sites of clefting, depending on their relationship to the sagittal midline [5]. According to Gorlin et al. [6], craniofacial clefting associated with frontonasal malformations was first described by Hoppe [7] in 1859.

An aspect of the clinical spectrum of craniofacial clefting associated with frontonasal malformations that has received less attention is that of the presence of possible speech problems. Very few authors reported detailed analysis of the articulation and resonance in children with this rare type of clefting. As Shprintzen [8] pointed out, recognition of the spectrum, natural history and prognosis of a genetic disorder is critical for proper patient care. The present study aims to perform a detailed analysis of the speech characteristics (overall speech intelligibility, resonance and articulation) in an English-speaking Ugandan boy of 4.5 years

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with a lip, palatal, nasal and maxillary cleft and associated hypertelorism (no. 2 of the classification of Tessier). Detailed information may lead to the better guidance of pediatric management programs, especially in poor countries (like Uganda) where very few specialized expertise clinics in cleft palate treatment and very few speech language pathologists exist. Also, knowledge of the speech characteristics in children with rare clefts in developed countries can provide useful information (especially regarding prognosis) for reconstructive surgeons and speech language pathologists, and may lead to more adequate tailor-made protocols. According to a study by van den Elzen et al. [9], long-term results are often still far from ideal and could be improved, especially in regards to esthetics and speech. To the best of our knowledge, no studies were found regarding speech characteristics in children with this rare craniofacial cleft.

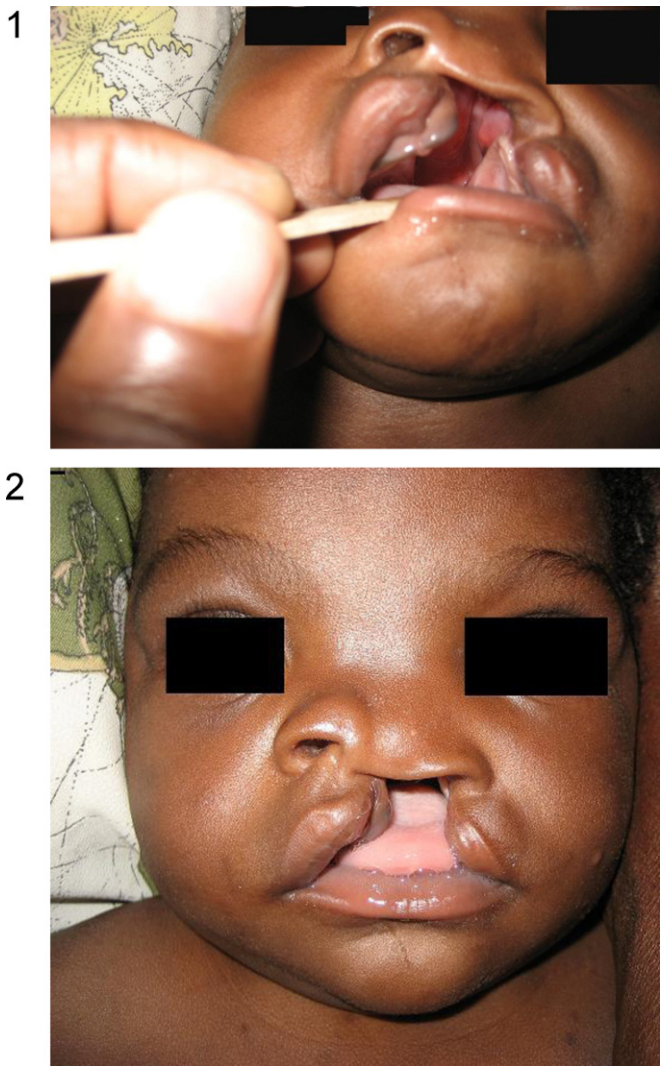
2. Methods and materials

This research was approved by the local institutional review and the ethical board (B67020096013) and an informed consent was signed by the parents.

2.1. Case history

2.1.1. Child with the rare craniofacial cleft

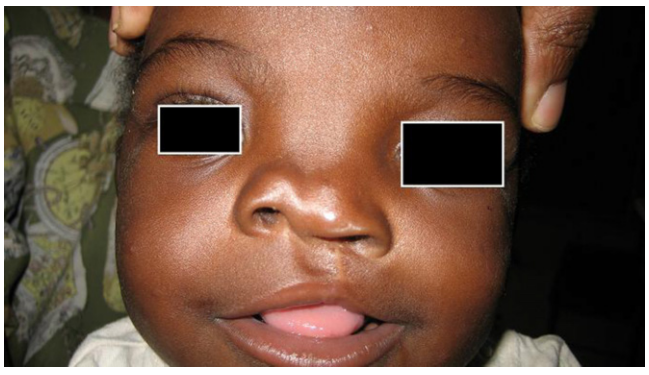
G., a Ugandan boy, is the first child of healthy non-consanguineous parents and was born at 37 weeks of gestation after an uncomplicated pregnancy and delivery. The family history is uneventful. Clinical evaluation of the newborn at the age of 1 month revealed a left cleft lip, alveolus and palate, a nasal cleft, and hypertelorism (Pictures 1 and 2). According to Tessier's [5] classification, this is a no. 2 cleft. Breastfeeding was not possible due to severe sucking problems. The child was fed with a spoon but nasal regurgitation was still present. Due to the presence of the left cleft lip and palate (CLP) and the nasal cleft, the child and the mother were abandoned by the community because – according to traditional Ugandan beliefs – the mother and child were cursed. Closure of the lip, together with the closure of the hard and soft palate (one-stage palatal closure), was performed at the age of 5 months (Picture 3). Postsurgical complications were not present. No alveolar bone grafting or surgical corrections of the hypertelorism or nose were performed. At the age of 4.5 years, G. (Picture 4) consulted the cleft palate center together with his grandmother because of decreased speech intelligibility, especially for strangers, in both Luganda (mother tongue and an official language in Uganda) and English. G. was attending a regular school and he was able to produce and comprehend the English language. Normal mental abilities, normal language, normal motor development and normal hearing were present, according to the grandmother and clinical observations. Open mouth behavior with mouth breathing and tongue sucking was present. A CT scan at the age of 4.5 years (Picture 5) revealed a bone defect extending through the nasal bone, hard palate and maxilla bone on the left side of the face with the presence of a slight hypertelorism. Consensus evaluation (AH, KVL) of the velopharyngeal mechanism (intraoral observation) during the production of the /a/ revealed a normal morphology of the soft palate, with a lifting of the soft palate and a medial constriction of the pharyngeal walls. Muscle activity of the posterior pharyngeal wall could not be observed. Equipment to perform nasopharyngoscopic and multi-view videofluoroscopic evaluation of the velopharyngeal mechanism during speech was not available in this poor Ugandan country. G. was assessed for articulation and resonance at the cleft palate center. Since very few speech language pathologists are available in Uganda, no speech therapy was provided.



Pictures 1 and 2. Clinical evaluation of the newborn at the age of 1 month revealed a left cleft lip, alveolus and palate, a nasal cleft and hypertelorism.

2.1.2. Surgical technique

The child with this rare craniofacial cleft consulted the cleft palate center in Uganda and was operated on by an experienced surgeon (AH), who performed a lip closure and one-stage palatal closure (soft and hard palate) at the age of 5 months. A Modified Millard repair and a Modified Mulliken technique were applied to



Picture 3. Closure of the lip together with the closure of the hard and soft palate (one stage palatal closure) was performed at the age of 5 months.