

Case Report

Relato de Caso

Laryngeal cleft type I in neonate: case report

Cleft laríngeo tipo I em neonato: relato de caso

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ABSTRACT

Laryngeal cleft (LC) is a congenital malformation that leads to the unusual communication between the esophagus and the laryngotracheal complex. It is a rare disease, mostly prevalent among male individuals. The goal of this study was to describe the evaluation and intervention by the speech language pathologist of a female newborn diagnosed with LC type I, admitted on the University Hospital of Universidade de São Paulo, in her second hospitalization due to small weight gain and pneumonia. She was submitted to a bedside clinical evaluation of the swallowing and the most important occurrence was frequent gagging. The videofluoroscopy swallowing study showed laryngotracheal aspiration level 8 for thin liquid and level 1 for thickened liquid, according to the Penetration-Aspiration Scale. The newborn was submitted to a microlaryngoscopy, in which the presence of LC type I was found. After the diagnosis, the speech language pathologist offered thickened liquid at 6% and, in 8 days, the newborn was discharged with exclusive oral diet without gagging. Eight outpatient consultations were carried out for 11 months, with emphasis on reintroduction of thin liquids. The treatment was discontinued and the patient was put on general diet for the age without modifications. Throughout follow-up, the patient remained asymptomatic and showed no respiratory complications.

RESUMO

O *cleft* laríngeo (CL) é uma malformação congênita que resulta em uma incomum comunicação entre esôfago e o complexo laringotraqueal. É uma doença rara, de maior prevalência no gênero masculino. O objetivo deste estudo foi relatar a atuação fonoaudiológica em um caso de um paciente neonato com diagnóstico de CL tipo I, admitido no Berçário do Hospital Universitário da Universidade de São Paulo, em sua segunda internação por baixo ganho ponderal e quadro de pneumonia. Foi realizada a avaliação clínica da disfagia infantil em beira de leito, cuja principal ocorrência foi a presença de engasgos frequentes. O exame de videofluoroscopia da deglutição evidenciou presença de aspiração laringotraqueal escore 8 para líquido fino e escore 1 para líquido engrossado, segundo a Escala de Penetração-Aspiração. O neonato foi submetido a microlaringoscopia, na qual foi constatada a presença de CL tipo I. Após o diagnóstico, a conduta fonoaudiológica foi de engrossar a fórmula láctea a 6% e, em oito dias, o neonato recebeu alta com dieta exclusiva por via oral, com ausência de engasgos. Foram realizadas oito consultas ambulatoriais durante 11 meses de acompanhamento, com ênfase na reintrodução de líquidos finos. A paciente recebeu alta fonoaudiológica com dieta geral para a idade, sem modificações. Durante todo o gerenciamento, a paciente permaneceu assintomática e não apresentou quadros respiratórios.

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INTRODUCTION

Laryngeal cleft (LC), by definition, is a congenital malformation that results in the unusual communication between the esophagus and the laryngotracheal complex. It is a rare disease, and it affects 1:10,000/20,000 live births, mostly among the male gender⁽¹⁻⁹⁾.

LC is usually associated with other malformations (16–88%)^(1,4,6-8), especially in the digestive tract⁽⁷⁾, which can be presented alone or resulting from genetic syndrome. Four genetic syndromes are frequently associated with LC: Opitz–Friaiz syndrome and Pallister-Hall syndrome^(1,3,6-8), DiGeorge syndrome⁽⁸⁾, and CHARGE syndrome^(6,8), besides the VACTERL association^(1,6,8).

The extension level of this anatomical impairment varies, and it starts with a small opening considered to be an anatomic variant, then becomes a full communication between the trachea and the esophagus⁽⁷⁾. The most accepted classification system is proposed by Benjamin and Inglis, modified by Sandu, in 2006^(1,3,5-8). Five types are presented, from 0 to IV, ranging from submucous LC to the one that extends toward the thoracic trachea. Symptomatology is varied and unspecific. Literature describes the most common symptoms for each type, and severity increases according to cleft extension, which has a direct implication on the prognosis^(1,6-8).

Generally, usual symptoms are choking events, coughing, cyanosis, episodes of aspiration, respiratory stridor, x-ray thoracic images with changes suggestive of bronchoaspiration, or pneumonia^(1,2,4,6-8). Some of these symptoms are usually taken for sucking, swallowing, and breathing incoordination, especially when it comes to newborns. The diagnosis becomes difficult and, sometimes, late^(2,6).

The LC diagnosis is conducted by imaging examinations and endoscopy^(1-3,5,7,8). With these tests, it is possible to verify the existence of other malformations, such as fistulae, laryngomalacia, laryngeal paralysis, gastroesophageal reflux disease, and changes in deglutition^(1,2,5,6,8). The gold standard examination is direct microlaryngoscopy^(1,3,6,7).

Medical evaluation considers not only the extension of LC but also the severity of symptoms and their frequency, as well as complications caused to the respiratory tract. Only after that, the most indicated therapy is chosen. In minor LCs, the first indication is the nonsurgical treatment, by conservative therapy. In the other cases, there is surgical indication, and the approach can be diverse^(1,2,5-8).

Because of the unspecific symptomatology, the course with other diseases, and the presence of signs indicating changes in deglutition, literature points out to the importance of differential diagnosis^(1,2,6,7), which should also involve professionals from correlated fields, including a speech language pathologist^(2,7).

The objective of this study was to report the work of the speech language pathologist in the case of a newborn diagnosed with LC type I.

CLINICAL CASE PRESENTATION

The project was approved by the research ethics committee of the University Hospital at Universidade de São Paulo (USP) — registration CEP 830/08 — and all of the criteria from Resolution 196/96, from the National Health Council, were fulfilled. People who were legally in charge of the participant signed the informed consent.

This is the case of a female newborn, born at term, adequate for gestational age. Cesarean section was performed due to suspicion of fetal suffering. The mother was 29 years old and it was her second gestation, with 8/9/10 Apgar, 3,250 g, presence of meconium fluid (++/4+), and need for upper airway aspiration. After 24 hours, the newborn had fever and respiratory discomfort. During the first hospitalization, she presented with sepsis and needed volemic expansion; she underwent oxygen therapy for 6 days. After 10 days in the hospital, she improved and was discharged, exclusively on breast-feeding.

Eighteen days after this first hospitalization, the newborn was admitted to the nursery in the same hospital, presenting with small weight gain, complaints of choking and coughing while breast-feeding, leading to crisis of cyanosis. Right thoracic x-rays and tomography were conducted, which showed bronchopneumonia. A speech language pathology and audiology evaluation was requested because these symptoms persisted during breast-feeding.

The evaluation conducted by the speech language pathologist did not show orofacial structural changes, and the frequency of saliva swallowing was adequate. During non-nutritive suction, the following were observed: adequate suction, satisfactory rhythm, and adequate intraoral pressure. The newborn did not present changes in physiological parameters or in cervical auscultation.

In the orthodontic bottle test, the patient had effective suction, adequate bottle nipple grip, 1:1 suction/deglutition frequency, and absence of spontaneous pauses. However, she choked twice, which led the speech language team to suspect changes in the pharyngeal swallowing phase and to discuss the request of a swallowing videofluoroscopy from the medical team. The speech language team also oriented the mother and the nursing staff to make pauses after three suction to avoid choking events.

The test was conducted in the hospital, on March 27, 2012, and was in accordance with the service protocol. The newborn was placed in raised decubitus with a compression bandage, which allowed safe retention and lateral view, being as close as possible to the tabletop and intensifier. Therefore, it was possible to avoid distortions in the fluoroscopic image. The area of interest was observed, with exposure parameters that were automatically defined by the ionization chamber, where it is possible to see the best fluoroscopic image with the lowest radiation dose. Contrast was administered by an orthodontic nipple from NUK®, with hole for thin liquid, and the dilution of 30% of barium sulfate at 100% (Bariogel®) was used, as well as 70% of warm milk (thin liquid), in the total volume prescribed

by the neonatal physician. The focus of the fluoroscopic image was limited by the lips, in the anterior area, and by the nasal cavity, in the upper area; by the spine, in the posterior area, and by the bifurcation of the airway and the cervical esophagus in the lower area.

Swallowing videofluoroscopy

In the oral swallowing phase, the newborn presented with adequate lip competence and tongue movement, as well as efficient suction and 1:1 suction/deglutition frequency. In the laryngeal phase, she did not have contrast in the rhinopharynx, and it was possible to observe decreasing elevation and anteriorization of the hyolaryngeal complex (qualitative observation). There were three episodes of laryngeal penetration and two episodes of silent laryngeal aspiration. Afterward, she was offered thickened milk (2%) in an orthodontic bottle, with a hole for thin liquids, and she performed well, without penetration and/or aspiration. The images led to the diagnosis of discrete/moderate dysphagia, according to the Dysphagia Severity Rating Scale⁽¹⁰⁾, and, according to the Penetration-Aspiration Scale^(10,11), she got score 8 for thin liquid and score 1 for thickened liquid. It is worth mentioning that there are no specific scales for swallowing videofluoroscopy in the infant population, therefore two scales addressed to classifying dysphagia among adults were used⁽¹²⁾. In the esophageal phase, images indicated the presence of esophageal reflux up to the cervical portion of the esophagus.

On the basis of these data, and considering the fact that the mother's milk production had reduced, the multiprofessional team chose to use milk formula for terms, thickened in 2% with rice cream. Therefore, it was possible to observe fewer choking events and oxygen saturation falls, besides more weight gain.

With the objective of clarifying the diagnosis, a direct microlaryngoscopy was conducted on March 30, 2012, which showed the presence of LC type I (arytenoid cleft), and larynx had normal morphology, except for the absence of the posterior interarytenoid fold, presence of precommissure, and presence of milk in the airways. The medical team considered there was no indication for surgery, therefore the conservative therapy was adopted. The multiprofessional team chose to thicken the milk in 6% with rice cream.

Eight days after the direct microlaryngoscopy and changes in the diet, the newborn was discharged from the hospital and got better, presenting with stable respiratory pattern, and absence of chokes and oxygen desaturation.

After hospital discharge, the patient was referred to the Speech Language Pathology and Audiology Outpatient clinic in the same hospital, and attended eight appointments in 11 months. During this treatment, the patient performed well with thickened milk at 6%, with rice cream. She underwent therapy to restart with thin liquids (water and juice), beginning with a spoon, then to the bottle with reduced flow. She did not have clinical signs suggestive of penetration and/or aspiration with thin liquids at the end of therapy. Besides, pasty foods were introduced with a spoon. The patient showed good performance and absence of clinical signs suggesting penetration and/or aspiration, thus progressing to a general diet for the age,

without using thickeners after discontinuation of the treatment. During the whole period of speech language pathology and audiology, as well as pediatric follow-up, the patient remained asymptomatic and did not present respiratory problems.

DISCUSSION

LC is a low incidence congenital structural anomaly, mostly prevalent among male individuals⁽¹⁻⁹⁾. This study is in accordance with literature, because it presents the case of a newborn diagnosed with congenital malformation; however, it is different from the references because this is the case of a female newborn.

Even though the presence of LC is strongly associated with other malformations or genetic syndromes^(1,4,6-8), the patient in this study did not have any additional diagnoses.

The symptoms of this disease are unspecific, therefore diagnosis can be difficult^(2,3,8). The symptomatology presented in the reported case is in accordance with findings in literature referring to the presence of LC type I, that is, crises of cyanosis and changes in deglutition^(1,2,6-8). In the first medical and speech language clinical evaluations, these symptoms were attributed to swallowing incoordination presented during breast-feeding⁽⁶⁻⁸⁾. However, the newborn was at term, hygid, and there was no justification as to the patient's maturation for such incoordination; that is why the hypothesis of changes in the pharyngeal swallowing phase came up. These observations are corroborated by the fact that the mentioned studies indicate the following common symptoms in oropharyngeal dysphagia: O₂ saturation falls while breast-feeding, choking events, coughing, changes in cervical auscultation, and crises of cyanosis, which justifies the speech language intervention^(5,9,13).

The presence of changes in deglutition leads to increasing risks for pulmonary chronic diseases, for the children, as well as changes regarding nutrition, neuromotor development, besides stressful interactions with caretakers. It is also important to consider the delay and the interference in typical patterns of eating/swallowing development, which can lead to food refusal^(5,9,13).

In this study, LC caused pulmonar complications (pneumonia), as described in literature^(1,2,4). The early cleft diagnosis minimized morbidity and mortality rates^(3,6-8), and speech language intervention aiming at adapting the deglutition of the newborn^(5,9,13) was essential to guide the diagnosis, once imaging examinations were performed after the case was discussed with the medical team.

Studies^(1-4,6-8) indicate that LCs are diagnosed and classified by imaging examinations, once the unspecific symptomatology of these changes masks the base illness. They are also important for the choice of safe therapy, which, in the case of the aforementioned newborn, was directly related with speech language intervention^(1,2). The use of imaging examinations was also essential to elucidate the family with regard to the proposal to change diets, and, to create more adherence to treatment.

The treatment elected by the speech language team was to thicken the diet of the newborn with the objective of preventing new aspiration episodes. Literature^(2,3,8) mentions thickened foods as a strategy to prevent aspiration in smaller clefts, such

as type I. This strategy is part of the conservative treatment and does not involve surgical interventions. Besides, the use of anti-reflux medicines and positioning techniques during meals are also part of this treatment^(1,7,14,15). The adopted intervention was efficient, because during speech language pathology and pediatric follow-up, the patient remained asymptomatic and did not present with respiratory problems.

CONCLUSION

The presence of a speech language pathologist in the neonatal staff is reinforced by the presented case, once his active participation during the intervention process with the patient allowed the early diagnosis by the speech language pathology clinical evaluation and by the participation in the indication of a gold standard examination for the accurate diagnosis of deglutition (swallowing videofluoroscopy) and the indication to change diets, with the objective of preventing aspirations and pulmonary complications. It is worth to mention the work of the speech language team with regard to elucidation and adherence of family members to the intervention. When speech language pathologists are part of a neonatal clinical staff, they play an important role concerning the factor time of hospitalization, thus reducing that period, both in the quality of life of the babies and family members, because they work with orientations and outpatient clinic follow-up, when necessary, aiming at preventing new hospitalizations.

**MSR and KEBB performed the speech language and audiology follow-up of the case; GABV and JPO provided medical care and the objective evaluation of the structures involved in deglutition; SCOL oriented the paper; all of the authors actively participated in the discussion and in the preparation of the manuscript.*

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