Case Report

Relato de Caso

Intensive treatment of speech disorders in robin sequence: a case report

Maria Daniela Borro Pinto[1]
Maria Inês Pegoraro-Krook[1]
Laura Katarine Félix de Andrade[1]
Ana Paula Carvalho Correa[1]
Linda Iris Rosa-Lugo[2]
Jeniffer de Cássia Rillo Dutka[1]

Keywords

Pierre Robin Sequence
Speech
Velopharyngeal Dysfunction
Cleft Palate
Speech Bulb
Speech Therapy

ABSTRACT

Purpose: To describe the speech of a patient with Pierre Robin Sequence (PRS) and severe speech disorders before and after participating in an Intensive Speech Therapy Program (ISTP). **Methods:** The ISTP consisted of two daily sessions of therapy over a 36-week period, resulting in a total of 360 therapy sessions. The sessions included the phases of establishment, generalization, and maintenance. A combination of strategies, such as modified contrast therapy and speech sound perception training, were used to elicit adequate place of articulation. The ISTP addressed correction of place of production of oral consonants and maximization of movement of the pharyngeal walls with a speech bulb reduction program. Therapy targets were addressed at the phonetic level with a gradual increase in the complexity of the productions hierarchically (e.g., syllables, words, phrases, conversation) while simultaneously addressing the velopharyngeal hypodynamism with speech bulb reductions. **Results:** Re-evaluation after the ISTP revealed normal speech resonance and articulation with the speech bulb. Nasoendoscopic assessment indicated consistent velopharyngeal closure for all oral sounds with the speech bulb in place. **Conclusion:** Intensive speech therapy, combined with the use of the speech bulb, yielded positive outcomes in the rehabilitation of a clinical case with severe speech disorders associated with velopharyngeal dysfunction in Pierre Robin Sequence.

Correspondence address:
Jeniffer de Cássia Rillo Dutka
Hospital for Rehabilitation of Craniofacial Anomalies, Universidade de São Paulo – USP
Rua Silvio Marchione 3-20, Vila Universitária, Bauru (SP), Brazil, CEP: 17012-900.
E-mail: jdutka@usp.br

Received: April 29, 2016

Accepted: April 30, 2017

Study conducted at the Hospital for Rehabilitation of Craniofacial Anomalies, Universidade de São Paulo – USP - Bauru (SP), Brazil, in partnership with the Bauru School of Dentistry at the Universidade de São Paulo Paulo – USP - Bauru (SP), Brazil, and the Department of Communication Sciences and Disorders at the University of Central Florida – UCF - Orlando (FL), USA.

[1] Universidade de São Paulo – USP - Bauru (SP), Brazil.

[2] University of Central Florida – UFC - Orlando (FL), USA.

Financial support: nothing to declare.

Conflict of interests: nothing to declare.

INTRODUCTION

Pierre Robin Sequence (PRS) is characterized by the presence of micrognathia, glossoptosis, and cleft palate occurring in isolation or associated with other syndrome. Problems often associated with PRS include airway obstruction and feeding difficulties, which are more severe in the neonatal period. The manifestation of these difficulties in children occurs heterogeneously and may cause severe asphyxia, which can lead to death if not promptly treated[1]. Cleft palate can be present in 90% of PRS cases, with 70% involving extensive, complete U-shaped clefts and 30% involving narrow, V-shaped clefts[1]. Since the presence of a cleft palate can have a negative impact on speech, primary palatoplasty is recommended as soon as possible and ideally should be conducted within the first year of life[1]. The presence of airway obstruction and feeding difficulty, however, can have an impact on the timing of surgery, considering that normal breathing and feeding are required prior to surgery[1]. According to Prado et al.[2], hypernasality after primary palatoplasty in patients with PRS can vary between 26% and 53% depending on the surgical technique used. Velopharyngeal dysfunction (VPD) can involve a structural etiology, such as insufficient tissue, and a functional etiology, such as velopharyngeal hypodynamism[3].

Correction of velopharyngeal insufficiency requires physical treatment (surgery or a pharyngeal obturator), whereas correction of velopharyngeal hypodynamism (VPH) can only be accomplished with behavioral treatment (speech therapy). Velopharyngeal insufficiency and hypodynamism critically impact middle ear functioning and speech production, both resulting in a disorder known as *cleft palate speech*, which is characterized by the presence of hypernasality, audible nasal air emission, weak intra-oral pressure, and use of atypical place of articulation (compensatory articulation). Whenever the disorder involves errors of speech sound production and/or velopharyngeal mislearning (particularly velopharyngeal hypodynamism), speech therapy is the only option to establish intelligible speech.

For children presenting velopharyngeal dysfunction after palatal repair, the timing of primary palatoplasty may be delayed considering that normal breathing and feeding must be established prior to surgery[1]. The longer the delay in establishing a functional velopharyngeal mechanism, the higher the risk for development of velopharyngeal hypodynamism, that is, because speech development requires control of different amounts of air pressure for adequate sound production, coupling of the oral and nasal cavities associated with backing of the tongue predispose children with PRS to produce speech using compensatory mechanisms, leading to velopharyngeal mislearning for speech. Behavioral management of velopharyngeal mislearning for speech in this population requires adequate, specialized, intensive speech therapy. Golding-Kushner[4], suggests that the success of speech therapy is directly associated with the frequency at which the activities proposed during therapy are adequately practiced. The literature addressing speech therapy interventions for children with cleft palate is limited, particularly regarding information related to intensive treatment[5-13]. Lima et al.[9], Pamplona et al.[8], and Pinto[12] reported treatment outcome that supports the importance of conducting intensive speech therapy to resolve speech disorders associated with cleft lip and palate (CLP). Another study reported the findings for a 16-year-old patient with VPD treated with speech bulb associated with a 20-week-long speech therapy program, and showed adequacy of speech nasality previously classified as hypernasal[14]. More recently, a doctoral dissertation[12] reported speech outcome for 67 patients with history of cleft palate before and after an intensive speech therapy program (ISTP) to correct the use of glottal stop (GS) and pharyngeal fricative (PF). After the ISTP, adequate oral place of articulation for all targets was observed in 39% of the patients, whereas reduced use of GS and PF was noted in 48% of the patients, thereby revealing improved articulatory production for most of the patients. Only 13% of the patients in the ISTP maintained the same speech errors they presented prior to participating in the therapy. These findings[12] suggest a correlation between correction or improvement of articulatory production and ISTP. Nonetheless, the optimal duration and intensity of therapy to correct speech disorders associated with CLP still need further investigation.

The objective of this case report is to describe the unique features of the intensive therapeutic approach implemented for management of the communication disorders associated with velopharyngeal dysfunction in a patient with PRS and history of cleft palate. The efficacy of the intensive speech therapy program, however, could not be established in this single subject study. More specifically, a case control was not possible for comparison and, because the data were retrieved retrospectively, the control of variables such as exact duration of each therapy and the specific strategies implemented by the mother during home practice were not registered. Therefore, this case report characterizes an uncontrolled descriptive report of events and outcomes in a single case (ASHA, 2017a)[15] and does not propose to test a hypothesis.

CASE REPORT

This case report was approved by the Ethics Committee of Hospital de Reabilitação de Anomalias Craniofaciais at the Universidade de São Paulo (HRAC-USP) (Protocol: CAAE-35213414.6.0000.5441) and has been written as an individual study describing the intervention approach used in the ISTP implemented at the Palatal Prosthesis Services (PPS) for management of severe speech disorders associated with velopharyngeal hypodynamism. Individual studies are important sources of evidence that can guide treatment decisions, particularly when clinical practice guidelines or systematic reviews are not available (ASHA, 2017b)[16]. Therefore, the objective of this case report was to describe the management of speech disorders associated with CLP and velopharyngeal hypodynamism for a patient with PRS submitted to an intensive speech therapy program involving the use of speech bulb for management of velopharyngeal dysfunction.

Measures of speech nasality and articulatory production, as established by a speech language-pathologist (SLP), and identification of displacement of the velopharyngeal structures (rated by a single SLP during nasoendoscopy) were used to compare speech outcome before and after treatment in this case report. The patient studied presented incomplete cleft palate,

micrognathia and glossoptosis and was diagnosed with PRS. According to the information verified in the patient's medical records, the primary palatoplasty had to be postponed until the age of three years and 11 months because of respiratory and feeding difficulties associated with PRS. Between three and six years of age, the patient underwent periodic speech and hearing clinical evaluations at the institution where she received surgical treatment. Review of the medical records indicated that between three and six years of age, the patient's speech was already characterized by the use of atypical place of production (glottal stop), weak intraoral air pressure, and moderate hypernasality. The patient was referred for speech therapy to be conducted in her hometown with the primary goal of managing the articulatory errors; however, according to the mother, the patient did not receive speech therapy because of shortage of speech-language pathologists in their hometown.

The patient was continuously monitored by an otolaryngologist because of continuous complaints of recurrent ear infections. At the age of four, pressure equalizing tubes were placed in both ears. The diagnosis of VPD with both insufficiency and learning etiologies was established when the patient was 7 years old. At that time, the use of a speech bulb combined with speech therapy were recommended to correct the insufficiency, velopharyngeal mislearning, and hypodynamism.

At the age of eight, the patient presented persistent hearing complaints and a tympanoplasty was conducted in the right ear. At the age of nine, a tympanoplasty was conducted in the left ear. Audiological evaluations revealed moderate mixed hearing loss bilaterally. This diagnosis led to a recommendation for the use of an Individual Sound Amplification Device (AASI), bilaterally.

The mother also reported difficulties in school performance. These difficulties were often due to the patient's recurring ear infections and need to attend hospital appointments for treatment, causing absences from school. The patient also experienced difficulties interacting with other children because of reduced speech intelligibility and hearing loss.

At 10 years and eight months of age, the patient returned for an evaluation at the hospital to determine her progress in speech development. Even with an installed speech bulb the patient continued to demonstrate moderate hypernasality and replacement of all oral plosive, fricative and affricate sounds with glottal stops. Nasoendoscopic evaluation revealed absence of movement of the pharyngeal walls and reduced movement of the soft palate, with a large velopharyngeal gap, featuring hypodynamic velopharynx along with consistently posteriorized tongue positioning[4,17].

Owing to the lack of speech therapy in the patient's hometown, the family agreed to participate in an Intensive Speech Therapy Program (ISTP) at the PPS of HRAC-USP. The family was able to obtain resources to continue the treatment in the city of Bauru (away from their hometown) from funding provided by Healthcare Services. Before the ISTP, a speech assessment was conducted by a single SLP, not involved in the treatment and experienced in the diagnosis and treatment of speech disorders associated with VPD. As reported by the SLP, the patient presented with a speech disorder characterized by moderate hypernasality and replacement of the oral sounds /k/, /s/, /z/, /ʃ/, /ʒ/, /ʦ/, /ʤ/, /p/, /b/, /t/, /d/, /g/, /f/, and /v/ with glottal stops, as well as distortion of the sounds /l/, /r/, /ʎ/. Prior to starting the ISTP, a new speech bulb was installed to establish structural conditions for velopharyngeal sufficiency and also to be used as a strategy to foster movement of the velum and pharyngeal walls. At the age of 11 the patient returned to school while attending the ISTP.

A total of 360 sessions of therapy were conducted by two speech therapists under the supervision of a senior SLP experienced in the treatment of velopharyngeal hypodynamism. The program consisted of two daily 30-minute sessions over a 9-month period, resulting in a total of 36 weeks of treatment. The initial goals of the ISTP were to provide this patient with three months of intensive treatment consisting of frequent revaluations and to prepare the child to continue therapy at her hometown. However, the family chose to live near the craniofacial center to continue the intensive therapy program, resulting in a total of nine months of intervention.

The focus of therapy was to facilitate oral place of articulation and promote velopharyngeal closure for speech in order to correct speech nasality. As proposed in most approaches for treatment of speech sound disorders, therapy began at the establishment phase and progressed to the generalization and maintenance phases. Therapeutic approaches similar to the one used in this ISTP have been described in the literature[6,8,9,11]. Golding-Kushner[4,11], particularly, describes strategies for correction of glottal stops involving the target approximation model for controlling articulatory production to elicit adequate displacement of the velopharyngeal function. Some authors also contributed suggestions regarding therapy intensity, frequency, target selection, and use of facilitators to elicit adequate articulatory production[4-14]. The utilization of strategies for correcting the use of post uvular place of articulation, such as GS and PF, combined with a bulb reduction program to correct velopharyngeal hypodynamism are still not well described in the literature[17].

The approach involved eliciting the adequate oral place of articulation for all sounds while focusing on the elements lacking or overproduced in the patient's repertoire, starting at the phonetic level and increasing the complexity of the productions hierarchically (e.g., syllables, words, phrases/sentences, stories, conversation). Speech sound perception training was used to stabilize the *new speech pattern* while contrasting it with the *old speech pattern.* Facilitating cues (auditory, visual, and tactile) were used to promote self-monitoring of speech and self-correction of errors. Two SLPs alternated implementing the daily therapies under the guidance of a senior SLP experienced in behavioral treatment of velopharyngeal hypodynamism and correction of atypical place of articulation. The mother was prepared to reinforce the practice of well-established speech targets.

To address the velopharyngeal hypodynamism, a modified contrast therapy approach was used to develop awareness of oral and nasal levels of sound pressure, as well as the presence and absence of nasal air emission during each level of speech production. Initially the targets were practiced with and without the speech bulb (bulb 1) in place. Once nasal air escape was eliminated with the original speech bulb (bulb 1), a slightly

smaller bulb (bulb 2) was used with the modified contrast therapy approach. The contrast was implemented alternating between the larger (bulb 1) and the smaller (bulb 2) speech bulbs, until all oral speech sounds were produced with adequate place of articulation and without nasal air emission while using the smaller bulb (bulb 2).

Considering the history of micrognathism and glossoptosis, the risk for unheard backing of the tongue and use of post-uvular place of articulation were carefully monitored by nasoendoscopic evaluation of the velopharyngeal function for speech. Nasoendoscopy was used to monitor velopharyngeal closure (with the speech bulb in place) and as a biofeedback to provide the patient with awareness of displacement of the velum and pharyngeal walls during guided speech productions (Figure 1). Once velopharyngeal closure against the smaller speech bulb (bulb 2) was observed, a slightly smaller bulb (bulb 3) was introduced and the contrast was again implemented alternating between bulb 2 and bulb 3 until oral sounds were produced without nasal air emission using the smallest bulb (bulb 3). The process was repeated once again with bulb 4 (smaller than bulb 3) and bulb 5 (smaller than bulb 4). The patient reached maximum displacement of the pharyngeal walls without nasal air emission and with adequate oral place of production with bulb 5, that is, during the 36 weeks of intensive treatment a total of four bulb reductions were implemented in this patient.

The outcome of the ISTP was monitored with auditory-perceptual evaluation and nasoendoscopic assessment of the velopharyngeal function during speech production. The same speech therapist who conducted the auditory-perceptual assessment before the ISTP conducted the evaluation at the 9-month daily intervention. The findings revealed normal resonance and adequate oral articulation of speech sounds. The nasoendoscopic evaluation of the velopharyngeal function, after the ISTP, revealed consistent velopharyngeal closure during production of oral sounds with the smallest size speech bulb in place.

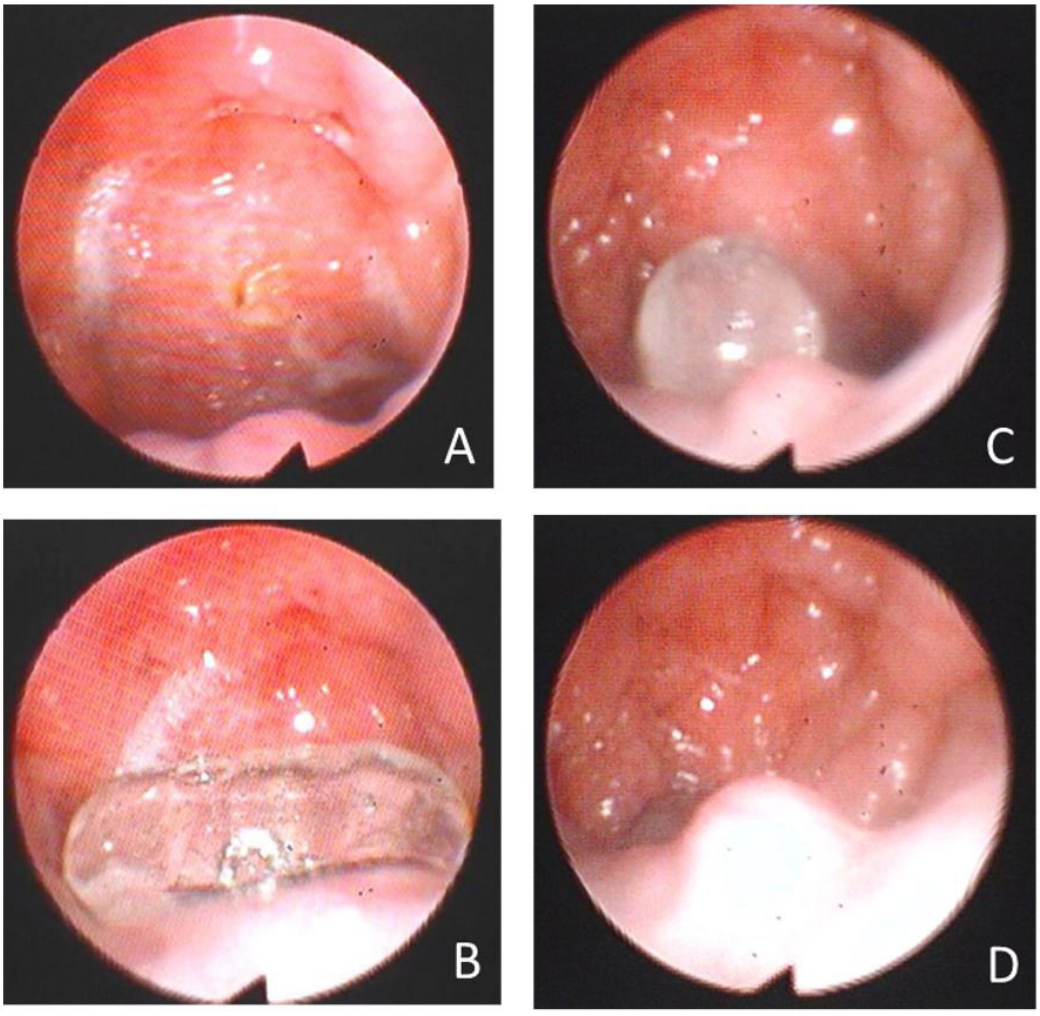

Figure 1. Superior view of the velopharynx during the nasoendoscopic evaluation. (A) Velopharyngeal mechanism at REST without speech bulb; (B) Velopharyngeal mechanism at REST with BULB 1 (largest) BEFORE the Intensive Speech Therapy Program (REST-BEFORE ISTP); (C) Velopharyngeal mechanism at REST with BULB 5 (smallest) AFTER the Intensive Speech Therapy Program (REST-AFTER ISTP); (D) Velopharyngeal mechanism during speech indicating CLOSURE with BULB 5 (smallest) at the end of the Intensive Speech Therapy Program (SPEECH-AFTER ISTP)

DISCUSSION

A critical component of speech development in individuals with cleft lip and palate is the age at which a primary palatoplasty is performed. In this case study, the surgery to correct the cleft was delayed because of the respiratory difficulties associated with micrognathia and glossoptosis compromising the development of speech and hearing in this patient with Pierre Robin Sequence. The cleft was not operated until the patient was approximately four years old, and the patient remained with velopharyngeal dysfunction until 10 years of age, leading to a hypodynamic velopharynx with persisting use of glottal stops and pharyngeal fricatives. Notably, the patient did not receive speech therapy until she was 10 years old, particularly because of lack of specialized speech therapy in her hometown. The combination of intensive speech therapy and speech bulb reduction for this patient required the collaboration of an interdisciplinary team including SLPs, dentists, plastic surgeons, and psychologists. In this case, the involvement of the patient and her mother in treatment decision and planning was essential to ensure adherence, considering that the intensive program required commitment to daily practice.

Intensive treatment for correction of cleft palate speech, described in this case report and in the literature[6-10,12], is important for populations that do not have access to specialized treatment in their hometowns. Correction of speech mislearning associated with VPD is complex and requires strategies that involve contrasting the *old* with the *new speech patterns* while alternating between different sizes of speech bulbs to address velopharyngeal hypodynamism [3,17].

The history of this patient corroborates the importance of prevention of atypical speech errors (such as replacement of oral speech sounds with glottal stops) in order to reduce the burden of care involved in the management of CLP. Timing and successful outcome of primary palatoplasty play an important role in the development of adequate speech and should be a priority at most craniofacial centers. Whereas treatment of passive speech errors, such as nasal air emission and hypernasality (obligatory in the presence of VPD and unoperated cleft palate), requires physical management for establishment of the possibility of velopharyngeal closure during speech[4], management of articulatory errors involving the use of atypical post-uvular place of production (compensatory articulation) can be prevented, or at least minimized. Phonetic-based, parent-administered treatment approaches such as the corrective babbling speech treatment program[13], have been shown to help reduce cleft-related speech errors in some children with history of cleft palate and velopharyngeal dysfunction.

FINAL COMMENTS

In this case report, it was possible to observe that the patient demonstrated typical resonance and normal articulatory production after an ISTP involving a considerably large number of sessions (360) offered within a period of nine months. Late and unsuccessful primary palatoplasty, as well as lack of adequate speech therapy intervention, can explain the complexity of this case. Intensive speech therapy for treatment of speech disorders associated with VPD is not always a viable option for patients who do not have access to treatment in craniofacial centers or specialized care in their hometowns. In cases where there is a possibility of a partnership between the craniofacial team and speech therapists in the patient's hometown, a speech therapy program can be implemented using an alternative method such as the Tele-Partnership. The use of Teletherapy can be valuable in training specialists and beneficial in minimizing the use of public resources.

REFERENCES

1. Marques IL, Sousa TV, Carneiro AF, Peres SP, Barbieri MA, Bettiol H. Seqüência de Robin – protocolo único de tratamento. J Pediatr. 2005;81(1):14-22. PMid:15742081. http://dx.doi.org/10.2223/1277.
2. Prado-Oliveira R, Marques IL, Souza L, Souza-Brosco TV, Dutka JC. Assessment of Speech nasality in children with Robin Sequence. CoDAS. 2015;27(1):51-7. PMid:25885197. http://dx.doi.org/10.1590/2317-1782/20152014055.
3. Dutka JCR, Uemeoka E, Aferri HC, Pegoraro-Krook MI, Marino VCC. Total obturation of velopharynx for treatment of velopharyngeal hypodynamism: case report. Cleft Palate Craniofac J. 2012;49(4):488-93. PMid:21417778. http://dx.doi.org/10.1597/09-240.
4. Golding-Kushner KJ. Treatment of articulation and resonance disorders associated with cleft palate and VPI. In: Shprintzen RJ, Bardach J. Cleft palate speech management: a multidisciplinary approach. Saint Louis: Mosby; 1995. p. 327-51.
5. Bessell A, Sell D, Whiting P, Roulstone S, Albery L, Persson M, et al. Speech and language therapy interventions for children with cleft palate: a systematic review. Cleft Palate Craniofac J. 2013;50(1):e1-e17. PMid:22433039. http://dx.doi.org/10.1597/11-202.
6. Pamplona M, Ysunza A, Patino C, Ramírez E, Drucker M, Mazón JJ. Speech summer camp for treating articulation disorders in cleft palate patients. Int J Pediatr Otorhinolaryngol. 2005;69(3):351-9. PMid:15733594. http://dx.doi.org/10.1016/j.ijporl.2004.10.012.
7. Pamplona MC, Ysunza A, Perez G, Vergara S. Summer school speech therapy for children with cleft palate and language disorder. Gac Med Mex. 2009;145(6):475-9. PMid:20077865.
8. Pamplona MC, Ysunza A, Chavelas K, Arámburu E, Patiño C, Martí F, et al. A study of strategies for treating compensatory articulation in patients with cleft palate. J Maxillofac Oral Surg. 2012;11(2):144-51. PMid:23730060. http://dx.doi.org/10.1007/s12663-011-0314-x.
9. Lima MRF, Leal FB, Araújo SVS, Matos EF, Ninno CQMS, Britto ATBO. Atendimento fonoaudiológico intensivo em pacientes operados de fissura labiopalatina: relato de casos. Rev Soc Bras Fonoaudiol. 2007;12(3):240-6. http://dx.doi.org/10.1590/S1516-80342007000300012.
10. Bispo NHM, Whitaker ME, Aferri HC, Neves JDA, Dutka JCR, Pegoraro-Krook MI. Speech therapy for compensatory articulations and velopharyngeal function: a case report. J Appl Oral Sci. 2011;19(6):679-84. PMid:22231007. http://dx.doi.org/10.1590/S1678-77572011000600023.
11. Golding-Kushner KJ. Therapy techniques for cleft palate speech and related disorders. San Diego: Singular; 2001.
12. Pinto MDB. Resultados da fonoterapia intensiva para correção da oclusiva glotal e fricativa faríngea na fissura labiopalatina [tese]. Bauru: Universidade de São Paulo; 2016.
13. Dobbelsteyn C, Bird EK, Parker J, Griffiths C, Budden A, Flood K, et al. Effectiveness of the corrective babbling speech treatment program for children with a history of cleft palate or velopharyngeal dysfunction. Cleft Palate Craniofac J. 2014;51(2):129-44. PMid:24010867. http://dx.doi.org/10.1597/12-188.
14. Shin YJ, Ko SO. Successful and rapid response of speech bulb reduction program combined with speech therapy in velopharyngeal dysfunction: a case report. Maxillofac Plast Reconstr Surg. 2015;37(1):22. PMid:26258115. http://dx.doi.org/10.1186/s40902-015-0022-4.
15. ASHA: American Speech-Language-Hearing Association [Internet]. Steps in the process of evidence-based practice. Rockville: ASHA; 2017a

[cited 2017 Sep 25]. Available from: http://www.asha.org/Research/EBP/Finding-the-Evidence/

16. ASHA: American Speech-Language-Hearing Association [Internet]. Individual studies. Rockville: ASHA; 2017b [cited 2017 Sep 25]. Available from: http://www.asha.org/Research/EBP/Finding-the-Evidence/#individual
17. Polzin17 ACZ. Estratégias terapêuticas para abordar o funcionamento velofaríngeo durante tratamento de distúrbios da fala relacionados à FLP e DVF: relato descritivo [monografia]. Bauru: Residência Multiprofissional em Saúde: Síndromes e Anomalias Craniofaciais, Hospital de Reabilitação de Anomalias Craniofaciais, Universidade de São Paulo; 2015.

Author contributions

MDBP, LKFA and APCC and were involved in the speech therapy program and in the preparation and design of the study; they were also involved in collecting, preparing, and comparing data and in the preparation and review of this manuscript; JCRD and MIPK were responsible for the coordination of the speech therapy program and for the study design; they prepared, compared, and analyzed the data and were involved in the manuscript preparation, translation, and revision; LIRL was involved in the study design, preparation of the manuscript, comparison and analysis of the data, as well as in the translation and revision of this manuscript.