



**Research Article**

**REHABILITATION OF ORAL FEEDING ABILITY IN A TRACHEOSTOMIZED ADOLESCENT WITH NF2: FROM MECHANICAL VENTILATION TO AMBIENT AIR**

**Nishi TCS<sup>1\*</sup>, Bortolatto NO<sup>1</sup>, da Silva NS<sup>2</sup>, Cappellano AM<sup>2</sup>, Petrilli RBN<sup>3</sup>, Pinho RS<sup>4</sup>, Dastoli PA<sup>5</sup>, Cavalheiro S<sup>5</sup>, and Gonçalves MIR<sup>1</sup>**

<sup>1</sup>Department of Speech-Language Pathology, Institute of Pediatric Oncology (IOP-GRAACC/UNIFESP), São Paulo, Brazil

<sup>2</sup> Department of Neuro Oncology of Institute of Pediatric Oncology (IOP-GRAACC/UNIFESP), São Paulo, Brazil

<sup>3</sup>Otorhinolaryngologist of Institute of Pediatric Oncology (IOP-GRAACC/UNIFESP), São Paulo, Brazil

<sup>4</sup>Pediatric Neurologist of Institute of Pediatric Oncology (IOP-GRAACC/UNIFESP), São Paulo, Brazil

<sup>5</sup>Pediatric Neurosurgeon of Institute of Pediatric Oncology (IOP-GRAACC/UNIFESP), São Paulo, Brazil

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**ABSTRACT**

The objective of this manuscript was to report the speech-language pathology (SLP) rehabilitation in a tracheostomized patient, presenting the possibility of early intervention from mechanical ventilation. This is a retrospective clinical case study, by means of data collection of medical records, of a 15 year-old tetraparetic male with upper limbs paraesthesia, global muscle weakness and cutaneous nodules, diagnosed with neurofibromatosis type 2; and 3 different histological types of brain tumors in three different locations. After two tumoral resections and one cervical traction, he presented impairment of the spine and progressive motor and respiratory distress, evolving with pneumonia and prolonged orotracheal intubation (OTI). Two cervical arthrodesis for cervical deformity and vertebral slip were required, as well as a tracheostomy, leading to communication and swallowing difficult. It was required SLP intervention to rehabilitate feeding and oral communication, initially in continuous mechanical ventilation, then progressing to ambient air. We observed, not only less risk of aspiration to the patient, but also the family participation had fundamental importance in rehabilitation.

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**INTRODUCTION**

Neurofibromatosis (NF) is a common tumor predisposition syndrome, classified in types 1 and 2 (Brossier & Gutmann, 2015) and NF2 prevalence is estimated to be 1 per 35,000 persons/year, with clinical symptoms that compromise the skin, nervous system, bones, eyes and other sites characterized by the occurrence of bilateral vestibular schwannomas and other central nervous system (CNS) tumors (Eldridge, 1981; Evans, Huson & Donnai, 1992; Antonio, Goloni-Bertollo & Trídico, 2013). Brain cancers and other cancers of the nervous system are the second most common type (26%) among childhood neoplasms and the most common solid tumors in the pediatric age group (Siegel, Miller & Jemal, 2016). Symptoms of brain tumors are highly variable and are dependent on its location, size and treatment. Several studies have reported dysphagia (swallowing disorder) as a primary or secondary complaint in patients with brain tumors (Frank *et al.*, 1989; Straube & Witt, 1990; Hensel *et al.*, 1997; Daniels, 2006), especially those located in the posterior fossa (Albert *et al.*, 2004).

Prolonged orotracheal intubation (OTI), defined as an intubation lasting more than 48 hours (Brown *et al.*, 2011), may also be responsible for dysphagia (Heffner, 2010), but literature shows a prevalence of 3 to 62% in intubated patients (Skoretz, Flowers & Martino, 2010).

Patients of different diagnoses who required prolonged intubation were found to have a higher incidence of post extubation dysphagia (PED) compared to postoperative patients with a shorter duration of OTI (Rassameehiran *et al.*, 2015). The mechanisms of PED are multifactorial, causing the dysregulation of the swallowing reflex and the increase in risk factors (Goldsmith, 2000). Researches verified that more than 50% of patients with PED were intubated for over 48 hours, showing that the prolonged mechanical ventilation is one of the factors that can cause laryngeal irritation and inflammation (Leder, Cohn & Moller, 1998; Barker *et al.*, 2009). Various objective exams are used for evaluation of PED, such as videofluoroscopic swallow study (VFSS) and fiberoptic endoscopic evaluation of swallowing (FEES). FEES, mainly used at bedside evaluations, allows a direct observation of the pharyngeal phase of swallowing via a fiberoptic nasopharyngolaryngoscope with detailed information also

**\*Corresponding author: Talita Cristina de Sousa**

Department of Speech-Language Pathology, Institute of Pediatric Oncology of Federal University of São Paulo (IOP-GRAACC/UNIFESP), São Paulo, Brazil

regarding laryngeal function, relevant for airway protection (Bastian, 1991; Hanners, 2013).

**Case Report**

Patient R.O.P., male, 15 years and 9 months old, from Roraima State, Brazil, was referred to São Paulo Hospital (HSP) with features suggestive of type 2 neurofibromatosis. Then, he was admitted to the Pediatric Oncology Institute (IOP-GRAACC/UNIFESP) in São Paulo city, Brazil, on April 1st 2015, with paraesthesia in the upper limbs for about 4 months, initially on fingers, progressing to weakness of the distal upper and lower limbs, associated with weight loss (10Kg in 4 months), muscular hypotrophy and cutaneous nodules in the trunk and upper limbs.

Magnetic resonance imaging (MRI) of the skull and spine showed an intramedullary expansive process of about 15cm in length, with anatomopathology of bulb-cervical schwannoma and also left frontal meningeoma and bilateral acoustic neurinoma (Fig. 1). In clinical neurological evaluation it was observed tetraparesis assymetrical with deep tendon reflexes absent, atrophy of upper limbs, spasticity and clonus after dorsiflexion of the foot. The electromyography evidenced multiple motora-sensory mononeuropathy.

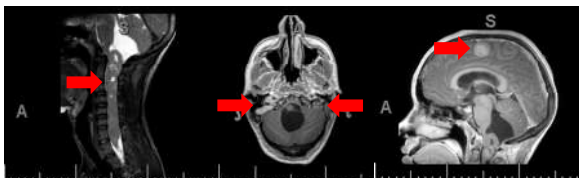


Fig 1 MRI showing the three primary sites of brain tumors diagnosed.

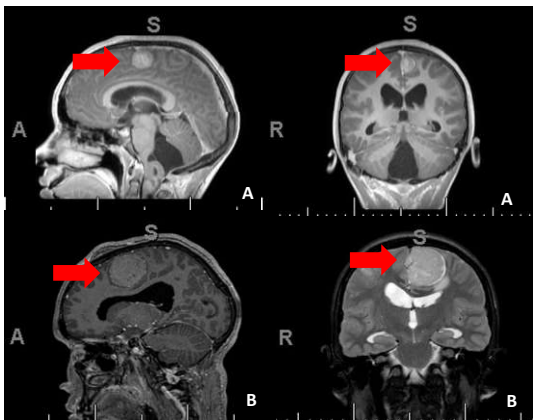


Fig 2 MRI showing the increase in the meningeoma dimensions, between March (A) and November (B) 2016

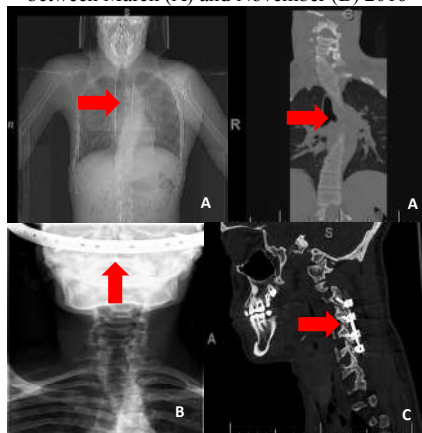


Fig 3 CT scan showing deformity in the cervico-thoracic spine (A), cervical traction (B) and posterior arthrodesis (C).

R. was submitted to neurosurgery for total resection of the bulb-cervical intramedullary tumor on April 10th, requiring orotracheal intubation for two days due to atelectasis. After 4 days of surgery, showed improvement of left hemibody movement, and was clinically evaluated by the speech-language pathologist (SLP) and physical therapy, receiving hospital discharge on April 17th.

One month after discharge, in neurological evaluation, R. presented signs of improvement of trunk and cervical control (axial hypotonia), but maintained spastic tetraparesis, although in rehabilitation with physical therapy. Patient was released to return to his home state in October.

One year later, the patient returned to follow-up appointments, with several complaints: worsening of weakness in upper limb right and axial hypotonia. His audiometry showed bilateral sensorineural hearing loss (6 and 8KHz on the right and 3 to 8kHz on the left). Due to increase of meningeoma dimensions (Fig. 2), it was performed total surgical resection on December 9th 2016, again without signs of dysphagia, being hospital discharged 3 days later.

At the end of June, R. presented worsening of weakness and of sensitivity in upper and lower limbs, with difficulty for walking and performing daily activities, and fatigue. After control computed tomography (CT) of cervical and thoracic spine it was observed a worsening of the listese (vertebral slip) and cervical kyphosis (deviation in the spine associated with scoliosis), intensifying the compressive effect in cervical cord. It was performed cervical traction and posterior C3-C6 arthrodesis with cage disc placement in C3-C4 on June 23rd for correction of deformity, with hospital discharge 4 days after procedure (Fig. 3). However, on August 2nd, the patient reported significant respiratory distress and loss of motor power in the 4 limbs for 4 weeks, being hospitalized and submitted to new traction after evidence of arthrodesis release and worsen of cervical deformity.

The patient showed signs of motor and sensory dysfunction and respiratory distress, even in non-invasive ventilation due to pneumonia, requiring orotracheal intubation (OTI) on August 13th and extension of the arthrodesis to C2-C7 on August 22nd. After 28 days of OTI and two extubation failures, the patient was submitted to tracheostomy. The clinical bedside swallowing assessment was performed 9 days after the tracheostomy, with the patient in continuous mechanical ventilation (CMV) with inflated cuff, being verified risk of bronchoaspiration due to reduced strength, mobility and coordination of oropharyngeal structures, fatigue, and respiratory discomfort. Swallowing therapy consisted of: direct therapy with food intake maneuvers; specific exercises aiming to reestablish strength, mobility and coordination of oropharyngeal structures; and use of a Passy Muir® speaking valve (PMV), when he could tolerate deflated cuff, to assist verbal communication. Oral feeding with pasty and liquid consistencies was possible on the fifth day; full oral diet occurred within 9 days of rehabilitation, on September 29th, under CMV conditions and inflated cuff due to risk of lack of coordination between swallowing and breathing.

FEES was performed by the otorhinolaryngologist, with SLP attendance, for objective swallowing evaluation due to odynophagia, on October 24th and showed candidiasis, absence of salivary stasis, presence of hypopharynx sensitivity,

mobile vocal folds and preserved swallowing, without aspiration of any consistencies.

These findings set new goals regarding SLP intervention. With the aim of assisting in verbal communication without respiratory discomfort, we added exercises to improve airflow control, pneumophonoarticulatory coordination and glottic coaptation during PMV training in mechanical ventilation, with progressive improvement and good performance with the device upon reaching 1 hour of PMV on October 31st.

Feeding and speech with PMV in nebulization occurred in November, progressing quickly to ambient air, with R. reaching above 2 hours in 2 periods of the day. On January 16th 2018, patient was able to receive oral feeding without restrictions in ambient air, being hospital discharged 2 days later.

In his first outpatient return in February, R. reported good food acceptance with complaint of fatigue during speaking with PMV, which was then used 3 to 4 hours a day. In the second outpatient return, in May, the patient related at least 6 hour a day of PMV, without complaints, use of positive pressure airway with BIPAP for sleep only and to be exclusively oral feeding with weight gain (Fig. 4).

## DISCUSSION

Patients admitted to the intensive care unit (ICU) usually present alterations of different functions and abilities; respiratory distress, muscle atrophy, decreased sensitivity of oropharyngeal structures and need for tracheostomy, as also occurred initially in this case, are risk factors for complications in the swallowing process ( DeVita & Spierer-Rundback, 1990; Elpern *et al.*, 1994; Tolep, Getch & Criner, 1996). Contrary to what other studies that reported, an average of two weeks for the deglutition assessment and one month for full diet reintroduction after performing tracheostomy (Freeman-Sanderson *et al.*, 2011), this patient progressed in less than twenty days after the procedure. Weight (approximately 3kg) and muscle mass gain were also observed one month after starting oral feeding on this case.

Literature shows that early intervention under mechanical ventilatory conditions contributes to the reduction of the risk for aspiration related to pulmonary distress and to nutritional recovery and, consequently, to the general health of patients in intensive care (Macht *et al.*, 2013; Rodrigues *et al.*, 2015). The training and participation of family members were another important aspect for the rapid progression of the diet; the good understanding of the guidelines, from the ideal patient positioning to the control of volume and frequency of supply, allowed the patient to receive support during all meals.

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