REVISTA CEFAC
SPEECH, LANGUAGE, HEARING SCIENCES AND EDUCATION JOURNAL
Rev. CEFAC. 2016 Maio-Jun; 18(3):721-726 doi: 10.1590/1982-0216201618314615

Original articles

Oropharyngeal dysphagia in Wallenberg's syndrome – case series

Disfagia orofaríngea na Síndrome de Wallenberg – série de casos

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Conflict of interest: non-existent

Received on: September 22, 2015
Accepted on: February 16, 2016

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ABSTRACT

Purpose: to characterize the changes of swallowing function in patients with Wallenberg's syndrome.

Methods: case series of seven patients with diagnosis of this syndrome referred for clinical assessment. To evaluate the degree of dysphagia used to Gugging Swallowing Screen scale and to assess the level of oral ingestion used the Oral Intake Functional Scale.

Results: the mean age was 60.57 years; all subjects showed changes in the function of severe degree of swallowing (71.42%) to moderate (28.58%); 85.71% required alternative pathway of food, and, 71.43% were fed exclusively by nasoenteric probe; all required speech therapy.

Conclusion: this study concluded that the oropharyngeal dysphagia in Wallenberg syndrome presents itself as moderate to severe disorder, being necessary the use of alternative pathway of food in most cases.

Keywords: Deglutition Disorders; Stroke; Lateral Medullary Syndrome

RESUMO

Objetivo: caracterizar o quadro de comprometimento da função da deglutição em pacientes com Síndrome de Wallenberg.

Métodos: série de casos de sete pacientes, com diagnóstico dessa síndrome, encaminhados para avaliação fonoaudiológica. Para avaliação do grau de disfagia utilizou-se a escala *Gugging Swallowing Screen* e para avaliar o nível de ingestão oral utilizou-se a *Functional Oral Intake Scale*.

Resultados: a média de idade foi de 60,57 anos; todos os sujeitos apresentaram alteração na função da deglutição de grau grave (71,42%) a moderado (28,58%); 85,71% necessitaram de Via Alternativa de Alimentação, sendo que, 71,43% eram alimentados exclusivamente por sonda nasoentérica; todos necessitaram de acompanhamento fonoaudiológico.

Conclusão: este estudo concluiu que a disfagia orofaríngea na Síndrome de Wallenberg apresenta-se como um distúrbio de grau grave a moderado, sendo necessária a utilização de Via Alternativa de Alimentação na maioria dos casos.

Descritores: Transtornos de Deglutição; Acidente Vascular Cerebral; Síndrome de Wallenberg

INTRODUCTION

The Wallenberg Syndrome (WS), also called Lateral Bulbar Syndrome, is retro-olivary injury generally resulting from an Arterial Ischemic Stroke (AIS) in intracranial portion of the Vertebral Artery or its Cerebellar Posterior Inferior branch, which is responsible for the vascularization of the dorsolateral region of the bulb. The initial manifestations in WS are: limb ataxia, nausea, dizziness, vomiting, nystagmus, difficulty in balance and walking, dysarthria, dysphonia, oropharyngeal dysphagia (OD) neurogenic, being the percentage of occurrence of the last manifestation ranging from 51 to 94% [1-4].

Neurogenic OD is a secondary symptom to an underlying disease or neurological trauma that cause, in most cases, a sensory-motor impairment in the oral and/or pharyngeal swallowing. Taken together, these changes can result in dehydration, malnutrition and aspiration pneumonia due to laryngeal penetration and tracheal aspiration[5,6]. Mortality after episodes of aspiration pneumonia is significant, with occurrence ranging from 7.5 to 72%[7].

In WS, among different kinds of impairment, cranial nerves Trigeminal (V) are affected, being them responsible for the muscles of chewing, tensor muscle of the soft palate and sensitivity of the face and 2/3 of anterior part of the tongue; Glossopharyngeal (IX), responsible for the sensitivity and taste of the posterior third of the tongue and innervation of the constrictor muscles of the pharynx and stylopharyngeus muscle; and Vago (X), responsible for motor and sensory functions of the pharynx and larynx, and branches of the last two nerves form the pharyngeal plexus[1,6]. Thus, lesions in these cranial nerves interfere with the swallowing process, they can cause uncontrollable sobs; ipsilateral paralysis to the lesion, palate and vocal cords; ipsilateral facial hypalgesia and possible loss of taste in hemi-tongue[8,9].

OD after WS is frequently classified as a severe degree, affecting the pharyngeal phase of swallowing, and the prognosis depends on the extent and location of the lesion, which may vary from complete recovery to a permanent vegetative basis[10-14]. Furthermore, patients with neurogenic OD might present other neurological symptoms and deficits in cognitive abilities due to injury in areas of the central nervous system, which may complicate the clinical condition[15].

The speech therapy in swallowing disorders aims at the early detection of dysphagia, at the elimination of the possible risk associated complications and, therefore, at stabilizing the nutritional status[16]. The treatment of dysphagia in WS is based in signs and symptoms, as the focus of therapy is the reduction of aspiration risk and not removing the cause, it may be necessary to recommend the use of an alternative feeding route (AFR). This, together a swallowing rehabilitation program based on techniques of oral stimulation, facilitating maneuvers and postural maneuvers, may bring benefits to the patient[10,11,13].

Consequently, the aim of this study was to characterize the impairment condition of the swallowing process in patients with WS.

METHODS

It is a study of retrospective case series, approved by the Ethics and Research Committee (ERC) of *Universidade Federal de Ciências da Saúde de Porto Alegre (UFCSPA)* under the number 362795, according to the rules established by the resolution No. 196/96 of the National Health Council and its subsequent resolutions. Thus, as it is a search in databases, all researchers have signed a data privacy statement.

Data from subjects were collected from the Database of academic activities of the Speech, Hearing and Language Sciences Major of UFCSPA at *Santa Clara Hospital* at *Santa Casa de Misericordia Hospital Complex in Porto Alegre* (RS, Brazil). Data were analyzed from the following inclusion criteria: being admitted to the neurology sector of the hospital during the period from January 2012 to August 2013; have been diagnosed with WS by the group of neurology, which was based on clinical criteria and confirmed by neuroimaging (MRI); receiving speech therapy, performed by the interns of the Speech, Hearing and Language Sciences Major, who were well trained and interns who were under supervision of an expert speech therapist professor. Finally, all subjects hospitalized in the period, with this diagnosis, contemplated the above criteria and they were included in the survey.

The data collected from medical and speech therapy records of seven patients with WS diagnosis were: date of birth, date of hospital admission and discharge, medical staff responsible for patient referral, previous medical history, medical admission diagnosis, lesion location, date clinical assessment, level of oral intake at the time of evaluation and hospital discharge, initial degree of dysphagia and hospital discharge, alternative feeding route use (AFR) in the evaluation and discharge; use of mechanical ventilation (MV); tracheostomy (TR), date of the last speech therapy and laboratory tests related to swallowing.

In order to classify the degree of dysphagia, the Gugging Swallowing Screen scale (*GUSS*) was used[17]. This, in turn, is divided into two stages: direct and indirect assessment of swallowing. Thus, through the score, it is possible to classify the swallowing in normal or mild dysphagia with no or with minimal risk of aspiration (20 points), mild dysphagia with low risk of aspiration (15 to 19 points), moderate dysphagia with risk of aspiration (ten to 14 points) and severe dysphagia with a high risk of aspiration (zero to nine points).

To assess the level of oral ingestion Functional Oral Intake Scale (*FOIS*) was used[18]. It is used to scale, at levels from one to seven, the amount of intake by mouth (oral), while *FOIS 1* provides "not oral;" *FOIS 2* "dependent on alternative route with minima (oral) food or liquid"; *FOIS 3* "dependent alternative route with consistent food or liquid (oral)"; *FOIS 4* "(oral) total of a single consistency"; *FOIS 5* "(oral) complete with multiple consistencies, but in need of special preparation or compensation"; *FOIS 6* "(oral) complete with multiple consistencies, but without special preparation or compensation, but with restrictions for some food" and *FOIS 7* "total (oral) without restrictions."

The subjects were evaluated according to the routine of the institution, initially in the degree of dysphagia (*GUSS I*) and the level of oral intake (*FOIS I*) and those who underwent speech therapy were assessed at discharge (*GUSS II* and *FOIS II*).

After tabulating the data by using the Microsoft Office Excel spreadsheet, a descriptive analysis in absolute and relative values was performed.

RESULTS

For this case series, there were six men and one woman, identified as S1, S2, S3, S4, S5, S6 and S7, averaging 60.57 years (minimum of 38 and maximum of 76). In Table 1 the data of the subjects are reported.

From seven subjects, four (57%) presented systemic arterial hypertension (SAH), two (28.57%) had diabetes, one (14.28%) was obese, one (14, 28%) presented cardiomyopathy and one (14.24%) used to smoke. These were considered risk factors for Cerebral Vascular Accident (CVA). Only one patient (14.28%) had a diagnosis of previous stroke.

Table 1. Characterization of subjects with wallenberg syndrome

Subject	Genre	Age	Diagnosis	Location	AFR	TR
S1	M	76	IS; WS	Right Brainstem and Cerebellum	NET	Yes
S2	M	58	IS/h; WS	Right Brainstem and Cerebellum	NET	No
S3	M	72	IS; WS	Left Bulb	NET	Yes
S4	M	59	IS; WS	Left Brainstem and Cerebellum	NET	No
S5	M	70	IS; WS	Left Bulb	NET	No
S6	F	38	IS; WS	Left Bulb	No	No
S7	M	51	IS; WS	Left Brainstem and Cerebellum	NET	No

Caption: Diagnosis: Medical diagnostic; Location: Location of the lesion from Neuroimaging; AFR: Alternative Feeding Route use; TR: Tracheostomy use ; M: male; F: female; IS: Ischemic stroke; WS: Wallenberg Syndrome; IS/h: Ischemic stroke with hemorrhagic transformation; NET: Nasoenteric tube.

At the time of phonological assessment, five (71.43%) subjects presented degree of severe dysphagia and two (29.57%) had moderate dysphagia. Six subjects (85.71%) needed AFR. From these ones, five (71.43%) used exclusively AFR (*FOIS 1*) and (14.28%) used AFR in pasty consistency (oral) (*FOIS 2*). One (14.28%) subject received exclusive oral feeding in paste consistency (*FOIS 4*).

Two (28.57%) subjects made use of tracheostomy and, one of them (50%) required prolonged mechanical ventilation.

Regarding speech therapy conduct, all patients received speech therapy indication and four (57.14%) needed further investigation by swallowing videofluoroscopy (VFS), however, due to the routine of the service, no subject carried out the objective evaluation of swallowing.

From seven patients, five (71.43%) remain hospitalized with speech therapy follow-up three times a week, and from these subjects, two (40%) improved from severe dysphagia and *FOIS 1* to moderate or mild dysphagia and *FOIS 2* or *FOIS 7* respectively, while the

other three subjects (60%) did not present improvement in the degree of dysphagia and level of oral intake. Two (28.57%) were discharged after clinical assessment, by appointment of the medical staff and therefore they did not receive speech therapy intervention while hospitalized. One (50%) of the subjects had oral diet indication in paste consistency with thickened liquid, and one (50%) had no oral feeding condition and received AFR indication. Both were referred to a speech therapy ambulatory. It is noteworthy that during the hospital stay, patients received speech therapy only three times a week due to the institution routine.

Table 2. Characterization of speech therapy evaluations of subjects with wallenberg syndrome

Subject	TH/STA (days)	GUSS I (pre)	FOIS I (pre)	TST (days)	GUSS II (post)	FOIS II (post)
S1	16	Severe dysphagia	1	80	Severe dysphagia	1
S2	7	Severe dysphagia	1	14	Mild dysphagia	7
S3	35	Severe dysphagia	1	1	NR	NR
S4	6	Severe dysphagia	1	17	Moderate dysphagia	2
S5	9	Severe dysphagia	1	11	Severe dysphagia	1
S6	7	Moderate dysphagia	4	1	NR	NR
S7	8	Moderate dysphagia	2	8	Moderate dysphagia	2

Caption: TH/STA: time between hospitalization and speech therapy evaluation; *GUSS* I: initial degree of dysphagia; *FOIS* I: level of oral intake at the time of evaluation; TST: time between speech therapy evaluation and discharge; *GUSS* II: dysphagia degree in discharge *FOIS* II: level of oral intake at the time of hospital discharge; NR: not reevaluated.

DISCUSSION

OD can be defined as a secondary symptom of the underlying disease, which prevents the correct food transportation. So, it is directly associated with the interruption of food pleasure and it may cause deficits in proper nutrition and hydration of patients affected by such symptom[5,19,20].

Studies show that the WS affects the oral and oral preparatory phases, due to motor and sensory impairments, which associated with changes in the intrinsic and extrinsic muscles of the larynx results in significant disturbances in the pharyngeal phase, which is considered the main phase of swallowing[8,9, 12-14,21,22].

A study evaluated 20 patients with WS by electromyography, verifying the occurrence of dysphagia in 95% of cases[12]. These findings are similar to the present study, which found dysphagia in all the cases evaluated with WS. Still, in relation to the severity of OD, for the aforementioned research, 45% of the subjects were diagnosed with severe dysphagia, while in this study 71.42% received the same diagnosis. This difference between the studies may be attributed to the use of different criteria for OD classification, as in other research, the degree of severe dysphagia was found by clinical evaluation, and a total of 11 patients with WS, was 63.63 % approaching more of this study data[23].

In this research, there was not oral intake evaluation (*FOIS 1*) or minimum (*FOIS 2*) for the six subjects, indicating the need for AFR initially. In the literature, other studies have described the inability of the patient to feed themselves orally only after the onset of OD signals[8,9,12,14,23,24] . The AFR chosen for all patients in this study was nasoenteric tube (NET), according to data from another reaearch[25]. However, due to the severity of dysphagia and its slow recovery, gastrostomy indication would be the most appropriate to these subjects.

This work showed that adult subjects had lower levels of OD and greater oral intake levels at baseline, whereas subjects in middle-aged and elderly had severe OD degree at baseline and *FOIS 1*. Therefore, it is important to note that the physiological changes in the swallowing process, due to aging, associated with a vulnerability to chronic diseases makes them sensitive to swallowing disorders and greater negative impact when afflicted with neurological diseases[20,26].

In this study, all subjects had speech therapy indication. However, due to the discharge of two subjects, only five received speech therapy during hospitalization. Of these five subjects, only two had reduction in the degree of OD and evolution at the level of oral intake. After hospital discharge, the five reevaluated subjects received referral to outpatient speech

therapy. The literature shows that the recovery of OD can be very slow and can take several months to years or even not present evolution[8,12,27,28]. Some studies have compared dysphagia in WS with dysphagia in hemispheric stroke, showing that in WS the dysphagia tends to be more severe and its recovery slower[12,29].

Still, in relation to speech therapy in patients who were referred for early evaluation, six and seven days showed the evolution in both the degree of OD, as the level of oral intake. It is known that early intervention in dysphagia minimizes the risk of complications, and provide the benefits as speech therapy and nutritional aspects[16,30].

In a study of 208 with dysphagic patients from different etiologies it was found that after swallowing therapy, 30% of patients with WS still needed alternative feeding route[29]. In this study, except for one subject who had complete evolution of swallowing passing from *FOIS 1* to *FOIS 7*, the other four, even undergoing speech therapy, remained with some food restriction and needing also of AFR. In addition, all required speech therapy after hospital discharge, according to the opinion of the team that treated the cases.

Some studies suggest the benefit of different therapies for dysphagia in WS such as repetitive transcranial magnetic stimulation; injection of botulinum toxin in the salivary glands; rehabilitation program based on techniques of oral tactile and thermal stimulation; postural maneuvers; pharyngeal maneuvers and facilitating strategies[8,13,21,23,27].

CONCLUSION

This study concluded that OD in SW presented itself as a serious degree of disturbance to moderate, requiring the use of AFR in most cases. It identified the importance of early assessment and speech therapy in order to prevent risks of lung and / or nutritional complications, and rehabilitate the function of swallowing. The multidisciplinary effort can ensure comprehensive care and promote better quality of life to these subjects.

More investigations related to dysphagia in WS are required for different evaluation methods and especially with larger groups of patients, as found in the scientific literature, most studies refers to the case of a single subject reports.

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