

# PRACTICE OF SPEECH THERAPY IN CONGENITAL HYDROCEPHALUS WITH VENTRICULOPERITONEAL SHUNT: A CASE REPORT

## *Atuação fonoaudiológica na hidrocefalia congênita com derivação ventrículo peritoneal: relato de caso*

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### ABSTRACT

Hydrocephalus is defined as a disorder in the cerebrospinal fluid hydrodynamics with increased intracranial compartment volume, ventricular dilatation and intracranial pressure. The treatment of hydrocephalus is made with the use of valves periventricular (shunt). If not treated, the child with hydrocephalus may display macrocephaly, neurodevelopmental delay, infections, meningitis, difficulties of learning and eating (dysphagia). To report the main Speech Pathology found diagnosed with congenital hydrocephalus, treated with ventriculoperitoneal shunt later. It is a retrospective study, descriptive of the data contained in medical records. The patient showed changes articulators, drooling, jaw clenching and Neurogenic Oropharyngeal Dysphagia grade Moderate/Severe. Exercises deep stimulation, cryotherapy, motor points of the face, facilitating maneuvers of swallowing and postural correction were performed during the offer orally. The literature for speech therapy in hydrocephalus is scarce, however, through this case, it was possible to report major speech pathology and therapeutic interventions found in Congenital Hydrocephalus.

**KEYWORDS:** Hydrocephalus; Speech, Language and Hearing Sciences; Deglutition Disorders

### ■ INTRODUCTION

Congenital Malformations in the Central Nervous System are more common in Child Neurosurgery field<sup>1-4</sup>; available data show that congenital malformations rates in the US population is 1: 1,000 live births<sup>2</sup>. According to ECLAMC – *Estudio Colaborativo Latinoamericano De Malformaciones Congenitas* (Argentina, Brazil, Chile, Colombia, Ecuador, Venezuela, Bolivia and Uruguay) – the number of malformed people, born in 2008, was 4.821<sup>5</sup>, while in Brazil the incidence rate is varied, occurring in 0,83: 1000 to 1,87: 1.000<sup>3</sup>. Among these anomalies, neural tube defects, myelomeningocele and hydrocephalus are highlighted<sup>2-4</sup>.

Hydrocephalus is the most common abnormality<sup>6</sup> occurring in 0.3 to 1.0: 2.000 births<sup>7</sup> and can be defined by a disturbance in the cerebrospinal fluid circulation, which leads to increased intraventricular pressure and volume of cerebrospinal fluid (CSF), leading to the dilation of the ventricles and nervous tissue compression<sup>3,6,8</sup>. Its etiology may be related to genetic factors such as obstruction of the Sylvius aqueduct, Dandy-Walker syndrome, Arnold Chiari malformation, agenesis cerebellar and spine bifida; as well as infectious factors: toxoplasmosis, cytomegalovirus and syphilis, or intracranial bleeding, multifactorial inheritance or congenital abnormality<sup>3,7,8</sup>.

The diagnosis can be made in during pregnancy by ultrasound (US), by puncture of umbilical cord or by karyotype analysis by collecting blood or after birth<sup>3,6,7,9</sup>, through Skull computerized tomography (CT) or a Magnetic Resonance Imaging (MRI)<sup>6</sup>.

Treatment of congenital hydrocephalus is made, in most cases, using periventricular bypass valves

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(*shunt*), which is a mechanism by which excess liquid is unidirectionally derived for other body cavities (atrium and peritoneum, for instance)<sup>10</sup>.

The child with untreated hydrocephalus may have macrocephaly, severe mental retardation, physical disabilities, a *shunt* malfunctioning, infections, meningitis, psychomotor development delay, problems for learning and visual, difficulties for food, persistent vomiting and / or convulsions and oral amendments, oral hygiene and dysphagia<sup>6,10-12</sup>, which is a swallowing disorder in with uncontrolled coordination of breathing and feeding, due to congenital or acquired neurological damage<sup>13</sup>.

The health of patients with hydrocephalus requires special care, for the prevention of post-operative complications and reduced sequelae<sup>12</sup>.

The speech therapist in hospitals is responsible for patient in bed, a preventive and intensive way, pre and post-surgery care, with technical support for interdisciplinary team, in order to prevent or minimize the consequences in neurological disorders in feeding (dysphagia) and / or communication<sup>13,14</sup>. This study aims to report the main speech pathologies found in an individual diagnosed with congenital hydrocephalus, treated late with peritoneal shunt ventricle.

## ■ CASE PRESENTATION

This study examined a male patient, 1 year and 1 month old, diagnosed with congenital hydrocephalus admitted at a pediatric emergency department of a university hospital in Goiânia-GO, in October 2012. The Ethics Committee for Research on Human Beings of Hospital das Clínicas, Federal university of Goiás, and protocol approved this study. 556,373.

In pregnancy history, the child's mother reported that had not done prenatal because the family rejected the pregnancy causing her a depression. She also reported that, in the family, there was a case of hydrocephalus in a cousin of third degree. His maternal great-grandmother and great-aunt had seizures.

According to the mother, with a difficult pregnancy, an emergency USG was held, and fetal hydrocephalus was detected. At 40 weeks and 3 days of gestation, a cesarean delivery was performed, the newborn (NB) weighed 2.535g, 35 cm of head circumference (HC), height 45,5 cm and APGAR index 7 and 9 for the first and fifth minutes, respectively. The transfontanellar USG proved severe hydrocephalus. Physiological neonatal jaundice signal was observed and the newborn was referred to the NICU (Neonatal Intensive Care Unit). After 15 days in the NICU, the newborn was discharged without guidance on medication or surgical treatment

of hydrocephalus for Ventriculoperitoneal Derivation (VPD) installation

In the hospital, the child weighed 15kg, with HC = 89cm, heart rate (HR) of 108 bpm and respiratory rate of 24 breaths per minute. At the neuro-developmental examination (NPPE), the child did not set his eyes, did not articulate words, cried a lot, did not crawl, did not move or sit. In Glasgow scale, the index was equal to 13 indicating minimal neurological injury.

A clinical speech therapist assessment was required, because his mother reported that the child had feeding difficulties for soft diet. In the physical examination he was awake, whining, with stiffness in upper and lower limbs, oxygen saturation level (Sat O<sub>2</sub>) = 86%, HR = 54 bpm.

The speech therapy was performed two days before the surgery to place the VPD, to evaluate myofunctional bodies, identify changes in the dynamics of swallowing, characterize clinical suggestive signs of penetration / aspiration, presence of dysphagia and establish procedures from these results.

The child showed pathological reflex in the bite, the resting tongue posture was between the arcades and parted lips, deciduous teeth in poor condition, symmetrical and hypotonic cheeks and mixed type breathing. The mobility of sound articulators (MSA) was limited and performed masticatory movements upright and the cervical auscultation, the stasis saliva/ secretion was evidenced in the laryngeal region, not discarding aspiration hazard. There was no respiratory distress.

For the clinical evaluation of swallowing was used liquid-soft diet (banana milkshake with thickener), offered in the spoon : the patient did not capture the food, the lip sealing was ineffective, exacerbated lingual protrusion during chewing, swallowing , oral escape previous, food remains in the mouth after swallowing and cervical auscultation showed food stasis in the larynx with presence of choking. These characteristics above, with a pasty consistency, were present but was not detected food stasis or choking.

Due to data in the evaluation, it was possible to diagnose in functional swallowing to pasty consistency and neurogenic oropharyngeal dysphagia moderate for pasty liquid. In a speech therapy approach, it was shown a pasty thickened liquid diet for safe food, and speech exercises were performed to the patient rehabilitation, such as deep brain stimulation MSA, cryotherapy, facial motor points (neuromuscular activation), active exercises, facilitating maneuvers of swallowing and postural correction during the diet offer orally due to the

difficulty created by the sharp growth of the HC and the weight of the head.

The surgery for VPD placement was performed two days later. Within 11 days, the CP had decreased about 10 cm and the patient was feeding with soft diet by mouth (orally) with no complications. The patient was discharged by the programming team and came back to his hometown with referral report for speech therapy.

The patient returned to the emergency room with 1 year and 4 months, with VPD infection and weight loss, abdominal pain, difficulty accepting the diet orally and reported that he had not performed speech therapy in his city.

In clinical assessment, these aspects were observed: clenching jaw, dental breaks and in poor condition, drooling, and grunting and refusal the diet, by changing the O<sub>2</sub> Sat before, during and after swallowing, being compatible with the Oropharyngeal Neurogenic Dysphagia with degree of Moderate / Severe and diet indication by Nasoenteric probe (NEP).

## ■ RESULTS

New treatment plans were developed in order to reduce drooling and improve the oromyofunctional standard, while the medical team investigated the VPD infection.

After 19 days in hospital due to infection diagnosed as meningitis, he also received daily speech therapy interventions, we observed significant reduction in drooling. On the same day, there was a Sat O<sub>2</sub> fall, followed by cyanosis, which led to continuous oximetry and supply of O<sub>2</sub> by nasal catheter. The speech therapy was discontinued and did not improve the dysphagia, the patient continued in use of NEP. A therapeutic treatment with antibiotic was performed for 53, when there was an improvement of infection, the patient was referred back to his city and for outpatient care with pediatric neurologist. There were no more therapeutic sessions recorded.

## ■ DISCUSSION

Prenatal is an important follow up and can detect fetal abnormalities during the pregnancy<sup>1-3,6,7,9</sup>; however, the patient's mother in this case did not perform prenatal, discovering Hydrocephalus just hours before delivery.

According to the gender distribution, several studies identify slight male predominance for Congenital Hydrocephalus<sup>1,15</sup> being limited in fetal period<sup>3,6,9,15-17</sup>, being this case report in the findings. The mean gestational age (GA) found was 36.6 weeks<sup>7</sup>, with the IG's case over the average (40.3

weeks) and the case of HC (35cm) was within the thresholds (26cm to 57cm) found in the literature<sup>7</sup>.

All neurological alterations found in this case, from NPPE to talk or feeding skills have been described in the literature, with some kind of alteration or incapacity<sup>6,10,11,13,18</sup>.

The description of speech therapy in the literature is found in neurological pathologies such as Cerebral paralysis<sup>19</sup>, with the characterization of stomatognathic system functions and rehabilitation of dysphagia, but did not find any study to investigate in Hydrocephalus Congenital, changes in swallowing or dysphagia. Only one study showed the speech disorders of communicative and orofacial profile in hydrocephalus, with reports of orofacial muscle tone changes and strategies used to fit<sup>20</sup>.

The Brazilian Speech Therapy Protocol of Risk Assessment for Dysphagia (PRAD) has been used in speech therapy to research the swallowing disorders<sup>21</sup>. The use of this protocol was not described for data collection, however, it was possible to compare medical records according to PRAD parameters.

According to the National Institute of Neurological Disorders and Stroke – NINDS, infections in the bypass valves are commonly found, and among the best known, meningitis<sup>22</sup>, as the patient in this study.

Children with global motor development changes may show abnormalities in the development of oral motor control, negatively influencing functions performance of chewing and swallowing and consequently the swallowing control saliva<sup>23</sup>, providing the drooling, as was found in this case.

Therapeutic interventions performed by a speech therapist, according to American Speech-Language-Hearing Association – ASHA, may involve instruction to the diet supply and / or modification of its consistency, the stimulation of MSA and the use of adaptive utensils and food / or alternative nutrition<sup>24</sup>, justifying the use of strategies and alternative ways to feed our patient.

Prevention of postoperative complications should be performed in an interdisciplinary way, aiming a better quality of life as the body hydration, nutrition, safeness and effective supply route (if there is neurological damage), prevention of pressure ulcers in the head and skin care<sup>11,22,24</sup>.

## ■ FINAL CONSIDERATIONS

The speech therapist role in inter and multidisciplinary team showed differential in the patient care with congenital hydrocephalus, which can be

observed temporary improvement due to worsening of neurological symptoms, however, it is noteworthy that this action is very important in order to minimize sequelae and / or maintain oral functions, focusing

on better quality of life. The complications of the patient and the difficulty for taking care by distance from his homeland interrupted the intervention continuity.

## RESUMO

A Hidrocefalia é definida como um transtorno na hidrodinâmica liquórica, com aumento do seu volume no compartimento intracraniano, da dilatação ventricular e da pressão intracraniana. O tratamento da hidrocefalia é feito com uso de válvulas de derivação periventricular (*shunt*). Se não for tratada, a criança com hidrocefalia poderá apresentar macrocefalia, retardo do desenvolvimento neuropsicomotor, infecções, meningites, dificuldades para aprender e alimentar-se (disfagia). Este estudo busca relatar as principais alterações fonoaudiológicas encontradas em um indivíduo diagnosticado com hidrocefalia congênita, tratada tardiamente com derivação ventrículo peritoneal. Trata-se de um estudo retrospectivo, descritivo dos dados contidos no prontuário. O paciente apresentou alterações de órgãos fonoarticulatórios, sialorréia, apertamento mandibular e Disfagia Orofaríngea Neurogênica de grau Moderado/Severo. Foram realizados exercícios de estimulação profunda, crioterapia, pontos motores da face, manobras facilitadoras da deglutição e correção postural durante a oferta de via oral. A literatura para atuação fonoaudiológica na hidrocefalia é escassa, porém, com este relato de caso, foi possível relatar as principais alterações fonoaudiológicas e intervenções terapêuticas encontradas na Hidrocefalia Congênita.

**DESCRITORES:** Hidrocefalia; Fonoaudiologia; Transtornos da Deglutição

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