Epidemiology of the amyotrophic lateral sclerosis
Bahia - Brazil

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Abstract
The Amyotrophic Lateral Sclerosis is a neurological disorder, with the degeneration of the upper and lower motor neurons. The aim is investigate the start of the symptoms, describe the findings and study the survival period of patients with ALS. We analyzed 70 patients. The patients’ average age was 49.68 years old and we found 43 patients (61.4%) who were white, 22 who were grayish brown (31.4%) and 5 who were black (7.1%). Regarding the start of the symptoms, 51 patients (72.9%) showed a distal start, 31 a proximal one (44.3%) and 8 of them (11.4%) showed a bulbar start. The survival period, after de diagnosis, was of 64.11 months. The mean age, signs and symptoms and the patients’ survival period we found, are compatible with the ones found in the literature, except for the number of black patients, that was bigger in our survey.

Key words: Amyotrophic lateral sclerosis, Characteristics, Charcot’s disease, Epidemiology.

Resumo
A esclerose lateral amiotrófica (ELA) é uma desordem neurológica com degeneração dos neurônios motores superiores e inferiores. Objetivo: investigar o inicio dos sintomas, descrever a evolução e os achados neurológicos e estudar a sobrevida dos pacientes com ELA. Método: Analisamos 70 pacientes entre 1996 e 2007, que preencheram os critérios propostos no El Escorial, sendo 52 do sexo masculino e 18 do sexo feminino. A média de idade dos pacientes era de 49,6857 anos, encontramos 43 (61,4%) brancos, 22 (31,4%) pardos e 5 (7,1%) negros. Quanto ao início dos sintomas, 51 (72,9%) dos pacientes apresentaram início distal, 31 (44,3%) de forma proximal e 8 (11,4%) de forma bulbar. Os sintomas mais comuns foram as fraqueza muscular, atrofia muscular e miofasciculações presente em 69 (98,6%) pacientes. A sobrevida após diagnóstico foi de 64,116 meses. Conclusão: A idade média, os sinais e sintomas e a sobrevida dos pacientes analisados são compatíveis com os encontrados na literatura, exceto pela quantidade de pacientes da raça negra, que foi maior.

Palavras-chave: Esclerose lateral amiotrófica, epidemiologia, doença de Charcot.
Introduction

Amyotrophic lateral sclerosis (ALS) is a neurological disorder with the degeneration of upper and lower motor neurons. ALS is characterized by progressive fatigue, with atrophy, fasciculation, hyporeflexia or hyperreflexia and lipglossopharyngeal paralysis, that cause disability and death, in average, 5 years after the start of the symptoms. This disease was completely described - clinically and pathologically - by Charcot (1874) and since then, little information was added to his description. ALS is a neuromuscular disease, whose etiology is unknown, which is histo-pathologically characterized by a lesion in the motor nucleus of the cerebral trunk and of the corn of the spinal medulla. There is loss of upper and lower neurons that culminates in respiratory insufficiency and death after 3 to 5 years. Its incidence is of 0.5 to 3/100,000 people per year, in different regions in the world, except for endemic areas. It is a disease that generally affects adults, mainly between 50 and 60 years old, mainly in men (3:2) and only 10% of the cases occur before the age of 40 years old. The etiology of sporadic ALS is unknown and there are many hypotheses that may explain it, as: virus, metals, toxins, self-immune origin and endocrine dysfunction. A benchmark was the discovery, in 1993, of the mutations in the genetic code for the superoxide dismutase 1 from Cu/Zn (SOD1), in familial cases - it guided the research of ALS and elucidated the mechanism mediated by SOD1. Amyotrophic lateral sclerosis is a disease with poor prognostic and there is no cure for it. Information about the prognostics is very useful for parents, relatives and especially clinicians, because they give a notion of the most effective treatment. If we are aware about the factors that affect the prognostic, we can provide the patient with much and better information about the probable clinical course he/she will follow. The prognostic is associated with many factors, as age when the disease started, sex, how the disease started and evolution of the symptoms. The variety of the clinical presentation, that can arise prematurely, makes the absolute diagnosis very difficult. The diagnosis of ALS is based on the clinical presentation, but there is not a specific biological marker, nor neuroradiological or neuropsychiologic specific characteristics. Generally, the diagnosis is given through El Escorial that had been revised and proposed by the World Federation of Neurology. El Escorial was created through a consensus among experts in neuromuscular diseases that gathered in the city of El Escorial, in Spain. Nevertheless, two studies in New Jersey and in Italy showed that 24% of the patients in New Jersey and 40% of the patients in Italy had, in the beginning, an incorrect diagnosis. The objective of this study is to investigate the start of the symptoms, to describe their evolution and the neurologic findings and, to study the survival period of the patients with ALS.

Materials and Methods

In the present study, we used El Escorial’s criteria, revised in 1998 by the World Federation of Neurology. The diagnosis was made through anamnesis and neurological exam that excluded the possibility of other diseases. To help with this diagnosis, there were done Nuclear Magnetic Resonances, Computed Tomographies and Electromyographies, whenever necessary. This is a longitudinal study, and it was carried through filling out of a protocol with patients who suffered from ALS, in the first ambulatory appointment. The patients came from the Ambulatory of Diseases of the Motor Neuron - Foundation of Neurology and Neurosurgery - Institute of the Brain, that is located in Salvador, Bahia, Brazil, during the period between 1996 and 2007. Sixty-six patients with diagnosis of ALS defined by El Escorial were selected. Some of these patients were followed until their death. Criterias for inclusion: Patients with ALS defined through El Escorial, with 18 years old or more, and who had undergone, at least, an image exam and an electromyography. Criterias for exclusion: Exclusion of hypothesis for other diseases that can lead to the degeneration of upper and lower motor neurons; presence of another pathology that may have the same symptoms as the ones for ALS; impossibility to answer the questionnaire and doubts related to the patient’s diagnosis. Instrument: The specific protocol for ALS was used. This protocol contains the patient’s personal information, duration of disease, medical antecedents and familial cases found in physical exams, type of start of disease, associated diseases and instructional background. Analysis: In this case, the analysis will be evaluated through average frequency measures, standard deviations and the test χ², when the evaluation of the statistical significance to test hypothesis that categorized variables are independent.

Results

Seventy-four patients were selected, but 04 of them were excluded from the study for the following reasons: 2 of them because they did not fulfill the protocol and 2 for not having passed in the inclusion criteria in this study. Among the total number of patients, 52 (74,3%) were male and 18 (25,7%) were female. Patients’ average age was 49,68 years old - for men this average was 48,5 and for women it was 53,11, and there was not a significant difference between these two groups. When we subdivided the groups in races, we had 43 (61,4%) white people, 22 (31,4%) grayish brown and 5 black people.

Regarding the start of the symptoms, the Syndrome of the Motor Neuron and the signs and symptoms of ALS, the result can be seen in the table 01. Patients’ familial history was checked out and only seven (07) patients (10%) reported some familial case.

The great majority of patients were followed during the treatment, until their death - or they were alive when this study was finished. This way, Kaplan-Meier’s curve was used to evaluate the patients’ survival period, after the diagnosis. A patient’s average time of life, after the diagnosis, was of 64,116 months and median of 48 months, as shown on Graph 1.

Discussion

The average age found in our study was...
compatible with the average that had been reported in other studies. In the studies by Belsh and Schiffman, Zoccolella et al. and del Aguaiga et al. the average age reported was 53.3, 64.5 and 61.3 years old, respectively. In a review study, about the epidemiology of ALS, Beghi et al. reassured this information and he says that the start of this disease (ALS) before the age of 30 years old is rare, once it begins more commonly on a patient’s 5th and 6th decade of life. It demonstrates that ALS usually appears in middle aged and elderly people. In the present study, there was no difference in the way of start, signs and symptoms, average age and familial case, when the studied population was divided into two groups regarding the sex. The ratio among the sexes was of 2.88 men for 1 woman, what shows the predominance of ALS in men. But, Begghi et al., showed that this difference, as years pass by, is decreasing, and he reported studies from 1965-1974 in which the same ratio was 2.6:1, and more recent studies, 1995-1997 where this ratio was 1.3:1. The referred author also reports that due to women’s exposure to the same risk factors as men’s is due to women’s inclusion in the working market.

The symptom’s distal start was present in almost half of the patients; however, some patients showed a distal start and a proximal one as well. It was not possible to compare these data, because there were not available ones. In spite of it, Lee et al. showed in his study that 31% of the cases start in the upper members and 37% in the lower members. Zoccolella et al. found 35% for the upper members and 36% for the lower members. A bigger difference was described by Belsh and Schiffman with 52% in the upper members and 36% in the lower members. A bigger difference was described by Belsh and Schiffman with 52% in the upper members and 36% in the lower members. Although we did not evaluate the patients this way, in our results we found that more than one-fifth of our patients started to show the symptoms in the upper or lower members.

Ethnicity is another factor that seems to be important in the definition of ALS. Our group of patients had a great percentage of white people, when compared top grayish brown and, especially black people. Del Aguila et al., in his work with work with 180 patients, had 170 white people and 10 classified as non-white. Lee et al., in his group of patients had 90% of white people, 5% of Hispanic and black people only. We could say that this high prevalence in white patients might be due to the characteristics of the population of the countries were these pieces of work were done. However, although the region where our study was done, showed a very big black population, this same study showed that most of the patients who had ALS were white. So, this result can indicate that the white race has a bigger predisposition for ALS.

The familial history is present in around 5 to 10% according to literature. However, it might happen that a familial ALS be classified as sporadic, and a sporadic ALS be considered familial, and vice versa, because to affirm that ALS is familial, it depends on the diagnosis of the disease in more than one member of the family according to Mulder et al. In our study, we considered as a positive familial history when the patient had, at least, a relative with the same clinical picture. In spite of it, we found, in the medical literature, a very close percentage - according to Belsh and Schiffman familial history was present in 12.5% of the patients. In the work of Lee et al. familial history was positive for 5% of the patients, a little less than what we found, but in the percentage described in the literature. The signs and symptoms that were found in our work agree with all the ALS’s patients’ classical clinic. Zoccolella et al. in his work showed that fatigue was present in 71% of the cases, cramps in 16%, fasciculations in 13%, and dysarthria in 27% and dysphagia in 10% of the patients eligible for ALS. In our group, this percentage was bigger than the ones compared to the work of Zoccolella et al., due to the delay of the patient in searching for medical services - so, our patients were evaluated in a further stage of the disease. For this reason, a good part of the patients already showed almost all signs and symptoms of ALS - even the symptoms of the bulbar syndrome, as dysphagia and dysarthria. However, if we observe the start of the disease, we will notice that the great part of patients started, mainly, with signs and symptoms of the upper and lower motor neuron. In addition, the great majority of patients (between 97,1% to 98,6%), with the evolution of the disease, will show a lessening of the muscular power, miosfaciculations, muscular atrophy and motor deficit. Our results also show that dysarthria, dysphagia and difficulty in whistling, are less frequent characteris-

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**Graph 1:** Kaplan-Meier’s survival period’s curve.
vived for 10 years—they belonged to these exceptions, maybe deviating the average of survival period higher. However, the difference between literature and our study was not significant. (Table 2).

### Table 2: Types of start of disease, found syndromes and signs, familial cases. Comparison made between the sexes and the total number of patients

<table>
<thead>
<tr>
<th>Start</th>
<th>Male</th>
<th>Female</th>
<th>Total n</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>Distal</td>
<td>39 (76,5%)</td>
<td>12 (66,7%)</td>
<td>51 (72,9%)</td>
<td>0,4931</td>
</tr>
<tr>
<td>Proximal</td>
<td>21 (40,4%)</td>
<td>10 (55,6%)</td>
<td>31 (44,3%)</td>
<td>0,2640</td>
</tr>
<tr>
<td>Bulbar</td>
<td>8 (15,4%)</td>
<td>0 (0%)</td>
<td>8 (11,4%)</td>
<td>0,0770</td>
</tr>
<tr>
<td><strong>Syndromes</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>UMN (Upper Motor Neuron)</td>
<td>48 (92,3%)</td>
<td>15 (83,3%)</td>
<td>63 (90%)</td>
<td>0,2740</td>
</tr>
<tr>
<td>LMN (Lower motor neuron)</td>
<td>48 (92,3%)</td>
<td>16 (88,9%)</td>
<td>64 (91,4%)</td>
<td>0,6551</td>
</tr>
<tr>
<td>Bulbar</td>
<td>39 (75%)</td>
<td>12 (66,7%)</td>
<td>51 (72,9%)</td>
<td>0,4931</td>
</tr>
<tr>
<td><strong>Signs and Symptoms</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Muscular Fatigue</td>
<td>52 (100%)</td>
<td>17 (94,4%)</td>
<td>69 (98,6%)</td>
<td>0,0869</td>
</tr>
<tr>
<td>Microfasciculations</td>
<td>52 (100%)</td>
<td>17 (94,4%)</td>
<td>69 (98,6%)</td>
<td>0,0869</td>
</tr>
<tr>
<td>Muscular Atrophy</td>
<td>51 (98,1%)</td>
<td>18 (100%)</td>
<td>69 (98,6%)</td>
<td>0,5534</td>
</tr>
<tr>
<td>Motor Deficit</td>
<td>51 (98,1%)</td>
<td>17 (94,4%)</td>
<td>68 (97,1%)</td>
<td>0,4252</td>
</tr>
<tr>
<td>Difficulties in whistling and sucking</td>
<td>24 (46,2%)</td>
<td>10 (55,6%)</td>
<td>34 (46,6%)</td>
<td>0,4915</td>
</tr>
<tr>
<td>Dysphagia</td>
<td>30 (57,7%)</td>
<td>12 (66,7%)</td>
<td>42 (60%)</td>
<td>0,5029</td>
</tr>
<tr>
<td>Dysarthria</td>
<td>41 (78,8%)</td>
<td>12 (66,7%)</td>
<td>53 (75,7%)</td>
<td>0,2989</td>
</tr>
<tr>
<td>Hyporeflexia in the upper members</td>
<td>34 (65,4%)</td>
<td>8 (44,4%)</td>
<td>42 (60%)</td>
<td>0,1180</td>
</tr>
<tr>
<td>Hyporeflexia in the lower members</td>
<td>40 (76,9%)</td>
<td>11 (61,1%)</td>
<td>51 (73,9%)</td>
<td>0,1935</td>
</tr>
<tr>
<td>Mandibular Hyporeflexia</td>
<td>19 (36,5%)</td>
<td>5 (27,8%)</td>
<td>24 (34,3%)</td>
<td>0,4997</td>
</tr>
<tr>
<td>Hyporeflexia in the upper members</td>
<td>13 (25%)</td>
<td>8 (44,4%)</td>
<td>21 (30%)</td>
<td>0,1207</td>
</tr>
<tr>
<td>Hyporeflexia in the lower members</td>
<td>10 (19,2%)</td>
<td>7 (38,9%)</td>
<td>17 (24,3%)</td>
<td>0,0936</td>
</tr>
<tr>
<td>Hoffman’s Sign</td>
<td>21 (40,4%)</td>
<td>4 (22,2%)</td>
<td>25 (35,7%)</td>
<td>0,1657</td>
</tr>
<tr>
<td>Babinski’s Sign</td>
<td>42 (80,8%)</td>
<td>9 (50%)</td>
<td>51 (72,9%)</td>
<td>0,0114</td>
</tr>
<tr>
<td><strong>Familial Cases</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Present</td>
<td>5 (9,6%)</td>
<td>2 (11,1%)</td>
<td>7 (10%)</td>
<td>0,8553</td>
</tr>
</tbody>
</table>

### Reference


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Meningioma clinoideo

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Resumen

Los meningiomas clinoideos son tumores benignos originados en la leptomeninges que rodea el proceso clinoideo anterior, representando el 17% de los basales. Con el objetivo de caracterizarlos se estudiaron los pacientes operados en el Servicio de Neurocirugía del Hospital Hermanos Ameijeiras (2000 al 2010). La serie estuvo integrada por 10 pacientes, 9 mujeres y un hombre, la edad promedio fue de 49,8 años, clínicamente caracterizados por cefalea (10 pacientes) y déficit de la agudeza visual (9 pacientes). A todos se les practicó un abordaje frontotemporal-orbitozigomático (FTOZ) alcanzándose 7 resecciones totales y 3 subtotales, 3 pacientes sufrieron complicaciones, al alta ocho de los diez operados egresaron en excelente estado, no tuvimos recidiva ni crecimiento tumoral en los casos con resecciones parciales y al año de seguimiento la calidad de vida según la escala de Karnofsky fue superior o igual a 80 puntos en todos los casos. Concluimos que los meningiomas clinoideos por su asiento en el centro de la base craneal relacionados anatómicamente con estructuras neurovasculares críticas y por las grandes dimensiones que alcanzan en el momento de su diagnóstico, representan un desafío en la práctica neuroquirúrgica, comportándose en nuestra serie más frecuentes entre las mujeres con la cefalea y el déficit visual monocular como síntomas principales. Las técnicas de base de cráneo y en nuestras manos la craneotomía FTOZ complementadas con osteotomías basales de diferentes grados, han demostrado ser una alternativa eficaz para mejorar la suficiencia de la resección tumoral con mínima morbilidad, ausencia de recurrencias y mortalidad.

Abstract

Clinoidal meningiomas are benign tumors originated in the liptomeninge surrounding the anterior clinoidal process representing 17% of basal tumors. With the objective to characterize it, studied were patients operated in the neurosurgical service of Hermanos Ameijeiras hospital from 2000 to 2010. The series were composed of 10 patients, 9 female and a male, the mean age was 49.8 years and were clinically characterized with headache (10 patients) and visual deficit (9 patients). To all were practiced a frontotemporal-orbitozygomatic approach (FTOZ) reaching 7 total and 3 subtotal removal, 3 patients suffered complications, 8 out of 10 patients operated were discharged in good condition. There was no recurrence after total removal and after a year of follow up, the life quality following Karnofsky scale was superior or equal to 80 points in all the cases. In conclusion Clinoidal meningiomas due to its placement in the cranial base with anatomical relations with critical neurovascular structures and also the great dimension to reach in the moment of its diagnosis represents a challenge to neurosurgical practice. It comports in our series with more frequency within females presenting headache and mono ocular visual defect as the principal symptoms. Cranial base techniques and in our case frontotemporal-orbitozygomatic craniotomy complimented with basal osteotomy of different grades, had demonstrated to be an efficient alternative to better the technique of total removal with less mobility, zero recurrence and mortality.