

1st Latin American Symposium and 3rd Brazilian Symposium on Frontotemporal Lobar Degeneration

Belo Horizonte, Minas Gerais, Brazil
June 14-16, 2012

Frontotemporal Lobar Degeneration (FTLD) is the second most frequent cause of early-onset dementia, after Alzheimer's disease, and includes three major clinical subtypes, namely behavioral variant frontotemporal dementia (bvFTD), primary progressive aphasia (PPA) and semantic dementia (SD). These dementia syndromes share as a common feature the presence of focal atrophy affecting the anterior parts of the frontal and/or temporal lobes, presenting clinically as a progressive impairment in social function, personality changes and executive dysfunction, characterizing the bvFTD, or a decline in language abilities, which occurs in PPA and in SD.

Considerable research efforts have been made in the last years in order to disentangle the clinical, genetic and pathological basis of FTLD syndromes. These advances are crucial for the development of new diagnostic tools, which may allow early and better detection of these disorders, and will also help to build up and test new therapies for such devastating conditions.

This issue of *Dementia & Neuropsychologia* presents the abstracts of the 1st Latin American Symposium and 3rd Brazilian Symposium on FTLD, which was held in Belo Horizonte, Minas Gerais, Brazil, in June 14 to 16, 2012. Researchers from Argentina, Brazil, Chile, Colombia, Peru, Uruguay, and also investigators from France, have gathered together to present and discuss their works, at conferences, round tables and poster sessions.

The Symposium has been organized by the Cognitive Neurology and Ageing Department of the Brazilian Academy of Neurology, which has *Dementia & Neuropsychologia* as its official scientific journal.

We thank our colleagues from the scientific committee, the guest lecturers, as well as all investigators and participants for their significant contributions to the Symposium, which demonstrate the important role that researchers from our continent play in this field.

Ricardo Nitrini

Chair of the 1st Latin American Symposium on FTLD

Paulo Caramelli

Chair of the 3rd Brazilian Symposium on FTLD

PROGRAMAÇÃO CIENTÍFICA

14/06/2012 (Quinta-feira / Jueves)		
13h00	Recepção e entrega de materiais	
14h00-15h30	Mesa redonda: Introdução à Degeneração Lobar Frontotemporal (DLFT)	Coordenador: Nilton Santos Custodio Capuñay (Peru)
	Classificação clínica e epidemiologia da DLFT na América Latina	Nilton Santos Custodio Capuñay (Peru)
	Novos critérios diagnósticos de DLFT	Facundo Manes (Argentina)
	Correlações clínico-patológicas na DLFT	Valéria Santoro Bahia (Brasil)
15h30-16h30	Apresentação de pôsteres	
16h30-17h00	Intervalo	
17h00-18h30	Mesa redonda: Avaliação cognitiva e funcional na DLFT	Coordenadora: Letícia Lessa Mansur (Brasil)
	• Testes de rastreio cognitivo para a variante comportamental da DLFT	Rogério Gomes Beato (Brasil)
	• Novos testes neuropsicológicos para diagnóstico da variante comportamental da DLFT	Teresa Torralva (Argentina)
	• Funcionalidade na DLFT e sua relação com as alterações cognitivas	Letícia Lessa Mansur (Brasil)
19h30-19h45	Abertura	Facundo Manes (Argentina) Paulo Caramelli (Brasil) Ricardo Nitrini (Brasil)
19h45-20h30	Conferência	Coordenador: Ricardo Nitrini (Brasil)
	• Demências de início precoce: panorama geral	Florence Pasquier (França)
20h30 - 21h30	Coquetel	
15/06/2012 (Sexta-feira / Viernes)		
8h00-9h30	Mesa redonda: Neurobiologia da cognição social e do comportamento	Coordenador: Henrique Cerqueira Guimarães (Brasil)
	• Bases neurobiológicas da empatia/teoria da mente	Leonardo Ferreira Caixeta (Brasil)
	• Apatia e desinibição	Henrique Cerqueira Guimarães (Brasil)
	• Julgamento moral	Jorge Moll Neto (Brasil)
9h30-10h00	Conferência	Coordenador: Rogério Gomes Beato (Brasil)
	• Genética da DLFT	Julien Chapuis (França)
10h00-10h30	Intervalo	
10h30-12h00	Mesa redonda: Desafios no diagnóstico diferencial da DLFT	Coordenadora: Sonia Maria Dozzi Brucki (Brasil)
	• Diagnóstico diferencial entre DLFT e transtornos psiquiátricos	Andrea Slachevsky (Chile)
	• Cognição social contextual e a variante comportamental da DLFT	Facundo Manes (Argentina)
	• Diagnóstico diferencial entre variante comportamental da DLFT de início tardio e variante frontal da doença de Alzheimer	Sonia Maria Dozzi Brucki (Brasil)
12h00-13h30	Intervalo para almoço	
13h30-14h00	Conferência	Coordenador: Paulo Caramelli (Brasil)
	• Biomarcadores na DLFT	Luc Buée (França)
14h00-14h30	Conferência	Coordenadora: Valéria Santoro Bahia (Brasil)
	• Neuropatologia da DLFT	Vincent Deramecourt (França)

14h30-16h00	Mesa redonda: As múltiplas trajetórias clínicas da variante comportamental da DLFT	Coordenador: Luis Fontán Scheitler (Uruguai)
	• Estadiamento da variante comportamental da DLFT	Paulo Caramelli (Brasil)
	• Fenocópia ou formas não progressivas da variante comportamental da DLFT	Ricardo Nitri (Brasil)
	• Comparação do perfil neuropsicológico da deterioração cognitiva do transtorno bipolar e da variante comportamental da DLFT	Luis Fontán Scheitler (Uruguai)
16h00-16h30	Intervalo	
16h30-18h00	Mesa redonda: Neuroimagem - até onde pode auxiliar no diagnóstico?	Coordenadora: Andrea Slachevsky (Chile)
	• Padrões de atrofia à ressonância magnética nos diferentes subtipos clínicos de DLFT	Jorge Moll Neto (Brasil)
	• Alterações de substância branca na DLFT	Fernanda Tovar-Moll (Brasil)
	• Neuroimagem funcional: estado atual e perspectivas	Geraldo Busatto Filho (Brasil)
18h00-19h00	Apresentação e discussão de casos clínicos de DLFT (1ª parte)	Coordenadora: Thais Helena Machado (Brasil) Facundo Manes (Argentina) Ricardo Nitri (Brasil) Thais Helena Machado (Brasil)
16/06/2012 (Sábado)		
8h00-8h45	Conferência	Coordenadora: Thais Helena Machado (Brasil)
	• Afasia progressiva primária: atualização e relato de experiência de acompanhamento de 90 casos	Mirna Lie Hosogi-Senaha (Brasil)
8h45-10h15	Mesa redonda: Afasia progressiva primária: experiência latino-americana	Coordenador: Paulo Caramelli (Brasil)
	• Experiência argentina	Teresa Torralva (Argentina)
	• Experiência chilena	Andrea Slachevsky (Chile)
10h15-10h45	Intervalo	
10h45-12h00	Apresentação e discussão de casos clínicos de DLFT (2ª parte)	Coordenadora: Thais Helena Machado (Brasil) Paulo Caramelli (Brasil) Ricardo Nitri (Brasil) Thais Helena Machado (Brasil)
12h00-13h30	Intervalo para almoço	
13h30-14h30	Mesa redonda: O que é possível oferecer para o tratamento da DLFT?	Coordenadora: Teresa Torralva (Argentina)
	• Reabilitação na afasia progressiva primária	Maria Teresa Carthery-Goulart (Brasil)
	• Intervenções com cuidadores e familiares de pacientes com DLFT	Marcella Guimarães Assis Tirado (Brasil)
14h30-15h00	Conferência	Coordenador: Facundo Manes (Argentina)
	• Tratamento farmacológico da variante comportamental da DLFT: presente e futuro	Florence Pasquier (França)
15h00	Encerramento	

NEUROLINGUISTIC COMPARATIVE PROFILE OF ALZHEIMER'S DISEASE AND SPECTRUM OF FRONTOTEMPORAL LOBAR DEGENERATION

SOARES CD, SOARES VLD, CAIXETA L

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Background: The national survey of dementia and its implications for language, although in small numbers, line up with the international literature in the early trends of research.

Objective: The aim of this study is intended to draw a profile comparison between the differential Neurolinguistics of Alzheimer's Disease (AD) and Frontotemporal Lobar Degeneration (FTLD) spectrum.

Methods: The sample consisted of 60 participants; 16 Frontotemporal Dementia (FTD), 10 Primary Progressive Aphasia (PPA), 4 Semantic Dementia (SD) and 30 AD. We applied the Boston Diagnostic Aphasia Examination, Boston Naming Test, Semantic and Phonemic Fluency, Token Test, subtests of Vocabulary and Similarities/WAIS-R. To compare the performance of the groups, we used Mann-Whitney test.

Results: All groups showed substantial linguistic differences. The PPA group stood out from the other groups, especially when compared to AD, showing that fluency, vocabulary, abstraction of ideas, understanding, reading and writing are more impaired.

Conclusions: The FTLD group presented dysfunction in semantic and phonemic verbal fluency, SD group showed a statistically significant abstractive capacity with respect to the AD group. In FTLD oral expression was shown to be noticeably compromised compared to AD patients.

GENOTYPING OF APOE GENE IN PATIENTS WITH FRONTOTEMPORAL LOBAR DEGENERATION

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BAHIA VS, LIN CJ, NITRINI R

UNIVERSIDADE DE SÃO PAULO

Background: For over 20 years, the $\epsilon 4$ allele of the gene APOE has been reported as a risk factor for Alzheimer's disease (AD) and related to early onset of dementia associated with amyloid plaques. Some studies have suggested that $\epsilon 4$ might also be a risk factor in frontotemporal lobar degeneration (FTLD). A recent meta-analysis demonstrated an increased susceptibility in $\epsilon 2$ carriers to develop FTLD, however, the role for APOE alleles in FTLD is still unclear.

Objective: To investigate the presence of APOE alleles $\epsilon 2$ and $\epsilon 4$ in patients with FTLD (n=43) and AD (n=15) and compare this data with meta-analysis of Brazilian studies for controls (n=360).

Methods: FTLD and AD have been diagnosed by Neary criteria and NINCDS/ADRDA criteria, respectively. Genomic DNA was extracted from peripheral lymphocytes. APOE polymorphism genotyping was based on differential amplification by means of Real Time PCR.

Results: The presence of a copy of the $\epsilon 2$ and $\epsilon 4$ alleles were calculated in patients with FTLD ($\epsilon 2$ =23.3%; $\epsilon 4$ =25.6%), patients with AD ($\epsilon 2$ =6.7%; $\epsilon 4$ =40%) and controls ($\epsilon 2$ =11.1%; $\epsilon 4$ =23%), however, statistical differences (chi-square) were not still obtained due to small sample.

Conclusions: Preliminary data indicates that the presence of a copy of $\epsilon 2$ allele may have an implication in FTLD. Samples are still being collected. More cases of AD and FTLD will be genotyped and controls will be recruited for this study.

EXECUTIVE FUNCTIONS AND EPISODIC MEMORY AS PREDICTORS OF FUNCTIONAL PERFORMANCE IN MCI AND DEMENTIA

DE PAULA JJ, BERTOLA L, MALLOY-DINIZ LF

UNIVERSIDADE FEDERAL DE MINAS GERAIS

Background: Cognitive impairment and depressive symptoms are associated with worse functionality phenotype in elderly with dementia and Mild Cognitive Impairment (MCI).

Objective: To investigate the association between executive and memory impairment, and depressive symptoms, with functionality of these patients.

Methods: 113 patients with dementia (86 Alzheimer's-AD, 7 Frontotemporal, 5 Lewy-Bodies, 15 vascular) and 41 with MCI underwent neuropsychological examination covering various domains of episodic memory and executive functions and answered the Geriatric Depression Scale (GDS). Caregivers answered a scale of activities of daily living (DLAs). The cognitive variables were reduced through factor analysis to three components (executive functions, episodic memory and working memory) and, along with GDS, were used as predictors in a regression model.

Results: Only executive functions were related to functionality ($R^2=0.27$). Group analysis indicates that non-AD demented patients showed worse executive functions' performance ($p<0.01$) and functional status ($p<0.01$), but not depressive symptoms, episodic and working memory ($p>0.05$) than AD or MCI patients.

Discussion: These results suggest that executive functions are particularly relevant for functionality of these patients. Non-AD demented patients are especially impaired in the performance of DLAs due to executive dysfunction.

NAMING AND FLUENCY IN DIFFERENT CATEGORIES: A POSSIBLE DISSOCIATION IN A CASE OF SEMANTIC DEMENTIA

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Background: Semantic Dementia is characterized mainly by changes in semantic memory, as difficulties in naming, comprehension, categorization and production of spontaneous speech.

Objective: Compare the performance of one patient in different categories of naming tasks and fluency.

Methods: Female patient, 71 years, one year of schooling, with left frontotemporal atrophy, underwent tests as: Token Test (TT), Naming Test (NT), Semantic Verbal Fluency (SVF) and Phonemic (FVF). Additionally, performed an experimental battery of naming, consisting of six categories with 20 stimuli each (action, food, animals, body parts, objects and clothes). The categories were presented through line drawings, photos and real objects.

Results: The patient achieved a performance of more than 2SD below the mean in the TT, FVS, FVF and NT. In the experimental battery, her performance showed significant dissociation ($p < 0.001$) between body parts (16-18/20) and actions (11-9/20) when compared to animals (3-2/20), food (2-3/20), clothing (2-2/20) and objects (3-2/20).

Conclusions: The results suggest distinct impairments of different semantic categories in this patient. This is supported by literature, which suggests that the representation of body and actions are located in the parietal lobe and basal ganglia, unlike other categories evaluated that are represented predominantly on the temporal lobe.

PREVALENCE OF FRONTOTEMPORAL LOBAR DEGENERATION AMONG 460 OUTPATIENTS IN CENTRAL BRAZIL

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Background: Compared with other major dementias, very little is known about the prevalence and risk factors of FTLD. Recent improvement in FTLD diagnostic assessment suggests that it is more common than previously noticed.

Objective: The aims of this study were to determine the prevalence of FTLD in a Brazilian sample of outpatients consecutively attended in an academic medical center, as well as search for probable risk factors.

Methods: Four hundred sixty consecutive dementia

outpatients were investigated using clinical, neuroimaging (MRI and SPECT in all) and neuropsychological methods. We adopted the diagnostic criteria recommended by Neary et al. for FTLD and standard diagnostic criteria for other dementias.

Results: Four hundred and ten were included at sample. The prevalence of FTLD in overall sample was 17.5%, among these 25.9% were younger than 65 years and 12.1% were older than 65 years. The sex ratio (M:F) was 31/41 and the mean of age was 66.93 ± 10.8 . The risk factors associated with FTLD more common were hypertension, family history and history of depressive symptoms.

Conclusions: FTLD represented the second most common dementia in all demented patients, being only surpassed by Alzheimer's disease. The estimate of DLFT prevalence was higher than previously reported in Brazilian studies. Further studies are needed to confirm this rate.

PHENOMENOLOGICAL STUDY OF REALISTIC PICTURE COPY IN A CASE OF ADVANCED SEMANTIC DEMENTIA

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Background: Semantic dementia (SD) is a clinical form of FTLD, with verbal and nonverbal semantic disorder. Patients have shown preserved artistic skills even after disease onset. Miller et al. (1998, 2000 and 2006) reported the appearance of visual representations despite the cognitive deterioration.

Objective: To describe the preservation of picture copying abilities in severe DS.

Methods: 81 year old male with advanced SD, with assessment through the battery LOTCA e DAD with severe loss of verbal and visual semantic memory, associative visual agnosia for objects, with perception of shapes, colors, repetitive patterns, the concept of symmetry in the expressions and semi-dependent for self-care with compulsion eating, apathy and isolation.

Results: [1] Realistic copy: does not interact with the semantic content of the figure, only the formal elements (line, direction, contours, and colors) with visuospatial skills preserved. [2] The patient paints only in enclosed spaces in an impulsive way, with sense of color distribution and action automatic.

Conclusions: Artistic activities can be tools for intervention in advanced stages, with productions that generate well-being, self-esteem and social recognition, supporting the personality. The assessment of artistic production in dementia may be a valuable useful resource in the evaluation of residual cognitive processing.

NONLITERAL LANGUAGE AND THEORY OF MIND IN FRONTOTEMPORAL DEMENTIA: PRELIMINARY RESULTS

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Background: Nonliteral language (NLL) is the ability to think about their own language and includes the abilities of metaphor and proverb interpretation and irony recognition. Theory of Mind (ToM) is the ability to infer the contents of other people's minds, including beliefs and intentions. Patients with Frontotemporal dementia (FTD) appear to have difficulties in these abilities due to the alterations of language and executive functions.

Objective: Investigate a possible impairment in NLL and ToM in patients with FTD and their relationship with frontal symptoms.

Methods: Thirteen patients with FTD were evaluated using the Test of Language Competence – Figurative Language, Irony Test, Screening Test for Alzheimer's disease with Proverbs and Happé's Test. Qualitative evaluation was done through the frontal symptoms present during the testing.

Results: All patients presented low performance in all tests. Patients did not use the abstraction to proverb interpretation. Concrete thought was observed in metaphor comprehension as well. In irony test most patients failed in recognizing ironic sentences. Patients failed to infer mental states of characters. The most prevalent frontal symptoms were economy of effort, concrete thought and perseveration.

Conclusions: Evidences show that patients with FTD have major impairment in NLL and ToM, especially because of the frontal symptoms and language alterations.

COGNITIVE PERFORMANCE AND FUNCTIONAL PROFILE ARE ASSOCIATED IN BEHAVIORAL VARIANT FRONTOTEMPORAL DEMENTIA (bvFTD)

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Background: There is limited information regarding the functional profile of individuals diagnosed with behavioral variant frontotemporal dementia (bvFTD).

Objective: To examine the association between cognitive and functional performance in bvFTD.

Methods: The sample consisted of 15 patients diagnosed with bvFTD (mean age= 1.0±9.6 years, mean schooling=10.9±6.18 years, mean time of diagnosis=17.1 months). The exclusion criteria were: advanced dementia, other causes of dementia other than bvFTD and presence of psychiatric illnesses. The protocol in-

cluded the Mini-Mental State Examination (MMSE), the Addenbrooke's Cognitive Examination-Revised (ACE-R), the Executive Interview (EXIT-25) and the Direct Assessment of Functional Status-Revised (DAFS-R).

Results: Results indicated a robust association between cognitive and functional performance (correlations DAFS-R and MMSE=0.89, $p<0.001$, DAFS-R and ACE-R=0.89, $p<0.001$, DAFS-R and EXIT-25= -0.91, $p<0.001$). Functional performance assessed objectively was significantly associated with all aspects of cognition assessed by the ACE-R.

Conclusions: Results suggest functional impairment in bvFTD is highly associated with the cognitive deficits present in the disease.

AMYOTROPHIC LATERAL SCLEROSIS AND DEMENTIA: REPORT OF 3 CASES WITH ONSET AFTER THE AGE OF 65

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Background: Dementia occurs in approximately 14% of patients with Amyotrophic Lateral Sclerosis (ALS), frontotemporal dementia being the most common etiology.

Objective: To report three cases of dementia and ALS that started after the age of 65.

Methods: Cases report.

Results: Case 1 – A 70-year-old man was seen with an one-year-history of social isolation and loss of performance in their work. MMSE: 25/30; FAB 9/18. MRI frontal atrophy. After one year of follow-up, fasciculations in upper limbs and weakness were observed. Electroneuromyography (ENMG) suggestive of ALS. Case 2 – A 79-year-old woman presented with a 6-year-history of walking difficulty and urinary incontinence, which were treated in another hospital as normal pressure hydrocephalus without improvement. In the last six months, her gait deteriorated even more and she developed dysphagia, anarthria, applause sign, tetraparesis with brisk reflexes and fasciculations. CT revealed hydrocephalus and mild frontal atrophy. ENMG: suggestive of ALS. Case 3 – A 84-year-old man presented with a 5-year-history of amnesia for recent events, more severe in the last 2 years, which was associated with dyspnea in the last year. MMSE: 17/30; proximal tetraparesis with fasciculations in the four members and hyperreflexia. MR showed atrophy of hippocampus. ENMG: suggestive of ALS.

Conclusions: We call attention to the association of ALS with dementia, which can occur also in older individuals, including cases with amnesic initial complain.

CLINICAL PROFILE ON FIRST-TIME EVALUATION OF BEHAVIORAL VARIANT FRONTOTEMPORAL DEMENTIA (bvFTD) PATIENTS IN A SPECIALIZED TERTIARY OUTPATIENT UNIT: PRELIMINARY FINDINGS

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DE MATOS VP, CARVALHO VA, MACHADO TH,
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Introduction: The bvFTD is a clinical syndrome characterized by marked personality changes, social inadequacy and cognitive impairment. Despite recent advances in the characterization of its phenotype, patients with this disorder usually receive an accurate diagnosis only lately on disease course.

Objective: To describe the clinical profile on the first-time evaluation of 18 patients with bvFTD.

Methods: We selected 18 patients from our tertiary care outpatient specialized unit and reviewed their medical charts on admission sheets. All patients fulfilled the international consensus criteria for probable or possible bvFTD.

Results: Patients' mean age was 67.6 years; 11 of them were men. The mean number of years of schooling was of 6.5 years. The mean age at onset was 59.9 years. Seven patients (38.9%) presented with initial behavioral complaints only, seven patients presented with memory and behavioral complaints, two patients had only with memory disturbances and one patient presented with memory, behavioral and language problems. The mean Mini-Mental State Examination score was 16.7 points. The median score on the Pfeffer Functional Assessment Questionnaire was 22 points.

Conclusions: When first time evaluated in a specialized unit, most of the patients displayed moderate impairment on cognitive screening tests and activities of daily living measures. They presented with both memory and behavioral complaints in combination or alone in the majority of cases.

NEUROPSYCHIATRIC SYMPTOMS EVALUATED IN FRONTOTEMPORAL DEMENTIA PATIENTS

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Background: Behavioral symptoms are frequently observed in dementia patients and are associated to highest distress for patients and caregivers.

Objective: The objective was to evaluate the prevalence, severity and distress of neuropsychiatric symptoms in a sample of frontotemporal dementia patients.

Methods: Nine frontotemporal dementia patients

(six male and three female) and their caregivers were consecutively included in the study by the Dementia Outpatient clinic of a University Hospital. The Neuropsychiatric Inventory (NPI) was applied to evaluate behavioral symptoms and their impact upon caregivers.

Results: The patients had a mean of 60 years (± 6) of age when beginning the evaluation in the Dementia Outpatient clinic, but symptoms started a mean of 5 years (± 2) before. Apathy and agitation were the most prevalent symptoms (55%) Apathy was responsible for the highest severity, and agitation for the highest caregiver distress level in this sample.

Conclusions: In the present study, agitation was the most distressing symptom. Comparing with our previous study with Alzheimer's disease patients (Godinho et al., 2008), apathy remained one of the most prevalent and severe symptom, and then independent of the type of dementia. These findings emphasize the need for prevention and treatment of these symptoms.

CLINICAL AND SOCIODEMOGRAPHIC DIFFERENCES IN EARLY- AND LATE-ONSET FRONTOTEMPORAL LOBAR DEGENERATION

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Background: The different clinical and sociodemographic patterns related to age of disease onset in FTLD have been fairly studied.

Objective: To compare clinical and sociodemographic characteristics in patients with early-onset and late-onset FTLD.

Methods: There were 72 consecutive patients with FTLD (according Neary et al., criteria), 42 had early-onset dementia, 29 with behavioural variant frontotemporal dementia (bvFTD), 10 with progressive nonfluent aphasia (PNFA), 3 with semantic dementia (SMD) and 30 had late-onset dementia (age of onset ≥ 65), 23 with bvFTD and 7 with PNFA. Studied variables were sex, education, duration from onset to consultation, ethnicity, socioeconomic level, marital status, Clinical Dementia Ratio (CDR) scores, Mini-Mental State Examination (MMSE) scores, Pfeffer Functional Activities Questionnaire scores and presence of some factor of risk.

Results: There were no significant differences in sex ratio, duration from onset to consultation, ethnicity, socioeconomic level, marital status, CDR scores and MMSE scores. There were significant differences on Pfeffer questionnaire, education and factor of risk as hypertension, depression and cardiovascular disease.

Conclusions: Late-onset FTLD patients may have more notorious functional decline, hypertension, car-

diovascular disease and less depression than early-onset FTLD. Our findings suggest there is clinical heterogeneity explained by age at disease onset in FTLD.

ROSS-CULTURAL ADAPTATION OF THE INECO FRONTAL SCREENING: FINAL TRANSLATION

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Background: The INECO Frontal Screening is a sensitive and specific tool for assessment of executive functions in dementia. It is an easy and fast screening application, consisting of eight subtests: motor programming, conflicted instruction, go-no go, digits backward, verbal and spatial working memory, proverbs interpretation and verbal inhibitory control, with a total of 30 points.

Objectives: To present the Brazilian final version and preliminary results with Frontotemporal Lobar Degeneration (FTLD) patients.

Methods: Translation and back-translation, and a final version after a pilot study. Participants were 12 patients with FTLD (mean age=59.58±8.62).

Results: We used items of Hayling Test (brazilian version), and we opted to brazilian proverbs presented in increasing level of complexity. The FTLD group had an average of 11.62±7.37 points in the full scale. The means of subtests were: motor programming=1.58±1.16; conflicted instruction=1.75±1.29; go-no go=0.5±1.0; digit backward=2.08±0.9; verbal working memory=1.0±0.95; spatial working memory=2.08±1.0; proverbs interpretation=0.62±1.05 and verbal inhibition control=2.0±1.17.

Conclusions: Preliminary results corroborate the findings of the original study concerning frontotemporal dementia patients (15.6±4.2) and indicated the need for better proverbs semantic adaptation. The final translation is completed.

POSTERIOR PICKS DISEASE WITH POSTERIOR CORTICAL ATROPHY PHENOCOPY: A CLINICO-PATHOLOGICAL CASE STUDY

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Background: Atypical presentations of Pick's disease do not include significant occipital atrophy.

Objective: Present a case considered to represent a rare case of Pick's disease of the posterior type, presenting as a syndrome of progressive posterior cortical dysfunction (PPCD). The disease progressed to more anterior, frontotemporal, areas.

Methods: Case report of a rare presentation of posterior Pick's disease with pathological confirmation. MRI,

SPECT, neuropsychological and autopsy evaluations are presented.

Results: 56yo patient, initially presented with higher order visuospatial dysfunction, Balint and Gerstmann syndromes, environmental agnosia, transcortical sensory aphasia and a rapid dementia. Then emerged severe behavior alterations. CT and MRI revealed predominant atrophy of the posterior cortex with isolated posterior ventricular enlargement, similar to PPCD. Posterior fibrillary gliosis resembled to "progressive subcortical gliosis". Brain weight was 750g. Severe loss of nerve cells was observed in occipital, frontal lobes, insula, gyrus cinguli, head of caudate nucleus and partial areas of temporal lobes; Pick cells and Pick bodies were found. Mild loss was observed in the parietal lobes. Primary motor and sensitive cortex were almost spared.

Conclusions: Our case expands the list of etiologic possibilities that may be associated with PPCD. A posterior variant of Pick's disease is reported.

SWALLOWING IN FRONTOTEMPORAL DEMENTIA: COGNITIVE AND BEHAVIORAL ASPECTS

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¹ UNIVERSIDADE FEDERAL DE SÃO PAULO, ² UNIVERSIDADE DE SÃO PAULO

Background: Swallowing in frontotemporal dementia (FTD) can be influenced by cognitive and behavioral aspects.

Objectives: To investigate swallowing difficulties in mild, moderate and severe stages of FTD and its correlates with cognitive and behavioral aspects.

Methods: Difficulty in swallowing was evaluated in 14 FTD patients by the Assessment of Feeding and Swallowing Difficulties in Dementia (AFSDD), that was completed by caregivers. To assess the stage of dementia, we used the Clinical Dementia Rating (CDR).

Results: Of 5 patients in the mild stage, 40% were slowed, 20% were distracted during feeding and 100% had chewing problems. Of 3 patients in the moderate stage, 66.6% had binge eating and a preference for sweets, 66.6% also had chewing problems and cough, and 100% ate very fast and stirred for food. Of 6 patients in the severe stage, 66.6% of patients were slowed, 84% were passive, 50% were distracted in a feeding situation, and 84% had chewing problems and cough.

Conclusions: In moderate stage all patients with chewing problems and cough were quick and stirred for feeding, while in mild and severe stages most patients with chewing problems and cough during feeding were slowed. Better knowledge of swallowing and its aspects is important to guide caregivers and to promote actions to maintain an oral feeding safer.

APATHY SYMPTOMS IN THE BEHAVIORAL VARIANT OF FRONTOTEMPORAL DEMENTIA: PREVALENCE AND NEUROPSYCHOLOGICAL CORRELATES

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Background: Apathy is a typical pathological manifestation associated with neurodegeneration of the frontal lobes. Previous studies have shown a high prevalence of apathy symptoms in bvFTD. However, little is known about the clinical and neurocognitive characteristics of such symptoms.

Objectives: To assess the presence of apathy symptoms in patients with behavioral variant Frontotemporal Dementia (bvFTD) and analyze their relation with neuropsychological performance.

Methods: Patients with bvFTD (n=18) were studied using the Apathy Scale [Starkstein et al., 2006] and the Apathy subdomain of the Frontal Systems Behavior Scale (FrSBe). In both cases, the patients' relatives completed the questionnaires. Patients were assessed with a comprehensive neuropsychological battery in order to assess basic cognitive functioning, and more specifically, executive functions. Patients with Alzheimer disease (AD, n=8) were assessed with the same tools as a control group.

Results: In accordance with previous findings, a high number of bvFTD patients showed clinically significant apathy symptoms (72.2%). Both apathy scales showed a significant correlation between each other ($r=0.488$; $p=0.040$). Apathy symptoms within the bvFTD group were significantly higher after onset of the disease relative to premorbid scores. BvFTD patients had significantly higher apathy scores than AD patients ($U=36.00$, $p=0.045$). Significant correlations were found with neuropsychological measures.

Conclusions: Apathy is a significant symptomatic domain in bvFTD associated with impaired cognitive and executive performance.

COMPARING THE CLINICAL USEFULNESS OF THE INSTITUTE OF COGNITIVE NEUROLOGY (INECO) FRONTAL SCREENING (IFS) AND THE FRONTAL ASSESSMENT BATTERY (FAB) IN AMIOTROPHIC LATERAL SCLEROSIS (ALS)

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Background: The IFS (Torralva et al., 2009) and the FAB (Dubois et al., 2000) are brief screening measures of executive functions. Both instruments showed to be

useful to detect executive impairments in patients with ELA (Oskarsson et al., 2010; Sierra et al., 2011). However, to our knowledge no study compared the specificity and sensitivity of both measures.

Objective: The aim of this study is to examine which measure is better to detect executive impairments in the early stages of ELA.

Methods: 32 persons participate in this study: 13 who meet Escorial criteria of Amiotrophic Lateral Sclerosis (ALS) in the early stages of their disease and 19 age and education matched controls (CTR). All participants were assessed with a standar cognitive evaluation wich include attention (Trail Making Test -A); memory (Rey Auditorial Verbal Learning Test); language (Boston naming test, semantic verbal fluency); visuospatial ability (Rey Complex Figure); executive functions (Trail Making Test - B, Wisconsin Card Sorting Test; phonologic verbal fluency) and the IFS and FAB.

Results: Relative to the FAB, the IFS showed (a) better capability to differentiate between ELA and controls; (b) higher sensitivity and specificity for the detection of executive dysfunction; (c) stronger correlations with standard executive tasks.

Conclusions: We conclude that while both tools are brief and specific for the detection of early executive dysfunction, the IFS is more sensitive and specific in differentiating ELA from controls, and its use in everyday clinical practice can contribute to identified the ELA's executive dysfunction.

COMPARISON OF COGNITIVE FUNCTIONING IN PATIENTS WITH PARKINSON DISEASE AND FRONTOTEMPORAL DEMENTIA BEHAVIORAL VARIANT

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Background: The executive dysfunction found in early PD has mainly been attributed to a disruption of the cortico-striatal loops that connect the basal ganglia and frontal-lobes. However, it is completely unclear whether these executive deficits reflect PD cortical (frontal) or subcortical (striatal) pathology. A possible approach to this problem is to compare performance in frontal and executive tasks between patients with early PD and a group of patients with predominantly fronto-cortical pathology, such as the behavioral variant of fronto-temporal dementia.

Objective: The objective of this study is to determine whether there are significant differences in performance between a group of patients with Parkinson's Disease

(PD) and one with the behavioral variant of Frontotemporal Dementia (bvFTD) in cognitive tasks that have been previously associated with the frontal lobes.

Design/Methods: A group of patients with PD (n=33) was compared to a group of patients with bvFTD (n=35) in a battery composed of frontal lobe-dependent functions.

Results: Patients with bvFDT performed significantly worse than patients with PD in tests of cognitive flexibility (p=0.01), set shifting (Trail B p<0.01), Theory of the Mind (p<0.01), Multitasking (p<0.01) and decision making (p<0.01).

Conclusions: Even though both groups of patients showed deficits in executive and frontal tasks, cognitive performance differed, with bvFTD patients showing a significantly lower performance than patients with PD.

DEVELOPING A BEHAVIORAL INVENTORY FOR THE DIAGNOSIS OF THE BEHAVIORAL VARIANT FRONTOTEMPORAL DEMENTIA

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Introduction and Objective: Along with the introduction of the new diagnostic criteria for the behavioral variant frontotemporal dementia (bvFTD) has emerged a need for systematic assessment of cognitive and behavioral changes in this patient population. Brief and psychometrically effective questionnaires aimed at this goal can contribute to this objective.

Methods: We manually selected individual items that corresponded to each of the new diagnostic criteria for bvFTD from a pool of 230 items, drawn from eight questionnaires widely used in the field of dementia. For each criterion, stepwise discriminant analysis was performed on a sample of 100 patients, of which 42 were bvFTD and 68 had been diagnosed with other non-behavioral variants of FTD. Items with the highest discriminatory power were pooled together to create a questionnaire.

Results: Twenty items were obtained whose internal consistency was 0.83 (Cronbach's alpha). A high canonical correlation (0.80) was obtained, which explained 64.4% of variability in linear regression and was significantly different between patients with bvFTD and other dementias [F(20.79)=7.1, p<0.0001]. ROC curve for the total score showed high discriminatory accuracy (AuC=0.79, p<0.001). Scores higher than 8/20 showed a sensitivity to detect bvFTD of 83.7% and a specificity of 84.1%.

Conclusions: The 20-item questionnaire hereby introduced has the potential to become a useful, brief, sensitive and specific tool to detect patients with bvFTD according to the new diagnostic consensus criteria.

DIFFERENT KINDS OF IMPULSIVITY IN PATIENTS WITH BEHAVIORAL VARIANT FRONTOTEMPORAL DEMENTIA (BVFTD)

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Background: Behavioral disorders constitute a major clinical feature of FTD. Patients frequently exhibit a wide array of behavioral disturbances such as inflexibility, disinhibition, impulsivity and perseverative behavior. These behavioral abnormalities are associated with cognitive deficits involving executive functioning. We hypothesized that orbitofrontal dysfunction provides a unifying framework to explain the origin of similar behavioral/cognitive symptoms.

Objective: The objective of the present study was to explore cognitive / behavioral measures of impulsivity in patients with bvFTD including pencil-and-paper tasks, computational tasks, and behavioral measures.

Methods: Patients with established diagnosis of bvFTD (n=15) and mild Alzheimer's disease (AD; n=15), were assessed with cognitive tests and behavioral questionnaires aimed at tapping on different aspects of impulsivity.

Results: Compared to AD, bvFTD patients showed significantly lower scores on the motor/ verbal inhibitory control subtests of the INECO Frontal Screening (IFS), more perseverative errors in the WCST, a worse performance on the IOWA Gambling Task, and more commission errors in a control inhibition test. In behavioral questionnaires, they presented elevated scores in the subscales of disinhibition and dysexecutive, which were strongly correlated with performance on executive tests. The Eating Behavior Questionnaire correlated only to the verbal inhibitory control subtest.

Conclusions: There seems to be a shared underlying basis for the cognitive and behavioral measures of impulsivity.

INTELLIGENCE AND EXECUTIVE FUNCTIONS IN FRONTOTEMPORAL DEMENTIA

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Background and Objective: Recently, we used the re-

relationship with general intelligence (Spearman's g) to define two sets of frontal lobe or "executive" tests. For one group, including Wisconsin Card Sorting and Verbal Fluency, reduction in g entirely explained the deficits found in frontal patients. For another group, including tests of social cognition and multitasking, frontal deficits remained even after correction for g . In the present study we develop this distinction in the context of behavioural-variant frontotemporal dementia (bvFTD), a disorder in which different executive tests decline at different stages of the disease: while tests of social cognition and multitasking decline from the early stage of the disease, classical executive tests, such as Wisconsin Card Sorting and Verbal Fluency, show deficits only later on.

Methods: We assessed 35 patients with bvFTD and 14 control subjects with a range of frontal tests. A measure of g was obtained from a general test battery. Statistical analysis asked which deficits in frontal tests remain, after correction for g .

Results: We show that, while deficits in the classical executive tests are entirely explained by g , deficits in social cognition, multitasking and decision-making under uncertainty are not.

Conclusions: The results suggest an early phase of relatively selective deficit, likely linked to atrophy in ventromedial and anterior frontal cortex, followed by more widespread cognitive decline as the disease spreads to other frontal regions.

PERSONALITY IN FTD

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Background: Changes in personality constitute some of the early signs of bvFTD. While these changes in personality have been previously studied in Alzheimer's disease (AD) and psychiatric disorders, systematic research in bvFTD patients regarding this issue is scarce.

Objective: To study personality changes in patients diagnosed with behavioral variant of Frontotemporal Dementia (bvFTD).

Methods: Relatives of sixteen patients with bvFTD and nineteen patients with AD completed the NEO-PI-R questionnaire both for personality traits before the onset of the disease and current personality. The NEO-PI-R domains were compared across patient groups, as well as the different facets involved in each personality domain.

Results: There was a significant difference between

previous and current personality in bvFTD patients for the extraversion domain ($p=0.024$; significant facets: assertiveness, activity, emotion-seeking), openness ($p=0.013$; ideas), and conscientiousness ($p<0.001$; competence, dutifulness, self-discipline). AD patients showed significant differences on neuroticism ($p<0.001$; anxiety, hostility, depression, impulsiveness and vulnerability), conscientiousness ($p<0.001$; all facets) and extraversion ($p<0.01$; assertiveness, activity).

Conclusions: The pattern of personality changes in bvFTD differed from the observed in AD patients. This study reveals the importance of characterizing personality changes in bvFTD patients with solid tools such as the NEO-PI-R in order to contribute to the early diagnosis.

CHARACTERIZATION OF THE COGNITIVE AND FUNCTIONAL PERFORMANCE OF PATIENTS WITH BEHAVIORAL VARIANT FRONTOTEMPORAL DEMENTIA (BVFTD)

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Background: There are few studies describing the functional changes in behavioral variant frontotemporal dementia (bvFTD) and it is not clear which aspects of functionality are affected.

Objective: To characterize the functional profile of patients diagnosed with bvFTD and to compare it to the functional profile of elderly patients with Alzheimer's disease (AD) and normal controls (NC) described in previous studies.

Methods: 15 patients diagnosed with bvFTD (mean age=61.0±9.6 years, mean schooling=10.9±6.18 years, mean time of diagnosis=17.1 months). The protocol included the Mini-Mental State Examination (MMSE), the Addenbrooke's Cognitive Examination-Revised (ACE-R), the Executive Interview (EXIT-25), the Direct Assessment of Functional Status-Revised (DAFS-R).

Results: Compared to previous studies, bvFTD patients were younger, yet, performed worse than NC but better than AD patients in the MMSE, DAFS-R total score, DAFS-R Time Orientation, Communication Skills and Dealing with Finances. Correlational analyses revealed a strong association between time of diagnosis and cognitive and functional performance.

Conclusions: The functional impairment observed in patients with bvFTD may be attributed to executive dysfunction whereas functional tasks which rely on episodic memory may be intact.

SCREENING FOR MUTATIONS IN *TARDBP* GENE IN PATIENTS WITH FRONTOTEMPORAL LOBAR DEGENERATION AND/OR AMYOTROPHIC LATERAL SCLEROSIS

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Background: TAR DNA-binding protein 43 (TDP-43) is a highly conserved 43kDa protein of 414 amino-acids, encoded by the *TARDBP* gene (6 exons) on chromosome 1p36.2. TDP-43 is involved in regulation of gene expression and splicing, normally localized in the nucleus, but in pathological conditions, its cleaved form is mainly present in the cytoplasm. Ubiquitinated TDP-43 is one of the major abnormal proteins found in frontotemporal lobar degeneration (FTLD) and amyotrophic lateral sclerosis (ALS). Recent genetic studies have identified mutations in *TARDBP* in patients with familial and sporadic ALS and in patients with FTLD with and without ALS.

Objective: To investigate the presence of *TARDBP* mutations in a cohort of FTLD and ALS.

Methods: Neary et al. and El Escorial criteria were used for the diagnosis of FTLD and ALS, respectively. Genomic DNA was extracted from peripheral lymphocytes. Briefly, extracted DNA was amplified (PCR) and direct nucleotide sequencing was performed using an ABI 3130xl Genetic Analyzer.

Results: Up to date, we have sequenced 43 patients with FTLD (bvFTD, n=30; SD, n=5; PNFA, n=5; FTLD-ALS, n=3) and 101 patients with ALS. No mutations have been identified in both FTLD and ALS cases.

Conclusions: Patients are still being recruited and more cases of FTLD and ALS will be analyzed for the presence of mutations in the gene *TARDBP*.

FRONTOTEMPORAL DEMENTIA DUE TO *C9orf72* AND *GRN* MUTATIONS: REPORT OF TWO BRAZILIAN KINDREDS

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Background: Around 15% of patients with frontotemporal dementia (FTD) spectrum disorders have a family history with an autosomal dominant pattern of inheritance. The main FTD-causing genes are *MAPT*, *GRN* and *C9orf72*.

Objective: To describe 2 Brazilian FTD kindreds.

Methods: We collected clinical data from chart review. Testing for *GRN* mutations was performed through exon amplification by PCR and direct sequencing. Presence of GGGGCC expansion in *C9orf72* was confirmed by repeat-primed PCR and Southern blot.

Results: Kindred 1: The *C9orf72* repeat expansion was found in 4 members of this family. Age of onset of symptoms was highly variable, ranging from 38 to 59 years. A few patients developed subtle behavioral changes years or decades before the diagnosis of dementia. The phenotypes observed were behavioral variant FTD and FTD-motor neuron disease. Kindred 2: A *GRN* mutation (Q300X) was found in the proband of this family, who developed symptoms at age 63. Her clinical presentation was consistent with a diagnosis of progressive non-fluent aphasia (PNFA), later evolving into a corticobasal syndrome. Her father and all of her 7 siblings developed symptoms, with significant variation in clinical presentation.

Conclusions: *GRN* and *C9orf72* mutations are present in Brazilian kindreds and further research is necessary to assess the frequency of these mutations in our population.

NEUROPSYCHOLOGICAL EVALUATION IN THE DIFFERENTIAL DIAGNOSIS OF DEMENTIA

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Introduction: Neuropsychological evaluation is useful for the syndromic diagnosis and to address the differentiation between the countless types of dementia.

Objective: To draw the characteristic neuropsychological of patients with dementia, in order to investigate which elements allow adequate clinical differentiation.

Methods: Data from 60 patients were retrospectively evaluated. The sample was divided in three groups: Alzheimer's disease (AD), Frontotemporal dementia (FTD) and pre-senile subcortical dementia (SD). All individuals were submitted to neuropsychological evaluation. The results were transcribed in tables and quantified.

Results: Significant differences emerged between groups, with AD patients presenting memory deficits initially. FTD patients, besides conduct changes, presented deficits in executive functions in the initial phas-

es. SD was characterized by expressive attention deficits in the initial phases of the disease and, in agreement with the topography of the lesion, other deficits varied, regardless of duration of the disease.

Conclusions: Neuropsychological evaluation is an effective instrument to differentiate various types of dementia, allowing anatomical and functional correlations, with diagnostic implications.

COMPARISON OF FRONTOTEMPORAL DEMENTIA AND ALZHEIMER'S DISEASE PATIENTS ON THE ADDENBROOKE'S COGNITIVE EXAMINATION-REVISED (ACE-R) AND ON THE VLOM RATIO

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Background: The ACE-R allows a brief evaluation of five cognitive domains, being particularly useful in differentiating Alzheimer's disease (AD) from frontotemporal dementia (FTD). Additionally, the battery also offers the VLOM subscore that comprises the ratio of

the scores of verbal fluency plus language to orientation plus name and address delayed recall memory (V+L)/(O+M).

Objective: To compare the performance of patients with mild FTD, mild AD and healthy controls in the Brazilian version of the ACE-R and its VLOM ratio.

Methods: The test was administered to 26 FTD patients, 25 probable AD subjects and to 58 cognitively healthy individuals. The groups were compared and cut-off points for ACE-R total score and VLOM ratio were extracted by investigation their accuracy using ROC curve analysis.

Results: The cut-off score (Sensitivity/Specificity) for differentiating AD from FTD in the ACE-R was >76 (40%/96%) and for VLOM Ratio ≤ 2.61 (66%/84%), respectively. In the same order, for FTD vs. controls they were ≤ 79 (70%/82%) and ≤ 2.12 (44%/89%). For AD vs. controls, ACE-R score was ≤ 77 (100%/89%) and VLOM ratio was >2.83 (74%/88%).

Conclusions: These preliminary results suggest that the Brazilian version of the ACE-R displayed good accuracy in discriminating FTD and AD patients, although further investigation is still necessary