Analysis of the linguistic profile in down syndrome using the arizona battery for communication disorders of dementia – a pilot study

ABSTRACT

Purpose: To characterize the linguistic profile of adults and elderly with Down Syndrome (DS) using the Arizona Battery for Communication Disorders of Dementia (ABCD). Methods: Thirty adult individuals with DS were evaluated through the MoCA cognitive battery, four functional scales (Pfeffer, Lawton-IADL, Katz-IADL and IQCODE) and the ABCD battery, which evaluates Mental State, Episodic Memory, Linguistic Expression, Linguistic Comprehension and Visuospatial Construction. The scores obtained by the individuals in the ABCD were correlated to those obtained on the Lawton-IADL scale. Results: Individuals with DS had significantly lower performance than cognitively normal adults and elderly as described in Brazilian studies. Due to the lack of similar studies in our country, we compared our results to those of elderly with Alzheimer’s Disease (AD), verifying that the performance of the DS population is similar to that of AD patients, although the former presented better scores on episodic immediate memory tests. There was a significant positive correlation between the scores obtained in the Lawton-IADL and those on the constructs Mental State, Episodic Memory, Linguistic Comprehension and Total ABCD. Conclusion: The ABCD battery is a useful tool in the evaluation of adults and elderly with DS and the performance of individuals in this battery correlates with indices of functionality. This is a pioneer study in Brazil, and it points to the need for a better characterization of the linguistic abilities of individuals with DS, in order to allow the elaboration of strategies that stimulate their communicative abilities as to promote greater social insertion for this population.

RESUMO

Objetivo: Caracterizar o perfil linguístico de adultos e idosos com Síndrome de Down (SD) através da Bateria Arizona para Desordens da Comunicação em Demência (ABCD). Método: Foram avaliados trinta indivíduos adultos e idosos com SD, através da bateria cognitiva MoCA, 4 escalas funcionais (Pfeffer, Lawton-IADL, Katz-IADL e IQCODE), e a bateria ABCD, que avalia os domínios Estado Mental, Memória Episódica, Expressão Linguística, Compreensão Linguística e Construção Visuoespacial. Os escores obtidos pelos indivíduos na ABCD foram correlacionados aos obtidos na escala Lawton-IADL. Resultados: Os indivíduos com SD apresentaram desempenho bastante inferior ao de adultos e idosos cognitivamente normais descrito na literatura brasileira. Na ausência de estudos semelhantes em nosso meio, comparamos os resultados aos de idosos com Doença de Alzheimer (DA), verificando que o desempenho da população com SD é equiparável ao de indivíduos com DA, embora os primeiros apresentem melhores escores em provas de memória episódica imediata. Houve correlação positiva e significante entre os escores obtidos na escala Lawton-IADL e os escores dos construtos Estado Mental, Memória Episódica, Compreensão Linguística e Total da ABCD. Conclusão: A bateria ABCD é um instrumento útil na avaliação de adultos e idosos com SD e o desempenho dos indivíduos nesta bateria apresenta correlação com índices de funcionalidade. Este estudo é pioneiro em nosso meio e mostra a necessidade de melhor caracterização das habilidades linguísticas de indivíduos com SD, a fim de permitir a elaboração de estratégias de estímulo de suas habilidades comunicativas a fim de proporcionar maior inserção social desta população.

Study carried out at the Aging Department, Associação de Pais e Amigos dos Excepcionais – APAE - São Paulo (SP), Brazil.

Conflict of interests: nothing to declare.
INTRODUCTION

Population aging is a worldwide phenomenon. Currently, the elderly population in Brazil represents 10.8% of the population\(^1\). This demographic transformation is happening rapidly and it has become a reality that affects people with intellectual disabilities (ID).

According to the American Association on Intellectual and Developmental Disability\(^2\), an individual with ID has a lower than average intellectual functioning (intellectual quotient - IQ), associated to adaptive limitations in at least two of the following areas: communicative skills, self-care, home life, social adjustment, health and safety, difficulties in the use of community resources, as well as impairments in academic, leisure and work functioning.

ID may be the result of genetic, gestational alterations, as well as perinatal and postnatal injuries, whose etiology may or not be recognized. Down Syndrome (DS) is the second leading cause of ID (1 per 700-1000 live births), and it is caused by a trisomy in chromosome 21\(^3\).

Aging of people with DS is atypical and precocious, leading to functional impairments that can jeopardize the person’s general health and quality of life. Some studies\(^4\) report signs of aging beginning at 30 to 40 years old in this population, which may account for limitations at such premature age.

Regarding language, the literature points that expressive language may be compromised in individuals with DS, being characterized by simplification and reduction of sentence length (i.e., use of short sentences), difficulties with auxiliary verbs, and syntactic errors\(^5\). Moreover, there are communication deficits in the SD population that require speech-language evaluation and rehabilitation as speech apraxia\(^6\) and impairments in oral expression, especially in individuals with concomitant DS and AD.

DS poses some challenges regarding the early detection of dementia\(^7,8\). Due to intellectual deficits throughout life, there is a difficulty in determining the first characteristic signs of cognitive decline associated with age and dementia\(^9\). This is mainly due to the inadequacy of most evaluation tools. The early assessment of dementia in individuals with DS requires the acknowledgment of cognitive impairment before the first signs of dementia\(^10\). Also, it is essential to analyze the existence of functional decline and to follow the cognitive-functional profile over a period\(^11\). The diagnosis of dementia in DS demands the intervention of a multidisciplinary team\(^12\).

Communication alterations DS may be more frequent compared to the elderly population in general. These alterations may be due to premature aging and to the genetic predisposition for the development of AD\(^13\).

Although sometimes challenging, the language assessment in patients with dementia (without ID) is supported by a lot of studies focusing on all aspects of language alterations in this population. However, in dementia associated with DS, the challenge becomes greater, due to the lack of studies addressing communication abilities in this population. Such studies would allow the development of adequate evaluation tools and guide the rehabilitation efforts for language disorders. Thus, this study aimed to describe the linguistic profile of individuals with DS and to correlate the linguistic performance to functional parameters.

METHODS

This is an open, prospective study conducted from April 2015 to November 2016. All data were collected at the Aging Department of Associação de Pais e Amigos dos Excepcionais de São Paulo – APAE DE SÃO PAULO, in São Paulo - Brazil.

The project was submitted to the Research Ethics Committee of Federal University of São Paulo and approved under registry number 1.009.370 (2/4/2015) and CONEP n° 43097214.7.000.5505. All participants and/or legal guardians received detailed information about the study and signed the Free and Informed Consent Form. Participants were clearly informed about the research objectives, as well as about the non-compulsory participation and absence of remuneration.

The study comprised 30 individuals with DS diagnosed according to the ICD-10 (Q90), aged 35 years and over, native speakers of Brazilian Portuguese, and residents in São Paulo. Cutoff scores were not established for cognitive and functional screening tests, due to the absence of standard data for the Brazilian population with DS. Education level was not computed due to the difficulty of establishing equivalence parameters regarding formal education and the lack of information on the years of schooling from the sample.

Exclusion criteria were: other etiologies of ID, previous or current psychiatric disorders, alcoholism, drug abuse, non-controlled systemic disease, evidence of cognitive or functional decline that precluded the performance of the tests, non-correctible perceptual deficits (visual or auditory), and use of psychotropic medications in doses that could impact cognitive performance (clinical judgment based on interview and physical examination).

Individuals were initially submitted to the Montreal Cognitive Assessment (MoCA)\(^14\), a global cognitive screening test, aiming to assess their cognitive profile, followed by the Arizona Battery for Communication Disorders in Dementia (ABCD)\(^15\). The ABCD is a useful tool in the identification and characterization of linguistic and communicative deficits in all stages of dementia. It consists of 18 subtests, divided into five constructs: mental state (temporal and spatial orientation), episodic memory (immediate and delayed story retelling and word learning), linguistic expression (object description, visual confrontation naming, and concept definition), linguistic comprehension (following commands, comparative questions, repetition and reading comprehension), and visuospatial construction (generative drawing and figure copying). The ABCD was translated and adapted to Portuguese and was tested in a sample of Portuguese native speakers\(^16,17\) with replication in populations with the AD and late depression\(^18\) prior to its use in the current study, which is the first to employ this instrument to assess individuals with DS in Brazil.

The duration of each evaluation session was approximately 1 hour. The main researcher was committed to providing a quiet environment, without interferences that could compromise the individuals’ performance during the evaluation. In order to facilitate the comprehension of the instructions, the examiner adapted the communication style to the needs of each individual. During the evaluation session, there were no signs suggestive of fatigue, discomfort, or lack of interest on the part of the examinee.

Functional performance of individuals was measured through four instruments: the Informant Questionnaire on Cognition Decline in the Elderly (IQCODE)\(^19\), the full version Lawton...
Instrumental Activities of Daily Living (Lawton-IADL)\(^{(20)}\), the Pfeffer Functional Activities Questionnaire (FAQ)\(^{(21)}\), and the Katz Index of Independence in Activities of Daily Living (Katz-IADL)\(^{(22)}\) was employed as a functional measure in this study. The IQCODE is a screening questionnaire\(^{(19)}\), which is applied to the caregiver (informant) and can provide information about the evaluated person over time (10 years). It includes 26 items, scored from 1 (much better) to 5 (much worse). The final score is a weighted mean, where values above 3 indicate changes from the baseline.

The Lawton IADL Scale\(^{(20)}\), assesses eight daily living activities, including the ability to use the telephone, shopping, food preparation, housekeeping, laundry, mode of transportation, responsibility for own medications, and ability to handle finances. Each item is scored 0 or 1 (maximum: 90 points) and the individual is then classified as independent, partially dependent and totally dependent according to clinical judgment.

The Pfeffer FAQ\(^{(21)}\), consists of items related to cognitive / social functions such as: handling finances, shopping, working on a hobby, making coffee, preparing a meal, keeping track of current news, paying attention to TV, remembering appointments, travelling out of neighborhood, and it is scored from 0 to 30, where higher scores denote poorer functional skills. The lower the score, the greater the independence and autonomy. Scores equal to or greater than 5 have good specificity and sensitivity for the diagnosis of functional decline in people with DS.

The KATZ-IADL\(^{(22)}\) was developed to measure the physical functioning of people with chronic diseases and their recovery in the functional performance of six activities: bathing, dressing, toileting, transferring, continence, and feeding. Scoring ranges from 0 to 6, depending on how many activities the individual can perform (1 point each). Classification varies from 0 (dependent for all activities) to 6 (independent for all activities).

**STATISTICAL ANALYSIS**

To characterize the demographic profile and population performance in cognitive, linguistic, and functional tests, a descriptive statistical analysis was performed (means and standard deviation were calculated).

Spearman correlation test was used to verify the association between cognitive/linguistic (ABCD constructs) and functional (Lawton-IADL) performance. A significance level of 0.05 was adopted.

**RESULTS**

The sample comprised 30 adults (18 men and 12 women) with a mean age of 47.8 (6.7) years. Twenty-seven individuals were right-handed, two were left-handed, and one was ambidextrous.

Scores obtained in cognitive and functional scales are displayed in Table 1. Table 2 displays the performance of the individuals in the ABCD.

There was a significant positive correlation between Mental State, Episodic Memory, Linguistic Comprehension and total scores in the ABCD and the Lawton-IADL scores. These data can be visualized in Table 3.
DISCUSSION

Communication is one of the fundamental aspects of human life. It is through communication that human beings interact and express their feelings. Impairments in cognitive abilities (memory, problem-solving and verbal reasoning) may interfere with the functional use of language. The role of the speech therapist in dementia consists in the maintenance of communicative aiming at the promotion of quality of life.

The objective of the present study was to describe the linguistic profile of individuals with DS in the aging process using the ABCD and to verify its association with the degree of functionality in this population.

Early aging, such as observed in DS, contribute to the progressive decline of cognitive functions, such as memory, language, executive functions, the speed of information processing, attention and visuospatial abilities. Individuals with DS may present biological changes in brain development. Among them, alterations in neocortical and hippocampal development can be found. Premature deposition of Alzheimer's disease (AD) biomarkers, such as senile plaques and neurofibrillary tangles in young adults (at 17 years of age) with DS, has already been described. These biological differences range from structural modifications at the cellular level to changes in neurochemical parameters and, ultimately, impact brain function. In addition to the biological changes inherent to the aging process, deficits in functional abilities also occur, which predisposes such individuals to greater impairment in general health and higher prevalence of dementia.

It is known that there is a close correlation between AD and DS. The prevalence of AD in people with DS is described as varying from 6% to 75%. Among the relevant factors for understanding the etiology of dementia are genetic factors. The chromosomal alterations present in individuals with DS are related to the early development of dementia in these individuals, particularly in those younger than 65 years.

Although still scarce, there is evidence that both comprehensive and expressive language is impaired in individuals with DS throughout the aging process. The decline in cognitive and language skills are a cardinal marker of functional decline in this population. However, cognitive evaluation in DS remains a major challenge due to difficulties in standardization of assessment instruments, and the need to consider the pre-existing cognitive impairment. Performance of individuals in the MoCA test was somewhat erratic and the highest scores obtained fell much below the cutoff score that indicates cognitive impairment in adults without ID, which is 26. But we must notice that the MoCA was not developed for the assessment of people with ID and that individuals in our sample had different levels of literacy and cognitive baseline skills.

The ABCD proved to be a useful instrument in adults and elderly with DS, although some adaptations in the administration might be necessary, such as repetition or simplification of instructions, due to an increased time for information processing and difficulty in understanding commands.

Due to the lack of similar studies in the literature, we compared our findings to those described in studies using the ABCD in Brazilian AD patients, based on the aforementioned link between AD and DS. Overall, individuals in our sample presented a performance comparable to those of non-DS AD patients in most tasks. However, it is worth noting that the individuals with DS performed better in the “immediate story retelling” and “word learning – free recall” tasks, but worse in “repetition”, comprehension of words and sentences, and all visuospatial tasks. These data are consistent previous findings reported in the literature that show cognitive and linguistic deficits in this population, as well as differences between DS adults / elderly with and without dementia. They most probably reflect, on one side, that memory codification is more preserved in DS persons, in spite of their cognitive limitations, but are also difficult to interpret when taking into account the heterogeneity of cognitive/educational background of our population.

Clinical assessment in DS should focus on functional communication in daily routine and main life settings. This may be accomplished, at first, by measuring basic communication skills, such as providing simple “yes/no”, “I like/I dislike” responses, but also by evaluating interactions of increasing complexity such as to initiate, develop, sustain, and terminate a conversation in a proper manner. Social communication skills must be emphasized, as they are essential to promote the individual’s insertion in his/her environment.

Individuals with DS are usually interested in starting conversations with different interlocutors, but frequently lack the ability to do so; in many cases, difficulties to make solicitations or ask for information may be present. Hence, an adequate speech and language evaluation addressing multiple aspects of comprehensive and expressive language have an enormous potential to contribute to the development of tailored speech therapy interventions.

In this study, we employed four different functional instruments to obtain a combined measurement of the individuals’ functional status. From them, the Lawton-IADL proved to be more suitable to provide detailed data regarding both functional abilities and dependency level, and this led us to choose it to perform the correlation analysis. We were then able to verify an association between scores in the Lawton-IADL and performance in Mental Status, Episodic Memory, Linguistic Comprehension, and Total ABCD. Although the existence of correlations does not imply a cause-effect relation, we believe that they reflect the interdependency between language skills and the ability to perform daily tasks in an independent manner, especially those tasks that demand social interaction, such as shopping, traveling and, most certainly, using the telephone.

We understand that the person with DS needs to be socially included. However, this inclusion will be limited if there is no incentive and/or promotion of communication in their space of coexistence. In Brazil, there are some projects and programs of inclusion for Adult Intellectual Disability (ID), including DS.
in the labor market. To make it possible, it is imperative that the level of independence and autonomy in daily life activities of this population may be traced. Language skills, on which all communication depends, is of extreme relevance at any stage of life.

Limitations of this work include the small sample, the heterogeneity of literacy among the participants, and the use of cognitive and functional batteries that were not designed for the evaluation of this specific population.

CONCLUSION

To the best of our knowledge, this is the first study addressing language abilities in adult and elderly DS population in Brazil. We conducted a preliminary study in order to verify the suitability of the ABCD (an instrument designed for dementia population) in the evaluation of individuals with DS. We concluded that the ABCD might be a useful tool to assess adult individuals with DS. We found a significant positive correlation between communicative abilities and measures of autonomy and independence in the activities of daily living in our DS sample. Further studies addressing the interrelation between communication and functional skills in the adult Brazilian DS population are warranted.

REFERENCES


Author contributions

CLC was responsible for data collection and tabulation, and manuscript writing; LRC supervised data collection; AFRB, and MR collaborated with data analysis, interpretation of results, and overall supervision of the stages of manuscript writing and elaboration.