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# Cognitive-Linguistic Profiling in Young Onset Dementia: A case Study

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**Case Report**
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## Abstract

The onset of dementia is after 65 years. Though there are evidence reported in the past regarding the onset of dementia before this age, it is very rare. Studies carried out in the past have concentrated on the site of lesion, genetic base for the young onset dementia and the studies focusing on cognitive linguistic profiling are extremely rare. The current study focussed on the cognitive linguistic profiling and profiling the effect of dementia on the activities of daily living. The findings suggested that the cognitive linguistic deficit was very severe and had a note-worthy impact on the activities of daily living. This is pivotal in designing the plan of intervention and deciding the course of treatment.

**Keywords:** Cognitive Decline; Rate of Decline; Deficits; Futuristic Strategy; Profiling

**Abbreviations:** APP: Amyloid Precursor Protein; PSEN1 and PSEN2: Presenilin-1 and Presenilin-2; MAPT: Microtubule-Associated Protein Tau.

## Introduction

Dementia is the loss of cognitive functioning over time. It is an umbrella term used to refer to a cluster of cognitive linguistic deficits [1]. The onset of dementia is after 65 years [2]. Dementia in younger individuals is extremely rare and very few exceptional cases have been reported in individuals below 60-65 years [3]. The term pre-senile dementia was used as a diagnostic label for the condition, however the term is sparsely used in the recent years. The term young onset dementia has replaced this term contemporarily. The term young onset dementia is used when the onset of dementia is before the age of 65 years [4]. Though the age of 65 years is arbitrary, it is the informal criterion used for diagnosis of young onset dementia [5].

As far as the epidemiological findings is concerned, there is a clear dearth of prevalence studies is concerned. A study carried by Harvey and colleagues estimated that the prevalence of young onset dementia is 54 per 100,000 [6]. Alzheimer's disease was the most common cause for the young onset dementia. A prevalence figure in Japan was mentioned by Ikejima C, et al. [7]. They reported a prevalence rate of 42 per 100,000. The prevalence figure was almost the same as the aforementioned study. The prevalence rate suggests that the condition is rare and worthy to be profiled.

The cause of young onset dementia is profiled by some authors. The common causes of young onset dementia is Alzheimer's disease, vascular disease, fronto-temporal dementia, and Lewy body dementia [8]. The same causes are listed for Dementia seen in relatively older individuals also

hence there are no exclusive causes of young-onset dementia. The clinical features would differentiate the feature of early onset dementia with dementia seen in older individuals as they can differ from the standard prototype. A seminal study carried out by Blessed and colleagues reported a case of Alzheimer's who were 52 at the time of reporting [9]. A detailed genetic analysis revealed mutations in the amyloid precursor protein (APP) and presenilin-1 and presenilin-2 (PSEN1 and PSEN2) genes in this case [10].

Fronto-temporal lobar degeneration is yet another common cause of dementia in general. It refers to a set of conditions associated with atrophy confined to the frontal and temporal regions of the brain. Within the fronto-temporal lobar degeneration three variants of dementia have been recognised [11]. The three variants include the behavioural variant, semantic variant (characterised by problems in single word comprehension and the progressive non fluent variant of dementia (associated with effortful speech similar to the condition seen in Broca's or any type of non-fluent aphasia). The fronto-temporal lobe degeneration can be the common cause of young onset dementia also. Some studies have highlighted the positive family history associated with this condition. With gene mutations in microtubule-associated protein tau (MAPT) and progranulin (GRN) gene mutations [10,11].

As the cause of dementia is fronto-temporal degeneration in the client reported here in the current study, fronto-temporal lobar degeneration is profiled in detail. A series of examination including behavioural examination, neurological assessment, cognitive linguistic assessment, blood tests and genetic tests has to be carried out in clients diagnosed with fronto-temporal degeneration based on the severity of symptoms. The cognitive linguistic profiling is an essential crux is young onset dementia regardless of the causes as the patients may present varied set of symptoms and the manifestation of the symptoms may differ with the conventional dementia [12]. Though there are data-banks corroborating details of a variety of clients diagnosed with Dementia, there is a clear death of cases of early onset dementia. The manifestation of symptoms in young onset dementia can be heterogeneous [13] as it varies across the individuals affected by it. In addition to this, it is also reported that, patients with a younger onset of dementia may have a variety of co-morbid in addition the core cognitive-linguistic deficit [14,15]. Hence the current study deals with the cognitive linguistic profiling of a 42 year old client diagnosed with fronto-temporal lobe degeneration.

## Case Report

Participant details: The client was 42 year old lady when she reported to a premier institute in speech and hearing. She was B-Com Graduate and was married for the last 22 years. She was a home maker. Her husband was working as Manager in a private company. The couple had twins (one boy and one girl) who were 15 year old. The client had an elder brother who was reportedly normal. The husband revealed that the client's mother 9his mother in law) was diagnosed with Dementia after the age of 60 years. Detailed evaluation was not carried out with the mother due to financial constraints. The client's mother expired when she was 65 year old and had severe dementia at the time of her death. She had behavioural and cognitive linguistic deficits apparently but no details was available in specific.

The client's main complaint was forgetfulness and change in behaviours since 2 years. In the initial one year after the symptoms developed, it did not interfere her activities of daily living, in other words it was only in the last one year that the symptoms turned severe to cause interference with the activities of daily living. Even after developing the symptoms the client was never aggressive and was very polite.

Her problem was first identified by the husband and was observed during meal time. She was making mistakes in choosing ingredients and she would add few ingredients in more quantity, when questioned she was denying it. Following this she was facing difficulties in retrieving objects and was apparently misplacing objects and was finding very difficult in finding these objects. She was apparently getting irritated for trivial things. She lost interest in pursuing her hobbies like singing and knitting. In a year, her symptoms started turning severe. She failed in identifying neighbours, friends and relatives. She lost the ability to categorise things (like the item belonged to the living room, kitchen etc).

She also developed symptoms like word finding difficulty, confusions and confabulations. She had problems in performing her activities of daily living like cooking, wearing saree, she used to loose ways within the road which she lived. She was not able to call her children by their names. She got confused with the locations within the house eventually. She was in need of assistance for almost all the activities at home.

Her diet was normal. The husband complained that she developed craving for sweets. She did not have a history of emotional liability, delusions and hallucinations, seizures and myo-clonic jerk. The husband reported that few members of the family from her paternal side had similar problems in addition to her mother having a similar condition, but no reports were available at the time of testing and none of them were alive too. A detailed cognitive linguistic profiling was done with the intent of quantifying the cognitive-linguistic deficits and choosing an intervention strategy.

Montreal Cognitive Assessment- Kannada version was carried out on the client, the client obtained a score of 2 on 30. The client could not follow the instructions on most of the occasions. The client was able to get a score of 1 on immediate recall and 1 on naming. Hence the findings on the test showed that the client had severe cognitive impairment.

In addition to this, cognitive-linguistic profiling was carried out. In order to assess attention and memory, recall task was carried. Recall task taps encoding, storage and retrieval. Encoding taps attention, while storage and retrieval taps memory. 4 item chunks was given to test recall. The client was not able to channelize attention, she was not able to retrieve the items on most of the occasion. At times she was able to retrieve the first item in the chunk reflecting primacy effect.

Picture description was carried out. The picture stimulus of Western Aphasia Battery was used for tapping picture description. The client was able to identify individual items like boy, man, dog etc. She was not able to identify the actions associated with the noun and produce sentences. She struggled with the retrieval of verbs compared to nouns. Her phrase length was confined to 1-2 words.

Conversation task was carried out. Few questions related to her daily routine was asked. Few of the questions like '"What is your husband's name"? Where do you do leave etc. tapped memory also. She was not able to answer to most of the questions, she repeated the words in the questions again. Following the conversation task Repetition task was carried. Repetition was tested at various levels, like word repetition, phrase repetition and sentence repetition. She was able to repeat only the words on most occasions. However phrase and sentence repetition was affected. Her discourse skills was also affected though she was maintaining eye to eye contact during conversation. Her topic navigation, coherence, cohesion was affected.

## Discussion

Young onset dementia is a rare condition. The prevalence figures carried out in the western countries6 and Asian countries 7 show that the prevalence rate is extremely rare. Evidence' pertaining to young onset dementia is purely based on case-studies. The case study on young dementia has focussed on the site of lesion and the genetic bases of these cases. Studies on genetic base have identified protein tau (MAPT) and progranulin (GRN) gene mutations [11]. There was a strong genetic base in the client as the mother and other relatives from the maternal side had similar problems.

There was practically very few studies on the cognitive linguistic profiling in these cases which necessitated the current study. Detailed assessment regarding the activities of daily living showed that the impact of dementia on this skill. The client had problems in orientation, was not able to find things kept in the confined places. Her object knowledge was depleting with time. She was not able to identify objects based on their function. Her navigation skills looked compromised. She was not able to differentiate kitchen with the leaving room. The disease progression followed a rapid rate. Rate of decline was faster or equal to the rate of cognitive decline in older dementia cases.

The cognitive skill was probed in detailed to figure out the intervention strategy to be used in this client, as mentioned in the case report section. All the cognitive skills were severely affected. Her attention was confined to about 5 seconds. She was not able to channelize her attention for conversation also. The same question in the conversation had to be repeated three to four times to evoke a response. Rephrasing did not favour. Owing to the deficits in this skill even her functional communication was affected. She was not able to hold more than one or two items in her memory. This further complicated information retrieval. Her long term memory was not affected to a greater extent as reflected on the conversation task. Though she was able to identify her family members, she was not able to identify them by names. She was not able to remember her personal details.

She did not have any co-morbid conditions other than the cognitive-linguistic impairment. She did not have history of seizures, myoclonic jerks. Hr walking, posture and motor stability was unaffected. This contradicted the findings mentioned in the other case studies [12]. After the detailed evaluation, the immediate care giver was counselled regarding the importance of cognitive-linguistic intervention in her case. As the disease progression was happening at a rapid rate and also that the cognitive linguistic skills were affected to a greater extent, futuristic strategy like Augmentative and Alternative Communication was suggested as a part of intervention.

## Conclusions

42 year old client with a positive family history dementia reported the out-patient department of a premier speech and hearing institute in India. Her condition lasted from the last two years. The disease progression was happening at a fast rate. The main intent of the family was to seek cognitive-linguistic intervention. A detailed evaluation was carried out in two parts. The first part elicited details regarding the impact of dementia on activities of daily living and it was that condition was severe to intrude the activities of daily living. Second part of the evaluation focussed on the cognitive linguistic profiling. Detailed profiling revealed that the cognitive skills were again severely affected. Following

this, use of Alternative and Augmentative communication was suggested for the case.

## Authors' contribution

Abhishek B P was involved in concept development, study design, analysis of the results and writing the manuscript. Haripriya was responsible for proof reading.

## Ethical Statement

Ethical standards: The manuscript adheres to the ethical standards according to the Declaration of Helsinki. Ethical Approval: All procedures performed in this study were in accordance with the ethical guidelines of bio-behavioral research involving human subjects of the All India Institute of Speech and Hearing, Mysore.

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## Conflict of interest

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

## Informed Consent

Informed consent was obtained from the patient/ caregiver to participate in the study.

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