Telemedicine for Delivery of Care in Frontotemporal Lobar Degeneration During COVID-19 Pandemic: Results from Southern Italy

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- 11 Abstract.
- 12 Background: The COVID-19 pandemic is changing clinical practice in neurology, after the governments decided the introduc-
- tion of social distancing and interruption of medical non-emergency services in many countries. Teleneurology is an effective
- tool for the remote evaluation of patients but its adoption for frontotemporal lobar dementia (FTD) is in a preliminary stage.
- ¹⁵ **Objective:** We evaluated multidisciplinary assessment of patients with FTD using telehealth during the COVID-19 pandemic.
- Methods: All patients received a diagnosis of FTD during 2018-2019 according to international criteria. A structured questionnaire and Clinical Dementia Rating Scale (CDR)–FTD were used by the neurologist with patients and/or caregivers.
- ¹⁸ Index symptoms of COVID-19 infection were searched.
- 19 **Results:** Twenty-eight clinical interviews were completed with caregivers and four with both patients/caregivers. Most patients
- and caregivers were satisfied with the neurological interview and expressed their willingness to continue to be included in
- remote evaluation programs (90%). Fifty percent of patients experienced significant worsening of clinical picture and quality
- of life since the start of social distancing. The CDR-FTD scale revealed a significant worsening of behavior (p = 0.01) and
- p_{23} language functions (p = 0.009), compared to the last in-person evaluation at the center. One patient presented index symptoms
- of COVID-19 infection and was confirmed to be positive for COVID-19 with pharyngeal swab.
- 25 **Conclusion:** The study was conducted in Italy, one of the countries hit particularly hard by the COVID-19 pandemic, with
- interruption of all non-emergency medical services. Our study indicates that telemedicine is a valid tool to triage patients
- ²⁷ with FTD to increase practice outreach and efficiency.
- 28 Keywords: COVID-19, frontotemporal lobar dementia, multidisciplinary care, pandemic, quality life, telemedicine

INTRODUCTION

The 2019 coronavirus disease (COVID-19) outbreak has important implications for clinical care in Alzheimer's disease (AD) and frontotemporal lobar dementia (FTD). Many countries have implemented social distancing measures, such as quarantines

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within urban areas, prohibition of travel to and from
certain countries, and suspension of broad range of
activities, involving large numbers of people such
as outpatients visits in public and private hospitals.
All these decisions were aimed to limit the risk of
COVID-19 infection.

Government containment maneuvers to reduce 41 infections included the ban of all non-urgent clini-42 cal activities, thus limiting the access to hospitals, 43 with significant discomfort for older people affected 44 by chronic conditions. Not unexpectedly, such pop-45 ulation segments, with high comorbidities and being 46 particularly frail, have the highest mortality rate from 47 COVID-19 [1]. FTD is an umbrella term encompass-48 ing a wide spectrum of clinical pictures characterized 49 by progressive changes in behavior, personality, 50 and/or language functions associated with degen-51 eration of the frontal and temporal lobes [2-4]. 52 Several clinical FTD variants have been described. 53 The behavioral variant of FTD (bvFTD) and pri-54 mary progressive aphasia (PPA) represent the most 55 frequently recognized clinical syndromes, both in 56 tertiary centers [5, 6] and in population-based set-57 tings [7]. Progressive supranuclear palsy (PSP) and 58 corticobasal degeneration syndrome (CBD), and oth-59 ers which are considered to be part of the spectrum 60 of tauopathies, have been shown to overlap both 61 clinically and neuropathologically, and are consid-62 ered under the same label of the FTD spectrum [8]. 63 FTD also often overlaps with amyotrophic lateral 64 sclerosis (ALS), with symptoms of FTD occurring 65 in 15%- 41% of patients with ALS and features 66 of ALS occurring in 15% of FTD patients [9, 10]. 67 Multiple system atrophy (MSA) may also present 68 clinically and pathologically as an FTD [11]. More-69 over, patients with FTD represent a segment that 70 could be particularly affected by both the epidemic 71 and the rules to reduce its transmission due to 72 the specificity of FTD clinical features; patients 73 have special difficulties in understanding changes 74 in the surrounding environment and social rules 75 [12]. 76

It is well recognized that people with FTD ben-77 efit from multidisciplinary care in terms of survival 78 and quality of life [13, 14]. However, access to multi-79 disciplinary clinics may be difficult for patients with 80 FTD due to distance from the nearest tertiary refer-81 ral center and costs of travel [15]. Patients with FTD 82 have been particularly limited by the new rules during 83 the COVID-19 epidemic. Moreover, in our country, a 84 significant proportion of patients with dementia live 85 in medically served residencies with high incidence 86

of COVID-19 infections and an extraordinary high mortality [16].

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Telehealth is a rapidly progressing field that is constantly exploring patient care with the support of multiple applications and services including remote videoconferencing, e-mail, apps, and other forms of technology. Teleneurology is an effective tool for the rapid evaluation of patients in remote locations [17] with a well-established use in acute stroke [18, 19] and epilepsy [20, 21] and more recently in neurodegenerative disorders, such as Parkinson's disease [22] and dementias [23–27]. Recent studies supported the use of telemedicine in cognitive rehabilitation in mild cognitive impairment, AD, and FTD [28].

Telehealth in the current COVID-19 pandemic has become the primary way to deliver care, especially as means of reducing the risk of cross-contamination caused by close contact [24]. Telemedicine approach in the era of COVID-19 has been used for monitoring of persons potentially affected by the virus [1, 24], but to date, no study on FTD is available. A recent editorial of the Alzheimer Association Society and other similar advocacy groups encouraged novel public health initiatives, including telehealth, from countries with large or growing populations with dementia in order to protect these vulnerable patients and their caregivers during the COVID-19 outbreak [29].

The aim of our study was to evaluate whether the assessment of patients with FTD using telemedicine is feasible and acceptable to patients and caregivers in the era of the COVID-19 pandemic.

METHODS

All study subjects were patients receiving multi-120 disciplinary care at the Center for Neurodegenerative 121 Disease and The Aging Brain of the University 122 of Bari at the Hospital "Card. G. Panico" Tri-123 case (Lecce). Subjects were enrolled within the 124 "SLAP-Dem Study", a population-based study on 125 rare neurodegenerative disease that was approved by 126 the Ethics Committee for Medical Research at Lecce 127 on 25 May 2017. Both patients and their caregivers 128 provided their informed consent verbally [Italian 129 Guarantor for the protection of personal data has pro-130 vided that for treatments for "treatment purposes" 131 it is not necessary to acquire the patient's consent 132 (Measures Registries n. 55, 7 March 2019)]. Patients 133 with a diagnosis of bvFTD [30] and PPA [31], also 134 overlapping with PSP [32] or other syndromes such 135

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as CBD, MSA, and ALS, according to the respec-136 tive international criteria [33-35], were selected and 137 contacted for the study. In particular, bvFTD and 138 PPA plus Parkinsonism are defined as the presence 139 of extrapyramidal signs not sufficient to meet diag-140 nostic criteria for PSP, CBD, and MSA. All patients 141 received a diagnosis in 2018 and 2019 after a com-142 plete clinical and paraclinical assessment and had at 143 least one access for follow-up after the baseline visit 144 at the Center during the year 2019; new cases with 145 diagnosis after their first visit in the Center still pend-146 ing or patients not referring to the Center in the last 147 year were not included as it was assumed that they/we 148 would not have a basis for comparison of their previ-149 ous care. Patients were included regardless of disease 150 severity and distance from the clinic. 151

The telemedicine approach (tele/cell-phone) from 152 the clinic to patient's home was performed by the 153 interaction of the neurologist with the patient (if 154 it was possible) or the caregiver (defined as the 155 person dedicated to the patient's care for at least 156 8-10 hours a day). The telemedicine assessment in 157 this study was based on a structured questionnaire 158 designed to collect demographics and clinical infor-159 mation [diagnosis, onset of disease, clinical status 160 and changes since last visit in the following domains: 161 cognition, behavior and personality, language, sleep 162 disturbances, nutritional status, swallowing capac-163 ity, respiratory function, access to rehabilitation 164 and speech therapy, pharmacological therapy] with 165 the aim to identify significant changes or prob-166 lems since the last multidisciplinary evaluation 167 (see Supplementary Material). Disease progression 168 was evaluated using the Clinical Dementia Rating 169 Scale-FrontoTemporal Dementia (CDR-FTD) con-170 sidering both SoB (sum of boxes) and the Behavior 171 and Language items [36]. Results obtained during 172 the teleconsultation were compared with those of 173 the last in person evaluation at the center. We also 174 assessed the distress induced on caregivers in the last 175 period by the COVID-19 pandemic and subsequent 176 governmental rules. The global caregivers' distress 177 was evaluated on a Likert scale from 0 (none) to 5 178 (severe) with the caregiver distress question of the 179 Neuropsychiatric Inventory [37]. 180

The questionnaire also included information concerning possible signs/symptoms of COVID-19 infection in patients and/or caregivers/family. One week after the teleconsultation, a survey was administered via email or via WhatsApp to the patient or caregiver to ask participants about their opinion of the telemedicine interview.

RESULTS

Data were collected during the COVID-19 emergency from the period between 10 April and 30 April 2020. We identified 80 FTD consecutive cases, who were referred to the center during the calendar years 2018 and 2019 for diagnosis. Based on the study criteria. 33 patients were selected and all of them accepted to participate to the study. Overall, 32 telemedicine clinical interviews were completed. In one case, the encounter was not completed because the patient affected by bvFTD had died due to respiratory failure 3 months before the teleconsultation. Nearly all caregivers were patients' daughters/sons (n = 15) and spouses/partners (n = 15); one was a daughter-in-law and one a nephew. Finally, four interviews were also conducted directly with the patients. All patients included in the study lived at home. A video interaction was offered but refused by all patients because the large majority of participants did not own a computer or smartphone or, while they owned a cellphone, they were not familiar with this type of technology. All subjects contacted for the study felt comfortable during the interaction with the clinician. The total length of the telehealth visits was 60-90 minutes.

Demographic and clinical characteristics are summarized in Table 1. The patients include 18 males and 14 females (sex ratio = 1.3) with mean age at evaluation of 66 years and mean duration of disease from onset of 73 months (Table 1). Clinical diagnoses were distributed as follow: 12 bvFTLD (39%), 9 bvFTD plus Parkinsonism (28%), 2 PPA (6%), 3 PPA plus Parkinsonism (9%), 2 bvFTD-PSP (6%), 2 bvFTD-MSA (6%), and 2 bvFTD-ALS (6%; Fig. 1). Among all domains explored during the telehealth evaluation, we found significant worsening since last visits mostly in behavior (56%), language (47%), and cog-

Table 1				
Demographic and clinical features of FTD patients $(n = 32)$				
Sex M/F	18/14			
Age at evaluation (mean \pm SD, y)	66.25 ± 9.76			
Disease duration since disease	73.22 ± 46.70			
onset (mean \pm SD, months)				
Time since last visit (mean \pm SD, months)	6.78 ± 3.84			
Type diagnosis				
PPA	N 2			
PPA-Parkinsonism	N 3			
bvFTD	N 12			
bvFTD-Parkinsonism	N 9			
bvFTD-PSP	N 2			
bvFTD-MSA	N 2			
bvFTD-ALS	N 2			

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Fig. 1. Distribution of different FTD clinical phenotypes of patients enrolled in the teleconsultation. The clinical diagnoses of our cohort were distributed as follow: 12 bvFTD, 9 bvFTD plus Parkinsonism, 2 PPA, 3 PPA plus Parkinsonism, 2 bvFTD-PSP, 2 bvFTD-MSA, 2 bvFTD-ALS.

nitive functions (53%; Fig. 2). Memory was described as worsened in 17 out 32 patients and (3 were patients with PPA (18%) and 14 patients with bvFTD (82%); Fig. 3). Significant sleep disturbances (problems in falling asleep, overall sleep quality) were reported in 25% of patients since the start of quarantine time (Fig. 2). All patients reporting sleep problems as disabling received new sleeping therapy prescriptions. Among all patients reporting the onset or worsening of preexisting behavior disorders, 78% of these were 232 bvFTD and 22% PPA (considering all the FTD spec-233 trum together); while among all patients reporting 234 the onset or worsening of preexisting language prob-235 lems, 67% were bvFTD and 33% PPA (Figs. 4 and 5). 236 Nearly 30% of patients with the spectrum of bvFTD 237 reported no changes since the last visits; 14 out 27 238 patients presented behavioral changes (apathy, iras-239 cibility, verbal aggressiveness, anger attacks), 7 out 240 27 patients presented with both language (anomia, 241 reduced fluency) and behavior changes, 13 out 27 242 patients presented with both behavior and cogni-243 tive changes with prevalent memory disorders. All 244 patients with PPA spectrum complained of a wors-245 ening of language functions since the last visit, in 246 particular they reported more anomia and reduced 247 verbal fluency. A significant worsening of behavior 248 and language functions at CDR-FTD scale performed 249 during teleconsultation compared to the CDR-FTD 250 performed during last evaluation at the Center was 251 reported (p = 0.01 and p = 0.009, respectively); while, 252 no differences were found for CDR SoB (Table 2). 253

In 8 out 32 patients (2 PPA plus parkinsonism, 6 bvFTD plus parkinsonism A), a mild worsening of bradykinesia and limb rigidity since last evaluation at the center was reported, while initial problems

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Fig. 2. Percentage of changes reported by patients/caregivers in the main domains investigate during multidisciplinary teleconsultation compared to last visit. During the telehealth evaluation, a significant worsening since last in person visit was observed mostly in behavior (56%), language (47%), and cognitive functions (53%).



Fig. 3. Cognitive decline distribution among different behavioral and language clinical phenotypes. Memory was reported as worsened in 17 out 32 patients. Among these, 14 patients were affected bvFTD (82%) and 3 were patients with PPA (18%).



Fig. 4. Behavior worsening distribution among different behavioral and language clinical phenotypes. 78% of patients reporting the onset or worsening of preexisting behavior disorders were affected by bvFTD and 22% by PPA (considering all different bvFTD and PPA spectrum together); while among all patients reporting the onset or worsening of preexisting language problems, 67% were bvFTD and 33% PPA.



Fig. 5. Language worsening distribution among different behavioral and language clinical phenotypes. 67% of patients reporting the onset or worsening of preexisting language problems were affected by bvFTD and 33% by PPA.

Table 2
Functional evaluation of FTD patients enrolled in the teleconsul-
tation by clinical Dementia Rating Scale-FTD

	Last visit	Teleconsultation	t-test
$\overline{\text{CDR SoB}, \text{mean} \pm \text{SD}}$	5.30 ± 3.13	5.41 ± 3.11	0.13
CDR-Behavior	1.41 ± 0.70	1.58 ± 0.77	0.01
CDR-Language	0.69 ± 0.66	0.86 ± 0.79	0.009
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Data are mean \pm SD.



Fig. 6. Global level of satisfaction about the telehealth consultation of FTD spectrum patients/caregivers participating to the likingsurvey. Results from the satisfaction surveys indicated that most of the patients were consistently satisfied with the telemedicine visits (88%). No patient was unsatisfied at all by the teleconsultation.

in swallowing was reported in 3 patients (1 bvPSP and 2 bvMSA). None of the patients enrolled in the study reported significant nutritional or respiratory changes, including the 2 patients with bvFTD-ALS.

With regards to treatment, the therapy was arbitrarily suspended in 3 patients by their caregivers and then promptly restored after the teleconsultation; only one patient with bvFTD-ALS reported that the therapy plan for riluzole was expired so it was renewed and then sent directly to the territorial pharmacy by email, in order to guarantee minimal exposure to virus infection of patients and their caregivers.

Sixteen patients with bvFTD associated with parkinsonian syndromes and all the 5 patients with PPA were performing physiotherapy and speech therapy at home before the outbreak and all of them reported a subjective feeling of discomfort after its discontinuation because of COVID-19 pandemic restrictions.

Based on the overall clinical information reported during the telehealth, a follow-up visit either in person or by telemedicine was ordered to be scheduled within three months for 26 out of the 32 patients.

All caregivers interviewed reported the presence of psychological and emotional distress; in particular 15 out 28 (53%) reported a minimum or mild distress (1–2 on scale), while 13 out 28 (47%) reported a moderate-severe distress (from 3 to 5 on scale).

Furthermore, we investigated the presence of any symptoms/signs potentially indicative of COVID-19 infection (fever, cough, and other flu symptoms) in patients and in their caregivers. A patient with bvFTD-MSA was reported to present with fever and cough for a few days before the teleconsultation occurred on 10 April. A detailed epidemiological interview was reconstructed with the caregiver (the wife) who reported that the patient had been hospitalized until the beginning of March in a rehabilitation clinic in Lombardy in the Red Area of Italy (high 258

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experience					
	Very satisfied	Fairly satisfied	Not at all satisfied		
Are you satisfied with the quality of the voice sound during the telehealth consultation?	81%	19%	0%		
Are you satisfied with the Neurologist's skill to understand your problems during the telehealth consultation?	88%	12%	0%		
Globally, are you satisfied with your experience with telehealth consultation?	81%	19%	0%		
	Yes	No			
Did you seem to be talking with the Neurologist as you were face-to-face with him?	82%	18%			
Did you feel comfortable during the telehealth consultation?	89.5%	10.5%			
Did you think you have saved money by avoiding travelling to the center?	88%	12%			
Would you like to continue follow-up also by telehealth consultation in the future?	90%	10%			

Table 3 Survey's satisfaction and agreement level about telehealth

prevalence of COVID-19 infection in the general 297 population) during the COVID-19 pandemic. We 298 contacted the family doctor of the patient who ordered 299 an oropharyngeal swab that resulted positive for a 300 COVID-19 infection. The patient did not need hos-301 pitalization and healed completely; furthermore, two 302 consecutive swabs resulted negative for the infection 303 in the following 2 weeks. None of his family members 304 presented with COVID-19 symptoms/signs. 305

After a few days, 21 out of 32 subjects (66%) 306 participated in the satisfaction surveys. Most were 307 consistently satisfied with the telemedicine visits 308 (Fig. 6). In particular, they were satisfied with the 309 clinical interview made by the neurologist (88%), 310 and by the possibility to interact directly and more 311 easily with the clinician while being at home (88%). 312 The majority of subjects (88%, Table 3) expressed 313 their willingness to continue to be placed in the 314 telemedicine program; they also underlined the cost 315 reduction resulting from avoiding travel to the clinic, 316 considering that more than 70% of the patients lived 317 more than 30 km from the center 318

319 DISCUSSION

The pandemic due to COVID-19 is changing clinical practice in neurology, after governments

introduced social distancing, interrupting medical 322 non-emergency services in many countries [38]. 323 These decisions are particularly critical, in particular 324 for elderly patients with chronic neurodegenerative 325 diseases like dementias, because of the presence of 326 physical frailty and comorbidities. To our knowledge, 327 this is the first study that aimed to analyze the role of 328 telemedicine as a valid tool to deliver and support care 329 to people with FTD during the COVID-19 pandemic 330 that placed Italy among the most seriously hit coun-331 tries in the world. It is well known that dementia is 332 a pandemic particularly relevant in consequent mor-333 bidity and mortality in Europe and North America, in 334 countries with aging societies with high prevalence 335 of elderly subjects [39]. The double hit of dementia 336 and COVID-19 pandemics has generated great wor-337 ries and issues for people with dementia and their 338 families. All the issues are important for subjects at 339 home and especially for people living in long-term 340 residential care facilities, where the rate of spread and 341 the reported mortality of COVID-19 were particularly 342 high in all Europe [40]. 343

Teleneurology is an effective tool for the rapid evaluation of patients in remote locations [17], with a well-established use in acute stroke [18, 19] and epilepsy [20, 21]. Telehealth is in the process of exponentially expanding as clinicians aim to preserve patients' access to clinical care during the COVID-19 pandemic [41].

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Previous reports supported telemedicine as a useful tool for elderly patients with chronic illnesses such as dementias and their caregivers [42, 43]. In areas where there is inadequate presence and distribution of clinical resources and community health systems, telemedicine can improve access to specialists in tertiary centers, especially for patients who live in rural areas or have difficulties to reach the hospital [44]. Consistently, in a recent report the comparison of inperson and teletherapy revealed similar outcomes in treating PPA [45].

In the present study, at the peak of the COVID-19 epidemic, we found that almost one out of two patients experienced a significant worsening of clinical picture and quality of life since the start of social distancing and quarantine time, due to behavioral changes and language and memory dysfunctions. We also identified in one out four patients sleep disturbances interfering with both patients and their caregivers' quality of daily life already compromised by forced cohabitation all day for the pandemic lockdown. Moreover, we observed an increase in the level of anxiety among caregivers at homes, under the dual stress of fear of infection and worries about
the patients' condition, developing signs of exhaustion after about two months of full lockdown. Almost
50% of caregivers interviewed reported a moderatesevere emotional and psychological distress in the last
period after the onset of the COVID-19 epidemic.

Notably, since we also performed a survey on pos-380 sible COVID-19 symptoms, we identified one patient, 381 reporting fever and cough after recently returning 382 from Lombardy, an area with the highest prevalence 383 of the epidemic. The patient did not need hospitaliza-384 tion and healed completely at home in the following 385 2 weeks. We wrote this report in the days immedi-386 ately after the peak of COVID-19 in Italy with more 387 than 200,000 cases and more than 30,000 deaths in 388 the whole country on May 13. The survey was con-389 ducted, however, in Puglia in the South-East of Italy, 390 where the epidemic had much lower prevalence and 391 mortality, compared to Lombardy and Veneto, North-392 East Italy. This study offers a great opportunity to 393 underline to the patients and caregivers the rules and 394 recommendations to prevent virus infection in sub-395 jects at high risk of infection with negative outcomes 396 including death from COVID-19, as older adults with 397 dementia and other underlying chronic health prob-398 lems [39]. 399

Telehealth is defined as "the use of electronic 400 information and telecommunication technologies to 401 support long-distance clinical healthcare, patient and 402 professional health-related education, public health 403 and health administration" [46]. People with prior 404 known chronic neurologic diseases, including FTD, 405 continue to need support and care even if not acutely 406 ill. Telemedicine might be a well-suited instrument 407 for ongoing management of such patients, par-408 ticularly during a time when social distancing is 409 encouraged [41, 47]. 410

The use of an extensive and structured questionnaire assessing the main clinical domains in FTD and its administration by a specialist with a very high level of expertise in dementia are important strengths of the study. One of the main successes of our survey has been the prompt identification of a patient with COVID-19 hallmarks.

Strengths of our study are the relatively large 418 number of participants, considering that FTD is a 419 rare disease, the high response rate and the real-420 life recruitment of patients. Teleconsultation use 421 also permitted us to bypass the limited Internet 422 access for patients in a rural area. The area we 423 surveyed is probably representative of a popula-424 tion with low web literacy and with limited use 425

of new communication technologies. Indeed, all the patients/caregivers chose to interact via telephone or mail and refused video telemedicine because they did not have a smartphone and/or were not familiar with this type of technology. This points out that a social divide may be an important element to be considered when planning to implement advanced telemedicine systems [48]. Interestingly, all patients lived at home and none were in long-term residential facilities. This is probably linked to the characteristics of the system of social support in Southern Italy, still largely based on the family.

However, our study has some limitations, including: 1) the lack of opportunity for the telemedicine physician to perform both a physical and neurological examination; 2) the difficulties in the evaluation of some CDR-FTD subitems (as language assessment). The large majority of participants indeed did not own a computer or smartphone and we could not provide any specific support due to the outbreak restrictions. Future projects with telemedicine system and adequate devices (including video connection) would improve such assessments.

In conclusion, the COVID-19 pandemic has determined the preventive isolation of entire populations, disrupting the usual care of patients with chronic diseases. Unfortunately, Italy does not include telemedicine in the services granted by the National Health service [49]. Although no firm conclusions can be drawn given the observational design of our study, our findings with data collected during the peak of this pandemic seem to indicate that teleneurology is an effective tool for remote evaluation of patients with FTD, can serve as a safe and effective alternative to in-person care, and has a high level of satisfaction. The results of our study indeed showed a significant worsening of clinical feature and quality of life in half of patients with FTD and in a significant percentage of patients' caregivers.

Our study supports the use of telemedicine to better face both future epidemics and the usual care of complex patients. In our area, we are aiming with the project named Tecnopolo to build upon an up-to-date telemedicine system with adequate devices to supply language rehabilitation [45].

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483 SUPPLEMENTARY MATERIAL

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