Published in final edited form as:
Curr Opin Neurol. 2011 December ; 24(6): . doi:10.1097/WCO.0b013e32834cd42a.

Personality and Social Cognition in Neurodegenerative Disease

Tal Shany-Ur and **Katherine P. Rankin**
Memory and Aging Center, Department of Neurology, University of California San Francisco

Abstract

Purpose of review—Neurodegenerative diseases often cause focal damage to brain structures mediating social cognition and personality, resulting in altered interpersonal communication and behavior. We review recent research describing this phenomenon in various aspects of social cognition.

Recent findings—Corresponding to their pervasive social-emotional deficits, patients with frontotemporal dementia perform poorly on lab-based tasks including recognizing emotions, attending to salient information that guides social behavior, representing social knowledge, comprehending others' mental states, and maintaining insight to their own difficulties. Together with poor executive and regulation mechanisms, these social cognition deficits ultimately impact behavior. Patients with logopenic and nonfluent primary progressive aphasia have some deficits recognizing emotional prosody, while those with the semantic variant show more widespread deficits in social comprehension. While Alzheimer's disease patients perform poorly on some social cognition tasks, this typically reflects general cognitive impairment, and their real-life social functioning is less affected than in diseases targeting frontotemporal structures. Studies in motor diseases such as Parkinson's suggest some degradation of emotion recognition and social comprehension which should be investigated further.

Summary—We summarize recent findings concerning perception and evaluation of socioemotional information, social knowledge storage and access, advanced information processing mechanisms, and behavioral response selection and regulation across various neurodegenerative diseases.

Keywords

Neurodegenerative diseases; dementia; frontotemporal; social cognition; personality

Introduction

"Social cognition" is comprised of many psychological processes that enable an individual to participate in social interactions [1]. These include perceiving and recognizing social and emotional signals, evaluating the personal emotional relevance of everyday information, maintaining and accessing common social knowledge (e.g., norms), processing higher-order information about beliefs and intentions, and generating and selecting behavioral responses [2,3]. These processes are selectively vulnerable in certain neurodegenerative diseases with prominent altered personality and social behavior, including behavioral variant frontotemporal dementia (bvFTD) [4] and right temporal disease, variably diagnosed [5] as a variant of bvFTD or semantic variant primary progressive aphasia (svPPA) [6]. In Alzheimer's disease (AD), conversely, social cognition is relatively spared. This review will

Correspondence concerning this article should be addressed to Katherine P. Rankin, Memory and Aging Center, Department of Neurology, University of California, San Francisco, 350 Parnassus Avenue, Suite 905, San Francisco, CA 94143-1207, USA. Phone: (415) 502-0619. Fax: (415) 476-4800. krankin@memory.ucsf.edu.

summarize recent studies characterizing these changes and their neural correlates in neurodegenerative disease patients, organized by social- cognitive mechanism.

Perception of social and emotional signals

Some neurodegenerative disease patients develop deficits in the initial stages of social perception, misperceiving socially relevant visual or auditory cues, which leads to distortions in their downstream processing and response to these signals. Domain-specific social perception in the visual modality has been primarily studied using photos of facial emotions. While previous research has demonstrated impaired face perception and facial emotion recognition in patients with frontal and temporal variants of frontotemporal lobar degeneration (FTLD) compared with healthy controls (NC), a recent study used voxel based morphometry (VBM) to directly link these deficits with grey matter volume (GM) in FTLD patients. Right anterior fusiform gyrus volume predicted face perception deficits, corresponding to well-documented evidence for the fusiform face area's function, and insula volume directly corresponded with patients' ability to read angry faces [7]. bvFTD patients also perform worse than controls on a more demanding task requiring them to determine people's social emotions based on photographs of the eye area of the face [8]. When patients were asked to continuously track shifting emotions in dynamically changing facial stimuli, those with FTLD, AD, corticobasal syndrome (CBS) and progressive supranuclear palsy (PSP) showed a similar level of impairment, and poor scores related to GM loss in right lateral orbitofrontal cortex (OFC), suggesting this region may be involved in adapting behavior in response to changing perceptual input [9]. Impaired emotion recognition correlates with GM in bilateral OFC in Parkinson's disease (PD) patients as well [10]. Recent large studies of Huntington's disease (HD) patients have convincingly established that they are impaired in recognizing facial emotions, particularly anger [11], disgust, and fear [12,13], and that these deficits relate to reduced cingulate volume [14].

While decreased recognition of static emotions has often been reported in AD patients [15–17], these studies either did not control for patients' general cognitive impairment [17], or reported that emotion labeling was predicted by executive functions (EF) and MMSE scores [16]. Variable results in such studies over the past decade suggest that AD patients' failure on tests measuring facial emotion identification may often reflect general cognitive or perceptual impairment rather than a focal emotion processing deficit. Additionally, though bvFTD patients have poor recognition of eye gaze direction, this is intact in AD [17]. Some studies have found that AD patients' ability to recognize emotions displayed in more ecologically realistic, dynamic modalities does not differ from that of NCs [18]. Recent evidence from studies of altered intrinsic connectivity in the brains of AD patients suggests emotion sensitivity may actually be preserved or even heightened in AD as a result of increased salience network tone corresponding to decreased default mode network activity [19]. Alteration in AD patients' emotion sensitivity may also reflect an exaggeration of normal aging processes, in which visually-presented emotional information shifts from primarily medial temporal processing to reliance on a more widespread fronto-insular network [20]. Although decreased visual perception of social signals in AD patients with posterior cortical atrophy may not cause real-life social behavior deficits [3], recent data suggest that these patients' poor emotion recognition on testing correlates with lower self-ratings of quality of life [16]. Similar to the findings in AD, emotion identification deficits in PSP patients were not related to a specific face perception deficit but rather to general cognitive impairment [21].

With regard to auditory signal perception, primarily progressive aphasia (PPA) patients of both nonfluent (i.e., left frontal-opercular) and logopenic (left angular gyrus) subtypes show particularly poor emotion recognition from vocal prosody relative to reading facial

emotions. Processing of emotional voice prosody correlates with volume in a network of frontal, temporal, limbic and parietal regions in these patients [22]. svPPA and bvFTD patients have difficulty identifying emotions in music, a deficit which correlates with GM loss in insula, OFC, anterior cingulate cortex (ACC) and medial prefrontal cortex (PFC), as well as regions in the temporal and parietal cortices, amygdala and subcortical mesolimbic system [23]. svPPA patients also have impaired comprehension of sarcasm based specifically on (primarily vocal) paralinguistic cues [24]. Additionally, HD patients show poor recognition of negative [12] as well as positive [25] vocal emotional signals. In contrast, AD patients' ability to read emotions in both voice prosody and music is preserved [26]. Unlike FTLD patients, whose social perception in auditory modalities appears compromised, patients with AD may compensate for visual-based emotion recognition deficits with intact perception of auditory-based emotional signals, leading to preserved ability to recognize emotions presented in a realistically multimodal manner [18]. This is consistent with the hypothesis that when a social situation is perceived through multiple input channels, it can elicit the same experience even when some of the bottom-up signals are degraded [27].

Evaluating personal relevance of social and emotional signals

Most of the social behavioral deficits in bvFTD occur when these patients sustain selective degeneration of amygdala, insula, ACC, and OFC. Recent research shows this decimates a specific intrinsic functional network that acts in healthy adults to filter environmental input, rapidly identifying personally salient social signals that will be the target of sustained attention and higher-level processing [3,28]. Neuroimaging demonstrates decreased intrinsic connectivity in this "salience network" among bvFTD patients [19]. A direct consequence of damage to this network may be that bvFTD patients lose the capacity to balance reward and punishment signals, resulting in decreased sensitivity to the negative consequences of their social decisions [29]. For instance, bvFTD patients tend to judge negatively-valenced social norm violations as more acceptable than NCs do, even though the bvFTD patients could still accurately judge neutral situations or norm violations with a positive valence, a pattern associated with ventral PFC atrophy [30]. bvFTD patients may also show new-onset pathological gambling, attributed to decreased sensitivity to possible negative outcomes of risky acts [31], potentially due to lack of emotional arousal in response to punishment signals that normally guide behavior [32]. Accordingly, while NCs benefit from feedback and learn to make long-term advantageous financial choices on gambling tasks, bvFTD patients continue to make disadvantageous decisions [8]. Case illustrations show that bvFTD patients are predisposed to criminal violations, even in the context of intact knowledge of social rules, particularly when they have right anterior temporal hypometabolism [33]. Decreased attention to salient social signals might also explain some patients' interpersonal deficits. In a study using eye gaze to indicate where people direct attention in an interpersonal situation, researchers quantified shared eye gaze exhibited by participants with bvFTD, svPPA, AD, or older controls during conversations with their partners. bvFTD patients spent less time attending to their partners' eyes than NCs did, svPPA patients had more shared eye gaze than NCs, and ADs did not differ from NCs [34]. Altogether, these data suggest that many of the behavioral symptoms in bvFTD may result from failure to correctly identify social and emotional signals from their environment with a potential reward or punishment value, which normally help people avoid negative social outcomes.

Social knowledge awareness

Acquired social knowledge, in the form of sociolinguistic concepts, behavioral norms, and schemas of common social events, is essential for knowing how to behave in a social setting. Social semantic knowledge is at least partly mediated by the superior anterior temporal lobes

[35,36] and social schemas are accessed via the medial prefrontal cortex [27]. bvFTD patients, particularly those with right lateral anterior temporal damage, have a specific semantic deficit for social concepts [37]. Though studies show bvFTD patients make unusual judgments about the acceptability of social behaviors [30,38], it is unclear whether degradation of social semantic knowledge or diminished access to this knowledge prevents them from making correct decisions involving norm violations. In contrast, AD patients make social decisions similar to those made by NC and MCI participants, suggesting their access to social knowledge may be preserved [39].

Higher-order social information processing

Once social and emotional input has undergone basic perceptual processing, assuming that social knowledge is relatively intact, more advanced cognitive mechanisms are often required to perform higher-order analysis, which can be impacted by various neurodegenerative diseases [40]. One such complex social cognitive process is theory of mind (ToM), which allows accurate representation of one's own and others' beliefs, opinions, and emotions, often relying on a form of mental "time travel" in which past experiences are used to predict others' future social behavior and read their intentions. A decade of studies has demonstrated that bvFTD patients have difficulties with all aspects of ToM, exhibiting poor performance on tests of first- and second-order false belief, ToM cartoons and stories, faux pas comprehension, and reading social emotions based on photographs of eyes. Moreover, these ToM deficits remain after accounting for patients' EF deficits [41]. A recent study found that these ToM deficits caused bvFTD patients to misinterpret insincere speech (sarcasm and lies) despite normal comprehension of sincere speech. PSP patients had difficulties representing others' verbalized opinions and comprehending their intentions. AD patients, conversely, did not display deficits when controlling for their cognitive deficits (Shany-Ur et al., submitted). This pattern holds true in other studies of ToM in AD, in which patients' performance on simple "false belief" ToM tasks remains intact, while they perform poorly on more complex ToM tasks such as second-order false belief stories and reading people's eyes for social emotions. However, their deficits correlate with poor memory and EF, particularly abstract reasoning and inferential reasoning [42], thus may simply be a byproduct of general cognitive deficits rather than a focal, domain-specific deficit like in bvFTD. However, the default mode network is involved in ToM processing [43], and is vulnerable in AD [19], suggesting closer investigation of ToM in AD may be required. ToM impairments have also recently been demonstrated in early PD [44], though their deficit has not yet been clearly characterized [45]. HD patients also show deficits on both cognitive and emotional ToM tests[12,46], though it remains unclear whether these deficits result from executive dysfunction [46] or remain after accounting for intellectual and executive functioning [47].

Personal moral reasoning is another higher-order social-cognitive process that shares mechanisms with ToM. While moral reasoning is complex and depends in part on accessing social rules, personal moral decisionmaking requires predicting others' emotions and weighing the social consequences of potential choices in a moral dilemma. Though their decisions in impersonal moral dilemmas are normal, bvFTD patients tend to make more utilitarian decisions than ADs or NCs in response to personal, emotionally loaded moral dilemmas [38], and this deficit correlates with poorer emotional ToM [48], suggesting a link between personal moral reasoning and empathy.

While ToM is primarily conceptualized as the ability to accurately represent others' thoughts and emotions, it overlaps with the ability to have insight into one's own mental and emotional state. Patients with bvFTD, svPPA, AD, CBS and PSP have higher levels of alexithymia than healthy older adults, i.e., they have trouble identifying and describing their

feelings. Alexithymia correlates with right pregenual ACC, temporal, and parietal volume in older NCs [49]. Similarly, inaccurate appraisal of one's abilities (anosognosia) in neurodegenerative patients correlates with right ventral PFC volume, suggesting that diminished emotional reaction in response to signals of poor performance (e.g., errors) may contribute to impoverished self-awareness [50].

Behavioral response selection, regulation, and personality

Perhaps the best indicator of intact social cognition is the ability to enact an appropriate behavioral response to a social situation, which involves both selecting an effective behavior and performing it in an appropriately moderated manner. These processes depend in part upon executive control and top-down regulation [51]. Habitual patterns of social behavior are known as personality traits, and recent research has quantified these traits in neurodegenerative disease patients in order to identify their anatomic and clinical correlates. For instance, compared to their premorbid functioning, patients with both bvFTD and svPPA become less warm, agreeable, open to new experiences, dominant, and extraverted as their disease progresses, while AD patients primarily show progressive decreases in social dominance [52,53]. While warmth [54] and emotional empathy [51] are primarily related to right medial temporal regions and do not covary with EF in patients [55], traits such as dominance and cognitive empathy rely to a greater degree on frontal regions and EF capacities [51]. Together, anatomic studies suggest that personality changes in neurodegenerative disease patients result from imbalance across networks of frontal and temporal structures, in which specific traits emerge from complex functional patterns involving both preserved and damaged regions [53,54]. The relationship between social behavior and EF was further elucidated in a study demonstrating that while patients' degree of socio-emotional disinhibition was predicted by primarily right OFC thickness, their cognitive control was mediated by separate dorsolateral PFC structures [56].

An effective social response often requires regulation and modulation of the initial emotional reaction. A recent study induced a startle response in patients with AD and FTLD and controls to examine their spontaneous emotion regulation. When subjects were not warned that a startling stimulus would occur, all groups showed a similar immediate emotional reaction on their faces; however, when forewarned, FTLDs showed less regulation of immediate emotional expression than ADs or NCs, suggesting less spontaneous self-monitoring. Finally, when forewarned and explicitly asked to down-regulate their facial response, both FTLD and AD patients showed less regulation of their emotional reaction than NCs. The authors hypothesized this pattern may reflect a loss of top-down executive regulation in AD but decreased monitoring of bottom-up emotional signals in FTLD [57]. Similarly, another study showed that increased neuroticism in FTLD, which reflects impaired emotional regulation, is correlated to GM loss in OFC and ACC regions [53]. Finally, PSP patients may show disinhibited social behavior as well, presumed to reflect executive impairment [58].

Summary and Conclusions

While it is well-known that bvFTD patients exhibit extensive real-life social dysfunction, recent studies have elucidated the underlying social cognitive deficits, including impaired recognition of primary emotional signals, decreased attention to relevant "warning signs" about potential negative consequences, decreased social knowledge, and inability to represent their own and others' perspectives and emotions. These social cognitive impairments combine with a dysexecutive syndrome and poor emotional and behavioral regulation to result in aberrant behavior. New research clarifying social cognition deficits in other patients with neurodegenerative syndromes have revealed that lvPPA and nfPPA

patients have selective deficits reading emotion from vocal prosody, while svPPA patients demonstrate more widespread deficits in social comprehension. Recent research has also shown that while AD patients may fail tests of social cognition, this often occurs as a result of general cognitive deficits, but that these patients have very few focal deficits in social cognition, and may actually develop a paradoxically heightened sense of social and emotional salience leading to temporarily increased social sensitivity. Studies also suggest that HD and PD patients have impairments in recognizing emotional signals, though studies of advanced social-cognitive processing in these and other motor-disordered patients are still needed.

Acknowledgments

The authors have received funding in the form of research grants from NIH during the time this article was written, including 1R01 AG029577, HHSN271200623661C, and P01 AG019724.

References and recommended reading

1. Frith CD. Social cognition. Philos Trans R Soc Lond B Biol Sci. 2008; 363:2033–2039. [PubMed: 18292063]
2. Adolphs R. Conceptual challenges and directions for social neuroscience. Neuron. 2010; 65:752–767. [PubMed: 20346753]
3. Sollberger M, Rankin KP, Miller BL. Social Cognition. Continuum Lifelong Learning Neurol. 2010; 16:69–85. * This review exemplifies the different social-cognitive mechanisms using case illustrations of neurodegenerative diseases patients.
4. Neary D, Snowden JS, Gustafson L, et al. Frontotemporal lobar degeneration: a consensus on clinical diagnostic criteria. Neurology. 1998; 51:1546–1554. [PubMed: 9855500]
5. Josephs KA, Whitwell JL, Knopman DS, et al. Two distinct subtypes of right temporal variant frontotemporal dementia. Neurology. 2009; 73:1443–1450. [PubMed: 19884571]
6. Gorno-Tempini ML, Hillis AE, Weintraub S, et al. Classification of primary progressive aphasia and its variants. Neurology. 2011; 76:1006–1014. [PubMed: 21325651]
7. Omar R, Rohrer JD, Hailstone JC, Warren JD. Structural neuroanatomy of face processing in frontotemporal lobar degeneration. J Neurol Neurosurg Psychiatry. 2010 * This study examines the neural correlates of facial emotional processing in FTLD, providing patient-based evidence for brain-behavior relationships underlying visual emotional perception.
8. Torralva T, Roca M, Gleichgerrcht E, et al. A neuropsychological battery to detect specific executive and social cognitive impairments in early frontotemporal dementia. Brain. 2009; 132:1299–1309. [PubMed: 19336463]
9. Goodkind MS, Sollberger M, Gyurak A, et al. Tracking emotional valence: The role of the orbitofrontal cortex. Hum Brain Mapp. 2011
10. Ibarretxe-Bilbao N, Junque C, Tolosa E, et al. Neuroanatomical correlates of impaired decision-making and facial emotion recognition in early Parkinson's disease. Eur J Neurosci. 2009; 30:1162–1171. [PubMed: 19735293]
11. Henley SM, Novak MJ, Frost C, et al. Emotion recognition in Huntington's disease: A systematic review. Neurosci Biobehav Rev. 2011
12. Calder AJ, Keane J, Young AW, et al. The relation between anger and different forms of disgust: implications for emotion recognition impairments in Huntington's disease. Neuropsychologia. 2010; 48:2719–2729. [PubMed: 20580641]
13. Ille R, Holl AK, Kapfhammer HP, et al. Emotion recognition and experience in Huntington's disease: Is there a differential impairment? Psychiatry Res. 2011
14. Hobbs NZ, Pedrick AV, Say MJ, et al. The structural involvement of the cingulate cortex in premanifest and early Huntington's disease. Mov Disord. 2011
15. Wiechetek Ostos M, Schenk F, Baenziger T, von Gunten A. An Exploratory Study on Facial Emotion Recognition Capacity in Beginning Alzheimer's Disease. Eur Neurol. 2011; 65:361–367. [PubMed: 21625142]

16. Phillips LH, Scott C, Henry JD, et al. Emotion perception in Alzheimer's disease and mood disorder in old age. Psychol Aging. 2010; 25:38–47. [PubMed: 20230126]

17. Bediou B, Ryff I, Mercier B, et al. Impaired social cognition in mild Alzheimer disease. J Geriatr Psychiatry Neurol. 2009; 22:130–140. [PubMed: 19321881]

18. Henry JD, Ruffman T, McDonald S, et al. Recognition of disgust is selectively preserved in Alzheimer's disease. Neuropsychologia. 2008; 46:1363–1370. [PubMed: 18241898]

19. Zhou J, Greicius MD, Gennatas ED, et al. Divergent network connectivity changes in behavioural variant frontotemporal dementia and Alzheimer's disease. Brain. 2010; 133:1352–1367. [PubMed: 20410145] ** This fMRI study showed that in bvFTD, intrinsic connectivity increases in the salience network and decreases in the default mode network, while in AD this pattern is reversed.

20. Fischer H, Nyberg L, Backman L. Age-related differences in brain regions supporting successful encoding of emotional faces. Cortex. 2010; 46:490–497. [PubMed: 19560133]

21. Ghosh BC, Rowe JB, Calder AJ, et al. Emotion recognition in progressive supranuclear palsy. J Neurol Neurosurg Psychiatry. 2009; 80:1143–1145. [PubMed: 19762901]

22. Rohrer JD, Sauter D, Scott S, et al. Receptive prosody in nonfluent primary progressive aphasias. Cortex. 2010 * This study describes relationships between grey matter volume of specific structures and ability to detect different emotions through prosodic auditory signals.

23. Omar R, Henley SM, Bartlett JW, et al. The structural neuroanatomy of music emotion recognition: evidence from frontotemporal lobar degeneration. Neuroimage. 2011; 56:1814–1821. [PubMed: 21385617]

24. Rankin KP, Salazar A, Gorno-Tempini ML, et al. Detecting sarcasm from paralinguistic cues: anatomic and cognitive correlates in neurodegenerative disease. Neuroimage. 2009; 47:2005–2015. [PubMed: 19501175]

25. Robotham L, Sauter DA, Bachoud-Levi AC, Trinkler I. The impairment of emotion recognition in Huntington's disease extends to positive emotions. Cortex. 2011; 47:880–884. [PubMed: 21429482]

26. Drapeau J, Gosselin N, Gagnon L, et al. Emotional recognition from face, voice, and music in dementia of the Alzheimer type. Ann N Y Acad Sci. 2009; 1169:342–345. [PubMed: 19673804]

27. Krueger F, Barbey AK, Grafman J. The medial prefrontal cortex mediates social event knowledge. Trends Cogn Sci. 2009; 13:103–109. [PubMed: 19223228]

28. Seeley WW, Crawford RK, Zhou J, et al. Neurodegenerative diseases target large-scale human brain networks. Neuron. 2009; 62:42–52. [PubMed: 19376066]

29. Seeley WW, Zhou J, Kim EJ. Frontotemporal Dementia: What Can the Behavioral Variant Teach Us about Human Brain Organization? Neuroscientist. 2011 * This is a comprehensive review of bvFTD symptoms, neuroanatomic correlates, and neuropathology, including information on salience network dysfunction and changes to Von Economo neurons.

30. Grossman M, Eslinger PJ, Troiani V, et al. The role of ventral medial prefrontal cortex in social decisions: converging evidence from fMRI and frontotemporal lobar degeneration. Neuropsychologia. 2010; 48:3505–3512. [PubMed: 20691197] * This study showed a greater tendency in bvFTD patients to find acts violating social norms to be acceptable, and related this to ventromedial PFC structures.

31. Manes FF, Torralva T, Roca M, et al. Frontotemporal dementia presenting as pathological gambling. Nat Rev Neurol. 2010; 6:347–352. [PubMed: 20440289]

32. Damasio AR. The somatic marker hypothesis and the possible functions of the prefrontal cortex. Philos Trans R Soc Lond B Biol Sci. 1996; 351:1413–1420. [PubMed: 8941953]

33. Mendez MF. The unique predisposition to criminal violations in frontotemporal dementia. J Am Acad Psychiatry Law. 2010; 38:318–323. [PubMed: 20852216] * This case review discusses the prevalence of criminal behavior in bvFTD, its origins and implications.

34. Sturm VE, McCarthy ME, Yun I, et al. Mutual gaze in Alzheimer's disease, frontotemporal and semantic dementia couples. Soc Cogn Affect Neurosci. 2011; 6:359–367. [PubMed: 20587598] ** This study used a quantitative approach to characterize decreased gaze direction towards significant others, during interpersonal conversations, in bvFTD patients, providing evidence for impaired attention to salient socio-emotional information in this disease.

35. Zahn R, Moll J, Krueger F, et al. Social concepts are represented in the superior anterior temporal cortex. Proc Natl Acad Sci U S A. 2007; 104:6430–6435. [PubMed: 17404215]
36. Ross LA, Olson IR. Social cognition and the anterior temporal lobes. Neuroimage. 2010; 49:3452–3462. [PubMed: 19931397]
37. Zahn R, Moll J, Iyengar V, et al. Social conceptual impairments in frontotemporal lobar degeneration with right anterior temporal hypometabolism. Brain. 2009; 132:604–616. [PubMed: 19153155]
38. Mendez MF, Shapira JS. Altered emotional morality in frontotemporal dementia. Cogn Neuropsychiatry. 2009; 14:165–179. [PubMed: 19499384]
39. Bosch-Domenech A, Nagel R, Sanchez-Andres JV. Prosocial capabilities in Alzheimer's patients. J Gerontol B Psychol Sci Soc Sci. 2010; 65B:119–128. [PubMed: 19468059]
40. Gleichgerrcht E, Ibanez A, Roca M, et al. Decision-making cognition in neurodegenerative diseases. Nat Rev Neurol. 2010; 6:611–623. [PubMed: 21045795]
41. Adenzato M, Cavallo M, Enrici I. Theory of mind ability in the behavioural variant of frontotemporal dementia: an analysis of the neural, cognitive, and social levels. Neuropsychologia. 2010; 48:2–12. [PubMed: 19666039] * This review summarizes existing literature concerning theory of mind deficits in bvFTD.
42. Castelli I, Pini A, Alberoni M, et al. Mapping levels of theory of mind in Alzheimer's disease: a preliminary study. Aging Ment Health. 2011; 15:157–168. [PubMed: 21140304]
43. Spreng RN, Grady CL. Patterns of brain activity supporting autobiographical memory, prospection, and theory of mind, and their relationship to the default mode network. J Cogn Neurosci. 2010; 22:1112–1123. [PubMed: 19580387]
44. Roca M, Torralva T, Gleichgerrcht E, et al. Impairments in social cognition in early medicated and unmedicated Parkinson disease. Cogn Behav Neurol. 2010; 23:152–158. [PubMed: 20829664]
45. Freedman M, Stuss DT. Theory of Mind in Parkinson's disease. J Neurol Sci. 2011
46. Allain P, Havet-Thomassin V, Verny C, et al. Evidence for deficits on different components of theory of mind in Huntington's disease. Neuropsychology. 2011
47. Brune M, Blank K, Witthaus H, Saft C. "Theory of mind" is impaired in Huntington's disease. Mov Disord. 2011; 26:671–678. [PubMed: 21384426]
48. Gleichgerrcht E, Torralva T, Roca M, et al. The role of social cognition in moral judgment in frontotemporal dementia. Soc Neurosci. 2011; 6:113–122. [PubMed: 20706963]
49. Sturm VE, Levenson RW. Alexithymia in neurodegenerative disease. Neurocase. 2011; 17:242–250. [PubMed: 21432723]
50. Rosen HJ. Anosognosia in neurodegenerative disease. Neurocase. 2011; 17:231–241. [PubMed: 21667396]
51. Eslinger PJ, Moore P, Anderson C, Grossman M. Social cognition, executive functioning, and neuroimaging correlates of empathic deficits in frontotemporal dementia. J Neuropsychiatry Clin Neurosci. 2011; 23:74–82. [PubMed: 21304142]
52. Sollberger M, Neuhaus J, Ketelle R, et al. Interpersonal traits change as a function of disease type and severity in degenerative brain diseases. J Neurol Neurosurg Psychiatry. 2011; 82:732–739. [PubMed: 21172858]
53. Mahoney CJ, Rohrer JD, Omar R, et al. Neuroanatomical profiles of personality change in frontotemporal lobar degeneration. Br J Psychiatry. 2011; 198:365–372. [PubMed: 21372059]
54. Sollberger M, Stanley CM, Wilson SM, et al. Neural basis of interpersonal traits in neurodegenerative diseases. Neuropsychologia. 2009; 47:2812–2827. [PubMed: 19540253]
55. Sollberger M, Stanley CM, Ketelle R, et al. Neuropsychological correlates of dominance, warmth, and extraversion in neurodegenerative disease. Cortex. 2011
56. Krueger CE, Laluz V, Rosen HJ, et al. Double dissociation in the anatomy of socioemotional disinhibition and executive functioning in dementia. Neuropsychology. 2011; 25:249–259. [PubMed: 21381829]
57. Goodkind MS, Gyurak A, McCarthy M, et al. Emotion regulation deficits in frontotemporal lobar degeneration and Alzheimer's disease. Psychol Aging. 2010; 25:30–37. [PubMed: 20230125]

58. Kertesz A, McMonagle P. Behavior and cognition in corticobasal degeneration and progressive supranuclear palsy. J Neurol Sci. 2010; 289:138–143. [PubMed: 19733862]

Key Points

- Recent studies have begun to more precisely link the socioemotional changes in neurodegenerative diseases with specific neuroanatomic structures and networks, particularly in the medial temporal, medial frontal, and ventral frontal regions.
- bvFTD patients show early, focal deficits in most aspects of social cognition, while patients with AD and other neurodegenerative diseases often fail social cognition tests due to general cognitive decline, showing relative preservation of specific socioemotional functions until late in their disease.
- Aphasia patients such as those with lvPPA and nfPPA evidence focal deficits in aspects of social perception relating to language systems, such as decoding emotional prosody, while svPPA patients show more widespread alteration of social cognition and behavior.
- Recent large-scale studies have confirmed that HD patients show altered emotion recognition and regulation, and new research suggests they may also have theory of mind deficits.
- Some focal deficits in social cognition may occur in patients with other neurodegenerative diseases traditionally viewed as primary motor disorders, such as PD and PSP, but additional investigation of this phenomenon is needed.