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Emotion Regulation Deficits in Frontotemporal Lobar Degeneration and Alzheimer's Disease

Madeleine S. Goodkind, Anett Gyurak, and Megan McCarthy
University of California, Berkeley

Bruce L. Miller
University of California, San Francisco

Robert W. Levenson
University of California, Berkeley

We examined instructed and spontaneous emotion regulation in patients with frontotemporal lobar degeneration (FTLD, $N = 32$), which presents with profound emotional and personality changes; patients with Alzheimer's disease (AD, $N = 17$), which presents with profound memory impairment; and neurologically normal controls ($N = 25$). Participants were exposed to an aversive acoustic startle stimulus (115 dB) under 3 different conditions: (a) unwarned without instructions to down-regulate, (b) warned without instructions to down-regulate, and (c) warned with instructions to down-regulate. In the last 2 conditions, the warning took the form of a 20-s countdown. In all conditions, visible aspects of the startle response were assessed by measuring overall somatic activity and coding emotional facial expressions. FTLD patients, AD patients, and control participants showed similar patterns of down-regulation in somatic activity across the 3 startle trials. However, differences between the 3 groups emerged in the amount of emotional facial behavior expressed in the startle trials. There were no group differences in response in the unwarned condition, indicating that the startle response was intact in the patients. In the warned with instructions condition, both FTLD and AD patients were moderately impaired in down-regulatory ability compared with controls. In the warned without instructions condition, AD patients and normal controls spontaneously down-regulated their emotional responses, but FTLD patients did not. These findings illuminate specific problems that these patients have in the emotional realm.

Keywords: emotion, emotion regulation, frontotemporal dementia, Alzheimer's disease, dementia

The ability to regulate emotions is a critical aspect of emotional functioning. Adjusting emotional responses so that they are appropriate to the situation is crucially linked to social adeptness and to the ability to get along with others (Keltner & Kring, 1998). Specifically, as human beings, we often adjust or regulate our emotional expressions in the service of communicating our emotions. People deploy a combination of controlled and automatic processes to maintain, amplify, or reduce their emotional responses (Gross & Thompson, 2007). For example, we feign interest when bored, control anger when provoked, hide reactions to disappointments, and diminish fearful responses when threatened. The intricate process of modulating complex emotional responses draws upon a wide range of abilities, including allocating attention to environmental cues, self-monitoring, and controlling behavior. These processes rely on the integrity of a diverse set of neural circuits. Given the complexity of skills and neural circuits involved, it is not surprising that emotion regulatory difficulties are observed in a large number of neurological (and psychiatric) conditions.

One of the first descriptions of deficits in emotion regulation following brain damage was the case of Phineas Gage (Harlow, 1848), who exhibited clear signs of emotion dysregulation after suffering a severe injury in which a tamping iron penetrated his skull and damaged his frontal lobes. Since Gage, the belief that the frontal lobes play a critical role in emotion regulation has been foundational in affective neuroscience (Davidson, Fox, & Kalin, 2007; Ochsner & Gross, 2007), deriving support from converging evidence from imaging (Goldin, McRae, Ramel, & Gross, 2008; Ochsner et al., 2004) and brain injury studies (Bechara, 2005).

Brain damage that affects emotion regulation comes in many forms (e.g., focal lesions, traumatic brain injuries, and vascular events). Neurodegenerative diseases (e.g., Alzheimer's disease [AD] and frontotemporal lobar degeneration [FTLD]) can also cause circumscribed loss of brain tissue. Indeed, clinicians and caregivers often report problems with emotion regulation in patients with these disorders; however, precise characterization of

Madeleine S. Goodkind, Anett Gyurak, Megan McCarthy, and Robert W. Levenson, Department of Psychology, University of California, Berkeley; Bruce L. Miller, Department of Neurology, University of California, San Francisco.

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Correspondence concerning this article should be addressed to Robert W. Levenson, Department of Psychology, University of California, Berkeley, 3210 Tolman Hall, #1650, Berkeley, CA 94720-1650. E-mail: boblev@berkeley.edu

these deficits has been rare. The present study applies laboratory methods derived from basic affective science (Levenson et al., 2007) to study deficits in emotion regulation in AD and FTLD.

Emotion Regulation in Normal Aging

Because AD and FTLD typically occur in middle to late adulthood, changes in cognitive and emotional functioning must be evaluated against the backdrop of the normal aging process. Although declines in many areas of cognitive and physical functioning have been documented as part of normal aging (Salthouse, 2004), emotional functioning, particularly emotion regulation, may be relatively spared (Levenson, 2000). In a study using survey methodology (Gross et al., 1997), older adults reported being better able to control their emotions than earlier in life. In one of the few laboratory studies of emotion regulation and age, we (Kunzmann, Kupperbusch, & Levenson, 2005) found no difference between healthy elderly and young adults in ability to comply with instructions to amplify or suppress emotional expression in response to disgust-eliciting film clips.

Emotional Functioning in AD and FTLD

Among older adults, AD and FTLD together make up 65% of neurodegenerative diseases (Cummings & Benson, 1992; Rosen et al., 2002). Each disease is associated with different patterns of behavioral change, reflecting differences in underlying neuropathology.

AD accounts for over 50% of dementia cases (Cummings & Benson, 1992; Rosen et al., 2002). In AD, neurodegeneration is most prominent in the hippocampus, temporal lobe, entorhinal cortex, precuneus, and posterior cingulate cortex (Braak & Braak, 1991; Greicius, Srivastava, Reiss, & Menon, 2004; Minoshima et al., 1997) Frontal brain areas are largely spared in the early stages of the disease but often become involved in later stages (Tikofsky, Hellman, & Parks, 1993), with attendant deficits in executive functioning (Perry & Hodges, 1999). AD is characterized primarily by cognitive symptoms, including deterioration of memory and visuospatial function (Katzman, 1986; McKhann et al., 1984; Mendez, Mastri, Sung, & Frey, 1990). Emotional behavior and social abilities remain relatively intact in the early stages of AD (Varma et al., 1999). For example, AD patients show relatively preserved abilities to recognize emotions in photographs (Bucks & Radford, 2004; Lavenu, Pasquier, Lebert, Petit, & Van der Linden, 1999).

FTLD accounts for 15% of all neurodegenerative diseases (Rosen et al., 2002) and may be as common as AD in patients under the age of 65 (Ratnavalli, Brayne, Dawson, & Hodges, 2002). In FTLD, deterioration is most prominent in frontal and anterior temporal regions (Kertesz, Davidson, & Munoz, 1999; Neary et al., 1998). FTLD is characterized primarily by behavioral symptoms, including dysregulated social and personal conduct and emotional unconcern (Neary et al., 1998). Clinically, FTLD patients are often described as disinhibited, impulsive, and emotionally indifferent (Harciarek & Jodzio, 2005). Cognitive functions such as memory are relatively preserved in the early stages of FTLD (Rascovsky et al., 2002). Prior experimental investigations of emotional functioning in FTLD indicate that emotional reactivity to simple emotional stimuli (such as sudden loud noises and emotional films with very simple themes) is relatively preserved in the early stages of this disease (Sturm, Rosen, Allison, Miller, & Levenson, 2006; Werner et al., 2007), however, deficits are found in more complex and self-referential forms of emotional reactivity (e.g., embarrassment; Sturm et al., 2006) and in recognizing the emotions of others (Werner et al., 2007).

To our knowledge, there have been no prior laboratory evaluations of emotion regulation in AD or FTLD patients, although clinician and caregiver observations suggest that FTLD patients have difficulties regulating emotion. For example, FTLD patients have been described as disinhibited or impulsive (Bozeat, Gregory, Ralph, & Hodges, 2000), and these patients often engage in socially inappropriate behaviors (Levenson & Miller, 2007). The symptoms suggest a lack of behavioral control, and they may arise due to impaired emotion regulatory abilities. Moreover, patterns of neurodegeneration in FTLD typically involve frontal brain structures (Levenson & Miller, 2007) that are thought to be critically involved in emotion regulation (Davidson et al., 2007; Ochsner & Gross, 2007). There is reason to expect greater preservation of emotion regulation in AD than FTLD, in part because frontal brain regions critical for emotion regulation are relatively preserved in the early stages of AD. Consistent with this, clinician and caregiver reports suggest that emotional functioning is comparatively preserved in the early stages of AD (Bozeat et al., 2000).

The Current Study

The goal of the current study was to assess several aspects of emotion regulation in a sample of FTLD patients, AD patients, and neurologically normal age-matched controls. We used emotion regulation tasks that are part of a comprehensive battery we have developed for assessing emotional functioning in neurological patients (Levenson et al., 2007). An acoustic startle stimulus known to elicit a strong emotional response (Ekman, Friesen, & Simons, 1985; Hagemann, Levenson, & Gross, 2006) was administered under three different conditions: (a) without warning and without instructions to down-regulate; (b) with warning and without instructions to down-regulate; and (c) with warning and with explicit instructions to down-regulate. On the basis of our previous research using the startle stimulus (Sturm et al., 2006) and the neural underpinnings of the startle response, which involves brainstem circuits (Davis, Gendelman, Tischler, & Gendelman, 1982) that are relatively spared in the early stages of FTLD and AD, we did not expect group differences in the unwarned condition. On the basis of clinician and caregiver reports suggesting a lack of emotional changes in AD (Harciarek & Jodzio, 2005), we did not expect to find deficits in patients with AD in the two regulation conditions (warned without instructions to down-regulate and warned with instructions to down-regulate). However, we did expect patients with FTLD to show deficits in emotion regulation ability due to the degeneration of critical frontal regions (Rosen et al., 2005).

Method

Participants

FTLD and AD patients were recruited through the Memory and Aging Center at the University of California, San Francisco. Di-

agnoses were determined using clinical interviews, rating scales, structural MRIs of the brain, and neuropsychological tests. FTLD was diagnosed using the Neary clinical criteria (Neary et al., 1998). AD was diagnosed using the criteria of the National Institute of Neurological and Communication Diseases and Stroke/Alzheimer's Disease and Related Disorders Association (McKhann et al., 1984). Normal controls were recruited through newspaper ads and confirmed to have no neurological or psychiatric conditions. Thirty-two FTLD patients, 17 AD patients, and 25 controls were studied. Analyses of variance revealed significant age differences between the groups, $F(2, 71) = 3.65$, $p = .03$. Post hoc analyses revealed that controls were significantly older than FTLD and AD patients; consequently, age was included as a covariate in all analyses. There were no differences among the groups in years of education, $F(2, 71) = 1.47$, *ns*, or sex, $\chi^2(2, N = 74) = 3.51$, *ns*. General functioning was assessed using the Mini Mental State Exam (MMSE; Folstein, Folstein, & McHugh, 1975), a brief measure of overall cognitive functioning. As expected, there were significant differences in Mini Mental State Exam scores between the three groups, $F(2, 71) = 28.33$, $p < .01$; post hoc analyses revealed that all three groups significantly differed from one another (all ps $< .01$). Control participants scored near the highest possible score, indicating no cognitive impairments. Patients with FTLD scored in the range of no to mild cognitive impairment. Patients with AD were more impaired than either the control participants or the FTLD patients, scoring in the range of mild to moderate impairment. Demographic data are summarized in Table 1.

Procedure

A day-long laboratory procedure designed to provide a comprehensive assessment of emotional functioning (Levenson, 2007) was conducted at the Berkeley Psychophysiology Laboratory. Upon arrival, participants and their caregivers signed consent forms (approved by the Committee for the Protection of Human Subjects at the University of California, Berkeley) that delineated the experimental tasks (including hearing a loud noise). The present article focuses on the data obtained using the three variants of the acoustic startle, a task that took place approximately 2 hr into the 6-hr evaluation. Participants were seated in a comfortable chair in a well-lit 3 × 6 m room, and physiological recording devices were attached. All participants received $30 for their participation.

Startle Trials

Visual stimuli and instructions were presented on a 21-in. (53-cm) television screen placed 1.75 m from the participant. Acoustic startle stimuli were administered through two loudspeakers located behind the participant's head. The acoustic startle stimulus was a 115-dB, 100-ms burst of white noise, which can be likened to a gunshot. Baseline data were collected for 1 min before each trial and 2 min after each trial. A self-report emotion inventory (described below) followed each posttrial baseline.

The startle stimulus was presented in three different ways on separate trials.

Unwarned without instructions to down-regulate. The trial began with a 1-min baseline in which an *X* was presented on the television screen and participants were instructed to relax. The acoustic startle stimulus was then presented without warning. This was followed by a 2-min poststartle period during which the *X* reappeared on the screen. This condition was included to assess response magnitude when there is no opportunity for prestimulus down-regulation.

Warned without instructions to down-regulate. At the start of the trial, participants were informed that the startle stimulus would be presented at the end of a countdown. Following a 1-min baseline in which an *X* was presented on the television screen and participants were instructed to relax, a 20-s countdown from 10 to 0 was presented via the television's screen and speaker. The acoustic startle stimulus was then presented, followed by a 2-min poststartle period during which the *X* reappeared on the screen. Because no instructions to regulate were given, this condition assesses spontaneous emotion regulation. Prior studies show that people typically down-regulate their motor response spontaneously under these conditions (Ekman et al., 1985; Hagemann et al., 2006; Keltner & Ekman, 1996).

Warned with instructions to down-regulate. The trial was identical to the warned without instructions to down-regulate trial except that participants received the additional instruction to "hide your emotional reactions during the trial" and to "pretend someone is watching you and you don't want him to know how you feel." Following the 1-min baseline and prior to the onset of the 20-s countdown, participants were reminded (on the television's screen and speakers) to hide their emotions. This condition assesses capacity to down-regulate when instructed.

Because of difficulties inherent in recruiting large numbers of patients for these kinds of studies and advantages inherent in using participants as their own controls, we adopted a within-subject

Table 1
Participant Demographic Data

Variable	Normal aging control ($n = 25$)	AD ($n = 17$)	FTLD ($n = 32$)	Test statistics
Males	13	12	24	$\chi^2(2, N = 74) = 3.51$, *ns*
Age (*SD*)	66.74_a (8.34)	61.12_b (8.65)	61.39_b (7.84)	$F(2, 71) = 3.65$, $p < .05$
Education (*SD*)	17.48 (2.21)	16.00 (4.03)	16.84 (2.24)	$F(2, 71) = 1.47$, *ns*
MMSE	29.72_a (.46)	19.94_b (6.59)	25.50_c (4.07)	$F(2, 71) = 28.33$, $p < .01$

Note. Groups with different subscripts differed from each other at $p < .05$. FTLD = frontotemporal lobar degeneration; AD = Alzheimer's disease; MMSE = Mini-Mental State Examination; *ns* = not significant ($p > .05$).

design in which all participants were exposed to the startle under all three conditions. The nature of the conditions dictated that we use a fixed order of presentation. The unwarned condition had to come first so that this startle would be completely unexpected. The warned with instructions to down-regulate regulation condition had to come last so that there would be no carryover of the instructional set to the other conditions.

Measures of Startle Response

Although a number of peripheral physiological measures were obtained from each participant, the emphasis of the present study was on regulation of visible features of the startle response. Thus, we focused specifically on emotional facial behavior and overall somatic activity.

Emotional facial behavior. A frontal view of the participant's face and torso was recorded using a remotely controlled high-resolution video camera partially concealed behind darkened glass and embedded in a bookshelf. A team of trained research assistants used the Expressive Emotional Behavior Coding System (Gross & Levenson, 1993) to code emotional facial expressions. The Expressive Emotional Behavior Coding System is based on the Facial Action Coding System (Ekman & Friesen, 1976) and uses descriptions of specific facial actions to assign emotion codes on a second-by-second basis. Following procedures we have used in other studies with the acoustic startle (Hagemann, Levenson, & Gross, 2006; Soto, Levenson, & Ebling, 2005), the coding epoch for startle-related behaviors was set at 5 s, beginning with the onset of the startle stimulus. Coders, who were blind to the diagnostic status of the participant as well as to the particular startle trial they were coding, watched the videotapes without sound. Coders rated 10 different kinds of emotional expression (anger, contempt, confusion, disgust, fear, happiness, embarrassment, interest, sadness, and surprise) each second during the 5-s startle coding epoch using a 4-point intensity scale (*none*, *slight*, *moderate*, and *strong*). Interrater reliability among the coders for the full set of codes was high (intraclass correlation coefficient = .79). To obtain an index of overall facial emotional response and to encompass individual differences in the particular emotions shown (Ekman et al., 1985), we created an emotional facial behavior composite by averaging across all 10 emotion codes.

Somatic activity. A transducer attached to a platform under the participants' chair was used to measure overall body movement. The transducer generated an electrical signal proportional to the amount of movement in any direction. The electrical activity was amplified using a Grass Model 7 polygraph (Grass Technologies, West Warwick, RI) and averaged each second in arbitrary units by a computer program written by one of the authors (Robert W. Levenson). To parallel the epoch used with facial data, we calculated a somatic activity score by averaging over the 5-s period, beginning with the startle stimulus.

Results

Manipulation Check: Down-Regulation in the Warned Conditions

To establish that down-regulation was in fact the normative response in the warned without instructions to down-regulate and warned with instructions to down-regulate conditions, we used paired t tests to compare mean level of emotional facial and somatic response between each of the warned trials and the unwarned trial, using data only from the control participants (the two patient groups were not included). These analyses revealed significantly smaller response for the warned without instructions to down-regulate trial compared with the unwarned trial for both emotional facial expression, $t(24) = -3.84$, $p < .002$, and somatic activity, $t(24) = -3.86$, $p < .002$. Additionally, control participants showed significantly smaller responses for the warned with instructions to down-regulate trial compared with the unwarned trial for both emotional facial expression, $t(24) = -4.69$, $p < .001$, and somatic activity, $t(24) = -3.73$, $p < .002$. Comparing the two warned trials, there was a trend in the direction of less emotional facial behavior in the warned with instructions to down-regulate trial compared with the warned without instructions to down-regulate trial, $t(24) = -2.04$, $p < .06$, but no differences in somatic activity, $t(24) = -0.36$, *ns*.

Subjective Emotional Response

After each startle trial, participants used a 3-point scale (*none*, *a little*, *a lot*) to describe how much of each of seven emotions (afraid, angry, disgusted, embarrassed, happy, sad, and surprised) they felt in response to the loud noise. In response to the unwarned startle stimulus, the most commonly reported emotions were "surprised" and "afraid," with 69 (93%) of 74 participants reporting they felt surprised and 34 (46%) of 74 participants reporting that they felt afraid. The percentage of participants reporting they felt surprised or afraid did not differ across the three diagnostic groups, $\chi^2(2, N = 74) = 3.21$, *ns*, and $\chi^2(2, N = 74) = 1.16$, *ns*, for surprised and afraid, respectively. Thus, the prevalence of the two most commonly reported emotions in response to the unwarned startle did not differ by disease status. Examining the other two startle conditions, in the warned without instructions to down-regulate trial, 38% of participants reported feeling surprised; and in the warned with instructions to down-regulate trial, 32% reported feeling surprised. These percentages did not differ significantly across the three diagnostic groups in either the warned without instructions to down-regulate trial, $\chi^2(2, N = 74) = 2.17$, *ns*, or the warned with instructions to down regulate trial, $\chi^2(2, N = 74) = 1.25$, *ns*.

Analytic Strategy

Emotional facial expression and somatic activity were compared separately using a 3 × 2 × 3 (Diagnostic Group × Sex × Startle Trial) analysis of covariance (ANCOVA). In these analyses diagnostic group and sex were treated as between-subjects factors and startle trial was treated as a within-subject factor. Age, which had been found to differ between the diagnosis groups, was included as a covariate. Significant main effects were followed up using pairwise comparisons. Because there were no significant main effects for sex or significant interactions involving sex, we collapsed across sex in all subsequent analyses. Significant interactions of diagnostic group by startle trial were followed up using separate one-way (diagnostic group) ANCOVAs for each of the three startle trials. Significant main effects for diagnostic group in these ANCOVAs were followed up using least significant difference tests

(adjusted for multiple comparisons). Partial eta squares representing the portion of explained variance in the dependent variable are reported for all ANCOVA effects.

Emotional Facial Behavior

The ANCOVA revealed no significant main effects for diagnostic group, $F(2, 67) = 1.36$, $p = .26$, $\eta^2 = .04$; sex, $F(1, 67) = 1.54$, $p = .22$, $\eta^2 = .02$; or startle trial, $F(2, 134) = 1.36$, $p = .26$, $\eta^2 = .02$. There was a significant interaction between diagnostic group and startle trial, $F(4, 140) = 3.66$, $p < .01$, $\eta^2 = .10$, indicating that the three diagnostic groups did not show the same pattern of emotional facial behavior across the three startle trials. This interaction was decomposed using one-way ANCOVAs for each startle trial.[1] Covariate corrected means and standard errors are presented in Table 2.

Unwarned without instructions to down-regulate. The main effect for diagnostic group was not significant, $F(2, 70) < 1$, $\eta^2 = .018$.[2] Thus, our expectation that unregulated emotional reactivity would not differ between groups was confirmed.

Warned without instructions to down-regulate. The main effect for diagnostic group was significant, $F(2, 70) = 5.00$, $p < .01$, $\eta^2 = .13$. Follow-up least significant difference analyses revealed that FTLD patients showed less down-regulation (i.e., more emotional facial behavior) than AD patients ($p = .01$) and normal controls ($p = .01$). AD patients did not differ from normal aging participants ($p > .05$). These results are consistent with our hypotheses that FTLD patients would show impaired spontaneous emotion down-regulation and that AD patients would not be impaired. See Figure 1.

Warned with instructions to down-regulate trial. The main effect for diagnostic group was significant, $F(2, 70) = 3.59$, $p < .05$, $\eta^2 = .09$. Follow-up least significant difference analyses revealed that FTLD patients showed less down-regulation (i.e., more emotional facial behavior) than control participants ($p < .02$). AD patients also showed significantly less down-regulation than controls ($p < .04$) and did not differ from FTLD patients ($p > .05$). These results are consistent with the hypothesis that FTLD patients would show impaired emotion regulation. However, the finding that AD and FTLD patients were comparably impaired in this kind of instructed emotion regulation was not hypothesized (see Figure 1).

Somatic Activity

The ANCOVA revealed no significant main effects for diagnostic group ($F < 1$, $\eta^2 = .03$), sex ($F < 1$, $\eta^2 = .01$), or startle trial, $F(2, 134) = 2.31$, $p = .10$, $\eta^2 = .03$. The interaction between diagnostic group and startle trial was not significant ($F < 1$, $\eta^2 = .03$); thus, to our surprise, there was no indication that patients had any more difficulty down-regulating somatic activity than controls. Covariate corrected means and standard errors are presented in Table 2.

Discussion

We applied laboratory methods derived from contemporary affective science (Levenson et al., 2007) to examine specific emotion regulatory deficits in two common forms of dementia, FTLD and AD. We used a highly aversive acoustic startle as a stimulus, administered the startle under three different instructional conditions, and measured two aspects of visible emotional response (general somatic activity and emotional facial behavior). For somatic activity, there were no differences between the three diagnostic groups in their pattern of responding across the three startle trials. Thus, patients with FTLD or AD are comparable to controls in their ability to regulate this aspect of the visible response to the acoustic startle. For emotional facial behavior, in contrast, there were differences between the three diagnostic groups in their pattern of responding across the three startle trials. We turn next to these differences.

Consistent with previous research reporting intact emotional reactivity to simple stimuli in FTLD patients (Sturm, et al., 2006; Werner et al., 2007) and preserved emotional functioning in AD patients (Bucks & Radford, 2004; Harciarek & Jodzio, 2005; Lavenu et al., 1999), we found no differences between FTLD patients, AD patients, and neurologically normal control participants in emotional facial behavior when the startle stimulus was presented without warning and without instructions to regulate. This preservation of startle reactivity is consistent with the fact that the startle response is mediated by brainstem circuits (Davis et al., 1982) that are not affected in the early stages of these disorders (Hidgon et al., 2004).

Presenting the startle with warning but without any explicit instructions to down-regulate emotion provided an opportunity to examine if patients naturally down-regulate their emotional responses. On this trial, we found that patients with FTLD showed less spontaneous down-regulation of the outward expression of emotion than patients with AD and control participants. Thus, FTLD patients did not show the kind of spontaneous down-regulation of emotion when warned of an upcoming aversive stimulus that was shown by our AD patients and controls and that has been reported by others (e.g., Ekman et al., 1985; Keltner & Ekman, 1996). This kind of spontaneous down-regulation likely serves both personal (e.g., "don't overreact") and social (e.g., "don't alarm others") needs and is consistent with the deterioration of behaviors that protect self and others, the lack of social concern,

[1] Our primary analysis of this interaction involved examining differences within startle trials among diagnostic groups. Visual examination of the group means in Figure 1 suggested that AD patients might actually show more emotional behavior on the warned with instructions to down-regulate trial than on the warned without instructions to down-regulate trial. However, decomposing the interaction within diagnostic groups among startle trials revealed that this difference was not significant, $t(16) = 1.24$, $p > .05$. AD patients showed a significant reduction in emotional facial behavior from the unanticipated trial to warned without instructions to down-regulate trial, $t(16) = 3.17$, $p < .02$; this suggests that similar to normal control participants, AD patients down-regulated their emotional responses in the absence of instructions to do so. However, AD patients were not able to effectively comply with the instructions to down-regulate their emotional responses, showing emotional behavior that was comparable to the warned without instructions to down-regulate trial and to the unwarned trial, $t(16) = -1.19$, $p > .05$.

[2] Partial eta squares representing the portion of explained variance in the dependent variable are reported for each significant effect. The following eta squares correspond with small (.10), medium (.25), and large (.40) effect sizes (f), respectively: $\eta^2 = .01$, $\eta^2 = .06$, $\eta^2 = .14$ (Cohen, 1988).

Table 2
Emotional Facial Behavior Composite Score and Somatic Activity in the Three Startle Trials

Variable	Normal aging controls (n = 25) M (SE)	AD (n = 17) M (SE)	FTLD (n = 32) M (SE)
Emotional facial behavior			
Unwarned without instructions to down-regulate	$.82_a$ (.12)	$.60_a$ (.15)	$.71_a$ (.11)
Warned without instructions to down-regulate	$.28_a$ (.10)	$.24_a$ (.12)	$.62_b$ (.09)
Warned with instructions to down-regulate	$.08_a$ (.10)	$.41_b$ (.17)	$.40_b$ (.09)
Somatic activity			
Unwarned without instructions to down-regulate	3.03 (.48)	3.29 (.56)	3.43 (.41)
Unwarned without instructions to down-regulate	1.32 (.33)	1.51 (.39)	2.56 (.29)
Unwarned without instructions to down-regulate	1.12 (.22)	1.22 (.26)	1.62 (.19)

Note. Values are the emotional facial behavior scores in each startle trial for each diagnostic group; higher values indicate more emotion expressed during the 5-s epoch. Groups with different subscripts differed from each other at $p < .05$. Significance levels are from generalized linear model analyses of covariance comparing the three diagnostic groups. When the omnibus test was significant, pairwise comparisons were performed. FTLD = frontotemporal lobar degeneration; AD = Alzheimer's disease; *ns* = not significant ($p > .05$).

and the social inappropriateness often observed in FTLD patients (Levenson & Miller, 2007). Moreover, successful spontaneous emotion regulation requires the ability to self-monitor and self-regulate. Prior research has shown that processes related to the self (self-related emotions such as embarrassment; Sturm et al., 2006) are clearly deficient in FTLD patients—again, likely reflecting damage to frontal regions critical for such processes (Beer, Heerey, Keltner, Scabini, & Knight, 2003; Mitchell, Banaji, & Macrae, 2005).

In terms of instructed down-regulation on the warned with down-regulation instructions trial (on this trial participants knew when the startle stimulus would occur and were given explicit instructions to down-regulate their reactions), both FLTD and AD patients showed less down-regulation of emotional facial behavior than normal control participants. We think that this deficit may have different bases in the two dementias. Suppressing an emotional response is known to deplete other cognitive resources (e.g., memory in Richards & Gross, 2000). Thus, for AD patients, the relatively greater cognitive demands inherent in the instructed down-regulation trial, which involved remembering about the countdown, tracking its progress, remembering the instruction to down-regulate, and monitoring compliance, may have overwhelmed their limited cognitive abilities. For example, all participants were reminded immediately before the 20-s countdown of the instructions to hide their emotions, but holding this information in working memory for 20 s may have been too long for AD patients. However, for FTLD patients, the provision of the explicit instruction to down-regulate might have compensated for their inability to read social cues and devise appropriate regulatory strategies. The fact that they still had a measurable deficit may relate to problems with self-monitoring and self-regulation, skills that are still necessary to comply with the down-regulation instructions.

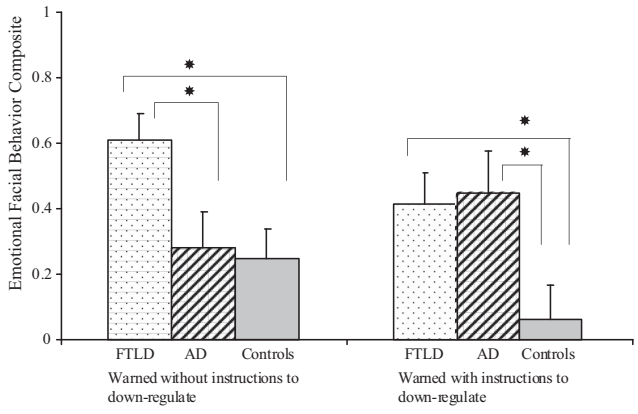

Figure 1. Mean facial expression composites (+*SE*) for frontotemporal lobar degeneration (FTLD) patients (n = 32), Alzheimer's disease (AD) patients (n = 17), and normal aging controls (n = 25) in the warned without instructions to down-regulate and warned with instructions to down-regulate startle trials. * $p < .05$.

This study revealed important similarities and differences in the emotional regulatory deficits found in two common dementias, FTLD and AD. The emerging picture is broadly consistent with clinician and caregiver reports of compromised emotion regulatory ability but provides greater detail as to the specific areas of preservation and loss of functioning in these two dementias. Deficits in emotion regulation in dementia may be even more striking given that emotion regulatory abilities have been found to show little decline in normal aging both in survey studies (Gross et al., 1997) and in laboratory studies (Kunzmann et al., 2005). Findings that AD patients may be able to regulate their emotions successfully in situations that do not overtax memory and other cognitive resources may be helpful to families and clinicians alike. Findings that FTLD patients can regulate emotion to some extent when explicitly told to do so but have trouble doing so spontaneously may be similarly helpful (although perhaps still frustrating to caregivers). It should be noted that these deficits were limited to the ability to down-regulate emotional facial behavior and were not found for the ability to down-regulate somatic activity. The specificity of this deficit suggests the possibility that different neural circuits are involved in down-regulating these two aspects of the visible emotional response and that these two neurodegenerative diseases relatively spare brain regions important for regulating somatic activity. This is clearly an unsettled issue worthy of

exploration in future research. For those interested in the accurate diagnosis of FTLD and AD, these findings add to the growing body of research showing areas of preserved and compromised functioning in these two important late-life dementias.

In terms of understanding the neural substrates of emotional functioning, the finding of striking deterioration of the facial expressive aspects of emotion down-regulation in FTLD patients underscores the important role that frontal brain regions (which are clearly targeted by FTLD) play in emotion regulation and self-monitoring. Although the data that would enable us to correlate the extent of regulatory deficits with neural loss in particular brain regions are not yet available with this sample of patients, we expect that they would show a link between the extent of frontal neural loss and the magnitude of deficits in emotion regulation. Evidence from other sources showing that disinhibited behavior is correlated with atrophy in orbitofrontal cortex (Peters et al., 2006; Rosen et al., 2005) and ventromedial prefrontal cortex (Lough & Hodges, 2002) and that response inhibition is correlated with levels of right inferior prefrontal cortex activation (Konishi et al., 1999) is certainly consistent with this view.

Limitations

Two limitations should be considered when interpreting the findings of this study. First, for all participants, the startle trials were presented in the same order, and therefore we cannot rule out the possibility that habituation effects might have affected the three groups differently. Second, we did not explicitly assess whether participants retained the regulation instructions over the course of the startle trials. Thus, although we speculate that regulation deficits seen in AD patients were due to problems they had in remembering the instructions, this cannot be confirmed.

Conclusions

This study illustrates the utility of using laboratory methods derived from basic affective science to assess emotional functioning in neurological patients (Levenson et al., 2007). Applied to a sample of dementia patients with AD and FTLD, this approach revealed nuances of preservation and loss of emotional functioning that have not been apparent in descriptions derived from clinician and caregiver reports. When provided with explicit instructions to down-regulate emotion and knowledge of precisely when an aversive event would occur, both AD and FLTD patients showed impairment in ability to down-regulate emotional facial behavior compared to normal control participants. When told when the aversive event would occur but not given instructions about emotion regulation, AD patients spontaneously down-regulated emotional facial behavior in ways quite similar to normal controls. In contrast, FTLD patients showed little evidence of spontaneous down-regulation. There was no evidence of deficits in ability to down-regulate somatic activity in either group. We believe these findings may be directly useful in discriminating between AD and FTLD patients in terms of subtle differences in their emotional functioning and indirectly useful in understanding the neuroanatomy of complex emotional behaviors such as spontaneous and instructed emotional down-regulation.

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