INTRODUCTION

Denial of deficit (anosognosia) is a well-described disorder, encompassing a range of unusual phenomena (see Bisiach and Geminiani, 1991; Feinberg, 1997; Gainotti, 1972; McGlynn and Schacter, 1989, Prigatano and Schacter, 1991 for review). However, until recently, the disorder was often regarded as little more than “an enigmatic clinical curiosity” (Ramachandran, 1996, p.124). In its extreme form the disorder presents as a manifest denial of paralysis, despite clear evidence to the contrary, where denial occurs even after explicit demonstration of the deficit by the examiner. In other circumstances, perhaps to be regarded as milder forms (anosodiaphoria) the patient may become aware of their hemiparesis, but deny that it causes them any functional disability (c.f. House and Hodges, 1988).

In some cases the denial of deficit is held with such strong conviction that the false beliefs persist in the face of all contradictory evidence and logical argument, even if this results in strange beliefs or perceptual experiences (Bisiach et al., 1986; Halligan et al., 1995; Ramachandran and Blakeslee, 1998; Weinstein and Kahn, 1995). It is clear that anosognosia can present in a number of ways (Marcel et al., in press). Thus, while mismatches between intended actions and outcomes “may be necessary for anosognosia they do not seem to be sufficient” (Frith et al., 2000, p.1782).

Another potentially fruitful proposal focuses on issues of motivation and emotion. There are some theoretically-driven grounds for this idea – for example the recent focus on the role of secondary somato-sensory cortex, in the right-hemisphere, in the higher aspects of homeostasis, somato-sensory representation, and emotion regulation (Damasio, 1994 pp. 62-69; 1999 pp. 209-213; 1996). This argument is supported by a well-established literature suggesting a central role for the right hemisphere in the perception and expression of emotion (e.g. Borod, 2000). For example, there are well-described effects of right-sided intra-carotid amobarbital, which produces anosognosia in a high proportion of cases (Brier, 1995; Lu et al., 1997, Meador et al., 2000), and has long been known to differ from the more emotionally appropriate outcomes that follow from left-sided amobarbital (e.g. Gainotti, 1972, see Feinberg, 1997 for review).

There is also a long-standing clinical literature on patients with right-sided brain lesions, showing that patients with anosognosia often do not show the negative emotional responses to their paresis that one might expect. Most notably, they have fewer so-called ‘catastrophic’ reactions (episodes of tearfulness and emotional breakdown) that are more frequently seen in patients with left hemisphere lesions (e.g. Fedoroff et al., 1992; House et al., 1990; Gainotti, 1972; 1997; Robinson and Jorge, 1994). Also, anosognosics are often not merely unaware of their deficits, but are sometimes
unnecessarily optimistic about their medical condition (i.e. ‘euphoric-maniacal’, Gainotti, 1997), and may also over-emphasize their abilities with the paretic limb (e.g. Ramachandran and Blakselee, 1998, pp.138-139).

The absence of a negative attitude towards impairment in patients with right-sided lesions, together with reports of low mood after left-sided lesions (Fedoroff et al., 1992; Gainotti, 1972; Robinson and Jorge, 1994) has led to Davidson et al.’s (e.g. Davidson, 2001; Davidson and Irwin, 1999) suggestion of a right frontal system involved in negative (withdrawal-related) emotional states, with left frontal regions associated with positive (approach-related) emotion. Thus, Davidson et al. have suggested that depression might result from disruption of a (left-sided) positive emotion system (e.g. Davidson and Irwin, 1999, p. 13). On this argument anosognosia would result from a disruption of negative emotion systems, leaving the patient with only a (left-sided) positive emotion system.

However, there are several reasons to doubt this possible link between an absence of negative emotions and anosognosia. For example, a disruption of negative emotions would explain only the absence of emotion in relation to paresis, not why the patient might actively deny their paresis and explicit evidence thereof (Marcel et al., in press). It has also been pointed out (see Gainotti, 1997 for review) that the low mood seen in patients with left-sided lesions is likely to result from an emotionally-appropriate response to their substantial levels of disability – which typically involves hemiparesis and non-fluent aphasia. A further line of argument is that there are some circumstances when right-sided lesions produce an increase, rather than a disruption, in negative emotion. For example, the occasional finding of explicit dislike or obsessional hatred of the paretic limb (‘misoplegia’, Critchley, 1974) seen after right-sided lesions, which is discordant with a loss-of-negative-emotions account. Finally, there have been reports of frank depression after right convexity lesions in patients who were also anosognosic (e.g. Kaplan-Solms and Solms, 2001; Starkstein et al., 1990; see also Turnbull et al., 2002) reported patients who experienced transient awareness of their deficit, including “sudden moments of tearfulness and pre-tearfulness” (p. 166) which appeared to be preceded by themes of ‘loss’ – even if the cause of such loss was apparently unrelated to the hemiparesis. Similar examples have been reported by Ross and Rush (1981), and Starkstein et al. (1988). This suggests that negative emotions may be intact in at least some instances of anosognosia. However, we note also that awareness of hemiplegia may appear to vary, depending on the nature of the question asked of the patient (see Marcel et al., in press), so that it is important to base examples of fluctuations in emotion on the same class of question asked at different times.

The fluctuations in emotion seen in these patients suggests that they might show the full range of emotional experience appropriate to their neurological deficits, including anger and sadness. This runs contrary to any claim (e.g. Davidson, 2001) for a right hemisphere system specialized for negative emotions, and a role for loss of negative emotions in anosognosia. The present study investigates an anosognosic patient (IW) on direct and indirect measures of emotion, especially focusing on three classes of negative emotion.

1The question of exactly how to define a ‘negative’ emotion is a complex one. Some authors (e.g. Davidson) classify all experiences that appear to be aversive in a unitary ‘negative’ category. However, recent work separating independent emotions suggest that there may be several distinct classes of negative emotion, with fear, anger and sadness (and perhaps others) as likely candidates (e.g. Calder et al., 2001; Damasio et al., 2000; Le Doux, 1996; Panksepp, 1998). All such states appear to result from ‘aversive’ events or potential events: for example, anger with frustration of goal-directed action; fear with risk to bodily harm; and sadness with separation and loss (e.g. Panksepp, 1998, p.52). A second line of evidence that supports the claim that these emotions have a negative valence is that the core anatomy of the purported anger, fear and sadness systems (such as the amygdala, hypothalamus, and dorsal peri-aqueductal gray matter) appear to have their substrate in regions that produce aversive responses when electrically stimulated (Bandler and Shipley, 1994; DePaulis and Bandler, 1991; Panksepp, 1999).

CASE REPORT

IW was a 70-year-old, right-handed, man, who had long been regarded as an upstanding member of the community, and had spent his life in various forms of public service (Royal Air Force, Police Service, and political office). He was married, with several children. In his younger years he had also been an enthusiastic sportsman.
At age 68 he suffered a right fronto-parietal cerebro-vascular accident (see Figure 1). Cortically, this involved the right motor cortex, frontal eye fields, frontal and parietal operculum, inferior parietal lobule, and the anterior portion of the inferior middle and superior temporal gyrus. There was also some involvement of the insula, a substantial portion of the frontal white matter, and the globus pallidus (sparing the internal capsule). IW was left with a substantial left hemiparesis, largely sparing the face and involving the arm and leg equally. The paresis had undergone a limited recovery. When he was assessed (from 20-24 months after the stroke) he retained a substantial paresis—such that he could move the upper arm and shoulder well, but had no movement in the distal portion of the arm and hand.

IW had shown marked anosognosia for his paresis in the acute period after the stroke. When assessed at 20-24 months this situation had improved slightly, but was variable. There were times when he was frankly anosognosic. On other occasions he appeared to be aware of his hemiparesis, but was wildly inaccurate about the severity: for example claiming that it was “95% returned to normal” (i.e. anosodiaphoria). At these times his presentation resembled the dissociation between awareness of disability and handicap reported by House and Hodges (1988, pp. 114-115), such that he could be made aware of his paresis on direct questioning, but denied that it was causing him any real handicap. Finally, there were occasions where he showed far greater awareness of his hemiparesis and its implication. At such times he felt enormous frustration at his disability, and such episodes often led to brief episodes of tearfulness. However, when questioned later during the same session on this topic, he typically returned to the argument that his disability was minor. Thus, IW was anosognosic or anosodiaphoric during each assessment, with brief periods of awareness of deficit during three of the five testing sessions. A possible explanation of the apparent differences in the level of his anosognosia, for at least some of the instances, may be the exact nature of the question asked of him (see Marcel et al., in press).

NEUROPSYCHOLOGICAL ASSESSMENT

Visuo-Spatial Ability

Neglect and Extinction on confrontation (Heilman et al., 2003, pp. 297-298): IW showed no signs of hemi-spatial neglect on confrontation testing in any sense modality (visual, auditory and tactile). However, he showed clear signs of extinction of the leftward stimulus on bilateral stimulation. In 5/6 ‘bilateral’ trials in the visual modality he reported the event as ‘right-sided’ only. A similar effect was seen in 5/7 ‘bilateral’ trials in the tactile modality. There were no signs of extinction in the auditory modality.

Line cancellation (Albert, 1973): IW showed no signs of neglect on the Albert line cancellation task.

Line bisection (Heilman et al., 2003): IW bisected 3/4 lines right of center. However, the effect of lateral bias was small, his errors (on lines of 12cm) being 2%, 3% and 8% right of center, with his one leftward errors being 2% left of center.

Rey Complex Figure Recall: IW’s memory of this figure (Figure 2) showed good recall of the basic elements, though it was impoverished in terms of detail (15/36). It was also rotated through 90 degrees relative to the original (see Solms et al. 1998; Turnbull et al., 1997a; 1997b for further discussion of this sign).

Executive Function

Block Design: IW performed very poorly on the Block Design subtest of the WAIS-R, failing to correctly complete any items successfully before the test was terminated after item 6. His errors were of three types: (i) failure to complete the figures into the ‘square’ pattern of the model (items 1-4), (ii) producing the correct overall ‘square’ shape, but with a single block rotated or incorrectly chosen, and (iii) a clear 90 degree rotation of one item.

Picture Completion: IW performed within normal limits (Scaled Score 11).

Rey Figure Copy: IW produced a reasonable final version of the figure, scored as 30/36. He used a poor (Type 3) strategy and omitted the
leftmost item from the figure, in spite of specific prompting to be certain that he had completed the entire figure.

Reitan’s Word Finding Test (Reitan, 1972). IW performed well within normal limits on this task (50/75) on this test of verbal problem solving. The task requires that the participant make a ‘guess’ at the meaning of a nonsense word (‘grobnick’) after consecutive clues (see Walsh, 1985, pp.155-156 for clinical examples). E.g.: Without the grobnick we would not live very long. The grobnick is very big. The grobnick is very far away. We cannot see the grobnick at night. The grobnick is hot and bright.

Austin Maze: IW showed a clear ability to learn the route through the maze, but with a shallow learning curve. He scored 12, 11, 10, 3, 10, 5, 5, 3 before testing was terminated. He showed no impulsivity, and no systematic errors on choice points, so that his poor performance seemed more a result of visuo-spatial than executive difficulties.

Comment

IW suffered a large right middle cerebral artery infarction, and showed a range of neurological and neuropsychological deficits consistent with this lesion site: a substantial left hemiparesis, some neglect-related and visuo-spatial disorders, and denial of deficit (anosognosia). He did not appear to be impaired on tests of executive function. IW’s visuo-spatial deficits appear to be relatively minor, and seem unlikely to be playing a substantial role in generating his anosognosia.

Assessment of Emotion and Personality

A variety of approaches were adopted in order to establish the nature of the changes to IW’s emotional life. These consisted of (1) a self-rating measure of emotion (Affective Neuroscience Personality Scale), (2) a series of memory probes relating to different classes of emotion (Affective Story Recall), and (3) interviews with two members of IW’s family, attempting to establish the nature of IW’s pre-morbid personality, and personality changes.

Interviews with IW’s family (his wife and daughter) suggested a man who placed great emphasis on authority figures, and on the institutions of society in general. He had always believed that there was a ‘right’ way to do things, and that one should constantly strive to become a better person. This account is of some interest given that denial of deficit has been reported to be more common in those who were pre-morbidly “conscientious, highly work-oriented, orderly, disciplined people” (Prigatano and Weinstein, 1996, pp. 318-319; Weinstein, 1996; Weinstein and Kahn, 1953; see also Gainotti, 1975).

From the perspective of IW’s family, the most striking effect of his stroke was a change in personality. His family reported that IW was easily moved to tears. For example, when singing “sad laments and other sad songs” in the church choir, IW was frequently overcome with sadness, most notably when he had returned for his first post-morbid concert with his choir. IW burst into tears at the beginning of a song, was incapable of joining in, and merely stood still in the midst of his fellow choristers, with tears streaming down his face, for the remainder of the song. At the start of the next song he was able to regain his composure and continue to sing. When asked what the song in question had been, he said that it was about ‘autumn’, with the leaves falling from the trees. IW then clearly suggested (though he could not quite say this out loud, for fear of becoming overcome with emotion again when describing the event) that the associated thought was one of the end of life. IW and his family described many other examples of this sort – such as when he read or saw a television report of someone being involved in an accident. He was notably more anxious when a member of his family might be exposed to danger, such as his daughter driving long-distances in her car.

There were no reports of IW behaving in a disinhibited way (i.e. suddenly acting on impulse). However, there were times when his behaviour seemed fully planned, but ill-judged. Thus, on one occasion he began telling a joke of a sexual nature to a group of women. His sister and wife were horrified, and said that such an event would have been unimaginable to him pre-morbidly.

Affective Neuroscience Personality Scale

IW was asked to complete a multi-dimensional emotion and personality inventory (Davis and Panksepp, 1999; Davis et al., 2003), derived from the basic categories of emotion that have emerged from the recent neuroscience literature (Damasio et al., 2000; Cloninger, 1994; Le Doux, 1996; Phan et al., 2002; Panksepp, 1998; Rolls, 1999). This measure is especially useful in the present circumstances because it tracks negative emotions of three separate types. The negative emotions are (1) Anger: e.g. “When I am frustrated I often get angry”, “If I am blocked from getting what I want, I usually just accept it”; (2) Fear: e.g. “people who know me well would say I am an anxious person”, “I am frequently more tense inside than others realize”; and (3) Sadness: e.g. “I often feel sad”, “Moving away from my friends would not upset me”.

The scale also includes one category for positive emotion, nowadays well-understood to be related to dopamine - motivating us to investigate our environment, with the expectation of reward (e.g. Robbins and Everitt, 1992). The system has been variously referred to over the years as a ‘reward’ (Schultz, 2001), ‘preparation’ (Hobel,
On the three negative emotion categories, IW produced scores that were divergent from neurologically-normal controls only in that he reported elevated levels of emotion. On measures of fear IW scored 33/42 (> 84th percentile), and for sadness, IW scored 34/42 (> 98th percentile, a higher score than any male control subject) and on anger, IW scored 16/42 (< 16th percentile). On the positive emotion items, IW scored 39/42 (> 98th percentile).

Comment

IW scored highly on two of the three categories measuring negative emotion, including an extremely high score on the sadness measure. He also produced a high score for the positive emotion category. These data are not consistent with a claim that IW has a selective disruption of negative emotions. However, we note also that self-report measures of emotion are readily open to bias, and also that IW’s increased ratings may result from a low level of insight into his own emotional state.

AFFECTIVE STORY RECALL

In addition to a questionnaire-based approach, we also thought it appropriate to design a task that addressed the issue of IW’s emotional experience more indirectly. In an Affective Story Recall task, IW was asked to recall a series of personal experiences from events in his life, which matched a particular emotional category. The list of such prompts was designed around the categories used in the questionnaire reported above. IW was asked to recall memories of four types, following the prompts was designed around the categories used in the questionnaire reported above. IW was asked to recall memories of four types, following the

Results

The stories produced by the neurological patient control group tended to focus on the nature of their disability, and its implications for their everyday life. Thus, a typical patient described anger because he could not garden any more (a previous hobby), and sadness because of his reduced social contact. He was also fearful because he might be knocked over, or trip, when going up the stairs in his local pub. His seeking stories were dominated by his wish to return to full health, and that he had expected to show the sort of full recovery seen in cases of a broken leg.

Those produced by the neurologically-normal control group tended to focus on events related to how they felt in situations related to themselves or close family. For example, one control described fear when she heard on the radio that there had been a massive explosion at her husband’s factory.

IW’s stories were dominated by emotion of various types, but differed from those of controls in that they focused on the disability experienced by others. A typical example was his Anger story: When climbing in the mountains, impending bad weather, IW and colleagues chanced upon a climber totally unprepared for the possible conditions. IW warned him not to continue, but the climber did not heed his advice. Later, at IW’s insistence, he and his colleagues tracked back to find that climber had slipped and been killed. IW chose this as an example of the emotion ‘anger’, he said, because “the climber should have taken advice from people who knew”.

Because IW’s stories focused especially on the serious consequences were typically experienced by others, the data were analysed in terms of the ‘object’ of the emotion, in addition to quantifying its magnitude (see below).

The data were blind-rated by two control subjects who had no history of neurological disease. The raters read each story and rated it in terms of its perceived emotional content in each of the four categories (e.g. “How much of the content relates to fear or anxiety?”) on a 0-10 scale. They also rated each story in terms of the object of the emotion – i.e. whether the primary object/person which the story referred to was that of the person telling the story (self), or another individual. The
scores for each emotion, in each category of story, were averaged across raters.

**Absolute Levels of Emotion**

An important preliminary analysis focuses on whether the absolute levels of emotion differ between IW and two control groups (Figure 2). The levels of emotion did not differ greatly across the four emotion categories, though IW’s level of rated emotion were lower than those of the two control groups for three of the four emotion categories, but higher for separation-distress. Using ANOVA to compare the magnitude of overall emotion across categories, IW was not significantly different to the neurological patient control group \( F (1, 14) = 0.036, p > 0.05 \), or the neurologically-normal control group \( F (1, 22) = 1.387, p > 0.05 \).

**The ‘Object’ of the Emotion**

While IW’s overall level of emotion did not differ from those of controls, it is of some note that the object of the emotions experienced by IW appeared to differ from those of controls. In each case, the emotions experienced by controls appeared to be rated more highly in relation to their self, for example in that they felt anger, anxiety, and feelings of loss in relation their paresis (see Figure 3) – a result that we have reported based on data from another group of patients (Turnbull et al., 2002). However, IW’s profile showed that such feelings was far more commonly directed at others.

When comparing IW to the neurological patient control group, ANOVA showed no main effect of self vs. other \( F (1, 14) = 2.29, p > 0.05 \), no main effect of group \( F (1, 14) = 2.22, p > 0.05 \), however, there was a substantial interaction \( F (1, 14) = 34.5, p < 0.001 \). *Post-hoc* analyses showed a significant difference between IW and controls for both self \( t (14) = -4.6, p < 0.001 \) and others \( t (14) = -4.2, p < 0.001 \).

When comparing IW to the neurologically normal control group, ANOVA showed a main effect of self . other \( F (1, 22) = 23.51, p < 0.001 \), a main effect of group \( F (1, 22) = 109.5, p < 0.001 \), and an interaction \( F (1, 22) = 8.84, p < 0.01 \). *Post-hoc* again analyses showed a significant difference between IW and controls for both self \( t (22) = -7.5, p < 0.001 \) and others \( t (22) = -11.8, p < 0.001 \).

Analysing these data at the level of individual emotion types would result in a loss of statistical power. However, as can be seen in Figure 4, a few general trends are evident. Firstly, as regards the tendency for IW to have lower ratings than controls on ‘self’ items, this trend was evident for all four emotion classes, relative to both the neurological, and the neurologically normal control group. In each case IW’s performance was in the lower 10th percentile, and was below the 1st percentile relative to neurologically normal controls in terms of sadness, fear and seeking. As regards the tendency for IW to have higher ratings than controls on ‘other’ items, this trend was also evident for all four emotion classes, relative to both the neurological, and the neurologically normal control group. Again, IW’s performance was always in the lower 10th percentile, and was again below the 1st percentile relative to neurologically normal controls in terms of sadness, fear and seeking.

**Comment**

The overall magnitude of emotion in Affective Story Recall for IW was not significantly different.
from that of controls, suggesting that IW has a relatively normal range of emotional report, at least as measured by this task. It is of especial note that the negative emotions (sadness-distress, anger-rage and fear-anxiety) did not differ from controls. Indeed, in the category of separation-distress, IW exhibited higher levels than both control groups. This suggests that IW continues to experience a magnitude of negative emotions that is consistent with that of others, including neurological patients with hemiparesis.

However, there was some evidence that IW’s emotions appear to be more commonly directed at a different object – such that emotional content of the story is directed far more at others than at himself. As shown in Figure 4, this effect seems to be consistent across each of the categories of emotion.

**DISCUSSION**

The primary goal of the investigation was to formally establish the magnitude of emotions experienced by IW, and in particular whether he experienced negative emotions. His emotional experience, measured by self-report questionnaire (the Affective Neuroscience Personality Scale), produced scores at the higher end of the normal range for three of the four emotion categories (only anger was low), and was especially high for the category of sadness. A more indirect assessment of his emotional state, using the same categories of emotion (Affective Story Recall), again produced scores not significantly different from two control groups across all emotion categories. Indeed, it was lowest on the positive emotion category of seeking.

Thus, across two quite different assessment methods, IW shows a range of emotional experience that is consistent with that of controls, especially for negative emotions (anger, fear and sadness). Taken together with other reports of negative emotion, and even frank depression, in anosognosia (Kaplan-Solms and Solms, 2000; Starkstein et al., 1990), this result runs counter to the argument that anosognosia might result from a loss of negative emotions. However, it is possible that anosognosia is a diverse phenomenon (e.g. Marcel et al., in press) that includes negative emotions (anger-rage and fear-anxiety) that have been revived more recently, suggesting that anosognosia might be a form of defense (Marcel et al., in press; Kaplan-Solms and Solms, 2000; Ramachandran, 1996; Turnbull et al., 2002). Were this true, then at least some anosognosic patients would have a demonstrable implicit awareness of their disability (see Feinberg, 1997, p.384 for further discussion).

An interesting aspect of IW’s performance on the task of Affective Story Recall was his tendency to produce the same magnitude of emotion as controls, but for this to be directed at an external ‘object’ – a finding that we have reported elsewhere (Turnbull et al., 2002). For example, IW’s production of emotion-related stories carry a recurring theme of loss (especially death and injury), almost regardless of the emotional topic suggested to him. One explanation of this finding might be that IW's attempts to consciously link his feelings of sadness with his hemiparesis are aversive and/or intolerable to him (c.f. Anderson and Green, 2001). Arguments of this type have previously been proposed (Goldstein, 1939; Schilder, 1935; Weinstein, 1991; Weinstein and Kahn, 1955), and have been revived more recently, suggesting that anosognosia might be a form of defense (Marcel et al., in press; Kaplan-Solms and Solms, 2000; Ramachandran, 1996; Turnbull et al., 2002). Were this true, then at least some anosognosic patients would have a demonstrable implicit awareness of their disability (see Feinberg, 1997, p.384 for further discussion). However, the evidence for this claim remains indirect, and more explicit attempts to measure the extent of awareness of disability would form a further useful topic for future research.

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