Clinical Postcard

Preserved fluid intelligence in the context of impaired executive functions

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Fluid intelligence (Gf) is a key feature of human cognition. Its relationship to other cognitive domains, such as executive functions, and its brain basis, however, remains contentious. It has been suggested that widely distributed cortical networks underpin Gf (Colom et al., 2010). More specifically, a large fronto-parietal network, the Multiple Demand network (MD), has been proposed as the seat of Gf (e.g. Woolgar et al., 2010). However, on this approach deficits on some executive tasks are not explained solely by impairment on Gf (e.g., Roca et al., 2010). We recently reported that performance on the Raven Advanced Progressive Matrices (APM), a commonly used test of Gf, is underpinned by a set of localised right frontal regions (Cipolotti et al., in press). This raises the possibility of finding isolated sparing of Gf processes. We present an observation of two patients with Gf sparing and profound executive impairment.

Both patients (P1 and P2; 56 and 66 years old, respectively) met the international consensus criteria for the behavioural variant of fronto-temporal dementia (FTD; Rascovsky et al., 2011). P1's MRI showed frontal atrophy, more pronounced on the left (the original imaging is no longer available). P2's MRI showed widespread atrophy, quantified from a volumetric study, revealing a striking regional pattern. There was marked bilateral atrophy of temporal and, to a lesser extent posterior and subcortical regions, with left-sided frontal changes but relative preservation of right frontal and bilateral parietal cortices (Fig. 1).

Both patients presented with 3-to-4-years history of progressive deterioration of behaviour, as reported by their spouses. Thus, both displayed: inappropriate social behaviour (e.g. P2 enjoyed telling 'embarrassing jokes' with sexual innuendos, even in front of children), diminished response to

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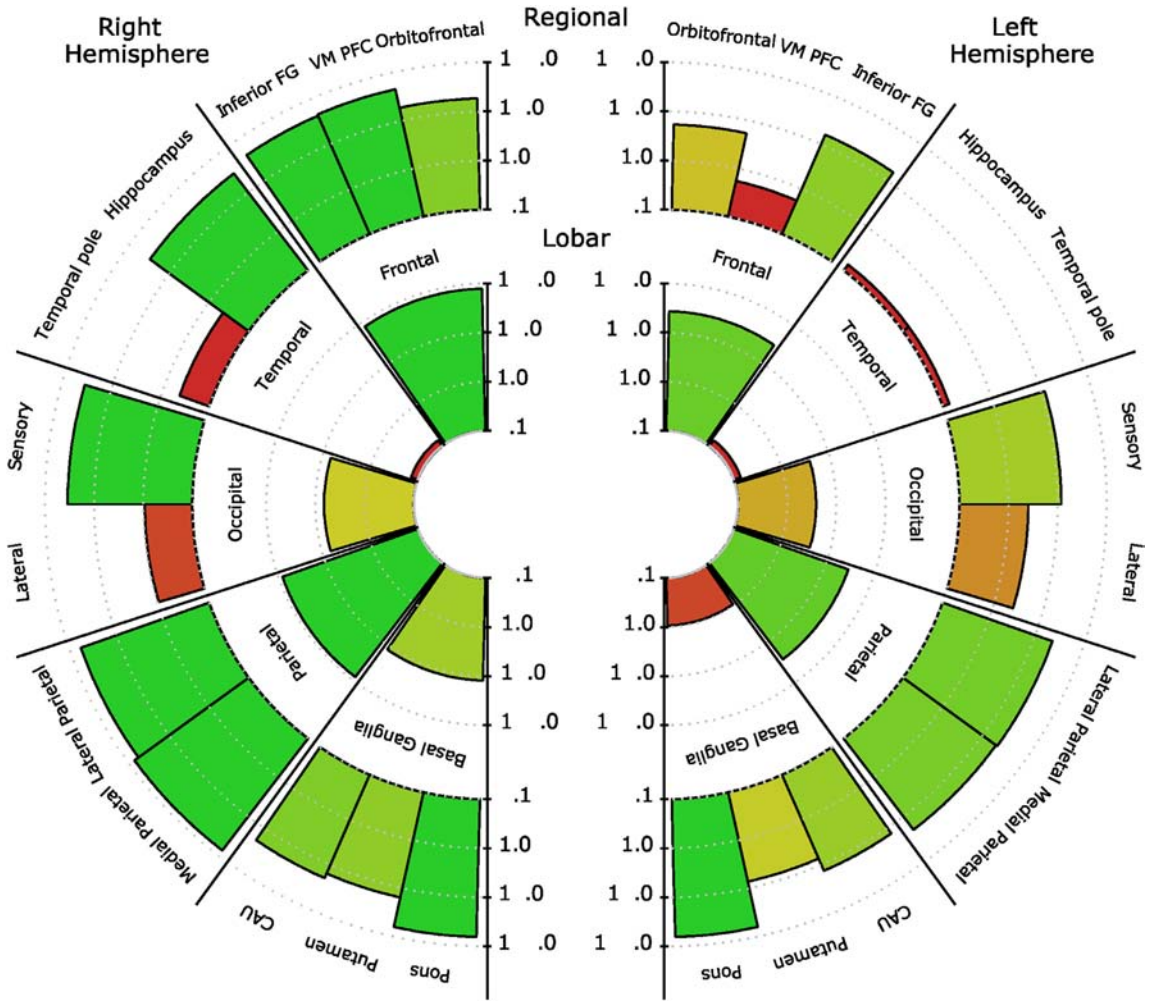

Fig. 1 Representation of regional volumes quantified with geodesic information flows and expressed in terms of percentiles of an age matched reference distribution (Cardoso et al., 2015). Note pronounced temporal and posterior atrophy, with comparative preservation of right frontal and parietal regions.

peoples' needs and feelings (e.g. P1 was inappropriately chatting and jocular in the outpatient waiting area), dietary changes (e.g. eating huge number of chocolates and sweets), stereotyped and compulsive/ritualistic behaviour (e.g., P2 vacuum cleaned his bedroom several times a day) and loss of manner or decorum (e.g., both developed a keen interest in pornography and discussed it openly).

Both patients scored within the normal range on perceptual tests from Visual Object and Space Perception Battery (Warrington & James, 1991) and on the Graded Difficulty Arithmetic Test (Jackson & Warrington, 1986). Memory for ongoing events was satisfactory. Both presented with a progressive language impairment which developed, in a similar manner, approximately three years after the findings reported here for P1 and was already present at the time of the examination for P2. Thus, P2's spontaneous speech was well articulated, with a normal prosody and, at times, somewhat tangential. Some stereotyped, grammatically correct sentences were present. Performance was impaired on a test of receptive grammar (Bishop, 1989). Severe nominal difficulties were noted. For example, when describing the cookie theft picture from the Boston Diagnostic Aphasia Examination (Goodglass et al., 2001), he said " ..quite an interesting one .it looks like for some reason .this is pouring out … (pointing to the overflowing sink)..the son is just about to fall over which is not very good (pointing to the boy falling off the stool). On naming tests (see Cipolotti, 2000; McKenna & Warrington, 1980), P2 produced circumlocution (e.g. map of USA- "this is where New Yok is"; monocle; "goes in the eye"); semantic (e.g. rabbit- "monkey") and superordinate errors (e.g. boat- "vehicle").

Both patients showed very marked executive impairment on no less than 5 verbal and nonverbal executive tests (see Supplementary Table 1). When normative data was unavailable, patients' performance was compared against two individually age and education matched groups of 10 healthy controls. Both patients were impaired on nonverbal fluency tasks. P1 made several perseverative errors. P2 could generate a small number of designs and made rule break errors. Remarkably, both patients failed even the Weigl, usually administered to patients with severe deficits (MacPherson et al., 2015). This could not be explained by perseveration. For example, although P2 initially perseverated on the shape

Table 1 e Fluid intelligence tests.

	P1		P1 HCs (n ¼ 10)	P2		P2 HCs (n ¼ 10)
	Raw score	Percentiles	Mean (SD)	Raw score	Percentiles	Mean (SD)
RAPM (correct/12; Raven, 1976) *	7	32nd %ile		9	64^{h} %ile	
RCPM (correct/36; Raven, 1949) *	NT			36		
RSPM (correct/60; Raven, 1958) *	41	75th-90th %ile		NT		
Cattell Culture Fair Test 1 (correct/12; IPAT, 1973)	8	20th %ile	9.10 (1.20)	10	60th %ile	9.50 (1.78)
Cattell Culture Fair Test II (correct/14; IPAT, 1973)	7	34th %ile	8.40 (3.13)	8	50th %ile	8.00 (2.58)
Cattell Culture Fair Test III (correct/12; IPAT, 1973)	9	54th %ile	8.80 (1.75)	9	73rd %ile	8.00 (1.49)
Cattell Culture Fair Test IV (correct/8; IPAT, 1973)	5	43rd %ile	5.30 (1.64)	6	57th %ile	5.60 (2.01)

Legend: HCs ¼ Healthy controls; SD ¼ standard deviation; RAPM ¼ Ravens' Advanced Progressive Matrices; RCPM ¼ Raven's Coloured Progressive Matrices; NT ¼ not tested; RSPM ¼ Raven's Standard Progressive Matrices; IPAT ¼ Institute for Personality and Ability Testing. See supplementary material for test references.
Where possible, patients' performance was compared against published normative data (indicated by *). On the Cattell tests, when normative data were unavailable, modified t-tests (see Crawford & Garthwaite, 2002) showed that neither P1 and P2's performance significantly differed from age and education matched HCs (P1 HCs mean age ¼ 54.80; SD ¼ 2.82; mean years of education ¼ 14.20; SD ¼ 1.23; P2 HCs mean age ¼ 65.00; SD ¼ 2.05; mean years of education ¼ 13.90; SD ¼ 1.85).

sorting principle, when prompted, he was still unable to obtain the second principle and resorted to producing a column. Both patients' performance was effortful and with many errors on the incongruent condition of a simplified Stoop test. Moreover, P1 was unable to suppress the dominant response on Section 2 of the Hayling test, despite being able to understand and repeat the instructions. P2 failed the Luria's hand sequence test (learning only 2 movements) and the reverse tapping test. P1 was perseverative on a phonemic fluency test.

Remarkably, both patients' performance was completely normal on three Gf tasks, relative to normative data and two groups of 10 healthy controls (see Table 1). Both patients performed within the normal range on various versions of the Raven tests and on all the four subtests of the Cattell test.

Our results suggest that performance on two sets of key Gf tests can be selectively spared in FTD in the context of grossly impaired behaviour on other executive tests. This complements the findings of several group studies which have suggested that frontal executive impairments cannot all be explained by a Gf impairment (Roca et al., 2010: Mole & Cipolotti, 2021). Our current observations show that despite sparing of Gf, severe executive impairment can occur. This supports the notion that a single factor eGf - cannot account for all executive performance. More positively, it raises the possibility that there may be a separation in the architecture of frontal functions between processes mainly involved in Gf and those involved in other executive components.

Our imaging analysis of P1 revealed relative preservation of right frontal areas, which are now known to play a critical role in Gf (Cipolotti et al., in press). This may help to explain why our patients performed well on Gf tasks. Further research may sharpen our understanding of the core processes underlying Gf.

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Data availability

Data will be made available on request.

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