Are Socio-Affective Impairments in Ataxia Related to Language and Processing Speed Deficits?

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Declaration of interests

The authors declare no competing interests.

Abstract

Over the last decade a number of studies have demonstrated the implication of the cerebellum in cognition, including verbal memory, executive function, and language. Social cognition abilities like emotion attribution and theory of mind are essential in social interaction and rely on verbal abilities and executive functions. Involvement in social cognition has been explored in the most common forms of Spinocerebellar Ataxia (SCA), but several aspects remain unclear. The present study tested whether socio-affective impairments are observed in SCA patients by using different tasks and to determine whether these impairments were associated with reduced verbal processing and/or processing speed. 13 patients (SCA1, n=1; SC2, n=5; SCA3, n=7) were matched with 13 controls for gender, age and education. Verbal and non-verbal theory of mind abilities were tested (validated French versions of an attribution of intention test, faux-pas test), and emotion attribution. Language efficacy was explored in a word fluency test and processing speed in two non-motor tasks. Results revealed no difference between SCA2 and SCA3 patients in neither socio-affective or cognitive test. Performance on all tests was on contrary significantly reduced in the SCA patient group compared to controls. SCA patients' performance was positively correlated between the three social cognition tests, indicating a somewhat generalized impairment. Their performance rate in each of three social cognition tests was further correlated with processing speed but not with word fluency. In the verbal theory of mind task they displayed however comprehension deficits of the faux-pas and of general control questions. Taken together the data suggest that processing speed and comprehension difficulties might account, partly at least, for socio-affective and cognitive

deficits in both genotypes. This conclusion was modulated by the observation that the neuropsychological features did not correspond well with age of illness diagnosis and illness duration, indicating that there is a degree of heterogeneity in the cognitive profiles and social cognitive impairments in SCA patients.

Keywords.

Theory of mind, emotion attribution, cognitive speed, fluency, SCA2, SCA3.

1. Introduction

Since Schmahmann's seminal contribution[1], an increasing number of neuropsychological studies have investigated the role of the cerebellum in cognition in patients with congenital and acquired damage of the cerebellum. The spinocerebellar ataxia (SCAs) is a rare (prevalence rate of 1 to 4 in 100,000) neurodegenerative disorder of autosomal dominant inheritance[2]. The most common of the 26 sub-types, SCA1, SCA2, SCA3, SCA6, and SCA7 are characterized by degeneration of the cerebellum and its corticocerebellar connections leading to cerebellar ataxia. Sub-types 1, 2 and 7 for instance share common clinical features, in particular pyramidal and extrapyramidal signs, oculomotor abnormalities and amyotrophy, though several clinical features and olivopontocerebellar atrophy vary between these subtypes, being less severe in SCA1 compared to SCA2 patients[3-5].

Cognitive deficits in the most common SCA1, SCA2, and SCA3 types – of interest in the present study - affect more especially verbal memory, executive function, language and attention[4,6-9]. A heterogeneity in the types of cognitive impairments has been reported for the different SCA subtypes in studies using large cognitive assessment batteries. Little deficits or selective deficits were reported in SCA6 and SCA7 patients and severe/rapidly declining impairments in SCA1, SCA2 and SCA3 patients[5,10]. The decline over time also appears to be specific of the phenotype[8]. The neuropsychological profiles and cognitive decline over time of SCA patients correspond generally with the severity of pathological and clinical features[5, 8], but dissociations between cognitive and motor impairments have also been reported[4].

More recently, the involvement of the cerebellum in social cognition abilities has also been explored in the most common forms of SCA. Social cognition abilities like emotion attribution and theory of mind are essential in social interaction. According to Baron-Cohen and co-workers[11] Theory of mind tests evaluate how well participants can put themselves into the mind of another person: "This ability is the main way in which we make sense of or predict another person's behavior". Studies exploring social cognition abilities in ataxia reported

emotion attribution impairments in SCA2 and SCA7 patients but not in SCA1, SCA3 and SCA6 patients, who all showed in contrast a Theory of Mind deficit[5,10]. According to these studies, socio-affective impairments are not homogeneous among the various SCA genotypes, in line with the findings of differential cognitive and executive impairments according to subtype. Other studies reported theory of mind comprehension disturbances in different patient groups (SCA1, 2, 3, 6, 7) without obvious group differences. The authors propose that social cognition might to be one component of the SCA profile, and might not be associated with clinical or other neuropsychological measures[8]. Likewise, specific recognition deficits of social facial emotion expressions in SCA2, 6, 7 and 8 patients were independent of clinical and demographic features, leading to the proposal of an involvement of the corticocerebellar network in socio-cognitive abilities[7]. Altered connectivity to cerebral regions has thus been reported to be related to cognition and emotion in a group of SCA2 patients[12].

The involvement of the cerebellum in social cognition abilities was also documented in healthy subjects and in other clinical conditions. Neuroimaging studies have reported a cerebellar activation in healthy controls during theory of mind testing[13-15] and facial expression processing[16,17]. Conversely, patients suffering from isolated cerebellar degeneration suffer from theory of mind deficits[18]. Patients suffering from schizophrenia and autism, two conditions that are associated with structural and functional abnormalities of the cerebellum, also show social cognitive dysfunction, including deficits in facial emotion recognition and of theory of mind among the core features. More especially, neuroimaging studies consistently report reduced cerebellar activity in schizophrenia patients during facial emotion processing, though inconsistent results were reported for theory of mind in this clinical population[19], with for instance divergent results according to task variables[20] (verbal *vs.* non-verbal. In patients with autism a processing deficit of facial emotional expressions was also associated with an altered activation of cerebellum, limbic and cortical regions compared to control subjects. Furthermore, a cerebellar–parietal network has been shown to be directly related to social cognition and social functioning in autism and to account for individual variance in emotional reasoning in patients with psychotic disorders[21].

Despite supporting evidence, the widespread nature of ataxia and the heterogeneity of the cognitive profile of SCA patients *per se* and over time[5,8,10] make it difficult to attribute the cognitive deficits unequivocally to cerebellar dysfunction[6,10]. This cautious point of view seems all the more relevant when a specific cognitive domain, *i.e.* the socio-affective area is considered. Contradictory interpretations were proposed in the literature to account for social cognition disorders in SCA patients; the idea of a common social cognitive impairment profile

irrespective of the subtype[8] contrasting with the observation of a heterogeneous social cognition impairment in the different genotypes[5,10]. Several factors might account for the contradictory findings in the literature. Decreased abilities in a higher order cognitive area may for instance be the consequence of lower order cognitive dysfunction or of a dysfunction outside of the social-emotional area. It is now well established that theory of mind for instance is a complex ability and that successful task completion involves distinct cognitive processes relating both to executive functions, *i.e.* working memory to manipulate information in an ongoing social situation, inhibition of one's own perspective, flexibility to switch to the other person's point of view[22], and to instrumental abilities including semantic abilities to understand the context and processing speed. Processing speed is an important construct in understanding cognition and has been shown to mediate working memory and executive functions[23]. Likewise, verbal fluency performance is determined by cognitive speed and by several abilities and processes related to semantic memory (*i.e.* word productivity) and to executive function (i.e. response initiation according to a specific rule, cognitive flexibility) (for a review[24]). Hence, in SCA patients verbal fluency, executive and processing speed deficits have been consistently reported[1,4,6-10]. Some studies have described these abilities more extensively in specific genotypes. A case study observed slowed processing speed and reduced mental flexibility in SCA2, 3 and 6 patients[25]. Significant deficits in executive function processing speed and letter fluency were described in SCA3 patients, and letter fluency was shown to be correlated with functional neuroimaging measures[26]. Further, processing speed in a nonverbal memory task predicted SCA patients' recall accuracy[27]. In patients with schizophrenia, slower theory of mind performance reaction times and delayed activation in theory of mind related brain areas have been interpreted as reflecting slower cognitive processing during mentalizing[28].

Cognitive speed, verbal and executive disturbances reported in specific SCA subtypes also vary according to clinical variables and in particular illness onset and duration. In SCA2 patients for instance no or slight impairments of word fluency and executive function were observed after one to three years of illness[12], verbal memory and fluency impairments after 6 years of illness[29]. A follow-up study reported some impairments in SCA2 patients on tasks of speed, attention and executive function after ten to fifteen years of illness and relatively little change over an additional 7-year period suggesting a rather stable neuropsychological profile in SCA2 patients[8], also reported by Le Pira et al's study after 8.5 years of follow-up in SCA2 patients. In contrast, SCA3 patients exhibited a decline of these abilities after the same follow-up period and their deficits were already more pronounced at baseline, presumably in relation to early onset of disease in this subtype. They also reported some verbal recognition memory

impairments in SCA2 patients and more severe nonverbal recognition memory decline in SCA3 patients. On contrary, they reported the greatest degree of neuropsychological impairment over time (attention, speed, executive function) in SCA1 patients, exhibiting a more severe atrophy. Interestingly, all subgroups exhibited comprehension deficits while performing a theory of mind task. Taken together, these observations favour the idea that reduced cognitive speed but also impaired comprehension and executive function might be determinant factors of SCA patients' performance in tasks exploring socio-affective abilities.

In the present study we assessed SCA2 and 3 patients' performance in two theory of mind tasks and in an emotion attribution task in order to test whether SCA patients' performance *a)* was impaired on both types of tasks thereby indicating a socio-affective cognition deficit in these subtypes on contrary to the dissociation reported in the literature between impaired theory of mind performance and preserved emotion attribution performance in SCA2 patients[5] and the opposite dissociation in SCA3 patients[10]. In light of verbal fluency and comprehension deficits reported in the literature, *b)* performance was expected to be preserved in the nonverbal but not in the verbal theory of mind task task *c)* and to be significantly related to language efficacy and cognitive speed suggesting that instrumental difficulties might account partly at least for the observed social cognition deficits.

2. Methods

2.1. Subjects

Thirteen genetically confirmed SCA1 (n = 2), SCA2 (n = 5) and SCA3 (n = 7) patients, all caused by CAG repeat expansion, and 13 control subjects gave their informed consent to participate in the study. Subject groups were matched for age (mean 47.77; 13.45), gender (5 males) and number of years of formal education (range 7 to 18; median 12). Age, age at disease onset, symptom duration, demographic characteristics and salient clinical details about each patient in the study are indicated in Table 1. Onset age and disease duration were used to assess disease severity[30] as they have been shown to explain disease severity in SCA1 (60.4%), SCA2 (45.5%) and SCA3 (46.8%). Sex distribution was as follows: SCA1, 1 male; SCA2, 2 males; SCA3, 2 males. Psychological tests were carefully selected in order to obtain valid responding, i.e. avoiding a motor response.

Table 1. Demographic and clinical characteristics of SCA1, SCA2 and SCA3 patients

No.	*SCA*	*Sex*	*Years of Education*	*Age (yrs)*	*Age at disease onset*	*Age at diagnosis*	*Disease duration*	*Motor signs*	*Nonmotor signs*	*Comorbid condition*
1	1	M	17	37	30	26	7	Speech Gate Balance		
2	2	F	12	22	11	12	11	Speech Gate Balance Posture	Comprehension Memory Planification	
3	2	F	14	41	34	41	7	Speech Balance	Memory Fatigue	
4	2	F	14	57	51	51	6	Speech Gate Balance		Hypertension
5	2	M	11	58	50	50	8	Balance	Memory	Depression (secondary)
6	2	M	7	69	30	68	39	Gate Balance	Memory Comprehension	
7	3	F	12	27	24	24	3	Balance		
8	3	F	12	45	26	32	19	Balance	Reaction Time	Crohn's disease
9	3	M	11	49	45	46	4	Balance	Memory Language	
10	3	F	16	50	45	46	5	Balance		
11	3	M	14	51	45	49	6	Balance		
12	3	F	12	52	47	42	5	Speech Balance		Tachycardia
13	3	F	9	63	43	55	20	Gate Balance	Nausea	

2.2. Materiel

Neuropsychological testing was completed individually at participants' homes, in a quiet room, and lasted about one hour. The interview started by the participant signing the informed consent form. Tests were chosen which are extensively used in clinical neuropsychological assessment and can be administered to individuals of various ages and different levels of education[31], and to reduce the influence of upper-limb symptoms suffered by the patients.

Measures of socio-affective abilities

The ability to attribute mental states to others was assessed with a nonverbal and with a verbal theory of mind task. Emotion attribution was explored with a nonverbal task.

- The nonverbal theory of mind task assessed the attribution of intentions to others, and required both the visual perception of human figures and the understanding of their intentions[32]. Twenty-eight black-and-white comic strips, were presented one-by-one. Each

comic strip was comprised of three pictures depicting a brief story that were displayed in the upper half of the sheet. Three answer-pictures were displayed in the lower half of the sheet. The subjects were required to choose spontaneously the logical ending of the story from these answers without looking for traps or humor. Only one of the answers ended the story logically and scored 1 (the maximum score was 28).

- The verbal theory of mind performance was evaluated with the faux-pas test[33] (Stone, Baron-Cohen, Knight, 1998; French version[34], in which participants were asked to identify whether someone in a fictitious social situation had said something wrong or had upset others. Twenty short stories of different social situations, 10 with a social and 10 without, were narrated to the participant. He/she then answered questions on each story. The participant was first asked whether someone had said something he or she should not have said, or something awkward that could have offended others (faux-pas detection). Following stories they identified as having a faux-pas, they were then asked five questions to assess their understanding of the faux-pas (character, rationale 1 and 2, unintentionality, associated emotion) and two to ascertain their comprehension of the story. Following stories they identified as not having a faux-pas participants were asked only general control questions. The maximum score of the faux-pas detection was 10 while the faux-pas understanding questions were quoted in percentages in regard to the correct faux-pas detection. The maximum score for the non faux-pas detection and for general comprehension control questions was 20 and 40 respectively.

Emotion attribution was explored using the reading the mind in the eyes-test, assessing how well a participant can put themselves into the mind of another person and tune into their mental state[11]. The participant is presented with a series of 36 black-and-white photographs of the eye-region of the face of different actors and actresses and is asked to choose which of four words best describes what the person in the photograph is thinking or feeling. Each correct response scored 1 and the maximum score was 36.

Executive function measures

Conventionally, the term of response speed refers to the duration between the onset time of a stimulus and the behavioral response and including sensory and motor time. In the present study we used the term of cognitive speed as tests were chosen to have minimal demands on motor functions, *i.e*. participants had to produce verbal responses, and the number of responses produced in a given time-interval was of interest rather than single-item response speed. The Stroop test[35] was used to assess processing speed. Commonly the colour-word interference

subtest (Card C) is used to assess cognitive inhibitory functions: Participants are presented with a series of colour words printed in different coloured ink (e.g.,"red" written in blue) and have to name as quickly and accurately as possible the ink colour, inhibiting the automatic reading of the incongruent colour word. In the present study, cards A and B (100 items per card) were used. Participants were asked to read colour words printed in black ink (card A) and to name coloured rectangles (card B) respectively as quickly and accurately as possible. Both subtests were time-constrained and the response score corresponded to the number of words/colours correctly read/named during a 45-sec delay.

Cardebat's Verbal Fluency test was used to test language efficacy[36]. The task consists of generating as many words as possible in a 2-min interval with a semantic restriction. The participant is asked to name as many words as possible belonging to a particular semantic group (e.g., animals).

2.3. Data analysis

Nonparametric statistics were used to compare mean raw test scores of the patient group and of the control group (test of Mann-Whitney). Spearman's correlation test was used to assess relationships of social cognition performance with executive functions and with clinical features in the group of patients.

3. Results

The comparison of the raw test scores of SCA2 and SCA3 patient sub-groups (Table 2) revealed no significant difference (Mann-Whitney test, in each case $p > .1$) for any neuropsychological test nor for age (49.4 and 48.1 respectively) or any clinical feature (age at disease onset: 35.2 and 39.3, diagnosis: 44.4 and 42.0, duration:14.2 and 8.9). Disease duration in the SCA2 subgroup dropped from 14.2 to 8.4 years when excluding the patient with the longest illness duration (39 years). Socio-demographic data and the scores of clinical assessment are shown in Table 3 for the group of patients and for the group of control subjects. Test scores are reported as raw scores and results of statistical analyses (Mann-Whitney test) figure in the last column. The patient and control groups were matched on the basis of relevant control variables of gender, age (mean 47.8 years in both groups), and education level (mean 12 to 13 years in both groups) and comparable ranges for the latter two variables.

Table 2. Mean (sd) scores on socio-affective and cognitive tests for SCA2 and SCA3 patients

	Emotion attribution	Nonverbal ToM	Verbal ToM	Semantic fluency	Word reading	Colour naming
SCA2 (*n* = 5)	18.6 (6.06)	33.0 (23.05)	21.2 (6.72)	18.8 (9.03)	69.2 (17.59)	47.4 (7.92)
SCA3 (*n* = 7)	23.43 (6.37)	42.43 (14.57)	22.71 (7.34)	16.43 (5.97)	76.71 (25.24)	58.0 (15.91)

3.1. Socio-affective performance

Table 3 shows that the mean raw performance scores on the three social cognition tests were lower in SCA patients compared to the matched controls. Statistical analyses revealed that SCA patients' performance was impaired in the emotion attribution task (M_{SCA} = 21.85 *vs.* M_{Ctr} = 27.38; p = .014), and in the nonverbal theory of mind task (M_{SCA} = 22.46 *vs.* M_{Ctr} = 27.15; p = .002). They also had lower scores in the verbal theory of mind task concerning the faux-pas detection (M_{SCA} = 7.69 *vs.* M_{Ctr} = 9.00) and the faux-pas understanding (M_{SCA} = 39.69 *vs.* M_{Ctr} = 52.92; p = .034), but only the faux-pas understanding score departed significantly from the one found in the control group (p = .034).

Table 3. Summary of socio-demographic data and group performance on cognitive tests for SCA patients and Controls

Neuropsychological tests	SCA patients (*n* = 13)		Controls (*n* = 13)		Statistics (Mann-Whitney test)	
	Mean (sd)	Range	Mean (sd)	Range	U	*p*-value
Age	47.77(13.45)	22 – 69	47.77(13.45)	22 – 69	84.5	.97
Level of Education	12.38(2.69)	7 – 17	12.77(2.38)	7 – 17	84.5	.97
Emotion attribution						
Reading the mind (/36)	21.85(6.38)	8 - 30	27.38(2.87)	22 – 31	37.5	.014
Nonverbal theory of mind						
Attribution of intention (/28)	22.46(6.67)	9 - 28	27.15(0.80)	26 – 28	26.5	.002
Verbal theory of mind						
Faux-pas detection (/10)	7.69(2.84)	0 – 10	9.00(0.91)	7 – 10	62.00	.26
FP understanding (%)	39.69(17.98)	0 – 57	52.92(5.87)	48 – 60	43.00	.034
Without FP detection (/20)	18.62(2.50)	12 – 20	18.92(1.75)	14 – 20	83.00	.96
Comprehension (/40)	37.15(4.36)	25 – 40	38.31(5.51)	20 – 40	40.50	.02
Verbal efficacy						
Semantic fluency (2 min)	18.00(7.13)	5 – 25	33.08(10.60)	15 – 53	15.0	< .001
Cognitive processing speed						
Word reading (45 s)	75.54(22.03)	25 – 101	100.23(18.32)	74 – 109	23.00	.001
Colour naming (45 s)	55.54(14.98)	40 – 77	76.46(11.19)	57 – 90	26.5	.002

Means and SDs of raw scores; * significance level $p < .05$

3.2. Executive function performance (Table 3)

Executive functioning was assessed by word fluency and cognitive speed tests (subtests A and B of the test of Stroop). The patients uttered roughly half as many words than controls in the fluency test (M_{SCA} = 18.00 *vs.* M_{Ctr} = 33.08; p = .002), indicating impaired verbal efficacy (p < .001). They also responded less accurately the general control questions in the faux-pas test (M_{SCA} = 37.15 *vs.* M_{Ctr} = 38.31; p = .02), suggesting a somewhat impaired comprehension of the fictious social situations described in the test. Their cognitive speed was also significantly decreased compared to the controls, as indicated by slower word-reading (M_{SCA} = 75.54 *vs.* M_{Ctr} = 100.23; p = .001) and color-naming in the Stroop test (M_{SCA} = 55.54 *vs.* M_{Ctr} = 76.46; p = .002).

3.3. Relationships of socio-affective performance with executive impairment and clinical features

Table 4 details the results for the correlation analyses of performance for the different neuropsychological tests. The first analysis revealed highly significant positive correlations of the scores in the two theory of mind tasks and of each one with emotion attribution performance (in each case, p < .001). The relationships with executive function were also significant, with a positive link between cognitive speed in the colour naming task and scores in the nonverbal (p = .001) and verbal theory of mind tasks (p = .026), and with emotion attribution performance (p = .001). They indicated that the more rapidly patients processed in the Stroop task, the higher were their socio-affective abilities. A similar relationship occurred between response speed in the word reading task and nonverbal theory of mind performance (p = .034), while it fell short of significance with emotion attribution performance (p = .056) and was nonsignificant with verbal theory of mind performance (p > .1). On contrary neither social cognition score was significantly linked to the word fluency score (p > .1), while semantic fluency and word reading speed were positively correlated (p = .009).

Finally, investigation of relations between test performance and clinical features showed no significant links with disease duration or age of diagnosis for any neuropsychological test score. A significant positive correlation was, however, observed between age of symptom onset and verbal fluency (ρ = .627, p = .012), indicating that the later symptoms occurred during the patients' life, the better their verbal fluency was and vice versa.

Table 4. Correlation analyses between social cognition abilities and executive function in the SCA patient group

	Nonverbal ToM	Verbal ToM	Semantic fluency	Word reading	Colour naming
Emotion attribution	.927*** (.001)	.787 *** (.001)	.245 (.420)	.541 (.056)	.810*** (.001)
Nonverbal ToM	/	.787*** (.001)	.218 (.474)	.588* (.034)	.823*** (.001)
Verbal ToM	/	/	.457 (.116)	.421 (.152)	.611* (.026)
Semantic fluency	/	/	/	.687** (.009)	.502 (.080)
Word reading	/	/	/		.890*** (.001)

*** stands for $p < .001$; ** stands for $p < .01$; * stands for $p < .05$;

ToM stands for theory of mind

4. Discussion

The present study assessed whether socio-cognitive performance was affected in patients with ataxia, both in a verbal and nonverbal task and whether performance impairments in this highly specific cognitive area could be accounted for by impairments in instrumental processes, *i.e.* speed and language efficacy. Overall, results revealed socio-affective and executive function impairments in SCA2 and 3 patients and significant relationships between the scores in the two task types but not with typical clinical features.

Results indicate that both theory of mind and emotion attribution were impaired in the patient group, on contrary to the dissociation reported in the literature between emotion attribution and theory of mind performance in different genotypes[5,10]. Furthermore, they do not support the finding of an opposite socio-affective profile according to genotype, with a specific emotion attribution impairment in SCA2 patients and a selective theory of mind impairment in SCA1 and 3 patients, that has been reported in the same studies. Rather, the present results indicate a generalized impairment in socio-affective abilities in this neurological condition, as subgroups displayed comparable impairments in the three tasks assessing social cognition abilities. In line with these findings, theory of mind deficits have been observed in different SCA genotypes, including SCA1, 2 and 3 genotypes, without apparent differences between subtypes[8]. These authors found no significant relationships of theory of mind performance with other neuropsychological measures or clinical features and proposed that social cognition impairments might be an important component of the SCA profile independently of clinical and demographic features[8]. Likewise, impaired social (but not basic) facial emotion recognition has been reported in different SCA genotypes, again independently of other cognitive abilities, including processing speed, flexibility and fluency[7].

On contrary to the results reported by D'Agata and co-workers[7], SCA2 and 3 patients participating in the present study displayed executive function impairments in addition to socio-affective disturbances. More especially, their cognitive speed was reduced in two versions of a verbal naming task and their word fluency decreased compared to controls. These findings are in line with previous research reporting impaired processing speed and fluency in SCA2 patients[29], in SCA3 patients[26], and in both genotypes[6,25]. In a follow-up study, Moriarty and collaborators[8] reported more important difficulties in SCA3 compared to SCA2 patients. At baseline, after 12 to 13 years of illness, they observed minor differences at odds with the present findings of slight but nonsignificant lower performance in SCA2 than SCA3 patients after a mean illness duration of 8 to 9 years. The differences were more marked at follow-up, seven to eight years later, particularly in a face recognition task, consistently with poor performance on nonverbal visual tasks associated with hypoperfusion in visual cerebral areas reported in SCA3 patients[8,37]. Though a more rapid cognitive decline in SCA3 patients may not be excluded, another study reported comparable performance for SCA2 and 3 patients in various neuropsychological tests, including verbal fluency and processing speed tasks after 8 and 13 years of illness respectively[6].

Furthermore, as expected, patients' socio-affective abilities were significantly associated with their processing speed. Their cognitive speed in a color-naming task was thus positively correlated with their performance in both theory of mind tasks and in the emotion attribution task. Even though correlation analyses demonstrate no causal relationship, the finding of a significant relationship with the three social cognition tasks indicates a robust link with cognitive speed. A similar relationship occurred also with the the word reading task but only for the nonverbal theory of mind task. As the tasks differ not only in respect with the verbal *vs.* nonverbal nature of the material and response modalities, but also by virtue of the processes explored, *i.e.* intention attribution, faux-pas detection and mental state attribution, the results favor the idea that reduced processing speed might account for socio-affective disturbances in these genotypes. Furthermore, the patients' word fluency performance was also correlated with processing speed in agreement with a prior study indicating that speed is a key-component of verbal fluency[24]. Interestingly, fronto-cerebellar connectivity has been shown to specifically mediate cognitive speed, indicating that an altered connectivity of this pathway might account for, or contribute to the socio-affective and cognitive impairments associated with cerebellar dysfunction[38].

Further, on contrary to our predictions, socio-affective abilities were not significantly associated with another instrumental ability, *i.e.* word fluency. Despite this negative finding,

comprehension difficulties were observed in the patients as in the verbal theory of mind task they displayed significant lower scores of faux-pas understanding and on the general control questions compared to controls. The latter results are in line with the comprehension impairments reported in several subtypes of ataxia[8]. Taken together, the present results suggest that impaired semantic processing and slower cognitive speed might more generally account for socio-cognitive disturbances in ataxia.

Clearly, future studies performing more systematic investigations and including a larger number of participants are necessary to further test this hypothesis based on preliminary data obtained in a reduced number of participants. Another limitation of the present study concerns the small numbers of patients within each SCA subtype, making comparisons difficult. Further, the Scale for the Rating and Assessment of Ataxia (SARA) had only been used in five patients to evaluate physical symptoms, so that onset age and disease duration were used to assess disease severity in patients. Interestingly however, a significant correlation was observed between word fluency and the age of symptom onset, but not with the age of illness diagnosis or with illness duration. In future studies it might thus be important to include the possibility of less formal clinical features, like symptom onset in addition to more traditional clinical features to assess relationships with cognition in ataxia. Including neuroanatomical results would also have been useful to discuss cognitive impairments in SCA patients in light of anatomical correlates, in particular of the cerebellum.

5. Conclusion

A generalized impairment in socio-affective abilities observed in SCA patients was significantly related to processing speed and comprehension difficulties (but not verbal fluency). Deficits in both cognitive areas were observed independently of the genetic sub-type and of clinical features (age of illness diagnosis or illness duration). Accordingly, though socio-affective disorders may represent a key component of the neuropsychological profile of this neurological condition as has been suggested by others, it may be mediated via executive function disturbances that have been largely documented in the literature. The implications of these findings may be important for cognitive remediation training in SCA patients.

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