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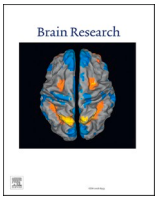

Review

A systematic review of social cognition in hereditary ataxia patients: Evidence from neuroimaging studies

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1. Introduction

Cerebellar ataxias represent a heterogeneous group of complex conditions in which cerebellum and connectivity networks are compromised (Manto et al., 2020; Palau and Arpa, 2023). The main characteristic symptoms are lack of coordinated movements and accuracy-related difficulties, leading to a wide range of motor signs including gait and posture, limb movements, speech and oculomotor abnormalities (Grimaldi, 2013). A broad nosological spectrum has been described for different diagnoses taking into consideration its etiology: degenerative, inherited or acquired disorders (Manto et al., 2020). According to their inheritance pattern, they can be classified as autosomal recessive cerebellar ataxia (ARCA), autosomal dominant cerebellar ataxia (ADCA), mitochondrial ataxia, X-linked forms and sporadic ataxia diagnosis (Palau and Espinós, 2013).

Scientific research has predominantly been focused on the genetic basis of the disease and the motor symptomatology (Bodranghien et al., 2016; Ilg et al., 2014). However, the involvement of the cerebellum in cognitive and affective processes is now widely accepted (Schmahmann, 2019). Regardless of the etiology, lesions or damage on the posterior cerebellar lobe, vermal area or deep nuclei could lead to the emergence of the neurocognitive and neuropsychiatric symptomatology, which would develop into the cerebellar cognitive-affective syndrome (CCAS) (Schmahmann, 2019; Schmahmann and Sherman, 1998). This syndrome includes a set of symptoms characterised by executive disturbances, linguistic deficits, visuospatial impairments, and affective dysregulation (Schmahmann and Sherman, 1998). In a study subsequent to the CCAS proposal, these authors further developed the affective component of the syndrome through a series of case studies (Schmahmann et al., 2007). Five major domains of symptomatology were established, each with positive and negative symptoms, including attentional control, emotional control, autism spectrum, psychosis spectrum and social skill set (Koziol et al., 2014; Schmahmann et al., 2007).

The role of the cerebellum in emotion has been discussed in a consensus paper, in which the experts identified specific cerebellar regions and their involvement in different stages of affective processes (Adamaszek et al., 2017). Neuroimaging studies have found some key areas dedicated to emotional processing and regulation, and mentalizing processes, in particular, lateral cerebellar hemispheres (lobules VI, Crus I, Crus II and IX), vermis and deep nuclei (Adamaszek et al., 2017; Baumann and Mattingley, 2012; Stoodley, 2012; Stoodley and Schmahmann, 2023). These regions are not only involved in perceptual and recognition stages, but also in integrative stages of affective evaluation (Adamaszek et al., 2017). This view is also supported by previous studies that highlighted the role of the vermis, the fastigial nucleus and the flocculonodular lobe in the emotion regulation, even considering them as part of Papez circuit (Schutter and van Honk, 2005). In this sense, anatomical relationships between the cerebellum and limbic and paralimbic brain regions have been identified, which would underlie the cerebellar participation in emotional and affective processes (Benagiano et al., 2018; Blatt et al., 2013), as well as the intrinsic connectivity between lobule IX and the default mode network, which is implicated in social cognition (Habas et al., 2013). Moreover, neuroscience research has evidenced both structural and functional anomalies in the cerebellum of patients with neuropsychiatric disorders (Schutter and van Honk, 2005).

Within the emotional or affective component related to cerebellum, the study of social cognition domain has received a growing interest in recent years (Van Overwalle et al., 2020). Researchers have found a worse performance on emotion attribution tasks in ataxia patients compared to healthy controls, suggesting that individuals with damage on the cerebellum can exhibit poor social skills (Hoche et al., 2016; Sokolovsky, 2010). Likewise, deficient theory of mind (ToM) and social emotion recognition capacity has been reported in spinocerebellar ataxia (SCA) patients (D'Agata et al., 2011; Garrard et al., 2008). Despite the increase in research on cognitive and affective functioning in ataxia patients, knowledge about social-cognitive profile in these pathologies remains scarce (Giocondo & Curcio, 2018). Moreover, there are studies

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that investigate neural correlates in cerebellar lesions, but patients' samples include different diagnoses such as cerebellar infarctions (Adamaszek et al., 2015) or neuropsychiatric disorders (Olivito et al., 2023). These issues limit the drawing of clear conclusions about a common social-cognitive profile for different cerebellar ataxia diagnoses. Moreover, they also highlight the importance of conducting studies with cerebellar ataxia patients in order to learn how the diagnosis affects such a necessary area as social cognition is in daily life.

Social behavioural expression is dependent on adequate emotional processing, that is why the aim of this study is to conduct a systematic review of previous research focused on social cognition assessment in patients with hereditary ataxia. We have limited our search to published works that include neuroimaging protocols in order to support the social-cognitive performance of ataxia patients with structural or functional findings.

2. Method

This study has been conducted considering the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) methodology.

2.1. Search strategy

A systematic review was carried out to identify original empirical studies that used social cognitive tests and neuroimaging in ataxia patients. We queried the following online databases: Pubmed, ProQuest, Scopus and Web of Science. The searching terms were (1) Ataxia, (2) Neuroimaging, (3) Neuroimage, (4) Social cognition, (5) Theory of mind, (6) Social thinking, (7) Mentalizing, (8) Social cognitive functioning and (9) Social skills. Search strategy was made with all of the possible combinations between these keywords (see Table 1 for details). The literature search was conducted until June the 1st, 2023.

2.2. Inclusion and exclusion criteria

Studies were included if (a) reported human cases diagnosed with hereditary ataxia, (b) used social cognition tests or questionnaires, c) used neuroimaging techniques and (d) were published in the last 15 years. However, literature reviews, *meta*-analysis and conference papers were excluded, along with papers not written in english or spanish and other non-hereditary ataxia diagnosis.

2.3. Data extraction

The search strategy identified 1678 articles. After removing 1194 duplicates, 447 articles were left for screening. A total of 37 papers were selected because they met inclusion criteria, although only 9 eligible articles included social cognition assessments (see Fig. 1 for flow diagram). Studies that met inclusion criteria were summarised according to (i) the authors, (ii) study location, (iii) experimental and control sample size, (iv) ataxia diagnosis, (v) demographic variables, (vi) illness status, (vii) social cognition assessment, (viii) neuroimaging findings and (ix) conclusions (Table 2).

Table 1
Search strategy for the databases.

First column (Title/Abstract/ Keywords)	Second column (Full text)	Third column (Full text)
Ataxia	Neuroimage Neuroimaging	Social cognition Theory of Mind Social thinking Mentalizing Social cognitive functioning Social skills

Note. Search strategy combined the terms in the first, second and third column.

2.4. Risk of bias

The modified Newcastle-Ottawa Scale (Bawor et al., 2014) was used to evaluate the risk of bias of the eligible articles. The aim of this scale is to examine the quality of each study in the following domains: sample selection, control confounding, statistical and outcomes (Table 3).

3. Results

3.1. General overview

All the articles were case-control series, in exception of one paper which was a longitudinal study without control group (Moriarty et al., 2016) (Table 2). Hereditary ataxia diagnosis differed in each article; the most prevalent diagnosis was SCA 2, with a total of 54 patients (Clausi et al., 2019; Clausi et al., 2021a; Clausi et al., 2021b; Mercadillo et al., 2015; Moriarty et al., 2016), SCA1 was diagnosed in 22 patients (Clausi et al., 2019; Clausi et al., 2021b; Contali et al., 2023; Moriarty et al., 2016), 22 patients were diagnosed with Friedreich ataxia (FRDA) (Clausi et al., 2019b; Dogan et al., 2016), a total of 12 patients were diagnosed with spastic paraplegia (SPG) 7 (Clausi et al., 2021b; Lupo et al., 2020), SCA6 was diagnosed in 5 patients (Clausi et al., 2021b; Moriarty et al., 2016), SCA15 was diagnosed in 4 patients (Clausi et al., 2019; Clausi et al., 2021b), 3 patients were diagnosed with SCA3 (Moriarty et al., 2016) and SCA7 (Moriarty et al., 2016) and, finally, 2 patients were diagnosed with SCA28 (Clausi et al., 2019; Clausi et al., 2021b). There was one article that did not specify the number of participants in each hereditary ataxia type, although it did mention that it counted with SCA1, SCA3 and SCA6 diagnoses (Lopes da Cunha et al., 2022). Relative to the sample, 63 participants identified as female whereas 56 as men, although two papers did not specify this information (Lopes da Cunha et al., 2022; Moriarty et al., 2016). The mean age of the sample was constant in all the articles, 39.94 was the minimum mean age (Moriarty et al., 2016), while 46.69 was the maximum (Clausi et al., 2019). Moreover, sample size was small in most cases, with less than 20 participants (Clausi et al., 2021a; Contali et al., 2023; Dogan et al., 2016; Lupo et al., 2020; Lopes da Cunha et al., 2022; Mercadillo et al., 2015; Moriarty et al., 2016), in exception of two articles (Clausi et al., 2019; Clausi et al., 2021b). Illness status was assessed by different methods, such as the Scale for the Assessment and Rating of Ataxia (SARA) (Contali et al., 2023; Dogan et al., 2016; Lopes da Cunha et al., 2022; Mercadillo et al., 2015; Moriarty et al., 2016), the International Cooperative Ataxia rating Scale (ICARS) (Clausi et al., 2019; Clausi et al., 2021a; Clausi et al., 2021b; Lupo et al., 2020) and the Inventory of Non-Ataxia Signs (INAS) (Dogan et al., 2016; Moriarty et al., 2016). The severity of the illness ranged between articles; SARA's mean punctuation ranged from 11.53 (Contali et al., 2023) to 20.1 (Dogan et al., 2016), ICARS' mean also varied from 25.17 being the minimum score (Lupo et al., 2020) to 34 being the maximum (Clausi et al., 2019) and INAS' mean punctuations also ranged from 2.38 (Moriarty et al., 2016) to 4.73 (Dogant et al., 2016).

The analysis of risk of bias of the eligible articles showed that the main domains that had compromised evidence were methods for selecting sample and control confounding (Table 3). Future studies should focus on improving these methodological issues.

3.2. Social cognition

Social cognition tests selected to assess social cognition were Faux Pas test, The Advanced ToM test, the Emotion Attribution (EA) test and the Reading the Mind (RME) test, which are some of the most common tools used to evaluate this matter. In the following paragraphs, specific information about the results obtained in each test will be given.

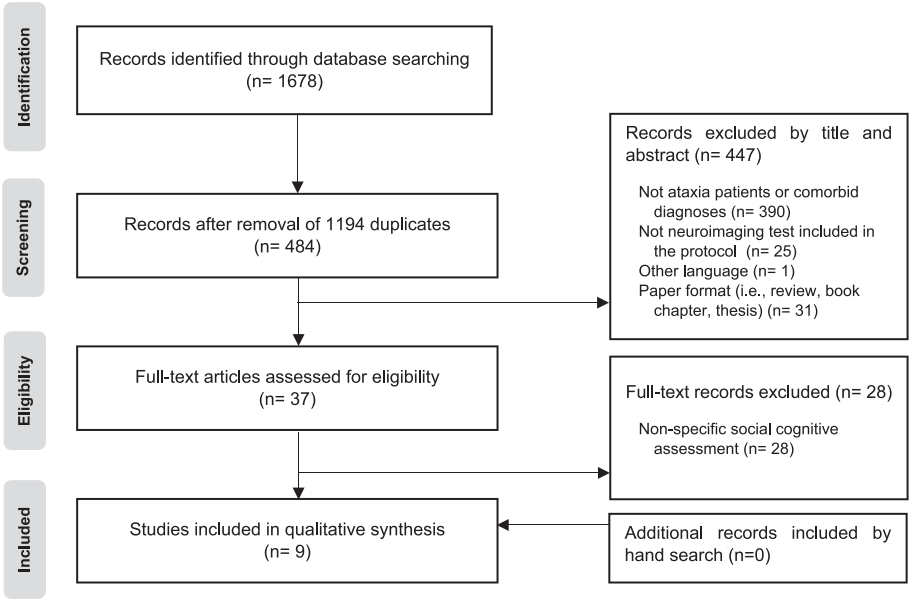

Fig. 1. Flow diagram of the literature search.

3.3. *Faux Pas Test*

Faux Pas test (Stone et al., 1998) is one of the most commonly tools to assess ToM and consists of short stories that represent social scenarios where the participant has to infer the thoughts and feelings of others in a 'faux pas' situation (a declaration or action that accidentally offends another person) (Faísca et al., 2016). A total of 4 articles used this test (Clausi et al., 2019; Clausi et al., 2021a; Clausi et al., 2021b; Dogan et al., 2016). In first place, patients with FRDA diagnosis obtained lower scores in "faux-pas" stories (Clausi et al., 2019, Clausi et al., 2021b; Dogan et al., 2016) and in the cognitive component (Clausi et al., 2021b; Dogan et al., 2016). More specifically, Dogan et al. (2016) reported worst performance in the assessment of intentions and empathy with no significant differences in neutral stories or control questions. Moreover, Faux Pas test was assessed to SCA2 patients, which also obtained lower scores in "faux-pas" stories (Clausi et al., 2019; Clausi et al., 2021a; Clausi et al., 2021b), specifically in the cognitive component (Clausi et al., 2021a; Clausi et al., 2021b). In other types of ataxia, such as SPG7, SCA6, SCA1, SCA15 and SCA28 coincident results were found with lower scores in "faux-pas" stories (Clausi et al., 2019; Clausi et al., 2021b).

3.4. *The Advanced ToM Test*

The Advanced ToM test (Happè, 1994) is used to assess more advanced concepts of ToM, such as double bluff, white lies and persuasion (Clausi et al., 2019). This test was used in 3 articles (Clausi et al., 2019; Lupo et al., 2020; Moriarty et al., 2016). Results regarding patients with SPG7 diagnosis obtained pathological results in 66 % of the cases (Lupo et al., 2020), whereas patients with SCA1, SCA2, SCA15, SCA28 and FRDA also obtained impaired results (Clausi et al., 2019). Moreover, Moriarty et al. (2016) evaluated longitudinally patients with SCA1, SCA2 and SCA7 and reported worsening of results to varying degrees compared with baseline scores. The SCA7 group demonstrated the smallest decrease in score over time.

3.5. *The Emotion Attribution Test*

The EA test (Blair and Cipolotti, 2000) is used to assess the ability to attribute emotions to others in a social context. A total of 4 articles assessed their sample with the EA test (Clausi et al., 2019; Clausi et al., 2021a; Contali et al., 2023; Lupo et al., 2020). Regarding SCA1 diagnosis results were not consistent among articles. In the study by Contali et al. (2023), 25 % of the sample showed impaired performance, specifically recognition of fear and anger was mostly affected, whereas Clausi et al. (2019) did not find impairment in EA test total score. In the same line, contradictory results were also found regarding SCA2 participants; while Clausi et al., (2021a) reported worse performance when the stories represented "anger", in previous work from the same authors did not find impairment in EA test scores (Clausi et al., 2019). Moreover, only Lupo et al. (2020) assessed the EA test on SPG7 participants, which obtained pathological results in the "embarrassment" emotion. Other diagnoses, such as SCA15, SCA28 and FRDA did not show worse performance in this test (Clausi et al., 2019).

3.6. *Reading the Mind Eye Test*

The RME test (Baron-Cohen et al., 2001) assesses the ability of a person to recognize another person's mental state. This test was used in 4 articles (Clausi et al., 2019; Clausi et al., 2021a; Clausi et al., 2021b; Lupo et al., 2020). SCA2 participants obtained impaired performance (Clausi et al., 2019; Clausi et al., 2021; Clausi et al., 2021), as also did SPG7 participants (Clausi et al., 2021b; Lupo et al., 2020). Other diagnoses such as SCA1, SCA6, SCA15, SCA28 and FRDA showed low scores (Clausi et al., 2019; Clausi et al., 2021b).

3.7. *Other assessments*

Two articles did not use a standardised test to assess social cognition (Lopes da Cunha et al., 2022; Mercadillo et al., 2015). Lopes da Cunha et al. (2022) evaluated the level of comprehension with two stories: a social text, which contained interpersonal events, and a non-social text, presenting actions from a single character. The results showed that SCA participants scored worse in the social text, but there were no significant results for non-social text results. Moreover, statistical analysis confirmed that the selective deficit in social text processing was not driven by mnesic skills. Secondly, Mercadillo et al. (2015) used semi-structured interviews that focused on social cognition issues and concluded that social abilities, cognitive and emotional alterations were present in ataxia patients.

Table 2
Summary of the eligible articles to review.

Author	Study location	*N (E/C)*	Ataxia diagnosis	Demographic variables		Illness status	Social cognition assessment	Neuroimaging findings	Conclusions
				Age (M ±SD) (min. - max.)	Gender (n/%)	M±SD (min. - max.)			
Mercadillo et al. (2015)	Mexico	15/15	SCA-2	37.2 ± 15.9 (15-65)	F: 9 (60%) M: 6 (40%)	SARA: 17.2 ± 8.5 (2-33.5)	Semi-structured interviews (patients and relatives) and participant observation, focused on social cognition issues. Social abilities, cognitive and emotional alterations.	MRI (3T): *VBM* Degeneration in cerebellar vermis (uvula IX) and brainstem, in cortical and subcortical regions (insula, parahippocampal gyrus, precentral, frontal, parietal, temporal cortices).	Cortical regions are involved in social interactions. Correlation between social cognition assessment and neuroimaging findings not specified.
Dogan et al. (2016)	Germany	15/15	FRDA	37.73 ±13.57	F: 7 (46.7%) M: 8 (53.3%)	SARA: 20.1 ±7.34 INAS: 4.73 ±2.37	Faux Pas FRDA obtained lower scores in the general result, especially in the assessment of intentions and empathy. No significant results in neutral stories or control questions.	MRI (3T): *VBM* Cerebellar GM atrophy in the bilateral lobule VI. GM reductions in posterior cerebellar lobules bilaterally (VI, Crus I/II, VIIb) and in left motor cortex. WM degeneration in the medulla extending bilaterally to the inferior cerebellar peduncle and in the bilateral SCP, adjacent to the dentate nucleus area. Atrophy in the midbrain. No significant increases in GM or WM in patients vs controls. *DTI* Widespread decreased FA in bilateral SCP, ICP, corticospinal tract, cerebral peduncles, fornix, posterior thalamic radiation, corpus callosum, internal capsule, corona radiata, forceps major, inferior longitudinal and fronto-occipital fasciculus and left hippocampal cingulum.	Correlation between social cognition assessments and neuroimaging findings not specified.
Moriarty et al. (2016)	UK	13/-	SCA1: 2 SCA2: 2 SCA3: 3	At onset: 36.94 ±6.99 (25-59)	-	SARA: Baseline: 13.3±4.3	The Advanced ToM SCA1, SCA2 and SCA7 above threshold of impairment at baseline, whereas at follow-up, all groups scored below threshold to varying degrees. SCA7 group demonstrated the smallest decrease in scores over time.	MRI (1.5 and 3T): General cerebral atrophy in all patients, but in two SCA3 cases moderate or severe atrophy. Brainstem atrophy moderate or severe in all but two cases.	Correlation between social cognition assessments and neuroimaging findings not specified.

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Table 2 (*continued*)

Author	Study location	*N (E/C)*	Ataxia diagnosis	Demographic variables		Illness status	Social cognition assessment	Neuroimaging findings	Conclusions
				Age (M ±SD) (min. - max.)	Gender (n/%)	M±SD (min. - max.)			
			SCA6: 4			(9-23.5)		SCA1 and SCA2 greatest degree of cerebellar atrophy.	
			SCA7: 3	At baseline:		Follow-up:		SCA6 patient with cerebellar atrophy but no brainstem atrophy.	
				43.66 ±5.95		18.92 ±4.83		9 patients subtle linear sagittal hyperintensities in the midline of the pons.	
				(36-65)		(13-28)		Mild cortex atrophy in 6 patients.	
								SCA2 patient with mild parietal cortex atrophy.	
						INAS		Mild subcortical atrophy in all SCA7 patients.	
						Baseline: 2.38±1.6 (0-5) Follow-up: 4±2.41 (1-9)			
Clausi et al. (2019)	Italy	27/27	SCA1: 2	46.4 ±10.8	F: 21 (77.7%)	ICARS:	RME; EA; Faux Pas; The Advanced ToM	MRI (3T):	Correlation between social cognition assessment and neuroimaging findings not specified.
			SCA2: 11	(24-60)	M: 6 (22.3%)	34±34.6	Impaired performance on RME and Advanced ToM task. In the faux pas test, lower scores in 'faux-pas' stories.	*VBM*	
			SCA15: 2			(8-59)	Normal scores in the EA test total score.	Decreased GM volumes in the left and right lobules I-IV (anterior cerebellum), in the right lobule VI with extension in the left side and vermis portion, and in the left and right Crus I-II.	
			SCA28: 1					Decreased GM volumes in the bilateral hemispheric lobule VIIIa and vermis VIIIa with extension in vermis IX.	
			FRDA: 2 ICA: 8 Cerebellitis: 1					*Seed-Based analyses* (fMRI) Anterior cerebellum: decreased FC between lobules I-IV and precentral gyrus, postcentral gyrus, rolandic operculum, inferior frontal gyrus (motor and somatosensory cortex); supramarginal gyrus, anterior and posterior cingulate cortex, left orbitofrontal cortex and middle frontal gyrus (mentalizing processes). Intermediate cerebellum: decreased FC between lobule VI and middle frontal gyrus, left premotor cortex, inferior frontal gyrus and temporal pole area. Posterior cerebellum: decreased FC between left crus I-II and middle frontal gyrus, dorsomedial prefrontal cortex, superior frontal gyrus and orbitofrontal cortex; and decreased FC between right	

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Table 2 (*continued*)

Author	Study location	*N (E/C)*	Ataxia diagnosis	Demographic variables		Illness status	Social cognition assessment	Neuroimaging findings	Conclusions
				Age (M ±SD) (min. - max.)	Gender (n/%)	M±SD (min. - max.)			
								crus I and left inferior frontal gyrus and precentral gyrus (abstract aspects of social cognition). Vermis: decreased FC between VI, VIIIa and IX and middle frontal gyrus, anterior cingulate cortex, premotor cortex and supplementary motor areas, orbitofronatal cortex, inferior frontal gyrus, angular gyrus and superior temporal sulcus (emotional processing and mentalizing networks).	
Lupo et al. (2020)	Italy	6/30[a]	SPG7	46.33 ±12.93 (23-55)	F: 0 (0%) M: 6 (100%)	ICARS: 25.17 ±18.49 (7-56)	RME; EA; The Advanced ToM SPG7 obtained pathological results in the Tom (66%), the RME (83%) and the "embarrassed" emotion of the EA test.	MRI (3T): *VBM* GM atrophy in the cerebellar cortex (right lobule V and bilateral lobule VI with extension in the right Crus I). GM atrophy in the cerebral cortex (cortical and subcortical regions in the left and right hemispheres). *Seed-Based analysis* FC alterations (increased FC between the left and right DN and ipsilateral cerebral regions).	Correlation between social cognition assessments and neuroimaging findings not specified.
Clausi et al. (2021a)	Italy	13/33[b]	SCA-2	46.69 ±8.69 (3-65)	F: 7 (53.8%) M: 6 (46.2%)	ICARS: 28.31 ±7.38 (17-47)	RME; EA; Faux Pas SCA2 showed worse performance in RME.	MRI (3T): *VBM*	RME total score positively correlated with the mean FA value from MCP and form the right SCP. The patient's "anger" score from the EA was negatively correlated with mean AD and MD values from the MCP. The same value positively correlated with the GM volume in the right lobule VIIIB, the right lobule IX and the left Crus II.
							SCA2 performed worse in EA when the stories elicited "anger".	GM loss in SCA2 in the lobules I-IV of the anterior cerebellum, the right lobule VI, the left crus II and the lobule IX bilaterally.	Total score and the cognitive component score of the Faux Pas positively correlated with the GM volumes in the right Crus II. No correlation between any considered diffusion parameter and patients' Faux Pas scores.
							SCA2 performed worse in Faux Pas in the cognitive component of the Faux Pas stories.	Reduced GM volume in the left occipital fusiform gyrus. No significant differences in total GM volume.	

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Table 2 (*continued*)

Author	Study location	*N* (*E*/*C*)	Ataxia diagnosis	Demographic variables		Illness status	Social cognition assessment	Neuroimaging findings	Conclusions
				Age (M ±SD) (min. - max.)	Gender (n/%)	M±SD (min. - max.)			
								DTI FA reduced and increased MD, AD and RD in the MCP and in the SCP.	
Clausi et al. (2021b)	Italy	36/67	SCA1: 2 SCA2: 13 SCA6: 1 SCA15: 2 SCA28: 1 FRDA: 2 SPG7: 6 ICA: 8 Cerebellitis:1	46.97 ±10.7 (24-64)	F: 21 (58.3%) M: 15 (41.7%)	ICARS: 26.52 ±12.4	RME; Faux Pas Lower scores on RME and Faux Pas total and cognitive component scores.	MRI (3T)[c] *VBM* Reduced GM volumes at anterior and posterior cerebellar hemispheres (right and left lobules I-IV and right lobule VI, with extension to the right Crus I-II, left Crus I-II). Reduced GM in the putamen and caudate, orbitofrontal cortex, superior frontal gyrus, lingual gyrus and fusiform gyrus.	No correlation between ToM scores and GM volume.
Lopes da Cunha et al. (2022)	Chile	15/29	SCA1 SCA3 SAC6	-	-	SARA: 13.50 ±8.32	All participants listen to two stories: a social text, highlighting interpersonal events, and a non-social text, presenting actions of a single character. SCA participants obtained lower scores in social text outcomes and no significant results for non-social text results.	MRI: *VBM*	No significant association was found between the SCA participants and the non-social text outcomes. Analysis that collapsed the SCA group and the HC group correlated positively with connectivity between the cerebellar culmen and the cerebellar tonsil with posterior regions.
Contaldi et al. (2023)	Italy	16/16	SCA-1	47.69 ±8.16	F: 7 (43.7%) M: 9 (56.3%)	SARA: 11.53 ±5.14	EA 25% of the sample showed impaired performance on EA (recognition of fear and anger was mostly affected).	*Seed-Based analysis* (fMRI) ERP (Keypoint™ software); EEG Increased latencies of N200 and P300, decreased amplitudes of N100 and P300.	Significant inverse correlation between P300 latency and EA score.

Note. ICA and cerebellitis diagnoses reported by Clausi et al. (2019) and Clausi et al., (2021b) were excluded from the total sample due to not meeting the inclusion criteria.

[a] Only MRI analysis; [b] Only 26 healthy subjects for the behavioral and magnetic resonance imaging study; [c] Only 32 underwent MRI protocol.

AD: Axial diffusivity; DTI: Diffusion Tensor Imaging; DN: Dentate Nucleus; E/C: Experimental/Control; EA: The Emotion Attribution test; EEG: Electroencephalography; ERP: Event Related brain Potentials; FA: Fractional anisotropy; FC: Functional Conectivity; fMRI: Functional Magnetic Resonance Imaging; FRDA: Friedreich's Ataxia; GM: Grey Matter; HS: Healthy subjets; ICA: Idiopathic Cerebellar Atrophy; ICARS: International Cooperative Ataxia Rating Scale; ICP: Inferior Cerebellar Peduncle; INAS: Inventory of Non-Ataxia Signs; MCP: Middle cerebellar peduncle; MD: Mean diffusivity; MRI: Magent Ressonance Imaging; RD: Radial diffusivity; ROI: Region Of Interest; SARA: Scale for the Assessment and Rating of Ataxia; SCA: Spinocerebellar ataxia; SCP: Superior cerebellar peduncle; SPG: Spastic Paraplegia; RD: Radial diffusivity; RME: The Reading Mind in the Eyes Test; ToM: The Theory of Mind; VAS: Visual Analogue Test; VBM: Voxel-Based Morphometry; WM: White Mater.

4. Neuroimaging

4.1. Voxel Based Morphometry

Voxel Based Morphometry (VBM) measures differences in local concentrations of brain tissue through a voxel-wise comparison of multiple brain images. This technique has been the most used among the articles reviewed. On the one hand, SCA2 participants showed cerebellar degeneration in the cerebellar vermis (uvula IX) (Mercadillo et al., 2015), grey matter (GM) loss in the lobules I-IV of the anterior

Table 3
Risk of bias assessment for reviewed studies.

	Method for selecting sample	Methods to control confounding		Statistical methods		Methods for measuring outcomes	
		Sample size	Identification of confounders	Appropriate analyses	Missing data	Outcome measures	Objective assessment
Mercadillo et al. (2015)	Moderate	High	High	High	Moderate	High	Moderate
Dogan et al. (2016)	Moderate	High	Moderate	Low	Low	Moderate	Low
Moriarty et al. (2016)	Moderate	High	Moderate	High	Low	Moderate	Moderate
Clausi et al. (2019)	Moderate	Moderate	Moderate	Low	Low	Low	Low
Lupo et al. (2020)	Moderate	High	Moderate	High	Low	Low	Low
Clausi et al., (2021a)	Moderate	High	Low	Low	Low	Low	Low
Clausi et al., (2021b)	Moderate	Low	Low	Moderate	Low	Low	Low
Lopes da Cunha et al. (2022)	Moderate	High	Moderate	High	Moderate	Low	Low
Contaldi et al. (2023)	Moderate	High	Moderate	Low	Low	Low	Low

Note. The modified Newcastle-Ottawa scale (Bawor et al., 2014) has been used to analyze the risk of bias of the studies reviewed.
High risk of bias: High; Moderate risk of bias: Moderate; Low risk of bias: Low.

cerebellum (with extension to the left lobules V, VI and Crus I), the right lobule VI (with extension to the right Crus I and II), the left Crus II and the lobule IX bilaterally (with extension to lobule VIIb) (Clausi et al., 2021a). Moreover, other cerebral regions affected were the brainstem and cortical and subcortical regions, such as the insula, parahippocampal gyrus, precentral, frontal, parietal and temporal cortices (Mercadillo et al., 2015), although other studies found reduced GM volume in the left occipital fusiform gyrus but no significant differences in total GM volume (Clausi et al., 2021a) and mild parietal cortex atrophy (Moriarty et al., 2016). Moriarty et al. (2016) compared neuroimaging findings between different ataxia diagnoses and concluded that SCA2 and SCA1 participants showed the greatest degree of cerebellar atrophy. In FRDA participants, VBM results had cerebellar GM atrophy in the bilateral lobule VI, together with the midbrain, and GM reductions in posterior cerebellar lobules bilaterally (VI, Crus I/II, VIIb) and in the left motor cortex. Furthermore, white matter degeneration was found in the medulla extending bilaterally to the inferior cerebellar peduncle (ICP) and in the bilateral superior cerebellar peduncle (SCP), adjacent to the dentate nucleus area (Dogan et al., 2016). Regarding SPG7 participants, GM atrophy was found in the cerebellar cortex (right lobule V and bilateral lobule VI with extension in the right Crus I) and also in the cerebral cortex (cortical and subcortical regions in both hemispheres). Moreover, Moriarty et al. (2016) counted with a heterogeneous ataxia diagnoses sample and therefore obtained different neuroimaging findings. For instance, a SCA6 patient showed cerebellar atrophy but no brainstem degeneration, whereas SCA7 participants had mild subcortical atrophy. General findings regarding all the sample were mild cortex atrophy, found in 6 patients, subtle linear sagittal hyperintensities in the midline of the pons in 9 participants and general cerebral atrophy in all participant but in two SCA3 cases which showed moderate or severe atrophy (Moriarty et al., 2016). Finally, other studies did not specify what neuroimaging findings referred to certain diagnosis, instead gave a general overview regarding different types of ataxia entities. For instance, Clausi et al. (2019) included in their sample diagnoses such as SCA1, SCA2, SCA15, SCA28 and FRDA and found decreased GM volumes in the cerebellar cortex; in the left and right lobules I-IV (anterior cerebellum), the right lobule VI with extension in the left side and vermis portion, and in the left and right Crus-I-II.

4.2. Diffusion Tensor Imaging

Diffusion Tensor Imaging (DTI) is used to study white matter architecture. In this review, only two articles used this neuroimaging technique. Dogan et al. (2016) detected widespread decreased Fractional Anisotropy (FA) in patients with FRDA in bilateral SCP, ICP, corticospinal tract, cerebral peduncles, fornix, posterior thalamic radiation, corpus callosum, internal capsule, corona radiata, forceps major, inferior longitudinal and fronto-occipital fasciculus and left hippocampal cingulum. The second article evaluated patients with SCA2 diagnosis and showed reduced FA and increased mean diffusivity (MD), axial diffusivity (AD) and radial diffusivity (RD) in the middle cerebellar peduncle (MCP) and in the SCP (Clausi et al., 2021a).

4.3. Seed-based analysis

Seed-based analysis explores functional connectivity (FC) within the brain. Lupo et al. (2020) used seed-based analysis and detected increased FC between the left dentate nucleus (DN) and ipsilateral regions, including the lateral occipital cortex, the supramarginal gyrus, and the superior parietal lobule. Similarly, there was also increased FC between the right DN and ipsilateral regions, involving the lateral occipital cortex and the precuneus. Furthermore, Clausi et al. (2019) also used this technique to evaluate SCA1, SCA2, SCA15, SCA28 and FRDA and obtained general neuroimaging findings. In particular, certain cerebellar regions were detected with decreased FC. In first place, the anterior cerebellum showed reduced FC between lobules I-IV and precentral gyrus, postcentral gyrus, rolandic operculum, inferior frontal gyrus (motor and somatosensory functions), supramarginal gyrus, anterior and posterior cingulate cortex, left orbitofrontal cortex and middle frontal gyrus (mentalizing functions). Secondly, the intermediate cerebellum showed decreased FC between lobule VI and middle frontal gyrus, left premotor cortex, inferior frontal gyrus and temporal pole area. Moreover, the posterior cerebellum had decreased FC between left Crus I-II and middle frontal gyrus, dorsomedial prefrontal cortex, superior frontal gyrus and orbitofrontal cortex; and decreased FC between right Crus I and left inferior frontal gyrus and precentral gyrus (abstract aspects of social cognition). Finally, the vermis showed decreased FC between VI, VIIIa and IX and middle frontal gyrus, anterior cingulate cortex, premotor cortex and supplementary motor areas, orbitofrontal cortex, inferior frontal gyrus and superior temporal sulcus (emotional processing and mentalizing networks) (Clausi et al., 2019).

4.4. Social cognition and neuroimaging

Out of the 9 chosen articles, only 4 articles carried out statistical correlations between social cognition findings and neuroimaging results (Clausi et al., 2021a; Clausi et al., 2021b; Contali et al., 2023). Mercadillo et al. (2015) also suggested neuroanatomic regions such as cortical areas that were involved in social interaction, however, they did not use statistical analysis to verify these results. Clausi et al., (2021a) were the ones that provided more information regarding SCA2 participants. Specifically, RME test total score positively correlated with the mean FA value from MCP and the right SCP. Moreover, the patient's "anger" score from the EA test was negatively correlated with AD and MD mean values from the MCP. This value also correlated with the GM volume in the right lobule VIIIb, the right lobule IX and the left Crus II. Finally, total

score and the cognitive component score of the Faux Pas positively correlated with the GM volumes in the right Crus II. In contrast, no correlation was found between any considered diffusion parameter and patients' Faux Pas scores, and The Advanced ToM test scores and GM volume. Moreover, no correlation was also found by the same authors between The Advanced ToM test scores and GM volume in another study (Clausi et al., 2021b). On the other hand, Lopes da Cunha et al. (2022) obtained significant correlations collapsing the SCA group and the HC group between the cerebellar culmen and the cerebellar tonsil with posterior regions, such as calcarine, right superior temporal, supramarginal and transverse temporal gyri and bilateral cuneus. However, the authors did not mention any correlations between the social outcomes and the SCA group subtype (Lopes da Cunha et al., 2022).

5. Discussion

Over the past decade, interest in the emotional or affective component related to the cerebellum has increased. To our knowledge, no critical analysis about social cognition and neuroimaging findings associated with cerebellar ataxias has previously been performed. Unfortunately, this systematic review revealed there is limited empirical research in investigating the role of the cerebellum in social cognition supported by neuroimaging techniques. Moreover, the studies that do have numerous limitations. First, sample size was small in most cases and the number of participants diagnosed with different subtypes of cerebellar ataxia differed significantly between studies, being SCA2 the most represented subtype (see Table 2 for the rest of the diagnosis). Moreover, the studies reviewed did not consider the severity of the condition, which could play a crucial role on the social cognition functioning. In total, 128 participants were included in this review and age ranged between 15 and 65 years old. All of the articles were case-studies, although sample size was small in most studies. Second, social cognition protocol was different in each article, although four main tests were used: Faux Pas test, The Advanced ToM test, the EA test and the RME test. Illness status was also assessed with different instruments, limiting the comparison of the symptom's severity. Moreover, this limitation makes it difficult to relate cognitive performance with the degree of severity of the disease, which would be very useful for the better understanding of the pathology course. Only one study followed participants' social cognition performance longitudinally, which allowed to know the progression of each cerebellar ataxia subtype (Moriarty et al., 2016). Finally, we acknowledge some risk of bias in extracting results from the articles included in this review, mainly in sample selection and confounders identification. The final search included 9 eligible articles for reviewing which reflected the deficits in social cognition through neuropsychological assessments and neuroimaging techniques that people with cerebellar ataxia diagnosis have.

Regarding social-cognitive outcomes, generally all participants, regardless of the ataxia subtype, obtained lower scores in "faux-pas" stories, which indicates a deficit in identifying stories that offend someone in some way. Specifically, the cognitive component was also impaired in FRDA and SCA2 (Clausi et al., 2021b; Dogan et al., 2016). Moreover, the assessment of intentions and empathy were reported to be worse in cerebellar ataxia participants. In contrast, Sayah et al. (2018) assessed FRDA participants and the results were similar to the general population. Instead, cerebellar hereditary and non-hereditary disorders showed impaired scores in the Faux Pas test compared to healthy controls (Tamaš et al., 2021). In other cerebellar pathologies, such as non-demented patients with isolated cerebellar degenerative diseases, participants performed worse in belief questions and in the Faux Pas test, which is widely used and requires a high level of complexity in ToM capacity (Abel et al., 2007).

Contradictory results were found in this review regarding the EA test. Specifically, the same authors obtained contradictory results in two articles written by their research team, although the newest study confirmed that SCA2 participants performed worse when the stories represented "anger", while in the previous one the sample obtained normal total scores (Clausi et al., 2019; Clausi et al., 2021a). SPG7 participants obtained pathological results in the "embarrassment" emotion (Lupo et al., 2020), while the other subtypes of ataxia did not show impaired performance. Therefore, these results suggest that an impaired performance in the EA test may depend on the ataxia subtype and, moreover, there would be certain diagnoses in which this function would be preserved. In other studies, patients with SCA2 and SCA7 showed impaired performances in story-based assessments of emotion attribution (Sokolovsky et al., 2010), whereas SCA6 did not (Garrad et al., 2008). Moreover, picture-based face emotion attribution tasks also reported worst scores in SCA2, SCA6, SCA7 and SCA8 in the recognition of emotional stimuli, highlighting a more pronounced impairment for negative, such as guilt, and positive emotions compared to neutral (D'Agata et al., 2011). Same results were found by Costabile et al. (2018), where FRDA patients failed in the recognition of sadness and three social emotions (pleasure, amusement and interest) (Costabile et al., 2018). In this review, worst performances were found regarding negative emotions, not mentioning positive stimuli. In the same line, Hoche et al. (2016) also corroborated this hypothesis by using the RME test and pointed out, as well, that cerebellar pathology alone is sufficient to produce these deficits in emotion attribution. Moreover, these authors found an even larger impairment for positive emotions (Hoche et al., 2019), which is contradictory with previous studies of whole-face perception that did not demonstrate this deficit in decoding positive stimuli (Adamaszek et al., 2014) and as shown in this review. Therefore, these findings suggest cerebellar patients utilise additional perioral facial cues to decode positive emotions, such as mouth regions (Hoche et al., 2016). In the same line, all participants that were assessed with the RME test test showed low scores regardless of the ataxia subtype. Story-based tasks allow subjects to utilise other reasoning abilities to compensate for a weakness in the EA test, meanwhile the RME test is a more pure measure of emotion attribution. In other pathologies, such as cerebellar stroke, patients also showed impairment in the ability to recognize emotions, particularly for negative stimuli (Adamaszek et al., 2014). Other authors found that patients with cerebellar hereditary and nonhereditary disorders obtained significantly lower scores than healthy controls in the RME test (Tamaš et al., 2021).

As in other cognitive assessments, ataxia participants performed worse than controls in The Advanced ToM test. Even so, a longitudinal study demonstrated worsening of the scores in SCA1, SCA2 and SCA7 participants, even though the last group had the smallest decrease over time. These results go in line with the ones found by García et al. (2022), who demonstrated that hereditary ataxia participants performed worse in The Advanced ToM test and such results were not influenced by physical or psychological status of the clinical group.

Regarding the accumulating evidences through the neuroimaging techniques, it is well known that cognitive tasks engage different cerebro-cerebellar circuits, including lobules VI and VII and the prefrontal and parietal cortices (Stoodley et al., 2012). A *meta*-analysis of cerebellar activation patterns demonstrated that emotional processing paradigms activated bilateral clusters in VI-Crus I and the midline posterior region in VIIAt (Stoodley and Schmahmann, 2009). Consistent with the idea that the cerebellum plays an important role in social cognition, cerebellar structural alterations interfere with the modulatory function of the cerebellum on the cerebral areas involved in social cognition (Van Overwalle et al., 2020). Hoche et al. (2016) demonstrated that cerebellar pathology is sufficient to generate deficits in emotion attribution similar to pathology located in cerebral hemispheric limbic structures, consistent with the notion of a limbic cerebellum and that cerebellar-limbic pathway allows the cerebellum to influence on emotion recognition (Schmahmann, 1991). Accumulating findings show cerebellar structural and functional alterations caused by a cerebellar pathology lead to behavioural issues consistent with a compromised ToM (Olivito et al., 2023b). Specifically, cerebrocerebellar patterns of connectivity such as the bilateral posterior cerebellar lobes with the

temporoparietal junction and the cerebellum with the precuneus, play a crucial role in automatic understanding, prediction and error correction of behavioural sequences (Van Overwalle et al., 2019). For instance, understanding a person's mental state or mentalizing and their personality traits strongly activates posterolateral cerebellar hemispheres, mainly lobules VI and VII (Crus I-II) (Andreasen and Piersons; 2008; Stoodley and Schmahmann, 2010; Helven et al., 2019). As shown in this review, total score and the cognitive component of the Faux Pas positively correlated with GM volumes in the right Crus II in SCA2 participants (Clausi et al., 2021a). Another specific region is the vermis, which is the principal target for limbic connections and is implicated in the modulation of primitive emotions (Stoodley and Schmahmann, 2010). More specifically, the salience network encompasses vermal and hemispheric parts of the lobule VI as well as the adjacent region of the Crus I and the dentate nuclei (Habas et al., 2009). Therefore, these cerebellar regions would be each one associated with a certain emotion, which are referred to as the "medial limbic" part of the cerebellum (Adamaszek et al., 2016). Buckner et al. (2011) showed that the cerebellum is involved in the default/mentalizing network, which connects cerebrum regions like the medial prefrontal cortex, the posterior cingulate cortex and the temporoparietal junction, which have an important role in mentalizing tasks (Van Overwalle et al., 2015). In particular, the main areas coupled with the default network are the Crus I-II and the lobule IX (Habas et al., 2013). For instance, the EA test "anger" score from SCA2 participants correlated with the GM volume in the right lobule VIIIb, the right lobule IX and the left Crus II, exemplifying the relevance of such regions in mentalizing. Other studies that evidence the involvement of the Crus I-II thanks to seed-based analysis, showing decreased FC between these cerebellar areas and the medial prefrontal cortex (Clausi et al., 2019). Another main areas are the middle and the superior cerebellar peduncle, which are responsible for the cerebellar-cortical connections and the feedback and feedforward systems (Olivito et al., 2023b). Therefore, Clausi and colleagues (2021a) suggested that the association between the cerebellar damage and the RME test results could be explained by the predictive role that the cerebellum has, where the WM degeneration of the peduncles would be interfering in the cerebello-cortial interactions that may be responsible for the patient's impairment. Accumulating evidence shows that a constatable relationship between the cerebellum and social cognition, although understanding the underlying mechanisms involved is still a challenge for the scientific community. Only four studies reviewed have carried out statistical analysis correlating neuroimaging findings with social cognition results, which does not allow to achieve a clear response to our research goal.

There were some limitations in this study that should be noted, being the main one the lack of studies covering this topic and, therefore, this review emphasises the scarcity of empirical research on the role of the cerebellum in social cognition using neuroimaging. Sample size was also an issue but, more specifically, the variety of ataxia subtypes interfered with the possibility of characterising each subtype in a cognitive level, not allowing the detection of specific deficits to each subtype. Another limitation was age variability and the degree of severity, which were not taken into account when obtaining results. Factors such as simple size, ataxia subtype and severity of condition could limit the generalizability of the findings and not represent the broader population of hereditary ataxia patients. In the same line, only one study evaluated longitudinally their sample, showing the progression that each ataxia subtype had over time. Moreover, although there is no standard protocol to evaluate social cognition in cerebellar disease patients there was a majority consensus on the choice of tests. Nonetheless, there is still a lack of tests that evaluate the different components of social cognition. Subsequently, insufficient studies carried out statistical correlations between social cognition findings and neuroimaging results, which was the main goal of this review. To direct future research, it is necessary to take into consideration all of the limitations mentioned previously. Mainly, research should focus on correlating social cognition and neuroimaging techniques, using a battery of standardised tests. Also, it would be important to study each ataxia subtype separately, which would allow to draw the different progressions of the disease course related to social cognition.

6. Conclusion

This review supports the role of the cerebellum in social cognition and its important role in the neuroanatomic pathway in emotional processing. However, the vast majority of the studies reviewed did not provide specific information regarding the correlation between social cognition outcomes and neuroimaging findings, most of them did find activation or irregularities in specific areas of the cerebellum, thus suggesting the involvement of the cerebellum in social cognition. Moreover, it is necessary to go further and understand how these deficits affect the day-to-day life of ataxia patients, and therefore, take them into consideration for therapeutic approaches.

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CRediT authorship contribution statement

Mercè Pallarès-Sastre: Writing – review & editing, Writing – original draft, Methodology, Conceptualization. **Maitane García:** Writing – review & editing, Writing – original draft, Methodology, Conceptualization. **Idoia Rouco-Axpe:** Writing – review & editing, Conceptualization. **Imanol Amayra:** Writing – original draft, Conceptualization.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

Data availability

No data was used for the research described in the article.

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