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ORIGINAL PAPER

The Working Life of People with Degenerative Cerebellar Ataxia

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Abstract

The aim of the present study was to characterize and analyze the most important individual and organizational variables associated with job accommodation in subjects with degenerative cerebellar ataxia by administering a series of international and validated work activity-related scales. Twenty-four workers (W) and 58 non-workers (NW) were recruited: 34 with autosomal dominant ataxia and 48 with autosomal recessive ataxia (27 with Friedreich ataxia and 21 with sporadic adult-onset ataxia of unknown etiology). The severity of ataxia was rated using the Scale for the Assessment and Rating of Ataxia. Our results showed that the ataxic W were predominantly middle-aged (41–50 years), high school graduate, and married men with a permanent work contract, who had been working for more than 7 years. The W with ataxia exhibited a good level of residual working capacity, irrespective of gender, age range, and duration of the disease, and they were observed to have a low or average-to-low job stress-related risk. Supporting patients with ataxia to find an appropriate job is an important priority because about 78% of NW search for a job and W and NW have the same potential work abilities (no relevant differences were found in terms of disease characteristics, gender, and work resilience). In this view, introducing NW to work-life may have a potential rehabilitative aspect. Findings of this study highlight that equal job opportunities for subjects affected by cerebellar ataxia are recommended.

Keywords Degenerative cerebellar ataxia · Working life · Work activity related scales

Introduction

Degenerative cerebellar ataxias are a heterogeneous group of disorders, affecting primarily the cerebellum and/or the cerebellar afferent pathways, with an estimated prevalence ranging from 5.2 to 18.5 per 100,000 inhabitants (1–3). A vast amount of hereditary forms of ataxia have been described so far including dominant, recessive and X-linked (4). The typical cerebellar motor syndrome includes a wide range of features such as dysmetria, asynergia or dyssynergia, a- or dysdiadochokinesia, tremor, oculomotor abnormalities, speech disturbances, hypotonia, and abnormalities of posture and gait (5). The cerebellum also contributes to the regulation of certain non-motor functions such as linguistic, cognitive (i.e., executive functions, processing speed, visuo- spatial functions, social skills), affective, and autonomic (5). All these symptoms have a great influence on the patient's independence in daily life activities (6) and working capacities.

Since in many patients the onset of disease symptoms falls within the working age, starting in the childhood-adolescent or youth periods (7–13), this may determine a series of problems linked to the work activity including the employability, work complications, or premature work activity interruption (14, 15). It should also be considered that the gradual decline of the motor ability expected over time (5) determines the interruption of the habitual routines and long-term plans and the engagement of demanding and persistent rehabilitation treatments (7).

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Taking into account these considerations, the knowledge of the specific work-related problems in these patients and, consequently, the attempts to improve their working life, should be an essential part of patients' health care in terms of psychological, social, and health wellness. Indeed, it has been demonstrated that searching for an appropriate job and avoiding the premature exit from it plays a beneficial key role in pathological subjects' overall quality of life (6, 16, 17). This can be attributable to the possibility to fully exploit the positive role of the vocational rehabilitation (18–26), by overcoming barriers to accessing, maintaining, or returning to employment (Vocational Rehabilitation Association of the UK). This possibility is also supported by the contextual ability of these subjects to maintain an effective motor strategy by adopting different compensatory behaviors during the course of the disease, in spite of disease progression and motor decline (27–30). Furthermore, designing an adequate job accommodation support allows an increase of self-esteem and of social wellness and a reduction of people's prejudice at workplace towards disabled people (31–33). Lastly, improving, among others, the social environment, support from coworkers and supervisors, job expectations, family- and disability-friendly policies, ergonomic interventions, and physical conditions (34) implies the reduction of experienced risk for stress-related disorders (35).

So far, no studies have focused on the work activity life in patients with cerebellar ataxia. The aim of the present study was to fully characterize and analyze the most important individual and organizational variables regarding the job accommodation in a relatively large sample of working and non-working subjects with degenerative cerebellar ataxia by administering a series of international and validated work activity-related scales.

Methods

Participants

This was a bi-centric, prospective, observational study involving two centers for degenerative ataxias in the University of Rome, Sapienza, and University of Naples, Federico II. Eighty-two out of 102 subjects with cerebellar ataxia consecutively evaluated were included in the study. Twenty subjects were excluded because they were not interested in participating in the study (6) or not able to fulfill (14) the questionnaires due to the cognitive deficit.

Thirty-four patients were diagnosed with autosomal dominant ataxia (spinocerebellar ataxia [SCA]), 27 with autosomal recessive ataxia (i.e., Friedreich ataxia [FRDA]) and 21 with sporadic adult-onset ataxia of unknown etiology (SAOA). All patients with SAOA showed slowly progressive cerebellar ataxia without clinical evidence of autonomic impairment. Twenty-four were workers (W) and fifty-eight were non-workers (NW) (see Table S1, supplementary online material).

All patients underwent a complete neurological assessment that included the following: cognitive evaluation, cranial nerve evaluation, muscle tone evaluation, muscle strength evaluation, joint coordination evaluation, sensory examination, tendon reflex elicitation, and disease severity. As for the cognitive assessment, a neuropsychological assessment was conducted by experienced neurologists (CS, MS, AR, AF). This included an in-depth clinical interview and an assessment of basic cognitive functions, such as motor and processing speed, spatio-temporal orientation, attention, comprehension, memory, and executive functions such as fluency and working memory. None of the assessed patients showed clear features of dementia. The severity of ataxia was rated using the Scale for the Assessment and Rating of Ataxia (SARA) (36).

A detailed description of the subjects' clinical characteristics is reported in Table S1 (supplementary online material).

All subjects gave informed consent prior to taking part in the study, which complied with the Helsinki Declaration and had local ethics committee approval.

Assessments

For the purposes of our research, we prepared two sets of surveys, one for W and one for NW. Both sets contained a socio-demographic, organizational (gender, age range, civil status, education, work contract, employment duration), and clinical diagnostic section (duration of disease, work injuries, other pathologies). Furthermore, both tests included the Resilience Scale for Adults (RSA) questionnaire (37). The surveys for W were composed of the Work Ability Index (WAI) (38) and the Health and Safety Executive (HSE) Management Standards Indicator Tool (39). Instead, the questionnaire for NW consisted of the Search for Work Self-Efficacy Scale (SWSES) (40). The RSA, WAI, HSE, and SWSES are thoroughly described in the supplementary online material.

Statistical Analysis

All the analyses were performed by using the IBM SPSS Statistics version 21 software (SPSS Inc. Chicago, IL, USA).

The Shapiro–Wilk and Kolmogorov–Smirnov tests were used to analyze the normal distribution of the data. Then, to highlight the presence of statistically significant differences between the mean values obtained for each tool (RSA, WAI, HSE, and SWSES), we performed the following tests: the parametric *t* test or the non-parametric Mann–Whitney test by splitting the sample by gender and duration of disease in years (less than 10 and 11–30) both for W and NW; the parametric *t* test or the non-parametric Mann–Whitney test by splitting the sample by age range in years (18–49, and) for

W; the parametric test ANOVA or the non-parametric Kruskal Wallis test, by subdividing the subjects by age range in years (18–40, 41–50, and 51, and beyond) for NW and post hoc analyses with Bonferroni's corrections, when significant differences were observed in the ANOVA or in the non-parametric Kruskal Wallis test. The aforementioned splitting of the data based on the duration of disease and age range was performed to obtain homogeneous subgroups in terms of the number of subjects and for higher statistical reliability.

When considering the RSA, we applied the parametric *t* test or the non-parametric Mann–Whitney test, by splitting the sample also by occupational status (W vs NW).

Furthermore, we investigated the effect of gender and occupational status, as well as their interaction, on the clinical scale (SARA) by using a two-way analysis of covariance (ANCOVA), considering age and duration of disease as covariates. Before the ANCOVA was run, all assumptions for this test were verified: the linear relationship between the dependent variable and covariates for each combination of the groups of the two independent variables, homogeneity of regression slopes, Levene's test of homogeneity of variance, and the Shapiro–Wilk test of normality.

Finally, the Spearman test was used to investigate any correlations among SARA and total and dimensions scores of each tool. Correlations among dimensions belonging to the same tool were not considered.

Values of $p < 0.05$ are the criteria for significance.

Results

Socio-demographic Data

The results of socio-demographic and working data in both W and NW are reported in Table 1, while Table 2 reports the clinical and diagnostic data for W and NW.

Table 1 Socio-demographic (gender, age range, civil status, and education) and working data (work contract and employment duration) for W and NW

	Workers ($n = 24$)	Non-workers ($n = 58$)
Gender		
Men	16 (66.7%)	25 (43.1%)
Women	8 (33.3%)	33 (56.9%)
Age range (years)		
18–40	8 (33.3%)	21 (36.2%)
41–50	16 (66.7%)	21 (36.3%)
51 and beyond	0 (0.0%)	14 (24.1%)
Missing	0 (0.0%)	2 (3.4%)
Mean ± SD	43.38 ± 6.90	46.28 ± 10.42
Civil status		
Unmarried	9 (37.5%)	18 (31.0%)
Married or cohabiting	12 (50.0%)	31 (53.5%)
Separated or divorced	3 (12.5%)	7 (12.1%)
Widower	0 (0.0%)	2 (3.4%)
Education		
Elementary school diploma	0 (0.0%)	3 (5.2%)
Junior high school	5 (20.8%)	26 (44.8%)
High school diploma	15 (62.5%)	25 (43.1%)
Bachelor's degree	1 (4.2%)	2 (3.5%)
Master's degree	3 (12.5%)	1 (1.7%)
Other	0 (0.0%)	1 (1.7%)
Work contract		
Permanent	21 (87.5%)	–
Fixed-term	1 (4.2%)	–
Collaboration/Fellowship	2 (8.3%)	–
Employment duration (years)		
Less than 6	1 (4.2%)	2 (3.4%)
7–15	8 (33.3%)	4 (6.9%)
16–25	6 (25.0%)	4 (6.9%)
26 and beyond	9 (37.5%)	2 (3.5%)
Missing	0 (0.0%)	46 (79.3%)

RSA

Scores of the RSA showed a moderate level (score range 78–121) of total resilience for both W (97.38 ± 10.63) and NW (96.36 ± 12.08). Tables 3 and 4 show the results of total RSA and the six dimensions of RSA for W and NW, respectively. No significant differences emerged for gender, age range, and duration of disease considering each dimension and the total resilience considering both W (Table 3) and NW (Table 4). Furthermore, the non-parametric Mann–Whitney test showed no significant differences between W and NW for total RSA ($p = 0.988$) and for each dimension (PS $p = 0.342$, PF $p = 0.198$, SC $p = 0.176$, SS $p = 0.650$, FC $p = 0.129$, SR $p = 0.257$).

WAI

In the 24 ataxic W, the WAI test showed an "excellent" level of residual working capacity for 16.7% of the subjects, "good" for 50.0%, "mediocre" for 25.0% and "poor" for 8.3%. The mean value (37.63 ± 5.66) obtained in W showed a "good" level of residual working capacity (37–43) (Table 3).

Table 3 shows the mean values of scores of total WAI and of the seven dimensions of WAI obtained considering the whole population of W and splitting the sample by gender, age range, and duration of disease.

Table 3 also shows the *p* values of the parametric or non-parametric tests. Particularly, no significant differences emerged for gender and duration of disease, when considering

Table 2 Health state data (year of diagnosis of ataxia, duration of disease, work injuries, and other pathologies) for W and NW

	Workers	Non-workers
Duration of disease (years)		
Less than 10	12 (50.0%)	7 (12.1%)
11–30	11 (45.8%)	44 (75.8%)
Missing	1 (4.2%)	7 (12.1%)
Mean ± SD	12.30 ± 6.90	16.25 ± 5.13
Work injuries		
Yes	18 (75.0%)	15 (25.9%)
	Falls (35%) Burns (30%) Crushing (15%) Others (20%)	
No	6 (25.0%)	0 (0.0%)
Missing	0 (0.0%)	43 (74.1%)
Other pathologies		
Cardiovascular	0 (0.0%)	3 (5.2%)
Genitourinary	1 (4.2%)	1 (1.7%)
Diseases of nervous system	0 (0.0%)	3 (5.2%)
Respiratory	2 (8.3%)	18 (31.0%)
Gastrointestinal	0 (0%)	1 (1.7%)
Other	3 (12.5%)	5 (8.6%)
Nothing	13 (54.2%)	16 (27.6%)
Missing	5 (20.8%)	12 (20.7%)

each dimension and the total WAI (Table 3), whereas a significant difference ($p = 0.011$) was observed with respect to age range only when considering the dimension 1 that regards the current work ability as opposed to lifetime best (Table 3). Particularly, the mean value in the group with age range 18–40 years (8.88) was significantly higher than that in the group with age range 41–50 years (7.38).

HSE

The average value for each dimension, the standard deviation, and its reference color generated by comparison with percentiles of reference (48) are described in Table 5.

Table 3 shows the scores of the seven dimensions of HSE obtained considering the whole population and splitting the sample by gender, age range, and duration of disease. Table 3 also shows the p values of the parametric or non-parametric tests. Particularly, a significant difference was found for gender only, when considering the control ($p = 0.026$) and managerial support ($p < 0.001$) dimensions: in the men's group, the mean values of both control (4.28) and managerial support (3.84) dimensions were significantly higher than in the women's group (3.56 and 3.60, respectively).

Furthermore, a significant difference emerged for age range for the relationships dimension ($p = 0.019$): mean value in the group of age range 41–50 years (4.32) was significantly higher than in the group of age range 18–40 (3.58).

Finally, significant differences were found, when considering the duration of disease for total HSE ($p = 0.007$), relationships ($p = 0.007$), and change ($p = 0.017$) dimensions: all the mean values obtained in the group of disease duration in the range of 11–30 years (4.35, 4.53, and 4.15, respectively) were significantly higher than the mean values of the group of disease duration less than 10 years (3.80, 3.75, and 3.53, respectively).

SWSES

The subjects who responded to the questionnaire were 77.6%, of which 46.7% were men and 53.3% were women, because not all subjects were planning or intended to seek a job.

The results show values that are placed in medium-high scores, which are higher than the perception that people have of their ability in seeking a job (see Table 4).

Table 4 shows the scores of the four dimensions of SWSES obtained when considering the whole population and splitting the sample by gender, age range, and duration of disease. Table 4 also shows the p values of the parametric or non-parametric tests. Particularly, no significant differences emerged for the duration of disease, when considering each dimension (see Table 4), while a significant difference was found for gender only, when considering the dimension of frustration coping ($p = 0.016$): the mean value of frustration coping dimension in the men's group (9.97) was significantly higher than the mean value in the women's group (8.79). Furthermore, a significant difference was found also for age range considering dimension regarding proactive career planning ($p = 0.021$). Particularly, post hoc analysis showed a significant difference between the group of age range 41–50 years and the group with 51 years and beyond: the mean value of the first (12.04) was significantly higher than the second (8.06).

SARA

There was no statistically significant interaction between gender and occupational status on SARA, when controlling for age and duration of disease ($F(1, 66) = 0.112$, $p = 0.739$, $p\eta^2 = 0.02$). There was no statistically significant main effect of the independent variable of gender ($F(1, 66) = 2.079$, $p = 0.154$, $p\eta^2 = 0.031$), but there was a statistically significant main effect of occupational status ($F(1, 66) = 7.041$, $p = 0.010$, $p\eta^2 = 0.096$), when controlling for age and duration of disease. In particular, in the W group, the mean value of SARA (12.99) was higher than the mean values reached in the NW groups (8.11), when controlling for age and duration of disease (Table 6).

Table 3 Scores of RSA, WAI, and HSE obtained for W, considering the whole population and splitting the sample by gender, age range, and duration of disease. *p* values regard the results of the non-parametric test of Mann–Whitney (a) and the parametric *t* test (b). Italicized value indicates statistical significance

		W	Gender			Age range (years)			Duration of disease (years)		
			Men ($n = 16$)	Women ($n = 8$)	*p* value	18–40 ($n = 8$)	41–50 ($n = 16$)	*p* value	Less than 10 ($n = 12$)	11–30 ($n = 11$)	*p* value
RSA total		97.38 ± 10.63	95.75 ± 11.90	100.62 ± 7.06	0.326[a]	97.13 ± 2.30	97.50 ± 13.07	0.913 [b]	100.33 ± 6.09	94.27 ± 14.06	0.188[b]
RSA dimensions	PS	2.87 ± 0.44	2.77 ± 0.37	3.06 ± 0.53	0.130[b]	2.87 ± 0.33	2.86 ± 0.50	0.958[b]	2.99 ± 0.47	2.74 ± 0.42	0.204[b]
	PF	3.18 ± 0.62	3.26 ± 0.67	3.00 ± 0.48	0.333[b]	3.16 ± 0.71	3.19 ± 0.59	0.910[b]	3.35 ± 0.57	3.00 ± 0.67	0.185[b]
	SC	2.72 ± 0.42	2.72 ± 0.42	2.71 ± 0.43	0.732[a]	2.58 ± 0.34	2.79 ± 0.44	0.281[b]	2.74 ± 0.36	2.68 ± 0.50	0.767[b]
	SS	2.96 ± 0.48	2.83 ± 0.43	3.22 ± 0.51	0.109[a]	3.16 ± 0.42	2.86 ± 0.49	0.159[b]	3.06 ± 0.35	2.86 ± 0.61	0.343[b]
	FC	3.38 ± 0.51	3.27 ± 0.58	3.58 ± 0.28	0.162[a]	3.38 ± 0.36	3.37 ± 0.58	0.999[b]	3.44 ± 0.40	3.30 ± 0.64	0.531[b]
	SR	2.73 ± 0.55	2.69 ± 0.55	2.80 ± 0.58	0.636[b]	2.70 ± 0.34	2.74 ± 0.64	0.856[b]	2.81 ± 0.48	2.63 ± 0.65	0.473[b]
WAI total		37.63 ± 5.66	37.09 ± 6.02	38.69 ± 5.08	0.634[b]	38.94 ± 7.18	36.97 ± 4.87	0.424[a]	35.67 ± 6.47	39.73 ± 4.27	0.093[b]
WAI dimensions	1	7.88 ± 1.68	7.50 ± 1.55	8.63 ± 1.77	0.105[a]	8.88 ± 2.10	7.38 ± 1.20	*0.011*[a]	7.58 ± 1.92	8.27 ± 1.42	0.363[a]
	2	8.00 ± 1.49	7.84 ± 1.55	8.31 ± 1.41	0.999[a]	7.94 ± 1.84	8.03 ± 1.35	0.888[b]	7.42 ± 1.70	8.64 ± 1.03	0.079[a]
	3	4.71 ± 1.60	4.68 ± 1.45	4.75 ± 1.98	0.700[a]	5.00 ± 1.51	4.56 ± 1.67	0.477[a]	4.83 ± 1.59	4.64 ± 1.75	0.743[a]
	4	4.46 ± 1.38	4.56 ± 1.15	4.25 ± 1.83	0.658[b]	4.38 ± 1.85	4.50 ± 1.15	0.645[a]	4.08 ± 1.38	4.82 ± 1.40	0.115[a]
	5	3.42 ± 0.88	3.50 ± 0.97	3.25 ± 0.71	0.948[a]	3.13 ± 0.99	3.56 ± 0.81	0.214[a]	3.17 ± 0.94	3.64 ± 0.81	0.222[a]
	6	5.88 ± 1.96	5.88 ± 1.86	5.88 ± 2.23	0.948[a]	6.25 ± 1.39	5.69 ± 2.18	0.645[a]	5.25 ± 2.38	6.46 ± 1.21	0.183[a]
	7	3.29 ± 0.62	3.13 ± 0.62	3.63 ± 0.52	0.063[a]	3.38 ± 0.74	3.25 ± 0.58	0.558[a]	3.33 ± 0.65	3.27 ± 0.65	0.810[a]
HSE total		4.05 ± 0.51	4.18 ± 0.34	3.79 ± 0.71	0.171[b]	3.95 ± 0.71	4.10 ± 0.40	0.521[b]	3.80 ± 0.55	4.35 ± 0.29	*0.007*[b]
HSE dimensions	1*	3.90 ± 0.60	4.00 ± 0.54	3.69 ± 0.69	0.281[b]	3.76 ± 0.73	3.96 ± 0.54	0.450[b]	3.67 ± 0.63	4.15 ± 0.51	0.057[b]
	2	4.04 ± 0.80	4.28 ± 0.50	3.56 ± 1.09	*0.026*[b]	3.98 ± 1.08	4.07 ± 0.66	0.794[b]	3.75 ± 1.00	4.36 ± 0.41	0.070[b]
	3	3.76 ± 0.74	3.84 ± 0.28	3.60 ± 1.26	*< 0.001*[b]	3.90 ± 0.97	3.69 ± 0.62	0.517[b]	3.55 ± 0.91	4.00 ± 0.47	0.513[a]
	4	4.09 ± 0.57	4.17 ± 0.51	3.94 ± 0.69	0.411[b]	4.19 ± 0.66	4.05 ± 0.53	0.580[b]	3.96 ± 0.61	4.25 ± 0.54	0.239[b]
	5*	4.07 ± 0.75	4.25 ± 0.68	3.73 ± 0.80	0.140[a]	3.58 ± 0.79	4.32 ± 0.61	*0.019*[b]	3.75 ± 0.80	4.53 ± 0.34	*0.007*[b]
	6	4.58 ± 0.55	4.58 ± 0.47	4.60 ± 0.73	0.306[a]	4.53 ± 0.74	4.61 ± 0.46	0.723[b]	4.45 ± 0.65	4.78 ± 0.37	0.175[a]
	7	3.81 ± 0.68	3.92 ± 0.59	3.58 ± 0.83	0.341[b]	4.00 ± 0.76	3.71 ± 0.64	0.333[b]	3.53 ± 0.72	4.15 ± 0.50	0.017[a]

RSA dimensions: *PS*, perception of self; *PF*, planned future; *SC*, social competence; *SS*, structured style; *FC*, family cohesion; *SR*, social resources

WAI dimensions: 1, current work ability as opposed to lifetime best; 2, work ability in relation to the demands of the job; 3, number of current diseases diagnosed by a physician; 4, estimated work impairment due to disease; 5, sick leave during the past year (12 months); 6, own prognosis of work ability 2 years from now; 7, mental resources

HSE dimensions: 1, demands; 2, control; 3, managerial support; 4, peer support; 5, relationships; 6, role; 7, change

*The questions for dimensions demands and relationships are negatively phrased, but in order to allow comparison across the other dimensions, the scores have been reversed so that higher value indicates less risk of stress at work, like for the other dimensions

Table 4 Scores of RSA and SWSES obtained for NW, considering the whole population and splitting the sample by gender, age range, and duration of disease. *p* values regard the results of the non-parametric test of Mann–Whitney (a), the parametric *t* test (b), the non-parametric Kruskal Wallis test, (c) and the parametric test ANOVA (d). Italicized value indicates statistical significance

		NW	Gender			Age range (years)				Duration of disease (years)		
			Men ($n = 25$)	Women ($n = 33$)	*p* value	18–40 ($n = 21$)	41–50 ($n = 21$)	51 and beyond ($n = 14$)	*p* value	Less than 10 ($n = 7$)	11–30 ($n = 44$)	*p* value
	Total RSA	96.36 ± 12.08	93.16 ± 15.09	98.79 ± 8.66	0.484[a]	97.04 ± 9.58	96.48 ± 9.53	93.43 ± 18.16	0.985[c]	95.00 ± 15.73	96.32 ± 11.86	0.763[b]
RSA dimensions	PS	2.93 ± 0.51	2.77 ± 0.55	3.05 ± 0.44	0.056[a]	3.10 ± 0.48	2.84 ± 0.30	2.78 ± 0.72	0.145[d]	2.98 ± 0.41	2.91 ± 0.55	0.689[b]
	PF	3.32 ± 0.76	3.26 ± 0.92	3.37 ± 0.59	0.585[a]	3.29 ± 0.78	3.28 ± 0.67	3.37 ± 0.86	0.932[d]	3.46 ± 0.98	3.24 ± 0.72	0.535[b]
	SC	2.55 ± 0.46	2.43 ± 0.39	2.64 ± 0.48	0.491[b]	2.53 ± 0.31	2.54 ± 0.37	2.56 ± 0.73	0.957[c]	2.69 ± 0.70	2.52 ± 0.40	0.637[b]
	SS	2.88 ± 0.70	2.60 ± 0.66	3.09 ± 0.66	0.695[b]	2.98 ± 0.67	2.71 ± 0.54	2.87 ± 0.92	0.480[d]	2.61 ± 0.54	2.90 ± 0.76	0.269[b]
	FC	3.16 ± 0.64	3.12 ± 0.65	3.20 ± 0.63	0.521[a]	3.23 ± 0.66	3.20 ± 0.52	2.98 ± 0.76	0.426[c]	2.83 ± 0.83	3.24 ± 0.55	0.181[b]
	SR	2.81 ± 0.53	2.83 ± 0.54	2.80 ± 0.53	0.585[a]	2.69 ± 0.53	3.00 ± 0.47	2.64 ± 0.56	0.061[c]	2.82 ± 0.69	2.82 ± 0.43	0.772[b]
SWSES dimensions	FrC	9.30 ± 2.24	9.97 ± 1.58	8.79 ± 2.53	*0.016*[a]	9.28 ± 2.65	11.12 ± 1.13	8.53 ± 2.53	0.057[c]	9.68 ± 1.02	9.47 ± 2.18	0.405[a]
	EE	10.49 ± 2.39	11.08 ± 2.11	10.04 ± 2.52	0.206[b]	10.24 ± 3.22	10.89 ± 1.45	10.32 ± 1.79	0.242[c]	10.70 ± 0.57	10.69 ± 2.44	0.708[a]
	PCP	9.56 ± 3.29	9.81 ± 2.46	9.36 ± 3.82	0.568[a]	9.53 ± 3.85	12.04 ± 2.42	8.06 ± 2.79	*0.021*[c]	9.56 ± 0.55	9.77 ± 3.49	0.804[a]
	RI	11.76 ± 2.22	12.16 ± 1.96	11.45 ± 2.38	0.110[a]	12.31 ± 2.53	8.76 ± 1.42	10.91 ± 2.59	0.163[c]	14.33 ± 0.47	11.97 ± 2.14	0.598[a]

RSA dimensions: *PS*, perception of self; *PF*, planned future; *SC*, social competence; *SS*, structured style; *FC*, family cohesion; *SR*, social resources

SWSES dimensions: *FrC*, frustration coping; *EE*, enterprising exploration; *PCP*, proactive career planning; *RI*, relational integration

Table 5 Dimensions of HSE: mean, SD, and color code for 24 workers

	Demands	Control	Managerial support	Peer support	Relationships	Role	Change
Mean	3.90	4.04	3.76	4.09	4.07	4.58	3.81
SD	0.60	0.80	0.74	0.57	0.75	0.55	0.68
Percentiles*							
20th	2.88	2.83	3.00	3.00	3.00	4.00	2.67
50th	3.50	3.50	3.80	3.75	4.00	4.40	3.33
80th	4.00	4.17	4.40	4.25	4.50	5.00	4.00
Risk	Average low	Average low	Average high	Average low	Average low	Average low	Average low

*Percentiles 20th, 50th, and 80th values (48)

Correlations

The correlation analysis for W showed the following:

- Perception of self (PS), an RSA dimension, was negatively correlated with dimension 6 (own prognosis of work ability 2 years from now) of WAI ($R = -0.662$, $p < 0.001$).
- Social competence (SC), an RSA dimension, was negatively correlated with dimension 6 (own prognosis of work ability 2 years from now) of WAI ($R = -0.441$, $p = 0.031$), control ($R = -0.567$, $p = 0.004$), managerial support ($R = -0.561$, $p = 0.004$), peer support ($R = -0.532$, $p = 0.007$), role ($R = -0.406$, $p = 0.049$), change ($R = -0.452$, $p = 0.027$) dimensions of HSE, and HSE total ($R = -0.535$, $p = 0.007$).
- Dimension 2 (work ability in relation to the demands of the job) of WAI was positively correlated with change dimension of HSE ($R = 0.479$, $p = 0.018$).
- SARA was positively correlated with peer support ($R = 0.412$, $p = 0.045$) and demands ($R = 0.522$, $p = 0.009$) dimensions of HSE, and HSE total ($R = 0.494$, $p = 0.014$);
- There were no correlations between total RSA, total WAI, total HSE, and SARA.

The correlation analysis for NW showed the following:

- Structured style (SS), an RSA dimension, was negatively correlated with frustration coping (FrC) ($R = -0.339$, $p = 0.009$) and proactive career planning (PCP) ($R = -0.335$, $p = 0.010$) (both SWSES dimensions).
- Social resources (SR), an RSA dimension, was negatively correlated with SARA ($R = -0.408$, $p = 0.002$).

Discussion

In our study, we have analyzed the most important features and problems related to the work activity in a sample of working and non-working subjects affected by degenerative cerebellar ataxia to possibly identify those aspects that are perceived as most critical. To our knowledge, this is the first study of patients with ataxia regarding the occupational area.

We found that the ataxic workers were predominantly middle-aged (41–50 years), high school graduate, and married men with a permanent work contract, who had been working for more than 7 years. These results suggest that, although degenerative ataxia, i.e., SCA and FA, are progressive diseases leading to an inexorable decline in the motor function (5, 27), patients with CA can fully face work-life. This evidence is further reinforced by the fact that the adaptability in the face of their serious health problems and workplace stressors is moderately present and that a high percentage of ataxic subjects displayed a good-to-excellent level of residual working capacity (Table 3), irrespective to the gender, age range, and duration of disease. This residual skill should be leveraged to avoid work interruptions. Among other possible interventions, specific environmental adjustments at the workplace, according to the principle of "reasonable job accommodations," could be sufficient to accommodate W's needs. On

Table 6 Mean (SD) of SARA unadjusted and adjusted for age and duration of disease. Two-way ANCOVA (VI: gender and occupational status; covariates: age and duration of disease).

SARA		Male	Female	Total	Sig.
Unadjusted M (SD)	W	11.95 (7.41)	13.33 (9.630)	12.43 (8.06)	
	NW	6.76 (4.07)	9.72 (5.79)	8.45 (5.29)	
	Total	8.92 (6.18)	10.52 (6.83)	9.72 (6.52)	
$M_{adj(age,\ duration)}$ (SE)	W	12.01 (1.64)	13.97 (2.47)	12.99* (1.50)	* 0.010
	NW	6.56 (1.42)	9.66 (1.20)	8.11* (0.94)	
	Total	9.28 (1.08)	11.81 (1.35)		

the other hand, these strategies should support work performance over an individual's working life. To do this, strategies such as targeted rehabilitation, corrective ergonomics, and training interventions should be scheduled regularly.

In addition, W with ataxia show a low or average low class of job stress-related risk as a total score (Table 5). Unexpectedly, we found a negative correlation between the disease severity and stress-related risk: the most severe the disease, the less the job stress-related risk. The interpretation of such a result may have a double-fold explanation. On one side, the most severely affected W with CA may be more cautious and thus adopt safety strategy to move in the workplace. On the other side, the risk perception may be reduced in these W due to cognitive decline (5).

Interestingly, we found an age-related difference regarding the current work ability as opposed to lifetime best, the older W perceiving the worsening of their current work abilities compared with the younger ones (Table 3). Conversely, younger W with shorter disease duration showed a higher risk of stress development due to work social relationships compared with older W (Table 3). Moreover, W with shorter disease duration also showed a higher risk of stress development due to changes in work activity (Table 3). These last findings suggest that younger W and W with shorter disease duration, thus those who are less adapted to the disease consequences, are more predisposed to the stress-related risk due to social relationships and changes at work.

Lastly, we found that men show less risk of stress development than women in the work management and managerial support (Table 3), in accordance with some studies that underlie the men-women different vulnerability to various occupational health outcomes (i.e., accidents or stress) (50).

In the present study, a high percentage of W (75%) reported work-related minor injuries, e.g., burns, falls, and wounds, possibly due to their poor motor coordination, leading to irregular and imprecise upper limb and hand movements as well as locomotion instability (51, 52). In this regard, it is a crucial point to identify the individual and environmental risk factors for these patients with the aim to find a solution in terms of risk prevention. For instance, since the balance disorder is one of the main problems in CA, it could be useful to adopt all the design and corrective ergonomic interventions (53), mainly on critical working tasks such as manual lifting and patient handling, pushing and pulling and repetitive activities, awkward and/or sustained postures, and prolonged sitting (54–60).

In addition, whereas the high percentage of W reported work-related injuries, only a low percentage (33%) of those asked turned to insurance assistance, which is obligatorily required by the Italian Workers' Compensation Authority (INAIL). The low rate of W patients requiring assurance may be due to the fear of losing their job or being discriminated by their managers or colleagues.

Although the W represent 29% of our sample, we should consider that a large quote of the NW could be included in the work-life as well. Indeed, NW do not differ from W in the working resilience scoring (moderate level in RSA, Tables 3 and 4) and perceive themselves as able in seeking a job based on the total score (medium-high scores in SWSES, Table 4).

Since a high percentage (about 78%) of NW search for a job, helping patients with ataxia in finding an appropriate job is an important priority if we consider that no relevant differences were found between W and NW in terms of disease characteristics, gender, and work resilience (Tables 3 and 4). Furthermore, no significant impact of the disease severity was present on the work resilience and seeking job adequacy. All these findings indicate that both W and NW have the same potential work abilities. Moreover, we found that the younger NW showed a higher degree of adequacy in the Proactive Career Planning dimension than the older NW (Table 4). This finding suggests that younger NW with CA are properly motivated in planning their future working career.

Furthermore, we found higher scores in women than in men in the dimension of feeling of frustration for selecting a job. This finding confirms the presence, highlighted in different studies, of the men-women differences in exposures and health effects in working life (50, 61).

Regarding NW, a job placement of these patients would allow an increase of self-esteem and of social wellness and a reduction of people's prejudice at the workplace towards disabled people (31–33). In this view, introducing NW to work-life may have a potential rehabilitative perspective. Finally, SARA values indicate that NW subjects can be considered employable for work due to the comparable clinical status with respect to W.

Findings of this study, also strived by the principles contained in the Convention on the Rights of Persons with Disabilities (United Nations 2008), highlight that equal job opportunities for subjects affected by CA are recommended.

Social policies for subjects with disabilities are worldwide supported by the World Report on Disability (WHO 2011, (62)), the principles stated within the "Convention on the Rights of Persons with Disabilities (United Nations 2008)" and guidelines from the International Labor Organization (ILO 2002). As reported by the Americans with Disabilities Act of 1990 (ADA), expanded with the adoption of the Americans with Disabilities Act Amendments Act of 2008 (ADAAA), "reasonable job accommodations," by reducing the obstacles faced by subjects with disabilities across many domains, represent the most effective way to mitigate the costly benefit dependence and social exclusion (63). The Job Accommodation Network (JAN), a service of the US Department of Labor's Office of Disability Employment Policy associated with the ADA, provides to employees with disabilities and employers useful guidance and documents with specific accommodation suggestions. Employees with

disabilities are supported to improve their employability, while employers to optimize the value that people with disabilities add to the workplace. Ataxia is considered in JAN. Within its website, you can find suggestions about flexible work schedules, liberal use of leave time and rest breaks, reduction of physical exertion and workplace stress, and implementation of an ergonomic workstation design. Specific solutions for computer use and standing and walking difficulties are given. In particular, the benefits of using key guards and typing aids, alternative input devices, adjustable workstations, wheelchairs, or scooters, in accommodating limitations when moving around a workplace, are highlighted. The European Disability Strategy (2010–2020) is aimed at unifying the approach for the inclusion of people with disabilities, in order to significantly raise the employment percentage (EC European Disability Strategy 2014). Moreover, the European Union Employment Directives issued in 1989 and 2000 provided guidelines addressed to people with disabilities for workplace inclusion, adaptation for accessibility (EC Directive 89/654/EEC, 2015, EC Employment Equality Framework Directive 2000/78/EC Council Directive 2000/78/EC, 2015) and for equal treatment by empowering employers towards a "reasonable accommodation" (64). In Italy, despite the employment of individuals with disabilities is regulated by Law No. 68 from 1999 ("Regulations for the right to work of persons with disabilities"), based on job-matching (65), only 16% of people with disabilities have a job (66, 67).

Furthermore, recent results show that in Italy, the workplace is not considered well- adapted for people with disabilities (63%), with a percentage higher than that found in Spain (62%), Belgium (61%), Slovenia (49%), Poland (47%), and UK (31%) (Moody 2016). Among the adaptations found at the workplace, the most common ones are physical adaptations to the buildings, whereas few adaptations are made to the way the jobs are carried out (changes to job tasks, role, pace, and working hours) (67). Only half of people with disabilities are well accommodated in terms of workplace design. Moreover, poor knowledge about adaptations for workers with neurological pathologies is shown (67). Some typical comments in Italy were that it is very difficult to find companies that are able to offer ergonomic furniture for people with disabilities (67). Fears about the cost to the employer were perceived surprisingly frequently (50%), despite Italian employers having stated an interest in training themselves and their organizations in creating ergonomic workplaces (86%) (67).

The INAIL has designed two 3-year research programs (2013–2015 and 2016–2018), focused on job accommodation of subjects affected by neurological pathologies, involving the cerebellum and the pyramidal and extrapyramidal system. The first research program was aimed at a full motor characterization of these subjects (27, 52, 68–76).

The INAIL, together with two reference centers for the study of ataxia, the Sapienza University of Rome and the Federico II University of Naples, shows the first results of the second research program regarding the characteristics of ataxic people. These include intrapersonal and interpersonal protective resources facilitating the tolerance to stress, work ability, sick leave, mental resources, demands referring workload and work patterns, control, managerial support, peer support, relationships, role, and other factors able to detect any problems perceived in terms of capacity in seeking employment.

Limitations of the Study

As the Italian version of RSA refers to healthy subjects and not those with neurological pathologies, the results should be interpreted with caution. Similarly, HSE, designed for the assessment of work-related stress risk within the organization in a sample of healthy subjects, was selected for this study as a tool for the evaluation of the subjective perception of ataxic workers within their work environment. Consequently, also in this case, results should be interpreted with caution (39).

History of employment for any group, details of the actual job and income of W patients, and disease-related or unrelated reasons why NW stopped working were not collected. These factors could be aspects of a future development of our study.

All the investigated patients were able to understand and complete the self-administered questionnaires, which implies preserved cognitive function in our patient sample. Furthermore, at the clinical interview, no clear features of dementia were revealed. However, we did not perform a test battery to thoroughly investigate the different cognitive domains including working memory, verbal and visuospatial long-term memory, attention, executive functions, language, and intelligence level. Thus, whether specific aspects of cognition may have influenced the questionnaire scores could not be inferred by the present study. Further studies are needed to investigate the interaction between cognitive function and work-life activity.

Conclusion

A high percentage of ataxic subjects, despite difficulties related to the symptoms of their disorder, display moderate levels of work ability. However, workplace accessibility remains a critical issue that must be addressed through targeted rehabilitation of ataxic subjects as well as ergonomic and training interventions. Such interventions are likely to be effective because ataxic workers positively perceive their current abilities, strengths, planning and control capacity, and ability to achieve their goals in the future. Moreover, they would allow ataxic workers to perceive any tension in relationships within their

organizations to a lesser extent. The aim of the INAIL, the Italian Workers' Compensation Authority, is to promote campaigns aimed (i) at improving the quality of work activity of patients with neurological pathologies, particularly of ataxic workers, and (ii) at helping non-workers find employment and go back to work. These aims will also be achieved by creating a dedicated job accommodation network, designed to provide neurological subjects and employers with the best suggestions for interventions based on the characteristics of their pathologies.

Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflict of interests.

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