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Psychological symptoms and Quality of Life in adults with Chiari malformation type I: an Assessment by the Italian version of Chiari Symptom Profile

*Loredana Benedetto [1], John J. Oró [2], Diane M. Mueller [2], Deborah Panarello [1], Erica Lo Turco [3], Gerardo Caruso [3], Antonino Francesco Germanò [3], Massimo Ingrassia [1]**

Abstract

Chiari malformation type I (CM-I) is a rare condition with physical and neurological manifestation changing across people. Being a chronic and debilitating disease, a comprehensive multidisciplinary approach is needed for evaluating patient's experienced Quality of Life (QoL) and psychological correlates of CM.

Objectives: The aim of this study was to develop the Italian adaptation of Chiari Symptom Profile (CSP), a questionnaire assessing the core symptoms of Chiari malformation and their impact on people's lives. Secondly, the occurrence of anxiety/depression symptoms and associations with patient-reported QoL were explored.

Methods: 172 adults with diagnosed CM-I (*N*=79 with neurosurgery) completed an online questionnaire measuring general QoL (WHOQOL-brief), disease-related QoL (CSP), and symptoms of anxiety/depression (HADS). Participant's demographic and clinical data were also collected.

Results: The Italian version of CSP showed excellent reliability both in total (Cronbach alpha = .97) and factorial scores (alphas from .87 to .95) assessing four domains of Chiari-related QoL (functional, physical, social, and psychological). For construct validity, significant correlations ($p < .001$) resulted among severity of CM symptoms, social and daily limitations assessed by CSP and general QoL. Participants' age at neurosurgery and condition (with/without neurosurgery treatment) did not significantly impact QoL scores, but perceived QoL worsened with increasing age. Among participants, 32% reported clinical anxiety and 14% depression symptoms (with higher incidence of depression in non-operated participants). Participants with clinical anxiety/depression reported a worse QoL in all domains of CSP (functional, physical, social, and psychological).

Conclusion: Findings suggest the need to include in CM treatment a continuous psychological support, identifying the patients most at risk who, in time, they may experience greater psychological suffering.

[1] Department of Clinical and Experimental Medicine, University of Messina, Messina, Italy

[2] Colorado Chiari Institute. Aurora, Colorado, USA

[3] Department of Biomedical and Dental Sciences and Morphofunctional Imaging, Unit of Neurosurgery, University of Messina, Messina, Italy

E-mail corresponding author: massimo.ingrassia@unime.it

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1. Introduction

Chiari malformation (CM) is a central nervous system malformation characterized by caudal displacement of the cerebellar tonsils exceeding 5mm below the foramen magnum with or without syringomyelia. CMs can present with diverse clinical manifestations, secondary to the involvement of the cerebellum, brain stem, spinal cord, lower cranial nerves, and altered CSF flow dynamics. Chiari malformation type I (CM-I), the most common, shows a caudal displacement (>5 mm) of the cerebellar tonsils through the foramen magnum into the cervical canal, and is without an associated meningomyelocele (Caffo et al., 2019). Chiari malformation type II (CM-II) is characterized by compression of the medulla and cerebellar tonsils into the upper cervical spinal canal and an associated meningomyelocele (Geerdink et al., 2012). Type III has the features of type II with an additional herniation of the entire cerebellum through the bony defect involving the foramen magnum, forming an encephalocele (Hadley, 2002). Type IV is characterized by cerebellar hypoplasia or aplasia and occipital encephalocele (Haddad et al., 2018).

Indications for surgery include the presence of neurological symptoms, their progression, and/or headache caused by herniation of the cerebellar tonsils and significantly deteriorating the patients' quality of life. However, even under surgical treatment, some patients may have little or no benefit (Almotairi et al., 2009; Durham et al., 1998, Landridge et al., 2017). Chronic pain is the most frequent symptom reported (Aghakhani et al., 2009; Garcia et al., 2021); other common complaints are fatigue, vertigo, headache, neck pain, dizziness or weakness in extremities (Cohodarevic et al., 2000; Mueller & Oró, 2004). Other comorbidities impacting patient's functioning are cognitive and memory deficits, aphasia, and sleep disturbance (Almotairi et al., 2020; Ferré Masó et al., 2014; Garcia et al., 2018), with consequences on daily activities (Meeker et al., 2015).

Being a chronic and debilitating disease, CM-I brings with it not only pain and physical symptoms, but also psychological consequences and reduced quality of life (QoL). From a psychological point of view, fear is the most common emotion experienced by people after the diagnosis (Fischbein et al., 2015; Garcia et al., 2019), and subsequent adaptation to illness is linked to clinical variability of symptoms, more favourable diagnosis, patient's coping strategies and general psychological functioning (Burke et al., 2015; Garcia et al., 2019; Lazzari et al., 2019). Conversely, studies document a high incidence of problems such as depression (Chen et al., 2009), affective pain, stress, and anxiety (Garcia et al., 2019) in patients with CM. Particularly, psychiatric conditions such as anxiety and depression increase patient's discomfort and are associated with poorer QoL (Garcia et al., 2021; Mestres et al., 2012). Following the well-known

definition by the World Health Organization (WHOQOL Group, 1998), the Quality of Life describes the individual's perception of overall well-being, both mental and social, not only the absence of the disease. In this meaning QoL is a multidimensional construct that poses the emphasis on what the individual considers important to his/her life, that is, emotional needs, relationships, self-determination, expectancies and values, together with the physical health conditions. The Chiari syndrome involves clinical manifestations and functional deficits that can impact all the dimensions of person's QoL (physical and psychological health, social relationships, and environment). The presentation of physical symptoms is very heterogeneous, and often the severity of consequences self-reported by patients does not correspond to objective neurological conditions (namely, patients with minimal herniation can accuse severe symptoms; Bezuidenhout et al., 2018; Hofkes et al., 2007). In addition, symptoms can improve to various degrees or remain unchanged after surgical intervention (Greenberg et al., 2015), differentially impacting subjective QoL. Patients with CM-I report lower QoL than healthy individuals and perceived that QoL ameliorates in most of cases after surgical decompression (Almotairi et al., 2020), regardless of the patient's age and the presence of syringomyelia (Mueller & Oró, 2005). However, due to the complex brain abnormalities of CM and associated comorbidities, patients may report no improvement in some self-evaluated symptoms or even worsening years later neurosurgical intervention (Garcia et al., 2021). For example, as Martínez-Sabater et al. (2018) refer, symptoms linked to brainstem or cerebellum compression tend to ameliorate more than those attributable to syringomyelia (i.e., scoliosis or loss of sensitivity). Beyond the objective clinical evaluation, the patient can perceive no benefit after surgery, continuing to experience emotional distress and difficulties in daily activities. Therefore, QoL is a crucial conceptual perspective for comprehensively evaluating health problems and their relevance starting from the patient's experience, needs, and desire to achieve better well-being. Secondly, researchers and practitioners agree that assessing the QoL is vital for planning interventions (both medical and psychosocial) and for evaluating their effectiveness in improving an individual's QoL. There are two approaches assessing the health-related QoL, the generic QoL scales or disease-specific scales (Patrick & Deyo, 1989; Lin et al., 2013). The generic measures provide a subjective evaluation in all the components of health – physical, emotional, environmental, and social – and have the advantage to compare QoL between patients and healthy individuals, or across different diseases, treatments, or interventions. Conversely, diseases specific measures examine symptoms and conditions typical of the respective disease. Therefore, a disease-specific approach is more responsive to patient-reported functional well-being, and derived questionnaires are more sensitive than generic questionnaires to measure changes in QoL following treatments and interventions.

The Chiari Symptom Profile (CSP) is a questionnaire developed by Mueller and Oró (2013) to assess the presence and severity of the wide range of symptoms of CM/syringomyelia and how they affect an individual's daily functioning. The 57 items use an ordinary and straightforward language, without medical terms (i.e., I have neck pain, ringing in my ears, etc.), to describe problems the person can experience, and their impact on physical, functional, emotional, and social well-being. Following a disease-specific approach, the CSP provides a measure of QoL, together with a patient-evaluated measure of the degree of perceived disability. The questionnaire is a reliable and valid measure of different aspects of QoL in adults affected by CM/syringomyelia, and it is also sensitive to evaluating changes in QoL after neurosurgery intervention (Mueller & Oró, 2013). For these features, the CSP is a practical instrument that can be used to determine the person's health needs, following a comprehensive approach beyond the diagnosis of physical problems. Not the least being a self-administered questionnaire, the CSP does not require much time to be filled in by the patient and can be proposed by many physicians and professionals within the multidisciplinary teams, not exclusively by dedicated psychologists.

The main scope of this study was to contribute to the development of the Italian version of the CSP for measuring the domains of the QoL (physical, functional, psychological, and social) in adults with CM-I. All the domains of QoL, together with perceived disability degree, were compared between individuals with/without neurosurgical intervention. Secondly, since the literature exploring the risk for psychological disorders in people with CM is scarce (*see*, for example, Bakim et al., 2013; Fischbein et al., 2015; García et al., 2019), the study assessed the prevalence of anxiety/depression symptoms and the association between anxiety or depression and patient's surgical condition (with/without neurosurgical intervention).

2. Method

2.1 Participants and procedure

Participants were recruited with the collaboration of AISMAC (*Associazione Italiana Siringomielia e Arnold Chiari*), an Italian association of families and individuals affected by CM and syringomyelia. The inclusion criteria were a diagnosed CM and age over 18 years old. An invitation to participate in the study, with a brief presentation of its aim and procedure, was published on AISMAC's website. Participants were volunteers who, after declaring their interest in collaborating on the research, received a link with the informed consent and the questionnaires to fill online. One hundred seventy-four people replied, but two were excluded because they were under the age of 18 years. The characteristics of the remaining 172 participants are reported in Table 1. The final sample comprised mainly females (83%) with

CM-I (92%). About half of the patients (n = 79) underwent decompression surgery, and two were waiting for surgery. Ages ranged between 18-75 y.o. for males (*M* = 41.4, *SD* = 16.01) and 18-78 y.o. for females (*M* = 42.2, *SD* = 13.9). Regarding age distribution, the groups were young adults (18-35 years, *n* =59, 34%), middle-aged adults (36-55 years, *n* =84, 49%), and adults (more than 55 years, *n* =29, 17%).

Table 1. Demographic and clinical characteristics of participants

		M (*DS*)	Number of cases (%)
Mean age, years (*SD*)		42.1 (14.2)	
Mean age at diagnosis, years (*SD*)		32.6 (13.6)	
Mean age at surgery, years (*SD*)		33.6 (14.4)	
Gender	Male		29 (17%)
	Famale		143 (83%)
Marital status	Unmarried		69 (40.1%)
	Married/ Cohabitant		88 (51.1%)
	Separate/ divorced		14 (8.2%)
	Widower		1 (0 .6%)
Educational status	1st grade Secondary School		28 (16.3%)
	2nd grade Secondary School		94 (54.7%)
	Graduation		40 (23.3%)
	Post Graduate Specialization		10 (5.7%)
Chiari malformation type	Type I		158 (92%)
	Type II		11 (6%)
	Type III		1 (0 .6%)
	Diagnosis in course		2 (1%)
Decompression surgery	Yes		79 (46%)
	No		91 (53%)
	Waiting for intervention		2 (1%)
Neurological condition	Syringomyelia		79 (46%)
	Hydrocephalus		26 (15%)
Medical comorbidities	Scoliosis		49 (28,5%)
	Tinnitus		50 (29.1%)
Other diagnosed disorders	Vision		64 (37%)
	Language		20 (11.6%)
	Learning disabilities		20 (11.6%)

2.2 Measures

WHOQOL-brief The Italian version of the WHOQOL-brief (De Girolamo et al., 2011) was used for the dual purpose of evaluating general QoL (WHOQOL Group,1998) and having a measure to assess the construct validity of the Italian translation of the CSP. The WHOQOL-brief is a self-rating questionnaire (26 item) giving an overall score of QoL and four domains: physical health (e.g., fatigue, sleep quality, need for medication, etc.), psychological health (e.g., positive and negative feelings, personal beliefs, self-esteem), social relationships (e.g., participation, social support, and sexual activity), and environment (e.g., security at home, transportation, financial resources, and healthcare assistance). Items ask respondents to rate their QoL during the last 15 days with a 5-point Likert scale. In this study, the internal consistency of measures resulted good (Cronbach's alphas from .62 for social relationships to .87 for physical health), similarly to the Italian and the original WHOQOL-brief questionnaire. The questionnaire does not provide a cut-off point for better or worse QoL, but scores have a positive direction, so the higher the score, the better the perceived quality of life.

Chiari Symptom Profile (CSP, Mueller & Oró, 2013) is a self-report questionnaire measuring symptoms of CM and their impact on the people's lives through a quantitative analysis. The questionnaire is composed of 57 items describing core symptoms of CM and syringomyelia. However, there are no medical terms, so the simple language makes it easy for patients to recognize the described conditions and difficulties. Items refer to four dimensions: physical (pain, numbness, for example "I have headache"), functional (daily activities, "I need help to bathe and dress"), psychological (anxiety, negative feelings, "I feel generally tired or fatigued"), and social (communication and social relationships, "I have difficulty speaking clearly"). Respondents rate symptoms' presence and severity according to a Likert-type scale (0=never, 1=rarely, 2=some of the time, 3=most of the time, and 4=all of the time). Higher overall scores indicate worse QoL/symptom outcomes. A score between 0-60 corresponds to *no disability,* 61-115 is *a mild disability*, 116-173 is *a moderate disability,* and 174-228 indicates *severe disability* (Mueller & Oró, 2013). There are two additional items. The first item asks the patient to evaluate to what extent Chiari symptoms impact their life (*Perceived disability*: from 0= "I fill like I am not disabled - I am able to function independently and do the activities I enjoy" to 3= "I feel like I am totally disabled, I am not able to function independently, and need help with all activities"). Secondly, the patient rates their overall quality of life by choosing a score from 1 (worse) to 10 (best). Higher scores correspond to better QoL.

Translation process. After the authors' permission, two psychology researchers independently translated the English questionnaire into Italian. The two translations were then compared with

the help of physicians in order to discuss minor conceptual disagreement in some items and obtain a shared version. This preliminary version was revised with a back-translation procedure by a native-speaker teacher for the final version of the questionnaire that was again submitted to the doctors' opinion. The items of the Italian CSP questionnaire (CSP-Ita) are listed in Table 2.

Table 2. Results of the Principal-components analysis (Varimax rotation) for the Italian Chiari Symptoms Profile questionnaire (CSP-Ita)

	Factors			
	Functional	Physical	Social	Psychological
1. I have headaches [*Ho mal di testa*]		.71		
2. I have neck pain [*Ho dolore al collo*]		.69		
3. I have arm pain [*Ho dolore al braccio*]		.54		
4. I have back pain [*Ho mal di schiena*]		.53		
5. I have dizziness or feel faint [*Ho le vertigini o mi sento svenire*]		.47		
6. I have ringing in my ears [*Ho ronzii alle orecchie*]			.63	
7. I have trouble swallowing [*Ho difficoltà a deglutire*]			.46	
8. I have trouble reading due to blurred vision [*Ho difficoltà a leggere a causa della visione offuscata*]		.54		
9. I have trouble with my balance while walking [*Perdo l'equilibrio mentre cammino*]	.60			
10. My symptoms prevent me from participating in activities I enjoy [*I miei sintomi mi impediscono di partecipare alle attività che mi piacciono*]	57			
11. My symptoms prevent me from exercising regularly [*I miei sintomi mi impediscono di allenarmi regolarmente*]	.49			
12. I need help to bathe and dress [*Ho bisogno di aiuto per lavarmi e vestirmi*]	.71			
13. I need someone else/cane/walker to help me walk [*Ho bisogno di qualcuno/un bastone/un accompagnatore che mi aiuti a camminare*]	.82			
14. I feel sad or depressed [*Mi sento triste o depresso*]				.76
15. I feel angry [*Mi sento arrabbiato*]				.76
16. I need help to take care of my family [*Ho bisogno di aiuto per prendermi cura della mia famiglia*]	.58			
17. I have difficulty concentrating, thinking and problem solving [*Ho difficoltà a concentrami, a pensare e a risolvere problemi*]			.60	
18. I have difficulty reading and understanding letters/books or newspapers [*Ho difficoltà a leggere e comprendere lettere/libri e giornali*]			.48	
19. I have difficulty speaking clearly [*Ho difficoltà a parlare chiaramente*]			.72	
20. I cannot walk more than 10 minutes without stopping to rest [*Non riesco a camminare più di 10 minuti senza fermarmi a riposare*]	.74			
21. I need to lie down during the day to rest [*Ho bisogno di sdraiarmi durante il giorno per riposare*]				.54
22. I am working shorter or limited amount of hours due to my symptoms [*Sto lavorando un numero di ore minore o limitato a causa dei miei sintomi*]	.53			
23. I have difficulty sleeping at night [*Ho difficoltà a dormire la notte*]				.52
24. I feel my heart racing or have palpitations [*Sento il mio cuore battere forte oppure ho le palpitazioni*]				.50
25. I feel generally tired or fatigued [*In generale mi sento stanco o affaticato*]				.55
26. I need assistance with shopping [*Ho bisogno di assistenza per fare shopping*]	.76			

27. I need someone else to drive me to the store/appointments [*Ho bisogno di qualcuno che mi porti nei negozi/agli appuntamenti*]	.77			
28. I feel irritable [*Mi sento irritabile*]				.76
29. I need help to do housework (laundry, vacuuming, and dusting) [*Ho bisogno di aiuto per svolgere i lavori di casa (fare la lavatrice, passare l'aspirapolvere, spolverare)*]	.60			
30. I have difficulty holding objects in my hands [*Ho difficoltà a tenere oggetti nelle mani*]	53			
31. I feel short of breath or have difficulty breathing [*Sento il fiato corto oppure ho difficoltà a respirare*]	.49			
32. I am confused and forget what I am doing [*Sono confuso e dimentico ciò che sto facendo*]			.54	
33. I have trouble finding the right words to communicate my needs [*Ho difficoltà a trovare le parole giuste per comunicare i miei bisogni*]			.66	
34. I lose control of my bowel/bladder [*Perdo il controllo del mio intestino o della mia vescica*]	.61			
35. I have trouble with fine motor tasks such as buttoning buttons [*Ho difficoltà con le abilità fini-motorie come abbottonare i bottoni*]	.58			
36. Food does not taste normal [*Il cibo non ha un sapore normale*]	.42			
37. I choke when I try to swallow liquids [*Soffoco quando provo a ingoiare i liquidi*]	.51			
38. I have been told that I snore loudly at night [*Mi è stato detto che russo rumorosamente durante la notte*]			.44	
39. I have been told that I stop breathing at night or while lying flat [*Mi è stato detto che smetto di respirare di notte o quando sono disteso*]			.52	
40. I feel nauseated or sick to my stomach [*Mi sento nauseato o malato di stomaco*]		.65		
41. I feel like the room is spinning around me [*Sento come se la stanza giri intorno a me*]		.49		
42. My face feels numb [*La mia faccia sembra addormentata*]		.64		
43. I have double vision [*Ho una visione doppia*]	.48			
44. Bright lights hurt my eyes [*Le luci intense mi fanno male agli occhi*]		.56		
45. I *have* head pain when I cough/sneeze or strain [*Ho mal di testa quando tossisco/starnutisco o mi sforzo*]			.54	
46. I have head *pain* when I bend forward or lean over [*Ho mal di testa quando mi piego in avanti o mi chino*]			.54	
47. I have head pain when I look up at the sky or the top shelf [*Ho mal di testa quando guardo in alto il cielo oppure i ripiani più alti*]		48		
48. My tongue is numb or tingly [*La mia lingua è intorpidita o sento formicolii*]		.50		
49. I get a headache when I stand up after lying down [*Mi viene mal di testa quando mi alzo dopo essermi sdraiato*]		.60		
50. I have hiccups [*Ho il singhiozzo*]		.52		
51. My eyes twitch or jump [*I miei occhi si chiudono o si muovono (involontariamente)*]				
52. My arm is numb [*Il mio braccio è intorpidito/addormentato*]		.47		
53. I have heartburn or indigestion [*Ho bruciore di stomaco o indigestione]*		.61		
54. I need to take pain medication to get through the day [*Ho bisogno di prendere antidolorifici per superare la giornata*]		.52		
55. I have generalized body pain (all over my body) [*Ho un dolore al corpo generalizzato (in tutto il corpo)*]		.47		
56. I have constipation (difficulty or pain having a bowel movement)	.43			

[*Sono costipato (ho difficoltà o dolore dovuto al movimento intestinale)*]				
57. Overall, I feel unhappy and/or frustrated about my health [*In generale, mi sento infelice e/o frustrato per la mia salute*]				.71
Eigenvalue	20.94	3.48	2.90	2.09
Percentages of variance	16.34	14.39	10.82	10.04
Standardized Cronbach Alpha	.95	.92	.87	.88

Note. The number of items corresponds to the original English questionnaire. The Italian items are in parentheses.

Hospital Anxiety and Depression Scale (HADS; Zigmond & Snaith,1983; It. ad. Costantini et al., 1999) is a screening questionnaire for psychiatric disorders both in clinical and community populations. The questionnaire is composed of 14 items and two scales assessing anxiety (i.e., "Worrying thoughts go through my mind", the response from 0= *Only occasionally* to 3= *A great deal of the time*) and depression symptoms ("I feel as if I am slowed down", response from 0= *Not at all* to 3= *Nearly all the time*). The questionnaire has a high internal consistency (in this sample Cronbach's alphas .81 and .83 for anxiety and depression, respectively). In this study, we assumed the ten score as the cut-off for the presence of anxiety or depression (Costantini et al., 1999).

Ethics. The study was planned following the ethical standards of the 1964 Declaration of Helsinki, the Ethical Code for Italian psychologists (L. 18.02.1989, n. 56), and Italian law for data privacy (DLGS 196/2003). Approval by the Ethical Committee of the Psychological Research and Intervention Center (CeRIP) of the University of Messina was obtained (nr. prot. 125435, 2020).

2.3 Statistical analysis

Data were processed with IBM SPSS Statistics for Windows 19.0. The first step of data analysis was validating the questionnaire in the Italian version (CSP-Ita). An exploratory factor analysis (EFA), with the principal-components method, was conducted with the scope to derive the dimensionality of the CSP-Ita questionnaire. The criteria for the extraction of factors (varimax-rotated solution) were: a) eigenvalues ≥ 1 (Kaiser's criterion), b) scree test, c) coherence and interpretability of factors. The internal consistency of extracted factors (both total and subscales scores) was tested with the Cronbach's alpha. Inter-correlations among CSP-Ita measures (Total and subscale scores), Perceived disability scores, and overall QoL scores - were checked by Pearson's coefficients. Secondly, the construct validity was tested correlating (Pearson's coefficients) CSP-Ita and WHOQOL-brief scores. Significant negative associations (p <.05) between the severity of CM outcomes (CSP-Ita) and the general QoL (WHOQOL-brief) were assumed, since the two scales provide measure with opposite directions (divergent validity).

Pearson's correlations also tested the associations between CSP-Ita measures (total score and subscales) and participants' age/age at neurosurgery.

Third, descriptive statistics (*M* and *DS*) were calculated for CSP-Ita measures, and differences between patients not operated/operated on were tested by ANOVA (overall score) or MANOVA (subscales). Based on CSP-Ita total scores, participants were classified into four levels of disability (from *no disability* to *severe disability*). The number (and percentages) of people with different disability grades were calculated, and a chi-square test checked the independence of distributions between not operated/operated participants. Finally, the number (and percentages) of participants who achieved the cut-off score (HADS) for psychiatric disorders (anxiety or depression) was calculated. The independence of the distributions (χ^2) was checked according to a contingency table 2 (patients with/without neurosurgery) x 2 (presence/absence of psychiatric disorders) for anxiety and depression, separately. Differences (*F* test by MANOVA or ANOVA) for CSP-Ita measures (both subscales and total score) as a function of neurosurgery (with/without) and psychiatric disorders (presence/absence for anxiety and depression) were estimated. Lastly, Pearson's correlations were tested between anxiety/depression levels and participants' age, age at CM diagnosis, age at neurosurgery, and severity of CM outcomes (CSP Total).

3. Results

3.1 CSP-Ita reliability and validity

Preliminarily, the sampling adequacy was checked. The Kaiser-Meyer-Olkin's measure resulted in excellent (KMO = .91), and the Bartlett's test of sphericity significant, $\chi^2(1596) = 6919.32$, $p = .001$. Therefore, the EFA on the scores of 57 items of the CSP-Ita was calculated. Four factors explaining 51.59% of the overall variance emerged. Based on the items with the highest saturations (minimum factor loading of .40), the EFA identified four factorial dimensions that correspond to the constructs by Mueller and Oró (2013) in the original English questionnaire (Table 2). All items were included, with the only exception of the item 51 which does not reach saturations above .40 in any of the factors. The internal reliabilities of factors (Cronbach's alphas) ranged from very good for the Social dimension (.87) to excellent for the Functional dimension (.95). The reliability of the total CSP-Ita scale (all 57 items) resulted in excellent (.97) similarly to the English questionnaire (.96). Regarding the validity check, strong inter-correlations ($p<.001$) resulted among the factorial dimensions of CSP-Ita (from .54 to .71), and between factorial dimensions and CSP-Ita full scale (from .75 to .93; Table 3). In addition, as hypothesized, highly significant associations ($p<.001$) with a negative direction were found between the CSP-Ita scores and the general QoL (WHOQOL-brief) scores.

Table 3. Internal consistency and construct validity for the CSP-Ita questionnaire (Pearson's coefficients). Means (and Standard Deviations) for scores of health-related (CSP) and general (WHOQOL) quality of life.

Measure	Chiari Symptom Profile (CSP)					General QoL (WHOQOL)				
	1.	2.	3.	4.	5.	6.	7.	8.	9.	10.
1. CSP - Physical	1									
2. CSP - Social	.71**	1								
3. CSP - Functional	.70**	.70**	1							
4. CSP- Psychological	.60**	.66**	.54**	1						
5. CSP – Total [1]	.88**	.93**	.85**	.75**	1					
6. WHOQOL – Physical Health	-.63**	-.76**	-.62**	-.73**	-.78**	1				
7. WHOQOL – Phycological Health	-.36**	-.48**	-.47**	-.69**	-.53**	.63**	1			
8. WHOQOL – Social relationships	-.23**	-.32**	-.33**	-.49**	-.36**	.43**	.59**	1		
9. WHOQOL – Environment	-.39**	-.49**	-.47**	-.62**	-.54**	.60**	.66**	.55**	1	
10. WHOQOL – General	-.61**	-.62**	-.48**	-.58**	-.67**	.68**	.52**	.34**	.43**	1
M (*SD*)	1.73 (.67)	1.24 (.87)	1.50 (.80)	2.11 (.78)	89.29 (40.39)	21.69 (5.51)	18.76 (4.19)	9.74 (2.61)	24.27 (4.93)	3.09 (.95)

Note. [1] CSP Total score is calculated on all 57 items

** Correlation is significant at the .001 level (two- tailed)

The participants' age resulted correlated negatively with overall QoL (r = -.18, p <.05), and positively with CSP total score (r = .23, p <.01) and perceived disability (r = .24, p <.01). A significant positive association resulted between CSP total score and perceived disability (r = .69, p <.001).

3.2 Descriptive statistics

Statistics (*M* and *DS*) in CSP-Ita total scores and perceived disability scores were calculated as a function of neurosurgery condition (not operated/operated). No significant differences (p > .05) emerged in CSP-Ita total score between not operated (M = 90., SD = 38.03) and operated participants (M = 87.70, SD = 43.19). Similarly, no significant differences emerged (p > .05) in the Perceived disability scores: not operated (M = 6.38, SD = 1.72), operated (M = 6.58, SD = 1.98).

The prevalence of disability levels in the sample according to CSP scores is reported in Table 4. There is no significant association between disability grade and neurosurgery intervention, χ^2 (3, N = 172) = 3.81, p > .05.

Table 4. Occurrence of levels of disability according to the CSP-Ita questionnaire measure (Total score)

Levels of Disability	Not operated (N = 93)	Operated (N = 79)	Total (N = 172)
No disability	22 (12.8%)	25 (14.5%)	47 (27.3 %)
Mild disability	46 (26.7%)	34 (19.8%)	80 (46.5%)
Moderate disability	24 (14 %)	17 (9.9%)	41 (23.8%)
Severe disability	1 (0.6%)	2 (1.7%)	4 (2.3%)

3.3 Psychiatric symptoms and QoL

Participants who reported elevated anxiety (according HADS score) resulted 55/172 (32%). Among them, 22/55 (40%) underwent the decompression intervention, and 33/55 did not (60%). Chi-square test of independence showed that there was no significant association between anxiety and neurosurgery intervention, χ^2 (1, N = 172) = 1.15, p > .05. Regarding depression, participants who reached the clinical threshold (HADS score) resulted in 25/172 (14%), and 18/25 (72%) did not undergo neurosurgery, whereas 7/25 (28%) were operated. The relationship between the two variables is significant, and depressed subjects are likelier to have not been operated, χ^2 (1, N = 172) = 3.79, p = .05.

Statistics (M and DS) for severity of CM outcomes (CSP subscales) as a function of neurosurgery condition (not operated/operated) and anxiety levels (normal/clinical range) are presented in Table 5. By the MANOVA significant differences (p < .001) emerged for "anxiety level" factor, and participants with clinical anxiety reported higher scores in all dimensions of the CSP questionnaire, indicating worse QoL: Physical F(1, 168) = 24.88, η_p^2 = .13; Social F(1, 168) = 18,22, η_p^2 = .10; Functional F(1, 168)= 11.99, η_p^2 =.07; Psychological F(1, 168)= 56.26, η_p^2 = .25. For "neurosurgery condition" factor significant differences emerged in Social subscale [F(1, 168)= 4.37, η_p^2 = .03, p= .04] with lower scores in operated (M= 1.65, SD= .68) than not operated participants (M = 1.81, SD = .66). Finally, a significant interaction "anxiety level" x "neurosurgery condition" resulted in Psychological dimension [F(1, 168)= 4.19, η_p^2 = .02, p = .04]. With a normal anxiety level, the scores of not operated participants (M = 1.95) were higher than operated participants ones (M = 1.75); conversely, with a clinical anxiety level the scores of operated participants (M = 2.82) resulted higher than not-operated (M = 2.56), but in both cases the mean differences were not significant (ps > .05).

Table 5. Mean (and Standard Deviations) scores for the Chiari Symptom Profile (CSP) measures as a function of Neurosurgery condition and Anxiety/Depression levels

Neurosurgery condition	Symptoms levels	Anxiety		Depression	
		M	*SD*	*M*	*SD*
CSP Physical					
Not operated	Normal	1.65	.67	1.68	.62
	Clinical range	2.10	.52	2.36	.51
Operated	Normal	1.49	.61	1.59	.67
	Clinical range	2.07	.67	2.27	.44
CSP Social					
Not operated	Normal	1.04	.78	.98	.67
	Clinical range	2.09	.58	1.98	.67
Operated	Normal	1.04	.78	1.24	.90
	Clinical range	1.41	.79	1.98	.78
CSP - Functional					
Not operated	Normal	1.48	.82	1.42	.76
	Clinical range	1.72	.69	2.15	.58
Operated	Normal	1.61	.69	1.34	.78
	Clinical range	1.88	.94	2.15	.80
CSP - Psychological					
Not operated	Normal	1.95	.69	2.04	.67
	Clinical range	2.56	.58	2.70	.64
Operated	Normal	1.75	.73	1.97	.83
	Clinical range	2.82	.62	2.88	.60

Results (*M* and *DS*) for CSP subscales as a function of neurosurgery condition and depression levels are summarized in Table 5. From the MANOVA, significant differences ($p < .001$) emerge for "depression level factor", with higher scores in all dimensions of the CSP questionnaire for participants who fall within the clinical range of depression: Physical $F(1, 168) = 21.14$, $\eta_p^2 = .11$; Social $F(1, 168) = 29{,}73$, $\eta_p^2 = .15$; Functional $F(1, 168) = 20.3$, $\eta_p^2 = .11$; Psychological $F(1, 168) = 19.87$, $\eta_p^2 = .11$. No correlation emerges between anxiety/depression levels and age at neurosurgery or diagnosis, but a positive correlation between anxiety and participant's age (Tab. 6). Severity of Chiari outcomes (CSP Total) correlates positively ($p < .001$) with anxiety ($r = .49$) and depression levels ($r = .64$). Similarly, perceived disability results positively associated ($p = .001$) with anxiety ($r = .25$) and depression symptoms ($r = .47$).

Table 6. Pearson's correlation among Chiari Symptom Profile (CSP) measures, anxiety/depression symptoms (HADS) and participant's age factors

Measure	1	2.	3.	4.	5.	6.	7.	8.
1. CSP Total [1]	1							
2. CSP – Perceived disability	.69 **	1						
3. CSP – QoL	-.62**	-.59**	1					
4. HADS – Anxiety	.49**	.25**	-.40**	1				
5. HADS- Depression	.64**	.47**	-.61**	.59**	1			
6. Age	.23**	.24**	-.18*	.08	.23**	1		
7. Age at diagnosis	.05	.02	-.02	.06	.14	.78**	1	
8. Age at Neurosurgery	-.06	.2. -.02	.06	.02	.12	.81**	.96**	1
M (*SD*)	88.9 (39.9)	.87 (.82)	6.5 (1.8)	8.5 (4.4)	5.9 (4.1)	42.1 (14.2)	32.5 (13.6)	33.6 (15.4)

Note: [1] CSP Total score is calculated on all 57 items

* Correlation is significant at the .05 level (two-tailed)

** Correlation is significant at the .01 level (two-tailed)

4. Discussion

The study aimed to develop and make available to professionals the Italian version of the Chiari Symptom Profile (CSP-Ita), a valid questionnaire assessing the severity of symptoms of CM and their consequences on people's QoL. Since the clinical manifestations of CMs are very heterogeneous, the CSP as a diseases-specific instrument can be very useful for evaluating patient-reported symptoms, their psychological impact, and concrete limitations that can derive from social, physical, and daily activities. The psychometric qualities of the CSP-Ita resulted globally adequate, confirming the reliability and validity of the questionnaire with Italian-speaking respondents. A four-dimensional factorial structure was found that includes all items with the only exception of item 51. Factors correspond to the original dimensions – physical, functional, psychological, and social – identified by Mueller and Oró (2013) and confirmed by Martínez-Sabater et al. (2017) with the Spanish questionnaire. The Cronbach's alphas resulted very good for both the total score and subscales, indicating excellent internal consistency similar to the original English questionnaire. Regarding the construct validity, as hypothesized, the CSP-Ita measures obtained strong negative correlations with general QoL scores, indicating an inverse association between perceived limitations due to CM symptoms and personal well-being domains (physical and psychological health, social relationships, and environment).

Differences in the CSP dimensions did not emerge between people who had undergone decompression neurosurgery or had not been surgically treated. This result does not confirm what was hypothesized, but it suggests an "alignment" of perceived QoL between operated and non-operated people. In other words, it is possible that in the treated group, the more severe symptoms improved to a similar level to the untreated group, as observed in another samples (García et al., 2019).

The literature reports mixed results about the improvements that the patients perceive after the decompression surgery. Some studies indicated significant improvements in QoL assessed one year after (Parker et al., 2013), whereas other found that after decompression symptoms can persist even in 20-40% of cases (Durham et al., 1998). Differences across studies about the benefits after the intervention can depend on the sample's clinical characteristics (Aghakhani et al., 2020), or the methods for assessing surgery outcomes (i.e., patient's retrospective self-evaluation *vs.* pre-post and follow-up measures; Greenberg et al., 2015; Mueller & Oró, 2005). In addition, studies differ in the aspects of QoL specifically measured, such as daily functioning, perceived disability, or general QoL (Almotairi et al., 2020; Bakim et al., 2013; Meeker et al., 2015). Therefore, a standardized instrument such as CSP specifically developed for measuring the QoL related to CM can be extremely useful. First, as disease-specific scale, the CSP can be advantageous for assessing and comparing data from different clinical samples. Secondly, the CSP can allow the clinicians to evaluate patient's daily difficulties and changes in perceived QoL along time. In this regard, a significant result of current study is that perceived QoL worsens with increasing age. This data is to be taken into consideration and underlines the importance of monitoring patients' health conditions beyond the post-operative phase. From a global care perspective, it is recommended to "accompany" the patient to recognize signs of worsening disease or problems and needs (such as familiar, job issues) for which psychological support can be helpful. Interestingly, recent studies evidence that age-related changes in the morphometric cerebellar anatomy, such as a reduction of anterior CSF space in older patients, are associated with increased self-perceived pain, particularly in not operated patients (García et al., 2022). In addition, in the analysed sample significant associations emerged between the perceived severity of symptoms and the levels of anxiety/depression. The prevalence of psychological disorders (participants with HADS scores above the clinical threshold) resulted 32% for anxiety and 14% for depression, with a greater risk of non-operated people presenting clinical signs of depression. These results add to the few studies examining the co-occurrence of psychological symptomatology in patients with CM. Fischbein et al. (2015) found that among 768 adults with CM-I 32% self-reported symptoms of depression and 20% of anxiety. García et al. (2019) found rates of 44% for depression and 60% for anxiety (both at moderate-severe levels), with a higher

likelihood of reaching the clinical cut-off for anxiety among non-operated people. Generally, anxiety and depression are associated with persistent pain, as has been observed in patients with chronic diseases (i.e., rheumatic heart disease; Sheikh et al., 2019). Particularly headache and neck pain, which are the most common symptoms in CM and interfere with cognitive and adaptive functioning (Allen et al., 2014; Almotairi et al., 2020). According to the Fear-Avoidance model of pain, the person tends to reduce or avoid activities that generate intense pain because dysfunctional thoughts can intervene in the perception of pain itself (Gatchel et al., 2016; Lethem et al., 1983). In the short term, the avoidance of physical activities eliminates the proprioceptive sensation of pain. Still, over time it creates a vicious circle, with a sense of frustration, altered mood, helplessness, and increased disability of the person (limitations in self-care, daily living skills, mobility, etc.). Other studies following a neuropsychological approach underline the link between the neuro-anatomic manifestations of CM and deficits in emotional processing observed in patients (such as agitation and panic, hopelessness, dysphoria, and negative mood) particularly the involvement of the cerebellum in affective regulation (Lázaro et al., 2018). In this perspective, the psychological manifestations associated with CM, such as anxiety and depression, may represent not only a reaction to illness and distressing limitations that people perceive on daily functioning by may be a part of CM disease itself and its neuro-anatomic impairment. These studies open a promising perspective to clarify how structural cerebellar abnormalities of CM can contribute to deficits in the emotional processing (Houston et al., 2018) and the vulnerability of CM patients for affective disorders (Lázaro et al., 2018).

5. Conclusions

The presence of psychological disorders in people with CM stresses the need for an accurate multidisciplinary approach, and other interventions besides medical must be considered. Findings from current sample indicate that participants with elevated anxiety/ depression, regardless the neurosurgical history (operated/not operated), report worse QoL in all domains assessed by the Chiari Symptoms Profile questionnaire, with more severe physical symptoms, affective suffering, and reduced daily and social functioning. These results add to other studies suggesting a bidirectional influence on psychological symptoms, physical well-being, and overall QoL. In fact, in the presence of psychiatric disorders, patients with CM-I report more physical pain, discomfort, and lower levels in all domains of QoL (Bakim et al., 2013). In this regard Feghali et al. (2020) recommend investigating and diagnosing the comorbidity between CM disease and a depressive disorder. In fact, some somatic symptoms of depression, such as fatigue, weakness, or headache, may overlap with those of CM and therefore not improve despite surgery.

6. Limitations and future perspectives

The study has a cross-sectional design. Despite having separated the participants on the basis of their neurosurgical history (with/without decompression intervention), we cannot draw conclusions about possible changes in Chiari-related QoL or attribute their actual condition to neurosurgery. In other word, we did not know the severity of CM-I symptoms in the surgically treated group before the intervention, nor how much they changed after neurosurgery. Similarly, we do not know the "natural" course of symptoms (Langridge et al., 2017) in the group of participants who did not undergo surgery. Longitudinal design is needed to further explore to what extent CM patient-reported improved symptoms after intervention, or in which domain (physical, daily activities, etc.) the patient did not feel any benefit. The use of a standardized, disorder-specific measure such as the CSP questionnaire could be very advantageous for this purpose. Secondly, the sample was predominantly female (83%), but this disparity between gender is similar to analogous studies reflecting the higher incidence of CM among women (Fischbein et al., 2015; Martínez-Sabater et al., 2018). In addition, the data were web collected and clinical information were self-reported. The affiliation of participants to an Association of people affected by Arnold Chiari ensured their involvement and motivation for research (there were no dropouts by participants or incomplete questionnaires). However, from a methodological point of view, bias in the comprehension of questions or imprecisions in reporting anamnestic and clinical data cannot be excluded. Finally, the online procedure did not allow researchers to carry out a debriefings phase after completing the questionnaires, so leaving space to the emotional resonances and the expression of needs by the participants. Despite these limitations, this study provides a contribution for the validation of the CSP in Italian language, offering to professional a disorder-specific questionnaire aimed to assessing the severity of Chiari symptoms and their impact on patient's life experience. An evaluation of all domains of QoL, including not only physical symptoms but also emotive and environmental well-being, is useful to quantify the amelioration in post-surgical follow-ups, and to identify those domains in which complementary interventions can be required. For example, high score in the psychological domain of CSP can suggest the need of a psychological intervention for reducing preoperative anxiety before undergoing neurosurgery. Combined medical (i.e., pharmacological pain management) and psychological interventions for patients with CM are recommended, with the common aim of improving individual's well-being and reducing the psychosocial impact of the disease. Empirically supported interventions can include cognitive-behavioral techniques coping with chronic pain, and more recent approaches based on acceptance and commitment therapy (ACT; Holmes et al., 2019). ACT encourages patient's acceptance of the disease condition (i.e., limitations in daily activities) and flexibility (i.e.,

awareness of present condition instead of hyper-alert and anticipation of feeling of pain), and clinical evidences document the efficacy of these interventions in reducing pain and anxiety in chronic diseases (Bailey et al., 2019). However, clinical trials focused on psychological treatment of CM are still scarce. Research on this rare pathology must continue, to explore the factors that predict better outcomes not only from a neurological but also a psychological point of view, and to evaluate the effectiveness of psychological intervention in improving the quality of life of patients with CM.

Conflict of Interest Statement

The authors declare that the research was conducted in the absence of any potential conflict of interest.

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