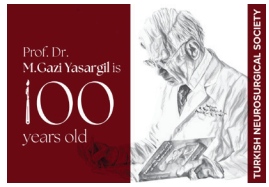
Turk Neurosurg 35(3):503-511, 2025
DOI: 10.5137/1019-5149.JTN.47107-24.2

Original Investigation

General Neurosurgery and Miscellaneous-Others

Received: 09.06.2024
Accepted: 04.10.2024
Published Online: 30.04.2025



Using Patient-Reported Outcome Measures for Adult Chiari Malformation Type 1

Guilherme Lucas de OLIVEIRA LIMA[1,2], Marlon Cesar SOUZA-FILHO[1,2], Luiz Gonzaga de OLIVEIRA-JUNIOR[1], Daniel Felipe Fernandes PAIVA[3], Roger Schmidt BROCK[2], Eberval Gadelha FIGUEIREDO[2]

[1]Nervous System Unit - Federal University of Rio Grande do Norte, Onofre Lopes University Hospital, Natal, Brazil
[2]University of São Paulo, Departament of Neurology, São Paulo, Brazil
[3]Universidade Estadual de Campinas, Faculdade de Odontologia de Piracicaba, Departament of Bioscience, Piracicaba, Brazil

Corresponding author: Guilherme Lucas de OLIVEIRA LIMA ✉ guilhermelimaneurocirurgia@gmail.com

ABSTRACT

AIM: To validate the Chicago Chiari Outcome Scale (CCOS) as a patient-reported outcome measures (PROMs) tool and identify the cutoffs of CCOS-PROM and EuroQoL-5D-3L score index (EQ-SI) against Gestalt in adults who underwent chiari malformation type 1 (CM1) treatment.

MATERIAL and METHODS: This cross-sectional study analyzed the health status of CM1 adults after a 12-month surgery follow-up. An electronic form was sent to groups of CM patients. The self-Gestalt good perception was distributed as a cure or improvement for validation purposes. AUROC was evaluated to verify the ability of each PROM to predict Gestalt outcomes. Ethical approval registry: 64148822.0.0000.5292.

RESULTS: The 85 subjects had a mean age of 42.1(±10.6) years and a follow-up of 4.8(±4.1) years. Gestalt outcome was good for 23(27%) patients. The median CCOS-PROM of 74 patients was 11(IQR 4). The median EQ-SI of 84 patients was 0.52(IQR 0.29). CCOS-PROM score of 13 was the optimal cutoff value (sensitivity 82.6%; specificity 78.4%). For EQ-SI, this value was 0.64 (sensitivity 82.6%; specificity 75.4%). The AUROC for CCOS-PROM and EQ-SI were 0.90 and 0.86, respectively.

CONCLUSION: CCOS-PROM is valid for assessing the outcome of CM1 surgery in adults. CCOS-PROM score of 13 and EQ-SI of 0.64 were the best tradeoff values.

KEYWORDS: Arnold-Chiari malformation, Treatment outcome, Cauda equina, Posterior cranial fossa, Surveys and questionnaires

INTRODUCTION

Surgical treatment of Chiari malformation type 1 (CM1) aims to prevent deterioration and improve symptoms (3,8,26). However, assessing CM1 surgical outcomes is a challenging task due to the use of different approaches, such as general versus disease-specific scales (19), and patient- versus physician-centered perspectives (12). Patient-centered assessment is essential for practice (27), yet few studies have presented patient-reported outcome measures (PROMs) for CM1 treatment (11,15).

PROMs are scarce in neurosurgery (12). The EuroQoL-5D-3L assesses general health-related quality of life across five dimensions: mobility, self-care, usual activities, pain or discomfort, and anxiety/depression. The scale employs a three-level severity scale. The EQ-5D-score index (EQ-SI) summarizes 243 possible health states (27). Several studies have discussed the validation of this tool in CM1 (13,15,19,30), but an optimal cutoff point for a good outcome remains undefined.

Guilherme Lucas de OLIVEIRA LIMA : 0000-0001-6633-8723
Marlon Cesar SOUZA-FILHO : 0009-0004-4961-0042
Luiz Gonzaga de OLIVEIRA-JUNIOR : 0000-0002-8065-3866
Daniel Felipe Fernandes PAIVA : 0000-0003-4186-9856
Roger Schmidt BROCK : 0000-0002-6186-3200
Eberval Gadelha FIGUEIREDO : 0000-0001-7754-4178

The Chicago Chiari Outcome Scale (CCOS) is a disease-specific and physician-centered tool designed as a flowchart (1), and recently validated for CM1 adults (2). This scale had high interrater reliability and consistency in detecting good overall outcomes (1,2,15,31). An advantage of the CCOS is time efficiency over the other scales (1,19), due to its summed score of four items, giving a simple range of 4 to 16, the optimal score varies under validation studies between 13 (1,2), and 14 (31). These studies discuss the interpretation bias in a retrospective analysis of patient medical records. The CCOS flowchart can be adapted as a PROM to avoid this bias.

This study aimed to validate the CCOS as a PROM tool and identify the optimal cutoffs of CCOS-PROM and EQ-SI, compared against patients' self-assessment of their overall health (referred to as 'gestalt' outcomes), in adults undergoing CM1 treatment.

MATERIAL and METHODS

The Institutional Research Ethics Committee approved the study under the reference number 64148822.0.0000.5292 on December 15, 2022.

A cross-sectional study was conducted using a self-administered online questionnaire from adult patients who had undergone surgical treatment for CM1 at least 12 months prior to the investigation. Participants were recruited from CM1 patient support groups in two rounds of invitations, with a 15-day interval between each round.

After consent was obtained, the patients completed a questionnaire. The questionnaire included inquiries regarding sex, current age, age at the time of the most recent surgical procedure, the type of CM, the presence of comorbidities (basilar invagination, hydrocephalus, or syrinx), preoperative symptoms, the type of surgical procedure, postoperative complications, current self-assessed clinical status (gestalt), self-reported CCOS (CCOS-PROM), and the EuroQoL-5D-3L scale. The follow-up period was calculated in years based on the patient's current age and age at their last surgical procedure.

Additionally, patients were asked whether they experienced a level of improvement in their general health (gestalt) following surgery (meaningful effectiveness). The gestalt outcomes were distributed into three categories: 0 for worse, 1 for stable, and 2 for improved or resolved symptoms.

The CCOS is a four-level disease-specific scale distributed across four categories: painful symptoms, non-painful symptoms, functionality, and complications (1,2,31). Originally developed as a flowchart (1), CCOS was converted for use as a PROM with affirmative sentences (Table I). The score is calculated by summing up the four items, with a total ranging from 4 to 16 (1,2,14,31). The EQ-SI was calculated using a previously defined scoring algorithm (27).

We grouped the gestalt categories for worse or unchanged to assess the accuracy of the CCOS-PROM and EQ-SI in identifying patients who reported improvement or resolution of symptoms. The area under the receiver operating characteristic curve (AUROC) was calculated using a previously described methodology (4,31). The AUROC was interpreted as follows: a score of 1.0 indicated a perfect test, 0.9–0.99 indicated an excellent test, 0.8–0.89 indicated a good test, 0.7–0.79 indicated a fair test, and scores under 0.7 indicated non-useful tests (6). Interrater reliability assessment was unnecessary, as the patients self-reported their clinical outcomes. The Youden's index (sensitivity + specificity - 1) was employed to identify the optimal cutoff value for each PROM in detecting favorable outcomes.

Anktowiak et al. propose using the EQ-SI tool to validate CCOS (2). Our study yielded a secondary result: the indirect comparison of the CCOS-PROM with the EQ-SI tool, following the same methodology described for the validation using the AUROC. In this case, the CCOS-PROM was calibrated with the description of health or illness obtained from the EQ-SI cutoff score.

Descriptive statistical analyses were conducted on the entire sample, regardless of the type of surgery performed. The scores were presented as the median with interquartile range (IQR). The statistical software Jamovi (version 2.3.26) was employed for data analysis, with $p<0.05$ indicating statistical significance.

RESULTS

During the recruitment period, 153 questionnaires were received. Fifty-eight were excluded for absence of surgery, seven for being under-age, and three for being Chiari malformation type II. Eleven patients did not complete the CCOS, and the remaining 74 patients for CCOS-PROM validation were based on self-reported gestalt perception of improvement. One respondent failed to complete the EQ-5D, and the remaining 84 patients were included in the EQ-SI validation group. The characterization of each PROM validation group is presented in Table II.

Eighty-five patients self-reported gestalt outcomes. Of these, 23 (27%) perceived an improvement, 37 (44%) stabilization, and 25 (29%) a worsening of general health.

The median CCOS-PROM score was 11 (IQR 6). Figure 1 illustrates the relative distribution of CCOS-PROM scores in relation to gestalt outcomes. All 22 cases of aggravation of symptoms occurred with a score of under 11. A score of 13 represented the optimal cutoff for gestalt self-perception (Table III). The AUROC for CCOS was 0.90 (Figure 3A).

The calculated EQ-SI median was 0.520 (IQR 0.294). The relative distribution of EQ-SI in relation to the gestalt response is presented in Figure 2.

The cutoff value for discriminating self-reported overall outcomes was 0.64 (Table IV). The AUROC for EQ-SI was 0.86. (Figure 3B).

A CCOS-PROM score of 12 was the optimal cut-off to determine the subgroup of patients with an EQ-SI greater than 0.64 (Table V). The AUROC for CCOS-PROM was 0.93 (Figure 4).

Table I: Adaptation of CCOS Flowchart Statements into CCOS-PROM with Respective Scores

CCOS	CCOS-PROM	Score
Is the patient completely without any Chiari-related pain/ non-pain symptoms?	I am completely without any Chiari-related pain/ non-pain symptoms.	4
Are the patient's preoperative Chiari-related symptoms improved or well managed medically? Or Are any new postoperative Chiari-related pain/ non-pain symptoms managed so as not to interfere with the patient's activity?	My preoperative Chiari-related symptoms improved or are well managed medically. Or Are any new postoperative Chiari-related pain/ non-pain symptoms managed so as not to interfere with my activity?	3
Are the patient's preoperative Chiari-related symptoms unchanged?	I have my Chiari-related symptoms unchanged.	2
The patient is worse or there are new pain/ non-pain symptoms refractory to medical treatment.	I am worse or there are new pain/ non-pain symptoms refractory to medical treatment.	1
Does the patient experience no interference in their daily activities of living from their Chiari-related symptoms?	I experience no interference in my daily activities of living from my Chiari-related symptoms.	4
Is the patient able to attend work or school the majority (>50%) of the time	I can attend work or school for more than half a week.	3
Is the patient able to attend but his/her attendance is significantly impaired (<50% of the time) by his/her Chiari-related symptoms?	I can attend but my attendance is significantly impaired (less than half days of a week) by Chiari-related symptoms.	2
The patient is unable to attend work or school due to Chiari-related symptoms	I am unable to attend work or school due to Chiari-related symptoms.	1
Did the patient experience no complications after surgery?	I experienced no complications after surgery.	4
Did the patient experience transient complications after surgery that have since been resolved?	I experienced transient complications after surgery that have since been resolved.	3
Does the patient experience persistent complications after surgery that can be managed medically or surgically?	I experience persistent complications after surgery that can be managed medically or surgically.	2
There are persistent and poorly controlled post-operative complications.	I have persistent and poorly controlled post-operative complications.	1

CCOS: *Chicago Chiari outcome scale,* ***PROM:*** *Patient-reported outcome measures.*

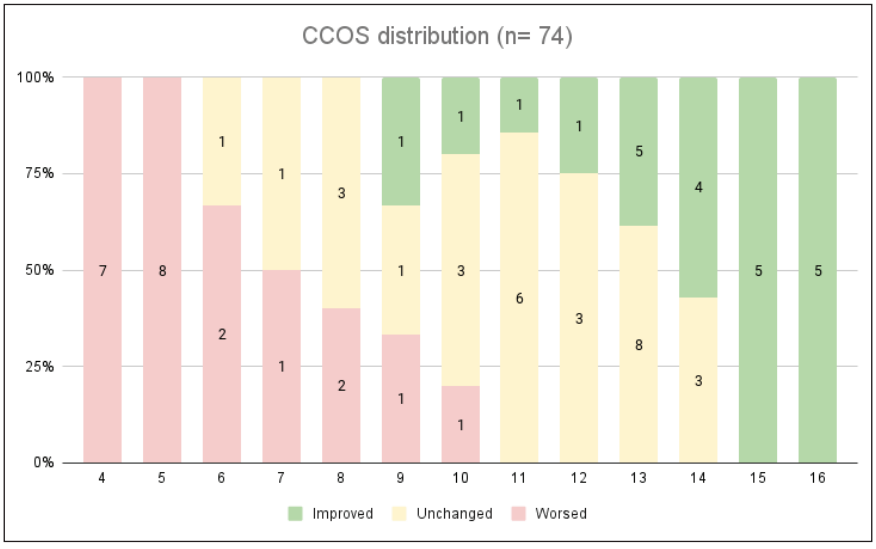


Figure 1: Distribution of Chicago Chiari outcome scale - patient-reported outcome measures (CCOS-PROM) scores related to gestalt outcome categories.

Table II: Characterization of the Groups for Validation of Each PROM Related to Gestalt

	CCOS (N=74)	EQ-SI (N=84)
Sex		
Female	58 (78.4)	66 (77.6)
Age (Years at surgery)	41.7 ± 10.8	41.6 ± 10.7
Chiari I subtypes		
0	4 (5.4)	5 (6.0)
1	64 (86.5)	72 (85.7)
1,5	6 (8.1)	7 (8.3)
Comorbidities	60 (81)	68 (91)
Symptoms		
Occipital headache	51 (68)	54 (64)
Other headache	57 (76)	63 (75)
Cervicobrachial pain	58 (77)	63 (75)
Dysestesias	41 (55)	47 (56)
Otovestibular	61 (81)	66 (79)
Swallowing/ Speech	49 (65)	51 (61)
Cerebelar	42 (56)	47 (56)
Paresia	65 (87)	72 (86)
Parestesia	58 (77)	66 (79)
Surgery		
PFD	35 (47.3)	43 (51.2)
SFT	25 (33.8)	25 (29.8)
Other or multiple	14 (17.6)	16 (19.0)
Follow-up (Years)	4.66 ± 4.17	4.84 ± 4.11
Complications	18 (24)	21 (25)

Values are presented as median (range) or number (%). ***CCOS:*** *Chicago Chiari outcome scale;* ***PROM:*** *patient-reported outcome measures;* ***EQ-SI:*** *EuroQoL-5D-score index;* ***PFD:*** *Posterior Fossa Decompression;* ***SFT:*** *Sectioning of filum terminale.*

DISCUSSION

The literature reveals a paucity of validated and reliable patient-centered assessment tools for the surgical outcomes of CM1 (2,8,11,13,15,19). We present the CCOS and EQ-SI as valuable PROM tools for assessing the surgical treatment of CM1 in adults and have determined the ability of each PROM to identify patients with improved gestalt outcomes. Furthermore, we were also able to provide an optimal EQ-SI cutoff value for CM1.

Recently, PROMs have gained popularity and now play an important role in the enhanced evaluation of treatment effectiveness, patient satisfaction, communication (12,27), and several other parameters that have shifted the focus of analysis from the physician's perspective to the patient's. CCOS-PROM brings a patient-centered perception of meaningful effectiveness for the treatment of CM1. Given the range of CM classifications and therapeutic options (8,9), patient-centered outcome assessment tools are important to objectively establish cutoffs for meaningful outcomes. In this context, a CCOS-PROM score of 13 was identified as the optimal trade-off value for detecting favorable self-reported gestalt outcomes in adults, comparable to those validated recently through retrospective chart analysis (2). This CCOS-PROM cutoff provides a reasonable measure for interpreting patients' perceptions of CM treatment independent of any decision criteria (23), such as the surgical procedure performed.

Concerning quality of life, the EuroQoL-5D-3L is a CM-validated PROM that includes a visual analog scale (VAS) (12,13). However, the EQ-VAS is not reliable enough to be used as part of routine clinical studies, according to the EuroQol group (21). The poor accuracy in VAS results led to the adoption of the EQ-SI as the most appropriate measure. The EQ-SI provides good accuracy against gestalt measures, with similar AUROC as a previous validation study (13). Despite a broader range in EQ-SI (from 0.57 to 0.69) for the discrimination of good general health status (Figure 2), a cutoff point of 0.64 was identified as the optimal balance between sensitivity and specificity.

The CCOS validation study discusses the limitations of gestalt measures in defining good outcomes due to its known subjectivity (1,2), suggesting the EQ-SI as a more appropriate tool (2). Our data allowed such validation, showing a CCOS-PROM score of 12 as the threshold for a better quality of life. This patient-centered shift may bring to light a different outcome perspective and might explain the underperformance in the aforementioned group. However, these data present a secondary interpretation of the results.

Using a CM support group provided a direct response from the patient's perspective concerning the disease. However, this convenience sampling may lead to a less-than-optimal outcome (28) and an imbalanced representation of variables. Nevertheless, our sample represented many CM characteristics, such as female predominance, a multitude of CM types, comorbidities, symptoms, and surgical treatments. The reason for the prevalence of female sex, as reported in the literature (2,4,24), remains unclear, and further research is needed to establish any possible underlying genetic or hormonal factors.

The other findings may, in part, be explained by the proper definition of CM as a heterogeneous group of disorders related to the obstructed flow of cerebrospinal fluid at the foramen magnum (8). Moreover, several variations of CM differ from the classic threshold of 5mm of herniation below the foramen magnum, such as CM type 0 (16), or involve only the tonsil's herniation, such as in CM type 1.5 (7,9,22,29). The lower frequency of CM type 1.5 observed in our series compared with the literature may have influenced improved outcomes (9,29). However, the elevated number of comorbidities, including syrinx and craniovertebral junction distortion (5,9,20,26), may have counterbalanced this effect.

Table III: CCOS-PROM Validity for Gestalt Prediction in 74 Patients

Cutoff point	Sensitivity (%)	Specificity (%)	PPV (%)	NPV (%)	Youden's index	AUROC
4	100	0	31.08	-	0	0.90
5	100	13.73	34.33	100	0.137	
6	100	29.41	38.98	100	0.294	
7	100	35.29	41.07	100	0.353	
8	100	39.22	42.59	100	0.392	
9	100	49.02	46.94	100	0.490	
10	95.65	52.94	47.83	96.43	0.486	
11	91.3	60.78	51.22	93.94	0.521	
12	86.96	72.55	58.82	92.5	0.595	
13*	**82.61**	**78.43**	**63.33**	**90.91**	**0.610**	
14	60.87	94.12	82.35	84.21	0.550	
15	43.48	100	100	79.69	0.435	
16	21.74	100	100	73.91	0.217	

CCOS: *Chicago Chiari outcome scale,* ***PROM:*** *Patient-reported outcome measures,* ***PPV:*** *Positive predictive value,* ***NPV:*** *Negative predictive value,* ***AUROC:*** *Area under the receiver operating characteristic curve.*

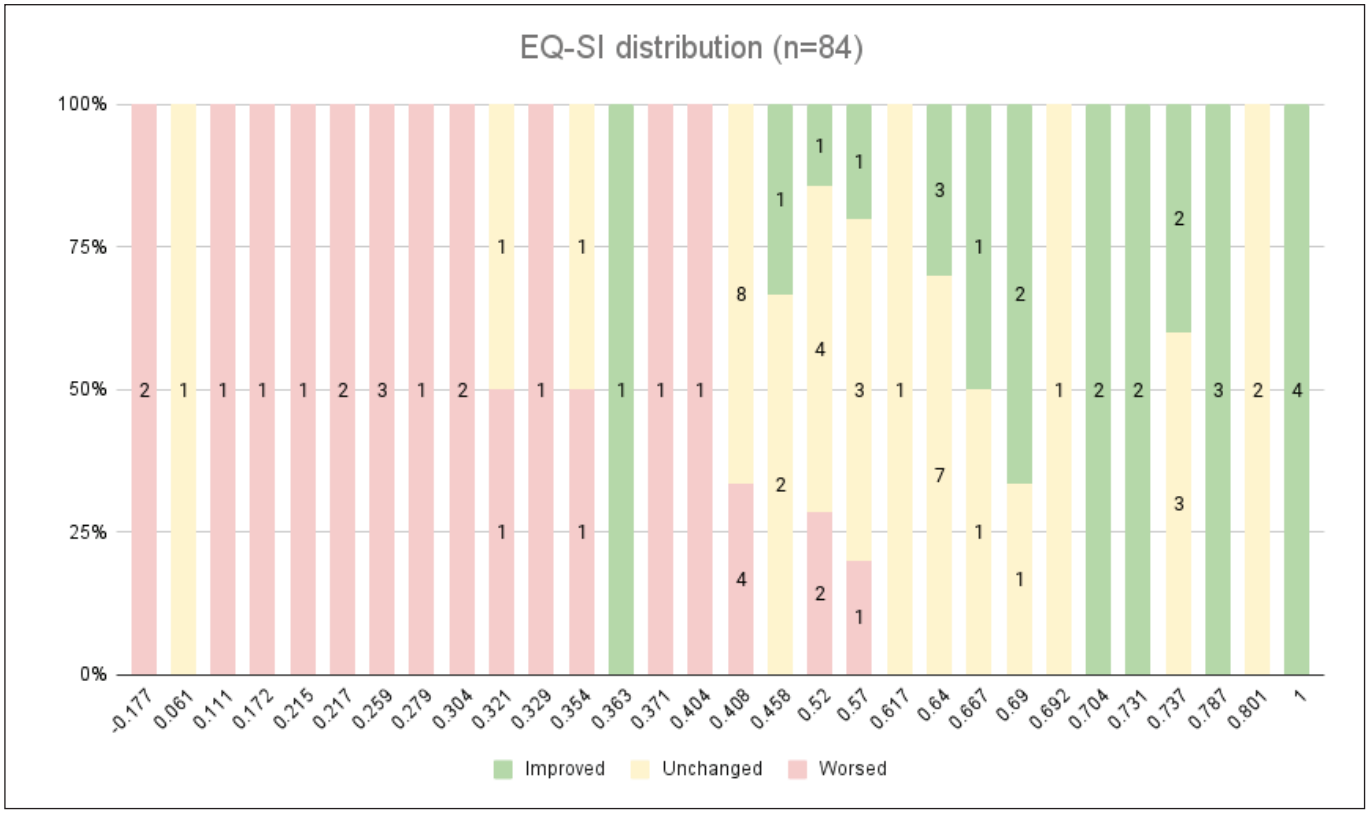


Figure 2: Distribution of EuroQoL-5D-score index (EQ-SI) scores related to gestalt outcome categories.

Table IV: EQ-SI Validity for Gestalt Prediction in 84 Patients

Cutoff point	Sensitivity (%)	Specificity (%)	PPV (%)	NPV (%)	Youden's index	AUROC
-0.177	100	0	27.38	-	0	0.86
0.061	100	3.28	28.05	100	0.032	
0.354	100	27.87	34.33	100	0.279	
0.363	100	31.15	35.38	100	0.312	
0.371	95.65	31.15	34.38	95	0.268	
0.404	95.65	32.79	34.92	95.24	0.284	
0.408	95.65	34.43	35.48	95.45	0.301	
0.458	95.65	54.1	44	97.06	0.498	
0.52	91.3	57.38	44.68	94.59	0487	
0.57	86.96	67.21	50	93.18	0.542	
0.617	82.61	73.77	54.29	91.84	0.564	
0.64*	**82.61**	**75.41**	**55.88**	**92**	**0.580**	
0.667	69.57	86.89	66.67	88.33	0.565	
0.69	65.22	88.52	68.18	87.1	0.537	
0.692	56.52	90.16	68.42	84.62	0.467	
0.704	56.52	91.8	72.22	84.85	0.483	
0.731	47.83	91.8	68.75	82.35	0.396	
0.737	39.13	91.8	64.29	80	0.309	
0.787	30.43	96.72	77.78	78.67	0.272	
0.801	17.39	96.72	66.67	75.64	0.141	
1	17.39	100	100	76.25	0.174	

EQ-SI: *EuroQoL-5D-score index,* ***PPV:*** *Positive predictive value,* ***NPV:*** *Negative predictive value,* ***AUROC:*** *area under the receiver operating characteristic curve.*

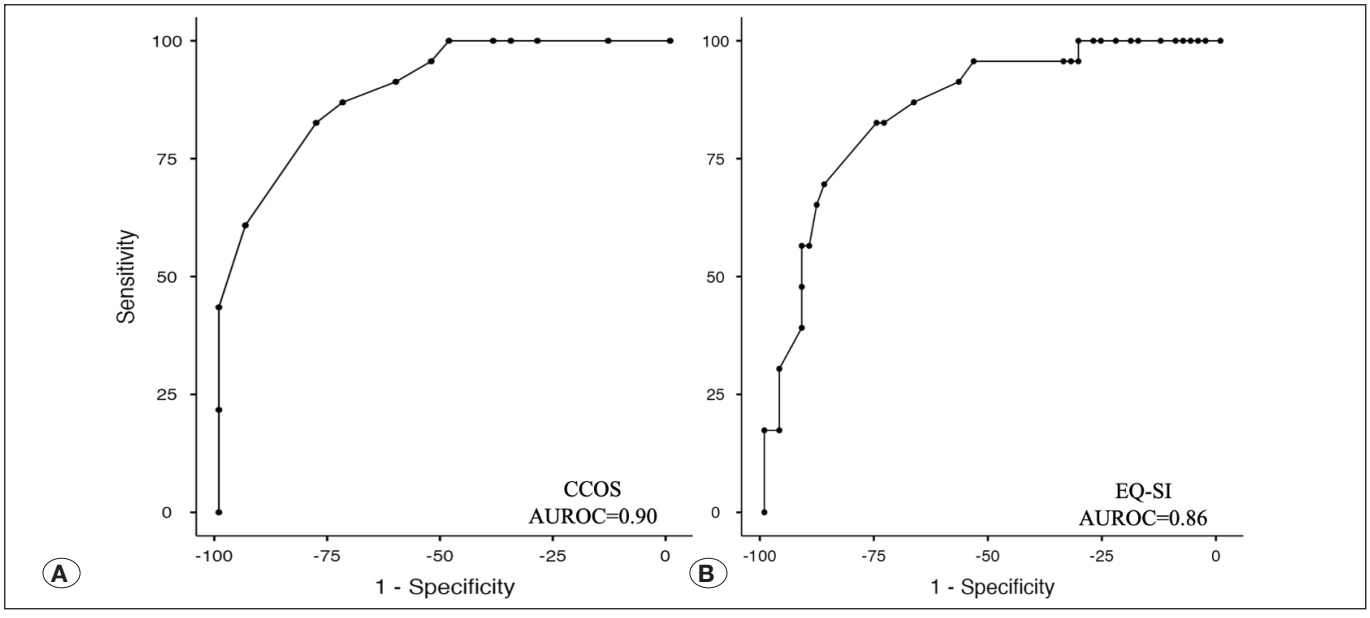


Figure 3: ROC curves for gestalt improvement. **A)** Chicago Chiari outcome scale - patient-reported outcome measures (CCOS-PROM) (AUROC = 0.90). **B)** EuroQoL-5D-score index (EQ-SI) (AUROC = 0.86).

Table V: CCOS-PROM Cutoff Against EQ-SI Discriminative Score for Better Quality of Life in 74 Patients

Cutoff point	Sensitivity (%)	Specificity (%)	PPV (%)	NPV (%)	Youden's index	AUROC
4	100	0	42.47	-	0	0.93
5	100	16.67	46.97	100	0.167	
6	100	35.71	53.45	100	0.357	
7	100	42.86	56.36	100	0.429	
8	96.77	45.24	56.6	95	0.420	
9	96.77	57.14	62.5	96	0.539	
10	96.77	61.9	65.22	96.3	0.587	
11	96.77	73.81	73.17	96.88	0.706	
12*	**90.32**	**85.71**	**82.35**	**92.31**	**0.760**	
13	77.42	85.71	80	83.72	0.631	
14	51.61	97.62	94.12	73.21	0.492	
15	32.26	100	100	66.67	0.323	
16	16.13	100	100	61.76	0.161	

CCOS: *Chicago Chiari outcome scale,* ***PROM:*** *Patient-reported outcome measures,* ***EQ-SI:*** *EuroQoL-5D-score index,* ***PPV:*** *Positive predictive value,* ***NPV:*** *Negative predictive value,* ***AUROC:*** *Area under the receiver operating characteristic curve.*

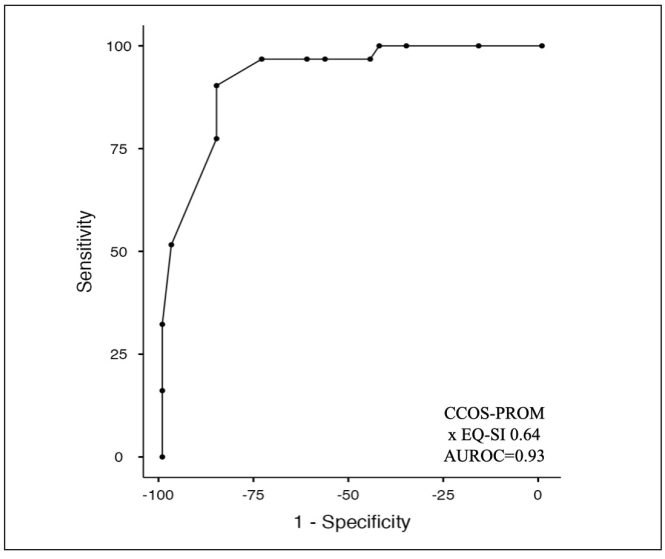


Figure 4: Receiver operating characteristic (ROC) curve for the ability of Chicago Chiari outcome scale - patient-reported outcome measures (CCOS-PROM) to identify better outcomes in quality of life (EuroQoL-5D-score index EQ-SI superior to 0.64) after Chiari malformation surgery in 74 patients. AUROC = 0.93.

The reported symptoms of CM1 result from the distortion of nervous system structures, including the cerebellum, brainstem, and spinal cord (18). The frequency of headaches among our cohort was comparable to that reported in the literature. However, other symptoms, particularly occipital pain triggered by Valsalva's maneuver, vestibulocochlear abnormalities, swallowing, or speech difficulties, were more frequent in our series (10,17). This combination, including spinal disturbances and sphincter dysfunction, has been controversially described as Neuro-Cranio-Vertebral syndrome (24,25).

Concerning treatment, one-fifth of our sample underwent sequential surgeries to alleviate symptoms or prevent deterioration (8,18,22). As the surgeries were performed at multiple centers, complications may have occurred due to variability in the surgeons' experience, or the techniques used.

This study has limitations that should be considered when interpreting the findings. First, the use of postal questionnaires to obtain outcome scores may not fully reflect the results of clinical examinations or personal interviews. Second, cross-sectional data collection raises concerns about generalizability. Regarding the validation of CCOS-PROM against EQ-SI, it is important to note that although the statistical results were promising, the sample was non-specific, preventing firm conclusions. Therefore, it is necessary to conduct a specific evaluation to confirm the current results.

This study is innovative in recommending the CCOS-PROM as a specific patient-centered tool in adults with CM. It has a similar cutoff point to the original physician-centered version (a CCOS score of 13). The study also contributes to the literature by defining an optimal EQ-SI 0.64 cutoff point for this disorder.

CONCLUSION

CCOS-PROM and EQ-SI show good accuracy for gestalt outcomes in surgically treated adults with CM1. The proposed cutoff point of 13 for CCOS-PROM was found to be appropriate

for assessing surgical outcomes, and a score of 0.64 for EQ-SI was appropriate for assessing quality of life after treatment. Further studies assessing the CCOS-PROM against the EQ-SI cutoff of 0.64 are needed to confirm our secondary findings.

ACKNOWLEDGEMENTS

We thank the Supporting Groups of Chiari Malformation and Rare Diseases (Monikeyt Ferreira and Rafaela Forner) for their contribution.

Declarations

Funding: There were no grants or funding for this research.

Availability of data and materials: The datasets generated and/or analyzed during the current study are available from the corresponding author by reasonable request.

Disclosure: The authors declare no competing interests.

AUTHORSHIP CONTRIBUTION

Study conception and design: GLOL, DFFP, RSB, EGF

Data collection: GLOL, DFFP, MCSF, LGOJ

Analysis and interpretation of results: GLOL, MCSF, LGOJ, DFFP, RSB, EGF

Draft manuscript preparation: GLOL, MCSF, LGOJ, DFFP, RSB, EGF

Critical revision of the article:

All authors (GLOL, MCSF, LGOJ, DFFP, RSB, EGF) reviewed the results and approved the final version of the manuscript.

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