
Article:
Piper, R.J. orcid.org/0000-0002-6422-5853, Afshari, F.T., Soon, W.C. et al. (23 more authors) (2025) Quality-of-life outcomes in adults and children with Chiari 1 malformation and in those managed without surgery: a prospective, multicenter, observational study. Neurosurgery. ISSN: 0148-396X

https://doi.org/10.1227/neu.0000000000003718

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Quality-of-Life Outcomes in Adults and Children With Chiari 1 Malformation and in Those Managed Without Surgery: A Prospective, Multicenter, Observational Study

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Oral abstract presented at the British Neurosurgical Research Group Meeting, Oxford, UK, November 7-8, 2024.

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Received, April 15, 2025; **Accepted,** June 24, 2025; **Published Online,** September 9, 2025.

Neurosurgery 00:1–16, 2025 https://doi.org/10.1227/neu.0000000000003718

BACKGROUND AND OBJECTIVES: Chiari 1 malformation (CM1) is a common MRI finding and a frequent reason for neurosurgical consultation. Although many studies have investigated surgical outcomes for patients with CM1, outcomes for those treated without surgery have been less frequently reported. The UK Chiari 1 Study reports the quality of life of adults and children with CM1 treated without surgery, 12 months after the first neurosurgical clinic visit.

METHODS: The UK Chiari 1 Study was a prospective, multicenter cohort study of adults (≥16 years) and children (<16 years) with CM1. This was an observational study that did not alter the course of clinical care. Symptoms and quality-of-life data (using Short-Form 36 in adults and the Pediatric Quality of Life Inventory™ in children) were collected at baseline and 12 months after the first clinical review for all participants.

RESULTS: One hundred ninety-two patients with CM1 (146 females; 148 adults) were studied at baseline, and 113 patients with CM1 treated without surgery were studied at a 12-month follow-up. Baseline quality-of-life scores in the study cohort were significantly lower in every domain compared with normative control data, in both adults and children. There were no decreases in quality-of-life subscores after 12 months in this cohort of adults and children with CM1 treated without surgery. Social functioning ($t = -40$, $P < .001$) and bodily pain ($t = -2.9$; $P = .03$) Short-Form 36 scores showed improvements at 12 months in adult patients treated without surgery.

CONCLUSION: This study demonstrates the stability of quality-of-life domains in adults and children with CM1 after 12 months who have been managed without surgery. Further studies are required to understand the determinants of poor quality of life in patients with CM1 and to investigate interventions for improving quality of life. There is a further need for robust comparison of surgical and nonsurgical management for patients with CM1.

KEY WORDS: Chiari 1 malformation, Patient-reported outcome measures, Pediatrics, Quality of life

ABBREVIATIONS: CM1, Chiari 1 malformation; **FMD,** foramen magnum decompression; **HRQoL,** health-related quality of life; **PedsQL,** Pediatric Quality of Life Inventory; **SF36,** Short-Form 36; **UKC1S,** Chiari 1 Study.

Supplemental digital content is available for this article at neurosurgery-online.com.

Chiari 1 malformation (CM1) is a structural abnormality of the hindbrain characterized by descent of the cerebellar tonsils through the foramen magnum.[1,2] CM1 is a common finding estimated to be present in 8 in 1000 people, affecting both children and adults.[3]

Patients with significant or worsening symptoms believed to be related to the CM1 may undergo further investigation or intervention. These include diagnostic intracranial pressure monitoring,[4] foramen magnum decompression (FMD),[5] and cerebrospinal fluid diversion (ventriculoperitoneal shunt or endoscopic third ventriculostomy[6,7]). For many patients, nonoperative management is chosen. CM1 is often encountered incidentally when cross-sectional imaging of the brain or cervical spinal cord is undertaken for other reasons.[3]

CM1 is a frequent reason for neurosurgical consultation. Despite the relatively high volume of patients with CM1 in general neurosurgical practice, recent survey-based studies of neurosurgical practice suggest uncertainty surrounding investigation and decision-making, particularly regarding who would most benefit from surgical intervention.[8-10] Previous studies of the management of CM1 have mostly been small, single-center, and retrospective. Many studies have investigated outcomes after surgery for CM1, but there are only a small number of studies of outcomes after the first assessment in clinic and of nonoperative management.[11] The outcome measures used in previous studies have typically been surgeon-reported, and very few studies have included patient-reported outcome measures. The limitations of current evidence are an important reason for the lack of consensus in management.

This first data analysis of the UK Chiari 1 Study ('UKC1S') reports the baseline data of the study, including symptoms, signs, radiological findings, and the variation of UK neurosurgical management of the total UKC1S patient cohort. The study primarily investigates health-related quality of life (HRQoL) in patients with CM1 managed without surgery at first clinic appointment and 12 months later.

METHODS

Study Design

The *UKC1S* was a prospective, multicenter, observational, cohort study of patients of all ages with CM1, conducted according to a predefined published clinical investigation protocol[12] and reported according to the STROBE (Strengthening the Reporting of Oservational Studies in Epidemiology) Checklist[13] (**Supplementary File 1,** http://links.lww.com/NEU/E970). This study collected patient- or parent-reported (hereafter collectively referred to as patient-reported) and surgeon-reported longitudinal data at 3 time points: (1) baseline (within 45 days of the first neurosurgical encounter), (2) 12-month follow-up, and (3) 12 months postoperatively, if applicable. These first results of the study focus on data from patients treated without surgery.

The study was led and delivered by the British Neurosurgical Trainee Research Collaborative (https://www.bntrc.org.uk/) and followed a model for collaborative and trainee-led research.[14-16] All UK neurosurgical units were invited to collaborate on this study.

Study Registration and Ethical Approval

This study received approval from the East Midlands Leicester South Research Ethics Committee (reference: 20/EM/0053), and each participating clinical site granted local research governance approvals. The study followed the principles outlined in the Declaration of Helsinki. Informed and signed consent was acquired from all patients and/or parents as appropriate for age.

Participant Eligibility

Patients were identified and screened for eligibility by the local neurosurgical team by monitoring records from neurosurgical outpatient clinics. Patients included in this study were those of any age attending their first neurosurgical clinic or consult within the past 45 days. Patients 16 years and older are henceforth described as adults. Radiological diagnosis of CM1 was confirmed by the neurosurgical team. Exclusion criteria included the following: (1) alternative Chiari malformation or other conditions not fitting criteria for CM1, (2) a history of spinal dysraphism, (3) previous neurosurgical consultation for CM1, or (4) previous neurosurgical intervention (other than lumbar puncture) for any reason.

Data Collection

All data were recorded using REDCap (https://projectredcap.org/).[17]

Patients were asked to report their symptoms, family history, and HRQoL. Adults (16 years and older) were asked to complete the 36-Item Short Form Health Survey (SF-36; UK Version #1). Parents of the children aged 2-15 years were asked to complete the Pediatric Quality of Life Inventory™ (PedsQL™; UK version 4.0) (www.mapi-trust.org). Higher Short-Form 36 (SF36) and PedsQL scores are associated with a higher quality of life.

Surgeons from each neurosurgical unit were asked to report details of the patients' demographics, medical history, radiological findings, clinical decision-making, and, if applicable, the surgical techniques used. The template case report form documentation has previously been made openly available as a supplement to the study protocol.[12]

Imaging

The date of diagnosis of CM1 was taken as the date of first imaging confirming CM1.

The collaborating surgeons from each neurosurgical unit measured cerebellar tonsillar descent on the patient's first MRI by measuring the

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maximum herniation below McRae's line to the nearest millimeter. Additional radiological variables for both the first and follow-up MRI studies were requested, including the presence of hydrocephalus and the presence and levels of syringomyelia.

Data Analysis

Demographic and imaging data are presented to characterize the whole UKC1S cohort and the nonoperative cohort. For reference, baseline data are also presented for the group treated with FMD. Data are reported as percentages or median and IQR values unless otherwise specified. The baseline clinical characteristics of the patients treated without surgery were compared with those treated with FMD using either a nonpaired Welch *t*-test (continuous data) and the Fisher's exact count test (categorical data).

The baseline HRQoL data were compared with normative data sets using an unpaired Welch *t*-test. The baseline SF36 data of adults with CM1 were compared with those of a normative cohort of Welsh adults, one of the most recent data sets using the same UK version of the SF36 assessment.[18] The sample of the adult normative cohort was different for each subscore (with a range of n = 837-1308 responses). Baseline PedsQL data were compared with a matching normative data set of 665 healthy Welsh children.[19] As PedsQL only allows for data from those 5 years and older to be amalgamated, patients younger than 5 years were excluded from this analysis. Normative data for both the adult and pediatric cohorts were available only as mean and SD values, so a paired student's *t*-test was used to compare the total and each domain of the PedsQL data between children with CM1 and controls.

Longitudinal analyses were only performed for patients for whom both the surgeon-reported and patient-reported data were complete. This was to ensure the treatments each patient received were known. Paired *t*-tests were used to compare each subscore of the quality-of-life subscores between baseline and 12 months in patients with CM1 treated without surgery.

In the adult group, a multivariate linear regression analysis was performed to identify the clinical variables associated with baseline SF36 subscores. Clinical variables classically associated with CM1 were used to build the model, including headache (any type), classic CM1 headache (Valsalva-induced and occipital), back pain, neck pain, limb pain, paresthesia, weakness, poor balance, poor coordination, coexisting psychiatric diagnosis, and presence of a syrinx.

Patients were removed from the analyses for which their respective data were missing, with reporting and transparency of missing data elements throughout.

Throughout, statistical analyses were performed using R (Version 4.1.0). Statistical significance was confirmed when statistical tests provided a value of $P < .05$. Data are presented to 1 decimal place unless the value is lower than .1. Correction for multiple comparisons was performed using the *Bonferroni* method.

Data availability

Patient consent has not been acquired for externally sharing individual patient data.

RESULTS

Recruitment and *UKC1S* Characteristics

Two hundred seventy-five patients from 24 recruiting units were recruited between 8-Oct-20 and 7-Apr-22, of which 211 patients were found to be eligible (**Supplementary Figure 1 and Supplementary Table 1,** http://links.lww.com/NEU/E970). One hundred ninety-two patients had complete patient-reported and surgeon-reported data at baseline (Figure 1).

In the total UKC1S cohort (n = 192), 146 patients (76.0%) were female, with a median age of 28.2 (IQR 16.6–38.6) years. One hundred forty-eight were adults (126/148 females, median age 32.5 [IQR 25·8-40.8] years), and 44 were children (20/44 females, median age = 11.1 [IQR 5·7-13.9] years). The most frequent reasons for referral were symptoms attributed to CM1 (59.6%) or an incidental finding of a CM1 (32.1%). Other referral reasons are shown in Table 1. The median time between radiological diagnosis and referral was 27.5 (7·0-85.0) days (data available for 130/192). The median time between referral and consultation was 82.0 (35·0-132.5) days (data available for 147/192).

A family history of CM1 was reported in 4.7% of patients in the overall cohort, and this was higher in the pediatric group compared with the adult group (9.1% vs 3.4%, respectively).

Symptoms and Signs at Baseline

Headache was the most prevalent symptom in the patient cohort. Of 192 patients, 152 (79.2%) had headache, 125 (65.1%) had an occipital headache, and 104 (54.2%) had Valsalva headaches (ie, occipital headaches made worse with coughing and straining). Both overall headache (88.5% [131/148] vs 70.5% [31/44], respectively) and Valsalva headaches (63.5% [94/148] vs 20.5% [9/44], respectively) were more prevalent in the adult cohort compared with the childhood cohort. Figure 2 presents all the patient-reported symptoms in both adults and children.

From the entire cohort, papilledema was detected or known about by the consulting neurosurgeon and/or the referring clinician in 5.2% (10/192), excluded in 48.9% (94/192), and unknown in the remainder.

Of the total baseline cohort, 4.7% (9/192) was found to have scoliosis at the first clinic visit, all of whom were children (20.4% of the childhood cohort [9/44]).

Imaging Findings at Baseline

By the 12-month follow-up, 97.9% (188/192) of the patients had undergone imaging with a brain MRI. The median maximum cerebellar tonsillar descent was 10 (IQR 7-13) mm. At least 80.2% of patients had a CM1 measuring 5 mm or more below McRae's line (measurement unavailable for 16.1%). There was no correlation between the extent of tonsillar herniation and the severity of headache on a visual analog scale (Spearman's Rho = −0.007; $P = .9$).

Syringomyelia was present in 25.2% (37/147 were available for surgeons to review) of patients who underwent spinal MRI. The distribution by vertebral level is shown in Table 2.

Management and Patient Cohorts

The clinical management decisions of the consulting neurosurgeon at the first clinic visit were most frequently reported as nonoperative management (43.2% [83/192]), further investigation

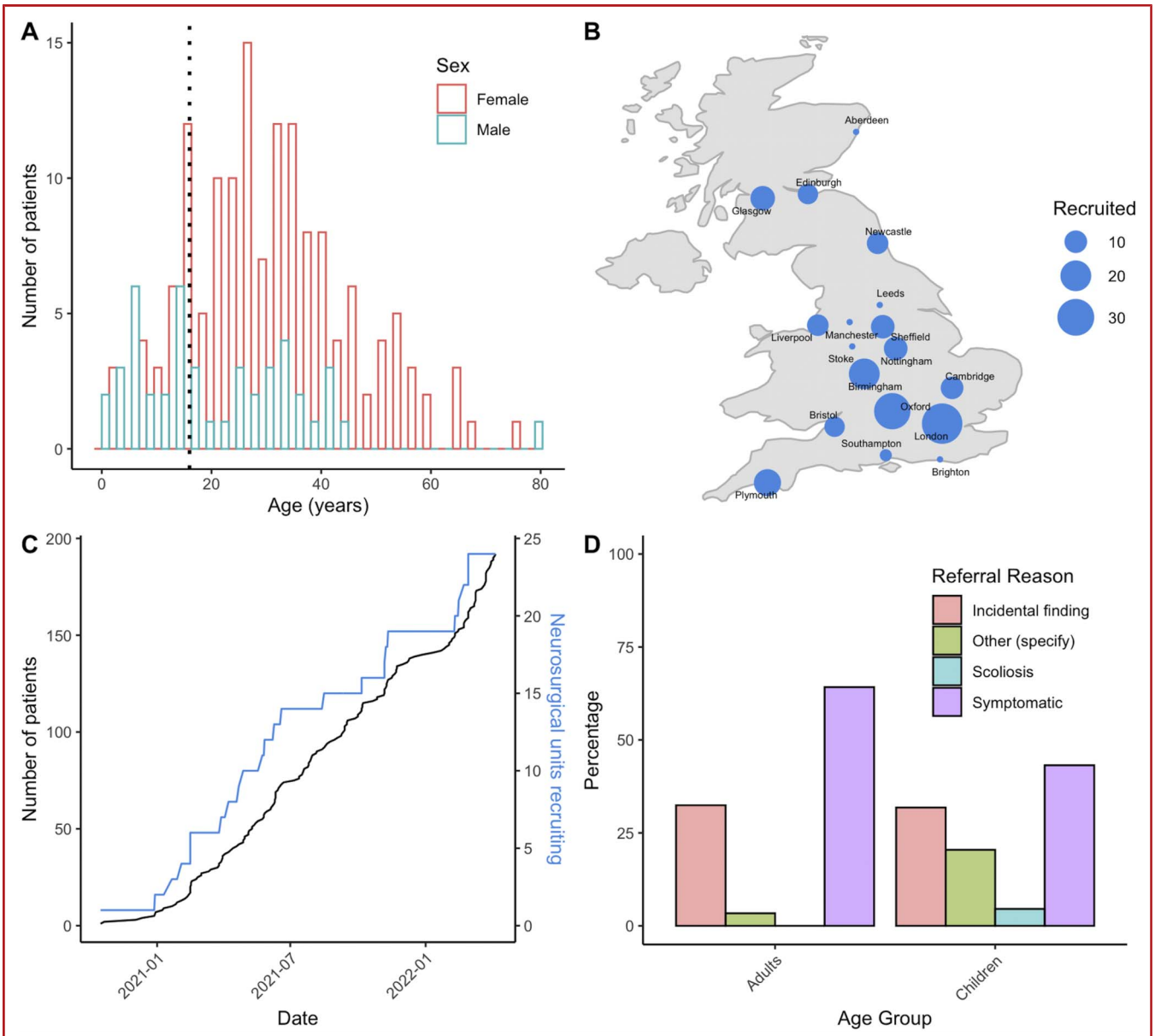

FIGURE 1. *Demographics and recruitment data. Baseline data elements.* **A**, *A histogram of the participants within the UK Chiari 1 Study cohort—including females (red) and males (teal). A dotted vertical line at 16 years shows the age at which participants are categorized as adults rather than children.* **B**, *A UK map with dots (scaled to the number of recruits) at the cities recruiting patients (neurosurgical units within each city have been combined).* **C**, *The number of recruits over the time study period.* **D**, *The reasons for referral in the adult and childhood cohorts.*

(45.3 [87/192]), schedule for FMD (15.1% [29/192]), and referral to a different specialty (13.0% [25/192]). A comparison of management plans at baseline and after 12 months is presented in Figure 3.

An FMD was performed in 12.0% (23/192) of patients before their follow-up survey. The clinical characteristics of this group are included in Tables 1 and 2.

Quality of Life at Baseline

Adults with CM1 at baseline (n = 148) had lower SF36 HRQoL scores across all domains, compared with normative controls ($P < .001$) (Figure 4; Table 3). The adults with CM1 treated without surgery (n = 96) also had lower (worse) SF36 HRQoL scores across all domains, compared with normative controls ($P < .001$).

TABLE 1. Demographics, Referral Sources, and Management Plans for the Baseline Cohort

	UKC1S cohort			Treated with FMD			Treated without surgery[a]					
Factors	**All**	**Adults**	**Children**	**All**	**Adults**	**Children**	**All**	***P*-value [CI]**	**Adults**	***P*-value**	**Children**	***P*-value**
Number of patients	192	148	44	23	19	4	113	—	96	—	17	—
Age in years, median (IQR)	28.2 (16.6 to 38.6)	32.5 (25.8 to 40.8)	11.1 (5.7 to 13.9)	33.7 (18.8 to 44.7)	40.7 (25.3 to 45.3)	9.8 (6.0 to 13.2)	31.3 (21.4 to 39.2)	.9[b] [−7.7 to 7.1]	32.8 (26.1 to 40.9)	.8[b] [−7.6 to 5.8]	8.2 (5.4 to 12.5)	.8[b] [−8.7 to 9.4]
Sex (number)												
Female	146	126	20	20	17	3	92	.8[c] [0.12 to 2.5]	84	1.0[c] [.08 to 4.3]	8	.6[c] [.01 to 4.8]
Male	46	22	24	3	2	1	21		12		9	
Referring clinician (%)												
Neurologist[d]	28.1	33.1	11.4	26.1	31.6	0.0	31.0	.4[c] [NA]	33.3	.10[c] [NA]	17.6	.4[c] [NA]
General Practitioner	27.1	34.5	2.3	34.8	42.1	0.0	28.3		32.3		5.9	
Pediatrician[e]	15.1	2.0	59.1	13.0	5.3	50.0	10.6		1.0		64.7	
Physician	7.3	9.5	0	4.3	5.3	0.0	7.1		8.3			
Orthopedic surgeon	5.7	2.7	15.9	8.7	5.3	25.0	3.5		3.1		5.9	
Ophthalmologist	5.2	5.4	4.5	4.3	0.0	25.0	8.0		8.3		5.9	
Ear, nose & throat surgeon	4.2	4.7	2.3	0.0	0.0	0.0	5.3		6.3		0.0	
Emergency department doctor	3.2	3.4	2.3	4.3	5.3	0.0	1.8		2.1		0.0	
Other/unknown	4.2	3.4	2.3	4.3	5.3	0.0	4.4		5.2		0.0	
Reason for CM1 referral (%)												
Symptomatic	59.6	64.4	43.2	87.0	94.7	50.0	56.6	.4[c] [NA]	61.5	.01[c] [NA]	29.4	.1[c] [NA]
Incidental	32.1	32.2	31.8	4.3	5.3	0.0	35.4		34.4		41.2	
Scoliosis	1.0	0.0	4.5	4.3	0.0	25.0	0.0		0.0		0.0	
Other	7.3	3.4	20.5	4.3	0.0	25.0	8.0		29.4		29.4	
Duration between diagnosis and referral, median (IQR) d [available data points]	27.5 (7.0 to 85.0) [130/192]	33.0 (10.0 to 95.8) [92/148]	18.5 (6.0 to 44.8) [38/44]	19.0 (8.5 to 44.0) [15/23]	19.5 (9.3 to 58.5) [12/19]	19.0 (10.5 to 22.0) [3/4]	31.0 (10.5 to 111.5) [74/113]	.03[b] [19.9 to 423.8]	39.0 (11.5 to 147.8) [60/86]	.04[b] [11.9 to 508.3]	23.0 (10.0 -44.3) [14/17]	.1[b] [−17.1 to 124.5]
Duration between referral and the clinic visit, median (IQR) d [available data points]	82.0 (35.0 to 132.5) [147/192]	90.0 (34.0 to 144.8) [106/148]	81.0 (56.0 to 113.0) [41/44]	54.0 (28.3 to 84.5) [16/23]	42.0 (26.0 to 83.0) [13/19]	66.0 (50.5 to 77.5) [3/4]	101.0 (39.5 to 149.5) [87/113]	<.001[b] [37.3 to 104.7]	110.0 (37·0-174·0) [71/86]	<.001[b] [40.0 to 121.0]	87 (62 to 114) [16/17]	.2[b] [−22.2 to 80.2]

TABLE 1. Continued.

Factors	UKC1S cohort			Treated with FMD			Treated without surgery[a]				
	All	Adults	Children	All	Adults	Children	All	*P*-value [CI]	Adults	*P*-value	Children	*P*-value
Patient-reported family history of CM1 (%)	4.7	3.4	9.1	4.3	5.3	0.0	4.4	1.0[c] [0.02 to 9.4]	3.1	.5[c] [.03 to 22.7]	11.8	1.0[c] [.0 to 24.6]
Coexisting diagnoses known at the first clinic visit (%)												
Migraine	7.3	8.1	4.5	8.7	10.5	0.0	8.0	.4[c] [NA]	8.1	.3[c] [NA]	11.8	1.0[c] [NA]
Anxiety and/or depression	5.2	5.4	4.5	4.3	5.3	0.0	4.4		4.7		5.9	
Asthma	2.6	2.0	4.5	8.7	10.5	0.0	0.9		1.2		0.0	
Other psychiatric conditions	2.6	3.4	0.0	4.3	5.3	0.0	3.5		4.7		0.0	
Fibromyalgia	2.1	2.7	0.0	8.7	10.5	0.0	0.9		1.2		0.0	
Degenerative spinal condition	2.1	2.7	0.0	8.7	10.5	0.0	0.9		1.2		0.0	
Epilepsy	2.1	0.0	9.1	0.0	0.0	0.0	1.8		0.0		11.8	
Idiopathic intracranial hypertension	1.6	2.0	0.0	0.0	0.0	0.0	2.7		3.5		0.0	
Autism	1.6	1.4	2.3	0.0	0.0	0.0	1.8		2.3		0.0	
Cardiac condition	1.6	2.0	0.0	0.0	0.0	0.0	2.7		3.5		0.0	
Hypertension	1.6	2.0	0.0	0.0	0.0	0.0	1.8		2.3		0.0	
Genetic condition	1.0	1.4	0.0	0.0	0.0	0.0	0.9		1.2		0.0	
Connective tissue disorder	1.0	1.4	0.0	0.0	0.0	0.0	0.9		1.2		0.0	
Congenital condition	1.0	0.0	4.5	0.0	0.0	0.0	0.0		0.0		0.0	
Craniosynostosis	0.5	0.0	2.3	0.0	0.0	0.0	0.0		0.0		0.0	
Personality disorder	0.5	0.7	0.0	0.0	0.0	0.0	0.9		1.2		0.0	
Other craniofacial disorders	0.0	0.0	0.0	0.0	0.0	0.0	0.0		0.0		0.0	
Other conditions	28.6	29.7	25.0	34.8	31.6	50.0	31.0		34.9		29.4	

CM1, Chiari 1 malformation; CSF, cerebrospinal fluid; FMD, foramen magnum decompression; UKC1S, UK Chiari 1 Study.
[a]Nonoperative treatment cohort limited to those with 12-month longitudinal data available. Statistical testing was performed to compare the baseline clinical characteristics of the respective groups (all, adults, and children) treated with FMD and without surgery.
[b]Welch 2 sample *t*-test.
[c]Fisher's exact count test. CI = 95% CIs.
[d]Neurologists may be pediatric neurologists.
[e]Management decisions are not mutually exclusive, and multiple options could be selected.

Children with CM1 at presentation (n = 35/44 in age categories to allow group-level analysis) had lower (worse) PedsQL scores across all domains compared with normative controls ($P < .001$) (Figure 4; Table 4). The children with CM1 treated without surgery (n = 13/77 in age categories to allow group-level analysis) did not have significantly different PedsQL scores compared with the normative cohort.

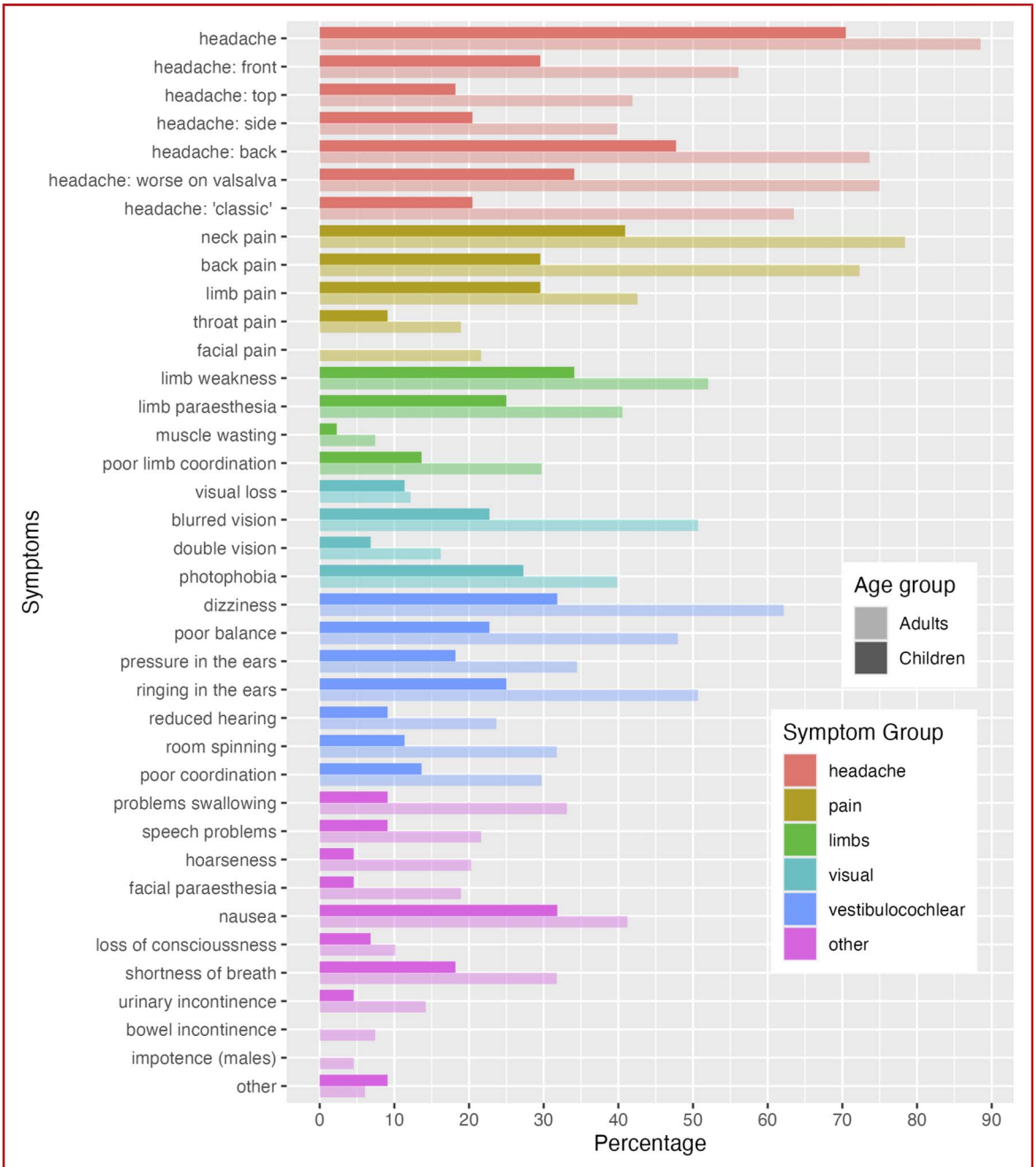

FIGURE 2. *Prevalence of patient-reported symptoms at the first neurosurgical clinic visit in the entire CM1 cohort. Prevalence is given as a percentage (%) on the x-axis. The prevalence of "classic" CM1 headache as a symptom was defined by the prevalence of patients who had a combination of a headache "at the back of the head" that worsened on Valsalva. CM1, Chiari 1 malformation.*

A linear regression analysis of clinical factors traditionally associated with CM1 associated with baseline SF36 quality-of-life subscores showed that both limb pain and neck pain had a significant relationship to 7 of 8 subscores. Back pain, a "classical" CM1 headache, poor balance, poor coordination, the presence of syringomyelia, and a coexisting psychiatric diagnosis had a relationship with low SF36 scores in at least 1 domain (Figure 5).

TABLE 2. Imaging Findings in the Baseline Cohort

	UKC1S cohort	FMD cohort	Nonoperative cohort[a]	Difference nonoperative and FMD cohorts (*P*-value [CI])
Cohort sample size	192	23	113[a]	—
First scan				
First scan modality to diagnose CM1				
CT of the head	16.1%	17.4%	17.7%	.06[b] [NA]
MRI of the head	74.5%	60.9%	76.1%	
Other	8.9%	21.7%	6.2%	
Unknown	0.5%	0.0%	0.0%	
Hydrocephalus on diagnosing scan				
Yes	0.5%	4.3%	0.0%	.03[b] [NA]
No	94.3%	87.0%	98.2%	
Unknown	5.2%	8.7%	1.8%	
First MRI head scan				
MRI of head performed within 12 months of the first clinic visit				
Yes	97.9%	91.3%	100%	.03[b] [0.0 to 1.2]
Maximum cerebellar tonsillar descent median (IQR) (nearest mm) [available cases]	10 (7 to 13) mm [161/192 cases]	11.0 (8 to 15) mm [18/23 cases]	10 (7 to 13) mm [93/113 cases]	0.5[c] [-5.0 to 11.9]
Ventriculomegaly				
Yes	2.1%	4.3%	0.9%	.3[b] [NA]
No	82.3%	73.9%	82.3%	
Unknown	15.6%	21.7%	16.8%	
Syringobulbia				
Yes	2.1%	8.7%	1.8%	0.04[b] [NA]
No	91.7%	78.3%	93.8%	
Unknown	6.3%	13.0%	4.4%	
First spine MRI scan				
MRI of spine performed within 12 months of the first clinic visit				
Yes	80.7%	78.3%	78.8%	.9[b] [NA]
No	17.2%	21.7%	18.6%	
Unknown	2.1%	0.0%	2.7%	
MRI spinal segments performed				
Cervical	74.0%	73.9%	70.8%	1.0[b] [0.4 to 3.9]
Thoracic	64.6%	65.2%	62.8%	1.0[b] [0.4 to 3.3]
Lumbar	59.9%	65.2%	53.9%	.4[b] [0.6 to 4.7]
Sacral	57.3%	65.2%	50.4%	.3[b] [0.7 to 5.4]

TABLE 2. Continued.

	UKC1S cohort	FMD cohort	Nonoperative cohort[a]	Difference nonoperative and FMD cohorts (*P*-value [CI])
Presence of syringomyelia				
Yes	19.3%	56.5%	11.5%	<.001[b] [NA]
No	57.3%	21.7%	61.1%	
Unknown	23.4%	21.7%	27.4%	
Syringomyelia vertebral levels (excluding cases with unavailable or did not undergo spine imaging)				
C1	2.0%	16.7%	0%	<.01 [2.1 to inf.]
C2	12.9%	50.0%	6.1%	<.001[b] [3.5 to 58.9]
C3	13.6%	44.4%	8.5%	<.001[b] [2.2 to 29.8]
C4	15.6%	44.4%	8.5%	<.001[b] [2.2 to 29.8]
C5	17.0%	50.0%	8.5%	<.001[b] [2.7 to 35.4]
C6	19.7%	50.0%	11.0%	<.001[b] [2.2 to 24.9]
C7	19.0%	50.0%	11.0%	<.001[b] [2.2 to 24.9]
T1	16.3%	44.4%	8.5%	<.001[b] [2.2 to 29.8]
T2	15.6%	44.4%	8.5%	<.001[b] [2.2 to 29.8]
T3	14.3%	38.9%	7.3%	<.01[b] [1.9 to 31.4]
T4	13.6%	38.9%	6.1%	<.001[b] [2.2 to 41.6]
T5	13.6%	38.9%	7.3%	<.01[b] [1.9 to 31.4]
T6	12.9%	38.9%	7.3%	<.01[b] [1.9 to 31.4]
T7	12.2%	38.9%	6.1%	<.001[b] [2.2 to 41.6]
T8	9.5%	38.9%	3.7%	<.001[b] [3.2 to 102.4]
T9	9.5%	33.3%	3.7%	<.001[b] [2.4 to 84.8]
T10	8.2%	33.3%	3.7%	<.001[b] [2.4 to 84.8]
T11	6.1%	27.8%	1.2%	<.001[b] [3.1 to 1469.9]
T12	4.1%	22.2%	1.2%	<.01[b] [2.1 to 1162.2]
Other	2.0%	11.1%	0.0%	<.001[b] [0.01 to 0.4]
Other imaging				
Other imaging performed within 12 months of the clinic visit				
MRI flow studies	16.7%	4.3%	15.9%	.2[b] [0.6 to 181.3]
Flexion/extension x-rays	6.8%	8.7%	7.1%	.7[b] [0.1 to 8.3]
CT venography	1.6%	0.0%	2.7%	<.001[b] [0.005 to 0.1]
MRI venography	7.8%	0.0%	9.7%	<.001[b] [0.04 to 0.3]
Other	7.8%	8.7%	4.4%	.3[b] [0.07 to 5.5]

CM1, Chiari 1 malformation; CT, computed tomography; FMD, foramen magnum decompression; UKC1S, UK Chiari 1 Study; Vertebral levels: C, cervical; T, thoracic.
[a]Nonoperative treatment cohort limited to those with 12-mo longitudinal data available. Statistical testing was performed to compare the baseline clinical characteristics of the groups treated with FMD and without surgery.
[b]Fisher's exact count test. CI = 95% CIs.
[c]Welch 2 sample *t*-test.

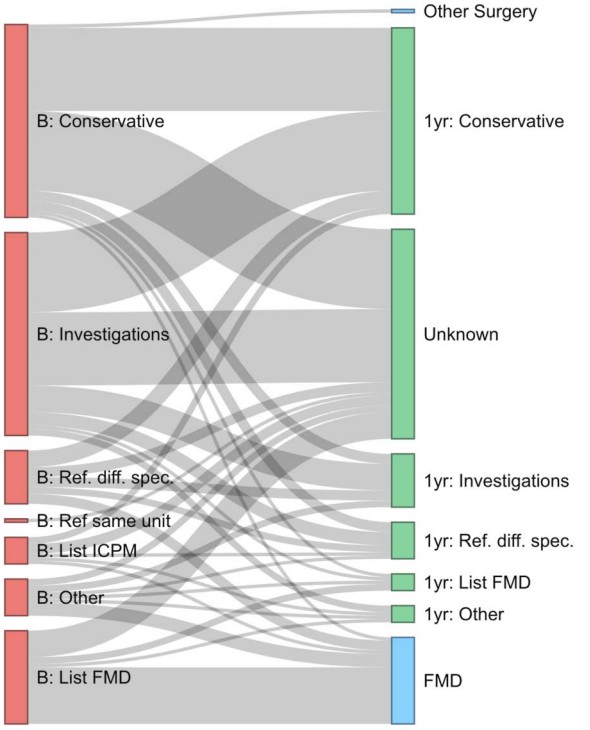

FIGURE 3. *Management plans at the first clinic visit (B = baseline) and a follow-up clinic visit held after 12 months (1 yr). For those patients without a follow-up clinic visit, they are coded as "Unknown". Management decisions at baseline are not mutually exclusive (multiple options could be chosen). FMD, foramen magnum decompression; ICPM, intracranial pressure monitoring; Ref. diff. spec., refer to a different specialty.*

Change in Quality of Life of Those Treated Without Surgery

A total of 138/192 patients in the UKC1S completed the 12-month questionnaire (median 398 (IQR 381-421) days).

In 96/148 adults treated without surgery, there were no significant group-level declines (worsening) in any of the SF36 subscores at 12 months compared with baseline. Two subscores showed a significant improvement: social functioning ($t = -4.0$, $P < .001$) and bodily pain ($t = -2.9$, $P = .03$) (Figure 4; Table 3). There were no significant differences in any of the scores between baseline and 12 months in children with CM1 treated without surgery (n = 13/17 in age categories to allow group-level analysis) (Figure 4; Table 4).

DISCUSSION

CM1 is a common condition and a frequent reason for consultation in the neurosurgical clinic. Many patients are treated without surgery, but, despite this, most studies of CM1 have focused on surgical outcomes or techniques.[5] Very few studies have followed patients managed without surgery or even those waiting for surgery. A systematic review identified only 15 studies of nonoperative management and natural history.[11] Previous studies have reported that some SF36 subscores in adults with CM1 were low preoperatively and improved after FMD,[20] but did not follow a nonoperative cohort. The *UKC1S* was a prospective, multicenter study of patient-reported outcomes of CM1 that has initially studied outcomes after nonsurgical management. This was an observational study and did not make alteration to patient care.

Baseline HRQoL in the whole study group is lower in both adults (SF36) and children (PedsQL) compared with normative data sets. It is crucial for us to understand better the reasons for poor HRQoL at presentation so that we may improve our care for these patients. An analysis of the clinical factors typically associated with CM1 identified that back pain and limb pain were associated with poor HRQoL scores in 7 of 8 SF36 subscores in adults with CM1. Other factors associated with poor HRQoL scores included poor balance, poor coordination, the presence of syringomyelia, and a coexisting psychiatric diagnosis.

This study conducted a longitudinal analysis of HRQoL scores in the patients who were managed by their surgeons without surgery between baseline (first clinic visit) and after 12 months. In the adult cohort, no significant declines in SF36 subscores were observed, and 2 subscores—social functioning and bodily pain—improved. The reasons for improving subscores in adults are unclear, but may relate to counselling, better understanding of the condition, treatment of coexisting conditions (such as primary headache), or other symptom-based treatment. Further research is required to investigate the factors associated with improvement that can be achieved without surgery. In the childhood cohort, no significant declines or improvements were found between baseline and after 12 months. In children, this corroborates previous studies suggesting that the natural history of CM1 in children is more favorable than hitherto believed.[21] Overall, it is a reassuring finding that HRQoL is stable after 12 months; however, a direct comparison with surgical management in this observational study was not possible.

The strengths of this study are a prospective design, a focus on patient-reported outcome measures, and inclusion of the patient group treated without surgery. Furthermore, this is a multicenter study providing wide geographical coverage and recruitment of patients across the United Kingdom.

Limitations

Limitations of the study include that the patient cohort is not a consecutive series, and therefore, the clinical characteristics provided are a description of the cohort rather than a claim on prevalence in the real-world population. Selection bias is a key limitation of this study. This means that the study cannot make conclusions about the epidemiology of the condition or the correct distribution of its clinical features. For example, there were more females in the study, with a female-to-male ratio of 3.2:1. However, a recent review has found that this ratio ranges from 1.3:1 to 4.1:1,[22] and a previous study has suggested that female

patients are more likely to be symptomatic from CM1.[23] Another key limitation and potential bias is the level of dropout from the study, with only 138 of 192 patients (72%) completing the 12-month follow-up. Patients were contacted through telephone when their 12-month follow-up was due, but not all patients opted to complete the follow-up questionnaire. The investigators acknowledge that CM1 is a chronic anomaly, and as such, the 12-month follow-up is a limited snapshot of the natural history. The decision to use this period of follow-up was pragmatic and based on the ability to deliver a timely study using the British Neurosurgical Trainee Research Collaborative model and framework. A total of 92.7% of patients consented to be contacted for future studies, and this might serve as a source of longer-term data in a cohort that has been carefully studied at baseline and 12 months. In addition, although this is the largest prospective UK study of patients with CM1, the study is still limited by a low sample size,

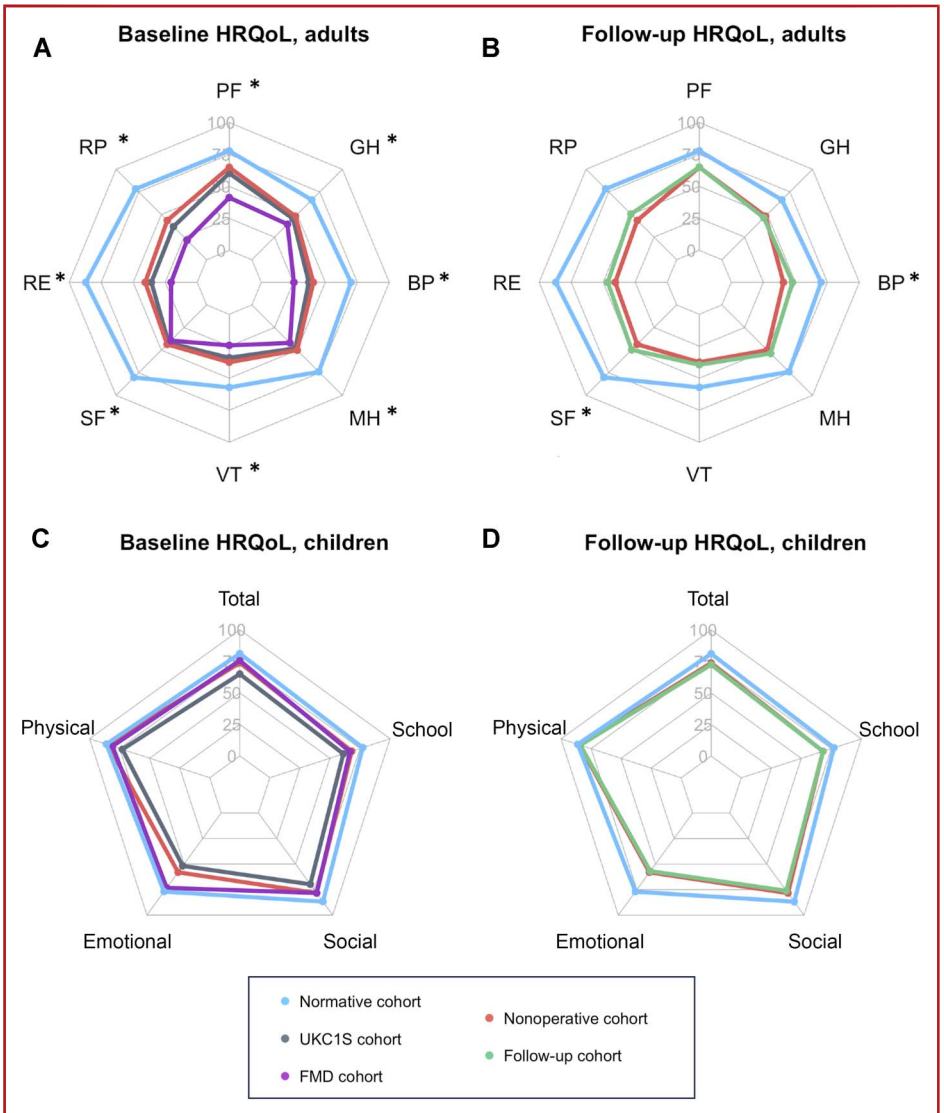

FIGURE 4. *HRQoL, measured with the HRQoL data, in patients with Chiari 1 malformation. Spider plots of each mean health subscore are shown for the normative cohort (blue), the entire UKC1S cohort at baseline (gray), and the nonoperative cohort at baseline (red) and the 12-month follow-up (green). The baseline HRQoL data for the patients treated with FMD are given for reference (purple).* **A** *and* **C**, *Data for the studied cohorts in adults and children, respectively;* **B** *and* **D**, *data for the nonoperative cohort with available 12-month data, for adults and children, respectively. BD, bodily pain; FMD, foramen magnum decompression; GH, general health; HRQoL, health-related quality of life; MH, mental health; PF, physical functioning; RE, role limitations (emotional); RP, role limitations (physical); SF, social functioning; UKC1S UK Chiari 1 Study; VT, energy and vitality.*

TABLE 3. Health-Related Quality-of-Life Data for Adults With CM1 at Baseline (First Neurosurgical Clinic Visit) Compared With a Normative Control Cohort[19]

	Normative controls [available data points][a]	*UKC1S* cohort at baseline [n = 148]	*UKC1S* cohort at baseline vs normative cohort (*t*-value; *P*-value)[b]	FMD cohort at baseline [n = 19/148][c]	Nonoperative cohort at baseline [n = 96/148][c]	Nonoperative vs FMD cohorts at baseline (*t*-value; *P*-value)[b]	Nonoperative cohort at baseline vs normative cohort (*t*-value; *P*-value)[b]	Nonoperative cohort at the 12-mo follow-up [n = 96/148]	Difference between the nonoperative cohort at baseline and follow-up (*t*-value; *P*-value)[d]
SF36: physical functioning (mean SD)	77.8 (30.0) [n = 1308]	60.4 (30.5)	*t* = 6.6; **P < .001**	41.3 (32.1)	64.9 (29.5)	*t* = 3.0; *P* = .1	*t* = 4.1; **P < .001**	65.1 (30.3)	*t* = −0.1, *P* = 1.0
SF36: role limitations (physical) (mean (SD))	78.3 (32.3) [n = 913]	36.6 (38.4)	*t* = 12.5; **P < .001**	21.6 (34.2)	43.4 (40.7)	*t* = 2.5; *P* = .2	*t* = 8.1; **P < .001**	50.4 (40.5)	*t* = −2.1, *P* = .3
SF36: role limitations (emotional) (mean (SD))	87.0 (26.0) [n = 900]	35.8 (36.6)	*t* = 16.4; **P < .001**	20.4 (28.9)	40.6 (37.7)	*t* = 2.6; *P* = .1	*t* = 11.8; **P < .001**	46.5 (39.0)	*t* = −1.7, *P* = .7
SF36: energy and vitality (mean (SD))	57.2 (22.3) [680]	34.0 (23.6)	*t* = 10.9; **P < .001**	24.3 (21.3)	37.5 (24.1)	*t* = 2.4; *P* = .2	*t* = 7.5; **P < .001**	39.5 (23.3)	*t* = −1.1, *P* = 1.0
SF36: mental health (mean (SD))	74.0 (18.9) [n = 761]	48.0 (22.9)	*t* = 13.0; **P < .001**	42.0 (24.2)	50.0 (23.0)	*t* = 1.3; *P* = 1.0	*t* = 9.8; **P < .001**	53.7 (23.4)	*t* = −1.9, *P* = .5
SF36: social functioning (mean (SD))	80.2 (28.1) [n = 514]	41.9 (10.7)	*t* = 25.2; **P < .001**	39.5 (8.3)	43.6 (11.3)	*t* = 1.8; *P* = .6	*t* = 21.6; **P < .001**	49.7 (11.4)	*t* = −4.0, **P < .001**
SF36: bodily pain (mean (SD))	70.1 (32.3) [n = 526]	37.2 (25.0)	*t* = 13.2; **P < .001**	25.5 (22.2)	40.7 (26.1)	*t* = 2.6; *P* = .1	*t* = 9.7; **P < .001**	47.8 (25.9)	*t* = −2.9, **P = .03**
SF36: general health (mean (SD))	66.2 (24.0) [n = 837]	45.4 (23.9)	*t* = 9.7; **P < .001**	39.3 (24.2)	48.1 (24.4)	*t* = 1.5; *P* = 1.0	*t* = 6.9; **P < .001**	46.3 (23.8)	*t* = 1.0, *P* = 1.0

CM1, Chiari 1 malformation; FMD, foramen magnum decompression; SF36, Short-Form 36; UKC1S, UK Chiari 1 Study
[a]Available data points differ between each subscore.
[b]Welch *t*-test (nonpaired).
[c]Only patients with complete 12-month survey data are included.
[d]Paired *t*-test.
P-values are given after correction for multiple comparisons per group.
Bold *P*-values indicate statistical significance.

particularly in the pediatric group. Larger-scale studies are needed, and a registry should be considered to collect more systematic and longer-term follow-up data.[24] Participant numbers were not equal across the recruiting neurosurgical centers (breakdown given Figure 1B and listed in **Supplementary Table 1,** http://links.lww.com/NEU/E970), dependent on study start time per center, and are not a reflection on geographical representation of CM1 across the United Kingdom. Geographical skew might have introduced further bias, with centers recruiting more patients influencing patient selection to a higher degree. An intentional deviation from

TABLE 4. Comparison of Baseline PedsQL Data in Children With CM1 With Normative Healthy Control Children

	Normative controls [n = 665]	*UKC1S* cohort at baseline [n = 35]	*UKC1S* cohort at baseline vs normative cohort (*t*-value; *P*-value)[a]	FMD cohort at baseline [n = 3/35]	Nonoperative cohort at baseline [13/17][b]	Nonoperative vs FMD cohorts at baseline (*t*-value; *P*-value)[a]	Nonoperative cohort at baseline vs normative cohort (*t*-value; *P*-value)[a]	Nonoperative cohort at the 12-mo follow-up [13/17]	Difference in the nonoperative group between baseline and follow-up (*t*-value; *P*-value)[c]
PedsQL: total (mean (SD))	81.1 (13.9)	62.3 (20.8)	$t = 5.3$; **$P = .001$**	75.1 (20.0)	73.8 (17.9)	$t = -0.1$; $P = 1.0$	$t = 1.5$; $P = .8$	72.5 (22.0)	$t = 0.3$, $P = 1.0$
PedsQL: physical (mean (SD))	86.1 (16.1)	62.5 (25.1)	$t = 5.5$; **$P = .02$**	80.9 (21.6)	84.6 (16.9)	$t = -0.3$; $P = 1.0$	$t = 0.3$; $P = 1.0$	83.9 (22.4)	$t = 0.2$, $P = 1.0$
PedsQL: emotional (mean (SD))	80.1 (17.7)	54.6 (23.1)	$t = 5.6$; **$P < 0.001$**	73.3 (22.5)	58.1 (26.7)	$t = -1.0$; $P = 1.0$	$t = 2.5$; $P = .1$	56.9 (28.3)	$t = 0.2$, $P = 1.0$
PedsQL: social (mean (SD))	86.9 (17.2)	71.6 (22.5)	$t = 3.9$; **$P < .001$**	78.3 (18.9)	78.5 (20.9)	$t = 0.01$; $P = 1.0$	$t = 1.4$; $P = .9$	76.2 (21.4)	$t = 0.4$, $P = 1.0$
PedsQL: school (mean (SD))	77.3 (18.5)	60.3 (23.9)	$t = 4.2$; **$P = .004$**	66.7 (30.6)	68.1 (24.5)	$t = 0.1$; $P = 1.0$	$t = 1.3$; $P = 1.0$	68.1 (29.2)	$t = 0.0$, $P = 1.0$

CM1, Chiari 1 malformation; FMD, foramen magnum decompression; PedsQL, Pediatric Quality of Life Inventory; UKC1S, UK Chiari 1 Study.
[a]Welch *t*-test (nonpaired).
[b]Only patients with complete 12-month survey data are included.
[c]Paired *t*-test.
P-values are given after correction for multiple comparisons per group. Six hundred sixty-six of the children in the normative group had returned proxy (parent) forms.
Bold *P*-values indicate statistical significance.

the protocolized statistical analysis plan[12] is the inclusion of follow-up questionnaires completed more than 45 days after the 12-month mark, which was done to avoid a significant dropout and removal of valuable data points, which might have introduced bias. Dividing adult and childhood groups at the age of 16 years is arbitrary but was chosen as a threshold given that 16 years is the age where patients provide their own consent for both clinical decisions and research participation. Finally, the normative data sets were chosen as the most recent and largest-scale UK data sets reporting QOL values but were limited by their geography and date of completion.

This was a pragmatic study, designed to reflect current practice and address certain issues of clinical contention. For example, some neurosurgeons may only diagnose CM1 when there is more than 5 mm of tonsillar herniation below the foramen magnum. However, the definition of CM1 is contentious, and therefore, as detailed in our protocol,[9] a radiological threshold value for the distance of herniation of the cerebellar tonsils through the foramen magnum was not implemented. To sample current practice, the study protocol left the definition of CM1 to the collaborators and included any patients who the attending surgeon considered to have a CM1. There is a potential bias introduced in this approach. However, at least 80% of patients in our study had tonsillar descent of at least 5 mm.

CONCLUSION

This prospective study of patients with CM1 managed without surgery, providing patient-reported outcome measures, showed that there was no decline in HRQoL in adults or children treated without surgery. HRQoL of patients with CM1 remained stable

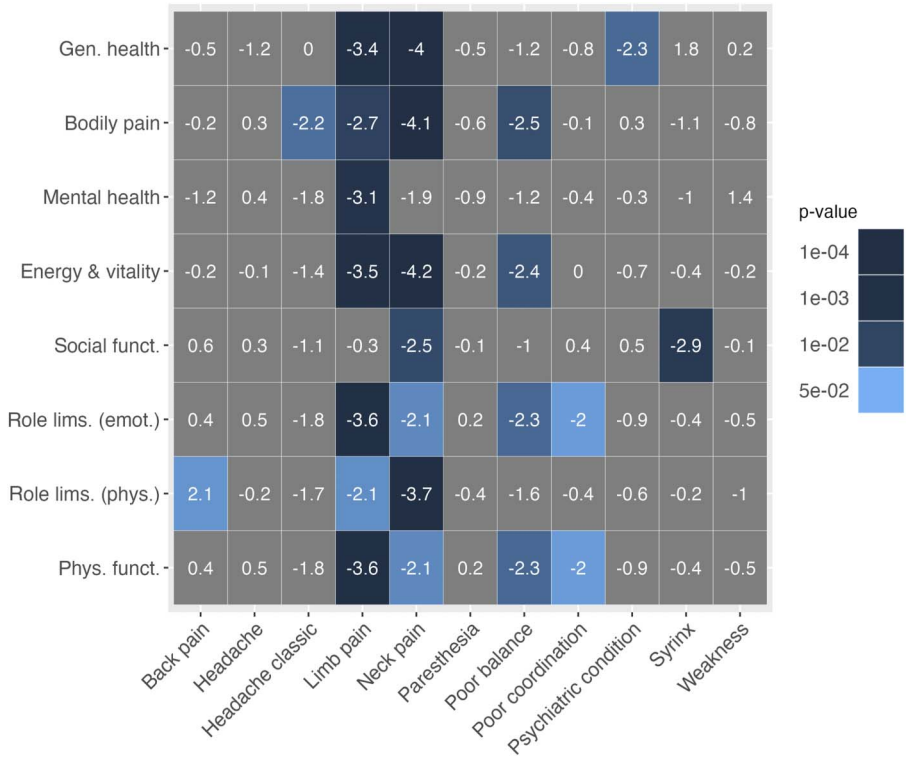

FIGURE 5. *Clinical variables associated with quality of life in the whole UK Chiari 1 Study adult cohort patients with CM1. A linear regression model of clinical variables (x-axis) associated with CM1 with each of the subscores/domains of the Short-Form 36 quality-of-life survey (y-axis) at baseline. The values in each cell are the t-values derived from the general linear model, and the colors are the P-values for the significance, with each association derived from the general linear model. CM1, Chiari 1 malformation.*

12 months after enrollment at the first neurosurgical clinic visit. Adults showed improvement in 2 subscores of HRQoL (social functioning and bodily pain) in this group treated without surgery. Further studies with larger patient populations are needed to make robust comparisons with patients treated surgically.

Funding

This research was supported by the Ann Conroy Trust.

Disclosures

Linda D'Antona received funding from the Academic Clinical Fellowship by NIHR and is a recipient of a research fellowship by B. Braun. Laurence Watkins received funding from Codman Integra and Medtronic and receives funding from B. Braun Aesculap. The other authors have no personal, financial, or institutional interest in any of the drugs, materials, or devices described in this article.

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Acknowledgments

Thank you to the British Neurosurgical Trainee Research Collaborative (BNTRC), the Society of British Neurological Surgeons, and the British Syringomyelia-Chiari Group (Oxford, England, United Kingdom) for their guidance. BNTRC collaborators are named as PubMed-citable/PubMed-searchable collaborators (in alphabetical order, excluding those in the author line). BNTRC Collaborators to be added as PubMed-citable collaborators are as follows: Hazem Abuhusain, Zena Al-Saadi, Andrew Alalade, Amin Andalib, Adriana Baritchii, Pragnesh Bhatt, Stana Bojanic, Andrew Brodbelt, Aabir Chakraborty, David Choi, Shuvamay Chowdhury, Ronan Dardis, Joe M Das, John Duddy, Richard Edwards, Shady Elsayed, Frederick Ewbank, Jonathan Funnell, Pasquale Gallo, John Goodden, Lauren Harris, Ibrahim Jalloh, Greg James, Samuel Jeffery, Edward Jerome St George, Vivek Josan, Chandrasekaran Kaliaperumal, Muhammad Ahmad Kamal, Jothy Kandasamy, Howra Ktayen, Ashwin Kumaria, Simon Lammy, Hugo Layard Horsfall, Paul Leach, Rhannon Lobo, Milan Makwana, Hani Marcus, Joseph Merola, Gabriel Metcalf-Cuenca, Ali Nader-Sephai, Mohammad Naushahi, Mark Nowell, Benedetta Pettorini, Arup Ray, David Rowland, Teresa Scott, Asim Sheikh, Navneet Singh, Saurabh Sinha, James Stewart, Rosa Sun, Dominic Thompson, Lewis Thorne, Ahmed K Toma, Sandhya Trichinopoly Krishna, Kevin Tsang, Georgios Tsermoulas, Ioannis Tsonis, Nikolaos Tzerakis, Micaela Uberti, Siddharth Vankipuram, Babar Vaqas, Alexandros Vyziotis, Ryan Waters, Barrie White, and Ardalan Zolnourian. Author contributions: RJP, FTA, WCS, ED, JE, MDC, MF, SK, MB-C, LD'A, DT, MH, JW, HS, SR, SIH, and AC contributed to data collection, data analysis, and manuscript composition. MZT, SM, GF, NH, AK, LW, RL, WBL, JJ, BSCG, and BNTRC contributed to data analysis and manuscript composition.

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Supplementary Figure 1. Recruitment and exclusion summary for the UK Chiari 1 Study.

COMMENTS

The authors are to be congratulated for a great idea and execution of a nonsurgical study on CMI. That alone is an accomplishment. The UKC1S strength is that it was a prospective study. It was a multicenter study of patient-reported outcomes of CM1 that has initially studied outcomes following nonsurgical management. It was an observational study and did not make alteration to patient care. However, for this, reviewer selection was likely influenced by surgeon bias from diagnosis to completion. However, even with that limitation caveat, I found the conclusions useful for counseling patients with mild symptoms and borderline Chiari morphology.

The authors can say most powerfully that observation at 12 months of CMI patients did not hasten neurological deterioration of those patients with mild Chiari I. This population may have a more benign natural history than surgeons who counsel their patients in detail originally thought. Thus, the common patient question of will my borderline CMI worsen with nonoperative care has been addressed, and the answer is as predicted, likely one will remain stable.

Rich Ellenbogen
Seattle, Washington, USA

In this manuscript, the authors address important issues pertinent to decision-making in the management of patients with Chiari 1 malformations (CM1). They employ the innovative combination of patient- and provider-reported outcomes at 1 year for those who did not undergo surgical treatment. In so doing, they provide valuable quality-of-life data. It is a prospective multicenter observational study that has a number of shortfalls that will be apparent to our readers. Nonetheless, it is these shortfalls that provide strong direction as to what are essential topics to address in future trials as well as the needed follow-up analyses the authors plan to perform. The good discussion of these important issues included increase in the value and validity of this study. The concerns that will propel additional analyses include length of follow-up, the rationale used to decide who initially went to surgery or were followed conservatively, crossovers to surgery, uniformity of imaging at admission and final evaluation, and the large number of dropouts. We look forward to the investigators' next report on their total cohort and to new studies that expand on their approach and findings, improve the thoroughness of future clinical investigations and advance development of clear guidelines for the treatment of CM1.

Cameron Sadegh and James E. Boggan
Sacramento, California, USA

This paper examines the quality of life of patients who present with a radiographic Chiari 1 malformation (CM1). The investigators present a study focusing on the quality of life of adults and children with CM1 managed without surgery in a prospective multicenter study. The authors have recruited a relatively substantial number of patients (275) and ultimately report 12 months of data on 113 who reached the 12-month follow-up. Strengths of this paper include the detailed ascertainment of quality-of-life information, important in the consideration of management of individuals who are found to have radiographic Chiari malformation. Other strengths include its prospective nature and multicenter recruitment. The authors show stability in the quality-of-life domains in adults and children with CM1 after 12 months managed without surgery.

Although the follow-up in this publication is relatively brief for CM1, the authors are to be commended for the contribution of evidence to the understanding of the natural history of the malformation. This reviewer looks forward to the next publications from this group in hopes of learning about the outcomes of the surgical patients and of the longer follow-up for the nonsurgical group.

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