

Functional Abnormalities of the Cervical Cord and Lower Medulla and Their Effect on Pain: Observations in Chronic Pain Patients With Incidental Mild Chiari I Malformation and Moderate to Severe Cervical Cord Compression

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Abstract:

Objective: Abnormalities of central sensory processing may play a role in the pathogenesis of chronic pain. The *Chiari I malformation* is a congenital hindbrain anomaly characterized by protrusion of the cerebellar tonsils into the upper cervical canal, with variable effects on the lower brain stem and cervical cord. The purpose of this study was to compare sensory function and pain among patients with chronic pain who had these disorders incidentally diagnosed, to assess the effect on pain in these patients in comparison with those without central nervous system disease.

Design: Retrospective study in which pain, mood, and sensory function in 32 patients with chronic pain who had mild Chiari I malformation were compared with that in 53 patients with chronic pain who had moderate to severe compression of the cervical spinal cord and 52 patients with chronic pain who had no apparent central nervous system disorder. Data had been collected previously as part of standard clinical assessments, including clinical neurological examinations, quantitative sensory testing, pain drawings, and psychometric testing with the Symptom Checklist 90.

Patients: All subjects were patients of a hospital-based pain management practice who had been accepted for treatment over a 5-year period.

Results: Both the Chiari I and cervical compression groups had long tract signs evident on clinical neurological examination. Quantitative sensory testing indicated elevations in the trigeminal territory among patients with Chiari I malformation and on the neck, hands, and feet in both the Chiari I and cervical compression groups. The extent of pain and mood disturbance was greatest in the Chiari I group and least in the group with no central nervous system disorder. Complex regional pain syndrome, fibromyalgia, and temporal mandibular joint disorder were more common among the Chiari I malformation group than among the other groups.

Conclusions: Quantitative sensory analysis indicates sensory dysfunction associated with Chiari I malformation and cervical cord compression. The pattern of sensory abnormality is consistent with medullary dysfunction among the patients with Chiari I malformation and cervical cord dysfunction among cord compression patients. There were differences in the types and extent of pain and the associated disorders of mood observed among the cohorts defined above. These differences may be partly due to the presence and location of central sensory dysfunction.

Key Words: Cervical cord compression—Chiari I malformation—Complex regional pain syndrome—Fibromyalgia.

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The *Chiari I malformation* is a congenital, or acquired, displacement of the cerebellar tonsils into the upper cervical canal.¹⁻⁴ Symptoms (headache, weakness, numbness, ataxia, vertigo, blurred vision, dysphagia, and

pain) usually occur in adulthood secondary to medullary compression, central cord syndrome, cerebellar dysfunction, and raised intracranial pressure.⁴⁻⁸ There is a consensus that malformations lacking associated brain stem compression or syringomyelia are inconsequential, although few clinical investigations in this area have been performed.^{1,2,4}

Some investigators have unveiled associations between mild Chiari I anomalies and chronic pain. Thimineur et al. found that 5% of patients with complex regional pain syndrome (CRPS) had occult mild Chiari I malformations, a prevalence greater than that estimated in the general population.^{1,9} Studies of patients with fibromyalgia have also shown an increased prevalence of mild Chiari I malformation as well as abnormalities of spinal fluid nociceptive neurotransmitters among such patients.^{10,11} These observations suggest a potential link between mild Chiari I malformation and syndromes of widespread pain and hyperalgesia. However, the mechanisms of such associations remain to be elucidated.

Compression of the cervical spinal cord is well known to cause neurological consequences at and below the level of compression, related to dysfunction of ascending and descending sensory and motor pathways. The possible effects on chronic pain, which may occur as the result of dysfunction of descending nociceptive modulation, have not been specifically investigated. Such an analysis is complicated because many such patients experience pain at the level of compression because of effects on spinal roots, the cord, or structural changes in the spinal column.

MATERIALS AND METHODS

Database

Over a 5-year period, a relational database of clinical measurements (e.g., history, results of neurological examination, and pain diagnosis), quantitative neurological test results, psychometric questionnaire scores, and other clinical testing results had been maintained fastidiously in a single physician's practice (M.T.). During this time, 1,572 patients had been evaluated and accepted for treatment into the pain management program. All data from these patients was entered into the database as initial baseline data or subsequently, as the results of evaluations and testing became available. The study patients and comparison cohorts were from a retrospective database analysis of these 1,572 patients. No new measurements or additional studies were undertaken involving any subject for the purposes of this study.

Study subjects (n = 32)

Analysis of the database disclosed 42 patients with Chiari I malformations. Seven patients had been referred

with a known diagnosis of Chiari I for pain associated with syringomyelia (4 patients), headache (1 patient), facial pain (1 patient), and multifocal pain (1 patient). Five of these patients had undergone suboccipital decompression with cerebrospinal fluid shunts. Diagnoses for the remaining 35 patients were established on the basis of the pain management evaluation. All incidentally diagnosed malformations were mild and were not causative of medullary compression or syringomyelia. Three patients' histories were consistent with possible head injury. The subjects of this study were 32 patients who had mild Chiari I malformations, incidentally diagnosed during pain management evaluation, but had no history of head injury or other apparent central nervous system (CNS) comorbidity.

Comparison cohort 1 (n = 52)

These were randomly selected patients from the same database who met the following criteria.

1. There was no known CNS disease, and pain center evaluation did not uncover the presence of CNS disease.
2. There was no history of head injury.

Comparison cohort 2 (n = 53)

Analysis of the same database disclosed 137 patients with moderate to severe compression of the cervical spinal cord. Of these cases, 66 had been incidentally diagnosed as the result of pain management evaluation, and these patients had not undergone previous cervical spine surgery. Eleven of these patients had a history of possible head injury and two had a history of cerebrovascular accident. The 53 patients with incidentally diagnosed cervical compressions but no history of head injury or other apparent CNS comorbidity were used as comparisons.

Control subjects (n = 25)

These data had been acquired in a previously published study by the first author and concerned individuals who were full time used, were not impaired or disabled, had no acute injuries, and had no history of neurological conditions.¹⁸ No additional measurements were obtained for these individuals for the current study.

Clinical evaluation

Pain syndromes

During the 5 years in which subjects had been evaluated as new patients, the first author used the standards established by the International Association of the Study of Pain and the American College of Rheumatology to diagnose fibromyalgia and CRPS.^{13,14} The diagnosis of

low back pain was established for patients whose primary complaint was of low back pain or low back and leg pain. A large number of patients in the Chiari I group had previously diagnosed temporal mandibular joint dysfunction syndrome. For the purposes of this study, the medical histories of all subjects were retrospectively checked for a prior diagnosis of temporal mandibular joint dysfunction syndrome by a dentist or maxillofacial surgery specialist.

Clinical neurological examination

As part of standard clinical practice for all patients, a neurological examination was performed and the results were entered into the database. This baseline examination included evaluation of cranial nerves, muscle power, reflexes, and sensation. Olfaction and visual acuity were usually omitted. Hearing was assessed with Rinne and Weber tests with a 256-Hz tuning fork. Muscle power was assessed with standard confrontation testing. The sensory examination included pin-prick with a pin; kinesthesia testing, involving asking the patient to identify passive joint movement performed manually at the thumbs and great toes; and testing of vibration sense with a 128-Hz tuning fork at the thumbs and great toes.

Quantitative sensory examination

Thermal sensory thresholds

As part of the standard practice of pain evaluation, all new patients of the first author are tested for quantitative thermal perception thresholds with the protocol described below, and results are entered into the database. This was typically done within 2 months of initial evaluation and acceptance in the pain management program.

Thermal perception threshold measurement is useful in assessing the status of central and peripheral pain and temperature pathways.^{15,16} Warming and cooling perception thresholds were established with a thermal sensory analyzer (TSA-2001; Medoc, Ramat Yishay, Israel), which is capable of maintaining a linear temperature change through a feedback mechanism. A Peltier thermode was placed in contact with the skin at the forehead, maxillae, C4 dermatome, hand, and foot dorsum. Each area was tested bilaterally. Stimulator temperature range was 0°C to 50°C, and the skin adaptation temperature was a constant 32°C.

The method of limits was performed with increasing stimuli, directed from adaptation range toward the sensation range. Subjects were asked to depress a switch (held in the free hand) at the instant they perceived a specific sensation. At each site, five readings were obtained and averaged to determine a single threshold score for each side.

Psychometric studies

The Symptom Checklist 90 (SCL-90) and pain drawing are administered to every patient in the practice at the initial evaluation.

The Symptom Checklist inventory revised

The Symptom Checklist inventory revised (SCL-90R) is a 90-item self-reported inventory of symptoms, designed to reflect the psychological symptom patterns of psychiatric and medical patients. The revised version of the scale was introduced by Derogatis et al.¹⁷ Each item of the scale is rated along a 5-point continuum of distress (0–4), ranging from not at all to extreme. Subjects are asked to rate their level of perceived distress for the preceding 7 days.

The SCL-90R is scored and interpreted in terms of 9 primary symptom dimensions, as follows: (1) somatization; (2) obsessive-compulsive; (3) interpersonal sensitivity; (4) depression; (5) anxiety; (6) hostility; (7) phobic anxiety; (8) paranoid ideation; and (9) psychoticism. Three global indices of distress are also obtained: (1) global severity index, (2) positive symptom distress index, and (3) positive symptom total. Normative values have been obtained from several sample groups, including nonpatients, psychiatric inpatients and outpatients, and pain patients.¹⁸

Pain drawing

Pain drawings have been used extensively in the clinical evaluation and study of chronic pain.^{19–23} Subjects were asked to draw a picture of their pain by shading dorsal and anterior silhouettes of the human body. Pain drawings were scored in three ways: total area of pain, total number of pain sites, and individual sites of pain. The total area of pain was assessed by means of a transparency with a standard grid containing uniform 8 mm × 8 mm cells and counting the total number of cells with any shading. The number of cells with shading was divided by the cell total, to yield the percentage of body pain. The total number of pain sites and the establishment of sites of pain were scored by means of a transparency with nine defined anatomic areas: head, neck, shoulder and arm, thoracic, abdominal, low back, leg, pelvic, and genital. Shading within a defined area was taken to mean the patient had pain in that area of the body.

Statistical analysis

To control for the possibility that differences of age may have confounded group comparison, parametric tests were performed on adjusted means with use of an analysis of covariance model for all continuous variables. Side-to-side variance within groups for quantitative sensory testing measurements was analyzed with use

of the *t* test for dependent samples and was found to be nonsignificant. Post hoc means comparisons were analyzed according to the Tukey test for unequal-size groups. Differences between groups for pain syndromes (CRPS, fibromyalgia, and temporal mandibular joint dysfunction syndrome) were analyzed with a Pearson χ^2 analysis. A Yates-corrected χ^2 analysis was used to compare the prevalence of clinical neurologic signs and painful areas of the body on pain drawings.

RESULTS

Basic demographics

There were significant differences in age between cohorts and a significantly greater fraction of females in the Chiari I group (Table 1). Because we excluded patients with history of possible head injury (such as patients who had a prior motor vehicle accident), there was very little litigation in any group. The fraction of patients receiving workers' compensation was similar in the three groups.

Magnetic resonance imaging

Retrospective review of magnetic resonance imaging studies confirmed that all 32 patients in the Chiari I group met radiologic criteria for mild Chiari I malformation, with tonsillar ectopia ranging from 5 to 10 mm below the foramen magnum. No subject had syringomyelia or brain stem/upper cervical cord compression. One patient had associated occipitalization of the atlas. All 53 of the subjects with cord compression had moderate to severe cord compression due to spondylosis of the cervical spine, disc herniation, or a combination of those. No patient had compression above the C3/4 level.

Spinal comorbidity

One of the patients with Chiari malformation had moderate thoracic spine scoliosis, and another had undergone a lumbar discectomy. Eleven patients with cervical myelopathy had moderate to severe lumbar spine stenosis. Five additional patients with cervical myelopathy had undergone lumbar spine surgery for decompression of stenosis or discectomy. Fifteen of the patients who did not have CNS disease had undergone one or

TABLE 1. Characteristics of the three study groups

Characteristic	Chiari I malformation	Cervical compression	No CNS disease
Age (y), mean	37	55	48
Female (%)	70	45	48
In litigation (%)	3	2	0
Receiving worker's compensation (%)	16	29	23

CNS, central nervous system.

TABLE 2. Clinical diagnoses in the study groups

Study group	Percentage of group with diagnosed condition			
	CRPS	FM	TMJ	LBP
Chiari I malformation	34.4	25.0	28.1	6.3
No CNS disease	1.9	1.9	0.0	44.2
Cervical compression	9.4	5.7	0.0	49.1
<i>p</i> Value	<0.0000	<0.0000	<0.0000	—

CRPS, complex regional pain syndrome; FM, fibromyalgia; LBP, low back pain; TMJ, temporomandibular joint syndrome.

more lumbar spine surgeries for fusion or discectomy, and three additional patients had moderate to severe lumbar stenosis. On the basis of the criterion of previous lumbar surgery or the presence of moderate to severe lumbar stenosis, lumbar spine comorbidity was lowest in the Chiari I group (3%) and comparable in the cervical cord compression (30%) and non-CNS-disease groups (35%).

Pain syndromes and pain drawings

The three groups exhibited distinctly different pain drawings and syndromes (Tables 2 and 3; Fig. 1). The number of painful sites and the extent of the painful area were significantly greater in the Chiari I group than in the other groups and also were significantly greater in the cord compression group versus the non-CNS-disease group. Pain in the head was significantly greater in the Chiari I group ($p < 0.0000$) than in both the other cohorts. Pain in the neck was significantly greater in the Chiari I and cord compression groups than in the non-CNS-disease group ($p < 0.003$ and $p < 0.01$, respectively). The only other significant elevations were more upper extremity and thoracic pain in the Chiari I group than in the non-CNS-disease group.

Despite the negligible amount of lumbar spine disease in the Chiari I group, a higher percentage of these patients indicated pain in the low back and legs than in the comparison groups, although the difference was not significant. Pain was isolated to two or fewer areas on the pain drawing in 60% of the non-CNS-disease comparison group, compared with 30% in the cervical myelopathy group and only 3% of the Chiari I group. Conversely, 77% of the Chiari I group had pain in five or more areas, versus 25% in the cervical myelopathy group and 13% in the non-CNS-disease group.

Temporomandibular joint disorder had been previously diagnosed in 28% of the Chiari I patients, but the diagnosis had never been made in either comparison group. There were far more Chiari I patients who met criteria for CRPS and fibromyalgia than in the other groups. These observations were statistically significant. The one patient in the non-CNS-disease group who had

TABLE 3. Pain drawings

Variable	Chain I (1)	Cord compression (2)	No CNS disease (3)	p value	Comparisons
No. (SD) of painful sites	5.6 (1.2)	3.3 (1.4)	2.4 (1.5)	<0.000000	1 >2,3, <i>p</i> <0.0000, 2 >3, <i>p</i> <0.02
% Painful body area (%)	31	16	9	0.0000	1 >2,3, <i>p</i> <0.0000, 2 >3, <i>p</i> >0.003
Head (%)	84	11	10	0.0000	1 >2,3, <i>p</i> <0.0000
Neck (%)	94	70	27	0.0000	1 >3, <i>p</i> <0.003, 2 >3, <i>p</i> <0.01
Upper extremity (%)	91	53	39	0.0004	1 >3, <i>p</i> <0.003
Lower extremity (%)	88	68	67	NS	NS
Low back (%)	88	79	62	NS	NS
Thoracic (%)	81	43	23	0.0000	1 >3, <i>p</i> <0.004
Abdomen (%)	19	8	6	NS	NS
Pelvic or groin (%)	13	4	8	NS	NS
Genital (%)	3	0	4	NS	NS

Data are percentages of patient groups, except as otherwise indicated. NS, not significant.

met criteria for the CRPS diagnosis was subsequently found to have a locally invasive lymphoma of the affected upper extremity. Although fibromyalgia and CRPS were slightly more frequent in the cervical myelopathy group than in the non-CNS-disease group, the differences did not reach statistical significance.

Clinical examination

Besides trigeminal sensory deficit, cranial nerve abnormalities were present in a small number of Chiari patients (three cases of absent gag reflex and three of exaggerated anisocoria) but were absent in the other groups (Table 4). These observations were not statistically significant. Chiari I patients had significantly more hypoesthesia than both the cord compression and non-CNS-disease groups (*p* <0.05 and *p* <0.0000, respectively), whereas the cord compression group had significantly more than only the non-CNS-disease group (*p* <0.002).

Although variable deficiencies of vibration and position sense were noted in a small number of Chiari I and cervical myelopathy subjects, these observations were not significant. Diminished strength (4+ or less on the 5-point rating scale) in two or more extremities and a positive Hoffman reflex were noted in a high fraction of Chiari I patients and those with cervical myelopathy. These differences in strength, sensation, and reflexes were statistically significant between the Chiari group and the non-CNS-disease group (*p* <0.0000) and between the cord compression group and the non-CNS-disease group (*p* <0.0000) but were not significant between the Chiari and cord compression groups.

Quantitative sensory testing and the Symptom Checklist 90

Thermal perception thresholds were elevated in the trigeminal territory only in the Chiari I patients. Cervical myelopathy and Chiari I patients had comparable eleva-

tion of thermal perception thresholds in the neck, hands, and feet as compared with the non-CNS-disease group and normal comparison group (Table 5; Fig. 3). Mean scores on the SCL-90 were significantly elevated on all scales for the Chiari I subjects versus the other groups (Table 6; Fig. 2), whereas the cervical myelopathy group exhibited less significant elevations than the non-CNS-disease group.

DISCUSSION

The difference in average age among the three cohorts appears easy to reconcile. The Chiari malformation is a congenital malformation that often manifests symptoms in young adulthood, whereas cervical cord compression from degenerative spinal changes is a condition of middle and old age. Through random selection of patients without apparent CNS disorders, we formed a cohort whose average age was between that of the others and very close to the average age of patients with chronic pain in the practice (47 years). The sex difference is not as easy to explain. Within our practice, there are slightly more female patients (52%) than males (48%). It is

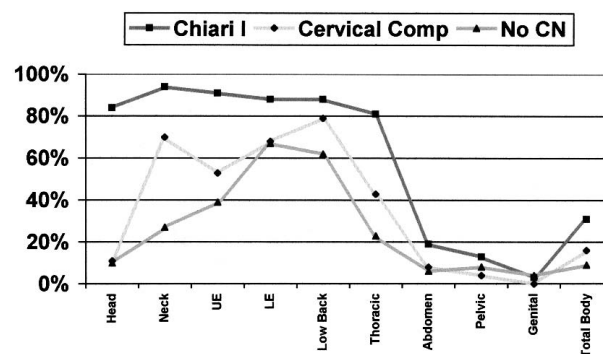


FIG 1. Pain drawing: percentage of subjects indicating pain in these areas. UE and LE, upper and lower extremities, respectively.

TABLE 4. Neurological examination findings (% of patients)

Clinical sign	Chiari I (%)	Cord compression (%)	No CNS disease (%)	Statistics
Absent gag reflex	9.4	0.0	0.0	NS
Anisocoria	9.4	0.0	0.0	NS
Hypesthesia	75.0	34.0	1.9	1 >2, <i>p</i> <0.05, 2 >3, <i>p</i> <0.002, 1 >3, <i>p</i> <0.0000
Proprioception deficits	12.5	11.3	0.0	NS
Decreased strength	75.0	88.7	0.0	1 >3, <i>p</i> <0.0000, 2 >3, <i>p</i> <0.0000
Hoffman reflex	56.3	66.0	3.8	1 >3, <i>p</i> <0.0000, 2 >3, <i>p</i> <0.0000

NS, not significant.

known that the Chiari I malformation is more common in females, and this may account for the additional females in that cohort.

The neurological consequences of moderate to large Chiari I malformations have been well described, whereas those of mild malformations have been less well characterized.^{1-8,24} To understand the possible effects of mild Chiari I malformation and cervical cord compression on pain and sensory alteration, we compared patients with chronic pain who had (1) incidental mild Chiari I malformation, (2) incidental cervical cord compression, or (3) no apparent CNS comorbidity. To assess the effects of mild Chiari I malformation and cervical myelopathy on pain and temperature processing, we compared thermal perception thresholds on the face

and body. Although subjectivity is inevitable in sensory evaluation, psychophysical indicators of deficit can be considered objective if they are anatomically and physiologically consistent.²⁵

The central primary afferent trigeminal pathways subserving pain and temperature descend ipsilaterally into the medulla and upper cervical cord, terminating in the nucleus caudalis, whereas second-order neurons ascend contralaterally to thalamic relays and higher centers.^{26,27} Various abnormalities of the medulla are well known to affect pain and temperature perception on the face as well as the body, because the spinothalamic system is closely allied to these central trigeminal tracts.^{9,26,28} Both the Chiari I and cervical compression patients had long tract signs evident on clinical examination. How-

TABLE 5. Quantitative somatosensory thermotest values

Variable	Side	Mean thermal perception threshold				<i>p</i> value	Group comparison
		Chiari (1)	Cord compression (2)	No CNS disease (3)	Normal subjects (4)		
VI							
Cool	Right	27.4 (3.9)	30.3 (1.9)	30.6 (1.0)	31.0 (0.5)	<0.000000	1 >2,3,4, <i>p</i> <0.000
Cool	Left	27.1 (6.1)	30.7 (1.1)	30.9 (0.8)	30.3 (3.4)	<0.00001	1 <2,3,4, <i>p</i> <0.002
Warm	Right	39.4 (4.7)	34.8 (2.7)	34.5 (1.9)	33.7 (1.4)	<0.000000	1 >2,3,4, <i>p</i> <0.000
Warm	Left	38.7 (4.5)	34.5 (2.4)	34.4 (1.7)	33.9 (1.6)	<0.000000	1 >2,3,4, <i>p</i> <0.000
V2							
Cool	Right	28.7 (3.4)	31.0 (0.6)	30.9 (0.9)	31.3 (0.7)	<0.000000	1 <2,3,4, <i>p</i> <0.000
Cool	Left	28.2 (6.1)	31.0 (0.6)	31.1 (0.6)	31.3 (0.8)	<0.00003	1 >2,3,4, <i>p</i> <0.000
Warm	Right	35.6 (2.8)	33.6 (1.4)	33.4 (1.0)	33.0 (0.7)	<0.000000	1 >2,3,4, <i>p</i> <0.000
Warm	Left	35.4 (3.4)	33.6 (0.8)	33.3 (0.8)	32.9 (0.7)	<0.000000	1 >2,3,4, <i>p</i> <0.000
Cervical							
Cool	Right	25.7 (6.2)	27.8 (5.1)	239.7 (1.7)	30.0 (1.9)	<0.00004	1 >3,4, <i>p</i> <0.001, 3 >4, <i>p</i> <0.001
Cool	Left	25.0 (7.7)	28.4 (2.9)	29.5 (2.7)	30.3 (1.3)	<0.00004	1 >2,3,4, <i>p</i> <0.006
Warm	Right	39.5 (4.4)	37.4 (2.7)	36.3 (2.3)	36.1 (2.0)	<0.000007	1 >2,3,4, <i>p</i> <0.023
Warm	Left	39.8 (4.7)	38.2 (3.9)	36.7 (2.6)	35.7 (2.4)	<0.00001	1 >3,4, <i>p</i> <0.001
Hand							
Cool	Right	26.1 (5.4)	26.0 (5.2)	29.1 (3.6)	29.1 (3.6)	<0.0003	1 >3,4, <i>p</i> <0.027
Cool	Left	25.6 (7.5)	26.5 (5.9)	29.7 (2.0)	30.9 (0.8)	<0.00001	1 <3,4, <i>p</i> <0.004, 2 <3,4, <i>p</i> <0.005
Warm	Right	40.6 (5.0)	41.5 (5.8)	37.4 (3.9)	34.5 (1.5)	<0.000000	1 >3,4, <i>p</i> <0.026, 2 >3,4, <i>p</i> <0.000
Warm	Left	40.3 (4.3)	41.7 (4.9)	37.1 (3.4)	34.3 (2.3)	<0.000000	1 >3,4, <i>p</i> <0.006, 2 >3,4, <i>p</i> <0.000
Foot							
Cool	Right	24.1 (5.4)	22.1 (7.6)	26.9 (6.0)	28.2 (5.1)	<0.0014	2 >3,4, <i>p</i> <0.002
Cool	Left	23.6 (8.2)	20.6 (8.1)	26.9 (6.6)	29.7 (1.6)	<0.00002	1 >4, <i>p</i> <0.009, 2 >3,4, <i>p</i> <0.000
Warm	Right	42.5 (4.4)	44.1 (4.3)	39.7 (4.3)	37.0 (3.2)	<0.000000	1 >3,4, <i>p</i> <0.026, 2 >3,4, <i>p</i> <0.000
Warm	Left	42.4 (3.9)	44.1 (4.0)	39.7 (3.8)	37.0 (3.2)	<0.000000	1,2 >3,4, <i>p</i> <0.000

Values in parentheses are standard deviation.

VI, trigeminal ophthalmic division; V2, trigeminal maxillary division.

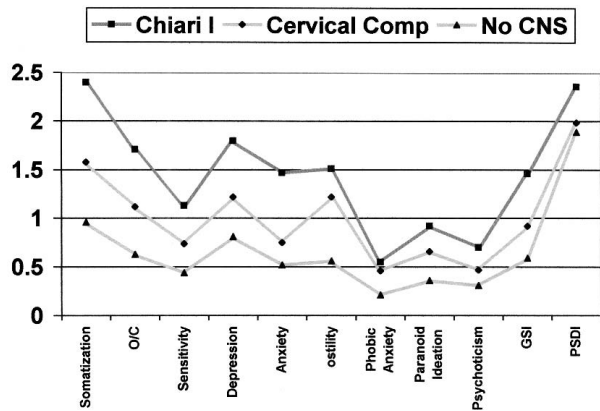


FIG. 2. Symptom Checklist 90 scores. O/C, obsessive/compulsive; GSI, Global Severity Index; PSDI, Positive Symptom Distress Index.

ever, only the Chiari patients exhibited thermohypoesthesia of both the face and body. The simplest explanation would be that the Chiari malformations were causative of dysfunction within the lower brain stem, whereas cord compression was causative of dysfunction within the cervical spinal cord.

There may be several reasonable explanations for expansive pain that was associated with the Chiari I group. Large malformations can cause craniospinal pressure dissociation, resulting in a characteristic headache.⁸ However, since there was no syringomyelia or compression of the upper cervical cord and lower brain stem in any patient, the pathogenesis of head and neck pain likely lies elsewhere. This mechanism would also not explain pains in multiple other areas of the body in these patients. Comparison of pain patterns and sensory measurements in Chiari I patients versus those with cervical myelopathy can provide insight into the pathophysiology of pain in these two groups of patients.

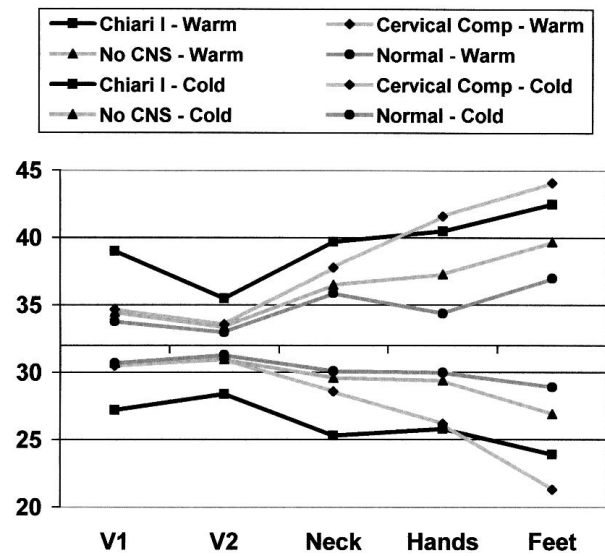


FIG. 3. Somatosensory thermotest. V1, trigeminal ophthalmic division; V2, trigeminal maxillary division.

The rostral ventral medulla (RVM) plays a critical role in the modulation of pain and maintenance of pain threshold. Descending pathways from the RVM project directly to the trigeminal nucleus caudalis and spinal cord dorsal horn.²⁹⁻³³ Chronic hyperalgesia may be determined by an imbalance of inhibitory and facilitatory inputs to the spinal cord dorsal horn and trigeminal nucleus caudalis exerted by neurons originating in the RVM.³⁴ An imbalance of this system has been suggested in studies of fibromyalgia in patients with incidental Chiari I malformations in whom high levels of substance P and low levels of serotonin were noted in spinal fluid assays.¹² As suggested by patterns of thermal hypoesthesia, the Chiari I patients in this study exhibit a picture of sensory abnormality consistent with dysfunction of

TABLE 6. Mean SCL-90 scores

SCL-90 scale parameter	Mean SCL-90 score per group			p value	Group comparison
	Chiari I (1)	Cord compression (2)	No CNS disease (3)		
Somatization	2.24 (0.67)	1.58 (0.59)	0.96 (0.61)	<0.000000	1 >2,3, p <0.02, 2 <3, p <0.02
Obsessive—compulsive	1.71 (1.0)	1.12 (0.85)	0.63 (0.74)	<0.000000	1 >2,3, p <0.02, 2 >3, p <0.02
Interpersonal sensitivity	1.13 (0.84)	0.74 (0.67)	0.44 (0.65)	<0.0011	1 >3, p <0.00
Depression	1.80 (0.91)	1.22 (0.85)	0.81 (0.81)	<0.000027	1 >2,3, p <0.02
Anxiety	1.47 (0.88)	0.75 (0.71)	0.52 (0.61)	<0.000003	1 >2,3, p <0.00
Hostility	1.51 (0.98)	1.22 (1.19)	0.56 (0.72)	<0.000105	1 >2,3, p <0.00
Panic—anxiety	0.55 (0.66)	0.46 (0.82)	0.21 (0.56)	NS	
Paranoid ideation	0.92 (1.0)	0.66 (0.77)	0.36 (0.67)	<0.0205	1 >3, p <0.01
Psychoticism	0.79 (0.67)	0.47 (0.46)	0.31 (0.54)	<0.0082	1 >3, p <0.01
GSI	1.46 (0.73)	0.92 (0.60)	0.59 (0.57)	<0.000001	1 >2,3, p <0.00, 2 >3, p <0.03
PST	53.29 (17.06)	39.83 (19.76)	27.46 (20.82)	<0.000002	1 >2,3, p <0.02, 2 >3, p <0.01
PSDI	2.36 (0.71)	1.99 (0.59)	1.89 (0.60)	<0.021377	1 >3, p <0.01

Values in parentheses are standard deviation.

GSI, Global Severity Index; NS, not significant; PSDI, Positive Symptom Distress Index; PST, Positive Symptom Total; SCL, Symptom Checklist.

medullary sensory pathways. It would be reasonable to suggest that medullary antinociceptive functions are affected and that descending inhibitory inputs to the dorsal horn and trigeminal nucleus are impaired. A consequence of abnormal RVM-mediated pain modulation may be a tendency toward widespread hyperalgesia and persistent pain, which would logically include the trigeminal territory.

In contrast, compression of the cervical spinal cord should affect only the axonal tracts of the sensory system, at and below the compression. It can be assumed that descending antinociceptive projections from the RVM, which travel in the dorsolateral funiculus, may be adversely affected. In comparison with the situation of medullary dysfunction, the relative effect of cervical cord dysfunction on the descending antinociceptive system may be less profound, and the regulation of the trigeminal system should be spared.

The pain drawings of these patients may reflect this. There was very little pain in the trigeminal territory and a surprising amount of pain in territories well below the level of cord compression (thoracic, low back, and leg pain).

Complex regional pain syndrome and fibromyalgia are pain syndromes of expansive hyperalgesia and autonomic disturbance. Although many diverse pathologies and mechanisms may underlie these syndromes, there is growing evidence of a supraspinal origin for the associated hyperalgesia and autonomic abnormalities.^{9,11,12,35-40} Observations in this study suggest that subtle dysfunction of lower brain stem processes and perhaps also dysfunction of the cervical cord may be factors that predispose to the pains and symptoms of CRPS or fibromyalgia. Likewise, observations by some investigators that the syndrome of temporomandibular joint dysfunction syndrome is frequently accompanied by pain outside the head are perhaps due to a more widespread problem with sensory processing.⁴¹

This study provides evidence that anatomic abnormalities of the cervical spinal canal and hindbrain can influence the extent and character of pain in patients with chronic pain. The prevalence of Chiari I malformation in this population is less than 1 in 200, whereas cervical cord compression is present in 7% of individuals greater than 40 years of age.^{2,42} Although mild malformations are often considered inconsequential, this investigation suggests that some cases may be associated with dysfunction of medullary sensory processing.

As such, the expansive pain and hyperalgesia in such patients may derive from a disturbance of descending pain modulation from the brain stem. Abnormal integration of autonomic responses to nociceptive input is also likely, since autonomic outflow abnormalities have been

associated with the Chiari I malformation.²⁴ Although less clear from this work, dysfunction of descending pain modulation may be an important if not obvious consequence of cervical cord compression. Clearly, these issues need further study to uncover the mechanisms of our observations.

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