Operative treatment of anterior thoracic spinal cord herniation

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Operative Treatment of Anterior Thoracic Spinal Cord Herniation: Three New Cases and an Individual Patient Data Meta-analysis of 126 Case Reports

OBJECTIVE: Anterior thoracic spinal cord herniation is a rare cause of progressive myelopathy. Much has been speculated about the best operative treatment. However, no evidence in favor of any of the promoted techniques is available to date. Therefore, we decided to analyze treatment procedures and treatment outcomes of anterior thoracic spinal cord herniation to identify those factors that determine postoperative outcome.

METHODS: An individual patient data meta-analysis was conducted, focusing on age, gender, vertebral segment of herniation, preoperative neurological status, operative interval, operative findings, operative techniques, intraoperative neurophysiological monitoring, postoperative imaging, neurological outcome and follow-up. Three cases from our own institution were added to the material collected. Bivariate analysis tests and multivariate logistic regression tests were used so as to define which variables were associated with outcome after surgical treatment of anterior thoracic spinal cord herniation.

RESULTS: Brown-Séquard syndrome and release of the herniated spinal cord appeared to be strong independent factors, associated with favorable postoperative outcome. Widening of the dura defect is associated with the highest prevalence of postoperative motor function improvement when compared with the application of an anterior dura patch ($P < 0.036$).

CONCLUSION: Most patients with anterior thoracic spinal cord herniation require operative treatment because of progressive myelopathy. Patients with Brown-Séquard syndrome have a better prognosis with respect to postoperative motor function improvement. In this review, spinal cord release and subsequent widening of the dura defect were associated with the highest prevalence of motor function improvement. D-wave recording can be a very useful tool for the surgeon during operative treatment of this disorder.

KEY WORDS: Anterior thoracic spinal cord herniation, Brown-Séquard syndrome, Cord release, D waves, Dura widening, Follow-up, Intraoperative neurophysiological monitoring

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Anterior thoracic spinal cord herniation (ATSCH) is a rare cause of progressive myelopathy. Since the introduction of magnetic resonance imaging (MRI), ATSCH has been increasingly recognized and reported in the international medical literature. It is characterized by herniation of the thoracic spinal cord through an anterior or anterolateral dura defect. Most patients present with a gradually progressive sensorimotor Brown-Séquard-like syndrome, which may evolve into a severe paraparesis (56). Because of its rarity, clinical experience with ATSCH within a single institution is very limited. As a result, treatment

ABBREVIATIONS: ADP, anterior dura patch; **ATSCH**, anterior thoracic spinal cord herniation; **CI**, confidence interval; **CR**, cord release; **CSF**, cerebrospinal fluid; **CTM**, computed tomographic myelography; **DS**, direct suture; **eMEP**, epidural motor evoked potential; **IONM**, intraoperative neurophysiological monitoring; **IPD**, individual participant data; **mMEP**, muscular motor evoked potential; **MRI**, magnetic resonance imaging; **PAC**, posterior arachnoid cyst; **SSEP**, somatosensory evoked potential; **WDD**, widening of the dura defect

strategies are based on individual cases and reports in the literature, or they result from ad hoc decisions during operative exploration of preoperatively unrecognized cases. Surgical treatment of spinal cord herniation is usually followed by stabilization or improvement in the neurological symptomatology, but unfavorable postoperative outcomes are also reported (4, 9, 14, 24). During the operation, release of the herniated part of the spinal cord and subsequent repositioning of the cord into the normal anatomic position is advocated as essential (10). To prevent reherniation/reincarceration of the spinal cord, enlargement of the ventral dura defect has been promoted by several authors (49, 51, 58, 61, 68, 76, 85, 86). Others strongly advocate closure of the defect by direct suturing (15, 34, 42, 86, 89) or by insertion of a patch (8, 10, 17, 22, 24, 25, 43–45, 47, 48, 52, 53, 55, 56, 72, 78, 79). However, the literature has not provided evidence in favor of any of these strategies from either controlled studies or meta-analysis. To remedy this, we performed a meta-analysis using the case report data for 126 individual patients described in the international medical literature and added to this sample 3 patients who were treated in our own department. To identify those factors that determine postoperative outcome, special attention was paid to neurological presentation, interval until diagnosis, operative findings and technique, including intraoperative neurophysiological monitoring (IONM), and postoperative imaging findings and neurological outcome.

ILLUSTRATIVE CASES

Patient 1

A 42-year-old man presented with a 5-year history of numbness and burning sensation in the right foot and lower leg, followed in subsequent years by similar sensations in the left leg and dysesthesia on the left side of his abdomen. Numbness in both legs progressed, with a sensory level at the T12 dermatome. For 2 years, he was unable to run or take part in sports because of weakness and clumsiness in the left leg. In addition, a change in the pattern of defecation and micturition was noted. On MRI, a posterior arachnoid cyst (PAC) with spinal cord compression was suspected, and an exploratory laminectomy was performed in another hospital. Postoperatively, the neurological condition of the patient continued to deteriorate. At the time of presentation at our department, an incomplete Brown-Séquard syndrome was present, with weakness of the left leg (hamstring and lower leg muscles Medical Research Council Grade 4), a decrease in pain and temperature sensation below T7–T8, and a decrease in vibration sensation in both legs. MRI showed anterior displacement of the spinal cord at T5–T6, with anterior transdural cord herniation (Fig. 1A). An operation was performed, using IONM, recording somatosensory evoked potentials (SSEPs) and using transcranial electrical motor cortex stimulation to elicit muscular and epidural motor evoked potentials (mMEPs and eMEPs). Laminectomy was performed, and intradural exploration of the spinal cord revealed cord herniation through a ventral dural defect. The spinal cord was released microsurgically, and the herniated part of the cord was redressed intradurally. During the final part of release of the very adherent cord, mMEPs in the lower extremities were lost. At the same time, D-wave amplitude of the distal eMEP decreased dramatically to 30% from baseline, whereas the proximal eMEP amplitude and latency time remained unchanged. The latency time was increased

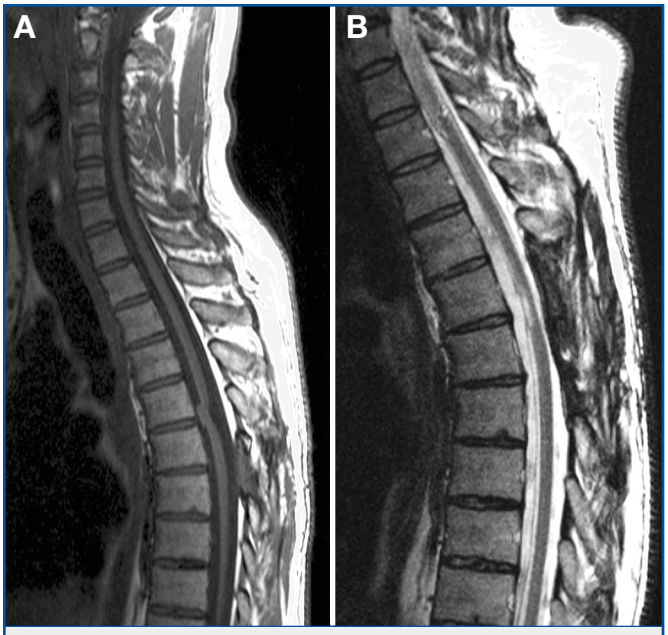

FIGURE 1. A, *preoperative sagittal T1-weighted unenhanced magnetic resonance imaging (MRI) scan showing anterior displacement of the spinal cord at the T5–T6 vertebral level, with anterior transdural cord herniation.* **B**, *postoperative sagittal T2-weighted MRI scan of the same area showing realignment of the spinal cord, with some atrophy and myelopathic changes at the site of previous herniation.*

by 25% over the cord area between both spinal epidural electrodes. After a pause, the amplitude of the distally recorded D waves returned to 60% from baseline level, and at the end of the monitoring, the latency time recovered slightly to 15%. A GORE-TEX sleeve was inserted to cover the dura defect, ventral to the spinal cord, to prevent reherniation. The patch was held in place with stay sutures on both sides. After the patch was brought into place, mMEPs returned in both lower legs and in the quadriceps muscle on the right. SSEPs remained stable during the operation. Immediately after surgery, the patient experienced diffuse paresis of his left leg (MRC Grade 4). Three months after the operation, sensory disturbances had improved, and defecation and micturition had normalized. Postoperative MRI showed realignment of the spinal cord and cord atrophy with myelopathic changes at the site of previous herniation (T5–T6) (Fig. 1B). At 1-year follow-up, improvement in motor function of the proximal left leg was noted. According to the patient, functionality of the left leg was judged to be worse than before the operation. One year later, the patient returned, reporting an increase in the sensory deficit and a slight increase in the paresis of the left lower leg. This was confirmed after neurological examination (sensory level at T9 on the right side and at T4 on the left side and an increase in the paresis of the flexors and extensors [MRC Grade 3–4] of the left foot). MRI did not reveal any changes. Neurological examination 30 months after the second operation was stable.

Patient 2

This 68-year-old man had a 5-year history of paresthesia in the left leg and later in the right leg as well. He gradually noticed stiffness in both legs and problems with walking. Later on, he experienced intermittent, uncontrollable spasms in both legs and, finally, problems with bladder

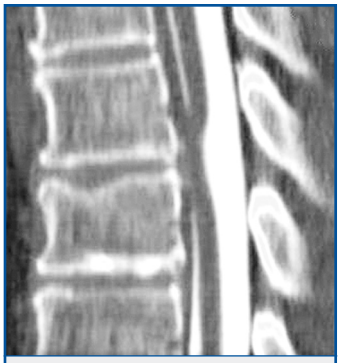

FIGURE 2. *Preoperative sagittal reconstruction of computed tomographic myelography scan, with interruption of the contrast line at the site of cord herniation, anterior to the cord and posterior to the T6–T7 intervertebral disc space.*

control. Neurological examination revealed hyperreflexia in both legs, with ankle clonus and Babinski's sign on the left side along with numbness of the left upper leg. Sequential computed tomography myelography (CTM) and MRI confirmed anterior spinal cord herniation at T7–T8 and ruled out the presence of a PAC (Fig. 2). Laminectomy T7–T9 was performed with the use of IONM. An anterolateral transdural herniation of the spinal cord was found (Fig. 3A). The herniated cord was released from the dura edges and redressed intradurally, and a GORE-TEX sleeve was inserted as previously described (Fig. 3B). No events occurred during IONM. Postoperatively, this patient made a very quick recovery, with improvement in motor function of the left leg, improvement in sensory disturbances, disappearance of spasms, and relief of urinary dysfunction. Because of cardiac dysrhythmia, a pacemaker had been inserted, which frustrated postoperative evaluation with MRI. Because neurological improvement was evident, the patient refused postoperative CTM. Ten months after operation, his neurological condition is stable.

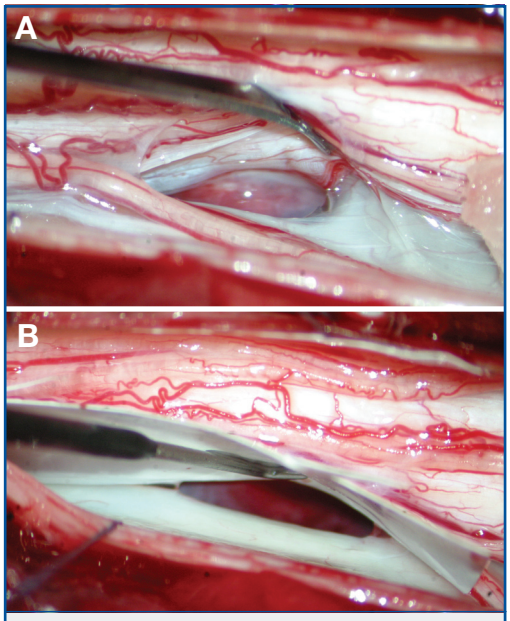

FIGURE 3. *Intraoperative photographs showing herniation of the spinal cord through an oval anterior dural defect at T7–T8 (**A**) and the situation after microsurgical release of the spinal cord and reduction of cord herniation (**B**). A Gore-Tex sleeve is inserted ventral to the spinal cord, covering the dura defect, in order to prevent reherniation.*

Patient 3

The details of our third patient are listed in Table 1.

MATERIAL AND METHODS

Search Strategy

To identify relevant literature, computerized databases of MEDLINE, PubMed, EMBASE, and ISI Web of Science were searched, using the search terms "idiopathic spinal cord herniation" and "spontaneous spinal cord herniation." In addition, the reference lists of the selected papers were checked for related articles. Articles in foreign languages were translated and checked for the subject, and, when necessary, the authors were contacted to provide additional information. This search resulted in a total of 83 potentially relevant papers.

Inclusion and Exclusion of Publications

Nine papers were excluded from this study (postoperative or traumatic cases, insufficient documentation, and double publication). From the remaining 74 articles, a total of 126 cases of ATSCH were identified and were included in this analysis. Age, sex, time interval between onset of symptoms and establishment of the diagnosis and operation (operative interval), vertebral segment of cord herniation, imaging studies (pre- and postoperative), surgical approach, surgical findings, surgical technique, IONM, and postoperative result were analyzed. In almost all the papers, detailed motor scores were missing. As a result, standardized classification of functionality was not possible. We decided to use the motor function (if applicable) for scaling postoperative outcome (improved, unchanged, or worse) because motor dysfunction in general has the greatest impact on the patient in his or her daily life. All relevant data for the 126 cases from the literature and for our own 3 patients are listed in Table 1.

Study Design

The design of the study is that of a meta-analysis done by collecting the complete set of raw data from case reports across all the studies concerning all the variables of interest (including potential confounding variables and censoring information, where appropriate), which is known as individual participant (sometimes patient) data (IPD) meta-analysis (90).

Data Collection

Data were extracted from the publications included using a data extraction sheet developed and piloted by the medical review team. In this data extraction sheet, all relevant patient characteristics and clinical data were defined and used to assess each eligible case report. All study variables are shown in Table 1.

Statistical Analysis

Patient characteristics, stratified by surgical treatment result, were compared with the χ^2 test (Fisher's exact test when appropriate) and the difference of proportions test (59) for nominal variables, which are presented as numbers and percentages. For differences in proportions, Cohen's effect size statistic w was used with a threshold of less than 0.10 for trivial, greater than 0.10 to less than 0.30 for small, greater than 0.30 to less than 0.50 for medium, and greater than 0.50 for large differences (19). All statistical tests were 2 tailed. A P value of less than 0.05 was used for all tests to indicate statistical significance.

A multivariate logistic regression was used to define the independent association between postoperative improvement and nonimprovement

TABLE 1. Relevant data for cases of anterior thoracic spinal cord herniation[a]

Case no.	Series (ref. no.)	Age (y)/sex	Interval (y)	Symptoms	Level	CTM/MRI	Surgical finding	Surgical technique	IONM	Operative result	MRI	FU (mo)
1	Abe et al. (1)	58/M	4	B-S	T8–T9	+/+	VDD, SCH	CR, WDD		imprSM	RC	36
2	Aizawa et al. (3)	44/M	5	B-S	T8–T9	+/+	DDM, SCH	CR, WDD		imprM, unchS	RC	12
3	Aizawa et al. (3)	60/F	3	B-S	T4–T5	+/+	DDM, SCH	CR, WDD		imprM, unchS	RC	12
4	Aizawa et al. (3)	59/F	20	SM para	T4–T5	+/+	VDD, SCH	CR, WDD		imprM, unchS	RC	12
5	Ammar et al. (4)	50/F	1	B-S	T7–T8	−/+	VDD, SCH	CR, WDD		imprM, unchS	RC	84
6	Aquilina et al. (5)	37/F	1	SM para	T4–T5	−/+	VDD, SCH	CR, ADP		imprSM	RC	3
7	Arts et al. (6)	58/F	?	B-S	T7–T8	−/+	VDD, SCH	CR, ADP		Unchanged		
8	Arts et al. (6)	43/M	?	B-S	T4–T5	−/+	1PAC/2VDD, SCH	1NE/2CR, ADP		imprSM	RC	
9	Bandai et al. (7)	63/F	5	SM para	T2–T3	+/+	DDM, SCH	CR, ADP		imprSM		
10	Barbagallo et al. (8)	28/F	5	SM para	T6	−/+	VDD, SCH	CR, ADP		Unchanged	RC	6
11	Barbagallo et al. (8)	64/M	10	SM para	T6	−/+	BD, VDD, SCH	BDF, CR, ADP		Worse		
12	Barrenechea et al. (9)	65/F	3	B-S	T4–T5	−/+	1PAC/2VDD, SCH	1Cyst/2CR, ADP	SSEP, mMEP	Unchanged		60
13	Barrenechea et al. (9)	32/M	1	SM para	T7–T8	−/+	1PAC/2VDD, SCH	1Cyst/2CR, ADP	SSEP, mMEP	Worse		144
14	Barrenechea et al. (9)	54/F	6,5	B-S	T2–T3	−/+	1PAC/2VDD, SCH	1Cyst/2CR, ADP	SSEP, mMEP+ eMEP	Unchanged		16
15	Barrenechea et al. (9)	60/F	2,5	B-S	T2–T3	−/+	VDD, SCH	CR, ADP		imprSM		42
16	Barrenechea et al. (9)	59/F	1,5	B-S	T5–T6	−/+	VDD, SCH	CR, ADP	SSEP, mMEP	Worse		40
17	Barrenechea et al. (9)	34/M	5	SM para	T7–T8	−/+	VDD, SCH	CR, ADP		Unchanged		32
18	Barrenechea et al. (9)	72/M	5	B-S	T4–T5	−/+	VDD, SCH	CR, ADP	SSEP, mMEP	Unchanged		10
19	Bartolomei et al. (10)	61/F	10	B-S	T3–T4	+/+	VDD, SCH	CR, ADP		imprM, unchS	RC	3
20	Batzdorf (11)	23/F	2	B-S	T6	?	1PAC/2VDD, SCH	1Cyst/2CR, ADP		imprSM		
21	Baur et al. (12)	66/F	7	B-S	T10	+/+	VDD, SCH	CR, DS		imprSM	RC	
22	Berbel et al. (13)	56/M	?	B-S	T5–T6	−/+	PAC, VDD, SCH	NE		Unchanged		1
23	Bode et al. (14)	60/F	2	SM para	T8	+/+	VDD, SCH	CR, ADP		Worse	RC	60
24	Bode et al. (14)	33/F	4	SM para	T5	+/+	VDD, SCH	1CR, ADP/2CR		Unchanged		3
25	Borges et al. (15)	68/F	12	B-S	T7–T8	−/+	VDD, SCH	CR, DS		imprSM		12
26	Borges et al. (15)	68/M	8	B-S	T2–T3	−/+	VDD, SCH	CR, ADP		imprSM		
27	Borges et al. (15)	48/F	9	B-S	T7	−/+	VDD, SCH	CR, DS		imprM, unchS		2
28	Brugieres et al. (16)	54/F	5	B-S	T6	−/+	VDD, SCH	CB, CR, DS		imprSM		
29	Brugieres et al. (16)	70/M	0,5	S bilat	T5–T6	−/+	DDM, SCH	CR, DS		Improved	RC	
30	Cellerini et al. (17)	53/M	1	B-S	T8–T9	−/+	VDD, SCH	CR, ADP		imprM, unchS		3
31	Cellerini et al. (17)	37/F	0,5	B-S	T4–T5	−/+	VDD, SCH	CR, ADP		imprSM		3
32	Darbar et al. (20)	41/M	3	B-S	T5	−/+	VDD, SCH	CR, DS		imprSM		1

Continues

TABLE 1. Continued

Case no.	Series (ref. no.)	Age (y)/sex	Interval (y)	Symptoms	Level	CTM/MRI	Surgical finding	Surgical technique	IONM	Operative result	MRI	FU (mo)
33	Darbar et al. (20)	63/F	?	B-S	T4–T6	−/+	1PAC/2VDD, SCH	1Cyst/2CR, ADP		imprM, unchS		
34	Darbar et al. (20)	34/F	5	SM para	T7–T8	Myelo	VDD, SCH	CR, ADP		imprM		
35	Dietemann et al. (21)	68/M	?	SM para	T5–T6	+/+	VDD, SCH	CR, ?		?		
36	Dix et al. (22)	44/F	?	B-S	T7–T8	+/+	VDD, SCH	CR, ADP		imprSM		6
37	Ellger et al. (23)	59/f	2,5	B-S	T2–T3	+/+	VDD, SCH	CR, DS		imprSM	RC	4
38	Ewald et al. (24, 25)	51/F	2	B-S	T5–T6	+/+	VDD, SCH	1CR, ADP/2NE		Worse	RC	2
39	Ewald et al. (25)	46/M	3	B-S	T2	−/+	VDD, SCH	CR, ADP		Unchanged		
40	Ewald et al. (25)	50/F	8	SM para	T6	−/+	VDD, SCH	CR, ADP		Unchanged		
41	Ferré et al. (26)	70/M	1,5	SM para	T10–T11	−/+	VDD, SCH	CR, ADP		imprSM	PAD	24
42	Ferré et al. (26)	75/F	1	B-S	T5–T6	−/+	VDD, SCH	CR, WDD		Worse	RC	12
43	Francis et al. (27)	28/F	1,5	B-S	T6	−/+	VDD, SCH	CR, WDD		imprSM	RC	
44	Gwinn and Henderson (30)	47/F	6	B-S	T7–T8	−/+	VDD, SCH	CR, ADP	SSEP	imprSM	RC	3
45	Gwinn and Henderson (30)	51/F	?	B-S	T8	−/+	VDD, SCH	CR, ADP	SSEP	imprSM	PAD	3
46	Gwinn and Henderson (30)	55/M	0,5	B-S	T6–T7	−/+	VDD, SCH	CR, ADP		imprSM		3
47	Hausmann and Moseley (32)	58/F	8	B-S	T6	+/+	BD, VDD, SCH	BDF, CR, ADP		imprSM		
48	Hausmann and Moseley (32)	36/M	7	SM para	T6–T7	+/+	SCT	CB		Worse	RC	
49	van den Hauwe et al. (83)	65/F	?	B-S	T5–T6	+/+	PAC, VDD, SCH	CR, ADP		?		
50	Inoue et al. (34)	21/M	2	B-S	T3	+/+	VDD, SCH	CR, DS		imprSM		24
51	Isu et al. (35)	43/F	1	B-S	T5–T6	+/+	PAC	Cyst		unchM, imprS		
52	Isu et al. (35)	45/F	2	SM para	T2–T3	+/+	PAC	Cyst		unchM, imprS		
53	Iyer et al. (36)	59/F	3	B-S	T3–T4	−/+	1NE/2VDD, SCH	1NE/2CR, ADP		imprSM	RC	6
54	Karadeniz-Bilgili et al. (37)	36/F	1,5	M left leg	T2–T3	−/+	1NE/2VDD, SCH	1Cyst/2CR, ADP		Improved	RC	2
55	Kumar et al. (42)	38/M	2	B-S	T7–T8	−/+	VDD, SCH	CB, CR, ADP		imprSM	RC	2
56	Maira et al. (43)	31/F	7	SM para	T8–T9	−/+	VDD, SCH	CR, WDD		imprSM	RC	156
57	Maira et al. (43)	54/F	8	B-S	T5	+/+	VDD, SCH	CR, ADP		imprSM	RC	132
58	Maira et al. (43)	45/F	24	B-S	T4–T5	−/+	VDD, SCH, DP	CR, ADP, discotomy		Worse	RC	48
59	Maira et al. (43)	50/M	2	SM para	T6–T7	−/+	VDD, SCH	CR, WDD		imprSM	RC	36
60	Maira et al. (43)	57/F	6	B-S	T4–T5	−/+	VDD, SCH	CR, ADP		imprSM	RC	24
61	Marshman et al. (45)	55/F	14	SM para	T7–T8	−/+	VDD, SCH	CR, ADP		imprSM		12
62	Maruichi et al. (46)	53/F	5	S right leg	T4–T5	+/+	VDD, SCH	CR, ADP		?	RC	7
63	Massicotte et al. (47)	39/F	?	SM para	T6–T7	−/+	VDD, SCH	CR, ADP		imprSM	RC	

Continues

TABLE 1. Continued

Case no.	Series (ref. no.)	Age (y)/sex	Interval (y)	Symptoms	Level	CTM/MRI	Surgical finding	Surgical technique	IONM	Operative result	MRI	FU (mo)
64	Massicotte et al. (47)	44/F	7	B-S	T5–T6	−/+	1NE/2VDD, SCH	1NE/2CR, ADP	EP	Unchanged		48
65	Massicotte et al. (47)	57/F	8	SM para	T6	−/+	1PAC/2VDD, SCH	1Cyst/2CR, ADP		Unchanged		36
66	Massicotte et al. (47)	27/M	1	B-S	T9	−/+	VDD, SCH	CR, ADP		imprM, unchS		12
67	Masuzawa et al. (48)	36/M	3	B-S	T4	+/+	LDD, SCH	CB, CR, ADP, PDP		imprSM		36
68	Matsumura et al. (49)	63/F	?	B-S	T3–T4	−/+	PAC, DDM, SCH	CR, WDD		Improved		
69	Miura et al. (51)	49/M	1	SM para	T5–T6	−/+	VDD, SCH	CR, WDD		imprSM	RC	6
70	Miyaguchi et al. (52)	54/F	2	B-S	T3–T4	+/+	DP, VDD, SCH	discect, CR, ADP	SSEP	imprSM	RC	6
71	Miyake et al. (53)	45/F	4	B-S	T3–T4	+/+	VDD, SCH	CR, ADP		imprSM	RC	1
72	Miyake et al. (53)	53/M	6	B-S	T2–T3	+/+	VDD, SCH	CR, ADP		imprSM	RC	1
73	Morley et al. (54)	28/F	2	B-S	T5–T6	−/+	VDD, SCH	CR, ADP		imprM, unchS		24
74	Morokoff et al. (55)	33/F	8	B-S	T8	−/+	VDD, SCH	CR, WDD		unchM, imprS		3
75	Najjar et al. (56)	32/M	5	B-S	T8–T9	−/+	1PAC/2VDD, SCH	1Cyst/2CR, ADP		Improved	RC	2
76	Nakagawa et al. (57)	77/F	9	SM para	T6–T7	+/+	VDD, SCH	BDF, CR, ADP		imprSM	RC	12
77	Nakazawa et al. (58)	43/F	5	B-S	T2	−/+	DDM, SCH	CR, WDD		unchM, imprS	RC	48
78	Nakazawa et al. (58)	39/F	4	B-S	T4	+/+	DDM, SCH	CR, WDD		imprSM	RC	
79	Novak et al. (60)	64/F	6	SM para	T6	−/+	VDD, SCH	CR, ADP	mMEP +eMEP	Unchanged	RC	12
80	Novak et al. (60)	45/F	2	SM para	T4–T5	−/+	VDD, SCH	CR, ADP	mMEP	Improved	RC	3
81	Novak et al. (60)	73/M	6	SM para	T4–T5	−/+	VDD, SCH	CR, ADP	mMEP +eMEP	Unchanged		9
82	Oe et al. (61)	61/M	?	SM para	T4	?/?	PAC, DDM, SCH	CR, WDD		Unchanged		
83	Pereira et al. (62)	55/M	4	SM para	T2–T3	−/+	VDD, SCH	CR, ADP		imprSM	RC	18
84	Rivas et al. (64)	49/M	3	B-S	T6–T7	+/+	VDD, SCH	CR, ADP		imprM, unchS		48
85	Roland et al. (65)	50/F	?	B-S	T4	+/+	VDD, SCH	CR, ADP		imprSM		
86	Sagiuchi et al. (66)	48/M	20	B-S	T7–T8	+/+	DP, VDD, SCH	CR, ADP		imprSM		
87	Sahl et al. (67)	51/M	16	SM para	T3–T4	+/+	VDD, SCH	CR, DS		imprSM		12
88	Saito et al. (69)	57/M	13	SM para	T2–T3	−/+	DDM, SCH	CR, ADP		imprSM	RC	1
89	Saito et al. (68)	68/F	36	SM para	T6–T7	−/+	VDD, SCH	CR, WDD		Unchanged	PAD	40
90	Sasaoka et al. (71)	57/M	15	SM para	T2–T3	+/+	VDD, SCH	CR, ADP, PDP	EP	unch, pain disapp	RC	24
91	Sioutos et al. (72)	34/F	1	SM para	T6–T7	−/+	PAC, VDD, SCH	CB, CR, ADP		Worse	RC	3
92	Slavotineket al. (73)	22/F	4	B-S	T5	−/+	1PAC/2VDD, SCH	1Cyst/2CR, fat graft		imprSM		
93	Spissu et al. (74)	56/F	1	B-S	T7	+/+	VDD, SCH	CR, DS		imprM, unchS		12

Continues

TABLE 1. Continued

Case no.	Series (ref. no.)	Age (y)/sex	Interval (y)	Symptoms	Level	CTM/MRI	Surgical finding	Surgical technique	IONM	Operative result	MRI	FU (mo)
94	Srinivasan et al. (75)	69/F	?	B-S	T3–T4	+/+	?	?		?		
95	Sugimoto et al. (76)	48/M	1	SM para	T4–T5	−/+	DDM, SCH	CR, WDD		imprSM	RC	12
96	Taghipour et al. (77)	32/M	3	B-S	T10–T11	−/+	DDM, SCH	CR,WDD		imprSM		
97	Tekkök (78)	49/F	3	B-S	T3–T4	−/+	VDD, SCH	CR, ADP		imprM, unchS	RC	5
98	Tronnier et al. (79)	45/F	4	S level T5	T3–T4	−/+	PAC, VDD, SCH	Cyst, CR, ADP		Worse	RC	4
99	Uchino et al. (80)	71/F	2	B-S	T4–T5	−/+	PAC, VDD, SCH	CR, DS		Unchanged	RC	
100	Uchino et al. (80)	61/F	2	B-S	T5–T6	+/+	NE	NE		Improved	PAD	
101	Urbach et al. (81)	44/M	2	S level T6	T5–T6	−/+	VDD, SCH	CR, ADP		Improved	RC	
102	Vallee et al. (82)	28/F	2	B-S	T3–T4	+/+	DDM, SCH	CR, WDD		imprSM	PAD	2
103	Vallee et al. (82)	58/F	7	B-S	T4–T5	−/+	DDM, SCH	CR, WDD		imprSM	PAD	2
104	Vallee et al. (82)	40/F	1,5	B-S	T5–T6	+/+	VDD, SCH	CR, ADP	SSEP	Unchanged		6
105	Vallee et al. (82)	49/F	3,5	B-S	T4–T5	+/+	VDD, SCH	CR, DS, PDP	SSEP	Unchanged		6
106	Verny et al. (84)	28/F	1,5	B-S	T3–T4	+/+	DDM, SCH	CR, WDD		imprSM	PAD	2
107	Verny et al. (84)	58/F	6	B-S	T4–T5	−/+	DDM, SCH	CR, ADP		imprSM	RC	
108	Wada et al. (85)	59/M	4	B-S	T4–T5	+/+	PAC, VDD, SCH	Cyst, CR, WDD		imprM, unchS	RC	48
109	Wada et al. (85)	48/M	2	B-S	T5–T6	+/+	VDD, SCH	CR, WDD		imprSM	RC	48
110	Wada et al. (85)	63/F	10	B-S	T3–T4	+/+	VDD, SCH	CR, WDD		imprM	RC	48
111	Watanabe et al. (86)	43/F	5	B-S	T4	+/+	DDM, SCH	CR, WDD		Improved	RC	156
112	Watanabe et al. (86)	39/F	3	B-S	T3	+/+	DDM, SCH	CR, WDD		Improved	RC	132
113	Watanabe et al. (86)	54/F	4	B-S	T4	+/+	DDM, SCH	CR, WDD		Improved	RC	24
114	Watanabe et al. (86)	71/F	5	SM para	T4	+/+	DDM, SCH	CR, WDD		Worse		24
115	Watanabe et al. (86)	49/M	5	B-S	T4	+/+	DDM, SCH	CR, WDD		Improved	RC	21
116	Watanabe et al. (86)	47/F	5	B-S	T5	+/+	DDM, SCH	CR, WDD		Improved	RC	18
117	Watanabe et al. (86)	78/F	4	SM para	T4	+/+	DDM, SCH	CR, WDD		Improved	RC	12
118	Watanabe et al. (86)	56/M	2	B-S	T6	+/+	1PAC/2DDM, SCH	1Cyst/2CR, WDD		Improved	RC	9
119	Watanabe et al. (86)	47/F	3	SM para	T3	+/+	DDM, SCH	CR, WDD		Improved	RC	6
120	Watters et al. (87)	55/F	10	B-S	T3–T4	−/+	1NE/2VDD, SCH	1NE/2CR, DS		imprSM	RC	18
121	White and Firth (88)	61/F	1,5	B-S	T4	−/+	VDD, SCH	CR, ADP		Unchanged	RC	
122	White and Firth (88)	39/M	1,5	B-S	T8	−/+	VDD, SCH	CR, ADP		Unchanged	RC	
123	White and Tsegaye (89)	61/M	4	B-S	T7	−/+	VDD, SCH	CR, ADP		imprSM		12
124	White and Tsegaye (89)	62/F	1,5	B-S	T6–T7	−/+	VDD, SCH	CR, ADP, PDP	MEP	imprSM	RC	12
125	White and Tsegaye (89)	66/F	8	SM para	T7	−/+	BD, VDD, SCH	BDF, CR, ADP, PDP	MEP	imprSM	RC	9
126	Wortzman et al. (91)	63/M	2	SM para	T7	Myelo	BD, VDD, SCH	CB, CR, DS		imprSM		24

Continues

TABLE 1. Continued

Case no.	Series (ref. no.)	Age (y)/sex	Interval (y)	Symptoms	Level	CTM/MRI	Surgical finding	Surgical technique	IONM	Operative result	MRI	FU (mo)
127	Case 1	42/M	5	B-S	T5–T6	−/+	1PAC/2VDD, SCH	1Cyst/2CR, ADP	SSEP, mMep +eMEP	Worse	RC	30
128	Case 2	60/M	2	B-S	T6–T7	+/+	VDD, SCH	CR, ADP	SSEP, mMEP +eMEP	imprSM	RC	20
129	Case 3	69/M	5	SM para	T7–T8	+/+	VDD, SCH	CR, ADP	SSEP, mMep +eMEP	imprSM	RC	10

[a] CTM, computed tomography myelography; MRI, magnetic resonance imaging; IONM, intraoperative neurophysiological monitoring; FU, follow-up; B-S, Brown-Séquard syndrome; VDD, ventral dura defect; SCH, spinal cord herniation; CR, cord release; WDD, widening dura defect; imprSM, motor and sensory function improvement; RC, realigned spinal cord; DDM, duplicated dura mater; imprM, motor function improvement; unchS, unchanged sensory function; SM para, paraparesis and sensory deficit; ?, unknown; ADP, anterior dura patch; 1, first exploration; PAC, posterior arachnoid cyst; 2, second exploration; NE, negative exploration; BD, bony defect; BDF, bony defect filling; Cyst, excision of arachnoid cyst; SSEP, somatosensory evoked potentials; mMEP, muscular recorded motor evoked potentials; eMEP, epidurally recorded motor evoked potentials; DS, direct suture dura defect; CB, cord biopsy; S bilat, bilateral sensory disturbance; PAD, persistent anterior displacement; SCT, suspicion of cord tumor; unchM, motor function unchanged; imprS, sensory function improved; Mleft leg, isolated motor dysfunction left leg; EP, evoked potential monitoring (unspecified); LDD, lateral dura defect; PDP, posterior dura patch; DP, disc prolapse; discect, discectomy; Myelo, myelography.

(unchanged or worse) in motor function as the binary dependent variable. After adjusting for age, gender, operative interval, and vertebral segment of cord herniation, we analyzed outcomes of imaging, surgical approach, surgical findings, and surgical technique as independent variables to allow us to define which variables were associated with improvement or no improvement (unchanged) or with deterioration (worsening) after surgical intervention. A P value of 0.05 was used as a cutoff for sequentially entering and removing each variable. We reported the odds ratios with 95% confidence intervals (CIs) and applied the Hosmer-Lemeshow test to evaluate the model calibration. All statistical analyses were performed using SPSS 14.0 for Windows (SPSS Inc., Chicago, IL).

RESULTS

All 129 patients were adults. Age was normally distributed (Shapiro-Wilk test, $P < 0.05$) and ranged from 21 to 78 years (mean age, 51 years). Women (n = 80) were more affected (64%) than men (male-to-female ratio, 1:1.8). Brown-Séquard syndrome was present in 85 cases (66%), paraparesis in 39 cases (30%), isolated sensory deficit in 4 cases (3%), and isolated motor deficit in 1 patient (1%). Operative interval (interval between the onset of symptoms and surgical treatment) ranged from 6 months to 36 years (mean interval, 5.2 years). In 17 cases, the operative interval was unknown. Segmental distribution is shown in Figure 4. ATSCH has a predisposition for the T3–T7 vertebral segments (80%; $P < 0.001$). In 129 patients, 218 vertebral segments were involved: 40 dura defects were at the level of the vertebral body; in 88 patients, the dura defect was on the level of the intervertebral disc; and in 1 patient, the dura defect extended over 3 segments (T4–T6) (20).

Operative results were analyzed in 121 patients. Two patients that worsened after spinal cord biopsy (32, 33, 72), 2 patients after negative exploration (13, 80), and 4 patients with unknown operative results (21, 46, 75, 83) were excluded. Improvement in (sensory) motor function was reported in 88 patients (73%). In 24 patients (20%), (sensory) motor function did not change after the operation, and in 9 patients (7%), motor function deteriorated because of operative intervention (Table 2).

Follow-up period was specified in 93 cases (72%), ranging from 1 month to 13 years. Mean follow-up was 24.4 months.

Improved patients did not differ from stable (unchanged) or deteriorated (worse) subjects in terms of gender, age, and operative interval. However, cord herniation at T3–T4 and T8–T9 had a statistically significant higher prevalence among improved patients (18.3% versus 4.2% and 8.3% versus 0%, respectively).

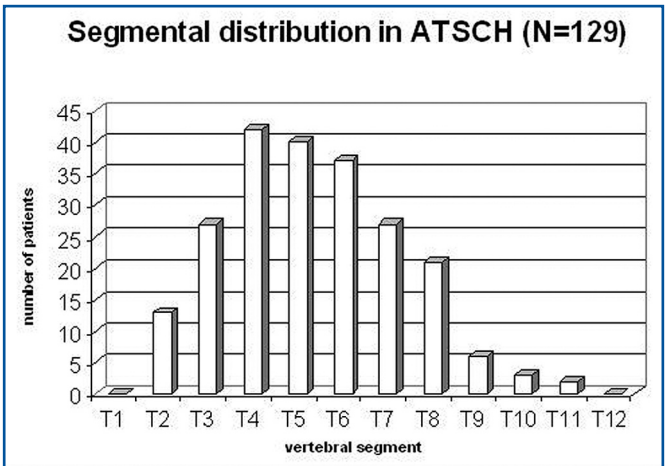

FIGURE 4. *Segmental distribution of anterior thoracic spinal cord herniation in 129 cases.*

TABLE 2. Operative results in anterior thoracic spinal cord herniation (n = 121)

	No. (%)
Improved	88 (68)
Unchanged	24 (19)
Worse	9 (7)
Total	121 (100)

Furthermore, Brown-Séquard syndrome was statistically significantly associated (Fisher's exact test, $P = 0.033$) with postoperative improvement in motor function, as three fourths (75.9%) of those patients improved compared with 55% of patients with paraparesis (95% CI, 2.42–39.5%). This can be denoted as a clinically relevant difference (Cohen's $w = 0.21$) compared with the outcome of patients who experienced paraparesis before the operation (Table 3). Unchanged or worsened motor function after surgical treatment was associated with the presence of a PAC, whereas improvement in motor function was associated with the observation of a duplicated dura mater. The difference in proportions of subjects with PAC (77.8%) and without PAC (26.7%) was statistically significant among patients with unchanged or worsened postoperative motor function (95% CI, 22.7–79.4%); this difference was clinically relevant (Cohen's $w = 0.29$). The difference between proportions of patients with duplicated dura mater (87.0%) and without duplicated dura mater (65.7%) was significant among improved subjects (95% CI, 4.71–37.8%) and clinically relevant (Cohen's $w = 0.18$).

Focusing on the operative technique, the operative results with respect to neurological outcome (motor function) were analyzed. In a bivariate analysis, we found that only cord release (CR) was associated with improvement in postoperative motor function ($P = 0.01$); 74.8% of patients with CR were improved versus 45.5% of patients without CR (95% CI, 6.97–51.7%), which was a clinically relevant difference (Cohen's $w = 0.24$).

Spinal CR, followed by direct suture (DS) of the dura defect, a widening of the dura defect (WDD), or application of an anterior dura patch (ADP), was performed in a total of 115 patients. Only 12 patients were treated by DS. Because both DS and ADP require significant manipulation of the spinal cord, we decided to compare DS and ADP operative results with the results after WDD. This did not reveal a statistically significant association between WDD and postoperative improvement in motor function (Fisher's exact test; $P = 0.173$). However, when DS was removed from the analysis and only the treatment outcome of ADP was compared with that of WDD, the operative result was statistically significant and had a clinically relevant difference across the used operative technique (Fisher's exact test, $P = 0.036$; Cohen's $w = 0.19$) (Table 4). WDD was associated with the lowest prevalence of unchanged/worsened motor function (difference of proportions unchanged/worsened between ADP and WDD was 17.5% (95% CI, 2.24–34.1%). A bias of favorable preoperative neurological condition was ruled out because Brown-Séquard syndrome and paraparesis

TABLE 3. Sociodemographic, clinical, and operative characteristics stratified by operative result[a]

	Improved (n = 87)	Unchanged or worse (n = 38)	
Sociodemographic and clinical			
Male, no. (%)	33 (37.9)	12 (31.6)	$P = 0.54$[b]
Female, no. (%)	54 (62.1)	26 (68.4)	
Age (y), mean ± SD	50.3 ± 12.8	51.5 ± 13.4	$P = 0.62$[c]
Brown-Séquard syndrome, no. (%)	63 (75.9)	20 (24.1)	$P = 0.033$[b]
Paraparesis, no. (%)	21 (55.3)	17 (44.7)	
Operative interval (mo), mean ± SD	56.5 ± 49.3	71.0 ± 82.8	$P = 0.25$[c]
CTM, no. (%)	40 (78.4)	11 (21.6)	$P = 0.75$[b]
MRI, no. (%)	83 (69.7)	36 (30.3)	$P = 0.52$[b]
Vertebral level cord herniation, no. (%)			95% CI[d]
T2–T3	8 (13.5)	3 (13.6)	−16.7 to 16.9%
T3–T4	11 (18.6)	1 (4.5)	−27.3 to 0.9%
T4–T5	12 (20.3)	6 (27.3)	−14.3 to 28.2%
T5–T6	5 (8.5)	6 (27.3)	−1.1 to 38.7%
T6–T7	7 (11.9)	3 (13.6)	−14.8 to 18.4%
T7–T8	9 (15.3)	3 (13.6)	−18.6 to 15.4%
T8–T9	5 (8.5)	0	−15.6 to 1.5%
T9–T10	2 (3.4)	0	−8.0 to 1.2%
Operative finding, no. (%)			
Ventral dura defect	56 (68.3)	26 (31.7)	$P = 0.69$
Spinal cord herniation	77 (72.6)	29 (27.4)	$P = 0.10$
Posterior arachnoid cyst	2 (22.2)	7 (77.8)	$P = 0.003$[b]
Duplicated dura mater	20 (87.0)	3 (13.0)	$P = 0.048$[b]
Disc prolapse	2 (66.7)	1 (33.3)	$P = 0.99$
Bony defect	3 (75.0)	1 (25.0)	$P = 0.99$
Suspicion cord tumor	0 (0.0)	1 (100.0)	$P = 0.30$
Lateral dura defect	1 (100)	0 (0.0)	$P = 0.99$
Operative technique, no. (%)			
Cord release	77 (74.8)	26 (25.2)	$P = 0.01$[b]
Widening dura defect	27 (81.8)	6 (18.2)	$P = 0.08$
Negative exploration	4 (57.1)	3 (42.9)	$P = 0.43$
Anterior dura patch	46 (63.9)	26 (36.1)	$P = 0.12$
Direct suture	12 (85.7)	2 (14.3)	$P = 0.22$
Bony defect filling	3 (75.0)	1 (25.0)	$P = 0.99$
Cord biopsy	4 (66.7)	2 (33.3)	$P = 0.99$
Cyst excision	1 (25.0)	3 (75.0)	$P = 0.08$
Posterior dura patch	3 (60.0)	2 (40.0)	$P = 0.99$
Fat graft	1 (100.0)	0 (0.0)	$P = 0.99$

[a] SD, standard deviation; CTM, computed tomography myelography; MRI, magnetic resonance imaging; CI, confidence interval.
[b] Fisher's exact test.
[c] Independent samples t test.
[d] Difference of proportions test.

TABLE 4. Operative result related to operative technique (n = 103)[a]

	Anterior dura patch	Widening dura defect	Total
Improved	45 (64.3%)	27 (81.8%)	72
Unchanged/worse	25 (35.7%)	6 (18.2%)	31
Total	70	33	103

[a] Fisher's exact test: $P = 0.036$.

TABLE 5. Preoperative neurological symptoms related to operative technique (n = 101)[a, b]

	Anterior dura patch	Widening dura defect	Total
Brown-Séquard syndrome	45 (66.2%)	23 (33.8%)	68
Paraparesis (SM)	23 (69.7%)	10 (30.3%)	33
Total	68	33	101

[a] SM, sensorimotor.
[b] Fisher's exact test: $P = 0.129$ (not significant).

were represented equally in both groups (ADP and WDD) (Table 5). Persistent anterior displacement or realignment of the spinal cord on postoperative MRI did not correlate with postoperative outcome. Also, no statistically significant association was found between operative techniques (WDD, DS, or ADP) and spinal cord position on postoperative MRI (realignment of the spinal cord or persistent anterior displacement).

Fifteen patients were operated on twice because of a misdiagnosis (6, 9, 11, 20, 36, 37, 47, 56, 73, 86, 87). At the initial operation, a PAC was detected and was thought to be responsible for the neurological symptoms in 11 cases. In 4 patients, no abnormalities were found (negative exploration). All 15 patients deteriorated postoperatively and were re-explored, which finally resulted in treatment of spinal cord herniation. Operative results in those patients were not significantly unfavorable compared with those patients who were operated on only once.

IONM was performed in 20 cases: unspecified evoked potentials were reported in 2 patients, SSEPs in 5 patients, mMEPs in 3 patients, mMEPs + eMEPs in 2 patients, SSEPs + mMEPs in 4 patients, and SSEPs + mMEPs + eMEPs in 4 patients. Because of the small number of cases and the absence of uniformity in monitoring, no statistical analysis was performed on this item (i.e., IONM in operative treatment of ATSCH).

After exclusion of cases with negative exploration (13, 80), cord biopsy (16, 32, 42, 48, 72, 91), absence of preoperative motor deficit (16, 46, 79, 81), arachnoid cyst removal without CR (21, 35, 75, 83), and unknown postoperative results (21, 75, 83), a sample of 111 patients was available for multivariable logistic regression to identify sociodemographic and clinical characteristics, operative findings, and surgical techniques as determinants of postoperative motor function improvement. Adjusted for age, gender, operative interval, and vertebral segment of cord herniation, only Brown-Séquard syndrome and CR showed a statistically significant association with motor improvement (odds ratio, 2.91; 95% CI, 1.15–7.39%; $P = 0.02$, Brown-Séquard syndrome; and odds ratio, 0.16; 95% CI, 0.05–0.52; $P = 0.02$, CR, respectively). Comparison of the number of patients actually classified in each group with the number predicted for each group was evaluated with the Hosmer-Lemeshow statistic, producing a nonsignificant χ^2 test result. The Hosmer-Lemeshow statistic for the model was 2.35 ($P = 0.93$), indicating a good fit.

DISCUSSION

ATSCH refers to herniation of the thoracic spinal cord through a mediolateral defect of the ventral (thoracic) dura mater. In the literature, different terminology was found to be in use (idiopathic spinal cord herniation, thoracic idiopathic spinal cord herniation, spontaneous spinal cord herniation, spontaneous thoracic spinal cord herniation) for the same entity. The cause of anterior thoracic spinal cord herniation remains speculative. A number of theories to explain the occurrence of a ventral thoracic dural defect have been postulated, such as a history of trauma (15, 27, 67, 81, 87, 91), pressure erosion of the ventral thoracic dura mater (35), thoracic disc herniation (32, 87, 91), congenital disorder (preexisting ventral meningocele) (48, 91), a duplication of the ventral dura mater (1, 3, 4, 43, 49, 51, 52, 58, 61, 68, 69, 76, 77, 82, 84–86), a congenital extradural arachnoid cyst (43), or an inflammatory process (56). Because the cause remains unknown, the rationale for a distinction between idiopathic or traumatic is missing. Therefore, we find it more appropriate to use the term ATSCH because this focuses on the radiological features that all these cases have in common.

Whatever the mechanism of the dura defect might be, the pathogenesis of herniation seems to be related to the fact that the spinal cord comes to lie against the defect and arachnoidal adhesions develop at the edges of the dura defect over time. Initially, cerebrospinal fluid (CSF) freely moves in and out of the defect with each pulsation. Possibly, herniation of an arachnoid membrane results in wedging and in widening of the dura defect. Conditions such as the negative pressure in the thoracic extradural space or variation in the force and amplitude of CSF pulsations during the cardiac cycle may also influence the course of the disorder (17). At a certain moment, the opening becomes blocked by the spinal cord, which will be sucked out by negative pressure in the epidural space (53) or pushed out by dorsal pressure (35) into the dura defect, finally leading to transdural spinal cord herniation (42, 77). Two out of 3 of our patients had a history of spinal trauma before the development of symptomatic ATSCH. It might be that a ventral dura rupture occurred at the time of trauma, possibly caused by an abrupt increase in intrathoracic and intradural pressure.

ATSCH predominantly occurs in middle-aged adults in the third and eighth thoracic vertebral segment and has a female preponderance (male-to-female ratio, 1:1.8). Patients usually present with Brown-Séquard syndrome. Initially, herniation of the lateral spinothalamic tract occurs, resulting in diminished

pain and temperature sensation, which is often unilateral and ascending. As herniation progresses, the corticospinal tracts also become involved, resulting in a gradually progressive weakness and spasticity of the leg(s). Paraparesis is present in almost one-third (30%) of the cases at the time of diagnosis. Isolated sensory or motor disturbance as the only symptom is rare. Before the advent of MRI, patients were misdiagnosed as having multiple sclerosis, transverse myelitis, some type of cord atrophy, or intradural adhesions (5, 63). Nowadays, diagnosis is made with axial MRI, which shows the dura defect and the cord herniation in most cases (1, 12, 16, 22, 23, 26, 35, 37, 54). CTM can show the attachment of the spinal cord to the ventral dura mater, and this is the method of choice to exclude an intradural arachnoid cyst (1, 12, 22, 51, 64, 74). In this review, 95% of the patients were diagnosed using MRI. CTM was performed in only 2 patients, and MRI was followed by CTM in 40%. Myelography was performed in only 2 cases (20, 91). Only 2 out of 80 papers on ATSCH were published before 1990, which means that ATSCH typically is a diagnosis from the MRI era.

Operative treatment of ATSCH is promoted by most authors; however, several cases of conservative treatment have been reported (2, 4, 26, 28, 32, 47). Ammar et al. (4) described 2 patients who were followed without operation and 1 patient who needed 4 operations because of retethering after initial CR and WDD, finally leading to nearly complete paraparesis after an initial operation and a second operation had resulted in temporary improvement (see also Hadley [31]). Batzdorf (11) reported on a patient who needed reoperation due to retethering of the spinal cord because of neurological deterioration 3 years after successful initial cord reduction and dura grafting. Ewald and Hassler (24) reported on a patient who deteriorated 6 weeks after operative treatment and needed re-exploration because of cord swelling, possibly due to cord strangulation by the dura patch. No strangulation, however, was found during reoperation (C. Ewald, personal communication). A posterior dura widening was performed, but symptoms did not improve. Bode et al. (14) reported a patient who experienced secondary deterioration caused by retethering after closure of the dura defect with DuraGen after initial postoperative improvement. In Adams and Anslow (2), the diagnosis was made using MRI and CTM. After 1 year, motor deficit had progressed, and MRI showed an increase in cord herniation and a signal change on the left side of the spinal cord. A similar evolution, illustrated by sequential MRI, was reported by Ewald et al. (25). Most authors promoted operative treatment of ATSCH, claiming up to an 85% favorable outcome (7, 24, 25, 60, 62).

The current study is based on case reports from the literature. So, by definition, it is retrospective, which results in a number of well-known drawbacks. However, because the prevalence and incidence of ATSCH are very low, it is impossible to evaluate the clinical outcomes in a cross-sectional or longitudinal observational study because such samples will not meet the level of sufficient statistical power. Consequently, prospective series from single departments or from multicenter study groups are not available, and evidence-based operative strategies are lacking. This means that the only available source of the information needed for quantitative research on this topic is a sample of patients with ATSCH who were described in sufficient detail in case reports that were published in international scientific medical journals. Therefore, we collected and analyzed data from a large sample of ATSCH patients following the method of IPD meta-analysis (90) that we used in an earlier study (29). With the inclusion of 126 patients who were identified in 74 publications, supplemented with 3 patients treated in the authors' neurosurgery department, we reached sufficient statistical power so we could use multivariable statistical analysis. One disadvantage of IPD meta-analysis compared with aggregated data meta-analysis is that IPD meta-analysis is time-consuming and prone to information bias as the definition of constructs may vary across countries. However, 2 great advantages of IDP meta-analysis are 1) the greater precision in measurement compared with meta-analysis in which aggregate data are pooled at study level and 2) the opportunity to use multivariate analyses on individual data and controlling for a number of confounding factors at patient level. Standardization of case reports would allow for better comparison of individual cases. However, at present, and in the absence of such a standard, the data used are the best available for analysis to improve understanding of this serious clinical disorder.

In our review, covering all cases that were reported in the international medical literature, postoperative improvement in at least motor function was found in 68% of all cases. In 19% of all cases, the neurological condition had stabilized after the operation, and in 7%, the operation had resulted in permanent neurological deterioration. We found that patients presenting with Brown-Séquard syndrome had better postoperative results than did those with spastic paraparesis. Nevertheless, it should be kept in mind that the data regarding pre- and postoperative neurological grading and follow-up are imprecise and incomplete in many of the reports. This lack of available detail does not permit correlation of the degree of preoperative neurological deficit with the degree of postoperative recovery. As a consequence, we can only give a global score for motor and/or sensory results: improved, unchanged, or worse.

Level (Vertebral Segment) of Cord Herniation

Among improved patients, spinal cord herniation at T3–T4 and T8–T9 had a statistically significant higher prevalence (18.3% versus 4.2% and 8.3% versus 0%, respectively). This might suggest that ATSCH between T5 and T7 has an unfavorable outcome, which would fit the classic view of the vasculature of the spinal cord that indicates that the watershed zone of ischemic vulnerability is in the mid-thoracic area (18). However, the majority of patients with ATSCH at T3–T4 and T8–T9 presented with Brown-Séquard syndrome. Because Brown-Séquard syndrome was identified as a strong independent factor associated with postoperative motor function improvement, we were unable to come to any conclusions about the influence of the anatomic level of ATSCH on postoperative outcome.

Surgical Strategy

Although conservative treatment was reported in 10 cases (2) (Cases 2 and 3 in Ammar et al. [4], Case 3 in Ferré [26], Cases 3

and 4 in Hausmann and Moseley [32], Cases 1, 3, 5, and 8 in Massicotte et al. [47]), most patients with ATSCH were indicated for operation. The aim of surgery is to release the strangulated spinal cord (CR), restore the cord to its normal intradural position, and prevent recurrence of cord herniation. The goal of surgical treatment is to stop and eventually reverse neurological deterioration.

Principally, there are 2 surgical strategies for preventing reherniation of the spinal cord. The first strategy is closure of the dura defect, either by DS of the ventral dural defect (12, 15, 16, 34, 80, 82, 87, 91) or by filling the dural defect with a fat graft (73), or by using a patch (fascia, pericard, or some other artificial material) to cover or suture the dural defect (ADP) (5, 8, 9, 11, 15, 17, 22, 24–26, 30, 32, 36, 37, 42–48, 52, 53, 56, 57, 62, 65, 66, 71, 72, 78, 79, 81, 82, 84, 88, 89) after releasing the spinal cord and reducing the herniation. Promoters of this strategy state that closure of the dura defect is necessary to prevent postoperative CSF circulation disturbances that may be produced by an extradural CSF collection (43) and because of the risk of recurrent spinal cord herniation (31, 78). Also, realignment of the spinal cord is thought to be essential, and it is claimed that this can only be achieved by covering or closure of the ventral dura defect (78).

The second strategy is WDD. This technique is mainly promoted in Japanese articles and is based on the perception of a so-called duplicated ventral dura mater. The spinal cord is said to be herniated through a defect in this dura mater duplication (1, 3, 4, 43, 51, 55, 58, 68, 69, 76, 77, 82, 84–86). Relief of the strangulated spinal cord can be obtained by resecting the dural ring around the cord, which at the same time results in a widening of the dura defect and also prevents reherniation and restrangulation of the cord. A number of authors claim that this technique is safer and is associated with the best operative outcome because the cord manipulations that are needed when closing the ventral dura defect (directly or by means of some type of patch) can be avoided (35, 56, 69, 86). Isu et al. (35) state that the aim of the operation is to resolve the incarceration of the spinal cord. In their opinion, it is not necessary to reduce the spinal cord herniation.

In multivariate logistic regression, spinal CR appears to be a strong independent factor associated with a favorable postoperative outcome (i.e., motor function improvement), irrespective of whether CR is followed by WDD, DS of the defect, or application of an ADP. After analysis of the data from 103 operative cases of ATSCH, we found statistical evidence in favor of WDD with respect to postoperative motor function improvement (Table 4). In 70 patients, postoperative MRI was performed to determine whether the spinal cord had returned to its natural position. In 7 patients, there was persistent anterior displacement (after WDD in 4 patients and after ADP in 3 patients), but this did not result in an unfavorable outcome compared with cases in which realignment of the spinal cord was confirmed. In this review, we were not able to find any support for the idea that closure of the ventral dura defect (by either DS or ADP) was in favor of WDD, when aiming for cord realignment on postoperative MRI. However, postoperative imaging was performed in only 56% of the 129 cases. To better define operative results from the perspective of operative technique, subsequent postoperative imaging (MRI or CTM) is needed to check whether cord herniation has been resolved and whether cord realignment has occurred.

It is evident that cord biopsy should not be part of the operative procedure in ATSCH.

Nowadays, ATSCH can be recognized and diagnosed preoperatively, using modern imaging techniques such as MRI and CTM. This helps avoid tissue sampling during exploration. In 6 cases, the spinal cord was biopsied during operation, which resulted in worsening of the neurological deficit in 2 patients (32, 33, 72) (separately reporting about the same patient). Interestingly, despite biopsy, 4 patients improved postoperatively (16, 42, 48, 91).

IONM

IONM is increasingly being used in operative treatment of spinal cord lesions (39–41). We monitored SSEPs and MEPs in all 3 patients. Motor potentials were evoked using transcranial electrical stimulation and were recorded epidurally as D waves (eMEPs) or muscularly as mMEPs. SSEP recording is not of much use in detecting and preventing a procedure-related motor deficit. In recent years, intraoperative spinal eMEP recording has proven very valuable in spinal cord tumor surgery because it allows monitoring of D waves and the intraoperative decrease in its amplitude (41, 50). The major advantage of combined mMEP and eMEP monitoring is that it identifies impairment of the functional integrity of the motor pathways before a permanent deficit has occurred (38, 40, 41, 70).

In our first patient, detethering of the spinal cord temporarily resulted in complete loss of motor potentials (mMEPs) and decrease in the distally recorded D-wave amplitude. After release of the cord and subsequent covering of the anterior dural defect with a GORE-TEX sleeve, the D waves returned to as much as 60% of the baseline, whereas mMEPs became visible again. Despite some improvement during the first postoperative year, motor function remained worse when compared with the preoperative condition. This was consistent with the decrease in mMEPs to the left leg at follow-up after 1 year. Novak et al. (60) reported significant change in MEPs (temporary loss of mMEPs and temporary reduction of eMEPs) in 1 patient with ATSCH (Case 1) at the time of insertion of the dural graft. Removal of the graft was followed by recovery of eMEPs and mMEPs. Reinsertion of the graft at the end of the procedure produced no new neurophysiological events. The postoperative outcome for this patient was initially worse, but during follow-up 1 year after the operation, neurological function had returned to preoperative status.

The IONM techniques used are based on the reported findings during spinal cord tumor surgery (41). Monitoring of eMEPs and mMEPs during operative treatment of ATSCH follows the same principles (38). In this review, 20 cases with IONM were identified, but only 6 recordings included both mMEPs and eMEPs. This very small number (n = 6) does not allow any statistical analysis or valid conclusions with regard

to the benefit of D-wave monitoring in ATSCH surgery. However, it is our experience that this type of monitoring can be a very useful tool for the surgeon in identifying impairment of the functional integrity of the motor pathways before a permanent spinal cord deficit occurs. Therefore, in our neurosurgical praxis, it has become routine in spinal cord surgery.

Follow-up

A small number of patients with secondary neurological deterioration after initial postoperative improvement were reported, 1 after WDD (4) and 3 after ADP (9, 14, 24). In 3 cases (4, 9, 14), reoperation revealed severe retethering of the spinal cord. It can be speculated that incomplete CR, reherniation, inadequate dura widening or induction of arachnoidal scarring by the graft material (Alloderm [9] and DuraGen [14]) were responsible for this. It remains unclear whether reoperation was performed in the Barrenechea et al. (9) case. Ewald and Hassler (24) suspected that strangulation of the edematous spinal cord by the dura patch was the cause of early deterioration (6 weeks after the initial operation), but this could not be confirmed at reoperation (personal communication). Ammar et al. (4) pointed out that the mean postoperative follow-up period for the 38 cases reported in their literature review was only 5.4 months, which is very short. Secondary deterioration in their patients occurred 18 months after the initial operation and finally resulted (after 3 reoperations, including attempted drainage of a suspected syrinx) in nearly complete paraparesis. This raises questions about the efficacy of surgical intervention. Because a number of cases with mild clinical symptoms that remain stable without operation (2) (Cases 2 and 3 in Ammar et al. [4], Case 3 in Ferré [26], Cases 3 and 4 in Hausmann and Moseley [32], Cases 1, 3, 5, and 8 in Massicotte et al. [47]) have been reported, it is mentioned in the literature that long-term follow-up is needed to further define the outcomes for those patients treated surgically. This would also help to better understand the natural history of the disease among those being managed conservatively (4). In 15 patients with thoracic myelopathy, however, a second operation was necessary because of progression of the neurological deficit. The initial operation failed to establish the proper diagnosis (11 patients with PAC and 4 patients with a negative exploration) and to resolve cord herniation. It seems plausible here that the initial diagnosis was incorrect and that those ATSCH cases (as in our index case) were misinterpreted as patients with a PAC. The diagnosis of ATSCH was made during reoperation, followed by release of the spinal cord and resolution of the cord herniation. This procedure resulted in improvement in 9 patients and in stabilization of the neurological deficit in 3 patients. Only 2 patients worsened (including our own Patient 1). In our opinion, this indicates that operative treatment is needed in the majority of patients with ATSCH and that conservative treatment is only an option in a limited number of cases. However, because the course of ATSCH is not well defined, the decision for operative treatment needs to be made on an individual basis.

CONCLUSION

ATSCH is a rare and, most probably, underdiagnosed disorder that is now receiving more attention with the increasing availability of MRI. The ventral dura defect plays a central role in the pathophysiology of ATSCH, but its cause remains unclear. The natural history is variable, but it typically presents as Brown-Séquard syndrome that may evolve over several years and can result in severe paraparesis. A few mild nonprogressive cases are reported, and it may be appropriate to treat such cases conservatively. However, operative treatment is indicated in the majority of cases, resulting in motor function improvement in as many as 70% of patients. The aim of surgery is spinal CR, realignment of the spinal cord, and prevention of reherniation. This can be achieved by WDD, or by release of the herniated cord, and closure/covering (DS) or some type of anterior patching (ADP) of the dura defect. Much dispute is found in the literature on this issue. However, in our analysis of 126 verified operative cases from the literature, together with 3 cases that were treated in our own department, we found a statistically significant association between motor function improvement and WDD and subsequent spinal CR. Postoperative cord realignment on MRI did not correlate with the operative technique used. Postoperative imaging, however, was performed in only 56% of the cases. Brown-Séquard syndrome appeared to be a strong independent factor associated with postoperative motor function improvement. This emphasizes the importance of early identification of ATSCH. Spinal CR in general (CR in combination with DS, ADP, or WDD) results in a high percentage of postoperative motor improvement (68% in the present analysis) or stabilization of the neurological deficit (19%).

In surgical treatment of ATSCH, IONM of the spinal cord, using transcranial electrical stimulation for recording of D waves, eMEPS, and mMEPS, appears to match the warning criteria used in the resection of intramedullary tumors. This type of monitoring can be very supportive for the surgeon because it provides online information about the functionality of the motor tracts and may be helpful in the preservation of motor function in surgery for ATSCH.

Because ATSCH is a rare disorder, clinical experience with treatment of this entity is very limited. Proper documentation is lacking in a significant number of reports, which hinders detailed analysis and obstructs the development of useful guidelines for proper treatment. Clinicians should be encouraged to report new cases and to describe the details that are discussed in the present review. Long-term follow-up of patients is essential, for both the patients who underwent operative treatment and the patients who were treated conservatively.

Disclosure

The authors have no personal financial or institutional interest in any of the drugs, materials, or devices described in this article.

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COMMENTS

Groen et al. offer a sincere attempt at making sense of an uncommon cause of thoracic paraparesis/myelopathy. The authors' retrospective review draws data together from multiple individual cases to give general information to clinicians faced with the management of similar patients. Even with this general insight, each patient must be managed individually. We agonized over the management of a single patient with thoracic cord herniation and neurological deficits for over 8 years. She had an aggressive connective tissue disease, which likely contributed to the ultimate failure. She responded well to our initial detethering procedure without suture or sling repair of the anterior defect for a long while. She developed recurrent symptoms and signs (and ultimately retethering) but was managed in her home state as a potential syrinx and was treated with attempted cyst to pleura shunting. When this failed and worsened her circumstances, she returned to us. We performed a second detethering procedure and this time placed an autographic fascia lata sling to cover the anterior defect and suspend the cord. She again improved for a period of time, only to return with remarkable contraction and scarring of the sling with constriction of her cord. We performed a third procedure, which eliminated the concentric constriction and detethered the cord yet again, but with a worse surgical and neurological outcome. Did her connective tissue disease contribute to advance the scarring and contraction of her own fascial graft and ultimately her poor outcome? (I think it, unfortunately, did.) The point is that individual patients must be managed individually even in the presence of medical evidence/reviews of treatment strategies about the entity from which they suffer.

The fact that thoracic cord herniation is identified on magnetic resonance imaging does not mean that surgery is indicated. We neuro-

surgeons do not treat x-rays, and, as the authors state, the procedure is directed at improving on or minimizing progressive thoracic spinal cord neurological loss in truly symptomatic patients. Two patients with documented thoracic cord herniation and intermittent symptoms without neurological signs, whom we have followed for years, have never undergone operations. This review suggests that careful microsurgical treatment does help most progressively symptomatic patients. We have subsequently treated 1 other markedly symptomatic patient with detethering only, with a good result and no late sequelae (at 3 years). We agree with the authors that surgical treatment should be based on documented progression of neurological symptoms and signs, not on the radiographic diagnosis alone.

The authors have collected and summarized meaningful Class III medical evidence on this subject. Wisely, they caution against drawing firm conclusions from their review. The authors do favor the recording of somatosensory evoked potentials and motor evoked potentials during these procedures. There is no convincing medical evidence (Class II or Class I) in support of the use of neurophysiological spinal cord monitoring in the management of patients with spinal cord tethering of any cause. I absolutely support their use as an adjunct during surgery if a surgeon feels that their use makes the patient "more comfortable" or "better" or improves the patient's outcome. I will just as sincerely defend those surgeons who do not use monitoring during spinal cord procedures. Neurophysiological monitoring remains an option for surgeons in the management of patients with spinal cord pathology who require surgical treatment. To recommend that somatosensory evoked potentials and motor evoked potentials be used during these procedures is an educated yet biased opinion (which is acceptable), not a medical fact or certainty. The authors present an exhaustive review of a difficult and variable pathological entity in a challenging and diverse group of patients with this uncommon disorder.

Mark N. Hadley
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Groen et al. have performed a review of the literature regarding operative management of anterior thoracic spinal cord herniation in addition to reporting 3 cases of their own. In a literature search, they located 74 articles with data on 126 patients. Specific areas of analysis included time interval between onset of symptoms and treatment, vertebral segment of cord herniation, imaging findings, surgical approach, operative technique and findings, intraoperative neurophysiological monitoring, and postoperative motor function. Two-thirds of the analyzed patients presented with Brown-Séquard syndrome, and one-third presented with lower-extremity paraparesis. The operative interval ranged from 6 months to 36 years from the time of symptom onset to treatment. Eighty percent of patients in the analysis experienced cord herniation between T3 and T7, with the dural defect located in the disc space twice as often as in the vertebral body.

Postoperative motor outcome was analyzed in 121 patients, with improvement noted in 73%, no change noted in 20%, and neurological deterioration seen in only 7%. Of the 93 cases followed for a mean of 24 months, cord herniation at T3–T4 or T8–T9 as well as Brown-Séquard syndrome on presentation had a statistically significant higher prevalence among improved subjects.

The surgical techniques used included cord release, direct suturing of the dural defect, anterior dural patch, and widening of the dural defect. The authors analyzed the results for 115 patients, and only cord release was associated with improved postoperative motor function. Postoperative magnetic resonance imaging findings of persistent anterior cord displacement or realignment of the cord did not correlate with postoperative outcome. Multivariate analysis was conducted on 111 patients, and only Brown-Séquard syndrome on presentation and spinal cord release as an operative technique were found to have a statistically significant association with motor improvement.

Absent from the authors' analysis is a description of complications from the various surgical techniques used, most notably cerebrospinal fluid leak and pleural fistulae associated with failure to close a ventral dural defect. Likewise, the mean follow-up interval of 24 months seems rather short for a disease process for which the natural history is not well defined but does appear to have the potential for a relapsing, remitting course, especially in light of the fact that, in the treatment of this condition, dural closure is not universally accepted as necessary or even beneficial.

Mark Dannenbaum
Daniel H. Kim
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In this article, Groen et al. present a very thorough meta-analysis of the current literature regarding the natural history and treatment of anterior thoracic spinal cord herniation. In addition, they present 3 new cases from their own institution. These data showed that presentation with Brown-Séquard syndrome and surgical treatment by widening of the dural defect are associated with a more favorable prognosis. Because anterior thoracic spinal cord herniation is a rare and relatively newly described condition, definitive statements regarding optimal treatment are difficult to make. However, analyses such as this one currently represent the most efficient way to aggregate and draw conclusions from the infrequent and widely distributed experiences of the global neurosurgical community.

Christopher Wolfla
Milwaukee, Wisconsin

Spinal cord herniation is a rare disease. It can present as a progressive myelopathy. The authors have performed a meta-analysis of the literature to determine those factors associated with better surgical outcomes. These factors were clinical presentation with a Brown-Séquard syndrome, surgical release of the spinal cord, and widening of the dural defect. Meta-analysis is 1 way of studying rare conditions. It can provide some insights into disease management. However, one must interpret the results of such studies with caution.

The major controversy in the management of spinal cord herniation is the appropriate surgical technique for repairing the herniation. Most surgeons with experience in managing this condition would agree that reducing or releasing the incarcerated spinal cord is absolutely necessary. The remaining question is how to manage the dural defect. Some authors, mainly those from Japan, have recommended widening the dural defect. In doing so, the surgeon eliminates the possibility of cord strangulation through a narrow defect. These studies report excellent results with this option. Alternatively, other surgeons have advocated primary or patch repair of the defect. My practice has been to repair the defect primarily if it can be done with a minimum of cord manipulation. I will use a patch graft or sling when this is not possible. Having treated 6 of these patients, I have yet to see a case in which I felt that either of these 2 options was not safe. I have concerns, perhaps unfounded, that simple expansion of the dural defect may not be durable. Still, the results of this study favor widening of the dural defect over dural repair. I think this is a good option when dural repair is not easy or safe.

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