Rheumatoid Arthritis of the Cervical Spine

A Long-Term Analysis with Predictors of Paralysis and Recovery*

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ABSTRACT: We analyzed the cases of seventy-three patients who were managed over a twenty-year period for rheumatoid involvement of the cervical spine and were followed for a minimum of two years, with an average follow-up of seven years. A neurological deficit did not develop in thirty-one patients (Ranawat et al. Class I) and paralysis developed in the remaining forty-two patients: Class II in eleven and Class III in thirty-one. Of the forty-two patients in whom paralysis developed, thirty-five had operative stabilization. Seven patients were managed with a soft cervical collar because they refused or were medically unable to have the operation; all of them had an increase in the severity of the paralysis.

The posterior atlanto-odontoid interval and the diameter of the subaxial sagittal canal measured on the cervical radiographs demonstrated statistically significant correlations with the presence and severity of paralysis. All of the patients who had a Class-III neurological deficit had a posterior atlanto-odontoid interval or diameter of the subaxial canal that was less than fourteen millimeters. In contrast, the anterior atlanto-odontoid interval, which has traditionally been reported, did not correlate with paralysis. The prognosis for neurological recovery following the operation was not affected by the duration of the paralysis but was influenced by the severity of the paralysis at the time of the operation. The most important predictor of the potential for neurological recovery after the operation was the preoperative posterior atlanto-odontoid interval. In patients who had paralysis due to atlanto-axial subluxation, no recovery occurred if the posterior atlanto-odontoid interval was less than ten millimeters, whereas recovery of at least one neurological class always occurred when the posterior atlanto-odontoid interval was at least ten millimeters. If basilar invagination was superimposed, clinically important neurological recovery occurred only when the posterior atlanto-odontoid interval was at least thirteen millimeters. All patients who had paralysis and a posterior atlanto-odontoid interval or diameter of the subaxial canal of fourteen millimeters had complete motor recovery after the operation.

In this series, although only patients who had a neurological deficit were operated on, we observed the range of the posterior atlanto-odontoid interval that was associated with poor or no recovery after the operation, and we identified the safe range on the basis of the patients in whom paralysis did not develop. Therefore, to minimize the potential risk of the development of irreversible paralysis, we recommend operative stabilization of the rheumatoid cervical spine, in the presence or absence of a neurological deficit, for patients who have atlanto-axial subluxation and a posterior atlanto-odontoid interval of fourteen millimeters or less, patients who have atlanto-axial subluxation and at least five millimeters of basilar invagination, and patients who have subaxial subluxation and a sagittal diameter of the spinal canal of fourteen millimeters or less.

The cervical spine is a common focus of destruction from rheumatoid arthritis, and the resultant instability and neural compression are known complications[10,28,45,46]. Instability in this disorder is secondary to destruction of articular and ligamentous structures by proliferative synovial tissue[2,27,36,52,56]. The prevalence of subluxation has been reported to be 43 to 86 per cent, and the magnitude has previously been related to the severity of the systemic disease process[15,47,56]. Atlanto-axial subluxation is the most common type of instability (50 to 70 per cent of cases) and is usually anterior, but posterior and lateral subluxations may also occur[3,26,47]. Subaxial subluxations compose 20 to 25 per cent, and basilar invagination is the least common, but most dangerous, type of cervical instability in rheumatoid patients[66,68]. The reported rate of neural impairment due to cervical instability has ranged from 11 to 58 per cent[14,72]. This discrepancy may be attributed to variability in neurological classification

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TABLE I
TYPE OF INSTABILITY ASSOCIATED WITH NEUROLOGICAL DEFICIT

	Patients Who Had a Deficit	Patients Who Did Not Have a Deficit	Total No. of Patients
Atlanto-axial subluxation			
Alone	12	28	40
With basilar invagination	19*	1	20
With subaxial subluxation	1	0	1
Total	32	29	61
Subaxial subluxation	8	2	10
Basilar invagination alone	2	0	2
Over-all total	42	31	73

*One of these patients also had subaxial subluxation.

systems as well as difficulty in the detection of subtle neurological deterioration in patients who have muscle weakness and atrophy secondary to chronic rheumatoid arthritis, which can mask the weakness caused by compression of the spinal cord.

Many studies have focused on the definition of the prevalence of radiographic subluxations or on reports of the results from various operative treatments of the unstable rheumatoid cervical spine, yet precise indications for operative intervention remain unclear. In 1981, Pellicci et al. noted that although 80 per cent of their rheumatoid patients who had cervical involvement had radiographic evidence of progression, only 36 per cent had progression of neurological disease[62]. In addition, since 50 per cent of their patients who had instability that was evident radiographically were asymptomatic, they recommended so-called conservatism — that is, not undertaking prophylactic stabilization on the basis of radiographic instability. Others have used the presence of a progressive neurological deficit as the primary indication for stabilization[81,89]. More recently, Clark et al. concluded that operative stabilization should be considered before potentially irreversible neurological changes occur in patients who have severe instability and abnormal findings on radiographic studies[11]. The wide variation in radiographic measurements and the overlap between symptomatic and asymptomatic patients has made a precise definition of radiographic evidence of substantial instability elusive. Furthermore, the poor correlation of neurological symptoms with such instability makes identification of an impending neurological deficit, and therefore of the ideal candidate for treatment, extremely difficult[13,62].

In the present investigation, we analyzed the cases of seventy-three patients who had rheumatoid arthritis involving the cervical spine and who were managed over a twenty-year period. To further define the operative indications for this disorder, our goals were to determine the radiographic parameters that most reliably predict the onset of paralysis and the potential for neurological recovery after an operation, and to discover whether the duration and severity of neurological involvement affect recovery.

Materials and Methods

Ninety-two consecutive patients who had rheumatoid arthritis involving the cervical spine were referred for orthopaedic consultation and had radiographs and medical records available for review. These patients were seen at The Johns Hopkins University Hospital between 1966 and 1972, inclusive, and at the Case Western Reserve University Hospitals between 1973 and 1987, inclusive. Seventy-three of these patients had adequate clinical and radiographic follow-up for a minimum of two years after the consultation, or until the time of death, and formed the basis of this retrospective analysis.

There were fifty women and twenty-three men, and the average age at the time of the consultation was sixty-one years (range, thirty-five to eighty-four years). The average duration of follow-up was seven years (range, two to sixteen years) for the patients who were alive at the end of the study period. Thirty-one patients were followed until the time of death, with an average duration of follow-up of five years (range, one month to twenty years). Seventeen patients died less than two years after the consultation; nine, between two and six years; two, between seven and twelve years; and three, between thirteen and twenty years.

Clinical Evaluation

Neurological deficits were classified by one of us (L. D. D.), an independent observer, using the system of Ranawat et al.[67]. At the time of the initial consultation, thirty-one patients had no neurological deficit (Class I); eleven patients had subjective weakness, hyperreflexia, and dysesthesias (Class II); and thirty-one patients had objective findings of weakness and long-tract signs (Class III), of whom eight had Class-IIIA deficits and could walk and twenty-three had Class-IIIB deficits and were quadriparetic and unable to walk. The deficits were classified again on the basis of the neurological examination at the time of the follow-up examination.

Radiographic Evaluation

Measurements were recorded on the original and follow-up flexion and extension radiographs of the cervical spine that had been made with a standard tube-distance of six feet (1.8 meters). Atlanto-axial subluxation was defined as an anterior atlanto-odontoid interval (the distance between the posterior edge of the anterior ring of the atlas and the anterior surface of the odontoid process, as measured along the transverse axis of the ring of the atlas) of more than three millimeters[30-32,35,41,51,76]. The posterior atlanto-odontoid interval (the distance between the posterior surface of the odon-

TABLE II

DATA ON THE FORTY-TWO PATIENTS WHO HAD A NEUROLOGICAL DEFICIT

Case	Sex, Age (Yrs.)	Duration of Paralysis (Mos.)	Radiographic Measurements* (mm)				Neurological Class[67]		Previous Op.	Op. Treatment	Complications	Duration of Follow-up (Yrs. + Mos.)
			C1-Odontoid Interval		Vertical Migration†	Diameter of Subaxial Canal‡	Initial	Latest Follow-up				
			Ant.	Post.								
C1-C2 sublux. alone												
1	M, 66	6	8	14	3	—	II	I	None	C1-C2 arthrodesis	None	3 + 4, died of unknown causes
2	F, 59	5	5	13	3	—	II	I	None	C1-C2 arthrodesis	None	2 + 2
3	F, 62	6	10	13	2	—	II	I	None	C1-C2 arthrodesis	None	2 + 6
4	M, 57	24	8	12	3	—	II	I	None	C1-C2 arthrodesis	None	8 + 0, died of unknown causes
5	M, 70	<1	9	12	3	—	IIIA	II	None	C1-C2 arthrodesis	None	4 + 6
6	M, 38	3	8	11	3	—	II	I	None	C1-C2 arthrodesis	None	4 + 10
7	F, 55	36	7	10	4	—	II	I	C1-C2 methacrylate fixation failed	C1-C2 arthrodesis	None	16 + 4
8	M, 60	2	13	7	3	—	IIIB	IIIB	None	C1-C2 arthrodesis	None	0 + 1, died of unknown causes
9	M, 50	6	9	5	3	—	IIIB	IIIB	None	C1-C2 arthrodesis	None	6 + 8, died of pneumonia
C1-C2 sublux., basilar invag.												
10	F, 57	13	5	17	15	—	IIIA	I	None	Occ.-C2 arthrodesis; C1 laminect.	None	16 + 0
11	F, 60	5	6	14	8	—	II	I	None	Occ.-C2 arthrodesis	Occ.-C2 pseudarthrosis	2 + 4
12	M, 55	4	7	13	8	—	IIIB	II	None	Occ.-C3 arthrodesis	None	7 + 5
13	F, 58	2	6	13	11	—	IIIB	II	None	C1 laminect.; occ. decompression	None	15 + 6
14	F, 63	<1	8	12	5	—	IIIB	II	C1-C2 arthrodesis; C2-C4 arthrodesis failed	Occ.-C4 arthrodesis	C1-C2 pseudarthrosis	4 + 4, died of pneumonia, cord compression
15	M, 75	1	10	12	7	—	IIIB	IIIB	None	Occ.-C2 arthrodesis	None	1 + 5, died of unknown causes
16	M, 75	3	8	12	8	—	IIIA	IIIA	None	Occ.-C2 arthrodesis	None	1 + 8, died of unknown causes
17	F, 51	36	6	12	8	—	IIIB	IIIB	None	Transoral odontoid resect.	Progressive paralysis	0 + 1, died of cord compression
18	F, 35	1	10	11	7	—	IIIB	II	C1-C2 methacrylate fixation failed	Occ.-C3 arthrodesis	None	11 + 2
19	F, 58	6	3	11	7	—	IIIB	IIIA	None	Occ.-C2 arthrodesis	None	5 + 4, died of unknown causes
20	M, 60	6	5	11	6	13 C6-C7	IIIB	IIIA	None	Occ.-C7 arthrodesis	None	5 + 10, died of pneumonia, cord compression, T1
21	F, 58	2	14	10	5	—	IIIB	IIIA	None	Occ.-C2 arthrodesis	None	3 + 6
22	F, 67	2	8	10	9	—	IIIA	IIIB	None	Occ.-C2 arthrodesis; C1 laminect.; transoral odontoid resect.	Progressive paralysis	0 + 1, died of pneumonia, cord compression
23	F, 54	6	5	9	6	—	IIIB	IIIA	None	Occ.-C3 arthrodesis	None	0 + 8, died of brain tumor

toid process and the anterior edge of the posterior ring of the atlas, as measured along the transverse axis of the ring of the atlas) was also measured in patients who had atlanto-axial subluxation. In patients who had substantial erosion of the odontoid process, the measurement was made from the base of the remaining portion of the

TABLE II *(continued)*

DATA ON THE FORTY-TWO PATIENTS WHO HAD A NEUROLOGICAL DEFICIT

Case	Sex, Age (Yrs.)	Duration of Paralysis (Mos.)	Radiographic Measurements* (mm)				Neurological Class[67]		Previous Op.	Op. Treatment	Complications	Duration of Follow-up (Yrs. + Mos.)
			C1-Odontoid Interval		Vertical Migration†	Diameter of Subaxial Canal‡	Initial	Latest Follow-up				
			Ant.	Post.								
24	F, 56	6	6	9	9	—	IIIB	IIIB	None	Occ.-C3 arthrodesis	None	0 + 5, died of perforated ulcer, cord compression
25	M, 69	1	5	8	6	—	IIIB	IIIB	None	Occ.-C2 arthrodesis	Occ.-C1 pseudarthrosis	2 + 2, died of unknown causes
26	F, 66	2	11	8	8	—	IIIB	IIIA	C1-C2 arthrodesis failed	Occ.-C2 arthrodesis; C1 laminect.	None	4 + 4
27	M, 52	16	10	7	5	—	IIIA	IIIB	C1-C3 methacrylate fixation failed	Occ.-C3 arthrodesis; C1 laminect.; transoral odontoid resect.	Progressive paralysis	0 + 8, died of progressive vertical migration of C2
28	F, 60	2	12	6	7	—	IIIB	IIIA	C1-C2 arthrodesis	Occ.-C3 arthrodesis	Progressive basilar invag.	2 + 8
Subaxial sublux.												
29	F, 71	12	—	—	—	14 C3-C4 14 C4-C5 (15)	II	I	None	C3-C5 ant. decompression; arthrodesis	None	8 + 4
30	M, 64	24	—	—	—	14 C4-C5 (20)	II	I	None	C4-C6 ant. decompression; arthrodesis	None	7 + 11
31	F, 70	1	6	13	4	13 C3-C4 (20)	IIIB	II	None	C1-C4 post. arthrodesis	None	2 + 2
32	M, 65	1	—	—	—	12 C5-C6 (30)	IIIB	IIIA	None	C5-C6 post. arthrodesis	None	2 + 8, died of unknown causes
33	F, 79	<1	—	—	—	12 C5-C6 (40)	IIIB	IIIB	None	C5-C6 post. arthrodesis	Postop. bowel infarct.	0 + 1, died of infarct., cord compression
34	F, 50	18	—	—	—	10 C4-C5 12 C5-C6 11 C6-C7 (25)	IIIB	II	None	C2-C7 post. arthrodesis	None	9 + 6
35	F, 51	<1	—	—	—	8 C3-C4 (40)	IIIB	II	None	C3-C5 ant. decompression; arthrodesis	None	2 + 10, died of unknown causes
Non-op. treatment												
36	F, 67	<1	10	9	4	—	IIIA	IIIA	None	—	Progressive paralysis	0 + 1, died of cord compression, pneumonia
37	M, 62	8	3	20	12	—	II	IIIA	None	—	Progressive paralysis	3 + 9, died of cord compression, pneumonia
38	F, 69	1	—	—	—	11 C4-C5	IIIA	IIIB	None	—	Progressive paralysis	0 + 1, died of myocardial infarct.
39	F, 84	<1	5	9	9	—	IIIB	IIIB	None	—	Progressive paralysis	2 + 2, died of heart failure, pneumonia
40	F, 56	<1	16	8	4	—	IIIA	IIIB	None	—	Progressive paralysis	0 + 1, died of cord compression
41	F, 84	3	10	9	4	—	II	IIIB	None	—	Progressive paralysis	1 + 10, died of cord compression
42	F, 50	1	—	—	—	10 C2-C3 11 C3-C4	IIIB	IIIB	None	—	Progressive paralysis	0 + 11, died of cord compression

*All radiographic measurements were normal unless otherwise indicated.

†Cephalad to the McGregor line.

‡Measured from the inner laminar base of the superior spinous process to the posterior aspect of the vertebral body inferiorly. Slippage, expressed as a percentage of anteroposterior vertebral body width, is given in parentheses.

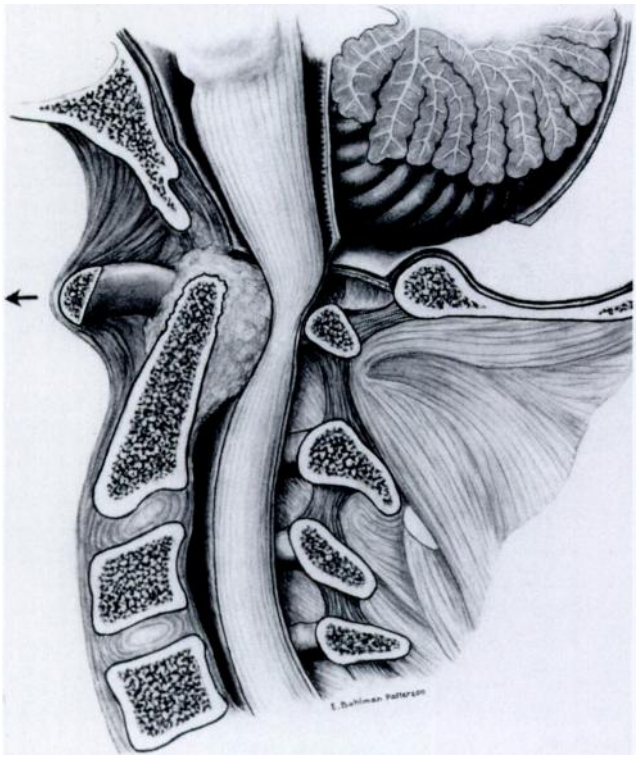

FIG. 1-A

Figs. 1-A through 1-D: Case 26. Atlanto-axial subluxation.

Fig. 1-A: Illustration demonstrating the forward subluxation of the atlas on the axis, pannus formation around the odontoid process, and osseous erosions. There is severe compression of the spinal cord between the pannus anteriorly and the arch of the atlas posteriorly.

second cervical vertebra. Basilar invagination was defined as migration of the odontoid tip more than 4.5 millimeters cephalad to the McGregor line[57,67,68,72] as drawn from the hard palate to the caudal surface of the basiocciput. Plain tomography was used to determine the severity of vertical migration of the odontoid. If subaxial subluxation was present, as defined for acute instability by White and Panjabi[86], the diameter of the subaxial canal was measured from the inner laminar cortex at the base of the superior spinous process to the posterior aspect of the vertebral body caudad at the most narrow part of the spinal canal. If osseous landmarks were uncertain on the plain radiographs, tomograms were made to confirm the measurements. All measurements were made by one of us (L. D. D.), an independent observer, before analysis of the data.

Myelography was used in all patients preoperatively to assess the degree and location of compression of the spinal cord. Pantopaque myelography was performed until 1980, when the use of intrathecal water-soluble contrast medium was instituted in conjunction with computerized axial tomography. Since 1984, magnetic resonance imaging has been helpful in visualization of the brain stem, the cervical spinal cord, and especially synovial pannus formation. Magnetic resonance imaging has also enabled visualization of the effect of cervical flexion and extension on atlanto-axial subluxation, basilar invagination, and subaxial subluxation.

Operative Management

All patients were managed non-operatively unless cervical instability with a neurological deficit developed. The type of operation performed was directed at the specific abnormality in each patient. Intubation was performed with the aid of a fiberoptic scope to avoid excessive passive manipulation in patients who had cervical instability. Patients who had isolated atlanto-axial subluxation had a posterior arthrodesis from the first to

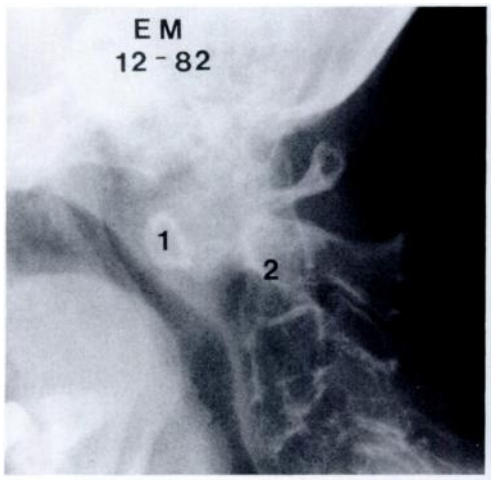

FIG. 1-B

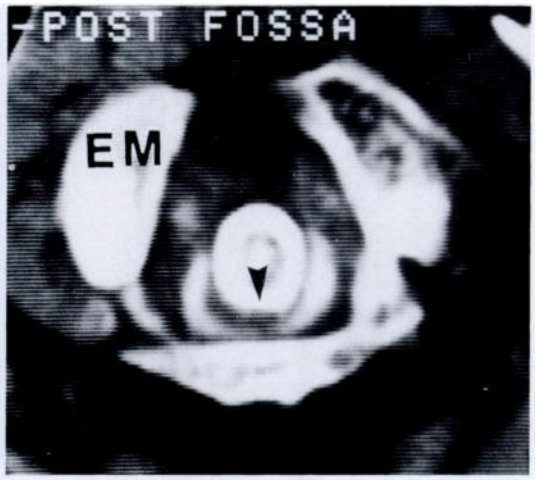

FIG. 1-C

Fig. 1-B: Lateral radiograph of a sixty-six-year-old woman who had had rheumatoid arthritis for thirty years. She began having increasing difficulty with weakness of the lower extremities, and she fell. After two visits to an emergency room, radiographs were made and she was noted to be quadriparetic, with an atlanto-axial subluxation. By the next month, she was wheelchair-bound and unable to walk. There is marked subluxation of the atlas on the axis, with very little room for the spinal cord, as evidenced by the narrow posterior atlanto-odontoid interval.

Fig. 1-C: Computerized tomographic scan, made after myelography, revealing the odontoid displacement posteriorly, severely compressing the spinal cord (arrowhead), which is surrounded by contrast medium.

TABLE III

RADIOGRAPHIC MEASUREMENTS RELATED TO INITIAL NEUROLOGICAL CLASSIFICATION

Class[67]	No. of Patients*	Radiographic Measurements (mm)							
		Atlanto-Odontoid Interval				Vertical Migration†		Diameter of Subaxial Canal‡	
		Anterior		Posterior					
		Range	Av.	Range	Av.	Range	Av.	Range	Av.
Occipitocervical levels									
I	29	3-16	7.5	11-21	14.8	2-8	3.7	—	—
II	9	3-10	7.2	9-20	12.9	2-12	4.6	—	—
IIIA	7	5-16	9.4	7-17	10.7	3-15	6.9	—	—
IIIB	18	3-14	7.9	5-13	9.7	3-11	6.7	—	—
Subaxial levels									
I	2	—	—	—	—	—	—	14-15	14.5
II	2	—	—	—	—	—	—	14	14
IIIA	1	—	—	—	—	—	—	11	11
IIIB	7	—	—	—	—	—	—	8-13	11

*The radiographic measurements for the two patients who had both atlanto-axial and subaxial instability are included in each part of the table.

†Cephalad to the McGregor line.

‡Measured from the inner laminar base of the cephalic spinous process to the posterior aspect of the vertebral body caudad.

the second cervical vertebra. For patients who also had basilar invagination, preoperative halo traction, with the head and neck in slight extension, was used in an attempt to obtain reduction before arthrodesis of the occiput to the second cervical vertebra. When it was impossible to reduce a fixed occipitocervical deformity and the posterior arch of the atlas was causing compression of the cord, a laminectomy of the atlas was performed with a high-speed diamond burr before posterior arthrodesis of the second cervical vertebra to the occiput. In patients who had severe fixed basilar invagination and atlanto-axial subluxation with anterior compression of the brain stem and myelopathy, the odontoid process was decompressed anteriorly through a transoral approach, although the extrapharyngeal approach is now a possible alternative[11,18,49,57,60]. Patients who had anterior subaxial subluxation and compression of the cord were managed with skeletal traction followed by a posterior arthrodesis. If the subaxial subluxation was not reducible, anterior decompression and arthrodesis were performed.

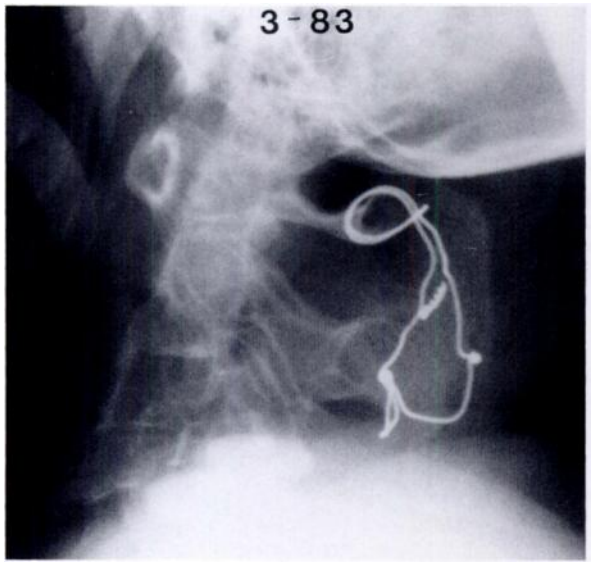

FIG. 1-D

Lateral radiograph made after reduction of the subluxation and atlanto-axial arthrodesis. The patient recovered neurological function and was able to walk.

Posterior arthrodesis between the atlas and the axis was performed with sublaminar wires and two autogenous corticocancellous rectangle-shaped grafts from the posterior aspect of the ilium. Posterior occipitocervical arthrodesis, sometimes accompanied by removal of the posterior arch of the atlas, involved the wiring of two autogenous corticocancellous iliac-crest grafts from the occiput to the second cervical vertebra[85]. Posterior subaxial arthrodesis was performed by wiring of the spinous processes and corticocancellous bone-grafting. Anterior subaxial decompression consisted of removal of the portion of the vertebral bodies that caused the neural compression and arthrodesis with tricortical autogenous iliac-crest graft.

The decision as to which postoperative orthosis to use was based on intraoperative assessment of the over-all stability of the arthrodesis construct. A cervicothoracic orthosis was used in most patients for three months. A halo vest was used when the surgeon judged the fixation to be tenuous.

Analysis of the Data

Radiographic measurements from the patients who did not have a neurological deficit (Class I) and from the patients who did have a neurological deficit (Classes II and III) were compared to determine the most reliable predictors of paralysis. The occurrence and degree of neurological recovery in the operatively managed

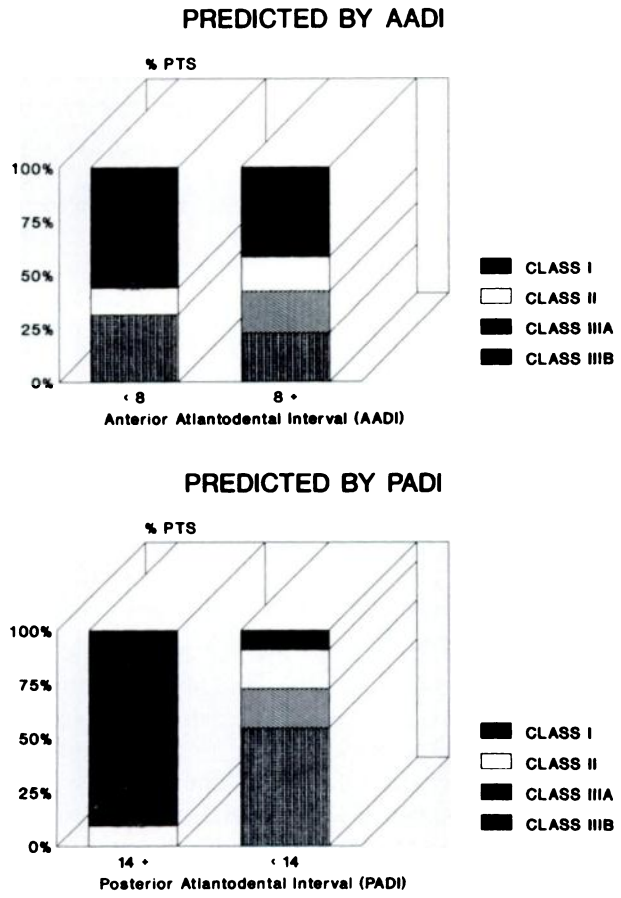

FIG. 2

Prediction of the development of paralysis in seventy-three patients who had rheumatoid arthritis. While the anterior atlanto-odontoid interval (AADI) did not correlate well with paralysis ($p > 0.10$), a posterior atlanto-odontoid interval (PADI) of fourteen millimeters clearly demarcated neurologically intact patients (Class I) from those in whom paralysis developed (Classes II and III) ($p = 0.000001$).

patients were analyzed on the basis of several variables: duration of paralysis, initial neurological class, anatomical location of the disease, and preoperative radiographic parameters. Comparison of the means of radiographic measurements between the neurological classes and determination of the effect of the radiographic parameters on the prediction of paralysis and recovery were made with use of analysis of variance (Stats+; Statsoft, Tulsa, Oklahoma). Differences in the mortality rates were compared with the chi-square test.

Results

Radiographic Analysis

Seventy-three patients were followed for a minimum of two years or until the time of death. Over-all, ten patients had isolated subaxial subluxation; sixty-one patients had atlanto-axial subluxation, twenty with basilar invagination and one with subaxial subluxation; and two patients had isolated basilar invagination. Of the forty-two patients in whom a neurological deficit developed, thirty-two had atlanto-axial subluxation, nineteen with basilar invagination and one with subaxial subluxation (Tables I and II). Eight of the patients who had paralysis had subaxial subluxation alone (Table II).

The anterior atlanto-odontoid interval, as measured on the radiographs of the cervical spine, showed a wide range of values within each class of paralysis; the interval was three to sixteen millimeters in Class I, three to

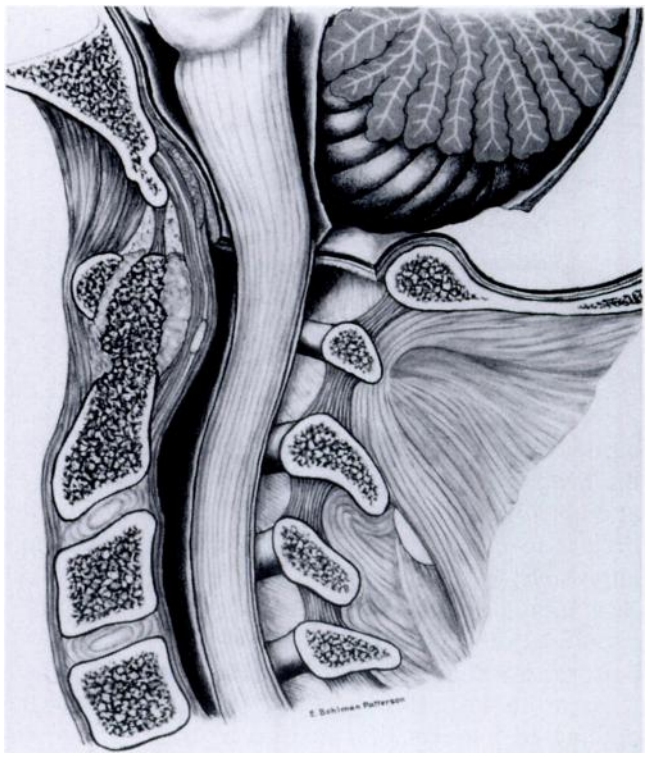

FIG. 3-A

Figs. 3-A through 3-D: Case 23. Erosion of the odontoid process.

Fig. 3-A: Illustration of the lateral view of the upper cervical spine in the early stages of involvement by rheumatoid disease, demonstrating pannus formation of the atlanto-axial joint eroding the odontoid process and dissolving the transverse ligament.

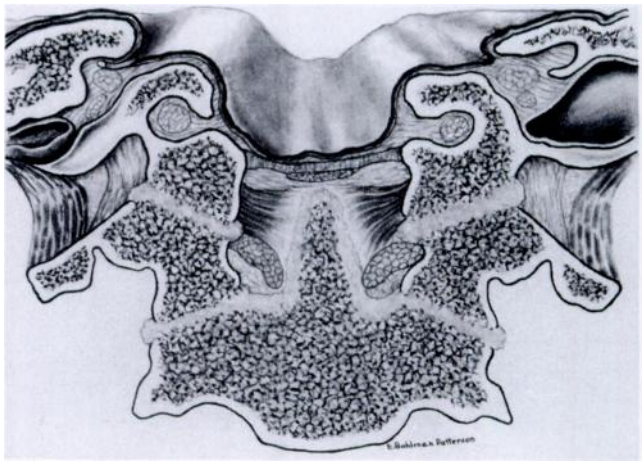

FIG. 3-B

Illustration of the erosions of the lateral mass of the atlas as well as destruction of the occipitocervical and atlanto-axial joints in the early stages of involvement.

ten millimeters in Class II, five to sixteen millimeters in Class IIIA, and three to fourteen millimeters in Class IIIB (Table III). We could not detect a significant difference ($p > 0.10$) in the anterior atlanto-odontoid interval among the different classes of paralysis with the small number of patients in our study. The mean anterior atlanto-odontoid interval for the patients in Class IIIA was 9.4 millimeters, slightly larger than for those in the other classes (7.5, 7.2, and 7.9 millimeters), but this was due to a single outlier in this small group of patients.

The posterior atlanto-odontoid interval showed excellent correlation ($p = 0.000001$) with the severity of paralysis. Of twenty-four patients who had atlanto-axial subluxation and a Class-III neurological deficit (Figs. 1-A through 1-D), twenty-three had a posterior atlanto-odontoid interval of thirteen millimeters or less (average, ten millimeters); the remaining patient had fifteen millimeters of basilar invagination. The patients who had a Class-II deficit or a Class-I rating had mean posterior atlanto-odontoid intervals of thirteen and fifteen millimeters, respectively. A posterior atlanto-odontoid interval of less than fourteen millimeters predicted the

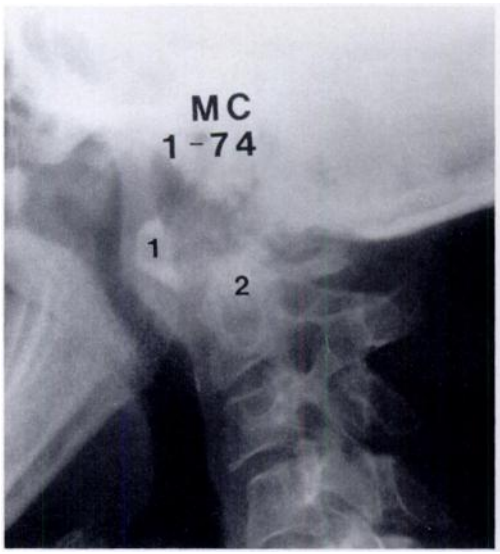

FIG. 3-C

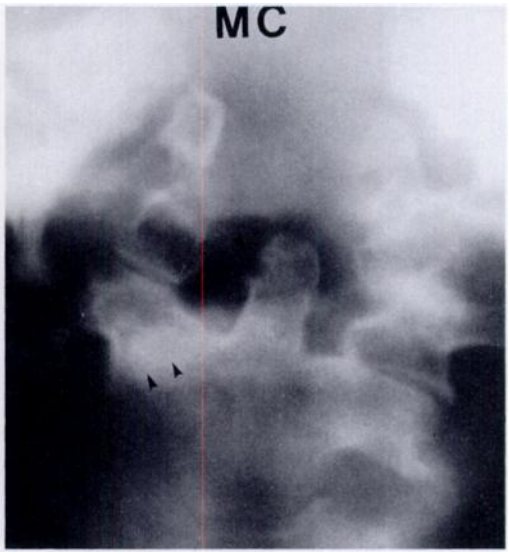

FIG. 3-D

Fig. 3-C: Lateral radiograph of a fifty-four-year-old woman who had had rheumatoid arthritis for at least twenty-five years. Note the erosion of the odontoid process and subluxation of the atlanto-axial joint. The patient was quadriparetic and unable to walk.

Fig. 3-D: Anteroposterior tomogram demonstrating erosions of the odontoid process and atlanto-axial joint (arrowheads).

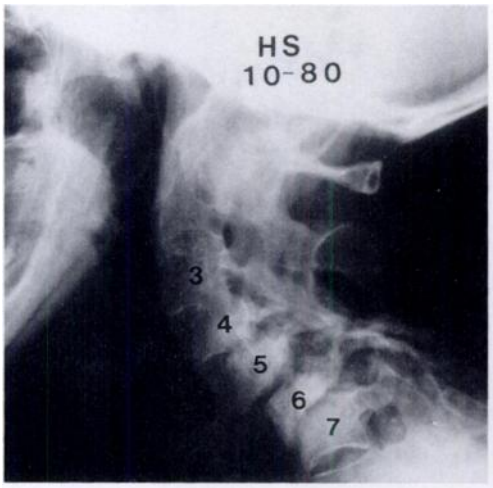

FIG. 4-A

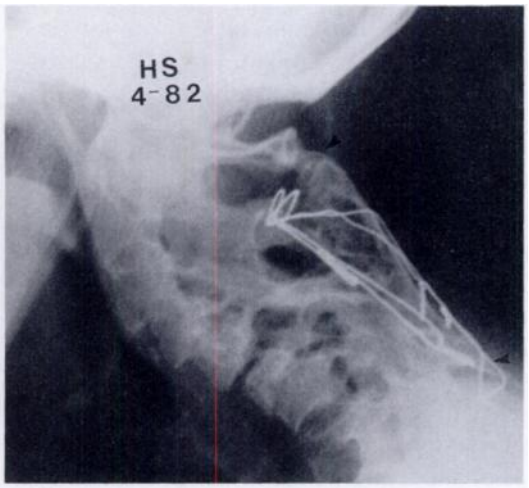

FIG. 4-B

Figs. 4-A and 4-B: Case 34.

Fig. 4-A: Lateral radiograph of a fifty-year-old patient who had had rheumatoid arthritis for many years, with progressive multiple-level subaxial subluxations from the third through seventh cervical vertebra. Over a period of two years, slowly progressive cervical myelopathy had developed and the patient was quadriparetic and unable to walk. Note the erosions of the facet joints and vertebral bodies by rheumatoid pannus.

Fig. 4-B: The patient was placed in skeletal traction that improved the quadriparesis, and a posterior cervical arthrodesis from the second cervical to the first thoracic vertebra was performed with autogenous iliac bone and wire. There was a solid posterior fusion (arrowheads) eighteen months after the operation. The patient recovered complete neurological function and became able to walk without the use of aids.

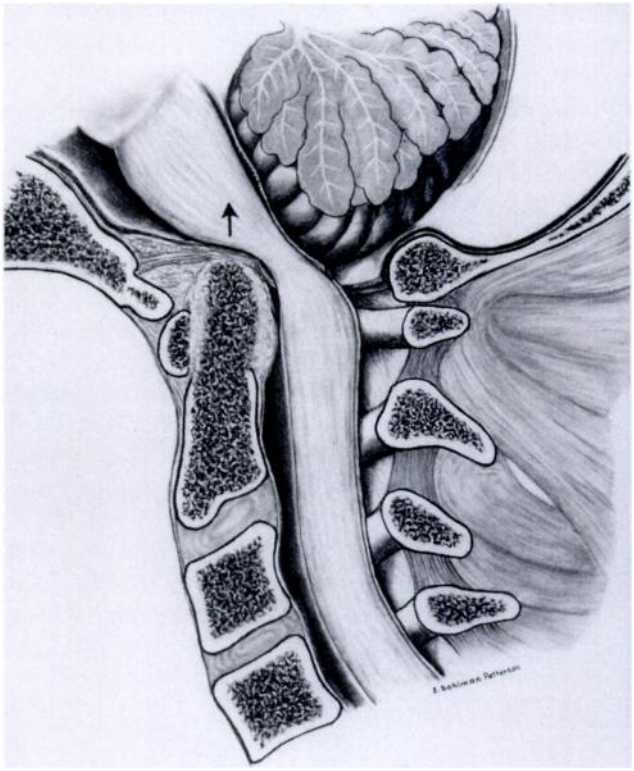

FIG. 5-A

Figs. 5-A through 5-G: Case 10. Basilar invagination.

Fig. 5-A: Illustration of the lateral view of the upper cervical spine, demonstrating basilar invagination (arrow) of the odontoid process into the foramen magnum. Note the compression of the spinal cord.

development of paralysis more accurately than did a large anterior atlanto-odontoid interval (Fig. 2).

Tomography was used to determine the severity of basilar invagination as well as the presence of osseous erosions. Of the sixty-one patients who had atlanto-axial subluxation, twenty-five had marked erosion of the odontoid process (Figs. 3-A through 3-D). The site of osseous destruction was most commonly on the postero-inferior surface of the odontoid process.

In the patients who had a subaxial subluxation, the diameter of the subaxial sagittal canal reflected the presence and degree of paralysis more often than did the percentage of vertebral slip (Table II). All patients who had a Class-III deficit had a diameter of the canal of thirteen millimeters or less (Figs. 4-A and 4-B). All patients who had a rating of Class I or II had a diameter of at least fourteen millimeters.

The prevalence of paralysis increased dramatically in the patients who had concurrent basilar invagination and atlanto-axial subluxation (Figs. 5-A through 5-G). Of the forty patients who had isolated atlanto-axial subluxation, a neurological deficit developed in twelve (30 per cent) (Table II). In contrast, paralysis developed in nineteen of the twenty patients who had combined atlanto-axial subluxation and basilar invagination (Table II). Only one patient who had a Class-I rating (no deficit) had radiographic evidence of basilar invagination, and this was not associated with substantial atlanto-axial subluxation.

Treatment and Predictors of Outcome

Paralysis did not develop in thirty-one patients who were followed and received periodic management for symptoms with a soft cervical collar and modification of the medications for systemic rheumatoid arthritis. Of the forty-two patients in whom a neurological deficit did develop, seven were managed non-operatively (Table II). Four patients refused an operation and were managed with a soft cervical collar; one patient who had

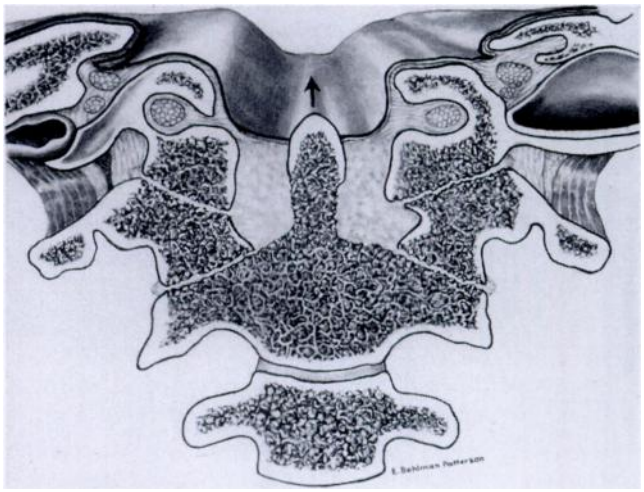

FIG. 5-B

Illustration of an anterior-posterior view of basilar invagination (arrow) of the odontoid process into the foramen magnum. Note the erosions and partial dissolution of the lateral masses of the atlas, allowing cephalad migration.

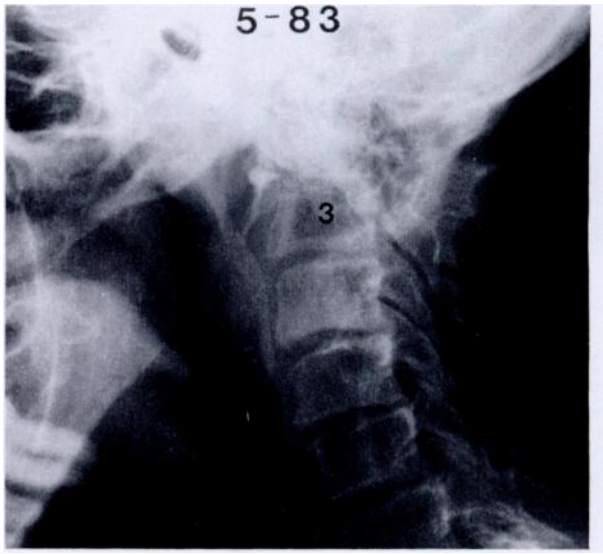

FIG. 5-C

Lateral radiograph of a fifty-seven-year-old woman who had had rheumatoid arthritis for many years. Multiple joint replacements had been performed. Note the basilar invagination of the second cervical vertebra into the skull, with the third cervical vertebra being the first visible vertebra. The patient was mildly quadriparetic.

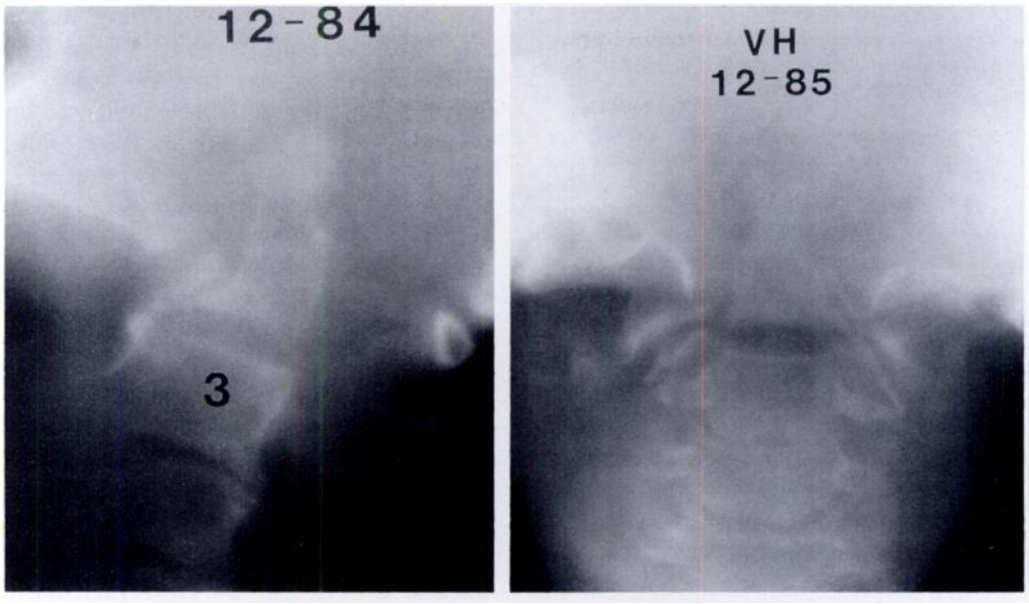

FIG. 5-D FIG. 5-E

Fig. 5-D: Lateral tomogram demonstrating complete migration of the odontoid process and the body of the axis into the foramen magnum and partial dissolution of the anterior arch of the atlas.

Fig. 5-E: Anterior-posterior tomogram demonstrating basilar invagination of the axis into the skull. Note the total dissolution of the lateral masses of the atlas.

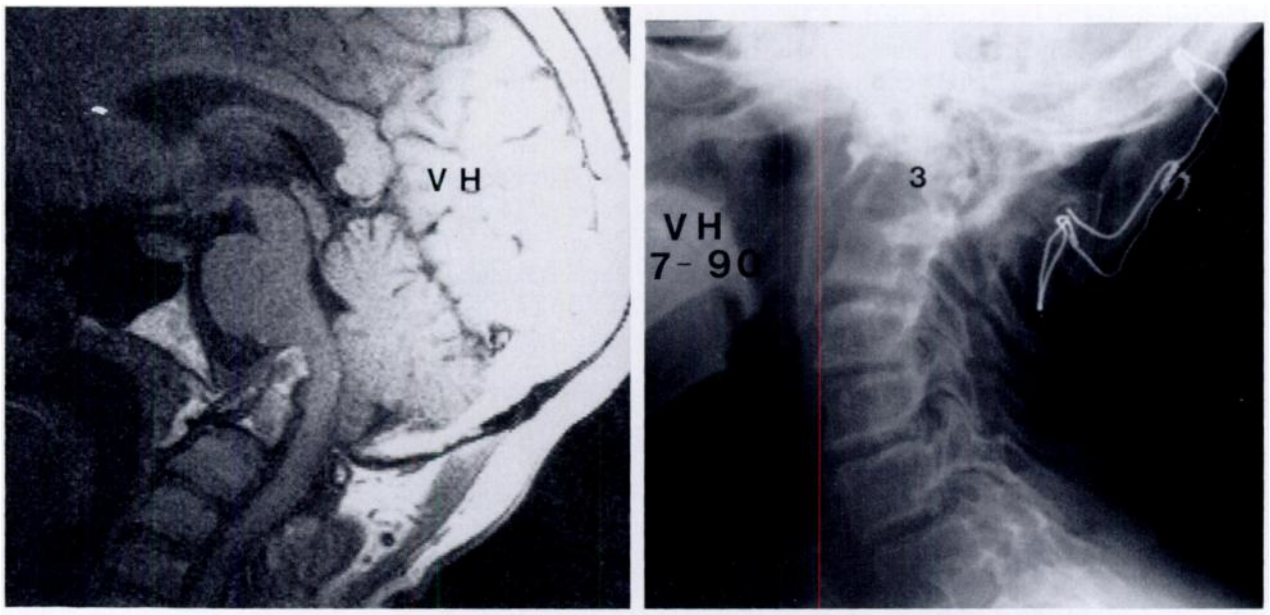

FIG. 5-F FIG. 5-G

Fig. 5-F: Sagittal magnetic-resonance scan demonstrating severe compression of the brain stem from the basilar invagination of the odontoid process. Note the relationship of the body of the axis to the clivus anteriorly.

Fig. 5-G: Lateral radiograph demonstrating solid cervical-occipital fusion; the operation was done to relieve cervical myelopathy. The patient recovered normal neurological function and was able to walk without aids.

severe quadriparesis died while in skeletal traction before an operation could be done; and the other two patients were deemed medically unstable. Three of the patients who were managed non-operatively died within one month after the onset of paralysis, two from compression of the spinal cord. The other four patients died within four years after the diagnosis, three from compression of the cord and one from cardiac disease. None of the patients who were managed without operative decompression or stabilization had neurological improvement. Four deteriorated at least one neurological class, and two others had progressive paralysis but did not change class because they had been in Class IIIB to begin with.

TABLE IV

MORTALITY RATE FOR PATIENTS MANAGED OPERATIVELY RELATIVE TO ORIGINAL NEUROLOGICAL CLASSIFICATION

Class[67]	No. of Patients	Average Duration of Follow-up (Mos.)	No. of Patients Who Died	Average Time from Consultation to Death (Mos.)
II	9	74	2	68
IIIA	5	145	3	10
IIIB	21	111	13	29

Thirty-five patients who had paralysis had operative stabilization or decompression, and twenty-five (71 per cent) improved by at least one neurological class, nine (26 per cent) improved by two classes, eight (23 per cent) remained unchanged, and two deteriorated by one class. The initial neurological class was related to neurological recovery (Fig. 6). All nine of the patients who had a Class-II deficit had resolution of subjective motor weakness. Of the twenty-six patients who had a Class-III deficit, sixteen (62 per cent) improved, eight (31 per cent) were unchanged, and two (8 per cent) worsened. The initial neurological class was also significantly related to the mortality rate ($p < 0.01$), as two of the nine patients who initially had a Class-II deficit and sixteen of the twenty-six patients who had a Class-III deficit died during the study period (Table IV). The mean age of the patients who died (sixty-one years) was not significantly different from the mean age of all of the patients studied (sixty years). We were unable to document the total number of patients who had rheumatoid arthritis and were seen at the two institutions over the twenty-year period; therefore, the prevalence of paralysis and the mortality rate in patients without involvement of the cervical spine were not calculated.

The preoperative duration of paralysis was not shown to affect the prognosis for neurological recovery after the operation. Ten of the fifteen patients who had an operation within two months of the onset of paralysis, ten of the thirteen patients who had an operation three to twelve months after the onset of paralysis, and five of the seven patients who had an operation more than one year after the onset of paralysis improved at least one neurological class following the operation.

The anatomical location of the disease had some effect on the prognosis for neurological recovery. Seven of the nine patients who had isolated atlanto-axial subluxation improved by at least one neurological class after the operation. Six of the seven patients who had subaxial subluxation and eleven of the nineteen who had combined atlanto-axial subluxation and basilar invagination improved by at least one neural class. Four of the five patients who had subaxial subluxation and a Class-III neurological deficit improved by at least one class. In contrast, only twelve of the twenty-one patients who had atlanto-axial subluxation alone or in combination with basilar invagination and had a Class-III deficit improved.

In the patients who had atlanto-axial subluxation, the most important predictor of the potential for neurological recovery was the preoperative radiographic measurement of the posterior atlanto-odontoid interval (Fig. 7). Of the patients who had isolated atlanto-axial subluxation, some recovery occurred in all patients in whom the posterior atlanto-odontoid interval was at least ten millimeters. In contrast, when the posterior atlanto-odontoid interval was less than ten millimeters, no neurological recovery occurred and both of the patients died. Of the patients who had concurrent basilar invagination, there was neurological recovery in all patients in whom the posterior atlanto-odontoid interval was at least thirteen millimeters, and no marked

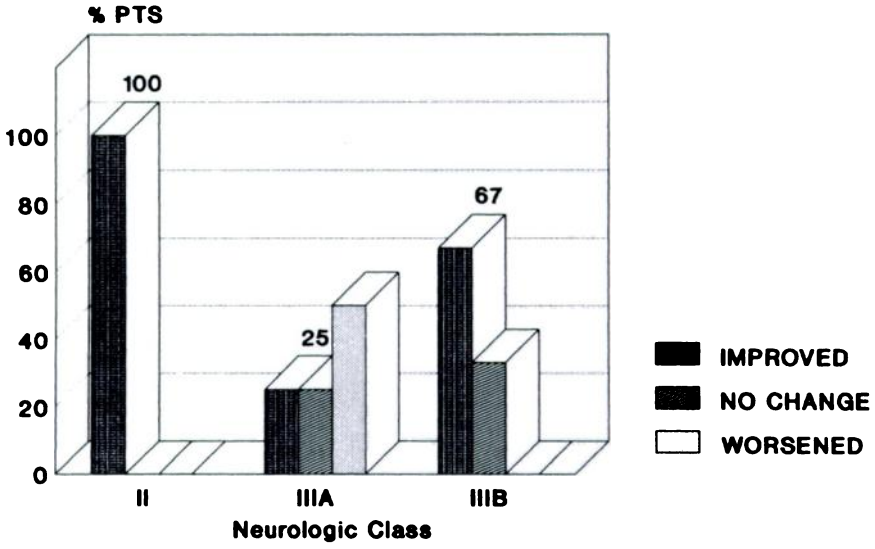

FIG. 6

Neurological recovery postoperatively in thirty-five rheumatoid patients, grouped by the original neurological class.

recovery occurred when the interval was less than ten millimeters.

Neurological recovery in the patients who had subaxial subluxation was related to the achievement of adequate decompression of the spinal canal. Three patients improved by two classes, three improved by one class, and one did not improve. Two patients had persistent Class-III deficits after the operation, while the remainder improved to Class I or II. In the two patients who had little or no recovery, the postoperative diameter of the subaxial canal was less than fourteen millimeters. Both patients had a posterior arthrodesis with incomplete reduction of the subluxation.

Operative Complications

Pseudarthrosis developed in two patients who had an onlay posterior occipitocervical arthrodesis without fixation with wire; presumably this was because they had a stable fibrous union. Neither needed repair. Pseudarthrosis developed between the first and second cervical vertebrae in a third patient after arthrodesis from the occiput to the fourth cervical vertebra. Accordingly, the rate of pseudarthrosis was 9 per cent (three of thirty-five patients). Three patients needed an extension of the posterior arthrodesis to include the occiput because of progressive basilar invagination. Three patients who had a transoral odontoid resection had progressive paralysis, and two died, from compression of the spinal cord, within four weeks after the operation; the third died eight months later from progressive vertical migration of the second cervical vertebra. Three other patients also died within six months after the operation, one from a bowel infarction and compression of the cord, one from a perforated ulcer with peritonitis, and one from an unknown cause. All six of these patients had severe Class-IIIB quadriparesis before death.

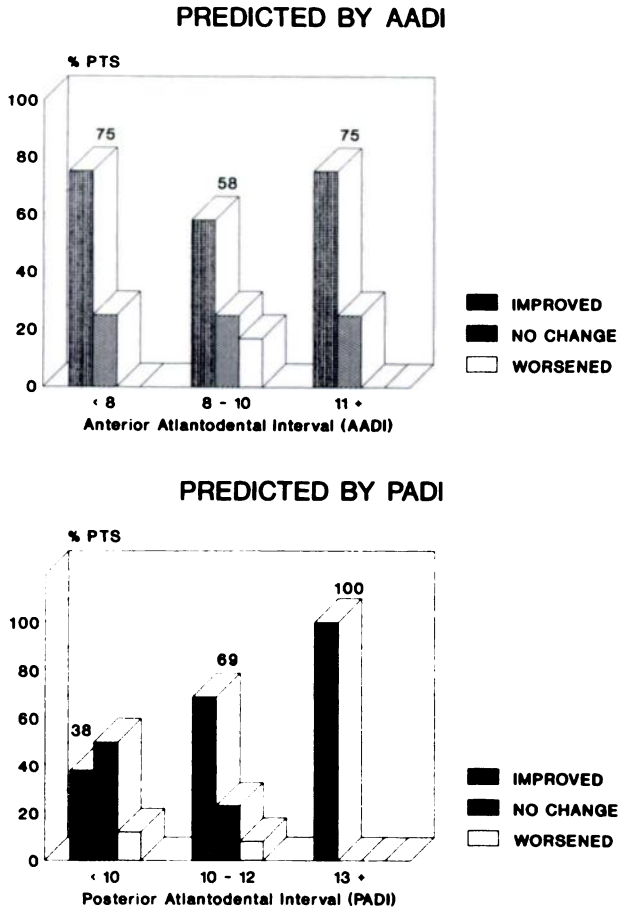

FIG. 7

Neurological recovery postoperatively in thirty-five rheumatoid patients, grouped by preoperative radiographic measurements. Although the anterior atlanto-odontoid interval (AADI) did not correlate with neurological recovery ($p > 0.10$), the posterior atlanto-odontoid interval (PADI) showed a strong correlation with recovery ($p = 0.0001$).

Discussion

Paralysis is a severe and a sometimes fatal complication in patients who have rheumatoid arthritis of the cervical spine[20,40,53,55,58,61,77,78,82,83,88]. Early neurological changes may be difficult to detect in patients who already have muscle weakness and atrophy secondary to chronic rheumatoid arthritis. Hyperreflexia may be masked due to severe disease of peripheral joints[8,45,47]. Similarly, the clinical assessment of neurological deterioration secondary to compression of the brain stem may be difficult to discern from compression of the vertebral artery with insufficiency of the basilar artery or from cranial-nerve palsies[14,33,42,50]. There is currently no reliable method to predict which patients who have radiographic evidence of instability will have progression of the cervical involvement[64]. Since some of the more severe deficits are not reversible, it would be advantageous to predict which patients are at the highest risk for paralysis and to perform operative stabilization before the onset of any irreversible neurological changes[11]. Although the literature is replete with reports of the results of various techniques for operative stabilization, the precise indications for an operation in patients who do not have a neurological deficit remain unclear.

A central problem is reflected by continuous reports emphasizing that the radiographic degree of subluxation does not reliably correspond to the development of neurological deficits[12,15,39,54,65,74]. The anterior atlanto-odontoid interval has traditionally been reported as the determinant of atlanto-axial instability. In the present series, this measurement showed poor correlation with neurological deficit; however, the posterior atlanto-odontoid interval was a reliable predictor of the development and severity of paralysis. This latter measurement more accurately reflects the space available for the spinal cord. While the posterior atlanto-odontoid interval has been described previously, it has not been evaluated in a large series of rheumatoid patients[72,86,87].

In the present series, nineteen (60 per cent) of thirty-two patients who had atlanto-axial subluxation and paralysis had an anterior atlanto-odontoid interval that

did not equal or exceed the common operative criteria of nine millimeters[11,13,65,84]. This weak correlation of the traditional anterior measurement may be explained by variation in the diameter of the atlas, as well as pannus formation behind the odontoid process. In contrast, all but one of the patients who had a Class-III deficit had a posterior atlanto-axial interval of thirteen millimeters or less (Fig. 2). Similarly, the diameter of the subaxial canal was found to correlate more closely with paralysis than with the degree of displacement of one vertebral body on another. All eight patients who had subaxial subluxation and a Class-III deficit had a diameter of the canal of thirteen millimeters or less. The critical diameter of the canal of thirteen millimeters was consistent with data from cervical spondylotic myelopathy as well as magnetic resonance-imaging studies in rheumatoid patients[4,6,7,22,23,43,44,63,71,86]. A unique component of rheumatoid arthritis, as compared with cervical spondylosis, is the variable soft-tissue pannus that can result in symptomatic compression of the cord before the osseous canal reaches a critical threshold diameter[90].

Our series confirmed previous reports that basilar invagination is associated with a higher frequency of severe neurological deficits that may not be reversible and can be fatal[19,25,37,69,72,73,79]. All but two of our patients who had paralysis and basilar invagination also had atlanto-axial subluxation. The severity of basilar invagination beyond five millimeters did not appear to affect the prognosis as much as the degree of atlanto-axial subluxation as reflected by the posterior atlanto-odontoid interval. Of note is the observation that none of the patients who remained intact neurologically had basilar invagination and marked atlanto-axial subluxation. Accordingly, when any evidence of basilar invagination is detected in patients who have marked atlanto-axial subluxation, early operative stabilization should be considered before the onset of neurological deficit.

The operative treatment of the unstable cervical spine in patients who have rheumatoid arthritis has been well documented[1,5,14,16,17,21,29,31,39,54,59,74,80]. Although our investigation was not prospective or randomized and our goal was not to evaluate the merits of any particular operative technique, several observations warrant elaboration. Of the seven patients in our series who refused or were unable to have a reduction and operative stabilization, none had any neurological improvement; all had progressive paralysis and eventually died (five from compression of the cord and two from cardiac disease). Three of the patients who were managed operatively were referred to us after failure of previous treatment with posterior methylmethacrylate fixation without a bone graft. These failures emphasize the need for that technique to be used with a bone graft as has been reported[9,11,24,48]. The results of transoral odontoid resection in this series were very poor, as two of three patients had a progressive neurological deficit and the other had no improvement; all three died[34,70,75]. The poor results in these patients may be due to the severity of the pre-existing disease rather than to the procedure itself, as others have reported more favorable results in less severely ill patients[11,18,57]. Alternatively, the extrapharyngeal approach for resection of the odontoid may be used[49]. Finally, although posterior arthrodesis for subaxial subluxation has been advocated instead of anterior arthrodesis[67], anterior decompression and arthrodesis in the present series was sometimes performed due to inadequate reduction of the subluxation with a posterior approach. Although previous authors believed that decompression was not needed in a rheumatoid patient who had myelopathy if a solid fusion were achieved, our results did not support this hypothesis[38]. When combined posterior arthrodesis and anterior decompression are indicated, we recommend that the posterior procedure be performed first. If the posterior arthrodesis results in only a partial reduction and a diameter of the subaxial canal that is still less than fourteen millimeters, an anterior decompression should also be considered.

Important variables in the prediction of neurological recovery after an operation for the unstable cervical spine in a patient who has rheumatoid arthritis have not been previously elucidated. Although Conaty and Mongan suggested that a delay in the diagnosis diminishes the chance for a satisfactory outcome[14], our data did not support this finding. Although no delay in diagnosis is theoretically advantageous, a longer duration of paralysis did not preclude substantial neurological recovery in our patients. The duration of paralysis was not a major predictor of recovery, but the severity of myelopathy at the time of the operation was important. More severe preoperative paralysis was associated with less recovery and a higher mortality rate over the long term.

The anatomical location of the impingement of the spinal cord was also relevant to neurological recovery. Although the patients who had basilar invagination tended to have more severe preoperative neurological deficits, the patients who had Class-III deficits and subaxial subluxation demonstrated better neurological recovery than the patients who had Class-III deficits and basilar invagination.

In patients who had atlanto-axial subluxation alone or with basilar invagination, the most dramatic predictor of neurological recovery after the operation was found to be the posterior atlanto-odontoid interval as measured on the preoperative radiograph. We were able to identify specific thresholds that helped to define the boundaries for potential neurological recovery. Patients can have neurological deficits before this threshold value is reached because of the contribution of soft-tissue pannus, but certainly the potential risk for compression of the cord is increased once these thresholds are reached. In patients who had paralysis, no substantial or predictable recovery occurred if the preoperative posterior atlanto-odontoid interval was less than

ten millimeters. In patients who had paralysis due to atlanto-axial subluxation alone, recovery of at least one neurological class always occurred when the posterior atlanto-odontoid interval was at least ten millimeters. If basilar invagination was superimposed, consistent neurological recovery only occurred when the posterior atlanto-odontoid interval was at least thirteen millimeters. All patients who had paralysis and a posterior atlanto-odontoid interval or diameter of the subaxial canal of fourteen millimeters had complete motor recovery after the operation. Fourteen millimeters appears to be a reasonable criterion for prophylactic operative stabilization, since the most severe deficit seen with this measurement was Class II and full motor recovery was realized in all of these patients after the operation.

The primary goal of arthrodesis in a patient who has rheumatoid arthritis and an unstable cervical spine is to prevent irreversible neurological compromise. Although we cannot prove that those patients would have a greater recovery with an earlier operation, our observations in patients in whom paralysis did not develop defined the range of the posterior atlanto-odontoid interval associated with low neurological risk, and our observations in patients who had residual deficits after the operation defined the limits of neurological recovery on the basis of this measurement. To minimize the potential risk of irreversible paralysis, we recommend operative stabilization of the rheumatoid cervical spine, in the presence or absence of a neurological deficit, for patients who have atlanto-axial subluxation and a posterior atlanto-odontoid interval of fourteen millimeters or less, patients who have atlanto-axial subluxation and at least five millimeters of basilar invagination, and patients who have subaxial subluxation and a sagittal diameter of the spinal canal of fourteen millimeters or less.

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