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ORIGINAL ARTICLE

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Atlantoaxial immobilization in rheumatoid arthritis: a prophylactic procedure?

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Abstract Timing of surgical intervention in atlantoaxial instability due to rheumatoid arthritis is still controversial. An aim of this study was to investigate whether atlantoaxial fusion can prevent progression of instability and upward migration of the dens. Thirty-two patients with rheumatoid arthritis, who underwent posterior atlantoaxial fixation due to instability, were clinically and radiologically examined after a minimum follow-up of 5 years. The radiological measurements focussed on the extent of cranial vertical migration after atlantoaxial fusion. In none of the 20 patients available for follow-up examination was a vertical cranial migration observed, in spite of the ongoing course of the disease. These findings are in concordance with findings in the literature, and strongly suggest that, with atlantoaxial stabilization, the inflammatory process with destruction of the lateral masses of the atlas is able to prevent further deterioration with vertical cranial migration.

Key words Atlantoaxial instability · Rheumatoid arthritis · Indication for surgery · Vertical migration

Introduction

The pathophysiology of rheumatoid arthritis (RA) involves inflammation of the synovial membrane. This leads to disintegration and destruction of bony, cartilaginous and ligamentous structures, and subsequent instability and deformation of a particular joint in a spinal unit. In the upper cervical spine, this process causes transverse atlantoaxial instability by insufficiency of the ligamentous apparatus, mainly the transverse and alar ligaments. Vertical instability or cranial settling is caused by bony destruction of the lateral masses of the atlas, allowing the dens to enter the cranium by decreasing the distance between the atlantoaxial joints and the base of the skull. This causes entrapment of the dens in the foramen and reduces the translational dislocation between the atlas and the axis. Erroneously, this fact may be interpreted as improvement and "spontaneous healing" of the atlantoaxial instability. In reality, there is a worsening of the situation, due to increasing pressure on the spinal cord and brainstem from the tip of the dens [22,25].

Vertical and horizontal instability of the suboccipital area present clinically with a variety of symptoms. Pain and (later on) myelopathic signs and symptoms are the most obvious features, but are not encountered consistently. Surgical procedures with decompression, reduction and stabilization are generally accepted if there is severe pain, progressive instability or increasing neurological symptomatology. Controversy exists as to the appropriate treatment in patients with radiographically confirmed instability but an absence of, or only mild, neck pain and no neurological symptoms. In this situation, most authors accept the clinical and radiological observations [22,25], in spite of the fact that progression of instability and deformity has been documented [27,28]. According to these authors, surgical intervention and stabilization is only indicated if there is atlantoaxial instability of more than 8 or 9 mm of the anterior atlanto-dental interval (ADI) [7, 10,30] or less than a 10-mm posterior ADI [4]. Although

aiming to avoid unnecessary surgery, this speculative approach may lead to late interventions, which have proved to worsen the outcome [1, 5, 29,30]. Once upward migration has occurred, the mortality rate [32] increases drastically, even after adequate surgery with decompression and stabilization.

The determination of timing of atlantoaxial arthrodesis in rheumatoid patients with C1/2 instability remains a crucial issue with respect to cervical lesions in RA.

The aim of this study was to evaluate the influence of atlantoaxial fusion on the progression of upward migration of the dens, and to discuss the potential prophylactic value of this procedure.

Materials and methods

Thirty-two patients with RA, as classified by the criteria of the American Rheumatism Association, and atlantoaxial subluxation were operated on using selective atlantoaxial fusion between 1987 and 1990. Patients who required more extensive surgery of the cervical spine, such as occipitocervical fusion, extended fusion to the lower cervical spine, or simultaneous anterior decompression, were excluded from the study. Twenty patients were able to attend a clinical and radiological examination, with an average follow-up of 82 (range 61–108) months. Five patients died in the follow-up period due to reasons unrelated to the surgery of their neck. Two patients were lost to follow-up, and five declared themselves unable (due to impairment caused by the disease; four patients) or unwilling (one patient) to undergo the follow-up examination.

In all 20 patients, the indication for surgery was atlantoaxial instability due to RA after unsuccessful adequate conservative treatment. With the exception of one patient who was reoperated because of pseudarthrosis after atlantoaxial wiring, none of the 20 patients had undergone previous surgery of the cervical spine. There were 13 women and 7 men. The average age at the time of surgery was 58 (range 34–71) years. The duration of the disease at the time of surgery ranged from 3 to 45 years (mean 17 years). The preoperative anterior atlanto-dental interval (ADI) averaged 10.5 mm (range 8–14 mm). Of the 20 patients, 5 were Ranawat stage II and 15 were Ranawat stage IIIA. Intractable pain was the indication for atlantoaxial fusion in 14 patients. In two patients, progressive instability with moderate pain, and in a further two, instability without pain indicated the fusion. There were no neurological deficits attributable to the atlantoaxial instability in this patient series. In no case was there any indication for additional decompressive procedures. An anatomical reduction of the translational instability in the extension radiographs was noted in all patients. The adjacent segments in the subaxial spine showed no pathology that would justify surgical stabilization. Therefore, in all 20 patients an isolated posterior atlantoaxial fusion was performed.

Operative technique

The fusion was performed by one surgeon according to the Magerl technique, which has been described extensively in the literature [14]. Two set-screws were inserted through the atlantoaxial facets by a posterior approach. An additional midline bone graft inserted between the atlas and the axis completed the procedure, providing optimal stability by a three-point fixation of the segment [13]. Prior to the screw insertion, a manual reduction of the translational dislocation was attempted. Postoperative external immobilization was performed with a soft collar for 6 weeks, whilst the preoperative medication for RA was re-established on the 1st postoperative day.

Radiographic evaluation

On the lateral radiographs of all patients, the postero-inferior rim of the vertebral body of the axis was clearly visible and was chosen for the distal point of reference. To overcome difficulties with different magnification factors in different radiographs, the length of the C2 endplate in the preoperative radiograph served as an individual reference. The subsequent measurements were calculated in relation to this reference for each patient. As the purpose of the study was to investigate the amount of cranial vertical migration after atlantoaxial fusion, a point of reference on the occiput had to be defined. However, the preoperative lateral radiographs did not uniformly show the hard palate necessary to apply the measurement technique of McGregor [23], and it was not possible to identify the foramen magnum with satisfactory precision on all the radiographs. In contrast, the tip of the mastoid processes were visible with sufficient accuracy on all the radiographs. In lateral radiographs, the projection of one process was overlying the other and the caudal summit of the contour was chosen as point of reference. In radiographs without a true lateral view, and therefore double projection of the processes, the point of reference was defined as the midpoint of a line connecting the summits (Fig. 1, Fig. 2).

Measurements were performed after digitization of the radiographs, and distances were measured in pixels. The evaluation of all pre-and postoperative radiographs was performed by the same person, thus minimizing errors resulting from different definitions of the anatomical landmarks. These measurements were made by a paramedic who was not informed of the purpose of the study.

The values pre- and postoperatively are mean values based on the available radiographs (1) from the period covering a maximum of 6 (range 0–6) months before surgery, (2) between 2 and 12 weeks following surgery, and (3) at follow-up, an average of 82 (range 61–108) months.

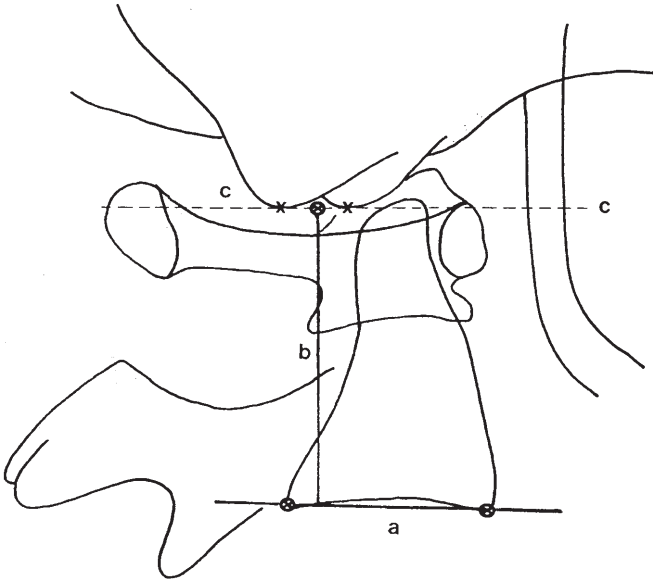

Fig. 1 Technique of radiological measurement. After digitization of the radiographs, the distances between the postero-inferior border and the occiput were measured. The baseline of C2 (*a*) served as reference to exclude errors due to different magnification factors of the radiographs. In true lateral projections, the summit of the two overlying projections of the mastoid process served as point of reference. If two silhouettes of mastoid process were visible, due to slight rotation, the midpoint of a line connecting the two summits (*c*) was the point of reference for the distance *b*

Fig. 2 Radiograph showing the technique of measurement according to explanation in Fig. 1

Clinical evaluation

The general course of the disease following surgery of the cervical spine was evaluated by questionnaires sent to patients and their treating physicians. The use of medication, the number of recurrent flare-ups and the general course of disability were assessed.

Results

In all 20 patients, a radiographically solid fusion was achieved with no apparent motion on functional radiographs. The anterior ADI was reduced to the anatomic position (not more than 2 mm) in 14 patients; 6 patients had a remaining ADI of an average of 4 mm (range 3–5 mm). Neck pain was reduced from 7.8 (range 0–10) preoperatively to 2.1 (range 0–5) postoperatively on a ten-point visual analogue pain scale [34].

The evaluation by questionnaire of the treating physician revealed a progression of the disease in nine patients, while in nine patients a steady course with an unchanged number of flare-ups per year since surgery was observed. In two patients, a slight improvement with a decreased number of flare-ups was reported. Fourteen patients needed an unchanged number of intra-articular corticosteroid injections per year following their cervical surgery, and six required an increased number of injections. None of the patients had to undergo further surgery of the upper cervical spine.

The second questionnaire was completed by the patients themselves. In their subjective view, 4 noted an improvement in the course of the disease, 4 had a worsening and 12 reported unchanged symptoms. The results describing restrictions in the ability to perform daily activities preoperatively and at follow-up are shown in Table 1. “No or slight restriction” was defined as independence in most daily activities. “Moderate restriction” refers to independent walking (<15 min) and help with most of the household activities. If the patient was dependent on help for carrying out basic activities such as eating, dressing and personal hygiene, and was non-walking, the situation was graded as “severely restricted”. It was concluded from the results of these questionnaire items that the general course of the disease was steady or worsening during the follow-up period.

Evaluation of pain was done using a visual analogue pain scale (from 0 to 10). Preoperatively, the neck pain averaged 7.8 (range 0–10) and postoperatively 2.1 (range 0–5). Eighteen patients complained preoperatively about a painful noise during flexion and extension of their neck, which was eliminated in all patients at follow-up. Only 1 out of 11 patients who had complained about sleeping disturbances due to their neck pain preoperatively indicated difficulties in finding a comfortable position at nighttime at follow-up.

The results of the measurements of the two mastoid processes and the baseline of C2 are shown in Table 2. If

Table 1 Results of questionnaire items on restrictions in ability to carry out daily activities, preoperatively and at follow-up, to determine the general course of the disease

Level of restriction	Preoperatively (no. of patients)	Follow-up (no. of patients)
No/slight	4	4
Moderate	16	12
Severe	0	4

Table 2 Distances between the base of the vertebral body C2 and the mastoid process (in pixels). The values were measured on radiographs preoperatively, 2–4 weeks postoperatively and at follow-up (min. 5 years.) (Values not measurable due to insufficient quality of the radiographs or insufficient precision of identification are indicated as *not meas*)

Patient no.	Preop. Mean ± SD	Postop. Mean ± SD	Follow-up Mean ± SD
39590	178.8	183.40	180.27 ± 2.32
42894	179.5 ± 5.50	188.10	188.10 ± 9.90
48004	109.3	122.00	104.60
48009	171.7 ± 34.10	164.60 ± 0.70	190.80
51554	186.0	189.90	186.50 ± 4.07
66234	109.2 ± 17.20	153.00	134.70 ± 14.91
66648	179.3	176.40	172.00
17108	not meas.	125.00	99.20
22827	216.6	221.00	228.00
34941	197.7 ± 7.75	196.90	202.05 ± 30.75
52674	not meas.	200.00	206.90 ± 4.10
54005	210.0 ± 4.05	197.05 ± 0.05	283.30
56006	151.8 ± 2.55	148.15 ± 4.45	158.40
57710	228.0	223.70 ± 0.80	220.30
64412	173.2 ± 0.75	172.60	173.05 ± 0.75
26466	227.2	224.00	224.45 ± 0.45
36233	not meas.	215.75 ± 11.25	231.20 ± 9.03
42871	not meas.	270.80	279.40 ± 5.75
49248	not meas.	194.40	201.90
55357	179.4	198.60	190.87 ± 3.75

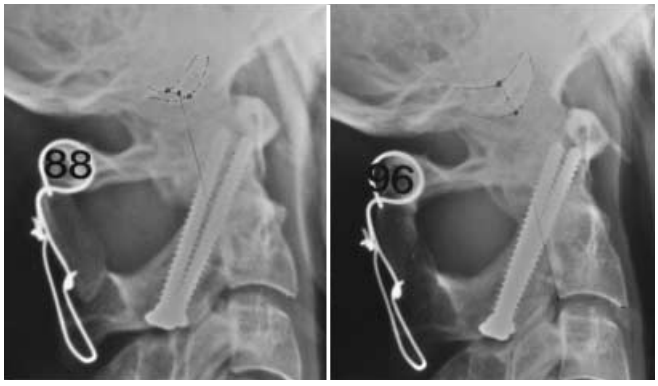

Fig. 3 A 54-year-old woman patient, R.S. Eight years after posterior atlantoaxial screw fixation and fusion, there is no change in the position of the atlantoaxial segment in relation to the occiput in spite of worsening of the clinical course of the disease

there was more than one radiograph available, the mean of the measured values is indicated together with the standard deviation.

In none of the patients was a progression of the upward migration of the dens noted at follow-up (Fig. 3, Fig. 4).

Complications

There were no intraoperative complications and no worsening of the neurological status. One patient had to undergo surgery of the lower cervical spine during the follow-up period because of painful instability of the segment C4/5. In one patient, the iliac bone graft was positioned too cranially and caused irritation with intermittent neck pain, but no intervention was considered necessary.

Discussion

The generally accepted advantage of atlantoaxial fusion in RA consists primarily of pain reduction and restoration of anatomical configuration, thus avoiding late complications, such as myelopathy due to continuous compression of the spinal cord. In addition, recent reports in the literature show that infectious tissue such as retrodental pannus disappears after atlantoaxial immobilization [16,36], in spite of the ongoing systemic course of the disease [16]. Peridental pannus represents hypertrophy of the synovial membrane, providing an inflammatory joint fluid containing several different types of enzyme, which, together with the pannus, cause joint destruction. The articular cartilage is slowly destroyed and the cartilage matrix is digested by the proteases produced by the synovial fluid. The integrity of the subchondral bone, joint capsule and adjacent ligamentous structures is threatened by direct destructive invasion of inflammatory tissue [2].

This process creates a tendency for progressive disintegration of the cervical spine, a finding that is well documented in the literature [3, 11, 26, 27,28]. Therefore, nonoperative management of C1/2 instability in RA is known to have a considerable failure rate, with figures of up to 50% for the mortality rate once myelopathy is present [21, 22, 24,35]. These fatal outcomes have been attributed to persistent repetitive trauma by both retrodental pannus and the dislocation and subsequent upward migration of the odontoid [9, 17, 21, 27, 28,29]. This has led to acceptance of surgical stabilization of the atlantoaxial instability. However, controversy remains in relation to the appropriate timing of surgery.

In the present study, atlantoaxial screw fixation proved to be an appropriate procedure without any complications

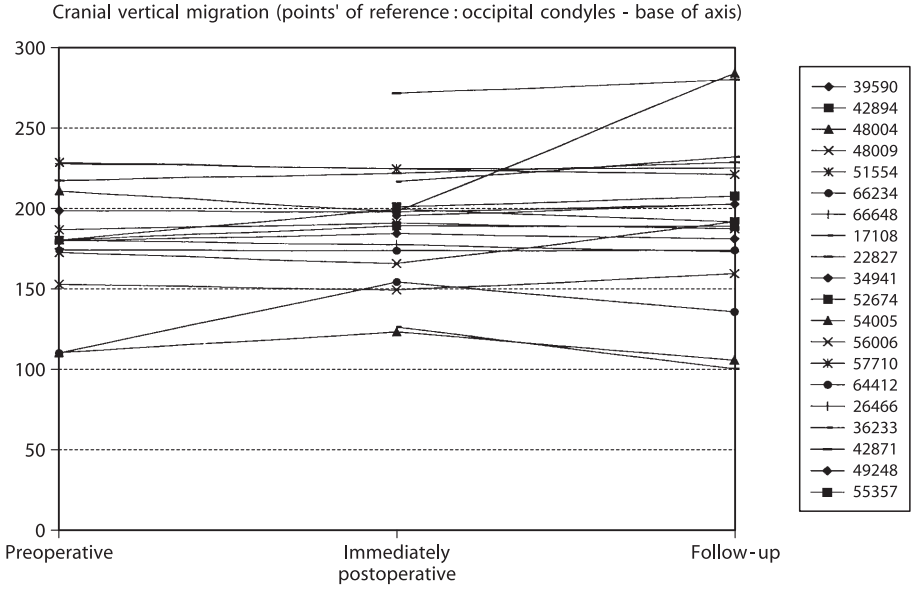

Fig. 4 Chart of the data presented in Table 2. The distance between the postero-inferior border and the occiput was measured pre- and postoperatively as well as at follow-up

directly related to the screw insertion. This technique has been in clinical use for almost 20 years [20], and has been shown to be superior in reducing transverse deformities of the C1/2 segment and in achieving solid bony union compared with conventional posterior wiring techniques [12,13].

Considering the fact that, after immobilization of the atlantoaxial segment, the pannus disappears, it might be hypothesized that the local process of destruction of bony and capsulo-ligamentous structures is similarly suppressed. The results of our study support this hypothesis: there is an absence of progression of the upward migration of the dens (as a consequence of loss of substance of the lateral masses of the atlas), in spite of the unchanged systemic course of the disease during the 5-year follow-up period. This conclusion has to be considered in the context of a follow-up rate of 62%. In addition, the natural course of the ongoing disease is not known individually, and therefore the true incidence of cranial migration in these patients without atlantoaxial fusion remains unknown. However, our results are supported by similar results published by Morizono et al. [26], where after atlantoaxial fusion of nine rheumatoid patients no changes of the Redlund-Johnell index were found. The same observation was made more recently by Kraus et al. [18] and Agarwal et al. [1]. They conclude that early stabilization can decrease the risk of progression of cervical instability and cranial migration.

Surgical atlantoaxial fusion in patients with inflammatory-induced instability therefore fulfils two purposes:

1. It eliminates pain and prevents continuous compression of the medulla by eliminating instability and atlantoaxial subluxation, and
2. It prevents progression of the deformity with cranial settling by pannus reduction and, thus, elimination of tissue destruction.

In conclusion, atlantoaxial fusion is not only a therapeutic modality, but represents a true prophylactic procedure. If the intervention is performed at a later stage of the disease, extensive surgery with anterior transoral decompression might be necessary to relieve pressure on the brain stem and allow neurological recovery. This implicates extension of the posterior fusion to the occiput. Late results reported in the literature of this extensive occipitocervical fusion are less favorable than isolated atlantoaxial fusions, due to postoperative decompensation of the subaxial cervical spine induced by increased lever arms [1, 7, 8, 15,18].

Several techniques to measure the upward migration of the dens in respect to the base of the skull [10,31], or as a change of the atlantoaxial relation [30], have been published. Most of these methods accept plain radiographs in neutral position or in flexion/extension as evidence of instability [6]. However, bony destruction or severe osteopenia due to RA may cause difficulties in interpretation of the radiographs. Landmarks such as the anterior or posterior cortex of the dens [10,31], the projection of the contours of the articular masses of C2 [30] or the tip of the dens [31] may not be clearly identifiable in all patients, thus influencing the reliability of the results.

Extended radiographic studies with, for instance, computed tomography (CT) or magnetic resonance imaging (MRI) are ideal to measure exact values of the individual anatomic situation [10, 19,33]. However, the problems with feasibility and availability at most centers, as well as the cost of these investigations, does not permit their wide use for patient screening and follow-up studies. In 1987, preoperative CT or MRI investigation was not available, therefore we had to rely on bony landmarks in conventional lateral radiographs. The tip of the mastoid process and the base of the vertebral body of the axis were clearly identifiable on all radiographs.

This study has drawbacks in relation to the clinical situation. We were not able to establish a comparative patient group. This scientifically accepted method had to be abandoned for ethical reasons. To expose a group of patients to a treatment less effective than a known accepted procedure was unacceptable in this situation of manifest and progressive atlantoaxial instability. However, the existence of a spontaneous benign course of the disease, which could have caused an inhibition of progression of bony destruction in the suboccipital area, was refuted by the results of the questionnaire administered to the physicians and patients reporting the clinical course of RA.

Conclusion

The results of this study serve to justify the expansion of indications for atlantoaxial arthrodesis in RA to an early stage of the disease, thus avoiding progression of instability with further deformity requiring more extensive surgery in the suboccipital area.

References

1. Agarwal AK, Peppelman WC, Kraus DR, Pollock BH, Stolzer BL, Eisenbeis CH, Donsaldson WF (1992) Recurrence of cervical spine instability in rheumatoid arthritis following previous fusion: can disease progression be prevented by early fusion? J Rheumatol 19:1364–1370
2. Allard SA, Muirden KD, Maini RN (1991) Correlation of histopathological features of pannus with patterns of damage in different joints in rheumatoid arthritis. Ann Rheum Dis 50:278–283
3. Awerbuch MS, Henderson DRF, Milazzo SC, White RG, Utturkar AB (1988) Long term follow-up of posterior cervical fusion for atlanto-axial subluxation in rheumatoid arthritis. J Rheumatol 8:423–432
4. Boden SC, Dodge LD, Bohlmann HH, Rechtine GL (1993) Rheumatoid arthritis of the cervical spine. Long term analysis with predictors of paralysis and recovery. J Bone Joint Surg Am 75:1282–1297
5. Casey AT, Crockard HA (1995) Vital reconstruction in rheumatoid arthritis. In: Baumgartner H, Dvorak J, Grob D, Munzinger U, Simmen BR (eds) Rheumatoid arthritis: current trends in diagnostics, conservative treatment, and surgical reconstruction. Thieme, Stuttgart New York, pp138–149
6. Clark CR (1995) The cervical spine in rheumatoid arthritis: natural history and diagnostic criteria for operation. In: Baumgartner H, Dvorak , Grob D, et al (eds) Rheumatoid arthritis. Thieme, Stuttgart New York, pp 132–138
7. Clark CR, Goetz DD, Menezes AH (1989) Arthrodesis of the cervical spine in rheumatoid arthritis. J Bone Joint Surg Am 71:381–391
8. Conaty JP, Morgans ES (1981) Cervical fusion in rheumatoid arthritis. J Bone Joint Surg Am 63:1218–1227
9. Crockard HA, Pozo JL, Ransford AO, Stevens JM, Kendall BE, Essigman WK (1986) Transoral decompression and posterior fusion for rheumatoid atlanto-axial subluxation. J Bone Joint Surg Br 68:350–356
10. Dvorak J, Grob D, Baumgartner H, Gschwend N, Grauer W, Larsson S (1989) Functional evaluation of the spinal cord by magnetic resonance imaging in patients with rheumatoid arthritis and instability of upper cervical spine. Spine 14:1057–1064
11. Fielding JW (1983) Rheumatoid arthritis of the cervical spine. pp 114–131
12. Grob D, Dvorak J, Gschwend N, Fröhlich M (1990) Posterior occipito-cervical fusion in rheumatoid arthritis. Arch Orthop Trauma Surg 110:38–44
13. Grob D, Dvorak J, Panjabi MM, Froehlich M, Hayek J (1991) Posterior occipito-cervical fusion. Spine 16:S17-S24
14. Grob D, Jeanneret B, Aebi M, Markwalder T (1991) Atlantoaxial fusion with transarticular screw-fixation. J Bone Joint Surg Br 73:972–976
15. Grob D, Schütz U, Plötz G (1999) Occipitocervical fusion in patients with rheumatoid arthritis. Clin Orthop 366: 46–53
16. Grob D, Würsch R, Grauer W, Sturzenegger J, Dvorak J (1997) Atlantoaxial fusion and retrodental pannus in rheumatoid arthritis. Spine 22:1580–1584
17. Henderson FC, Geddes JF, Crockard HA (1993) Neuropathology of the brainstem and spinal cord in end stage rheumatoid arthritis: implications for treatment. Ann Rheum Dis 52:629–637
18. Kraus DR, Peppelman WC, Agarwal AK, DeLeeuw HW, Donaldson WF (1991) Incidence of subaxial subluxation in patients with generalized rheumatoid arthritis who have had previous occipital cervical fusions. Spine 16:486–489
19. Lipson SJ (1989) Rheumatoid arthritis in the cervical spine. Clin Orthop 239: 121–127
20. Magerl F, Seemann P (1986) Stabile posterior fusion of the atlas and axis by transarticular screw fixation. In: Kehr P, Weidner A (eds) Cervical spine. Springer, Vienna New York, pp 322–327
21. Marks JS, Sharp J (1981) Rheumatoid cervical myelopathy. Q J Med 50:307
22. Mathews JA (1974) Atlanto-axial subluxation in rheumatoid arthritis. A five year follow-up study. Ann Rheum Dis 33:526–531
23. McGregor M (1948) The significance of certain measurements of the skull in the diagnosis of basilar impression. Radiology 21-B:171–181
24. Meijers KAE, Cats A, Kremer HPE, Luidendijk W, Onvlee GJ, Thomeer RTW (1984) Cervical myelopathy in rheumatoid arthritis. Clin Exp Rheumatol 2:239–245
25. Menezes AH, van Gilder JC, Clark CR, elKhoury G (1985) Odontoid upward migration in rheumatoid arthritis: an analysis of 45 patients with cranial settling. J Neurosurg 63:500–566
26. Morizono Y, Sakou T, Kawaida H (1987) Upper cervical involvement in rheumatoid arthritis. Spine 12:721–725
27. Oda T, Fujiwara K, Yonenobu K, Azuma B, Takahiro O (1995) Natural course of cervical spine lesions in rheumatoid arthritis. Spine 20:1128–1135
28. Pellici PM, Ranawat CS, Tsairis P, Bryan WJ (1981) A prospective study of the progression of rheumatoid arthritis of the cervical spine. J Bone Joint Surg Am 63:342–350
29. Rana NA, Hancock DO, Taylor AR, S HAG (1973) Atlanto-axial subluxation in rheumatoid arthritis. J Bone Joint Surg Br 55:458–470
30. Ranawat CS, O'Leary P, Pellici P, Tsairis P, Marchisella P, Dorr L (1979) Cervical spine fusion in rheumatoid arthritis. J Bone Joint Surg Am 61: 1003–1010
31. Redlund-Johnell I (1984) Posterior atlanto-axial dislocation in rheumatoid arthritis. Scand J Rheumatol 13:337–341
32. Santavirta S, Kankaapää U J S (1987) Evaluation of patients with rheumatoid cervical spine. Scand J Rheumatol 16: 9–16
33. Santavirta S, Kontinnen YT, Laasonen E, Honkanen V, Antti-Poika I, Kappui M (1991) Ten year results of operations for rheumatoid cervical spine disorders. J Bone Joint Surg Br 73:116–120
34. Scott JS, Huskisson EC (1967) Graphic representation of pain. Pain 2:175–184
35. Sumi M, Kataoka O, Ikeda M, Sawamura S, Uno K, Siba R (1997) Atlanto-axial dislocation. A follow-up study of surgical results. Spine 22:759–764
36. Zygmunt S, Saveland H, Brattström H, Ljunggren B, Larsson EM, Wollheim F (1988) Reduction of rheumatoid periodontoid pannus following posterior occipito-cervical fusion visualised by magnetic resonance imaging. Br J Neurosurg 2:315–320