Cervical spine

- Instability and/or dislocation between cervical vertebrae frequently occurs in patients with rheumatoid arthritis (RA).
- Severe pain and/or impending or existing clinical signs of spinal cord involvement are generally accepted as indications for surgery.
- In advanced rheumatoid arthritis there is an association between radiological findings and neurological symptoms. These patients should receive clinical and radiological examinations on a regular basis, particularly those at greatest risk for progressive malalignment, ie, patients with erosive small joint arthritis and on long-term steroid medication.
- Although nerve roots and the spinal cord are seldom affected, it is important to surgically stabilize the upper cervical spine and/or decompress the spinal cord in patients with stenosis before they show signs of neurological involvement.
- Rheumatic patients about to have hip or knee prosthetic surgery often show radiological signs of C1-C2 instability, and it is important to consider this in conjunction with anesthesia.
- Fusion of the upper cervical spine is a safe and effective method of decreasing pain and the risk of spinal cord injury, and the operation involves little risk for complications.

Chronic synovial inflammation may destroy joints, ligaments, and bone, mainly in the upper cervical spine. Atlantoaxial horizontal dislocation/instability directed forward and exceeding 3 mm due to destruction of ligaments may be observed after only a few years of disease. Malalignment may also be directed toward the side or, less frequently, backward. However, since the spinal cord canal is rather wide at this point, spinal cord involvement seldom occurs, and many patients with positive radiological findings do not have clinical symptoms.

Bone loss in the joints between the base of the skull and C1, and between C1 and C2, may result in vertical malalignment, and the tooth-like prominence (dens) will stick out into and through the exit for the spinal cord in the base of the skull (foramen magnum). Although concurrent erosion of the dens tip is common, more dramatic neurological symptoms involving weakness or, in some cases, total paralysis in both the upper and lower extremities (tetraparesis/plegia) are found in this patient group. Instability and malalignment at lower (subaxial) levels of the cervical spine are somewhat less common, but may also cause considerable neurological symptoms due to the more narrow conditions at these levels. Spontaneous stiffening sometimes occurs at these levels and may avert the threat to the spinal cord. Although some patients have isolated subaxial malalignment or instability, combinations of atlantoaxial horizontal and vertical malalignment/instability and subaxial changes are more common.

The earliest and most common symptom of inflammation in the joints between the uppermost cervical vertebrae is pain in the neck and back of the head, in some cases radiating toward the eyes. This can be explained by the granulation tissue which is secondary to the instability, and which apart from pressure on the spinal cord may also result in compression on the C2 nerve roots. A snapping sensation when turning the head, and a sense of fatigue in the neck, are other common, but non-specific, symptoms. In advanced cases with spinal cord involvement, neurological symptoms may appear, eg, a tingling sensation in the arms and legs, peripheral numbness, stiffness (spasticity), followed by muscular weakness, gait difficulties, and bladder involvement. It may be difficult to discover early signs of spinal cord involvement in patients having a disease which itself causes weakness and stiffness in muscles and

joints. Spontaneously or surgically fused joints may render neurological assessment difficult. Furthermore, polyneuropathy is often present and may partly explain the sensory involvement.

The space in the spinal cord canal is best mapped by magnetic resonance imaging (MRI) [26]. This method has substantially improved the ability to establish the indications for surgery and is also useful in showing the involution of inflammatory tissue after stabilizing procedures [40]. MRI can also be used to measure the shape and diameter of the spinal cord. Preoperative MRI investigation of the rheumatic cervical spine should be mandatory.

Although the spinal cord canal is relatively wide, instability or malalignment in combination with inflamed soft tissue may restrict space and affect the spinal cord to the extent that surgical treatment is required.

Scope of the problem

Changes in the joints between the occiput and C1, and between C1 and C2, have been found to occur in 10% to 70% of patients with rheumatoid arthritis, depending on which patient database is used [3,4,28,38]. The most reliable information is found in a Finnish study that estimates the prevalence of rheumatic cervical spine changes in a population of 13,000 with 103 detected cases of RA [15]. Anterior horizontal atlantoaxial instability/dislocation occurred in 33%, lateral in 14%, and posterior in 2%. Vertical dislocation was found in 14% to 27% depending on the method of measurement, and subaxial instability/dislocation was found in 21%. Similar distribution of instability types was reported in a Swedish study [27]. Despite the frequency of instability and malalignment, only a small number of patients actually develop symptoms with spinal cord involvement.

It is important to remember that rheumatic patients who are to undergo hip or knee prosthetic surgery often show radiological signs of C1-C2 instability, which must be considered in conjunction with anesthesia [7].

The natural history of rheumatic changes in the cervical spine is relatively unknown since prospective studies are lacking. What is known, however, is that although most patients show a radiological progression, the neurological symptoms have a less pronounced progression [24]. Patients with erosive small joint arthritis and long-term steroid medication, run the greatest risk for progressive malalignment [36]. Most likely, the spinal cord can tolerate many years of constriction before its function is affected. Hence, radiological progression is not an immediate indication for surgery. In advanced rheumatoid arthritis, there is an association between radiological findings and neurological symptoms, but otherwise the association between radiological findings and clinical signs of spinal cord involvement is weak, and therefore the indication for surgery may be prophylactic only in exceptional cases (in vertical dislocation) [1].

During 1993, around 90 cervical spine operations were performed in rheumatic patients in Sweden, 10 of which were reoperations. Half were C1-C2 fusions and the remainder were fusions involving the base of the skull or procedures performed at the subaxial level. In light of their prevalence, cervical fusions should probably be performed earlier in the course of disease and more often than is currently the case.

Indications and surgical methods

Analgesics and a cervical collar provide initial relief of symptoms in most patients. When involvement of the upper cervical nerve roots is suspected, a peripheral nerve block or root block may be attempted with the help of fluoroscopy. However, if the patient has disabling pain and/or symptoms of spinal cord compression, or if MRI reveals stenosis in an asymptomatic patient, operative decompression and stabilization must be considered. Major instability is always a more serious condition than a major, but stable, dislocation.

Traditionally, the indications for surgery have been based on the distance (measured by radiology) between the first cervical vertebra and dens on the second cervical vertebra, and on the degree of vertical dislocation. However, the most interesting measurement is the space available for the spinal cord. An American study shows that the risk for developing irreversible neurological damage can be minimized if operative stabilization, regardless of the presence of neurological symptoms, is performed in all patients with C1-C2 or subaxial malalignment causing spinal cord space to be less

than 14 mm, or with C1-C2 instability and concurrent vertical dislocation exceeding 5 mm [3]. Patients with clinically obvious spinal cord involvement have a very poor prognosis for survival [20,21].

In addition to threatening spinal cord injury, another indication for surgery is pain which does not respond to conservative treatment. Depending on the extent of bone damage, fusion is performed either between the base of the skull and the upper cervical vertebrae, or between the first and second cervical vertebra only. Numerous methods are described [2,31]. The dominant procedures in Sweden are C1-C2 fusion with wire and bone transplant, and occipitocervical fusion with wire, bone cement, and bone transplant [40]. Only in rare cases is arthrodesis performed from the front, due to the usually poor quality of bone. Usually, bone from the patient's iliac crest is used to promote bone healing. Although the bone does not always heal completely, most patients are helped, probably because connective tissue healing provides sufficient stability [6,22,28].

If C1-C2 fusion is performed early, it may prevent further progression of vertical dislocation [2,16]. However, vertical dislocation in patients with well preserved dens calls for occipitocervical fusion.

Fusing the cervical spine at a high level does not appear to increase the risk for instability at the adjacent lower levels [17,27]. Progressive, so-called *staircase*, deformity is common in both operated and nonoperated patients [6]. One study reports that 36% of patients in whom fusion included the base of the skull later developed subaxial instability requiring surgery, compared to 6% of those in whom fusion involved C1 and C2 alone [16].

Instability and dislocation at subaxial levels of the cervical spine seldom result in spinal cord symptoms. In these cases, the possibility exists to surgically relieve pressure by removing one or more vertebral arches (laminectomy). However, the possibility for fusion is poor since bone quality is usually poor.

Rehabilitation

Postoperatively, patients are treated with various stable external fixation methods, depending on the stability achieved by surgery. When bone cement is used, a soft cervical collar is sufficient while the wound is healing. If only wire and transplanted bone are used, a fixed cervical collar is usually recommended for 8 to 10 weeks postoperatively. Elsewhere in the world, rigid external fixation, eg, a halo vest, is more common during the healing period. However, this type of treatment is not tolerated well by patients with multiple joint disease. Hence, in these patients it is common to use surgical methods which offer immediate and secure fixation, possibly with bone cement around the fusion.

Surgical treatment results

Horizontal instability of C1-C2 and vertical dislocation of dens

Most studies report results from a mix of C1-C2 and occipitocervical fusion procedures. Radiological and clinical outcomes from series published later than 1980 are shown in Table 1 [3,5,6,8,10, 11,13,19,21,22,29,33,34,35,37,39]. Of 250 patients who received C1-C2 fusion surgery, 75% (range: 50% to 100%) showed bone healing, 82% (range: 53% to 100%) improved clinically (pain relief and/or decreased neurological symptoms), and peroperative and postoperative mortality was 3% (range: 0% to 11%). Of the 319 patients who received occipitocervical fusion surgery, 84% (range: 62% to 100%) showed bone healing, while 73% (range: 50% to 100%) improved clinically, and early mortality was 6% (range: 0% to 25%). These studies included small numbers of patients, except for the Swedish study which thoroughly analyzed postoperative complications in 163 occipitocervical fusion cases [38]. On average, the patients were 61 years of age at the time of surgery, and the average followup period was 4.5 years. Clinical improvement was observed in 88% of the cases. In 7% of the cases, problems remained unchanged, and in 5% the clinical symptoms progressed despite stabilizing surgery. No patients died in conjunction with the operation or during the first 4 months postoperatively. Seventy-nine patients suffered one or more complications (eg, a break in the wire, fracture, infection). Most complications were not dangerous, and only

Table 1. Radiological and clinical results after stabilizing surgery in the upper cervical spine.

Author	Number of patients	Mean-age at time of operation	Follow-up (years)	Bone-healing (%)	Fibrous healing (%)	Failure-to heal (%)	Clinical improve-ment (%)	Mortality (< 3 months) (%)
C1-C2 fixation								
Conaty&Mongan (1981)	17	50	5	71	18	11	71	0
Thompson&Meyer (1984)	10	53	3	90	0	10	84	10
Fehring&Brooks (1985)	12	–	1–8	67	0	33	–	0
Larsson&Toolanen (1986)	23	56	>1	52	32	16	96	0
Zoma et al. (1987)	17	60	5	59	0	41	53	6
Clark et al. (1989)	20	57	>2	75	10	15	65	0
Milbrink&Wigren(1989)	19	57	2	67	11	12	89	5
Papadopoulos et.al. (1991)	17	60	1	76	12	12	–	6
Santavirta et al. (1991)	24	57	10	50	17	33	91	0
Chan et al. (1992)	11	55	5	91	9	0	100	0
Boden et al. (1993)	9	57	>2	100	0	0	78	11
Peppelman et al. (1993)	55	60	5	85	0	15	95	0
Stirrat&Fyfe (1993)	16	61	3	87	13	0	75	0
Occipitocervical fusion								
Conaty&Mongan (1981)	10	54	3	80	0	10	60	10
Fehring&Brooks (1985)	5	–	1-8	75	0	25	–	0
Larsson&Toolanen (1986)	4	60	>1	100	0	0	75	0
Zoma et al. (1987)	13	60	5	62	0	38	77	6
Heywood et al. (1988)	8	–	–	88	0	12	50	13
Clark et al. (1989)	16	56	> 2	100	–	–	69	0
Grob et al. (1990)	26	56	4	73	15	12	62	4
Santavirta et al. (1991)	4	47	>10	100	–	–	100	0
Vanden Berghe et al. (1991)	22	56	4	69	31	0	88	9
Chan et al. (1992)	8	54	5	100	–	–	63	25
Boden et al. (1993)	18	60	> 2	82	12	6	67	6
Peppelman et al. (1993)	22	60	6	81	0	9	76	9
Zygmunt et al. (1995)	163	61	6	–	–	–	88	0

- = data lacking

24 (15%) reoperations had to be performed. Deep infection was reported in five patients and superficial wound infection in 11 patients. Among the patients who suffered complications and in some cases reoperation, 84% had improved from the procedure, 9% remained unchanged, and 7% were reported to be worse than before the first operation. Hence, it appears as if occipitocervical fusion, despite a rather high rate of complications, yields good clinical results in stabilizing the upper cervical vertebra.

The wire technique is not always successful in repositioning C1 and C2, and in the light of the relatively high frequency of broken wires, other alternatives have emerged which may further improve stability and bone healing [12,13].

Only rarely is dens removed via the oral cavity (transoral decompression) in combination with posterior C1-C2 fusion. The method is advocated by some, but is associated with several risks. An English study recommends the procedure in patients with fixed C1-C2 malalignment, large dens granuloma, and vertical malalignment [9]. Of 68 patients, four died in the early postoperative stage, 3 had deteriorated clinically, and of the remaining 61, one third were greatly improved while the others improved only slightly. These findings are not as strong as those presented above or in Table 1.

Many providers in Sweden use occipitocervical fusion by wire and bone cement to treat both C1-C2 instability alone and in combination with vertical dislocation. Occipitocervical fusion probably prevents the progression of vertical malalignment [31]. It is also possible that C1-C2 fixation is better guaranteed if the base of the skull is included in the fusion procedure.

Most reports in the literature are retrospective and include data on radiological healing and pain

relief, while data on functional outcomes are neglected. One exception, however, is a Swedish study of 20 patients who received occipitocervical fusion [14]. Although most reported decreased pain and improved neurological symptoms, the gains in functional capacity were insignificant.

Subaxial dislocations

Subaxial dislocation usually occurs at several levels (C3-C4 is the most common), at times in combination with C1-C2 instability. It may be difficult to ascertain the level from which clinical symptoms originate, although MRI may be of help. Laminectomy and removing granulation tissue may effectively relieve pressure. Attempts are made to combine laminectomy at several levels with fusion. Posterior fusion at subaxial levels is technically difficult to achieve, although numerous methods being described [31]. Anterior fusion is usually impossible due to poor bone quality.

Although a Japanese study reports clinical improvement in 18 of 26 patients who received laminectomy and posterior or anterior fusion, and a Finnish study of 16 patients reports one early death, good pain relief in all other patients, and neurological improvement in most, it appears that the clinical results of laminectomy and fusion at multiple subaxial levels are inferior to the atlantoaxial results [18,30,37].

At least in the short term, laminectomy without concurrent posterior fusion appears to be a potential method in patients having subaxial dislocation without instability.

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