Rheumatoid arthritis affects 0.5–1% of the Caucasian population; however, the incidence around the world varies considerably with some North American Indians having an incidence of 5–7% and the disease being almost unheard of in South Africa and Nigeria [1]. It has not always been seen in Europe; there is little evidence of its prevalence before Renoir’s self portrait and Garrod’s descriptions about 120 yrs ago [2, 3].

The polyarthropathy of rheumatoid arthritis affects the joints of the spine, and particularly the upper cervical spine. A rare but devastating complication is the development of progressive neurological disability leading ultimately to paralysis and a lingering death. In the early days, this complication was under-diagnosed and disability was blamed on peripheral neuropathy due to drugs and nerve compression by diseased peripheral joint capsules. Sudden deaths were attributed to strokes or heart attacks until Redlund-Johnell [4] reviewed the coroners post mortems in Southern Sweden and found that over half of the ‘sudden deaths’ in rheumatoid arthritis were actually associated with instability of the cervical spine and cord compression. The increasing availability of CT, and then MRI scans, revealed that the brain stem and upper cervical cord compression of rheumatoid arthritis was more common than previously realized. Henderson et al. [5] clearly demonstrated that it was the mechanical problem of spinal cord compression and not vasculitis that caused disability in these patients, and through the databases collected by Corbett et al. [6] and Kauppi et al. [7], the natural history of this complication was determined. Cervical myelopathy rarely appeared in the first decade from the onset of disease because the aetiology was probably repeated minor cord injuries over time [8]. Two or three previous large-joint operations usually preceded the development of myelopathy [9]. Once established, however, neurological deterioration could be rapid with half of those who developed myelopathy dying within a year and most patients dying within 7 yrs [9]. In the ‘80s, it became apparent that patients and clinicians had to be made more aware of the problems of rheumatoid cervical instability. More emphasis was now placed on the importance of treatment before irreversible neurological damage had occurred [10].

The timing and extent of surgery, however, were issues which remained for debate. It was clear that surgery for the bed-bound quadriparetic patient was not only dangerous, with a mortality of up to 30%, but also did not improve the disability often; it was ‘too much and too late’ [10], whilst others with atlantoaxial subluxation might remain asymptomatic for many years without evidence of neurological deterioration. Therefore, if a proposed operation had a significant mortality, then the risks of surgery outweighed the dangers of atlantoaxial subluxation. The true danger of atlantoaxial subluxation could not be assessed with flexion and extension X-rays alone; once the movement exceeded 3 mm, it did not really matter how much subluxation there was. Only with the advent of MRI scans could the true extent of subluxation and cord compression be appreciated, allowing rational decisions to be made regarding surgery. MR imaging, for example, reveals compression by rheumatoid pannus and vertical translocation, which can occur with only small increases in anterior atlantoaxial intervals on cervical X-rays.

Casey and Crockard [11] calculated that there might be more than 62 000 people in the United Kingdom at risk of neurological compression from rheumatoid cervical instability, and extrapolating to the United States, a figure of a quarter of a million [11]. This was calculated from records going back to the ‘60s and ‘70s when the patients first developed their disease. In a more recent study at the turn of this century, Hamilton et al. [12] showed that the incidence is actually 14 times lower. There has been a step-like change in the disease prevalence.

During the same time period, the numbers of large-joint replacements have also dramatically fallen. Ward [13] in California and de Silva et al. [14] in Minnesota revealed a clear decrease in the numbers of knee and hip arthroplasties performed in patients with rheumatoid arthritis. These changes have coincided with the introduction of new disease modifying anti-rheumatic drugs (DMARDs) and the decreased use of steroids in rheumatoid arthritis. Many of the bony and ligamentous changes seen in the rheumatoid spine resemble those of long-term steroid usage. Indeed, it might be argued that some of the problems associated with rheumatoid changes in the neck might be attributed to the long-term use of steroids rather than the disease itself. Over the last 20 yrs, five factors have emerged, changing the way we treat cervical rheumatoid disease:

(i) Changes in medical management—we now use DMARDs, anti-tumour necrosis factor and anti-interleukin1 agents, and have moved away from using steroids for chronic disease.

(ii) The expression of the disease itself may have become less aggressive.

(iii) A change in patients’ expectations—patients’ perception of treatment as being for a potentially lethal complication of an incurable disease has evolved to patients’ demands for treatment to maintain their posture, improve deformity and minimize or abolish neck pain.

(iv) Medical teams have realized that surgery should be performed to prevent deformity and neurological symptoms and to treat specific neck pain.

(v) Surgical decision-making has moved from complex heroic operations on patients who are too ill to survive, to elective preventative operations in patients who are relatively well [15, 16].

The surgical management of cervical rheumatoid disease has evolved considerably from the days of long posterior fusions in the severely debilitated and occasional surgery through the mouth to decompress the medulla [17, 18] towards early surgery to prevent disease progression [19]. The results of surgery in specialized centres has improved over time, with 66–89% of symptomatic patients benefiting from surgery and a decrease in the rate of surgical complications [12, 20].

The realization that in many cases the occipitoatlantal joint is not involved in the early stages has led to the concept of ‘segment-saving’ surgery; for example, fusing the atlantoaxial joint whilst preserving the occipitoatlantal joint.

It is important that the surgery is carried out by teams that are experienced in modern surgical techniques and in treating...
rheumatoid patients. There is no place for the occasional operator. A survey in Scandinavia clearly demonstrated the high incidence of complications and requirement for surgical revision after surgery was performed in smaller units [21]. Patients with rheumatoid arthritis should be treated by a multi-disciplinary team that has access to specialist cervical spine surgeons who are well-versed in the possibilities, problems and complications of surgery in rheumatoid patients.

With these caveats, cervical surgery in rheumatoid arthritis should now be offered in the same way that large-joint replacement surgery is. No longer should it be a last-ditch heroic attempt at treating irreversible paralysis but, in expert hands, instability, deformity and some forms of neck pain may be treated by planned elective surgery, with low mortality and morbidity, to improve a patient’s quality of life. Spine surgery for rheumatoid disease has come a long way in the past 20 yrs.

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