DISLOCATION OF THE CERVICAL SPINE
WITH CORD COMPRESSION IN RHEUMATOID ARTHRITIS

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In rheumatoid arthritis chronic proliferative inflammatory changes may involve all of the synovial tissue in the cervical spine (Ball 1963, Ball and Sharp 1971). Radiographs of the upper cervical area may reveal erosions of the dens, narrowing of the atlanto-axial and atlanto-occipital joints with erosion of the adjacent bone, and abnormal mobility of the atlas on flexion. The distance between the anterior margin of the dens and the anterior arch of the atlas may exceed the reported normal values, which are said to be 2.5 to 3 millimetres (Brocher 1966); 4 millimetres for persons under forty-five and 3 millimetres over that age (Sharp and Purser 1961); and between 2 and 2.2 millimetres (Hinck and Hopkins 1960).

Below C2 the disc spaces may be narrowed and bone plate erosion without osteophytosis is seen, resulting in abnormal mobility and dislocation at multiple levels (Sharp, Purser and Lawrence 1958). All these abnormalities can result in damage to the spinal cord, usually at the C1–2 level. This occurs in only a small proportion of cases; exact figures are lacking.

Many cases of spinal cord compression have been reported (Coste, Auquier and Civatte 1952; Vignon and Patet 1955; de Séze, Djian and Caroit 1957; Coste, Merle d’Aubigné and Garcin 1960; Graham 1960; Martel 1963; de Leeuw and Meijers 1967), some with a fatal outcome due to compression of the medulla oblongata (Davis and Markley 1951; Hauge 1958; Martel and Abell 1963; Webb, Hickman and Brew 1968; Whaley and Dick 1968). In one case obstruction of the vertebral canal was demonstrated (Webb et al. 1968).

A few larger series with C1–2 slips have been published. Serre, Simon, Janicot and Levy (1963) noted such a slip in twenty-two out of fifty-nine patients in hospital with rheumatoid arthritis. Neck pain was not a prominent feature. Signs of a partial cord lesion were present in two cases; in one of these cases a collar was worn for eight months; in the other no specific treatment was given.

Sharp and Purser (1961), however, reported neck pain in eight out of twenty-six patients with rheumatoid arthritis; all showed displacement at C1–2. Definite neurological signs were present in six of the eight. Four of those with cord compression underwent operation. Of the two patients who were not treated surgically, one recovered while wearing a collar, and in the other the signs regressed with skull traction but relapsed during immobilisation in a Minerva plaster. The remaining patients either received no specific treatment or were provided with a collar.

Crelin, Maccabe and Hamilton (1970) described nineteen rheumatoid patients with severe slips either in the C1–2 area or at lower levels, and with neck pain a prominent feature. Eleven were treated surgically; nine had symptoms and signs of spinal cord compression and two had symptoms of vertebral artery insufficiency. Traction was thought to be harmful and was abandoned before operation. According to Bland (1967), traction has a deleterious effect on vertebral slipping in rheumatoid arthritis. In his opinion the atlanto-axial area can be stabilised by the constant use of a firm plastic collar for several months.

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CLINICAL MATERIAL

Between February 1961 and December 1969 fourteen patients with rheumatoid arthritis were treated for cervical cord lesions due to severe destructive changes with dislocation in the cervical spine. Thirteen were women. Their ages ranged between forty-five and seventy years, with an average of fifty-nine. The Rose-Waaler and latex fixation tests were positive in all cases. Ten patients were confined to a wheelchair; four could walk but were very much restricted in their daily living. The relevant details of the whole series are given in Table I. Neurological features—It is evident that patients with a crippling form of rheumatoid arthritis complain of pain at various sites and also of muscle weakness. This may mask the symptoms of a cord lesion for some time, even though the pattern of complaints has changed in a quite characteristic way. In analysing the case histories we were struck by the frequency of complaints which appeared to indicate spinal cord damage. It is important to stress these “alarm signals”: 1) severe neck pain, often radiating to the occiput (nine cases); 2) disturbed bladder function, varying from incontinence to urinary retention (ten cases); 3) diminished motor power in arms and legs (ten cases); 4) jumping legs, as a consequence of spinal automatism (six cases); 5) tingling of the fingers and feet (six cases) or only numbness (another six cases); 6) a “marble sensation” in the limbs and trunk. This peculiar complaint was volunteered in two cases with a disturbance of deep sensibility. None of our patients had noticed these symptoms previously.

Neurological examination of crippled rheumatoid patients can be very difficult. The inflamed or deformed joints often make it impossible to judge motor power and to elicit reflexes. A thorough examination of sensibility is possible, however. A disturbance of sensibility in combination with the alarm signals will establish the diagnosis of a cervical cord lesion. In these crippled patients lumbar puncture and contrast myelography were felt to be too severe a burden.

In our fourteen patients we found an incomplete transverse lesion in one case, partial unilateral or bilateral paresis in eleven cases, partial loss of all modes of sensibility in nine cases, and dissociated loss of protopathic or epticritic sensibility in three cases.

ILLUSTRATIVE CASE REPORTS

Case 2—A woman aged sixty-five had suffered from severe destructive rheumatoid arthritis for six years when she started to complain of headache, pain in the neck, heaviness of the arms and right leg and impaired continence of urine. On examination, disturbances of sensibility were found in the cervical dermatomes, vibration sense was impaired in the legs, and there was a severe spastic paresis of both arms and right leg. A slip at C1–2 was found, the anterior part of the atlas lying in front of the vertebral body of the axis close to the C2–3 disc. The dens was severely eroded, the remnants being situated centrally in the foramen magnum and protruding 4 millimetres into the cranial cavity, with pseudobasilar impression (Fig. 4). The development of these lesions in radiographs taken four years and one year before admission is shown in Figures 1 to 3.

Skull traction was applied. This had only a slight effect on the subluxation but the neurological state improved: normal micturition was restored, and power in the right leg and sensibility were improved.

At operation ten weeks later the posterior arch of the atlas was found to be completely detached. Laminectomy was carried out at C1, the foramen magnum was enlarged, and fusion from occiput to C3 was performed. After operation transient tetraplegia and some mental disturbance persisted for a fortnight. The patient was discharged after six months with only slight paresis of both arms still present. Bony fusion had occurred.

Five years later no neurological lesion could be found. Despite her many handicaps the patient was happy with the result of the operation. She died of gastric haemorrhage six years after the operation. At autopsy the bones in the surgical field were found to be completely fused; C4, 5 and 6 were also ankylosed (Fig. 5). Granulation tissue was found between the vertebral bodies of C3 and 4 and in the corresponding posterior intervertebral joints. On mid-sagittal section the cord appeared to be somewhat compressed at the C3–4 level. At this level the diameter of the cord was 6.5 millimetres and just above and below this segment 8.5 millimetres; measurements of the transverse diameter were not possible.
**TABLE 1**

**DETAILS OF THE SERIES OF FOURTEEN CASES**

<table>
<thead>
<tr>
<th>Case number</th>
<th>Age (years)</th>
<th>Sex</th>
<th>Duration of R.A. (years)</th>
<th>Neurological syndrome</th>
<th>Level and degree of subluxation</th>
<th>Traction: duration and effect</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>60</td>
<td>M</td>
<td>7</td>
<td>Incomplete transverse lesion of cord</td>
<td>C.1-2: in flexion 15 mm, in extension 10 mm; C.2-3: minor slip</td>
<td>6 weeks; no neurological improvement. Slip still 10 mm in extension</td>
</tr>
<tr>
<td>2</td>
<td>65</td>
<td>F</td>
<td>6</td>
<td>Spastic and epicritic sensory disturbance; micturition impaired</td>
<td>C.1-2: C.1 tilted forward, anterior arch in front of C.2 and C.3; dens eroded away; pseudobasilar impression; posterior arch of C.1 loose at operation</td>
<td>10 weeks; good neurological improvement. Slip unchanged</td>
</tr>
<tr>
<td>3</td>
<td>67</td>
<td>F</td>
<td>20</td>
<td>Spastic and mixed sensory disturbance; micturition impaired</td>
<td>C.1-2: C.1 tilted forward, anterior arch in front of C.2 and C.3; absence of dens; pseudobasilar impression</td>
<td>4 weeks; neurological improvement. Slip unchanged</td>
</tr>
<tr>
<td>4</td>
<td>67</td>
<td>F</td>
<td>23</td>
<td>Occipital neuralgia: epicritic sensory disturbance; micturition impaired</td>
<td>C.1-2: in flexion 15 mm, in extension 12 mm</td>
<td>4 weeks; neurological improvement. Slip reduced to 8 mm</td>
</tr>
<tr>
<td>5</td>
<td>56</td>
<td>F</td>
<td>7</td>
<td>Spastic disturbance; micturition impaired</td>
<td>C.1-2: 8 mm</td>
<td>2 weeks; neurological improvement. Slip reduced to 5 mm</td>
</tr>
<tr>
<td>6</td>
<td>57</td>
<td>F</td>
<td>13</td>
<td>Spastic and epicritic sensory disturbance</td>
<td>C.1-2: in flexion 17 mm, in extension 5 mm</td>
<td>3 days; neurological improvement lying supine; slip reduced to 5 mm</td>
</tr>
<tr>
<td>7</td>
<td>65</td>
<td>F</td>
<td>16</td>
<td>Spastic and mixed sensory disturbance; micturition impaired</td>
<td>C.1-2-3: severe destruction; kyphosis and shortening of cervical spine</td>
<td>Daily intermittent traction for one year; slight improvement, sudden deterioration</td>
</tr>
<tr>
<td>8</td>
<td>45</td>
<td>F</td>
<td>9</td>
<td>Occipital neuralgia: mixed sensory disturbance</td>
<td>C.1-2: in flexion 11 mm, in extension 5 mm</td>
<td>1 week; complete neurological recovery; slip reduced to 5 mm</td>
</tr>
<tr>
<td>9</td>
<td>58</td>
<td>F</td>
<td>24</td>
<td>Occipital neuralgia: mixed sensory disturbance</td>
<td>C.1-2: in flexion 13 mm, in extension 3 mm</td>
<td>2 days; improvement of slip, lying supine, to 5 mm</td>
</tr>
<tr>
<td>10</td>
<td>53</td>
<td>F</td>
<td>11</td>
<td>Occipital neuralgia: mixed sensory disturbance; paresis of the arms only; micturition impaired</td>
<td>C.1-2: C.1 tilted forward, anterior arch in front of C.2 and C.3; destruction of atlanto-axial joints and dens; pseudobasilar impression</td>
<td>4 weeks; complete neurological recovery; forward tilt unaltered</td>
</tr>
<tr>
<td>11</td>
<td>61</td>
<td>F</td>
<td>9</td>
<td>Mixed sensory disturbance; paresis of the arms only; micturition impaired</td>
<td>C.1-2: 7 mm; erosion of dens; destruction of both atlanto-axial joints</td>
<td>2 weeks; neurological improvement; slip unchanged</td>
</tr>
<tr>
<td>12</td>
<td>48</td>
<td>F</td>
<td>20</td>
<td>Spastic disturbance; micturition impaired</td>
<td>C.6-7: 6 mm in flexion and in extension; destruction of intervertebral joints</td>
<td>Several weeks; neurological improvement; complete reduction of slip</td>
</tr>
<tr>
<td>13</td>
<td>62</td>
<td>F</td>
<td>22</td>
<td>Spastic and sensory disturbance; micturition impaired</td>
<td>C.4-5: 6 mm in flexion and in extension</td>
<td>1 week; no effect</td>
</tr>
<tr>
<td>14</td>
<td>70</td>
<td>F</td>
<td>21</td>
<td>Spastic and sensory disturbance; micturition impaired</td>
<td>C.1-2: 7 mm; C.4-5: minor slip. Block C.2-3-4; shortening of cervical spine</td>
<td>4 weeks; no effect</td>
</tr>
</tbody>
</table>

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<table>
<thead>
<tr>
<th>Surgical treatment</th>
<th>Neurological result</th>
<th>Orthopaedic result</th>
<th>Follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>Laminctomy C.1; spondylodesis from occiput to C.2 and 3</td>
<td>Sensory recovery; slight spastic paresis right leg</td>
<td>Pseudarthrosis</td>
<td>Recurrence of neurological symptoms after 11 years; sudden death at home after 2 years</td>
</tr>
<tr>
<td>Laminctomy C.1; foramen magnum enlarged; spondylodesis from occiput to C.2 and 3</td>
<td>Post-operative quadriplegia which improved to slight paresis of arms only; sensory recovery</td>
<td>Bone fusion occiput to C.2 and C.3; position of C.1 as before operation</td>
<td>Complete recovery; at 5 years no neurological abnormalities; died at 6 years of gastric bleeding</td>
</tr>
<tr>
<td>Laminctomy C.1; foramen magnum enlarged; spondylodesis from occiput to C.2, 3 and 4</td>
<td>Slight sensory disturbance in fingers, motor function recovered</td>
<td>Bone fusion occiput to C.4; position of C.1 as before operation. Later, slip of C.7 on D.1</td>
<td>Alive; 4 years after operation, dysaesthesia in both hands, minimal sensory loss in the right arm C.7–Th.1 with cord symptoms</td>
</tr>
<tr>
<td>Laminctomy C.1; spondylodesis from occiput to C.2, 3 and 4</td>
<td>Slight sensory disturbance in fingers</td>
<td>Bone fusion doubtful</td>
<td>Died 2 years later of perforated gastric ulcer and cardiac failure</td>
</tr>
<tr>
<td>Spondylodesis C.1–2</td>
<td>Complete recovery</td>
<td>Bony fusion. Subluxation C.2–3 and narrowed discs below C.2 after 7 years</td>
<td>Died at home 8 years after operation probably from pneumonia</td>
</tr>
<tr>
<td>Spondylodesis from occiput to C.2, 3 and 4</td>
<td>Early recovery during the first 2 weeks</td>
<td>—</td>
<td>Died of pulmonary embolism 15 days after operation</td>
</tr>
<tr>
<td>Laminctomy C.1, 2 and 3; spondylodesis from occiput to C.4 and 5</td>
<td>Sensory disturbance remained; motor power improved greatly</td>
<td>Bony fusion</td>
<td>Died of heart failure 1 year after operation</td>
</tr>
<tr>
<td>Spondylodesis from occiput to C.1, 2 and 3</td>
<td>Complete recovery</td>
<td>Bony fusion; slip C.1–2, 7 mm</td>
<td>Alive and walking; after 4 years no neurological abnormalities</td>
</tr>
<tr>
<td>Spondylodesis from occiput to C.1, 2 and 3</td>
<td>Complete recovery</td>
<td>Bony fusion; slip C.1–2, 3 mm</td>
<td>Alive and walking; 3 years after operation no neurological abnormalities</td>
</tr>
<tr>
<td>Laminctomy C.1; foramen magnum enlarged; spondylodesis from occiput to C.2 and 3</td>
<td>Complete recovery</td>
<td>Bony fusion; position of C.1 as before operation</td>
<td>Alive 3 years after operation; no neurological abnormalities</td>
</tr>
<tr>
<td>Spondylodesis from occiput to C.1, 2 and 3</td>
<td>Complete recovery</td>
<td>Bony fusion; slip C.1–2 reduced to 2 mm</td>
<td>Died of septicaemia 3 years after operation</td>
</tr>
<tr>
<td>Not performed</td>
<td>Marked improvement while in hospital</td>
<td>—</td>
<td>Died at home 6 months after discharge, probably due to a recurrence of cord compression</td>
</tr>
<tr>
<td>Laminctomy C.4 and 5; spondylodesis C.2–7</td>
<td>After improvement, slow recurrence of transverse lesion</td>
<td>Bony fusion</td>
<td>Died of cardiac failure 1 year after operation</td>
</tr>
<tr>
<td>Not performed</td>
<td>Progressive transverse lesion</td>
<td>—</td>
<td>Died 6 months after discharge due to complications of a complete cord lesion</td>
</tr>
</tbody>
</table>
Case 2. Figure 1—A lateral radiograph of the cervical spine taken in a neutral position in 1960. There is no sign of a slip of C.1 on C.2 at this time. Figure 2—A lateral radiograph taken in flexion in 1963. Forward tilting of C.1 on C.2 is now well shown, together with narrowing of the C.2-3 and C.3-4 discs.

Case 2. Figure 3—The lateral radiograph in extension taken in 1963. The forward slip of C.1 on C.2 is less marked than in flexion. Note the slight backward slipping of C.2 on C.3, of C.3 on C.4, and of C.5 on C.6. Figure 4—A lateral tomograph taken before operation in 1964. The forward tilt of C.1 on C.2 is well shown.
Case 6—This woman aged fifty-seven had suffered for thirteen years from severe polyarticular rheumatoid arthritis. For two years she had been unable to walk because of a peroneal neuropathy on both sides. A few months before admission she had complained of odd sensations in both feet, together with tingling and numbness of the hands. The strength of her arms had diminished. She had no complaints concerning her neck.

Fig. 5
Case 2—The radiograph of a sagittal slab obtained at necropsy in 1970, six years after the operation in 1964 when laminectomy of C.1, enlargement of the foramen magnum, and grafting of the occiput to C.2 and C.3 were performed. The anterior arch of C.1 can be distinguished in front of the body of C.2, quite close to the C.2-3 disc, which is greatly narrowed. A small remnant of the posterior arch of C.1 can be seen. The arch of C.3 is atrophied and there is a translucent zone between it and the arch of C.4. The bodies of C.3 and 4 appear to be crushed together. The arches and the bodies of C.4, 5 and 6 appear to have fused.

On neurological examination a spastic paresis of the right leg with a positive Babinski sign and impaired vibration sense in both legs were found. Radiographs showed a mobile slip of C.1 on C.2 of 17 millimetres in flexion and 5 millimetres in extension (Figs. 6 and 7). Lying supine as much as possible during the period before operation gave relief of the symptoms in the hands and feet, especially on the right side. She gained strength in the right arm and could lift heavier objects than before. Neurological examination now revealed normal vibration sense in both legs and the Babinski signs negative.

A few days before operation skull traction was applied. Laminectomy of C.1 was not considered necessary. Cervico-occipital fusion down to C.4 was performed.
This patient died suddenly fifteen days after the operation, and at autopsy massive pulmonary embolism was found. The cervical spine was removed. The dens showed some bone proliferation and was surrounded by granulation tissue. The size of the spinal cord was markedly reduced at the level of the dens (Fig. 8).

RADIOLOGY

The radiographs of the cervical spine were taken in two directions: lateral films in flexion and extension, usually with tomography, and antero-posterior films. The latter were often difficult to read, and even the tomograms.

The abnormalities found can be classified as follows: 1) dislocation at the C.1–2 level in nine cases (Figs. 2, 3, 4, 6 and 7); 2) dislocation below C.2 in two cases; and 3) dislocation at the C.1–2 level as well as below C.2 in three cases.

The C.1–2 level is involved the most frequently, but severe destruction often makes it difficult to discern the various
structures. In general, two types of dislocation can be distinguished: 1) A forward slip of the atlas and occiput in flexion; in extension the slip is diminished. In this series the distance in flexion between the anterior border of the dens and the anterior arch of the atlas varied between 7 and 17 millimetres. The tip of the dens is located centrally in the foramen magnum, usually just below that level, thus causing a marked narrowing of the spinal canal. 2) A forward tilting of the atlas, its anterior part being displaced downward to a position in front of the axis close to the C.2-3 disc. The dens is often completely eroded. The body of C.2, sometimes with remnants of the dens, is located centrally in the foramen magnum and protrudes into the cranial cavity (pseudobasilar impression). The spinal canal is not only narrowed in an antero-posterior direction, but the distance between the C.2-3 disc and the foramen magnum is reduced (see Case 2, Figures 2 to 4).

Dislocations in the area caudal from C.2 occurred in only two cases, in one at the C.4-5 level and in the other at C.6-7. In both cases destruction of the disc, the adjacent vertebral plates and the intervertebral joints was visible as well as a forward slip of 5 to 6 millimetres. Severe narrowing of the spinal canal had occurred.

**TREATMENT**

It seems desirable to avoid a burdensome operation in these patients, many of whom are severely crippled. To provide them instead with a collar as an external support might seem preferable (Bland 1967). But such a collar must be applied firmly to the mandible, the occiput and the shoulders. Pain and discomfort in the temporo-mandibular region and restricted movement of the scapulo-humeral joint often render this pressure unsupportable. Furthermore, it seems unlikely that a collar can hold a slip reduced (Sharp and Purser 1961). Consequently, operation was considered to be the treatment of choice.

Pre-operative period—All patients had skull traction applied through a Blackburn caliper before operation. As most patients were severely crippled by their rheumatoid arthritis, ambulant after-treatment was not considered. In these circumstances we felt that halo traction would be of no advantage and this technique was not used for this type of case.

The duration of traction varied widely. In three patients skull traction was applied for a few days before operation. In two of them (Cases 6 and 9) most of the signs and symptoms disappeared under nursing in a strictly supine position without traction. All of these patients had a mobile slip, with a distance of 4 or 5 millimetres between the anterior margin of C.2 and the posterior margin of the anterior arch of C.1 in extension. In one patient with severe neurological disturbance and gross lesions of the cervical spine (Case 7) traction was not expected to have any beneficial effect and was therefore applied only the day before operation.

In eleven patients skull traction was applied for one to ten weeks. In eight the neurological signs and symptoms subsided; in four the slip was reduced as compared with the position in extension before traction (Cases 4, 5, 8 and 12); and in the remaining four no reduction occurred (Cases 2, 3, 10 and 11). In three patients the neurological signs and symptoms remained unaltered (Cases 1, 13 and 14) and the slip was not reduced.

Surgical treatment was abandoned in two cases owing to bad general health involving too great an operative risk: Case 12 mainly because of severely reduced pulmonary function, and Case 14 because of a combination of extensive destruction in the cervical spine, the complete failure of skull traction, general oedema and the age of seventy.

Operation—This was performed in twelve patients (Table I). Operation for a slip of C.1 on C.2 consisted of occipito-cervical fusion using two autogenous bone grafts from the iliac crest (seven cases) or the tibia (five cases) held by steel wire. The tibial grafts did fuse except for Case 1 where a pseudarthrosis developed. When decompression was done by removing the lamina of C.1, with enlargement of the foramen magnum when necessary for pseudobasilar impression (Cases 2, 3 and 10), the grafts were fixed with wire both to the occiput and to the cervical vertebrae (Figs. 9 and 10). If decompression was not needed, C.1 was fixed with steel wire to C.2 or C.3 and a single broad graft was fixed to the vertebrae only (Figs. 11 and 12).

Post-operative treatment—After the operation most of the patients were nursed on a Stryker circo-electric bed with continuous skull traction for two to five months (average two and a half months). This required great patience and endurance from the patients and from all those
who cared for them. Routine physiotherapy was given. Consolidation of the graft proved difficult to verify radiographically. Our initial approach seems to have been over-cautious, and three months in traction has proved to be sufficient. Before removal of the traction device the patient was provided with a collar to control movement of the neck.

**Complications**—One patient (Case 6) died on the fifteenth day from pulmonary embolism. One patient (Case 2) developed quadriplegia immediately after the operation but this resolved after a fortnight. Infection of one of the caliper holes developed twice (in Cases 8 and 4, after two and five months respectively) and the Blackburn calipers had to be removed, much earlier than planned for Case 8. There were no wound complications except for one infected donor site (Case 7).

**RESULTS**

**Patients treated without operation**—As already mentioned, two patients (Cases 12 and 14) were not treated surgically because of their poor general condition. In Case 12 a good position of C.6 on C.7 was achieved by skull traction and the neurological symptoms and signs disappeared. A supporting shell for the occiput and back was applied with straps round the forehead and chest. About six months after discharge this patient died at home from an unknown cause, most probably a recurrence of cord compression. In Case 14 skull traction was not successful; the patient was discharged and died six months later from the complications of a cord lesion.

**Patients treated by operation**—In eleven patients the neurological alarm signals, such as severe neck pain, disturbed bladder function and jumping legs, disappeared and the motor power

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**FIG. 9**

Case 10. Figure 9—A lateral radiograph four years after operation. Two steel wires have been passed through drill holes in the occipital bone. One wire fixes the occipital bone to C.2. The other is used to fix the iliac bone grafts at both ends to the occipital bone and to the spinous process of C.3. Figure 10—A diagram to illustrate in principle the technique used when laminectomy of C.1 is done. The wires are either passed through drill holes in the occipital bone or through one drill hole and the occipital foramen. They are used to draw the occipital bone towards the spinous process of C.2 or C.3 and at the same time to fix the bone grafts at both ends.
soon increased. The paraesthesia of the fingers and feet did not always disappear and is designated as "few complaints". In one patient the "marble sensation" over the whole body remained.

**Surgical treatment of a slip of C.1 on C.2 (eleven patients)** (Table 1)—Eight patients with few or no neurological complaints at follow-up had sound fusion of the grafted area two to eight years after the operation, and neurological examination revealed only minor lesions. In four of these cases progression of the rheumatoid lesions was observed. The spine of one patient (Case 5) showed a slip at the C.2-3 level seven years after the operation. The patient died at home from a pneumonia eight years after the operation. In Case 2 gastric bleeding proved fatal six years after the operation; autopsy revealed ankylosis of C.1-2-3 and of C.4-5-6-7, and rheumatoid granulation tissue between C.3 and C.4 (Fig. 5). In Case 9 the lower part of the cervical spine ankylosed and the neck is now completely immobile. In Case 3 cord

*Fig. 11*
Case 8—A lateral radiograph taken after operation, when the lamina of C.1 was retained. The steel wire, forming a U-bend under the spinous process of C.3 and then encircling the lamina of C.1, finally transfixes the bone graft at two points before being twisted.

*Fig. 12*
A diagram to illustrate the technique of operation when the lamina of C.1 is retained.
symptoms recurred four years after operation due to a slip of 5 millimetres between C.7 and D.1. One patient (Case 1) had few persisting neurological complaints after the operation but eighteen months later they recurred; instability of the grafted area with pseudarthrosis and at lower levels narrowing of the cervical canal were found. This patient died suddenly at home two years after the operation, the probable cause being cord compression.

One patient (Case 7), with extensive changes in the upper cervical spine, was operated on for changes at the two levels C.1–2 and C.2–3. Sound bony fusion was obtained. The chief complaint of “marble sensation” of the body remained unaltered, but micturition improved as well as the spasms of the legs. The patient died from bronchial infection and cardiac insufficiency one year after operation.

**Surgical treatment of a slip at a lower level**—One patient (Case 13) treated surgically for changes at a lower level (C.4–5) had improvement immediately after the operation but slowly developed recurrence of all the symptoms of cord compression. This patient died from cardiac insufficiency a year after the operation.

**DISCUSSION**

Dislocation of the cervical spine in rheumatoid arthritis is well known. This can occur at all levels; in our series twelve slips were found at the level of C.1–2 (in three cases combined with C.2–3 slips) and two slips at a lower level (C.4–5 and C.6–7). In a hospital population the prevalence of a slip between C.1 and C.2 in rheumatoid arthritis varies between 18 and 34 per cent (Sharp and Purser 1961; Martel 1963; Conlon, Isdale and Rose 1966). In our rheumatism clinic (Sole Mio at Noordwijk) we found a similar figure of 28 per cent. Neurological signs and symptoms occur in only a minority of these rheumatoid patients: in two patients out of a series of eighty-nine described by Serre et al. (1963), in six out of twenty-six patients reported by Sharp and Purser (1961), and in nine out of nineteen mentioned by Crelin et al. (1970).

It is not yet possible to predict which patients will develop this complication. In a six-year follow-up study Isdale and Conlon (1971) frequently found an increase of the radiological changes in the cervical spine in the absence of neurological disturbances. Of their original 333 patients, 171 were still available for study; none of the fifty-nine deaths had been due to a cord lesion.

The onset of a cord lesion can be insidious as well as quite sudden. As soon as the patient’s complaints start to deviate from the usual, it is advisable to watch for alarm signs, when often the whole gamut will be disclosed. There may be a striking discrepancy not only between minimal neurological signs and symptoms and severe cervical dislocation, but also between numerous complaints and slight neurological findings, despite considerable disorganisation of the cervical spine.

The cause of the neurological symptoms and signs is not yet clear, but one possibility is impairment of the blood flow in the medullary vessels secondary to compression. Compatible with this idea is our observation that fixation of the spine can lead to good neurological improvement even with a seemingly unreduced slip. A case of probably long-standing cord damage was observed at autopsy on the patient (Case 6) who died on the fifteenth day after operation. This patient had nevertheless shown an impressive degree of neurological recovery while being nursed in the strictly supine position before operation.

Crelin et al. (1970) and Ball and Sharp (1971) have stressed the point that the cervical slip may interfere with blood flow in the vertebral arteries, thus giving rise to vestibular dysfunction, diplopia and other disorders. A case of obstruction of the vertebral artery was found at autopsy by Webb et al. (1968). In our series signs and symptoms indicating involvement of the vertebral artery were not observed.

In the C.1–2 area there are several joints, all of which can become affected by the rheumatoid process with destruction of the adjacent bones. Radiological signs may therefore
differ considerably. We distinguish two main types: a) a forward slip of the atlas in relation to C.2, and b) downward tilting of the atlas in relation to C.2.

Ball (1963) demonstrated in a normal spine that section of the transverse ligament will produce an atlanto-axial slip of 4 to 5 millimetres. If the atlanto-axial joint capsules are cut, the slip may increase to 10 millimetres. In our group of forward slips some measured almost 10 millimetres in flexion and others even exceeded this figure. It has to be accepted that in all of them not only the transverse ligament and the odontoid process but also the atlanto-axial joints are altered by the chronic inflammatory process.

In cases of a forward slip this could be demonstrated radiologically. We have observed two types of forward slip—mobile and relatively fixed. In the mobile type the distance C.1–C.2 is markedly reduced in extension (Cases 6, 8 and 9). In the more fixed type, the distance C.1–C.2 remains the same in extension or is slightly reduced (Cases 1 to 4). Fixation might be due to blocking of the atlanto-axial joints. In those with a tilted atlas, the atlanto-axial joints were severely destroyed and the odontoid process was usually eroded away. Slips only at a lower level in the cervical spine were encountered less frequently. They are probably due to a complex mechanism—destruction of the disc as well as of the adjacent intervertebral joints (Sharp et al. 1958, Ball 1963).

In several patients (Cases 1, 7 and 14) destructive lesions were found in the lower part of the cervical spine in addition to changes at the C.1–2 level. It was difficult to discern which of these changes—those below C.2 or those at the C.1–2 level were responsible for the cord lesion. The possibility of a summation effect cannot be excluded. In the most pronounced example of this type (Case 7) the vertebral bodies zigzagged and the arches of C.1, 2 and 3 were reduced to small remnants lying loose in fibrous tissue.

Crellin et al. (1970) found the severe slips predominantly at the C.4–5 level. In our comparable two cases the slips were localised at the C.4–5 and C.6–7 levels and no detectable radiological changes were found at C.1–2. The C.6–7 level is often not visible on routine radiographs. It is important to keep this fact in mind with regard to a rheumatoid patient having symptoms and signs of a cord lesion but an apparently normal cervical spine.

As already mentioned, fusion ultimately became the treatment of choice in this series. Unlike Crellin et al. (1970) and Bland (1967), we did not see any objection to the use of preoperative skull traction to reduce the slip, which also permits observation of any neurological improvement that would obviate laminectomy.

In some patients, even those with severe slips, lying supine continuously before skull traction was applied produced marked neurological improvement. In these cases the period of traction amounted to only a few days. But even then we think it advisable in order to accustom the patient to it and to the Stryker circo-electric bed in which he will be nursed for several months after the operation.

All but one of the twelve patients recovered from the operation. The immediate neurological results in all cases were very satisfactory. In one patient, however (Case 7), the subjective "marble sensation" remained unaltered. After one year and eighteen months, two patients (Cases 1 and 13) and after four years one patient (Case 3) had a recurrence of the neurological signs and symptoms. For the remaining patients the follow-up periods varied from one to eight years. They had only slight neurological complaints or none at all and were satisfied with the results, even when radiographs demonstrated progression of the rheumatoid lesions in the part of the cervical spine not operated upon.

SUMMARY

1. The treatment is described of a consecutive series of fourteen patients suffering from rheumatoid arthritis causing a slip of the cervical vertebrae, mainly at the C.1–2 level, and a cord lesion.
2. The importance of a change of symptoms and signs in these patients, especially the occurrence of the so-called “alarm signs”, is stressed.
3. In twelve patients the combination of this kind of cervical instability with a cord lesion was treated by fusion, most frequently of occiput to C.2 and 3.
4. In the eleven patients who survived the operation the results with a follow-up period of one to eight years are considered highly gratifying.
5. Technical details and various problems associated with treatment are discussed.

REFERENCES


