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# Intramedullary spinal cord tumours: a clinical outcome and radiological follow-up study

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## Summary

*Principles:* Intramedullary spinal cord tumours are rare. The long-term results depend on their varying natural histories and the surgical approach. Less extensive tumour resection avoids greater postoperative neurological impairment without a negative impact on postoperative outcome.

*Methods:* Twenty-seven patients who underwent a total of 34 surgical interventions (including 7 reoperations) were clinically and radiologically reinvestigated. Histology revealed 19 glial, 4 non-glial and 4 miscellaneous tumours.

*Results:* Postoperative long-term clinical follow-up (mean 62 months postoperatively) in 25 patients revealed functional improvement in 2 cases, stable conditions in 17 and deterioration in 6. Although there was residual tumour on MRI in 19 of the 22 patients reexamined, stable radiological studies were seen in 15 cases. Despite the high percentage of partial resections or biopsies, good long-term clinical results were found in 19 patients (70%).

*Conclusion:* The long-term outcome depends on tumour biology and the type of surgery. For low-grade astrocytomas we propose partial resection without incurring the risk of major postoperative neurological deficits, with semi-annual and, after 5 years, annual follow-up. Despite the fact that ependymomas are amenable to complete surgical resection, this was achieved in only one of six cases in this series. Postoperative MRI follow-up of intramedullary tumours must be protracted, as most of these tumours are slow-growing. An increase in the extent and intensity of contrast enhancement of the tumours was defined as tumour recurrence or progressive tumour growth.

*Key words: intramedullary tumour; spinal cord neoplasm; outcome*

## Introduction

Intramedullary spinal cord tumours (IMT) are rare and present with varying natural histories depending on their heterogeneous histological types. Resection techniques vary from biopsy to partial, subtotal, and gross total resection. Although there is disagreement in the literature concerning the extent of surgery, only a few long-term studies have been reported [1–11]. However, radical resection of intramedullary astrocytomas does not necessarily correlate with improved long-term outcomes [3, 4].

## Patients and methods

Twenty-seven patients with IMT, exclusive of filum terminale lesions, were treated between 1984 and 1998. Three had had prior biopsies performed at outside institutions but none of them had undergone radiotherapy before. There were 17 males and 10 females with an average age of 41 years. All patients gave informed consent.

A total of 34 surgical interventions were performed. Symptom duration averaged 37 months (range 3 weeks to 19 years). The most common complaint was back pain with paravertebral spasms (29/34, 85%), followed by sensory (28/34, 82%), motor (26/34, 76%), and sphincter complaints (4/34, 11%). Nocturnal pain occurred in 3 cases (8%). Preoperative signs are shown in table 2. Preoperative functional status was graded by the Cooper and Epstein scale [3, 12] taking both sensory and motor deficits into account (table 1).

**Table 1**

Scale of the functional status of upper and lower extremities [3, 12].

| | |
|---|---|
| **Function of the upper extremities** | |
| Grade 0: | Intact |
| Grade 1: | Sensory symptoms only |
| Grade 2: | Mild motor deficit with some functional impairment |
| Grade 3: | Major functional impairment in at least one upper extremity but upper extremities useful for simple tasks |
| Grade 4: | No movement or flicker of movement; no useful function |
| **Function of lower extremities** | |
| Grade 0: | Intact |
| Grade 1: | Walks independently but not normally |
| Grade 2: | Walks but needs cane or walker |
| Grade 3: | Stands but cannot walk |
| Grade 4: | Slight movement but cannot stand or walk |
| Grade 5: | Paralysis |

**Table 2**

Preoperative signs in 27 patients with 34 spinal surgeries.

| Clinical signs | number | |
|---|---|---|
| Sensory impairment | n = 30 | (88%) |
| Slight to moderate motor deficit | n = 21 | (61%) |
| Severe motor deficit | n = 7 | (20%) |
| Genitourinary dysfunction | n = 4 | (12%) |
| Spinal ataxia | n = 3 | (9%) |

Blind retrospective histological review of the tumour specimens was completed by one neuropathologist using the WHO classification system for central nervous system tumours [13]. The lesions were grouped into glial, non-glial and miscellanous tumours, as proposed by Jouvet [14].

Functional motor status was assessed in our outpatient clinic. At the same time patients were reevaluated radiologically by MRI. MR imaging follow-up examinations were performed at 1.5 T using superficial coils. The spine imaging protocol consisted of sagittal and axial spin-echo (SE) T1-weighted (TR: 450–600 msec; TE: 15–20 msec) and sagittal T2-weighted (Turbo-SE: TR: 3000–5000 msec; TE: 90–120 msec) images. Additionally enhanced sagittal and axial T1-weighted SE images were obtained employing the same T1 parameters. MR data were interpreted by one neuroradiologist who was blinded to the histology, surgical therapy and level of the primary lesion. In each patient the follow-up examinations were analysed before and independently of the preoperative scan and compared subsequently.

# Results

## Histology and initial surgery

The results of the histological diagnosis at initial surgery are summarised in table 3. Glial origin of the tumour was found in 70% (n = 19), non-glial in 15% (n = 4) and miscellaneous in 15% (n = 4) of the cases. In 2 of the 27 patients tumours with malignant growth patterns were found (both were recurrences of a glioblastoma multiforme of the thoracic spinal cord operated on previously in another clinic). Five astrocytomas were located in the cervical cord, two of them spreading into the medulla oblongata. Ependymomas were equally distributed (2 each in the cervical, thoracic and conus regions). Table 3 summarises the relationship between the extent of resection and histology. At initial surgery, cystic cavitation or syringomyelia was seen in 52% and an intratumoral haematoma in 19%.

## Histology at reoperation

A total of 7 patients required reoperation (time interval 1–65 months). Histological type or grade did not change except in one case of secondary rapid progressive malignant anaplastic ganglioglioma at T12 in a 17-year-old girl (time interval 20 months).

## Immediate postoperative functional outcome and adjuvant therapy

There was no operative mortality in this series. Pre- and immediate postoperative functional statuses following the 34 interventions (including 7 reoperations) are summarised in figure 1a and 1b. The functional outcome of the arms improved in one patient, remained stable in 31 cases and deteriorated in another 2. Sensorimotor function of the lower extremities improved in 3 cases, did not change in 24 patients and worsened in 7 patients in the immediate postoperative phase, though 6 of these patients described increased motor capacity not clinically objectifiable.

Six patients with astrocytomas and one patient with a malignant ganglioglioma received postoperative radiotherapy (38–56 Gy) without signs of myelopathy at follow-up.

## Long-term clinical follow-up

A total of 25 of the 27 patients were reinvestigated, the mean follow-up period being 62 months (3 months–14 years). Figure 2a and 2b is a synopsis of the long-term follow-up investigation. Long-term conditions improved in 2 patients, although another 2 patients described better motor function

**Table 3**

Degree of tumour resection in comparison to histological diagnosis at initial surgery. The tumours were classified using the WHO classification system for central nervous system tumours [13] and grouped as proposed by Jouvet [14].

| Tumour classification | number n = 27 | total resection n = 4 | partial resection n = 10 | biopsy n = 13 |
|---|---|---|---|---|
| Glial tumours | 19 | | | |
| – Astrocytoma | | | | |
| • Fibrillary, WHO 2 | 10 | | 3 | 7 |
| • Glioblastoma multiforme, WHO 4 | 2 | | | 2 |
| – Ependymoma | | | | |
| • Myxopapillary, WHO 1 | 2 | | 1 | 1 |
| • Classic, WHO 2 | 4 | 1 | 1 | 2 |
| – Ganglioglioma, WHO 2 | 1 | | | 1 |
| Non-glial tumours | 4 | | | |
| – Lipoma | 3 | | 3 | |
| – Haemangioblastoma | 1 | 1 | | |
| Pseudotumours | 4 | | | |
| – Epidermoid | 1 | | 1 | |
| – Dermoid | 1 | | 1 | |
| – Cavernoma | 2 | 2 | | |

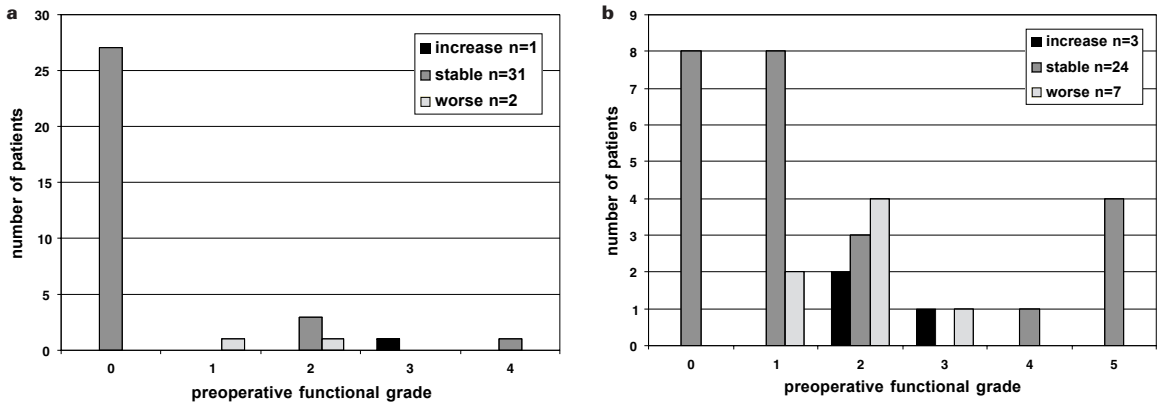


**Figure 1**

Preoperative and immediate postoperative neurological results following 34 spinal surgeries of a) the upper and b) the lower extremities. Results are classified according to the Cooper and Epstein scale [3, 12].

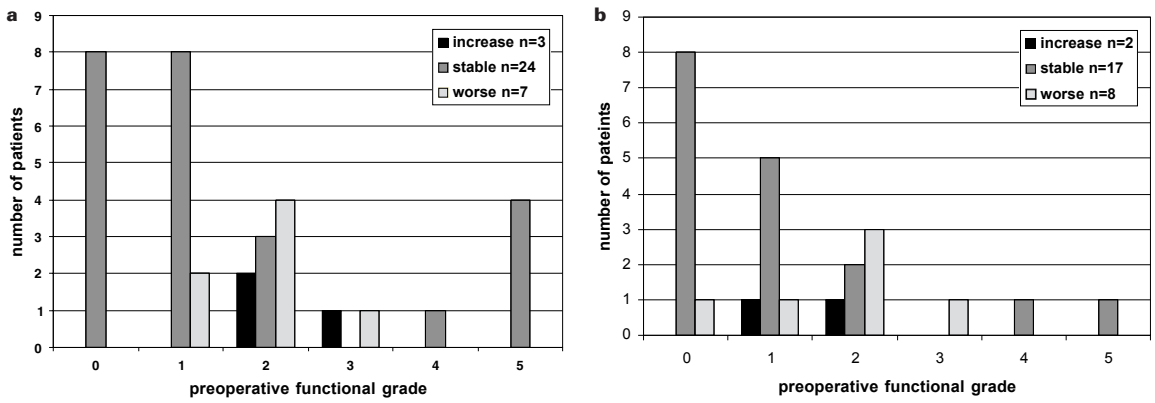


**Figure 2**

Preoperative and long-term functional status following 34 spinal surgeries of a) the upper and b) the lower extremities. Neurological results are classified according to the Cooper and Epstein scale [3, 12].

which could not be confirmed during clinical examination. Sensorimotor function of the upper extremities remained stable in 21 patients and decreased in 2 cases, whereas neurological findings in the lower extremities remained stable in 17 patients and decreased in 6. In 4 of 7 ependymomas the long-term outcome was either stable or increased motor function.

Six patients were pain-free at long-term follow-up. Four patients complained of marked urinary sphincter disturbance requiring catheterisation, while a further 4 had dysuria including sexual dysfunction.

Postoperative spinal kyphoscoliosis requiring stabilisation occurred in one patient following laminectomy T12 to L1 for resection of an astrocytoma.

Eight patients died during the follow-up period within 5 years of IMT diagnosis. All 3 patients with malignant IMT died within 2 years due to tumour-induced pulmonary dysfunction (involvement of the phrenic nerves, metastases into the lungs). Two patients with low-grade IMT died within 2 and 4 years of pulmonary dysfunction due to tumour progression.

## Radiological follow-up

Mean radiological follow-up by MRI of the 22 reinvestigated patients was 52 months (range: 6–132 months). In no case was there evidence of intradural seeding. In cases with no residual tumour the level of resection was observed as a region of segmental narrowing (figure 3).

In 15 clinically stable patients there was no increase in tumour size or diameter of the spinal cord at the level of the residual IMT. No tumour progression was noted in 9 reinvestigated low-grade

**Figure 3**

Grade II ependymoma at the level of T10–T12.
a. Sagittal T2-weighted images (left, middle) and postcontrast T1-weighted images (right). The initial (left) and follow-up examinations 3 years after biopsy (middle, right) demonstrate an inhomogeneous, partially cystic tumor with hyperintense signal on T2- and iso-/ hypointense signal on T1-weighted images. Central areas of signal loss are characteristic of haemosiderin due to repetitive haemorrhages. A progressive syrinx up to T6 can be detected on the follow-up examinations.
b. Sagittal T2-weighted images (left) and T1-weighted images (right) 4 years after complete tumour removal. A segmental spinal cord atrophy on the level of T9–T11 with a cystic intramedullary cavity can be seen.

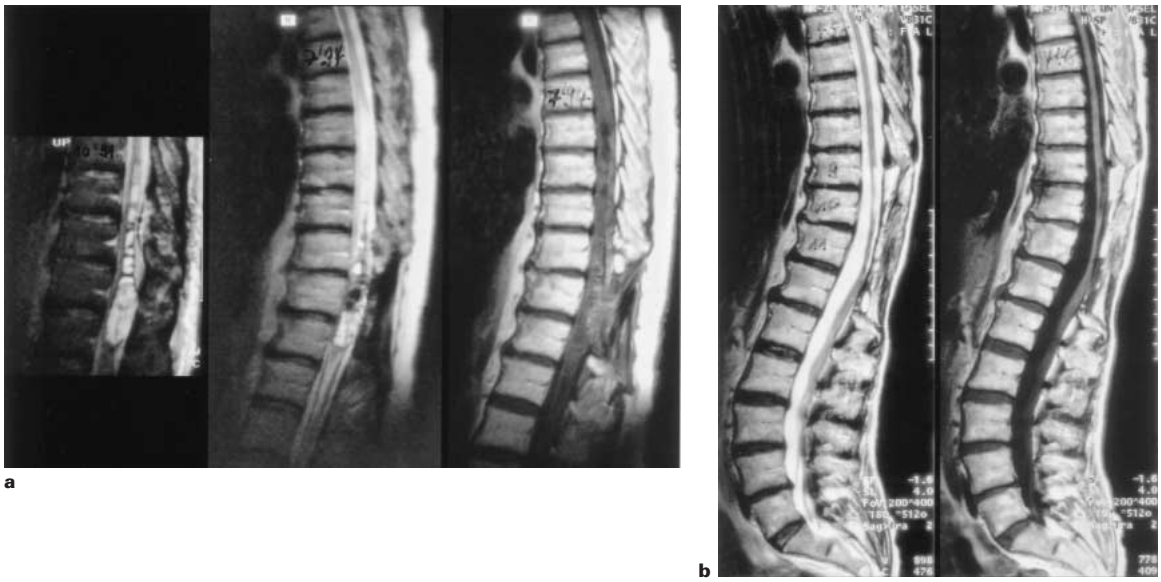

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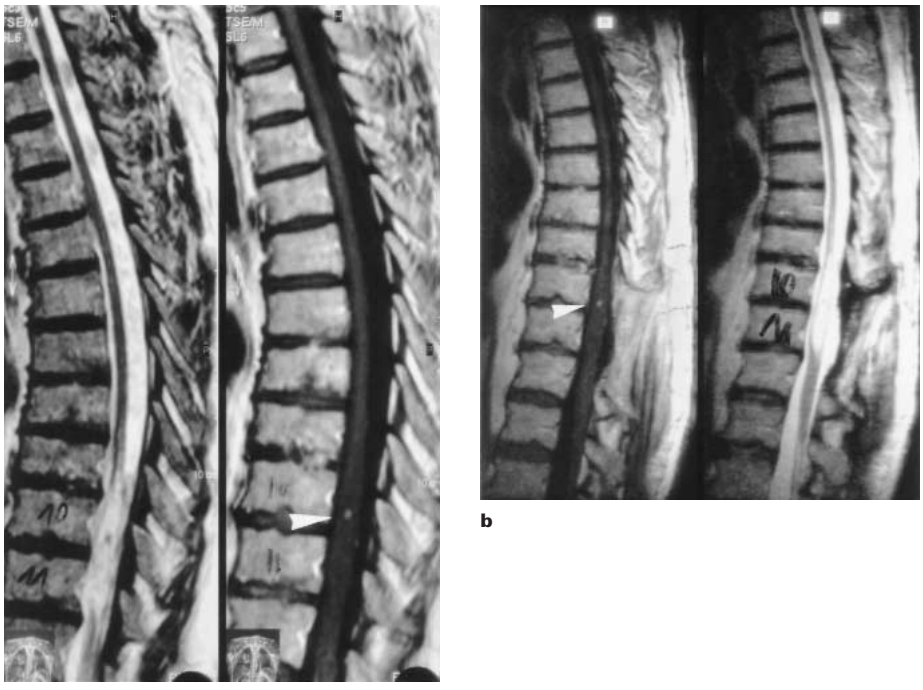
**Figure 4**

Grade II astrocytoma of the conus.
a. Preoperative sagittal T2-weighted images (left) and T1-weighted images after contrast application (right). T2-weighted images are capable of demonstrating the precise tumour limits of this high-signal lesion. On the T1-weighted images a small enhancing nodule within the lesion can be detected (arrowhead).
b. Sagittal T2-weighted images (right) and postcontrast T1-weighted images (left) on follow-up examinations 2 years after biopsy alone. There is no tumour progression in this clinically stable patient, the enhancing nodule is unchanged in size (arrowhead).



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astrocytomas of which 4 had undergone previous biopsy alone (figure 4). In 4 cases of tumour recurrence the diameter of the spinal cord increased segmentally, the craniocaudal extension was enlarged and the intensity of contrast enhancement was increased. Clinical deterioration was seen in all 4 recurrences. An accompanying syrinx was found in 13 reinvestigated IMT on MRI. In 2 cases a newly developed cystic tumour component could be seen. Oedema of the spinal cord was found only in astrocytomas.

## Discussion

We adopted a less aggressive approach to glial intramedullary tumours than that proposed by other authors [2, 5–7, 10–12, 15]. Although aggressive surgical management is avoided in many cases, the long-term results are good.

We noted a favourable long-term outcome in low-grade astrocytomas even though none of them was resected radically. On MRI there was no tumour progression in the reinvestigated low-grade astrocytomas. This observation favours less aggressive tumour removal in cases of poor cleavage and signs of benign histological behaviour. Having reviewed his final long-term results, Cooper points out that total tumour resection is unreliable and tumour recurrence is common despite radical resection [3]. A recent report confirms our finding that although microsurgical complete tumour removal may achieve long-term tumour control, an incompletely resected low-grade astrocytoma may also be associated with improved postoperative survival [9]. Other authors propose radical resection of these biologically unpredictable tumours [15]. However, in a recent paper the former group reports a better long-term outcome in subtotally compared to radically resected low-grade astrocytomas in young adults [2]. Postoperative radiotherapy is suggested as an effective mode of therapy [11]. In our opinion radiotherapy should be used very restrictively in low-grade astrocytomas. In agreement with the literature, we found no histological "upgrading" of astrocytomas in cases of second surgery [15]. It is not known why the change in malignancy from low-grade to high-grade glioma is not seen in spinal as in cerebral astrocytomas. The prognosis of malignant gliomas remains dismal. There is no evidence-based standard for treating high-grade spinal gliomas [2, 3].

In our series, 4 of 6 patients with an ependymoma presented with a stable or enhanced functional condition, even though in the majority only partial tumour removal was achieved. Patients complained primarily of back, interscapular or radicular pain, followed by dysaesthesia and motor weakness. None of our radiological follow-up investigations showed signs of tumour dissemination along the spinal cord. The incidence of intradural dissemination for primary spinal ependymomas is reported to be 12.5% [16]. In our series postoperative radiotherapy was employed only in cases with evidence of increased mitotic activity, but proved to be of no clear benefit. The role of radiotherapy in spinal ependymomas remains less clear [1, 5, 8, 9].

Gangliogliomas occur typically in young patients and usually present as slow-growing benign lesions [17]. However, malignant transformation, as in our case, is exceptional [18]. For cavernomas complete removal is recommended after onset of the first, even minor, symptoms, to avoid irreversible neurological damage following further haemorrhage [19–21].

We do not recommend radical resection of intramedullary lipomas, as their course is typically slow and attempted total resection markedly increases morbidity. However, they should regularly be followed up postoperatively by MRI to detect minor signs of morphological tumour recurrence before precipitous decline sets in. In agreement with another report, patients in our series with lipoma had a long and indolent history with a tendency to secondary rapid progressive decline [22]. In our series, patients with lipoma presented in their forties and none had signs of spinal dysraphism.

Severe postoperative spinal deformity is an important point to take into account in evaluating functional results. Postlaminectomy kyphoscoliosis depends not only on the extent of laminectomy but also on the tumour's location in relation to the innervation of the muscles attached to the spine. Adjuvant radiotherapy favours the development of postoperative spinal deformity [6]. Osteoplastic laminotomy may forestall the development of progessive spinal deformity. If spinal instability is suspected, one-step spinal stabilisation is recommended. Although the effects of surgical dural decompression alone may initially improve tumour-related results after partial resection or biopsy, this cannot explain favourable long-term results of a less aggressive approach.

Postoperative dysaesthesia and pain is a well known phenomenon and is often difficult to manage [6]. Postoperative hyperaesthesia and allodynia may be caused by disturbances of the dorsal columns, as tumour removal is usually performed by midline spreading and retraction of the posterior columns. Postoperative position-sense disturbance causing gait disturbance is usually transient and improves within 3 months [6].

We are fully aware that we approach the controversial questions surrounding the extent of resection of intramedullary gliomas in a much more conservative way than other authors [2, 5–7, 10–12, 15]. Aggressive resection reduces the risk of local tumour recurrence but involves a danger

of major postoperative neurological impairment. A disadvantage of our policy is that in the event of tumour progression the secondary, and then usually more aggressive, tumour resection is complicated by scar tissue of the first surgical intervention, whereas aggressive initial tumour resection does not face scar tissue-related problems. Advances in microsurgical techniques such as the ultrasound aspirator, laser, intraoperative ultrasound, and intraoperative neurophysiological monitoring facilitate radical tumour resection and obviate adjuvant treatment for low-grade IMT. Despite all these technological improvements our aim was to achieve resection of the tumour mass without the risk of neurological impairment, even when this implies partial resection or only biopsy.

In conclusion, we propose that in cases which are radiologically suspect for low-grade astrocytoma, histological diagnosis should be performed by biopsy or, if achievable, by partial tumour resection. In these cases the risk of major postoperative neurological deficits which may be associated with radical tumour resection should be avoided. In view of the slow-growing behaviour of these tumours, patients can be followed clinically and radiologically semi-annually and, after 5 years, annually. In the event of tumour progression a further resection can then be performed. While ependymomas are reportedly amenable to complete removal, due to their growth behaviour with a distinct tumour border compared with astrocytomas, in our series this was achieved only in 1/6. Cavernomas should be radically resected, whereas radical resection of lipomas may markedly increase postoperative morbidity.

Acknowledgment: The authors thank Alexandra Palatini and Daniel Sigg, MD for reviewing this manuscript.

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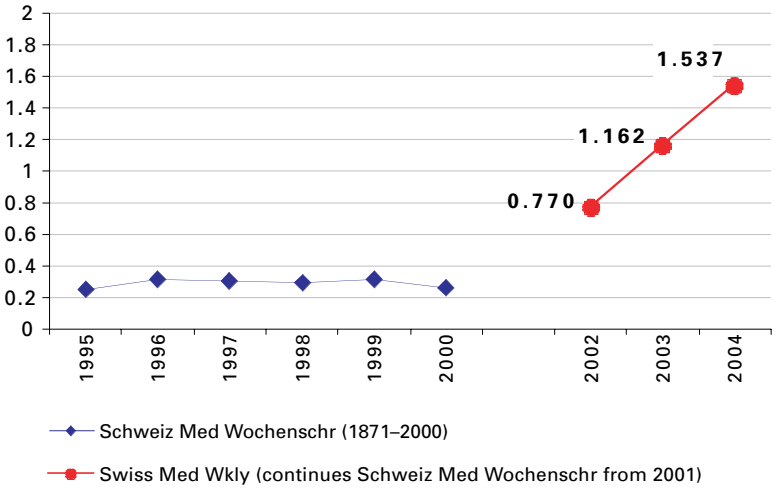


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