

# Intramedullary tumors in children

## Analysis of 24 operated cases

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### ABSTRACT

Intramedullary tumors are rare. The authors reviewed 24 cases operated between 1996 and 2006. The study assessed the clinical characteristics and surgical results based upon the neurological function. **Method:** Medical records of patients with intramedullary astrocytoma and ependymoma were reviewed. The minimal follow up time was 6 months and, at the end of this period, a comparative analysis of the neurological function was performed based using the McCormick scale score. **Results:** Most patients had astrocytoma (75%). Male gender was more prevalent (58.3%). The most common type of tumor was graded as I or II, and in three cases these were malignant. The total resection of the tumor was achieved in 20.8% of the cases. The statistical analysis did not show a statistically significant difference between preoperative and postoperative grades at McCormick scale. **Conclusion:** The authors concluded that microsurgery to intramedullary tumors did not significantly alter the neurological function after six months.

**Key words:** astrocytoma, ependymoma, microsurgery, spinal cord.

### Tumores intramedulares em crianças: análise de 24 casos operados

### RESUMO

Os tumores intramedulares são doenças raras. Os autores analisaram 24 casos operados entre 1996 e 2006. O estudo analisou as características clínicas e o resultado da cirurgia quanto à função neurológica. **Método:** Foram analisados pacientes com astrocitomas e ependimomas intramedulares. O tempo mínimo de acompanhamento foi de 6 meses e ao final deste período foi realizada a avaliação comparativa da variação do estado neurológico baseado na escala de McCormick. **Resultados:** A maioria dos pacientes era de astrocitoma (75%). O gênero masculino foi mais prevalente (58,3%). A maioria dos tumores era de grau I ou II, 3 casos eram malignos. A ressecção total do tumor ocorreu em 20,8% dos casos. A avaliação estatística demonstrou que não houve diferença significativa entre o estado neurológico na escala de McCormick pré-operatória e pós-operatória. **Conclusões:** Os autores concluem que a microcirurgia para ressecção dos tumores intramedulares não ocasionou variação funcional significativa nos pacientes após seis meses da cirurgia.

**Palavras-chave:** astrocitoma, ependimoma, microcirurgia, medula espinhal.

Spinal cord tumors represent 4 to 8% of central nervous system tumors in the pediatric population<sup>1</sup>. Despite this low prevalence, there are doubts about the clinical management of these cases. In children, astrocytomas are 3 times more frequent than ependymomas. Due to the slow growth in the natural history of many of these tumors, neurosurgeons usually do not make a definitive statement regarding

treatment<sup>2</sup>. In the pediatric population, at the time of diagnosis, some patients have few symptoms, leading to medical concerns regarding radical surgery<sup>3</sup>.

Advances in surgical techniques and the widespread availability of improved imaging techniques have enhanced the ability of neurosurgeons to perform radical resections of these tumors<sup>4</sup>.

This study aimed to analyze the neuro-

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logical outcomes after surgery performed to treat spinal cord tumors in children.

## METHOD

A consecutive series of 24 children, aged 1 to 17 years old at the time of diagnosis, with pathologically confirmed astrocytomas or ependymomas were studied. These children were treated at the Sarah Network of Rehabilitation Hospitals, from 1996 until December 2006. Inclusion criteria were: age below eighteen years old, diagnosis of intramedullary astrocytoma or ependymoma, and a follow up of six months or more. All of these cases were operated by the neurosurgical team at Sarah Hospitals. Patients who were operated at other hospitals, but followed in the rehabilitation spinal cord program, were evaluated by the neurosurgical team at Sarah Hospitals but were excluded from this study. Patients with other types of intramedullary tumors or those who had less than six months follow up after the surgical procedure were excluded from this assessment. Prior to surgery, all cases underwent magnetic resonance imaging (MRI). Following the surgical procedure, all patients underwent MRI scans to assess the volume of the resected tumor. The pictures were studied by independent group of radiologists of Sarah Hospitals.

The children were assessed using the following variables: age at diagnosis, type of the tumor, histological grade (World Health Organization classification), time of diagnosis, volume of tumor resection, McCormick grade before and after surgery. The preoperative functional status was assessed by McCormick classic scale. The postoperative outcome grades were performed at one and six months<sup>5</sup>.

Patients underwent laminectomy and, more recently, laminotomies, whenever the tumor extended to more than two vertebral segments. Both the dorsal myelotomy and the surgical resection were made using microsurgical techniques. The tumor dissection was carried out along the cleavage plane with the spinal cord for as far as it could be identified. Ultrasonic aspiration was used to excavate the tumor from inside out, until its interface with the white matter was reached. Sensory evoked and, recently motor evoked potentials were used routinely for on-line feedback to the surgeon.

Volume of surgical resection was classified in biopsy when only 50% of the tumor was resected, or when more than 50% was resected. When less than 90% of the tumor was removed, it was considered to have been a partial resection. A subtotal resection meant that more 90% of the tumor was resected, but the MRI showed some residual tumor. Total resection was achieved only when MRI did not show any residual tumor (Fig 1).

The statistical analyses were performed by the software SPSS version 13.0, with the assistance of the statistics group of Sarah Hospitals. The Wilcoxon test was used

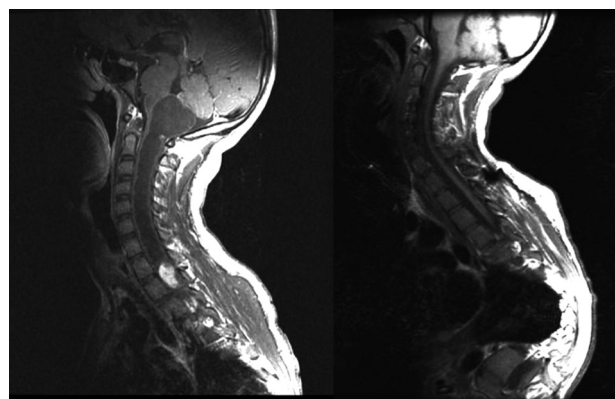


Fig 1. Preoperative and postoperative MRI showing total resection of the tumor in the cervical spinal cord.

Table. Clinical and surgical characteristics of patients.

Clinical features	Number	Percentage
Astrocytoma	18	75.0%
Grade I	8	33.3%
Grade II	8	33.3%
Grade III	2	8.3%
Ependymomas	6	25.0%
Grade I	3	12.5%
Grade II	2	8.3%
Grade III	1	4.2%
Tumor location		
Cervical	8	33.3%
Thoracic	13	54.2%
Lumbar	3	12.5%
Age		
0 to 3 years	4	16.7%
4 to 10 years	5	20.8%
11 to 18 years	15	62.5%
Scoliosis		
Yes	15	62.5%
No	9	37.5%
Sex		
Female	10	41.7%
Male	14	58.3%
Motor deficit		
Yes	19	79.2%
No	5	20.8%
Tumor resection		
Biopsy	0	0%
Partial	10	41.7%
Subtotal	9	37.5%
Total	5	20.8%

for analysis of variation in the clinical status after surgery, at one and six months.

## RESULTS

### Preoperative clinical status

Between 1996 to 2006, 24 patients aged under 18 years, with spinal cord tumors, were studied. These pa-

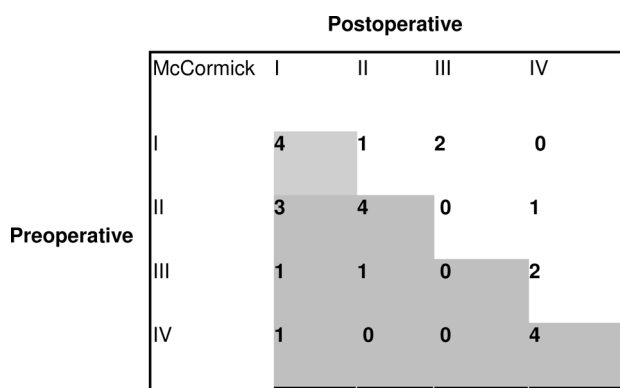


Fig 2. Diagram showing the relationship between preoperative and postoperative neurological function after six months.

tients were divided into 18 cases of astrocytomas and 6 cases of ependymomas. The average age of these patients was 11 years old. We divided the ages of patients in three groups. The population in the first group (0 to 3 years) consisted of only four cases, and most of patients were older than eleven years old (Table). The median time prior to the procedure was 14 months, varying from 28 days to 26 months. The Table provides a summary of the main clinical features in our patients. The most common histological type of tumor was astrocytoma grade I. No cases of anaplastic multiform glioblastoma were found in our series. Ependymomas were found only in patients older than 10 years. The most common location for tumors was the thoracic region (Table). Scoliosis was diagnosed in 15 radiological films. Motor deficit was identified in 19 cases at the time of admission. McCormick classification<sup>5</sup> was performed in all of cases to define the clinical preoperative status and it was used again for comparison with the postoperative status.

### Postoperative clinical status

All of these 24 patients were submitted to surgical treatment. Biopsies were not performed in 10 cases in which only a partial resection was achieved. Total resection was done in 5 cases and subtotal in 9 children (Table).

The McCormick scale was used to assess the surgical results and the impact of the procedure in the clinical status at one and six months after surgery (Fig 2). Most the patients maintained the same neurological function as in the preoperative assessment. For the group of children with preoperative grade 1 or 2 on the McCormick scale, three cases presented a worse grading in the postoperative assessments. Patients with preoperative grades 3 or 4 kept the same clinical status, with exception of three cases that showed improvement. In one case, a child with paraplegia, the ability to walk was recovered after surgery (Fig 2)

The statistical analysis performed by non parametric test showed no difference in the preoperative and postoperative groups ( $p=0.773$ ), six months after the surgery. This result showed that the patients kept the same neurological grade after the surgical procedure, and the surgery in itself was not responsible for any change in neurological function.

### DISCUSSION

Intramedullary tumors are rare in children, with an annual incidence of about one case per million. Astrocytomas and ependymomas are the most frequent intramedullary tumors, accounting for about 80% of all tumors in this area<sup>16</sup>. For the pediatric population, there is an increased number of spinal cord tumors in comparison with other spinal neoplasms<sup>7</sup>.

Surgical treatment is the initial approach for treating spinal cord tumors, with the objective of resecting most of the tumor, while attempting to keep the patient in the same neurological function grade<sup>8</sup>. Epstein published his work in spinal cord tumors showing the possibility of complete resection in most of 75% of his cases<sup>7-9</sup>. Nad-Kani and Rekate performed a systematic review of this subject, and no evidence was found that total resection of spinal cord tumors could improve de survival rates<sup>1</sup>.

The French multicenter study showed two prognostic factors: time to the diagnosis (relative risk – 4.93) and histological type of tumor (relative risk – 7.69)<sup>10</sup>. The other variables analyzed in that study did not show statistical difference, and gross total removal of intramedullary tumors did not correlate with increased survival rates<sup>10</sup>. More recently, another study from England with 453 adults patients showed that extension of tumor resection did not correlate with improvement in survival rates<sup>11</sup>. We believe that the aim of surgery in intramedullary tumors should be radical removal, although this is possible only in cases where there is cleavage plane. A clear cleavage plane is usually found in ependymomas, where radical tumor removal should be the objective of surgery<sup>5</sup>. Total removal is rarely possible in astrocytomas which, even when viewed under a microscope, rarely have a clear cleavage plane<sup>5</sup>. This is the reason for the better surgical outcome from spinal ependymomas in comparison with spinal astrocytomas.

The limitations on the use of radiotherapy and chemotherapy, as well as the fact that there is no clear evidence that these treatments may improve outcomes in children, have made surgery the best option for treatment of spinal cord tumors<sup>11,12,13</sup>. Concerns regarding neurological function are a very important issue. Most cases are astrocytomas grade 1 or 2, and the survival rate in this type of tumor is very good, so the clinical status after surgery in these patients will be of great importance to their quality of life<sup>10,14</sup>.

In the present study, it was observed that, after 6 months, the patients still maintained the same functional neurological grade on the McCormick scale. No statistical difference was noticed in the preoperative and postoperative groups. The present study used the McCormick scale to compare the neurological status, since this is the most common method used in the literature to assess clinical function after surgical resection<sup>5,15</sup>.

One hundred cases of spinal tumors were studied by Ferreira et al<sup>16</sup>. In this series of cases from Porto Alegre, ependymomas (4%) and astrocytomas (2%) were reported<sup>16</sup>. The pediatric population differs from the adult cases. Taricco et al published results from a series of forty-eight patients with primary spinal cord tumors. The thoracic cervical cord was most common site involved, and microsurgical removal was achieved in 71% of the patients without added neurological morbidity<sup>17</sup>. Ependymoma was the most common tumor found in their paper. This is an important difference, since the pediatric population presented astrocytoma as the most frequent tumor, and resection of these lesions is more complex. Koerbel et al studied 35 patients with spinal cord tumors<sup>18</sup>. Total resection was more often obtained among patients with ependymomas (13 out of 17) than with astrocytomas (5 out of 12). However, the degree of resection and tumor histology did not interfere in the postoperative clinical morbidity, although the authors found that the tumors located at the thoracic level were associated with higher morbidity ( $p=0.021$ )<sup>18</sup>.

This study collected 24 cases with spinal cord tumors in pediatric patients. There is no comparable series of cases in the Brazilian literature, and the present results showed the importance of surgery, which did not alter the neurological function of the patients.

In conclusion, the surgical resection did not alter the neurological function six months after surgery. Early diagnosis and treatment improved the surgical morbidity rate. The strongest predicting factor of functional outcome was the preoperative clinical status, as assessed by the McCormick scale. The neurosurgical team aimed to remove most of the tumor, while maintaining neurological function.

The larger numbers of patients followed for longer periods is helping to clarify the results from surgery for spinal cord tumors. It is important to emphasize that the natural history of these tumors is variable, affecting even patients who were considered to have had total tumor removal. With longer follow-up periods, the natural history and the effect of the surgical treatment may become clearer in the future.

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