Resection of intramedullary spinal cord tumors in children: assessment of long-term motor and sensory deficits

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Object. With modern surgical advances, radical resection of pediatric intramedullary spinal cord tumors (IMSCTs) can be expected to preserve long-term neurological function. Nevertheless, postoperative neurological decline is not uncommon after surgery, and many patients continue to experience long-term dysesthetic symptoms. Preoperative predictors of postoperative neurological decline and sensory syndromes have not been investigated and may serve as a guide for surgical risk stratification.

Methods. Neurological function (as determined using the modified McCormick Scale [mMS]) preoperatively, postoperatively, and 3 months after surgery was retrospectively recorded from patient charts in 164 consecutive patients undergoing resection of IMSCTs. A median 4 years (interquartile range [IQR] 1–8 years) after surgery, long-term motor and sensory symptoms were assessed by telephone interviews and corroborated by subsequent medical visits in 120 available patients. This long-term assessment was retrospectively reviewed for the purposes of this study. The authors reviewed this series to assess long-term motor, sensory, and urinary outcomes and to determine independent risk factors of postoperative neurological decline and long-term sensory dysfunction.

Results. Patients were 8.6 ± 5.7 years old and presented with a median mMS of 2 (IQR 2–4). Three months after surgery, 38 patients (23%) continued to experience decreased neurological function (1 mMS point) incurred perioperatively. Increasing age (p = 0.028), unilateral symptoms (p = 0.046), and urinary dysfunction at presentation (p = 0.004) independently predicted persistent 3-month perioperative decline. At long-term follow-up (median 4 years), 39 (33%) exhibited improvements in their mMS scores, 13 (30%) had improvement in their urinary dysfunction, and 27 (30%) had resolution of their dysesthesias. Seventy-eight patients (65%) experienced long-term dysesthetic symptoms. Increasing age (p = 0.024), preoperative symptom duration ≥ 12 months (p = 0.027), and worsened postoperative mMS score at hospital discharge (p = 0.013) independently increased the risk of long-term dysesthesias.

Conclusions. In the authors' experience, nearly one third of patients may experience improvement in motor, sensory, and urinary dysfunction years after IMSCT resection, whereas the majority will continue to experience long-term dysesthetic symptoms. Improvement in motor deficits preceded improvement in sensory syndromes, and urinary dysfunction typically resolved much longer after surgery. The risk of persistent perioperative motor decline was increased with older age, unilateral symptoms, preoperative urinary symptoms, and less severe preoperative neurological deficit.

The risk of long-term dysesthesias was increased with older age, increased duration of symptoms prior to resection, and greater postoperative neurological deficit. (DOI: 10.3171/PED-08/01/063)

Key words: intramedullary spinal cord tumor, outcome, pediatric neurosurgery, predictor

Intramedullary spinal cord tumors account for ~35% of all intraspinal tumors in children.1,2,16 Historically, treatment of these tumors consisted of biopsy, bone decompression, and/or radiation therapy. This approach resulted in only moderate improvements in slowing disease progression. Modern advances in microsurgical technology have allowed for increasingly aggressive resection of these tumors, often permitting radical resection3,4,8–10 and resulting in increased long-term survival and improved quality of life.3,4,8–10,15

Despite improvements in surgical outcomes, overall morbidity following IMSCT resection remains relatively high.3,14 Acute postoperative neurological decline may occur in up to a third of patients after surgery, although many patients continue to experience long-term dysesthetic symptoms despite successful IMSCT resection.3,4,5,11,15 Studies identifying preoperative predictors of neurological outcome and long-term dysesthesias in the pediatric population remain relatively unstudied. In this study, we characterize changes in motor and sensory deficits as a function of time after surgery and set out to ascertain preoperative predictors of postoperative motor...
and sensory decline, which may serve as a guide for surgical risk stratification.

Clinical Material and Methods

One hundred sixty-four consecutive patients with IMSCTs surgically treated between 1980 and 1994 at a single academic institution were interviewed and examined at initial presentation, at hospital discharge, and at the 3-month follow-up visit. All pre- and postoperative clinical and imaging variables were retrospectively recorded from patient charts. Patients underwent pre- and postoperative MR imaging in all cases. Cases with no residual enhancement noted on postoperative MR imaging were classified as radical resection. Cases with residual enhancement noted along the resection cavity were defined as subtotal resection.

Neurological function (as graded using the mMS [Table 1]) was retrospectively assessed from the physical examination and patient interview documented in the medical charts of patients from their preoperative, postoperative, and 3-month clinic visits after surgery. Patients experiencing a decline in neurological function (by $\geq 1$ mMS point) after surgery and not returning to baseline by 3 months of follow-up were classified as experiencing "postoperative neurological decline." Long-term assessment was available for 120 patients. For long-term assessment, patients and their families were contacted by phone by a trained nurse practitioner, and they completed a questionnaire assessing neurological function, paresthesia, and dysesthesia. The medical records of patients were obtained to confirm patient questionnaires and phone interviews near the time of the phone interview. The data collected from this long-term assessment were later retrospectively reviewed, and mMS and Karnofsky Performance Scale scores were assigned for the purposes of this study. Improvement in motor, sensory, or urinaiary function was based on the patients’ perception of their function. Dysesthetic symptoms were defined as sensory discomfort arising from burning, tingling, or hyperesthesia.

Surgical Technique

Children undergoing primary or revision surgery underwent laminectomy spanning the length of the tumor in all cases. Only medial facet joint exposure was needed in the vast majority of cases and an effort was made to preserve the facet joint capsule in all cases. Facetectomy was not performed in any case. For the last 20 cases during the study period, children who underwent surgery for their IMSCT, respectively. Of the patients who underwent prior surgery, 76 (46%) had undergone biopsy and 27 (16%) had undergone prior resection. The median preoperative mMS score was 2 (IQR 2–4). Symptoms were present for $<1$ month prior to surgery in 24 patients (15%) and $>12$ months in 47 (29%). One hundred forty-nine patients (91%) presented with motor symptoms, 120 (73%) with sensory symptoms, and 53 (32%) with urinary incontinence. Motor deficits were unilateral in 45 patients (27%) and involved only the upper extremities in 19 (12%). The tumor involved $6 \pm 3$ levels (mean $\pm$ standard deviation). One hundred eighteen patients (72%) had a tumor-associated syrinx noted on MR images. Twenty-four tumors (15%) were confined to the cervical spine, and 42 (26%) were confined to the thoracic spine. Sixty-four (39%) involved the cervicothoracic spine and 40 (24%) the thoracolumbar spine. Six tumors (4%) involved both the cervicothoracic and thoracolumbar spine.

Radical resection was achieved in 125 patients (76%) and subtotal resection in 33 (20%). Six patients (4%) underwent open biopsy only. The number of surgical levels was $7.5 \pm 3$ (mean $\pm$ standard deviation). Pathologically, 66 tumors (40%) were World Health Organization Grade I or II, 14 (8%) were Grade III, and 4 (2%) were Grade IV astrocytoma. In addition, there were 44 gangliogiomas (27%), 19 ependymomas (12%), 10 ganglioneurocytomas (6%), 3 glioleioblastomas (2%), 3 myxopapillary ependymoma (2%), and 1 primitive neuroectodermal tumor (1%).

Statistical Analysis

For intergroup comparison the Student t-test was used for parametric data, and the Mann–Whitney U-test was used for nonparametric data. Percentages were compared via chi-square tests. The univariate association of all recorded variables to postoperative neurological decline and long-term sensory syndromes was assessed via logistic regression analysis (Statview, SAS, Inc.). To assess independent risk factors of postoperative neurological decline and long-term sensory syndromes, all variables with a p value $< 0.10$ in univariate analysis were included in a multivariate logistic regression model (Statview). Variables with a p value $> 0.05$ were removed from the multivariable model.

Results

Patient Population

One hundred sixty-four patients underwent surgery for an IMSCT during the reviewed period and were observed for a median of 4 years (IQR 1–8 years). The mean age at the time of surgery was $8.6 \pm 5.7$ years, and 100 (61%) were boys. In addition, 13 (8%), 48 (29%), and 103 (63%) patients had surgery for their IMSCT, respectively. Of the patients who underwent prior surgery, 76 (46%) had undergone biopsy and 27 (16%) had undergone prior resection. The median mMS score was 2 (IQR 1–4). Symptoms were present for $<1$ month prior to surgery in 24 patients (15%) and $>12$ months in 47 (29%). One hundred forty-nine patients (91%) presented with motor symptoms, 120 (73%) with sensory symptoms, and 53 (32%) with urinary incontinence. Motor deficits were unilateral in 45 patients (27%) and involved only the upper extremities in 19 (12%). The tumor involved $6 \pm 3$ levels (mean $\pm$ standard deviation). One hundred eighteen patients (72%) had a tumor-associated syrinx noted on MR images. Twenty-four tumors (15%) were confined to the cervical spine, and 42 (26%) were confined to the thoracic spine. Sixty-four (39%) involved the cervicothoracic spine and 40 (24%) the thoracolumbar spine. Six tumors (4%) involved both the cervicothoracic and thoracolumbar spine.

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**TABLE 1**

The mMS for functional evaluation of patients with IMSCTs

<table>
<thead>
<tr>
<th>Grade</th>
<th>Explanation</th>
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<tbody>
<tr>
<td>I</td>
<td>Neurologically intact, ambulates normally, may have minimal dysesthesia</td>
</tr>
<tr>
<td>II</td>
<td>Mild motor or sensory deficit, patient maintains functional independence</td>
</tr>
<tr>
<td>III</td>
<td>Moderate deficit, limitation of function, independent w/ external aid</td>
</tr>
<tr>
<td>IV</td>
<td>Severe motor or sensory deficit, limit of function w/ a dependent patient</td>
</tr>
<tr>
<td>V</td>
<td>Paraplegic or quadriplegic, even if there is flickering movement</td>
</tr>
</tbody>
</table>

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M. J. McGirt et al.
Motor and sensory outcomes after resection of IMSCTs

Three-Month Neurological Outcome

Three months after surgery, the median mMS score was 3 (IQR 2–4). Thirty-eight patients (23%) had experienced a neurological decline by ≥ 1 mMS point compared with the preoperative score, 99 (60%) remained stable, and 27 (16%) demonstrated an improvement in mMS score. Each increasing year in patient age at the time of surgery was independently associated with a 9% increase in the odds of 3-month postoperative decline (Table 2). Less severe preoperative neurological deficit (each decrease in preoperative mMS score) was independently associated with a 68% increase in the odds of 3-month postoperative decline (Table 2). Unilateral symptoms and urinary dysfunction at presentation also increased the odds of 3-month postoperative decline (Table 2). All other clinical, imaging, and operative variables were not associated with 3-month neurological decline.

Long-Term Neurological Outcome

At a median of 4 years (IQR 1–8 years) after surgery, 120 patients (73%) were available for long-term assessment (34 died, and 10 were lost to follow-up). In these patients, long-term mMS scores were improved compared with 3-month mMS score (2 [IQR 2–4] compared with 3 [IQR 2–4], p = 0.07). A high functional status was maintained in these patients with a median Karnofsky Performance Scale score of 90 (IQR 80–100). Thirty-nine patients (32%) demonstrated an improvement in mMS score, 13 (11%) demonstrated an improvement in urinary symptoms, and 27 (22%) demonstrated an improvement in dysesthetic symptoms. Improvement in motor deficits preceded improvement in dysesthesia, and resolution of urinary dysfunction typically occurred at a longer time after surgery. Improvement in motor deficits preceded improvement in dysesthesia, whereas resolution of urinary dysfunction typically occurred at a longer time after surgery. Increasing age, presence of unilateral symptoms, and urinary dysfunction at presentation independently predicted 3-month postoperative neurological decline. At long-term follow-up, 65% of the patients who underwent IMSCT resection experienced long-term dysesthetic symptoms. These dysesthetic symptoms usually became evident within the 1st month after surgery. Typically, these patients had a component of sensory dysfunction preoperatively that worsened with surgery and then subsequently resolved or persisted to some degree. De novo sensory syndromes typically did not occur late after resection. Increasing age, preoperative symptom duration > 12 months, and worsening postoperative mMS score independently increased the risk of these long-term dysesthesias syndromes.

Discussion

In this study, we retrospectively analyzed a cohort of 164 consecutive patients to determine independent risk factors of postoperative neurological decline and long-term sensory dysfunction. Three months after surgery, 23% experienced a neurological decline, 60% remained stable, and 16% demonstrated an improvement in their mMS score. Improvement in motor deficits preceded improvement in dysesthesia, whereas resolution of urinary dysfunction typically occurred at a longer time after surgery. Increasing age, presence of unilateral symptoms, and urinary dysfunction at presentation independently predicted 3-month postoperative neurological decline. At long-term follow-up, 65% of the patients who underwent IMSCT resection experienced long-term dysesthetic symptoms. These dysesthetic symptoms usually became evident within the 1st month after surgery. Typically, these patients had a component of sensory dysfunction preoperatively that worsened with surgery and then subsequently resolved or persisted to some degree. De novo sensory syndromes typically did not occur late after resection. Increasing age, preoperative symptom duration > 12 months, and worsening postoperative mMS score independently increased the risk of these long-term dysesthesias syndromes.

TABLE 3
Independent predictors of long-term dysesthetic symptoms a median of 4 years (IQR 1–8 years) after resection of pediatric IMSCTs

<table>
<thead>
<tr>
<th>Variable*</th>
<th>Multivariate Logistic Regression</th>
</tr>
</thead>
<tbody>
<tr>
<td>OR (95% CI)</td>
<td>p Value</td>
</tr>
<tr>
<td>increasing age (yrs)</td>
<td>1.09 (1.01–1.19)</td>
</tr>
<tr>
<td>preop symptoms &gt;12 mos</td>
<td>2.82 (1.12–7.06)</td>
</tr>
<tr>
<td>increasing postop mMS</td>
<td>1.59 (1.11–2.26)</td>
</tr>
</tbody>
</table>

* These variables independently increased the risk of long-term dysesthetic symptoms.
The ultimate goal for patients undergoing surgery for IMSCT is to preserve neurological function. After resection, a minority of patients experience postoperative neurological decline. In the pediatric population, this rate varies between 10 and 30% in several studies, which is consistent with this study. The authors of these prior studies, however, did not perform statistical analyses to evaluate independent factors that may predict neurological decline following IMSCT resection. An additional complication of IMSCT resection is the development of long-term dysesthesias. The incidence has been reported to be as high as 70% in some studies. Studies investigating risk stratification of postoperative sensory syndromes are lacking.

Increasing age at the time of surgery was associated with an increased risk of postoperative neurological decline. As with the brain, a less mature spinal cord may possess increased plasticity and adaptability to compensate for neurological injury. It has been shown that infants and young children who sustain brain injuries can adopt alternative profiles of neurological organization to minimize the effects of the initial injury on neurological function and development. Therefore, a less mature spine, as with the brain, may have a better ability to compensate for injuries incurred during surgery or as a result of the tumor itself. In fact, Constantini et al. reported that the conditions of 7% of the children they studied who were <3 years of age deteriorated postoperatively, compared with another study in which the conditions of ~24% of the patients <21 years of age deteriorated postoperatively. This study supports the feasibility of pursuing resection in younger children as documented in previous studies, as well as advocates for pursuing surgery when the children are younger to maximize the chance of preserved neurological outcomes.

The presence of unilateral symptoms was associated with higher rates of neurological decline following resection. This may be due to a combination of factors. Unilateral or eccentrically located tumors may predispose the normal hemicord to increased risk of surgical injury, where inadvertent injury occurs with either resection or manipulation of normal neurological tissue. On the other hand, centrally located tumors are usually technically easier to resect and are associated with less risk of injury to adjacent neurological tissue, when using an inside-out technique, as described previously. Histologically, astrocytomas, as opposed to ependymomas, are also typically eccentrically located and often present with unilateral symptoms. Astrocytomas, due to their infiltrative nature, are associated with worse neurological outcomes after resection.

Urinary dysfunction prior to surgery was also associated with a higher incidence of perioperative neurological decline. Urinary dysfunction is not commonly a presenting symptom for patients with IMSCTs, with rates that range between 10 and 20% in several studies. Urinary dysfunction is often a late clinical finding and may signify advanced disease. In our series, urinary symptoms never occurred in isolation but were always associated with motor and sensory symptoms. Hence, urinary symptoms only occurred when both motor and sensory symptoms had already developed, marking more advanced disease. Our findings support early surgical intervention before urinary incontinence occurs to maximize the chance of preserved and/or improved neurological outcome.

Interestingly, patients with less severe preoperative neurological deficits were more likely to develop postoperative neurological decline. We hypothesize that this may be due to the fact that subtle changes following resection may be more apparent in patients who have normal or close-to-normal functioning. This may also mean that patients with normal preoperative function may actually be compensating, and thus, masking any subtle underlying neurological deficits. Surgical cord manipulation may cause these patients to decompensate more easily. Patients with more severe neurological deficits preoperatively may require a greater degree of surgical cord injury to manifest a decline in an entire mMS grade. Similar to independently predicting neurological decline, increasing age at the time of surgery also independently predicted the presence of long-term dysesthesias. As with the brain, a less mature spinal cord may possess an increased ability to adapt to neurological injury. This ability to compensate may be the reason why younger patients are less likely to incur long-term dysesthesias. In fact, prior studies have shown that the most common symptom in adult patients with IMSCTs is sensory dysesthesias with values that range between 50 and 70%. Much lower rates of dysesthesias are reported in studies on children (~30%).

Increasing duration of preoperative symptoms was also independently associated with long-term dysesthesias. This finding not only supports the need for early recognition of symptoms but also advocates for prompt resection to minimize the chance that patients will develop long-term dysesthesias. Authors of prior studies have also advocated the need for early surgical intervention. These authors contend that surgery should be pursued before the onset of severe neurological deficits, given that most children remain at their same preoperative neurological status following resection. Therefore, although authors of other studies have stressed the need to pursue surgery to preserve neurological function, we have focused on the need for prompt surgery to prevent the development of debilitating dysesthesias.

Finally, a worse postoperative mMS score was independently associated with the presence of long-term dysesthesias. The mMS is a scale designed to reflect the functional status of a patient with an IMSCT and does not necessarily incorporate the presence of dysesthetic symptoms. This study shows that patients who incur postoperative motor function decline will also likely develop long-term dysesthesias, sometimes regardless of subsequent motor improvement. This demonstrates that neurological function and the presence or absence of dysesthesias may be interrelated.

The importance of intraoperative neuromonitoring cannot be over emphasized. We use both muscle and epidural electrode monitoring in all cases for MEPs. This gives us immediate feedback with regard to tumor–spinal cord interface to guide the degree of resection. We have found that a sustained decrease in MEPs <50% of the epidural or D-wave baseline will likely result in temporary postoperative motor changes, whereas a greater degree of MEP change results in permanent deficits. The intraoperative monitoring, however, has had no predictive value for postoperative sensory dysfunction. This is particularly true for cases of postoperative dysesthesia syndromes, which typically occur shortly after surgery; we initiate treatment with neurotin as soon as symptoms present and escalate doses as needed. In these patients, subsequent improvement in sensory syn-
Motor and sensory outcomes after resection of IMSCTs

dromes vary greatly and appear to be associated with the factors reported in this study.
In addition, the laminectomy and/or laminotomy should be as limited as possible to provide adequate exposure but not to remove the more posterior elements as necessary to help avoid the future spinal deformity.

Conclusions
In our experience, nearly one third of patients may experience improvement in motor, sensory, and urinary dysfunction years after IMSCT resection, whereas the majority will continue to experience long-term dysesthetic symptoms. Improvement in motor deficits precedes improvement in dysesthesias, and urinary dysfunction typically resolves at a longer duration after surgery. The risk of motor decline is greatest in older children who have unilateral symptoms, preoperative urinary symptoms, and less severe preoperative neurological deficit. The risk of long-term dysesthesias is greatest in older children who have increased duration of symptoms prior to resection and more severe perioperative neurological deficit. Early resection of IMSCTs after symptom onset and at an earlier age may be associated with less long-term motor and sensory deficits. Patients with motor function decline after surgery may be at an increased risk of developing long-term dysesthesias.

Acknowledgment
We would like to recognize Fred Epstein, M.D., for his role as senior-most surgeon in the care of all patients included in this paper.

References

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