

The Current Management of Intramedullary Neoplasms in Children and Young Adults

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SUMMARY

AIM: The authors discuss the current management of intramedullary spinal cord tumors in children and young adults. Intramedullary spinal cord tumors of the pediatric population are rare and comprise only a small percentage of all central nervous system neoplasms. There has been much controversy with the surgical management of these tumors.

METHODS: The majority of these neoplasms are histologically benign with large percentage being astrocytomas or gangliogliomas. Ependymomas, which are common in adults, are relatively rare in children. The majority of these neoplasms occur in the cervical region with pain or a motor deficit as the chief complaint. Surgery on these neoplasms can be performed with safety using surgical adjuncts such as the ultrasonic aspirator, contact laser and neurophysiological monitoring.

RESULTS: A radical resection of these tumors results in a good long-term outcome, since the majority is histologically benign. Adjuvant radiation therapy should only be administered for the high grade or malignant tumor. Malignant tumors have a dismal outcome and surgery in these patients should be a conservative debulking. Spinal deformity is a concern in these children as a 35% of children may require a stabilization procedure.

CONCLUSION: Radical surgery for intramedullary neoplasms with the avoidance of adjuvant radiotherapy results in long-term survival with minimal morbidity.

KEYWORDS: Intramedullary, Neoplasm, Pediatric, Spinal Cord

INTRODUCTION

Intramedullary spinal cord tumors are rare central nervous system neoplasms. In particular the intramedullary location is an uncommon site for tumors. Only 35-40% of all intraspinal tumors in children are found at the intramedullary site. After several

early reports advocating aggressive surgery, postoperative neurological deficits were significant. Thereafter, many neurosurgeons recommended a conservative strategy with biopsy, dural grafting and radiation therapy regardless of histological diagnosis and age of the patient (47). With the advent of modern neurosurgical

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instruments, operating microscope, imaging technology, and intraoperative neurophysiology, the treatment strategy for these intramedullary neoplasms has again become more aggressive. This is particularly important as the majority of intramedullary tumors are histologically benign (8, 35) and the radical removal results in long-term survival with an acceptable morbidity (8, 11, 14, 15, 17, 22, 23).

In a recent study of 164 children with intramedullary spinal cord tumors operated on by the senior author (FE), the median age at presentation was 10 years (Table 1). The male to female ratio was 1:1.1. The majority of tumors were histologically benign (8). Two-thirds of these tumors were astrocytomas or gangliogliomas. The remainder was ependymomas, mixed gliomas, and miscellaneous tumors. Ependymomas were not found in any child less than 3 years of age. A malignant or high grade tumor was seen in only 9% of children.

A review of the intramedullary spinal cord database at a single institution between 1991 to 1998 yielded 294 cases in adults and children (35). The 294 tumors included 117 removed from children under the age of 21 years, and 177 from patients 21 years and older (Table 2). The majority of these pediatric tumors were operated upon by the senior author (FE). The most common single tumor type in this study population was the fibrillary astrocytoma, which

accounted for 45 or 39% of all tumors. There were 31 gangliogliomas, 19 ependymomas, 5 of which were examples of myxopapillary ependymomas. In this large number of children there were only 13 high-grade astrocytomas.

Age	10.4 yrs
Sex	
males	79
females	85
Location	
cervico-medullary	14
cervical	26
cervico-thoracic	44
thoracic	64
conus	16
No. of vertebrae levels	5.4

Table 1: Characteristics of 164 children with intramedullary spinal cord tumors

CLINICAL PRESENTATION

Intramedullary tumors may remain asymptomatic for a long time or cause nonspecific complaints, which make the diagnosis difficult in children. The onset of symptoms is often insidious and symptoms are typically present for many months. High grade or malignant neoplasms typically have a shorter prodrome (median, 4.5 months). Some children

may complain of symptoms following a trivial injury. The most common symptom is pain. The pain may be diffuse or radicular in nature. Thus there is no characteristic feature for the pain in children with intramedullary tumors. Young infants may even present with abdominal pain and undergo extensive gastrointestinal investigations before the diagnosis of intramedullary tumor.

Children can present with some motor deficits. These deficits can initially be seen as clumsiness, weakness or frequent falls. In young infants, this may manifest as motor regression such as refusal to stand or crawl after having learned to walk. Complaints of sensory dysfunction are quite uncommon in children.

Scoliosis can also be a presenting complaint. This is seen in one third of patients (49). The scoliosis curve is not specific in any direction. Children with scoliosis typically have paraspinal pain which is otherwise unusual for intramedullary tumors.

DIAGNOSTIC STUDIES

Magnetic resonance imaging (MRI) is the study of choice to identify an intramedullary spinal cord neoplasm (3, 12). MRI scans should be performed with intravenous contrast agents and in multiple planes. These images demonstrate the solid tumor component, associated cysts, edema, and on occasion the scoliosis. Although MRI does not provide the histological diagnosis, there are

some typical patterns of appearance for intramedullary tumors. Astrocytomas and gangliogliomas have a heterogeneous enhancement pattern (Figure 1). These tumors are often eccentrically located and produce an asymmetric enlargement of the spinal cord. Ependymomas, on the other hand, tend to enhance brightly and homogeneously with contrast. They are often associated with rostral and caudal cysts. These tumors are centrally located within the spinal cord.

Plain radiographs are mandatory only for children who present with scoliosis. These initial films serve as a baseline for the future management of spinal deformity. In addition, children who undergo an extensive laminotomy or laminectomy should be followed with serial radiographs and we therefore advocate immediate postoperative films.

SURGICAL MANAGEMENT

Surgical Instruments

The traditional method for the removal of an intramedullary neoplasm is now supplemented by special microinstruments. These instruments have become essential for the microsurgical resection of spinal cord tumors.

The Cavitron ultrasonic aspirator (CUSA) uses high frequency sound waves to fragment and then suction tumor from the tip of this device (7, 20). This surgical adjunct now has microtips which allow for intraspinal use (26). This allows for tumor removal with only

minimal manipulation of the adjacent spinal cord tissue.



Fig. 1a

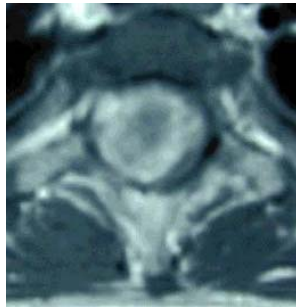


Fig. 1b

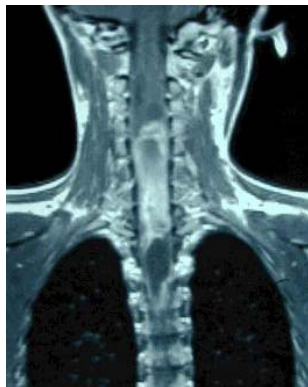


Fig. 1c

Figure 1: MRI scan in a 9 year old boy who presented with neck pain. (A) Sagittal T1-weighted images demonstrates the heterogeneously enhancing cervicothoracic tumor. (B) Axial T1-weighted image demonstrates the eccentric location. (C) Coronal study better demonstrates the eccentric location and extent of the tumor.

The laser is another excellent surgical instrument for intramedullary surgery. We prefer the Nd:YAG Contact Laser System (SLT, Montgomeryville, PA) to other available systems. This system has a handpiece and various contact probes. The contact probes are useful as a scalpel to perform the myelotomy, demarcate the glial-tumor interface and to remove any residual fragments. Unlike other laser systems, there is minimal associated char and smoke generation. This contact laser is effective in preventing small arteriole or capillary bleeding. This laser essentially works as a micro-instrument.

Surgical Technique

The surgical approach for all intramedullary tumors is a laminectomy or osteoplastic laminotomy with the child in the prone position. For cervical or cervicothoracic tumors, the child's head is fixed in a Sugita head holder. The child is positioned on soft gel-rolls to minimize any venous hypertension. We avoid the horseshoe headrest for fear of facial and eye abrasions.

A craniotome is used to cut the laminae bilaterally. In children who have been previously operated upon, the laminae may not be present for repositioning. The opening is made large enough to expose the solid component of the tumor. The rostral and caudal cysts do not need to be fully exposed. This opening is planned with X-ray

control and verified with the intraoperative ultrasound. The ultrasound allows the surgeon to visualize the spinal cord in two dimensions (18). Intramedullary astrocytomas and gangliogliomas have a similar echogenic pattern as the spinal cord. However, the cord will appear expanded in the area of the tumor. In contrast, ependymomas tend to be hyperechogenic and can readily be differentiated from spinal cord. The ultrasound is helpful to identify the associated cyst(s). If the bone removal is not adequate to visualize the full extent of the tumor, the laminotomy is extended prior to opening the dura.

The dura is then opened in the midline. The spinal cord is expanded and may occasionally be rotated. The asymmetric expansion and rotation of the spinal cord may make the identification of the midline difficult. In those cases of an asymmetric tumor and rotated spinal cord, a myelotomy may be performed through the dorsal root entry zone.

The neoplasm is typically located several millimeters underneath the dorsal surface. The contact laser is used to perform the myelotomy with minimal neural injury. Intramedullary tumors have different appearances, such as texture and color, which help the neurosurgeon differentiate the tumor type.

Astrocytomas or gangliogliomas have a gray-yellow appearance. A true plane between tumor and normal spinal cord does

not exist. The surgeon should make no effort to define this "true" interface because it results in hazardous manipulation of normal spinal cord tissue. Ependymomas are typically red-gray in color and these tumors are well demarcated from the surrounding spinal tissue. This interface can be separated with a plated bayonet (19) or the scalpel probe on the contact laser.

Once the tumor is exposed a biopsy is taken for immediate histological examination. This information may, if a malignant glioma or inflammatory process is a possible diagnosis, be crucial in deciding the extent of tumor resection. For malignant gliomas a more conservative approach, to limit any potential motor deficits, is undertaken. The goal is a debulking with preservation of motor function.

Tumor removal for low-grade astrocytomas and gangliogliomas begins after the initial myelotomy is performed. An internal debulking with the CUSA is done to reduce the tumor volume. The resection of astrocytomas is initiated at the midportion rather than the tumor poles. The rostral and caudal poles are the least voluminous and manipulation at these locations may be the most dangerous to the normal cord tissue. Then using the suction or contact laser the tumor is gently removed from the surrounding spinal tissue. These tumors do not have a cleavage plane, although in some areas a plane may exist between tumor and normal spinal tissue. These tumors tend to

displace the motor tracts anteriorly or laterally. The surgeon should be aware of these pathways during tumor resection.

The rostral-caudal length of the tumor does not influence the functional outcome after tumor resection. We have found the removal of small tumors with a wide girth to be more difficult and hazardous than the holocord narrow tumors. This observation corresponds to previous reports that spinal cord atrophy is a poor prognostic factor in terms of neurologic dysfunction (24).

Miscellaneous Tumors

Hemangioblastomas are quite uncommon in children unless associated with von Hippel-Lindau disease. The mean age for an isolated intramedullary tumor is 31.5 years. Hemangioblastomas (9), regardless of size, in the spinal cord are often associated with significant edema and syrinx formation. These lesions should be resected in a circumferential fashion. The tumor surface can be coagulated to allow for the manipulation of the lesion, however this tumor should not and cannot be debulked from within.

Cavernous malformations, similar to hemangioblastomas, are typically located on the dorsal surface of the spinal cord (11). These lesions are typically located in the cervical cord. This vascular malformation is resected in an inside-out fashion, similar to the technique for astrocytomas. These lesions do not usually bleed during

the resection, thus the CUSA or suction cautery can be safely used for their removal. These vascular malformations are quite uncommon in children. When these lesions present in the pediatric population, there is high chance for multiple intracranial lesions (11).

Intramedullary lipomas (31) require a different strategy than glial neoplasms. Although this tumor may appear well demarcated from the adjacent spinal cord tissue, these lesions are densely adherent. Thus total removal is fraught with neurological compromise. The contact laser is used to debulk these tumors. The laser vaporizes the fatty tissue without any surgical trauma to the spinal cord.

Tumor Type	Children
Juvenile Pilocytic Astrocytoma	4 (3%)
Fibrillary Astrocytomas	45 (38%)
Low Grade	32
Anaplastic	10
Glioblastoma	3
Ependymomas	14 (12%)
Myxopapillary	5
Subependymoma	0
Oligodendroglioma	1
Mixed Glioma	3
Ganglioglioma	31 (27%)
Miscellaneous	14 (12%)
Neuronal	10
Hemangioblastoma	3
Others	1
Total	117

Table 2: Histological Diagnoses of intramedullary Neoplasms at a single Institution between 1991-1998

Following intramedullary tumor removal, the dura is closed primarily in a watertight fashion. If an osteoplastic laminotomy was performed the laminae are replaced and secured with a nonabsorbable suture. One tissue layer must be closed in a cerebrospinal fluid tight fashion. The muscle and fascial closure must not be under tension (51). Children who have had previous surgery and radiation therapy are at increased risk for wound dehiscence and cerebrospinal fluid leak.

INTRAOPERATIVE NEUROPHYSIOLOGICAL MONITORING

Intraoperative monitoring with motor evoked potentials (MEPs) allows direct monitoring of the corticospinal tracts (2, 5, 10, 30, 37). This monitoring is predictive of functional motor outcome for intrinsic spinal cord tumor surgery (30). Motor potentials are evoked with transcranial stimulation of the motor cortex. A single electrical impulse results in the direct activation of fast conducting axons by a single stimulus. This potential, called D-wave (40), is recorded by an epidurally-placed electrode just distal to the intramedullary tumor. The D-wave amplitude is a relative measure of the number of functioning fast conducting corticospinal fibers. When this amplitude drops 50%, then 50% of these fibers have been injured (37). The muscle MEPs is elicited with a

short high frequency train of 5-7 electrical stimuli (46). The responses are recorded with needle electrodes from limb muscles with heavy pyramidal innervation, such as the thenar muscles, the tibialis anterior and short toe flexors. These motor potentials follow an on-off pattern; their presence indicates intact motor control and their absence or loss is highly indicative of temporary loss of motor function (30).

The D-wave and muscle MEPs must be interpreted together (28, 29, 30). Loss of muscle MEPs during a tumor resection indicates a temporary disruption of motor function. The incremental change of the D-wave amplitude allows for further interpretation and resection of motor outcome for an intramedullary tumor. As long as the D-wave amplitude remains above the 50%, the patient is likely awakened with a motor deficit. However, the deficit will recover within hours, days or weeks. The intraoperative monitoring using these two electrophysiological parameters has significantly improved the safety of complete resection of intramedullary neoplasms.

Somatosensory evoked potentials (SEPs) is used to assess the functional integrity of the sensory system. The correlation of SEPs with pre and post-operative motor function is poor (28). In many cases of intramedullary tumors, the SEPs disappear after the myelotomy is performed.

OUTCOME FOLLOWING SURGERY

Neurologic Outcome

The most feared complication following intramedullary tumor surgery is paralysis. The incidence of this occurrence is related to the preoperative motor status. Patients who have no or minimal preoperative motor deficits have less than 1% incidence of this postoperative complication. Almost all patients undergoing gross total resection of intramedullary spinal cord tumors experience some postoperative deterioration of neurological function, and about 1/3 have a significant temporary motor deficit. This neurological deterioration typically recovers within a few weeks (4, 21, 28, 30).

In our group of 164 children, there were no deaths due to surgery. The tumors were located throughout the spinal axis. Most children remained at their preoperative grade level or improved. Of the patients who deteriorated 67% deteriorated by only one grade. The children who had a deterioration in grade already had significant preoperative motor deficits (8). At a mean follow-up time of 157 months, more than 60% of the children were functioning at a Grade I or II level of the modified McCormick Scale (34). Thus it is essential that children with known intramedullary tumors be operated upon before the development of severe neurological deficits.

It appears that neurologic improvement after surgery is more likely in patients undergoing total resection than in patients undergoing partial resection (48). In our series, the extent of resection gross total (>95%) or subtotal resection (80-95%) did not significantly affect the long-term outcome. Only patients who underwent a partial resection (<80%) fared significantly worse than those with radically removed tumors. These conclusions for intramedullary astrocytomas have been supported by others (38).

Oncologic Outcome

Despite gross total resections for intramedullary neoplasms, residual microscopic fragments are left in the resection bed. These residual fragments may remain dormant or involute over time. There is abundant evidence that radiation has deleterious effects on the nervous and osseous system (6, 13, 32). The alterations in motor and sensory evoked potentials in patients who have received radiation therapy have been documented (37). Some authors still recommend radiotherapy for intramedullary neoplasms regardless of the histological diagnosis (36, 39). There is no evidence that radiation therapy indeed improves the outcome of low grade astrocytomas or ependymomas (16, 17, 25, 44). We recommend radiation therapy only for malignant tumors, children with documented postoperative rapid tumor regrowth and in those cases where substantial tumor remains

and further surgery is not safely feasible. Unfortunately, despite this adjuvant radiation therapy these neoplasms invariably progress.

SPINAL DEFORMITY

Scoliosis and kyphosis may evolve following laminectomy (42, 49, 50). Many children, approximately two-thirds, developed a spinal deformity following laminectomy for their intramedullary neoplasm. However, only one-third of these children required a stabilization procedure. The only significant factors associated with progressive deformity were cystic tumors, prior radiotherapy, and age less than 7 years. The mean time to an orthopedic stabilization procedure was 3.4 years in our series. Other examined parameters such as location, histology, extent of laminectomy, and imaging characteristics were not significantly associated with subsequent stabilization surgery. Spinal deformity has been reported as a complication of radiation therapy used to treat epidural tumors (27, 43). There was a higher rate of spinal deformity in children who were irradiated at a younger age and doses greater than 20cGy for neuroblastomas (33). In another study of 58 patients younger than 25 years who underwent laminectomies for intraspinal tumors, deformity occurred in 46% of patients younger than 15 years and in only 6% of

patients older than 15. In addition there was a higher incidence for a progressive deformity in the cervical region (45). Several authors have recommended osteoplastic laminotomies for all children to reduce the incidence of spinal deformity (1, 41), however we could not evaluate this parameter in our series. It is essential that patients with intramedullary neoplasms be followed for possible progression of spinal deformity. All patients should undergo routine serial plain radiographs.

CONCLUSION

The surgical resection for intramedullary neoplasms has evolved since the initial reports. With the advent of microsurgical technique, imaging technology and intraoperative electrophysiology, the radical resection of intramedullary neoplasms is a safe and effective treatment. In particular, the electrophysiological monitoring of motor pathways is extremely helpful to achieve a radical resection for these intramedullary tumors. The functional outcome of surgery is best correlated with the preoperative status, thus surgery should be performed early prior to onset of severe motor deficits. The majority of these neoplasms are typically benign in children. Adjuvant radiation and chemotherapy should only be administered to malignant gliomas.

REFERENCES

- 1 Abbott R, Feldstein N, Wisoff JH, Epstein FJ: Osteoplastic laminotomy in children. [Pediatr Neurosurg 18:153-156, 1992.](#)
- 2 Boyd SG, Rothwell JC, Cowan JMA, Webb PJ, Morley T, Asselman P, Marsden CD: A method of monitoring function in corticospinal pathways during scoliosis surgery with a note on motor conduction velocities. [J Neurol Neurosurg Psychiatry 49:251-257, 1986.](#)
- 3 Brotchi J, Dewitte O, Levivier M, Baleriaux D, Vandesteene A, Raftopoulos C, Flament-Durand J, Noterman J: A survey of 65 tumors within the spinal cord: surgical results and the importance of preoperative magnetic resonance imaging. [Neurosurgery 29:651-656; discussion 656-657, 1991.](#)
- 4 Brotchi J, Lefranc F: Current management of spinal cord tumors. **Contemp Neurosurg** 21:1-7, 1999.
- 5 Burke D, Hicks R, Stephen J, Woodforth I, Crawford M: Assessment of corticospinal and somatosensory conduction simultaneously during scoliosis surgery. [Electroenceph Clin Neurophysiol 85:388-396, 1992.](#)
- 6 Clayton PE, Shalet SM: The evolution of spinal growth after irradiation. [Clinical Oncology 3:220-222, 1991.](#)
- 7 Constantini S, Epstein FJ: Ultrasonic dissection in Neurosurgery, in RH Wilkins and SS Rengachary (ed): *Neurosurgery*. New York, McGraw-Hill, 1996, pp 607-608.
- 8 Constantini S, Miller DC, Allen JC, Rorke LB, Freed D, Epstein FJ: Radical excision of intramedullary spinal cord tumors: surgical morbidity and long-term follow-up evaluation in 164 children and young adults. [J Neurosurg \(Spine\) 93:183-193, 2000.](#)
- 9 Cristante L, Herrmann H-D: Surgical management of intramedullary hemangioblastoma of the spinal cord. [Acta Neurochir \(Wien\) 141:333-340, 1999.](#)
- 10 Deletis V: Intraoperative monitoring of the functional integrity of the motor pathways, in O Devinsky, A Beric and M Dogali (ed): *Electrical and Magnetic Stimulation of the Brain and Spinal Cord*. New York, Raven Press, 1993, pp 201-214.
- 11 Deutsch H, Jallo GI, Faktorovich A, Epstein F: Spinal intramedullary cavernoma: clinical presentation and surgical outcome. [J Neurosurg \(Spine\) 93: 65-70, 2000.](#)
- 12 Dillon WP, Norman D, Newton TH, Bolla K, Mark A: Intradural spinal cord lesions: Gd-DTPA-enhanced MR imaging. [Radiology 170:229-237, 1989.](#)
- 13 Duffner PK, Horowitz ME, Krischer JP, Friedman HS, Burger PC, Cohen ME, Sanford RA, Mulhern RK, James HE, Freeman CR, Seidel FG, Kun LE: Postoperative chemotherapy and delayed radiation in children less than three years of age with malignant brain tumors. [N Engl J Med 328:1725-1731, 1993.](#)
- 14 Epstein FJ, Epstein N: [Surgical treatment of spinal cord astrocytomas of childhood. J Neurosurg 57:685-689, 1982.](#)

- 15 Epstein FJ, Farmer J-P: Pediatric spinal cord tumor surgery. [Neurosurg Clin North Am 1:569-590, 1990.](#)
- 16 Epstein FJ, Farmer J-P, Freed D: Adult intramedullary astrocytomas of the spinal cord. [J Neurosurg 77:355-359, 1992.](#)
- 17 Epstein FJ, Farmer J-P, Freed D: Adult intramedullary spinal cord ependymoma: the result of surgery in 38 patients. [J Neurosurg 79:204-209, 1993.](#)
- 18 Epstein FJ, Farmer J-P, Schneider SJ: Intraoperative ultrasonography: an important surgical adjunct for intramedullary tumors. [J Neurosurg 74:729-733, 1991.](#)
- 19 Epstein FJ, Ozek F: The plated bayonet: a new instrument to facilitate surgery for intra-axial neoplasms of the spinal cord and brain stem. Technical note. [J Neurosurg 78:505-507, 1993.](#)
- 20 Flamm ES, Ransohoff JP, Wuchinich D, Broadwin A: Preliminary experience with ultrasonic aspiration in neurosurgery. [Neurosurgery 2:240-245, 1978.](#)
- 21 Goh KYC, Velasquez L, Epstein FJ: Pediatric intramedullary spinal cord tumors: is surgery alone enough? [Pediatr Neurosurg 27:34-39, 1997.](#)
- 22 Greenwood J: Surgical removal of intramedullary tumors. [J Neurosurg 26:276-282, 1967.](#)
- 23 Guidetti B, Mercuri S, Vagnozzi R: Long-term results of the surgical treatment of 129 intramedullary spinal gliomas. [J Neurosurg 54:323-330, 1981.](#)
- 24 Hoshimaru M, Koyama T, Hashimoto N, Kikuchi H: Results of microsurgical treatment for intramedullary spinal cord ependymomas: analysis of 36 cases. Clinical study. [Neurosurgery 44:264-269, 1999.](#)
- 25 Isaacson SR: Radiation therapy and the management of intramedullary spinal cord tumors. [J Neurooncol 47:231-238, 2000.](#)
- 26 Jallo G: CUSA Excel ultrasonic aspiration system. [Neurosurgery 48:695-697, 2001.](#)
- 27 Katzman H, Waug T, Berdon W: Skeletal changes following irradiation of childhood tumors. [J Bone Joint Surg \(Am\) 51:825-842, 1969.](#)
- 28 Kothbauer K, Deletis V, Epstein FJ: Intraoperative spinal cord monitoring for intramedullary surgery: an essential adjunct. [Pediatr Neurosurg 26:247-254, 1997.](#)
- 29 Kothbauer K, Deletis V, Epstein FJ: Intraoperative neurophysiological monitoring, in A Crockard, R Hayward and JT Hoff (ed): *Neurosurgery The scientific basis of clinical practice*. Oxford, Blackwell Science, 2000, pp 1041-1057.
- 30 Kothbauer KF, Deletis V, Epstein FJ: Motor evoked potential monitoring for intramedullary spinal cord tumor surgery: correlation of clinical and neurophysiological data in a series of 100 consecutive procedures. *Neurosurg Focus* 4:Article 1 (<http://www.neurosurgery.org/focus/may98/4-5-1.html>), 1998.
- 31 Lee M, Rezai AR, Abbott R, Coelho DH, Epstein FJ: Intramedullary spinal cord lipomas. [J Neurosurg 82:394-400, 1995.](#)

- 32 Marcus RB, Million RR: The incidence of myelitis after irradiation of the cervical spinal cord. [International Journal of Radiation Oncology and Biological Physics 19:3-8, 1990.](#)
- 33 Mayfield JK, Riseborough EJ, Jaffe N, Nehme AME: Spinal deformity in children treated for neuroblastoma. [J Bone Joint Surg \(Am\) 63:183-193, 1981.](#)
- 34 McCormick PC, Torres R, Post KD, Stein BM: Intramedullary ependymoma of the spinal cord. [J Neurosurg 72:523-532, 1990.](#)
- 35 Miller DC: Surgical pathology of intramedullary spinal cord neoplasms. [J Neurooncol 47:189-194, 2000.](#)
- 36 Minehan KJ, Shaw EG, Scheithauer BW, Davis DL, Onofrio BM: Spinal cord astrocytoma: pathological and treatment considerations. [J Neurosurg 83:590-595, 1995.](#)
- 37 Morota N, Deletis V, Constantini S, Kofler M, Cohen H, Epstein FJ: The role of motor evoked potentials during surgery for intramedullary spinal cord tumors. [Neurosurgery 41:1327-1336, 1997.](#)
- 38 Nadkarni TD, Rekate HL: Pediatric intramedullary spinal cord tumors: critical review of the literature. [Child's Nerv Syst 15:17-28, 1999.](#)
- 39 O'Sullivan C, Jenkin RD, Doherty MA, Hoffman HJ, Greenberg ML: Spinal cord tumors in children: long-term results of combined surgical and radiation treatment. [J Neurosurg 81:507-512, 1994.](#)
- 40 Patton HD, Amassian VE: Single-and multiple unit analysis of cortical stage of pyramidal tract activation. [J Neurophysiol 17:345-363, 1954.](#)
- 41 Raimondi AJ, Gutierrez FA, Di Rocco C: Laminotomy and total reconstruction of the posterior spinal arch for spinal canal surgery in childhood. [J Neurosurg 45:555-560, 1976.](#)
- 42 Reimer R, Onofrio BM: Astrocytomas of the spinal cord in children and adolescents. [J Neurosurg 63:669-675, 1985.](#)
- 43 Riseborough EJ, Grabias SL, Burton RI, Jaffe N: Skeletal alterations following irradiation for Wilm's tumor. [J Bone Joint Surg \(Am\) 58:526-536, 1976.](#)
- 44 Stein BM: Intramedullary spinal cord tumors. [Clin Neurosurgery 30:717-741, 1983.](#)
- 45 Tachdjian MO, Matson DD: Orthopedic aspects of intraspinal tumors in infants and children. [J Bone Joint Surg \(Am\) 49:713-720, 1965.](#)
- 46 Taniguchi M, Cedzich C, Schramm J: Modification of cortical stimulation for motor evoked potentials under general anesthesia: Technical description. [Neurosurgery 32:219-226, 1993.](#)
- 47 Wood EH, Berne AS, Taveras JM: The value of radiation therapy in the management of intrinsic tumors of the spinal cord. [Radiology 63:11-24, 1954.](#)
- 48 Xu QW, Bao WM, Mao RL, Yang GY: Aggressive surgery for intramedullary tumor of cervical spinal cord. [Surg Neurol 46:322-328, 1996.](#)

49 Yao K, Kothbauer KF, Yao K, Kothbauer KF, Constantini S, Epstein FJ, Jallo GI: Spinal deformity and intramedullary tumor surgery. *Child's Nerv Syst* 16:530, 2000.

50 Yasuoka S, Peterson HA, MacCarty CS: Incidence of spinal column deformity after multilevel laminectomy in children and adults. [J Neurosurg 57:441-445, 1982.](#)

51 Zide BM, Wisoff JH, Epstein FJ: Closure of extensive and complicated laminectomy wounds. [J Neurosurg 67:59-64, 1987.](#)

Comments

The authors illustrate the formidable challenge that tumours of this type can present. Their rarity and often nebulous clinical picture means that symptoms have often been present for some time before the diagnosis is made. As a consequence the lesions are often extensive. The majority are low-grade tumours, which will respond poorly to adjuvant therapy, and the extent of surgical resection influences outcome. However, astrocytomas, the most common lesions, have no capsule and attempts at gross total resection may risk paralysis. Spinal deformity is a serious risk after resection, particularly in those with persisting neurological deficit or who have undergone radiotherapy.

Intra-operative ultrasound and neurophysiological monitoring have been major advances in surgery of this difficult disease. There is no longer a place for the "occasional" operator in this field. The treatment of this condition should be confined to institutions that have sufficient throughput of cases, not only to allow safe surgery, but to ensure that facilities exist for rehabilitation and

the management of long term complications.

**Robert Macfarlane MD FRCS
Cambridge**

This is a very good paper on the current management of intramedullary neoplasms. However, it is important that children are separated from young adults in when reporting. Most Authors consider children to be under the age of 15 years.

**SM Eljamel MD FRCSI FRCS(SN)
FABI
Dundee**

Authors reply: Thank you for your interest in the paper and your comments. In our previous paper, Constantini et al., *Radical excision of of intramedullary spinal cord tumors: surgical morbidity and long-term follow-up evaluation in 164 children and young adults*, (8) we found no correlation with patient's age and progression-free and overall survival. Thus the age does not correlate with histology, prognosis or outcome. In our institution those below 21 years are admitted to pediatric ward.