Long-term incidence and risk factors for development of spinal deformity following resection of pediatric intramedullary spinal cord tumors

Clinical article

RAHEEL AHMED, M.D., PH.D.,¹ ARNOLD H. MENEZES, M.D.,¹ OLATILEWA O. AWE, M.D.,¹
KELLY B. MAHANEY, M.D.,¹ JAMES C. TORNER, PH.D.,² AND STUART L. WEINSTEIN, M.D.³

Departments of ¹Neurosurgery, ²Epidemiology, and ³Orthopedics and Rehabilitation, University of Iowa Hospitals and Clinics, Iowa City, Iowa

Object. Spinal deformity in pediatric patients with intramedullary spinal cord tumors (IMSCTs) may be either due to neurogenic disability or due to secondary effects of spinal decompression. It is associated with functional decline and impairment in health-related quality-of-life measures. The authors sought to identify the long-term incidence of spinal deformity in individuals who had undergone surgery for IMSCTs as pediatric patients and the risk factors and overall outcomes in this population.

Methods. Treatment records for pediatric patients (age < 21 years) who underwent surgical treatment for histology-proven primary IMSCTs between 1975 and 2010 were reviewed. All patients were evaluated in consultation with the pediatric orthopedics service. Clinical records were reviewed for baseline and follow-up imaging studies, surgical fusion treatment, and long-term skeletal and disease outcomes.

Results. The authors identified 55 patients (30 males and 25 females) who were treated for pediatric IMSCTs between January 1975 and January 2010. The mean duration of follow-up ($\pm$ SEM) was 11.4 ± 1.3 years (median 9.3 years, range 0.2–37.2 years). Preoperative skeletal deformity was diagnosed in 11 (20%) of the 55 patients, and new-onset postoperative deformity was noted in 9 (16%). Conservative management with observation or external bracing was sufficient in 8 (40%) of these 20 cases. Surgical fusion was necessary in 11 (55%). Posterior surgical fusion was sufficient in 6 (55%) of these 11 cases, while combined anterior and posterior fusion was undertaken in 5 (45%). Univariate and multivariate analysis of clinical and surgical treatment variables indicated that preoperative kyphoscoliosis ($p = 0.0032$) and laminectomy/laminoplasty at more than 4 levels ($p = 0.05$) were independently associated with development of spinal deformity that necessitated surgical fusion. Functional scores and 10-year disease survival outcomes were similar between the 2 groups.

Conclusions. Long-term follow-up is essential to monitor for delayed development of spinal deformity, and regular surveillance imaging is recommended for patients with underlying deformity. The authors' extended follow-up highlights the risk factors associated with development of spinal deformity in patients treated for pediatric IMSCTs. Surgical fusion allows patients who develop progressive deformity to achieve long-term functional and survival outcomes comparable to those of patients who do not develop progressive deformity.

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KEY WORDS • intramedullary tumors • laminoplasty • spinal deformity • fusion • oncology • spine

ADVANCEMENTS in neurosurgical techniques and neurophysiological monitoring have enabled improvements in resection of pediatric intramedullary spinal cord tumors (IMSCTs). This has enabled improved tumor control with favorable disease survival and neurological function outcomes. These surgical approaches are complicated, however, by the development of secondary spinal deformity. Spinal deformity associated with the development and treatment of pediatric IMSCt is an impor-

tant determinant of overall disease outcomes. Progression in spinal deformity is directly linked with health-related quality of life measures and functional scores.^{26,31}

There is wide variation in the pediatric incidence of postlaminectomy deformity following resection of intramedullary tumors, ranging from 24% to 100%.^{3,5,7,12,16,27,29,33,35}

The pathogenesis of secondary spinal deformity for these patients is unlike the development of iatrogenic postlaminectomy kyphosis in adults undergoing cervical decompression for cervical myelopathy, given the unique biomechanical properties of the spine in this age group.

Abbreviations used in this paper: IMSCt = intramedullary spinal cord tumor; XRT = radiation therapy.

Neuromuscular dysfunction associated with the underlying intramedullary tumor may contribute to neurogenic spinal deformity at baseline.^{16,33} Ligamentous structures provide predominant biomechanical stability in the face of skeletal immaturity in this age group and may be disrupted by surgical intervention.² Ventral subluxation due to loss of dorsal laminae and musculoligamentous support may also contribute to deformity.¹⁷ The pubertal growth spurt may further aggravate spinal deformity.³³ Postlaminectomy kyphosis in children may develop through increased load bearing on anterior elements leading to wedging of vertebral bodies.

Various measures, including closer surveillance, concurrent fusion in patient subgroups at risk, and osteoplastic laminoplasties, have been advocated to address adverse consequences of secondary deformity.^{4,18,27,28} Studies to date focusing on the comparative roles of spinal laminectomy versus osteoplastic laminoplasty have been limited by short-term follow-up.¹⁸ Studies have documented onset of postoperative deformity at 1.8–7 years following spinal laminectomy.^{22,35} Moreover, onset of deformity following adjunct radiation therapy (XRT) is typically delayed.⁷ Even after attainment of skeletal maturity and final growth, there is continued risk of deformity progression. Given the risk of deformity progression in pediatric patients, often extending into adulthood, long-term assessment of spinal deformity is essential to effectively monitor the delayed development and/or progression of spinal deformity and to characterize disease outcomes. Extended follow-up of our study group, beyond the 1st decade, enables us to address long-term spinal growth patterns past the adolescent growth spurt.

This article is the second of 2 companion articles being published together. In the first article,¹ we reported on the clinical and treatment determinants affecting long-term disease survival and neurological outcomes in the same group of patients reported on in the present paper. In this second article, our goal was therefore to determine the long-term consequences of spinal decompression and resection of IMSTs on spinal alignment and secondary deformity in the pediatric age group.

Methods

This retrospective study was approved by the University of Iowa Human Subjects Office Institutional Review Board.

Pediatric patients (age < 21 years at presentation) with a histopathological diagnosis of IMST were identified through a retrospective search of the neurosurgery inpatient database at the University of Iowa Hospitals and Clinics for the period from January 1975 to January 2010. Patient records were reviewed for: 1) patient demographics, 2) clinical presentation, 3) surgical treatment received, 4) chemotherapy and/or radiation therapy, and 5) final clinical outcome (based on clinical and radiographic evaluation by the Neurosurgery and Pediatric Oncology and Pediatric Orthopedic Services).

Case records were reviewed to obtain demographic, clinical, radiographic, treatment, and follow-up details. Baseline and follow-up clinical and radiographic assess-

ment of skeletal deformity and determination and timing of skeletal fusion when needed were undertaken in consultation with the Pediatric Orthopedics Service. Patients who developed progressive deformity on interval imaging despite external bracing were considered for deformity correction surgery.

Deficit score on presentation was calculated through the presence or absence of motor, sensory, bladder, or bowel deficits, with the presence of each deficit counting for 1 point, such that the scores could range from 0 (no deficits) to 4 (deficits in each category). If either C-7 or T-1 was involved in the laminectomy, the cervicothoracic junction was considered to be involved. If either T-12 or L-1 was involved in the laminectomy, the thoracolumbar junction was considered to be involved.

In treating patients with IMSTs at our institution, an osteoplastic laminoplasty is planned unless preoperative imaging studies indicate significant mass effect that necessitates extensive decompression, often with the aid of a fascial graft, or in cases in which preoperative imaging studies indicate a high probability of incomplete resection given a lack of clearly defined tumor margins.

Statistical Analysis

Progression-free and disease-survival rates were determined with Kaplan-Meier survival estimates and 95% confidence intervals. Statistical associations were assessed by means of log-rank Mantel Cox analysis using GraphPad Prism version 5.04 for Windows and SAS version 9.3. Statistical significance was determined at $\alpha \leq 0.05$.

Results

A total of 55 patients (30 male [55%] and 25 female [45%]) were selected over the study period from 1975 to 2010. Their mean age at presentation was 10 ± 1 years (range 0.5–21 years).

Astrocytomas were the most prevalent histological tumor subtype, present in 53% of the cases ($n = 29$), followed by ependymomas (22%, $n = 12$) and gangliogliomas (13%, $n = 7$). The tumors were predominantly low grade (43 tumors [78%]). High-grade tumors were diagnosed in 22% of the study group (12 patients).

Skeletal Deformity

The incidence of preoperative, baseline skeletal deformity as evident on preoperative imaging was 20% (11 of 55 cases). New-onset postoperative skeletal deformity was noted in 16% of the patients (9 of 55). Nonoperative management with observation or external bracing was sufficient in 40% (8) of the 20 patients with pre- or postoperative skeletal deformity. Surgical fusion was necessary in 55% (11 patients); resolution was noted in 1 patient (5%).

Of the 10 patients with significant kyphosis, 4 (40%) were treated conservatively, while the remaining 6 (60%) underwent surgical fusion procedures. The mean kyphotic angle deformity in the conservatively treated group was $40^\circ \pm 8^\circ$ (range 30° – 64°), as compared with $80^\circ \pm 8^\circ$ (range 47° – 105°) in the surgically treated group ($p =$

Skeletal deformity and pediatric intramedullary tumors

0.007). Of the 18 patients with scoliosis, 5 (28%) were treated conservatively and 13 (72%) were treated surgically. The mean scoliotic deformity measured $27^\circ \pm 7^\circ$ (range 10° – 50°) in the conservatively managed group as compared with $60^\circ \pm 9^\circ$ (range 16° – 110°) in the surgically treated group ($p = 0.04$).

Surgical Treatment

Surgical treatment details are summarized in Table 1. A total of 43 patients (78%) underwent surgical laminectomy, while 12 patients (22%) underwent laminoplasty. The average age of patients in the 2 groups was similar— 8.8 ± 1.8 years in laminoplasty group and 10.3 ± 1.1 years in laminectomy group ($p = 0.5078$). The number of spinal levels involved was also similar: a mean ($\pm$ SEM) of 4.1 ± 0.3 in the laminoplasty group and 4.7 ± 0.5 in the laminectomy group ($p = 0.4841$). Spinal fusion procedures were performed in 9 (21%) of the 43 patients who underwent spinal laminectomy and 2 (17%) of the 12 patients who underwent osteoplastic laminoplasty ($p = 0.7179$; Table 2).

Of the 11 patients who underwent surgical fusion for spinal deformity, 7 (64%) had preoperative kyphoscoliosis. The other 4 patients (37%) developed clinically significant kyphoscoliosis at a mean interval of 26 months (range 2–84 months) postoperatively. Preoperative baseline surgical deformity characteristics included a mean kyphotic deformity of $63^\circ \pm 12^\circ$ in 3 patients (range 41° – 82°) and $49^\circ \pm 9^\circ$ scoliosis in the other 4 patients (range 32° – 70° scoliosis).

Eight of 11 patients (73%) with surgically treated deformity underwent a single fusion procedure, while the remaining 3 patients (27%) underwent 2 fusion procedures. Within the latter group, the underlying spinal deformity had progressed despite the initial fusion procedure. The mean interval between tumor resection and surgical fusion was 4.4 ± 0.9 years in the postoperative period (range 0.7–10 years). Posterior surgical fusion was sufficient in 6 patients (55%), while combined anterior and posterior fusion was undertaken in 5 (45%).

Long-Term Outcomes

The mean follow-up period ($\pm$ SEM) for the study group was 11.4 ± 1.3 years (median 9.3 years, interquartile range 3.8–15.5 years, overall range 0.2–37.2 years). Univariate and multivariate analyses of the clinical and surgical treatment variables indicated that preoperative kyphoscoliosis ($p = 0.0032$) and laminectomy/laminoplasty spanning more than 4 levels ($p = 0.05$) was independently associated with a higher risk of developing spinal deformity that necessitates surgical fusion (Table 2).

The proportion of patients with normal or mild functional limitations (McCormick Grades I–II) was similar in the subgroup that underwent deformity correction surgery (8 [72%] of 11 patients) and those who did not develop surgical deformity (40 [90%] of 44, $p = 0.1056$). Overall survival was not significantly different between the group of patients who underwent deformity correction surgery and those who did not require surgical fusion. The 10-year overall survival rate was 91% (95% CI

TABLE 1: Summary of surgical treatment*

Variable	Value
no. of tumor surgeries	
1	39 (71)
2	11 (20)
3–4	5 (9)
extent of resection	
GTR	21 (38)
STR	34 (62)
LM	43 (78)
mean no. of levels	4.1
range of no. of levels	1–11
LP	12 (22)
mean no. of levels	4.7
range of no. of levels	3–6
spinal level for LP/LM	
cervicomedullary	4 (7)
cervical	16 (29)
cervicothoracic†	5 (9)
thoracic	15 (27)
thoracolumbar‡	13 (24)
lumbar	2 (4)
no. of myelotomy levels	
mean $\pm$ SEM	3.7 ± 0.2
range	1–9
onset of deformity	
preexistent	11 (20)
postop	9 (16)
time from op to onset of postop deformity	
mean $\pm$ SEM	18 ± 9
range	0–84
course of preexistent deformity (n = 11)	
observation/bracing	4 (36)
surgery	7 (64)
course of postop deformity (n = 9)	
observation/bracing	4 (44)
surgery	4 (44)
resolution	1 (12)
management of deformity (n = 20)	
observation/bracing	8 (40)
surgery	11 (55)
resolution	1 (5)
time interval from tumor resection to fusion surgery (yrs)	
mean $\pm$ SEM	4.5 ± 0.9
range	0.7–10
no. of fusion surgeries (n = 11)	
1	8 (72)
>1	3 (28)

(continued)

TABLE 1: Summary of surgical treatment* (continued)

* Values represent numbers of cases (%) unless otherwise indicated. GTR = gross-total resection; LM = laminectomy; LP = laminoplasty; STR = subtotal resection.

† If either C-7 or T-1 was involved in the laminectomy, the cervicothoracic junction was considered to be involved.

‡ If either T-12 or L-1 was involved in the laminectomy, the thoracolumbar junction was considered to be involved.

51%–98%) for patients with surgical deformity as compared with 69% (95% CI 50%–83%) for patients without surgical deformity ($p = 0.4545$) (Fig. 1).

Discussion

Our results highlight the long-term incidence of and risk factors for spinal deformity in patients with pediatric IMSCTs. Development of secondary deformity is associated with functional decline, with neurological deficits developing due to spinal cord compression.³³ In turn, the resultant neurological decline tends to aggravate the skeletal deformity. In this respect, long-term follow-up of pediatric patients with IMSCTs is essential, given the immediate and delayed lifetime risk of progressive skeletal deformity of the developing spine. Extended follow-up of our study cohort, for the first time beyond the 1st decade, enabled us to undertake long-term assessment. Our results indicate that the long-term risk of developing surgical deformity is 20% and is significantly associated with spinal decompression extending beyond 4 spinal segments and with preexisting spinal deformity. Long-term survival and functional outcomes were not adversely affected by the secondary development and surgical treatment of spinal deformity.

Preoperative Deformity

In our study group, the overall prevalence of underlying deformity on presentation was 20%. This is similar to previous reports of preoperative deformity in up to 33% of patients.^{16,33} Our results indicate that preoperative spinal deformity, when present, is most significantly associated with the lifetime risk of developing progressive deformity that necessitates surgical fusion. In our series, the incidence of surgical fusion within the subgroup of patients with underlying deformity on presentation was 64%. Preoperative spinal deformity, when present, has also been previously reported to be an independent risk factor for progressive deformity following surgery.^{12,33}

The pathogenesis of spinal deformity on presentation in patients with IMSCTs is in part due to the underlying neuromuscular dysfunction. This in turn predisposes to development or progression of postoperative spinal deformity.¹¹ Irreversible changes in musculoskeletal loading may also persist despite resection of IMSCTs. Of note, the functional grade on presentation was not significantly different in the two groups in our study group. This indicates that functional grade assessment may not correlate with underlying neurogenic disability that leads to skeletal deformity.

Osteoplastic Laminoplasty and Risk of Spinal Deformity

Osteoplastic laminoplasties have been proposed to reduce the risk of future development of spinal deformity. The postulated protective effects include restoration of the posterior tension band and bony re-fusion leading to structural reestablishment of spinal integrity. Through reattachment of the paraspinal musculature, laminoplasty may also decrease the incidence of postoperative musculoskeletal pain.⁶ In pediatric IMSCT patients, the overall incidence of progressive spinal deformity was reported to be 5% in patients treated with laminoplasty as compared with 30% in patients having undergone a laminectomy.¹⁸ Although both groups had similar functional outcomes at most recent follow-up, this study reported a beneficial role for laminoplasty in averting postoperative spinal deformity.

Nevertheless, data regarding the role of laminoplasty in averting the risk of postoperative deformity are conflicting.

A meta-analysis of postoperative spinal deformity following laminoplasty indicated significant rates of progression in cervical deformity (35%) as well as de novo postoperative kyphosis (10%), indicating limited efficacy of laminoplasties in averting future progression of deformity.²⁴ Other studies with pooled data from spinal tumor resections within all age groups have demonstrated similar rates of deformity between these two comparison groups.¹⁹ Previous studies may have been limited by the short duration of follow-up and may have underestimated the true long-term incidence of deformity in these patients.^{18,19}

In our opinion, the definitive benefits of laminoplasty are yet to be proven. Laminoplasty may possibly reduce the incidence of deformity, without completely eliminating the lifetime risk of postoperative deformity, as evident on our extended follow-up. It may still be recommended since it may promote wound healing and reduce postoperative CSF leaks without incurring any significant additional morbidity.^{19,20,36} Importantly, it provides a safer extradural plane of surgical dissection during exposure in patients who undergo repeat resections.

Role of Skeletal Factors in Postoperative Deformity

Our results indicate that spinal decompression extending beyond 4 spinal segments is also associated with the risk of progressive deformity over long-term follow-up ($p = 0.05$ approaches significance). The risk of postoperative deformity or instability has been shown to correlate with the total span of the laminectomy defect, preservation of facet joint integrity,^{12,22,37} and preoperative loss of cervical lordosis.¹¹ Additional risk factors identified in other studies include presence of a tumor-associated syrinx and preoperative spinal deformity.³³ The latter, in particular, may be associated with irreversible changes in musculoskeletal loading that may persist despite resection of intramedullary tumor. Yet other studies have shown that the extent of bony removal, including facetectomy, does not correlate with the incidence of postoperative spinal deformity in children.³⁴

The risk of deformity in the pediatric age group is

Skeletal deformity and pediatric intramedullary tumors

TABLE 2: Factors determining operative deformity*

Characteristic	Surgical Deformity		Total	Univariate Analysis p Value	Multivariate Analysis p Value
	Present	Absent			
patient age at presentation					
<5 yrs	5 (25)	15 (75)	20 (36)	0.2111	
6–13 yrs	5 (28)	13 (72)	18 (33)		
14–21 yrs	1 (6)	16 (94)	17 (31)		
tumor pathology					
histological type					
astrocytoma	8 (26)	23 (74)	31 (56)	0.2211	
other†	3 (13)	21 (87)	24 (44)		
grade					
high grade	1 (8)	11 (92)	12 (22)	0.2532	
low grade	10 (24)	33 (77)	43 (78)		
clinical presentation					
time from Sx onset to presentation in mos					
mean	7.7	7.2	7.3	0.8949	
range	0.7–14.7	3.9–10.5	4.4–10.2		
motor deficit					
present	7 (21)	26 (79)	33 (60)	0.8473	
absent	4 (18)	18 (82)	22 (40)		
sensory deficit					
present	9 (21)	33 (79)	42 (76)	0.7179	
absent	2 (15)	11 (85)	13 (24)		
bladder deficit					
present	3 (14)	18 (86)	21 (38)	0.3758	
absent	8 (24)	26 (76)	34 (62)		
bowel deficit					
present	3 (30)	7 (70)	10 (18)	0.4022	
absent	8 (18)	37 (82)	45 (82)		
deficit score					
≤2	8 (21)	31 (79)	39 (71)	0.8820	
>2	3 (19)	13 (81)	16 (29)		
functional grade on presentation					
I–II	6 (16)	31 (84)	37 (67)	0.3145	
III–V	5 (28)	13 (72)	18 (33)		
preop kyphoscoliosis					
present	7 (64)	4 (36)	11 (20)	<0.0001	0.0032
absent	4 (9)	40 (91)	44 (80)		
tumor location					
cervical	6 (22)	21 (78)	27 (49)	0.5024	
thoracic	5 (22)	18 (78)	23 (42)		
lumbar	0 (0)	5 (100)	5 (9)		
syrinx					
yes	4 (25)	12 (75)	16 (36)	0.2127	
no	3 (11)	25 (89)	28 (64)		
margins					
circumscribed	1 (7)	14 (93)	15 (34)	0.2280	
diffuse	6 (21)	23 (79)	29 (66)		

(continued)

TABLE 2: Factors determining operative deformity* (continued)

Characteristic	Surgical Deformity		Total	Univariate Analysis p Value	Multivariate Analysis p Value
	Present	Absent			
clinical presentation (<i>continued</i>)					
enhancement					
yes	7 (18)	33 (88)	40 (91)	0.3616	
no	0 (0)	4 (100)	4 (9)		
no. of levels spanned					
1–3	2 (9)	20 (91)	22 (40)	0.2473	
4–6	6 (26)	17 (74)	23 (42)		
>7	3 (30)	7 (70)	10 (18)		
surgical treatment					
resection					
GTR	3 (14)	18 (86)	21 (38)	0.4051	
STR	8 (24)	26 (76)	34 (62)		
shunt placement					
yes	4 (33)	8 (67)	12 (22)	0.1916	
no	7 (16)	36 (84)	43 (78)		
no. of tumor surgeries					
1	7 (18)	32 (82)	39 (71)	0.5527	
>1	4 (25)	12 (75)	16 (29)		
spinal decompression					
LM	9 (21)	33 (79)	42 (78)	0.7179	
LP	2 (17)	10 (83)	12 (22)		
no. of levels w/ LM or LP					
≤4	2 (7)	28 (93)	30 (56)	0.0052	0.05
>4	9 (38)	15 (63)	24 (44)		
mean no. of levels w/ LM/LP (± SEM)	6 ± 0.8	4 ± 0.4	4.6 ± 0.4	0.0251	
spinal level of LM/LP					
cervical	6 (24)	19 (76)	25 (45)	0.6602	
thoracic	5 (18)	23 (82)	28 (51)		
lumbar	0 (0)	2 (100)	2 (4)		
LM/LP involves spinal transition level‡					
yes	4 (18)	18 (82)	22 (40)	0.7831	
no	7 (21)	26 (79)	33 (60)		
mean no. of surgical myelotomy levels (± SEM)	4 ± 0.5	3.6 ± 0.3	3.7 ± 0.3	0.3486	
spinal levels of myelotomy					
cervical	6 (25)	18 (75)	24 (44)	0.4936	
thoracic	5 (19)	22 (81)	27 (49)		
lumbar	0 (0)	4 (100)	4 (7)		
onset of deformity					
preop	3 (50)	3 (50)	6 (11)	<0.0001	
postop	8 (62)	5 (38)	13 (24)		
none	0 (0)	36 (100)	36 (65)		
clinical outcomes					
course at last follow-up					
stable	1 (13)	7 (87)	8 (15)	0.7602	
decline	3 (18)	14 (82)	17 (31)		
improvement	7 (23)	23 (77)	30 (55)		

(continued)

TABLE 2: Factors determining operative deformity* (*continued*)

Characteristic	Surgical Deformity		Total	Univariate Analysis p Value	Multivariate Analysis p Value
	Present	Absent			
clinical outcomes (<i>continued</i>)					
functional status at last follow-up					
I–II	8 (17)	40 (83)	48 (87)	0.1056	
III–V	3 (43)	4 (57)	7 (13)		
adjuvant therapy					
type of therapy					
chemo w/ or w/o XRT	4 (17)	19 (83)	23 (42)	0.6818	
none	7 (22)	25 (78)	32 (58)		
indication for chemo					
not used	8 (21)	31 (79)	39 (72)	0.9666	
adjuvant or recurrence therapy	3 (20)	12 (80)	15 (28)		
indication for XRT					
not used	3 (11)	24 (89)	27 (49)	0.1056	
adjuvant or recurrence therapy	8 (29)	20 (71)	28 (51)		

* Values represent numbers of cases (%) unless otherwise indicated. chemo = chemotherapy; Sx = symptom.

† Includes ependymomas, gangliogliomas, and other histopathology subtypes.

‡ Involvement of cervicothoracic or thoracolumbar region.

higher for patients with cervical tumors,³⁵ although it can occur after surgery at any spinal level.⁷ Other studies have shown higher rates of secondary deformity with thoracolumbar tumor locations.^{33,36} The risk of secondary deformity is especially high in the pediatric age group.^{7,12,16,22,34,35} The underlying ligamentous laxity, reduced paraspinal musculature support, horizontal orientation of the facet joints, and disproportionate load bearing due to the relative cranium to body mass ratio have all been known to underlie the susceptibility of the pediatric spine.² Age less than 13 years may be an independent risk factor for secondary deformity^{33,35} and may additionally be independent of the extent of bony resection or history of irradiation.³⁴ Heterogeneity in the pathogenic role of these factors may be due to differences in patient popula-

tion and variation in tumor grade, preoperative neurological disability, and adjunct treatment factors.

Role of Concurrent Fusion

Laminoplasty with concurrent posterolateral fusion has been reported to decrease the incidence of postoperative deformity.^{4,27} However, concurrent posterior fusion has been documented to decrease the development of postlaminectomy kyphosis only within the subset of pediatric patients with no evidence of tumor progression or recurrence.²⁸ The protective effects of prophylactic fusion were lost when deformity rates were pooled from the entire study group.⁴

There are limitations of the prophylactic fusion approach in pediatric IMSCT patients. Disease recurrence leading to neuromuscular scoliosis can negate any beneficial effects of the posterior structural fusion. This approach is hence limited, given the natural history of pediatric IMSCTs that are complicated by recurrence.^{10,13,32} Additionally, the presence of posterior fusion following laminectomy is associated with the risk of “crank-shaft” deformity in skeletally immature children, due to continued growth of the anterior spine column and a high rate of pseudarthrosis.^{15,25} Conversely, anterior augmentation of spinal fusion can halt deformity progression, but it is associated with significant operative morbidity that is difficult to justify on a prophylactic basis in pediatric IMSCT patients.^{14,23}

Effects of XRT

Radiation therapy is independently associated with a higher risk for posttreatment deformity, even in patients with no history of spinal decompression or laminectomy.^{8,30} Radiation-induced damage to the growth plate leads to decreased growth potential. Irregular growth patterns can

Fig. 1. Comparison of disease survival between patients with (dashed line) and without (solid line) surgical deformity, as determined with the Kaplan-Meier method. Vertical hatches indicate censored events. The p value was nonsignificant.

result from asymmetrical involvement of the growth plate, with resultant progression in spinal deformity.^{7,35} Onset of postradiation spinal deformity is later when compared with postlaminectomy cases (mean of 77 months post-XRT vs 5–15 months following surgical laminectomy).⁷ The risk of secondary deformity is probably compounded in patients who undergo laminectomy and XRT.²¹

External Orthosis

The efficacy of external orthosis in averting the subsequent development of spinal deformity is unclear. Postoperative bracing may help delay the onset of progression without reducing the overall risk of developing postoperative deformity.²¹ Postoperative bracing may be necessary following laminoplasty to promote anatomical alignment of the laminae block.²¹ It should also be used following fusion procedures until bony fusion occurs.⁷ In our series, 10 of 13 patients with postoperative braces needed fusion surgery. We recommend bracing in all patients who undergo surgical fusion, in patients with mild scoliotic defects, and as an adjunctive treatment to delay surgical fusion, when feasible, in patients before their adolescent growth spurt.

Conclusions

Late neurological deterioration following development of postlaminectomy kyphosis has been well described¹⁷ and is attributed, in part, to spinal compression from anterior draping over the kyphotic deformity. Correction of deformity aids in cosmesis and improves psychological development in children.³¹ It is also associated with significant improvement in pulmonary function.²⁶ Treatment options include anterior approaches for kyphotic deformities, which are typically supplanted by posterior fusion for more complex deformity curves.⁹ Long-term follow-up is essential to monitor for delayed development of spinal deformity, and surveillance imaging is needed for patients with underlying known deformity. Our extended follow-up highlights the risk factors associated with development of spinal deformity in pediatric IMSCT patients. Surgical fusion allows patients who develop progressive deformity to have long-term functional and survival outcomes that are comparable to those of patients who do not develop progressive deformity.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Menezes, Ahmed. Acquisition of data: Menezes, Ahmed, Awe. Analysis and interpretation of data: all authors. Drafting the article: Menezes, Ahmed, Awe, Mahaney, Torner. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Ahmed. Statistical analysis: Ahmed, Mahaney, Torner. Study supervision: Menezes.

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Skeletal deformity and pediatric intramedullary tumors

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Address correspondence to: Arnold H. Menezes, M.D., Department of Neurosurgery, University of Iowa Hospitals and Clinics, 200 Hawkins Dr., 1824 JPP, Iowa City, IA 52242. email: arnold-menezes@uiowa.edu.