

# Spinal Cord Compromise: An Important but Underdiagnosed Condition in People with Mental Retardation

ROD CURTIS, MD<sup>a,b,c</sup>  
PER FREITAG, MD, PhD<sup>d</sup>  
JAMES J. LA GUARDIA, MD<sup>e</sup>  
STEVEN THORNTON, MD<sup>c</sup>  
SANDRA VICARI, PhD<sup>a</sup>  
STEPHEN MARKWELL, MA<sup>f</sup>

The Surgeon General's 2002 report *Closing the Gap: A National Blueprint to Improve the Health of Persons with Mental Retardation* clearly delineates the need for enhancements in medical education, medical research, and direct care for people with neurodevelopmental disabilities.<sup>1</sup> For clinicians actively involved in this challenging field, the report and its recommendations have been welcomed, along with expectations of greater resources and improved clinical outcomes for all people with neurodevelopmental disabilities.

It is well appreciated that, as a group, people with developmental disabilities experience poorer health than the general population.<sup>1</sup> Issues such as complex psychiatric conditions, seizure disorders, aspiration pneumonia, bowel obstruction, congenital anomalies, and injuries sustained from repeated traumas—including those resulting from physical abuse, self-injurious behaviors, and accidents—contribute to significant morbidity, early mortality, and poor quality of life. In addition, as addressed in the Surgeon General's report, modifiable conditions that commonly lead to preventable disorders are not actively addressed in this population; these include poor fitness, excessive sun exposure, smoking, sexually transmitted diseases, substance abuse, and domestic and sexual abuse.<sup>1</sup> Health care screening for serious and possibly treatable conditions, such as malignancies of the breast, prostate, colon, and skin, as well as routine screening for hypertension, diabetes, and hyperlipidemia represent yet another area that must be addressed and accommodated for people with neurodevelopmental disabilities.

Relying on the Surgeon General's report as a guide, a group of clinicians affiliated with a large intermediate care facility began exploring the prevalence and etiology of several conditions that result in serious complications and death for the developmentally disabled population. Systems have been developed to identify early predictors of these conditions. At our center we have developed medical protocols that have led to prompt diagnosis and treatment, which have resulted in reductions in serious morbidity and mortality from these conditions. This process was facilitated by collaborative relationships among various medical specialties, including neurology, psychiatry, orthopedics, and primary care.

We initially focused on bowel obstruction, pneumonia, and osteoporosis. In evaluating the residents of the center for osteoporosis, we identified a large number who presented with progressive limitations of functional abilities, musculoskeletal pathology

<sup>a</sup>Division of Developmental Disabilities, Department of Psychiatry, Southern Illinois University School of Medicine, Springfield, IL

<sup>b</sup>Department of Medicine, Southern Illinois University School of Medicine, Springfield, IL

<sup>c</sup>Jacksonville Developmental Center, Jacksonville, IL

<sup>d</sup>Division of Orthopedic Surgery, Department of Surgery, Southern Illinois University School of Medicine, Springfield, IL

<sup>e</sup>Department of Neurology, Southern Illinois University School of Medicine, Springfield, IL

<sup>f</sup>Department of Statistics and Research, Southern Illinois University School of Medicine, Springfield, IL

Address correspondence to Rod Curtis, MD, Dept. of Psychiatry, Southern Illinois Univ. School of Medicine, 901 W. Jefferson, Springfield, IL 62702; tel. 217-545-3935.

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and gait abnormalities. We began a retrospective review of the medical files of these residents. Upon closer evaluation, many individuals that were initially reported to have cerebral palsy or other congenital anomalies were discovered not to have these conditions and no clear explanation for their function decline.

Clinicians at the center began an extensive work-up of these individuals to identify the etiology of their functional decline, using a multimodal approach tailored for each person. We quickly realized that degenerative spine disease and prolapsed disc disease leading to spinal cord compromise (myelopathy) was likely to be at least a contributing factor to the observed decline in functional ability.

In people with neurodevelopmental disabilities, progressive functional decline—evident by gait abnormalities, loss of motor abilities, sensory impairment, urinary and fecal incontinence, and a decline in the ability to participate in activities of daily living—is usually attributed to various dementing processes. Alzheimer's disease, vascular and subcortical dementias, and toxic effects of prolonged use of certain medications (such as psychotropic and anticonvulsant drugs) can obviously contribute to loss of function. In our experience, however, spinal cord compromise, regardless of its etiology, is a serious and underdiagnosed condition in individuals with neurodevelopmental disabilities and may be an important contributor to functional decline and early death. With the exception of upper cervical spine subluxation (instability of the upper vertebrae) in people with Down's syndrome<sup>2,3</sup> and cervical spinal stenosis in people with cerebral palsy,<sup>4</sup> little has been reported on spinal cord compromise, such as degenerative spine disease, prolapsed disc disease, and vertebral fractures, in people with neurodevelopmental disabilities.



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Indicators of spinal cord compromise range from mild discomfort, numbness, and tingling to more severe symptoms such as pain, urinary and fecal incontinence, loss of motor and sensory function, and total paralysis. Spinal cord compromise may occur from an acute trauma to the spine, or develop insidiously from chronic degenerative disease secondary to a host of conditions (including osteoporosis involving the vertebrae,<sup>5,6</sup> arthritis, and/or prolapsed disc disease<sup>7-9</sup>). Finally, complications from various congenital anomalies of the spine, including scoliosis and cerebral palsy may also be causative factors.<sup>10</sup> The medical literature suggests that people with neurodevelopmental disabilities are at a greater risk for osteoporosis<sup>11-16</sup> and sustain a higher rate of traumatic fractures<sup>11,12,17-21</sup> than the general population. Both conditions may be important contributors to the development of spinal cord compromise.

Unfortunately, many people with neurodevelopmental disabilities are not able to accurately communicate their symptoms, and observable signs of progressive pathology may be so insidious that direct care staff and family members may not identify subtle changes over time. Individuals with moderate to profound mental retardation often manifest discomfort by behavior exacerbations that may be treated psychiatrically and without a thorough review for possible medical causes. In many cases, the initial signs and symptoms of degenerative spine disease are addressed with behavior programming and/or the administration of psychotropic medications.

### SPINAL CORD INJURY IN AN INTERMEDIATE CARE SETTING

Recognizing the need to better identify individuals with spinal cord compromise, we began clinically evaluating center residents with progressive neuromuscular changes, as indicated by declines in functional ability. A request to direct care staff to identify individuals with observable changes in functional status—including changes in gait, changes in performance of activities of daily living, bladder and bowel incontinence, and worsening spasticity or contracture—resulted in 48 initial referrals for evaluation. Physical examinations of these residents, including thorough neurological examinations when possible and reviews of their clinical records, were supplemented by additional diagnostic studies as indicated. Depending on the patient's presentation and evidence of other causes for their decline, x-ray, CT, or MRI studies were obtained of one or more areas of the spine to determine the presence of degenerative spine disease, spinal stenosis, disc disease, and/or spinal cord compression with myelopathy.

We report here on pooled data for the first 48 individuals triaged at the center, including information on age; sex; level of mental retardation; history of a psychiatric diagnosis, seizure disorder, and/or cerebral palsy; current use of antiepileptic medications; and the results of bone density testing and imaging studies of the spine. Pooled data were stripped of identifying information. The center's executive committee and the Springfield (IL) Committee for Research in Human Subjects gave approval for the chart review.

The mean age of the 48 residents was 49 years, with a range of 21 to 75 years; 37 were male (77%) and 11 were

female (23%). Level of mental retardation, according to clinical records, ranged from mild to profound (mild 14%, moderate 21%, severe 19%, and profound 46%). Psychiatric diagnoses were recorded for 26 (54%) individuals, seizure disorders were diagnosed in 18 (38%) individuals, and 10 (21%) individuals had diagnoses of cerebral palsy.

Of the 48 individuals reviewed, 44 had previously been screened for osteoporosis by heel densitometry. A total of 19 (43%) individuals had a diagnosis of osteoporosis (bone density T-score  $\leq -2.5$ ), another 12 (27%) were known to have osteopenia (T-score  $> -2.5 \leq -1.1$ ), while only 13 (30%) individuals had normal bone density (T-score  $> -1.1$ ).

One or more antiepileptic drugs had been prescribed for about half of the individuals screened (25, or 52%): 16 were on valproic acid (Depakote), 11 were on carbamazepine (Tegretol), four were on zonisamide (Zonegran), two were on levetiracetam (Keppra), and two were on phenytoin (Dilantin).

An x-ray, MRI, or CT of one or more areas of the spine (cervical, thoracic, and/or lumbar region) was clinically indicated for 44 of the 48 individuals identified as having progressive functional changes; 19 had one or more MRIs, three had one or more CTs, and 39 individuals underwent one or more x-rays of the spine. MRIs are helpful in identifying spinal stenosis, prolapsed disc disease, and spinal cord compression; CTs help with diagnosing fractures and spinal stenosis; while x-rays help identify spinal subluxation, degenerative spine disease, and vertebral fractures.

The evaluation of these 48 intellectually disabled adults residing at a developmental center suggests a correlation between functional decline and positive findings on imaging studies of the spine. Of the 44 individuals who underwent imaging studies, 14 (32%) were without pathology, nine (20%) demonstrated mild pathology, and 21 (48%) were identified as having moderate to severe degenerative disease. Seven (16%) of the 44 were determined to have severe spinal cord compression and myelopathy. Thus, evidence of spinal cord compromise was found for 30 of the 44 individuals who underwent imaging studies. Of significance is the number of individuals with spinal cord compression (myelopathy), which in most cases would benefit from surgical intervention.

The Table shows the characteristics of individuals identified as having spinal stenosis or other non-specific degenerative spine disease; prolapsed intervertebral disc disease; or spinal cord compression. A statistically significant relationship was found between anticonvulsant medication and each of the three conditions. As expected, there were statistically significant relationships between bone density T-scores and both spinal stenosis/degenerative disease and prolapsed disc. The data also show a statistically significant relationship between age and spinal cord compression. The distribution of severity of mental impairment differed for those with and without prolapsed disc.

These findings suggest that the use of some antiepileptic drugs, which are frequently prescribed in this population not only for seizure disorders but also for behavioral disorders, may be an important contributor to the disease process. An important consideration regarding antiepileptic drugs and their potential correlation with spine disease is

that many individuals may have recently been switched from older agents to newer medications, and the effect reported here may be residual from the previous drugs. This, however, needs further study.

The relationship between low bone density and both prolapsed disc and spinal cord compression suggests that it may be advantageous to closely monitor people with neurodevelopmental disabilities for early signs of osteoporosis by measuring bone density early on. Larger prospective studies will be necessary to identify the significance of this correlation.

In the general population, the standard of care for mild to moderate spine disease includes non-surgical approaches such as nonsteroidal anti-inflammatory medications, specialized physical therapy and exercises, epidural steroid injections, or a combination of all of these options. In patients with objective findings of myelopathy, where a fall or minor trauma could result in paralysis, decompression surgery and stabilization of the spine is indicated. Normally, clinicians rely on the patient voicing his or her concerns of pain and other symptoms in deciding which treatment to recommend. However, many people with neurodevelopmental disabilities are not able to accurately communicate their symptoms. For this reason, the physician must decide on treatment options based upon functional deterioration and behavior changes, as well as on findings from a comprehensive physical examination and results of imaging studies.

Another quandary with regard to treatment of individuals with neurodevelopmental disabilities is that many individuals have developed secondary complications, including paralysis, contractures, and spasticity many years earlier, and surgical intervention may initially appear futile. However, if the disease process continues or the individual sustains a trauma to the spine from a minor fall, an assault, or self-injurious behavior, there is little room within the spinal canal to accommodate additional stenosis or edema. This would potentially cause total compression of the spinal cord resulting in respiratory insufficiency, total paralysis, or death. Survivors of such an event usually require long-term artificial ventilation and around-the-clock care. Early surgical intervention helps prevent progressive deterioration of function, and helps to protect the individual from complications secondary to minor trauma. In our experience, the four individuals who underwent early surgical intervention for myelopathy had exceptional recovery and were back to their daily activities within one week or less.

Our experience suggests that individuals with identified spinal pathology improve behaviorally following surgical and nonsurgical intervention or appropriate conservative measures. Several individuals who were placed on anti-inflammatory medications were reported to have significant reduction or total abatement of their undesirable behaviors. Individuals who underwent early surgical intervention were also noted to improve behaviorally. One such individual was known to have had significant self-injurious and aggressive behaviors for many years. This individual would bite his hand repeatedly or bite anyone who would attempt to assist him. It was later realized that his actions were a defense mechanism to protect himself from excruciating pain caused by his condition and exacerbated by subtle movements. Following surgical intervention, the behaviors have mostly resolved.

**Table. Attributes contributing to risk of degenerative spine disease**

<i>Bone density (T-scores) and age means and standard deviations</i>										
		<i>Spinal stenosis/degenerative disease (Number of individuals)</i>			<i>Prolapsed disc (Number of individuals)</i>			<i>Spinal cord compression (Number of individuals)</i>		
		<i>Yes</i>	<i>No</i>	<i>(p-value)</i>	<i>Yes</i>	<i>No</i>	<i>(p-value)</i>	<i>Yes</i>	<i>No</i>	<i>(p-value)</i>
T-scores	Mean	-2.65	-1.57	(0.034)	-2.89	-1.82	(0.042)	-2.66	-2.10	
	SD	1.56	1.64		1.41	1.70		1.12	1.75	
Age	Mean	50.89	45.62		52.35	46.52		59.29	46.76	(0.012)
	SD	11.89	12.61		10.26	13.06		9.76	11.90	
<i>Risk variable</i>										
		<i>Spinal stenosis/degenerative disease (Number of individuals)</i>			<i>Prolapsed disc (Number of individuals)</i>			<i>Spinal cord compression (Number of individuals)</i>		
		<i>Yes</i>	<i>No</i>	<i>Odds ratio (p-value)</i>	<i>Yes</i>	<i>No</i>	<i>Odds ratio (p-value)</i>	<i>Yes</i>	<i>No</i>	<i>Odds ratio (p-value)</i>
Sex	Male	21	16	1.09	7	19	0.44	6	31	1.94
	Female	6	5		10	12		1	10	
Psychiatric illness	Yes	12	14	0.40	7	19	0.44	3	23	0.59
	No	15	7		10	12		4	18	
Seizures	Yes	10	8	0.88	6	12	0.82	2	16	0.60
	No	17	12		11	18		5	24	
Cerebral palsy	Yes	6	4	1.14	4	6	1.23	2	8	1.60
	No	21	16		13	24		5	32	
Severe MR	Yes	9	7	0.93	3	13	0.28	2	14	0.74
	No	18	13		14	17		5	26	
Antiepileptic drug therapy	Yes	21	8	5.69	14	15	4.98	7	22	7.00 <sup>a</sup>
	No	6	13	(0.01)	3	16	(0.03)	0	19	(0.03)

NOTE: Continuity adjusted chi-square or Fisher's exact tests, as appropriate, were used to determine the statistical significance of the odds ratios describing the relationship of the risk variable and each of three forms of degenerative spine disease.

<sup>a</sup>The true odds ratio is not calculatable because there are no cases of spinal cord compression among individuals who were not being treated with antiepileptic drug therapy. An estimated odds ratio of 7.00 results from adding one to each cell.

SD = standard deviation

## CONCLUSION

Our clinical findings suggest that spinal cord compromise secondary to degenerative spine disease, disc prolapse, and/or trauma may be an important contributor to life-altering and potentially life-threatening functional decline in people with neurodevelopmental disabilities. Large, well-controlled prospective studies are needed to determine the prevalence of spinal cord pathology and the etiology of functional decline in this population. Importantly, our findings imply the need for prompt evaluation of all intellectually impaired individuals with either new onset or progressive functional decline and the inclusion of spinal cord pathology in the differential diagnosis.

Our findings support former Surgeon General Satcher's recommendation for improvements in clinical services for this population through enhanced clinical research, medical education, and care delivery systems. Only by addressing clinical issues systematically through a multidisciplinary approach, involving primary care physicians and sub-specialists, will we be able to address the many known and the yet unknown medical conditions that result in poor quality of life, early mortality, and significant expenditure of clinical resources.

Long term solutions leading to improvements in health care delivery for this important population will require medical schools to develop efficacious educational curricula designed to assist medical students and residents in gaining a better understanding of the unique health care issues of people with neurodevelopmental disabilities.

Spinal cord compromise is just one of many under-recognized and under-treated conditions in people with neurodevelopmental disabilities and related disorders. Such conditions may progress slowly for years, manifesting behaviorally and being treated psychiatrically. Behavior issues are known to be a major factor in preventing individuals from community integration and in the consumption of significant resources. If medical conditions were recognized and effectively treated promptly, there would be significant improvements with regard to behavior issues and reductions in the utilization of important resource that could be applied in different domains in the support of people with neurodevelopmental disabilities. More important, efficacious treatment would reduce pain and suffering and enable people to reach their maximum potential as individuals and in society.

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