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Few patients with neurodegenerative disorders require spinal surgery

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Abstract

Background: Few patients with neurodegenerative disorders (ND) (e.g., Multiple Sclerosis (MS), Amyotrophic Lateral Sclerosis (ALS), and Postpolio Syndrome (PPS)) require spinal surgery. Typically, their neurological symptoms and signs reflect their underlying neurologic disorders rather than structural spinal pathology reported on magnetic resonance images (MR) or computed tomographic scans (CT).

Methods: The first author, a neurosurgeon, reviewed 437 spinal consultations performed over a 20-month period. Of 254 patients seen in first opinion (e.g., had not been seen by a spinal surgeon), 9 had MS, while 2 had ALS. Of 183 patients seen in second opinion (e.g., prior spinal surgeons recommended surgery), 4 had MS, 2 had ALS, and 1 had PPS. We performed this study to establish how often patients with ND, seen in first or second opinion, require spinal surgery. We focused on whether second opinions from spinal surgeons would limit the number of operations offered to these patients.

Results: Two of 11 patients with ND seen in first opinion required surgery. The first patient required a C5-7 laminectomy/C2-T2 fusion, followed by a L2-S1 laminectomy/L5S1 fusion. The second patient required a L2-L3 laminectomy/diskectomy/fusion. However, none of the seven patients seen in second opinion, who were previously told by outside surgeons they needed spinal surgery, required operations.

Conclusions: Few patients with neurodegenerative syndromes (MS, ALS, PPS) and reported "significant" spondyloitic spinal disease interpreted on MR/CT studies required surgery. Great caution should be exercised in offering patients with ND spinal surgery, and second opinions should be encouraged to limit "unnecessary" procedures.

Key Words: Amyotrophic lateral sclerosis, first opinions, multiple sclerosis, post-polio syndrome, second opinions: Limiting spinal surgery, spinal surgery

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INTRODUCTION

Patients with neurodegenerative disorders (ND) (e.g., Multiple Sclerosis (MS), Amyotrophic Lateral Sclerosis (ALS), and Postpolio Syndrome (PPS)) present with progressive neurological deterioration typically attributed to their underlying diseases rather than surgical spinal pathology. Nevertheless, when they do deteriorate,

they often undergo magnetic resonance imaging (MR) and/or computed tomographic (CT) examinations that often show some degree of spondylotic pathology. As these degenerative findings on MR and/or CT are often over-interpreted by some radiologists/neuroradiologists and spinal surgeons, they may inadvertently be advised to undergo spinal operations they do not need. In these cases, seeking a second opinion from another spinal surgeon may avoid some of these operations.

MATERIALS AND METHODS

We evaluated how often patients with underlying ND (e.g., MS, ALS, or PPS) come in for first (no prior spinal surgical evaluation) or second opinions (where a previous spinal surgeon recommended surgery) regarding the need for spinal surgery. Our main premise was that although patients with MS, ALS, or PPS may exhibit degenerative spondylotic features on MR or CT studies, their deficits are largely attributable to their underlying ND disorders.

Over a 20-month period, prospectively, 437 patients with cervical or lumbar complaints were evaluated. Spondylotic changes (stenosis, spondylosis, disc disease, instability) seen on MR and/or CT studies were correlated with patients' clinical symptoms and signs. Out of 254 patients seen in first opinion, where no prior surgeon had evaluated the patient, we found that 11 had ND syndromes; 9 had MS, and 2 had ALS. Out of 183 patients seen in second opinion, where other spine surgeons previously told patients that they needed surgery, 7 patients had ND disorders: 4 had MS, 2 had ALS, and 1 had PPS.

We asked two questions. First, how often did first opinion patients with ND require spinal surgery? Second, how often did patients who were previously told they needed spinal surgery by other spine surgeons actually require spinal surgery?,

Determining whether MR studies documented "surgical spinal pathology" not only required stringent reassessment of studies that were previously over-interpreted by prior radiologists/neuroradiologists or other surgeons, but also required obtaining better quality or newer studies (MR, CT) and/or consultation with a specialized spinal neuroradiologist.

RESULTS

First opinion results for patients with neurodegenerative disease

Of the 254 patients undergoing first opinions, 11 had ND; 9 had MS, while 2 had ALS [Table 1]. Patients averaged 60.5 years of age with a range of 53-77. There were six males and five females. For patients with ND radiologists/neuroradiologists interpretation of spondylotic changes on MR and/or CT studies. For 11 ND patients, radiologists'/neuroradiologists' interpretation of significant spondylotic spinal disease on MR/CT studies prompted referrals by primary care physicians/neurologists for first opinion surgical spinal consultations.For these 11 patients, studies had been interpreted as demonstrating; multilevel posterior cervical cord compression (3 patients), single or multilevel anterior cervical disc disease/cord compression (3 patients), or multilevel lumbar disc disease/stenosis resulting in thecal sac/nerve root/cauda equina compression (5 patients).

Table 1: Clinical data for first opinion patients with or without neurodegenerative disease

Variable	Patients without neurodegenerative diseases	Patients with neurodegenerative syndromes
Number	243	11
MS		9
ALS		2
Sex		
Males	114	6
Females	129	5
Average age	57.4	60.5
Range	(25-88)	(53-77)
Std dev	14.2	7.45
No surgical disease	106 patients	9 patients
Surgical disease	137 patients	2 patients
	25 Lumbar laminectomies and fusions	Patient 1: Diskectomy/ laminectomy/fusion
	71 Lumbar laminectomies	L2-L3 (Grade I slip)
	28 Cervical laminectomies and fusions	Patient 2: Cervical laminectomy C5-C7 and posterior fusion C2-T2
	1 Tumor	and (6 months later)
	12 ACDF	Lumbar laminectomies L2-S1 with *In situ* L4-S1 fusion

MS: Multiple sclerosis, ALS: Amyotrophic lateral sclerosis

2 Of 11 first opinion ND patients had surgery

Two patients had "significant" spinal spondylotic changes on MR and/or CT findings to warrant surgical intervention. Of interest, both underwent up-dated MR and CT examinations that were reviewed with a specialized spinal neuroradiologist. The first patient exhibited myeloradiculopathy that correlated with both MR/CT documented dorsolateral cord compression with inward shingling warranting a C5-C7 cervical laminectomy with posterior C2-T2 instrumented fusion. [Figures 1-5]. Postoperatively, although her new myelopathic findings resolved, most of her long-standing MS-related deficits remained unchanged. Additionally, 6 months later, she required a lumbar laminectomy

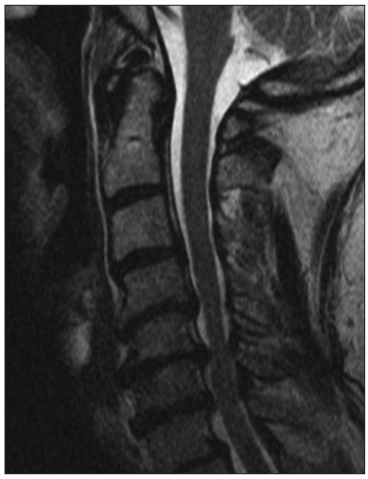

Figure 1: Preoperative Cervical Sagittal T2-Weighted MRI The preoperative sagittal T2 MRI showed spinal stenosis at the C56/C67 levels. Dorsolateral compression was due to inward shingling of the lamina and OYL. Ventral disease was attributed to hypertrophy of the posterior longitudinal ligament (not fully ossified on the CT), and not disc disease. She had a laminectomy C5-C7 with posterior fusion C2-T2 to maintain alignment. Postoperatively, her myelopathy (recent) improved, but prior deficits from MS remained unchanged

Figure 2: Preoperative Midline Sagittal 2D CT Study. The parasagittal preoperative CT study demonstrated adequate preservation of a cervical lordosis without kyphosis, posterolateral inward shingling of the lamina of C5-C7, without OPLL anteriorly or OYL posteriorly. The vertebral bodies of C4-C6 were also spontaneously fused. The preoperative MRI documented multilevel cord compression from C4-C7 that was adequately decompressed following a C5, C6, C7 laminectomy with undercutting of C4 and T1 to remove hypertrophied yellow ligament

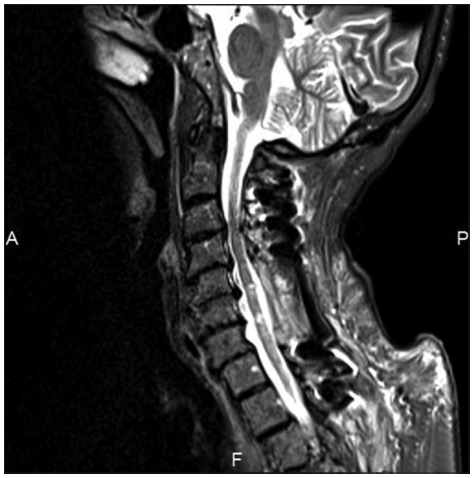

Figure 3: Postoperative T2-Weighted MRI Scan of Patient with Multiple Sclerosis Following a Cervical Laminectomy C5, C6, C7 Followed by C2-T2 Fusion. The study demonstrated chronic cord atrophy and marked focal intracord T2 hyperintensity from C4-C7, most prominent at/immediately below the C45 level. A central less intense signal was seen at C3 without cord expansion/pathological enhancement, consistent with the history of MS and chronic myelomalacia. Excellent decompression of the spinal canal was seen at all levels without recurrent/residual spinal stenosis/extrinsic cord deformity

Figure 4: Preoperative Parasagittal 2D CT Study of Lumbar Stenosis. The parasagittal 2D preoperative CT scan documented marked stenosis at the levels of L2-S1 with Grade I spondylolisthesis at the L45 level and Grade I-II spondylolisthesis at the L5S1 level. Ossification of the yellow ligament contributing to both central and bilateral lateral recess stenosis that was also confirmed on the preoperative MRI

L2-S1 for stenosis and an *in situ* fusion L4-S1 for Grade I or I/II spondylolisthesis (L45/L5S1) respectively. Of interest, one week following the lumbar surgery, she sustained a mild exacerbation of her long-standing right hemiparesis attributed to an exacerbation of her underlying MS. The second patient underwent a L2-L3 diskectomy with laminectomy for stenosis with an in-situ fusion for grade I spondylolisthesis; he did well without further sequelae.

9 First opinion ND patients without surgical disease

Nine ND patients with spinal "spondylosis" seen in first opinion did not warrant surgery. The first author, a spinal neurosurgeon, came to this conclusion following careful review of all diagnostic studies with a specialized spinal neuroradiologist combined with obtaining additional better quality/updated MR (7 patients) and/or CT (3 patients) examinations.

Typically, in the cervical spine, the spondylotic changes included single or multilevel calcified/ossified

Figure 5: Lumbar Preoperative Parasagittal T2-Weighted MRI Study. The preoperative T2 MRI similarly documented multilevel stenosis from L2-S1. There was diffuse ventral intrusion from degenerative discal changes accompanied by dorsolateral ossification/hypertrophy of the yellow ligament; these findings contributed to marked lateral recess compromise. Also noted was the Grade I spondylolisthesis at the L45 level and Grade I-II spondylolisthesis at the L5S1 level

spurs, or degenerated "black" or hypointense discs that intruded only on the subarachnoid space/dura, without contributing to significant spinal cord and/or nerve root compression. Similarly, in the lumbar spine, degenerative spondylotic/stenotic changes, hypointense changes consistent with "black discs", and mild olisthy were often again noted, but did not contribute to significant thecal sac or focal nerve root compression.

7 Second opinion ND patients told they needed "Unnecessary" Spinal Surgery

Seven of 183 patients coming in for second spinal surgical opinions, where prior spinal surgeons recommended surgery, had underlying MS (3 patients), ALS (2 patients), or PPS (1 patient) [Table 2 and Figures 6-8]. These patients averaged 54.6 years of age (range 33-70 years), and included three males and four females. Based on outside MR/CT findings, patients had been offered; two lumbar procedures (1 laminectomy, 1 laminectomy/fusion), two posterior decompressions for cervical stenosis/multilevel cord compression, and three single/multilevel anterior cervical diskectomy/fusions for disc disease/stenosis and cord compression. Typically, however, spondylotic pathology only contributed to mild intrusions on the cervical or lumbar subarachnoid space, without "significant" focal cord, nerve root, or cauda equina compression. None of the spondylotic changes identified on the original outside MR studies appeared to warrant spinal surgery. However, before finalizing these opinions, the author ordered new and often better quality MR studies (5 patients) and/or selective CT examinations (3 patients). All studies were then reviewed with a specialized spinal neuroradiologist who confirmed that none exhibited significant neurological compression to warrant an operation; the author agreed that none of these patients warranted spinal surgery.

Table 2: Clinical data for second opinion patients without or with neurodegenerative disease

Variable	Patients without neurodegenerative diseases	Patients with neurodegenerative diseases
Number (age ranges)	176 total	7 total
MS		4
ALS		2
PPS		1
Sex		
Males	64	3
Females	112	4
Average age	55.4	54.6
Range	24-88	33-70
Std dev	14.5	12.9
No surgical disease	104 (59%) patients "unnecessary" surgery advised by initial consultant	7 (100%) patients "unnecessary" surgery advised by initial consultant
		2 cervical laminectomies and posterior/fusions (stenosis)
		3 single or multilevel anterior cervical diskectomies/fusions
		2 lumbar laminectomies/fusions
Surgical disease present	61 (34.7%) "wrong operations" advised by initial consultant	
	11 (6.3%) "right operations" advised by initial consultant	

*TLIF: Transforaminal lumbar interbody fusion, **PLIF: Posterior lumbar interbody fusion, MS: Multiple sclerosis, ALS: Amyotrophic lateral sclerosis, PPS: Post polio syndrome

176 Patients without ND told they needed spinal surgery

There were 176 patients remaining in the second opinion category without ND who were told by outside surgeons that they required spinal surgery. The second opinion surgeon determined that for 104 (59%) patients, the proposed operations were "unnecessary", (e.g., pain only, no neurological deficit, no significantly abnormal radiographic findings). Additionally, for 61 patients (34.7%), an operation was indicated, but not the one proposed by the outside surgeon (e.g., too extensive, wrong access route). Finally, only 11 patients (6.3%) were told the same operation, recommended by the second opinion surgeon [Table 2].

Figure 6: Midline T2-Weighted Sagittal MRI Study in Patient Presenting For Second Opinion Advised to Undergo Cervical Laminectomy with Fusion This 70-year-old female with ALS developed increased cervical myelopathy. Although she was advised to undergo a cervical laminectomy/fusion, the midline sagittal T2 MRI showed no significant ventral or dorsal cord compression; there was only mild dorsal shingling of the laminae of C6C7, without an increased cord signal. She was advised not to undergo cervical surgery

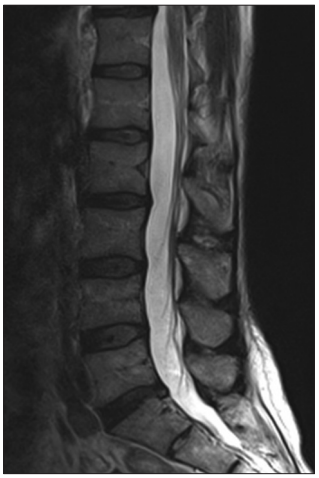

Figure 7: T2-Weighted Sagittal Lumbar MRI Showing No Surgical Disease This 50-year-old female with an underlying diagnosis of MS was offered an L5S1 posterior lumbar interbody fusion for disc disease. Her neurological examination demonstrated a foot drop unrelated to this MRI study as on this sagittal and the axial images there was only loss of disc hydration (minimal degenerative changes) without significant thecal sac or neural compression from disc disease or stenosis. The patient was advised not to undergo lumbar surgery

DISCUSSION

In this study performed over 20 months, we prospectively evaluated 437 patients presenting for consultation with cervical or lumbar spinal complaints; 18 (4.1%) had underlying ND; (MS (13 patients), ALS (4 patients), and PPS (1 patient)). We found that the need for spinal surgery among ND patients was extremely rare. Only 2 of 11 first opinion ND patients (out of a total of 254) required spinal surgery, while none of 7 ND second opinion patients warranted surgical intervention. For the latter patients, pursing a second spinal surgical opinion avoided "unnecessary" operations.

There are several potential explanations as to why patients consult spinal surgeons as first or second opinions. First, and foremost, they want to know if they have a surgical problem. First opinion patients are typically referred by medical consultants or neurologists based on radiologists' or neuroradiologists' over interpretations of MR findings. This is also the case for patients coming in for second surgical spinal opinions, but here, the major problem is that the previous surgeons who recommended surgery may not have independently read or could not adequately read the MR/CT studies to independently determine whether the radiologists'/neuroradiologists' interpretations were correct (vs. exaggerated or over-interpreted). Certainly, the ability to independently read these studies should be considered crucial if spinal surgeons are to do their job. However, radiologists'/neuroradiologists' often focus on over reporting nonessential and clinically insignificant findings in order to avoid "missing" anything as it could result in suits. Some spinal surgeons, on the other hand, may not have the training/ability to distinguish surgical from non-surgical disease, a shortcoming that may lead to "unnecessary spinal surgery". For the 7 patients presenting for second opinions, the inability of their first opinion surgeons to adequately interpret their diagnostic studies almost led to "unnecessary" surgery. Reassessment by the second opinion surgeon, and a specialized spinal neuroradiologist prevented this from happening in these 7 cases. However, there must be many more out there that were not as fortunate.

ADDITIONAL LITERATURE REVIEW

Efficacy of spinal surgery in patients with both spondylotic myelopathy and histories of multiple sclerosis

Differentiating between cervical spondylotic myelopathy (CSM) and MS is often difficult, as both may exhibit similar symptoms and signs of myelopathy.[1,9] For MS patients, spinal cord lesions can cause radicular pain through involvement of the dorsal root entry zone, thus mimicking structural disease. The analysis of these patients is often further complicated when intramedullary MS cord lesions and spinal cord compression occur at the same level.

Surgical lesions in patients with both MS and CSM

In the Arnold *et al*. study, 15 patients (10 females and 5 males, averaging 50.1 years of age) with both CSM and MS underwent spinal surgery that included decompressions and fusions.[1] Although patients were known to have MS, all exhibited progressive myelopathy attributed to CSM that correlated with MR-documented cord and/or nerve root compression;

Figure 8: Cervical Midline Sagittal T2-Weighted Study. This 49-year-old male with a history of polio presented for a second opinion after having been advised to undergo an anterior cervical diskectomy/fusion at the C5-C6 level. However, the midline sagittal (and axial) T2-weighted MRI documented only ventral intrusion on the thecal sac at the C56 level without significant cord or root compression and there was no increased signal in the cord. The patient was diagnosed with Postpolio Syndrome (PPS) and advised not to undergo any cervical procedure

all improved with surgical decompression/fusion. Over an average 47-month postoperative follow-up period, 13 demonstrated decreased neck/upper extremity pain/parenthesis, motor function improved in 13 patients and stabilized in two, but bladder incontinence remained. The authors concluded that patients with MS, but clearly documented myelopathy attributed to significant, radiographically documented CSM, may benefit from surgery. Young similarly noted that spine surgery was an effective adjunct in select older patients with both CSM and ND (MS or ALS) where patients exhibited "significant" MR findings of cord and/or root compression (correlating with discs, osteophytes/stenosis with ligamentous hypertrophy).[9]

Difficulty in differentiating between ALS vs. CSM vs. other neurological disorders

Rowland noted that in order to establish the diagnosis of ALS, it is critical to utilize electrodiagnostic techniques that can differentiate between multifocal motor neuropathy vs. CSM.[5] Furthermore, ALS had to be distinguished from other conditions that included; benign fasciculation (Denny–Brown, Foley syndrome), paraneoplastic syndromes, lymphoproliferative disease, radiation damage, monomelic amyotrophy (Hirayama syndrome), Parkinsonism, dementia, and other multisystem neurological disorders. Magistris added the following to the list of differential diagnostic considerations (neurodegenerative and/or endocrine disorders) of ALS: Multifocal motor neuropathy, Kennedy's bulbospinal atrophy, CSM, hyperthyroidism, hyperparathyroidism, PPS, and postradiation syndromes.[2] He, like Rowland, agreed that electrodiagnostic techniques should, when correlated with clinical findings and other markers, ultimately led to correctly establishing the diagnosis of ALS.

Nearly half of ALS patients are misdiagnosed with cervical spondylosis

Yamada *et al.* observed that nearly half of ALS patients are misdiagnosed with cervical spondylosis.[8] As both groups of patients typically exhibit progressive neurological deterioration, those with ALS alone should not be subjected to "unnecessary" spinal surgery. Looking at 63 middle to older aged patients with ALS, the majority had CSM (30 patients), while others had lumbar spondylosis (7 patients), ossification of the posterior longitudinal ligament (OPLL) (4 patients), or ossification of the yellow ligament (OYL) (4 patients). Spinal surgery was performed shortly after the diagnosis of ALS was established in six patients: Five with cervical disease (7.9%), and one with lumbar pathology (1.6%). Surgery was of "questionable benefit", as most exhibited progressive worsening of motor symptoms within a relatively short duration due to their underlying ALS.

Tandem spinal stenosis and ALS both contribute to progressive muscle weakness

Opstelten and Boon evaluated two patients, aged 35 and 72, with progressive muscle weakness characterized by upper and lower motor neuron signs, without sensory findings.[4] However, the clinical examination, although consistent with ALS, did not include the typical brain stem signs. As both patients exhibited significant tandem cervical and lumbar spondylotic/stenotic lesions on MRI/CT scans, they both underwent tandem spinal surgery. Postoperatively, the patients' deficits remained unchanged, prompting the authors to emphasize that although tandem spinal stenosis must be considered along with ALS, improvement with spinal surgery cannot be assured.

Trapezius motor evoked potentials help distinguish between ASL and CSM

Truffert *et al.* utilized transcranial magnetic stimulation of the trapezius and limb muscles to evaluate and differentiate ALS (10 patients) from CSM (9 patients) in 19 patients, while also comparing them with 23 normal control patients.[6] Central motor conduction times, amplitude ratios, and trapezius inter-side asymmetry were all evaluated. Limb motor evoked potentials (MEPs) were abnormal in most ALS and CSM patients (17/19). However, trapezius MEPs proved helpful in differentiating between ALS and CSM; trapezius MEPs were abnormal in all ALS patients, but normal in 8 of 9 patients with CSM.

Diagnosing and differentiating postpolio syndrome vs. CSM

Pain considerations in postpolio syndrome

Although patients with CSM and PPS may exhibit similar pain syndromes, those with PPS must have a prior history

(often remote) of an acute polio infection.[7] Predominant symptoms include; progressive motor deficit/weakness, atrophy, fatigue and pain (the latter more frequent in younger females using the Visual Analog Scale (VAS). In Werhagen and Borg's study, patients underwent neurological examinations, and answered Short-Form 36 (SF-36) and VAS questionnaires. Pain, observed in 77 (68%) patients, appeared to significantly impact SF-36 Vitality and General Health scales as well as VAS scores, and appeared to be a prominent factor in patients with PPS.

Function and structure of the neuromuscular junction in postpolio syndrome differentiate it from csm

Patients with CSM may be differentiated from those with PPS utilizing neurodiagnostic testing. For example, Maselli *et al*. evaluated the morphology and electrophysiology of the neuromuscular junction utilizing muscle biopsies from 10 patients with PPS.[3] Intracellular microelectrode recordings, histologic evaluations, and electron microscopy showed different types/degrees of failure of transmission at the level of the neuromuscular junction. Nevertheless, although functional and structural abnormalities were frequently documented, they were not uniformly noted, and therefore, "do not appear to be a necessary condition to define the post-poliomyelitis syndrome".

CONCLUSION

In this series, 18 of 437 patients seen by a single spine surgeon over a 20-month period for cervical and lumbar complaints had underlying ND syndromes. Eleven of the 254 patients seen in first opinion, wherein they had not previously consulted a spinal surgeon, had MS or ALS; only 2 required spinal surgery. Seven of 183 patients seen in second opinion, where a prior surgeon recommended a spinal operation, had MS, ALS, or PPS; none required spinal surgery. Therefore, very few patients with MS, ALS, or PPS presenting for first or second spinal surgical opinions actually required spinal surgery. This must be attributed to the fact that MR studies were often over-interpreted by radiologists/neuroradiologists and some spinal surgeons. Being able to distinguish nonsurgical disease from truly surgical pathology on neurodiagnostic studies should not only be the purview of the radiologists/neuroradiologists, but should also be considered a critical part of the expertise/training of the spinal surgeon who must use these studies to determine whether or not an operation should be performed. How else will we avoid "unnecessary" spinal surgery in patients with ND syndromes?

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