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Dystonic Movement Disorders and Spinal Degenerative Disease

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Choreoathetosis · Dystonia · Movement disorders · Spine

Abstract
The occurrence of degenerative spinal disease subsequent to dystonic movement disorders has been neglected and has received more attention only recently. Spinal surgery is challenging with regard to continuous mechanical stress when treatment of the underlying movement disorder is insufficient. To characterize better the particular features of degenerative spinal disease in patients with dystonia and to analyze operative strategies, we reviewed the available published data. Epidemiologic studies reveal that degenerative spinal disorders in patients with dystonia and choreoathetosis occur much earlier than in the physiological aging process. Dystonic movement disorders more often affect the spine at higher cervical levels ($C_{2–5}$), in contrast to spinal degeneration with age which manifests more frequently at the middle and lower cervical spine ($C_{5–7}$). Degenerative changes of the cervical spine are more likely to occur on the side where the chin is rotated or tilted to. Various operative approaches for treatment of spinal pathologies have been advocated in patients with dystonic movement disorders. The available data do not allow making firm statements regarding the superiority of one approach over the other. Posterior approaches were first used for decompression, but additional anterior fusion became necessary in many instances. Anterior approaches with or without instrumented fusion yielded more favorable results, but drawbacks are pseudarthrosis and adjacent-level disease. Parallel to the development of posterior fusion techniques, circumferential surgery was suggested to provide a maximum degree of cord decompression and a higher fusion rate. Perioperative local injections of botulinum toxin were used initially to enhance patient comfort with halo immobilization, but they are also applied in patients without external fixation nowadays. Treatment algorithms directed at the underlying movement disorder itself, taking advantage of new techniques of functional neurosurgery, combined with spinal surgery have recently been introduced and show promising results.

Movement disorders affecting the neck can result in premature cervical spondylosis and other degenerative spinal pathology [1–11]. In particular, dystonic movement disorders can cause severe degenerative disease of the upper cervical spine including even spontaneous craniocervical osseous fusion, fractures and subluxation [1, 6, 11, 12]. Nevertheless, the occurrence of degenerative

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spinal pathology still is often neglected in movement disorders, and patients are referred only late or not at all for surgical treatment [2]. Cervical myelopathy and radiculopathies due to cervical spondylosis may result in secondary neurological deficits and additional disability [6]. Patients with spinal pathology due to poorly controlled movement disorders usually are not considered good candidates for spinal surgery, first because it is often assumed that only limited benefit will be achieved, and second because it is thought that surgical complications and the risk of spinal instability are higher than in other patients. Several case reports and series on spinal surgery in patients with movement disorders have been published within the past three decades. Treatment concepts, however, differ markedly regarding the surgical approach, the extent of decompression, the need for postoperative fusion or instrumented stabilization, and the methods used for spinal stabilization. Only recently has more attention been paid to concurrent treatment of the underlying movement disorder by functional neurosurgical procedures.

Here, we review the available data on the occurrence and the frequency of spinal pathology in dystonic movement disorders both from a neurological and a neurosurgical perspective. We also analyze published data on surgical strategies and postoperative outcome, and suggest treatment strategies for patients who need spinal surgery.

Dystonic Movement Disorders and Degenerative Spinal Disease

Abnormal neck movements in patients with generalized or focal dystonia tend to accelerate degeneration of the cervical spine due to mechanical overstress to the disks and zygapophysial joints. Cervical spondylosis with radiculopathy or myelopathy has been reported in patients with generalized dystonia [10, 13–26] and in patients with cervical dystonia (CD) and segmental dystonia [2, 3, 5, 26–28]. There is little epidemiologic data on the incidence of degenerative spinal disorders in patients with dystonic movement disorders. Overall, the prevalence has been estimated to range between 18 and 41% [6]. Comparison with control groups, however, is mostly lacking and patient selection is subject to bias.

In 1962, Anderson et al. [14] were the first to describe the occurrence of cervical spondylosis in patients with congenital athetosis. Soon thereafter, Levine et al. [21] reported a high incidence of degenerative spinal disorders in a study on 20 patients with dystonic movement disorders (mean age 41.5 years; range, 21–56 years). Relevant degenerative changes were demonstrated in 50% of their patients on X-rays. Those with advanced degenerative changes had a longer history of movement disorders. Further epidemiological data were provided mainly by Japanese authors in the 1980s [17, 19, 22]. Harada et al. [18] compared X-rays of 180 patients with athetoid cerebral palsy (CP) to those of 417 unselected controls with spinal disorders. In their series, 43 out of the 180 CP patients were operated on for cervical myelopathy. Marked to severe disk degeneration (grades 3 and 4 according to their classification) was found in 65% of operated CP patients and in 46% of nonoperated CP patients, whereas disk degeneration grades 3 and 4 was seen in only 6% of the control group. In a recent systematic CT study on patients with CD (mean age 53.5 years), 14 out of 34 patients (41%) had moderate to severe signs of cervical spine degeneration [2]. The extent and severity of the degenerative changes clearly exceeded that which would have been expected in a reference population of the same age [29, 30].

Radiculopathies or myelopathy manifest at a younger age in patients with movement disorders. In series of patients with early-onset generalized dystonia or choreoathetosis the mean age ranged between 33 and 38 years [18, 19, 22]. In CD patients, radiculopathies or myelopathy were noted at a mean age of 48 years [5]. Mean duration from onset of CD until diagnosis of radiculopathy or myelopathy was 5.8 years. When a group of CD patients with no or slight degenerative changes of the cervical spine was compared to another group of CD patients with moderate or severe changes, no significant difference in age, sex, duration of torticollis, degree of disability, or pain was found [2]. For the group with moderate or severe spinal degeneration, however, the mean interval between onset of the movement disorder and effective treatment was significantly longer (10.1 years), and head tremor was severer.

Several studies have shown that dystonic movement disorders tend to affect the cervical spine at higher levels more frequently [10, 13, 14, 18–21, 23, 31]. In CD patients, degenerative changes were reported predominantly at the $C_{2/3}$ and $C_{3/4}$ levels [2]. Physiological movements in normal aging, in contrast, have their main degenerative impact on the lower cervical spine [9, 32, 33]. Degenerative changes usually occur in the segments C_3–C_7 and involve most frequently the uncovertebral joints at $C_{5/6}$. The segment $C_{6/7}$ is the second most frequently affected, whereas the segment $C_{2/3}$ is most rarely involved. Thus, the degenerative changes in the segments $C_{2/3}$ in CD pa-

tients appear not to be related to an accelerated aging process. The $C_{2/3}$ intervertebral joints are a transition zone between the upper cervical spine, which is mainly mobile in the horizontal plane (rotation), and the lower cervical spine, where movements in the sagittal plane predominate [34]. Therefore, biomechanical stress by involuntary, particularly complex rotatory movements in CD and CP patients have more impact on the upper cervical spine [19, 31, 35–37]. The mobility of the $C_{0/1}$ and the $C_{1/2}$ segments does not decrease with age, but on the contrary increases somewhat, whereas ankylosis gradually develops age-dependently in the lower cervical segments [38]. In addition, the combination of abnormal lateral flexion of the neck and rotation is likely to be detrimental in dystonia patients [36, 38]. Thickening of the ligamentum flavum is another important factor for the development of radiculopathy and myelopathy [19, 39]. Degenerative changes often are asymmetrical, and they are found more frequently on the side where the head is rotated or tilted to [2, 21]. Instability of the cervical spine also tends to be more frequent in patients with dystonic movement disorders. Harada et al. [18] reported listhetic instability at the $C_{3/4}$ level in 17%, and at the $C_{4/5}$ level in 27% of their CP patients, whereas it was six times, respectively eight times, less frequent in their control group. A systematic study of cervical instability was performed in another series of 57 patients with athetoid CP by dynamic X-rays [31]. Segmental instability, defined as anterior or posterior slippage of more than 3 mm or increased rotation, was found to be frequent and at multiple levels of the cervical spine. It was present at $C_{2/3}$ in 13% of patients, at $C_{3/4}$ in 26%, at $C_{4/5}$ in 35%, at $C_{5/6}$ in 15%, and at $C_{6/7}$ in 4%. When velocity and acceleration during flexion/extension motion of the cervical spine was analyzed in patients with athetoid CP, a sudden increase in velocity and acceleration occurred during rapid movements at various levels, resulting in a larger overall range of motion of the cervical spine in CP patients but not in controls [35]. The greater shearing forces and bending stress were suggested to predispose the cervical spine to premature instability.

Recently, it has been postulated that scoliosis, which is idiopathic in most cases, but which might also be due to degenerative spinal disease, could be a risk factor for the development of CD. A multicenter case-control study showed that idiopathic scoliosis developing in middle or late childhood or at around puberty is associated more frequently with CD independent of age, duration of disease, education, other spinal diseases and family history of dystonia [40]. The authors suggested that static changes may induce compensatory involuntary postures or might alter peripheral sensory input from the trunk leading to central cortical and subcortical reorganization.

Spinal Surgery in Dystonic Movement Disorders

The first attempt at spinal surgery in a patient with dystonia was reported by Anderson et al. in 1962 [14]. The operation was discontinued, however, due to intraoperative complications, and it took several years until this issue was reconsidered. A chronological survey over the studies on surgery of degenerative spinal disorders in dystonic movement disorder patients is summarized in table 1 [4, 5, 8, 13–22, 24, 28, 41–46]. Note that reports in Japanese were not included in this analysis. Overall, approximately 200 patients with dystonic movement disorders undergoing spinal surgery are described in the literature written in English. Interpretation and comparison of reports, however, is difficult because of several methodological limitations, such as lack of clear differentiation between focal, segmental and generalized forms of dystonia and the absence of standardized and comparable scales to measure outcome. Furthermore, some studies include only few patients, the study design is retrospective, and information on follow-up is limited. In what follows, we will review the impact of different approaches and techniques as far as possible.

Posterior Approach

Laminectomy for decompression of the cervical spinal cord and the foramina – introduced in the early 1970s – was the first technique which was routinely used in patients with dystonic movement disorders [16, 20, 21, 43]. In early series, postoperative results were only rarely satisfactory. At least partially, this was due to poor patient selection and in some cases it was recognized that surgery had been carried out 'too late' in patients who were already plegic and had irreversible neurological damage [21]. A major disadvantage of posterior decompression in early series was the lack of technical equipment to perform stabilization via a posterior approach. Although early postoperative improvement was achieved after posterior decompression, it became necessary to add anterior fusion techniques in some instances when deformity increased with subsequent instability on long-term follow-up [22]. This led to the general belief that laminectomy would be contraindicated in patients with dystonia [22].

Table 1. Literature review of surgery for treatment of degenerative spinal disorders in dystonic movement disorder patients (listed in chronological order)

Authors, year of publication	Patients	Age at surgery, years	MD	Duration of MD prior to surgery, years	Level of pathology	Approach anterior (A) posterior (P)	Operative procedure	Length of FU after surgery	Functional outcome	Outcome of spinal surgery
Levine et al. [21], 1970	1	38	CA	38	$C_{3–7}$	P	extensive laminectomy (C_2–T_1)	weeks	unchanged ('downhill course continued')	NA
Brown et al. [41], 1982	8	11–30	CA	NA	thoraco-lumbar scoliosis	A + P	anterior discectomy + instrumented fusion; posterior instrumented fusion	1–7 years (mean 3.9 years)	mild imp.	correction of scoliosis in most patients; solid fusion in 82% of patients; problems with instrumentation in 50%
McCluer [43], 1982	3	41–60	CA	41–60	$C_{2–7}$; $C_{3–7}$; $C_{4/5}$	A (1); P (2)	discectomy (1); laminectomy (2)	NA	unchanged or worsening	NA
Hirose and Kadoya [19], 1984	4	27–41	CA	27–32	$C_{3/4}$ (3); $C_{3–5}$ (1)	A	discectomy; iliac crest grafting (3)	1.4–4.5 years	moderate imp.	solid fusion
Nishihara et al. [22], 1984	9	19–52	CA	NA	$C_{2/3}$ (1); $C_{3–5}$ (7); $C_{4/5}$ (1)	A (7); A + P (2)	discectomy + iliac crest grafting (9); laminectomy (2)	1–12 years (mean 5.4 years)	moderate imp. (7) mild imp. (1) worsening (1)	good fusion with anterior interbody fusion and halo (7); instability after laminectomy (additional anterior operation after 3 + 6 years)
Fuji et al. [17], 1987	10	30–58	CA	NA	predominantly upper cervical spine	A (7); A + P (3)	corporectomy or discectomy; iliac crest grafting; interspinous wiring (3)	2–6 years (mean 5 years)	marked imp. (6) mild imp. (3) no imp. (1)	good fusion (6); collapse of the graft (2); kyphotic deformity (1); pseudarthrosis (1)
Kidron et al. [20], 1987	2	45, 64	CA	NA	$C_{5–7}$; $C_{4–7}$	P	laminectomy	NA	worsening	NA
El-Mallakh et al. [16], 1989	2	48, 68	CA	48, 68	$C_{3–7}$; $C_{3–6}$	P	laminectomy	1 + 2 years	mild imp.	NA
Traynelis et al. [28], 1992	2	34, 64	CD	34, 15	$C_{4–7}$; $C_{0/1}$	P	laminectomy + fusion with lateral mass plates $C_{4–7}$ (1); occipitocervical fusion (1)	4–6 months	NA	solid fusion after 6 months of halo vest immobilization (1)
Adler et al. [13], 1996	3	34–62	CD (2); GD (1)	20–22	$C_{4–6}$ (2); $C_{3–6}$ (1)	A + P (1); A (1); P (1)	anterior and posterior fusion (1); discectomy + fusion (1); laminectomy (1)	2–9 months	unchanged (1) mild imp. (2)	stable fusion (2; CD, GD); no instability after laminectomy in CD
Harada et al. [18], 1996	43	19–60	CA	NA	$C_{2–7}$	NA	NA	NA	NA	NA
Mikawa et al. [44], 1997	2	35, 61	CA	35, 61	$C_{3–6}$; $C_{4–7}$	A + P	discectomy + iliac crest grafting (1); anterior corporectomy iliac crest grafting + posterior instrumental fusion (1)	3 + 4 years	moderate imp.	repeat surgery after anterior approach only + second repeat surgery with posterior fusion; finally solid fusion in both patients
Pollak et al. [10], 1998	4	31–42	CA	31–42	$C_{3/4}$ (2); $C_{4/5}$ (1); $C_{5/6}$ (1)	A	discectomy + iliac crest grafting (3); discectomy + 'bone cement' (1)	1–7 years	mild imp. (2) unchanged (2)	solid anterior fusion (3); graft displacement (bone cement) requiring reoperation (1)

Table 1 (continued)

Authors, year of publication	Patients	Age at surgery, years	MD	Duration of MD prior to surgery, years	Level of pathology	Approach anterior (A) posterior (P)	Operative procedure	Length of FU after surgery	Functional outcome	Outcome of spinal surgery
Racette et al. [24], 1998	2	38, 46	CA	38–46	$C_{4–6}$; $C_{3–7}$	A; A + P	anterior corporectomy with fibular allograft fusion + plating (1); discectomy + iliac crest grafting + plating + laminectomy + fusion	9 + 12 months	mild imp.	solid fusion
Epstein [42], 1999	2	48, 52	CA	48, 52	$C_{2–7}$; $C_{3–7}$	A + P	anterior corporectomy with fusion and plating; posterior wiring and fusion	9 + 12 months	mild imp.	solid fusion; anterior graft displacement (1)
Onari [45], 2000	17	19–62	CA	19–62	predominantly $C_{3–6}$	A + P	discectomy; iliac crest grafting and plating + posterior instrumented fusion (iliac bone + wave-shaped rods)	5–15.5 years	moderate imp. (3) mild imp. (11) worsening (3)	solid fusion
Fricka et al. [4], 2001	1	15	GD	9	C_0-sacrum (lordosis)	A + P	instrumented fusion anteriorly from $T_{7–12}$ + posteriorly from C_3–L_2	3 years	mild imp.	repeat surgery at 8 months postoperatively with fusion of $C_{0–5}$; correction of deformity at last follow-up
Krauss et al. [8], 2002[1]	3	40–46	CA (2); GD (1)	34–46	$C_{2–7}$; $C_{3–5}$; C_7–T_1	P (2); A (1)	laminectomies (2); multilevel corporectomy + cage fusion/plating (1)	20 months	mild imp.	solid fusion (1)
Onari et al. [46], 2002[2]	23	19–66	CA	19–66	predominantly $C_{3–6}$	A + P	discectomy; iliac crest grafting and plating + posterior instrumented fusion (iliac bone + wave-shaped rods)	5–17 years	good imp. (3) moderate imp. (11) mild imp. (3) unchanged (3)	solid fusion

CA = Choreoathetosis; FU = follow-up; GD = generalized dystonia; imp = improvement; MD = movement disorder; NA = not available. Figures in parentheses indicate number of patients.

[1] Patients underwent pallidal stimulation for treatment of their movement disorders.

[2] Extended series.

Nevertheless, several surgeons continued to perform posterior decompression in selected patients with acceptable results [8, 13]. In particular, favorable outcomes were reported in patients who underwent limited-level laminectomy for focal posterior osseous or ligamentous compression.

Anterior Approach

Anterior approaches and fusion techniques in patients with dystonic movement disorders were first promoted in the early 1980s by Japanese groups [17, 19, 22]. Hirose and Kadoya [19] achieved anterior decompression via multilevel discectomies and interbody fusion by autologous iliac bone grafts in a series of athetoid CP patients with cervical radiculopathy and myelopathy. They reported moderate improvement of motor function, walking ability and bladder dysfunction up to 4.5 years postoperatively, whereas sensory status remained unchanged. In another series of athetoid CP patients, postoperative outcome showed mild to moderate recovery of neurological deficits in 8 out of 9 cases after anterior decompression and iliac crest fusion [22]. Fusion was supported by immobilization with a halo device. This concept was also adopted to perform corporectomies in patients with athetoid CP and cervical degenerative pathology [17]. In this series, postoperative improvement classified on the basis of the former activities of daily living was considered marked in 6 patients, mild in 3 and poor in 1. Postoperative complications included pseudarthrosis and adjacent-level instability. Therefore, the au-

thors recommended interspinous wiring as a supplement to anterior spinal fusion allowing earlier removal of halo fixation.

Despite the drawbacks of anterior fusion which became more evident over the years, mainly pseudarthrosis and adjacent-level disease, anterior decompression and fusion techniques have continued to be used as a single procedure in patients with dystonia. More recently, for example, Pollak et al. [10] reported on the long-term follow-up of anterior fusion in 4 young patients with athetoid CP. Immobilization by Philadelphia collars for 2–3 months after surgery was considered to be sufficient for fusion when iliac crest was used as confirmed on cervical spine X-rays. Graft displacement, however, occurred in a patient in whom methyl methacrylate was used, and a reoperation 5 years after the first intervention was necessary. Partial recovery of neurological deficits due to radiculomyelopathy was reported in all patients.

Combined Approaches

Realizing the relative advantages and disadvantages of the two approaches, anterior versus posterior decompression and stabilization, the concept of combining both approaches became more popular [13, 24, 42, 44, 45]. The introduction of this treatment concept was also fueled by the development of new hardware for posterior stabilization. Parallel to its inception and acceptance in clinical practice, posterior stabilization techniques evolved from simple interspinous and interlaminar wiring techniques to using rods and plates, and finally pedicular and transarticular screws [47–49]. Circumferential stabilization was initially used as a staged procedure in those patients in whom anterior or posterior decompression alone had failed [22]. Since the mid 1990s, circumferential stabilization was recommended as the procedure of choice to achieve optimal cord decompression with the highest fusion rate in patients with dystonic movement disorders [44, 45]. In particular, with this technique it became possible to perform more extensive spinal reconstructions and deformity correction, even in patients with advanced multilevel spinal degeneration.

The largest series, including 23 adult patients with generalized athetosis due to CP suffering from cervical myelopathy or radiculopathy, has recently been reported by Onari et al. [46]. They used wave-shaped rods for posterior stabilization and a screw-plating technique, or alternatively a screw-wiring technique, for anterior stabilization. Fusion was promoted by tricortical iliac crest grafts for interbody fusion, and cancellous/cortical iliac bone grafts for interlaminar fusion. With this technique, consistent functional improvement was achieved in almost all patients. At 1-year follow-up, most patients had relief of upper extremity pain, deltoid muscle weakness, and they were able to feed themselves. Improvement was maintained at a mean follow-up of 8.7 years. Surgery-related complications were described as minor, and there were few hardware failures which did not result in reoperations. Postoperatively, there were minor changes of the mean angular motion at segments adjacent to the stabilization and between C_1 and C_2, which resulted in slight atlantoaxial subluxation in some instances.

Peri- and Postoperative Immobilization

Early after the introduction of decompressive spinal surgery for patients with dystonic movement disorders, it was recognized that postoperative stabilization and fixation is necessary to achieve solid fusion and good long-term functional outcome [22]. In particular, when the movement disorder itself is not being treated, there is increased stress and strain on the decompressed spinal segments as well as on adjacent levels. Immobilization with a halo vest has been shown to be useful to facilitate interbody fusion in patients in whom no spinal instrumentation was applied. It has also been used, however, in cases who underwent extensive spinal reconstruction with multilevel instrumented fusion [28]. With the increasing use of plating techniques for anterior fusion, and wiring and plating techniques for posterior instrumentation and fusion, halo immobilization has been replaced largely by hard collars for several weeks [10, 13]. The more rigid constructs in circumferential stabilization allow patients to wear soft collars resulting in less discomfort in the postoperative period [46].

Botulinum toxin (BTX) was introduced in the 1980s for treatment of movement disorders, and it has been shown to be particularly beneficial in focal dystonia such as CD [50]. The average duration of benefit after intramuscular BTX injection is 3–4 months in CD patients [51]. The initial purpose of using BTX injections in dystonic patients who underwent spinal surgery was to assist in postoperative external immobilization and enhance patients' tolerance and comfort with halo vests [28]. Injections were repeated when longer periods of immobilization were necessary. Subsequently, perioperative BTX injections were also performed by other groups and they are administered on a routine basis nowadays in several centers performing spinal surgery in dystonic patients. With its more widespread use, the concept of perioperative BTX injection is applied also for patients with rigid anterior or posterior instrumentation without external

immobilization to reduce dystonic movements postoperatively and thus facilitate fusion.

Functional Neurosurgery

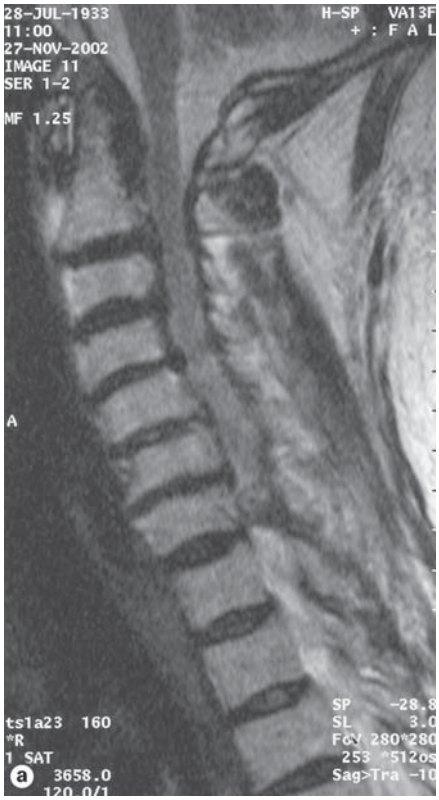
The concept that treatment algorithms for patients with dystonic movement disorders suffering from spinal degenerative disorders should also incorporate adequate measures to treat the movement disorder itself, taking advantage of the armamentarium of functional neurosurgery, gained more attention only recently. Concerning surgery of the cervical spine, two techniques are of particular interest – pallidal deep brain stimulation (DBS) [52, 53] and selective peripheral denervation and myotomy/myectomy. Since we introduced pallidal DBS for patients with otherwise refractory CD, its efficacy and safety have been confirmed by various groups [53–56]. Pallidal DBS results in a long-term improvement of about 60–70% of the movement disorder itself, dystonia-associated neck pain, and disability according to standard rating scales. While phasic dystonic movements may improve within days after surgery, the full extent of amelioration of the tonic-dystonic elements is appreciated more often with a delay of weeks and months. The benefit of pallidal DBS is less pronounced in patients with secondary dystonia. In a recent study, we reported on combining chronic pallidal stimulation and spinal surgery in 3 patients with rapidly progressive cervical myelopathy secondary to severe cervical dyskinesias and dystonia in the context of a generalized dystonic movement disorder [8]. Pallidal DBS was performed before spinal surgery in 2 patients and 1 month thereafter in the third patient. Two patients with choreoathetoid CP underwent multilevel laminectomies. The third patient with primary genetic generalized dystonia had a spinal reconstruction including C_3–C_6 corporectomy, stabilization with a titanium cage filled with autologous bone, C_2–C_7 anterior plating, and halo immobilization for 6 months. In both patients with choreoathetoid CP, the severity and frequency of cervical dyskinesias improved with pallidal DBS. The patient with prominent CD in the setting of generalized dystonia had almost complete relief of dystonic posturing. Progression of cervical myelopathy was halted in all 3 patients at a mean follow-up of 1.5 years. Although no significant change was found in formal assessment of functional disability, patients subjectively reported better performance in activities of daily living.

There is only very little published on the experience with other functional neurosurgical procedures in movement disorder patients with degenerative spinal pathology. A patient with a 'fixed' spine due to craniocervical osseous fusion caused by long-standing CD had marked relief of neck pain after partial myectomy of the dystonic sternocleidomastoid muscle and selective posterior ramisectomy of the contralateral posterior neck muscles despite the position of the head was not corrected [11].

Discussion

Dystonic movement disorders frequently entail degenerative changes of the spine which more often affect upper cervical levels compared to the normal population and which appear at a younger age. The reasons why secondary spinal pathology is diagnosed often only with a delay in these patients are multifold. New neurological signs and symptoms are occasionally not appreciated in the right context, in particular, when they are embedded in a clinical picture with other neurological problems, such as in patients with choreoathetoid CP. Neck pain and pseudoradicular pain, on the other hand, are primary symptoms of CD itself. Good imaging studies are often hard to obtain without sedation or even general anesthesia. Other issues that can be relevant include discontinuous care by several health care providers over the years and sometimes restricted communication with the disabled patient. It is important to note that the duration of inadequate treatment, independent of age and duration of disease, is a significant risk factor for the development of degenerative spinal disease [2]. One of the primary goals of the present review was to increase the awareness of the occurrence of degenerative spinal disease in dystonic movement disorders and to disencourage therapeutic nihilism.

Despite the methodological limitations of most of the studies on spinal surgery in patients with dystonic movement disorders, it can be concluded that, overall, patients achieve mild to moderate benefit, and that progression of secondary symptoms appears to be halted. Apart from yielding immediate improvement of disability, prevention of further decline of functional disability is another important goal in this setting which, however, is much more difficult to measure and to quantify. The disability at the time of surgery most likely is of crucial importance to predict future outcome. While early after the introduction of spinal surgery for patients with movement disorders patients were referred only late or 'too late' for surgery, apparently there has been a shift to earlier referral for surgery within the past decade according to published data. Over the decades, surgical treatment has evolved from simple posterior decompression to anterior decom-



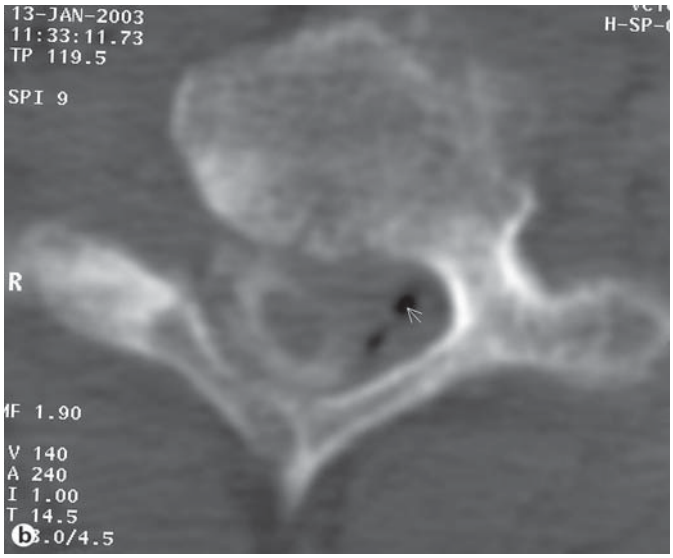
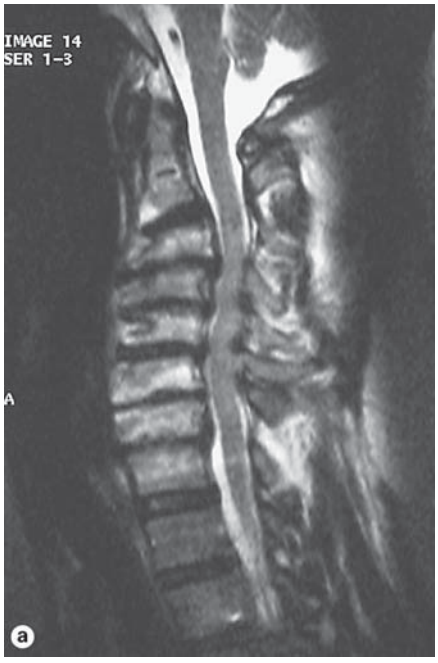
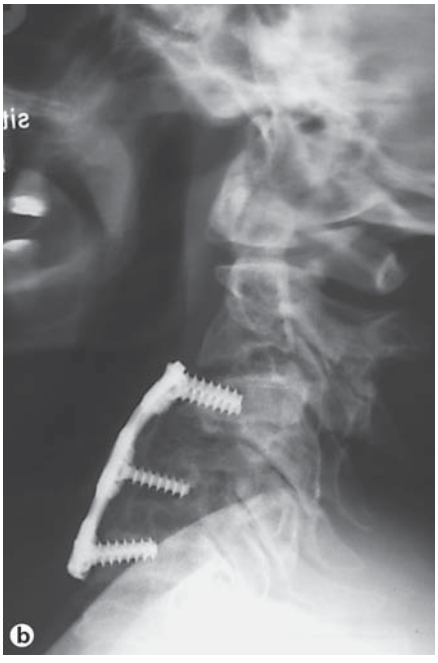


Fig. 1. Sagittal MR scans of the cervical spine (**a**) and axial postmyelographic CT at the C_7 level (**b**) of a 72-year-old man with segmental tonic dystonia due to infantile cerebral palsy. This patient presented with a left-sided C_8 monoradiculopathy. Imaging studies show a synovial cyst with marked compression of the corresponding nerve root. In the presence of relatively mild and predominantly tonic cervical dystonia and with regard to his age, he underwent decompression of the synovial cyst via a posterior unilateral interlaminar fenestration. Two years postoperatively, his pain is alleviated.

Fig. 2. Preoperative sagittal MR scans (**a**) and postoperative X-rays of the cervical spine (**b**) of a 44-year-old man with segmental cervical/orofacial dystonia due to infantile cerebral palsy. This patient presented with deterioration of his gait disorder and C_5 and C_6 radiculopathies of his right arm. Imaging studies showed severe multisegmental spondylosis, spondylarthrosis and osteochondrosis of his cervical spine resulting in marked spinal canal stenosis at C_4–C_5. The movement disorder was predominated by mainly tonic tilt of his head to the left shoulder and rotation of the chin to the right. Therefore, he first underwent selective sternocleidomastoid denervation/myotomy on the left side combined with contralateral C_1–C_6 ramisectomy, which resulted in an 80% relief of his cervical dystonia. One month later, spinal decompression was performed via an anterior approach and two-level corporectomy with subsequent autologous bony fusion and cervical plate stabilization. Postoperatively, both the gait disturbance and the radicular deficits improved, and his condition remained stable over 2 years of follow-up.





pression, circumferential stabilization, and lately to concurrent treatment of the movement disorder itself. This evolution of surgical technique has been paralleled and made possible only by the technical development of spinal surgery and implantation techniques. It appears that primary circumferential stabilization and fusion is superior to simple decompression techniques in subgroups of patients with dystonic movement disorders. Nevertheless, the available data do not allow to draw firm conclusions and establish solid guidelines for treatment. An important question is whether or not we will have better data in the near future and whether the principles of evidence-based medicine may be applied to this peculiar group of patients.

Outcome most often has been evaluated according to composite scores in these patients including relief of symptoms, change of disability and quality of life. As indicated above, 'minor improvement' can be a sufficient goal to achieve under these circumstances. Patient discomfort in the postoperative period also has to be considered in the presence of movement disorders. In that regard, both the introduction of more rigid constraints for spinal stabilization and the concomitant perioperative treatment with BTX injections have been a major progress.

Side effects of spinal surgery have been mentioned only rarely in patients with dystonic movement disorders. However, they may have been underestimated and underreported. In a recent retrospective study on patients with Parkinson's disease who underwent spinal surgery, the number of complications was clearly higher than anticipated [57]. In particular, a markedly increased rate of instrument-related complications and failure was noted. Postoperative deterioration of neurological signs and symptoms may be misinterpreted as a result of progression of the underlying neurological disorder itself. In most studies on spinal surgery for dystonic patients, the follow-up period was relatively short, and thus instrument-related problems may have gone unnoticed.

The progress of functional neurosurgery will have a pivotal impact on treatment algorithms for patients with spinal pathology secondary to dystonic movement disorders. Given the diversity of the phenomenology of dystonia and the spinal pathology in the individual patient, we suggest to tailor treatment plans, similar as has been proposed for denervation procedures in CD [58]. Thus far, the authors have adopted this concept in a recent series of 16 patients with spinal disorders due to dystonia [1, 8]. Data on technical details and outcome will be shown elsewhere. For exemplary cases, see figures 1–3. Such a concept may involve staged surgical procedures – not only for treatment of the movement disorder but also for the spinal problem. Whenever possible, action should be taken to treat the movement disorder first. The plan for spinal surgery should focus both on the present clinical symptoms and the spinal pathomorphology. While monoradiculopathy may be treated adequately by posterior foraminotomy in a patient with prominent torticollis, more extensive decompression and stabilization may be necessary in patients with generalized choreoathetosis with multilevel compression, instability and myelopathy. Circumferential decompression and stabilization certainly yields the highest rate of fusion, but its higher surgical risk and the long-term risks of secondary instability and deformity have to be considered.

Careful evaluation of side effects of spinal surgery and of outcome is necessary along different axes under these special circumstances. Development of techniques and hardware for spinal instrumentation is a rapidly evolving field. Improved techniques have opened new horizons in these multidisabled patients.

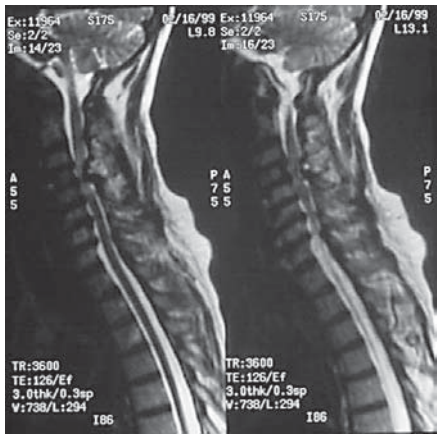


Fig. 3. Preoperative sagittal MR scans of a 40-year-old woman with severe cervical dyskinesias due to choreoathetoid infantile cerebral palsy. This patient suffered from progression of paraparesis over several years, and she was wheelchair bound on presentation. MR studies show multilevel spondylosis and spinal stenosis with cervical myelopathy. There were continuous severe dyskinesias with writhing movements of her neck. She underwent bilateral pallidal DBS, which resulted in moderate improvement in the severity and the range of the dyskinetic movements of her neck. Three months thereafter, she underwent a three-level laminectomy via a posterior approach. The progression of cervical myelopathy was halted over a follow-up of 5 years.

Table 2. General recommendations for surgical treatment of patients with spinal degenerative disease secondary to dystonic movement disorders

Diagnostic challenges	Deterioration of neurological deficits in patients with a stable condition for years may indicate secondary spinal degenerative disease; further investigation should include MR imaging and flexion/extension roentgenological studies for evaluation of instability
Timing of surgery	Spinal surgery should be performed before irreversible neurological deficits are present; in some instances, the more limited goal will be to halt further progression of secondary neurological deficits
	Whenever possible treatment of the movement disorder itself should be considered first (medication, BTX, functional neurosurgery including denervation and myotomy procedures or stereotactic operations)
	In elderly patients with focal spinal pathology and stable movement disorders, spinal surgery only might be sufficient
	Tailored and individualized treatment concepts may be superior to standard approaches
Spinal surgery techniques	Simple limited decompression techniques (anterior or posterior approaches) may be used for treatment of focal pathologic changes
	Anterior decompression with instrumented fusion or circumferential stabilization is the preferred treatment option in patients with instability or multilevel degenerative disease
Postoperative treatment	BTX injections are a useful adjunct to control transiently both dystonic postures and movements.
	Cervical or theses such as soft or hard collars suffice in the majority of cases and are recommended for several weeks
	Halo immobilization may be necessary in special circumstances when the movement disorder may not be controlled adequately or with long and unusual instrumented constructs

Conclusion

The treatment of spinal degenerative diseases in patients with dystonic movement disorders remains a formidable challenge. Clinicians should be aware that neurosurgery offers unique perspectives in these patients given the option to combine spinal and functional neurosurgical techniques to achieve optimal benefit. Some general guidelines on treatment algorithms are summarized in table 2. Given the paucity of data and the rapid development of new technology, however, we have to recognize the preliminary nature of these recommendations.

References

1 Al-Jishi A, Sreekantaswamy: Dystonia associated with atlantoaxial subluxation. Clin Neurol Neurosurg 2000;102:233–235.
2 Chawda SJ, Münchau A, Johnson D, Bhatia K, Quinn NP, Stevens J, Lees AJ, Palmer JD: Pattern of premature degenerative changes of the cervical spine in patients with spasmodic torticollis and the impact on the outcome of selective peripheral denervation. J Neurol Neurosurg Psychiatry 2000;68:465–471.
3 Defazio G, Lepore V, Melpignano C, Lamberti P, Livrea P, Ferrari E: Cervical radiculopathy following botulinum toxin therapy for cervical dystonia. Funct Neurol 1993;8:193–196.
4 Fricka KB, Kim C, Newton PO: Spinal lordosis with marked opisthotonus secondary to dystonia musculorum deformans. Spine 2001;20:2283–2288.
5 Hagenah JM, Vieregge A, Vieregge P: Radiculopathy and myelopathy in patients with primary cervical dystonia. Eur Neurol 2001;45:236–240.
6 Konrad C, Vollmer-Haase J, Anneken K, Knecht S: Orthopedic and neurological complications of cervical dystonia. Acta Neurol Scand 2004;109:369–373.
7 Krauss JK, Jankovic J: Severe motor tics causing cervical myelopathy in Tourette's syndrome. Mov Disord 1996;11:563–566.

8 Krauss JK, Loher TJ, Pohle T, Weber S, Taub E, Bärlocher CB, Burgunder JM: Pallidal deep brain stimulation in patients with cervical dystonia and severe cervical dyskinesias with cervical myelopathy. J Neurol Neurosurg Psychiatry 2002;72:249–256.
9 McCormack BM, Weinstein PR: Cervical spondylosis. West J Med 1996;165:43–51.
10 Pollak L, Schiffer J, Klein C, Mirovsky Y, Copeliovich L, Rabey JM: Neurosurgical intervention for cervical disk disease in dystonic cerebral palsy. Mov Disord 1998;13:713–717.
11 Weigel R, Rittmann M, Krauss JK: Spontaneous craniocervical osseous fusion caused by cervical dystonia. J Neurosurg 2001;95:115–118.
12 Konrad C, Vollmer-Haase J, Gaubitz M, Nabavi DG, Reilmann R, Knecht S: Fracture of the odontoid process complicating tardive dystonia. Mov Disord 2004;19:983–985.
13 Adler CH, Zimmerman RS, Lyons MK, Simeone F, Brin MF: Perioperative use of botulinum toxin for movement disorder-induced cervical spine disease. Mov Disord 1996;11:79–81.
14 Anderson WW, Wise BL, Itabashi HH, Jones M: Cervical spondylosis in patients with athetosis. Neurology 1962;12:410–412.
15 Angelini L, Broggi G, Nardocci N, Savoiardo M: Subacute cervical myelopathy in a child with cerebral palsy. Secondary to torsion dystonia? Childs Brain 1982;9:354–357.
16 El-Mallakh RS, Rao K, Barwick M: Cervical myelopathy secondary to movement disorders: case report. Neurosurgery 1989;24:902–905.
17 Fuji T, Yonenobu K, Fujiwara K, Yamashita K, Ebara S, Ono K, Okada K: Cervical radiculopathy or myelopathy secondary to athetoid cerebral palsy. J Bone Joint Surg 1987;69:815–821.
18 Harada T, Ebara S, Anwar MM, Okawa A, Kajiura I, Hiroshima K, Ono K: The cervical spine in athetoid cerebral palsy. A radiological study of 180 patients. J Bone Joint Surg 1996;78:613–619.
19 Hirose G, Kadoya S: Cervical spondylotic radiculo-myelopathy in patients with athetoid-dystonic cerebral palsy: clinical evaluation and surgical treatment. J Neurol Neurosurg Psychiatry 1984;47:775–780.
20 Kidron D, Steiner I, Melamed E: Late-onset progressive radiculomyelopathy in patients with cervical athetoid-dystonic cerebral palsy. Eur Neurol 1987;27:164–166.
21 Levine RA, Rosenbaum AE, Waltz JM, Scheinberg LC: Cervical spondylosis and dyskinesias. Neurology 1970;20:1194–1199.
22 Nishihara N, Tanabe G, Nakahara S, Imai T, Murakawa H: Surgical treatment of cervical spondylotic myelopathy complicating athetoid cerebral palsy. J Bone Joint Surg 1984;66:504–508.
23 Polk JL, Maragos VA, Nicholas JJ: Cervical spondylotic myeloradiculopathy in dystonia. Arch Phys Med Rehabil 1992;73:389–392.
24 Racette BA, Lauryssen C, Perlmutter JS: Preoperative treatment with botulinum toxin to facilitate cervical fusion in dystonic cerebral palsy. J Neurosurg 1998;88:328–330.
25 Tunkel AR, Pasupuleti R, Acosta WR: Improvement of idiopathic torsion dystonia following dystonia-induced cervical subluxation. J Neurol Neurosurg Psychiatry 1986;49:957.
26 Waterston JA, Swash M, Watkins ES: Idiopathic dystonia and cervical spondylotic myelopathy. J Neurol Neurosurg Psychiatry 1989;52:1424–1426.
27 Adler CH, Kumar R: Pharmacological and surgical options for the treatment of cervical dystonia. Neurology 2000;55(suppl 5):S9–S14.
28 Traynelis VC, Ryken T, Rodnitzky RL, Menezes AH: Botulinum toxin enhancement of postoperative immobilization in patients with cervical dystonia. J Neurosurg 1992;77:808–809.
29 Van Saase JL, van Romunde LK, Cats A, Vandenbroucke JP, Valkenburg HA: Epidemiology of osteoarthritis: Zoetermeer survey. Comparison of radiological osteoarthritis in a Dutch population with that in 10 other populations. Ann Rheum Dis 1989;48:271–280.
30 Zapletal J, de Valois JC: Radiologic prevalence of advanced lateral C1–C2 osteoarthritis. Spine 1997;22:2511–2513.
31 Ebara S, Harada T, Yamazaki Y, Hosono N, Yonenobu K, Hiroshima K, Ono K: Unstable cervical spine in athetoid cerebral palsy. Spine 1989;14:1154–1159.
32 Dvorak J, Antinnes JA, Panjabi M, Loustalot D, Bonomo M: Age- and gender-related normal motion of the cervical spine. Spine 1992;17:S393–S398.
33 Lestini WF, Wiesel SW: The pathogenesis of cervical spondylosis. Clin Orthop 1989;239:69–93.
34 Mestdagh H: Morphological aspects and biomechanical properties of the vertebroaxial joint (C2–C3). Acta Morphol Neerl Scand 1976;14:19–30.
35 Ebara S, Yamazaki Y, Harada T, Hosono N, Morimoto Y, Tang L, Seguchi Y, Ono K: Motion analysis of the cervical spine in athetoid cerebral palsy. Extension-flexion motion. Spine 1990;15:1097–1103.
36 Panjabi MM, White AA: Basic biomechanics of the spine. Neurosurgery 1980;7:76–93.
37 Panjabi M, Dvorak J, Duranceau J, Yamamoto I, Gerber M, Rauschning W, Bueff HU: Three-dimensional movements of the upper cervical spine. Spine 1988;13:726–730.
38 Dvorak J, Penning L, Hayek J, Panjabi MM, Grob D, Zehnder R: Functional diagnostics of the cervical spine using computer tomography. Neuroradiology 1988;30:132–137.
39 Stoltmann HF, Blackwood W: The role of the ligamenta flava in the pathogenesis of myelopathy in cervical spondylosis. Brain 1964;87:45–50.
40 Defazio G, Abbruzzese G, Girlanda P, Buccafusca M, Curra A, Marchese R, Martino D, Masi G, Mazzella L, Vacca L, Livrea P, Berardelli A: Primary cervical dystonia and scoliosis: a multicenter case-control study. Neurology 2003;60:1012–1015.
41 Brown JC, Swank S, Specht L: Combined anterior and posterior spine fusion in cerebral palsy. Spine 1982;7:570–573.
42 Epstein NE: Circumferential cervical surgery for spondylostenosis with kyphosis in two patients with athetoid cerebral palsy. Surg Neurol 1999;52:339–344.
43 McCluer S: Cervical spondylosis with myelopathy as a complication of cerebral palsy. Paraplegia 1982;20:308–312.
44 Mikawa Y, Watanabe R, Shikata J: Cervical myelo-radiculopathy in athetoid cerebral palsy. Arch Orthop Trauma Surg 1997;116:116–118.
45 Onari K: Surgical treatment for cervical spondylotic myelopathy associated with athetoid cerebral palsy. J Orthop Sci 2000;5:439–448.
46 Onari K, Kondo S, Mihara H, Iwamura Y: Combined anterior-posterior fusion for cervical spondylotic myelopathy in patients with athetoid cerebral palsy. J Neurosurg 2002;97:13–19.
47 Epstein N: Posterior approaches in the management of cervical spondylosis and ossification of the posterior longitudinal ligament. Surg Neurol 2002;58:194–207.
48 Harms J, Melcher RP: Posterior C1–C2 fusion with polyaxial screw and rod fixation. Spine 2001;15:2467–2471.
49 Schmidt R, Wilke HJ, Claes L, Puhl W, Richter M: Pedicle screws enhance primary stability in multilevel cervical corpectomies: biomechanical in vitro comparison of different implants including constrained and nonconstrained posterior instrumentations. Spine 2003;15:1821–1828.
50 Jankovic J, Orman J: Botulinum A toxin for cranial-cervical dystonia: a double-blind, placebo-controlled study. Neurology 1987;37:616–623.
51 Dauer WT, Burke RE, Greene P, Fahn S: Current concepts on the clinical features, aetiology and management of idiopathic cervical dystonia. Brain 1998;121:547–560.
52 Coubes P, Roubertie A, Vayssiere N, Hemm S, Echenne B: Treatment of DYT1-generalised dystonia by stimulation of the internal globus pallidus. Lancet 2000;355:2220–2221.
53 Krauss JK, Pohle T, Weber S, Ozdoba C, Burgunder JM: Bilateral stimulation of globus pallidus internus for treatment of cervical dystonia. Lancet 1999;354:837–838.
54 Eltahawy HA, Saint-Cyr J, Poon YY, Moro E, Lang AE, Lozano AM: Pallidal deep brain stimulation in cervical dystonia: clinical outcome in four cases. Can J Neurol Sci 2004;31:328–332.
55 Krauss JK, Yianni J, Loher TJ, Aziz TZ: Deep brain stimulation for dystonia. J Clin Neurophysiol 2004;21:18–30.
56 Parkin S, Aziz T, Gregory R, Bain P: Bilateral internal globus pallidus stimulation for the treatment of spasmodic torticollis. Mov Disord 2001;6:489–493.
57 Babat LB, McLain RF, Bingaman W, Kalfas I, Young P, Rufo-Smith C: Spinal surgery in patients with Parkinson's disease: construct failure and progressive deformity. Spine 2004;29:2006–2012.
58 Krauss JK, Toups EG, Jankovic J, Grossman RG: Symptomatic and functional outcome of surgical treatment of cervical dystonia. J Neurol Neurosurg Psychiatry 1997;63:642–648.