Movement Disorders

84

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Abstract

Movement disorders in persons with intellectual and developmental disabilities are pervasive, often disabling and can be associated with adverse effects of medication as well as intrinsic CNS dysfunction. Recognizing and addressing movement disorders effectively can prevent injuries, tardive dyskinesias and long term disabilities. Video recording and accurate historical documentation of the movements is an important part of being able to diagnose and treat these individuals long term.

Introduction

Individuals with perinatal injury or prenatally determined developmental and intellectual disabilities (IDD) (often genetic or related to placental insufficiency) are susceptible to movement disorders due to abnormal neural networks involving the frontal lobes, basal ganglia and cerebellum. Motor control is determined by three specific networks (see Fig. 84.1). The motor planning areas include the frontal cortex, thalamus, limbic nuclei, parietal sensory cortex and occipital cortex. The motor integration center predominantly involves the basal ganglia (caudate and putamen, globus pallidus, subthalamic nucleus) and cerebellum. The motor execution system consists of the outflow tracts from above including the brainstem, spinal cord, nerve roots, joint proprioceptors and muscles.

Autism spectrum disorders (ASD) appear to have anatomic changes in the frontal lobes impacting the motor plan and attentional matrix, often causing problems with fine motor coordination, initiation of movement evidenced as 'impulsive' turns and transfers and repetitive patterns of movement with compulsive features (i.e. stereotypies and self-injurious behaviors) [1, 2] (Table 84.1).

Basal ganglia related disorders are most common in cerebral palsy (CP) as the lenticulostriate perforating vessels are vulnerable to ischemia from hypoperfusion, intracranial hemorrhage due to prematurity, and hydrocephalus from intraventricular bleeding at birth [3]. These injuries result in hemiplegia, hemidystonia, dystonic diplegia, spasticity and facial dyskinesias (see Table 84.1). Similar problems also occur more commonly in

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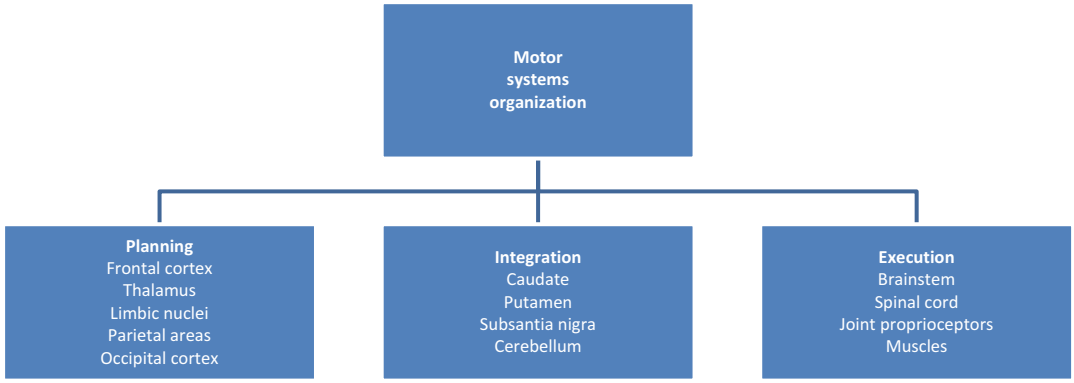

Fig. 84.1 Motor control is determined by three specific networks

Table 84.1 Definitions

Tremor- rhythmic oscillations
Rigidity- stiffness of limbs with active or passive movement
Chorea- dance-like nonpatterned movements
Dystonia- sustained or writhing postures of the trunk, limbs or head and neck, often one sided
Tics- twitches or vocalizations, briefly suppressible, associated with premonitory urges
Blepharospasm- involuntary forceful blinking
Orobuccolingual dyskinesia- darting, writhing tongue and lip movements often caused by medications
Ataxia- swaying trunk or limb movements "like a drunk"
Parkinsonism- shuffling gait, stiffness and slowness, tremors, flexed posture, balance problems
Stereotypies- repetitive purposeless hand movements, self-stimulating
Spasticity- tight resistance, legs > arms, worse with rapid movement

Table 84.2 Drugs implicated in causing movement disorders

Acute and tardive dyskinesias and akathisia:
Antiemetics- metochlopramide, droperidol, promethazine, prochlorperazine
SSRIs
TCAs
Carbamazepine
Lithium
Reserpine, alpha-methyldopa
Neuroleptics (typical and atypical)
Acute and tardive dystonia:
Antiemetics- droperidol, metochlopramide, promethazine, prochlorperazine
Amoxapine
Neuroleptics (typical and atypical)
Acute and tardive Parkinsonism:
Antiemetics-droperidol, metochlopramide, prochlorperazine, promethazine
Valproate
Reserpine, alpha-methyldopa
Amoxapine
Neuroleptics (typical and atypical)
Cinnarizine, flunarizine
Pimozide
Tetrabenazine

drug-induced movement disorders (see Table 84.2) due to increased serotonergic activity (SSRI's, SNRI's and TCA's) or dopaminergic blockade (neuroleptics, metoclopramide, valproate, antiarrhythmics, flunarizine, and more) in areas which are more sensitive to adverse effects due to aberrant neural networks developmentally [4, 5].

Cerebellar malformations (disordered cellular layers, atrophy or tonsillar elongation) result in problems with axial and limb control especially with transfers and walking. This is often evidenced as delayed gross motor development, balance problems and limb dysmetria on distal targeting. Cerebellar dysmorphism is especially common in genetic syndromes such as Velocardio-facial syndrome, Angelman's syndrome, Down syndrome, Joubert's syndrome, Dandy Walker malformations and Arnold-Chiari syndromes, Fragile X and more rare disorders [6].

Recognizing aspects of movement disorders requires a vocabulary to describe the appearance of such movements including parts of the body involved, aggravating and mitigating factors, developmentally normal movement and patterns of movement (see Table 84.1). Videotaping individuals (even a brief video on a smart phone) during examination or at home can allow more accurate assessment as parents or staff may describe movements in their own terms. There is often confusion between 'spasms' and 'seizures', 'twitches' and 'tremors' and 'spasticity' vs. 'rigidity'.

CW was a 23 year old male with ASD and mild IDD, partial complex seizure disorder, cystic acne and increasing SIB consisting of banging his head on walls and doorways, biting his hands and gouging his face. He had been moved from his family home to residential care at age 22 years and his parents were concerned that he was having more seizures because he appeared to have a lot of 'spasms' when they visited him. He was on valproate for seizure disorder and recently dextroamphetamine salts for possible ADD as he appeared to be distractible and fidgety.

On examination, he had frequent blinking, grimacing and vocalizations of grunts and clicking noises. Head flinging and fidgety movements with his fingers were also observed, especially when he was trying to walk on tiptoes. His valproate trough level was 85 and he had no signs of extrapyramidal tone or tremors. Staff who had been observing him closely since he joined the residence did not report any episodes of lapses of responsiveness or staring spells. He had exhibited increasing obsessional rituals before going to his day program and at bedtime but seemed to be adjusting well otherwise.

It was apparent that CW had simple motor and vocal tics in the setting of new exposure to stimulant therapy during a period of increased stress and adjustment difficulties. His behavioral profile along with more than 1 year of motor and vocal tics were consistent with a possible diagnosis of Tourettism but there was no documentation of tics prior to age 22 years to establish whether it was primary or secondary. Family members were reassured that he was not having more seizures. The neurologist observed at the visit that CW's father also had excessive blinking and nasal flaring while watching the examination, suggestive of a familial predisposition to tic disorder. It was recommended that the amphetamine salts be discontinued. Instead a behavioral plan was implemented to help with relaxation, lorazepam was used on a prn basis for stressful events such as doctor's visits and the family was educated about the nature of Tourettism and the possibility of the 'spasms' continuing long term.

Gait Disorders

Gait in particular is difficult to describe as it is involves multiple components including stance, static and dynamic balance, base and stride pattern, speed and cadence, steppage and turning pattern, truncal and limb posture and changes with 'stressed gait' if possible. Many individuals with IDD who appear to have a normal gait walking slowly will have problems when moving faster, walking on tiptoes or heels or attempting to tandem walk (heel to toe in a line) [7]. Acute changes in gait pattern can reflect problems in any part of the central or peripheral nervous system, including toxic encephalopathy, spinal injury, or pain from any source (even dental), joint subluxation, and peripheral nerve compression.

Common patterns of gait disorder in people with IDD include spastic steppage, ataxic gait, dystonic and parkinsonian gait. CP in particular predisposes to the first three patterns while drug therapy and age related changes cause parkinsonism. All of the above can result in falls especially with distraction, moving too quickly or too many obstacles in the walking path [8, 9]. In general, gait deterioration with advanced age is multifactorial and associated with joint and spinal deformity accelerated by unbalanced mechanical stress and often osteomalacia from medications.

Dystonic gait is usually action induced with unusual postures and twisting movements of the limbs occurring with weight bearing activity such as walking, lifting objects or with transfers. Parkinsonian gait is also more often associated with drug side effects and may evolve so slowly that it goes unrecognized until balance is compromised. Cardinal features are truncal stiffness (turning 'en bloc' as if the upper body is a single block that cannot move separately from the lower body), decreased arm swings and forward jutting posture of the head and neck rather than simply flexion of the neck. Rest tremor or pill-rolling, shuffling feet and propulsiveness are less frequent in drug induced Parkinsonism than idiopathic Parkinson's disease.

SC is a 45 year old male with a history of CP, mild ID, ASD, seizure disorder, and expressive language deficits who was living in a supported apartment setting but had a recent deterioration in his personal hygiene and was noted to be more irritable and aggressive with staff.

He started having tonic seizures as a newborn heralded by his earlobes turning blue. He had a bout of status epilepticus at 14 months and transient left hemiparesis followed by marked slowing of development. He was felt to have "intractable partial and generalized seizures" as he reported frequent lapses of awareness and multiple trips to the ED for head injuries after falling. Over the years he has been tried on many combinations of anticonvulsants and only had an observed reduction in his spells with vagal nerve stimulation. His EEGs showed right frontal focal epileptiform activity but he did not have videomonitoring. Over the past 2 years his antiepileptic medication (AED) regimen was carbamazepine, zonisamide, levatiracetam, chlorazepate and clonazepam.

When his behavior changed, he was given citalopram and later risperidone was added which helped 'calm him down' for a few weeks. Increasing the doses of both seemed to make him more irritable and he continued to have two episodes a month of falling down. He was never witnessed to have any tonic-clonic movements, urinary or bowel incontinence or tongue biting. On examination he had right hemispasticity, left hemidystonia with head tilt to the right and rotation left, poor balance with mild limb rigidity and asterixis and myoclonus in his outstretched hands. He also had dysmetria on finger to nose and impulsive targeting at end point as well as somnolence when not stimulated and bilateral extensor plantar responses. He was reported as taking frequent naps during the day, had gained 30 lb in the past 2 years and had not had any anticonvulsant levels done outside of the ED visits randomly.

He was diagnosed with a toxic encephalopathy superimposed on a static encephalopathy (CP) causing his falling episodes. He also was felt to have probable obstructive sleep apnea, drug -induced behavioral changes (levatiracetam, zonisamide and benzodiazepine) and mild Parkinsonism. Gradual elimination of the zonisamide and levatiracetam as well as the chlorazepate and risperidone resulted in substantial improvement in his behavior and balance over 4 months' time. He moved to a shared living situation where rare seizures consistent with his former tonic episodes were noted and were aborted by VNS activation. Weight loss and elevation of the head of his bed resolved the sleep apnea issue and improved his energy level in the daytime. His behavior reverted to his previous baseline of anxiety in novel situations but general compliance with caretaker's requests.

Age Related Changes in Motor Control

Gait problems from age related changes such as atlantoaxial instability in Down syndrome, spondylolithesis and spinal stenosis, multisensory deficits such as visual loss, vestibular problems from otoschlerosis or head trauma and dementia or Parkinsonism are all relatively common in this population and occur at a younger age than average individuals. Cumulative injuries over time also impact gait and can result in progressive

debility and nonambulatory status [10]. Physical and occupational therapy can improve overall function and advise regarding adaptive equipment that can help to maintain functional independence longer in many cases.

Most important in diagnosing age related change is identifying the individual's normal baseline by description of caretakers who have long term familiarity with the individual [11]. Often the deterioration appears to be sudden when there has actually been a gradual loss of function reaching a critical threshold where the ability to transfer, stand or walk has been lost. It is not uncommon for the earlier changes to be attributed to 'bad behavior' due to refusals to do routine tasks. Addition of medication to treat the behavior may contribute to more rapid decline in gait if they are sedating, impact basal ganglia function or cause drug-drug interactions with other prescribed regimens.

Drug Induced Movement Disorders

Drug induced movement disorders are very common among people with IDD due to lack of brain plasticity in response to changes in the neurochemical environment (see Table 84.2). 'Paradoxical' responses to medication, such as hyperactivity from phenobarbital and increased anxiety from SSRIs are often predictable based on the individual's neural network abnormalities. Data relating to psychotropic medication side effects and benefits in people with IDD is very limited in clinical literature and most is based on small study groups or short term observations [12]. Often the initial effect of a psychotropic drug is sedation which may appear to improve the behavioral problems being targeted. Over several half-lives of the drug (usually 1–2 weeks), that effect is lost and behaviors reemerge, prompting an increase in the dose. Eventually undesirable side effects become apparent and result in new behaviors that are problematic. Unfortunately, abrupt discontinuation of these drugs can cause significant 'emergent withdrawal' symptoms (EWS) akin to neuroleptic malignant syndrome (see Table 84.3). It may take months for EWS to resolve on its own, while prompt reinstitution of the offending drug and very gradual tapering off is better tolerated.

Table 84.3 Neuroleptic malignant syndrome diagnostic criteria: Expert panel consensus

Diagnostic criterion	Priority score[a]
Exposure to dopamine antagonist, or dopamine agonist withdrawal, within past 72 h	20
Hyperthermia (>100.4 °F or >38.0 °C on at least 2 occasions, measured orally)	18
Rigidity	17
Mental status alteration (reduced or fluctuating level of consciousness)	13
Creatine kinase elevation (at least 4 times the upper limit of normal)	10
Sympathetic nervous system lability, defined as at least 2 of the following:	
Blood pressure elevation (systolic or diastolic ≥25 % above baseline)	
Blood pressure fluctuation (≥20 mmHg diastolic change or ≥25 mmHg systolic change within 24 h)	
Diaphoresis	
Urinary incontinence	
Hypermetabolism, defined as heart-rate increase (≥25 % above baseline) AND respiratory-rate increase (≥50 % above baseline)	5
Negative work-up for infectious, toxic, metabolic, or neurologic causes	7
	Total: 100

[a]The mean priority score indexes each criterion according to its relative importance in making a diagnosis of neuroleptic malignant syndrome according to an expert panel Gurrera et al. [13]. Copyright © 2011, Physicians Postgraduate Press. (need to obtain permission for this text.)
Abnormal Involuntary Movement Scale
Munetz and Benjamen [14]

AT is a 30 year old female with Angelman's syndrome living in a group home with moderate assistance for most ADLs, mild gait ataxia and anxiety disorder. She was on carbamaze-

pine for generalized tonic-clonic seizure disorder and recently put on olanzapine for anxiety during transport to her day program. She began exhibiting strange behavior over the past 2 months, "refusing to walk" and bumping around on her buttocks to get around. The olanzapine was gradually increased to 10 mg a day but her behavior did not improve and she started refusing to eat, screaming and sleeping poorly. Her seizures did not increase but she was steadily losing weight and appeared distraught. She was admitted to a dedicated inpatient unit for individuals with IDD where she could be medically evaluated.

On admission she was restless, unable to maintain an upright posture in a chair and had urinary retention, mild spasticity in the legs and general tremulousness. She was not able to follow directions verbally or visually and had a low grade fever. Her CPK was 18,000 attributed by the admitting house officer to bruising on her buttocks from bumping around on the floor. After being examined by neurology, she was diagnosed with neuroleptic malignant syndrome, transferred to the ICU and olanzapine was discontinued and lorazepam introduced for agitation. Her CPK levels peaked at 56,000 and she had acute tubular renal failure due to rhabdomyolysis which resolved with supportive care. After 6 weeks she was discharged back to the group home and gradually weaned off lorazepam. She went home after 4 months, ambulatory and able to communicate with gestures again as well as eating a soft diet and regaining weight.

Table 84.4 Classification of tics

Physiologic tics
Mannerisms
Pathologic tics
Primary
Sporadic
Transient motor or phonic tics (<1 year)
Chronic motor or phonic tics (>1 year)
Adult-onset (recurrent) tics
Tourette's syndrome
Inherited
Tourette's syndrome
Huntington's disease
Primary dystonia
Neuroacanthocytosis
Secondary ("Tourettism")
Infections: Encephalitis, Creutzfeldt-Jakob disease, Sydenham chorea
Drugs: Stimulants, levodopa, carbamazepine, phenytoin, phenobarbital, antipsychotics (tardive tics)
Toxins: Carbon monoxide
Developmental: Static encephalopathy, mental retardation syndromes, chromosomal abnormalities
Other: Head trauma, stroke, neurocutaneous syndromes, chromosomal abnormalities, schizophrenia, neuroacanthocytosis degenerative disorders
Related disorders
Stereotypies
Self-injurious behaviors
Hyperactivity syndrome
Compulsions
Excessive startle
Jumping Frenchman, Latah, Myriachit

Adapted from: Jankovic [20] (need to obtain permission for reprint from Up to Date.)

The types of drug induced movements that occur in this population are similar to what is observed with chronic therapy of schizophrenia, psychotic depression, bipolar disorder and advanced Alzheimer's dementia. Orobuccolingual dyskinesias and 'rabbit mouth' (upper lip quivering) are seen with neuroleptics including the atypicals such as olanzipine, risperidone and paliperidone [15–17]. Cervical dystonia is more common with neuroleptics in young males while high doses of SSRIs may produce this pattern in young women. Tremor in the limbs is seen with valproate, lithium and SNRIs, stimulants and phenytoin, which can merge with extrapyramidal signs as the doses are increased or exposure time lengthens [18, 19]. Myoclonus and tics are more common with stimulants, but also occur with toxic levels of numerous anticonvulsants, especially the epoxide of carbamazepine, free phenytoin, felbamate and lacosamide (see Table 84.4). At baseline,

Table 84.5 Pharmacologic management of Tourette's syndrome

Treatment of tics:	
1.	Clonidine- 0.1, 0.2, 0.3 mg ½-1 tablet bid or tid, later converting to 0.1, 0.2, 0.3 mg patch q5 to 7 days if tolerated
	Side effects sedation, hypotension, bradycardia, headache, insomnia, skin rash, minor ECG changes
2.	Clonazepam- 0.5–1.0 mg tablets ½-1 qhs or bid
	Side effects sedation, imbalance, dependence and tolerance, diarrhea
3.	Pimozide- 2 mg tablets ½-1 po bid to tid
	Side effects sedation, weight gain or QT interval changes, requires ECG monitoring
4.	Guanfacine- 1, 2 mg tablets ½-1 po bid to tid; 1, 2, 3, 4 mg ER tablets 1 poqd
	Side effects sedation, hypotension, GI distress
5.	Aripiprazole- 5, 10, 15, 20, 30 mg tablets ½-1 po qd to start, also available in IM, liquid, depo form
	Side effects sedation, headache, weight gain, anxiety, agitation, EPS
6.	Risperidone- 0.25, 0.5, 1, 2, 3, 4 mg tablets, also po solution, ODT, depo forms
	Side effects weight gain, sedation, ECG changes, EPS and dystonia, cognitive blunting, impotence

Adjunctive therapy for ADHD, OCD, anxiety and depression per usual pharmacotherapy guidelines, with attention to drug-drug interactions and additive side effects

there is a significant proportion of individuals with IDD who evidence involuntary movements without exposure to any of the aforementioned drugs. The relevance of determining whether the movements are drug induced or not lies in their impact on ADLs and the potential for worsening over time with long term exposure to the drugs (i.e. become 'tardive') [21]. As with gait disorders, it is helpful to establish whether there are any involuntary movements before initiating a CNS acting medication using either standardised measurement scales (see Table 84.5) or video recording (AIMS exam).

Dysphagia, dysarthria and akathisia are movement disorders infrequently identified as adverse effects of drug therapy as they may be present at baseline and worsen gradually after introduction of many classes of drugs, especially neuroleptics. Dysphagia often accompanies other signs of drug induced Parkinsonism while dysarthria is seen with drugs that cause sedation, dry mouth, cerebellar signs and Parkinsonism. Articulation changes can aggravate swallowing problems and swallowing problems similarly can result in excessive salivary pooling, drooling and slurred speech. Akathisia, on the other hand, may appear to be anxiety driven behaviors such as rocking, pacing, hyperactivity with purposeless repetitive gestures (flapping, clapping, spinning, crossing and uncrossing the legs repetitively, etc.) and worsen with interruption or increases in the offending drug. Not infrequently, these movements are misinterpreted as 'behaviors' that warrant medication adjustment, creating a cycle of drug induced adverse effects.

Stereotypies, Tics and Movement Mimics

Specific types of unusual movements are more characteristic of individuals with IDD than in typically developing persons with psychiatric or dementing illnesses [22]. These include repetitive purposeless, often self-stimulatory gestures and patterned movements variably labeled as stereotypies or obsessional rituals. The former are seen most often in ASD in early childhood and the latter in older children/young adults. As seen in the case of akathisia, the 'driven' quality of the movements can only be ascertained indirectly by attempting to interrupt or suppress them [10]. Often the stereotypy will simply resume when the distraction is gone whereas the obsessional ritual will become more intense in an effort to correct the disturbance and 'fix' the sequence of events. Obsessional slowness reported in Down Syndrome may be a form of bradykinetic movement disorder but is not associated with extrapyramidal tone [23].

Pisa syndrome is a form of axial dystonia induced by neuroleptics that appears as if the individual is purposely and progressively leaning backwards as they walk, making balance precarious. It may occur acutely on exposure or with increasing doses of the offending drug but is often hard to resolve even with drug discontinuation. Camptocormia is just the reverse as progressive truncal flexion occurs on walking, often to

one side. Both forms of truncal dystonia are more likely to occur in people with severe IDD or elderly persons with IDD and be associated with spinal deformity over time [11]. Surgical correction of the spine does not result in improvement of the abnormal posturing on walking and can lead to significant pain issues afterwards. Treatment with anticholinergics (benztropine, trihexylphenidyl) has occasionally been reported as helpful. Sensory tricks such as holding hands in front of the Pisa subject or using a tall walker for the camptocormia subject have also been reported as partially effective in overcoming the dystonia [12]. Fear of falling can make the individual resistant to such interventions however.

Tourettism (TS) and the spectrum of tic disorders are variably classified as an involuntary movement disorder, an 'involuntary'/psychiatric disorder or a behavioral/psychiatric disorder depending on one's point of view [24]. While diagnostic criteria have been defined in the DSM-IV (see Table 84.4), the impact on functioning and the suppressibility of the tics is hard to define in most ID/DD individuals. The fact that it is more commonly seen as a bilineal inherited disorder in families with a male preponderance suggests a strong biological basis for Tourette's syndrome. Comorbid conditions such as Attention Deficit Hyperactivity Disorder (ADHD), bipolar disorder, Obsessive Compulsive Disorder (OCD), anxiety and depression, learning disability and conduct disorder may be implicated in the same neural networks that appear to cause the tic disorder [24].

Tourette Syndrome (TS) has been conceptualized as a developmental expression of underlying aberrant dopaminergic pathways. TS has been demonstrated to be associated with loss of grey matter volume in the left caudate, decreased D2 receptor density and decreased inhibitory activity of the limbic system. The typical presentation is between ages 5 and 14, occurs 4 times more frequently in boys than girls, and fluctuates in regards to how often the tics appear each day. Patterns of tics, or the 'repertoire' each individual develops, may be changeable or cumulative with periods of apparent remission and a gradual increase in the suppressibility of the tics as the individual gets older. Often tics appear to dissipate in adult years though comorbid behavioral and mood issues may become more prominent.

Social disability is a greater concern in elementary school years and prompts intervention with medications but may not be required long term. It is unusual to see evidence of drug induced extrapyramidal effects with neuroleptics in TS but impacts on learning can limit their utility. The discontinuation syndrome from weaning off neuroleptics may result in transient worsening of tics for several months. The choice of drug therapies is dependent on how disabling the tics are and whether there is accompanying ADHD, OCD or behavioral problems [21]. Nonpharmacologic treatments such as habit reversal training, supportive counseling and environmental modification, TS support groups and education of parents, teachers and peers complement drug therapy and may be all that is needed for mild Tourettism [25].

Other movement 'mimics' include seizures with myoclonus or nocturnal dystonia, self-injurious behavior (SIB) and compulsive habits such as hair twirling, pencil chewing and skin picking. The motor changes seen in neuroleptic malignant syndrome (NMS) are the major mimic of concern, as it can be life threatening if not recognized early. In people with IDD, the first signs may not be dysautonomia but rather a general deterioration in level of arousal or agitation and behavioral decompensation (see case of AT above). Clinical biomarkers, such as elevated WBC count, increased CPK and electrolyte imbalances as well as hyperthermia and dehydration, are typically seen within the first few days of NMS emergence but may be delayed by days to weeks after introduction or augmentation of the offending neuroleptic [26]. Serotonergic signs such as diarrhea, nausea and vomiting may suggest gastroenteritis or an acute bowel syndrome rather than serotonin syndrome (SS) which closely resembles NMS [27]. A high index of suspicion and prompt medical intervention have resulted in lower morbidity and mortality in NMS and SS in recent years.

Other Interventions for Movement Disorders

Botulinum toxin injections (BOTOX), Deep Brain Stimulator therapy (DBS) and intrathecal baclofen pump therapy (ITB) can improve motor control with fewer CNS side effects than seen with oral pharmacotherapy. Botulinum toxin is very effective for severe lower extremity spasticity, segmental dystonia and focal tics, as well as contractures prior to release surgery, allowing relaxation of the muscles long enough for the tendons and muscles involved to heal [28]. Unfortunately, the Botulinum wears off in 12–15 weeks requiring frequent painful injection protocols and often sedatives or topical analgesics prior to the procedure each time.

Deep Brain Stimulator implants in the thalamus or globus pallidus have been recently trialed for CP, dystonic storms associated with metabolic disorders and drug withdrawal, disabling tremors and severe opisthotonus [29]. Results are modest with 30–50 % improvement of the postures overall and complications such as device infection, lead fracture and frequent battery replacements may limit its utility [30].

Intrathecal baclofen therapy (ITB) is also effective for spastic diplegia, dystonic diplegia and spastic paraparesis in CP with long term benefit being well established after >20 years of study [31]. The intrathecal catheter is threaded from the lumbar level to the upper thoracic level, in most cases, and titration of the dose is gauged by reduction in spasticity or dystonia without emergence of urinary retention, loss of truncal control or severe limb weakness. As pump technology has improved, the period of time between reservoir refills has increased along with introduction of compounded solutions such as baclofen with morphine or diazepam.

Finally, physical and occupational therapy as well as orthopedic consultation, behavioral interventions and environmental modifications all contribute to the success of treatment for movement problems. Access to a dedicated team of IDD specialists may be limited, but there are many online resources and instructional videos that help with outreach to remote areas.

Summary

Movement disorders are very common among individuals with IDD, both primary and secondary types, and can result in functional disability and behavioral deterioration in a significant number of them. Simple tools such as video recording of the individual's gait and movements before medications are introduced, as well as accurate descriptions of the movements using consistent terminology can be invaluable in assessing any adverse events later. Treatment of movement disorders is directed towards improving overall function without adding drug side effects. This requires reassessment at regular intervals by a clinician familiar with the individual long term. While the movements are often not eliminated they can be managed using a multidisciplinary approach including neurology, developmental psychology, orthopedics, physical and occupational therapy, pediatrics and neurosurgery.

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