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Neurologic Conditions in Individuals with Intellectual Disability

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Abstract

Individuals with intellectual disabilities (ID) frequently have co-occurring neurologic conditions, which are more likely to be complex in nature when compared to the general population. These individuals are more likely to suffer from conditions such as seizure disorders, attention-deficit/hyperactive disorder, autism spectrum disorder, dementia, delirium, and motor disorders (e.g., tics, antipsychotic-induced). ID is commonly comorbid with other neurodevelopmental disorders, which often present in early developmental years. Sometimes ID is associated with the development of neurocognitive disorders later in life. ID may be associated with a known environmental factor (e.g. prenatal alcohol exposure) or genetic condition that can predispose to other neurologic problems (e.g., Down Syndrome is a risk factor for Alzheimer's dementia). Neurologic disorders can be difficult to diagnose in those with ID due to barriers in communication and decreased self-report. Because these disorders increase impairment of personal, social, academic or occupational functioning, early identification and intervention can help clinicians improve treatment and outcomes.

Keywords: Intellectual Disability; Neurology; Cognitive; Delirium; Encephalopathy

Seizure Disorders

Individuals with ID suffer from epilepsy up to twenty times that of the general population. For example, among those with cerebral palsy, about 30% have ID and, of those, about 40% have seizures. Additionally, Angelman syndrome, homocystinuria, Krabbes's disease, Lesch-Nyhan syndrome, Lennox-Gastaut syndrome, neurofibromatosis, phenylketonuria, Rett syndrome, Sturge-Weber syndrome, and tuberous sclerosis are also associated with both ID and seizures [1,2]. The prevalence of seizures increases with the severity of ID and nearly one half of individuals with severe ID suffer from seizures [3]. Up to one third of individuals with ID do not develop seizures until adolescence or early adulthood; seizure are more likely to be treatment resistant in ID [12]. Psychogenic non-epileptic seizures are more common in those with ID and epilepsy than the general population. Non-epileptic seizure-like activity may be due to transient ischemic attack (TIA), complicated migraine, syncope, hypoglycemia, narcolepsy, myoclonus, or conversion disorder. Of note, individuals with ASD who do not suffer from seizures may show abnormal rhythms and other nonspecific changes on electroencephalogram. Complex partial seizures are the most common seizures among patients with ID and ASD.

Dementia

A neurocognitive disorder—particularly a slow, progressive dementia—may be difficult to diagnose in those with ID due to overlapping symptoms, but it should be suspected if there is a decline from a previously obtained level. Many individuals with ID are at increased risk for cognitive decline; many psychologists do not have specific training and experience in neuropsychological testing in patients with ID. For example, Trisomy 21 is associated with early-onset Alzheimer's dementia—likely due to the amyloid precursor protein gene located on chromosome 21—with up to 77% of individuals with Down syndrome developing Alzheimer's, and up to 55% developing it before the age of 50 [4]. There is no cure for dementia, though lowering stroke risk factors (e.g., diabetes, hypertension, hyperlipidemia, smoking) may help prevent dementia, and acetylcholine esterase inhibitors and memantine may slow the rate of decline.

Delirium/Toxic Metabolic Encephalopathy

Individuals with ID—particularly severe and profound ID—are at increased risk of developing delirium [5]. Common underlying causes of delirium among those with ID are summarized in Table 1. Electroencephalography (EEG) often shows generalized slowing, or less commonly, abnormally fast activity (NADD, 2016). Individuals with ID may experience hyper- or hypo-active delirium, though medication-induced delirium is rarely hyperactive [5]. Unfortunately, delirium in this population is more likely to go undiagnosed resulting in delayed treatment. Reasons for this may be misattributing cognitive or behavioral changes to ID, and/or difficulty for those with ID to effectively communicate symptoms they experience. While ID and delirium may both involve disturbances in attention and cognition, a key distinction from ID is that delirium involves a disturbance in awareness (i.e., reduced orientation to the environment). Furthermore, these disturbances represent a change from baseline,

Table 1: Common underlying causes of delirium in the ID population

Infection
• Respiratory tract
• Urinary tract
Medications
• Anticholinergics
• Antipsychotics
• Anticonvulsants
Electrolyte imbalances
Dehydration
Fecal impaction

and tend to fluctuate in severity during the course of the day [6]. Patients with delirium are at increased risk of falls, longer hospital stays, and increased cognitive decline and increased mortality rates during the twelve months following hospitalization [7].

Treatment of Delirium

Definitive treatment of delirium requires identification of the etiology. However, this is not always immediately identifiable. Additional objective data is useful in this patient population due to decreased subjective information because of communication difficulties. Once the underlying cause is known, treatment involves reversing or addressing this underlying cause. Regardless of the cause, medications such as sedatives or anticholinergic medications should be used only with extreme caution in ID during the course of delirium.

Antipsychotics historically been the treatment of choice for agitation associated with delirium. It is important to explain to staff members and family members that the goal of treatment with antipsychotics in a delirious patient is not simply to sedate the patient, but rather to prevent the individual from harming themselves or others. Current thinking is that they should be avoided if it is possible to keep the patient safe without their use. "As needed" antipsychotics should not be administered for convenience but rather for dangerousness. Haloperidol is the most commonly utilized agent when managing hyperactive delirium. Acute delirium with agitation generally requires parental rather than oral antipsychotics, and haloperidol has both intravenous (IV) and intramuscular (IM) routes of administration. IV is preferable to IM haloperidol as it allows for more reliable absorption and, when an IV lock is present, may decrease the likelihood of causing distress to a delirious patient that might be confused or paranoid [7]. It is appropriate to monitor for QT prolongation with EKG or cardiac monitor, and monitor vital signs and mental status regularly.

Environmental measures are also important for delirium-related agitation or "sundowning." Table 2 summarizes commonly used environmental measures.

Antipsychotic-induced Movement Disorders

Individuals with ID are at an increased risk of developing medication-induced extrapyramidal symptoms due to their sensitive central nervous system; this is especially the case if the patient has underlying muscular conditions such a cerebral palsy [3]. The four common extrapyramidal adverse effects of antipsychotics include acute dystonia, akathisia, parkinsonism, and tardive dyskinesia (see Table 3). The side effects may be treated by tapering and discontinuing the antipsychotic, and switching to another agent (e.g., from a potent first generation to a second-generation antipsychotic). However, in some cases (e.g., comorbid psychotic or bipolar disorders, ineffectiveness or worse adverse effects with other agents), continuing the agent and starting another agent to manage adverse effects may be appropriate.

Table 2: Environmental measures for delirium-related agitation

Orientation
• Place clocks and calendars in sight
• Open curtains during daylight
• Dim lights at night
• Frequently orient the patient to the time of day
Comfort
• Ask caregivers to bring treasured objects from home such as a favorite blanket or pictures of loved ones
• Encourage caregivers to visit frequently during the daytime
• Closely monitor and treat issues such as constipation or pain
Avoid over-stimulation
Minimize disruptions in nighttime sleep

Table 3: Antipsychotic-induced extrapyramidal symptoms

	Characteristics	Onset	Treatment
Acute Dystonia	Muscle spasm or stiffness (e.g., torticollis, trismus), tongue protrusions, oculogyric crisis	Usually occurs within the first hours or days of treatment and is most common in young males	Diphenhydramine, anticholinergics (benztropine, trihexyphenidyl)
Akathisia	Subjective feeling of restlessness and is in constant motion, unable to sit still, pacing, alternating sitting and standing	Usually occurs within the first few days of treatment. Most commonly occurs with the atypical antipsychotic Aripiprazole	Propranolol, benzodiazepines
Parkinsonism	Stiffness, cogwheel rigidity, shuffling gait, mask-like facies and is most common in elderly females	Usually occurs within the first few months of treatment, related to antipsychotic-induced dopamine depletion	Diphenhydramine, anticholinergics (benztropine, trihexyphenidyl)
Tardive Dyskinesia	Perioral movements (darting or protruding movements of the tongue, chewing, grimacing, puckering). choreoathetosis of the head, limbs, trunk.	Usually presents after years of treatment	Valbenazine, deutetrabenazine, some antipsychotics may suppress symptoms (e.g., quetiapine, clozapine)

ADHD

ADHD is three times more prevalent in children with ID, three to five times more common in children with comorbid ID and epilepsy, and more common among adults with ID (NADD, 2016). Fetal alcohol syndrome, Fragile X syndrome and DiGeorge syndrome are each associated with both ID and ADHD. The inattentive subtype of ADHD is more common in ID patients; however, those with hyperactive/impulsive type have worse outcomes and are more likely to suffer from conduct disorder, or oppositional defiant disorder (NADD, 2016) [8]. Mood dysregulation and aggression are common among those with comorbid ADHD and ID, and these individuals are more likely to be misdiagnosed with bipolar disorder (NADD, 2016) [8]. Likewise, to ensure accuracy of ADHD diagnosis, it is important to assess individuals' academic performance and community integration in comparison to neurotypical peers (rather than chronological age). According to the DM-ID-2 (NADD, 2016) [8], the ADHD diagnostic criteria adapted for persons with ID states that both inattentive and hyperactive/impulsive symptoms must persist to a degree that causes a direct negative impact and is inconsistent with developmental level.

Treatment of ADHD

Stimulants including methylphenidate or amphetamines are considered first-line treatments for ADHD. Stimulants are generally well tolerated but adverse effects can include anorexia, nausea, insomnia, anxiety, irritability, and hallucinations. It is important to note that stimulants may induce or worsen tics therefore caution should be used in individuals with comorbid ADHD and tic disorders. Common non-stimulant medications used in the treatment of ADHD include atomoxetine, bupropion, and alpha-2 agonists (i.e., clonidine, guanfacine). Behavioral interventions for ADHD include individualized education plans, psychotherapy, skills training, exercise, and parent education and training [9-11].

Tic Disorders

Tic disorders are common among individuals with ID (NADD, 2016). Individuals with tic disorders display motor or vocal tics, which are sudden, rapid, recurrent, nonrhythmic, stereotyped motor movements or vocalizations. Tourette's disorder commonly presents with comorbid obsessive-compulsive disorder, ADHD, self-injurious behaviors, anxiety disorders, and depressive disorders (NADD, 2016). The DM-ID-2 did not suggest adaptations for individuals with ID regarding Tourette's disorder, persistent (chronic) motor or vocal tic disorder, and provisional

tic disorder. However, DSM-5 noted "age of recognition and duration may be difficult to establish, especially if there are multiple genetic, neurodevelopmental, or medical conditions that may not be a direct contribution to tics (eg. tics in individuals with Fragile X or trisomy 21)" (APA, 2013).

Treatment of Tic Disorders

Second generation antipsychotics have been commonly used for the treatment of Tourette's Disorder, though have limited efficacy for common comorbid disorders such as ADHD and OCD. Alpha-2 agonists are effective in decreasing both the severity and frequency of tics and also treat ADHD.

Summary

Neurologic conditions and intellectual disability are closely intertwined and frequently co-occur. Communication limitations and decreased self-report in many patients with ID may complicate the assessment and diagnostic process. Until the existence of best practices and evidence based medicine principles specific to ID, clinicians should utilize treatment interventions based in science for the general population with added caution and conservatism.

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