Psychiatric Assessment of Severe Presentations in Autism Spectrum Disorders and Intellectual Disability

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KEYWORDS

• Autism • Intellectual disability • Self-injury • Aggression • Hyperactivity • Psychiatric evaluation

KEY POINTS

- Psychiatric illnesses are common in autism spectrum disorder (ASD)/intellectual disability (ID).
- Externalizing behaviors are common presenting symptoms but are etiologically nonspecific.
- Genetic conditions associated with ASD/ID may inform medical surveillance as well as potential therapeutics.
- Co-occurring medical conditions are common in ASD/ID and may contribute to symptom presentation.
- Environmental factors, for example, change in caregiver or experience of trauma, may be particularly significant in the setting of ASD/ID.

INTRODUCTION

Decades ago, Sovner and Hurley[1] somewhat rhetorically debated whether individuals with ID experience affective illness. Although the answer then as now is an unequivocal yes, uncertainty does remain as to how the presentation of psychiatric

Disclosure Statement: B.H. King has received research funding and has served as a consultant for Seaside Therapeutics and Roche. Drs N. de Lacy and M. Siegel report no financial disclosures.

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Child Adolesc Psychiatric Clin N Am 23 (2014) 1–14
http://dx.doi.org/10.1016/j.chc.2013.07.001 **childpsych.theclinics.com**
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disorders may be altered in the setting of atypical intellectual development. Moreover, as the genetic underpinnings of neuropsychiatric illness are revealed and certain behavioral phenotypes elaborated in association with particular genetic abnormalities, questions have been raised about the appropriateness of applying nonspecific illness labels to behaviors that occur in specific contexts. In cases of hyperphagia and restricted interests in Prader-Willi syndrome, for example, is value added by superimposing the psychiatric diagnoses of impulse control disorder or obsessive-compulsive disorder?

Conversely, there is every bit as much heterogeneity in symptom expression in the context of genetic syndromes as for the general population. Not everyone with Prader-Willi syndrome has significant skin-picking behavior nor does Lesch-Nyhan syndrome guarantee aggression, although these are common symptom-syndrome correlations. Taking a step back, the same can be said for persons with idiopathic ID: not everyone with severe ID is aggressive, self-injurious, or hyperactive. These problems occur only in a minority of this population.

Howe[2] observed that "there are some among the lowest class of idiots who seem to have a superabundance of innervation, who are consequently very active. They appear like insane persons in a state of excitement, and yet they have no speech, and no reasoning faculties."[2] Hurd,[3] who was superintendent of the Eastern Michigan Asylum, wrote that irritability, violence, and impulsivity alone are insufficient grounds for the diagnosis of insanity in this "lowest grade of imbeciles," but impulsive acts, "morbid propensities," and "acts of suicidal intent" (eg, "attempting to dash one's brains"), "occurring in higher grades of imbecility" are symptoms consistent with "actual insanity," even in the absence of delusions.

The recognition that even severe ID neither protects nor precludes an individual from experiencing psychiatric illness is thus one of the earliest observations from clinicians working in this field. Modern studies generally estimate that having ID increases the risk of psychiatric illness at least 3-fold or 4-fold relative to the general population,[4,5] thereby underscoring the importance of careful psychiatric assessment in this population.

PREVALENCE OF PSYCHIATRIC ILLNESS

Estimates of the overall prevalence of psychiatric disorders in individuals with ID range from 10% to 39%.[6,7] In children and adolescents, emotional and behavioral problems occur up to 7 times more frequently than in typically developing youth.[8]

Specific factors that place individuals with ID at increased risk for developing comorbid psychopathology include severity of disability, lower adaptive behavior, language impairments, poor socialization, low socioeconomic status, and families with only 1 biologic parent.[9] In general, developmental and genetic disorders are associated with elevated rates of depression and anxiety. Specific genetic syndromes are also associated with increased rates of particular disorders, such as higher rates of depression in Down syndrome (DS),[10] anxiety, and ADHD in individuals with Williams syndrome and increased rates of schizophrenia in velocardiofacial syndrome (22q11.2 deletion syndrome).[11,12]

Psychiatric illness is also clearly more prevalent in the ASD population than in the general population. Prevalence estimates vary based on the type of measure used. When diagnostic instruments developed for the neurotypical population are used for individuals with ASD, high rates of comorbid psychiatric illness are found. Using the Kiddie Schedule for Affective Disorders and Schizophrenia (K-SADS) for School-Age Children,[13] in a psychiatrically referred population, produced an estimate of

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