Problems and Questions in the Dual Diagnosis of Schizophrenia and Intellectual Disability

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Although the Diagnostic and Statistical Manual of Mental Disorders, Fourth Edition (DSM-IV)³ defines intellectual disability as the global cognitive impairment of an individual that begins before age 18. DSM-IV designates cognitive impairments that occur or worsen after 18, “dementias.” There are numerous classification systems for the diagnosis of intellectual disability, including DSM, ICD-10, and AAMR. These all consider standardized testing but also emphasize to varying degrees the concurrent evaluation of the patient’s adaptive functioning. The latter is related but not identical to intellectual ability in standardized tests. DSM-IV labels those below the third percentile, equivalent to an IQ of 70 on individually administered tests, as having intellectual disability. This implies that 3% of the population has intellectual disability, but intellectual disability is actually much less prevalent because of altered survival rates among those with extremely low IQ. Though some investigators have reported higher rates of intellectual disability amongst males, this finding may be a product of male persons’ requiring more services.¹⁹,²⁸

DSM-IV divides intellectual disability into levels of impairment: mild, moderate, severe, and profound. It also requires that the person have at least two impaired domains amongst communication, self-care, home living, social/interpersonal skills, use of community resources, self-direction, functional academic skills, work, leisure, health, or safety. The diagnosis of intellectual disability does not exclude other diagnoses, including dementia. Were the person with intellectual disability to experience declines before the age of 18 from previous global intellectual functioning, he or she would receive both the diagnosis of intellectual disability and that of dementia. However, if the person experienced all his deficits after the age of 18, then in the DSM-IV system he or she would only receive the diagnosis of dementia. Furthermore, intellectual disability does not exclude the diagnosis of a developmental disorder such as autistic disorder (75% of autistic individuals have intellectual disability, for example). Although the cause of intellectual disability is often multi-factorial, the clinician can identify a probable etiology in many patients. The likelihood of identifying a cause is proportional to the severity of the intellectual disability: the clinician identifies the condition’s etiology in 80% of patients with severe intellectual disability, but in only 25-30% of those with mild intellectual disability.¹⁹

Schizophrenia is a syndrome defined by symptoms and history rather than any known pathophysiology: thus, its diagnostic framework resembles that of intellectual disability. DSM-IV requires that the patient have one month of acute symptoms and six months of residual symptoms. The acute symptoms must include at least two amongst delusions (fixed false, culturally and developmentally inappropriate ideas), hallucinations (false perceptions with impaired reality testing), disorganized speech, disorganized behavior, or negative symptoms like flattening of affect, alogia, or avolition. Residual symptoms are impairments in interpersonal relations, work, or self-care. A large number of different presentations fit this definition, and DSM-IV divides the disorder into subtypes, which include paranoid, disorganized, catatonic,
undifferentiated, and residual. At this time, there is no clear evidence that any known factors influence the occurrence of one or another subtype, but persons with the disorganized subtype have a worse prognosis than the paranoid one and some have associated it with a higher genetic load (as measured by illness in family members). Though cognitive dysfunction is frequent in persons with schizophrenia, it disproportionately reflects impairments in executive function. Decrement in global cognitive functioning accompany severe schizophrenia however, and in this sense, schizophrenia is a dementing process. There is extensive literature on possible pathophysiologies of schizophrenia, but there is no identified etiology and there may, in fact, be a spectrum of schizophrenias or different types that are unrelated. Presently, many researchers have found evidence that schizophrenia resembles a neurodevelopmental disorder.30 Evidence for this view includes events in the second trimester of development that correlate with eventual schizophrenia, schizophrenic individuals’ tendency to minor physical anomalies, typical age of onset of schizophrenia in the second decade when the brain is at the peak of re-organization, and absence of neurodegenerative disease in deceased affected individuals. The most commonly reported neuroanatomical finding has been varying degrees of ventricular dilatation and widened cortical sulci in those people affected, particularly left-right asymmetry. Many investigators attribute this to reduced temporal lobe structures. Some also suggest that illness severity correlates with the extent of this non-progressive hydrocephalus ex vacuo.5,19,25

The diagnoses of schizophrenia and intellectual disability are not technically mutually exclusive in DSM-IV. Not surprisingly therefore, clinicians diagnose many patients with both syndromes. However, because the diagnosis of schizophrenia depends in large part on adequate communication with the patient and an accurate history, the identification of these two separate syndromes in any one patient is idiosyncratic.56 For example, although there is no logical reason why the pathophysiology that leads to schizophrenia should not be possible in patients with severe intellectual disability, many clinicians find it impossible to identify the required symptoms in this population because patients with intellectual disability have impaired communication skills.41 Adequate history is essential to satisfy the differing history requirements for each of the diagnoses, and while theoretically this should be possible, in fact persons with intellectual disability frequently present without accurate histories of their specific symptoms.20,54

Jablensky14 represented the prevalence of schizophrenia in the general population as .4%. Numerous (but not all) surveys suggest a prevalence of schizophrenia in patients with intellectual disability to be 3%, which is greater than random assortment of the two conditions would suggest.38,56 No one knows why schizophrenia appears to occur more frequently in patients with intellectual disability than in the general population. The heterogeneity of the disorder’s past diagnosis may be one explanation. For example, no community survey separately evaluated the different subtypes of schizophrenia.9,12,18,29,56 This is problematic because the reliability of the diagnosis of schizophrenia in intellectual disability might differ according to the apparent subtype, because of the poor separation of undifferentiated and disorganized subtypes’ symptoms from intellectual disability-associated behaviors. Most surveys also did not attempt to differentiate autistic symptoms from psychotic ones.10,12,18,20,34,56 Yet fragile X and other common intellectual disability etiologies are disproportionately associated with autistic symptoms. An example of the importance of this oversight is that Lund29 found much lower schizophrenia prevalence when he separated out “atypical psychosis” in his survey, a category that included persons with autistic symptoms but not the complete autism syndrome. Doody12 did not separate out the various sub-types of schizophrenia either. On the contrary, he found more negative symptoms in his co-morbid group, and more positive symptoms in his group with schizophrenia alone. This is exactly what one would predict if he had misidentified the autistic symptoms of intellectual disability as the negative symptoms of schizophrenia. Therefore, there may be a group of putatively schizophrenic patients with intellectual disability who merely lie on the autistic spectrum, rather than the psychotic one.

Psychiatrists have long observed the coincidence of poor intellectual function and schizophrenia, but their explanation for the two conditions occurring together has evolved over time. Despite Kraepelin’s (1902) idea that schizophrenia and intellectual disability might be interrelated when they co-occur, Luther (1913)
spoke for many by identifying schizophrenia and intellectual disability as two separate illnesses.\(^{31,40,56}\) Brugger (1928) and Kallmann (1941) later separately confirmed this with family studies that showed no increased frequency of intellectual disability in relatives of persons with schizophrenia.\(^{21,36}\) May\(^{33}\) stated a third view, that psychosis in patients with intellectual disability is a condition entirely different from schizophrenia and reflects that population’s diathesis for psychosis. Yet a fourth view suggests that early effects of psychosis on child development contribute to the evolution of intellectual disability.\(^{50}\)

Schizophrenia is a phenomenological diagnosis exclusively based on signs and symptoms, but its diagnosis presumes to some extent a unitary origin for the condition. For example, DSM-IV specifically excludes symptoms due to a mood disorder, substance intoxication, general medical condition, or pervasive developmental disorder.\(^{19}\) Schizophrenia diagnoses in patients with intellectual disability imply that they suffer from at least two distinct pathophysiological processes or problems (intellectual disability may result from none or several diseases). Though a person exhibits symptoms that appear diagnostic of schizophrenia, the goal of modern psychiatric diagnosis is for the label to parallel an as-yet-to-be-discovered pathophysiology.

**Problem**

There have been several attempts to improve the precision of psychiatric diagnosis for persons with intellectual disability and psychotic symptoms, especially diagnostic scales like the Psychopathology Instrument for Mentally Retarded Adults or the Reiss Screen for Maladaptive Behavior.\(^{19,44}\) The research community has extensively studied and utilized these instruments and they are undoubtedly important tools in both diagnosis research and treatment. Any scale or instrument can be valid in three ways. Content validity is the accurate measurement of what one wishes to measure, for instance psychoticism. Criterion validity is measurement that adequately correlates with some standard, for instance a scale’s agreeing with structured interviews. Construct validity is accurately identifying the internal condition one seeks, in this case, “true” schizophrenia.\(^{14}\) Although some investigators have found structured diagnosis of schizophrenia in intellectual disability to have content validity and criterion validity, very few have attempted to demonstrate its construct validity, or the construct validity of the diagnosis of schizophrenia in intellectual disability by any means.

Whether persons with intellectual disability who are diagnosed with schizophrenia in fact have schizophrenia, is more than a strictly heuristic question. While there are numerous evidence-based interventions for schizophrenia in persons without intellectual disability, these treatments might not be the most helpful for persons with intellectual disability and psychiatric illness. There are many reports for example that illustrate neuroleptics’ efficacy in persons with both schizophrenia and intellectual disability, but they do not elucidate whether neuroleptics are more beneficial for patients with intellectual disability and psychiatric illness, than for those with just target symptoms (such as aggressiveness or suspiciousness) but no second diagnosis. If neuroleptics were not more helpful for patients with intellectual disability and psychiatric illness, this would violate the principle of “parsimony of diagnosis” because the extra diagnosis did not contribute to the treatment. Nor do studies clarify whether other treatments besides neuroleptics are more appropriate. Though there exist extensive and responsible guidelines for the treatment of patients with intellectual disability, the research they are derived from is flawed.\(^{4,13,32,45}\) There are many more specific reasons that the superfluous diagnosis of schizophrenia in persons with intellectual disability might be harmful.

The diagnosis of schizophrenia may make the prescription of antipsychotics more likely in preference to stimulants, lithium, antidepressants, anxiolytics, or non-medical interventions. If psychotic phenomena result in part from sensory impairments, treatments to address these deficits (blindness and/or deafness) like hearing aids might be more effective than medical therapy. Ten percent of persons with intellectual disability are deaf, for example.\(^{19}\) There is a possibility furthermore that less medicalization of persons with intellectual disability might be beneficial. In a study of interdisciplinary approaches to intellectual disability, Glaser and Morreau\(^{17}\) found that the interdisciplinary review of medical regimens reduced medication amounts without increasing injuries, compared to physician management alone. Whether superfluous diagnoses were
responsible for the patients' excessive regimens warrants further study.

Its diagnosis may mislead family members about prognosis. While numerous persons with schizophrenia exhibit the so-called Kraepelinian course, persons with both intellectual disability and schizophrenia do not necessarily have the same evolution. For example, paranoid symptoms often remit in persons with intellectual disability. Moreover, the schizophrenia diagnosis carries a considerable social stigma.

Antipsychotics safely and effectively treat schizophrenia in the general population better than psychotherapy, but this may not be true in persons with intellectual disability. While psychotherapy plays a limited role in the management of schizophrenia, it may potentially play a far larger role in the management of persons with intellectual disability and psychiatric illness. To my knowledge, no one has ever undertaken a trial of psychotherapy specifically for psychotic symptoms in persons with intellectual disability. At least one team has looked at behavioral intervention for a psychotic symptom (bizarre speech) however and found it effective.

The identification of schizophrenia in patients with intellectual disability leads to a large number of false-positive diagnoses. Until we can understand the pathophysiology of schizophrenia, at which time undoubtedly we will identify numerous phenotypes and varying degrees of penetrance, we should apply as rigid a definition as possible to maximize the utility of the schizophrenic disease construct. This will also maximize the usefulness of evidence-based treatments as we develop them. Baldessarini's2 entreaties for the integrity of bipolar disorder diagnoses should apply to schizophrenia as well. His essay, which deserves wider dissemination, challenged Hagop Akiskal and other's elaboration of bipolar disease sub-types, without any support from Koch Postulate-like evidence that these syndromes are indeed related.

A challenge to the diagnosis of schizophrenia in persons with intellectual disability appears to risk diagnostic overshadowing, the phenomenon of clinicians' overlooking co-morbid psychiatric illnesses in persons with intellectual disability. But this is not entirely accurate. First, no one has clearly defined the degree to which diagnostic overshadowing occurs in the real world, or its clinical significance. Second, though Reiss (who developed the concept) does not distinguish between diagnostic overshadowing in affective disorders from that in psychotic disorders, he clearly emphasizes the importance of affective disorders. Third, to accept that many of the symptoms that we identify as schizophrenic in persons with intellectual disability probably have a different etiology is not to diminish the importance of these symptoms. However, it does open the door to the pursuit of novel approaches to treatment that we might otherwise ignore.

**Review**

How can we demonstrate construct validity for the schizophrenia diagnosis? Robins and Guze48 proposed that one evaluate a psychiatric illness' construct validity in five phases: clinical picture, prognosis, differentiation from other disorders, objective testing, and family history. To apply this to schizophrenia in intellectual disability, we must show that schizophrenia in patients with intellectual disability is the same illness as schizophrenia in the general population. We also have to demonstrate that schizophrenia in persons with intellectual disability and psychiatric illness signifies a useful construct: this is diagnostic parsimony, inherent in the Robin and Guze criteria and the goal of any modern system of disease classification.

Experts have given widely differing descriptions of the clinical pictures and prognosis of each of the subtypes of schizophrenia when they occur in patients with intellectual disability, in part due to confusion over the nosology of paranoid disorders. For instance, the variety of conditions that cause paranoia has complicated the diagnosis of patients with intellectual disability and hallucinations or delusions. According to Reid and others, the paranoid patient with intellectual disability is by and large female, well into adulthood, functioned well until the onset of paranoia, and often improves after the psychosis remits.10,20,21,40,42 The person with paranoid schizophrenia in the normal population, however, presents in very early adulthood and is as likely to be male as female.15,16 This reflects the average and not special syndromes like late-onset schizophrenia. Therefore, the two paranoid schizophrenias do not resemble one another from the standpoint of average age of onset or gender distribution. Furthermore, Reid45 identified sensory deficits (blindness or deafness) in 42% of his paranoid patients, a finding distinctive from the general population with schizophrenia, as well as from the general population with intellectual
disability, which suggests an especially important link between sensory impairment and psychosis in patients with intellectual disability. The general population experiences several psychotic syndromes related to sensory impairment, notably Charles Bonnet and Anton Syndromes, and numerous articles testify to the link between sensory impairment and late-life paranoia.\textsuperscript{7,8} Hearing and vision impairments might be more likely to affect patients with intellectual disability due to their reduced ability to selectively attend, and might be more important in the evolution of their psychoses.

Reid wrote that persons with disorganized or undifferentiated schizophrenia and intellectual disability had the same age of onset, equal sex distribution, and poor prognosis compared to paranoid schizophrenia, as that of corresponding persons in the general population.\textsuperscript{20,21,40,42} Yet there is no precise evidence to suggest that the difference would be comparable to the differences in prognosis between the subtypes of schizophrenia in the population without intellectual disability. Furthermore, no one has explained whether persons with both hebephrenia and intellectual disability have a “poor prognosis” due to their schizophrenia or due to the non-specific severity of their intellectual disability symptoms, whose extent might even make multiple diagnoses more likely. For example there is some evidence that patients with both intellectual disability and a diagnosis of schizophrenia are more brain-injured than would be expected from the coincidence of the two separate and unrelated mental illnesses. In one study, patients with both intellectual disability and schizophrenia experienced more epilepsy and demonstrated more neurological soft signs than singly affected individuals.\textsuperscript{12} Therefore, whether disorganized or undifferentiated types resemble one another in the two populations is ambiguous. The age of onset and gender distribution is the same, while their prognoses may or may not be similar.

Differentiating schizophrenia from intellectual disability is difficult because of the similarities between intellectual disability’s autism and anxiety symptoms and schizophrenia’s deficit symptoms.\textsuperscript{46} Hallucinations and delusions do not appear to have this problem, but paranoid schizophrenia (in whom these are by definition the most prominent symptoms) is the least studied subtype in persons with intellectual disability.\textsuperscript{40} On the contrary, most studies of persons with both diagnoses disproportionately included persons with undifferentiated schizophrenia, in whom no one set of symptoms predominate.

Intellectual disability mimics the deficit symptoms of schizophrenia, but also the positive ones. For example, Reid\textsuperscript{12} factor-analyzed persons with severe intellectual disability to describe behavioral syndromes within this population. He identified a syndrome of social withdrawal and suspiciousness with equal numbers of males and females, and the vignettes he offers to illustrate the complex suggest that symptoms began in early childhood. Either Reid failed to call this syndrome schizophrenia when he should have, or alternatively, he showed how much suspiciousness and aggressiveness could owe to intellectual disability alone, without a superimposed mental illness. Given the childhood onset of the symptoms in these people and the lack of a history of decline from previous milestones, one might conclude at least they did not experience what we label schizophrenia in the rest of the population.

Treatment response to medication, the mainstay of schizophrenia therapy, is another domain that for some might validate schizophrenia in persons with intellectual disability and psychiatric illness. The lack of specificity with antipsychotics complicates this. Clinicians have successfully deployed antipsychotics in many conditions, not only schizophrenia and intellectual disability. Furthermore, antipsychotics have non-specific effects in the population with intellectual disability and suppress non-psychotic as well as apparently psychotic behaviors.\textsuperscript{3,32} Therefore, a post hoc diagnosis of patients based on their treatment response would not be informative.\textsuperscript{48}

The most convincing objective testing in Robin and Guze’s system must be pathology, as it would be in any non-psychiatric illness. Unfortunately, only one study has looked at the differences between brains of patients with both schizophrenia and intellectual disability, and those persons with only intellectual disability. That study, published in Lancet in 1999, utilized MRI to find that each group demonstrated varying degrees of hydrocephalus. Patients with intellectual disability showed the most hydrocephalus, followed by patients with both schizophrenia and intellectual disability, and then patients with only schizophrenia. The latter two categories were closest in several parameters. The
authors concluded persons with intellectual disability and psychiatric illness owed their intellectual disability to their schizophrenia, since they most closely resembled the persons with schizophrenia alone. This paper suffered from several methodological problems, including that the authors did not report the statistical significance of each finding, correct for multiple tests of brain volume (the Bonferroni correction), or provide IQ randomization data. Most important, the authors failed to elucidate the history of subjects’ illnesses though the evolution of their disease would have been the strongest support for the authors’ surprising conclusion.\textsuperscript{51} No one has replicated this finding (or duplicated the study for that matter), and virtually no one has accepted that persons with both intellectual disability and schizophrenia owe their intellectual disability to schizophrenia. The idea recalls Kraepelin’s concept of “pfropschizophrenia,” which suggests schizophrenia to be a neurodevelopmental disorder that might accompany intellectual disability. Kraepelin otherwise believed greater than 90% of schizophrenia to be neurodegenerative. Though pfropschizophrenia is an archaic diagnosis, that schizophrenia might be a neurodevelopmental problem is the prevalent viewpoint today.\textsuperscript{6,19,31}

Robins and Guze\textsuperscript{48} took family history of persons with putative schizophrenia to be the most important proof of diagnostic homogeneity. If schizophrenia without intellectual disability occurs more frequently in the family of persons with both intellectual disability and schizophrenia than in the general population, this would suggest the equivalence of the two schizophrenias. Heaton-Ward\textsuperscript{20} identified a small number of patients with both conditions who had a family history of psychotic illness (eleven of forty-two patients), but he did not publish further details. Hucker\textsuperscript{21} found 12% of parents and siblings of 24 patients with both intellectual disability and schizophrenia to have experienced schizophrenia, reportedly comparable to persons with schizophrenia but no intellectual disability. He did not provide details of how he determined this such as the numbers of relatives involved, and his data were retrospective and selective. Doody\textsuperscript{12} identified equal genetic loads in co-morbid and schizophrenic patients (35 and 31 respectively) as measured by numbers of affected family members, about 30%. Unusual exclusion criteria such as Down syndrome, un-blinded raters, an exceptionally high reported rate of familial schizophrenia in all groups including those with only intellectual disability, and significant differences in neurological disease between groups (co-morbid patients the most affected), all weaken the author’s conclusion that co-morbid patients have a higher genetic propensity to schizophrenia than those with intellectual disability alone. Furthermore, a much larger un-translated Chinese family study found no increase in schizophrenia frequency over the rate in the general population, amongst 354 immediate relatives of 58 patients with both intellectual disability and schizophrenia.\textsuperscript{35}

\textbf{Conclusions}

This review has outlined how no one has demonstrated schizophrenia in persons with intellectual disability to be equivalent to schizophrenia in the general population by any means of determining a psychiatric illness’ validity: clinical picture, prognosis, differentiation from other disorders, objective testing, or family studies. Additionally, no one has demonstrated the utility of its diagnosis in altering treatment, which raises the issue of diagnostic parsimony. Its most easily appreciated symptoms, hallucinations and delusions, are neither stable in persons with developmental impairment nor do they predict psychotic illness.\textsuperscript{22,27} Even in the general population, hallucinations have a variable relationship to psychotic disorders (particularly in children), and their significance in persons with delayed cognitive development is very unclear.\textsuperscript{11,23,39,32} There is evidence that what appears to be schizophrenia in patients with intellectual disability has heterogeneous etiologies. Blindness or deafness, both of which may be risk factors for paranoia, are both important conditions in persons with intellectual disability.\textsuperscript{7,8,19} Environmental stresses might be responsible to a much greater degree for psychosis-like complaints in patients with intellectual disability than we ordinarily imagine possible, even in the stress-diathesis model of psychiatric illness.

Furthermore, the reported heightened prevalence of schizophrenia in the population with intellectual disability may be artifactual. Since intellectual disability is not a disease but the endpoint of various insults to brain development, one would imagine the connection from intellectual disability to schizophrenia (if there is one) should be through intellectual disability’s numerous etiologies. For example, velocardiofacial
syndrome has been identified as a risk factor for schizophrenia, but there is no statistical association between intellectual disability and schizophrenia in patients with the condition (not all patients with velocardiofacial syndrome have intellectual disability).\textsuperscript{37} Intellectual disability's most important causes, on the other hand, are not risk factors for schizophrenia.\textsuperscript{20} For example, fetal alcohol exposure, the most common cause of intellectual disability in the world, does not lead to schizophrenia according to the only study that has examined this connection.\textsuperscript{19,49} In the absence of a link between schizophrenia and the important causes of intellectual disability, the difference in prevalence of schizophrenia between persons with intellectual disability and persons without can be due to over-diagnosis and misattribution of symptoms to schizophrenia.

There may in fact be a subset of persons with intellectual disability diagnosed with schizophrenia that has the schizophrenia observed in the ordinary population. They have severe negative symptoms that are stable over time, well-differentiated from pre-morbid personality, and suggestive of poor prognosis, just as they are in the rest of the population.\textsuperscript{27} Nevertheless, negative symptoms are the traits of schizophrenia least responsive to antipsychotics.\textsuperscript{5,47} If persons with intellectual disability with prominent negative symptoms have "true" schizophrenia, antipsychotics may still be less effective than other interventions. Reid\textsuperscript{40} observed (and was satisfied with) a 25% response to antipsychotics amongst his patients, considerably less than we observe in the general population with schizophrenia. Menolascino\textsuperscript{36} found the opposite in a small study when he observed antipsychotics to be more effective in persons with intellectual disability and schizophrenia than in persons with schizophrenia alone. However, his subjects were overwhelmingly "undifferentiated," so we do not know which symptoms responded best and which did not respond at all (he provided results for the BPRS sub-scales, but these are not specific enough). No one has repeated his finding. Quantifying the precise risk-to-benefit ratio of antipsychotics is important. Though antipsychotics are amongst the safest drugs physicians prescribe, they are not benign. They contribute to sudden death, weight gain, diabetes, and tardive dyskinesia. Institutions employ them as a type of chemical restraint, reducing patients' quality of life. There exists a consensus that clinicians must use them only when safer alternatives do not exist.

The above discussion focuses on the dearth of knowledge of the co-occurrence of schizophrenia and intellectual disability. Its objections are all negative and there is no evidence that schizophrenia cannot occur in patients with intellectual disability. Nevertheless, we need to answer several key questions:

1. Do persons with intellectual disability and psychiatric illness have an increased incidence of schizophrenia in their relatives?
2. Does this vary with the subtype of schizophrenia they appear to have?
3. Are antipsychotics more effective for patients with schizophrenia and intellectual disability than for patients with intellectual disability but without schizophrenia, and for which symptoms?
4. Are there alternative treatments like psychotherapy or behavioral therapy that are just as effective as antipsychotics for these patients' schizophrenia-like symptoms?
5. Is aggressive treatment of sensory deficits as effective as medication to reduce paranoid symptoms?
6. With respect to the risk for diagnostic overshadowing, could many seemingly psychotic symptoms in persons with intellectual disability actually be indicators of mood or anxiety disorders?
7. Would antidepressants be more effective for these symptoms? One covariate analysis of antidepressant responders, for example, identified psychosis as an independent predictor of positive response to tricyclics in persons with intellectual disability.\textsuperscript{26}

In the absence of the answers to these questions, the evidence we do have suggests there are three groups of persons with intellectual disability diagnosed with schizophrenia. Stress or over-stimulation overwhelms the first group, many of whom have impairments of hearing or sight, because of a reduced ability to process environmental stimuli. They develop paranoia late in life, both because infirmities worsen with time, but also because aging patients with intellectual disability face the loss of caregivers and other dramatic changes. They have no genetic predisposition to psychosis. They benefit from
stress management, modification of their sensory infirmities, and change of environment. They may need antidepressants, even if they are catatonic.54

The second group consists of autistic spectrum patients and anxiety-disordered patients. Either they have inaccurate histories that suggest a decline of function, or challenges to their rigidity brought out their unappreciated autistic symptoms. They have primary deficit symptoms due to whatever arrest of development or lesion causes autistic spectrum disorders. Many of them may owe their intellectual disability to fragile X disease, which some have associated with autistic symptoms.

The third group may in fact have the same schizophrenia that the remainder of the population is at risk for. They experience a decline in their second or third decade and become disorganized or bizarre. These patients may benefit substantially from neuroleptics.36 They may report auditory hallucinations or delusions but their other symptoms are very prominent.

We should investigate the potential for novel interventions like hearing aids, vision correction, antidepressants or even sleep hygiene to modify apparently psychotic disorders, and compare them with the usual medication strategies.55 Another approach would be to consider patients with particular symptoms like auditory hallucinations, rather than diagnoses, because of the heterogeneity of application of DSM criteria in the population with intellectual disability. Therefore, we might investigate the characteristics of patients with complaints of auditory hallucinations, their family history, their response to medication, their prognosis, etc., rather than “patients diagnosed with schizophrenia.” This approach might provide more clinically useful information.

Because construct-valid criteria for eliciting psychotic perceptions and differentiating schizophrenia in persons with intellectual disability remain to be developed, I recommend a skeptical attitude towards the dual diagnosis of schizophrenia and intellectual disability. Sovner54 has already suggested the privileging of family history in the diagnosis of affective disorders with intellectual disability. The clinician should probably reserve the diagnosis of schizophrenia and intellectual disability for persons with strong family histories of psychotic illness, or whose unmistakable clinical course follows relatively preserved premorbid personality and communication. By doing so, we can increase the utility of precise behavioral diagnosis in patients with intellectual disability, a cornerstone of modern psychiatric care.

References


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