Psychotic disorders in People with Mental Retardation: Diagnostic and Treatment Issues

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Psychotic disorders manifest themselves with hallucinations, delusions, loss of contact with reality, and strange behavior, and may be difficult to diagnose in people with mental retardation (MR). A careful history including associated psychiatric symptoms, behaviors observed by caretakers, medical or neurological disorders, current medications and possible substance abuse is needed to make an accurate diagnosis and to determine when antipsychotic medications are indicated. This paper presents, with case examples, a detailed description of psychiatric disorders with psychoses known to occur in people with MR. Most of these disorders are improved by antipsychotic medications, at times in combination with other psychotropic medications.

Although it may take considerable effort to identify psychotic symptoms, the occurrence of psychotic disorders, with hallucinations, delusions, loss of contact with reality, and strange behavior, in people with mental retardation (MR) has been recognized since early in this century. Since the 1950's, neuroleptic or antipsychotic medications have been established to be effective in the treatment of psychotic disorders, which are thought to be mediated in the brain via dopamine (DA) pathways through projections in the temporal lobe, subcortical striatum, and frontal lobe. The action of antipsychotic drugs is probably primarily via D2 dopamine receptor blockade.

With the use of antipsychotic drugs in people with MR being viewed negatively due to past misuse, their valid, appropriate, timely, and therapeutic use in those with MR and psychotic disorders may be overlooked. The diagnosis of a psychotic disorder in a person with MR thereby deserves careful attention. In this paper, we will examine the diagnostic aspects of psychotic disorders and other conditions in which a psychosis may appear in those with MR.

A psychotic disorder includes impaired reality testing (severely distorted perception of reality and lack of awareness of distortions), delusions (fixed false idiosyncratic beliefs), hallucinations (false perceptions, usually auditory or visual), and bizarre behavior. These mental phenomena can be recognized in people with MR, especially those with mild to moderate degrees of MR. In contrast, a psychotic state is short-lived and largely limited to impaired reality testing and is not reliable for use in people with MR because the cognitive limitations already present are misleading. The psychotic disorders to be considered are those disorders which, at some time in their course, have hallucinations, delusions, impaired reality testing and bizarre behavior, and which are known to be responsive to antipsychotic (neuroleptic) drugs.

Although the recognition of these four criteria establish the diagnosis of a psychotic disorder, it is usually the associated psychopathology and time course that will identify the type of psychotic disorder. Acute psychotic disorders occur in hours to weeks; chronic psychotic disorders are usually at least six months or longer. Periodicity of disturbed functioning and deterioration in social/adaptive functioning from higher premorbid levels are clues to diagnosis. The person with mild MR and a psychosis will appear similar to the intellectually normal person with psychosis, while the person with profound MR and a psychosis may be difficult or impossible to recognize except for the major deterioration in functioning. With time and the assistance of others who know the person with MR well, one can come to understand his/her perceptions more clearly so as to arrive more closely at an accurate diagnosis and to establish the likely presence of a psychotic disorder. Standardized questionnaires have been used in the detection and quantification of psychosis in the MR population.

While there is some evidence for an increased prevalence for psychiatric disorders in people with MR in comparison with the general population, there is also some evidence that people with MR are at great risk for psychotic disorders.
example, it has been observed that between 5.6% and 12.4% of individuals with MR at the time of admission to MR institutions have been displaying psychotic symptomatology.25,44 The point prevalence rate of schizophrenia in people with MR is estimated to be 3%;22,48,61,63 that for schizophrenia in the NIMH Epidemiological Catchment Area (ECA) study in the general population is 0.5 to 0.8%.40 The point prevalence for major affective disorders in people with MR is estimated to be 1 to 3.5%;62,63 the point prevalence for bipolar disorders in the ECA study on the general population is 0.4 to 0.6%, and for major depressive disorders is 1.8 to 2.6%.46 Acute psychotic disorders may be more frequent in people with MR admitted for psychiatric disorders than for intellectually normal individuals.39 However, the reliability of these statistics in the MR population is open to question, and more extensive research will be necessary to clarify the epidemiology of psychotic disorders in people with MR.

A number of acute and chronic psychotic disorders in people with MR need to be discriminated from each other. This paper will focus on differential diagnostic issues of these psychotic disorders as presently recognized by DSM-IV14 and known to occur in the MR population.

**PSYCHOTIC DISORDERS**

1. **Schizophrenia**

Since the introduction of diagnostic criteria provided in 1980 in DSM-III-R,12 schizophrenia is now more accurately diagnosed. The criteria of schizophrenia include at least one month of hallucinations, delusions, thought disorder (disorganized speech), catatonic behavior (immobility and/or unusual posturing), and negative symptoms including flat or inappropriate affect, lack of drive, and decreased speech. There must be continuous signs or symptoms and social/occupational dysfunction lasting at least six months. Schizophrenia often moves on to become a chronic disorder lasting years with negative symptoms, personality change, social isolation, impairment in self-care and role functioning remaining prominent despite traditional antipsychotic medications. Newer antipsychotic medications have reduced the social disability of the negative symptoms.4

With these criteria, it is possible to diagnose schizophrenia more accurately in people with MR. Although the form of the psychopathology in MR is similar to that in the general population, the elaboration or expression of psychopathology in MR is less complex. It often presents as bizarre behavior, persistent withdrawal, and blunted affect that appears in a person who had previously functioned at a higher level.38 Delusions and hallucinations may be naive and poorly sustained or florid and fantastic.46 These criteria apply primarily to those with mild MR whose language skills allow for diagnosis of schizophrenia.36 It is extremely difficult, if not impossible, to identify schizophrenia in the person with profound MR except for the deterioration from a better level of function often associated with aggressive and/or self-injurious behavior.47,50 "Picroschizophrenia" was a term used to describe mannerisms in persons with MR and is not now felt to be related to schizophrenia.49

Schizophrenia and MR have been reported to be found in a number of chromosome abnormalities, including trisomy 21, XXY and XXYY syndromes in males, and Turner syndrome in females.26,30,49,57,61 However Reid49 asserts no specific relationship to have been established between schizophrenia and these disorders. Schizophrenic-like syndromes may occur in people with temporal lobe epilepsy,56 and other seizure disorders,60 and in people with MR and epilepsy.11,15,31 Paranoid delusions, auditory hallucinations, and sometimes a thought disorder occur, but affect and social relatedness remain intact.

The major treatment for schizophrenia in the general population and in those with MR, is antipsychotic medications continued on a long-term basis.4 The typical antipsychotics have been most effective with the positive symptoms of hallucinations, delusions, and thought disorder but less with the negative symptoms of flat or inappropriate affect, lack of drive, and decreased speech. This apathy and social withdrawal of schizophrenia has been more responsive to the new atypical antipsychotics (clozapine, risperidone, olanzapine, quetiapine).20 These atypical antipsychotics are now being used extensively for all psychotic disorders occurring in the general population.19
Although studies have been limited, there is evidence of schizophrenia in those with MR being responsive to the atypical antipsychotics\textsuperscript{54} as well as to typical antipsychotics.\textsuperscript{38} Apart from clozapine, which requires weekly to biweekly white cell counts, the side-effects, including tardive dyskinesias and sedation, are lower in the atypical than the typical antipsychotics.\textsuperscript{19,27} The young man in this case presentation responded well to risperidone:

Mr. A. was a 22-year-old twin with moderate MR secondary to fragile X syndrome. Since age seven he has been in multiple foster homes. At age 22, he suddenly clawed another foster son, threw a wastebasket at the foster mother, and began soiling, sucking his thumb and eating excessively. He was transferred to a new foster home as the only handicapped person in the home. Because of continuing tension he was placed on buspirone (Buspar\textsuperscript{®}) up to 45mg/day, settled in to the new foster home with gradual disappearance of soiling, increasing participation in chores and recreation with the family, and few anger outbursts over five years. Then his behavior deteriorated markedly; for example, he suddenly became angry after being criticized for taking his father's pills, a new behavior for him, progressing ultimately to biting his father's hand. As his illness progressed, further assaults occurred, he was noted to talk to nonexistent people, talk to objects (e.g., his stereo), he withdrew from social contact, and appeared depressed. On Mental Status Exam, Mr. A. was aloof and could/would not answer questions except those that were simple, neutral and concrete. He was started on risperidone up to 3mg/day and over the next year gradually improved, becoming more outgoing, and happier and no longer displayed any psychotic symptoms. He still, however, remains socially aloof compared to his previous friendliness, showing characteristic negative symptoms of schizophrenia.

2. DELUSIONAL DISORDER (PARANOID DISORDER)

A delusional disorder refers to a disorder in which a person has a non-bizarre delusion that is not due to any other disorder and is not associated with odd behavior or change in affective intensity. Delusional themes include erotomania, grandiosity, jealousy, persecution and somatization. A person with MR and delusional disorder may be misdiagnosed as a schizophrenic. Reid\textsuperscript{49} notes that people with deafness or blindness and MR have an increased risk for paranoid disorders, as do members of the general population, particularly older persons with these sensory impairments. Paranoid disorders may be rational reactions to unsafe institutional settings. Cases of erotomania in people with MR are reported by Bhauvik and Collacott\textsuperscript{50} and summarized by Collacott and Napier.\textsuperscript{5} A delusional disorder is usually responsive to antipsychotic medications, as in the following case:

Mr. B. was a 25-year-old young man with mild MR who had been in numerous foster homes and left his adoptive home at age 18 after sexually abusing a sister. He moved back to foster care and ultimately was able to hold a job in a nursing home laundry, move into an apartment on his own and buy his own car. At age 25, he developed a delusion that a female co-worker knew all his personal life and was spreading news about him to her friends who were following him. This delusion persisted for six weeks. He was admitted to the hospital where Mental Status Exam showed a guarded and suspicious young man who firmly believed in the above delusion but did not have any other delusions, hallucinations, psychotic symptoms, or mood disturbance. Unsuccessful trials of risperidone and olanzapine led to his placement on clozapine with dramatic improvement of both the delusion and his social responsiveness. Mr. B. returned to his previous level of functioning with no further delusions.

3. SCHIZOPHRENIFORM PSYCHOSIS

The symptomatology of this psychosis is similar to that of schizophrenia but the duration is one to six months. Environmental precipitating factors may well be present. Schizophreniform psychoses, which have been successfully treated by antipsychotics, have been reported in people with MR.\textsuperscript{54,62} A schizophreniform psychosis may persist and become recognized as chronic schizophrenia which requires long-term antipsychotics.

4. BRIEF PSYCHOTIC DISORDER

Psychotic symptoms often suddenly appear in response to acute stress and last from a day to a month. Symptoms include delusions, hallucinations, disorganized speech, and grossly disorganized or catatonic behavior.\textsuperscript{14} People with MR may be more prone to what were called "reactive psychoses" than the general population.\textsuperscript{39,47} Those with personality disorders (particularly schizotypal, schizoid, paranoid, and borderline) are particularly at risk for brief psychotic episodes.\textsuperscript{54} Brief psychotic episodes
usually respond to a short period of antipsychotic medication. This woman has had several brief psychotic episodes, only recently receiving antipsychotic medication.

Ms. C. is a 46-year-old woman with borderline intelligence, schizoaffective disorder, and hearing loss requiring aids. For many years she drove with her parents to their winter home in Florida. Each fall she had to do all the planning and packing for the trip and drive one of the cars to Florida, an overwhelming task for her. After her parents died she continued to make the trip by air with a caretaker/companion. Each October she showed strange behaviors, becoming anxious, rearranging items in her home, putting the toaster in the bathroom, putting all her clothes into the tub to soak, and filling the bathroom drawers with water. Her communication suggested that she was disoriented. Her behavior for the rest of the year was not unusual with a normal mental exam in April. The following October, after a phone call from a cousin in Florida, she became anxious and disorganized, thinking she was pregnant with twins and obsessed with the need to prepare for the trip to Florida. Mental Status Exam revealed her to be anxious, laughing and silly, concentrating poorly and disoriented for time. She was severely thought disordered and incoherent. She was placed on risperidone 2mg/day and promptly became calmer, oriented, coherent with no delusions of pregnancy or hallucinations. By mid-December the risperidone was stopped, and she went on the trip to Florida with her companion without problems.

5. **Shared Psychotic Disorder (Folie à Deux)**

This is a rare psychotic disorder with delusions shared by two people, usually two family members. For example, a relative in an extended family develops a psychotic disorder such as schizophrenia; the relative with MR also accepts and acts in accordance with the delusion. It has been reported in Down syndrome by Ghaziuddin and Mazzoli. The psychotic disorder in the individual with MR usually will resolve with pharmacotherapy of the relative but sometimes with separation of the two individuals.

6. **Mood Disorders with Psychosis**

Mood disorders are divided into bipolar disorders and depressive disorders. The essential features of bipolar disorder are one or more manic episodes and usually a history of major depressive episodes; for depressive or unipolar disorders is one or more periods of depression without mania. Psychotic features can occur both in manic episodes and major depressive episodes and usually are short-lived.

The treatment for those with bipolar mood disorders is primarily mood stabilizers (lithium, carbamazepine, and valproic acid) and for depression, both bipolar and unipolar, antidepressants. The short-term or long-term use of antipsychotic medication in combination with mood stabilizers and/or antidepressants may be necessary in the presence of psychotic symptoms. Antipsychotics in combination with mood stabilizers and/or antidepressants are also useful in mood disorders with psychosis in the MR population. The use of multiple psychotropic medications in mood disorders is not inappropriate. However, one does not need to use more than one antipsychotic medication.

**6a. Manic Episode**

A manic episode or mania is defined as a distinct period of abnormally and persistently elevated, expansive or irritable mood plus at least three of the following: inflated self-esteem, insomnia, increased and/or pressured talkativeness, flight of ideas, distractibility, increase in goal directed activity or psychomotor agitation, and excessive involvement in pleasurable activities, e.g., buying or sexual sprees. Delusions and hallucinations of a grandiose nature usually occur in association with expansive mood and are short-lived, usually occurring for less than two weeks. Signs and symptoms of mania in people with MR resemble those in the general population, but are simplified. Euphoria, hostility, irritability, insomnia, psychomotor hyperactivity, agitation, aggression, and hyperverbosity may be quite striking and long-lived. Delusions and hallucinations are florid, naive, and grandiose and usually short-lived. A close study of the natural history of the symptoms, its seasonality and recurrent, linguistic analysis as compared to schizophrenia, and the positive family history of mood disorder should suggest the correct diagnosis in the person with MR and manic symptoms.

A specific syndrome of MR, bipolar disorder, and flexion deformities of the hands has been reported.

While psychiatric hospitalization is often required during a severe manic episode, recurrent
admissions can be prevented with stable long-
term medication management. The following
case illustrates a man who had a florid manic
episode (bipolar I disorder, with psychotic
features) followed by many years of almost
complete control requiring an antipsychotic
medication:

Mr. D. was a 46-year-old man with mild MR and
cerebral palsy (spastic paraplegia) who lived with
his mother until at age 34 he began to be loud,
talkative and hostile. He was often euphoric and
hyperverbal, delighting in wearing numerous
necklaces and hearing "God, Jesus, Mary, and
Joseph" (and sometimes angels) talk to him "telling
him to be good." Mental Status Exam revealed an
animated, euphoric, loud, hyperverbal man
wearing eight necklaces. His speech was
pressured and shifted rapidly from one topic to
another often not making sense. He admitted to
hearing voices as described above. Sometimes God
came through the wall telling him stories. He was
hospitalized and placed on lithium with full control
of above symptoms except some hallucinations
which resolved with molindone (Mobane®). He
moved to a community residence and has been
followed by the author for 12 years. Apart from
periodic pressured speech, irritability and
hallucinations which have responded to
adjustments in molindone, he has never required
further psychiatric hospitalization.

6b. MAJOR DEPRESSIVE EPISODE

The diagnosis of a major depressive episode
consists of depressed mood and/or loss of
interest or pleasure lasting two or more weeks,
plus at least four of the following: significant
weight loss or gain related to decreased or
increased appetite, insomnia or hypersomnia,
psychomotor agitation or retardation, fatigue or
loss of energy, feelings of worthlessness or
excessive and inappropriate guilt, diminished
ability to think or concentrate, or recurrent
thoughts of death and/or suicide. Delusions
and hallucinations may occur, with themes of
personal inadequacy, guilt, disease, death,
nihilism or deserved punishment. The diagnosis
of major depression in a person with MR,
particularly a person with severe or profound MR,
may be difficult. Depression may be manifest as
a sad immobile facial expression with withdrawal,
sleep disorder, impaired appetite, and
psychomotor-immobility or agitation but may be
even more atypical. Symptoms also include
somatic complaints, behavioral regression, self-
injurious behavior or aggression. Appetite
disturbance may present as a weight loss. Sleep
disorder may present as behavior disturbance at
night. Loss of interest may appear as apathy and
fatigue; loss of energy as poor productivity in day
program or loss of self-care abilities. Inactivity
may not be recognized until social withdrawal and
immobility are prominent. While guilt,
worthlessness, and self-deprecation may not be
verbalized, clues may be as an example, "I'm
dumb...stupid...no one likes me" expressed
repetitively. School or work refusal may suggest
depression. Infrequently, hallucinations and
delusions may be prominent, suggesting
schizophrenia or schizoaffective disorder. A
careful history may reveal the concurrent
depressive symptoms to indicate the diagnosis is
a major depression with psychosis.

There are suspicions that major depression
(sometimes including psychotic symptoms) occurs
with somewhat increased frequency in people
with Down syndrome, but further studies are
needed.

The treatment of major depression in the
general population is antidepressants and
psychotherapy. Antipsychotic medication is
indicated when psychotic symptoms are present.
The following case illustrates a major depression
that recurred sometime after the antidepressant
was stopped even though the psychosis was
controlled with an antipsychotic:

Ms. E. is a 68-year-old woman with mild MR and
limited gait, living in institutional and then
community residence who had always been active
and sociable. Eight months before evaluation, her
smiling, talking, eating, and sleeping decreased.
Her self-care skills declined; she became
withdrawn with frequent crying; she became
inactive and moved slowly; she no longer enjoyed
or was interested in her usual activities; she had
increased self-injurious behaviors. Concurrently
she had been seeing and hearing people she
called the "grandmothers," who stared into her
window frightening her, stole her clothes, and kept
her from sleeping at night. Mental Status Exam
revealed an elderly woman with a walker who
looked depressed and talked about the
"grandmothers talking to her and to heaven." She
was placed on haloperidol 5mg/night and
paroxetine 20mg/day. The paroxetine gave her
abdominal distress and was discontinued, yet her
depressive symptoms had regressed. Her
hallucinations resolved after the haloperidol was
increased to 10mg/day and she seemed back to
her old self for five months. She then had a recurrence of depression with anorexia, fatigue, insomnia, withdrawal, but no hallucinations. Fluoxetine 40mg/day was added to the haloperidol and she has recovered from the depressive symptoms, returning to her previous self for the past year.

7. **Schizoaffective Disorder**

Schizoaffective disorder is diagnosed when a person has the psychotic symptoms of schizophrenia in extensive concurrence with a manic or major depressive disorder. Delusions and hallucinations last for more than two weeks in the absence of mood symptoms, yet the mood disorder is present for a substantial part of the illness. The content of the hallucinations and delusions is often not congruent with the mood symptoms. Little is known about schizoaffective disorder in people with MR. Treatment invariably requires antipsychotic medication in combination with mood stabilizers including lithium, and/or antidepressants.

**Secondary Psychotic Disorders**

8. **Cognitive Disorders with Psychosis (Delirium; Dementia)**

Once termed "organic mental disorder" prior to 1980, a group of acute and/or chronic mental disorders with cognitive impairments, including delirium and dementia, may be superimposed on the cognitive deficits of MR. These disorders can usually be recognized as secondary to a non-psychiatric medical disorder, to substance use, or to medications used for medical or psychiatric purposes. A psychotic component manifested by hallucinations and delusions is commonly part of delirium and dementia and may be the most prominent feature.

Those with cognitive disorders with psychoses (delirium and dementia) require primary treatment to the disorder if that is available. An antipsychotic for a brief time may help in the management of agitation and hallucinations in person with delirium and for a longer time for dementia. These principles apply when abuse of drugs or medications are involved; remove or reduce the causal agent and consider an antipsychotic only where agitation is dangerous.

Careful management of environmental stimulation to reduce disruptive and dangerous behaviors is critical in all people with MR and secondary psychotic disorders and can frequently eliminate the need for antipsychotic medication. The physician prescribing the antipsychotic in each clinical situation should discuss with the caretakers the relative need for antipsychotics and the alternative approaches available for that situation.

8a. **Delirium**

Delirium is characterized by a disturbance in consciousness and a change in cognition that develop over a short period of time of minutes to hours. The disturbance in consciousness is manifested by a reduced clarity of awareness of external stimuli with an inability to focus, sustain or shift attention. The accompanying change in cognition may include recent memory loss, disorientation to person, place, and time, language impairment such as dysnomias, dysgraphias and perceptual disturbances including illusions and hallucinations. It may fluctuate over the day-night cycle and is often worse toward evening.

The Mini-Mental Screen is a useful measure in the general population for recognizing the cognitive impairments of delirium as manifested by a score below 24 out of 30. A person with mild MR usually scores above 24, but, in the presence of an acute psychiatric disorder and a MMS score below 24, would be suspected to have a delirium. The MMS is not useful in those with moderate to profound MR. Although not often done, the diagnosis of delirium can be confirmed with electroencephalogram (EEG) demonstrating excessive slow waves.

Once referred to as mental confusion, toxic psychosis, or acute brain syndrome, delirium is frequently observed in patients in hospitals whose conditions include medical conditions such as infections, cardiovascular disease and cancers, metabolic disorders such as hypoxemia, hypoglycemia and uremia, neurological conditions such as brain trauma and stroke, or substances such as alcohol, drugs, or medications. Abrupt withdrawal of substances such as alcohol, diazepam (Valium®), or diphénylhydantoin (Dilantin®) may lead to a withdrawal syndrome including delirium (known as delirium tremens -DT's - when alcohol is withdrawn after chronic abuse). Little is known about people with MR and delirium except to suspect it is frequent in those in hospital with similar conditions.
8b. DEMENTIA

The essential feature of dementia is the appearance of multiple cognitive deficits that are always irreversible. Usually occurring in clear consciousness, these deficits include impairment of memory, judgment, personality, and higher cortical functions such as aphasia, agnosia and constructional apraxias. The disturbance is severe enough to interfere with work and relationships with others. Hallucinations and delusions may be an initial presentation, but frequently appear in the later stages of mental deterioration.

Causes of dementia include Alzheimer's disease, vascular disease, central nervous system infections (including syphilis, tuberculosis, fungus, viral), the latter including AIDS and Jakob-Creutzfeld disease, trauma, toxic-metabolic (such as pernicious anemia and hypothyroidism), normal pressure hydrocephalus, and neurologic diseases such as Huntington's chorea, multiple sclerosis, cerebellar degeneration, Parkinson's disease and postanoxic and posthypoglycemic states. Reversible causes of dementia include hypothyroidism, pernicious anemia, and chronic use of one or more medications, including diazepam (Valium®).

Dementia may occur in a person with MR and is a deterioration of cognitive abilities (especially memory) from a previous level of functioning. A child with MR whose cognitive abilities fail to improve with age and whose IQ thus drops proportionately is less likely to have a dementia than one who actually loses cognitive skills. Each person with MR who shows a clinical and psychometric deterioration in cognitive functioning should have a careful neurological examination, psychological/neuropsychological testing, and appropriate testing such as EEG, MRI, and CT scanning and blood studies to establish the diagnosis and to identify treatable conditions.

People with MR and dementia sometimes present with hallucinations somewhere during the course and are helped with a course of antipsychotic medication.

9. SUBSTANCES CAUSING PSYCHOSIS
(DELIRIUM/DEMENTIA)

Any person with MR who presents to an emergency room with a new acute psychosis with hallucinations and delusions should be carefully examined for the possibility of the clouded consciousness of delirium or the cognitive deterioration of dementia. A careful history including all psychiatric symptoms, substance abuse, associated medical or neurological disorders, and all medications is essential. Physical and neurological examinations and toxic screens of blood and urine, including levels of current medications, may provide the accurate diagnosis and etiology of the acute psychiatric symptoms. An emergency MRI or CT scanning may be indicated.

There are a number of substances, those that are abused and those that are therapeutic medications, that may lead to the clinical picture of delirium and/or dementia with the psychotic symptoms of hallucinations and/or delusions. Substances that are abused that cause this clinical picture include intoxication or withdrawal of alcohol, hallucinogens, inhalants, stimulants, cocaine and opioids.

There are also many medications whose toxic effects include delirium and/or dementia. Psychotropic medications include antipsychotics, antidepressants, lithium, anticonvulsants used as mood stabilizers, sedatives, antianxiety medications, and anticholinergic medications.

Treatment is as indicated above: remove the toxin; reduce or stop the medication; carefully support the individual while clouded and/or agitated; and use small doses of antipsychotic medication where psychosis is problematic.

10. SECONDARY PSYCHOSES IN MR SYNDROMES

a) Homocystinuria. Homocystinuria, an autosomal recessive disorder of methionine metabolism, presents with some of these signs: ectopic lens, Marfan-like features, cerebrovascular accidents, MR, and psychotic episodes. While several cases of "schizophrenia" and homocystinuria have been reported, it would appear that the episodic psychoses do not truly represent schizophrenia, but rather delirium or secondary psychosis.

b) Other hereditary biochemical abnormalities. A familial occurrence of Argininosuccinic Aciduria with paranoid psychosis and MR has been reported. Cystathioninuria and Hartnup Disease, with accompanying MR have been reported to have superimposed psychotic episodes. Persons with untreated phenylketonuria have been variously described as
being hyperactive with tantrums, withdrawn, bizarre, schizoid, psychotic, or schizophrenic. The phenomenology of these psychoses were not clearly reported and schizophrenia is unlikely in any one of them. Secondary psychoses is perhaps a more appropriate diagnosis. Treatment of psychotic symptoms with antipsychotic may relieve troublesome symptomatology.

**DISCUSSION**

Careful diagnosis of psychiatric disorders has been increasingly emphasized since specific interventions for identified psychotic disorders in the general population have become available and particularly since the introduction of the DSM-III, DSM-III-R and DSM-IV. Although sometimes with difficulty, these diagnostic categories are also applicable to the MR population.

The diagnosis of psychotic disorders in people with MR is, however, more difficult than in those with normal abilities. One must utilize every resource available, particularly the observations of caretakers in addition to mental status examinations (often must be repeated), physical and neurological examinations and laboratory studies such as medication blood levels and toxic screens of blood and urine, to arrive at an accurate diagnosis and the appropriate treatment, whether pharmacological or psychological. Standardized measures may help identify those with psychotic disorders and measure their response to therapy.

There are a number of psychiatric disorders, often with withdrawal, agitation, and/or strange behavior that may simulate or mimic psychotic disorders. Most of these conditions are treated with other interventions, both psychotropic medications and psychological measures. A separate paper discusses these conditions, giving case examples.

**TREATMENT**

With a more accurate diagnosis of the psychiatric disorder in the person with MR with or without psychotic symptoms, one can arrive at more appropriate management decisions. In this way one can avoid the indiscriminate use of antipsychotic medications in all persons with MR and psychiatric disorders, as has been a pattern of treatment of the past.

Neuroleptic or antipsychotic medications continue to be the major agents in the treatment of psychotic disorders whether in the general population, where there is extensive research, or the MR populations, where there is not. Other psychotropic medications will be indicated, depending on the diagnosis. These include antidepressants, lithium, other anticonvulsants used as mood stabilizers (carbamazepine, valproic acid, lamotrigine, and neurotin), antianxiety drugs (benzodiazepines and buspiron), and/or sedatives (benzodiazepines, barbiturates, chloral hydrate and others). The medications indicated for each psychiatric diagnosis in the general population are probably the same for MR. However the number of controlled studies for psychiatric disorders in the MR population has been limited compared to the extensive studies done in psychiatric disorders in the general population.

Those with severe and profound MR and persistent disturbed behavior that is not responsive to behavioral measures and nonpsychotic psychotropic medications and that presents dangers to self and others - self-injurious and aggressive behaviors - may in fact represent psychotic disorders, as demonstrated in the case of Mr. A. Antipsychotic medication may be indicated where other medications are insufficient to control the dangerous behavior. One must however look carefully for specific psychiatric disorders, particularly mood disorders, both mania and depression, where the mood stabilizers and antidepressants are the primary medications and antipsychotics are secondary. Major depression in severe and profound MR, often associated with aggressive and self-injurious behavior, is more likely to respond to antidepressants alone where psychosis does not seem to be present.

With the introduction of new atypical antipsychotics, unacceptable side-effects can be avoided. These medications, including risperidone (Risperdal®), olanzapine (Zyprexa®), quetiapine (Seroquel®) and clozapine (Clozaril®) produce less sedation, less tardive dyskinesia and other movement disorders than the typical antipsychotics. However, Clozapine, a potent and effective antipsychotic, does require weekly or biweekly white cell counts because of the risk of dangerously reduced counts. While controlled studies are limited so far in MR, increasing reports support the value in dangerous and
problematic behaviors in MR as well as psychotic disorders.

The degree of legal and emotional barriers to prompt and appropriate use of antipsychotic medications for psychotic disorders as described above should be reexamined within the MR and legal communities. Laws that require substitute consent by the courts for incompetent persons with MR for antipsychotic medication can become a roadblock to prompt treatment of a psychosis. While emergency clauses may exist allowing for immediate treatment, the support staff may be fearful of following through with psychiatric recommendations because of the belief that antipsychotic medications have no place in the care of people with MR.

This paper is presented with the hope that the appropriate place for the prompt use of antipsychotic medications in people with MR with psychotic disorders may become clear. With careful psychiatric diagnosis, the appropriate use of psychotropic (all types of psychiatric) medications, and the selective use of antipsychotic medications, one can relieve unnecessary burdens and suffering for the individual with MR and psychosis and for those involved with him/her.

REFERENCES


42. Myers BA. Psychiatric disorders mimicking psychotic disorders in people with mental retardation. Ment Health Asp Dev Disabil, in press.


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