Psychiatric Disorders Mimicking Psychotic Disorders in People With Mental Retardation

Beverly A. Myers, M.D.

The accurate diagnosis of psychotic disorders (or any psychiatric disorder) in people with mental retardation (MR) is especially challenging because of cognitive limitations in the interview process. There are a number of psychiatric disorders which may mimic a psychotic disorder, and possibly lead to inappropriate use of antipsychotic medications. These include depressive disorder, autistic disorder, stereotypy/habit disorder, aggressive behaviors, movement disorders (acute dystonia, akathisia), temporal lobe seizures, dissociative disorders, anxiety disorders, fantasies and imaginary playmates, semantic-pragmatic deficit disorder, obsessive compulsive disorder, hearing loss/deafness. Each of these psychiatric disorders occurring with MR is described with case examples given.

When a person with mental retardation (MR) displays strange and unusual behavior, the question of a psychotic disorder being present may be raised. Differentiating between those with a psychotic disorder and those whose symptoms simulate a psychosis can be difficult due to impairments in language and behavior associated with developmental delay. This paper focuses on those conditions in people with MR which may look like a psychotic disorder but are not.

The criteria for recognizing psychotic disorder occurring in a person with MR are no different than in those with normal intelligence, as defined by the DSM-IV. 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. A psychotic state is short-lived and largely limited to impaired reality testing and is not reliably diagnosable in people with MR because of their cognitive limitations. Although identification may take considerable effort, the occurrence of psychotic disorders in people with MR has been recognized since early in this century and has been discussed in a previous paper.

Since the 1950's, neuroleptic or antipsychotic medications have been established as effective in the treatment of psychotic disorders, which are thought to take place in the brain via dopamine (DA) neurotransmitter pathways through projections in the temporal lobe, subcortical striatum, and frontal lobe. The action of antipsychotic drugs is believed to take place via dopamine receptor blockade. Although research on the use of antipsychotic medications in psychotic disorders in people with MR has been limited, they do appear to be the same medications for psychotic disorders that have been well researched in the general population. With the use of antipsychotic drugs in people with MR being viewed so negatively in these times, their valid, appropriate, and therapeutic use in those with MR and psychotic disorders may be overlooked.

While genuine psychotic disorders in people with MR will more likely, but not always, require an antipsychotic drug, disorders mimicking psychotic disorders seldom require one. Distinction is necessary therefore between those with MR and a psychotic disorder and those with MR and a psychiatric disorder which simulates a psychosis and which is less likely to respond to an antipsychotic drug. Many of these latter patients may be improved by some other psychotropic, but not antipsychotic, agent. The recognition of those disorders which simulate a psychosis and their differentiation from a psychotic disorder in a person with MR thereby deserves careful attention. The following psychiatric disorders may occur in persons with MR, where the severity of the disorder, associated unusual behaviors, or peculiarity of the symptoms have at some time mimicked or suggested a psychotic disorder.

Psychiatric Disorders

1. Major Depressive Disorder

Major depression without psychosis can be very dramatic and intense in a person with MR, even suggesting a psychosis by the presence of
unusual and withdrawn behavior. Careful review of symptoms given as criteria of Major Depressive Disorder in DSM-IV must address not only objective symptoms but also subjective complaints that can be identified through observations by support staff and families. \(^29\) At least one of two following symptoms, lasting two weeks, are essential according to DSM-IV: (a) depression manifested by complaints of depression, crying, and/or looking depressed; and/or (b) loss of interest or enjoyment in usual activities lasting at least two weeks. Other criteria include changes in sleep; in appetite, often with weight changes; slowed activity; fatigue or loss of energy; poor concentration; preoccupation with suicide, death, or suicidal attempts; feelings of worthlessness or guilt. Five of these nine criteria are needed for the diagnosis of major depression and can be observed in those with severe as well as mild MR. \(^21\) Newly appearing or increased self-injurious and aggressive behavior may also be associated with major depression. \(^15\) The appropriate treatment for major depression without psychosis is antidepressant medication, plus psychotherapy or counseling. \(^5\) The following case has a typical pattern of major depression without psychosis. The dramatic severity with aggression may be mistaken for a psychotic illness.

Ms. A. was a 30-year-old woman with moderate MR who lived at home and was compulsively neat and orderly in caring for herself at home and at a vocational site. Six months previously, she had become anxious and wanted to run away. She frequently became angry with tantrums, smashing her dolls and furniture and hitting other people. Her appetite declined and she lost 30 pounds; she frequently wandered at night. She complained of fatigue and seemed to have no energy for daily activities. She lost interest in TV, movies, and community trips and became socially withdrawn, avoiding all interaction with others. She cried frequently and giggled for no reason. She stated that she hated herself. Her neat and orderly ways declined and she cared little for her hygiene needs. With the diagnosis of major depression, she was placed on paroxetine (Paxil\(^\text{®}\)) 10mg/day. All the above symptoms dramatically improved and she returned to her old self in two months.

2. AUTISTIC DISORDER/PERVERSIVE DEVELOPMENTAL DISORDER

Once considered to be childhood psychosis or childhood schizophrenia, this group of conditions are not now considered to be psychotic disorders. \(^4\) The criteria for autistic disorder include severe impairments in (a) reciprocal social interaction, (b) verbal and nonverbal communication, and (c) markedly restricted, repetitive, and stereotyped repertoire of behavior, interests, and activities. \(^4\)

Although the child and adult with autistic disorder has some of the behaviors of the schizophrenic (i.e., social isolation and withdrawal, peculiar behavior, blunted or inappropriate affect, and oddities in language), he or she does not have the hallucinations, delusions, and type of thought disorder of schizophrenia. \(^5\) A person with autistic disorder may be thought to show a "psychotic state" with a very distorted perception of reality, but the absence of hallucinations and delusions rules out a psychotic disorder. Nevertheless, schizophrenia can co-occur in a person with autistic disorder, having been noted in autistic adults, with the appearance of hallucinations and delusions, and a further deterioration of adaptive functioning. The risk of schizophrenia in people with autistic disorders is probably no more frequent than that of schizophrenia in the general population, suggesting that autistic disorder and schizophrenia are not inter-related. \(^3\) Antipsychotic medications are sometimes used in people with autistic disorder with MR to control the dangerous self-injurious and/or aggressive behavior that is often co-occurring. \(^12\) Mr. B. was a 27-year-old man with moderate MR and autistic disorder, showing a severe disturbance in social relationships with marked social withdrawal, severe limitation in the use of language for communication, and repetitive behaviors since early childhood. Despite intensive behavior modification interventions, he retained a severe disturbance in social and communication skills but he had never revealed any hallucinations or delusions. Intellectual testing as a adult showed that he was functioning in the moderate to severe MR range with his scores in the two to seven-year-old level. His visuomotor skills were stronger than his verbal skills. Communication skills were at a two year level with limited use of language to express his needs, and frequent echolalia, suggesting poor comprehension. Educational skills ranged from three to seven years with good reading skills without comprehension, and good math skills with poor understanding of the concepts. Problematic behaviors had been banging objects to the point of breaking them, and jumping and aggression, particularly in crowded or noisy situations, pica of mild degree and some hand to face self-stimulation. These have improved somewhat with behavioral interventions but compulsive behaviors were reduced further with clomipramine (Anafranil\(^\text{®}\)) 125mg/day, andgetQuery (Stelazine\(^\text{®}\)) 6mg/day. His diagnosis was autistic disorder with compulsive behaviors and aggression.

3. STEREOTYPY/HABIT DISORDER

It once was frequently suggested that the non-relating individual with severe or profound MR
and stereotypic behavior had schizophrenia. This is not likely to be so. The stereotypic behavior of the person with profound MR is defined as the frequent or sustained repetition of a pattern of behavior which appears to the observer to be inappropriate, useless or harmful in the reality situation of the individual concerned. Stereotypy can be differentiated from the posturing (catatonia) of the person with schizophrenia and is usually associated with limited ability to relate and communicate, with autistic or pervasive developmental disorders, and/or with severe or profound MR. Treatment should be focused primarily on behavioral interventions, fostering more adaptive behavior, with pharmacotherapy reserved for those with serious self-injurious or aggressive behavior.

Mr. C. was a 45-year-old man with profound MR, cerebral palsy (dystonic quadriplegia), and severe visual impairment with one lens replacement. He is wheelchair bound but can walk with braces and two person assistance. He is nonverbal and does not interact with others. Throughout his life he has had many self-stimulatory movements, including hand flapping, rubbing his lips, holding his ears or head, sucking his thumb, scratching his head or chest, covering his eyes, shaking his head, making noises, biting himself, clacking his jaw, and kicking his feet. He responds mostly to inner stimuli and reacts negatively to any demands which involve touching his skin, such as brushing his hair, shaving him, washing his hands. Yet he does help with dressing, toiletting on schedule, and has recently learned to feed himself soft foods with a spoon. His behaviors were not dangerous and thus pharmacotherapy was not indicated.

4. AGGRESSIVE BEHAVIORS

Aggressive behavior is one of the major behavior problems in people with MR and probably occurs in relation to many psychiatric disorders, whether psychotic or non-psychotic, with both environmental stressors and organic causes. Environmental stressors include harsh or abusive treatment, excessive expectations, disruptive or hostile circumstances, and insensitive care. Aggression may be part of manic or depressive disorders, of schizophrenia, of delirium, or dementia. Aggressive behavior may also be a long-standing behavior disorder emerging from a combination of environmental and/or organic factors and without evidence of psychosis. Behavioral approaches are essential in dealing with all aggressive behavior. Specific medications are indicated in specific psychiatric disorders, including antidepressants for depression, mood stabilizers (lithium or anticonvulsants) for mania and antipsychotic medications for psychotic disorders. For non-specific aggression, buspirone (Buspar®), carbamazepine (Tegretal®), and valproic acid (Depakote®) medications are often used. Sometimes, particularly when the individual is dangerous, treatment includes antipsychotic medication, even in the absence of psychosis. Since it may be very difficult to discern hallucinations or delusions in an aggressive person with severe to profound MR, a psychotic disorder may actually be present and responsive to neuroleptics (antipsychotics). Thus neuroleptics are occasionally necessary in combination with behavioral interventions for aggressive behavior where other medications and/or behavior modification alone cannot help enough. The two cases presented below show differing outcomes for long-term aggressive behavior:

Mr. D. was a 28-year-old man with mild MR diagnosed with intermittent explosive disorder. He had lived in a stressful home with his dysfunctional family until leaving after a fight. One year before, he had become infatuated with a staff person at the workshop he attended. Despite her departure his obsession continued; he threatened suicide if he could not be with her; he had frequent angry and violent outbursts, often over her. The outbursts continued despite moving to a group home supported by a community agency and the introduction of behavioral intervention and counseling. Buspirone up to 30mg/day was tried initially for the anger outbursts with little success. Although carbamazepine 1600mg/day up to a blood level of 8-9mg% has markedly reduced the anger outbursts, he continues to be obsessed with the same woman and cannot accept her unavailability despite continued counseling. He has never had hallucinations. Antipsychotic medication has never been utilized.

Mr. E. was a 22-year-old man with moderate MR related to fragile-X syndrome. At age 22, after numerous foster homes since childhood, he suddenly, without provocation, attacked 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 where he was the only handicapped person. Because of continuing tension, he was placed on buspirone up to 45mg/day. He settled into the new foster home with disappearance of soiling, happy incorporation into the family, and no anger outbursts for five years. He suddenly became angry after being criticized for taking his father's pills, a new behavior for him, and remained angry and demanding all day long, ultimately biting his father's hand. He hit a staff person at the day program, mumbled, not making sense, and seemed to be talking to a nonexistent person. He became depressed, withdrawn, talking to his stereo. On exam he was aloof and could not or would not answer questions, including those about hallucinations. He was started on risperidone (Risperdal®) up to 3mg/day and over the last six months has gradually improved, becoming more outgoing, happier and no longer talking to his stereo. His initial diagnosis was generalized anxiety disorder, but became schizophreniform disorder and may become schizophrenic with longer duration.
5. MOVEMENT DISORDERS

(a) Acute Dystonia

The sudden appearance of persisting deviating eye movements, or spasms of the neck or extremities in a person with MR can be very startling and frightening. With no apparent environmental reason for these bizarre movements, the person may even be thought to be "crazy." The usual explanation is an acute oculogyric (eye deviations) crisis or dystonia (acute muscle spasms) secondary to the recent ingestion of antipsychotic drugs such as haloperidol (Haldol®), perphenazine (Trilafon®), or trifluoperazine (Stelazine®), or antiemetic drugs such as prochlorperazine (Compazine®). The "cure" is prompt oral or parenteral use of an anticholinergic agent such as benzotropine (Cogentin®) or diphenhydramine (Benadryl®), which removes the spasms in a few minutes. The emergency room doctor needs a high index of suspicion for this problem, which may appear mysteriously (for example, when the child with MR has taken a parent's pills for schizophrenia or when prochlorperazine given for vomiting is followed by bizarre body posturing and misinterpreted as schizophrenia.) Complex behaviors may be very confusing. The symptoms of the following teenager were misinterpreted as all psychological and the dystonic tongue movements were not recognized and treated.17

Ms. F. is a 17-year-old verbal adolescent in a wheelchair with spina bifida and moderate MR who presented to the ER having bitten her tongue several times. The tongue was sutured and she was told she would return to the nursing home where she lived and which she disliked. She then bit her tongue almost completely through and fifty percent of her tongue had to be removed with primary closure. The tongue biting was interpreted as a psychological reaction to her dislike of the nursing home. Review of the nursing home records later showed that she had had fewer, sore throat, and vomiting for seven days preceding the ER visit. During this time she had been given prochlorperazine 15mg/day for her vomiting. On the night before the ER visit, she had received an extra 5mg of prochlorperazine. Throughout the night she was protruding her tongue repeatedly, frequently biting it in agitation and was weeping. Tongue protrusion was not recognized as an acute dystonic reaction to prochlorperazine, which can be promptly stopped by oral or parenteral anticholinergic such as benzotropine or diphenhydramine.17 Her depression about returning to the nursing home she disliked did not cause the tongue protrusion.

(b) Akathisia

People with MR who are treated with antipsychotic medications may also experience intense restlessness, anxiety, or inability to sit or stand still, known as akathisia.7 Frequent shuffling movements of the feet (while sitting or standing) of a person on antipsychotic medications may be interpreted as a worsening of his or her psychotic or behavior disorder rather than as akathisia produced by the neuroleptics. A reduction rather than an increase in the antipsychotic is usually indicated; beta blockers will often alleviate this problem.

Mr. G. was a 25-year-old man with borderline intelligence who was able to hold a job in a nursing home laundry, move into an apartment on his own and buy his own car. At 25 he had a six week history of believing a female co-worker knew all about his personal life and was spreading news about him. He thought her friends were following him. Mental status examination in the hospital showed a guarded and suspicious young man who firmly believed in the above delusion but did not have any hallucinations. He was started on risperdone, up to 2mg/day but complaints of fatigue led to a switch to olanzapine (Zyprexa®) 15mg/day, with reduction of delusions and return to work. After four months of being fairly well, he suddenly developed intense restlessness, anxiety and an inability to sit still that persisted over several hours. His delusional beliefs were very mild. Olanzapine was dropped to 10mg/day and propranolol (Inderal®) 60mg/day was started with considerable improvement over the next 24 hours. His acute restlessness was diagnosed as akathisia in a man with delusional psychotic disorder.

6. TEMPORAL LOBE SEIZURE DISORDER (COMPLEX PARTIAL SEIZURES)

Seizures in the temporal lobe area can produce psychiatric symptomatology that sometimes can look like a psychotic disorder. The typical picture is one of confusion followed by a post-ictal(post seizure) confused state. Common ictal emotions include fear, depression, elation, anxiety, anger, and a sense of doom. Delusions, especially paranoid, can present with rage reactions or extreme fearfulness. Hallucinations are reported as well. These signs and symptoms are recurrent and stereotyped and are associated with ictal or post-ictal confusions, amnesia, and automatism.34 EEG abnormalities are often found in the temporal lobe but not always. Antiepileptics rather than antipsychotics are usually indicated.

Mr. H. is a 27-year-old man with Down syndrome and moderate MR who lived alone with his mother after his father's sudden death one year previous. In the past year he had become depressed, with crying, agitation, insomnia, ruminations about his father, self-deprecatory thoughts, self-destructive behavior (biting his arm), and paranoid thinking. He no longer was sociable and had to stop his part time work in the community. On exam he was withdrawn and depressed but had no hallucinations or delusions. Desipramine (Norpramin®) up to 150mg for six weeks was unsuccessful, but clomipramine up to
150mg led to complete recovery from depression. However, within 3 months he began to have sudden episodes of waking up very frightened, saying with conviction that he was in a disaster, and that he saw flames all around him. EEG with temporal leads demonstrated a temporal lobe seizure disorder. He was switched to fluoxetine (Prozac®) 20mg/day with complete disappearance of night time seizures and continued control of his depression. Psychiatric diagnoses included major depression without psychosis, temporal lobe epilepsy secondary to clonazepam.

6. DISSOCIATIVE DISORDERS

The sudden onset of dissociated states or trance-like states (altered states of consciousness with markedly diminished or selectively focused responsiveness to environmental stimuli) can occur with MR. This disruption in consciousness, memory, identity, or perception of the environment manifest in fugue, amnesia, and depersonalization disorders may be quite dramatic and bizarre. It occurs in relation to present or past stressful and abusive events to such an extent that it may be difficult to separate from a brief psychosis (reactive), or acute delirium (toxic psychosis). Moreover, hallucinations, delusions, and thought disorder have been identified to be part of the extensive psychopathology of dissociative identity disorder (multiple personality disorder), another dissociative disorder which may be difficult to separate from schizophrenia. Psychotherapy is a prominent component of treatment, with specific medications in associated symptomatology, e.g., antidepressants. The following case is confusing but probably represents a young woman who experienced more than one traumatic episode with one fusing into another.

Ms. I. was a 33-year-old woman with mild MR living with a couple (relatives) and their young daughter, who was seen because of her behavior. Three years before, she had moved to her relatives' house and began to care for the two-year-old girl while the parents worked. She infantilized the child and was possessive with her, behaving as if the child were her own. This led to a power struggle between Ms. I. and her relative, with Ms. I. undermining the relative's authority with her daughter. The relative informed her that she could no longer live with the family. This led to Ms. I. calling 911, saying she had been raped. She stated the rape occurred somewhere else when she had not left the house of the relative. She had had exploitative and abusive experiences with other relatives in the past. Exam revealed a disheveled young woman who was crying and initially talking in an incoherent manner about the rape as if it had just happened but rapidly shifted to talk about the past with no admission of any problems at present in her relatives' house. She had no hallucinations and did not appear to be psychotic. After a brief hospitalization with no medications given, she was placed in respite care, pending a supervised home placement.

Her psychiatric diagnoses included posttraumatic stress disorder and dissociative fugue state.

7. PANIC DISORDER/SOCIAL PHOBIA/OTHER ANXIETY DISORDERS

Children and adults with MR may experience anxiety to the point of being severely withdrawn or even mute. It may be extremely difficult to discern whether the person has a psychotic disorder or an anxiety state because the nature of his or her inner experiences cannot be identified. If the child or adult is able to confide in a familiar other, it may be possible to determine if he or she is anxious or psychotic. Anxiety, fearfulness, inability to speak in some settings, speaking only with familiar people, in the absence of associated strange or unusual thinking, are more likely to be characteristic of an anxiety disorder, whether separation anxiety disorder, selective mutism, panic disorder, or social phobia. In the person with MR and fearful withdrawal, it may take several assessment sessions plus extensive information from familiar others to determine the nature of the problem. Behavior modification, counseling, antidepressants, benzodiazepines (preferably avoided), or buspiron are some of the approaches possible in treating anxiety disorders. In the following case, an antipsychotic medication was used briefly when the young woman was home, aggressive, and suspicious, but, after moving into a group home, it soon became clear that she did not have a psychotic disorder:

Ms. J. was a 22-year-old young woman with mild MR and severe expressive aphasia who lived with her mother. Ms. J. had witnessed her father's sudden death when she was 12 and she became increasingly fearful of her mother leaving her. She and her mother moved several times, with much stress for both mother and daughter. Mother worked, with Ms. J. remaining home until a day program became available. She became increasingly fearful and distraught with her mother's departures, then increasingly demanding, with constant tantrums and occasional aggression, progressively impairing the mother's ability to cope. Ms. J. grew depressed, neglected her hygiene, showed less independence and ate excessively, gaining 60 lbs in the previous year, but sleep was no problem. She talked about people looking at her, calling her names, and being mean to her. With the introduction of a partial day program and of paroxetine (Paxil®) 20mg and risperidone 2mg (for aggression), she gradually became happier during the time away from mother, but continued to have severe separation anxiety with tantrums when with mother. As mother's ability to cope declined further, Ms. J. was able to move into a community home where she has had an excellent adjustment, becoming cheerful, with no separation problems or suspiciousness. Risperidone and later paroxetine were discontinued, with no recurrence of any psychiatric symptoms. Psychiatric diagnoses include separation anxiety disorder and major depressive disorder.
8. Fantasies/Imaginary Playmates

Since imaginary playmates tend to occur in normal children at the mental ages of three to five, the person with MR and imaginary playmates may be functioning at a similar level. Under stress, his or her imaginary playmates may become prominent and thus be thought to be hallucinations and/or delusions of schizophrenia, but they are an exaggeration of the fantasies. Examination of the nature and duration of the symptomatology reveals that the long-time existence of imaginary playmates, in the absence of social withdrawal or a deterioration in social functioning, argue against the diagnosis of schizophrenia or other psychotic disorder. Medication does not play a role in the management of imaginary playmates. The following case describes a young woman with mild MR and imaginary friends in the presence of a generalized anxiety disorder.

Ms. K. was a 22-year-old young woman with mild MR who was born with genitorespiratory deformities. Although she had surgical repairs, she had chronic constipation which involved frequent enemas for many years and led to intense conflict with her parents. Despite academic abilities at a fifth grade level and independent self-help skills (apart from enemas), she presented at age 18 with intense anxiety, social withdrawal with talking to herself, self-injuring, holding herself with pencils and running away. Residential placement was arranged and, despite improvement in many of her problems, she has continued to have a major problem with anxiety and involvement with fantasy as a means of avoiding tension-related issues. The fantasy usually has centered around Disney stories, listening to tapes, reading Disney books, watching Disney movies, and drawing pictures. She often inserted herself into the scene she was imagining and became so involved she was not able to separate fantasy from reality. Mental status examination showed a fearful young woman who cried to the staff psychologist and had a hard time looking at the examiner. She apparently did not have hallucinations. Gradually the staff have redirected her activities to those more reality based. She became able to recognize when she used fantasy and sometimes could speak to herself to attempt to shift away from her fantasy. Clonazepam (Klonopin®) has been unsuccessful in reducing her anxiety as she tends to cry to staff, who continue supportively to use behavioral approaches to help her relate to the real world.

9. Language Disorder (Semantic-Pragmatic Deficit Disorder)

Children and adolescents with this developmental language disorder articulate clearly, usually in well-formed sentences but make little sense and have severely impaired pragmatic skills (verbal and nonverbal aspects of communication, particularly comprehension), with functioning in the mentally retarded range.

This language disorder has sometimes been confused with schizophrenia because of an apparent “thought disorder” (incoherence of speech) and poor ability to relate. According to Resnick and Rapin, in the semantic-pragmatic syndrome verbal expression is often superior to comprehension. Individuals understand language in a concrete way and have particular difficulty comprehending open-ended questions such as why, when, and how. At times they cannot answer a question, although they know the answer. Yet vocabulary is adequate and they can identify objects by name. In addition to impairments in verbal communication, nonverbal communication (pragmatics of language) is impaired, and include such difficulties as defective turn-taking in conversation, unawareness of effect on others of their constant chatter, narrow choice of topics, and repetitive utterances or echolalia. By contrast, in childhood schizophrenia one sees the presence of an earlier normal (or more normal) social and language development, a later deterioration (beyond age five) with the appearance of withdrawal, delusions, hallucinations, sometimes a thought disorder, a flatter and less responsive affect, a family history of schizophrenia, and a better response to neuroleptics. Caution is needed in making the diagnosis of schizophrenia in a child with a “thought disorder,” where there has been early language delay but fluent speech, poor relating, and no hallucinations or delusions, and where severe impairment of comprehension is a prominent symptom.

Mr. L. was a 53-year-old man with moderate MR living with some relatives, his mother having died. All his life he had spoken with language that did not make sense and was blatantly untrue. Although his articulation was clear and individual sentences were coherent, much of what he said was unrelated to reality and he firmly insisted was true. For example “I like my own office in...” “I am a cop.” Much of what he said was related to TV and TV characters but there was no coherent or consistent story. On exam he related warmly and was cooperative with the examiner, constantly making comments that were incoherent. He had difficulties comprehending larger and more abstract questions, making irrelevant answers and had greater ability to answer short concrete questions correctly. He had no auditory or visual hallucinations. He had presented to the author with a three month history of anger outbreaks and threats of violence without actual aggression, poor sleep and appetite problems. These symptoms appeared to relate to severe tensions at home. A brief period on clonazepam, continuing psychotherapy, and reduction of home tension led to cessation of problematic behaviors, but his irrelevant conversations persisted. Psychiatric diagnoses included adjustment disorder with aggressive behavior and language disorder of the semantic pragmatic type.
10. OBSESSIVE-COMPULSIVE DISORDER (OCD)

The person with MR and repetitive behavior that seems strange may be suspected to have schizophrenia or some other psychotic disorder unless the pattern is recognized to be one of obsessions (repetitive involuntary thoughts) and compulsions (repetitive behaviors). The normally intelligent person with obsessions and compulsions is usually able to say that strange thoughts (violence, contamination, doubt) occur to him involuntarily, and he feels compelled to carry out repetitive acts (e.g., hand washing, counting, checking rituals), no matter how senseless they seem. It is his or her recognition or insight that his or her thoughts, which are not hallucinations, are intrusive and his or her behavior is senseless that precludes the diagnosis of a psychotic disorder. Despite occasional "psychotic states" where the person is unable to respond to the observer because of his or her preoccupation with his or her obsessions, OCD is not considered a psychotic disorder, and delusions and hallucinations do not occur in OCD. In the person with MR, similar patterns of obsessions and compulsions may be recognized, even though he or she usually does not have insight and cannot verbalize their silliness. Compulsive behaviors are particularly common in people with autistic or pervasive developmental disorders and in association with Tourette's disorder. In addition to behavior modification approaches, OCD is usually responsive to serotonergic antidepressants. The first case presented a challenge to his parents and his workshop staff with a characteristic OCD pattern, while the second case shows a compulsively destructive behavior:

Mr. M., a 27-year-old male with Down syndrome and moderate MR living at home, developed rituals and compulsive behavior as an adolescent. He dressed, cleaned, and bathed himself in a slow meticulous manner with frequent hand rituals, and folded his clothing neatly and kept his room tidy. His rituals included greeting his toys one by one in the morning and evening. He resisted distraction, frequently got stuck in the rituals, repeating them, so that he was regularly late. Pressuring him led to many conflicts with his parents. Clomipramine (up to 125mg/day) led to moderate improvement, but side effects of falling episodes, constipation, and abdominal pain led to discontinuation. Fluoxetine 40mg/day led to moderate improvement in his rituals. Ultimately sertraline (Zoloft®) 50mg, a reduction of parental pressure, and a day program involving more interaction with the community have been most effective in diminishing the negative impact of his rituals. His psychiatric diagnosis was obsessive-compulsive disorder.

Mr. N., a 47-year-old nonverbal man with severe MR, had a lifetime history of ripping clothes and shredding shoes. He had been in a home-supported by an agency for five years and had been shredding his shoes at up to 5.5 pairs of shoes per month. Behavioral approaches to reduce the shredding had had a moderate impact with destruction down to two pairs per month. Because he began relapsing to a rate of three pairs per month, he was placed on fluoxetine (Prozac®) 20mg/day with a dramatic reduction of shoe destruction to less than one every month. Discontinuing the fluoxetine briefly returned the destruction to two per month. His psychiatric diagnosis was obsessive compulsive behavior.

11. HEARING LOSS/DEAFNESS

The child with apparent unresponsiveness to another's oral beckoning should be suspected to have a hearing loss even though other problems may be suspected, e.g., autistic disorder. One should suspect hearing loss particularly when language development is well behind gross and fine motor and social development and should be among the possible diagnoses when there is a deterioration of function as in this case.

Ms. O. was a 16-year-old girl with moderate MR who was referred because of a deterioration in behavior and school performance. She also had short attention span, loss of interest in her environment, anorexia, 20 pound weight loss, headaches, staring spells, hand shaking and emotional lability. She thought she had bugs on her (a tactile hallucination). Exam showed a socially unresponsive teenager with hand waving and tooth grinding. She had impaired receptive and expressive skills, repeating single words only, a deterioration for her. WISC-R failed to produce a scoreable response on verbal testing. A Stanford-Binet basal of two years could not be achieved. At 15, her WISC Full Scale was 41, with Verbal 45 and Performance 48. Her physical exam revealed dysmorphic features of synophrys, pronathus, protuberance of the palate and hyperactive deep tendon reflexes. Pure tone testing on the same day revealed right conductive hearing loss and unreliable testing on the left. A large amount of wax was removed from both ear canals on the same day. Over the next month she had an immediate and dramatic improvement, talking and eating more. She became more attentive and returned to her usual level of school work. Exam revealed a teenager who was alert, spoke in short sentences and laughed appropriately. She had no hand waving or tooth grinding. Audiolological testing revealed normal hearing. Six months later she was reading at third grade level, doing math at fourth grade level. Estimated WISC-R scaled scores were Full scale of 44, Verbal 46, and Performance 51. Her psychiatric diagnosis was temporary dementia, and hearing loss caused by wax in ear canals.

DISCUSSION

Accurate diagnosis of psychiatric, including psychotic, disorders in people with MR has practical implications for management when psychotropic medications are involved. In discriminating between those whose delusions, hallucinations, and bizarre behavior represent a
psychotic disorder and those whose condition mimics a psychotic disorder, one can arrive at more specific treatments for each psychiatric disorder. Accurate diagnosis and use of the appropriate psychotropic medication is likely to reduce suffering in many psychiatric disorders, whether in the general population or the MR population. Hence, a careful, educated, and continuing approach to the diagnosis of all psychiatric disorders in those with MR is critical, no matter how difficult the ongoing task.

When the person with MR has limited ability to speak about his or her internal state, a number of measures are needed to supplement the basic history taking and mental status examination. These include the taking of extended history from familiar observers from all settings (home, group home, school, work, workshop, and in the community), numerical records of target behaviors over time, standardized behavioral questionnaires, repeated mental status examinations, and toxic screens. Repeated reconsideration of the psychiatric diagnosis on each clinical visit is necessary, because the diagnosis may have to be changed in the course of the disorder as more information becomes available.

Careful diagnosis of a psychotic disorder and appropriate use of the antipsychotic medication will improve the psychotic disorder. In the absence of danger to self or others in the person with a suspected psychotic disorder and MR, the use of antipsychotic medications need not be immediate. It is essential to include the important others (parent, guardian, case worker, psychologist, group home manager and, legal representatives) in arriving at a consensus that the use of an antipsychotic medication is necessary for the person with a psychotic disorder. This appropriate and indicated use of an antipsychotic drug can reduce the excessive use of such antipsychotic medications, which has been such a problem in the past in the MR population.

The accurate diagnosis of those disorders simulating a psychotic disorder in persons with MR should also reduce the inappropriate use of antipsychotic medications and lead to the more specific treatment of these non-psychotic disorders, whether by pharmacotherapy, behavioral therapy, or other interventions. The scientific study of the use of psychotropic medications for psychiatric disorders in the general population has moved along dramatically in the last 25 years, while study of their use in the MR population has barely begun. These cases have demonstrated the value of medications other than antipsychotic medications in people with MR population who have psychiatric disorders that simulate psychotic disorders. Further scientific study of these medications is urgently needed to determine appropriate treatments for non-psychotic psychiatric disorders in those with MR.

REFERENCES


Dr. Beverly Myers is a Clinical Assistant Professor at Brown University School of Medicine and a psychiatric consultant for the Child Developmental Center, Rhode Island Hospital, 503 Eddy Street, Providence, RI 02903. She has a private practice of child and adult psychiatry in Warwick, RI.