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THE MENTAL STATUS EXAMINATION IN PATIENTS WITH MENTAL RETARDATION AND DEVELOPMENTAL DISABILITIES

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The Mental Status Examination (MSE) is a crucial part of the evaluation of any patient, providing clues to diagnosis beyond what is found in the history and serving as a check or confirmation of caregivers' reports and observations. In the case of patients with mild or moderate mental retardation, the standard MSE format can be usefully followed, although some observations may differ in significance from similar findings in patients who do not have mental retardation and developmental disabilities (MR/DD), and some phenomena may take a different form. For patients with severe or profound mental retardation the MSE format must be altered significantly to be adapted to their needs and the range of phenomena they display. It is nevertheless a useful way to organize and standardize the priceless data of patient observation.

Keywords: developmental disabilities, intellectual, mental retardation, Mental Status Examination, psychiatric assessment

Psychiatric treatment begins with the Initial Psychiatric Diagnostic Evaluation. The clinician meets with the patient and establishes a relationship, obtains information, conducts a Mental Status Examination (MSE), gives feedback and a proposed treatment plan. The MSE is analogous to the internist's physical examination: a "systematic observation and recording about a person's thinking, emotions and behavior."⁶ The MSE is a fundamental part of mental health clinical practice, taught, for example, to all medical students on their required basic psychiatry rotation. It is part of the introductory clinical material in every standard textbook of psychiatry, and was among the earliest topics addressed when mental health clinicians in large numbers were turning to the problems of diagnosis and treatment of patients with mental retardation and developmental disabilities (MR/DD).^{14,15} For people with MR/DD, major advances in psychiatric assessment have begun, including development of one structured clinical interview and companion scales.⁸⁻¹⁰ A differential diagnosis—the list of possible diagnoses of the patient—can only be approached by combining the information from the history of the present illness, past history, and the MSE.

For patients with MR/DD, the clinician must substantially alter the examination and his/her expectations of the patient. The exam assumes that the patient functions normally with regard to

intellectual ability, and he or she is able to understand questions and answer correctly. For patients with MR/DD, and for children, these assumptions may be incorrect. Even if mildly intellectually handicapped, patients with MR/DD may not understand the contextual aspects of even simple questions. Further, they may be pressed to answer "yes" to please an examiner/authority figure, much as is expected with children.^{7,20} Observations of behavior may be thought of as aberrant, when it may be normal within the context of developmental disability. Lastly, many clinicians, faced with even very evident psychopathology, may fail to make a diagnosis in a patient with MR/DD, because the disability "overshadows" the obvious diagnosis.¹³

In this paper, we will explore the application of the MSE to patients with MR/DD and make recommendations for its use by clinicians with this population.

THE MENTAL STATUS EXAMINATION

The MSE consists of observation of the following:

- Appearance
- Psychomotor Behavior
- Intelligence
- Speech
- Emotional Expression
- Thinking and Perception

- Sensorium
- Attention/Concentration/Memory
- Insight
- Judgment

Often diagnosis of a person with MR/DD, particularly an individual with severe or profound mental retardation (S/PMR), is made without the clinician making such a systematic observation of the patient, or in some cases without the clinician even seeing the patient (a dangerous and unethical practice). The usual approach and formulation of the MSE may be thought to be inapplicable to persons with MR/DD, because so much of it appears to depend on a patient's advanced cognition, interaction and verbal report. It is incorrectly assumed that a formal MSE must take place within a traditional formal psychiatric interview setting, but in fact it is simply a way of organizing data, not a formula for obtaining it. Observations could take place in a formal interview, a classroom, a work site, or any other place the patient is encountered, or anywhere the patient is comfortable. The MSE itself must be modified and adapted to the purposes and capabilities of persons with mild and moderate mental retardation (MM/MR) and those with S/PMR.

This paper proposes modifications of the MSE applicable to patients with MM/MR and S/PMR, with suggestions for observations relevant to differential diagnosis of major psychiatric disorders in each range of functioning. It is assumed throughout that the reader is familiar with the basic MSE as described in **Kaplan and Saddock's Comprehensive Textbook of Psychiatry, Seventh Edition** (or previous).⁶ Discussion will be directed to the American Standard Mental Status Examination and the Diagnostic and Statistical Manual of Mental Disorders, Fourth Edition (DSM-IV).²

GENERAL APPROACH TO THE DIAGNOSTIC INTERVIEW

The initial diagnostic interview has four goals: establish a relationship, obtain information, assess psychopathology, and provide feedback.¹⁷ When evaluating a patient with MR/DD, substantial alterations in these assumptions may occur.

ESTABLISHING A RELATIONSHIP

First, because the vast majority of individuals are brought to the appointment by a family member or caregivers, similar to child psychiatric practice, the establishment of a therapeutic relationship must include the caregivers as well

as the patient. This presents problems on several levels. First, the job is more complex; one has not only to relate well to the patient, but to other persons in relationships with the patient. These individuals may be supportive persons in the patient's life, be part of the problem, may or may not be trusted, or may be caregivers who do not know the patient well.

Another difficulty ensues due to the level of disability. For those patients who are in the moderate to profound range of MR/DD, establishing a relationship during an appointment may be very difficult. Personal conversation and discourse, occurring typically with intellectually normal patients, does not occur in the same way. Rather, excessive dependence on caregivers may be expected.

It is important that the clinician, if at all possible, spend time alone with the patient with MR/DD. During this time, the clinician may talk, if the person has MM/MR, but otherwise the clinician may rely on other techniques. The office should have materials available, such as paper and crayons, simple puzzles, or other craft material. Doing a short, simple "project" with the patient may help to establish rapport, and will also allow the clinician to see more of what the person may be like in his or her normal environment. The office is not "normal" and is often quite intimidating to the person with MR/DD.

OBTAINING INFORMATION

The clinician must obtain a history of the presenting problem, in addition to other developmental and health information. Traditionally the clinician obtains this history of the problem from the patient. When working with patients who have MR/DD, this is not the case. Even if very mildly impaired, the ability to accurately give a history and answer questions will be inadequate to this purpose. Further, mental health clinicians are trained to use techniques that are not direct. Clinicians are

trained to ask “open-ended questions” and to explore feelings and thoughts. One can ask a neurotypical patient, “How did your depression develop over the last several years?” and expect a full and revealing answer. The person with MM/MR could only answer in a fairly concrete fashion, with possible distortions of timeline, or misunderstanding of the basic concept of depression.

ASSESSING PSYCHOPATHOLOGY

The clinician assesses psychopathology through his or her impressions during the diagnostic interview, through obtaining of history, the MSE, and through any additional tests that might be ordered. With the exception of the specialization in Psychiatry of Learning Disabilities in the UK, mental health clinicians do not have adequate training in MR/DD.¹⁹ Thus, when evaluating such patients, many misconceptions occur. As noted above, many clinicians fail to diagnose psychopathology because of an assumption that any abnormalities are the result of MR/DD, rather than a psychiatric illness, a phenomenon called “diagnostic overshadowing” by Reiss and his colleagues.¹³ On the other hand, the existence of very worrisome behaviors such as self-injury or aggression, may lead to “behavioral overshadowing,” that is, concluding that the problem is just a “behavior problem.”⁵

GIVING FEEDBACK

Feedback to the patient at the end of the diagnostic interview should be reassuring and provide an outline of the treatment plan. For a patient with MR/DD, this must be done in a very concrete fashion, while at the same time answering the patient’s or any caregivers’ questions.

MENTAL STATUS EXAMINATION OF THE PERSON WITH MILD OR MODERATE MENTAL RETARDATION

Most persons with MM/MR will be evaluated in a more or less formal interview setting, and modification of the standard MSE will be minimal. However, one must include observation of details of more significance and relevance to this population.

APPEARANCE

Typically one begins with an observation of the age of the patient and appropriateness of appearance to age, relative neatness or dishevelment, and any peculiarities of appearance or dress. Persons with MM/MR may live in independent apartment settings or more supervised situations. The vast majority would be expected to perform self-care chores with some degree of independence, but appearance might (and this should be established when history is taken) be in part the product of response to caregiver supervision. In depression one would expect to see almost complete neglect of appearance, with withdrawal from self-care; persons in acute manic episodes would continue to perform self-care tasks in disorganized or haphazard way (e.g., many layers of clothing, over-elaborate or sloppy makeup). Persons with schizophrenia might present a bizarre appearance, with self-care performed in a negligent way, or in a way suggestive of compliance with some but not all caregiver supervision.

One should comment about the use of crutches, a cane or a wheelchair. Sometimes, a person arrives at the clinic in a wheelchair, but this is not the ordinary means of transportation. For example, when there is a considerable distance from the parking lot to the clinic, staff may transport an older individual in a wheelchair for convenience. The clinician should ask if the wheelchair is just for today or is needed all of the time. Even the use of a wheelchair as a temporary vehicle may mean that the person tires much more easily or is more unsteady on his or her feet.

The clinician should comment on obvious lesions, sutures, scars, scratch marks, cuts, misshapen ears and especially any missing fingernails or toenails (e.g., self-removed nails raises the possibility of Smith-Magenis syndrome). Numerous superficial cuts may be a clue of someone with borderline personality disorder. Scratch marks could represent self-injurious behavior or scratching secondary to a drug allergy. Sutures not only mean a recent injury, but also that the injury was of at least moderate severity.

Observations of dysmorphisms may be suggestive of genetic syndromes. For example, most clinicians are familiar with the characteristic

facial and body appearance of someone with Down syndrome. Many other genetic conditions are also associated with abnormal facial appearance or body development.

Mode of relating is part of the patient's overall presentation. Since the evaluation may not have been the patient's own idea, he or she might relate in a *hostile* manner. Since many persons with MR/DD see anyone in authority as a caregiver, the patient may relate in an *overfamiliar* manner which, in a person with normal cognitive functioning, would be taken for inappropriate friendliness indicative of mania or loss of ego boundaries. Patients answering "I don't know" to most or all questions are often assumed to be relating in a *guarded* or *mistrustful* manner; persons with MM/MR may in fact not know the answers to many standard psychiatric interview questions, or not understand the questions. A patient with MM/MR may be *overcompliant*, pleasing the interviewer by giving answers he or she thinks the interviewer wants to hear.^{5,10} Persons with Autism Spectrum (Pervasive Developmental) Disorders might be passively gaze-avoidant, talking with no regard for eye contact with the interviewer (as opposed to actively avoiding direct gaze).

INTELLIGENCE

The clinician often must globally assess the patient's intelligence by impression. Usually, the level of educational background is noted, and the patient's ability to function within the interview is then compared with the expected functioning. For patients with MR/DD, the clinician may have the *range of cognitive functioning* noted by caregivers, but ideally a report of past formal testing by a psychologist should be obtained.

PSYCHOMOTOR BEHAVIOR

The clinician must be alert to motoric appearance. Here, the clinician notes if the patient has good posture or is slumped. Any abnormalities of gait or use of extremities is noted. The patient may be hyperactive or show the opposite, psychomotor retardation. One would note level of activity, involuntary movements (e.g., tics or stereotypies, tremor, choreoathetoid movements, paresis or spasticity), if any.

To test the level of activity, the interviewer can ask the person to try to sit still, and determine if

he or she can stay still for sixty seconds. Persons with akathisia or mania will not be able to sit still for a minute. Alternatively, compare the person's movements with others in the room. It is unusual for a person to go more than a couple of minutes without some movement. If the individual's movement is much less than others in the room, it may be a clue to psychomotor retardation.

Patients with MR/DD typically have many psychomotor abnormalities because MR/DD is the result of some abnormality in the brain, often affecting the many neuromotor systems as well. It is common for gait and fine motor speed and dexterity to be at least somewhat abnormal. Individuals with Autism Spectrum Disorders often show many stereotypies, such as finger wiggling or hand-flapping, that aid in diagnosis of this condition.

It is important that the clinician know the history of the motoric impairment. For individuals who are intellectually normal, motoric impairment is quite significant in the diagnostic process. For those with MR/DD, abnormalities may be the result of neurodevelopmental impairment rather than significant psychiatric illness. For example, many individuals with Autistic Disorder have been quite hyperactive since early childhood; this hyperactivity then would not necessarily indicate manic excitement in an adult.

The first lines of the MSE of a person with MM/MR might read:

"31-year-old-man, functioning in the mild range of mental retardation, with long face and large, prominent ears, relating in overcompliant manner. He is actively gaze-avoidant, with occasional hand-flapping. There are periods of extreme overactivity."

SPEECH

This section "describes the physical production of speech, not the ideas being conveyed."⁶ Speech impediments, styles, rate and intensity (including "*pressured*" speech typical of mania), amount (including *logorrhea* typical of mania, *underproduction* typical of depression) are all noted. There may be *palatal speech*, typical of syndromes causing cleft palate (e.g., Velocardiofacial Syndrome), or *stuttering*, or *cluttering* (bursts of words or phrases).

For patients with MR/DD, speech may be quite abnormal due to the developmental delay, not due to psychiatric illness. Thus, speaking too

quickly or slowly may be a life-long disability, rather than an indicator of mood or psychotic disorder. Some types of abnormal speech, such as echolalia, are characteristic of Autism Spectrum Disorders, or persons with S/PMR. Persons with Autism Spectrum Disorders may have monotone speech (dysprosody) that has little of the normal inflections and syllabic stresses that most people unconsciously use. The result of dysprosody is *robotic speech*. *Pronominal reversal* (e.g., using “you” instead of “me”) can occur in Autism Spectrum Disorders.

Language comprehension can be tested by asking two or three step requests: “Turn this paper over, fold it in half, and put it on the floor. For example, one person with moderate MR who had difficulty in the workshop was given the three step request. He went completely out of sequence and started by putting the paper on the floor as the first step. Then he stooped over and tried to fold a corner of the paper while it was lying on the floor. Afterwards, the staff commented that his difficulties made much more sense when they witnessed the problems with sequential tasks.

A caregiver should be present to interpret impeded speech. The patient’s comprehension of interviewers’ questions must be checked at this point.

“Speech is pressured, with logorrhea and cluttering. There are occasional ‘robotic’ phrases.”

EMOTIONAL EXPRESSION

Manley⁶ defines mood as “the prevailing emotional state” and affect as “the expression and expressivity of a patient’s emotions” (p. 657). There has long been debate as to the exact meaning of these terms and the ability of an interviewer reliably to assess what is an internal state. Manley proposes that these concepts be replaced with the patient’s subjective report of his or her feeling at the time of interview, and the interviewer’s objective report of apparent mood and expression. The latter may include range, intensity, appropriateness and *lability* (changeableness) of mood.

One method to test the person’s understanding of emotional states is to draw faces (happy, sad, mad, fearful, and neutral). Ask the person to identify the happy face, etc. If the individual can do that, ask to point to the face that shows how he or she feels today. Sometimes,

a person will say he or she is happy when the facial expression is one of sadness. “Happy” may be the response that the person thinks is the expected one. The clinician may comment, “You look sad, but you say you’re happy.” This may be a bit complex for some people, but it allows the person to consider an alternative (i.e., more accurate) response.

The patient with MM/MR may not be comfortable expressing what he or she fears are negative mood states, and may seek permission from caregivers or require encouragement or reassurance from the interviewer, and this is important to report.⁶ Persons with Autism Spectrum Disorders may be unable to label emotions (a symptom called *alexithymia*).

Objectively the patient with MM/MR may appear bland in the expression of mood, or expression may lack nuance. Persons with Autism Spectrum Disorders may seem to lack mood altogether. All of these can be mistaken for the *flat affect* of schizophrenia. The intensity of depression can be missed. Manic intensity, on the other hand, is difficult to miss because of the uncharacteristic nature of this intensity in this population; it may, however, be confused with the more episodic disinhibition or incontinence anger seen in some personality disorders.

“Subjectively he reports feeling ‘fine.’ Objectively emotional expression is full range, with periods of elation and tearfulness; mood is quite labile.”

THINKING AND PERCEPTION

For the neurotypical patient, aberrations in thought and perception are easily recognized. For those with MR/DD, the clinician may be unable to sort out which abnormalities are a reflection of developmental delay, and which represent recent onset of abnormal thoughts and perceptions. Many individuals with MM/MR continue to display behaviors normal in young children, but considered abnormal in adults.³ Talking out loud to oneself is a normal part of speech development, which becomes subsumed as private speech in early childhood. We all talk out loud to ourselves occasionally, but persons with MM/MR do so much more often and even more so when they are upset—as when experiencing a psychiatric disorder. This can be very disconcerting to caregivers, and can be perceived as the patient having hallucinations when he or she is not.

Patients with MM/MR also may engage in extreme fantasy as adults in a way more characteristic of children. We all engage in fantasy and daydreaming. But the person with MM/MR may be overinvolved with TV soap operas, or the World Wrestling Federation, in a way that is disturbing to caregivers. Again, this may be a normal manifestation of the developmental delay rather than psychotic behavior. Similarly, the continuation of an imaginary friend into adulthood is seen among adults with MM/MR, while it is reported quite rarely among neurotypical adults. Clinicians must be extremely careful not to mistake such developmental delay phenomena with psychotic thought.³

Characteristic psychopathological forms of thought are discussed below.

Thought Form or Process “refers to the way in which ideas are linked, not the ideas themselves.”⁶ Thinking may be *concrete*, limited to observable, not inferred, relationships between things; such thinking is typical of persons with MM/MR. *Circumstantiality* (inclusion of irrelevant concrete details) and *tangentiality* (a reply appropriate to the general topic of a question, but which does not answer the question) may be observed; this is also typical of persons with MM/MR and may not be, as it is in persons with neurotypical cognition, indicative of psychopathology. Thoughts may be logically connected and goal directed, or disorganized in ways that preserve logical links between individual thoughts but fail to preserve goal direction (“*Flight of Ideas*”). Ideas may follow each other without apparent connection, or with only nonlogical connections (e.g., connected by rhyme, an intervening thought which is not expressed, or by idiosyncratic link—“*Loosening of Associations*” or “*Derailment*”). *Perseveration* (repetition of an answer, no matter how irrelevant) may be observed.

“*Thinking is concrete, circumstantial, with flight of ideas and perseveration.*”

Other abnormalities of thought form, called Formal Thought Disorders, may be found in Manley⁶ (p. 658, Table 7.1-2), with examples.

Thought Content “describes a patient’s ideas.”⁶ Abnormalities include:

- *Delusions*: Fixed, false beliefs, not shared by others in a patient’s family, subculture, ethnic, national or religious group, rigidly held despite all evidence of their falsity.

No type of delusion is any more than indicative of any disorder; none is diagnostic. *Delusions of guilt* (self-accusation of incredible responsibilities and failures) and *somatic delusions* (delusions of illness) are more typical of major depression, whereas *grandiose delusions* (beliefs that one is of great importance or consequence) are more often seen in mania. *Persecutory delusions* (belief in a conspiracy against oneself or loved ones) may be seen in schizophrenia or mania. All of these may occur in any disorder, or be missed or misdiagnosed in persons with MM/MR.

Persons with MM/MR may express ideas easily mistaken for delusions because they cannot support these ideas with evidence, or may report experiences thought to be beyond their level of ability. For example a man with MM/MR, when asked what he did at his group home, said, “I helped build it.” Only when clinicians discovered that his family and he had participated in the home’s construction did they abandon the idea that this was a grandiose delusion. Only a complete history, and a review of such statements with knowledgeable caregivers, can prevent such errors.

In persons with MM/MR, *grandiose delusions* may take the form of a belief that the person has skills that are typically seen as normal. For example, the patient may believe that he or she can drive a car or cook elaborate meals when he or she cannot. For people of normal intellectual ability, such achievements would not be grandiose, but for someone with MM/MR, they would be. Similarly, delusional guilt may take the form of a belief that one will, or has, run out of money, or bankrupted the family or residential program, that one is a failure at work—all within the realm of possibility and not bizarre in any way. One can only determine the delusional nature of such beliefs by reviewing their basis with knowledgeable caregivers, and observing the patient’s response to confrontation or reassurance.

One must always ask a patient to clarify what he or she means. For example, caregiving staff reported that a 50-year-old man with moderate

MR and a diagnosis of schizophrenia had delusions. They described the delusions as beliefs that he had special powers. At interview the man said he had special powers. When asked if others had special powers, he said, "yes." It turned out that everyone in the room had special powers. Clearly, his concept of "special powers" was not so special—and not delusional.

Somatic delusions may take the form of constant minor complaints and clinging, constant pain, repeated demands to see medical personnel, and other vague complaints suggestive of a *somatization disorder*, rather than the bizarre somatic and somatosensory experiences reported by neurotypical persons with schizophrenia (e.g., a belief that "a cat is living in my belly"). However, somatic delusions in persons with MM/MR and Major Depressive Disorder may take the dramatic form of a belief that the person him or herself is dead or dying, that loved ones and other important persons in the patient's life are dead or dying, that caregivers, friends and partners no longer love or care for the patient. In one instance a woman with MM/MR due to Williams Syndrome had a lifelong history of having two imaginary friends (possibly based on dolls she had once owned), clowns named "Glad" and "Happy." In the course of an episode of Major Depressive Disorder she reported that the two clowns were dead.

Persecutory delusions may take the form of simple insistence that others are "bothering" the patient; there may be none of the bizarre elaboration of conspiracies typical of the schizophrenic who does not have MR/DD. Again, the belief is well within the realm of possibility and can only be seen to be false if checked with caregivers, who may be able to verify that the people "bothering" the patient are nowhere nearby or have never been observed to have any interaction with the patient.

- *Ideas of reference* are beliefs that the innocent gestures of others (scratching, blinking) or such things as radio or TV broadcasts, are signals and communications of significance to the patient personally.

The patient with MM/MR and schizophrenia might believe that an episode of coughing in his workplace means he is going to come to harm. This is only an example of an *idea of reference* if the patient means this is a signal of impending

harm, or a signal to attack him. If it is based on a fear of contagion, it is an *obsession*.

- *Obsessions* are unwanted, repetitive, intrusive thoughts, experienced as beyond one's control. Usually the thoughts are disturbing in some way, for example, fears of harm or impending harm to oneself or a loved one, a fear of doing something embarrassing or inappropriate.

People with MM/MR rarely report *obsessions* spontaneously; they must be actively elicited. They are distinguished from *preoccupations* in that the latter are recurrent thoughts to which the person returns voluntarily. *Obsessions* are involuntary, to the point that persons with MM/MR may attribute the thoughts to others, or accuse others of putting these thoughts in his or her head. This can be difficult to distinguish from a *persecutory delusion*. In the case of delusions the patient tends to focus on the conspiracy, or on the person "bothering" him or her; in the case of obsession the patient tends to focus on the distress engendered by the obsessive thought. They may be experienced as voices giving simple commands (e.g., "Turn off the radio").

Obsessive thoughts may drive a person to action to avoid them; for example, the person with the obsession of contagion might run from the room when faced with another person coughing, or perhaps violently push the other person away. The interviewer must work backward from the violent act to the obsession or delusion behind it.

- *Suicidal and Homicidal Ideation* is, simply, thoughts of killing oneself or others.

The ability of persons with MM/MR who actually kill themselves or others is easily underestimated. Assessment of suicidal or homicidal ideation in a person with MM/MR is no different from such assessment in any other patient. The patient must be asked directly about any homicidal or suicidal threats reported by caregivers, or asked directly about such thoughts if no threats are reported. The patient can be reassured that this is a standard part of the evaluation, a question everyone is asked. If the patient has made such threats or has such thoughts, he or she must be asked how the threat would be carried out and an assessment made with consultation with caregivers, of how realistic the threat is. A threat to use a gun, when the

patient has no access to firearms, is not as immediate a threat as a plan to throw oneself or another from a car or a height, or a plan to overdose with someone's medication, or to hang oneself. Overdose threats or plans are particularly dangerous coming from an individual who may have no idea what would constitute a lethal dose, a fact that has turned many a *suicide gesture* into a completed suicide. Persons who fail to kill themselves by jumping from windows, hanging or other strangulation may cause themselves severe brain and other injury instead.

Even when a suicide or homicide threat poses no real risk due to the patient's level of supervision, it is always a measure of the intensity of distress, and in persons with bland mood, or the moodlessness and alexithymia of Autism Spectrum Disorder, suicidal or homicidal ideation may be the only clue to the intensity of the patient's distress.

"He believes he can get a chauffeur's license and drive a bus; he sometimes expresses the belief that he has such a license, and a job with a bus company. He expresses thoughts of killing anyone who stops him from driving the bus; later he tearfully expresses a wish to die if he cannot be a bus driver."

Abnormalities of *Perception* principally include *illusions* and *hallucinations*.

- *Illusions* are misprocessed or misinterpreted sensory stimuli.

The important thing to note is that there is a real sensory input in which the *illusion* originates. Common *illusions* include such things as mistaking a similar-appearing stranger for a deceased loved one, or mistaking a sound in the environment for someone calling one's name; perhaps more serious is the mistaking of fluttering curtains for monsters. There is no reason to think persons with MM/MR are more than usually prone to illusions, but they may be more prone to act on them, and in ways that can be mistaken for a sign of psychiatric disorder.

Illusions are common in delirium and states of mild intoxication, including intoxication caused by anticonvulsants and some other medications in common use by persons with MM/MR, and in withdrawal states. One often needs a complete history to reliably distinguish an *illusion* from a *hallucination*.

- *Hallucinations* are perceptual experiences generated within the central nervous system; there is no current external sensory basis for the experience. *Hallucinations* can occur in any and all sensory modalities, and with two important exceptions the sensory modality has little diagnostic significance.

Auditory hallucinations are experienced as heard through the ears, not as a voice inside the head. This can be an important feature distinguishing *auditory hallucinations* from the phenomenon of *auditorization of thought*. The latter, also referred to by Szymanski¹⁸ as "soliloquizing," refers to the common experience of persons with MM/MR (and of us all, under sufficient stress) hearing the voices of self, caregivers or admired authority figures from life or fiction (e.g., a deceased parent or grandparent, the Lone Ranger, Superman) encouraging them to "be good," "be brave," or alternatively "don't get angry," or "stay cool." These encouraging voices, heard within one's own head, auditorize the person's own thoughts. The strategy of "reauditorization" is in fact often taught to persons with learning problems of all kinds; it is often discovered spontaneously, and used not only by persons with MM/MR, but by everyone. Persons with MM/MR, however, when asked if they "hear voices," will often report these experiences, which are than mistaken for *auditory hallucinations*.

In contrast, *auditory hallucinations* are experienced as if heard through the ears, and seldom if ever have such benign content. The voice may be described as that of God or the Devil. The content may be bizarre, auditorizing the content of *delusions*, or commenting adversely on the person's life, threatening self or others, or commanding the person to perform acts they experience as not wanting to do (*command hallucinations*), the most serious of which can include *suicidal* or *homicidal command hallucinations*. *Command hallucinations* are as serious a sign in persons with MM/MR as in any other persons, and are good reason to consider hospitalization. Voices of deceased family members, with malign content or dangerous *command hallucinations*, are not to be mistaken for *auditorization of thought*, the content of which is almost invariably benign and encouraging.

Two kinds of hallucinations with special diagnostic significance are *formication* and *olfactory hallucinations*. *Formication*, a tactile sensation described by patients as “ants crawling under my skin,” is a sign of a withdrawal syndrome, from alcohol or other central nervous system depressant. *Olfactory hallucinations* are odors, by far most commonly unpleasant odors, originating in seizure foci or tumors deep in the brain adjacent to the olfactory nerves. They are a premonitory sign, or *aura* of an impending partial complex seizure. When the odor is of fire or burning substances, the person may run; the period of confusion following the actual seizure may be mistaken for a psychotic state. The clinician must think of this possibility whenever there is a history of episodes of brief confusion or combativeness, or of apparently motiveless running; the patient must be asked about *olfactory hallucinations*. Only an EEG with nasotemporal leads can definitively rule partial seizures in or out, but an invariant (that is the identical odor every time) olfactory hallucination preceding behavioral episodes is excellent presumptive evidence.

Visual hallucinations were once thought to be more indicative of structural brain pathology than of psychiatric disorder, but this proves not to be the case. One in seven of Reid’s group of persons with manic depression had visual hallucinations.¹¹ One important exception is the usually invariable (unchanging) *visual aura*, analogous to the *olfactory hallucination*, preceding a seizure. The visual experience can be simple (e.g. patterns of stripes) or quite elaborate (a tiger), or can be elaborated by the patient; the important point is that it is always the same, always moves across the visual field in the same way, and precedes episodes of altered consciousness that are the seizure.

Hallucinations are such dramatic symptoms that they can lead caregivers and clinicians alike to mistaken diagnoses of major psychiatric disorders, principally schizophrenia, despite absence of any other diagnostic criteria, especially when the patient cannot articulate the details of his or her experience. It is important as part of the MSE to ask, “Do you hear voices?” and even more important to elaborate on that question. First, the person should be asked if the meaning

of the question is clear—“voices in your head of people who aren’t there.” If the answer is “yes,” it should be clarified as to whether the voices are heard inside the head or through the ears, whose voice or voices are heard, and what the voices are saying. The person should always be asked how he or she feels about the voices, since *hallucinations* are almost always unpleasant, whereas the *auditorization of thought* is almost always pleasant or encouraging. Next, hallucinations in other sensory modalities should be inquired about; if present, it should be clarified whether these are multiple or invariable, and the person and caregivers should be questioned about “blackouts,” periods of involuntary movement and of confusion, and memory lapses (all possible indicators of partial seizure).

“He describes hearing the voice of God saying he can drive any car in the world; he also hears his mother’s voice in his head saying to be careful, and not to steal a bus or a car.”

SENSORIUM

The standard MSE includes gross assessments of intactness of central nervous system functioning in order to differentiate psychiatric disorders characterized by *change in level of consciousness*, usually a sign of delirium, dementia, intoxication, or structural brain lesion.

- *Alertness* refers to the person’s place on a scale ranging from fully awake to comatose. Alertness may be *stable* or *fluctuating*; the latter is more suggestive of an acute intoxication or metabolic disturbance. Intoxication, again, may be caused by medications to which people with MM/MR are frequently exposed, and by metabolic abnormalities, as well as by alcohol or drugs of abuse.

- *Orientation* refers to the person’s understanding of who and where he or she is, and when in time the interview is taking place. People with MM/MR of course know who they are (*orientation to person*), but persons with an Autism Spectrum Disorder may refer to themselves only by name, or in the third person. A person with MM/MR may know he or she is in a hospital or doctor’s office, but may not be able to identify the exact hospital (*orientation to place*). The person may know his or her address or the name of his or her group home. Orientation to time depends on

the degree of independence vs. supervision in a person's life. A person with MM/MR may or may not know the exact date, but may know the month, the day of the week, whether it is a weekday or a weekend day, the season, the next major holiday, or his or her own birthday. A very significant finding on MSE is a change in orientation; e.g., a person who once could identify his or her exact location no longer can do so.

- *Concentration* is the ability to sustain attention. *Distraction* (by extraneous stimuli) may be noted. Formal testing of concentration is usually done by asking the person to count backwards by 7's or 3's from 100 ("serial 7's"). This may be beyond the ability of most persons with MM/MR; instead, they can be asked to count backwards from 100, or 20. Sometimes, the concept of counting backwards is confusing. The interviewer can start the sequence with "I'm going to count backwards, beginning from 25: 25,24,23,22,21,20. Please continue from 20 down to one." One can quantify problems in concentration by timing how long it takes to count backwards or to say the months of the year forwards (or backwards). Concentration can be assessed in the course of a sustained interview as well. Patients might be asked to perform a simple, manual repetitive task in the office to assess concentration. The point is it must be a sustained task.

- *Memory* is divided into three functions: *immediate recall*, *recent memory*, and *remote memory*. *Recall* can be tested by having the person repeat sequences of from 3 to 6 numbers forwards and backwards (normal recall is 5 to 6 forward, 3

cognitive function and distractibility). with level of cognitive function and distractibility). *Recent memory* can be assessed by giving three or four unrelated objects (car, apple, umbrella), asking the person to repeat them, then asking the person to remember them 5 and 10 minutes later. Alternatively, the person can be asked what he or she ate at the last meal. Many people with MM/MR will require reassurance that these are not tests "like in school"; they are "just memory tests." *Remote memory* can be assessed in the course of interview with questions about events two or more years in the past, or in childhood. Persons with Autism Spectrum Disorders may

exhibit *hypermnesia*, extremely exact and detailed memories. Verbal memory items are very difficult for people with MM/MR. Asking him or her to remember ball, cat or shoe might be possible. More practically, a possession or interesting object might be hidden in the office in front of the patient, calling his or her attention to the object and its whereabouts; after 20 minutes, the patient will be asked to find it. Alternatively, the patient can be asked to copy a triangle then asked to draw the triangle from memory after a few minutes.

- Remaining elements of the MSE, *calculations*, *fund of knowledge*, *abstract reasoning*, *insight* and *judgment*, are of limited usefulness with persons with MM/MR because of the lack of formal investigation of what the norms of functioning should be for a person with MM/MR. Changes in any of these areas should be noted in the history as being of possible significance; loss of abilities or worsening judgment could be due to acute onset of many psychiatric disorders, intoxication, dementia, or brain trauma. Fund of knowledge testing may include asking if the person knows the name(s) of the medicine(s) he or she takes. Judgment can be tested by relating the questions to everyday issues. "What do you need to do to get a job, an apartment, a girlfriend or boyfriend," etc.

"The patient is alert and awake throughout the interview. He is oriented to person, to place and to day of the week. He is easily distracted. Recent and remote memory are intact."

The complete MSE of the patient described in the sections above would read:

"31-year-old man, functioning in the mild range of mental retardation, with long face and large, prominent ears, relating in overcompliant manner. He is actively gaze-avoidant, with occasional hand-flapping. There are periods of extreme overactivity. Speech is pressured, with logorrhea and cluttering. There are occasional 'robotic' phrases. Subjectively he reports feeling 'fine.' Objectively emotional expression is full range, with periods of elation and tearfulness; mood is quite labile. Thinking is concrete, circumstantial, with flight of ideas and perseveration. He believes he can get a chauffeur's license and drive a bus; he sometimes expresses the belief that he has such a license, and a job with a bus company. He expresses thoughts of killing anyone who stops him from driving the bus; later he tearfully

expresses a wish to die if he cannot be a bus driver. He describes hearing the voice of God saying he can drive any car in the world; he also hears his mother's voice in his head saying to be careful, and not to steal a bus or a car. The patient is alert and awake throughout the interview. He is oriented to person, to place and to day of the week. He is easily distracted. Recent and remote memory are intact."

The MSE describes a man with the overcompliance, auditorization of thought, and partial time-orientation characteristic of the person with MM/MR, the overactivity, pressured speech, logorrhea, elation, lability and intensity of mood, grandiosity and auditory hallucinations of bipolar disorder, manic type with psychotic features, and the long face, large, prominent ears, gaze-avoidance, hand-flapping, and "cluttering" of speech and "robotic" speech possibly consistent with Fragile X syndrome. Even without a history, an MSE can sometimes be quite revealing. And while one might be able to make a diagnosis of mania from history alone, one would be unlikely to consider the possibility of Fragile X syndrome without observing the patient directly.

MAJOR ALTERATIONS IN THE MENTAL STATUS EXAMINATION OF THE PERSON WITH SEVERE OR PROFOUND MENTAL RETARDATION

The MSE was never conceived with the needs of people with S/PMR in mind. However, any organized observation of a patient with S/PMR can be devised. The advantage of adapting and following, as much as is possible, the standard format of the MSE is that by doing so we take another step towards bringing psychiatric care of persons with S/PMR into the mainstream of mental health practice.

For persons with S/PMR, few settings could be more unnatural than the standard one-to-one psychiatric interview. Almost invariably a caregiver is present in every aspect of their lives, and to be left alone with a stranger can be extremely frightening, distorting the actual mental status and needlessly skewing the observation in the direction of pathology. There is no reason not to have a caregiver present; on the contrary, part of the MSE of a person with S/PMR would be differences between relatedness with a familiar caregiver and with the interviewer. Even more so than with a person with MM/MR, one would be

prepared to observe the person with S/PMR in any setting: home, school, work, institutional day-room, clinician's waiting room, or a van or other familiar setting the person will not leave.

Persons with S/PMR are usually characterized by limitations in communication rendering uncertain, or when speech is completely lacking, impossible, any access to internal experience. The person cannot describe *mood* reliably, cannot give information on *hallucinations*, *delusions* or *obsessions*. Thus the standard MSE must be modified for persons with S/PMR. The clinician may be limited to the parts of the MSE which depend strictly on observation. The MSE serves as a check upon or confirmation of the observations of caregivers reported in the history. It is the place to observe the patient for stigmata of mental retardation syndromes and other medical problems.

Persons with Autism Spectrum Disorders may be classified as having S/PMR due to a lack of responsiveness to standardized testing, or to communication limitations that do not, however, limit access to internal experience. One of the chief purposes of the MSE in persons with S/PMR is diagnosis of individuals with Autism Spectrum Disorders who may not have been previously diagnosed as such; numerous behavioral problems typical of persons with Autism Spectrum Disorders mimic or are mistaken for signs of psychiatric disorder. Fortunately much of what leads the clinician to a suspicion of a diagnosis of an Autism Spectrum Disorders is observable at the MSE.

GENERAL APPROACH

APPEARANCE

One would begin the MSE as one would with a patient with MM/MR, noting age, range of cognitive functioning, and dysmorphisms suggestive of genetic syndromes if any. Degree of mobility (is the patient independently ambulatory, in need of a support device, or needing a wheelchair for ambulation) and sensory handicaps if any should be recorded. Often persons with S/PMR are so classified because sensory or motor handicaps prevent them from responding to standardized cognitive testing; tests adapted for persons with sensory handicaps are available.

PSYCHOMOTOR BEHAVIOR

As with a patient with MM/MR, one would note *level of activity* and involuntary movements (e.g., *tics* or *stereotypies*, *tremor*, *choreoathetosis*, *paresis* or *spasticity*). Overactivity is more typical of manic states, hypoactivity of depression. Caregivers can confirm whether the observed state is typical or atypical for the patient.

One type of involuntary movement for which the clinician should be on constant alert is the stereotyped series of movements of head, neck, eyes and sometimes arms and trunk, accompanied by change in level of consciousness, typical of partial simple or partial complex seizures.

"This is a 37-year-old African-American female, legally blind on the left, with cataract on the right, partially hearing-impaired and wheelchair-bound due to spastic paraparesis. There is kyphoscoliosis as well. She is underactive and covers her eyes."

RELATEDNESS

Relatedness, for lack of a better term, is the quality, style and intensity of the patient's interaction with others in the immediate environment. One begins with observation of interaction with familiar caregivers, observing for eye-contact, spontaneous seeking of interaction or comfort, and response to caregiver's approaches. Persons with Autism Spectrum Disorders avoid direct eye-to-eye gaze. This can be difficult to assess in persons with severe degrees of blindness. A person with S/PMR may or may not respond to speech; unless deaf, however, most persons with S/PMR will respond to his or her name or to a familiar voice. A person with S/PMR may respond warmly to affectionate approach by a caregiver, and seek to prolong the interaction; the exception to this is the *touch-avoidant* patient with Autism Spectrum Disorders. In cases of abuse or past abuse, a patient may have a fearful response to the caregiver's approach or voice. It can be difficult to appreciate the difference between fearfulness and *touch-avoidance*; such an observation must be integrated with the rest of the history and MSE, and there is still the risk of misinterpretation. A withdrawal response to approach may signal depression, especially if accompanied by eye- or head-covering.

Next, the interviewer should seek to engage the patient. A standard offer to shake hands while introducing oneself provides observation of response to eye-to-eye gaze, auditory and tactile stimuli. The response may be the same as to a familiar caregiver, or different. The patient may be *responsive*, and shift attention to the interviewer, or *aloof* to the approach, and ignore the interviewer entirely. If the patient avoids eye-to-eye gaze, an attempt should be made to engage the patient face to face; the patient may be inattentive (*passive gaze-avoidance*) or may actively seek to end the interaction (*active gaze-avoidance*). Aloofness and gaze-avoidance is most typical of persons with Autism Spectrum Disorders. Initial responsiveness and active withdrawal should suggest the possibility of depression. Irritable rejection of interaction could signal Autism Spectrum Disorders, depression, or both.

- *Ability to engage* can be assessed by simple interactive play, with a ball or plush toy. The interviewer can quickly determine whether the patient rejects interaction, engages passively, actively seeks to prolong the interaction, or engages in *reciprocal interaction*, (e.g., "plays catch" with a ball, or hands a toy back and forth). Few persons with Autism Spectrum Disorders will seek to prolong a play interaction; fewer will engage in reciprocal play. Persons with depression may withdraw after brief interaction. Persons with mania may be unable to sustain play interaction, or may display disorganized response.

- *Ability to learn* can be assessed by using an oral reinforcer to teach a simple task, such as greeting. Persons with Autism Spectrum Disorders may be more responsive to this sort of interaction than to a play interaction.

It is important to note if the patient tries to hit, pinch or kick others during the exam and the context in which the aggression occurs. One individual with Autism Spectrum Disorders had a dramatic decrease in aggression. The clinician extended a hand to congratulate the individual and she tried to pinch him. Then she continued to try to pinch. It appears that the person interpreted the clinician's attempt to shake hands as an assaultive gesture that invaded her space.

"She spontaneously seeks her caregivers' hands; when approached by them she responds and seeks to prolong

the interaction, but soon withdraws. Gaze-avoidance is difficult to assess due to her limited vision, but she orients to sound and engages briefly in simple interactive play. She withdraws and covers her eyes when this is prolonged."

SPEECH/VERBAL OUTPUT AND OTHER COMMUNICATION

Many persons with S/PMR have some speech. There may be communicative speech or various kinds of stereotypy: sounds, single words, complex phrases, perseverative or echolalic speech.

- *Vocal stereotypy* are sounds other than language. The sounds may be repetitive, and may or may not have meanings recognized by caregivers.

- Single words and complex, or at least multi-word, phrases; these may or may not have meaning or communicative value recognized by caregivers.

- *Perseverative speech* consists of repetitive words or phrases, used in multiple contexts and usually without communicative intent or value. An exception is the *perseverative questioning* seen in many persons with Autism Spectrum Disorders; this is usually either a compulsion, when the question is meaningful (e.g., a person informed of an impending event might repeatedly ask about when it was going to happen), or a signal of anxiety, when it is not (e.g., a person might repeat "Mommy's coming?" whenever approached).

- *Echolalia* is the repetition of all or part of what was just heard, instead of a response (e.g., a person asked "What's your name?" replies "Your name"). *Delayed echolalia* is repetition, tape-recorder like, of words or phrases heard in the past. Both are typical of persons with Autism Spectrum Disorders. Phrases may include directions, orders, angry responses, or reassurances and pet phrases, heard in the person's past; whether in the distant or immediate past, and of what significance, may be impossible to determine.

The patient's verbal output may be a mixture of any or all of the above, plus communicative speech.

The patient may have gestural communication understood by caregivers, or may have a

vocabulary of standard sign language signs. There may be mixtures of noncommunicative verbal output of the above types and communicative nonverbal speech.

"The patient displays vocal stereotypy and a few single words, one used communicatively. She has gestural communication understood by caregivers."

EMOTIONAL EXPRESSION

Mood may be difficult to assess in persons with S/PMR. Subjective mood is seldom accessible; even persons with Autism Spectrum Disorders who may have language exhibit *alexithymia* (inability to describe mood). Mood expression is most often attenuated. Facial expression may be distorted by dysmorphism or paresis. Persons with some types of cerebral palsy (athetoid or pseudobulbar palsy) may have exaggerated emotional expression. Persons with Autism Spectrum Disorders may have brief, apparently unmotivated outbursts of laughter or tears (*"unexplained affective displays"*). Still, tearfulness, withdrawal, elated or euphoric laughter, irritability and lability may all be observed, and compared to history of mood expression in the past.

"Patient becomes irritable and finally tearful as the interview progresses."

THINKING AND PERCEPTION

Thought content and sensory response are beyond the ability of persons with S/PMR to report. Sensory responses and at least one form of thought can, however, be observed.

- In persons with S/PMR and intact sensory systems, one may test simple sensory responses: visual tracking, auditory locating (turning of the head in response to sound), and response to vibratory sensation. Persons with Autism Spectrum Disorders may avoid visual stimuli, and may respond inconsistently to auditory stimuli, but respond enthusiastically to vibration and seek to prolong vibratory stimuli. Persons with Autism Spectrum Disorders may also smell and mouth objects.

- Persons with S/PMR cannot report *obsessions*, but can in the course of observation demonstrate *compulsions*. These may include picking at skin or threads, getting up to empty

trash, picking up tiny pieces of paper or dust, tearing or repeatedly readjusting clothing, or playing stereotypically with some preferred object. This can be demonstrated by providing objects or situations known to elicit the behavior.

“When active even briefly, the patient picks at threads in her sleeves; when not prevented, she puts these in her mouth.”

SENSORIUM

For persons with S/PMR, level of alertness can also be assessed by observation.

- *Orientation* is usually assessed through discourse, but for persons with S/PMR or Autism Spectrum Disorders, this may be inferred through observation of behavior during the exam. Does the person seem to know preferred caregivers? Do they react to the new mental health clinician appropriately (as a new person)? Only by caregiver report can orientation to time be assessed, as the person seems to behave appropriately for general known times of the day, such as morning, afternoon, meal times, and nighttime.

- *Concentration* can only be inferred from caregiver report. Does the person conduct his or her level of activities of daily living (ADLs) as always, or is there a change? Does he or she seem to react to environmental events as if they are distracting or disturbing?

- *Memory* can be assessed by caregiver report as well. If the person is in a familiar environment, does he or she seem to know where his or her room is, where meals are served, and other geographic locations? In the office, the mental health clinician may also attempt to hide an interesting object, and assess if the person can find it on delayed recall.

- The remaining elements of the MSE, calculations, fund of knowledge, abstract reasoning, insight and judgment cannot be assessed appropriately in persons with S/PMR.

USE OF DATA AND SCALES

Given the major limitations in standard assessment practices for persons with S/PMR, it is recommended that clinicians use formal scales and data collection devices. These may be published forms, or those developed by individual clinicians and hospitals. There are no scales that

sufficiently capture the full state of possible psychiatric illness in persons with MR/DD, but the completion of stand scales will offer the clinician valuable observations of caregivers in a standard format. The Reiss Scales, Aberrant Behavior Checklist, and PAS-ADD package have been used by many for initial assessments and later follow-up measures.^{1,3,4,8-10,12} Reports of ABC sheets, a Bipolar Mood Chart or Sleep Data forms are extremely useful, as these patients cannot report their symptoms.¹⁶

The complete MSE of the patient described in the sections above would read:

“This is a 37-year-old African-American female, legally blind on the left, with cataract on the right, partially hearing-impaired and wheelchair-bound due to spastic paraparesis. There is kyphoscoliosis as well. She is underactive and covers her eyes. She spontaneously seeks her caregivers’ hands; when approached by them, she responds and seeks to prolong the interaction, but soon withdraws. Gaze-avoidance is difficult to assess due to her limited vision, but she orients to sound and engages briefly in simple interactive play. She withdraws and covers her eyes when this is prolonged. The patient displays vocal stereotypy and a few single words, one used communicatively. She has gestural communication understood by caregivers. Patient becomes irritable and finally tearful as the interview progresses. When active even briefly, the patient picks at threads in her sleeves; when not prevented, she puts these in her mouth.”

It describes a woman with a cataract consistent with Congenital Rubella Syndrome; her age is consistent with this as well, pointing to birth during the Rubella epidemic of the early 1960’s which was particularly virulent. She shows every sign of warm relatedness, attenuated by withdrawal perhaps due to depression. Tearfulness, irritability and compulsive picking of threads might also be consistent with Major Depressive Disorder.

CONCLUDING REMARKS

The MSE is a crucial part of the evaluation of any patient, providing clues to diagnosis beyond what is found in the history and serving as a check upon or confirmation of caregivers’ reports and observations. In the case of patients with MM/MR, the standard MSE format can be usefully followed, although some observations may differ in significance from similar findings in

patients who do not have MR/DD, and some phenomena may take a different form.

For patients with S/PMR the MSE format must be altered significantly to be adapted to their needs and the range of phenomena they display. It is nevertheless a useful way to organize and standardize the priceless data of patient observation.

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