Practice Parameters for the Assessment and Treatment of Children, Adolescents, and Adults With Mental Retardation and Comorbid Mental Disorders

ABSTRACT

Mental retardation (MR) is a heterogeneous condition defined by significantly subaverage intellectual and adaptive functioning and onset before age 18 years. With an approach underscored by principles of normalization and the availability of appropriate education and habilitation, persons with MR generally live, are educated, and work in the community. Mental disorders occur more commonly in persons with MR than in the general population. However, the disorders themselves are essentially the same. Clinical presentations can be modified by poor language skills and by life circumstances, so a diagnosis might hinge more heavily on observable behavioral symptoms. The diagnostic assessment considers and synthesizes the biological, psychological, and psychosocial context of mental disorders. Comprehensive treatment integrating various approaches, including family counseling, pharmacological, educational, habilitative, and milieu interventions is the rule. *J. Am. Acad. Child Adolesc. Psychiatry*, 1999, 38(12 Supplement):5S–31S. **Key Words:** dual diagnosis, developmental disabilities, mental illness, psychiatry, mental health, practice parameters, guidelines, children, adolescents, adults.

In the past several decades, the patterns of service delivery for persons with mental retardation (MR) have changed dramatically. At present, an overwhelming majority live in the community and are expected to use community-based medical services. Yet barriers to successful community living exist. Mental disorders are more prevalent in persons with MR than in the general population and are a primary reason for failure to adapt to community living. Furthermore, relatively few mental health clinicians receive training specific to the needs of this population. The goal of these parameters is to aid clinicians who are called upon to provide mental health services to persons who have mental disorders comorbid with MR.

The primary focus of these practice parameters is on children, adolescents, and young adults up to 21 to 22 years of age,

an upper age limit of eligibility for public special education and related services in some states. Issues related to older individuals with MR are also highlighted to provide an understanding of the natural history of MR.

Mental retardation is not a single entity, but includes a heterogeneous group of individuals with a broad spectrum of levels of functioning, disabilities, and strengths. Therefore, these parameters are general in nature and have to be adapted to a particular person's needs.

These parameters consist of 2 major sections:

Part I: Review of General Knowledge on MR. Mental health clinicians usually are not the primary professionals responsible for the diagnosis and treatment of MR. However, the understanding of the biological basis and nature of the MR, its natural history, and the availability of and entitlement for services is necessary for every mental health clinician working with these individuals.

Part II: Mental Disorders Comorbid With MR. This section describes the modifications of psychiatric diagnostic and treatment techniques that may be necessary for patients with significant cognitive impairment. It also details some variations in the clinical presentation of major mental disorders in this population and the principles of psychiatric interventions including general treatment and habilitation programs.

EXECUTIVE SUMMARY

GENERAL ASPECTS OF MENTAL RETARDATION

The diagnosis of MR requires the finding of significantly subaverage current intellectual functioning, for example, IQ

below 70 to 75 on a standardized, individually administered test; significant impairment in present adaptive functioning; and onset prior to age 18 years. Its presence is noted on Axis II in the multiaxial classification scheme. MR is not a single disorder but a heterogeneous condition defined by a person's functioning. Its prevalence is estimated at about 1% of the population, and about 85% of those with this condition have IQs within the mild MR range. In about 35% a genetic causation is found, and in fewer than 10% a malformation syndrome of unknown origin may be identified. External, prenatal, perinatal, or postnatal factors including infections, trauma, toxins, delivery problems, and prematurity account for about one third. In the remainder the etiology is unknown.

The cause of the MR should be identified if possible, as it may clarify the prognosis, sometimes suggest treatment, or alert the clinician to possible medical and behavioral complications that may be more common in certain conditions.

Assessment of Mental Retardation

The assessment of a person with MR is typically multidisciplinary. Increasingly, developmental disorders clinics are established in which psychologists may perform cognitive assessment, developmental pediatricians and clinical geneticists may complete physical diagnostic evaluations, and psychiatrists and behavioral psychologists may assess psychological and behavioral function. University-Affiliated Programs (Appendix) have been established throughout the United States to bring together such expertise, but available resources will influence the roles of various clinicians in the evaluation process. The comprehensive assessment includes:

- Diagnosis of MR using standardized intelligence testing and evaluation of adaptive skills through testing or clinical evaluation.
- Biomedical evaluation, including family, pregnancy, perinatal, developmental, health, social, and educational history; physical and neurodevelopmental examination; and laboratory tests. Laboratory tests are usually indicated by the findings in the history and physical examination and may include chromosomal analysis (including fragile X by DNA analysis), brain imaging (computed tomography, magnetic resonance imaging), EEG, urinary amino acids, blood organic acids and lead level, and appropriate biochemical tests for inborn errors of metabolism.
- Assessment of psychological and behavioral functioning.

Habilitation and Treatment of Persons With Mental Retardation

The habilitation of persons with MR is based on the principles of normalization and community-based care, with additional supports as needed. Federal legislation, for example, the Individuals With Disabilities Education Act (IDEA), entitles disabled children and adolescents to a full range of diagnostic, educational, and support services from birth to age 21 years. Specialized treatments are also provided if necessary, as is done for persons with severe visual and auditory impairment. Additional entitlements may be provided by state laws. The parents of children and adolescents with MR are entitled by these laws to receive support services and to be active participants in treatment planning. Some parents and older patients are not aware of their rights to obtain services. The clinician has an important role in such instances to educate and, if needed, to refer to a "patient advocate" or "educational advocate." In recent practice, children and adolescents are educated in special classes in regular school or in inclusionary programs (in age-appropriate regular classes, with additional supports as needed). In the United States, children with MR are now rarely (if ever) placed in residential institutions and separate schools. Adults with MR of all levels live in the community, in settings varying from their own apartments with supports as needed, to small (4–8 residents) group homes. They are employed in specialized settings or, increasingly, in the competitive job market. Habilitation and treatment include:

- Specific treatment of the underlying condition, if known, to prevent or to minimize brain insults that result in MR (e.g., shunting in the case of hydrocephalus).
- Early intervention, education, and ancillary therapies (such as physical, occupational, and language therapies), family support, and other services, as needed.
- Treatment of comorbid physical conditions, such as hypothyroidism, congenital cataracts or heart defects in children with Down syndrome, treatment of seizures in persons with tuberous sclerosis, etc.
- Treatment of comorbid mental disorders.

ASSESSMENT OF MENTAL ILLNESS IN PERSONS WITH MENTAL RETARDATION

Mental illness is frequently comorbid with MR, with prevalence estimates ranging from 30% to 70%. Virtually all categories of mental disorders have been reported in this population. An accurate psychiatric diagnosis provides the foundation for understanding the patient and for treatment planning.

The psychiatric diagnostic evaluation of persons who have MR is in principle the same as for persons who do not have retardation. The diagnostic approaches are modified, depending on the patient's cognitive level and especially communication skills. For persons who have mild MR and good verbal skills the approach does not differ much from diagnosing persons with average cognitive skills. The poorer the communication skills, the more one has to depend on information provided by caregivers familiar with the patient and on direct behavioral observations.

The assessment includes the following components.

Comprehensive History

The history taken from the patient and from several caregivers in different settings covers:

- Presenting symptoms including concrete descriptions of specific behaviors in various situations and settings, their change over time, antecedent events, and the way the various caregivers handle the behaviors.
- Psychiatric review of systems including premorbid and current behavioral and personality patterns, adaptive functioning, self-care, communication, and social functioning.
- Details of previous psychiatric treatment, with particular emphasis on medication side effects that could cause the presenting symptoms.
- Past and present educational and habilitative interventions, work programs, and living situation: their quality, consistency, and appropriateness; availability of supportive services; and long-term plans for the patient's care.
- Parents'/caregivers' attitudes toward the patient, their understanding of his/her disability, and support for growth versus overprotection.
- Review of past cognitive tests and evaluations or request for new ones if needed.

Patient Interview

Ample time must be allotted for the patient interview, which typically takes longer than with patients without MR. Sufficient time is needed to put the patient at ease.

The verbal examination should be adapted to the patient's communication skills and should use clear and concrete language, structure, reassurance, and support. Leading questions and questions requiring yes or no answers should be avoided, and the interviewer should ensure that questions are understood.

Patients with sensory impairments such as blindness or deafness must be approached in a manner that recognizes their needs through the use of appropriate interpreters or communication devices.

Mental status may be assessed in the context of conversation, rather than in a formal examination. It is often helpful to start the interview with a discussion of a patient's strengths and interests, rather than problems, and later focus on the patient's understanding of disability, limitations, and reasons for the referral.

Nonverbal aspects of the interview include observations of performance on selected tasks, relatedness, expression of affect, impulse control, attention span, activity level, and the presence of unusual behaviors or seizures.

Medical Review

This review should include developmental and medical history, past etiological assessments, and coexisting general medical disorders and their treatments. The latter is particularly important, since undiagnosed medical conditions are frequent in this population and may lead to behavioral symptoms.

Diagnostic Formulation

Data from the assessments should be interpreted in light of developmental level, communication skills, associated handicaps, life experiences, education, and family and sociocultural factors. A particular behavior may suggest an underlying mental disorder if it is part of a pattern of a defined mental disorder syndrome. The possibility of sexual or other abuse that the patient cannot report should be considered. A *DSM-IV* diagnosis (in addition to MR) should be made if the appropriate criteria are met. The diagnostic statement should include a description of the person's strengths, deficiencies, and needs including intellectual, adaptive behavior, communication, health, and psychosocial domains. A comprehensive assessment should yield a multiaxial diagnostic formulation with appropriate differential, and supporting evidence for diagnoses should be highlighted.

SPECIFIC DIAGNOSIS OF COMMON COMORBID MENTAL DISORDERS

Pervasive Developmental Disorders

The majority of children with pervasive developmental disorders (PDDs) also have MR. However, children with MR alone do not have significant impairments in reciprocal social interaction and can engage in social communication, verbal or nonverbal (such as gestures and eye contact), appropriate to their developmental level.

Attention-Deficit/Hyperactivity Disorder

The diagnostic criteria for attention-deficit/hyperactivity disorder (ADHD) are based on observable behavior as reported by multiple informants and thus can be applied to nonverbal children. ADHD should be differentiated from situation-specific inattentiveness, such as at school if the academic expectations are too high, and medication side effects. In assessing "noncompliance"—not following commands of caregivers—one should consider the child's ability to understand social rules and the presence of sufficient skills to communicate opposition.

Tic Disorders and Stereotypic Movement Disorder

In Tourette's disorder, the movements, as opposed to self-stimulatory stereotypies seen in persons with severe MR, are less complex and appear involuntary. For the latter, the diagnosis of stereotypic movement disorder may be used if other mental disorders are excluded. The specifier "with self-injurious behavior," is added if bodily damage results. Self-injurious behavior is common in certain MR syndromes, especially Lesch-Nyhan syndrome.

Mental Disorders Due to a General Medical Condition

MR, in and of itself, does not constitute a medical condition to which aberrant behavior or emotional disturbance

should be ascribed. The attribution of behavioral or emotional disturbance to a general medical condition should be used only when there is evidence from history, physical examination, or laboratory findings that the disturbance is a direct consequence of a specific medical condition. For example, in the case of Down syndrome, hypothyroidism may present as symptoms of depression. In such cases, the medical disorder to which the depression should be attributed is hypothyroidism, not Down syndrome.

Schizophrenia and Other Psychotic Disorders

Schizophrenia can be diagnosed in the usual manner in verbal persons with mild MR, but rarely if at all in persons with more severe retardation. For the latter, the less specific diagnosis of psychotic disorder not otherwise specified (NOS) may be made, if behavioral features such as grossly disorganized behavior and negative signs are present, but were absent in the premorbid period. Conversation with an imaginary friend should not be confused with hallucinations.

Mood Disorders

Mood disorders, especially depressive disorders, are quite common in persons with MR. In verbal persons with mild MR the complaints are simple and concrete. History obtained from caregivers and evidence of neurovegetative signs help in assessing the mood change. Depression may also be manifest by aggressive behavior. Environmental events, such as a precipitous move to a new setting or change in care provider, may trigger a depressive episode. Medication side effects should be considered, for example, depression resulting from β-blockers or agitation associated with akathisia from a neuroleptic drug.

Anxiety Disorders

Verbal persons with mild MR can report on subjective feelings of anxiety; in nonverbal ones symptoms such as avoidance behaviors and agitation might suggest the diagnosis. The tendency toward anxiety and social avoidance is also a part of the behavioral phenotype of fragile X syndrome.

Posttraumatic Stress Disorder

Posttraumatic stress disorder (PTSD) in persons with MR might be quite frequent and should be routinely considered in the differential diagnosis. These individuals are vulnerable to abuse because of difficulties in reporting it and a tendency to want to please others.

Obsessive-Compulsive Disorder

The diagnosis of obsessive-compulsive disorder (OCD) may be difficult in nonverbal persons who cannot report on obsessional thoughts underlying their compulsions. Some repetitive behaviors, for example, hoarding objects, flicking lights on and off, and tidying and arranging, all have been suggested as indi-

cations of OCD in persons with MR. Self-restraint, for example, insistence upon wearing a helmet or other protective device, is a behavior that has been described in persons who also exhibit self-injurious behavior and might suggest the ego-dystonic nature of self-injury. A connection between some self-injurious behaviors and OCD has thus been postulated.

Eating Disorders

Anorexia and bulimia nervosa are relatively rare in the context of MR, particularly moderate to severe MR, but MR is a predisposing factor for other eating disorders such as pica and rumination. The ingestion of nonnutritive substances, pica, and the regurgitation and rechewing of food, rumination, occur with greater frequency as the severity of cognitive disability increases. When these behaviors are a focus of clinical attention, the diagnoses should be indicated.

TREATMENT

The principles of psychiatric treatment are the same as for persons without MR, but modification of techniques may be necessary according to the individual patient's developmental level, especially communication skills. Medical, habilitative, and educational interventions should be coordinated within an overall treatment program. The mental health clinicians should actively participate with other professionals in the development of the various treatment interventions.

The Behavioral Emergency

When the clinician is called to assist with a behavioral emergency, the first task is generally to ensure the safety of the patient and others. For example, in the case of severe self-injurious or aggressive behavior, if the usual attempts at redirection fail and the patient continues to pose an imminent risk, it may be necessary to temporarily use physical restraint. In some instances this may require admission to a psychiatric hospital. The use of emergency medication may also be considered after adequate diagnostic assessment. Medical causes for an acute behavioral exacerbation must always be considered. It is not uncommon for even simple problems such as constipation, infection, or even occult injury to set the stage for behavioral problems. Medication side effects such as akathisia from neuroleptics or disinhibition from sedative-hypnotic agents can be expressed in aggressive and self-injurious behaviors. When a temporizing measure is necessary, it is generally advisable to use a drug with which the patient has had a positive experience, typically a neuroleptic or benzodiazepine. The need for emergency treatment should prompt a comprehensive diagnostic assessment including the evaluation of environmental influences. Approaches should also be considered to prevent recurrence of such emergency situations for a given individual.

Psychosocial Interventions

Persons with MR may benefit from group, individual, and family psychotherapy. Concrete goals should be established, with the overall aim to achieve a maximally feasible quality of life. Disruptive behaviors should not merely be suppressed, but they should be replaced with constructive, adaptive behaviors and skills. Patients should learn to understand their own disability, focus on strengths, and develop a positive self-image, a realistic striving for independence, and age-appropriate social skills. The treatment techniques include focus on current reality, directiveness and structure to maintain focus, and activities adapted to chronological age. A therapist with training in developmental disorders is best equipped to accomplish these goals and to guide the patient to develop his or her own understanding. Therapists should be active, directive, and flexible, perhaps using themselves as examples, and should be prepared to give concrete advice.

Family therapy typically focuses on the parents' identification and support of their child's strengths and independence and the provision of opportunities for success. Parents of recently diagnosed children need careful explanation of their child's condition. Concrete advice in management and resource finding is important, as is help in obtaining educational supports to which the child is entitled under federal and local laws. Parents of adolescents and young adults need help in coming to terms with emergent sexuality and in emotionally separating and preparing them to move to out-of-family living in the community.

Pharmacotherapy

Medication effects generally are not different from those expected in the absence of MR. The adage, "start low, go slow," reflects the observation that shifts in dose-response in certain contexts are far more likely than changes in the mechanism of action of a compound; for example, persons with Down syndrome may be exquisitely sensitive to anticholinergic drugs, and some persons with MR may be more sensitive to the disinhibiting effects of sedative-hypnotic agents.

Several problems with pharmacotherapy are frequently encountered with persons with MR:

- Some clinicians appear to prescribe medication with inadequate information, aiming, for example, for symptom suppression seemingly without consideration of the potential negative impact on habilitative function or overall quality of life. Risks/benefits for medication appear not to be fully considered (e.g., drugs that adversely affect cognition, either directly or through sedation, appear to be used without clear justification and careful monitoring).
- There is no evidence of informed consent to support the prescription or administration of psychotropic medication.
- Medication appears not to be integrated as part of a comprehensive treatment plan (e.g., there is no evidence of a behavior plan; there is no evidence of communication between

prescribing physician and other therapists; behavioral data are not collected).
- Medication may not appear to be appropriate for the diagnosis of record (e.g., a patient with a diagnosis of a mood disorder is receiving only thioridazine).
- Drug exposure appears to be excessive or poorly justified. Medication appears to be prescribed for extended periods for nonspecific indications for which other active treatments or environmental supports are needed. Multiple representatives from the same medication class and other complex polypharmacy regimens are used or no attempt can be seen over time to adjust medication doses to document ongoing need or the minimum dose at which a medication remains effective. In some cases treatment effectiveness is never clearly established.
- No evidence can be found for the active monitoring for emergent side effects, particularly in nonverbal patients.

Treatment Follow-up

A common problem in the treatment of persons with MR is assessing its effectiveness, which may be viewed differently by various caregivers. Therefore, discrete treatment goals should be agreed upon by the clinician and caregivers, as well as target or "index" symptoms. Interdisciplinary collaboration of professionals and caregivers is essential. Various mental health clinicians might function in the team as direct care providers, team leaders, or consultants to other professionals. Among them, clinicians with medically and psychologically oriented training are often prepared to function as synthesizers of treatment modalities of various disciplines. Follow-up includes patient interview/observation and obtaining comprehensive interim information. If the patient is not experiencing improvement, the accuracy and completeness of the biopsychosocial diagnosis should be reviewed, as well as the consistency of implementation of treatment by the caregivers.

LITERATURE REVIEW

The literature review was based on *Medline* and *PsychInfo* searches using key words *mental retardation, mental illness, psychiatry,* and specific mental illness entities; references from major review articles and book chapters; and books on MR and mental illness comorbid with MR. In addition, the authors and consultants brought their cumulative clinical experience from years of work with individuals with MR and related systems of care.

PART I: MENTAL RETARDATION

THE CONCEPT OF MENTAL RETARDATION

Mental retardation does not denote a single disease or entity with a single cause, mechanism, natural course, or prognosis.

TABLE 1
DSM-IV Criteria for Diagnosis of Mental Retardation in Children, Adolescents, and Adults

Criterion	As Evidenced By
Significantly subaverage intellectual functioning	An IQ of approximately 70 or below on an individually administered IQ test (for infants, a clinical judgment of significantly subaverage intellectual functioning)
Concurrent deficits or impairments in present adaptive functioning (i.e., the person's effectiveness in meeting the standards expected for his or her age by his or her cultural group)	At least 2 affected areas: communication, self-care, home living, social/interpersonal skills, use of community resources, self-direction, functional academic skills, work, leisure, health and safety
Onset before age 18 years	

It refers to a heterogeneous behavioral syndrome, characterized by impairments in a person's current level of intellectual and adaptive skills. MR is not necessarily lifelong. Persons who carry this diagnosis present with a wide spectrum of abilities, disabilities, and clinical and behavioral patterns.

DEFINITION OF MENTAL RETARDATION

The tridimensional definition, developed by the American Association on Mental Retardation (AAMR, 1992), is widely accepted: "Mental retardation refers to substantial limitations in present functioning. It is characterized by significantly subaverage functioning, existing concurrently with related limitations in 2 or more of the following applicable adaptive skill areas: communication, self care, home living, social skills, community use, self-direction, health and safety, functional academics, leisure, and work. Mental retardation manifests before age 18" (p. 1). This definition subclassifies MR according to the intensity and nature of needed supports. It emphasizes the need for detailed assessment in all relevant domains, including psychological and emotional. It is the first modern definition that stresses the importance of diagnosing comorbid mental disorders in the assessment of MR.

The *DSM-IV* criteria for MR (American Psychiatric Association, 1994) are summarized in Table 1. The *DSM-IV* subclassifies MR according to its severity as illustrated in Table 2.

MR and its severity are coded on Axis II. The etiology, if known (e.g., trisomy 21, Down syndrome) is coded on Axis III. The current classification does not recognize the past category of "borderline retardation." Instead, the code for "borderline intellectual functioning" (V62.89) may be used if appropriate.

FEATURES OF THE DIAGNOSIS OF MENTAL RETARDATION

Intellectual Impairment

Measures of intellectual function are generally expressed as the ratio of measured to expected performance for age on a standardized test, such as the Wechsler Intelligence Scales or Stanford-Binet. The AAMR and *DSM-IV* definitions of MR both require that the intelligence tests be individually administered. In the choice of cognitive testing instruments, sociocultural background and native language of the individual should be considered to avoid a falsely positive diagnosis of MR in children from cultural and linguistic minorities (*DSM-IV*; American Psychiatric Association, 1994). Measures to that effect have been developed but await validation (AAMR, 1992). The IQ score is a composite of subscores. The profile of these subtests might be much more revealing than the global IQ, as it can reveal both cognitive strengths and weaknesses and thus highlight areas of needed support. For example, a nonverbal person may have a low global IQ despite having adequate learning abilities as evidenced by intact nonverbal scores.

Impairments in Present Adaptive Functioning

Adaptive behavior refers to a person's effectiveness in functioning at the level expected for his or her age and cultural group or setting. Various cultures may also hold their own views regarding the causation, nature, and treatment of disabilities. These factors should be considered and approached in a sensitive manner, and a consultant familiar with a particular culture may be very helpful (Lynch and Hanson, 1992). Both the *DSM-IV* and the AAMR definitions list 10 areas of functioning, and significant impairments in at least 2 of these domains are required for the diagnosis of MR. While there are standardized scales to measure adaptive behaviors, such as Vineland Adaptive Behavior Scale (Sparrow et al., 1984) and AAMD

TABLE 2
Mental Retardation: Classification by Severity

DSM-IV Code	Severity	Approximate IQ Range
317	Mild	55 to approx. 70
318.0	Moderate	35–40 to approx. 50–55
318.1	Severe	20–25 to approx. 35–40
318.2	Profound	Below 20–25
319	Undetermined	Not determined

Adaptive Behavior Scales (Nihira et al., 1974), these measures do not capture all of the above functional domains. Thus, adaptive functioning may be subject to clinical assessment.

The Concept of Developmental Disability

The term *developmental disability* is sometimes used interchangeably with MR, when referring to entitlement to services. However, developmental disability is actually not a medical but a legislative/legal concept referring to a broad spectrum of disorders of development. Originally it included specific diagnoses, such as MR, epilepsy, and autism. The Rehabilitation, Comprehensive Services, and Developmental Disabilities Amendments of 1975 (PL 95-602 of 1978) redefined developmental disability in functional terms, as a severe disability related to mental and/or physical impairment, manifested before age 22 years, and resulting in substantial functional limitation in at least 3 major areas of life activity (Kiernan et al., 1986).

EPIDEMIOLOGY OF MENTAL RETARDATION

Estimates of the prevalence of MR vary, depending on the diagnostic criteria, study design, and methods of ascertainment. When the diagnosis is based on IQ alone, a prevalence of approximately 3% is obtained, but when the current tridimensional definition is used, and with multiple methods of ascertainment, the prevalence in the United States appears to be closer to 1% of the population (reviews by McLaren and Bryson, 1987; King et al., 1999; Roeleveld et al., 1997). About 85% of persons with MR are thought to have mild MR and the remainder, moderate, severe, and profound MR, although these estimates are disputed. The prevalence varies in different age groups, the highest being at school age. The time of diagnosis is also age-dependent, with more severe retardation diagnosed earlier than milder forms. Many of the persons in whom MR is diagnosed in childhood develop adaptive functional skills that are high enough by adulthood that the diagnosis ceases to be appropriate.

With greater severity of retardation comes a higher prevalence of associated disorders. Seizure disorders are seen in 15% to 30% of persons with severe or greater MR, motor handicaps (including cerebral palsy) in 20% to 30%, and sensory impairments (including impairments of hearing and vision) in 10% to 20% (McLaren and Bryson, 1987).

Retardation associated with genetic syndromes, physical disabilities, and general medical conditions with characteristic phenotypical features tends to be diagnosed earlier in life. For example, Down syndrome is usually diagnosed in the neonatal period on the basis of phenotypical features and karyotyping, while mild MR of unknown origin might be recognized only at school age, when learning difficulties become evident.

CAUSATION AND BIOLOGICAL ASPECTS OF MENTAL RETARDATION

Knowledge of the etiology of MR in a particular case is important: it may be treatable and/or preventable; it may be associated with a particular "behavioral phenotype"; or it may be associated with an increased risk for a medical disorder. In most cases the etiology of a disability is multifactorial (Institute of Medicine, 1991).

In 58% to 78% of mild and in 23% to 43% of severe MR, no causation is ascertained with current diagnostic techniques (McLaren and Bryson, 1987). Existing classifications of etiology are based on the timing of the presumed insult to the developing organism, the type of causal factor (AAMR, 1992), or a combination of both approaches as described in Table 3. The frequency numbers for each category vary in different studies, depending on the population and methodology.

NATURAL HISTORY AND COURSE OF MENTAL RETARDATION

MR is a heterogeneous entity without uniform natural history and course. The prognosis or outcome for a given individual typically reflects the interaction of 3 factors: biomedical, psychological, and environmental (Garrard and Richmond, 1965; Szymanski and Wilska, 1996).

Biomedical Factors

The nature of the disorder causing the MR will affect the lifespan and abilities of the individual. Degenerative disorders—for example, certain inborn metabolic disorders—might limit life expectancy, and associated conditions such as epilepsy or motor disabilities may affect a person's functioning and adaptation, regardless of the cognitive level. However, even well-defined biomedical disorders, such as Down syndrome, are not homogeneous and encompass a spectrum of individuals from the severely disabled requiring continuous supervision and care to those who become relatively independent.

Psychological Factors

These factors include the cognitive abilities or, more accurately, the balance between cognitive impairments and strengths. The level of communication skills is important in the ultimate adaptation. Comorbid mental disorders are one of the most important factors influencing adaptation and quality of life.

Environmental Factors

These factors include the availability of educational, habilitative, medical, and other services from as early as possible. The expectations and attitudes of caregivers, especially encouraging and supporting independence as well as socioeconomic opportunities and social exposure, can be important influences.

TABLE 3
Identifiable Causes of Mental Retardation and Estimated Frequency

Subdivision	Example	Pathology	Presentation	Behavioral Features[b]
1. Prenatal causes—genetic disorders[a]: 32%				
1.1: Chromosomal aberrations	Down syndrome	95%: Trisomy 21 (not inherited); 5%: translocation (may be inherited)	Upward slanted eyes, epicanthal folds, simian crease, hypotonia, microcephaly, may have congenital heart defects, cataracts, hypothyroidism	MR: MI to P; behavior is variable, at risk for Alzheimer dementia (but clinical dementia usually not before 50 years)
1.2: Monogenic mutations	Tuberous sclerosis	Single gene dominant	Brain tumors, typical facial skin lesions, seizures	MR: none to P
	PKU	Autosomal recessive; lack phenylalanine hydroxylase	Normal at birth; seizures, vomiting, microcephaly	MR: S to P, if not treated by diet; psychosis in some adults
	Fragile X	X-linked; CGG repeats > 230	Long, narrow face; postpubertal: large testes, large ears	MR: Mi to Mo; gaze aversion, ADHD-like
1.3: Multifactorial	"Familial" MR	Mixed: genetic, environmental, other	Nonspecific	MR: Mi, nonspecific
1.4: Malformation syndromes due to known microdeletion	Williams syndrome	Microdeletion chr 7	Short stature, congenital heart defects, typical facies; full lips, long philtrum	MR: low avg to Mi; outgoing, talkative, language problems
	Prader-Willi syndrome	Deletion on chr 15 of paternal origin	Hypotonia, obesity, short stature, hypogonadism, small hands and feet, almond-shaped eyes	MR: low to avg to S; hyperphagia, irritability, compulsive behaviors, tantrums, skin picking
2. Malformations of unknown causation: 8%				
2.1: Malformations of the CNS	Neural tube defects	May be associated with hydrocephalus	Variable neurological signs and symptoms	MR: variable
2.2: Multiple malformation syndromes	Cornelia de Lange syndrome	Unknown	Hirsutism, microcephaly, prominent philtrum, synophrys	MR: S to P; stereotypies, SIB
3. External prenatal causes: 12%				
3.1: Maternal infections	HIV	Viral encephalopathy	Failure to thrive, infections, microcephaly, neurological deterioration	Variable developmental delay
3.2: Toxins	Fetal alcohol syndrome	Exposure to alcohol in utero	Microcephaly, short palpebral fissures, long philtrum, thin upper lip, congenital heart defects	MR: usually Mi to Mo; irritability, short attention span, hyperactivity, conduct problems
3.3: Toxemia, placental insufficiency	Prematurity	Variable, multifactorial	Growth retardation, variable insult to CNS	Variable
4. Perinatal causes: 11%				
4.1: Infections	Encephalitis	Herpes simplex 2	Variable neurological deficits	MR: S
4.2: Delivery problems	Neonatal asphyxia	Variable, CNS infarctions	Variable neurological deficits, seizures	MR: variable, may be severe, associated with motor deficits, risk for ADHD
4.3: Other	Hyperbilirubinemia	Maternal–child blood group incompatibility	Variable, motor and hearing deficits	MR: variable
5. Postnatal causes: 8%				
5.1: Infections	Encephalitis	Viral	Variable	Variable; ADHD symptoms
5.2: Toxins	Lead poisoning	Exposure to lead (pica)	Acute: weight loss, vomiting, headache; late: seizures, developmental delay	Variable; ADHD symptoms
5.3: Psychosocial	Deprivation	Malnutrition, neglect, abuse	Dependent on causative factor	MR: generally Mi; behavioral problems reflect psychological and environmental causation
5.4: Other	Trauma, tumor	Dependent on nature and extent of causative factors	Dependent on localization of injury to CNS	MR: none to P; behavioral symptoms depend on localization of brain injury
6. Unknown causes: 25%				

Note: These are only illustrative examples, not a full and detailed listing of important MR-associated conditions. Excellent reviews are available (Harris, 1995; Jones, 1997; Szymanski and Wilska, 1996; Szymanski and Kaplan, 1997). The frequency data are estimates based on survey data from Finland (Szymanski and Wilska, 1996). In 4% of the surveyed population, no data were available. PKU = phenylketonuria; MR = mental retardation; HIV = human immunodeficiency virus; chr = chromosome; ADHD = attention-deficit/hyperactivity disorder; SIB = self-injurious behavior.
[a] Characterized by changes in the genetic material, which may or may not have been inherited from the parents.
[b] MR: Mi = mild; Mo = moderate; S = severe; P = profound.

DIAGNOSTIC ASSESSMENT OF MENTAL RETARDATION

Patients with MR are typically referred to mental health professionals because of behavioral/emotional symptoms. Psychiatrists and nonmedical mental health professionals are seldom expected to do the initial diagnosis and assessment of MR as such. However, for some patients MR might have been undiagnosed, and even if the patient is presented as having MR, a critical review of this diagnosis may be indicated. The diagnosis could have preceded the characterization of some MR-associated syndromes, and relevant diagnostic techniques might not have been available. For example, the diagnosis of fragile X syndrome became possible only in the late 1970s (Sutherland, 1977), and certain behavioral patterns may now suggest "behavioral phenotypes" associated with certain syndromes, rather than nonspecific psychopathology (State et al. 1997). A patient who in the past received a diagnosis of psychogenic obesity and MR of unknown origin might actually have Prader-Willi syndrome (Harris, 1995, Vol 2). Some MR-associated conditions are also known to predispose to certain general medical conditions that influence behavioral or emotional symptoms (such as hypothyroidism in Down syndrome, which may present with symptoms of depression). The approach to the patient with MR is summarized in Table 4.

A useful outline for the diagnostic assessment of persons with MR has been described in the diagnostic manual of the AAMR (1992). Medical assessment of children with developmental delays is also reviewed in detail by Levy (1996).

PRINCIPLES OF PREVENTION AND TREATMENT OF MENTAL RETARDATION

Again reflecting heterogeneity, there is no uniform treatment of MR per se. Interventions with persons who have MR include:
- Primary prevention, whenever possible, to prevent the condition that might result in MR. Examples might include abstinence from alcohol and other substances during pregnancy, and immunizations for congenital rubella or measles. Prevention of lead poisoning, folic acid supplementation in pregnancy to prevent neural tube defects, the use of child car seats, the use of bicycle helmets, and good prenatal care are all examples of primary prevention.
- Specific treatment of an underlying condition, if known, to prevent or to minimize brain insult that results in MR: e.g., diet for phenylketonuria (PKU) and shunting in the case of hydrocephalus. These strategies would constitute secondary prevention of MR.
- Provision of early intervention, education, habilitation, and ancillary therapies (such as physical, occupational, and language therapies), family support, and other services, as needed. The goal of these interventions is to minimize functional retardation and maximize a person's abilities (Rowitz, 1986). These approaches would be considered tertiary prevention of MR.

- Medical care to treat and/or prevent general medical conditions that complicate certain MR syndromes and reduce the person's functioning, such as congenital cataracts and deafness in children with Down syndrome, seizures, otitis, or other painful conditions that may induce a nonverbal person to self-injure.
- Treatment and prevention of psychosocial dysfunction (including mental illness). Such consideration is essential because mental disorders are the most important cause of maladaptation (Szymanski, 1987).

The approaches to the treatment and care of persons with MR have changed dramatically in the past 25 years. One common misconception had been that most persons with significant MR lived in institutional settings, since they could not adapt to life in the community and had a deleterious effect on their families. In fact, in this country no more than 10% of persons with MR have ever resided in institutions. Currently, in most states children with MR are not admitted to public institutions at all, and adults only rarely. When they are so admitted, it is usually because of severe mental disorder comorbid with the MR or massive medical needs. Several states have closed all of their large public residential care facilities, and the remaining states have active deinstitutionalization policies. With proper services, the overwhelming majority of persons with MR do well in the community, living with their families or in out-of-family placements, such as family care (foster home), community residences (group homes), and apartments with varying degrees of supports. A minority of children and adolescents might end up in private residential schools, usually because of difficult-to-manage behaviors and lack of adequate support services in the community.

The current philosophy underlying the services for persons with MR is based on the "normalization principle" (Nirje, 1969, p. 181): "making available to the mentally retarded patterns and conditions of everyday life which are as close as possible to the norms and patterns of the mainstream of society." Thus, normalization does not imply making persons with MR "normal," but enabling them to live in as normal conditions as possible.

For children, the normalization approach has focused attention on the right to appropriate education. Public education and related comprehensive services are guaranteed for children with MR and other developmental disabilities by the Education for All Handicapped Children Act (PL 94-142 of 1975). With later amendments it is now referred to as IDEA (Individuals With Disabilities Education Act) and provides for diagnostic, educational, and support services from birth to age 21 years. These services may include early intervention, respite, and specialized treatments if needed (such as for persons with severe visual and auditory impairment). At 3 years of age the child becomes eligible for a full range of educational and related support services. The parents have the right to participate in developing an Individualized Education Program. In general, the educational approach has changed in recent years from that of

TABLE 4
Approach to the Patient With Mental Retardation

Step	Stage	Procedures
Diagnosis of mental retardation	Assessment of intellectual skills; assessment of adaptive skills	Standardized, individual intelligence testing; consider language and cultural aspects; clinical or standardized tests, e.g., Vineland Adaptive Behavior Scales (Sparrow et al., 1984), specific cognitive, vocational, and other relevant instruments
Classification and description	Assessment of personal strengths, weaknesses, and psychopathology	Clinical evaluation by history and patient interview; personality testing, if relevant; formal *DSM-IV* diagnosis
Medical evaluation of etiology	Health status, associated disabilities	History, including family, pregnancy, perinatal, developmental, health; physical and neurological examination; laboratory tests, usually as indicated clinically, may include genetic (including chromosomes and DNA for fragile X), brain imaging (CT scan, MRI), EEG, urinary amino acids, blood organic acids and lead, biochemical tests for inborn errors of metabolism
Medical evaluation of etiology	Evaluation of environmental issues	History, interview of caregivers/service providers, direct observation: focus on physical and psychosocial environments; educational, habilitative, work, and other services
Needed supports	Intellectual/adaptive skills, including communication, self-care, home living, social and interpersonal skills, use of community resources, self-direction, functional academic skills, work, leisure, health and safety	As per initial assessment: identification of nature and intensity of relevant services, therapies, and other supports to maximize strengths and compensate for impairments in each of skill domains
Needed supports	Psychological/psychopathology Health needs Environmental supports	Counseling, psychotherapy, medications Medical and specialty care: treatment of identified disorders; ancillary therapies; prosthetics Education, living environment, work, habilitative, recreational, other milieu supports

Note: Adapted from American Association on Mental Retardation, 1992. CT = computed tomography; MRI = magnetic resonance imaging.

mainstreaming children in regular schools to "inclusion," or placement of all children with disabilities in age-appropriate classrooms. This strategy creates a new challenge for child mental health clinicians since behavioral problems are the chief reason for the failure of such placements.

Besides federal law, the states have additional policies concerning services to persons with developmental disabilities, and clinicians should be familiar with these in order to provide families with effective advice and advocacy. Information relevant to each state's early intervention programs is usually available from state departments of MR, education, or public health.

For adults, the normalization approach has led to an integration into the community. It is expected that these individuals, regardless of the level of MR, will live in the community with their families, in small group homes, or in

their own apartments. Varying degrees of supports, as needed, are provided, although these resources vary by region. These adults are expected to use community-based, "generic" services, such as health, education, employment, recreation, and mental health. With proper vocational training most can be employed in the open job market or in special settings such as workshops. As with people with other disabilities, they are protected by the Americans with Disabilities Act of 1992, which prohibits discrimination and requires reasonable accommodation for special needs due to the disability.

A somewhat troubling development has been a recent study in California that found that persons with MR living in the community had higher mortality rates than those living in institutions (Kastner et al., 1993). While further research is needed before generalizations can be made, it is clear that mere placement in the community is not a sufficient end in itself.

The provision of adequate health care and other services is necessary. Still, exposure to additional risks of *normal* life may come with the territory of living in a normalized setting.

PART II: MENTAL DISORDERS

COMORBIDITY OF MENTAL ILLNESS AND MENTAL RETARDATION

Although mental illness comorbid with MR was described in the last century, until relatively recently most mental health professionals felt that diagnosing mental disorders in the presence of significant retardation was not possible. Two developments paved the way to diagnosing mental disorders in persons with limited cognitive and language skills. One was the introduction of *DSM-III* in 1978, with behaviorally and phenomenologically based diagnostic criteria. These criteria relied less heavily on subjective psychological symptoms and conflicts which required exploration through the verbal interchange between clinician and patient. Second, progress in research on the presentation of mental disorders in childhood prompted allowances for developmental influences on symptom expression. Thus, standard diagnostic nomenclature could be applied in both children and adolescents as well as for persons of all ages with developmental delay. At the present it is well accepted that MR and mental illness do coexist. In fact, persons with MR are at increased risk for mental disorders. Mental disorders may be an important, even primary, factor limiting functioning of persons with MR, their quality of life, and their adaptation to community life (Reiss, 1994).

Mental disorders in persons with MR are often underdiagnosed. Several factors might be responsible. Clinicians may narrow their focus to the symptomatic treatment of disruptive behaviors, neglecting to evaluate the global picture. "Diagnostic overshadowing," or the tendency to write off symptoms as merely expressions of MR and not mental disorders, will also lead to underdiagnosis (Reiss et al., 1982b).

Having a formal diagnosis of a mental disorder, rather than a nonspecific description of a "behavior disorder" or "challenging behavior," is important with this population. The comprehensive evaluation leading to diagnosis may indicate a specific treatment. The patient can be seen as ill rather than merely "bad." Accurate diagnosis is important for research and administrative purposes including planning for services. The diagnosis, as all medical diagnoses, should be used to characterize disorders, not persons. The term *dual diagnosis* is frequently used for persons with comorbid mental disorders and MR, but it may be confusing, as it also refers to comorbidity of mental illness and substance abuse. It may be stigmatizing and interfere with obtaining services (Szymanski and Grossman, 1984).

The term *behavior disorder* is frequently used by nonmedical professionals working with persons with MR. It has no generally accepted definition. Commonly, this term is meant to refer to disruptive behaviors which pose problems for the caregiver and which are thought to be deliberate, due to environmental factors, performed to avoid certain tasks, or performed to obtain attention. Persons with MR are often referred specifically to discriminate whether they have a "behavior disorder" or a mental disorder. Some caregivers believe that the former should be treated with behavior modification and the latter with psychopharmacological intervention. However, such a dichotomy is simplistic and inaccurate. In virtually every mental disorder there might be behavioral manifestations that are learned, conditioned by environmental factors, and under voluntary control. In addition, every behavioral presentation that is serious enough to cause significant discomfort or dysfunction can be characterized by a *DSM-IV* designation, even if a relatively nonspecific one or a V code is used. In recent years the concept of *behavioral phenotype* has been defined: "the heightened probability or likelihood that people with a given syndrome will exhibit certain behavioral and developmental sequelae relative to those without the syndrome" (Dykens, 1995, p. 523; Hodapp, 1997). For instance, the behavioral presentation frequently associated with Williams syndrome includes strengths in expressive language and superficial sociability, with a tendency to anxiety, perseveration, and short attention span. Behavioral features of selected syndromes are described in Table 3.

EPIDEMIOLOGY OF MENTAL DISORDERS COMORBID WITH MENTAL RETARDATION

There is a wide discrepancy in the reported prevalence rates of psychopathology in persons with MR, due to variations in methodology, diagnostic definitions, and population sampling strategies. A landmark early study comparing the prevalence of psychopathology in children with and without MR in an unselected population on the Isle of Wight (Rutter et al., 1976) indicated that the prevalence was 5 or more times higher in children with MR than in those without MR. With studies using *DSM-III* and *DSM-III-R* criteria, the general impression is that a high percentage of persons who have MR may have symptoms that warrant the diagnosis of an additional mental disorder (Borthwick-Duffy and Eyman, 1990; Eaton and Menolascino, 1982; Einfeld and Tonge, 1996; Lund, 1985; Reiss, 1990). A review by Bregman (1991) of representative studies showed reports of prevalence ranging from 27% to 71%. Virtually all categories of mental disorders have been reported in this population.

PSYCHIATRIC ASSESSMENT OF MENTAL DISORDERS COMORBID WITH MENTAL RETARDATION

Referral to a Mental Health Clinician

Specific referral for mental health consultation usually occurs because of the presence of manifest behavioral problems, most

often disruptive behaviors. Emotional symptoms such as depression and anxiety, if not associated with behavioral problems, lead less often to referral. Specific causes for referral vary with factors such as age, degree of disability, and caregivers' tolerance and concerns. In very young children, irritability and sleep and eating problems are common referral causes. In school-age children, lack of progress, impulsivity, short attention span, and aggression are common reasons for referral. In adolescents and adults, aggression, social problems, self-injurious behaviors, and symptoms related to depression and other specific mental disorders lead to referral.

Psychiatric Diagnostic Evaluation

The evaluation of persons who have MR should follow the general rules of psychiatric assessment. Some modifications may be necessary, depending on the patient's cognitive level, communication abilities, associated disabilities, and life circumstances. However, in principle, the diagnosis of mental disorders in persons who have mild MR and good verbal skills does not differ much from diagnosing these disorders in persons with average cognitive skills. If the retardation is significant, the diagnosis might be more difficult, and in particular the delineation of disorder subtypes, such as for schizophrenia, might not be possible (Reid, 1993).

That a diagnosis must be viewed in a comprehensive context has been long emphasized in the literature (Szymanski, 1980b). Persons with MR often have associated disabilities and disorders, such as seizures, sensory impairments, and motor impairments, and the syndromes underlying the retardation might be associated with a certain behavioral phenotype. These individuals are usually dependent on multiple service providers and supportive services. All these biological, sociocultural, and psychological factors interact with one another to influence clinical presentation and functional adaptation (Garrard and Richmond, 1965). Therefore, for the psychiatric evaluation to be meaningful, the clinical symptoms must be assessed in the broad context of a person's functioning, considering the person's deficits, strengths, and relevant biopsychosocial factors (AAMR, 1992). For persons with poor or no communication skills, the function of the behavior in question must be considered. A disruptive behavior may be a means of communication, serving as a vehicle for obtaining a caregiver's attention or avoiding an unwanted task.

Thus, the psychiatric diagnostic assessment must be comprehensive and multifocal, rather than a "medication evaluation" focused only on the choice of drug to suppress a disruptive behavior. Because of the complexity of many of these cases and the need to interview multiple informants, the diagnostic evaluation may require significantly more time than the evaluation of persons of normal cognition. The clinician must feel that all important areas were sufficiently explored to provide for a comprehensive understanding of the patient and for a detailed discussion of the findings and recommendations with the patient and caregivers.

History

The history is the cornerstone of the diagnostic process. Persons with MR may have difficulties in reliable self-reporting even if the retardation is mild (Reiss, 1994). They often look for outside approval (Zigler and Burack, 1989) and may readily agree with an interviewer's leading questions. They may try to present themselves in the best possible manner and deny having any difficulties or disabilities. The clinical manifestations, especially maladaptive behaviors, may be situation-specific. For example, disruptive behaviors may occur only in the unstructured home situation but not at school, where greater supports are available. Therefore, several informants/care providers (such as parents and teachers) may have to be interviewed to obtain comprehensive historical data. Schools and workshops often collect helpful standardized behavioral data. Various informants may have different concerns and expectations of the psychiatric consultation, which should be explored as well. Table 5 presents an outline of the desired history (based on, among others, Reiss, 1994; Sovner and Hurley, 1983a,b; Szymanski, 1980b; Szymanski and Wilska, 1996).

Medical disorders may be twice as prevalent among persons with MR compared with other mental health referral populations (King et al., 1994; Ryan and Sunada, 1997). Thus, it is important to assess the pattern of psychiatric symptom change over time in the context of concurrent biopsychosocial factors. For example, regression from a previous level of function might indicate a progressive CNS disorder.

Just as mental health needs are often neglected for the population with MR (Einfeld and Tonge, 1996), so, too, general medical care is often lacking (Beange et al., 1995). Certain MR syndromes are associated with increased risk for physical problems, e.g., Down syndrome with thyroid disorders which may result in psychiatric symptoms, hearing impairment, and cataracts. Such problems are often undiagnosed in routine examinations unless physicians are alerted to look for them.

Behavioral Symptoms Related to Physical Illness

Physical illness can be missed in persons with MR mostly because of their difficulties in reporting symptoms. In one study, up to 75% of persons with MR referred for psychiatric assessment were found to have previously undiagnosed or undertreated medical conditions and 45.5% received nonpsychiatric medication that could produce psychiatric symptoms (Ryan and Sunada, 1997). Many medical illnesses have psychiatric symptoms (Hall et al., 1980; Lipkin, 1989). Hyperthyroidism can present with anxiety, depression, or manic symptoms; Wilson disease may present with mood swings and dementia; systemic lupus erythematosus can include mood symptoms; and hyperactivity can be prominent in lead poisoning. Medical

TABLE 5
Components of a Comprehensive History

Domain	Information Needed
Presenting symptoms	Behavioral description of symptoms in various settings, situations, from different caregivers; concrete examples; their evolution over time; antecedent events; management measures used and results
Assessments	Critical review of past cognitive tests and other tests; focus on profile of cognitive strengths and weaknesses rather than on IQ
Medical review	Review of past assessments; request new if past inadequate or out of date
Personality patterns	Premorbid and current patterns of behavior, personality, emotional style
Adaptive skills	Strengths and impairments in communication, self-care, social/interpersonal, community integration, self-direction, functional academic skills, work, leisure, health and safety
Mental health	Past diagnoses, psychotherapies, use of psychotropic drugs; their beneficial and adverse effects; behavioral and other treatments
Environmental supports and stressors	Past and present living, educational, habilitative, work settings: nature, consistency, ancillary therapies; their appropriateness; family events, school transfers, losses of important caregivers; possibility of physical and sexual abuse
Family/caregiving	Understanding of, and reaction to, the patient's mental retardation; overprotection vs. encouraging growth; behavioral management and consistency; disability's effects on the family dynamics, lifestyle, and siblings; long-term plans; consideration of cultural factors

illnesses may also result in nonspecific behavioral symptoms or exacerbate preexisting ones (Gardner, 1996). An example would include a painful condition in a nonverbal person, which may lead to self-injurious behavior (for example, head banging related to otitis media or migraine) as well as to nonspecific aggression. Not infrequently, while the medical illness leads initially to the behavior in question, secondary gain, such as increased attention, may maintain the problem even if the illness has been treated. Thus, multimodal medical and behavioral assessment and treatment may be necessary (Gardner, 1996). Many nonpsychiatric drugs can result in behavioral or emotional symptoms. Commonly cited examples include β-blockers, which may produce depressive symptoms, and phenobarbital used for treating epilepsy, which may result in impulsive, aggressive behavior. Estrogen and progesterone preparations and corticosteroids may also produce affective symptoms.

Patient Interview

Direct patient interview or observation may have to be more extended and done in a "natural" setting such as school, home, or a community residence if possible. Some patients may be very anxious in the office setting. Specific observation and interviewing techniques of persons with MR have been developed and described (Reiss, 1994; Sovner and Hurley, 1983a,b; Szymanski, 1980b). Both verbal and nonverbal techniques are used, depending on the patient's cognitive level, age, and especially communication skills. If a patient's intelligibility is poor, a care-

giver may be used as an "interpreter." Persons who have MR may see a formal, structured mental status examination as an attempt to show that they are incompetent. Therefore, sometimes it may be better to assess the mental status in the context of a nonstructured conversation and interaction that incorporates the questions normally asked in the formal mental status examination. The verbal examination should be adapted to the patient's actual communication level, not to what the interviewer expects for reported IQ. With the use of clear and concrete communications, structure, reassurance, and support, even persons with limited language will respond. In addition to standard components of the mental status, the patient's communication skills, understanding of disability, limitations, and the reasons for the referral, self-image, and ability to relate should be assessed. Confidentiality and a respectful attitude are very important, even if the language used is necessarily simple and concrete. Questions requiring yes and no answers should be avoided because the interviewer's last statement may frequently be endorsed. One should ensure that questions are understood. For example, to the question, "Do you hear voices?" a patient might answer yes, but upon clarification reveal that he means that he is not deaf.

Nonverbal interview techniques include observations of behavior such as performance on selected tasks, relatedness, expression of affect, impulse control, attention span, activity level, presence of unusual behaviors, and seizures, stereotypies, and stigmata of self-injury. A play interview using dolls to "act

out" experiences that cannot verbally be described might be helpful. However, some adult patients may appropriately feel insulted if they are asked to play as if they were children. Artwork, such as drawings, may be helpful.

Rating Scales

A number of psychopathology instruments have been developed specifically for assessment of persons with MR (Hurley et al., 1998; Reiss, 1994). They are useful for screening purposes to identify persons needing mental health consultation, for measuring severity of symptom change in the course of treatment, as guides for obtaining historical and examination data, and for research. While there might be a correlation with a clinical diagnosis, generally they were not intended for such a purpose. The following scales are among those most commonly used. The Reiss Screen for Maladaptive Behavior (Reiss, 1988a,b) provides scores for 8 psychopathology scales and for 6 maladaptive behaviors, as well as a total score reflecting severity. This total score appears to discriminate between people with and without a psychiatric diagnosis. The Reiss Scales (Reiss and Valenti-Hein, 1994) is a child and adolescent version. The Psychopathology Inventory for Mentally Retarded Adults (Senatore et al., 1985) was developed to reflect *DSM-III* categories, and it has separate informant and self-report scales. The Aberrant Behavior Checklist (Aman et al., 1985) appears to be useful in tracking the effects of psychopharmacological agents on persons who have moderate to severe MR. It has 58 items rating behaviors which yield scores on 5 subscales and is an informant-based survey which thus is particularly useful with nonverbal patients. This scale can be useful to measure a person's treatment progress over time. A community version is also available for outpatient use (Aman and Singh, 1994).

Diagnostic Formulation

Clinical data should be interpreted in the context of the patient's developmental level, communication skills, associated handicaps (e.g., sensory and/or motor), life experiences, education, and family and sociocultural factors (Szymanski, 1980b). Not every unusual behavior equates with psychopathology. A particular behavior may suggest an underlying mental disorder if it is a part of a pattern of a defined mental disorder syndrome (*DSM-IV*; American Psychiatric Association, 1994) and if it is associated with distress or disability, and not just a temporary reaction or adaptation to a given situation (such as lack of structure and attention in an understaffed facility). To be clinically useful, the diagnostic statement should not be limited to coding the formal name of a disorder, but should include a description of the person's strengths, deficiencies, and needs including intellectual, adaptive behavior, communication, health, and psychosocial domains (AAMR, 1992; Szymanski and Crocker, 1989). With this population it is also particularly useful to summarize the criteria on which diagnoses are derived in indi-

vidual cases. The diagnosis of a mood disorder will carry more credence for outside agencies or care providers if specific mention is made of neurovegetative symptoms, which will also assist others in monitoring the course of treatment. The practice of categorizing MR as a primary disorder and the comorbid mental disorder as secondary (or vice versa) is neither useful nor based on scientific grounds (Reiss, 1994; Reiss et al., 1982a).

Formal *DSM-IV* Diagnosis

Whenever appropriate, an Axis I *DSM-IV* diagnosis should be made. Factors limiting the reliability of such a diagnosis include experience and training of clinicians, a patient's limited communication skills, lack of reliable history, and lack of sufficient time for the diagnostic assessment. While most of the *DSM-IV* criteria consider a patient's verbal productions, there are also provisions for situations in which the patient does not have sufficient language to describe symptoms. For example, in major depression, depressive mood may be indicated by "observation by others." For individuals with more severe MR, especially if they are nonverbal, the less specific diagnoses, such as those with NOS designations, may have to be used.

DIAGNOSIS OF MAJOR MENTAL DISORDERS COMORBID WITH MENTAL RETARDATION

Pervasive Developmental Disorders

About 75% of autistic children also meet criteria for MR, and both can be associated with the same disorders, such as congenital rubella syndrome, tuberous sclerosis complex, and PKU (Fombonne, 1997; Tanguay, 1980). As opposed to uncomplicated MR, children with a PDD have significant impairments in reciprocal social interaction and social communication, verbal or nonverbal. Similarly, in individuals with significant MR who have poor verbal language and who exhibit self-stimulatory behaviors, the diagnosis of a PDD should not be made unless they also have significant impairments in their social skills, considering their developmental level. The diagnostic criteria for these disorders were modified in the *DSM-IV* to minimize false-positives in these circumstances (Volkmar et al., 1994). The diagnosis of autistic disorder in adults may be quite difficult and requires a detailed early developmental history.

Attention-Deficit/Hyperactivity Disorder

Poor attention and hyperactivity are common reasons for psychiatric referral of persons with MR. The prevalence of ADHD in this population is similar to that in the general population, estimated at between 4% and 11% (Feinstein and Reiss, 1996). The formal diagnosis of ADHD should be made only if the *DSM-IV* diagnostic criteria are fully met. Most of these criteria are based on observable behavior and thus may be applied for nonverbal children. The criteria specify that the symptoms be considered in light of the patient's developmental level, but

use of chronological age instead has been suggested (Pearson and Aman, 1994). In the differential diagnosis, one should consider inattentiveness that is situation-specific (such as at school if the academic expectations are too high); hyperactivity associated with certain syndromes, such as fragile X (Baumgardner et al., 1995, Feinstein and Reiss, 1996); and side effects of certain medications (such as phenobarbital).

Conduct Disorders

The prevalence of conduct disorder in children and adolescents with mild MR has been reported at 33% (Richardson et al., 1985). Persons with MR are frequently referred because of "noncompliance," or not following commands of caregivers. In such cases diagnoses of conduct and oppositional defiant disorders should be considered in the context of an individual's circumstances, ability to understand social rules, and possession of sufficient skills to communicate opposition. Contrary to common belief, conduct disorders in this population are not correlated with CNS impairment (for review, see Feinstein and Reiss, 1996).

Tic Disorders

Tourette's disorder can be comorbid with MR. Differentiation of tics from self-stimulatory stereotypic movements common in persons with severe MR may be difficult. The latter are usually more complex than tics and appear intentional, although proving intentionality may be difficult in nonverbal persons.

Stereotypic Movement Disorder

This category can be used for self-stimulatory, nonfunctional, motor behaviors, usually comorbid with severe MR, if they are a focus of treatment and if other relevant mental disorders are excluded. The *DSM-IV* permits a specifier "with self-injurious behavior," if bodily damage results. While not a specific symptom of any diagnosis or underlying disorder, a tendency to self-injury is common in certain MR syndromes, especially Lesch-Nyhan, Prader-Willi, and to lesser degree Cornelia de Lange and fragile X syndromes.

Mental Disorders Due to a General Medical Condition

This term replaced in the *DSM-IV* the categories of "organic" mental disorders, which were often inappropriately used as a "wastebasket" or default diagnosis for any inappropriate behaviors of persons with MR. The rationale was that MR is due to a "brain injury" from which all associated unusual behaviors derive. The *DSM-IV* criteria require that "there is evidence from history, physical examination, or laboratory findings that the disturbance is caused by the direct physiological consequences of the general medical condition" (coded on Axis III). Thus, mere coexistence of MR and abnormal behaviors is not sufficient for this diagnosis, but it might be justified by temporal relationship, such as the emergence of seizures together with personality change.

Certain conditions underlying MR are associated with increased incidence of medical conditions that result in behavioral/emotional symptoms. For example, Down syndrome is associated with increased incidence of hypothyroidism that may present with symptoms of depression (Pueschel, 1982). It is also associated with Alzheimer-type dementia, although the clinical symptoms rarely are apparent prior to 45 to 50 years of age, and as many as 75% may be clinically affected by age 60 years (Hodapp, 1996; Lai and Williams, 1989).

Schizophrenia and Other Psychotic Disorders

Earlier views that stereotypic behaviors seen in persons with severe retardation were diagnostic of schizophrenia (Hayman, 1939; Kraepelin, 1896) are no longer accepted. The prevalence of schizophrenia is estimated to be similar in persons with and without MR (Reiss, 1994). Most of the *DSM-IV* criteria for schizophrenia require some degree of language skills to document existence of delusions, hallucinations, or grossly disorganized speech. Therefore, it is generally accepted that while schizophrenia can be diagnosed in the usual manner in persons with mild MR, this diagnosis and subtype cannot be made in the presence of more severe retardation (Reid, 1993). In those cases the less specific diagnosis of psychotic disorder NOS may be made, if features such as grossly disorganized behavior and negative signs are present but were absent in the premorbid period. One must be careful not to confuse conversation with an imaginary friend and hallucination (Szymanski, 1980b).

Mood Disorders

Mood disorders, especially depressive disorders, are probably underdiagnosed in persons with MR, as referents tend to focus on persons who are disruptive. Earlier studies probably also underestimated the prevalence of affective disorders in persons with MR (Feinstein and Reiss, 1996), but it is now generally accepted that these individuals suffer these disorders with equal or greater frequency than the general population (Reid and Naylor, 1976; Reiss, 1994; Reiss and Benson, 1985; Sovner and Hurley, 1983c; Sovner and Pary, 1993). Depressed mood has been described as more frequent in children with MR than in children without retardation (Schloss et al., 1988). In persons with mild MR the symptoms are similar to those among patients with normal intelligence, although the complaints may be simpler and more concrete, for example, feeling sick rather than sad. The *DSM-IV* criteria make the diagnosis of mood disorders easier in persons with limited verbal skills by allowing observer ratings of mood change in place of self-report. Depression in these individuals may also manifest in aggressive behavior (Reiss and Rojahn, 1993). Suicidality has also been reported (Walters et al., 1995). The behavioral symptoms have to be assessed in the context of change over time (compared with the premorbid status) and of a person's realities. For example, pressured speech in a manic episode may be replaced with increased

vocalization if the person is nonverbal (Szymanski et al., 1998). Environmental events frequently trigger a major depressive episode. These may include a precipitous move to new setting and, for adults, the realization that one is "overtaken" by younger sibling. Rapid-cycling bipolar disorder has been described in children and adults with significant MR with mood, behavior, and sleep shifts as frequent as every few days (Jan et al., 1994; Glue, 1989; Lowry and Sovner, 1992).

Anxiety Disorders

A broad range of anxiety disorders, including generalized anxiety disorder, phobias, panic disorder, PTSD, and OCD, has been reported in persons with MR, with a prevalence of about 25% in an outpatient sample (Benson, 1985; review by Feinstein and Reiss, 1996). Feinstein and Reiss (1996) highlighted the importance of psychosocial stress factors, including fragile self-esteem, fears of failing, and loss of caregivers, as likely contributing to the development or expression of anxiety disorders in this population. Patients may talk about disliking certain situations, rather than report that they are anxiety-provoking. The tendency to anxiety and social avoidance is also a part of the behavioral phenotype of fragile X syndrome (Baumgardner et al., 1995). The diagnosis of disorders in this category is easier in verbal persons who can report on subjective feelings associated with anxiety episodes. In persons with little or no verbal ability, the diagnosis may be only postulated on the basis of observed behaviors, which may include avoidance behaviors, such as school or work refusal and signs of autonomic arousal.

Posttraumatic Stress Disorder

PTSD in persons with MR has been described (Ryan, 1994). Like children, this population is vulnerable to abuse because of difficulties in reporting it, dependency, a tendency to want to please others, and lack of understanding of their rights. For these reasons PTSD is probably significantly underdiagnosed in this population, and it should be routinely considered in differential diagnosis. A common defense on behalf of the perpetrator is that the alleged victim is too retarded to be a reliable witness.

Obsessive-Compulsive Disorder

Differentiating between self-stimulatory, stereotypic behaviors and compulsions may be difficult in nonverbal persons who cannot describe obsessional thoughts or identify compulsive behaviors as responses to obsessions. Some repetitive behaviors, for example, hoarding objects, flicking lights on and off, and tidying and arranging, all have been suggested as indications of OCD in persons with MR. Some forms of self-injurious behavior are accompanied by self-restraint, for example, tightly binding the arms in clothing or insisting on wearing protective devices such as helmets or gloves. In these cases, where self-restraining behavior might suggest the ego-dystonic nature of

self-injury, a connection between self-injurious behavior and OCD has been postulated (King, 1993).

Eating Disorders

Eating and feeding disorders occur in persons with MR. While anorexia and bulimia nervosa are relatively rare in the context of MR, particularly MR of moderate to severe severity, MR is a predisposing factor for other eating disorders such as pica and rumination. The ingestion of nonnutritive substances, pica, and the regurgitation and rechewing of food, rumination, occur with greater frequency as the severity of cognitive disability increases. When these behaviors are a focus of clinical attention, the diagnoses should be indicated.

Personality Disorders

Although there is no personality pattern unique to MR, traits such as conduct problems, distrust and suspiciousness, attention-seeking, and dependency are common in this population. Often these are related to environmental factors (necessary dependence on others, lack of social experiences) or neurological factors (such as personality traits related to temporal lobe epilepsy), and these factors have to be considered when making a diagnosis. Personality disorders have been found to occur in a high percentage of persons with MR (Reid and Ballinger, 1987; Reiss, 1992). It has been suggested that this diagnosis should not be made in persons functioning below the mild MR level (Dana, 1993).

TREATMENT

Treatment should flow from a comprehensive, biopsychosocial diagnostic assessment. The basic principles of psychiatric treatment for a given mental disorder are essentially the same, whether the patient has MR or normal intelligence. However, modification of treatment approaches and specific techniques are necessary, according to the individual patient's needs, life circumstances, and cognitive and communication skills. The goal of treatment is not merely removal of symptoms, but helping the patient to achieve maximally feasible quality of life (Stark and Goldsbury, 1990).

The Behavioral Emergency

When the psychiatrist is called to assist with a behavioral emergency, the principles of treatment flowing from a complete evaluation still apply. However, it is usually necessary to ensure a patient's and others' safety first. In the case of severe self-injurious or aggressive behavior, if the usual attempts at redirection fail and the patient continues to pose an imminent risk, it may be necessary to temporarily use physical restraint. In some instances this may require admission to a psychiatric hospital. Medical causes for an acute behavioral exacerbation must always be considered. It is not uncommon for constipation or infection to contribute to irritability, which sets the stage for behavioral problems. Moreover, medication side effects such as

akathisia from neuroleptics or disinhibition from sedative-hypnotic agents can be expressed in aggressive and self-injurious behaviors. In situations in which a temporizing measure is necessary, it is generally advisable to use a drug with which the patient has positive experience, typically a neuroleptic or benzodiazepine, pending the completion of a more definitive diagnostic and treatment plan. Emergencies are generally not the occasions to experiment with new treatments. Furthermore, the use of such emergency medication should be considered only after adequate attention can be given to a diagnostic assessment. The need for emergency treatment should prompt a comprehensive diagnostic assessment including the evaluation of environmental influences. Approaches should also be considered to prevent recurrence or refine the approach to such emergency situations should they occur again.

The "Medication Evaluation"

In some systems that serve persons with developmental disabilities, there is a strong emphasis on compliance with staff rules and skills acquisition. This approach can represent a type of "affectionless control" which worsens conditions such as depression and anxiety disorders. Individuals in these circumstances may be referred to a psychiatrist with a request for a "medication evaluation," which usually means obtaining a medication that will suppress behavior deemed disturbing by the referring caregiver, sometimes even at the expense of benefit from the educational or habilitation program. Psychiatrists who approach a "medication evaluation" as a "patient evaluation" can seize an opportunity to educate staff about a person's overall biopsychosocial well-being and avoid inappropriate medication administration or expectations.

Psychosocial Treatments

Typical goals of psychotherapy may include symptom relief through altering patterns of maladaptive behavior and helping the patient to understand his or her own disability and associated feelings, balanced by a recognition of strengths, to constructively resolve internalized conflicts and develop realistic expectations for self. These expectations might include striving for independence; learning to recognize, manage, and communicate emotions; learning to recognize the effect of one's own behavior on others; developing age-appropriate social skills; and learning to constructively handle developmental crises and challenges. Thus, the focus is usually on current reality rather than on uncovering the past. However, with persons with mild MR and good language, a more exploratory and dynamic approach aiming to integrate past experiences may be possible. The therapy can be viewed as an educational process that uses communication means and techniques appropriate for the patient's language, cognitive, and interactional skills. Developing a positive self-image is essential in order to avoid self-destructive behaviors which are used in a futile attempt to prove self-worth

and which make these patients vulnerable to exploitation. "Pressure-proofing," or teaching how to resist peer pressure to protect oneself from engaging in inappropriate or dangerous behavior, can be a very useful part of therapy. Most verbal patients can understand their limitations, if presented as a lack of "talent" in certain areas, counterbalanced by a discussion of their unique gifts or strengths.

Studies on psychotherapy techniques and outcomes with persons with MR have long been available (anthology by Stacey and DeMartino, 1957; Thorne, 1948). Although most of the outcome literature is anecdotal, there is consensus that these individuals can benefit from individual, family, and group psychotherapy, providing that they have the minimal necessary communication skills and psychotherapeutic approaches are modified according to their needs (Harris, 1995; Hurley, 1989; Reiss, 1994; Szymanski, 1980a; Szymanski and Rosefsky, 1980). The principles of psychotherapeutic management of persons with comorbid mental disorders and MR, as elucidated in the literature, are summarized below in terms of prerequisites, context, and techniques.

Prerequisites. The patient should be sufficiently communicative to permit a meaningful interchange with the therapist, verbally or nonverbally. An accurate and comprehensive diagnostic assessment should be completed. The therapist should be trained in developmental disabilities, familiar with the medical problems of the patient, and willing to work with persons with cognitive disability.

Context. The therapy should be an integral part of a comprehensive treatment program. The mental health clinician should actively participate with other professionals in the development of the treatment across settings. For example, on the basis of knowledge of the patient's poor self-esteem, the clinician might explore a change to a classroom or job more appropriate to the patient's abilities that will ensure success and reduce frustration and aggressive outbursts.

Techniques. These techniques have to be adapted to a patient's chronological age as well as communication and cognitive skills. The communications should be clear and concrete. The use of familiar examples may be helpful, and the therapist should ascertain the patient's understanding. Other techniques include directiveness and structure to maintain focus, but not at the expense of spontaneity. Long silences may be perceived as frustrating and punitive. Play and activity may be useful, if adapted to chronological age. For example, an adolescent may engage in creating a "TV show" in a "model house," rather than in doll play. The patient should be guided to discover interpretations, rather than be told statements that may be rote-memorized as "pat" phrases such as, "You're angry because you have lost control."

The therapist should see and respect the patient as a person with dignity, be flexible, be able to use himself or herself as an example, give concrete advice, avoid overprotection, resist rescue fantasies, and be able to work with other caregivers.

Group Therapy. In addition to individual therapies, group therapy is particularly useful with adolescents and young adults who have relatively good verbal skills. Social skills training can also be helpful in this context (Hollins et al., 1994; Szymanski and Rosefsky, 1980). Multiple family group therapy provides the family and the child with a supportive microcosm of the society at large, represented by other families in the group (Szymanski and Kiernan, 1983).

Family Therapy. The family should be seen as treatment team members. The family is needed to provide information the patient cannot provide and to generalize the therapy across environments. Goals have to be consistent with those for the patient and consonant with the family's values. These goals usually include recognizing the patient's strengths, avoiding guilt feelings and overprotection, supporting a child's pathways to independence, and providing opportunities for success. Learning about the nature of the child's disability, help in finding resources including information about entitlement for services, advocacy, empathy but not paternalistic sympathy, and concrete advice in management are important.

Over time, the needs of families change, depending on a child's age and condition. Parents of young children, recently diagnosed, need detailed explanations of their child's condition, coupled with a second opinion if necessary, and help in understanding that the child is not hopeless and will always continue to learn. Parents may react with grief, anger, and despair with the initial diagnosis. They need guidance in finding services, such as early intervention and various therapies. If the psychiatrist cannot be helpful in this respect, a referral should be made to an appropriate professional or an agency. Parent organizations, such as The ARC (formerly Association for Retarded Citizens), provide invaluable support, information, and advocacy. Parents of older children, especially those with challenging behaviors, need guidance in managing the child in order to balance teaching with building positive self-image through encouraging development of existing abilities, rather than focusing only on the impairments. Concrete and ongoing help in developing behavior management programs and generalizing to the home the programs instituted at school is essential. Families also need help in obtaining educational supports to which the child is entitled under federal and local laws. Parents of adolescents need help in coming to terms with a child's sexuality and with helping the child to develop as much independence as possible.

Parents of adults need help in emotionally separating from their children, in permitting them to move to out-of-family living arrangements, and in finding appropriate placement in the community. Some parents see the latter as abandoning their children, and they may need help resolving feelings of guilt. An emerging group are elderly parents who themselves become feeble and in need of care, with whom adult or elderly children with MR still live. This group may require considerable external support services.

Collaboration With Other Professionals. Teachers and other service providers have to be actively included as team members with families in the therapy process. They should be involved in developing therapy goals, and their reports are necessary in monitoring its progress. They are also critical in developing opportunities for patients to practice newly acquired skills, such as activities of daily life, that will promote positive self-image. Close collaboration and consultation with family physicians, pediatricians, speech and other professionals, workshop counselors, and others are important.

Behavior Modification. These techniques, if properly applied, provide a consistent and structured framework for teaching appropriate behavioral patterns as well as various adaptive life skills. Details of specific behavioral treatment techniques are beyond the scope of these parameters and are nicely reviewed elsewhere (Reiss, 1994; Schroeder, 1989). Behavioral treatment based on restraint and aversive procedures is currently only, if ever, used for brief periods in rare cases of severe problems. Such circumstances might be intractable aggression or self-injurious behaviors which could result in irreparable damage such as blindness from head hitting and which have not responded to other measures. Generally, behavior therapy for aggression or self-injury flows from an analysis of the functions which such maladaptive behavior might serve and the variables which reinforce that behavior. An individual whose aggression is motivated by anxiety and reinforced by the clearing of a room, for example, would very likely be taught other ways of indicating fear as part of his behavior plan. He might also be given opportunities to take himself to a quiet setting in lieu of creating one in consequence of his aggression. To be effective, a behavior management program should be generalized and consistent in all settings, such as home and school. It should focus, first of all, on teaching appropriate skills and behaviors to replace maladaptive behaviors rather than merely to suppress them.

Milieu

Environment influences behavior. It is important that the provision of work, educational opportunities, and living setting be commensurate with the patient's needs and abilities. For example, classroom placement with sufficient educational supports and an opportunity to succeed may obviate the need for medication for a child presenting with disruptive behavior and expressed hopelessness.

Pharmacotherapy

There is no evidence that MR per se changes the mechanism of action of psychotropic drugs. Medication effects in this population generally are not different from those expected in the absence of MR, and while a drug's effects might be influenced by a medical/neurological disorder associated with MR, few such situations have been described. The adage "start low, go slow" reflects the experience that shifts in dose-response in

certain contexts are far more likely than changes in the mechanism of action of a compound. Thus, persons with Down syndrome may be exquisitely sensitive to anticholinergic drugs and some persons with MR may be more sensitive to the disinhibiting effects of sedative-hypnotic agents than the population in general. Table 6 summarizes some of the behavioral and other side effects that have been associated with medications in persons with MR. It is not always clear that the risk of these side effects is specifically increased in association with MR, however, as many side effects come from case reports and not population surveys with requisite controls. The choice of a drug in persons with MR should follow the usual principles of rational psychopharmacology and proceed from an accurate diagnosis. Nonspecific indications such as prescribing a drug for "aggression" should be avoided if possible, because this symptom may be only one aspect of a variety of disorders including psychosis, mood disorder, or disruptive disorders. As needed or p.r.n. prescriptions should be given with caution, as medications so administered bring considerable potential for abuse.

As in any medical treatment, the risk/benefit ratio should be considered. The benefits of the drug should be weighed versus potential adverse effects. Medications should be prescribed only for the treatment of specific mental disorders and as a part of a comprehensive treatment plan. They should not be used for the convenience of the caregivers or as a substitute for appropriate services. The clinician should be alert to the possibility of service systems seeking a medication to cause a person to become more compliant with a living or work setting which may be inappropriate for the individual and thus anxiety-provoking. Overuse of medications and the use of more than one drug, unless there is a specific indication and proof of effectiveness, should be avoided. The lowest effective dose should be used. Opportunities for simplification and consolidation of medication regimens in the context of high medical comorbidity should be considered. For example, if a patient has a seizure disorder, perhaps an anticonvulsant drug may be changed to one that also has a desired psychotherapeutic effect, and thus the use of 2 medications is avoided.

Careful follow-up for side effects, particularly tardive dyskinesia, akathisia, and extrapyramidal symptoms, is most important because patients with significant MR may be unable to self-report symptoms. Scales measuring involuntary movements such as the Abnormal Involuntary Movement Scale are helpful. Many patients with significant MR engage in stereotypic movements that may be difficult to differentiate from involuntary movements of tardive dyskinesia. Pretreatment videotap-

TABLE 6
Medication Side Effects With Possible Increased Frequency in Persons With Mental Retardation

Drug	Side Effect(s) Reported in Patients With MR
Aminophylline (ethylenediamine)	Aggression
Anticholinergic drugs	Greater likelihood of cognitive impairment, delirium in persons with Down syndrome
Carbamazepine	Inconspicuous elevation of carbamazepine-epoxide levels during polytherapy with seizure exacerbation; hyponatremia; hypovitaminosis D; folic acid and riboflavin deficiency in persons with marginal diet; irritability
Clobazam	Aggression, agitation, SIB, insomnia, hyperactivity
Gabapentin	Aggression; choreoathetosis reported in persons with significant brain abnormality
Lithium	Cognitive dulling, increased likelihood of toxicity due to erratic fluid intake or regulation of same
Lorazepam, other benzodiazepines	Hyperactivity, SIB, withdrawal-induced manic symptoms
Methylphenidate	Social withdrawal, motor tics
Neuroleptic drugs	Greater likelihood for development of tardive and other dyskinesia, parkinsonism, withdrawal irritability, self-injury, akathisia
Phenobarbital	Irritability, SIB, aggression, hyperactivity, propensity to osteomalacia
Phenytoin	Increased susceptibility to intoxication; cerebellar, brainstem atrophy; osteomalacia
Valproate	Pancreatitis, hepatotoxicity, myelodysplasia

Note: MR = mental retardation; SIB = self-injurious behavior.

TABLE 7
HCFA Guidelines for Psychotropic Medication Use in Persons With Mental Retardation

Prior to prescribing psychotropic medication:
- Medical, environmental, and other causes of the behavioral problem must be ruled out
- A detailed description of symptoms and differential diagnosis is required
- Behavioral data should be collected
- The least intrusive and most positive interventions should be used, including as applicable behavior therapy, psychotherapy, and habilitation/education; medications might be the least intrusive and most positive intervention in some cases

When medication is prescribed:
- It should be an integral part of an overall individual active treatment program
- It should not diminish the patient's functional status
- The lowest effective dose should be used
- A gradual dose reduction should be periodically considered (at least annually) unless clinically contraindicated
- Adverse drug effects should be monitored
- Data should be collected documenting that the drug achieves the desired outcome (including patient's quality of life)

Note: HCFA = Health Care Financing Administration.

ing of the patient may be helpful in these cases. Drugs should be used only as long as there is clear evidence that they have benefits that outweigh their side effects. Periodic trials of dose reduction and discontinuation can be helpful to assess ongoing need. Dose reduction and drug discontinuation should be gradual, particularly if there may be withdrawal effects from the drug in question. There is no established standard of practice for optimal rate of drug taper toward discontinuation. In general, if a drug confers no identifiable benefit or the risks of a drug outweigh its benefit, a more rapid taper is indicated. If a taper is part of an effort to identify a minimum effective dose in the context of previously demonstrated effectiveness or the patient has a history of withdrawal-emergent symptoms, a more gradual reduction strategy will apply. Reiss and Aman (1998) offer an excellent review of clinical and social aspects of psychopharmacology of developmental disabilities.

Regulations applicable to Intermediate Care Facilities for the Mentally Retarded (Title 42 of the Code of Federal Regulations, Part 483, Subpart I, Sections 400 through 480) were originally written more than 10 years ago, but in 1997 the Health Care Financing Administration (HCFA) published *Psychopharmacological Medications: Safety Precautions for Persons With Developmental Disabilities* (HCFA, 1997). This is a manual for surveyors who assess a facility's compliance with the regulaions. This manual interprets the original regulations in light of modern approaches to MR. The Committee on Mental Retardation and Developmental Disabilities of the American Psychiatric Association had considerable input into this manual, which can be obtained from HSQB, Center for Long Term Care, HCFA, s2-20-03, 7500 Security Boulevard, Baltimore, MD 21244-1850. The main points concerning psychotropic medication use are summarized in Table 7. The manual includes the standard mention that the use of drugs should be "approved" by an interdisciplinary team. This point may be criticized as tantamount to suggesting the practice of medicine by nonphysicians. The manual does not condone such a practice, and it

refers rather to coordination with the interdisciplinary team of which the physician is an important member. In the end, however, the physician, not "the team," must assume the final authority concerning medical decisions (Davis et al., 1998).

Pharmacological Treatment of Specific Disorders

Depression. Selective serotonin reuptake inhibitors have gained favor with this population because of their relatively benign side effects profile (Sovner et al., 1998). Tricyclics are used less often, and their potential undesirable effects on seizure threshold, cardiac rhythm, and cognition are particularly salient in persons with MR and comorbid medical conditions. Monoamine oxidase inhibitors are rarely used because of the difficulty in ensuring that the patient and the caregivers will be able to comply with dietary restrictions (Sovner et al., 1998). Electroconvulsive therapy is quite controversial and rarely used in this population, possibly because of difficulties with consent and a perceived potential for abuse.

Anxiety Disorders. Persons with MR are particularly sensitive to the cognitive and other effects of benzodiazepines (impaired acquisition of memory, interference with respiration, and disinhibition), and thus these are not usually used as first-line treatments for chronic anxiety. Barron and Sandman (1983) observed that fully two thirds of persons with stereotypies and self-injury drawn from a developmental center population were described as having paradoxical reactions to sedative-hypnotic agents used as pretreatment for dental or other procedures. Though with minimal, if any, literature support, medications such as buspirone (Werry, 1998), antidepressants with anxiolytic effects, such as fluoxetine or paroxetine (Cook et al., 1992; Davanzo et al., 1998), or antiepileptic drugs, such as carbamazepine, are often used for anxiety disorders. Clonidine and various β-blockers may be useful for short-term treatment of anxiety (Fraser et al., 1998). However, over the long-term, side effects of depression, sleep disturbance, cardiac disturbances, and cognitive dulling may make these medications

intolerable. The treatment of OCD is similar to that for persons without MR. However, selective serotonin reuptake inhibitors are the initial drugs of choice over clomipramine because of their more favorable side effect profiles (Sovner et al., 1998).

Bipolar Disorder. Lithium, while effective (Poindexter et al., 1998), may not be well tolerated because of the magnified significance of cognitive dulling. First-line treatments usually include antiepileptic drugs such as carbamazepine and valproic acid (Poindexter et al., 1998). Newer anticonvulsants such as gabapentin (Ryback and Ryback, 1995) and lamotrigine (Uvebrant and Bauziene, 1994) may show some promise in persons with MR, but more investigation is needed.

In many settings that serve persons with MR, there are serious difficulties with sleep hygiene which may destabilize the patient with bipolar disorder. This may include frequently waking the person or roommate for toileting, lack of a comfortable sleep setting, frequent awakenings due to noise caused by staff or colleague activities, or even nighttime employment. Education of the team regarding issues of sleep hygiene is important.

Schizophrenia. Persons with MR are more susceptible than others to tardive dyskinesia, tardive akathisia, and other toxic effects of conventional antipsychotic agents (Gualtieri et al., 1984, 1986). For this reason the newer, atypical neuroleptics are gaining favor as first-line treatments. Antipsychotic medications remain appropriate as first-line treatment of psychosis generally and for schizophrenia in particular (Baumeister et al., 1998).

Disorders of Impulse Control. Perhaps the most common clinical justification for the use of psychotropic medication in persons with MR is disruptive behavior. Three of the most commonly identified types—self-injurious behaviors, stereotyped behaviors, and aggression—have been the subject of considerable attention. Although these behaviors are often grouped as outcome measures for treatment studies, their pathogenesis is likely to be quite different and there is no uniform response to pharmacological agents.

Neuroleptics have been the most widely used agents in the treatment of destructive behaviors (Baumeister et al., 1998). With respect to both self-injurious behavior and aggression, the majority of reports have concluded that such behaviors may be effectively suppressed by these agents, but the number of well-controlled studies is limited. It is generally believed that symptom suppression may be nonspecific in that improvement has been reported for multiple target behaviors simultaneously (Baumeister et al., 1993; Osman and Loschen, 1992). Studies of stereotyped behaviors, on the other hand, have produced more consistent and reliable results. Neuroleptics have been shown to be clearly and specifically effective in decreasing these behaviors (Aman et al., 1989). Because of the significant risks for side effects associated with neuroleptic agents, alternatives are continually being explored. Opiate receptor antagonists such as naltrexone, for example, have received considerable attention and may be particularly useful for particular types of

self-injurious behaviors such as self-biting (reviewed by Sandman et al., 1998). However, results have not been uniformly encouraging. Self-injurious behavior may be initially worsened by opiate blockers (Benjamin et al., 1995), and a recent, relatively large-scale double-blind study found no positive clinical effects in more than 30 adults with MR and self-injurious behaviors. Moreover, naltrexone appeared to exacerbate stereotypic behaviors in some of the patients studied (Willemsen-Swinkels et al., 1995).

Interest in the role of serotonergic agents in moderating aggression, stereotypies, self-injurious behaviors, and compulsive behaviors has been increasing. Several studies have demonstrated the efficacy of clomipramine in the treatment of repetitive behaviors in children with autism and persons with MR (Gordon et al., 1993; Lewis et al., 1996). Additional medications occasionally used in the treatment of aggression include lithium and β-blockers (Fraser et al., 1998). Several studies have shown high rates of response to lithium (Bregman, 1991). Because of the relatively narrow toxicity window for lithium and because hydration status can be erratic in this population, some clinicians are reluctant to use this agent as readily as may be the case for persons without MR.

Sleep Disorders. Persons with MR, particularly those with multiple impairments such as congenital rubella syndrome that include blindness, may present with sleep disturbance. Sleep disorders have also been described in Smith-Magenis syndrome (insomnia) and in Prader-Willi syndrome (excessive daytime somnolence). There are no approaches to the treatment of sleep disturbance that are unique to persons with MR. The use of benzodiazepine hypnotics should be approached cautiously because of the potential for disinhibition.

Nonestablished Dietary Treatments

Numerous dietary treatment approaches to MR have been proposed over the years, but their effectiveness generally is not supported by methodologically sound research, if any (Singh et al., 1998). These treatments include vitamin and mineral supplements and various dietary restrictions, for example yeast and gluten-free regimens. Folic acid, initially suggested as leading to marked improvement in children with fragile X syndrome, has not been found to be an effective treatment in well-designed studies. Similarly, there is no clear evidence from methodologically sound studies for the effectiveness of vitamin B_6 in this or any MR syndrome. In certain metabolic disorders, such as PKU, appropriate diet is clearly necessary and may even prevent the development of MR. Reports of low-phenylalanine diets providing help in the treatment of behavioral symptoms in adults with MR due to untreated PKU also exist. Children with developmental disabilities often have peculiar eating preferences. In such cases a nutritional assessment is advisable to rule out malnutrition, including a lack of certain essential vitamins and nutrients.

Choosing and Implementing a Treatment Plan

Treatment priorities should be established with more general interventions such as improving services initiated first. More complex and intrusive measures may be added later, as needed. Some interventions may have synergistic effects, for instance, behavioral techniques may be more effective when a child's hyperactivity and short attention span are improved with the use of an appropriate medication. Persons with relatively mild retardation but with severe motor and communication impairments that severely limit their interaction with others, such as nonverbal, nonambulatory individuals with severe cerebral palsy, can become severely depressed, although their depression may go unrecognized. As much as possible, the relevant impairment should be addressed as part of a comprehensive treatment program. For example, provision of alternative communication for a depressed or disruptive, nonverbal person, such as signing, may be an effective antidepressant intervention.

Continuation and Maintenance Treatment

A common problem in the treatment of persons with MR is assessing its effectiveness. Various caregivers may have different opinions which may reflect their subjective expectations of the results as well as the patient's actual progress (Szymanski and Wilska, 1996). For example, an adolescent recovering from a depressive episode may become more active and demanding of attention, which might be viewed by a teacher as deterioration. To prevent such misperceptions, discrete treatment goals should be established collaboratively by the clinician and other caregivers for each identified problem area. A problem-oriented, follow-up mechanism should be developed which includes selecting target or "index" symptoms and means to assess their change over the course of treatment. The collection of well-defined behavioral data can form a basis for a decision regarding whether a treatment in question is working and should be continued or not.

The patient should be seen for follow-up as needed. Patients receiving stable psychotropic medication regimens should be seen at least every 3 months, and annual reviews can be helpful for persons with previous drug exposure to establish familiarity and a baseline level of function. In many cases, the generation of the annual Individual Service Plan provides an opportunity for yearly psychiatric input. During these and other visits, sufficient information should be obtained to assess not only the changes in the problem areas and target symptoms, but also emergent side effects, changes in clinical presentation, patient progress in all areas, and relevant environmental events. Clinicians of other disciplines, such as behavioral psychology, are expert in collecting concrete behavioral data, and collaboration with them can be very valuable. These data and the rationale behind any changes in therapy that are recommended should be clearly recorded and available to other involved professionals.

It is reasonable to expect that the person receiving treatment will experience reduction in symptoms of concern and improved quality of life as assessed by physical health, living situation, and vocational and social functioning. If the person is not experiencing improvement, the accuracy and completeness of the biopsychosocial diagnosis should be reviewed, as well as the consistency of implementation of treatment by the caregivers.

Factors Influencing Treatment Choice and Results

Cognitive Skills. The severity of MR will influence, to some degree, the choice of treatment. Severe MR and lack of communication skills will likely make verbally based therapies impractical, although these individuals may benefit from behavior modification and milieu manipulation.

Concurrent General Medical Disorders. The more severe the retardation, the higher the prevalence of associated medical, neurological, and sensory impairments which will complicate psychiatric treatment. Many of these individuals receive a variety of medications for their medical condition, which may have behavioral side effects and may cause adverse interactions with psychotropic drugs.

Collaboration of Professionals and Caregivers. Persons who have MR, because of their multiple needs, usually receive services from a number of caregivers and professionals of different disciplines. Close collaboration within this team of professionals is essential to the success of the treatment program. Treatments prescribed by various professionals should be coordinated and integrated, and all should be aware of their colleagues' roles. This interdisciplinary team approach, although somewhat time-consuming, is in fact required by various consent decrees concerning institutional care and by HCFA standards. In a truly interdisciplinary team, all disciplines are expected to contribute to all aspects of the program, although each has primary responsibility in its own area of expertise (Szymanski and Wilska, 1996). Effective treatment requires the synthesis of developmental and psychological interventions.

Mental health clinicians may participate in an interdisciplinary team in a variety of roles including direct care provision, as team leaders or members, or as consultants to other professionals. These clinicians should be active members of the team who share responsibility for leadership and decision-making, rather than being limited to "medication reviews." In many cases, especially in the community, the clinician will see the patient in his or her private office and there is no identified, regularly meeting team. In such cases it is essential to maintain close communication and collaboration with other professionals and caregivers who are involved with the patient.

Occasionally, arrangements for the care of patients who have MR involve nonphysicians conducting therapy, consulting with the caregivers, and in effect remaining the primary decision makers. Psychiatrists are consulted to prescribe medications only. Psychiatrists who elect to participate in such arrangements

should be aware that they remain legally responsible and accountable for the overall treatment. In such cases it is essential to have the opportunity to see the patient for follow-up as often and as long as necessary, to obtain sufficient information from all relevant sources, and to maintain close collaborations with all professionals and caregivers. A clear, preferably written, agreement should specify the responsibility of each professional, availability for emergencies, and their interaction (Woodward et al., 1993). Various roles of child and adolescent clinicians working with persons with MR are explored in a policy statement adopted by the American Academy of Child and Adolescent Psychiatry in 1987.

Guardianship

Parents are usually the legal guardians of children under the age of majority. However, they are not automatically legal guardians of their incompetent children over that age and must be appointed by a court to that position. Increasingly, the courts put incompetent persons under partial guardianship, such as for medical decisions only. Informed consent of the patient or a legal guardian is needed for the initiation of or major changes in the psychiatric treatment. If the patient is a minor, such consent is provided by a parent. Patients over the age of majority and who do not have a court-appointed guardian can provide this consent themselves, if the clinician judges that they are capable of making such decisions. The clinician has to be fully satisfied in making this judgment to avoid possible future liability for treatment without valid permission. If there are doubts about a patient's competency, the caregivers should be asked to initiate legal proceedings to assess competency and need for guardianship. In emergency situations a temporary medical guardianship can be obtained.

Competency and Psychotropic Drugs

The procedures for prescribing psychotropic drugs for incompetent persons differ in different states, and clinicians should be aware of applicable statutes in their jurisdictions. In some, a guardian's consent is sufficient, but in others, such as Massachusetts, separate court permission is required for the administration of antipsychotics to incompetent adults. However, if there is a justified emergency such as danger to the patient or to others, treatment may usually be initiated before legal procedures are completed.

Decisions Regarding the Site of Treatment

Treatment should occur in the least restrictive environment of care. In most cases outpatient settings are fine if appropriate services, even respite, can be secured. A mental disorder with major behavioral manifestations is no longer a common reason for a person with MR to be admitted to an institution. Private residential schools may admit children with special educational needs, including children with MR. The most common reason

for admission is the presence of a challenging behavior that makes the child's integration in the day school difficult. These facilities should be used only if all possible community-based placements and settings fail and the child needs a very structured, around-the-clock setting. The usual goal of these schools is to provide education and treatment for as short a period as possible, until the child can return to the community.

Other inpatient treatment settings include psychiatric hospitals, day hospitals, and respite facilities. The indications for hospitalization include risk of immediate danger to self or others or the need for observation and treatment in a very structured environment which cannot be secured on an outpatient basis. Unfortunately, not all inpatient psychiatric settings are familiar with the needs of persons with comorbid MR or equipped to provide them with an appropriate therapeutic and social milieu, habilitative and recreational programs, and other necessary services. However, inpatient and day hospital units specializing in patients with MR are being increasingly developed. In many localities "crisis teams" are established to respond to behavioral crises of persons with MR who live in the community. Such teams often have well-staffed respite residences where patients can be stabilized, perhaps averting hospitalization.

Compliance

To ensure the implementation and compliance with a treatment program, the education of and ongoing support for caregivers and patients (according to their cognitive abilities and understanding) is important. This support is particularly relevant to psychopharmacological and behavioral interventions. If the caregivers and patients do not bring up their concerns about it, this should be actively explored by the clinician.

CONFLICT OF INTEREST

As a matter of policy, some of the authors of these practice parameters are in active clinical practice and may have received income related to treatments discussed in these parameters. Some authors may be involved primarily in research or other academic endeavors and also may have received income related to treatments discussed in these parameters. To minimize the potential for these parameters to contain biased recommendations due to conflict of interest, the parameters were reviewed extensively by Work Group members, consultants, and Academy members; authors and reviewers were asked to base their recommendations on an objective evaluation of the available evidence; and authors and reviewers who believed that they might have a conflict of interest that would bias, or appear to bias, their work on these parameters were asked to notify the Academy.

SCIENTIFIC DATA AND CLINICAL CONSENSUS

Practice parameters are strategies for patient management, developed to assist clinicians in psychiatric decision-making.

These parameters, based on evaluation of the scientific literature and relevant clinical consensus, describe generally accepted approaches to assess and treat specific disorders, or to perform specific medical procedures. The validity of scientific findings was judged by design, sample selection and size, inclusion of comparison groups, generalizability, and agreement with other studies. Clinical consensus was determined through extensive review by the members of the Work Group on Quality Issues, child and adolescent psychiatry consultants with expertise in the content area, the entire Academy membership, and the Academy Assembly and Council.

These parameters are not intended to define the standard of care; nor should they be deemed inclusive of all proper methods of care or exclusive of other methods of care directed at obtaining the desired results. The ultimate judgment regarding the care of a particular patient must be made by the clinician in light of all the circumstances presented by the patient and his or her family, the diagnostic and treatment options available, and available resources. Given inevitable changes in scientific information and technology, these parameters will be reviewed periodically and updated when appropriate.

APPENDIX: MENTAL RETARDATION RESOURCES

Listing the following resources does not constitute their endorsement by AACAP. They are included here for convenience and as a potential source of information about other resources and services (including educational advocates) in a given locality.

Local Sources of Information

Depending on the state, clinicians and families can obtain information on local services, other resources, and entitlement to services from the state department of mental retardation, the state department of public health, or the state department of education.

University-Affiliated Programs for Persons With Developmental Disabilities

These programs, located in all states, focus on provision of clinical services, training professionals, and research related to developmental disabilities. Clinicians might refer patients to such programs and/or use them as sources of information about services in their areas. The addresses and details about the University-Affiliated Programs may be obtained from:

American Association of University Affiliated Programs for Persons With Developmental Disabilities
8630 Fenton Street, Suite 410
Silver Spring, MD 20910-3803
(301) 588-8252
http://www.aauap.org

National Organizations Concerned With Services for Persons With Developmental Disabilities and Their Families

These offices can provide addresses of the local chapters, which might be a good source of information concerning local services:

The ARC (formerly Association for Retarded Citizens)
500 East Border Street, Suite 300
Arlington, TX 76010
(617) 261-6003
http://www.thearc.org

Autism Society of America
7910 Woodmont Avenue, Suite 650
Bethesda, MD 20814-3015
(800) 3AUTISM x150
http://autism-society.org/

National Down Syndrome Congress
7000 Peachtree-Dunwoody Road, N.E.
Lake Ridge 400 Office Park
Building 5-Suite 100
Atlanta, GA 30328
(800) 232-NDSC

Williams Syndrome Association
1312 N Campbell, Suite 34
Royal Oak, MI
(248) 541-3630
http://www.williams-syndrome.org

National Fragile X Foundation
1441 York Street
Denver, CO 80206
(800) 688-8765

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