Psychiatric Disorders in Patients with Mental Retardation

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INTRODUCTION

Until recently, the diagnosis and treatment of psychiatric disorders in individuals with mental retardation has been a long-neglected aspect of the practice of psychiatry. Individuals with mental retardation and developmental delays experience the same psychiatric disorders and associated morbidities as the general population. In fact, the prevalence of mental illness associated with mental retardation is considered to be several times greater than that of individuals with no significant deficit in cognitive ability and adaptive functioning. A complete and competent psychiatric evaluation of patients with mental retardation does not greatly differ from the assessment of those without mental retardation. With mindful consideration of a patient’s developmental abilities, largely related to language use and comprehension, the general psychiatrist has the capacity to profoundly improve the quality of life for those with the greatest of needs.

MENTAL RETARDATION

DEFINITION

Mental retardation is a generalized deficit in cognitive ability together with adaptive functioning below the average range. The Diagnostic and Statistical Manual of Mental Disorders, 4th edition, text revision (DSM-IV-TR) and the 10th revision of the International Statistical Classification of Diseases and Related Health Problems consider an IQ of 70 or less to indicate mental retardation. Impairment in social adaptation and functioning also must be apparent in order to meet diagnostic criteria. Adaptive skills include abilities on the job and at school, as well as with friends or at home. Adaptive functioning is diminished when the individual has the ability to perform certain behaviors, but the behaviors are not reliably executed. In order to meet the diagnostic criteria for mental retardation, the diagnosis must occur before age 18 years. The American Association on Mental Retardation’s definition is similar to that of the DSM-IV-TR.

The DSM-IV-TR divides mental retardation into 4 categories of severity (Table 1): mild, moderate, severe, and profound. Of those with mental retardation, approximately 85% have the mild form of the disorder, 10% have the moderate form, 3% to 4% have the severe form, and 1% to 2% have the profound form.

EPIDEMIOLOGY

According to most estimates, approximately 1% of the US population meets the criteria for mental retardation. Mental retardation is approximately 1.5 times more common in males than in females. An IQ below 70 is equivalent to 2 standard deviations below the mean on most psychometric tests. If IQ were the sole criterion for diagnosing mental retardation, the prevalence rate would increase to 3%, thus demonstrating the importance of adaptive functioning impairment in making the diagnosis. These prevalence rates assume that IQ remains constant, but in some cases that assumption is incorrect. Persons with Down syndrome may demonstrate their highest IQ scores within their first year of life. These scores gradually decline until middle childhood. Boys with fragile X syndrome often begin to show a decline in IQ when they are between the ages of 10 and 15 years.

ETIOLOGY

The most common known causes of mental retardation are Down syndrome, fragile X syndrome, and fetal alcohol syndrome (FAS). In combination, these 3 causes are responsible for 30% of identified cases of mental retardation. The body of knowledge regarding the many etiologies of mental retardation continues to expand. These causes include genetic syndromes (eg, Down syndrome, fragile X syndrome), a variety of developmental and metabolic disorders, and pre- and postnatal toxic
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Exposures (eg, FAS). The etiology of mild cases of mental retardation is unclear, but cases of severe and profound mental retardation are more likely to have one or more identifiable causes for their delay and often show comorbid neurologic disease.

Some chromosomal abnormalities are inherited from a parent, but most occur de novo. Unfortunately, in 30% to 50% of all cases of mental retardation, the etiology is unidentified even after a thorough diagnostic evaluation.5

Chromosomal Abnormalities and Genetic Syndromes

Down syndrome. The occurrence of Down syndrome is thought to be 1 in every 600 to 800 live births. There is a positive correlation between Down syndrome and advanced maternal age.³ Down syndrome is the most common chromosomal abnormality leading to mental retardation; the syndrome results from an extra copy of chromosome 21, which is usually a sporadic event. Two percent of cases may be inherited from a balanced translocation carrier parent.

Early clinical manifestations of Down syndrome include a flat facial profile, hypotonia, up-slanting palpebral fissures, small ears, in-curving fifth fingers, and single transverse palmar creases. Language is often significantly impaired in those with Down syndrome. Visual processing remains relatively intact, compared with normal control groups, but auditory processing can be significantly impaired. Persons with Down syndrome are often described as considerably social.

Individuals with Down syndrome have an increased risk of developing thyroid abnormalities and congenital heart disease. A possible overproduction of β-amyloid caused by a defect at band 21q21.1 leads to an Alzheimer’s-type dementia in adulthood. Commonly noted behavioral problems in children with Down syndrome include attention difficulties, impulsivity, hyperactivity, and occasional aggression. Depression, autism, and pervasive developmental disorders appear to be relatively rare, although they are likely more common among persons with Down syndrome than in unaffected control groups.4

Fragile X syndrome. Fragile X syndrome is the most common inherited cause of mental retardation, affecting 1 in 1000 male births and 1 in 3000 female births.5 Males are more severely affected by the disorder than are females. The pathophysiology involves an inactivation of the FMR-1 gene located on the X chromosome at band Xq27.3 caused by CGG (cytosine, guanine, guanine) base repeats.6 The process of excessive repetition of a 3-nucleotide sequence in DNA is called amplification.6 Triple repeat exceeding 200 will likely result in the full phenotypic syndrome. Numbers of repeats between 50 and 200 are termed premutations. As many as 1 in 259 women in the general population may carry the pre-mutation.4

Clinical manifestations of the disorder include macrocephaly, large ears, macroorchidism, and hyperextensible fingers.³ Fragile X syndrome also may be associated with mitral valve prolapse.
Approximately 50% of fully affected females show mild to moderate mental retardation, and the other 50% have average IQ scores. Those with average intelligence, however, often show social dysfunction or anxiety, as well as significant difficulties with planning, memory, and attention. Fully affected males often demonstrate autistic-like symptoms such as language delay, stereotypy, self-injurious behavior, poor eye contact, and tactile sensitivity. Although controlled studies show that only approximately 5% of males with fragile X syndrome have autistic disorders, many display social anxiety and avoidance common with anxiety disorders or pervasive developmental disorders. In addition, symptoms of attention-deficit/hyperactivity disorder (ADHD) are seen in the vast majority of both boys and girls with fragile X syndrome.

**Prader-Willi syndrome.** Prader-Willi syndrome affects approximately 1 in 10,000 to 15,000 live births. It is caused by a deletion of paternal origin at band 15q12. Ninety percent of cases are sporadic. The syndrome is associated with borderline to moderate mental retardation.

Clinical characteristics in infants with Prader-Willi syndrome include hypotonia and feeding difficulties. Beginning around 2 years of age, children frequently develop hyperphagia and are often known to forage and hoard food. The leading causes of death for people with this syndrome are complications relating to obesity, which is likely caused by a hypothalamic abnormality resulting in lack of satiety. Other clinical features of the disorder include microorchidism, cryptorchidism, short stature, fair hair and skin, scoliosis, and orthopaedic problems. Psychiatric comorbidities include compulsive behavior, impulsivity, emotional lability, daytime sedation, skin picking, and anxiety.

**Angelman's syndrome.** Angelman’s syndrome involves a maternal deletion at band 15q12. Its prevalence is estimated to be 1 in every 20,000 to 30,000 births. Persons with Angelman’s syndrome are usually profoundly mentally retarded compared with persons who have Prader-Willi syndrome (caused by the same chromosomal deletion but of paternal origin), who usually have borderline to moderate mental retardation.

Clinical features of the disorder include fair hair and blue eyes in 66% of those affected, with a wide smiling mouth, thin upper lip, and pointed chin. Epilepsy occurs in 90% of persons with Angelman’s syndrome, and 25% of patients show microcephaly; ataxia also is frequently present. Behavioral characteristics of those with Angelman’s syndrome include a reportedly happy disposition, paroxysmal laughter, and hand flapping and clapping.

**Williams syndrome.** Williams syndrome occurs in approximately 1 in 20,000 births and is caused by an autosomal dominant hemizygous deletion at band 7q11.23. IQ in individuals with Williams syndrome usually ranges from 40 to 80, placing most affected individuals in a mild to moderate category of mental retardation. Clinical features of the disorder include short stature and elfin-like facies (eg, broad forehead, depressed nasal bridge, widely spaced teeth, full lips). Medical comorbidities include hypercalcemia and renal, cardiovascular, and thyroid abnormalities. Behavioral phenotypes include anxiety and hyperactivity. Persons with Williams syndrome are often known to be outgoing and sociable, with verbal skills exceeding their visual-spatial skills.

**Toxic Exposures**

FAS, the toxic exposure syndrome most commonly responsible for mental retardation, is caused by the mother’s alcohol consumption during pregnancy. Between 0.05 and 3 in every 1000 births are diagnosed with FAS annually in the United States. The diagnosis is usually made at birth and is based on the mother’s medical history and the baby’s facial features. Typical features include medial epicanthal folds, a wide nasal bridge, small upturned nose, long philtrum, and narrow upper lip. Low birth weight and body measurements also are common. FAS usually involves mild to moderate mental retardation, but the severity of central nervous system dysfunction is related to the amount of maternal alcohol consumption. Poor academic achievement and behavioral problems are typical for children with FAS. Psychiatric conditions associated with fetal alcohol syndrome include ADHD, seizures, irritability, autism, and memory impairment.

Lead poisoning is another important cause of mental retardation and is usually a result of ingesting flakes of old lead-based paint. Young children are most at risk because of the continued development of the immature central nervous system. Headache and gastrointestinal symptoms of lead poisoning predominate acutely. Late symptoms include developmental retardation, seizures, ataxia, and personality changes.

Prenatal exposure to cocaine may lead to a wide variation of intellectual capacity, but persons exposed often seem to have difficulties with attention, poor impulse control, and decreased academic achievement.

**Metabolic Abnormalities**

Inborn errors of metabolism are known to be a significant cause of mental retardation. Some of the more
common metabolic disorders associated with mental retardation include Lesch-Nyhan syndrome, phenylketonuria (PKU), homocystinuria, and Tay-Sachs disease. The clinical disorder is often the result of a genetic enzyme defect. The key clinical features of these disorders are more fully described in Table 2.

**Lesch-Nyhan syndrome.** Lesch-Nyhan syndrome is an X-linked recessive disorder characterized by a defect in the production of hypoxanthine-guanine phosphoribosyltransferase, resulting in an accumulation of uric acid. The site of the genetic defect is band Xq26-27. The syndrome occurs in 1 in every 10,000 to 38,000 births. Lesch-Nyhan syndrome is often associated with ataxia, chorea, renal failure, and gout. Individuals with the disorder often show mild to moderate mental retardation, anxiety, aggression, and, most notably, severe self-biting behavior.

### Table 2. Examples of Inborn Errors of Metabolism Causing Mental Retardation*

<table>
<thead>
<tr>
<th>Disorder</th>
<th>Enzyme Defect†</th>
<th>Onset/Life Expectancy</th>
<th>Clinical Features</th>
<th>Laboratory Diagnosis‡</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Aminoacidurias</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Phenylketonuria (PKU)</td>
<td>Phenylalanine hydroxylase deficiency</td>
<td>I/A</td>
<td>If not on diet low in phenylalanine: vomiting, musty odor, eczema, seizures, tremors, psychosis</td>
<td>U: ferric chloride test; gene locus 12q22-24</td>
</tr>
<tr>
<td>Homocystinuria</td>
<td>Cystathionine β-synthase deficiency</td>
<td>I/A</td>
<td>Seizures, venous thromboses, cerebrovascular accidents, Marfan’s habitus, malar flush, lens subluxation, often MR</td>
<td>U: cyanide-nitroprusside test</td>
</tr>
<tr>
<td><strong>Mucopolysaccharidoses</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>MPS I (Hurler’s syndrome)</td>
<td>L-Iduronidase deficiency</td>
<td>I/10 y</td>
<td>Early facial feature coarsening, hepatosplenomegaly, growth failure, corneal clouding, skeletal changes</td>
<td>U: heparan sulfate, dermatan sulfate</td>
</tr>
<tr>
<td>MPS II (Hunter’s syndrome)</td>
<td>Iduronate-2-sulfatase deficiency</td>
<td>I/15 y</td>
<td>Symptoms milder and progression slower than in MPS I</td>
<td>U: heparan sulfate, dermatan sulfate</td>
</tr>
<tr>
<td><strong>Sphingolipidoses</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Tay-Sachs disease (GM1)</td>
<td>GM1 ganglioside-N-acetylhexosaminidase</td>
<td>3–6 mo/2–3 y</td>
<td>Vision and hearing deficits, macular “cherry red” spot, muscular atrophy and paralysis, death usually by age 5</td>
<td>Serum hexosaminidase assay</td>
</tr>
<tr>
<td>Metachromatic leukodystrophy</td>
<td>Arylsulfatase A deficiency</td>
<td>1–4 y/10–15 y</td>
<td>Hypotonia, seizures, blindness, rigidity, paralysis, and mental deterioration</td>
<td>U: metachromatic cells, sulfatase A assay; sural nerve biopsy</td>
</tr>
</tbody>
</table>

A = adulthood; I = infancy; MR = mental retardation; U = urinary.

*All listed disorders cause mental retardation except homocystinuria, in which mental retardation does not occur in every case.

†Inheritance of all listed disorders is autosomal recessive, except for MPS II, which is X-linked.

‡Prenatal diagnosis is available for all listed disorders.

Phenylketonuria. PKU is an autosomal recessive disorder caused by a deficiency of phenylalanine hydroxylase resulting in an accumulation of phenylalanine. Occurring in approximately 1 in every 11,500 births, PKU is linked with a defect at band 12q22-24.1. All children in the United States are screened for PKU at birth. Symptoms are absent at birth, with seizure developing later in childhood. PKU is characterized by very fair skin, blue eyes, and blonde hair. If individuals with PKU are left untreated without diet restrictions, mild to moderate mental retardation may result. Other aspects of the behavioral phenotype of PKU include language delay, destructiveness, self-injury, and hyperactivity.

PSYCHIATRIC EVALUATION OF PATIENTS WITH MENTAL RETARDATION

MEDICAL EVALUATION

Although the medical care of a patient with mental retardation is ideally managed by the patient’s primary care physician, the psychiatrist evaluating a mentally retarded person with mental health concerns may be in a unique position to ensure that the appropriate initial medical work-up has been completed. This evaluation should include a thorough history, physical examination, structural neuroimaging, and appropriate laboratory studies, including genetic testing.

History

Knowing the underlying cause of a patient’s disability is helpful when evaluating patients with mental retardation, because some medical and psychiatric diagnoses are more prevalent in specific disabling conditions. As in all evaluations, obtaining an accurate clinical history of each patient is of the utmost importance; this information should include a prenatal and birth history, as well as any complications or exposures to toxic substances. Obtaining a 3-generation family history also is imperative. Pertinent questions regard relatives with learning problems, mental retardation, neurologic disorders, psychiatric disorders, or other problems resembling those of the patient.

Physical Examination

A thorough physical examination also is a critical part of patient evaluation. Patients should be examined closely for dysmorphic features or abnormalities. Most minor abnormalities involve the face, ears, hands, or feet and are easily recognized. The presence of 3 or more minor abnormalities in a newborn correlates with a 90% frequency of coexisting major abnormalities.

Additional Diagnostic Evaluations

Findings from the medical history and physical examination will help determine the next appropriate diagnostic tests or referrals to obtain. Metabolic studies, muscle biopsies, DNA molecular studies, chromosome analysis, and testing for fragile X syndrome may be necessary. Any abnormal neurologic findings will likely warrant neuroimaging. Magnetic resonance imaging (MRI) typically provides better anatomical resolution compared with computed tomography (CT), but a CT scan may be more useful than an MRI in locating possible intracranial calcification. Neuroimaging is indicated in patients with seizures, microcephaly, macrocephaly, neurologic signs (eg, spasticity, dystonia, altered reflexes), or loss of previously acquired skills. A referral for an evaluation by a geneticist should be considered if the diagnosis is not firmly established.

DIAGNOSING PSYCHIATRIC DISORDERS

Defining and diagnosing psychiatric disorders in patients with mental retardation have been longstanding challenges for psychiatrists and require a developmental approach. This approach calls for an assessment based on intellectual or developmental age rather than chronologic age. The possibility of differing expressions of subjective emotional content in patients with a limited repertoire of such expression must be fully considered. Limited communicative ability is often the greatest obstacle in diagnosing psychiatric disorders in patients with severe or profound mental retardation. Therefore, a collateral history of adaptive functioning obtained from multiple sources such as primary caregivers, school personnel, neuropsychiatric testing, and adaptive behavior ratings is the cornerstone of evaluation.

A factor analytic study of the Diagnostic Assessment for the Severely Handicapped Scale, involving 506 severely and profoundly mentally retarded persons, yielded 6 major factor scales in categorizing behavior in this population. These scales included (1) tantrums, (2) aggression/conduct disorder, (3) language disorder/verbal aggression, (4) social withdrawal/stereotypy, (5) eating disorders, and (6) sleep disorders. These 6 categories were found to be the most common causes of psychiatric consultation. Diagnostic outcome relied more heavily on vegetative symptoms than on more traditional methods of diagnosis. Another study involved 251 patients with severe and profound mental retardation who were consecutively referred for psychiatric consultation and grouped into 6 categories on the basis of chief complaint.
The categories used were similar to those just described. The most frequently made diagnoses following consultation were impulse control disorders, anxiety disorders, and mood disorders. The presence of these conditions in this population underscores the vital nature of psychiatric involvement in the multidisciplinary assessment and treatment of patients with mental retardation.

COMORBID PSYCHIATRIC DISORDERS

ATTENTION-DEFICIT/HYPERACTIVITY DISORDER

The prevalence rates of ADHD in patients with mental retardation are estimated to be between 8% and 19%. The presence of hyperactivity in patients who do not meet full diagnostic criteria for ADHD is even more common. Unfortunately, children and adults with cognitive disabilities often are not included in the research that has been conducted on the treatment of ADHD. For persons with mental retardation, the diagnosis of ADHD depends upon the presence of symptoms excessive for an individual’s mental age or remarkable compared with peers who have similar levels of retardation.

Stimulant treatment for ADHD may exacerbate conditions such as epilepsy, anxiety, tics, or obsessive behavior to a greater extent in mentally retarded patients than in unaffected control groups. Common adverse effects of stimulant medications (eg, sleep and appetite disturbance) also may be exaggerated in patients with mental retardation.

PERVASIVE DEVELOPMENTAL DISORDERS AND AUTISM

The pervasive developmental disorders (PDDs) are a group of clinical syndromes that present as both developmental delays and deviations from normotypic behaviors. PDDs differ from mental retardation in that they represent a unique set of aberrant behaviors. Key features include limited idiosyncratic language patterns, lack of social relatedness, persistent need for sameness, and frequent repetitive activities. The PDDs include autism, Rett syndrome, childhood disintegrative disorder, Asperger’s syndrome, and pervasive developmental disorder not otherwise specified. PDDs occur much more commonly in males than in females, and the majority of individuals with autism or PDDs are mentally retarded as well. Only 20% to 30% of persons with autism have an IQ of 70 or greater.

Stone et al studied the play and imitation skills of 22 autistic children, compared with the same play performance of 15 mentally retarded children, 15 hearing-impaired children, 19 language-impaired children, and 20 nonhandicapped children. All subjects were between 3 and 6 years of age. Motor imitation ability was the most important characteristic differentiating the autistic children from the others. The failure to use toys in a functional manner also was an indicator of autism in young children. These indicators may prove to be important areas for screening, given that the characteristics were relatively specific to autism.

ANXIETY DISORDERS

Anxiety is a common symptom in mental retardation and in PDDs and is frequently associated with obsessive and compulsive adherence to a routine. Because the diagnosis of specific anxiety disorders relies on an individual’s ability to communicate subjective symptoms of anxiety, making a diagnosis in patients with severe communication deficits is often difficult. Anxiety disorders appear to be underdiagnosed in persons with mental retardation; reports of prevalence rates between 1% and 25% in this population demonstrate the likely difficulty of establishing a diagnosis. Patients who are avoidant and exhibit excessive autonomic arousal in the face of stimuli that peers find tolerable are likely experiencing anxiety. Frequent symptoms of anxiety in mentally retarded persons include aggression, agitation, self-injury, and insomnia. These symptoms are often difficult to distinguish from impulse control disorders. A diagnosis of posttraumatic stress disorder is always an important consideration in a population that is at high risk for abuse.

PSYCHOSIS

A certain level of verbal competence is required to describe the typical clinical features of delusions and hallucinations in psychosis. Consequently, it is quite difficult to diagnose psychosis in patients with an IQ below 45. Some individuals may seem to respond to hallucinations or display catatonic postures, which may suggest psychosis. Irrespective of the difficulty in diagnosis, patients with intellectual impairment are at a significantly increased risk for schizophrenia, bipolar disorder, and other illnesses having the positive symptoms of psychosis and thought disorder. Likewise, patients with developmental disabilities are most likely to experience adverse effects from antipsychotic medication use, such as extrapyramidal side effects, tardive dyskinesia, and other movement disorders.

AFFECTIVE DISORDERS

Expression of the psychological symptoms of mood disorders may be limited in persons with mental retardation, especially among those with poor verbal skills. However, changes in neurovegetative symptoms are useful.
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in making a diagnosis. Changes in sleep, appetite, activity level, and interest often can cause sufficient impairment in daily activities to warrant psychiatric treatment. Aggression and self-injurious behavior may be symptoms of depressed mood; symptoms such as irritability, mood lability, aggression, and sleep disturbance may indicate bipolar disorder. Both rapid cycling and mixed affective states are more common in mentally retarded persons than in control populations. Learning problems, social skills deficits, poor coping capacity, low self-esteem, and real or perceived social isolation are risk factors for the development of mood disorders. Subclinical hypothyroidism and temporal lobe epilepsy are common medical comorbidities in mentally retarded persons and are frequently associated with rapid cycling bipolar disorder. Correction or treatment of the underlying abnormality could substantially improve the patient’s affective symptoms.

SLEEP DISORDERS

Sleep disorders are a persistent problem in persons with mental retardation, especially in the presence of sensory impairments such as deafness and blindness. Sleep hygiene and anxiety management are appropriate behavioral approaches for the treatment of sleep disorders. Benzodiazepine therapy for sleep disorders often can worsen behavior for those with mental retardation through confusion or disinhibition, leading to aggression. Benzodiazepines are essentially contraindicated in these patients.

DELIRIUM

Delirium is an important clinical consideration in diagnosing psychiatric disorders in patients with mental retardation. Clinicians should rule out delirium as a cause of behavioral deterioration by performing a physical examination, obtaining a medical history, and carefully evaluating a patient’s prescribed medications.

PSYCHIATRIC TREATMENT OF PATIENTS WITH MENTAL RETARDATION

A wide variety of therapeutic techniques have served to treat psychiatric disorders among patients with mental retardation. Those techniques most widely employed include behavioral treatments; individual, group, and family-oriented psychotherapies; and psychopharmacologic treatments.

BEHAVIORAL TECHNIQUES AND PSYCHOTHERAPY

Behavioral treatment techniques involve efforts aimed at increasing or enhancing desired behaviors and reducing or limiting behavioral excesses. These techniques are based largely on the principle of operant conditioning. Other behavioral interventions include desensitization, modeling, and social skills training.

In any form of psychotherapy, the clinician must use an active participatory style with concrete and supportive interventions, carefully regarding the language ability and developmental level of the patient. With these considerations, psychotherapy can be quite beneficial to patients with mental retardation. All therapeutic interventions are most effective when they are conducted in the context of a treatment plan that includes educators, clinicians, caretakers, and, especially, the patient’s family members.

PSYCHOPHARMACOLOGIC THERAPY

Relatively few well-controlled studies have examined the use of medication to manage mental disorders in mentally retarded persons. Conventional practice dictates that these patients respond in a pattern similar to that of normotypic individuals; however, there are some exceptions. Individuals with mental retardation often are quite sensitive to the adverse effects of psychotropic medication, and physicians should use caution when determining the correct dosage to administer to these patients.

Double-blind crossover studies have shown methylphenidate to be superior to placebo in treating ADHD among mentally retarded children. One study involved children with IQ scores ranging from 50 to 74 who were rated on the Conners’ Hyperactivity Index. Seventy-five percent of subjects showed significant increases in work output and attention skills with methylphenidate use. The rates of efficacy were similar to those in the non-mentally retarded population.15

Several open trials have demonstrated the efficacy of selective serotonin reuptake inhibitors in treating anxiety and depressive disorders. One open trial of fluoxetine showed significantly reduced scores on a Hamilton Depression Rating Scale in adults with mental retardation.16

An open nonblind trial of clomipramine reduced adventitious movements and compulsions in 5 prepubertal boys with autism and severe mental retardation.17 Another placebo-controlled study demonstrated that clomipramine use improved symptoms of hyperactivity, rituals, anger, and stereotypic movements in prepubertal boys with autistic disorder and severe mental retardation.12

Stereotypic movements also have been treated frequently with antipsychotic agents, specifically to decrease destructive behaviors. Clearly, because of the risk of tardive dyskinesia, atypical antipsychotic agents are
preferred to conventional neuroleptics in such conditions.

Several clinical trials have found lithium and valproate to be beneficial in treating mentally retarded patients with aggression, mood lability, and self-injurious behavior. Valproate is currently the drug of choice in managing bipolar symptoms. Case reports have described valproate as an effective therapy in treating aggression without any evidence of emotional arousal or anger, which is occasionally seen in patients with severe and profound mental retardation. Such behavior has been termed nonaffective aggression.

β-Blockers and buspirone have been used with mixed results in treating aggression and irritability. The opioid antagonist naltrexone hydrochloride has been hypothesized to improve self-injurious behavior in mentally retarded patients, but reported results have varied considerably. Several case reports have demonstrated dramatic improvement in severe self-destructive behavior with naltrexone treatment. However, naltrexone was tested in a double-blind placebo-controlled study involving a group of 32 mentally retarded adults, and was found to have no clinical value in reducing self-injurious behavior.

Since research data are limited, it is advisable to treat each patient on an individual basis, starting with the safest and most commonly used drugs in low doses and increasing the dosage gradually. Differing classes of medications should be used in a systematic manner.

MEDICAL COMORBIDITIES

PREVALENCE OF MEDICAL DISORDERS IN PATIENTS WITH MENTAL RETARDATION

Several developmental disorders have special health issues that must be considered in the diagnosis and treatment of psychiatric disorders. Almost 100% of persons with Down syndrome demonstrate early Alzheimer’s disease by the age of 40 years. Down syndrome is associated with extensive medical disease, including deafness (75% of patients), eye disease (65%), congenital heart disease (50%), thyroid disease (15%), seizure disorders (12%–50%), and gastrointestinal atresias (12%). Diabetes mellitus also is especially prevalent in patients with Down syndrome.

In men and boys with fragile X syndrome, there is a 60% prevalence of recurrent serous otitis media, a 22% to 77% prevalence of cardiac disorders (including mitral valve prolapse), a 30% to 56% prevalence of ocular disorders, and a 40% to 50% prevalence of seizures. FAS is linked to disorders of vision in 94% of persons with the syndrome, recurrent otitis media in 93%, heart defects in 29% to 41%, hearing loss in 6%, and significant genitourinary tract abnormalities in 10%.

Obesity is quite common in persons with mental retardation. The prevalence of obesity in women with Down syndrome is 3 times as high as that of the general population. Obesity occurs in men with Down syndrome twice as often as in the general population.

Epilepsy is the most common medical disorder in patients with mental retardation, occurring in up to 18% of persons with mild mental retardation and 36% of persons with severe impairment. A study of psychiatric disorders in 98 children and adolescents with mental retardation and active epilepsy demonstrated that 59% had at least 1 psychiatric disorder; 33% could not be classified because of profound or severe mental retardation. Thus, 90% of children with epilepsy whose mentally handicapping conditions could be categorized met criteria for at least 1 psychiatric disorder. The most common psychiatric diagnosis was autistic disorder (27% of patients), with a male-female ratio of 2 to 1. Sixty-seven percent of those children with autistic disorder had severe mental retardation, and 51% of the children displayed self-injurious behavior. Hyperactivity, rage, antisocial behavior, and psychosis have been reported particularly in connection with temporal lobe epilepsy.

MEDICAL CONSIDERATIONS IN PSYCHOPHARMACOLOGIC THERAPY

It is critical to be mindful of comorbid medical disorders when prescribing psychotropic medication to patients. Tricyclic antidepressants and some antipsychotic agents may prolong the QT interval, placing patients with cardiac anomalies at increased risk. Lithium may worsen the thyroid functioning of a patient already diagnosed with primary hypothyroidism. Thyroid dysfunction caused by medication is more common among patients with mental retardation than among the general population. Clinicians prescribing atypical antipsychotic agents such as risperidone and olanzapine must monitor weight gain closely in mentally retarded patients.

IMMUNIZATIONS

Immunization status must be optimized in all patients with mental retardation. Most mentally retarded patients benefit from influenza and pneumococcal vaccines. Patients with mental retardation are at a higher risk for hepatitis A and B, and immunizations against both diseases should be offered.
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LEGAL CONSIDERATIONS

Several important legal issues affect the lives of persons with mental retardation. One of the most well-known laws, the Americans with Disabilities Act of 1990, provides a clear mandate for the elimination of discrimination against persons with disabilities, provides enforceable standards addressing discrimination, and ensures that the federal government will enforce those standards on behalf of the disabled.

Section 504 of the Rehabilitation Act of 1973, the Individuals with Disabilities Education Act of 1990, and the Americans with Disabilities Act collectively provide for developmental assessments of children older than 3 years in every school district. These laws mandate that all children receive a free, appropriate public education in the least restrictive environment and that reasonable accommodation is made for the employment of adults with disabilities as well.

Physicians also must be aware of local guardianship, custodial care, informed consent, and abuse reporting laws in order to accurately serve their patients with mental retardation. Local advocacy organizations and legal agencies such as boards for mental retardation can be helpful in educating physicians regarding these issues (Table 3). Issues of sexuality, family planning, and estate planning are important considerations for the patient and his or her family. Despite issues of medical guardianship, clinicians must make every effort to discuss medical decision making with their patients on a developmentally appropriate level.

BOARD REVIEW QUESTIONS

Choose the single best answer for each question.

1. Which of the following is the most common inherited cause of mental retardation?
   A) Down syndrome
   B) Fragile X syndrome
   C) Prader-Willi syndrome
   D) Williams syndrome

2. Which of the following terms describes the process of excessive repetition of a 3-nucleotide sequence in DNA?
   A) Amplification
   B) Anticipation
   C) Premutation
   D) Translocation

3. Which of the following statements regarding Angelman’s syndrome is INCORRECT?
   A) Angelman’s syndrome involves a paternal deletion at band 15q12

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Table 3. Advocacy Groups and Information Sources for Mental Retardation

<table>
<thead>
<tr>
<th>The Arc of the United States</th>
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<tbody>
<tr>
<td>1010 Wayne Avenue, Suite 650</td>
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<tr>
<td>Silver Spring, MD 20910</td>
</tr>
<tr>
<td>(301) 565-3842</td>
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<td><a href="http://www.thearc.org">http://www.thearc.org</a></td>
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<tr>
<th>American Association on Mental Retardation</th>
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<tbody>
<tr>
<td>444 North Capitol Street, NW</td>
</tr>
<tr>
<td>Suite 846</td>
</tr>
<tr>
<td>Washington, DC 20001-1512</td>
</tr>
<tr>
<td>(800) 424-3688</td>
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<td><a href="http://www.aamr.org">http://www.aamr.org</a></td>
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<th>CAPP National Resource Center</th>
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<tr>
<td>Federation for Children with Special Needs</td>
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<tr>
<td>1135 Tremont Street, Suite 420</td>
</tr>
<tr>
<td>Boston, MA 02120</td>
</tr>
<tr>
<td>(617) 236-7210</td>
</tr>
<tr>
<td>(800) 331-0688</td>
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<tr>
<td><a href="http://www.fcsn.org">http://www.fcsn.org</a></td>
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<tr>
<th>Clearinghouse on Disability Information</th>
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<tr>
<td>Office of Special Education and Rehabilitative Services</td>
</tr>
<tr>
<td>330 C Street, SW</td>
</tr>
<tr>
<td>Switzer Building, Room 3132</td>
</tr>
<tr>
<td>Washington, DC 20202-2524</td>
</tr>
<tr>
<td>(202) 205-8241</td>
</tr>
<tr>
<td><a href="http://www.ed.gov/offices/OSERS/">http://www.ed.gov/offices/OSERS/</a></td>
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<tr>
<th>FRAXA Research Foundation, Inc. (for fragile X syndrome)</th>
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<tbody>
<tr>
<td>45 Pleasant Street</td>
</tr>
<tr>
<td>Newburyport, MA 01950</td>
</tr>
<tr>
<td>(978) 462-1866</td>
</tr>
<tr>
<td><a href="http://www.fraxa.org">http://www.fraxa.org</a></td>
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<th>National Organization on Fetal Alcohol Syndrome</th>
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<tr>
<td>216 G Street, NE</td>
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<tr>
<td>Washington, DC 20002</td>
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<tr>
<td>(202) 785-4585</td>
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<tr>
<td><a href="http://www.nofas.org">http://www.nofas.org</a></td>
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LEGAL CONSIDERATIONS

Several important legal issues affect the lives of persons with mental retardation. One of the most well-known laws, the Americans with Disabilities Act of 1990, provides a clear mandate for the elimination of discrimination against persons with disabilities, provides enforceable standards addressing discrimination, and ensures that the federal government will enforce those standards on behalf of the disabled.

Section 504 of the Rehabilitation Act of 1973, the Individuals with Disabilities Education Act of 1990, and the Americans with Disabilities Act collectively provide for developmental assessments of children older than 3 years in every school district. These laws mandate that all children receive a free, appropriate public education in the least restrictive environment and that reasonable accommodation is made for the employment of adults with disabilities as well.

Physicians also must be aware of local guardianship, custodial care, informed consent, and abuse reporting laws in order to accurately serve their patients with mental retardation. Local advocacy organizations and legal agencies such as boards for mental retardation can be helpful in educating physicians regarding these issues (Table 3). Issues of sexuality, family planning, and estate planning are important considerations for the patient and his or her family. Despite issues of medical guardianship, clinicians must make every effort to discuss medical decision making with their patients on a developmentally appropriate level.
B) Behavioral characteristics of Angelman’s syndrome include paroxysmal laughter and hand flapping
C) Clinical features of Angelman’s syndrome include fair hair and blue eyes
D) Persons with Angelman’s syndrome are usually profoundly mentally retarded

4. A possible overproduction of which of the following substances by a defect at band 21q21.1 leads to an Alzheimer’s-type dementia in adulthood for individuals with Down syndrome?
   A) Aluminum
   B) Apolipoprotein E-2
   C) β-Amyloid
   D) Presenilin 2

5. Which of the following criteria is consistent with the DSM-IV-TR diagnostic criteria for mental retardation?
   A) An IQ of approximately 70 or below must be present
   B) Impairment in adaptive functioning in at least 2 areas must be present
   C) Onset of mental retardation occurs before 5 years of age
   D) Profound mental retardation is based on an IQ of 35 or below

ANSWERS

1. B
2. A
3. A
4. C
5. B

REFERENCES


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