CALIFORNIA STATE BOARD OF PHARMACY

Special Edition in cooperation with
CALIFORNIA HEALTH AND HUMAN SERVICES AGENCY

HEALTH NOTES

Care of Children & Adults with Developmental Disabilities

Consumer Education and Communication Committee

Dedicated to building bridges of communication with those Californians whose health depends on proper drug therapy, compliance with a treatment regimen and a healthier lifestyle.

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Introduction

HEALTH NOTES is published by the California State Board of Pharmacy's Consumer Education and Communication Committee to assist California pharmacists and other health care providers to be better informed on subjects of importance to California consumers.

This issue of *HEALTH NOTES* addresses the pharmacist's role in providing counseling, health information, and health-related services to a unique and often underserved group of individuals–children and adults with developmental disabilities.

In California, the term "developmental disability" is defined by the State and refers to a severe and chronic disability that is attributable to a mental or physical impairment. The disability must begin before the person's 18th birthday, be expected to continue indefinitely, and present a substantial disability. Also, the disability must be due to one of the following conditions: mental retardation, cerebral palsy, epilepsy, autism, or a disabling condition closely related to mental retardation or requiring similar treatment. The Department of Developmental Services is the agency through which the State of California provides services and support to children and adults with developmental disabilities.

Because of communication barriers, physical limitations, drug sensitivity, and other health-related issues, individuals with developmental disabilities can be especially vulnerable to the incorrect use of medications.

Pharmacists can have a valuable influence on the health of Californians with developmental disabilities by providing medication information and therapeutic resources that will help persons with developmental disabilities and their caregivers to properly manage and monitor medications and to ensure that drug therapy outcomes are effective.

We anticipate that this issue of *HEALTH NOTES* will be a valuable resource to assist pharmacists and other health care providers to better understand the special health care needs of individuals with developmental disabilities.

Marilyn Standifer Shreve
Editor, Health Notes
Chair, Consumer Education and Communication Committee

Statewide Overview

I am honored to be a part of the State Board of Pharmacy's publication *HEALTH NOTES*, which is highlighting the important issue of caring for children and adults with developmental disabilities.

As Secretary of the California Health and Human Services Agency, I oversee thirteen State departments and two boards. These include the Departments of Aging, Alcohol and Drug Programs, Community Services and Development, Developmental Services, Employment Development, Health Services, Mental Health, Rehabilitation, and Social Services as well as the California Workplace Investment Board, Emergency Medical Services Authority, Health and Welfare Data Center, Managed Risk Medical Insurance Board, Office of Statewide Health Planning and Development, and the Department of Child Support.

Making sure all Californians are knowledgeable about the proper use of medication, especially for those with developmental disabilities, is of the utmost importance to me. Vulnerable individuals oftentimes have complex medical issues and medication regimens. Because of these medical issues, people with developmental disabilities often require additional assistance with their care.

This assistance includes the pharmacist providing special consultation and education to the person with the developmental disabilities, family members, and caregivers regarding the medication prescribed. It also requires additional monitoring of the prescribed medication for side effects and appropriate utilization.

I am pleased that the State Board of Pharmacy and the Department of Developmental Services have recognized the importance of continuing education for pharmacists and professionals who serve people with developmental disabilities. I am confident that you will find the following articles both educational and helpful as you continue to provide service to Californians with developmental disabilities.

Grantland Johnson
Secretary, California Health and Human Services Agency

HEALTH NOTES

VOLUME 1, NUMBER 4

Care of Children & Adults with Developmental Disabilities

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** The views expressed by the individual authors are not necessarily the view of the Board of Pharmacy.*

The Department of Developmental Services (DDS) is one of 13 departments composing the California Health and Human Services Agency. With an annual budget of over $1.9 billion, DDS provides services to more than 150,000 children and adults with developmental disabilities. These services are provided through State-operated developmental centers and contracts with 21 nonprofit agencies known as regional centers. Through our Wellness Initiative we are developing partnerships with State agencies, universities, and professionals to promote the health and well-being of Californians' with developmental disabilities.

DDS is collaborating on many educational projects with regional centers, medical schools, and professional associations. We are pleased to be a part of the State Board of Pharmacy's educational edition of the *HEALTH NOTES* "Care of Children & Adults with Developmental Disabilities." We hope you enjoy the articles and find them useful. Thank you for your continuing dedication and commitment to providing needed medical services to individuals with developmental disabilities.

Cliff Allenby
Director, Department of Developmental Services

TO FAMILIES AND CAREGIVERS:

The purpose of this *HEALTH NOTES* is to open the door to better communication between California pharmacists and their patients. We know that successful communication between people requires the use of language that all participants understand. In addition, it is helpful for everyone to have an understanding of each other's circumstances.

In order to be most helpful, pharmacists need to know about their patients–this includes how they live, their eating and sleeping habits, how they get around in the community, and special problems they might have such as allergies or difficulty with swallowing. Similarly, patients and their caregivers need to know about their pharmacists–the medical terms they use, the types of physical symptoms they watch for, and especially their ability to clarify directions for the use of medications.

We hope that you will look through this monograph and find articles that interest you, and that teach you new things about people with developmental disabilities. If you use this information, I know that you and the people you live and work with will be healthier.

One important mission of the Board of Pharmacy is to promote better health through increased communication between pharmacists and the people who look to them for their health needs. We hope you find that this issue of *HEALTH NOTES* helps you work with your pharmacist more closely.

Good Health to You!

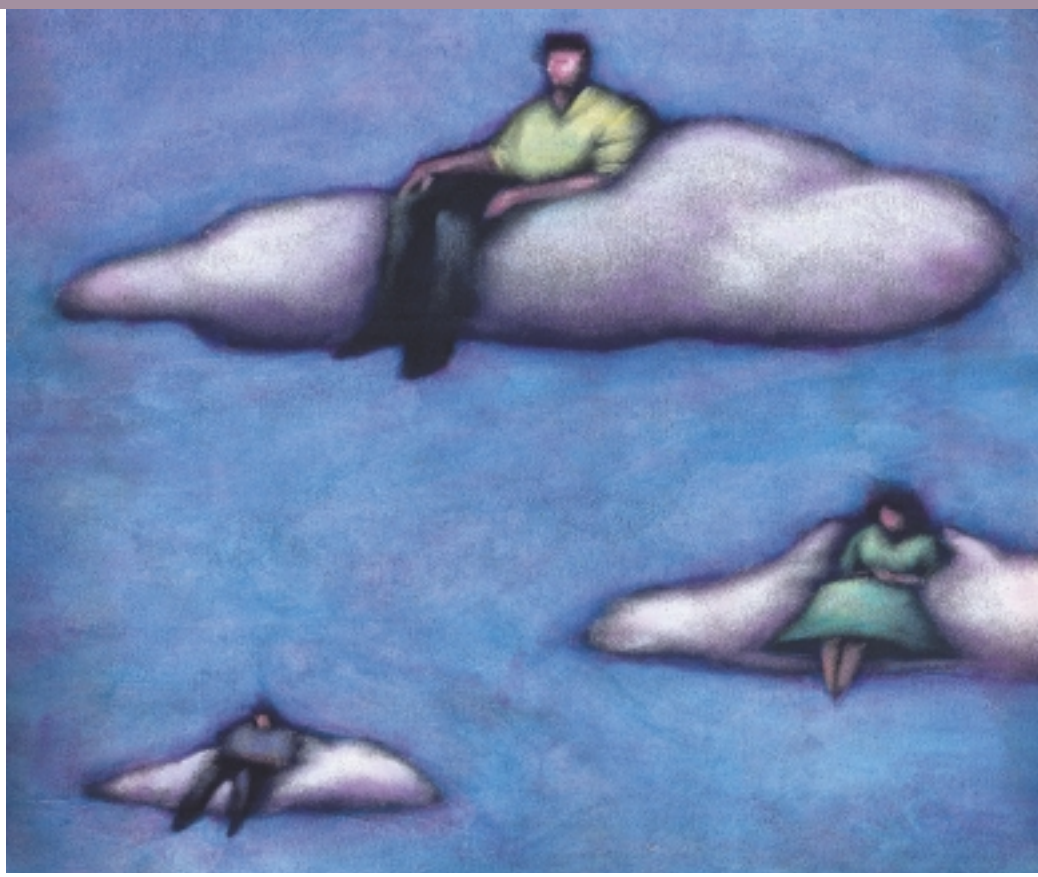
Sandra K. Bauer
Public Member, Board of Pharmacy

PART ONE:

Different People, Different Challenges

Five Life Stories:

Joe, Carrie, Judy, Terry and Donald



Californians with developmental disabilities are a diverse group in terms of lifestyles, ages, personalities, support, living conditions, and medical conditions. These five fictional life stories are intended to illustrate some of the challenges and opportunities caregivers and health care practitioners encounter as they work to improve the lives of those in their care.

Joe

Joe is a 55-year-old man with Down syndrome. He is independent, capable of taking care of his personal hygiene needs, and can use the local transportation system. For the last fifteen years, Joe has lived in a community care home with five other men and has attended art classes at a local activity center. He has always taken pride in his artwork and enjoys socializing with his friends at the center. However, he worries about his routine being changed and frequently asks center staff the same questions repeatedly. Several of his pottery pieces have been sold in a local art gallery specializing in art made by persons with developmental disabilities.

Two months ago, Joe's attendance at the activity center decreased. One day, after ten years of riding the same route, he took the wrong bus home from the activity center. When found, he was disoriented and agitated. A few days later he got into a verbal battle with an activity center teacher. His mood is less stable and he seems more anxious lately. These changes are most evident in the evening.

Joe's caregiver states that she now has to assist him in basic daily

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living skills. Joe needs reminders and help to brush his teeth, comb his hair, and get fully dressed. Joe's caregiver decided to take him to the doctor for an evaluation to determine why there have been changes in his behavior and level of functioning. If these problems persist, Joe may be in jeopardy of losing his residential and day program placements.

His current medications include digoxin (Lanoxin®) and furosemide (Lasix®) for his heart function and lorazepam (Ativan®) for anxiety. After seeing his physician, he was prescribed a new medication, risperidone (Risperdal®). A staff member of the care home was surprised that he was a given a medication for severe mental illness (antipsychotic) and worried he might be "overdosed" on such a "strong" medication. This concern was brought to the attention of Joe's pharmacist.

Did you know that...

- Individuals with Down syndrome may begin to lose their ability to reason and adapt by age 50.
- The onset of Alzheimer's disease in persons with Down syndrome may occur at a younger age and may result in a more rapid decline than in the general population. Drug treatment may help slow the rate of decline.
- People with declining mental function adapt poorly to changes in their environment. A treatment goal is to maintain consistency in daily activities and caregivers.
- Anxiety disorders and phobias are common among people with mild and moderate levels of developmental disabilities.
- Heart and thyroid problems are common among persons with Down syndrome.

Medication Issues

- The amount of digoxin in the blood should be checked to help evaluate changes in Joe's mental status. Other laboratory tests such as potassium level and kidney (renal) function may also be helpful in determining whether drug toxicity is present. If so, the dosage may need to be adjusted, and/or other medications may need to be added for treating Joe's heart condition.
- The use of a new family of antipsychotics is increasing for treating Alzheimer's disease. Risperdone (Risperdal®) is used in low doses and may be a good choice for Joe, who is showing signs of Alzheimer's-type dementia.
- Benzodiazepines such as lorazepam (Ativan®) are a family of drugs used for anxiety, insomnia, seizures, and muscle spasms. Any of these medications may cause amnesia, increased anxiety, and confusion, rather than decreasing these symptoms. This is more common in the elderly. Risk of falls also increases in this age group when treated with benzodiazepines, and fractures that result can be a serious health issue. Most uses for benzodiazepines are short-term or only "as needed," although some anxiety disorders may need long-term therapy. It may be reasonable to consider lowering the dose or discontinuing lorazepam in Joe's case.
- Antidepressants (once thought of as only for depression) such as Prozac®, Paxil®, Zoloft®, and Effexor® are useful for treating anxiety disorders. They need to be taken on a regular basis, not when needed ("prn"), as is common with benzodiazepines. It takes several weeks of continuous therapy for beneficial effects to be seen.

What pharmacists do...

- Pharmacies keep a record of all prescription medications dispensed to a patient. This patient profile may also include over-the-counter (nonprescription) medications and herbal or alternative therapies.
- It is important to use *one* pharmacy to maintain *one* complete medication record. Patronizing one pharmacy will allow the pharmacist to keep a complete profile with all prescription medications, over-the-counter drugs, and alternative/herbal therapies being taken by the individual with developmental disabilities. The pharmacist is then able to evaluate the complete drug regimen and check for drug interactions, multiple medications being used for the same condition, appropriate doses, and so forth. This is sometimes referred to as "drug regimen review" or "drug use evaluation."
- California law requires that pharmacists offer to counsel patients on all new prescriptions. The pharmacist should provide directions on how to take the medication, storage instructions, and precautions about potential adverse effects. When appropriate, the pharmacist will provide additional information and advice on the use of unusual dosage forms (suppositories, inhalers, patches) or what to do if a dose of medication is missed. If a prescription is delivered to a person's home, the pharmacist will most likely provide written information. Your pharmacist should also be available by telephone for additional questions about your prescription.

Carrie

Carrie is a 21-year-old woman with epilepsy and asthma. She is able to perform her basic self-care needs but often requires reminding to wash her hair and clean her clothes. She has been reluctant to brush her teeth lately due to bleeding gums. Socialization is very important for Carrie, so she lives in a large apartment complex. A roommate and staff from an agency that assists individuals with developmental disabilities support her throughout the day. With the help of her roommate and the agency worker, Carrie keeps up the apartment, goes grocery shopping, and cooks. She works part-time at a chain restaurant where she sets up the dining room before opening.

Carrie has daily seizures that cause brief loss of consciousness (absence seizures), and she has more severe seizures that involve movement, jerking of arms and legs, and brief loss of consciousness (myoclonic seizures). Due to her myoclonic seizures, Carrie must wear a protective helmet during the day. While wearing a helmet protects her head from harm, she refuses to wear it consistently because it makes her stand out in public.

Carrie is friendly, but can be overly assertive and uncooperative at times. These moments are often followed by a myoclonic seizure. Afterward, Carrie is usually extremely weak and sleepy. She takes several antiseizure (anticonvulsant) medications: phenytoin (Dilantin®), carbamazepine (Tegretol®), and divalproex (Depakote®). There are many days when Carrie is too tired to get out of bed, possibly due to a combination of her medication regimen and seizures at night.

Carrie's asthma is controlled with oral theophylline and inhaled albuterol (Ventolin®). Without help, Carrie cannot properly inhale her medication. Her wheezing is worse when she is emotionally upset or on poor air quality days.

Carrie also takes birth control pills to help regulate her menstrual cycle, to reduce pain from menstrual cramps, and for contraception.

Did you know that...

- Caregivers and family members need education about seizures so they can accurately observe and document seizure characteristics, frequency, possible triggers, etc. This is important information for medical professionals who don't witness seizures.
- Myoclonic seizures occur more frequently in persons with developmental disabilities.
- People with seizures should carry identification such as a MedicAlert bracelet with information on medication usage, allergies, and past adverse reactions to medications.
- Asthma is treated with two types of drugs. "Symptom reliever" medications include albuterol (a beta-agonist) or theophylline. Long-term controllers or asthma "preventer" medications include corticosteroids, cromolyn, or leukotriene modifiers. Some asthma medications are swallowed (tablets and capsules), and others are inhaled directly into the lungs. Mild asthma may only require treatment with a symptom reliever drug used as needed ("prn") and an asthma preventer drug taken on a regular basis, both in the form of inhalers.
- The pharmacist can teach and evaluate proper inhaler technique. This training should be regularly reinforced to ensure that the medication gets to the right spots in the lungs. The use of a device called a "spacer," which holds the drug before it is inhaled, makes sure the full amount of drug reaches the lungs.
- Women with developmental disabilities need the same "routine" health care as nondisabled females. This includes periodic breast exams, pelvic exams, and PAP smears.

Medication Issues:

- A goal of antiseizure (anticonvulsant) therapy is to control seizure activity with the fewest agents possible. Because these drugs have many side effects, it is especially important to avoid adding medications without evaluating whether other drugs can be discontinued.
- Blood tests are common with anticonvulsant drugs to ensure that the proper dose is prescribed and to avoid toxic side effects. They can also aid in assessing whether patients are taking their medications as prescribed.
- An evaluation by a neurologist may be valuable because Carrie's seizures are not well controlled on her current three-drug combination, and she complains of oversedation. Carbamazepine may worsen absence seizures, and discontinuing this drug may be helpful. Taking more doses toward the evening or at bedtime may decrease daytime drowsiness.
- Regarding Carrie's bleeding gums, phenytoin can cause a condition of gum overgrowth called "gingival hyperplasia" which can contribute to dental cavities, bad breath and chronic gum infection. A good dental hygiene plan is needed to minimize this condition.

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Judy

Judy is a thin 40-year-old woman with cerebral palsy who cannot walk or perform daily basic care needs. She lives in a six-bed home that specializes in caring for people with nursing needs.

Judy requires transportation assistance to visit friends at a local adult activity center for people with developmental disabilities. She enjoys going shopping at the mall with activity leaders and eating at McDonald's. Her favorite activity is relaxing in the water at the community pool. Judy uses a reclining wheelchair that keeps her positioned properly for activities and must be repositioned by a caregiver throughout the day to prevent pressure sores.

The medications she takes are lorazepam (Ativan®) to relax her muscles; cisapride (Propulsid®) to assist the movement of food through her digestive system; docusate (Colace®), a stool softener; bisacodyl (Dulcolax®), a laxative; calcium carbonate, a dietary supplement to prevent osteoporosis; cotrimazole (Bactrim®) to prevent urinary tract infections; and oxybutynin (Ditropan®) for bladder spasms.

> Judy's favorite activity is relaxing in the water at the community pool.

Did you know that…

- People with cerebral palsy or Down syndrome may experience difficulty with eating and swallowing due to a loss of control of their throat muscles. Individuals who experience frequent muscle spasms can be at high risk for choking and inhaling food from the mouth into the lungs (aspiration).
- Gastroesophageal reflux disease (GERD) is common in people with developmental disabilities. Symptoms include heartburn and pain or ulcers when stomach fluid leaks back into the esophagus. This can result in serious complications, such as aspiration pneumonia.
- Decreased movement, not drinking enough fluid, or not eating enough fiber make people with cerebral palsy especially prone to severe constipation. Ongoing bowel and bladder problems will intensify with age. Help with a daily individualized exercise routine and use of a bowel regimen (a stool softener and a stimulant laxative such as one with bisacodyl or senna) is essential.
- Nutritional deficits, hormonal or bone abnormalities, limited muscle activity, and medication usage place people with cerebral palsy or other developmental disabilities at risk for developing osteoporosis.
- Muscle relaxants, including benzodiazepines like lorazepam (Ativan®) are used to help relieve rigidity and muscle spasms in people with cerebral palsy.

Medication Issues

- Cisapride (Propulsid®) can cause life-threatening heart rhythm disturbances (arrhythmias) when taken with certain foods and medications. It should not be used with grapefruit juice, antifungals, erythromycin, clarithromycin (Biaxin®), cimetidine (Tagamet®), some antidepressants, and some drugs for heart rhythm problems (antiarrhythmics). A pharmacist can provide a complete list of drug interactions. {Author's Note: As of press date, the manufacturer has announced that it will initiate a limited-access proram and will voluntarily stop marketing Propulsid® in the U.S. by July 2000. Your pharmacist can suggest an alternative medication if necesary.}
- Judy may need to be evaluated for GERD. Drugs that block the release of acid into the stomach–the proton pump inhibitors Prilosec®, Prevacid®, Aciphex®–are the most effective treatment for GERD. It also may be beneficial to minimize the use of coffee, alcohol, fats, chocolate, nicotine, and drugs that slow intestinal movement such as oxybutynin (Ditropan®).
- Judy may benefit from a bone mineral density test to evaluate the effectiveness of her calcium supplementation. This measure of osteoporosis is sometimes available in pharmacies as well as in doctor's offices.
- It is important to have adequate fluid intake to minimize constipation and allow the safe elimination of Bactrim® by the kidneys. Bactrim® can cause sensitivity to ultraviolet light, so sunscreens and protective clothing should be worn when outdoors.

Terry

Terry is an 8-year-old boy in second grade. He has epilepsy and mild mental retardation. He has recently been diagnosed as having an attention deficit hyperactivity disorder (ADHD). His teachers report that he has difficulty completing assignments and is disruptive during class. He is easily distracted and several times has run into the street. In order for Terry to stay in regular education, he has been assigned a tutor to assist him in class. Terry is an active boy who enjoys playing with his friends and going to school, but he has trouble waiting his turn or engaging in leisure activities quietly.

Terry lives with his single mother who takes full custodial care of him and participates in all areas of his life. She coordinates all of his medical appointments, and attends school meetings and program planning meetings with his caseworker.

Terry has started methylphenidate (Ritalin®) for ADHD. He is also on phenytoin (Dilantin®) and phenobarbital for his seizures and must be careful about the kind of stimulation he receives. Flashing lights or too much excitement has caused Terry to have prolonged seizures that required emergency hospitalization.

Because of Terry's special challenges, it has been difficult for his mother to find a day care provider who will watch him before and after school. To solve this problem, she quit her job and opened her own licensed in-home day care for school-age children. Terry's mother wonders if there is any other medication that might be helpful, as life is very stressful for her and Terry with his current behaviors.

> ADHD is the most common children's psychiatric disorder and is more common in boys than girls.

Did you know that...

- ADHD is the most common children's psychiatric disorder and is more common in boys than girls. It occurs in 3-5% of school-age children nationwide and is divided into two subtypes: mainly very active (hyperactive-impulsive) or mainly distracted (inattentive). Terry appears to have the hyperactive subtype.
- Occasionally other conditions coexist with ADHD, such as Tourette's syndrome (a condition characterized by uncontrollable verbal outbursts or tics) or other behavioral and developmental disorders.
- Treatment for ADHD includes family therapy, parent education and support, behavioral therapy, and drug therapy. Pharmacists can help to dispel the fears about medication harming the child. Recent studies show that children with ADHD who are treated with stimulants are far less likely to abuse drugs later in life than children with ADHD who are untreated.
- Children may outgrow ADHD or it may continue into adulthood.
- Dietary factors probably do not play a role in ADHD.

Medication Issues

- Phenobarbital can contribute to and even be the cause of behavioral disorders such as overactivity and irritability. Another anticonvulsant that will not aggravate ADHD may be a better choice for Terry. If, after evaluation, it is determined that phenobarbital plays a role in Terry's behavior, the dose should be slowly reduced and finally discontinued. Terry should be observed without phenobarbital to see if his behavior improves. His response may affect his diagnosis of ADHD and subsequent treatment.
- Stimulants are the drugs most often used to treat ADHD. They work by decreasing movement and improving coordination. Learning is increased due to an increase in efficiency and attentiveness. Insomnia and loss of appetite (anorexia) are common side effects. There are several stimulants available. Adderall® is a sustained-release form of methylphenidate that has longer lasting effects. This may be an advantage for school-age children in that they may not need to take a dose while at school. Pemoline (Cylert®) is no longer considered a good first choice for ADHD, due to the risk of liver toxicity. If pemoline is used, a consent form should be signed before starting therapy, and blood tests to determine liver function should be done every two weeks.
- Some agents other than stimulants may be useful in ADHD. For example, clonidine (Catapres®) has been used to calm aggressive, impulsive or extremely overactive children.

Donald

Donald is a 30-year-old man with significant cognitive impairment who lives in a 60-bed skilled nursing facility. He communicates with gestures, some sign language, and a few words. At regular times during the day he will loudly bang on the table, wall or cupboard doors. He also shoves people without warning. At times his movements are jerky and abrupt, and occasionally he cries out and makes sharp, guttural sounds. Donald's banging on the table and pushing of others have occurred since childhood. He has a diagnosis of Tourette's syndrome, but caregivers feel that some of his behavior may be attributed to pain. He takes ibuprofen (Motrin) twice a day for this "pain or arthritis-like condition."

Lately, he has been observed to shrink away, as if from visions that other people do not see. When asked what is wrong, Donald can't make clear what he feels or sees. He says "bad" and has scratched his arms severely. Shrinking from people and harming himself are new behaviors for him.

After an evaluation that included a functional assessment to rule out environmental factors, it was determined that Donald is now psychotic. The psychiatrist recommended that Donald receive an antipsychotic medication, but his family insists that he also take herbal medications that contain ingredients such as ma huang, gingko biloba, and ginseng. His medications now include risperidone (Risperdal®), clonidine (Catapres®) for Tourette's syndrome, hydroxyzine (Atarax®) for sedation, levothyroxine (Synthroid®) as a thyroid supplement, and the herbal therapies mentioned above. He also takes pravastatin (Pravacol®) for high cholesterol and hydrocholorothiazide (Esidrix®) for hypertension.

> When asked what is wrong, Donald can't make clear what he feels or sees.

Did you know that...

- Persons with developmental disabilities are vulnerable to all psychiatric disorders, just like the general population.
- About 20% to 30% of individuals with developmental disabilities living in the community and in residential settings have a psychiatric disorder. People with developmental disabilities residing in developmental centers may have higher rates. Anxiety disorders and depression are the most frequent conditions, but more severe mental disorders (psychoses) can also occur. Common symptoms that lead to a referral for psychiatric services are aggression, self-injury or self-stimulation.
- The recognition and management of psychiatric disorders in persons with developmental disabilities who use little or no spoken communication is possible. However, in such cases, it takes more time, patience, and observation to gather clues about what symptoms mean. Environmental, medical, or drug-induced causes all need to be ruled out.
- It is a myth that psychotropic drug therapy is needed for all behavioral problems. Psychotropic medications will decrease only those behavioral problems directly associated with psychiatric illness. Some behaviors that may not respond to drug therapy include undressing, not following instructions, cursing, running away, and not being able to perform tasks.
- When medication is used, it should be part of a treatment plan that is coordinated among caregivers and includes multiple approaches to address behavioral problems. Specific "target symptoms" need to be identified and monitored in the nursing care plan.
- Educating the family about the treatment plan and gaining the support of involved members is an important part of a successful medication treatment plan.
- Reducing high cholesterol and high blood pressure lowers the risk of stroke, heart attack, and death from heart disease.

Medication Issues

- People with developmental disabilities are more sensitive to side effects of psychotropic drugs. It is best to start with low doses and increase, decrease, or discontinue the drug slowly.
- The new antipsychotics such as risperidone (Risperdal®) are increasingly being used as the first choice for treating psychosis. The risk of side effects, especially movement disorders (dystonia, Parkinsonism, akathisia or tardive dyskinesia) are much lower.
- Hypothyroidism can cause changes in mental stability. A blood test to determine Donald's thyroid function is needed to monitor the use of levothyroxine as replacement therapy.
- Haloperidol (Haldol®) and pimozide (Orap®) have traditionally been used to treat Tourette's syndrome. Orap® has new warnings about causing irregular heart rhythm and should probably not be used except as a last resort. Clonidine (Catapres®), guanfacine (Tenex®), antidepressants, and nicotine gum are also used.
- Antihistamines such as hydroxyzine (Vistaril®, Atarax®) or diphenhydramine (Benadryl®) are not useful for behavioral problems. People develop resistance to the general sedative effect, but side effects such as dry mouth, blurry vision, constipation, and trouble urinating continue to be problems. Donald's hydroxyzine should probably be slowly discontinued.

- Herbal medications are not always harmless and need to be assessed by a health professional because they can cause, as well as treat, medical and psychiatric conditions. Products that contain more than one herbal ingredient should be avoided. Pharmacists need to know about alternative therapies in order to evaluate their use for an individual's medical and psychiatric conditions and with their medication therapies. Some herbal products can interact with warfarin and digoxin, or affect blood pressure, mood, or electrolytes such as potassium.
- There is no such thing as generic equivalence with herbals, and potency of "active" ingredients varies widely among products, or even batches of the same product.
- Ma huang contains ephedrine and in high doses can cause toxicity in the central nervous system. Some studies show gingko may have a role in slowing Alzheimer's disease by stimulating and protecting nerve cells and improving blood flow to the brain. Even though these effects appear promising, very little research is available. Gingko can also cause stomach or intestinal (GI) complaints, headache, dizziness, or restlessness. Ginseng can affect blood pressure and heart rate and cause insomnia and anxiety. There are many types of ginseng; composition and effects vary widely.
- Chronic Motrin® or other nonsteroidal anti-inflammatory (NSAID) therapy has risks of gastrointestinal (GI) bleeding and ulcers. Drug therapy to prevent gastrointestinal bleeding and ulcers is recommended for people taking high doses of NSAIDs, those who have a prior history of ulcers or GI bleeding, or individuals taking steroids or anticoagulants. Drugs such as omeprazole (Prilosec®) or misoprostol (Cytotec®) may be used for this preventive therapy.
- Donald will need periodic blood tests to evaluate cholesterol levels, triglyceride levels, and liver function. These tests are necessary with his pravastatin (Pravacol®) therapy.

> With the help of her roommate and the agency worker, Carrie keeps up the apartment, goes grocery shopping, and cooks.

Carrie

Continued from page 9

- Many anticonvulsants (phenytoin, carbamazepine, and phenobarbital) can decrease the effectiveness of birth control pills (oral contraceptives). Breakthrough vaginal bleeding may signal the need for a stronger estrogen pill, but estrogens cause seizures in some women, and high doses may increase side effects. The effectiveness of medroxyprogesterone (Depo-Provera®) doesn't seem to be reduced when used with anticonvulsants and may be a good birth control choice for Carrie.
- Theophylline blood level measurements are needed to help ensure effectiveness and prevent toxicity. Theophylline, which Carrie takes for her asthma, can increase the frequency of seizures by altering brain chemistry. Many drugs interact with theophylline to change its blood level unpredictably. Blood levels go up when theophylline is combined with cimetidine, erythromycin, ciprofloxacin, and other medications. Levels go down with phenytoin, phenobarbital, nicotine (smoking) and other drugs. Carbamazepine can raise or lower theophylline levels. A pharmacist's review can help prevent adverse drug interactions.
- Carrie may not need theophylline if her inhaler technique can be improved. A goal would be to control her asthma by use of an inhaled "reliever" medication as needed and an inhaled "preventer" medication daily. This would minimize drug side effects, toxicities, and interactions.

Summary

People with developmental disabilities constitute a unique and vulnerable population that deserves special attention from pharmacists. A pharmacist may be the best person to review the appropriateness of a medication regimen and to serve as the intermediary between other health providers, the developmentally disabled person, and the caregiver. This means that sometimes the pharmacist will need to intervene on behalf of a patient with well meaning but uninformed family members or health professionals.

Pharmacists may need to develop "developmental disability sensitivity" and be on the lookout for lack of treatment for medical problems, poor preventive medical and dental care, general abuse and neglect, overtreatment with medication to control behavior, and the misconception that those with developmental disabilities don't feel pain or are incapable of having a mental illness.

It is important to assess and treat each person individually because the response to drug therapy that is usually observed may not apply to people with developmental disabilities. The pharmacist must understand the special circumstances of this group of people and rise to the challenges of communication, education, and rational drug use. Pharmacists should be alert for polypharmacy; inappropriate drug selection, dosing, or treatment length; drug interactions; adverse drug reactions; poor compliance; etc., and should not hesitate to act as advocates for their patients. Monitoring for drug regimen efficacy means discontinuing harmful or unneeded drug therapies and suggesting new or more appropriate therapies. Both strategies can make big improvements in a person's quality of life. Practicing pharmaceutical care can be both professionally and personally rewarding! ✦

PART TWO:

Information for the Caregiver

How to Get the Most From Your Pharmacist and Pharmacy

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The pharmacist can be an excellent resource for the caregiver, and a close relationship should be established between them at the earliest opportunity. Pharmacists have been educated in all aspects of drug therapy and are uniquely qualified to address all issues regarding the use of drugs and their administration. Specifically, the pharmacist can provide the caregiver with information on side effects, drug interactions, appropriate timing and dosing of medications, and what to expect from therapy. In order to maximize the pharmacist-caregiver-consumer relationship, open communication should be maintained at all times. Understanding the pharmacist's role will aid the caregiver in using this resource to its fullest effect.

When a person with developmental disabilities or a caregiver first seeks the advice of a pharmacist, the pharmacist will need to ask several questions and take a complete drug history of the person with the developmental disability. A drug history provides the pharmacist with a wealth of information. Often, the pharmacist is called upon to recommend drug therapy for a particular condition to the individual's physician. Knowing what medications the person is taking, as well as the response to medications taken in the past, will help guide the pharmacist in making a recommendation. The pharmacist needs to know about all medications the person with developmental disabilities takes including prescription, over-the-counter, herbal, and alternative therapies. A complete list of the consumer's current therapy will allow the pharmacist to look for drug interactions and duplications. One of the pharmacist's most important contributions is maximizing the effectiveness of drug therapy while minimizing the number of drugs taken and the number of side effects.

When taking the drug history, the pharmacist will ask about previous allergies to medications. It is important for the pharmacist to know what drug caused the reaction, how long ago it happened, and the nature of the reaction (rash vs. shortness of breath, etc.). When assessing drug allergies, the pharmacist and caregiver need to distinguish a true drug allergy from a side effect. For example, many people say that they are allergic to codeine because it gives them an upset stomach. An upset stomach is a very common side effect of codeine, but is not an allergy, even though it may provide a reason not to use the drug again. By contrast, a person who develops hives while taking penicillin would be considered to have a true drug allergy. The pharmacist uses his or her knowledge of a person's drug allergy history to prevent future occurrences.

There is some basic medical information the pharmacist needs to

know that the caregiver can provide. The consumer's age, height and weight will help the pharmacist verify that the drugs are being given at the correct dose. The pharmacist will want an overview of the individual's medical history, such as diseases or conditions for which the person has been treated, a list of surgeries or procedures that the individual has undergone, and a complete list of all the physicians responsible for the person's care. The pharmacist may ask the consumer's legal guardian or conservator to sign a waiver that will allow the pharmacist access to the consumer's medical records. This allows the pharmacist and physician to freely share information about the consumer and work as a team in coordinating care.

Once the pharmacist-consumer-caregiver relationship has been established, ongoing follow-up should occur at intervals that best meet the consumer's and the caregiver's needs. Whenever the caregiver presents a new prescription to the pharmacist, there is an excellent opportunity for review. At this time the caregiver will also learn about the new medication being prescribed: the name of the medication, what it is used for, how to properly take the medication, common side effects, and how to properly store the medication. Most pharmacies can provide all this information in written form for review at home. If anything is unclear, it is crucial for the caregiver to ask the pharmacist for clarification as soon as possible.

There are some side effects and problems with drug therapy that a pharmacist can help minimize by counseling the consumer or by recommending an over-the-counter remedy. Other problems need to be referred to a physician, and the pharmacist should be prepared to make this referral. For example, if an individual develops shortness of breath after taking one dose of a new antibiotic, the pharmacist should immediately refer the person and the caregiver to an emergency room. Alternatively, if the individual begins complaining of an upset stomach when taking the same newly prescribed antibiotic, the pharmacist may simply recommend the antibiotic be taken with food rather than on an empty stomach. Continued discussion between the caregiver and the pharmacist will ensure that new medications are effectively used and tolerated by the consumer. The individual's primary caregiver is responsible to ensure that all of the individual's needs, including drug therapy, are met and that the individual's family is informed of issues relating to the person's care. The pharmacist should be able to freely communicate directly with the family as well as the caregiver. Everyone involved in the health and welfare of the individual should keep in frequent contact to ensure that the person receives the highest level of care.

Assisting with Self-Medication

In addition to being careful with what medications a person takes, it is also important that the medication is, in fact, taken. Many factors are involved in giving medication correctly. The caregiver should be knowledgable about how best to give each drug the person is taking. Likewise, the caregiver should take steps to ensure that dosing errors are minimized. An informed and conscientious caregiver is the individual's best protection against potentially serious problems with medication use.

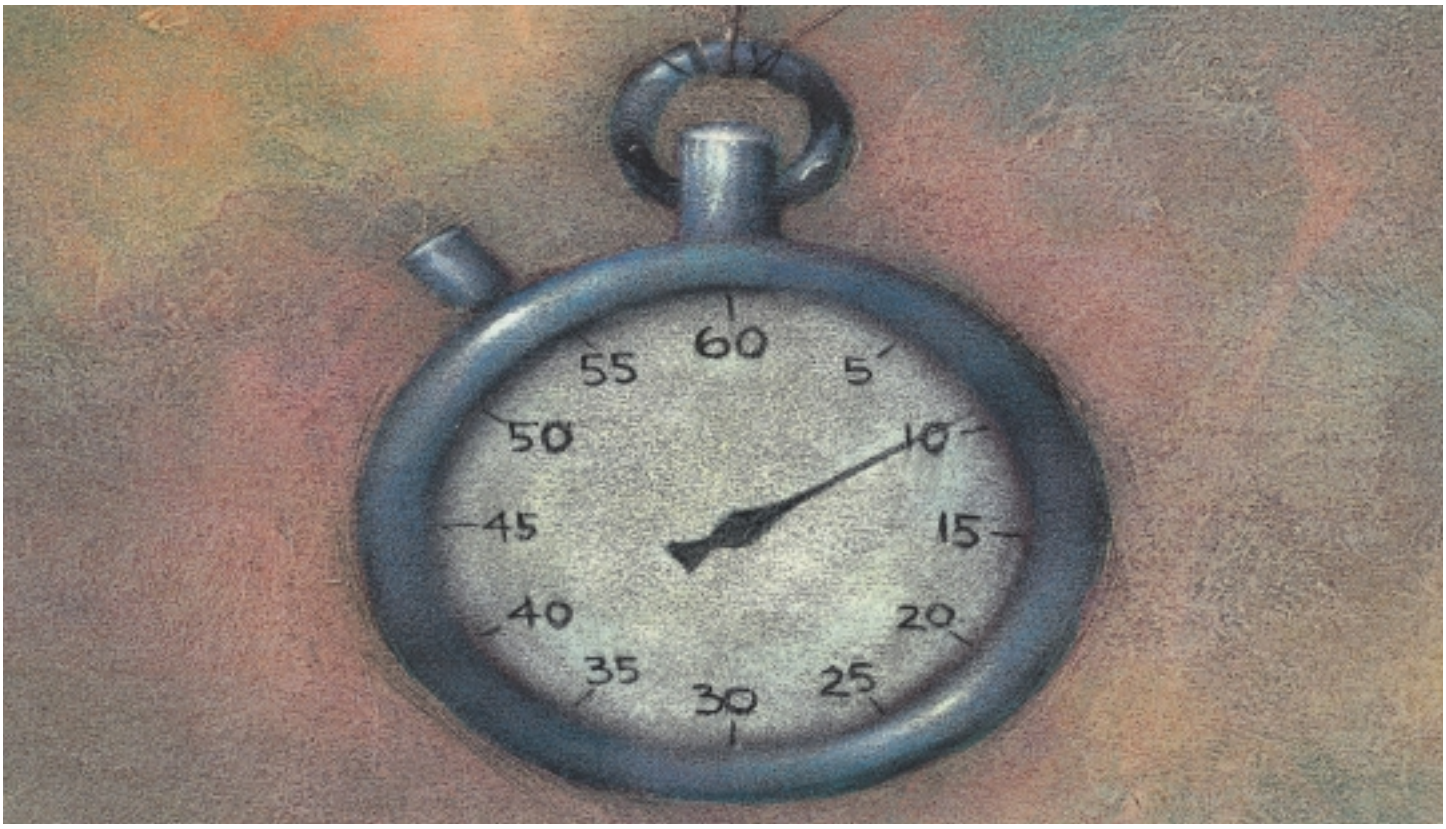
The caregiver should become familiar with the information on each prescription label. By law, the prescription label must contain the following information: the consumer's name, prescription number, date the medication was filled, prescribing physician, name and strength of the medication, directions for use, quantity contained in the package, number of refills remaining, and expiration date of the medication. The label will also contain the pharmacist's initials, and the name, address, and phone number of the dispensing pharmacy. The caregiver should be able to readily identify the name of the drug and the directions for use, as well as the prescription number used to call for refills. If any portion of the label is unclear, the caregiver should get an explanation from the pharmacist.

Several types of containers are used to hold medications. The most common is a vial to which a label as described above is attached. Medications may also come packaged in a "unit dose card" with each dose contained in a plastic bubble for the caregiver to punch out at the appropriate time. The advantage of these bubbles is that the caregiver can note the time the dose was given right on the card next to the empty bubble. This aids in recordkeeping within the home. Medications may sometimes be supplied in "medisets," trays organized by the time of day and by the day of the week. All doses to be taken at a particular time are put in one slot of the mediset. Most medisets can hold up to one week's worth of medication and can be filled either by the caregiver or the pharmacist. Medisets must include the labels of all prescriptions contained in them or identify the contents in some other manner. Licensed caregivers should contact their licensing agencies for rules and restrictions regarding the use of medication containers.

TABLE 1

Medication Administration Record Example

MEDICATION	TIMES	DAYS OF THE MONTH																														NAME OF THE MONTH _____
		1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18	19	20	21	22	23	24	25	26	27	28	29	30	31
Name, dose:	8 am																															
Frequency:	12 pm																															
Directions:	4 pm																															
	8 pm																															



Medications often need to be taken at specific times or under specific conditions to maximize their effectiveness. The caregiver should become familiar with proper administration technique for each medication that the consumer takes. If the label itself does not provide this information, a pharmacist should be consulted. Some general rules of thumb can be followed: if the directions read to take the medication twice daily, then it should be taken every 12 hours, or as close to that time frame as is reasonably possible. If the directions read to take the medication three times daily, then the medication should be taken every 8 hours, or as close to that time frame as possible (not necessarily at breakfast, lunch, and dinner, as many people assume). If a medication is to be taken once daily, it is best to try to take it at the same time each day. Certain medications are best taken at a specific time during the day. For example, certain drugs used to lower cholesterol are best taken with the evening meal.

The caregiver should be certain to find out what effect food has on the drugs taken by the individual with developmental disabilities. Food can sometimes alter the absorption of a drug, minimize a drug's effectiveness, enhance its effectiveness, or merely be recommended to decrease stomach upset. If a medication is to be taken on an empty stomach, this means either one hour before or two hours after a meal. Most prescriptions that need to be taken with or without food will have an extra label attached to the vial. Typically, plenty of water means eight ounces (an average full glass). Once again, the pharmacist should instruct the caregiver on the appropriateness of fluids with respect to medication.

Caregivers are often faced with the dilemma of individuals who will not take their medication. This difficult situation requires patience on the part of the caregiver, who should do his or her best to ensure that each dose of medication is taken. It is important, however, to keep in mind that the consumer does have a right to refuse to take a medication.

If the person with developmental disabilities has difficulty swallowing, ask the pharmacist if the medication is available in liquid form. Certain pharmacies may be able to prepare such a liquid if one is not commercially available. Another alternative may be to crush the medication and sprinkle it on a food that the consumer will readily eat. A pharmacist should be consulted prior to this practice, as some medications should not be crushed. The consumer or the consumer's representative, as well as the physician, need to give their permission in writing for a medication to be crushed. Regardless of the individual's problems with medication administration, the caregiver should make every effort to ensure that medications are taken consistently. The pharmacist can be an excellent resource in assisting the caregiver with this challenge.

Drug errors have the potential to result in tragic outcomes. Many drug errors are easily preventable; therefore caregivers should take measures to ensure that risks associated with medication administration are minimized to the extent possible. Caregivers should thoroughly familiarize themselves with the colors, sizes, shapes and names of all medications their consumers routinely take. If a refill arrives from the pharmacy that looks different from what was expected, an inquiry should be made. Many drugs have generic counterparts that look different from the brand name product and from each other. Dispensing errors do occasionally occur, but a simple telephone call can usually resolve any discrepancy before the individual takes a dose of the new medication.

When an individual with developmental disabilities resides in a Community Care Facility (CCF) or Intermediate Care Facility (ICF), medications cannot be self-administered unless the physician, the nurse, and the caring staff agree that it is safe to do so. Before ICF staff members can administer medications, they must pass a training course in medications and their administration and it is recommended that CCF staff receive similar training.

Five basic rules - the "Five Rights" - apply when giving any medication. Before giving any medication, ask yourself:

- Is this the RIGHT individual to receive this medication?
- Is it the RIGHT drug?
- Is it the RIGHT dose?
- Is it the RIGHT time?
- Is this the RIGHT route of administration?

In healthcare facilities, safeguards are in place to ensure that the right medications are administered. Typically a Medication Administration Record (MAR) is created to provide a place to document that the five RIGHTS were carried out. Table 1 is an example of a an MAR that shows the drugs to be given and the times of administration on the left-hand side of the form, with the days of the month across the top. When the medications are administered, the caregiver records who gave what, when, how many, and to whom in the appropriate square on the form.

A caregiver in the home should develop a similar system for documenting when doses for medications are given. This is especially important if an individual is on multiple medications or the caregiver is responsible for the care of more than one person. Documentation should occur immediately after a dose is given, and the medication administration record should be readily accessible at all times. If the caregiver administers medications to more than one person, he or she should take a few seconds prior to giving any medication to verify that the drug in hand is in fact for that particular individual. Medications should be kept in a well-lighted area and in a locked cabinet to prevent access to them by individuals with developmental disabilities and/or other people in the home. Refill requests should be made well in advance to prevent missed doses due to inadequate supply.

Medication administration requires special attention since it is such an important aspect of an individual's care. With planning and organization, many of the common problems associated with administering drugs can be minimized. As always, the pharmacist is an excellent resource in guiding the caregiver in the appropriate steps to take in a particular home environment. ✦

Library for Caregivers

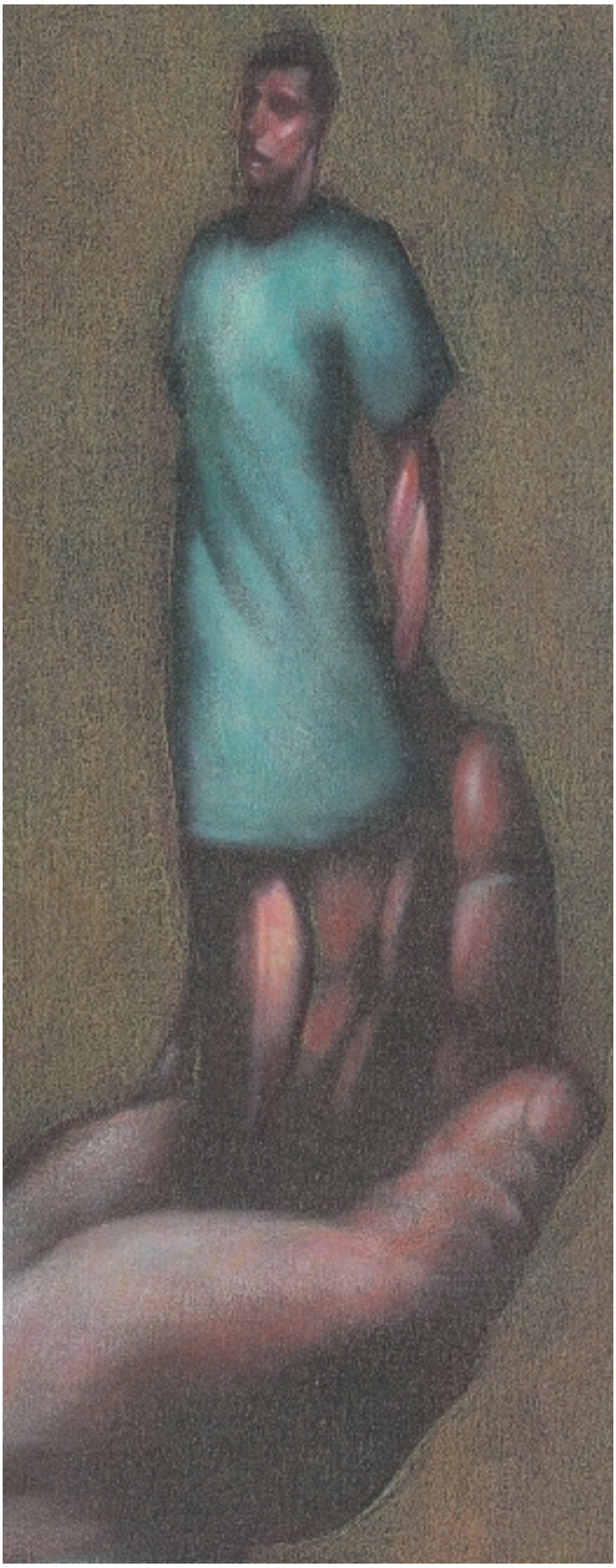
Drug Information for the Allied Health Care Professional: With Indication Therapeutic Category Index: 1998-99, ISBN 0916589633

The PDR Family Guide to Prescription Drugs, 6th Edition, ISBN 0609803565

1999 Physician's Desk Reference, ISBN 1563632888

Advice For the Patient: Drug Information in Lay Language, 19th Edition, Volume 2, ISBN 156363323X

1999 PDR For Non Prescription Drugs and Dietary Supplements, ISBN 1563632985



PART THREE:

Information and Resources for the Caregiver, the Pharmacist, and Other Health Care Professionals

"People First" Language

Like many aspects of health care, there are terms often applied to people with developmental disabilities that they, their families or their caregivers may find offensive. Choosing appropriate words is important when pharmacists counsel persons with developmental disabilities or their caregivers. Being sensitive to terminology will enhance the patient consultation experience.

Language is a reflection of how we see each other, and sometimes the words we use can hurt. It is best to think, "People first," and now responsible communicators are choosing language that recognizes the dignity of people with disabilities...words that put the person first, rather than the disability.

Examples of what to say...

- "People with disabilities" … Avoid "handicapped" or "the disabled."
- "People with mental retardation" or "has cognitive impairment" … Avoid "the mentally retarded" or "he's/she's retarded."
- "My son has autism" … Avoid "my son is autistic."
- "She has Down syndrome" … Avoid "she is a Down syndrome."
- "A congenital disability" … Avoid "a birth defect."
- "Uses a wheelchair" or is "a wheelchair user" … Avoid "wheelchair bound" or "confined to a wheelchair."
- "She has a developmental disability" … Avoid "she is developmentally delayed" or "he is developmentally disabled."
- "Orthopedic disability" … Avoid "he's crippled" or "lame."
- "She is with short stature" … Avoid "she's a dwarf" or "a midget."
- "He is nonverbal" … Avoid "mute."
- "Has a learning disability" … Avoid "learning disabled."
- "Accessible parking" … Avoid "handicapped parking."
- "Person who has" … Avoid "afflicted with," "suffers from," "a victim of…"
- "She has an emotional disability" … Avoid "she's emotionally disturbed" or "crazy."
- "Typical" or "nondisabled" … Avoid "normal" and "healthy."
- "He has quadriplegia" or "paraplegia" … Avoid "he's a quadriplegic" or "paraplegic."

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The California Service Delivery System for Individuals With Developmental Disabilities

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Overview

The Department of Developmental Services (DDS) is one of thirteen departments composing the California Health and Human Services Agency in the Governor's administration. DDS is responsible, under the California Lanterman Developmental Disabilities Services Act, for designing and coordinating services for California residents with developmental disabilities. The Lanterman Developmental Disabilities Services Act defines the rights of persons with developmental disabilities, ensures that eligible individuals will receive appropriate services, and establishes how those services will be delivered.

The state of California provides criteria to qualify for developmental disability services. To be eligible for services, the disability must begin before the individual's 18th birthday, be expected to continue indefinitely, and present a significant disability. The disability must be due to one of the following conditions: mental retardation, cerebral palsy, epilepsy, autism, or be a disabling condition closely related to mental retardation that requires similar treatment. Infants and toddlers (age 0 to 36 months) who are at risk of a developmental disability may be eligible for referral for regional center services. In addition, parents who are at risk of having a child with a developmental disability may also be eligible for genetic diagnosis, counseling, and other prevention services. In California, developmental disability does not include other disabling conditions that are solely physical in nature.

DDS Mission Statement

The Department of Developmental Services will provide leadership and direction to help ensure that individuals with developmental disabilities have the opportunity to lead more independent, productive, and normal lives as envisioned by the Lanterman Developmental Disabilities Services Act.

DDS Vision Statement

Building Partnerships, Supporting Choices: Vision for a Preferred Future for Persons with Developmental Disabilities

People with developmental disabilities will participate in valued ways with their friends, neighbors, and coworkers in all areas of community life. Services and supports are provided to enable them to have real choices in where to live, work, and socialize. Families are

respected and supported in their role of primary decision-makers on behalf of their minor children. Professionals join in partnership with families, supporting their cultural preferences, values, and lifestyles.

Individuals with developmental disabilities who reside in the state developmental centers receive services appropriate to their needs that allow them to develop and achieve their full potential.

Those who reside in the community are supported in natural settings with opportunities to live in their own homes, to be involved in meaningful activities in integrated settings and to participate in community life.

The service system for persons with developmental disabilities supports rather than controls individuals and families entering into partnerships that promote self-determination and interdependence.

Service Delivery System

Services to individuals with developmental disabilities are provided through 21 regional centers and 5 state developmental centers. Regional centers are nonprofit private corporations that have offices throughout California. They are a local resource to help find and access services for individuals and their families. The state developmental centers are operated directly by DDS and provide services to individuals who have been determined by regional centers to require care, treatment, and supervision in a structured health facility setting on a 24-hour basis.

Rights of Individuals with Developmental Disabilities

Each individual with a developmental disability is entitled to the same rights, protections, and responsibilities as all other persons under the laws and Constitution of the State of California, and under the laws and the Constitution of the United States. These rights include, but are not limited to, the following:

- A right to treatment and habilitation services.
- A right to dignity, privacy, and humane care.
- A right to participate in an appropriate program of publicly supported education, regardless of the degree of handicap.
- A right to religious freedom and practice, including the right to attend services or to refuse attendance.
- A right to prompt and appropriate medical care and treatment.
- A right to social interaction and participation in community activities.
- A right to physical exercise and recreational opportunities.
- A right to be free from harm, including unnecessary physical restraint, or from isolation, excessive medication, abuse, or neglect. Medication shall not be used as punishment, for convenience of staff, as a substitute for a treatment program, or in quantities that interfere with the treatment program.
- A right to be free from hazardous procedures.
- A right to advocacy services.
- A right to be free from discrimination.
- A right to have access to the courts.

TABLE 1

Living Situations of Californians with Developmental Disabilities

~155,000 people, total population

~100,000	live with family members
~34,000	live in a community care facility, an intermediate care facility or skilled nursing facility
~13,000	receive independent living or supported living services in the community
~4,000	live in developmental centers
~1,000	live in other places, such as foster family homes and county homes

TABLE 2

*Number of Californians with Specific Diagnostic Conditions, July 1999**

Condition	Individuals With Condition Present	
Mental Retardation	110,810	84%
Cerebral Palsy	29,002	22%
Autism	11,233	8%
Epilepsy	31,184	24%
Other Types of Developmental Disabilities	11,411	9%
Visual Problems	17,846	13%
Hearing Problems	6,276	5%
Visual & Hearing Problems	5,828	4%
Ambulatory	103,634	78%
Medical Problems	28,806	22%
Special Aids Needed	32,719	25%
Technology Dependent	5,139	4%
Dual Diagnosis	10,945	8%
Severe Behaviors	9,038	7%
Behavioral Drugs	13,154	10%

*Individuals with multiple conditions may be counted in multiple categories.

Note: Figures represent the number of individuals with a current and active status with the Department of Developmental Services.

CDER - The Client Development Evaluation Report is an instrument used to obtain diagnostic and evaluation information on individuals with developmental disabilities.

Regional Center Locations

Color corresponds to area served by each Regional Center

Courtesy of Associations of Regional Center Agencies (ARCA)

Regional Centers

Regional centers have contracts with DDS to provide or coordinate the following services for individuals with developmental disabilities:

- Information and referral
- Assessment and diagnosis
- Counseling
- Lifelong individualized planning and service coordination
- Purchase of necessary services included in the individual program plan
- Assistance in finding and using community and other resources
- Early intervention services for at-risk infants and their families
- Genetic counseling
- Family support
- Planning, placement, and monitoring for 24-hour out-of-home care
- Training and educational opportunities for individuals and families
- Community education about developmental disabilities

There is no charge for diagnosis and assessment for eligibility. Once eligibility is determined, most services are free, regardless of age or income. There is, however, a requirement for parents to share the cost of 24-hour out-of-home placements for children under age 18 with the amount depending on the parent's ability to pay. A copayment may also be required for other selected services.

Regional centers are required by law to provide services in the most cost-effective way possible. They must use all other resources, including generic resources, before using any regional center funds. A generic resource is a service provided by an agency that has a legal responsibility to provide services to the general public and receives public funds. Such agencies include school districts, county social service departments, Medi-Cal, Social Security, Department of Rehabilitation, etc.

Anyone who uses regional center services has a planning team that includes the individual utilizing services, family members, regional center staff, and anyone else who is asked to be there by the individual. An Individual Program Plan or Individual Family Service Plan (for children age 0 to 36 months) is developed that lists the individual's goals, the services needed, and who will pay for services. A focus on the individual (i.e., Person-Centered Planning) ensures that the identified services and supports meet the needs and preferences of each person with a developmental disability. All services listed in the plan will be provided by a generic resource, a regional center vendor (a business approved by the regional center), or directly by the regional center.

TABLE 3

California Regional Centers Demographic Information*

Regional Center	Total Population
Alta	10,185
Central Valley	9,343
East Bay	9,974
East Los Angeles	5,111
Far Northern	4,313
Golden Gate	5,874
Harbor	7,246
Inland	14,431
Kern	4,373
Lanterman	4,964
North Bay	5,302
North Los Angeles	9,084
Orange	10,269
Redwood Coast	2,113
San Andreas	7,656
San Diego	12,162
San Gabriel/Pomona	7,413
South Central Los Angeles	6,768
Tri-Counties	7,168
Valley Mountain	6,578
Westside	4,817
Totals	155,144

* Source: DDS Client File, October 6, 1998

Residential Services

Person-Centered Planning is used in making decisions regarding where a person with developmental disabilities will live and the kinds of services and supports that may be needed. The following is a list of living arrangements and supportive services available for persons with developmental disabilities:

- Supported Living Services – where an individual lives in an apartment or home and receives support from a supportive living agency.
- Community Care Facilities – typically a six-bed home operated by a care provider and licensed by the Department of Social Services, Community Care Licensing.
- Family Home Agency – where an adult lives with a family that is monitored through a family home agency.
- Foster Family Agency – where a child lives in a family home monitored through a foster family agency.
- Independent Living – where an individual lives alone and receives training on specific daily living skills.
- Intermediate Care Facilities – typically a six-bed home, licensed by the Department of Health Services, that specializes in caring for individuals with medical and self-care needs.

Personal Rights for Individuals in Care Facilities or Developmental Centers

Individuals with developmental disabilities who have been admitted or committed to state developmental centers, community care facilities, or health care facilities shall have rights which include, but are not limited to, the following:

- To keep and be allowed to spend their own money for personal and incidental needs.
- To keep and wear their own clothing.
- To keep and use their own personal possessions.
- To have access to individual storage space for their private use.
- To see visitors each day.
- To have reasonable access to telephones, both to make and receive confidential calls, and to have calls made for them upon request.
- To mail and receive unopened correspondence and to have ready access to letter-writing materials, including stamps.
- To refuse electroconvulsive therapy (ECT).
- To refuse behavior modification techniques that cause pain or trauma.
- To refuse psychosurgery.

Day Program Services

Day programs are community-based programs for individuals served by a regional center. They are available when those services are included in a person's Individual Program Plan developed with the Person-Centered Planning process. Day program services may be at a fixed location or out in the community. Types of services available through a day program include:

- Developing and maintaining self-help and self-care skills.
- Developing the ability to interact with others, making one's needs known, and responding to instructions.
- Developing self-advocacy and employment skills.
- Developing community integration skills, such as accessing community services.
- Developing social and recreational skills.
- Developing behavior management skills.
- Vocational rehabilitation, supported employment, and work activity training (programs administered by the California Department of Rehabilitation).

Foster Grandparent and Senior Companion Program

The Foster Grandparent Program started in 1967 at Lanterman Developmental Center and is now in operation at all developmental centers, Napa State Hospital, and in the community. The program establishes a relationship between elderly persons in the community and children with developmental disabilities. Foster grandparents bring a lifetime of experience to their children and provide unconditional love and consistency, while establishing a sense of trust.

In 1973, the Senior Companion Program was established. This program shares the same philosophy and goals as the Foster Grandparent program, except that it serves individuals over the age of 21. Adults with developmental disabilities derive many benefits from these one-on-one relationships. Currently, 381 Foster Grandparents and 287 Senior Companions are working throughout the state.

Respite Services

In-Home Respite is intermittent or regularly scheduled temporary nonmedical care and/or supervision provided in the person's home. In-Home Respite services typically include:

- Assisting the family members to enable a person with developmental disabilities to stay at home.
- Providing appropriate care and supervision to protect that person's safety in the absence of a family member(s).
- Relieving family members from the constant responsibility of providing care.
- Attending to basic self-help needs and other activities that would ordinarily be performed by the family member.

Prevention Programs

In recognition of the fact that up to fifty percent of mental retardation, birth defects, and other developmental disabilities can be prevented, the Department of Developmental Services has assumed a leadership role in the area of prevention. Prevention programs provide services such as genetic screening, and prenatal and other genetic and diagnostic procedures, including public awareness programs and outreach to parents at high risk of having a child with a developmental disability.

One of the most significant programs in this area is the Early Start Program. The mission of the Early Start Program is "to establish a statewide coordinated interagency system for infants and toddlers with disabilities and infants at high risk, and their families, based on existing resources and innovative approaches to comprehensive, family-focused early intervention services."

The Early Start Program is California's response to federal legislation to ensure that services to infants and toddlers with developmental disabilities and their families are provided in a coordinated, family-friendly manner. The vision for California's program includes family and professional collaboration and support, along with the coordination of services provided by community agencies and service providers.

Wellness Initiative

The Department of Developmental Services launched the Wellness Initiative to help ensure access to quality medical, dental, and mental health services for persons with developmental disabilities through special projects and activities. As a part of this mission,

TABLE 4

Advocacy Organizations and Associations

California Department of Developmental Services
P.O. Box 944202
Sacramento, CA 94244-2020
(916) 654-2205
www.dds.ca.gov

Association of Regional Center Agencies
915 L Street, Ste. 1050
Sacramento, CA 95814
(916) 446-7961
www.arcanet.org

Protection and Advocacy, Inc.
100 Howe Avenue, Ste. 235N
Sacramento, CA 95825
(916) 488-9950
www.pai-ca.org

State Council on Developmental Disabilities
2000 O Street, Ste. 100
Sacramento, CA 95818
(916) 322-8481
www.scdd.ca.gov

ARC California
120 I Street, Second Floor
Sacramento, CA 95814-2213
(916) 552-6619

Organization of Area Boards
3000 S Street, Ste. 210
Sacramento, CA 95816-7055
(916) 227-2148
www.ns.net/OAB

People First of California
1225 8th Street, Ste. 590
Sacramento, CA 95814
(916) 552-6625

DDS has founded the California Wellness Partnership. In conceptualizing the Wellness Initiative, DDS realized that many experts and public leaders in the field of health and wellness could be of great assistance to our mission and that we needed to reach outside our customary borders and forge partnerships with allied organizations. With these partnerships and special projects, DDS is continually working to improve the health care system for individuals with developmental disabilities.

In addition, DDS has created the *Wellness Digest*. Recent publications of this digest have included Dental Health, Infection Prevention and Control, and Medications. To request a free subscription to the *Wellness Digest*, call 1-877-DDS-HEAL (1-877-337-4325) toll free.

The service delivery system for individuals with developmental disabilities in California is unique. No other state has passed legislation similar to the Lanterman Developmental Disabilities Services Act. More information regarding individuals with developmental disabilities, DDS, and California's service delivery system, can be found on the DDS website at *www.dds.ca.gov*.

Physician Assistance

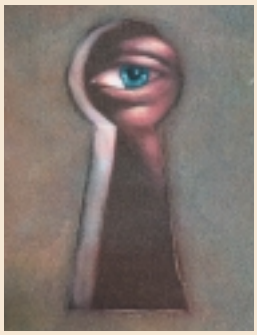
The Physician Assistance, Consultation and Training Network (PACT Net) is now available for physicians who are treating patients with developmental disabilities and complex medical conditions. Specialists at the University of California, Davis are available to provide consultations to community physicians via a toll-free telephone "warm line." Specialists are available in the areas of: developmental pediatrics, gastroenterology, medical genetics, neurology, orthopedics, pharmacology, physical medicine and rehabilitation, psychiatry, and pulmonary medicine. Physicians can obtain a consultation by calling 1-800-4-UC-Davis. At this number they will pick from a menu of options, one of which is the physician consultation (#3 prompt). For more information about the PACT Net program, please contact Andrea Verdon at the M.I.N.D. Institute, UC Davis Health System, at (916) 734-5153 or 888-883-0961 (toll-free).

Medication-Related Legislative Mandates

In 1998, several changes were made to the Welfare and Institutions Code that related to individuals with developmental disabilities. One statute, Senate Bill 1038, added a health status review (with consumer consent) to the Individual Program Plan meeting. The review consists of a medical, dental, and mental health needs assessment of the adult or child. It also includes a discussion of the consumer's current medication regimen, any observed side effects, and the date of the last review of the medications prescribed. If concerns are noted during the review, referrals are to be made to regional center clinicians or to the consumer's physician, as appropriate.

References:

Information in this article was obtained from various Department of Developmental Services' publications and the DDS web page. ✦



Sources of Information for Treating Adults & Children with Developmental Disabilities

The Council on Quality and Leadership in Supports for People with Disabilities (formerly the Accreditation Council for People with Developmental Disabilities — ACDD), *www.thecouncil.org.*

Joint Commission of Accreditation of Healthcare Organizations (JCAHO), *www.JCAHO.org.*

Psychopharmacological Medications - Safety Precautions for Persons with Developmental Disabilities published by the Health Care Financing Administration (HCFA), Health Standards and Quality Bureau, Center for Long Term Care, *www.hcfa.gov.*

National Alliance for the Mentally Ill (NAMI), *www.nami.org.* Develops position papers to assist advocates in efforts to improve the lives of those with mental illness.

American Psychiatric Association (APA), *www.psych.org.* Maintains fact sheets and position papers on diverse topics such as St. John's Wort, children's health, patient protection and managed care, medical records privacy, psychiatry and mental retardation, etc.

National Institutes of Health, *www.nih.gov.*

Psychotropic Medications and Developmental Disabilities: The International Consensus Handbook, edited by Steven Reiss and Michael Aman, Columbus, Ohio: The Ohio State University Nisonger Center, 1998.

Applied Therapeutics: The Clinical Use of Drugs, 6th Edition, Chapters 78 and 79, edited by Lloyd Young and Mary Anne Koda-Kimble, Applied Therapeutics, Inc, Vancouver, WA, 1995.

PART FOUR:

Information for the Pharmacist and Other Health Care Professionals

Common Causes of Mental Retardation

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Within the California Regional Center system, the established diagnosis for eligibility for services and supports include the following five categories: mental retardation, cerebral palsy, epilepsy, autistic disorder, and conditions similar to mental retardation or requiring treatment similar to the needs of an individual with mental retardation ("5th category"). All these conditions must have been established before the age of 18 years. Individuals with the diagnosis of cerebral palsy and epilepsy must meet criteria of substantially disabling in order to be eligible.

The estimated percentage breakout for each established diagnoses is: 57% mental retardation, 14.4% cerebral palsy, 14.8% epilepsy, 3.2% autistic disorder, and 10.3% 5th category.

Mental retardation by definition refers to significantly subaverage general intellectual functioning existing concurrently with deficits in adaptive behavior. The prevalence of mental retardation in any population is 3%, with the majority (80%) being within the mild range of mental retardation. Categories of etiology for mental retardation include: chromosomal (27%), single gene disorders (15%), brain malformation and other idiopathic syndromes (14%), environmental (15%), and unexplained (25+%). Normal intelligence quotients (IQ) are defined within two standard deviations between IQ 70 and IQ 130, with the mean IQ of 100. The ranges of mental retardation start below these two standard deviations and are the following: IQ 70-55 is mild mental retardation, IQ 55-40 is moderate mental retardation, IQ 40-25 is severe mental retardation, and IQ less than 25 is profound mental retardation.

Over the past 10 years, I have reviewed the diagnosable causes of mental retardation at Alta California Regional Center, which is one of the larger of the 21 California Regional Centers, with a current population just over 10,000 consumers. The leading causes of identifiable mental retardation are included in Table 1. Each of these diagnoses is briefly summarized in the following discussion that highlights important clinical information for pharmacists and other health care providers.

TABLE 1

Common Causes of Mental Retardation

- Down Syndrome (Trisomy 21)
- Fetal Alcohol Syndrome
- Fragile-X Syndrome
- Fetal Anticonvulsant Syndrome (Fetal Hydantoin Syndrome)
- Noonan Syndrome
- Tuberous Sclerosis
- Klinefelter Syndrome
- Williams Syndrome
- Prader-Willi Syndrome (H3O)
- Angelman Syndrome

Down Syndrome (Trisomy 21)

Down syndrome is the most common recognizable chromosomal disorder, occurring at a frequency in all populations of 1/800. It is important that each infant suspected to have Down syndrome have a karyotype (chromosome test) to determine the genetic mechanism. Ninety-five percent have an additional chromosome 21 (Trisomy 21), 3 to 4% have a translocated chromosome 21 (21 attached to another chromosome), and 1% have mosaic Down syndrome (some cells with an additional chromosome 21 and some cells with the normal count). This is important, since the recurrence risk for the translocation form of Down syndrome is much higher if one of the parents has a balanced translocation (where one of their chromosome 21 is attached to another chromosome, i.e. chromosome 15). The parent is balanced (has all the genetic material), but can make eggs or sperm that is unbalanced (with an additional chromosome 21 or missing one chromosome 21 [for example, monosomy 21, a nonsurvivable state]).

The clinical features of this condition are very recognizable and include flat midface, protuding tongue, upward slant to the eyes, extreme hypotonia (floppiness), incurved fifth fingers, single palmar creases, and a gap between the great and the second toes.

Children with Down syndrome are much more prone to the following medical problems: congenital heart disease (50%), excessive recurrent upper and lower respiratory infections (90%), cataracts (2-3%), atlanto-axial instability symptoms (first and second cervical vertebral instability with symptoms of spinal cord compression) (1-3%), epilepsy (10%),

gastroenterology obstruction (8%), leukemia (1%), hypothyroidism (variable up to 15% by adult life), hydrocephaly (1%), hearing deficits (10-30%), sleep apnea (5%), and Alzheimer disease (very common into adult life).

Routine medical evaluations should check for hearing and vision problems, symptoms of neck subluxation (neck pain, head tilt, back pain with radiation down legs, bowel-bladder incontinence, spasm, and increased reflexes of the lower extremities), hypothyroidism, dental disease, and nutritional deficiencies.

Even though most individuals with Down syndrome are placid, affectionate, good-natured, and have a good imagination and mimicry, by adulthood it is estimated that 20% of these individuals are prone to depression, and have obsessive-compulsive qualities and some autistic-like behavior with avoidant and oppositional components.

Controversial therapies for Down syndrome include cosmetic surgery, megavitamins, fetal cell infusion, antioxidant vitamin-mineral-amino acid preparations, and piracetam. Also known as nootropin, piracetam has received testimonial claims of enhancing cognition. This has not been proven scientifically, so the agent is not FDA-approved or available in the United States. Donepezil (Aricept®), an acetylcholinesterase inhibitor that has been used for the protection and treatment of cognitive impairment in adults with Down syndrome who are prone to Alzheimer disease, has not had adequate clinical trials to confirm its efficacy.

Fetal Alcohol Syndrome (FAS)

Since its specific recognition and documentation in the early 1970s, it appears that the incidence of severe fetal alcohol syndrome is 3/1000. Alcohol, more than any other substance of abuse, has devastating effects on brain growth and maturation. Characteristic features of this syndrome include prenatal and postnatal growth deficiency; microcephal; moderate mental deficiency; short eyelid openings (palpebral fissures); short upturned nose; flat midface; smooth upper lip without the development of philtral folds; heart, brain, kidney, and limb malformation; and irritable hyperactive behavior.

The estimated risk of fetal alcohol syndrome, based on one drink equals 12 oz. of beer equals 4 oz. of wine equals 1 oz. of hard liquor, is as follows:

- 4 to 6 drinks per day or more equals 40% risk
- 2 to 4 drinks per day equals 10% risk
- 1 to 2 drinks per day equals 1% risk

A concern recently expressed is the overdiagnosis of Fetal Alcohol Effect in children who have some features of the syndrome but do not meet all the criteria. The concern is stigmatizing the child, terminating diagnostic studies to ascertain the true diagnosis, and loss of educational and social supports for the child and mother.

Because of the extreme hyperactive behavioral problems demonstrated by those with fetal alcohol syndrome, various medications have been tried, including methylphenidate (Ritalin®) and clonidine (Catapres®), in combination with behavioral training.

Fragile X Syndrome

This disorder is caused by an expansion gene with excessive CGG triplet code repeats at the tip of the X chromosome and is one of the most common causes of mental retardation. Its cause explains the variable incidence in males (1/1250 affected) and females (1/2500 affected), and this discovery has improved our understanding of other neurogenetic disorders characterized by expansion phenomena of triplet codes (e.g., Huntington disease and myotonic dystrophy).

Individuals who have fewer than 50 CGG triplet repeats in the active gene area are not affected. Transmitting males and carrier females have a gene area expanded to an unstable number of CGG repeats (50 to 200). They are not affected, but in subsequent generations the unstable gene area can expand even further (more than 220), and the gene becomes nonfunctional. This gene is thought to be a carrier protein for messenger RNA that is important in maturation of fetal and early infant brain neurotransmission connectors. Because females have a second X chromosome that may function normally, they often are less affected. It is also now appreciated that males with fully expanded genes (beyond 220) may have borderline or low average intelligence on psychometric testing, rather than actual retardation. This is explained by the fact that these males have some mosaicism (mixed population of functional and nonfunctional cells), and/or the switch that activates the gene (by chemical methylation) may still function.

As adults, fully affected males display typical physical features that include tall, slender body style, elongated face with a high forehead and enlarged head, large ears, arched roof of mouth, flexible joints, and macro-orchidism (very large testes).

The behavior profile for fully affected males is characteristic and reveals autistic-like behavior, including gaze avoidance, hand flapping, resistance to change, and impulsiveness. Speech often is cluttered with a repetition of sounds and words, with articulation errors, an inability to stay with the topic, and the making of tangential comments. Interpersonal interaction improves after early childhood, as does the performance of daily living and self-care activities. But for many, independent living is not possible.

Affected females seem to have more psychiatric problems than males. These include depression, schizotypal symptoms with an odd communication pattern, inappropriate affect, unusual thought content, and increased emotional liability. Females do not often have the altered physical features observed in males.

Drug therapy includes the use of stimulants or beta-blockers for hyperactivity, carbamazepine (Tegretol®) and divalproex (Depakote®) for mood liability and aggression, and SSRIs for aggression, anxiety, and obsessive-compulsive behavior.

Fetal Anticonvulsant Syndrome (Fetal Hydantoin Syndrome)

Epilepsy is not uncommon in the general population (1/200). It has been recognized since the early 1970s that maternal exposure to various anticonvulsant medications during pregnancy has resulted in the birth of infants with a variety of congenital defects (brain, heart, kidney, limb, and clefts of lip and palate), prenatal and postnatal

growth deficiencies, and varying degrees of mental retardation.

Characteristic features after exposure to phenytoin (Dilantin®) and other hydantoin anticonvulsants are growth deficiency with microcephaly, midface hypoplasia with flat nasal bridge, a false impression of widely spaced eyes, undergrowth or lack of fingernails and toenails, and stippling calcification of bone growth centers and cartilaginous tissues. The exact cause of these effects is unknown but is thought to be associated with epioxide hydrolase metabolites of the anticonvulsant. It is estimated that 10% of children exposed to hydantoin anticonvulsants in the perinatal period have growth and congenital defects and that another 30% may have mental retardation and/or severe learning problems. Two nonhydantoin anticonvulsants, carbamazepine (Tegretol®) and valproic acid (Depakene®), have been more commonly associated with neural tube defects, i.e. spina bifida.

Treatment of maternal epilepsy includes monotherapy with the anticonvulsant most appropriate for the type of seizure at the lowest effective dose. It appears that pre-pregnancy use of folic acid by the mother may have a role in preventing or decreasing the development of fetal anticonvulsant syndrome.

Noonan Syndrome

Noonan syndrome is a much more common disorder than is generally recognized with an estimated frequency of 1/2500. Half the time it appears to be dominant in families with a wide range of clinical findings in affected individuals and half the time it appears spontaneously. One-fourth to one-third of affected individuals have mild mental retardation or significant learning disability. A defective gene on chromosome 12p is now thought to be the cause of this syndrome.

Males and females are equally affected and have a similar phenotype to that described in Turner syndrome. Their physical appearance is characterized by short stature with a short and webbed neck, droopy eyelids (ptosis), low-set and cupped ears, wide-spaced and downward slanting eyes, and a receded chin. The chest sternum is sunken (pectus excavatum), there is outward (valgus) positioning of the extremities, various congenital heart and kidney malformations, and notable edema at birth. Other features of Noonan syndrome are failure to thrive due to poor feeding patterns, gastrointestinal reflux, congestive heart failure, platelet adhesion problems with increased risk of bleeding (aspirin should be avoided), delayed puberty, and reduced fertility in males.

Behavior varies from stubborn and repetitive with preoccupations and immaturity, to lovable and unflappable with overconcern for the well-being of others.

Tuberous Sclerosis (TS)

Tuberous sclerosis resulting in mental retardation (50%) with autistic features and seizures (90%) has an occurrence of 1/10,000. Recent discoveries have identified two gene areas in which coexisting defects result in tuberous sclerosis. These areas are chromosome 16 (the tuberin gene), and chromosome 9q34, (the hamartin gene). Eighty-five percent of patients have had spontaneous mutations on these chromosomes and do not have a family history of TS. At least 25% of infants who present with infantile spasm seizures will have TS. If seizures (including infantile spasm seizures) occur before the age of 18 months, the incidence of mental retardation is near 90%.

Tuberous sclerosis is a classic neurocutaneous syndrome. Definitive diagnostic criteria include facial angiofibromas (small tumors over the cheeks that appear like acne), digital nail bed tumors, eye tumors (retinal hamartoma), brain tumors (cortical and subependymal tumors [around the ventricles]), and kidney tumors (angiomyolipomas). Supporting evidence includes hypopigmented "ashleaf" spots, shagreen patches (skin plaques of fibrous tissue), gingival fibromas, dental enamel pits, kidney cysts, cardiac muscle tumors (rhabdomyoma), pulmonary cystic lesions, infantile spasms, and a member of the immediate family with TS.

Care includes management of seizures; dermabrasion for excessive facial angiofibromas; and monitoring for intracranial ventricular obstruction, hypertension, cardiac arrhythmias, and congestive heart failure in infants with cardiac rhabdomyomas, renal tumors, pulmonary cystic lesions with pneumothorax, and pulmonary hypertension.

Behavior disorders associated with TS include hyperkinetic states with aggression and sleep disturbances. Acute hydrocephaly should always be ruled out in patients who bang their heads.

Klinefelter Syndrome (XXY)

This syndrome occurs only in males and has an overall occurrence of 1/1000, with a low percentage of individuals having borderline to mild mental retardation. Chromosome testing reveals an extra X chromosome, making the karyotype XXY. The etiology of this syndrome is nondisjunction of the X chromosome during either first or second meiotic division in either parent, but more commonly comes from the mother if her age is advanced.

Physical features include tallness; mild obesity in adult life; male gynecomastia (breast enlargement) in 40-50% with a risk of breast cancer in 4%; small (2 cm or less), firm, fibrotic testes with infertility and diminished sex drive; and increased incidence of pulmonary disease, varicose veins, and autoimmune disorders.

Behavioral and psychiatric disorders often occur before adolescence. Children often are immature and shy, have language and learning problems, are anxious and aggressive, and may engage in antisocial acts, i.e., fire setting and cruelty to animals. As adults they may express aggressive and antisocial behavior.

Treatment includes providing educational supports, behavior management, social skills training, testosterone replacement beginning at adolescence, and reduction mammoplasty.

Williams Syndrome

This disorder is more commonly recognized since the recent micro-gene deletion discovery on chromosome 7q minus 11.23. This is known as the elastin gene, and failure of its expression results in the Williams syndrome. The incidence is thought to be near 1/10,000.

Characteristic physical features include the so-called elfin-like facial features with prominent lips and cheeks and the appearance of edema around the eyes, broad nasal bridge and nasal tip, prominent ears, stellate patterning of the iris, hoarse voice, relative short stature, infantile hypercalcemia, and various cardiac structural defects. These defects include supra-valvular aortic stenosis (narrowing of the aortic valve outlet), pulmonic arterial stenosis (narrowing of the pulmonary artery), and septal defects (opening in the muscle wall between the pumping chambers).

Behaviors characteristic of this syndrome include mild to moderate mental retardation, short attention span, precocious vocabulary with excessive inappropriate cliches and stereotyped phrases, chattiness, and outgoing personality with socially disinhibited responses. The engaging personalities of individuals with this disorder can result in an overestimation of their general cognitive abilities. They usually have poorer relationships with peers than with adults, toward whom they are outgoing and excessively affectionate.

Treatment includes decreasing calcium and vitamin D in the diet if hypercalcemia is present and monitoring for hypertension. Some individuals may require cardiac structural correction and medication for hypertension and congestive heart failure. Appropriate behavior modifications and individualized education programs are important.

Prader-Willi Syndrome (H3O)

The estimated occurrence of this syndrome is 1/10,000, and the clinical features are particularly striking. Many of these infants are born with a history of poor in utero movement. They may have extreme hypotonia at birth requiring mechanical ventilation and may appear to have congenital muscle myopathy. During infancy the hypotonia persists, causing feeding difficulties and significant failure to thrive. Both males and females have hypogonadism with micropenis and cryptorchism in males and hypoplasia of the labia in females. In childhood they develop an extreme obsessive-compulsive eating disorder and become quite obese if their diet is not monitored. Some individuals weigh hundreds of pounds. Often parents have to put locks on food cabinets and the refrigerator door. Their passion for food leads to stealing and hoarding food and at times even eating inanimate objects.

Additional physical features include short stature, small hands and feet, bitemporal narrowing of the skull, almond-shaped eyes, and a triangular-shaped mouth.

Behavioral traits are outstanding as well. Beyond the striking obsessive-compulsive eating habits, they often have a lot of emotional lability with outbursts of rage, aggression (being especially aggressive to those withholding food), stubbornness, and belligerence. Between episodes, the difficult behavior improves, and they become good natured, placid, cooperative and friendly, but impulsively chatty. Fifty percent are mildly retarded, and only a few individuals with this disorder can function independently in adult life. They have difficulties in social relationships with peers and are described as immature, lonely, and isolated. Additionally they may have self-injurious behavior, deliberately picking and scratching their skin as well as rectal digging.

The genetic defect in this syndrome is now understood. Seventy-five percent of individuals with this disorder have a microdeletion of

the paternal chromosome area 15q11-q13. Another 20% have 2 maternal chromosomes 15 (uniparental dizomy), thus illustrating a new genetic principle in which only one allele gene of a parent actively expresses the gene product. Another few percent have either a submicroscopic deletion in the active gene area or methylation switch abnormalities (imprinting) that turn the gene off.

Treatment includes enforcing lifelong strict nutritional guidelines and exercise programs to avoid morbid obesity, monitoring for the complications of diabetes, hypertension, cardiac disease, sleep apnea, and self-mutilation. Social relationship skills training, behavioral strategies, and the use of various psychotropic medications for obsessive traits and occasional psychiatric symptoms all have a role. To date no medications have shown long-term effectiveness in controlling appetite in individuals with Prader-Willi syndrone. Growth hormone, although controversial, has been beneficial in decreasing body mass index. Sex hormone replacement has not been proven beneficial in controlled trials and may result in increased aggressive behavior.

Angelman Syndrome

This is another syndrome that has striking physical and behavioral features. The occurrence of the disorder is estimated also at 1/10,000. The genetic features of this disorder are similar to those described in Prader-Willi syndrome. In this disorder, however, the microdeletion is on the maternal chromosome 15 q11q13 gene in 75% of the cases. Another 3 to 4% have two paternal chromosomes 15 (uniparental dizomy), and the remainder is thought to have undetectable deletions or methylation switch abnormalities (the chemical process that activates or inactivates gene expression).

Physical features include a stiff, jerky, ataxic gait, microcephaly with a flat back of the head, and a prominent jaw with a protruding tongue.

Mental retardation is usually severe, and expressive speech is never achieved. These individuals have the behavioral trait of frequent outbursts of laughter, along with the stiff jerky movements. Eighty percent develop seizures that disappear in adult life.

Supportive care includes control of seizures (valproic acid seems to be the drug of choice), occupational and physical therapy, development of nonverbal ways to communicate, behavior management, respite care, and safety adaptations to the environment.

Conclusion

This review has summarized the known etiologies of mental retardation, a condition that occurs in three percent of the population. The etiology and the best strategies for medical, behavioral, mental health, and educational treatments are now better understood for many of the developmental disability syndromes associated with mental retardation. Making a specific diagnosis is helpful in providing specialized treatment and genetic counseling about recurrence risks, natural history, and prenatal diagnosis. The Regional Center System of California serves the needs of individuals with mental retardation and their families.

Suggested Readings and Sources of Information for Health Care Providers

General Discussion of Developmental Disability

Arnold J. Capute and Pasquale J. Accardo, *Developmental Disabilities in Infancy and Childhood*, Paul H. Brookes, Baltimore, 1991

Steven Parker and Barry Zuckerman, *Behavioral and Developmental Pediatrics, A Handbook for Primary Care*, Little, Brown and Co. Boston, 1995

Individual Developmental Disabilities

Mary Louise Buyse, *Birth Defects Encyclopedia*, Blackwell Scientific Publications, Cambridge MA, 1990

Kenneth Lyon Jones, *Smith's Recognizable Patterns of Human Malformation – 5th Edition*, WB Saunders Co, 1997

Roger E. Stevenson, Judith G. Hall, Richard M. Goodman, *Human Malformations and Related Anomalies - Vol. 1.* Oxford University Press, NY, Oxford. 1993

Sam Goldstein and Cecil R. Reynolds, *Handbook of Neurodevelopmental and Genetic Disorders in Children*. The Guilford Press, NY, London. 1999

Gregory O'Brien and William Yule, *Behavior Phenotypes. Clinics in Developmental Medicine No. 138*. Mackeith Press, Cambridge University Press, NY. 1995

Siegfried M Pueschel and Jeanette K. Pueschel, *Biomedical Concerns in Persons with Down Syndrome*, Paul H. Brookes, Baltimore. 1992

Randi Jenssen Hagerman and Amy Cronister, *Fragile X Syndrome, Diagnosis, Treatment, and Research – 2nd Edition*, The John Hopkins University Press, Baltimore and London. 1996

Web Sites

Alta California Regional Center
www.altaregional.org

California Department of Developmental Services
www.dds.cahwnet.gov

Association of Regional Centers
www.arcanet.org

Medical Care for Conditions of Developmental Disability
www.ddhealthinfor.org

Psychotropic Medication

Use in People with Developmental Disabilities

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Editor's Note: Medication references contained in this article may need to be updated over time as additional research is completed.

Abstract

Psychotropic medications are frequently used to treat undesirable behaviors in persons with developmental disabilities. Successful use of these drugs is dependent on accurate assessment of the psychiatric disorder or behavioral problem. Treatment of aggression and self-injurious behavior and the use of antipsychotics, antidepressants, mood stabilizers, anxiolytics, beta-blocking agents, and naltrexone will be discussed.

Introduction

Psychotropic medications are frequently employed in persons with developmental disabilities to treat psychiatric disorders such as schizophrenia, depression, bipolar disorder, and anxiety disorders much as they are used in the general population. The prevalence of psychotropic medication use in this population varies considerably depending on the setting. The mean prevalence rate reported by Baumeister et al., is 57%, 41%, and 22% in institutions, community, and school settings, respectively.[1] Antipsychotic drugs are the most frequently used.[1,2]

Psychotropic drugs are also often employed in persons with developmental disabilities to control or manage "maladaptive" behaviors including aggression, hyperactivity, self-injury, and disruptive or inappropriate social behavior.[1,2] These are vague and frequently poorly defined behaviors rather than psychiatric syndromes. The use of drugs for these behaviors is controversial. The efficacy of psychotropic medications for these purposes is poorly documented and ill defined.

The key to appropriate use of psychotropic medications in people with developmental disabilities is accurate assessment of the psychiatric disorder or behavior problem. It is crucial to rule out environmental causes of the problematic behavior prior to initiation of pharmacologic therapy.[3] When other causes cannot be found, or it is difficult or impossible to give a psychiatric diagnosis to a given individual with a developmental disability, target symptoms should be specifically defined. Length of a medication trial depends on the type of drug used and its purpose. Most antipsychotic medications take at least a week to reach steady state and may take longer to reach maximum therapeutic effect. Other drugs with much longer half-lives, such as fluoxetine, may take up to a month to reach steady state and will require a longer trial. Medication should be maintained at least two to three months at therapeutic doses or levels to adequately determine efficacy. A drug should be discontinued if there is no evidence of improvement or if side effects outweigh its benefit.

Aggression and Self-injurious Behavior

People with developmental disabilities are frequently referred to psychiatrists for treatment of aggression and self-injurious behavior (SIB).[4] SIB is defined as any chronic, repetitive behavior that results in physical injury to a person's own body and is more common in those with severe and profound developmental delay. Aggression may take many forms, including physical assault, verbal aggression, or property destruction. Self-injurious or aggressive behavior may be caused by inability to communicate and/or environmental influences; therefore, examination of environmental factors is crucial. The cause of problem behavior may also be a result of a medical illness, and a thorough review of potential medical causes, including a physical examination, should be performed.[3] Aggression and SIB may be caused by pain from dental problems, dysmenorrhea, gastrointestinal upset, or otitis media. Aggression and self-injury may also result from psychiatric disorders such as mood or psychotic disorders.

Antipsychotic medications are frequently employed to treat these behaviors. There are reports in the literature that these drugs will decrease SIB and aggression in people with developmental disabilities. However, it is unclear that these effects are specific.[4] The sedative effects of antipsychotic medication may simply suppress all behavior. Additionally, these agents may decrease learning in some individuals who have a developmental disability.[4]

Side effects of the antipsychotic medications must also be considered when they are used in this population. People with developmental disabilities experience the same problems as others in the general population. Identification of side effects may be more difficult due to communication problems. Akathisia, a subjective sense of restlessness and an inability to sit still, is frequently unrecognized and may be confused with an increase in maladaptive behaviors or hyperactivity.[5] Tardive dyskinesia, akathisia, and dystonia are seen in people with

developmental disabilities as a result of antipsychotic administration. Symptoms of the tardive syndromes may mimic or be confused with stereotypy or an underlying neurologic problem. A baseline assessment and follow up are crucial.

The atypical antipsychotic medications clozapine, risperidone, and olanzepine may be of use in selected individuals that exhibit aggressive behavior. Clozapine has been reported to improve aggressive and self-injurious behavior in people with developmental disabilities.[6,7,8,9] Risperidone has also been reported to reduce aggression and self-injurious behavior.[10,11] No published reports on the effectiveness of olanzepine or sertindole (due to be released) could be found.

There are numerous reports on the use of naltrexone to treat self-injurious behavior in developmentally delayed people. Theoretically, a person who engages in self-injury releases endogenous opioids and receives positive reinforcement from the narcotic effect of these endogenous substrates.[12-14] Naltrexone blocks the effect of exogenous narcotics and endogenous opioids. Most of the literature describing the use of the opioid antagonists comes from case reports and open trials. It appears that some individuals may benefit from the use of naltrexone, but not everyone who engages in self-injury will benefit from the drug. A trial of naltrexone may be warranted in an individual for whom no apparent environmental, physical, or psychiatric cause is identified. The dose of naltrexone used for this purpose is usually 1.0 mg/kg/day to 2.0 mg/kg/day.[12-15]

Beta-blocking medications such as propranolol, metoprolol, and nadolol have been reported to decrease aggression and self-injury in people with developmental disabilities.[16-19] However, most of the published reports are also case reports and open studies. Some persons may benefit from the use of these drugs, especially if the aggression and SIB are caused by neuroleptic-induced akathisia.[15] Doses of these agents reported to treat these behaviors in individuals with developmental disabilities vary from 200 mg to 1000 mg per day.[19]

Many other drugs such as antidepressants, anxiolytics, and anticonvulsants have been employed to treat or control aggressive and self-injurious behavior. Unfortunately, none of these drugs specifically treats these behaviors. They may be effective in a specific individual; however, there is no one medication that will eliminate these maladaptive behaviors.

Psychotic Disorders

Persons with developmental disabilities suffer from the same psychotic disorders as the general population. Manifestation of the illness may be different in this population. The person's ability to communicate and verbal and cognitive sophistication must be evaluated prior to establishing a diagnosis.

The choice of a medication should be based on the individual's health status and medication history. A previous response to a medication may predict a response. Medications such as thioridazine

(Mellaril®), chlorpromazine (Thorazine®), loxapine (Loxitane®), and clozapine (Clozapine®) may lower the seizure threshold in individuals with a seizure disorder and should be used with caution. The presence of extrapyramidal symptoms such as pseudoparkinsonism and akathisia may make the higher potency medications (haloperidol, thiothixene, fluphenazine) less desirable. Lower potency medications (chlorpromazine, thioridazine, mesoridazine) cause more sedation, constipation, urinary retention, and cognitive impairment.

Antipsychotic medications should be used at the lowest dose possible. This is important not just for side effect and therapeutic reasons, but also satisfies state and federal guidelines for using the least restrictive intervention.[20]

Some literature suggests that the newer medications may be beneficial in this population.[6-11] Risperidone, used at low doses, may have fewer side effects than the traditional agents and is well tolerated in this population.[10,11] Clozapine should be reserved as a last line agent because of its potential toxicity. Currently, there are no published reports of olanzapine use in this population; therefore, its place in therapy is not yet established.

Mood Disorders

Mood disorders may also present differently in people with developmental disabilities than in the general population. Again, depending on the functioning level of the individual, it may be difficult to make definitive diagnosis of bipolar disorder or depression. A history of behavioral cycling, irritability, and sleep problems may indicate the presence of a mood disorder.[21] Changes in adaptive functioning, self-injury, and aggression may also be present in people with mood disorders.[22]

Valproic acid, carbamazepine, and lithium are effective medications for treating people with bipolar disorder regardless of level of functioning. However, the literature is sparse regarding the use of these drugs in treating people with developmental disabilities. One open study and several case reports show good results using valproic acid in people with developmental disabilities for treatment of what appears to be affective disorders.[21-25] Serum valproic acid levels ranged from 64 µ/ml to 144 µ/ml in these reports. Valproic acid is usually well tolerated, however, many people experience gastrointestinal upset and sedation. Thrombocytopenia is associated with the use of valproic acid and should be periodically monitored. Other side effects that have been reported in people with developmental disabilities include pancreatitis and cholecystitis.[26]

The Food and Drug Administration has not approved carbamazepine for use in bipolar disorder; however, it is effective in treating people with this disorder.[27] Carbamazepine has a reputation as having "antiaggressive" properties, however, there is little literature to support this concept. Carbamazepine appears to be most effective in people who display symptoms consistent with bipolar disorder and also have a seizure disorder.[28-30] Carbamazepine has many side effects, including gastrointestinal upset, ataxia, and diplopia.[31] These side effects can be minimized by starting the drug at low doses and increasing slowly to the desired serum concentration. Other side effects that need to be monitored include leukopenia, jaundice, and electrolyte imbalances. Because of the potential for agranulocytosis and leukopenia, concomitant administration of carbamazepine and other drugs that can also cause bone marrow suppression should be avoided.[31] Clinically significant drug interactions occur with carbamazepine. This drug is a potent inducer of liver enzymes in the cytochrome P-450 system and reduces the serum concentrations of other drugs metabolized in that system. Drugs that are significantly affected by carbamazepine include, but are not limited to, phenytoin, phenobarbital, primidone, valproic acid, antipsychotic medications, oral contraceptives, and theophylline.[31]

Lithium has been extensively used in persons with developmental disabilities for many purposes.[32-34] Again, as with the other drugs used to treat people with mood disorders, lithium appears to be most effective in people who have cyclical behavioral symptoms. Pary reviewed the literature up to 1991 for the use of lithium in this population and provided the following guidelines: Serum concentrations of at least 0.5 mEq/ml to 1 mEq/ml should be obtained unless the person responds or side effects prevent achievement of that level. The trial should last for at least six to eight weeks at adequate serum concentrations before concluding that the person will not respond.[32] Lithium can cause many side effects.[35] People taking this drug frequently have gastrointestinal upset, tremor, muscle weakness, and fatigue. Hypothyroidism, leukocytosis, polyuria, and polydipsia can also occur. Lithium is excreted exclusively through the kidney and this must be taken into consideration in people with impaired renal function.

Antidepressant medications have also been used to treat people with developmental disabilities. The amount of formal research with these drugs in this population is limited. Tricyclic antidepressants have been reported to be beneficial in persons exhibiting symptoms of depression, psychosis, and tantrum behavior, and in those who have a high social age.[36] The tricyclic antidepressants have significant side effects, including dry mouth, urinary retention, constipation, and blurry vision. They also cause significant sedation, hypotension, cardiac arrhythmias, and a decrease in the seizure threshold. The tricyclic antidepressants may also cause deterioration in symptoms of irritability, lethargy, social withdrawal, and hyperactivity.[37] These side effects may limit their use in people with developmental disabilities.

The selective serotonin reuptake inhibitors (SSRIs) are a relatively new group of antidepressants. This class of drugs includes fluoxetine, paroxetine, sertraline, clomipramine, and fluvoxamine. These drugs have been used to treat people with developmental disabilities who appear to have symptoms of depression.[38-42] Symptoms of depression may present in a person with developmental disabilities as social withdrawal, sleep and eating changes, self-injury, and aggression.[38] The SSRIs are not without side effects. Two studies report increased irritability, hyperactivity, agitation, anorexia, and insomnia in developmentally delayed people who received fluoxetine.[41,42] Other side effects include gastrointestinal upset and sexual dysfunction. These drugs may be safer and less toxic than the tricyclic antidepressants. The clinician must be aware, however, of the potential for worsening of behavior problems, especially with fluoxetine. Drug interactions include significant inhibition of specific isoenzymes of the cytochrome P-450 system. Concomitant use of other serotonergic drugs may result in the serotonin syndrome. These symptoms include mental status changes, hyperreflexia, tremor, restlessness, and diaphoresis.

Antidepressants and mood-stabilizing agents are effective in treating affective symptoms in people with developmental disabilities. The choice of medication must be based on the person's presenting symptoms and medical status. Valproic acid and carbamazepine may be more effective than lithium in treating cyclical disorders in people with seizure disorders and brain damage. Tricyclic antidepressants should be avoided in people with active seizure disorders. The SSRIs may be useful in treating people with symptoms of depression and cause less toxicity than the tricyclic antidepressants, but may also exacerbate some behavioral problems.

Anxiety Disorders

Anxiety disorders, including obsessive-compulsive disorder (OCD), occur in people with developmental disabilities. OCD has been described and treated in people with developmental disabilities.[43-46] Obsessive symptoms may be difficult to ascertain due to communication limitations. However, the compulsive symptoms may be evident in ritualistic behavior. Currently, several SSRIs are indicated for use in OCD. These include clomipramine, fluoxetine, fluvoxamine, and paroxetine. Any of these may be used to treat a person who appears to be engaging in ritualistic behavior that is interfering with activities of daily living. The same cautions and potential side effects listed above apply for the use of these drugs for this indication. Clomipramine is a tricyclic antidepressant and has the same side effects as that class of drugs. It is also of concern because of its potential to cause seizures.

Benzodiazepines have been used for "as needed" treatment for any type of behavioral disorder. The use of these drugs is declining for a few reasons: there is little evidence that they control or reduce problem behaviors in people with developmental disabilities, they may actually exacerbate agitation and aggression, and long-term use may lead to tolerance and withdrawal symptoms upon discontinuation.[47] Chemical restraint for the control of an acutely violent or agitated person is sometimes required. Benzodiazepines are useful in these situations, but should be avoided for long-term use.

Buspirone may be useful in people with developmental disabilities and anxiety. A few studies have found it to be useful in treating anxious, aggressive, and self-injurious behavior in this population.[48,49] The doses usually employed are between 15 mg/day and 45 mg/day.[48,49] Buspirone has a favorable side effect profile, however, clinicians must keep in mind that no well-controlled methodogically sound studies have assessed the effects of this drug in people with developmental disabilities.

Conclusion

Appropriate use of psychotropic medications depends on accurate assessment of the underlying psychiatric disorder. The environment can also influence behavior and must be taken into consideration when prescribing medications. Unfortunately, good controlled, methodogically sound studies are rare. Frequently, the use of psychotropic medications is empiric at best. The clinician treating these persons must be aware of the limitations of these drugs and use them only when clinically necessary and when efficacy is demonstrated by symptom resolution.

References for this article are available upon written request to: California State Board of Pharmacy, Attn.: *Health Notes* Developmental Disabilities References, 400 R Street, Suite 4070, Sacramento, CA 95814.

Selected Tips for Counseling Individuals with Developmental Disabilities

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Children and adults with developmental disabilities who live in the community are a unique and greatly underserved group in our society. The pharmacist has an opportunity to make meaningful contributions to the care of these individuals by improving the use of medications.

For a variety of reasons, including federal regulations (HCFA and Health and Human Services), practice guidelines (ACDD), and class action suits, there has been a significant movement in the United States to transfer individuals with developmental disabilities out of institutions (developmental centers) and into the community. Additional pressure from advocacy groups desiring placement of individuals closer to their homes and in less restrictive environments has led to new levels of care. This has resulted in a demand for new types of services, which has in turn created opportunities for pharmacists. Serving individuals with developmental disabilities is a fine example of pharmacists making a significant difference by providing cognitive services in addition to prescription services.

In California, most individuals with developmental disabilities are living in the community and are cared for under the auspices of one of the 21 Regional Centers spread throughout the state. These centers are nonprofit agencies that have responsibility for providing care and shelter to the thousands of individuals who reside in the community or have been returned to the community from a State Developmental Center. There are numerous agencies and support groups linked to these Regional Centers that provide support, training, teaching, daily care, and community-based work programs for individuals with developmental disabilities.

The pharmacist has an excellent opportunity to provide a variety of services to adults and children with developmental disabilities living in the local community. This article will describe how the pharmacist can counsel and assist these individuals, improve the delivery of their

TABLE 1

*Barriers to Effective Care for People with Developmental Disabilities**

Difficulty of Cognitively Impaired Patients

Cognitively impaired patients:

- may not be able to communicate history of symptoms.
- may be unable to tolerate waiting room stays.
- may be threatened by aspects of the office or hospital environment, such as needles, physical examination of private areas, unfamiliar clinicians.
- may resist or fight when confronted with an examination.
- may require ancillary or family help for even the smallest procedure.

Overall, a history and physical requires three times longer than average.

Difficulty of Physically Impaired Patients

Patients with physical impairments:

- may require ancillary help to gain position, because many patients (despite normal intelligence) are severely compromised in movement.
- similarly, may have severe communication difficulties.
- because of their appearance, and/or unintelligible speech, are misperceived as cognitively impaired.

Attitudes of Health Care Professionals

Providers of care:

- may not have had proper training to see difficult patients; training programs and curricula may not have emphasized or even included these patient's special needs.
- hold negative attitudes toward caring for people with disabilities, based on fear/apprehension or a few time-consuming and difficult personal encounters.

The current health care delivery system:

- is ill-equipped to handle the deinstitutionalized patient population.
- is inadequately reimbursed by Medicaid, Medicare, and/or private insurance agencies that do not recognize the greater time and effort required to provide adequate care for the developmentally disabled.

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medications, increase awareness of their special needs, and serve as their advocate in regard to medication-related issues.

Table 1 lists some barriers to effective care for the individual with developmental disabilities.

Meeting Patients with Developmental Disabilities

In many instances the person with a developmental disability is not the individual the pharmacist is likely to first encounter. More often, the pharmacist will encounter a family member or care provider, case manager, residential home manager, or another member of the community who has oversight and some responsibility for the individual.

Pharmacists are encouraged to make every effort to meet the individual with developmental disabilities. It is always helpful to put a face with the prescription. In fact, the authors believe it is imperative that, time and practice site permitting, the pharmacist ask to meet the individual. Meeting and learning about the patient can help dispel inaccurate assumptions about people with disabilities and increase the comfort level for the pharmacist. A friendly greeting and handshake can be a first step in the communication process. Meeting the patient will also help the pharmacist better understand the myriad of responsibilities assumed by the local care managers, providers, and/or family members who have direct and indirect responsibility for the well-being of the patient.

Time spent by the pharmacist in building relationships can be tremendously beneficial, because the pharmacist can serve as an important link between the patient/caregiver and the health care system. Enhancing communication among all parties is a key element in caring for people with developmental disabilities.

It is important to realize that although an individual has developmental disabilities, it does not necessarily mean they cannot communicate with the pharmacist. Individuals with developmental disabilities have a wide range of intelligence and understanding of their surroundings, and they often have unique ways of communicating. Regardless of the intelligence level of the individual with a developmental disability, the pharmacist should remember that he or she can hear, unless the disability includes a hearing deficit.

Patient Records

Persons with developmental disabilities may not have complete medical records in one place. Many individuals with developmental disabilities are taken to the doctor's office by people who do not have complete details of their health issues and may not fully know why this particular person is seeing the doctor. In other instances the person taking the client to the doctor may have several health care issues to address with the physician or nurse practitioner in a short period of time – often 15 minutes or less. Finally, many individuals with developmental disabilities are moved to a new location without the benefit of their medical records being transferred. These same individuals may see multiple physicians who prescribe medications without completely knowing what other doctors have prescribed.

The pharmacy may be the only place where a complete medication record is maintained. Drug-drug, drug-food and drug-disease interactions, therapeutic duplications, excessive doses, and other therapeutic complications can and will be found in the pharmacy medication profiles of many of these individuals. Pharmacists can play an important liaison role with physicians, linking multiple medical diagnoses with the safest drug combinations.

Working with Care Providers

Care providers generally want as much information as they can obtain about medications, alternative health care options, and herbal remedies because they have seen side effects or adverse consequences from medications taken by their clients. They are eager for information on how to know if a medication is working, or how to spot adverse effects early. Individuals with developmental disabilities have the same health problems common in the general population, such as hypertension, diabetes, and arthritis. They also have additional

medical concerns more specifically related to their disabilities. Complying with the drug treatment needed for multiple conditions can be a daily challenge for caregivers.

There are recognized intelligence quotient (IQ) ranges that are used to classify developmental disabilities. Generally, an individual must have an IQ of less than 70 to be classified as having a developmental disability. Individuals with an IQ between 70-80 are classified with borderline intelligence, and those with an IQ above 80 carry the classification of normal intelligence. The *Diagnostic and Statistical Manual*, 4th edition, from the American Psychiatric Association (DSM-IV) is an excellent reference and contains the diagnostic criteria for developmental disorders.

Not all people served by the regional centers have low IQ's, since individuals qualify for services for a variety of reasons. Individuals may have genetic disorders, refractory seizures, head trauma from an accident in early life, or similar life events that have qualified them to receive care and support from the regional centers.

Many individuals with developmental disabilities are able to maintain their own apartments and care for themselves with minimal support. Some require supervision by a parent or guardian in order to live outside a care facility. Others live in residential care facilities that may be controlled (i.e., entry and exits are restricted for individuals' protection) and have high staffing ratios. Regardless of the setting, these individuals and their care providers are in need of the pharmacist's knowledge and skills.

The pharmacist will find that most providers caring for individuals with developmental disabilities in the community have limited formal training in medication or disease management, but have a strong desire to help maintain a good quality of life for their clients. Consequently, the pharmacist needs to explain complex and technical medical information in a way that the layperson can understand. Pharmacists are in an excellent position to provide medication information and training needed by care providers.

Care providers keenly observe patient's behavior, are knowledgeable about the history of these behaviors, and know how to successfully intercede with the patient when things start to get "out of control." They are the front-line people who observe and document seizures (for example, length, frequency, type), falls, food intake, and other signs of health and wellness or medical conditions. Care providers input to pharmacists can be very valuable in helping to assess the effectiveness of the patient's drug therapy.

Common Predispositions in Individuals with Developmental Disabilities

GI Issues

Individuals with development disabilities may have abnormalities in processing foods and drugs through the gastrointestinal (GI) tract. Constipation is a common GI problem, and a planned or daily bowel regimen is often necessary. Since the time of the developmental insult may influence later development of the liver and bowel, drug metabolism is significantly more variable in this population than in other individuals. Pharmacists must consider the pharmacokinetics and pharmacodynamics of all medications, including their interaction with food. Changes in transit time or the administration of drugs

Education and Counseling Tips

There are numerous ways to determine an individual's understanding of scientific information in order to communicate with that person in the most appropriate way. It is important that patients or caregivers are able to read prescription labels, interpret prescription instructions, and understand commonly used patient education materials. The pharmacist can most effectively promote drug therapy compliance by ensuring that information imparted is truly understood by the caregiver and/or the patient.

You could ask the patient or caregiver, "Do you understand what I just discussed with you?" This is an example of a close-ended question, one that yields minimum information by limiting an answer or preventing a detailed response. It may not be the best approach when you are trying to get as much information as possible because the most common response is a simple, "Yes." Rarely will you hear, "No." Generally, members of the public will defer to a health care professional, even if they really don't understand.

Alternatively, you can use an open-ended question such as "Please tell me in your own words what you understand about what I just told you." From the response to an open-ended question, the pharmacist will usually gather enough information to know what the patient or care provider comprehended.

Pay attention to the vocabulary, language, and nonverbal clues that the care provider or patient uses to tell you what he or she understands. This will help you gain insight into any confusion or misunderstanding that the patient/provider might have regarding your counseling and may prevent embarrassment.

Another approach is to explore the reasons for which the patient went to the physician in the first place, e.g., "Tell me why you went (or took the patient) to see the doctor." "How long have you (or has the patient) had this problem?" "What can you tell me about therapies that have been tried in the past?" And, "What has been your experience with drugs prescribed in the past?" These approaches require minimal time for pharmacists to gather meaningful, yet extremely important information that they can use to help patients or caregivers understand medical and drug directions.

There are many ways to help patients and caregivers improve compliance with a multiple-drug therapy regimen. One way might be color-coding the prescription bottles to indicate time of day or the number of times per day to take medicines. Other suggestions include gluing pictures of the pills to the medicine box, or creating a chart with other symbols or stickers – such as fork and plate for meals, and moon or stars for bedtime dosing.

with certain foods or other drugs may influence the absorption of medications, resulting in increases or decreases in drug levels.

Comorbid Disease States and Physical Limitations

Individuals with developmental disabilities may have comorbid disease states or physical limitations that impact their ability to properly take certain types of medications or operate an asthma inhaler or a spacing device such as an Aerochamber®. The use of special medication administration devices that are commonly used in the general population may not be optimal for this population.

It is not uncommon for individuals with developmental disabilities to have a small stature, which is important to consider for medications that are dose dependent on size or body surface area. An adult dose may not always be indicated. Certain oral dosage forms may also be problematic, especially bad-tasting liquids and pills that are large or have an unusual shape so they may be hard to swallow.

Certain genetic abnormalities can predispose patients to unusual drug effects. For example, persons with Down syndrome may be less able to eliminate aspirin, exhibit a greater response to the pupil-dilating effects of atropine, and be more sensitive to the toxicity of methotrexate.

Muscle strictures or severely tightened muscle masses, fused joints, and deformed limbs or digits are just a few examples of problems that may hamper proper drug administration or utilization. Poor eyesight, lack of intellect, or even medication side effects may preclude the patient from remembering your comments or reading the prescription label.

Pharmacists need to know about the patients' limitations in order to recommend the most appropriate drug and dosage form to meet their specific needs.

Drug Sensitivity

As a result of developmental insults to the nervous system or trauma early in life, individuals with developmental disorders may be extremely sensitive to the effects of many drugs, especially psychotropic medications. Sensitivity to psychotropic drugs may result in excessive drowsiness, confusion, impaired thinking, or unexpected opposite (paradoxical) effects, such as rage reactions. The common (anticholinergic) side effects of many psychotropic agents can contribute to other medical problems, such as gastroesophageal reflux disease or cognitive difficulties, or lead to constipation, urinary retention, dry eyes or blurred eyesight, and poor salivation. Psychotropics may also predispose patients to heat stroke, especially in hot weather.

A good rule of thumb is to expect the unexpected when monitoring drug effects in individuals with developmental disabilities.

It is not uncommon for individuals with developmental disabilities to have multiple medical problems, such as seizure disorders, malabsorption syndromes or other gastrointestinal conditions, cardiovascular abnormalities, skeletal malformations, or autonomic nervous system dysregulation. These many conditions may be challenging to treat, since it is rare that a single physician is responsible for all the medical conditions as well as the behavioral and/or psychiatric disorders. Many individuals receive multiple medications that may result in drug-drug interactions. Pharmacists can play an important role in assuring that all practitioners are aware of all medications being used, their indications, and monitoring requirements.

Handling Drug Interactions

Drug-drug interactions in individuals with developmental disabilities should be handled in the same way as in the general population. It is important to be aware of interacting drugs and the clinical importance of their interactions. Genetic factors, age, nutrition, liver disease, and hormones are just some of the influences on drugs and their effects. The pharmacist has a wealth of information, in computer programs and reference texts, that can be used to make sure that potential drug interactions don't result in adverse effects.

A change in the dose of one seizure medication (anticonvulsant) can cause a change in the effectiveness of other drugs the patient may be taking. Therefore, dose changes need to be viewed in the context of the total drug therapy for that individual. When considering drug interactions, it is important to review the cytochrome P-450 enzymes, of which the most commonly cited subgroup for producing drug interactions is the 3A4 isozyme. Some agents that commonly inhibit CYP3A4 are erythromycin, clarithromycin (Biaxin®), itraconazole (Sporonox®), nefazodone (Serzone®) and grapefruit juice. By inhibiting CYP3A4, these agents can raise the levels and increase the risk of toxicity of drugs such as cisapride (Propulsid®), cyclosporin, calcium channel blockers, some statins, and many others. Agents that commonly induce CYP3A4 are carbamazepine (Tegretol®), dexamethasone, griseofulvin, phenytoin (Dilantin®), and troglitazone (Actos®). These drugs can decrease the levels and possible effectiveness of the substrate drugs listed above.

There are also other isoenzyme families that can play a role in drug interactions (CYP1A2, CYP2C9, CYP2C19, CYP2D6). Drugs that affect these enzymes can impact the effectiveness or toxicity of drugs metabolized by them. For example, cimetidine (Tagamet®) strongly inhibits the breakdown (metabolism) of many drugs. Caffeine and nicotine (smoking) may also impact drug therapy. Caffeine may compete for metabolism with drugs such as clozapine (Clozaril®), olanzapine

Pharmaceutical Care – A Case Study

Pharmacists providing pharmaceutical care for persons with developmental disabilities will be presented with unique challenges. Ongoing review of drug regimens is necessary and interventions may be frequent, but the quality of life for the individual will be improved. The following case demonstrates the pharmacist's role in the proper care of individuals with developmental disabilities.

A consulting pharmacist for a regional center was referred to a 45-year-old woman who was living on her own in the community. At the time the pharmacist interviewed the patient, she was receiving 22 medications, all filled by the same local pharmacy. These medications and the patient's diagnoses are shown in the chart below.

Case Study Patient Information

Medication Profile

omeprazole (Prilosec®)
cisapride (Propulsid®)
oxybutynin (Ditropan®)
amlodipine (Norvasc®)
levothyroxine (Levoxyl®)
conjugated estrogens (Premarin®)
carbamazepine (Tegretol®)
paroxetine (Paxil®)
metoclopramide (Reglan®)
tramadol (Ultram®)
naproxen (Naprosyn®)
ibuprofen (Motrin®)
prochlorperazine (Compazine®)
sumitriptan (Imitrex®)
dicyclomine (Bentyl®)
promethazine (Phenergan®)
meclizine (Antivert®)
psyllium (Metamucil®)
diphenhydramine (Benadryl®)
hydrocortisone ointment
flunisolide nasal steroid inhaler (Nasalide®)
neomycin/polymyxin/bacitracin (Neosporin®) ear drops
steroid ear drop (Cortisporin®)

Diagnoses

- Seizure disorder
- Gastroesophageal reflux disease
- Chronic lower back pain
- Migraine headaches
- Hypothyroidism
- Hypertension
- Chronic nausea and vomiting related to motion sickness
- Vertigo
- Allergies, including food allergies to tomatoes and citrus, drug allergy to naproxen, and environmental allergies
- Urinary incontinence
- Hormone replacement therapy required due to previous surgery
- Depression
- Anxiety
- Ear infection

The consulting pharmacist visited the patient at home and inspected all her prescription and over-the-counter medications. She had medications prescribed by three different physicians but dispensed by one pharmacy.

During the evaluation, the pharmacist found that three medications were expired (Ultram®, Bentyl®, and Nasalide®). Three additional medications (Naprosyn®, Phenergan®, and Reglan®) were causing significant side effects and warranted discontinuation.

Several therapeutic duplications were identified. Phenergan®, Benadryl® and Bentyl® had all been prescribed for motion sickness although these agents are not indicated for this problem and were not effective. Compazine® and Phenergan® were both prescribed for nausea. Naproxen, ibuprofen, and Ultram® were all prescribed for pain, in addition to over-the-counter acetaminophen (Tylenol®).

Other medications were determined to be ineffective or inappropriate and were thought to be causing significant problems in thermoregulatory areas, balance, and memory. In total, the pharmacist's intervention resulted in elimination of nine different medications. Bentyl®, Naprosyn®, Ibuprofen®, Ultram®, Compazine®, Phenergan®, Benadryl®, Norvasc®, and Reglan® were all discontinued. Tenormin® was substituted for Norvasc® due to its effectiveness in lowering blood pressure and reducing the incidence of migraines. Allegra® was substituted for Benadryl® to minimize side effects such as dry mouth and to treat the patient's symptoms of allergies not relieved by Benadryl®.

The patient continued to have severe motion sickness. The pharmacist recommended that she try ginger capsules for one week, and inform the pharmacist of any positive or negative results. During the subsequent visit, the patient informed the pharmacist that the ginger capsules worked and that she had been able to travel in a car without experiencing nausea and vomiting for the first time in over four years. Of course not all recommendations will be this successful. As with all nonprescription medications, drug interactions should be kept in mind when pharmacists recommend any herbal or alternative medication.

The patient and her doctor were given a written copy of the pharmacist's consultation, and efforts are currently under way to address her health care issues in a collaborative fashion between the regional center, the patient, the doctor and the consulting pharmacist. These efforts will be ongoing. This case is just one example of how a pharmacist can provide pharmaceutical care that improves the quality of care for children and adults with developmental disabilities living in the community.

TABLE 2

Objectives for Best Practice in Treating Persons with Developmentally Disabilities*

1. Explore our biases and stereotypes with respect to people with developmental disabilities.
2. Understand techniques for caring for people with disabilities.
3. Understand patient and family expectations.
4. Understand the importance of preventive health care for patients with disabilities.
5. Explore techniques to enlist other health care providers, the family, or the guardian in assisting and providing health care.
6. Explore ethical and management controversies that arise in caring for people with disabilities.

** This table was first published in the* Western Journal of Medicine *(D. Doostan, M.Wilkes, Treating the Developmentally Disabled,* WJM *1999;171:92-96) and is reproduced by permission of the* WJM.

(Zyprexa®), or theophylline (TheoDur®, others) causing increased drug levels and possible toxicity. Smoking can increase the metabolism of drugs such as clozapine, olanzapine, and theophylline.

A complete review for drug-drug interactions needs to include alternative medications, herbal therapies, over-the-counter (OTC) medications, and nutritional or dietary supplements. More and more individuals in general are using alternative treatments, such as glucosamine for osteoarthritis, St. John's wort for depression, and gingko biloba for mental alertness. Some of these agents have side effects or could alter the metabolism of prescription drugs. Pharmacists need to use their drug information resources to help patients and caregivers make good choices with alternative medications.

Sometimes the families of individuals with developmental disabilities hear of unconventional and often expensive dietary, medical, or other therapies that have unknown efficacy. Advertising appeals to their emotions or the wish to "cure" developmental disabilities. Pharmacists can be helpful in counseling families and promoting caution in considering the use of unproven therapy. Information can be found on websites for national associations. For example, in early 1999, when the GI hormone secretin was in the news for treating autism, the Autism Society of America and the Autism Research Institute provided solid information from experts, putting new or possible therapies in perspective.

Psychotropic Medication Tips

People with developmental disabilities have the same mental illnesses seen in the general population, such as major depression, bipolar disorder, anxiety disorders, attention-deficit-hyperactivity, and psychotic disorders.

Not all problem behaviors require treatment with psychotropic medications. Some behaviors are the result of feelings like grief or sadness that are normal events for all people. A person with a developmental disability experiences these same feelings but may not be able to adequately communicate what is bothering them.

When working with individuals with developmental disabilities, remember the ABC's:

ANTECEDENT: Always look for an antecedent. There may have been an event that triggered the behavior that appears to be problematic. Look for things in the environment such as new teachers, helpers, and roommates. Caregivers and family can check for simple things like clothing that doesn't fit, changes in wheelchairs, sunburn, bee stings, or injuries.

BEHAVIOR: Identify the behavior itself, and if possible, try to witness the behavior, if it is one that occurs frequently. Caregivers and family can provide important feedback here.

CONSEQUENCE: Consider that the consequence of the behavior, as maladaptive as it may be, could be a cry for attention or a means of having the individual's needs met. In such a situation, giving a medication may be perceived as a punishment for attempting to communicate. Behavioral treatment plans may be very effective in decreasing some problem behaviors.

Remember that in many cases individuals with developmental disorders have diminished cognitive ability and may use rather childish means for getting attention or what they want. Never assume that a person who has a developmental disability does not hear well or is unable to communicate. Pharmacists are responsible to use their skills and experience and do all they can to understand the patient's needs.

Diagnosing a mental illness in a person with mental retardation or another developmental disability can be a challenge. A reasonable approach is to develop a list of problem behaviors and communicate these to the patient's physician or other qualified health professional. The most common reasons for referral to psychiatric services are self-injurious behavior, aggression, or self-stimulatory behaviors. Medications should be used to treat a specific cluster of target symptoms. The treatment plan must include monitoring these target symptoms, so that response to a medication can be assessed. Families, caregivers, and patients need to know the treatment plan and what to expect from medications.

Pharmacists who desire to learn more about developmental disabilities can access the NADD website at *www.thenadd.org*. NADD is a national, nonprofit organization whose goals include bridging the gap for individuals with mental illness and mental retardation. At this site you can become a member, learn about upcoming conferences, find educational resources, and be directed to related links.

Conclusion

Assisting children or adults with developmental disabilities is a wonderful opportunity for the pharmacist to provide cognitive services and pharmaceutical care to a group of people who are underserved by the health care community.

Most individuals with developmental disabilities living in the community have seen many physicians over the years. Their medical records are not necessarily found in one medical practitioner's office, but many of these same individuals have their prescriptions filled in just one pharmacy.

Pharmacists can provide a wealth of services to people with developmental disabilities and offer them a healthier life through the appropriate, careful, and rational use of medications for their medical conditions.

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LEARNING OBJECTIVES:

After reading these articles, you should be able to:

1. List the five diagnoses used to establish eligibility for services and support within the California Regional Center System and state their relative frequency.
2. Differentiate between the most common causes of mental retardation.
3. Recognize potential barriers to effective care for people with cognitive and/or physical impairments.
4. Identify medication issues that are common in people with developmental disabilities.
5. Discuss the appropriate use of psychotropic medications in people with developmental disabilities.
6. Give examples of "people first" communication as part of the patient consultation process.
7. Describe the pharmacist's role in the care of people with developmental disabilities.

DIRECTIONS FOR OBTAINING CE:

- Read the articles and take the test to receive three (3) hours of continuing education credit. Return the entire test page and include a self-addressed, stamped envelope. Please note that the passing grade is 70%. If necessary, one test will be readministered.
- Type or print your name, address, and license number in the space provided on the form below.
- This continuing education monograph is jointly sponsored by CSHP and CPhA, and you may return your answer sheet to EITHER:

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California Society of Health-System Pharmacists
CE Exam
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Sacramento, CA 95816

PLEASE NOTE: It is not necessary to send the exam to both organizations. Select one. Remember to enclose a self-addressed, stamped envelope.

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Please type or print neatly:

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How long did it take you to complete the reading and the test? _____ minutes

Additional comments are appreciated: _____

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CONTINUING EDUCATION CERTIFICATE OF COMPLETION

Date: _____ Course Name: The Care of Children & Adults with Developmental Disabilities. ACPE Universal Program #126-113-99-078-HO4.

This is to certify that the above-mentioned continuing education course was completed by:

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The California Pharmacists Association Educational Foundation and the California Society of Health-System Pharmacists are approved by the American Council on Pharmaceutical Education as providers of continuing pharmaceutical education. This course provides three hours of credit, Universal Program #126-113-99-078-HO4. Pharmacists completing this course prior to April 1, 2003 may receive credit.

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25. _____

TEST QUESTIONS

1) The number of Californians with developmental disabilities is:
   a) 15,500
   b) 155,000
   c) 300,000
   d) 1.5 million

2) The majority of Californians with developmental disabilities:
   a) live in developmental centers
   b) live in a community care facility, an intermediate care facility, or skilled nursing facility
   c) live with family members
   d) receive independent living or supported living services in the community
   e) live in other places, such as foster family homes and county homes

3) The categories of etiology for mental retardation include:
   a) Chromosomal
   b) Brain malformation
   c) Environmental
   d) All of the above

4) Children with Down syndrome are prone to recurrent respiratory infections.
   a) True
   b) False

5) Which of the following syndromes is associated with an extreme obsessive-compulsive eating disorder?
   a) Angelman
   b) Klinefelter
   c) Prader-Willi
   d) Noonan

6) Mental retardation occurs in what percent of the population?
   a) 1%
   b) 3%
   c) 6%
   d) 9%

7) At least 25% of infants who present with infantile spasm seizures will have tuberous sclerosis.
   a) True
   b) False

8) Barriers to effective care for the cognitively impaired include:
   a) They may be unable to communicate.
   b) They may feel threatened by unfamiliar clinicians.
   c) They may resist physical examination.
   d) All of the above.

9) Meeting the individual with developmental disabilities is an important first step in the communication process.
   a) True
   b) False

10) Which of the following statements would be the best to use to determine that an individual understands information about his or her medications?
   a) "Do you understand what I just discussed with you?"
   b) "Please tell me, in your own words, what you understand of what I just told you."

11) Which of the following are ways that pharmacists can help clients and caregivers improve compliance with multiple drug therapy regimens:
   a) Color-code prescription bottles to indicate time of day or the number of times per day to take medications
   b) Glue pictures of the pills to the medicine box
   c) Create a chart with other symbols or stickers – such as a fork and plate for meals, moon or stars for bedtime
   d) All of the above

12) Small body stature, certain genetic abnormalities, and poor eyesight are examples of factors that can affect the choice of drug or dosage form for individuals with developmental disabilities.
   a) True
   b) False

13) It is not uncommon for individuals with developmental disabilities to have multiple medical problems, such as seizure disorders, malabsorption syndromes or other gastrointestinal conditions, cardiovascular abnormalities, skeletal malformations, or autonomic nervous system dysregulation.
   a) True
   b) False

14) Drug-drug interactions in individuals with developmental disabilities should be handled differently than they are handled in individuals within the general population.
   a) True
   b) False

15) The approximate percentage of individuals with developmental disabilities who have psychiatric disorders and are living in the community or in residential settings is:
   a) 10-20%
   b) 20-30%
   c) 30-40%
   d) 40-50%

16) Which of the following is NOT an example of how a pharmacist can improve the quality of care for the individual with developmental disabilities?
   a) Taking a complete medication history and sharing that information with caregivers, family, and the appropriate physicians
   b) Recommending changes in the medication regimen, such as stopping medications that are duplicative, or replacing medications that are causing problematic side effects with more appropriate choices
   c) Assuming that the individual can not hear you when conversing with family or caregivers
   d) Visiting the patient at home

17) A Medication Administration Record (MAR) is a document used for all of the following, except:
   a) to record the drug administered and the individual who received the medication
   b) to track inventory of medications
   c) to record the dose administered
   d) to record the time the drug was administered
   e) to record the route of administration

18) The key to appropriate selection of psychotropic medications in people with developmental disabilities is accurate assessment of the psychiatric disorder or behavior problem.
   a) True
   b) False

19) Psychotropic drugs are only used for persons with developmental disabilities to control or manage "maladaptive" behaviors such as aggression, hyperactivity, self-injury and disruptive or inappropriate social behavior.
   a) True
   b) False

20) Drugs indicated for the treatment of obsessive-compulsive disorders (OCD) in persons with developmental disabilities include:
   a) Clomipramine, fluoxetine, fluvoxamine, and paroxetine
   b) Buspirone, lithium, benzodiazepines, and carbamazepine
   c) Clozapine, risperidone, and olanzepine
   d) Thioridazine, chlorpromazine, loxapine, and clozapine

21) Which of the following groups of medications may lower the seizure threshold in individuals with a seizure disorder?
   a) Fluoxetine, fluvoxamine, and paroxetine
   b) Buspirone, benzodiazepines, and naltrexone
   c) Carbamazepine, valproic acid and lithium
   d) Thioridazine, chlorpromazine, and clozapine

22) The following medications have been used to treat aggression and self-injurious behavior (SIB):
   a) Clozapine, risperidone, and olanzepine
   b) Naltrexone
   c) Propanolol, metoprolol, and nadolol
   d) Antidepressants, anxiolytics, and anticonvulsants
   e) All of the above

23) Lithium is most effective in people who have cyclical behavior symptoms and:
   a) Has not been used extensively in persons with developmental disabilities
   b) Should be administered to achieve serum concentrations of 10mEq/L to 15mEq/L
   c) Should be administered for at least six to eight weeks at adequate serum concentrations (0.5 mEq/L to 1 mEq/L) before concluding that a person will not respond.
   d) Has very few side effects

24) The selective serotonin uptake inhibitors (SSRIs) have been used to treat people with developmental disabilities who:
   a) Have symptoms of depression
   b) Exhibit aggressive behaviors
   c) Have symptoms of anxiety disorders, such as obsessive-compulsive disorder (OCD)
   d) a & c

25) The use of psychotropic medications in persons with developmental disabilities has been thoroughly investigated with well-controlled, methodologically sound studies.
   a) True
   b) False

HEALTH NOTES
Care of Children & Adults with Developmental Disabilities

Making a Difference

This issue of *HEALTH NOTES* is a collaborative effort of the **California State Board of Pharmacy** and the **California Health and Human Services Agency**. We would like to thank **Grantland Johnson**, Secretary, Health and Human Services Agency, **Cliff Allenby**, Director, Department of Developmental Services, and **Patricia Harris**, Executive Director, Board of Pharmacy, for their assistance with this joint project.

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Continuing Education

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