The Face of Intellectual Disability

Segment I – Discovering Patterns

What Is Intellectual Disability?

Intellectual disability is a developmental disability in which an individual has difficulty learning and using daily life skills. As a result, development and learning are slower than average, and do not reach the levels of higher order reasoning that are found in typically developing individuals. Intellectual disability occurs during the developmental period before age 18, and the extent of impairment ranges from mild to profound.

An individual is considered to have an intellectual disability if he or she meets the following three criteria:

- Significant limitations in general intellectual functioning (Intelligence Quotient [IQ] is below 70-75, using a comprehensive individual intelligence test).
- There are significant limitations in two or more adaptive skill areas (such as self-care, daily living skills, functional communication, or social skills).
- The condition is present before the age of 18.

A psychological assessment using at least one reliable standardized intelligence test determines intellectual functioning. This means that the assessment is conducted in a standardized manner, using an evaluation tool which is administered the same way each time, by a well-trained professional, in a culturally sensitive manner.

These tests assess verbal comprehension, problem-solving and expression, non-verbal thinking and perception, attention and concentration, planning and memory. Results are expressed as intelligence quotients (or IQ scores), and determine the level of intellectual disability. There are four levels:

- **Mild** – Intelligence test scores are 50-55 to 70, accompanied by limitations in multiple adaptive skill areas
- **Moderate** – Test scores range from 35-40 to 50-55, along with related limitations in multiple adaptive skill areas
- **Severe** – Test scores are 20-25 to 35-40, along with related limitations in multiple adaptive skill areas
- **Profound** – Scores fall below 20-25, along with related limitations in multiple adaptive skill areas

What Does Intellectual Disability Look Like?

A person with an intellectual disability may demonstrate different levels of ability or different

degrees of impairment in various areas of functioning. While intellectual disability is a condition that is expected to continue lifelong, individuals with an intellectual disability continue to learn and develop throughout childhood and adulthood.

An individual with an intellectual disability has limited intellectual functioning, along with limitations in what we call adaptive skills. Adaptive skills are those areas of development that we need to live, work and play in the community. They include such things as:

- Communication
- Self-care
- Home living
- Social skills
- Leisure
- Health and safety
- Self-direction
- Functional academics (reading, writing, basic math)
- Work

While an individual may have limitations in certain cognitive and social adaptive areas, he or she may simultaneously have strengths in other adaptive skills. Given appropriate help and support, these individuals will generally improve their skills, function successfully in the community, and lead rewarding, productive lives.

It's important to note that a person who has limits in intellectual functioning, but who does not have limits in adaptive skill areas, may not be considered to be substantially disabled, and cannot receive a diagnosis of intellectual disability.

The Diagnostic Detective

There are basically three steps in diagnosing a person as having an intellectual disability:

1. Assessment of the individual's ability on one or more standardized intelligence tests and a standardized adaptive skills test.

2. A description of the individual's strengths and challenges in four areas:

    - Intellectual and adaptive behavior
    - Psychological/emotional
    - Physical/health
    - Environmental considerations

    This is usually accomplished through a combination of formal testing, observation, talking with key people in the person's life, and interacting with the individual.

    The purpose of this detective work is to determine whether factors other than primary developmental disability are either responsible for below average cognitive scores, and/or are further compromising or exacerbating the primary disability.

3. A determination of what supports are needed in these four areas, as well as the assignment of a level of intensity: intermittent, limited, extensive, pervasive.

Segment II – Class Dis-Mythed…Focus on Facts

What Intellectual disability Is Not

Because intellectual disability manifests in many ways, a number of myths and misconceptions have grown up around the disorder.

Let's Dis-myth Them

- Intellectual disability is not a developmental delay – it is not simply a need to "catch up" to a certain level of functioning. The term developmental delay is used for infants and very young children for whom early intervention may be effective. After age three, the diagnosis of intellectual disability is given only when functioning has been diagnosed as significantly below average, and is anticipated to permanently remain as significantly below average (though many adaptive abilities may continue to improve).

- Intellectual disability is not a learning disability. The term learning disability implies that an individual has difficulty in one type of ability, with general intelligence that is average or above, while a diagnosis of intellectual disability means that the individual functions significantly below average across several areas of capabilities, including the cognitive area (especially general reasoning ability) and at least two adaptive areas.

- Intellectual disability is not a mental illness, and it is not associated with any particular type of personality. Persons with mental illness commonly have intelligence that is average or above. However, an individual with intellectual disability may also develop mental illness, or emotional problems due to difficulty with communication skills, and exposure to environmental stressors such as isolation, loneliness and discrimination.

- Intellectual disability is not a disease. In most cases, the conceptual model for defining intellectual disability is functional rather than medical. Intellectual disability may occur as the result of a specific cause or medical condition, such as Down syndrome, but the specific cause does not alone determine the limits of the individual's destiny, or ability to function.

It's important to realize that someone with intellectual disability may not necessarily function in a way that meets the common stereotype of intellectual disability. There are many faces of intellectual disability. As with all disabilities – the person is an individual and needs to be treated as such.

Segment III – Exploring the Unknown

What Are the Causes?

Intellectual disability can be caused by any condition, which impairs development of the brain before birth, during birth, or in the developmental period of life, which spans the childhood years.

Its causes are grouped as follows:

Genetic Conditions

Intellectual disability, which has been caused by a genetic condition, begins at the time of conception. These conditions result from abnormal genes inherited from parents, errors when genes combine, or other genetic disorders caused during pregnancy by infections, overexposure to x-rays, and other factors. More than 500 genetic diseases are associated with intellectual disability. Among them are: PKU (phenylketonuria), a single gene disorder caused by a defective enzyme; Down syndrome, the result of a chromosome disorder; and Fragile X syndrome, a single gene disorder located on the X chromosome.

Down syndrome is caused by the presence of an extra chromosome 21, which results in distinctive physical characteristics and limited intellectual ability. This most common type of Down syndrome is also sometimes referred to as Trisomy 21. The estimated incidence of Down syndrome is 1 in 800-1,100 births. Down syndrome is usually diagnosed at birth or shortly thereafter. However, it can be identified between the 16th and 20th weeks of pregnancy through an amniocentesis.

Each year approximately 3,000-5,000 children are born with this disorder, and about 250,000 families in the United States are affected. Down syndrome is one of the most common clinical causes of intellectual disability in the world – it is not related to race, nationality, religion or socio-economic status.

Although women over the age of 35 are more likely to give birth to a baby with Down syndrome, more than 80% of the children with Down syndrome are born to women under 35.

Children with Down syndrome are often smaller, and their physical and mental development is slower. They walk a little later – between 15-26 months – than those who develop more typically. Language development is also delayed.

There is a wide variation in mental abilities, behavior and physical development. Some function cognitively in the borderline to low average range, and others may have severe intellectual disabilities. However, the majority function in the mild to moderate range of intellectual disability.

Some other common conditions associated with Down syndrome may include hearing deficits, congenital heart disease, congenital abnormalities of the gastrointestinal system,

eye problems and thyroid dysfunction.

Some theories on cause include hormonal abnormalities, X-rays, viral infections, immunologic problems, and genetic predisposition.

Pre-natal testing can diagnose Down syndrome, thereby allowing the family to obtain information about the condition for planning and preparation. Genetic testing may also be useful to determine whether Down syndrome is likely to recur within the family, if either parent is a carrier of the translocation chromosome.

Early detection allows for early intervention so that physical, occupational and speech therapies can be started at the youngest possible age. Throughout an individual's life, services such as appropriate education, vocational training, job placement, and social and recreational programs can help him or her achieve independence.

Fragile X syndrome is an inherited genetic condition. It involves a break, or weakness, on the long arm of the X chromosome.

The abnormal X chromosome can be transmitted from parent to child. The term "ex-linked" or "X-linked" inheritance is used in the medical literature. This means that mothers are carriers of this chromosome, and their sons are at risk of being affected, while daughters are at risk of being carriers and sometimes mildly affected. While more boys than girls are affected by Fragile X, it is not transmitted from father to son.

Fragile X is the most common inherited cause of intellectual disability known to exist, and may account for up to 10% of intellectual disabilities. Prenatal testing can determine whether a fetus is affected by this condition, and the degree of intellectual disability which can be expected.

Affected persons may have features such as large ears, a prominent jaw, and a long face. Older males may have large testicles. There is currently no cure for Fragile X, but a variety of medications can improve attention span, concentration, hyperactivity, aggressive behavior and other problems.

Problems during pregnancy

Prenatal exposure to certain substances during pregnancy, including drugs, alcohol, or cigarettes, can lead to damage of the developing brain, and to lifelong developmental disabilities. Other risks for the pregnant mother and the development of her baby include malnutrition, certain environmental contaminants, and exposure to certain diseases such as toxoplasmosis, cytomegalovirus, rubella and syphilis.

One of the most preventable causes of intellectual disability is Fetal Alcohol Syndrome, or Fetal Alcohol Effects (FAS/FAE).

Fetal Alcohol Syndrome (FAS) – is a pattern of malformations and disabilities resulting from when a pregnant woman drinks heavily during her pregnancy.

Women who drink during pregnancy may experience spontaneous abortions, miscarriages or stillbirths.

The children who are born with Fetal Alcohol Syndrome often experience:

- Low birth weight, birth length and small head circumference
- Increased birth defects and developmental delays
- Decreased IQ
- Central nervous system involvement – intellectual disability, learning disabilities
- Distinctive, unusual facial patterns
- Failure to thrive
- Poor wake and sleep patterns
- Hyperactivity, distractibility and attention deficits
- Impulsiveness, temper tantrums

Fetal Alcohol Effects (FAE) is a condition where children are born with many of the same physical, behavioral and psycho-social characteristics as those with FAS, but to a lesser degree. Many individuals with FAE, while looking quite normal, have significant deficits in their intellectual, behavioral and social abilities which prevent them from leading normal lives.

Problems at Birth

Any birth condition of unusual stress may injure the infant's brain, particularly prematurity and low birth weight.

Problems After Birth and During Childhood

Intellectual disability can occur as the result of some trauma or damage to the brain during infancy or childhood. Childhood diseases such as whooping cough, chicken pox, measles and Hib disease can damage the brain, as can accidents such as a blow to the head or near drowning. Lead, mercury and other environmental toxins can cause irreparable damage to the brain and nervous system as well.

Poverty and cultural deprivation – Children in poor families may become mentally retarded because of malnutrition, disease-producing conditions, inadequate medical care, and environmental health hazards.

What Is the Incidence?

Intellectual disability cuts across the lines of racial, ethnic, education, social and economic backgrounds. It can occur in any family.

According to the U.S. Department of Health and Human Services, “the usual national percentages are estimated to be 1% (which usually includes all or most persons currently receiving services in the “MR/ID” Service System), based on best estimates from various authorities in the field.”

A study by the Centers for Disease Control and Prevention (CDC) and the Health Resources and Services Administration (HRSA), "Trends in the Prevalence of Developmental Disabilities

in U.S. Children, 1997–2008," found that the prevalence of ***any*** type of developmental disability, (including learning disabilities, attention deficit disorder, autism, and other developmental delay) was 13.7%. Of this total, the prevalence of "other developmental delay" was 3.65%.

Can It Be Prevented?

Yes. Many of the known causes of intellectual disability are preventable. These include prematurity, prenatal exposure to drugs and alcohol, genetic conditions, environmental toxins, infections to the brain, and traumatic head injury.

Early comprehensive prenatal care and preventive measures prior to and during pregnancy increase a woman's chances of preventing intellectual disability. Early intervention for infants with established high risks can prevent developmental delays from becoming lifelong developmental disabilities, or can minimize the effects. Education and prevention related to infectious diseases, traumatic brain injuries, and exposure to drugs and alcohol can play an important role in preventing intellectual disabilities, as can proper use of child safety seats and bicycle helmets. Intellectual disabilities can also be diminished by removing lead or other toxins from the environment.

There has been significant progress in research over the last 30 years or so. For example, each year in the United States we prevent more than 10,000 instances of intellectual disability through the use of effective prenatal and infant screening, childhood vaccinations, and other measures. For example, intellectual disabilities which would have occurred in the past as a result of PKU or hypothyroidism can now be effectively prevented through routine screening of newborns, and timely implementation of dietary or hormone treatment. Intellectual disabilities which in the past were the result of measles encephalitis, rubella, or Hib disease are now preventable through the use of vaccines.

What Are the Effects?

Just as in a typically developing population, the effects of intellectual disability vary considerably among individuals – and the ability levels can be very different.

About 87% of those with intellectual disabilities will have a mild form of the disability, while the remaining 13% or so are within the moderate, severe, or profound range of impairment. For individuals with mild intellectual disability, learning does occur more slowly. When provided with appropriate opportunities for development, learning, and inclusion in regular day-to-day community life, however, these individuals can benefit greatly. Adults with mild intellectual disability may need limited amounts of support and can achieve a good degree of independence.

Individuals who are in the moderate, severe, or profound range of intellectual disability will demonstrate more significant limitations in cognitive skills – that is, their ability to learn new information and skills - and in their adaptive skills- such as the ability to communicate, socialize, and perform activities of daily living. However, these individuals will also benefit from appropriate early intervention, special education, and appropriate lifelong supports, to allow them to develop the highest level of independence and quality of life.

Segment IV – Reaching Beyond Syndromes to Treatment

The individual with intellectual disability can be highly responsive to his or her physical and social environment. People with Down syndrome in particular have a high social quotient, which helps them to adapt successfully into society. The person with intellectual disability also has the same emotions and needs as the typically developing person.

With appropriate intervention and support over a sustained period, the functional abilities of a person with intellectual disability generally improve. This requires education to improve existing skills, as well as support or accommodation in their environment.

What Are Some Typical Treatments?

A caring and enriching home environment, early intervention and focused education efforts will help everyone with intellectual disabilities. Treatment is determined by an individualized program of special education including speech therapy, physical therapy and vocational preparation.

Today, early intervention programs, pre-schools and integrated special education strategies have demonstrated that youngsters with intellectual disabilities can participate in many learning experiences which positively influence their overall functioning. Research has shown that early intervention, environmental enrichment and assistance to families results in remarkable progress for infants who have such educational and stimulating experiences.

Like all children, children with intellectual disabilities can benefit from sensory stimulation, specific exercises involving gross and fine motor activities, and instruction in cognitive development. Also, preschool experiences play an important role in the young child's life, since exploring the environment beyond the home enables the child to participate in a broader world.

Segment V – Ask the Experts

What Research Is Being Conducted?

Research has increasingly made us aware of measures to take before and during pregnancy to prevent prematurity.

Recent advances in molecular biology and the successful mapping of the human genome now make it feasible to examine the genetic basis for Down syndrome and other forms of genetic intellectual disability. This, in turn, may lead to a medical therapy.

Genetic and prenatal testing techniques have made it possible for prediction and prenatal diagnosis of conditions, which in turn allows parents more time to obtain information and counseling, and to plan accordingly.

Research is also being pursued to promote surgical intervention while an effected fetus is

still in the womb, which may prevent intellectual disability. For example, surgical intervention is being explored for fetal hydrocephalus diagnosed during pregnancy, in the hopes of decreasing the possibility of related cognitive impairment.

Segment VI – Understanding People, Exploring Possibilities

While much remains to be done, there is continual progress in helping children and adults with intellectual disability. Today, people with intellectual disabilities hold jobs, live independently, and enjoy recreational opportunities in their communities. They also date, socialize and form ongoing relationships, including marrying.

More each year continue their education. Many are becoming actively involved in volunteerism, self advocacy, and leadership roles, to enhance awareness, understanding, and services for themselves and others with intellectual disabilities.