

How to Provide a Safe and Stimulating Classroom Environment for Students with Down Syndrome-A Medical and Educational Perspective

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Down Syndrome

- ❖ Dr. John Langdon Down described the syndrome in 1866.
- ❖ Diagnosis was made clinically until 1959
- ❖ The chromosome abnormality was discovered in 1959.
- ❖ Down syndrome is one of the first symptom complexes associated with mental retardation to be identified as a syndrome.

Down Syndrome

Overview

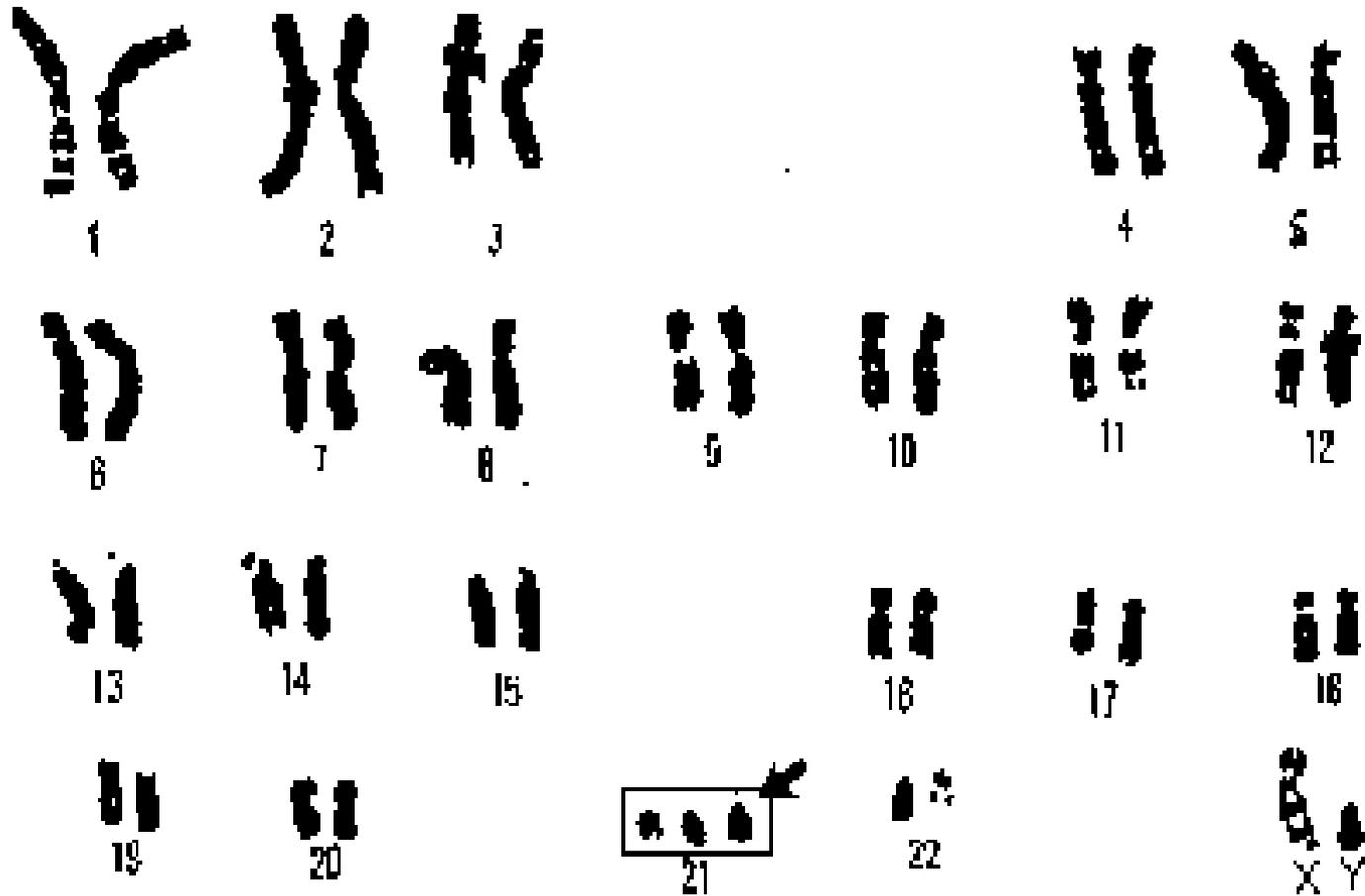
- ❖ It is the most common pattern of human malformation with an incidence in the general population of 1 in 800-1000 live births.
- ❖ Over 350,000 people in the United States have Down syndrome.
- ❖ A high correlation exists between increasing maternal age and the presence of an extra chromosome in the offspring.
- ❖ Therefore, the expected rate of Down syndrome in a woman 20 years of age is 1 in 1,925 compared to an expected rate of Down syndrome in a woman 45 years of age of 1 in 20.

Down Syndrome

Genetics

- ❖ All persons with Down syndrome have duplicated chromosome 21 material
- ❖ The origin of the duplicated genetic material can vary
- ❖ **Nondisjunction** (about 96% of cases)
 - ♦ The extra chromosome may arise by abnormal sorting to produce an extra, free-standing chromosome (trisomy 21)
- ❖ **Translocation** (3 % of cases)
 - ♦ a joining of chromosomes to produce extra chromosome 21 material that is attached to another chromosome
- ❖ **Mosaicism** (1-2%)
 - ♦ Patients with mixtures of normal and trisomic cells (mosaic Down syndrome) often have milder phenotypes.
 - ♦ Percentages of normal cells within the blood sample used for chromosome studies may differ from the percentages of normal cells in other tissues like brain or heart.

Trisomy 21 Karyotype



- ❖ Extensive mapping of genes on this "critical region" of chromosome 21 is under way as part of the human genome initiative.
- ❖ The mechanisms by which increased dosage of these genes leads to the Down syndrome phenotype are as yet unknown.

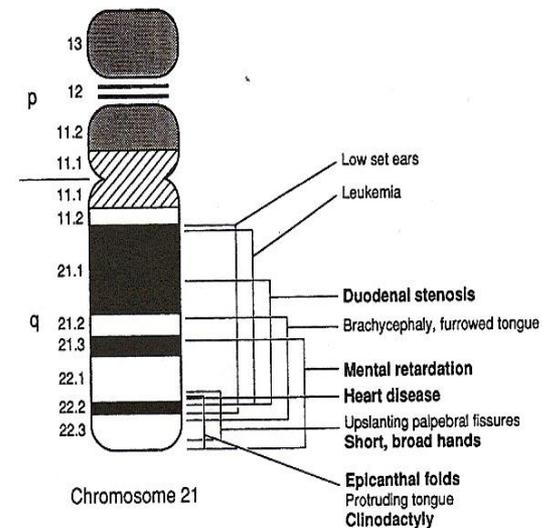


Figure 17.2. The "critical region" for the Down syndrome phenotype is on the long arm (q11.1-q22.3) of chromosome #21. Specific malformations are noted as being localized to the bracketed regions. The most clear associations are bolded. (From Epstein, C.E. [Ed.]. [1991, January 17-18]. *The morphogenesis of Down syndrome [Proceedings of the National Down Syndrome Society Conference on Morphogenesis and Down Syndrome, New York]* [p. 44]. Copyright © 1991 John Wiley & Sons; adapted by permission.)

Clinical Features

- ❖ Extra and loose neck skin
- ❖ Single creases on the palms
- ❖ Clinodactyly (curved fifth finger)
- ❖ Broad space between the first and second toes
- ❖ A deep plantar crease
- ❖ The tongue often protrudes, more because of low muscle tone than true enlargement.



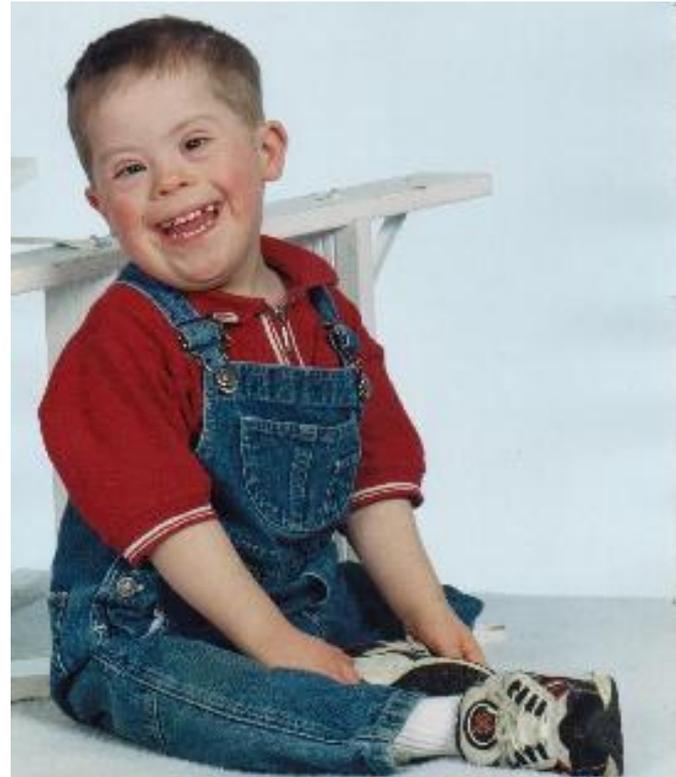
Clinical Features

- ❖ As with all syndromes, the pattern of minor and major defects in Down syndrome varies from individual to individual.
- ❖ None of the anomalies taken alone are specific or diagnostic for Down syndrome.
- ❖ When Down syndrome is suspected, a chromosome study (karyotype) will confirm or exclude the diagnosis.



Clinical Features

- ❖ Central hair whorl (cowlick)
- ❖ Flat occiput (back of the head)
- ❖ Upslanting eyes
- ❖ Epicanthal folds (folds around the corner of the eye)
- ❖ White spots in the iris of the eye (Brushfield spots)
- ❖ Upturned nose



Related Conditions

- ❖ Congenital Heart Disease
- ❖ Seizures
- ❖ Eye Anomalies
- ❖ Hearing Loss
- ❖ Genito-Urinary Tract Anomalies
- ❖ Respiratory Disease
- ❖ Obesity
- ❖ Sleep Apnea
- ❖ Developmental Delays
- ❖ Blood Disease
- ❖ Endocrine Disease
- ❖ Blood Disorders
- ❖ Alzheimer's Disease

Heart Defects

40% of individuals with Down syndrome have some form of congenital heart defect

- ❖ AV (atrioventricular) Septal defect
- ❖ Ventricular Septal Defect (VSD)
- ❖ Patent Ductus Arteriosus (PDA)
- ❖ Atrial Septal Defect (ASD)
- ❖ Tetralogy of Fallot
- ❖ Aberrant Subclavian Artery
- ❖ Pulmonary Hypertension
- ❖ Adolescents and young adults can develop heart valve dysfunction even when they have had no history of congenital heart problems.
- ❖ SBE prophylaxis may be necessary.

Ophthalmologic Features

- ❖ Brushfield spots
(speckling of the iris)
- ❖ Fine Lens Opacities
- ❖ Nystagmus
- ❖ Strabismus
- ❖ Keratoconus
- ❖ Ptosis
- ❖ Cataracts
- ❖ Astigmatism
- ❖ Hyperopia (far sightedness)
- ❖ Myopia (near sightedness)
- ❖ Blepharitis
- ❖ Tear Duct Obstruction

Ophthalmologic Features

Brushfield Spots



Keratoconus



Hearing Loss

- ❖ Can be conductive, sensorineural or both
- ❖ Can be unilateral or bilateral
- ❖ Often undiagnosed
 - ◆ Change in learning or behavior-consider hearing screen
- ❖ Etiology can be multifactorial
 - ◆ Narrow posterior throat structures
 - ◆ Subtle immune deficiency resulting in recurrent ear infections

Gastrointestinal Disease

- ❖ Pyloric stenosis
- ❖ Tracheo-esophageal fistula
- ❖ Esophageal atresia (obstruction of the esophagus)
- ❖ Duodenal atresia (obstruction of the duodenum).
- ❖ Gastroesophageal Reflux (GERD)
- ❖ Constipation
- ❖ Hirschprung's disease
- ❖ Celiac disease
- ❖ Imperforate Anus
- ❖ Neonatal Liver Disease (rare)

Respiratory Disorders

- ❖ Otitis media
- ❖ Sinusitis
- ❖ Pneumonia
- ❖ Sleep apnea
- ❖ Dysphagia/oromotor dysfunction
- ❖ Aspiration
- ❖ Pulmonary Hypertension

Dental Diseases

- ❖ Microdontias
- ❖ Missing Teeth/Fused Teeth
- ❖ Delayed tooth eruption (1-2 years later than average)
- ❖ Malocclusions
- ❖ Periodontal disease
 - ◆ Gingivitis leading to loss of Alveolar bone
- ❖ Dental Caries occur with lower prevalence than in the general population

Genitourinary Tract Anomalies

- ❖ Micropenis
- ❖ Cryptorchidism
- ❖ Infertility
- ❖ Cystitis/Urinary Tract Infections
- ❖ Renal Anomalies
- ❖ Wilm's Tumor

Endocrine Disorders

- ❖ Congenital hypothyroidism
- ❖ Thyroid Disease
- ❖ Diabetes
- ❖ Failure to Thrive- in infancy and early childhood
- ❖ Obesity
- ❖ Short Stature
- ❖ Lipid abnormalities

Blood Disorders

- ❖ Immune Dysfunction

- ❖ Myeloid Proliferation

- ❖ Leukemia

- ◆ 1 in 150 in children with Down syndrome compared with 1 in 2800 in children at large

- ❖ Platelet Disorders

- ❖ White blood cell impairment

- ❖ Erythrocytosis (secondary to chronic hypoxia)

Skin Conditions

Most related to Immune dysfunction/Inflammatory response

- ❖ Atopic Dermatitis (eczema)
- ❖ Seborrheic Dermatitis
- ❖ Vitiligo
- ❖ Cheilitis
- ❖ Ichthyosis
- ❖ Xerosis
- ❖ Alopecia Areata
- ❖ Syringomas
- ❖ Onychomycosis

The Neurology of Down Syndrome

The nervous system is always affected in Down syndrome.

- ❖ Developmental Delays
- ❖ Hypotonia
- ❖ Atlantoaxial instability
 - ◆ Symptomatic -vs- Asymptomatic
- ❖ Seizures
- ❖ Mental Health Disorders
- ❖ Alzheimer's Disease

Brain pathology

- ❖ Slightly smaller brain size for age.
- ❖ Shorter diameter for the anterior-posterior brain measurement.
- ❖ An unusually steep slope to the posterior portions of the brain.
- ❖ Insufficiently developed superior temporal gyrus.
- ❖ It is not known in what way these features contribute to the developmental disabilities of Down syndrome.

Brain Pathology

- ❖ Post-mortem brain studies show that virtually all persons over 50 with Down syndrome have pathological plaques and neurofibrillary tangles- the hallmarks of Alzheimer's disease, However only 15-20% of individuals with Down syndrome have clinical signs of the disorder
- ❖ The prevalence of Alzheimer's is much more prevalent than in the general population.
- ❖ It is important to rule out treatable causes of decline in mental functioning (thyroid problems, B vitamin deficiencies, vision and hearing problems, depression, sleep apnea, polypharmacy, etc.) before jumping to the conclusion that an individual has Alzheimer's.

Neurodevelopmental and behavior Impairments

- ❖ Aggressive Behavior (7%)
- ❖ ADHD (6%)
- ❖ Conduct/Oppositional Disorder (5%)
- ❖ Anxiety Disorders (5%)
- ❖ Self Injurious Behavior (1%)
- ❖ Autism (1%)

Seizures

- ❖ Seizures occur in 6% of individuals with Down syndrome
- ❖ Most common seizure type is generalized tonic clonic
- ❖ Age of onset typically bimodal distribution
 - ◆ Onset before age 3
 - ◆ Onset after age 13

Neurologic

- ❖ Language Delays

- ❖ Receptive language tends to be better than expressive

- ❖ Central Hypotonia

- ❖ Gross motor Delays

- ❖ Oral motor dysfunction

- ❖ Poor oral motor coordination
 - ❖ Decreased sensory awareness

- Most people with Down syndrome have some level of intellectual disability.
- The level usually falls into the mild to moderate range.
- Not indicative of the many strengths and talents that each individual possesses.
- Children with Down syndrome learn to sit, walk, talk, play, toilet train and do most other activities only somewhat later than their peers without down syndrome.

- Early intervention services, which begin shortly after birth, help children with Down syndrome develop to their full potential.
- Quality educational programs, along with a stimulating home environment and good medical care enable people with Down syndrome to become contributing members of their families and communities.

Early Intervention Works!

Just a decade ago, the prognosis for these children was not as bright as it is today.

Early Intervention programs from birth to age 3 have shown impressive results.

Families receive training in how to help their children learn to maximize the development that occurs in the early years.

In addition, many children receive:

- ♦ PT
- ♦ OT
- ♦ Speech Therapy
- ♦ Aqua therapy
- ♦ Hippotherapy
- ♦ Music therapy
- ♦ Infant education individually and in groups

Inclusion

- ❖ Studies have demonstrated social, academic, and behavioral benefits for students with disabilities who are placed in inclusive settings, without negatively impacting the educational experience of the other students. (Becker, Dumas, and Roberts ,2000)
- ❖ Many children with Down syndrome, if they were in the special education or self-contained classroom, would not have the same amount of language development as their non-disabled peers.
- ❖ "The learning characteristics of students with Down Syndrome are more similar to their regular education peers than they are different.
- ❖ However, language and motivational deficiencies may necessitate more highly structured, sequenced activities, with smaller bits of information presented at a time, and lots of rewards and praise built into the design of the lesson" (Wolpert, 2001).

Learning strengths

- ❖ Strong short-term visual memory
- ❖ High social/interpersonal intelligence

Learning weaknesses

- ❖ Poor short-term auditory memory
- ❖ Difficulty with basic math skills

Short Term Auditory Memory

“Working Memory”

“Verbal Memory”

- ❖ It relates to the speed with which we can hold process understand and assimilate language
- ❖ It relates directly with the speed with which we can articulate words, influences the speed at which children learn new words and learn to read.

- ❖ Short-term auditory processing impairment occurs almost universally in people with Down syndrome of all ages.
- ❖ Visual tools that support the short-term processing of concepts are amongst the most useful resources any school can implement to support students with Down syndrome and their teachers.

Make Learning Visual

- ❖ Accompany keywords and checklists with pictures or symbols.
- ❖ Use keywords icons or diagrams rather than full text.
- ❖ Underline and highlight key words.

Communication through

- sign language,
- picture cards and
- assistive communication devices

promotes cognitive and language development even when children lack the oral-motor skills to speak.

Teaching Strategies

- **Short attention spans** are also prevalent among students with Down's. Direct instruction in short periods of time along with smaller chunks of activities will help to support learning. Introducing new material slowly, sequentially and in a step by step fashion will help to ensure maximum learning occurs.
- **Distractibility:** Down's students are often easily distracted. You'll need to employ strategies that work to minimize distractions such as keeping the student away from the window, using a slightly more structured environment, keeping the noise level down and having an orderly classroom where students are free from surprises and know what your expectations, routines and rules are.
- **Speech and Language:** Down's students all suffer from serious problems such as hearing difficulties and articulation problems. Sometimes they will require speech/language intervention and a great deal of direct instruction. In some cases, augmentative or facilitated communication will be a good alternative for communication. Use patience and model appropriate interactions at all times.
- **Behavior management techniques** used for other students should not differ for the student with Down's Syndrome. Again, positive reinforcement is a much better method than anything punitive. Reinforcers need to be meaningful.
- **Social Skills Training** -Appropriate Boundaries and Sexuality training should start in Elementary grades

Things to keep in mind

- ❖ When behavior changes, escalates or new behaviors develop, think about pain or undiagnosed/untreated medical conditions first.
- ❖ Make sure the routine is consistent and predictable.
- ❖ Make sure accommodations for special communication needs are being addressed.

Services Available

- Continuum of Care Project 925-2350
- NM SAFE Program 272-0285
- LINC-Library Information Network Center
272-3000
- CDD-Center for Development and
Disability 272-3000
- PRO-Parents Reaching Out 247-0192
- DDSD-DOH 841-5000

