

## Health Topics

- Alzheimer's Disease
- Associated Conditions
- Atlantoaxial Instability
- Blood Diseases
- Cosmetic Surgery
- Dental Care
- Dental-Periodontal Disease
- Dimethyl Sulfoxide
- DS Clinics and UCEDDs
- Dual Diagnosis
- Ear, Nose, Throat
- Early Intervention
- Endocrine
- Growth Charts
- Healthcare Guidelines
- Healthcare Guidelines - How to Use Them
- Heart
- Interferon and Down Syndrome
- Neurology
- Physical Therapy
- Piracetam
- Sexuality
- Speech
- Thyroid
- Vision & Hearing
- Vitamin Related Therapies
- Your Baby and DS

## Health Programs

## Neurology

By: Dr. Ira Lott Professor

Neurology, the study of the human nervous system and its disorders, is important in the clinical care treatment of a person with Down syndrome. The nervous system is always affected in Down syndrome; among the most common involvements of the nervous system are developmental disabilities (retardation in mental development and motor capabilities), hypotonia, atlantoaxial dislocation and seizures.

### What does the term "developmental disabilities" mean in relation to Down syndrome?

This term refers to incomplete development of the brain, which leads to both mental retardation and to slowed and/or incomplete mastery of physical coordination.

### What is known about brain pathology in regard to developmental difficulties?

Pathology in the Down syndrome brain includes a slightly smaller brain size for age, a shorter diameter for the anterior-posterior brain measurement, an unusually steep slope to the posterior portions of the brain and an insufficiently developed superior temporal gyrus. It is not known in what way these features contribute to the developmental disabilities of Down syndrome.

### Has any progress been made in the pathology of the Down syndrome brain through research?

Research in this area is being actively pursued, especially since it may yield clues to Alzheimer's disease and to the neuroscientific effects on learning and memory.

### Is IQ (Intelligence Quotient) a meaningful measure in Down syndrome?

No. IQ is not an adequate measure of the functional status of people with Down syndrome. For example, individuals with

SEARCH

Donate

Shop

- Changing Lives Partners
- Changing Lives

Down syndrome may have difficulty with grammar, but understand individual words, or have a sense of when to speak, at a par with their mental age.

---

### **What can be done to help people with Down syndrome to achieve their full potential in the face of such disabilities?**

A variety of intervention programs, designed to begin in infancy and continue throughout learning years, help children with Down syndrome maximize their capabilities.

### **What is hypotonia?**

Hypotonia refers to the reduced muscle tone that occurs in virtually all infants with Down syndrome. It is commonly seen in the flexor group, muscles which act to flex a joint. The muscle weakness ranges from mild to moderate to severe.

### **How is hypotonia diagnosed?**

Common hypotonic symptoms are a lagging head when the child is pulled into a sitting position and arching of the back when carried upright or lying on the stomach. In addition, hypotonic children will tire more easily and adopt movement patterns requiring the least expenditure of energy.

### **Are there any effective interventions for hypotonia?**

Hypotonic conditions tend to improve with age. Gross motor programs offered by occupational and physical therapists have been shown to diminish hypotonic symptoms. These programs improve large body movements, such as walking, turning, sitting, standing and climbing stairs. Enrollment of children in an early intervention program which includes gross motor programs is recommended at the earliest feasible age, generally between four and six weeks of age.

### **What is atlantoaxial dislocation?**

Atlantoaxial dislocation refers to a problem caused by hypotonia. The ligaments at the first two cervical vertebra are more relaxed than they should be, putting the individual at risk of spinal cord compression and injury.

### **How common is atlantoaxial dislocation?**

The cervical spine instability occurs in 10 to 20 percent of children with Down syndrome; however, actual spinal compression is rare.

### **What are the symptoms of atlantoaxial dislocation?**

If symptoms are present, they include neck pain, change in gait, onset of weakness in the extremities, spasticity, limited

neck movement and bowel/bladder incontinence (particularly after toilet training has been accomplished). However, most children with x-ray evidence of atlantoaxial dislocation have no apparent symptoms.

#### **What preventive measures should be taken?**

Since most children with atlantoaxial dislocation do not exhibit symptoms, lateral cervical x-rays are recommended for all children with Down syndrome after age two. If the instability is present, but there are no symptoms, the appropriate precaution is to limit "high risk" activities which might over-stress the neck. These activities include high-jumping, diving, gymnastics, trampoline, and butterfly strokes in swimming. To devise a safe exercise program for a symptom-free child with atlantoaxial dislocation, consult your doctor.

#### **Is there any treatment for atlantoaxial dislocation?**

If symptoms are present, the atlantoaxial joint can be surgically fused.

#### **What is the incidence of seizures in Down syndrome?**

In the young child with Down syndrome, seizures are no more common than in the general population. Beginning at age 20-30, the incidence of seizure disorder rises substantially in the Down syndrome population. Research is ongoing as to whether the frequencies of seizures in individuals with Down syndrome is related to the aging of the brain.

#### **What is the typical seizure for a person with Down syndrome?**

Seizures for individuals with Down syndrome commonly look like epileptic seizures: jerking of arms and legs and loss of consciousness. Seizures can also take a mixed form, with staring spells and momentary lapses of attention.

#### **What is the treatment for seizures?**

Seizures can be controlled with standard anti-convulsant medication.

#### **Summary:**

- The most common involvements of the nervous system in Down syndrome are hypotonia, atlantoaxial dislocation and seizures.
- The reduced muscle tone of hypotonia affects most infants with Down syndrome and ranges from mild to severe.
- Hypotonic conditions improve with age; however, gross motor movement programs provided by physical and occupational therapists have been proven useful in

diminishing the symptoms of hypotonia.

- Atlantoaxial dislocation refers to a weakness of neck muscles that affects approximately 10 to 20 percent of children with Down syndrome. Most cases are asymptomatic. Preventive measures against neck injuries call for lateral cervical x-rays for all children with Down syndrome after age two. If atlantoaxial dislocation is present, high-risk activities such as jumping, diving, etc., should be limited.
- In cases of atlantoaxial dislocation where symptoms are present, the atlantoaxial joint can be surgically fused.
- Seizures are equally common in the child with Down syndrome as compared to the general same-age population. After age 20, the incidence of seizures in the Down syndrome population increases substantially as compared to the general population. Seizures can be controlled by standard anti-convulsant medication.

### BOOKS & NEWSLETTERS

"The Central Nervous System" In: *Medical Care in Down Syndrome: A Preventive Medicine Approach*. Rogers, P. and Coleman, M. (1992). New York: Marcel Dekker, Inc., pp. 201-224. Available through Marcel Dekker, Inc., 270 Madison Ave., 4th Floor, New York, NY 10016; tel.: (212) 696-9000.

This chapter provides medical information about the brain's structure and function in individuals with Down syndrome. Specific topics discussed include sleep disturbances, seizure disorders, behavior disorders, aging and Alzheimer's disease.

"223 Neurologic Abnormalities," Florez, J. In: *Biomedical Concerns in Persons with Down Syndrome*. Pueschel, S. and Pueschel, J., (Eds.) (1992). Baltimore, MD: Paul H. Brookes Publishing Co., pp. 159-173. Available through Paul H. Brookes Publishing Co., P.O. Box 10624, Baltimore, MD 21285-0624; tel.: (800) 638-3775.

This chapter focuses on information about morphologic alterations, neurochemical alterations and neurophysiologic alterations. Also discussed are brain activity related to learning and language and epilepsy and seizures in individuals with Down syndrome.

"Neurological and Neurobehavioral Disorders in Down Syndrome," Lott, I. In: *Down Syndrome: Advances in Medical Care*. Lott, I., and McCoy, E. (Eds.) (1992). New York, Wiley-Liss, Inc., pp. 103-109. Available through Wiley-Liss, 1 Wiley Dr., Somerset, NJ 08875; tel.: (800) 225-5945.

This chapter explains the brain function in individuals with Down syndrome. Seizure disorders, hypotonia, atlantoaxial instability and neurobehavioral aspects of Down syndrome are discussed briefly.

"Neurology of Children with Down Syndrome," Mattheis, P. In: Medical & Surgical Care for Children with Down Syndrome: A Guide for Parents. Van Dyke, D., Mattheis, P., Eberly, S., Williams, J. (Eds.) (1995). Bethesda, MD: Woodbine House, Inc., pp. 267-287. Available through Woodbine House, Inc., 6510 Bells Mill Rd., Bethesda, MD 20817; tel.: (800) 843-7323.

This chapter discusses the ways in which Down syndrome affects the nervous system, including differences in thought, speech, learning and muscle control. The author provides an overview of some of the neurologic conditions which occur more frequently in individuals with Down syndrome and some of the available treatment alternatives.

The Neurology of Down Syndrome, Lott, I. (1995). New York: National Down Syndrome Society. Society. Available through the National Down Syndrome Society, 666 Broadway, New York, NY 10012; tel.: (800) 221-4602 or (212) 460-9330.

This booklet answers questions about how the nervous system is affected in individuals with Down syndrome. The most common impacts, including developmental disabilities, hypotonia, atlantoaxial dislocation and seizures, are discussed.

---

About the Author: Pediatrics University of California at Irvine