

## RETT'S DISORDER



### DEFINITION:

Rett's disorder, also known as Rett's syndrome, is a severe degenerative condition diagnosed by deceleration of head growth, loss of previously acquired hand skills, loss of interest in the social environment, appearance of stereotyped hand movements and gait and coordination problems, and subsequent development of severe impairments in language and psychomotor function. Rett's disorder has been reported only in females and is estimated to occur in 1 in 10,000 to 1 in 15,000 female births.

### CAUSE:

The cause of Rett's disorder is not known. However, because only females are affected, it is believed to be a result of a dominant mutation on a Y-linked gene.

### DIAGNOSTIC CRITERIA:

Each of the following:

- Apparently normal prenatal and perinatal development
- Apparently normal psychomotor development through the first 5 to 6 months
- Normal head circumference at birth

Onset of each of the following occurs after the period of normal development:

- Deceleration of head growth between ages 5 months and 4 years
- Loss of previously acquired purposeful hand skills between ages 5 and 30 months and the subsequent development of stereotyping hand movements
- Early loss of social interaction
- Appearance of poorly coordinated gait or trunk movements between ages 1 and 4 years of age
- Severely impaired expressive and receptive language development with severe psychomotor retardation

### TEACHING TIPS AND SUGGESTED ACTIVITIES:

- Develop a highly structured environment.
- Use the same organization and equipment each day.
- Use proper safety equipment at all times.
- Teach safety often and be very specific.

- Obtain the student's full attention.
- Eliminate unnecessary external stimuli.
- Limit the amount of relevant stimuli presented at one time.
- Utilize sensory stimulation to increase attention span and decrease self-stimulation.
- Use vigorous aerobic exercise to reduce self-stimulatory and off task behaviors.
- Parents and professionals should use the same rules, expectations, and behavior management systems including positive reinforcement of desired behaviors.
- Utilize the Premack principle (pairing something liked with something disliked).
- Teach in a game-like environment to facilitate generalization.
- Improve motor skill acquisition by using reinforcement, task analysis, and physical Prompting.
- Describe, discuss, and/or role play difficult situations.
- Use transitions that depict activities between each activity.

\*\* Refer to the fact sheets on MENTAL RETARDATION and AUTISM for possible signs, symptoms, and characteristics and more helpful hints and suggested activities.

*Information on this sheet contains only suggested guidelines. Each student must be considered individually, and in many cases, a physician's written consent should be obtained.*