

MOBILITY AND GROSS MOTOR ABILITIES: THE ROLE OF PHYSICAL THERAPY

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A wobbly gait or stance is often the first feature of A-T to be recognized as abnormal. Children with A-T typically walk at a normal age, but the instability of early walking does not improve with age. Many children with A-T are mis-diagnosed as having cerebral palsy, which then predicts that motor skills will remain delayed or slowly improve. The physical therapist may be the first to document worsening motor skills, and thus can be instrumental in initiating the re-evaluation that leads to the correct diagnosis. This early importance of the physical therapist continues throughout the life of the child with A-T, as progression of the disease brings new challenges to physical function.

Physical therapists are dedicated to helping with difficulties in gross motor ability, including problems with walking, mobility and posture. For most patients and problems, physical therapists work closely with occupational therapists, who direct their attention to functions of the arms, hands, head and neck. As A-T worsens, changes in neurologic impairment, age, maturity, environment, personal goals and family needs will necessarily change and physical therapy needs will change with them.

The Physical Therapy Evaluation

A physical therapy evaluation includes assessment of muscle tone, involuntary movement, compensations, strength, range of motion, access to automatic movement reactions (such as balance reactions), and functional ability in different environments. The potential benefit from adaptive equipment or orthotics (braces) is considered in consultation with occupational therapists and physicians. Evaluation of current fitness activities is also an important component of the physical therapy evaluation. Whenever possible, objective measurements of motor performance should be made to document status. Unfortunately, this may be difficult in children with A-T because commonly used pediatric standards are not well designed to assess their special problems. Some useful parameters may include number of steps out of specified length and width path, time able to stand still within a confined space (such as 12 inch square), and time to complete a transfer into or out of wheelchair).

General Findings

Children with A-T stand and walk in an unusual fashion. Standing quietly in one place is often difficult, with frequent stagger reactions to maintain

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- √ muscle tone
- √ involuntary
- √ movement
- √ compensations
- √ strength
- √ range of motion
- √ access to automatic movement reactions
- √ functional ability

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balance. Most children with A-T begin walking at a normal age, but their walk never attains a normal fluid motion. They wobble and fall more often than expected for age. The gait may appear to be propelled, leaning forward over the toes with each step. Instead of striking the floor first with the heel, children may step forward onto a flat foot with a “stomping” gait, or step forward toe first. There may be difficulty with clearing one foot around the other, or clearing the floor, with each step. There are differences in the particulars of gait abnormalities between different children. Sometimes the foot is rotated in (pigeon toed), sometimes the foot is bent inward at the ankle-walking on the outside of the foot, and in some the foot is turned outward or excessively flat-footed. Muscle tone is usually loose in the hips and knees, but stiff at the feet and ankles. Often, children with A-T are described as “running everywhere” and using stagger reactions to maintain balance. The gait in A-T may be complicated by other abnormalities of tone, or extra movements that distort walking in unusual ways. There may be abnormal tone or sustained abnormal posture (dystonia) or other abnormality that is a form of adventitious movement. (See chapter 4) As the children begin to fall more frequently, safety becomes a concern, and there is an increase in the rate of minor injuries.

Over time children with AT discover that they are better able to use their arms for play activities if they sit in reverse tailor fashion (“W sitting”) on the floor. This serves to widen their base of support and thereby improve their stability. Some children don’t find the “W” sitting position comfortable, and may achieve the same stability by sitting “cross-legged”. Either of these sitting postures are acceptable and neither should be discouraged out of concern for maldevelopment of the bones or joints.

With time, children are less able to cope with increasing impairment. It can be so tiring to constantly work against instability that fatigue limits overall function. Fatigue is a significant problem in A-T even at younger ages when it may be manifest in more subtle ways. Fatigue is responsible for variability of performance, whether gross motor, fine motor control, speech or academic. Sometimes the “good days” and “bad days” are unpredictable, but all parents know that time of day, inadequate rest, illness and other challenges present special problems for children and adults with A-T.

Orthopedic deformities may develop, and appear to be related to both central (brain) and peripheral (nerves in the body) nervous system disease. The most common deformities occur at the foot and ankle. The foot tends to point toes downward with restriction of ankle joint mobility. The ankle may deform as well, most commonly becoming restricted to an inward position. These foot deformities can cause skin breakdown and pain with walking. The distorted posture makes it even more difficult to stand and walk. Most individuals with A-T will have sufficient muscle power for many decades to allow them to walk short distances around the home or bear weight with transfers if someone assists them. Painful deformity of the ankle may prevent this. Deformity of the foot may be more easily delayed or prevented than corrected.

Appropriate braces may slow the progression of deformity, and may give added stability for walking in some cases. Operative tendon transfers by an orthopedic surgeon may be necessary to balance muscular forces around the ankle, and may also improve walking.

There is a low incidence of spinal curvature (scoliosis) seen in the A-T population, but the rate of occurrence is greater than what is found in the general population of children and teenagers. The cause is not clear, but may be due to unequal muscle tone on one side of the body compared to the opposite side. Once an early curvature is detected, the child should be referred to an orthopedist experienced in treating children and adults with neurologic impairment.

Physical Therapy Recommendations

Braces

Braces or Orthotics are devices that are applied to the body to improve function or maintain proper alignment and range of motion. Braces that apply to the back of the calf and under the foot (known as an AFO, for Ankle-Foot Orthoses) may help to improve walking, particularly if there is a tendency to roll outward or inward over an unstable ankle. These braces can be rigid, or can be fashioned with a hinge at the ankle that allows for up and downward movement of the ankle (an articulated AFO) while it restricts sideways deflection of the ankle. AFOs can be worn for either or both of two purposes. First, an AFO can be worn during the daytime to help with the gait. It may be worthwhile to minimize tripping due to toe walking or difficulty with foot clearance from the floor as the leg swing forward, or help provide stability at the ankle to minimize turning the ankle with an errant step. If this is the indication for the AFO, the best way to assess whether it works is to try it! Children will know if they feel more secure, and a reduction in falls, improvement in gait and reduced fatigue with walking can be evidence that a brace is working. Other persons, who do not walk much, nonetheless appreciate the stability of an AFO while sitting. Again, the indication is comfort, and whether it works should be evident.

A second indication for an AFO is to slow the progression of deformity. Usually there is benefit for the gait as well, though this may not always be the case. The brace will resist the deforming forces of weight bearing and abnormal tone around the foot and ankle. It is not possible to improve existing foot deformity with a brace; such a brace would be intolerable due to pain. Whether or not an AFO is successful in slowing the progression of deformity is a decision for medical evaluation. Braces for either of these purposes may be fashioned by physical therapists or may be made by special brace shops depending upon local circumstances.

How often and how long the braces are worn each day depends on the reason for which they are being used. If the purpose is to help improve

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functional activities such as walking and transferring, they will work only when worn. If the purpose is to slow the development of deformity, the schedule of wearing should be determined by the physician who prescribed them. In some cases, the benefit of stretching can be achieved with night-time wearing only.

Sometimes it is possible to achieve some of the benefit of an AFO by proper choice of shoes. Boots can give additional support to the ankles, preventing deflection of the ankle off to one side with an errant step. They may be fashionable, however, and certainly for some children this matters! An AFO can be hidden fairly effectively inside the pant leg and shoes, for those children who are concerned about appearance. One curious fact about shoes is that sticky "Air-Jordan" like soles may not be better for the individual with ataxia. If there is a tendency to stumble because of catching a foot on the floor as it passes to the next step, a sticky sole may make this worse. Instead, sometimes shoes with smooth soles (old Keds, or slick leather soles) may be less likely to cause a trip-up. Everyone tends to walk more carefully with a smooth soled shoe; this caution may be better than the false security of sticky soled shoes that grab and trip with errant steps.

Only a few patients with A-T develop scoliosis. The rate and amount of curvature of the spine, and the nature of the forces that appear to be driving it to one side, determines the best therapy. In some cases, use of a TLSO (for thoraco-lumbar-sacral-orthoses, or stiff body jacket) is indicated. Referral to a specialty center that cares for children with spinal deformity is essential for any individual with a curvature greater than 30 degrees.

Assistive Devices

The child's ability to be independent with mobility within his or her peer group is always of paramount importance. Fatigue may be the most important reason to introduce an assistive device. Safety, energy efficiency, and the level of physical independence and its psychological implications should be constantly monitored to assist with selection of the appropriate devices.

Use of a walker is indicated when falling increases or the child is too tired to participate as a result of walking to an activity. The Kaye Products 4 wheel posture walker with rear wheel ratcheting (which works as a brake protecting against staggering or falling backward) is the walker of choice for most children with A-T. Those children who have fairly stable trunk control and don't stagger from side to side are often successful for at least some period of time or in certain environments utilizing the "swivel front wheel option".

As the child grows older, distance and speed requirements increase and fatigue becomes limiting even with the use of a walker. When that happens, power mobility should be considered. This may be indicated for children in certain environmental circumstances as early as age 4 years. Power mobility is initially utilized for travelling long distances in the community. For example, children who are still walking at home and at school may use a power scooter

or power wheelchair at the mall or on class trips. This focused use of power mobility will promote independence, and does *not* lead to increased weakness or loss of skills. Once power mobility is under consideration, an evaluation should be undertaken by an assistive technology team experienced in caring for children with neurologic impairments and abnormalities of muscle tone. There is an ever expanding plethora of power adaptive equipment. An assistive technology team can recommend the most appropriate control mechanism and electronics package, seating system, and power mobility base that meet the child's needs. For example, electronic packages should be selected to offer the option of customizing such parameters as acceleration, torque, tuning speed, forward and reverse speed, braking and sensitivity (tremor dampening). Whenever it becomes feasible under the durable medical provision of the child's health care insurance, a lightweight manual wheelchair with adaptive seating system should be obtained as a back up to the power mobility base.

Children are very practical in their approach to mobility, and this should be encouraged to optimize their independence. There is no one right way to move around. Their abilities, the environment and the demands of the situation tend to dictate the safest and most efficient means of mobility for a given child at a given point in time. For example, the same child may be scooting or crawling for very short distances to get across a room, while at the same time walking around the house balancing on walls and furniture, walking with a posture walker at school, and using a power wheelchair outdoors for longer distances.

Children tend to like and readily accept anything that helps them be more independent and do things more easily and safely. Parents relate that their child's reaction to beginning the use of power mobility is almost always very positive. Children with A-T see the wheelchair as a device that liberates them from the difficult task of getting from one point to another, and enabling them to more easily participate in activities and keep up with family and friends. In fact, the children with A-T are usually more accepting of the introduction of assistive devices than their family members. It is in the child's best interest to introduce assistive devices in a timely fashion so that function and quality of life are optimized. Even if the family home is inaccessible or the family cannot transport a power mobility system, it is helpful for the child to utilize power mobility at school and on class trips.

Compensations and Therapeutic Activities

Once independent ambulation with assistive devices is no longer feasible, the family should be trained in the safest method of assisted walking for short distances and stairs, as needed in the home. Transfer techniques need to be monitored and adjusted as indicated over time with the overall goal of maximum independence, safety and efficiency. It is critical throughout life to maintain the ability of the A-T patient to accept full weight in supported standing so that it is rarely if ever necessary for a caretaker to physically lift the person.

A-T patients, just like everyone else, need a fitness and recreation program to promote aerobic fitness and to maintain optimal strength, endurance and function.

Potential recreational activities might include:

- √ swimming
- √ horseback riding
- √ stationary bicycle
- √ rowing machine
- √ weight lifting
- √ aerobics done in a sitting position.

In general, it is recommended that people of all ages be involved in some type of aerobic exercise that is recreationally appealing. A-T patients, just like everyone else, need a fitness and recreation program to promote aerobic fitness and to maintain optimal strength, endurance and function. The type of activity will obviously vary with age, interest, physical ability and available resources. Young children who are constantly moving tend to keep themselves fit through spontaneous play. In older children with A-T, this becomes more of a challenge. They can participate in adapted physical education classes at school. Other potential recreational activities might include swimming with appropriate flotation devices, adapted horseback riding, use of adapted exercise equipment such as a stationary cycle or rowing machine, weight lifting and aerobics done in a sitting position. Reclining cycles with back support, seat belt, and toe loops tend to be safe and fun. During all fitness activities, it is critical to avoid exercising to the point of fatigue.

Home and Environmental Modifications

The environment should be accessible to allow maximum participation and independence. Families who are planning to build or remodel an existing home should make certain that they are collaborating with a contractor/architect who is knowledgeable about “universal design”. Physical and occupational therapists should be able to recommend appropriate consultants.

Guidelines for accessible buildings can be easily accessed through the internet.

- The “Accessibility Guidelines for Buildings and Facilities” contains detailed information and building codes (such as the acceptable grade for ramps): www.access-board.gov/bfdg/adaag.htm
- The “Do-Able Renewable Home” website is designed specifically for the geriatric population, however much of the information is applicable to all persons with physical impairments: www.usc.edu/dept/gero/hmap/library/drhome/
- A paper entitled “Universal Design in Housing” published in 1998 can be obtained by writing to Ronald L. Mace, F.A.I.A. at the Center for Universal Design, School of Design, North Carolina State University, Box 8613, Raleigh, NC 27695-8613.