



Physical Therapy and Occupational Therapy in Progeria

Information for Families and Caretakers from The Progeria Research Foundation

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Physical and Occupational Therapy in Progeria

Hutchinson-Gilford Progeria Syndrome (“Progeria”, or “HGPS”) is a rare, fatal genetic condition characterized by an appearance of accelerated aging in children. Children with Progeria need Physical Therapy (PT) and Occupational Therapy (OT) as often as possible (optimally 2- 3 times each per week) to ensure maximum range of motion and optimal daily functioning throughout their lives. Services can be provided in a traditional medical setting or a school setting depending on eligibility criteria in individual states. The role of the PT/OT in the school setting is to maintain range of motion, strength, and functional status so a student can access the school building and playground and participate with peers in classroom activities, at recess, and in Physical Education class. Proactive PT and OT are important, since all children with Progeria develop restrictions in range of motion in a progressive manner. Bony abnormalities are almost always evident in X-rays by the age of 2 years. Range of motion may be restricted for several reasons. Children with HGPS experience progressive joint contractures, primarily in the knees, ankles, and fingers due to tendonous abnormalities. They also experience hip abnormalities due primarily to progressive coxa valga and shoulder restrictions due to clavicular resorption. Finally, tightened skin can restrict range of motion. Skin tightening can be almost absent in some children, or can be severe and restrict chest wall motion and gastric capacity in others.

Each regimen should be tailored to the child's individual needs, and tailored according to cardiac status. It is appropriate to have oxygen available at all times (usually kept in the school nurse's office). The child may need oxygen in case of angina, cyanosis, dyspnea, or if the child simply cannot catch his/her breath in the normal way. Medical personnel will be able to assess the need for further treatment.

Common protocols for PT and OT include but are not limited to the following exercises. Tracking progress through regular joint range of motion measurements is advised at least every 3-4 months. Due to the orthopedic conditions commonly seen in the hip and shoulder, range of motion in these joints should be closely monitored. Tightness is also seen in the heel cords, low back muscles, finger flexors, and triceps muscles. To maintain range of motion, a combination of myofascial release techniques followed by more traditional passive, active, and active-assisted stretching exercises have been found to be effective. Due to the possibility of weakened joint integrity due to coxa valga in the hips and clavicular resorption in the shoulders, it is advisable to avoid passive stretching in these joints and instead focus on active stretching. Weight bearing activities in hands and knees are helpful for stretching finger flexors. To help maintain range of motion, traditional stretching can be followed by functional activities like scooter board games, reaching activities on the big ball, and ball skills like throwing and catching to help make therapy sessions more enjoyable.

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Strengthening activities should target core strengthening for the hips and abdominals with activities such as sit-ups, bridges, and leg lifts. Again strengthening can be done traditionally as well as through developmental sequence activities, scooter board games, tricycle riding, obstacle courses, and ball skills.

Due to orthopedic deformities as well as tendonous, muscular and skin tightness, gait deviations may occur. It is advisable to focus on maintaining heel cord flexibility and hip internal rotation to minimize gait deviations. Heel cord flexibility can be addressed through a runner's stretch, stretching off a stair, and heel walking short distances. Hip rotation can be addressed through mat exercises as well as tandem walking activities on a line or balance beam. Traditional gait training with verbal cueing and the use of mirrors can also be helpful.

Depending on a child's medical status, participation in Physical Education class and recess activities is encouraged. The Physical Education teacher and classroom teacher should be made aware of any precautions so modifications can be done on an ongoing basis. Therapists should emphasize the importance of warm-up activities and shoulder, arm, and leg stretches at the beginning of class to help decrease the incidence of muscle pulls in class activities.

Depending on a child's medical status, therapy sessions should include the practice of age appropriate gross motor skills such as running, jumping, climbing, and pedaling a tricycle or small bicycle as well as ball skills to help these children better participate with age level peers in the school setting.

Swimming activities are highly recommended, although they are usually not available in the school setting. Since children with Progeria have very little body fat, water temperature should be on the warmer side, and layers or insulated suits are advisable. Several Progeria families have used "warm belly wetsuits" at www.warmbelly.com. Other sources of wetsuits may be as appropriate or more appropriate. Pros and cons: warm belly wetsuits have flexible fit due to velcro straps, but have a tank top design which does not cover any part of the arms. Other wetsuits have arm coverage but tend to be higher priced. PRF does not endorse any particular company or source of clothing.

As with any child with therapy needs, therapists should have ongoing communication with families to be made aware of any medical conditions and resultant precautions and the implications regarding therapy treatment. With creative strategies, therapy sessions should be made fun to help these children maximize their participation and level of benefit. Ongoing consultation should occur with both the teachers and the family to maximize carryover. An emphasis is placed on compliance with a home exercise program for stretching on a regular basis and this should be encouraged early on in the treatment relationship.

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