13. Occupational Therapy (OT)

Generally, occupational therapy (OT) promotes health with a focus on fine motor skills. Occupational and physical therapists often work together for optimal whole body treatment.

Please also refer to *Living with Progeria*, section 17, for additional advice on physical adaptations from parents of children with Progeria.

As joint contractures progress, children use alternative methods or assistance devices to perform activities such as putting on socks. This helps to maintain independence.

Evaluation

Children with Progeria should have yearly assessments by a pediatric occupational therapist. The evaluation should include the following areas:

- Physical measures (range of motion, strength)
- Coordination
- Functional skills
- Visual perceptual
- Visual motor integration skills

There have been no studies on the effectiveness of occupational therapy interventions with this population and the recommendations in this handbook are based on clinical observations and discussion with the patients and their health care providers. Any sudden change in range of motion, hand strength, or ability to participate in functional activities should be evaluated by a physician even if there is no traumatic event.

Physical findings

Physical findings vary markedly within age groups and age spans among children with Progeria. Body functions and structures that affect upper extremity use and functional activities often include the following:

- Joint contractures of all upper extremity joints
- Upper extremity asymmetries
- Prone to shoulder dislocations
- Reduced upper extremity strength
- Wrists typically have limited dorsiflexion (bending upward)
- Some children's thumbs do not go into carpometalcarpal (CMC) extension plane
- Most children's thumbs are used with the thumb against the distal interphalangeal joint of the index finger (the joint closest to the tip of the finger)
- On occasion hyperextension of the thumbs' interphalangeal joints (joint closest to the tip of the finger) is seen
- Metecarpalphalangeal joints most often have limited flexion (joints closest to the hand)
- Distal and proximal interphalangeal joints (the middle joint and the joint closest to the tip of the finger) tend to have flexion contractures
- Resorption of the distal phalangeals

Maximum finger extension in a child with Progeria

Small size, difficulty with supination, contracted finers, lack of fat, and prominent veins in a child with Progeria (below) compared with an age-matched chid without Progeria (above)

- Distal phalangeals are often painful with pressure
- Decreased fat deposits within the hand (most notably at the thumb and finger tips)
- Short in stature
- Increased bony prominences
- Difficulty tolerating extreme hot or cold temperatures (i.e., weather, water)
- Some have decreased fine motor coordination
- Some have visual perceptual and visual motor integration deficits

Areas of occupational therapy include self-care, education, work, play, leisure, and social participation. Children with Progeria have a very large array of activities that they enjoy participating in. They do have some difficulty performing some tasks and there are a few patterns that were noted and reviewed below. The limitations appear in relationship to the child's physical findings from their occupational, physical, and medical examinations. Participation in functional activities requires a skilled therapist who should fully probe to ascertain what the child can do.

The following sections review common areas of occupation in which these children have difficulty and/or limitations, and offer some intervention strategies to increase their participation:

Self-care

> Dressing

Children with Progeria often have difficulty with lower extremity dressing (putting on shoes, socks, and pants below the knees).This appears to be related to lower extremity joint contractures. Some children also have difficulty with mastering fasteners as quickly as other children their age. Reasons for this include limited exposure to fasteners due to the style of clothing they wear, cultural/parenting style, decreased strength, and coordination. Children with Progeria often need assistance with the lower extremity dressing. They often develop adaptive dressing strategies such as positional changes or the use of adaptive equipment such as reachers that can help them to be independent with donning lower extremity clothing. A sock aid can be used to put on socks, while a long-handled shoe horn may assist with putting on their shoes independently.

> Hygiene

Most children with Progeria are independent with age-appropriate hygiene by the age of 4 or 5; however, they require some environmental adaptations to assist with height obstacles and with what appears to be postural instability (hesitant on step stool). In the bathroom, stools should be placed at the toilet and the sink. Parents may assist or supervise when they are getting in and out of the tub or shower due to safety concerns. Rarely do the children require adaptive equipment to assist with hygiene tasks such as bathing. However, equipment such as long handle sponges may be used to assist with lower extremity washing. Some children are not able to wipe themselves after toileting due to range of motion limitations and difficulty with balance. Aides such as long handled tongs (tongs with toilet paper wrapped around them) or wet wipes to decrease the amount of wiping can be helpful. Toilet seat inserts may increase the child's comfort due to the child's size and difficulty with balance. Padded toilet seats may also be used to address discomfort with prolonged sitting due to increased bony prominences. With grooming or oral hygiene, an electric or battery-operated toothbrush may be used as the children may fatigue with brushing due to decreased strength and range of motion limitations. Flossing sticks and automated hands-free toothpaste dispensers may also be helpful. Please refer to *Dental Recommendations*, section 9, for further information on tooth hygiene. Although it is important for the children to participate in brushing their own teeth, it is recommended that this activity be supervised and parents assist to ensure optimal hygiene.

> Feeding

Children with Progeria become independent self-feeders. Early signs of decreased motor coordination or the effects of joint limitations can be noted during feeding with a utensil but do not generally interrupt food intake. Use of a rocker knife may assist some children with cutting. Children with reduced hand strength or coordination often find a straight knife, such as the Amefa straight knife, very helpful and parents seem to feel safe with the use of this knife.

> Meal preparation and eating

Children with Progeria often have limited participation with basic meal preparation as compared to age-matched peers. This may be due to height limitations and parenting style. Some families have arranged a section where snack items can be at a height the child can reach. Snacks should be removed from original packaging and placed in easily opened containers.

Modifications can also be made to allow children to pour their own drinks, as standard drink containers are typically too heavy and are difficult to grasp due to range of motion limitations. These modifications include placing drinks in a small partially-filled container with a spout. Stool(s) placed in the kitchen also allow for access to counter tops and the sink. If the child is starting to cook and there are difficulties, seek out an OT assessment for further assistance with bowl and pan holders, electric peelers, and other cooking aides. Adapted seats such as tripp-trapp or right-height chairs with additional foot plates allow the children to sit at the dinner table with their families.

Encourage your child's independence by removing snacks from their original packaging and placing in easy-to-open containers, placing stools in the kitchen, and having adaptive kitchen utensils on-hand.

> House management

Some children have difficulty managing basic home functions due to height limitations. Recommendations include adapted light switches with hanging strings or plastic devices, adapted door knobs (due to difficulty with hand positioning and strength to open the door independently), and automatic doors, which may also assist with children getting out of their house in case of an emergency.

Education

> Positioning

Children often complain of pain while sitting for prolonged periods of time, which appears to be related to their bony prominences. Seat cushions and frequent rest breaks, allowing them to stand if needed, are recommended. Chairs within the classroom setting should allow them to be at standard seat high with their feet supported. The use of chairs such as a tripp-trapp or right-height chair, with an additional foot plate to allow them to get in and out of the chair safely, are also recommended. These special chairs are important as they allow for the child to be an active participant and socialize with their peers within the classroom. Being at the same height as their peers also allows them to visually scan the classroom and see the chalkboard or whiteboard.

> Handwriting

Children with Progeria often complain of hand fatigue or pain during writing or coloring activities. The reasons for this are unclear, but appears to be related to joint limitations, reduced fatty pads, and the functional position of the carpometacarpal thumb joint (which remains fixed in mid abduction or extension) and their limited wrist positioning (neutral to slight palmar

Children with Progeria can successfully meet the demands of the school day with some accommodation in the areas of seating, classroom tools, and lunch room considerations.

flexion). Some parents report reduced motor control during handwriting. Others report difficulty with mastering writing. In most of the children, this appears to be a result of abnormal wrist and hand positioning and decreased strength rather than visual perceptual, visual motor integrative, and/or fine motor incoordination. OT intervention often helps children with Progeria master handwriting, with improved motor control. Children can benefit from an individualized strengthening program, including stretching exercises and activities to enhance in-hand manipulation skills along with dexterity skills. Some children also benefit from using unique crayons and pencils that are shorter and narrower, to assist with the structure of their hands and their decreased strength. Padded pencil grips or padded pens may be used to decrease the amount of finger pain that is often experienced from the pressure of the writing utensil, due to the lack of fat deposits in fingertips. The use of a vertical surface is recommended to improve wrist dorsiflexion (the ability to bend backwards) and strength. Slant boards should only be used at the recommendation of a therapist after full evaluation, due to possible contraindications. Many children report fatigue and hand pain with lengthy writing assignments. Early education and exposure to keyboarding may increase the amount of written output the child can produce. Older children may benefit from voice-activated software if they experience motor problems with keyboarding and writing.

> Scissors

Some children with smaller hand size demonstrate difficulty mastering scissor cutting, and benefit from a smaller size scissor proportional to their hand size.

> Carrying objects

Many children with Progeria are not able to carry their own school bag or books to and from school or during the school day. Those with difficulty in this area require accommodations such as a second set of books (one set at home and the second set in the appropriate classroom). Bags can then be lightweight, as all they need to carry are their notebooks or paperwork. If the child does wear a backpack, the bag should be no more than 15% of their body weight and should be placed over both shoulders. Additional accommodations include use of a backpack bag with wheels. The school therapist should complete a cafeteria assessment for lunch room adaptations that keep the child actively involved with of their peers (for example, ways to access the table tops or carry lunch trays). The children also often have difficulty walking and carrying moderately weighted objects. Most

frequently they are unable to carry objects up or down stairs and thus require help from a peer, teacher, or parent.

Social participation

Most children report participation in sports, playing on the playground, and other leisure activities. There is no evidence suggesting that these children should not participate in these activities unless it impacts their health. Activities such as contact sports, team sports, or leisure activities with their peers may require some adaptation to accommodate for their abilities and medical conditions. At times the activity demands may be too great or the child may need specialized equipment. Please refer to *Physical Therapy*, section 12, for further recommendations on physical activities.

Many children with Progeria experience fatigue when walking extended distances. In addition, they may not be able to keep up with their peers or family pace due to their shorter stride; this may impact their socialization. Use of functional mobility devices such as strollers, manual wheelchairs, or power wheelchairs may be needed in various environments. The child's therapist should complete a functional mobility assessment and provide the child and family with ways to allow the child to have optimal modes of mobility. For example, power wheelchair options (such as the Permobil which has a seat elevator and a chair-to-floor option) allow for increased independence. This chair allows the child to get in and out of the chair safely and to reach items at different heights, as well as navigate within the classroom, home, and community.

Treatment approach

After completion of an occupational therapy evaluation, a treatment program should be recommended. This may include direct services, home programming with follow-up, or ongoing consultation. Many children with Progeria will not require weekly services, but will require ongoing treatment with parent and child education.

The occupational therapist should provide evaluation and treatment to assist the children in all areas of function (self-care, education, work, play, leisure, and social participation). Children under the age of 6 years should be seen twice a year for an assessment by an occupational therapist. Children 6 years and older should be seen yearly for an occupational therapy

evaluation. If there is a significant change in function or other concern, the family should contact the therapist sooner. The treating therapist should have current medical history and be aware of all precautions. Ongoing communication is needed between the occupational and physical therapist, and may require combined treatment sessions at times. Accommodation or environmental changes may require minimum intervention but provide the child with optimal independence. An occupational therapy treatment program should include use of traditional physical disabilities treatment approaches, including passive range of motion with particular emphasis on the thumb, wrist, and fingers. At this time it is unknown if hand static splinting will improve range of motion; this should not be tried without the child first being seen for assessment by a pediatric hand specialist (MD). The therapist should provide the pediatric hand specialist with a comprehensive hand assessment that includes range of motion, strength, functional grasping, dexterity items, and activities of daily living.

Children with Progeria enjoy a very large array of activities. Despite their unique body functions and structural differences, there are many ways to accommodate their environment and tasks with adaptive devices and other changes that allow them to increase their independence and participation in activities of self-care, education, work, play, leisure, and social participation. Their involvement in these areas with their peers and their increased independence is important, especially as they become pre-adolescent.

Summary of environmental changes to help children with Progeria

> House

- Steps for bathroom
- Adapted switches and knobs
- Lower the placement of items for food preparation

> Mobility

- Adaptations differ depending on environment: home vs. neighborhood vs. larger community

> Allow for functional mobility

- Ease of mobility from place to place
- Ability to keep up with peers
- Mobility allows for socialization

> Recreation

- Adjust for safety or parents' concern
- Bike and/or tricycle

> School

- See *Going to School*, section 16