



## **PHYSICAL THERAPY FOR CHILDREN WITH MPS AND RELATED DISEASES INFORMATION FOR PROFESSIONALS**

**Stacey C. Dusing, PT, MS**  
**Angela Rosenberg, DrPH, PT**  
**Sadye Paez, PT, MS**

Children diagnosed with MPS and related diseases have a wide range of bony, joint, and muscle impairments that contribute to decreased motor abilities. The degree and type of motor impairment is specific to the child's diagnosed disease. In addition, the rate of progression of MPS varies significantly depending on the diagnosed MPS disease and the availability of medical treatments such as Hematopoietic Stem Cell Transplantation (HSCT) and Enzyme Replacement Therapy (ERT). The purpose of this fact sheet is to provide general guidelines for physical therapists that can be individualized to each child's specific needs.

Children with MPS and related diseases have multi-system medical problems that may impact physical therapy examinations and interventions. Physical therapists should be in contact with the child's primary care doctors and specialists to identify parameters for exercise and report any problems or changes observed during the course of therapy. Physical therapists should be aware of safety precautions specific to children with MPS. Respiratory compromise may result from airway obstruction and changes in the shape and size of the rib cage. Cardiac compromise may occur resulting in decreased endurance and tolerance to exercise. Given these precautions, heart and respiratory rate, as well as work of breathing, should be monitored in children with MPS throughout examinations and interventions. Hydrocephalus can also be an onset complication of some MPS diseases or may develop as the disorder progresses and may require shunt placement. Signs of hydrocephalus or shunt malfunction include: nausea, vomiting, irritability, changes in tone, and sun setting eyes. Children with MPS that have significant skeletal compromise, (MPS I, II, IV) may also be at risk of atlanto-axial subluxation, spinal cord compression, and carpal tunnel syndrome. These children need to be identified prior to initiation of therapy and precautions observed during therapy, in addition to yearly x-rays and nerve conduction studies as recommended by the primary care or orthopedic physicians.

Children diagnosed with MPS and related diseases should be referred to a physical therapist as soon as possible after diagnosis. The physical therapist must conduct a detailed review of the child's medical history and review medical records prior to the initial evaluation. Obtaining copies of all pertinent medical records prior to the initial evaluation will improve the accuracy of the history and documenting safety precautions that must be observed. The physical therapy examination includes a comprehensive assessment of gross and fine motor abilities using standardized measurement tools and passive joint range of motion assessment. Children with MPS may have discrepancies in their gross and fine motor abilities warranting the use of assessment tools that have distinct fine and gross motor scales. Standardized measures are beneficial for documenting changes in motor abilities over time or a lack of regression in skill for children who have not received HSCT or ERT. In addition to standardized measures, parents and children should be included as much as possible to obtain supplementary information regarding the child's functional abilities.

Parents and children with MPS are an integral part of the evaluation process and provide the foundation for functional goals and intervention/treatment strategies

Children with MPS and related diseases may benefit from a variety of treatment options based on the impairments and activity limitations identified in the examination. Gait training, static and dynamic balance, and activities of daily living should all be addressed in a functional context. Functional skills such as moving between positions or completing daily routines should be emphasized. Strengthening activities are especially important for children who have had a HSCT as multiple medications may contribute to a decline in strength needed for functional activities. It is important to encourage prolonged stretching in a functional context multiple times each day to preserve range of motion. Examples include long sitting or wedge standing while watching TV or reading to address knee extension and ankle dorsiflexion range of motion. Applying splints for nightwear or during naptime may also be effective at maintaining or regaining range of motion. Therefore, physical therapists may also need to assess the child's need for orthotics and/or braces. In general, joint motions must be restricted as little as possible while maintaining lower extremity alignment. Repeated joint motion over the course of the day is beneficial to maintaining joint motion and should not be restricted unless it is contributing to poor alignment. Children should not be forced into a brace for daily use that does not accommodate their current joint deformity. Thus having a child wear an ankle foot orthosis that is set at neutral when their ankle can not be dorsiflexed to neutral will only damage their foot rather than improve their ankle range of motion. Children who have more involved needs may need to be assessed for adaptive equipment such as wheelchairs, special car seats, bath chairs, hand splint, adaptive eating utensils, etc. It is recommended that physical therapist consider working with other professionals such as occupational therapists, speech therapists, augmentative communication specialists, and psychologists to evaluate children with MPS. Evaluating children with MPS with an interdisciplinary team approach will maximize the skills of all participating therapists and ensure the child receives all needed services.

As with intervention strategies, the frequency of physical therapy intervention should be individualized. As a general guide, children who are being treated to maintain skills and prevent the regression of skills may benefit from 1-2 times per week physical therapy with a function-based home program the family can complete on a daily basis. In contrast, children who have received a HSCT or are receiving ERT may benefit from more intensive therapy to capitalize on their potential for skeletal changes and improving motor patterns.

The goal of physical therapy for children with MPS and related diseases is to maximize the children's functional abilities. Physical therapy will not cure MPS and will not stop the progression of the disease. However, physical therapy can be beneficial to limit the impact that changes in the body structures have on functional abilities. More importantly, physical therapy can facilitate independence and participation in age appropriate activities.

In summary, physical therapy is a vital component of the care plan for children with MPS and related diseases. Physical therapists must be aware of safety precautions specific to each child's participation, be in frequent communication with the child's medical team, conduct standardized and functional assessments, and develop functional goals in collaboration with the child and family.