

Physiotherapy: developing and improving motor functions

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Children and adults with Rett syndrome (hereafter, called patients) benefit from physiotherapy, which helps them develop and maintain motor functions.

Body-structure and functional motor-skills assessments give an indication of appropriate treatment and preventative measures. Stakeholders (parents, therapists, caregivers) should aim to motivate patients during therapy and daily living. Persons within patients' daily-living networks are crucial because they can encourage the patients to use their acquired functional gross motor skills and aids on a daily basis.

Standing and walking

Patients experience increased, decreased and/or alternating muscle tone. As a result, joint deformities may develop that can increase total energy consumption and contribute to functional impairments and physical limitations. The most effective approach to address these complaints is to maintain or increase soft-tissue extensibility and long-term elongation, for example via foot braces and standing aids.

Physical activity maintains tissue elasticity and muscle strength. Base-line activities, such as standing and walking – with or without aids – are critical success factors.

Encourage patients to walk on uneven surfaces, such as on forest paths, because this promotes tissue elasticity and natural leg-muscle elongation. Treadmills also help because the speed increases step length and thus the range of motion in hips, knees and ankles.

If balance and muscle strength are poor, try to arrange standing and walking exercises in a pool – this is an excellent option. Water in the pool has weight-bearing properties, which facilitates standing and walking activities. Weight cuffs around the ankles enable the feet to maintain contact with the pool floor.

Consider using a walking harness connected to a ceiling hoist. Stations that provide various sensory experiences or dancing to music may increase motivation to walk around in the room.

Note: Rett syndrome is not a progressive disorder. Gross motor skills, such as walking, *might* deteriorate or fade away in adulthood. Nevertheless, *do not* expect deterioration to occur. Secondary causes, such as poor health, severe *contractures* (muscle shortening) and inactivity might diminish skills. However, researchers report that regaining walking capability *has* occurred in adulthood – after many years in a wheelchair [1] [2]

Spine

The risk of developing spine deformities is high among patients. Double curved or c-shaped scoliosis can appear in varying degrees. Some patients develop *kyphosis* (an increased upper-back curve). Standing and walking – with or without aids – are very important for delaying deformity developments in the spine. Strengthening neck and back muscles is crucial; to do so, encourage patients to lie on their stomachs on top of floating devices in pools. A balanced sitting position

(with the feet firmly positioned on the floor and the buttocks evenly balanced on the seat of a chair) affects the position of the spine positively. Many other solutions enable stable sitting positions.

Assessments of each patient's needs and regular follow-ups are vital. Expert consensus and clinical evidence have enabled the creation of guidelines for scoliosis management for patients with Rett syndrome [3].

Feet

Stakeholders must be aware of the importance of foot positioning. Depending on muscle tone, foot weight can either be loaded (placed) on the outward or inward part of foot soles. This, in turn, affects knee and hip positions, which trigger unnecessary leg tension.

Standing high up on the toes – due to high muscle tone in calf muscles – is an undesirable foot position, because it diminishes the loading base, which, in turn, affects balance when standing and walking.

Assist patients in keeping their feet flat on the floor or on the footrest of a wheelchair, because this allows them to keep their trunk in an erect position for as long as possible.

Keep an eye on the feet. If any deviations occur, take immediate action to prevent permanent deformations.

Note: If sturdy shoes, insoles and orthopaedic shoes are insufficient for stabilizing the feet, inquire about foot braces for keeping the feet in a neutral position. Brace selection requires an individual assessment to obtain the best effects. Prosthetists/orthotists shape braces in various ways and in various materials.

If misalignment is extensive, and if neutral alignment cannot be restored, then a Botox injection can relax the foot and calf muscles and increase the ability to stretch the muscles. Sometimes, orthopaedic surgery is the only solution for improving gross motor function because it can elongate muscles or re-position tendons.

Dyspraxia and latency

Encourage patients to use and maintain their acquired functional gross motor skills that might be hidden due to *dyspraxia* (difficulty in initiating purpose movements).

Guide patients into a movement (get them started). Once in motion, they can use existing skills.

Note: Repetitive functional experiences might assist in overcoming dyspraxia. Execution of movements and transitions are sometimes slow, therefore requiring more time for independent achievement.

Balance between physical activity and recovery

Daily living must contain a balance between physical activity and recovery. Both should be included and alternated, with the expectation of active participation and activities that are less demanding.

There can be both physical and psychological reasons/needs for recovery. For example, relaxation might enable a feeling of comfort if muscle tone is high and the body is very tense. Relaxation is particularly important if anxiety and/or stress occur or if the goal is to prevent anxiety and stress.

Use touch, gentle pressure, vibration or warmth to stimulate skin receptors in order to achieve a sensation of calmness in the body.

Note: Patients have their own preferences regarding what makes them feel good and relaxed. Some like tactile massage. Others prefer floating in a warm pool or taking a footbath. Swinging, listening to calming music or sitting close to someone and having a nice chat are also relaxing activities.

Positioning facilitates relaxation in bed. Joint deformities and tense muscles may interfere with relaxation. Use pillows and/or rolled up towels to control the forces that affect the body (eg. gravity and muscle tone). The ultimate goals are to stabilize body sections, achieve a symmetric position, reduce the risk of pressure and pain and obtain a comfortable position.

References

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