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Neuroorthopaedics

1. Diseases of the brain, the spinal cord, nerves, and muscles influence function and form of the musculoskeletal system

Generally Neuroorthopaedics is not been regarded as clearly defined subspecialty of the field of orthopaedics. It is rather a way of thinking and managing complex static and movement disorders of a group of cerebral and neuromuscular diseases.
Neuroorthopaedic specialists are diagnosing, analysing, treating and caring for children and adults with different kinds of motor disabilities.
Most common cerebral disorders are hemiplegia after stroke in adults and cerebral palsy in children. Spinal cord dysfunctions are caused by congenital dysraphy disorders, like myelomeningocele, hereditary spinal tract disorders, and traumatic spine lesions.
Poliomyelitis is an infection of the anterior cell and spinal muscular atrophy is a congenital diseases of these cells.
Neuropathies might be due to rare hereditary disorders like Charcot-Marie-Tooth-disease, a hereditary motor-sensor neuropathy, or due to very common toxic influences like diabetes or alcoholism.
Muscle disorders are either congenital non-progressive or progressive, like Duchenne muscle dystrophy, or caused by metabolic or inflammatory processes.

All of these neuromuscular diseases produce typical disorders of the musculoskeletal system that are influencing activities of daily life of the patients.
Improving quality of life is one of the main goals of Neuroorthopaedics. Considering motor activities, sensory cognitive, emotional, and psychological functions are important for attaining new possibilities. A multidisciplinary team approach has turned out to be essential (Stotz 2000).

Example: Cerebral Palsy
Cerebral palsy is among the most frequent disorders of the growing motor system. It is the most common cause of physical disability affecting children. The incidence is 2,0 - 3,0 per 1000 live births. Severity of the motor disorder is dependent on the extension of the central nervous damage. According to the local affection cerebral palsy may be subclassified as hemiparesis, diparesis and tetraparesis. Each of these clinical entities has its typical symptoms, pathophysiology, and prognosis. The term Paresis means an incomplete loss of muscle function, for example spastic or dystonic paresis, whereas Plegia is associated with a total loss of function, for example after damage of the spinal cord or peripheral nerve. Nevertheless in literature the terms paresis and plegia are commonly used as synonym.
Depending on the lesion of the central nerve system structure different clinical characteristics of muscle dysfunction may be observed: spasticity, weakness, lack of motor control, ataxia, dystonia, athetosis, rigidity and most commonly mixed types.
Hemiparesis is defined by unilateral dysfunction of distal muscles of the upper and lower extremity. It is caused by cerebral ischemia or haemorrhage before, during, or after birth. Children usually start to walk between 18 and 24 months. Without treatment they are disabled by a slight progressive motor and musculoskeletal asymmetry.

Diparesis is the internationally most common form of cerebral palsy. Prematurity is associated with the disease in more than 60 percent. After a period of muscular hypotonia and delayed motor development, spasticity usually starts during the second year and increases by the forth year of life.
Motor development improves gradually and most of the children will be able to walk freely between 3 to 7 years. Independent walking ability is subject to the primary neurological deficit, spasticity, associated sensory deficits, disorders of balance and motor symmetry, and to secondary muscle shortening, contractures, and bone and joint deformities.

Tetraparesis is the most severe entity of cerebral palsy. It is caused by an extended congenital or secondary brain damage. Due to lack of motor control of the head, trunk, and pelvic muscles children are usually not able to develop free sitting or standing or walking function.
Associated sensory, proprioceptive, visual, vestibular, auditory, cognitive, emotional and psychic disorders are very common in this group.

Clinical features of all forms of cerebral palsy include typical deformities of the upper and lower limbs. Weakness as well as spasticity due to lack of selective neuronal control causes functional impairment and additional mechanisms of compensation, retardation of motor development, secondary deformities of muscles and soft tissues due to a failure of muscle growth, instability and dislocation of joints, early osteoarthritis, and pain.

Main problems of motor function in cerebral palsy are crouch gait and stiff-knee gait, flexion and extension deformities, and disorders of feet, hips, and the spine. For successful treatment the pathologic mechanisms have to be fully understood. Special examination tools and therapy options are applied to gain satisfying long-term results. Goal is improvement of quality of life like pain free mobility for social integration (Döderlein 2007).

2. Different neuromuscular diseases cause similar patterns of pathologic motor function and disabilities

Form follows function: for example normal development of the knee joint, leg biomechanics, standing and walking is dependent on physiologic use, weight bearing, muscular power and balance due to voluntary gross and fine motor function of the whole lower extremity.
Prerequisites of normal lower limb function are the daily use of full range of motion, standing by a slight combined extension and rotation of the joint, and energy cost efficient walking. Voluntary motor control plays an important role in normal muscular and power development.
Recent research has shown that the ability to extend the knee during swing is dependent on the selective voluntary motor control. Stance limb muscle strength appears not to be the limiting factor for achieving adequate knee extension (Goldberg 2009).

Concerning the motor system cerebral palsy may be defined as a disorder of selective neuronal control of muscles. Weakness as well as spasticity due to this lack of control causes functional impairment and additional mechanisms of compensation, retardation of motor development, secondary deformities of muscles and soft tissues due to a failure of muscle growth, instability and dislocation of joints, early osteoarthritis, and pain.
This means developing a vitious cycle of primary, secondary and tertiary deformities.

Neuromuscular foot deformities
Neuromuscular disorders of feet in children and adults are common. In many cases they are the first symptoms that lead to the diagnosis of a neurologic disease. Progressive foot deformities may cause severe limitations in daily living activities. Identification of hidden neuromuscular diseases and understanding the pathophysiology are crucial for planning treatment and estimating prognosis of any foot deformity. Before exact indication for conservative and surgical therapeutic options we need to analyse the pathogenesis of the neurogenic pes equines, flat foot, club foot or pes cavuvarus.
The very commonly observed *equinus deformity* is caused by hyperactivity of the plantar flexors or severe weakness of foot extensor muscles.
Clubfoot deformity is produced by weakness of the pronator group, for example by complete paresis of the peroneal nerve, or by supinating hyperactivity.
Flatfoot deformities are commonly seen in patients with lack of control of the hind- and midfoot stabilizing muscles, like in spastic diparetic form of cerebral palsy.
Cavovarus deformity may be caused by deficiency of short foot muscles, for example in all kinds of neuropathies.

Systematic decision making in indication may help to find out the best therapy for each kind and stage of foot deformities. Balance of muscular forces, stability and range of movement of the foot skeleton have to be considered.
Treatment options include specific orthopaedic adaptions of shoes, custom-made orthoses for daily activities, supporting physical and movement therapy, drugs, and surgical procedures.
Goals of treatment are maintenance and improvement of the patient´s mobility by lack of pain and easiness of shoe-wearing through all periods of life.

Knee flexion deformity
Flexion deformity of the knee may be primary, caused by spasticity and shortening of the hamstrings. It may be secondary to compensate equinus deformity of the ankle and flexion deformity of the hip. Or it may be functional to lower the center of gravity to achieve balance as it is seen in patients with triceps surae weakness following Achilles tendon lengthening (Tachdijan 1990).
Structural shortening of muscles and contracture of the posterior knee joint capsular are insidious problems which may complicate the management of patients with cerebral palsy. Flexion contractures of more than 20° induce severe disabling degenerative arthritis in adults. Many of these patients have been asymptomatic until the age of 35 to 40 years (Bleck 1987) is common in patients with crouch gait. Clinical tenderness and fragmentation of the distal pole of the patella in radiographs develops due to the constant pull of the patellar tendon. These problems interfere with transfer standing and energy cost efficient walking. Secondary pain may ensue as a result of fragmentation of the patella and tibial tubercle. Furthermore flexion deformities of hips and lumbar lordosis as well as pseudo-equinus of the ankles may develop.

Crouch gait
By k-means cluster analysis Rozumalski and Schwartz (2009) found five different clusters among children with excessive knee flexion at initial contact. They labelled these clusters in order of increasing gait pathology:

- Mild crouch with mild equinus
- Moderate crouch
- Moderate crouch with anterior pelvic tilt
- Moderate crouch with equinus
- Severe crouch

Age, range of motion, strength, selective motor control, and spasticity were significantly different between the clusters. So the authors recommend this classification for treatment decision making and outcome assessment.
Stiff-knee gait is the common gait pattern in patients with rectus femoris spasticity. High energy consumption reduces walking range and mobility in daily life. Rectus femoris spasticity and shortening causes reduced range of motion of the knee joint. Reduction of ROM in swing phase influences step length and biomechanical function of bilateral hip and ankle joints as well as pathology of the femoropatellar joint.

Neuromuscular hip instability
Progressive hip dislocation occurs in several neuromuscular disorders with lack of control of the pelvic and lower extremity muscles. It is common in patients with cerebral palsy GMFCS IV and V and with lumbar spinal cord lesions.
Physiologic motor and biomechanical parameters are prerequisites for normal hip development and hip function. Disorders of muscle activity and lack of weight bearing due to neuromuscular diseases may cause clinical symptoms like an unstable hip or reduced ROM. Disability and handicap because of pain, hip dislocation, osteoarthritis, gait disorders, problems in seating and positioning and of caring are depending on the severity of the disease, on the time of occurrence, and on the means of prevention and treatment. Preserving pain free and stable hip joints should be gained by balancing muscular forces and by prevention of progressive dislocation.
Helpful prognostic criterias for development of the hip joints are the extent of central neurogenic damage, active and passive range of movement of the joint, effect of gravitation and stage of decentration of the

femoral head at the start of therapy.

Neuromuscular spine deformities
Spinal deformities may develop on the one hand as result of muscular weakness, spasticity, or lack of trunk control and on the other hand as associated disorders combined with congenital spinal cord malformations.
Excessive kyphosis and lordosis are common in patients with insufficient control of spinal extensors. Upright position should be obtained to improve sensory functions and to reduce back pain.
Neuromuscular scoliosis are commonly seen in patients with reduced motor trunk control. Progressive pulmonary dysfunction and lack of seating ability following pressure sores have to be prevented.

3. Neuroorthopaedic evaluation has to include motor, sensor, cognitive, and psychic abilities and activities in daily life

GMFCS
The Gross Motor Classification System helps to distinguish patients with neuromuscular disorders according to their gross motor function abilities (see Fig. 1):

- **Level I**: free walking, climbing stairs, running and jumping; speed, balance and coordination are limited
- **Level II**: walking with aids, limitations for long distances, for running and jumping
- **Level III**: walking with the support of walkers, wheelchairs used for long distances
- **Level IV**: transfer standing and walking with support for short distances, transported in a wheelchair or powered mobility
- **Level V**: transported in a wheelchair, lacking head, arm, and trunk control.

DLA daily living activities, social environment
Knowledge about the patient´s social integration and daily activities and skills are important for decision making whether therapy programs, casts, splints, drugs, or surgical procedures will be the appropriate mean to reach the defined goal of functional improvement. According to the systemic approach the specialist will draw into consideration as many factors as possible.

Sensory system and cognitive function
The clinical examination of a neuroorthopaedic patient contents gathering information about basic neurogenic functions like sensory and proprioceptive function, equilibrium, vision, auditory, cognitive and communication abilities.

Musculoskeletal and motor system
Observation of the individual walking pattern, active and passive joint mobility, ROM and measurement of muscle length are documented. Axial bone deformities, capsular joint contractures, structural and dynamic shortening of muscles have to be distinguished.
Sliding of the patella in the patellar-femoral groove has to be examined by full flexion and extension of the knee.
Muscle spasticity is documented according to the Ashworth scale (Richardson 1998).
Muscle strength is measured according to the Medical Research Counsil Scale (Hislop 1995).

Length and spasticity of the hamstrings is evaluated by measuring the *popliteal angle test.* To palpate the hamstrings and get a better estimate of shortening, hip is flexed to 90° and then the knee is extended to the limit permitted by the hamstrings. The angle between the tibia and the vertical line is measured or estimated – it is the popliteal angle. Ten Berge (2007) did not see any differences in reliability between visual estimation and goniometric measurement. 20° is considered to be normal. A recent study did not find any change between examination in the clinic versus under anesthesia McMulkin (2008).

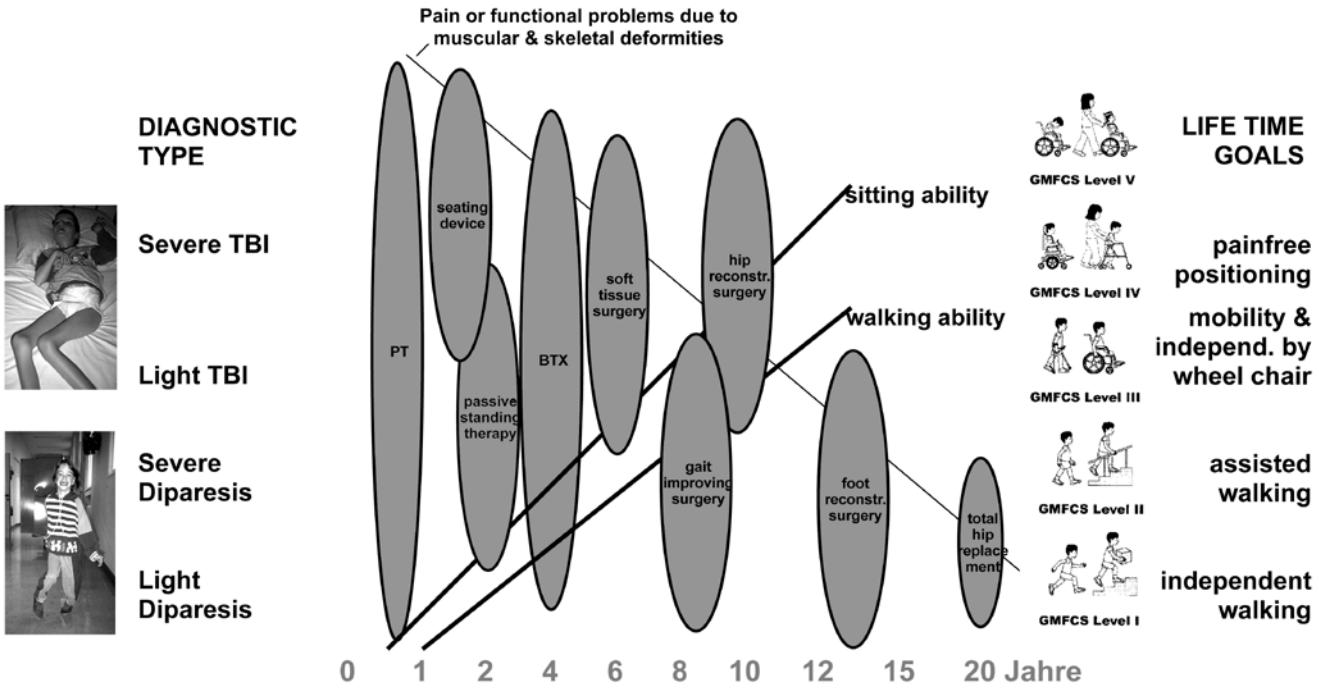

Fig. 1 Life of children with cerebral palsy: Time for therapeutic indications depending on age, Gross Motor Function Level, and corresponding life time goals (Strobl 2011)

Length and spasticity of the rectus femoris muscle is examined by the *Duncan-Ely test* in prone position flexing the knee to the buttocks. The examiner slowly flexes the child`s knee on one side of his or her body. If there is a rectus femoris contracture the pelvis slowly rises off the examination table as the knee is flexed. This reaction is the resultof the rectus femoris crossing both the hip and the knee joint. In addition to testing contracture, the test also can examine the rectus muscle for spasticity but the quadriceps stretch has been shown to elicit a reflexive iliopsoas firing that may also cause hip flexion.

The amount of a *flexion contracture of the knee joint* is measured while the limb lies flat on the table. If firm pressure on the anterior aspect of the knee fails to extend the joint in 0° a contracture of the posterior capsular in addition to the hamstring shortening is likely.

Length of gastrocnemius and soleus is evaluated by the *Silverskjöld test.* Passive ankle dorsiflexion is estimated with (soleus) and without (gastrocnemius) knee flexion. McMulkin (2008) described a significant change of dorsiflexion angle between examinations in the clinic versus under anesthesia in children younger than 11years.

Radiographs
In neuroorthopaedic patients radiographic evaluation of the spine, hip, knee, and feet are routinely used to diagnose spine malformations, hip abnormalities, axial deformities, disturbances of growth plates, disorders of the feet. Additional investigations of the musculoskeletal system in neuromuscular diseases are made by MR or CTscan.

Videofilms
The evaluation of films with slow motion functions helps to differentiate functional gait disorders. Furthermore films play an important role in the documentation of the treatment and rehabilitation process and should belong to the routine equipment of a neuroorthopaedic clinic.

3D-Gait Analysis
The role of gait analysis in the management of neuromuscular patients is controversial. In the evaluation of walking dysfunctions in neuromuscular patients concerning decision making for surgery the authors recommend its routine clinical use. Clinical gait analysis helps to distinguish ambulatory children with neuromuscular disorders who would benefit from surgery from those in whom non-operative management was appropriate (Gough 2008). It is useful in defining indications for surgery that is clinically proposed, and for excluding or delaying surgery that is clinically proposed.

Dynamic electromyography
Dynamic EMG datas give insight in the function of muscles during the gait cycle. Spastic muscle activities of the lower limbs as cause of reduced stance phase stability or swing phase mobility may be identified. Its routine use eases the composition of therapy programs and the indication for surgical procedures.

4. Neuroorthopaedic treatment and caring need a team approach

Principles
Weakness as well as spasticity due to lack of selective neuronal control causes functional impairment and additional mechanisms of compensation, retardation of motor development, secondary deformities of muscles and soft tissues due to a failure of muscle growth, subluxation/dislocation of joints, early osteoarthritis, and pain.
Prevention of this vitious cycle of primary, secondary and tertiary deformities may be defined as the main goal of caring for children and young adults with spasticity.
Early detection of walking disorders, for example a progressive knee flexion contracture, is crucial in qualified orthopaedic screening programs for neuroorthopaedic patients. Optimal function of the knee joint is one of the most important prerequisites for life-time transfer standing and walking with adequate energy consumption (see Fig. 2).

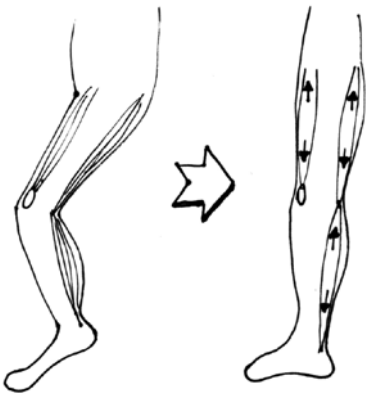

Fig. 2 Therapy goal even for severly disabled neuroorthopaedic patients: obtaining the ability of transfer standing and assisted walking by single-stage-multi-level-surgery and orthoses (Strobl 2011).

By clinical orthopaedic examination an exact differentiation of conditions with increased muscle tone or reduced ROM has to be performed:

- reduced muscle tone may need stabilizing treatment like orthoses
- normal tone will not need treatment
- increased muscle tone may be treated by physical means like warmth, massage, continuous active or passive movement
- spasticity (typical clinical diagnosis) is the indication for treatment by BTX A
- dynamic shortening of muscles may be treated by stretching exercises or/and casts in combination with BTX
- structural shortening of muscles is to be treated only by surgical intramuscular or aponeurotic lengthening in combination with exercises, casts and orthoses
- capsular contractures may need additional osseous procedures
- bone and joint deformities will need combined soft tissue and osseous reconstructional surgery followed by activating long-term rehabilitation.

Quality of life of children and adults with neuromuscular diseases can be improved by support of their daily living motor activities. Regular joint movement, weight bearing, improvement of motor control and strength training are main topics.

Increased muscle tone may be reduced by physical exercises, by individually adapted orthoses, especially ankle-foot-orthoses (AFO), walkers, and sometimes wheelchairs for long distances, by manual therapy, serial casting and in certain cases by systemic drugs or by multiple-stage surgical procedures.
Instable joints may be treated by orthoses or orthopaedic surgery.

Operative treatment is indicated when conservative measurement is failing to reduce spasticity and orthoses are not capable to counteract weakness in order to improve motor development and maturing of personality.
In the past first surgical procedures in children with CP have commonly become necessary between the age of 3 to 5 years. Careful multi-level surgery procedures like adductor release, hamstring release, and gastrocnemius-lengthening have been perfomed in that age, sometimes done at the right age to reach independent walking. Until today studies describe that surgical intervention in selected young children can result in improvements in gait and function in the short to mid-term compared with non-operative management (Gough 2008).
Today these procedures are still indicated, but by injections of Botulinum Toxine A in this age group surgery may be delayed to early puberty. So a decreasing number of children are in need of repeated muscle releases during their growth.
Usually one multi-level procedure including rotational deformities is performed to improve walking, to get better motor symmetry and to reduce energy consumption in daily life´s locomotion.

In the long run quality of life for adults with any neuromuscular disorder is determined by self assured mobility for adequate social integration and pain free walking or at least transfer standing without progressive deformities up to a higher age. All of them will be in need of specialized physical therapy and rehabilitation programs

Sports, daily exercises, and physical therapy
Daily mobility and activity help all patients with neuromuscular disorders to prevent loss of strength, coordination and progressive muscular dysfunction and shortening.
Reduction of increased muscle tone and spasticity may be obtained by application of warm temperature, massage, reflex therapy, osteopathic handling, and continuous active and passive movement therapy.
Physical stretching exercises are improving dynamic shortening of spastic muscles and reducing spasticity.
The effect of *electrical stimuation* is controversial. Combined with passive stretching it is described to be marginally more effective than passive stretching alone (Khalili 2008).
Strength training may improve walking function and alignment in patients for whom weakness is a major contributor to their gait deficits. Due to the variability of outcomes in several strengthening studies in CP analytical approaches to determine the causes of weakness are needed to identify those individuals who are most likely to benefit from strengthening (Damiano 2009).
Only for patients with progressive muscle weakness due to dystrophinopathies, like Duchenne or Becker-Kiener disease, strength exercises are contraindicated.

Orthoses for positioning and improving hand and sensor function
Standing devices
Short or long leg standing orthoses are offered for patients with lack of motor control of the lower limbs.
In the case of lacking control of pelvis and trunk hip-knee-foot orthoses are necessary to stabilize the total body in order to improve hand function and head control. Standing orthoses or standing frames may satisfy these demands.
Standing represents an elementary part of people moving their body. The stages at which humans are able to weigth bearing primarily by using their upper extremities, and finally standing freely are important milestones in learning to control the movements of their body. Mobility and upright position are prerequisites for proper function of cardiopulmonary, gastrointestinal, and urinary tract organs, thus being a substantial precondition for achieving adequate quality in life. Suitable measures to prevent the postural and locomotor system from being damaged should already be taken in childhood.

Physiologic function of the following organ systems is dependent on the fact that the human body is temporarily able to stand or can be brought into a vertical position under the influence of gravity:

- function of cardiovascular system, vascular system and blood pressure regulation
- pulmonary function and oxygen supply
- gastrointestinal system
- urinary tract function
- calcium-phosphate metabolism of lower-extremity bones
- cartilaginous tissue of lower-extremity joints
- metabolism of the muscular apparatus
- additional functions during the growth period:
- growth plates of lower extremities
- physiologic growth of lower-extremity muscles
- physiological development of the vertebral column as well as of hip, knee and ankle joints

Seating devices
Patients with neuromucular disorders are not able to adapt their sitting posture continously. Seating devices, like seating orthoses, braces, seating shells, and custom-made cushions for wheelchairs, however may improve their quality of life by stabilizing their pelvis and trunk.
Sitting should be regarded as a dynamic process regulated by motor reactions of trunk and pelvic muscles due to endogenic and exogenic influences. Prerequisites for the indication of high quality and cost effective seating devices are guidelines for planning and fitting which consider both pathomorphologic mechanisms and the patient´s personality. In order to avoid functional problems and pain caused by an insufficient seating device it is necessary to pay attention to the exact indication, time, and combination of technical options.
Planning within a seating clinic needs teamwork. Primarily the goal of treatment is defined; it depends on the functional deficit, on the daily living activities of the patient, and on the social environmental factors. Secondly fitting of the devices follows defined treatment guidelines.
By examination of the sensor and musculoskeletal system it is possible to classify the patient´s sitting or seating ability for simplifying indication: three groups of ACTIVE sitters who are able to change position of trunk and pelvis actively are differentiated from three groups of PASSIVE sitters who have to be seated.

Orthoses for improving walking
Orthopaedic shoes and foot orthoses (FO) may improve pain and pressure sores of deformed feet. Goal is stabilizing the hind- and midfoot in neutral position for easing standing and walking. In walking patients orthopaedic shoe wear may improve knee and hip extension in stance phase.

Studies indicate the potential clinical utility of tuning using wedges to correct knee hyperextension during the stance phase (Jagadamma 2009).

Ankle-foot orthoses (AFO)
AFOs improve toe-walking in children with CP. Studies comparing different kinds of dynamic orthoses describe improvement of gait pattern but only small differences between the configurations of the orthoses. Hinged and dynamic AFOs are equally effective for improving ankle kinematics and kinetics in GMFCS level I children.
In quadriplegic children the use of an AFO results in a significant decrease in energy cost of walking compared with barefoot walking. It is related to both a faster and more efficient walking pattern. The improvements in efficiency are reflected in changes of stance and swing phase knee motion towards a typical normal range.
The floor-reaction ankle-foot orthosis is commonly prescribed for CP children who walk with excessive knee flexion and ankle dorsiflexion during the stance phase of gait. It is effective in restricting sagittal plane ankle motion during stance. Best outcomes are reported in subjects with knee and hip flexion contracture of less than 10 degrees. Contractures of more than 15 degrees should be considered as contraindication.

Hip-ankle-foot orthoses (HAFO) are necessary to stabilize lower extremities in spite of lacking motor control of the pelvis.

Orthoses for guiding growth
Ankle-foot orthoses (AFO)
Used as night time splints this group of orthoses is well tolerated by patients and may improve foot and hand function by reducing muscular imbalance, dynamic shortening, and by improving strength and reducing overlength of their antagonists.

Knee-ankle-foot orthoses (KAFO)
By supporting physical stretching exercises KAFOs may prevent knee flexion deformities and increase knee extension in stance. They can be temporarily used as day or night splints. Cartilage nutrition and patient´s compliance may be improved by the use of elastic joints which allow defined motion of spastic hamstrings or biceps femoris muscles.

Serial casting
Application of short leg casts is regarded as a simple, safe, cost effective, and well established procedure to reduce spasticity and improve walking in patients with spastic diparesis. The tonic stretch reflex of spastic muscles is used as treatment principle. Dynamic lengthening of both gastrocnemius heads improves knee extension in stance. Casts are applied for about four weeks. Shortened muscles are gradually stretched by serial casts which are applied one or two times per week. Special padding is necessary to avoid skin breakdown. Producing iatrogenic flatfoot- or clubfoot deformities have to be prevented by casting in slight overcorrection of the hindfoot. Serial casts can be applied even in children with mental subnormality having all three major joints involved bilaterally. In contrary to more sophisticated orthoses this procedure is also commonly used in developmental projects because of its described advantages.

Drugs
Systemic drugs
The centrally effective antispastic drug Baclofen is used as systemic tone reducing agent. Stiff-knee gait as part of general spasticity is reported and experienced to be improved significantly. Application of higher dosages may cause side effects like fatigue and reducing the patient´s vigilance.

Local drugs
Neurotoxins like phenole and botulinum toxine A are applicated directly to the nerve or muscle in order to reduce spasticity and muscular imbalance and to improve the potential underlying voluntary motor activity.

Botulinum toxine A
BTX A is suggested to be one of the most powerful and useful tools in reducing spasticity. Repeated injections have a long-term effect on gross motor function in children with CP.
Limitations of BTX treatment have been reported in studies: Glanzmann (2004) found that in children with spastic equinus deformity casting demonstrated a significantly more robust impact on ROM than BTX-A alone. Kay (2004) stated that serial casting alone is preferable for the treatment of fixed equinus contactures. Concerning the treatment of progressive hip dislocations there is a significant difference between children with CP treated by multi-level soft-tissue-surgery and those treated by series of BTX-injections of the same muscles. They suppose the anti-spastic effect of BTX has not been as continuous as surgical tone reduction (Molenaers 2005; Strobl 2005).

Neuroorthopaedic surgery
Principles
Surgery in neuromuscular disorders needs specialized knowledge and experience. Functional problems have to be detected as early as possible, they have to be analysed, pathologies have to be understood and addressed at the right age by the optimal surgical method in a well-dosed manner. The paediatric orthopaedic surgeon Mercer Rang (1986) used to comment to the point: the indication is more important than the incision.
Surgeons have to be aware of postoperative complications like nerve palsy especially in non-communicative and non-ambulatory adolescents, persistent pain especially in cognitive disabled individuals, temporarily or persisting increased spasticity, skin breakdown at heel area especially in patients with sensory and/or communicative deficits, deformity recurrence, persistent muscle weakness in spite of training programs, necessity for walking aids, delayed rehabilitation process
Optimal postoperative pain control eases spasticity and avoids the vitious spasm-pain-cycle.
Immobilization always has to include adequate padding of risk sites for skin breakdown like heels, ankle joint, patella, and the dorsal proximal femur region.
Vigilance in patients with epidural pain control to avoid nerve palsies following excessive knee extension and hip flexion is warranted. In the case of symptoms immediate knee flexion is necessary to avoid persistent motor and sensory deficits.
Despite the normalization of ROM after surgery there is an early postoperative period of functional gait deterioration which has to be considered. Early mobilization and strength training needs perfect team work and interdisciplinary management between surgeons, rehabilitation specialists, physical and occupational therapists, and orthopaedic technologists.

Surgical procedures

Muscle lengthening is performed to obtain physiologic length of short and hyperactive, spastic muscles.

For example: Intramuscular medial hamstring lengthening in cases when hamstring spasticity causes a dynamic knee flexion deformity which is an indication for this procedure. This prohibits knee extension during the stance phase of gait and interferes with efficient ambulation. Progressive structural shortening produces severe crouch gait, an indication for surgery. Reducing knee pain, improving function and independence may be expected.

Myofasziotomies are percutaneously performed releases of fibrous tissue within the muscle bellies. In selected patients this minimal invasive technique may be superior to open lengthening, studies describe the short- and long-term outcome to be sufficient.

Muscle shortening is performed in weak and too long muscles to obtain physiologic length and strength.

For example: Patellar tendon advancement is indicated in the case of persistent crouch gait in adolescents and young adults, quadriceps insufficiency due to quadriceps spasticity and structural shortening, patella alta, and elongated patellar tendon.

Muscle transfers are performed to achieve an improved, balanced muscle function.

For example: Distal transfer of rectus femoris muscle is indicated in the case when rectus femoris spasticity causes reduced knee flexion during the swing phase. This prohibits adequate step length and interferes with efficient ambulation resulting in a stiff-legged gait. To achieve adequate limb clearance compensatory circumduction of the involved limb or pathologic pelvic movement is necessary.

Tendon transfers are performed to balance muscle power and stabilize joint functions.

For example: Tibialis anterior transfer to the dorsal aspect of the foot is indicated in patients with spinal S1-lesions, gastrocnemius- and soleus muscle plegia, and consecutive calcaneal foot deformity. So the tibialis anterior muscle will work as plantar flexor instead of the paralysed triceps surae.

Split-transfers of muscles may be indicated to stabilize joints without sufficient motor control. Goal is a *functional arthrodesis.*

For example: A tibialis posterior tendon transfer is indicated in non-structural spastic clubfoot deformities to obtain a normal foot position.

Joint repositions and reconstructions are very commonly indicated in the cases of severe dislocations and non-structural foot deformities and hip dislocations to obtain stable and pain free joints for weight bearing, improved standing and walking ability.

For example: Chopart- or Triple-Arthrodeses are indicated in neuromuscular clubfeet, flatfeet, and cavovarus deformities.

Arthrodesis is indicated in severe structural foot deformities to obtain normal shape without danger for pressure sores. Goal is ease of shoe wear and improved standing and walking function. Osseous arthrodesis should be avoided in big joints like the hip, knee, and malleolar joint whenever possible because minimal movement of joints is necessary for the equilibrium of movements.

Osteotomies of long bones may be indicated to improve lever arm dysfunctions.

For example: Distal supracondylar femoral extension osteotomies are efficient for increasing knee extension and improving walking in patients with fixed knee flexion deformities, even with severe capsular contractures.

Spondylodesis is performed in patients with progressive neuromuscular scoliosis to improve seating and to prevent the danger of decreased pulmonary function. In patients with muscular dystrophy the indication for spinal stabilization starts as early as COBB angle reaches 20-30 degrees.

Selective dorsal rhizotomy (SDR) is a dissection of selected nerve roots. It is indicated in the rare cases of high spasticity that overlies very well voluntary muscle activity especially in children with spastic diparesis. Crucial is exact diagnostics and evaluation by GMFCS, daily living activities, ROM, spasticity according to Ashworth scale, muscle strength according to Oxford scale, radiographs, 3D gait analysis, and dynamic electromyography. Complications are uncontrollable weakness and increased sensory dysfunction.

A special issue: Therapy of neuromuscular hip instability

The most striking factor for sufficient therapy is movement of the joint. Supporting sensomotoric development, muscle strength and coordination as well as inhibition of pathologic muscle activity may improve the musclar balance at the joint. Orthoses, standing and walking aids support active and passive movements. They may prevent secondary defects due to positioning in non-walking patients.

Intramuscular injections of Botulinum Toxine as an adjuvant therapy may help to reduce spasticity and dystonia in order to reach movement therapeutic, orthetic, but also surgical goals easier or by optimized timing.

As conservative options are not sufficient soft tissue surgery may prevent hip dislocation in a considerably high percentage.

Combination of exactly indicated surgical procedures, parts of a *multi-level-surgery*, and the following rehabilitation period with movement therapy and orthoses are crucial for the patient´s benefit.

Intertrochanteric varization and derotation osteotomies of the proximal femur are indicated to improve biomechanics of the hip joint, correcting an excessive antetorsion and treat a severe gait dysfunction. Combinations with multi-level-soft-tissue-surgery result in encouraging longterm outcomes.

Complex reconstructions (see Fig. 3) of dislocated hip joints include multi-level-soft-tissue-procedures, femoral and pelvic osteotomies and open reduction. Walkers may profit by walking with a stable hip joint, non-walkers may profit by painfree weight.bearing and standing possibility for transfers. Most important factors for an excellent outcome are peri- and postoperative caring and movement therapy (Strobl 2004).

Femoral head resection is a palliative procedure for gaining painfree hips without stability for transfer standing. Complicattions include heterotopic ossifications, extensive resections and muscle flap interpositioning are recommended.

Total hip arthroplasty is indicated in walking cerebral palsy patients with arthritic, subluxates or dislocated hip joints and sufficient muscular stability.

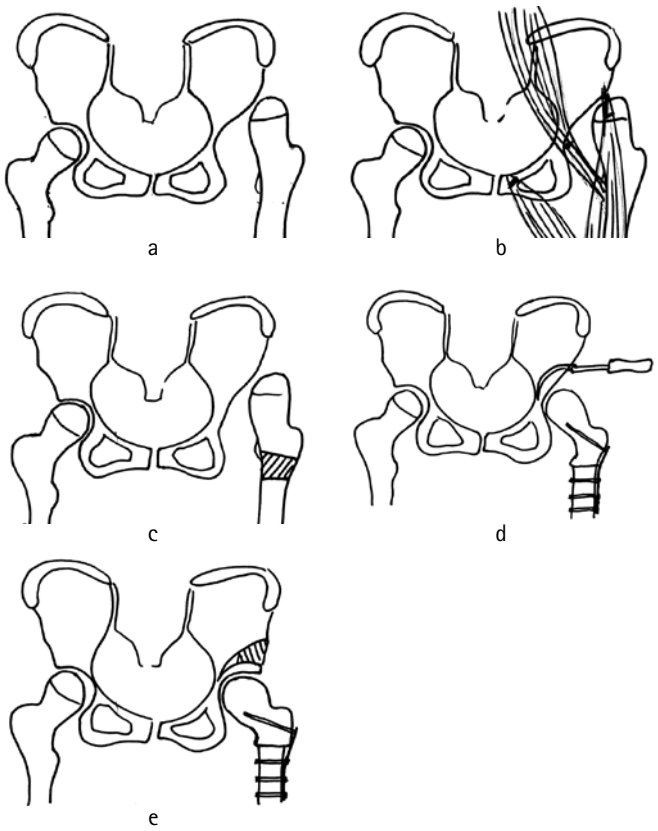

Fig. 3 Neuromuscular hip dislocation (a): reconstructive surgery by muscular lengthening (b), proximal femoral derotation-varisation-shortening osteotomy (c), open reposition acetabuloplasty (d) and femoral bone interpositioning and capsular suture (e).

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