REPORT
Professional Assignment Project 3.1/3.2

Production of a brochure for children related to physiotherapy

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1. Introduction

Children often don’t know what physiotherapy is and what they have to expect from such a treatment. Also they might not understand why physiotherapy is important for them. This can interfere with the treatment and especially the treatment goals, because if they don’t see the importance and need to follow the treatment they might be unmotivated and for example not do homework exercises the physiotherapist asks them to do. With the brochure we want to address those problems.

The goals of this report are to explain the medical background of the brochure’s content and to discuss new developments. It will inform the reader - parents, health professionals and people interested in more detailed and scientific background material - about the developmental process of the brochure containing a scientific explanation of the brochures content and the evidence based background literature. Found evidence is used to justify the physiotherapeutic modalities described in the brochure. The content of this report is mainly based on guidelines, the clinic’s protocols and scientific articles. Furthermore we used books describing the standards of paediatric physiotherapy.

In the Asklepios clinic, there is no patient education material concerning physiotherapy especially addressing children. After discussion with our client, Sabine Steinstraß (head of the physiotherapy department in the Asklepios clinic), we decided on the three main topics Juvenile Rheumatoid Arthritis, Cystic Fibrosis and Scoliosis, because those are conditions frequently treated in the Asklepios clinic.

“Juvenile rheumatoid arthritis (JRA) is the most common paediatric rheumatic disease and is a leading cause of childhood duality. Physical therapists play a crucial role in the treatment of these children and serve as essential members of the interdisciplinary treatment team” (Rhodes, 1991).

One in 2500 children born to white parents is diagnosed with Cystic Fibrosis. Today patients commonly reach their 30th to 40th (Campbell et al, 2006). Physiotherapy plays an important role in the multidisciplinary approach of Cystic fibrosis. The Physiotherapeutic treatment starts early in life and has to be adapted according to changing situations from infancy to adulthood (Prasad et al, 1999).

Infantile scoliosis, at the age of four and 10, compromises for 10-15% of all idiopathic scoliosis. Physiotherapy for scoliosis is depending on the way the patient presents. Most commonly used interventions are probably exercise therapy, electrical stimulation and orthotic management. In the extreme cases where operation is necessary physiotherapists will have to be involved in the recovery process of the children.

During the developmental process of the brochure, we had to keep the adequate presentation of medical information to children in mind. Therefore the first part of this report will cover the communication between health professionals and children, followed by the background information regarding paediatric physiotherapy in general, Juvenile Rheumatoid Arthritis, Cystic Fibrosis and Scoliosis.
2. Communication with the patient - writing a patient leaflet

Instructional and informational leaflets are integral aspects of patient self-management plans, they provide important information for the patient in a cost-effective way for the health professional (Wilson & Park, 2008). Before starting to write a patient leaflet, four points have to be clear:

1. which professions have to be involved
2. why the leaflet is needed
3. the content that has to be conveyed
4. the design has to be conveyed

These concepts are important because the understanding of the addressed population is related to the clarity of the communication (Ivnik & Jett, 2008). It is best when professionals are involved who identified the need for that leaflet. That also includes assistants who are involved in dealing with the addressed patient group (Ivnik & Jett, 2008).

The purpose of a brochure should be clear, it can be descriptive - meaning it describes a procedure -, focused on management of a condition, skill-specific, instructive - meaning it informs the reader about the optimal use of a tool - or a decision making tool meant to help the patient and his family to make a decision (Ivnik & Jett, 2008).

The content of a leaflet should be written in an easy way to understand and remember for the patient (Wilson & Park, 2008). Active language should therefore be used, it is less formal and more direct. Using a direct approach like saying ‘I’, ‘You’ or ‘We’ to directly address a patient and using personal pronouns creates the effect of direct communication. The writer should try to write as if he would speak to the patient.

For the patient it is not necessary to know everything that might be nice to know, but provide the reader with information he needs to know.

In case medical jargon has to be used, it needs to be explained to not leave the patient with unclear information. The information should be free of most medical terms. Information about skills should be written in a step-by-step manner so that the patient feels safe in performing the skill (Ivnik & Jett, 2008).

To write a leaflet for children about what to expect from the Physiotherapy treatment, all sentences have to be short and should contain information that is essential. Children will not understand sentences that give them possibilities of how things might go. Such sentences should not be constructed like ‘either this or that might be a reason’, but rather say ‘this is the reason for’. It is helpful to construct a story around a problem so that the child hears something that is similar to his situation and will identify with it.

Pictures and Illustrations can be used if they clarify the text they are added to, but should be left out if there is no direct connection (according to Petra Tillmann; kindergarden teacher, specialised in children with special needs).
3. Paediatric Physiotherapy

Physiotherapy is defined as a systematic method of assessing musculoskeletal, cardiovascular, respiratory and neurological disorders of function, including pain and those of psychosomatic origin and of dealing with or preventing those problems by natural methods based essentially on movement, manual therapy and physical agencies (Eckersley, 1993).

Physical therapy modalities are used to correct, prevent and alleviate movement dysfunction of anatomic or physiologic origin. Paediatric Physiotherapy is a specialty concerned with maintaining health and providing care to children from neonate to adolescence.

Physiotherapy nowadays works following “The International Classification of Functioning, Disability and Health” (ICF) (WHO, 2009). The ICF is a form to report the patients need on more then just physical level. It includes impaired body functions as well as Body structures. Furthermore it reports activities and participation information mentioned by the patient (WHO, 2009) In Germany this approach is not yet commonly used, and for the brochure the ICF does not play an important role.

The brochure aims at children in the Asklepios Clinic in Germany and children who receive treatment are most likely in an acute stage of their disease, meaning that the Physiotherapists at the Asklepios Clinic are less likely to work with the participation problems of the children. This work will be taken over by Physiotherapists working in a rehabilitation setting or private practice.

The brochure will use the word “Krankengymnastik” instead of Physiotherapy. “Krankengymnast/ Krankengymnastin” is the old German term, still frequently used for Physiotherapy. There is no difference in the meaning of the two words. The term “krankengymnmatik” was replaced 1994 due to the adjustment to the international linguistic use and the reunion of east and west Germany. In the “DDR” the term physiotherapy was already commonly used before the reunion. (Rudolph, 2006)

The decision to use the term "Krankengymnastik" is based on a discussion we had with our client. We decided together that children would understand the word “Krankengymnastik” better.

Additionally in Germany the majority of people use the term “Krankengymnastik” and therefore the term will be used to prevent confusion and misunderstanding.

3.1 Goals of Paediatric Physiotherapy

The overall goal of paediatric physiotherapists is to encourage the child to the best abilities in order to live the fullest life possible (Eckersley, 1993).

In physiotherapy the same constellation of impairments can lead to different setting of goals. They can focus more on activity improvements or participation improvements (Campbell et al, 2006). Goals can be set as prevention of worsening of the situation as well as ameliorative (Campbell et al, 2006). The goals differ with every child, making it difficult to define general goals of Paediatric Physiotherapy, leading us to the conclusion that the specific goals will be mentioned with the specific pathologies following. For all goals it is important that risk factors are eliminated or reduced to a minimum (Campbell et al, 2006).

3.2 Physiotherapeutic working field

The working field of paediatric physiotherapists is very broad. They work in all kinds of settings like hospitals, clinics, health centres, child assessment and support units,
nurseries, preschool, special and mainstream schools and at the children’s homes (Eckersley, 1993). The therapist is involved in the management of children with physical and learning difficulties, the acutely and chronically ill child and children sustaining injury or with joint and soft tissue problems (Eckersley, 1993). The treatment can start with the neonate and might extend to adolescence. Most of the times the Physiotherapist is part of a multidisciplinary team which meets the personal, clinical and statutory needs of a child (Eckersley, 1993).

The family situation, the child’s environment as well as the cultural background and the motivation for treatment are important information for the paediatric physiotherapist to ensure the treatment goals are specific for the child. Knowing the environmental circumstances is an important fact concerning the ICF form introduced by the WHO in May 2001. The ICF allows to produce records of the environmental factors influencing the Patients functioning during Active Daily Living (WHO, 2009). Knowing about the environment and participation problems of the patient will be important for setting long term goals for treatment.

Treatment will start with the gathering of information like the history and any medical data provided by other health professions. An assessment to determine the aims of treatment, a diagnosis and define long and short term goals for treatment will follow (Campbell et al, 2006). The assessment will be recorded so that the Physiotherapist can compare treatment outcomes to a baseline, and has the possibility to adjust treatment goals according to the changes the child experiences. If the needs of the child change, the goals have to be adjusted. This might mean that the treatment method has to change as well. The assessment will include observations of the movement pattern of the child as well as of specific joints, palpations and special tests (detailed information about the assessment is mentioned for each pathology). If necessary there might be an assessment of aids, equipment and the child’s normal living environment to improve the treatment outcomes.

A decision for the treatment mode and the design of the method will be made by the therapist. It might be that not the physiotherapist himself performs the treatment but another member of the care team or the parents who are taught by the therapist (Eckersley, 1993). Counselling is an aspect of Physiotherapy, in case family members have questions or are in need of information. In times where everyone uses the internet to get an idea it is even more important to clarify information to reduce the anxiety (Eckersley, 1993).

The Physiotherapist decides about a discharge from treatment in case therapy is not needed acutely (Campbell et al, 2006). The child will be in a program so it can be referred back to Physiotherapy by a health profession or family members to restart therapy at any needed time (Eckersley, 1993).

3.3 Recommendations for Physiotherapy

There are multiple reasons why a child might need Physiotherapy. There are short-term difficulties which will improve, long-term permanent problems which cannot be cured ranging from mild to severe or even life threatening (Burns & MacDonald, 1996). Early Physiotherapy can limit primary impairments therewith preventing activity limitations that might have occurred without therapy. Prevention involves attempts to limit impairments from the lesion or disorder and help to develop appropriate functional abilities (Campbell et al, 2006). Early intervention should lead to a reduction in secondary impairments as well.

When limitations persist for long periods of time, children may fail to succeed in active daily living, meaning they have problems performing in roles like participation
at school, playing or within the family, these changes or problems on participation level should be mentioned in the ICF. Therapy aims on not just try to improve the physical condition but also to adjust the environment to the needs of the child. In this case therapy can help to find solutions and show modifications of the daily life so the child is able to fulfil its tasks. This approach will need a multidisciplinary team to ensure all parts necessary are covered. The family will be involved as well to establish a network for the child that can help him succeed situations (Campbell et al, 2006). In this case the Physiotherapist will work as counsellor and advisor to make sure the child’s needs are respected and transformed into treatment goals to provide the best abilities in order to live the fullest life possible (Eckersley, 1993).
4. Juvenile Rheumatoid Arthritis

4.1 Pathophysiology

Juvenile Rheumatoid Arthritis (JRA) is the most common chronic childhood rheumatoid disease affecting the cartilage. JRA is divided into three subtypes, which are differentiated by the affected joints, by the type of antibodies found in the blood and other effects (Gould, 2006) and/or onset, namely systemic onset. JRA is also called Still’s disease (Gould, 2006), Pauciarticular-onset JRA and Polyarticular JRA (Effgen, 2005).

Even though specific tests might be helpful they are not diagnostic caused by the fact that the rheumatoid factor will not show up in children’s blood. Still leukocytosis and antinuclear antibodies as general signs of inflammation are frequently found in the blood of affected children (Gould, 2006).

The course of the disease might have remission and exacerbation periods and may last months or years. Inflammatory arthritis develops with the invasion of the synovium by either immune complexes or a putative antigen. This initiates the cascade of inflammation, while the lymphocytes lose their ability to distinguish between antigens and healthy tissue leading to the release of lysosomal enzymes and collagens into the joint fluid and herewith destroying the synovial membrane (Rhodes, 1991).

Systemic-onset JRA is the least common but most painful onset of JRA. ‘Patients with SJIA (Systemic Juvenile Idiopathic Arthritis) do not show signs of lymphocyte-mediated antigen-specific immune responses. The clinical signs of SJIA are rather associated with granulocytosis, thrombocytosis and up-regulation of acute-phase reactants indicating an uncontrolled activation of the innate immune system’ (Fosh & Roth, 2007). It comes along with high fever and patients might even present with heart infection, swollen glands, enlarged spleen, abdominal pain, anaemia and growth retardation (Effgen, 2005). There are two peaks in incidence, one being from 2-5 years and the second being from 9-12 years. Persistent arthritis is a prognosis in case arthritis continues after the systemic signs subside. This development can lead to severe deformities.

Both Pauciarticular-onset JRA and Polyarticular JRA do not include fever, but they are based on the distribution of the disease. Pauciarticular-onset JRA affects four joints six months after the onset of the JRA. The prognosis varies; permanent joint space narrowing is frequently seen after remission. Still many patients do not have severe chronic disabilities after remission (Effgen, 2005). Polyarticular JRA is defined as affecting five or more joints. The onset is between the 1st and 3rd year and again in early adolescence. A symmetrical pattern is often seen after the first year of the disease. The prognosis is moderate if started in childhood but poor if the disease is contracted in adolescence. 50% of patients develop severe destructive arthritis.

In all three types joint inflammation can involve few or many joints. The proliferation of the joint synovium leads to pannus (massive overgrowth) eroding the adjacent cartilage and bone. Joint adhesions and osteophytes are possible as well. Periods of inflammation can stretch and erode the ligaments leading to joint instability and ligamentous laxity. Surrounding muscles and tendons might get inflamed as well and cause restriction in ROM by the intensity of pain. A decreased ROM causes articular cartilage damage and compounds destructive forces. The normal skeletal alignment can be destroyed, leading to additional destructive forces on the body (Effgen, 2005).
Overgrowth, stimulation of adjacent growth plates or early physeal closure and subsequent limb shortening can result from inflammatory hyperaemia. Leg length discrepancies are a common effect of the subsequent limb shortening. It occurs more often in monoarticular and pauciarticular JRA. Overgrowth and leg length discrepancy diseases are commonly seen in the knee, if the disease onset is earlier than 9 years of age. It then arrests of the acetabulum and femoral head. The knee overgrowth does normally not exceed 3 cm. Is the diseases onset after the age of 9, premature closing of the growth plate and subsequent shortening with a discrepancy of up to 6 cm are seen more often (Effgen, 2005). Iridocyclitis (chronic eye inflammation) is a common symptom of JRA especially for systemic-onset JRA. Large Joints like knees and elbows are more often involved; cervical spine ankylosis is seen frequently.

Typical hand and wrist deformities are subluxation and radial deviation with ulnar deviation of the fingers (Rhodes, 1991). Extremely prevalent in children with JRA are foot deformities as well as differences in pressure distribution while walking. The most noted deformities are pronated rear foot and midfoot as well as toe valgus. ROM limitations and joint stiffening at the metatarsophalangeal joint affect the push off phase during walking. ROM limitations are common in the spine as well, in which radiographic changes show cervical spinal abnormalities, like subluxation or ankylosing, leading to limitations in extension direction (Effgen, 2005).

4.2 Etiology
The cause of JRA is unknown (Gould 2006). ‘The prevailing theory is that it is a autoimmune inflammatory disease activated by an external trigger in a genetically predisposed host’ (Campbell et al, 2006). This hypothesis is also supported by Rhodes (1991). It might result from any of a heterogeneous group of diseases as well as diseases like juvenile psoriatic arthritis, juvenile ankylosing spondylitis and other enthesitis-related forms of arthritis (Campbell et al, 2006). This information leads to the idea that JRA is not a single disorder but different factors might be responsible for different pathologies presenting as to the combined picture of JRA (Campbell et al, 2006).

Three hypothetical causes are a viral infectious agent, an immunological abnormality or a combination of the two (Effgen, 2005).

Pauciarticular-onset of JRA is the most common onset, it is most often diagnosed in the age of 2 and it affects girls 4 times more often than boys. The onset of systemic-onset JRA is between the 5th and 15th year and is equally distributed in girls and boys. Systemic-onset JRA is the least common form of JRA.

4.3 Assessment
Every examination will start with the history of the child, containing the onset of the disease, patterns of fatigue, pain and stiffness are asked to create a picture of the child’s day. Information about the independence of the child during active daily living as well as performance in a social environment like school, sports or any other recreational activities, are reported. If applicable, an analysis and evaluation of aids, splinting or exercise programs that are already present should be performed by the therapist (Rhodes, 1991).

It is very important that before starting the treatment an extensive assessment is done to realise the start of deformities, and to get a differential diagnosis on the patient’s condition. The assessment gives information of the causes of deformities
and helps to create a very patient specific treatment plan (Hüter-Becker & Dölken, 2005).
However an extensive assessment will not be possible during the acute stage of inflammation. The extensive assessment will be performed as soon as the acute stage has resolved (Burns & MacDonalds, 1996). During the acute stage, the assessment will include the musculoskeletal system to identify the major problems (Burns & MacDonalds, 1996). It is most important to decrease the pain, stiffness and swelling and to move the involved joints carefully out of their protective position to decrease the risk of deformities (Hüter-Becker & Dölken, 2005). The symptomatic relieve of pain; swelling and stiffness will be achieved by non-steroidal anti-inflammatory drugs (Burns & MacDonalds, 1996).
The assessment will also include the evaluation of functional skills, strength, bony deformities, ROM - observed actively and tested passively with goniometric measures - fatigue and pain (Effgen, 2005). The observation of the normal movement of the child is best done by looking at the way the child moves while playing, sitting down or dressing. If the child is very young, it might not allow getting touched by the therapist. Therefore it is important that the child and the family gain trust in the therapist. This can be done by talking to the parents while playing with the child. It is important that the child trusts the therapist to allow the therapist to touch the painful joints, and do special tests.
A joint inspection with a comparison of both sides will be performed to compare swellings, check for atrophies, bursitis or growth interferences. The palpation will give more information about inflammation of joints and tendons, and the muscle tonus can be tested as well. The range of motion of all joints should be tested quickly to detect even little variances as early as possible and prevent primary or secondary limitations. The joints are tested actively and passively to test the free range of motion.

**Table 1: Checklist for assessment** (Hüter-Becker & Dölken, 2005, p. 411)

<table>
<thead>
<tr>
<th>Joint</th>
<th>Assessment Details</th>
</tr>
</thead>
<tbody>
<tr>
<td>Jaw</td>
<td>Opening of the mouth: 3 vertical fingers of the child</td>
</tr>
<tr>
<td>Cervical Spine</td>
<td>Extension: face close to horizontal</td>
</tr>
<tr>
<td>Shoulder</td>
<td>Flexion: 180° humerus without additional movements next to ears</td>
</tr>
<tr>
<td>Elbow</td>
<td>Flexion: 150° fingers should touch shoulder</td>
</tr>
<tr>
<td></td>
<td>Extension: often 10° hyperextension</td>
</tr>
<tr>
<td>Wrist</td>
<td>Dorsiflexion. 90° support yourself on your handy</td>
</tr>
<tr>
<td></td>
<td>Palmarflexion: 80-90°</td>
</tr>
<tr>
<td>Fingers</td>
<td>Flexion: making a fist</td>
</tr>
<tr>
<td></td>
<td>Extension: “star” wrist dorsiflexion, finger extension thumb 90° abduction, reposition and extension</td>
</tr>
<tr>
<td>Thumb</td>
<td>Abduction, reposition, extension 90°</td>
</tr>
<tr>
<td>Hip</td>
<td>Flexion: 150-160°, femur to abdominals</td>
</tr>
<tr>
<td></td>
<td>External rotation till 90° (with young children)</td>
</tr>
<tr>
<td></td>
<td>Internal rotation 50-60°</td>
</tr>
<tr>
<td>Knee</td>
<td>Flexion: 150-160° Calcaneus to buttocks</td>
</tr>
<tr>
<td></td>
<td>Extension: 5-10° hyperextension -&gt; long</td>
</tr>
</tbody>
</table>
sitting position when the knees touch the ground the heel is in the air

<table>
<thead>
<tr>
<th>Ankle</th>
<th>Plantarflexion 60°, Pronation:30-40° Toes II-V: equally on both sides, equal toe and forefoot pressure distribution on both feet.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Toe I</td>
<td>Dorsiflexion: MTP joint: 60-70°</td>
</tr>
<tr>
<td>Spine:</td>
<td>Thoracic spine: according to size of child 2-3cm separation Lumbar spine: 4-6cm separation</td>
</tr>
</tbody>
</table>

Findings of joint limitations are recorded. If muscle atrophies are visible, the circumference has to be measured and reported. A reduced muscle circumference relates commonly to joint dysfunctions. Deformities are named and described to establish a baseline measure for re-evaluations. For this purpose it is also useful to use a grading system.

Grading system (Hüter-Becker & Dölken, 2005, p. 413):
1. actively correctable
2. passively correctable, which can be hold in the corrected position actively
3. correctable passively but position can not be maintained actively
4. not completely correctable passively
5. fixed deformity

The lower extremities will be checked for axis and pressure lines as well. It is important to see if the axis and pressure lines can be changed actively. Additional information like x-ray, ultrasound and sonography are useful tools to detect discrepancies and inflammations.

After each acute stage has resolved there has to be a reported re-assessment. The treatment goals and priorities might change after the assessment, according to the changes.

**Specific assessment of the different levels of impairment**

**PARTICIPATION AND ACTIVITY RESTRICTIONS**

The impact of the disease on participation and activities of the child depends on the extent of the disease, the child’s resiliency and desire to be independent. The exact type of disease plays an important role: a child with Pauci-JRA may have few functional limitations while a child with severe poly-JRA needs assistance with ADL’s. The participation of the child will depend on the quality of aids, equipment and available services (Campbell et al, 2006).

**JOINT STRUCTURE AND FUNCTION**

Swelling, end range stress pain and stiffness are the major signs of inflammation. Swelling of a joint might be due to intra-articular effusion, synovial hypertrophy, and soft tissue oedema or periarticular tenosynovitis. A joint might also be enlarged due to bone overgrowth or increased blood flow to this area. Swelling and muscle spasms create pain and inactivity. Stiffness is most often recognised in the morning and after
long periods of sitting. If the inflammation is chronic, joint deformities occur. Joint swelling and limitations can be marked on a stick figure (Campbell et al, 2006).

**Muscular Structure and Function**

Muscle atrophy and weakness are most often observed close to inflamed joints, but might also occur further away. They can persist over a long period of time after remission of the arthritis. Common patterns are: weakness in the hip extensors and abductors, knee extension, plantar flexion, shoulder abduction and flexion, elbow flexion and extension, wrist extension and grip.

Muscle bulk circumference, strength and endurance should be tested regularly. In young children muscle strength can be assessed by performing age specific functional tasks. In older children isometric manual muscle testing will be done in an acute stage when the child has pain while moving the limb against resistance. When there is no sign of inflammation dynamic muscle testing can be performed. Endurance can be tested through repetition of movement (Campbell et al, 2006).

**Aerobic Capacity and Function**

Impaired aerobic fitness does not appear to be significantly related to the severity of the disease, but may be due to hypoactivity secondary to disease symptoms. Physiological factors like anaemia, muscle atrophy, generalized weakness and stiffness result in poor mechanical efficiency. According to Takken et al (2001), the 6-minute walking test is a useful tool to measure waking tolerance in children.

**Pain**

Pain is the major cause for restriction in children with JRA. Children above the age of 7 can use a numeric rating scale, horizontal word graphic scale or a visual analogue scale to evaluate their level of pain. For smaller children the Varni/Thompson Paediatric Pain Questionnaire will provide the best outcome. It includes both parents and child reports (Campbell et al, 2006).

**Growth Disturbances and Postural Abnormalities**

A retardation of linear growth is seen with extended periods of active disease and is exacerbated by the long-term use of steroids, puberty and secondary sex characteristics. Osteopenia may be due to inadequate bone formation for age, low bone turnover and depressed bone formation. Increased blood supply to the inflamed joints might lead to bony overgrowth (Campbell et al, 2006).

Leg length discrepancies are common in unilateral knee arthritis and can lead to hip asymmetries as well as thorax misalignment. The therapist has to check the knee line, trochanter level, spina iliaca posterior superior, level of the crista iliaca and the spine. If the leg length difference is more than 0.5 cm, the difference has to be adjusted with orthopaedic shoes (Burns & MacDonalds, 1996). The child might develop a functional scoliosis. Premature closure of the growth plates may also occur. The child’s posture in standing might be affected by hip and knee flexion contractures, genu valgus and foot deformities. Muscle imbalances might occur due to postures adopted to relieve the pain (Campbell et al, 2006).

**Gait Impairments**

Gait anomalies are a very common symptom which should be taken care of during assessment to find the cause of the anomaly. Specific attention should be drawn to a decrease in velocity and cadence which might be due to stiff sluggish joints. The decreased stride length may relate to decreased hip extension and decreased
plantar flexion at the end of the stance phase (Burns & MacDonalds, 1996). A anterior pelvic tilt is recorded commonly. This deviation occurs due to tight hip flexors and weak abdominal muscles (Burns & MacDonalds, 1996).

4.4 Treatment

**GOALS (EFFGEN, 2005; CAMPBELL ET AL, 2006)**
- Family education
- Pain management
- Deformity prevention
- Promotion of functional abilities
- Physical fitness
- Maintain or improve function

Treatment should always be below the level of pain. The grip should be close to the joint and soft. Little children do not normally describe pain verbally; therefore it is important that the therapist realizes the changes in the face and body language of the child. The position to start with should be chosen in a way that the inflamed joints are not under pressure.

**PAIN MANAGEMENT**

The treatment should start with interventions to reduce the pain (Hüter-Becker & Dölken, 2005). Short hydrotherapy sessions are very effective in the acute stage. The warmth and buoyancy can relieve the pain and alleviate movement through relaxation of muscle spasms. The child can maintain flexibility as well as strength, fitness and functional performance (Burns & MacDonalds, 1996).

Before starting the treatment it might be helpful to put the joints in the most relaxed and least painful position. During the acute phase, all involved and inflamed joints should be positioned on towels, pillows and cuddly toys till they are able to overly pain free. Passive and active movements of the joints in a protective position, starting from the least painful position with frequent repetitions, decrease the pain, leading to improved movement. During the acute stage, all involved joints should be moved passively or active-assistive out of their protective position. If possible, the child should try to contract the muscle to hold the corrected position. If the child has heart problems during the acute stage this has to be taken into consideration of the child’s general fitness.

Traction of the large joints (e.g. hip), eventually intermittent traction, can reduce pain. It should not be performed if the child mentions pain while the movement is performed.

Taking away the weight of the extremities by using a so called “Schlingentisch” improves movement, as well as improving metabolism to the specific joints, leading to decreased pain.

Superficial heat applied with a hot pack or warm moist towels for 20 minutes before exercise can reduce muscle spasms and improve muscle extensibility through the increased blood flow to the specific area. Stiffness and pain of the hands might be reduced with Paraffin wax “painted” on the hand. “Painting” wax on the hands makes treatment more related to the activities children like.

Chyrotherapy is used to reduce inflammation. Therefore cold packs are applied for 10-15 minutes on the hot and inflamed joint. A few minutes are enough to reduce
pain. It is important that hyperaemia leading to overheating of the joint does not occur.

Electrotherapy, especially interferential and high intensity tens, is also used to reduce pain. Tens machines can be taken home and are easy to apply. The child and family can be trained how to use the machine. Massage can be used to reduce muscle spasms. It can break up fascial adhesions (Hücker-Becker & Dölken, 2005).

Balanced rest and exercise programmes, restful sleep, resting splints, exercises after a warm bath, and sleeping in a sleeping bag to keep the body warm all reduce pain and morning stiffness. These interventions also improve well being of the child (Campbell et al, 2006).

**MANAGEMENT OF JOINT IMPAIRMENTS**

Joint mobilisations improve the movement. Passive or active assisted movements improve the range of motion. The treatment is very close to the pain barrier but never above the threshold. It will take some time till improvement will be visible (Hüter-Becker & Dölken, 2005). On a daily basis, once to twice a day, each arthritic and adjacent joint should be moved through its ROM (Campbell et al, 2006).

Traction with sliding component can be performed according to Kaltenborn (1982), tightening the soft tissue surrounding the joint to prevent subluxation.

Stretching of the hypertone musculature is performed passively in little children. In older children, active stretches are performed as well. It is important the the joint is in a relaxed position before starting the stretch and the passive stretch should always be performed with a soft and wide based grip, close to the joint to prevent subluxation. Stretching can be achieved by positions like lying prone for 30 minutes. This will lengthen the flexor muscles of the lower limb by making use of gravity (Campbell et al, 2006). To activate the hypotone muscles, the child should try to hold the stretched position with an isometric contraction. With this technique the child learns to activate inactive muscles and correct his own false position first static and after some time the child will be able to move out of the false position through the new gained range of motion. In addition the child has to be taught to use the new gained range of motion in his daily life. It is important to break up complex movements to be able to correct the child if it wants to move again in his old pattern. The new, physiologically normal pattern has to be trained to enable the child to use it constantly.

Parents and child will be educated in a home programme which is discussed with both the children and parents to find solutions on how to integrate the treatment into the child’s normal day. The home programme can be a cause for arguments between child and parents. This should be avoided as far as possible and the child should be able to decide together with his parents how the treatment will work best. If the children are younger, parents will be trained in handling techniques by the therapist so that they are able to perform a few exercises at home.

Additionally functional and positional aids, orthotics, orthopaedic shoes, tobbi collar, jaw splint as well as walking aids or support like a tricycle for little children and a bike for older children might be necessary and helpful tools to give the child the greatest independence possible (Hüter-Becker & Dölken, 2005).

**TREATMENT AFTER OPERATIONS**

Operations are carefully planned and aim at preserving joint health. In older children reconstructive surgery may relieve pain and restore function. ROM exercises start 48 hours after the operation unless there are any complications. For operations of the
lower extremity, gait training starts as early as possible, long periods of sitting should be avoided, the patient should lay prone as much as possible. As soon as the soft tissue inflammation resolves, a strengthening programme can be started (Campbell et al, 2006).

**HYDROTHERAPY**

During a hydrotherapy session, gait anomalies and their causes can be addressed easily. Due to the facilitated relaxation of the soft tissues a greater velocity can be gained (Burns & MacDonalds, 1996). In children who had exercised in water for 6 weeks, hip rotation angles were significantly improved and other ranges of motion were also enhanced (Bolukbas, 2005).

**STRENGTHENING AND AEROBIC EXERCISES**

Exercises for strengthening should be performed for the muscles surrounding and supporting arthritic joints. During the acute phase, isometric contractions are recommended. The child is taught to hold the contraction for up to 6 seconds. It is important that the child exhales during contraction and inhales during relaxation. It is sufficient to perform five to ten repetitions a day to maintain muscle bulk and strength. If necessary, EMG biofeedback might be used to train the child in regulating the intensity of the contraction, which should be sub maximal (Campbell et al, 2006). Strengthening programmes during the chronic stage should include upper and lower abdominal muscle training to increase postural stability. Progressive resisted exercises for all involved joints should be performed to increase the child’s functional ability and fitness (Burns & MacDonald, 1996). Eccentric and concentric exercises, functional movement patterns and programmes like Pilates, Yoga or Tai chi can be included in the programme to increase the variety of the training. Resisted exercises have been shown to improve contraction speed, functional status, disability, and performance of timed tasks (Campbell et al, 2006).

Each training should start with light aerobic and flexibility exercises. The session should be finished with a cool down and stretching. Twice a week resistance exercises can be performed. Enough time should be planned in between these sessions to allow time for rest and recovery (Campbell et al, 2006).

Exercises performed to improve aerobic capacity should be moderate in intensity and should not last more than 30 minutes per day (Bolukbas, 2005). Extensive aerobic exercises like dancing, cycling, swimming and others, result in improved functioning of cardio vascular and respiratory systems (Bolukbas, 2005). A heart rate of 60%-85% of the maximum should be acquired twice a week for at least 30 minutes. Intensity, duration and frequency seem to be more important in improving aerobic fitness that the mode of exercises (Campbell et al, 2006; Klepper, 2003).

Weight bearing exercises are vital for bone density and bone growth. Therefore children at the expected age should be encouraged to walk (Campbell et al, 2006).

**INDEPENDENCE AND RECREATIONAL ACTIVITIES**

A primary goal is that the child has the greatest possible self independence. The therapist should be informed about the activities of the child and help with making decisions on how to adjust the child’s environment, like school. It might be necessary that the child uses two sets of books, one at school and the other one at home. Parents, the child and the physiotherapist should discuss these topics together to find the best way (Campbell et al, 2006).

The child should also be encouraged by the Physiotherapist to participate in recreational and endurance activities to improve fitness, functional performance and
socialization (Burns & MacDonalds, 1996). High impact sports like soccer, hockey or basketball should be avoided, because of their high impact on the inflamed joints as well as their high risk of injury (Campbell et al, 2006).

4.5 Evidence based discussion
Juvenile Rheumatoid Arthritis is nowadays called Juvenile Idiopathic Arthritis, due to the fact that the underlying cause is still unclear. The diagnostic picture for Juvenile Rheumatoid Arthritis differs a lot from the picture of Rheumatoid Arthritis in adults which is another factor why the name changed to idiopathic arthritis. In the brochure we will use the term Juvenile Rheumatoid Arthritis. This is the term used at the Asklepios Klinik when giving a diagnosis to a child and the family. According to our Client it is easier for children to have a name for a disease, meaning children would say "I have Rheuma" instead of saying “I have an idiopathic disease”. The term is integrated into the children’s lives so we decided to stick to it for the brochure and the report. Campbell et al (2006) state that ‘deep heat, including ultrasound and short-wave diathermy is not used for pain control in JRA’ while Hüter-Becker & Dölken (2005) state that the thermal approach of the Ultrasound has to be eliminated in children to protect the growth plate. This is done with a pulsed setting which showed to be very effective in flexion synovitis of the fingers and tendinopathies. The lack of evidence about treatment techniques makes it difficult to prove either one or the other statement right or wrong. These two statements are kind of combinable by the fact that even Hüter-Becker & Dölken know about the negative effects of ultrasound for the growth plate and therefore describe the way a setting can look if electrotherapy is the chosen intervention. It is difficult to describe the best way of practices in this field for the simple reason that research often does not prove effectiveness of treatment techniques. The used authors otherwise have the same opinion about the way a treatment should look like.
Rhodes (1991) as well as Frosh and Roth (2007) state that with new insights in the pathophysiology of the disease, new ways of treatment will develop and medical prognosises will be more precise. The new insight would mainly change the medication and medical treatment the patients will receive.

4.6 Conclusion
Juvenile Rheumatoid Arthritis is a live changing condition. Children being diagnosed with JRA experienced a lot of pain already; they go through many medical examinations and a lot of hospital stays. This makes children and parents afraid of what is coming next. The children are as well afraid of more pain due to treatment, therefore the brochure tries to tell the truth. It says that pain might be experienced but it also gives an explanation why it is important to go through a little bit of pain to get better afterwards. The “Schlingentisch” might seem very scary to children therefore the explanation of floating like an astronaut decrease the fear of the children. In JRA the Physiotherapist will not just be in the hospital but children will go on with Physiotherapy after they leave the hospital. The brochure also tries to inform parents about what is normal during Physiotherapeutic treatments; it might be very difficult for parents to see that their children are suffering pain.
The brochure contains information and instructions for the parents which might be helpful for the parents as soon as the child can leave the hospital and go back home. The aim is to get parents involved in the treatment and help the family to not feel helpless even though the child is in a lot of pain. This will lead to a more relaxed family situation also improving the mental status of the child.
5. Cystic Fibrosis

5.1 Pathophysiology
Cystic fibrosis (CF), also called mucoviscidosis, is a common inherited disorder. The gene responsible for the disease is located on the seventh chromosome which is related to a protein involved in the chloride ion transport in the cell membrane, the cystic fibrosis transmembrane conductance regulator (CFTR) protein. The abnormality in the CFTR protein leads to abnormalities in the cell membrane, namely chloride impermeability and sodium hyperpermeability. This change in permeability induces an abnormal amount of fluid being removed from the airway lumen.

The primary feature of CF is the obstruction of the mucus-secreting exocrine glands by hyperviscous secretions (MacLusky et al, 1987) which inhibits the delivery of exocrine gland products to target tissues and organs creating abnormalities in these systems (Campbell et al, 2006). Effects are seen in the lungs and pancreas where the sticky, tenacious mucus obstructs the passages, as well as reproductive system, sinuses and sweat gland involvement.

Respiratory manifestations of CF are primarily caused by the abnormal expression of CFTR protein in the airway epithelial cells (Campbell et al, 2006). The excessive mucus obstructs the airflow in the bronchioles and small bronchi, causing permanent damage to the bronchial walls by air trapping or atelectasis. Air trapping and atelectasis in the small bronchi cause ventilation and perfusion mismatching, leading to hypoxemia. Infections are a very common secondary problem. The stagnant mucus is an excellent medium for bacterial growth. This complication adds to the destruction of lung tissue. Bacteria which lead to pneumonia most often are Staphylococcus aureus, Haemophilus influenzae and Pseudomonas aeruginosa (Grossman & Grossman, 2005).

Bronchiectasis and emphysematous changes are seen frequently as fibrosis and obstructions advance (Gould, 2006). A combination of bronchiectasis and small airway obstruction reduces the vital capacity and tidal volume, resulting in decreased airflow volume at alveolar level, and a progressive increase of arterial carbon dioxide tension (PaCO2) leading to hypercapnic respiratory failure (Yankaskas, 1992). Respiratory failure is the most common mortality factor in CF accounting for 95% of death in CF.

Malnutrition of the respiratory muscles leads to impairment in those and has an impact on functionality of the respiratory system. Hypoxemia over longer times may result in artery hypertension and cor pulmonale (right sided heart failure).

In the upper respiratory tract sinusitis is a moderately seen symptom. In the advanced stage of pulmonary disease hypertrophic pulmonary osteoarthropathy is most noticeable in the rounded hypertrophic changes in the terminal phalanges of the fingers and toes (Campbell et al, 2006).

The first indicator of abnormality in the digestive tract is meconium ileus, presenting in 20% of infants with cystic fibrosis in the new born. It occurs when the meconium becomes inspissated and obstructs the distal ileum (Hekmatnia et al, 2005). In some older patients a distal intestinal obstruction might be seen, caused by abnormal intestinal secretion and increased adherence of mucus in the intestines. Obstruction of the pancreatic duct in utero by the viscosé secretion leads to periductal inflammation and fibrosis causing the loss of the pancreatic exocrine function causing itself malabsorption and pancreatic insufficiency. (Campbell et al, 2006).

Pancreatic insufficiency causes malnutrition through the malabsorption of nutrients which leads to insulin dependent diabetes in 15% of adults with CF. Poor nutrition
has a negative influence on the pulmonary course, therefore a high calorie diet as well as pancreatic enzyme replacement therapy is needed to maximize gastrointestinal function (Effgen, 2005).

Infertility in males is seen in 98% of cases (Wlischeski et al, 1996). In females the fertility rate is 20% less than in non affected females (Flume & Yankaskas, 1999). It is caused by the obstruction of the vas deferens in males and obstruction of the cervix in females (Effgen, 2005). Puberty is often delayed in both sexes (Grossman & Grossman, 2005).

5.2 Epidemiology
Annually about 1000 children in the United States of America are diagnosed with Cystic Fibrosis. The disease is much more common in the white population from northern Europe. A high proportion of asymptomatic carriers are common in these populations (Gould, 2006).

The cystic fibrosis transmembrane conductance regulator (CFTR) protein is located on the seventh chromosome and transmitted as an autosomal recessive disorder. The protein has a high incidence on the membrane surface of epithelial cells of the respiratory, gastrointestinal, reproductive and sweat glands (Collins, 1992; Campbell et al, 2006). More than 1000 possible changes can occur in CFTR to cause Cystic Fibrosis. Approximately 70% of all cystic fibrosis patients have the same defect, called F508 which causes the deletion of three bases leading to the loss of the protein phenylalanine (Grossman & Grossman, 2005). The defect can be diagnosed prenatal and in carriers with reliable results, this is done if a family history of Cystic Fibrosis is present.

5.3 Assessment
The goals of the assessment of a patient suffering from Cystic Fibrosis are: identifying individual problems and needs, provision of information necessary for the formulation of treatment plans and the evaluation of treatment intervention. Key points for the assessment are the evaluation of respiration, including also changes of chest and posture, evaluation of the vital capacity and thorax mobility. ‘Direct observational methods are especially useful in order to make an accurate characterization and quantification of energy expenditure’ (Klijn, 2003). The KNGF guidelines on COPD recommend that ‘in addition, patients’ personal goals and expectations should be formulated, and their willingness, motivation, confidence in the ability to succeed or barriers against engaging in behavioral change should be determined’ (KNGF, 2008). According to the CFT, ‘patients should be reviewed at regular intervals and treatment regimens adapted according to their changing needs’ (CFT, 2002).

5.4 Treatment
Cystic Fibrosis is a chronic disorder and therefore the disease can’t be treated causally but symptomatically. Goals of the physiotherapeutic care are the removal of excess bronchial secretions, the relief of the respiratory musculature and the maintenance of thoracic mobility. Those goals contribute to the main goal of maintaining the best possible pulmonary condition (Hüter-Becker & Dölken, 2005). ‘The role of the physiotherapist is not limited to airway clearance but also includes encouragement and advice on exercise, posture and mobility, inhalation therapy and,
in the later stages of the disease process, non-invasive respiratory support’ (Prasad et al, 2000). Furthermore the physiotherapist tries to maintain the patient’s participation level as high as possible, as the disease progresses. He can give advice on possible recreation activities and adjust the functional training according to the patient’s condition.

Airway clearance techniques build an essential part in the physiotherapeutic treatment. ‘By removing obstructive secretions they aim, in the short-term, to reduce airway obstruction, airway resistance and improve ventilation. In the long-term chest physiotherapy aims to delay the progression of respiratory disease and maintain optimal respiratory function, as removal of mucopurulent secretions may help to reduce the elastase mediated damage to the airways’ (Cystic Fibrosis Trust, 2002).

The Cystic Fibrosis Trust (CFT) guidelines recommend the following airway clearance techniques: Active cycle of breathing techniques, autogenic drainage, modified autogenic drainage, oscillating positive expiratory pressure (Flutter, Cornet), high frequency chest wall oscillation, intra pulmonary percussive ventilation, positive expiratory pressure, high pressure positive expiratory pressure, postural drainage and percussion. The techniques frequently used in the Asklepios clinic will be explained in more detail.

5.4.1 Airway Clearance Techniques

**ACTIVE CYCLE OF BREATHING**

The active cycle of breathing consists of a combination of thoracic expansion exercises, forced expiratory technique and breathing control. It is the most commonly used airway clearance technique in the treatment of Cystic Fibrosis. ‘This technique has been shown to have beneficial effects on both airway clearance and lung function’ (CFT, 2002). The patient starts usually with gentle breathing at tidal volume using the lower chest. As a reference, the patient can be asked to lay his hand on the abdomen. Thoracic expansion exercises consist of several deep breaths, expanding the lower chest, an inspiratory hold and ending with a quiet expiration. ‘Lung volume is increased, reducing resistance to airflow within the distal airways and collateral channels, allowing air to assist in the mobilisation of secretions’ (CFT, 2002). During this technique the physiotherapist or the patient himself can apply chest clapping to further assist the mobilisation of secretions. Forced expiratory techniques mobilize peripheral secretions which can then be cleared with a huff or cough. However, those techniques can lead to an increased airflow obstruction.

**AUTGENIC DRAINAGE**

The autogenic drainage is an unassisted technique which allows the patient to clear secretions without the help of a therapist. It uses expiratory airflow to mobilize secretions in the peripheral airways and moves them centrally. The technique consists of three phases (McCool & Rosen, 2006):

1. “Unsticking” the mucus in the smaller airways by breathing at low lung volumes
2. “Collecting” the mucus from the intermediate-sized airways by breathing at low to middle lung volumes
3. “Evacuating” the mucus from the central airways by breathing at middle to high lung volumes.
At the end of the technique, mucus is cleared from the large airways with a huff or a cough. ‘There is some evidence to suggest that autogenic drainage is as effective as other airway clearance techniques’ (CFT, 2002). McCool and Rosen (2006) recommend that ‘autogenic drainage should be taught as an adjunct to postural drainage as a method to clear sputum because it has the advantage of being performed without assistance and in one position’.

**MODIFIED AUTOCGENIC DRAINAGE**
The modified form of the autogenic drainage places less emphasis on the three phases of the original autogenic drainage and allows therefore a better teaching and learning. Modified Autogenic Drainage is often used together with inhalation therapies.

**OSCILLATING POSITIVE EXPIRATORY PRESSURE**

**Flutter**
The Flutter is a commonly used device in respiratory physiotherapy, mostly used in patients with chronic mucus retention. It is a small plastic device which contains a large ball bearing which repeatedly interrupts the outward flow of air. Expiration through the devise produces a controlled oscillating positive pressure, which results in mobilisation of secretions. According to Hüter-Becker and Dölken (2005), the Flutter should be used at least 15 to 30 minutes a day, with a small break after 10 to 20 inspirations. ‘Though some evidence exists to suggest that this device has a beneficial effect on lung function and sputum clearance in patients with Cystic Fibrosis other studies suggest no benefit over other techniques such as active cycle of breathing and it may be less effective in terms of maintaining pulmonary function’ (CFT, 2002).

**Cornet**
The main difference between the Flutter and the Cornet is that the Cornet is a gravity independent device and can therefore be used in any therapeutic position. The expiration volume passes entirely through the inner tube and is converted into pressure and flow oscillations. According to the CFT, the device should be used 10 to 15 minutes and can be introduced from the age of two years.

**POSITIVE EXPIRATORY PRESSURE**

During positive expiratory pressure breathing, the patient breathes against a resistance via a facemask or mouthpiece. This technique promotes the mobilisation of secretions, increases lung ventilation and trains the respiratory musculature. The patient uses the device in a sitting position and is asked to breathe through the mask at tidal volume 10 to 12 times, followed by a period of huffs, coughs and breathing control. ‘In patients with CF, PEP is recommended over conventional chest physiotherapy because it is approximately as effective as chest physiotherapy, and is inexpensive, safe, and can be self-administered’ (McCool & Rosen, 2006).

**POSTURAL DRAINAGE AND PERCUSSION**
Postural drainage and percussion are the traditionally used techniques in the treatment of Cystic Fibrosis and are commonly known as conventional physiotherapy. ‘Postural drainage can be an additional intervention when large amounts of mucus are being retained’ (KNGF, 2008). It is a passive form of treatment and requires assistance by a therapist. It involves positioning to allow gravity to assist drainage of secretions from the peripheral to more central airways. According to Hüter-Becker and Dölken (2005), infants should, if possible, be treated on the lab of the therapist
while older children can be treated on a mat using a wedge to make use of gravity. It is also recommended that the posture should be held at least 3 to 5 minutes. The combination of postural drainage with manual techniques, such as vibrations and percussions can facilitate the mobilisation of secretions. Percussion involves single or double handed rhythmical chest wall percussion, applied comfortably with a cupped hand. According to the KNGF (2008), chest percussion and mechanical vibration are appropriate for patients unable to cooperate with active treatment. Postural drainage in combination with percussion is of great benefit especially in infants or children who cannot actively participate in the therapy. However there are some contraindications described by the CFT. Those include:

- osteopaenia or osteoporosis (low bone mineral density)
- uncontrolled thoracic pain
- rib fractures/flail segment.
- incisions
- implantable venous access devices

'In patients with CF, chest physiotherapy is recommended as an effective technique to increase mucus clearance, but the effects of each treatment are relatively modest and the longterm benefits unproven' (McCool & Rosen, 2006). The KNGF (2008) states that, ‘although the use of postural drainage, chest percussion or vibration, and positive expiratory pressure has not been unequivocally substantiated by the literature, various combinations of these techniques may be effective in individual patients. If these procedures have not proved effective after 6 sessions, their continued use is no longer meaningful’.

5.4.2 Exercise
Next to airway clearance techniques, exercise plays an important part in the physiotherapeutic management of Cystic Fibrosis. ‘It can increase cardio respiratory fitness, maintain the patient’s strength, endurance and mobility and promote a good body image’ (Rogers et al, 2003). The CFT (2002) states that every patient - from mild to severe disease - can exercise, but that exercise programmes must be ‘tailored to the individual, based on disease severity, level of fitness and patient preference’. The benefits of exercise include:

- increased cardiorespiratory fitness
- increased ventilatory muscle endurance
- decreased breathlessness
- enhanced sputum clearance
- improved body image through increased muscle mass and muscle strength
- improved morale
- enhanced quality of life

Exercise programmes should include endurance as well as strength training. The frequency and duration of training is different for every individual but should, according to the CFT, progress to 20-30 minutes 3-4 days a week. Because of the differences in relation to growth, muscles and fat, children should not exercise like adults. Exercises should vary and it should be ensured that joints are not subjected to repetitive stress. ‘Exercise programmes should be tailored to the individual taking into consideration disease severity, level of fitness and the patient’s preferences to
exercise activities’ (CFT, 2002). Furthermore, Klijn (2003) claims that ‘exercise programs should be primarily based on the preference of the individual child’.

5.5 Evidence Based Discussion
Based on the idea that ‘early intervention may both delay the onset and progression of lung disease and that establishing a routine early on in the child’s life may improve long term adherence’ (Cystic Fibrosis Trust, 2002), patients are traditionally treated twice a day. However, to this date, there is no evidence supporting a twice-daily routine. Prasad et al (2000) state that ‘daily airway clearance regimens are time consuming and burdensome’ and that ‘scientifically proven justification of therapy might serve not only to persuade patients and carers of the value of a routine which often does not offer immediate or obvious benefit, but also reassure therapists that their practice is worthwhile’. However several authors agree that a study, comparing treatment versus no treatment, would be of ethical concern and therefore such a study is very unlikely to be conducted.

Most patients admitted to the Asklepios clinic, are in an acute exacerbation period. Therefore the treatment goals are usually aiming on improvement in the short term. Because of that, we believe that a twice-daily routine is reasonable.

5.6 Conclusion
Children who suffer from cystic fibrosis face a life that is marked by many hospital stays, examinations and treatments. Therefore it is essential that as a physiotherapist we try to create the best possible treatment environment and make sure that the treatment remains interesting and motivating. The treatment has to be adjusted continuously as the child’s condition worsens. But also the child’s age is of great importance, because an older child will have different goals, especially regarding the participation level, than a younger child. An important task of the physiotherapist is to improve the child’s general condition and consequently preparing the child for sporting activities. ‘Exercise programs should be primarily based on the preference of the individual child’ (Klijn, 2003). Another important fact to consider is the psychosocial situation of the child. During work with cystic fibrosis patients, we often experienced that the parents are afraid that exercise could harm the child. Our brochure can have an educative effect for parents, as we explain that sport is not contraindicated but even promoting the condition of the child.

As mentioned before, children with cystic fibrosis face continuous medical treatment and are confronted with physicians on a regular basis. At times examinations and procedures can be very stressful, uncomfortable or even painful. Therefore it can happen easily that the children are afraid when being confronted with another health professional, the physiotherapist. Our brochure can take that fear and make children understand that it is an important treatment and that it is meant to help them also to manage their daily life.
6. Scoliosis

6.1 Pathophysiology
Scoliosis is a lateral curvature of the spine of at least 10°. It can either be structural or non-structural. With a primary structural scoliosis there is no flexibility of the spine while a non-structural scoliosis is flexible (Effgen, 2005). It is secondary to another problem like unequal leg length. 80% of the cases with structural scoliosis are idiopathic. A genetic factor might play a role in the idiopathic form. Girls are approximately 5 times more affected than males (Gould, 2006). Congenital scoliosis results from developmental defects, approximately 60% of all cases are girls. The curvature becomes greater during growth spurts and might come along with a rotational component.

Early effects are unequal hip and shoulder levels, and rotations of the vertebrae affecting the thorax and pelvis. It might also lead to rigid ribs restricting ventilation if the fixation and rotation is severe (Gould, 2006).

Five types of scoliosis can be recognised (Adams & Hemblen, 2001):

1. infantile scoliosis, a type seen in very young children which may either resolve to normal or become progressively worse
2. primary or idiopathic scoliosis, a well defined group of unknown causes arising in children
3. secondary structural scoliosis, a miscellaneous group in which the curvature is secondary to a demonstrable underlying disorder
4. compensatory scoliosis, neuromuscular scoliosis
5. sciatic scoliosis, a temporary deformity

INFANTILE SCOLIOSIS
It usually begins in the first year of life till the 3rd year as a simple curve. Mostly it is convex to the left. The cause is unknown. The radiographic examination shows if it either resolves or progresses (Mehta, 1972). It is more frequent in male infants and in many cases the curvature will resolve. If it does progress it will lead to severe deformities (Campbell et al, 2006).

IDIOPATHIC STRUCTURAL SCOLIOSIS
This type is the most common and most important type of scoliosis. It starts in childhood (3-6 years) called juvenile idiopathic scoliosis, or adolescence (adolescent idiopathic scoliosis) between the age of 10-12 and progresses until the cessation of growth is reached. Girls are more affected than boys.

There is a primary structural curve and secondary compensatory curves above and below the primary curve. A rotational component always goes along with this lateral curvature in which the body of the vertebra rotates towards the convexity of the curve while the spinous process rotates towards the concavity (Adams & Hemblen, 2001). Three types are recognised: lumbar scoliosis, thoraco-lumbar scoliosis and thoracic scoliosis. In juvenile idiopathic scoliosis the curve is most often right thoracic and has a high rate of progression (Campbell et al, 2006).

SECONDARY STRUCTURAL SCOLIOSIS
This type of scoliosis is formed by underlying causes. The three most common causes are congenital abnormalities, presenting with a sharp angulation at the site of the anomaly and compensatory curvatures above and below (Adam & Hemblen, 2001). Anomalous vertebral development leads to congenital scoliotic curvatures.
The malformation can lead to failure of vertebral segmentation or failure of vertebral formation. Both processes are often seen in the same spine either at the same or different levels. Adjacent vertebrae which do not completely separate, and thereby produce a unsegmented bar without a disc in between them, present a segmentation defect, which leads to severe lateral progressive scoliosis. Circumferential segmental failure results in the loss of segmental motion and loss of longitudinal vertebral growth, without rotational or angular deformities (Campbell et al, 2006).

Poliomyelitis or other neuromuscular disorders with residual weakness of the spinal muscles cause unequal pulling on the sides of the spine. Neuromuscular curvatures typically develop at a young age and often present as a “S”-shape curve. 90-100% of spinal deformities occur with dystrophy and in spinal cord injuries leading to quadriplegia in infants and young children (Campbell et al, 2006). Neurofibromatosis leading to spinal curvatures have an unknown mechanism (Adams & Hemblen, 2001).

**COMPENSATORY SCOLIOSIS**
Compensatory scoliosis presents with leg length discrepancy or abduction or adduction deformity at either hip. Normally it will resolve when the underlying cause is corrected, in case the condition did not last so long that the scoliosis became fixed.

**SCIATIC SCOLIOSIS**
This curve is normally located in the lumbar spine and caused by a prolapsed disc. It is a protective action of muscles to reduce the pain as far as possible caused by the affected nerve or joint. The position is assumed involuntarily (Adams & Hemblen 2001).

### 6.2 Etiology
The underlying conditions are unknown, but a familiar tendency is expected. Environmental factors are implicated to be a factor contributing to infantile idiopathic scoliosis for the reason that it is common in England and northern Europe but rare in the United States (Campbell et al, 2006).

Adolescent idiopathic scoliosis accounts for 80% of all cases, the female to male ratio for curvatures of 10° is nearly equal (1.4:1), while the ratio for curvature of 20° is 5:1 female to male, and females having a higher percentage with curvature progression (Campbell et al, 2006).

The etiology of congenital scoliosis involves foetal environmental factors, especially for the development from day 45-60 after fertilization.

In neuromuscular scoliosis the cause is unclear. Hypothesis are loss of strength in the muscles and lost of proprioception as well as loss of spinal stability which is directly related to the condition of the end support of the spine and inversely proportional to the flexibility in the spine and to the approximate square column length of the spine (Campbell et al, 2006).

### 6.3 Assessment
The goals for the assessment of a child with scoliosis are to monitor curve progression and to detect individual problems, like respiratory difficulties or pain. Assessment is important in order to develop a treatment plan. Scoliosis is often detected through the Adam’s forward-bend test, the use of a scoliometer or clinical examination of asymmetry. According to Cote et al (1998), the Adam’s forward-bend
test is more sensitive than the scoliometer and is therefore considered the best non-invasive clinical test to evaluate scoliosis. A typical sign seen in scoliosis patients when performing the test is a rib hump. During the assessment, the physiotherapist should also do a leg length examination, because ‘a leg length asymmetry can cause a non-structural curve’ (Effgen, 2005). Furthermore, the therapist should look for uneven hip or shoulder heights during standing or an increased space between the elbow and the trunk on the concave side of the curve. Those are signs, typically seen in scoliosis patients. If an asymmetry is found, the scoliometer can be used to determine the angle of trunk rotation. ‘The scoliometer measures the angle of trunk inclination and has a high interobserver reproducibility, which permits the determination of cut-off points above which radiographic study is indicated’ (Negrini et al, 2005). An angle of 7° or more is considered significant. According to Farady (1983), ‘the presence of structural change is evidenced by the patient’s inability to correct the curve when flexing laterally toward the convex side’.

6.4 Treatment

The management of scoliosis can be either nonoperative or operative. Nonoperative intervention is also considered as conservative management. Its main goal is to stop curvature progression. Sub goals are the prevention and treatment of secondary functional impairments (restrictive ventilatory disorder and reduced cardiopulmonary performance due to reduced mobility of the ribs), reducing pain and cosmetic improvement.

Next to scoliosis intensive rehabilitation and brace treatment, physiotherapy is one of the three conservative treatments of scoliosis. ‘It has to be emphasized that (1) physical therapy for scoliosis is not just general exercises but rather one of the cited methods designed to address the particular nuances of spinal deformity, and (2) application of such methods requires therapists and clinicians specifically trained and certified in those scoliosis specific conservative intervention methods’ (Weiss et al, 2006). Physiotherapy modalities used in the treatment of scoliosis are exercise, electrical stimulation and respiratory exercises. Exercises are aimed at maintaining and improving spine flexibility and trunk strength. ‘Exercise therapy is generally the sole treatment of patients with a curvature (Cobb) angle up to 20°’ (Weiss, 1992). Respiratory exercises are important in patients with large thoracic level curves, ‘because the curve may impede thoracic expansion and limit breathing’ (Effgen, 2005). According to Weiss et al (2006), a Cobb angle of more than 35° is an indication for orthotic intervention. In that case, the physiotherapist is responsible for the teaching of breathing exercises during orthotic wear as well as trunk strengthening and trunk flexibility exercises in and out of the orthosis. ‘Exercises in the brace are intended to help the wearer adapt to the orthosis, aid in the development of postural awareness, and contribute to the overall effectiveness of the brace by encouraging the active correction of lateral and rotatory deformities and the reduction of lumbar lordoses’ (Farady, 1983). ‘Exercises performed out of the brace are intended to correct preexisting joint range of motion and muscle strength limitations, to maintain or increase the range of motion and muscle strength of those joints and muscles that are immobilized by the brace, to ensure adequate chest mobility, and to develop the corrected posture’ (Farady, 1983). Furthermore the child should be encouraged to stay physically active during brace intervention. Negrini et al (2005) state that ‘general sports activities are recommended which offer patients aspecific benefits in terms of psychological, neuromotor and general organic well-
being’. They furthermore recommend that ‘sports activities should be continued also during brace treatment because of the physical and psychological benefits these activities provide’ (Negrini et al., 2005). Cardiopulmonary fitness can be achieved through specific exercises as well as group activities like swimming, cycling or dancing. ‘Exercises in groups is performed during Scoliosis Inpatient Rehabilitation (SIR) in Germany and Barcelone, but also in Switzerland and Israel. The positive psychological impact on scoliosis patients who are rather alone with their deformity helps to cope with the disorder.’ (Weiss et al., 2006).

According to Effgen (2005) children who belong to one of the following groups, are treated operatively. ‘These are children who are still growing and have a curve of more than 40° to 45°, children with curve progression of 40° to 45° despite nonoperative intervention, and mature adolescents with a curve of more than 50° to 60°. In the inpatient setting, after the surgery, the physiotherapist’s role is to teach bed mobility, transfers and ADL skills like dressing. Directly after the surgery, spinal rotation is contraindicated. Therefore it is important that the patient is taught log rolling. Further exercises that have to be taught are general ROM and strengthening exercises. ‘Postoperative bed rest after major surgery puts the child at risk for orthostatic hypotension’ (Effgen, 2005). Therefore care has to be taken when starting to exercise with the child and the physiotherapist should instruct the child in performing circulation promoting exercises, like ankle pumps. Especially after surgery, body awareness training is an important part of the rehabilitation because the child has to get used to his new body image. ‘The use of proprioception, tactile stimulation to improve neurodynamics and self perception is important and is an integral part of many treatment programs to facilitate 3D correction’ (Weiss, 2006).

### 6.5 Evidence Based Discussion

In the literature dealing with scoliosis treatment modalities, there is a lot of discussion concerning the effectiveness of physiotherapy in the rehabilitation process of scoliosis. In older papers, physiotherapy is usually considered ineffective. ‘Physiotherapy by itself has no part to play in the conservative management of scoliosis. Its use is based on the misguided belief that it is possible to strengthen unilaterally the muscles on the convexity of the curve, so creating a muscle imbalance which would correct the deformity. This, however, is impossible because exercises strengthen all of the spinal muscles equally’ (McMaster, 1982). Those papers regard the treatment with braces as much more important than physiotherapy. In a recent paper, Romano and Negrini (2008) state: ‘The most recent review published in 2005, asserts that at this state of the art, physical therapy and the application of a brace have the same reliability value’. This shows that the opinion regarding physiotherapy has changed dramatically over time. Weiss et al (2006) even declare that ‘although discussed from contrasting viewpoints in the international literature, there is some evidence for the effectiveness of scoliosis treatment by physical therapy alone’.

### 6.6 Conclusion

In the treatment of scoliosis, the physiotherapist plays a major role. Especially in the postoperative rehabilitation process he fulfils an important part. The children lived their whole life with a curved spine and suddenly have a more or less straight back. Therefore they have to get used to a totally new body image. Through proprioception and body awareness training, the physiotherapist can help the child to feel
comfortable in his own body. The brochure gives the children an idea about what will happen after an operation. Most scoliosis patients in the Asklepios clinic are admitted for an operation. In the brochure we therefore focused mainly on the postoperative treatment. Furthermore the patients already stay at the hospital several days before the actual surgery. That means that the brochure could be given to them upfront, taking their fear and preparing them mentally on the time after the operation. The brochure also explains what the children have to take care of when leaving the hospital, for example that they should not do sports in the first time after the surgery. The brochure informs the child about the entire time spend in the hospital, including the preparatory phase and the postoperative rehabilitation.
7. General Conclusion

Our goal was to provide children with appropriate information material about physiotherapeutic treatment. We wanted to decrease their fear, make them understand what they have to expect and why physiotherapy can be very important. After finishing the content of the brochure one of our external advisors read the text to a group of children, aged four to 12, and their parents. We received very positive feedback from both the children and parents. Because of that, we believe that we reached our goal. Another goal was to learn how to produce a client-focused information brochure. Throughout the development of the brochure we gained a lot of experience in writing patient information leaflets, how to approach a specific patient group and the development of a layout and design.

For this project it was essential to meet the client’s expectations. Sabine Steinstraß was satisfied with the process and the result. Regarding the language of the brochure, we tried to write it according to the before mentioned guidelines (see chapter 2). We tried not to use any medical terms and keep the sentences short. If medical terms had to be used, such as goniometer, we explained them in an illustrative way. In order to give the children the opportunity of identification, we chose for three fictional characters, namely Ben, Lisa and Tina who experience physiotherapeutic treatment for their disease. In the introductive parts of the brochure, the reader is directly addressed by the use of “I”, “You” and “We”, creating the effect of direct communication.

Nowadays a brochure might seem old fashioned and other media might be more effective times of playstation, internet and nintendo wii. Therefore it might be an idea for the future to use such media as information material. For example it could be an option to develop a computer game or an interesting, animated website. For the before mentioned reasons, we also recommend to digitalize the brochure.

For our brochure we decided on three specific diseases. Those diseases were chosen in agreement with our client. We decided for JRA, Cystic Fibrosis and Scoliosis as those are diseases very frequently treated in the clinic. An idea for future projects would be to write patient leaflets for more specific diseases such as heart conditions. We belief that a brochure like the one we wrote would definitely be helpful in the German Childrens Heart Centre. On one hand there are more babies treated, for who a brochure wouldn’t make sense, but the few older children are often very afraid of physiotherapy. Especially after a surgery they do not like to be touched.

To sum up we can state that we are very satisfied with our end product. Both of us could imagine to work in a similar project again.
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9. References


