

OI in Motion

**Rehabilitation and Physiotherapy in
Osteogenesis Imperfecta**

**Rheinsberg, Germany
November 20-22, 2009**

ABSTRACTS



Foreword

It is with tremendous pleasure and pride that we welcome you to OI in Motion. After our previous meeting in Rome about orthopaedics this second topical meeting takes physiotherapy and rehabilitation as its theme.

Initiated by OIFE and this time also organized by OIFE, it is the first international meeting of its kind on the topic of physiotherapy and OI. Within the "OI-sciences" physiotherapy deals directly with the person with OI - and his or her family - often so for many years. In this respect physiotherapy is special and different to some other disciplines. On the other hand rehabilitation does not stand alone; it needs to be seen in close connection to other forms of treatment such as rodding and scoliosis surgery, occupational therapy, psychological assistance, bisphosphonate treatment and nutritional advice. In an ideal scenario, different fields work together to determine the optimal treatment for each individual with OI. Children with OI experience most fractures and may therefore receive most attention from physiotherapists. However OI does not stop at 18 and adults still benefit from physiotherapy, especially as the ageing process sets in and accumulated physical problems start to take their toll on fitness.

A specialized field like physiotherapy and rehabilitation in OI is entitled to its own place. A place we hope to have created with OI in Motion. A title that symbolizes that none of us should sit still, neither people with OI nor those treating people with OI. We express the hope that you will have ample opportunity to exchange experiences and ideas, connect with each other and return home truly inspired.

At this point I wish to thank my partners from the organization team; Carina Svenheden and Simona Paveri, as well as our secretary Steffi Wagner and OIFE president Ute Wallentin for their tremendous work in the past year! Also a big thank you goes to Oliver Semler our very knowledgeable friend from the Children's Hospital in Cologne who happily accepted to be the chair of our meeting. Finally we thank all speakers, volunteers and sponsors who made this happen.

Thank you all and welcome to Rheinsberg ! Enjoy your stay.

Taco van Welzenis, chair of the organizing committee

Contents

Foreword - T. van Welzenis

Osteogenesis Imperfecta Federation Europe (OIFE) - U. Wallentin

Abstracts of presentations

Rehabilitation following Fassier-Duval intramedullary rodding of the femur/tibia - J. Ruck

The spine and spinal fusion in Osteogenesis Imperfecta – S. Pochintesta

Different physiotherapeutic strategies in OI - The Cologne Concept – O. Semler & T. Petersen.

Rehabilitation throughout the years of the child and adolescent with OI – J.Ruck & K. Montpetit

Measurements of muscles and bones and their use to evaluate therapeutic concepts - O. Semler & T. Petersen

Equipment and devices for children with OI – K. Montpetit, and J. Ruck.

Special aspects in follow up and (re)habilitation of adults with osteogenesis imperfecta - L. Lande Wekre

Psychological aspect of OI – I-L Andresen

Experience with OI at "La Nostra Famiglia" in Bosisio Parini and "Centro Vojta" in Rome - P. Fraschini

Rehabilitation of children with OI in Italy - T. Baratella

The Vojta Center experience in Rome: A complete approach in the Rehabilitation of O.I. outcomes - M. de Bari

Physiotherapeutic approach with O.I. child and education to autonomy in O.I. adolescent and adults - O. Vitale

Workshop abstracts

"Glassfit" - A physiotherapy program for people with OI – W. Hagelstein

Galileo workshop – O. Semler, A. Meiswinkel & T. Petersen

Poster abstracts

Assessed and perceived physical function in adults with Osteogenesis Imperfecta (OI)
V. Balkefors, E. Mattson, Y. Pernow & M. Sääf

Time management as a tool to fight exhaustion and to keep fit in OI adults
Experiences from practice and role of a patient organization – T. van Welzenis

TRS – National Resource Center for Rare Disorders – L. Lande Wekre

Osteogenesis Imperfecta Federation Europe (OIFE)

OI is an inherited disease of connective tissue affecting around 1:15.000 people. Its most striking features are brittle bones, blue sclera, short stature, deformity, deafness and dental abnormalities. OI results from genetic defects that cause the body to make insufficient amounts and / or imperfectly formed collagen, a major building block of connective tissue. As OI is a relatively rare condition, the number of people it affects, let alone specialists treating it, is not very large in each country. The need for specialized help however is considerable, especially given the huge variation in symptoms and severity that make each case unique.

In 1993 several of the often small national European OI organizations decided to join forces and work together in a European Federation. Thus OIFE was born. At this moment OIFE has 18 European member societies and 5 associated non-European members from Australia, Ecuador, Mexico, Peru and the USA. The godparents-program PadrinoS-OI, focusing on Latin-America and the HOI foundation (Help OI) are supporting members of OIFE with whom we work closely together. In total OIFE represents more than 7500 people with OI.

Each OIFE member is represented by a national delegate. The executive committee (EC) consists of the president, vice-presidents and treasurer. So far this work is all done by volunteers! The EC is further assisted and advised by a secretary, webmaster and youth delegate and several medical experts.

Objectives of the OIFE:

- Networking between professional OI-specialists, treatment centres, national OI associations and OI patients worldwide
- Support of member societies by the exchange of information and experiences
- To support people with OI in countries without existing OI societies and to help establish such organizations there
- To support international exchange between people with OI
- Representation of its members on a European level – as a member of the European umbrella organisation for rare disorders „EURORDIS“
- Presentation of problems and needs of people with OI to national and international organisations
- Promotion of research on all aspects of OI – in cooperation with the international OI-registry at the Kennedy Krieger Institute in the US
- Collection and publication of information about OI
- Promotion of public awareness of OI

Recent activities include: European youth meetings, an OI-passport with information in 16 languages, international training for doctors and other OI specialists, a multilingual website and digital newsletters. Immediate future plans include the subtitling of an educational DVD and a student exchange project.

If you would like to know more or see a way to support our work, please contact us:

OIFE
Ute Wallentin
Sturzstr. 17
96049 Bamberg
Germany
+49 – 951 - 60 33 16
office@oife.org www.oife.org



Abstracts of presentations

Rehabilitation following Fassier-Duval intramedullary rodding of the femur/tibia

Joanne Ruck, MSc PT

Shriners Hospitals for Children – Montreal, Quebec, Canada

The purpose of this presentation is to describe the rehabilitation and outcomes following Fassier-Duval (FD) rodding of the lower extremities for children with Osteogenesis Imperfecta who have never walked.

The criteria for surgery at the Shriners Hospitals for Children – Canada (SHC) include: significant femoral and tibial bowing, repeated fractures of the lower extremities, an adequate level of bone mineral density and a child who demonstrates the ability of pull to stand.

The surgery is carried out in two stages: one leg followed by the other one week apart and moulding of long leg braces in the operating room after the second surgery. Physiotherapy rehabilitation occurs four weeks after the first surgery. During that week the child receives the braces, is verticalized receives active range of motion exercises of the legs, gentle hip flexor and heel cord stretches and takes steps with a walker.

Functional outcomes of this surgery for 60 children will be presented:

Active ROM 5 weeks post rodding hip: 100° knee: 100°

FAQ ambulation score: pre-op: 2 (some stepping with human assistance) to 5.9 (at least a household ambulatory at the one year level).

The GMFM results show a significant improvement in standing and walking domains from 6 months to 4 years post op.

The spine and spinal fusion in Osteogenesis Imperfecta

Simona Pochintesta

Associazione “la Nostra Famiglia” - IRCCS “Eugenio Medea”, Bosisio Parini, Italy

Incidence of spinal deformity is very high in O.I.: from 26% to 80%. Important spinal deformity concerns mainly the severe types of O.I. The onset of scoliosis and kyphosis comes very early in the life of patients. Untreated spinal deformity continues to worsen in adulthood with severe complications and disabilities. The most important complications are respiratory restriction and ventilatory failure. The treatment with braces is generally ineffective and causes some complications. Surgical treatment with spinal fusion can preserve respiratory function and improve sitting balance or walking abilities. In the preoperative and postoperative phases patients need a specific physiotherapy in a rehabilitation centre.

Different physiotherapeutic strategies in OI - The Cologne Concept

O. Semler¹, T. Petersen²,

1 Children's hospital, university Cologne, Germany

2 medifitreha, university Cologne, Germany

The Cologne Rehabilitation Concept for children and adolescent with OI is a new approach to combine different therapeutic strategies to strengthen muscles and bones and to improve mobility. All therapeutic strategies should focus on improvements of independency and on an increase of life quality in these patients.

Intervention: Patients participate in an intensive multimodular rehabilitation program. The program includes six months of whole body vibration training at home and two stays as inpatients at the rehabilitation centre (13 days at start and 6 days after three months). During the time at the centre physiotherapeutic treatment include: NDT, gait training, treadmill training, pool therapy and training in a specially equipped fitness gym. One important part at the centre is a whole body vibration training with the side alternating platform Galileo®. On this platform special exercises have to be conducted by the patients to train their muscles. During the first stay at the centre patients and parents were taught to train in the appropriate positions. During the second stay correct positioning during the vibration training is checked and exercises are intensified where possible. After the end of training and after further six months outpatient visits are performed to assess the long-term improvements.

Conclusion: In the first analysis the Cologne Rehabilitation Concept for children, adolescents and adults with OI including a whole body vibration training was beneficial for the participants and led to an increase in muscle- and bone mass. Additionally the patients showed improvements of mobility and became more independent in activities of daily living.

Rehabilitation throughout the years of the child and adolescent with OI

Joanne Ruck, MScPT, Kathleen Montpetit, MScOT

Shriners Hospital for Children, Montreal, Quebec, Canada

With the introduction of bisphosphonates and the use of more effective surgical techniques with shorter periods of immobilization the role of rehab has greatly increased for children with all types of OI. The new treatments reduced the fragility and consequently the anxiety promoting the development of interdisciplinary teams. This presentation will demonstrate the rehabilitation philosophy developed at our center. Our approach consists of

- setting attainable short term goals which is optimal for a fragile child with overprotective parents.
- Individualized goals according to child's clinical presentation
- Respect for milestones of normal growth while considering the musculoskeletal characteristics of OI.
- Gradual progression of goals

Various strategies for exercises, positioning and adaptive equipment will be described for the following developmental stages and transition points

- Infancy
- Learning to sit
- First mobility
- Standing
- Ambulation
- Adapted mobility
- Upper extremity

- Self care
- Strengthening and sports
- School

Measurements of muscles and bones and their use to evaluate therapeutic concepts

Oliver Semler¹, Tanja Petersen²

1 Children's hospital, university Cologne, Germany

2 medifitrea, university Cologne, Germany

When children are treated with drugs there are strict regulations regarding safety and effectiveness. Primary and secondary endpoints have to be defined and it is specified, which improvements should be achieved in a given time period. In physiotherapy therapeutic aims, time periods and endpoints are rarely defined. Sometimes physiotherapists and patients do not realize the importance of defined endpoints and sometimes they have problems to find appropriate measurements. Assessment of motor functions in children with OI with validated scales is sometimes difficult due to fractures and deformities. Nonetheless there are possibilities to measure muscle force, muscle function and bone stability even in children with OI. Dual energy X-ray absorption is a method primarily to assess bone density but it can also be used to measure other skeletal parameters. The method can be used to quantify bone mass and bone mineral density but also muscle mass. This method allows to determine muscle mass of the whole body and of specific regions of interest like legs or trunk. By using DXA it is possible to quantify changes in muscle mass in the course of therapeutic interventions.

In growing children with OI the influence of physiological growth and changes of intramedullary rods have to be considered as influencing factors. It has to be kept in mind that muscle mass is not identical with muscle function. Muscle function and mobility and not an increase of muscle mass are the common therapeutic aims in rehabilitation and the parameters physiotherapists focus on. To assess improvements of muscle function a "ground reaction force plate" can be used. This platform can measure the force a patient can apply to the ground while standing up from a chair, shifting weight or while jumping. For patients not able to bare their whole weight the platform can be combined with a tilting table.

For patients who can walk a gait analysis can be used to record and quantify changes of mobility during training. Assessing, recording and analyzing changes in motor function should become an indispensable part of physiotherapy like it is in other medical fields already. This seems to be the only possibility to improve physiotherapeutic strategies in the future based on a scientific background and to ensure adequate funding for rehabilitation in the future.

Equipment and devices for children with OI

Kathleen Montpetit, MScOT and Joanne Ruck.MscPT

Shriners Hospital for Children, Montreal, Quebec, Canada

The use of appropriate adaptive equipment and devices can improve the performance and ultimately the independence of the child with Osteogenesis Imperfecta. Exploration and experimentation at all stages of development are essential ingredients for independence.

The challenge is to encourage meaningful activity while minimizing the risk of fractures and increasing the child's sense of accomplishment. These are key elements if the child is to achieve independence in gross motor skills, self care, mobility and domestic life. This presentation will outline the general categories of equipment that can be useful at various stages of development and describe equipment and devices that promote safety, early mobility, independence, and optimal body alignment. Furthermore this presentation will explain how adaptive equipment can reduce the care giving burden for parents, meet the aesthetic and social needs of families ultimately improving their quality of life.

Psychological aspect of OI

Inger-Lise Andresen

TRS National Resource Center for Rare Disorders, Norway

Psychological aspects of a disability are often described in general terms, regardless of diagnosis: Crisis, grief, depression, anxiety. This can be relevant and important and health care professionals can be helpful if the problems are extensive. But different diagnoses have different psychological implications and it is important that health professionals know which specific challenges a diagnosis implies, also on the psychological level.

Osteogenesis Imperfecta is: Lifelong; Hereditary; Rare; Complex; Visible for some – invisible for others. OI implies: Many contacts with the health care system; Restrictions and demands; FRACTURES and Pain. These aspects may cause challenges that will be discussed in the presentation.

The challenges of OI will be experienced in very different ways for different persons; and what at one time is quite easy to cope with, may at other times be very difficult. It is important that health professionals are aware of the psychological challenges as well as the medical, and take consideration of them in our treatment and advice. But it is also important that we acknowledge that we meet persons with OI in very specific situations, their lives may be quite different and have more manageable challenges when they do not seek our help.

Special aspects in follow up and (re)habilitation of adults with osteogenesis imperfecta

Lena Lande Wekre, MD

TRS, National Resource Center for Rare Disorders, Sunnaas Rehabilitation Hospital, Norway

OI is a genetic disorder of increased bone fragility and other connective-tissue manifestations with a wide spectrum of clinical expression. Ageing is said to be the accumulation of changes in an organism or object over time. It is a multidimensional process of physical, psychological and social change. Some dimensions of ageing grow and expand over time, while others decline. Distinctions may be made between the biologic ageing process (age changes that all people share), and "probabilistic ageing" (age changes that may happen to some, but not all people as they grow older). The biologic ageing process begins at the age of 20, and leads to a 50 % reduction of the organ-reservoirs by the age of 70. However, OI may have influence on the "probabilistic ageing" process, but we know little about how, and in what degree. These perspectives have to be taken into consideration when making follow up and (re)habilitation programs for adults with OI. As a connective tissue disorder, OI affects different parts of the body. Even though the main problems come from the skeleton and muscles, we have to make an overall examination including hearing, sight, teeth, lungs, heart and gastrointestinal organs to establish optimal follow up and (re)habilitation programs.

Experience with OI at "La Nostra Famiglia" in Bosisio Parini and "Centro Vojta" in Rome

Paolo Fraschini

Associazione "la Nostra Famiglia" - IRCCS "Eugenio Medea",
Bosisio Parini, Italy

Pediatric rehabilitation as a discipline is rapidly changing, especially during the last decades. This is reflected among other things in the increasing number of pediatric measures and instruments specifically geared to the paediatric rehabilitation profession, GMFM, WeeFIM, PEDI. We tested our group both with GMFM and WeeFIM: both have good levels of validity reliability and responsiveness.

Osteogenesis Imperfecta is a genetically heterogeneous disorder with similar skeletal manifestations that are the result of different genetic defects. Clinical features vary widely not only between types, but within types, and even within the same family.

Physical therapy should begin as soon as it is evident that an infant has muscle weakness or motor skill delay when compared with same-age peers, and continue until a child reaches appropriate physical therapy goals. The long-term goal for children with OI is independence in all life functions with adaptive devices as needed. Occupational therapy can help with fine motor skills and adaptive equipment for daily living. As a child with OI grows older and gains more independence, he or she will benefit from continued physical activity, such as adapted physical education. Adults with OI also benefit from safe exercise to maintain bone and muscle mass. Swimming and water therapy are particularly well-suited for people with OI of all ages, as they allow independent movement with little fracture risk.

Many children with OI undergo a surgical procedure known as rodding, in which metal rods are inserted into the long bones to control fractures and improve deformities that interfere with function. Progressive scoliosis may cause respiratory problems. Bracing is generally not recommended. Spinal rodding may be appropriate in severe cases.

The most significant medical advance in the treatment of children with OI has been the introduction of bisphosphonates therapy. Worldwide experiences show a reduction in fracture and an increase in BMD and vertebral shape mainly in young patients. Further studies from other groups including our Italian group confirmed those data improving functional values in WeeFIM evaluation, however we had delayed healing or pseudarthrosis on bone x-rays in 8 patients despite an increase in their BMD of the same magnitude as in the others.

Rehabilitation of children with OI in Italy

Tamara Baratella

Associazione "la Nostra Famiglia" - IRCCS "Eugenio Medea",
Bosisio Parini, Italy

The scientific research institute "E.Medea, La Nostra Famiglia" is a rehabilitation centre that takes care of disabled people and their families. Its main goal is the rehabilitation during developmental age and not just to treat the growth and improve the functional skills. One of the diseases that is given particular importance is Osteogenesis Imperfecta, a genetic disease that causes brittle bones.

Rehabilitative treatment depends on the severity of disease and the age of patient. Severe Osteogenesis Imperfecta is a hard challenge to face for doctors, therapists and all the personnel involved. The cooperation between personnel and the patient's family is very important during childhood.

The therapist has got to help the child and his family to develop an optimistic view of life and to teach it how to become as independent as possible.

This task can be achieved using a step by step approach towards the treatment, treating the child in a confident and relaxed manner in order to put the child at ease. The children can enjoy and move without being afraid when playing on a carpet on the floor.

The following part of the treatment aims at preventing fractures, developing motor skills, increasing the degree of joint mobility and muscle strength and gradually increasing weight bearing. The therapist as well plays a fundamental role after surgery which may lead to a variety of problems such as pain, fear, articular blocks, immobility, loss of weight bearing and walking and reduction of autonomy in daily life. In these specific cases the treatment has the following targets: to regain joint function, to regain strength and muscle control, to improve postural alignment, to rehabilitate or acquire independence during postural steps and transfers, to [recover the load] become weight bearing again, to gain/acquire a functional walking for short trips and independence both in indoor and outdoor.

All this can be achieved through: active and passive mobilization, active muscle strengthening exercises, electro stimulation with Kotz currents, gradual increase of weight bearing using a static table, execution of postural control of upright posture and exercises to assisted recovery or acquisition of walking ability. In addition to traditional therapy, the patient with Osteogenesis Imperfecta has a great benefit and advantage from rehabilitation in water which uses: the absence or reduction of weight bearing, the hydrostatic pressure and temperature (water at 34 °). Through specific therapeutic exercises and gradual use of floats, weights and other hydrotherapy can reach the goals with less effort and quicker.

The Vojta Center experience in Rome: A complete approach in the Rehabilitation of O.I. outcomes

Mariapia de Bari

Centro di Riabilitazione Neuromotoria Vaclav Vojta, Roma, Italy

Rehabilitation and physiotherapy of O.I. patients are very difficult, since this disease presents not only orthopaedic, traumatologic, respiratory problems, but also impairment of the psychomotor development.

The first year of life has a huge importance in babies growth: new born babies discover their own limbs and body through the eye-hand co-ordination.

They take contact with the surrounding ambient through their motion, starting to control their head (2 months), trunk (6 months), using two first forms of global moving: the rolling and the crawling.

Afterwards, about 7 months and a half of life, babies acquire a well-developed form of global moving: the on all fours deambulation which will turn in the bipedal walking (about 12 months).

In babies with O.I., deformities and recurrent fractures, even if occurred after the twelfth month of life, retard and/or impair the acquisition of one or more motor stages.

The growth through those motor stages (rolling, crawling, on all four deambulation, etc.) is essential for the baby, not only for a physiological psychomotor development, but also to build a muscular balance and a correct posture.

Further more the motor performance of O.I. babies is extremely influenced by all psychological aspects related to this disease: fear of pain, sense of inadequacy, family anxiety, etc., all elements that compromise and reduce babies motion's potentiality.

For all these reasons, rehabilitation of O.I. outcomes, at any level of seriousness and in any age is never simple and recovering strength, endurance and the degree of a stiff joint after a fracture is not the only purpose: O.I. patients deserve a global and specific approach in which the physiotherapist plays a very important role.

The principal ways the physiotherapist has to face O.I. outcomes and to get the patient confidence are the technical strategies.

By technical strategies we mean high professional knowledge of physiotherapeutic instruments. The more instruments the physiotherapist has, the more possibilities O.I. patients have to achieve results up to the highest possible level of autonomy.

The Vojta Center experience about physiotherapy in O.I., is actually based on a complete use of exercises extrapolated from various methods.

Vojta and Kabath method, hydrokinesitherapy, technics of manual therapy and global exercises of postural re-education are all based on principles of muscular physiology and neurophysiology and are opportunely chosen according to the needs of the patient and after a functional evaluation.

In order to apply powerfully and successfully the above mentioned complete approach, all methods have to be deeply studied and must be well known by the physiotherapist.

The physiotherapist will choose the best method to reach the patient's target.

An open minded use of all methods mixed with high sensitiveness is the main peculiarity of the physiotherapist job.

Physiotherapeutic approach with O.I. child and education to autonomy in O.I. adolescent and adults

Orazio Vitale

Associazione "la Nostra Famiglia" - IRCCS "Eugenio Medea",
Bosisio Parini, Italy

Osteogenesis Imperfecta can't be compared to other diseases, although similar, because its clinical features needs a special attention especially considering the psychological dynamics. one's weakness and deformities are present in almost all cases from birth, producing situation of tension and anxiety both in patients and parents. This particular state generates a "fear of fracture" that will last for all their lives. The first difficult task for a physiotherapist will be to establish a relationship based on trust with the child, or the young patient and his or her family. From this relationship will depend the outcome of all the future treatments. In O.I. people in addition to bone fragility, determined by the structural genetic defect, it is necessary to consider also the so-called "secondary osteoporosis"

due to the immobility of skeletal segments, caused by the analgesic attitude and from supine prolonged immobilization of fractures in chalk.

Stillness and restriction of movement should be avoided, therefore it is essential that the therapist with the parents help the OI child to find the motivation to movement. Stillness would certainly damage him or her both psychologically and physically increasing bone fragility. Before deciding a treatment plan and setting goals for the patient it is very important to know his or her history from birth up to present day, to know the number and the kind of fractures incurred, and which body districts have been affected. Then the therapist should ask about the number of hospitalizations and about the way the whole family experienced pain and life with O.I.

The aim will be, for the children and the young O.I., to make them as independent as possible, living a balanced confident life. Autonomy will be the goal we should try to achieve most of all during adolescence and adult age. Adolescence is, for certain aspects, the more difficult and critic period for a boy or a girl, it will be even more difficult if he or she is affected by O.I. An O.I. adolescent faces society carrying his past made of pain, hospitalizations, tension and his own and his family's fears; his future is unsure and a bit frightening. Physiotherapists must take into account all this.

The two fundamental moments on the way toward autonomy for an adolescent and for an adult can be summarized in two words: rehabilitation and prevention. Achieving goals will give, or give back, autonomy (especially in late forms) Physiotherapy, surgery and orthesical treatments, if necessary, should be used to limit, as much as possible, the immobility in order to avoid any kind of important muscular problem. In adolescence goals will be: deformity prevention and trophism and muscular power maintained.

For the adult, goals will be linked to the improvement of physical ability, muscular balance and autonomy.

In any case, there will be a special attention for movement (for example: constant activity in water) and for load (important element of contrast against osteoporosis).

Workshop abstracts

“Glassfit” - A physiotherapy program for people with OI

Willy Hagelstein,

Developer of Glassfit; Lauf Germany

Based on my experience I developed a physiotherapy program for people with OI, called Glassfit. Together with two physiotherapists and a doctor, all experienced in OI, we published Glassfit in 1997. In the meantime Glassfit has already been translated into English, Georgian, and recently translated into Russian. The Glassfit 2, published in 2001, has also been translated into Spanish.

The Idea of Glassfit is to give people affected by Osteogenesis Imperfecta a support for their daily life training and to give the therapists assistance in the treatment.

Galileo workshop

Oliver Semler^{1,2}, Annika Meiswinkel², Tanja Petersen²

1 Children's hospital, university Cologne, Germany

2 medifitrea, university Cologne, Germany

Whole body vibration therapy with the Galileo system is an important part in the rehabilitation concept for children with OI in Cologne. The Galileo system is a side alternating vibration system to strengthen muscles. It can be used as a standing device or in a special adaptation in combination with a tilt table. This adaptation allows children who are not able to stand to use the system to train their muscles. The Galileo-System has shown to improve musculomass and muscleforce in adults and children. Following the increased muscleforce the bone mass is increasing by training with the Galileo system.

During this workshop we will present the theoretical background of the side alternating vibration platform and we will demonstrate our experiences with this training method in children with OI. Most importantly we will have some Galileo standing devices and a Galileo

tilt table available. All participants will have the opportunity to test the Galileo System on their own and can try different exercise. We will discuss different therapeutic exercises which can be performed on the platform to train specific muscles which are needed for new patterns of movement.

Poster abstracts

Assessed and perceived physical function in adults with Osteogenesis Imperfecta (OI)

Veronica Balkefors,⁽¹⁾ PhD-student, PT, Eva Mattsson,⁽¹⁾ PhD, PT, Ylva Pernow,⁽²⁾ PhD, MD, Maria Sääf,⁽²⁾ PhD, MD
Department for Neurobiology, Care sciences and Society ⁽¹⁾ and Department of Molecular Medicin and Surgery,⁽²⁾ Karolinska Institutet, Sweden.

The aim was to describe joint mobility, muscle function, pain, physical activity, quality and satisfaction of life and how adults experience living with mild/moderate form of Osteogenesis Imperfecta, OI.

Participants were invited through Clinic's patient register or via the Swedish OI Association. The participants included had to be able to walk and diagnosed with mild/moderate OI. Forty, men and women between 21-71 years, were identified and twenty-nine agreed to participate. Evaluation of joint-, muscle function, life satisfaction, pain, daily physical activity and self-perceived disabilities was performed partly by self-administrated questionnaires along with taped interview and partly by clinical examination.

The result shows that 25/29 of the participants had pain. Eight persons self-estimated low level of physical activity assessed by International Physical Activity Questionnaire (IPAQ) meaning they did not reach recommended physical activity level associated with great health benefits. Several of the persons perceived not be able to run or walk fast as a disability, and estimate it high on the Disability Rating Index (DRI). Health related quality of life, assessed with Short Form 36 (SF-36), was lower in the areas physical functioning, median 75(0-100), role physical, median 88(0-100), bodily pain, median 62(0-100), general health, median 72(35-100) and vitality, median 60(25-90) than the Swedish norm (95, 100, 84, 82 respective 75). The interviews of the person's experiences resulted in categories showing the experience of living with OI and the solutions they adopted in daily life.

We conclude that OI affects the person on all levels according to ICF (impairment, disability, activity limitation and participation restriction) WHO. Reported physical limitations involve pain, trouble running/walking fast, heavy lifting and the feeling of fear of falling. The functional ability differs between, as well as within individuals and over time.

Time management as tool to fight exhaustion and to keep fit in OI adults - experiences from practice and role of a patient organization

Taco van Welzenis
VOI – Dutch OI association, Utrecht, The Netherlands
Netherlands@oife.org

In recent years there has been increased attention to the needs of adults in the Dutch OI association. Several meetings were organized for this target group. Frequent fractures are often no longer the main problem; instead many OI adults indicate (sometimes severe) problems due to declining energy levels and increased pain. Limited energy is frustrating

when one wants to pursue a career, raise a family and lead a social life. Constant feelings of exhaustion may contribute to burnout and depression. Pain levels increased with tiredness. Adults receive less physiotherapeutic attention because they have fewer fractures. At the same time lack of energy, pain and fear of fractures make them less inclined to participate in sports and activities that could help to optimize their fitness.

Risk factors: fractures and long periods of inactivity damage the bones and cause wasting of muscles. Leg length discrepancy, scoliosis and bowing of bones cause parts of the body to receive excessive amounts of physical stress. Muscles need to contract with more force to produce the same result when tendons and joints are weaker. Hearing problems and the need to be aware of dangers take a constant toll on concentration. Cardiovascular problems play a role in some. Once fracture rate declines after puberty young adults like to believe OI is a thing of the past. The belief can lead to overburdening. People with less visible forms of OI experience social pressure to perform as if they were completely healthy.

VOI members exchanged tips on how to deal with these issues looking from the perspective of time-management, trying to balance between activity and rest, and thus limit the risks posed by overburdening on the one hand and inactivity on the other. Tips were clustered into 8 solution strategies:

- Rest: good night rest, relaxing activities, meditation, powernaps.
- Activity: regular exercise without overdoing it, activity keeps your mind of the pain.
- Balance activity and rest: Don't go till you drop, build in breaks, protect rest moments (unplug the phone!).
- Prioritize: skip things and people that drain energy.
- Adapt activities: delegate heavy parts, plan efficient, use aids, occupational therapy.
- Support: explain problems that are not visible, build friendships, join an OI group.
- Psychology: enjoy life, exert control, lower demands, choose for yourself even when that is less "social".
- Medical: watch your weight, eat healthy, pain medication, stay updated on OI

TRS - National Resource Center for Rare Disorders

Lena Lande Wekre, MD

TRS, National Resource Center for Rare Disorders, Sunnaas Rehabilitation Hospital, Norway

TRS is a national resource centre that offers services to seven diagnostic groups of rare, congenital disorders; Osteogenesis Imperfecta is one of them. The centre is a part of the Sunnaas Rehabilitation Hospital, and is located just outside Oslo, Norway.

The centre works to increase knowledge about Osteogenesis Imperfecta, and offers guidance and counselling on medical, psychological, social and educational issues.

No medical referrals are required to use the services of the centre, and all services at TRS are free of charge for the users. Multidisciplinary approach is used, and the centre offers life-long services to all age-groups.

OIFE member associations



Denmark



Portugal



Mexico



The Netherlands



Ecuador



Germany



France



Belgium - Franc.



Belgium – Flemish



Peru



Switzerland



Italy



PadrinoS-OI



UK



Norway



Australia



Croatia



Finland



USA



Georgia



HOI



Sweden



Spain