

---

## 3rd CNE International Cystinosis Conference

### How to improve muscle function in cystinosis: **IMPACT study**

Christian Koepl  
Physiotherapist B.A.  
Interdisciplinary Cystinosis Clinic  
RoMed Kliniken Rosenheim

# Galileo training

## Galileo plate

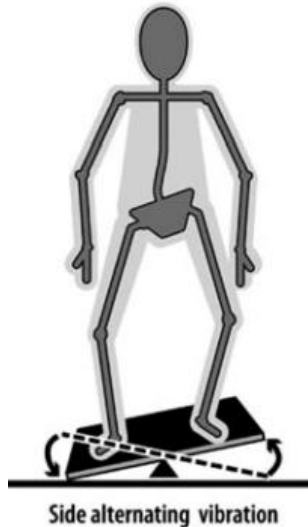


## Galileo dumbbell



### Whole-Body Vibration (WBV) is an effective and safe application

- Strengthening the muscles in a short training time
- Exercises can be done at home



- Hoyer-Kuhn H et al. A specialized rehabilitation approach improves mobility in children with osteogenesis imperfecta. J Musculoskelet Neuronal Interact. 2014;14(4):445-453.
- Matute-Llorente A. Effect of whole-body vibration therapy on health-related physical fitness in children and adolescents with disabilities: a systematic review. J Adolesc Health. 2014;54(4):385-396.

## Primary objectives

Does regular home practice, using Galileo vibration training platforms or vibrating dumbbells

- positively impacts muscle strength
- and increases cardiorespiratory performance in cystinosis patients.

## Secondary objectives

Does the training

- result in a general increase in everyday activity
- and improvement in the quality of life.
- Is it possible to integrate the training into the day-to-day life of chronically ill patients.

## Study group

Inclusion criteria

confirmed diagnosis of cystinosis  
children, prepubertal 5-13 years  
and adults > 18 years  
informed consent

Exclusion criteria

likelihood of serious intervention  
(e.g., transplantation, orthopedic intervention )  
contraindication according to the Galileo manual

## Study design

Randomized controlled study

Matched-pair design

(age, sex, weight, length, renal function,  
dialysis, previous surgery)

Intervention Group

n= 20 (Galileo Plate)

Control group

n= 20 (Galileo dumbbells)

## Data basis

**Accelerometric data (Pedometer – 7 days)** (before M1, M2, M3, M4)

**M<sub>1</sub>**

### Baseline measurement

Interdisciplinary cystinosis clinic (Rosenheim)  
(including laboratory data, pQCT measurement)

**M<sub>1,2,3,4</sub>**

**Jumping power : Leonardo Mechanograph<sup>®</sup>, single two- legged jump (3x)  
multiple one leg hopping (3x), Heel rise test (3x)  
Grip strength and Finger strength: Keygrip, Two-point-grip (right and left (3x))  
6-minute walking test (2x)  
Balance tests (Romberg, Semi tandem, Tandem and one leg stand)  
Surveys: Lifestyle (structured questionnaires)  
Quality of life questionnaires (SF-36 or KINDL-R)  
Patient training diaries**

**M<sub>2</sub>**

after 14 weeks training (Mainz and Milan)

**M<sub>3</sub>**

after 28 weeks (Mainz and Milan)

**M<sub>4</sub>**

after 12 months (= M<sub>1</sub>) (Rosenheim)

# IMPACT

<b>Randomization</b>	training platform/dumbbells LMU Munich
Initial training course Delivery	Rosenheim of the training tools to the patients' homes
<b>14 weeks of training</b>	10 short training sessions/week
<b>Exercises</b>	training platform (n= 4) dumbbells (n= 3) each exercise 30 sec. - 2 min.
<b>Training session</b>	5 -10 minutes max. 2 training session/day
<b>Training Frequency</b>	20 Hz up to 28 Hz
Regular supervision	Physiotherapist or sports student



## **Due to COVID**

the study group was separated into two groups

- adults                      start 09/2020 - last visit 09/2021
- children                    start 09/2021- last visit 09/2022

**and patients had less overall daily activity.**

# IMPACT – study participants (adults)

Parameter	Number of patients	male	female
Number of patients	23 (100 %)	14 (60.87 %)	9 (39.13 %)**
Age at last visit	28.09	29.15	26.79
Age at diagnosis (month)*	17 [6 - 39]	18 [8 - 39]	15 [6 - 27]
ESRD:	14 (60.87 %)	8 (57.14 %)	6 (66.67 %)
ESRD: age**	12.75 [7.14 - 24.81]	13.52 [7.14 - 24.81]	11.87 [9.06 - 23.23]
No cysteamine therapy	5		
5-10		1	3
> 15 years			1

\* Median [Minimum - Maximum]

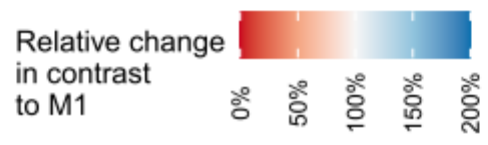
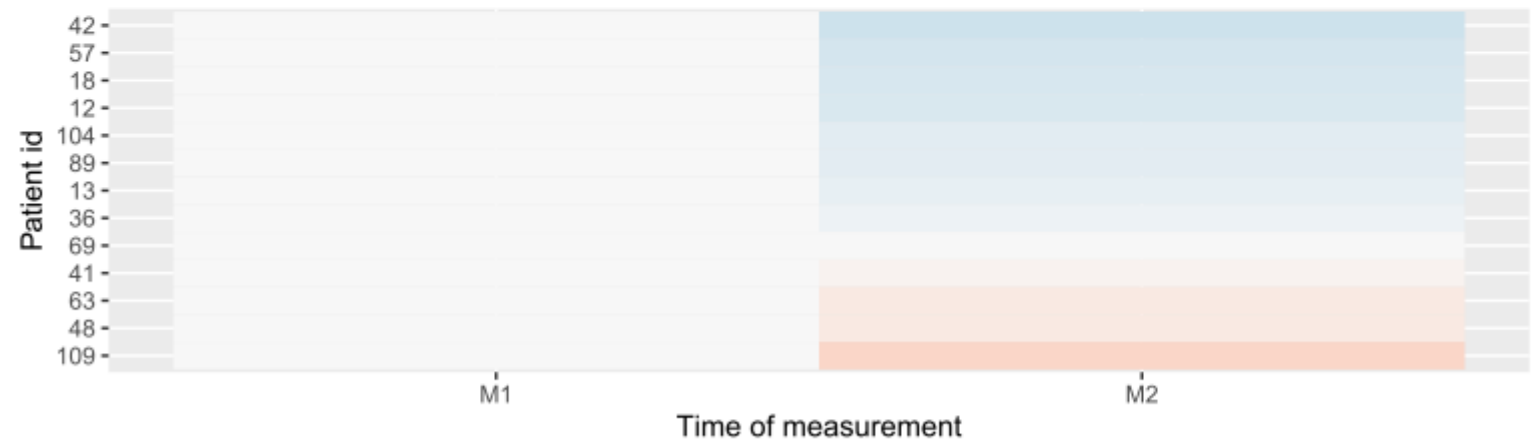
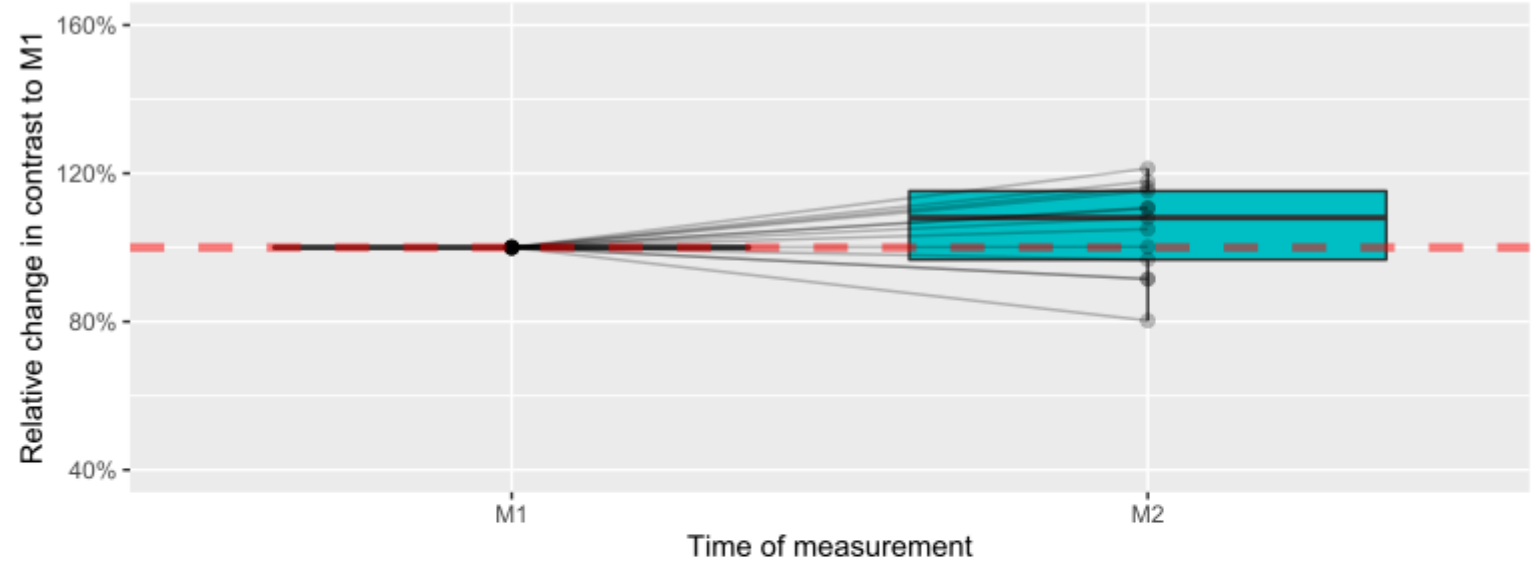
\*\* Mittelwert [Minimum - Maximum]

\*\*\* drop out n=1



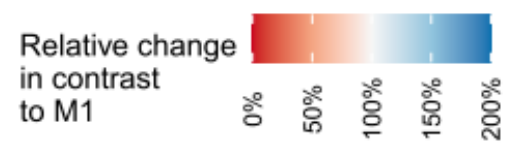
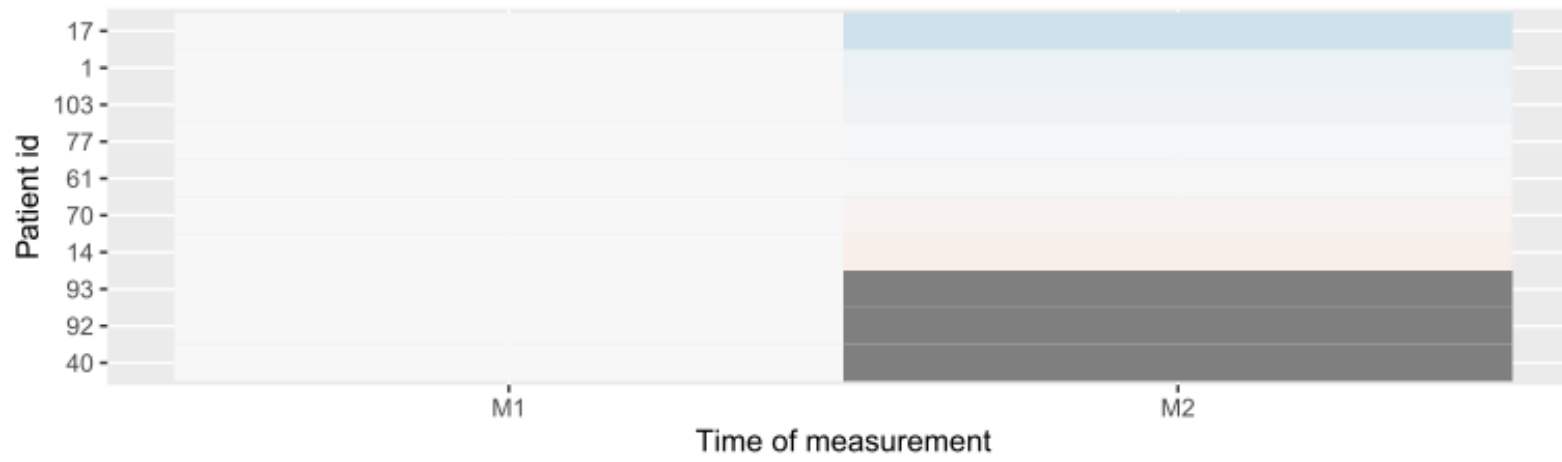
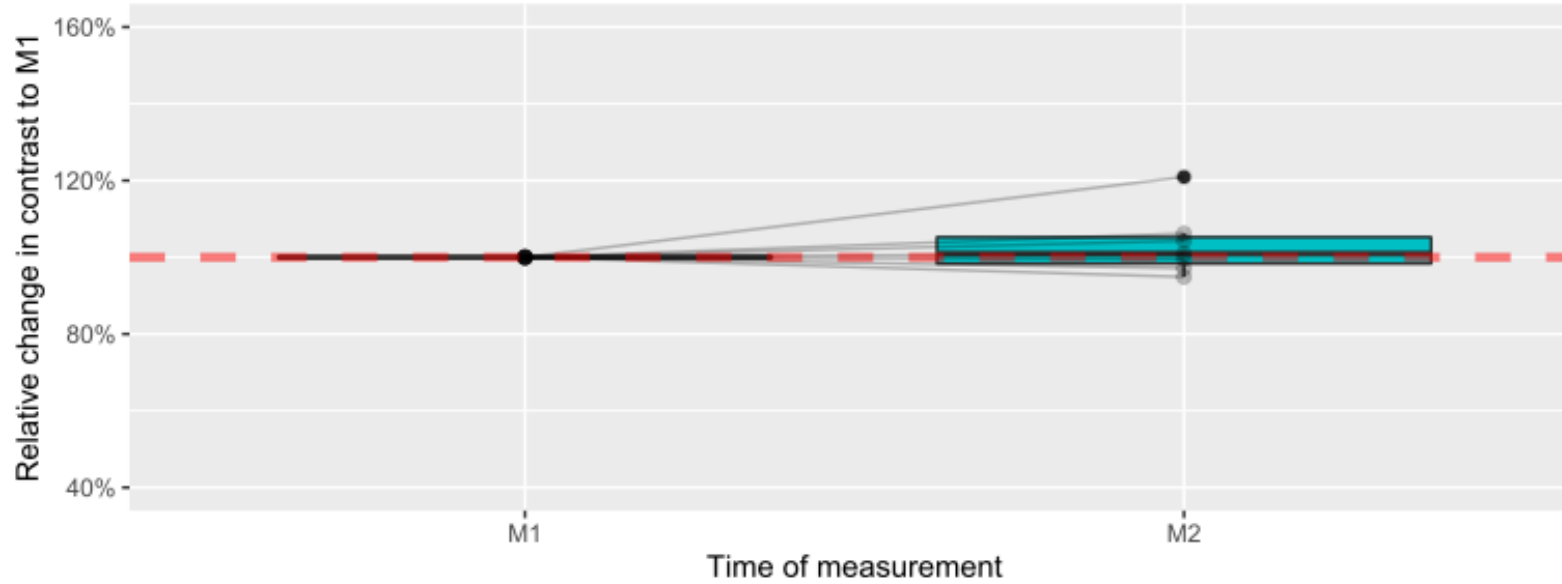
# Gripstrength: relative to M1

Training device: dumbbell



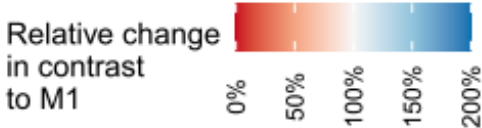
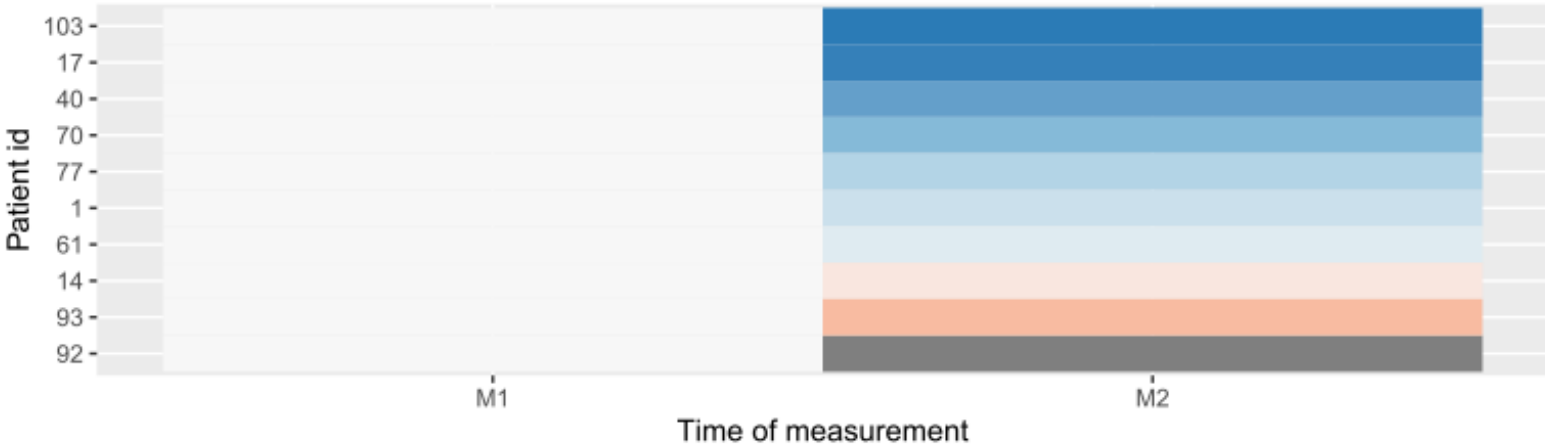
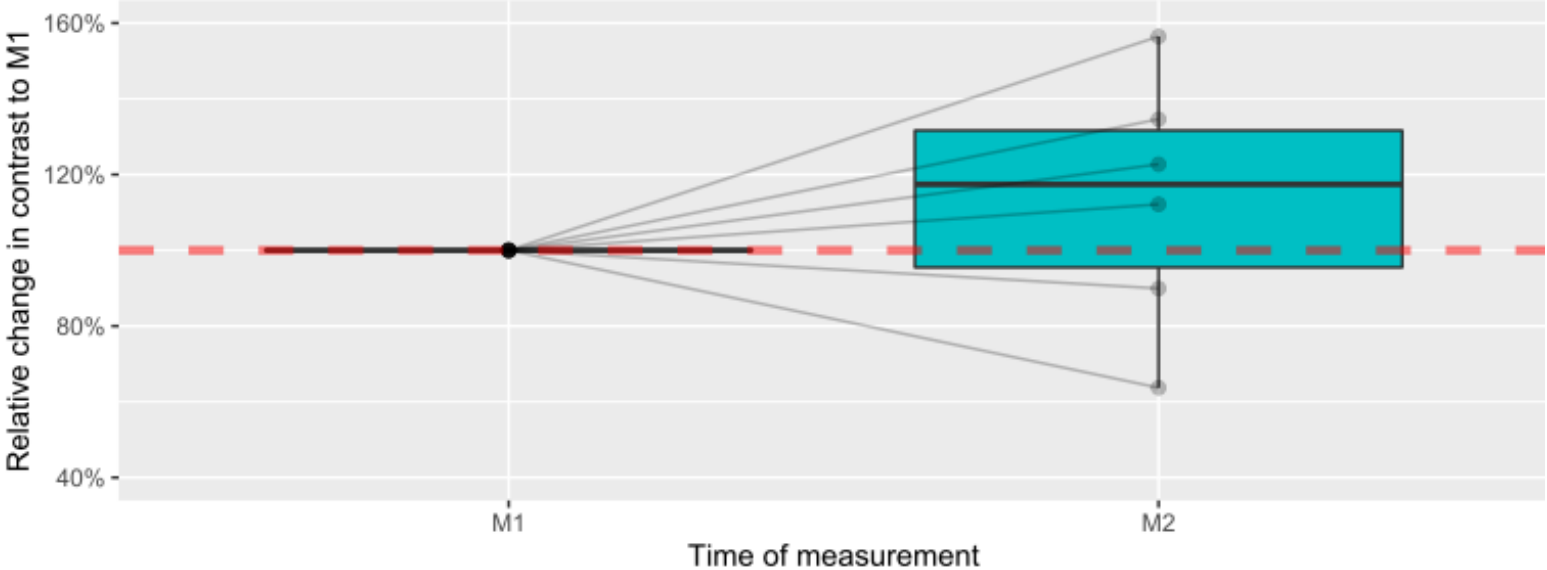
# Single two leg jump: relative to M1

Training device: platform



# Semitandem eyes opened: relative to M1

Training device: platform



# Training diaries

The participants noted their completed training sessions with comments in a diary

A large proportion of the participants stated that they had trained very thoroughly and regularly

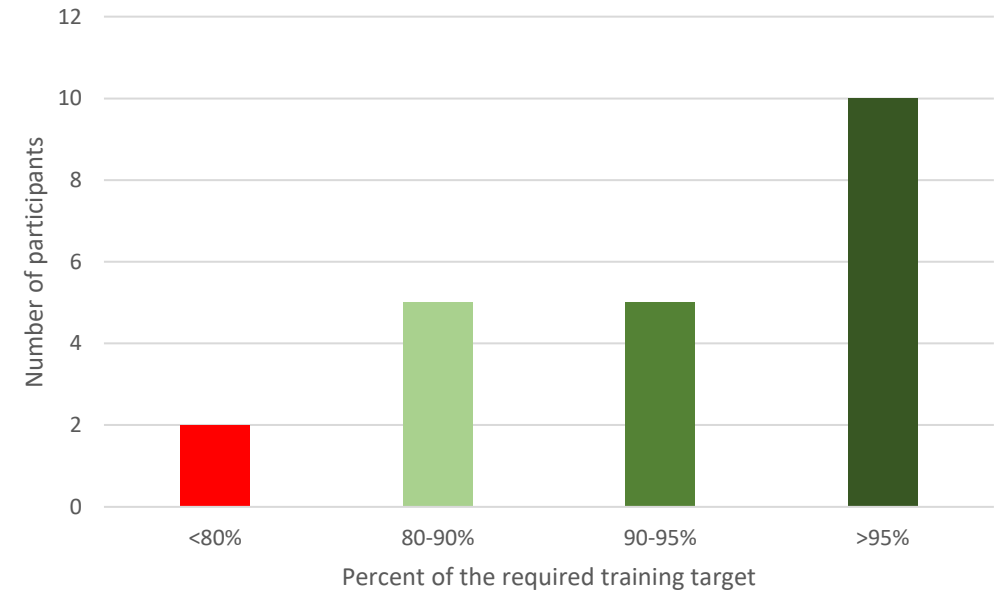
Training period: 14 weeks, 10 units/week

Adults:

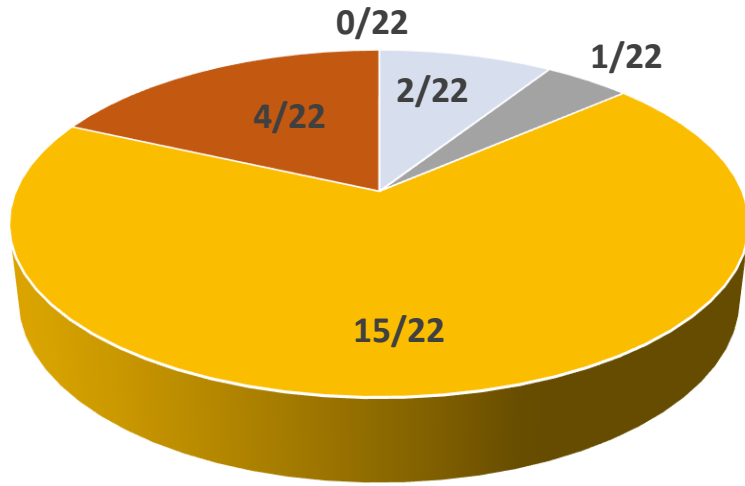
more than 95% of the required training target: 10 / 23

more than 80% of the required training target: 20 / 23

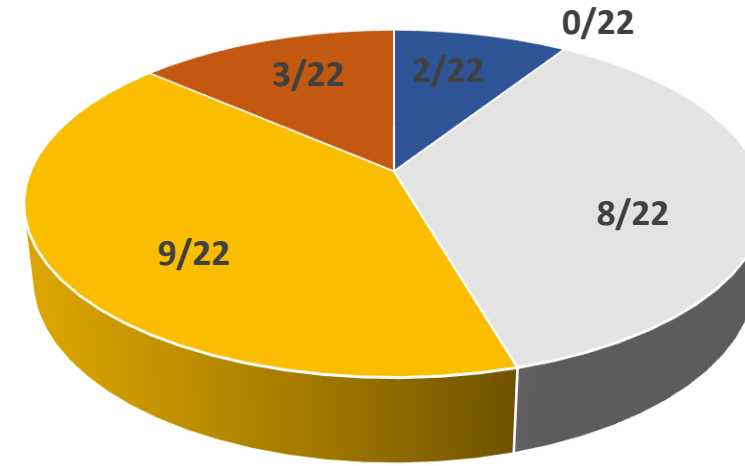
All participants reported no serious side effects or accidents during the training or serious discomfort caused by the training.



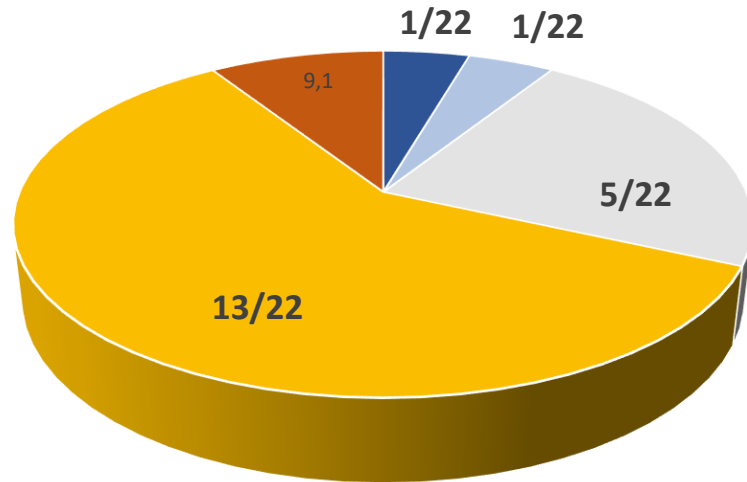
# Integration into every day life



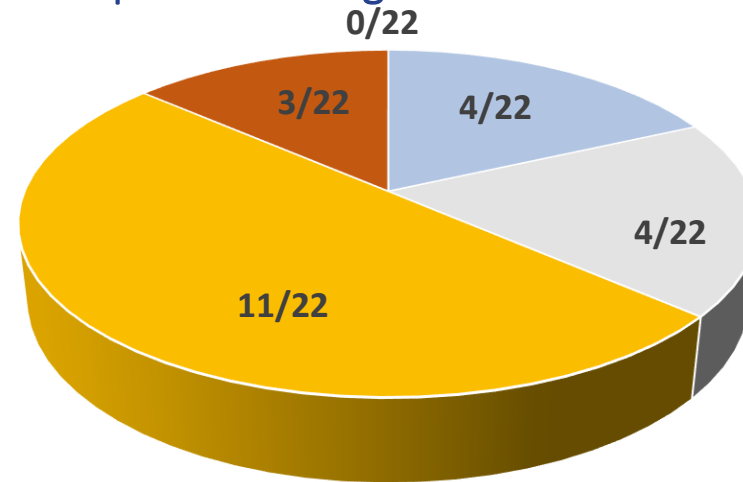
I was able to fit the training into my everyday life.



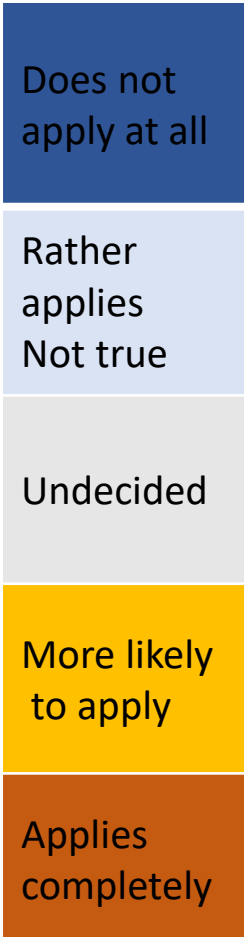
I have noticed positive changes because of the training.



I would continue to do this training regularly.



I now resolve to continue to do sport/exercise regularly.



---

Regular training could be integrated well into the daily life of the participants.

The number of training sessions per week was too much and should be reduced.

Improvements in muscle strength and balance could be achieved through the training.

Christian Koeppel

Anne-Laure Boulesteix, Tim Landfarth

Susanne Bechtold-dalla Pozza

Inga Jarosch

Harald Schubert

Johannes Willenecker

Nadine Herzig

Katharina Hohenfellner

Dep. of Physiotherapy, SPZ, Traunstein

Department of Medical Informatics, Biometry and Epidemiology (IBE), Munich

Dep. of Endocrinology, Dr. v. Haunersche Kinderklinik, Munich

Schoen Clinic, Schoenau a.K.

Novotec Medical GmbH

Novotec Medical GmbH

Dep. of Orthopedics Schoen Clinic Harlaching, Munich

RoMed Kliniken, Pediatric Nephrology, Rosenheim

Tessa Schneeberger

Daniela Leitl

Jessica Wendel

Andreas Zech

Moritz Berghaus

Pauline Käser

Schoen Clinic, Schoenau a.K.

Schoen Clinic, Schoenau a.K.

Schoen Clinic, Schoenau a.K.

Schoen Clinic, Schoenau a.K.

Student, University of Frankfurt

Physiotherapist, Rosenheim

Heike Holla

Sonja Froschauer

Study nurse, Cystinosis Foundation, Germany

Cystinosis Foundation, Germany

Thank you  
for your attention



CYSTINOSIS  
RESEARCH NETWORK

CYS  
CYSTINOSE  
STIFTUNG



cystinosis  
ireland