

**PRECEPTORSHIP PROGRAMME ON TREATABLE LYSOSOMAL STORAGE DISEASES (LSDs) -
MANAGEMENT AND TREATMENT**

**29th of June – 1st of July 2020
Salzburg Medical Simulation Center, Austria**

Organizers:

Dr. med Christina Lampe
Prof. Maurizio Scarpa MD PhD
PD Florian Lagler, MD

Institute of Metabolic Diseases of the Paracelsus Medical University
Clinical Research Center Salzburg GmbH

Do you sometimes think that managing patients with lysosomal storage diseases is too complex and work-intensive to be covered in addition to all your other tasks?

AND – Do you feel that the many disease complications arising during LSDs management go beyond your clinical field of expertise?

Well, most probably your concerns are legitimate, because the management of patients with LSDs commonly demands interdisciplinary teams of many different specialists. Metabolic physician, neurologists and neuropsychologist, ENT physician, cardiologist, general and neuro surgeon are only the most commonly needed specialists. In an ideal world all of the specialists in such teams are LSD experts. But this is rarely the case.

The next best organizational model is an interdisciplinary team lead by a LSD expert who is capable to lead and coordinate the team. To do so, you need to know about medical challenges and solutions in these diseases. Moreover you need to be competent in setting up and implementing safe and efficient working conditions and processes for critical and potentially critical situations.

Our preceptorship programme offers you the knowledge and training you need to provide state of the art medical care in an interdisciplinary team to your LSD patients.

We are an international faculty of very experienced LSD experts from different disciplines around Dr. Christina Lampe, Professor Maurizio Scarpa and PD Dr. Florian Lagler, who created this programme in 2014 – in the first step with a focus on Mucopolysaccharidoses.

Since that we have welcomed participants from all over the world and from rather diverse working environments and health systems. Culture, human and technical resources, educational and training standards as well as many other factors that impact clinical practice differ substantially in the different working environments of our participants. So it is very obvious that there is no single best solution to most of the challenges.

Therefore our preceptorship is designed to provide information and opportunity to make experiences that are of highest relevance for the specific clinical setting of the participants.

The following three elements are most important for achieving this goal:

1. Our faculty consists of international LSD experts with many years of experience in metabolic medicine, genetics, neurology, orthopedics, surgery etc.. They come from different settings and disciplines and provide a deep insight in the clinical challenges of LSDs and the tools and protocols they use in their own disciplines to master these.

2. Participants are invited to present their most challenging cases and discuss it with the experts.

3. In the simulation training we simulate critical cases in a training environment that corresponds with the everyday clinical setting of the participants. So they can apply and improve their skills and knowledge und realistic circumstances.

So after your participation, you will know what diagnostic and therapy challenges you can expect with your patients and how you can best prepare yourself and your team to master these in your own setting.

The Agenda

1st day – 29th of June 2020

- 13.00 Welcome and introductory round
- 13.20 Introduction into Simulation- and Communication Training
- 13.40 Short overview on Communication Challenges in Fabry disease
- 14.00 Overview on Principles of Communication
- 14.20 Communication Scenarios
- 15.30 Coffee
- 15.45 Communication Scenarios continued
- 18.15 Basic Principles of Patient Safety in LSD and other diseases
- 18.30 Familiarization with the Simulation Environment
- 19.00 Wrap Up
- 19.30 Dinner

2nd day – 30th of June 2020

- 9.00 Introductory round with Simulation Team
- 9.30 Simulation Warm Up
- 10.00 Management ERT and mild IRR
Presentation „ERT management and management IRR in all LSDs“
Simulation Scenarios
Presentation „Airways in MPS and other LSDs“
- 13.00 Lunch
- 14.00 Management Perioperative Complications in MPS
Simulation Scenario
Presentation „Anesthesia in MPS“
- 15.30 Coffee
- 15.45 Lung and Airways in Pompe diseases and other LSDs
Simulation Scenario
Presentation „Lung and Airways in Pompe diseases and other LSDs “
- 17.15 Presentation “Transition in LSDs”
- 17.45 4 Case Presentations
- 18.45 Wrap Up
- 19.30 Dinner

3rd day – 1st of July 2020

- 8.30 Musculoskeletal System in LSDs
Presentation “Bone Disease and Orthopedics in LSDs”
with corresponding case presentation
- 9.30 Neurology in LSDs
Presentation „Neurology in LSDs“
with corresponding case presentation
- 10.30 Coffee
- 10.45 Case Presentations
- 11.45 Simulation Scenario
- 12.30 Lunch
- 13.30 The Heart in LSDs
Presentation “The Heart in LSDs”
Simulation Scenarios
- 15.30 Feedback and Wrap Up
- 16.00 Farewell

By reading the agenda of our preceptorship you may have wondered: Why should I do simulation training for critical situations in LSD patients? Neither my clinical responsibility includes emergency cases or intensive care. Nor have I ever experienced a critical situation in one of my LSD patients. These must be very rare incidents.

Well, still cardiac and respiratory incidents are by far number one mortality causes in MPS, Fabry and Pompe patients and not always these are unpreventable.

This is why our programme is designed to make you familiar with factors and situations that can lead into critical situations with MPS patients. Even more importantly we will discuss how preventive measures, early recognition and competent management can prevent patient harm.

BUT patient safety research clearly indicates that medical knowledge is only one success factor needed AND as a matter of fact 70% of preventable medical harms are caused by failures of situational awareness, team work, leadership, communication etc. You've probably experienced yourself that situational awareness is particularly challenging in rarely occurring events and that efficient team work can be hard if several team members don't know the patient and the disease very well. Unfortunately, both are rather the rule than the exception in LSDs.

For that reason, in our programme, we will simulate real LSD cases with patient actors and simulation manikins so you can practice your knowledge and skills in realistic high stake situations. You will learn how to identify and minimize the risk for critical situations and how to deal with overwhelming situations. You will reflect your experiences within the video-debrief and discuss factors that contributed to success or failure with your co-delegates and the experts. By this you will create a plan how to implement the principles of state of the art LSD patient management and patient safety into your daily work. Clearly, experiencing critical situations under observation causes stress in all of us. But we are competent in keeping the stress on level that improves learning and retention and in creating a positive learning experience.

Participants: The participants, in a limited number of max 24 physicians (3 physicians each scenario, by bringing in each scenario 1 junior, 1 senior and one specialist), should be fluent in English and experienced in the management of these patients and providing ERT. Participants are required to attend for the entire length of the preceptorship.

Registration:

Please contact Ms. Andrea Lughofer (a.lughofer@cracs.at) if you're interested – you'll get a link and login data for registration on www.cracs.at

We are looking forward to welcome you in Salzburg.

Dr. med. Christina Lampe, MD
Prof. Maurizio Scarpa, MD, PhD
PD Dr med. Florian Lagler, MD

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