



SWISS **LUNG** ASSOCIATION



Lay Summary

Management of upper airway disease in PCD: current practices and development of guidelines

Research management

1. Main applicant: PD Dr Myrofora Goutaki, MD-PhD, Institute of Social and Preventive Medicine, University of Bern, Switzerland
2. Partner institutions involved in the research project: Clinical centres contributing patients to the Ear-Nose-Throat (ENT) Prospective International Cohort of Patients with PCD (EPIC-PCD)

Short title

Upper airways care in primary ciliary dyskinesia

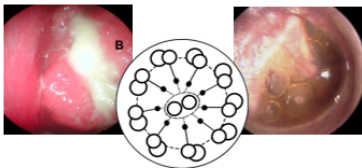
Synopsis

Upper airway problems are common in primary ciliary dyskinesia, but their management varies greatly between clinics. We aim to describe current clinical practices and develop international consensus guidelines for use in different settings, including recommendations for referral to specialists.




SWISS **LUNG** ASSOCIATION


**Management of upper airway disease
in **primary ciliary dyskinesia (PCD)****



Chronic upper airway
problems in all patients



BUT no recommendations
for management or
referral to specialists



Develop guidelines to
support clinical decisions
by treating physicians

Project description

- **Context / background:**

Primary ciliary dyskinesia (PCD) is a rare genetic disease that causes different problems, especially from the airways. In few countries, patients are treated in national centres by large teams including different specialists. Away from these centres however, and in most countries, patients still do not receive optimal care. Respiratory specialists are required to make decisions also about treating upper airway problems patients face. However, management of the upper airways is not standardised and there are no existing treatment guidelines.

- **Objectives and methods**

This project aims to describe how upper airway problems are currently treated in children and adults with PCD and how these treatment decisions are made. We will use data from a large international clinical cohort study of patients followed-up regularly in specialist PCD centres. The study collects details on tests, examinations, and prescribed treatments from the clinics, and information on symptoms directly from patients, and parents of children with PCD. These findings will inform an international expert panel to develop management

guidelines, using a series of surveys to reach consensus. We want to provide recommendations for different settings, including not only management by specialists, but also regular care.

- **Significance**

Upper and lower airway problems should be managed together as common entity in PCD. It is therefore important to develop guidelines also for respiratory physicians who are usually the main treating physicians of patients with PCD, to improve routine care and have clear recommendations about when a specialist referral is required. This project is timely and will bring together expertise of different specialists, healthcare professionals, and patients themselves to ensure the guidelines are useful in clinical practice for different settings.

- **Start and duration**

Dec 2, 2024; 24 months.

- **Amount of funding**

The SLA makes available a total amount of **93 000 CHF**

