ISSUE 3| AUGUST 2021

BEAT-PCD NEWSLETTER

What's inside:

We want to hear from you

We value the opinion of our readers and we are looking forward to receive your feedback about this newsletter.

Please send us a personal email with your suggestions or comments to the following address:

Attn: Stephanie Gallant
BEATPCD@ersnet.org

ERS EUROPEAN RESPIRATORY SOCIETY
every breath counts

The BEAT-PCD Clinical Research Collaboration (CRC) is funded by the European Respiratory Society.

ANNUAL EVENTS

Online BEAT-PCD training school 2021

September 4th 2021
13:00 CET/12.00 UK

Online BEAT-PCD annual meeting 2021

September 9th 2021
13:00 CET/ 12.00 UK

EVENT AGENDAS INCLUDED

save the date

Registrations for both events, agendas and further information is available on our website:
http://beat-pcd.squarespace.com/

The BEAT-PCD annual events take place on either side of ERS international congress which for 2021 will be a virtual event. Additional info for the ERS virtual event is provided here.

Message from the BEAT PCD chairs

Welcome to the 3rd edition of the BEAT-PCD newsletter.

It is with great pleasure that we invite you in the upcoming online BEAT-PCD events. The BEAT-PCD Training School will take place on September 4th while the annual BEAT-PCD event will take place on September 9th. The Training School will include sessions on PCD diagnosis and management while the annual event will provide updates on PCD genetics, international collaborative projects, impact of COVID-19 on PCD and many more.

In this edition, you may find detailed agendas for both events as well as a list of all talks and poster presentations that are relevant to PCD and will take place in the ERS virtual meeting (4-8 September). In addition, you can read about recent scientific publications from PCD collaborative projects as well as find out more about special issues focusing on diagnostic and therapeutic approaches of PCD.

Looking forward meeting you online soon and we hope that you will enjoy reading this edition.

Amelia & Myrona

BEAT-PCD Training School - Agenda

Online Event - ZOOM

Saturday September 4th 2021

13:00 - 17:00 Central European Time

13:00 - Welcome

Amelia Shoemark

Myrona Goutaki

Claire Hogg

1st session:

Chairs: Suzanne Crowley and Ana Reula

13:15 - Recognition and diagnosis of PCD in neonates and infants

Claire Hogg, Royal Brompton Hospital

Reena Bhatt, Royal Brompton Hospital

14:00 - nNO measurement in preschool age

Jane Lucas, University of Southampton

Nicole Beydon, Hospital Armand-Trousseau

14:45 - Break

2nd session:

Chairs: Eleonora Dehlink and Nagehan Emiralioglu

15:00 - Recognising and treating PCD in adults with bronchiectasis

Amelia Shoemark, University of Dundee

James Chalmers, University of Dundee

15:45 - Diagnostic dilemmas – route to resolution case reviews

Marie Legendre, Sorbonne University & Inserm

Hannah Mitchison, UCL Institute of Child Health

16:30 - Closing Remarks

Register here:

https://www.eventbrite.co.uk/e/beat-pcd-training-school-tickets-161086960435?aff=ebdsoporgprofile

BEAT-PCD annual meeting 2021 - Agenda

Online Event - ZOOM
Thursday, September 9th, 2021
13:00 - 18:00 Central European Time

13:00 - Welcome, meeting rules and introduction
Amelia Shoemark and Myrona Goutaki

13:15 - Work package summaries (10 min each including questions)
Work Package Leads

14:15 - 30 minute break/interactive sessions

14:45 - PCD genetics and molecular therapies (3x10 min talks including questions)
Chairs: Heymut Omran and Mathieu Bottier

Development and first results of the BEAT-PCD international Primary Ciliary Dyskinesia gene variant database: CiliaVar - *Amelia Shoemark*

High-content screening for rare respiratory diseases: read through therapy in primary ciliary dyskinesia - *Chris O'Callaghan*

A messenger RNA (mRNA)-based therapeutic designed o treat PCD - *Caroline Woo*

15:15 - Genome-wide analysis of PCD gene variant frequencies: estimated prevalence and ethnic heterogeneity (15 min, 5 min questions) - *Will Hannah*

15:35 - Cohort studies on PCD (2 x 10 min talks including questions)
Chairs: Claudia Kuehni and Renate Kos

The EPIC-PCD study: time to focus on the upper airways - *Myrona Goutaki*
COVID-PCD: results and future plans - *Eva Pedersen*

15:55 - Highlights from the ERS meeting (5 x 10 min talks including questions)
Chairs: Jane Lucas and Rob Hirst

The short term influence of chest physiotherapy on lung function parameters in pediatric bronchiectasis - *Mieke Boon*

The international primary ciliary dyskinesia (PCD) registry reports genotype/phenotype correlations - *Johanna Raidt*

Service delivery models for primary ciliary dyskinesia, an international comparison *Bruna Rubbo*

Diagnosis of Primary Ciliary Dyskinesia: Discrepancy according to different algorithms - *Mirjam Nussbaumer*

The prevalence of primary ciliary dyskinesia (PCD) in adult patients with bronchiectasis in Dnipro (Ukraine) - *Kseniia Suska*

16.45 - Closing Remarks
Amelia Shoemark and Myrona Goutaki

17:00 -Break out meetings for specific projects (until 17:30-17:45)

Register here:
https://www.eventbrite.co.uk/e/beat-pcd-annual-meeting-tickets-161083887243?aff=ebdsoporgprofile

Detailed PCD programme at the international virtual ERS 2021 congress

All times refer to Central European Time (CET)
*All e-posters are available at the conference platform after August 23rd.

Sunday 05/09/2021

Oral Presentations

Session: Psychological and behavioural issues in respiratory care

Mental health during COVID-19 in Dutch adolescents and adults with cystic fibrosis (CF) or primary ciliary dyskinesia (PCD) and their caregivers. (Channel 4, 11:45)
Speaker: M. Verkleij, Netherlands

Session: Mechanistic pathways in chronic and new lung diseases
Diagnosis of Primary Ciliary Dyskinesia: a multi-center experience. (Channel 5, 15:25)
Speaker: R. Hjeij (Germany)

Session: Bronchiectasis heterogeneity: assessing the endotypes
Mapping inflammatory endotypes of bronchiectasis associated with impaired mucociliary clearance. (Channel 4, 17:00)
Speaker: M. Bottier (United Kingdom)

E-Posters Discussion

Session: Posters in Pediatric Bronchology (13:15-14:15)

Does the type of bacterial colonization in lower airways affect the lung function in patients with Primary Ciliary Dyskinesia (PCD)?
Speaker: V. Fein (Germany)

Anxiety of children with primary ciliary dyskinesia and their mothers during COVID-19 pandemic.
Speaker: P. Asfuroglu (Turkey)

Genotype-phenotype correlation in a group of patients with primary ciliary dyskinesia in Italy.
Speaker: L. Petrarca (Italy)

The short term influence of chest physiotherapy on lung function parameters in pediatric bronchiectasis.
Speaker: M. Boon (Belgium)

Session: Burden and epidemiology of COVID-19 (13:15-14:15)

SARS-CoV-2 infections in people with PCD.
Speaker: E. Pedersen (Switzerland)

Session: Lung function evaluation and assessment of airway diseases (13:15-14:15)

Genetic mutations and ciliary ultrastructure as possible determinants of lung function evolution in primary ciliary dyskinesia: a 40-year observational study.
Speaker: M. Holgersen (Denmark)

Session: Peripheral muscle function, exercise capacity and functional status in respiratory disease (13:15-14:15)

Exercise capacity, peripheral muscle strength, and balance are related in primary ciliary dyskinesia
Speaker: H. Sonbahar Ulu (Turkey)

Detailed PCD programme at the international virtual ERS 2021 congress

All times refer to Central European Time (CET)
*All e-posters are available at the conference platform after August 23rd.

Monday 06/09/2021

Oral Presentations

Session: Patient-reported outcome measures (PROMs): easy tools in the management of chronic respiratory diseases

A Parent Reported Quality of Life Measure for Young Children with Primary Ciliary Dyskinesia: QOL-PCDPR (Channel 2, 11:45)
Speaker: L. Behan (Ireland)

Session: Paediatric lung disease: novel mechanisms, methods and translational studies
Defects in outer dynein arm docking machinery cause primary ciliary dyskinesia. (Ch 4, 11:25)
Speaker: R. Hjeij (Germany)

E-Posters Discussion

Session: Pathogenic progress in airway diseases (13:15-14:15)

Transcriptome analysis of ciliary differentiation in airway epithelium
Speaker: J. Legebeke (United Kingdom)

Session: Environment and air pollution (13:15-14:15)

Bushfire smoke exposure cause impairment to airway muco-ciliary function
Speaker: R. Suryadinata (Australia)

Session: Bronchiectasis and pleural infection (13:15-14:15)

Vestibular function in adult patients with primary ciliary dyskinesia.
Speaker: N. Winthereik (Denmark)

Session: Back to infection basics (13:15-14:15)

Ciliary videomicroscopy at room temperature might lack sensitivity for primary ciliary dyskinesia diagnosis.
Speaker: N. Bricmont (Belgium)

Session: Bronchiectasis (13:15-14:15)

International BEAT-PCD Consensus Statement for infection prevention and control in Primary Ciliary Dyskinesia in collaboration with ERN-LUNG PCD Core Network and patient representatives.
Speaker: J. Marthin (Denmark)

Characteristics and outcomes of adults with primary ciliary dyskinesia (PCD): an EMBARC/BEAT-PCD analysis
Speaker: A. Shoemark (United Kingdom)

Systematic review of lung clearance index (LCI) in non-cystic fibrosis (CF), non-primary ciliary dyskinesia (PCD) bronchiectasis (Bx)
Speaker: C. Hine (United Kingdom)

Session: Comorbidities in airway obstructive diseases (13:15-14:15)
The prevalence of primary ciliary dyskinesia (PCD) in adult patients with bronchiectasis in Dnipro.
Speaker: K. Suska (Ukraine)

The international primary ciliary dyskinesia (PCD) registry reports genotype/phenotype-correlations.
Speaker: J. Raidt (Germany)

Session: Cystic Fibrosis

In vitro effect of combined hypertonic saline and salbutamol on ciliary beat frequency of human nasal epithelial cells.
Speaker: L. Müller (Switzerland)

Detailed PCD programme at the international virtual ERS 2021 congress

All times refer to Central European Time (CET)
*All e-posters are available at the conference platform after August 23rd.

Tuesday 07/09/2021

Oral Presentations

Session: Causes and consequences of paediatric respiratory diseases

Respiratory symptoms of Swiss people with Primary Ciliary Dyskinesia. (Channel 4, 11:20)
Speaker: M. Goutaki (Switzerland)

The Palestinian primary ciliary dyskinesia (PCD) cohort: clinical, diagnostic and genetic spectrum. (Channel 4, 11:25)
N. Rumman (Palestine)

E-Posters Discussion

Session: Paediatric respiratory infections: chronic infection, vaccination and lung function (13:15-14:15)

Nasal Nitric Oxide Sampling In 0-5-year-old Patients With Cystic Fibrosis, Primary Ciliary Dyskinesia and Healthy Controls
Speaker: M. Kragh (Copenhagen, Denmark)

Session: What's new in respiratory problems in rare and systemic diseases (13:15-14:15)

Genetic analysis of 17 children with Primary Ciliary Dyskinesia (PCD)
Speaker: S. Iniesta González (Spain)

Session: Cystic fibrosis in children: various aspects (13:15-14:15)

Evidence for ciliary dyskinesia in cystic fibrosis
Speaker: R. Bonhiver (Belgium)

Session: Primary ciliary dyskinesia and tracheostomie (13:15-14:15)

Development and first results of the BEAT-PCD international Primary Ciliary Dyskinesia gene variant database: CiliaVar
Speaker: M. Rahma (Tunisia)

Diagnosis of Primary Ciliary Dyskinesia: Discrepancy according to different algorithms
Speaker: L. Müller (Switzerland)

Comparison Of Lung Clearence Index, Impulse Oscillometry And Spirometry in Children With Primary Ciliary Dyskinesia.
Speaker: P. Ergenekon (Turkey)

Service delivery models for primary ciliary dyskinesia, an international comparison
Speaker: B. Rubbo (United Kingdom)

Respiratory symptoms and physical activity in patients with Primary Ciliary Dyskinesia in Switzerland.
Speaker: Y. Lam (Switzerland)

Does nasal irrigation (NI) improve outcomes in children with primary ciliary dyskinesia (PCD)?: a scoping review.
Speaker: A. Harris (United Kingdom)

The role of genetics in diagnosing primary ciliary dyskinesia (PCD) in pediatric patients in Gothenburg, Sweden.
Speaker: C. Kavouridou (Sweden)

Case series of 25 children with PCD: evolution over the last 33 years.
Speaker: D. Quintero García (Spain)

Face mask usage against SARS-CoV-2 among people with primary ciliary duyskinesia
Speaker: E. Pedersen (Switzerland)

Detailed PCD programme at the international virtual ERS 2021 congress

All times refer to Central European Time (CET)
*All e-posters are available at the conference platform after August 23rd.

Wednesday 08/09/2021

Oral Presentations

Session: Emerging respiratory diseases: state-of-the-art studies of SARS-CoV-2 infection

A novel short isoform of ACE2 is expressed in ciliated airway epithelium and is induced by interferon and rhinovirus infection but not SARS-COV-2; implications for COVID-19 interferon treatment. (Channel 2, 11:30)
Speaker: C. Jackson (United Kingdom)

European Respiratory Society (ERS) International Congress 2021

Important information

Registration information:
https://www.ersnet.org/congress-and-events/congress/registration/

Full programme:
https://www.ersnet.org/congress-and-events/congress/ers-international-congress-programme/

ERS award winners 2021:
https://www.ersnet.org/the-society/awards-and-fellows/award-winners/

Featured Research

BEAT-PCD groups have been busy over the previous months completing collaborative projects to improve diagnosis and treatment for patients with PCD. Here we highlight some recent publications from the group:

International BEAT-PCD consensus statement for infection prevention and control for primary ciliary dyskinesia in collaboration with ERN-LUNG PCD Core Network and patient representatives

June K Marthin, Jane S Lucas, Mieke Boon, Carmen Casaulta, Suzanne Crowley, Damien M S Destouches, Ernst Eber, Amparo Escribano, Eric Haarman, Claire Hogg, Bernard Maitre, Gemma Marsh, Vendula Martinu, Antonio Moreno-Galdó, Huda Mussaffi, Heymut Omran, Petr Pohunek, Bernhard Rindlisbacher, Phil Robinson, Deborah Snijders, Woolf T Walker, Panayiotis Yiallouros, Helle Krogh Johansen, Kim G Nielsen

This article describes the first ever international consensus statement on infection prevention and control, intended specifically for PCD. Among others, it includes suggested actions for microbiological identification, indications for treatment of Pseudomonas aeruginosa, Burkholderia cepacia and nontuberculous mycobacteria and suggested segregation aspects aimed to minimise patient-to-patient transmission of infections whether in-hospital, in PCD clinics or wards, or out of hospital at meetings between people with PCD. The statement also includes segregation aspects adapted to the current coronavirus disease 2019 (COVID-19) pandemic. You can read the published manuscript **here**.

SARS-CoV-2 infections in people with primary ciliary dyskinesia: neither frequent, nor particularly severe

Eva S L Pedersen, Myrofora Goutaki, Amanda L Harris, Lucy Dixon, Michele Manion, Bernhard Rindlisbacher, Covid-PCD Patient Advisory Group, Jane S Lucas, Claudia E Kuehni

This article provides the first data on risk and severity of SARS-CoV-2 infections among study participants for the time period between 30 May 2020 and 5 March 2021. In the COVID-PCD study, people with PCD worldwide are followed weekly via online questionnaires. The incidence rate of COVID-19 and the proportion of participants infected were low, and the observed severity of infection mostly mild. People with PCD reported careful shielding behaviour which partly explains the results. You can read the published manuscript **here**.

Special Issue: "Primary Ciliary Dyskinesia: Genetics, Molecular Mechanisms, Diagnostic and Therapeutic Perspectives"

Journal: **International Journal of Molecular Sciences**
Guest Editors: Prof. Michał P. Witt, Prof. Ewa Ziętkiewicz, Dr. Zuzanna Bukowy-Bieryłło
Deadline for manuscript submissions: **30 December 2021**

You can find out more about the special issue and read the scientific articles already published **here**.

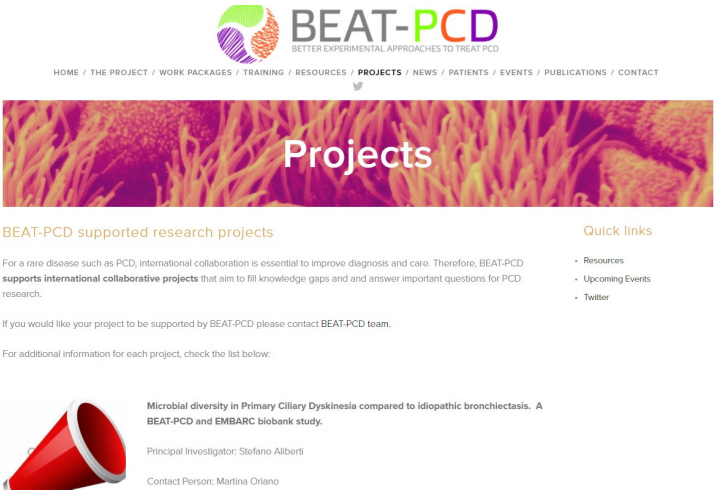
Special Issue: Progress in Diagnosing and Managing Primary Ciliary Dyskinesia

Journal: **Diagnostics**
Guest Editors: Dr. Thomas Burgoyne
Deadline for manuscript submissions: **31 July 2021 (closed)**

You can find out more about the special issue and read the scientific articles already published **here**.

BEAT-PCD Website

The BEAT-PCD website is available in English language and is updated regularly. It provides an overview of the network and describes the the project's aims and objectives, introduces the seven Work Packages, provides resources for both health professionals and patients and is frequently updated with BEAT-PCD related research projects, training activities as well as networking and other events.



In addition, the BEAT-PCD website aims to showcase all PCD related research, published from members of the network. To acknowledge the participation to BEAT-PCD, authors are invited to include the following statement in the declarations section of their manuscript:

"Study authors and data contributors participate in the BEAT-PCD clinical research collaboration, supported by the European Respiratory Society."

Find us online

BEAT-PCD ON SOCIAL MEDIA