



NEWSLETTER

6th EDITION, AUGUST 2018

INTRODUCTION

Welcome to the latest edition of the BEAT-PCD newsletter. This edition highlights the BEAT-PCD meeting which will be held Saturday September 15th, and the ERS Congress which will run 15-19th September, both in Paris. The BEAT-PCD meeting not only includes an exciting international programme, but also provides a great opportunity to relax together over lunch and an evening meal. PCD continues to be well represented at the ERS Congress through symposia and 37 oral and poster presentations. Please get in contact with the newsletter team if you have ideas for the next edition.

I look forward to seeing you in Paris!

Jane Lucas, Chair of BEAT-PCD

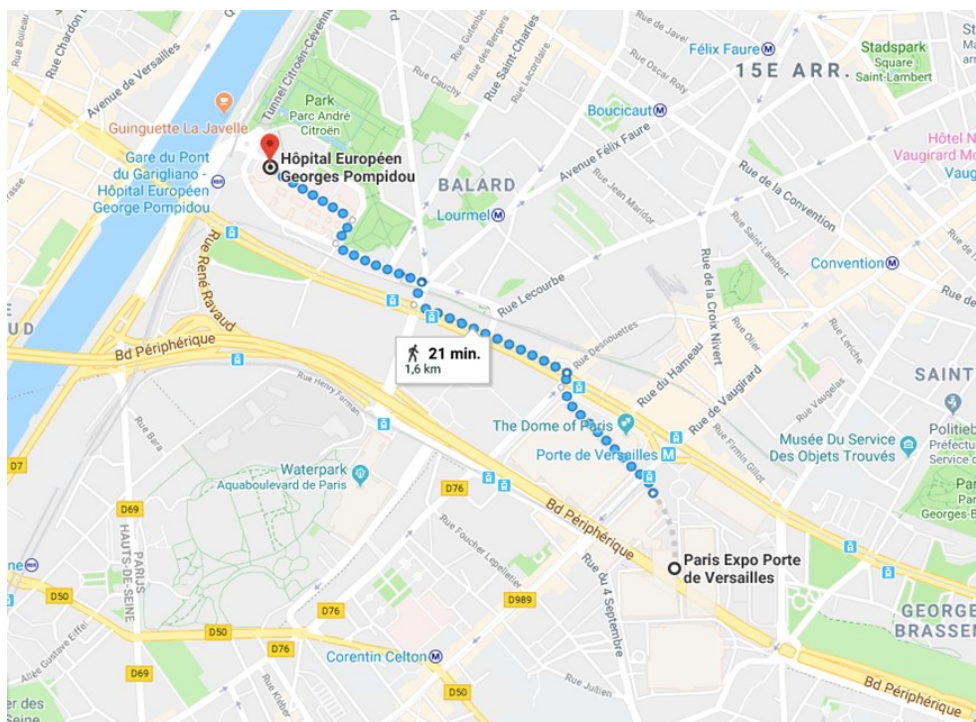


ERS PCD MEETING (15th SEPTEMBER 2018)

When: Saturday, Sept 15, 2018; 12.00-18:00

Location: Auditorium, Hôpital Européen Georges Pompidou

Address: 20 Rue Leblanc, 75015 Paris





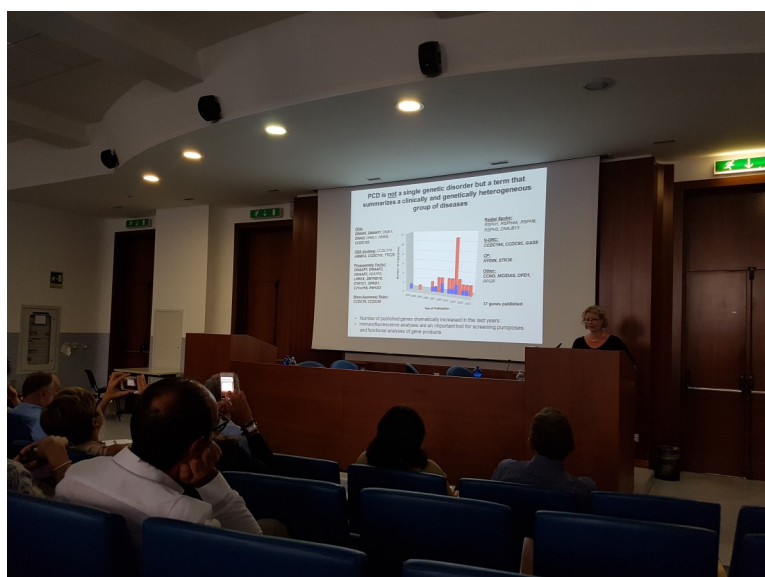
ERS PCD MEETING

The ERS PCD meeting will take place on **Saturday, 15th September 2018 in Paris**. The meeting will kick-off with a light lunch from 12pm, followed by brief updates on BEAT-PCD Action (Jane Lucas) and Training School (Claire Hogg).

June Marthin and Nicole Beydon will present the latest developments on nasal nitric oxide measurements in children, followed by a discussion on the usefulness of the diagnostic test in infants with suspected PCD. Adam Shapiro and Jane Lucas will discuss the diagnostic evaluation for PCD according to the (recently published) North American and the European guidelines, highlighting similarities and differences in diagnostic approach. A series of short presentations by researchers from different countries on the latest research findings and potential collaborative projects will follow. The meeting will conclude with a discussion led by Heymut Omran on a potential new COST-Action application. Participants will then have an opportunity to network with colleagues over an informal dinner.



Amelia Shoemark (top) and **Petra Pennekamp** (bottom) present the latest developments on immunofluorescence analysis in clinical practice at last year's ERS PCD meeting in Milan, Italy.





ERS PCD MEETING: AGENDA (PRELIMINARY)

12:00-13:00 Get-together with light lunch

Introduction and updates on BEAT-PCD

13:00 Introduction and greetings (**Bernard Maitre and Annick Clément, Paris**)

13:10 Updates on BEAT-PCD Action (**Jane Lucas, Southampton**) and BEAT-PCD Training school (**Claire Hogg, London**)

How useful is nNO for diagnosing PCD in infants? (two centres' experience and discussion)

Chairs: Estelle Escudier (Paris) and Florian Singer (Bern)

13:30 Choice of nasal nitric oxide technique as first-line test for primary ciliary dyskinesia (**June Marthin, Copenhagen**)

13:45 Technical and practical issues for tidal breathing measurements of nasal nitric oxide in infants and children (**Nicole Beydon, Paris**)

14:00 **Discussion** (further standardization, potential collaborative project?)

14:40 Coffee break (30 minutes)

Transatlantic discussion on diagnostic evaluation (decision-tree) for PCD – agreements, differences, and the way forward

Chairs: Claudia Kuehni (Bern) and Tom Ferkol (St. Louis)

15:10 The North American (ATS) approach: why we chose what we chose (**Adam Shapiro, Montreal**)

15:20 The European (ERS) approach: why we chose what we chose (**Jane Lucas, Southampton**)

15:30 Discussion

Short presentations

Chairs: Bruna Rubbo (Southampton) and Myrona Goutaki (Bern)

16:00 Ependymal cilia beating induces cytoskeleton rearrangement to protect the cilia base against shear stress (**Nathalie Delgehyr, Paris**)

16:20 Primary ciliary dyskinesia in adults with bronchiectasis: Data from the EMBARC registry (**James Chalmers/ Amelia Shoemark, Dundee**)

16:40 The nation-wide survey of primary ciliary dyskinesia in Japan (**N. Hashimoto, Nagoya**)

16:50 Clinical features & management of 334 children with primary ciliary dyskinesia in the English National Cohort (**Woolf Walker, Southampton**)

Discussion of plans for new COST application

Chairs: Jane Lucas (Southampton) and Claudia Kuehni (Bern)

17:00 Plans and outline of new application: **Heymut Omran, Münster (20 minutes)**

17:20 Discussion

18:00 **Closure of the meeting**

19:30 Informal dinner: After the meeting there is an informal dinner



ERS CONGRESS (15th—19th SEPTEMBER 2018)

The 2018 European Respiratory Society (ERS) International Congress will take place in Paris, France between 15th and 19th of September. During the congress, participants will have the opportunity to attend several PCD-related activities. Activities range from poster discussions to oral presentations and also include a symposium dedicated to PCD. The symposium titled “Recent advances in Primary Ciliary Dyskinesia” will take place on Tuesday 18/09 at 10:45 and will bring together experienced clinicians and researchers that will provide updates on PCD genetics (Heymut Omran, University of Munster), PCD diagnosis (Eric Haarman, VU University), clinical characteristics (Claudia Kuehni, University of Bern) and PCD management (Pierre Burgel, Paris Descartes University). Furthermore, during a second symposium titled “State of the art session: Paediatric Respiratory diseases” (Wednesday, 19/09 at 08:30) Jane Lucas (University of Southampton) will provide an overview of the state of the art in PCD.

Lastly, a total of 34 PCD-related poster and oral presentations will also take place throughout the congress duration. Additional information can be found in the detailed schedule available below (pages 4 and 5 of this edition).

this edition).

			Poster discussion	Thematic poster	Oral presentation				Symposium				
Day	Session	Title	Author (country)	8.30	10.30	10.45	12.45	12.50	13.15	14.40	14.45	16.45	
Sunday	Poster discussion: The global impact of bronchiectasis and nontuberculous mycobacteria (NTM)	AMPK is inhibited in severe bronchiectasis and may relate to reduced ciliary beat frequency	Y. Giam (Dundee, UK)										
		Neutrophil elastase increases ciliary beat frequency ex-vivo: implications for the bronchiectasis airway	M. Contarini (Gorgonzola, Italy)										
		Primary ciliary dyskinesia in adults with bronchiectasis: Data from the Embarc registry	A. Shoemark (Dundee, UK)										
	Poster discussion: Assessing pathophysiology in children	Functional dead space in children with cystic fibrosis or primary ciliary dyskinesia.	F. Singer (Bern, Switzerland)										
		Natural variability of lung clearance index and FEV1 in children and young adults with Primary Ciliary Dyskinesia	F. Singer (Bern, Switzerland)										
	Poster discussion: Orphan diseases in children	Ciliary beating patterns in PCD patients of Greek origin: a four-year experience	G. Chatziparasidis (Trikala, Greece)										
	Thematic poster: Management of bronchiectasis and large airway diseases	Diagnostic contribution of PICADAR score in etiological evaluation of patients with bronchiectasis of unknown etiology	E. Çakir (Istanbul, Turkey)										
		Prophylactic azithromycin in patients with primary ciliary dyskinesia.	A. Massiot (Levallois Perret, France)										
	Thematic poster: Role of omics in population studies	Mutations of BRWD1 may be associated with primary ciliary dyskinesia	T. Guo (Chang Sha, China)										
	Thematic poster Chronic respiratory disease : determinants of physical activity in patients	Continence Assessment in Paediatric Patients with Primary Ciliary Dyskinesia	A. Harris (Southampton, UK)										
	Thematic poster: Chronic respiratory disease: effects of rehabilitation interventions in patients	Quality of life and exercise capacity in patients with primary ciliary dyskinesia	M. Firat (Ankara, Turkey)										
	Thematic poster: Cystic fibrosis in adults: current research	Assessment of mental health status in primary ciliary dyskinesia and cystic fibrosis patients and their parents	S. Graziano (Rome, Italy)										
	Thematic poster: The bad and the ugly in paediatric asthma: comorbidities and exacerbations	Functional status of ciliated epithelium during asthma exacerbation and remission in children	I. Ozerskaia (Moscow, Russia)										
	Thematic poster: Clinical problem in paediatric pulmonology	Psychometric validation of QOL-PCD, adolescent, child and parent proxy questionnaires assessed their reliability and validity	L. Behan (Cork, Ireland)										



Day	Session	Title	Author (country)	8.30	10.30	10.45	12.45	12.50	13.15	14.40	14.45	16.45
Monday	Oral presentation: A fresh view on airway differentiation and regeneration	Functional analysis of PIERCE1, in motile ciliogenesis	P. Anujan (Sheffield, UK)									
		LSC - 2018 - miR449 protects airway cilia and healthy lung ageing	M. Lizé (Göttingen, Germany)									
		From Dictyostelium to human airway epithelium: adenine nucleotide translocase enhances cellular respiration and ciliary function disrupted by cigarette smoke	J. Nguyen (Baltimore, USA)									
		Differentiation of Induced Pluripotent Stem Cells (iPSC) into human bronchial epithelium from healthy and severe COPD patients.	E. Ahmed (Montpellier, France)									
	Oral presentation: New methods for diagnosing and treating lung diseases	To the diagnosis of primary ciliary dyskinesia without situs inversus	V. Kobylansky (Moscow, Russia)									
	Poster discussion: Latest developments in cystic fibrosis	Plasma ceramides and sphingomyelins increase in primary ciliary dyskinesia but decrease in cystic fibrosis	D. Bal Topcu (Ankara, Turkey)									
Day	Session	Title	Author (country)	8.30	10.30	10.45	12.45	12.50	13.15	14.40	14.45	16.45
	Symposium: Recent advances in primary ciliary dyskinesia	Genetics of PCD	H. Omran (Münster, Germany)									
		Diagnosis of PCD	E. Haarman (Amsterdam, Netherlands)									
		Clinical characteristics of PCD	C. Kuehni (Bern, Switzerland)									
		Management of PCD in young people and adults	P. Burgel (Sorbonne Paris Cité, France)									
	Thematic poster: Airway disease: recent discoveries	Primary ciliary dyskinesia diagnosis in adult patients with bronchiectasis in Sao Paulo, Brazil.	M. Kowal Olm (São Paulo, Brazil)									
		The nation-wide survey of primary ciliary dyskinesia in Japan	N. Hashimoto (Nagoya, Japan)									
		Motile cilia structure and function in patients with mutations in the outer dynein arm heavy chain DNAH9	A. Shoemark (Dundee, UK)									
		Does nasal rinsing reduce the need for repeat nasal brushing biopsies in a primary ciliary dyskinesia (PCD) diagnostic service? A pilot study.	A. Harris (Southampton, UK)									
Tuesday	Thematic poster: Insights in primary ciliary dyskinesia, asthma and lung function testing	Comparison of diagnostic accuracy between the chemiluminescence and electrochemical methods for nasal nitric oxide measurements in PCD	P. Kouis (Nicosia, Cyprus)									
		Characterising nutrition status in a regional cohort of children with primary ciliary dyskinesia	W. Walker (Southampton UK)									
		Comparison of conventional chest physiotherapy and vibratory positive pressure treatment system method in diagnosis of primary ciliary dyskinesia	E. Atag (Istanbul, Turkey)									
		The effect of conventional chest physiotherapy and vibratory positive pressure therapy system on lung clearance index and spirometry in patients with primary ciliary dyskinesia	E. Atag (Istanbul, Turkey)									
	Poster discussion: Primary ciliary dyskinesia and lung function tests in respiratory morbidities	Evaluation of quality of life in patients with primary ciliary dyskinesia	P. Ergenekon (Istanbul, Turkey)									
		Neonatal manifestations in Primary Ciliary Dyskinesia: a multinational cohort study	M. Goutaki (Bern, Switzerland)									
		Accuracy of high-speed video microscopy to diagnose primary ciliary dyskinesia	B. Rubbo (Southampton, UK)									
		Prevalence and Impact of Lung Resection in Primary Ciliary Dyskinesia: a cohort and nested case-control study	P. Viallourous (Nicosia, Cyprus)									
		Clinical features & management of 334 children with primary ciliary dyskinesia in the English National Cohort	W. Walker (Southampton, UK)									
		Associations between PCD-specific health-related quality of life (QOL-PCD) outcomes and clinical measures	H. Kobbernagel (Copenhagen, Denmark)									
Wednesday	Symposium: State of the art session: Paediatric respiratory diseases	Primary ciliary dyskinesia	J. Lucas (Southampton, UK)									



WE NEED YOUR CONTRIBUTION!

We would like to invite early-career researchers (ECRs) and senior researchers to send us material that could be of interest to the wider PCD community. These could include recent publications, meetings, achievements, local events, news items or photos that could be featured in the next newsletter.

ECRs in particular are encouraged to get in touch as we are working to further develop the BEAT-PCD ECR network. Among others, there will be opportunities to contribute to the next proceedings manuscript, chair sessions and organise conferences.

Last but not least, we would like to thank ECR **Agatha Wisse** for collaborating with the team to develop this edition of the BEAT-PCD newsletter. Agatha wrote the section on the ERS PCD Meeting and provided the preliminary agenda.

Please do not hesitate to get in touch with us at:

b.rubbo@soton.ac.uk and **myrofora.goutaki@ispm.unibe.ch**

We will be very happy to hear from you.

BEAT-PCD Newsletter team

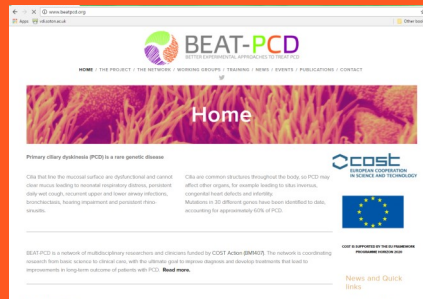
Bruna Rubbo, Myrona Goutaki, Ana Reula, Panayiotis Kouis, Maciej Dabrowski, Nisreen Rumman, Florian Halbeisen and Regan Doherty

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