



## EDITORIALS

- 1597 Bronchiectasthma and asthmectomy!**  
Joan B. Sorianó and José Serrano
- 1601 Adding biological markers to COPD categorisation schemes: a way towards more personalised care?**  
Nicolas Roche
- 1606 COPD treatment: about collateral channels and collapsing airways**  
Ralf-Harto Hubner and Dominik Herzog
- 1611 When insulin has to work hard to keep the sugar at bay the upper airway collapses away**  
Danny J. Eckert and Arie Oliven
- 1615 Sildenafil for pulmonary hypertension complicating idiopathic pulmonary fibrosis: a rationale grounded in basic science**  
Paul M. Hassoun and Steven D. Nathan
- 1618 How to follow-up patients with hereditary haemorrhagic telangiectasia and suspected pulmonary arteriovenous malformations**  
Thierry Chinet
- 1622 Interstitial pneumonitis with autoimmune features (IPAF): a work in progress**  
Fabrizio Luppi and Athol U. Wells
- 1625 Beryllium disease and sarcoidosis: still besties after all these years?**  
Daniel A. Culver
- 1629 Genetic studies yield clues to the pathogenesis of Langerhans cell histiocytosis**  
Li Zhang and Joel Moss
- 1632 A wider pathological network underlying breathlessness and respiratory failure in amyotrophic lateral sclerosis**  
Kyle T.S. Pattinson and Martin R. Turner

## ORIGINAL ARTICLES

### COPD

- 1635 Prognostic assessment in COPD without lung function: the B-AE-D indices**  
Lucas Boeck, Joan B. Sorianó, Marjolein Brusse-Keizer, Francesco Blasi, Konstantinos Kostikas, Wim Boersma, Branislava Milenkovic, Renaud Louïs, Alicia Lacoma, Remco Djamin, Joachim Aerts, Antoni Torres, Gernot Rohde, Tobias Welte, Pablo Martinez-Camblor, Janko Rakic, Andreas Scherr, Michael Koller, Job van der Palen, Jose M. Marin, Inmaculada Alfageme, Pere Almagro, Ciro Casanova, Cristobal Esteban, Juan J. Soler-Cataluña, Juan P. de-Torres, Marc Miravitles, Bartolome R. Celli, Michael Tamm and Daiana Stolz
- 1645 Linoleic acid-derived lipid mediators increase in a female-dominated subphenotype of COPD**  
David Balgoma, Mingxing Yang, Marcus Sjödin, Stuart Snowden, Reza Karimi, Bettina Levänen, Heta Merikallio, Riitta Kaarteenaho, Lena Palmberg, Kjell Larsson, David J. Erle, Sven-Erik Dahlén, Barbro Dahlén, C. Magnus Sköld, Åsa M. Wheelock and Craig E. Wheelock

- 1657 Collapse phenomenon during Chartis collateral ventilation assessment**  
Wolfgang Gesierich, Konstantinos Samitas, Frank Reichenberger and Juergen Behr

- 1668 Real-life assessment of the multidimensional nature of dyspnoea in COPD outpatients**  
Capucine Morelot-Panzini, Hélène Gilet, Bernard Aguilaniu, Philippe Devillier, Alain Didier, Thierry Perez, Christophe Pignier, Benoit Arnould and Thomas Similowski

### Asthma and bronchiectasis

- 1680 Asthma and bronchiectasis exacerbation**  
Bei Mao, Jia-Wei Yang, Hai-Wen Lu and Jin-Fu Xu

### Paediatric pulmonology

- 1687 Tidal flow variability measured by impedance pneumography relates to childhood asthma risk**  
Ville-Pekka Seppä, Anna S. Pelkonen, Anne Kotaniemi-Syrjänen, Jari Viik, Mika J. Mäkelä and L. Pekka Malmberg

### Cystic fibrosis

- 1697 Forecasting US ivacaftor outcomes and cost in cystic fibrosis patients with G551D mutation**  
Piyameth Dilokthornsakul, Ryan N. Hansen and Jonathan D. Campbell

- 1706 Multicentre chest computed tomography standardisation in children and adolescents with cystic fibrosis: the way forward**  
Wieying Kuo, Mariette P.C. Kemner-van de Corput, Adria Perez-Rovira, Marleen de Bruijne, Isabelle Fajac, Harm A.W.M. Tiddens and Marcel van Straten on behalf of the ECFS-CTN/SCIFI CF study group

### Sleep

- 1718 Pharyngeal collapsibility during sleep is elevated in insulin-resistant females with morbid obesity**  
Oscar L. Llanos, Panagis Galitsatos, Edmarie Guzmán-Vélez, Susheel P. Patil, Philip L. Smith, Thomas Magnusson, Michael Schweitzer, Kimberley Steele, Vsevolod Y. Polotsky and Alan R. Schwartz

## ORIGINAL ARTICLES (CONTINUED)

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### Pulmonary vascular diseases

#### 1727 Initial dual oral combination therapy in pulmonary arterial hypertension

Olivier Sitbon, Caroline Sattler, Laurent Bertoletti, Laurent Savale, Vincent Cottin, Xavier Jaïs, Pascal De Groote, Ari Chaouat, Céline Chabannes, Emmanuel Bergot, Hélène Bouvaist, Claire Dauphin, Arnaud Bourdin, Fabrice Bauer, David Montani, Marc Humbert and Gérald Simonneau

#### 1737 Vascular effects of sildenafil in patients with pulmonary fibrosis and pulmonary hypertension: an ex vivo/in vitro study

Javier Milara, Juan Escrivá, José Luis Ortiz, Gustavo Juan, Enrique Artigues, Esteban Morcillo and Julio Cortijo

#### 1750 Follow-up of pulmonary right-to-left shunt in hereditary haemorrhagic telangiectasia

Veronique M.M. Vorselaars, Sebastiaan Velthuis, Repke J. Snijder, Cornelis J.J. Westermann, Jan A. Vos, Johannes J. Mager and Martijn C. Post

### Tuberculosis

#### 1758 Comparison of effectiveness and safety of imipenem/clavulanate- versus meropenem/clavulanate-containing regimens in the treatment of MDR- and XDR-TB

Simon Tiberi, Giovanni Sotgiu, Lia D'Ambrosio, Rosella Centis, Marcos Abdo Arbex, Edith Alarcon Arrascue, Jan Willem Alffenaar, Jose A. Caminero, Mina Gaga, Gina Gualano, Alena Skrahina, Ivan Solovic, Giorgia Sulis, Marina Tadolini, Valentina Alarcon Guizado, Saverio De Lorenzo, Aurora Jazmín Roby Arias, Anna Scardigli, Onno W. Akkerman, Alena Aleksa, Janina Artsukevich, Vera Auchynka, Eduardo Henrique Bonini, Félix Antonio Chong Marin, Lorena Collahuazo López, Gerard de Vries, Simone Dore, Heinke Kunst, Alberto Matteelli, Charalampos Moschos, Fabrizio Palmieri, Apostolos Papavasileiou, Marie-Christine Payen, Andrea Piana, Antonio Spanevello, Dante Vargas Vasquez, Pietro Viggiani, Veronica White, Alimuddin Zumla and Giovanni Battista Migliori

### Interstitial lung diseases

#### 1767 Characterisation of patients with interstitial pneumonia with autoimmune features

Justin M. Oldham, Ayodeji Adegunsoye, Eleanor Valenzi, Cathryn Lee, Leah Witt, Lena Chen, Aliya N. Husain, Steven Montner, Jonathan H. Chung, Vincent Cottin, Aryeh Fischer, Imre Noth, Rekha Vij and Mary E. Strek

#### 1776 Unfavourable effects of medically indicated oral anticoagulants on survival in idiopathic pulmonary fibrosis

Michael Kreuter, Marlies S. Wijsenbeek, Martina Vasakova, Paolo Spagnolo, Martin Kolb, Ulrich Costabel, Derek Weycker, Klaus-Uwe Kirchgaessner and Toby M. Maher

### Orphan lung disease

#### 1785 Recurrent NRAS mutations in pulmonary Langerhans cell histiocytosis

Samia Mourah, Alexandre How-Kit, Véronique Meignin, Dominique Gossot, Gwenaël Lorillon, Emmanuelle Bugnet, Florence Mauger, Celeste Lebbe, Sylvie Chevret, Jörg Tost and Abdellatif Tazi

### Beryllium-induced lung disease

#### 1797 Beryllium-induced lung disease exhibits expression profiles similar to sarcoidosis

Li Li, Lori J. Silveira, Nabeel Hamzeh, May Gillespie, Peggy M. Mroz, Annyce S. Mayer, Tasha E. Fingerlin and Lisa A. Maier

### Lung cancer

#### 1809 In-hospital mortality following lung cancer resection: nationwide administrative database

Pierre-Benoit Pagès, Jonathan Cottenet, Anne-Sophie Mariet, Alain Bernard and Catherine Quantin

### Dyspnoea and amyotrophic lateral sclerosis

#### 1818 Cortical drive to breathe in amyotrophic lateral sclerosis: a dyspnoea-worsening defence?

Marjolaine Georges, Elise Moravieic, Mathieu Raux, Jésus Gonzalez-Bermejo, Pierre-François Pradat, Thomas Similowski and Capucine Morélot-Panzini

## REVIEW

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#### 1829 Diffuse idiopathic pulmonary neuroendocrine cell hyperplasia syndrome

Giulio Rossi, Alberto Cavazza, Paolo Spagnolo, Nicola Sverzellati, Lucia Longo, Agita Jukna, Gloria Montanari, Cristiano Carbonelli, Giada Vincenzi, Giuseppe Bogina, Renato Franco, Marcello Tiseo, Vincent Cottin and Thomas V. Colby

## BACK TO BASICS

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#### 1842 "Scar-cinema": viewing the fibrotic lung mesenchymal cell in the context of cancer biology

Jeffrey C. Horowitz, John J. Osterholzer, Antonia Marazioti and Georgios T. Stathopoulos

## AGORA

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### Research letters

- 1855 Airflow obstruction is associated with increased smooth muscle extracellular matrix**  
Robyn L. Jones, Peter B. Noble, John G. Elliot, Howard W. Mitchell, Peter K. McFawn, James C. Hogg and Alan L. James
- 1857 An outreach model of tertiary difficult asthma care reduces adverse asthma outcomes and healthcare utilisation costs**  
Veeresh K. Patil, Christine Townshend, Frances Mitchell and Ramesh J. Kurukulaaratchy
- 1861 Use of the modified Borg scale and numerical rating scale to measure chronic breathlessness: a pooled data analysis**  
Miriam J. Johnson, Lucy Close, Suzie C. Gillon, Alex Molassiotis, Paul H. Lee and Morag C. Farquhar on behalf of the Breathlessness Research Interest Group (BRIG)
- 1864 Asthmatics with high levels of serum surfactant protein D have more severe disease**  
Alida Benfante, Salvatore Battaglia, Stefania Principe, Chiara Di Mitri, Alessandra Paternò, Mario Spatafora and Nicola Scichilone
- 1867 Incorporating therapeutic drug monitoring into the World Health Organization hierarchy of tuberculosis diagnostics**  
Samiksha Ghimire, Matthieu S. Bolhuis, Marieke G.G. Sturkenboom, Onno W. Akkerman, Wiel C.M. de Lange, Tjip S. van der Werf and Jan-Willem C. Alffenaar
- 1870 Low yield of screening asylum seekers from countries with a tuberculosis incidence of <50 per 100 000 population**  
Gerard de Vries, Job van Rest, Wieneke Meijer, Bert Wolters and Rob van Hest
- 1873 A 380-gene meta-signature of active tuberculosis compared with healthy controls**  
Simon Blankley, Christine M. Graham, Joe Levin, Jacob Turner, Matthew P.R. Berry, Chloe I. Bloom, Zhaohui Xu, Virginia Pascual, Jacques Banchereau, Damien Chaussabel, Ronan Breen, George Santis, Derek M. Blankenship, Marc Lipman and Anne O'Garra
- 1876 Implementation of digital technology solutions for a lung health trial in rural Malawi**  
Blessings Chisunkha, Hastings Banda, Rachael Thomson, S. Bertel Squire and Kevin Mortimer
- 1879 Somatic DICER1 mutations in adult-onset pulmonary blastoma**  
Leanne de Kock, Ismaël Bah, Joan Brunet, Harriet Druker, Itziar Astigarraga, Joaquim Bosch-Barrera, Dorothée Bouron-Dal Soglio, Van-Hung Nguyen, David Malkin, John R. Priest and William D. Foulkes
- 1883 Should lymphangioleiomyomatosis patients be permitted to donate blood?**  
Anne F. Eder, Jay H. Ryu and Joel Moss
- Correspondence**
- 1885 Risk of COPD in smokers with low transfer factor**  
Martin R. Miller and Vito Brusasco; Philip H. Quanjer; Ben-Gary Harvey, Yael Strulovici-Barel, Robert J. Kaner, Abraham Sanders, Thomas L. Vincent, Jason G. Mezey and Ronald G. Crystal
- 1890 Sensitivity to chemotherapy/tyrosine kinase inhibitors of mucinous lepidic adenocarcinoma should be tested in a phase III trial?**  
Jacques Cadanel, Marie Wislez, Alexandra Langlais, Franck Morin and Gérard Zalcman
- 1892 AUTHOR CORRECTIONS**