




EARCO Registry Update

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European Respiratory Society statement: diagnosis and treatment of pulmonary disease in α_1 -antitrypsin deficiency

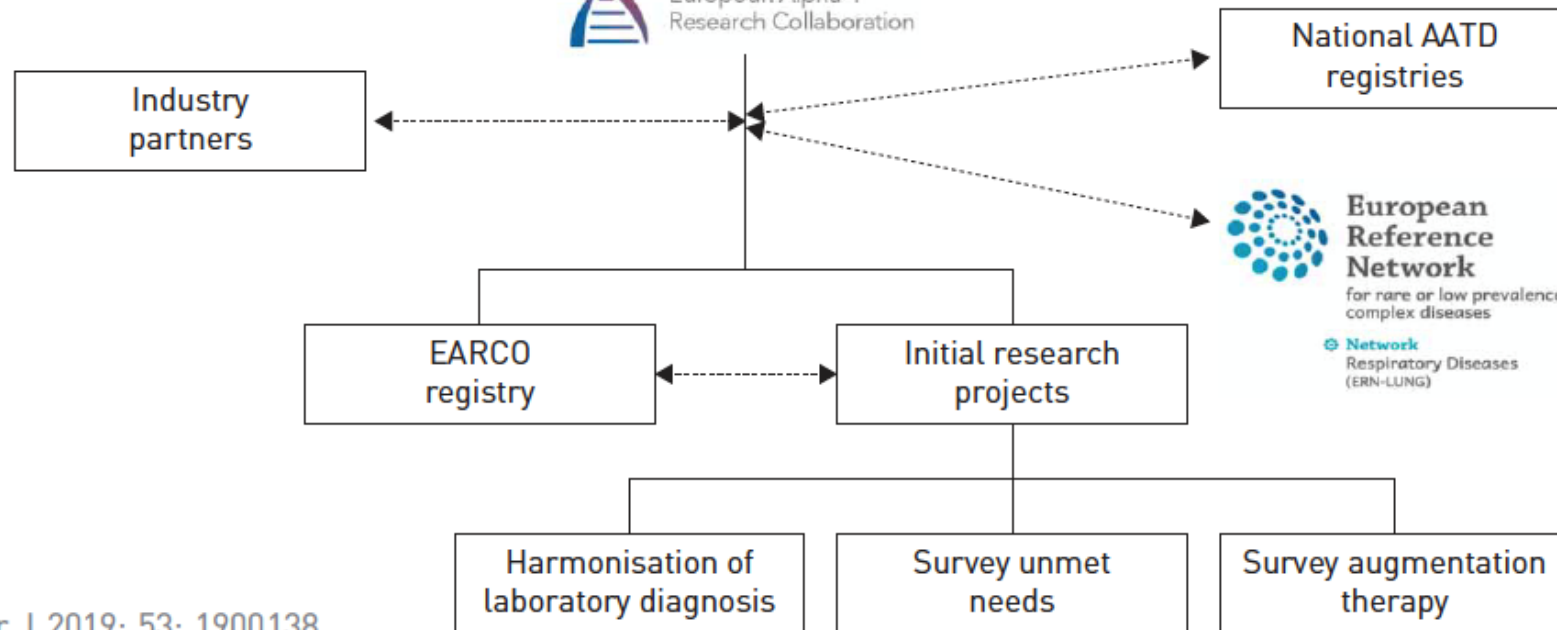
Marc Miravittles  (co-chair)¹, Asger Dirksen², Ilaria Ferrarotti³, Vladimir Koblizek⁴, Peter Lange⁵, Ravi Mahadeva⁶, Noel G. McElvaney⁷, David Parr⁸, Eeva Piitulainen⁹, Nicolas Roche¹⁰, Jan Stolk¹¹, Gabriel Thabut^{12,13}, Alice Turner¹⁴, Claus Vogelmeier¹⁵ and Robert A. Stockley (co-chair)¹⁶

Statement

- According to the European Council, management of patients with AATD should be supervised by reference centres of excellence at a national or regional level.
- The systematic collection of data concerning clinical characteristics and natural history of patients with AATD in national and international registries will enhance knowledge about the evolution of this disease and its optimal management.
- For many AATD individuals a respiratory service is the first point of diagnosis. The operational pathway includes varying assessments and follow-up depending on personalising the patients' risk and defining the respiratory phenotype. Links to multidisciplinary teams will ensure the best quality of care.

The European Alpha-1 Research Collaboration (EARCO): a new ERS Clinical Research Collaboration to promote research in alpha-1 antitrypsin deficiency

Marc Miravittles ¹, Joanna Chorostowska-Wynimko ², Ilaria Ferrarotti ³, Noel G. McElvaney ⁴, Karen O'Hara ^{5,6}, Jan Stolk ⁷, Robert A. Stockley ⁸, Alice Turner ⁹, Marion Wilkens ^{6,10} and Timm Greulich ¹¹ on behalf of the EARCO Clinical Research Collaboration ¹²





**Frankfurt
EARCO 2018**





Research priorities in α_1 -antitrypsin deficiency: results of a patients' and healthcare providers' international survey from the EARCO Clinical Research Collaboration






Miriam Barrecheguren¹, Karen O'Hara², Marion Wilkens³, Jeanette Boyd⁴, Ewa Kolda⁵, Beatriz Lara ⁶, Joanna Chorostowska-Wynimko⁷, Ilaria Ferrarotti ⁸, Jan Chlumský⁹, Christian Clarenbach ¹⁰, Timm Greulich ¹¹, Marc Miravittles ¹ and Maria Sucena¹², on behalf of EARCO

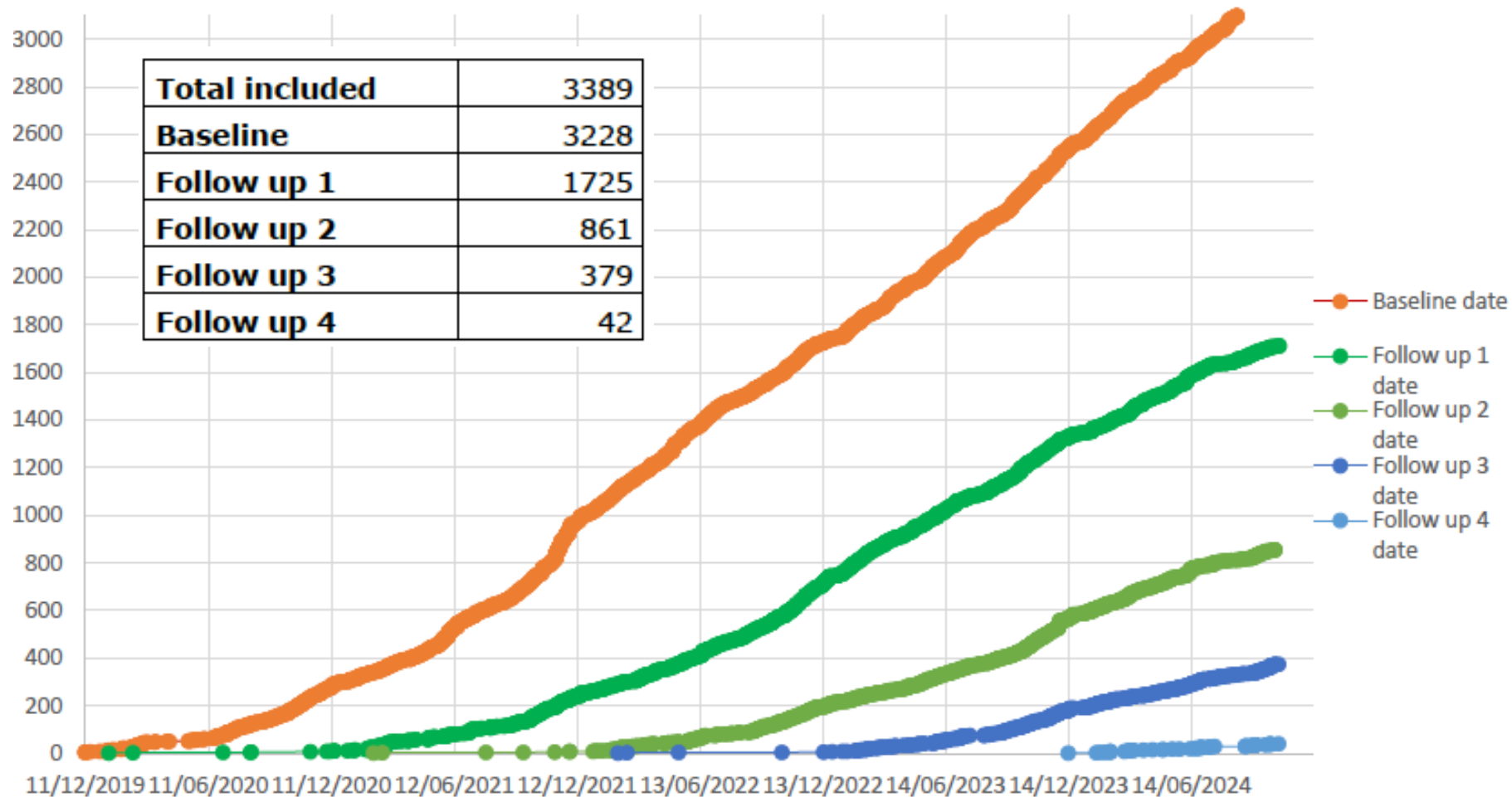
TABLE 4 Most important research areas as rated by patients and caregivers

Research area	Total	Non-reimbursed countries	Reimbursed countries
Improving knowledge of AATD, in particular among general practitioners	99	99	99
Access to AATD specialised centres	97	99	96
Access to reliable, easy to understand information about living with AATD	97	99	96
Being able to recognise an exacerbation	97	98	97
Targeted screening programmes: COPD and asthma patients	96	94	99
Having an action plan for exacerbations and easy access to healthcare during episodes	96	99	95
A personalised integrated care plan including therapeutic physical activity	95	91	98
Education for physicians on diagnostic techniques, algorithm, interpretation of results	95	95	97

Recruitment

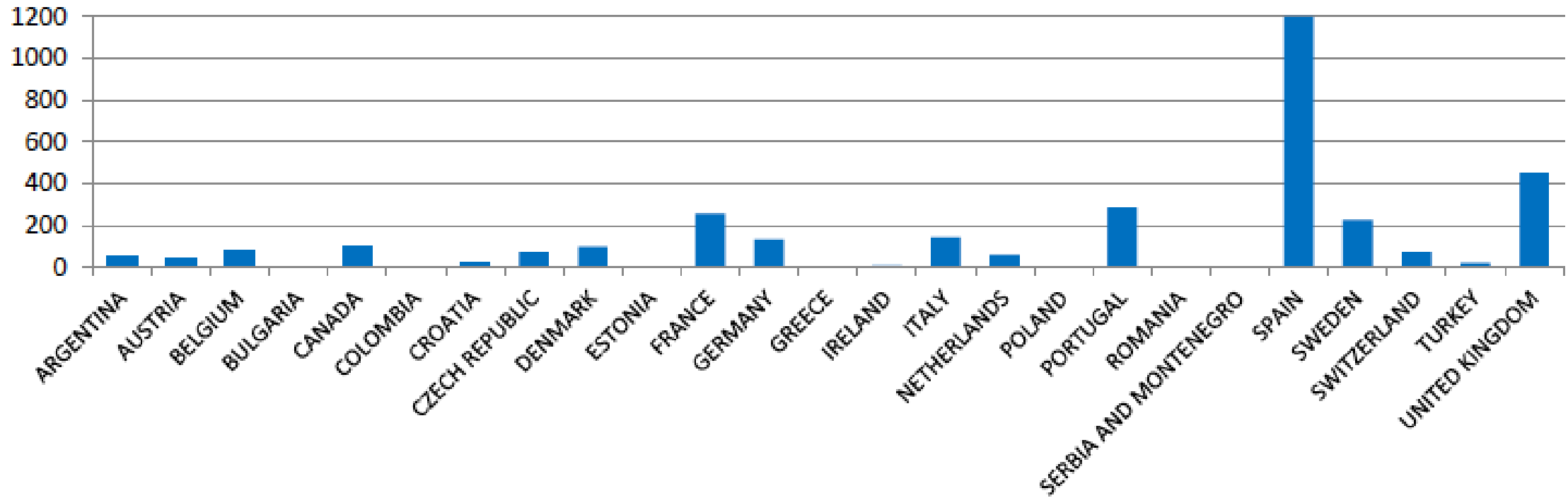
Recruitment summary:

Patient included: 3389



Recruitment

Number of patients included by country





About EARCO

EARCO stands for European Alpha-1 Research Collaboration. It is a pan-European network committed to promoting clinical research and education in alpha-1 antitrypsin deficiency (AATD).

[See more](#) ↗



Hospitals

Site	Location	Investigator	Email
ARGENTINA			
Hospital Dr Antonio Cetrángolo	Buenos Aires	Mariano Fernández Acquier	acquier@hotmail.com
Centro Médico Capital	La Plata	Andrés L. Echazarreta	aechaza@gmail.com
AUSTRIA			
Karl-Landsteiner Research Institute at Klinik Ottakring	Vienna	Karin Schmid-Scherzer Funk Georg-Christian	karinschmid.197@gmx.at georg-christian.funk@gesundheitsverbund.at
Medizinische Universität Wien	Vienna	Marco Idzko	marco.idzko@meduniwien.ac.at
BELGIUM			
Universitair Ziekenhuis Leuven	Leuven	Wim Janssens	wim.janssens@uzleuven.be
Hôpital Erasme - ULB	Bruxelles	Sylvia Perez-Bogerd	silvia.perez.bogerd@erasme.ulb.ac.be

Country	Number of open sites
ARGENTINA	2
AUSTRIA	1
BELGIUM	3
BULGARIA	1
CANADA	1
COLOMBIA	1
CROATIA	1
CZECH REPUBLIC	1
DENMARK	1
ESTONIA	2
FRANCE	7
GERMANY	9
GREECE	1
IRELAND	1
ITALY	4
NETHERLANDS	1
POLAND	1
PORTUGAL	14
ROMANIA	1
SERBIA AND MONTENEGRO	1
SPAIN	34
SWEDEN	1
SWITZERLAND	1
TURKEY	3
UNITED KINGDOM	9

Site	Patients included
Hospital Álvaro Cunqueiro, Vigo (Spain)	308
Skåne University Hospital, Malmö (Sweden)	227
Queen Elizabeth Hospital Birmingham (United Kingdom)	222
Complejo Hospitalario Universitario de Santiago (Spain)	186
Hospital Universitari Vall d' Hebron, Barcelona (Spain)	145
Hospital Clínico San Carlos, Madrid (Spain)	110
Gentofte Hospital (Denmark)	101
IRCCS Policlinico San Matteo Foundation, Pavia (Italy)	100
Hospital Universitario Virgen del Rocío, Sevilla (Spain)	97
Inspiration Research, Toronto (Canada)	90
Coventry University Hospital (United Kingdom)	88
University Hospital Zurich (Switzerland)	85
Thomayer Hospital, Prague (Czech Republic)	84
Hospital Universitario Ntra Sra de Candelaria, Tenerife (Spain)	79
Hospital Senhora de Oliveira (Portugal)	76
Hôpital Louis Pradel - Lyon (France)	74
Hospital Universitario Central de Asturias (Spain)	72
Royal Free London NHS Foundation Trust (United Kingdom)	71
Thoraxklinik, Heidelberg (Germany)	69
Hospital Universitario de Burgos (Spain)	67
LUMC, Leiden (Netherlands)	66
Centro Hospitalar de Vila Nova de Gaia/Espinho (Portugal)	63
Centro Hospitalar e Universitario de Coimbra (Portugal)	60
University Hospital Leuven (Belgium)	59
Hospital Bichat Claude Bernard (France)	59

**101 sites
in 25 countries**

Research projects

<p>RESEARCH PROJECT EARCO Registry</p>	<p>RESEARCH PROJECT Research Priorities in alpha-1 antitrypsin deficiency: International survey from EARCO.</p>	<p>RESEARCH PROJECT Descriptive Analysis of the Expert Opinion Regarding the Initiation of Augmentation Therapy in Alpha-1-Antitrypsin-Deficiency</p>	<p>RESEARCH PROJECT European external quality control program for laboratory diagnosis of alpha1-antitrypsin deficiency</p>	<p>RESEARCH PROJECT Impact of COVID-19 infection in patients with alpha-1 antitrypsin deficiency</p>
<p>RESEARCH PROJECT Analysis of the prevalence and impact of bronchiectasis in patients with AATD</p>	<p>RESEARCH PROJECT Pathways of Care in Alpha-1 antitrypsin Deficiency</p>	<p>RESEARCH PROJECT Prognosis of co-existent lung and liver disease in alpha-1 antitrypsin deficiency</p>	<p>RESEARCH PROJECT Defining Rapid Decliners : Implications for identifying clinical benefit in AATD</p>	<p>RESEARCH PROJECT Exacerbations of AATD lung disease: impact and role of augmentation therapy</p>
<p>RESEARCH PROJECT Impact of augmentation in patients with AATD with PiSZ or with null or rare genotypes.</p>	<p>RESEARCH PROJECT Alpha-1 antitrypsin deficiency associated with the Mmalton variant – characterization and prognosis</p>	<p>RESEARCH PROJECT Alpha-1 antitrypsin related disease risk for the Pi*SS genotype</p>	<p>RESEARCH PROJECT Analysis of routine blood biomarkers of systemic inflammation in patients Pi*ZZ and Pi*SZ.</p>	<p>RESEARCH PROJECT Clinical AATD clusters from the EARCO registry</p>

28 Research projects

Research projects

RESEARCH PROJECT

Monitoring of plasma levels of two different specific fibrinopeptides of patients with AATD-associated panniculitis

RESEARCH PROJECT

Endobronchial valves in the treatment of AATD

RESEARCH PROJECT

Gender aspects in alpha-1 antitrypsin deficiency

RESEARCH PROJECT

Relationships between Z-alpha1 antitrypsin levels and routine blood biomarkers of systemic inflammation and lung function in patients Pi*ZZ

RESEARCH PROJECT

Characteristics and risk factors for lung disease in never smokers with alpha-1 antitrypsin deficiency and Pi*ZZ genotype

RESEARCH PROJECT

FLARE = Future of alpha-1 Antitrypsin deficiency = Prediction Of Mortality Or Lung Transplantation In Patients With AATD

RESEARCH PROJECT

Molecular characterization of Alpha 1-Antitrypsin deficiency-related liver cancer

RESEARCH PROJECT

Characteristics of Bronchiectasis in Alpha-1-Antitrypsin Deficiency Patients with genotype other than Pi*ZZ

RESEARCH PROJECT

Actionable Aspects Towards Disease modification – Proposal for a holistic treatment concept for patients with AATD

RESEARCH PROJECT

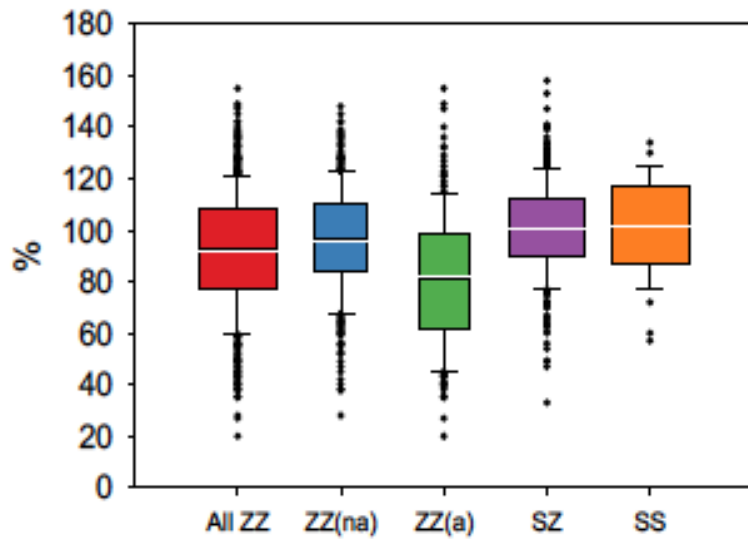
Self-reported physical activity in patients with alpha-1 antitrypsin deficiency

28 Research projects

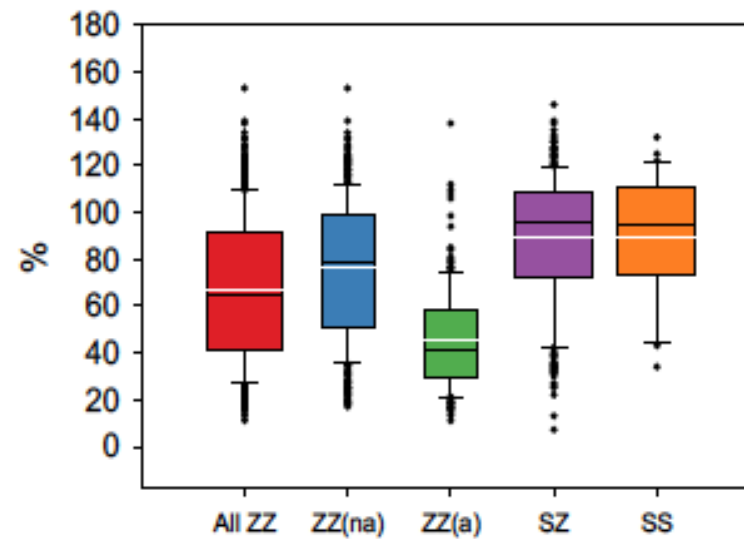


Clinical and functional characteristics of individuals with alpha-1 antitrypsin deficiency: EARCO international registry

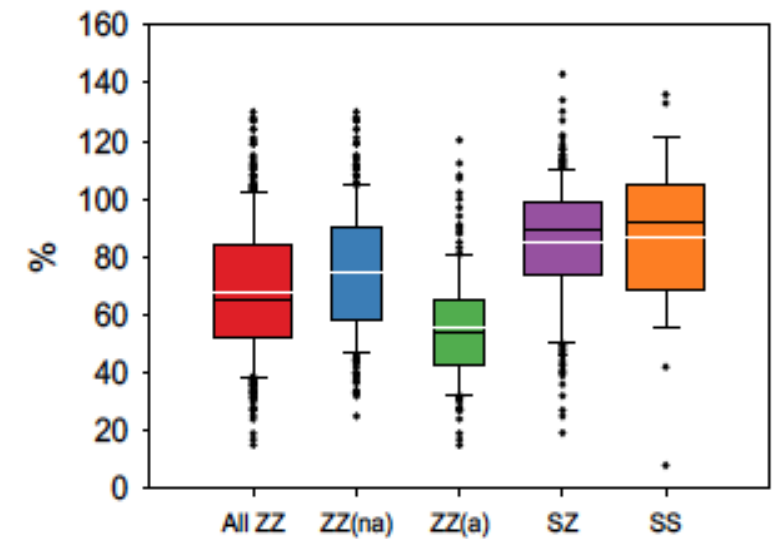
Marc Miravittles^{1,2*}, Alice M. Turner^{3,4}, María Torres-Duran⁵, Hanan Tanash⁶, Carlota Rodríguez-García⁷, José Luis López-Campos^{2,8}, Jan Chlumsky⁹, Catarina Guimaraes¹⁰, Juan Luis Rodríguez-Hermosa^{11,12}, Angelo Corsico^{13,14}, Cristina Martínez-González¹⁵, José María Hernández-Pérez¹⁶, Ana Bustamante¹⁷, David G. Parr¹⁸, Francisco Casas-Maldonado¹⁹, Ana Hecimovic^{20,21}, Wim Janssens^{22,23}, Beatriz Lara¹⁸, Miriam Barrecheguren¹, Cruz González²⁴, Jan Stolk²⁵, Cristina Esquinas¹ and Christian F. Clarenbach²⁶



FVC



FEV1

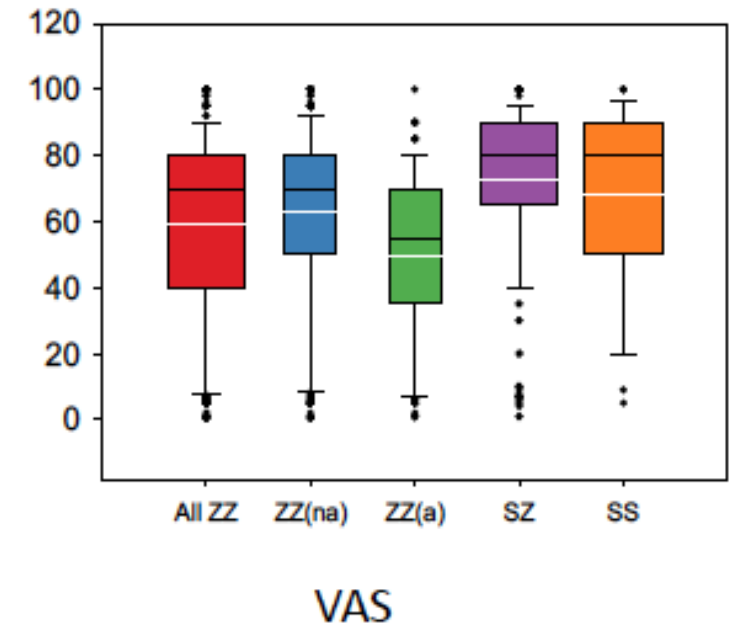
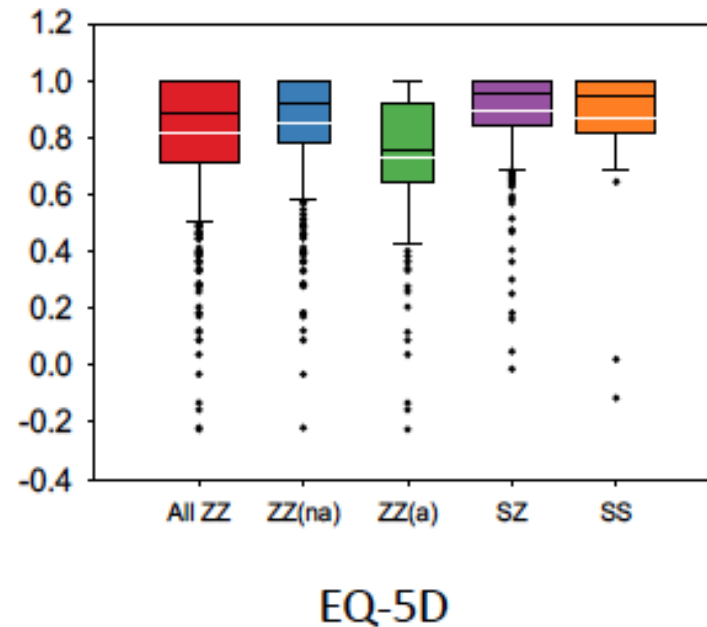
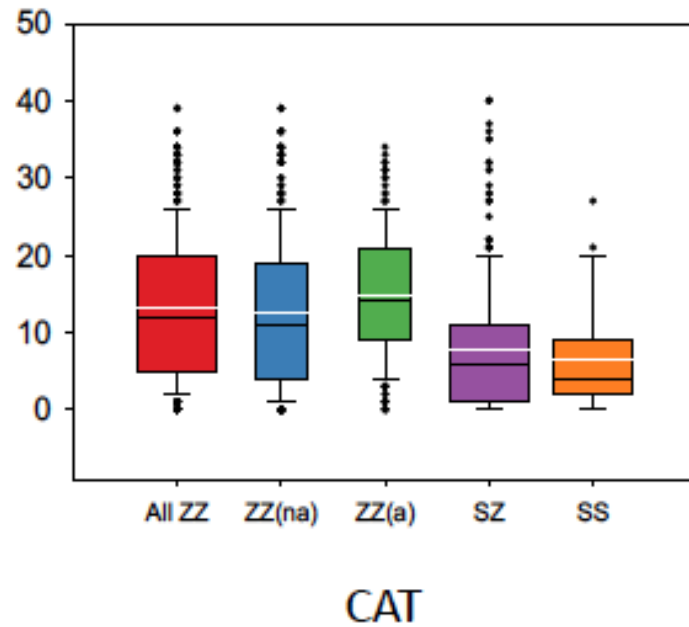


KCO



Clinical and functional characteristics of individuals with alpha-1 antitrypsin deficiency: EARCO international registry

Marc Miravittles^{1,2*}, Alice M. Turner^{3,4}, María Torres-Duran⁵, Hanan Tanash⁶, Carlota Rodríguez-García⁷, José Luis López-Campos^{2,8}, Jan Chlumsky⁹, Catarina Guimaraes¹⁰, Juan Luis Rodríguez-Hermosa^{11,12}, Angelo Corsico^{13,14}, Cristina Martínez-González¹⁵, José María Hernández-Pérez¹⁶, Ana Bustamante¹⁷, David G. Parr¹⁸, Francisco Casas-Maldonado¹⁹, Ana Hecimovic^{20,21}, Wim Janssens^{22,23}, Beatriz Lara¹⁸, Miriam Barrecheguren¹, Cruz González²⁴, Jan Stolk²⁵, Cristina Esquinas¹ and Christian F. Clarenbach²⁶





Characteristics of individuals with alpha-1 antitrypsin deficiency from Northern and Southern European countries: EARCO international registry

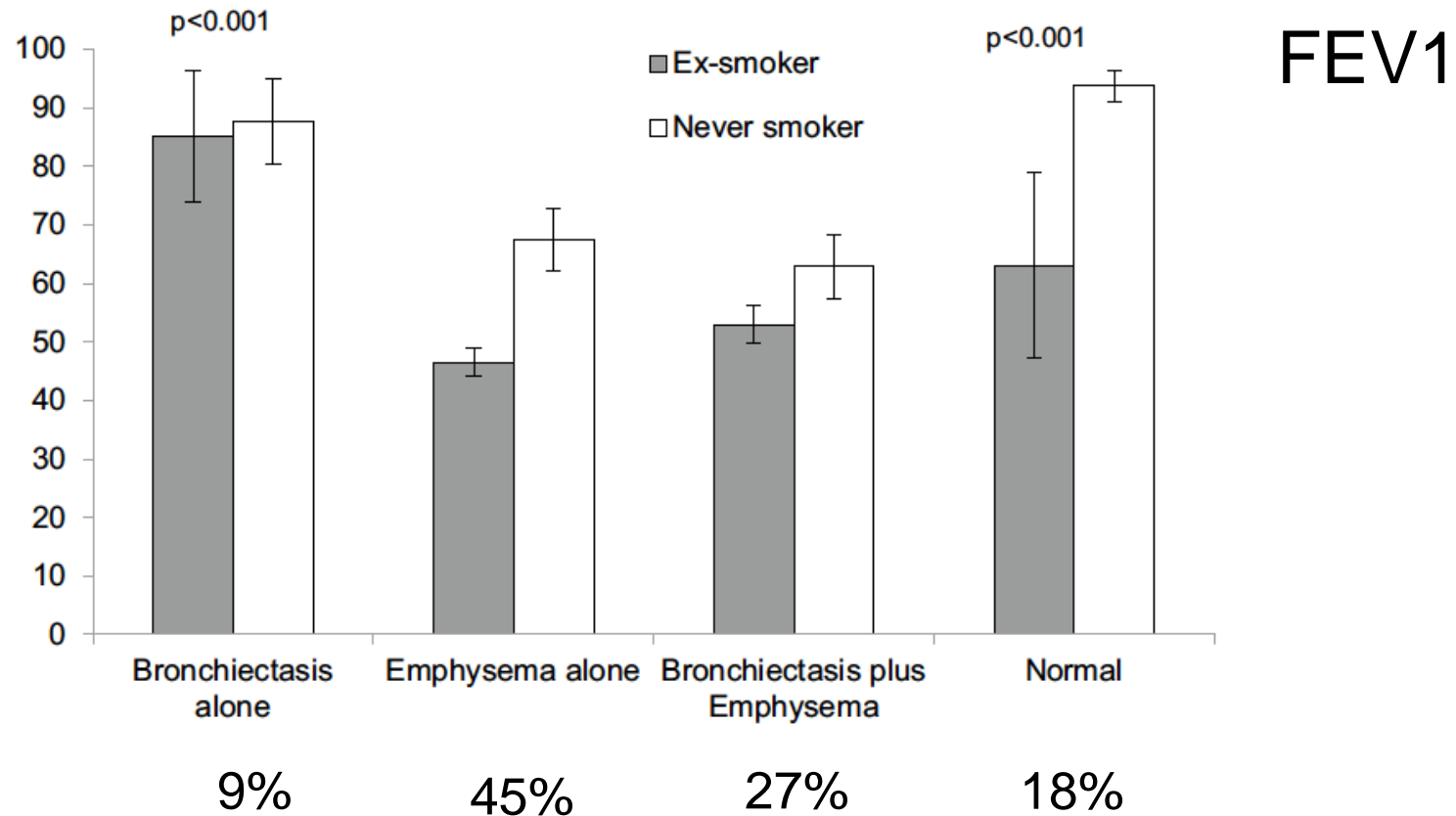
Eur Respir J 2023; 61: 2201949

	All PI*ZZ	PI*ZZ in Northern countries	PI*ZZ in Southern countries	p-value
Subjects	629	285	251	
Age, years	55.6±13.2	57.1±11.8	55.5±14.4	0.441
Male	324 (51.5)	137 (48.1)	128 (51)	0.499
BMI, kg·m ⁻²	26±4.8	26.2±4.9	26±4.8	0.955
Smoking				
Smokers	11 (1.8)	2 (0.7)	7 (2.8)	0.089
Ex-smokers	356 (56.8)	148 (51.9)	149 (59.8)	0.067
Never-smokers	260 (41.5)	135 (47.4)	93 (37.3)	0.020
Tobacco exposure, pack-years	17.8±14	14.3±12.9	20.7±14.3	<0.001
FVC, %	91.8±23.7	91.8±21.2	94.6±24.8	0.052
FEV ₁ , %	66.9±30.7	67.3±27.9	70.3±32.4	0.410
FEV ₁ /FVC	0.54±0.2	0.57±0.2	0.58±0.2	0.485
K _{CO} , %	68±23.2	71.2±23.1	68.7±22.8	0.394
CAT	13.2±9.3	16±9.2	9.7±8.5	<0.001
Serum AAT, mg·dL ⁻¹	23.8±8.3	24.3±8.7	23±8.2	0.135
Age at onset of symptoms, years	43.7±14.7	45±13.3	44.4±16.4	0.868
Age at diagnosis of AATD, years	44.7±16.7	44.1±16.9	45.4±17.2	0.280
Index case	457 (72.8)	213 (74.7)	196 (78.4)	0.319
CT scan available	529 (84.1)	231 (82.5)	214 (85.9)	0.279
Chronic respiratory disease	508 (81.4)	222 (78.4)	205 (82.7)	0.222
COPD	349 (55.9)	135 (47.7)	144 (58.1)	0.017
Emphysema	357 (57.2)	133 (47)	154 (62.1)	<0.001
Bronchiectasis	137 (22)	50 (17.7)	79 (31.9)	<0.001
Asthma	88 (14.1)	49 (17.3)	31 (12.5)	0.122
Liver disease	99 (16)	36 (12.9)	43 (17.4)	0.149
Charlson index (age corrected)	3.3 (1.9)	3.2 (1.7)	3.3 (2)	0.773
History of pneumonia	173 (29.3)	76 (29.9)	62 (25.1)	0.227
Augmentation therapy	190 (30.2)	4 (1.4)	131 (52.2)	<0.001



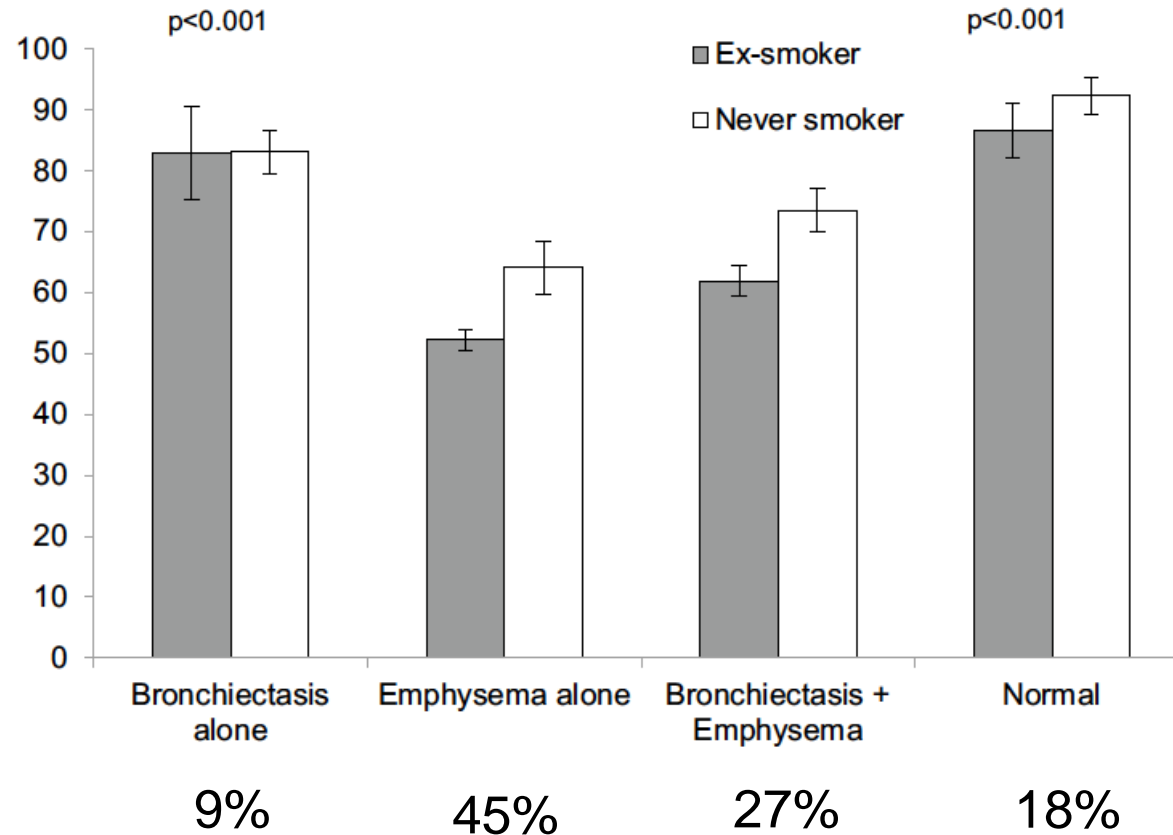
The prevalence of bronchiectasis in patients with alpha-1 antitrypsin deficiency: initial report of EARCO

Robert A. Stockley^{1*}, Anita Pye², Joshua De Soyza², Alice M. Turner² and Marc Miravittles³ on behalf of the EARCO study investigators



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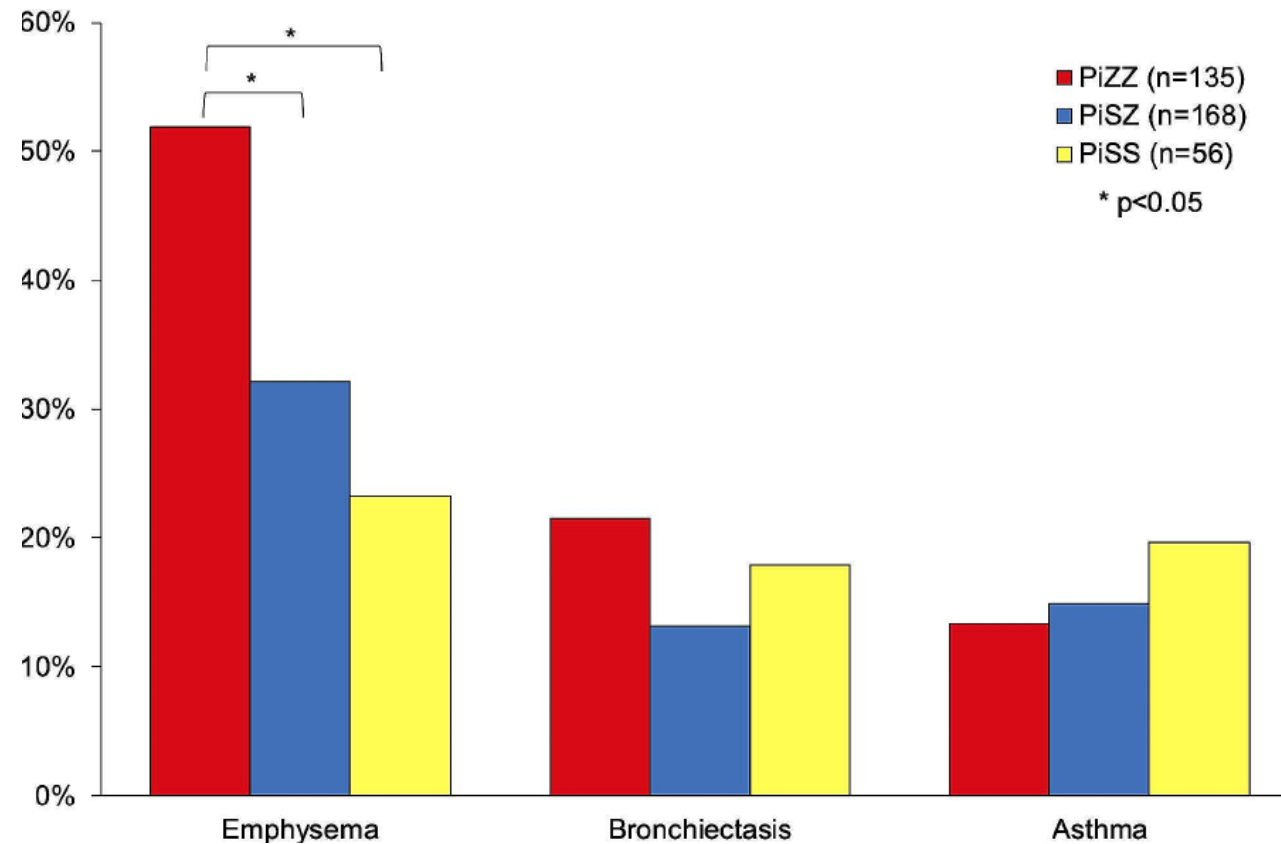


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Risk of lung disease in the PI*SS genotype of alpha-1 antitrypsin: an EARCO research project

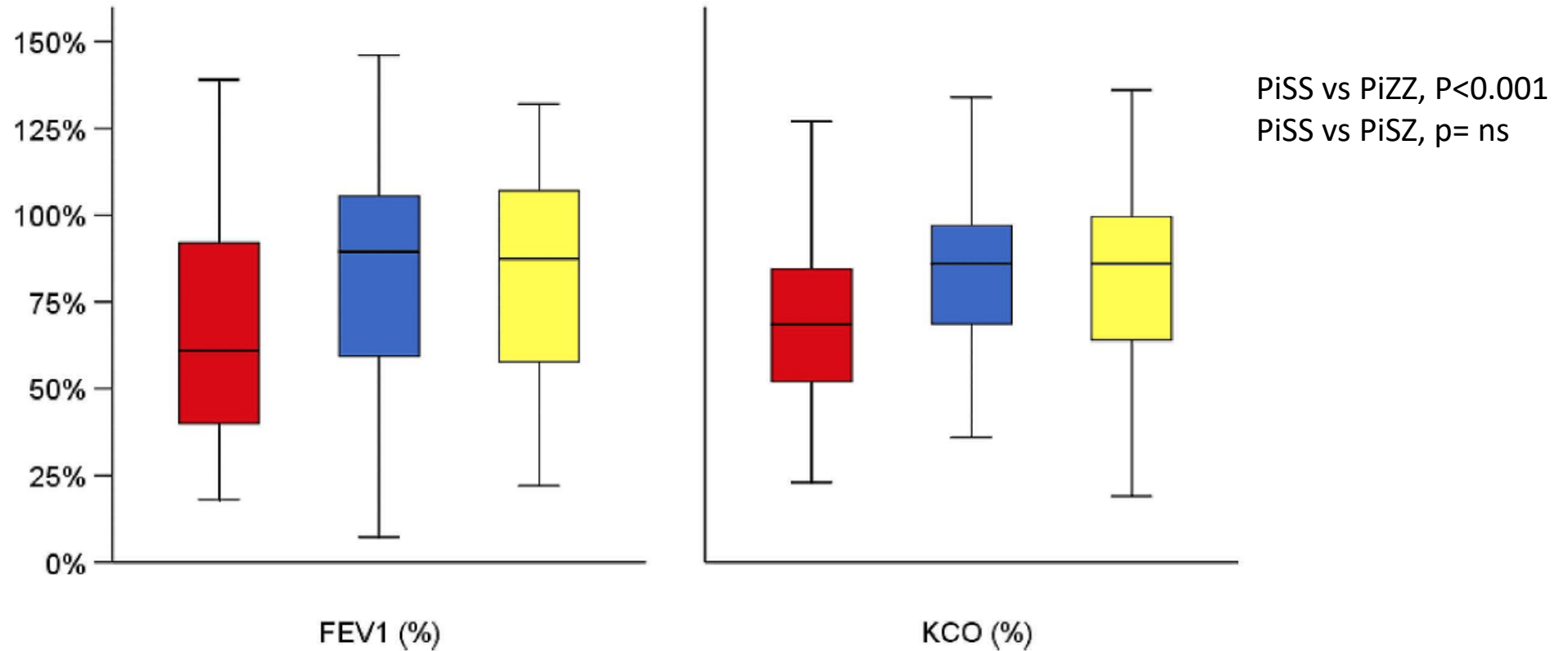
Teresa Martín^{1*}, Catarina Guimarães², Cristina Esquinas³, Maria Torres-Duran⁴, Alice M. Turner^{5,6}, Hanan Tanash⁷, Carlota Rodríguez-García⁸, Angelo Corsico^{9,10}, José Luis López-Campos^{11,12}, Eva Bartošovská¹³, Jens-Ulrik Stæhr Jensen^{14,15}, José María Hernández-Pérez¹⁶, Maria Sucena¹⁷ and Marc Miravittles¹⁸





Risk of lung disease in the PI*SS genotype of alpha-1 antitrypsin: an EARCO research project

Teresa Martín^{1*}, Catarina Guimarães², Cristina Esquinas³, Maria Torres-Duran⁴, Alice M. Turner^{5,6}, Hanan Tanash⁷, Carlota Rodríguez-García⁸, Angelo Corsico^{9,10}, José Luis López-Campos^{11,12}, Eva Bartošovská¹³, Jens-Ulrik Stæhr Jensen^{14,15}, José María Hernández-Pérez¹⁶, Maria Sucena¹⁷ and Marc Miravittles¹⁸

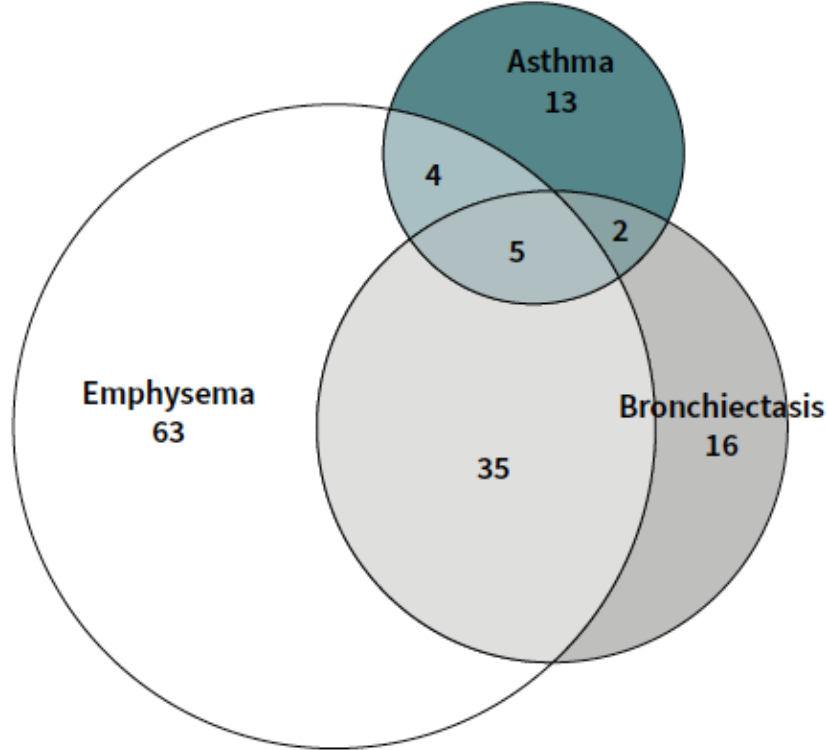




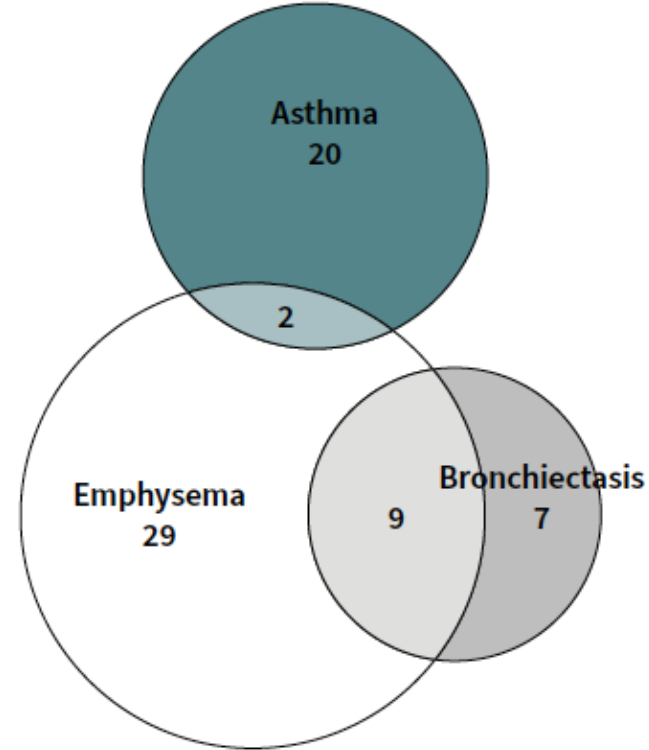
Demographic and clinical characteristics of patients with α_1 -antitrypsin deficiency genotypes PI*ZZ and PI*SZ in the Spanish registry of EARCO

María Torres-Durán¹, José Luis López-Campos ^{2,3}, Juan Luis Rodríguez-Hermosa^{4,5}, Cristina Esquinas^{2,6}, Cristina Martínez-González ⁷, José María Hernández-Pérez ⁸, Carlota Rodríguez⁹, Ana Bustamante ¹⁰, Francisco Casas-Maldonado¹¹, Miriam Barrecheguren^{2,6}, Cruz González ¹² and Marc Miravittles ^{2,6}

a)



b)









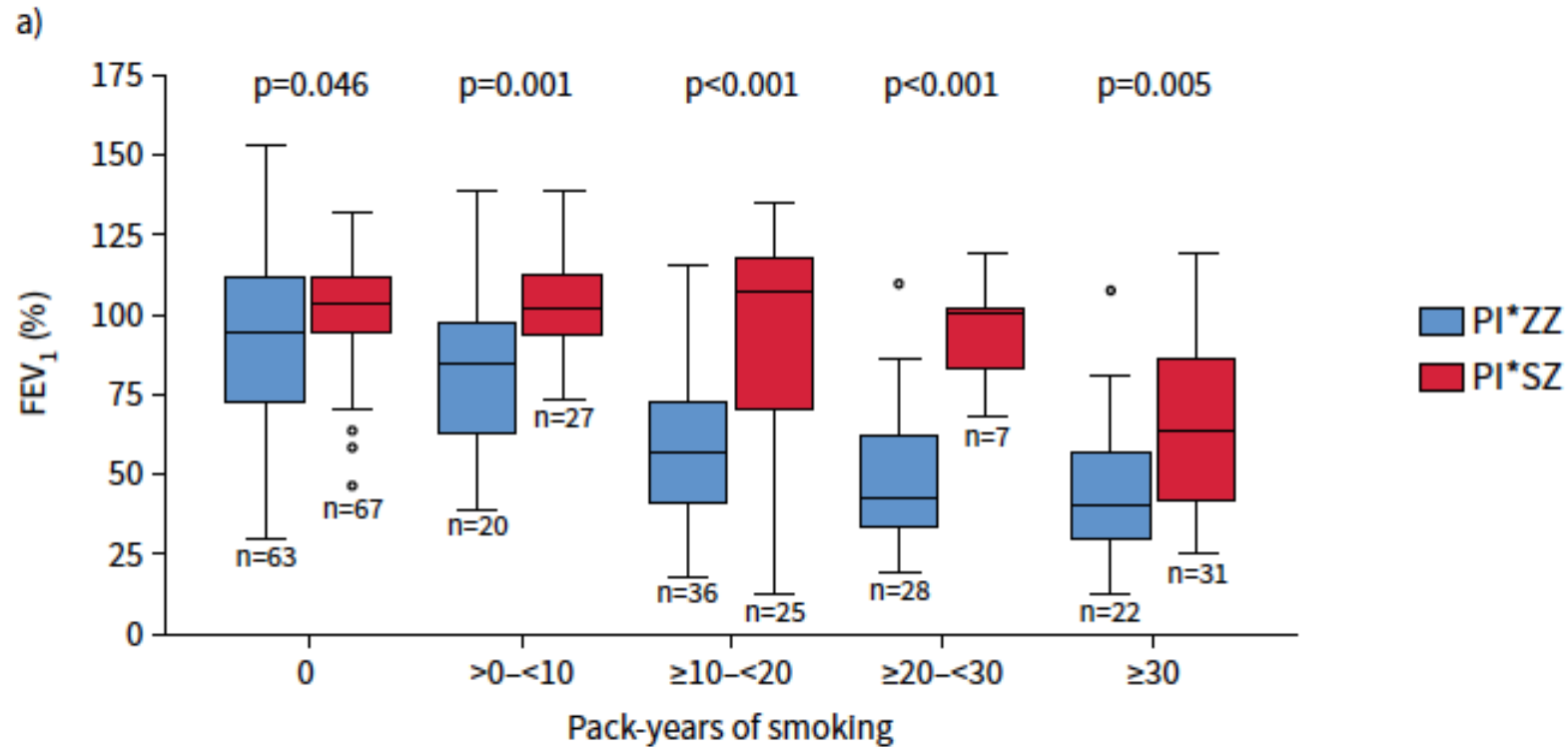
REDAVT
Registro Español de Pacientes
con Déficit de Alfa-1 Antitripsina

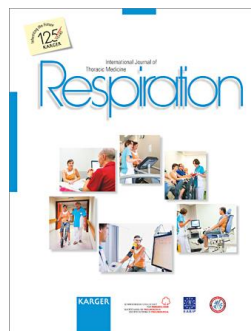
Sociedad Española
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y **Cirugía Torácica**
SEPAR



Demographic and clinical characteristics of patients with α_1 -antitrypsin deficiency genotypes PI*ZZ and PI*SZ in the Spanish registry of EARCO

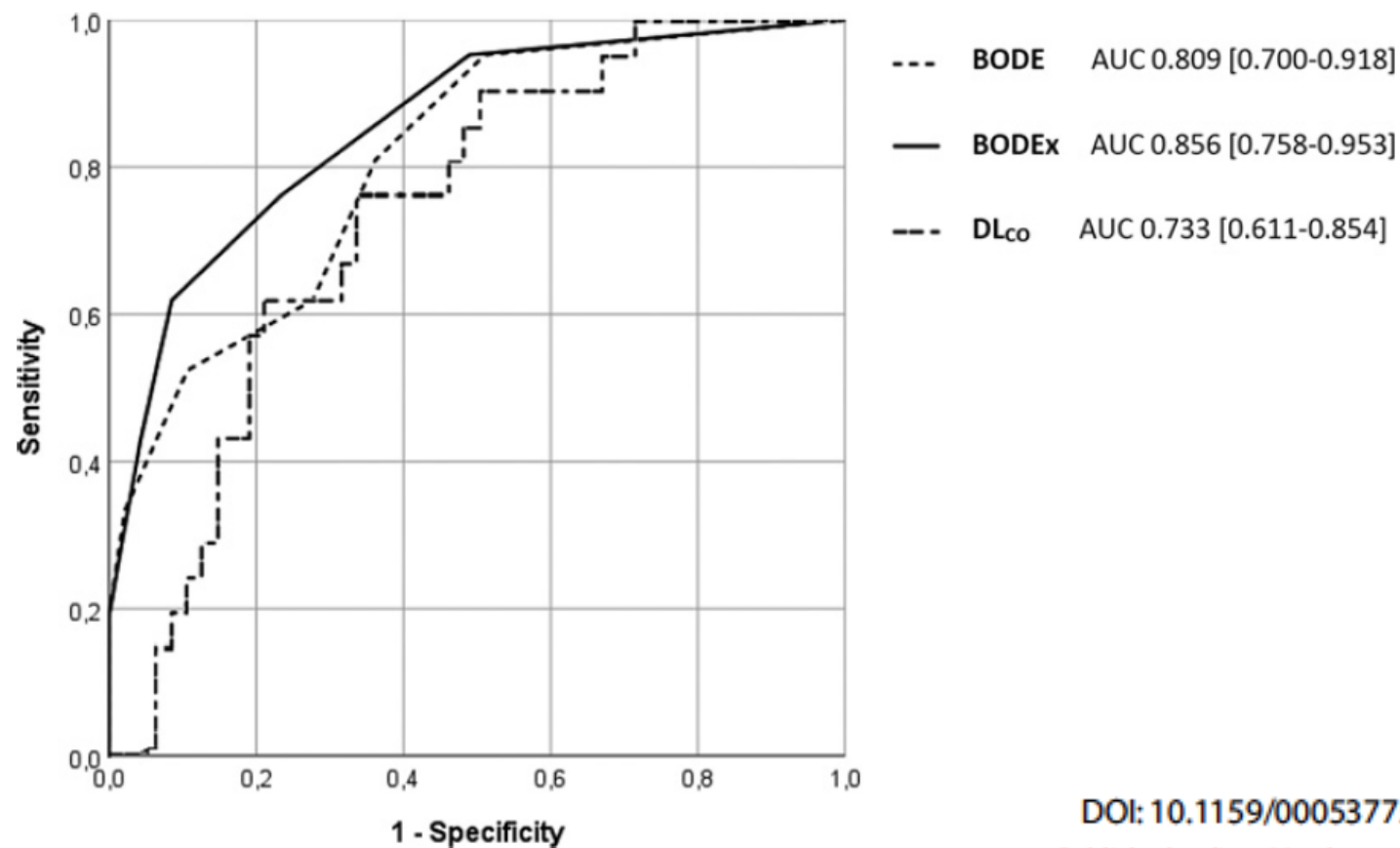
María Torres-Durán¹, José Luis López-Campos ^{2,3}, Juan Luis Rodríguez-Hermosa^{4,5}, Cristina Esquinas^{2,6}, Cristina Martínez-González ⁷, José María Hernández-Pérez ⁸, Carlota Rodríguez⁹, Ana Bustamante ¹⁰, Francisco Casas-Maldonado¹¹, Miriam Barrecheguren^{2,6}, Cruz González ¹² and Marc Miravittles ^{2,6}





Predicting Exacerbations in Alpha-1 Antitrypsin Deficiency Using Clinical and Pulmonary Function Tests: Portuguese EARCO Registry

Nuno Faria^a Joana Gomes^a Catarina Guimarães^b Raquel Marçôa^c
Beatriz Ferraz^{a,d} Maria Sucena^a



DOI: 10.1159/000537759

Published online: March 26, 2024



Assessment and monitoring of lung disease in patients with severe alpha 1 antitrypsin deficiency: a european delphi consensus of the EARCO group

Marc Miravittles^{1*}, Alice M. Turner^{2,3}, Maria Sucena⁴, Jean-François Mornex⁵, Timm Greulich⁶, Marion Wencker⁷ and N. Gerard McElvaney⁸

Initiation of augmentation therapy	● Before commencing augmentation therapy, patients should abstain from smoking for > 6 months	Agreement	5.8
	● AAT levels should be measured before commencement of augmentation therapy	Consensus	6.6
	● Augmentation therapy should only be initiated where AAT levels are < 11 μ M	Agreement	5.8
	● The age of the patient should be considered prior to initiating augmentation therapy	Agreement	5.1
	● Augmentation therapy should only be initiated after emphysema is confirmed on CT scan	Agreement	5.1
	● The patient's deterioration in FEV ₁ should determine whether to initiate augmentation therapy	Agreement	5.1
	● Extended interval dosing (e.g, biweekly) is an acceptable treatment strategy	Agreement	5.3
	● Augmentation therapy should be initiated in patients who have had a successful lung transplant	Agreement in negative	2.9
	● After initiation of augmentation therapy, AAT levels should be monitored after a change of therapy dose or interval	Agreement	5.6
	● After initiation of augmentation therapy, AAT levels should be monitored after a significant change in body weight	Agreement	5.4
● When monitoring AAT levels, levels should be recorded at trough level	Agreement	5.4	

19 publications

22-04-2021

EDITORIAL

The European Alpha-1 Research Collaboration (EARCO): a new ERS Clinical Research Collaboration to promote research in alpha-1 antitrypsin deficiency.

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22-04-2021

GUIDELINE/STATEMENT

European Respiratory Society statement: diagnosis and treatment of pulmonary disease in α 1-antitrypsin deficiency.

[SEE MORE ↗](#)

22-04-2021

REVIEW

New Patient-Centric Approaches to the Management of Alpha-1 Antitrypsin Deficiency.

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00-00-0000

ORIGINAL RESEARCH

Research priorities in α 1-antitrypsin deficiency: results of a patients' and healthcare providers' international survey from the EARCO Clinical Research Collaboration.

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EDITORIAL

Spanish Implementation of the New International Alpha-1 Antitrypsin Deficiency International Registry: The European Alpha-1 Research Collaboration (EARCO).

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EDITORIAL

Implementation of European Alpha-1 Research Collaboration (EARCO) in Portugal: the future starts now.

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ORIGINAL RESEARCH

Opinions and attitudes of pulmonologists about augmentation therapy in patients with alpha-1 antitrypsin deficiency. A survey of the EARCO Group.

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ORIGINAL RESEARCH

Impact of COVID-19 in Patients With Severe Alpha-1 Antitrypsin Deficiency: The IMCAI Study of the EARCO Clinical Research Collaboration

[SEE MORE ↗](#)

27-06-2024

ORIGINAL RESEARCH

Risk of lung disease in the PI*SS genotype of alpha-1 antitrypsin: an EARCO research project

[SEE MORE ↗](#)

06-04-2024

ORIGINAL RESEARCH

Predicting exacerbations in α 1-antitrypsin deficiency using clinical and pulmonary function tests: Portuguese EARCO registry

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12-02-2024

COFFEE-TALK

The prevalence of bronchiectasis in patients with alpha-1 antitrypsin deficiency: initial report of EARCO.

[SEE MORE ↗](#)

18-08-2023

ORIGINAL RESEARCH

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[SEE MORE ↗](#)

16-04-2023

ORIGINAL RESEARCH

Characteristics of individuals with alpha-1 antitrypsin deficiency from Northern and Southern European countries: EARCO international registry.

[SEE MORE ↗](#)

21-12-2022

ORIGINAL RESEARCH

Clinical and functional characteristics of individuals with alpha-1 antitrypsin deficiency: EARCO International Registry

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01-10-2022

ORIGINAL RESEARCH

Demographic and clinical characteristics of patients with α 1-antitrypsin deficiency genotypes PI*ZZ and PI*SZ in the Spanish registry of EARCO

[SEE MORE ↗](#)

22-04-2021

ORIGINAL RESEARCH

Protocol for the EARCO Registry: a pan-European observational study in patients with α 1-antitrypsin deficiency.

[SEE MORE ↗](#)

EARCO registry

- Launched: **Feb 2020**
- Recruiting centers: **101**
- Countries: **25**
- Patients: **3389**

Young investigators

- Call for ECM
- Identified **15 ECMs** from **9 countries**
- Now **30 ECMs** from **12 countries**
- Six ECM leading research projects

Research

- Projects: **28**
- Finished and published: **9**
- Ongoing: **12**
- Design: **7**

Community

- Partnership with ELF
- Collaboration with patient organisations
- Partnership with pharma industry

EARCO 2022



EARCO 2 (2023-2025)



- 5. Consolidate the relationship with industry and incorporate new Gold or Silver sponsors to the EARCO project to secure funding for the ongoing and future research activities.
- 6. Consider the development and implementation of a biobank with samples from patients included in the EARCO registry.
- 7. Facilitate exchange of information with the EASL European Registry of Liver disease in alpha-1 antitrypsin deficiency.