EMBARC
The European Bronchiectasis Registry

iABC
inhaled Antibiotics in Bronchiectasis and Cystic Fibrosis

ERS EUROPEAN RESPIRATORY SOCIETY

EMBARC Case Report Form
Annual Follow-up

Version 2.0 April 2016

BASIC CASE INFORMATION

Case Identifier _____

Gender: ☐ Male ☐ Female Date of birth: _____ (dd/mm/yyyy)

Center: _____

How long has the patient had bronchiectasis?

☐ Unknown
☐ < 5 years
☐ 5-10 years
☐ 11-15 years
☐ 16-20 years
☐ >20 years

Date of Review: _____ (dd/mm/yyyy)

Is the patient still under clinical follow-up? ☐ Yes ☐ No

If not, please provide reason _____

Has the patient died since last visit? ☐ Yes ☐ No

If yes, Date of Death _____ (dd/mm/yyyy)

Cause of Death _____

****

Red text indicates data fields which must be updated (questions are box highlighted for black and white printing)

Black text areas will autocomplete on the eCRF based on the data provided in the previous year.

Only edit black sections where data has changed from the previous visit.

****

CO-MORBIDITIES

Have any new comorbidities been diagnosed in the past 12 months? ☐ **Yes** ☐ **No**
(*If no, please move to next question regarding non-respiratory medications*)

Cardiovascular diseases	☐ **Yes**	☐ **No**
If yes;		
- Myocardial infarction	☐ Yes	☐ No
- Angina	☐ Yes	☐ No
- Stroke or Transient Ischaemic Attack	☐ Yes	☐ No
- Coronary artery bypass graft	☐ Yes	☐ No
- Congestive cardiac failure	☐ Yes	☐ No
- Pulmonary hypertension	☐ Yes	☐ No
- Atrial fibrillation	☐ Yes	☐ No
- Others_____		
Liver Cirrhosis	☐ Yes	☐ No
Osteoporosis	☐ Yes	☐ No
Depression	☐ Yes	☐ No
Anxiety	☐ Yes	☐ No
Chronic renal failure	☐ **Yes**	☐ **No**
If yes;		
- Haemodialysis	☐ Yes	☐ No
Neoplastic disease	☐ **Yes**	☐ **No**
If yes;		
- Active	☐ Yes	☐ No
- Haematological	☐ Yes	☐ No
- Site	☐ Lung ☐ Breast ☐ Prostate ☐ Colon ☐ Pancreas ☐ Bone ☐ Skin ☐ Brain ☐ Other ☐ Unknown	
Diabetes	☐ **Yes**	☐ **No**
If yes;		
- Type	☐ Type I ☐ Type II ☐ Unknown	
- Treatment	☐ Insulin ☐ Sulphonylurea ☐ Metformin ☐ Other	

NON RESPIRATORY MEDICATIONS

Have any changes been made to non-respiratory medications in the past 12 months?
☐ **Yes** ☐ **No**

If yes, please complete;

Statin	☐ Yes	☐ No
Angiotensin-converting-enzyme inhibitor	☐ Yes	☐ No
Angiotensin II receptor blocker	☐ Yes	☐ No
Aspirin	☐ Yes	☐ No
Non-aspirin platelet inhibitors eg, Clopidogrel	☐ Yes	☐ No
Warfarin/Oral anticoagulants	☐ Yes	☐ No
β-Blocker	☐ Yes	☐ No
Proton pump inhibitor	☐ Yes	☐ No

Additional medications can be recorded in the respiratory treatments section.

BRONCHIECTASIS BACKGROUND INFORMATION

****

Spirometry, MRC score and Exacerbation history are deemed Essential Data and will not autopopulate in the eCRF. These must be updated annually. Cases may be rejected from the registry in the absence of Essential Data.

****

Weight (Kg) _____ ☐ N/A Height (cm) _____ ☐ N/A BMI (Kg/m2) _____ ☐ N/A
(BMI autocalculated by eCRF)

FEV_1 L (recorded) _____ ☐ N/A FEV_1 L (% predicted) _____ ☐ N/A
(% predicted values as autocalculated by eCRF)

FVC L (recorded) _____ ☐ N/A FVC L (% predicted) _____ ☐ N/A
(% predicted values as autocalculated by eCRF)

Bronchodilator Status
☐ Pre-Bronchodilator
☐ Post-Bronchodilator
☐ Unknown
(where possible, post-bronchodilator values are preferred)

If spirometry has not been completed in the past 12months, please give a reason in the box below

Were any additional lung function tests performed? ☐ Yes ☐ No

If yes;
Total Lung Capacity (L)_____ ☐ N/A Diffusing capacity (DLCO) (L)_____ ☐ N/A

Residual Volume (L)_____ ☐ N/A Inspiratory capacity (DLCO) (L)_____ ☐ N/A

Modified MRC dyspnoea score:

☐ 0 (I only get breathless with strenuous exercise)
☐ 1 (I get short of breath when hurrying on level ground or walking up a slight hill)
☐ 2 (On the level ground I walk slower than people of the same age because of breathlessness or I have to stop for breath when walking at my own pace on the level)
☐ 3 (I stop for breath after walking about 100 yards or after a few minutes on the level ground)
☐ 4 (I am too breathless to leave the house or I am breathless when dressing)

Asthma: ☐ Yes ☐ No COPD: ☐ Yes ☐ No

Nasal polyps: ☐ Yes ☐ No Rhinosinusitis: ☐ Yes ☐ No

Sputum color when stable: ☐ Mucoid ☐ Mucopurulent ☐ Purulent ☐ Purulent (severe)

Usual daily sputum volume:_____(ml/day)

Smoking status: ☐ Current ☐ Ex ☐ Never

Approximate Pack years: ☐ 0 - 4 ☐ 5 – 9 ☐ 10 - 20 ☐ 21 – 40 ☐ More than 40

Number of exacerbations **not** requiring secondary care in the last year:
☐ 0 ☐ 1 ☐ 2 ☐ 3 ☐ 4 ☐ 5 ☐ 6 ☐ 7 ☐ 8 ☐ 9 ☐ 10 ☐ 11 ☐ 12
Source of this data:
☐ Patient history ☐ Antibiotic prescription data ☐ Hospital records

Number of exacerbations requiring **hospital admission** in the last year:
☐ 0 ☐ 1 ☐ 2 ☐ 3 ☐ 4 ☐ 5 ☐ 6 ☐ 7 ☐ 8 ☐ 9 ☐ 10 ☐ 11 ☐ 12
Source of this data:
☐ Patient history ☐ Antibiotic prescription data ☐ Hospital records

Number of respiratory related **emergency department visits not resulting in hospitalisation** in the last year:
☐ 0 ☐ 1 ☐ 2 ☐ 3 ☐ 4 ☐ 5 ☐ 6 ☐ 7 ☐ 8 ☐ 9 ☐ 10 ☐ 11 ☐ 12
Source of this data:
☐ Patient history ☐ Antibiotic prescription data ☐ Hospital records

Has the patient ever been hospitalised for bronchiectasis? ☐ Yes ☐ No

Has the patient received outpatient intravenous antibiotics in the last year? ☐ Yes ☐ No

Has the patient ever had major haemoptysis requiring hospital admission? ☐ Yes ☐ No

Has the patient participated in a clinical trial for bronchiectasis (other than the registry)? ☐ Yes ☐ No

QOL-B QUESTIONNAIRE

Please provide recent QoL-B data (not more than 12 months old). QOL-B data is deemed Essential Data for those with appropriately verified translations.

☐ English-UK ☐ Danish-Denmark ☐ Dutch-Belgium ☐ Dutch-Netherlands ☐ Finnish
☐ French-Belgium ☐ French-France ☐ German ☐ Hungarian ☐ Italian ☐ Lithuanian
☐ Norwegian ☐ Polish ☐ Portuguese ☐ Romanian ☐ Russian-Israel ☐ Russian- Russia
☐ Serbian ☐ Spanish-Latin ☐ Spanish-Spain

Date of completion: _____ (dd/mm/yyyy)

Q1 _____	Q2 _____	Q3 _____	Q4 _____	Q5 _____
Q6 _____	Q7 _____	Q8 _____	Q9 _____	Q10_____
Q11_____	Q12_____	Q13_____	Q14_____	Q15_____
Q16_____	Q17_____	Q18_____	Q19_____	Q20_____
Q21_____	Q22_____	Q23_____	Q24_____	Q25_____
Q26_____	Q27_____	Q28_____	Q29_____	Q30_____
Q31_____	Q32_____	Q33_____	Q34_____	Q35_____
Q36_____	Q37_____			

AETIOLOGY AND LABORATORY TESTING

Has the patient had any new aetiology tests in the past 12 months? ☐ **Yes** ☐ **No**
(If not, please move to next question)

ABPA ☐ **Yes** ☐ **No**
If yes;

- Serum eosinophil count ☐ Elevated ☐ Normal ☐ Not tested
- Total IgE _____iu/mL ☐ Normal ☐ Elevated ☐ Not tested
- Specific IgE to aspergillus ☐ Raised ☐ Normal ☐ Not tested
- Aspergillus IgG ☐ Raised ☐ Normal ☐ Not tested
- Aspergillus Skin prick test ☐ Raised ☐ Normal ☐ Not tested

Cystic Fibrosis ☐ **Yes** ☐ **No**
If yes;

- Sweat test ☐ Positive ☐ Intermediate ☐ Negative ☐ Not performed
- Genetics ☐ Homozygous ☐ Heterozygous ☐ No mutations ☐ Not performed

Serum Immunoglobulins ☐ **Yes** ☐ **No**
If yes;

- Serum level IgM ☐ Normal ☐ Low ☐ High ☐ Not tested
- Serum level IgG ☐ Normal ☐ Low ☐ High ☐ Not tested
- Serum level IgA ☐ Normal ☐ Low ☐ High ☐ Not tested
- Serum level IgG1 ☐ Normal ☐ Low ☐ High ☐ Not tested
- Serum level IgG2 ☐ Normal ☐ Low ☐ High ☐ Not tested
- Serum level IgG3 ☐ Normal ☐ Low ☐ High ☐ Not tested
- Serum level IgG4 ☐ Normal ☐ Low ☐ High ☐ Not tested

α-1 antitrypsin deficiency ☐ **Yes** ☐ **No**
If yes;

- Level ☐ Normal ☐ Low ☐ High ☐ Not tested
- Genetics ☐ PiMM (Normal) ☐PiMS ☐ PiSS ☐ PiMZ ☐ PiSZ ☐ PiZZ ☐ Not performed

Functional antibodies to Pneumococcal/H influenza vaccine ☐ **Yes** ☐ **No**
If yes;

- Result ☐ Normal ☐ Abnormal

Serum electrophoresis ☐ **Yes** ☐ **No**
If yes;

- Result ☐ Normal ☐ Abnormal

Tests of ciliary function ☐ **Yes** ☐ **No**

If yes;

- Nasal eNO ☐ Positive ☐ Intermediate ☐ Negative ☐ Not performed
- Saccharin test ☐ Positive ☐ Intermediate ☐ Negative ☐ Not performed
- Scintigraphic mucociliary clearence ☐ Positive ☐ Intermediate ☐ Negative ☐ Not performed
- Biopsy for electron microscopy ☐ Positive ☐ Intermediate ☐ Negative ☐ Not performed
- Biopsy for analysis of ciliary beat patter/frequency ☐ Positive ☐ Intermediate ☐ Negative ☐ Not performed
- Genetics ☐ Positive ☐ Intermediate ☐ Negative ☐ Not performed

Bronchoscopy ☐ **Yes** ☐ **No**

Autoantibody testing ☐ **Yes** ☐ **No**

If yes;

- CCP screen results ☐ Positive ☐ Intermediate ☐ Negative ☐ Not performed
- ANA screen results ☐ Positive ☐ Intermediate ☐ Negative ☐ Not performed
- ENA screen results ☐ Positive ☐ Intermediate ☐ Negative ☐ Not performed
- ANCA ☐ Positive ☐ Intermediate ☐ Negative ☐ Not performed
- Additional tests performed _____

Has the patient been diagnosed with any of the following in the past 12 months?
(If not, please move to microbiology section) ☐ **Yes** ☐ **No**

Pneumonia ☐ Yes ☐ No

Whooping cough/pertussis ☐ Yes ☐ No

Other childhood/respiratory infection ☐ Yes ☐ No

Tuberculosis ☐ **Yes** ☐ **No**
If yes;
- Infection ☐ Current ☐ Previous
- Treatment received ☐ Yes ☐ No ☐ Unknown

Atypical mycobacterial infection ☐ **Yes** ☐ **No**
If yes;
- Infection ☐ Current ☐ Previous
- Treatment received ☐ Yes ☐ No ☐ Unknown

Rheumatoid arthritis ☐ Yes ☐ No

Other connective tissue disease ☐ **Yes** ☐ **No**
If yes;

☐ Systemic lupus erythematosus
☐ Systemic sclerosis/scleroderma
☐ Ehlers_danlos syndrome
☐ Mixed connective tissue disease
☐ Stills disease
☐ Sjogrens syndrome
☐ Poly/dermatomyositis
☐ Juvenile idiopathic asthritis
☐ Relapsing polychondritis
☐ Other

Inflammatory bowel disease ☐ **Yes** ☐ **No**
If yes;
- Ulcerative colitis ☐ Yes ☐ No
- Crohns disease ☐ Yes ☐ No

HIV ☐ Yes ☐ No

Immunodeficiency ☐ **Yes** ☐ **No**

If yes;

- B-cell deficiencies:
  - ☐ Common variable immunodeficiency
  - ☐ X-linked agammaglobulinaemia
  - ☐ Thymoma with antibody deficiency
  - ☐ Hyper IgM syndrome
  - ☐ Activate PI3K-delta syndrome
  - ☐ Selective IgA deficiency
  - ☐ IgG subclass deficiency
  - ☐ Specific antibody deficiency
  - ☐ Other

- T-cell and combined deficiencies
  - ☐ Severe combined immunodeficiency
  - ☐ DiGeorge syndrome
  - ☐ X-linked lymphoproliferative syndrome
  - ☐ Hyper IgM syndrome (CD40 ligand)
  - ☐ MHC class II deficiency
  - ☐ Ataxia-telangiectasis
  - ☐ Wiskott-Aldrich syndrome
  - ☐ Chronic mucocutaneous candidiasis
  - ☐ TAP deficiency
  - ☐ IPEX (immune dysfunction, polyendocrinopathy, eneteropathy, X-linked)
  - ☐ ALPS (autoimmune lymphoproliferative syndrome)
  - ☐ WHIM syndrome
  - ☐ Other

- Secondary immunodeficiencies
  - ☐ Chronic Lymphocytic leukemia
  - ☐ Multiple Myeloma
  - ☐ Immunodeficiency associated with haematological malignancy
  - ☐ Immunodeficiency secondary to systemic chemotherapy
  - ☐ Immunodeficiency secondary to immunosuppressive drugs
  - ☐ Stem cell transplantation
  - ☐ Solid organ transplantation
  - ☐ Other

- Phagocyte deficiencies
  - ☐ Chronic granulomatous disease
  - ☐ Familial Haemophagocytic lymphohistiocytosis
  - ☐ Congenital agranulocytosis
  - ☐ Cyclic neutropenia
  - ☐ Leucocyte adhesion deficiency
  - ☐ Chediak-Higashi syndrome
  - ☐ Griscelli's syndrome
  - ☐ Hyper IgE syndrome
  - ☐ Interferon gamma/IL-12 rec
  - ☐ Other cytokine deficiencies

- Complement deficiencies
  - ☐ Mannose binding lectin (MBL) deficiency
  - ☐ Properdin deficiency
  - ☐ Complement C3 deficiency
  - ☐ Terminal complement component deficiency
  - ☐ Other

Primary ciliary dyskinesia ☐ Yes ☐ No

Aspiration ☐ Yes ☐ No

Gastro-oesophageal reflux disease ☐ Yes ☐ No

Congenital airway abnormality ☐ Yes ☐ No
If yes, please specify: _____

Foreign body inhalation or obstruction ☐ Yes ☐ No

EMBARC
The European Bronchiectasis Registry

iABC inhaled Antibiotics in Bronchiectasis and Cystic Fibrosis

ERS EUROPEAN RESPIRATORY SOCIETY

MICROBIOLOGY

****

Microbiology is deemed Essential Data. This must be updated annually.
Cases may be rejected from the registry in the absence of Essential Data.

****

Have any new microbiology samples been obtained in the past 12 months? ☐ Yes ☐ No

If no;
Microbiology samples should be collected from the bronchiectasis patients at least once per year, if no sample has been collected please provide a reason: _____
_____

If yes, complete the following:
Samples are dividd into those performed when clinically stable and those performed during exacerbation. If it is uncertain whether patients were stable or not at the time of sampling please record under "clinically stable".

While clinically stable

Please provide details of all sputum results while stable over the last 12 months including negative cultures (use additional sheets where necessary)

Date of sample: _____ (mm/yyyy)

Source:
☐ Sputum
☐ BAL
☐ Induced sputum
☐ Throat swab

☐ No organism isolated
☐ Organism:_____

Antibiotic: Sensitive:_____
Sensitive:_____
Sensitive:_____
Sensitive:_____
Sensitive:_____
Resistant:_____
Resistant:_____
Resistant:_____
Resistant:_____

☐ Organism:_____

Antibiotic: Sensitive:_____
Sensitive:_____
Sensitive:_____
Sensitive:_____
Resistant:_____
Resistant:_____
Resistant:_____

During Exacerbations

Please provide details of <u>all</u> sputum results during exacerbations over the last 12 months (use additional sheets where necessary)

Date of sample: _____ (mm/yyyy)

Source:
☐ Sputum
☐ BAL
☐ Induced sputum
☐ Throat swab

☐ No organism isolated
☐ Organism:_____

Antibiotic: Sensitive:_____
Sensitive:_____
Sensitive:_____
Sensitive:_____
Sensitive:_____
Resistant:_____
Resistant:_____
Resistant:_____
Resistant:_____

☐ Organism:_____

Antibiotic: Sensitive:_____
Sensitive:_____
Sensitive:_____
Sensitive:_____
Sensitive:_____
Resistant:_____
Resistant:_____
Resistant:_____
Resistant:_____

Mycobacterial samples

Please provide details of <u>all</u> sputum results for acid fast bacilli/mycobacterial culture over the last 12 months (use additional sheets where necessary).

Date of sample: _____ (mm/yyyy)

Source:
☐ Sputum
☐ BAL
☐ Induced sputum
☐ Throat swab

☐ No organism isolated
☐ Organism:_____

RADIOLOGY

Has the patient had an updated CT scan in the past 12 months? ☐ **Yes** ☐ **No**

If yes:

Date of CT scan: _____ (dd/mm/yyyy)

Type of imaging:
☐ High resolution CT scan (HRCT)
☐ CT Thorax

Is there CT evidence of Bronchiectasis in;

Right upper lobe:
☐ No Bronchiectasis
☐ Cylindrical
☐ Varicose
☐ Cystic
☐ Unknown Severity

Left upper lobe:
☐ No Bronchiectasis
☐ Cylindrical
☐ Varicose
☐ Cystic
☐ Unknown Severity

Right middle lobe:
☐ No Bronchiectasis
☐ Cylindrical
☐ Varicose
☐ Cystic
☐ Unknown Severity

Lingula:
☐ No Bronchiectasis
☐ Cylindrical
☐ Varicose
☐ Cystic
☐ Unknown Severity

Right lower lobe:
☐ No Bronchiectasis
☐ Cylindrical
☐ Varicose
☐ Cystic
☐ Unknown Severity

Left lower lobe:
☐ No Bronchiectasis
☐ Cylindrical
☐ Varicose
☐ Cystic
☐ Unknown Severity

RESPIRATORY TREATMENTS

Have there been any changes to respiratory medication in the past 12 months? ☐ **Yes** ☐ **No**

Long term oxygen therapy: ☐ Yes ☐ No

Non invasive ventilation: ☐ Yes ☐ No

The patient has regular respiratory treatments: ☐ **Yes** ☐ **No**
If yes, complete below;

Respiratory Medications

☐ Inhaled steroid	Drug:_____
☐ Inhaled steroid/Long acting beta agonist	Drug:_____
☐ Intravenous Immunoglobulin	Drug:_____
☐ Itraconazole	Drug:_____
☐ Leukotriene receptor antagonist	Drug:_____
☐ Long acting anti-muscarinic	Drug:_____
☐ Long acting beta agonist/Long acting anti-muscarinic	Drug:_____
☐ Long acting beta agonist	Drug:_____
☐ Long term (>28 days) Oral corticosteroids	Drug:_____
☐ Monoclonal antibody	Drug:_____
☐ Mucolytic	Drug:_____
☐ Nebulised bronchodilators	Drug:_____
☐ Oral theophylline	

Antibiotic Medications

☐ Inhaled/Nebulised antibiotics	Drug:_____
☐ Long term (>28 days) Oral antibiotics	Drug:_____
☐ Cyclical antibiotic therapy	Drug:_____

Physiotherapy Adjuncts

☐ DNAase	☐ Nebulised Normal saline
☐ Inhaled mannitol	☐ Sodium Hyaluronate
☐ Nebulised Hypertonic saline	

Vaccination

Pneumococcal polysaccharide vaccine (e.g.: PSV23):	☐ Yes	☐ No
Pneumococcal conjugate vaccine (e.g.: PCV13):	☐ Yes	☐ No
Influenza vaccine:	☐ Yes	☐ No

PHYSIOTHERAPY AND ACTIVITY

Within the last 12months has the patient

- Seen a specialist physiotherapist ☐ Yes ☐ No
- Had a self management plan written ☐ Yes ☐ No
- Practiced regular chest physiotherapy ☐ Yes ☐ No

Manual airway clearance:
☐ Active cycle of breathing technique
☐ Autogenic drainage
☐ Postural drainage
☐ Assisted cough
☐ Manual vibration
☐ Percussion
☐ ELTGOL
☐ None
☐ Regular physical exercise

Devices:
☐ Positive expiratory pressure (PEP) device
☐ Flutter device
☐ Cornet
☐ Acapella
☐ Mechanical vibration
☐ Percussionaire
☐ High frequency chest wall oscillation
☐ Other
☐ None

Has the patient attended pulmonary rehabilitation?
☐ Yes
☐ Not referred
☐ Not fit due to co-morbidities
☐ Patient refused
☐ Patient failed to attend

ADDITIONAL INFORMATION

Provide any additional required information in the free text provided:

_____
_____
_____
_____
_____
_____
_____

Disclaimer
In using this paper case report form to record identifiable patient data, the user accepts all responsibility for the secure storage of this data and disposal of this data in accordance with local ethical approvals and policies.

Acknowledgements
EMBARC is a European Respiratory Society Clinical Research Collaboration.
This case report form was created with the kind support of:

- Innovative Medicines Initiative
- European Respiratory Society
- The United States COPD Foundation Bronchiectasis Registry
- Bronch-UK, the UK bronchiectasis network
- The European Cystic Fibrosis Society
- Italian Society of Respiratory Medicine (SIP)
- Lung Foundation of Australia
- The European Lung Foundation
- Bayer HealthCare
- Novartis Pharma AG
- Aradigm Corporation
- The Prospective German Non-CF Bronchiectasis (PROGNOSIS) Registry
- The Group for Research and Education in Pneumo-Infectious Diseases (GREPI), France