

# ***Building the European CF Registry***

***The challenge***

# ***Board of the ECFS European CF Registry***

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# ***The aims of the ECFS European CF Registry***

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Providing essential information for the ECFS confined to answer the following questions:

- 👉 How many CF patients are there in Europe?
- 👉 What is their clinical status, hence their needs?
- 👉 What are the annual trends of survival?

This information will facilitate long-term planning of health expenditure allocations and developing pan European support systems.

## ***The aims of the European CF Registry cont.***

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Building an infrastructure for data collection for countries that do not yet have national registries (most of the European countries).

## ***The aims of the European CF Registry cont.***

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- Introducing standardization of data collection for the national registries.
- Promoting standardization of care.

# ***The aims of the European CF Registry cont.***

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Comparing clinical data from different centers and different countries in order to improve the care of the patients.

- 👉 Providing data resources so that center directors can compare their own center to other European centers.
- 👉 Providing an audit instrument that will enable the identification of centers requiring support by the ECFS.
- 👉 Responding to the patients' right to know if they are receiving the standard of care treatment.

# ***Long term aims of the European CF Registry cont.***

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- Facilitating research on rare CF presentations.
- Facilitating research on uncommon complications.
- Enabling identification of patients or centers that are suitable for clinical studies on new therapies or genetic research.
- Enabling the design of studies with strict inclusion criteria that require the participation of few patients from multiple centers.
- Providing tools to identify topics that need to be discussed in the ECFS conferences.
- Providing tools to identify areas that need further research.

# ***The Characteristics of the European CF Registry***

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- ✓ Simple,
- ✓ Usable
- ✓ Manageable
- ✓ Non-expensive
- ✓ Compatible with the already existing national registries
- ✓ Expandable



## ***Two level action:***

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Merging data from the already existing registries.

Building registries in centers that are not associated with national registries.

## ***Other Important Issues***

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- Standardization of data collection  
**There is an urgent need to develop an International CF Standard for data collection**
- Constitution and ethical issue

# ***Basic data*** (7 fields)

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Country code number + patient's original code

Month and year of birth (mm/yyyy)

Gender (1=male; 2= female)

Genotype 1

Genotype 2

# *Diagnosis*

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- Age at diagnosis (decimals)
- Sweat chloride meq/L [highest test], positive, not done

# ***Mode of presentation***

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- Neonatal screening
- Family history
- Meconium ileus
- Gastrointestinal/FTT/malnutrition
- Respiratory infections symptoms
- Anemia/hypoproteinemia
- Electrolyte imbalance
- Chronic sinusitis
- Nasal polyps
- Male infertility
- Recurrent pancreatitis
- Other

# Yearly measurement

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1. Patient alive 1=yes, 2=no
2. Pancreatic status (PS=1, PI= 2)
3. Height (cm) [last test ]
4. Height (%tile) [last test]
5. Weight (kg) [last test]
6. Weight (%tile) [last test]
7. BMI [last test]
8. BMI (%tile) [last test]
9. FVC liter [last test]
10. FEV1 liter [last test]

# ***Bacteriology***

***(if present in the last year at least once)***

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11. Chronic *Pseudomonas aeruginosa*
12. Multi-resistant Chronic *Pseudomonas aeruginosa*
13. *Pseudomonas aeruginosa* not chronic
14. *Staph aureus*
15. *Staph aureus* (MRSA)
16. Nontuberculous mycobacteria
17. *Stenotrophomonas maltophilia*
18. *Alcaligenes xylosoxidans*
19. *Burkholderia cepacia* complex
20. *Hemophilus influenzae*

# ***Current Therapy***

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21. Inhaled bronchodilators
22. Inhaled steroids
23. Inhaled antibiotic (1=none; 2=intermittent; 3= continuous)
24. Pulmozyme
25. Inhaled hypertonic NaCl
26. Continuous Oral antibiotics
27. Ursolit
28. Antacid (H2 antagonists. PPI)
29. Systemic steroids
30. Oxygen therapy
31. Taurine



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32. Total CF clinic visits in last year
  33. Hospitalizations, 'in hospital'-number per year,
  34. Hospitalizations, 'in hospital'-days/year.
  35. Total number of days on IV antibiotics

# ***Complications***

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36. Hemoptysis – major over 250 ml (1=no; 2=once; 3=recurrent)
37. Pneumothorax requiring chest drain
38. ABPA
39. Nasal polyps
40. Pancreatitis
41. DIOS
42. GER requiring treatment
43. Fibrosing colonopathy
44. CFRD (1= no; 2= IGGT; 3= insulin treated)
45. Late complications of diabetes
46. Elevated liver enzymes (1.5 times the upper limit)
47. Biliary cirrhosis\ Portal hypertension Bleeding varices
48. CF related bone disease
49. Urinary incontinence
50. Kidney stones
51. CF related vasculitis, arthropathy
52. Renal failure requiring dialysis / transplant
53. Malignancy
54. Liver transplant
55. Lung transplant /lung+heart

## **Social report**

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- 56. **Marital status** (if over 18yrs) (1=single; 2=living together; )
  
- 57. **Employment** (if > 18yrs) (1 =yes; 2=student; 3=unable to work; 4 = unemployed)
  
- 58. **Children – number**