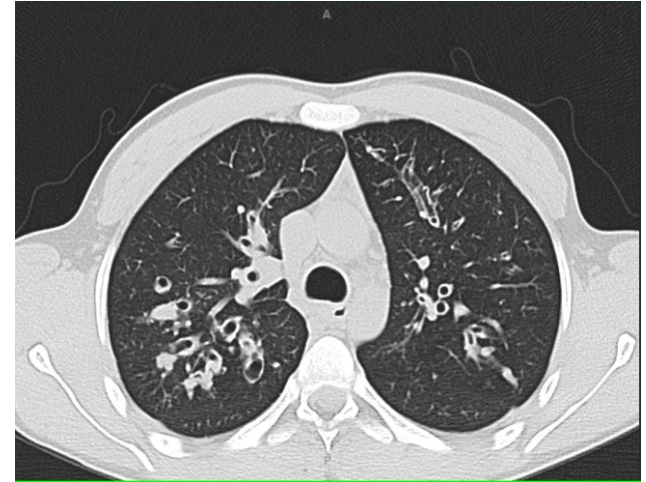


Cystic Fibrosis

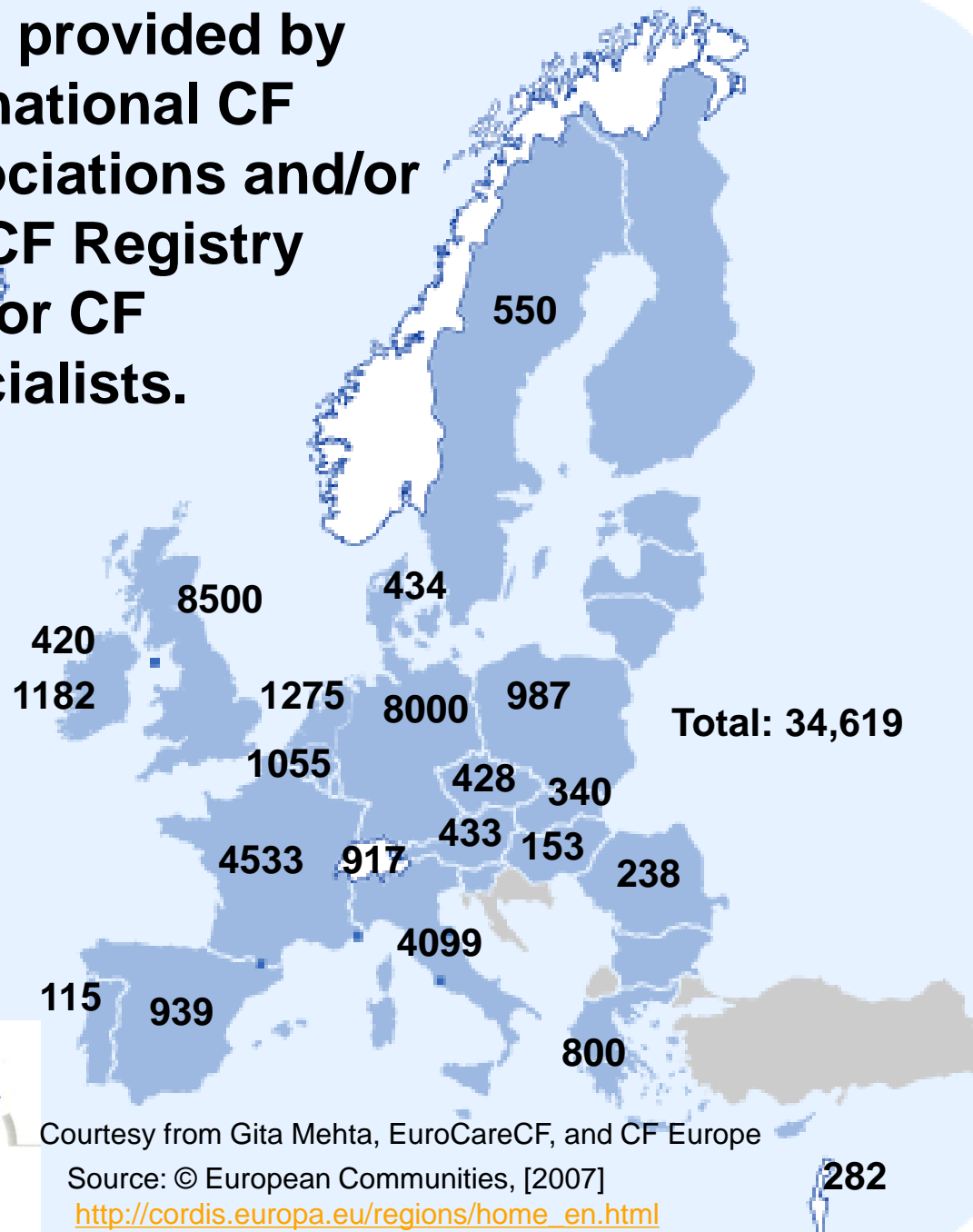
Demographic Challenges for Europe



J Stuart Elborn
Centre of Infection and Immunity, QUB
and
Adult CF Centre, Belfast HSCT



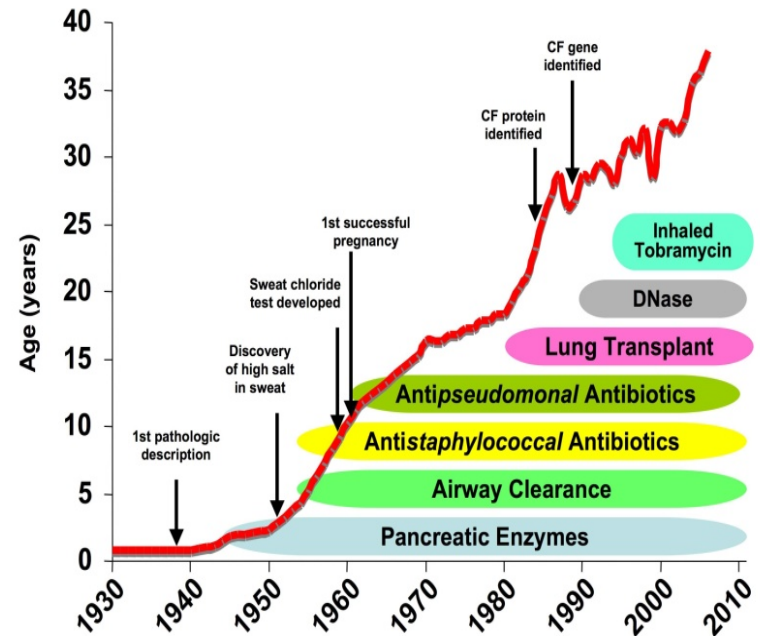
**Data provided by
the national CF
associations and/or
the CF Registry
and/or CF
specialists.**



- Austria
- Belgium
- Czech R
- Denmark
- France
- Germany
- Greece
- Hungary
- Ireland
- Israel
- Italy
- Netherlands
- Poland
- Portugal
- Romania
- Slovakia
- Spain
- Sweden
- Switzerland
- UK

Improvements in survival

- Median age at death has increased dramatically
- In the UK children born in the 21st century are estimated to have a median survival of over 50 years of age (Dodge 2007)
- “under the influence of factors such as improved treatment and management, improved nutrition and better living conditions”



Adapted From 2005 Annual Data Report to the Center Directors. Cystic Fibrosis Patient Registry, Bethesda, MD
<http://knol.google.com/k/jerry-nick/cystic-fibrosis/Utl7gr91/HU2blw#> (24 April 09)

1940's



1980's



1990's



REMARKABLE CHANGE IN PROGNOSIS FOR PEOPLE WITH CF

Ben is now the poster boy – the new look, if you will – of cystic fibrosis (CF), the incurable life-threatening genetic condition

Read more:

<http://www.dailymail.co.uk/health/article-2193518/Could-male-pin-Ben-Mudges-incredible-pack-really-key-conquering-cystic-fibrosis.html#ixzz25lQqUCTc>

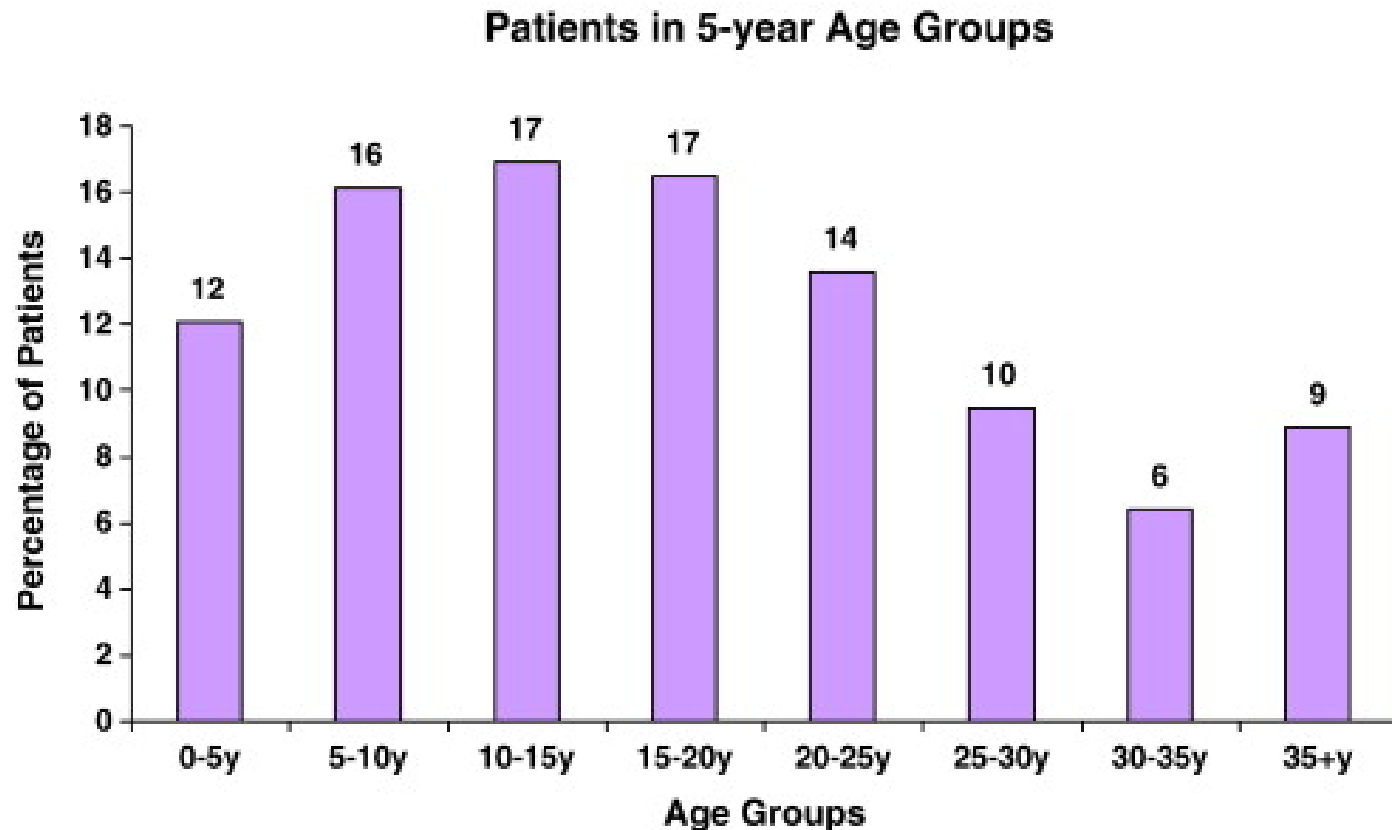


© Matthew Pover



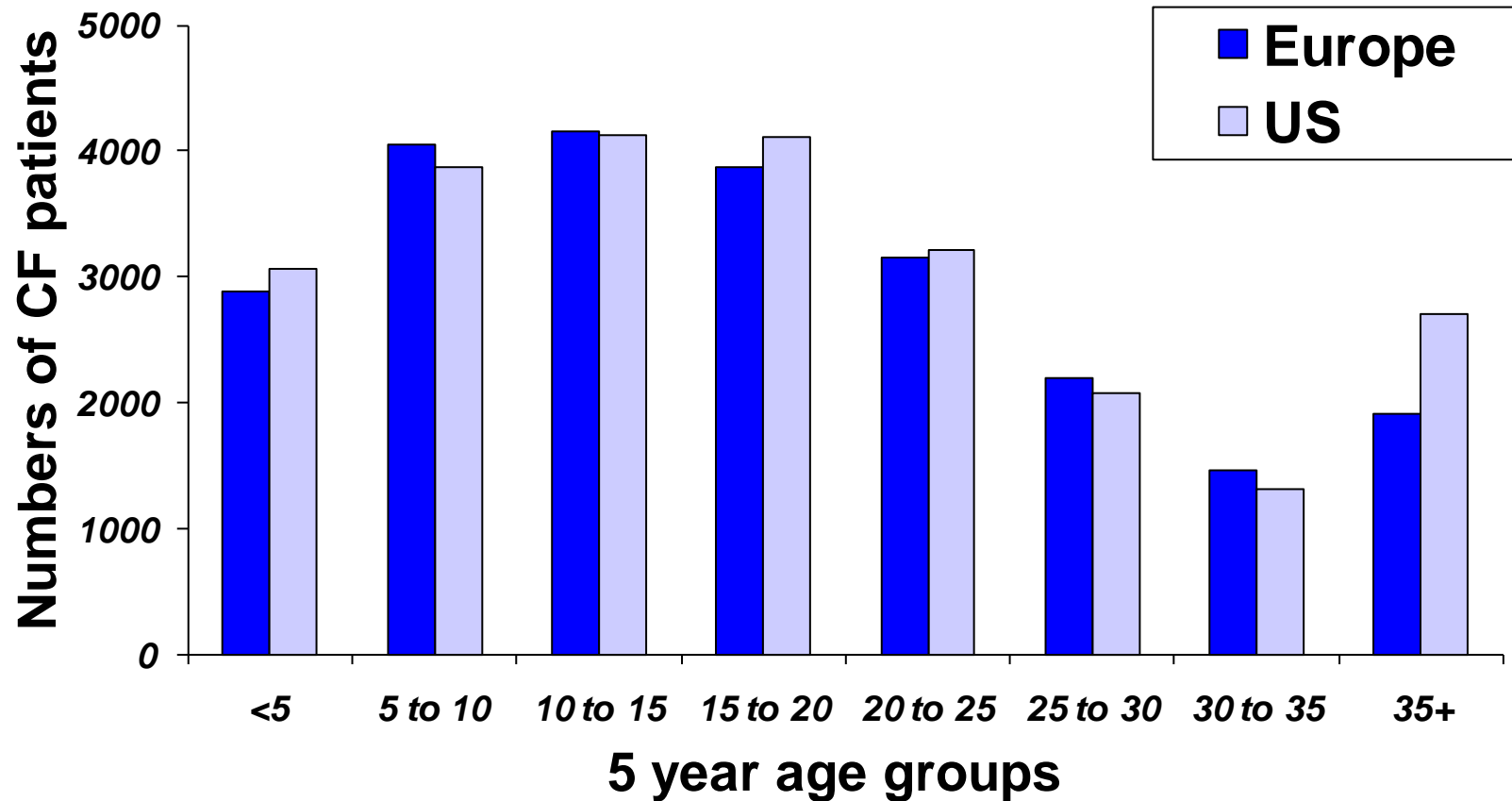
© Matthew Pover

Age distribution in Europe



0-5y	5-10y	10-15y	15-20y	20-25y	25-30y	30-35y	>35y	Total
3524	4672	4920	4806	3959	2774	1856	2584	29,095

Europe / US Age Distributions

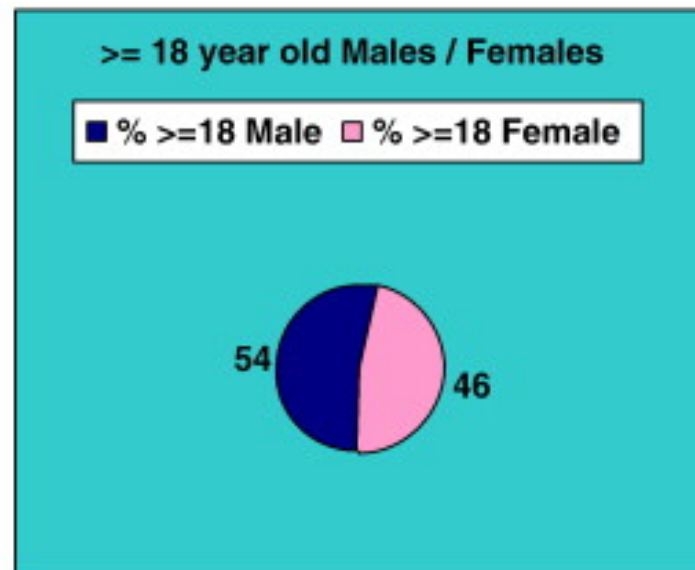
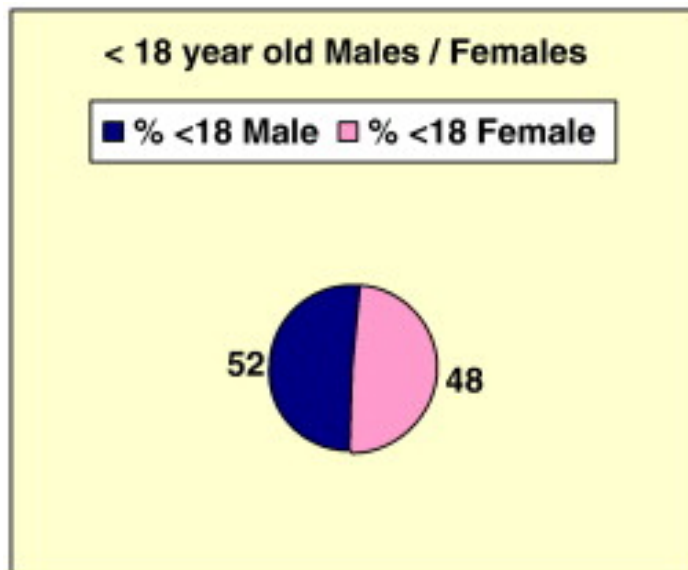


Patients from Europe: 23693, US: 24487, Total: 48180

21 countries from Europe + US data shown

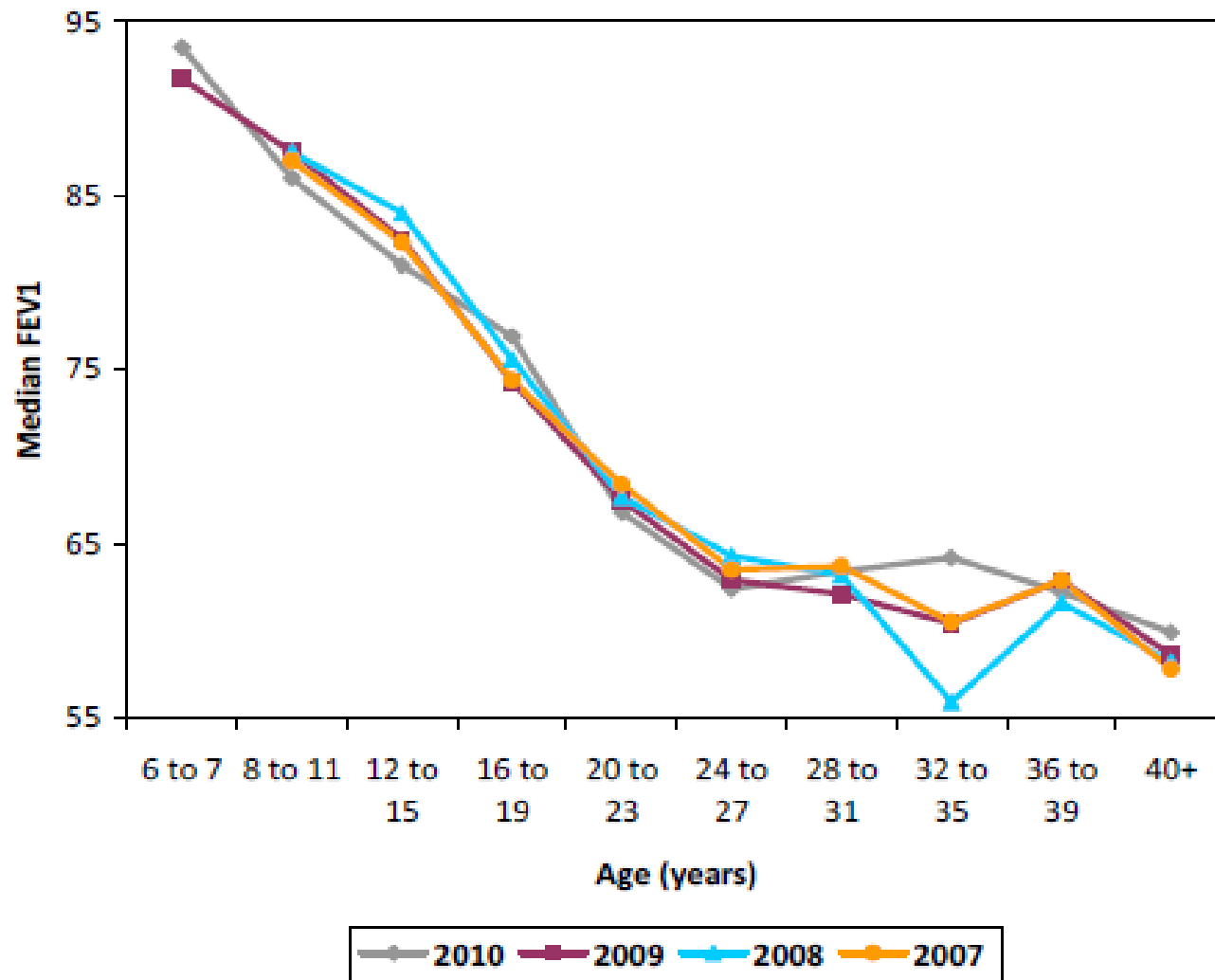
Data from CFF Registry, ECFS & EuroCareCF

Gender in Europe

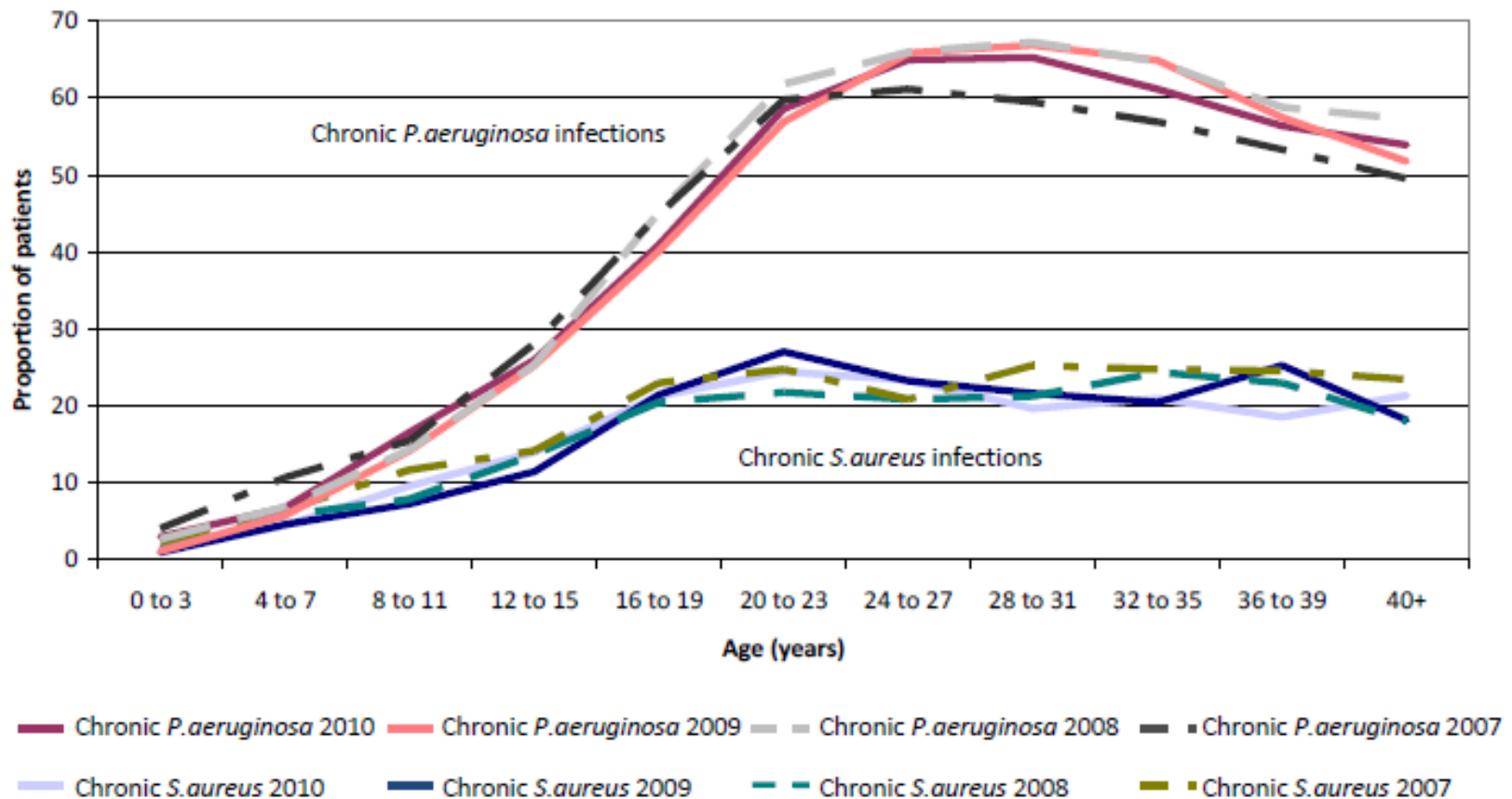


<18 year old	<18 year old	>=18 year old	>=18 year old	Total
Males	Females	Males	Females	Patients
8497	7982	6805	5811	29,095
Total <18 year olds: 16,479		Total >=18 year olds: 12,616		

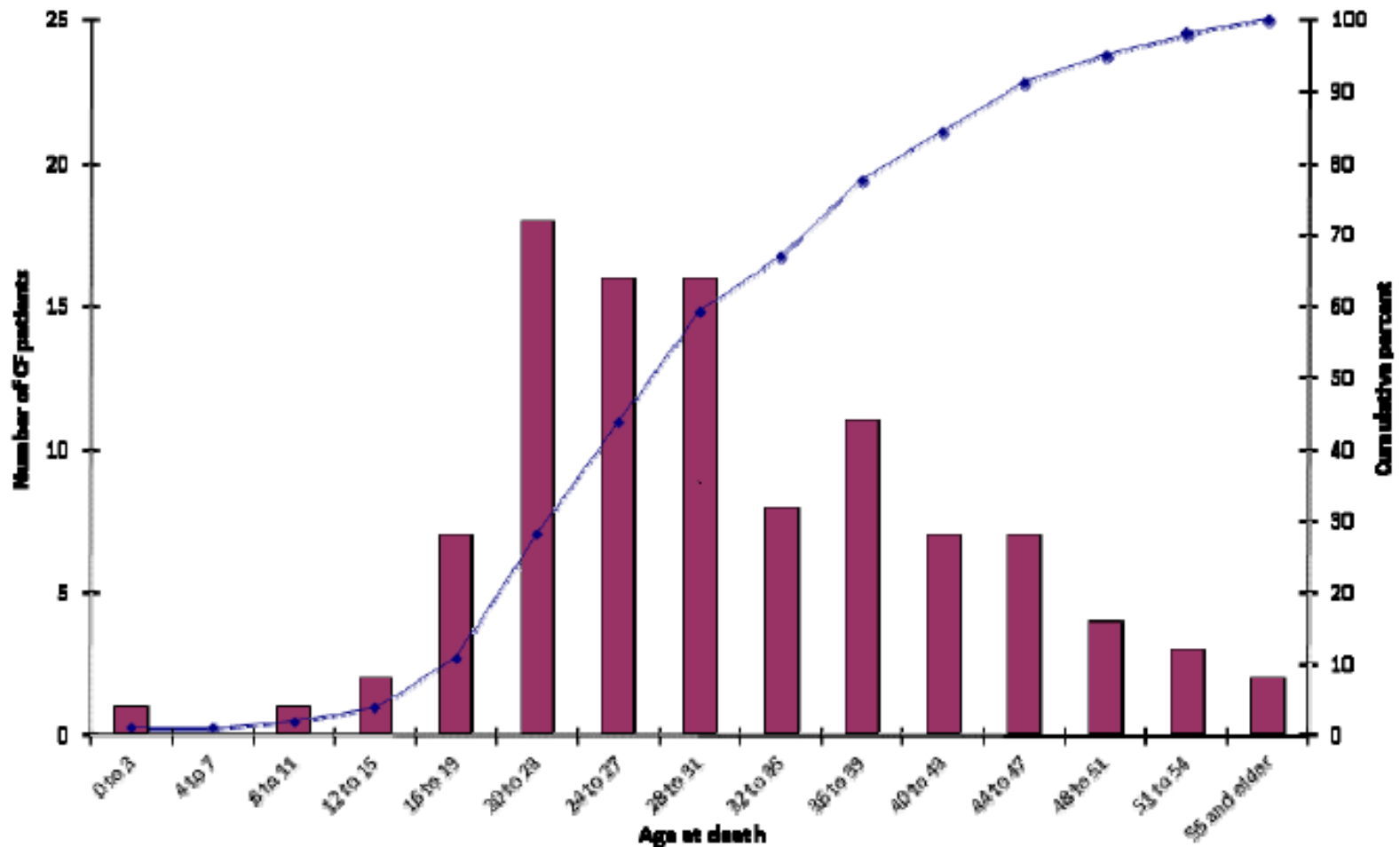
Median FEV₁ (%predicted) among patients 6yrs and older by year since 2007



Trends in lung infections since 2007



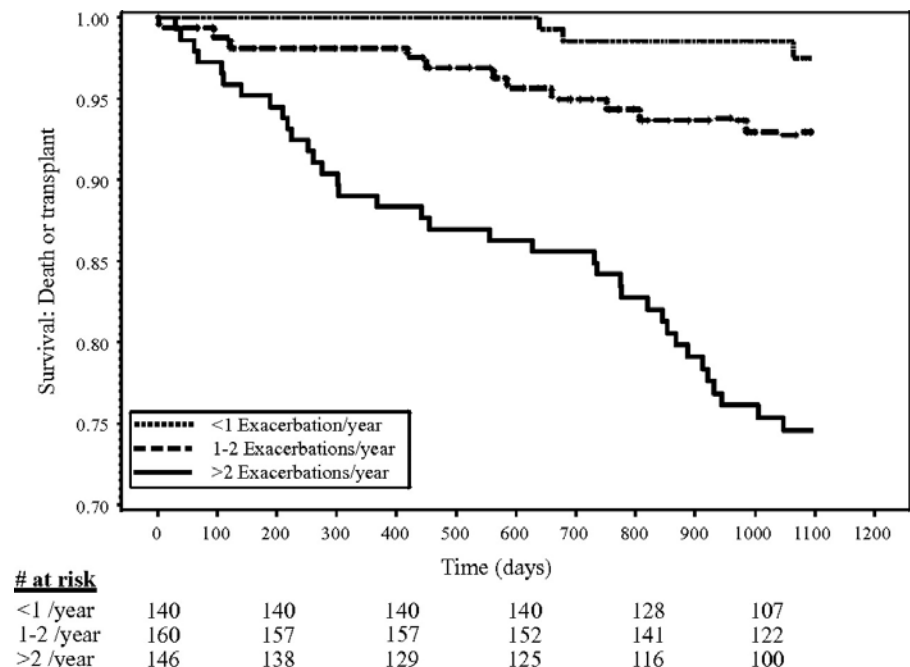
Age distribution of deaths in 2010



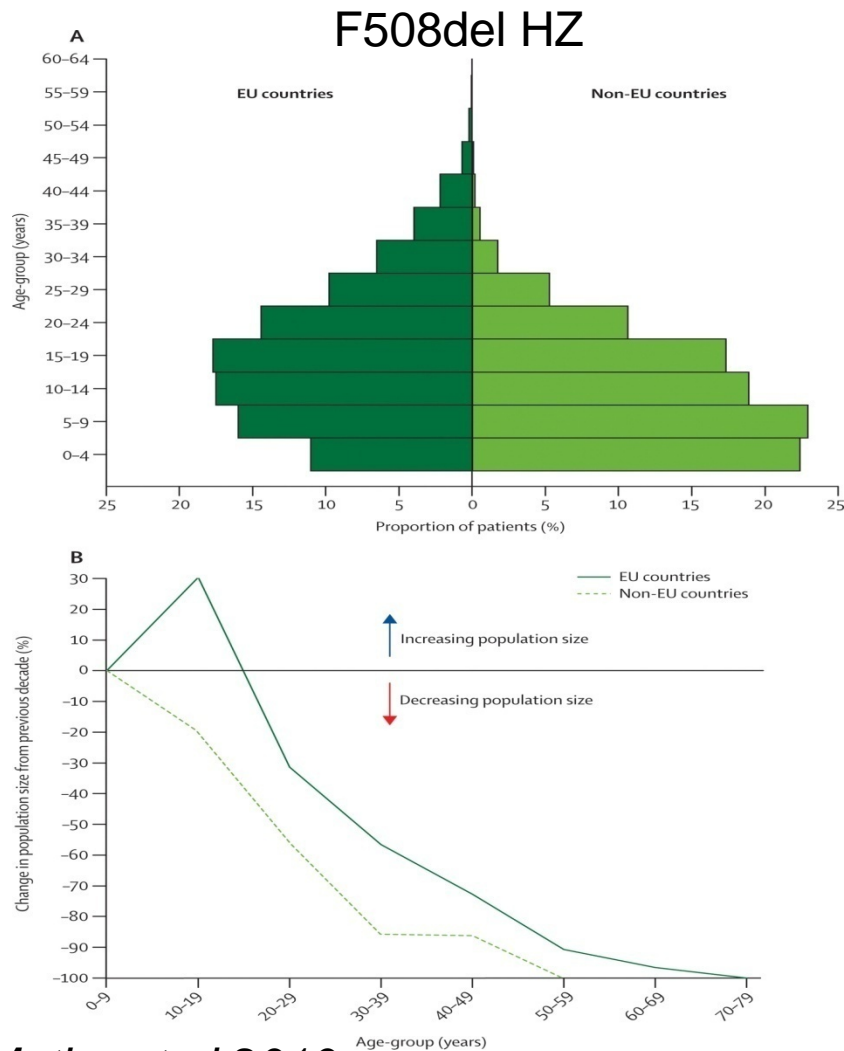
There were 103 recorded deaths in 2010. The median age at death was 29 years (min = 0 yrs; max = 61 years).

Factors associated with worse survival

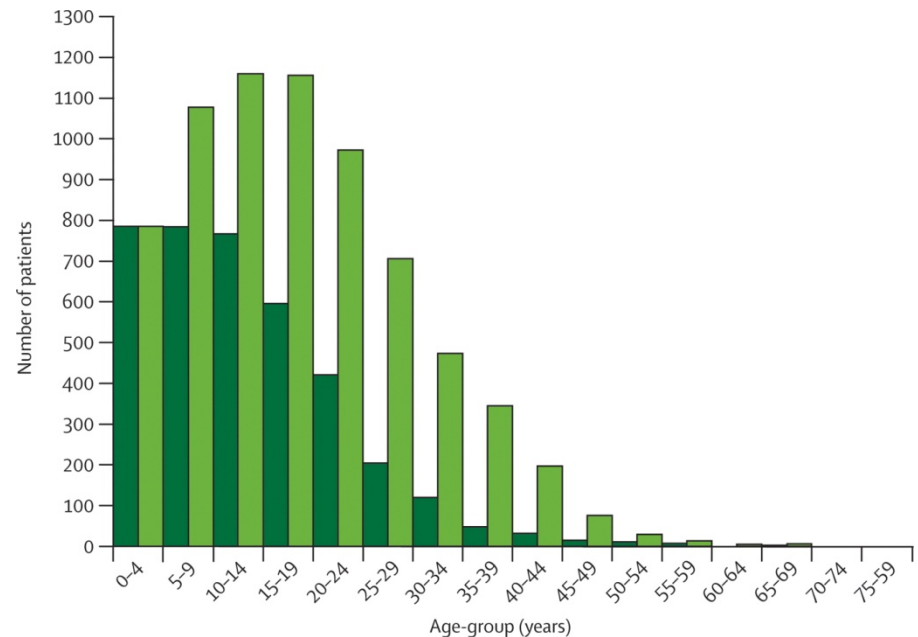
- Lung function/ FEV₁
- Exacerbation frequency
- *P aeruginosa*
- *B cenocepacia*
- Nutrition (BMI)
- CFRD
- CFLD
- Female gender

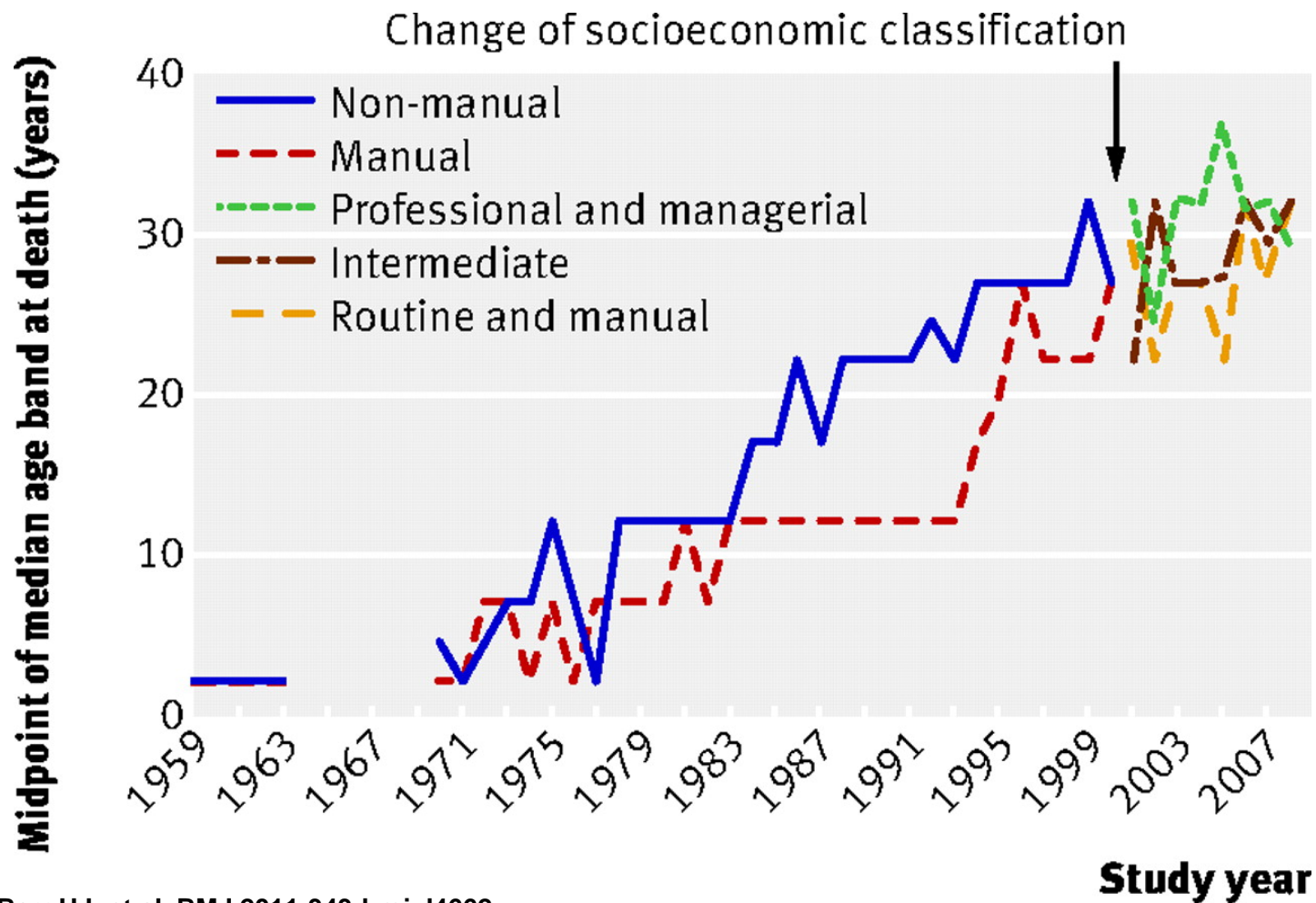


Comparison of patient numbers in EU and non EU countries



3212 patients (84%)





SOCIOECONOMIC STATUS AND RISK OF DEATH FROM CYSTIC FIBROSIS AT AGE GREATER THAN MEDIAN AGE AT DEATH

Study year	Total no of deaths	Risk of death at age greater than median age at death, by socioeconomic status				
		Manual	Non-manual*	Routine and manual	Intermediate*	Professional and managerial*
1959-63†	969	1.00	1.90 (1.35 to 2.67)	–	–	–
1970-79	1425	1.00	2.05 (1.57 to 2.69)	–	–	–
1980-89	1215	1.00	3.04 (2.26 to 4.10)	–	–	–
1990-2000	1277	1.00	2.06 (1.52 to 2.80)	–	–	–
2001-08‡	873	–	–	1.00	1.07 (0.66 to 1.72)	1.89 (1.20 to 2.97)

Data are odds ratio or adjusted odds ratio (95% confidence intervals) unless stated otherwise.

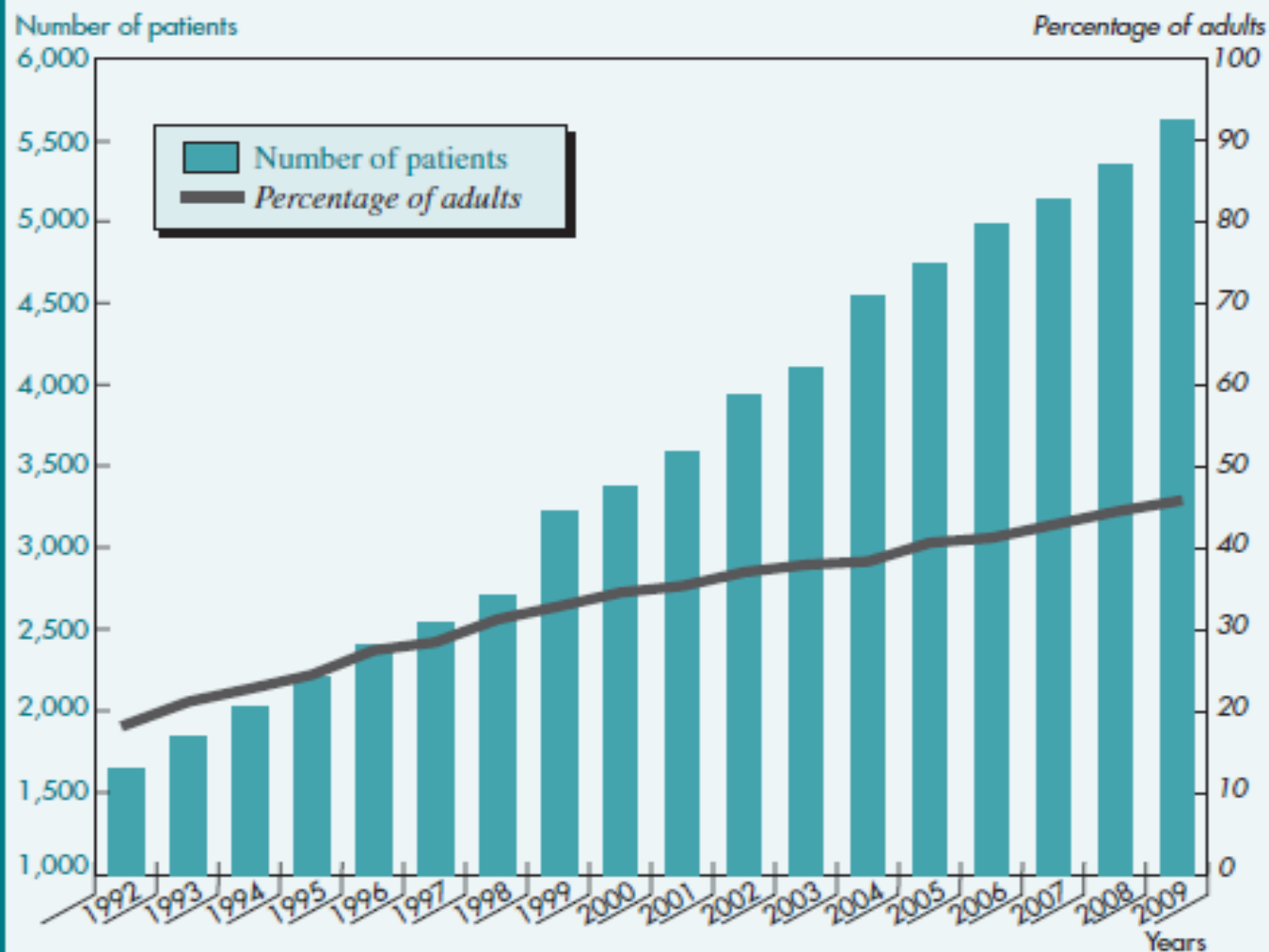
*Odds ratios adjusted for sex.

†Socioeconomic status not classified in 1964-69.

‡Socioeconomic status coding changed after 2000.

Barr H L et al. BMJ 2011;343:bmj.d4662

Figure 1 – French CF Registry 2009.
Number of patients seen during the year
and percentage of adults (a): trend since 1992

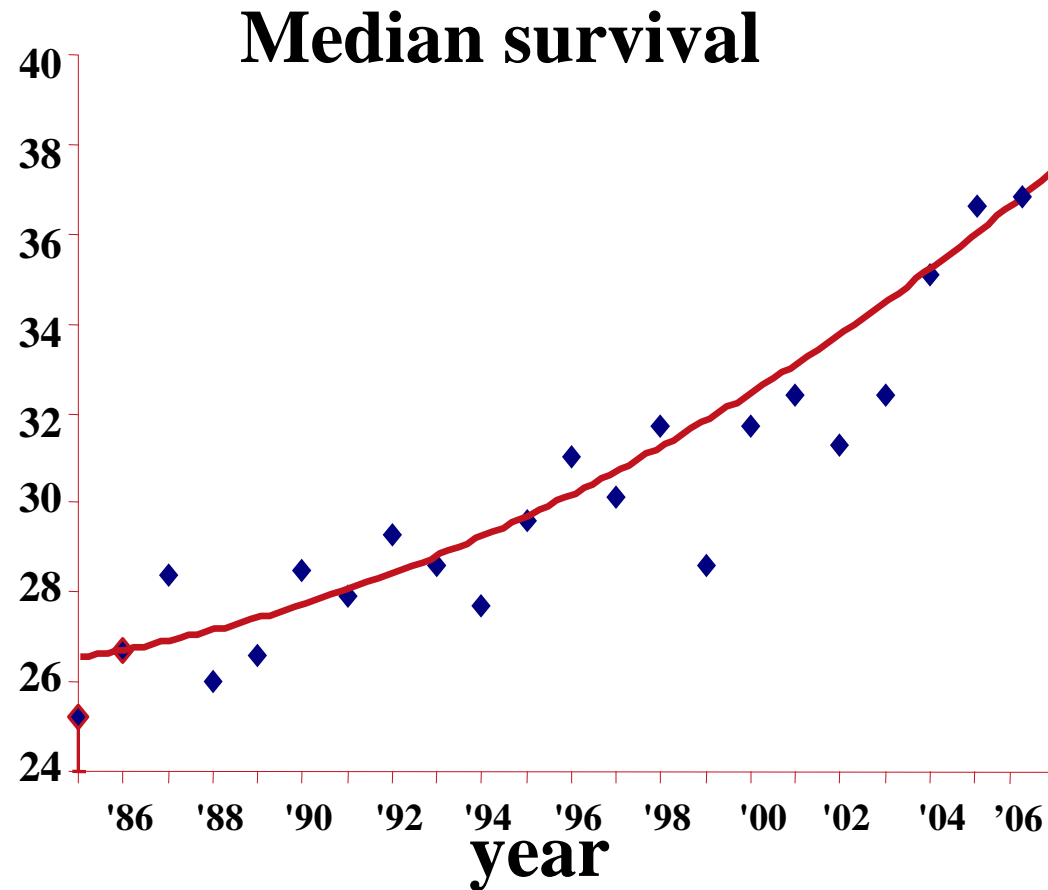


Note : (a) Patients aged 18 or above.

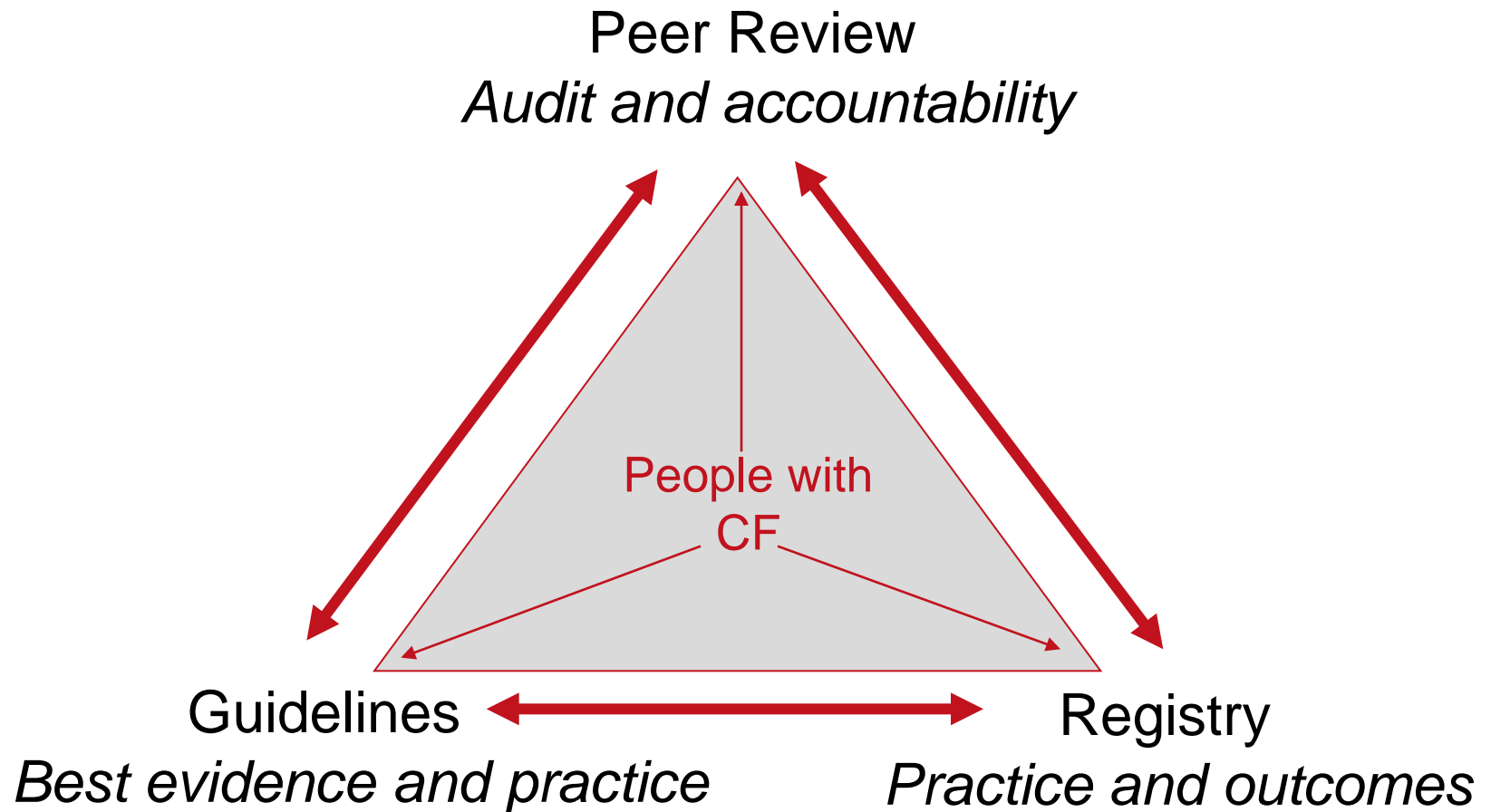
Expertise and capacity

Clinical
Expertise

- Capacity and Training
 - Europe additional 800 adults/year.
 - USA 600 additional adults/year.
- Future challenges for adult care! If we don't have the expertise we can't deliver care.



Virtuous triad of evidence based practice in Cystic Fibrosis



Summary and conclusions

- Survival in CF is increasing
- Disease factors are important: FEV₁, PEx, BMI and chronic PA
- Social and economic factors have a major impact on outcomes
- The majority of people with CF in Europe are adults
- We need more capacity to deliver adult care: adult CF teams/centres?



Clinical Care & Research in CF

