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/UtI7gr91/HU2blw# (24 April 09)

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#ixzz25IQqUCTc](http://www.dailymail.co.uk/health/article-2193518/Could-male-pin-Ben-Mudges-incredible-pack-really-key-conquering-cystic-fibrosis.html#ixzz25IQqUCTc)
Age distribution in Europe

Mehta et al 2010
Patients from Europe: 23693, US: 24487, Total: 48180
21 countries from Europe + US data shown
Data from CFF Registry, ECFS & EuroCareCF
Gender in Europe

<table>
<thead>
<tr>
<th>&lt;18 year old</th>
<th>&gt;=18 year old</th>
</tr>
</thead>
<tbody>
<tr>
<td>Males</td>
<td>Males</td>
</tr>
<tr>
<td>8497</td>
<td>6805</td>
</tr>
<tr>
<td>Females</td>
<td>Females</td>
</tr>
<tr>
<td>7982</td>
<td>5811</td>
</tr>
<tr>
<td>Total Patients</td>
<td>Total Patients</td>
</tr>
<tr>
<td>16,479</td>
<td>12,616</td>
</tr>
</tbody>
</table>
Median $\text{FEV}_1(%\text{predicted})$ 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/ FEV$_1$
- Exacerbation frequency
- *P. aeruginosa*
- *B. cepacia*
- Nutrition (BMI)
- CFRD
- CFLD
- Female gender

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de Boer K et al. Thorax 2011;66:680-685
Comparison of patient numbers in EU and non EU countries

F508del HZ

3212 patients (84%)

Metha et al 2010
### SOCIOECONOMIC STATUS AND RISK OF DEATH FROM CYSTIC FIBROSIS AT AGE GREATER THAN MEDIAN AGE AT DEATH

<table>
<thead>
<tr>
<th>Study year</th>
<th>Total no of deaths</th>
<th>Manual</th>
<th>Non-manual*</th>
<th>Routine and manual</th>
<th>Intermediate*</th>
<th>Professional and managerial*</th>
</tr>
</thead>
<tbody>
<tr>
<td>1959-63†</td>
<td>969</td>
<td>1.00</td>
<td>1.90 (1.35 to 2.67)</td>
<td>–</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>1970-79</td>
<td>1425</td>
<td>1.00</td>
<td>2.05 (1.57 to 2.69)</td>
<td>–</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>1980-89</td>
<td>1215</td>
<td>1.00</td>
<td>3.04 (2.26 to 4.10)</td>
<td>–</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>1990-2000</td>
<td>1277</td>
<td>1.00</td>
<td>2.06 (1.52 to 2.84)</td>
<td>–</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>2001-08‡</td>
<td>873</td>
<td>–</td>
<td>1.00</td>
<td>1.07 (0.66 to 1.72)</td>
<td>–</td>
<td>1.89 (1.20 to 2.97)</td>
</tr>
</tbody>
</table>

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.
Figure 1 – French CF Registry 2009. Number of patients seen during the year and percentage of adults (a): trend since 19922

Note: (a) Patients aged 18 or above.
Expertise and capacity

- 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

Peer Review
Audit and accountability

People with CF

Guidelines
Best evidence and practice

Registry
Practice and outcomes
Summary and conclusions

• Survival in CF is increasing
• Disease factors are important: FEV$_1$, 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