



UK CF Registry Annual Data Report 2007

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UK CF Registry
Annual Data Report 2007**

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**Cystic Fibrosis Trust
11 London Road
Bromley Kent
BR1 1BY**

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PREFACE



We are pleased to present the *Annual Data Report 2007* – the first from the new UK CF Registry.

The past three years have been a period of great change and significant achievement for the CF Registry in the UK. Since the last Annual Data Report for 2004, the new web-based technology of Port CF has been implemented in over 100 specialist CF centres and clinics throughout the UK for the collection of patient data for the CF Registry.

The primary purpose of the CF Registry/Port CF is to help drive up standards of clinical care throughout the UK. Port CF contributes to this objective by offering more user-friendly and flexible data entry and direct access to information, facilitating clinical audit and real time use of the patient data in each care centre.

The Annual Data Report provides a national 'snapshot' of demographic and clinical outcome data. More will be added to the Annual Data Report in future years; however, we hope you will find that the *Annual Data Report 2007* provides a firm basis for development and for monitoring improvements in outcomes over the coming years. Our intention is to produce a larger report for 2008 that will be available before the end of 2009.

A highlight from the *Annual Data Report 2007* is that the median predicted survival for 2007 is 35.2 years. This is the first time that this figure has been calculated from the CF Registry data for the UK, and represents a significant improvement over the currently quoted figure of 31 years.

The implementation of Port CF throughout the UK has required a greater amount of time, effort, and commitment than usual from the multidisciplinary teams at care centres, who have little of these resources to spare. Our thanks are due to them all for engaging with us and making the time to learn how to use Port CF and, of course, for entering the patient data. In particular, we would like to acknowledge and thank the Port CF administrators at each location who have been instrumental in this process.

Producing the *Annual Data Report 2007* would not have been possible without the contributions of many others including: the patients and families who agree to contribute their data to the Registry; our colleagues at the Department of Occupational and Environmental Medicine at the National Heart and Lung Institute, Imperial College for providing the analyses and charts; and the Registry team at the Cystic Fibrosis Trust.

Finally, we would like to acknowledge and thank the Patient Registry team at the CF Foundation in the US for their help, guidance and encouragement throughout this project.

A handwritten signature in black ink, appearing to read 'Diana Bilton'.

Dr Diana Bilton
Chair
CF Registry Steering Committee

A handwritten signature in black ink, appearing to read 'Alan Larsen'.

Alan Larsen
Director of Research and Finance
Cystic Fibrosis Trust

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Summary of the UK Cystic Fibrosis Registry

	<u>2002</u>	<u>2003</u>	<u>2004</u>	<u>2007</u>
CF patients registered	6932	6861	7046	8080 ¹
CF patients with complete data	5301	4875	5561	4408 ²
Age in years; median	17.4	16.1	16.4	18 ³
Newly diagnosed patients	159	142	164	239 ⁴
Age at diagnosis in months; median	5	5	5	5 ^{3,5}
Adults aged 16 yrs and over; %	50.1	50.8	51.4	56.7 ³
Males; %	53.9	53.8	53.4	53.9 ³
Genotyped; %	95	95	95	92.6 ³
Median predicted survival in years (95% Confidence interval)				35.2 ⁶ (31.0, 42.6)
Total deaths reported	94	103	123	106
Age at death in years; median	23	24.2	25.6	24 ⁷

In 2007, UK data were entered on the newly established PortCF system. Definitions may not be consistent between years.

Notes:

¹ In 2007, this was calculated as the number of patients on the database who satisfied the following criteria:

- were born and diagnosed with CF prior to 1 January 2008; and
- had no recorded date of death before 1 January 2007

² "Complete data" was defined as having a clinical encounter when "well" in 2007.

³ Calculated for patients with complete data.

⁴ Calculated for all patients registered. Among the 4408 patients with complete data in 2007, 83 were diagnosed in 2007.

⁵ Three patients diagnosed prenatally had age at diagnosis set to 0 months.

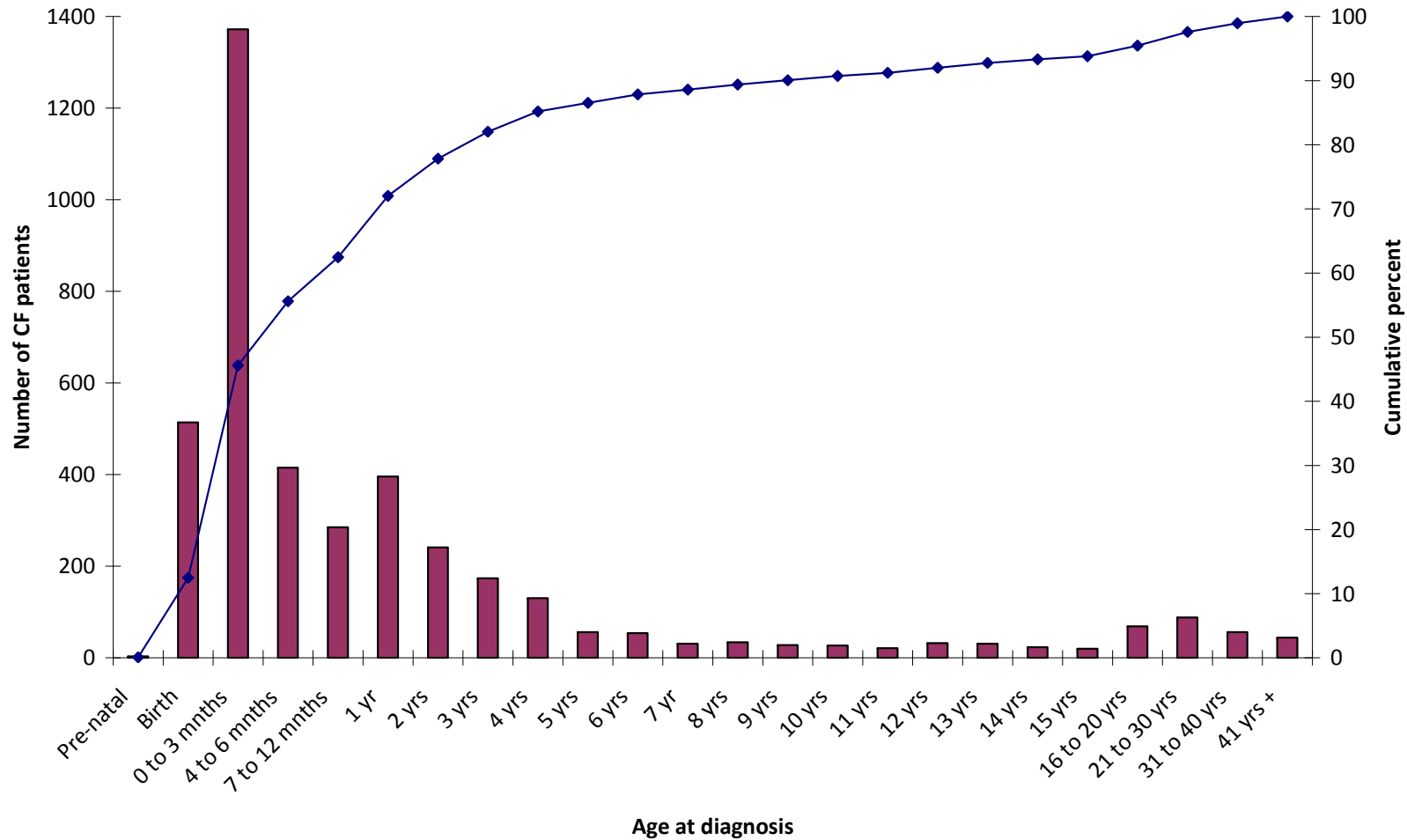
⁶ This represents the age beyond which half of the current UK CF Registry patients would be expected to live, given the ages of CF patients in the Registry and the mortality distribution of deaths in 2007.

⁷ Age at death ranged from 3 years to 61 years.

Section 1: All UK Patients

(based on 4408 patients with complete data for 2007)

1.1 Age at diagnosis



Date of diagnosis was available for 4144 of the 4408 patients with complete data. Three patients were diagnosed using prenatal screening.

1.2 Diagnosis and screening statistics

Age at diagnosis; n (%)

<i>Pre-natal</i>	3 (0.1%)
<i>Birth</i>	514 (12.4%)
<i>0-3 months</i>	1372 (33.1 %)
<i>4-6 months</i>	415 (10.0%)
<i>7-12 months</i>	285 (6.9%)
<i>1 yr</i>	396 (9.6%)
<i>2 yrs</i>	241 (5.8%)
<i>3 yrs</i>	174 (4.2%)
<i>4 yrs</i>	130 (3.1%)
<i>5 yrs</i>	56 (1.4%)
<i>6 yrs</i>	54 (1.3%)
<i>7 yrs</i>	31 (0.8%)
<i>8 yrs</i>	34 (0.8%)
<i>9 yrs</i>	28 (0.7%)
<i>10 yrs</i>	27 (0.7%)
<i>11 yrs</i>	21 (0.5%)
<i>12 yrs</i>	32 (0.8%)
<i>13 yrs</i>	31 (0.8%)
<i>14 yrs</i>	23 (0.6%)
<i>15 yrs</i>	20 (0.5%)
<i>16 – 20 yrs</i>	69 (1.7%)
<i>21 – 30 yrs</i>	88 (2.1%)
<i>31 – 40 yrs</i>	56 (1.4%)
<i>41 yrs +</i>	44 (1.1%)

The median (range) age at diagnosis is 5 months (birth – 73 yrs).

464 patients were identified using neonatal screening.

Of the 31 patients born in 2007, 21 were identified using neonatal screening.

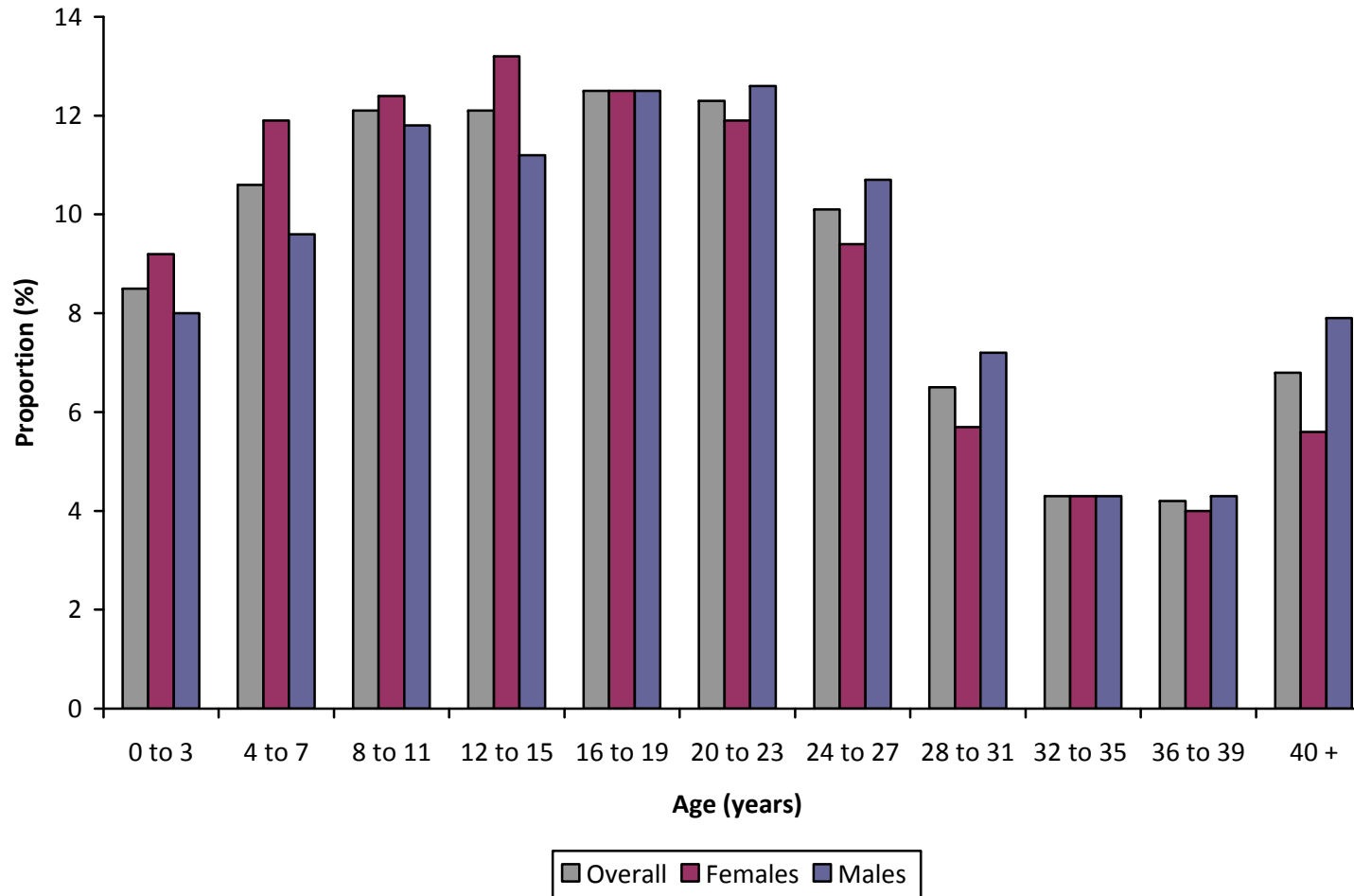
1.3 Genotyping

4081 patients have been genotyped with a recorded value

ΔF508 Mutations; n (%)

Homozygous ΔF508	2218 (54.4%)
Heterozygous ΔF508	1547 (37.9%)
No ΔF508 or both unidentified	316 (7.7%)

1.4 Age distribution

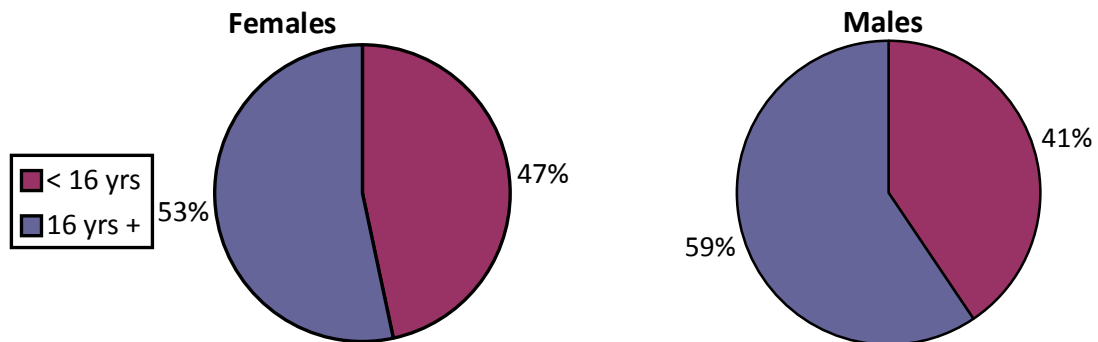


Age is calculated as the age at the clinical encounter associated with the annual review.

1.5 Age and sex distribution statistics

Age	Overall N=4408	Female N=2033	Male N=2375
0-3 yrs	375 (8.5)	186 (9.2)	189 (8.0)
4-7	469 (10.6)	242 (11.9)	227 (9.6)
8-11	532 (12.1)	251 (12.4)	281 (11.8)
12-15	534 (12.1)	268 (13.2)	266 (11.2)
16-19	552 (12.5)	255 (12.5)	297 (12.5)
20-23	540 (12.3)	242 (11.9)	298 (12.5)
24-27	445 (10.1)	191 (9.4)	254 (10.7)
28-31	286 (6.5)	116 (5.7)	170 (7.2)
32-35	191 (4.3)	88 (4.3)	103 (4.3)
36-39	184 (4.2)	81 (4.0)	103 (4.3)
40+	300 (6.8)	113 (5.6)	187 (7.9)
Median (range)	18 yrs (0 – 75 yrs)	17 yrs (0 – 69 yrs)	18 yrs (0 – 75 yrs)

1.6 Age distribution by sex



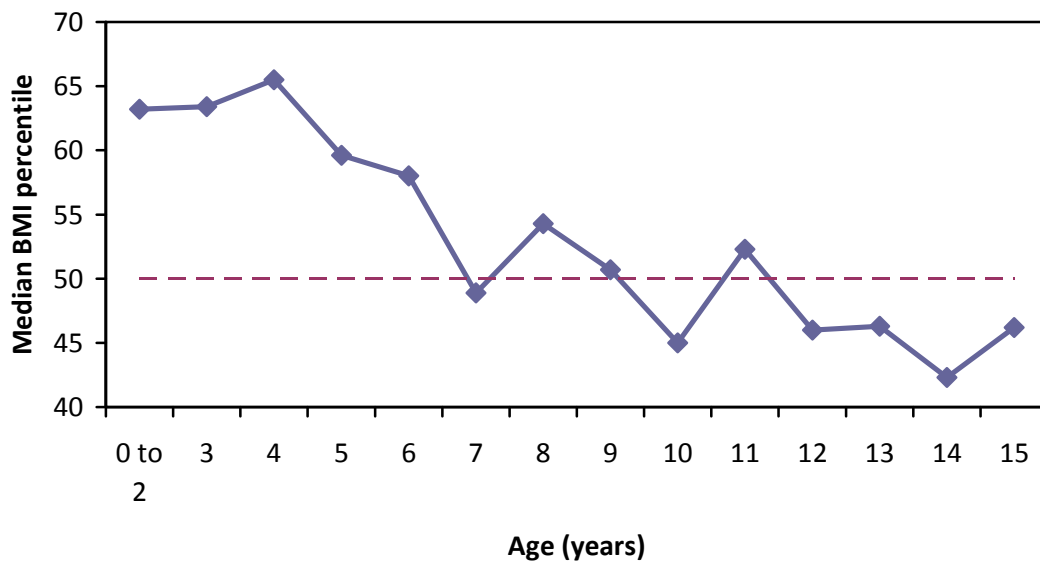
1.7 Employment status among adults aged 16yrs+

	<u>Number of patients</u>
Full-time working	745
Part-time working	266
Student	437
Homemaker	84
Unemployed	376
“Disabled”	65
Retired	23
Unknown	384

Note that these groups are not mutually exclusive and that these data were collected from the formal annual review which was not recorded for all patients.

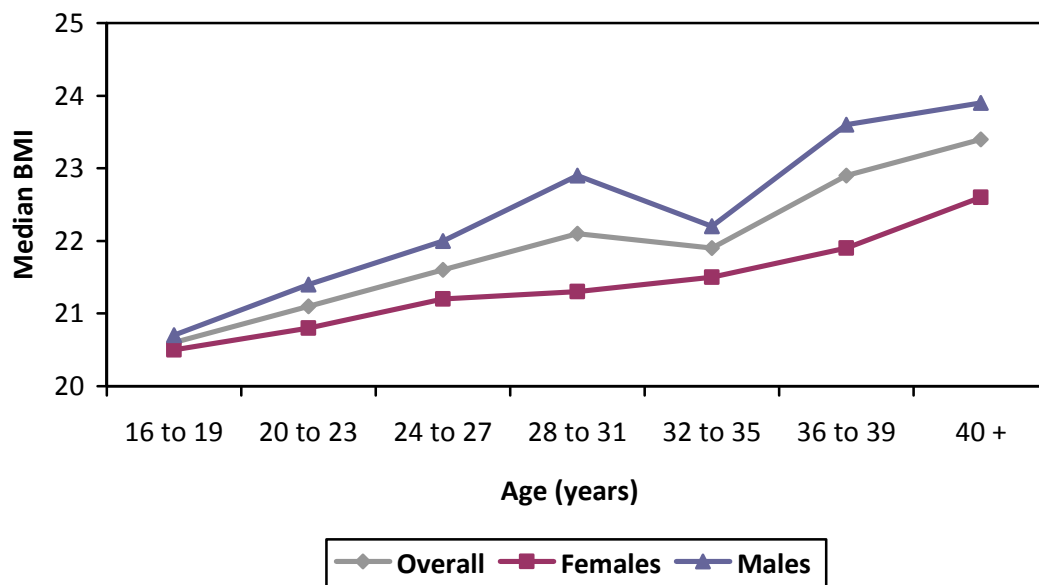
Of the 1950 adults aged 16yrs and older for whom an employment status questionnaire was completed (excluding “unknown”), 1422 (72.9%) reported being in work or study.

1.8 Median BMI percentiles among children (n=1738)



Age (years)	Number of observations	Median BMI percentile (range)
0-2	99	63.2 (1.1, 100)
3	122	63.4 (2.6, 100)
4	109	65.5 (0.1, 99.6)
5	103	59.6 (0, 98.3)
6	132	58.0 (0.7, 98.6)
7	121	48.9 (0.02, 99.3)
8	130	54.3 (0, 99.0)
9	119	50.7 (0.02, 97.8)
10	121	45.0 (3.4, 98.5)
11	154	52.3 (0.8, 98.4)
12	133	46.0 (0.4, 95.4)
13	141	46.3 (0.02, 98.8)
14	139	42.3 (2.9, 96.3)
15	115	46.2 (6.7, 96.7)
Overall	1738	53.3 (0, 100)

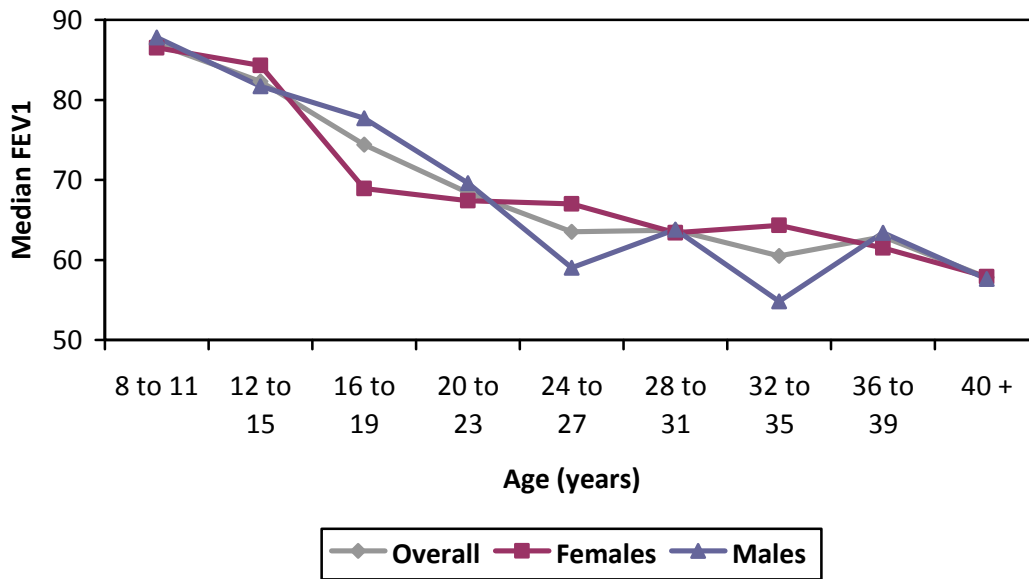
1.9 Median BMI values among adults (n=2404)



Age	Overall		Female		Male	
	N	Median (range)	N	Median (range)	N	Median (range)
16-19	531	20.6 (14.1, 35.3)	243	20.5 (14.1, 35.3)	288	20.7 (15.5, 29.6)
20-23	519	21.1 (14.1, 37.4)	233	20.8 (14.1, 30.6)	286	21.4 (15.8, 37.4)
24-27	428	21.6 (14.9, 40.8)	183	21.2 (15.7, 40.8)	245	22.0 (14.9, 31.6)
28-31	278	22.1 (16.0, 51.1)	112	21.3 (16.7, 41.0)	166	22.9 (16.0, 51.1)
32-35	182	21.9 (16.0, 35.6)	83	21.5 (16.1, 35.6)	99	22.2 (16.0, 31.3)
36-39	177	22.9 (14.4, 41.0)	76	21.9 (14.4, 31.1)	101	23.6 (15.9, 41.0)
40+	289	23.4 (16.6, 39.4)	110	22.6 (16.6, 33.4)	179	23.9 (16.8, 39.4)
Overall	2404	21.6 (14.1, 51.1)	1040	21.2 (14.1, 41.0)	1364	22.0 (14.9, 51.1)

N refers to the number of observations in each age/sex category

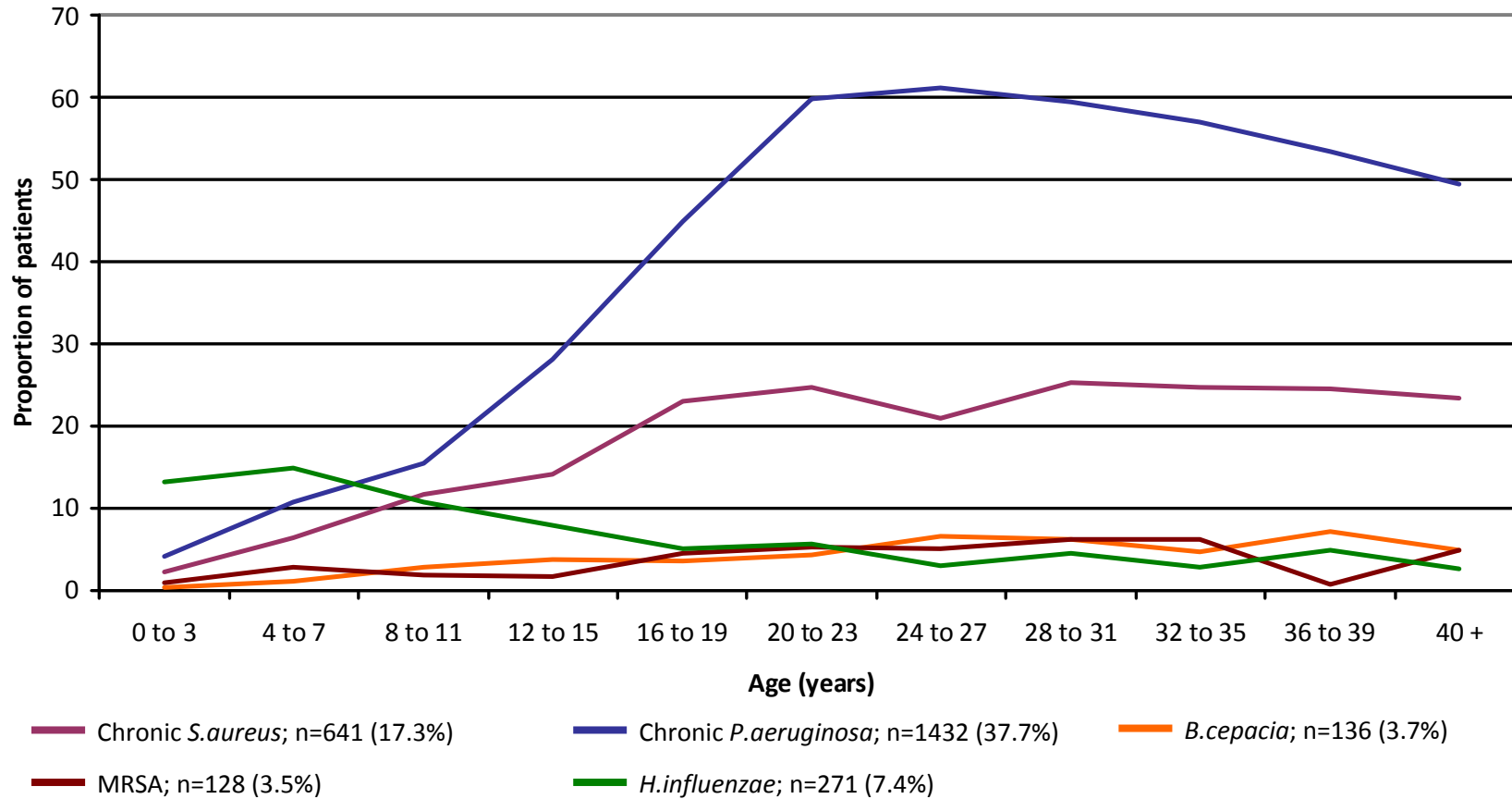
1.10 Median FEV₁ (% predicted) among patients aged 8 years and older (n=3404)



Age	Overall		Female		Male	
	N	Median (range)	N	Median (range)	N	Median (range)
8-11	501	87.0 (19.6, 140.8)	239	86.5 (19.6, 140.8)	262	87.8 (35.0, 121.1)
12-15	512	82.3 (12.2, 130.9)	258	84.3 (20.7, 130.1)	254	81.7 (12.2, 130.9)
16-19	523	74.4 (15.8, 186.1)	242	68.9 (15.8, 186.1)	281	77.7 (22.7, 121.2)
20-23	520	68.4 (15.4, 173.2)	233	67.4 (15.4, 173.2)	287	69.5 (17.0, 139.3)
24-27	423	63.5 (12.6, 145.2)	179	67.0 (18.5, 145.2)	244	59.0 (12.6, 133.7)
28-31	277	63.7 (12.5, 163.2)	111	63.4 (12.5, 163.2)	166	63.8 (14.6, 114.6)
32-35	181	60.5 (19.1, 137.9)	85	64.3 (20.2, 137.9)	96	54.8 (19.1, 114.2)
36-39	178	62.9 (16.0, 160.1)	77	61.5 (20.7, 160.1)	101	63.4 (16.0, 120.8)
40+	289	57.8 (13.5, 143.7)	110	57.9 (21.9, 143.7)	179	57.6 (13.5, 134.4)
Overall	3404	73.2 (12.2, 186.1)	1534	72.9 (12.5, 186.1)	1870	73.3 (12.2, 139.3)

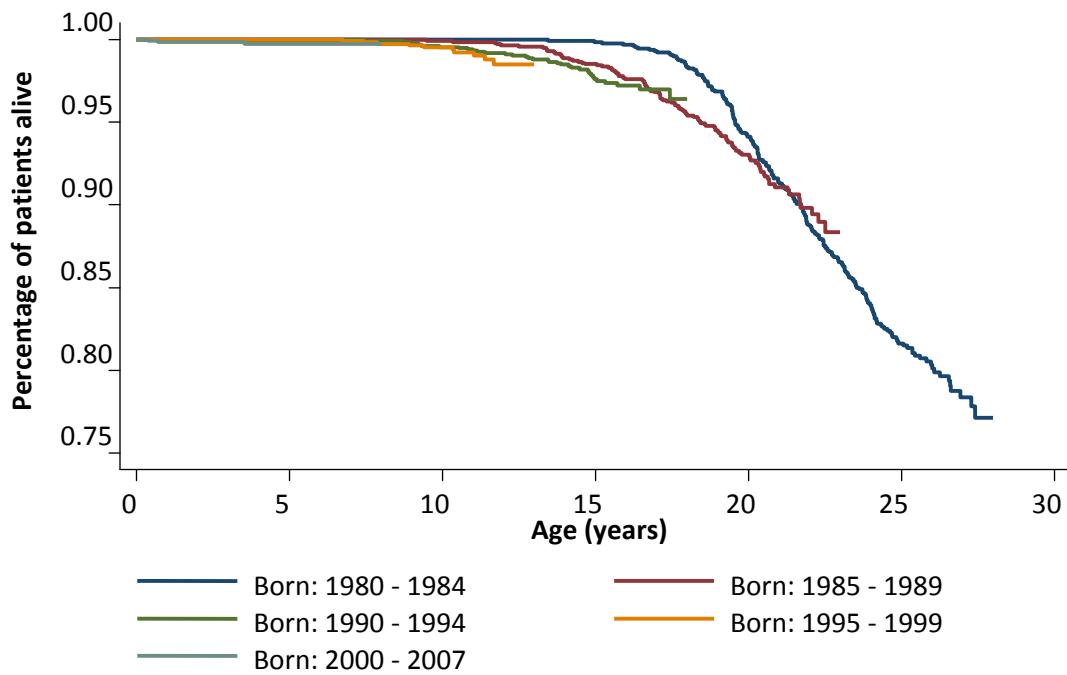
N refers to the number of observations in each age/sex category

1.11 Infections



Chronic infection with *S. aureus* or *P. aeruginosa* was identified from annual review. Data on *B. cepacia*, MRSA and *H. influenzae* were collected from culture results at the clinical encounter associated with the annual review.

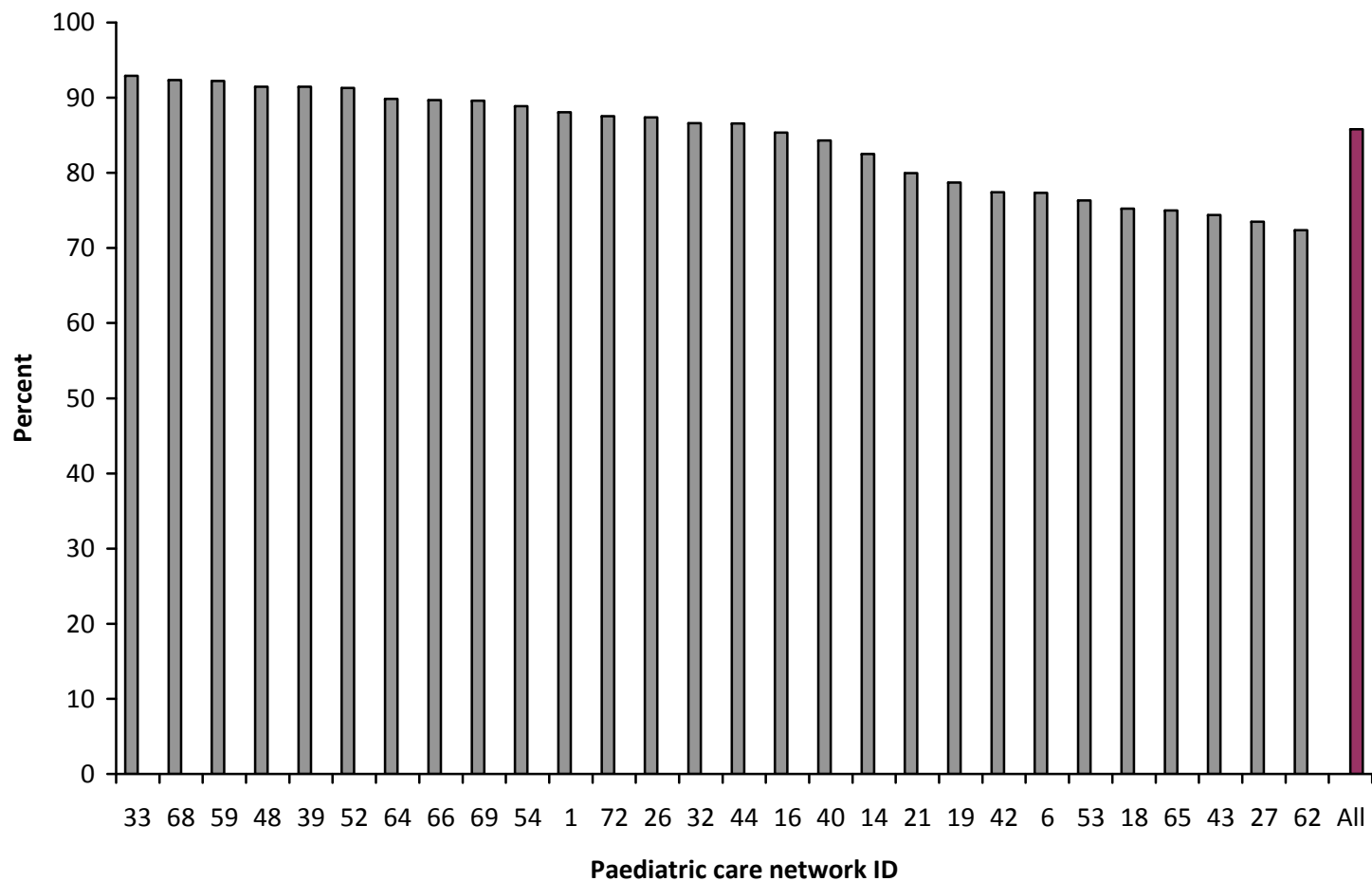
11.12 Actuarial survival by birth cohort



Actuarial survival from birth of all patients on the UK CF Registry in 2007, regardless of whether they have complete data. These findings may not represent all patients with CF during these periods.

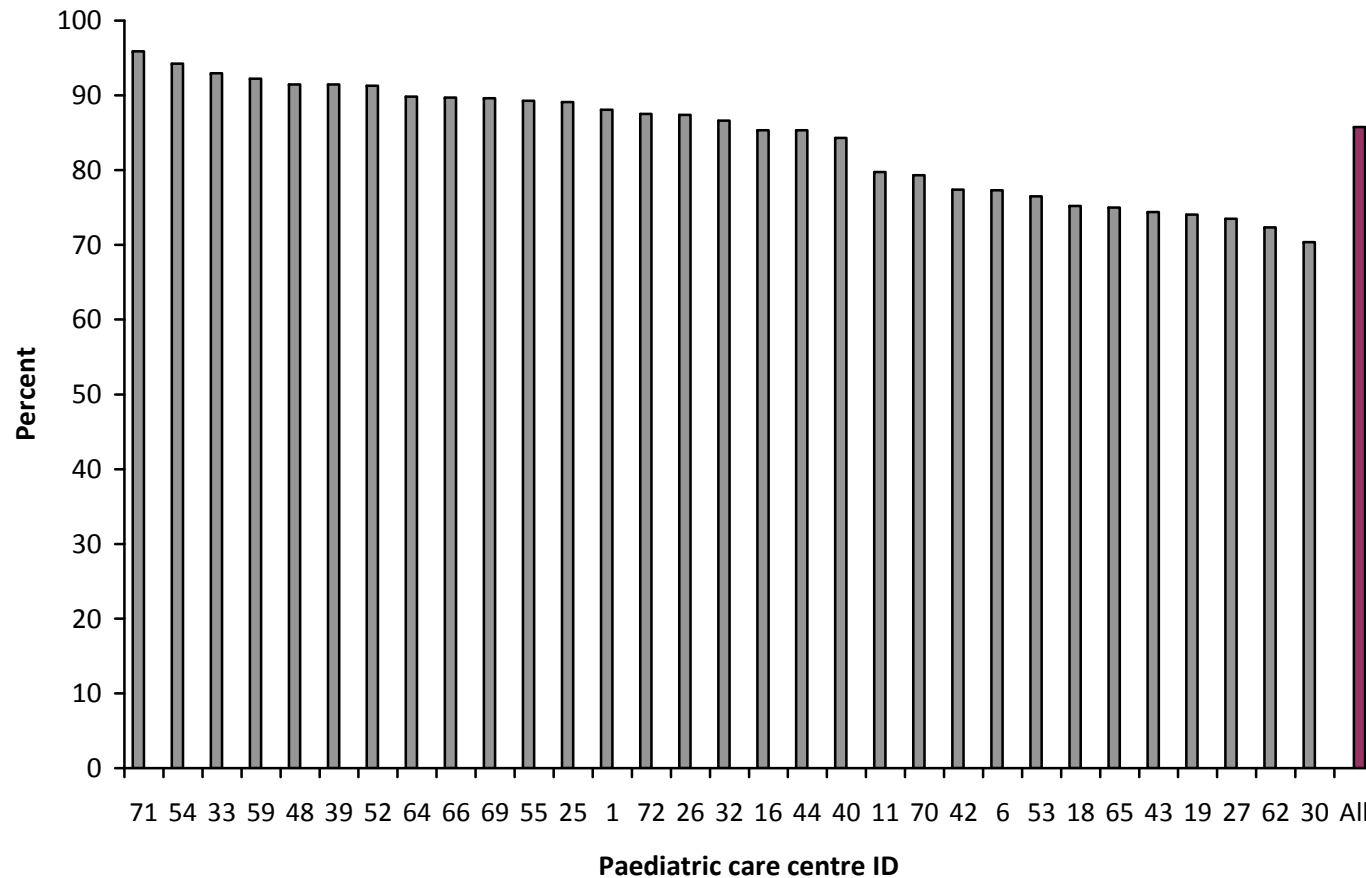
Section 2: Analyses by care centre **(based on 4408 patients with complete data in 2007)**

2.1 Median FEV₁ (% predicted) by paediatric care network or stand-alone paediatric centre/clinic



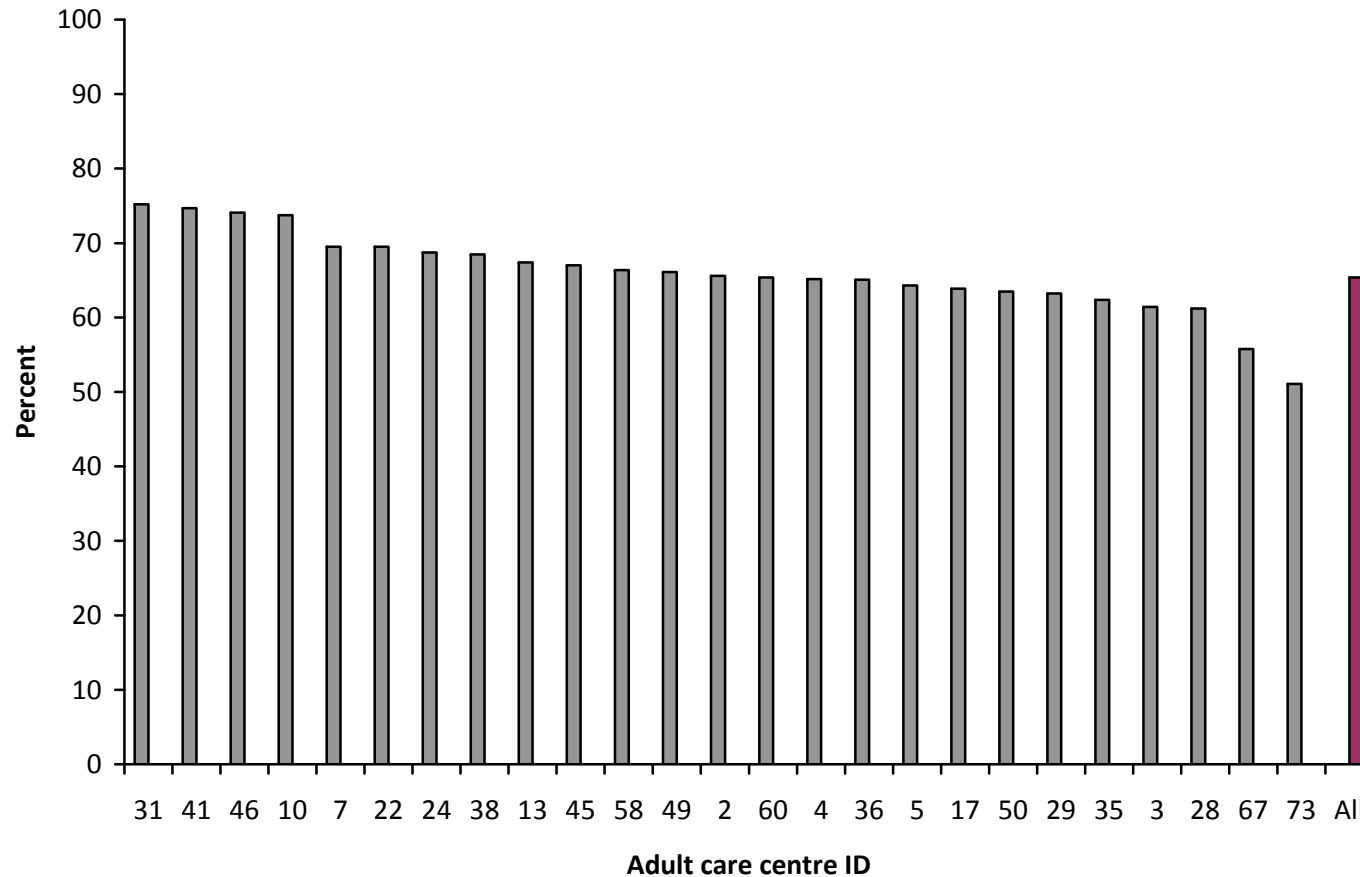
The median FEV₁ (% predicted) for paediatric care networks and stand-alone paediatric centres/clinics is 86.6% (min=72.3%, max=92.9%).

2.2 Median FEV₁ (% predicted) by paediatric centre/clinic with at least 20 patients at annual review



The median FEV₁ (% predicted) for paediatric care centres/clinics with at least 20 patients is 86.6% (min=70.3%, max=95.9%).

2.3 Median FEV₁ (% predicted) by adult centre/clinic with at least 20 patients at annual review



The median FEV₁ (% predicted) for adult care centres with at least 20 patients is 65.6% (min=51.1%, max=75.2%).

Section 3: Acknowledgements

3.1 Paediatric care centres/clinics providing data in 2007

England

Addenbrookes Hospital, Cambridge
Hinchingsbrooke Hospital, Huntingdon
West Suffolk General Hospital, Bury St Edmunds
Alder Hey Children's Hospital, Liverpool
Warrington District General Hospital
Royal Albert Edward Infirmary, Wigan
Birmingham Children's Hospital
Walsgrave Hospital, Coventry
New Cross Hospital, Wolverhampton
Booth Hall Children's Hospital, Manchester
Bristol Royal Hospital for Children
Derriford Hospital, Plymouth
Great Ormond Street Hospital for Sick Children, London
Hull Royal Infirmary
John Radcliffe Hospital, Oxford
King's College Hospital, London
Leicester Royal Infirmary
Nottingham City Hospital
Pilgrim Hospital, Boston
Chesterfield Hospital
Derby Hospital
Kings Mill Hospital, Sutton-In-Ashfield
Lincoln Hospital
Queen Mary's Hospital for Children, Carshalton
Royal Cornwall Hospital, Truro
Royal Devon & Exeter Hospital
Royal Brompton Hospital, London
Royal London Hospital
Royal Victoria Infirmary, Newcastle
Bishop Auckland General Hospital
West Cumberland Hospital, Whitehaven
Sunderland Royal Hospital
University Hospital Lewisham, London
St James's University Hospital, Leeds
Sheffield Children's Hospital
Southampton General Hospital
University Hospital of North Staffordshire, Stoke-on-Trent

Scotland

Crosshouse Hospital, Ayr
Ninewells Hospital, Dundee
Raigmore Hospital, Inverness
Royal Aberdeen Children's Hospital
Royal Hospital for Sick Children, Edinburgh
Royal Hospital for Sick Children, Glasgow

Indented hospitals are associated network clinics

3.1 Paediatric care centres/clinics providing data in 2007 (continued)

Northern Ireland

Royal Belfast Hospital for Sick Children

Wales

Children's Hospital for Wales, Cardiff

Bronglais Hospital, Aberystwyth

Hereford County Hospital

Nevill Hall Hospital, Abergavenny

Princess of Wales Hospital, Bridgend

Royal Glamorgan Hospital, Llantrisant

Singleton Hospital, Swansea

West Wales General Hospital, Carmarthen

Withybush General Hospital, Haverfordwest

Indented hospitals are associated network clinics

3.2 Adult care centres/clinics providing data in 2007

England

Birmingham Heartlands Hospital
New Cross Hospital, Wolverhampton
Bristol Royal Infirmary
Churchill Hospital, Oxford
Derriford Hospital, Plymouth
Frimley Park Hospital, Camberley
Glenfield Hospital, Leicester
King's College Hospital, London
Liverpool Heart and Chest Hospital
London Chest Hospital
Norfolk & Norwich University Hospital, Norwich
Northern General Hospital, Sheffield
Nottingham City Hospital
Papworth Hospital
Royal Brompton Hospital, London
Royal Cornwall Hospital, Truro
Royal Devon & Exeter Hospital
Royal Victoria Infirmary, Newcastle
St James's University Hospital, Leeds
Southampton General Hospital
Poole Hospital
University Hospital Lewisham, London
University Hospital of North Staffordshire, Stoke-on-Trent
Wythenshawe Hospital, Manchester

Northern Ireland

Belfast City Hospital

Scotland

Aberdeen Royal Infirmary
Gartnavel General Hospital, Glasgow
Western General Hospital, Edinburgh
Dumfries and Galloway Royal Infirmary, Dumfries
Ninewells Hospital, Dundee
Raigmore Hospital, Inverness

Wales

Llandough Hospital

Indented hospitals are associated outreach clinics

3.3 UK CF Registry Steering Committee

Dr Diana Bilton (<i>Chair</i>)	Consultant Physician, Adult CF Unit Royal Brompton Hospital, London
Mrs Rosie Barnes	Chief Executive, Cystic Fibrosis Trust
Dr Keith Brownlee	Consultant Paediatrician, Regional CF Unit St James's University Hospital, Leeds
Ms Diana Cargill	Senior Commissioner South West Specialist Commissioning Group
Dr Geoffrey Carroll	Medical Director, Health Commission Wales
Ms Kathy Collins	Nursing and Quality Adviser National Services Division, Scotland
Mrs Marian Dmochowska	Parent Representative
Dr Iolo Doull	Consultant Paediatrician, Paediatric CF Unit Children's Hospital for Wales, Cardiff
Professor Stuart Elborn	Consultant Physician, Adult CF Unit Belfast City Hospital
Dr Caroline Elston	Consultant Physician, Adult CF Unit King's College Hospital, London
Mr Alan Larsen	Director of Research and Finance, Cystic Fibrosis Trust
Dr Jim Littlewood	Chairman Cystic Fibrosis Trust Retired Consultant Paediatrician
Ms Sue McLellen	Head of Specialised Commissioning, London Specialised Commissioning Group
Dr Sarah Walters	Patient Representative Epidemiologist, Birmingham University
Ms Jacquie Westwood	Director of Specialised Services, South East London
Dr Martin Wildman	Consultant Physician, Adult CF Centre Northern General Hospital, Sheffield

3.4 Data analysis

Data analysis was performed by:

Paul Cullinan MD, FRCP – Professor in Occupational and Environmental Respiratory Disease

Stephanie MacNeil PhD – Medical Statistician

Department of Occupational and Environmental Medicine

National Heart and Lung Institute

Imperial College

London