



CYSTIC FIBROSIS IN AUSTRALIA 2006

**Annual Report from the Australian
Cystic Fibrosis Data Registry**

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Preface

I am pleased to present the ninth annual report from the Australian Cystic Fibrosis Data Registry.

Once again I offer my thanks to all of the people in CF treatment centres who work hard at providing the information for the registry. CFA, and users of the new reporting features of the Data Registry, are especially grateful for their hard work.

During the past year CFA has continued to consult the Data Registry Advisory Committee and has also engaged the broader community of CF Specialist Centre Directors in discussions about the future directions for the Australian registry. A major item for discussion during the year has been investigation of the option of adopting the PortCF system that is used by the Cystic Fibrosis Foundation for the United States CF patient registry. While the existing PortCF system has been implemented in the United Kingdom, and has been investigated by other national and international registries, further development of PortCF has opened up new options for harmonisation and sharing of CF registry data internationally, without necessarily abandoning current national registry systems. Developments are in the early stages and will be managed in consultation with the Advisory Committee.

Although there continues to be some under-reporting of data for this report, it is pleasing that, since the launch of the online data registry, centres that had not been contributors, and some whose data entry had been interrupted for some years, have (re)commenced contributing data. The fruits of this will be seen in even better coverage of the registry in future reporting years.

Once again I invite all in the Australian cystic fibrosis community to make maximum use of the Data Registry for the benefit of people with CF. Further data analysis can be done on request and applications from researchers to use registry data are most welcome.

Terry Stewart
Chief Executive Officer
Cystic Fibrosis Australia

May 2009

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Many thanks go to the Australasian Cystic Fibrosis Data Registry Advisory Committee, whose members have shown a commitment to making the Data Registry a useful tool for improving health and standards in the CF community. Members are:

Dr Scott Bell – The Prince Charles Hospital, Brisbane QLD
Ms Zez Stankovic – National Data Registry Coordinator, CFA
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LJ Hooker Cystic Fibrosis National Sponsor for Research

Participating Centres

The Data Registry relies on the tireless work of people in the following CF Centres who enter data and handle edit queries for quality control of the annual collection of data:

New South Wales

Sydney Children's Hospital
Royal Prince Alfred Hospital, Sydney
The Children's Hospital, Westmead
Westmead Hospital - Adults
Gosford Hospital
John Hunter Hospital - Newcastle

Victoria

Royal Children's Hospital, Melbourne
The Alfred Hospital, Melbourne
Monash Medical Centre, Clayton

Queensland

Royal Children's Hospital, Brisbane
The Prince Charles Hospital, Brisbane
Mater Hospital - Children, Brisbane
Mater Hospital - Adults, Brisbane
Southport Hospital - Children, Gold Coast
Southport Hospital - Adults, Gold Coast

South Australia

Royal Adelaide Hospital
Women's and Children's Hospital, Adelaide

Western Australia

Princess Margaret Hospital for Children, Perth
Sir Charles Gairdner Hospital, Perth

Tasmania

Royal Hobart Hospital
Launceston General Hospital
CF Clinic, Burnie

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1 People with cystic fibrosis

1.1 Overview

At 31 December 2006 the Australian Cystic Fibrosis Data Registry (ACFDR) held records of 2467 people with cystic fibrosis.

The average age of the Registry population was 17.8 years at 31 December 2006. This is up from 17.2 years at the end of 2005 and 16.9 years in 2004. The proportion of the registry population that is adult (18 years and over) increased to 44 percent by the end of 2006. The proportion of adults recorded in the registry was 27 per cent in 1998, the year that the Registry first reported.

Within the Registry population, the median age at 31 December 2006 was 16.0 years, an increase from 15.3 at the end of 2005. The median age for males (17 years) remained higher than that for females (15.5 years).

The number of new diagnoses of CF notified to the Registry for 2006 was 63, including 53 diagnosed at age less than one year. This is similar than the 63 overall and 55 infant diagnoses reported in 2005.

The number of deaths reported to the National Mortality Database decreased from 39 in 2005 to 24 in 2006, less than half of the 60 deaths reported in 1998.

Overall, the coverage of demographic information about persons with CF is high for 2006. While all specialist CF centres in Australia have agreed to participate in the Data Registry, reporting from a small number of adult centres had not begun in 2006, leaving a gap in coverage that has been estimated at about 80 adults.

Individuals who do not attend specialist CF treatment centres would not be covered by statistics in this report. The number of such persons is unknown but not expected to be large.

Tables reflecting personal information about patients, presented in Sections 1.3 and 1.4, are affected by non-reporting for a high proportion of adult patients. Missing data are generally the for the complete patient population of certain centres that have failed to report this information.

1.2 Age distribution

Figure 1.1 shows the age distribution of Australian residents recorded in the CF Data Registry at 31 December 2006. Details are in Table 1.1.

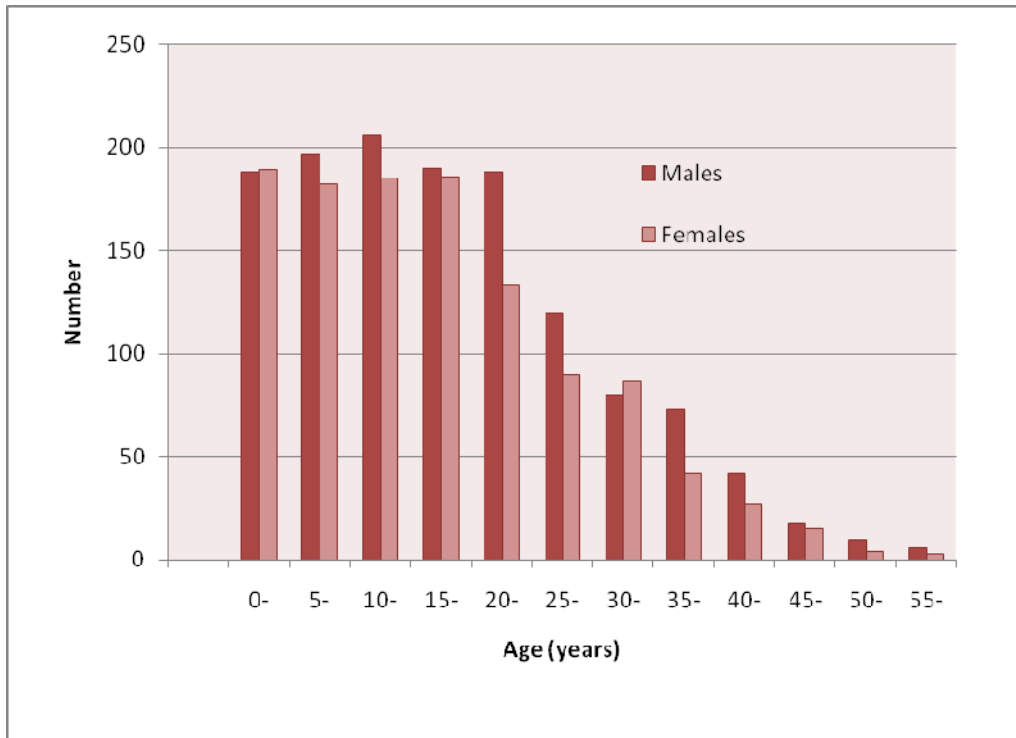


Figure 1.1: ACFDR 2006: Age distribution by sex

Table 1.1: ACFDR 2006: Age and sex of registrants at 31 December 2006

Age	Males	Females	Persons	Per cent male
0	38	34	72	
1	42	44	86	
2	27	37	64	
3	32	44	76	
4	49	30	79	
<i>0 - 4 years</i>	188	189	377	49.9
5	38	34	72	
6	55	38	93	
7	27	37	64	
8	40	35	75	
9	37	38	75	
<i>5 - 9 years</i>	197	182	379	52.0
10	42	38	80	
11	33	34	67	
12	41	37	78	
13	43	29	72	
14	47	47	94	
<i>10 - 14 years</i>	206	185	391	52.7
15	37	47	84	
16	34	39	73	
17	41	41	82	
18	49	27	76	
19	29	32	61	
<i>15 - 19 years</i>	190	186	376	50.5
20	42	23	65	
21	42	29	71	
22	36	27	63	
23	41	32	73	
24	27	22	49	
<i>20 - 24 years</i>	188	133	321	58.6
25	32	25	57	
26	23	12	35	
27	27	18	45	
28	20	19	39	
29	18	16	34	
<i>25 - 29 years</i>	120	90	210	57.1
30	14	22	36	
31	16	25	41	
32	20	17	37	
33	22	13	35	
34	8	10	18	
<i>30 - 34 years</i>	80	87	167	47.9
<i>35 - 39 years</i>	73	42	115	63.5
<i>40 - 44 years</i>	42	27	69	60.9
<i>45 - 49 years</i>	18	15	33	54.5
<i>50 - 54 years</i>	10	4	14	71.4
<i>55 - 59 years</i>	6	3	9	66.7
<i>60 + years</i>	0	6	6	0.0
Total	1318	1149	2467	53.4

The proportion of adults in the Registry as a whole was 44 per cent at 31 December 2006. In most jurisdictions the proportion is between 40 and 50 per cent. Lower proportions in New South Wales and the Australian Capital Territory reflect known gaps that remained in 2006 in reporting from centres treating adult patients.

Table 1.2: ACFDR 31 December 2006: Adult status by State/Territory of residence

State or Territory of residence	Child/adolescent	Adult	Total	Per cent adult
New South Wales	457	296	753	39.3
Victoria	264	249	513	48.5
Queensland	314	249	563	44.2
Western Australia	159	138	297	46.5
South Australia	151	108	259	41.7
Tasmania	28	37	65	56.9
Australian Capital Territory	7	1	8	12.5
Northern Territory	3	3	6	50.0
Overseas	3	0	3	0.0
<i>Total</i>	<i>1,386</i>	<i>1,081</i>	<i>2,467</i>	<i>43.8</i>

At 31 December 2006, males made up 53.4% and females 46.6% of the Australian Registry population. This has remained a consistent proportion since establishment of the Registry in 1998.

Table 1.3: ACFDR 31 December 2006: Sex distribution by State/Territory of residence

Place of residence	Males	Females	Persons	Per cent male
New South Wales	379	374	753	50.3
Victoria	292	221	513	56.9
Queensland	295	268	563	52.4
Western Australia	160	137	297	53.9
South Australia	147	112	259	56.8
Tasmania	40	25	65	61.5
Australian Capital Territory	3	5	8	37.5
Northern Territory	1	5	6	16.7
Overseas	1	2	3	33.3
<i>Total</i>	<i>1,318</i>	<i>1,149</i>	<i>2,467</i>	<i>53.4</i>

1.3 Siblings with cystic fibrosis

Around one in 6 persons with cystic fibrosis reported having siblings who also have the disease. (Table 1.4)

Table 1.4 ACFDR 31 December 2006: Patients who have siblings with cystic fibrosis

State or Territory of residence:	Yes	No	Unknown	Not reported	Total	Per cent with CF sibling(a)
New South Wales	84	389	0	280	753	17.8
Victoria	42	238	1	232	513	15.0
Queensland	67	324	14	158	563	17.1
Western Australia	48	221	5	23	297	17.8
South Australia	31	129	2	97	259	19.4
Tasmania	14	40	3	8	65	25.9
Australian Capital Territory	2	6	0	0	8	25.0
Northern Territory	1	5	0	0	6	16.7
Overseas	0	3	0	0	3	0.0
<i>Total</i>	<i>289</i>	<i>1,355</i>	<i>25</i>	<i>798</i>	<i>2,467</i>	<i>17.6</i>

(a) excludes not reported from total

1.4 Adults: marital status, education and activity

Management of cystic fibrosis allows an increasing proportion of persons with the disease to continue with normal activities into their adult life, despite the degenerative effects of the disease. Personal information is recorded for 68% of the CF patient population. While reporting of current personal information is incomplete, gaps in coverage largely reflect lack of information from a small number of treatment centres. Information compiled below therefore reflects largely complete information from a subset of CF treatment centres.

Table 1.5 shows that around 37% of adult patients for whom marital status is known were in a formal or informal marriage relationship.

Table 1.5: ACFDR 31 December 2006: Marital status of adults

Marital status	Males		Females	
	Number	Per cent	Number	Per cent
Married (includes de facto)	103	31.2	111	44.8
Not married	227	68.8	137	55.2
		100.0		100.0
Unknown (includes not recorded)	285	46.3	218	46.8
<i>Total</i>	<i>615</i>		<i>466</i>	

Table 1.6 reports the number of adult CF patients who have children. Nearly one in 7 adult CF patients are recorded as having children.

Table 1.6: ACFDR 31 December 2006: Number of children

CF patients with:	Sex of CF patient	
	Males	Females
No children	288	206
1 child	20	33
2 children	19	13
3 or more children	3	3
Unknown/not reported	285	211
<i>Total</i>	<i>615</i>	<i>466</i>

Many people with cystic fibrosis continue with education beyond school level, with 12% of adult CF patients having university qualifications and a further 12% having completed other study beyond school.

Table 1.7: ACFDR 31 December 2006: Educational attainment of adults

	Number	Per cent
Junior Secondary (Year 10)	54	11.1
Senior Secondary (Year 12)	170	34.8
Tertiary Certificate or Diploma	58	11.9
University Degree	61	12.5
Left school prior to Year 10	10	2.1
Not applicable (a)	135	27.7
		100.0
Unknown (b)	593	54.9
<i>Total</i>	<i>1,081</i>	<i>100.0</i>

(a) currently studying

(b) includes not recorded

Nearly two thirds of the Registry's adult population were in either full-time or part-time paid employment during 2006.

Table 1.8: ACFDR 31 December 2006: Activity status of adults

	Number	Per cent
Employed, full time paid	184	34.0
Employed, part time paid	175	31.8
Voluntary work only	4	0.7
Unemployed	15	2.8
Pensioner	75	13.9
Others not in labour force (a)	92	16.8
		100.0
Unknown/not reported	536	49.6
<i>Total</i>	<i>1,081</i>	<i>100.0</i>

(a) includes homemakers, students

2 Diagnosis

2.1 Overview

In the reference year 2006, 63 newly diagnosed cases of cystic fibrosis were included in the Australasian Cystic Fibrosis Data Registry. This is the same as new cases reported in 2005. Most diagnoses were made during infancy, that is before the first birthday (see Table 2.1).

Presentation and diagnostic information was recorded for 93% of the CF patient population in 2006. Information was available for at least two thirds of patients in all centres with missing diagnostic information concentrated in a small number of treatment centres.

How is cystic fibrosis diagnosed?

The majority of Australian cystic fibrosis diagnoses occur within or soon after the neonatal period. Neonatal screening programs test a blood sample taken from all newborn babies at around 3 days. Where immuno-reactive trypsin (IRT), a product of the pancreas, is found in the blood at a level above the 99th percentile, genetic testing is carried out to screen for the most common gene mutation or mutations associated with cystic fibrosis.

Where two copies of a CF gene mutation are found, cystic fibrosis is diagnosed. Where one copy is found, a sweat test for elevated electrolyte levels is undertaken to establish diagnosis. If no copies of a CF gene mutation are found, it is considered unlikely that the child has inherited cystic fibrosis. However, only the most common CF gene mutations from a very large range of mostly rare possibilities are screened. Thus a child may present with other symptoms later. These could be respiratory distress, pancreatic insufficiency, rectal prolapse or failure to thrive. Some may present much later with symptoms indicating infertility.

Some other early signs may aid diagnosis. The most common of these is meconium ileus, or failure to pass meconium from the intestines within 48 hours of birth. Where there is already a CF sibling, diagnosis may be established by antenatal testing of fetal DNA obtained at amniocentesis.

2.2 Age at diagnosis

Figure 2.1 shows the large proportion of infant diagnoses that are completed in the first three months, the result of neonatal screening programs that operate in all States and Territories of Australia.

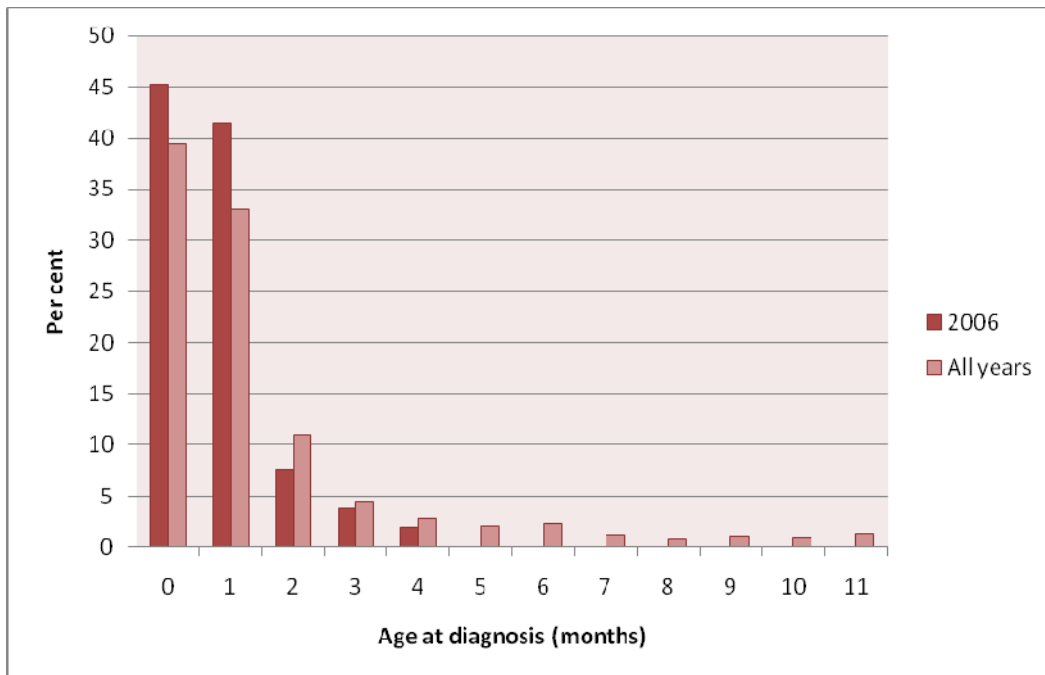


Figure 2.1: ACFDR 31 December 2006: Infant diagnosis by months of age (per cent distribution)

Australian CF centres reported 4 new cases diagnosed in early childhood (1 to 4 years), one child aged 5 to 9 years, 10 to 14 years and 15-19 years, two aged 25 to 34 years and one new diagnoses in an adult aged over 35 years (Table 2.1).

Table 2.1: ACFDR 31 December 2006: Age at diagnosis

	All years	2003	2004	2005	2006	All years	2003	2004	2005	2006
	<i>Number</i>					<i>Per cent</i>				
0 months	576	34	16	37	24	31.0	38.6	27.6	45.1	38.1
1 month	483	29	24	25	22	26.0	33.0	41.4	30.5	34.9
2 months	160	3	6	5	4	8.6	3.4	10.3	6.1	6.4
3 months	64	0	1	0	2	3.4	0.0	1.7	0.0	3.2
4 months	39	2	1	0	1	2.1	2.3	1.7	0.0	1.6
5 months	30	0	0	0	0	1.6	0.0	0.0	0.0	0.0
6 - 11 months	109	5	0	2	0	5.9	5.7	0.0	2.4	0.0
1 - 4 years	211	5	2	6	4	11.4	5.7	3.5	7.3	6.4
5 - 9 years	73	4	3	2	1	3.9	4.6	5.2	2.4	1.6
10 - 14 years	41	0	3	0	1	2.2	0.0	5.2	0.0	1.6
15 - 19 years	17	2	1	0	1	0.9	2.3	1.7	0.0	1.6
20-24 years	21	3	0	3	0	1.1	3.4	0.0	3.7	0.0
25 - 34 years	14	0	0	0	2	0.8	0.0	0.0	0.0	3.2
35+ years	20	1	1	2	1	1.1	1.1	1.7	2.4	1.6
Not known (a)	609	0	0	0	0					
Total	2,467	88	58	82	63	100.0	100.0	100.0	100.0	100.0

(a) Not known have been excluded when calculating percentages.

2.3 Presentation and diagnosis

Nearly 80% of new cases in 2006 included neonatal screening as a mode of presentation. Next most common was respiratory symptoms with 14% of cases and then meconium ileus reported in 13% of new diagnoses (Table 2.2).

Table 2.2 ACFDR 31 December 2006: Mode of presentation (a) by year of diagnosis

	All years					All years				
	2003	2004	2005	2006	2003	2004	2005	2006		
	<i>Number</i>					<i>Per cent</i>				
Neonatal screening	916	56	39	52	50	50.6	63.6	67.2	63.4	79.4
Respiratory symptoms	379	14	10	9	9	20.9	15.9	17.2	11	14.3
Gastrointestinal symptoms	311	12	3	6	4	17.2	13.6	5.2	7.3	6.3
Meconium ileus	273	12	8	20	8	15.1	13.6	13.8	24.4	12.7
CF sibling	160	3	4	6	5	8.8	3.4	6.9	7.3	7.9
Minor manifestations	18	0	1	2	2	1	0	1.7	2.4	3.2
Pre-natal diagnosis	18	1	0	3	0	1	1.1	0	3.7	0
Infertility	9	0	0	0	1	0.5	0	0	0	1.6
Other	262	9	7	7	6	14.5	10.2	12.1	8.5	9.5
Not known(b)	51	0	0	0	0					
Total	1862	88	58	82	63	100.0	100.0	100.0	100.0	100.0

(a) More than one mode of presentation can be recorded for a patient so numbers in this section add to more than the total number of registrants and percentage columns add to more than 100.0.

(b) Not known have been excluded when calculating percentages.

All new cases of cystic fibrosis identified in 2006 have been genotyped.

Table 2.3: ACFDR 31 December 2006: Whether patient genotyped, by year of diagnosis

	All years					All years				
	2003	2004	2005	2006	2003	2004	2005	2006		
	<i>Number</i>					<i>Per cent</i>				
Genotyped	1,976	86	55	78	63	80.2	97.7	94.8	95.1	100.0
Not genotyped	101	1	2	0	0	4.1	1.1	3.5	0.0	0.0
Unknown/Not recorded	390	1	1	4	0	15.7	1.1	1.7	4.9	0.0
Total	2,467	88	58	82	63	100.0	100.0	100.0	100.0	100.0

Three patients diagnosed in 2006 recorded both sodium and chloride levels below diagnostic thresholds of 60 mmol/L. All of these were genotyped.

Table 2.4: ACFDR 31 December 2006: Sweat electrolyte levels, by year of diagnosis

	All years					All years				
	2003	2004	2005	2006	2003	2004	2005	2006		
	<i>Number</i>					<i>Per cent</i>				
Sodium > 60mmol/l	811	43	23	43	34	32.9	48.9	39.7	52.4	54.0
Sodium <= 60mmol/l	166	14	13	10	6	6.7	15.9	22.4	12.2	9.5
Not tested / not recorded	1,490	31	22	29	23	60.4	35.2	37.9	35.4	36.5
<i>Total</i>	<i>2,467</i>	<i>88</i>	<i>58</i>	<i>82</i>	<i>63</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>
Chloride > 60 mmol/l	850	50	31	46	37	34.5	56.8	53.5	56.1	58.7
Chloride <= 60 mmol/l	104	15	11	7	4	4.2	17.1	19.0	8.5	6.4
Not tested / not recorded	1513	23	16	29	22	61.3	26.1	27.6	35.4	34.9
<i>Total</i>	<i>2467</i>	<i>88</i>	<i>58</i>	<i>82</i>	<i>63</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>

2.4 Genotyping

The genetic mutation $\Delta F508$ has been identified as at least one of the paired genes responsible for the inheritance of cystic fibrosis in 94% of patients who have been genotyped. Just over half (51%) are reported as homozygous for $\Delta F508$.

Table 2.5: ACFDR 31 December 2006: Genotype

	$\Delta F508$	G551D	Mutation 1				Other	Total
			R117H	G542X	R553X			
	<i>Number</i>							
Mutation 2:								
$\Delta F508$	968							
G551D	106	2						
R117H	39	7	0					
G542X	9	0	0	0				
R553X	46	1	0	0	4			
Other	202	10	5	1	4	32		
Unknown / missing	417	9	9	4	5	18		
<i>Total</i>	<i>1,787</i>	<i>29</i>	<i>14</i>	<i>5</i>	<i>13</i>	<i>50</i>	<i>1,898</i>	
	<i>Per cent</i>							
Mutation 2:								
$\Delta F508$	51.0							
G551D	5.6	0.1						
R117H	2.1	0.4	0.0					
G542X	0.5	0.0	0.0	0.0				
R553X	2.4	0.1	0.0	0.0	0.2			
Other	10.6	0.5	0.3	0.1	0.2	1.7		
Unknown / missing	22.0	0.5	0.5	0.2	0.3	1.0		
<i>Total</i>	<i>94.2</i>	<i>1.5</i>	<i>0.7</i>	<i>0.3</i>	<i>0.7</i>	<i>2.6</i>	<i>100.0</i>	

2.5 Pancreatic insufficiency

Table 2.6: ACFDR 2006: Pancreatic insufficiency

	Males	Females	Persons	Males	Females	Persons
	<i>Number</i>			<i>Per cent</i>		
0 - 4 years	139	148	287	88.5	86.6	87.5
5 - 9 years	152	145	297	84.0	86.3	85.1
10 - 14 years	166	145	311	84.7	81.9	83.4
15 - 19 years	136	144	280	77.3	82.3	79.8
20 - 24 years	135	100	235	83.9	85.5	84.5
25 - 29 years	92	69	161	82.9	82.1	82.6
30 - 34 years	60	66	126	81.1	81.5	81.3
35 + years	109	62	171	80.2	75.6	78.4
<i>Total</i>	<i>989</i>	<i>879</i>	<i>1,868</i>	<i>83.0</i>	<i>83.3</i>	<i>83.1</i>

The overall proportion of patients who are pancreatic insufficient is just over 83 percent, with no difference evident between the sexes.

3 Health and functioning

Information in this chapter covers respiratory infections, medical complications, lung function and nutritional measures. For a number of centres microbiology results from respiratory samples, lung function and height and weight measures were taken from data that are now being supplied to the registry for each test or measurement. However, data from one paediatric centre was provided, for clinical measures only, in annual summary format similar to that recorded on former collection instruments. Information reported in this chapter reflects good response rates of 94%. While all centres have good coverage the non-reporting is mainly within two centres.

3.1 Respiratory infections

Table 3.1 ACFDR 2006: Number of sputum and BAL/bronchoscopy cultures

	0 - 4 years	5 - 9 years	10 - 14 years	15 - 19 years	20 - 24 years	25 - 29 years	30 - 34 years	35 + years	All ages
<i>Per cent of patients tested (a)</i>									
Sputum cultures:									
None	36.8	12.4	0.0	0.0	2.5	5.0	0.0	3.2	9.6
1	15.2	20.8	23.4	24.7	28.6	24.0	26.2	26.9	22.8
2	13.0	15.5	19.6	19.1	14.9	19.0	21.5	14.0	16.7
3	8.2	13.3	16.6	16.5	10.6	8.0	16.9	15.1	13.0
4	5.6	10.2	9.4	11.9	13.0	12.0	7.7	11.8	10.0
5	6.5	8.0	9.4	8.3	4.4	7.0	1.5	2.2	6.7
6	2.2	7.1	7.2	6.7	5.6	5.0	1.5	4.3	5.4
7 or more	12.6	12.8	14.5	12.9	20.5	20.0	24.6	22.6	15.9
<i>Total</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>
BAL/bronchoscopy:									
None	63.6	81.0	97.5	100.0	98.8	99.0	100.0	98.9	89.5
1	31.6	15.0	2.6	0.0	0.6	1.0	0.0	1.1	8.9
2	3.9	3.1	0.0	0.0	0.6	0.0	0.0	0.0	1.3
3 or more	0.9	0.9	0.0	0.0	0.0	0.0	0.0	0.0	0.3
<i>Total</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>
<i>Number of patients</i>									
<i>Patients tested (a)</i>	231	226	235	194	161	100	65	93	1,305
Culture not done	119	123	125	144	156	105	92	144	1,008
<i>Total reported</i>	<i>350</i>	<i>349</i>	<i>360</i>	<i>338</i>	<i>317</i>	<i>205</i>	<i>157</i>	<i>237</i>	<i>2,313</i>
Not reported	27	30	31	38	4	5	10	9	154
<i>Total patients</i>	<i>377</i>	<i>379</i>	<i>391</i>	<i>376</i>	<i>321</i>	<i>210</i>	<i>167</i>	<i>246</i>	<i>2,467</i>

(a) By any method of obtaining culture.

Table 3.1 shows the distribution of CF patients according to the number of both sputum and BAL/bronchoscopy samples examined during 2006. The latter method is used mainly on smaller children. Taking sputum samples alone, 68 per cent of patients tested had at least two sputum samples in 2006. It can also be seen that cultures were not done for around 44% of patients.

The most commonly identified organisms in respiratory specimens are various species and forms of *Pseudomonas* (see Table 3.2). Six out of ten patients tested produced positive *Pseudomonas* cultures, with the mucoid form of *Pseudomonas aeruginosa* showing in 44 per cent. Its presence is greater in adult patients. The table shows proportions of around 80 per cent of adult CF patients produced samples indicating the mucoid form of *Pseudomonas aeruginosa* in 2006.

Table 3.2: ACFDR 2006: Pseudomonas infection by age group

	0 - 4	5 - 9	10 - 14	15 - 19	20 - 24	25 - 29	30 - 34	35 +	All
	years	years	years	years	years	years	years	years	ages
<i>Number</i>									
Pseudomonas aeruginosa:									
Mucoid	8	36	83	115	134	80	51	72	579
Rough/non-mucoid	29	44	74	81	68	48	23	30	397
Not differentiated	19	29	35	30	35	27	17	22	214
Any Ps aeruginosa	48	83	136	151	144	90	58	79	789
Pseudomonas other species	0	2	5	8	4	2	2	5	28
<i>Patients tested</i>	231	226	235	194	161	100	65	93	1,305
Culture not done	119	123	125	144	156	105	92	144	1,008
<i>Total reported</i>	350	349	360	338	317	205	157	237	2,313
Not reported	27	30	31	38	4	5	10	9	154
<i>Total patients</i>	377	379	391	376	321	210	167	246	2,467
<i>Per cent</i>									
Pseudomonas aeruginosa:									
Mucoid	3.5	15.9	35.3	59.3	83.2	80.0	78.5	77.4	44.4
Rough/non-mucoid	12.6	19.5	31.5	41.8	42.2	48.0	35.4	32.3	30.4
Not differentiated	8.2	12.8	14.9	15.5	21.7	27.0	26.2	23.7	16.4
Any Ps aeruginosa	20.8	36.7	57.9	77.8	89.4	90.0	89.2	84.9	60.5
Pseudomonas other species	0.0	0.9	2.1	4.1	2.5	2.0	3.1	5.4	2.1
<i>Patients tested</i>	100.0	100.0	100.0	100.0	100.0	100.0	100.0	100.0	100.0

While prevalence of *Pseudomonas* organisms is lower in children than in adults, though increasing with rising age, young children are more likely than adult patients to produce cultures showing presence of *Staphylococcus aureus*. About half of all child patients had this bacterial infection. *Haemophilus influenzae* is also evident in relatively high proportions of child patients, even the youngest. This organism was present in a quarter of all children in the 0 to 4 years age group and 21 per cent in the 5 to 9 years age group, but it is less prevalent at older ages. The 0 to 4 years age group also had the highest proportions (19 per cent) with positive cultures of the bacteria *Escherichia coli*.

Table 3.3: ACFDR 2006: Other respiratory culture by age group

	0 - 4 years	5 - 9 years	10 - 14 years	15 - 19 years	20 - 24 years	25 - 29 years	30 - 34 years	35+ years	Total
<i>Number</i>									
Bacteria:									
Staphylococcus aureus	92	126	146	104	56	37	25	23	609
Haemophilus influenzae	58	47	26	15	9	6	0	7	168
Burkholderia cepacia (Ps cepacia)	3	5	5	9	7	4	5	6	44
Stenotrophomonas maltophilia	3	18	18	11	6	9	3	5	73
Escherichia coli	43	10	14	3	0	0	1	0	71
MRSA (a)	2	8	7	12	8	5	4	2	48
Alcaligenes xylosoxidans	0	3	3	2	8	3	6	4	29
Serratia marcescens	4	3	4	4	1	1	1	1	19
Klebsiella (any species)	16	2	3	1	0	0	0	0	22
Non-tuberculous mycobacterium	0	0	0	1	7	1	3	4	16
Fungi:									
Candida	41	65	80	85	26	15	9	19	340
Aspergillus (any species)	9	53	72	47	40	27	21	28	297
Scediosporium (any species)	1	9	7	11	3	3	5	3	42
Other organisms not listed above	85	104	111	82	51	39	24	36	532
Normal flora only	200	194	207	161	90	51	42	50	995
No growth/sterile culture	41	29	12	20	48	39	22	30	241
<i>Patients tested</i>	231	226	235	194	161	100	65	93	1,305
Culture not done	119	123	125	144	156	105	92	144	1,008
<i>Total reported</i>	350	349	360	338	317	205	157	237	2,313
Not reported	27	30	31	38	4	5	10	9	154
<i>Total patients</i>	377	379	391	376	321	210	167	246	2,467
<i>Per cent of patients tested</i>									
Bacteria:									
Staphylococcus aureus	39.8	55.8	62.1	53.6	34.8	37.0	38.5	24.7	46.7
Haemophilus influenzae	25.1	20.8	11.1	7.7	5.6	6.0	0.0	7.5	12.9
Burkholderia cepacia (Ps cepacia)	1.3	2.2	2.1	4.6	4.3	4.0	7.7	6.5	3.4
Stenotrophomonas maltophilia	1.3	8.0	7.7	5.7	3.7	9.0	4.6	5.4	5.6
Escherichia coli	18.6	4.4	6.0	1.5	0.0	0.0	1.5	0.0	5.4
MRSA (a)	0.9	3.5	3.0	6.2	5.0	5.0	6.2	2.2	3.7
Alcaligenes xylosoxidans	0.0	1.3	1.3	1.0	5.0	3.0	9.2	4.3	2.2
Serratia marcescens	1.7	1.3	1.7	2.1	0.6	1.0	1.5	1.1	1.5
Klebsiella (any species)	6.9	0.9	1.3	0.5	0.0	0.0	0.0	0.0	1.7
Non-tuberculous mycobacterium	0.0	0.0	0.0	0.5	4.3	1.0	4.6	4.3	1.2
Fungi:									
Candida	17.7	28.8	34.0	43.8	16.1	15.0	13.8	20.4	26.1
Aspergillus (any species)	3.9	23.5	30.6	24.2	24.8	27.0	32.3	30.1	22.8
Scediosporium (any species)	0.4	4.0	3.0	5.7	1.9	3.0	7.7	3.2	3.2
Other organisms not listed above	36.8	46.0	47.2	42.3	31.7	39.0	36.9	38.7	40.8
Normal flora only	86.6	85.8	88.1	83.0	55.9	51.0	64.6	53.8	76.2
No growth/sterile culture	17.7	12.8	5.1	10.3	29.8	39.0	33.8	32.3	18.5

(a) Methicillin-resistant Staphylococcus aureus

3.2 Other medical complications

Complications data were subject to about 35 per cent under-reporting in 2006. One major adult centre did not report complications information for its patients. The numbers in the following table show how the prevalence of medical complications increases with age in CF patients. For instance, around 33 per cent of adult patients aged 25 years or over suffer gastro-oesophageal reflux, around 18 per cent have chronic insulin-dependent diabetes and nearly 50 per cent have osteoporosis or osteopenia. More than 20 per cent of adult patients have chronic insulin-dependent diabetes.

The proportion for whom none of the selected complications shown in Table 3.4 have been reported is high for very young children, near 80 per cent, but declines to around 20 per cent or lower in older CF patients.

Table 3.4 ACFDR 2006: Medical complications

	0 - 4 years	5 - 9 years	10 - 14 years	15 - 19 years	20 - 24 years	25 - 29 years	30 - 34 years	35 + years	Total
<i>Number</i>									
Pulmonary:									
Major haemoptysis	3	2	5	7	9	5	4	10	45
Massive haemoptysis	1	0	4	0	5	3	1	3	17
Therapeutic bronchial artery embolisation	1	0	0	1	6	1	0	2	11
Pneumothorax	1	0	0	1	4	0	2	0	8
Any pulmonary above	6	2	8	9	20	8	6	13	72
Gastro-intestinal:									
Gastro-oesophageal reflux	14	14	21	30	53	38	28	37	235
- proven at endoscopy	3	2	5	7	14	10	5	11	57
Abnormal liver function test	13	22	32	47	44	29	21	28	236
Cirrhosis or portal hypertension	0	3	11	13	8	11	2	2	50
Pancreatitis	0	0	2	3	4	1	1	2	13
Any Gastro-intestinal above	26	34	46	67	82	56	40	50	401
Endocrine:									
Chronic insulin-dependent diabetes	0	3	9	22	20	24	16	23	117
Intermittent insulin-dependent diabetes	0	2	4	3	8	6	1	4	28
Other glucose abnormality	8	13	17	16	20	10	12	18	114
Any Endocrine above	8	18	28	41	47	37	29	43	251
Osteo:									
Osteoporosis	1	1	4	20	23	14	5	20	88
Osteopenia	1	1	9	29	55	40	29	42	206
Fracture this year	0	0	1	3	1	3	0	2	10
Any Osteo above	1	1	10	35	65	50	32	55	249
Other:									
Cancer	0	2	0	2	1	0	1	2	8
None of the above	265	218	224	163	47	23	16	15	971
<i>Total reported</i>	<i>302</i>	<i>269</i>	<i>296</i>	<i>264</i>	<i>179</i>	<i>108</i>	<i>73</i>	<i>107</i>	<i>1598</i>
Unknown or not stated	75	110	95	112	142	102	94	139	869
<i>Total patients</i>	<i>377</i>	<i>379</i>	<i>391</i>	<i>376</i>	<i>321</i>	<i>210</i>	<i>167</i>	<i>246</i>	<i>2467</i>

Table 3.4 (cont.)ACFDR 2006: Medical complications

	0 - 4	5 - 9	10 - 14	15 - 19	20 - 24	25 - 29	30 - 34	35 +	Total
	years	years	years	years	years	years	years	years	
	<i>Per cent</i>								
Pulmonary:									
Major haemoptysis	1.0	0.7	1.7	2.7	5.0	4.6	5.5	9.3	2.8
Massive haemoptysis	0.3	0.0	1.4	0.0	2.8	2.8	1.4	2.8	1.1
Therapeutic bronchial artery embolisation	0.3	0.0	0.0	0.4	3.4	0.9	0.0	1.9	0.7
Pneumothorax	0.3	0.0	0.0	0.4	2.2	0.0	2.7	0.0	0.5
Any pulmonary above	2.0	0.7	2.7	3.4	11.2	7.4	8.2	12.1	4.5
Gastro-intestinal:									
Gastro-oesophageal reflux	4.6	5.2	7.1	11.4	29.6	35.2	38.4	34.6	14.7
- proven at endoscopy	1.0	0.7	1.7	2.7	7.8	9.3	6.8	10.3	3.6
Abnormal liver function test	4.3	8.2	10.8	17.8	24.6	26.9	28.8	26.2	14.8
Cirrhosis or portal hypertension	0.0	1.1	3.7	4.9	4.5	10.2	2.7	1.9	3.1
Pancreatitis	0.0	0.0	0.7	1.1	2.2	0.9	1.4	1.9	0.8
Any Gastro-intestinal above	8.6	12.6	15.5	25.4	45.8	51.9	54.8	46.7	25.1
Endocrine:									
Chronic insulin-dependent diabetes	0.0	1.1	3.0	8.3	11.2	22.2	21.9	21.5	7.3
Intermittent insulin-dependent diabetes	0.0	0.7	1.4	1.1	4.5	5.6	1.4	3.7	1.8
Other glucose abnormality	2.6	4.8	5.7	6.1	11.2	9.3	16.4	16.8	7.1
Any Endocrine above	2.6	6.7	9.5	15.5	26.3	34.3	39.7	40.2	15.7
Osteo:									
Osteoporosis	0.3	0.4	1.4	7.6	12.8	13.0	6.8	18.7	5.5
Osteopenia	0.3	0.4	3.0	11.0	30.7	37.0	39.7	39.3	12.9
Fracture this year	0.0	0.0	0.3	1.1	0.6	2.8	0	1.9	0.6
Any Osteo above	0.3	0.4	3.4	13.3	36.3	46.3	43.8	51.4	15.6
Other:									
Cancer	0.0	0.7	0.0	0.8	0.6	0.0	1.4	1.9	0.5
None of the above	87.7	81.0	75.7	61.7	26.3	21.3	21.9	14.0	60.8
<i>Total reported</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>

Note: Patient may have had more than one complication. Percentages add to more than 100.0.

3.3 Lung function

Table 3.5: ACFDR 2006: FEV1 per cent predicted - children and adolescents

	Number	Per cent	Cumulative %	Number	Per cent	Cumulative %
	Males			Females		
FEV1 % predicted:						
<10%	0	0.0	0.0	0	0.0	0.0
10 - 19.99%	0	0.0	0.0	0	0.0	0.0
20 - 29.99%	2	0.6	0.6	0	0.0	0.0
30 - 39.99%	4	1.1	1.6	4	1.1	1.1
40 - 49.99%	3	0.8	2.5	7	1.9	3.0
50 - 59.99%	10	2.7	5.2	16	4.3	7.3
60 - 69.99%	27	7.4	12.6	26	7.1	14.4
70 - 79.99%	42	11.5	24.1	42	11.4	25.8
80 - 89.99%	68	18.6	42.7	67	18.2	43.9
90 - 99.99%	92	25.2	68.0	80	21.7	65.6
>=100%	117	32.1	100.0	127	34.4	100.0
<i>Total measured</i>	<i>365</i>	<i>100.0</i>		<i>369</i>	<i>100.0</i>	

Note: Per cent predicted calculations included only patients aged 6 years and over for whom lung function test results were available.

The usual measure of lung function is forced expiratory volume in one second (FEV1). Per cent predicted FEV1 values, based on 'normal' values observed in studies of healthy subjects, decline along with disease development. The FEV1 values reported in these tables are the 'best' measures reported during the year. Lung function was reported for around 72 per cent of adult patients and 81 per cent of children/adolescents aged 6 years and over.

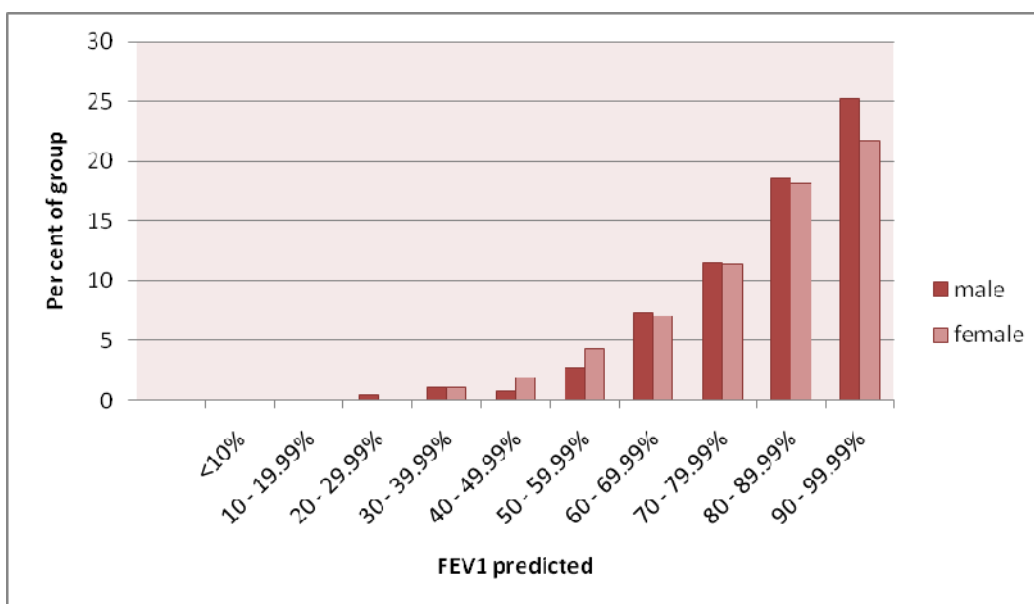


Figure 3.1: ACFDR 2006: Distribution of FEV1 per cent predicted - children and adolescents aged 6 to 17 years

Predicted values for lung function are based on research by Wang et al (1993) and Hankinson et al (1999) – see Technical Notes. Using the new predicted values, Figure 3.1 demonstrates that child CF lung function measured as FEV1 is skewed towards lower than predicted normal levels. From Table 3.5, 13 per cent of males and 14 per cent of females have FEV1 values that are below 70% of predicted values.

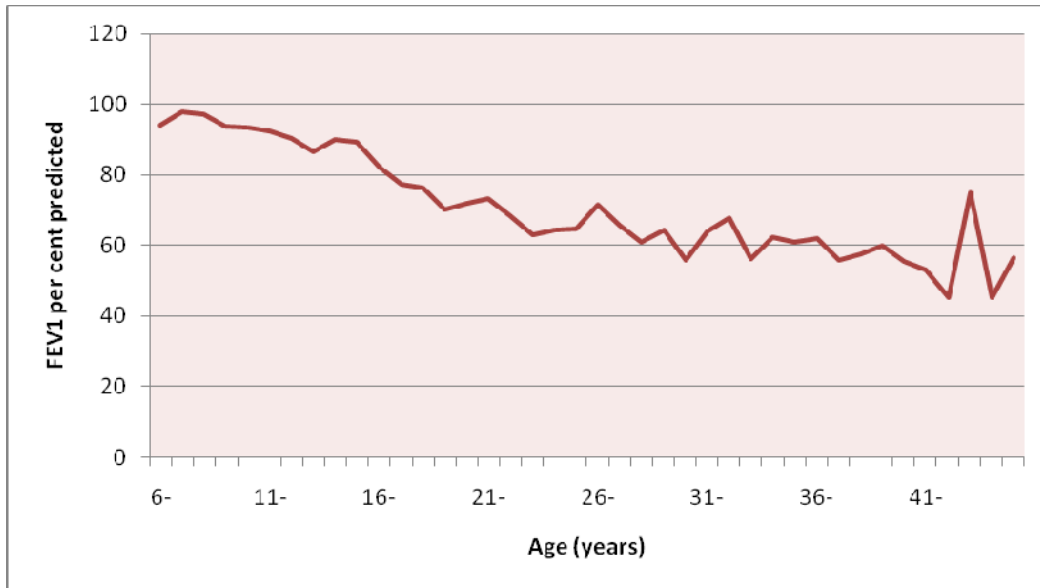


Figure 3.2: ACFDR 2006: Lung function (FEV1 % predicted) by age

Figure 3.2 shows that lung function in is around 75% of predicted values for older children and adolescents and around 60% for adults with CF. Almost 60 per cent of both male and female adults have percent predicted lung function values below 70% (Table 3.6)

Table 3.6 ACFDR 2006: FEV1 per cent predicted - adults

	Number	Per cent	Cumulative %	Number	Per cent	Cumulative %
	Males			Females		
FEV1 % predicted:						
<10%	0	0.0	0.0	0	0.0	0.0
10 - 19.99%	3	0.7	0.7	0	0.0	0.0
20 - 29.99%	24	5.5	6.1	10	3.2	3.2
30 - 39.99%	45	10.2	16.4	29	9.2	12.4
40 - 49.99%	52	11.8	28.2	39	12.4	24.8
50 - 59.99%	50	11.4	39.6	60	19.1	44.0
60 - 69.99%	69	15.7	55.2	44	14.0	58.0
70 - 79.99%	72	16.4	71.6	48	15.3	73.3
80 - 89.99%	62	14.1	85.7	46	14.7	87.9
90 - 99.99%	39	8.9	94.6	24	7.6	95.5
>=100%	24	5.45	100.0	14	4.5	100.0
<i>Total measured</i>	<i>440</i>	<i>100</i>		<i>314</i>	<i>100</i>	

The decline in lung function with age can be seen also in comparison of Figures 3.1 (children and adolescents) and 3.3 (adults). For adult patients the modal point for the distribution against predicted FEV1 values is in a lower range than is seen for children and adolescents, for both male and female patients.

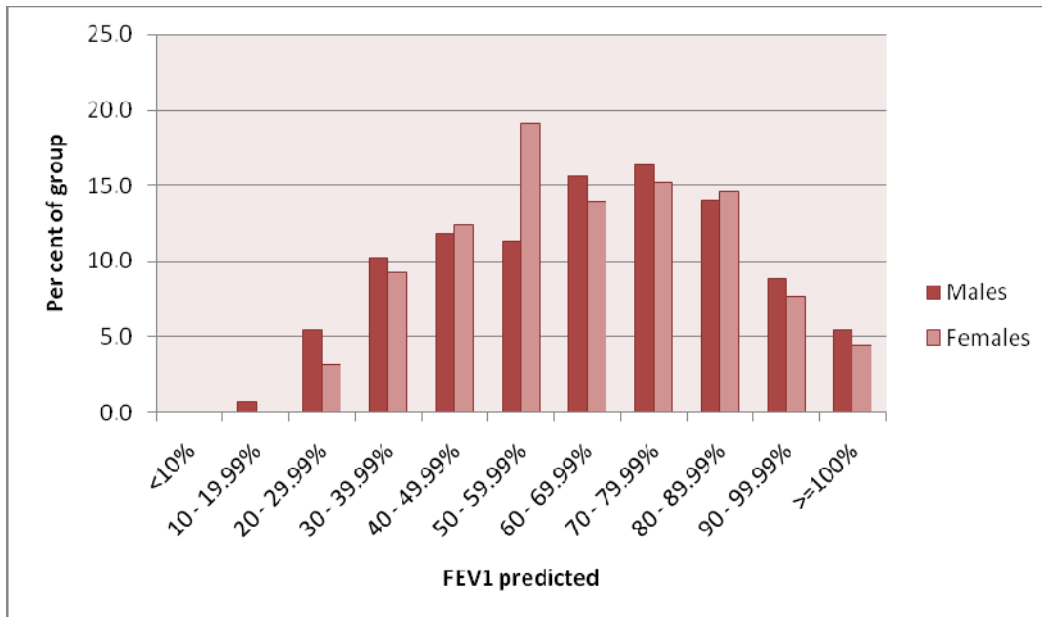


Figure 3.3: ACFDR 2006: Distribution of FEV1 per cent predicted - adults

An alternate view of lung function is provided in the following tables and charts, in which the distributions are summarised into categories of normal lung function (90% of predicted FEV1 and above), mild impairment (below 90% but not below 70%), moderate (below 70% but not below 40%) and severe (below 40%). A higher proportion of male children (57.3%) than females (56.1%) have lung function within the normal range (Table 3.7). This is also evident in Figure 3.4 on the following page.

Table 3.7: ACFDR 2006: Lung function impairment by age and sex - children and adolescents

Age group/sex	Severe	Moderate	Mild	Normal	Total	Severe	Moderate	Mild	Normal	Total
	<i>Number</i>					<i>Per cent</i>				
5 - 9 years	1	19	66	162	248	0.0	6.5	25.8	67.7	100.0
10 - 14 years	2	43	99	180	324	1.3	12.3	30.4	56.0	100.0
15 - 17 years	6	34	72	60	172	3.5	20.0	34.7	41.8	100.0
<i>Total measured</i>	9	96	237	402	744	1.4	12.1	29.8	56.7	100.0
<i>Males</i>	6	40	110	209	365	1.6	11.0	30.1	57.3	100.0
<i>Females</i>	4	49	109	207	369	1.1	13.3	29.5	56.1	100.0

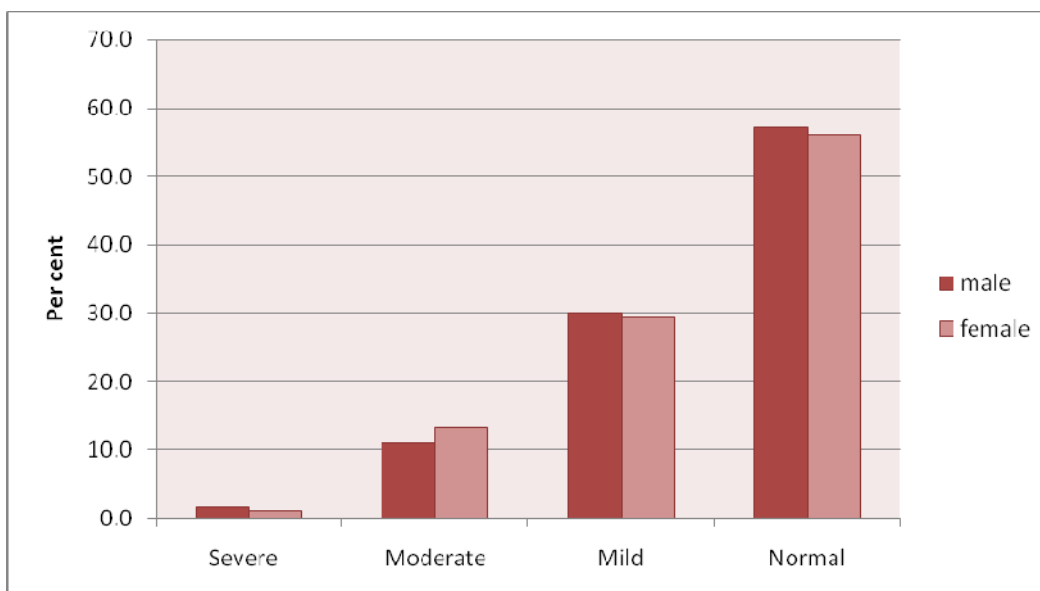


Figure 3.4: ACFDR 2006: Lung function impairment - children and adolescents

Table 3.8 shows generally greater proportions of patients with severe lung function impairment in successive older age groups. The proportion of adult male patients with severe lung function impairment (16%) is greater than the proportion of female patients in this severity category (12%).

Table 3.8: ACFDR 2006: Lung function impairment by age and sex - adults

	Severe	Moderate	Mild	Normal	Total	Severe	Moderate	Mild	Normal	Total
	<i>Number</i>					<i>Per cent</i>				
Age/sex group:										
18 - 19 years	3	42	40	18	103	2.9	40.8	38.8	17.5	100.0
20 - 24 years	31	87	82	45	245	12.7	35.5	33.5	18.4	100.0
25 - 29 years	18	67	45	15	145	12.4	46.2	31.0	10.3	100.0
30 - 34 years	22	50	28	12	112	19.6	44.6	25.0	10.7	100.0
35 + years	37	68	33	11	149	24.8	45.6	22.1	7.4	100.0
<i>Total measured</i>	111	314	228	101	754	14.7	41.6	30.2	13.4	100.0
<i>Males</i>	72	171	134	63	440	16.4	38.9	30.5	14.3	100.0
<i>Females</i>	39	143	94	38	314	12.4	45.5	29.9	12.1	100.0

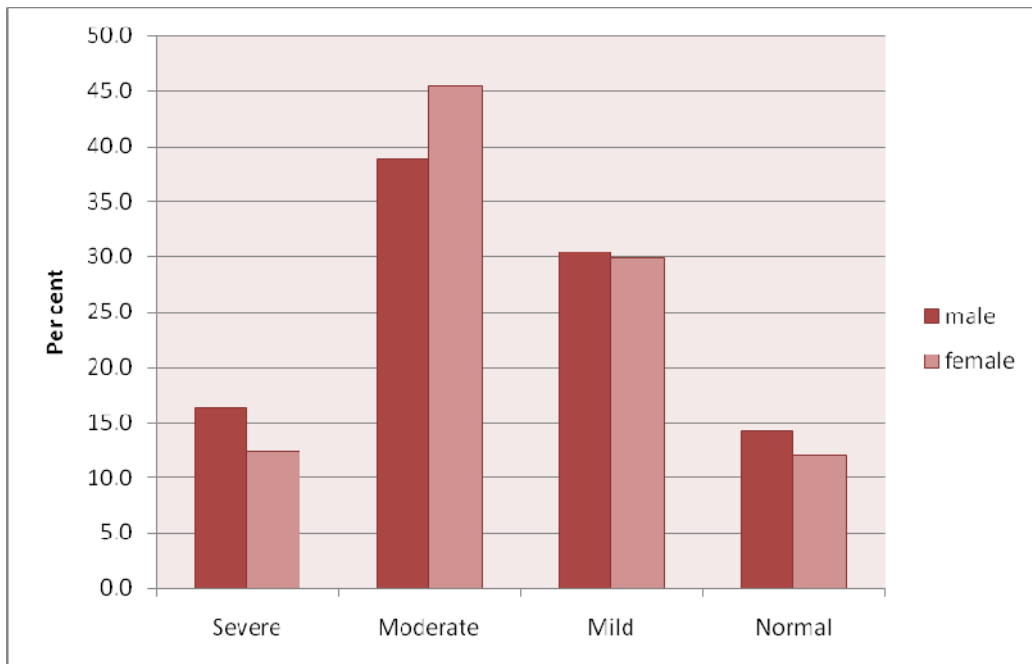


Figure 3.5: ACFDR 2006: Lung function impairment - adults

3.4 Height and weight

Regular monitoring of nutrition outcome by measures of height and weight and calculation of body mass index (BMI) is undertaken within cystic fibrosis clinics, yielding the information presented in this section. Height and weight measures are selected from the date of best BMI. Data were reported for 93 percent of children/adolescents aged 2 to 17 years and 76 per cent of adults. Distributions shown in tables and charts exclude patients for whom data were not reported.

Table 3.9: ACFDR 2006: Child and adolescent height, weight and BMI - mean percentiles by age and sex

	Height	Weight	BMI
<i>Males</i>			
2 years	50.0	58.7	61.6
3 years	50.3	66.6	70.6
4 years	46.8	52.1	59.3
5 years	37.2	49.4	65.9
6 years	39.3	45.4	60.4
7 years	46.9	46.5	48.5
8 years	44.2	56.3	60.2
9 years	34.9	50.1	57.9
10 years	44.3	42.3	46.7
11 years	38.7	44.9	52.7
12 years	37.5	35.8	42.6
13 years	34.0	38.4	47.8
14 years	34.4	37.2	42.6
15 years	37.3	44.4	49.9
16 years	28.3	35.8	41.7
17 years	43.7	41.1	41.3
<i>Females</i>			
2 years	54.5	54.7	53.1
3 years	44.3	52.4	62.9
4 years	50.6	56.7	66.3
5 years	35.3	47.1	62.0
6 years	38.0	46.7	58.6
7 years	44.7	51.9	57.5
8 years	30.5	43.0	55.8
9 years	39.9	39.9	47.6
10 years	48.2	45.2	50.2
11 years	44.7	39.3	42.4
12 years	38.7	43.0	49.7
13 years	31.9	35.9	41.3
14 years	41.9	53.4	57.2
15 years	46.9	49.9	49.9
16 years	49.9	55.2	56.7
17 years	43.8	50.3	53.9

Mean height and weight percentiles for boys were highest for those aged 3 years (50 and 66 per cent respectively). For girls, the highest mean height percentile was 55 per cent at age 2 and highest mean weight percentile was 57 per cent at age 4. The lowest height and weight percentiles for boys were generally recorded by those aged in early adolescence. For girls, lowest height percentiles were recorded by those aged 8 years and 13 years and for weight by those in early adolescence.

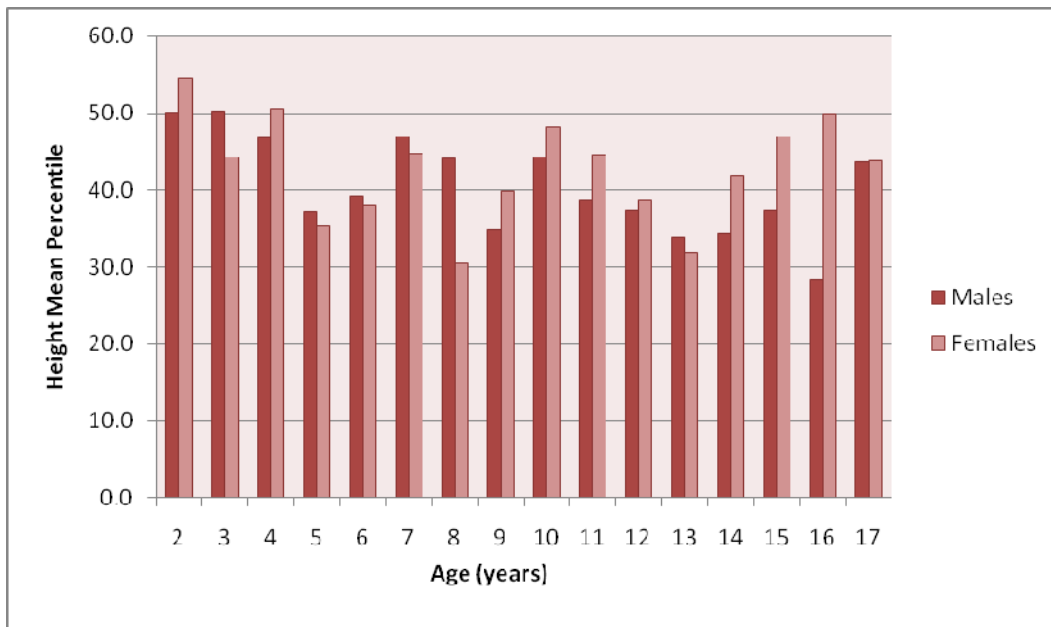


Figure 3.6: ACFDR 2006: Mean percentile height by age - children and adolescents

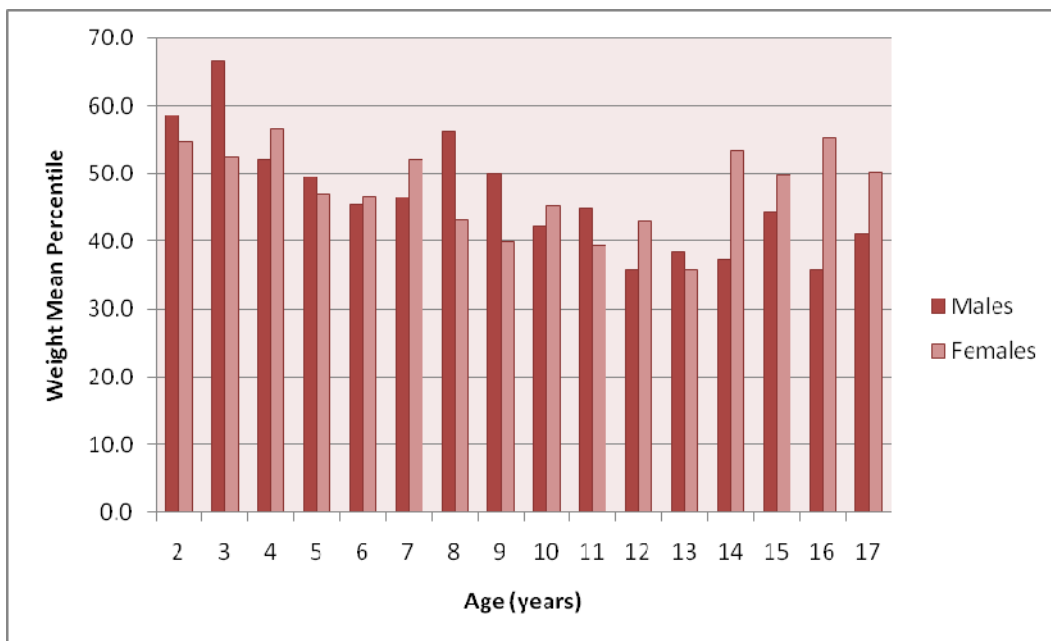


Figure 3.7: ACFDR 2006: Mean percentile weight by age - children and adolescents

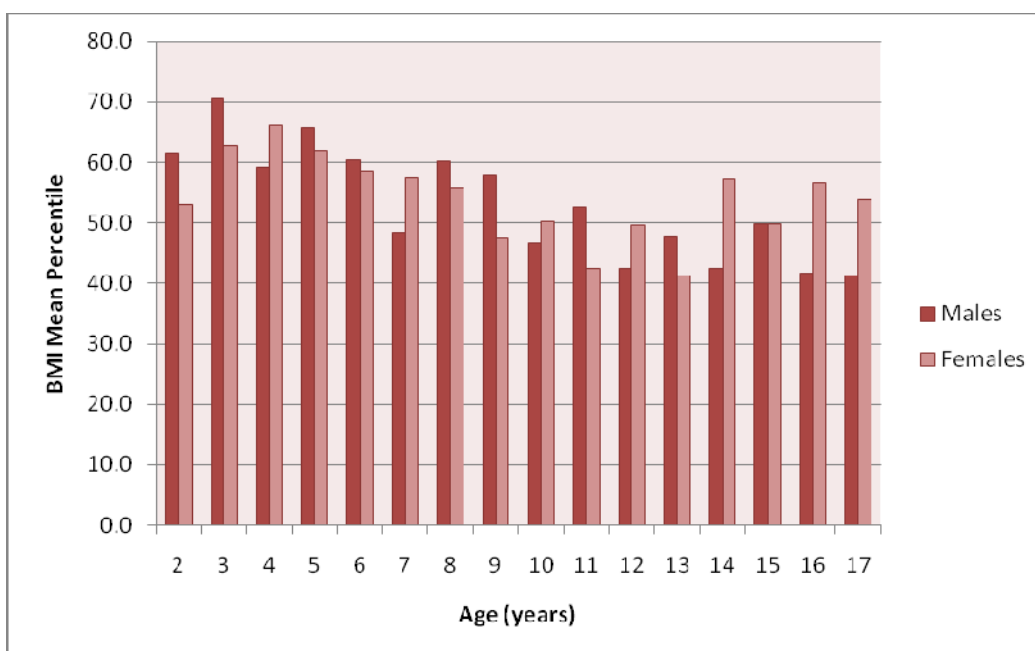


Figure 3.8: ACFDR 2006: Mean percentile BMI by age – children and adolescents

Table 3.10: ACFDR 2006: Child and adolescent height, weight and BMI - percentile distribution by sex

	Height		Weight		BMI	
	Number	Per cent	Number	Per cent	Number	Per cent
<i>Males</i>						
< 5th	60	12.6	34	7.2	17	3.6
5th - 9.99th	28	5.9	31	6.5	16	3.4
10th - 24.99th	92	19.4	82	17.3	59	12.4
25th - 49.99th	113	23.8	108	22.7	132	27.8
50th - 74.99th	102	21.5	118	24.8	126	26.5
75th - 89.99th	53	11.2	62	13.1	77	16.2
90th - 94.99th	16	3.4	22	4.6	27	5.7
>= 95th	11	2.3	18	3.8	21	4.4
<i>Males measured</i>	<i>475</i>	<i>100.0</i>	<i>475</i>	<i>100.0</i>	<i>475</i>	<i>100</i>
<i>Females</i>						
< 5th	51	10.6	36	7.5	14	2.9
5th - 9.99th	25	5.2	24	5.0	21	4.4
10th - 24.99th	82	17.1	63	13.1	51	10.6
25th - 49.99th	125	26.0	131	27.3	120	25.0
50th - 74.99th	114	23.8	127	26.5	152	31.7
75th - 89.99th	54	11.3	65	13.5	82	17.1
90th - 94.99th	19	4.0	19	4.0	16	3.3
>= 95th	10	2.1	15	3.1	24	5.0
<i>Females measured</i>	<i>480</i>	<i>100.0</i>	<i>480</i>	<i>100.0</i>	<i>480</i>	<i>100.0</i>

Alternative presentations of child and adolescent height and weight distributions, by percentile and z-scores are presented in Tables 3.10 and 3.11. In terms of percentile distributions, 62% of male and 59% of female children and adolescents are below the 50th percentile for height. Corresponding figures for weight are 54% and 53%.

Table 3.11: ACFDR 2006: Height and weight z-score distribution for children and adolescents

	Height		Weight	
	Number	Per cent	Number	Per cent
	<i>Males</i>			
Less than -3	3	0.6	6	1.3
From -3 to <-2	30	6.3	9	1.9
From -2 to <-1	91	19.2	89	18.7
From -1 to <0	169	35.6	151	31.8
From 0 to <1	138	29.1	157	33.1
From 1 to <2	37	7.8	56	11.8
From 2 to <3	7	1.5	7	1.5
Greater or =3	0	0.0	0	0.0
<i>Males measured</i>	<i>475</i>	<i>100</i>	<i>475</i>	<i>100</i>
	<i>Females</i>			
Less than -3	7	1.5	5	1.0
From -3 to <-2	22	4.6	16	3.3
From -2 to <-1	82	17.1	63	13.1
From -1 to <0	172	35.8	170	35.4
From 0 to <1	148	30.8	165	34.4
From 1 to <2	44	9.2	53	11.0
From 2 to <3	5	1.0	8	1.7
Greater or =3	0	0.0	0	0.0
<i>Females measured</i>	<i>480</i>	<i>100.0</i>	<i>480</i>	<i>100.0</i>

Table 3.12: ACFDR 2006: Height and weight z-scores for children and adolescents

	Height			Weight		
	Number	Mean z-score	Standard error	Number	Mean z-score	Standard error
<i>Males</i>						
Age:						
2 years	15	-0.01	0.80	15	0.28	0.80
3 years	29	0.02	0.67	29	0.52	0.73
4 years	36	-0.14	1.06	36	0.09	1.09
5 years	31	-0.38	1.02	31	0.01	0.98
6 years	33	-0.41	1.02	33	-0.13	0.95
7 years	29	-0.11	1.22	29	-0.12	0.95
8 years	26	-0.25	0.90	26	0.17	1.03
9 years	28	-0.51	0.85	28	-0.05	1.08
10 years	33	-0.22	1.24	33	-0.23	1.17
11 years	25	-0.43	0.95	25	-0.30	1.08
12 years	39	-0.44	0.90	39	-0.45	0.86
13 years	35	-0.63	1.12	35	-0.40	1.04
14 years	34	-0.60	1.17	34	-0.42	0.88
15 years	28	-0.53	1.10	28	-0.22	0.96
16 years	22	-0.96	1.25	22	-0.84	1.59
17 years	32	-0.22	1.32	32	-0.35	1.11
<i>Females</i>						
2 years	31	0.14	0.77	31	0.13	0.84
3 years	31	-0.15	1.12	31	0.12	1.08
4 years	20	0.00	0.95	20	0.22	1.10
5 years	26	-0.47	0.76	26	-0.07	0.89
6 years	33	-0.52	1.12	33	-0.20	1.12
7 years	30	-0.16	0.88	30	0.11	0.91
8 years	30	-0.83	1.27	30	-0.33	1.15
9 years	34	-0.37	0.96	34	-0.27	1.01
10 years	32	-0.12	1.01	32	-0.22	0.95
11 years	27	-0.17	0.79	27	-0.35	0.72
12 years	29	-0.48	1.21	29	-0.33	1.10
13 years	31	-0.66	1.07	31	-0.50	0.95
14 years	31	-0.49	1.34	31	-0.13	1.45
15 years	30	-0.15	1.15	30	-0.10	1.15
16 years	37	-0.03	1.00	37	0.15	0.94
17 years	28	-0.30	1.09	28	-0.10	1.13

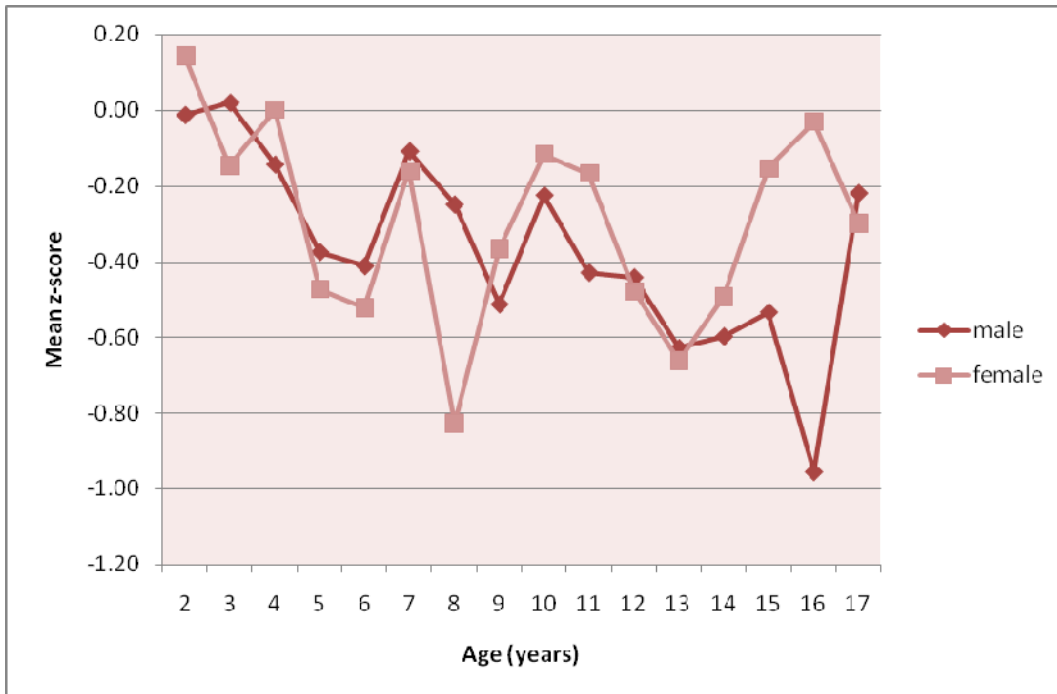
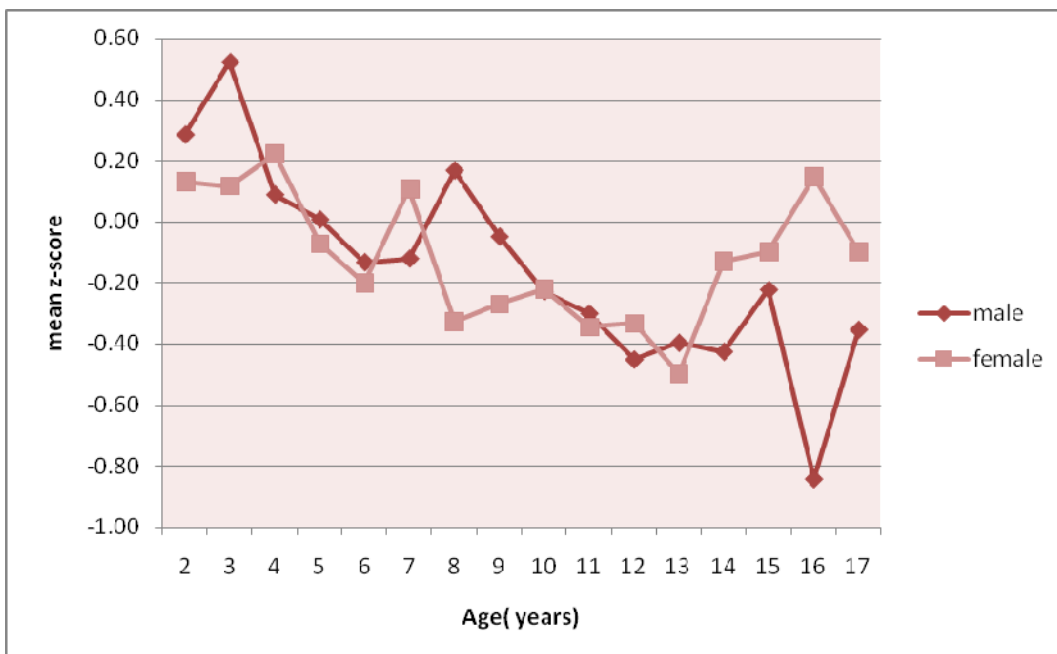


Figure 3.9: ACFDR 2006: Mean height z-score by age of children and adolescents

Figure 3.10: ACFDR 2005: Mean weight z-score by age of children and adolescents



Adult Body Mass Index scores are shown in Figure 3.11 and Table 3.13. More than 57% of both males and females have a BMI in the range 20 to less than 25. A higher proportion of females than males have BMI scores below 20.

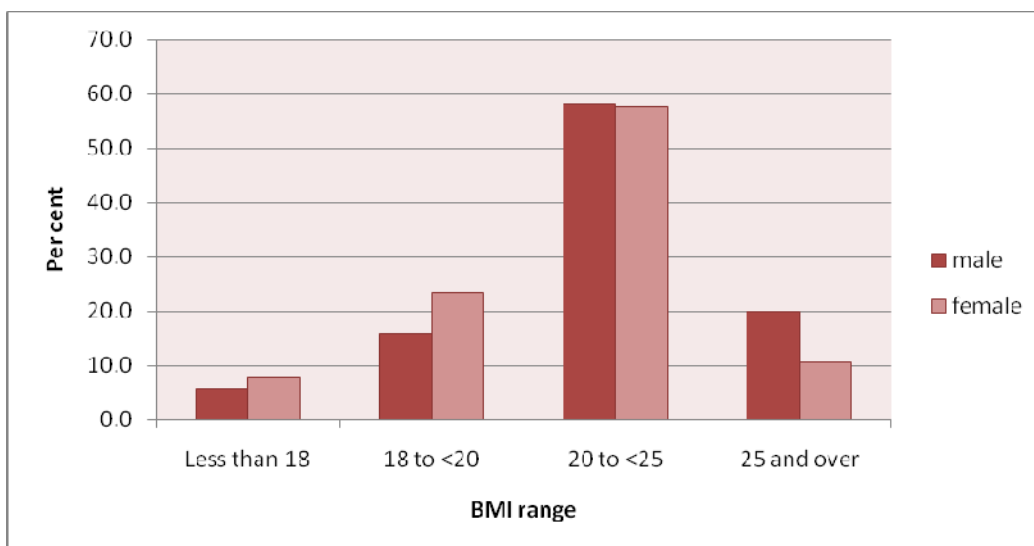


Figure 3.11: ACFDR 2006: Per cent distribution of adult BMI

Table 3.13: ACFDR 2006 Adult BMI distribution

	BMI range				Total
	Less than 18	From 18 to <20	From 20 to <25	25 and over	
<i>Males: number</i>					
18 - 19 years	7	8	35	7	57
20 - 24 years	18	24	90	19	151
25 - 29 years	3	9	59	17	88
30 - 34 years	3	6	42	12	63
35 + years	3	7	48	40	98
<i>Adults measured</i>	<i>34</i>	<i>54</i>	<i>274</i>	<i>95</i>	<i>457</i>
<i>Females: number</i>					
18 - 19 years	4	14	25	2	45
20 - 24 years	11	25	57	7	100
25 - 29 years	5	12	41	7	65
30 - 34 years	4	13	39	7	63
35 + years	3	15	32	13	63
<i>Adults measured</i>	<i>27</i>	<i>79</i>	<i>194</i>	<i>36</i>	<i>336</i>
<i>Males: per cent</i>					
18 - 19 years	14.8	25.9	51.9	7.4	100.0
20 - 24 years	5.3	22.7	56.8	15.2	100.0
25 - 29 years	3.7	12.4	65.4	18.5	100.0
30 - 34 years	9.5	4.8	63.5	22.2	100.0
35 + years	0.0	11.6	53.5	34.9	100.0
<i>Adults measured</i>	<i>5.8</i>	<i>16.1</i>	<i>58.2</i>	<i>20.0</i>	<i>100.0</i>
<i>Females: per cent</i>					
18 - 19 years	8.9	31.1	55.6	4.4	100.0
20 - 24 years	11.0	25.0	57.0	7.0	100.0
25 - 29 years	7.7	18.5	63.1	10.8	100.0
30 - 34 years	6.4	20.6	61.9	11.1	100.0
35 + years	4.8	23.8	50.8	20.6	100.0
<i>Adults measured</i>	<i>8.0</i>	<i>23.5</i>	<i>57.7</i>	<i>10.7</i>	<i>100.0</i>

4 Treatment of cystic fibrosis

This Chapter describes the treatments and therapies recorded for patients in the Australasian Cystic Fibrosis Registry.

4.1 Visits to clinics

Table 4.1: ACFDR 2006: Outpatient visits to CF clinics

CF patients making:	Children and Adolescents		Adults	
	Number	Per cent	Number	Per cent
No visits	13	1.2	50.0	7.1
1 visit (a)	100	9.0	82.0	11.6
2 visits	143	12.9	85.0	12.1
3 visits	164	14.8	61.0	8.7
4 visits	235	21.2	82.0	11.6
5 visits	149	13.4	55.0	7.8
6 visits	99	8.9	68.0	9.7
7 visits	52	4.7	46.0	6.5
8 visits	40	3.6	39.0	5.5
9 visits	28	2.5	32.0	4.5
10 visits	22	2.0	18.0	2.6
11 visits	17	1.5	14.0	2.0
12 or more visits	48	4.3	73.0	10.4
<i>Total with number of visits reported</i>	<i>1,110</i>	<i>100</i>	<i>705</i>	<i>100</i>

(a) See text below

Table 4.1 demonstrates that some patients make a large number of visits to specialist clinics over a year. The average number of visits for children and adolescents was 4.6 and for adults 4.2. The median number of visits to clinics was 4 for children and adolescents and 3 for adults in 2006. Only one clinic visit per patient was reported by one adult centre where it is likely that multiple visits occurred. Counts of clinic visits have not been included in Table 4.1 for this centre.

4.2 Therapy for cystic fibrosis patients

Table 4.2 shows that the Data Registry recorded 62 cystic fibrosis patients who had at least one course of intravenous antibiotic therapy administered at home during 2006. Around three times as many had courses administered both in hospital and at home.

Table 4.2: ACFDR 2006: Whether had home IV antibiotic therapy by age group

	0 to 4 years	5 to 9 years	10 to 14 years	15 to 19 years	20 to 24 years	25 to 29 years	30 to 34 years	35 + years	All ages
Courses at home:									
	<i>Number of patients</i>								
1	1	6	4	3	6	3	4	3	30
2	0	2	2	1	6	4	4	4	23
3 or more	0	0	0	0	2	3	1	3	9
<i>Total</i>	<i>1</i>	<i>8</i>	<i>6</i>	<i>4</i>	<i>14</i>	<i>10</i>	<i>9</i>	<i>10</i>	<i>62</i>
Courses at both hospital and home									
1	21	21	19	12	17	12	12	9	123
2	6	5	11	8	7	3	3	6	49
3 or more	1	2	4	10	2	2	1	2	24
<i>Total</i>	<i>28</i>	<i>28</i>	<i>34</i>	<i>30</i>	<i>26</i>	<i>17</i>	<i>16</i>	<i>17</i>	<i>196</i>

Almost all cystic fibrosis patients use antibiotic therapy in some form. Table 4.3 shows the proportion to be 96 per cent of patients for whom information was reported. Use of therapies was reported for two thirds of patients in 2006.

Table 4.3: ACFDR 2006: Antibiotic therapy by age group

	0 to 4 years	5 to 9 years	10 to 14 years	15 to 19 years	20 to 24 years	25 to 30 years	30 to 35 years	35 + years	All ages
<i>Number</i>									
Antibiotic use:									
Yes	274	277	281	250	179	112	76	110	1,559
No	25	8	13	14	4	3	1	2	70
Total reported	299	285	294	264	183	115	77	112	1,629
Not reported	78	94	97	112	138	95	90	134	838
<i>Total patients</i>	<i>377</i>	<i>379</i>	<i>391</i>	<i>376</i>	<i>321</i>	<i>210</i>	<i>167</i>	<i>246</i>	<i>2,467</i>
<i>Per cent</i>									
Yes	91.6	97.2	95.6	94.7	97.8	97.4	98.7	98.2	95.7
No	8.4	2.8	4.4	5.3	2.2	2.6	1.3	1.8	4.3
<i>Total reported</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>

Oral antibiotics were used by 96 per cent of patients who were reported as using antibiotics. Of these, 18 per cent had used oral antibiotics continuously. The age groups with the highest proportions of continuous oral antibiotics users were young children and adolescents. Within both of these groups, around 25 per cent of oral antibiotics users took them on a continuous basis (Table 4.4).

Table 4.4: ACFDR 2006: Oral antibiotic therapy by age group

	0 to 4 years	5 to 9 years	10 to 14 years	15 to 19 years	20 to 24 years	25 to 30 years	30 to 35 years	35 + years	All ages
<i>Per cent</i>									
Oral antibiotics use:									
Yes	98.2	98.9	98.6	96.0	90.8	92.5	93.2	95.4	96.3
No	1.9	1.1	1.4	2.8	4.1	3.8	2.7	1.9	2.2
Unknown (a)	0.0	0.0	0.0	1.2	5.2	3.8	4.1	2.8	1.4
<i>Total antibiotics users</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>
<i>Number</i>									
<i>Total antibiotics users</i>	<i>271</i>	<i>272</i>	<i>281</i>	<i>248</i>	<i>173</i>	<i>106</i>	<i>73</i>	<i>108</i>	<i>1,532</i>
<i>Per cent</i>									
Mode of use:									
As needed (PRN)	79.4	83.2	74.7	70.9	92.4	95.1	96.4	94.4	82.1
Continuous	20.6	16.8	25.3	29.1	7.6	4.9	3.6	5.6	18.0
<i>Total oral antibiotics users(b)</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>
<i>Number</i>									
<i>Total oral antibiotics users</i>	<i>266</i>	<i>269</i>	<i>277</i>	<i>238</i>	<i>157</i>	<i>98</i>	<i>68</i>	<i>103</i>	<i>1476</i>

(a) includes not recorded

(b) More than one mode of use can be recorded so numbers add to more than 100.0. As well, mode of use was not recorded for all patients where oral antibiotics were reported.

Inhaled antibiotics were used by 56% of CF patients during 2006, with those in the 0 to 4 years age group being lowest at 23% (Table 4.5). Amongst inhaled antibiotics users, 19% used on a continuous basis.

Table 4.5: ACFDR 2006: Inhaled antibiotics by age group

	0 to 4 years	5 to 9 years	10 to 14 years	15 to 19 years	20 to 24 years	25 to 30 years	30 to 35 years	35 + years	All ages
<i>Per cent</i>									
Inhaled antibiotics									
Yes	23.1	50.2	64.9	66.8	73.3	69.2	70.8	61.5	56.4
No	76.9	49.1	34.8	32.0	23.0	23.1	23.6	35.6	41.8
Unknown (a)	0.0	0.8	0.4	1.2	3.6	7.7	5.6	2.9	1.8
<i>Total antibiotics users</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>
<i>Number</i>									
<i>Total antibiotics users</i>	<i>268</i>	<i>265</i>	<i>279</i>	<i>244</i>	<i>165</i>	<i>104</i>	<i>72</i>	<i>104</i>	<i>1,501</i>
Mode of use									
As needed (PRN)	90.3	90.8	81.0	64.2	81.3	85.7	83.0	93.1	81.4
Continuous	9.7	9.2	19.0	35.9	18.8	14.3	17.0	6.9	18.6
<i>Total inhaled antibiotics users(b)</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>
<i>Number</i>									
<i>Total inhaled antibiotics users</i>	<i>62</i>	<i>133</i>	<i>181</i>	<i>163</i>	<i>121</i>	<i>72</i>	<i>51</i>	<i>64</i>	<i>847</i>

(a) includes not recorded

(b) More than one mode of use can be recorded so numbers add to more than 100.0. As well, mode of use was not recorded for all patients where oral antibiotics were reported.

Almost all CF patients use a range of other therapies to manage conditions other than infections, and many take nutritional supplements. Therapies used by the highest proportions of patients include pancreatic enzymes (86% of children/adolescents and 88% of adults), vitamin supplements (75% and 80% respectively and bronchodilators (42% and 64%).

Table 4.6: ACFDR 2006: Other therapy by type

	Child/adolescent		Adult	
	Number	Per cent	Number	Per cent
Other therapies used:				
Yes	1010	96.0	579	98.6
No	42	4.0	8	1.4
<i>Total reported</i>	<i>1052</i>	<i>100.0</i>	<i>587</i>	<i>100.0</i>
Not reported	334		494	
<i>Total patients</i>	<i>1386</i>		<i>1081</i>	
Pulmozyme	268	25.5	263	44.8
Pancreatic enzymes	903	85.8	515	87.7
Vitamin supplements	786	74.7	466	79.4
Bronchodilators	442	42.0	375	63.9
Corticosteroids inhaled	233	22.1	254	43.3
Corticosteroids oral	84	8.0	65	11.1
Insulin	24	2.3	101	17.2
Macrolides	95	9.0	305	52.0
Salt tablets	349	33.2	140	23.9
Antihypercalcaemics	3	0.3	25	4.3
Gastric acid secretion reducers	111	10.6	154	26.2
Other	466	44.3	323	55.0
<i>Total reported</i>	<i>1052</i>	<i>100.0</i>	<i>587</i>	<i>100.0</i>

Note: individuals may use more than one type of therapy; percentages by type of therapy add to more than 100.0.

Nearly one quarter of CF patients also use nutritional supplements. The highest proportion, over one third of patients, occurred in the age groups 20 to 24 years and 30 to 34 years.

Table 4.7: ACFDR 2006: Nutritional supplements by age group

	0 to 4 years	5 to 9 years	10 to 14 years	15 to 19 years	20 to 24 years	25 to 30 years	30 to 34 years	35 + years	All ages
<i>Number</i>									
Oral (prescribed)	49	49	46	37	47	20	22	17	287
Nasogastric	6	1	2	1	6	4	4	0	24
TPN	3	0	1	0	0	0	0	0	4
Gastrostomy tube/button	5	10	17	25	14	3	1	1	76
Total using nutritional supplements	55	57	62	62	64	27	27	18	372
Not using nutritional supplements	243	214	231	196	109	73	39	81	1186
<i>Total reported</i>	298	271	293	258	173	100	66	99	1558
Not reported	79	108	98	118	148	110	101	147	909
<i>Total patients</i>	377	379	391	376	321	210	167	246	2467
<i>Per cent</i>									
Oral (prescribed)	16.4	18.1	15.7	14.3	27.2	20.0	33.3	17.2	18.4
Nasogastric	2.0	0.4	0.7	0.4	3.5	4.0	6.1	0.0	1.5
TPN	1.0	0.0	0.3	0.0	0.0	0.0	0.0	0.0	0.3
Gastrostomy tube/button	1.7	3.7	5.8	9.7	8.1	3.0	1.5	1.0	4.9
Total using nutritional supplements	18.5	21.0	21.2	24.0	37.0	27.0	40.9	18.2	23.9
Not using nutritional supplements	81.5	79.0	78.8	76.0	63.0	73.0	59.1	81.8	76.1
<i>Total reported</i>	100.0	100.0	100.0	100.0	100.0	100.0	100.0	100.0	100.0

Note: Individuals may use more than one type

Tables 4.8 and 4.9 show numbers of patients reported to have commenced or continued long term oxygen therapy or non-invasive ventilation during 2006. Numbers commencing are similar to 2005, but the large number of persons for which no information on this topic was reported (almost 35%) could be expected to contain others using oxygen or non-invasive ventilation therapy.

Table 4.8: ACFDR 2006: Oxygen therapy

	Child/adolescent		Adult		All Ages	
	Number	Per cent	Number	Per cent	Number	Per cent
Long-term oxygen therapy:						
Commenced during 2006	4	0.4	11	1.9	15	0.9
Commenced before 2006	2	0.2	6	1.1	8	0.5
No	1,024	98.9	540	94.7	1,564	97.5
Unknown	5	0.5	13	2.3	18	1.1
<i>Total reported</i>	<i>1,035</i>	<i>100.0</i>	<i>511</i>	<i>100.0</i>	<i>1,605</i>	<i>100.0</i>
Not reported	351		1,081		862	
<i>Total patients</i>	<i>1,386</i>		<i>1,104</i>		<i>2,467</i>	

Table 4.9: ACFDR 2006: Non-invasive ventilation

	Child/adolescent		Adult		All Ages	
	Number	Per cent	Number	Per cent	Number	Per cent
Commenced during 2005	0	0.0	4	0.7	4	0.3
Commenced before 2005	0	0.0	4	0.7	4	0.3
No	1,028	99.3	553	96.9	1,581	98.4
Unknown	7	0.7	10	1.8	17	1.1
<i>Total reported</i>	<i>1,035</i>	<i>100.0</i>	<i>571</i>	<i>100.0</i>	<i>1,606</i>	<i>100.0</i>
Not reported	351		510		861	
<i>Total patients</i>	<i>1,386</i>		<i>1,081</i>		<i>2,467</i>	

4.3 Hospital treatment

Not all CF patients experience hospitalisation during a year. The manner of collection of the data in 2006 does not allow a distinction to be drawn between 'no hospitalisation' and non-response. A total of 856 patients, or 35 per cent of patients recorded in the Data Registry, recorded at least one hospitalisation.

Table 4.10: ACFDR 2006: Hospitalisation, all causes

	Persons aged								All ages
	0 - 4 years	5 - 9 years	10 - 14 years	15 - 19 years	20 - 24 years	25 - 29 years	30 - 34 years	35 + years	
	<i>Per cent of persons in age group</i>								
Number of hospitalisations:									
1	57.1	63.6	53.4	38.8	33.3	47.9	35.6	45.0	48.7
2	28.0	16.7	22.1	25.6	25.0	21.1	22.2	33.3	24.1
3	11.3	10.6	11.5	12.4	21.7	14.1	20.0	13.3	13.7
4	1.2	3.8	6.1	10.9	6.7	7.0	15.6	3.3	6.0
5	0.6	3.8	2.3	7.8	5.8	2.8	0.0	3.3	3.5
6	0.6	0.8	3.1	1.6	4.2	5.6	4.4	1.7	2.3
More than 6	1.2	0.8	1.5	3.1	3.3	1.4	2.2	0.0	1.8
<i>Total</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>
	<i>Number of persons in age group</i>								
<i>Total</i>	<i>168</i>	<i>132</i>	<i>131</i>	<i>129</i>	<i>120</i>	<i>71</i>	<i>45</i>	<i>60</i>	<i>856</i>

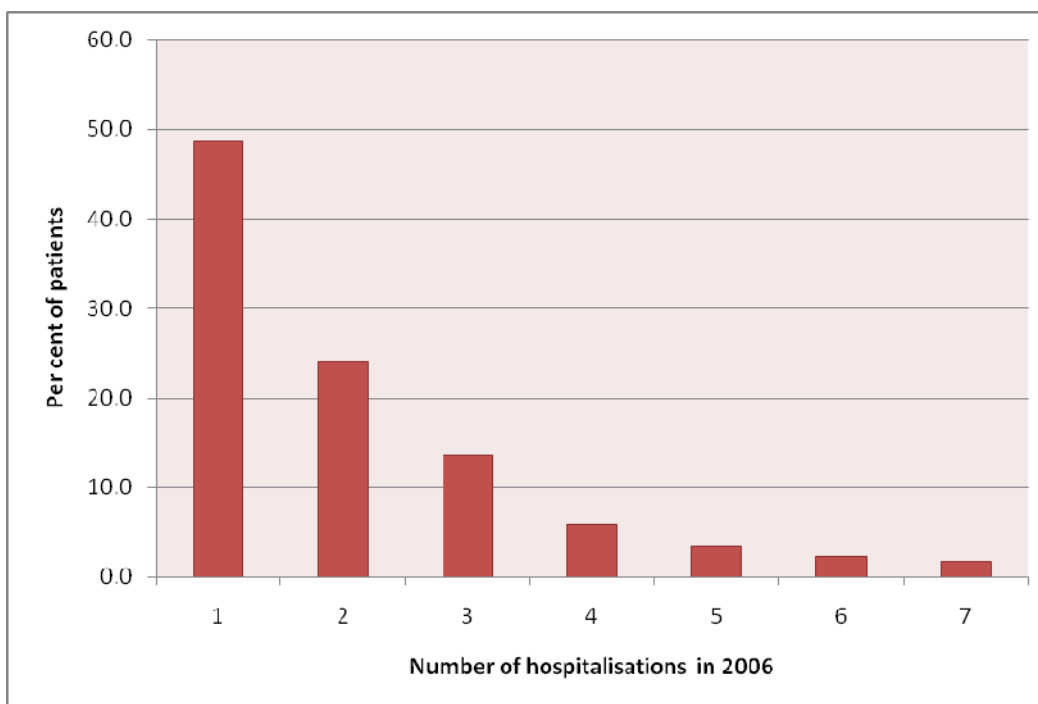


Figure 4.1: ACFDR 2006: Hospitalisations

Table 4.11: ACFDR 2006: Hospitalisation related to cystic fibrosis - respiratory

	Persons aged								All ages
	0 to 4 years	5 to 9 years	10 to 14 years	15 to 19 years	20 to 24 years	25 to 29 years	30 to 34 years	35 + years	
<i>Per cent of persons in age group</i>									
Number of hospitalisations:									
1	68.1	61.1	57.1	44.2	35.8	50.0	43.6	59.6	53.5
2	22.9	18.5	19.6	21.7	24.8	21.2	25.6	25.0	22.0
3	6.3	11.1	11.6	14.2	22.0	13.6	15.4	11.5	12.8
4	0.7	5.6	6.3	10.0	8.3	7.6	10.3	0.0	5.9
5	0.0	3.7	1.8	8.3	3.7	3.0	0.0	1.9	3.1
6	1.4	0.0	1.8	0.0	3.7	4.6	5.1	1.9	1.9
More than 6	0.7	0.0	1.8	1.7	1.8	0.0	0.0	0.0	0.9
<i>Total</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>
<i>Number of persons</i>									
<i>Total</i>	<i>144</i>	<i>108</i>	<i>112</i>	<i>120</i>	<i>109</i>	<i>66</i>	<i>39</i>	<i>52</i>	<i>750</i>

The total time spent in hospital over the year 2006 is reported in Table 4.12 for patients whose hospitalisation experience was reported. Mean and median times (20 and 14 days respectively) are comparable with 2005 and reinforce the fact that some patients spend a high proportion of their time hospitalised. For those reported as hospitalised in 2006, over half spent more than two weeks in hospital.

Table 4.12: ACFDR 2006: Total time in hospital for persons hospitalised

	Persons aged:								All ages
	0 to 4 years	5 to 9 years	10 to 14 years	15 to 19 years	20 to 24 years	25 to 29 years	30 to 34 years	35 + years	
<i>Per cent of persons in age group</i>									
Total time in hospital:									
Less than 1 week	52.9	37.9	19.9	9.9	8.3	13.7	20.0	21.7	25.6
From 1 to < 2 weeks	17.7	18.9	24.4	18.9	24.2	21.9	28.9	25.0	21.4
From 2 to < 4 weeks	20.6	26.5	29.8	34.1	28.3	39.7	20.0	31.7	28.4
4 weeks or more	8.8	16.7	26.0	37.1	39.2	24.7	31.1	21.7	24.6
<i>Total persons hospitalised</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>	<i>100.0</i>
<i>Number of persons</i>									
<i>Total persons hospitalised</i>	<i>170</i>	<i>132</i>	<i>131</i>	<i>132</i>	<i>120</i>	<i>73</i>	<i>45</i>	<i>60</i>	<i>863</i>
<i>Number of days</i>									
Mean	11.5	15.1	21.8	29.4	26.9	22.8	22.2	19.4	20.3
Median	5.0	11.0	14.0	21.0	23.0	17.0	14.0	14.0	14.0

4.4 Non-transplant surgery

Table 4.13 shows the age distribution of persons reported as having undergone selected non-transplant surgery during 2006. In view of the incompleteness of reporting, these numbers are likely to be under-estimates.

Table 4.13: ACFDR 2006: Non-transplant surgery during year

	0 to 1 years	2 to 5 years	6 to 10 years	11 to 17 years	18 years and over	Total
IV access devices	0	4	6	10	19	39
Gall bladder disease	0	0	0	1	4	5
Gastrostomy	1	1	2	1	4	9
Intestinal obstruction	9	0	2	2	2	15
Nasal (any surgery)	0	3	7	7	13	30
Other	9	19	11	16	31	86

5 Organ Transplants

5.1 Patients assessed for transplant in 2006

Cystic Fibrosis centres reported that 14 patients had been assessed for organ transplant during 2006. Assessments were for bilateral lung transplants (9 persons), combination heart/lung (3) and single lung (1). Organ type was not reported for the remaining patient assessed.

Table 5.1: ACFDR 2006: Patients assessed for transplant

Age group:	Males	Females	Persons
10 - 14 years	0	1	1
15 - 19 years	0	2	2
20 - 24 years	1	3	4
25 - 29 years	0	4	4
30 - 34 years	0	2	2
35 + years	0	1	1
<i>All ages</i>	<i>1</i>	<i>13</i>	<i>14</i>

5.2 Transplants during 2006

The number of organ transplants reported to the Data Registry for the reference year 2005 was 11, the same as in 2005. All of these patients survived the transplant operation.

The majority of the transplants completed (10) were bilateral lung type. The remaining patient had a heart/lung/liver transplant.

Table 5.2 ACFDR 2006: Patients receiving transplants

Age group:	Males	Females	Persons
15 - 19 years	0	2	2
20 - 24 years	1	3	4
25 - 29 years	0	2	2
30 - 34 years	1	1	2
35 + years	1	0	1
<i>All ages</i>	<i>3</i>	<i>8</i>	<i>11</i>

Note: Significant under-counting of both patients assessed and patients transplanted is suspect in the above tables. One major CF specialist centre treating adult patients has not reported transplant information for 2006.

6 Mortality

6.1 Deaths recorded in the Australian CF Data Registry

During 2006 there were 24 deaths of people with cystic fibrosis notified to the Australian CF Data Registry. Although this was 3 more than were reported in 2005, a retrospective review of deaths recorded found that some deaths had not been reported in several reference years, including 2006. The tables below incorporate information from the review.

There was one death reported of a person with CF who was aged less than 15 years. This death was due to a cause unrelated to CF. The median age at death for persons with CF who died in 2006 was 33 years.

Table 6.1: ACFDR 2006: Deaths, by age and sex

Age group:	Males	Females	Persons
0 - 4 years	1	0	1
15 - 19 years	0	2	2
20 - 24 years	1	3	4
25 - 30 years	2	1	3
30 - 35 years	2	1	3
35 + years	6	5	11
<i>All ages</i>	12	12	24

6.2 Causes of death

Table 6.2: ACFDR 2006: Cause of death

	Males	Females	Persons
Related to CF:			
Pulmonary	4	7	11
Other and not stated	6	5	11
Unrelated to CF	2	0	2
<i>All causes</i>	12	12	24

(a) Reported as 'post-transplant' but neither was not a proximate transplant death.

Appendix 1- Technical notes

Data collection and editing

Data for the Australasian Cystic Fibrosis Data Registry were collected from cystic fibrosis specialist treatment centres in Australia after the close of the reference year 2006. Participating centres extracted information from patient medical records. The new online data entry system introduced during data collection for the 2004 reference year was used for 2006. Some centres also used the Registry's data importing facility (using html and excel files) to upload clinical measures and microbiology data. Geoff Sims Consulting Pty Ltd extracted and tabulated information from the online database for this report. Data were processed using STATA software, Release 10. (StataCorp, College Station TX 77845 USA)

Collection instrument

The online data collection instrument arranges data into logical property groups accessible by opening tabs on a web page. The property groups are

- Patient and Survey Identification
- Birth/Diagnosis
- Clinic Visit and Clinical Measurements
- Microbiology
- Hospital and Home Therapy
- Treatment
- Complications
- Personal Information
- Transplant
- Death/Transfer

Contributing centres have secure access arrangements for entering data. Further information about data set contents and system specifications is available on request from CFA.

Identification and resolution of duplicate records

Duplicate records can occur where a patient is seen by more than one clinic within the reference year, either because of a transfer recorded during the year, because the patient re-establishes contact with a different clinic without referral or because the patient attends more than one clinic.

Duplicate records were identified through automated 'dedupe' procedures followed by visual examination of records with common initials (four letters from each person ID, comprising first two letters of last and first names) or common sex and dates of birth. Information from patient transfers between clinics as well as other personal characteristics were also used to identify possible duplicates.

Data derivation

Age derivations

The method of deriving patient age varied according to the analytical requirement. For demographic data, age was calculated by subtracting date of birth from the reference date, 31 December 2006 and rounding to completed years. Age at death and age at diagnosis were calculated in a similar manner, by deducting date of birth from the respective death and diagnosis dates and rounding to completed years.

Age groups are generally 5 year aggregations from birth or combinations thereof, as used in national health and demographic statistics.

Months of infant age, reported for age at diagnosis, were calculated as twelfths of a year (taken to be 365.25 days).

The age calculated for reporting of information about lung function and nutritional information is as at the date of best lung function or best nutritional status respectively, to permit accurate compilation of predicted values for lung function and population percentiles for height, weight and BMI.

Lung functions norms

Per cent predicted values for FEV1 and FVC for children and adolescents were calculated using the formula published by Wang et al (1993), for males aged 6 to 17 years and females aged 6 to 15 years and by Hankinson et al (1999) for males aged 18 years and over and females aged 16 years and over, according to recommendations adopted by the US Cystic Fibrosis Foundation for its National Patient Registry. Further information is available on request to CFA.

Height, weight and BMI norms

Population norms were calculated from the 'Nutstat' module of Epi Info 2002, a program developed by the Epidemiology Program Office of the US Centres for Disease Control and made available via its website – www.cdc.gov.

Registry data quality

Population coverage

Participating centres cover most of the specialist cystic fibrosis treatment centres in Australia and data are requested in respect of all of their patients. Nevertheless, there are shortfalls in coverage of the total Australian cystic fibrosis population.

Firstly, there are people with cystic fibrosis who do not attend specialist CF treatment centres. These include some who have had transplants and whose care is managed by a transplant centre, and an unknown number in private care.

Secondly some specialist treatment centres in major centres outside of the six State capital cities are not yet contributing data. These include adult centres in Newcastle and Canberra and paediatric clinics in Canberra and Darwin. Their participation is anticipated in the future but their absence from this report means that there will be a degree of under-coverage of the treatment centre population.

Thirdly, pressures on contributing centres have inevitably limited the completeness of reporting by some centres from time to time. Comments on the coverage of data are made at the beginning of each section of the report.

Data editing

Logical and range edit checks have been incorporated into the online data entry system, which presents real time queries to persons submitting data. Further checks and post-collection cleaning are undertaken in conjunction with processing the data for reporting.

References

Wang X, Dockery DW, Wypij D, Fay ME, and Ferris BG Jr (1993), *Pulmonary function between 6 and 18 years of age*, Paediatric Pulmonology 15:75-88.

Hankinson JL, Odencrantz JR and Fedan KB (1999), *Spirometric reference values from a sample of the general U.S. population*, Am J Respir Crit Care Med 159:179-187.

Research access

Requests for additional information from the Australasian Cystic Fibrosis Data Registry are welcome. Application should be made to Cystic Fibrosis Australia (CFA). In the case of more complex requests a fee may be charged to recover the cost of database services.

Bona-fide researchers proposing to undertake unit record analysis may be granted access to de-identified records, subject to ethics committee approval and researcher agreement to CFA's conditions of use. Interested researchers are advised to contact CFA for details and to arrange consideration of their research proposal.

Detailed information about data elements, coding schemes and detailed methodology can be provided on request to CFA.

All communication about additional data requirements and research access should be addressed to:

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