Cystic Fibrosis in Australia 2013

16TH ANNUAL REPORT AUSTRALIAN CYSTIC FIBROSIS DATA REGISTRY

CYSTIC FIBROSIS Australia

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16th Annual Report from the Australian Cystic Fibrosis Data Registry

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Preface

I am very pleased to deliver this 16th annual report from the Australian Cystic Fibrosis Data Registry, for the year 2013.

The cystic fibrosis population in Australia continues to grow, both in overall numbers and in its proportion of adults – now just a touch under 50 per cent. Continuing strong growth in Australia's adult CF population, of around 4 per cent per this year, is a feature that will not be news to Directors and staff at the adult CF treatment centres in Australia, but may not be as well recognised in the general community.

Cystic fibrosis is certainly no longer a disease of childhood. Children and adolescents with CF are now highly likely to survive into adulthood, a factor that will contribute to the growth in the adult population into the future.

Other good news in this report includes improvements over time in the lung function of child and adolescent cohorts, shown in the chart on page 22. Such messages in the data are strong motivation to CFA, enabled by the supporters acknowledged over the page, to maintain its strong collaboration with CF Centre Directors to provide this data registry resource. The registry sits alongside other collaborative activity that delivers Australian CF Standards of Care, peer reviews of CF centres and biennial Australasian CF Conferences, all playing a part to inform efforts to improve the life chances of people with cystic fibrosis.

CFA would not have the facts highlighted above, and the many other indicators that are reported herein, without the dedicated efforts of the staff at CF centres. I thank them for their efforts. Although this report is appearing somewhat later than anticipated, affected by staffing changes at the registry's contracted manager and at CFA, it is anticipated that more timely output will be reinstated soon. CFA owes this to the staff who enter the data promptly.

As a newly appointed CEO of Cystic Fibrosis Australia, I will make every effort to lobby government for support of the registry and related endeavours. I can point out that CFA's long-established and strong collaboration with the CF Centre Directors is delivering for CF care the kind of clinical quality management information that governments espouse in national health care reforms. Governments and the general population are clear stakeholders in this work.

Nettie Burke
Chief Executive Officer
Cystic Fibrosis Australia

April 2015

Acknowledgements

Many thanks go to the ACFDR Advisory Committee, whose members are:

Dr Scott Bell – The Prince Charles Hospital, Brisbane QLD
Dr Peter Bye – Royal Prince Alfred Hospital, Camperdown NSW
Dr Peter Cooper – The Children's Hospital, Westmead NSW
Dr Adam Jaffe – Sydney Children's Hospital
Dr Guy Marks – Liverpool Hospital NSW
Dr James Martin – Women's and Children's Hospital, Adelaide SA
Dr Sarath Ranganathan – Royal Children's Hospital, Melbourne VIC
Dr Phil Robinson – Royal Children's Hospital, Melbourne VIC
Dr Gerard Ryan – Sir Charles Gairdner Hospital, Perth WA

Further acknowledgement must go to the following people and organisations:

Ms Ann-Maree Bosch for logistical arrangements and minutes of proceedings of the Advisory Committee

Mr Geoff Sims and Miss Leticia Good of Australian Clinical Registries for database management and reporting

CFA also acknowledges generous support from **Mylan, Novartis, Roche, Vertex Pharmaceuticals** and the participants in **The Great Escape car rally for CF.**

Participating Centres

The ACFDR 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
Gosford Hospital
John Hunter Children's Hospital, Newcastle
John Hunter Hospital, Newcastle

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

Queensland
Royal Children's Hospital, Brisbane
The Prince Charles Hospital, Brisbane
Mater Hospital – Children, Brisbane
Mater Hospital – Adults, Brisbane
Gold Coast University Hospital, Southport

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

Australian Capital Territory
The Canberra Hospital

Contents

1 People with cystic fibrosis

1.1 Overview

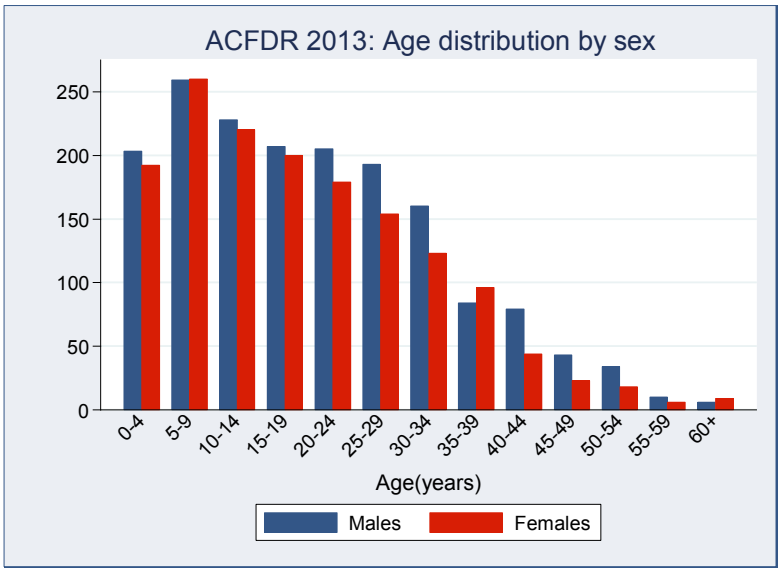
At 31 December 2013 the Australian Cystic Fibrosis Data Registry (ACFDR) held records of 3,235 people with cystic fibrosis.

The mean age of the registry population was 20.0 years at 31 December 2013. This was up from 19.5 years in 2012, following steady increases over preceding years. Reflecting this trend, the proportion of the registry population that is adult (18 years and over) increased to 49.9 per cent in 2013, from 49.4 per cent in 2012. Only one third of patients were adult when the registry commenced in 1998.

The median age of 17.9 years at 31 December 2013 is slightly higher than at the end of previous years, having been 17.7 in 2012 and 17.6 in 2011. Median age for males (18.9 years) remained higher than that for females (17.1 years) in 2013.

Note that some earlier year figures quoted in the previous paragraphs have had minor revision since first being published.

An increase of 80 in the overall number of registrants in 2013 is not only due to an excess of new diagnoses (92) over deaths (38) reported for the year. There were 11 new patients who had been diagnosed with CF in 2012 but not reported until 2013. Four patients withdrew their consent to reporting of their data by their CF treatment centre and eight were removed because they are no longer considered to have CF. The change also includes the net effect of patients arriving from and leaving for overseas countries, patients lost to follow-up in 2013 and returning patients not reported as current during 2012.



1.2 Age distribution

ACFDR 2013: Age and sex of registrants at 31 December 2013

Age group	Males	Females	Persons	Per cent male
Standard demographic age groups:				
0 - 4 years	203	192	395	51.4
5 - 9 years	259	260	519	49.9
10 - 14 years	228	220	448	50.9
15 - 19 years	207	200	407	50.9
20 - 24 years	205	179	384	53.4
25 - 29 years	193	154	347	55.6
30 - 34 years	160	123	283	56.5
35 - 39 years	84	96	180	46.7
40 - 44 years	79	44	123	64.2
45 - 49 years	43	23	66	65.2
50 - 54 years	34	18	52	65.4
55 - 59 years	10	6	16	62.5
60 + years	6	9	15	40.0
Alternative CF age groups and totals:				
0 - 1 years	76	62	138	55.1
2 - 5 years	177	182	359	49.3
6 - 11 years	302	290	592	51.0
12 - 17 years	267	266	533	50.1
Children and adolescents	*822*	*800*	*1622*	*50.7*
18 - 29 years	473	405	878	53.9
30 + years	416	319	735	56.6
Adults	*889*	*724*	*1613*	*55.1*
Total, all ages	*1,711*	*1,524*	*3,235*	*52.9*

The lower table area shows age dissections that have been recommended for international comparison of CF data. Many of the tables and charts later in this report use this age dissection.

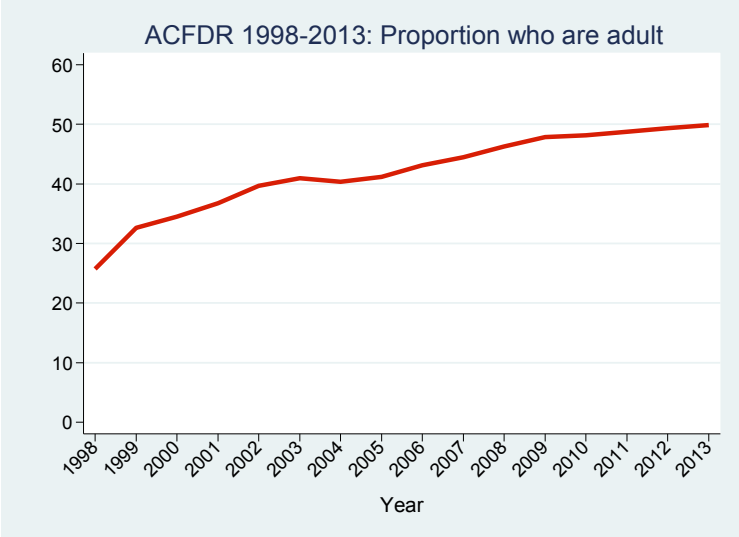
The proportion of males in the Australian CF population shows generally better survival of males in the Australian CF population, which reflects international experience.

At 31 December 2013, males made up 52.9 per cent and females 47.1 per cent of the ACFDR population. This has remained a consistent proportion since establishment of the registry in 1998. The proportion of males is higher amongst the adult population (55.1%) than in the child and adolescent population (50.7%).

The proportion of adults in the Registry as a whole was 49.9 per cent at 31 December 2013. A lower proportion shown for the Australian Capital Territory in 2013 reflects reporting issues.

ACFDR 31 December 2013: Adult status by State/Territory of residence

State or Territory of residence	Child/adolescent	Adult	Total	Per cent adult
New South Wales	493	474	967	49.0
Victoria	320	369	689	53.6
Queensland	401	395	796	49.6
Western Australia	184	169	353	47.9
South Australia	146	141	287	49.1
Tasmania	50	52	102	51.0
Australian Capital Territory	20	8	28	28.6
Northern Territory	3	5	8	62.5
Overseas	5	0	5	0.0
Total	*1,622*	*1,613*	*3,235*	*49.9*



1.3 Adult marital status, education and activity

Improved disease management allows an increasing proportion of persons with CF to continue normal activities into their adult life.

Thirty eight per cent of male adult patients and 48 per cent of adult female patients for whom marital status was reported were in a formal or informal marriage relationship. One adult centre was excluded from this analysis because of almost incomplete reporting. Even after this exclusion, marital status was either unknown or not reported for around one quarter of adults, as three other centres had data missing for respectively one half, one third and one quarter of their patients.

Sixteen per cent of adult CF patients for whom parenthood status was reported (75% of the adult population) had children. Three centres did not report parenthood status for, respectively, 58, 45 and 24 per cent their adult patients.

ACFDR 31 December 2013: Marital status of adults[(a)]

	Males		Females	
Marital status	**Number**	**Per cent**	**Number**	**Per cent**
Married (includes de facto)	230	38.0	245	47.9
Not married	376	62.0	267	52.1
		100.0		*100.0*
Unknown (includes not recorded at some centres)	196	24.4	*162*	24.0
Total	*802*		*674*	

(a) One centre was excluded from the analysis.

Many people with cystic fibrosis continue with education beyond senior secondary school level, with 19 per cent of adult CF patients for whom educational attainment was reported having university qualifications and a further 21 per cent having completed other study beyond high school. Data presented in the table exclude 3 centres that did not report education attainment for more than half of their patients.

Sixty five per cent of adults with CF were in either full-time or part-time paid employment during 2013. Four centres for whom activity status was either missing or unknown (mainly the former) for over half of their patients were excluded from the analysis. One other had missing data for about one quarter of its patients.

ACFDR 31 December 2013: Educational attainment of adults[(a)]

	Number	**Per cent**
Junior Secondary (Year 10)	98	13.9
Senior Secondary (Year 12)	311	44.2
Tertiary Certificate or Diploma	146	20.8
University Degree	134	19.1
Left school prior to Year 10	3	0.4
Not applicable (b)	11	1.6
Total reported	*703*	*100.0*
Unknown/not reported (incl. as % of total below)	377	34.9
Total	*1,080*	*100.0*

(a) Three CF centres were excluded from analysis
(b) Currently studying

ACFDR 31 December 2013: Activity status of adults [(a)]

	Number	**Per cent**
Employed, full time paid	278	36.6
Employed, part time paid	214	28.2
Voluntary work only	5	0.7
Unemployed	47	6.2
Pensioner	68	9.0
Others not in labour force (b)	148	19.5
Total reported	*760*	*100.0*
Unknown/not reported (incl. as % of total below)	148	16.3
Total	*908*	*100.0*

(a) Four CF centres were excluded from analysis
(b) includes homemakers, students

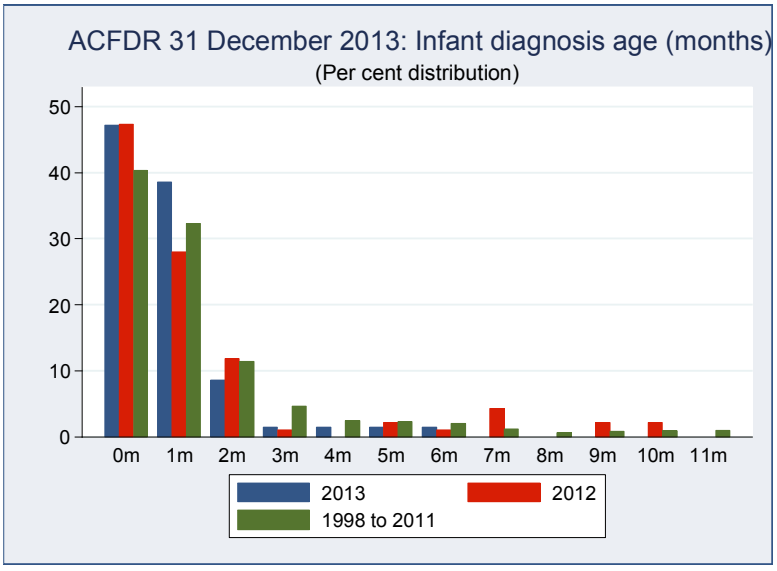
2 Diagnosis

2.1 Overview

The number of new diagnoses of cystic fibrosis (CF) notified to the Registry for 2013 was 92, including 80 diagnosed less than one year of age.

2.2 Age at diagnosis

All but 4 of the infant diagnoses where a diagnosis date was reported (70 out of the 80 new infant diagnoses) were completed by three months of age, assisted by neonatal screening programs that operate in all States and Territories of Australia. There were 10 infant diagnoses in 2013 where a diagnosis date was not reported but where the fact of infant diagnosis was inferred from age less than 12 months at the end of the year.



Australian CF centres reported 5 new cases that were diagnosed in early childhood (1 to 4 years), 1 aged 10 to 14 years and 6 aged 20 years and over.

2.3 Presentation and diagnosis

Over 70 per cent of new cases of CF diagnosed in 2013 included neonatal screening as a mode of presentation and 11 per cent reported meconium ileus.

Respiratory symptoms were reported in 12 per cent, and gastrointestinal symptoms in 3 per cent.

ACFDR 31 December 2013: Mode of presentation [a] by year of diagnosis

	All years	2013	All years	2013
	Number		*Per cent*	
Neonatal screening	1,649	65	53.6	71.4
Respiratory symptoms	463	11	15.0	12.1
Gastrointestinal symptoms	365	3	11.9	3.3
Meconium ileus	405	10	13.2	11.0
CF sibling	243	8	7.9	8.8
Minor manifestations	30	0	1.0	0.0
Pre-natal diagnosis	44	3	1.4	3.3
Infertility	18	0	0.6	0.0
Other	320	7	10.4	7.7
Unknown (b)	157	1		
Total	*3,235*	*92*	*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.

2.4 Pancreatic insufficiency

The overall proportion of patients who are pancreatic insufficient is 81.1 percent, based on consolidated data across all years of reporting. Related information later in this report show that 85 per cent of adults and 79 per cent of children/adolescents are using pancreatic enzymes

2.5 Genotyping

Mutation information consolidated across reporting years was available for 92.9 per cent of patients in the registry at the end of 2013. More detailed tables follow in the following two pages.

ACFDR 31 December 2013: Whether patient genotyped, by year of diagnosis

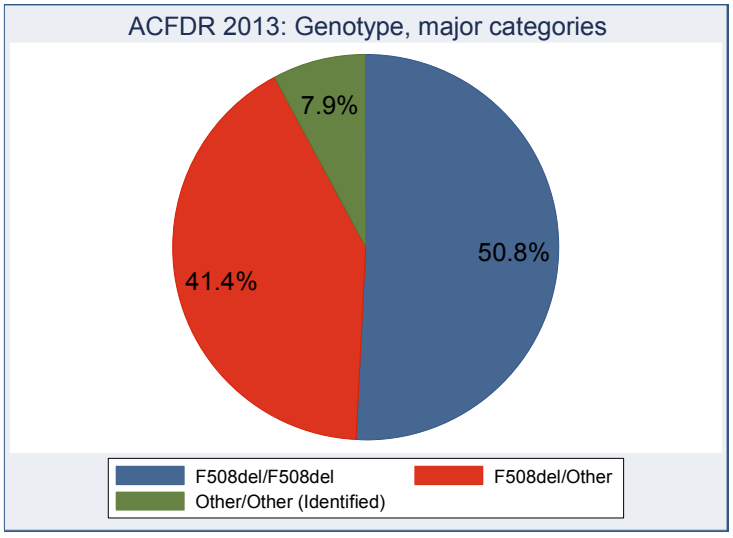
	All years	2013	All years	2013
Genotyped	3,006	82	92.9	89.1
Not genotyped	55	1	1.7	1.1
Unknown/Not recorded	174	9	5.4	9.8
Total	*3,235*	*92*	*100.0*	*100.0*

The genetic mutation F508del has been identified as at least one of the paired mutations responsible for the inheritance of cystic fibrosis in 92.1 per cent of patients for whom genotype details have been reported. Over half (50.8%) are reported as homozygous for F508del. G551D was the next most prevalent mutation, with 7.7 per cent of the CF population having this mutation, mostly in combination with F508del.

ACFDR 31 December 2013: Genotype[a]

	Mutation 1										
	F508 del	G 542 X	G 551 D	N 1303 K	W 1282 X	R 117 H	1717 -1G ->A	621 +1G ->T	Other NEC	Unk-nown	Total
	Per cent										
Mutation 2:											
F508del	50.8										50.8
G542X	1.9	0.1									2.0
G551D	6.0	0.2	0.2								6.4
N1303K	1.1	0.1	0.1	0.1							1.3
W1282X	0.5	0.0	0.0	0.0	0.2						0.7
R117H	3.2	0.0	0.1	0.0	0.0	0.1					3.4
1717-1G->A	1.3	0.0	0.0	0.0	0.0	0.0	0.0				1.4
621+1G->T	1.1	0.1	0.0	0.0	0.0	0.0	0.0	0.0			1.3
Other NEC	14.7	0.2	0.6	0.1	0.1	0.2	0.2	0.2	1.5		17.7
Unknown	11.4	0.3	0.5	0.0	0.0	0.1	0.0	0.0	1.0	1.6	14.9
Total	*92.1*	*1.0*	*1.5*	*0.2*	*0.3*	*0.4*	*0.2*	*0.2*	*2.6*	*1.6*	*100.0*

(a) Patients with missing genotype data for both alleles were excluded from analysis



The table over the page shows population and allele prevalence of the most common CFTR mutations found in the Australian CF population. A more extended list is available on request. Just 21 mutations have a population prevalence of 10 or more in Australia.

ACFDR 2013: Patients and alleles – most common CFTR mutations in Australian CF population

CFTR Mutation	Patient Number	Patient Per cent	Homozygous Patient Number	Allele Number
F508del	2,770	92.1	1,527	4,297
G551D	230	7.7	6	236
R117H	113	3.8	2	115
G542X	87	2.9	3	90
1717-1G->A	48	1.6	0	48
N1303K	43	1.4	2	45
621+1G->T	44	1.5	1	45
W1282X	25	0.8	5	30
R553X	22	0.7	0	22
5T	21	0.7	0	21
D1152H	17	0.6	0	17
P67L	17	0.6	0	17
2789+2insA	16	0.5	0	16
G85E	12	0.4	0	12
Q493X	12	0.4	0	12
E60X	11	0.4	1	12
R560T	11	0.4	0	11
R1162X	10	0.3	0	10
I507del	10	0.3	0	10
R334W	10	0.3	0	10
1078delT	10	0.3	0	10
2789+5G->A	9	0.3	0	9
1898+1G->A	9	0.3	0	9
3659delC	9	0.3	0	9
3272-26A->G	9	0.3	0	9
V520F	9	0.3	0	9
3849+10kbC->T	8	0.3	0	8
R347P	8	0.3	0	8
A455E	8	0.3	0	8
2183AA->G	4	0.1	3	7
2184delA	7	0.2	0	7
S549N	7	0.2	0	7
R1066C	6	0.2	0	6
1154insTC	6	0.2	0	6
R75Q	4	0.1	1	5
394delTT	4	0.1	0	4
R347H	4	0.1	0	4
R352Q	4	0.1	0	4
2622+1G->A	4	0.1	0	4
711+3A->G	4	0.1	0	4
3791delC	4	0.1	0	4
R1070W	4	0.1	0	4
Other mutations , not listed above	281	9.3		
Unknown mutation	449	14.9		
Total patients genotyped	*3,006*	*100.0*		

3 Health and functioning

Information in this chapter covers respiratory infections, medical complications, lung function and nutritional measures. Two adult centres did not submit microbiology information for their patients in 2013 and are excluded from the analysis. All centres submitted data for multiple occasions where clinical measures were taken - height, weight and lung function.

3.1 Respiratory infections

Patients who were tested for respiratory infections in 2013 had a mean of 3.7 tests of all types during the year. The median number of tests was 3, with little variation across age groups. Two adult centres did not provide data for respiratory infections and were excluded from the analysis.

ACFDR 2013: Number of sputum and BAL/bronchoscopy cultures[a]

	0 - 1 years	2 - 5 years	6 - 11 years	12 - 17 years	18 - 29 years	30 + years	All ages
			Per cent of patients tested (b)				
Sputum cultures:							
None	49.0	36.8	14.0	1.6	0.6	2.1	11.1
1	9.6	17.7	15.9	16.4	25.1	24.6	19.6
2	1.9	8.4	13.4	13.8	17.9	19.8	14.4
3	8.7	9.7	10.6	12.2	15.0	17.2	12.9
4	7.7	8.7	8.9	13.1	9.3	10.9	10.1
5	8.7	7.4	13.2	12.2	7.8	6.1	9.5
6	8.7	4.0	8.7	10.5	7.4	6.1	7.7
7 or more	5.8	7.4	15.3	20.3	16.9	13.2	14.8
Total	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*
BAL/bronchoscopy:							
None	59.6	58.5	88.8	97.2	99.4	97.6	89.0
1	34.6	35.5	10.2	2.1	0.6	1.6	9.4
2	4.8	5.0	1.1	0.7	0.0	0.5	1.4
3 or more	1.0	1.0	0.0	0.0	0.0	0.3	0.2
Total	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*
			Number of patients				
Patients tested (b)	*104*	*299*	*471*	*428*	*526*	*378*	*2,206*
Culture not done	34	60	121	105	146	165	631
Total reported	*138*	*359*	*591*	*533*	*672*	*542*	*2,835*
Not reported	0	0	1	0	0	1	2
Total patients	*138*	*359*	*592*	*533*	*672*	*543*	*2,837*

(a) Two adult CF centres were excluded from analysis
(b) By any method of obtaining culture.

The table on the previous page shows the distribution of CF patients according to the number of both sputum and BAL/bronchoscopy samples examined during 2013. The latter method is used mainly on smaller children. Taking sputum samples alone, more than two thirds (69 per cent) of the patients tested had at least two sputum samples in 2013. Respiratory cultures were not done for 22 per cent of patients, including for about one third of patients aged 30 years and over.

Denominators for percentages shown in the tables and chart in this section are the total number of persons for whom any respiratory culture results were reported, from any method of testing.

The most commonly identified organisms in respiratory specimens are various species and forms of *Pseudomonas*. It can be seen that half of all patients who were tested produced positive *Pseudomonas aeruginosa* cultures, with the mucoid form showing in one third. Its prevalence is greater in adult patients. The table shows that 56 per cent of patients aged 18 to 29 years and 76 percent of those aged 30 years and over produced samples indicating the mucoid form of *Pseudomonas aeruginosa*, two to three times the proportion for adolescents and much higher than that for children.

ACFDR 2013: Pseudomonas infection by age group[(a)(b)]

	0 - 1 years	2 - 5 years	6 - 11 years	12 - 17 years	18 - 29 years	30 + years	All ages
				Per cent			
Pseudomonas aeruginosa:							
Mucoid	0.0	2.3	7.4	26.9	55.9	75.9	33.5
Rough/non-mucoid	2.9	10.7	11.3	21.5	30.4	31.5	20.8
Not differentiated	4.8	11.4	12.7	21.3	23.0	24.1	18.2
Any Ps aeruginosa	7.7	21.1	24.8	45.6	72.6	87.0	49.6
Pseudomonas other species	1.9	0.7	1.1	2.6	3.4	2.4	2.1
				Number of patients			
Patients tested	*104*	*299*	*471*	*428*	*526*	*378*	*2,206*

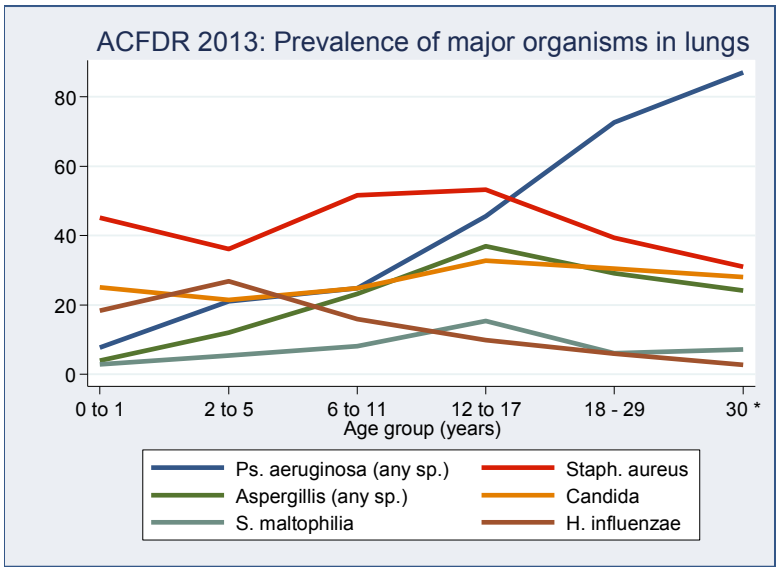
(a) Two adult CF centres were excluded from analysis
(b) Patient may have had more than one type of Pseudomonas infection. Percentages for individual types may add to more than totals.

While prevalence of *Pseudomonas* organisms is lower in children than in adults, although increasing with rising age (see table above), young children are more likely than adult patients to produce cultures showing presence of *Staphylococcus aureus* (see table opposite). Half of all child patients and adolescent patients aged 6 to 17 years had this bacterial infection. *Haemophilus influenzae* is also evident in relatively high proportions of child patients, highest in children aged from 2 to 5 years, where this organism was cultured for over one quarter of children. The youngest age groups also had the highest proportions with positive cultures of the bacteria *Escherichia coli*; 26 per cent for those aged less than 2 years being the highest.

ACFDR 2013: Other respiratory culture by age group (a)

	0 - 1 years	2 - 5 years	6 - 11 years	12 - 17 years	18 - 29 years	30 + years	Total
			Per cent of patients tested (b)				
Bacteria:							
Staphylococcus aureus	45.2	36.1	51.6	53.3	39.4	31.0	43.1
Haemophilus influenzae	18.3	26.8	15.9	9.8	5.9	2.6	11.7
Burkholderia cepacia (Ps cepacia)	0.0	0.3	1.5	2.3	3.4	4.0	2.3
Stenotrophomonas maltophilia	2.9	5.4	8.1	15.4	6.1	7.1	8.3
Escherichia coli	26.0	7.0	4.9	2.3	0.8	0.5	3.9
MRSA (c)	1.0	2.0	2.8	3.5	4.0	4.0	3.2
Alcaligenes xylosoxidans	0.0	0.7	3.0	4.4	5.7	4.2	3.7
Serratia marcescens	1.0	1.7	0.8	0.7	0.8	1.1	1.0
Klebsiella (any species)	7.7	1.3	0.4	0.5	0.2	0.3	0.8
Non-tuberculous mycobacterium	0.0	0.3	0.4	3.0	2.3	3.4	1.9
Fungi:							
Candida	25.0	21.4	24.8	32.7	30.4	28.0	27.8
Aspergillus (any species)	3.8	12.0	23.1	36.9	29.1	24.1	25.0
Scediosporium (any species)	0.0	0.7	5.5	6.5	3.8	3.7	4.1
Other organisms not listed above	29.8	34.4	28.7	25.7	15.0	15.1	23.3
Normal flora only	84.6	78.9	80.5	78.7	35.4	28.6	60.5
No growth/sterile culture	*13.5*	*10.7*	*8.7*	*5.1*	*4.8*	*5.3*	*7.0*
			Number of patients				
Patients tested	*104*	*299*	*471*	*428*	*526*	*378*	*2,206*

(a) Two adult CF Centres were excluded from analysis
(b) Note: Patients may have multiple infections during the year. Percentages may add to more than 100.0.
(c) Methicillin-resistant Staphylococcus aureus



3.2 Other medical complications

Complications data were again subject to under-reporting in 2013, with 3 centres that reported complication data for less than 50 percent of their patients being excluded from the analysis. Prevalence of medical complications increases with age in CF patients. For instance, 37 per cent of adult patients suffer gastro-oesophageal reflux, over one quarter of patients aged 30 years and over experience chronic insulin-dependent diabetes and almost 40 per cent of the same age group have osteoporosis or osteopenia.

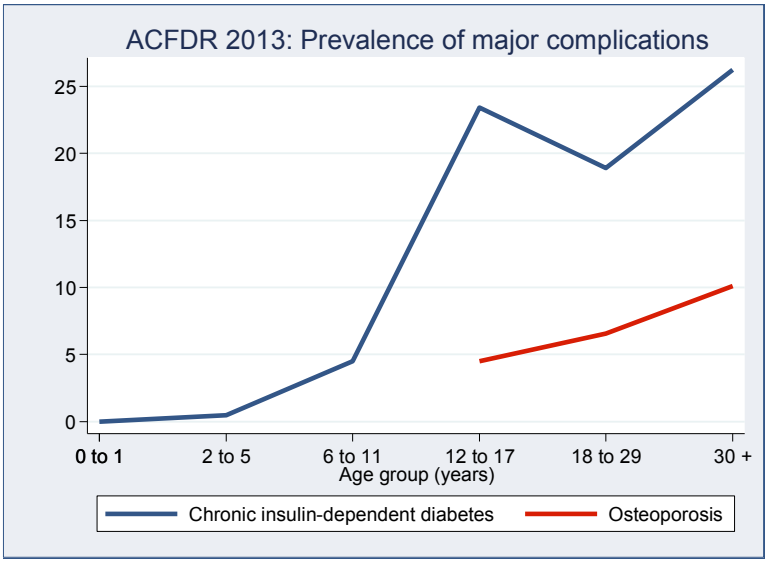
The proportion for which none of the selected complications shown in the following table have been reported is over 85 per cent for children less than 6 years, but declines to 17 per cent in CF patients aged 30 and over.

ACFDR 2013: Medical complications[(a)]

	0 - 1 years	2 - 5 years	6 - 11 years	12 - 17 years	18 - 29 years	30 + years	Total
				Per cent			
Pulmonary:							
Major haemoptysis	0.0	0.5	0.3	1.3	6.5	7.8	4.0
Massive haemoptysis	1.2	0.0	0.0	0.3	1.1	1.6	0.8
Therapeutic bronchial artery embolisation	0.0	0.0	0.0	0.6	2.2	2.7	1.3
Pneumothorax	1.2	0.0	0.3	2.6	6.4	10.3	4.7
Any pulmonary above	1.2	0.5	0.6	4.2	12.7	19.3	8.9
Gastro-intestinal:							
Gastro-oesophageal reflux	11.9	6.2	8.1	12.5	36.7	37.4	23.7
- proven at endoscopy	1.2	1.9	2.4	4.2	5.1	4.0	3.7
Abnormal liver function test	3.6	6.2	12.3	17.0	27.1	22.2	18.5
Cirrhosis or portal hypertension	0.0	0.0	1.8	5.4	4.5	1.8	2.9
Pancreatitis	1.2	0.0	0.0	0.0	1.1	2.5	0.9
Any Gastro-intestinal above	14.3	11.4	18.6	26.6	54.4	49.8	36.3
Endocrine:							
Chronic insulin-dependent diabetes	0.0	0.5	4.5	23.4	18.9	26.2	16.0
Intermittent insulin-dependent diabetes	0.0	0.0	1.5	2.6	2.9	1.8	1.9
Other glucose abnormality	0.0	0.0	7.5	16.0	17.8	13.9	12.1
Any Endocrine above	0.0	0.5	12.9	39.7	38.4	40.1	28.8
Osteo:							
Osteoporosis	0.0	0.0	0.6	4.5	6.5	10.1	5.0
Osteopenia	0.0	0.0	1.2	13.5	21.5	28.7	15.1
Fracture this year	0.0	0.0	0.0	0.0	1.3	1.6	0.7
Any Osteo above	0.0	0.0	1.2	14.1	26.4	38.8	18.9
Other:							
Cancer	0.0	0.0	0.3	0.3	0.4	0.7	0.4
None of the above	85.7	87.7	70.0	38.8	20.4	17.0	41.3
Total reported (b)	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*
				Number			
Total reported	*84*	*211*	*333*	*312*	*550*	*446*	*1,936*
Unknown or not stated	10	61	100	95	311	283	860
Total patients (a)	*94*	*272*	*433*	*407*	*861*	*729*	*2,796*

(a) Two paediatric CF centres and one small mixed centre were excluded from analysis
(b) Patient may have had more than one complication. Percentages add to more than 100.0.

Although some prevalence of osteoporosis at younger ages is reported in the table above, this is not displayed on the following chart because of uncertainty about diagnosis at younger ages.



3.3 Lung function

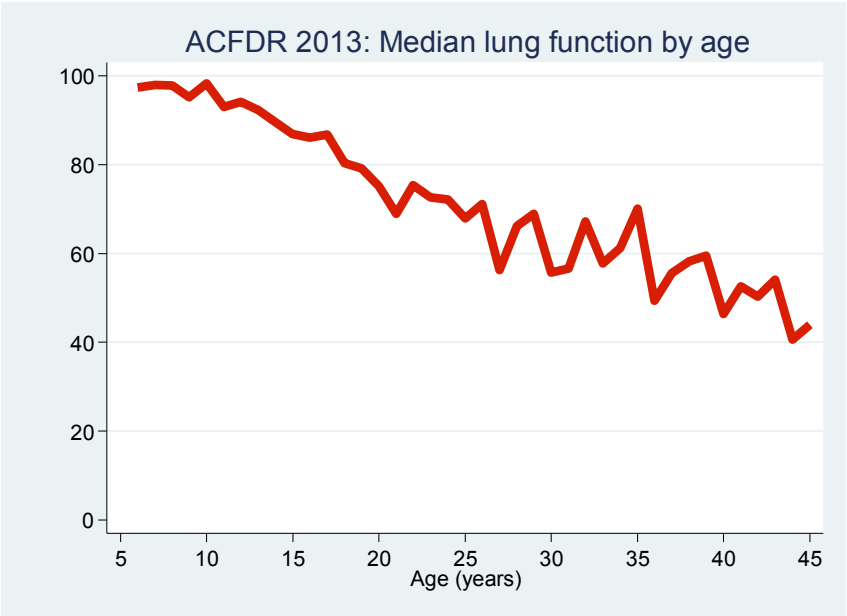
Lung function measures compiled for this report are aligned with methods used in the United States' Cystic Fibrosis Foundation's Patient Registry, that is the lung function measure included for each patient is the average of the highest FEV_1 per cent predicted value recorded in each quarter of the year.

Lung Function categories described in tables and charts:

- Normal – 90% of predicted FEV_1 and above
- Mild impairment – at least 70% but below 90% of predicted FEV_1
- Moderate impairment – at least 40% but below 70% of predicted FEV_1
- Severe impairment – below 40% of predicted FEV_1

Predicted values are based on Wang and Hankinson formulae – see References

Median CF lung function measured as FEV_1 percent predicted is within the normal range for young children but is lower than 70 per cent of normal, the level at which moderate lung function impairment is experienced, in adult age ranges. Fewer than 5 per cent of children aged 6 to 11 years have FEV_1 values that are below 70 per cent of predicted values but 16 per cent of older children and adolescents are in this category.

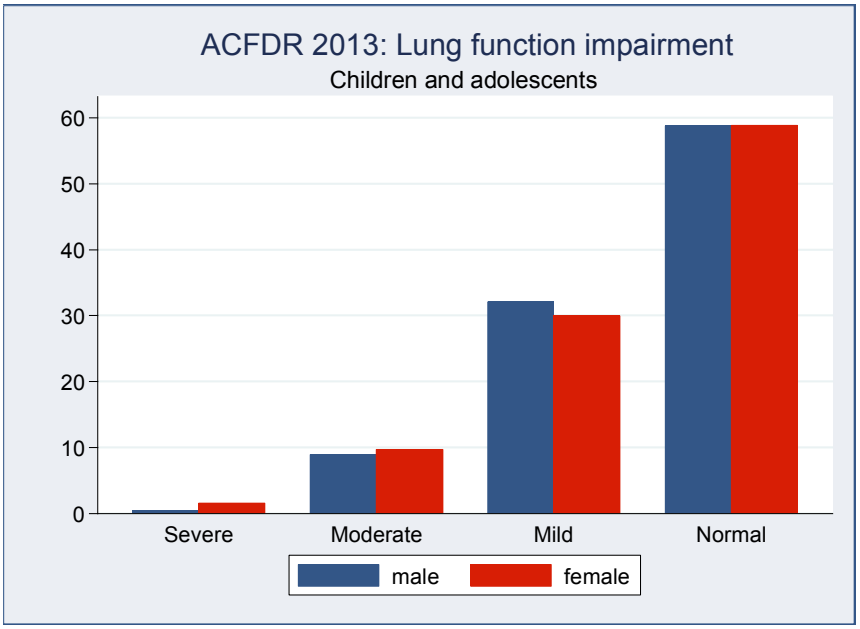


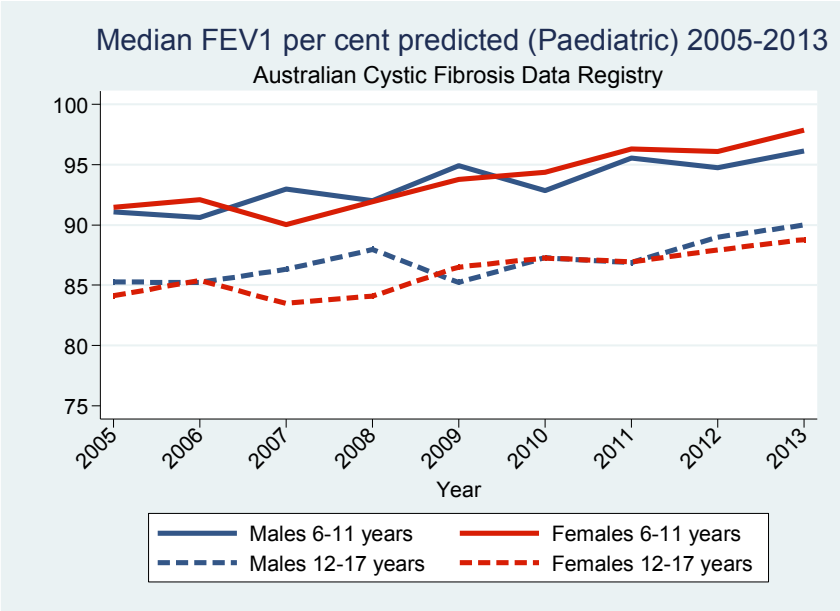
Generally greater proportions of patients have severe lung function impairment in successive older age groups. The proportion of adult male patients with severe lung function impairment (19%) is greater than the proportion of female patients in this severity category (12%). Both proportions are lower than those reported in 2013 (21% and 14%, respectively). Details for finer age groups in 2013 are in the table opposite.

ACFDR 2013: Lung function impairment by age group and sex

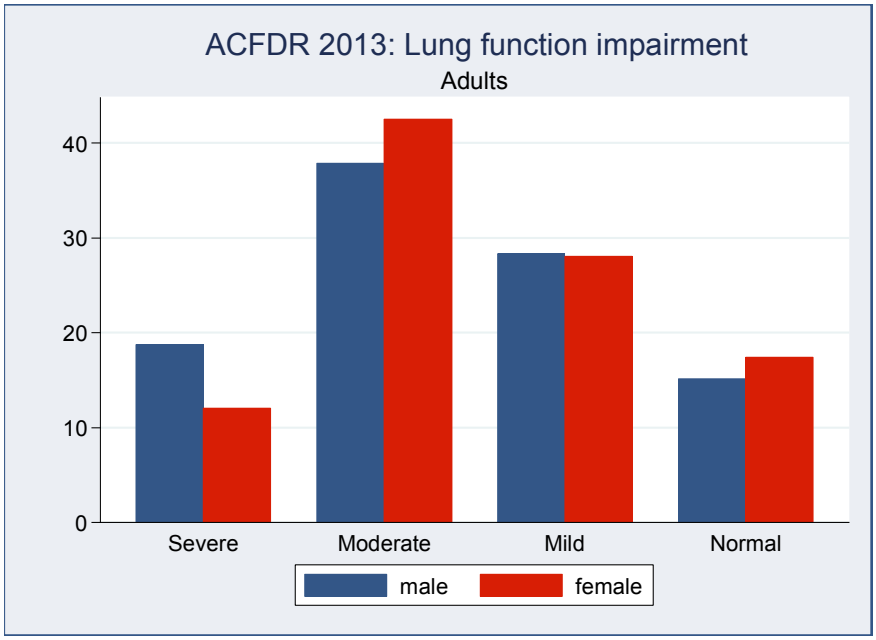
	Severe	Moderate	Mild	Normal	Total	Severe	Moderate	Mild	Normal	Total
			Number					*Per cent*		
Males:										
6 - 11 years	0	11	80	186	277	0.0	4.0	28.9	67.2	100.0
12 - 17 years	2	37	93	131	263	0.8	14.1	35.4	49.8	100.0
18 - 29 years	49	150	145	89	433	11.3	34.6	33.5	20.6	100.0
30 + years	97	145	76	29	347	28.0	41.8	21.9	8.4	100.0
Total measured	*148*	*343*	*394*	*435*	*1,320*	*11.2*	*26.0*	*29.9*	*33.0*	*100.0*
Females:										
6 - 11 years	2	11	61	185	259	0.8	4.3	23.6	71.4	100.0
12 - 17 years	6	39	94	119	258	2.3	15.1	36.4	46.1	100.0
18 - 29 years	33	144	108	81	366	9.0	39.3	29.5	22.1	100.0
30 + years	41	117	64	26	248	16.5	47.2	25.8	10.5	100.0
Total measured	*82*	*311*	*327*	*411*	*1,131*	*7.3*	*27.5*	*28.9*	*36.3*	*100.0*
Persons:										
Total measured	*230*	*654*	*721*	*846*	*2,451*	*9.4*	*26.7*	*29.4*	*34.5*	*100.0*

The following chart shows categories of lung function impairment experienced by the child and adolescent CF population as a whole. Fifty nine per cent of children and adolescents had lung function within the normal range, the same for males and females. This is a higher proportion than was reported for either males (56 per cent) or females (54 per cent) in 2012 and reflects the continuation of improving trends in lung function that are shown for broad age groups in the chart at the top of the following page.





For adults with cystic fibrosis, a different pattern of lung function impairment is evident, with just 15 per cent of adult males and 17 per cent of adult females having normal lung function in 2013. Severe lung function impairment was experience by 19 per cent of male adults and 12 per cent of female adults.



Trend data for adult lung function (not shown) indicate improvement for the 18 to 29 years age group since 2005, though less marked than for children and adolescents. A flatter trend for adults aged 30 and over may be confounded by possible increased survival within this age group.

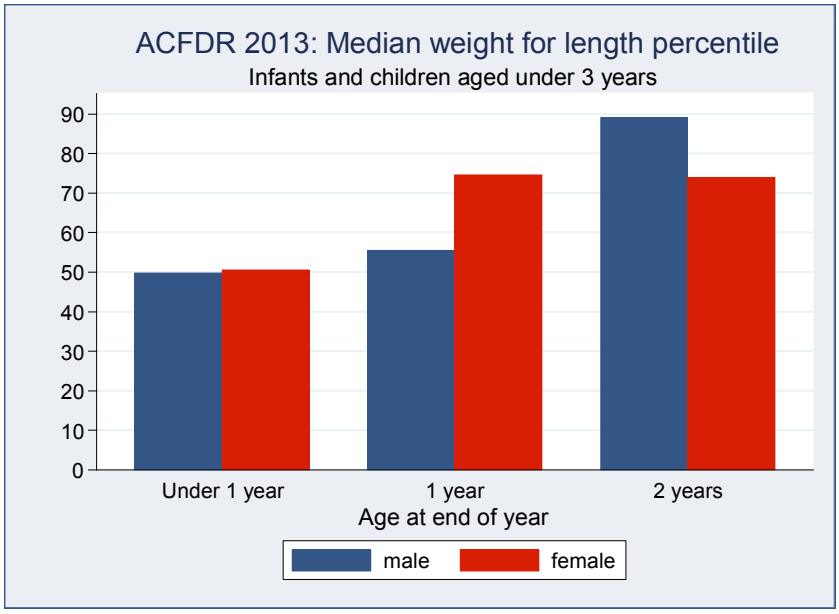
3.4 Nutrition: height and body mass index

Methodological note

As for lung function measures reported above, values reported in this section are compiled from each patients average of their highest value recorded in each quarter of the year.

Infants and young children aged under 3 years

Nutritional outcomes for children aged under 3 years have been compiled for the first time for this report. The median value of weight for length is at the 50th percentile for both male and female infants and at higher percentiles for those aged 1 and 2 years. Percentiles are derived from World Health Organisation Child Growth Standards (WHO 2006) – see References.



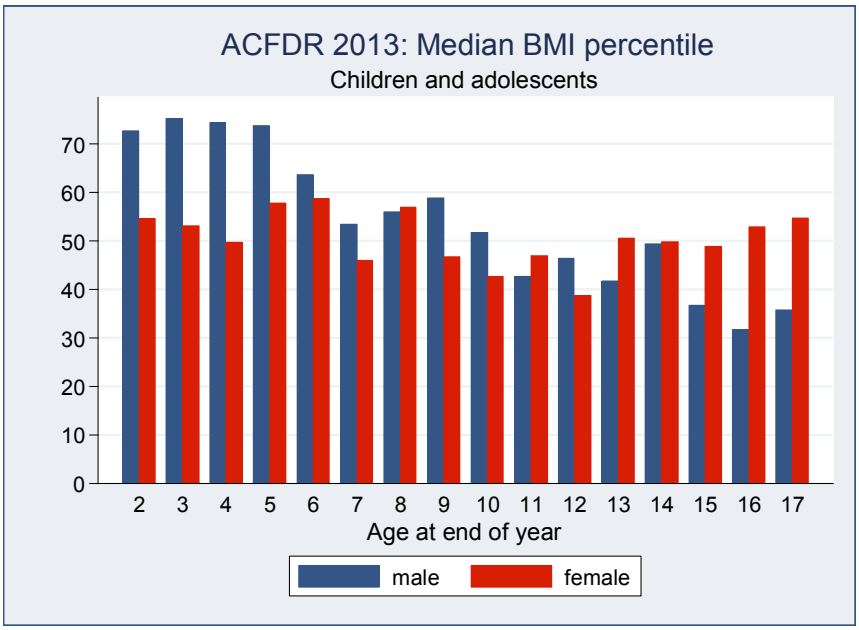
Children and adolescents

Also for the first time, median height percentiles are presented alongside median BMI percentiles for children and adolescents. Median height percentiles are similar to those of the reference population for young children, but are below the 40th percentile for both male and female adolescents in 2013. These indicators may help interpretation of median BMI percentile values, which are higher than reference population values at young ages (at the 74th percentile for boys and 55th for girls aged 2 to 5 years) but are lower than general population norms for adolescent boys but close to population norms for adolescent girls.

ACFDR 2013: Child and adolescent BMI: median percentiles by age group and sex

	Height	BMI
Males		
2 - 5 years	49.6	74.0
6 - 11 years	46.3	55.4
12 - 17 years	37.9	39.8
Females		
2 - 5 years	48.9	54.6
6 - 11 years	48.4	49.8
12 - 17 years	38.5	48.8

BMI percentiles across individual ages show a generally consistent pattern of lower values at higher ages, but a cohort of adolescent girls have BMIs above the 50th percentile.



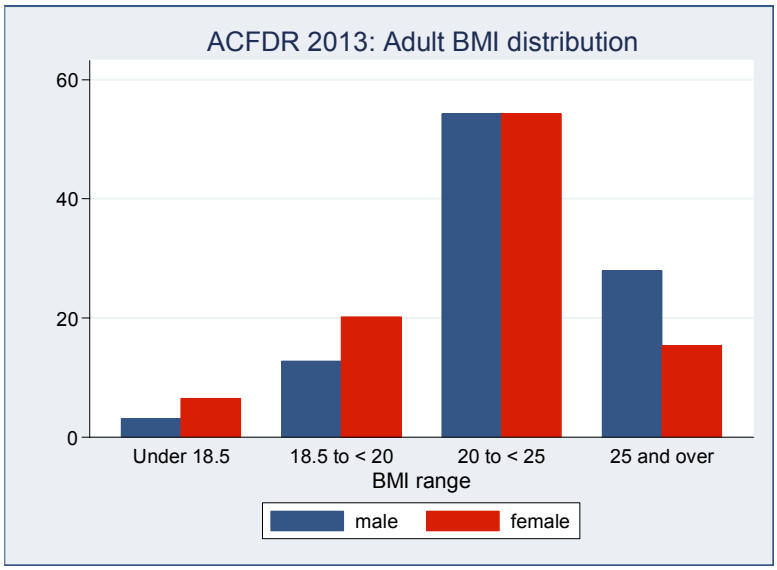
Overall, just under half (46 per cent of males and 49 per cent of females) were below the 50th percentile for BMI in 2013. The distribution is shown in the table opposite.

ACFDR 2013: Child and adolescent height and BMI percentile distributions by sex

	Height			BMI		
	Males	Females	Persons	Males	Females	Persons
	Per cent			*Per cent*		
< 3rd	4.5	4.6	4.5	1.8	1.0	1.4
3rd - 4.99th	1.8	2.3	2.1	1.0	0.9	0.9
5th - 9.99th	6.0	6.5	6.3	4.3	3.6	4.0
10th - 24.99th	17.9	15.1	16.5	12.6	11.2	11.9
25th - 49.99th	25.5	26.9	26.2	26.6	32.7	29.6
50th - 74.99th	24.8	26.0	25.4	27.3	31.2	29.2
75th - 89.99th	12.3	13.0	12.6	18.5	13.7	16.1
90th - 94.99th	2.9	3.5	3.2	5.0	3.6	4.3
95th - 96.99th	2.1	1.0	1.6	0.7	1.0	0.9
>= 97th	2.1	1.2	1.6	2.1	1.2	1.6
Total	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*
	Number			*Number*		
Total	*715*	*695*	*1,410*	*715*	*695*	*1,410*

Adult body mass index

Adult Body Mass Index scores show 55 per cent of males and 56 percent of females had an average quarterly BMI score in the range 20 to less than 25. The proportion of females who had BMI scores below 20 (28 per cent) is higher than the proportion of males (16 per cent). Over a quarter (28.5 per cent) of adult males had a BMI above 25.

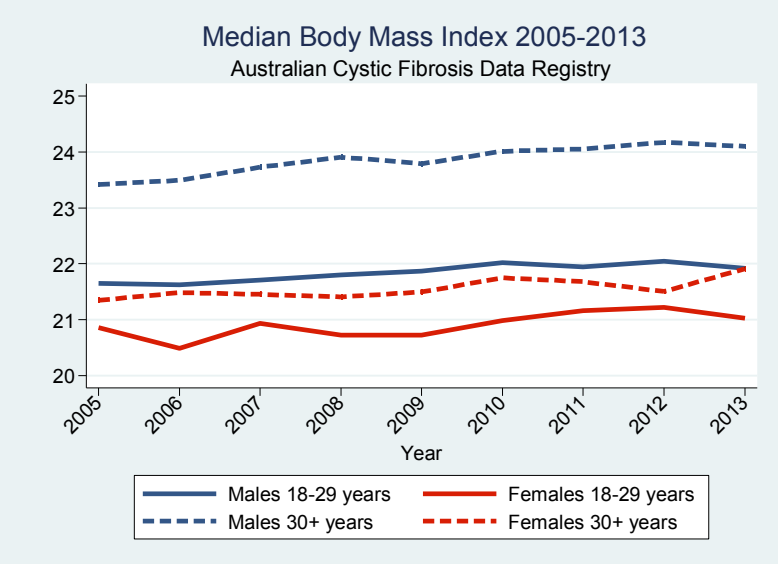


ACFDR 2013: Adult BMI distribution

	BMI range				
	Less than 18.5	**From 18.5 to <20**	**From 20 to <25**	**25 and over**	**Total**
			Males: per cent		
18 - 29 years	4.7	18.2	58.6	18.4	100.0
30 + years	1.2	6.5	51.3	41.0	100.0
Male adults measured	*3.2*	*13.0*	*55.4*	*28.5*	*100.0*
			Females: per cent		
18 - 29 years	9.0	23.5	55.7	11.9	100.0
30 + years	3.4	17.3	57.4	21.9	100.0
Female adults measured	*6.8*	*21.0*	*56.4*	*15.9*	*100.0*

Note that the lower BMI range has been changed from 'less than 18' to 'less than 18.5' to bring the BMI ranges shown in the table and chart above into accord with the International Classification of adult underweight, overweight and obesity as published by the World Health Organisation (see References), and as recommended in advice submitted by the Dietitians Association of Australia.

The following chart shows general increases in median BMI values for grouped adult age data since 2005.



4 Treatment of cystic fibrosis

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

4.1 Visits to clinics

The average number of visits for both children and adolescents and for adults was 4.9 in 2013. This figure should be treated with some caution as it may have been affected by different practices in recording clinic visits at contributing centres. The median number of visits to clinics is less likely to be so affected, and stood at 4 for children and adolescents and also the same for adults in 2013.

4.2 Therapy for cystic fibrosis patients

Antibiotic therapy was prescribed for most CF patients, including 92 per cent of the youngest patients, under age 2 years, and for at least 95 per cent of patients in all other age groups. These proportions, and the numbers and proportions that follow in this section, were compiled from therapy usage information supplied for 2311 patients, 71.4 per cent of all patients in the registry. Missing data were concentrated more in adult than child and adolescent patients, as 2 major adult centres did not supply therapy data for 2013. Proportions of adult and child/adolescent patients for whom therapy data were missing were 41 and 16 per cent respectively.

Oral antibiotic therapy was prescribed for 94 per cent of antibiotics users. Both PRN (as needed) and continuous usage was prescribed for these patients at some time during 2013, as shown in the following table. Higher proportions of adolescents (39.2%) and very young children (45.9%) than those in other age groups were prescribed oral antibiotics for continuous use.

ACFDR 2013: Oral antibiotic therapy - mode of use by age group [(a)]

	0 - 1 years	2 - 5 years	6 - 11 years	12 - 17 years	18 - 29 years	30 + years	All ages
				Per cent			
Mode of use:							
As needed (PRN)	58.7	77.7	77.9	70.1	86.1	85.4	78.3
Continuous	45.9	25.8	28.2	39.2	30.2	28.6	31.5
Mode of use unknown	0.9	0.0	0.2	0.5	0.4	0.0	0.3
Total oral antibiotics users(b)	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*
				Number			
Total oral antibiotics users	109	287	461	411	460	322	*2,050*

(a) Patients for whom no treatment information was provided (29 per cent of total) were excluded from analysis.
(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.

One half (49.8%) of antibiotics users used inhaled antibiotics in 2013, with proportions generally greater in successively older age groups.

ACFDR 2013: Inhaled antibiotics by age group[a]

	0 - 1 years	2 - 5 years	6 - 11 years	12 - 17 years	18 - 29 years	30 + years	All ages
				Per cent			
Inhaled antibiotics							
Yes	11.0	26.1	38.7	58.7	62.8	66.0	49.8
No	62.4	52.9	39.2	26.9	25.6	23.6	34.0
Unknown	6.4	2.1	2.7	1.9	0.0	0.8	1.7
Total antibiotics users	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*
				Number			
Total antibiotics users	*109*	*291*	*475*	*424*	*508*	*368*	*2,175*
Mode of use							
As needed (PRN)	100.0	81.6	72.8	60.2	70.2	74.1	70.4
Continuous	0.0	19.7	27.7	41.8	34.8	29.2	32.5
Mode of use unknown	0.0	0.0	0.0	0.4	0.3	0.4	0.3
Total inhaled antibiotics users(b)	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*
				Number			
Total inhaled antibiotics users	*12*	*76*	*184*	*249*	*319*	*243*	*1,083*

(a) Patients for whom no treatment information was provided (29 per cent of total) were excluded from analysis.
(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 proportion of patients include pancreatic enzymes (85% of children/adolescents and 79% of adults), vitamin supplements (78% and 75% respectively), bronchodilators (38% and 64%) and salt tablets (46% and 22%).

ACFDR 2013: Other therapy by type[a][b]

	Child/adolescent		Adult	
	Number	Per cent	Number	Per cent
Dornase alpha	514	37.8	432	45.4
Pancreatic enzymes	1,157	85.1	755	79.3
Vitamin supplements	1,053	77.5	717	75.3
Bronchodilators	511	37.6	608	63.9
Corticosteroids inhaled	274	20.2	343	36.0
Corticosteroids oral	80	5.9	91	9.6
Insulin	129	9.5	194	20.4
Macrolides	154	11.3	564	59.2
Salt tablets	619	45.5	205	21.5
Antihypercalcaemics	5	0.4	33	3.5
Gastric acid secretion reducers	311	22.9	339	35.6
Other	745	54.8	341	35.8
Patients with therapies reported	*1,359*	*100.0*	*952*	*100.0*

(a) Patients for whom no treatment information was provided (29 per cent of total) were excluded from analysis.
(b) Individuals may use more than one type of therapy; percentages by type of therapy add to more than 100.0.

ACFDR 2013: Nutritional supplements by age group[(a)(b)]

	0 to 1 year	2 to 5 years	6 to 11 years	12 to 17 years	18 to 29 years	30+ years	All ages
				Per cent			
Oral (prescribed)	13.3	28.4	38.8	39.2	33.2	30.5	33.4
Nasogastric	3.6	0.5	1.7	2.8	1.2	2.1	1.8
Total Parenteral Nutrition (TPN)	1.2	0.0	0.0	0.0	0.0	0.3	0.1
Gastrostomy tube/button	1.2	2.8	8.9	14.8	4.9	1.6	6.3
Nutritional supp. type unknown	0.0	0.0	0.0	0.0	0.2	0.0	0.1
Total using nutritional supplements	16.9	31.3	45.4	49.4	37.3	31.6	38.2
Not using nutritional supplements	83.1	68.7	54.6	50.6	62.7	68.4	61.8
Patients with nutritional supplements reported	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*
				Number			
Total with nutritional supplements. reported	*83*	*211*	*361*	*324*	*512*	*380*	*1,871*
Nutritional supps. not reported	38	97	132	113	38	22	440
Patients with therapies reported	*121*	*308*	*493*	*437*	*550*	*402*	*2,311*

(a) Patients for whom no treatment information was provided (29 per cent of total) were excluded from analysis.
(b) Individuals may use more than one type.

Of the 2,311 patients for whom treatment data were reported in 2013, nine were reported to have commenced oxygen therapy during 2013 and 23 remained on oxygen therapy commenced in a previous year. The majority (7 and 17 respectively) were adults.

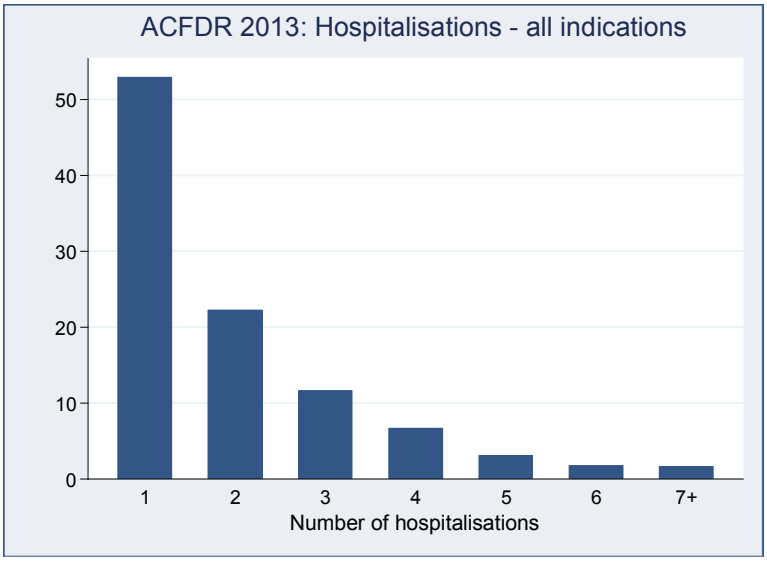
Eight patients commenced using non-invasive ventilation in 2013 and 11 had commenced in earlier years. All of those commencing during 2013 were aged 18 years and over.

For both non-invasive ventilation and oxygen therapy, it is likely that the numbers reported are less than complete, as the full patient population of two adult CF centres is missing from reported data.

4.3 Hospital treatment

The manner of collection of hospitalisation data for the registry does not allow a clear distinction to be drawn between 'no hospitalisation' and missing data in relation to a patient. One adult hospital was excluded from the analysis because no hospitalisation data were provided. An estimate from examination of data from other hospitals is that of the order of 100 paediatric patients and 70 adult patients may have had admitted time in hospital that was not reported.

A total of 1262 patients, or 42.5 per cent of those attending hospitals that provided data, recorded at least one hospitalisation during 2013. The tables and charts in this section describe the hospital experiences of that population.

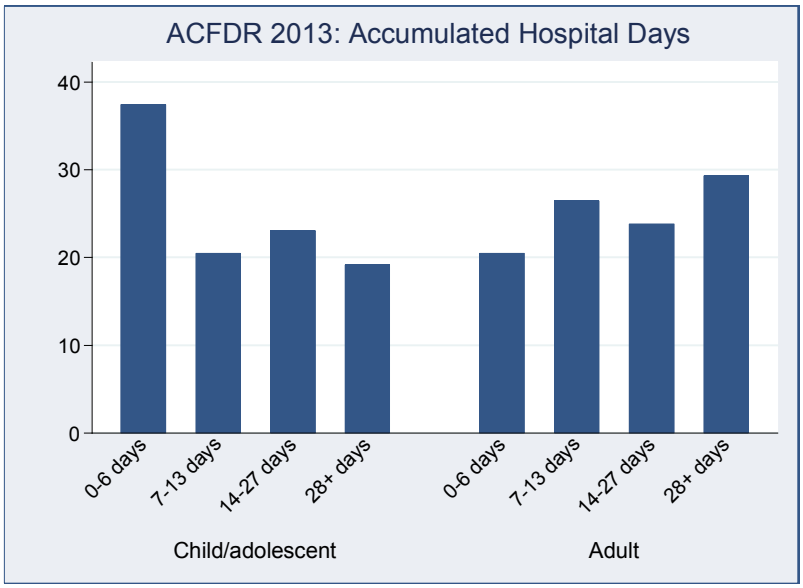


ACFDR 2013: Hospitalisation related to cystic fibrosis, respiratory causes[a]

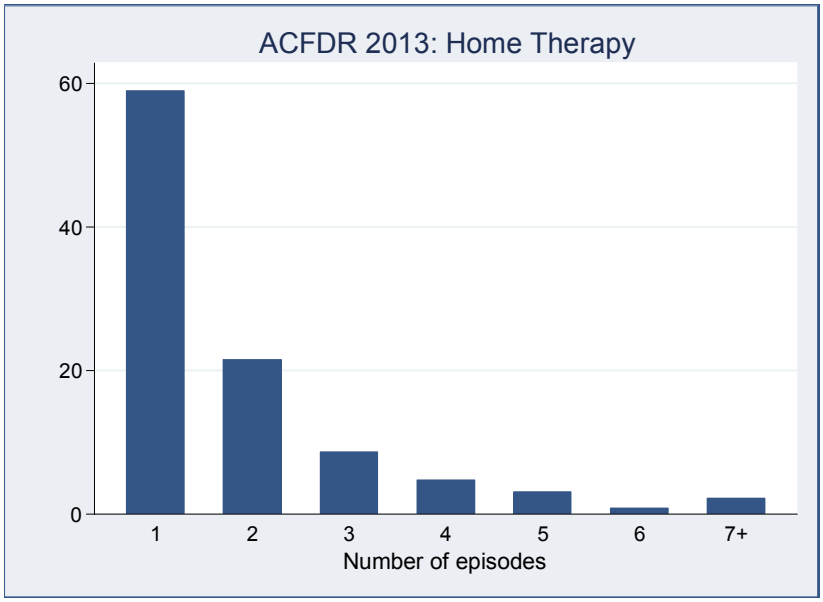
	Persons aged						
	0 - 1 years	2 - 5 years	6 - 11 years	12 - 17 years	18 - 29 years	30 + years	All ages
	Per cent of persons in age group						
Number of hospitalisations:							
None or none reported	51.4	63.0	70.4	56.1	59.4	67.2	62.6
1	26.8	24.0	16.7	21.4	19.5	17.2	19.7
2	13.8	8.6	8.4	11.3	9.6	8.2	9.5
3	4.3	3.1	2.2	4.7	4.7	3.3	3.7
4	2.9	0.8	1.0	4.1	2.8	2.2	2.3
5	0.7	0.3	0.7	1.3	1.9	1.0	1.1
6	0.0	0.3	0.0	0.9	0.9	0.3	0.5
More than 6	0.0	0.0	0.5	0.2	1.2	0.5	0.5
Total	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*
	Number of persons						
Total	138	359	592	533	748	598	2,968

(a) One adult CF centre was excluded from analysis

Of the 1262 persons known to have been hospitalised during 2013, almost half (47.1%) accumulated at least 14 admitted days through the year. Mean and median days, of 20 and 13 respectively for these people, underline the fact that some CF patients spend considerable periods of time hospitalised. Adult patients (mean 24 days, median 14 days) generally spent more days as admitted patients in hospital than children adolescents (16 and 11 days respectively)

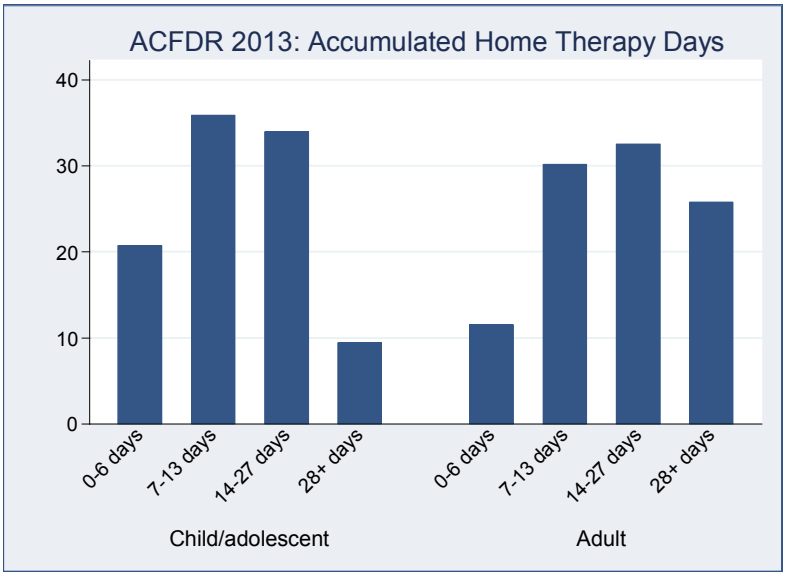


4.4 Home therapy



Charts in this section show the distribution of episode number and days for 358 persons (106 children/adolescents and 252 adults) for whom details were provided of intravenous antibiotic treatment at home. As the number of patients

who have experienced home therapy is considerably less than the number hospitalised in 2013, it is more difficult to make a judgement about whether a centre has missing data or has had few patients in home therapy. However, some missing data is suspected.



4.5 Non-transplant surgery

The following table shows the age distribution of persons reported as having undergone selected non-transplant surgery during 2013. In view of the incompleteness of reporting, these numbers are likely to be under-estimates.

ACFDR 2013: Non-transplant surgery during the year[(a)]

	0 - 1 years	2 - 5 years	6 - 11 years	12 - 17 years	18 - 29 years	30 + years	Total
IV access devices	0	1	7	9	17	8	42
Gall bladder disease	0	0	0	0	0	0	0
Gastrostomy	1	1	10	9	4	0	25
Intestinal obstruction	7	0	0	2	2	1	12
Nasal (any surgery)	0	0	4	7	9	3	23
Other	12	11	21	27	37	29	137

(a) Patients for whom no treatment information was provided (29 per cent of total) were excluded from analysis.

5 Organ Transplants

5.1 Patients assessed for transplant in 2013

Cystic fibrosis centres reported 33 patients had been assessed for organ transplant during 2013. Twenty eight were listed as waiting for a bilateral lung transplant. The organ to be transplanted was not specified for the other five.

Of the total, four were reported as having received a transplant during 2013 and are included in the table below. Of the remainder, 10 were waiting at the end of the year, 11 had their transplant deferred and one had died. The end of year transplant status was not reported for 7 patients assessed for transplant during the year.

5.2 Transplants during 2013

Twenty eight bilateral lung transplants were reported by CF centres as having occurred in 2013. Most of these transplants (18) were performed on patients aged 30 years and over. One of the patients also received a liver transplant. No transplants of other organs were reported during 2013.

ACFDR 2013: Patients receiving lung transplants in 2013

Age group:	Males	Females	Persons
12 - 17 years	0	2	2
18 - 29 years	3	5	8
30 years and over	9	9	18
All ages	*12*	*16*	*28*

6 Mortality

6.1 Deaths recorded in 2013

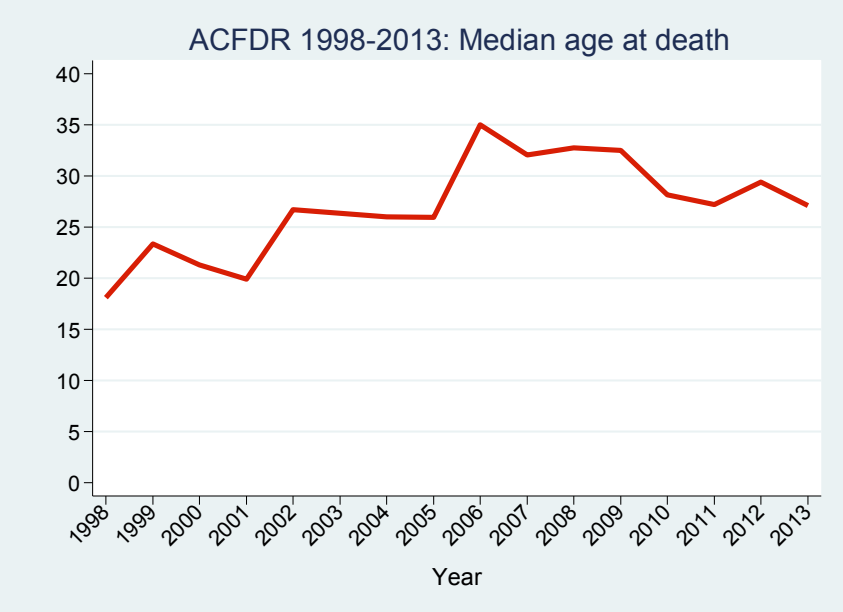
The number of deaths reported to the Registry in 2013 was 38, two fewer than the number reported in 2012 but higher than the numbers reported in any year since 2000. Two of the deaths reported in 2013 were of people aged less than 18 years, both female patients in late adolescence.

ACFDR 2013: Deaths, by age and sex

Age group:	Males	Females	Persons
12 - 17 years	0	2	2
18 - 29 years	13	8	21
30 + years	9	6	15
All ages	*22*	*16*	*38*

By state and territory of residence, the highest number of deaths was reported for people residing in Queensland (14). Nine deaths were reported in New South Wales, seven in Victoria, three each in South Australia and Western Australia and two in Tasmania.

The median age at death for patients tho died on 2013 was 27.3 years. A rising trend shown in this indicator from 1998 appears to have stabilised or even reversed from around 2006, when the median age at death was 35 years. Such apparent trends should be interpreted conservatively, because of the relatively small number of deaths each year.



6.2 Causes of death

Twenty one deaths reported in 2013 were due to pulmonary causes.

ACFDR 2013: Cause of death

	Males	Females	Persons
Related to CF:			
Pulmonary	12	9	21
Other (including post-transplant0	2	1	3
Specific cause not stated	1	1	2
Unrelated to CF	1	2	3
Cause unknown or not stated	6	3	9
All causes	*22*	*16*	*38*

Notes

Supplementary tables and technical notes

A range of supplementary tables and technical notes may be accessed on the Cystic Fibrosis in Australia website: http://www.cysticfibrosis.org.au/data-registry.

Technical notes cover:

- Data collection and editing
- Collection instrument
- Identification and resolution of duplicate records
- Derivations for age, lung function and nutrition data
- Registry data quality

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

Note that, in relation to application of reference equations for FEV1 percent predicted (see Lung Function, Chapter 3), formulae published by Wang et al (1993) are used for males aged from 6 to 17 years and for females aged from 6 to 15 years. Formulae published by Hankinson et al (1999) are used for males aged 18 years and over and females aged 16 years and over.

References

Hankinson JL, Odencrantz JR and Fedan KB. Spirometric Reference Values from a Sample of the General U.S. Population. Am J Respir Crit Care Med 159:179-187 (1999)

Kuczmarski RJ, Ogden CL, Guo SS, et al. *2000 CDC growth charts for the United States: Methods and development*. National Center for Health Statistics. Vital Health Stat 11(246). 2002

Wang X, Dockery DW, Wypij D, Fay ME, and Ferris BG. *Pulmonary Function Between 6 and 18 Years of Age*. Pediatric Pulmonology 15:75-88 (1993)

WHO Global Database on Body Mass Index – BMI Classification, accessed on 20 April 2015 from website http://apps.who.int/bmi/

WHO Multicentre Growth Reference Study Group. *WHO Child Growth Standards: Length/height-for-age, weight-for-age, weight-for-length, weight-for-height and body mass index-for-age: Methods and development*. Geneva: World Health Organization, 2006

Abbreviations

ACFDR	Australian Cystic Fibrosis Data Registry
BAL	Bronchioalveolar lavage
BMI	Body mass index
CF	Cystic fibrosis
CFA	Cystic Fibrosis Australia
FEV_1	Forced expiratory volume (litres) in 1 second
MRSA	Methicillin-resistant Staphylococcus aureus
TPN	Total parenteral nutrition

Access to registry data

Requests for additional information from the Australian Cystic Fibrosis Data Registry are welcome. Application should be made to Cystic Fibrosis Australia (CFA). In accordance with a CFA policy on charging for ACFDR data services, a fee may be charged to recover costs.

Researchers proposing to undertake analysis of unit records may be granted access to de-identified patient records, subject to approval by the Registry's medical advisory and ethics committees, and to 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.

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

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