

Original Research

Do children with cystic fibrosis receiving outreach care have poorer clinical outcomes than those treated at a specialist cystic fibrosis centre?

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Abstract

Introduction: Although cystic fibrosis (CF) centre care is generally considered ideal, children living in regional Australia receive outreach care supported by the academic CF centres.

Methods: This is a retrospective database review of children with CF treated at the Royal Children's Hospital in Melbourne and its outreach clinics in Albury (Victoria), and Tasmania. The aim was to compare the outcomes of children with CF managed at an academic centre with that of outreach care, using lung function, nutritional status and *Pseudomonas aeruginosa* colonisation. Three models of care, namely CF centre care, Shared care and predominantly Local care, were compared, based on the level of involvement of CF centre multidisciplinary team. In our analyses, we controlled for potential confounders, such as socio-economic status and the degree of remoteness, to determine its effect on the outcome measures.

Results: There was no difference in lung function, i.e. forced expiratory volume in 1 s (FEV₁), the prevalence of *Pseudomonas aeruginosa* colonisation or nutritional status (body mass index (BMI)) between those receiving CF centre care and various modes of outreach care. Neither socio-economic status, measured by the Socio-Economic Index for Area (SEIFA) for disadvantage, nor

distance from an urban centre (Australian Standard for Geographical Classification (ASGC)) were associated with lung function and nutritional outcome measures. There was however an association between increased *Pseudomonas aeruginosa* colonisation and poorer socio-economic status.

Conclusion: Outcomes in children with CF in regional and remote areas receiving outreach care supported by an academic CF centre were no different from children receiving CF centre care.

KEY WORDS: comparative study, cystic fibrosis, cystic fibrosis centre, outreach care, paediatric.

Introduction

It is projected that with the significant improvement in survival and without further advances in cystic fibrosis (CF) care, children with CF born in the 21st century will have a median survival to the 5th decade.¹ Expert multidisciplinary team CF care largely based at academic hospitals, referred to as CF centre (CFC) care, is considered a major contributor to this improved outcome.² As about a half of the world's population and about a third of Australians live outside the major urban centres, outreach care becomes a major consideration to ensure similar life expectancies.^{3,4}

Most national CF guidelines^{5–7}, which includes those from USA, Europe and Australia, propose that the best model of care is that delivered by a CFC. This conclusion is based on studies using mortality as the outcome measure,^{8,9} and other studies involving predominantly adults.^{10–12} The results of more recent paediatric studies

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Accepted for publication 31 August 2016.

What is known on the topic:

- A multidisciplinary team approach is the ideal form of care in cystic fibrosis.
- Most major CF organisations promote academic centre care as the ideal mode of delivering this type of care.
- A third of Australians and half the world's population live outside major urban centres and do not have easy access to academic centre care.

What this paper adds:

- There is no difference in clinical and lung function outcomes in children with CF receiving outreach care, supported by an academic centre, compared to those receiving academic CF centre care.
- This is the first study that evaluated the role of confounding factors in this situation and determined that the outcome measures were not confounded by socio-economic status, or distance from a major urban centre.

have challenged this notion, with some showing no difference in clinical outcomes between those children with CF receiving academic centre care and outreach care. However, other studies have found improved lung function with centralised care.^{12–16} In contrast, Thomas *et al.*¹⁷ demonstrated better quality of life scores with outreach CF care than CFC care. A growing body of literature has shown an association between poor socio-economic status (SES) and worse outcomes in patients with CF.^{18,19} However, the confounding effect of SES was not considered in previous studies comparing CF outreach care to CFC care.

This study compared the clinical outcome and lung function of children receiving care at an academic CF centre with those from two different modes of outreach care: Shared care and predominantly Local care. It also explored the influence of potential confounders, namely, SES and remoteness, on clinical outcomes.

Methods

Settings & Subjects

The CF Australia database on children with CF attending a central academic facility and its

accompanying outreach CF clinics during 2010 provided clinical information. A CFC is defined as having access to all the necessary supportive and subspecialty services, caring for at least 50 CF patients under the auspices of a CF director.⁶ The CFC at the Royal Children's Hospital in Melbourne has provided paediatric different models of CF outreach services to Albury, 500 kilometres north of Melbourne, and to the three Tasmanian hospitals over the last 18 years, as outlined in Table 1. Outreach CF clinics in Tasmania are supported by a statewide CF coordinator, monthly videoconference educational sessions and resources provided on a local intranet website.

In classifying remoteness, Australian Standard for Geographical Classification (ASGC)²⁰ quintiles compiled by the Australian Institute of Health and Welfare (AIHW), representing the physical distance from the nearest urban centre, were used. For SES, postal code was used to divide patients into quintiles representing SES categories, based on the Australian Bureau of Statistics (ABS) Socio-economic index for area (SEIFA)²¹ categories for disadvantage (Index of Relative Socio-economic Disadvantage (IRSD)). In this study, the

TABLE 1: Levels of Paediatric CF care at CF clinics in Victoria and Tasmania

Levels of CF Care	Predominant levels of outreach care	Details of delivery of CF care	Specific hospitals (Location)
CFC care		At least 3 monthly review by MDT team Patients receive all care at CFC	RCH (Melbourne)
Outreach care	Shared care	Regular care by local MDT At least two annual visits by CFC MDT	LGH (Launceston) NWRH (Burnie) Albury Hospital (Albury)
	Local care	Majority of care provided by local CF MDT Annual visits from CFC Director	RHH (Hobart)

CFC, Cystic Fibrosis Centre; LGH, Launceston General Hospital; MDT, multidisciplinary team; NWRH, Northwest Regional Hospital; RCH, Royal Children's Hospital; RHH, Royal Hobart Hospital. MDT consists of: CF nurse, physiotherapist, dietician, psychologist, social worker, respiratory paediatrician (CFC) and general paediatrician (outreach care).

number of SEIFA categories was reduced from five to two to avoid sparse numbers in some of the cells.

Clinical outcomes

Lung functions included the best forced expiratory volume in 1 s (FEV₁), forced vital capacity (FVC) and forced expiratory flow rate over 25% to 75% of FVC (FEF_{25–75%}) on any one occasion over the year. The mean of the best forced expiratory volume in 1 s percentage (FEV₁%) values for the year was calculated from the means of the best FEV₁% for each quarter. Anthropometric measures (weight and height) recorded at the time of the best lung functions were used.

To compare nutritional status, body mass index (BMI) was used for children over 2 years of age and z-scores were compared in different age groups. Two positive cultures for *P. aeruginosa* (routinely collected) were considered as chronic infection.

Statistics

Categorical data were expressed as frequencies and percentages, while medians (*M*) and interquartile ranges (IQR) were used for continuous data. Chi-square and Fisher's exact tests were used for categorical data. Wilcoxon rank-sum and Kruskal–Wallis statistical tests were used for continuous data using STATA statistical software, release 12 (StataCorp, College Station TX 77845 USA). Tests of significance were two-tailed, and a *P*-value <0.05 was considered statistically significant.

Ethics

Informed consent was obtained for entry of information into the national Australian CF database and ethics approval was given by the institutional review boards of the Universities of Tasmania (approval #H11975), Melbourne (approval #32055A) and Cape Town (dissertation approval #616/2012).

Results

Baseline characteristics

Of 350 children with CF managed at Royal Children's Hospital and its outreach clinics, the median age was 10.3 years (IQR: 5–15.3 years). Included is a patient who received a lung transplant, two deaths at the CFC and one death in the Shared care group. In Table 2, which describes the baseline characteristics, gender, age and pancreatic insufficiency were similar in those receiving various levels of CF care. A similar number of patients (65%) in each model of care were identified via newborn screening.

There were more patients in the two most advantaged SES quintiles receiving CFC care (CFC 44.2%, Shared care 24.5% and Local care 32.2%), which is not evident when combining SES categories as in Table 2. More patients receiving CFC care were categorised as living in major cities and inner regional areas (Table 2).

Management

The median number of outpatient clinic visits per annum (CFC *M* = 4, IQR 3, 5 versus CF outreach *M* = 4 IQR 4,5), sputum cultures (CFC *M* = 3 IQR 1, 5 versus *M* = 2, IQR 0, 5) and intravenous courses of antibiotics (CFC *M* = 2, IQR 1, 2 versus CF outreach *M* = 2, IQR 1, 2) did not differ between those receiving CFC or combined CF outreach care. More patients at the CFC received continuous antibiotics (CFC 49.4% versus CF outreach 27.1% (*P* = 0.006)). The use of Pulmozyme was more prevalent among those children who received outreach care (CFC 28.7% versus CF outreach 48.7% (*P* = 0.011)).

Bacterial colonisation

P. aeruginosa colonisation did not differ across types of CF care. Although *Burkholderia cepacia* and methicillin-resistant *Staphylococcus aureus* (MRSA) differed across the modes of CF care, when combined across modes of outreach care, there was no longer a difference detected (*Burkholderia cepacia*: CFC 2.4% versus CF outreach 3.0% (*P* = 0.65))(MRSA: CFC 2.0% versus CF outreach 3.0% (*P* = 0.62)).

Outcome measures

Lung function

A similar proportion of (60%) of patients in each outreach category were able to perform adequate lung functions. The outcome measures, as outlined in Table 3, demonstrated no statistically significant lung function differences across types of care. A similar proportion of patients in each model CF care had evidence of airway obstruction, as evidenced by a FEV₁/FVC ratio below 80% (CFC 39.3%, Shared care 32.3% and Local care 42.1%). In Table 4, we compared clinical outcomes across SES and remoteness categories. There appears to be a trend for worse clinical outcomes for those children with CF of lower socio-economic status, although only *P. aeruginosa* colonisation difference reached statistical significance. The distance from major urban centres did not appear to have an impact on clinical outcome measures in this study. Lung function parameters were also not statistically significantly different when compared by modes of CF care, after controlling for SES (Table 5).

TABLE 2: Underlying characteristics of children with CF being treated at three levels of care in Australia

	Cystic Fibrosis Centre	Shared	Local	P-value
Number of patients	272	49	29	
Gender				
Males (%)	142 (52.2%)	30 (61.2%)	16 (55.2%)	—
Female (%)	130 (47.8%)	19 (38.8%)	13 (44.8%)	<i>P</i> = 0.43
Age (years) [median (M), (IQR)]	10.45 (5.2, 15.9)	11.7 (5.0, 14.7)	8.7 (4.0, 12.8)	<i>P</i> = 0.005
Neonatal screening	177 (65.1%)	35 (71.4%)	16 (55.2%)	<i>P</i> = 0.79
Pancreatic insufficiency (%)	225 (82.7%)	45 (91.8%)	26 (90.0%)	<i>P</i> = 0.35
Genetics				
dF508/dF508 (%)	115 (47.9%)	19 (45.2%)	7 (26.9%)	—
dF508/other (%)	104 (43.3%)	22 (52.4%)	16 (61.5%)	—
other/other (%)	21 (8.8%)	1 (2.4%)	3 (11.1%)	<i>P</i> = 0.16
Missing	32	7	3	
SEIFA score for disadvantage (Binary)				
SES 1, 2 and 3 (most disadvantaged)	138 (50.9%)	37 (75.5%)	18 (67.9%)	—
SES 4 and 5 (<i>least disadvantaged</i>)	133 (49.1%)	12 (24.5%)	9 (32.1%)	<i>P</i> = 0.003
Missing SEIFA score	1	0	1	
ASGC categories				
1 (Major city)	133 (48.9%)	12 (24.5%)	1 (3.4%)	—
2 (Inner regional)	111 (40.8%)	18 (36.7%)	23 (79.3%)	—
3 (Outer regional, remote and very remote)	28 (10.3%)	19 (38.8%)	5 (17.2%)	<i>P</i> < 0.001
Nutrition: weight (kg) [median, (IQR)].	27.7 (18.0, 45.6)	35.0 (16.9, 49.1)	27.2 (15.0, 42.7)	<i>P</i> = 0.68
Height (cm)	130.0 (106.0, 157.0)	142.2 (100.5, 157.2)	129.2 (93.0, 152.7)	<i>P</i> = 0.69
BMI (kg m ⁻²)	18.1 (16.1, 20.3)	16.8 (15.2, 18.4)	18.1 (17.7, 19.2)	<i>P</i> = 0.42
Weight median z-score	-0.02 (-0.68, 0.57)	-0.01 (-0.46, 0.38)	-0.06 (-0.26, 0.38)	<i>P</i> = 0.20
Height median z-score	-0.16 (-0.84, 0.43)	-0.35 (-0.79, 0.22)	-0.24 (-0.87, 0.28)	<i>P</i> = 0.32
CF-related diabetes mellitus	18/265 (6.8%)	1/46 (2.2%)	3/29 (10.3%)	<i>P</i> = 0.62
Management:				
Continuous oral antibiotic	112/247 (49.4%)	14/43 (32.6%)	5/27 (18.5%)	<i>P</i> = 0.002
Macrolide	62 (22.8%)	10 (20.4%)	1 (3.4%)	<i>P</i> = 0.08
DNAse	78 (28.7%)	21 (42.9%)	17 (58.6%)	<i>P</i> < 0.001
Airway colonisation				
<i>P. aeruginosa</i> cultures				
Negative	171 (69.5%)	29 (65.9%)	9 (60.0%)	—
Single	26 (10.6%)	7 (15.9%)	1 (6.7%)	—
≥ 2 positive	49 (19.9%)	8 (18.2%)	5 (33.3%)	<i>P</i> = 0.12
MRSA	5/246 (2.0%)	2/44 (4.6%)	0/15 (0%)	<i>P</i> = 0.05
<i>Burkholderia cepacia</i>	6/248 (2.4%)	2/44 (4.6%)	0/15 (0%)	<i>P</i> = 0.01

Medians (M) and interquartile ranges (IQR) reported in brackets, unless otherwise stated. The *P*-value is for any statistically significant difference between two of the groups. dF508, delta F508; SEIFA, Socio-economic index for areas; BMI, body mass index; *P. aeruginosa*, *Pseudomonas aeruginosa*; MRSA, methicillin-resistant *Staphylococcus aureus*; SES, socio-economic status.

TABLE 3: Clinical outcome in children with cystic fibrosis able to perform lung functions, by level of care

	Cystic Fibrosis Centre	Shared	Local	P-value
Number	163	31	19	
Males (%)	81 (49.7%)	19 (61.3%)	12 (63.2%)	<i>P</i> = 0.66
Age [median years]	12.8 (9.5, 16.2)	12.6 (9.5, 14.9)	11.8 (8.7, 13.7)	<i>P</i> = 0.21
Pancreatic insufficiency (%)	134 (82.2%)	28 (90.3%)	17 (89.5%)	<i>P</i> = 0.21
Anthropometry				
Height (cm)	148.3 (129.0, 163.3)	150.0 (130.0, 157.2)	148.0 (129.2, 154.7)	<i>P</i> = 0.69
Height z-score	-0.14 (-0.84, 0.43)	-0.59 (-0.89, 0.12)	-0.24 (-0.76, 0.38)	<i>P</i> = 0.32
BMI [†] (kg m ⁻²)	17.9 (15.9, 20.2)	17.1 (15.9, 19.0)	18.1 (17.6, 19.9)	<i>P</i> = 0.42
BMI z-scores	-0.12 (-0.74, 0.49)	-0.28 (-1.14, 0.42)	0.12 (-0.24, 0.9)	<i>P</i> = 0.22
<i>P. aeruginosa</i> (%)				
None	68 (64.8%)	15 (68.2%)	5 (55.6%)	—
Present	9 (8.6%)	2 (9.1%)	1 (11.1%)	—
Chronic	28 (26.7%)	5 (22.7%)	3 (33.3%)	<i>P</i> = 0.41
Lung function tests				
FEV ₁ (l)	1.95 (1.4, 2.8)	2.10(1.60, 2.70)	1.80 (1.60, 2.40)	<i>P</i> = 0.85
FEV ₁ % predicted	88.8%(75.8, 99.7)	97.8 (80.6, 105.9)	90.8% (77.6, 101.0)	<i>P</i> = 0.22
Mean FEV ₁ %	83.2 (70.4, 94.3)	87.3 (73.2, 101.1)	83.8(66.5, 97.9)	<i>P</i> = 0.36
FVC (litres)	2.34 (1.73, 3.41)	2.01(1.79, 3.55)	2.33 (1.82, 2.99)	<i>P</i> = 0.93
FEF _{25-75%}	1.7 (1.2, 2.7)	2.0 (1.3, 2.8)	1.9 (1.2, 2.6)	<i>P</i> = 0.71

Medians(M) and interquartile ranges (IQR) reported in brackets, unless otherwise stated. The *P*-value is for any statistically significant difference in clinical outcome measures between the two of the groups of SES and Remoteness categories. BMI, body mass index; *P. aeruginosa*, *Pseudomonas aeruginosa*; FEV₁, forced expiratory volume in 1 s; FVC, forced vital capacity; FEF₂₅₋₇₅, forced expiratory flow rate 25% to 75% of FVC.

TABLE 4: Comparing lung function (FEV₁), body mass index (BMI) and *P. aeruginosa* colonisation across socio-economic categories (SEIFA quintiles) and remoteness (ASGC classification) categories

	<i>n</i> (%)	FEV ₁ (l)	BMI (kg m ⁻²)	Ps Aer ≥2
Socio-economic status – SEIFA (Disadvantage) categories				
Binary				
1, 2 and 3 (Most disadvantaged)	194 (55.8%)	1.82 (1.41–2.62)	17.7 (16.0–20.1)	69 (35.6%)
4 and 5 (Least disadvantaged)	154 (44.2%)	2.06 (1.48–2.76)	18.3 (16.2–19.9)	29 (24.0%)
		<i>P</i> = 0.39	<i>P</i> = 0.37	<i>P</i> = 0.02
Remoteness (ASGC categories)				
Major cities	136 (41.1%)	1.90 (1.38–2.65)	17.34 (15.76–19.86)	40 (30.2%)
Inner Regional	144 (43.5%)	2.04 (1.48–2.62)	18.2 (16.36–19.91)	46 (30.3%)
Outer regional – very remote	51 (15.4%)	1.96 (1.42–2.95)	17.89 (15.56–20.42)	17 (32.7%)
		<i>P</i> = 0.69	<i>P</i> = 0.77	<i>P</i> = 0.76

Medians (M) and interquartile ranges (IQR) reported in brackets, unless otherwise stated. The *P*-value is for any statistically significant difference in clinical outcome measures between the two of the groups of SES and Remoteness categories. ASGC, Australian Standard for Geographical Classification; BMI, body mass index; FEV₁, forced expiratory volume in 1 s; Ps Aer ≥2, two or more positive cultures of *Pseudomonas aeruginosa*; SEIFA, Socio-economic index for area (disadvantage); SES, Socio-economic status.

Other outcome measures

The standardised scores for all nutritional parameters and the prevalence of *P. aeruginosa* colonisation were

not statistically significantly different across the three different levels of CF care (Table 3). Those with no *P. aeruginosa* isolation had a median BMI z-score of 0.13 (IQR -0.53, 0.62), while those with two or more

TABLE 5: Lung function in children with cystic fibrosis able to perform lung functions, by level of care, stratified by SES

Lung function parameters	Cystic Fibrosis Centre	Shared care	Local care	P-value
FEV ₁ (l)				
SES 1–3	1.8 (1.39, 2.64)	2.1 (1.52, 2.57)	1.7 (1.54, 2.25)	—
SES 4–5	2.1 (1.41, 2.76)	2.0 (1.87–3.1)	2.1 (1.61, 2.5)	<i>P</i> = 0.54
FEV ₁ % predicted (%)				
SES 1–3	90.7 (76.1, 102.9)	89.3 (80.6, 103.9)	85.0 (67.44, 94.54)	—
SES 4–5	87.5 (75.7, 96.3)	102.6 (92.2, 117.6)	99.3 (85.32, 106.31)	<i>P</i> = 0.41
FVC (l)				
SES 1–3	2.2 (1.73, 3.27)	2.6 (1.79, 3.01)	2.3 (1.85, 3.04)	—
SES 4–5	2.5 (1.78, 3.44)	2.5 (2.16, 3.55)	2.6 (1.82, 2.99)	<i>P</i> = 0.77
FEF _{25–75} (l/s)				
SES 1–3	1.7 (1.27, 2.7)	1.8 (1.22, 2.76)	1.7 (1.13, 2.06)	—
SES 4–5	1.7 (1.13, 2.74)	2.4 (1.65, 3.34)	2.6 (2.19, 2.64)	<i>P</i> = 0.83
FEV ₁ /FVC <80%				
SES 1–3	27/81 (33.3%)	9/25 (36.0%)	8/19 (42.1%)	—
SES 4–5	37/82 (45.1%)	1/6 (16.7%)	0/9 (0%)	<i>P</i> = 0.92

Medians (M) and interquartile ranges (IQR) reported in brackets, unless otherwise stated. The *P*-value is for any statistically significant difference in lung function outcome measures between two of the SES groups. FEV₁, forced expiratory volume in 1 s; FVC, forced vital capacity; FEF_{25–75}, forced expiratory flow rate 25% to 75% (mid-expiratory flow rate); SES, Socio-economic status; SEIFA (Disadvantage), Socio-economic index for area classification for disadvantage.

infections had a lower median BMI z-score of -0.35 (IQR -0.62, 0.26) (*P* = 0.038).

Combining outreach care groups

A combination of the outreach models, compared to CFC care, provided no statistically significant difference in median FEV₁% (CFC 89.2%, IQR 75.8, 99.7% versus CF outreach 94.3%, IQR 80.6, 103.9%). Median nutritional measures (BMI) did not differ between CFC and outreach care, (CFC: 18.1 kg m⁻², IQR 16.1, 20.3 versus CF outreach 17.6 kg m⁻², IQR 15.8, 18.6). Colonisation with *P. aeruginosa* on at least one occasion was not statistically different between the two groups (CFC 43.7% versus CF outreach 38.9% (*P* = 0.77)).

Discussion

This study found no statistically significant differences in lung function outcomes between children receiving academic centre care (CFC) and CF outreach care. Similarly, no differences in nutritional status and *P. aeruginosa* colonisation were noted. The underlying clinical characteristics across models of care were similar, although there were differences in particularly the socio-economic status of the patient population which needed to be controlled for. These findings suggest that children with CF in regional

areas receiving care from a local multidisciplinary team, supported by an academic CFC, have similar clinical outcomes.

Lung function

Equivalent lung function outcomes in our study for children with CF receiving various levels of outreach care, has been similarly reported in two Australian studies.^{12,14} In the only longitudinal study, from the Netherlands, no statistical difference in clinical outcome across different level of CF care was found.¹³ Two subsequent European reports by Doull *et al.*¹⁵ and Lebecque *et al.*¹⁶ reported better lung function outcomes for children receiving predominantly academic or centralised care. However, in the study by Lebecque *et al.*¹⁶, centralised academic care was compared to unregulated primary care without the involvement of a CFC. In spite of evidence, all the major national CF organisations propose that superior care is delivered at a CFC.^{5–7}

Although some differences in the therapeutic approach were noted across various levels of CF care in our study, none had a significant impact on the measured clinical outcomes. In addition, the concern of an increased risk of *P. aeruginosa* colonisation in patients receiving continuous antibiotics was not supported in our study. It is not clear which aspects of care affect clinical outcomes the most, although

Doull *et al.*¹⁵ have attributed this to the frequency of multidisciplinary team visits.

Other clinical measures

The effective management of nutritional status at local level has been described elsewhere.^{12,13,15} In contrast to the higher rates of *P. aeruginosa* acquisition in academic centres highlighted by Mahadeva *et al.*¹⁰, our study, like others, have found similar infection rates when centralised care was compared to outreach care.^{12,13}

Socio-economic status/Remoteness

The lack of statistically significant differences in lung function or nutritional status across SES categories or distance from a major urban centre was notable. In Canada, with a similar health care system to Australia which ensures universal access to health care, no difference across SES categories in its primary outcome measure (hospitalisation rates) was detected.²² On the other hand, US studies found better clinical outcomes in CF patients with private health care insurance than in those without private health care insurance.¹⁹ Interestingly, in the current study, more patients in the lower socio-economic were colonised with *P. aeruginosa*, a finding which requires further study. More generally, the model of outreach care in the current study could inform the management of other chronic diseases in regional or remote areas.

Limitations

Small differences in lung function and nutritional status could not be demonstrated as the study was not adequately powered to detect this. Another limitation is that with the slower decline in lung function with improved care, larger sample sizes are required to detect subtler changes in lung function. Also, lung function changes have been found to be a relatively insensitive marker of early structural lung disease.²³

The postcode method for evaluating SES (SEIFA) and remoteness (ASGC) are aggregate measures based on census districts and are also less accurate in rural areas than in urban areas as the spatial differences across census districts are larger and the rural communities per census district are more heterogeneous.²⁴

Finally, the current study faces the problem of temporality, a problem often associated with cross-sectional studies. Although the current study has similar limitations to comparable studies, unlike these studies it evaluated the role of potential confounders. A randomised prospective study would be impossible for practical and ethical reasons.

Conclusion

This study shows that children with CF managed in outreach centres which are appropriately staffed do not have significant differences in clinical and lung function outcomes from those in academic centres. These results, while welcome, especially for those receiving predominantly Local CF care, should be interpreted with caution until more evidence becomes available. Socio-economic status and distance from major urban centres did not influence lung function and nutritional outcomes in children with CF.

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Title:

Do children with cystic fibrosis receiving outreach care have poorer clinical outcomes than those treated at a specialist cystic fibrosis centre?

Date:

2017-02-01

Citation:

Weber, H. C., Robinson, P. F., Saxby, N., Beggs, S. A., Els, I. & Ehrlich, R. I. (2017). Do children with cystic fibrosis receiving outreach care have poorer clinical outcomes than those treated at a specialist cystic fibrosis centre?. AUSTRALIAN JOURNAL OF RURAL HEALTH, 25 (1), pp.34-41. <https://doi.org/10.1111/ajr.12334>.

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