Original Research

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

Heinrich C. Weber, MBChB, DCH(SA), MMed(Paed), FCPaed(SA)(Cert Pulmonology), Dip Allerg(SA), MPH, FRACP, Philip F. Robinson, B Med Sc, MBBS, MD, PhD, FRAC, Nicole Saxby, B Bio Med Sci, M Nut Diet, Grad Cert Health, Sean A. Beggs, MBBS, MPH, FRACP, Ingrid Els, MBChB, MMed(Paed), FCPaed(SA), FRACP, and Rodney I. Ehrlich, MBChB, DOH, FCPHM(SA), PhD

1Faculty of Health, University of Tasmania, Rural Clinical School, Burnie, Tasmania, 2Paediatrics - Respiratory Medicine, The Royal Children’s Hospital, Melbourne, Victoria, 3Dietetics/Cystic Fibrosis, Royal Hobart Hospital, Hobart, 4Paediatrics, Royal Hobart Hospital, Hobart, 5Paediatrics, Launceston General Hospital, Launceston, Tasmania, Australia and 6School of Public Health and Family Medicine, University of Cape Town, Cape Town, South Africa

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 (FEV$_1$), 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. 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. 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.

Most national CF guidelines, 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, and other studies involving predominantly adults. The results of more recent paediatric studies...
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. In contrast, Thomas et al. 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. 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. 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) 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) 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 | LGH (Launceston) | |
| | | At least two annual visits by CFC MDT | NWRH (Burnie) | |
| | Local care | Majority of care provided by local CF MDT | Albury Hospital (Albury) | |
| | | 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, psychologists, social workers, 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 (FEF₂₅₋₇₅%) 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 *Burkhodella 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 (*Burkhodella 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%) | P = 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) | P = 0.005 |
| Neonatal screening     | 177 (65.1%) | 35 (71.4%) | 16 (55.2%) | P = 0.79 |
| Pancreatic insufficiency (%) | 225 (82.7%) | 45 (91.8%) | 26 (90.0%) | P = 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%) | P = 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 (least disadvantaged) | 133 (49.1%) | 12 (24.5%) | 9 (32.1%) | P = 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%) | P < 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) | P = 0.68 |
| Height (cm)            | 130.0 (106.0, 157.0) | 142.2 (100.5, 157.2) | 129.2 (93.0, 152.7) | P = 0.69 |
| BMI (kg m⁻²)           | 18.1 (16.1, 20.3) | 16.8 (15.2, 18.4) | 18.1 (17.7, 19.2) | P = 0.42 |
| Weight median z-score  | −0.02 (−0.68, 0.57) | −0.01 (−0.46, 0.38) | −0.06 (−0.26, 0.38) | P = 0.20 |
| Height median z-score  | −0.16 (−0.84, 0.43) | −0.35 (−0.79, 0.22) | −0.24 (−0.87, 0.28) | P = 0.32 |
| CF-related diabetes mellitus | 18/265 (6.8%) | 1/46 (2.2%) | 3/29 (10.3%) | P = 0.62 |
| Management:            |        |       |         |         |
| Continuous oral antibiotic | 112/247 (49.4%) | 14/43 (32.6%) | 5/27 (18.5%) | P = 0.002 |
| Macrolide              | 62 (22.8%) | 10 (20.4%) | 1 (3.4%) | P = 0.08 |
| DNAse                  | 78 (28.7%) | 21 (42.9%) | 17 (58.6%) | P < 0.001 |
| Airway colonisation    |        |       |         |         |
| P. aeruginosa 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%) | P = 0.12 |
| MRSA                   | 5/246 (2.0%) | 2/44 (4.6%) | 0/15 (0%) | P = 0.05 |
| Burkholderia cepacia   | 6/248 (2.4%) | 2/44 (4.6%) | 0/15 (0%) | P = 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.
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

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**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%) | P = 0.66 |
| Age [median years]     | 12.8 (9.5, 16.2)       | 12.6 (9.5, 14.9) | 11.8 (8.7, 13.7) | P = 0.21 |
| Pancreatic insufficiency (%) | 134 (82.2%)       | 28 (90.3%) | 17 (89.5%) | P = 0.21 |

**Anthropometry**

|                        | Cystic Fibrosis Centre | Shared | Local | P-value |
|------------------------|------------------------|--------|-------|---------|
| Height (cm)            | 148.3 (129.0, 163.3)   | 150.0 (130.0, 157.2) | 148.0 (129.2, 154.7) | P = 0.69 |
| Height z-score         | -0.14 (-0.84, 0.43)    | -0.59 (-0.89, 0.12) | -0.24 (-0.76, 0.38) | P = 0.32 |
| BMI (kg m⁻²)           | 17.9 (15.9, 20.2)      | 17.1 (15.9, 19.0) | 18.1 (17.6, 19.9) | P = 0.42 |
| BMI z-scores           | -0.12 (-0.74, 0.49)    | -0.28 (-1.14, 0.42) | 0.12 (-0.24, 0.9) | P = 0.22 |

**P. aeruginosa (%)**

|                        | Cystic Fibrosis Centre | Shared | Local | P-value |
|------------------------|------------------------|--------|-------|---------|
| 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%) | P = 0.41 |

**Lung function tests**

|                        | Cystic Fibrosis Centre | Shared | Local | P-value |
|------------------------|------------------------|--------|-------|---------|
| FEV₁ (l)               | 1.95 (1.4, 2.8)        | 2.10 (1.60, 2.70) | 1.80 (1.60, 2.40) | P = 0.85 |
| FEV₁% predicted        | 88.8% (75.8, 99.7)     | 97.8 (80.6, 105.9) | 90.8% (77.6, 101.0) | P = 0.22 |
| Mean FEV₁%             | 83.2 (70.4, 94.3)      | 87.3 (73.2, 101.1) | 83.8 (66.5, 97.9) | P = 0.36 |
| FVC (litres)           | 2.34 (1.73, 3.41)      | 2.01 (1.79, 3.55) | 2.33 (1.82, 2.99) | P = 0.93 |
| FEF₂₅₋₇₅%             | 1.7 (1.2, 2.7)         | 2.0 (1.3, 2.8) | 1.9 (1.2, 2.6) | P = 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

|                          | n (%) | FEV₁ (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%) |
| P = 0.39                 |       |          |              |           |
| 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%) |
| P = 0.69                 |       |          |              |           |

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.

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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

### 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) | $P = 0.54$ |
| 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) | $P = 0.41$ |
| 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) | $P = 0.77$ |
| FEV$_{25-75}$ (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) | $P = 0.83$ |
| 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%)   | $P = 0.92$ |

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. FEV$_1$, forced expiratory volume in 1 s; FVC, forced vital capacity; FEV$_{25-75}$, 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.

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Doull et al. 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. In contrast to the higher rates of P. aeruginosa acquisition in academic centres highlighted by Mahadeva et al., our study, like others, have found similar infection rates when centralised care was compared to outreach care.

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. 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. 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.

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.

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.

References

1 Dodge JA, Lewis PA, Stanton M, Wilsher J. Cystic fibrosis mortality and survival in the UK. 1947-2003. European Respiratory Journal 2007; 29: 522–526.
2 Australia Institute of Health & Welfare (AIHW). Australia’s Health 2006. AIHW 2006 Cat.no AUS73. Canberra: AIHW. [Cited 19 Nov 2014]. Available from URL: http://www.abs.gov.au/ausstats/abs@.nsf/md/2039.0.55.001
3 Population Division of the Department of Economic and Social Affairs of the United Nations Secretariat. World population prospects: the 2002 revision and World urbanization prospects: the 2003 revision. [Cited 25 Oct 2014]. Available from URL: http://www.un.org/esa/population/publications/wup2003/WUP2003Report.pdf
4 CFF. Clinical Practice Guidelines for Cystic Fibrosis. Bethesda, MD, US; Cystic Fibrosis Foundation, 1997. [Cited 30 Sep 2015]. Available from URL: http://www.un.org/esa/population/publications/wup2003/WUP2003Report.pdf
5 Bell SC, Robinson PJ, eds. Cystic Fibrosis Standards of Care. Sydney: Cystic Fibrosis Australia, 2008.
6 Kerem E, Conway S, Elborn S, Heijerman H. Standards of care for patients with cystic fibrosis: a European consensus. Journal of Cystic Fibrosis 2003; 4: 7–26.
7 Merelle ME, Schouten JP, Gerritsen J, Dankert-Roelse JE. Influence of neonatal screening and centralized treatment on long-term clinical outcome and survival of CF patients. European Respiratory Journal 2001; 18: 306–315.
8 Nielsen OH, Thomsen BL, Green A, Andersen PK, Hauge M, Schiotz PO. Cystic fibrosis in Denmark 1945 to 1985. An analysis of incidence, mortality and influence of centralized treatment on survival. Acta paediatrica Scandinavica 1988; 77: 836–841.
9 Phelan P, Hey E. Cystic fibrosis mortality in England and Wales and in Victoria, Australia 1976-80. Archives of Disease in Childhood 1984; 59: 71–73.
10 Mahadeva R, Webb K, Westerbeek RC, Carroll NR, Dodd ME, Bilton D, et al. Clinical outcome in relation to care in centres specialising in cystic fibrosis: cross sectional study. BMJ 1998; 316: 1771–1775.
11 Walters S, Britton J, Hodson ME. Hospital care for adults with cystic fibrosis: an overview and comparison between special cystic fibrosis clinics and general clinics using a patient questionnaire. Thorax 1994; 49: 300–306.
12 Thomas CL, O’Rourke PK, Wainwright CE. Clinical outcomes of Queensland children with cystic fibrosis: a comparison between tertiary centre and outreach services. Medical Journal of Australia 2008; 188: 135–139.

13 van Koolwijk LME, Uiterwaal CSPM, van der Laag J, Hoekstra JH, Gulmans VAM, van der Ent CK. Treatment of children with cystic fibrosis: central, local or both? Acta Paediatrica 2002; 91: 972–977.

14 Collins CE, MacDonald-Wicks L, Rowe S, O’Loughlin EV, Henry RL. Normal growth in cystic fibrosis associated with a specialised centre. Archives of Disease in Childhood 1999; 81: 241–246.

15 Doull I, Evans H. Full, shared and hybrid paediatric care for cystic fibrosis in South and Mid Wales. Archives of Disease in Childhood 2012; 97: 17–20.

16 Lebecque P, Leonard A, De Boeck K, De Baets F, Malfroot A, Casimir G, et al. Early referral to cystic fibrosis specialist centre impacts on respiratory outcome. Journal of Cystic Fibrosis 2009; 8: 26–30.

17 Thomas C, Mitchell P, O’Rourke P, Wainwright C. Quality-of-life in children and adolescents with cystic fibrosis managed in both regional outreach and cystic fibrosis center settings in Queensland. Journal of Pediatrics 2006; 148: 508–516.

18 Barr HL, Britton J, Smyth AR, Fogarty AW. Association between socioeconomic status, sex, and age at death from cystic fibrosis in England and Wales (1959 to 2008): cross sectional study. BMJ 2011; 343: d4662.

19 Schechter MS, Shelton BJ, Margolis PA, Fitzsimmons SC. The association of socioeconomic status with outcomes in cystic fibrosis patients in the United States. American Journal of Respiratory and Critical Care Medicine 2001; 163: 1331–1337.

20 Australian Bureau of Statistics. Australian Geographical Classification (ASGC) (ABS cat. No. 1216.0.2011) Canberra ABS 2011. [Cited 20 Jan 2015]. Available from URL: http://www.abs.gov.au/ausstats/abs@.nsf/mf/1216.0

21 Australian Bureau of Statistics. Socio-economic index for Areas (SEIFA) Technical Paper. (ABS cat no. 2039.0.55.001.2006) Canberra ABS 2006. [Cited 16 Nov 2014]. Available from URL: http://www.abs.gov.au/ausstats/abs@.nsf/mf/2039.0.55.001

22 Stephenson A, Hux J, Tullis E, Austin PC, Corey M, Ray J. Socioeconomic status and risk of hospitalization among individuals with cystic fibrosis in Ontario, Canada. Pediatric Pulmonology 2011; 46: 376–384.

23 Tiddens HA, Donaldson SH, Rosenfeld M, Pare PD. Cystic fibrosis lung disease starts in the small airways: can we treat it more effectively? Pediatric Pulmonology 2010; 45: 107–117.

24 Australian Bureau of Statistics (ABS). ASGC Remoteness Classification: Purpose and Use, 2003, census paper no. 03/01, ABS Canberra.
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Author/s:
Weber, HC; Robinson, PF; Saxby, N; Beggs, SA; Els, I; Ehrlich, RI

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