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Do New Zealand Children With Non-Cystic Fibrosis Bronchiectasis Show Disease Progression?

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Summary. Background: There is minimal literature available on the long-term outcome of pediatric non-cystic fibrosis (CF) bronchiectasis. Aim: To document 5-year outcomes of children with chest computerized tomography (CT) scan diagnosed bronchiectasis from a tertiary New Zealand (NZ) respiratory clinic. Methods: Review of a clinical database identified 91 children. Demographics, clinical data, lung function, chest X-ray (CXR), sputum, presumed etiology, admission data, and the NZ deprivation index (NZDep) were collected. Univariate and multivariate regression were used to correlate clinical findings with lung function data and CXR scores using the Brasfield Scoring System. Results: Of the 91 children, 53 (59%) were Pacific Island, 22 (24%) Maori, 14 (15%) European, and 2 (2%) Other. The median follow-up period was 6.7 years (range 5.0–15.3 years) and median age at diagnosis was 7.3 years (range 11 months–16 years). Lung function data ($n = 64$) showed a mean decline of −1.6% predicted/year. In 30 children lung function declined (mean FEV_1 −4.4% predicted/year, range 1–17%), remained stable in 13 and improved in 21 children (mean FEV_1 of +3% predicted/year, range 1–15%). Reduced lung function was associated with male gender, chronic *Haemophilus influenzae* infection, longevity of disease, and Maori and Pacific Island ethnicity. There was a significant correlation with FEV_1 and CXR score at beginning ($n = 47$, $r = 0.45$, $P = 0.001$) and end ($n = 26$, $r = 0.59$, $P = 0.002$) of the follow-up period. The only variable consistently related to CXR score was chronic *Haemophilus influenzae* infection occurring in 27 (30%) ($r^2 = 0.52$, $P = <0.0001$). Only four children were chronically infected with *Pseudomonas* species. Six children died. Conclusion: In our experience despite management in a tertiary multidisciplinary bronchiectasis clinic, progression of lung disease continues in a group of children and young adults. **Pediatr Pulmonol. 2011; 46:131–138.**

Key words: ethnicity; outcome; lung function; radiology; colonization.

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INTRODUCTION

In New Zealand (NZ), the incidence of diagnosis of pediatric non-cystic fibrosis (CF) bronchiectasis has been described as the equivalent of 1 in 1,700 births, twice the incidence of CF disease.[1] The ''incidence'' of bronchiectasis referred to is the incidence of bronchiectasis diagnosis, rather than disease in the population as, unlike CF, bronchiectasis is not screened for but presents when symptoms are sufficient to warrant a visit to primary care, complex enough to warrant referral to specialty care and severe enough to warrant radiographic evaluation. This is 7 times higher than the reported diagnosis incidence rate in Finland, and 18 times higher than the United Kingdom (UK), the only countries to publish specific rates.[2–4] Since the first published review of pediatric bronchiectasis in Auckland NZ,[5] between 2000 and 2008 the number of children with bronchiectasis under active review has increased 280% (60–170). This could be due to increased recognition or a true increase in diagnosis of disease.

In 1994 bronchiectasis was reported to be the seventh highest cause of death in adult Pacific women,[6] and since then hospital admission rates have been climbing (Fig. 1).[7] Cohort studies from the 1940s and 1950s report mortality rates of 3–5% over 10- to 12-year follow-up periods.[8,9] Contemporary cohort studies, that also include children, report a much lower mortality rate of 0–1%.[10,11] Indeed, case reports have described resolution and/or improvement in this previously supposed irreversible disease.[10,12,13] Pediatric specialist respiratory centers outside NZ have reported short-term outcome data,[14,15]

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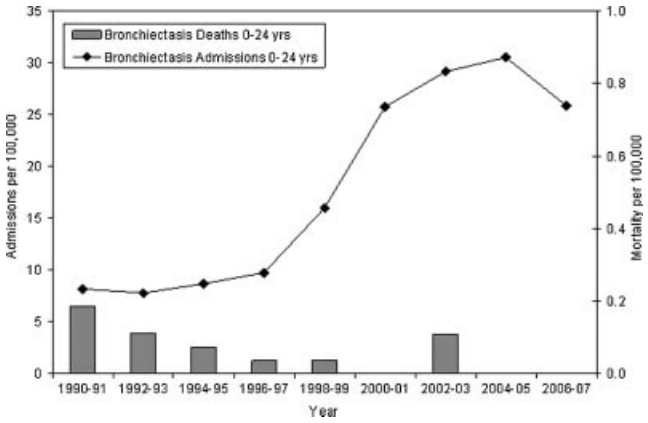

Fig. 1. Hospital admissions (1990–2007) and deaths (1990–2005) due to bronchiectasis in New Zealand children and young people 0–24 years. (Figure provided by Dr Elizabeth Craig, NZ Child and Youth Epidemiology Service 2008. Source: Numerators: National Minimum Dataset and Mortality Collection; Denominator: Census. Note: Mortality data unavailable for 2006–2007.)

but otherwise there is limited longer term outcome data since Field's work more than 30 years ago.[16]

Disease progression monitoring using standard parameters (lung function testing, radiology, sputum) has long become usual practice with CF. The first study, published in 1966 to use lung function tests (LFTs) as a measure of bronchiectasis disease progression, found no change with age.[17] In 1981 Ellis published results of long-term follow-up (mean duration 14 years) on 116 adults with bronchiectasis.[18] Compared to time of diagnosis, of the 94 survivors, 30% were symptomatically better and only 11% were symptomatically worse. The decline in forced expiratory volume in 1 sec (FEV_1) was no greater than seen in normal adults for 80% of the cohort.[18] More recently, follow-up data from King et al.[19] found that adult patients had persistent symptoms of dyspnea and sputum volume which were worse on follow-up and an excess loss of FEV_1 at an average of 49 ml/year (>1% predicted/year). Other adult data have also shown a mean decline of −3.8% predicted/year but with 25% of the cohort having a much higher decline of >−15% predicted/year.[20] Similarly in children we have previously shown over the short term (<5 years) a decline in lung function of −1.9% predicted/year.[21] In contrast, a recent study of 59 children, mean age 8.2 years, had no change in their forced vital capacity (FVC) (slope 0.21) and FEV_1 z scores (slope 0.19) over 2 years.[14]

Better knowledge of progression is needed for more accurate clinical and prognostic evaluation and to determine appropriate intervention trial outcome measures. The aim of this study was to document outcomes for NZ children and young people with bronchiectasis following at least 5 years of management by a tertiary respiratory service. The endpoints measured were lung function, CXR score, and clinical features of disease severity.

METHODS

Study Population

Data on children on the computerized bronchiectasis database[5,22] at the Starship Children's Health, Auckland, New Zealand was retrospectively reviewed. The database was established in 1998 and recorded diagnoses from 1991. All children had high-resolution computerized tomography (HRCT) scan diagnosed bronchiectasis and cases identified had been followed for at least 5 years (December 31, 2006). They had undergone a series of investigations to exclude CF and identify the presumed etiology for the bronchiectasis. The data fields were determined at establishment of the database, and where data were available was collected systematically. The data on the individual child were revised when needed at the clinic reviews—most particularly being checked at an ''annual review'' visit. When possible, data were also collected on those who had transitioned to adult care or had moved within NZ by sending out questionnaires to their current physicians.

Data Collected

Demographics, growth (height, weight, body mass index (BMI)), sputum analysis, presumed etiology, and admission data were collected. Children were defined as being chronically infected with a particular organism if they had a positive sputum or bronchoalveolar lavage (BAL) culture on three or more occasions each at least 1 month apart over a 12-month period. Children able to participate had lung function testing carried out at each clinic or inpatient stay by one of two pediatric respiratory technologists performed on a system 62000 Autobox DL Plethysmograph (Sensormedics Corporation, Yorba Linda, CA) according to American Thoracic Society criteria.[23] FVC and FEV_1 were collected. The Polgar reference equation was used to determine percent predicted as this covered the entire age range.[24] Only children who had more than one LFT at least 12 months apart were included in the final analysis and the ''best'' annual (individuals best spirometric result for each calendar year) was used. Asthma was diagnosed with at least a 15% improvement in FEV1 post-bronchodilator.[25] Chest X-rays (CXRs) were evaluated by pediatric radiologists (HJ and DP), blinded to clinical data, at the beginning and end of the follow-up period and scored using the Brasfield score.[26] The growth parameters used were those recorded with lung function data at diagnosis and end of follow-up period. The NZ index of deprivation score for each child was noted at the beginning (NZDep96) and end (NZDep2001) of the follow-up

period.[27] This has a range from 1 (10% least deprived in NZ) to 10 (10% most deprived in NZ) and is determined by address using a mesh block of 100 persons where possible and combining the following variable in decreasing weight—income, employment, communication, transport, support, qualifications, living space, home ownership.

Management

Each child has a named lead consultant and previously described investigation and management strategies with 3–6 monthly clinic follow-up.[28–30]

Statistical Analysis

Statistical analyses were undertaken with JMP software (SAS Institute, Inc., Cary, NC). Univariate comparisons of variables between children were undertaken with Fisher's Exact Test. The growth parameters were presented as *z* scores for height, weight, BMI, and change over time (www.cdc.gov/growthcharts/zscore.htm). Differences in lung function at diagnosis, between subgroups and correlations to clinical findings were investigated by univariate one-way ANOVA and linear regression. Changes in lung function over time were determined by univariate linear regression on a per patient basis (thus correcting for the different number and time periods of observations between patients), and differences between subgroups and correlations to clinical findings investigated by one-way ANOVA. Multivariate least squares modeling was undertaken to investigate the relative roles of demographic predictors of lung function. However, the relatively low sample size of this study limits the power to investigate multivariate relationships robustly and the focus of this investigation was on univariate associations. Correlations with CXRs are presented as Pearson correlation coefficients (r) for the continuous measures of lung function (which have a non-skewed distribution), and ''coefficients of determination'' (r^2) for categorical variables. For all tests, a *P*-value of less than 0.05 was used for nominal statistical significance.

It was determined that ethical approval was not required as the data collection was made retrospectively and anonymously and was not additional to normally collected data.

RESULTS

Patient Characteristics

Ninety-one children (49 boys) were identified to have been followed for ≥5 years since diagnosis. Patient characteristics are summarized in Table 1. This included 53 Pacific Island (25 Samoan, 12 Cook Island Maori, 10 Tongan, 2 Niuean, 1 Fijian, and 3 not further specified), 22 Maori, 14 European, and 2 Other. Thirty-three (36%) children live in areas with an NZDep score of 10 (most deprived) and 60 (67%) children live in areas scoring 8–10. There was no significant difference in scores over time with a mean NZDep score for the cohort of 9 in 2001 and 8 in 2006.

Thirty-eight (42%) children remain with the Respiratory Service (Starship Children's Health), 18 (20%) had transferred to adult care (age range 16–23 years at end of follow-up), and 1 transferred back to general practitioner care. Eight (9%) children moved from Auckland to within NZ with responses on progress received from 4 cases, 7 (8%) children emigrated to Australia, and 13 (14%) were lost to follow-up.

TABLE 1— Study Group Baseline Characteristics

Baseline characteristics	Patients studied (n = 91)
Gender (males/females)	49:42
Ethnicity	
European	14 (15%)
Maori	22 (24%)
Pacific Island	53 (59%)
Other	2 (2%)
Etiology	
Unknown	41 (45%)
Post-infectious	21 (23%)
Primary immunodeficiency	8 (9%)
Post-oncology disease	10 (11%)
Other	11 (12%)
Follow-up period (years)[1]	6.7 (5–15.3)
Age of diagnosis (years)[1]	7.3 (0.9–16)
NZDep score (range)	8 (2–10)
CXR (Brasfield) score (n = 70)	18 (11–25)
Chronic HI infection	27 (30%)
Chronic *Pseudomonas* infection	4 (4%)
Digital clubbing	37 (41%)
Chronic chest deformity	38 (42%)
Asthma	85 (44%)
Weight *z*-score[2]	0.1 (−2.9 to +2.8)
Height *z*-score[2]	−0.1 (−2.8 to +2.9)
BMI *z*-score[2]	0.3 (−2.6 to +2.5)
FVC % predicted[2] (n = 64)	72 (17–123)
FEV1 % predicted[2] (n = 64)	66 (18–116)

NZDep score, New Zealand index of deprivation score; HI, *Haemophilus influenzae*; BMI, body mass index; FEV_1, forced expiratory volume in 1 sec; FVC, forced vital capacity; Other = 7 aspiration, 2 foreign body, 1 burns, and 1 Marfans.
[1]Median.
[2]Mean.

Etiology

No underlying cause was found for almost half the cases of bronchiectasis (45%) despite extensive investigation. Table 1 outlines the identified underlying etiologies. There was no statistical association between etiology and lung function at baseline or over time ($P > 0.05$).

Morbidity and Mortality

Thirty-one children (34%) had both finger clubbing and chest wall deformity, six had only finger clubbing (7%), and seven (8%) had chest wall deformity alone. Thirty-nine (43%) had neither sign of chronic lung disease. Presence or absence of these signs was not documented in eight children (9%). Thirty-seven (44%) patients had comorbid asthma (n = 85). Overall growth parameters were normal at diagnosis (n = 77) mean height *z*-score of −0.1 (range −2.8 to +2.8), a mean weight *z*-score of 0.1 (range −2.9 to +2.8) and a mean BMI *z*-score of 0.3 (range −2.6 to +2.5). The change in growth over the follow−up period presented as change in *z*-score per year for height was 0.00 (range −0.40 to +0.32), for weight was 0.04 (range −0.44 to +0.79), and for BMI was 0.02 (range −0.82 to +0.64). There was a significant difference between those of European descent (n = 12), compared to those of Maori (n = 19) and Pacific descent (n = 44) for weight ($P = 0.02$) and BMI ($P = 0.0002$) at diagnosis. The presence of clubbing had a significant correlation with lower *z* scores for height ($P = 0.008$), weight ($P = 0.004$), and BMI ($P = 0.05$). Lower *z* scores for weight were also seen with presence of chest wall deformity ($P = 0.01$) and higher (least deprived) NZDep scores of 1–3 ($P = 0.02$).

Six (7%) children died during this follow-up period; two as a direct result of bronchiectasis and 4 from comorbidities.

Clinic and Hospital Attendance

Children attended a median of seven clinic visits during the follow-up period (range 1–26) with a ''did not attend'' rate of 28% for the cohort. Admission data were available for 85 children. Twenty-five (29%) had never required admission for bronchiectasis management, 16 (19%) had required one admission, and 32 (38%) children had required at least annual admissions with a small minority of 5 (7%) needing 3–5 admissions per year. Thirty-three (39%) children had only acute admissions, 14 (16%) had only ''elective'' admissions, and 13 (15%) had a combination. By the end of the follow-up period, there were 10 (11%) with regular ''elective'' admissions for intensive treatment commencing between 2000 and 2006 when the children were aged 7–14 years; 5 with 3 monthly admissions, 3 with 6 monthly admissions, and 2 with annual admissions.

Sputum Culture

Sputum culture and/or BAL culture was available for 88 children (97%); 42 with sputum cultures, 43 children with both, and 3 with only BAL samples. Figure 2 shows the organisms cultured at diagnosis and then most recently with no significant differences between the beginning and end of the period. *Haemophilus influenzae* was the most commonly cultured organism with 27 (30%) found to be chronically infected. Only four children were chronically infected with *Pseudomonas* species.

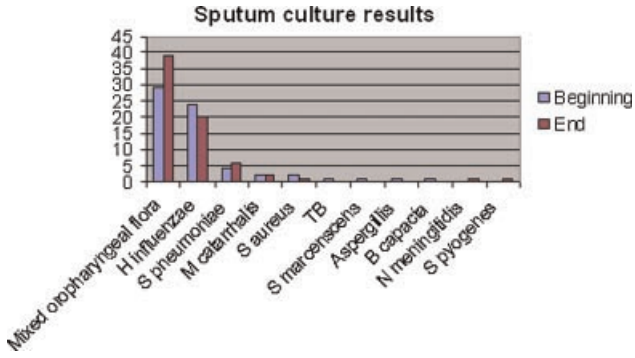

Fig. 2. Sputum culture and/or BAL culture at diagnosis and most recent.

Lung Function

LFTs data were ultimately analyzed in 64 (70%) children who had more than one test, more than 12 months apart. This lung function criteria was not met for 27 children (7 with comorbid developmental delay and 20 who were unable to be followed—7 moved away, 9 lost to follow-up, 4 died). There were a median of 15 LFTs per child (range 2–112) with mean LFT follow-up of 5.8 years (range 1–11.5 years). Table 2 summarizes data for subgroup analysis of bronchiectasis and FEV_1. Male gender, comorbid asthma, presence of digital clubbing and chest wall deformity had a significantly lower mean FEV_1. There was a similar trend with mean FVC (clubbing $P = 0.05$, 95% CI 59–74 vs. 69–83; chest deformity $P = 0.04$, 95% CI 58–73 vs. 70–83). Maori and Pacific Island ethnicity had significantly lower FVC than European children at diagnosis ($P = 0.03$, mean FVC European 88% CI 76–100; Maori 70% CI 61–80; Pacific Island 69% CI 63–75). FVC was not significantly associated with gender, NZDep, etiology, chronic infection, or asthma. A group of children had a more rapid decline in lung function (41%) with a mean FEV1 loss of −4.4%/year and this was associated with female gender.

The variables used in univariate analysis were gender, ethnicity, NZDep score, etiology, chronic *Haemophilus influenzae* infection, *Pseudomonas* colonization, routine admissions, clubbing, chest wall deformity, and asthma. These analyses provided evidence that ethnicity was the greatest predictor of FEV_1 at diagnosis explaining about 11% of the variance. Multivariate least squares modeling indicated that ethnicity, gender and NZDep score all had some independent predictive value on FEV_1 at diagnosis explaining about 13%, 5%, and 5% of the variance, respectively ($P = 0.01$, 0.05, and 0.11). There was no significant difference in FEV_1 related to etiology or

TABLE 2—Subgroup Analysis of Bronchiectasis and Lung Function Tests (Polgar Reference)

Group	% Predicted FEV1[1] (CI)	*P*-value	% Predicted FEV1/year[1]	*P*-value
Gender				
Male	61 (54–68)	0.04	−1.2 (−2.7 to +0.4)	ns
Female	72 (64–79)		−1.1 (−2.7 to +0.9)	
Ethnicity				
European	81 (68–94)	0.03	−0.9 (−4.1 to +1.9)	ns
Pacific Island	66 (59–72)		−0.7 (−2.1 to +1.1)	
Maori	60 (50–70)		−2.2 (−4.5 to 0.0)	
Other	41 (10–71)		—	
Etiology				
Post-infectious	64 (54–74)	ns	−2.2 (−4.6 to −0.2)	ns
Primary immunodeficiency	67 (51–83)		−2.4 (−6.0 to +1.1)	
Post-oncology disease	84 (68–100)		−2.4 (−5.2 to +1.9)	
Other	61 (45–77)		−1.6 (−5.8 to +2.5)	
Unknown	64 (56–72)		+0.3 (−1.4 to +2.2)	
Chronic *Haemophilus influenzae*	62 (54–71)	ns	−1.5 (−3.1 to +0.5)	ns
No *Haemophilus influenzae*	68 (62–75)		−0.9 (−2.4 to +0.7)	
Asthma	60 (52–68)	0.02	−1.4 (−3.2 to +0.3)	ns
No asthma	72 (65–79)		−1.0 (−2.4 to +0.9)	
Chest deformity	59 (51–67)	0.01	−2.1 (−3.8 to −0.4)	ns
No chest deformity	72 (65–79)		−0.5 (−2.1 to +1.4)	
Digital clubbing	59 (52–67)	0.02	−2.2 (−3.9 to −0.4)	ns
No digital clubbing	72 (65–79)		−0.2 (−1.9 to +1.6)	
NZDep				
1–3	50 (30–70)	ns	−0.5 (−4.8 to 3.7)	ns
4–7	74 (64–84)		−0.1 (−2.6 to 2.1)	
8–10	65 (58–71)		−1.7 (−3.0 to −0.1)	
Chronic *Pseudomonas* infection	47 (21–73)	ns	−2.8 (−8.3 to 2.6)	ns
No *Pseudomonas*	67 (62–72)		−1.0 (−2.2 to 0.2)	
Routine admissions	58 (44–74)	ns	−1.9 (−4.7 to 1.3)	ns
No routine admissions	67 (62–73)		−1.0 (−2.2 to 0.3)	

CI = 95% confidence interval; ns = $P > 0.05$; NZDep = NZ index of deprivation score.
[1]Mean best annual.

chronic sputum bacterial infection. FVC was not significant for all comparisons.

FEV_1 for the cohort as a whole over the follow-up period declined by a mean of −1.6% predicted/year (Fig. 3). The FEV_1 of 30 children declined by a mean of −4.4% predicted/year (range −1 to −17%); FEV_1 of 13 children remained unchanged; 21 children improved by a mean of +3% predicted/year (range +1% to +15%). There was a trend towards greater reduction in FEV_1 over time associated with Maori ethnicity, high NZDep score, presence of digital clubbing, or chest wall deformity though these did not reach statistical significance ($P > 0.05$).

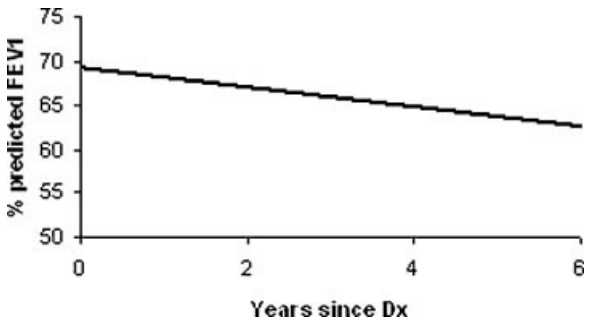

Fig. 3. Pictorial representation illustrating the trend in percent predicted FEV_1 over time for all patients (n = 64). (NB: The graph represents an ''average'' patient and assumes a linear relationship extrapolated over 6 years).

FVC declined by a mean of −0.7 predicted/year. The FVC of 17 children declined by a mean of −5.8% predicted/year (range −1% to −13%); FVC of 10 children remained unchanged; 37 children improved by a mean of +6.4% predicted/year (range +1% to +22%). FVC was not significant for all comparisons.

CXR Scores

The mean Brasfield score for CXR at diagnosis (n = 70) was 18 (range 11–25) and 17.5 (range 6–22.5) at the end (n = 38) of the follow-up period ($P = > 0.05$). There was a significant correlation with beginning CXR score and concurrent FEV_1 and FVC (n = 47, FEV_1 r = 0.45, $P = 0.001$; FVC r = 0.47, $P = 0.0008$) and end CXR score and concurrent FEV_1 and a similar trend with FVC

($n = 26$, FEV_1 $r = 0.59$, $P = 0.002$; FVC $r = 0.40$, $P = 0.05$). The rate of change of CXR score versus rate of change of FEV_1 and FVC was not significant ($n = 26$, FEV_1 $r = 0.04$, $P = 0.86$; FVC $r = 0.24$, $P = 0.24$). The presence of chronic *Haemophilus influenzae* infection was significantly correlated to the CXR score at the end ($n = 26$, $P = < 0.0001$, $r^2 = 0.52$) and the change in CXR score over time ($P = 0.002$, $r^2 = 0.33$). There were no significant relationships with ethnicity, age gender, presumed etiology, digital clubbing, chest wall deformity, asthma, or growth of *Pseudomonas* species for FEV_1 or FVC.

DISCUSSION

The main finding of this review is that almost half the children and young people with non-CF bronchiectasis (45%) have declining lung function over time despite follow-up in a tertiary respiratory clinic. However, chronic colonization with *Pseudomonas* species was rarely seen. Stigmata of chronic lung disease and asthma were associated with poor lung function. Ethnicity, gender, and socioeconomic status appear to be predictive of poor lung function. Sadly, NZ children and young people continue to die from bronchiectasis.

In 1969 Field suggested "it is clear that the patients as a whole improve in the second decade, during puberty, and remain stationary in the third decade."[16] Bastardo et al.[14] have recently described lung function over 2 years ($n = 59$) as stable (FVC and FEV_1 slopes 0.21 and 0.17/annum, respectively). A subgroup ($n = 31$) had data spanning 4 years and these also remained stable (FVC and FEV_1 slopes 0.009 and 0.107/annum, respectively). The mean deterioration in our group over at least 5 years was -0.7% per annum for FVC and -1.6% per annum for FEV_1. This is in keeping with our previous study which showed no change for FVC but a mean decline of -1.9% per annum for FEV_1 in the 44 children with bronchiectasis which was not as great as the corresponding mean decline of -1.7% per annum for FVC and of -2.9% per annum for FEV_1 in the comparison group of 44 children with CF.[21] Yet the FEV_1 at baseline was similar in both the Bastardo et al. and this current study—68.3% predicted (prediction equation not stated) and 66% predicted (Polgar), respectively.

We found a group of children that appeared to have a more rapid LFT decline (41%) with a mean FEV1 loss of -4.4%/year. This was associated with female gender with a trend for other parameters to have an effect including Māori ethnicity, high NZDep score, presence of digital or chest wall deformity. Bastardo et al. suggested that a more rapid decline was present in those who had a worse FEV_1 at baseline.[21] Two papers in adult patients with bronchiectasis have shown that more rapid LFT decline was associated with chronic colonization with *Pseudomonas*, more frequent severe exacerbations, more systemic inflammation, greater volume of sputum and increased bronchodilator use.[19,31] There was no association between etiology and lung function at baseline or over time which is surprising but may be due to small numbers in some etiological categories. Disappointingly, the 10 children who have regular elective admissions for intensive treatment of their bronchiectasis with physiotherapy and IV antibiotics also showed no improvement in lung function over time. We recognize that the findings regarding lung function pertain only to the older and able pediatric group and the findings of deterioration may not apply to the children <5 years of age unable to be measured by this parameter.

At baseline, we found lower LFTs were associated with male gender, longevity of disease, Maori and Pacific ethnicity, digital clubbing, chest wall deformity, and comorbid asthma. Digital clubbing was early recognized to be a poor prognostic sign.[32] More recently both digital clubbing and chest wall deformity have been associated with higher (more abnormal) HRCT chest scores.[33] In a cross-sectional paper in adults, lung function was inversely associated with duration of chronic cough and therefore worse in the group with childhood onset of adult disease.[34] Interestingly Field 30 years ago described worsening of symptoms in those with comorbid asthma, and 2 papers from Turkey ascribe asthma as a cause of bronchiectasis.[10,35] We found that asthma predicted a slower decline in an earlier study, but the presence of asthma was associated with lower FEV_1 at diagnosis in this study.

This study could be criticized for scoring CXRs rather than repeat HRCT scans. Also we used the Brasfield score originally created as a CXR score for CF[26] in the absence of a CXR score specifically developed for pediatric non-CF bronchiectasis, which includes some features that may not apply to non-CF disease (e.g., nodular lesions). However, scoring repeated CXRs has been standard practice in CF for many years. Most children with non-CF bronchiectasis in this study had several X-rays over time, whereas only 18 had repeat HRCT scans. While there was a relationship between the CXR scores at beginning and end with FEV_1, there did not appear to be a correlation over time as the CXR score did not change significantly while the FEV_1 deteriorated. This may indicate its insensitivity in particular regarding the area of the lung covered by the domes of the diaphragm and behind the heart.

The microbiological results of sputum and BAL cultures are similar to other studies.[1,5,14,19] This contrasts with recent adult studies where *Pseudomonas* has been described in 12–33% patients.[36–39] In the current study chronic *Haemophilus influenzae* infection was associated with a poorer CXR score and in a previous study with more severe disease.[21]

Unlike in the CF population, failure to thrive is not a regular feature of disease in bronchiectasis. While in central Australia children with bronchiectasis were three times more likely to have had malnutrition in early childhood,[40] this has not been reported in our previous series, Alaska, Turkey, or the UK.[5,10,41,42] Bastardo et al.[14] found that those with reduced FEV_1 at baseline also had reduced weight and BMI *z* scores with the anthropometric measurements not altering over 2 years of follow-up. We found that overall the growth parameters were normal, although were significantly different depending on the ethnic origin of the child. Lower *z* scores were seen with the presence of clubbing and chest wall deformity, but contradictorily with higher (least deprived) NZDep scores.

Children and young people with bronchiectasis in our population are itinerant and the number of outpatient reviews missed at 28% plus 14% lost to follow-up is disappointing. This indicates that follow-up is not optimal, particularly in comparison with the model of care seen in CF.[30,43] Building good relationships with children and their families/whanau through cultural competency and the use of culturally appropriate support workers may improve engagement with medical care, as would improving socioeconomic factors (transport, travel costs) aiding healthcare access. Also depressing is that childhood mortality remained significant with 7% of our cohort dying during the follow-up period—high when compared to other pediatric series. In addition, 20% of our cohort transferred to Adult Respiratory Services and now there are between 7 and 12 young people transitioning each year with a formal transition process recently in place. These young people enter their adult lives with significant obstructive lung disease.

This study is limited by its retrospective nature. Smoke exposure, active or passive, use of inhaled corticosteroids, the number of prescriptions for, and adherence with, antibiotics was not recorded in this review. Another important factor not addressed is quality of life.

Despite management in a tertiary multidisciplinary clinic, almost half the children with bronchiectasis have declining lung function over time. The effect of asthma on bronchiectasis remains to be determined. Although we have a standard approach to care, not dissimilar to a CF care model, we have difficulties in deploying this in our patient group and we are failing to arrest the disease. Better methods to engage and/or improve adherence or new ''easy to deliver'' treatments are needed.

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