BRONCHIECTASIS

Introduction

The term bronchiectasis is derived from two Greek words literally meaning 'stretching of the windpipe'. Bronchiectasis is characterised by abnormal dilation and distortion of the bronchial tree and is the end result of a number of conditions which lead to difficulty clearing secretions, recurrent infections and a vicious circle of infection and inflammation producing airway injury and remodelling [138].

Bronchiectasis is usually a progressive disease, and typically results in a persistent wet cough with purulent sputum (in older children, as younger children tend not to expectorate the sputum) and recurrent infectious exacerbations. Children with extensive bronchiectasis often have reduced exercise capacity, chest wall deformity, finger clubbing and persistent coarse crackles on examination and they may have slower growth. The disease results in significant morbidity, lost schooldays and parental absences from work [139]. Extensive disease may also produce effects beyond the respiratory system, including cardiac and psychological effects, and may lead to respiratory failure and premature death [140].

The incidence of bronchiectasis in New Zealand children is considerably higher than that reported for Finland or the United Kingdom [141,142,143]. By their 15th birthday 1 in 1700 New Zealand children will have been diagnosed with bronchiectasis. Compared to European children, the incidence in Māori children is three times higher and the incidence in Pacific children is 12 times higher [144]. Bronchiectasis also shows a marked socio-economic gradient, with one Auckland study finding that 67% of affected children were living in NZDep deciles 8–10 (the most deprived 30% of areas) and that this percentage had not changed significantly over time [141]. The same study also found that in Auckland between 2000 and 2008, the number of children with bronchiectasis under active review had increased 280%, although it was unclear whether this was due to increased recognition (e.g. as a result of the increased use of high resolution CT) or a true increase in the burden of disease. Further, despite recent advances in diagnosis, the study found that the aetiology of bronchiectasis was often unclear, with 45% of cases in this study being of unknown aetiology, 23% being post-infectious, 9% being due to primary immunodeficiency and 11% being due to post-oncology disease [141].

The following section explores bronchiectasis rates in children and young people using information from the National Minimum Dataset and Mortality Collection. It concludes with a brief overview of evidence-based review documents and guidelines which consider the prevention or management of bronchiectasis in children and young people.

Data Sources and Methods

Indicator

1. *Acute and Semi Acute Hospital Admissions for Children and Young People Aged 0–24 Years with (non-Cystic Fibrosis) Bronchiectasis listed in any of their first 15 diagnoses.*

Numerator: National Minimum Dataset: Acute and semi-acute hospital admissions for children and young people aged 0–24 years with Bronchiectasis (ICD-10-AM J47) in any of the first 15 diagnoses. Admissions with Cystic Fibrosis (ICD-10 E84) in any of the first 15 diagnoses were excluded.

Denominator: Statistics NZ Estimated Resident Population (with linear extrapolation being used to calculate denominators between Census years).

2. Mortality from (non-Cystic Fibrosis) Bronchiectasis in Children and Young People Aged 0–24 Years

Numerator: National Mortality Collection; Deaths in children and young people aged 0–24 years where the main underlying cause of death was Bronchiectasis (ICD-10-AM J47) and where Cystic Fibrosis (ICD-10-AM E84) was not listed as a contributory cause.

Denominator: Statistics NZ Estimated Resident Population (with linear extrapolation being used to calculate denominators between Census years).

Notes on Interpretation

Note 1: Unless otherwise specified, this analysis focuses on hospital admissions for children and young people with bronchiectasis listed in any of the first 15 diagnoses (rather than on the subset of admissions where bronchiectasis was listed only as the primary diagnosis). The rationale for this wider focus was the fact that

many children and young people with bronchiectasis will not be hospitalised for their bronchiectasis per se, but rather for one of its predisposing conditions or resulting complications. For example, during 2005–2009, only 55.4% of hospitalisations for children and young people with bronchiectasis had bronchiectasis listed as the primary diagnosis, with 11.5% having agranulocytosis or immune deficiencies listed as the primary diagnosis, and a further 19.8% having pneumonia and/or other diseases of the respiratory system listed as the primary reason for admission [145].

Note 2: Because children and young people with cystic fibrosis usually develop bronchiectasis over time, and because the epidemiology of cystic fibrosis and non-cystic fibrosis bronchiectasis differ, admissions where cystic fibrosis was mentioned in any of the first 15 diagnoses have been excluded from this analysis.

Note 3: An acute admission is an unplanned admission occurring on the day of presentation, while a semi-acute admission (referred to in the NMDS as an arranged admission) is a non-acute admission with an admission date <7 days after the date the decision was made that the admission was necessary.

Note 4: **Appendix 3** outlines the limitations of the hospital admission data used. The reader is urged to review this Appendix before interpreting any trends based on hospital admission data.

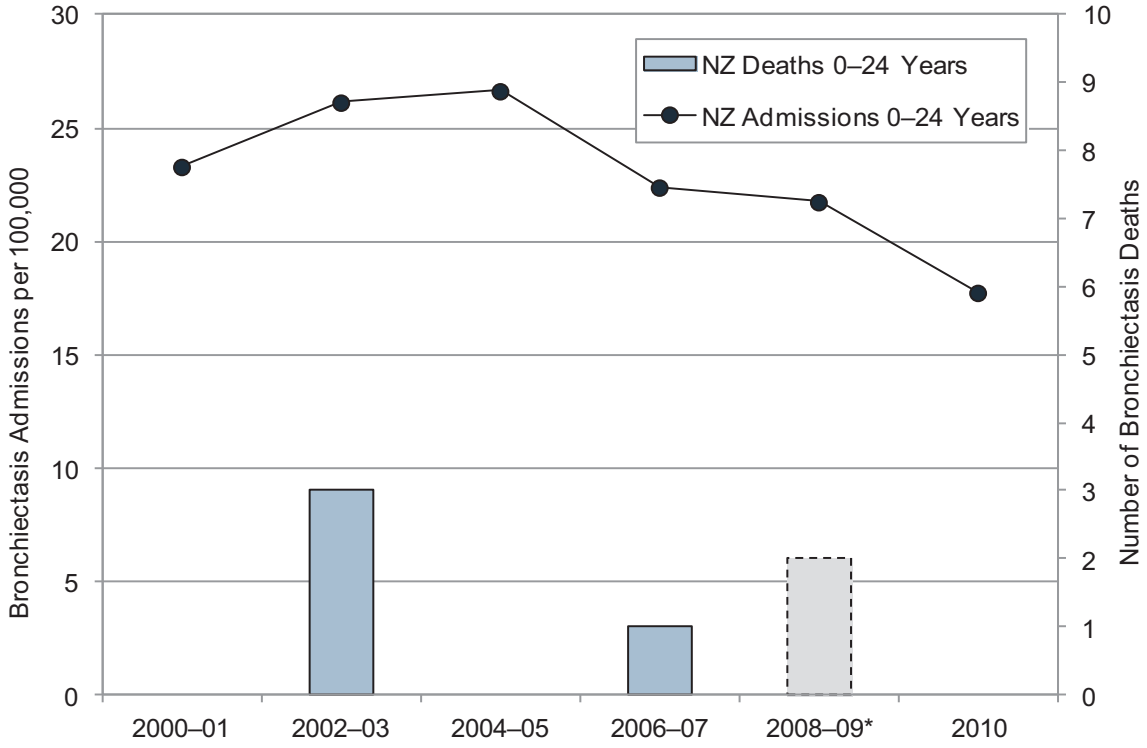
Note 5: 95% confidence intervals have been provided for the rate ratios in this section and where appropriate, the terms *significant* or not *significant* have been used to communicate the significance of the observed associations. Tests of statistical significance have not been applied to other data in this section, and thus (unless the terms *significant* or non-*significant* are specifically used) the associations described do not imply statistical significance or non-significance (see **Appendix 2** for further discussion of this issue).

New Zealand Distribution and Trends

New Zealand Trends

In New Zealand, hospital admissions for children and young people with bronchiectasis increased during the early 2000s, reached a peak in 2004–05 and then declined. During 2000–2008, a total of six New Zealand children or young people had bronchiectasis listed as their main underlying cause of death (**Figure 68**).

Figure 68. Acute and Semi-Acute Hospital Admissions (2000−2010) and Deaths (2000−2008) for New Zealand Children and Young People Aged 0−24 Years with Bronchiectasis

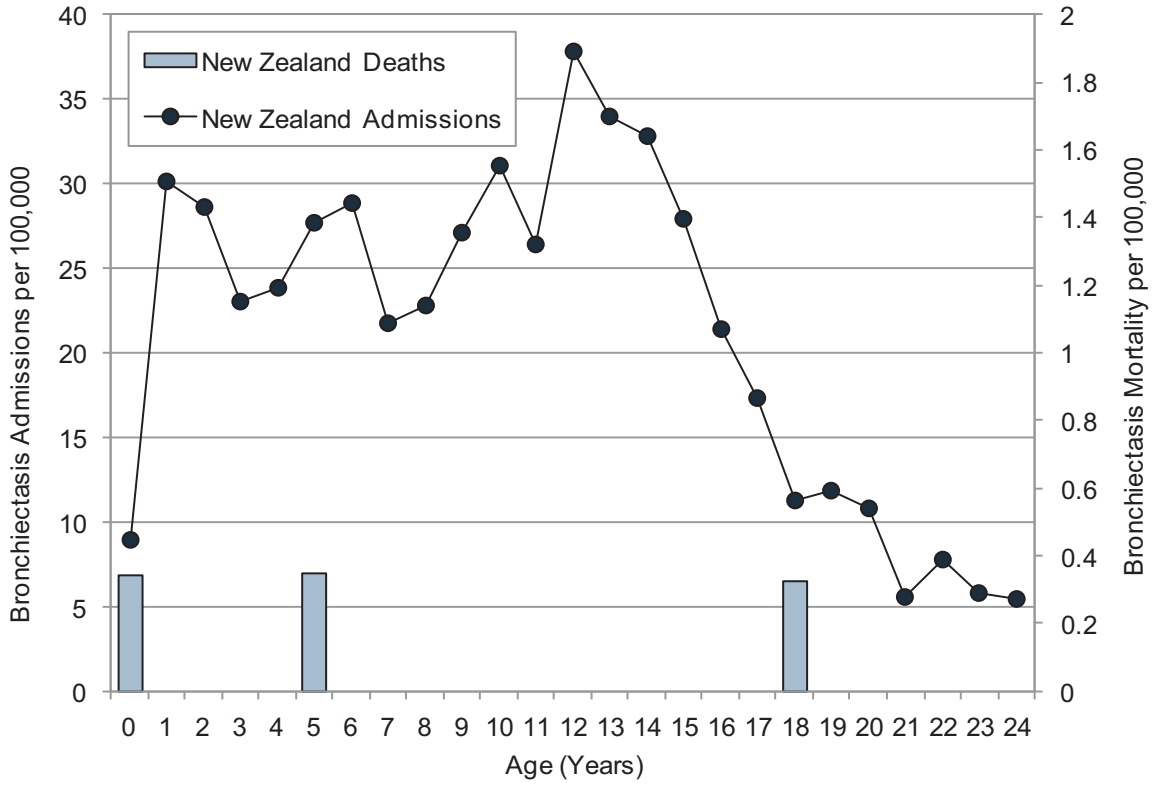


Source: Numerators: National Minimum Dataset (Acute and semi-acute admissions only) and National Mortality Collection (bronchiectasis listed in any of first 15 diagnoses, cystic fibrosis cases excluded); Denominator: Statistics NZ Estimated Resident Population. *Note: Number of Deaths is per 2 year period, with the exception of 2008–09, which is for a single year (2008) only.

New Zealand Distribution by Age

In New Zealand during 2006–2010, hospital admissions for children and young people with bronchiectasis increased rapidly after the first year of life, with rates remaining elevated during childhood, but dropping away amongst those in their teens and early twenties. No consistent age related patterns were evident however, for bronchiectasis deaths during 2004–2008 (**Figure 69**).

Figure 69. Acute and Semi-Acute Hospital Admissions (2006−2010) and Deaths (2004−2008) for New Zealand Children and Young People with Bronchiectasis by Age



Source: Numerators: National Minimum Dataset (Acute and semi-acute admissions only) and National Mortality Collection (bronchiectasis listed in any of first 15 diagnoses, cystic fibrosis cases excluded); Denominator: Statistics NZ Estimated Resident Population

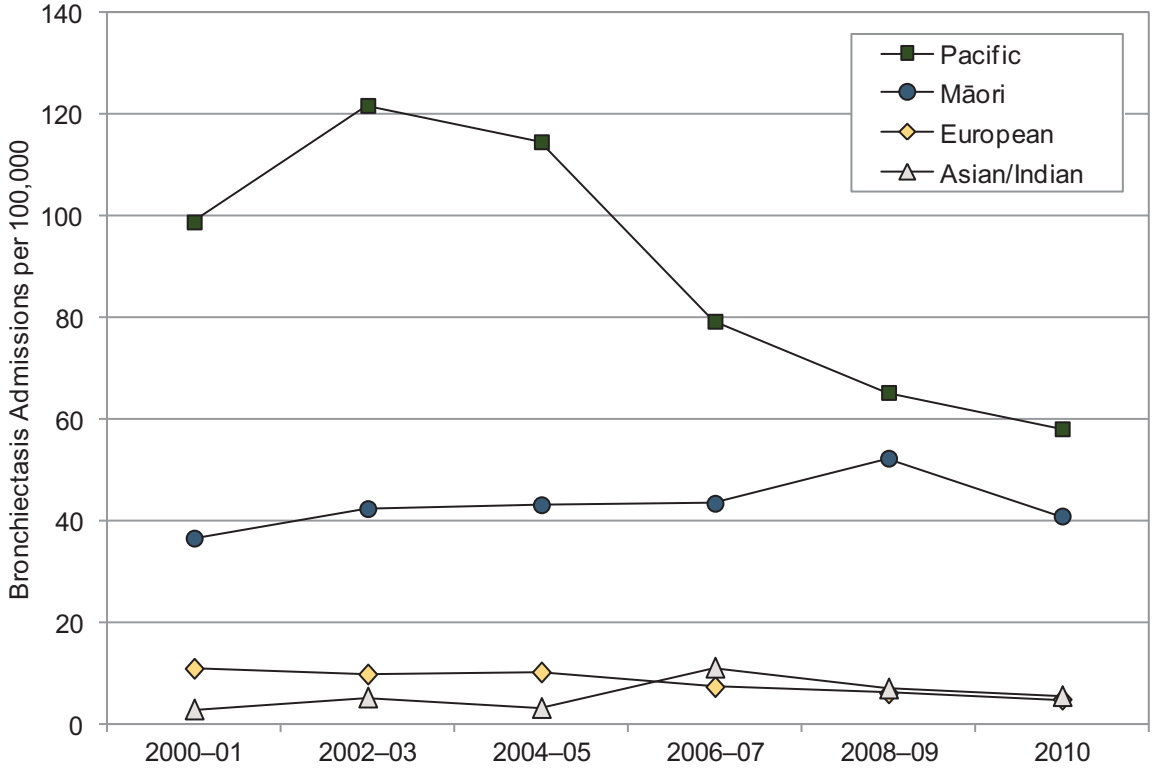
New Zealand Distribution by Ethnicity, NZDep Index Decile and Gender

In New Zealand during 2006–2010, hospital admissions for children and young people with bronchiectasis were *significantly* higher for Pacific > Māori > Asian/Indian > European children and young people and those in average-to-more deprived (NZDep decile 3–10) areas (**Table 77**). Similarly, during 2000–2010 hospital admissions were higher for Pacific > Māori > Asian/Indian and European children and young people, although admissions for Pacific children and young people declined rapidly during the mid-late 2000s (**Figure 70**).

New Zealand Distribution by Season

In New Zealand during 2006–2010, hospital admissions for children and young people with bronchiectasis were generally lower over the summer months (**Figure 71**).

Figure 70. Acute and Semi-Acute Hospital Admissions for Children and Young People Aged 0−24 Years with Bronchiectasis by Ethnicity, New Zealand 2000−2010



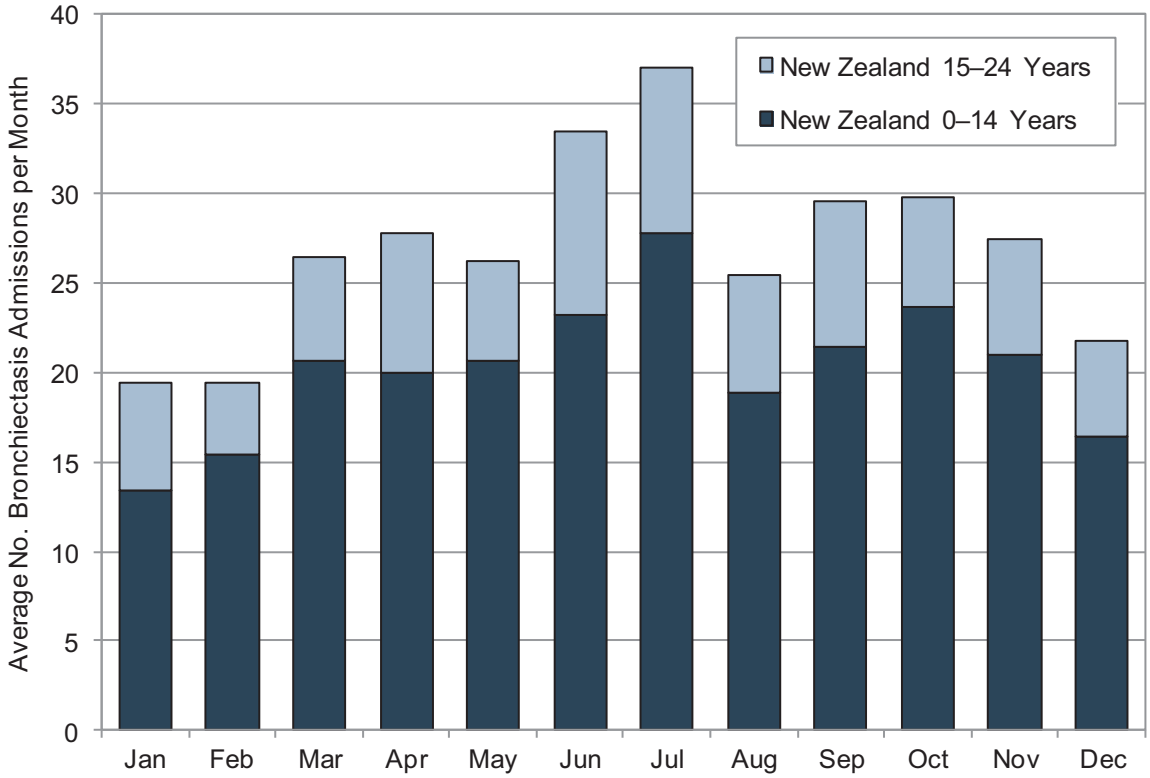
Source: Numerator: National Minimum Dataset (Acute and semi-acute admissions with bronchiectasis listed in any of the first 15 diagnoses (cystic fibrosis cases excluded)); Denominator: Statistics NZ Estimated Resident Population. Note: Ethnicity is Level 1 Prioritised

Table 77. Acute and Semi-Acute Hospital Admissions for Children and Young People Aged 0−24 Years with Bronchiectasis by Ethnicity, NZ Deprivation Index Decile and Gender, New Zealand 2006−2010

Variable	Rate	Rate Ratio	95% CI	Variable	Rate	Rate Ratio	95% CI
Bronchiectasis 0–24 Years							
NZ Deprivation Index Quintile				Prioritised Ethnicity			
Decile 1–2	4.63	1.00		European	6.17	1.00	
Decile 3–4	6.46	1.40	1.02–1.92	Māori	46.4	7.51	6.53–8.63
Decile 5–6	15.3	3.30	2.50–4.36	Pacific	69.1	11.2	9.63–13.0
Decile 7–8	20.6	4.46	3.41–5.82	Asian/Indian	8.06	1.30	1.01–1.69
Decile 9–10	49.7	10.7	8.35–13.8				
Gender							
Female	20.7	1.00		Male	21.7	1.05	0.95–1.15

Source: Numerator: National Minimum Dataset (Acute and semi-acute admissions with bronchiectasis listed in any of the first 15 diagnoses (cystic fibrosis cases excluded)); Denominator: Statistics NZ Estimated Resident Population. Note: Rate is per 100,000; Ethnicity is Level 1 Prioritised; Decile is NZDep2001.

Figure 71. Average Number of Acute and Semi-Acute Hospital Admissions for Children and Young People Aged 0−24 Years with Bronchiectasis by Month, New Zealand 2006−2010



Source: National Minimum Dataset (Acute and semi-acute admissions with bronchiectasis listed in any of the first 15 diagnoses (cystic fibrosis cases excluded))

South Island Distribution and Trends

South Island DHBs vs. New Zealand

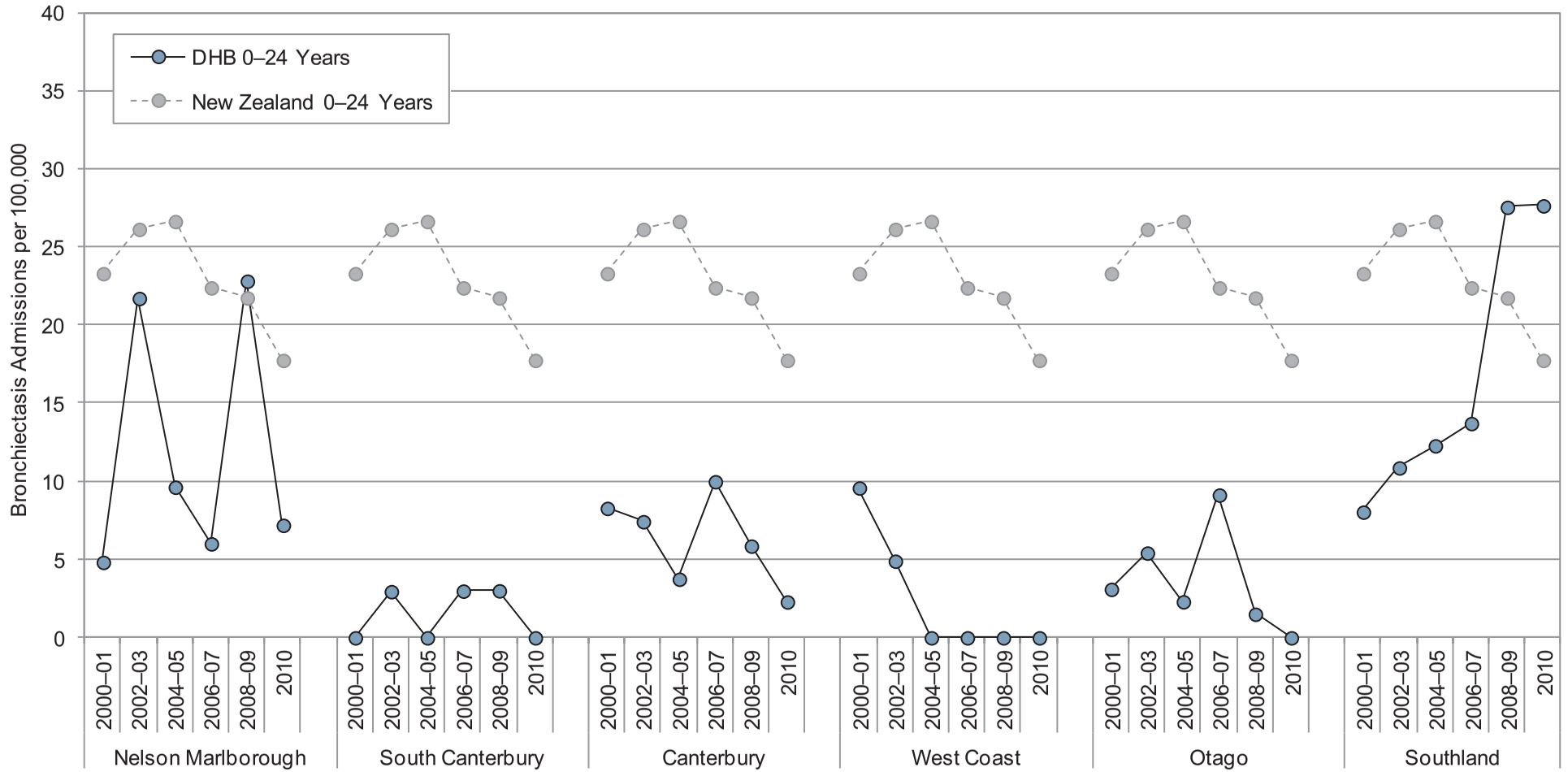
In Nelson Marlborough, Canterbury and Otago during 2006–2010, hospital admissions for children and young people with bronchiectasis were *significantly* lower than the New Zealand rate, while in Southland admission rates were similar. Small numbers precluded a valid analysis in South Canterbury, while no admissions occurred in the West Coast during this period (**Table 78**).

Table 78. Acute and Semi-Acute Hospital Admissions for Children and Young People Aged 0−24 Years with Bronchiectasis, South Island DHBs vs. New Zealand 2006−2010

DHB	Number: Total 2006–2010	Number: Annual Average	Rate per 100,000	Rate Ratio	95% CI
Bronchiectasis 0–24 Years					
Nelson Marlborough	27	5.4	13.0	0.61	0.42–0.90
West Coast	0	0.0	–	–	–
Canterbury	57	11.4	6.75	0.32	0.24–0.41
South Canterbury	<3	s	s	s	s
Otago	14	2.8	4.24	0.20	0.12–0.34
Southland	40	8.0	22.0	1.04	0.76–1.42
New Zealand	1,618	323.6	21.2	1.00	

Source: Numerator: National Minimum Dataset (Acute and semi-acute admissions with bronchiectasis listed in any of the first 15 diagnoses (cystic fibrosis cases excluded)); Denominator: Statistics NZ Estimated Resident Population. Note: s: suppressed due to small numbers.

Figure 72. Acute and Semi-Acute Hospital Admissions for Children and Young People Aged 0−24 Years with Bronchiectasis, South Island DHBs vs. New Zealand 2000−2010



Source: Numerator: National Minimum Dataset (Acute and semi-acute admissions with bronchiectasis listed in any of the first 15 diagnoses (cystic fibrosis cases excluded)); Denominator: Statistics NZ Estimated Resident Population.

South Island Trends

In Nelson Marlborough, South Canterbury, Canterbury, the West Coast and Otago during 2000–2010, large year to year variations (as the result of small numbers) made trends in hospital admissions for children and young people with bronchiectasis difficult to interpret. In Southland however, rates increased, with the most rapid increases being seen between 2006–07 and 2008–09. (**Figure 72**).

Summary

In New Zealand, hospital admissions for children and young people with bronchiectasis increased during the early 2000s, reached a peak in 2004–05 and then declined, with six children or young people having bronchiectasis listed as their main underlying cause of death during 2000–2008. During 2006–2010, admission rates increased rapidly after the first year of life, with rates remaining elevated during childhood, but dropping away amongst those in their teens and early twenties. Admissions were also *significantly* higher for Pacific > Māori > Asian/Indian > European children and young people and those in average-to-more deprived (NZDep decile 3–10) areas.

In Nelson Marlborough, South Canterbury, Canterbury, the West Coast and Otago during 2000–2010, large year to year variations (as the result of small numbers) made trends in hospital admissions for children and young people with bronchiectasis difficult to interpret. In Southland however, rates increased, with the most rapid increases being seen between 2006–07 and 2008–09. During 2006–2010, admissions were *significantly* lower than the New Zealand rate in Nelson Marlborough, Canterbury and Otago, while in Southland admission rates were similar. Small numbers precluded a valid analysis in South Canterbury, while no admissions occurred in the West Coast during this period.

Evidence-Based Reviews Relevant to the Prevention and Management of Bronchiectasis

In New Zealand there are no policy documents which focus solely on the prevention of bronchiectasis in children and young people. A range of documents however consider approaches to respiratory and infectious diseases more generally, and these are reviewed in other sections of this report:

1. **Generic Approaches to Infectious and Respiratory Disease:** Table 46 on Page 166
2. **The Prevention of Second Hand Smoke Exposure:** Table 47 on Page 168
3. **Interventions Aimed at Housing and Household Crowding:** Table 48 on Page 170
4. **Interventions to Improve Breastfeeding:** Table 27 on Page 107

A range of international reviews and guidelines also consider the prevention and management of bronchiectasis in children and young people and these are considered in **Table 79**.

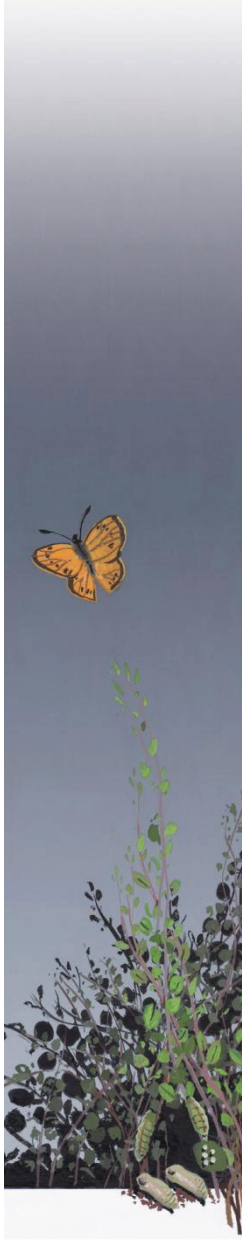


Table 79. Evidence-Based Reviews Relevant to the Prevention and Management of Bronchiectasis

International Guidelines
Pasteur MC, Bilton D, Hill AT, et al. 2010. **British Thoracic Society guideline for non-CF bronchiectasis**. *Thorax* 65 Suppl 1 i1-58. http://thorax.bmj.com/content/65/Suppl_1/i1.full.pdf The aims of these British guidelines were 1) to identify relevant studies in non-cystic fibrosis bronchiectasis; 2) to provide management guidelines based on published studies where possible or a consensus view otherwise and 3) to identify gaps in the knowledge base and identify areas for future research. They cover causes, assessment and investigations, management and complications. The evidence levels for the papers cited and the grading system for recommendations follow the system developed by the Scottish Intercollegiate Guidelines Network (SIGN) and used in the British Thoracic Society (BTS)/SIGN British guideline on the management of asthma.
Chang AB, Bell SC, Byrnes CA, et al. 2010. **Chronic suppurative lung disease and bronchiectasis in children and adults in Australia and New Zealand A position statement from the Thoracic Society of Australia and New Zealand and the Australian Lung Foundation**. *Medical Journal of Australia* 193(6) 356-65. https://www.mja.com.au/public/issues/193_06_200910/cha10303_fm.pdf These Australasian guidelines were produced as a result of a multidisciplinary workshop with the aims of 1) increasing awareness of chronic suppurative lung disease (CSLD) and bronchiectasis in adults and children, 2) promoting earlier diagnosis and improved management of these conditions and 3) presenting an Australian and New Zealand consensus on the management of these conditions. Recommendations are followed by an indication of the level of evidence and the strength of the recommendation (using the GRADE approach).The guidelines cover aetiology and investigations, management, and public health issues. The guidelines state that immunisations that prevent acute respiratory infections are recommended despite the lack of specific evidence for benefit with regard to bronchiectasis. The guidelines include a useful table summarising the evidence for the various possible interventions.
Rosen MJ. 2006. **Chronic cough due to bronchiectasis: ACCP evidence-based clinical practice guidelines**. *Chest* 129(1 Suppl) 122S-31S. These concise American guidelines are based on a systematic review of the literature. Recommendations are followed by an indication of the evidence level, the degree of benefit and the grade of recommendation. They cover diagnosis, specific causes, and treatment.
Systematic and Other Reviews from the International Literature
Irons JY, Kenny DT, Chang AB. 2010. **Singing for children and adults with bronchiectasis**. Cochrane Database of Systematic Reviews 2010(2) Art. No.: CD007729. DOI: 10.1002/14651858.CD007729.pub2. In their introduction, the authors state that a holistic approach is needed in the management of bronchiectasis and its negative effect on quality of life and that therapies involving breathing manoeuvres such as singing may improve both respiratory function and psychological well-being. The authors were unable identify any RCTs assessing singing as a therapy for bronchiectasis and so were unable to draw any conclusions about its benefits or otherwise.
Chang CC, Singleton RJ, Morris PS, et al. 2009. **Pneumococcal vaccines for children and adults with bronchiectasis**. Cochrane Database of Systematic Reviews 2009(2) Art. No.: CD006316. DOI: 10.1002/14651858.CD006316.pub3. The authors of this review identified one randomised controlled open label study in adults (167 participants) with chronic lung disease (bronchiectasis and other associated diseases) which compared 23-valent pneumococcal vaccine (PV) plus influenza vaccine with influenza vaccine alone. There was a significant reduction in acute respiratory exacerbations in the PV group compared to the control group, OR 0.48, 95% CI 0.26-0.88, number needed to treat for benefit=6, 95% CI 4-32, over two years. There was, however, no difference in episodes of pneumonia between the two groups and there was no data on pulmonary decline. One non-randomised Russian study reported that in 25 children with chronic lung disease (including bronchiectasis) who were vaccinated with PPV-23, a year after vaccination *Streptococcus pneumoniae* was isolated in monoculture from only 3 children. This suggests vaccination was beneficial but no clinical effect was described. The review authors concluded that there was limited evidence that the use of 23-valent pneumococcal vaccination was beneficial for adults with bronchiectasis and circumstantial evidence that it was beneficial for children. Due to the absence of evidence they recommend health providers adhere to national guidelines regarding how often the vaccine should be given.
French J, Bilton D, Campbell F. 2003. **Nurse specialist care for bronchiectasis**. Cochrane Database of Systematic Reviews 2003(1) Art. No.: CD004359. DOI: 10.1002/14651858.CD004359. Content updated after new search for studies (no change to conclusions), published in Issue 4, 2008. This review assessed the effectiveness of nurse-led care in the management of bronchiectasis. The review included one randomised cross over trial (80 patients) which found no significant differences in clinical outcomes between nurse-led and doctor-led care in a specialist clinic setting but did find that nurse-treated participants used more healthcare resources (hospital admissions and intravenous antibiotics) in the first arm of the study. The review authors, who also conducted the study, concluded that long term studies are needed to determine whether changes in cost-effectiveness observed over time are due to learning by the nurses, changes in physician behaviour or a carry-over effect and that further studies in other settings including primary care are also needed.

Chang CC, Morris PS, Chang AB. 2007. **Influenza vaccine for children and adults with bronchiectasis.** Cochrane Database of Systematic Reviews 2007(3) Art. No.: CD006218. DOI: 10.1002/14651858.CD006218.pub2.

The authors of this review evaluated the effectiveness of routine influenza vaccination for adults and children with bronchiectasis in reducing the reducing the frequency and severity of respiratory exacerbations and in reducing pulmonary decline. They found that there was no evidence from RCTs for or against vaccination for these purposes but they state that a Cochrane review found that there was evidence that influenza vaccination for people with COPD was beneficial and, given the significant overlap between COPD and bronchiectasis, there is justification for recommending annual influenza vaccination for people with bronchiectasis whilst taking into account individual responses and the potential for adverse effects.

Bradley J, Moran F, Greenstone M. 2002. **Physical training for bronchiectasis**. Cochrane Database of Systematic Reviews 2002(2) Art. No.: CD002166. DOI: 10.1002/14651858.CD002166.

This review considered the effectiveness of a prescribed regime of physical training (compared to no physical training) for either producing improvements or reducing deterioration in physiological and clinical outcomes in people with bronchiectasis. Results from two randomised controlled studies (with a total of 52 participants) published in abstract form only showed that inspiratory muscle training (compared to sham or no training) improved endurance exercise capacity, maximum inspiratory pressure and quality of life. The authors concluded that the only type of physical training for which there was evidence of benefit was inspiratory muscle training.

In terms of the medical management of bronchiectasis, the following interventions have been the subject of **Cochrane Reviews**: Inhaled non-steroidal anti-inflammatories, short acting beta2-agonists, inhaled steroids, oral non-steroidal anti-inflammatories, mucolytics, short courses of antibiotics, anticholinergic therapy, inhaled hyperosmolar agents, long acting beta2 agonists, oral corticosteroids, oral methylxanthines, prolonged antibiotics, surgical treatment, leukotriene receptor antagonists and bronchial hygiene physical therapy.

Other Relevant Publications

The Asthma and Respiratory Foundation of New Zealand, Innes Asher and Cass Byrnes, editors. 2006. **Trying to Catch our Breath: The burden of preventable breathing disorders in children and young people**. Wellington: The Asthma and Respiratory Foundation of New Zealand. http://www.asthmanz.co.nz/files/PDF-files/Burden_FullDocument.pdf

Chapter 11 of this publication considers bronchiectasis. It notes that most children with bronchiectasis (80%) are Māori or Pacific children. Recommendations for prevention are: improvement in socio-economic conditions for the most deprived, reductions in overcrowding and smoking, and improvement in vaccination coverage. Recommendations for management are: increasing awareness among the public and general medical staff, early investigation of children with persistent (>6 weeks) productive or wet cough and improved management based on research.