Bronchiectasis
A Growing Problem
Bronchiectasis (in Children)

• What is it?
• Why such a concern in NZ?
• What to look out for?
• Management
• Positives?

Just a note that the bar for diagnosis, referral and management are all set too high before instigated.

*NOTE: in online version have removed the Photos of children that were in presented version.*
What is it?

- Airway damage
- Mucus retention
- Recurrent infections
- Damage progresses
What is it?

Diagnosed by Chest CT scan

Clinical Diagnosis
- cough
- mucus
- infections

Don’t wait!
# Delay in diagnosis

<table>
<thead>
<tr>
<th>Countries</th>
<th>numbers</th>
<th>Age onset symptoms</th>
<th>Age of Dx</th>
</tr>
</thead>
<tbody>
<tr>
<td>Italy, 2009</td>
<td>105</td>
<td>0.5yrs</td>
<td>7 yrs</td>
</tr>
<tr>
<td>Saudi Arabia, 2007</td>
<td>151</td>
<td>3yrs</td>
<td>7.3yrs</td>
</tr>
<tr>
<td>NZ, 2005</td>
<td>65</td>
<td>2.3yrs</td>
<td>5.2yrs</td>
</tr>
<tr>
<td>Turkey, 2005</td>
<td>111</td>
<td>2.5yrs</td>
<td>7.4yrs</td>
</tr>
<tr>
<td>Turkey, 2005</td>
<td>204</td>
<td>2.3yrs</td>
<td>8 yrs</td>
</tr>
<tr>
<td>UK, 2004</td>
<td>93</td>
<td>1.1yrs</td>
<td>7.2yrs</td>
</tr>
<tr>
<td>NZ, 2003</td>
<td>60</td>
<td>1yr</td>
<td>8 yrs</td>
</tr>
<tr>
<td>Australia, 2003</td>
<td>59</td>
<td>0.5yrs</td>
<td>4.8yrs</td>
</tr>
<tr>
<td>Alaska, USA, 2000</td>
<td>46</td>
<td>0.4yrs</td>
<td>4.8yrs</td>
</tr>
</tbody>
</table>

Kapur et al, Paed Resp Rev 2011
Why such a concern in NZ?

1 in 625 Pacifica
1 in 1300 Māori

2009-2013 Children 0-14 years
• 136 admissions per year
• 1 death every 18 months

Hospital admissions & deaths to 2004
NZ Child & Youth Epidemiology Service 2006
Why such a concern in NZ?

In ADULTS (international data)
- 15-30% COPD, chronic bronchitis in primary care
- 29-50% severe COPD
- 40% difficult to control asthma

One year in South Auckland
- 307 admissions in 152 adults with bronchiectasis
- 46% at least one readmission
- 21% died

Gupta S, Chest 2000
Patel IS, AJRCCM 2004
OBrien C, Thorax 2001
Martinez Garcia MA, AJRCCM 2013

Roberts ME, Internal Med J 2012
Why such a concern in NZ? comparative data

<table>
<thead>
<tr>
<th>At diagnosis</th>
<th>Australia</th>
<th>Alaska</th>
<th>NZ</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bilateral disease</td>
<td>15.3%</td>
<td>38%</td>
<td>83%</td>
</tr>
<tr>
<td></td>
<td>50%</td>
<td>57%</td>
<td>87%</td>
</tr>
<tr>
<td>Widespread disease</td>
<td></td>
<td></td>
<td>64%</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>At diagnosis</th>
<th>Australia</th>
<th>Alaska</th>
<th>NZ</th>
</tr>
</thead>
<tbody>
<tr>
<td>Median LRTI</td>
<td>9</td>
<td>15</td>
<td>13 (3-19)</td>
</tr>
<tr>
<td>Median LRTI admissions</td>
<td>3</td>
<td>3</td>
<td>6 (0-17)</td>
</tr>
</tbody>
</table>

Munro K, Current Paeds 2009
Singleton R, Ped Pulm 2014
Why such a concern in NZ?

Persisting symptoms:
Children < 2 yrs age admitted with severe LRTI seen one year later at time of ‘health’

94 children: 67% had: chronic cough, abnormal exam, persisting CXR abnormalities

_Trenholme AA, Ped Pulm 2013_

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<table>
<thead>
<tr>
<th>Our Data</th>
<th>Age Dx</th>
<th>Severity</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pillarisetti 2015</td>
<td>3.2 yrs</td>
<td>84% bilateral, 66% generalised</td>
</tr>
<tr>
<td>Twiss 2005</td>
<td>5.2 yrs</td>
<td>83% bilateral, 61% generalised</td>
</tr>
<tr>
<td>Edwards 2003</td>
<td>8 yrs</td>
<td>87% bilateral, 83% generalised</td>
</tr>
</tbody>
</table>
At 10 years, FEV1: CF=85%, Bx=68%

FEV1 decline in CF – 2.9%
FEV1 decline in Bx – 1.9%

Associations with accelerated decline (FEV1 ↓4.4%):
• Female, Maori, low SES, chest deformity or clubbing
What to look out for?

COUGH

GROWTH

CLUBBING

CHEST

DEFORMITY

CRACKLES

Note: although these are the traditional features associated with paediatric bronchiectasis - by the time the children are presenting with these features – they already have significant disease and really it is too late to be recognising it.
Chronic suppurative lung disease and bronchiectasis in children and adults in Australia and New Zealand

Chang AB, Bell SC, Torzillo PJ, King PT, Maguire GP
Byrnes CA, Holland AE, O’Mara P, Grimwood K
& extended voting group

Referral in children:

- Wet cough not responding to 4 weeks antibx
- > 3 episodes of cough lasting > 4 weeks
- CXR abnormality persisting > 6 weeks with treatment
What to look out for?

- Cough – persistent, recurrent, wet
- Recurrent resp infections
- Hospital admissions for LRTI
- Recurrent antibx use
- School absenteeism
- SOB with sport
- Asthma – poorly responsive
- Growth

Don’t wait!
Thoracic Society of Australia and New Zealand guidelines

Chronic suppurative lung disease and bronchiectasis in children and adults in Australia and New Zealand

Chang AB et al, MJA 2015

Pasteur MC et al, 2010

1. Antibiotics
2. Airway clearance – physiotherapy, exercise, mucolytics,
3. Environment – housing, smoking
4. Immunizations
5. Nutrition
Azithromycin for prevention of exacerbations in non-cystic fibrosis bronchiectasis (EMBRACE): a randomised, double-blind, placebo-controlled trial

Conroy Wong, Lata Jayaram, Noel Karalus, Tam Eaton, Cecilia Tong, Hans Hockey, David Milne, Wendy Fergusson, Christine Tuffery, Paul Sexton, Louanne Storey, Toni Ashton

- Exacerbations ↓ 62% over 6 months
- Still ↓ 42% over 12 months
Long-term azithromycin for Indigenous children with non-cystic-fibrosis bronchiectasis or chronic suppurative lung disease (Bronchiectasis Intervention Study): a multicentre, double-blind, randomised controlled trial

Valery PC, Respiratory Lancet 2013

<table>
<thead>
<tr>
<th>RESULTS</th>
<th>AZM vs placebo</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Respiratory Infections</td>
<td>Halved</td>
<td>P&lt;0.0001</td>
</tr>
<tr>
<td>Admissions</td>
<td>Decreased by a third</td>
<td>P&lt;0.06</td>
</tr>
<tr>
<td>Antibx for other illnesses</td>
<td>Halved</td>
<td>P=0.003</td>
</tr>
<tr>
<td>Weight for Age</td>
<td>Increased by a third</td>
<td>P=0.005</td>
</tr>
</tbody>
</table>

Valery PC, Respiratory Lancet 2013
Prolonged antibiotics for non-cystic fibrosis bronchiectasis in children and adults (Review)

Cochrane Review, Hnin K et al 2014

- 275 fewer exacerbations per 1000 treated
- 50 fewer hospital admission per 1000 treated

Thoracic Society of Australia and New Zealand guidelines

Chronic suppurative lung disease and bronchiectasis in children and adults in Australia and New Zealand 2015

Trial: those with frequent exacerbations:
- 3 per year,
- or > 2 hospitalisations per year
Nebulised antibiotics

Studies - range of antibxs, range of duration

Combined results:

• ↓ bacteria, ↓inflammation, longer to next infection
• ↑ exercise tolerance, ↑ QoL (some)
• No change in lung function

Lin AMJRCCM 1997
Orriols Resp Med 1999
Barker AJRCCM 2000
Drobnic Ann Pharm 2005
Scheinberg Chest  2005
Bilton Chest 2006
Dhar Thorax 2010
Wilson ERJ 2013
Haworth AJRCCM 2014
Barker Lancet Resp 2014
Nebulised antibiotics

Gentamicin neb 12 months in adults:
• ↓ bacteria in sputum
• ↓ hospitalisation
• ↓ infections
• ↑ exercise ability

Murray AJRCCM 2011

Gentamicin vs placebo neb 3 months cross over
• ↓ inflammation
• ↓ use of other antibiotics

Twiss, ATS 2009
Stay Positive!
Toward making inroads in reducing the disparity of lung health in Australian Indigenous and New Zealand Māori children

Anne B. Chang1,2*, Robyn L. Marsh3, John W. Upham3,4, Lucas R. Hoffman5,6, Heidi Smith-Vaughan1, Deborah Holt1, Maree Toombs1,7, Catherine Bynes8, Stephanie T. Yorkovich1,4,9, Paul J. Torzillo10, Kerry-Ann F. O’Grady2 and Keith Grimwood11 on behalf of the CRE extended group

Factors affecting clinical outcomes or consequences of illness
- Quality care, access, service and family factors
- Microbial factors
- Modifiable factors*
- Host factors

*Modifiable factors:
- Hygiene practices
- Health education
- Socio-economic
- ETS/pollutants
- Vaccinations
- Nutrition
- Housing

Predisposing factors
- Low birth weight
- In-utero ETS
- Genetics

Acute Respiratory Infections (ARIs)

Recurrent+/persistent infection & inflammation

CSLD COPD

Bronchiectasis COPD (severe)
Severe LRTI
Recurrent LRTI
Chronic symptoms
Bx

Chang, Byrnes, Everard Paed Resp Reviews 2011
Serial CT scan comparison

N=52 paired scans

- **IMPROVED**: 17/52 (32.6%)
- **STABLE**: 24/52 (46.1%)
- **PROGRESSED**: 11/52 (21.2%)

*Kim Michelle Williams & Russell Metcalfe
Starship Radiology Service*

(note – not yet published)
2003 (14 month female)
2004 (11 month male)

2008
THE BRONCHIECTASIS FOUNDATION

To create a place for families, siblings, parents, children and individuals affected by this condition “Bronchiectasis” to seek guidance, resources and initiative to manage their health in the best possible way for “them”.

Esther-Jordan Muriwai

Courage led to foundation

Bronchiectasis Foundation Launch
7th April 2015

Foundation launched in memory of Esther — A3
TE HĀ ORA: THE BREATH OF LIFE
NATIONAL RESPIRATORY STRATEGY
RAUTAKI ROMAHĀ A-MOTU