“Bronchiectasis” originates from Greek literally meaning ‘stretching of the windpipe.’ It is defined as an abnormal, irreversible dilatation of one or more bronchi. Bronchiectasis can be caused by a variety of respiratory insults. Even after extensive investigations the aetiology of non-cystic fibrosis (CF) bronchiectasis remains unknown in 50%. Twenty five percent of cases seen at the Starship clinic are related to previous severe childhood pneumonia. Other causes include primary immune deficiency, recurrent aspiration (gastroesophageal reflux and foreign body inhalation), and other rare causes.

In 2000, a retrospective review of children attending the Starship Bronchiectasis Clinic found a crude prevalence rate of 1:6000 in the Auckland paediatric population. A prospective, national study reporting all new cases diagnosed 2001-2003 using the NZ Paediatric Surveillance Unit gave an incidence of 3.7/100,000 children per year. This is seven times higher than the only other comparable national study from Finland. Eighty percent of the children were Maori or Pacific peoples indicating a disproportionate prevalence of bronchiectasis at 1/1875 for Pacific peoples, 1/4244 for Maori and 1/24,900 for European groups. This also means that bronchiectasis is nearly twice as common in Pacific peoples as CF is in the general population.

There are >170 children with non-CF bronchiectasis in the Starship Bronchiectasis Clinic (May 2013). Most have widespread lung disease involving at least three parts of the lung. Without treatment bronchiectasis is almost always a progressive condition resulting in a shortened life expectancy. Bronchiectasis also causes high morbidity; with frequent school absenteeism, poor exercise tolerance, recurrent primary health care presentations, and repeated hospital admissions. Over a quarter of the families do not speak English as their first language and interpreters may be needed to establish an adequate history.

**Clinical features**

- A persistent or recurrent moist cough.
- Increased volume or production of sputum – but remember most children less than school age are unable to expectorate sputum which is usually swallowed.
- Alteration in sputum colour – clear to yellow to green.
- Haemoptysis (rare) – reported more commonly in adults. If reported get a clear history of amount (teaspoons etc), fresh, old clots etc.
- Fever (not always).
- Shortness of breath.
- Wheeze – there may be a coexistent history of asthma in approximately a third of the children with bronchiectasis. Use of inhalers and devices should be assessed as for asthma.
Examination

- There may be no temperature despite an intercurrent exacerbation
- \( \text{SaO}_2 \) – may be low or borderline 90-92\% during the day with an acute exacerbation. It is important to ensure that nocturnal oxygen saturations are adequate if daytime levels are normal or borderline.
- Increased respiratory rate.
- Increased work of breathing.
- Crackles – may be widespread but may be localised to the areas involved.
- Wheeze - may not be bronchospasm as can occur with airflow turbulence caused by excessive bronchial secretions.
- Chest wall abnormalities (60\%) e.g. hyperinflation, Harrison’s sulci, pectus carinatum.
- Digital clubbing (up to 50\%).
- Failure to thrive is unusual in children with bronchiectasis. This may be seen in advanced disease or with specific causes (e.g. immunodeficiency or severe gastroesophageal reflux).

Causative Organisms

- The commonest reason for an acute respiratory exacerbation is a viral infection.
- Three quarters of the children chronically grow \textit{Haemophilus influenzae} (non-typeable) from their sputum.
- A few children concurrently grow \textit{Streptococcus pneumoniae} or \textit{Moraxella catarrhalis}.
- \textit{Staphylococcus aureus} or gram-negative organisms are uncommon.
- There is a small number of children who are chronically infected with \textit{Pseudomonas aeruginosa} and who require antipseudomonal therapy.

Investigation

- Ensure oxygen saturations are adequate in air (>94\%).
- Sputum - probably the most useful test to perform - send for microscopy, bacterial culture and sensitivities. A nasopharyngeal aspirate may be useful in a younger child who is unable to expectorate sputum
- Inflammatory indicators (FBC, CRP, ESR) are frequently elevated even when the child is well and may not effect management.
- Chest x-ray - If you suspect a pleural effusion or a pneumothorax or the child has not had a CXR in the last 12 months then a repeat CXR may be indicated.
- Lung function is used for monitoring and is not normally done acutely
BRONCHIECTASIS - ACUTE RESPIRATORY EXACERBATION

Treatment

Most children managed by the Starship Respiratory Service have action plans for treatment of intercurrent infections.

Antibiotics

- Give 2 weeks of a broad-spectrum oral antibiotic with intensification of chest physiotherapy (at least 2 treatments per day while on antibiotic).
- Even if you suspect a viral aetiology antibiotics are normally prescribed to reduce the microbial load.
- **Antibiotic choice** - where possible should be determined by sputum bacterial culture and sensitivity. Review previous sputum results where possible.
- H.influenzae is frequently found and is usually sensitive to oral Augmentin® (amoxicillin/clavulanic acid) / co-trimoxazole. Use amoxicillin if sensitive.
- If the child is allergic to penicillin, oral cefaclor or erythromycin should be used.
- If the child is still very productive or the sputum purulent, then a further two weeks oral antibiotic should be prescribed.
- If at the end of that 2 weeks of antibiotics the child is no better, or if the child’s clinical condition deteriorates during that 2 weeks of antibiotics, consideration should be given to in-patient intravenous antibiotics and more intensive chest physiotherapy.

Chest Physiotherapy

- Chest physiotherapy is central to treatment and prevention of disease progression.
- Compliance is often an issue.
- Most children should be performing chest physiotherapy once daily and at least twice daily during exacerbations as per their action plan.
- Many children use active cycle of breathing and postural drainage techniques (ACBT) but others use such options as positive expiratory pressure (‘PEP’) devices or sometimes Acapella devices.

Respiratory Support

There are a few children under the respiratory team who have very severe respiratory disease (type I or II respiratory failure). They may be supported at home at night on oxygen which may need to be increased and monitored closely during acute exacerbations. Some are supported on CPAP and Bilevel mask ventilatory support (VPAP/BiPAP) at night. Please consult with the on call respiratory consultant for these children.
Follow-up and Contacts

- Please ensure that you have discussed the child with the Respiratory Registrar during the day, or the on call Respiratory Consultant after hours, before admission or discharge from CED.
- Please check the family have transport and a phone. If not, ensure that other measures have been taken so that contact can be made with emergency services.
- Ensure they have follow-up arranged and clear instructions on when they should seek further medical review if the child’s symptoms deteriorate or just don’t improve. Children may be reviewed by home care nurses in the community.
- If you are not sure what to do, you may contact the Bronchiectasis Nurse Specialist in working hours mobile 021412734

Bronchiectasis Clinics

- The Starship Bronchiectasis clinics are run by a team of health professionals. The clinics are coordinated by the Bronchiectasis Nurse Specialist (Una Wainevetau). The paediatricians are Dr Cass Byrnes Dr Naveen Pillarissetti and Dr Innes Asher from the Starship Respiratory service along with a Respiratory Fellow or Respiratory Registrar. The Bronchiectasis physiotherapist also attends
- The clinics are held weekly on a Tuesday morning
- Most children are seen 3-6 monthly and some have shared care between the respiratory service and their primary general paediatrician

Further Reading


