

PCD Day, June 2008

Pippa Hall, Children's Respiratory Nurse at the Royal Brompton Hospital came to talk to us. She gave an overview of PCD, explaining that it involved chest infections, ENT problems and school issues.

Cilia

She said that PCD was a genetic disorder that mainly affects the lungs due to abnormal cilia. Cilia are found in the lungs, ears, sinuses, the female reproductive tract and sperm. Cilia line the primary bronchus, and their function is to keep the lungs clean. The cilia should beat and circulate mucus in an upward fashion. This mucus is necessary for breathing and any debris is moved along by the cilia. However, in PCD this mucus is not moved in the normal way, and can become a prime ground for infection.

Signs and Symptoms

1. Breathing problems at birth
2. Snuffly at birth
3. Feeding difficulties
4. 50% of children have dextrocardia (mirror image arrangement of organs)
5. Long term cough
6. Runny nose
7. Bronchiectasis (lung damage)
8. Sinus problems
9. Glue ear/ear infections
10. Hearing problems
11. Sub fertility and infertility

Diagnosis of PCD

1. History
2. Nasal nitric oxide test (can be done after the age of 5), abnormally low levels of this gas in the nose can indicate PCD
3. Cilia biopsy/brushing

Aim of treatment

1. Main aim is to prevent chest infections and bronchiectasis (permanent scarring of lung and widening of airways)

Treatment

1. Physiotherapy
2. Regular exercise
3. Antibiotic therapy, oral and intravenous. For some a few courses are necessary, and for others prophylactic regular/preventive antibiotics are necessary
4. Nebulised therapy

5. Inhalers
6. Avoid passive smoke

Medical Management

1. Regular hospital check ups
2. Lung function tests (over 5 years old)
3. Regular physiotherapy reviews
4. Sputum checks
5. ENT reviews - this usually improves with age
6. Hearing tests
7. Hospital admissions
8. Intravenous antibiotics

What are the signs of chest infection?

1. Increase or new cough
2. Increase or change in colour of sputum
3. Temperature
4. Tiredness
5. Loss of appetite
6. Reduced exercise tolerance
7. Painful chest

What to do

1. Call local nurse for advice
2. See GP and provide sputum sample if possible
3. Increase the amount of physiotherapy
4. Make appointment to see the hospital specialist doctor

ENT advice

1. Regular review by ENT doctor
2. Treat ear infections
3. Clean ear avoiding water
4. Topical antibiotic drops can be used
5. Grommets are not recommended
6. Discuss any hearing concerns, speech and language specialists may be needed
7. SALT assessment can be done if there is speech delay
8. Saline douches for runny nose

School issues

1. Encourage normal school activities
2. Encourage sport
3. Address bullying as soon as possible
4. Keep school informed of any illness/hospital appointments
5. Physio may be done at school

Other issues

1. Try not to run out of prescribed medicines
2. Discuss having a spare supply of antibiotics at home
3. Keep up to date with immunisations
4. Make a list of questions before clinic appointments
5. May be eligible for Disability Living Allowance
6. If going on holiday, get letters for the plane if need medication and get medical insurance advice

Outlook

If appropriate and diligent care is undertaken from an early age, children and adults with PCD can lead normal lives. She stressed the importance of having the flu jab but said the BCG jab (for TB) was only advised in high risk areas or where you are exposed to people who travel a lot to where TB is still prevalent. In winter with central heating there may be more bugs around which can cause more chest infections.

Hannah Jay, Senior Physiotherapist on the Royal Brompton Paediatric Team followed on from Pippa. Hannah also has experience of working with adults at RBH.

Treating PCD

1. Physiotherapy
2. Exercise
3. Antibiotics - nebulised, oral and intravenous
4. Inhalers

What is physiotherapy?

Everyone with PCD should do physio every day to clear secretions from the lungs. Even healthy lungs produce sputum, which the cilia beat up to the throat where it is swallowed. Because in PCD the cilia do not beat properly, the mucus settles, bugs grow in it, the lungs become inflamed and infection results. Physio helps to break this cycle, but infections can still occur.

Physio involves breathing techniques which move the secretions - active cycle of breathing. By taking deep breaths the air gets behind the secretions and moves them along.

Active Cycle of breathing

This involves taking deep breaths, resting and huffing(which allows you to assess how the lungs sound).Huffing should be done with an open mouth at the end of every cycle to cough and clear the lungs.

With young children (two and a half to three year olds) blowing games with bubbles and candles are recommended. Musical instruments are also recommended as a form of physiotherapy.

Postural drainage

Postural drainage uses gravity to help drain different areas of the lungs. A tipping bed,

two pillows, or a wedge can be used so that the head is lower than the hips. In PCD it is the middle and lower lungs that are mainly affected. Seven to ten minutes of physio is recommended in each position whilst breathing exercises are done. However, if a child has reflux despite medication, then tipping is not recommended.

Patting/shaking/clapping/vibrations help to loosen secretions. The clap is done whilst the patient takes four deep breaths, then a shake to squash down the lung volume, four sets of shaking, then huff - 5 -10 minutes on each side. Active cycle: 30 seconds of patting, 30 seconds of rest, huff and cough. Can sit up to cough. Good posture is needed for huffing and coughing to get more air in.

Adjuncts

When children get older they can use adjuncts. These devices use breathing out against positive pressure, which keeps the airways open. Children as young as 3 -4 years old can use a pep mask. Some devices like the acapella and flutter also use oscillations and vibrations. These vibrations should be felt in the chest and not the cheeks.

However, these devices can harbour germs and cause infection if they are not cleaned with soapy water every day, and sterilised every week. Ask about adjuncts at clinic. The PCD Family Support Group can help you get an adjunct if your clinic doesn't have them.

Autogenic drainage

This is a specialist way of breathing using different lung volumes, and requires concentration so is more appropriate for older children and adults.

How much physio and why?

Physio should be done twice a day for 15-30minutes. If you need longer maybe your techniques need reviewing. Listen to the huffs before and after physio, as they give an indication of how much dryer the chest is.

Inhalers

Blue inhalers and Dnase should be used before physio. Steroid inhalers and antibiotic inhalers should be used after physio.

Practical Tips

Physio should be a routine like brushing your teeth.

1. Reward charts with reward stickers can work very well
2. Blowing games
3. Trampoline
4. Tickling
5. Laughing
6. Acapella with cycle of breathing

What if physio is not working?

1. If sputum is too thick this may be due to lack of hydration - drink plenty of water
2. May need antibiotics

3. If wheezy an inhaler or pep might help
4. If bored may need new technique

You should ask to see a physiotherapist because the physio may need re-evaluation. If you have any problems ring the Brompton physiotherapists who have contacts and maybe able to help.

If unwell

1. Increase physio
2. Get sputum test/cough swab

Exercise is really important but should not replace physio. Timing exercise with physiotherapy makes the physio more effective.

Breathing control in children with PCD is better than normal children. Musical instruments that promote breathing control are important. Adjuncts are important for holiday. Acapellas go rusty, but the manufacturers say that it does no harm.

Sputum tests should be as fresh as possible. Can ring specified nurse and get results. If there is no feedback it should mean that there is no growth of bugs.

After a fantastic lunch Fiona Copeland chaired the AGM. The main points of the AGM was what a great year we have had for fund raising and raising awareness of PCD. For the next 12 months we are planning to produce new multimedia information about PCD for use by families and the medical profession.

The children were entertained with football tables, a bouncy castle and [Science Boffins](#).