

PCD Day 2014

Another successful PCD Day was held on Saturday, June 21st at the David Lloyd Centre, Milton Keynes. We had over 50 adults attend with 12 children.

Supported by



The day was split up by a number of speakers, a delicious lunch and the children being entertained by the Great Gappo and the David Lloyd Kidz Club.

Many thanks for all the speakers who gave up their time to come and talk and to answer questions and to Genetic Disorders UK for their grant.

This report is deliberately long for the benefit of those that could not attend. We have included post event feedback from some of the families that attended.

Any questions please contact Fiona
chair@pcdsupport.org.uk



Joanne Rimmer, Managing ENT issues

In PCD the problem caused by cilia issues is that the mucus does not move properly. In CF it is that the mucus itself is much thicker than normal. 70% of neonates with PCD are found to have respiratory distress.

It is common for all children to have snotty noses and ear problems. In PCD the snotty nose will usually be continuous; in other children the nose will be normal sometimes.

Nasal polyps are common in CF but less common in PCD. Adults may present at diagnosis with chronic sinusitis, glue ear, atypical asthma and chronic cough.

Ear Infections

All children may have recurrent ear infections, (acute otitis media), a bulging eardrum with lots of pus behind it. If left, the eardrum perforates, the muck comes out and the perforation heals itself in 99% of cases.

Studies show that giving antibiotics may reduce the infection by only one day. Antibiotics may be given to children less than one who are very unwell. If the ear infections recur it is advisable to try a one month course of low dose antibiotics.

For normal children who experience recurrent ear infections, grommets are inserted to let air into the ear. In PCD grommets are not normally successful as the ear will keep discharging and hearing will not improve.

Glue Ear

Glue ear (Otitis media with effusion) is not painful but is an infection and causes hearing loss.



20% of all children will get it, peak ages between 2 and 5 years old. If the hearing loss lasts more than 3 months there is concern for speech and language development. The hearing loss is *conductive*, as opposed to the loss experienced in old age which is *sensorineural*.

A hearing test will show the patient can hear much better with a bone conduction test, hearing through the bone, than through the ear, which is blocked up. For a child with PCD, hearing aids may be prescribed which will improve the hearing. Grommets may be given to an older child if they are stable medically.

Nose Issues/Rhinitis

Rhinitis is the inflammation of the lining of the nose. This can stop mucus from draining, so a saline douche (e.g. Neilmed Sinusrinse) may be suggested. This is a pinch of equal parts of salt and bicarbonate of soda, dissolved in warm water and sniffed up one nostril to drain out of the other and clean the nose. Sinusrinse sachets can be bought, or you can make your own mixture. The volume of water is important in shifting the mucus, a product like Sterimar spray will not be so effective.

Adenoids

Enlarged adenoids are rare in adults, they are at the back of the nose and reach their maximum size by age 5. They can cause snoring and/or sleep apnoea. If the lungs are healthy enough for a general anaesthetic then they may be removed.

Related directly to experiences, very systematic and easy to follow

Useful to know glue ear gives intermittent hearing loss

Rhinosinusitis

Cilia in the nose normally push mucus back and down the throat.

Cilia in the Maxillary sinus (cheeks) circulate the mucus round and out of a drain hole into the nasal cavity, rather than just using gravity. If the cilia don't work the mucus just sits at the bottom of the sinus. Sometimes surgery is done to create another opening lower down the sinus that is more gravity dependent, this also aids the saline douche to reach the sinuses better.

Polyps

This is inflammatory tissue that can cause a blockage, sometimes helped by saline douche or may need removal surgically.

Throat

Phlegm and post nasal drip is common and many people don't notice it. If people are aware they may cough to try to shift the mucus, this can irritate the throat. It is better to drink something to wash the mucus away.

Research

There has been a small study on the role of cilia in the balance system. The study showed that the cilia in the ears helps with balance but none of the patients who undertook the research had any issues or symptoms of lack of balance. It is thought that the brain learns to compensate for any reduced balance function in people with PCD. This report will soon be published in the Journal of Otolaryngology and Neurology.

Q&A

Steroids can be good if the nose lining is inflamed. Surgery is not normally performed on children/young adults until the sinus' is fully developed.

There are specialists in ENT (rhinology) at the Brompton, Southampton, Birmingham and Leeds.

It was recommended that PCD patients limit the use of decongestants, as they shrink the lining of the nose and use for only a few weeks can damage the lining and be hard to correct. Use only for a few days and discard.

Wendy Sale, Claiming DLA

DLA is non means tested, so it doesn't depend on how much money you have. It is meant to make life easier for the disabled person.

The benefit is assessed on the diagnosis of the individual, and how their mobility and care needs are affected.

The children's form is straightforward, and has a good guide leaflet which is well worth reading before filling in the form. Most PCD claims will be about care needs. There are 3 rates of care, e.g. if 2 x 30-minute physiotherapy sessions are needed every day that can qualify a child for the lowest rate of care.

The form is straightforward, with no trick questions. Make sure that you list any aids that a child may use e.g. acapella, a spacer for inhaler, nebulisers.

Department for Work and Pensions (DWP) will never see your child so write down in short sentences a breakdown of what you need to do for your child each day, so it can easily be understood by the decision maker. Medical letters from consultants are not always helpful as DWP just wants to know if your child needs help to dress etc. Put how the disability affects your child rather than generalisations about the condition. You can send a copy of your prescription form, or physiotherapy instructions, but too much paperwork can put DWP decision maker off!



It can be good to get the form, as it is hard to you do. A friend could Advice Bureau (CAB) or Make an appointment as

Useful tips
Good Update with clarification on PIP

someone to help you with look objectively at what help, or your local Citizens Community Law Centre. soon as you receive the

form as there is a deadline to get it back. If there is a delay, ring DWP to explain and give them the date you can get the form to them. Be accurate on the form as later alterations weakens your case and if someone fills in the form for you make sure they fill in their contact details on the form and/or provide a covering letter. This is important if the application goes to a tribunal. Keep a photocopy of the form and a proof of posting.

If you are refused, you will get a letter back, and you have one month from the date of the letter to write to them asking for 'a mandatory reconsideration, a complete statement of reasons the benefit has been refused and state that you are going to submit medical evidence to support your application'. It is very important that you use these words exactly.

It can take a long time for DWP to get back to you, they have no time limits on their side, and if the answer is still no you can submit an appeal to go to a tribunal. A tribunal is both a medical and legal person who will look at your case and make a decision. You can ring the DWP if things change while you are waiting and submit a completely new application. Your benefit will still be backdated if the tribunal finds in your favour.

Up to 16 it is DLA to apply for but after that it is Personal Independence Payment (PIP). All adults on DLA will eventually be transferred to PIP, even if they have been given an indefinite award. PIP is a tick box benefit, usually awarded after a medical assessment.

Bea Redfern, The Power of Positivity

Bea is a 22 year psychology student who has PCD. She spoke about how to manage your health



beyond the physiological aspects. Bea believes that the positive psychology can help reduce stress levels and that you should follow your strengths rather than your negatives.

Stress is bad for health, generally, and with PCD causing its own stresses, this can affect your effectiveness e.g. this can be particularly bad during exams.

Bea recommends that you do things you enjoy as positive emotions can help build up resilience.

Studies show that positivity, gratitude and mindfulness play a part in recovering from illness.

Just being at PCD Day is a positive, accepting your condition and trying to understand it better.

There are no text book rules, it is what works for you, and it is not about being happy ALL the time, which is unrealistic.

Mindfulness: pay attention to the "now" using concentrating on breathing to reach a calm state, if used regularly you can develop a mindfulness muscle, to help you cope with stress.

Try to be grateful for what you do have... e.g. one good thing that happened today, savour your food, thank someone, help others.

Q&A

There was a discussion about the benefit of being open about our health condition.

Laura Behan, PCD Researcher, Southampton

Laura invited everyone to help develop the Quality of Life Questionnaire that can be completed online or by post, at intervals over a period of time. It will help show the response to treatment and inform doctors for future use. There is also a new questionnaire about experiences with the diagnostic process, this will be on the PCD website soon.

Great that we are looking at all sorts of areas.

Very useful to hear how PCD affects Bea's life

Lynne Schofield, Effective Physiotherapy

Lynne spoke about how the importance of doing quality physiotherapy and not just doing it twice a day. She recommended that you become an 'expert' on your own chest so you know when it is good and when you might be getting a chest infection.

It is acknowledged by the PCD teams that keeping well and doing physiotherapy can be a challenge along with all the other things you want to do.

This is why it is so important that you regularly review your techniques to ensure that you are getting the best treatment.

Lynne demonstrated how to breathe in softly to get behind the sputum, pause, blow out. You can be using an acapella, flutter or other device while doing this. You don't need to count a specific set of breaths, do what feels right for you. Rest, then start again. Huffs help the sputum to move along so you only need to cough right at the end to bring the sputum out. Doing more huffs than coughs saves energy! Do slow huffs, then faster once you feel the sputum moving up. Where possible, spit the sputum out, so you can check the stickiness and colour. Rest if your airwaves are getting squeaky and tight, try again later.

Useful to see all the different pieces of equipment

Great use of animation – very good

Interesting overview and ideas

If you have a nebuliser to open airways, use before physio, or take Salbutamol. Take nebulised antibiotics *after* physio or you may end up coughing the antibiotic out again.

Do your physiotherapy when it is best for you, but preferably not straight after a meal. If sitting, don't slouch, allow the air to get right into your lungs. Don't have distractions, TV, etc, you need to concentrate on getting the breathing right.

Drinking can make a big difference to getting sputum to move, so make sure you are hydrated.

Try different positions, see what works for you. Do an exercise that you enjoy, anything that makes you breathless helps move the sputum. Focus on staying well, win the sputum battle!



Q&A

To help children take responsibility, try using different aids, do shorter sessions more often. If the child can feel the benefit of the physio it will help.

Using nebulisers abroad, check they will work? [See website](#)

Adults should ask for physio reviews regularly as they are not currently offered as in paediatrics. It is good to have a regular review as things can change subtly over time. Adults, reward yourself for physio too!

Milk: there is no evidence that milk consumption leads to more mucus production

Question and Answer Session

Sleep problems: good to have a physio session before bed, or in the position you will be sleeping in.

Nasal douching before bed can help stop post nasal drip. If a long term problem a sleep study could be done.

Bad breath: can be due to bad nasal hygiene so nasal douching can help.

Alcohol: individuals will work out what affects them. It is good to keep hydrated.

Vit D is show to help people fight infections.

Dental Care: rinse out mouth after taking any medication, puffer etc.

Salbutamol may not work for everyone, wheeze may be caused by sputum rather than a tight chest. If there's a rattle then it will be phlegm.



Anxiety Attacks: psychological support may be available, could be in schools. The [Ready Steady Go](#) transition programme to adult PCD care may help.

There was a discussion about whether we should have a children's discussion board... it was generally agreed that this might be difficult to manage. Please contact the PCD Family Support Group if you would like us

to put your child in contact with another child of the same age.

AGM

Fiona introduced herself and explained that this was here 11th PCD Day, and she has now been the Chairman for over 10 years! Thanks to Jeans for Genes for the grant to host the event.

Based on our objectives we have achieved the following in 2013/14:-

1. Continued to support families via:-
 - Email
 - Telephone
 - Newsletters
 - Facebook
 - Website
 - Family Days
 - Patient Surveys

Excellent and thoughtful answers to questions

Always good to give people chance to ask things

2. Raised awareness of PCD by:-
 - 27k visits to website
 - Facebook , Twitter & newsletters
 - Jeans for Genes Day
 - Attended Rare Disease Day at House of Commons
 - BTS Conference

3. Promoted research by:-
 - Supported the Biomedical Research Unit at the Royal Brompton Hospital with Patient and Public representation on the Chronic Suppurative Lung Disease Consortium
 - Continued to work as part of the Ciliopathy Alliance (the group will be represented at the Cilia 2014 Conference in Paris in November)
 - Helping recruit people for PCD research

4. Getting the best care:-
 - Helped to continue the development of the Diagnostic and Paediatric Management Service (all centres now up and running – annual assessments now underway)
 - Helped with the re-issue of the PCD Guidelines
 - Helped the Adult Teams develop a bid for funding for Adult care
 - Hosted PCD Medical Board Meeting (over 60 interested clinicians attended last meeting at the BMA in London).

5. Fundraising to support above activities by:-
 - London Marathon
 - Sponsored walks
 - Jewellery selling
 - Sponsored silence
 - Dryathon
 - Number of families asking for donations in lieu of gifts
 - Regular giving through Direct Debits



Thank you for the hard continued hard work and the excellent maintained focus of the group

Treasurer's report

Mick Wilkin (now in 10th year as Treasurer) explained:-

Cash reserves maintained at circa £20.1k

Fundraising and donations - £5,442 (£13,145 l/yr).

Income from NHS - £6,000 (£1,500 l/yr)

Interest received £4, (£4 l/yr)

Costs – £8,003 down from £15,119 including:

£2,997 for Web hosting fees

£1,092 on 2013 Summer Social and AGM.

£1,055 Conference Costs



Thanks were given to Ian Lowry of DNG who kindly independently examine our accounts free of charge. It was voted that they should be asked again for the 2014 accounts.

Mick explained that we only have regular income of NHS funding for website £6k per annum and some a few regular monthly contributes via direct debit. If anyone would like to set up a regular contribution this is easy to do very the Justgiving website. We would also welcome any fundraising activities to boost income – t-shirts, sponsorship instructions and collecting boxes all available.

Plans for 2014-2015

- Continue to work with the NHS to ensure the smooth running of the new paediatric PCD Management Service
- Helping to develop transition and adult service
- 'Northern' Family Day 11th October 2014
- Cilia 2014 Conference in Paris in November
- Fundraising plans include an [Annual Draw](#) and [Grand Union Challenge](#) London to Bletchley in June 2015 to raise funds and raise awareness. You are welcome to join in for all or part of the route.



Reappointment of Committee

The following committee members were reappointed for another year:-

Fiona Copeland (Chairman)
Mick Wilkin (Treasurer)
Myra Tipping (Secretary)

General Committee Members:- Sylvie Prouse, Liz Meleady, Gary Tipping, Terry Irwin & Sarah Kirk

Beatrice Redfern was also appointed as a general committee member.

Charlotte Kewell has volunteered to help with marketing.

Fiona to check with Justine Curry and Nicky Keech that they are happy to remain on the committee.

Fiona asked for volunteers to join the committee. We meet approximately 3 times a year in Milton Keynes on a Saturday morning. Please let us know if you would like to attend the next meeting on 6th September 2014.

Any Other Business

Amanda asked for feedback on our Ready Steady Go programme for newly diagnosed adults.