Northern PCD Day 11th October 2014

The second Northern PCD Day was a great success. It was held at the Thornbury Centre, Leeds Old Road, Bradford, and was organised by Lynne Schofield and the Leeds and Bradford team. It was well attended and attracted lots of new families.

The children had a great time and thoroughly enjoyed themselves. Barrington Powell, a children's entertainer' performed magic tricks and made balloon animals whilst the adults listened to the speakers.

Fiona Copeland, Chair of the PCD Family Support Group, opened the proceedings, and outlined the aims and role of the PCD Family Support Group which was set up in 1991. She explained that the group provides support to patients and carers, brings PCD to the attention of the medics, promotes research, supports the NHS to ensure that patients have diagnostic services and fundraises to support its activities. Support is offered by email, telephone, newsletters, Facebook, the PCD website, Family Days and patient surveys. She encouraged everyone to both buy and sell raffle tickets for the gold PCD angel necklace worth about £350. She explained that the organisation raises awareness through the website and this had 27,000 visits this year. Awareness is also raised via Facebook, Twitter, newsletters and Jeans for Genes Day, as well as Rare Disease Day at the House of Commons, the BTS Conference, and the British Lung Foundation. The group also support the Biomedical Research Unit at the Royal Brompton Hospital, are members of the Ciliopathy Alliance, host the PCD Medical Board Meeting and also recruit patients for PCD research. The group aims to help patients obtain the best care, and is currently helping the adult teams to obtain funding for a specialised adult care service.

All these activities are supported by fundraising including marathons, sponsored walks/silences/dryathons/jewellery sales and direct debits. The organisation is a going concern, and the finances are helped by these activities and by an income of £6000 from the NHS for using the PCD Family Support Group website.

Plans for the near future include working with the PCD Management Service, helping with the transition to the adult service, the Cilia2014 Conference in Paris and attending the BTS Conference. Fiona explained that the organisation is run by volunteers and appealed for new committee members.

Dr. Phil Chetcuti, Consultant Paediatrician and Neonatologist at Leeds Teaching Hospital and NHS Trust, gave an interesting and informative talk about PCD. He praised the achievement of the setting up of National Clinical Service with its centres at the Royal Brompton Hospital, Leeds, Bradford, Leicester and Southampton. There are about 350/400 children with PCD in the UK, 140 in the North of England and 70 in the area of Bradford. The service has enabled children to be assessed in Liverpool, Hull and Manchester too, and enabled home

nursing/physiotherapy visits. In the next year or so we will have a lot more information about PCD, more than anywhere in the world. This will hopefully advance management of the condition. There is interest in PCD in Europe and we will link with them and this will facilitate good management of the condition. It is challenging because we want PCD patients to be as healthy as possible, successful and happy, with the family involved.

He explained how cilia move, and demonstrated this with everyone waving their arms. In PCD the cilia do not move properly. There is not just one abnormality but many, and these can lead to faulty function of the cilia.

He stressed the importance of early diagnosis in order to prevent lung damage. Patients with dextrocardia are much easier to diagnose, and these account for about 40% of cases. Over 50% of PCD patients have breathing difficulties in the new born period. The inner arm and outer arm defect in the cilia is common, but there are a wide variety of other abnormalities.

By the age of six children are capable of doing lung function tests. These tests reflect the degree of airway damage, and are carefully monitored. The aim is to ensure that lung function does not decline. Chest X-rays only show lung damage, but CT scans are much clearer and give much more information. Usually in PCD it is the lower and middle lobes that are affected. Sputum cultures give very good information about bacteria present and antibiotic sensitivities. Cough swabs are not as good as sputum samples for showing this. Aggressive antibiotic treatment is required if pseudomonas is present.

Audiology is also important in PCD. Children should have an annual hearing test because education can be affected by poor hearing. Hearing usually improves as the children get older.

He went on to talk about transition from paediatric clinics to adult clinics. He pointed out that there are not enough respiratory physicians to deal with PCD, and that CF physicians may be best to deal with PCD patients if it is not possible to see a PCD specialist.

PCD patients have a wet cough every day, and this is the nature of the condition. Because of this, it can be difficult to decide when antibiotics are necessary. Generally if the cough is worse, with darker sputum for about 5-7 days, then antibiotics are likely to be required. Some coughs are caused by viruses and may settle down. Antibiotics are really important in order to avoid lung damage. It is a balancing act to decide which antibiotic, how long for,

and whether prophylactic antibiotics should be used.

Genetic studies have been undertaken and they may provide the pathway to treatments. Up to 50% of patients can now be identified from blood tests.

Good physiotherapy is key in the management of PCD, with early use of antibiotics, lots of exercise, good nutrition and regular assessment of ENT issues.

Dalvinder Helliwell, PCD paediatric dietician at Bradford Royal Infirmary, then gave an interesting talk on diet and PCD. She stressed the importance of assessing, monitoring and providing on-going support to PCD patients. The aim is to achieve normal growth. PCD patients lose energy by coughing and also there may be nutrition loss in sputum. They may lose weight and may have a reduced immune system due to recent infections. Children are often referred because they are not gaining weight and have a poor appetite. They may have increased requirements during illness and in a minority of cases may need tube feeding. Too much weight also needs management because obesity with a lung condition is a massive problem. A balanced diet is required, and a high calorie diet is needed if appetite is poor. Supplements are sometimes required.

She spoke about dietary beliefs and dietary symptoms and said that patients had implicated the following foods in relation to worsening symptoms (i.e. more coughing and more phlegm):- dairy products, fried foods, sweet dishes, bananas, oranges and grapes. Foods that were thought to be beneficial include fruits, boiled vegetables, ginger and honey. Remedies that were suggested were water, herbal teas and fennel seeds. She said that there was no evidence that milk caused extra mucus, and emphasised that if milk was avoided then the nutrients it provides needed to be replaced in the diet.

There is growing evidence to support beneficial links between antioxidant rich foods and respiratory symptoms. Antioxidant foods include those with vitamin A, C and E. Vitamin A is found in dark green leafy vegetables, cheese, milk, meat, liver, yellow and orange fruits and vegetables including sweet potatoes, apricots, pink grapefruit, carrots and pumpkin. Vitamin C is found in citrus fruits, red and green peppers, broccoli and some juices. Vitamin E is found in vegetable oils, margarine, nuts and seeds. She did not recommend eating grapefruit if the child is taking Azithromycin antibiotics.

She said that patients could ask to be seen by the CF dieticians. Honey was thought to be helpful if coughing at night. She said that multi-vitamins could be taken, but care should be taken not to exceed the recommended dose. She emphasised that they are intended only as a supplement, and not as a replacement for a nutritious diet. In PCD patients vitamin D

vitamins are often found to be low or reduced, and a supplement could be helpful here. She recommended that children should be exposed to the sun for ten minutes before applying sun cream, and that this would help with vitamin D levels.

She also spoke about the difficulties some patients experience with obtaining antibiotics from GPs and encouraged patients to obtain action plans and to take copies of clinic letters to the GP and to contact the consultant if there were problems.

Lunch ensued, and there was a lot of social activity with everyone (including the speakers) mingling and chatting together.

Lynne Schofield, Paediatric Physiotherapist at Leeds Hospital, then gave an interesting and imaginative talk. She stressed not only the importance of physio but also the quality of it, and the need to do 5 cycles, with ten breaths that would take about 15 minutes. She stressed the importance of doing physiotherapy until the chest is clear, and making the activity both fun and effective. She talked about "sputum police" and illustrated her talk with pictures of the Pink Panther. She talked about the active cycle of breathing and breathing in slowly through the nose if possible. She said that breathing in quickly could just push the phlegm down. She suggested sneaking the air in, pausing, and then blowing out. Various devices can be used like pep masks etc. to help. She explained that the big airways are usually the first to clear and the importance of clearing from the bottom of the chest. She said that huffs should be done using tummy muscles. It is important not to huff too hard because this could cause the chest to become squeaky and more difficult to clear. She explained the importance of having rests and relaxing the airways during physio. She stressed that if there are problems with physio, then it should be reviewed. She also said it is important to spit out the sputum because it can make the patient feel sick and that children are usually six before they are able to do this properly. The colour of the sputum should also be examined to check for infection. She recommended the use of nebulisers and saline before physio and nebulised antibiotics (if required) afterwards so that they can be more effective. Physio should not be done after eating and should be done without too many distractions because it is important to concentrate on the breathing techniques. She explained that mucus is like glue and if the child is dehydrated, then the sputum becomes thicker and more difficult to clear. Therefore drinking plenty of water is important. The smallest airways are just half a millimetre wide and can easily get blocked. Different positions are required to clear different parts of the lung.

She explained that exercise is very important and anything that makes you breathe harder is beneficial, ideally for 20 minutes or more.

She stressed the importance of focussing on staying well and winning the sputum battle.

Beatrice Redfern, a 23 year old PCD patient, and Family Support Group Committee Member, spoke about her experiences. She gave us a fascinating insight into her life with PCD. She explained that she was diagnosed when she was fifteen years old, although she had realised that she was "a bit different" before that. She became more aware of her cough and runny nose as she grew older. She now understands her condition, and therefore is more in control and able to manage it. She said that physio is the main focus for her, and she also takes regular exercise in the form of running. She emphasised the importance of managing stress in her life because when she is stressed she gets chest infections. She manages her stress levels by planning for exams. She said that by taking care of body and managing her stress, she is able to be well.

She went to university and initially did not reveal that she had PCD. However she has since concluded that it is better to be open about her condition because she was then able to have extra time in exams. She recommended obtaining a letter from the GP and then giving it to the new GP at university. She spoke of the need to be proactive. She also stressed the need for accommodation without damp as this would be detrimental to health. Her current concerns are job interviews, and whether to declare her medical condition. She asked for advice from the floor in relation to this.

We were then shown two videos, one with a ten year old and the other with an eleven year old describing how PCD affects them.

Saj Latif then spoke movingly about how PCD affects his five year old daughter and his family. He has two children, one of whom has PCD. When his daughter was born she spent the first 24 hours in intensive care, the next three weeks in a high dependency unit, and was given antibiotics and oxygen. When she was three weeks old she was very fortunate to be spotted by Dr Chetcuti, and diagnosed with PCD. He spoke about the difficulties of coming to terms with this. He explained that sometimes they had missed doing physio because his daughter did not like it, and they had not realised its importance. She had to have intravenous antibiotics in hospital and needed a lot of medication. However, after the first year they understood the importance of physio, and now never miss it. She has physio twice a day, and is as active as possible. She is now much better and not on medication apart from a nebuliser.

He went on to talk about the impact of PCD on family life and the difficulties of trying to explain to his family and extended family. His daughter is now in year 1 at school and she has physio for 20/25 minutes before breakfast and again later in the day. He stressed the importance of exercise and how this can be more difficult in the winter so they take her to

indoor play centres then. He also described the impact on her two year old sister who does not have PCD, but sometimes has physio too so that she does not feel left out.

A lively question and answer session then ensued with everyone getting involved and talking about PCD.