

What is PCD – leaflet for patients

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1.Pre-Birth

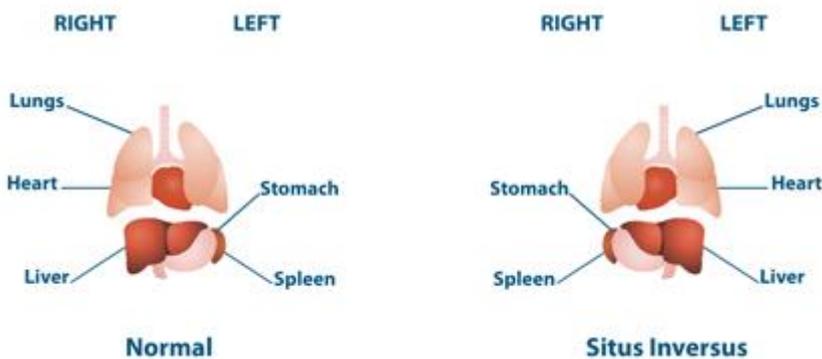
1.1 Pre Natal Scan

Situs Inversus, Dextrocardia

A diagnosis of situs inversus or other situs abnormalities may be made on antenatal scanning during pregnancy and such a finding may be a clue that the baby might have Primary Ciliary Dyskinesia [PCD].

In many cases this may result in the obstetrician referring the mother for specialist fetal echo scans to determine if there is a cardiac defect.

In any event the baby should be referred to a specialist diagnostic centre post-natally to exclude PCD, which is a common, but not the only cause of situs abnormalities.



1.2 Genetic Counselling

PCD is inherited via the autosomal recessive route, which means both parents pass a PCD gene to an affected child.

The chance that a child will be affected when both parents carry a PCD gene are 1:4. There is a 1:2 chance of their children being carriers and a 1:4 chance they will not inherit a PCD gene from either parent. In summary, any patient with PCD must have inherited a gene mutation from each parent, both of whom must be carriers, i.e. carry only one gene mutation, or have the condition themselves in which case they will also have two gene mutations.

In some communities, such as some Asian populations and some Caucasian communities, where intermarriage within the extended family is common the incidence of PCD may be significantly higher. This is because in any family carrying a genetic disorder there is a higher chance that the genes will be combined in a family and handed to their offspring.

This increase in incidence is typical of any recessive condition where intermarriage is common. It is becoming clear that prevalence 'hot-spots' are occurring where PCD is more prevalent in the community because of a higher degree of related marriages.

Genetic counselling can be sought from any specialist genetic centre, where advice can be sought for future family planning.

There are a large number of gene mutations causing ciliary defects resulting in PCD. Only some of these genes have been identified and as yet there is no genetic screening or diagnostic facility, although research continues rapidly in this aspect of diagnostics for PCD.

2. Birth to school

2.1. Special Care/Hospital Stays/IV antibiotic Treatment

If your child is very unwell he/she may be admitted to hospital for further investigations and treatment. The most common reason for a hospital stay is a chest infection, which needs to be treated with intravenous antibiotics every day for two weeks. That means your child will need to have a 'line' inserted, usually in a vein in their arm, for the antibiotics to go into their bloodstream. Your child will be given a local anaesthetic beforehand so it won't hurt, although it can be quite distressing for the parent to watch.

Your child will stay on a paediatric ward with other children. Sometimes if they are growing a specific bug in their sputum they will be given their own room to avoid cross infection with other children on the ward.

Many parents stay with their children and sleep on a camp bed by the child. Some hospitals also have special accommodation for parents.

Going into hospital can be very overwhelming for children, as they don't know what to expect. If they haven't been into hospital before there are books that explain what it is like and they may help reassure your child.

2.2 Hospital Stays and IV Antibiotic Treatment Continued

You'll also be allowed to stay with your child while they are having their treatment. There are lots of distractions around, like music, mobiles and toys and you can bring in your child's favourite games. If there's anything at all that you're worried about, talk to the nurses - **they're there to help.**

For young babies you will be supplied with bottles and teats. Most hospitals have a selection of baby foods and ready-made bottles of infant formula.

Whilst your child is in hospital they will be given regular physiotherapy sessions by the paediatric physiotherapists.

Paediatric Respiratory Nurse Specialists – he/she can offer support and advice to families with PCD children.

Hospital Social Workers – the social workers offer help and advice to parents and families. They can offer information about sources of help available and, where appropriate, support parents with applications, such as for financial assistance, and help when dealing with statutory and voluntary organisations like housing associations.

Interpreters – the hospital can arrange for interpreting services if you think you or your child will need them whilst you are staying in hospital. If possible please let the hospital know before your stay and which language you need the service for.

Some children who require frequent intravenous antibiotics may be offered a portacath, a device surgically implanted in the upper chest, just below the clavicle or collar bone, to allow easier access to the bloodstream. These devices last for approximately for 10 years if they are properly maintained. When they are not being used they have to be ‘flushed’ once a month – this will either require you going to the hospital or sometimes the community nurse will come to your house.

2.3. Diagnosis and acceptance of condition and possible future problems

Diagnosis is only made in only a few specialized centers around the world. In Cyprus diagnosis is made in the Department of Electron Microscopy and Molecular Pathology in Cyprus Institute of Neurology and Genetics, Nicosia, Cyprus.

Screening is available, via a non-invasive test measuring the level of nitric oxide [NO] from the nose, for most children greater than 5 years old. Very low levels of nasal NO are very specific for PCD, but the diagnosis can only be confirmed by analyzing the ciliary function and ultrastructure from a nasal brush biopsy.

Abnormal function or defects in the cilia ultrastructure provide a diagnosis in the majority of cases, although in some patients who have clinical PCD it may not yet be possible to confirm the diagnosis in this way. Research is ongoing to develop more advanced techniques to assist in these more difficult to diagnose cases.

All suspect patients in Cyprus are referred to the Pediatric Pulmonology clinic in Archbishop Macarios III and appointments are usually available within the next two weeks. Initial screening and ciliary function results are available the same day, but ultrastructural analysis takes about 4-6 weeks from the time of brushing.

For a small number of patients it may be difficult to confirm the diagnosis using current methods of diagnosis. In these situations where the clinical picture is typical and other evidence exists, such as low nasal NO levels or an abnormal ciliary beat pattern, patients should be treated in exactly the same way as confirmed cases.

The advent of genotyping for all PCD gene mutations, advanced electron microscopy techniques and molecular staining will hopefully overcome this diagnostic dilemma for many such cases as well as greatly enhance our knowledge of the underlying disease mechanisms.

2.4. Coming to terms with the diagnosis

Having a disabled child affects all members of a family. Mothers and fathers can sometimes react in different ways to the news that their child has a disability or medical condition. Many parents describe feelings of isolation; struggling to come to terms with the news; a lack of time for themselves and each other; problems balancing work and caring; increased financial worries; a lack of support and understanding from professionals and the wider family network; having to learn all about how the Cyprus national system works as well as worrying about your child.

It is really important that you talk to someone about the diagnosis. Try to make use of all your support networks - it might be your husband/wife or partner, relative, friend or neighbour. It might be helpful to try and just get some time to yourself or to spend some time alone with your partner.

Sometimes there is a key professional who can open the door to lots of information or contacts. This could be a health visitor, occupational therapist or person working for a voluntary organisation. Ask your GP, health visitor or social worker for more information or directly contact the Pediatric Pulmonology clinic in Archbishop Makarios III Hospital.

After the initial diagnosis you may have loads of new questions for your consultant. It may be helpful to make a list of the questions you have for your next consultation.

You might find a professional counselling service a helpful way of unloading some of your thoughts and feelings. Your GP should be able to tell you about any local services.

2.5. Immunisations/Vaccines

It is recommended all routine childhood vaccinations are given at the usual times and should be arranged by the general practitioner.

When to immunise	Diseases protected against	Vaccine given
Two months old	Diphtheria, tetanus, pertussis (whooping cough), polio and <i>Haemophilus influenzae</i> type b (Hib) Pneumococcal infection	DTaP/IPV/Hib + Pneumococcal conjugate vaccine, (PCV)
Three months old	Diphtheria, tetanus, pertussis, polio and <i>Haemophilus influenzae</i> type b (Hib) Meningitis C	DTaP/IPV/Hib + MenC
Four months old	Diphtheria, tetanus, pertussis, polio and <i>Haemophilus influenzae</i> type b (Hib) Meningitis C Pneumococcal infection	DTaP/IPV/Hib + MenC + PCV
Around 12 months	<i>Haemophilus influenzae</i> type b (Hib) Meningitis C	Hib/MenC
Around 13 months old	Measles, mumps and rubella Pneumococcal infection	MMR + PCV
Three years and four months or soon after	Diphtheria, tetanus, pertussis and polio Measles, mumps and rubella	DTaP/IPV or dTaP/IPV +MMR
Girls aged 12 to 13 years	Cervical cancer caused by human papillomavirus types 16 and 18.	HPV
13 to 18 years old	Diphtheria, tetanus, polio	Td/IPV

Influenza immunisation for children over 6 months of age is recommended and can be arranged by your GP. The vaccines are usually available in October each year. If a child is receiving it for the first time, a 2nd dose is repeated 4 weeks later, otherwise it is a single injection each year. For some of the needle phobic children, you can ask your hospital to carry out the immunisation. It is also recommended that parents receive the vaccine.

Pneumococcal vaccine (Pnuemovax) is often recommended for children and adults with PCD. Check with your consultant whether you should have it.

Check with your consultant what immunisations your child requires.

3. Starting School

3.1. School

It is important for you to choose a school that meets your child's educational and welfare needs. If possible find a school that is close to home or is easy for you to collect them if they are poorly or need to attend a hospital appointment.

Before your child starts at the school arrange to meet the head teacher and explain how the condition affects your child. Provide them with information in the form of leaflets and tell them that they can contact the Pediatric Pulmonology clinic in Archbishop Makarios III Hospital if they need any further information.

If your child requires physiotherapy at school find out if the school can do this. Then ask your Respiratory Nurse Specialist to arrange a school visit with a physiotherapist to explain what is required. If they cannot, ask your physiotherapist to review your current physio routine to see if any adaptations can be made.

These are the some of the questions you may want to ask the school:

- Find out if the school can give medication?
- Ask about the procedures to ensure your child gets the medication?
- Ask if your child can sit at the front of the class. If he/she has hearing loss.
- Does your child need to be reminded to wipe their nose? Can they have a box of tissues near their desk? Can you they have a bin near the desk to dispose of the tissues? Can they leave the room to blow their nose?
- Can your child not sit near the window with the sun on his/her back? This can cause overheating and then vomiting!
- If your child is unable to attend school – find out what the procedures are for them to catch up on work missed and how to advise the school.

3.2. Issues specific to deaf children (both those with neuro-sensorial and conductive loss)

Your local education authority will put you in touch with a Peripatetic Teacher of the Deaf (or a similar title) at an early stage. Make a good friend of this person; they will be hugely important to your child as they move into primary education.

This person should be in touch with you very soon after your child's hearing loss has been identified. Their role includes helping you identify a suitable nursery and school, acting as a contact point between the school and the education authority, arranging training of school staff, arranging provision of any equipment needed, and so much more. Ask questions of them and keep them **"on your side"**.

Think about the acoustics of the classrooms as well as the communal areas, corridors, halls, etc. Are carpets fitted or do floors have hard coverings? Check all the classrooms; remember

you want your child to stay at the same school for the next 7 years so more than one room needs to be suitable for them.

Think also about the lighting in the rooms. A deaf person needs to be able to see clearly the face of anyone who is speaking to them. Lip reading and other visual clues are essential for understanding spoken and non verbal communication.

Has the school any previous experience with deaf children? Are they **“deaf aware”** or will they need training before your child joins the school? Remember that all staff will need to be **“deaf aware”**, not only the specific class teacher; this includes any lunch staff and volunteers who may help out in school.

Who will support your child in checking hearing aids, using a radio aid and generally ensuring that they can fully access all sessions? There will need to be an appointed person to help your child with this. Trust your instinct; if you have any unease about how your child would be supported, look at other schools.

Ask if there are any children at the school with additional needs (not only deaf kids) and try to speak to the family about their experience of a particular school; this can often provide useful background information to help you make the right choice.

3.3. Absences

Many children with PCD have periods when they cannot attend school. If it is a planned period (e.g. you know they are going to start IV antibiotic treatment) ask the teachers to provide you with some work/games to do in hospital.

It is really important that your child doesn't get behind in their basic literacy and maths skills. Whilst in hospital or off school try to read with them every day. Ask them questions about the story, the pictures and about the characters. To practice their writing skills get them to draw pictures and write a few words on them to send to grandparents and friends. Help them to develop their maths skills by counting the number of beds, nurses and then timing the nurses on how long it takes to give IV's.

When your child returns to school arrange to borrow a friend's books to copy any work that they have missed. Ask the teacher to provide any important work that is missed. If you think your child is not coping with their school work then highlight to the teacher as soon as you can.

Some children find it difficult to go back to school after a few weeks – try to keep in contact with their friends whilst they are absent to make the transition back to school easier.

3.4. How to manage hospital appointments

Making the most out of consultants' time

Your consultant will be your doctor for a long time so it is important that you have a good relationship with them and their team. They will get to know you and your family very well so it is best to be open and honest with them from the start.

Let them get to know what is important to you. (e.g. annual holidays, sporting activities). The more that they get to know you as a person the better they will be able to help you overcome some of the obstacles that living with PCD present.

It is important to think about any issues/problems you have before you go to the appointment.

It is helpful if you can take another adult with you to the appointments so that you can concentrate on talking to the doctor whilst your child is being entertained by somebody else.

4. Development, 4 – 10 years

4.1. School Trips

It is important for children with PCD to experience 'normal' school activities and they should be encouraged to go on school trips.

If it is a day trip make sure that your child has his/her medication with them and that they have sufficient food and fluids for the day. It is important that the teacher in charge is aware of their condition and that they are aware of any limitations your child may have.

If your child is going on a residential trip then arrange to meet the person in charge of the trip.

It is important that your child continues to do their physiotherapy – you may have to train a teacher and give them written permission to do the treatment or go with them as a helper if this can be arranged.

Make sure that they have their medication (including nebulisers and physiotherapy adjuncts) with them. They may find it useful to take some nappy sacks to put tissues in that they have coughed into and then they can seal the bag and dispose of carefully.

4.2. Siblings

Siblings of children with chronic illness such as PCD, often feel that because of the additional care needs of their brother or sister, there is a lack of time for their needs. Everyday family activities may be limited as parents try to juggle the needs of all the children in the family and this can lead to more pressures at home.

It is important to take up offers of help from friends and relatives who may be able to offer some special time for brothers or sisters, or include them in activities which they would otherwise miss out on. It is also useful to have a relative/friend who can look after your child with PCD, including performing their physiotherapy and medical treatment, so that you can spend some time alone with your other children.

Many siblings have found it useful to join a local siblings group. Some of the benefits to siblings include meeting others in a similar position, sharing ideas about coping with difficult situations and having a good time.

4.3. Sleepovers

If your child is going on a sleepover, try to do their physiotherapy before they leave and as soon as they return. It is important that they take their medication with them and that the host is aware of their medical condition in case of an emergency.

4.4. Sport/PE, hobbies and playground games

Physical activity is an important part of development, for muscle strength, bone strength, posture, fitness and lung function. All types of exercise should be encouraged.

Lots of activities can be used including:

- Taking them to play areas and using the swings, slides, climbing frames and roundabouts.
- Teaching them to swim, use a trampoline, skip and play hopscotch.
- PE and sports at school.
- Whatever your child enjoys and gets them out of breath.
- Singing and playing wind instruments are also good for the lungs
- Trampolines are usually popular with children and have proved to be good at helping airway clearance.

5. Early teens, 10-15 years

5.1. Independence

Between the ages of 10 and 15 you will become more independent and will be expected to take charge of routine treatments, managing problems and learning independence. This will then help you to equip yourself with the skills to manage adult life and fully enjoy activities such as employment, going away to college or living with a partner.

This process is known as ‘transition’ in the medical world and should happen over a number of years and in that time, several things need to occur. The paediatric team should outline what the timetable of transition involves, including when the last paediatric clinic or admission will take place. This is usually between the ages of 16-18 years, but varies in different hospitals. Teenagers should be encouraged to participate in their care from an early age - for instance, they start to see the team members in clinic on their own for the first part of the consultation. They should be involved in all decisions about treatment.

The option of seeing an adult respiratory physician at a local district general hospital may appear more attractive than travelling to a specialist PCD hospital, especially if paediatric care has been received locally at a shared care clinic. It is unlikely, however, that the local consultant will have the experience of the full range of current PCD care and treatment. It is also unlikely that the local district general hospital will have a full multidisciplinary team that can provide the specialist nursing, ENT and physiotherapy input that is essential to deliver proper and safe PCD care.

It is important that the teenager understands as much as possible about their condition and recognises the signs when medical intervention (normally in the form of antibiotic treatment) is necessary. It is also important that as you grow more independent and spend less time with your parents/guardians that the people you are spending time with are aware of your condition. If not, then for safety purposes it is recommended that you wear a medical alert bracelet. This allows any medical team to have access to your current medical information when you may not be able to tell them.

5.2. Smoking

It is really important that if you have PCD or have a child with PCD that you don't smoke. As PCD causes mucus clearance to be delayed, the toxic constituents of cigarette smoke may be retained for longer in the lungs of PCD patients, with the risk of increasing damage.

Cigarette smoke also stimulates more mucus production, and the mucus is more sticky – this is why cigarette smokers have a wet “smokers cough”. These effects of cigarette smoke are disastrous in the PCD lung and will increase the risk of infection, damage the lung and cause deterioration in lung function.

It is therefore recommended that if you have a recently diagnosed child with PCD that you give up smoking as soon as possible.

5.3. Career Advice

Talk to your hospital consultant about what type of job you want to do. Ask them for a letter describing your medical fitness to put in with job application.

Some questions you may like to ask a potential employer are: Are there flexible working arrangements? These make life easier in terms of attending Clinic appointments etc. What is the working environment like? Do they have a staff canteen, rest-room or first aid room? Does the company have a Permanent Health Insurance plan?

5.4. Dealing with absences from school

GCSE Examinations and Coursework

Many children with PCD have absences from school. This becomes more difficult to manage when you have started in Senior School and start your GCSE course work.

In order for a pupil to be considered for concessions at GCSE level, they need to have documented their special needs over a number of years. If your child regularly has time off school due to medical reasons it may be worth talking to the school about how concessions may be applied for at an early stage. It is advisable to keep a record of any absences from school, so it can be presented at the request of the examining board.

The joint council for the GCSE (standing agreement number four) advises that 'Examining groups are required to take all reasonable steps to enable candidates with permanent, long-term, or temporary handicaps or indispositions to demonstrate their attainments.' This means that under suitable safeguards, a grade can be awarded if a candidate is absent from an exam for legitimate reasons. Additional time may be permitted (normally up to 25%) for all types of examination.

If necessary, a candidate can receive treatment during a supervised break. Arrangements can also be made (subject to examining board approval) for candidates to take examinations outside their own centre (ie: at home or in hospital).

Disabled candidates may be given additional time to complete course work, or assigned a reduced amount of course work. Again, this is subject to the approval of the examining board. Many of the larger specialist hospitals have hospital schools who will be able to give you further information on this subject.

5.5. Catching up with work missed

If you know you are going to be absent from school ask the teacher to provide you with details of the course work they will be covering. If you have unplanned absences on a regular basis then ask a friend to provide copies of their work/notes as soon as you return to school.

If you are doing a course that requires working on the school site such as D&T or Art – ask the teacher if you are permitted to work at lunchtimes or after school to keep up.

5.6. Physiotherapy - Taking responsibility for your own treatment

Physiotherapy is important to keep your lungs clear and prevent chest infections and lung damage. There are a number of airway clearance techniques to help move the clear the excess mucus from the lungs. As you get older you will become more in charge of your own physiotherapy treatment.

You will still need to do at least two treatment sessions a day but can choose which treatment type works best to suit you. If you find that you don't have time to do physiotherapy then talk to your physiotherapist who can help you find a more adaptable solution.

Your physiotherapist will teach you the **Active Cycle of Breathing Technique (ACBT)**. ACBT is a breathing technique that can be used in conjunction with postural drainage, percussion and vibrations during treatment sessions but can also be used on its own at a time to help clear mucus. It is an ideal technique to learn to make your treatment more independent.

The parts of the ACBT are:

- **Breathing Control:** This is normal gentle breathing using the lower chest, with relaxation of the upper chest and shoulders. It helps you to relax between the deep breathing and huffing.
- **Deep breathing :** These are slow deep breaths in followed by relaxed breaths out. 3 - 4 deep breaths are enough.
- **Huffing :** This is a medium sized breath in, followed by a fast breath out through an open mouth, using the muscles of the chest and stomach to force the breath out. This will move secretions along the airways to a point where you can cough them up.
- **Coughing :** This should follow 2 - 3 huffs OR a deep breath in. Don't cough unless secretions are ready to be cleared.

6. Early Adulthood, 15-20 years

6.1. Relationships and Sex

When you start a new relationship it is difficult to know when to tell your new partner about PCD. Although you may have reasons for not telling your new partner at an early stage about your health condition it is also important that you continue to see your health as priority and don't neglect your physiotherapy routine.

Sometimes having PCD can be difficult as when you get a chest infection you don't have the energy to socialise and you may have to change plans. The changing of plans last minute can put added stress on relationships and it can be difficult for partners to accept that your health might have to take precedence to going out. However most people try not to let PCD get in the way of a 'normal' life and the compromise is often to make the best of the times when you are well, whilst being kind to yourself when you aren't.

As for all people it is important to use barrier contraception to prevent contracting sexually transmitted diseases.

If you are a lady and take oral contraceptives you must remember that short term courses of oral or intravenous antibiotics can affect some oral contraceptive methods by changing how well they are absorbed into your body. During these times you should use a condom throughout the course of antibiotics and for seven days after.

Long term antibiotics used in PCD (e.g azithromycin, flucloxacillin) will not affect your contraceptive if you have been on them for a prolonged period, although you will need additional protection for a short period when starting these antibiotics for the first time.

Antibiotics, such as rifampicin, used to treat TB and related bacteria (called mycobacterium) can affect most methods of contraception, including the Depo-Provera injection and Implanon, so take further advice if this applies to you.

6.2. Use of recreational drugs

It is not advisable for anyone to take recreational drugs as all drugs are potentially dangerous.

This is because most drugs are illegal there's no way to control what goes into them.

Some drugs can cause more long term damage from a physical point of view - like heroin and crystal meth. While other drugs can have a stronger psychological impact and can trigger pre-existing mental conditions (like schizophrenia). There's no way of you knowing that beforehand.

6.3. Alcohol

It is important that if you follow the government recommended limits for drinking.

Also check that the medication you take is not affected by alcohol.

If you are going on a night out try and do your physiotherapy session before you go.

6.4. Stress/Exam time

Don't forget that stress reduces your immune levels.

It is really important that you manage your stress levels effectively in order that you don't become unwell – especially during exam periods.

Try and schedule some relaxation time into your life – getting some exercise is particularly beneficial as it helps clear your chest in an enjoyable way and releases endorphins which combat stress and make you feel good.

Make sure that you eat healthily and don't forget to schedule in your physiotherapy sessions.

6.5. University Choices

Either choose a university where you can easily walk to the campus from the halls or choose one where the halls are on the campus so walking to lectures is not far.

When applying for student accommodation, they usually ask if you have any special requirements for your room, and if you specify that you have a physiotherapy bed, they will make special allowances for you to get a slightly bigger room to accommodate this.

You can also specify if there is no lift in the halls, that you need a ground floor room so when you get ill, you never have to go too far.

When it comes to hospitals, it is helpful to have a consultant or a nurse as a point of call if you get sick and they can give you help to travel home to see your regular consultant.

This is just helpful in keeping your treatment consistent and more convenient rather than having to start afresh in a new clinic where they may not be familiar with the condition.

6.6. Learning that your limits are different to others but still pushing the boundaries

It is important to recognise that your energy levels are different from others, and to make allowances for this without necessarily spotlighting that you have a problem.

This can be managed by planning activities carefully so that there is rest/physio time in between.

Energy levels and degrees of wellness can be unpredictable, so plans may need to be changed at short notice. Whilst this may not be very palatable, it may be better to do this than to carry on anyway and make yourself worse so that you end up missing even more.

Being positive and living as normal and full a life as possible needs to be balanced against recognising your health needs and looking after yourself.

This balance constantly changes and needs to be carefully monitored to ensure that you get the very best out of life

7. Starting a Career, 20-25 years

7.1. Starting a Career, 20-25 years

When you start a career you will invest lots of time in your new job and won't have much time for other things.

As always with PCD it is important that your health remains a priority and that you find time for your physiotherapy sessions.

It is important that you share with your employer that you have PCD (give them a leaflet) and explain to them how PCD affects you on a day to day basis. This will help you become more empowered at work to manage your own condition.

Try and work from home if you have the option when bugs are flying around. Work by a window and try and avoid big air conditioned environments. Always keep a box of tissues on your desk so that you can blow your nose regularly.

Eat a varied diet to try and keep energy levels high. Try and keep up regular exercise (running around the block and getting a dose of fresh air is invaluable).

Manage your workload proactively to help you avoid stress. Basically, if you get run down, a chest infection will soon follow.

If you are finding it difficult to keep on top of your physiotherapy talk to your physiotherapists about alternative physiotherapy strategies that fit in better with your new lifestyle.

8. Raising a family, 30+ years

8.1. Fertility issues

Infertility is the inability to conceive after a year of trying and this affects 1 in 7 couples. At least 50 % of people affected by PCD can get pregnant without difficulty the remainder may need some sort of assistance.

There are several reasons for not conceiving and they can be divided for simplicity into 4 groups;

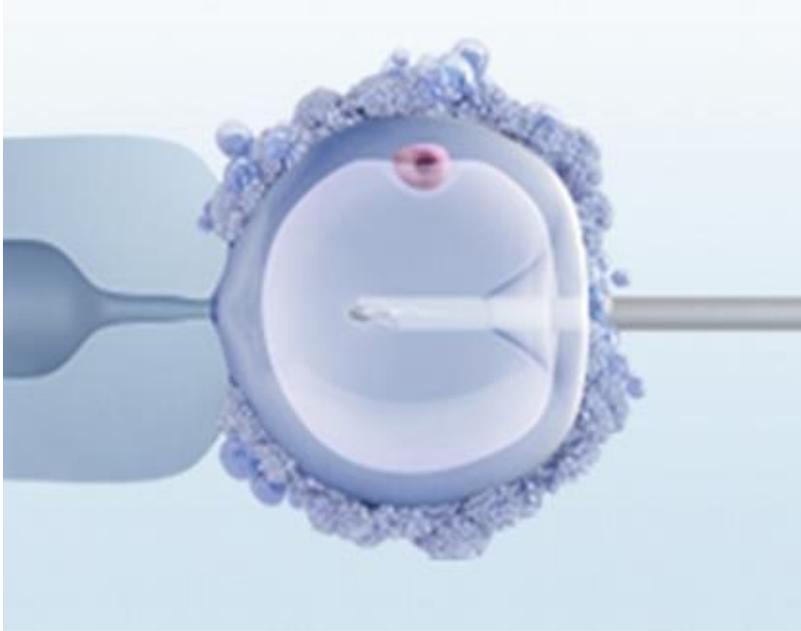
- **Male Factor**
- **Tubal Disease**
- **Anovulation**
- **Unexplained Fertility Issues**

Each of these conditions is explained on the next few pages.

8.2. Male Factor

This is when the sperm is poor quality. This affects 25% of couples. Either the count, the percentage swimming well (motility) or the number that look normal (morphology) are low. Sometimes all three are low. Occasionally a man may have no sperm in the ejaculate, this is called azoospermia.

In men with PCD the sperm may have poor motility and this reduces the chance of conceiving naturally. This can be overcome with in-vitro fertilization (IVF) and intracytoplasmic sperm injection (ICSI). The female partner is given drugs to make her produce lots of eggs. These are collected by a simple surgical procedure. A single good quality moving sperm is injected into each egg (ICSI) allowing fertilization to create embryos that can be replaced in the womb.



A drawing of a single sperm being injected into an egg (ICSI).

In azoospermia a surgical procedure can be performed and sperm can be successfully extracted from the testis in >50 %. Alternatively donor sperm may be used.

8.3. Tubal Disease

The sperm and egg meet in the fallopian tube and fertilization occurs then the fertilized egg (embryo) moves back into the womb. If the tubes are blocked or absent then this cannot occur. This is a cause of infertility in 20%.

The most common cause of tubal damage is pelvic inflammatory disease (PID) caused by chlamydia or gonorrhoea following unprotected intercourse. Other causes of tubal damage include surgery appendectomy, tubal ectopic, sterilization and ovarian surgery or infection/inflammation in the abdomen such as peritonitis. Rarely tubal surgery can be used to repair damaged tubes.



Damaged fallopian tube with adhesions (scar tissue) secondary to infection or surgery

In females with PCD the cilia in the tube may not move effectively and there may be tubal damage secondary to previous ectopic pregnancies. If the tubes are not functioning normally then IVF is recommended.

8.4. Anovulation

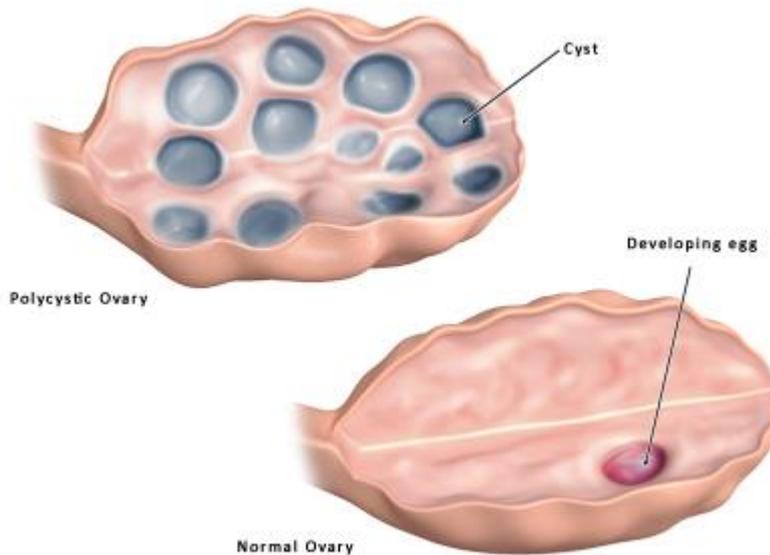
Anovulation

This is the inability to release an egg every month and is a cause of infertility in 25%. Commonly women who do not ovulate have irregular or infrequent menstrual cycles. Polycystic ovary syndrome is the commonest cause of anovulation.

Other causes include thyroid dysfunction, anorexia and obesity, disorders of the pituitary and hypothalamus (structures in the brain that control hormone levels). Lastly it can be caused by ovarian failure after chemotherapy, radiotherapy or surgery to the ovaries or chronic illness.

In women with PCD, like other chronic illnesses when you are unwell you may lose weight and the cycles may become irregular due to lack of ovulation. More rarely medication or serious illness may cause irreversible damage to the ovaries.

Restoring weight to normal can allow menstrual cycle to become regular again. Medication like clomiphene or treatments like intrauterine insemination (IUI) can be given to restore ovulation. If there is ovarian failure then egg donation is the only fertility option.



8.5. Unexplained Infertility

Unexplained infertility

This is when the sperm is normal, the tubes are patent and there is normal ovulation. This condition affects 25% of couples. If you are young it is sensible to continue trying naturally. If there is no success after 3 years IVF is the best option.

For anyone trying to get pregnant it is important to be healthy to have a normal weight (BMI > 18 < 30), to eat a healthy diet and exercise regularly, to avoid excessive alcohol, smoking and recreational drugs and for the women to take folic acid. The chances of getting pregnant decrease dramatically with age especially beyond the age of 39.

Access to fertility services can be arranged through your GP. The GP can organize basic test such as hormone blood levels and a semen analysis. If these are abnormal you should be referred to a fertility specialist.

Risks of IVF/ICSI must be considered. The main risk is multiple pregnancy (twins or triplets) if we put back more than one embryo. In women with PCD we would recommend avoiding multiple pregnancy by replacing only one embryo as all the risks of pregnancy are greater with twins. In particular there is a chance of decreased respiratory function as the increased size of the pregnancy compresses the lungs. Twins are also more likely to be born prematurely and could end up with long-term health problems.

An egg collection requires an anaesthetic or deep sedation so it is advisable for the respiratory function to be optimal in women with PCD when they start IVF/ICSI treatment

8.6. What is an Ectopic Pregnancy?

It happens when a woman's ovum (egg), when it has been fertilised by a man's sperm, remains "stuck" in the tube instead of moving down her fallopian tube into the womb to implant there. The fallopian tubes are narrow hollow muscular tubes lined with cells that are mucus-secreting and other cells that have cilia - tiny hair like projections which beat gently to propel these secretions towards the uterus.

The egg and sperm meet and fertilisation occurs in the tube. If the tube is damaged by inflammation or infection the pregnancy can implant in the tube to form an ectopic pregnancy.



The risk for ectopic pregnancy is greater in women with PCD compared to other women as the cilia in the tubes are less effective at propelling the embryo towards the uterus.

What are the Symptoms?

Mostly women present between the 4th and 10th week of pregnancy with any of the following symptoms:

- Abdominal pain
- Shoulder-tip pain
- Abnormal bleeding
- Missed or late period
- Pain when moving the bowels or on going to the toilet.
- Collapse

8.7. How is it managed?

If the woman is well and not in severe pain, she may be investigated with a blood hormone test done repeatedly over two to three days to establish whether there is an ectopic or not.

If an early diagnosis can be achieved before rupture of the tube then surgery or treatment with drugs can be offered. This allows a speedier recovery and may increase the woman's chance of future fertility.

The surgery is done laparoscopically (keyhole surgery) and the tube is either removed (salpingectomy) or the ectopic is removed leaving the tube (salpingostomy) .

Alternatively, the drug methotrexate, that destroys rapidly dividing cells such as pregnancy cells, can be used.

If rupture of the tube is suspected abdominal surgery normally laparoscopy (keyhole) surgery or occasionally open surgery is performed to remove the ectopic.

8. 8. Future chances of pregnancy?

The overall chances of a repeat ectopic are between 7–10%. If the tubes are both damaged IVF may be required.

8.9. Help with looking after your children

There are a number of people that can help out if you need extra support. These social workers work closely with the GP and also others such as Speech Therapists, School and District Nurses in all aspects of looking after small children. You can access your social visitor through your GP's surgery.

8.10. Get your partner to be more involved

Some people find this support helpful, where as others still prefer to keep their treatments more private.

It can also be difficult for partners to know how involved they should become in treatments, and some might prefer to stay very much on the periphery.

Others might want to offer as much help as possible, or may start to worry about whether you are keeping up with treatments effectively.

8.11. Stress Incontinence?

Stress incontinence is when urine leaks because there is a sudden extra pressure ('stress') on the bladder. (The diagram shows how the pelvic floor muscles support the bladder and nearby structures.)

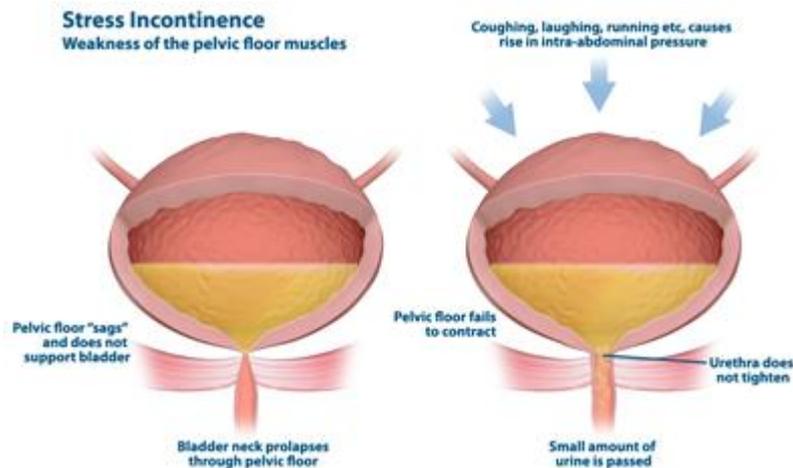
Stress incontinence develops because the pelvic floor muscles are weakened. Small amounts of urine may leak, but sometimes it can be quite a lot and can cause embarrassment.

Urine tends to leak most when you cough, laugh, or when you exercise (like when you jump or run). In these situations there is sudden extra pressure within the abdomen and on the bladder.

How common is Stress Incontinence?

Stress incontinence is the most common form of urinary incontinence in women.

It becomes more common in advanced age as many as 1 in 5 women over the age of 40 have some degree of stress incontinence.



8.12. What causes stress incontinence?

Causes

Most cases of stress incontinence are due to weakened pelvic floor muscles.

The commonest reason for the pelvic floor muscles to become weakened in women is childbirth in men it is prostate surgery. The pelvic floor muscles are a group of muscles that wrap around the underside of the bladder and rectum.

Stress incontinence is also more common with increasing age as the muscles become weaker, particularly in women after the menopause. Stress incontinence is also more common in those who are obese or those with a chronic cough.

What are the treatment options?

The usual first treatment is to strengthen the pelvic floor muscles. About 6 in 10 cases of stress incontinence can be cured or much improved with this treatment.

Strengthening the pelvic floor muscles - 'pelvic floor exercises'

It is important that you exercise the correct muscles. Your doctor may refer you to a continence advisor or physiotherapist for advice on the exercises.

Surgery

Various surgical operations are used to treat stress incontinence. They tend only to be used when the pelvic floor muscle exercises have not helped.

The operations aim to 'tighten' or support the muscles and structures below the bladder. Surgery is often successful.

9. General PCD Information

ENT Issues

9.1. ENT Issues

Problems with the ears and nose often lead to a diagnosis of PCD. Otitis Media with Effusion (OME) or Glue Ear is almost universal in PCD. This is due to mucus collecting in the Eustachian tube and not cleared as the cilia there are not working properly.

This means air does not get to the middle ear from the nose, fluid taking its place. This causes hearing loss as the sound has to travel through the liquid. In younger children there may be ear infections. Technically, the problem is just lack of air.

The normal management of a child with hearing loss due to glue ear is insertion of grommets (ventilation tubes). When grommets are put in the ear is no longer waterproof, so care must be taken when hair washing and swimming.

The tube can get infected and ear discharge may result. PCD patients with grommets tend to have persistent ear discharge.

The discharge mitigates any benefits from the grommets and makes wearing hearing aids not possible. For this reason normally grommets are not inserted in children with PCD except in special situations such as if the ear drum looks in danger of collapsing.

9.2. What can be done?

- For glue ear the Otovent works by getting the child to blow down one nostril into a balloon to inflate it, thus “popping” their ears. It is not something that has proved useful in long term use, but popping the ears can be helpful in improving hearing.
- Hearing Aids are recommended if both ears have significant hearing loss and there are signs that the loss is affecting the child’s general, school or speech and language development. If the loss is in one ear, then the other ear will be able to cope.
- With younger children it is important to help their speech development by talking to them from the front, at slightly raised level and sitting close so they have every chance to hear.

A study of all the children going through the clinic over time has shown that at 3, only 20% of the children have normal hearing, by 6, this is around 60% and by early teens, nearly all are normal.

9.3. PCD and the Nose

Mucus in the nose is there to trap things coming in, and drains down the throat. The sinuses in cheeks and forehead drain into the nose. In PCD the cilia do not move the mucus properly so it can get infected. If the nose is blocked it can lead to the sinuses getting infected. Sinuses develop as children grow which is why it is not a problem with small children.

What can be done?

Saline sprays can be helpful in clearing mucus. E.g. Sterimar. It can be used in the morning to help clear mucus that has gathered in the night, and may last half a day. Steroid nasal sprays, e.g. Flixonase are supposed to reduce nasal discharge; its effectiveness needs to be studied. In a case of recurrent sinusitis, the infected mucus in the sinuses can be removed surgically.

Additional points about ENT issues:

- Scuba diving deeper than 5 metres is not recommended.
- The amount of steroid in the nasal spray is very small and is safe in children. The child should have regular height and weight checks anyway.
- Ear wax is not normally a cause of hearing loss and can be kept down by using the drops for wax such as Sodium Bicarbonate found in the chemist shop.

9.4. Reflux Problems

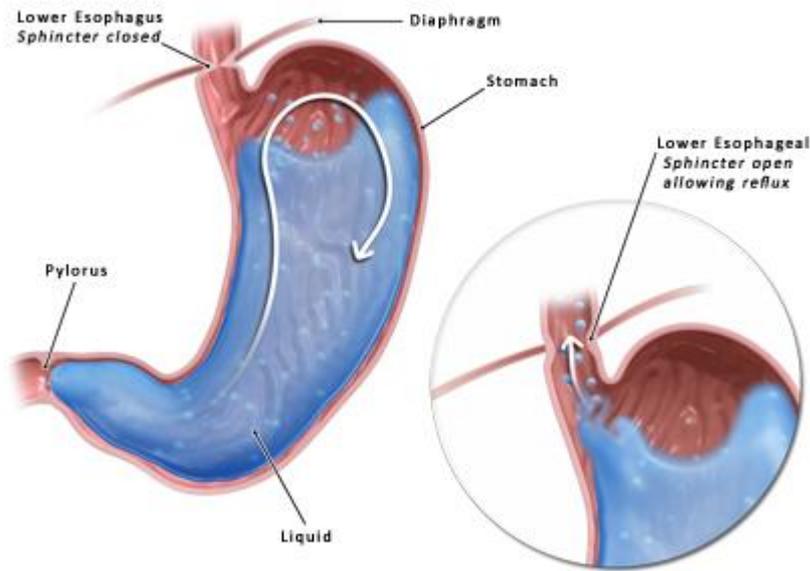
Gastro-oesophageal reflux [GOR] is a common companion for any patient with a chronic respiratory condition and may even be the cause of some chronic lung problems.

In PCD it may co-exist and it may exacerbate symptoms that already exist.

Anybody with a chronic cough is likely to have an increased risk of reflux due to raised intra-abdominal pressures during bouts of coughing. If patients appear not to be responding to maximal therapy to treat the lung consequences of PCD then GOR should always be looked for and treated. Reflux and aspiration may lead to infection and inflammation in the lungs than can greatly enhance the progression of the lung disease.

Treatment is usually medical with a combination of antacids and pro-kinetic agents. Neutralising gastric acid will reduce both the pain [heartburn] and inflammatory responses in the lungs if aspiration does occur. Prokinetics are agents that increase the gut motility [peristalsis] to enhance stomach emptying. Pooling of liquid and food in the stomach greatly increases the risk that stomach contents can wash up and down in the oesophagus and therefore lead to aspiration events.

Dual therapy is more common in young children, but can be useful in all ages. Occasionally surgical intervention will be required to prevent aspiration as a lung protective measure.



9.5. Baseline

It is helpful when you are attending a new hospital or doctor that you have an understanding of your own health.

Date	FEV1	FVC	Height	Weight	Sputum volume and colour	How do you feel?
00/00/00	From spirometry test	From spirometry test			Is it normal colour, yellow or green??	Good, not as good as normal, lousy etc.

Above is an example of a Baseline Table

Normal body temperatures can vary and are influenced by factors such as exercise, eating, sleeping and the time of the day - the lowest temperature is usually recorded at around 3am, the highest at 6pm. The average normal body temperature taken in the mouth is 37°C (98.6°F), but anywhere between 36.5°C and 37.2°C (97.7°F and 99°F) may be normal.

Normal armpit temperatures are 0.2°C to 0.3°C lower than this. It is recommended that if you have a long term condition like PCD that you know what your normal temperature is so that if you are unwell you know how raised it is and can act accordingly.

A temperature of 38°C (100.4°F) or above is usually considered to be a significant fever - you should measure it again after two to three hours.

9.6. Lung Function Tests

When you arrive at the hospital, the tester will measure your weight and height. If they can't measure your height (if you are in a wheelchair, for example), they will measure your arm span - this gives them a guide to your height.

For the tests, you will need to blow into a machine. With some machines, you blow into a tube; for others, you put a mouthpiece in your mouth, like a snorkel, and then blow. You will usually have to do this several times - but the tester will not rush you.

If you are breathing through your nose, and this is affecting your breathing test, you may be asked to wear a nose-clip.

If you wear dentures and they're loose, then you might need to take them out when you blow into the machine.

Occasionally, the tester might ask you to sit in a booth, which looks a bit like a small see-through shower cubicle. The door will be closed and the test will be done while you sit inside. This will help your tester get more detailed results.

Spirometry

This measures your FEV1 - which is how much air you can expel from your lungs in the first second of breathing out. It also measures your FVC - which is how much air you can get into your lungs and how long it takes for you to empty your lungs.

9.7. Blood gas tests

These are tests to see how well your lungs are getting oxygen into your blood and taking carbon dioxide (a waste gas) out.

The simplest test measures how much oxygen is in your blood using an oximeter (pictured) - a little gadget that clips on your fingertip or earlobe. Sometimes it is attached to a monitor that you wear on your wrist, like a watch.

The oximeter doesn't take blood. It shines a light through your fingertip or earlobe, and measures how red your blood is.

NB: If you wear nail varnish, this might block the light and affect the reading, so you'll be asked to take it off (on one finger only). False and acrylic nails can also cause a problem and are not so easily removed in an emergency situation.

9.8. Exercise tests

To check your breathing, the tester will ask you to do some exercise and take measurements while you're exercising, and afterwards.

This might involve:

- Walking at your own pace for six minutes, taking as many rests as you need.
- Doing a test called a 'shuttle walking test'. This involves walking between two points set 10 metres (about 30 feet) apart. This is timed with the times gradually get faster, until you can't keep up.
- Walking on a treadmill, while the tester monitors your heart and lungs.

- Doing a test on an exercise bike. This is done occasionally if the doctors need more detailed information about your breathing. Usually you will be asked to breathe through a mouthpiece while you cycle. The amount of oxygen you breathe in and the carbon dioxide you breathe out are measured, as well as your breathing rate, pulse and sometimes your blood gases

9.9. Treatment reversibility tests

Bronchodilators are designed to expand the bronchial tubes that carry air in and out of your lungs.

After you breathe in the medicine, you wait for about 20 minutes. Then you do the spirometry test again to see if your results have changed.

If you have asthma, your spirometry test improves after taking the bronchodilator.

These tests help to:

- Decide whether you may have asthma.
- Check how your lungs react to different medicines.
- Decide which treatment suits you best.

9.10. CT Scans and X-rays

Imaging in PCD is a useful adjunct to help the clinician assess a patient's status, but regular imaging is not necessary in the stable patient.

Chest x-rays can help confirm situs anomalies, and ultrasound should then be used to determine abdominal arrangement of organs where there is a suspicion that organs are arranged in an ambiguous arrangement [heterotaxy]. Chest x-rays are also useful if significant infections or pneumonias are suspected, and may be extremely useful in the diagnosis of allergic bronchopulmonary aspergillosis (ABPA).

CT scans can help the clinician with treatment options, but also be a useful way of reviewing the condition with patients who may not either understand the potential impact of PCD on their lungs or who may not be very engaged in their treatment.

In older children and adults where rhinosinusitis is a problem sinus scans may be required and can help the ENT surgeon determine when surgical intervention is required

„CT scan's are seldom needed as part of the diagnostic work up and their use is usually reserved for staging the lung disease.“

9.11. Are there any side-effects when performing lung function tests?

There should not be any side-effects from carrying out these tests.

As you are usually asked to perform the breathing exercises a few times, you may feel a little tired after the tests have been completed - this is quite normal and you should not worry.

The tired feeling will quickly disappear. However, if you have any chest pain or other problems during the tests, please tell a nurse.

Test results

Your lung function test results and a report, if needed, will usually be available almost immediately after the tests have been completed.

You can either take these results with you to the clinic or ward or they can be sent for you.

Do's and don'ts

- Do wear loose clothing which does not restrict your chest and abdominal movement.
- Do not smoke for at least 24 hours before the tests.
- Do not perform vigorous exercise for at least 30 minutes before the tests.
- Do not eat a large meal during the two hours before the tests. Please do not drink alcohol during the four hours before the tests.

10. Managing PCD in the home

10.1. Home administration of IVs

Your PCD Specialist nurse or consultant may suggest this to provide you with more flexibility.

The decision about whether home IV treatment is appropriate will be made each time the patient needs IVs.

Deciding the best place to receive IV antibiotic treatment is influenced by the patient's condition, availability of family and professional support, geographical location and personal needs and preferences.

If you are allowed to do home IV's the first dose is always given in hospital.

Learning how to do home IV therapy

A suitably qualified nurse or doctor will teach the patient or carer how to draw up drugs, safely administer IV antibiotics and look after their lines/portacaths.

Many patients will have had IV courses before and already be familiar with the procedure.

Teaching programmes are tailored for individuals at their own pace.

Reassessment may be necessary, particularly if IV courses are infrequent.

„It is essential that the patient or carer is both competent and confident before home IV therapy is considered.“

10.2. Practical considerations

You will need a cool, clean, dry shelf or cupboard out of reach of children and pets to store your drugs and equipment. Some drugs may need to be stored in a fridge – your pharmacist will advise you of this. Simple, user-friendly instructions should be issued to back up your teaching programme by your nurse or doctor. This should contain a section on dealing with problems.

Arrangements will be made for enough equipment to be issued to complete the course. A 'sharps bin' is required for safe disposal of needles, syringes and other sharp items and this must be sealed on completion. Information on disposal and/or collections of the box and any other items of equipment will be given by your nurse or doctor.

Monitoring and follow-up of home IV courses may involve home visits, phone calls or clinic appointments. Some drugs require a blood test to check dosage is correct; again, you will be advised of this.

You will need to be available for any follow-up care necessary to ensure treatment is both safe and effective. Reactions to drugs can occur, which is why the first one or two doses of any new drug are given in hospital under medical supervision.

10.3. What should I do if I have a reaction to the medication?

Should you experience any of the following problems you should stop the course and seek immediate advice from the 24-hour contact number provided by the hospital initiating the IV therapy:

- **rashes**
- **nausea**
- **severe diarrhoea**
- **any other unusual symptoms**
- **temperature**
- **vomiting**
- **swellings**

Severe reactions are very rare but must be treated as an emergency by calling an ambulance and going to the emergency department. This will include:

- **sudden collapse or illness**
- **severe difficulty breathing**
- **swelling around the neck**

Your hospital will let you know whether you need to have special training and / or emergency medication (anaphylaxis kit) to deal with a severe allergic reaction.

It may be possible to continue with employment or education whilst having a course of IV antibiotics and this can be discussed with the nurse or doctor initiating the course of therapy.

However physiotherapy, good nutrition and adequate rest are all essential towards achieving the best outcome from an IV antibiotic course.

Similarly, drug times may be adjusted to accommodate these activities. In some cases drug administration is possible at work or school.

It is advisable to inform your employer or school that an IV line is in place and an antibiotic course in progress as their permission may be required for health and safety reasons. In every case, good communication between all those involved is essential.

„Remember, giving IVs at home is an option. Hospital care is still available if you need it.“

10.4. Infection Control

Infection control when attending clinic appointments and staying in hospital is important to prevent cross infection between patients.

When you are in hospital you will be segregated from the other patients with any form of chronic suppurative lung disease (that is any lung disease that produces mucus such as PCD and Cystic Fibrosis). Certain bugs, such as pseudomonas, are potentially much more likely to cause progression of lung disease and every effort should be made to prevent cross infection.

This is achieved by having high levels of personal hygiene, separation of patients on wards and in clinic and avoiding sharing rooms for physiotherapy.

You should ensure that you clean and sterilise all nebulizer equipment as per the manufacturer's instructions. It is also important that you do not share any equipment with your brothers and sisters. If you are in hospital don't forget to wash and sterilise equipment – if this is a problem as the nurses to help.

In general it is a good idea to do the following to avoid infections:

- When you cough cover your mouth with your hand or a tissue. Dispose of the tissue quickly. Politely ask that other people do the same.
- Do not share drinking bottles, cups or glasses.
- Wash your hands and home and work surfaces regularly.

10.5. Vaccinations

Most is known about this problem in patients with cystic fibrosis, who are particularly vulnerable to both cross infection and to worsening symptoms when infected with certain organisms. Of particular concern is transmission with pseudomonas, although much less is known about the impact of this organism in PCD or other forms of bronchiectasis.

As a result in-patients with any form of chronic suppurative lung disease are segregated to prevent this happening. This is achieved by personal hygiene regimens, separation of patients on wards and in clinic and avoiding sharing rooms for physiotherapy. Cleaning all nebulizer equipment and not allowing siblings to share equipment is essential.

Preventing infection is also possible with vaccination and all PCD patients should follow guidance for both annual vaccination programmes, and more long term protection for other bacterial forms of pneumonia. The following guide is typical, but can change depending on emergence of new organisms and development of vaccines. Your medical team will be able to update you on what is current.

Influenza.

- Seasonal flu vaccines are developed every year in response to the virus that is most prevalent. It does not protect against all forms of influenza, and is only protective against the most likely virus to cause an epidemic that season.
- Swine flu vaccines have been developed in a very similar way to the seasonal flu vaccine in response to the worldwide spread [pandemic] of the H1N1 virus. This is a newly identified virus and less is known about how it will behave in any given season,

or what will happen when it is prevalent with another form of epidemic flu. It is safe and well tolerated and all PCD patients should be offered this protection.

Pneumonia.

- H. influenzae type B is now part of routine childhood vaccination programmes.
- Streptococcus pneumoniae is a common infection in PCD and all patients are advised to have the pneumococcal vaccine. This vaccine does not need to be given annually, and covers many but not all serotypes of the organism. Therefore, pneumococcal infections may still occur and should be treated aggressively.

10.6. Allergies and Irritants

Allergies + irritants

Patients with PCD are no less likely to be susceptible to allergies than any other individual, but occasionally atopy may co-exist and complicate or worsen the signs and symptoms they suffer.

Such patients might benefit from typical asthma therapies in conjunction with their other treatments for PCD.

Patients with chronic lung diseases can, however, be susceptible to a specific allergy to a fungal organism called aspergillus fumigates. This condition, called Allergic Bronchopulmonary Aspergillosis or ABPA, may present with shortness of breath, wheeze and feelings of chest tightness.

There may be a fall in lung function that is difficult to reverse with the usual treatments. All these symptoms are not typical of PCD alone, and so ABPA should be looked for using both sputum and blood tests to diagnose it.

Treatment involves a combination of anti-fungal agents and a limited course of oral steroids.

It is possible to reverse the features with treatment, but like many allergic conditions there may be exacerbations that need further treatment courses.

10.7. Dealing with GP's

It is important that you have a good relationship with them and that they are aware of your current medical condition.

You will need to become proactive in your own or your child's care, e.g. you may need to explain why you need another course of antibiotics and a minimum of a two week course. You will also need to explain what bugs you have recently grown and what they were treated with.

You will also need to ask for a sputum test so that antibiotics are targeted.

Give them PCD leaflets and recommend that they contact your hospital consultant for advice if required.

„Make sure that the GP feels that it's ok 'not to know all the answers' - the relationship then becomes 2 way.”

11. Healthy Lifestyle

11.1. Healthy Diet

Food is the fuel your body needs for everything you do, including breathing.

Your body uses food for energy as a part of a process called 'metabolism'. This is when food and oxygen are changed into energy and carbon dioxide.

Eating healthily also helps the body fight infections. Chest infections are illnesses that can be dangerous for people with PCD.

You can reduce your risk of infection by eating well.

11.2. Five essential nutrients

There are five nutrients that are essential for health and life.

1. *Proteins*

- Found in milk, eggs, meat, poultry, fish, nuts, pulses
- The body uses proteins to repair damaged tissue
- Remember to trim the fat off meat - too much fat is not good for you

2. *Carbohydrates*

- Found in bread, cereal, rice, pasta, fruit and potatoes
- They are the easiest form of energy to digest
- If they are high in fibre, they help with your bowel function and lower your cholesterol

3. *Fats*

- Found in butter, margarine, olive oil, vegetable oil, nuts
- They are mainly a source of energy and vitamins A D E and K

4. *Minerals*

- Found in liver, eggs, meat, fish and dried fruits such as raisins
- Milk and cheese are rich in calcium (and vitamin D)
- Calcium and vitamin D help with healthy bones and teeth. Iron is important for your blood
- Calcium is especially important for people who are on high amounts of steroid treatment

5. *Vitamins*

- A – improves eyesight and helps fight infections - it's found in butter and many dark green vegetables
- B – necessary for normal nerve function, good digestion, good appetite and healthy skin - found in wholegrain breads and cereals, meat, fish and dairy products
- C – good for fighting infection. The best sources are citrus fruits (oranges, grapefruit), strawberries and broccoli
- D – helps with healthy bones and teeth. Found in milk and cheese
- E – found in whole grain cereals
- K – regulates blood clotting - found in cauliflower, kale, spinach, Brussels sprouts, soya beans, meat and green tea

11.3. Your ideal weight

If you are overweight, your heart and lungs have to work harder to supply oxygen to your body.

Being underweight is not good either. You can feel weak and tired, and you may be more likely to develop a chest infection. It's important for you to eat enough calories to prevent muscle wasting or weakening.

Talk to your nurse or GP to find out your ideal weight.

11.4. Loss of appetite

If you feel unwell, if you're producing lots of mucus, or if you're breathless - you may lose your appetite.

Tips:

- Take your medicine with food unless advised otherwise. This prevents an upset tummy
- Vary the colours and textures of your food
- Use colourful garnishes to brighten food and make it more appealing
- Take regular exercise
- Take advantage of meals available at local community centres, clubs and churches
- Try high-energy drinks - they take less energy to digest
- Brush your teeth before meals

11.5. Loss of muscle tone

If you are unwell you may take less exercise. Your muscles may become weaker.

Try to eat a well balanced diet.

- Try to eat more high-protein foods - meat, fish, poultry and dairy products
- Add grated cheese to vegetables, soups and casseroles

- Use double-strength milk instead of water for cereals and milk puddings (double strength milk is one cup of whole milk with 1/3 cup of non-fat dry milk powder added)
- Add hard-boiled eggs to tuna and chicken for sandwiches/salads
- Add non-fat dried milk to casseroles, custards and puddings

11.6. Keep drinking

It's important to keep the secretions in your lungs thin and easy to cough up.

If you don't drink enough, your secretions will be thick and sticky and will increase your chances of an infection.

Drinking enough liquids also helps with digesting food and avoiding constipation.

Water is your best option.

- Drink at least 2.5 litres of liquid a day. (juices, milk shakes, water)
- When you have an infection or fever or during the hot weather, drink more.

"Water makes up about two-thirds of our body weight. And it's important for this to be maintained because most of the chemical reactions that happen in our cells need water."

11.7. Probiotics

Antibiotics may be effective at eliminating pathogens in the body but they also disrupt the balance of desirable and undesirable bacteria in the gut.

This can leave the digestive tract available for pathogenic bacteria to take up residence making it harder for the friendly bacteria to re-colonise to beneficial levels.

The symptoms of an imbalance of gut flora are: bloating, constipation, diarrhoea, indigestion, IBS and food intolerances. Friendly bacteria are found in cultured foods such as live yoghurt, cottage cheese, miso, sauerkraut, tofu, tamari and kefir.

However, these may not be enough, and a friendly bacteria supplement known as a probiotic may be beneficial. Friendly bacteria are crucial to our health and carry out a range of functions including:

- manufacture of B vitamins
- play an important part in our immune system by increasing numbers of immune cells
- increase the absorption of minerals
- produce lactase to digest milk
- enhance bowel function by promoting efficient passage of waste
- help control cholesterol levels
- act as anti-carcinogens
- produce antibiotics to control levels of bad bacteria, viruses and fungi.
- alter the acidity of the gut to make the environment undesirable for pathogens

- break down hormones and build hormones for re-use
- contribute to optimal metabolism
- help remove toxins

11.8. Exercise

Exercise is an essential part of a healthy lifestyle. Not only does regular exercise help to maintain overall fitness and manage weight loss, it can lower the risk of many life-threatening conditions.

People who exercise regularly are likely to live longer and enjoy a better quality of life. In fact, studies have shown that being physically unfit is just as dangerous as smoking in terms of lowering life expectancy.

Regular exercise also improves mental and emotional health. The chemicals and hormones that are released in the brain through exercise can help deal with stress and promote happiness.

For children it is important that they take regular exercise to develop muscle and bone strength, help posture, fitness and lung function. All types of exercise should be encouraged. Trampolines are particularly effective in helping to clear mucus.

All these health benefits are the reason why many people are encouraged to exercise. However, many people think exercise is too expensive or simply takes up too much time. In reality, as little as half an hour of moderate activity every day, such as brisk walking, can be enough to improve health and fitness. There are many different ways to exercise, and it is possible to find something to suit any kind of lifestyle.

Any type of aerobic exercise that encourages you to take deep breaths is good, such as football, rugby, swimming and running. It may also be helpful to try other activities that concentrate on breathing techniques and balance such as tai chi and yoga. If you are unable to get out then get a Nintendo Wii – they are great exercise for all the family.

“Exercise can also be used as part of your mucus clearance routine and will help to improve lung function and fitness.”

12. Going on Holiday

12.1. Insurance

It is important that you fill in the medical form in detail so that there is no risk of the company not reimbursing a potential claim. They also need to check that the policy does not exclude pre-existing illness.

Everyone needs a European Health Insurance Card (EHIC) in order to receive free emergency care in EU countries.

Some handy hints on getting insurance:

- You must telephone the medical pre screening line to ensure that you are covered – failure to do this might result in your insurance being invalid when trying to make a claim.
- They may ask you what the condition is and you will be told that they don't have P.C.D. on their systems! Don't panic – just tell them that another name for it is Kartagener's Syndrome and they will then be able to ask you relevant questions. If you tell them that the treatment is similar to Cystic Fibrosis you will struggle to get cover at a reasonable price.
- Don't forget to ring the medical screening line again before you travel if circumstances have changed since the time you first got cover
- It is unlikely if you take the correct medication with you that you will have an emergency situation because of your PCD – however, if you end up in hospital because of an accident such as breaking your arm you may not be covered by the insurance if you haven't told them about PCD.

12.2. Medication

Ensure that you take all your medication (including for an extra week) plus a suitable standby course of oral antibiotics.

Remember to keep some medication in hand luggage in case of delays in airports. rhDNase will need to be carried in a cool bag.

Don't forget that you will need to use sunblock if taking ciprofloxacin, doxycycline or voriconazole (and for 4 weeks after course has finished).

Nebulisers

In Europe (except for Cyprus, Gibraltar & Malta), the voltage for the nebuliser is the same as the UK (220v) and a standard travel plug adapter is all that is needed.

If travelling to USA, South America, Caribbean, Cyprus, Gibraltar & Malta, you will need a 110v nebuliser e.g. Port-a-Neb. A plug adapter is not enough.

Discuss this with your Physiotherapy Department well in advance of the holiday and they may be able to lend you one. Alternatively they are hireable in the USA from medical equipment companies.

Medical Letters

It is advisable to take a copy of your latest clinic letter with you to help explain why you are carrying medication and if you end up in an emergency situation.

If you don't have a copy of the latest letter then contact your Consultant's secretary and they can arrange one for you.

It is important that your consultant is happy that you are well enough to travel.

Fitness to Fly

A fitness to fly test may be suggested for patients with:

- History of needing oxygen during chest infections.
- Resting oxygen saturation < 94%.
- Spirometry FEV1 < 50% predicted.
- If on home oxygen, it will definitely be needed on the airplane, but a test can be used to determine flow rate necessary on the plane. Your consultant can arrange this test if required

13. General Physiotherapy Information

General Advice

13.1. General Advice

Mucus traps dust and bacteria in the lungs which is then swept out by normal cilia. However, in PCD mucus stays in the lung because the cilia do not work properly. In a normal person the cilia in the lung act like an escalator on the underground, but in PCD the cilia act like an escalator which is not moving. Therefore physiotherapy is needed to get rid of this mucus.

If PCD is managed well, then a normal and active life can follow. It is important to break the cycle of sputum getting trapped which causes infection and inflammation. This in turn causes airway damage, which then leads to more sputum being trapped.

Physiotherapy can help enormously to break this cycle, but lung damage can be irreversible. It is important to keep the chest clear which will reduce infection. Coughing is not enough, sputum needs help to move up.

Plugging of the airways occurs if sputum is not cleared which can cause inflammation, pain, infection, breathlessness and wheeze. Physio helps by delaying lung damage, reducing wheeze, reducing the rattle of cough, and helps prevent infection and inflammation.

„It needs to be stressed that each patient is an individual, and that there are many different kinds of physiotherapy to suit each individual.“

13.2. Active Cycle of Breathing Technique (ACBT)

This method is flexible, and can be used with a lot of other techniques.

It is easy to remember, and can be used in conjunction with postural drainage and blowing games etc.

The active cycle of breathing technique uses:-

- Breathing control
- Thoracic expansion
- Forced expiration/huffing



For babies and young children it helps to make physiotherapy fun. If you finish off a session with tickling it helps clear the chest and they enjoy it too. Playing hide and seek with a toy car/doll/hand puppet whilst the child is upside down on the wedge doing chest physio postural drainage.

The hand puppet can be used to give physio instructions, sometimes in funny voices, daft instructions, or hiding under clothes or behind the wedge. The puppet can sit on the child's his tummy in order to move in and out when tummy breaths (diaphragmatic breathing) are needed.

Another technique is to get the child to blow. Blowing anything: bubbles or cotton wool off a hand, or kazoos or blowing the hand puppet off the tummy or wedge can help.

13.3. Drainage

For this method the patient needs to be comfortable and lies on his/her side in a gravity assisted position.

It can be done on a physio table or pillows so that the patient lies in a tilted position which allows gravity to drain the lower lobe. If the patient lies further back then this drains the middle lobe. However this method should not be used if the patient has reflux problems.

It can be used in conjunction with the ACBT technique.

Autogenic Drainage

Is a 3 phase breathing technique which uses your air flow and gets you to breath at different levels to unstick, collect and evacuate mucus.

It can be completed in any position and can be done at work.

13.4. Using adjuncts for physiotherapy

Various devices can be used to increase the efficacy of physiotherapy, and can also help with compliance if there is reluctance to do it.

Oscillatory and positive expiratory pressure (PEP) devices such as the Pari PEP or Astra PEP help mobilise and move secretions. This positive pressure on breathing out helps to 'splint' the airways open, enabling more air to get in and out of the lungs increasing the efficiency of mucous clearance.

The Acapella Choice and Flutter are oscillatory PEP devices which provide both resistance and vibration as you exhale. It may be necessary to hold the cheeks to make sure that the vibration is in the lungs and not the cheeks. An Acapella can be used in the postural drainage position or the seated position. The Flutter is easiest used in a seated position.

Using the Flutter: The user simply exhales a number of times into the mouthpiece of the Flutter - causing a steel ball to vibrate in the casing. The pressure of blowing against this steel ball, and the vibrations that it makes travel back into the lungs, where they start to have a positive effect. The flutter can really only be used in the sitting position because it needs to be held upright.

These vibrations affect all the branches of the airways, and like "shaking apples from the branches of a tree", the mucus is loosened. Whilst this is happening, the changes in air pressure that makes the steel ball move are both keeping small airways open, and allowing air to move throughout the lungs. As a result of this movement, mucus is moved out from where it was previously stuck, into the larger airways and to where it can be coughed out. The Acapella has a similar effect but uses a seesaw mechanism to cause the vibration instead of a ball.

PEP devices (Positive Expiratory Pressure) help keep the airways open by blowing against resistance. It therefore make it easier to cough up the mucus.

Ask your physiotherapist if you can try any of these adjuncts.

Please note it is important that a physiotherapist assesses which device or technique may be most appropriate for you and teaches you how to use it. As every person with PCD is an individual it is important to find the right technique to suit you.

The physiotherapist can also help you work out how to fit your physiotherapy around your lifestyle.

„It is important once a week (to avoid unnecessary chest infections) that you sterilise your adjuncts as per the manufacturer's instructions.“

13.5. Additional Advice

Evidence also suggests that exercise is almost as effective as puffers. Trampolines are particularly effective. Activity is really important, and sport should be encouraged. It can be helpful to use a flutter before football.

There are various aids to loosen mucus, thereby making it easier to cough it up. These are mucolytics and include hypertonic saline (i.e. salty water) which can be inhaled by nebuliser. This is not suitable for everyone, and the airways need to be checked for toleration. There is also Mucodyne and Dnase medication.

In order of efficiency, the most efficient ways to clear the airways:

- Open airways (using inhalers)
- Use Mucolytics
- Airway clearance
- Steroid inhaler
- Inhaled antibiotics

The following questions should be asked so that the patient knows they are doing what is right for them and what works for them:

- Is my physiotherapy effective?
- Is my condition stable?
- Do my physiotherapy techniques fit with my life style?

Each patient should ask for a physio review periodically.

„The challenges for effective physiotherapy are adapting and tailoring techniques to age and circumstances e.g. transition experiences, adapting to work, relationships, pregnancy etc.“

14. Resources – Support Groups

The [Dutch PCD support group](#) have their own site with information provided in both Dutch and English

Susanne's [German PCD/KS Support Group](#)
Information in several languages including English

The [Centre for PCD diagnosis and therapy in Pisa](#), Italy have an Italian language website about their work.

The [P.C.D. Foundation](#) is a support group for P.C.D. patients in the United States