pkd charity newsletter
Polycystic Kidney Disease

The PKD Charity PO Box 141 Bishop Auckland County Durham DL14 6ZD
Tel 01388 665 004 Email info@pkdcharity.org.uk

www.pkdcharity.org.uk
Spring 2007

Registered Charity No. 1085662

Launch of patient support groups

Wirral Walkers raise over £5000 for kidney patients

Emma Kennedy has walked over 60 miles with friends and family and raised over £5000 to provide support for people with kidney disease.

See page 5 for the full story.

Three PKD support groups are to be piloted this spring in Twickenham and Richmond, Chester, and Moodiesburn in North Lanarkshire. We hope that more groups will be set up during 2007.

The groups will give people affected by polycystic kidney disease the opportunity to meet, receive information and give each other support.

The PKD Charity will monitor the pilot support groups in order to assess the level of financial and volunteer contribution involved. We will also explore the interests, needs and requirements of the people who join or are interested in joining a group–so do give us your feedback.

Meetings will be informal though professionally run, so that everyone who attends feels comfortable, valued, empowered and positive about managing his or her PKD.

There are other organisations and charities which address kidney conditions but the PKD Charity is the only one that focuses on polycystic kidney disease, which, as we know, is a genetic and a systemic condition that can affect other organs.

For further information please contact Maggie Pope, Support Group Coordinator:

Tel 0845 241 0964
Email maggie@pkdcharity.org.uk

Too few kidney transplants in UK

Kidney transplant is the preferred treatment for renal failure.

A transplant can transform your life, improve the quality of your health, increase life expectancy and free you and your family of the burden of dialysis. Survival rates for renal transplants have doubled in the last decade.

Yet, there are too few kidney transplant operations in the UK. In the 12 months before March 31, 2006, nearly 1,800 transplants were performed. But this represents less than one third of people waiting for a donor kidney, and a tenth of those on dialysis.

The UK has a meagre donation rate compared to other western countries. Waiting lists are growing and patients die while waiting for an organ.

We estimate there are around 4,000 people with PKD in the UK on dialysis, less than 200 of whom receive a donated kidney each year.

A cross-party group of politicians – the All-Party Parliamentary Kidney Group – has a particular focus on transplantation. During 2006, they held a summit and this resulted in the development of a Transplant Manifesto.

The Manifesto will be used to raise the issue in Parliament and challenge the Government. It is also a very useful reference for clinicians, patients and all kidney charities.

You can download a copy of the Transplant Manifesto from our website:

www.pkdcharity.org.uk

Or contact Tess to request a hard copy:

Tel 020 7387 0543
Email tess@pkdcharity.org.uk

Register now

Register now for our fourth PKD Information Meeting in Birmingham

Friday Jul 6 informal meet and greet

Saturday Jul 7 full programme of presentations and workshops on various aspects of PKD

Sunday Jul 8 support group workshops

Topics to be covered during the weekend include pregnancy, pain management, the progression of PKD, genetics and much more.

The event will be held at the Queen Elizabeth Medical Centre in Birmingham and information about local accommodation will be made available.

Contact justina@pkdcharity.org.uk or call 01246 823 468 to book your place and to find out more, or visit our website.

Understanding clinical trials

There is currently no cure for PKD, so new drugs and treatments are of great interest to us at the PKD Charity.

People with PKD may be asked to get involved in the clinical trial of a new treatment. Here, we give a guide to the trial process, questions to ask if you are considering getting involved, and an update on current trials.

The trial process

Clinical trials involve people in testing the safety and effectiveness of new drugs and treatments before they are made available. They are carried out in three phases, each with more people in increasingly exhaustive tests.

Phase I – Is it safe? The drug or treatment is tested with 20 to 80 people who do not necessarily have the disease to ensure it is safe, what its side effects are and to establish the maximum safe dose.

Phase II – Is it effective and safe? If phase one is successful, the treatment is trialled with a group of 100 to 300 people with the disease.

Phase III – Is it better than the current treatment? It is now tested with thousands of people at different centres over a longer period–typically two years or more. The treatment is compared with others, and sometimes placebos are used.

In Phase III and some Phase II trials, people are randomly selected to receive either the new treatment, the placebo or the current treatment.

Current drug trials

Tolvaptan People with PKD in the UK, and other countries, are being enrolled for Phase III trials. Ask your GP, consultant or nephrologist if you would like to participate in this highly interesting trial, which is looking at the effectiveness of Tolvaptan to reduce cyst growth.

Rapamune (Sirolimus/ Rapamycin/Everolimus) Rapamycin is already used to help prevent kidney rejection. Phase II studies have begun in Switzerland and the United States to investigate whether this drug also slows down cyst growth and the deterioration of kidney function in people with PKD.

PKD Gene Modifier Mapping Study - Canadian Institutes of Health Research This observational study is looking into the genetics of type I ADPKD. There are ten different participating ADPKD research centres in North America and Europe.

HALT PKD Treatment Network (US) This study is investigating the effect of two anti-hypertensive drugs on PKD and its cardiovascular complications. It will determine whether a combination of drugs is more effective than a single agent.

Statin Therapy (US) This trial will assess the effect of ACE inhibition on cyst growth in children and young adults with ADPKD. The results could impact on the standard of care for young people with ADPKD.

ARPKD and Congenital Hepatic Fibrosis The US National Human Genome Research Institute wants to collect comprehensive data on kidney and liver disease in ARPKD/CHF and follow patients over time to provide the groundwork for more focused studies and new treatments.

With thanks to Dr Patricia Wilson, Professor of Medicine (Nephrology) of the Mount Sinai School of Medicine, New York.

Sixteen questions to ask before you enrol for a clinical trial

1. How long is the trial?
2. Who is taking part in it?
3. What are the risks and benefits?
4. Are there likely to be side effects? If so, what are they?
5. How will my daily life be affected?
6. How often must I take the drug, when and for how long?
7. How often will I have to visit the clinic or surgery? What will happen at these visits?
8. Will I have extra tests?
9. What other medication can I take?
10. What can't I do or take during the trial?
11. What will happen if my condition worsens?
12. Will I be told the results of the trial?
13. Who is funding the trial?
14. Will my travel expenses be paid to take part?
15. Who can I talk to if I have any more questions?
16. Can I withdraw?

Tribute to former chair of the PKD Charity

Dr Peter Lockyer, former chairman of the PKD Charity, died in December 2006, aged 34. Peter is survived by his wife Julie and three children, Phoebe aged eight, Martha, four, and Molly who is three.

Peter got involved with the PKD Charity in 2002, first as scientific advisor and then as chairman, a role he left when he became ill in 2003. Tess Harris, current chairman of the charity, said, "Pete was dedicated to our cause. Every trustee of the charity, past and present, greatly values his contribution."

Peter organised the charity's first information day in 2004. It was a huge success and we continue to organise similar events annually. In 2005, he organised a two-day scientific conference, a major milestone in the charity's history, which was attended by leading researchers and clinicians from around the world.

Peter was a successful scientist who studied Biochemistry and Physiology at Southampton University, where he later completed his PhD.

Between 1997 and 2002, Peter continued his research as Post-Doctoral Fellow at Bristol University, where he was awarded a Beit Memorial Fellowship. Peter published several notable research papers during this time and was regarded as a rising star in his field.

In 2002, Peter began an independent research group at the Babraham Institute. Former colleague Dr Phill Hawkins said of Peter, "He was a fabulous guy, a great scientist."

Peter's family have asked that well wishers make donations to the PKD Charity

Want to support PKD research?

We set aside a percentage of donations to fund research into treatments and a cure for PKD. You can designate an entire donation for research–just let us know your wishes at the time of giving.

PKD – genetic testing and counselling

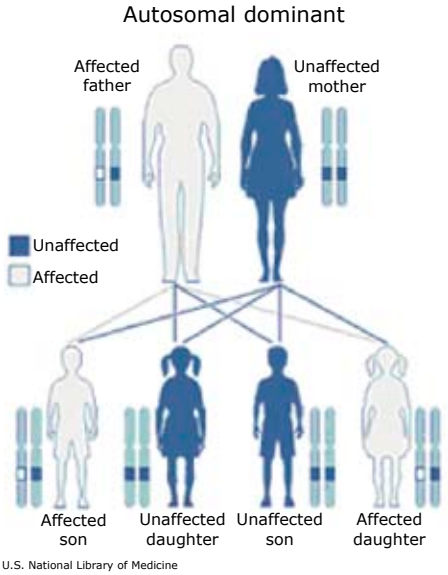
Polycystic kidney disease (PKD) is a genetic disorder caused by one or more genes functioning abnormally.

Genes exist in pairs. In autosomal dominant (AD) PKD, one abnormal copy of the pair is inherited, usually from one affected parent. The abnormal gene is found at either chromosome site PKD1 or PKD2.

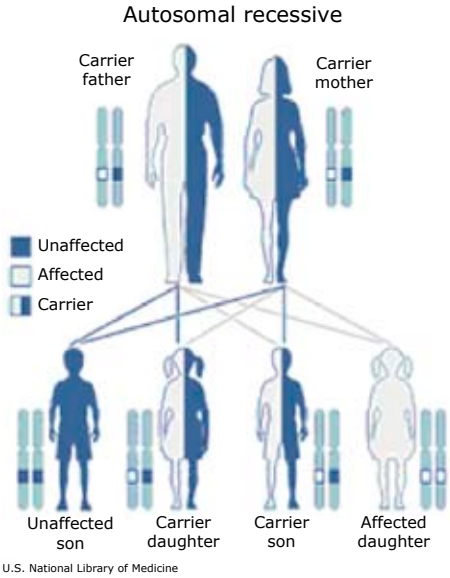
Autosomal recessive (AR) PKD is rarer and occurs when a child inherits two faulty genes - one from each parent. Parents are not usually affected as they carry one abnormal copy.

Genetic, or molecular, testing is of great interest to PKD patients. The test checks blood DNA for an abnormality that may indicate a predisposition to a disorder, or confirms a suspected mutation.

In PKD, a gene change must be found before testing family members, otherwise predictive testing is not possible.



Autosomal Dominant PKD – Two faulty genes have been found to cause ADPKD. PKD1 accounts for about 85% of affected individuals and is associated with renal failure up to 20 years earlier than those with PKD2, which accounts for about 15% of cases.



Autosomal Recessive (AR) PKD – It is believed that around 1 in 50 carry one copy of this gene. It is recessive, which means there is a 1 in 4 chance that any child born to parent-carriers will inherit two copies and will be affected by the condition.

Is an early or genetic test useful for me?

ADPKD

If you have ADPKD symptoms, a genetic or molecular test is not usually necessary. The condition is diagnosed by a scan (ultrasound, CT or MRI). The number of cysts, if any, together with your family history will be sufficient to confirm whether you have the disorder or not.

It is important to bear in mind that, while a scan or a molecular test may confirm that you have the disorder, it will not tell you when problems will begin, how severe they will be, the range of symptoms, or how quickly your kidneys may deteriorate.

While statistics suggest that PKD1 is more severe than PKD2, there is very wide variation in the severity of the disease overall. Knowing which gene change you inherited will not help you manage the condition better. In fact, there may be negative consequences. You must disclose a diagnosis when applying for insurance or if declaring medical conditions.

Generally, testing children for PKD is not advised unless they show symptoms. Pre-symptomatic diagnoses remove a child's right to choose and may cause them distress growing up.

On rare occasions, parents request a PKD gene test on a foetus. If there is no known gene change in the family this may not be possible to do. However, where there is a significant risk to the unborn child, genetic testing along with counselling, should be investigated.

Similarly, if a relative of someone with PKD is considering donating a kidney and scans are not conclusive, a genetic test may help the donor decide.

ARPKD

Prenatal or pre-implantation genetic diagnosis may be advisable when there is a family history of ARPKD, miscarriages, or if a previous child was born with the condition.

A diagnosis of ARPKD is made during pregnancy or soon after birth.

Learn more about genetics

To find out more, visit:

- www.bshg.org.uk
- www.ukgtn.org
- www.gig.org.uk
- www.phgu.org.uk

Genetic testing and counselling services

If you have a family history of PKD, you have a right to access genetic services through the NHS. A GP, paediatrician or renal consultant can refer you to a clinic for information and support from a specialist doctor, usually a clinical geneticist.

You may receive a questionnaire about your family history before your appointment to help the geneticist calculate your risk factors. When you meet, the geneticist will discuss your concerns and draw a family tree. You may also need a clinical examination.

These services are specialised and are not available at every hospital, but there are 22 genetics centres in the UK (2006 data) so there is no need for anyone to be denied access.

Details of these centres can be found at– www.bshg.org.uk.

Adopt the PKD Charity

We are looking for a business to adopt us. Interested? Email tess@pkdcharity.org.uk

Insurance–got a story?

We want to hear your experiences of applying for life, medical and travel insurance. Email tess@pkdcharity.org.uk

What your blood results tell you

Making sense of the many blood tests used in monitoring PKD isn't easy. Here we give a brief overview.

GFR

The most precise indication of kidney function is the glomerular filtration rate (GFR), which is normal at around 100 millilitres per minute. This figure will go down if your kidney function decreases.

GFR can be measured accurately, but it is more commonly estimated (eGFR) using a creatinine blood test, together with your age and sex. Current guidelines say that clinics should tell you your eGFR. If yours does not, you can use an online calculator, such as–www.renal.org/eGFRcalc/GFR.pl.

Creatinine

The creatinine test is the best routine way of seeing how well your kidneys are working. A normal level of creatinine in the blood is between 60 and 120 mmol/l. This figure rises as kidney function goes down.

Creatinine is excreted by muscle, so someone with little muscle may show normal levels until around 50% of their kidney function has been lost–eGFR will help give a clearer picture. Speak to your GP if you have any concerns about the accuracy of your GFR measurement.

Urea

Urea is produced from protein and excreted by kidneys. The level rises as kidneys fail. The urea measurement is not very accurate on its own but is useful together with a creatinine test.

Potassium (K)

Kidneys excrete potassium and levels are regularly checked to ensure they are safe. High levels are treated–initially a restricted potassium diet is used but, if you have persistently high levels, your GP or clinic will prescribe treatment.

Sodium (Na)

Increased sodium can cause high blood pressure, swelling and fluid retention in the lungs. When kidneys do not work so well, sodium, which comes from salt, increases in the body.

Bone disease

Bone disease can affect people who have had kidney disease for a long time. Unfortunately, the treatments for this can raise the amount of calcium in the blood to harmful levels so calcium, phosphate and parathyroid hormone levels in blood are frequently monitored.

Haemoglobin (Hb)

The kidneys help stimulate the production of red blood cells by producing EPO. Loss of EPO function in kidney failure causes severe anaemia but there has been a huge advance in treating this. Haemoglobin tests check the dose is right, detect bleeding and other potential problems.

Test	Normal result
GFR	100 ml/min approx
Creatinine	60-120µmol/l
Urea	2.5–6.6mmol/l
Potassium	3.6–5.0 mmol/l
Sodium	135–145 mmol/l
Calcium	2.1–2.6 mmol/l
Phosphate	0.8–1.4 mmol/l
Parathyroid hormone	According to lab
Haemoglobin	13–18 g/dl (men) 11.5–16.5 g/dl (women)

Blood pressure–take control

Raised blood pressure, or hypertension, is very common in people with ADPKD. It is not fully understood why, but we think that the cysts compress normal kidney tissue and this leads to excessive salt and water retention. Some research has suggested that increased activity of the reninangiotensin hormone system may play a part, though this remains controversial.

In the general population, raised blood pressure increases the risk of heart attacks and strokes. Rigorous blood pressure control is likely to reduce these complications.

So far, there is no research specifically on whether lowering blood pressure reduces the risk of heart attacks and strokes in people with ADPKD, though these are the major causes of death and disability in people with ADPKD.

There has been a study into a condition called left ventricular hypertrophy (LVH), where the heart muscle becomes thickened, which is common in people with ADPKD. It has been shown that people who had controlled blood pressure to 120/80 mmHg experienced greater reduction in LVH than those whose blood pressure was less tightly controlled.

So far there is not sufficient evidence to say whether lowered blood pressure also slows down the deterioration of kidney function in people with ADPKD.

Intracranial aneurysms – bulging of blood vessels within the brain – are more common in people with ADPKD than in the general population. Rupture of aneurysms can result in fatal bleeding, and treatment of raised blood pressure may also reduce the risk of this happening.

by Dr Tim Doulton, BSc MRCP, St George's Hospital, London

For more information, visit:

Blood Pressure Association
www.bpassoc.org.uk

British Hypertension Society
www.bhsoc.org

NICE for latest guidelines on hypertension management–www.nice.org.uk

Here's how

Aim for a blood pressure of 130/80 mmHg, or 130/75 if you have proteinuria.

- Reduce salt intake–less than six grams (a teaspoon) a day
- Exercise for 30 minutes three times per week
- Avoid smoking
- Don't drink alcohol excessively
- Eat a healthy diet and avoid saturated fat
- Maintain your ideal weight

Want to talk to others with PKD?

The PKD Online Support Group provides a friendly and informal way to share information and advice, exchange experiences and give and receive support–all via email.

Visit our website to sign up:

www.pkdcharity.org.uk/support.html

Dragon Ride

Image used with thanks to www.dgs-photography.co.uk

Chris Cottrell is a 46-year-old man with PKD and a keen cyclist. Here he describes the Dragon Ride, one of the UK's premier cycling events.

"I ride a bike to keep fit and help control my blood pressure. Last spring, I decided to enter the Dragon Ride. I asked a couple of friends if they would join me and help raise money for the PKD Charity. Jon Shaw, a software engineer and Mike Prior, senior police officer, agreed–two heroes in Lycra! I'm an account manager in the telecommunications industry, so all in all, we were an unlikely combination for a major bike riding challenge.

Using the great JustGiving service, the website was born. We initially set a modest fundraising target of £500, but we reached £1,500 before the event and at the time of writing it is looking like we've raised £2,500, with GiftAid!

The day of the ride dawned cool and sunny. The team converged on Bridgend ready for the 8 am start, pockets full of food and bottles full of water. The hooter sounded and we were off. The ride itself was great. It was a very long way; my cycle computer showed 101 miles in seven hours and thirteen minutes. My elapsed time was eight hours, four minutes and 59 seconds, including various stops for mechanicals and food. Was it hard work? Oh yes: 6,500 calories; 67,000 heartbeats. Did I enjoy it? The downhill bits were great. My top speed was 45 mph.

My PKD? Difficult to tell, I had a few aches and pains but I'm 46 and I had been riding a bike for eight hours. My blood pressure is great thanks to my medical team of Doctors Matthew Dolman and Chris Dudley.

Would I do it again? Yes, or something similar, for as long as I can. Cycling has very low impact on your body when you go steady. It is fun, great for general fitness and weight control. Why not try it?"

Thanks to Chris Cottrell.

If you would like to write an article or suggest a topic, contact tess@pkdcharity.org.uk.

Wirral Walkers raise over £5,000 for kidney patients

Emma Kennedy has walked over 60 miles with friends and family and raised over £5,000 to provide support for people with kidney disease.

A new trustee of the PKD Charity, Emma is a nurse at the Countess of Chester Hospital. She first took part in the Wirral Coastal Walk in 2003, in aid of the Haemodialysis Fund at the hospital. The following year she persuaded a team of her friends and family to join her and they continued walking in 2005 and 2006.

The money she raised up to 2005 was used to fund a new Kidney Patient Resource Room in the Renal Unit, where patients can read books or access the internet, or sit and relax while waiting for a consultant or dialysis. Last year, Emma's family donated the funds from the walk to the PKD Charity.

From January 2007, Emma is planning to hold PKD Support Groups in the Resource Room at the hospital. If you live in West Cheshire or South Wirral areas and would like to join the group, please contact Emma using the email or phone number below.

Emma, along with her friends and family, will be taking part in the Wirral Walk again this year on Sunday, May 20th.

This year they will be wearing fancy dress and Emma promises to be dressed as a giant kidney.

All money raised will go to the PKD Charity.

Contact Emma about the PKD support group at Chester Hospital.

Email info@pkdcharity.org.uk
Tel 0845 241 0964

If you would like to sponsor Emma, please visit the PKD website to see how:

www.pkdcharity.org.uk

Supporters tool-kit

There are a large number of ways you can help the PKD Charity to continue to provide a voice for people with polycystic kidney disease, fund research and enable people with PKD to meet and give each other information and support.

Our fundraising packs include newsletters, leaflets and sponsorship forms – use them to tell people about PKD and to raise money. These are available on request.

Sponsorship forms can also be downloaded from our website.

The website JustGiving enables supporters of the PKD Charity to provide an online sponsorship service. Add your fundraising event by setting up your own page and your friends, family and colleagues can sponsor you using their credit or debit card. It's simple to use, a secure, tried and tested service–and fun. Give it a try!

Just Giving
www.justgiving.com/pkd/raisemoney

For a fundraising pack contact
Email rebecca@pkdcharity.org.uk

Thanks to our donors

We would like to express our deepest gratitude to our generous donors without whose help the PKD Charity could not survive.

The Murphy Family, The Kennedy Family, St George's Medical Students' Rag, Awards for All, D'Oyly Carte Charitable Trust, The Kessler Foundation, Chris Cottrell, Otsuka Pharmaceutical, Fiona MacDonald's Brownie Pack, The Steve Morgan Foundation, John Mold, London Scottish, Rothschild's, Joanna Brown, Nicholas Reddington, Marina Franklin, Jacky Fitzsimons, in memory of her mother Gladys Nelson, friends and family of Stephen Howsley, Karen Robins & JWT, family and friends of Dr Peter Lockyer.

Donors - use Gift Aid

Please fill in a Gift Aid form when donating–the Government adds 28p to each £1 you give.

Remember us

When writing a will, please consider the PKD Charity. Contact us to find out more.

Recommended reading

The following books are made available from Class Publishing with a great discount to readers of the PKD Charity newsletter. Quote PKD6 when ordering.

Kidney Failure Explained (third edition)

by Dr Andy Stein and Janet Wild (£17.99 RRP)

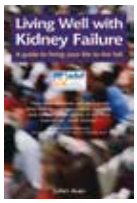
This handbook on kidney failure provides up-to-date, accurate information and practical advice from experts. It cuts through medical jargon, explaining terms in plain English and tackles the questions you may feel uneasy asking your doctor.

Eating Well with Kidney Failure

Helena Jackson, Annie Cassidy and Gavin James (£14.99 RRP)

This brilliant book provides a straightforward guide to eating well with kidney failure, including a collection of over 50 delicious recipes to show you how it can all work in practice.

The recipes have been analysed for their nutritional content and are coded to help you choose the most appropriate dishes for your particular requirements.

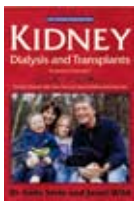


Living Well with Kidney Failure

Juliet Auer (£14.99 RRP)

This inspiring book will give you the confidence to live a full and rewarding life.

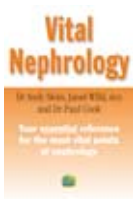
It draws on the experiences of a number of very different people with kidney failure, from all walks of life and ages and family situations. Their shared personal accounts celebrate the achievements, both small and large, of these people successfully living with kidney failure.



Kidney Dialysis and Transplants: Answers at your fingertips

Dr Andy Stein and Janet Wild RGN (£15.99 RRP)

This clear, comprehensive guide provides straightforward answers to over 450 patient questions, and is packed with positive advice about every aspect of dialysis and transplants, and how to carry on with an ordinary life.



Vital Nephrology: A Guide for Healthcare Professionals

Dr Andy Stein, Janet Wild and Dr Paul Cook (£14.99 RRP)

A 'fact file' of information about the most vital point of nephrology. It is an essential handbook for anyone on the renal team: clinician, nurse, dietician, social worker, counsellor, educator or expert patient.

Beating Depression

Dr Stefan Cembrowicz and Dr Dorcas Kingham (£17.99 RRP)

Beating Depression starts with the basics. It first answers real questions about what and why, then objectively explains all the available treatment options. It also advises on techniques for dealing with common symptoms such as insomnia, agitation, panic and apathy.

For further information please visit www.class.co.uk/books-kidneys-57

You can also buy these books by phone or mail-order. To order call 01256 302 699 or send a cheque to Class Publishing (Priority Service), FREEPOST, London W6 7BR. You must quote reference PKD6 when placing your order.

Help us spread the word

"I am a freelance journalist looking for health stories to feature as leads in Bella magazine's health pages." This is just one of many requests the PKD Charity receives from the press, radio and television every month.

We are now looking for a group of volunteers who are prepared to be interviewed on the subject of kidney disease and possibly photographed or filmed.

Most requests are for personal stories, explaining what it's like to live with kidney disease, how it affects relationships and family. The theme is often 'triumph over adversity'.

These articles and programmes provide an invaluable platform from which we can raise awareness of PKD and the PKD Charity, raise funds for research and campaign for more donor kidneys.

Are you willing to give it a go? We will provide a briefing and tips on working with the media to all, however, if you have some experience of press relations, we would particularly like to hear from you.

Please get in contact with Tess Harris if you are interested.

Email tess@pkdcharity.org.uk

Tell us your views

The PKD Charity is run by and for people with polycystic kidney disease, so we would like to hear from you.

- Send us an email – we may even publish it in the next newsletter
- Tell us what more we can do to help
- Give us feedback on our work

Email tess@pkdcharity.org.uk

New trustees

The PKD Charity welcomes five new trustees to its board: Cath Buchan, Chris Butler-Donnelly, John Eatough, Emma Kennedy and Christine Wallach.