pkd charity newsletter
Polycystic Kidney Disease
www.pkdcharity.org.uk Issue 11 - Autumn 2011

Rachel Green raised £272 on the first PKD Charity skydive (see page six)

Our survey says...

Thank you to everyone who found time to complete our recent survey. The response was amazing and we received over 300 completed forms, mainly from those with PKD but also from their carers, relatives and friends.

We asked specifically about the information people are given when they're diagnosed with PKD and what information they would have liked to receive, and also for feedback on the support the PKD Charity offers.

We received a huge amount of useful information, for which we're extremely grateful. Below is a summary of the key findings:

- 366 surveys were completed
  - 315 by people with PKD
  - 51 by carers / relatives
- 66% of respondents were female
- nearly 70% were either given 'no information' or 'not enough' on diagnosis
- minimal social, emotional and psychological information (including genetic counselling) was given
- very few respondents were told about charity or support groups by their doctor
- most had never been offered a Kidney Care Plan or other personalised healthcare plan
- there were many requests for local support groups
- more psychological help was felt to be needed.

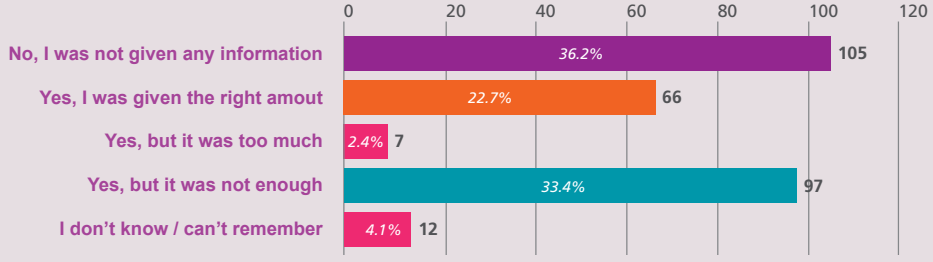
Many of you wrote extensively on the information and support you would have liked to receive, such as the impact PKD will have on your lives, what symptoms to look out for and how to stay as healthy as possible.

Sadly, many of you told us, 'I was just told I had PKD and that was all'. However, not all the responses were negative, and many positive points were made both about good doctors and the information we provide.

Once your replies have been fully analysed we'll use the results to:

- develop more / improve patient support materials
- tell the NHS about the information needed by people with PKD
- improve the support we offer to patients and their carers, relatives and friends.

When you first found out that you had PKD, were you given any information about the condition?



ARPKD family information day

We established the PKD Charity to support everyone affected by PKD but we often feel helpless about the families and children who inherit ARPKD because this condition is so rare and complex.

Over the past six months, we have been contacted by a number of families who have children affected by ARPKD: some are looking for support and others are fundraising and setting up groups, like the families of Madison Moore and Harrison Caines (see pages five and six for details). Our sincere thanks to both families.

One of our objectives for 2012 is to organise a family conference on ARPKD, with the help of some supportive paediatric nephrologists. More details about this event will be posted on the PKD website in the New Year.

Email tess@pkdcharity.org.uk to be kept updated.

Calendar

2011

October 16 Cardiff Half Marathon
Linda Cooke and Team
www.justigivng.com/linda-cooke

October 24
Sydney Harbour Bridge Climb
Caroline & Craig Kinshott
www.justgiving.com/
SydneyHarbourBridgeClimb

November 1 Deadline to apply for PKD's 2012 London Marathon place: esther@pkdcharity.org.uk

November 26 PKD Information Day Leicester Royal Infirmary.
More info: www.pkdcharity.org.uk

2012

January 27
James Voce's Kilimanjaro Climb

March 11
Adidas Silverstone Half Marathon More info:
esther@pkdcharity.org.uk

22 April London Marathon

20 May Great Manchester Run
More info:esther@pkdcharity.org.uk

Polycystic liver disease

Approximately half of all patients with ADPKD will have multiple liver cysts, which increase with age and the severity of kidney cysts.

For most people polycystic liver disease (PLD) causes no symptoms and liver cysts are detected only during an ultrasound scan. Whilst liver function is usually maintained, PLD can occasionally cause ill health and may very rarely be massive and life-threatening. Women tend to have larger cysts than men, this is thought to be associated with female hormones and multiple pregnancies (more than three babies).

Abdominal distension and bloating are common in PLD. Complications include cyst bleeding, infections and rupture. Infections may arise after kidney transplant or other surgery, or if on haemodialysis. People may experience early fullness on eating, heartburn or vomiting, which can result in malnutrition and loss of muscle/fat. A change in bowel pattern or haemorrhoids may occur. Other rarer complications include hernias, uterine prolapse, rib fractures, venous obstruction (hepatic, IVC, porta) and bile duct obstruction.

Management is 'conservative' as nothing can be done to stop the cysts forming and enlarging. Avoiding caffeine and oestrogen hormones is recommended. Women about to enter menopause and who are considering hormone replacement treatment should ask to see a specialist to discuss the best option.

There are some surgical treatments to relieve symptoms. Single, massive cysts can be drained followed by sclerosis (replacement of cyst fluid with ethyl alcohol). Some people with multiple cysts are offered surgical fenestration - opening or 'unroofing' the cyst and removing the fluid. In extreme cases, partial liver removal (a resection) or a full liver transplant may be necessary. Other newer treatments such as selective hepatic artery embolisation to reduce the mass of the liver are also being tried.

More promisingly, two types of drugs are being evaluated in clinical trials in a few overseas centres: Octreotide/Lanreotide and Sirolimus. Some reduction in liver volume has been observed.

Need financial help?

If you're struggling with the impact of PKD on your life, like covering the costs of your domestic bills, hospital travel, education and holidays, you could qualify for a grant from the British Kidney Patient Association.

More info: www.britishkidney-pa.co.uk/grants.html

Kidney transplants in the UK

Nearly 7,000 people are on the active kidney transplant list. 25,000 are on dialysis, 8% of whom have ADPKD. A transplant costs the NHS less per year than dialysis – £6,000 compared with £40,000. Alas, last year (to 31 March 2011), kidney transplant numbers hadn't increased, at just over 2,600. However, it's promising that Organ Donor Register numbers are rising annually, giving hope of more transplants from deceased donors. For details on kidney transplants and organ donation, visit www.organdonation.nhs.uk and click on 'Statistics'.

Focus on autosomal recessive PKD

Autosomal Recessive PKD or ARPKD is a rare condition affecting 1 in 20,000 live births, boys and girls equally, so potentially 3000+ in the UK. It causes enlarged kidneys (with or without cysts), liver enlargement and high blood pressure.

In ARPKD, small cysts form in a specialised part of the kidney tubule called the collecting duct.

The abnormality always involves both kidneys. Because thousands of cysts develop, the kidneys can become quite enlarged. In addition, the normal function of the collecting duct is disrupted. In the normal kidney, the collecting duct fine-tunes the amount of water and acid in the urine. In ARPKD the cystic collecting ducts cannot retrieve water efficiently, causing much more urine production than in children with normal kidneys.

ARPKD can be diagnosed prenatally or after birth. In utero, kidney function may be impaired, which decreases the level of amniotic fluid. Since amniotic fluid is vital for lung development, newborns with ARPKD often have underdeveloped lungs. 30-50% of these newborns die because of serious breathing difficulties and other lung complications. Some pregnancies are terminated.

Survival rates increase past infancy but most children with ARPKD face debilitating health problems and will likely develop kidney failure at a young age. For these children, approximately one-third will need dialysis or transplantation by the age of 10. High blood pressure is common and needs strict control with drugs.

The disease also affects the liver, causing a condition called congenital hepatic fibrosis (CHF). This disease may result in an enlarged liver and spleen that can cause life-threatening gastro-intestinal bleeding and low blood cell counts.

The genetics of ARPKD

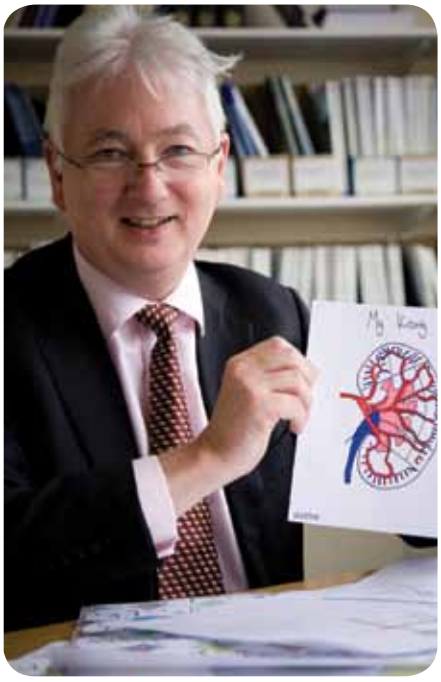
ARPKD is a recessive condition caused by a single defective gene called PKHD1. The affected child inherits a copy of the PKHD1 gene from each parent. Since the parents each have only one copy of the disease gene, they do not have the disease and are "carriers". For carrier parents, there is a 25% chance in each pregnancy that both copies of the disease gene will be transmitted to the baby.

Research and future therapies

ARPKD is incurable and there are no direct treatments. Dialysis and transplantation remain the only long-term life-saving treatments, combined with blood pressure management and ongoing monitoring/control of the other complications.

Ask three questions

Our regular advice column from Dr Donal O'Donoghue, Renal Tsar for England



If you're asked to make a choice about your healthcare, then make sure you always get the answers to these questions:

- What are my options?
- What are the benefits and possible risks?
- How likely are these risks and benefits?

These are the three questions that MAGIC (Making Good Decisions in Collaboration) is promoting. MAGIC is a Health Foundation programme for shared decision-making that has been running in Newcastle and Cardiff.

Recent surveys show that 48% of inpatients and 30% of outpatients want more involvement in decisions about their care. Greater involvement also results in better experience and outcomes - a Cochrane review in 2009 found that decision support leads to fewer patients being undecided and improves adherence to medication due to:

- improved knowledge
- more accurate understanding of risk
- increased participation and comfort with decisions.

Recently I was privileged to visit the MAGIC site in Newcastle. I assumed I'd spend the train journey battling with my inbox when to my delight I bumped into Sir John Oldham, a GP from Glossop and the DoH Quality, Innovation, Productivity, Prevention lead both for long-term conditions and acute care. We also travelled up with Alf Collins, Consultant in Pain Management, whose publication with Angela Coulter, "Making Shared Decision Making a Reality, no decision about me without me", had been released the day before and was receiving a lot of media attention.

The day's highlight was undoubtedly visiting Dave Tomson's surgery, where I saw the three questions in use and sat in on a shared decision-making skills workshop. It struck me how far ahead of hospital practice primary care is in terms of ensuring the skills needed to talk, understand and listen to patients are well taught. Clinical encounters may only be 10 minutes or less but, in addition to the dialogue, they're full of body language, unsaid messages, anxiety and anticipation. Offering choice, explaining options, providing decision support, eliciting preference and achieving good decisions is a real skill. Like all skills it needs practice before it becomes automatic and 'just happens'.

Back at the Royal Victoria Infirmary in Newcastle we heard from Richard Thompson (Professor of Epidemiology and Public Health) about shared decision-making in breast surgery, obstetrics and urology and discussed the importance of leadership, learning sets and team engagement in addition to clinical skills. The emerging lessons highlighted some of the NHS-wide barriers and disincentives that need to be addressed if we are to build on MAGIC's achievements and secure sustainability.

Shared decision-making is much more than patient decision aids. We need to embed shared decision-making within clinical pathways and to demonstrate its value to both patients and clinicians. The Health Foundation projects are using shared decision-making with quality improvement methods to drive up patient activation, measure patient experience and improve quality.

I'm looking forward to my next visit already. Although I don't think we need to wait until then to make the three questions OK in every surgery and clinic in the land.

This article is adapted from Donal's blog. You can read the full post and download a copy of Making Shared Decision-Making a Reality at: http://bit.ly/qNMjnx

International PKD alliance is launched

Patient groups representing 12.5 million people with polycystic kidney disease from around the world have formed an international alliance, with the goal of enhancing the lives of everyone affected by PKD. The PKD Charity is pleased to be a founder member of the alliance and we're very excited about the potential of this international cooperation.

PKD International is a non-profit, non-governmental association, based in Geneva, Switzerland. The alliance aims to unite support groups, researchers and medical professionals worldwide in the mission to find a cure for PKD.

The association is planning to provide the latest news on PKD, and to support the most promising research and clinical trials.

For more information, please visit www.pkdinternational.org.

Ciliopathy Alliance

PKD is one of several conditions, known as ciliopathies, in which the cilia, microscopic, hair-like structures on the surface of cells, are defective or dysfunctional. So far 20 ciliopathies have been identified, affecting nearly every organ in the body. In PKD, kidney cells are affected but cystic kidneys develop in other ciliopathies such as Bardet-Biedl.

The PKD Charity has become a founder member of the Ciliopathy Alliance, a collaboration of patient groups, researchers and clinicians, working together to promote research into ciliopathies and raise awareness. Find out more about cilia and the ciliopathies at www ciliopathyalliance.org.

Help on hand for balanced, PKD-safe ready-meals

Dietary advice for those affected by PKD differs depending on weight, blood tests and, where applicable, dialysis choice. While there is no such thing as a 'PKD Diet', we recommend you maintain an ideal body weight, exercise regularly, eat a balanced range of foods and pay particular attention to high levels of salt and fat in food. The information below is for general guidance only – always seek individual advice from a renal dietitian.

Most people know that too much salt (sodium) is bad for blood pressure and fluid retention. It can also increase thirst and make fluid limits hard to stick to if you're on dialysis. Some golden rules are given in the article on the right.

Blood potassium levels may also be high in severe renal failure, on dialysis, or if taking blood pressure medications called ARBs (angiotensin receptor blockers). Bodies need potassium, however, so only restrict your intake if tests show it's necessary. Beware of products labelled 'low sodium' as these are often high in potassium, added to retain the 'salty' taste. This site, created by a PKD patient, lists products to avoid: www.potassiumwatch.org.uk.

High phosphate levels can be another problem in the later stages of renal failure. Again, though, not everyone is affected, so if blood tests show you don't need to cut down on phosphate – don't.

Managing his diet is something that PKD sufferer Ray Winger understands all too well. Despite his lifetime's experience in the food industry, Ray struggled for four years. He told us, 'malnutrition is common among those with PKD because maintaining protein and calories on the ideal diet is hard. Cooking from scratch is tiresome – especially with the tiredness associated with PKD – and the food choices often seem so limited that meals became bland and repetitive'. Faced with this challenge, Ray decided to put his food technology skills to good use by developing a range of ready-meals that would both meet dietitian recommendations and make life easier.

Ray explained, 'the reason nobody has developed specific kidney-friendly meals before is that it's technically quite difficult to present foods meeting recommended levels of salt, potassium and phosphate that still taste good. However, I was convinced I could overcome this.' From this determination Ray's range of ready-meals and sauces, Cuisine For Me®, was born.

A special logo clearly identifies the purpose of the range as Ray wanted to make life easier for people, having spent hours reading packaging himself. Similarly, the food is quick and easy to prepare and, as it doesn't need to be stored in the fridge, is perfect for travelling or visits to friends.

"Ray decided to put his food technology skills to good use by developing a range of ready-meals that would both meet dietitian recommendations and make life easier"

It's inspiring to hear from someone who has used their skills and experience to help others with PKD. We've invited Ray to the next PKD information day at Leicester Royal Infirmary on 26th November to talk about his experiences and Cuisine For Me®.

Visit www.CuisineForMe.co.uk for more information.

Why reduce salt?

Maintaining low blood pressure is one of the most important ways someone with PKD can manage their condition and eating less salt is the quickest way to do this.

How much salt is safe?

Adults should eat no more than 6g of salt a day, roughly a level teaspoon. However, only 20% of the salt we eat is usually added to food at the table or during home-cooking, while 80% is hidden in prepared foods, like bread, biscuits, cereals, sauces, take-aways and ready meals.

How to reduce your salt intake

- Stop using salt at the table and don't add salt when cooking. Also, avoid soy sauce, curry powders and stock cubes, all of which contain high levels of salt
- Know the difference between sodium and salt (sodium is one of the chemicals in salt - 1g of sodium is the same as 2.5g of salt) and check ingredient labels
- When buying prepared food choose those with 0.3g salt (0.1g sodium) or less per 100g of food. Pick those with 0.3-1.5g salt (0.1-0.6g sodium) per 100g only occasionally
- Never buy foods with 1.5g salt or more per 100g or with 0.6g sodium or more per 100g
- If ingredients' amounts aren't given on the packet, see how near the top of the list salt appears. The higher it is on the list, the more salt it contains
- When eating out, ask the restaurant to reduce the amount of salt in your food, if they can

How to spice up your food without salt

- Use fresh herbs, zingy lemon or lime juice, fresh ginger, chilli and ground spices
- Get hold of a low-salt cookbook and experiment with new tastes - there are a number available

If your food tastes bland initially, don't worry. Your taste buds will adapt.

Laura's story

Headteacher Michael Kelly, head girl Rose Graham, Luke Green and Laura Marshall

Anyone with experience of PKD will remember how frightening it can be: the first diagnosis – or misdiagnosis – and the emotional rollercoaster that follows. Laura Marshall was just 13 years old when her father was rushed to hospital in excruciating pain. Below, in moving words taken from a fundraising speech she gave in front of 2000 people at her school, she tells us her inspiring story.

'Good afternoon, everyone. There's one thing I never thought I would say to a school hall of 2000 people: size matters.

'In the days after my father was taken to hospital there were blood tests and CT and MRI scans. Finally the diagnosis came back: PKD.

'I was told everything would be OK. However, that didn't stop me from becoming tearful every single day. It didn't stop me from asking myself, ''what if?". It didn't stop the impact on my family's daily life - it felt to us like the world had ended.

'I picked up a leaflet which said, "Imagine your kidneys grow to the size of rugby balls. They will fill your abdomen, crushing down and restricting the function of other organs. You will experience crippling pain, may struggle to breathe, walk or even stand up straight. Eventually they may fail completely. Without dialysis you will die. Your only hope is a transplant, if only it were that easy. Or a cure, if only there were one."

'I was devastated and thought there was no hope.

'Tests and my father's declining kidney continued for 18 months. I had to sit my GCSEs with all this in the back of my mind and to find the inner strength to keep my younger siblings from being upset.

'However, the worst part is there's no cure, no prevention. It's genetic. Children of those with the dominant form of PKD, like me, have a 50% chance of inheriting it.

'Sounds awful doesn't it? But, there has been a major upturn as in November 2010 my dad received a new kidney. Not only was he given a new kidney from a live donor, that donor was my mum. The fact that my dad's still here is all thanks to her.

'And, although PKD may not be curable, it CAN be coped with.'

The non-school-uniform day Laura organised and the parachute jump she made with her mum (a feat she claims was less scary than giving her speech in front of everyone at her school!), together raised £2,242.55.

A dream fulfilled

Before she died, Kristin MacEwan, who had PKD and was on dialysis for eight years, wanted to create an anthology of all her poetry and to see it printed. Her daughter Katriona MacEwan said, 'It has been my honour to fulfil this dream for her.' Entitled "This is the Morning of my Life - a Garland of Poems", the book will be launched on 29 October 2011 and all profits from sales are being donated to the PKD Charity. To order a copy, email Katriona: kmacewan@btinternet.com.

Maddie's story

Bethany Moore's five-year-old daughter, Madison, has ARPKD. Here, Beth tells us her story.

'Maddie was born at 35 weeks after three days' labour. She was blue (from collapsed lungs) and was taken from my arms after 30 seconds to be placed in an incubator. After five days, I was told she had ARPKD and I should make "appropriate arrangements" (i.e. christening and registering her).

'But Maddie kept fighting and, six very long months later, I was finally allowed to bring her home.

'At ten months she developed pneumonia and I was told she wouldn't make it through the night. A night that happened to be my birthday. Again, though, Maddie refused to give in. As the weeks and months went on she was developing a character you couldn't help but fall in love with: funny, independent, strong and yet unbelievably loving.

'Maddie's five now. She still has to go to hospital whenever she gets a cold, but the only thing she's scared of is the blood tests she has to have every four weeks.

'Sadly the disease has spread into her liver and she'll soon be on dialysis and will need a transplant. But, because of her reduced life expectancy, every minute is precious.

'Maddie really does bring every moment alive. She's such a happy-go-lucky little girl, despite everything.'

Over the summer, Beth and Maddie organised a sponsored walk to raise money for research into ARPKD. Unfortunately it had to be cancelled when Maddie became unwell, but they hope to rearrange it next year.

Sporting success

In April 2011, the PKD Charity was delighted to have not just one but two places in the prestigious London Marathon. Owing to a huge amount of interest for these places, we held a ballot for all interested runners and the trustees picked out two to run on behalf of the charity. The lucky winners were Simon Greenwood and Delyth Wanklin.

Delyth, her two brothers and mother are all affected by PKD but after lots of hard training and fundraising, Delyth completed her London Marathon in 4 hours, 45 minutes and raised over £1,300. Simon raised more than £2,255, completing the event in 3 hours, 32 minutes and he's planning to run the gruelling Marathon des Sables for us next.

Our sincere thanks to Delyth and Simon. If you would like to have the charity's 2012 London Marathon place, please email esther@pkdcharity.org.uk before 1 November.

May 2011 saw a number of PKD runners take part in the Great Manchester run, raising more than £600. The runners were the first to wear the new PKD running vest, and in the same lime green as the PKD logo, this ensured that the whole of Manchester saw them! Our thanks to Katie Phenix, Sarah Calin, Rebekah Garcha, Lena Murphy, Rebecca Murphy and Graham Johnston.

Delyth Wanklin


Simon Greenwood

Thank you to these amazing PKD fundraisers

In July, Andy Sykes took on the White Peak Walk, covering 26 miles in 12 hours, and we had our first ever PKD skydive: Mel Brakes and Rachel Green raised £272. Jane Bird became our third skydiver, making her jump in August. Tim Self's sponsored run raised £385; Tracey Firth asked for donations instead of gifts for her birthday and Andrew Hursthouse completed the Sheffield Half Marathon. Otsuka Pharmaceuticals, whose staff from across the UK organised a series of sponsored walks, raised more than £1,200. In September, John Mason and friends took on a 10km run, 20km cycle ride and a 5km kayak through the Cotswolds, while Lee Fitzsimmons took part in the Tatton Park Triathlon. Peter Harris, who has previously cycled the length of Wales for PKD, went further, cycling from Lands End to John O'Groats! Completing the trip in just 21 days and one hour, Peter raised more than £1,200. Joseph and Melanie Arazi organised a fabulous Magic Night with Nicholas Einhorn raising more than £7,000. Vanessa Russell and six friends raised over £300 through their Three Peaks Expedition: Russell Middleton and Peter Robinson completed this extraordinary feat!

Thank you to everyone who has supported the charity in 2011.

Correction: In the last issue, we mistakenly published a picture of Emma Peach with the caption Katie Peach and would like to point out that she completed the Robin Hood Marathon and not Half Marathon as listed.


Mel Brakes

Can you spare some time?

We need volunteers with enthusiasm for writing and interest in the internet for the following:

Planning and supervising the twice-yearly PKD Newsletter, working with Tess and the design agency.

Posting updates on the PKD Charity Facebook Page - to support our fundraisers and generate more awareness

Contact
tess@pkdcharity.org.uk
or ring 0300 111 1234

Left: More than £4,500 has been raised for the PKD Charity in memory of baby Harrison Caines, who was "already at peace" when born. His parents Katie and Adam have been touched by the fundraising support they have received, including a summer fete (£1,788) and a football match (£1,341).
More: www.justgiving.com/katie-caines