Vol 2, Issue 1 Summer 2004

Polycystic Kidney Disease
The PKD Charity
.....together the answer

newsletter

PKD Charity PO Box 141 Bishop Auckland County Durham DL14 6ZD

www.pkdcharity.co.uk

Registered Charity No. 1085662

Editor's intro

Welcome to the summer '04 issue of our newsletter. I'm pleased to say that the charity is going from strength to strength. The fundraising efforts of our supporters have been enormously helpful and once again the London Marathon runners have done us proud. You will also see that we have organised the first UK PKD patient information day to take place on 18th September. This promises both to be a most informative day plus a great opportunity to meet up with other members of our charity so I think it should be well worth attending.

We've already had a wonderful response to our first newsletter and it's encouraging to see our membership is increasing already. It's good to hear from patients around the UK and we'd really appreciate any comments or suggestions for the newsletter and website development. So please keep those letters coming in, whether it be to write an article for our next issue or whether it's simply to keep in touch.

Best wishes to you all and thank you for your continued support.

Pam Hooley

Patient Information Day 2004

We are holding the first UK PKD Patient Day on Sept 18th 2004 (see later). At the event you can receive information about the disease from a panel of experts, information that you may have found difficult to find from other sources. Listen to the latest research into the development of a potential drug treatment. Finally, attending the meeting will give you a chance to share your experiences of living with PKD with others similarly affected. If Cambridge seems too far to come for a one-day meeting then why not make a weekend of your visit and see the sites of the ancient city. We look forward to meeting you, hopefully on a sunny day in September!

The Babraham Institute, Cambridge

Mission Statement

We aim to support families who live with Polycystic Kidney Disease, and to provide information and advice to them and their healthcare professionals. We will fund research to provide better treatments and continue to search for a cure.

As a charity we also want to raise awareness of PKD, provide information about PKD to patients, the public, the medical community and the media.

We want your input

Please send in any articles, letters, photographs & jokes. Make your contribution to the success of this newsletter

Why we set up a UK group

When I was first diagnosed with PKD I began the long search for any information which might be helpful, both to me personally and for family and friends who were doing their best to be supportive. I had imagined that, since this is a relatively common disorder, information would be readily available and that I would come away from the outpatients' department armed with fact sheets and leaflets. It soon became apparent that this was not the case and that good sources of information were actually very thin on the ground. Not only was there an almost total lack of literature on the subject, but there were no local support groups or even national groups who could provide me with the information I was seeking. Although it would have helped enormously to be able to speak to other patients, it seemed that there was no easy way of making contact with them.

When I did an internet search I discovered that the only organisation worldwide that could really help me was based in Kansas, USA! The PKD Foundation's website was very impressive, both in terms of the information provided and the emphasis on bringing people together both to share information and experiences and to raise funds for research into PKD. There were groups, or chapters, in many states and even in France and Japan. Disappointingly, though, there was no UK chapter.

Nonetheless, encouraged by the work of the Foundation, I decided to make contact with Dan Larson, their president. I also contacted several of their group co-ordinators, for further information on how to set up a UK group. At the same time, through a mutual friend, I made contact with Dr Anand Saggar, Consultant in Clinical Genetics at St George's Hospital, London, and discovered that he was also very interested in establishing a UK PKD charity. Things were coming together.

After discussing the set up of a UK Charity with Anand and learning as much as we could from the PKD Foundation, we decided that the UK organisation really needed to be more than a "Friends" chapter. Patients and families in the UK needed more local support plus advice on matters which are specific to the UK (e.g. NHS-related, insurance and mortgage availability etc). Also, we felt that it was important to raise funds for the many worthy research projects here in the UK which would otherwise remain unfunded. After much deliberation we decided that the best approach would be to form a separate charity which would be affiliated with the Foundation. That way we could benefit from belonging to the US organisation whilst still remaining firmly rooted in the UK. We could fulfill our aims of raising awareness and providing support in this country whilst at the same time learning from the Foundation and working together to ensure that our research efforts were coordinated.

Now that we're up and running as a charity, we feel that this approach gives us the power of working as part of a much larger team, whilst still remaining flexible enough to respond to local needs and concerns. I suppose that it all depends upon striking a balance between uniting in a global battle against PKD and translating that into local action. We want to be part of a global effort to raise funds for research as we know that this is of paramount importance to all PKD patients. But our activities need to reflect the needs of patients in this country too. And by that I mean that I'm hoping that with our new charity we in the UK can join together, find our voice and start making a difference.

Pam Hooley

Knowledge is power

Would you like to know more about PKD?

We have an information booklet for patients and their families on Autosomal Dominant Polycystic Kidney Disease.

If you would like a copy of this booklet, write to the address below and consider sending a small donation to cover P+P.

Please send me a booklet
PKD Charity
PO Box 141
Bishop Auckland
County Durham DL14 6ZD

.....Also

In our last newsletter we promised a list of beneficiaries of your support. Thank you for your generosity. We have a new website developer and we are expanding the newsletter. Significant funds have been allocated to the Patient Awareness Day on Sept. 18th 2004. In addition, we are also planning to sponsor a major scientific meeting in London focusing on the new potential drug treatment for PKD in conjunction with the PKDF and the Novartis Foundation next year. Medical research is extremely expensive, for example a typical 3 year scientific research project currently costs in excess of £200k. We need your help to reach such a goal!

Your letters

I am in my 69th year and discovered that I had PKD some 40 years ago. I have been fortunate. Apart from infections I have been well and able to cope with 3 children and a teaching career. I also had the good fortune to be in the hands of a medical team with a particular interest in PKD.

Do we have a role as patients? What can we do for the newly established charity? What can it do for us? Obviously we must cooperate with the medical profession over on-going research. The NHS cannot fund every branch of research so the charity has an obvious duty to raise funds. The scope for this is endless. There is also need for support groups. It is good for the morale to take a positive and active interest and to share problems presented by other patients. The newsletter can provide that link. It helps to keep things in perspective (as I have learnt from the Parkinson's Disease Society since I was diagnosed 10 years ago).

It is not always easy to be optimistic but if one compares the medicine of today with the limitations of pre-NHS in this country then one can see how things have moved forward. Human beings are very resilient and cope with illness with remarkable, and surprising, fortitude. For those who have a faith there is an added spiritual dimension and the promise of hope.

J. Kitteringham, Warwickshire

In the news

PKD charity demands action on UK and European research spending

PKD is one of the most common life-threatening genetic diseases, far more common than better known conditions such as cystic fibrosis and multiple sclerosis. We estimate there are at least 60 000 individuals affected in the UK. In comparison to the US lead, scientific research into PKD in Europe is massively under-funded particularly in the UK. The Medical Research Council (MRC) is a government-funded agency with the remit to fund basic medical and clinical research in this country. They spend money that we pay in taxation. In the last set of figures from the MRC (2001-2002) there was a ZERO spend on basic research into PKD by our research council. This contrasts with millions of dollars spent every year by the US government who have a strategic plan to tackle the disease through their relevant research agency, the NIDDK.

We made this point very clearly to the world-wide scientific community in Nature Magazine, the most famous and widely-read scientific journal in the world, in a Headline Correspondence (dated 12th February 2004 Volume 427 P584). If you would like a copy of the letter please contact our Editor.

So where are the opportunities for British scientists to work on PKD?

Flora London Marathon 2004

Once again the PKD charity had a courageous and heroic bunch of runners raising funds on a wet and windy day in the capital. Thanks to you all and we hope to feature more of you in future newsletters.

Martin Bootman raised £1500 for PKD running the 2004 London Marathon and is shown here in training with his twin daughters. Martin is also researching the cause of PKD at The Babraham Institute.

An article by Jane Pugh in The Times supplement (Times 2)

Jane Pugh is a freelance writer with PKD who wrote a very personal account of living with PKD (Times 2, June 16th 2004). We did not see the story in advance of publication but Jane asked us to provide our contact address for readers who wanted further information. **This did not mean we endorsed Jane's individual view.**

The headline suggested that PKD kills 75 per cent of sufferers by their mid-forties **and was factually incorrect**, as was the same statement later in the article. On average PKD causes end-stage renal failure between the ages of 55 – 60 years. This requires management by kidney dialysis and can only be cured by transplantation. It is not the death sentence suggested by The Times and careful dialysis management can work for several decades. The risk of a ruptured aneurysm in PKD, usually of a type called a sub-arachnoid haemorrhage on the surface of the brain, is slightly increased compared to normal individuals and can have serious consequences depending on the severity of the incident. There is evidence that the risk is higher for PKD families with a history of this problem.

The best way to avoid such complications is to carefully control blood pressure and lead a healthy lifestyle.

Please visit our website or request our information booklet for more details.

We should point out that although Jane's article has upset some patients it has also been of value in highlighting PKD to the general public and media. Although we value your views on this account of PKD we suggest that specific complaints should be directed to The Times on this occasion.

June 16, 2004
Times 2, Cover story

Yes, life can begin at 40: even though I may be dead at 45

Twenty years ago Jane Pugh was told she had an incurable kidney disease that kills 75 per cent of sufferers by their mid-forties. She describes how she has fought the inevitable with energy, courage and humour

I'VE ALWAYS FELT that women who lie about their age are sad. What's the point? Surely claiming to be 35 when you're 55 just draws attention to the thing you were hoping to avoid.

I was going to be different. If asked, I would be honest; in fact, so at one with my advancing years that I wouldn't wait to be asked. I'd promote it; wear T-shirts with my age printed boldly on the front.

Who was I kidding? Hitting 40 for me had a greater significance. I wasn't going to pull this one off easily. It went beyond worrying about wobbly underarms, bums and boobs. Since the age of 20 I had known this day would come. When my GP casually announced "both kidneys are affected", I knew exactly what he meant. I ran home crying. I had polycystic kidney disease (PKD) — the same genetic condition that killed my grandmother and father, both aged 45. This was no fluke. Seventy-five per cent of those with PKD suffer renal failure in their mid-forties.

My eldest sister had it too. We chose different ways of dealing with it. Outwardly she was nonchalant. She devoted herself to toxic cleansing. Out went meat, white flour, in fact all forms of easily recognisable foods. In came hummus and coffee enemas (decaf, I think). She rejected conventional medicine. Doctors, she felt were "all stupid". She was going to beat it herself.

I smoked cigarettes and marijuana, got pissed, ate steak and kidney pies and partied all night long. That's what artistic people do, I argued; a form of expression. I did try colonic irrigation. Once. Performed by my sister, who'd taken a Colonic Irrigation Made Easy course. Except it wasn't easy. Our relationship was never the same after that.

The specialist didn't hold back. I was at risk of high blood pressure, heart disease, bowel problems and then, if none of those got me, renal failure. Whenever we met, he reminded me of the research being conducted into pig organ transplantation. Not long after this discovery I dropped out of university and opened a shop specialising in pig themed gifts. His advice was: "Get on with life." And so the countdown began.

Like my father, there was a sense of being on borrowed time. I was impulsive, reckless even. I leapt with enthusiasm from one "great idea" to the next. Having shelved my ambitions of becoming a commercial artist, and with the pig shop up and running, I seized the opportunity to expand by buying a failing health centre called the Hollywood Leisure Suite. I discovered on the day I took over, after a breathless phone caller asked for "Chantelle", that I had taken over a brothel. I found a bullwhip under the massage couch. I set about holding aerobic displays in the car park, to show that we were now legitimate.

After declaring myself bankrupt, I spent a good six months in my attic making jewellery and smoking. Now 25 and 20 years away from dying, I figured I had time to get healthy.

Later I climbed the corporate ladder. High. Then jumped off. With a baby to worry about, the pressure mounted. He would only be 13, when I was 45 (the age I was going to die). I gave up my secure, well-paid, glamorous job and became freelance, to spend more time with my boy.

At about 35 I noticed that I started to spend a lot of time with the mirror, chin up, stretching my neck really tight. The neck isn't something we can hide easily. And I wasn't prepared for chiffon scarves yet. The prospect of developing a goosy neck plagued me. I took to applying huge dollops of moisturiser, massaged upwards like the beauty experts tell us to do; though the urge to splat it on in any old direction eventually wins.

I was still 10 years away from dying. And there are bound to be big developments in pig organ transplantation in that time.

At 38 I developed tingling around my mouth and tongue. Doctors diagnosed a virus. Friends diagnosed that I talked too much. An MRI scan diagnosed three aneurysms. One of the "risks" the kidney specialist forgot to mention. I needed brain surgery. This involved making a seven-inch incision around my hairline, cutting a "window" in my skull and rummaging around in my brain to fit a clothes peg type clip around the neck of the aneurysms. Result? Two dead aneurysms. One was untreatable.

I was zombied for several months — recovery was slow. Having given up a freelance contract for the surgery, I lacked the energy to win new work. With my driving licence revoked because of postoperative seizures, I got in touch with my legs. I wouldn't be beaten. I perfected the art of cycling while carrying six bags of heavy shopping. Finally came the realisation that I had impaired sexual feeling. I had brain damage. Life changed.

OK, so now I have kidney disease, an aneurysm that could rupture at any moment and kill me, a dysfunctional clitoris, a crater in my head, a fear of my own neck, no career and soon I am going to be 40. Five years away from dying.

As my fortieth approached, friends and family inquired: "So, what are we doing for your birthday?" "Oh, I don't want any fuss," I'd say limply. They didn't buy it. "So are we having a party, then?" I put my foot down. I didn't want a birthday, and that was that. For me, life didn't begin at 40. This was the beginning of the end. So I pretended it wasn't happening; and started lying. If asked my age, with chin up and neck stretched tight, I would say "35."

I became bitter. Self-pitying. Depressed. Nobody loved me. Especially me. I was a failure. Why did I drop out of university? I could've been famous by now. My body is broken — ill, sexually inadequate. Whatever happened to that bouncy dynamo that I used to be? "Why don't we go to New York?" they persisted. Why don't you all f**k off, I thought, but didn't say. My boyfriend probed. He knew I'd always wanted an adventure 40th — abseiling, quad biking, gorge scrambling; getting dirty and being silly. "No, no, my aneurysm might rupture," I reminded him. "Just leave it alone." I sulked. Secretly I hoped he would plan something fab.

The day my birthday arrived I awoke full of evil. I lay in bed, with my neck smothered in Nivea, cursing everyone. I planned revenge on those who had wronged me. My best friend Flo called to ask if I wanted to drink champagne in our favourite bar. My sister called for a chat. I snarled at them.

Jollying me along, my boyfriend suggested we take the children (his two, and my eight-year-old — whom I haven't told yet) to look for a new fire surround. Whoopee. As an avid DIYer it's usual for me to get excited about a trip to B&Q, but today I shuffled pathetically around muttering expletives to myself. Back home, the children were getting on my nerves. I shouted extra loud instructions as we entered the house. Stomping through the door, I was deafened by the sudden noise.

Around 40 people were in my kitchen wearing big silly triumphant grins. I ran out. Emotional. Like celebs do on This Is Your Life. My boyfriend had bought the food earlier and given it to my sister and best friend to prepare while we were fireplace hunting. They'd all spent a good hour curled up on the kitchen floor giggling. The champagne flowed. I got drunk. It was a great day.

One month later something changed. Despite my sister's careful lifestyle we learnt that she has renal failure. Three years older and therefore closer to 45, she's always been my measure. It came as a shock, even to her. She's soon off to Thailand for intensive detoxing — where coffee enemas are a way of life. I believe her optimism will see her through.

Although contrasting events, my sister's ill-health and the surprise party have reinforced my sense that you never know what's round the corner. Now, I focus on the good things: my amazing son, the many great people around me, my beautiful home, my wrinkle-free neck. I am lucky. I have started writing — an earlier passion, shelved. Taken up yoga. The adventure day planned for the summer. A trip to New York. I think, even if I am going to die at 45, that's five years of living left to do.

Besides, they're now successfully transplanting pig organs into baboons. Snort, snort.

The polycystic kidney disease charity is at www.pkdcharity.co.uk, or call 01388 665004

New Scientist

'Hope for inherited kidney disease'

New Scientist magazine ran a story on March 6th 2004 highlighting the progress of a new drug that halts the growth of kidney cysts in rodent models of PKD. Dr Richard Sandford, University of Cambridge, offered his views of the research. You can hear more about the drug, and listen to Dr Sandford talk about PKD, at the Patient Meeting in September.

The PKD Charity Contact Information

Please contact Pam Hooley, The PKD Charity Chairman
Tel: 01388 665004
Email: Pam@search62.freeserve.co.uk
or write to
The PKD Charity
PO Box 141
Bishop Auckland
County Durham
DL14 6ZD