

Water therapy a breakthrough for PKD?



It's a well-known fact that humans are more than 50% water and that water is essential for life. Kidney specialists spend most of their time thinking about urine, which is mostly water. In PKD, ill health is a result of much of the normal kidney structure being replaced by cysts that are filled with water. Now we are confronted by the bizarre prospect that despite these water-filled cysts bloating the kidneys, water may be the 'cure' for this disease.

It is known that a certain molecule known as cAMP is involved in the enlargement of these cysts. Various studies on rodents have also shown that another molecule, called vasopressin, increases levels of cAMP. When mice with PKD were given a chemical that blocks vasopressin, there was an impressive decrease in kidney size and some preservation of kidney function. In the

same way, when mice had excessive consumption of water, (which decreases levels of vasopressin) a similar result was seen.

Subsequent research has supported this observation, and it has been noted that specific kidney cells are genetically predisposed to form cysts when exposed to cAMP and vasopressin. Vasopressin is important because the physiology of terrestrial animals must conserve water to survive – vasopressin levels in humans are usually consistently high enough to activate cAMP and as a result reduce the volume of urine and retain water.

Humans do not always mimic rodents and we won't know whether vasopressin inhibitors, such as water, work until clinical trials have been conducted.

Until then, nephrologists must grapple with a question that informed patients will undoubtedly ask: How

much water should I drink now? No one can give a definitive answer to that question, but common sense leads us to think that sufficient water should be drunk to keep these blood vasopressin levels down.

To know for certain how much of the 'water cure' is prudent therapy, a carefully controlled trial seems justified. And, with the assistance of more advanced molecular-based research strategies, we should expect to move much faster towards other highly targeted therapies.

This is why we are looking at ways of raising the money that is needed to help fund the groundbreaking water therapy research and trials. In the meantime, we shall be keeping a close eye on the subject and keep you updated of further findings.

With thanks to Neil Coutinho

In this issue



As chairman of the PKD Charity, I'd like to welcome you to the latest issue of our newsletter.

This time we have heartening news about water therapy; information and stories about kidney transplants; an excerpt from an online journal by a man on dialysis; plus tips on pain management, and pregnancy and PKD.

Get involved!

If you have any feedback or contributions please email me at tess@pkdcharity.org.uk

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Pregnancy and PKD



Following the success of Dr Richard Sandford's workshop at the PKD Information Day last summer, we thought we'd provide a brief introduction to the topic by answering a few commonly asked questions below. We'd also love to hear from any of you who'd like to share your experiences with readers in future issues!

Should I consider pregnancy if I have PKD?

The good news is that the majority of women with PKD (the commonly-cited figure is 80%) who become pregnant have successful pregnancies with no major complications. However, if you are considering having children, it is a good idea to discuss things with your GP first. They will be able to suggest whether pregnancy is advisable, and if necessary adjust your medicines. Also, as PKD is an inherited disease, you may be concerned about passing

the disease on to your children. If this is the case you should speak to a genetic counsellor before you become pregnant – your GP can arrange this for you.

Is there anything that increases the risk of complications, and what are these likely to be?

Although most women will not face any major complications, there will be some who will have an increased risk of serious complications – for both them and the baby. For example, uncontrolled high blood pressure at the start of pregnancy increases the risk of miscarriage. High blood pressure also increases the risk of pre-eclampsia, which in severe cases can be life-threatening for both mother and baby. Decreased kidney function can also lead to complications during pregnancy if the extra pressure they are under causes further damage. Women who expect to need dialysis in the future may find that pregnancy brings this forward – although it is very unusual to need to start dialysis during pregnancy. Basically the worse your kidney function, the higher your blood pressure, and the more protein in your urine, the higher the risk.

Is there anything I can do to reduce these risks?

Because of the possibility of complications it is vital that you are closely monitored during pregnancy

– in particular your blood pressure (pre-eclampsia can develop suddenly) and kidney function. Also, the strain placed on your kidneys increases with age, so older mothers should be particularly aware. Generally you should do your best to follow a healthy lifestyle – see your GP or Practise Nurse for tips.

Does the risk increase with each pregnancy?

Some research does suggest that the risk increases with the number of pregnancies – one study from the University of Colorado suggested that three or more pregnancies had a detrimental effect on kidney function, although this effect seemed to be confined to women with hypertension.

Should I stop taking my medicines?

Definitely not. The best way to keep your baby healthy is to keep healthy yourself. That's why it's important to see your GP regularly – they may need to change your medication once you are pregnant.

What will happen during my pregnancy? Will I need to see anyone special?

You will remain under the care of your GP and your usual Nephrology team, who will arrange for you to see specialist obstetricians.

You will find more about PKD and pregnancy on our website

Coping with pain: self-help information

Last issue we looked at managing pain with medications; this time it's the turn of natural methods. Ideally, though, we'd like to share readers' tips, so please do let us know your ideas for future issues.

Pain is felt when signals pass from nerve to nerve by the release of tiny amounts of chemicals called neurotransmitters. While some neurotransmitters intensify pain, others can reduce its intensity or even block it altogether. Some drugs can encourage these 'good' neurotransmitters but, fascinatingly, a positive attitude, exercise or simply laughing can also play a part.

Most people notice how pain is affected by their circumstances, and that being tired or stressed can make it even more unbearable. However, as living with pain can be both tiring

and depressing, this often creates a vicious circle – one that pain clinics and self-help treatments aim to break.

The British Pain Society, www.britishpainsociety.org, in particular is very helpful, providing general information, details of self-help groups and pain clinics, as well as publications such as 'Understanding and Managing Pain'. The Expert Patient Programme too is useful, running a self-management 'Persistent Pain Programme': www.expertpatients.co.uk.

As mentioned above, exercise can help – both improving your general health and directly blocking pain signals. Basically the better you understand yourself, the better equipped you are to manage your pain. What do you find stressful or relaxing? Simple relaxation

or visualisation techniques and breathing exercises really can make a difference, and there are many tapes or courses available to start you off.

Similarly many people living with pain find aromatherapy, homoeopathy, acupuncture (which is sometimes available on the NHS), massage or yoga helpful. Although do check that any potential therapist is registered with a reputable professional body – the Institute for Complementary Medicine provide a practitioner search facility on their website: www.i-c-m.org.uk.

Above all, remember The British Pain Society's words, 'The best pain control technique is probably your own determination that you will not allow it to take over your life. Pain may be part of your life, but it is not all of it. Life is still for living.'

Paired kidney donation – sharing the love

Living kidney donors who are incompatible can still help

Every year more and more people receive kidneys from living donors. Now, following changes to the systems for organ donations, the number is likely to grow.

When someone becomes sick due to kidney disease, it's not unusual for a family member to take the brave decision to donate one of their kidneys. However, more than half find their blood or tissue type make them incompatible with their loved-one, meaning they cannot go ahead.

'It is now legally possible to approve transplants between strangers'

Until recently this was the disappointing end of the story. By law, living donors were only allowed to give an organ to a genetic relative, or someone with whom they had a very close personal relationship. If their blood or tissue type was incompatible with this person, that was that.

But in 2004 a new Human Tissue Act was passed and, in 2006, the newly established Human Tissue Authority (HTA) set in place systems that were far more flexible. As the

HTA has created much tighter processes for making certain a donor understands what they are doing and are giving their consent freely, it is now legally possible to approve transplants between strangers.

As a result, incompatible donors and recipients can now swap kidneys with another pair of donors and recipients who are in a similar position – a process known as paired or pooled donation.

Additional good news is that living donor transplants are more successful than those from deceased donors, and the results of transplants between unrelated people are just as successful as they are between relations.

Given the severe shortage of kidneys available for transplant, this new way of donating kidneys is incredible news for people with chronic kidney disease.

UK Transplant manages organ transplants in the UK, running the National Transplant Database and the Organ Donor Register and providing a 24-hour matching and allocation service. By comparing the tissue and blood types of incompatible couples, UK Transplant can hopefully find a match so that both couples in need of

a transplant can be paired together to receive a compatible organ.

'Six kidney transplants were carried out in one go'

The first paired donation transplant in the UK took place in October 2007 and involved two couples in Cambridgeshire and Edinburgh. A landmark event, both transplants went well and the couple from Cambridgeshire were discharged from hospital after only five days.

On 5 April 2008, another first for paired donor kidney transplant took place. In Baltimore in the USA, twelve people were involved in six kidney transplants all at the John Hopkins Hospital. This, the first time six kidney transplants were carried out in one go, was set in motion when five separate transplant candidates visited the hospital for evaluation, each with their own willing donor with blood or tissue types that were incompatible with theirs.

Find out more about paired kidney donation:
www.uktransplant.org.uk
www.paireddonation.org

Should the rules change?

Kidney allocations and the HTA got a lot of media coverage recently. A story hit the headlines of an angry 39-year-old woman, Rachel Leake, who was refused her daughter's kidney after the 18 year old tragically died from an asthma attack. Many commentators felt this was wrong.

The policies that guide the HTA in approving donations do not allow for anyone to claim an organ from a deceased person, no matter how closely related they are. The policies are set up to ensure everyone who receives a kidney is in the greatest need and is most compatible.

The chief executive of the HTA said, 'There are people for whom getting a kidney is a matter of life and death. If we go down the path of saying you can direct who gets your

kidney after death, what if that person is in not as urgent a need as someone on the top of the waiting list?'

What do you think? Do you think the HTA made the right decision? Or that the policies should be changed? Or do you agree with the HTA? How would you feel in the same situation? Send your views to tess@pkdcharity.org.uk.

You can see the HTA's explanation of their position on their website.

http://www.hta.gov.uk/newsroom/media_releases.cfm

Donors – use Gift Aid

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

PKD information day 'highly informative'

Over 70 people attended the latest PKD Patient Information Day at the Royal Hospital in Edinburgh.

The audience of people with PKD and family members heard presentations on the clinical and genetic aspects of PKD, and attended workshops ranging from lifestyle tips to coping with PKD.

Of particular interest was the presentation by Professor Neil Turner on how research is carried out in the UK and the future prospects for more research into PKD, its treatment and cure. Delegates found the day to be 'highly informative' and enjoyed the opportunity to meet other people with PKD and share experiences.

The next information day will be on 18th October 2008 in Exeter.

Life on dialysis – how Jeff Sheridan stays ‘chipper’



Jeff Sheridan blogging during his 100th dialysis session

Although it's the kidneys' job to clean waste and extra fluid from the blood, if they fail then the process can be done artificially using dialysis, either haemodialysis or peritoneal dialysis (CAPD or CCPD).

So, that's the theory, but what does this mean in practice? What are the effects likely to be – not only on your health but on your emotions? On your family? Or on your sex life?

The best way of answering these questions is to hear from people with firsthand experience, either by joining a local or national support group or accessing some of the online resources. The following is US-based but has a useful discussion forum where you can compare experiences, and it should give you a good idea of what to expect, as well as useful tips for dialysis-friendly recipes or exercise.

www.davita.com/forum

There is also an excellent blog (online diary) by Jeff Sheridan, a 44-year-old American with ADPKD, which gives a day-by-day account of his life on haemodialysis. In it Jeff records the highs and the lows – or often just the monotony – of living with dialysis.

With Jeff's permission we've quoted the entries from his first days on dialysis to give you a flavour of what to expect and the emotions you might go through.

Day 1

I just got back from my first dialysis treatment. Other than some slight dizziness when I first stood up afterwards, everything went smoothly. My first treatment was only two hours, and the next one on Tuesday will be three hours.

Day 2

I was surprised at how much fluid they took off today... I drove home myself without any problems, but took a nap when I got home. Overall, dialysis was tolerable. On Thursday, I get bumped up to a four-hour treatment.

Day 3 & 4

Emotionally I crashed...

'I'm blogging here while on dialysis. It's a little awkward, as I have to type with one hand'

Day 5

Content. I'm blogging here while on dialysis. It's a little awkward, as I have to type with one hand... Physically, I feel noticeably improved since starting dialysis. Although I was resistant to having to do it, I'm glad it turned out the way it did.

'Nothing went wrong, but I just had a big case of the blahs at dialysis today'

Day 22

Not very positive today. Nothing went wrong, but I just had a big case of the blahs at dialysis today. I'm not sure why, but I just didn't feel like being there; maybe it's the monotony, or seeing the other people getting dialyzed that look sick, I don't know.

Day 23

Mood: chipper. No more 'blahs', thankfully. I was ready to ask my doctor for some chemical help, but that won't be necessary at this point. It's well documented that dialysis patients have a high rate of depression, but I prefer to 'suck it up' and avoid meds if possible. However, if it gets to be a persistent/long-lasting problem, I won't hesitate to discuss with my doctor.

Thank you to Jeff Sheridan for giving us permission to republish these excerpts. Since writing this article we were delighted to hear that Jeff has received a transplant and is doing well. Read more at:
<http://jsher.livejournal.com>.

Why not follow Jeff's example and keep a diary? It doesn't have to be made public. You may find it helps you to keep track of your emotions and manage them. Or, if you do fancy starting your own blog or website, take a look at the following tools:

www.livejournal.com
www.blogger.com
www.squarespace.com

Want to support PKD research?

We set aside a percentage of donations to fund research into treatments and a cure for PKD but if you want to designate an entire donation for research—just let us know your wishes at the time of giving.

Getting the most from your appointments



It is well known that involving patients in decisions about their healthcare can have beneficial effects on its results. However, for patients to make informed decisions about their treatment, they must be able to access and understand all the necessary information.

Download the leaflet from the DoH website: www.dh.gov.uk/en/Publicationsandstatistics/Publications/PublicationsPolicyAndGuidance/DH_079531
Or, call the orderline on 08701 555 455 (8am – 6pm Monday to Friday). Give the reference: 279234 – *Questions to ask leaflet*.

In response to this, in Autumn 2007 the Department of Health published *Questions to Ask*, a two-sided leaflet that aims to ensure both you, and your doctor or other health professional, get the most out of your appointment.

Although designed as a general guide, it is a strong starting point that offers a straightforward approach to things to consider before, during and after your consultation. Questions are listed on one side and tips to get the most from your appointment on the other – some of which we have highlighted on this page.

Next issue – How to see the consultant you want

Before your appointment...

- 1 List all your medicines (including vitamins and supplements)
- 2 List your symptoms
- 3 Write down all the questions you want to ask
- 4 Consider taking a friend or relative with you

During your appointment...

- 1 What happens next and, importantly, when
- 2 If any further tests are required – if so, how you will get the results, who you should contact if you don't, and who will explain them to you
- 3 What treatment, if any, is best for you. Are there any side-effects?
- 4 Are there alternatives? What will happen if you decide against treatment?
- 5 Anything you can do to improve your situation – lifestyle changes such as exercise, food etc
- 6 Who to contact if you have further questions or problems. Is there written information available or a support group you can contact?

Tolvaptan trial recruitment now underway



Last issue we reported on the phase three trial of Tolvaptan and provided information for those who wanted to volunteer when recruitment began. We can now report that recruitment is underway.

If you have ADPKD, live in the UK and are aged between 18 and 50, you may be eligible to take part in the trial. Full eligibility criteria is given on the trial website: www.tempostudy.com.

To get involved, you can download a pre-screening form from the study website and this should then be sent to the hospital – a list is given on the website – that is most convenient to you.

If you have never been involved in a clinical trial before, there is a fair bit to get to grips with, however, it's worth knowing that if you sign up you will be free to pull out at any time. In our Spring 2007 newsletter, we outlined

Research using mice with PKD gave scientists reason to think Tolvaptan could halt or reverse the progression of cyst growth. The TEMPO trial of Tolvaptan is the first of its kind for a treatment for PKD and looks very promising

16 questions to ask before joining a trial. You could use these questions when you next talk to your consultant or GP, or you could contact a participating hospital and run through the questions with them. You can download the Spring newsletter from our website: www.pkdcharity.org.uk/pkdnews.htm.

Not everyone who is admitted to the trial will receive the drug, as one third will be given a placebo. However, as Dr Albert Ong, a senior researcher at the University of Sheffield specialising in PKD, said, 'I hope many patients will volunteer to take part as that is the only way to get a clear answer as to whether the drug will work more effectively than a placebo.'

More information visit: www.tempostudy.com

After your appointment...

- 1 Keep notes of what was discussed and what will happen next
- 2 Book any tests and note them in your diary

Are you an experienced fundraiser?

We are looking for someone for 20 hours a week to develop and manage the Charity's future fundraising. Contact Tess Harris:

tess@pkdcharity.org.uk

World Kidney Day 2008



Andy Williamson and band provided music at the event

World Kidney day takes place in March and, to mark it, the PKD Charity joined forces with BODYWORLDS4 to hold its first national Awareness Event, with an evening reception at the Manchester Museum of Science and Industry (MOSI) on 14 March.

In the words of Dr Donal O'Donoghue, the Clinical Director of UK Renal Services, the event celebrated both 'amazing kidneys and the amazing people and families who have adult polycystic kidney disease. Families with polycystic kidney disease are in a special position because of their experience of renal failure in the family, [and] the previously thought inexorable decline in kidney function'.

Other speakers at the event, which aimed to raise awareness of kidney disease and to provide information in a fun setting, were Tess Harris, chairman of the PKD Charity; Andy Williamson, who recently received a live kidney organ transplant; and Dr Anand Saggur, co-founder of the PKD Charity, who in a fascinating talk showed what a polycystic kidney looks like using a real diseased kidney from the BODYWORLDS4 exhibition.

The kidney had undergone plasmation, a process pioneered by Dr von Hagens to preserve the body and organs by extracting bodily fluids and soluble fat and replacing them with substances such as rubber, silicone and epoxy. Other normal, abnormal and diseased kidneys were also on show in the main exhibition, which continues until June. (For further information see: www.mosi.org.uk/whats-on/body-worlds-4)

Dr Saggur also discussed current research into the treatment of PKD, including the Tolvaptan TEMPO trial and a new study in which water was found to reduce cyst size in rats with PKD. The PKD Charity intends to raise funds in the UK to sponsor a

clinical trial with water in the near future (see page one).

Jazz saxophonist Andy Williamson, together with musicians Kathy Dyson and Steve Berry, provided the music at the event. In 2006 Andy was diagnosed with PKD and put on dialysis before fellow band member, organist Maff Potts, offered to become a living donor.

Andy and Maff underwent the transplant operations in August 2007 and three weeks later recorded their fundraising single *Live Life Then Give Life* under the name Big Buzzard and the Organ Grinders.

An event celebrating 'amazing kidneys and the amazing people and families who have adult polycystic kidney disease'

Andy commented 'People aren't really familiar with this condition but it is very common. Often it goes undiagnosed. I have grandparents who died young who we now believe may have had it. Maff was incredibly generous to donate his kidney and it was a very moving experience for both of us. We are both now very keen to get as many people as possible thinking about organ donation – to sign up to the national register and help save someone's life.'

Andy and Maff are also campaigning to improve public understanding about the issues involved in living kidney donation – to find out more go to:

www.haveoneofmine.com

Tess Harris thanked everyone concerned for the success of the World Kidney Day event, in particular MOSI and the exhibition organisers. 'We are delighted that BodyWorlds gave us the opportunity to generate awareness of PKD and increase understanding of what it might be like to have this potentially devastating condition'.

Remember us

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

Easy Fundraising website – a new definition of shopping therapy!

If you've always wanted to raise money for the PKD Charity but couldn't quite face the London Marathon, thanks to www.easyfundraising.org.uk you can! Now, every time you shop online, you can help raise much needed extra funds for us without it costing you a penny. Although you may wonder what the catch is, we promise there isn't one! In fact, as many retailers give discounts or special offers for online shoppers, you may even find you save yourself money at the same time.

All you have to do is register on the website, in a straightforward process that takes less than five minutes. You simply choose your nominated charity from an alphabetical list (we've made sure the PKD Charity is on there!), enter your name, email address and a password. And that's it – you're ready to go. It really is as simple as the click of a mouse!

You still shop directly with each outlet in the normal way – choosing from 500 of the best-known UK retailers – but by using the links from the Easy Fundraising site to enter your favourite online shops, up to 15% from each purchase you make is donated to your nominated charity. (The precise amount is displayed next to each retailer listing.) Easy Fundraising simply note your online visit to the retailer and then reward the charity of your choice – The PKD Charity we hope – if you decide to make a purchase.

The site explains, 'It seems very generous, but when you buy online the cost to the retailer is much lower. As there's no shop, salesperson, heating, electricity, rates, not even the cost of the bag to put your goods in, retailers are more than happy to help good causes when new online customers are introduced to them via our website.'

So, if you're already an online shopper, or have been meaning to take the plunge for some time, get clicking now. By splashing out on that new DVD, jumper or sofa you can raise funds for a cause close to your heart – and help create a whole new meaning for shopping therapy at the same time!