EYE DONATION: ROGER DYER'S STORY

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Human rights for people registering as organ donors

Sir—In February, 2000, my son, Roger, died peacefully, at home. He was aged 18 years and had muscular dystrophy, so his death was not unexpected but my family still grieves for the loss of our much-loved son.

The family had discussed registration as an organ donor with each of my three children when they had raised the subject, generally after a secondary school lesson. We had all decided to register and to carry donor cards and we were aware of each other's wishes. We assumed our registered intention had been accepted by society as our right to donate.

My son died at home with his mother, brothers, and myself present on a Sunday morning. The emergency family physician was there for a short time and certified the death. In the afternoon my son's body was taken, by the undertakers, to the chapel of rest. His own family physician did not contact us other than to pass on the signed death certificate.

My son knew that he would probably not be able to donate his organs but he was aware that tissue donation remained a possibility. In the event, none of the professionals involved raised the issue of tissue donation with my family.

Does registration as an organ and tissue donor carry with it the automatic right to become a donor, provided there are no clinical contraindications at the time of death? Are such human rights routinely ignored? Why has organ and tissue donation never been a routine part of the dying process in this country? Is my family's experience unusual—I expect not. Organ and tissue donation campaigns are widespread, yet our society is unable to realise the potential of registered donors. My experience highlights the urgent need for society to recognise the human rights of those of us who have joined the donor register.

In the event, my son's wishes (rights) were achieved, but only because I am professionally involved in organ and tissue transplant services. I was able to telephone a colleague, who is a consultant ophthalmic surgeon, and he realised my son's rights. This has been a reassurance to my family and has helped us to cope with our tragedy. We are proud of our son's achievements in his short life and also that after his death he was able to facilitate the restoration of the sight of two other young people.

It is my opinion that all professionals involved in dealing with the dying and the dead must seriously consider why they are participating so little in realising the human rights of registered people who wish to donate organs and tissue at the time of their death.

The UK National Transplant Week is July 7–13, within which, July 10 is National Donor Day.

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Roger Dyer died in February 2000, aged eighteen. He had previously joined the Organ Donors' Register, and after his death one of his corneas was transplanted to a 28-year old man, the other to an 8-year-old girl.

Roger's life

His father, Phil Dyer, told me about Roger's life.

"Roger was born in June 1981. He was our first child, so we were very excited and we didn't have a child already to compare him with. With hindsight we should have realised that he was weak from the outset - but he was our first child, and it had been a normal birth. The maternity services at that stage were typical of the time and we didn't receive much advice. His brother Martin was born in August 1982, so there were only 15 months between them; and then it became fairly obvious that Roger was less robust."

When Roger was 18 months old, Phil was often away and holding down a very busy job, so it was his wife Elizabeth who was at home looking after the two small babies. She realised by now that something was not quite right about Roger's development, and took him to their GP, who in turn referred him to a gastroenterology specialist. Roger was tested for cystic fibrosis (which partly results from a malfunction of the lining of the intestine).

Roger had started to walk by this time but, Phil said, "We had to lift him up to get him on his feet, he really struggled. And six months later, it was obvious to us that he was walking strangely, showing lordosis - a curved back."

He was then referred to a neurologist, and eventually a muscle biopsy showed that Roger had a rare condition of congenital Muscular Dystrophy.

Roger and his parents were not the sort of people who give up. "We had to battle for NHS support, to get him a special wheelchair. He went to the local nursery along with is peers, but there was no question of a need for anything other than integration. His physical disability gave him a label, sometimes people would pat him on the head. Yet he was the healthiest member of the family, he rarely had a day off sick from school. By the time he was doing his GCSEs he'd been 15 years in a wheelchair - but then he started having headaches and difficulty with his breathing."

"Elizabeth didn't work, she delayed her career to look after Roger, but this brought benefits for her sons, her being at home." When Roger started having breathing difficulties, Elizabeth took him to a neurologist who explained that his breathing was deteriorating because of his posture due to his weakening muscles, and as a result the concentration of oxygen in his blood was too low. "We were fortunate," Phil said. "There was a new paediatric anaesthetist who knew about Bi-PAP [positive airway pressure] ventilation - a fan pushes normal air into the lungs at a higher rate overnight. So Roger then had the nightly ritual of putting on the face-mask, but it meant he was able to get a good night's sleep at last." Roger went on to take his A levels, and did

sufficiently well that he was able to go to the University of Manchester to read Economics.

"Roger means 'straight as a sword' -- and that's his personality, he was completely straightforward about everything."
He had his own budget for 24-hour support from the social services through the Direct Payments scheme, and when he was interviewing for a new carer, he would meet people in the Whitworth Art Gallery, which he jokingly called 'his office'; he chose Teddy, a Nigerian, who gave him 24-hour support.
"Roger called him 'his butler' - Teddy would have his clothes all folded neatly and his papers put in the right place. They got on really well, and Teddy was devoted to his job as a carer."
Roger had an amanuensis at college to take notes for him, and he also used a speech recognition programme on his computer because he was so slow at typing. Phil smiled when he told me, "He got away with murder. No-one in a pub could gauge his age and his pals used to get him to buy their drinks. He'd say of his disability 'You've got to balance it out on some occasions'. He had a good time at university, And he was an inspirational chap, he worked hard to compete with his peers."

On Millennium Eve Roger stayed out late with his friends, and he got very cold. A week later he had a pain in his chest.

Phil's own words describe Roger's final illness:
"His GP felt that we knew most about his condition and that we were prepared to fight our corner. At that time, because of his age, Roger was in the middle of a transfer from the paediatric services to the adult services, and so he was taken to the children's hospital - where he collapsed. They gave him oxygen, which was toxic at that level because his tissues had been starved of oxygen. Elizabeth had phoned me and so I was now driving up the M6, trying to get there as fast as I could. I got to the hospital and Roger was lying on his back, though we knew he was only comfortable on his side because his body was contorted and he often had flexures. Elizabeth was in tears as he was taken to Intensive Care. When I saw him I couldn't speak, for nearly an hour. I was in shock; it's incredible how it hits you... But after four days in IC he was taken off the ventilator and was able to come home. He even went back to university. I used to meet him at lunch and help him with various things. On Fridays he'd come home and have a bath and so on.
One Saturday, about five weeks after he'd come out of hospital, we took him out for a walk to get some fresh air, but he was very, very weak and didn't eat anything when we got home.
It was our younger son Douglas' birthday, and we were having a party in the village hall. Roger came, of course, but then he wanted to go home (we had a big Caravelle so we could fit in the wheelchair). I put him to bed and he said, 'Dad, do you think this is serious?' That was the last thing he said to me.
Elizabeth went to him in the morning, he was in the final stages, struggling to breathe. We took the ventilator off him, and we said to him, 'You've got to be able to do this yourself.'
We were surprisingly calm. He started Cheynes-Stoke breathing, and we rang the emergency GP, but we said, 'We know what's happening, we don't expect

you to call the blue light.' The emergency GP came later and certified his death.
We learnt a lot of things, such as dying on a Sunday morning is pretty good -- lots and lots of his pals were accessible.

"His life was happy, he was very positive, and full of the things he'd achieved. When we held his funeral in Manchester, people couldn't get in the doors. People who knew me even came from London. Afterwards his pals bombarded me with questions about him: that generation is very accepting, they hadn't questioned him about why he couldn't walk for example, they just had a fantastic acceptance of his disability.
The warden of the hall where Roger had lived asked me about having a memorial service - he said there were lots of very upset, lovely young people who wanted to remember Roger. The service was completely moving, so many of them said things about him. There was a huge chap, in the soccer team, who stood up and said, "He was a wizard on play-station - he could beat us all," and that they were going to institute an annual cup in his name.

"I have box-files full of correspondence telling how he and we had fought, won and lost battles with authority over provision of his care. He was never miserable, he was full of life and positivity. He was the first in so many ways, he was a pioneer - he was able to work his way through all sorts of irritating things that the system threw against us. And he was recognised as a pioneering voice, he knew about all the technical gizmos, and did wonders with his mobile phone.

"There's a lot of joviality, sometimes when we're trying to decide something we say 'What would Roger have decided?' We still talk naturally about him. We don't use words like 'sad' or 'illness' when we talk about him, we use positive words, nothing negative. He was great fun; thinking about him keeps me going. I dream about him a lot - without disability and at the age he would be now."

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Muscular Dystrophy is a condition that comes in several different forms and degrees of severity.
Roger's variant was found to be an autosomal recessive type, which means that although both Phil and Elizabeth are 'carriers' of the altered gene, it is over-ridden in each of them by the dominant, unaltered, version; in other words, neither of them show any symptoms of MD.
But if there are two copies of the altered gene - as in the fertilised egg which gave rise to Roger - this means that the gene's effects will be expressed.
"We received some rudimentary genetic counselling, telling us the implications if we were to have more children," Phil explained. "We were told we had a 1 in 4 chance in producing another child with the same condition. We didn't really discuss it, and when we had chance conversations we said 'we always wanted 3 or 4 kids'. The thing is, Roger's condition when he was young wasn't very challenging, so we thought it would be all right -- but the less sensible interpretation is that we were completely foolhardy! At that

stage, too, Martin was running around and healthy. Then Douglas was born in 1986 and when he was born, the doctor said 'He's a good 'un'."

Martin and Douglas have a two in three chance of being a 'carrier' of the recessive altered gene, so this has possible implications if they have children - although the chances of the other partner carrying a copy of the recessive gene will be no greater than in any other genetic condition which occurs in about 1 in 20-50,000 people.
"We're not a family that discusses its feelings," Phil told me, "A few years ago I sent Martin and his brother Douglas an email saying 'you need to think about this, and to be aware that the understanding of genetics has advanced'. For the last two years I've been trying to find the right opportunity to sit down with my sons and talk to them about it. But we believe that if and when the time comes we'll answer questions as best we can given the ongoing lack of knowledge of this condition."

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Phil also told me that "A sample of Roger's DNA was sent to Kay Davies (at Oxford University), and she said it would be archived for future study."

During a discussion at the ESRC Genomics Forum in December 2010, Dr Calum MacKellar of the Scottish Council on Human Bioethics noted that 'biobanks' had existed for years: "There are all kinds of tissue and DNA samples at the bottom of lab freezers, waiting for a PhD student to do some research on a particular topic." This presents huge difficulties from the ethical point of view, in terms of consent for example, because the records have often been poor. "Neither donors nor family know about this -- and it will be impossible to seek consent."

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Donating organs

Philip Dyer, Professor in Transplantation Science and Consultant Clinical Scientist, is the Director of the Histocompatibility and Immunogenetics Services for the Scottish National Blood Transfusion Service. He has had more than 30 years' research experience in organ and tissue transplantation support and is a past President of the British Transplantation Society; he has advised the Government on the Human Organ Transplants Act (1999) and the Human Tissue Act (2004).

He also gives many public talks about the importance and ethical implications of organ and tissue donation.
"When I give a talk about donating blood or tissues, my last slide is: 'Make your decision' - there's no sitting on the fence."
By the end of his talk, when he hands out information about Organ Donation and registration, very few of his audience are left undecided.

We can register as organ donors - but that does not necessarily mean that our organs and tissues will be retrieved and used.
"Organ-donor co-ordinators work in Intensive Care units - if they present the opportunities to a patient's family in the right context, they may be willing. But sometimes also families who have said 'no', later say that they regretted that decision."
Phil believes that, if someone has registered as an Organ Donor, and there are no medical reasons why their tissues cannot be used after death, it is in essence a contravention of their human rights to ignore that request.
His letter to the *Lancet*, reproduced above, shows that for people dying at home, these 'human rights' might be ignored; in the case of their son Roger, the Dyers had to exert pressure, hurriedly, to make sure his organs were used.
"Only Roger's corneas were used because of the way he died, at home. We knew his eyes were fine. But it would be different in hospital - they could take tissues, tendons, heart valves -- because the staff would know the circumstances and some details of the patient's life-style such as whether they were on narcotics. Biological screening is easy in hospital, and communicable disease are recorded."

As for Elizabeth Dyer, she now supports other disabled people who are "trying to wade through the systems. We were lucky," Phil says, "we had the ability and drive to deal with it, and now Elizabeth is using what she knows to help others. We observed the '33% rule': one-third give up at the first hurdle; another third give up at the second hurdle - and only the persistent final third get the necessary help."

Roger Dyer's legacy is not only two donated corneas, but the readiness of his parents to fight to improve the lives of other people.

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Further information about congenital Muscular Dystropy