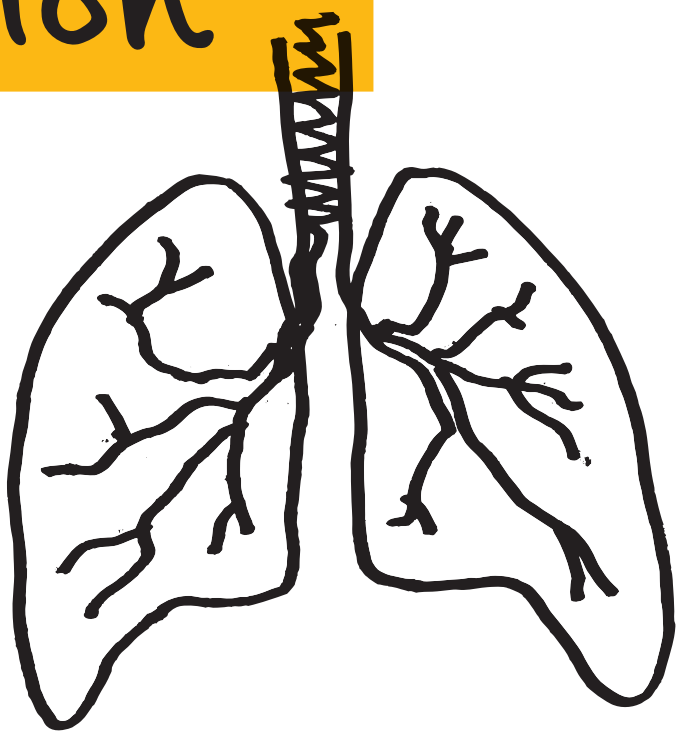
Cystic Fibrosis

a sticky, painful, suffocating condition



Hope for more

Improving access to lung transplantation and care for people with cystic fibrosis

David (left) with daughter Emma (right)

David's plea

I'm writing in support of all the people with cystic fibrosis who are on the lung transplant waiting list – and all those who will need a transplant to extend their length and quality of life in the future.

Currently one in three people with cystic fibrosis on the waiting list will die before they receive a life-saving transplant. I lost my daughter, Emma, in July 2013.

A month earlier, Emma took part in the Cystic Fibrosis Trust's campaign to improve the life chances of people with cystic fibrosis on the lung transplant waiting list. She shared her story in the hope that she could make a positive difference for everyone who may need to join the list. We were so proud of her.

Emma lived the first few years of her life as a happy and healthy child. At the age of seven she was diagnosed with cystic fibrosis, but she never let this define her.

Instead, she grew to be a funny, intelligent, vivacious and extremely talented young woman.

As with any family, everyone worked hard to keep Emma as well as possible despite her condition and she was determined to carry on with life. Emma always lived life with a smile, a smile that was warm, radiant and beautiful.

Through all of this, Emma knew that this awful disease was slowly attacking her lungs.

Emma spent the majority of 2012/13 in hospital. By this time, Emma's lungs were so scarred she was recommended for a transplant.

On 8 July 2013, she succumbed to an infection which took her life. She was 20 years old.

We were and always will be devastated – no parent should have to say goodbye to their child in that way.

Sadly, this story is all too common. Of the 10 people with cystic fibrosis who volunteered to share their stories during the Trust's campaign, four have since passed away without receiving the transplant they so desperately needed.

The Cystic Fibrosis Trust has produced this report calling for action from commissioners, NHS Blood & Transplant, NHS England, and others.

It is the result of the consultation that was launched at the Trust's parliamentary reception in June 2013.

The report contains actions that people working in health policy need to take. I am asking you, on behalf of the one in three people with cystic fibrosis on the transplant list to take those actions. Because I want to give hope to all those people on the transplant list now and in years to come.

David Kingston, father of Emma, who passed away last year aged 20.

Executive summary

One in three people with cystic fibrosis on the lung transplant list today will die before receiving a transplant.

Everyone with cystic fibrosis who has been assessed as eligible for and needing a lung transplant should receive one. There must also be a focus on the quality and standards set for lung transplantation so that everyone with cystic fibrosis that undergoes the procedure has access to the best information and is confident that they are receiving the best care available.

To do this we must focus on three key areas:

- Making the most out of the organs available now
- Increasing the number of organs available for transplantation
- Delivering a model of care that empowers the patient population

This report sets out 12 actions that we want policy makers and NHS professionals to take and three actions that we, the Cystic Fibrosis Trust, have committed to undertake.

Commitment

The Cystic Fibrosis Trust will continue to raise public awareness of the importance of organ donation and family consent to donation.

Recommendation

The Welsh Government and associated organisations must continue to involve the public in their scrutiny of the Human Transplantation (Wales) Act 2013.

Recommendation

NHSBT must develop a lung allocation system that balances the needs of medical urgency with longer-term outcomes, regardless of where a person is based in the UK.

Recommendation

Cardiothoracic transplant centres need to equip their surgeons with the skill to downsize lungs. Commissioners must promote a culture of controlled innovation to increase the number of lung transplants.

Recommendation

NHS England must fund an evaluation of the role of ECMO and Novalung in lung transplantation.

Recommendation

There must be a renewed focus on understanding what lungs are suitable for transplant. Patients should make a truly informed choice.

Commitment

The Cystic Fibrosis Trust will seek the views of those with cystic fibrosis from across the UK to evidence the community's views on acceptable limits in innovation and risk-taking.

Recommendation

The clinical reference groups for cardiothoracic transplant and cystic fibrosis must facilitate the development and implementation of a shared care protocol and standards of care.

Recommendation

Specialised commissioners must address gaps in the information, support and psychosocial services currently available through lung transplant and cystic fibrosis services.

Recommendation

NHS England, NHSBT and other stakeholders must discuss current reimbursement activity for organ donation, and agree on the steps needed to remove any financial barriers to achieving optimum donation.

Recommendation

Cystic fibrosis and transplant services staff should have the opportunity to observe patient progress through the full lung transplant assessment process.

Commitment

The Cystic Fibrosis Trust will facilitate opportunities for transplant and cystic fibrosis service staff to come together to share ideas and discuss the latest developments.

Recommendation

The Royal Colleges and specialist professional societies must improve opportunities for training, exchange, mentoring and shadowing schemes for junior and senior surgical staff within the UK cystic fibrosis and transplant services.

Recommendation

NHSBT must identify a common data set of essential information for people with cystic fibrosis considering a lung transplant.

Recommendation

NHSBT must make public its work assessing the risks and benefits of measuring success of UK lung transplant centres through survival measured from arrival on the waiting list and not solely from time of transplant.

"I was pretty healthy when I was younger. Through university my lung function started to plummet from 90%, to around 30%. I am in and out of hospital and have pseudomonas, which makes me unwell. I end up in hospital once a month for intravenous antibiotics for two weeks. Luckily for me I can walk without a wheelchair and at the moment don't need oxygen during the day, but I do have it at night. I take about 60 tablets a day.

"It's a tragedy that people die on the transplant waiting list if there are ways of overcoming the amount of organs that sadly get discarded."

23-year-old Poppy from Monmouth has been on the transplant waiting list since March 2012.

One in three people like Poppy will die before they can receive life-saving donor lungs. They are the reason this repot matters.

For Poppy it is a matter of waiting and hoping. In the meantime, she has to rely on a heavy burden of tablets and intravenous antibiotics to keep going. By doing what we can to increase the number of organs available for transplant, we can give Poppy, and many more like her, hope.

"It's not right that one in three people are still dying waiting for lungs, especially if people have been kind enough to donate them in the first place. I support the Trust's campaign to increase the rate of transplantation for people like me, because until you are on the waiting list, you can never picture what this is like."

Cystic fibrosis and lung transplantation

There are over 10,000 people with cystic fibrosis living in the UK. Cystic fibrosis is a life-shortening inherited disease caused by a faulty gene. This gene controls the movement of salt and water in and out of your cells, so the lungs and digestive system become clogged with mucus, making it hard to breathe and digest food.

Since the introduction of the newborn screening programme the condition is often diagnosed from birth and is managed through a demanding daily regime of drugs, physiotherapy and exercise. Half of those currently living with cystic fibrosis will live past 43 years of age. However, so many other people with cystic fibrosis still don't live to see their 40th birthday. In the year that we are marking our 50th anniversary, we think this is unacceptable.

So many other people with cystic fibrosis still don't live to see their 40th birthday. In the year that we are marking our 50th anniversary, we think this is unacceptable

The condition continues to claim the lives of younger adults, teenagers and children, and for the majority of people with cystic fibrosis, a lung transplant will, at some point, become the only means of significantly prolonging their life.

The wait for a lung transplant can be agonisingly long, at an average of 412 days for an adult. When added to the lung transplant list, people are commonly reliant on oxygen and have to endure a heavy burden of treatment to keep them alive. They are usually expected to live no more than two years if they don't receive a transplant.

412 days
is the average wait for adults on the lung transplant list

2 years
is the life expectancy for an adult on the transplant list who doesn't receive a transplant

1 in 3
People with cystic fibrosis on the lung transplant waiting list will die before receiving a transplant

For people with cystic fibrosis and those close to them, the risk of acute infection and rapid decline makes this period all the more stressful.

Cystic fibrosis is the third most common reason for lung transplantation (16.8%) and has the best survival rate (60% at five years). This is likely to be the result of a combination of factors for the disease cohort, including comparatively young age and greater levels of adherence due to a lifelong medical regime.

The status quo and its challenges

- Lungs from less than 25% of Donors after Brain Death (DBD) are utilised in transplantation. Within the remaining 75% there are clinically viable lungs which are suitable for use, and we need to maximise their usage.
- It is clear a large number of donor lungs are never used, despite consent being given by the next of kin.
- The UK continues to have one of the highest rates of family refusal to organ donation in the Western world[1], and 115 families overruled their relatives' consent to be organ donors in 2012/13[2].
- The current system for allocating lungs to transplant centres operates on a rota system. When a donor becomes available, the organs are offered to the closest transplant centre, when a matching recipient has been identified. If no suitable candidate is found, the organs will be offered to the next centre, as per the pre-agreed rota.

There is concern across the cystic fibrosis community about equity in lung allocation. There is a perception that the likelihood of getting a lung transplant, and the time frames for this vary according to where an individual is listed. A donated organ will not currently necessarily reach the most in-need candidate anywhere in the country.

Background and consultation

At our parliamentary reception in June 2013, we launched a consultation to gain expert insight into ways in which the rate of successful lung transplantation for people with cystic fibrosis could be increased.

It garnered 140 responses from across the spectrum of cystic fibrosis and transplant clinicians and health service personnel.

In response to the consultation, the Trust established a Project Advisory Group of expert cystic fibrosis and medical and surgical transplant specialists to ensure full consultation on our recommendations from the clinical community (see box).

To complement this group, we sought the views of people with cystic fibrosis and heard from a focus group that was made up of six individuals who have undergone a lung transplant, two people who remain on the waiting list, an individual who has been assessed twice but has not joined the transplant list and one person who has yet to consider transplantation for themselves.

We have had no contact with Intensive Care Unit (ITU) teams caring for prospective donors. However, commissioning for donation is a key action identified in this document. Consulting with and disseminating our recommendations to opinion leaders in Intensive Care Unit (ITU) teams will be an important output.

Project Advisory Group

Dr Mohamed Al-Aloul – Cardiothoracic transplant physician, University Hospital of South Manchester

Dr Amanda Brennan – Cystic fibrosis consultant, University Hospital of South Manchester

Dr Martin Carby – Cardiothoracic transplant physician, Harefield Hospital, Middlesex

Dr Paul Corris – Cardiothoracic transplant physician, Freeman Hospital, Newcastle

Dr Thomas Daniels – Centre Director and cystic fibrosis consultant, University Hospital Southampton

Ms Sarah Elworthy – Specialist nurse in cystic fibrosis, Royal Devon & Exeter Hospital, Exeter

Dr Gordon MacGregor – Cystic fibrosis consultant, Gartnavel General Hospital, Glasgow

Dr Jacqueline Rendall – Centre Director and cystic fibrosis consultant, Belfast City Hospital, Belfast

Dr Richard Thompson – Cardiothoracic transplant physician, Queen Elizabeth Hospital, Birmingham

Mr Nizar Yonan – Cardiothoracic transplant surgeon, University Hospital of South Manchester

Professor Andrew Fisher – Cardiothoracic transplant physician, Freeman Hospital, Newcastle

Professor John Dark – Cardiothoracic transplant surgeon, Freeman Hospital, Newcastle

1 - Taking Organ Transplant to 2020, NHS Blood & Transplant (NHSBT), October 2013
2 - NHSBT Potential Donor Audit

Maximising use of available donor organs

Organ donation

More names are always needed on the Organ Donor Register and the Trust is working alongside NHSBT and others to encourage more people to sign up to the Register.

Less than a third of donors are registered so lack of registration is not necessarily a bar to donation. However, having more people on the register is a good measure of progress in public support for organ donation.

The UK continues to have one of the highest rates of family refusal to organ donation in the Western world[1].

The number of times family members override the previously expressed wishes of a prospective donor and refuse organ donation is alarming. According to the 2012/13 NHSBT Potential Donor Audit, 115 families overruled their relatives' consent to be organ donors.

There is a great deal more to do to shift opinion among the general public and in society at large.

The current UK strategy states that "where individuals have given express consent, it is important that this consent is honoured and that families accept their relative's intention".

The public must be encouraged to talk to their loved ones and make their wishes known. Families will then have no doubt that they are honouring their loved ones' intentions when agreeing to donation.

We are very interested in the progress in organ donation and awareness that is expected from the Human Transplantation Act in Wales, which from 2015 introduces a system where consent to organ donation after death is deemed to be given in the absence of express consent.

We believe the shift to this system of consent will heighten the national debate around transplantation and encourage people to discuss organ donation with their friends and families while they are still alive.

The heightened public awareness means that family discussions are more likely to take place. These prior discussions make it more likely that family members will respect the wishes of their relatives to donate organs at the time of their death.

Recommendation

The Welsh Government and associated organisations, including NHSBT, must continue to involve the public in their scrutiny of the Human Transplantation (Wales) Act 2013, which comes into force in Wales in 2015. Progress in donation rates and public opinion must continually be reviewed.

Commitment

The Cystic Fibrosis Trust will continue to:

- raise public awareness of the importance of organ donation by engaging the media to inform opinion about the importance of transplantation and, vitally, to improve rates of family consent to donation;
- seek out communication opportunities, promote key messages, provide media-trained expert staff for interview and comment, work with expert medical contributors and promote case studies pre- and post- receipt of new lungs; and
- work with other organisations in the four nations, to maximise our impact.

1 – Taking Organ Transplant to 2020, NHS Blood & Transplant (NHSBT), October 2013

The lung allocation process

There is no national system to give lungs to the most urgent patients. Instead, lungs retrieved from a donor that are suitable for transplant are allocated to the closest transplant centre. Transplant clinicians at the centre will then assess the suitability of the candidates on their centre-specific waiting list for the procedure. If no suitable candidate is identified, the next transplant centre, in relation to a rota system, is alerted.

People should receive the transplanted lungs within eight hours.

We believe that a national system of allocation should be implemented. Where an individual lives in the UK should have no impact on the likelihood of receiving a lung transplant.

It is essential to implement a system for allocating lungs to potential recipients that minimises disparities in waiting time between individuals with comparable risk of death, and similar medical, geographic and demographic characteristics.

When considering a fair, safe and sustainable model for lung allocation, the quality of the donor organ, the wellness of the potential recipient, and predicted post-transplant survival all influence the weighting of an individual's suitability.

We believe that a national system of allocation should be implemented.

People with cystic fibrosis and clinicians feel left out of decision making by transplant centres because they do not know enough about the process. An improved allocation system, coupled with transparency and better communication, would help reassure people with cystic fibrosis that the process is actually working for them, with no in-built biases.

Lung allocation is an evolving situation in NHSBT, where it has been under active consideration. We keenly await the results of the analysis of the system being conducted by a subsection of the Cardiothoracic Transplant Advisory Group. We will scrutinise the outcome to ensure it is fair, equitable and transparent.

Any system must balance the needs of the person whose health is slowly declining on the list with those of someone whose health is suddenly deteriorating. The combination of clinical criteria, waiting time and illness severity make this particularly challenging.

The United States has a different system of lung allocation to the UK. Evidence accrued since the introduction of the Lung Allocation Score (LAS) in the US in 2005 shows the change in system has led to an increase in lung transplants for people with cystic fibrosis and a significant decrease in people with cystic fibrosis dying on the waiting list.

Super-urgent lung transplants

NHSBT has devised an add-on to the existing lung allocation system which is expected to begin operating in April 2014. It will consist of a Super-urgent Group, separate to the main waiting list. The people in this group will be a national priority and receive the first compatible lungs available from anywhere in the UK.

However, this change is only expected to impact small numbers of people, and is unlikely to have a significant impact on people with cystic fibrosis waiting for a lung transplant.

With around 60 people with cystic fibrosis waiting for a lung transplant at any one time, the numbers added to this list will not be enough to prevent the one in three people on this list today, who die waiting for a transplant. An even smaller proportion of the people with cystic fibrosis on the waiting list will reach the acute criteria required for Super Urgent listing.

The LAS balances estimates of medical urgency before transplant with the expected prospects for longer-term survival and success afterwards. The score gives priority to those most urgently in need but who are also expected to derive the greatest benefit from having a transplant.

Analysis of its impact in the US revealed dramatic reductions in waiting times, increased numbers of procedures and decreased mortality among listed patients.

Parts of Europe use a modified version that suits their local context, including making the system nationally applicable. A national model for the UK could develop the framework of the LAS in the US and end a system that allows where you live to be a factor in the likelihood of receiving a lung transplant.

Recommendation

NHSBT must progress towards a system that balances the needs of medical urgency with longer-term outcomes, regardless of where a person is based in the UK. This can be achieved through nationally coordinated allocation for all urgent and non-urgent individual cases. The lung allocation system must be made more fair and equitable.

Any mechanism introduced needs to be transparent. Data on how the system is working must be made available regularly, to enable the process to be modified in line with findings.

Increasing the number of organs transplanted

Innovation in techniques and surgery

New clinical techniques being developed mean surgeons can transplant lungs they would not have done before. This technological innovation has the potential to further increase the rates of successful transplant.

Ex-Vivo Lung Perfusion (EVLP) is a technique for the assessment and potential repair of sub-optimal lungs. The results of a current study at the University of Newcastle, funded by the Trust, could increase the number of lung transplants performed, reducing the number of people with cystic fibrosis who die on the transplant list waiting for suitable donor organs.

A more significant impact is expected to come from an increase in the re-sizing of lungs. The extension of this method of transplant also helps tackle the existing inequality made particularly acute by the small stature of many people with cystic fibrosis, especially women, compared to the donor pool.

The primary existing barrier to adequate provision of lung re-sizing is the skill needed by the surgeon to undertake this procedure. The UK does not have a culture where lung re-sizing is routine, despite evidence to support the value of this provision from other centres, throughout the world. This needs to change.

"I would really like to see more training and technology for resizing lungs so that organs can be made more widely available, especially for smaller people".

Vicky Pettersen, 43, from Cleveland, was put on the transplant list aged 28. She was fully oxygen dependent; she could not do anything with her son Anthony and husband Erik, and had a poor quality of life. Since receiving her transplant, Vicky has gone on to win silver medals in the British Transplant Games, and sits on the Freeman Heart & Lung Transplant Association Committee. She has also celebrated her 23rd wedding anniversary and seen her son go to university.

"If I hadn't received a transplant when I did," she says, "I wouldn't be here telling you my story, and my son would have grown up without a mother."

Techniques to artificially preserve critically ill patients and allow an extended time frame to receive donor lungs have been developed to prevent death for the individual and reduce mortality on the waiting list.

However, while an individual may benefit from Extra Corporeal Membrane Oxygenation (ECMO) or Novalung as a bridge to transplant there was no clear consensus in our consultation on how or if those techniques improve overall lung transplant rates.

Super-urgent allocation, currently proposed under the new lung allocation scheme, might increase demand for ECMO and evidence is being collected to confirm if this is the case.

Outlining the appropriate circumstances for ECMO or Novalung interventions to be undertaken is an important step in managing and controlling expected costs.

No one with cystic fibrosis should access ECMO without first being on the lung transplant waiting list. Only in those circumstances can transplant teams use ECMO efficiently as a bridge to transplant.

Recommendation

Cardiothoracic transplant centres must equip their surgeons with the skill to downsize lungs.

Commissioners must promote the provision of increased thoracic surgical capacity for lobectomy and promote a culture of controlled innovation to increase the number of lung transplants. Downsizing lungs must be a particular focus to address the inequity of access to transplant for smaller women.

Recommendation

NHS England must fund an evaluation of the role of ECMO and Novalung in lung transplantation.

Commitment

The Cystic Fibrosis Trust will establish what research is already available internationally in the area of lung transplantation, and tailor these findings to the UK situation. We will focus largely on the areas of lung preservation, reconditioning, trimming and lobar transplant, jointly identified and owned as priorities in lung transplant and cystic fibrosis. This would complement the Research and Development objective in the NHSBT's 'Taking Organ Donation to 2020'.

Maximising safe usage of donated lungs

Lungs from less than 25% of brain dead donors are utilised in clinical transplantation. Within the remaining 75% there are clinically viable lungs. We need to maximise their use. A large number of donor lungs are never used, despite consent being given by the next of kin.

Recent scientific guidance suggests 'extended criteria' lungs are key to progress in increasing rates of lung transplant. Research published over the past 10 years by a range of international centres, including teams from the University of Toronto, Hannover Medical School, University Hospital Zurich and the Royal Brompton & Harefield Trust, suggests that the use of selected extended criteria lungs may expand the donor pool without adverse consequences for lung transplantation.

Existing lung donor criteria were originally drawn up over 30 years ago, and while reviewed in 2001, have changed little. With the number of people waiting for a lung transplant increasing on a yearly basis, but the number of donor lungs (until recently) not following the same trend, there is an urgent need to expand the pool of useable donor organs for transplantation.

Many lungs designated as 'marginal' by the International Society for Heart and Lung Transplantation (ISHLT) criteria are now perceived by experts to be as clinically effective as standard criteria lungs. Several small, centre-specific reports have documented the use of this organ grouping, with outcomes comparable to those from studies featuring standard criteria donors. While the medical evidence on this front is continually evolving, public perceptions are falling behind.

There is an uncomfortable mismatch between expert transplant professionals' knowledge and the views of prospective recipients, who are naturally concerned about lungs that have been discussed as being a risk. Surgeons offer selected extended criteria lungs in the individuals' best interests and as a lifesaving procedure, but the individual may perceive risk or recklessness in the offer. The information on these issue is among expert groups. A better structured delivery of information to all parties is hugely important in rectifying this disparity.

Informed choice is an integral part of the process. The consequences of a decision to turn down donor lungs should be fully explored as part of assisting people with cystic fibrosis to make informed choices. Evidence must be shared with potential recipients who will be making informed choices to go on the transplant list and consent to transplant when lungs are available.

A large number of donor lungs are never used, despite consent being given by the next of kin

Recommendation

Everyone – from policymakers to those working on the frontline – must help ensure public and professional opinion is brought up to date with current scientific evidence. This evidence shows that the survival rates using extended criteria lungs are comparable to those using traditional criteria donor lungs.

There must be a renewed focus on understanding what lungs are suitable for transplant: there is strong evidence indicating that a new approach is increasing the rates of successful lung transplant elsewhere in the world.

Commitment

The Cystic Fibrosis Trust will seek the views of those with cystic fibrosis from across the UK to provide evidence of the community's views on acceptable limits in innovation and risk-taking.

75%
of clinically usable lungs are never used

Health service delivery

Ninety-one per cent of respondents to our online survey are in favour of developing pre- and post-lung transplant standards of care. This result was consistent across responses arising from transplant and cystic fibrosis services. Vitally, this should improve care and outcomes, although it is not expected to increase lung transplant rates.

Agreed standards of care would set a benchmark for services to meet, ensuring all people with cystic fibrosis have equal access to the highest level of multidisciplinary input in cystic fibrosis and transplant care, adequately resourced and based on the latest best evidence. This should be a document covering pre- and post-lung transplant, with comprehensive individual and family involvement in its design and development. The aim would be to improve quality of life on the waiting list and after transplant, as well as extend life expectancy post-transplant.

Standards of care can support benchmarking and form the basis for peer review and monitoring. This would aid commissioning across the breadth of services for people with cystic fibrosis who are on the lung transplant journey.

The standards of care would inform:

- Clinicians and other allied health professionals.
- Commissioners of cystic fibrosis and transplant services.
- People with cystic fibrosis, who should know what level of care they should expect.

The survey also showed 87% of respondents are in favour of developing a shared care protocol between cystic fibrosis and transplant services, across both pre- and post-transplant domains. Respondents saw several advantages, mostly in monitoring post-transplant but also in improving the management of people with cystic fibrosis on the waiting list. Over a third (37%) of respondents agreed that people are not referred for transplant early enough. Standardised transplant referral procedures, the third highest ranked suggestion for improving lung transplant rates, must be addressed within our proposed shared care model.

The commissioning of post-transplant care is a crucial area that must be carefully considered, because the on-going care of a post-transplant patient requires a substantial time and monetary commitment. This will be built into our review of a shared care protocol.

Of our survey respondents, 62% considered current psychosocial support during the transplant process to be insufficient. The demand for services that assist the family and individuals in coping with cystic fibrosis is drastically underestimated. The critical events on the lung transplant pathway – assessment, acceptance or rejection for transplant, life on the waiting list, false alarms, lung transplant itself and life after – are all deeply personally challenging. A lung transplant is not a cure for cystic fibrosis, and complications such as diabetes, osteoporosis and digestive issues will still remain.

Psychosocial care must be commissioned as a demand-led service to ensure a timely response to need. It must be locally accessible, for as long or as often as required and allow for self-referral and re-referral at any time.

Commissioning arrangements for organ donation were not initially identified as a key lever for improving lung transplant rates. However, it emerged that commissioning the ITU activity that facilitates all lung donation could be transformative.

Organ retrieval is commissioned by NHSBT, transplantation is commissioned by NHS England and equivalent bodies in Scotland, Wales and Northern Ireland, but early steps on the donation pathway are not specifically commissioned. Specialist nurses in organ donation are employed by NHSBT and have successfully increased donation rates. However, ITU clinical leads also need flexibility in funding to finance ad hoc services, staff, equipment and beds at short notice.

Such a change in commissioning could enable a more efficient use of potential organs for donation and provide capacity for all ITUs to support all donations, every time. There may be a case to create an Organ Donation Clinical Reference Group to fully explore this concept.

Recommendation

The clinical reference groups for cardiothoracic transplant and cystic fibrosis must take the lead in developing standards of care for people with cystic fibrosis throughout the transplant journey. All UK lung transplant centres are in England, and it is therefore important these standards of care facilitate excellent cross-border care. The standards must be subject to ongoing review, and be developed alongside contemporary medical evidence.

Recommendation

The clinical reference groups for cardiothoracic transplant and cystic fibrosis must facilitate the development and implementation of a shared care protocol to sit alongside the related standards of care.

Recommendation

Specialised commissioners must appraise the information, support and psychosocial services currently available through lung transplant or multidisciplinary cystic fibrosis services, and address gaps in provision.

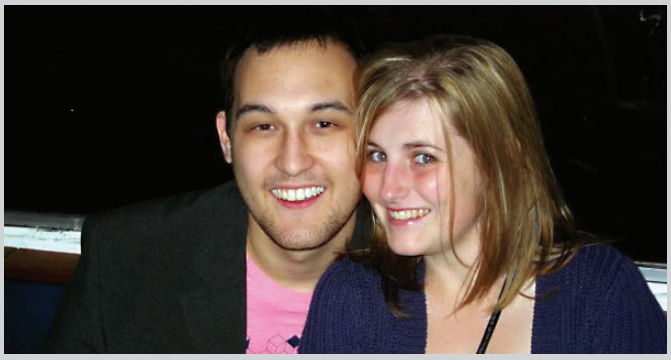
This service must be available from the moment an individual is advised to consider lung transplantation and continue indefinitely.

Recommendation

NHS England, NHSBT and other stakeholders must discuss current reimbursement activity for organ donation, and agree on the steps needed to remove any financial barriers to achieving optimum donation. This discussion should include analysis of advocating for service standards in intensive care that specify activities to support donation, extending further than the current provision of specialist nurses in organ donation alone.

"My hope is that when you need an organ there is more certainty, and it's not so much a lottery. The uncertainty of waiting for a transplant is enough."

Luke Yates, 27, lost his wife Sam on 4 April 2013. Sam was just 28 when she died, and had spent over three years on the waiting list for a double-lung transplant. Sadly, it never came.



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Communication, training and education

Inter-specialist uncertainty about practise in other areas is a clear problem, most obviously from cystic fibrosis services about the processes and practices in lung transplantation. Some staff do not feel suitably well prepared for discussing transplant with individuals prior to their operation, or caring for them again. A shared care protocol and the proposed standards of care would create greater inter-service familiarity and assist working relationships that enhance overall care; but more must be done.

Further education and professional development opportunities are needed to address this issue. There are many ways to deliver training and education in cystic fibrosis and transplant, with a good example being outreach clinics, where opportunities for patient-based expert discussion are available. Short term (three-month) professional rotations are popular as hands-on experience is of more educative value and relevance than shadowing. Additionally, we are currently sponsoring training fellowships to support those involved in cystic fibrosis care, treatment and research.

Our unique offer to professional education is our access to the cystic fibrosis community's voices and experience. We also have the ability to straddle the specialities – bringing together clinicians who might otherwise work in very separate fields.

Recommendation

Cystic fibrosis services staff should have the opportunity to observe patient progress through the full lung transplant assessment process as part of their ongoing training and education. In return, transplant staff should be similarly accommodated in cystic fibrosis clinics. NHS study leave, bursary and training budgets must be available to support this.

Recommendation

The Royal Colleges and specialist professional societies must improve opportunities for training, exchange, mentoring and shadowing schemes for junior and senior surgical staff within the UK cystic fibrosis and transplant services. This will improve surgical training and practice, enhance valuable knowledge-sharing and disseminate new techniques and practice as the evidence base develops.

Recommendation

NHSBT, in partnership with the Clinical Reference Group for transplant and cystic fibrosis, must identify a common data set of essential information for people with cystic fibrosis considering a lung transplant. It must include information on how to interpret centre-by-centre activity rates and outcomes data. It is vital that people with cystic fibrosis have comprehensive information on which to base their decisions regarding transplant.

Commitment

The Cystic Fibrosis Trust will facilitate opportunities for transplant and cystic fibrosis service staff to come together to share ideas and discuss the latest developments. Cross-specialism working will be encouraged to benefit the individual, and keep all involved up-to-date with current practise.

"I am grateful beyond words for the transplant I received, and I wish more people could benefit from organ donation the way I have."

Bernice, 22, from Haverhill in Suffolk, spent 15 months waiting for a call. She was staying in hospital last December when a nurse shook her awake at 3.30am, to let her know a donor had been found. Within three hours, Bernice was on the operating table receiving a new pair of lungs, which saved her life. Bernice is now planning the rest of her life, with a holiday high on the agenda. She also wants to continue her hairdressing apprenticeship after having to leave because she was too unwell to continue.

Bernice is an amazingly resilient and cheerful person. Speaking about the need to drive up the rate of transplantation, she said: **"It doesn't seem fair that there isn't enough training available or enough technology to resize organs for smaller people. A life is priceless."**

Bernice Perry and her sister

Measuring success

Currently, lung transplant success is measured according to the length of survival of an individual following transplant surgery, at various intervals from 1, 5 and 10 years post-transplant. This means that the effects of life on the waiting list are not reflected in outcome measures and therefore do not reflect clinical reality. Mortality on the list is not covered through this measurement.

The statistical focus on those who receive a donor organ obscures the importance to overall outcomes of management of people on the waiting list.

Recommendation

NHSBT must make public its work assessing the risks and benefits of measuring success of UK lung transplant centres through survival measured from arrival on the waiting list and not solely from time of transplant. This outcome measure will be important when assessing the effectiveness of alterations to the lung allocation system.

Changing the unit of measurement would optimise management of a group of seriously ill individuals. Their experience would form part of the success measure for the whole transplant journey. It would complement a change in the lung allocation system to prioritise the most seriously ill individuals with a chance of a good outcome.

We thank everyone who contributed to the consultation and look forward to helping to create positive change in transplantation for people with cystic fibrosis.

Cystic Fibrosis is a fight we must win

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