

Post print

This is a pre-copyedited, author-produced PDF of an article published in *Clinical Ethics*. The definitive publisher-authenticated version [Newson, A.J. (2009) "Clinical ethics committee case 7: Our young patient is in heart failure but has multiple co-morbidities. How can we best care for him and his family?" *Clinical Ethics*, 4(3): 111-115.] is available online at <http://cet.sagepub.com/content/4/3/111.extract>.

Clinical Ethics Committee Case 7:

Our young patient is in heart failure but has multiple co-morbidities. How can we best care for him and his family?

Ainsley J. Newson

Senior Lecturer in Biomedical Ethics, Centre for Ethics in Medicine, Department of Community Based Medicine, University of Bristol, UK
E-mail: ainsley.newson@bristol.ac.uk

Biographical Information

Dr Ainsley Newson is Senior Lecturer in Biomedical Ethics in the Centre for Ethics in Medicine at the University of Bristol. She has a PhD in Medical Ethics and Bachelors degrees in Science and Law. Her research interests include clinical and reproductive decision-making in genetics and synthetic biology. Ainsley is a member of the European Clinical Ethics Network and the Editorial Committee of this journal. She has been a member of Clinical Ethics Committees for 6 years; first at St Mary's Hospital in London and since 2006 at the Royal United Hospital in Bath.

Introduction

This is the seventh of a series of cases provided and discussed by UK clinical ethics committees. This series developed from the Virtual Ethics Committee, which discussed a case in each issue of the first two volumes of the journal. We invited all clinical ethics committees registered with the UK Clinical Ethics Network to volunteer to submit and then discuss a case, with cases allocated according to the experience of the committee. Committees have not discussed their own cases. The committees were given some guidance on how to generate a case, including the advice that unless consent from *all* parties to a case could be obtained, we would prefer fictional cases. The editorial committee took this decision to safeguard patient confidentiality. To the same end, we decided that the committee referring a case would not be identified (as this would provide a geographical indicator of identity), but we would name the committee discussing the case. A member of the editorial committee attends the discussion of the case and writes the summary to be published in *Clinical Ethics*, once the discussing committee and the journal editors have approved it. All committees and readers are invited to respond to this summary. We are also interested in publishing examples of good practice or guidelines on difficult areas that have been generated by clinical ethics committees during the course of their work.

Future series (starting with volume 5 in 2010) may examine some of the broad policy issues involving or affecting clinical ethics committees. Committees which would like to participate should contact the Case Studies editor Ainsley Newson (ainsley.newson@bristol.ac.uk).

The Clinical Ethics Committee at Royal United Hospital Bath agreed to discuss the case presented here. The Bath Clinic Ethics Committee (CEC) was established in 2000 and serves a large District General Hospital in the South West of England. Committee membership comprises hospital clinicians (doctors and nurses), a chaplain, a psychologist, a hospital manager, an academic ethicist and a lay member. The CEC provides a consultation service, education for hospital staff and offers review of hospital policies where ethics input is required (e.g. Jehovah's Witness, resuscitation). The committee has written a guide on ethical issues available to Hospital Staff. Although the committee meets in person only quarterly, members of the committee are available at any time through the hospital switchboard to receive referrals and give advice. In the nine years the committee has been operating it has discussed approximately 58 cases. 'Live' CEC consultations may take place at the bedside, at meetings of a small group from the committee, or by e-mail (with anonymised details). Cases have included end of life issues, refusal of treatment in adults and children and physician-patient (or family) conflict. All cases and notes of CEC deliberations are recorded in a pro-forma document that has been developed from North American models.

Referral to the Clinical Ethics Committee: A young patient with severe and progressive illness requiring organ support

T is 17 months old. He has been diagnosed with hypertrophic cardiomyopathy, an often-inherited life-limiting condition in which the patient's heart muscle hypertrophies (gets larger). As the muscle does not function properly, the heart cannot pump effectively. Of the few available treatments for this condition, heart transplantation can provide a means of achieving longer survival. T also has a rare inherited metabolic disorder called Complex IV Deficiency, characterised by a deficiency in the essential enzyme cytochrome C oxidase – involved in energy production. This disease can be localised or can affect multiple organs in the body and has an uncertain long-term neurological outcome. T is the youngest of four siblings. One of his older sisters died aged 18 months from cardiomyopathy and multi-organ failure.

Two months ago, T suffered damage to his heart such that it was unable to supply sufficient blood to his body. He was admitted to hospital, but then experienced an episode of an abnormally slow heart rate that required cardiopulmonary resuscitation. Following this, T was listed for a heart transplant and given Extra Corporeal Membrane Oxygenation (ECMO; effectively an artificial lung and heart, but a complex non-portable system that cannot be used for prolonged periods) to sustain him until a donor organ became available.

At this point, the treatment team were concerned about T being listed for heart transplant given that he also had a complex metabolic disease whose prognosis was uncertain. Very few, if any, heart transplants have been carried out in children with this combination of conditions. Although patients regularly survive with complex IV deficiency, it is not curable.

Despite this concern, a mechanical heart (a 'Berlin heart') was inserted to replace ECMO and support T's heart function. This device is smaller than the ECMO system, can be used for longer periods and can permit restricted movements of patients.

Soon after this procedure, concerns were expressed about an observed deterioration in T's neurological state. Although he did show some spontaneous signs of recovery (such as spontaneous eye opening) and his parents thought he was improving, the treatment team remained concerned. Their concerns were reinforced by a CT scan which showed irrecoverable loss of brain tissue at discrete sites. T's clinicians believe this damage would cause central vision impairment, paralysis on the right side of his body,

seizures and learning difficulties. Further tests also indicated renal and gut dysfunction, suggesting that T's metabolic condition was affecting multiple organs.

Because of this situation and its inherent uncertainties T's treatment team felt that he was no longer a good candidate for a heart transplant, particularly as the procedure may exacerbate his neurological condition. The metabolic team had also advised that they would not recommend transplantation solely on the basis of the metabolic condition, which they acknowledged would progress. Nonetheless a consensus agreement to continue treatment with the Berlin Heart, but to monitor neurological progress, was achieved.

T has now been on the Berlin heart for a further two weeks, with regular evaluation. The team have just met to discuss his prognosis. They noted some further changes to T's brain, with an uncertain cause. If the metabolic disease was involved, this would be a clear reason not to proceed to transplant. However if they were temporary, transplant may still be possible. T has exhibited some slow improvement, but there are still concerns that his disability will be severe. Further tests (such as lumbar puncture or MRI) are not possible due to the use of the Berlin heart.

T's treatment team feel they have three options: (1) relist him for transplant; (2) keep him on the Berlin hart and continue to evaluate him; or (3) discontinue T's Berlin heart treatment.

T's parents would like everything done for him. They were unhappy that he was taken off the transplant list, and did not understand why. They feel that they could handle a moderately disabled child but they need specific answers about what this means.

We are approaching the ethics committee, with the following questions in mind:

1. Two other children are 'urgently listed' for transplant. Should their claims trump T's?
2. What impact does or should neurological status have on heart transplant in children?
3. If T is no longer suitable for transplant, how long is it ethically justifiable to support him on the Berlin Heart?
4. What should we do to resolve this case and how should this be communicated to T's parents?

Response from Royal United Hospital Bath Clinical Ethics Committee

Thank you for your referral, which we considered at our meeting on 26th February 2009.¹ We found this case both interesting and difficult, particularly the questions of T's best interests and his quality of life, now and in the future. You proposed three options for clinical action and four questions for deliberation, which we used to structure our discussion.

Background to case

T appears to have two disorders; a metabolic disorder with serious morbidity and a neurological condition with relatively poor but unpredictable prognosis. T has then unfortunately also experienced a crisis precipitated by hypertrophic cardiomyopathy and heart failure. His life can most likely only be saved or prolonged with a heart transplant, as he is already being kept alive by way of a Berlin heart, a mechanical device that assists a failing heart by providing assistance to the cardiac ventricles (pumping chambers). This device is commonly regarded as a "bridge" to transplantation.

¹ Nb: The Case Studies Editor is a member of this Clinical Ethics Committee. She attended this meeting as a representative of *Clinical Ethics*.

We have assumed that: (i) the cardiomyopathy is unrelated to the metabolic disorder (although it could be a not-yet-described variant); and (ii) if a transplant were to go ahead it would be unlikely for the donor heart to acquire the same metabolic condition, although the disease might progress in other organs.

We discussed T's neurological state and the fact that he may deteriorate further. There is a requirement for clinical criteria to assess this deterioration, but there are also ethical issues inherent in this process – who should decide the quality of life criteria that should be used to assess T? What might be described as a reasonable quality of life for him? T is likely to have: severe learning difficulties, some paralysis and blindness, but if he can be kept comfortable and is loved and accepted by his family, should this be taken away from them? There will be some pre-existing clinical markers of deterioration, which should be taken into account, but we must also involve T's parents in these discussions.

T's parents' previous experience of the loss of T's older sister may also be relevant to their current request that the treatment team to 'do everything' and their disappointment with some of the decisions that have been made so far. This may give rise to a significant level of emotion which will require support. On the other hand, their previous experience of coping with their daughter's death could make them more resilient in the face of T's illness. Whichever is accurate, the loss of their daughter is likely to influence their decision-making for T. We don't have any information about the cultural or religious background of T or his family. This might be useful to determine what other support structures may be available to them.

Another potential difficulty for T's parents is that now that T has the Berlin heart, it may be more difficult for them to face a decision of withdrawing this treatment than it may have been to 'let him go' immediately after his cardiac episode. The distinction between withdrawing and withholding treatment is ethically contested but may be very real to T's parents.

In summary, we have interpreted T's medical condition as very serious. However the next stages and course of his metabolic illness are to a degree uncertain, as is any functioning that T may gain or retain were he to live.

We then turned to the questions you posed, and began by discussing the first option: relisting T for transplant.

1. Two other children are 'urgently listed' for transplant. Should their claims trump T's?

Transplantation is an expensive process with a finite resource of donated organs. It is therefore logical to ration this resource. However, the basis for rationing needs to be transparent. The Department of Health tends to use QALY (Quality Adjusted Life Years) to assess competing demands on resources; that is looking to determine who will get the greatest overall benefit from the finite resource - usually given to the person who has waited the longest. T has been taken off the list so his previous waiting time would presumably be lost. There are also principles from UK Transplant, but these do not explicitly address quality of life issues.²

² See: UK Transplant, Donor Organ Sharing Scheme Operating Principles for Cardiothoracic Transplant Units in the UK and Republic of Ireland "Allocation of Hearts to Paediatric Urgent Patients" http://www.uktransplant.org.uk/ukt/about_transplants/organ_allocation/cardiothoracic/cardiothoracic_organ_sharing_principles/cardiothoracic_organ_sharing_principles.jsp#c5 (Accessed 24 April 2009)

In T's case, we recognise that a heart transplant might give him a better quality of life and that it may enable him to go home. But we feel that his long-term prognosis remains poor. A heart transplant is a serious and risky procedure, with a higher clinical risk for children who are already seriously unwell. Assuming that the other children urgently listed for transplant do not have the neurological and metabolic co-morbidities of T, we believe these other children's claims would trump T's, however difficult this decision is to make in practice. We are mindful that a judgement comparing T's quality of life with that of other children is difficult, but we have a responsibility to all of our patients to ensure finite resources are distributed as fairly as possible, including maximising the potential benefit of those resources.

2. What impact does or should neurological status have on heart transplant in children?

Neurological status is a broad term and will have a varied clinical spectrum. A degree of compromise in neurological status should in general not be a barrier to organ transplantation, evidenced not least by heart transplantation in children with Down syndrome who have congenital heart problems. But listing and prioritising patients for transplant is a complex process and neurological status can risk further complications post-surgery. Rather than focussing merely on neurological status, you could consider T's broader quality of life. He will have some quality of life, but his neurological condition is serious and it may progress. To this end, we believe the degree of T's impairment and his other co-morbidities should be a relevant consideration for the decision of whether to re-list him for transplant.

Should T be re-listed for transplant?

Our answers to these questions and the above clinical assessment, lead us to the conclusion that T should not be re-listed for transplant. Although it is possible to argue that quality of life is subjective and difficult to assess, we must also recognise that we can harm children like T through going ahead with procedures that may lengthen his life but not provide any real gain. T may not subjectively feel worse but we would value the quality as well as the sanctity and length of his life.

Before this decision not to relist is made clinically, we would want to ensure that careful discussions had been held within the clinical team. These should take into account whether T is suffering with his current medical treatment, how long T might live with the transplant, the quality of the life he would have, and the risks that T might suffer further harm from the transplant than he would if it did not go ahead. The outcome of these discussions would then have to be carefully discussed with T's parents, addressing their fears and any gaps in knowledge. T's parents should also be able to talk to the transplant team if they wished to.

3. If T is no longer suitable for transplant, how long is it ethically justifiable to support him on the Berlin Heart?

This is perhaps the most difficult question to answer, as removing support for T's Berlin Heart will most likely also end his life. It forces us to examine both our professional and personal morality. We spent some time discussing the medical care for T in his current position. We discussed the nature of a Berlin heart and noted that although portable it comprises a sizeable cabinet, laptop computer, a pump and requires an abdominal incision. The use of this equipment would likely require an intensive care bed and there will be risks associated with its longer-term use, such as a risk of line infection. T may also require a PEG tube³ and a catheter, all of which carry risks. Given T's current status and the complexity of this

³ PEG stands for 'percutaneous endoscopic gastrostomy.' A PEG tube is a feeding tube that has been placed into the stomach via a minimally invasive abdominal incision. They are used to feed patients who cannot swallow.

equipment, we considered it unlikely that T would be able to be cared for at home while he has a Berlin heart.

T's parents have indicated that they would be able to cope with a child with moderate disability, but T's problems are likely to be greater than this. Given his age, T's lack of mobility with a Berlin heart may not be an issue, although it may become so in future. It is certainly not impossible for children to live for a long time with this support, but as T may experience deterioration due to his metabolic condition, long-term use may not be in T's best interests. Neither may it be the best use of this resource. There are real issues around rationing of this equipment and the clinical resources that will be required to support it.

Given that T is relatively stable at present, ceasing support now may not be appropriate. Several of us commented that it might be best were 'nature to take its course,' but this is not possible now that T has the Berlin Heart. We did wonder why these artificial support mechanisms were given in the first place, but concluded that this was probably done in an emergency situation when T's overall prognosis remained uncertain.

Withdrawing the Berlin heart will therefore become inevitable at some point, unless the parents very strongly object (in which case other courses of action, such as a legal determination of T's best interests, may be required). The decision to withdraw treatment is always difficult for both health professionals and family members. Treating very ill patients may be perceived as an easier option, as it allows you to offer the parents something and can avoid conflict. It will also give T's parents more time with their son. But even if his prognosis were better, maintaining T on a Berlin Heart exposes him to a risk of complications associated with immobility and the presence of invasive catheters.

This also raises a broader point about conceptions of quality of life, which have changed over time to encompass a wider range of states of health and well-being. However the issue of *who* makes decisions about best interests and quality of life can remain contested, particularly in paediatrics. For T, it is especially difficult as a decision about whether withdrawing the Berlin Heart is in his best interests will necessitate comparing his current life to no life at all. This decision cannot be made unilaterally by either T's treatment team or his parents. The reasons for not relisting T may not be the same as reasons to justify continuing or discontinuing his Berlin Heart support.

Advance planning should therefore be put in place now to determine what level of physical deterioration in T might justify withdrawal of this treatment. Clear criteria are needed (such as whether or when to give antibiotics, whether or when to change tubes and so on), coupled with regular reviews of T's overall situation. This should be undertaken with T's parents and the wider clinical team. T's parents may not even disagree with the recommendation to withdraw treatment, given their statements about caring for T. But open and frank discussions will help establish this and ensure good lines of communication.

Question 4: What should we do to resolve this case and how should this be communicated to T's parents?

T's parents are currently unhappy that he was removed from the transplant list, which suggests there may have been previous problems in communication. As we have already suggested, frank and open meetings with the family will be imperative to resolving this case (assuming that T's parents are not resistant to discussion). These meetings should be attended by those clinicians who know the parents. All known facts should be reviewed, including the benefits and drawbacks of the various options for proceeding and T's likely level of disability. Their overall family context, including support mechanisms, emotional and financial factors would also need to be raised. The family could also be invited to be put in

touch with other families who have experienced Complex IV deficiency to talk through their experiences. If you felt it appropriate, the clinical ethics committee (or some of its members) could also meet with the parents, to act as a further 'sounding board' for advice. However there would need to be value in this over and above the views of the clinical team and family that you will already be gathering.

T's best interests (both now and in the future) should form the focus for discussion. In addition to the aspects we've already identified, palliative care or pain management can also be discussed, as can bereavement counselling. The goal should be to reach a resolution on how to maximise T's quality of life given the competing considerations.

We also discussed, more generally, relatives who are particularly assertive in clinic. Some of us felt that at times, particularly in intensive care, it seems that we can in effect be treating a patient's relatives and not the patient. There may be two competing conceptions of best interests and it will be important to recognise this and try to address it. For T, we need to make sure that we are advocating for his best interests and not merely acquiescing to his parents' wishes.

We also considered that there may be a Human Rights aspect to this discussion. Although our committee does not provide legal advice, it is perhaps worthwhile to give some thought to this and to seek further advice if necessary. We also noted that the British Medical Association has produced a very helpful summary of the Human Rights Act and its possible impact on medical care.⁴

Concluding remarks

The resolution to the issues posed by T's case and his care is therefore a compromise between options (2): keeping him on the Berlin heart and continue to evaluate him, and (3): discontinue T's Berlin heart treatment. We are not convinced that further evaluation of T while on the Berlin heart will have clinical utility, although we are mindful that we have only seen a snapshot of the facts. But it also seems premature to discontinue treatment entirely, particularly given the current issues in the relationship with T's parents. What is needed, insofar as this is possible, is a clear understanding of T's likely quality of life and disability with respect to his ongoing treatment. Therefore we would propose that T be continued on the Berlin Heart for now, to allow more time for further discussion over the following days or weeks, but with a longer-term goal to withdraw support.

There needs to be frank and compassionate discussions with T's parents about his current and likely quality of life and an agreement on an outline timeframe for supporting T on the Berlin Heart and the deterioration criteria that would be met to justify withdrawal. The focus needs to be on T's best interests, even though this may involve the difficult decision to withdraw treatment.

Although the sanctity of life is an important consideration, the quality of any individuals' life can present at a variety of different points on a spectrum. T may currently appear settled, but at the same time there needs to be a realistic assessment of clinical futility and the right of patients, however young, to experience a good death. Accepting that treatment has failed is difficult but T's condition is terminal, so working with his parents to prepare for T's death is now imperative.

⁴ Ethics Department, British Medical Association. *The impact of the Human Rights Act 1998 on medical decision making*. 2007. London: BMA. Available at: http://www.bma.org.uk/health_promotion_ethics/human_rights/HumanRightsAct.jsp (accessed 26 February 2009).

Members of Royal United Hospital Bath Clinical Ethics Committee who discussed this case:

Dr Peter Rudd, Consultant Paediatrician (Chair)

Dr Monica Baird, Consultant Anaesthetist (Joint Vice-Chair)

Ms Vanessa Bishop, Committee Administrator

Dr Lindsey Dow, Consultant Geriatrician (Joint Vice-Chair)

Mr Julian Hunt, Consultant Nurse, Critical Care

Ms Tejal Mistry, Clinical Scientist Trainee (Internal observer)

Dr Barry O'Connell, Clinical Scientist Trainee (Internal observer)

Mr Stephen Roberts, Complaints and Litigation Manager

Mrs Sarah Symons, Solicitor and Lay Member

Ms Lizzie Tuckey, Medical Student, St George's Medical School (External Observer)

Dr Sarah Wexler, Consultant Haematologist