Current status of potential organ donation in cases of lethal fetal anomaly

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Key content
- Anencephaly is the most severe form of neural tube defect with most pregnancies terminated following antenatal diagnosis and those carrying to term often stillborn.
- Organ donation following anencephalic live birth has been undertaken worldwide, especially in the early years of infant transplantation. In the USA and Canada this ceased towards the end of the last century.
- Although few UK transplants occurred, and the practice has ceased, national guidelines still support organ retrieval from anencephalic babies once certified dead.
- With the current organ donation task-force linked drive, obstetricians are increasingly confronted with women carrying anencephalic fetuses who decline termination, perhaps for religious reasons, who would like to consider donation.
- Anencephalic donation does not occur in the UK, or indeed worldwide at present.

Learning objectives
- This paper seeks to review the current UK position and possible developments, and to provide guidance for obstetricians to reply to such requests.

Ethical issues
- Can a pregnant women elect for her anencephalic infant to be managed in such a manner – with organ support at delivery – that eventual donation after circulatory death can provide organs for others on the subsequent death of the baby?

Keywords: anencephaly / organ donation / severe fetal anomaly / transplantation

Introduction

Anencephaly occurs when the cephalic end of the neural tube fails to close, resulting in the absence of a major portion of the brain, skull, and scalp. Infants with this disorder are born without a forebrain or cerebrum and although some are born with a rudimentary brain stem the absence of cerebral function precludes consciousness, and infants are either stillborn or die shortly after birth.

Antenatal identification of anencephalic infants has progressed from α-fetoprotein screening, via static B ultrasound to almost universal diagnosis by high-quality real-time ultrasound, which permits cranial visualisation at 12–14 weeks. Following such diagnoses, the majority of anencephalic pregnancies are terminated. This, together with the use of folic acid to prevent neural tube defects, has lead to a 96% reduction in anencephaly rates over the period between 1975 and 1999.1 However, there are still in the region of 10–20 live born anencephalic infants in England and Wales per annum.1

Neonatal organ transplantation is, in itself, a rare undertaking in the UK – more so than, for example, in North America – as a result of UK policies on certification of infant death that inadvertently prevent organ donation in infancy.2

Whether organ donation from anencephalic infants that had been certified dead using neurological criteria ever occurred in the UK is unclear from the literature. In the US the controversial proposal of the removal of organs for transplantation from anencephalic infants before they were certified dead3 was short-lived, falling rapidly in the court of public opinion.4 Donation from formally ‘brain dead’ anencephalic infants continued for a time, however all programmes in the US have stopped following further debate and an influential Canadian ethical enquiry, which concurred that there are serious difficulties with using neurological criteria to certify death in anencephalic infants and that this ought to preclude donation after brain death (DBD).5 Indeed, no country seems to currently transplant organs from anencephalic infants with the most recent European transplant reported being in Italy in 1999.6 Somewhat surprisingly, therefore, the most recent UK guidelines from the Working Party on Organ Transplantation in Neonates,7 allows for such donation.
This guidance was unchanged, and therefore arguably endorsed, by both the British Paediatric Association document that established specific paediatric brain stem death guidance for the UK, and the contemporary Academy of Medical Royal Colleges national code of practice on the diagnosis and confirmation of death. The original Working party report suggested:

‘...that the absence of the forebrain in anencephalic infants together with apnoea shall be recognised as death.’

The document also stated that:

‘...organs for transplantation may be removed from anencephalic infants when two doctors who are not members of a transplant team agree that spontaneous respiration has ceased.’

Furthermore, such donation apparently remains lawful with parental consent, and causes no concerns for Her Majesty’s Coroner or the Human Tissue Authority (HTA) (Personal communication from Great Ormond St Hospital Solicitor, 2009 [after seeking senior legal advice] and both Coronial and HTA communication, 2009).

Dead donor rule

Apart from explicit living donation, the convention is that organ retrieval for donation can only be performed after the donor has been certified dead. Truog, amongst others, has questioned the inviolability of this ‘dead donor rule’, by arguing that brain death has no scientific basis, while also insisting that donation should continue from such patients who are, therefore, in the dying process rather than dead. In fact the entire question of the definitions of death, dying, and for that matter, human life are extremely complex, with only the procedure of organ donation driving exact times or indeed invasive testing to establish a diagnosis of death in the clinical setting.

Human death

It is worth setting any consideration of organ donation from anencephalic infants in the context of human death. Most people who die have death certified using what have been termed cardiorespiratory criteria, though this also involves confirmation of an absent pupillary response to light, as well as the absence of heart sounds, breath sounds and a pulse. Before organ support was possible, of course, cessation of respiration or circulation were usual precursors to human death with established absence used to ‘pronounce’ those who had died. However, historical recovery from such ‘states’ led to the setting up of devices to communicate from the grave and the building of charnel houses to allow witnessed putrefaction in those worried about being wrongly buried.

The onset of mechanical ventilation led to the evolution of a different method of the certification of human death: brain death. In the USA brain death is whole brain death, whereas in the UK the more bedside-orientated brain-stem death is used.

Brain death originally developed as a predictor of inevitable ‘somatic death’ in severely brain injured people on organ support – notably mechanical ventilation – but has, via clinical experience, a Presidential Commission in the US, professional guidance in the UK and court rulings, become an accepted method of certifying human death. Importantly, the development of brain death was remote from any concept of the provision of organs for transplantation, though certification of death using neurological criteria within intensive care has, without doubt, provided the majority of organs used for human transplantation.

One vital concept is that human death must be a uniform state, despite different methods being undertaken to determine it. Philosophers have suggested, logically, that one should first philosophically define human death, and only then suggest criteria for its diagnosis before agreeing tests to clinically verify death – whereas with brain death, things arguably evolved in the converse manner.

The pertinence of this to anencephalic donation is that for some philosophers anencephalic infants actually fulfil a conceptual definition of human death – namely the irreversible cessation of the capacity for consciousness – with its corollary that the presence of a circulation and respiration in a phenotypical human are not enough to define human existence. It follows that if we accept their neurological condition to be irreversible, those in a persistent vegetative state are also dead, with some suggesting that organ retrieval should be permitted in this circumstance.

Irrespective of such philosophical debates, the concept that anencephalic infants are dead at delivery due to their anatomy has been rejected by clinicians worldwide.

Anencephalic death

The current rather archaic UK guidelines permit certification of death using neurological criteria in an infant known to be anencephalic due to antenatal ultrasound findings once, after birth, the infant has ‘irreversible apnoea’. In essence this attempt at modifying standard brain stem testing, with antenatal imaging replacing conditions necessary for the diagnosis and confirmation of death as well as both motor and brainstem components of the clinical testing together with replacement of the apnoea test with the loose term irreversible apnoea, is clearly unworkable given the interim advances in medicine.

Furthermore, no other form of neurological condition is currently accepted by neonatal clinicians in the UK as equating to death, whereas in the US, Canada and Australia and indeed other European countries this is standard practice. © 2013 Royal College of Obstetricians and Gynaecologists
Of course all live-born anencephalic infants born in the UK die, having death certified using cardiorespiratory criteria. Practically, around half of these babies have inadequate brainstem formation and do not breathe adequately to support organ function after birth, and half breathe adequately for a short period then die in the subsequent hours.25

**Donation**

**Anencephalic donation after brain death (DBD)**

Anencephalic DBD has been successfully undertaken leading to cardiac and renal transplantation,24 but has now stopped, mainly due to the difficulties in certifying any form of neurological death in these infants.

Organ donation from anencephalic infants following brain death surely needs contemporary review, though whether the UK position on this following the organ retention scandal at Alder Hey Children’s Hospital would be different from elsewhere is unclear, however given current appropriate deference to parental autonomy in this area this is potentially possible.

As already highlighted, neonatal organ donation does not currently occur in the UK, though such practice is routine in mainland Europe, Canada, the USA and Australia. This has led to the anomaly of UK cardiac transplant teams retrieving organs from European infants to transplant into British babies, whilst babies dying in the UK cannot donate organs, irrespective of parental wishes because of neonatology practices.23 As part of a current ethical address of this by the Royal College of Paediatrics and Child Health’s Ethical and Law Advisory Committee and the Academy of Medical Royal College Donation Ethics Committee, amongst others, the entire topic of an agreed UK viewpoint on anencephalic DBD warrants attention.

**Anencephalic donation after circulatory death (DCD)**

Over the past decade, the shortage of organs for transplantation has led to a resurgence of DCD, whereby organ donation can occur within a short interval of death certified by cardiorespiratory criteria. As the circulation has been interrupted prior to death the organs are inevitably exposed to a period of warm ischaemia, which if prolonged or exacerbated by antecedent significant hypoxia and/or ischaemia during the dying process, can preclude successful transplantation. Consequently, most DCD donation occurs after elective withdrawal of organ support within the intensive care unit, and is standard in both adult and paediatric intensive care units. Importantly, fewer organs are currently transplantable following DCD, with no cardiac donation occurring in the UK. In fact, this is not undertaken in adults, though of course was involved with Barnard’s first cardiac transplant,26 but has recently been successfully reported in three infants in the US.27 For some, the ethical complexity of donating the heart after certifying death using circulatory criteria has been an issue,28 although death is surely a uniform state, however it is verified. There also remains significant concern about tolerable warm ischaemic times for successful DCD cardiac transplantation.

It is worth considering whether at parents’ request anencephalic infants might be managed in such a way to facilitate DCD. While this avoids any controversy about brain death certification, there are a number of practical steps, and indeed ethical considerations.

One initial ethical challenge is the need to provide organ support to such an infant at delivery. Currently, anencephalic infants are not intubated and ventilated at delivery, as this is considered futile in that it merely prolongs an inevitable dying process. The clinical standard is that children are managed in what is determined to be their ‘best interests’. For dying children, as with adults, it is rarely considered that attempts to provide invasive – and so burdensome – organ support is an appropriate intervention. However, if the parents of an anencephalic infant ask for ventilation of their baby to try to help another dying infant, surely this is compatible with the legal standard that parental responsibility permits parents to make decisions for their children, as they are considered the most likely to decide in their child’s best interests, unless their decision is overtly harmful to the child. While anencephalic children are not considered to be dead, it is unclear what degree of pain or suffering they could ever appreciate without a neocortex, especially given appropriate analgesia. Therefore, it is also unclear what harm would be caused to them. Furthermore, given extended best interests appropriately advanced by the courts29 with weight given to social and cultural best interests, not just limited medical best interests, familial altruism is potentially an overriding determinant in these cases.

Given the recent success of infant cardiac DCD it is worth exploring the limits of parental consent and whether an anencephalic infant might be practically and ethically managed in such a way that they may have intensive care support provided at delivery, and then withdrawn and their death certified in such a way that facilitated organ donation.

**Alder Hey and relevance**

Due to the controversy at Alder Hey and other children’s hospitals involving organ retention from children who had died without parents being aware or consenting the Human Tissue Act20 has changed UK practice. Whilst organ donation was not involved in this controversy, the subsequent legislation ensures that UK parents have unprecedented control over where and how organs and tissues from children that die are ultimately disposed. It cannot be logical to allow a family to receive their child’s liver after autopsy to their home address, yet not to permit another family to donate a healthy organ from their dying child to another person.
The future

Clearly the current guidance suggesting that irreversible apnoea in a newborn with known anencephaly equates to death is unsustainable, not least because irreversible apnoea remains undefined and practically challenging. Ventilation of anencephalic infants at delivery to permit more leisurely diagnosis of a neurological state compatible with death, akin to brain death, is feasible but internationally has been largely rejected.5 If the Rubicon of ventilating such infants (not in their narrow medical best interests, but in some extended best interests ethically supported by altruistic parental views), can be crossed, then DCD of heart and kidneys has already been successful, and liver (or hepatocyte), lung or other organs may be feasible. Indeed the FIGO Committee for the Ethical Aspects of Human Reproduction and Women’s Health has recently stated that:

- The purpose of organ donation constitutes an ethical ground for a woman to choose to maintain an anencephalic pregnancy. Counseling of women and couples regarding organ donation should be undertaken by persons with no conflict of interest.
- When an infant is born with signs of life but has no forebrain (anencephaly) and hence has no prospect of survival, with parental permission, the child may be placed on a ventilator for the purpose of organ donation following natural death. Any local legal definition of death is binding, but it may have to be reviewed in the light of scientific development of criteria concerning brain death in neonates.31

Conclusion

Organ donation from anencephalic infants does not currently occur in the UK, or indeed worldwide. However, national guidance does support such donation but clearly needs contemporary review given the events since it was provided. Internationally, the key ethical issue has been the determination of death before donation in anencephalic infants using neurological criteria, and it is generally accepted that this is not possible. However, DCD might be realised in such cases, though this requires research and new national guidance. Obstetricians approached by women carrying an anencephalic fetus should contact their local senior nurse in organ donation, though the advice is likely to be that organ donation is not currently possible, but that heart valve donation after death may be undertaken. Any other donation, such as stem cells or hepatocytes must be under the auspices of formally reviewed research projects.

Disclosure of interests

JB is the clinical lead for organ donation at Great Ormond St Hospital.

References

9 Academy of Medical Royal Colleges. A Code of Practice for the Diagnosis and Confirmation of Death. London: Academy of Medical Royal Colleges; 2008
Organ donation in fetal anomaly


