

Chapter

6

Dilemmas in current
practice: babies
needing intensive care

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Introduction

6.1 We have seen in the preceding chapters that in the UK, current practice rests on a consensus that if the outcome for a baby with a serious condition is uncertain after birth, life support and full intensive care should be instituted until the prognosis becomes clearer and the situation re-evaluated. This stage may be reached when the results of investigations are known, or after a period when the clinical situation changes, or even when the baby's parents have had some time to adjust to a diagnosis and prognosis. In this chapter, we focus on the very serious conditions that can lead parents and healthcare professionals to begin discussing whether intensive care should be continued or be withdrawn. Some babies in intensive care may have started life at the borderline of viability. Others may have been born later, but with serious health problems. The relief of discomfort and pain is an important consideration, whether during intensive care or as part of palliative care when intensive care treatment has been replaced with other forms of care. As before, we use hypothetical examples to illustrate the dilemmas that parents and professionals face in making decisions when a poor prognosis for a baby has been established.¹ We highlight some economic issues relating to critical care decisions after birth, although we have not been able to find sufficient data to compare the economic costs of outcomes for children who start life with different conditions, as we would have wished. Finally, we discuss the importance of data collection and analysis to help reduce uncertainty in the prediction of health outcomes.

The clinical perspective

- 6.2 In the past, debates about critical care decisions in the newborn have focused on whether babies with specific congenital malformations should or should not have life-saving surgery or life support. In 1973, 14% of the deaths in the neonatal intensive care unit in New Haven, USA, followed decisions to withdraw treatment.² Most of these babies had severe congenital abnormalities. Subsequently, debates about withdrawal of treatment and substitution with other forms of care have broadened to include babies with acquired brain injury and premature babies at the borderline of viability. In the UK and the Republic of Ireland in 1995, among extremely premature babies, approximately half of deaths were classed as "active withdrawal of intensive care",³ and experience would suggest that the proportion is now higher.⁴
- 6.3 The situations in which parents and clinical teams come to the point of deciding whether to withdraw life-sustaining treatment, after an initial decision to begin resuscitation and other forms of treatment, fall into three broad categories:
- (a) A baby for whom intensive care is proving futile, in that death appears inevitable. In these cases, intensive care serves only to extend the process of dying.

¹ We use examples that are representative of what occurs in hospital. They are not based on clinical cases. In the discussion of each example, issues are highlighted, some of which were drawn to the attention of members of the Working Party during fact-finding meetings. We acknowledge that the choice of the issues that we discuss after each example may influence how the examples themselves are perceived by different readers, depending upon the reader's own worldview.

² Duff RS and Campbell AG (1973) Moral and ethical dilemmas in the special-care nursery *N Engl J Med* **289**: 890–4.

³ Costeloe K, Hennessy E, Gibson AT, Marlow N, Wilkinson AR and the EPICure study group (2000) The EPICure study: outcomes to discharge from hospital for infants born at the threshold of viability *Pediatrics* **106**: 659–71. The majority of deaths took place in the first seven days after admission to a neonatal intensive care unit.

⁴ Personal communication to the Working Party from Professor Neil Marlow and Professor Andrew Whitelaw.

- (b) A baby who has suffered a severe brain injury and for whom there appears to a very high risk of severe disability as he or she grows up.
- (c) A baby who is discovered to have a serious malformation, dysplasia (abnormal development of tissues or organs) or a genetic condition with a serious outcome for which there is no treatment.⁵

The first example may be considered a judgement about the timing and mode of death in terms of how care is provided. The two other examples highlight the uncertainty of outcome that complicates decision making in cases involving many neonatal conditions. We begin by considering brain injuries, which can be acquired during pregnancy or close to the time of birth and which are a major cause of later disability.

Brain injuries in premature and term newborn babies

6.4 There is an extensive body of knowledge about the origin, diagnosis and prognosis of brain injury in newborn babies. Advances in diagnostic methods mean that it is now possible to identify the nature, location and extent of brain injury even in the smallest newborn baby (see Appendix 4 for details of common types of brain injury). A range of investigations may be carried out, depending on the circumstances, and the availability of facilities. For example, a baby may have to be transported some distance to another hospital to have magnetic resonance imaging (MRI). Severe brain injuries may be manifested differently in premature and term infants, which means that understanding the predictive value of the various tests is crucial for both parents and professionals who have to make decisions about a baby in intensive care.

The premature baby with brain injury

- 6.5 In the premature baby, brain injury usually occurs in two particularly vulnerable tissues:
- (a) **The white matter** is found in an area deep inside the brain which contains mainly nerve fibres, including those that allow the brain to control movement. As the white matter surrounds the ventricles, injury within it is called periventricular leucomalacia (literally softening of the white matter around the ventricles).
 - (b) **The germinal matrix** comprises fragile tissue that lies in the base of the fluid-filled spaces or ventricles found deep in the brain. Illness in the fetal or neonatal period can lead to bleeding in this tissue. Such bleeding usually resolves without serious problems later. However, a minority of cases are complicated either by hydrocephalus (literally 'water on the brain') due to blockage of the outflow of spinal fluid from the ventricles, causing them to enlarge, or by causing adjacent areas of brain tissue to die by obstructing their blood flow (known as infarction).

⁵ Four types of birth defect are recognised: **(1) Deformations:** resulting from abnormal mechanical forces acting to distort an otherwise normal structure. These often occur quite late in gestation after normal initial formation of organs, but the growth and subsequent development of these organs or structures is hampered by the mechanical force. **(2) Disruptions:** defects caused by destruction of previously normal tissue; sometimes consequent on haemorrhage or poor blood flow during development to a particular region of the developing fetus. Disruptional abnormalities generally affect several different tissue types within a well-demarcated anatomical region. **(3) Dysplasias:** abnormal cellular organisation or function within a specific tissue type throughout the body, resulting in clinically apparent structural changes; for example a skeletal dysplasia, resulting in 'dwarfing' where the patient's short stature is caused by a major gene mutation causing a dysplasia of the cartilage, with the result that the bones do not elongate. **(4) Malformations:** abnormalities caused by failure of the embryonic process; here the development of the particular tissue or organ is arrested, delayed or misdirected, causing permanent abnormalities of the structure which prevents normal development. Many malformations are the result of genetic mutations and can affect several different body systems causing a range of different clinical signs of birth defects in the individual patient. Unlike deformations and disruptions, malformations suggest an error occurring very early in gestation, either in tissue differentiation or during the development of individual organ systems. For simplicity, throughout this Report we chose to use the more general term **abnormality** for all types of defect.

- 6.6 Although all three types of brain injury described above (leucomalacia, hydrocephalus and infarction) are associated with later disability, the extent of injuries can vary greatly between different babies, and many children with these conditions grow up free from significant problems. Trying to decide if a lesion is likely to cause disability is difficult, unless the lesion is extensive. Generally, periventricular leucomalacia or infarction which occurs in the frontal part of the brain is not associated with serious disability, but where it occurs at the back, it is much more likely to be associated with cerebral palsy. Many lesions are situated centrally and these are difficult to assess. Thus a prognosis is rarely absolutely certain; it is a matter of risk which must be communicated to the parents to help them reach a decision.
- 6.7 Extensive lesions of the brain that have a more certain adverse prognosis tend to become evident earlier in a baby's postnatal course. This coincides with the period when babies are receiving maximal intensive care and when withdrawal of that support will almost certainly lead to death. The extent of less extensive lesions of the brain takes longer to become obvious. A secure diagnosis may not be made until the period when a baby requires maximal intensive care has passed, and often when he or she may no longer require intensive care at all. For example, hydrocephalus is not usually evident until after three weeks of age and the cysts that accompany serious periventricular leucomalacia take two to three weeks to develop. By this stage, generally a baby would no longer be receiving intensive care support.

The full term baby who acquires brain injury

- 6.8 Sometimes a baby acquires brain injury as a result of a period of inadequate oxygen supply during labour or birth. This deprivation of oxygen is known as intrapartum hypoxia, or more commonly 'birth asphyxia'. After birth, a baby with this type of brain injury passes through a well-described sequence of clinical phases. The set of neurological symptoms (neurological syndrome) is termed an 'encephalopathy'.⁶ As postnatal encephalopathy can sometimes be due to problems other than intrapartum hypoxia, it requires careful evaluation.
- 6.9 The clinical phases of postnatal encephalopathy may be categorised using a grading system first described by Sarnat and Sarnat in 1975.⁷ This system allows for three categories of clinical syndrome to be defined: mild, moderate and severe:
- All babies whose category is 'mild' universally recover and long-term consequences have not been reported.
 - As with very premature babies, the 'moderate' category is more difficult to define. Around 20% of those whose clinical grade is moderate will die or develop serious disability. The remaining 80% are generally considered as having the potential to do well, although there is good evidence that in some surviving children, significant learning and behavioural problems will arise at school age.⁸
 - Babies who develop a 'severe' encephalopathy and who have not improved to a moderate grade by 48 hours either do not survive or almost always are seriously disabled. This disability usually comprises spastic or athetoid cerebral palsy⁹ affecting all limbs and trunk,

⁶ When the encephalopathy results from intrapartum hypoxia, it is sometimes termed hypoxic ischaemic encephalopathy.

⁷ Sarnat HB and Sarnat MS (1975) Neonatal encephalopathy following fetal distress *Arch Neurol* **33**: 696–705.

⁸ Robertson CM and Finer NN (1993) Long-term follow-up of term neonates with perinatal asphyxia *Clin Perinatology* **20**: 483–500; Marlow N, Rose AS, Rands CE and Draper ES (2005) Neuropsychological and educational problems at school age associated with neonatal encephalopathy *Arch Dis Child Fetal Neonatal Ed* **90**: F380–7.

⁹ Spastic cerebral palsy resulting from hypoxia is usually caused by a relatively prolonged period of partial hypoxia. In cases where the baby suffers a short period of near total lack of oxygen, as opposed to a longer period of partial hypoxia, the resulting disability will be athetoid cerebral palsy, which may be accompanied by other physical and learning disabilities, although intellect may be preserved. These babies do not always have severe neonatal encephalopathy.

severe learning disabilities, epilepsy, and severe vision and hearing impairments. These are among the most severely disabled children encountered and doctors would normally advise discontinuation of active treatment after 48–72 hours of severe encephalopathy. Infants with this condition have lost their responsiveness and consciousness and are usually, but not always, ventilator-dependent, in which case withdrawal of ventilation will result in the infant dying within minutes or hours. It is therefore critically important that the diagnosis is accurate and a careful assessment made of other possible causes for a baby's encephalopathy, including biochemical disturbances, infection and congenital anomaly (see below).

Many of the investigations that are needed to identify the nature of the injury (Appendix 4) and clarify the prognosis take time. The period when it would be possible to withdraw treatment may pass (usually the first three days) while these tests are carried out, by which time the life of the baby may no longer be dependent on technological support.

The baby born with brain abnormalities

6.10 Although rare, babies can be born with a structural abnormality of the brain that has not been recognised before birth, with evidence of a brain injury that occurred at some time before labour, or with a condition where the prognosis for normal development is well established and known to be very unlikely. These are known as congenital¹⁰ conditions. Examples include:

- A child with **polymicrogyria**, which arises from abnormal organisation of brain cells very early on in gestation; children show poor body and brain growth, and have severe cerebral palsy.¹¹
- A twin who has survived the death of his or her co-twin at mid-gestation but at the time suffered a **major failure of the blood supply** to the whole of one side of the brain, usually the left hemisphere; these babies will develop a severe cerebral palsy.
- A child born with **trisomy 13** (also known as Patau syndrome) which is a genetic condition in which three copies of chromosome 13 are in each cell instead of the usual two. Trisomy 13 is usually associated with other serious abnormalities and learning disabilities. The majority of children (80–90%) with this syndrome do not survive infancy, and long-term survivors have not been identified.

6.11 Each child with brain abnormalities is given a careful individual assessment with a range of diagnostic procedures to establish the nature of the condition, the extent of any damage and the prognosis (Appendix 4). Doctors will discuss the option of withdrawing intensive care with the family in cases where a baby is severely affected and dependent on technological support and would die without it.

Other serious conditions in the newborn baby

6.12 Life-threatening conditions other than acquired brain injury or congenital brain abnormalities can arise. These conditions require careful individual assessment prior to any decision making. For example, a baby might have an abnormality in a major organ system that was present at birth or an acquired injury, either of which makes long-term survival or independence almost impossible. This group of conditions may affect the heart, lungs, bowel or kidneys. For example, complex congenital heart abnormalities are often considered inoperable and parents may decide not to subject their child to the pain and distress of repeated episodes of surgery

¹⁰ Congenital literally means 'born with'.

¹¹ See also footnote 9.

to no benefit in the long term. Some congenital abnormalities of the lung may become obvious during intensive care, such as alveolar capillary dysplasia where gas exchange is difficult because the blood vessels in the developing lung fail to grow in proportion to the airways. In all known cases, babies with this form of dysplasia have died. A lung biopsy can be performed in an affected baby, and intensive care may subsequently be withdrawn and substituted with palliative care (see paragraphs 6.18–6.22) in confirmed cases. However, most diagnoses are made after death, usually following an autopsy (see paragraph 6.51).

6.13 In some babies, acquired injuries to the bowel pose difficult dilemmas. Babies may be found to have too little remaining gut after lengths of intestine die such as when necrotising enterocolitis (a disease affecting the wall of the newborn gut) is complicated by peritonitis, an infection that occurs when the intestine perforates and spills its contents into the abdominal cavity. A similar situation can arise when the blood supply to the gut has been compromised because the bowel twists on its supporting membrane (mesentery) which contains the blood supply. In these situations a baby may be left with less than the minimum length of bowel known to be able to regrow and support feeding. Infants with these conditions require life-long intravenous feeding to survive and many develop serious side effects and have a shortened life expectancy.¹² Bowel transplants have recently become an option in such cases. However, an affected baby would need to take immunosuppressant drugs for the rest of his or her life, meaning he or she would often be susceptible to infections. Following surgery, the incidence of illness and death remains high.¹³ Difficult dilemmas also arise when babies are born with kidney failure. To survive, these babies must begin dialysis (which is technically difficult in the newborn) to await the age when a kidney transplant may be feasible. Critical care decisions are difficult in such cases because the prognosis is poor, yet a baby may not be dependent on a ventilator. In contrast to withdrawing ventilation, which would very quickly lead to a baby's death, withholding or withdrawing intravenous nutrition (see paragraph 6.22) or dialysis may mean that a baby takes several days to die, during which time he or she should receive palliative care. We say more about methods for relieving discomfort and pain, and palliative care in the next section.

Pain relief and developmental care in the neonatal intensive care unit

6.14 While neonatal intensive care saves the lives of many babies, it can be a stressful experience for them, especially when given for long periods. Many of the procedures that are frequently performed cause discomfort or pain, such as passing a tube into the windpipe, inserting fine tubes, or cannulae, into a baby's arteries and veins, and taking blood samples. A recent study in a neonatal intensive care unit in the Netherlands found that babies were subjected to an average of 14 painful procedures per day.¹⁴ In addition, the environment, often noisy and brightly lit, can be stressful for babies. Parents of babies receiving intensive care may also find it stressful, and concern about a baby experiencing pain has been identified as one factor contributing to stress.¹⁵

6.15 Recent research has provided increasing evidence that newborn babies, including those born prematurely, show strong responses to pain¹⁶ and that experiencing painful procedures without pain relief during the neonatal period is harmful, with potential both for short- and

¹² Kato T, Tzakis AG, Selvaggi G *et al.* (2006) Intestinal and multivisceral transplantation in children *Ann Surg* **243**: 756–64.

¹³ *Ibid.*

¹⁴ Simons SHP, van Dijk M, Anand KS, Roffthoof D, van Lingem RA and Tibboel D (2003) Do we still hurt newborn babies? *Arch Pediatr Adolesc Med* **157**: 1058–64.

¹⁵ Franck L, Cox S, Allen A and Winter I (2004) Parental concern and distress about infant pain *Arch Dis Child Fetal Neonatal Ed* **89**: F71–5.

¹⁶ Slater R, Cantarella A, Gallella S *et al.* (2006) Cortical pain responses in human infants *J Neurosci* **26**: 3662–6.

long-term effects.¹⁷ These effects can include a permanently altered response to pain.¹⁸ In the past, infant pain has been poorly understood and under-treated,¹⁹ but a better understanding of the risks of pain has led to research on how to observe infants for signs of pain, and how to give pain-relieving medicines safely.²⁰ Nevertheless a survey of practice found that pain is not monitored routinely and pain relief not used widely in neonatal units in the UK, and that there is considerable variation in practice.²¹

6.16 Pain can be difficult to recognise in an ill or premature baby, and effective methods for monitoring his or her level of pain using behavioural and physiological signs are not available. Several assessment tools have been developed²² and guidelines from the Royal College of Nursing (RCN) recommend that assessment of pain should include the use of a suitable method along with consideration of a baby's health status and parental views.²³ We note that other organisations have developed guidelines for clinical practice in the UK and internationally.²⁴ While assessing a baby's discomfort or pain is difficult, deciding which treatments are most effective and with least risk is even more challenging. Many pain-relieving medicines given to adults and children can be given to babies; however, they all have disadvantages, mainly related to the way they are administered or to adverse effects. For example, some medicines that can only be given by mouth cannot be used if a baby is not able to feed. Strong analgesics such as morphine, like other medicines, may carry some risks for a baby's development if they are used for a prolonged period. There is currently little research on whether the potential developmental risks from unrelieved pain outweigh those from prolonged exposure to medicines that act on the central nervous system. New analgesics are being sought, but there is no clear consensus about which of many potential drug targets in biological pain pathways should be pursued.²⁵ Moreover it may be many years before new medicines are tested in babies. Alternative means of pain relief can be used in conjunction

¹⁷ Research in animals suggests that, in the longer term, exposure to multiple painful experiences in the newborn period leads to more sensitivity to pain from major injury and less sensitivity to minor injury. Pain in early life is also associated with later abnormalities in the stress response system: see Fitzgerald M (2005) The development of nociceptive circuits *Nat Neurosci Rev* 6: 507–20. However, much less is known about how neonatal pain might affect human children or adults. Some clinical studies suggest that untreated pain suffered in early life may affect stress hormone responses in childhood or alter how children experience pain or other sensations: see Andrews K and Fitzgerald M (2002) Wound sensitivity as a measure of analgesic effects following surgery in human neonates and infants *Pain* 99: 185–95; Grunau RE (2002) Early pain in preterm infants A model of long-term effects *Clin Perinatol* 29: 373–94; Grunau RE, Weinberg J and Whitfield MF (2004) Neonatal procedural pain and preterm infant cortisol response to novelty at 8 months *Pediatrics* 114: e77–84; Peters JWB, Schouw R, Anand KJS (2005) Does neonatal surgery lead to increased pain sensitivity in later childhood? *Pain* 114: 444–54; Schmelzle-Lubiecki B *et al.* (2005) Abstracts: 11th World Congress on Pain (Seattle: IASP Press), pp 76–7.

¹⁸ Fitzgerald M (2005) The development of nociceptive circuits *Nat Rev Neurosci* 6: 507–20.

¹⁹ International Association for the Study of Pain (2005) Why children's pain matters *Pain – Clinical Updates* Vol. XIII, No. 4, available at: <http://www.iasp-pain.org/PCU05-4.pdf>, accessed on: 27 July 2006.

²⁰ *Ibid.*

²¹ The survey found that pain scales which can be used to assess pain and an infant's responsiveness were used relatively little and that less than 60% of units regularly used analgesia for pain relief. See Redshaw M and Hamilton K (2005) *A Survey of Current Neonatal Unit Organisation and Policy: Commissioned by BLISS – The Premature Baby Charity* (Oxford: National Perinatal Epidemiology Unit), available at: http://www.npeu.ox.ac.uk/neonatalunitsurvey/neonatalunitsurvey_downloads/BLISS%20Final%20Report.pdf, accessed on: 18 July 2006.

²² Some of these tools are compared in: Royal College of Nursing Institute (1999) *Clinical Guidelines for the Recognition and Assessment of Acute Pain in Children – Technical Report* (London: RCN).

²³ *Ibid.*

²⁴ Summarised in International Association for the Study of Pain (2005) Why children's pain matters *Pain – Clinical Updates* Vol. XIII, No. 4, available at: <http://www.iasp-pain.org/PCU05-4.pdf>, accessed on: 27 July 2006. In addition see Anand KJ and the International Evidence-Based Group for Neonatal Pain (2001) Consensus statement for the prevention and management of pain in the newborn *Arch Pediatr Adolesc Med* 155: 173–80; The Royal Australian College of Physicians (2005) *Guideline Statement: Management of procedure-related pain in neonates* (Sydney: The Royal Australian College of Physicians).

²⁵ Basbaum AI (2005) The future of pain therapy: something old, something new, something borrowed, and something blue, in *The Paths of Pain, 1975–2005*, Merskey H, Loeser JD and Dubner R (Editors) (Seattle: IASP Press), pp 513–32.

with analgesics; they include giving breast milk or a dummy, and using touch, warmth and positioning of a baby's body. Such interventions have been found to provide non-painful stimulation that activate the body's own pain-relieving chemicals or divert attention from pain.²⁶

- 6.17 Decreasing the stress of the neonatal care environment for babies is another evolving area of practice. One approach that has been found to have some benefit is 'developmental care'. This comprises a broad spectrum of interventions intended to reduce the stress of the neonatal intensive care unit through modification of the care-giving environment and processes of care giving to make them more appropriate to a baby's stage of development. Developmental care uses a range of strategies, including reduced lighting, noise and handling, and non-invasive monitoring. Parents are encouraged to become more involved in the care of their babies by helping them to recognise their baby's signals and responses, and to use interventions such as baby massage and kangaroo (skin to skin) care when their baby is well enough. Research studies have shown that developmental care leads to improved growth and bone development and has reduced some adverse health outcomes, although further studies on the scale and consistency of the benefits are needed.²⁷ **The Working Party considers that the reduction of pain and stress in neonatal units is an important area for improvement in clinical practice and that more needs to be done to apply current knowledge about how to assess, prevent and treat pain for babies receiving intensive care.** Our view is that research into the potential developmental effects of neonatal pain and stress and their treatments should be encouraged.

Palliative care

- 6.18 It will not always be appropriate to continue intensive care for all babies who are seriously ill. If they have serious life-threatening or life-limiting health problems and parents and doctors agree that they are unlikely to benefit either from the initiation or continuation of intensive care, they may instead receive another form of support, known as palliative care. This is the "active, total care of patients whose disease is not responsive to curative treatment. The goal of palliative care is achievement of the best quality of life for patients and their families."²⁸ The relief of pain and other distressing symptoms caused by disease or medical treatment is the primary focus of palliative care, together with psychological, social and spiritual support to assist patients and their families at the end of the patient's life. Modern palliative care began in the 1960s within the hospice movement in the UK.²⁹ More recently the importance of the implementation and development of palliative care specifically for children has been highlighted.³⁰
- 6.19 Palliative care involves a multidisciplinary team with specialist skills, including medical and nursing staff, staff from children's services, counsellors and voluntary organisations, and can be provided in a hospice, at home or in a hospital setting. Palliative care for newborn babies is indicated in three situations: (a) when a fetal abnormality known to cause death is diagnosed prenatally, and the fetus is born alive;³¹ (b) when a decision is made in the delivery

²⁶ Franck LS and Lawhon G (1998) Environmental and behavioral strategies to prevent and manage neonatal pain *Semin Perinatol* **22**: 434–43; Leslie A and Marlow N (2006) Non-pharmacological pain relief *Semin Fetal Neonatal Med* **11**: 246–50.

²⁷ Kenner C and McGrath J (Editors) (2004) *Developmental Care of Newborns and Infants: A guide for health professionals* (Mosby).

²⁸ For the World Health Organization definition of palliative care, see: <http://www.who.int/cancer/palliative/definition/en/>, accessed on: 31 July 2006; Gale G and Brooks A (2006) Implementing a palliative care program in a newborn intensive care unit *Adv Neonatal Care* **6**: 37–53.

²⁹ Saunders C (1996) Into the valley of the shadow of death: A personal therapeutic journey *Br Med J* **313**: 1599–601.

³⁰ Association for Children with Life-threatening or Terminal Conditions and their Families and Royal College of Paediatrics and Child Health (1997) *A Guide to the Development of Children's Palliative Care Services* (Bristol and London: Association for Children with Life-threatening or Terminal Conditions and their Families and Royal College of Paediatrics and Child Health).

³¹ Breeze AC, Lees CC, Kumar A, Missfelder-Lobos HH and Murdoch EM (2006) Palliative care for prenatally diagnosed lethal fetal abnormality *Arch Dis Child Fetal Neonatal Ed* doi:10.1136/adc.2005.092122.

room that it is not in baby's best interests to be resuscitated; or (c) when it is decided that intensive care is or has become futile and treatments should be withdrawn or withheld.

6.20 Guidance from the RCPCH on this subject reads: "Withholding or withdrawing life sustaining treatment does not imply that a child will receive no care. It should rather signal a change in focus towards palliative care making sure that the rest of the child's life is as comfortable as possible."³² BAPM guidance further states: "infants for whom life-sustaining support is withdrawn or withheld should continue to be kept warm, offered oral nourishment, and treated with dignity and love (comfort care). Their parents should be encouraged to be with their child as much as possible. They should be given every support during this distressing time."³³ The Department of Health and Department for Education and Skills has also recognised the value of this type of care, by including the following in the National Service Framework (NSF) for Children: "High quality palliative care services should be available for all children and young people who need them."³⁴

6.21 We understand, however, that healthcare professionals working in perinatal and neonatal intensive care do not receive mandatory training in palliative care and that access to teams who specialise in palliative care is extremely limited. The current use of techniques in palliative care for management of pain and symptoms in babies, and the availability of support for parents, vary greatly across different settings in the UK. **The Working Party therefore recommends that the NHS should train all professionals working in neonatal medicine in the basic principles of palliative care so that these can be applied when a need is identified. To complement this provision, the NHS should also facilitate access to specialist advice in palliative care for difficult cases in the same way that specialists would be consulted on complex problems in other areas of medicine.** It may be useful to draw upon examples from other countries of different ways in which comprehensive palliative care can be provided to babies and their families.³⁵ Meanwhile, the American Academy of Pediatrics has recommended minimum standards for paediatric palliative care, including multidisciplinary training, expert palliative care assistance to be available at all times, in-patient facilities and community outreach programmes.³⁶

6.22 The BAPM guidance quoted in paragraph 6.20 above specifically states that nourishment by mouth should continue to be offered when intensive care is withheld or withdrawn.³⁷ Newborn babies all require feeding by others, and in the UK, feeding by a stomach tube for a baby who cannot suck is often considered as basic nursing care, and not medical treatment.³⁸ In many cases hunger and dehydration may add to a baby's suffering, and artificial

³² Royal College of Paediatrics and Child Health (2004) *Withholding or Withdrawing Life Sustaining Treatment in Children: A framework for practice*, 2nd Edition (London: RCPCH), available at: http://www.rcpch.ac.uk/publications/recent_publications/Withholding.pdf, accessed on: 25 Sept 2006.

³³ British Association for Perinatal Medicine (2000) *Fetuses and Newborn Infants at the Threshold of Viability: A framework for practice* available at: <http://www.bapm.org/documents/publications/threshold.pdf>, accessed on: 31 July 2006.

³⁴ Department for Education and Skills and Department of Health (2004) *National Service Framework for Children, Young People and Maternity Services: Disabled children and young people and those with complex health needs* (London: Department of Health), available at: <http://www.dh.gov.uk/assetRoot/04/09/05/56/04090556.pdf>, accessed on: 19 July 2006.

³⁵ Gale G and Brooks A (2006) Implementing a palliative care program in a newborn intensive care unit *Adv Neonatal Care* **6**: 37–53; Craig F and Goldman A (2003) Home management of the dying NICU patient *Semin Neonatol* **8**: 177–83; Stringer M, Shaw VD and Savani RC (2004) Comfort care of neonates at the end of life *Neonatal Netw* **23**: 41–6.

³⁶ American Academy of Pediatrics (2000) Palliative care for children *Pediatrics* **106**: 351–7.

³⁷ British Association for Perinatal Medicine (2000) *Fetuses and Newborn Infants at the Threshold of Viability: A framework for practice* available at: <http://www.bapm.org/documents/publications/threshold.pdf>, accessed on: 31 July 2006.

³⁸ Under UK law a distinction (in adults) has been made between artificial nutrition as a medical treatment and as a non-medical procedure when considering the permissibility of withdrawing artificial nutrition and hydration. For example, withdrawal of artificial nutrition and hydration has been held lawful in the UK in most cases in which adult patients are in PVS (a persistent vegetative state) or what has been described as near-PVS. In the case of the young man Anthony Bland (see *Airedale NHS Trust v Bland* [1993]

nutrition and hydration would need to be continued if this suffering is to be avoided. However, we also recognise that if there is a failure of gastrointestinal function as a baby dies or if their gut is absent or damaged through disease, providing food and liquid may not be appropriate and could cause additional suffering. **We therefore conclude that oral nutrition and hydration should only be withheld from a baby in the exceptional circumstances when providing it causes discomfort and pain, for example when the baby has little functioning bowel or when death is imminent. The decision should only be taken after careful assessment and as part of a planned programme of palliative care designed to minimise suffering and make the baby as comfortable as possible.**

Some examples³⁹

6.23 We consider below several different hypothetical examples to highlight some of the complexities of making decisions about treatment when the prognosis for a baby has been established. The first two examples illustrate situations in which either the doctors or the parents would like intensive care to continue while the other party disagrees. The third example describes how the use of pain relief can create a dilemma.

Case 6: Danielle – continuation of intensive care on the advice of doctors

Danielle

Danielle was the third child in her family and was born at a gestational age of 27 weeks. Although early scans suggested that she had periventricular brain lesions, these seemed to resolve. She remained at risk of cerebral palsy which made her prognosis uncertain. Danielle did not need ventilator support after the first few days following her birth. On day 24 she began receiving air through a continuous positive airway pressure (CPAP) mask to her nose because her breathing had begun to stop for short spells (apnoea). Ultrasound scans showed bilateral cystic periventricular leucomalacia, an injury to the white matter surrounding the ventricles which indicated the probable development of cerebral palsy (see Appendix 4). Danielle's parents wished to withdraw CPAP support and provide Danielle with palliative care only. They said they could cope with physical disability alone but not with mental disability. The doctor advised that her prognosis remained uncertain and that she was unlikely to die if CPAP was withdrawn. However, she might become more distressed because extra handling would be needed to curtail her apnoeic spells. He explained that it was not certain that Danielle would have serious learning disabilities in the future although she was expected to have great difficulty with sitting and walking. She would benefit from specialist treatment and support for cerebral palsy. Life support was continued and Danielle survived. At three years old Danielle had severe spastic diplegia, a form of cerebral palsy affecting the legs and mild cognitive impairment.

Best interests

6.24 In Chapter 2 we explained that doctors are under a professional obligation to preserve life where and when they can, using appropriate treatment to achieve that end. However, they are not obliged to provide life-sustaining treatments when to do so would be futile. In cases like that of Danielle, it would generally be regarded as in a baby's best interests for clinicians to continue full intensive care while the prognosis remains uncertain. For Danielle, by the time the diagnosis became clearer, she was no longer on full ventilatory support, so the option of ceasing to maintain her life by withdrawing that support had passed. In deciding to continue CPAP treatment, the doctor was using his professional judgement to act in Danielle's best interests (paragraphs 2.21–2.32). However, the parents signalled their own best interests, when they asked for the implications for themselves and other family members to be

1 All ER 821) "the court considered there was overwhelming evidence that the provision of artificial feeding by means of a nasogastric tube was 'medical treatment' and that its discontinuance was in accord with good medical practice". See Mason JK and Laurie GT (2005) *Mason and McCall Smith's Law and Medical Ethics*, 7th Edition (Oxford: Oxford University Press), p 580. The situation for paediatric practice is unclear (Royal College of Paediatrics and Child Health (2004) *Withholding or Withdrawing Life Sustaining Treatment in Children: A framework for practice*, 2nd edition (London: RCPCH)). In one case the courts have sanctioned not providing artificial feeding for a baby Re C (a minor) whose death was imminent and inevitable (wardship: medical treatment) [1989] 2 A11 ER 782 (Mason & Laurie, p 549).

³⁹ See footnote 1.

considered. We noted that there are different kinds of interest, each of which should usually be given some weight when making decisions. In an analysis based on best interests, the conflict arises not because the parents and the doctor disagree about what is best for Danielle, but because her parents see their interests as outweighing Danielle's basic interests in being alive, with the doctor deciding that Danielle's interests are the ones that should be promoted. If intolerability rather than best interests were to be used as the basis for decision making, a similar outcome would probably arise, since in Danielle's case, at the time that the decision was made, it did not seem likely that her later quality of life would be 'intolerable' (paragraph 2.16).

Conveying information

6.25 It is clear that the doctor's decision to act against the parents' wishes created tension at the moment of clinical decision making. Several factors might have affected the quality of decision making. The parents might have been given insufficient information about what it means to live with a person with physical and mental disabilities. If this was the case, they may have underestimated their capacity to care for a disabled child. Such an omission is important because their potential in this regard needs to be considered in the decision-making process. Advice should have been offered to Danielle's parents about the kinds of support and possibilities available for a family with a disabled child (see Chapter 7), including opportunities for respite care and adoption. The example of Danielle demonstrates that professionals conveying information must handle the stressful situation in which parents find themselves with great sensitivity. Danielle's parents may feel that the clinical team has not thought properly about what it will mean for their family to have a daughter with disabilities. Specialist training in the communication of sensitive issues might have helped the team to talk this or other issues through with them and identify any need for further information or counselling. In Danielle's case the doctor has the difficult task of balancing an appreciation of the concerns of her mother and father with the need to convey to them that in his view, they seem not to have given sufficient weight to Danielle's best interests.

6.26 For neonatal care, an important general question is whether any decisions that are made jointly by healthcare professionals and families are based on shared comprehension of critical choices, actions and terminology. Few common interpretations appear to exist even of terms such as 'withdrawal of treatment', 'futility', 'quality of life' or 'consent'. Understanding how these terms have been used, and the importance attached to them in the decision-making process will differ widely between different parties.⁴⁰ Research suggests that different interpretations are to be expected, especially in situations where the outcome is uncertain. It might be the case, for example, that it is precisely these often subtle differences in interpretation that lead to overt conflict between different parties and cause them to seek resolution in the courts. **We conclude that further research is needed to clarify how the different parties interact with each other to further understanding, to provide an evidence base for identifying and applying changes in practice, and for the more effective resolution of differences of opinion.**

Legal issues

6.27 Danielle's case is one that might have gone to court. Her parents could have challenged the doctor's decision and sought an order requiring withdrawal of intensive care. They would probably have maintained that the doctor acted unlawfully in continuing CPAP without either parental consent or authority from the court to act in what they believed was Danielle's best interests (see Box 6.1). However, if this case had been taken to court, the

⁴⁰ Nettleton S (1995) *The Sociology of Health and Illness* (Cambridge: Polity Press); Annandale E (1998) *The Sociology of Health and Medicine* (Cambridge: Polity Press).

judges would have been likely to support continuation of CPAP. Under the law, the likely burdens to Danielle of continuing treatment are outweighed by her interests in survival and the pleasures that life should afford her, notwithstanding her disabling condition. We note that the quality of Danielle's life will depend very much on her parents. Would they be able to argue in court for the greater say in deciding what constitutes her best interests? This case is one where the parents and the healthcare professionals might benefit from access to mediation services.

Box 6.1: Going to court

Where doctors and parents reach irreconcilable disagreement about what treatment a baby should receive, a court may be asked to decide what care is in the best interests of the baby. As well as best interests, the judge must consider broader social issues. Cases about treatment decisions are generally heard in the Family Division of the High Court and the proceedings are usually started by the NHS trust. Occasionally the family may bring Judicial Review proceedings, asking the Administrative Court (part of the High Court) to examine whether the decision (made by the doctors employed by the NHS trust) to treat (or more usually refuse treatment) was properly arrived at.

The person (or NHS trust) who commences the proceedings* is known as the 'claimant' and the other parties are called the 'defendants'. In the usual case where the trust is the claimant, the baby will be the first defendant and one or both parents and any other closely involved family members will be the second or subsequent defendants. The baby will be represented by the Children and Family Court Advisory and Support Service (CAFCASS) whose functions include not only representing the baby, but also providing information, advice and other support for the baby and his or her family. Nonetheless, the parents are likely to want their own legal representation. They will need to find a firm of solicitors who specialise in this work. Parents on low incomes may qualify for Legal Aid (now called 'Public Funding').

Though it may not appear so to the parents and the doctors involved, these court proceedings are not in the form of conventional adversarial litigation, but are a process to establish a baby's best interests. The order the court is asked to make is often referred to as a 'best interests declaration' and takes a form that depends upon whether the claimant is asking to have treatment sanctioned, withdrawn or withheld. So in a case where the NHS trust is seeking the sanction of the court, it will apply for a declaration from the court, for example, that not commencing ventilation would be in the baby's best interests, despite the parents wish for the baby to be ventilated. Conversely, when the doctors feel the baby needs a particular treatment or operation and the parents disagree, the trust will seek a declaration that the particular treatment or operation is lawful, despite the lack of parental consent, and in the baby's best interests.

Once an application has been issued, a judge will give pre-trial directions, dealing for example with disclosure of the baby's medical records, the evidence† that is needed, when it should be produced, which witnesses should attend trial for cross examination, the date of the trial and the issue of publicity. Cases concerning children are usually heard in private, but recently (for example in the cases concerning Charlotte Wyatt and Luke Winston-Jones) the court has agreed, at the request of the parents, for the hearings to take place in 'open' court, that is in public. If the case is urgent, hearings are possible at any hour of the day or night, provided clear evidence is given to the court explaining why the case is urgent, and the degree of urgency involved.

Independent expert medical evidence based upon a baby's prognosis with and without the disputed treatment is often important for the court, the baby and the parents. A breakdown of communication between parents and the doctors treating the baby is a feature in some of the cases that reach the court. Independent paediatricians or neonatologists, retained as expert witnesses, in ideal circumstances may sometimes, in the course of detailed discussions with the parents, give new or clearer explanations that lead to shared knowledge, consensus or understanding and acceptance of different views. Alternatively parents may feel the experts are there simply to support their colleagues and assert medical authority over the lay public.

* By issuing an application summarising the orders sought from the court.

† For example, factual evidence from the doctors treating the baby, and the parents and expert evidence from independent doctors.

In the next example, doctors advised discontinuation of life support but the parents disagreed.

Case 7: Elliot – intensive care continued at the parents' wishes

Elliot

A first-time mother had labour induced at 41 weeks of pregnancy and needed an emergency Caesarean section. She gave birth to a baby she named Elliot who weighed 2,700g, which is relatively small for 41 weeks. Elliot did not breathe. He was resuscitated but developed seizures after four hours. After treatment with anticonvulsant drugs his seizures stopped 12 hours later, but he remained unresponsive and ventilator-dependent after a further 96 hours. An electro-encephalogram (EEG) revealed low brain activity and an abnormal blood flow in the brain. Cerebral MRI scans indicated serious brain injuries.

All the clinical evidence, with a high degree of certainty, indicated that if Elliot survived, he would have severe quadriplegia (paralysis of all four limbs) and learning difficulties as well as severe epilepsy. The neonatologists and nurses were in agreement that further life support was not justified when the prognosis was so poor. The clinicians based their prognosis on recent published evidence and their experience. All the relevant assessments were completed by 96 hours and the evidence indicated he would be unable to sit up or eat, and would be completely dependent on carers for all functions. Life would be a series of procedures with tubes, and uncomfortable handling. The parents did not agree to the withdrawal of life support, saying that in their view, life was sacred and only God should take it away. Elliot remained on mechanical ventilation. The next critically ill infant born at that hospital had to be transferred 50 miles to another city because there were not enough nurses to staff further intensive care cots. Elliot remained in the hospital for a further three months. Eventually he developed a serious infection and died in hospital without ever breathing on his own.

Sanctity of life

6.28 The clinical management of Elliot's condition is representative of normal practice in the UK when there is disagreement over whether full treatment should be withdrawn; that is, life support was continued. Elliot's parents valued life as sacred at any cost, which was in keeping with their religious beliefs. The values they hold correspond with the absolutist stance on sanctity of life that we identified in paragraph 2.9. They may also have been reluctant to accept the prognosis given by the doctor and hoped that Elliot would improve in time. In their view, even the most uncomfortable life with very limited communication was more valuable than death. Doctors are trained to try to do their best for their patient and to ignore consequences for other patients who are not directly under their care. From the doctor's perspective, maintaining Elliot's life was not in his best interests. The healthcare team could ensure that he did not suffer by providing palliative care.

Best interests

6.29 The conflict between the parents and the medical team about how to act in Elliot's best interests could not be resolved. After Elliot died, his parents may have some consolation from knowing that they acted in accordance with their beliefs and that everything possible had been done for Elliot. Even so, the situation would have been highly traumatic for them. For the clinical team, since their professional opinion was that the burden to Elliot exceeded the benefits of continuing treatment, their agreement to maintain a treatment that they thought futile may have come at a personal emotional cost.⁴¹

Pressure on resources

6.30 In addition to raising the issue of whether continuing ventilation was in Elliot's best interests, his case raises issues of justice and fairness. Elliot's treatment in intensive care was in retrospect unsuccessful and likely to be associated with serious disability later on. This was an outcome that the doctors felt was not in his best interests. The treatment was given to Elliot rather than another baby whose chances of survival without significant disability were greater. To the doctors, it might not seem fair to allocate resources to Elliot when another child could reasonably be expected to benefit more from them. The baby who had to be transported on a ventilator to another city, risked deterioration of his or her condition because of the transfer. So long as resources such as the provision of cots and staff within intensive care units are limited, issues about their fair use can be expected to arise.⁴²

⁴¹ Hefferman P and Heilig S (1999) Giving "moral distress" a voice: ethical concerns among neonatal intensive care unit personnel *Camb Q Healthc Ethics* 8: 173–8; Stutts A and Schloemann J (2002) Life-sustaining support: ethical, cultural, and spiritual conflicts Part II: Staff support: a neonatal case study *Neonatal Netw* 21: 27–34.

⁴² In the sense that the resources available for healthcare are finite, it will always be the case that care for one patient may affect that of another. Comments to the Working Party in fact-finding meetings indicated that although doctors do not allow resource implications to affect decision making, they are aware of the implications of their decisions.

6.31 This example provides another illustration of the pressure on healthcare resources that we described in paragraphs 5.45–5.51. If mechanical ventilation for Elliot had been discontinued, the equipment might have become available for a second baby, but the motive behind the doctors' advice might be questioned. From the perspective of the parents, the advice to consider allowing Elliot to die might be misunderstood for confusing ethical with economic issues; rationing resources rather than protecting their baby's best interests. We take the view that it is important to avoid arguments about 'bed-blocking' and instead to focus on the best interests of a baby. This is consistent with our recommendation that resource considerations should not affect decision making between the doctor and the parents of the patient (paragraph 2.43).⁴³ This is not to deny that decisions in practice are affected by cost, since the treatments that may be offered will necessarily depend on the facilities and staffing skills that are available in a particular neonatal unit. The capability of a particular unit depends in turn on decision making in healthcare provision at the national or regional or local (hospital) levels (macroeconomic or mesoeconomic levels, see Figure 3.4). The Working Party would expect considerations of 'fairness' and justice to be a part of decision making on the distribution of resources (paragraph 2.41).

Legal issues

6.32 This is another case which could have been brought before a court. However, the doctors and the NHS trust chose not to challenge the parents. In our meetings with healthcare professionals we found that most are unwilling to add to parents' trauma by recommending that the trust should initiate court proceedings unless they judge that the baby is suffering severely. Had the trust challenged the decision of Elliot's parents by going to court, the sole concern would be the best interests of Elliot himself, not the impact his survival would have on NHS resources or on other critically ill babies. Questions for a court would focus on whether Elliot experienced any meaningful human interaction in his limited life. For example, did he respond to his parents? The court would also be concerned with the burdens to Elliot of treatment. Was he subject to pain and distress?⁴⁴ If Elliot gained some benefit from his life, a judge might be hesitant to overrule his parents' wishes (see Chapter 8).

Economic issues relating to provision of intensive care

6.33 In this section, we consider first, early economic evaluations of the cost-effectiveness (see also paragraphs 5.45–5.51). Secondly, we examine the cost-effectiveness of a routinely-used treatment strategy, as an example of how more recent research into the cost-effectiveness of neonatal medicine has tended to focus on specific interventions.⁴⁵ There are several outcome measures incorporated into studies of cost-effectiveness. The quality of life (QALY) measure, which we describe in Appendix 8, is often used.

6.34 Early studies have been of variable quality in terms of their evidence for the effects of neonatal intensive care on mortality. One study from Canada⁴⁶ and another from Australia⁴⁷ analysed outcomes for selected groups of the population before and after access to neonatal intensive care was expanded. For babies born weighing less than 1,000 g, the studies estimated

⁴³ See the discussion of resources for healthcare in Mason JK and Laurie GT (2005) *Mason and McCall Smith's Law and Medical Ethics*, 7th Edition (Oxford: Oxford University Press), pp 571–4.

⁴⁴ Claims made in court cases about the pain experienced by babies with serious health problems are often not substantiated by proper assessment or measurement. Nor is it always clear if adequate treatment for pain has been given.

⁴⁵ Petrou S and Mugford M (2000) Predicting the costs of neonatal care in *Current Topics in Neonatology*, Hansen TN and McIntosh N (Editors), Volume 4 (London: WB Saunders), pp 149–74.

⁴⁶ Boyle MH, Torrance GW, Sinclair JC and Horwood SP (1983) Economic evaluation of neonatal intensive care of very-low-birthweight infants *N Engl J Med* **308**: 1330–7.

⁴⁷ Kitchen WH, Bowman E, Callanan C, *et al.* (1993) The cost of improving the outcome for infants of birthweight 500–999g in Victoria *J Paediatr Child Health* **29**: 56–62.

a cost per additional survivor of between £85,000 and £174,000 (at 1998 prices). When the outcome was measured in terms of an additional year of life gained, the extra cost fell to between £4,440 and £15,790. Alternatively, measuring the outcome in terms of an additional QALY gained, gives figures of between £4,190 and £38,030.

6.35 Evaluating the effects of treatment in economic terms to determine their cost-effectiveness is a complex task. The widespread introduction into neonatal intensive care of surfactant-replacement treatment in the early 1990s for less developmentally mature babies saved lives, which led to increased periods of stay in intensive care and thus increased costs.⁴⁸ It was also associated with increased costs in caring for survivors once they left hospital. However, in the case of more mature babies for whom it is equally effective, and for whom mortality and morbidity are low, surfactant led to shorter periods of stay and a clear cost benefit.⁴⁹ In the future, it is likely that economic analysts will have to evaluate the impact of recent developments in neonatal care such as head cooling, whole body cooling, liquid ventilation, intravenous immunoglobulin therapy and high-frequency oscillation ventilation.

Further example

Case 8: Freddie – pain relief and the consequence that death is hastened

Freddie

Freddie was born by normal vaginal delivery after a nine month pregnancy. It rapidly became apparent that he had the incurable rare inherited skin condition, recessive junctional (Herlitz type) epidermolysis bullosa (EB), which is lethal in infancy. The diagnosis was confirmed by skin biopsy and DNA analysis.

Here is a description by a mother of a child with this most serious form of the condition⁵⁰:

"A child with painful wounds similar to burns covering most of his or her body. Having to wrap each tiny little infant finger with Vaseline gauze and then cover it with gauze to prevent the hand from webbing and contracting.

Never being able to hold your child tight because if you did, their skin would blister or shear off.

A child who will never know what it's like to run, skip or jump, or to play games with other children because even the slightest physical contact will injure his or her skin.

A child who screams out each time it is bathed because the water touching its open wounds creates incredible pain.

A diet of only liquids or soft foods because blistering and scarring occur in the oesophagus.

An active baby with his knees soaked in blood from the normal act of crawling."

Freddie's parents read this description, and others, on the Internet.⁵¹ They realised that the severity of the condition varied and that there were different forms. Other sufferers might not be as badly affected. However, it was clear that their son had the most serious form, which occurs in only a small proportion of babies with epidermolysis bullosa. They accepted advice from doctors, who consulted specialists in the disorder, that there was no treatment for their son's condition. It was obvious that Freddie was suffering and he was given morphine⁵² for pain relief. The dose was increased to control the pain, until the point came that his breathing started to be affected. The parents were still very concerned that their child might be in severe pain and asked the medical staff to continue to increase the dose of morphine, which was done. They did not want him to be mechanically ventilated if he stopped breathing, and the clinicians agreed to this request. The parents were present when Freddie died peacefully.

⁴⁸ Respiratory distress syndrome, which almost always occurs in babies born at less than 37 weeks of gestation, is caused by a lack of surfactant. Surfactant is needed to help keep the air sacs of the lungs open but babies who are born early do not produce enough; they rapidly develop breathing difficulties and need supplemental oxygen. Surfactant replacement therapy is given into the lungs via a breathing tube; it supplements the baby's own surfactant until he or she can make it him or herself, usually after about 3–4 days.

⁴⁹ Schumacher R, Burchfield D, Vaughan R *et al.* (1991) Economic impact of two rescue doses of Exosurf neonatal in > 1350 gram infants *Pediatr Res* **29**: 264A, 1570; Mugford M, Piercy J and Chalmers I (1991) Cost implications of different approaches to the prevention of respiratory distress syndrome *Arch Dis Child* **66**: 757–64; Schwartz R, Luby A, Scanlon J *et al.* (1994) Effect of surfactant on mortality and resource use in infants weighing 500 to 1500g *N Engl J Med* **330**: 1476–80.

⁵⁰ See www.ebinfo.world.com.

⁵¹ For example DeBRA online, available at: <http://www.debra.org.uk/>. Organisations with websites such as this can provide information on particular conditions, enable people and families who are affected to establish contact, raise funds for research and raise awareness of the condition.

⁵² Morphine is widely used for pain relief in neonates. Other painkillers used for severe pain, including diamorphine, pethidine and fentanyl, would have similar effects on breathing.

Best interests

6.36 Arguments presented for Freddie's best interests were based on the concern about existing and future pain he would suffer and the poor prognosis for this incurable condition. For him, was survival worse than death? All those involved in the decision-making process agreed that Freddie's condition was 'intolerable' (paragraph 2.16). This condition is very distressing to witness, not just for the parents but also for clinical staff. It is crucially important that the diagnosis is correct and not confused with another type of the skin disorder which has a better prognosis, such as epidermolysis bullosa simplex.

The doctrine of double effect

6.37 Freddie's case illustrates the doctrine of double effect (paragraph 2.38) which becomes relevant where an action is taken with two outcomes, one good and the other bad. For such an action to be acceptable, the bad caused should not outweigh the good intended. The point at which the doctrine of double effect came to apply was when the dose of medication for pain had been increased to the point that Freddie's breathing became affected. The parents and the doctors wanted pain relief to be continued so that he should not suffer, but they also knew that increasing the dose would suppress his respiration. Here, giving morphine was intended to relieve pain but in the knowledge of the possibility of hastening death. While some people view such actions as equivalent to deliberately ending a life, others would disagree. **The Working Party takes the view that, provided the treatment in this case has been guided by the best interests of the baby, and has been agreed through joint decision making, pain-relieving treatments are morally acceptable, even if potentially life-shortening (paragraph 2.38).**

Active ending of life

6.38 Some might argue that Freddie should be given a lethal injection under sedation to allow him to die quickly. However, unlike voluntary euthanasia in adults, which is sometimes defended on the grounds that competent adults have a basic right to exercise choice, a baby cannot let his or her wishes be known. Thus any decision to end life would be on the basis of what others judged to be his or her best interests. This case also raises the question (accepting that it is not permissible by law) of whether it is ever morally acceptable actively to end life. Our position is that it is not (see paragraphs 2.37 and 8.40), and that to allow this practice would erode trust in doctors and the neutrality of their advice. There is further discussion of this issue in Chapter 8.

Decision making

6.39 All those involved in the decision-making process agreed that to continue life-sustaining treatment was not in Freddie's best interests, and arrangements were made to support his parents and for them to be with him while he was dying. Clear communication between the parents and the healthcare team about Freddie's condition and the options for clinical management was crucial to avoid later confusion or psychological distress if the parents subsequently experienced guilt in having 'allowed' Freddie to die.

6.40 This case exemplifies the complexity of decision making. First, decisions about the care of the newborn take place by means of an accumulating series of conversations, observations and interactions, sometimes quite minor, that contribute to a final decision.⁵³ Secondly, decision making does not entail a single decision that is made by the parents and doctors. In fact, a whole series of decisions need to be made, about what kind of care is given to a child at what stage and for how long.

⁵³ Decision making is understood by sociologists to be a staged process achieved through social interaction, and not a single act undertaken at one point in time by a single party.

6.41 During the decision-making process, parents sometimes complain of being deluged with conflicting information.⁵⁴ The nature of the information includes medical data about a child's diagnosis and prognosis, which is often complex and will include estimates of probabilities of particular outcomes.⁵⁵ Doctors sometimes use terminology that will not readily be understood unless it is carefully explained. Consequently parents will often fear that they have not understood it well enough to provide a basis for properly considered, informed consent (see paragraph 2.51 and Appendix 5). Further, doctors may be selective in what they tell parents, even for the best of reasons (see paragraph 6.42). How are parents to be sure they are being told everything they need to know?⁵⁶ Nevertheless, parents have also reported comprehending relevant medical information very quickly and acquiring an ability to participate in discussion of their child's condition.⁵⁷ Specifically in this case, Freddie's parents needed to gain a clear understanding of how epidermolysis bullosa can vary in severity, and that outcomes can differ between different patients. In conditions as rare as this form of epidermolysis bullosa, a specialist in palliative care would ideally be asked to advise the healthcare professionals looking after Freddie.

6.42 Some neonatal healthcare professionals believe that decisions which mean a choice between a baby surviving with a high probability of severe disability or a baby dying are too burdensome for some parents to take. Staff with these views might be inclined to present their advice in a way that leaves parents with little choice but to accept it. A study of 57 doctors reported that 58% preferred a joint approach to decision making and that only two thought that parents should take the full responsibility.⁵⁸ However, approximately 30% of the doctors considered that they alone should make decisions about critical care. The parents with whom we met⁵⁹ were clear that, whatever the magnitude of the decision, it was one that they should take as parents, although in practice decisions were jointly taken, or made by the doctors.⁶⁰ In a study involving parents for whom there had been discussion about limiting treatment for babies born at a range of gestations, 42% of parents considered that they had actually made the decision, 10% thought the decision was a joint one and 31% took the view that the doctors had decided.⁶¹ All parties need to be clear about how the decisions are being made, and each needs to be given the appropriate opportunity to participate. Nevertheless, it is entirely possible and even perhaps inevitable⁶² that when all concerned in a particular case believe that the parents were genuine partners in decision making, the parents may feel that they were led subtly towards a particular view. The balance of the information given, the tone of voice used, or the status of the person giving it can all be influential. One parent we met spoke of the "subtleties of the questions asked" (by both professionals and parents), and the complexities of the spoken words in the clinic, during a time of heightened emotion and

⁵⁴ Personal communication at fact-finding meeting (see Appendix 1).

⁵⁵ For discussion of views and experiences of parents and staff about sharing information and the care of babies with uncertain futures see Alderson P, Ehrich K, Hawthorne J, Killen M and Warren I (2004) *Foretelling futures: dilemmas in neonatal neurology* (London: Social Science Research Unit, Institute of Education, University of London).

⁵⁶ Later events may cause parents to question the adequacy of information provided at the time; see Case 6, Danielle.

⁵⁷ Personal communication from parents at fact-finding meetings (see Appendix 1).

⁵⁸ McHaffie HE and Fowle PW (1996) *Life, Death and Decisions: Doctors and Nurses Reflect on Neonatal Practice* (Hale: Hochland & Hochland Ltd).

⁵⁹ The Working Party met parents at fact-finding meetings. However, these were parents who had expressed a wish to meet the Working Party and we acknowledge that other parents might feel differently.

⁶⁰ McHaffie HE, Laing IA, Parker M and McMillan J (2001) Deciding for imperilled newborns: medical authority or parental autonomy? *J Med Ethics* 27: 104–9.

⁶¹ McHaffie H (2001) *Crucial Decisions at the Beginning of Life* (Abingdon: Radcliffe Medical Press).

⁶² There is also an imbalance of expertise and knowledge between the parents and healthcare professionals. This is a dilemma faced by all professional givers and lay receivers of advice. Those giving the information and advice are likely to be selective about what they impart. Their selection is likely to be influenced, consciously or subconsciously, by their own views about the 'right' outcome.

changing reactions.⁶³ In one study, a parent involved in decisions about her baby with brain damage described how “It makes you *feel* like you’ve made the final choice but you haven’t made the final choice”.⁶⁴

Supporting parents when a baby dies

6.43 When a decision has been made to change current treatment to palliative care, parents will need support. Ideally, nurses and doctors in a neonatal unit will provide immediate practical and emotional help at the time of death for bereaved families, as well as facilitating links to the community for longer-term support. For many severe disorders such as epidermolysis bullosa, specialist organisations can provide help. Most neonatal units offer parents assistance with funeral arrangements, the gathering and presentation of keepsakes, answering questions and providing information about the cause of death. They also offer leaflets and other general information about the grieving process. Nurses or counsellors will usually keep in contact by telephone or on a return visit to the hospital in the first few months after a baby’s death, and parents are encouraged to contact local community support groups (see paragraph 3.18).⁶⁵

Legal issues

6.44 Whatever the ethical arguments, we restate that it would be illegal to take active steps designed to hasten Freddie’s death. Any act intended to end life constitutes the crime of murder. However, the law accepts a version of the doctrine of double effect that permits doctors to administer necessary pain relief even in doses that are known to have the incidental effect of shortening life. Giving Freddie morphine to ensure he did not suffer is lawful in the UK (see paragraph 8.18). Having concluded our discussion of the types of decision that may need to be made for babies in intensive care treatment, we turn now to the broader question, of how more comprehensive and robust data can be gained on health outcomes. This information is crucial to enable doctors to provide more robust advice to parents on the prospects for individual cases.

Determining outcomes

6.45 A consistent theme throughout this Report has been the paucity of information on the prevalence, severity and causes of disability which have their origin before or at the time of birth, in the child population. A similar paucity of data has been noted in a recent major study in the USA.⁶⁶ In the UK, national statistics are limited to routinely collected data on birth and death. We noted in Chapter 5, that as the prospects for survival and outcomes for babies born at progressively earlier gestational ages improve, there is the possibility that the absolute number of survivors with some level of disability will increase. Significantly, the lack of information on health outcomes may become an increasing problem as advances in healthcare, medicines and technologies begin to improve the prospects of children with previously untreatable conditions. This includes, for example, children with cystic fibrosis, congenital birth defects, severe respiratory or heart conditions and cancers.

⁶³ Personal communication at a fact-finding meeting.

⁶⁴ McHaffie H (2001) *Crucial Decisions at the Beginning of Life* (Abingdon: Radcliffe Medical Press).

⁶⁵ The parents may sometimes want to contact organisations such as the Child Death Helpline (<http://www.childdeathhelpline.org.uk>, accessed on: 30 March 2006) and the Stillbirth and Neonatal Death Society (SANDS) (<http://www.uk-sands.org>, accessed on: 30 March 2006) which will allow them, for example, to share their experiences with others or obtain advice on, for example, going back to work or coping with the anniversary of a baby’s death. These organisations have links to other groups that offer similar help.

⁶⁶ The Institute of Medicine report noted a paucity of information on preventing, diagnosing, treating and minimising the long-term effects specifically of preterm labour. The recommendations of the report emphasise the need for research to address these issues and for the encouragement of large-scale prospective studies. The report draws attention to one such investigation that has been proposed, the US National Children’s Study, which would follow the health and development of 100,000 children up to the age of 21.

- 6.46 Neonatal critical care decisions are particularly difficult because of the lack of information about long-term follow up on which to base predictions of future health outcomes. Follow up is needed not only for groups of children diagnosed with serious clinical problems around the time of birth or who are born at the borderline of viability, but also for children who have minor manifestations during this period but who are at potential risk of late-onset problems.⁶⁷ EPICure and other birth cohort studies have provided some information at the national level and will remain an important means of addressing more subtle aspects of outcome, including cognitive function, behavioural difficulties and assessments of the quality of life for the children affected, and their families, at different stages in their development. **We conclude that further clinical research of this type is needed to identify outcomes relating to the quality of life for the children affected and their families at different ages. We note that these studies will require consent on a case-by-case basis.**
- 6.47 However, to monitor the outcome of neonatal care at the population level, basic data relating to birth and neonatal care needs to be documented, analysed on a routine basis and linked to subsequent information collected through the NHS such as hospital admission, child health records, attendance in general practice and educational placement. Without improved data on outcomes, it will not be possible to give parents and healthcare professionals a more robust prognosis to help with decision making. Such data are also needed to improve current understanding of the relationship between clinical care and outcome. The availability of linked information of this kind would also encourage clinical trials and associated follow-up studies. **The Working Party therefore takes the view that it is crucially important that the various existing datasets comprising clinical information collected at birth and subsequently during the neonatal period, should be integrated and linked.**⁶⁸ **We further recommend to the UK Departments of Health that there should be linkage with additional data collected to record later health outcomes, not only from childhood, but including adolescence through to adulthood. These data should be captured through NHS health records and educational records and will provide crucial information on health outcomes.** We note that training of healthcare professionals will be required to help ensure a consistent knowledge base for the identification and collection of the relevant data.
- 6.48 Recently, several initiatives to begin data collection have been established. We are encouraged by the establishment of a National Neonatal Audit Programme (NNAP) for England and similar plans for Wales, as well as the Maternity Services Dataset and other datasets collecting critical clinical data at birth and during the neonatal period. The NNAP will be administered by the RCPCH on behalf of the Healthcare Commission. A clinical dataset has been identified, based on a concise range of audit information around birth and aspects of neonatal care.⁶⁹ If further funding is secured for the programme, it is hoped that follow up of infants at two years of age will be conducted to compare outcomes in relation to specific interventions and the infant characteristics of children, between those who have experienced a period of care in a neonatal unit and other children.⁷⁰

⁶⁷ For example, babies who have acquired brain injury which could result in developmental problems.

⁶⁸ The new Maternity Services Dataset, the National Neonatal Audit Programme and the Neonatal Critical Care Minimum Dataset, and data collection being undertaken by neonatal networks/groups of networks, such as SEND (South East Neonatal Data) will all collect relevant information about what happens at birth and during the neonatal period.

⁶⁹ The initial aim of the NNAP programme is to identify areas for improvement in care in relation to delivery and outcomes and to provide a mechanism for ensuring consistency of care. For more information see: <http://www.ncap.org.uk/>, accessed on: 10 Aug 2006.

⁷⁰ Personal communication from the National Neonatal Audit Programme.

- 6.49 The current development of an electronic NHS database will make routine national data collection feasible, and the recent implementation of the NHS Numbers for Babies scheme⁷¹ will ensure that the electronic record includes health information from birth onwards.⁷² The real challenges will be to identify the essential data that should be recorded from the neonatal period and in the longer term. Examples of the early data that we regard as important to collect include gestational age at birth as well as birthweight, clinical status and details of neonatal care. Other valuable information which could be recorded includes the use of pre-conception fertility treatment, as this is associated with multiple births, and consequently with prematurity (see paragraph 3.4).⁷³ Data collected at a later stage will need to include indicators of health and educational progress. Moving to a system of routine, continuous data collection based on electronic NHS systems would allow a much more complete picture of practice in neonatal medicine to be obtained. We recognise that, by their nature, data on outcomes may only partly reflect current practice by the time they are analysed. Nevertheless, there is, in the view of the Working Party, an ethical imperative to analyse outcomes for neonatal care. **In summary, although the necessary electronic NHS systems are not yet in place, the view of the Working Party is that it is timely to identify the health-related questions that should be posed and the corresponding requirements for data collection.**
- 6.50 There is a further issue that needs to be taken into account when planning the routine collection of data for possible use in research studies. Interpretation of the Data Protection Act 1998 has limited the number of studies that are able to link NHS numbers to patient records in order to obtain particular health outcomes because the Act seeks to protect the identities of individuals. Normally, if information about patients is to be used for purposes beyond the delivery of personal treatment and care (as is the case when data are used for research), the consent of patients should be sought or the information should be anonymised. Although the intention has been to ensure that personal information is handled with appropriate care and respect, in practice the outcome has created a conservative culture of governance.⁷⁴ Proposals for such studies must be referred to the Department of Health Patient Information Advisory Group, a committee that decides on behalf of the Secretary of State whether monitoring may proceed without seeking consent⁷⁵ under Section 60 of the Health and Social Care Act 2001. Certain exceptional conditions must be satisfied to justify access. Broadly, a strong public interest must exist to justify access to patient-identifiable information where consent and anonymisation are impossible or inappropriate. **The view of the Working Party is that basic studies based on data linkage are required, to complement research studies aimed at determining more subtle aspects of outcome (paragraph 6.46). Such studies do not require contact with patients or their families. In view of the strong public interest in determining outcomes from critical care decisions, we recommend that that proposals for**

⁷¹ Under this scheme, which began in 2002, babies are now assigned a unique identifier, their NHS number, at birth, rather than after civil registration up to six weeks later. For further information, see <http://www.connectingforhealth.nhs.uk/nhsnumber/nn4b/>, accessed on: 3 Oct 2006.

⁷² In 2006 expectations were that the 'National Programme for IT' (electronic health records) would be phased in over the next ten years, see <http://www.connectingforhealth.nhs.uk/>, accessed on: 26 Sept 2006.

⁷³ A need to record outcomes following the use of fertility treatment has been recommended elsewhere to enable assessment of the impact on the health of children conceived using these techniques; Medical Research Council (2004) *Assisted Reproduction: A safe sound future* (London: MRC); Human Genetics Commission (2006) *Making Babies: Reproductive decisions and genetic technologies* (London: HGC). This would require the amendment of the Human Fertilisation and Embryology Act 1990, which is currently under review.

⁷⁴ The Academy of Medical Sciences (2006) *Personal Data for Public Good: Using health information in medical research* (London: Academy of Medical Sciences), available at: <http://www.acmedsci.ac.uk/images/publication/Personal.pdf>, accessed on: 6 June 2006.

⁷⁵ See <http://www.advisorybodies.doh.gov.uk/piag/>, accessed on: 26 May 2006.

studies based solely on data linkage should be referred to the Department of Health Patient Information Advisory Group to request access to the relevant patient information. It is crucial that decisions should be based on accurate and up-to-date evidence from research about the risks to and likely outcomes for babies in whom a birth abnormality or genetic disorder has been recognised antenatally or in the newborn period, as well as for extremely premature babies.

6.51 Finally, it is important not to overlook information of a different nature that may be of future help to parents and doctors. If a baby dies, information from an autopsy can be useful in confirming an original diagnosis, establishing a cause of death and providing further information about the presence of disease or abnormality. The Human Tissue Act 2004 regulates autopsies in England, Wales and Northern Ireland, other than those ordered by a coroner. An autopsy on a newborn baby can be conducted only with explicit consent from his or her parents, unless it has been ordered by a coroner. The Human Tissue (Scotland) Act applies in Scotland and similarly requires specific parental authorisation for autopsies. Autopsy data may also provide some insights into the cause of death and help parents in planning future pregnancies. Furthermore, if doctors can gain a better understanding of the causes underlying clinical conditions, other parents can be given more accurate information when making decisions, and research efforts can be directed towards obtaining more precise diagnoses. Recent studies suggest that autopsies provide valuable additional information in 25–30% of cases of neonatal death or termination for fetal abnormality.⁷⁶ We are aware that doctors can sometimes be reluctant to ask parents for consent to carry out an autopsy for a fetus or a child who has died, and that parents may also refuse their consent.^{77,78} Good communication is essential to help parents make a genuine choice about autopsy, and the subject must be introduced sensitively and appropriately. However, even though useful information can be gained from autopsy, rates have been declining since the mid-1990s.⁷⁹ Although this decline has been attributed to a general decline in trust in the patient–professional relationship in the wake of recent enquiries, it is also the case that many regions now lack specialist paediatric pathologists to perform autopsies. It is important to overcome this practical difficulty. The benefits of providing information for the future should not be underestimated and we encourage doctors and parents to continue to consider autopsy as an option.

⁷⁶ Boyd PA, Tondi F, Hicks NR and Chamberlain PF (2004) Autopsy after termination of pregnancy for fetal anomaly: retrospective cohort study *Br Med J* **328**: 137; Brodrie M, Laing IA, Keeling JW and McKenzie KJ (2002) Ten years of neonatal autopsies in tertiary referral centre: retrospective study *Br Med J* **324**: 761–3.

⁷⁷ There have been studies on parents' reasons for giving and refusing consent for autopsies, see for example McHaffie HE (2001) *Crucial Decisions at the Beginning of Life* (Abingdon: Radcliffe Medical Press). Common reasons given by parents include wanting to avoid further invasive procedures or because the parents themselves did not have any unanswered questions. In other cases, the information gained has been reported by parents as useful in helping to understand why their baby died, see Rankin J, Wright C and Lind T (2002) Cross sectional survey of parents' experience and views of the postmortem examination *Br Med J* **324**: 816–18.

⁷⁸ In 2003, autopsies were performed in 39% of cases of fetal or neonatal death between 20 weeks of gestation, and 28 days after birth. In 61% of cases an autopsy was not performed, because consent was not given by the parents; in 38% of cases an autopsy was not offered. Confidential Enquiry into Maternal and Child Health (2005) Stillbirth, Neonatal and Post-neonatal Mortality 2000–2003 – England, Wales and Northern Ireland (London: RCOG Press), available at: <http://www.cemach.org.uk/publications/CEMACHPerinatalMortalityReportApril2005.pdf>, accessed on: 31 May 2006.

⁷⁹ Confidential Enquiry into Maternal and Child Health (2005) Stillbirth, Neonatal and Post-neonatal Mortality 2000–2003 – England, Wales and Northern Ireland (London: RCOG Press), available at: <http://www.cemach.org.uk/publications/CEMACHPerinatalMortalityReportApril2005.pdf>, accessed on: 31 May 2006; Confidential Enquiry into Stillbirths and Deaths in Infancy (2001) 8th Annual Report (London: Maternal and Child Health Research Consortium), available at: <http://www.cemach.org.uk/publications/8threport/z-Complete%20file.pdf>, accessed on: 31 May 2006.