Abstract:

With the ever-increasing availability of aggressive medical treatment and technical support, neonatologists are offered an increasing ability to prolong life. While end-of-life decisions made in NICUs have been studied internationally, there is limited data available for Ireland. Through the auspices of the Irish Faculty of Paediatrics 2002 Neonatal Mortality Questionnaire, decisions made around the time of death in Irish Neonatal Intensive Care Units were examined. The overall response rate to the questionnaire was 96% (n=25). One hundred and eighty seven deaths were reported for 2002. Information pertaining to the mode of death was available in 53% of cases. Seventy seven percent of those paediatricians who answered this question, reported either withdrawing or withholding treatment in babies thought to have a hopeless outcome, with the greatest proportion of these deaths occurring in premature infants (n=30) and babies with congenital defects (n=40).

Background

Sometimes it is necessary to come to the conclusion that for an individual child – who might be a premature baby, a toddler, a child at primary or secondary school, a teenager or a young adult – the more humane path is one of palliation rather than a continuation of life-saving treatment. With the ever-increasing availability of aggressive medical treatment and endless amounts of technical support, doctors involved in intensive care, including neonatal intensive care, are offered an increasing ability to prolong life.

However, recent studies from North America, the UK and Europe suggest that most of the deaths occurring in the Neonatal Intensive Care Unit (NICU), are preceded by an active process of either withdrawal or limitation of critical care. While end-of-life decisions within NICUs have been studied internationally, there is limited data available for Ireland.

Aims

The aims of this study were to examine decisions made around the time of death in Irish Neonatal Intensive Care Units and to examine the relationship between the type of decision and the underlying medical condition.

Methods

Through the auspices of the Irish Faculty of Paediatrics, Neonatal Mortality questionnaire by post to all Paediatric and Neonatal units throughout Southern Ireland. They were sent by post to the Consultant-in-charge of each Neonatal Paediatric Unit and to the Consultant Pathologists of Our Lady’s Hospital for Sick Children, Crumlin and The Children’s University Hospital, Temple Street, Dublin. Telephone reminders were given to those who had not completed the questionnaire by the end of August 2003. Data was sought pertaining to the deaths of all live born infants born in 2002 with a birth weight greater than 500g but who died within the first 28 days of life. Neonatal deaths were categorised according to the Wigglesworth classification, into Congenital Malformations, Prematurity, Asphyxia and other Specific conditions. A specific section of the questionnaire examined the mode of death of the infants. One closed-ended question clarified whether the infant had died:

1. during or soon after full cardiopulmonary resuscitation (CPR);
2. following withdrawal of ventilatory support;
3. following a decision not to give CPR at birth;
4. following a decision not to give CPR after initial resuscitation; or
5. as a result of Sudden Infant Death Syndrome (SIDS). More complete details of this survey have been described elsewhere. Each hospital and patient was assigned a unique identification number and all data was entered anonymously into a Microsoft Access database for analysis.

Results

Twenty-five out of 26 centres, including the 3 Dublin maternity hospitals, responded to the Neonatal Mortality Survey giving a response rate of 96%. A total of one hundred and eighty seven neonatal deaths were reported in southern Ireland in 2002. The Neonatal Mortality Rate for 2002 was 3.1/1000 live births with a Corrected Neonatal Mortality rate of 1.8/1000 live births. Congenital Malformations represented the greatest proportion of deaths (44.5%; n=83) followed by prematurity (36%; n=67) and asphyxia (7.5%; n=14). Other causes of death (n=23) included meningitis (n=3), sepsis (n=3), Sudden infant Death Syndrome (SIDS) (n=3), necrotising enterocolitis (n=2) and hydrops fetalis (n=3).

Information pertaining to the mode of death was available in 53% of neonatal deaths (n=100). The remaining eight seven questionnaires did not contain any information on this issue. Twenty percent (n=20) of neonatal deaths occurred during or after full cardiopulmonary resuscitation (Table 1). Ten of these deaths were as a result of prematurity, 8 due to congenital defects, 2 as a result of SIDS or meningitis and one as a result of severe NEC. Of the 67 neonatal deaths classified as due to prematurity, 30 were born after 32 weeks’ gestation. Of the 67 neonatal deaths classified as due to prematurity, 30 were born after 32 weeks’ gestation. Of the 67 neonatal deaths classified as due to prematurity, 30 were born after 32 weeks’ gestation. Of the 67 neonatal deaths classified as due to prematurity, 30 were born after 32 weeks’ gestation.

Table 1: Data pertaining to modes of death (n=100)

<table>
<thead>
<tr>
<th>Mode of Death</th>
<th>Number (n)</th>
<th>Birthweight (g) Mean (Range)</th>
<th>Gestation (Wks) Mean (Range)</th>
<th>Age at death (d) Mean (Range)</th>
</tr>
</thead>
<tbody>
<tr>
<td>During/after CPR*</td>
<td>20</td>
<td>865 (430-1150)</td>
<td>25 (23-29)</td>
<td>3 (0.2-2.7)</td>
</tr>
<tr>
<td>Prematurity</td>
<td>10</td>
<td>2834 (1350-3450)</td>
<td>36.6 (33-40)</td>
<td>0.27 (0.25-6.0)</td>
</tr>
<tr>
<td>Withdrawal of Support</td>
<td>41</td>
<td>4005 (3660 - 4050)</td>
<td>40</td>
<td>1.4</td>
</tr>
<tr>
<td>Congenital Malformations</td>
<td>22</td>
<td>2053 (2650-3710)</td>
<td>40</td>
<td></td>
</tr>
<tr>
<td><strong>Asphyxia</strong></td>
<td>5</td>
<td>2540</td>
<td>35</td>
<td></td>
</tr>
<tr>
<td>Other (Meningitis)</td>
<td>1</td>
<td>2720</td>
<td>37</td>
<td></td>
</tr>
<tr>
<td>Other (NEC***)</td>
<td>1</td>
<td>2621 (550-780)</td>
<td>33 (22-25)</td>
<td>3.3 (10.4-10.22)</td>
</tr>
<tr>
<td>Prematurity</td>
<td>9</td>
<td>2186 (520-3760)</td>
<td>35 (23-43)</td>
<td>33.5 (6.03-168)</td>
</tr>
<tr>
<td>Congenital Malformations</td>
<td>20</td>
<td>2370 (2020-2850)</td>
<td>38.3 (38-39)</td>
<td>185.6 (7.5-538)</td>
</tr>
</tbody>
</table>

* CPR: Cardiopulmonary resuscitation. ** Asphyxia: Defined as the occurrence of a cardiac arrest before full CPR. *** NEC: Necrotising enterocolitis.
Seventy seven percent of NICU deaths occurred as a result of withholding or withdrawal of treatment. CPR was withheld from outset in 31% of infants and after initial resuscitation in 5%. In 41% cases, an active decision was made to withdraw support. The two main causes for withholding or withdrawal of support were congenital malformations in 40 of the 77 infants (52%) and prematurity in 30 (39%) infants. The mean gestation for withholding or withdrawing care due to prematurity was 25.9 weeks range from 22-32 weeks. Withholding initial resuscitation tended to occur in the most immature infants between 22-25 weeks gestation. Care was withheld or withdrawn in 40 of the 48 (83%) of infants who died with congenital abnormality. Of the 100 infants in the Responder Group, seven died from asphyxia, six of these in full term infants (mean weight 2738, range 2650-3710g). Support was actively withdrawn in 4 of these infants, the remaining 2 babies dying during/after CPR.

Fifty one percent (n=41) of deaths occurred following an active decision to withdraw support (Table 1). The greatest proportion of these deaths occurred in preterm infants (n=20) with a further 15 occurring as a result of congenital malformations (Table 2). Four deaths were due to asphyxia and 2 were as a result of meningitis (n=1) and necrotising enterocolitis (n=1).

Table 2 Modes of death in infants with congenital malformations (n=48)

<table>
<thead>
<tr>
<th>Mode of death</th>
<th>Congenital abnormalities</th>
</tr>
</thead>
<tbody>
<tr>
<td>During or after CPR</td>
<td>Pulmonary Hypoplasia (4)</td>
</tr>
<tr>
<td></td>
<td>Congenital Heart Disease(2)</td>
</tr>
<tr>
<td></td>
<td>Hydrops Fetalis (1)</td>
</tr>
<tr>
<td>Withdrawal of care</td>
<td>Ambiguous Genitalia/Multiple congenital abnormalities (1)</td>
</tr>
<tr>
<td>Decision not to give CPR</td>
<td>Trisomy 13 (3)</td>
</tr>
<tr>
<td></td>
<td>Trisomy 18 (2)</td>
</tr>
<tr>
<td></td>
<td>Trisomy 21 (1)</td>
</tr>
<tr>
<td></td>
<td>Neural Tube Defects (4)</td>
</tr>
<tr>
<td></td>
<td>Renal Agenesis (4)</td>
</tr>
<tr>
<td></td>
<td>Skeletal Dysplasia (3)</td>
</tr>
<tr>
<td></td>
<td>Pulmonary Hyperplastic (1)</td>
</tr>
<tr>
<td></td>
<td>Diaphragmatic Hernia (1)</td>
</tr>
<tr>
<td></td>
<td>Goners Syndrome (1)</td>
</tr>
<tr>
<td></td>
<td>Stickler Syndrome (1)</td>
</tr>
<tr>
<td></td>
<td>Congenital Anomaly (1)</td>
</tr>
<tr>
<td>No CPR after initial resuscitation</td>
<td>Tiresome 18 (1)</td>
</tr>
<tr>
<td></td>
<td>Tiresome 13 (1)</td>
</tr>
<tr>
<td></td>
<td>Anencephaly (1)</td>
</tr>
</tbody>
</table>

Thirty one percent (n=31) of neonatal deaths occurred following a decision not to give cardiopulmonary resuscitation at birth. The majority of these (n=22) babies had a congenital malformation (Table 2). Extreme prematurity accounted for the remaining 9 deaths in this group, the average gestation being 23 weeks (range 22 to 25 weeks). Death occurred between 0.4 to 10.2 hours of age (Table 1).

Five neonatal deaths occurred following a decision not to give CPR after initial resuscitation. Three of these deaths were due to congenital malformations, one due to prematurity (28 weeks gestation, birth weight 720g) and asphyxia (30 weeks gestation and birth weight 640g). Three deaths occurred as a result of Sudden Infant Death Syndrome.

In the non-responder group (n=47), 40% of infants died as a result of congenital malformations and 31% secondary to prematurity. Asphyxial deaths accounted for 8% (Table 3).

Table 3 Causes of neonatal mortality (n= 187), stratified according to those who responded specifically to the Mode of Death question (Responders) and those who did not (Non-responders).

<table>
<thead>
<tr>
<th>Causes of neonatal mortality</th>
<th>Responders (n=100)</th>
<th>Non responders (n=87)</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Congenital abnormalities</td>
<td>48% (48)</td>
<td>40% (35)</td>
<td>83</td>
</tr>
<tr>
<td>Prematurity</td>
<td>40% (40)</td>
<td>31% (27)</td>
<td>67</td>
</tr>
<tr>
<td>Asphyxia (including meconium aspiration and persistent fetal circulation in the absence of congenital heart disease)</td>
<td>7% (7)</td>
<td>8% (7)</td>
<td>14</td>
</tr>
<tr>
<td>Others-non specific *</td>
<td>5% (5)*</td>
<td>21% (18)</td>
<td>23</td>
</tr>
</tbody>
</table>

Discussion

With modern advances in the technical aspects of neonatal care, it is now possible to prolong the life of very ill or preterm infants. However concern has been raised that in certain cases, continuation of intensive care may be inappropriate and merely prolongs the dying process. Duff and Campbell in 1973, were one of the first groups to examine this sensitive issue. They reported that just 14% of all deaths in the Yale NICU occurred following a decision to withdraw care. In 1988 Whitelaw explored the option of withdrawal of active care in an NICU setting in the UK and reported that 30% of deaths in the NICU in the Rammermith Hospital had occurred after a decision was made to cease active management of a baby. In the early 1990s, Ryan et al examined neonatal deaths in a tertiary level NICU in Edmonton, Alberta on a regular basis. Sixty nine percent of deaths occurred subsequent to withdrawal of therapy and 17% without CPR at the time of death. Around the same time, Wall et al studied deaths at a Level 3 NICU in San Francisco over a three-year period. Seventy three percent of neonatal deaths occurred as a result of withholding or withdrawing treatment. In 74% of cases the most commonly documented reason for limitation of therapy was the neonatologists belief that continued treatment was futile with death imminent. Eighty percent of babies (n=72) deaths in the NICU of a Connecticut University Hospital between 1991 and 1994, had do not
resuscitate status. Support was actively withdrawn in seventy two percent of these babies.

In 1995, the American Academy of Paediatrics issued a framework document entitled 'The Initiation or Withdrawal of Treatment for High-Risk Newborns'. This attempted to address concerns regarding inappropriate prolongation of life in an NICU setting. It advocated an approach where the threshold of viability, little or no information has been available on modes of death in the Irish NICU setting. In this study only 53 percent of paediatricians completed this section of the questionnaire. However from the available data it is clear that in 78% (n=78) of neonatal deaths paediatricians report either withdrawing or withholding treatment in babies thought to have a hopeless outcome, with the greatest proportion of these deaths occurring in infants with congenital abnormalities, followed by prematurity. These figures are consistent with internationally available data, especially that from North America.

It is interesting to note that despite the fact that there was an overall response rate of 96% for the Neonatal Mortality Survey questionnaire, only 53% of respondents completed the question on mode of death. This may be a reflection of a degree of discomfort in addressing this question. Some paediatricians may feel that, while withdrawal of intensive care may be in the best interest of a baby deemed to have a catastrophic prognosis, this might be construed as an act of murder. This theory is supported by the EURONIC study published in 2000. This showed a great variability in physician attitude to withdrawal of support throughout Europe. While 61 to 98% of physicians reported having been involved at least once in a decision to limit care, there was a large variation (28-90%) in the number who decided to become involved in a proactive decision of withdrawing mechanical ventilation.

Making end-of-life decisions within an NICU setting is always difficult because they approach 'diagnostic certainty'. It demands a significant amount of time of the paediatrician with the parents and requires the ability and emotional resources to deal with their anguish together with that of the other neonatal staff involved. At present it is particularly difficult in an Irish setting due to a lack of uniformity in approaching such decisions. Rather, such decisions in Ireland appear to depend on the philosophy of each individual neonatal unit and neonatologist, using a mixture of US and UK guidelines. For many Irish paediatricians, there are still significant ethical and medicolegal implications and concerns. For this reason, Irish paediatricians should work towards establishing their own guidelines in this regard taking into consideration our different psychosocial, cultural and religious background. Emphasis should be placed on the need of a multidisciplinary approach to these ethical dilemmas with good communication being of the utmost priority. The result of these guidelines should go beyond the topic of withdrawal of neonatal intensive care and should also provide advice in dealing with the resuscitation of extremely low birth weight infants at the threshold of viability.

References