Counselling and management for anticipated extremely preterm birth

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Fetus and Newborn Committee
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Abstract

Extremely preterm birth (birth between $22^{0/7}$ and $25^{6/7}$ weeks' gestational age [GA]) often requires parents to make complex choices about the care of their infant. Health professionals have a significant role in providing information, guidance and support. Parents facing the birth of an extremely preterm infant should have the chance to meet with both obstetrical and paediatric/ neonatal care providers to receive accurate information about their infant’s prognosis, provided with clarity and compassion. Decision making between parents and health professionals should be an informed and shared process, with documentation of all management decisions. Consultation with and transfer to tertiary perinatal centres are important for the care of both mother and fetus. As the survival of infants born before or at 22 completed weeks’ GA remains uncommon, a noninterventional approach is recommended, whereas at 23, 24 and 25 weeks’ GA, counselling about outcomes and decision making should be individualized for each infant and family, using factors which influence prognosis. All extremely preterm infants who are not resuscitated, or for whom resuscitation is not successful, must receive compassionate palliative care.

Key Words: Antenatal counselling; Ethics; Extreme prematurity; Resuscitation

In 1994, the Canadian Paediatric Society and the Society of Obstetricians and Gynecologists of Canada published guidelines for managing the anticipated birth of infants between 22 and 26 completed weeks of gestation [1]. Since then, there have been significant changes in neonatal care practices, increased survival of extremely preterm infants, renewed concerns about long-term outcomes, ongoing ethical debate about the management of infants born at the “limit of viability” and further emphasis on the role of the family in decision making. This statement provides updated recommendations to help guide health professionals and families facing the anticipated birth of an extremely preterm infant. For the purposes of this document, the terms ‘parents’, ‘mother’ and ‘father’ all refer to those expecting the birth of a child, as well as those whose child has been born.

Extremely preterm birth raises complex issues of balancing what is best for the mother, the infant, the family and society. In the first trimester of pregnancy, respect for autonomy of the mother and consideration for her well-being usually take precedence in management decisions about the pregnancy. As pregnancy progresses, the survival and health of the fetus or newborn are increasingly considered. Decisions may be complicated by the differing legal status of the fetus and newly born infant in Canada. After birth, health professionals have the responsibility of ensuring that the best interests of the infant take precedence.

The imminent birth of an extremely preterm infant creates distress and uncertainty for families and health professionals. Parents face complex choices about the resuscitation and care of their infant. Health professionals wish to provide appropriate guidance and support. Concerns about long-term neurodevelopmental morbidity, quality of life and associated increased health care costs are complex and controversial, and health professionals vary greatly in their attitudes and opinions. Despite societal consultation and the development of consensus recommendations, such as those undertaken by the Nuffield Council on Bioethics in the United Kingdom [2] and the New South Wales Consensus Group in Australia [3], there is no universal agreement on approach and management. This statement provides recommendations developed by consensus despite these difficulties.

Methods

MEDLINE searches on the outcome and management of extremely preterm birth were performed and updated in 2011. Emphasis was placed on studies that reported outcomes by gestational age for extremely preterm infants born after 1995. Reference lists of publications were examined. Neonatologists, ethicists, parent advocates and international experts were consulted during statement development.
Definition of gestational age
This statement takes its definition of gestational age (GA) from the WHO’s International Classification of Diseases, 10th revision [4]. GA is expressed in terms of completed days and weeks from the first day of the last menstrual period. Thus “22 weeks” refers to the period between 22 weeks 0 days and 22 weeks 6 days. An extremely preterm birth is defined, in this statement, as one that occurs between 22 weeks 0 days and 25 weeks 6 days. In Canada, where GA is defined in accordance with the WHO’s definition, this GA range is interpreted as including babies born from 154 to 181 days inclusive.

Antenatal counselling and decision making
The goals of antenatal counselling include establishing a relationship between health care professionals and family, providing accurate information and assisting parents to make decisions about their infant’s care. Health professionals must be aware of their personal effect on parents, endeavour to understand parental experiences and values, help parents to consider their own values, foster a mutually respectful relationship, and strive to build a therapeutic alliance with parents to meet mutually understood goals. Those counselling must engage with families in a manner that is free of personal bias. Such discussions are especially challenging as they often take place in pressured and stressful situations.

Communication with parents
Parents facing the birth of an extremely preterm infant should have several opportunities to receive information and consider a plan, particularly if pregnancy continues or new information becomes available. Information that parents report as useful includes likelihood of survival, risk of disability, medical treatments, anticipated problems, the neonatal intensive care unit (NICU) experience, parenting and coping with stress [5-6]. Parents also seek compassion for and acknowledgement of their distress. Written information may help with parental understanding and recall [7].

Accurate and consistent information is crucial when many professionals are providing information to expectant parents. Joint counselling from obstetrical and neonatal teams promotes consistency of information and adherence to plans. Other strategies that can enhance consistency include interdisciplinary consensus guidelines [8] and staff information cards [9]. Case management rounds may help to resolve team disagreement and other difficult situations. Social workers and spiritual counsellors often provide support. Clearly documented and readily available antenatal consultations and updated management plans, including the management of labour, mode of delivery and care of the infant, ensure consistency and continuity of care [5-10].

Decision making
Shared decision making between health professionals and parents is strongly recommended in the perinatal setting [11] [12]. In reality, some parents are reluctant to carry the burden of decision making, others want to be involved and have the expertise to do so, while yet others do not know how to be involved. Health professionals are sometimes uncomfortable actively engaging with families in decision making and focus more on giving information than on addressing family values or preferences [13]. However, professionals have a responsibility to educate parents and to invite their engagement in shared decision making. At a minimum, professionals should find out how parents want to be involved. Patient decision aids (tools which provide information and help individuals clarify and share their values) may help parents to participate in making decisions. Patients and health professionals who use decision aids make better decisions; patients participate more, know more, and have more realistic expectations [14]. Decision aids to assist parents facing imminent preterm birth have been developed [15].

Care of the mother at risk of extremely preterm birth

Accurate assessment of gestational age
Ultrasound is the most accurate method for establishing GA [16]. Early ultrasound, between eight and 14 weeks’ gestation, when the crown-rump length is visible, is ideal. The accuracy of GA assessment by ultrasound is approximately ± 5 days during the first trimester and ± 10 days at 16 to 22 weeks’ gestation, with increasing imprecision with advancing GA [17]. Estimates of fetal weight have an accuracy of ± 10% and tend to be underestimates. Estimated fetal weight is not accurate enough for pregnancy dating at 22 to 25 weeks [18]. Obstetrical care providers should establish an estimated date of birth as early as possible and provide this information to the pregnant woman.

Place of care
Mortality and neonatal morbidity for extremely preterm infants are decreased when birth occurs at tertiary perinatal centres rather than at non-tertiary centres with subsequent transfer to Level 3 NICUs [19]. Transferring women with threatened extremely preterm birth to tertiary perinatal centres allows for consultation, maternal care and counselling by maternal-fetal medicine and neonatal-perinatal medicine specialists, as well as for the stabilization and care of the high-risk infant. Such care may prolong some pregnancies.

Antenatal therapies
The benefits of antenatal corticosteroids (ANCs) in at-risk pregnancies before 24 weeks’ GA are debated. The National Institutes of Health consensus statement excluded these infants because few ANCS trials included them [20]. However, some data show improved survival with ANCs [21]. Furthermore, regression modelling suggests benefits from ANCS for
infants between 22 and 25 weeks’ GA [22]. ANCS should be offered for women at risk of extremely preterm birth when active management of the neonate is considered. Short-term tocolysis should be considered to facilitate in-utero transfer to tertiary perinatal centres and to buy time for ANCS to take effect. Administering magnesium sulphate should be considered for fetal neuroprotection [23].

Mode of delivery

Decisions regarding fetal monitoring and mode of delivery at extremely early GA are difficult. A systematic review of elective Cesarean section compared with expectant management for preterm babies demonstrated increased maternal morbidity (eg, haemorrhage, infection) with no significant benefits for the neonate [24]. Cesarean section may be necessary for obstetrical indications. In such cases, a classical Cesarean section is usually necessary before 24 weeks’ GA and includes risks of maternal blood loss [24], impaired fertility and uterine rupture in future pregnancies. Decisions regarding method of delivery should be made jointly between the mother and her obstetrical care provider.

Management of the extremely preterm infant

Management decisions for extremely preterm infants are, in part, informed by an infant’s risk of mortality and long-term disability. Survival of extremely preterm infants has increased during the past 10 to 15 years [25,26] and the following paragraphs summarize recent data for survival and long-term outcome. Providing local or regional outcomes in addition to more global statistics is also important and more immediately relevant for some parents.

Infant outcomes: Survival

Interpreting survival data may be difficult when it is not possible to determine which infants form the denominator. Some cohorts include both stillbirths and live-births, some include only live-births, and yet others include only infants admitted to NICUs. Hospital or network-based studies often report better survival rates than do regional or population-based studies. Such potential for selection bias in cohort studies of extremely preterm infants may result in overestimation of survival [27].

Variable management practices for extremely preterm infants may also bias results. Studies that report live-births but not stillbirths may exclude pregnancies not monitored during labour. Therapies that improve outcome, such as antenatal corticosteroids (ANCS), may not be reported or may be withheld. The population of infants who die before NICU admission may include infants for whom a decision was made not to resuscitate, as well as those with unsuccessful resuscitation. Active management of extremely preterm infants may result in improved survival [28,29].

Several large, population-based studies reporting survival to NICU discharge for each week of gestation are summarized in Table 1. In all but one study [34], survival rates were adjusted so that the denominator is all live-births, including infants not resuscitated and infants who died in the delivery room. Different outcomes may, in part, reflect the extent to which sites adopted an active management strategy.

<table>
<thead>
<tr>
<th>Site</th>
<th>Total number of live-births</th>
<th>Survival</th>
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<tbody>
<tr>
<td></td>
<td>≤22 wks</td>
<td>23 wks</td>
</tr>
<tr>
<td>EPICure, 2000 [30]</td>
<td>United Kingdom and Republic of Ireland</td>
<td>1185</td>
</tr>
<tr>
<td>Markestad, 2005 [31]</td>
<td>Norway</td>
<td>182</td>
</tr>
<tr>
<td>Bolisetty, 2004 [32]</td>
<td>NSW and Australian Capital Territory, Australia</td>
<td>355</td>
</tr>
<tr>
<td>Field, 2008 [33]</td>
<td>Trent, UK</td>
<td>669</td>
</tr>
<tr>
<td>Itabashi, 2009 [34]</td>
<td>Japan</td>
<td>1303</td>
</tr>
<tr>
<td>EXPRESS, 2009 [35]</td>
<td>Sweden</td>
<td>501</td>
</tr>
</tbody>
</table>

Canadian statistics are reported by the Canadian Neonatal Network (CNN) for infants admitted to 29 Canadian NICUs. From 2005 to 2010, of a total of 2893 infants, survival was 8% at ≤22 weeks, 36% at 23 weeks, 62% at 24 weeks and 78% at 25 weeks [36]. Note that these data do not include delivery room deaths or infants not admitted to a CNN site.
Nineteen studies of hospital- and network-based cohorts reporting survival rates by GA for infants born after 1995 were reviewed in detail. Similar trends were noted across studies: increasing mean survival rates with increasing GA, increasing variability among studies of survival rate with decreasing GA, with the greatest difference between survival per live-birth and survival per NICU admission found for infants of lowest GA. Overall, most centres report survival rates for infants born at ≤ 22 weeks’ GA of < 5 %, with some reporting no survivors. Cohort studies also indicate that infants born at ≥ 25 weeks’ GA cannot be considered to have a poor likelihood of survival.

**Infant outcomes: Long-term disability**

Some studies report no differences in significant short-term neonatal morbidities that are associated with long-term disability (≥ Grade 3 intraventricular hemorrhage, periventricular leukomalacia, retinopathy of prematurity and chronic lung disease/bronchopulmonary dysplasia) among surviving infants born at 22/23, 24 and 25 weeks’ GA [28][42][47].

In most follow-up studies, severe adverse neurodevelopmental outcome at 18 to 24 months is defined by at least one of the following conditions: cerebral palsy, cognitive impairment (test score ≥ 2SD below mean), seizures, blindness and/or deafness. These outcomes may not necessarily reflect longer term outcomes [48]. The usefulness of follow-up data may be limited by the necessary time lag in reporting outcomes, usually a minimum of three to four years. Outcomes may be reported based on live-births, NICU admissions, NICU survivors, or percent of those followed.

Studies reporting outcomes by GA show that the rate of severe disability is higher in survivors born between 23 and 25 completed weeks’ GA compared to more mature preterm infants [49][55]. Some studies report disability rates in extremely preterm survivors that increase with decreasing GA [52][59][49] [54][55], while others report no differences among infants born at 23, 24 or 25 weeks [7][51]. The risk of severe disability ranges from 18% to 60% in the smallest and most immature infants. A large UK-wide study (EPICURE) showed that of the 283 survivors born at ≤ 25 completed weeks’ GA, 23% had a severe disability at 30 months corrected age [51] and 22% had a severe disability at age six years [56]. At 11 years of age [57], this cohort had serious cognitive impairment (score < 2 SD below mean) in 40% and functional disability in 45%. Approximately 50% of the children were free of serious disability. Similar results were reported at eight years of age for an Australian cohort [55].

Data from three Canadian cohorts reporting percentage of survivors assessed at 18 to 24 months who were free of severe adverse neurodevelopmental disability are shown in Table 2.

**Table 2**

<table>
<thead>
<tr>
<th>Study</th>
<th>Population</th>
<th>Year of birth</th>
<th>Survival free of severe adverse neurodevelopmental outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Jacobs [54]</td>
<td>Toronto, Ontario Level 3 units (regional referrals)</td>
<td>1990 to 1994</td>
<td>36% 57% 63%</td>
</tr>
<tr>
<td>Synnes [7]*</td>
<td>B.C. Women’s Hospital and Health Centre, British Columbia (all births)</td>
<td>1999 to 2006</td>
<td>35% 37% 37%</td>
</tr>
<tr>
<td>Robertson [58]*</td>
<td>Northern Alberta Level 3 units (all births)</td>
<td>1992 to 2003</td>
<td>50% 54% 70%</td>
</tr>
</tbody>
</table>

* Data approximated from tables

Regulatory and behavioural difficulties, such as attention-deficit hyperactivity disorder, language delays, health-related issues and hospital readmissions occur more frequently in preterms and also contribute to long-term morbidity [26].

A few small studies have followed infants < 1000 grams birth-weight through to adolescence and adulthood, but these may not be representative of the wider population. The individuals followed have lower educational attainment, increased neurosensory impairment and more functional limitations in cognition, sensation, mobility and self-care when compared with term controls, although no differences in self-esteem or self-perceived health-related quality of life [59][60][61][62] were found. One large Norwegian study of long-term preterm survivors showed increased mortality into childhood and lower reproduction rates compared to term controls [63].

**Individualizing prediction using additional variables**

While GA has the strongest effect on prognosis, additional factors that influence outcome include birth at a tertiary perinatal centre [19][64], ANCS therapy [21][22], female sex [22][65] and multiplicity [21][66]. Tyson et al used four factors (sex, ANCS, birthweight and multiple birth) to further refine prognosis based on GA alone [22]. Thus, some infants born at 25 weeks’
GA with additional risk factors have a greater likelihood of death or severe impairment than some infants born at 23 weeks gestation without these factors. More recently, a Canadian graphical tool for predicting survival without morbidity and using birthweight as well as GA, has been described [67].

Even with such additional information, prediction of outcome for individual babies remains imprecise.

**Management decisions**

Parents making decisions about resuscitation of their infant consider their infant’s prognosis as well as their own experiences, expectations, values and engagement with the health care team [12]. It is important that parents feel supported in their decisions. Decisions may change, particularly if additional information becomes available or if pregnancy continues. When the care plan is uncertain, individuals capable of caring for the infant and facilitating management choices should be present at the birth. Even after the birth of an extremely preterm infant, there are often ongoing opportunities to learn more about the infant’s prognosis and to re-evaluate care plans.

A decision not to resuscitate an infant mandates that parents understand how their infant will be cared for after birth, including comfort measures and opportunities for holding their infant. They should be aware that their infant may survive for several days. Comfort measures should also be provided for infants when resuscitation is not successful or when a decision is made to discontinue life-sustaining therapies.

**Recommendations using the GRADE recommendations format:**

(See Table 3)

1. Parents facing the birth of an extremely preterm infant should have the opportunity for face-to-face discussions with their obstetrical care provider and with a neonatologist or paediatrician. If the situation permits, parents should be able to meet with their health care providers on more than one occasion. (Strong Recommendation)

2. Parents must receive information that is as accurate as possible about their infant’s likelihood of survival and long-term outcome, presented in a compassionate and clear manner. Offering supplementary written information is encouraged. Parental understanding of all information should be verified. (Strong Recommendation)

3. Decision making between parents and health professionals should be an informed and shared process. Decision aids may be helpful for parents. (Strong Recommendation)

4. Discussions and decisions concerning management of labour, mode of delivery and intensity of neonatal intervention must be explicitly recorded and available to all members of the health care team. (Strong Recommendation)

5. Counselling should be ongoing. If decisions about care of the infant change, these changes must be clearly documented. If pregnancy continues, parents should be provided with updated information. (Strong Recommendation)

6. Caregivers must obtain the most accurate information about GA and estimated fetal weight available. (Strong Recommendation)

7. When a pregnant woman is believed to be at least 22 weeks’ GA and at risk of giving birth prematurely, her health professional should consult with a maternal-fetal medicine specialist and neonatologist. (Strong Recommendation)

8. Transfer to a tertiary level perinatal centre is recommended for all women with threatened preterm birth who are thought to be at least 23 weeks’ GA and are safe to transfer. In situations where, following consultation with a tertiary centre and complete and fully informed decision making by the family, a decision has been made not to resuscitate an extremely preterm infant, care providers and the family may agree not to transfer the mother. Antenatal corticosteroids should be administered when active management is considered. (Strong Recommendation)

9. Elective Cesarean section is not recommended before 24 weeks’ GA unless for maternal indications. (Strong Recommendation)

10. At 22 weeks’ GA, since survival is uncommon, a non-invasive approach is recommended with focus on comfort care. (Strong Recommendation)

11. At 23, 24 or 25 weeks’ GA, counselling about outcomes and decision making around whether to institute active treatment should be individualized for each infant and family. (Strong Recommendation)

12. At 23 and 24 weeks’ GA, active treatment is appropriate for some infants. (Weak Recommendation)

13. Most infants of 25 weeks’ GA have improved survival and neurodevelopmental outcomes and active treatment is appropriate for these infants except when there are significant additional risk factors. (Weak Recommendation)

14. When GA or the plan of care is uncertain, all births of extremely preterm infants must be attended by individuals capable of managing the infant. (Strong Recommendation)

15. All extremely preterm infants who are not resuscitated or for whom resuscitation is not successful must receive compassionate palliative care, including warmth and pain relief. (Strong Recommendation)
TABLE 3
Criteria for recommendations using GRADE (Grading of Recommendations Assessment, Development and Evaluation) [68]

<table>
<thead>
<tr>
<th>STRONG RECOMMENDATIONS</th>
<th>WEAK RECOMMENDATIONS</th>
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<tbody>
<tr>
<td>• based on moderate to high quality evidence</td>
<td>• based on low quality evidence</td>
</tr>
<tr>
<td>• benefits outweigh harms</td>
<td>• uncertainty in estimates of benefits and harms</td>
</tr>
<tr>
<td>• clinician and patient values and preferences are consistent</td>
<td>• clinician and patient values are not consistent</td>
</tr>
<tr>
<td>• cost of the intervention is reasonable</td>
<td>• cost of the intervention is high</td>
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Acknowledgements
This statement was reviewed by the Society of Obstetricians and Gynaecologists of Canada, as well as by the Bioethics and Community Paediatrics Committees of the Canadian Paediatric Society.

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