Caring for Fetal and Neonatal Patients (and their Families) when the Prognosis is Poor

Patrick M. Jones, MD, MA, FAAP
Outline

1. Context
2. Concepts
3. Case studies
Context #1—significant advances in fetal and neonatal medicine
We Save the Lives of Babes

See what Science has done for the world's little weaklings in

The Baby Incubators
An Exhibit of Intense Heart Interest
The Most Highly Educational Feature on the Pay Streak
Leading Children's Physicians, Highly Trained Nurses—Every Modern Surgical Appliance and Medical Appliance Constantly at Hand.

Hygienic Day Nursery for Children
Rest and Assurance for Tired Mothers

Baby Incubators
Complete installations as operated by us
For Sale
To Hospitals and Amusement Parks. Address
Infant Incubator Co.,
Dr. S. Fischel, Treasurer, Dreamland, Coney Island.
NEW KENNEDY SON IS HIT BY BREATHINGAILMENT

Officer Clear In Scott Case — Edwards
But Patrolman Is Taken Off Duty In Scout Cars

Auto Plunges Into River; 2 Women Die

Baby Rushed To Specialists
Born 5½ Weeks Early; Named Patrick Bouvier

19 Nations Sign A-Ban Pact Today

Baby Rushed To Specialists
Born 5½ Weeks Early; Named Patrick Bouvier

Other Pictures on Back Page

BOSTON—President Kennedy visited his ailing baby Wednesday night in the Boston Children’s Hospital Medical Center. The 5½-weeks premature infant had been rushed there by ambulance from Otis Air Force Base, Mass., where he was born by cesarean section earlier Wednesday.

The President went into a fifth-floor room set aside for him and his family, in a white

Vital Statistics
former 23 weeker

former 23 weeker

(Yes, these are real products, actually for sale)
Context #2—the culture of miracles
There are two ways to live your life.
One is as though nothing is a miracle.
The other is as though everything is a miracle.

Albert Einstein (1879–1955)
Miracles: Preexisting Conditions

• “Miracles still occur today as in ancient times”¹
  • 79%

• A person in a persistent vegetative state can be saved by a miracle²
  • 61%
  • 20% of trauma surgeons

• My faith in God will override any prognostic information³
  • 20%

• The surrogate decision I am making for my loved-one is based solely on the prognostic information given to me by a physician³
  • 2%

¹. Pew Research Center. Religion among the millennial*: less religiously active than older Americans, but fairly traditional in other ways. 2010.
Miracles: External Influences
“Religion, spirituality, and hope guided most parents’ decision-making. Regardless of the medical information, parents maintained hope that everything would be fine. They were encouraged by friends and family members to pray for miracles, to transfer to a hospital thought capable of miracles, or to trust that a miracle would happen despite the physicians.”
Miracles: Communication

• “Most parents felt that their decisions regarding delivery room resuscitation were not affected by physicians’ typically grim predictions regarding the infant’s possibility of survival or disability. In contrast, parents were influenced by their own sense of the possibility of survival or disability, which was nearly uniformly positive.”

• “Parents explained that what they needed most from health care providers was compassion and hope that the infant could survive…Parents felt abandoned by physicians who seemed untouched by the grief of the experience or who appeared to be ‘following protocol’ or ‘acting by the book.’ Parents described these interactions as motivating them to advocate for their baby ‘against’ the health care team.”

Context #3—dramatic reduction in childhood mortality
Figure 1: Death Rates for Children Aged 1-4 Years by Race, United States, 1935-2007

Figure 2: Death Rates for Children Aged 5-14 Years by Race, United States, 1935-2007
Contexts

1. Significant advances in fetal and neonatal medicine

2. A culture of miracles

3. Dramatic reduction in childhood mortality
Concept #1— ‘prognosticating ain’t easy’
Contemplate…

• You (or your partner/spouse/S.O.) is pregnant and premature labor begins at 23 weeks gestation. Do you think intensive care is in the best interest of the fetus → baby?

• You are providing medical care for a woman who is 25 weeks pregnant. She is refusing intensive care for her fetus → baby. Do you feel comfortable honoring this decision?
Prognostication—Prenatal Data

• EPICure Study
  • Initial report on survival (2000)—Pediatrics
  • Follow-up @ 30 months (2000)—NEJM
  • Follow-up @ 6 years (2005)—NEJM
EPICure Study—Survival

• Reported on the outcome of all infants before 26 weeks gestational age in the UK and Ireland during March-December 1995

• Approximate total births in UK and Ireland during the time period studied—584,000

• Total number of births that met criteria—4,004

• Total number studied (lived to admission)—811

• Percent of total births studied—0.14%
Epicure Study—Survival

TABLE 11. Survival to 7 and 28 Days for All Admissions and Probability of Survival to Discharge for Survivors at Both Ages

<table>
<thead>
<tr>
<th>Gestation (Weeks)</th>
<th>n Admissions</th>
<th>7 Days After Birth (Fig 1, Point A)</th>
<th>28 Days After Birth (Fig 1, Point B)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>% Alive at 7 Days (n)</td>
<td>% [95% CI] of 7-Day Survivors Discharged From Hospital (n)</td>
</tr>
<tr>
<td>≤23</td>
<td>156</td>
<td>38.1 (59)</td>
<td>47.5 [34.8–60.2] (28)</td>
</tr>
<tr>
<td>24</td>
<td>298</td>
<td>56.0 (167)</td>
<td>59.9 [52.5–67.3] (100)</td>
</tr>
<tr>
<td>25</td>
<td>357</td>
<td>70.5 (249)</td>
<td>74.7 [69.3–80.1] (186)</td>
</tr>
<tr>
<td>Total ≤25</td>
<td>811</td>
<td>58.9 (475)</td>
<td>66.1 [61.8–70.2] (314)</td>
</tr>
</tbody>
</table>
EPICure Study—Survival

![Bar graph showing survival rates at different gestational weeks.](image)

- **22 wk**: 14%, 9%, 9%
- **23 wk**: 29%, 21%, 20%
- **24 wk**: 43%, 36%, 34%
- **25 wk**: 59%, 54%, 52%

Legend:
- **28d**
- **EDD**
- **Discharge**
Contemplate…

• You (or your partner/spouse/S.O.) is pregnant and premature labor begins at 23 weeks gestation. Do you think intensive care is in the best interest of the fetus → baby?

• You are providing medical care for a woman who is 25 weeks pregnant. She is refusing intensive care for her fetus → baby. Do you feel comfortable honoring this decision?
EPICure Study—6 year Follow-Up

- 314 of the cohort discharged to home
- 6 died prior to study
- 15 moved out-of-country
- 52 failed to follow-up
- Total number studied: 241
- 78% of survivors evaluated in follow-up study
EPICure Study—6 year Follow-Up

• Methods

• If subject was in a mainstream school, it was attempted to match them with an age/sex control

• Resulted in 241 subjects with 160 controls

• Standardized cognitive and neurologic assessments performed at six years of age (not corrected for gestational age)

• Disability defined as severe, moderate, or mild according to predetermined criteria
Table 2. Neurocognitive Function and Degree of Disability at Six Years of Age among 241 Extremely Preterm Children and 160 Classmates Born at Full Term.

<table>
<thead>
<tr>
<th>Disability†</th>
<th>Comparison with Standardized Data</th>
<th>Comparison with Classmates</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Comparison Group</td>
<td>Extremely Preterm Group</td>
</tr>
<tr>
<td></td>
<td>no. % (95% CI)</td>
<td>no. % (95% CI)</td>
</tr>
<tr>
<td>Severe</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cerebral palsy, nonambulatory</td>
<td>0 15 6 (4–10)</td>
<td>0 15 6 (4–10)</td>
</tr>
<tr>
<td>IQ &gt; 3 SD below mean</td>
<td>0 27 11 (8–16)</td>
<td>0 50 21 (16–26)</td>
</tr>
<tr>
<td>Range, 39–54</td>
<td>0 27 11 (8–16)</td>
<td>0 50 21 (16–26)</td>
</tr>
<tr>
<td>Range, 39–69</td>
<td>0 27 11 (8–16)</td>
<td>0 50 21 (16–26)</td>
</tr>
<tr>
<td>Profound sensorineural hearing</td>
<td>0 7 3 (1–6)</td>
<td>0 7 3 (1–6)</td>
</tr>
<tr>
<td>loss</td>
<td>0 6 2 (1–5)</td>
<td>0 6 2 (1–5)</td>
</tr>
<tr>
<td>Blind</td>
<td>0 6 2 (1–5)</td>
<td>0 6 2 (1–5)</td>
</tr>
<tr>
<td>Any severe disability</td>
<td>0 32 13 (9–18)</td>
<td>0 53 22 (17–28)</td>
</tr>
<tr>
<td>Moderate</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Abnormal neurologic findings</td>
<td>0 17 7 (4–11)</td>
<td>0 17 7 (4–11)</td>
</tr>
<tr>
<td>with functional loss but</td>
<td></td>
<td></td>
</tr>
<tr>
<td>ambulatory</td>
<td></td>
<td></td>
</tr>
<tr>
<td>IQ &gt; 2 to 3 SD below mean</td>
<td>0 23 10 (6–14)</td>
<td>2 1 (0–4)</td>
</tr>
<tr>
<td>Range, 55–69</td>
<td>0 23 10 (6–14)</td>
<td>2 1 (0–4)</td>
</tr>
<tr>
<td>Range, 70–81</td>
<td>0 23 10 (6–14)</td>
<td>2 1 (0–4)</td>
</tr>
<tr>
<td>Sensorineural hearing</td>
<td>1 1 (0–3)</td>
<td>7 3 (1–6)</td>
</tr>
<tr>
<td>loss corrected with hearing</td>
<td></td>
<td></td>
</tr>
<tr>
<td>aids</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Impaired vision but ability to</td>
<td>0 11 5 (2–8)</td>
<td>0 11 5 (2–8)</td>
</tr>
<tr>
<td>see</td>
<td>0 11 5 (2–8)</td>
<td>0 11 5 (2–8)</td>
</tr>
<tr>
<td>Any moderate disability</td>
<td>1 1 (0–3)</td>
<td>27 11 (8–16)</td>
</tr>
<tr>
<td>Mild</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Neurologic signs, minimal</td>
<td>0 26 11 (7–15)</td>
<td>0 26 11 (7–15)</td>
</tr>
<tr>
<td>functional impairment</td>
<td></td>
<td></td>
</tr>
<tr>
<td>IQ &gt; 1 to 2 SD below mean</td>
<td>3 2 (0–5)</td>
<td>61 25 (20–31)</td>
</tr>
<tr>
<td>Range, 70–84</td>
<td>3 2 (0–5)</td>
<td>61 25 (20–31)</td>
</tr>
<tr>
<td>Range, 82–94</td>
<td>2 1 (0–4)</td>
<td>10 4 (2–7)</td>
</tr>
<tr>
<td>Mild hearing impairment</td>
<td>7 4 (2–8)</td>
<td>69 29 (23–35)</td>
</tr>
<tr>
<td>Squint or refractive error</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Any mild disability</td>
<td>9 6 (3–10)</td>
<td>71 29 (24–36)</td>
</tr>
<tr>
<td>No disability</td>
<td>150 94 (89–97)</td>
<td>111 46 (40–53)</td>
</tr>
</tbody>
</table>
EPICure Study—6 year Follow-Up

Overall Cognitive Disability

- none
- mild
- mod
- severe

23 wga
24 wga
25 wga
All infants

percentage

0
20
40
60
80
100
EPICure Study—6 year Follow-Up

Gestational Age (completed wk)

- Boys: N=7, N=37, N=78
- Girls: N=17, N=36, N=66

Comparison Group: N=71, N=89

Cognitive Score

- ≤23 wk
- 24 wk
- 25 wk
EPICure Study—Summary
Contemplate…

• You (or your partner/spouse/S.O.) is pregnant and premature labor begins at 23 weeks gestation. Do you think intensive care is in the best interest of the fetus → baby?

• You are providing medical care for a woman who is 25 weeks pregnant. She is refusing intensive care for her fetus → baby. Do you feel comfortable honoring this decision?
Concept #2— ‘simplicity on the other side of complexity’
How do I care for a baby with a serious illness if we are not going to do ________?

• Bag-mask ventilation, CPAP, intubation
• Chest compressions
• Orogastric/nasogastric tube placement
• Corrective surgeries (cardiac, neurosurgical, GI)
• Placement of IV access
• Resuscitative medications
• Diagnostic/lab testing
How do I care for a baby with a serious illness if we are not going to do ________?

The same as any other baby.
How do I care for a baby with a serious illness if we are not going to do __________?

Okay... maybe not the exact same as any other baby.

care of the dying neonate
Unpacking the black box

• Changing the focus
  • Does this intervention for my patient and/or the family maintain or improve their health?
  • Does this intervention for my patient and/or the family maintain or improve their comfort*?
• Rigorously approach the use of any intervention in light of this goal
• Reevaluate the effect of each intervention in light of this goal
Unpacking the black box—potential ways that interventions may provide comfort

• Bag-mask ventilation, CPAP, intubation

• Chest compressions

• Orogastric/nasogastric tube placement

• Corrective surgeries (cardiac, neurosurgical, GI)

• Placement of IV access

• Resuscitative medications

• Diagnostic/lab testing
Unpacking the black box—treating respiratory distress without your usual tools

- Morphine—0.1-0.2 mg/kg/dose (IV, PO, oral mucosa)
  - Onset: IV—5 minutes, oral—up to 30 minutes
  - Peak: 30 min - 1 hour
  - Duration: 3-4 hours
  - Mechanism of action: Blunts “panic” response to elevated CO2 and low pH

- Blow-by or nasal cannula O2
  - Limited usefulness
  - If no improvement, reevaluate in light of it’s potential to interfere with bonding
How do I care for a baby with a serious illness if we are not going to do ________?

• Simplicity—The same as any other baby

• Complexity—Okay, maybe not the exact same as any other baby.

• Simplicity—The same as any other baby, just with a different focus
Case Studies
Prenatal Consult—Overview

- Mother referred to Texas Fetal Center due to concern for CDH
- Further investigation revealed large left lung mass with hypoplastic right lung
- Mother strongly believes that “God will bring a miracle”
Perinatal Consult—Process

- Takes place in a private, quiet room
  - Diagnoses are taken into consideration re: location
  - Other commitments are minimized as much as possible
- Introductions are made
  - Family
  - Palliative care team
- Family is asked how they would like for us to refer to their fetus (frequent request is to use the future name)
Perinatal Consultation—Process

• The agenda is open but with plenty of available leading
  • “Help me know if we have done a good job of communicating…”
  • “Are there any things you definitely want to do or discuss today?”
  • “Tell me what you and your family have been talking about since you received the diagnosis of ________for ________.”

• Birth plan is started
  • Both benign and potentially emotional details
  • Often is ongoing project with family long after initial consult

• Tour of the NICU (as appropriate)
Miracles: Communication
Prenatal Consult—Results

• Patient delivered in presence of multi-disciplinary team

• Intubated in the delivery room and stabilized in NICU

• Successful surgical resection

• Palliative care team involved throughout stay to support family

• Palliative care also sees patient during readmissions to CMHH
Pediatric Consult—Overview

• Neonate transferred to Children’s Memorial Hermann Hospital at 14 days of life due to concerns for thanatophoric dysplasia

• Admitted after delivery on room air, progressed to CPAP of 10 cm H20, transferred after intubation on day of life 14

• Physical exam and radiographic findings consistent with diagnosis

• Primary team recommends removal of ventilator support

• Mother strongly believes that “God will bring a miracle if we wait”
Pediatric Consult—Process

- Common goals
  - Get patient home
  - Keep patient pain-free and with her family
- Weaning plan created with the family
  - Attempt to wean CPAP every 3 days, treat worsening respiratory distress with opioids instead of more CPAP
  - If unable to wean, remove CPAP and move patient to private room for additional privacy and intensive end-of-life care
Pediatric Consult—Results

• Plan revisited several times over a 2 week period

• Unable to get below CPAP of 6, patient taken to private room and taken off CPAP after spending time together as a family

• Several doses of morphine given during initial 6 hours to help ease respiratory distress

• Patient at 24 hours--pink and comfortable on room air

• Team celebrates, plan readdressed, discharged home with hospice support

• Patient dies peacefully at home three months later
Obligatory inspirational image...
...so you don’t leave depressed.