Treatment decisions for preemies and babies with trisomies

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Children’s Mercy Bioethics Center, KCMO
Goals

• First, talk about preemies and why we decide to treat or not to treat
• Second, talk (more briefly) about similar decisions for babies with trisomy 13 & 18.
• Discussion of the ethical issues
My talk will be mostly about this graph.
My talk will be mostly about this graph.

At 24-26 weeks, almost all babies received active treatment.

Rysavy et al, NEJM 2016
My talk will be mostly about this graph

Figure S1. Rates of Neonatal Active Treatment for Infants Born at 22 to 26 Weeks’ Gestation in 24 Hospitals in the NICHD Neonatal Research Network

At 23 weeks, every center offered treatment to at least some babies. Some offered treatment to all.

Rysavy et al, NEJM 2016
My talk will be mostly about this graph.

Figure S1. Rates of Neonatal Active Treatment for Infants Born at 22 to 26 Weeks’ Gestation in 24 Hospitals in the NICHD Neonatal Research Network

- 22 Weeks (n=357)
- 23 Weeks (n=755)
- 24 Weeks (n=1152)
- 25 Weeks (n=1262)
- 26 Weeks (n=1332)

5 hospitals provided “active treatment to 0% of 22 weekers

Rysavy et al, NEJM 2016
Most of my talk will be about this graph.

Figure S1. Rates of Neonatal Active Treatment for Infants Born at 22 to 26 Weeks’ Gestation in 24 Hospitals in the NICHD Neonatal Research Network

7 hospitals provided active treatment to 100% of 22 weekers.
Most of my talk will be about this graph.

Figure S1. Rates of Neonatal Active Treatment for Infants Born at 22 to 26 Weeks’ Gestation in 24 Hospitals in the NICHD Neonatal Research Network

12 hospitals provided active treatment to some but not all 22 weekers.
Hospital policies and mortality

• Strongest predictor of survival for babies under 25 weeks is whether the hospital in which they are born offers treatment.
  – Hospitals that treat more, save more
  – Policies to restrict treatment lead to avoidable deaths.
• The hospital with the best survival rates, University of Iowa, reports 40-50% survival for babies born at 22 weeks.

• And higher survival rates than most other places at 23 and 24 weeks.
## Survival of Inborn “Extremely Extreme Premature” Infants

22 - 24 weeks EGA (2006-2014)

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Survival of Inborn VLBW Infants
22 - 24 weeks EGA

NRN data: JAMA September 8, 2015; Volume 314, Number 10
How do they do it?
Begin at the Beginning

Antenatal Steroids are Critical
Interdisciplinary Teamwork with MFM

• ANS at 22 – 25 weeks gestation:
  ✓ Reduces death!
  ✓ Reduces severe IVH!
  ✓ Reduces the incidence of NDI at 18 to 22 months!

In the NICU
Standardization of Care
Golden Hour Protocol

Golden Hour Goals:
1) Admission temperature ≥ 36.0
2) Surfactant given
3) Dextrose infusion started
4) Antibiotics started
5) Communication post-delivery with mom
Standardized Ventilator Goals

1) 1\textsuperscript{st} Intention HFV Center at Iowa
   ✓ High Frequency Jet Ventilation for all infants < 25 weeks at birth

2) Critically important to avoid volutrauma (shear force injury) to the lung especially at 22 to 23 weeks gestation
   ✓ Follow pCO\textsubscript{2} levels closely with rigid adherence to goals to avoid fluctuations in Cerebral Blood Flow
     1) Target 45 - 55 first 3 days
     2) Target 45 - 60 next 4 days
     3) Gases Q2-3 hours or more frequently in the beginning
     4) After ventilator change, repeat in 20 minutes
These data raise an interesting question:

- Is there any other situation in which...
  - A group of patients have a disease that is uniformly fatal without treatment.
  - Some centers, trying an innovative approach, report 40-50% survival rates
  - Other centers
    - Do not offer the treatment.
    - Do not want to learn about the treatment.
    - Argue that it would be unethical to offer the treatment.
  - And many bioethicists support them.
Weird
I know what you are thinking

• Gestational age is a predictor of both mortality and severe neurocognitive impairment.
• And parents don’t want their tiny babies saved
• And it costs too much

• All these claims are wrong, some more than others.
Gestational age predicts morbidity

- Gestational age is a predictor of both mortality and severe neurocognitive impairment.
- But, at the threshold of viability (22-25 wks):
  - Gestational age is associated with survival
  - Gestational age is NOT associated with neurological outcomes among survivors
And in the UK: Disability Rates at 30 months of age
-- From Woods et al, NEJM 2000

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<tr>
<th>Gestational age (wks)</th>
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<tr>
<td>psychomotor</td>
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<td>No developmental</td>
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<td>disability</td>
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<tr>
<td>No neuromotor</td>
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<td></td>
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<tr>
<td>disability</td>
<td>85</td>
<td>74</td>
<td>76</td>
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<tr>
<td>No sensory</td>
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<tr>
<td>disability</td>
<td>58</td>
<td>64</td>
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Similar data from Iowa

• 22 weekers who survive have the same rates of IVH, BPD, and neurocognitive impairment as 23 and 24 weekers.
• If the goal is to reduce neuro impairment in society, we’d have to draw the line at 26 or 28 weeks.
A paradox

• More babies survive with disability at higher birth weights than at lower ones, even if rates of disability are higher in smaller babies.

• Consider 100 babies.
  – In one group: 90% survival; 20% severe disability
  – In one group: 20% survival; 50% disability

• 20% of 90 = 18; 50% of 20 = 10
People seem oddly impervious to the data
ACOG/SMFM statement

“Delivery before 23 weeks typically results in neonatal death irrespective of newborn resuscitation (5-6% survival) and, among rare survivors, significant morbidity is universal. (98-100%).”

After criticism, revised in 2017

- Among babies born between 22 and 24 weeks at 11 centers in the US from 2008=2011, most neonates died (64%) or were severely impaired (16%).

- A little misleading: most died, but, among survivors (36%), most were NOT severely impaired (20% vs. 16%).
Two (true) ways to “frame” the data

• Most babies (80%) will either die or survive with severe impairments.
• Many babies will die, most of the survivors do not have severe impairments.
Self-fulfilling prophecies??

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<td>22%</td>
<td>72%</td>
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Rysavy et al, NEJM, 2015
Of those, 23% survived

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Rysavy et al, NEJM, 2015
How many 500g, 23 week singletons survive unimpaired?

- Boys, no steroids: 5%
- Boys, steroids: 11%
- Girls, no steroids: 9%
- Girls steroids: 18%

Three things to note:
1. Fourfold difference in survival at same BW and GA.
2. Survival rates double if given steroids.
3. Doesn’t distinguish death from disability in survivors.

http://www.nichd.nih.gov/about/org/der/branches/ppb/programs/epbo/Pages/epbo_case.aspx
How many 500g, 23 week singletons who survive are unimpaired?

- Boys, no steroids  5%  50%
- Boys, steroids     11%  55%
- Girls, no steroids 9%  67%
- Girls steroids    18%  67%

Very different if the statistic is “disability among survivors,” rather than “overall survival without disability.”
Key question

Is it worse to have tried and failed than not to have tried at all?

OR, to put it another way

Is it better not to offer treatment and let a preemie die, or to offer a trial of therapy and withdraw treatment if things look bad?
Simply a bias against preemies?
Consider curious cases of consensus

• Case #1: A previously healthy 2-month-old baby develops fever, irritability, listlessness, and a rapidly spreading rash. A lumbar puncture shows that he has meningitis. If the baby survives, he will likely have severe neurological impairment.

• Case #2: A baby is born at 23 weeks of gestation and 550 grams, with Apgar scores of 3 and 6. He is intubated and given oxygen and his color and tone improve.

Two cases

• In many hospitals (as Rysavy showed), initial treatment would be considered optional for the baby in case #2.
• It would be automatic in all hospitals for Case #1
• Prognosis is better for the baby in case #2.
• Why the difference?

Another thought experiment

- Case #1: A baby is born at 22 weeks of gestation and 500 grams, with Apgar scores of 3 and 6. He is intubated and given oxygen and his color and tone improve.

- Case #2: An 84 year old (without an advance directive) comes to the ER. He is diaphoretic, short of breath, with chest pain and ST elevation on EEG.
Figure 2. **Unadjusted Rates of Survival to Hospital Discharge by Calendar Year.**

Observed (crude) rates for survival to discharge are shown for the overall cohort and separately for shockable cardiac-arrest rhythms (ventricular fibrillation [VF] and pulseless ventricular tachycardia [VT]) and nonshockable cardiac-arrest rhythms (asystole and pulseless electrical activity [PEA]).

P < 0.001 for trend for each survival curve.
Survival of Inborn “Extremely Extreme Premature” Infants
22 - 24 weeks EGA (2006-2014)

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There is something about preemies

1. *Residents:* All 172 residents in pediatrics and obstetrics in Quebec, Canada.

2. *Nurses:* 136 nurses of the McGill University Health Centre involved in perinatal and neonatal care: delivery room nurses, nurses working in the maternity hospital and the Children’s Hospital.

Something about preemies

• Two case vignettes
• One identified the patient as a 24 week preemie, the other didn’t, but simply gave outcomes that were those of a 24 weeks preemie
• “Would you resuscitate?”
Figure 1  Percentage of respondents replying that they would always or generally resuscitate a preterm baby known to be a 24-week infant compared to an infant with outcomes described but no gestational age given.

What do parents want?
Surveys of parent attitudes:

Parents generally want more treatment than doctors and nurses think is appropriate.
Most say they want “everything.”

Parent and professional agreement with the statement: “I believe an attempt should be made to save all infants regardless of birth weight.”

Streiner et al
Peds, 2001
More likely than HCWs to say we should try to save babies “at all costs.”


Copyright ©2009 American Academy of Pediatrics
More likely to rank “death” lower than “severe global impairment”

1. Death.

2. Severe global impairment – wheelchair, intelligence of 1y.o., unable to speak, read or write, incontinent, no independent ADLs.

5. Moderate global impairment – crutches, attends special school, cannot read or write, unable to live independently, continent.
Is severe disability is worse than death?

• Doctors and nurses - 55%
• Mothers of term babies – 40%
• Parents of preemies – 25%
A modest proposal

• Stop using gestational age as the basis for policies about which baby to treat in the DR.

• Instead....
A modest proposal

• Stop using gestational age as the basis for policies about which baby to treat in the DR.

• Instead....
  – Talk to the parents
  – Examine the patient
  – Make a recommendation
Two “elephants” in the room

1. Abortion politics
   – To save 22 weekers is to admit that Roe v. Wade got it wrong....and to raise questions about the rights of fetuses – and pregnant women - in the second trimester.
Two “elephants” in the room

1. Abortion politics
   – To save 22 weekers is to admit that Roe v. Wade got it wrong....and to raise questions about the rights of fetuses – and pregnant women - in the second trimester.

2. Institutional political culture
   – If we ask parents, many will want treatment
   – We will need to learn from places like Iowa about best practices –
   – Tiny baby units: NICUs on steroids
Conclusions

• Survival rates improving for 22 weekers.
• Non-treatment is a self-fulfilling prophecy
• A policy in favor of treatment will be welcomed by most parents.
Prediction

• We will see more centers treating more 22 weekers with better outcomes.
• More parents will want such treatment
• More centers will follow the Iowa model – a “small baby unit” within the NICU.

—And outcomes will improve. 😊
TRISOMY 13 AND 18: ethics in evolution
An Ethical Dilemma

“...it is morally and socially problematic to use resources and increase patients’ suffering without providing medical benefit. It is equally problematic to categorize a condition as lethal, if by doing so the use of potentially beneficial therapy is foreclosed.”

- J Perinatol 2011
Guiding Principles

• Good ethics begins with good facts
• Counseling and care should be individualized to each patient and family
• What are the facts? Are trisomies “lethal anomalies?”
AAP Recommendations

“Family members should be provided with an overview of the potential complications...They should be informed of the range of survival rates and of the types and rates of long-term disabilities that can be expected.

“Parents should be given the most accurate prognostic morbidity and mortality data available for their infant. In some situations, these may be hospital-specific data, and in other situations, regional or national data may be more appropriate.”

-Pediatrics, 2002 & 2015, Committee on Fetus and Newborn.

Similar principles may be applied to infants with trisomy 13 or 18
Another self-fulfilling prophecy?

- Because institutional approaches can affect outcomes, applying only local data may create a self-fulfilling prophecy. The number of such infants born at a given institution may be so low that local data may be hard to interpret; using both local and outside data may be helpful.

  - AAP Comm on Fetus and Newborn, Pediatrics, 2015
Trisomy 18 and 13: Historical Dogma

• Resuscitation at delivery and aggressive interventions are not warranted, and can be considered futile care.

• Cardiac surgery has not been justified by most institutions worldwide.
  – This is based on population-based studies citing $\geq 90\%$ mortality, with central apnea as the most common cause of death.
Caveats to the data

• Many babies dies of treatable conditions.
  – Bella Santorum story
    • Trisomy 13
    • After ten days in the NICU, Bella was stable on supplemental oxygen.
    • Plan – home on hospice.
    • Rick and Karen wanted to continue providing oxygen. (Karen had been a NICU nurse)
    • The doctors wanted to discontinue the oxygen. One doctor bluntly told the Santorums, “You realize that your child is going to die. You have to learn to let go.”
They got the oxygen. Bella survived. A few months later, she had a viral upper respiratory infection that led to a respiratory arrest. Her parents provided CPR. Again, she survived. She is now 8 years old.

Many parents like the Santorums.

• They understand that their babies may die.
• They want them to be treated like other babies.
• When treated, survival rates improve.
Survey of Parents: Description of Children with Trisomy 13 or 18

<table>
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<th>Characteristic</th>
<th>All Children (n = 272), n (%)</th>
<th>Full T13-18 (n = 216), n (%)</th>
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Janvier, Farlow, Wilfond, 2012
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Janvier, Farlow, Wilfond, 2012  (Note, this is among parents in on-line support groups – clearly an overestimate. We don’t know how much of an overestimate
Survey of Parents:
Survival Not Clearly Related to Level of Intervention

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<th>Level of Intervention</th>
<th>Died Before Discharge, n (%)</th>
<th>Died &lt;1 wk, n (%)</th>
<th>Died &lt;3 mo, n (%)</th>
<th>Lived &gt;1 y, n (%)</th>
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<tr>
<td>Full intervention (n = 53)</td>
<td>17 (32)</td>
<td>9 (17)</td>
<td>19 (36)</td>
<td>28 (53)</td>
<td>13 (25)</td>
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<tr>
<td>T13 (n = 19)</td>
<td>7 (37)</td>
<td>3 (16)</td>
<td>8 (42)</td>
<td>10 (53)</td>
<td>5 (26)</td>
</tr>
<tr>
<td>T18 (n = 34)</td>
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<td>6 (17)</td>
<td>11 (32)</td>
<td>18 (53)</td>
<td>8 (24)</td>
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<tr>
<td>In between full intervention and comfort care (n = 59)</td>
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<td>24 (41)</td>
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<td>15 (25)</td>
</tr>
<tr>
<td>Comfort care (n = 104)</td>
<td>37 (36)</td>
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Self-Fulfilling Prophecy?

- There *are* some long-term survivors with trisomy 13 and 18
- Cardiac defects and heart failure contribute to death in these infants
- Parents are requesting, and physicians providing, more aggressive interventions
Parental Perspectives

Parental hopes:
- Child born alive (80%)
- Child exceeds expectations, becomes one of the survivors (20%)
- Child remains comfortable, free from pain or suffering (10%)
- Child feels loved (12%)
- Hope for misdiagnosis, or a miracle (4%)

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Changing Paradigm

Intensive Cardiac Management in Patients With Trisomy 13 or Trisomy 18

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Neonatal Management of Trisomy 18: Clinical Details of 24 Patients Receiving Intensive Treatment

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Outcomes of cardiac surgery in trisomy 18 patients

Jun Muneuchi,1 Junko Yamamoto,1 Yasuhiko Takahashi,1 Mamie Watanabe,1 Tetsuji Yuge,1 Takuro Ohno,1 Yutaka Imoto,2 Akira Sese,2 Kunitaka Joo1

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An Analysis of Cardiac Defects and Surgical Interventions in 84 Cases with Full Trisomy 18

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Cardiac surgery for children with trisomies 13 and 18: Where are we now?
Annie Janvier, MD, PhD\textsuperscript{a,b,c,*}, Barbara Farlow, BEng, MBA\textsuperscript{d,e}, and Keith Barrington, MD\textsuperscript{a,c}

Effectiveness of Cardiac Surgery in Trisomies 13 and 18 (from the Pediatric Cardiac Care Consortium)
Eric M. Graham, MD, Scott M. Bradley, MD, Girish S. Shirali, MBBS, Christine B. Hills, BA, and Andrew M. Atz, MD

Evolving medical and surgical management of infants with trisomy 18
John M. Lorenz\textsuperscript{a} and George E. Hardart\textsuperscript{b}
Trisomy 18: A Single-Center Evaluation of Management Trends and Experience with Aggressive Obstetric or Neonatal Intervention


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Survival of Children with Trisomy 13 and Trisomy 18: A Multi-State Population-Based Study

Robert E. Meyer, Gang Liu, Suzanne M. Gilboa, Mary K. Ethen, Arthur S. Aylsworth, Cynthia M. Powell, Timothy J. Flood, Cara T. Mai, Ying Wang, and Mark A. Canfield

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Finding the Right Balance

- **Information**
  - Accurate, balanced

- **Respect**
  - For the infant, and for parental values

- **Collaboration with families**

- **Individualized care**
  - Provide appropriate therapies and interventions
  - Avoid unnecessarily burdensome treatments
  - *Focus on quality of life*
Thanks