Time, future, and genetics: the temporal complexity of disability

Rebecca Mueller, PhD, LCGC
ELSI Postdoctoral Fellow
Medical Ethics and Health Policy

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Twice-told Tales: Stories about Genetic Disorders

Abby Lippman* and Benjamin S. Wilfond†

*Department of Epidemiology and Biostatistics, McGill University, Montreal; and
†Department of Pediatrics, University of Wisconsin—Madison, Madison
Overview

• Medical perspective on cystic fibrosis’s prognosis
• Prognostic messages from perspective of CF patients
• How prognosis informs life plans
• Societal structures manifesting temporal ableism
• Key disability theory on time, future, and kinship
• Recommendations for specific stakeholders
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A brief history of Cystic Fibrosis

- Median Survival (yrs)
- Pathological Description
- CF Center Care
- CFTR Identified
- Highly effective CFTR modulator
- 1930: 0 years
  - 1940: 5 years
  - 1950: 10 years
  - 1960: 20 years
  - 1970: 30 years
  - 1980: 40 years

- Treatment Timeline:
  - 1930: Pancreatic Enzymes
  - 1950: Staph Antibiotics
  - 1960: Pseudomonal Antibiotics
  - 1970: rhDNase
  - 1980: TOBI
  - 1990: Chronic Azithromycin
  - 2000: 7% Saline
  - 2010: AZLI
FDA NEWS RELEASE

FDA approves new breakthrough therapy for cystic fibrosis

Treatment approved for approximately 90% of patients with cystic fibrosis, many of whom had no approved therapeutic options

For Immediate Release: October 21, 2019
A brief history of Cystic Fibrosis

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Pathological Description

CF Center Care

CFTR Identified

Highly effective CFTR modulator


0 10 20 30 40

AZLI
7% Saline
Chronic Azithromycin
TOBI
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Pseudomonal Antibiotics
Staph Antibiotics
Airway Clearance
Pancreatic Enzymes
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ALEX
The Life of a Child
Frank Deford

A true story of an extraordinary life

Based on the bestselling book by FRANK DEFord
Produced by NICOLAS BRASION
Directed by ROBERT MARRS
[At age six or seven] “…we were driving in the car, and I asked my mom how old people with CF live. And her whole body tensed up and she got really teary eyed. And she told me the average at that time was 25. And I definitely had that in my head growing up and I felt so compelled that I wanted to live a huge life in whatever time I had.”

-Hanna, 30s, mild FEV1
“…Like you have CF and you're how old and she just like did her treatments when we did our treatments and… it was an impactful moment because I could at that point imagine being an adult with CF. Like that is, you know, that will happen. I could go into the world and be an adult with CF.”

-Hanna, 30s, mild FEV1
Prognostic Imagination:
The visualized projections that an individual can see, hope, plan, or fear for their lives
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Prognostic Imagination:
The visualized projections that an individual can see, hope, plan, or fear for their lives.
“… I felt so compelled that I wanted to live a huge life in whatever time I had. I went to college at 15, full time. I worked full time. I got married a week after I turned 18, moved 900 miles from home….I wanted to fit everything in…”

-Hanna, 30s, mild FEV1
“Experience is seriously important to me, you know... I've done, I've done more than most people that are healthy do.”

- John, 40s, lung transplant
“Now I'm going to turn 50 and it's like, ‘okay, have I wasted my life?’ But then I think, well I got all the medical care I needed because I was on disability. I never had to worry like other people, but I couldn't work. I have a college degree, but I couldn't work. I couldn't get married because I’d lose my benefits. Like now what do I do? Because here I am 50, and they're like, ‘you know, this probably isn't going to kill you.’ It's like, really? This is 25 years too late for me to work or get a retirement plan or whatever you do ….

-Alexandra, 40s, mild FEV1
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## Erikson's Stages of Psychosocial Development

<table>
<thead>
<tr>
<th>Stage</th>
<th>Psychosocial Crisis/Task</th>
<th>What Happens at This Stage?</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Trust vs Mistrust</td>
<td>If needs are dependably met, infants develop a sense of basic trust.</td>
</tr>
<tr>
<td>2</td>
<td>Autonomy vs Shame/Doubt</td>
<td>Toddlers learn to exercise will and do things for themselves, or they doubt their abilities.</td>
</tr>
<tr>
<td>3</td>
<td>Initiative vs Guilt</td>
<td>Preschoolers learn to initiate tasks and carry out plans, or they feel guilty about efforts to be independent.</td>
</tr>
<tr>
<td>4</td>
<td>Industry vs Inferiority</td>
<td>Children learn the pleasure of applying themselves to tasks, or they feel inferior.</td>
</tr>
<tr>
<td>5</td>
<td>Identity vs Confusion</td>
<td>Teenagers work at refining a sense of self by testing roles and then integrating them to form a single identity, or they become confused about who they are.</td>
</tr>
<tr>
<td>6</td>
<td>Intimacy vs Isolation</td>
<td>Young adults struggle to form close relationships and to gain the capacity for intimate love, or they feel socially isolated.</td>
</tr>
<tr>
<td>7</td>
<td>Generativity vs Stagnation</td>
<td>The middle-aged discover a sense of contributing to the world, usually through family and work, or they may feel a lack of purpose.</td>
</tr>
<tr>
<td>8</td>
<td>Integrity vs Despair</td>
<td>When reflecting on his or her life, the older adult may feel a sense of satisfaction or failure.</td>
</tr>
</tbody>
</table>

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<table>
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<tr>
<th>Age Range</th>
<th>Developmental Tasks</th>
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| Infancy and Early Childhood | • Learn to walk  
• Learn to use the toilet  
• Learn to talk  
• Learn to form relationship with others |
| 0-5 years old               |                                                                                     |
| Middle Childhood            | • Learn school-related skills such as reading  
• Learn about conscience and values  
• Learn to be independent                                      |
| 6-12 years old              |                                                                                     |
| Adolescence                 | • Establish emotional independence  
• Learn skills needed for productive occupation  
• Achieve gender-based social role  
• Establish mature relationships with peers |
| 13-17 years old             |                                                                                     |
| Early Adulthood             | • Choose a life partner  
• Establish a family  
• Take care of a home  
• Establish a career                                             |
| 18-35 years old             |                                                                                     |
| Middle Age                  | • Maintain a standard of living  
• Perform civic and social responsibilities  
• Maintain a relationship with spouse  
• Adjust to physiological changes                              |
| 36-60 years old             |                                                                                     |
| Later Maturity              | • Adjust to deteriorating health  
• Adjust to retirement  
• Meet social and civil obligations  
• Adjust to loss of spouse                                      |
| Over 60 years old           |                                                                                     |
Curative time is not about any individual’s relationship with medical interventions or an individual desire for cure but rather “an understanding of disability that not only expects and assumes intervention but also cannot imagine or comprehend anything other than intervention.”

-Alison Kafer 2013, 27
The new kinship imaginary

“…As disabled kin move through the life cycle, their atypical experiences reverberate into the lives of their families in ways that reframe taken for granted assumptions. Family members find themselves recognizing and reorganizing tacit norms about familial relations and the temporality of the domestic cycle; in that process, the culturally ordered unfolding of a normative life course can no longer be assumed.”

-Ginsburg and Rapp 2017, 181
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Recommendations for clinicians

• Normative expectations underly many developmental models

• Normative developmental expectations like complete independence may cause harm or alienate certain patients and families given the realities of the disease

• Adapt models and expectations to promote interdependence
Recommendations for medical providers

• Think critically about prognostic communication beyond the moment of diagnosis
• Be proactive and thoughtful in supporting prognostic communication as a child grows up
• Embrace prognostic imagination to recognize that prognostic ideas impact decisions and plans for the future
Recommendations for researchers and advocacy organizations

• Consider views and needs of community stakeholders in research design and plans to communicate findings

• For example:
  • Ask who is helped and hurt with median life expectancy data
  • Think about how to aggregate and communicate data when the disease under study has a broad phenotypic spectrum
Recommendations for social policy research and policy-makers

• Rethink disability categories that direct accommodations and eligibility for social services to reflect variable temporalities of disability

• Ensure diagnoses create options rather than constrict possibilities

• Promote policy and societal change to build accessible futures
Thank you

Rebecca.mueller@pennmedicine.upenn.edu
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• See final sides for references and image credits
Key References – Further Reading


Key References – Further Reading


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