

Adulting with CCHS: Navigating Your Path Forward

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Disclosures

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- DSMB membership: Stimdia
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Outline and Goals

- What are the challenges in transitioning to an adult health system?
- Are there strategies that can smooth the transition?
- How do I find a doctor (and a team)?
- What else do I need to get good long-term care?
- How can the adult healthcare world address CCHS care challenges?

My background

- Medical school: Washington University in St. Louis, Residency in Internal Medicine: UCLA
- Pulmonary Disease and Critical Care Medicine Fellowship: UCSD
 - Sleep Medicine
- Research focused on physiology of sleep-related breathing disorders
- Founded a clinical program: Pulmonary Neuromuscular and Assisted Ventilation

My Philosophy: Supporting patients with breathing disorders is a foundation of adult pulmonary medicine



Adult-focused health systems differ from pediatric health systems

- Relatively few young patients
- Parents are not legally required to be involved
- Primary care is less available and often less comfortable with this group
- Lack of formalized multidisciplinary teams
- More “rigid” (?)
- Fewer social services
- Less “patient-centered”, more “disease-centered”
- Reimbursement and coverage challenges

CCHS specific challenges in adult care

- Very few adult CCHS patients
- Specialists who know CCHS are not widely available
- Lack of familiarity outside your specialist team
- Sleep labs may not accommodate
- Few institutions doing diaphragm pacemakers
- Variable needs
- Natural history of disease in adult is uncertain
- Insurance coverage for home nursing and monitors

Converging with the changes that come with adulthood

- Going out into the world – jobs, hobbies, friends
- Managing finances and schedules
- Spouses/roommates stepping in, parents stepping back
- Readiness for independence and autonomy

Transitions of care (Adult perspective)

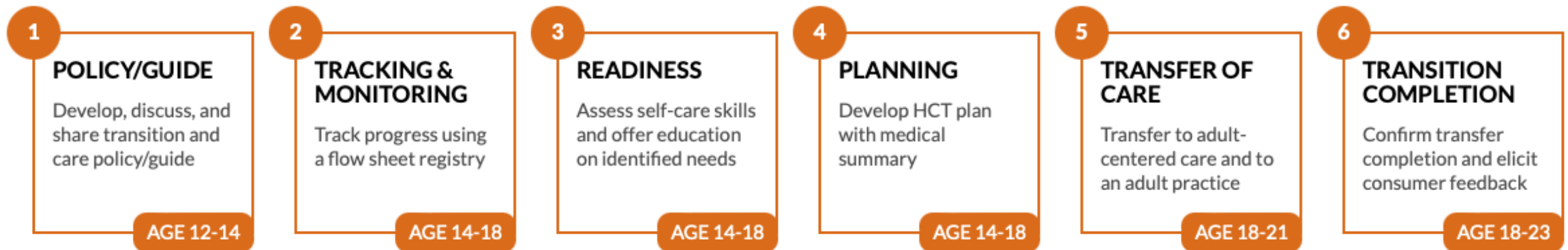
- Definition: ‘purposeful, planned, multidisciplinary movement of adolescents and young adults from child-versus-adult-oriented health care systems’
- Focus on children with “special healthcare needs”
- Has been best implemented for patients with cystic fibrosis
- Stressful time
- Vulnerable time – e.g. medication adherence drops
- CCHS patients often have multispecialty needs -> more complex transition

Pediatric to Adult Transition of Patients with Neuromuscular Disease in the upcoming Springer title “***Pulmonary Complications of Neuromuscular Disease***” – R. Amin, J. Chiang, K. Selby, J. Orr

- Blum RW, Garell D, Hodgman CH, et al. Transition from child-centered to adult health-care systems for adolescents with chronic conditions. A position paper of the Society for Adolescent Medicine. J Adolesc Health 1993;14(7):570-6. DOI: 10.1016/1054-139x(93)90143-d.

“Got Transition” is a federally funded resource center on health care transition

SIX CORE ELEMENTS™ APPROACH AND TIMELINE FOR YOUTH TRANSITIONING FROM PEDIATRIC TO ADULT HEALTH CARE



www.gottransition.org

Strategies for successful transition to adult health systems

- Early initiation: recognize that this is a multi-year process
- Consider use of readiness checklist
- Individualize
- Summary document – easier if shared electronic health record
- Speak up and advocate!

“Got Transition” has valuable resources



Sample Transition Readiness Assessment for Youth

Please fill out this form to help us see what you already know about your health, how to use health care, and the areas you want to learn more about. If you need help with this form, please ask your parent/caregiver or doctor.

Preferred name _____ Legal name _____ Date of birth _____ Today's date _____

TRANSITION IMPORTANCE & CONFIDENCE *Please circle the number that best describes how you feel now.*

The transfer to adult health care usually takes place between the ages of 18 and 22.

How important is it to you to move to a doctor who cares for adults before age 22?

0 _____ | 1 _____ | 2 _____ | 3 _____ | 4 _____ | 5 _____ | 6 _____ | 7 _____ | 8 _____ | 9 _____ | 10 _____
not _____ very

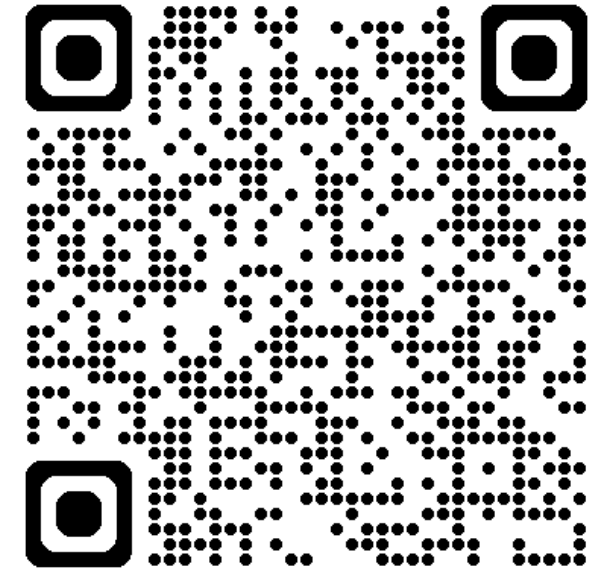
How confident do you feel about your ability to move to a doctor who cares for adults before age 22?

0 _____ | 1 _____ | 2 _____ | 3 _____ | 4 _____ | 5 _____ | 6 _____ | 7 _____ | 8 _____ | 9 _____ | 10 _____
not _____ very

MY HEALTH & HEALTH CARE *Please check the answer that best applies now.*

	NO	I WANT TO LEARN	YES
I can explain my health needs to others.	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
I know how to ask questions when I do not understand what my doctor says.	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
I know my allergies to medicines.	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
I know my family medical history.	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>

A FAMILY TOOLKIT: PEDIATRIC-TO-ADULT HEALTH CARE TRANSITION



How to find (or build) a CCHS adult team

- It might already exist: Ask pediatric providers and CCHS families/networks
- Find the “quarterback” – usually will be a Pulmonologist
 - Look for Assisted/Home Ventilation Programs at Academic Medical centers
- Includes a respiratory therapist (they know the most about equipment!)
 - Sometimes available in office, otherwise by DME
- Utilize resources available via insurance and other agencies
- Help your team help you

Ongoing care as an adult

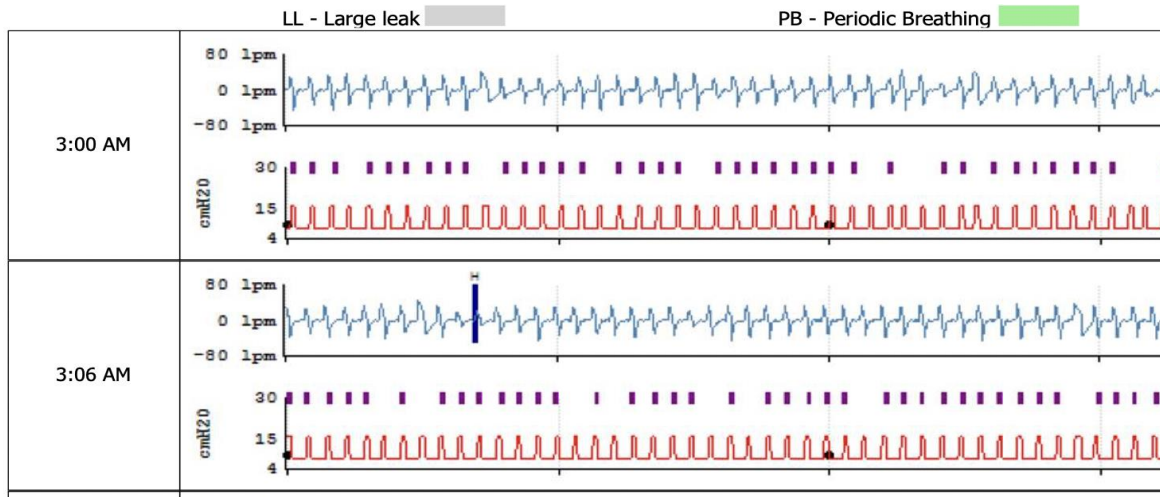
- Written copy of care plan
- Medical bracelet
- Power of attorney (default surrogate is legally defined by each state)
- Regular follow up – every 3-6 months
- Proactive about monitoring plan
- Report sentinel events
- Plan ahead for surgeries
- Stay involved
- Advocate and speak up!

What does the adult healthcare world need to do?

- Partner with pediatric CCHS programs
- Education of trainees (pulmonary and sleep fellows)
- Raise the profile via professional organizations
 - Symposia, practice guidelines, workshops
- Develop new strategies for monitoring
- Broader change to coverage
 - Outpatient respiratory care
 - Care coordination

Remote patient monitoring is an emerging tool for long-term care

- Monitor ongoing use, leak, ventilatory parameters, etc in near-real time



Therapy Report

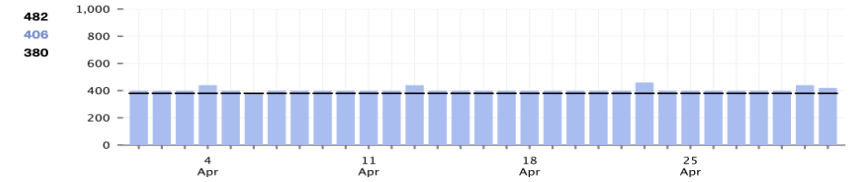
AirCurve 10 ST-A

SN: 22171081710

Tidal Volume (ml)

Maximum (avg)
95th % (avg)
Median (avg)

482
406
380



Respiratory Rate (breaths/min)

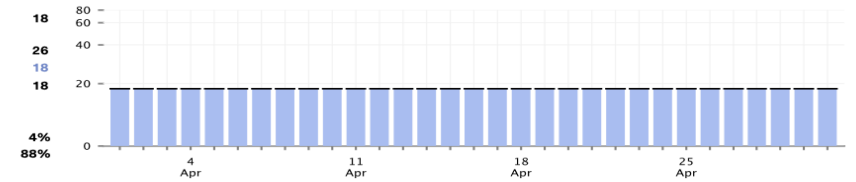
Target patient rate

Maximum (avg)
95th % (avg)
Median (avg)

18
26
18
18

Spontaneous breaths
Triggered
Cycled

4%
88%



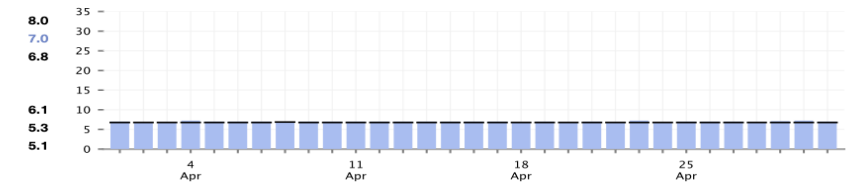
Minute Ventilation (L/min)

Maximum (avg)
95th % (avg)
Median (avg)

8.0
7.0
6.8

Alveolar Ventilation
Maximum (avg)
95th % (avg)
Median (avg)

6.1
5.3
5.1

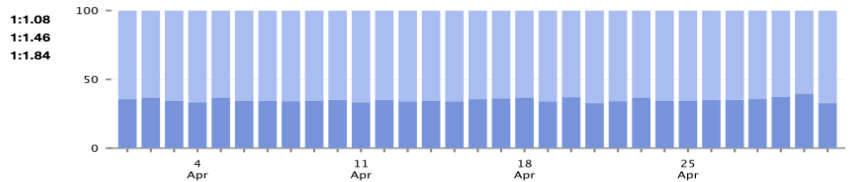


I:E Ratio (median in %)

Maximum (avg)
95th % (avg)
Median (avg)

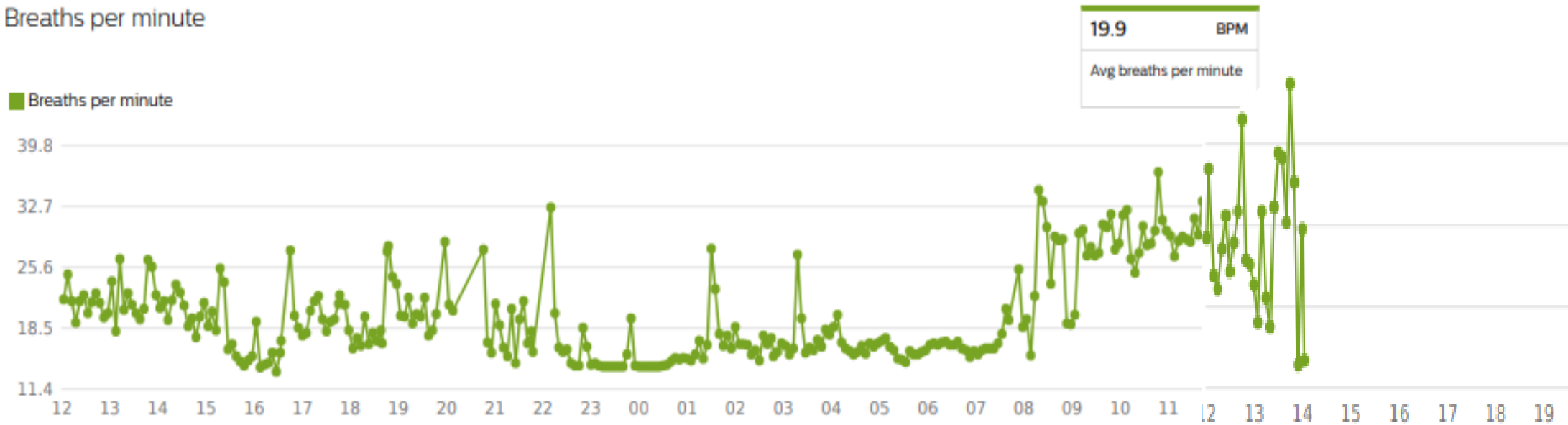
1:1.08
1:1.46
1:1.84

E%
I%

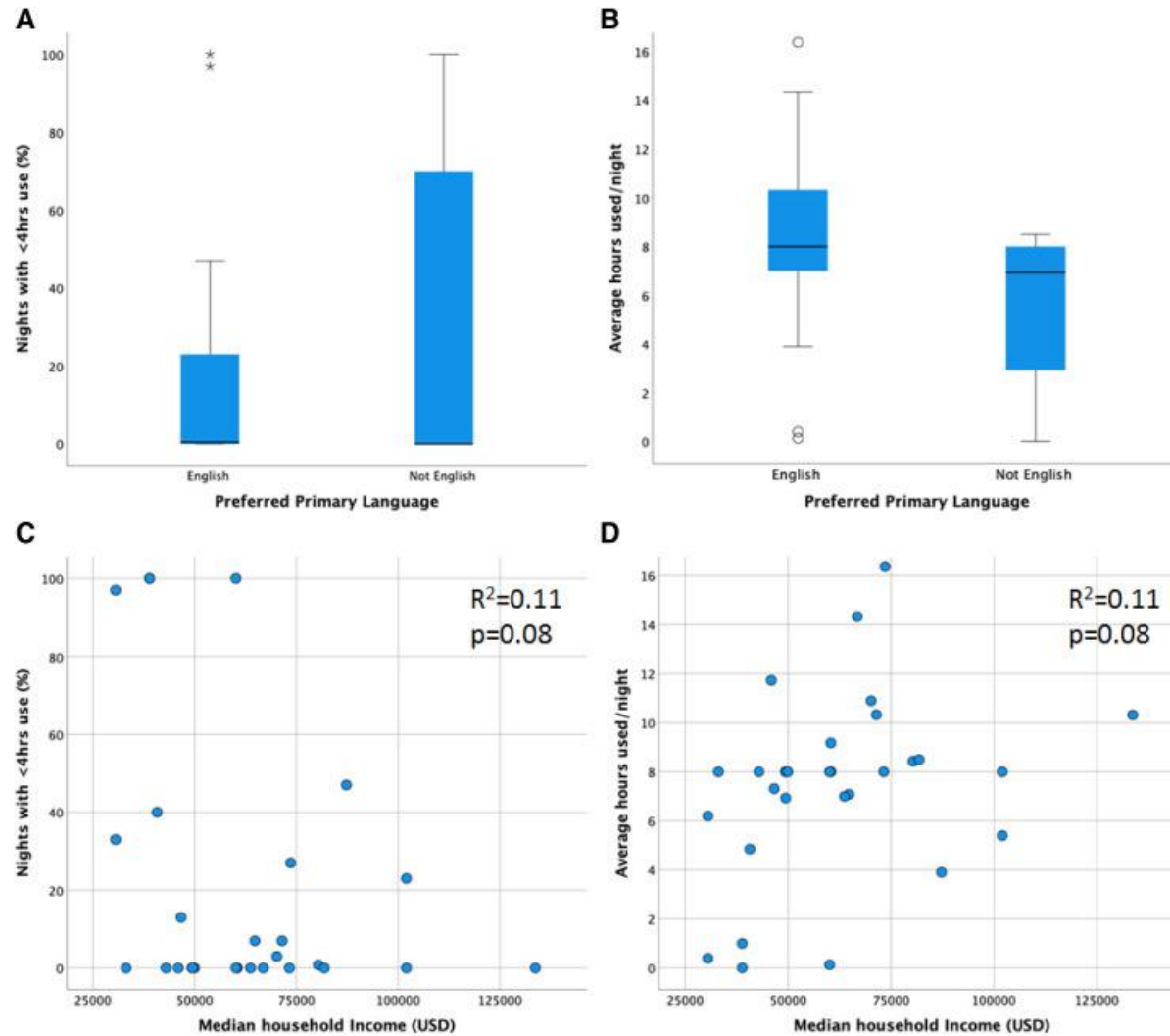


Remote monitoring technology might help detect issues

Breaths per minute



Social determinants of health are important



Independent predictors of NIV use:

- Lower lung function
- More ventilator support
- Higher estimated household income

Hurvitz MS, Bhattacharjee R, Lesser DJ, Skalsky AJ, Orr JE. Determinants of usage and nonadherence to noninvasive ventilation in children and adults with Duchenne muscular dystrophy. *J Clin Sleep Med*. 2021 Oct 1;17(10):1973-1980.

Thank you

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