Social Security Disability in the Congenital Heart Community: What are the Issues?
National Congenital Heart Coalition
Scope of Presentation

- Overarching Issues
- Issues in Pediatric Disability
- Issues in Adult Disability
- Summary of Recommendations
Overarching Problem

- Many CHD adults and families of children report
  - Severe problems qualifying for disability
  - Lack of knowledge of CHD by SSI staff/authorities
  - Denials despite disabling CHD
  - Burden of “re-proving” permanent conditions
NCHC Definition of CHD

- In-born problem with **structure** of the heart
  - Parts too big, too small, malformed, missing, in wrong place

- **Not** : In-born SCD syndromes (long QT, HCM)
- **Not** : Acquired (pediatric or adult)
CHD = #1 U.S. Birth Defect

- ~ 1 in 100 – 125 births
- ~ 1 born every 15 minutes
- ~ 40,000 born each year
- Born with 1 of 30+ possible CHDs
  - Many disorders in “one basket”
  - 7 more common lesions, 25+ rare ones
The CHD Pinwheel

*Other includes 20+ diagnoses with incidence < 5%
*Single Ventricle combines 6+ lesions with one ventricle (L, R, combo)
How many surgeries?

Number of Surgeries (CONCOR)

- 53% for one surgery
- 25% for two surgeries
- 22% for three or more surgeries

Mean age: 34
CONCOR Registry Data
Patients Reaching Age 18 with CHD

# CHD Patients 18+

- 1970: 325,000
- 1980: 500,000
- 1990: 750,000
- 2000: 1,000,000
- 2010: 1,300,000
Highly Complex CHD Population: Province of Quebec

- Single Ventricle
- Pulmonary Atresia
- Transposition Complexes
- Eisenmenger syndrome
- Cyanotic CHD

New CHD Landscape

- ~2 million CHD patients
- ~90% now reach 18
- More adults than children
- Population growing by ~ 5% yr
  - Due to increased survival not incidence
- Population growing more complex
  - Ex: First HLHS surgery mid-1980’s
  - First single, right ventricle pts now reaching late childhood/early adulthood
Diagnoses ≠ Outcomes

- Outcomes vary widely within diagnoses
- Factors affecting outcomes include:
  - Unique defect anatomy
  - Treatment(s)
  - Co-morbidities
  - Timing of interventions
  - Luck
New Anatomies need new standards

- Mismatch of current required assessment data and CHD physiology
- Ex: LV measure; LV EF often required
  - Many CHD patients have
    - Systemic right ventricle
    - Single ventricle
    - RV issues
- Standard EF/LV measures irrelevant
- Data relevant to anatomy not requested
NCHC Recommendation #1

Basic education for SSI staff on complex CHD as new chronic/disabling condition; CHD as structural problem, entirely unlike/unrelated to acquired cardiac problems
NCHC Recommendation #2

Any anatomic assessment in CHD should be anatomically appropriate; CHD patients should NOT be expected to submit irrelevant/inappropriate standard anatomic measures.
NCHC Recommendation #3

Disability determination should not assume correlation between diagnosis and disability, when possible should use functional measures, not diagnosis in isolation
CHD Categorization System

- 2000 - Bethesda 32\textsuperscript{nd} defined
- 2008 – Used in ACC/AHA ACHD Guidelines
- 3 Categories based on complexity
- Broadly correlates with impact/disability
Category 1: Simple CHD

Ex:
- PDA
- Simple/repaired ASD
- Small/repaired VSD
- ~50% of population

Least likely to be disabled
Category 2: Moderately Complex CHD

Ex:
- Coarctation
- Tetralogy of Fallot
- Pulmonary stenosis
- ~30 - 35% of population

More likely to be disabled
Category 3: Highly Complex CHD

Ex:
- Transposition disorders
- Single Ventricle disorders
- Palliated disorders
- Eisenmenger Syndrome
- ~15-20% of population

Most likely to be disabled
NCHC Recommendation #4

Disability determination in CHD should use existing CHD categorization system to help inform decisions.
Neurocognitive Issues

- 13% CHD pts syndromic
  - Major intellectual impairment common
- Many syndromes assoc. w/CHD
  - Downs (50%), 22Q, Noonans, Turners
- Disability criteria well-established for syndromes/major intellectual disability
- CHD pts w/syndromes eligible via syndrome
Neurocognitive Issues

- More subtle cognitive issues common
  - Up to 1/3 show learning issues, ADD, lower IQ
  - Difficult to measure in infant/young child
  - Become more visible in school-age children/adults

- Medical issues can impact mental health
  - Anxiety, depression, PTSD common in adults

- Likely to impact higher ed, employment
NCHC Recommendation #5

Disability assessment in CHD should include age-appropriate screening for associated neurocognitive and mental health issues.
Pediatric Disability Issues

- Majority of children now receive early detection and early intervention
- Significant # will still experience childhood disability
  - Permanent disability
  - Transient disability
    - Event-related - Many children with mod/highly complex CHD require childhood surgical intervention
- Many improve, may no longer meet adult guidelines for disability
Highly Complex CHD in Childhood

Almost all start out “blue” (cyanotic), need OHS (1 – 3 or more)

Two main groups (may overlap)
  - Transposition disorders –
    - Typically 1 OHS, less childhood impact
  - Single Ventricle Disorders
    - 2 + surgeries, more childhood impact
On-going Cyanosis

- Many CHD pts have pre-repair cyanosis
- Repair now corrects cyanosis in >90% pts
- Small % of children/greater % of adults have continued low O2 on effort or at rest

Permanent low O2 = permanent disability
NCHC Recommendation #6

CHD children and adults with documented low O2 at rest or with effort should be considered disabled.

Documentation should NOT require arterial stick as this is dangerous, painful, and unnecessarily invasive.
Single Ventricle in Childhood

- Highest level of mortality, morbidity, disability
- Complex multi-step infant/child surgeries
  - Typically Fontan procedure
  - Many additional problems post-Fontan
- 2 – 3 staged OHS
- Continued cardiac disability likely until surgeries complete
- Long/recurrent hospitalizations likely
- Continued complications likely
Single Ventricle in Childhood

- Common impacts on child during staging
  - Continued low O2 state
  - Developmental delay
  - Missed school
    - Intellectual, Social impact
  - Exercise capacity abnormal
  - Significant functional limitations
Single Ventricle in Childhood

- Common impact on Family
  - High medical costs
  - Impact employment
    - Lost work days
  - Impact on other children
  - High need for social, economic support
Single Ventricle in Childhood

- **Post-surgeries**
  - Good news: Many have near-normal childhood
  - Bad News - Palliative, not reparative
    - One ventricle system
    - Heart/lung system highly abnormal
      - Ex: loss of normal exercise response
    - Impacts other body systems
      - Gastronintestinal impact, PLE
      - Abnormal clotting, High risk of emboli/stroke
All CHD children and adults with documented single ventricle anatomy should be considered disabled
Mod. Complex CHD in Childhood

Ex: Coarctation, Tetralogy of Fallot, Pulmonary stenosis

- Most need infant/childhood intervention
  - OHS or cath-based, One or multiple
    - Bypass, multiple OHS raise risks to function
- Many have pre/peri/postoperative disability
- Many will have event-related disability
- Often stabilize/improve post-intervention
Mod. Complex CHD in Childhood

Issues post-intervention

- All need regular cardiac follow-up
  - Moderate-high intensity
- School absence common due to
  - Medical follow-up
    - Residue/sequelae ex: Rhythm issues
  - Medication dosing frequency and side-effects
  - Reintervention(s) may be needed
Mod. Complex CHD in Childhood

- Typically does not inhibit “normal” childhood activities
  - Minor functional limits Ex: no weight lifting
  - Normal/near-normal exercise capacity
    - Measurement is observation based in infancy/early childhood, Objective testing not feasible
    - Later childhood: V02 screen possible
- Most CHD patients show mild-moderate impairment
  - Even well-repaired, reporting no limits
    - Child “norms” to function
Mod. Complex CHD in Childhood

Significant minority of children:
- Less-successful repair
- Co-morbidities
- More residue/sequelae
- More re-intervention
- On-going cardiac disability
  - Can impact school, play, social development
- Significant impact on families
Simple CHD in Childhood

Ex: PDA, Simple ASD, small VSD

- May/may not need intervention
- Intervention typically uncomplicated
  - Cath-based or OHS
- Low-level cardiac follow-up needed
- Least likely to experience disability
  - **Exception**: Undiagnosed/Unrepaired patients
    - Prevalent in immigrant population
Suggestions for Assessment

• Allow appropriate time for stabilization following intervention/surgery(ies)
• Assessment might include
  • Appropriate anatomic assessments
  • Functional factors such as
    • loss of school days
    • #/frequency of hospitalizations
    • VO2 assessment in older children/teens/adults
    • Age appropriate neuro-development measures
NCHC Recommendation #8

Disability assessment in CHD population should include age-appropriate functional measures and better accommodate more subtle impairments and impact on schooling/work
Disability and Transition Issues

- Many CHD children currently lose SSI @ 18
  - Change from ped – adult SSI standard
  - Teen found to be employable
- Families often surprised/unprepared
  - Expectation: childhood SSI= adult SSI
- No previous planning for
  - Independence/employment
  - Insurance transition
- Teen
  - May lose health insurance
  - May be unprepared for work
NCHC Recommendation # 9

Educate patients and families about disparity between pediatric and adult disability guidelines to help with transition between two systems.
Adult Disability Issues

- Many issues same for adults/children
  - Pre/peri/post operative issues
  - Need for functional measures
  - Need for appropriate tests/anatomic measures
Adult Disability Issues

What’s Different?
- Morbidity/disability rises sharply
  - Even in those with no childhood problems
- Health issues grow more complex
  - Multiple re-operations, new co-morbidities
- Large majority of adults not in recommended care
  - Causes challenges in assessment
Age at Death for Adults with CHD

Oechslin EN et al AJC 2000.

- Tricuspid Atresia: Mean age 27 ± 5 years
- TGV: Mean age 27 ± 7 years
- COA (Coarctation of the Aorta): Mean age 29 ± 6 years

- Sudden death: 26%
- CHF: 21%
- Perioperative: 18%

n = 2609 patients
199 died
Mean age for all dx 37 ± 15 years
Adults with Congenital Heart Disease

Arrhythmias
- Atrial
- Ventricular
- Sudden Death

Heart Failure
- Right Heart Failure
- Left Heart Failure
  - Systolic
  - Diastolic
  - Pulmonary Hypertension

Vascular Lesions

Valvular Disease

Residual Shunts
Adult Disability Issues

- Adults with CHD get “typical” cardiac symptoms
  - Rhythm problems
  - Heart Failure
  - TIA/Stroke
- But
  - Atypical causes
  - Atypical presentation
  - Atypical impact
  - Atypical treatments
Rates of Rhythm Problems in ACHD (Supraventricular)

- TOF: 30%
- Eisenmenger's: 36%
- TGA: 48%
- Post-fontan: 49%
Rhythm Problems in CHD

- Arrhythmias most common problem in ACHD
  - Rates rise with age - >50% in some lesions
- May be caused by hemodynamic change
  - Reintervention may be indicated
- High rates of recurrence
  - Ablation, medication not as effective as in “regular” populations
  - New abnormal pathways often develop
- “Minor” rhythm problems can have major impact on
  - Function: ex:marginal ventricular function, lost preload
  - Ability to work
Heart Failure in CHD

- Common in adults
  - Complex CHD comes with ventricular weakness/vulnerability
  - Some consider all complex CHD = stage 1 failure
  - Often right-sided issues, and/or systemic RV issues
    - “standard” symptoms may be absent
  - Complex interaction with anatomy/rhythm
  - Functional challenges due to CHF rise with age
    - Fatigue
    - Decreased exercise capacity
    - ER, hospital, doc visits, meds, devices
Adult Disability Issues

- Adults with CHD show 2 – 3X higher health care usage such as
  - Doctor visits
  - Hospitalizations
  - ICU usage
  - ER visits
- Tracks with disease complexity
Age Range of Patients with CHD at Hospital Admission

Year of Admission

Number of Admissions

Age 12-19

Age 20-29

Age > 30

Warnes & Liberthson. JACC. 2001; 37: 1173.
## Health Service Utilization of 22,096 ACHD Patients, 1996 to 2000 inclusive

<table>
<thead>
<tr>
<th>Health Care Service</th>
<th>n* (%)</th>
<th>Median† (IQR)</th>
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<tbody>
<tr>
<td>General practitioner outpatient visits</td>
<td>20,131 (91.1)</td>
<td>15 visits (7-27)</td>
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<tr>
<td>Cardiologist outpatient visits</td>
<td>12,113 (54.8)</td>
<td>4 visits (2-8)</td>
</tr>
<tr>
<td>Specialist‡ outpatient visits</td>
<td>19,276 (87.2)</td>
<td>10 visits (4-22)</td>
</tr>
<tr>
<td>Emergency department visits</td>
<td>14,994 (67.9)</td>
<td>3 visits (1-6)</td>
</tr>
<tr>
<td>Hospitalization</td>
<td>11,332 (51.3)</td>
<td>9 days (4-26)</td>
</tr>
<tr>
<td>Critical care</td>
<td>3,536 (16.0)</td>
<td>5 days (3-10)</td>
</tr>
</tbody>
</table>

*Number of adults who used the service at least once from 1996 to 2000 inclusive.
†Median number of visits or inpatient days among users.

One-year hospitalization rate of patients with severe and other cardiac lesions compared with the adult population of Quebec (April 4, 1999 to March 31, 2000)

- Severe CHD: 354 cases per 1000 persons, p<0.0001 compared to the Quebec population
- Other CHD: 208 cases per 1000 persons, p<0.0001 compared to the Quebec population

Highly Complex CHD in Adults

- CHD = Multi-system disorder
  - Ex: Eisenmenger Syndrome, Liver/kidney impact, PLE
- Unexpected problems
  - “Premature aging” described
  - ? Related to autoimmune stress/treatment impact?
Mod Complex CHD in Adults

- New complications common
- Ex:
  - TOF – High rates of valve replacement, right-sided failure
  - Coarctation – re-stenosis, aneurysms
Simple CHD in Adults

- Repaired ASD/VSD can cause new problems, need Re-operation(s)
- Significant % of adults misdiagnosed as “simple”- early technology limited
- Unrepaired/undiagnosed ASD, VSD, PFO can cause major problems
Adult Disability Issues

- ~ 10% of CHD found in adulthood
  - Some simple – ex: PFO
  - Some complex - CCTGA
  - Severe symptoms can lead to diagnosis
  - OHS may be needed
  - PH, stroke, CHF = disability may occur
NCHC Recommendation #10

- Disability assessment in ACHD should accommodate for
  - greater impact of common disorders on abnormal anatomy
  - Impact of high rates of ER, hospitalizations, healthcare usage
  - All associated/related non-cardiac disorders
“Loss to Care” Challenge

- Majority of CHD patients drop from cardiac care in childhood
  - Feel well, perceive self as cured
  - Quebec study: Majority not in cardiac care by age 12
- Not in recommended ACHD care as adults
- Recommended for 50% of ACHD patients with mod/highly complex CHD, <10% receive it
Canada: Loss to Follow-up in CHD

- **< 6 years:**
  - Diagnosed by cardiologist: 643 (100%)
  - Seen by cardiologist: 413 (64%)
  - Attrition: 53 (8%)

- **6-12 years:**
  - Diagnosed by cardiologist: 643 (100%)
  - Attrition: 177 (28%)

- **13-17 years:**
  - Diagnosed by cardiologist: 466 (72%)
  - Seen by cardiologist: 292 (45%)
  - Attrition: 51 (8%)

- **18-22 years:**
  - Diagnosed by cardiologist: 343 (53%)
  - Seen by cardiologist: 249 (39%)
  - Attrition: 94 (15%)

ACHD Patients in USA vs Those in ACHD Clinics

Number Of Patients

- 787,000
- <50000 in ACHD


ACHA Clinic Directory Working Group 2007
Adult Disability Issues

- “Regular” adult cardiologists typically untrained in assessment, treatment of complex CHD
  - Highly specialized testing needed
- Testing likely to be inaccurate/irrelevant
  - Ex: Standard CAD tests done to assess anatomy, Systemic RV misdiagnosed/mismeasured
Adult Disability Issues

Many ACHD programs report that high % of new ACHD patients labeled “permanently disabled” improve with appropriate care/reintervention

Requiring ACHD center assessment would help get thousands of ACHD patients back to appropriate care and possible return to health
NCHC Recommendation #11

- Long-term goal:
  All mod/highly complex ACHD patients requesting disability should be referred to ACHD programs for assessment/review

- Difficult to achieve in current system but
  - Will save lives in the short run
  - Will save $$$ in long run
NCHC Recommendation #12

- Short term:
  Functional measures such as VO2 max exercise testing, medications, medical burden should be used rather than anatomic measures in complex CHD in adults to accommodate for poor quality of information likely to be submitted by non-ACHD centers
Summary: Challenges

1: CHD is complex
2: CHD survivors are a new population
3: Diagnosis ≠ Outcomes
4: New anatomies need new measures
5: Most ACHD patients “lost” to CHD care
Summary: Recommendations

1: Educate SSI staff on CHD
2: Only **Appropriate** anatomic measures
3: Assess function, not just diagnosis
4: Use complexity categories to guide
5: Assess neurocognitive/mental health
6: Single ventricle = disability
Summary: Recommendations

7: Cyanosis = disability
8: Look at “whole picture” of impact
9: Educate families for SSI transition
10: ACHD = > impact from common problems
11: ACHD programs review ACHD patients
12: Plan for poor ACHD data from “regular” cardiac care