Childhood Conditions All Grown Up – What Adult Hospitalists Need to Know

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Objectives

- Identify common conditions of childhood that carry into adulthood with case scenarios
- Recognize the appropriate approach to a specific clinical scenario in a hospitalized adult with a chronic childhood condition
- Characterize and share barriers to transition of care for patients with chronic childhood conditions

Clinical Case #1

- A 24 year old male with a past significant medical history of Trisomy 21 (Down's Syndrome) is presenting with a caregiver for a 7 day history of shortness of breath. Within the past 36 hours, intermittent low-grade fevers to 38.4 have been noted.
- Chronically on levothyroxine for hypothyroidism, but no other known medical issues. Works as a Volunteer 3 times per week.

Increasing Survival to Adulthood

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Survival Factoid</th>
</tr>
</thead>
<tbody>
<tr>
<td>Childhood cancers</td>
<td>46% of survivors are 20 to 40 years old; 18% of survivors are over age 40</td>
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<tr>
<td>Spina bifida</td>
<td>90% reach adulthood</td>
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<tr>
<td>Congenital heart disease</td>
<td>85% reach adulthood; adults with CHD&gt;1,000,000</td>
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<tr>
<td>Cystic fibrosis</td>
<td>37 years old; 50% are over age 18</td>
</tr>
<tr>
<td>Down syndrome</td>
<td>55-year life expectancy</td>
</tr>
<tr>
<td>Sickle cell disease</td>
<td>55-year life expectancy</td>
</tr>
<tr>
<td>Hemophilia</td>
<td>60-year life expectancy</td>
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</table>

Source: Chang W. Transitioning the Chronically Ill Child to Adult Care. Aug. 26, 2011
Clinical Case # 1

- On physical exam, vitals: 37.9 BP:105/67
  HR: 99
  89% O2 sat RA.
- Gen: Appears clinically dehydrated and weak
- Lungs: bilateral crackles with mild rhonchi
- CVS: tachycardia
- SKIN: decreased skin turgor

Down Syndrome – Trisomy 21

- Background
  - Most prevalent genetic disorder
  - 1:1000 births
  - Increased survival noted – mid-50s
- Outpatient Management
  - Multi-disciplinary approach
  - Similar health screening guidelines and immunizations

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**TABLE 1**

<table>
<thead>
<tr>
<th>Medical Conditions</th>
<th>Clinical Signs</th>
<th>Cardiac Disease</th>
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<tbody>
<tr>
<td>Endocrine</td>
<td>Cardiac failure</td>
<td>Mitral Regurgitation</td>
</tr>
<tr>
<td>Thyroid disease—hyperthyroidism</td>
<td>Abnormal sinus rhythm</td>
<td>Mitral Valve Prolapse</td>
</tr>
<tr>
<td>Diabetes mellitus</td>
<td>Angina pectoris</td>
<td>Clinical Signs</td>
</tr>
<tr>
<td>Mental health</td>
<td>Syncope</td>
<td>Echocardiography</td>
</tr>
<tr>
<td>Depression</td>
<td>Syncope</td>
<td>Valve repair?</td>
</tr>
<tr>
<td>Obsessive-compulsive disorder</td>
<td>Syncope</td>
<td></td>
</tr>
<tr>
<td>Abuse (physical or sexual)</td>
<td>Syncope</td>
<td></td>
</tr>
<tr>
<td>Conduct disorder</td>
<td>Syncope</td>
<td></td>
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</tbody>
</table>

*—Listed in approximate order of clinical importance.

**Down Syndrome – Trisomy 21**

- **Pulmonary Disease**
  - Increased risk of pneumonia and bronchiolitis
  - Leading hospitalization cause of Trisomy 21 patients
- **Obstructive Sleep Apnea**
  - Hypercapnea
  - Higher risk for mechanical ventilation

- **Musculoskeletal**
  - Atlantoaxial Instability
  - Clinical Signs
    - Gait abnormality
    - Limited Range of Motion
    - Paresthesias, Sensory Deficits
    - Hyperreflexia
    - Clonus
  - Implication of Hospitalized Patient

**Clinical Case #2**

- 19 year old female with past significant medical history of Sickle Cell Disease (Hb SS) admitted for a pain crisis secondary to bilateral leg pain and abdominal pain.
- She has not seen a Physician for 1 year. LFTs noted to be elevated, and found to have cholecystitis.
- On hospital day 2, pain is better controlled on a PCA, and she is being taken for Surgery for a Laparoscopic Cholecystectomy.
Clinical Case #2

- Pre-op, her Hgb was 8.0 and O2 sat was 90% with decreased air movement at bases thought to be secondary to atelectasis.
- Patient tolerated the procedure well, and has returned to the floor. Two hours later, you are informed that patient is acutely short of breath and has O2 sat on 3 L of 85%.

Sickle Cell Disease

- Age expectancy: 55 years
- More than just PAIN
- Limitations at transition to adult care
  - Lack of resources
  - Familiarity of Pediatric environment
  - Fear of new systems
  - Health insurance (after age 18)
  - At-risk age group

Newborn

- 5 years
- 15-18 years
- 20-60 years
- 40-60 years

- No symptoms
- Anemia
- Splenic Sequestration
- Splenic Involution
- Dactylitis (Hand-Foot Syndrome)
- Hepatobiliary Disease
- Acute Chest Syndrome
- Stroke
- Priapism
- Avascular Necrosis
- Pulmonary Hypertension

Sickle Cell Disease

- Acute Chest Syndrome
  - Most common cause of death from SCD
  - Clinical Findings
    - Fever
    - Chest Pain
    - Clinical Respiratory Decompensation
  - Multiple Triggers
  - Treat with supportive care
    - Possible exchange transfusion
  - HIGH mortality in Adults vs Children
Sickle Cell Disease

- **Acute Chest Syndrome**
  - Focus on Prevention
  - Supplemental Oxygen
  - Adequate hydration
  - Bronchodilators as needed
  - Analgesia – avoid respiratory splinting
  - Peri-operative Strategies
    - Avoid pre-op infection and assure baseline state of health
    - Supplemental oxygen & Incentive Spirometer
    - Be aware of significant intra-operative events
    - Analgesia
    - Preoperative transfusion?

- **Neurologic Complications**
  - Stroke (Pediatric vs Adult)

Clinical Case #3

- A 20 year old female with a past significant medical history of Anorexia Nervosa is brought to the hospital by her family members after she has had a relapse of her disease and lost over 20 lbs. in the past 3 months. Over the past week, she has had very little oral intake. Family is concerned as she is dizzy when she stands up and very weak.
- She has had multiple admissions as an adolescent at the local Children’s Hospital for complications related to Anorexia Nervosa, and was started on amitriptyline as outpatient 6 months ago for Depression.
Clinical Case #3

- On Physical exam, vitals are BP 95/45 (laying down) HR 52   RR 16   O2 Sat 95% RA. BMI 14
- Patient appears emaciated, disengaged, has temporal wasting, and does not allow a complete physical exam and refuses orthostatic vitals.
- Labs show a K+ of 3.1.

Eating Disorders

- Anorexia Nervosa
- Bulimia nervosa
- Eating Disorder NOS
- Binge-eating Disorder

Eating Disorders

- Multi-disciplinary outpatient management
- Criteria for Hospitalization
  — Hypothermia (T < 36 C)
  — Bradycardia (HR < 50 daytime or < 40 nighttime)
  — Hypotension
  — Orthostatic Changes
  — Arrhythmia (including prolonged QTC)
  — Electrolyte Abnormalities
  — Behavioral Changes

Eating Disorders

- Approach to Inpatient Management
  — Stabilize Patient
    - Hemodynamics
    - Electrolytes
    - Co-manage with Nutrition, Psychology, and/or Psychiatry
  — Nutritional Strategy
    - Maximize weight gain and minimize refeeding syndrome risk
    - Hypometabolic \(\rightarrow\) Hypermetabolic
Eating Disorders

- Starvation / Undernourished State – dependent on fat
  - Glucose as source of energy (shift from Fat)
    - High glucose levels → lead to acute rise in insulin secretion
      - Rapid Glucose Metabolism
      - Hypokalemia
      - Increased need for Phosphorus for cellular metabolism (glycolysis intermediates)
      - Hypophosphatemia
      - Other Electrolyte Abnormalities
      - Cardiac and Respiratory Complications

Eating Disorders

- Strategies to inpatient feeding
  - Goal to start calories at 1500 to 2000 kcal / day
  - Goal of weight gain of 0.2 kg/day
  - Electrolytes monitoring daily
  - Phosphorus Supplementation
  - Avoid vigorous IV Fluids
  - Telemetry

Clinical Case #4

- 24 year old female with a past significant medical history of Cystic Fibrosis recently has moved to the area, and presents to the ER with increased shortness of breath, fatigue, low-grade fevers.
- She has not set up with a Primary Care Physician or Pulmonologist (CF Center) in her area.
- Aside from these symptoms, she is constantly thirsty and feels as if she has lost weight over the past 2 months, but attributes it to stress related to the move.

Clinical Case #4

- On physical, BP 100/70 HR 109 RR 23 T 38.2 BMI 17 O2 sat 87% on RA
- Gen: Patient is awake, appears chronically ill
- HEENT: +temporal wasting
- CVS: +tachycardia; no m/r/g
- RESP: coarse breath sounds with rhonchi; mild intercostal retractions
- ABD: Soft; NT/ND; BS+
- EXT: no edema
Cystic Fibrosis

- Most common, life-shortening disease in Caucasians.
- Approximately 1000 new cases per year
- Improved survivability
  - Average survival age of 37 years of age
  - Nearly half are above age 18

Cystic Fibrosis

- Autosomal Recessive
- Pathophysiology
  - CFTR gene mutation
  - Defective transport of Sodium and Chloride
  - Results in Defective mucociliary clearance
  - Thick Secretions – difficult to clear from airways
  - Persistent infection

Cystic Fibrosis

- Pulmonary Exacerbations
  - Clinical Features
    - Cough
    - Increased sputum production
    - Dyspnea
    - Loss of weight
    - Lung Function Decline (monitored frequently in CF patients)
  - Other comorbidities requiring inpatient attention
    - Nutritional Needs
    - Glycemic Control
Cystic Fibrosis

- Pulmonary Exacerbations – management
  - Co-management with Pulmonary (CF Center?)
  - Continue all chronic therapy (inhaled medications and respiratory medications for lung health)
  - Airway clearance therapy
  - Antibiotics
    - *Pseudomonas aeruginosa*
    - Use aminoglycoside + Beta-lactam
    - Avoid PO antibiotics
    - Duration based on patient
    - Steroids?

Cystic Fibrosis

- Cystic Fibrosis-Related Diabetes (CFRD)
  - Increased prevalence post-adolescence
  - Unique disease process
    - Overlap of DM-Type 1 and DM-Type 2
  - Glycemic control as inpatient
    - Reduction in complications and mortality
    - Opportunities for education

Cystic Fibrosis

- Airway Clearance Therapy

Cystic Fibrosis

- Cystic Fibrosis-Related Diabetes (CFRD)
  - Screening
    - A1C not recommended for screening
    - Oral glucose tolerance test (OGTT)
      - Perform 6 weeks outside of an exacerbation
      - Cannot be performed during acute hospitalization
      - Start screening by age 10
Cystic Fibrosis

- CFRD Management in the hospital
  - Fasting hyperglycemia → Add basal insulin
  - Normal fasting blood glucose → Prandial insulin
    - Use a “Carbohydrate Counting” Method + correctional insulin
  - Titrate daily
  - Challenges in inpatient glycemic control
    - Variability in meals and consumption (erratic patterns)
- Discharge Treatment
  - Use insulin. Avoid oral medications.
  - Frequent monitoring
  - Can use A1C for monitoring (but NOT screening yet in CF)

- Hypoglycemia
  - Common in CF and CFRD patients
  - Treat as any other patient
  - Counsel and educate

Clinical Case #5

- A 32 year old female with a past significant medical history of Cerebral Palsy and Mental Retardation is brought to the ER by her Mother due to significant abdominal distention and generalized agitation.
- She has not had a hospitalization since adolescence when she was admitted for Pneumonia. She is fed exclusively through her PEG. Her mother is very well-versed on her care, and follows annually with a Neurologist and PCP as needed.
- Her medications include baclofen, miralax, dulcolax, protonix.

- On Physical Exam:
  - BP: 110/65  HR: 85  RR: 16  T: 36.8
  - Gen: Awake, moaning
  - HEENT: Dry mucous membranes
  - CVS: RRR; S1+S2+
  - RESP: good air movement b/l; no wheezes or rales
  - ABD: distended; hypoactive BS+
  - EXT: contractures of upper and lower extremities; diffuse pain on palpation of right leg
# Cerebral Palsy

- **Estimated 764,000 children and adults**
- **Abnormal brain development (pre- and post-natal)**
- **Clinical Features**
  - 70-80% have spasticity
  - 67% intellectual impairment
  - Growth problems
  - Overall, non-progressive
- **Goals of care**
  - Optimize functionality
  - Support as needed
    - Feeding
    - GU
    - Rehabilitation

## Cerebral Palsy

- **Pneumonia**
  - Most common reason for hospitalization of adults with CP
- **Risk Factors**
  - Pseudobulbar palsy
  - Aspiration of saliva or food contents
  - Poor cough reflex
  - Increased secretions
  - Scoliosis
  - Organisms similar (assess health-care acquired risk)
  - Treat as other patients
  - Vaccination prior to discharge (Influenza and Pneumococcal)

## Cerebral Palsy

- **Bone Fractures**
  - **Risk Factors**
    - Functional deterioration
    - Lack of weight bearing and ambulation
    - Osteoporosis / Osteopenia
  - **Clinical Presentation and Management**
    - Pain and acute distress
    - Plain film imaging
    - Orthopedic Co-management

## Cerebral Palsy

- **Digestive Disorders**
  - **Constipation**
    - Causes include dysmotility, hypotonia, medication side effects, and non-ambulation
    - Treat aggressively with bowel clean out, and keep on bowel regimen
  - **Delayed Gastric Emptying**
    - Diagnose and treat similar to other patients
    - Spinal abnormalities (scoliosis) worsen GI issues
      - Decreased gastric capacity
      - Increased GERD
      - Positioning for feedings
Cerebral Palsy

- Other common inpatient presentations
  - Urinary Tract Infection
  - Malnutrition / Dehydration
  - Mental Health Issues

Transition of Care

- Barriers to Transition
  - Lack of outpatient resources for adults with chronic diseases of childhood
    - IM / FP lack of familiarity and time
    - High complexity to most of these patients
    - Lack of training in these complex conditions
    - Lack of care coordination with subspecialists
    - Feeling of abandonment by patients and families

Transitions of Care

- Role of the Adult Hospitalist
  - Provide necessary inpatient care for the patient
  - Seek subspecialist consultation as indicated
  - Evaluate current outpatient follow-up
  - Identify outpatient providers in your area that will accept chronic patients
    - Med-Peds Providers
    - University Clinics (i.e. PM & R, Emory Cystic Fibrosis Center, Emory Downs Syndrome Center, Sickle Cell Clinics)
  - Coordinate care with the outpatient Physician
  - Collaborate with Multi-disciplinary team (Case Management, SW, Pharmacy)
  - Educate your Hospitalist Group on these patients that may return, and the plan of care. Consistency is key.
  - Create a Med-Peds Consult Team (long-term plan)

Transitions of Care

www.gottransition.org
Sources

- Young, Nancy. “Youth and Young Adults with Cerebral Palsy: Their Use of Physician and Hospital Services”. Arch Physical Medicine Rehab. June 2007.