The Long Road Home-
A Stability Guideline for Long Term Ventilator Dependent Children Discharging to Home
Jacquie Hanks, DNP, APRN-BC, CPNP AC/PC

Disclosures
• No commercial or financial relationships to disclose.

Learning Objectives
• Learners will recall basic concepts of discharge of long-term ventilated patients according to a guideline.
• Learners will be able to describe the complexities of discharging chronically ill ventilator-dependent children to home.
• Learners will be able to outline and discuss a guideline that will work for their institution, based on best practices.

Children with Chronic Lung Disease

Guidelines for the INITIAL Transition of Mechanically-Ventilated Pediatric Patients from Hospital to Home:
• Under certain circumstances, children with the need for long-term mechanical ventilation can be transferred directly from the hospital to home. To ensure that this transfer is carried out safely certain parameters must be met prior to discharge of the patient. These include the following:
• A formal consultation with the pulmonology service has been requested and the pulmonologist has been managing the ventilator for a period of at least 4 weeks.
• The patient will have obtained a level of clinical stability such that the transfer can be considered safe. Stability shall include the following criteria:
  1. Vital signs can be monitored every 12 hours without compromising safety.
  2. All feelings are consistently tolerated and are being delivered in a manner that is considered safe for long term care.
  3. The patient has been showing progressive and adequate weight gain and growth.
  4. Tracheostomy tube changes can be done without difficulty by nursing, RT and family members per existing policy.
  5. Children must not have evidence for “critical” airway, e.g. severe subglottic stenosis.

Prevalence Cost

<table>
<thead>
<tr>
<th>Intensive Care</th>
<th>Intermediate Care</th>
<th>Home Cost</th>
</tr>
</thead>
<tbody>
<tr>
<td>8000</td>
<td>3500</td>
<td>500</td>
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<td>2000</td>
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<td>6000</td>
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<td>9000</td>
<td>5000</td>
<td>1000</td>
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</tbody>
</table>
Guidelines (cont.)

• The patient has been clinically stable on their current ventilator for a period of not less than four weeks. Mechanical ventilation has been carried out using the same ventilator to be used in the home for 48 hours prior to discharge.
  – No emergency critical airway issues or cardiopulmonary arrest for the preceding 4 weeks.
  – No cyanotic or bradycardiac episodes requiring intervention beyond suctioning of the tracheostomy tube.
  – Capillary blood gas measurements (pCO2) repeatedly show levels of less than or equal to 60mmHg and pH within normal range (7.35–7.45).
  – No escalation of ventilation settings required during this period due to deterioration of patient status.
  – Inspired oxygen concentration is less than or equal to 40%.

• A home care company has been identified and has demonstrated that adequate home care staff will be available for, preferably, 24 hours a day initially but for a minimum of 16 hours a day for the first two weeks. Subsequently, a minimum of 8 hours per day coverage by nursing and/or RT staff who are skilled in managing children on ventilators will be available.

• A hospital based care conference has been carried out and has been attended by the appropriate hospital caregivers and the patient’s family members who will be caring for the child.

• Ventilator dependent children who will be transferred home for the first time and are in either the NICU or PICU need to be transitioned to a lower level of care based on acuity for an approximate two week period prior to discharge home.

• All family caregivers who will be responsible for taking care of the child with a tracheostomy tube at home will have completed the Children’s Hospital approved mandatory tracheostomy care teaching prior to discharge.

• The primary caregivers for home have roomed in at the hospital for at least 48 consecutive hours and shown competence with caring for the child without nursing or RT help, apart from documentation.

• Outpatient clinic follow up with pulmonology has been arranged and the date(s) provided to the family.

• As with all guidelines, patients may need slight variances from the above to accommodate a smooth transition of care.

• Any of the above parameters may be altered if there is consensus in advance to do so amongst the entire pulmonology provider team.
Where do we go from here?

References

Chaos in a Family with a Chronic Illness

Theresa Flint Rodgers, RN, CPNP-AC/PC, DNP
Nurse Practitioner
Division of Pulmonary and Sleep Medicine
UAB/Children’s of Alabama
Birmingham, AL

LEARNING OBJECTIVES

Though this case presentation, the participant will

• See how social and psychological issues impact outcomes in patients with chronic illness.
• Introduce tools that may help to determine anxiety and/or depression in patients and families

CASE PRESENTATION: KD

• 17 Year Old Caucasian Female With Cystic Fibrosis
• Diagnosed: August 1998
• Genetics: ΔF508/G542X
• Sweat test: 109

PERTINENT CLINICAL HISTORY

• Sputum Microbiology
  • P. aeruginosa
  • P. Floresens
  • MRSA
  • M. abscesses

RECENT MEDICAL HISTORY

• Hospitalized 11 times between June 2012 - April 2013 with the longest admission 12/5/13 – 2/4/13 (41 days)
• Pancreatic Insufficient
• CFRD (A1C 6.3)
• Intermittent Elevated Liver Enzymes w/o cirrhosis
• Low bone mineral density (z score = -1.9)
• Nutritional Status - at risk (BMI 27%)
• Chronic pain

DISCLOSURES

• No Disclosures
SOCIAL HISTORY

- KD lived in an apartment in a metropolitan area with mother and younger sibling. Father was not involved.
- Mother’s is involved in her children’s lives but was not a disciplinarian.
- KD no longer attended school but it is reported that she received home bound services.

CURRENT HOSPITALIZATION

- Presented for admission from ED with H/O “Pain all over”
- Lethargy
- Worsening pulmonary status

PHYSICAL EXAM

- General Appearance: acutely & chronically ill appearing, cushingoid face. Lethargic. Slurred speech.
- HEENT: NCAT. Pupil react to light, L 3mm, R 4mm. Tm’s intact. Mouth /posterior pharynx without erythema or exudate. Dry membranes
- Neck: Supple, trachea midline
- Respiratory Effort: increased WOB, suprasternal retractions.

LABORATORY RESULTS

- Na 133
- K 7.0
- Cl 94
- Glu 147
- BUN 34
- Cr 4.6
- Ca 6.3
- T. protein 6.9
- GGT 26
- ALT 2968
- AST 7917
- Alk phos 134
- T. bill 0.6
- PT 22.2 sec

TESTS

- EKG: NSR, abnormal T wave (nonspecific)
- CBG: 7.20 /57 /22.7
- CXR: coarse interstitial markings with bronchiectasis
**ASSESSMENT**

- Pulmonary Exacerbation, End stage lung disease
- AKI vs renal failure
- Dehydrated, hypovolemic
- Acute liver injury vs Liver failure

**HOSPITAL COURSE**

- NS boluses for hypovolemia
- Maintenance fluids
- IV antibiotics initiated but discontinued due to elevated BUN / Cr
- Pain management
- DNR / AND

**AKI vs Renal Failure**

- Nephrology consult
- Fluid resuscitation, IV maintenance fluids
- MEDS: Kayexalate, Continuous Albuterol, Calcitriol
- AKI r/t dehydration was the final diagnosis

<table>
<thead>
<tr>
<th>BUN</th>
<th>Phos</th>
<th>Cr</th>
<th>K</th>
</tr>
</thead>
<tbody>
<tr>
<td>12</td>
<td>3.0</td>
<td>0.7</td>
<td>2.4</td>
</tr>
</tbody>
</table>

BUN, Cr, Phos normalized within 3 days

**ACUTE LIVER INJURY VS LIVER FAILURE**

- GI consult
- Fluid resuscitation
- Acute Liver injury r/t dehydration

<table>
<thead>
<tr>
<th>AST</th>
<th>ALT</th>
</tr>
</thead>
<tbody>
<tr>
<td>37</td>
<td>275</td>
</tr>
</tbody>
</table>

**DEATH & DYING**

**CONCEPTS**

**MID-ADOLESCENTS:**
- Consolidation of self-image
- Feelings of achievement and power
- Experimentation
- Advancement of logical thought with capacity of abstract reasoning

- Perception
  - Adolescent has an adult understanding of death.
  - Death is viewed as an interruption; it is an enemy
  - Need permission to grieve
  - Represses sadness, feels anger, depression

Montgomery and et al (2006) in a case study reported that sildenafil improved exercise tolerance. The authors speculated that sildenafil improved pulmonary vascular responses to exercise.

**Management of Respiratory Failure**

- Oxygen
  - High flow O2 via nasal reservoir
  - Non-rebreather
- Morphine Aerosols
- Noninvasive Ventilation

**Chronic Pain**

- Followed by Palliative Care
- Medications: Oxycontin, Morphine, Ativan
  - MS Contin 60mg po q 12hrs
  - Morphine 30mg po q 3hrs prn breakthrough pain
  - Morphine 8mg IV q 2hrs prn
  - Lorazepam 2mg IV q 4hrs prn anxiety

**Other Diagnoses**

- CFRD
- Fluid Retention

**Types of Clinical Care**

- Preventive Care
  - IV and Oral antibiotics
  - Pulmonary Toiletries
  - Increase frequency and severity of exacerbations
  - Oxygen therapy (+/-)
  - Lab draws
- Therapeutic Care
  - Pain Management
  - Oxygen
  - Non-invasive ventilation
  - Vitamin / Pancreatic Enzymes
  - Chest physiotherapy

**Suicidal Attempts**

**A Family in Chaos: Suicidal Attempts**

**Addional Social History**

- KD's sibling has attempted suicide x 3 and is addicted to heroin.
- KD admitted x 1 for questionable suicide attempt.
- Mother has been admitted to a local hospital for drug induced stupor.

**Terminal prognosis, prolonged hospitalizations, intrusive daily regimens, impeded school attendance and socialization → poor self-image, social rejection, social isolation.**

Prevalence of depression and anxiety in patients with cystic fibrosis and parent caregivers

Psychological symptoms were reported by 6088 patients with CF and 4102 parents:
- Elevated symptoms of depression were found in 10% of adolescents, 19% of adults, 37% of mothers and 31% of fathers.
- Elevations in anxiety were found in 22% of adolescents, 32% of adults, 48% of mothers and 36% of fathers.
- Overall, elevations were 2–3 times those of community samples.


RECOMMENDATIONS

- Importance of measuring and treating mental health issues in patients and families coping with serious, chronic illnesses.
- Formation of an international committee, sponsored by both the European Cystic Fibrosis Society and the Cystic Fibrosis Foundation, to develop guidelines on mental health screening and treatment in CF.
- Annual screening of depression and anxiety in adolescents and adults with CF, and parent caregivers using the PHQ-7 and GAD-7.

PHQ (Patient Health Questionnaire)- 9

PHQ - 9

<table>
<thead>
<tr>
<th>Over the last 2 weeks, how often have you been bothered by any of the following problems?</th>
<th>Not at all</th>
<th>Several Days</th>
<th>More than half the days</th>
<th>Nearly Every Day</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Little interest or pleasure in doing things</td>
<td></td>
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<td></td>
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<tr>
<td>2. Feeling down, depressed or hopeless</td>
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<tr>
<td>3. Trouble falling or staying asleep, or sleeping too much</td>
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<tr>
<td>4. Feeling tired or having little energy</td>
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<tr>
<td>5. Poor appetite or overeating</td>
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<tr>
<td>6. Feeling bad about yourself—or that you are a failure or have let yourself or your family down</td>
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<tr>
<td>7. Trouble concentrating on things, such as reading the newspaper or watching television</td>
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<tr>
<td>8. Moving or speaking so slowly that other people could have noticed? Or the opposite—being so fidgety or restless that you have been moving around a lot more than usual</td>
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<tr>
<td>9. Thoughts that you would be better off dead or of hurting yourself in some other way</td>
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</tr>
</tbody>
</table>

If you checked any problems, how difficult have these problems made for you to do your work, take care of things at home, or get along with other people? □ Not difficult at all □ Somewhat difficult □ Very difficult □ Extremely difficult


GAD (Generalized Anxiety Disorder) - 9

GAD - 9

<table>
<thead>
<tr>
<th>Over the last 2 weeks, how often have you been bothered by any of the following problems?</th>
<th>Not at all</th>
<th>Several Days</th>
<th>More than half the days</th>
<th>Nearly Every Day</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Feeling nervous, anxious, or on edge</td>
<td></td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>2. Not being able to stop or control worrying</td>
<td></td>
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<td></td>
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<tr>
<td>3. Worrying too much about different things</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>4. Trouble relaxing</td>
<td></td>
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<tr>
<td>5. Being so restless that it’s hard to sit still</td>
<td></td>
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<td></td>
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<tr>
<td>6. Becoming easily annoyed or irritable</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>7. Feeling afraid as if something awful might happen</td>
<td></td>
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<td></td>
</tr>
</tbody>
</table>

If you checked any problems, how difficult have these problems made for you to do your work, take care of things at home, or get along with other people? □ Not difficult at all □ Somewhat difficult □ Very difficult □ Extremely difficult

CASE DEVELOPMENT

- 9 day hospital stay
  4/10/13 – 4/19/2013

- Expired from respiratory failure
  Last CBG: 7.24 / 98/ 42.3

- Life: 1/16/1995 – 4/19/2013

REFERENCES


Generalized Anxiety Disorder 7-item (GAD-7) scale

<table>
<thead>
<tr>
<th>Over the last 2 weeks, how often have you been bothered by the following problems?</th>
<th>Not at all sure</th>
<th>Several days</th>
<th>Over half the days</th>
<th>Nearly every day</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Feeling nervous, anxious, or on edge</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>2. Not being able to stop or control worrying</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>3. Worrying too much about different things</td>
<td>0</td>
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<td>2</td>
<td>3</td>
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<td>4. Trouble relaxing</td>
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<td>2</td>
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</tr>
<tr>
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<td>1</td>
<td>2</td>
<td>3</td>
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<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>7. Feeling afraid as if something awful might happen</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
</tbody>
</table>

Add the score for each column

Total Score (add your column scores) =

If you checked off any problems, how difficult have these made it for you to do your work, take care of things at home, or get along with other people?

Not difficult at all ________
Somewhat difficult ________
Very difficult ________
Extremely difficult ________

Patient Health Questionnaire (PHQ-9)

**Patient Name:** _________________________________________  **Date:** ________________

<table>
<thead>
<tr>
<th></th>
<th>Not at all</th>
<th>Several days</th>
<th>More than half the days</th>
<th>Nearly every day</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Over the <em>last 2 weeks</em>, how often have you been bothered by any of the following problems?</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>a. Little interest or pleasure in doing things</td>
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<tr>
<td>h. Moving or speaking so slowly that other people could have noticed. Or the opposite; being so fidgety or restless that you have been moving around a lot more than usual.</td>
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<tr>
<td>i. Thoughts that you would be better off dead or of hurting yourself in some way.</td>
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<td></td>
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</tbody>
</table>

2. If you checked off any problem on this questionnaire so far, how difficult have these problems made it for you to do your work, take care of things at home, or get along with other people?

**Not at all** | **Somewhat difficult** | **Very difficult** | **Extremely difficult**
---|---|---|---
|   |   |   |   |

PHQ-9* Questionnaire for Depression Scoring and Interpretation Guide

For physician use only

Scoring:
Count the number (#) of boxes checked in a column. Multiply that number by the value indicated below, then add the subtotal to produce a total score. The possible range is 0-27. Use the table below to interpret the PHQ-9 score.

Not at all (##) _____ x 0 = _____
Several days (##) _____ x 1 = _____
More than half the days (##) _____ x 2 = _____
Nearly every day (##) _____ x 3 = _____

Total score: _____

<table>
<thead>
<tr>
<th>Interpreting PHQ-9 Scores</th>
<th>Score</th>
<th>Actions Based on PH9 Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Minimal depression</td>
<td>0-4</td>
<td>&lt; 4 The score suggests the patient may not need depression treatment</td>
</tr>
<tr>
<td>Mild depression</td>
<td>5-9</td>
<td>&gt; 5 - 14 Physician uses clinical judgment about treatment, based on patient's duration of symptoms and functional impairment</td>
</tr>
<tr>
<td>Moderate depression</td>
<td>10-14</td>
<td>&gt; 15 Warrants treatment for depression, using antidepressant, psychotherapy and/or a combination of treatment.</td>
</tr>
<tr>
<td>Moderately severe depression</td>
<td>15-19</td>
<td></td>
</tr>
<tr>
<td>Severe depression</td>
<td>20-27</td>
<td></td>
</tr>
</tbody>
</table>

* PHQ-9 is described in more detail at the McArthur Institute on Depression & Primary Care website
www.depression-primarycare.org/clinicians/toolkits/materials/forms/phq9/
Learning Objectives

Project Goals

- Develop and implement a multidisciplinary Specialty Asthma Clinic for children between 5 and 18 years of age with persistent asthma referred to tertiary care by their Primary Care Provider (PCP)
- Provide national guideline based comprehensive care to improve asthma control and reduce asthma morbidity for children with persistent and poorly controlled asthma
- Evaluate PCP’s satisfaction with process

Population

- Convenience sample of children 5-18 years with persistent asthma, able to perform spirometry
- BMI < 25
- Exclude mild intermittent asthma, congenital airway, cardiac, or muscle disorders, immune disorders, sickle cell anemia & other chronic pulmonary conditions
- Families lived within 1-2 hours of clinic

Disclosures

- No financial disclosures

QI Project Selected Outcomes

- Asthma severity & control
  - Hospitalizations
  - Emergency Department visits
  - School absence
- Pulmonary Function: Spirometry
  - Oral steroid use
- Asthma Control Test scores (ACT)
- Medications filled/adherence

Guideline-based Asthma Care Provided

- Asthma control, severity and management was grounded in the National Heart Lung and Blood Institute guidelines of 2007
- INITIAL VISIT: PMH, FMH, SH, PE, spirometry, ACT or cACT administered
- Asthma severity, control, atopy status established
- Previous month (and previous years) asthma history (ED, hospitalization, school absence, medication use)
- Individualized Asthma Education provided
- Asthma Management Plan provided
Monthly Clinic Visits
- Provider evaluation included History and PE
- Hospitalizations
- ED visits
- Oral steroid use
- Asthma medication use
- School absence
- Asthma Education
- Dictated note to PCP
- Two weeks after clinic visit: phone f/u for questions or problems
- After 4 months of clinic
  - Detailed summary of child’s care sent to PCP
  - PCP questionnaire
  - Asthma Passport to family: wallet-size asthma care summary and information

Asthma Passport

Results; Visits
- Occasional missing visits; retrospective chart review

<table>
<thead>
<tr>
<th>VISITS COMPLETED</th>
<th># SUBJECTS</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Visit 1</td>
<td>15</td>
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<tr>
<td>Visit 2</td>
<td>15</td>
<td>100</td>
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<td>Visit 3</td>
<td>14</td>
<td>93.3</td>
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<tr>
<td>Visit 4</td>
<td>13</td>
<td>86.7</td>
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</table>

Summary

Selected Results

Demographics

<table>
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<tr>
<th>characteristic</th>
<th>categories</th>
<th>Number of subjects</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at enrollment</td>
<td>5 to &lt; 9 yrs</td>
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<td>66.7</td>
</tr>
<tr>
<td></td>
<td>9 to &lt; 13 yrs</td>
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<td>20.0</td>
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<tr>
<td></td>
<td>13 to &lt; 18 yrs</td>
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<td>13.3</td>
</tr>
<tr>
<td>Gender</td>
<td>Male</td>
<td>6</td>
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<td></td>
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<td>African Amer.</td>
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<td></td>
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<tr>
<td></td>
<td>Private/Other</td>
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<td>40.0</td>
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<tr>
<td>Attend School</td>
<td>Yes</td>
<td>15</td>
<td>100.0</td>
</tr>
<tr>
<td>Smokers in home</td>
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<tr>
<td></td>
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<td>12</td>
<td>80.0</td>
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</tbody>
</table>

Baseline Severity

<table>
<thead>
<tr>
<th>Severity of Asthma for Participants</th>
<th>Number of subjects</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mild Persistent</td>
<td>1</td>
<td>6.7</td>
</tr>
<tr>
<td>Moderate Persistent</td>
<td>8</td>
<td>53.3</td>
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<tr>
<td>Severe Persistent</td>
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Baseline & Follow-up Visits: Asthma Control

Asthma Control at Each Visit for Sample

Visit 1 Visit 2 Visit 3 Visit 4

Percentage of Patients with ER Visit

Visit 1 Visit 2 Visit 3 Visit 4

School Absences

Visit 1 Visit 2 Visit 3 Visit 4

FEV 1 at each visit

Visit 1 Visit 2 Visit 3 Visit 4

FEF 25-75% at each visit

Visit 1 Visit 2 Visit 3 Visit 4

ACT & Lung Function:
Baseline & Last Visit

<table>
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<tr>
<th>Outcome</th>
<th>Subjects with data at both visits</th>
<th>Visit 1 (Mean (SD))</th>
<th>Visit 4 (Mean (SD))</th>
<th>Change 1 Mean (SD)</th>
<th>Paired t-test</th>
<th>P-value</th>
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<td>ACT score</td>
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<td>96.7 (11.4)</td>
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<td>75.0 (21.7)</td>
<td>90.7 (22.2)</td>
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Outcomes; Baseline and Last Visit

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<th>MONEDER'S TEST EXACT P-VALUE</th>
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Conclusions

- Pilot Asthma clinic sought to improve asthma management and outcomes and reduce asthma morbidity in children with persistent asthma (ages 5–18)
- Population N of 15 represented a diverse group of children, majority covered through Medicaid
- At baseline, majority experienced poorly controlled or not well controlled asthma in setting of moderate to severe persistent disease with improvement in control at 4 months
- Although asthma control improved from baseline, we achieved well controlled asthma in only 45% after 4 visits but improved from 6% at baseline
- Participation was associated with significantly reduced ED visits, improved pulmonary function (FEV1 and FEF25–75%) per spirometry, and less missed school

References

Evaluation and Management of Children with Congenital Diaphragmatic Hernia (CDH)

Cheryl Samuels RN, MSN, CPNP-AE, Ashley Harmon, RN, MSN, FNP, Ricardo Mosquera MD, and Matthew Harting, MD, MS

Disclosures

- The authors have documented that they have no financial relationships to disclose or Conflicts of Interest (COIs) to resolve.

Learning Objectives

- Understand the anatomy and pathophysiology of CDH
- Describe the treatment and management of CDH, including pre- and post-operative interventions
- Recognize the long term pulmonary sequelae of CDH
- Introduce clinical pearls to aid primary providers

Congenital Diaphragmatic Hernia (CDH)

- A congenital defect of the diaphragm which allows abdominal organs to enter the chest cavity
- Impedes development of chest anatomy, including pulmonary structures and blood vessels
- Approximately 1 of 2,500 live births
- Multiple long term complications

Chest X-Ray

Staging of CDH

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<th>A</th>
<th>B</th>
<th>C</th>
<th>D</th>
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<td>Frequency</td>
<td>13%</td>
<td>44%</td>
<td>30%</td>
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<td>Survival</td>
<td>99%</td>
<td>96%</td>
<td>78%</td>
<td>58%</td>
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Neonatal Treatment

- If known defect, intubation at delivery or when CDH identified
- ECMO if needed to stabilize patient
- Surgical intervention when stabilized
  - Primary repair or patch

Patch Repair: Laparotomy Approach

Long-term morbidity in CDH

- Pulmonary: pulmonary hypoplasia and pulmonary hypertension
- Gastrointestinal: GERD, FTT, malrotation
- Musculoskeletal: Scoliosis, chest abnormalities
- Neurocognitive impairments

Pulmonary Sequelae

A combined average of 30% of all children with CDH will have pulmonary morbidity at discharge from NICU
Factors that increase risk include
- Earlier gestational age at birth and lower birth weight
- C/D defect size
- Patch repair
- Longer days of ventilation required
- ECMO
Pulmonary Morbidity

- Pulmonary Hypoplasia
- Chronic Lung Disease (80% have abnormal CXR in NICU)
  Obstructive Breathing Patterns (25% at 5 years)
  Restrictive Breathing Patterns
- Treatments: Oxygen, medications and even tracheostomy
- Increased risk for Pneumonia (7% in first year)

Pulmonary Hypertension

- Pathophysiology: Smaller pulmonary bed, thicker vessels, and less-responsive vessels
- Treatments: Nitric Oxide, Sildenafil, Bosentan, oxygen, diuretics, keep children calm
- Long term follow up care: ECHO, CXR and proBNP (monthly CBC/CMP for Bosentan)

High-risk multi Disciplinary Clinic for Congenital Diaphragmatic Hernia (HDC-CDH)

- We have a long term, multi-disciplinary follow up clinic
- Included: Pediatric Surgery, Pulmonary, GI, Cardiology, Neuro-Development, dietician and 2 NPs
- Multi-disciplinary presentations
- Designated contact person for questions or concerns

### Follow-up Schedule

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<tr>
<th>Time Period</th>
<th>Neurology</th>
<th>Neurodevelopmental</th>
<th>Pulmonary</th>
<th>Cardiology</th>
<th>Gastrointestinal</th>
<th>Orthopedic</th>
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High-risk multi Disciplinary Clinic for Congenital Diaphragmatic Hernia (HDC-CDH) A/B

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</table>
PCP Pearls for the Primary Care of CDH

- Pediatric Surgery: Every 2-6 months with issues, annual if doing well
- Pulmonary: First year then prn
  - Follow CXR, ECHO, pro BNP
  - Immune: (Synagis, Influenza, Pneumovax)
  - Follow PFT starting at age 5, may need VQ scan between 1-3 years if symptomatic
  - Treat airway obstructive disease, airway clearance
- GI: Follow growth velocity, look for GERD and increase risk of aspiration, dysmotility
- Recurrence: (Rate is 8-50%)
  - Increased risk is patch vs primary repair and stage of defect

Signs and Symptoms of Recurrence

The 2 major presentations of recurrence:

1. GI: emesis, bowel obstruction, increase GER
2. Pulmonary symptoms: increase in cough, increase WOB, hypoxemia

Additionally, can be subtle/silent thus the need for CXR surveillance

Conclusion

- CDH is an uncommon condition but important to recognize long term complications
- Following intense, early treatment there are long term complications
  Pulmonary & GI predominate
- Comprehensive evaluation and follow up is key

References


Contacts

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