The Speaking Valve and Diaphragm Pacing: Changing Lives

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Objectives

- Describe the physiological impact of respiratory insufficiency in persons with high spinal cord injuries, amyotrophic lateral sclerosis (ALS) and chronic central hypoventilation syndrome (CCHS)
- Discuss the application of diaphragm pacing as an alternative or adjunctive therapy to positive pressure ventilation
- Summarize the role of the Passy Muir Valve in optimizing outcomes for tracheostomy patients with a diaphragm pacing system.

What do these 3 people have in common??

- Mollee, an 18- year old high school senior takes dance classes and loves to play softball
- Kelli, a 42-year old veterinary assistant from Emporia, Kansas
- Trent, a 40- year old husband and father of 2 young girls

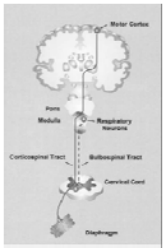
They all have respiratory insufficiency.



And they all have a diaphragm pacer.

How do we breathe?

- Volitionally (day time) breathing is controlled by brain's respiratory control center.
- Night breathing is regulated by CO_2 levels measured in the brain.
- Signals go down the nerves in the neck to the chest and down to the diaphragm.
- Phrenic nerve branches out into the muscles of the diaphragm, causing the diaphragm muscles to contract



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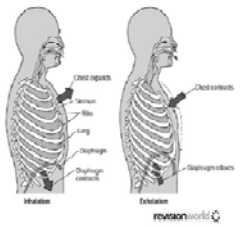
How do we breathe?

- The diaphragm contracts creating **negative pressure** in the chest cavity.
- To equalize the pressure, air rushes into the lungs.
- When the diaphragm relaxes, the elasticity of the lungs and chest wall pushes the air out of the lungs



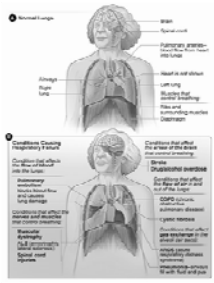
What causes respiratory insufficiency?

- **Acute injury or trauma**
  - an injury to the chest/ribs
  - an injury to the lungs: inhaling harmful fumes or smoke
- **Drug or alcohol overdose**
  - affects the area of the brain that controls breathing
- **Problems with the spine**
  - e.g. scoliosis
- **Lung diseases and conditions**
  - COPD, pneumonia
  - ARDS (acute respiratory distress syndrome)
  - pulmonary embolism, cystic fibrosis

What causes respiratory insufficiency?

Conditions that affect the nerves and muscles that control breathing

- spinal cord injuries (SCI)
- amyotrophic lateral sclerosis (ALS)

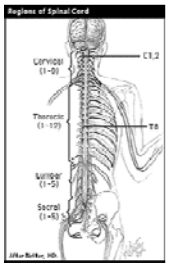


National Heart Lung and Blood Institute (NHLBI)

SCI: what are the facts?

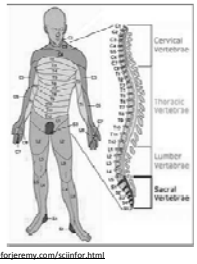
National Spinal Cord Injury Statistical Center (NSCISC)

- Approximately **12,000** new spinal cord injuries (SCI) each year in the United States.
- In 2008 there was an estimated **259,000** persons in the U.S. living with a SCI.
- Nearly half of the people with SCI exhibit tetraplegia
- Approximately 40% of all persons with C1-C4 SCI are vent dependent .



Impact of a high-cervical injury

- Bathing, dressing, grooming
- Meal preparation, eating
- Bowel care, bladder care
- Turning in bed
- Transfers
- Skin inspection
- Pressure relief, ROM
- Medications
- Suctioning



www.hopeforjeremy.com/sciinfor.html

So what does this mean to Trent?

- August, 2012 surgery to remove a mass on his spine: C3-C4
- Paralyzed, able only to turn his head and shrug his shoulders
- Total dependence on mechanical ventilator to stay alive
- Complete dependence for all of his cares
- Unable to eat or speak

What is ALS?

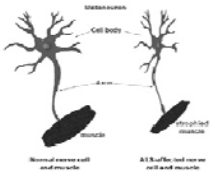
- Amyotrophic lateral sclerosis (ALS)
  - is also known as Lou Gehrig's Disease
  - or motor neuron disease.

http://www.alsa.org/about-als

- It is a relentlessly progressive and fatal disease
  - characterized by motor neuron degeneration
  - affecting nerve cells in the brain and the spinal cord.

What is ALS?

- Upper motor neurons (C1, C2) reach from the brain to the spinal cord
- Lower motor neurons (C3, C4) reach from the spinal cord to the muscles
- When the motor neurons die, the ability of the brain to initiate and control muscle movement is lost = functional paralysis



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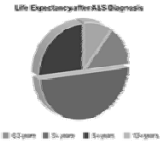
ALS: What are the facts?

- Approximately **5,600** people are diagnosed with ALS each year(U.S.)
- It is estimated as many as **30,000** Americans have the disease at any given time



- Half of all people affected with ALS live **three** or more years after diagnosis.
- Twenty percent live **five** years or more
- Up to 10% will live more than **ten** years.

Respiratory symptoms of ALS

Although ALS has no direct effect on the lungs, it has devastating effects on the mechanical function of the respiratory system.

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So what does this mean for Kelli?

- No longer able to work at the job she loves
- No longer able to live independently
- Mother is care giver: dressing, bathing
- Losing her ability to speak
- Eventually unable to eat or even swallow
- Decreased quality of life
- Significantly shortened life expectancy

What *is* Congenital Central Hypoventilation Syndrome?

- CCHS: previously known as "Ondine's Curse"
- Failure of autonomic control of breathing
- Sluggish or absent response to low blood oxygen saturation (hypoxemia) or to CO2 retention (hypercapnia).

(Remember how we breathe at night?

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So what does this mean to Mollee?

- Early childhood spent on a ventilator 24/7
- Infant and toddler: learning to speak, sit, walk; developmental milestones
- Adolescence: nighttime ventilator
- Quality of life limited: slumber parties, camping, swimming

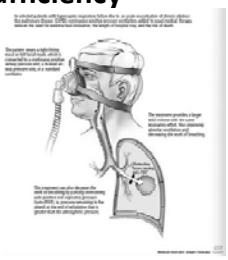
Treatment options for respiratory insufficiency

- Non-invasive ventilation (NIV)
- Tracheostomy mechanical ventilation (TMV)
- Diaphragm pacing

Treatment options for respiratory insufficiency

NIV

- May be difficult to fit/leaks
- Trial multiple interfaces
- Skin breakdown
- Limits eating, drinking
- Limits mobility
- Compliance



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Treatment options for respiratory insufficiency

TMV

- Mobility/transfers
- Tubing
- Humidity
- Trach/torque
- Disconnects
- Batteries

Impact of advanced ventilator technology

Improved technology = improved quality of life

- Improved portability & longer battery life = increased independence
- Increased travel = decreased isolation
- Decreased burden of care

Physiological risks of mechanical ventilation

<u>Positive</u> Pressure Ventilation forces air into the lungs

- Barotrauma and tracheal injury
- Posterior atelectasis
- Respiratory infections/VAP
- Respiratory muscle disuse atrophy
- Hemodynamic instability
  - Decreased venous return
  - Decreased cardiac output

What *is* diaphragm pacing?

The use of electrical signals to stimulate the diaphragm to contract.

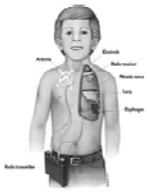
History of phrenic nerve pacing

- August 1966 Dr. William Glenn reported on the first use of radiofrequency coupled receivers and electrodes to pace the diaphragm
- First commercial distribution in the early 1970's (Dr. William Glenn and Avery Biomedical)
- FDA mandated the regulation of all medical devices sold in the U.S. in 1986

Phrenic nerve pacing

The Avery Breathing Device

- Cervical or thoracic surgical placement
- Electrodes are implanted on the phrenic nerves and attached to an implanted stimulator
- Powered by an external controller through the skin via a radiofrequency link



www.averybiomedical.com

Pacing the Diaphragm

NeuRx DPS®

- In development over 20 years
- First implant in March 2000
- Christopher Reeve 2003
- FDA approved 2008 for SCI
- FDA approved in 2011 for ALS

www.synapsebiomedical.com

NeuRx DPS®

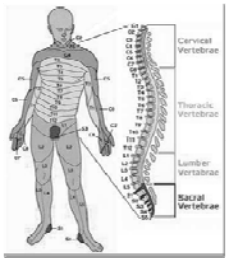
- Laparoscopic procedure
- Four electrodes implanted in the diaphragm, with a fifth electrode to complete the circuit
- Small external battery powered pulse generator
- Connector holder and cable

www.synapsebiomedical.com

NeuRx DPS® and the SCI patient

Review: impact of a cervical injury

- Impaired/loss of sensation and function below the level of the injury
- Phrenic nerve originates at C3-C5
- Lower motor neurons: C3-C5



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Will a diaphragm pacer work?

Candidates must have

- A stimulatable diaphragm
- Lower motor neurons intact
- Phrenic nerve function (phrenic nerve EMG)

Surgical procedure

- Performed by a general surgeon
- Minimally invasive, approx. 90 minutes
- 4 dime-sized holes in the abdominal region
- Laparoscope inserted to see muscle
- Typically done as outpatient or overnight stay

Surgical procedure

The first step:

- Motor point mapping
- Optimal site for electrodes
- Four electrodes
  - Two anterior
  - Two posterior

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NeuRx DPS® exit site

www.synapsebiomedical.com

Trent's rehab journey

Within 24 hours of his admission, Trent's SLP and RT successfully initiated the Passy Muir Valve.

For the first time in almost two months Trent and his family felt hope for their future.

Trent's rehab journey

Timeline:

- Admitted to Madonna on Sept. 7th, 2012 after spending 3 weeks in an ICU
- Initial team evaluations, including RT and SLP co-treat and assess for use of the PMV
- Immediate impact on Trent's motivation and willingness to participate in his rehab

Trent's rehab journey

- Surgery: at SERMC by Dr. Greg Fitzke Oct. 30th
- Overnight stay, returned to Madonna on Oct. 31st
- Started diaphragm pacing Nov. 1st
- Less than 3 weeks later completely ventilator-independent!

What about Kelli?

Diaphragm Pacer, SCI & ALS:

What's the same?

- The device
- The procedure

What's different?

- Everything else!

ALS: what's different?

Diaphragm Pacer Qualifying criteria

Phrenic nerve function as tested by <u>either:</u>

- Neurophsyiological testing
- Visualizing diaphragm contraction with fluoroscopy (a full motion x-ray)
- By radiographic techniques (such as ultrasound)

ALS: what's different?

Diaphragm Pacer Qualifying criteria

Chronic hypoventilation as measured by <u>either</u>:

- FVC less than 50% predicted
- MIP less than 60 cmH2O
- PCO2 greater than or equal to 45 mm Hg
- SaO2 less than 88% for 5 consecutive minutes during sleep

ALS: what's different?

Conditioning the diaphragm

- **Daytime**
  - When there is little respiratory compromise
- **Nighttime**
  - Any sleep disordered breathing
- **With NIV**
  - Use DPS® with NIV
- **Full time**
  - Respiratory instability
  - Moderate respiratory decline

ALS: what's different?

End goals

- Significant improvement in survival from diagnosis (by 16 months)and from the start of NIV (by 9 months) compared to standard –of-care NIV
- A 16-month survival from implant for patients with no other respiratory options (intolerant or unable to use NIV)
- Significant sleep improvement after 4 months of DPS conditioning

www.ClinicalTrials.gov

What about Mollee?

Diaphragm Pacer, SCI, ALS & CCHS:

What's the same?

- The device
- The procedure
- Initial programming: similar to ALS

What's the different?

- Everything else!
- CCHS is an off-label use of the NeuRx DPS®

CCHS: what's different?

Qualifying criteria

- The NeuRx DPS™ is considered an off-label use
  - HUE (Humanitarian Use Exception)
  - Required IRB approval
- Often make great candidates as typically have no underlying lung disease and a healthy diaphragm.
- Polysomnography:
  - SaO2 less than 88% for 5 consecutive minutes during sleep

CCHS: what's different?

Conditioning the diaphragm

- Little to no atrophy has taken place
- Build up to 8 hours daytime usage
- Adjust to sensation
- Adjust settings for maximum contract w/out pain

CCHS: what's different?

End goal

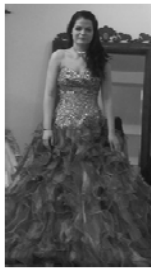
- Discontinue positive pressure mechanical ventilation
- Discontinue tracheostomy tube
- DPS® for nocturnal respiratory support

CCHS: what's different?

End goal

Helping Mollee reach her goal

- Condition the diaphragm with focus on adjusting to pacing for ≥8 hrs daytime
- Change from PMV® to trach cap
- Use of ventilator and DPS® at night, gradually wean the vent
- Trach capped (no DPS®) during the day, DPS® and PMV® at night
- Consider decannulation

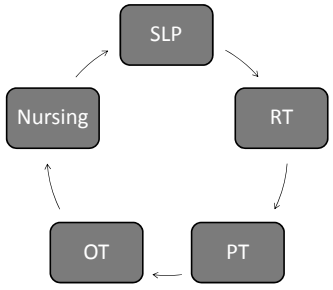


Care Planning for the DPS® patient

Interdisciplinary Team

- Assist in diaphragm conditioning and the transition from mechanical ventilator to DPS®
- Continue focus on functional gains and maximizing independence
- Patient and family education for transition to home

Team work = success!



Team collaboration is key

- Treatment team identifies 1-3 critical activities necessary for the patient to reach his/her goals
- Any discipline can initiate a strategy that is designed to address a critical activity
- All team members support the strategy by incorporating it into their treatment plan
- Results in a 24/7 rehab environment and the best possible outcomes

Interdisciplinary team roles

Collaboration is key to success!

OT & PT:
- pec stretches
- chest excursion
- positioning tactics
- adapt assistive devices

RT:
- measuring exhaled tidal volumes
- monitoring oximetry
- ABGs
- CO2 retention

Interdisciplinary team roles

RT & SLP:
- introduce use of PMV®
- transition to trach cap

- **RT & Nursing:**
- skin integrity
- exit site care
- monitoring vitals
- VAP prevention

Interdisciplinary team roles
therapeutic interventions

- Breath stacking
- Diaphragmatic breathing
- Expiratory trainer
- Incentive spirometry
- Pectoral stretch
- Vibratory PEP
- Sniffing

Interdisciplinary team roles
therapeutic interventions

- Breath stacking
- Diaphragmatic breathing
- Expiratory trainer
- Incentive spirometry
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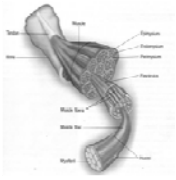
- Sniffing

Interdisciplinary team roles
positioning strategies

- Shoulder abduction and external rotation
- Pectoralis stretch
- Positioning for chest expansion
- Shoulder preparation for functional training
- "Iron cross" technique in bed/mat table

Interdisciplinary team roles
Conditioning the diaphragm

- Slow twitch muscle fibers: fatigue resistant, diaphragm must work 24/7
- Diaphragm is 70% slow twitch (oxidative)Type I
- Conversion of slow twitch to fast twitch with atrophy of diaphragm while on mechanical ventilation
- Convert fast twitch back to slow twitch



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Interdisciplinary team roles
SLP

- Eating and swallowing are complex neuromuscular activities
- A high cervical injury results in a loss of coordination between breathing and swallowing
- Use of the PMV® reduces the risks of aspiration during oral intake
- This is even more significant for the SCI/DPS® patient

Interdisciplinary team roles
SLP strategies

Interdisciplinary team roles
SLP & OT

- Computer access
- Assistive technology
  - Voice Activated Remote
  - Voice Recognition Software
  - Integrated Phone & PDA's
- Training in breath support, timing, phonation
  - Online bill paying, grocery shopping, Christmas shopping, social networking
  - Job skills, volunteer work, community participation

Interdisciplinary team roles
Neuropsychology

Kipp Ransom, Rehabilitation Counselor

"The benefit of verbal communication is that patients can express their thoughts and concerns as they did prior to their need for a tracheostomy.

If the PMV is not used, my sessions become more challenging as the patient has to rely on non-traditional forms of communication. The PMV has greatly enhanced my patients' ability to communicate with me and ultimately have successful outcomes."

Interdisciplinary team roles
Physical Therapy

Interdisciplinary team roles
Physical Therapy

Resources

- Chen, M L, et al: Diaphragm Pacers as a Treatment for Congenital Hypoventilation Syndrome. Expert Rev. Med. Devices 2(5), 577-585, 2005
- History of Phrenic Pacing/Avery Biomedical Devices, Inc. available at www.averylabs.com
- National Spinal Cord Injury Statistical Center, Spinal Cord Injury: Facts and Figures at a Glance. Available at www.nscisc.uab.edu
- Onders RP. Phrenic Nerve and Diaphragm Motor Pacing. Rice TW, editor. Pearson's Thoracic and Esophageal Surgery,
  Third Edition. Philadelphia, PA: Churchill Livingstone Elsevier, 1145-1457, 2007.
- Respiratory Management for Spinal Cord Injury, Consortium Guidelines, The Consortium for Spinal Cord Medicine. Available at www.PVA.org
- Onders R et al, Study Results of Diaphragm Pacing in Extremely Low Forced Vital Capacity Patients with Amyotrophic Lateral Sclerosis/Motor Neuron Disease: Is There a Role at End Stage ALS? ALS/MDA Sessions 2010

- Onders R et al, Study Results of Diaphragm Pacing in Extremely Low Forced Vital Capacity Patients with Amyotrophic Lateral Sclerosis/Motor Neuron Disease: Is there a ROLE at End Stage ALS? ALS/MDA Sessions 2010

  3 21 CFR § 814.118(a)(3).

* Clinical Indications for Noninvasive Positive Pressure Ventilation in Chronic Respiratory Failure Due to Restrictive Lung Disease, COPD, and Nocturnal Hypoventilation—A Consensus Conference Report. Chest, 1999. 116(2): p. 521-534.5. Miller, R.G., et al.,

- The care of the patient with amyotrophic lateral sclerosis (an evidence-based review):Neurology, 1999. 52(7): p. 1311-236 Lechtzin N et al.
- Hospitalization in amyotrophic lateral sclerosis Causes, costs, and outcomes, Neurology Volume 56 • Number 6 • March 27,2001Rello, Jordi, MD et al.
- "Epidemiology and Outcomes of Ventilator-Associated Pneumonia in a Large US Database." Chest, 122(6) 2002, pp.2115-2121.

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- You will have 5 days from the time this courses ends to complete the evaluation, which is required to receive credit.
  - Look in your email for a reminder link, or type this into your Internet browser's address bar:
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- If you are a **late registrant**, the meeting code is: **k2598p062**
  - If you are already registered, you do not need to use this code