

Standards of Care in Respiratory Management Duchenne MD

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A brief history of Standards of Care

- MDA never established a set of minimal standards
 - CF Foundation had done so decades earlier
- First Respiratory Care guidelines: 2004

American Thoracic Society Documents

Respiratory Care of the Patient with Duchenne Muscular Dystrophy

ATS Consensus Statement

THIS OFFICIAL STATEMENT OF THE AMERICAN THORACIC SOCIETY WAS APPROVED BY THE ATS BOARD OF DIRECTORS MARCH 2004.

AJRCCM, August 15, 2004

Gaining World-wide Acceptance...



Documenti dell'American Thoracic Society

Assistenza Respiratoria del paziente con Distrofia Muscolare Duchenne

IL DOCUMENTO UFFICIALE DELL'ATS È STATO APPROVATO DAL CONSIGLIO DEI DIRETTORI DELL'ATS NEL MARZO 2004.

Italian translation courtesy of PPMD Italy

The Respiratory Management of Patients With Duchenne Muscular Dystrophy: A DMD Care Considerations Working Group Specialty Article

David J. Birnkrant, MD,^{1*} Katharine M.D. Bushby,² Raouf S. Amin, MD,³ John R. Bach, MD,⁴ Joshua O. Benditt, MD,⁵ Michelle Eagle, PhD,² Jonathan D. Finder, MD,⁶ Maninder S. Kalra, MD,³ John T. Kissel, MD,⁷ Anastassios C. Koumbourlis, MD,⁸ and Richard M. Kravitz, MD⁹

Pediatr Pulmonol: 45:739-48, 2010.

Update: October 2018 Pediatrics:

Respiratory Care of Patients with Duchenne

Muscular Dystrophy. David J. Birnkrant, Daniel W.

Sheehan, Joshua O. Benditt, Michelle Eagle, Jonathan D.

Finder, John T. Kissel, Richard Kravitz, Hemant

Sawnani, Richard Shell, Michael D. Sussman, Lisa Wolfe

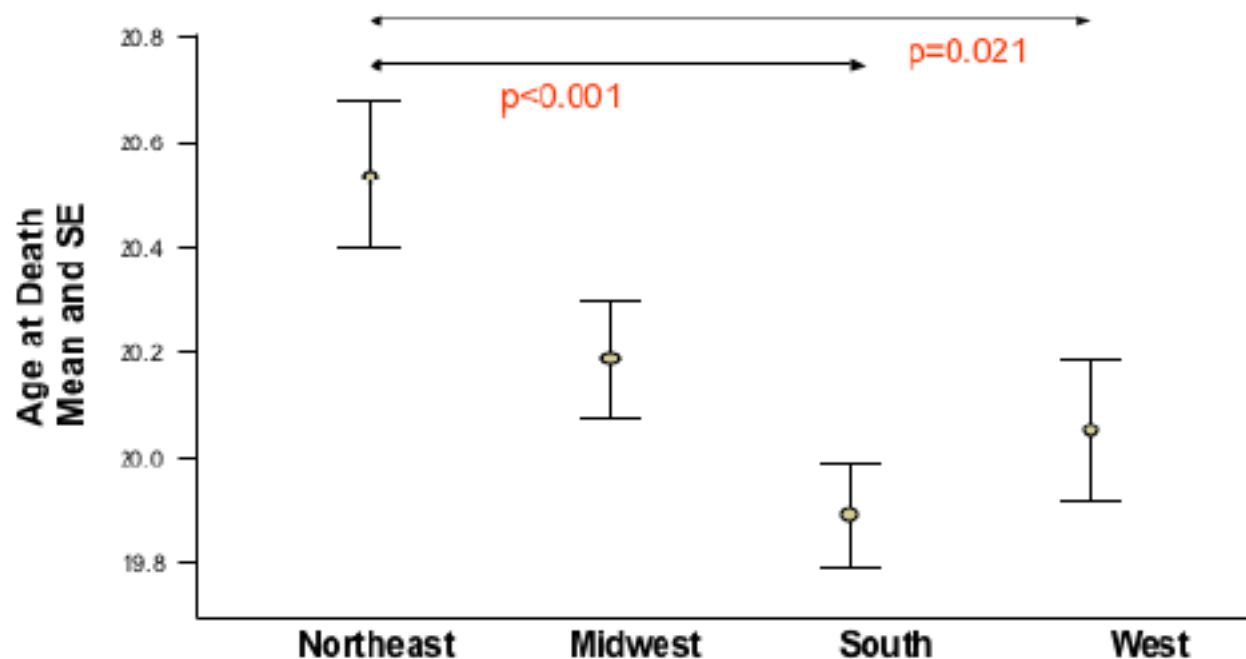
2018 Updates to Care Recommendations

- Simplified criteria for initiating of
 - Assisted coughing
 - Assisted ventilation
 - Starting therapy with higher levels of function
- Continued PREVENTIVE approach
- Lung volume recruitment FVC <60%

WHY Standards of Care?

- Survival used to vary greatly by center
- Some docs took a “hands off approach
- “Therapeutic nihilism” abounded
 - “take him home and love him, nothing can be done”
 - That approach is history

Figure 3: Regional Differences in Median Age at Death



Median	20	20	19	20
Mean	20.539	20.188	19.889	20.251
N	945	1334	1700	878

"a rising tide lifts all
boats"

You can thank PPMD for establishment of Standards of Care



PARENTS were
the main movers
to get things
BETTER for their
boys

Basics of Standards of Care

- Basic access to specialty care
 - Pulmonology, Cardiology among others
- Standardized approach to testing
 - Pulmonary function testing on a regular basis
 - Can head off trouble
 - Sleep studies
- An emphasis on non-invasive support

Basics Concepts of Respiratory Care

- Assisting cough when coughing muscles get weak
- Assisted breathing in sleep
- Assisted breathing awake
 - An emphasis on non-invasive support

Stages of respiratory dysfunction in Duchenne MD

1. Normal (age 0-10)
 - Vaccinate, educate
2. Inadequate cough (age 10-15)
3. Inadequate night time breathing (age 15-20)
4. Inadequate daytime breathing(>age 17)
 - These ages vary greatly!

Assisting Cough

- Access to assisted coughing is now universal
 - Prior to 2004 considered “experimental”
- Newer concept: LUNG RECRUITMENT
 - “pops” open areas of partial collapse
 - Prevents pneumonia
 - Stretches intercostal muscles to prevent chest wall rigidity

Assisting Breathing in Sleep

- Need predicted by FVC <30%
 - Sleep studies once FVC < 50%
- Signs include: A.M. headaches, Increasing awakenings, sleepiness, poor school performance, etc
- Low O₂ saturation on overnight oximetry
- High CO₂ on sleep study

Indicators of Need for Mechanical Ventilation in Duchenne Muscular Dystrophy and Spinal Muscular Atrophy*

Søren Lyager, MD; Birgit Steffensen, SRP; and Bent Juhl, DMedSc

Chest 1995;108:779-85

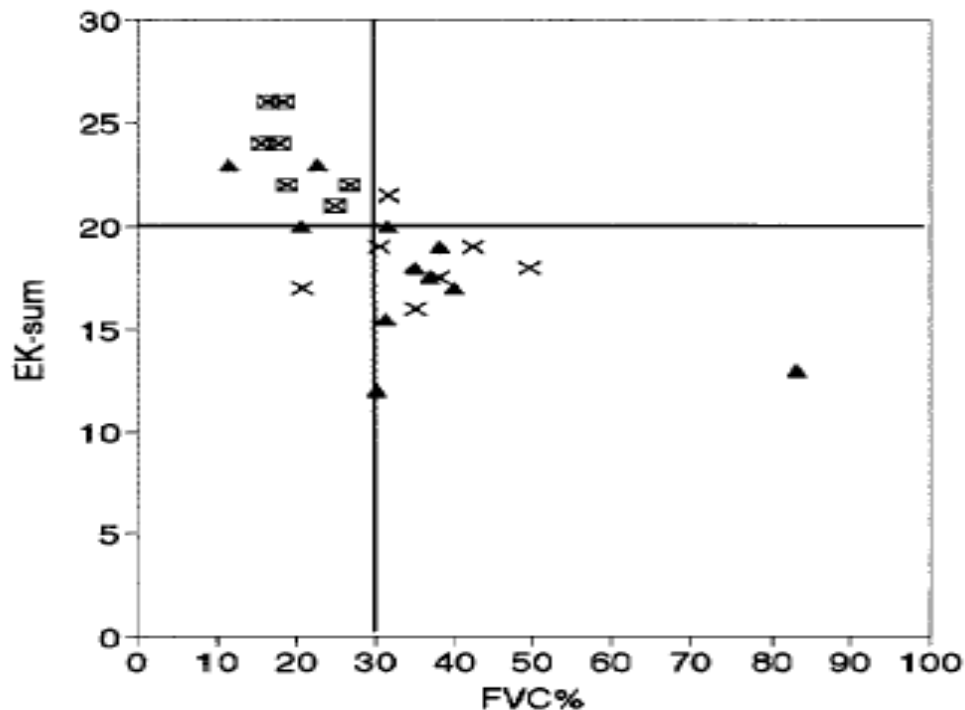


FIGURE 1. Relationship between disability score and the forced vital capacity in percent of measured value (FVC%) in the DMD and the SMA-II groups. EK sum vs FVC% for the patients with DMD (x) and the patients with SMA-II (triangles). Those who were later mechanically ventilated are identified by boxes. The vertical line indicates FVC%=30%; horizontal line indicates EK sum=20.

Note that FVC<30% in DMD correlates to ventilatory failure (but not in SMA)

Selected cohort FVC% and funct score pred of need for MV in DMD not SMA II (youngest pt 9 yrs)

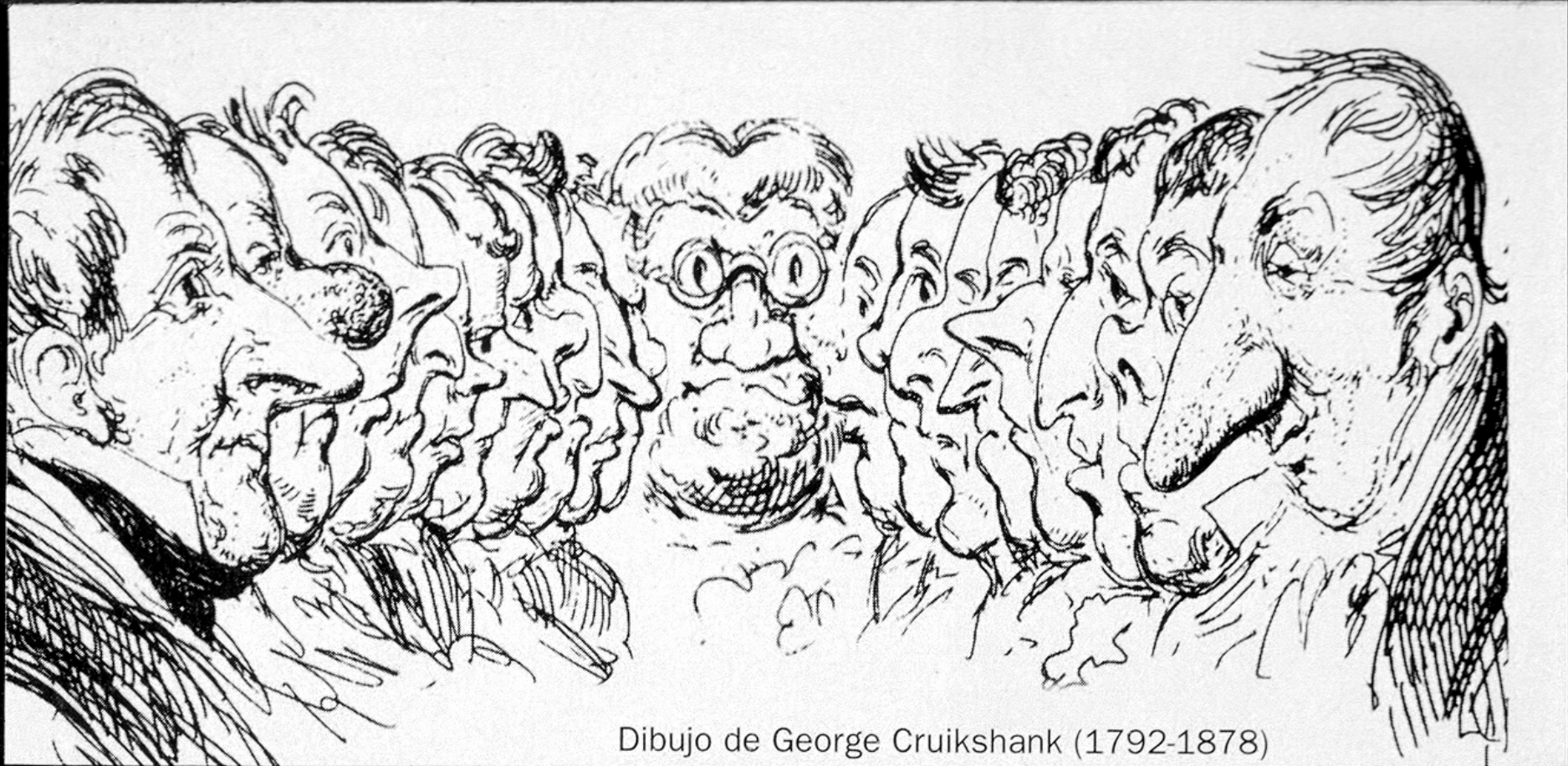
Special warning: O₂ can be hazardous to your health...

- Supplemental O₂ can be dangerous
- Suppresses respiratory drive
- Can precipitate respiratory failure
- Low saturation means increased airway clearance, need for increased ventilation
- Pulse oximeter very helpful – check CO₂!

Management of breathing in sleep

- Most avoid tracheostomy,
- Oxygen does NOT help
- BiPAP/VPAP/Ventilator
 - (NOT CPAP)
- Sleep studies
- Mask fit essential!

Bear in mind: Many faces -- many interfaces



Dibujo de George Cruikshank (1792-1878)



oronasal



nasal



pillows



total face



mouthpiece



helmet

Various interfaces used for noninvasive positive pressure ventilation.

Non-invasive ventilation is NOT new!

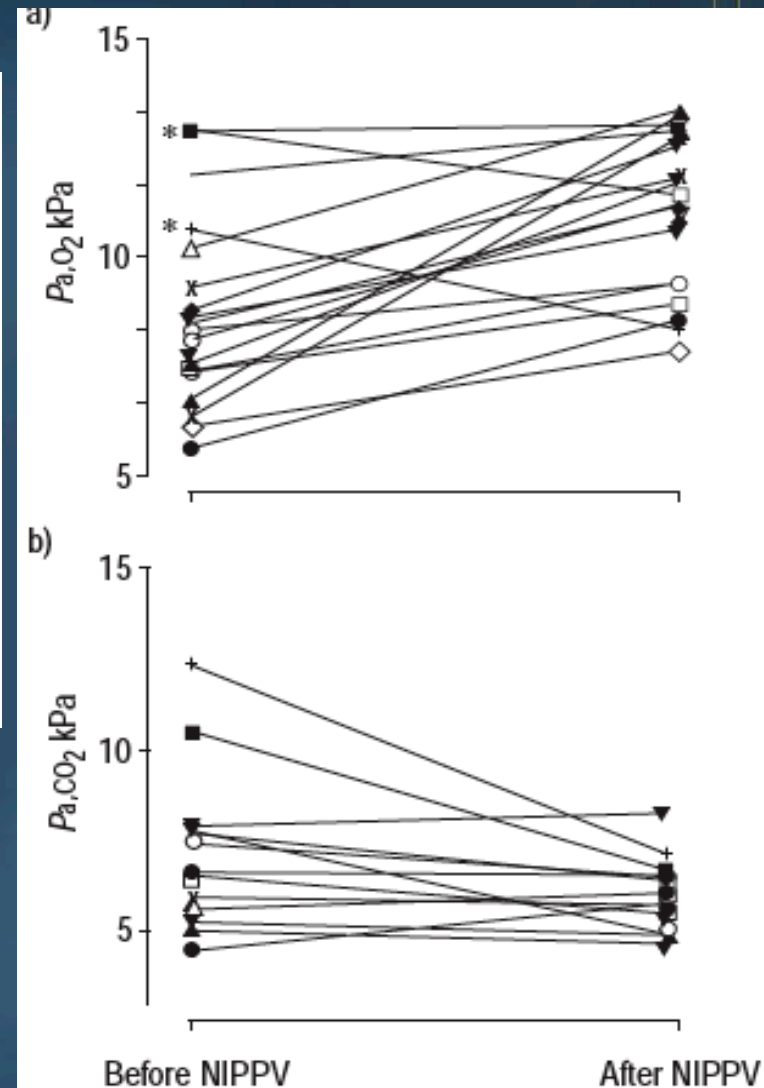


1952 Emerson infant "iron lung:" the first non-invasive ventilator

Breathing better at night: you breathe better awake!

Table 1. – Diagnostic categories, mean age at starting noninvasive positive pressure ventilation (NIPPV) and diurnal arterial oxygen (P_{a,O_2}) and carbon dioxide (P_{a,CO_2}) tensions before and after NIPPV by diagnostic categories

Diagnosis	Age yrs	Before NIPPV		After NIPPV	
		P_{a,O_2} kPa	P_{a,CO_2} kPa	P_{a,O_2} kPa	P_{a,CO_2} kPa
CMD	11.6±3.7	8.2±1.2	6.6±1.28	10.65±2.2	5.97±0.9
SMA	5.7±4.2	7.9±1.5	6.1±0.85	11.9±1.1	5.4±1.2
CM	7.3±3.3	8.2±0.6	7.73±1.25	11.3±1.15	6.00±0.9
DMD	13.9±1.5	9.3±2.3	6.33±1.13	12.17±1.46	5.75±0.41
Misc	9.4±4.8	9.4±2.3	7.6±1.37	9.33±1.87	6.33±1.1
Overall		8.5±1.8	7.0±1.6	10.9±1.7	5.9±0.8

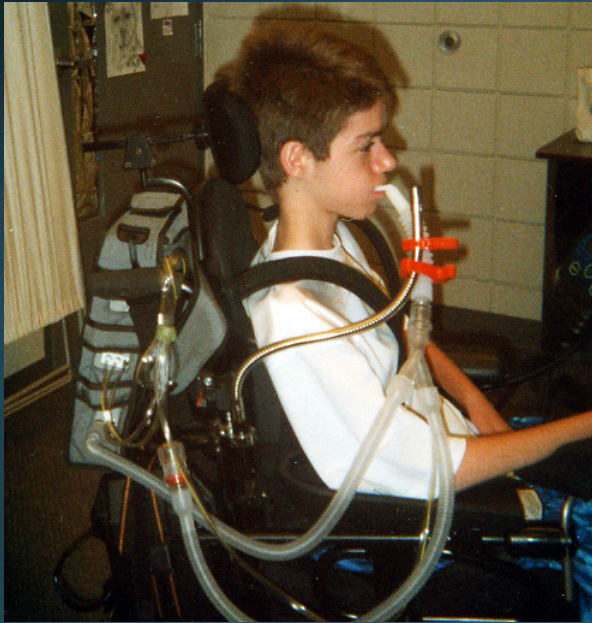


Daytime breathing support

- Most patients can be managed with mouthpiece ventilation
- Newer, lightweight ventilators facilitate portability and remaining in school or at work

Gomez-Merino, E., and J. R. Bach. 2002. Duchenne muscular dystrophy: prolongation of life by noninvasive ventilation and mechanically assisted coughing. *American Journal of Physical Medicine & Rehabilitation* 81(6): 411-5

Non-invasive breathing support



Portable ventilator with a mouthpiece attached
(like a microphone) to wheelchair

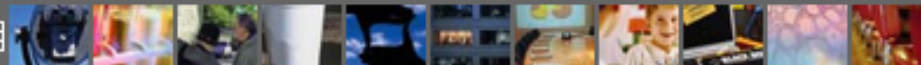
- Tremendous improvement in energy level and quality of life



Matt with his Trilogy ventilator (note the angled mouthpiece)

Patrick, age 26, graduating from Pitt Law





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M. Patrick Daniels is a staff attorney in the Pittsburgh office of Buchanan Ingersoll PC. He is a member of the firm's Commercial Litigation Section.

Before joining Buchanan Ingersoll, Patrick gained experience through several legal externships while attending law school. In 2001, he worked with the Disabilities Law Project, where he drafted a brochure designed for parents of disabled pre-school children to inform them of their rights regarding daycare services. He also drafted a memorandum discussing the use of "testers" in enforcing fair housing laws.

During 2002, Patrick worked in the U.S. Attorney's Office, Western District of Pennsylvania, in the criminal division of the narcotics/violent crime section. There, he drafted memoranda discussing whether specific facts warranted a particular charge, the application of the "knock-and-announce" rule to federal agents and the use of expert testimony in narcotics cases.

In 2003, Patrick added to his experience by working with the Honorable Chief Justice Ralph Cappy in the Pennsylvania Supreme Court. He drafted allocatur reports and researched material for a speech given by the chief justice titled "Candor to the Court."

Patrick earned his J.D. degree from the University of Pittsburgh School of Law in 2004. He received an Outstanding Achievement Award, as well as the CALI Excellence for the Future Award. He also participated in the Murray S. Love Trial Moot Court. Patrick earned his B.A. degree in political science from Wright State



THANK YOU!