

Swallowing and speech in children with neuromuscular diseases and effect of NIV

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As already emphasized, NIV is preferred over invasive ventilation, especially for patients with neuromuscular or skeletal disorders who require noncontinuous ventilation because of ease of administration, preservation of upper airway function, enhanced quality of life, and lower cost. Even patients with severely weakened or paralyzed respiratory muscles whose time off the ventilator is negligible may be treated with NIV.⁷⁰ However, invasive ventilation should be considered in patients who have persistent symptomatic hypoventilation despite repeated trials of NIV. Further, patients with more rapidly progressive neuromuscular syndromes that impair upper airway function, such as the Guillain-Barré syndrome, are usually treated with invasive ventilation when ventilatory support is indicated. For all patients, the decision to switch from noninvasive to invasive ventilation should be individualized and take patient and practitioner preferences and environmental resources into account.

Make Chest 2008

Recommendation

- ▶ NIV is the primary treatment for chronic respiratory failure in thoracic disease patients with CRF.
- ▶ The most important criteria for the advent of long-term NIV are hypercapnia in combination with the typical symptoms of ventilatory insufficiency, and the reduction in quality of life.
- ▶ For symptoms of hypoventilation in the absence of hypercapnia, a somnological examination should take place.
- ▶ Patients with severe, restrictive ventilatory dysfunction in the absence of manifest hypercapnia must be closely monitored.

Windisch Pneumologie 2010

L'apparition d'un déficit ventilatoire chronique justifie la mise en place d'une ventilation mécanique au long cours destinée à suppléer partiellement ou totalement les muscles respiratoires défaillants. La méthode proposée en première intention est une ventilation non invasive (VNI). Ce type de ventilation est à distinguer des hyperinsufflations périodiques ou IPPB (*intermittent positive pressure breathing*), parfois proposées dans le cadre de la rééducation respiratoire [1].

Les indications de la VNI chez les patients atteints de MNM sont basées sur des critères établis lors de conférences de consensus et/ou sur des avis d'experts. Le critère majeur d'initiation d'une VNI est l'existence d'une hypercapnie diurne, même modérée ($\text{PaCO}_2 > 45 \text{ mmHg}$). D'autres critères, plus précoces, ont été proposés, et comportent les symptômes d'hypoventilation alvéolaire nocturne (céphalées matinales, fatigue matinale, somnolence diurne, etc.).

**=> NON INVASIVE VENTILATION
= 1st-line Treatment**

Assisted ventilation

- ▶ Children with NMW resulting in symptomatic nocturnal hypoventilation or daytime hypercapnia should be supported with NIV. [C]
- ▶ A non-invasive approach should be considered in children needing daytime ventilation. [D]
- ▶ Clinical teams caring for children using home ventilators should become familiar with a small number of machines. For most children pressure-targeted machines work well and are simple to use. [√]

BTS 2015

ACCP 1998

Finder AJRCCM 2004

Conférence de consensus - Chest 1999

Conférence de consensus -

Rev Mal Respir 2006

Recommandations HAS - 2006

Make Chest 2008

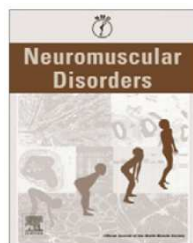
BTS - 2015

Intermittent Positive Pressure Ventilation via the Mouth as an Alternative to Tracheostomy for 257 Ventilator Users*

John R. Bach, M.D., F.C.C.P.;† Augusta S. Alba, M.D.;‡ and
Louis R. Saporito, B.A., R.R.T.

 **CHEST**™ JOURNAL
Official Publication of the American College of Chest Physicians

(*Chest* 1993; 103:174-82)



Duchenne muscular dystrophy: Survival by cardio-respiratory interventions

Yuka Ishikawa, Toshihiko Miura, Yukitoshi Ishikawa, Tomoyuki Aoyagi, Hitoko Ogata,
Satoshi Hamada, Ryoji Minami

Neuromuscular Disorders 21 (2011) 47–51

Eur Respir J 2006; 28: 549–555
DOI: 10.1183/09031936.06.00004906
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EUROPEAN RESPIRATORY *journal*
OFFICIAL SCIENTIFIC JOURNAL OF THE ERS

Diurnal ventilation *via* mouthpiece: survival in end-stage Duchenne patients

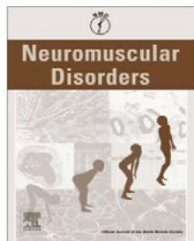
M. Toussaint, M. Steens, G. Wasteels and P. Soudon

**RESPIRATORY
CARE**

Full-Time Noninvasive Ventilation: Possible and Desirable

Joshua O Benditt MD

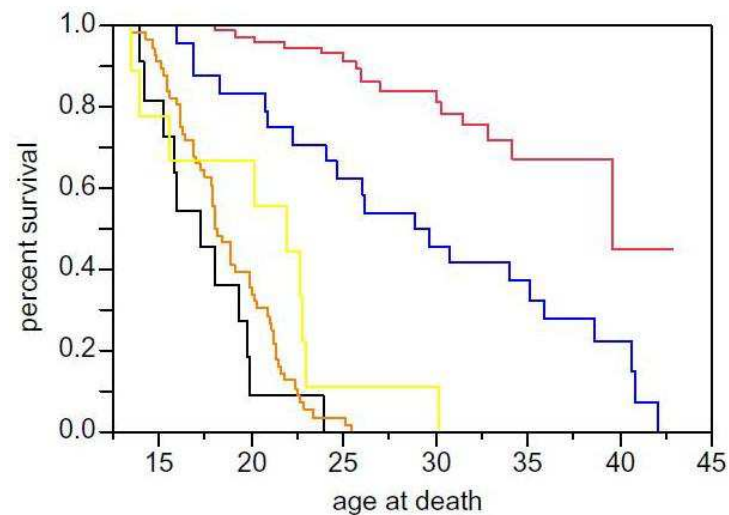
Respir Care (2006) 1005-15



Duchenne muscular dystrophy: Survival by cardio-respiratory interventions

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Neuromuscular Disorders 21 (2011) 47–51



- NIV from 1991 -> survival 39.6 ans
 - Tracheotomy from 1984 to 1991 -> survival 28.9 ans
 - No ventilation and dead after 1991
 - No ventilation and dead before 1984
 - No ventilation and dead from 1984 to 1991
- > survival 18.1 ans

Breathing and Swallowing Interaction

Normal Subject



Several successive phases with a critical and complex neurological control (cortex and TC)

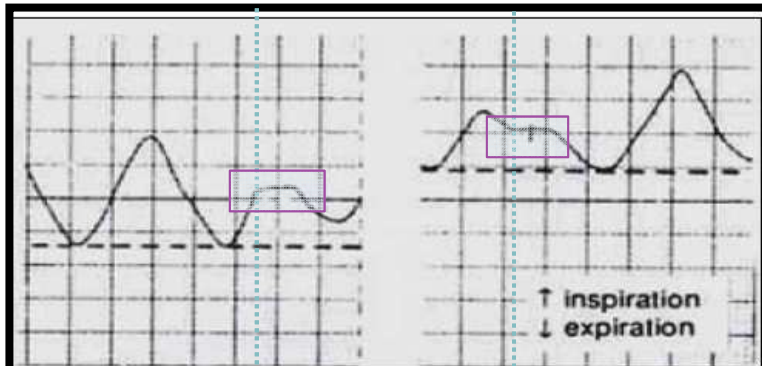
⇒ Critical phase: Oro-pharyngeal

Both voluntary and reflex event with modifications of the respiratory cycle

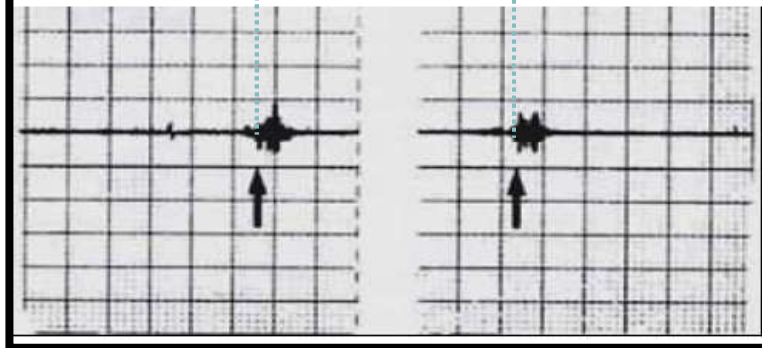
Breathing and swallowing Interaction

Normal Subject

Pletysomno.



EMG



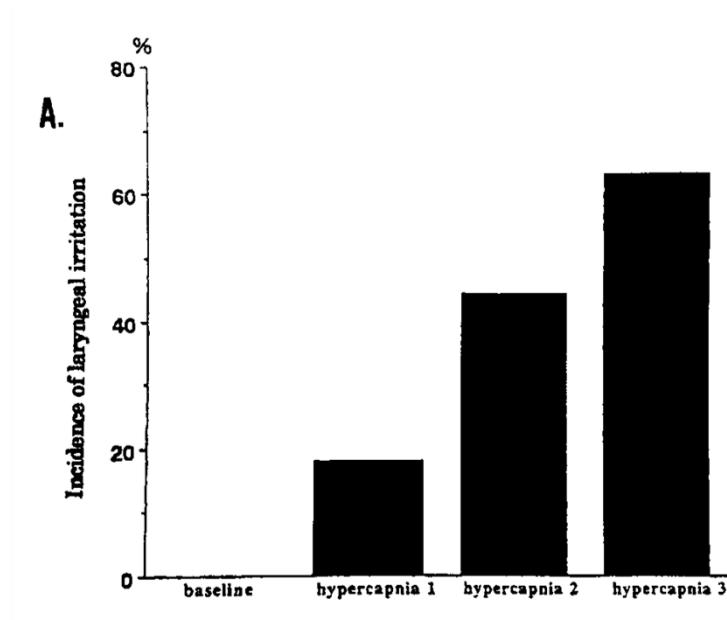
Interruption of respiratory cycle

=> Swallowing apnea

Breathing resumes at the end of inspiration **or** during expiration

Swallowing and respiratory failure

- Hypercapnia increases laryngeal inspiration



Nishino, AJRCCM, 1998

Swallowing and respiratory failure

- Respiratory failure in NM disorders and in COPD may be associated with swallowing disorders and breathing swallowing interactions dysfunction

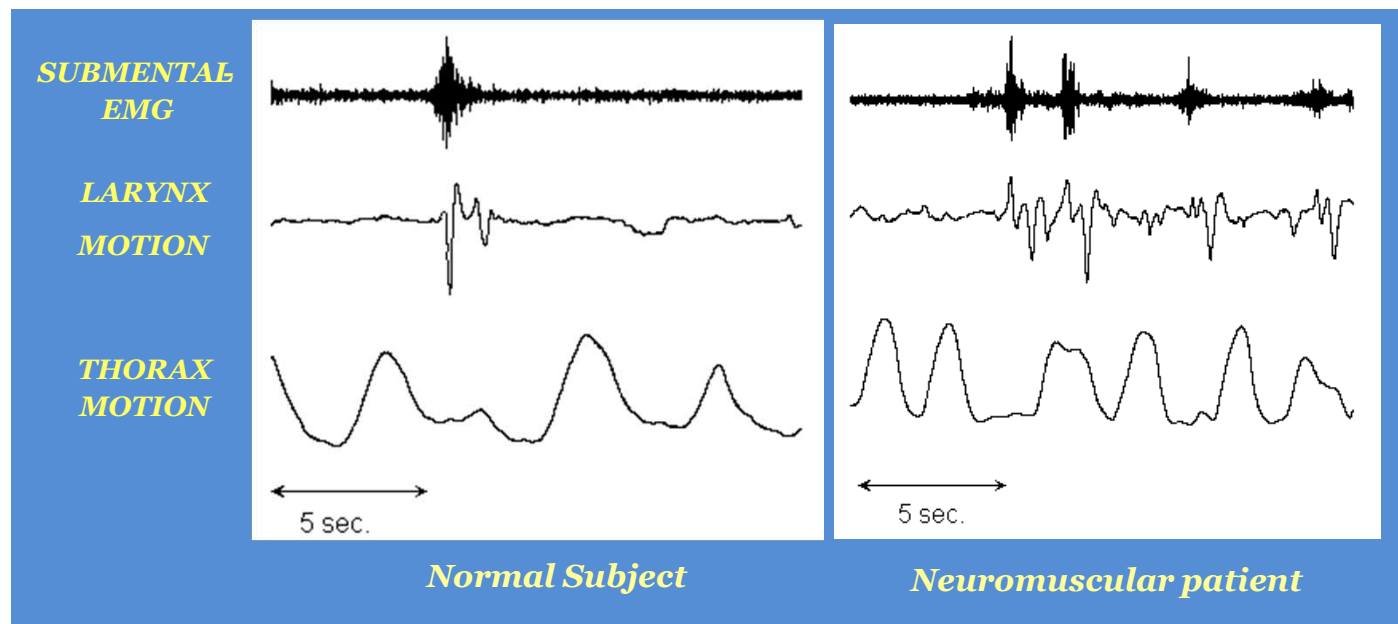
Shaker et al, American Journal of Physiol 1992

Terzi et al, AJRCCM 2007

Gross et al, AJRCCM 2009

Terzi et al, Neuromuscul Disord 2010

- In NM disorders



Terzi et al, AJRCCM 2007

Swallowing and respiratory failure

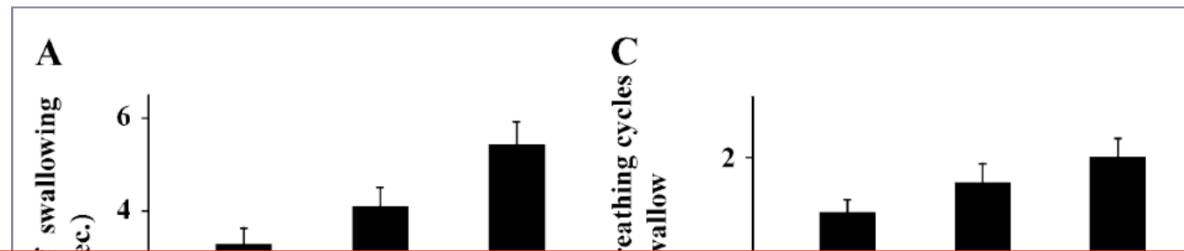
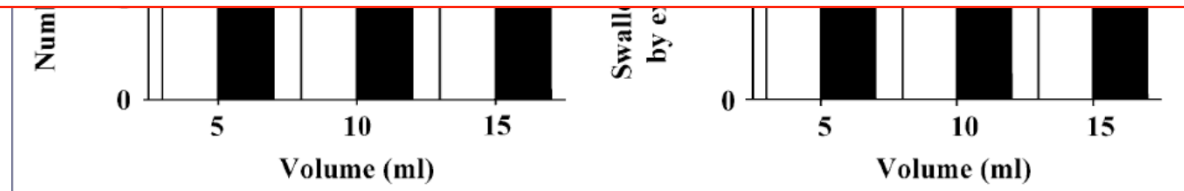


TABLE 3. UNIVARIATE REGRESSION ANALYSIS OF SWALLOWING VARIABLES ON INDICES OF LEVEL OF DISABILITY

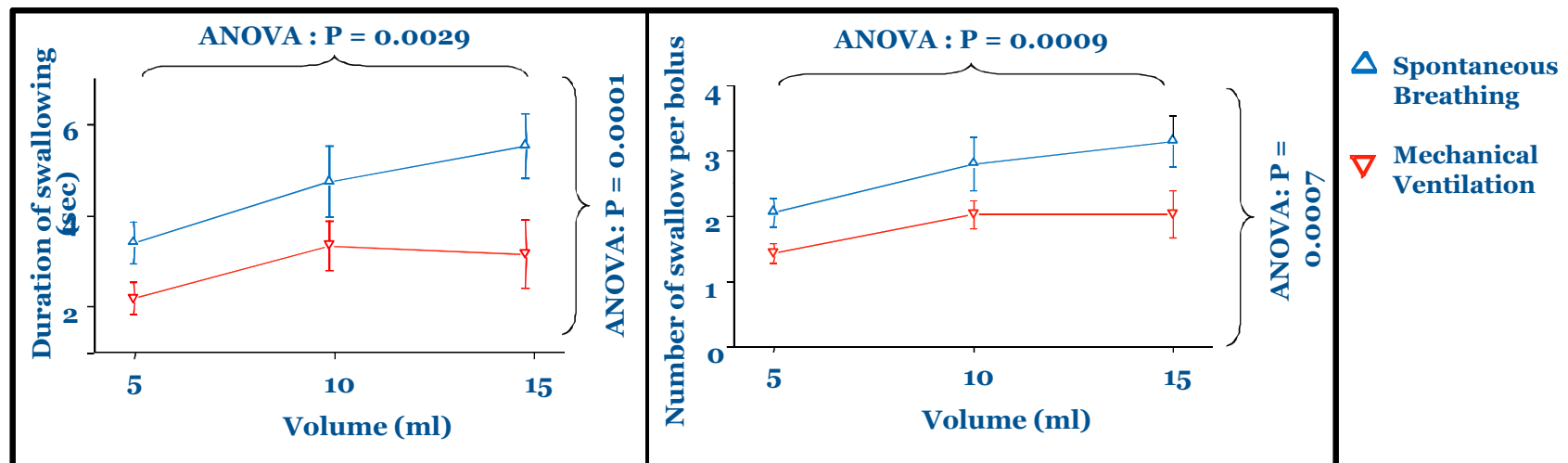
	Duration of Swallowing			Number of Swallows			Number of Breathing Cycles		
	Coefficient	R ²	p Value	Coefficient	R ²	p Value	Coefficient	R ²	p Value
VC	-0.23	0.05	0.22	-0.22	0.05	0.24	-0.39	0.15	0.03
MIP	-0.47	0.22	0.01	-0.56	0.30	0.002	-0.55	0.30	0.002
MEP	-0.43	0.19	0.02	-0.43	0.18	0.02	-0.53	0.28	0.004
Pa _{CO} ₂	0.22	0.048	0.29	0.181	0.03	0.39	0.24	0.05	0.25
AI	0.26	0.068	0.17	0.241	0.05	0.20	0.35	0.12	0.06
Dysphagia	0.20	0.04	0.30	0.13	0.018	0.48	0.11	0.012	0.56



Comparing Normal Subjects - NMD

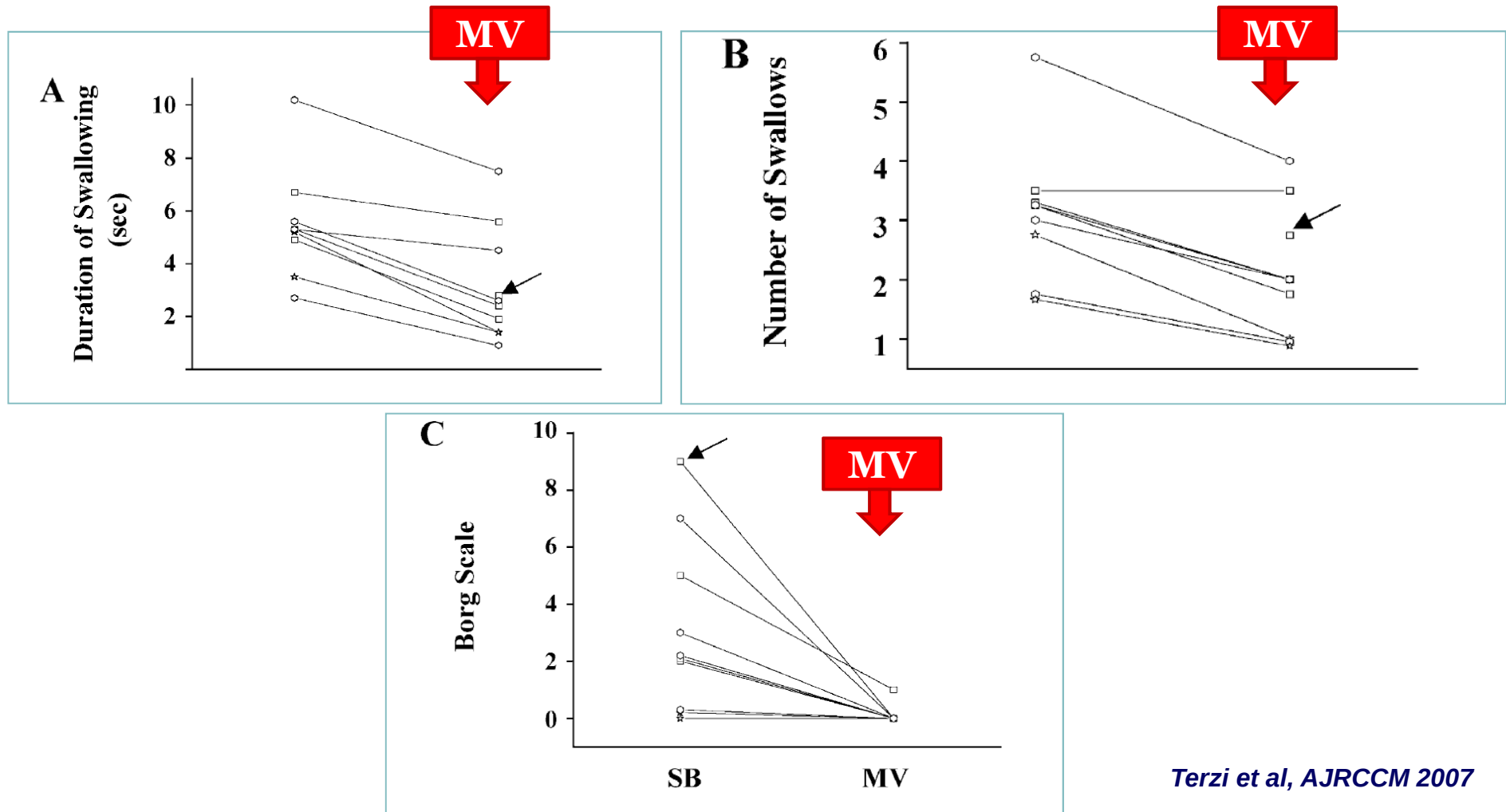
Swallowing and respiratory failure and ventilation

- Tracheostomized NM patients improve their swallowing parameters improve when swallowing **while ventilated**

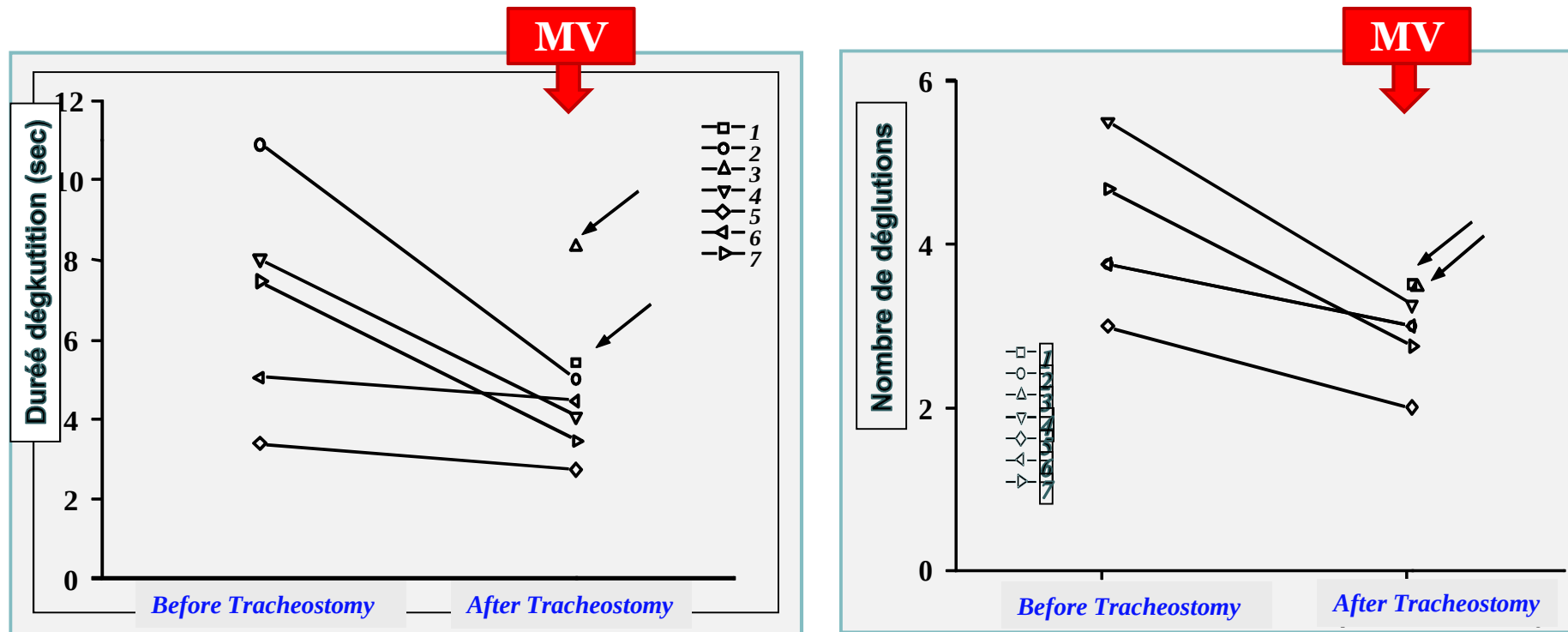


Swallowing and respiratory failure and ventilation

=> 10 tracheostomized subjects



Swallowing and respiratory failure and ventilation



=> Reduction of swallowing fragmentation **after** tracheostomy **while** ventilated

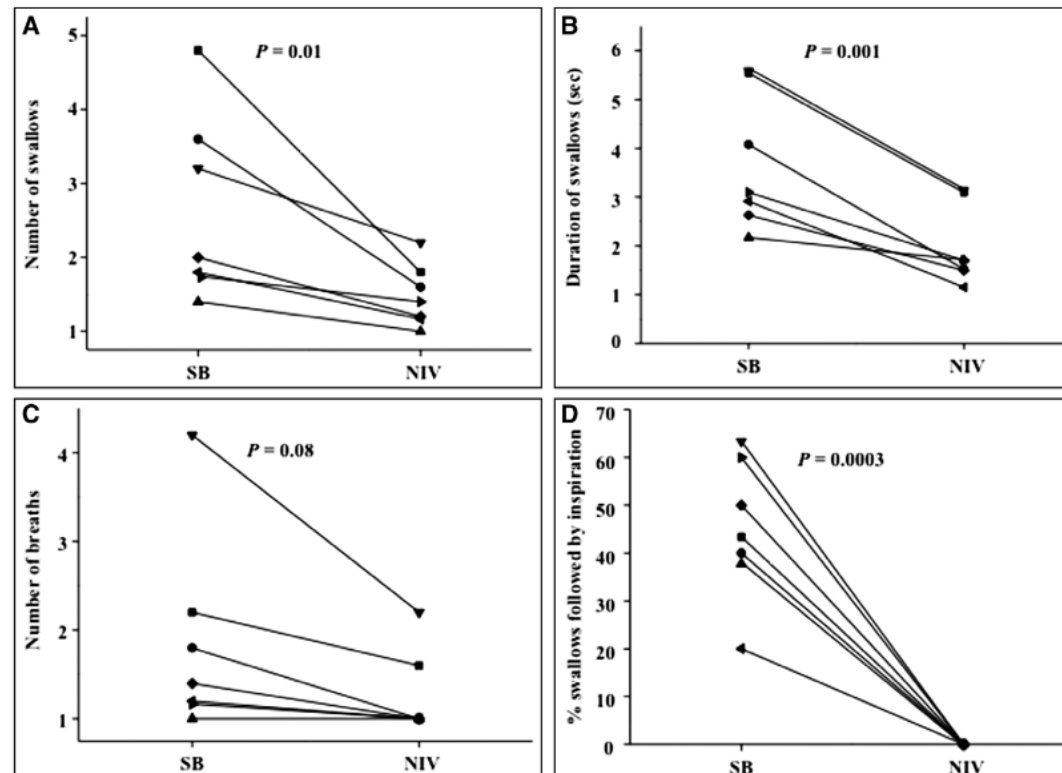


Swallowing and respiratory failure and ventilation

Can NIV use improve breathing-swallowing interactions in NM patients with severe respiratory failure?

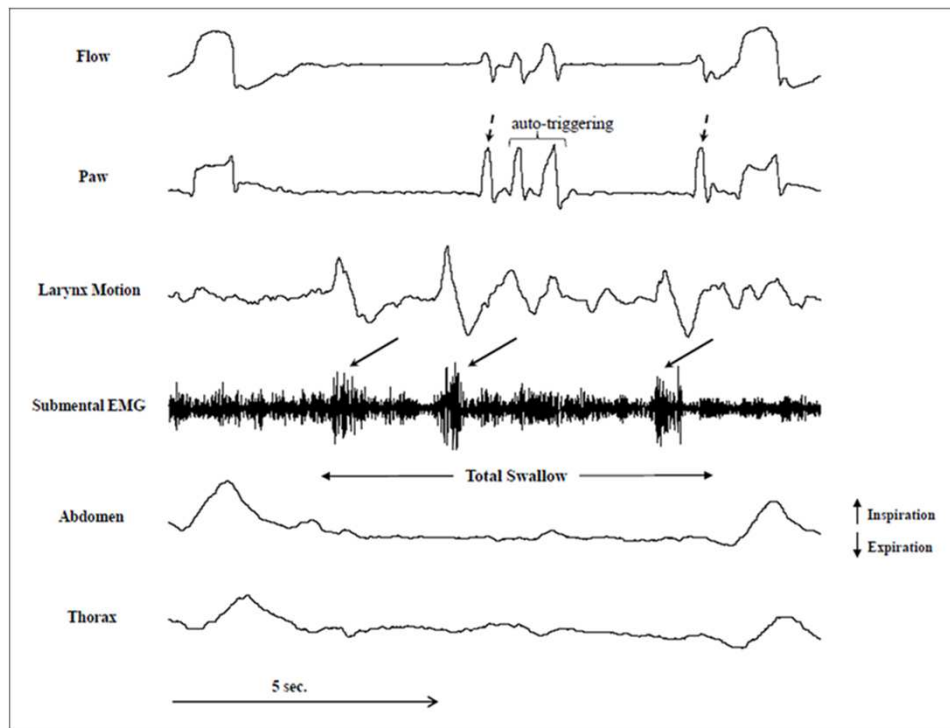
Swallowing and NIV

- **COPD** patients, during **acute** respiratory failure, improve breathing-swallowing interactions under NIV

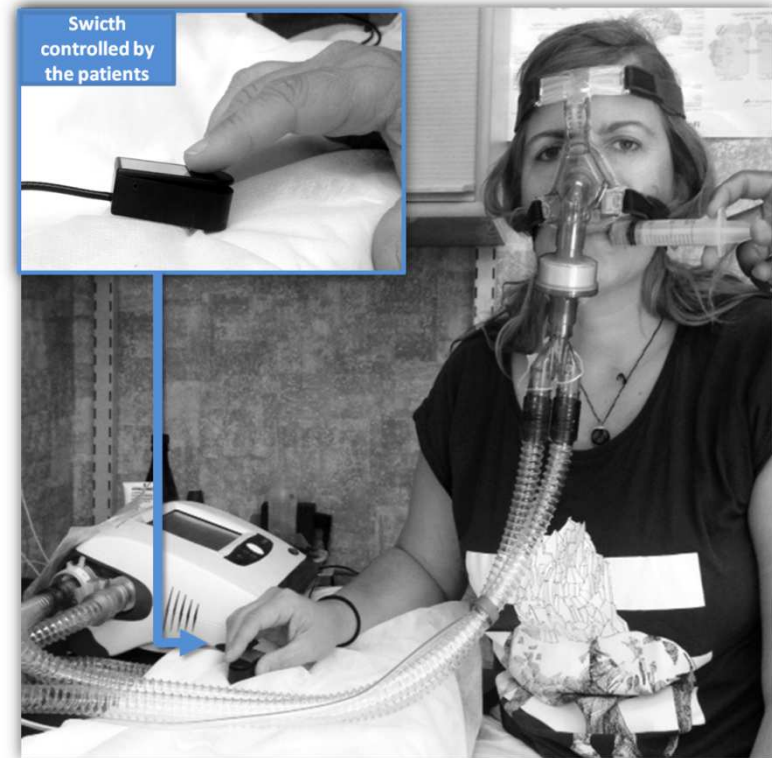


Swallowing and NIV

- **NMD**



Terzi et al, CCM 2014

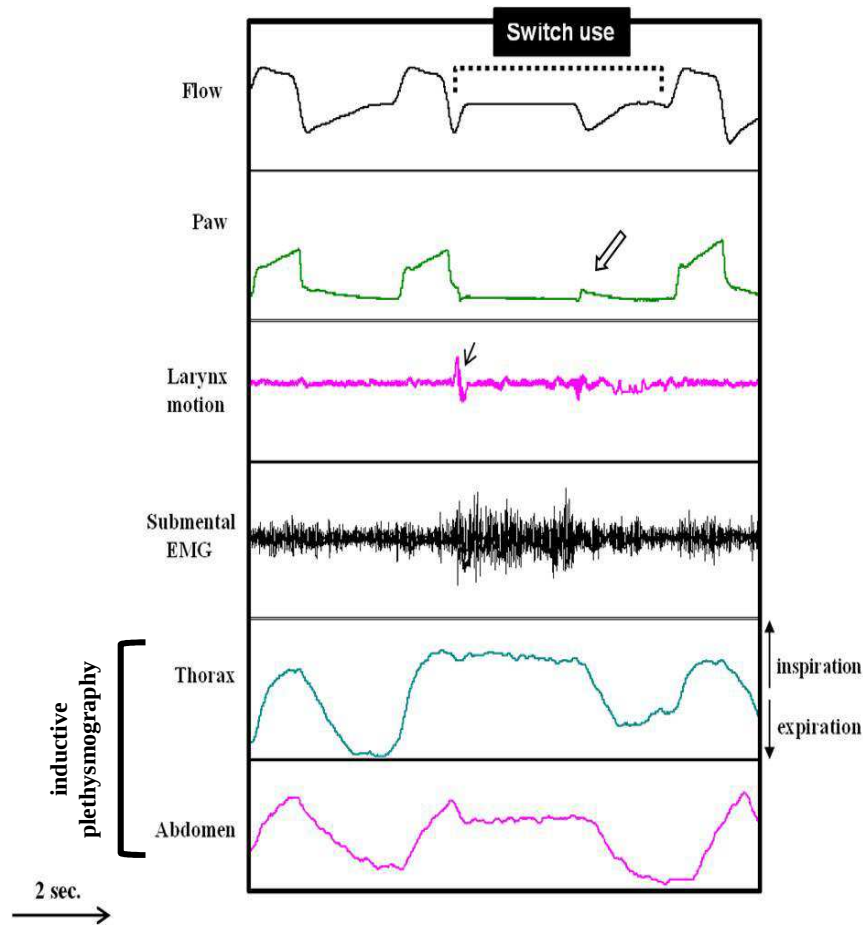


Modified home ventilator

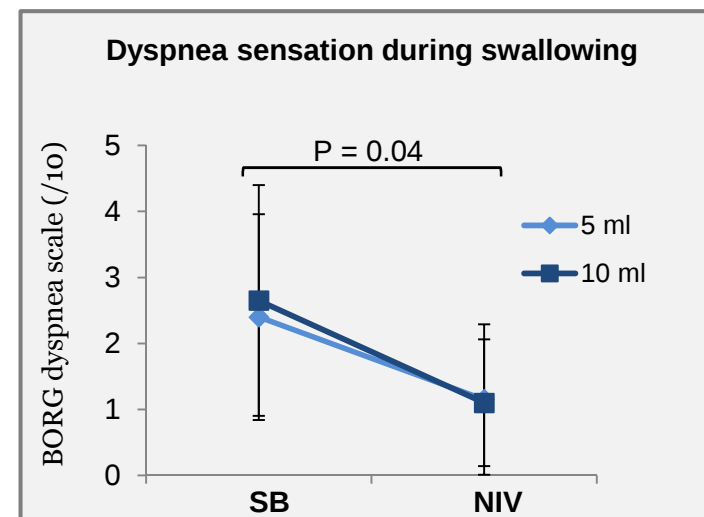
(Elysée 150, ResMed, San Diego, USA)

⇒ Switch activation withholds ventilation

Swallowing and NIV



- All patients found the device useful
- No episode of aspiration
- No episode of auto-triggering with device
- Swallowing comfort stable under NIV



Swallowing and NIV

	5 ml-bolus		10 ml-bolus		Yogurt		ANOVA (p)		
	SB	NIV	SB	NIV	SB	NIV	Size effect	Texture effect	NIV effect
Number of swallows (per bolus)	2.0±0.9	2.8±1.4	2.3±1.3	2.8±1.2	2.6 ±0.9	2.4± 1.1	0.4	0.6	0.07
Duration of swallowing (sec)	5.4±4.6	4.6±3.4	7.1±4.5	5.9±3.4	7.1±4.9	5.8±4.2	0.04	0.1	0.1
Swallowing fragmentation (respiratory events per bolus)	1.6±1.8	0.8±1.0	2.3±1.7	1.0±1.4	1.9±1.5	1.1±1.0	0.03	0.3	0.003
% of swallows followed by an inspiration	43.5±23.3	10.3±7.7	46.1±23.6	17.9±19.5	45.7±21.5	21.1 ±16.4	0.2	0.08	<0.0001

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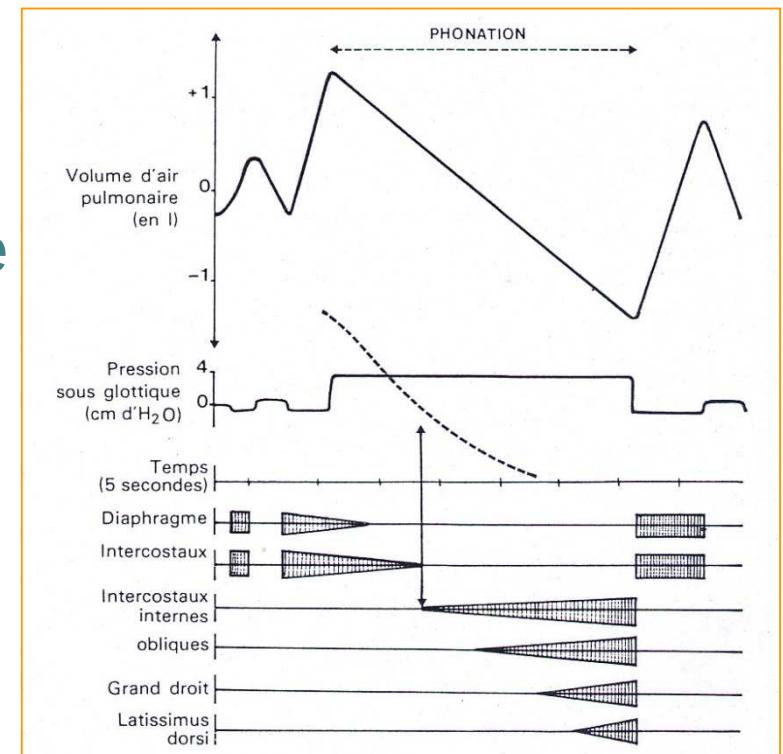
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% of swallows followed by an inspiration	<u>3.5±23.3</u>	10.3±7.7	<u>46.1±23.6</u>	17.9±19.5	<u>45.7±21.5</u>	21.1 ±16.4	0.2	0.08	<0.0001

Phonation

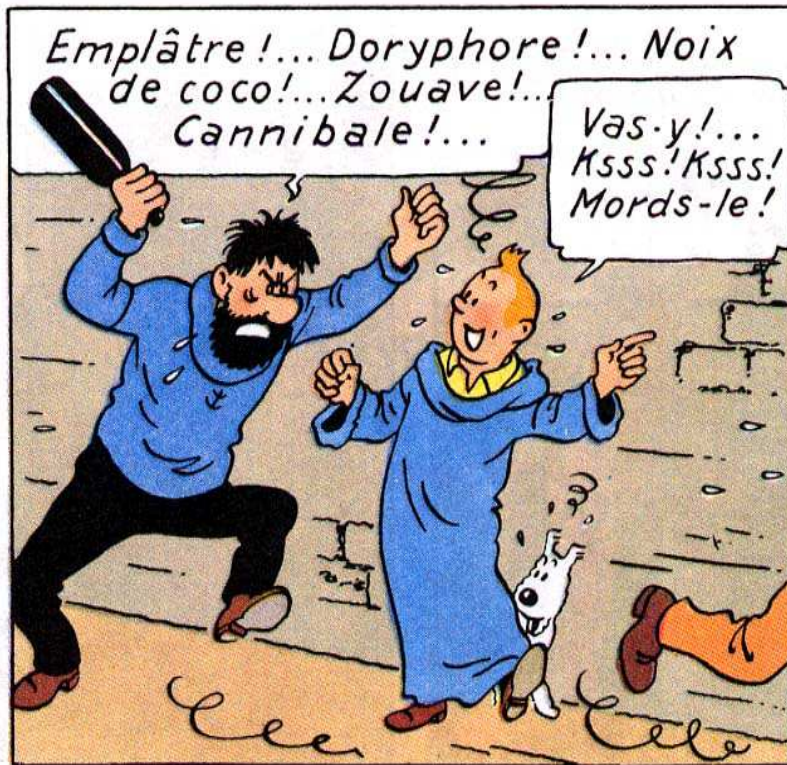
- Requires precise neuro-motor coordination : *laryngeal, pharyngo-bucco-labial, respiratory, postural muscles.*
- Exemple of **voluntary** control of ventilation
- Modification of the respiratory cycle and of respiratory muscles involvement in order to maintain the desired vocal production

Draper, 1959



Phonation and breathing interactions

Normal Subject



- **Expiratory event**
- $\uparrow$ RR ($TI \downarrow$ et $TE \uparrow \uparrow$)
- $\uparrow$ tidal volume
- $\Rightarrow \uparrow$ minute ventilation
- Alveolar Hyperventilation

Phonation and NMD

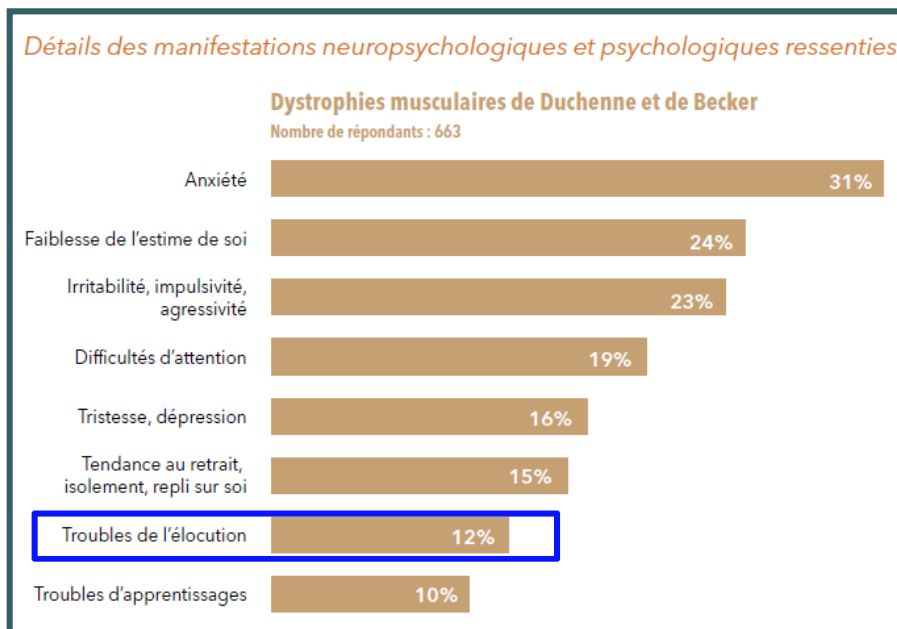
- NMD may impair phonation quality:

- UAW muscle dysfunction
- Facial muscle failure
- Macroglossia

⇒ articulation difficulties
⇒ poor intelligibility

- Respiratory failure?...

Phonation and NMD



- SCI patients: difficulties to control loudness

Draper et al, BMJ, 1960

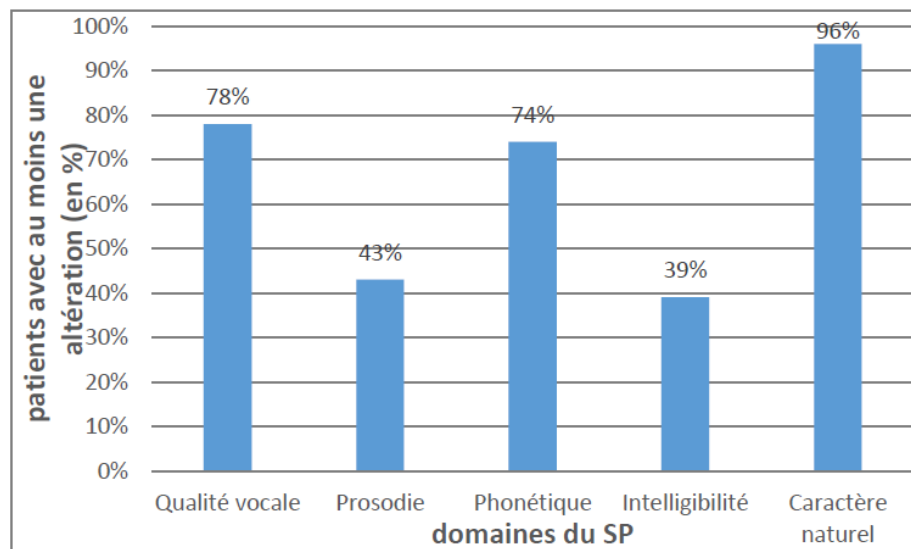
- In NMDs:
 - Decreased loudness and pitch
 - Shortness of breath and speaking-related dyspnea

Britton et al, Semin Speech Lang, 2016

Laakso et al, Int J Lang Commun Disord, 2011

Voice quality and NMD

- 27 Duchenne and Becker dystrophies studied during speech in natural breathing
- ⇒ VHI > 0 for 26/27, significantly altered in 26%
- ⇒ Deterioration of perception score:

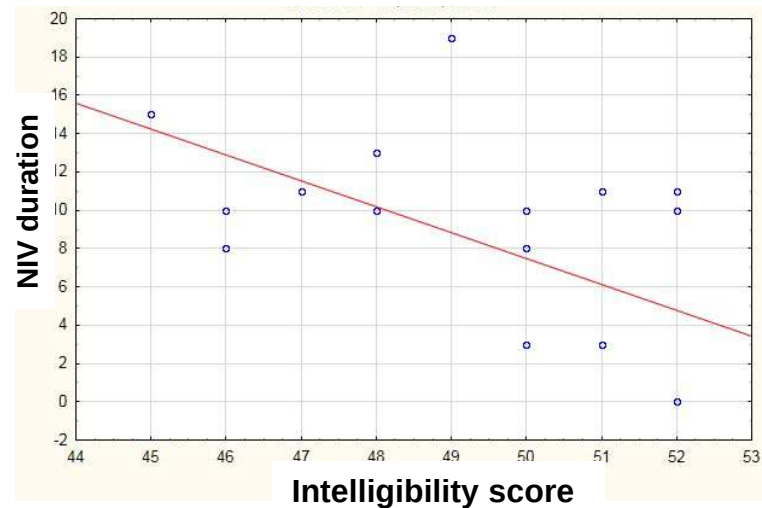
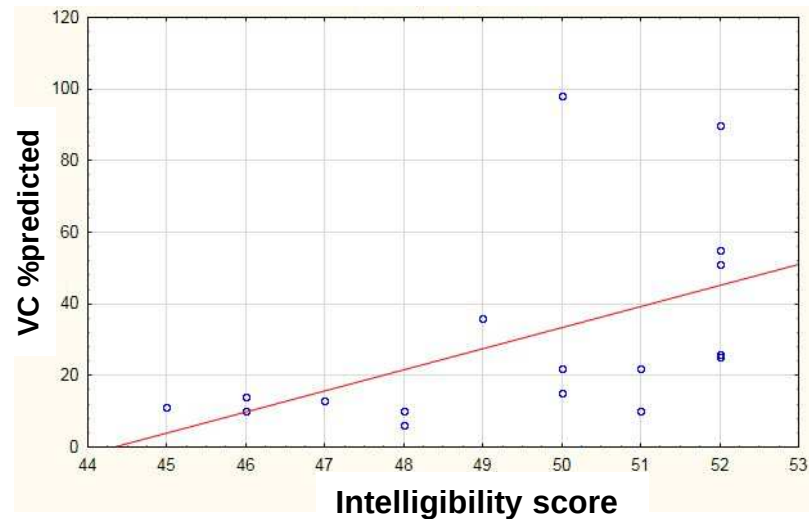


Personal data

Voice quality and respiratory failure

- 19 Duchenne patients' speech during spontaneous breathing

⇒ Inverse correlation between intelligibility and respiratory failure severity



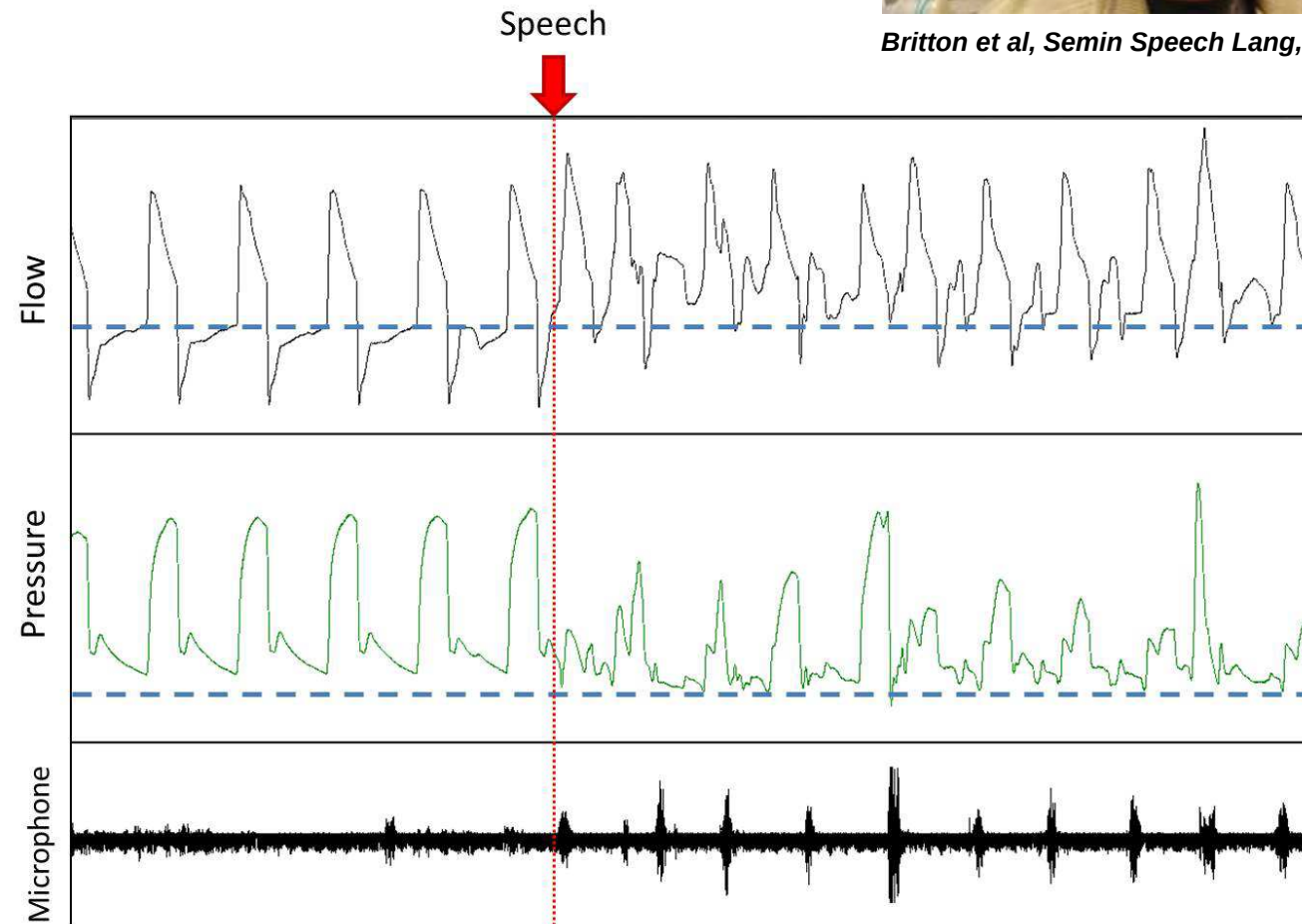
Personal data

Voice quality and NIV

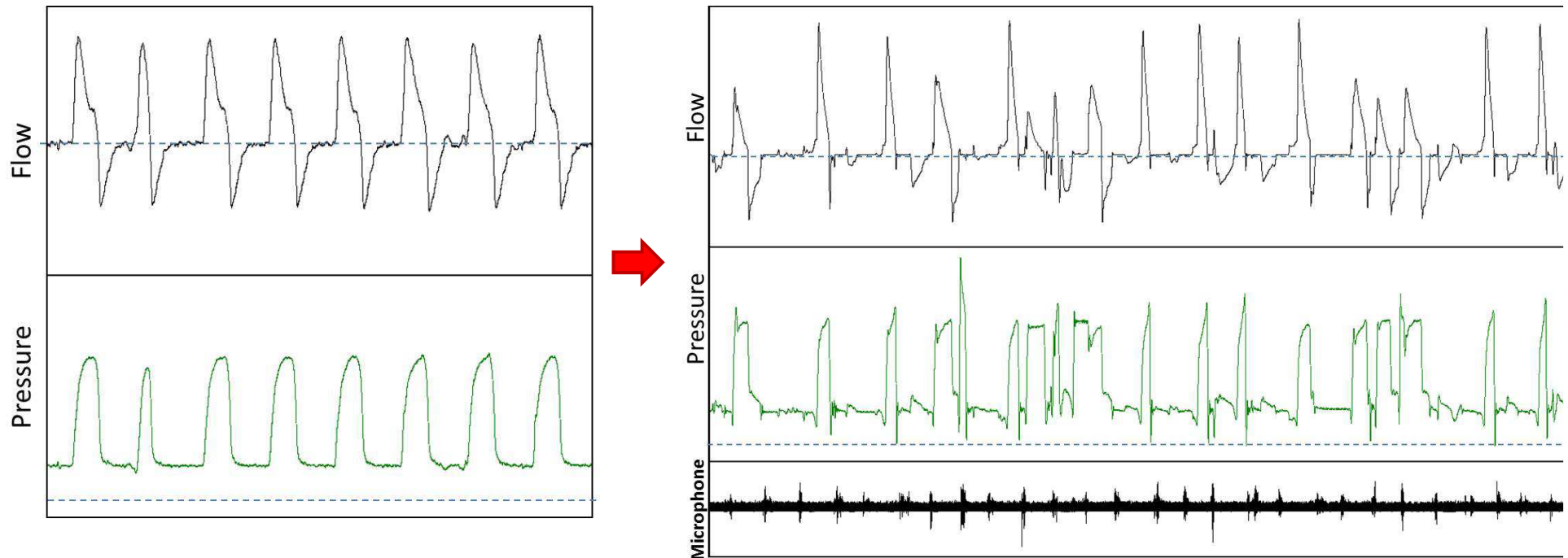


Britton et al, Semin Speech Lang, 2016

Nasal Mask



Voice quality and NIV



« It's like talking with someone plugging your nose »

With pressure controlled ventilation:

« When I try to talk, the air is leaking out of my mouth »

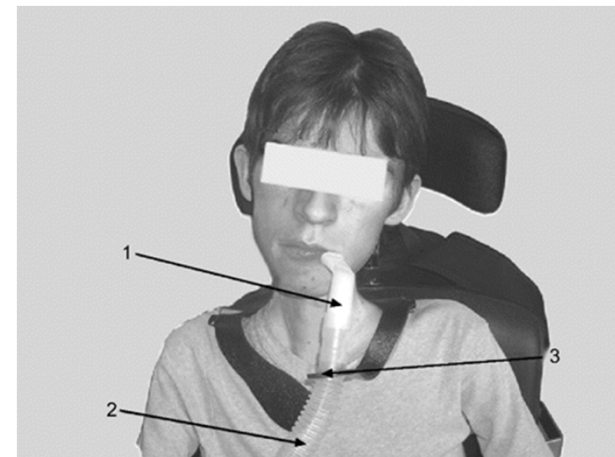
Voice quality and NIV

- Adaptation of the interface for optimized phonation

⇒ Mouthpiece ventilation



Britton et al, Semin Speech Lang, 2016



*Michel Toussaint's team
Belgium (ERJ 2006)*



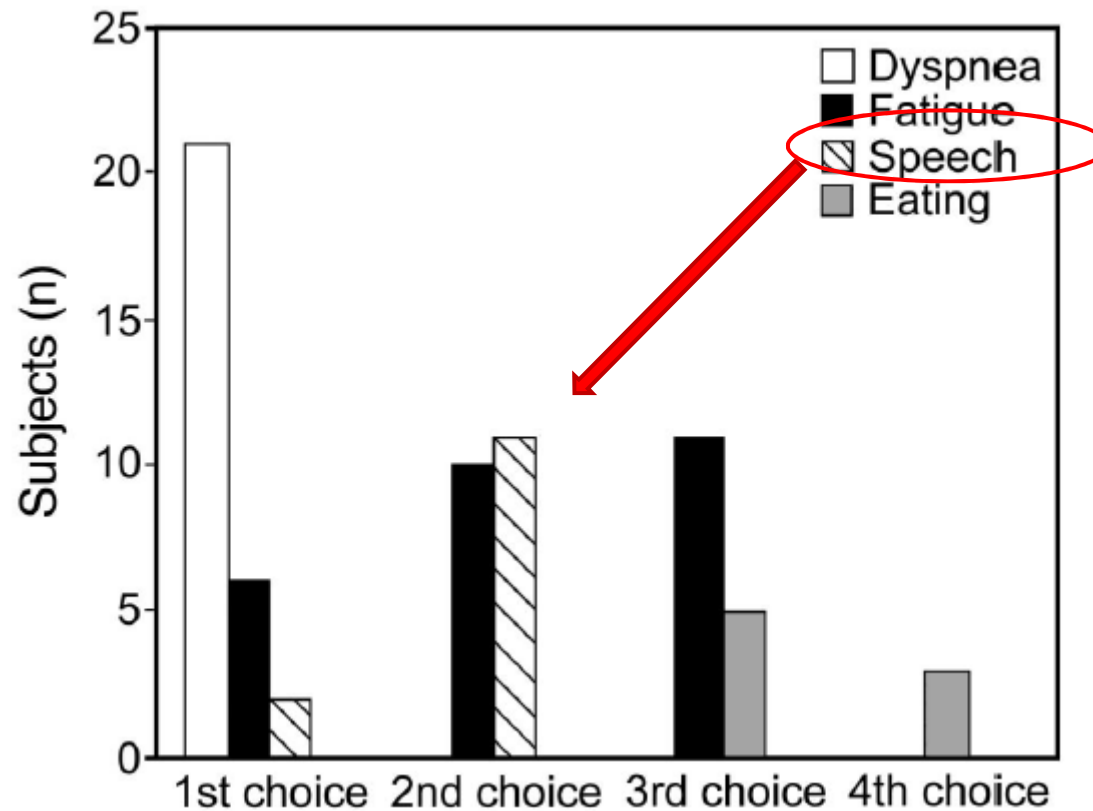
Voice quality and NIV: interface choice

Mouthpiece ventilation

- (1) less negative impact on the patient
- (2) no risk of skin breakdown
- (3) facilitates speech
- (4) facilitates eating and swallowing
- (5) better appearance and
- (6) is safer by permitting use of glossopharyngeal breathing in the event of sudden ventilator failure or accidental disconnection from the ventilator.

Voice quality and NIV: interface choice

Mouthpiece ventilation



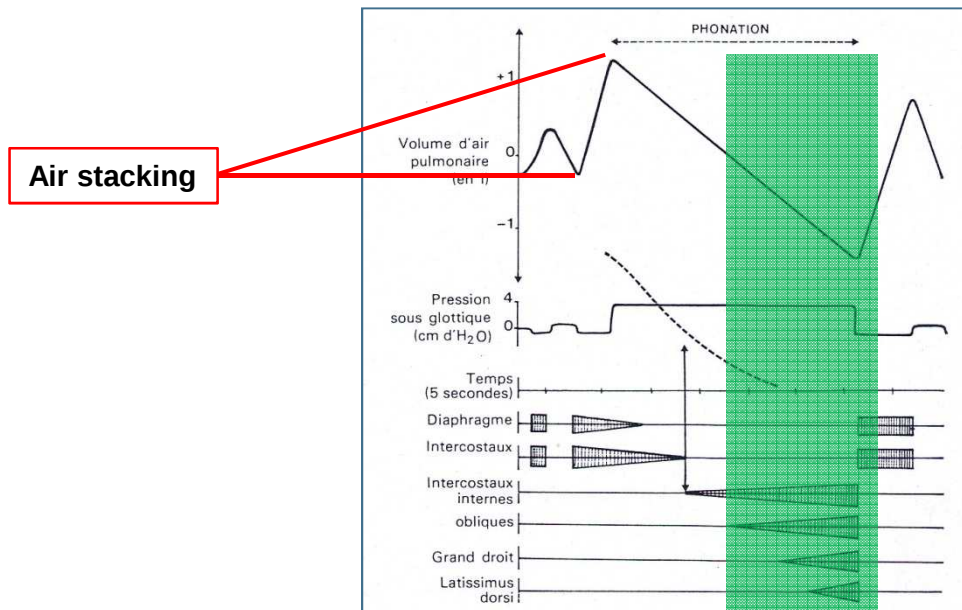
Followed by :

- 5) better vision,
- 6) reduction in skin injury,
- 7) facilitation of swallowing,
- 8) decrease of aerophagia

Voice quality and NIV: interface choice

Mouthpiece ventilation

- With volume controlled-ventilation, patients can use « breath-stacking » to increase loudness.

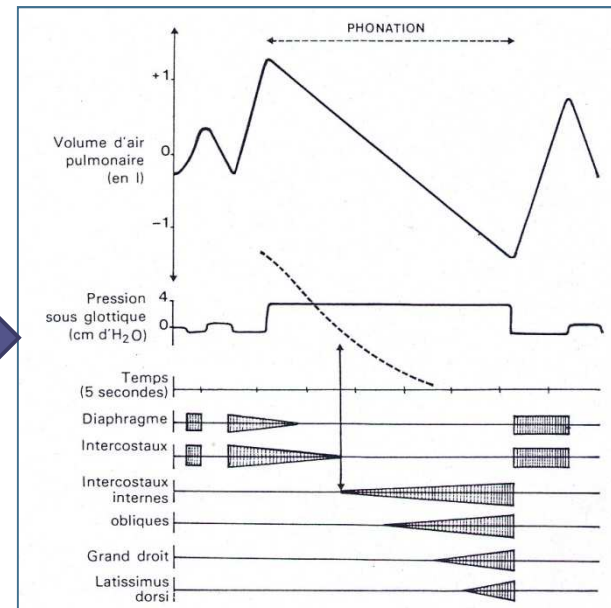


Britton et al, Semin Speech Lang, 2016

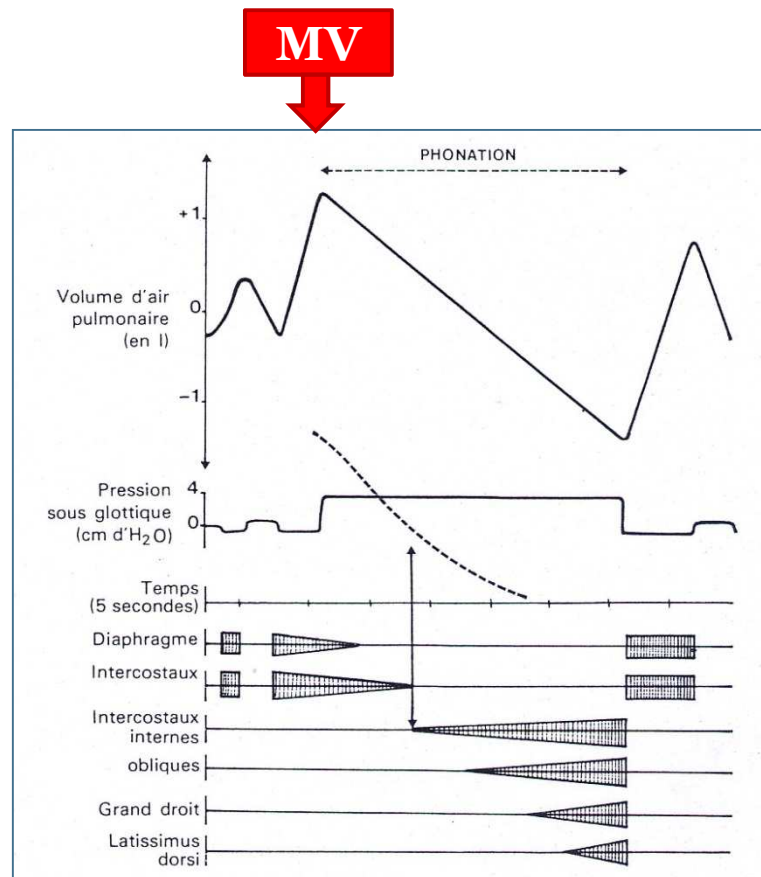
Voice quality and NIV:

- But with mouthpiece ventilation:

Speech occurs during spontaneous breathing **with** severe respiratory failure



Voice quality and NIV



Unpublished data

Voice quality and NIV

- 8 NMD patients tested under NIV while speaking
- No improvement:
 - Speech parameters
 - Respiratory and speech comfort

?

- **Interaction of nasal mask with voice quality**
- **Insufficient use of NIV for speech support**
- **Patients too severe to manage the increased inspiratory volume**



Conclusion

- Swallowing may improve with NIV in NMD patients with severe respiratory failure
- Mouthpiece ventilation ensures independant speech as patients may discontinue ventilation
- Speech may be altered by respiratory failure but NIV does not yet provide support for quality improvement

Thank you for your attention



Hergé, 1960