

Con il patrocinio di



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PNEUMOLOGIA 2016

Milano, 16 – 18 giugno 2016 · Centro Congressi Palazzo delle Stelline

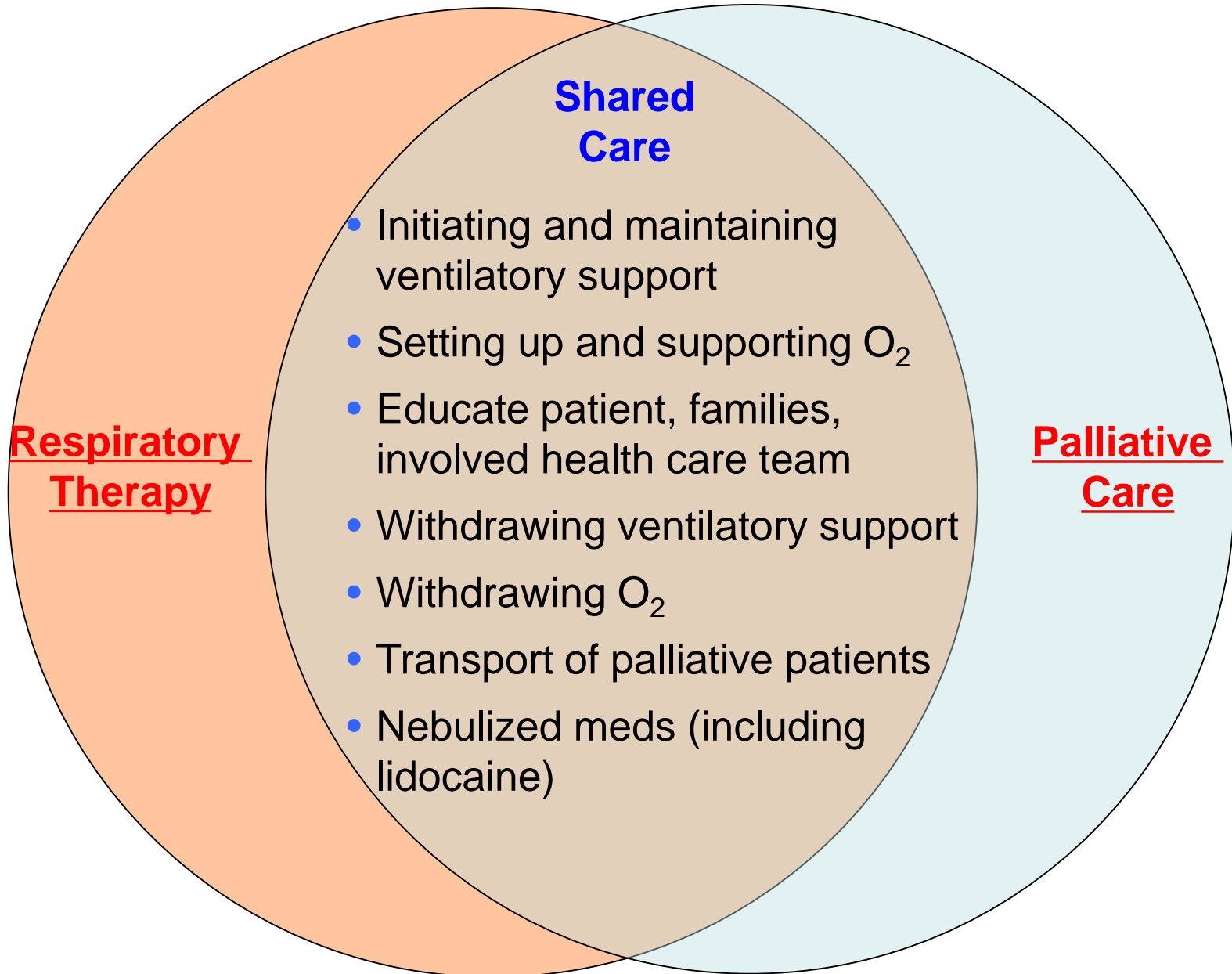
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La Ventilazione Non Invasiva può avere un ruolo nella palliazione?

Andrea Vianello
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Palliative Care is an **approach to care** which focuses on comfort and quality of life for those affected by life-limiting/life-threatening illness. Its goal is **much more than comfort in dying**; palliative care is about **living**, through meticulous attention to control of pain and other symptoms, supporting emotional, spiritual, and cultural needs, and maximizing functional status.



Shared care

1. Role of O₂ therapy
2. Noninvasive ventilation
3. Role of opioids in palliation of dyspnea
4. Transport of dying patients
5. Withdrawal of life-sustaining treatments – ventilatory support (invasive and noninvasive), oxygen

Shared care

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Use of Noninvasive Ventilation

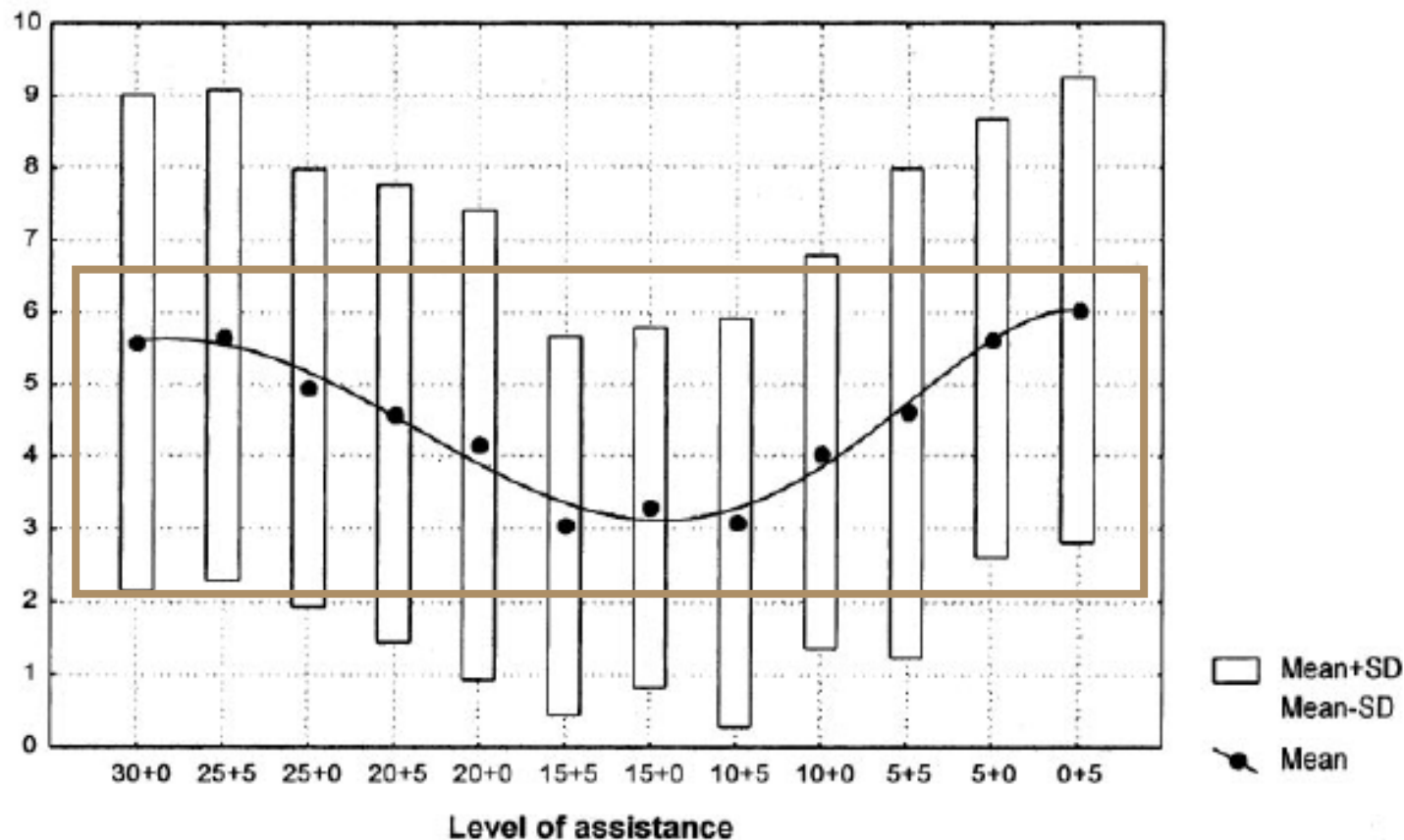
- Classified use of NIV for ARF into 3 categories:
 1. NIV as life support with no preset limitations on life sustaining treatments
 2. NIV as life support when patients/families have decided to forego ETT
 3. NIV as a palliative measure when patients/families have decided to forego all life support, receiving symptom management only to be comfortable

Use of Noninvasive Ventilation

- Discussing goals of using NPPV....
- Category 1:
 - goal is to restore health; will use intubation if necessary and indicated
- Category 2:
 - Goal is to restore health without using ETT and without causing unacceptable discomfort
- Category 3:
 - Goal is to maximize comfort while minimizing adverse effects of opiates

Assessment of Physiologic Variables and Subjective Comfort Under Different Levels of Pressure Support Ventilation*

Michele Vitacca, MD; Luca Bianchi, MD; Ercole Zanotti, MD;
Andrea Vianello, MD; Luca Barbano, MD; Roberto Porta, MD; and
Enrico Clini, MD, FCCP†



Advantages of Palliative NIV

- Symptom relief:
 - Decreased work WOB
 - Decreased dyspnea
- Buys time:
 - Allows family members to arrive
 - Allows for individuals to cope with the deterioration
- Improved level of consciousness:
 - Preserved communication between patient and family

Evidence and Behavior for Palliative NIV Use

- **Nava S et al. Eur Resp J 2007:** European survey, 40% of NIV was solely as palliative treatment
- **Sinuff T et al. Crit Care Med 2008:** North American survey, attitudes of MDs and RTs re use of NIV for ptns with ARF with DNI/DNR order; 62% MDs and 87% RTs included the potential use of NIV in their discussions
- >80 % used NIV for COPD and CPO patients with DNR and nearing end of life; 59% for underlying cancer

Observational Studies

- **Levy M et al. Crit Care Med 2004; 32**
- Recent multi-centre study; 43% survived and discharged from ICU
- ~50 % with COPD and 70% with CPO surviving at hospital discharge
- Better hospital survival if:
 - ✓ Higher level of baseline hypercapnia
 - ✓ Dx of COPD or CPO
 - ✓ Presence of strong cough and wakefulness

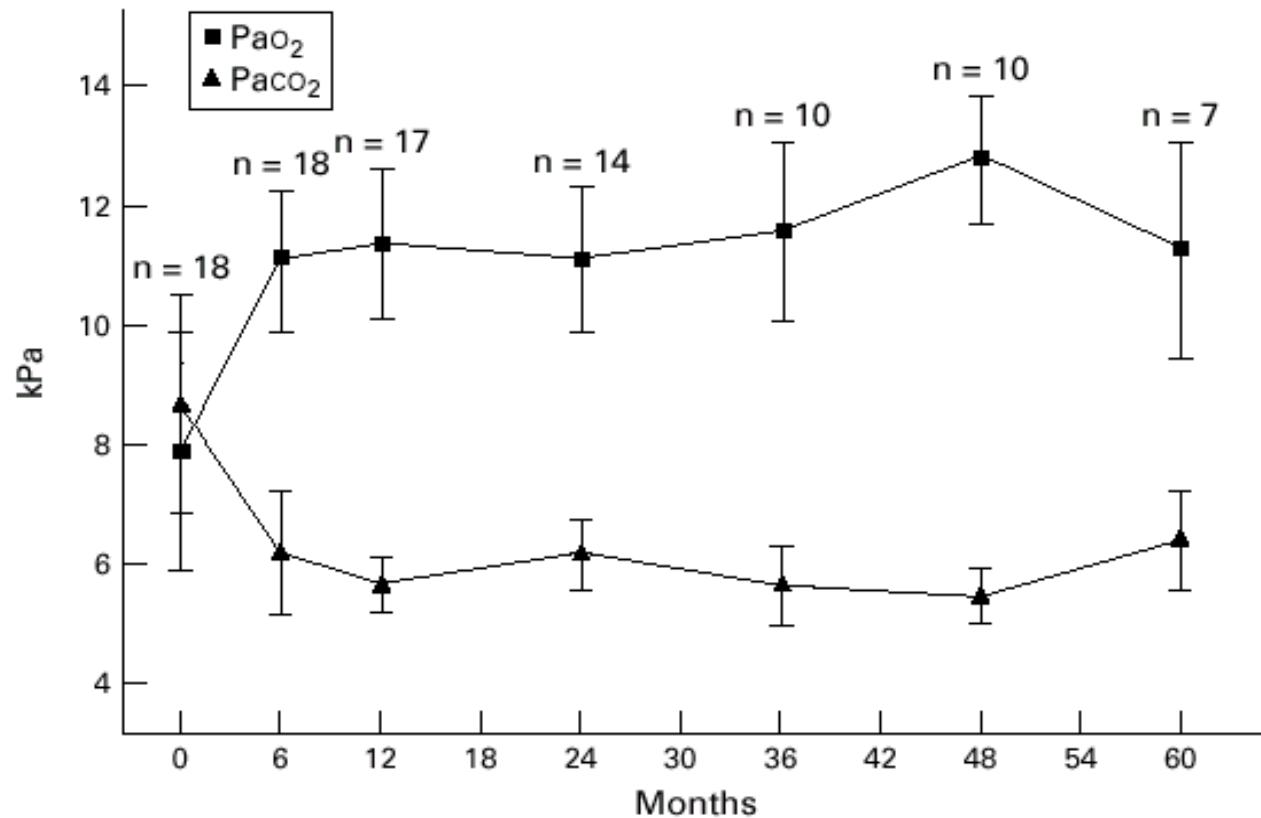
- **Schettino G et al. Crit Care Med 2005;33**
- Single-centre prospective observational study
131 patients
- Overall hospital mortality of 65%, poorer prognosis in those with advanced cancer (85%)
- COPD 63% & CPO 60% hospital survival rates

Underlying Respiratory Diseases

- Neuromuscular diseases and neurological disorders which impair respiratory function
- Respiratory complications produce burdensome symptoms
- NIV is very helpful in the early phases
 - ✓ Improves survival
 - ✓ Improves QOL
 - ✓ Decreases respiratory symptom burden
- Progressive disease on home NIV....almost all continue tx even in terminal stages....eventually tracheostomy and home invasive ventilation

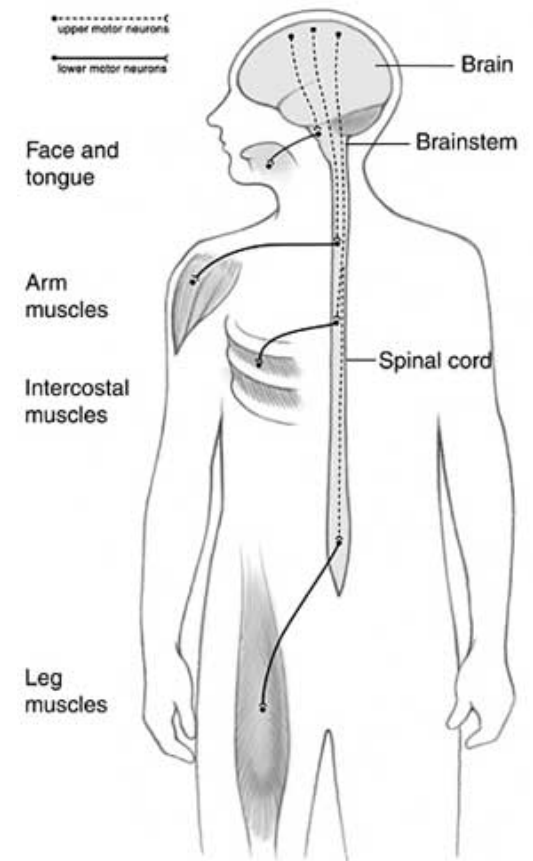
Impact of nasal ventilation on survival in hypercapnic Duchenne muscular dystrophy

A K Simonds, F Muntoni, S Heather, S Fielding



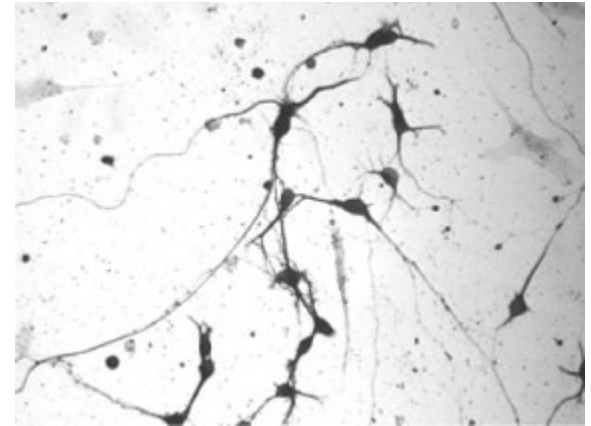
What is amyotrophic lateral sclerosis?

- It is a progressive neurological disease that affects the control of muscle movement due to its damaging effects on motor neurons in the spinal cord and the brain



Symptoms of ALS

- First signs and symptoms (frequently overlooked)
 - Twitching and cramping of muscles (especially in hands and feet)
 - Stiffness
 - Weakness (especially in hands, arms and legs)
 - Slurred speech



Picture taken from the National Institute of Aging

Table 8 Symptoms and signs of respiratory insufficiency in amyotrophic lateral sclerosis

Symptoms	Signs
Dyspnoea on minor exertion or talking	Tachypnoea
Orthopnoea	Use of auxilliary respiratory muscles
Frequent nocturnal awakenings	Paradoxical movement of the abdomen
Excessive daytime sleepiness	Decreased chest wall movement
Daytime fatigue	Weak cough
Morning headache	Sweating
Difficulty clearing secretions	Tachycardia
Apathy	Morning confusion, hallucinations
Poor appetite	Weight loss
Poor concentration and/or memory	Mouth dryness

Modified from Leigh *et al.* [28].

Clinical and Pulmonary Function evaluation

	ALS	NMD	
Symptoms resulting in the initial referral to pulmonary unit			
→ Difficulty clearing respiratory secretions	77.4	6	< .001
→ Dyspnea on exertion	77.4	68.9	.11
→ Disturbed sleep	58.5	83.6	< .001
Extreme fatigue	52.8	59	.28
Nocturnal awakenings	13.2	32.8	< .001
Irritability	9.4	19.7	.03
Excessive daytime sleepiness	9.4	19.7	.03
Difficulty in concentrating	9.4	21.3	.01
Impaired cognition	1.8	24.6	.002
Pulmonary function test performed at the initial evaluation			
Vital capacity sitting	98.1	91.9	.05
Blood gas analysis	98.1	95.2	.24
Nocturnal oximetry	88.7	83.9	.26
Maximum inspiratory/expiratory pressure	86.8	79	.10
Cardiorespiratory monitoring/polysomnography	64.2	74.2	.04
Peak cough flow	56.6	58.1	.79
Total lung capacity and inspiratory capacity	54.7	54.8	.98
Vital capacity lying	47.2	56.5	.10
Sniff inspiratory pressure	15.1	17.7	.55

Cause of death in 302 French patients with ALS

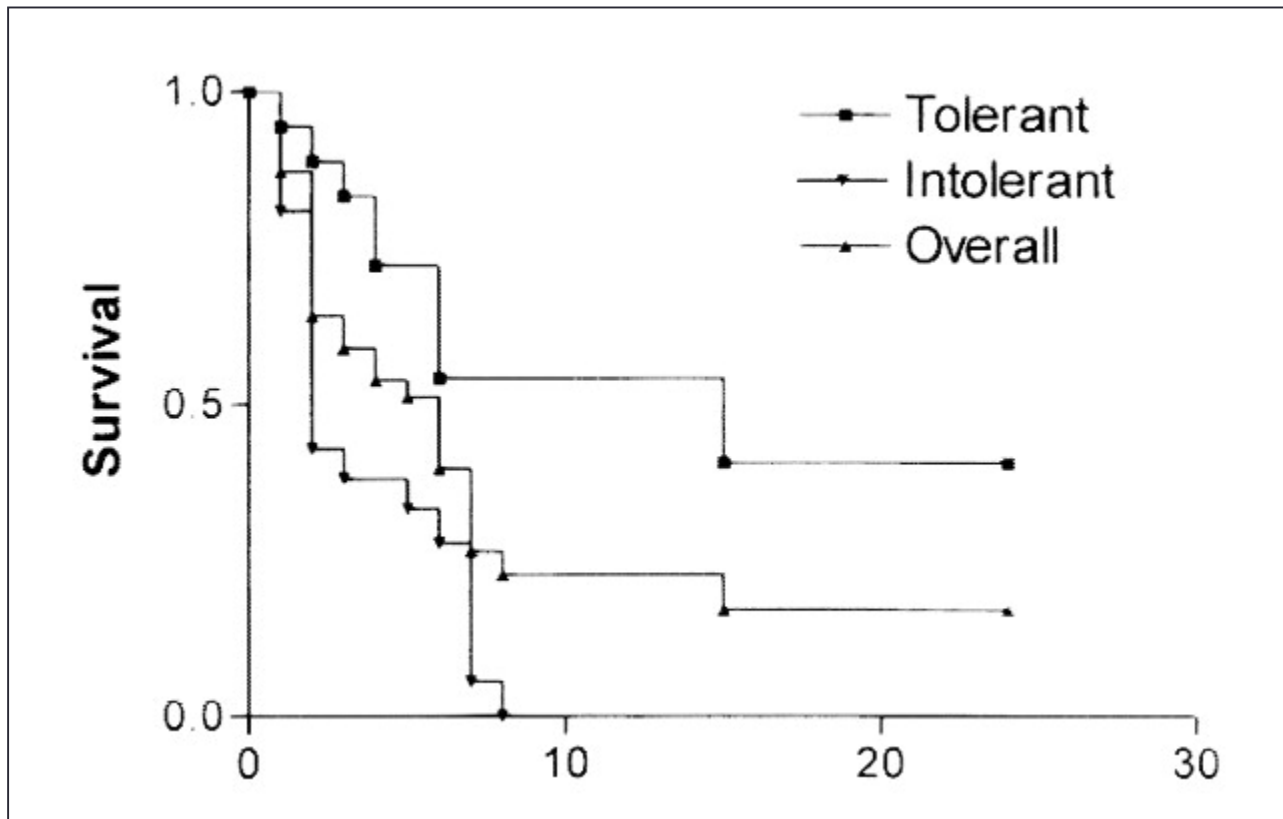
- Most patients (63%) died in a medical facility
- The most frequently reported cause of death was:
 - **respiratory failure (77%)** including:
 - terminal respiratory insufficiency (58%)
 - pneumonia (14%)
 - asphyxia due to a foreign body (3%)
 - pulmonary embolism (2%)
- Ten per cent of patients died from other causes: post-surgical or traumatic conditions (5%), cardiac causes (3.4%), suicide (1.3%) and sudden death of unknown origin (0.7%)
- The cause of death could not be determined in 13% of cases (6% inside a medical facility and 25% outside)

Gil J, Funalot B, Verschueren A et al. Causes of death amongst French patients with amyotrophic lateral sclerosis: a prospective study. Eur J Neurol 2008;15:1245–51.

Effect of Noninvasive Positive-Pressure Ventilation on Survival in Amyotrophic Lateral Sclerosis

L.S. ABOUSSAN, S.U. KHAN, D.P. MEEKER, K. STELMACH, H. MITSUMOTO

Kaplan-Meier survival plots from initiation of NIV in Amyotrophic Lateral Sclerosis

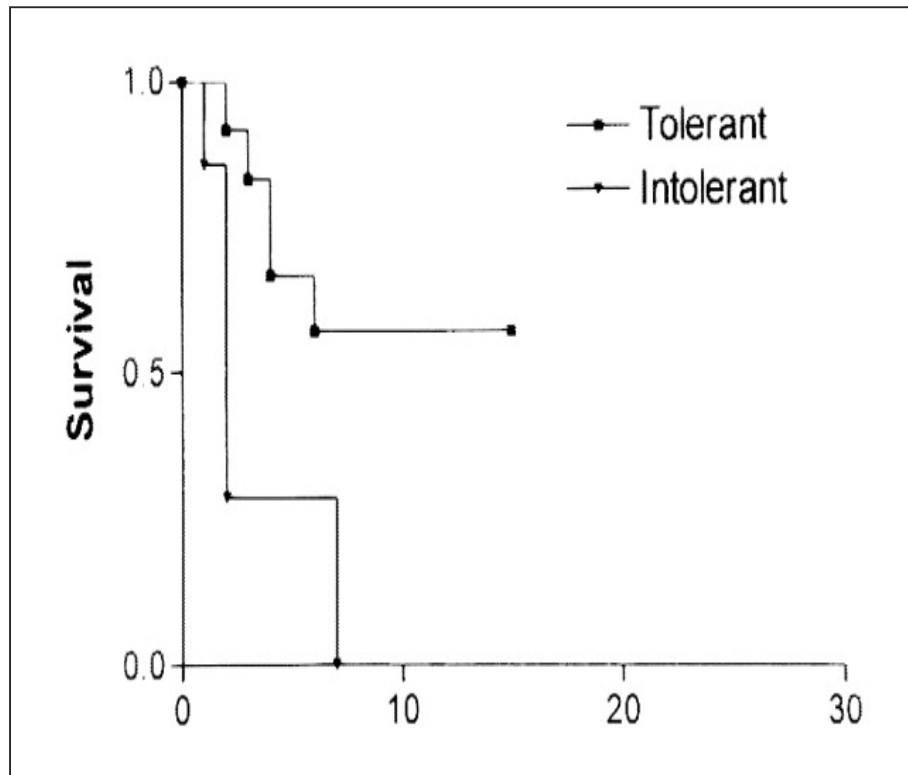




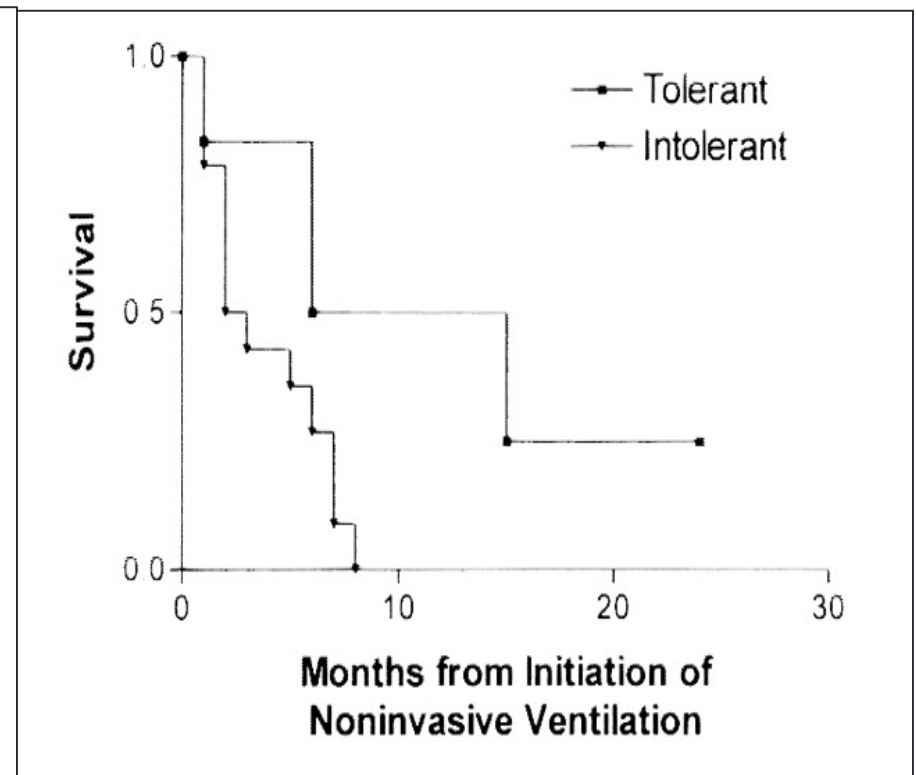
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Kaplan-Meier survival plots from initiation of NIV in ALS patients with moderate or severe bulbar symptoms



Moderate bulbar symptoms



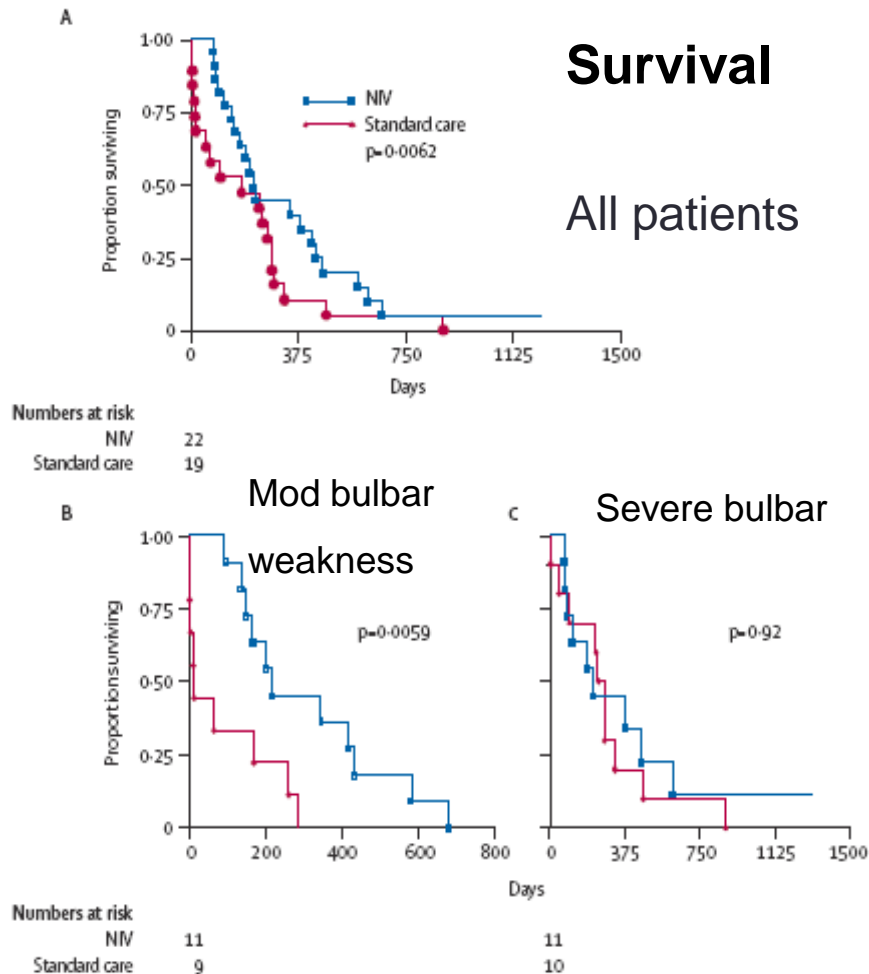
Severe bulbar symptoms

RCT of NIV in MND

Bourke SC Lancet Neurol 2006;5:140-7

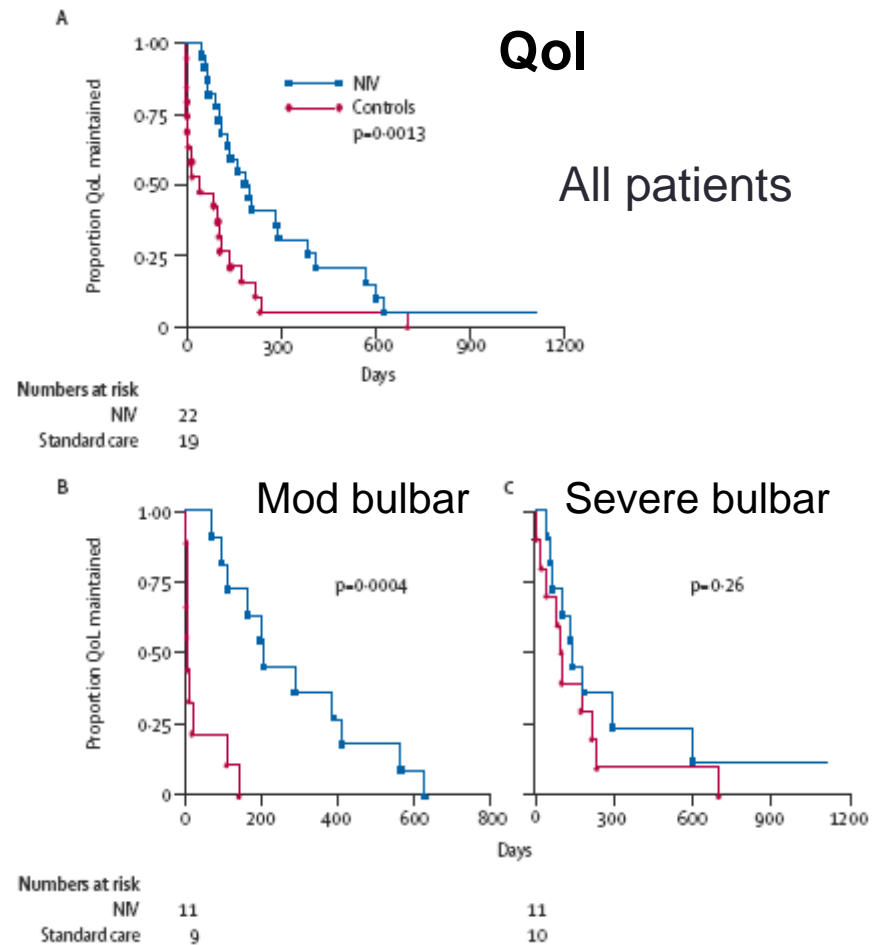
Survival

All patients



QoL

All patients



No survival advantage in severe bulbar patients but QoL improved

NIV in MND/ALS: Quality of life

- *Bourke et al Neurology 2003*: Assessment pre, 1, 3, 5 month after starting NIV
- Generic: Improvements in GWbS ($p=0.039$), SF36 emotional limitation, health perception
- Specific: Improvements Epworth SS, SAQLI, CRDQ dyspnoea, fatigue & mastery
- Improvements at 1 month maintained at 5 months despite disease progression
- Indices of sleep-related symptoms most responsive
- *Lyall et al Neurol 2001*: NIV increased Vitality domain (SF36) by 25% for up to 15 months despite disease progression

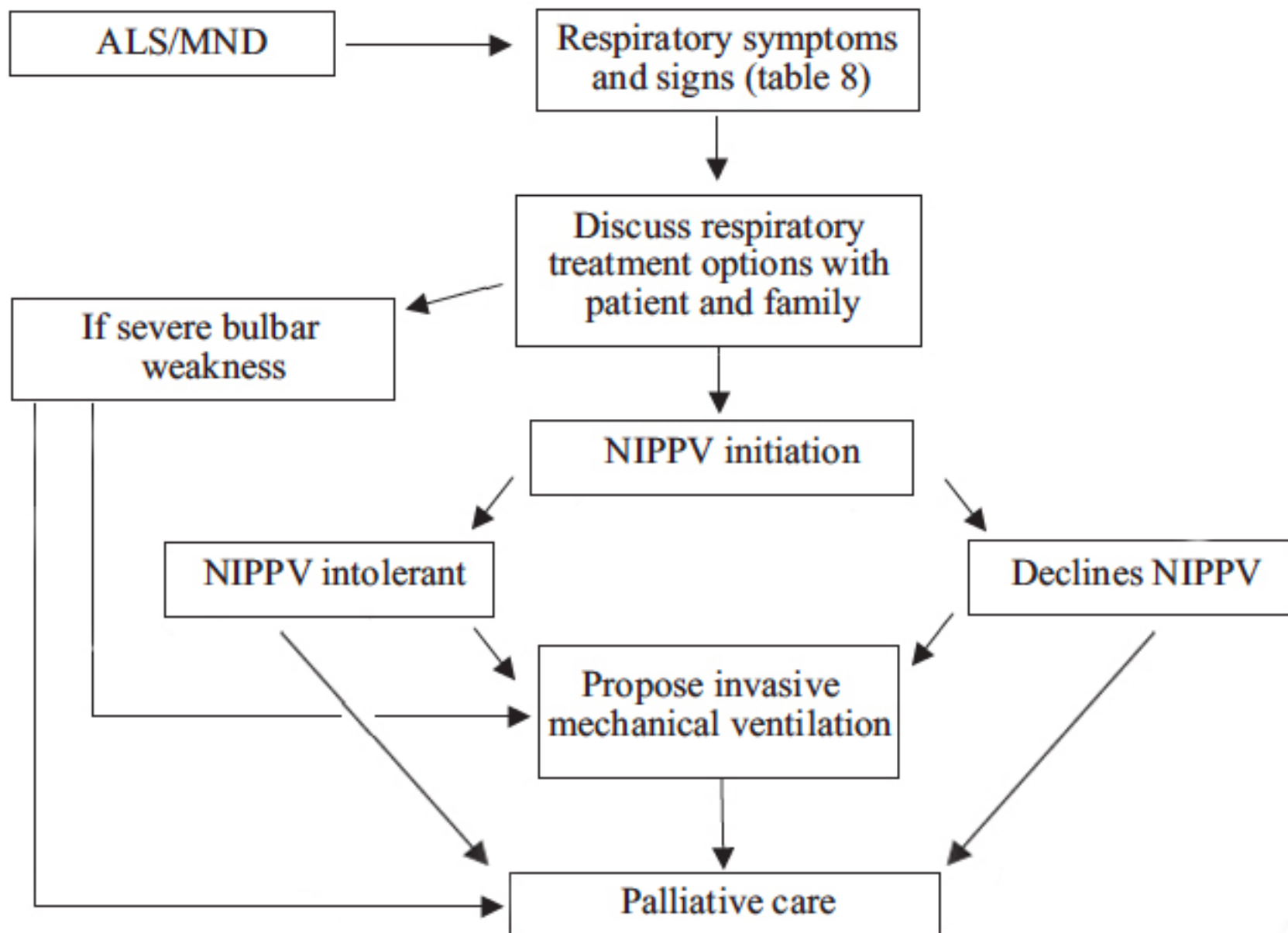
Noninvasive Ventilation in ALS

Table 3. Criteria for initiation of respiratory support in ALS patients

**Presence of
symptoms related to
respiratory failure
associated with one
of the following
objective criteria:**

- PaCO₂ greater than 45 mm Hg and/or
 - Vital capacity less than 50% of normal and/or
 - Max. sniff nasal insp. pressure < 60% normal and/or
 - Nocturnal O₂ desat. < 90% > 5% of the time
-

Gordon PH. *Amyotrophic Lateral Sclerosis: An update for 2013 Clinical Features, Pathophysiology, Management and Therapeutic Trials*. Aging Dis. 2013;4(5):295-310.



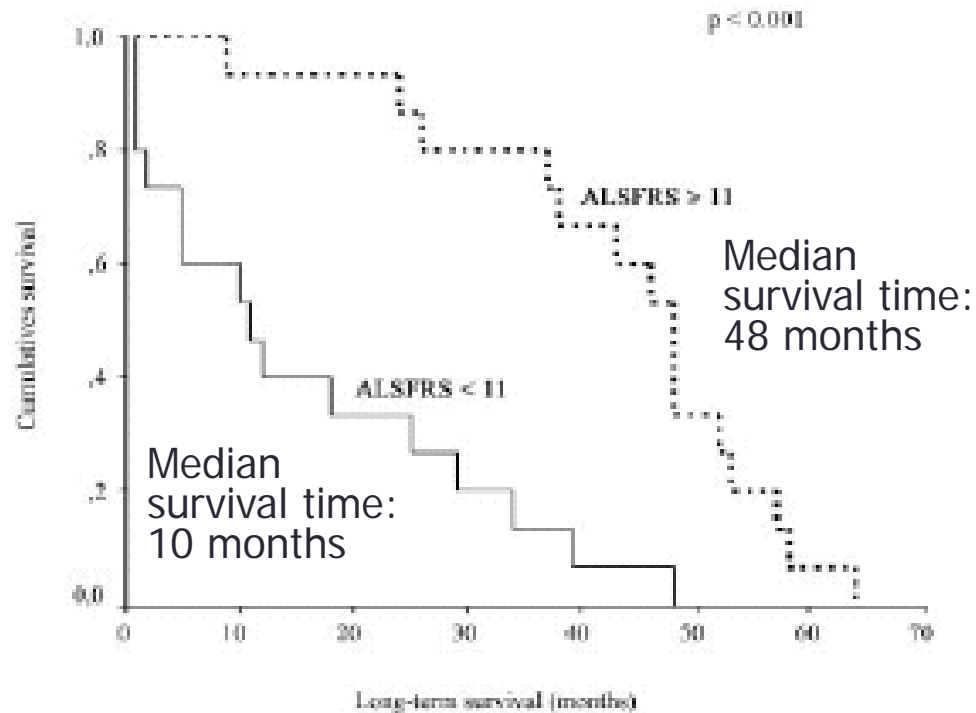
Report of an EFNS task force. Eur J Neurol. England; 2012;19(3):360-75

Palliative Considerations Regarding NIV in ALS

- tendency to gradually increase its use – eventually to 24/7; the implications of this does not seem to be commonly discussed
- patient may be completely dependent on NIV, and unable to remove mask in event of machine or power failure
- the very patients who selected NIV rather than tracheostomy often find themselves on “life-support” with NIV, having to decide about withdrawal of ventilatory support
- care setting for patients dependent on NIV need to address:
 - risk management around power / machine failure
 - ability to address symptoms in context of acute distress
 - ability to ensure comfort in context of withdrawal

The ALSFRS predicts survival in ALS patients on invasive mechanical ventilation

D. Lo Coco, V. La Bella, T. Piccoli, A. Lo Coco



Long-term survival after TIPPV according to ALSFRS score

Life Satisfaction Index –11 (Italian Version)

Items	Ad	HC	Li	Sh	Fr
1) Invecchiando, le cose sembrano meglio di quanto avevo pensato 1) As I grow older, things seem better than I thought they would be	Z	—	—	—	—
2) Nella vita ho avuto più opportunità della maggior parte della gente che conosco 2) I have gotten more of the breaks in life than most of the people I know	—	—	C	—	—
3) Questo è il periodo più triste della mia vita 3) This is the dreariest time of my life	M	—	—	—	M
4) Sono felice tanto quanto lo ero da più giovane 4) I am just as happy as when I was younger	M	M	M	M	M
5) La mia vita potrebbe essere più felice di adesso 5) My life could be happier than it is now	M	M	M	M	M
6) Questi sono gli anni migliori della mia vita 6) These are the best years of my life	M	M	M	M	M
7) La maggior parte delle cose che faccio sono seccanti o monotone 7) Most of the things I do are boring or monotonous	M	—	Z	Z/M	Z
8) Mi aspetto che in futuro mi accada qualcosa di interessante e piacevole 8) I expect some interesting and pleasant things to happen to me in the future	Z	F	Z	Z	Z
9) Le cose che faccio mi interessano tanto quanto prima 9) The things I do are as interesting to me as they ever were	Z	—	Z	Z/M	—
10) Mi sento vecchio e piuttosto stanco 10) I feel old and somewhat tired	Z	—	Z	Z	Z
11) Sento la mia età, ma ciò non mi preoccupa 11) I feel my age, but it does not bother me.	—	—	—	—	—
12) Se ripenso alla mia vita passata, sono abbastanza soddisfatto 12) As I look back on my life, I am fairly well satisfied	C	C	C	C	C
13) Anche potendo, non cambierei il mio passato 13) I would not change my past life even if I could	C	C	C	—	C
14) Rispetto ad altre persone della mia età, ho preso molte decisioni sciocche nella mia vita 14) Compared to other people my age, I've made a lot of foolish decisions in my life	—	—	—	—	—
15) Rispetto ad altre persone della mia età, ho un buon aspetto 15) Compared to other people my age, I make a good appearance	Z	—	—	—	—
16) Ho fatto progetti per cose da fare fra un mese o fra un anno 16) I have made plans for things I'll be doing a month or a year from now	Z	Z	—	Z	Z
17) Ripensando alla mia vita, non ho ottenuto la maggior parte delle cose importanti che volevo 17) When I think back over my life, I didn't get most of the important things I wanted	—	—	—	C	—
18) In confronto ad altre persone, mi sento troppo spesso giù di morale 18) Compared to other people, I get down in the dumps too often	M	—	—	Z/M	—
19) Ho avuto parecchio di ciò che mi aspettavo dalla vita 19) I've gotten pretty much what I expected out of life	C	C	C	C	C
20) Nonostante quello che si dice, la sorte dell'uomo medio sta peggiorando, piuttosto che migliorando 20) In spite of what people say, the lot of the average man is getting worse, not better	—	—	—	—	—

Item categories are disagree / don't know / agree. Depending on the individual items, a score 0, 1, or 2 is assigned to categories to allow higher scores to indicate a better condition. The table gives the items' factorial association in previous studies and in the proposed solution. Both Italian (upper row) and English (lower row) versions are presented. The 11 items of the short form of LSIA, LSI-11, are in bold type. Ad = Adams; HC = Hoyt and Creech; Li = Liang; Sh = Shmotkin (see References). Fr = present study. Factors: M = mood tone; Z = zest for life; C = congruence between desired and achieved goals.

The Life Satisfaction Index – 11 is a short form of LSI questionnaire providing a cumulative score acknowledged as an index of quality of life

Survival and quality of life after tracheostomy for acute respiratory failure in patients with amyotrophic lateral sclerosis☆

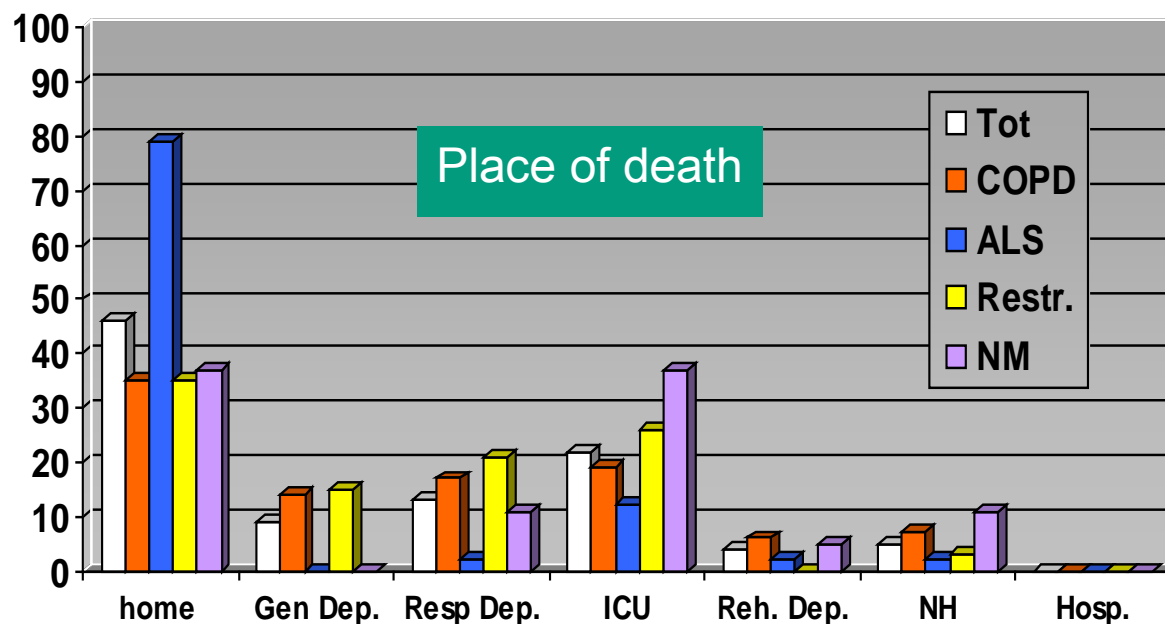
2011;26:329.e7-14

Andrea Vianello MD^{a,*}, Giovanna Arcaro MD^a, Arianna Palmieri PhD^b, Mario Ermani MD^b, Fausto Braccioni MD^a, Federico Gallan MD^a, Gianni Soraru' MD^b, Elena Pegoraro MD^b

	Trach ALS	ALS	Normal subject
Mood tone	1,57	1,60	3
Zest for life	4,14	4,10	4
Congruence between desired and achieved goals	2,85	3,15	3

Last 3 months of life in home-ventilated patients: the family perception

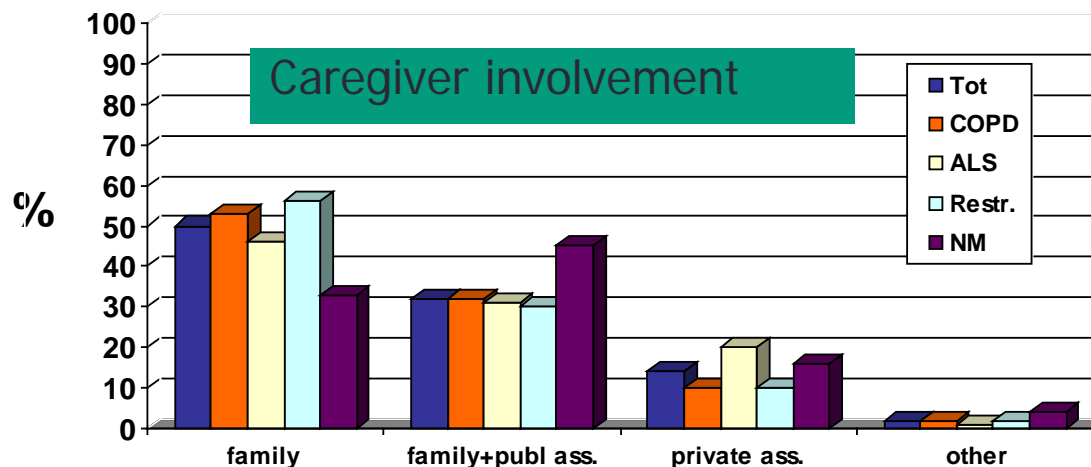
M. Vitacca, M. Grassi, L. Barbano, G. Galavotti, C. Sturani, A. Vianello, E. Zanotti, L. Ballerin, A. Potena, R. Scala, A. Peratoner, P. Ceriana, L. Di Buono, E. Clini, N. Ambrosino, N. Hill and S. Nava



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Italian multicenter survey on the last 3 months of life in 167 patients ventilated at home



Summary

1. There is good evidence that NIV may be used in terminally ill patients with different objectives (categories)
2. Up to 30% end stage chronic respiratory patients are receiving NIV in the last days of life (COPD, CPO)
3. Also being applied to the end stage cancer patients with good effect
4. About 50% of DNI patients with ARF may be successfully treated and discharged from hospital (mainly if COPD, CHF)

Summary

5. NIV can be used in people with ALS to improve symptoms and health perception; however, patient may become increasingly dependent and unable to remove mask
6. Patients with ALS transitioned to trach can live for many years and in most cases are happy with their choice