



ADVANCE DIRECTIVES AND PALLIATIVE CARE IN ALS PATIENTS: AN EPIDEMIOLOGICAL STUDY

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Introduction

Amyotrophic Lateral Sclerosis (ALS) is a neurodegenerative disorder involving both upper and lower motor neurons. The disease causes progressive limb weakness, bulbar palsy and respiratory failure; death occurs within 3 to 5 years from symptom onset (Beghi *et al*, 2006).

Since the disease has a fatal course, patients optimal management requires a palliative approach aimed at improving the quality of life (QoL) (Bede *et al*, 2011).

However, still nowadays in most countries palliative care is still not part of neurological training (Borasio, 2013). Moreover, a recent Italian study has shown that 58% of patients tracheostomized in the decade 1994-2005 had previously refused invasive ventilation (Veronese *et al*, 2014).

Aim

To evaluate the accessibility to Palliative Care Service (PCS), the diffusion of advance directives (ADs) and the respect of patients wishes in a population-based series of ALS patients.

Methods

All the 217 incident patients resident in the province of Turin, Piedmont, diagnosed with definite or probable ALS according to the revised El Escorial criteria (Brooks *et al*, 2000) over a period of three years (January 1st, 2008 - December 31th, 2010), were included in our study.

We investigated:

- how many patients died at the end of follow-up (June 1st 2014);
- the causes and place of death;
- how many patients referred to PCS;
- the completion rate of Ads form;
- the influence of gender, age at onset, ALS phenotype (familial vs. sporadic form), marital status, the use of non-invasive ventilation (NIPPV) and the presence of percutaneous gastrostomy (PEG/PRG) on patient's choice about tracheostomy.

The results of all quantitative data are reported as mean \pm standard deviation. Statistical analysis was performed using χ^2 -test and *t*-Student test, considering *p*-values less than 0.05 as statistically significant.

Results

Demographic and clinical characteristics of our patients are listed in Table 1.

At the end of follow-up 183 (84.3%) patients had deceased; among the 34 patients who were alive, 7 (20.6%) were tracheostomized (Figure 1). Place and cause of death are listed in Figure 2.

PCS support was activated for 50 patients (27.3% of deceased cases): 46 were followed up at home, 4 were admitted to the hospice. Patients referred to PCS showed no demographic differences compared to the other group of patients, but they more frequently used NIPPV (*p*=0.002) and died by palliative sedation (*p*<0.001)

n° (%)	Gender	
	m	f
122 (56.2)	122 (56.2)	95 (43.8)
years (SD)	Age at onset	
	m	f
66.6 (9.9)	66.6 (9.9)	67.4 (11.2)
n° (%)	Type of onset	
	Spinal	Bulbar
149 (68.7)	149 (68.7)	68 (31.3)
n° (%)	ALS phenotype	
	Sporadic	Familial
201 (92.6)	201 (92.6)	16 (7.4)

Table 1

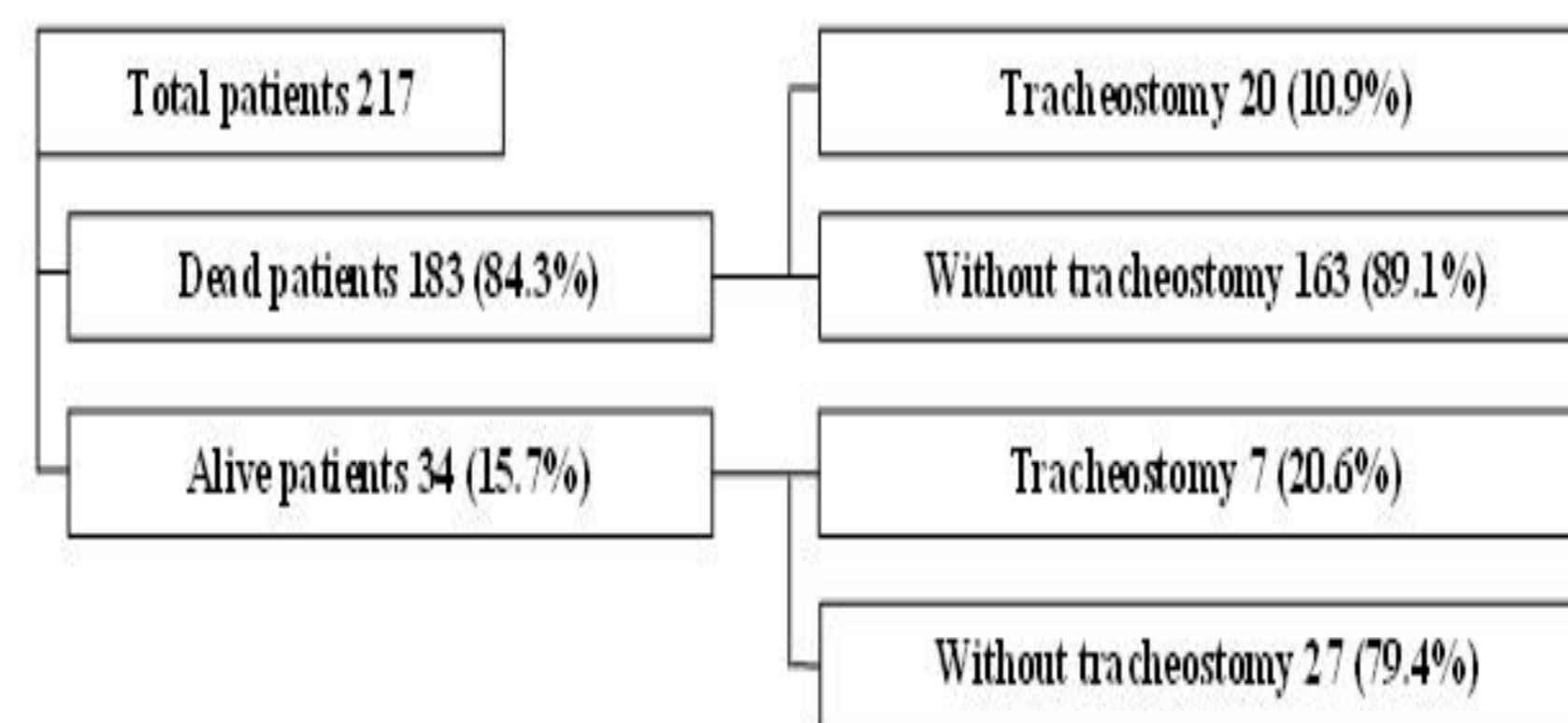


Figure 1

Place of Death	
Home	122 (66.7%)
Hospital	7 (3.8%)
Nursing Homes	4 (2.2%)
Hospice	1 (0.5%)
Suicide in Switzerland	1 (0.5%)
Unknow	21 (11.5%)

Cause of Death	
Palliative sedation	61 (33.3%)
Respiratory failure	60 (32.8%)
CCVA*	16 (8.7%)
Sepsis	12 (6.6%)
Neoplastic cachexia	1 (0.5%)
Assisted suicide	1 (0.5%)
Unknow	31 (16.9%)

Figure 2 - *CCVA: cardio-cerebral-vascular accidents

ADs form was completed in 105 cases (55.3% of deceased and/or tracheostomized patients) (Figure 3). At time of discussion, patients had a media ALSFRS score of 21 [IQR 16-30] and a median MITOS Stage of 2 [IQR 1-3].

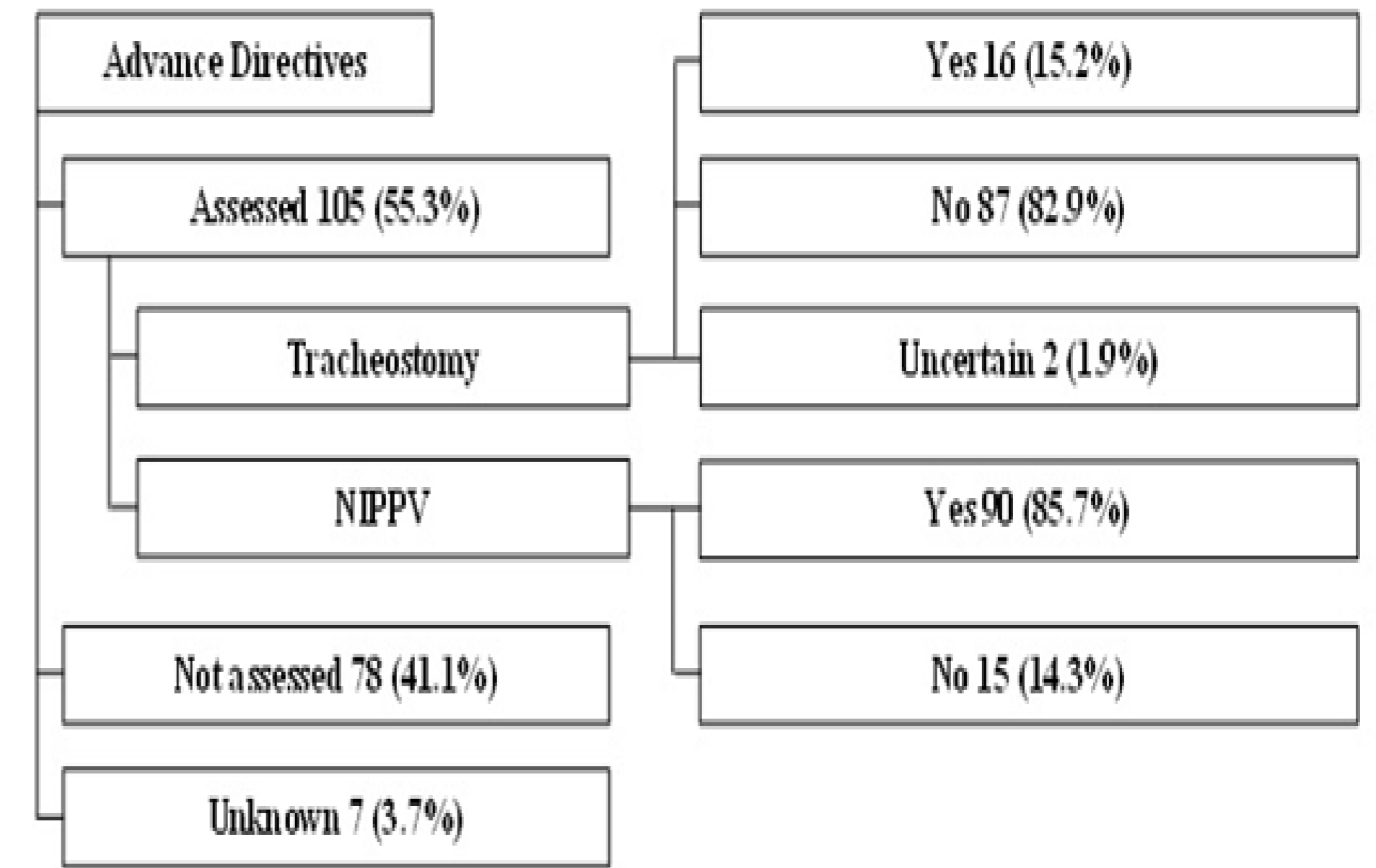


Figure 3: flow-chart of ADs patients decisions

Among patients who opted for tracheostomy, 3 did not undergo the procedure (2 cases because of sudden death, 1 for the opposition of a relative), while among those who refused tracheostomy, 7 were finally tracheostomized (5 were admitted at the Emergency Department because of respiratory failure and were tracheostomized upon medical decision, 1 upon the request of a relative, 1 patient changed his/her will during a crisis of respiratory failure). In our series patients' decisional autonomy was followed in 90.3% of the cases.

Tracheostomy was refused more frequently by older patients (*p*=0.001), while patients performing PEG/PGR were significantly more favorable to undergo tracheostomy (*p*<0.001). Gender, ALS phenotype, marital status and the use of NIPPV did not influence patients' choice about invasive ventilation.

Conclusion

More investigations are still required to better define the role that PCS could play in the management of ALS patients. Moreover, we deem it necessary to make further efforts to foster an early use of ADs.

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