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## 1 Ethical issues: invasive 2 ventilation in amyotrophic 3 lateral sclerosis 4

5 The first man was a school headmaster.  
6 By his eye-gaze system he is able to order  
7 coins to enlarge his 10-year collection. His  
8 grandchildren extort presents from him in  
9 exchange for help. The second was a pre-  
10 fect and even now insists that the flowers  
11 in the garden represent the national flag.  
12 In spite of his gastrostomy, he still likes to  
13 sip his espresso from porcelain cups. The  
14 ventilation circuit 24/7 has not changed his  
15 custom to read the newspapers and listen  
16 to classical music in the living room. That  
17 girl is a young mum. She is fed by a tube,  
18 but she always makes her little child laugh  
19 with the dialectal phrases that she writes  
20 on the screen. The surgeon, every day in  
21 his electric wheelchair, reaches the creek  
22 to admire the sea. He stays in the sun as  
23 long as the ventilatory battery allows him  
24 to. The teacher, on the contrary, lives in  
25 a bed. Around her, her husband mumbles  
26 and grumbles about their terrible fortune  
27 and never smiles. Another was a shepherd,  
28 and he died because an electric blackout  
29 turned off the ventilator. The boy's father,  
30 instead, is illiterate, but he is the best nurse  
31 you could possibly imagine.

32  
33 Proposing tracheostomy to amyotrophic  
34 lateral sclerosis (ALS) patients is not an easy  
35 task. While you are choosing the first words,  
36 all those different stories follow one another  
37 in your mind. As always, the phone is ring-  
38 ing in the room and many people are waiting  
39 for you outside the door. It seems a common  
40 conversation between doctor and patient, to  
41 obtain the informed consent, but the point  
42 is what must be chosen: death or tracheo-  
43 stomy? Years ago we had talked about the  
44 very modest benefit from Riluzole on the  
45 progression of the disease. Many other  
46 drugs since then have been tried unsuccess-  
47 fully, but unfortunately, at each new trial,  
48 the enthusiasm was soon substituted by  
49 disappointment. Visit after visit we observed  
50 the relentless advance of the disease that  
51 gradually stole all the forces and impaired  
52 speech, swallowing and respiratory func-  
53 tions. Every time a new problem arose, he  
54 had asked us the same question: 'what can  
55 we do?', receiving, then incredulous, at the  
56 same vague answer. Neurodegenerative dis-  
57 orders do not allow recovery. They deny  
58 the physician of the special privilege to treat  
59 a patient and give him a new lease of life.  
60 They disarm us. The last purpose of our  
61 job, then, is to relieve suffering and help to  
62 strengthen the patient's resources to go on.  
63 Most of ALS patients do not have cognitive  
64 or behavioural changes, preserving irony,  
65 character, intelligence and an unaltered dig-  
66 nity inside a body completely destroyed.  
67 They are ready to travel on a wheelchair, to  
68 eat by gastrostomy, to breathe with a mask

and to communicate by an eye-gaze system.  
It seems impossible, but the natural desire  
to live makes all these acrobatic solutions,  
and many more, acceptable. But despite all  
efforts, they will lose, and the natural course  
of the disease will end within 3–5 years,  
nearly invariably with severe respiratory  
failure. So here we are, in this room, at the  
final crossroads. He can demand a peaceful  
death or choose to undergo tracheostomy,  
as the last card to play to live many more  
years. Both choices are irreversible, since in  
Italy it is illegal to stop the ventilation sup-  
port, once started. Invasive ventilation in  
western countries is a procedure generally  
discouraged. The common opinion among  
physicians is that it prolongs the survival of  
the patients without slackening the disabili-  
ty progression that inexorably proceeds to  
the locked-in state. This condition is con-  
sidered incompatible with an acceptable  
quality of life. Paradoxically, the few stud-  
ies that until now have assessed the quality  
of life of tracheostomised ALS patients have  
shown scores comparable with the general  
population.<sup>1</sup> In fact, patients in the advanced  
stage of the disease suffer from respiratory  
symptoms and difficulty in mucus manage-  
ment, and often need to use a continuous  
ventilatory support by mask. From the per-  
spective of the patient, therefore, to pass  
from this really uncomfortable condition to  
invasive ventilation by tracheostomy prob-  
ably does not mean a worsening, especially  
if this change was previously considered  
and expected. It is common knowledge that  
tracheostomy ventilation provides a bet-  
ter mucus management than non-invasive  
ventilation) and also relieves the respiratory  
symptoms. Is it then fair displaying all these  
concerns about the quality of life? However,  
the American Academy of Neurology and the  
EFNS (European Federation of Neurological  
Societies) guidelines,<sup>2, 3</sup> recommend in-  
vasive ventilation to preserve quality of life  
of patients that want long-term ventilatory  
support. 'What the patient wants' is obvi-  
ously declared as a fundamental principle  
on end-of-life issues and seems to explain  
the remarkable differences on the attitudes  
towards invasive ventilation between differ-  
ent countries.<sup>4</sup> In Japan, the highest rate of  
use of tracheostomy—positive pressure ven-  
tilation in ALS (29%)<sup>5</sup>—is reported, while in  
the USA and North Europe only 2.1–5.4%  
of ALS patients receive this support in the  
terminal phase. This very meaningful differ-  
ence expresses, in the common opinion, the  
distance between the western health system  
in which the patient's will steers the medi-  
cal decisions, and the opposite attitude to be  
guided by physicians, that is typical in the  
Japanese culture. Following this line of rea-  
soning, invasive ventilation is so rarely used  
in western countries because almost all ALS  
patients in the terminal phase do not want  
to continue to live, and they freely decline to  
drag out their condition. Plenty of evidence

is inconsistent with this hypothesis. The  
first is that invasive ventilation by tracheo-  
stomy requires enormous costs for ventila-  
tion equipment and nursing care. In Japan,  
all these costs are fully covered by medical  
insurance. In North America, the health  
insurance hardly ever covers these costs, and  
in Europe, even when ventilatory support is  
provided (as is the case in Italy), the home  
nursing care weighs on the household bud-  
get. So, is the patient really free to decide?  
A distinct problem is that the patient and his  
caregivers do not receive any social or psy-  
chological support from the Health Systems.  
The human cost of such a devastating disease  
is incommensurable. By proposing tracheo-  
stomy, what are we asking of the patient?  
If he can bear the costs of the treatment? If  
his wife, any son, any brother or a friend are  
ready to put all their energies into his care?  
In this anomalous consent request, 'what the  
patient wants' is just one of the reasons of  
the choice. Besides the individual troubles,  
the collective dynamics affect the patient's  
decisions. Every word that the physician  
chooses to describe a procedure changes the  
idea that the patient forms about it and his  
agreement. It is natural that a doctor trusting  
to any operation will infuse the patient with  
faith, and vice versa. Unconsciously, physi-  
cians every day run the risk to point out their  
intuitive or personal vision of the outcomes,  
overlooking the contrary published eviden-  
ces. Equidistance is a very ambitious goal.  
The books written with eye-gaze system and  
the different personal stories of the patients  
prove that for some people also, a locked-in  
life is acceptable and even full of value. On  
the contrary, many other people think that  
a life without autonomy is not dignified.  
Did we dedicate enough time to know the  
values of the patient before discussing with  
him about his end of life? Talking with our  
patient, we carry the burden of a multitude  
of concerns. Explaining the risks and benefits  
of the procedure seems to be like walking  
on burning coals, trying to pick out the most  
sublimating words. Meanwhile, the patient  
in front of us stares into space. Unbelievably  
quiet.

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