

Patients with ALS may die from multiple causes. I'd classify them in two categories: *direct causes* and *indirect causes*.

1. **A) Direct causes:** The illness causes paralysis in our voluntary muscles. The only vital voluntary muscles in our bodies are respiratory muscles, including the diaphragm. That is why there's only one way to die directly from ALS: respiratory failure, or extrapulmonary restriction. A proper ventilator setup and tracheotomy perfectly replaces the mechanical functions of our respiratory muscles. Theoretically, we are out of harm's way. Yet, many of you will let me know that they lost a loved one in spite of a tracheotomy. Of course, tracheotomy does not make us immortal, and a ventilator doesn't prevent physiological lung problems.
1. **B) Indirect causes:** There are many indirect causes, which can also happen before tracheotomy. I won't be able to name them all here, so I'll only quote those which happened to me. Pulmonary embolism, sepsis, falls, peritonitis, acute malnutrition, and of course respiratory failure. All of these niceties can be life threatening. However, contrary to what I often read, *respiratory distress isn't possible when a patient has a proper ventilator setup and a tracheotomy, and if their lungs are healthy*. Respiratory problems will appear only if lungs are directly or indirectly affected (infection, embolism, spasms, edema...). This information comes from the hospital resuscitation department that took care of me and they were formal. I think reanimators are among the most qualified medical professionals to address post-tracheostomy lung problems.

Let's make that clear: ALS attacks neither lungs or bronchi. However, several post-tracheostomy acute lung impairments can happen in ALS. I will discuss 2 of them here.

Respiratory infections caused by germs, most often superbugs (antibiotic-resistant bacteria). Most of the time, these germs got inside the tracheostomy during cannula changes (here's why these changes should happen at home, a way safer place than the hospital on a septic point of view), or during tracheal suction (hence why a closed loop-system is needed), or finally during hygiene related questionable manipulations. This is why hygiene should be tantamount in all everyday gestures regarding tracheostomy. If those germs reach bronchi, they can cause a lot of harm.

The second risk is pulmonary aspiration. In addition to being a risk of choking, it represents a significant infectious risk. If food gets into the respiratory tract, it can constitute a considerable infectious hazard, and respiratory weakness will make it very difficult to expel. In this case, the germs that cause pulmonary infections are located inside the mouth, a septic cavity, hence the focus on oral hygiene.

This can happen before tracheostomy, and can be problematic post-tracheostomy. Hence the astonishment of some people who think tracheostomy protects them from pulmonary aspiration (which is the case) and get a pulmonary infection linked to a pulmonary aspiration event that happened before tracheostomy. That's why one should not delay tracheostomy when pulmonary aspirations get common.

I've also read that ALS attacks the heart, because it's a muscle. This is completely false, as any neurologist will tell you. However, the heart may fail for *reasons indirectly related* to ALS. For example, if one stays in respiratory distress for several days in a row, major fatigue can appear. And just as when anybody makes excessive effort, the risk of having a heart attack increases tenfold, especially if it finds a favorable ground.

I've also read that someone had been denied tracheotomy because "ALS was too far down". It takes a lot to discourage a resuscitator, yet when I arrived in intensive care, they did not believe I stood a chance. My wife almost had to get into a fight with them to comply with my advance directives. They didn't understand the aim of it all. However, after they tracheotomized me and got to know us, some became friends and everyone in the unit understood and accepted our choice. All of us accompanied us and in the end, the whole unit had changed their ways of seeing things. (Well, I stayed there for 2 months). They even made some administrative changes to the unit so that they could be more attentive to patients' and families' needs - wow that's amazing! They had also act because of a lack of information, thinking that having Lou Gehrig's disease meant that I was done for.

It's never too late. The saying "When there's life, there's hope" applies.

Lack of information is astounding, at every level. False information is spread, and this is even worse. I think this is one of the reasons that only 3% of us choose tracheostomy in France compared to 46% in Japan. Maybe the Japanese may have a very different philosophy towards welcoming sickness and death, but I don't think it's enough to explain such a gap.

Whether you choose tracheostomy or not, it doesn't matter to me, *it's your choice*. What matters to me is that you can do this choice while fully knowing all that this decision implies. My dream is that society switches from "Oh my God, they have ALS, they're done for!" to "Oh my God, they have ALS, it's tough but it's possible!".

I'm 45, I have 2 little daughters, a wife, I'm quadriplegic, tracheotomized, fed through a gastrostomy tube, and happy.

Have a good day, folks.

Your experience matters:

What are the statistics and attitudes on tracheostomy in your country?

What are the falsehoods you heard about tracheostomy?

Partager l'article !

Leave a comment with your Facebook account