

Choices

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One of the most important issues I can relay to parents of newly diagnosed Type I infants is that they do have a choice and that choice should be based on educated and accurate information. As I see it there are 3 possible choices that currently exist for children with SMA Type 1.

They can be left for nature to take its course and one can give them all their love and enjoy their time together;

they can undergo tracheotomy and very possibly survive in to adulthood but without the ability to breathe or speak on their own;

or one can use a newly described approach in an attempt to raise the children without resort to tracheostomy. The latter has both advantages and disadvantages.

The key points to understand in deciding to avoid tracheostomy for children with SMA is that eventually, whether having type I, II, or III, virtually all children will develop bronchitis that will cause the airway to be encumbered with airway secretions. This occurs because the abdominal muscles will not be powerful enough to generate sufficient cough flows to expel them. For children with SMA type I this usually happens before 1 year of age; for those with SMA type II, between 1 1/2 and 12 years of age, and for SMA type III, between 8 and 30 years of age.

For children too young to understand or cooperate with manually and mechanically assisted coughing, this usually means that intensive care and intubation are necessary. Most physicians think that once a small child with SMA is intubated, "weaning" from ventilator use may be impossible, and tracheotomy is necessary. However, we have demonstrated that this is not true. We have successfully extubated over 90% of small SMA type I and II children by following the "protocol" that we described in an article published in Chest in 2000. (See text of [Chest article](#)).

This "protocol" consists of basically not extubating until oxyhemoglobin saturation is normal without oxygen therapy, not giving oxygen after extubation, extubating to high span "BiPAP", and using mechanically assisted coughing both during the intubation and afterwards. When not using this approach the extubation success rate is 6% and this is the reason that most physicians think that a trach tube is necessary. We have shown that after the 3rd birthday, hospitalization rates for children without trach tubes is less than for those with them and that while virtually all children with tubes require ventilator use around-the-clock and can not speak, most without them can speak and only require nocturnal BiPAP. Although sometimes children need to be intubated on 6 or more

occasions before getting old enough to cooperate sufficiently with mechanically assisted coughing and oximetry to avoid pneumonia and respiratory failure, for SMA type I children after age 5 we have had only 1 hospitalization in over 25 patient-years.

On the other hand, for children for whom tracheostomy is avoided, intercurrent illnesses require much closer parental attention, often 24hr/day for many days, than for children with tracheostomy tubes because few nurses or other health care professionals truly understand how to manage acutely ill SMA kids without tubes as well as the parents.

Thus, the key to keeping SMA I and acutely ill SMA II kids without trach tubes is the proper management when they need intensive care. The key to keeping their lungs and chest walls growing properly is to use nocturnal high span BiPAP whenever they have "paradoxical breathing", that is, when their chests do not expand as their bellies protrude during inhalation.

I leave the choice to those living with SMA and give hope and support to their decision.