neuron disease causes stridor and upper airflow obstruction, whereas with SMA, there is little or no stridor; thus, some of the 20-year-olds in our series with SMA1 generate > 350 L/m of MIE expiratory flows. Thus, intubation/hospitalization rates decrease dramatically after 3 years of age.[2] The 23-year-old patient in Figure 5 was intubated 11 times; however, 10 intubations occurred by age $4^1/_2$ years and one subsequently.[2] It is time to stop assuming that all "ventilatory support" must be invasive, and "NIV" only bilevel PAP, and use full ventilator setting NVS instead.

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Rebuttal From Dr Panitch

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Dr Bach and I agree on the need for ventilatory support and airway clearance for infants with SMA1.[1] He has demonstrated success with his noninvasive approach, although his recommendations for some of the applied pressures used for ventilation and cough clearance seem both arbitrary and excessive. The pressures applied at the airway opening to achieve adequate tidal volumes for ventilation or inspiratory capacities for cough clearance must overcome the elastance of the lungs and chest wall, as well as the resistance of the extrathoracic and intrathoracic airways. The chest wall of infants and young children with neuromuscular disease is more compliant (has a lower elastance) than the chest wall of age-matched children, whereas lung compliance is similar.[2] Passive inflation of the respiratory system in healthy infants with 30 cm H_2O pressure at the airway opening results in a lung volume that approximates total lung capacity.[3] There is no reason to assume a priori that airways resistance should be elevated in infants and children with SMA compared with healthy children; thus, ample volumes and flows for airway clearance can be achieved at lower pressures than Dr Bach recommends, even when applied through pediatric tracheostomy tubes.[4] When delivering ventilatory support through a bilevel generator with a passive expiratory valve, expiratory pressure of 3 to 4 cm H_2O is required to guarantee expiratory flow to wash out circuit dead space and prevent CO_2 rebreathing. Inspiratory pressure should be applied to achieve adequate chest rise and to eliminate thoraco-abdominal paradox. This outcome is usually achieved by creating a difference between inspiratory and expiratory pressure of at least 10 cm H_2O.[5] Limiting positive pressure to the amount necessary to achieve these goals enhances patient comfort and can limit untoward side effects of noninvasive ventilation and airway clearance therapies.

Perhaps where Dr Bach and I differ the most is in our attitudes toward tracheostomy in the armamentarium of ventilation therapies for patients with SMA. He refers to the lack of ability of some parents to provide NVS and frequent, aggressive airway clearance as a cause for failure of his approach.[1] Indeed, we can agree that support of infants who have SMA1 requires tremendous commitment on the part of caregivers, but even tremendously committed caregivers may not possess the requisite skills to support their child noninvasively. Furthermore, the course of infants with SMA1 who require tracheostomy may be more severe than those who can be supported noninvasively.[6] Parents who face the difficult choice of whether to proceed to ventilation via tracheostomy should not have the added onus of perceived personal failure imposed upon their decision process. The well-described complications related to tracheostomy must be balanced with the risk of airway obstruction from secretions or ineffective bag-mask ventilation by lay caregivers during an acute event. Even when parents become expert at noninvasive airway and ventilatory management, the pool of skilled and unskilled caregivers who could provide them assistance and respite is often limited. As professionals who provide information and direction to these families, we

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should present all options, with their attributes and drawbacks, to families of children with SMA1 seeking such advice.

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