Laryngotracheal Reconstruction Outcomes in Hypotonic Children

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IMPORTANCE Children with poor muscle tone may demonstrate upper airway obstruction due to several mechanisms including obstructive sleep apnea, laryngopharyngeal reflux, and laryngomalacia. Though hypotonia has been shown to compromise the pediatric airway, and some authors suggest that neurologic deficits can compromise the success of laryngotracheal reconstruction (LTR), to our knowledge no studies have evaluated the effect of neurologic diagnoses or hypotonia on outcomes in LTR.

OBJECTIVE To determine whether hypotonic children with subglottic stenosis have lower rates of successful decannulation after LTR compared with children without neurologic deficit.

DESIGN, SETTING, AND PARTICIPANTS A retrospective medical chart review was conducted for 27 children aged 0 to 6 years, who underwent LTR for subglottic stenosis between December 2007 and December 2012 at a tertiary care children's hospital. Children were classified based on documented neurologic findings. Group 1 comprised those children without neurologic impairment (n = 16). Group 2 included those children with a documented neurocognitive or neuromuscular diagnosis but without evidence of hypotonia (n = 7). Group 3 comprised hypotonic children (n = 4).

INTERVENTIONS Laryngotracheal reconstruction.

MAIN OUTCOMES AND MEASURES The number of procedures performed after LTR to optimize the airway and whether the child was successfully decannulated.

RESULTS All 16 of the neurologically intact patients (100%) were decannulated. Among children with a neurologic deficit, 5 of 7 (71%) were ultimately decannulated. No hypotonic children 0 of 4 were decannulated. The difference in rates of decannulation between unaffected and normotonic children with a neurologic deficit was not statistically significant (P = .08). However, the difference in outcomes between hypotonic children and neurologically intact patients was statistically significant (P < .001).

CONCLUSIONS AND RELEVANCE Findings from this study suggest that hypotonic children may experience poorer rates of post-LTR decannulation compared with children without neurologic deficit. Dynamic upper airway obstruction may be unappreciated in hypotonic children. Future research may be directed at the appropriate evaluation and treatment of children with poor muscle tone and subglottic stenosis.
Centralized neurologic deficits have been shown to compromise the pediatric airway and complicate outcomes in the surgical management of upper airway obstruction.1-6 These neurologic disorders can affect the muscle tone required to dilate and maintain the pharyngeal airway, producing a clinical picture consistent with obstructive pathologic conditions such as laryngomalacia and obstructive sleep apnea.1-7 Advances in surgical management of subglotic stenosis have produced high rates of successful decannulation; however, children with neurologic deficit and upper airway obstruction do not achieve the same outcomes as unaffected children undergoing similar surgeries.3-5

Dynamic obstruction of the pharyngeal and laryngeal airway is common in infants and in older children with hypotonia. A 2005 study of children with obstructive sleep apnea demonstrated that dynamic airway collapse in infants without neurologic deficits decreased progressively as those children aged.8 However, dynamic airway obstruction in hypotonic children did not improve with time. By the age of 2 years, 83% of hypotonic children demonstrated a dynamic obstruction compared with only 17% of normotonic patients.

Laryngeal collapse is also thought to have a neurologic etiology. Recent evidence shows that an immature or otherwise abnormally integrated sensorimotor brainstem reflex is present in children with clinical laryngomalacia.9 In children without a diagnosed neurologic deficit, these symptoms frequently remit as the patient’s nervous system matures, usually resolving by the second year of life. Symptoms of upper airway obstruction that mimic laryngomalacia, however, can persist in older children and adults with centrally mediated neurologic compromise.7-9

Though hypotonia has been shown to produce upper airway obstruction, and some authors suggest that neurologic deficits can compromise the success of LTR,10 we did not find any studies that evaluated the association of neurologic diagnoses or hypotonia with outcomes in LTR. In the present study, we investigate the hypothesis that children with hypotonia who undergo LTR for subglottic stenosis have lower rates of decannulation than children with normal muscle tone.

Methods

A retrospective medical chart review approved by the institutional review board of The Albert Einstein College of Medicine was conducted of all laryngotracheal reconstructive procedures for subglottic stenosis performed between December of 2007 and December of 2011 at our tertiary care children’s hospital. All procedures were performed by 1 of 2 surgeons (J.P.B. and S.R.P.). Children were grouped based on documented neurologic findings. A patient was classified as hypotonic only if their documented physical examination described hypotonia or global weakness. Group 1 comprised children without neurologic impairment (n = 16). Group 2 included children with a documented neurocognitive or neuromuscular diagnosis but without hypotonia (n = 7). Group 3 comprised hypotonic children (n = 4). Data collected included age at time of surgery, concomitant airway lesions noted on laryngoscopy and bronchoscopy, and whether the child was successfully decannulated. In addition, any surgery undertaken to improve the patency of the airway after the LTR was tallied, including dilations, debridement of granulation tissue, and adenotonsillectomy, but not including direct laryngoscopy alone or removal of subglottic stent in the second stage of a 2-stage LTR.

Comparisons of the rates of decannulation between each group were made using the Fisher exact test. One patient in the neurologically intact group never underwent a tracheotomy and was considered decannulated for the purposes of these analyses. The median ages and number of post-LTR surgical procedures were compared between the 3 groups with the Kruskal-Wallis test. Analyses for the entire sample were repeated for the subgroup with double-stage LTRs.

Results

A total of 27 children underwent LTR between December 2007 and December 2011; 26 had been tracheotomy dependent prior to the procedure. Of the 16 neurologically intact children, all 16 (100%) were decannulated. Among normotonic children with a neurologic deficit, 5 of 7 (71%) were ultimately decannulated. No hypotonic children (0 of 4) were decannulated (Figure). The difference in rates of decannulation between unaffected and normotonic children with a neurologic deficit was not statistically significant (P = .08). However, the difference in outcomes between hypotonic children and neurologically intact patients was significant (P < .001). The patient-specific characteristics are displayed in the Table.

The median (interquartile range [IQR]) age of unaffected children at the time of surgery was 37.4 (23.8-42.3) months, which was significantly younger (P = .045) than the 66.0...
(55.7-67.7) months of the neurologically impaired and the 66.0 (55.7-67.7) months of hypotonic children. The median (IQR) number of surgical procedures performed after the LTR was not significantly different between groups 1, 2, and 3 (0.5 [0.0-1.8], 2.0 [2.0-3.0], and 1.0 [1.0-1.8], respectively) (P = .11). Neither age nor number of surgical procedures was significantly associated with decannulation (P = .11 and P > .99, respectively).

A single-stage reconstruction was performed on 13 children, while 14 children underwent the double-stage procedure.
dure. Of those neurologically intact children who underwent the double-stage procedure, 5 of 5 (100%) were decannulated. Among normotonic children with a neurologic deficit, 3 of 5 (60%) were decannulated after a double-stage LTR. None of the 4 hypotonic children who underwent double-stage LTR were decannulated (Figure). A comparison of the rates of decannulation between these groups shows a significant difference between neurologically intact children and hypotonic children \( P = .008 \). As in the full cohort, the difference in rates of decannulation between unaffected children and normotonic children with a neurologic deficit was not significant \( P = .44 \). However, the statistical power to identify significance was low owing to the small size of our cohort.

**Discussion**

Our data suggest a lower rate of decannulation in hypotonic children after LTR compared with the rate of decannulation for children without neurologic deficit. Children with neurologic deficit but without hypotonia were also decannulated less frequently than unaffected children. However, this trend was not statistically significant, possibly because of low statistical power. A comparison of the number of procedures performed to optimize the airway after LTR was not meaningfully different between the patient groups, suggesting there was not a strong bias on the part of the surgeons and guardians to intervene more aggressively on behalf of patients without a neurologic deficit.

Most analyses of outcomes in LTR demonstrate more favorable decannulation rates for the single-stage vs double-stage LTR.\(^{10,11}\) However, the double-stage procedure is recommended in children with complex stenosis (including high-grade or multilevel disease), significant neurologic deficits, or pulmonary disease. Accordingly, rates of decannulation among children undergoing double-stage LTR may be lower in part because of confounding patient factors. Studies in which these elements are controlled arrive at conflicting conclusions on whether the rates of decannulation differ between the single- and double-stage procedures.\(^{10,11}\)

To eliminate the potential effect of these varied procedures, we repeated the analysis in the subset of those children who underwent double-stage LTR. The difference in rate of decannulation between neurologically intact children and hypotonic children undergoing double-stage LTR remained significant.

Children with poor muscle tone may develop obstructive upper airway symptoms due to an inability to dilate and maintain the pharyngeal or laryngeal airway.\(^1\) On direct laryngoscopy, this dynamic collapse may not be apparent, and the contribution of this obstruction will not be appreciated. In such a scenario, surgical management of a fixed obstruction such as subglottic stenosis may have limited efficacy.

Despite the effects of hypotonia on the pediatric airway, we could find no previous study that evaluated the efficacy of LTR in patients with neurologic disorders. Data from children with severe laryngomalacia may offer some guidance regarding the surgical management of the hypotonic airway. In a 2012 review of supraglottoplasty outcomes in children with severe laryngomalacia, the authors reported on overall risk ratio of 7.14 for surgical failure among patients with congenital cardiac disease, neurologic compromise, and laryngopharyngeal reflux compared with patients with isolated laryngomalacia.\(^4\) In a separate review of supraglottoplasty outcomes, Schroeder et al\(^5\) found that 6 of 11 patients with cerebral palsy or hypotonia required additional surgical intervention after supraglottoplasty because of persistent upper airway obstruction. In the same study, only 2 of 41 neurologically intact children required further surgery, both of whom had mandibular hypoplasia. A similar study by Hoff et al\(^6\) revealed that 7 of 10 children with neurologic deficits who underwent supraglottoplasty required further surgical intervention, 6 of whom ultimately required tracheotomy.

Our study has several limitations. As an observational, retrospective study we cannot rule out latent confounding factors. Furthermore, with only 6 patients in the whole cohort and 4 in the double-stage subset who were not decannulated, we had low statistical power. Accordingly, multivariable adjustment for potential confounders including age, reflux disease, and concomitant airway lesions could not be performed. However, neither age nor number of surgical was significantly associated with decannulation, suggesting a low likelihood that these variables meaningfully confound the results. Importantly, despite low statistical power, we found significant differences in decannulation rates between the hypotonic and neurologically intact groups.

Our study suggests that LTR may be less effective in hypotonic children, perhaps owing to unappreciated dynamic upper airway obstruction. Future research may be directed at the appropriate evaluation and treatment of children with poor muscle tone and subglottic stenosis.

In conclusion, after LTR, children with neurologic deficits are decannulated at a lower rate than neurologically intact children, and this difference becomes statistically significant when comparing hypotonic with neurologically intact children. Further research is needed to determine the optimal evaluation and management of the hypotonic airway.
Conflict of Interest Disclosures: None reported.
Previous Presentation: This study was presented as a poster of the same title at the American Society of Pediatric Otolaryngology Annual Meeting; April 26-28, 2013; Arlington, Virginia.

REFERENCES