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Title: "Speech Outcomes After Sphincter Pharyngoplasty for Velopharyngeal Insufficiency"

Running Title: Speech Outcomes After SP for VPI

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Abstract:

Objective:

To investigate perceptual speech outcomes following sphincter pharyngoplasty (SP) and to identify patient characteristics associated with velopharyngeal insufficiency (VPI) resolution or improvement.

Methods:

Retrospective review of prospectively collected data was performed of consecutive patients that underwent SP for management of VPI between 1994 and 2016 at a single tertiary care pediatric hospital.

Demographic data, nasendoscopic findings and speech characteristics were recorded using a standardized protocol. Pre- and post-operative VPI was graded on a 5-point Likert scale. Frequency of post-operative VPI resolution and improvement was assessed and associations with patient characteristics were analyzed. The association between odds of VPI resolution or improvement and five patient characteristics identified *a priori* was performed controlling for confounding factors.

Results:

Two-hundred ninety-six subjects were included. All patients had at least minimal VPI pre-operatively; 72% were graded moderate or severe. Sixty-four percent experienced resolution

and 83% improved at least one point on the VPI-severity scale. Of the five patient characteristics, only history of cleft palate repair was significantly associated with decreased odds of VPI improvement but not resolution when controlling for other variables.

Conclusions:

Sphincter pharyngoplasty resulted in resolution of VPI in 64% and improvement in 83% of subjects. Children with a history of cleft palate had significantly decreased odds of VPI improvement compared to those without a history of cleft palate. Neither syndrome diagnosis nor 22q11 deletion had a significant association with speech outcomes after sphincter pharyngoplasty.

Key Words: Sphincter pharyngoplasty, velopharyngeal insufficiency, cleft palate, speech

Level of Evidence: Level 4

Introduction:

Velopharyngeal dysfunction (VPD) refers to any structural or functional difference that impedes complete velopharyngeal port closure during speech production resulting in hypernasal resonance and nasal air escape. Normal speech production requires complete closure of the velopharyngeal port for all consonants except the nasal consonants (m, n and ŋ). VPD etiology is often multifactorial and may involve anatomic abnormalities (velopharyngeal insufficiency), neuromuscular dysfunction (velopharyngeal incompetence) and compensatory/maladaptive articulation patterns (velopharyngeal mislearning).[1] The impact of velopharyngeal insufficiency (VPI) on speech intelligibility depends on velopharyngeal gap-size, with greater VPI severity often associated with larger gap-sizes.[2] Speech intelligibility may be further impaired if there is concomitant velopharyngeal mislearning (VPM). Multiple studies have found associations between VPI and measures of decreased quality of life, including impaired scholastic, social, behavioral, emotional and physical function.[3–5]

While speech therapy may be helpful for patients with VPM, treatment of VPI requires physical management provided only by intra-oral appliances or surgical intervention.[6,7] Various surgical approaches can narrow the velopharynx, facilitating velopharyngeal closure and eliminating nasal air escape.[8] Posterior pharyngeal flap and sphincter pharyngoplasty (SP) are the most common pharyngeal procedures aimed at managing VPI,[7] the latter of which is more commonly performed at our institution. SP creates a physiologic sphincter at the level of the

velum by raising lateral, superiorly-based myomucosal flaps from the posterior-lateral oropharynx and rotating them medially and superiorly to the posterior nasopharynx, ideally to the level at which the velum attempts to contact the posterior pharyngeal wall.[9]

The definition of success after SP varies widely and depends on the outcome measured.[8,10–13] A previous small study from our institution found that 63% of patients had complete resolution of VPI after SP.[11] Studies investigating characteristics associated with surgical outcomes after SP have found that greater age and VPI severity, history of cleft palate, and syndromic presence may be associated with poorer surgical outcomes, though results have not been consistent across studies.[8,10,12,13] Thus, patient factors affecting outcomes of SP remain poorly understood.

The purpose of this study was to describe VPI outcomes in a large group of children after SP, and to identify patient characteristics associated with VPI resolution and improvement after surgery. We hypothesized that failure to resolve or improve would be associated with 1) a history of palatal cleft, including submucous cleft, 2) any syndromic diagnosis (versus nonsyndromic), 3) a diagnosis of 22q11 deletion (versus nonsyndromic) 4) previous palatoplasty performed specifically for VPI, and 5) large pre-operative velopharyngeal gap on endoscopy.

Methods:

This study was approved by the Seattle Children's Hospital (SCH) Institutional Review Board (STUDY15330).

Data Collection:

Patient demographic data, perceptual VPI severity ratings and nasendoscopy findings were collected in a standardized manner by the pediatric otolaryngologist and speech-language pathologist (SLP) as part of routine clinical care at each patient's VPI clinic visits. All clinical data was entered into a secure database. Data from patients who underwent SP for VPI between February 1994 and October 2016 were downloaded from the database and additional chart review was performed to fill in missing variables. All surgeries were performed by one of 3 surgeons. The database was cross-referenced with a database of the SCH Craniofacial Center to determine syndrome diagnosis and presence of 22q11 deletion. Syndrome diagnosis was subdivided into five categories: 22q11 deletion, neuromuscular, syndromic cleft palate, central nervous system, or other (Table 1). This variable was dichotomized in two different ways: syndrome versus non-syndromic and 22q11 deletion versus non-syndromic for analysis.

For children with multiple follow-up visits, we selected the follow-up visit closest to 6 months after SP; we excluded children without a follow-up visit greater than 3 months after surgery. We also excluded children who had palatoplasty on the same day as SP, those for whom SP was a revision, and those for whom pre-operative or post-operative speech perception data was unavailable. We included children with a history of cleft of the secondary

and/or primary palate or submucous cleft that had been repaired, or previous Furlow

palatoplasty[14] for VPI (defined as that performed over the age of 2.5 years specifically to treat

VPI), as well as those with non-cleft causes of VPI.

Clinical VPI Evaluation:

Patients were evaluated according to standard clinical care at our institution, including

pre- and post-operative perceptual speech evaluations aimed at differential diagnosis of VPD

etiology by an SLP specializing in VPD diagnosis and management. Although VPI severity was

the outcome measure of interest for this study, additional speech characteristics including

degree of VPM and severity of motor planning and motor execution deficits were also

evaluated. Overall VPI severity was defined as the degree of passive nasal air emission and

hypernasality, in the absence of maladaptive articulation, present in a variety of speech tasks.

In cases where etiology of VPD was multifactorial, presence of VPI and candidacy for surgical

management was confirmed through nasendoscopy. Patients whose velopharyngeal port was

consistently open for consonants requiring complete closure were considered candidates for

surgical management. Passive nasal air emission not attributable to oronasal fistula(e) was

considered in the VPI-severity rating. Baseline and post-operative VPI were graded by the

evaluating SLP on a 5-point scale from 0-none to 1-minimal, 2-mild, 3-moderate and 4-severe

using a standardized institutional rubric (Table 2).[15]

The evaluating otolaryngologist performed nasendoscopy on each patient prior to SP and graded velopharyngeal gap-size as none, small, moderate, or large as described previously.[16–19] Gap-size was dichotomized as none/small versus moderate/large for analysis.

Outcome Measures:

The primary outcome measure was post-surgical VPI resolution, defined as post-operative VPI severity of "0-none" or "1-minimal" *and* improvement of at least one point on the 5-point perceptual severity scale. Thus, children who had a pre-operative score of 1 were required to improve to 0 to be considered resolved. VPI improvement, defined as a post-operative decrease of at least one point on the VPI-severity scale, was used as a secondary outcome measure.

Statistical Analyses:

Statistical analysis was conducted with Stata/SE 15 software (StataCorp LP, College Station, Texas). An alpha level of .05 was used for all statistical tests.

For bivariate analyses, Pearson's Chi-squared statistic for categorical variables and the Wilcoxon Rank-Sum test for continuous variables were used to test associations between each baseline variable with the two outcome measures, VPI resolution and improvement.

Multivariable logistic regression using robust standard errors was then performed to examine associations between the five pre-selected baseline patient factors hypothesized to

decrease odds of resolution and improvement, (1) history of palatal cleft (including submucous cleft); 2) any syndromic diagnosis (versus nonsyndromic); 3) diagnosis of 22q11 deletion (versus nonsyndromic); 4) previous palatoplasty performed specifically for VPI; and 5) large velopharyngeal gap on pre-operativeendoscopy, while controlling for age, sex, race, and baseline VPI severity.

Results:

Three hundred sixty-three subjects underwent SP during the study period. Twenty-six were excluded for concomitant palatoplasty and 41 were excluded for missing pre- or post-operative perceptual speech data, resulting in 296 included subjects.

Baseline Characteristics:

Baseline characteristics as well as results from bivariate analyses are provided in Table 3. Subjects ranged from 2 to 23 years old at the time of SP. Median follow-up time was 6.7 months after surgery. The majority were Caucasian and just over half were male (58%). Most had either a history of repaired cleft of the secondary and/or primary palate (41%), or submucous cleft (13%). Just over one-fourth (27%) had undergone a previous Furlow palatoplasty for VPI. The majority (64%) had no syndromic diagnosis. The most common syndrome was 22q11 deletion (15%). Half had a small velopharyngeal gap on pre-operative

nasendoscopy. All patients presented with at least minimal preoperative perceptual VPI, although the majority had moderate (29%) or severe (43%) pre-operative VPI.

VPI Severity:

Distribution of VPI severity before and after surgery is shown in Figure 1. Nearly two-thirds (64%) of subjects presented with resolution of VPI after SP and 83% presented with VPI improvement.

Bivariate Analyses:

Results from bivariate analyses are provided in Table 3.

Of factors tested in bivariate analysis, only pre-operative VPI severity and history of palatoplasty for VPI, were significantly associated with VPI resolution, though there was a trend for decreased VPI resolution among those with a history of cleft palate. The proportion of those with complete resolution of VPI was highest among those with mild baseline VPI severity (82%) while those with moderate or severe baseline VPI severity demonstrated resolution 66% and 54% of the time, respectively.

Only baseline VPI severity and history of cleft palate repair were significantly associated with VPI improvement on bivariate analysis. Those with minimal pre-operative VPI severity experienced resolution at the lowest rates (14%). Notably, of the 7 patients with minimal pre-operative VPI, 5 (71%) had a history of cleft palate repair and 5 (71%) had a syndrome

diagnosis; 1 (14%) had a moderate pre-operative velopharyngeal gap-size with the remaining

having small gaps. Proportion of VPI improvement was similar among those with small, medium

and large gap-sizes with greater than 80% of patients improving in each group (81%, 84% and

85%, respectively).

VPI resolution and improvement rates were higher among those without history of

palatal cleft than those with history of cleft palate repair. Resolution was demonstrated in 69%

of patients without a history of palatal cleft, versus 59% with a history of palatal cleft.

Improvement was noted in 88% without history of palatal cleft versus 78% with history of cleft.

We found no significant association with syndromic presence or 22q11 deletion and rates of VPI

resolution or improvement.

Multivariable Analyses:

Multivariable logistic regression was performed to examine associations between

baseline factors hypothesized to be associated with VPI resolution or improvement while

controlling for age, sex, race and VPI severity. Results are provided in Tables 4 and 5.

No significant associations between odds of VPI resolution and baseline factors were

noted on multivariable analysis.

With regards to VPI improvement, those with a history of cleft palate repair were

approximately 50% less likely to demonstrate improvement in post-operative VPI severity than

those without history of cleft palate, controlling for possible confounders. No other variables were significantly associated with decreased odds of VPI improvement.

Discussion:

We present a large series of patients who underwent SP for management of VPI. Preoperatively, all patients had VPI on perceptual speech assessment and transverse orientation of the levator veli palatini based on endoscopic findings. The outcome measures were based upon standardized assessments of VPI severity determined by SLPs with expertise in VPI and collected as part of routine clinical care.

Overall, 64% of children had resolution and 83% had improvement of their VPI after SP. Except for those with minimal baseline VPI, lower baseline VPI severity and lack of prior palatoplasty were associated with greater rates of VPI resolution following SP. Lower baseline VPI severity and absence of a history of cleft palate repair were associated with a greater likelihood of VPI improvement post-operatively. However, only history of palatal cleft was significantly associated with lack of VPI improvement when controlling for other patient factors.

Comparing these results to other similar studies is limited by use of different outcome measures. Losken et al. published a series of 250 patients with previously repaired cleft palate who underwent SP for VPI, using need for revision as the primary outcome. Their overall

revision rate was 13%, 78% of whom underwent revision SP for persistent VPI following initial SP. They found higher rates of revision in those with larger velopharyngeal gap-size and more severe hypernasal resonance pre-operatively. Patients with 22q11 deletion syndrome and cleft palate also appeared to have higher revision rates, though these were reported as statistically nonsignificant.[8] Carlisle et al. examined a cohort of 46 patients who underwent SP for VPI using rate of revision surgery as a proxy for VPI. The study found a 13% revision rate and no association with age, sex, cleft type, or syndrome. Notably, 57% of patients in their cohort underwent concomitant Furlow palatoplasty.[10] The revision rate was 25% in those who underwent SP alone.[10] Samoy et al. investigated speech outcomes of 62 patients who underwent VPI surgery with either SP or velopharyngoplasty. They also found no association with age, but did note poorer outcomes in patients with 22q11 deletion syndrome and those with more severe hypernasality.[13]

We sought to understand the outcome of SP and therefore excluded patients who had concomitant FP and SP. At our institution, combined FP and SP is typically offered to patients with hypodynamic velopharynx and evidence of sagittal orientation of the levator veli palatini. Therefore, the exclusion of these patients may have biased the outcomes reported, particularly for those patients with large gaps. Riski et al. described a series of 139 patients using nasal emissions on perceptual exam or nasal air escape on pressure-flow studies as a combined outcome measure. They found a success rate of 78%, with greater success in younger patients

and those with less pre-operative hypernasality but no association with velopharyngeal gap-size, type of cleft, or sex.[12] With regards to the association with age, it was hypothesized that "ingrained speech habits" (VPM) were to blame for lower success rates in older patients.[12] Studies have shown that velopharyngeal closure is different when using accurate oral placement for consonants compared to maladaptive articulation (i.e. glottal stops, pharyngeal fricatives, etc.).[20] Presumably, if accurate differential diagnosis of etiology of VPD is not made, VPI surgery may be tailored toward a gap size indicative of maladaptive articulation rather than gap size indicative of VPI. Accurate differential diagnosis of VPD vs. VPI vs. VPM is a primary focus of perceptual speech evaluations and care is taken to rate VPI severity based on appropriate speech characteristics. Thus our results may not be comparable to studies in which different methods are used for VPI diagnosis. The American Cleft Palate – Craniofacial Association developed the *Standards for Cleft Palate and Craniofacial Teams*, mandating the inclusion of SLPs among the core members of multidisciplinary craniofacial teams.[21] SLPs on these teams play a vital role in the assessment and management of patients with VPI. Like the present study, more recent studies likely include greater SLP involvement, and found no association between age at surgery and VPI outcomes.[8,10,13]

We found that patients with previous cleft palate repair were less likely to have improvement of VPI after SP. The previously repaired cleft palate group represents a very heterogenous population and it is likely that multiple factors contributed to this result. It is also

possible that patients with previous palate repair may have had more palatal scarring associated with less palatal mobility.

Evidence as to whether syndromic diagnoses are associated with poorer outcomes following SP is conflicting. While an association was suggested by Losken et al and Samoy et al, the present study, in which 39% carry a syndromic diagnosis, found no association.[8,13] In this study, 22q11 deletion syndrome represented 15% of the study population. We did not find a significant association between 22q11 deletion and the odds of VPI resolution or VPI improvement when controlling for age, sex, race, and baseline VPI severity. We had postulated that patients with deletion 22q11 and large gaps may have had combined Furlow palatoplasty and sphincter pharyngoplasty and were thereby excluded from this dataset. However, upon review of our data, only one patient with 22q11 deletion syndrome was excluded for undergoing simultaneous SP and Furlow palatoplasty.

The low rates of VPI resolution and improvement among patients with pre-operative minimal VPI was remarkable. At our institution, minimal VPI is assigned to those with nasal air escape detected by auscultation only. The low rates of resolution may reflect greater degree of neuromuscular discoordination contributing to VPI, as opposed to a predominantly structural or anatomic cause. The role for surgical intervention in this group should be carefully considered based upon these findings.

Median follow-up time after SP was 6.7-months. In order to improve data uniformity, data from the follow-up visit closest to 6 months was used for patients who had multiple follow-up visits. We selected this timepoint since patients that have VPI persisting beyond 6 months will usually be considered for revision surgery. The 6 month timepoint occurs after our standard 3 month instrumental speech assessment[6] but before 10 months, the reported mean time interval to revision SP.[8] It is important to point out however, that changes to the surgically manipulated tissues may continue beyond this point as surgical site healing and scarring progress.

Like other studies, the present investigation found a statistically significant association between baseline VPI severity and post-operative VPI resolution, with greater odds of resolution associated with milder baseline VPI, on bivariate analysis.[8,12,13] These findings are anticipated as one would expect those with less severe VPI to require less tissue volume in the transposition flaps. An association between greater velopharyngeal gap-size and decreased rate of VPI resolution would support this argument, and this was indeed demonstrated by Losken et al.[8] In the present study, while a tendency for children with larger velopharyngeal gap-size to have reduced VPI resolution rates was found, it was not statistically significant. These findings may have been affected by exclusion of those patients who had concomitant FP and SP.

This study has several other limitations, primarily those related to the retrospective nature of the research design and subjective nature of perceptual speech assessments. The

outcome measures in this study were limited to objective measures of (VPI resolution and improvement). While syndrome diagnosis was determined by a craniofacial pediatrician, we did not routinely test all patients for 22q11 deletion. Therefore, under-reporting of patients in this group is possible.

Conclusion:

In this study, SP is associated with VPI resolution in 64% and improvement in 83% of patients. Neither VPI resolution nor improvement were significantly associated with a history of syndrome (whether 22q11 or other syndromic diagnosis), history of previous palatoplasty for VPI, or large velopharyngeal gap-size. History of palatal cleft was significantly associated with decreased odds of VPI improvement but not resolution, independent of age, sex, race, and pre-operative VPI severity.

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Figure Legends:

Figure 1.

Figure 1.   Perceptual VPI severity at baseline and following sphincter pharyngoplasty.

VPI=velopharyngeal insufficiency
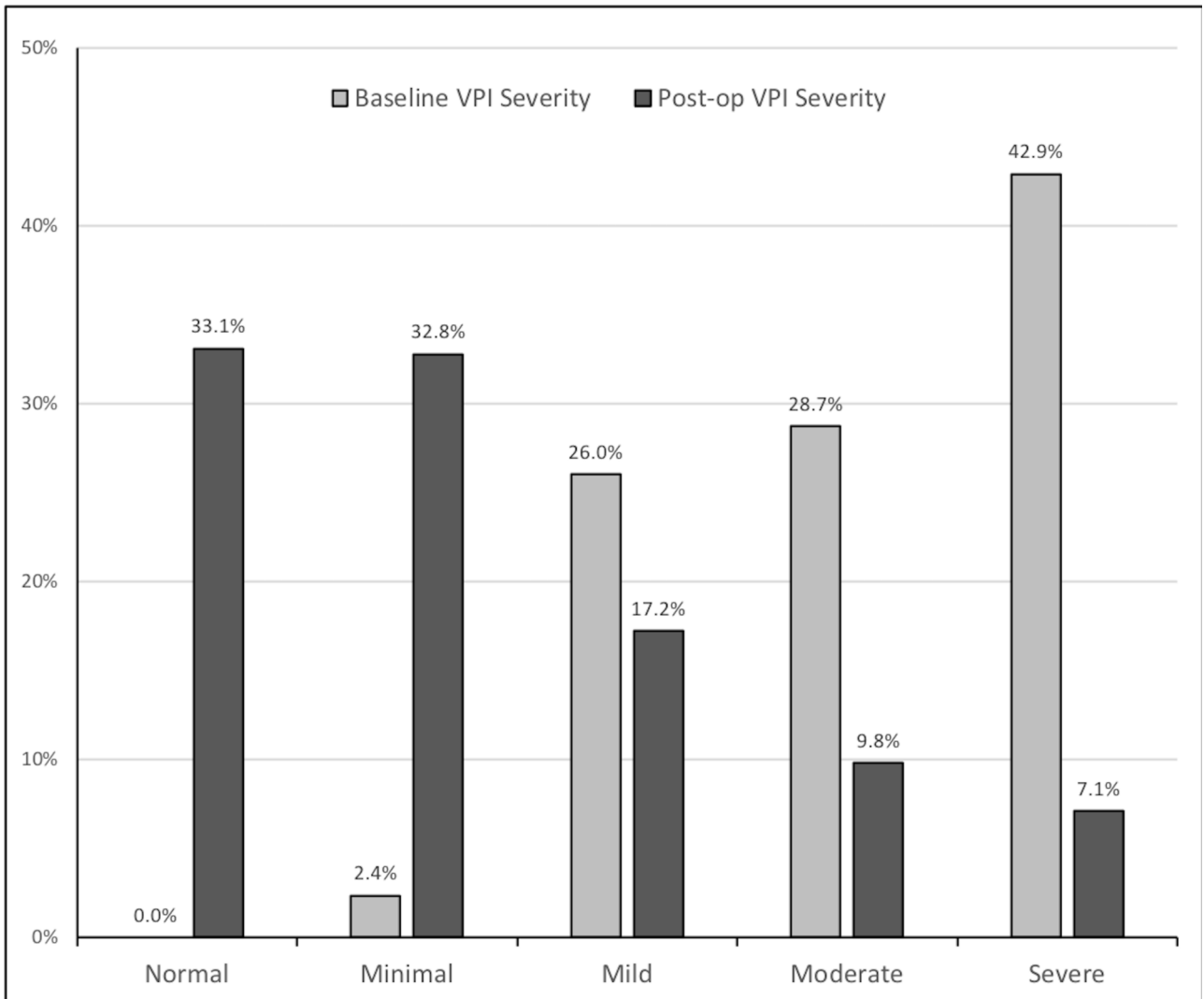
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Table 1. Categorization of syndromic diagnoses.

Syndrome Category	Included Diagnoses
22q11 Deletion	velocardiofacial, DiGeorge
Neuromuscular	myopathy, Moebius, pseudobulbar palsy
Syndromic Cleft Palate	Hays Wells, ectrodactyly-ectodermal dysplasia, Kabuki, Stickler, Pierre-Robin, and Van de Woude
Central Nervous System	history of encephalitis, brain tumor, hypotonia, cerebral palsy or traumatic brain injury
Other	amniotic band syndrome, arthrogryposis, CHARGE, congenital hypothyroidism, frontonasal dysplasia, Klippel Feil, macro/microcephaly, craniofacial microsomia, Down Syndrome, neurofibromatosis, Pallister-Hall, in-utero drug exposure

Table 2. Velopharyngeal insufficiency-severity rubric.

VPI Severity	Nasal Airflow (*Nasal Air Escape/Nasal Turbulence*)	Resonance (*Hypernasality*)
0 – None	Absent (normal intra-oral pressure for all pressure consonants)	Absent (balanced oral-nasal resonance)
1 – Minimal	Absent or on auscultation only[†]	Borderline/minimal (present only in nasal contexts)
	Occasional (infrequently heard)	Absent
2 – Mild	Absent or occasional	Mild (evident on high vowels only - /i, ɪ/)
3 – Moderate	Occasional or frequent (consistently heard)	Moderate (evident on high & mid-vowels - /u/)
4 – Severe	Frequent	Severe (present on all vowels and on voiced consonants)

VPI=velopharyngeal insufficiency

[†] Heard using listening tube.

Table 3. Characteristics of children overall and by velopharyngeal insufficiency outcome after sphincter pharyngoplasty.

Characteristic	All Subjects* (N=296)	Improved (n=245)	p	Resolved (n=189)	p
		VPI Outcome			
		Median (Interquartile Range) or n (%)			
Age at surgery (years)[†]	7 (5-11)	7 (5-11)	0.9	7 (5-10)	0.9
Follow up time (months)[†]	6.7 (4.4-13.1)	6.7 (4.4-13.0)	0.3	6.7 (4.5-13.3)	0.6
Male[‡]	171 (58)	142 (58)	0.9	105 (56)	0.3
Race/ethnicity[‡]			1.0		0.2
White	217 (73)	180 (74)		143 (76)	
Non-white	77 (26)	64 (26)		45 (24)	
Black	4 (1)	2 (1)		2 (1)	
Asian/Pacific Islander	22 (7)	19 (8)		11 (6)	
Hispanic	22 (7)	17 (7)		13 (7)	
Other	29 (10)	26 (11)		19 (10)	
Syndrome/co-morbidity[‡]			0.8		0.8
None	189 (64)	157 (65)		119 (64)	
Any syndrome below	105 (35)	86 (35)		68 (36)	
22q11deletion	45 (15)	40 (16)		31 (17)	
Neuromuscular	5 (2)	5 (2)		5 (3)	
Syndromic cleft	27 (9)	19 (8)		14 (7)	

CNS	8 (3)	6 (2)		5 (3)	
Other	20 (7)	16 (7)		13 (7)	
History of palatal cleft[‡]			*0.02*		0.07
None	137 (46)	121 (49)		95 (50)	
Palatal cleft	159 (54)	124 (51)		94 (50)	
Hard/soft palatal cleft	121 (41)	95 (44)		71 (43)	
Submucous only	38 (13)	29 (14)		23 (14)	
Previous palatoplasty for VPI[‡]	80 (27)	65 (27)	0.7	44 (23)	*0.05*
Gap size (endoscopy)[‡]			0.8		0.2
Small	148 (50)	120 (51)		102 (56)	
Moderate	106 (36)	89 (38)		61 (33)	
Large	33 (11)	28 (12)		20 (11)	
Baseline VPI severity[‡]			*<0.001*		*<0.001*
Minimal	7 (2)	1 (<1)		1 (1)	
Mild	77 (26)	63 (26)		63 (33)	
Moderate	85 (29)	72 (29)		56 (30)	
Severe	127 (43)	109 (44)		69 (37)	

VPI=velopharyngeal insufficiency, CNS=central nervous system

[†] Wilcoxon Rank-Sum test used.

[‡] Pearson's Chi-squared statistic used.

Medians are presented for continuous variables, with interquartile ranges in parenthesis. For

categorical variables, the number of patient's are presented, with percentages in parenthesis.

p – values refer to association between the given characteristic and either VPI improvement or

VPI resolution.

Column percentages may not total 100 due to missing data.

Table 4. Multivariable logistic regression of the association between baseline patient factors and velopharyngeal insufficiency _resolution_ controlling for age, sex, race (white vs. non-white), baseline VPI severity.

	Odds Ratio (95% Confidence Interval)	P value
History of cleft palate[†]	0.68 (0.41 – 1.12)	0.1
Any syndrome (vs. nonsyndromic)	1.04 (0.62 – 1.73)	0.9
22q11 deletion (vs. nonsyndromic)	1.18 (0.57 – 2.43)	0.7
Previous palatoplasty for VPI	0.65 (0.37 – 1.14)	0.1
Velopharyngeal gap size[‡]	0.74 (0.43 – 1.30)	0.3

VPI=velopharyngeal insufficiency

[†] Any cleft, including submucous cleft

[‡] Moderate/large vs. small

Table 5. Multivariable logistic regression of the association between baseline patient factors and velopharyngeal insufficiency *improvement* controlling for age, sex, race (white vs. non-white), baseline VPI severity.

	Odds Ratio (95% Confidence Interval)	*P* value
History of cleft palate[†]	0.46 (0.24 – 0.91)	***0.03***
Any syndrome (vs. nonsyndromic)	0.92 (0.48 – 1.74)	0.8
22q11 deletion (vs. nonsyndromic)	1.55 (0.55 – 4.38)	0.4
Previous palatoplasty for VPI	0.73 (0.35 – 1.53)	0.4
Velopharyngeal gap size[‡]	0.74 (0.38 – 1.44)	0.4

VPI=velopharyngeal insufficiency

[†] Any cleft, including submucous cleft

[‡] Moderate/large vs. small