# Speech Outcome after Palatal Repair in Nonsyndromic versus Syndromic Robin Sequence

Kamlesh B. Patel, M.D.
Stephen R. Sullivan, M.D., M.P.H.
Ananth S. Murthy, M.D.
Eileen Marrinan, M.S., M.P.H.
John B. Mulliken, M.D.

*Boston, Mass.*

**Background:** The authors' purpose was to document speech outcome after cleft palate repair in patients with syndromic versus nonsyndromic Robin sequence. Results of secondary correction of velopharyngeal insufficiency using a superiorly based pharyngeal flap or double-opposing Z-palatoplasty are also reported.

**Methods:** Charts of patients with Robin sequence and cleft palate between 1980 and 2007 were reviewed. Data collected included date of birth, sex, syndrome/association, cleft palatal type (Veau I or II), age at palatoplasty, incidence of palatal fistula, postoperative speech assessment, videofluoroscopic results, need for secondary operation for velopharyngeal insufficiency, and type of secondary operation (pharyngeal flap or double-opposing Z-palatoplasty).

**Results:** The authors identified 140 patients with Robin sequence who had palatal closure. Postoperative speech evaluation was available for 96 patients (69 percent). A syndrome or association was identified in 42 patients (30 percent). Primary palatoplasty was successful in 74 patients (77 percent); speech was characterized as competent and competent to borderline competent. The authors found a significantly higher incidence of velopharyngeal insufficiency following palatal repair for syndromic (38 percent) than nonsyndromic Robin sequence (16 percent). ($p$ = 0.039). In patients with velopharyngeal insufficiency, competent or borderline competent speech was determined after double-opposing Z-palatoplasty (two of five patients) or pharyngeal flap (eight of 10 patients).

**Conclusions:** The rate of velopharyngeal insufficiency in syndromic Robin sequence is significantly greater than in nonsyndromic Robin sequence. The authors prefer pharyngeal flap for velopharyngeal insufficiency in patients with Robin sequence, whether syndromic or nonsyndromic, without retrognathism or signs/symptoms of obstructive sleep apnea. (*Plast. Reconstr. Surg.* 130: 577e, 2012.)

**CLINICAL QUESTION/LEVEL OF EVIDENCE:** Risk, II.


Pierre Robin is traditionally credited as being the first to describe the clinical triad of micrognathia, glossoptosis, and upper airway obstruction.[1] This condition was called "Pierre Robin syndrome" for 50 years, and relabeled "Robin anomalad" for a brief time.[2] *Robin sequence* is a more precise term. The surname (e.g., Pierre) is not ordinarily used in medical eponyms. This developmental disorder is now thought to be initiated by mandibular undergrowth or retropositioning and is known to be pathogenically heterogeneous.[3] There is an associated syndrome in 34 to 46 percent of patients with Robin sequence.[4-7] These syndromes can be either monogenic (Stickler), chromosomal (deletion 4q), teratogenic (fetal alcohol), sporadic (hemifacial microsomia), or disruptive (amniotic

*From the Craniofacial Center and Department of Plastic and Oral Surgery, Children's Hospital and Harvard Medical School.*
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band).[8,9] It is clinically useful to categorize Robin sequence as either syndromic or nonsyndromic.

Cleft palate is present in up to 90 percent of patients with Robin sequence, although it is not an obligatory finding.[10] In nonsyndromic Robin sequence, the palatal processes are presumed to be intrinsically normal; the cleft is the result of an obstruction of fusion (8 to 12 weeks of gestation) caused by the superoposteriorly positioned tongue. A particular molecular alteration in syndromic Robin sequence might interfere with palatal fusion and elevation, such as collagen mutations in Stickler syndrome, hypotonia in velocardiofacial syndrome, neuromuscular hemihypoplasia in hemifacial microsomia, mandibular hypoplasia in Treacher Collins syndrome, or palatal agenesis in Nager syndrome.[11]

Our purpose is to characterize the outcome of palatal repair in Robin sequence, comparing syndromic to nonsyndromic patients. Our hypothesis is that speech would be better in children with nonsyndromic Robin sequence (because the palatal shelves are intrinsically normal) than in those with syndromic Robin sequence (in which the palatal shelves are developmentally malformed). We also documented the outcome of secondary correction of velopharyngeal insufficiency using a superiorly based pharyngeal flap versus double-opposing Z-palatoplasty.

## PATIENTS AND METHODS

After approval by the Committee on Clinical Investigation, we identified and reviewed the charts of all patients with a diagnosis of Robin sequence and cleft palate from 1980 to 2007. All operations were performed by the senior author (J.B.M.). We included only those patients who were at least 4 years of age at the time of review because children are more cooperative for speech assessment and unlikely to subsequently develop velopharyngeal insufficiency after that age. Data were collected for date of birth, sex, syndrome/association, palatal cleft type (Veau I or II), age at palatoplasty, palatal fistula, postoperative speech, videofluoroscopy, secondary operation because of velopharyngeal insufficiency, type of secondary operation (double-opposing Z-palatoplasty or pharyngeal flap), and interval to most recent evaluation. Operative techniques were as described previously.[12-14]

### Speech Assessment

Patients were followed annually in the cleft lip–cleft palate clinic; the senior author examined all patients. A speech pathologist, specializing in cleft palate, performed preoperative and postoperative perceptual assessments and scored the results using the Pittsburgh Weighted Values for Speech Symptoms Associated with Velopharyngeal Incompetence instrument.[15,16] Overall assessment of speech was graded as follows: 0, competent velopharyngeal mechanism; 1 to 2, competent to borderline competent; 3 to 6, borderline to borderline incompetent; and greater than or equal to 7, incompetent velopharyngeal mechanism. Competent and competent to borderline competent was categorized as a success, whereas borderline to borderline incompetent and incompetent were categorized as failures requiring secondary surgery (double-opposing Z-palatoplasty or pharyngeal flap).

Speech outcome following a secondary operation was documented. The need for a revisionary operation (e.g., postoperative tonsillectomy, adenoidectomy, flap division, or dilation of pharyngeal ports) to correct airway obstruction following pharyngeal flap was also recorded. Polysomnography was conducted if a child evidenced signs or symptoms of obstructive sleep apnea.

### Statistical Analyses

Patient characteristics and descriptive statistics were summarized and compared between syndromic and nonsyndromic patients. Continuous data were expressed as mean ± SD and compared with the $t$ test, and proportions were analyzed using Fisher's exact test. We used logistic regression to evaluate the relationship between age at palatoplasty and success or failure of palatoplasty. All calculated $p$ values were two-tailed and considered significant for values of $p < 0.05$. Statistical analyses were performed using Stata version 10.0 (StataCorp, College Station, Texas).

## RESULTS

### Patient Characteristics

We identified 140 patients with Robin sequence who had palatoplasty performed by the senior author. Postoperative follow-up assessment was available for 127 patients (91 percent), whereas postoperative speech evaluation with a complete speech score was available for 96 patients (69 percent). Patient characteristics are listed in Table 1. A syndrome or association was identified in 42 patients (30 percent) (Table 2).

### Speech Outcome

Successful palatoplasty was confirmed in 74 patients (77 percent); secondary operation was

**Table 1. Patient Characteristics**

| | All Patients (%) | Nonsyndromic Patients (%) | Syndromic Patients (%) | *p** |
|---|---|---|---|---|
| No. of patients | 96 | 67 | 29 | |
| Age at palatoplasty, mo | | | | |
| Mean | 11.0 | 10.1 | 13.0 | 0.23 |
| Range | 7.0–74.2 | 7.0–20.2 | 8.5–74.2 | |
| Male-to-female ratio | 48:48 (50:50) | 32:35 (48:52) | 16:13 (55:45) | 0.66 |
| Veau cleft type | | | | 0.40 |
| I | 18 (19) | 11 (16) | 7 (24) | |
| II | 78 (81) | 56 (84) | 22 (76) | |
| Interval to most recent follow-up after palatoplasty, yr | 7.2 | 6.9 | 7.6 | 0.22 |

*Continuous data were compared with the *t* test assuming unequal variances, and proportions were analyzed using Fisher's exact test.

**Table 2. Robin Sequence Diagnoses**

| | No. (%) |
|---|---|
| Total | 140 (100) |
| Nonsyndromic | 98 (70) |
| Syndromic | 42 (30) |
| Stickler | 19 |
| Unknown syndrome | 4 |
| Treacher Collins | 3 |
| Cornelia de Lange | 3 |
| Van der Woude | 2 |
| Velocardiofacial | 1 |
| Diamond-Blackfan anemia | 1 |
| Hemifacial microsomia | 1 |
| CHARGE association | 1 |
| Gordon | 1 |
| Carey-Fineman-Ziter | 1 |
| Ehlers Danlos | 1 |
| Dravet | 1 |
| Beckwith-Wiedemann | 1 |
| Seckel | 1 |
| Chromosome 16P11.2 deletion | 1 |

recommended in 22 patients (23 percent). We found a significant difference in the incidence of borderline to borderline incompetent or incompetent speech between patients with syndromic versus nonsyndromic Robin sequence ($p = 0.039$) (Fig. 1). Stickler syndrome was the most common associated condition [19 of 42 patients (45 percent)], with three of 12 (25 percent) patients developing velopharyngeal insufficiency.

The relationship between speech outcome and age at palatoplasty is shown in Figure 2 ($p = 0.13$). Four of seven patients (57 percent) who underwent palatoplasty when older than 15 months of age were noted to have velopharyngeal insufficiency, whereas when palatal closure was performed before 15 months of age, 18 of 89 (20 percent) developed velopharyngeal insufficiency. No significant difference in speech outcome was found between Veau I and II cleft palate ($p = 1.0$) (Fig. 3).

### Palatal Fistula

Postoperative palatal fistula occurred in eight patients (6.3 percent), all of whom had a Veau II cleft palate. These fistulas were slit-like or pinhole at the junction of the hard and soft palate. Three of the eight patients were syndromic (Stickler, Dravet, and Seckel). In eight patients with a Veau II cleft, a vomerine flap could not be used for

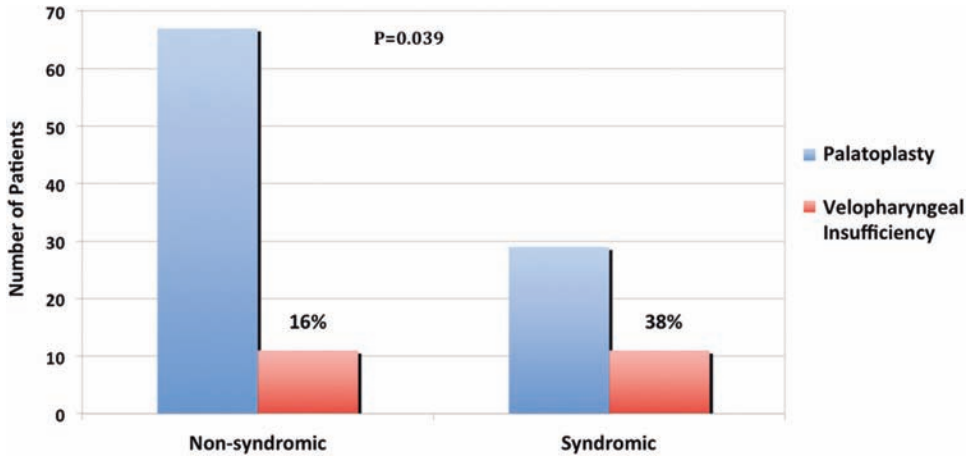


**Fig. 1.** Nonsyndromic versus syndromic diagnosis and incidence of velopharyngeal insufficiency. A higher proportion of syndromic patients [11 of 29 patients (38 percent)] failed palatoplasty when compared with nonsyndromic patients [11 of 67 patients (16 percent)] ($p = 0.039$).

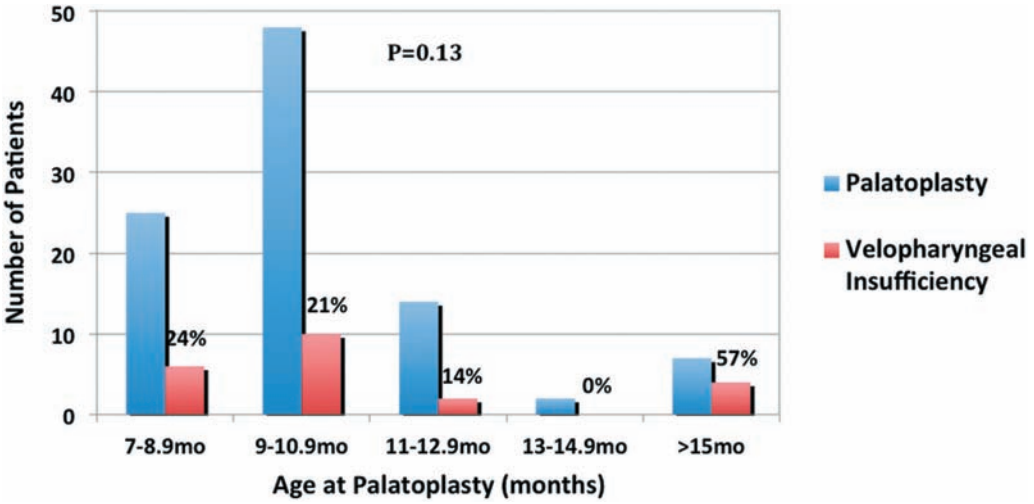


**Fig. 2.** Age at palatoplasty compared with patients who developed velopharyngeal insufficiency ($p = 0.13$). Patients who had palatoplasty at greater than 15 months of age were found to have a higher incidence of velopharyngeal insufficiency (57 percent versus 20 percent).

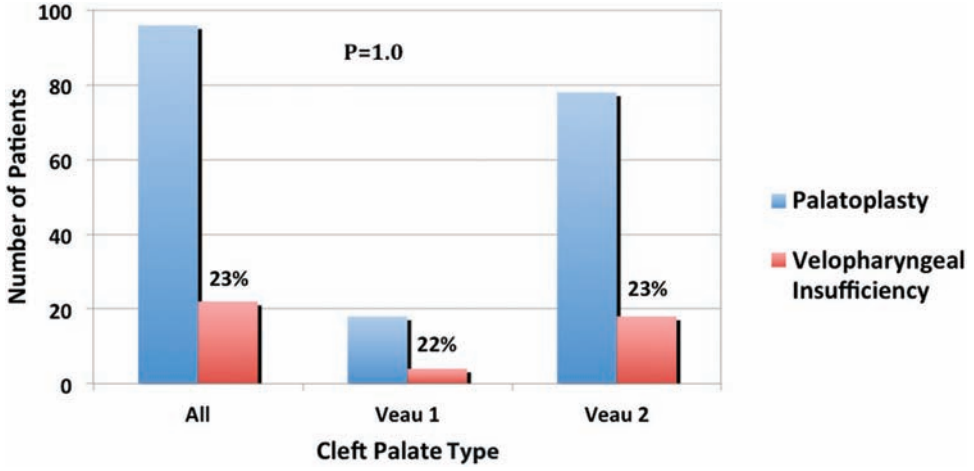


**Fig. 3.** Veau cleft type compared with velopharyngeal insufficiency. No association was identified between Veau classification and poor speech ($p = 1.0$).

nasal closure [three syndromic (7 percent) and five nonsyndromic patients (6 percent)], because the vomer was cephalad and diminutive; nevertheless, none of these patients had a fistula.

### Secondary Operations

Six patients with velopharyngeal insufficiency, three of whom were syndromic (Stickler and two unknown disorders), were managed with double-opposing Z-palatoplasty at a median age of 8.5 years (range, 5.0 to 12.9 years). Postoperative speech evaluation was available for five of six patients; two (unknown syndromes) had normal speech, whereas three patients (one Stickler and two nonsyndromic), although improved, had persistent velopharyngeal insufficiency.

Ten patients with velopharyngeal insufficiency (four syndromic and six nonsyndromic) were managed with a pharyngeal flap. The median preoperative lateral pharyngeal wall motion was 70 percent (range, 20 to 90 percent). Median age at surgery was 6.4 years (range, 4.8 to 12.2 years). Velopharyngeal function was normal or borderline competent in eight of 10 patients. One nonsyndromic patient had persistent borderline insufficiency, but had improved weighted speech score from 20 to 6. Another patient with Dravat syndrome continued to have borderline to borderline incompetent speech (weighted speech score from 10 to 4). All pharyngeal flaps healed without complications, and none of the patients developed signs or symptoms of obstructive sleep apnea.

## DISCUSSION

### Definitions

Different definitions of Robin sequence alter the frequencies of syndromic and nonsyndromic patients and result in conflicting speech outcomes.[17,18] Breugem and Courtemanche queried cleft palate teams about diagnostic criteria for Robin sequence and identified 14 different classifications; the most common were micrognathia, cleft palate, and glossoptosis.[18] In our unit, we use the criteria micrognathia, glossoptosis, and respiratory distress, as originally described by Robin.[1]

In syndromic Robin sequence, there can be several different for airway obstruction, leading to dissimilar incidences among cleft centers.[17] Our cohort of patients exemplifies the etiologic and pathogenic heterogeneity in syndromic Robin sequence.[2] For example, syndromic Robin sequence can be the result of intrinsic mandibular hypoplasia (Treacher Collins and hemifacial microsomia), connective tissue dysplasia (Stickler syndrome), retrognathia caused by obtuse cranial base angle (velocardiofacial syndrome), extrinsic disruption of mandibular growth (oligohydramnios), or diminished mandibular movement (congenital hypotonia).[9,19]

### Speech Outcome

The overall incidence of velopharyngeal insufficiency following palatoplasty in our series of patients with Robin sequence is comparable to those in three reports (29.3 to 34.6 percent).[4,6,20] We found the rate of velopharyngeal insufficiency for syndromic Robin sequence to be significantly greater than for nonsyndromic Robin sequence, contrary to two of these reports (Table 3).[4,6] Witt and colleagues assessed their rate of velopharyngeal insufficiency in syndromic and nonsyndromic Robin sequence to be 8 and 44 percent, respectively.[4] They designated their syndromic category as composed of two major or three minor associated malformations that are not explained on a familial basis. In contrast, we defined syndromic Robin sequence as a patient with multiple anomalies thought to have a shared pathogenesis.[21] Thus, Witt and colleagues excluded patients with major craniofacial malformations, such as hemifacial microsomia and Treacher Collins syndrome, as constituting syndromic Robin sequence. However, they included patients with myelomeningocele, autism, and club feet as being syndromic. Similarly, de Buys Roessingh and colleagues reported a lower rate of velopharyngeal insufficiency in syndromic (23 percent) versus nonsyndromic (36 percent) Robin sequence patients.[6] The most likely explanation for this discrepancy is their criteria used to diagnose Robin sequence. They defined Robin sequence as microretrognathia, glossoptosis, and cleft palate.

Stickler syndrome is the most common recognized condition associated with Robin sequence.[5,17,22] The incidence of velopharyngeal insufficiency is less in patients with Stickler syndrome (25 percent) compared with other types of syndromic Robin sequence (44 percent), but was not statistically significant ($p = 0.25$). Facial abnormalities (midfacial deficiency, small mandible, or cleft palate) are noted in 84 percent of patients with Stickler syndrome; however, they often do not exhibit feeding or airway problems.[5,22,23] Perhaps maxillary hypoplasia in Stickler syndrome results in a smaller resting velopharyngeal gap and explains the lower incidence of velopharyngeal insufficiency compared with other patients with syndromic Robin sequence.[24]

Isolated cleft palate (i.e., in the absence of an associated sequence or syndrome) is caused by an intrinsic defect in the developing maxillary-palatine shelves. In contrast, in nonsyndromic Robin sequence, it is believed that the superiorly retropositioned tongue prevents fusion of presumably normal palatal processes. Therefore, patients with nonsyndromic Robin sequence may have better speech outcomes following palatal repair than children with isolated (nonsyndromic) cleft palate. Khosla and colleagues found a lower incidence of velopharyngeal insufficiency in patients with nonsyndromic Robin sequence (8.8 percent) compared with patients with isolated cleft palate (13.6 percent), but the difference was not statistically significant.[25] In contrast, Witt and colleagues reported a higher rate of velopharyngeal

**Table 3. Speech Outcome in Patients with Robin Sequence**

| | No. of Patients (%) | | | No. of Patients with Velopharyngeal Insufficiency (%) | | |
|---|---|---|---|---|---|---|
| Authors | Total | Nonsyndromic | Syndromic | Total | Nonsyndromic | Syndromic |
| Patel et al. (present study) | 96 | 67 (70) | 29 (30) | 22 (23) | 11 (16) | 11 (38) |
| Witt et al., 19974 | 58 | 34 (59) | 24 (41) | 17 (29) | 15 (44) | 2 (8) |
| De Buys Roessingh et al., 2008[6] | 38 | 25 (66) | 13 (34) | 12 (32) | 9 (36) | 3 (23) |

insufficiency in patients with nonsyndromic Robin sequence (44 percent) versus isolated cleft palate (18 percent).[4] They proposed that the wide U-shaped cleft, as often portrayed in children with Robin sequence, might result in a short and immobile velum following repair. Of note, Rintala and colleagues documented that the incidence of a U-shaped cleft palate is the same in Robin sequence as in isolated cleft palate.[26]

The senior author and colleagues previously reported a 6.9 percent incidence of velopharyngeal insufficiency in 449 patients with isolated (nonsyndromic) Veau I and II cleft palate.[12] Thus, our hypothesis of better speech outcome following repair of Veau I/II cleft palate in children with nonsyndromic Robin sequence compared with children with isolated (nonsyndromic) cleft palate is untenable. Microscopic evaluation of isolated (nonsyndromic) cleft palatal muscle to noncleft specimens has shown decreased muscle mass, fiber size, and increased variability in fiber type.[27,28] This primary muscular abnormality could explain the irreducible incidence of velopharyngeal insufficiency after palatoplasty, regardless of the method of closure. There are no published histologic comparisons between palatal muscle in nonsyndromic or syndromic Robin sequence and isolated cleft palate.

A higher rate of velopharyngeal competence in Veau I compared with Veau II cleft palate has been reported in patients with an isolated cleft palate.[12,29,30] Nevertheless, we found no correlation in incidence of velopharyngeal insufficiency with Veau categories in our study. This finding is further evidence that there is likely intrinsic deficiency in cleft musculature in patients with Robin sequence, leading to similar rates of velopharyngeal insufficiency in Veau I and II cleft palate.

**Fistula Rate**

The frequency of palatal fistula in our series (6.3 percent) was lower than in other reports for Robin sequence (11.8 to 15.5 percent)[4,20] but higher compared with our previous report on isolated (nonsyndromic) cleft palate (2.9 percent).[12] Increased width of the cleft and hypoplastic palatal musculature probably explains the higher fistula rate. All fistulas were in Veau II clefts, where the cleft would be expected to be wider than in Veau I. In our patients with Robin sequence, we often noted that the vomer is diminutive, cranially displaced, and sometimes not available for construction of vomerine nasal lining flaps, in contrast to patients with isolated (nonsyndromic) cleft palate.

**Velopharyngeal Insufficiency**

Treatment of postpalatoplasty velopharyngeal insufficiency continues to be debated. Furlow recommends the double-opposing Z-palatoplasty for velopharyngeal insufficiency in patients with a small preoperative active pharyngeal gap (velar closing ratios greater than 80 percent).[31] Success rates, with resolution of velopharyngeal insufficiency, have been reported as high as 73 percent, and improvement in speech has been reported in 87 percent, depending on pharyngeal gap size in nonsyndromic cleft palate.[32] We used double-opposing Z-palatoplastic lengthening in a small number of patients with Robin sequence who had a small velopharyngeal gap and an increased likelihood for postoperative obstructive sleep apnea caused by persistent micrognathia (failure of "catch-up" growth). The double-opposing Z-palatoplasty corrected velopharyngeal insufficiency in two of five patients; thus, we cannot make definitive conclusions because of the small number of patients.

The superiorly based pharyngeal flap is the senior author's first-line method for correction of velopharyngeal insufficiency. We reported a 97 percent success rate for nonsyndromic isolated cleft palate.[14] Patients with Robin sequence, particularly if syndromic, might be expected to be at increased risk for obstructive sleep apnea following the pharyngeal flap procedure, as documented in a previous small case series.[33] Nevertheless, we believe the risk of sleep apnea after pharyngeal flap surgery is the same in patients with or without Robin sequence (syndromic or nonsyndromic), as anatomically the pharyngeal flap and the tongue base are at different levels in the oropharynx. Lehman and colleagues reviewed 23 patients with Robin sequence (12 were syndromic).[20] Of these six patients who had a pharyngeal flap, one required flap division because of sleep apnea. De Buys Roessingh and colleagues performed 12 pharyngeal flaps in 38 Robin sequence patients, and all 12 patients were reported to have good speech outcome without sleep apnea.[6] Likewise, none of the 10 patients with Robin sequence (four syndromic and six nonsyndromic) in our series had clinical signs or symptoms suggesting obstructive sleep apnea after pharyngeal flap.

In summary, we favor pharyngeal flap for velopharyngeal insufficiency in patients with Robin sequence without a retruded mandible or airway

evidence of obstructed sleep apnea. Another option is double-opposing Z-palatoplasty for the patient with documented obstructive sleep apnea and a small velopharyngeal gap size. If there is velopharyngeal insufficiency and obstructive sleep apnea caused by microretrognathia (usually, syndromic patients), mandibular advancement can be executed before double-opposing Z-palatoplasty or pharyngeal flap. Other alternatives to pharyngeal flap are palatal re-repair, radical dissection and retropositioning of velar muscles, or the double-opposing buccal flap for palatal lengthening.[34,35]

### Study Limitations

Complete speech evaluation with calculated speech score was not available for all patients. Although our speech pathologists specialize in cleft abnormality, more than one speech pathologist evaluated patients in this series. Speech pathologists can vary in their description and scoring of resonance, nasal emission, and intraoral pressure.

***John B. Mulliken, M.D.***
Department of Plastic and Oral Surgery
Children's Hospital
300 Longwood Avenue
Boston, Mass. 02115
john.mulliken@childrens.harvard.edu

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