RESULTS OF FURLOW Z-PLASTY IN PATIENTS WITH VELOCARDIOFACIAL SYNDROME

Sir:

Recently D'Antonio et al.[1] reported changes in velopharyngeal anatomy and function after Furlow Z-plasty for the treatment of velopharyngeal insufficiency. Specifically, the report documented increases in velar length and velar thickness following the procedure, and these changes were associated with improved velopharyngeal function. Several articles have shown that the Furlow double-opposing Z-plasty is an effective form of treatment for patients with velopharyngeal insufficiency.[1-4] However, the data indicate that the use of the Furlow Z-plasty to treat this condition is appropriate for select candidates only. D'Antonio et al.[1] suggested there may be important anatomic features that can be evaluated before surgery to predict which patients might be most likely to benefit from Furlow Z-plasty as a form of treatment. Over the past 10 years we have used the Furlow Z-plasty for treating velopharyngeal insufficiency in patients with minimal velopharyngeal gaps, good palate elevation, and muscle diastases indicative of submucous cleft palate or residual malpositioning of the levators.

Recent analysis of long-term follow-up, however, has identified a subset of patients who have consistently demonstrated a failure to achieve complete velopharyngeal closure after Furlow Z-plasty despite positive preoperative predictors. Specifically, we have noted that all of our patients with velocardiofacial syndrome have failed to achieve complete elimination of velopharyngeal symptoms following Furlow Z-plasty and have required further secondary palatal management.

Velocardiofacial syndrome is recognized as the most common syndrome associated with cleft palate and includes more than 100 clinical features.[5,6] The syndrome has been linked to a chromosomal microdeletion at 22q11.2 and can be iden-

tified by fluorescent in situ hybridization.[7] Some of the most common anomalies of importance in plastic surgery include cleft palate, deep pharynx, abnormal cranial base angle, conotruncal heart defects, displacement of the internal carotid artery, and syndrome-specific facies.[8]

It is a common observation among specialists in cleft lip and palate that traditional forms of secondary palatal management such as pharyngeal flap or sphincter pharyngoplasty are generally less successful in patients with velocardiofacial syndrome than in those without.[9,10] Therefore, we returned to our data concerning the outcome of the Furlow Z-plasty procedures performed on our population of patients with velopharyngeal insufficiency to assess whether there was a difference in surgical outcome between nonsyndromic patients with velopharyngeal insufficiency and patients with velocardiofacial syndrome.

To analyze surgical outcome, patients were divided into three groups: patients without any diagnosed syndrome or syndromic features (nonsyndromic), patients with velocardiofacial syndrome with a deletion at 22q11.2 confirmed by fluorescent in situ hybridization (VCFS), and patients who displayed many syndromic features including some phenotypic overlap with velocardiofacial syndrome but without a definitive diagnosis of that or another identifiable syndrome (syndromic). All patients were identified as having velopharyngeal insufficiency and required surgical correction based on a multi-method evaluation of velopharyngeal function, which included perceptual, aerodynamic, endoscopic, and cephalometric studies. After surgery, a full evaluation was repeated, and patients were categorized as having complete or incomplete velopharyngeal closure based on the results of the multi-method evaluation. Complete velopharyngeal closure was defined as the ability to close the velopharynx for correctly articulated sounds as determined endoscopically and aerodynamically and with no recommendation for further physical management.

Results of this analysis are shown in Figure 1. Seventy-eight percent ($n = 18$) of the nonsyndromic group and 38 percent ($n = 3$) of the syndromic group achieved complete velopharyngeal closure after Furlow Z-plasty. However, for the VCFS group, none of the four patients evaluated achieved complete closure after surgery. That is, all four of the patients with VCFS failed to achieve complete closure and required further secondary palatal management. Chi-square analysis indicates

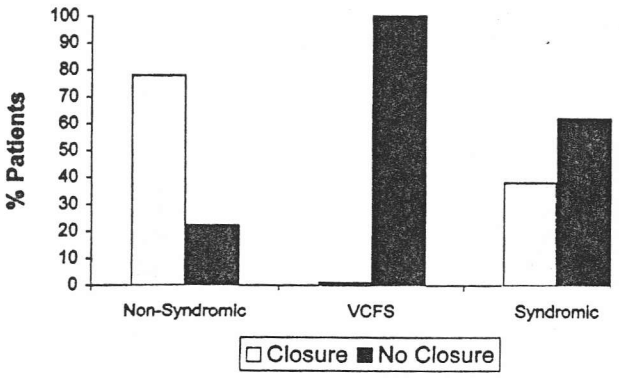

FIG. 1. Plot of percent of patients in each group (nonsyndromic, VCFS, and syndromic) that achieved complete velopharyngeal closure (closure) or incomplete velopharyngeal closure (no closure) after Furlow Z-plasty.

that there is a statistically significant difference among the groups for surgical outcome ($p = 0.004$). Although the difference in surgical outcome between the nonsyndromic and syndromic groups was not statistically significant ($p = 0.09$), the difference between the nonsyndromic and VCFS groups was statistically significant ($p = 0.01$). These data seem to indicate a gradation in surgical outcome for the three different groups. That is, Furlow Z-plasty seems to produce excellent results in the nonsyndromic group, varied results in the syndromic group, and poor results in the VCFS group.

These data support previous reports[9,10] suggesting a poorer outcome for treatment of velopharyngeal insufficiency in patients with velocardiofacial syndrome. These data also suggest that patients with velocardiofacial syndrome might be less likely to benefit from Furlow Z-plasty as a form of treatment for velopharyngeal insufficiency than their nonsyndromic counterparts. Two important conclusions can be drawn from these data. First, even when nonsyndromic children, syndromic children, and children with VCFS show similar patterns of velopharyngeal function evidenced by perceptual, aerodynamic, endoscopic, and cephalometric studies preoperatively, they may experience different levels of success following surgery. Post hoc assessment of the preoperative studies failed to demonstrate any features that could identify which patients might be expected to have negative outcomes. Therefore, the present data and the two previous studies[9,10] regarding surgical outcome in children with velocardiofacial syndrome show similar differences in outcome and strongly suggest that the diagnosis of this condition should alert the surgeon to be cautious when choosing the surgical procedure for treating velopharyngeal insufficiency in children with this syndrome. Despite the fact that the Furlow Z-plasty produces excellent results for nonsyndromic patients with velopharyngeal insufficiency, this procedure might have poor results for children with velocardiofacial syndrome and varied results in other children suspected of having a multi-malformation syndrome.

Second, the results pertaining to surgical outcome in children with velocardiofacial syndrome provide further data regarding the profile of speech impairment associated with this condition. Specifically, the data showing a consistent failure to eliminate velopharyngeal insufficiency in this population point to an underlying pathophysiology that differs from nonsyndromic or non-VCFS syndromic comparison groups. These data provide further support for the hypothesis given by Scherer et al.[11] and D'Antonio et al.,[12] which postulates that there may be quantitative and qualitative differences in the speech production mechanisms of children with velocardiofacial syndrome. For example, with respect to velopharyngeal function, D'Antonio et al.[12] reported that two of nine subjects in their VCFS group who had endoscopic evaluations showed no velar activity for speech, and four of nine demonstrated no observable lateral wall motion. However, none of the children in their non-VCFS comparison group demonstrated a complete absence of velar motion. These findings, combined with the present outcome data, support the hypothesis that there may be unique differences in the speech production mechanisms of some children with velocardiofacial syndrome compared with children with similar speech patterns who do not have a diagnosis of this syndrome.[11,12] These crucial quantitative and qualitative differences in the speech production mechanisms of children with velocardiofacial syndrome will require further investigation.

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