Impact of 22q Deletion Syndrome on Speech Outcomes Following Primary Surgery for Submucous Cleft Palate

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Background: Patients with 22q deletion syndrome are at increased risk of submucous cleft palate and velopharyngeal insufficiency (1-4). The authors' aim is to evaluate speech outcomes following primary Furlow palatoplasty or pharyngeal flap for correction of velopharyngeal insufficiency in submucous cleft palate patients with and without 22q deletion syndrome.

Methods: Records of submucous cleft palate patients who underwent primary surgery between 2001 and 2010 were reviewed. Data included 22q deletion syndrome diagnosis, age at surgery, procedure, preoperative nasopharyngoscopy and nasometry, speech outcomes, complications and secondary surgery rates. The primary study outcome was a perceptual assessment of resonance within normal limits. Secondary outcomes included the proportions of 22q deletion syndrome and nonsyndromic patients who (1) improved postoperatively with regards to perceptual assessment; (2) experienced a complication following surgery; or (3) required revision surgery for persistent velopharyngeal insufficiency.

Results: Seventy-eight submucous cleft palate patients were identified. Twenty-three patients had 22q deletion syndrome. Fewer 22q deletion syndrome patients obtained normal resonance on perceptual assessment compared to nonsyndromic patients (74 percent versus 88 percent). A similar difference existed based on postoperative nasometric scores. Among 22q deletion syndrome patients, similar success rates were achieved with Furlow palatoplasty and pharyngeal flap. No difference in the proportion improved postoperatively was noted between 22q deletion syndrome and nonsyndromic groups. One complication was experienced per group. More revision surgeries were indicated in the 22q deletion syndrome group (17 percent) compared to nonsyndromic group (4 percent). Median times to normal resonance for 22q deletion syndrome and nonsyndromic patients were 150 weeks and 34 weeks, respectively (Figure 1). Adjusting for multiple variables, 22q deletion syndrome patients were 3.6 times less likely to develop normal resonance.

Conclusions: Careful selection of Furlow palatoplasty or pharyngeal flap for primary repair of submucous cleft palate is highly effective in 22q deletion syndrome patients and yields results approaching those of nonsyndromic patients.

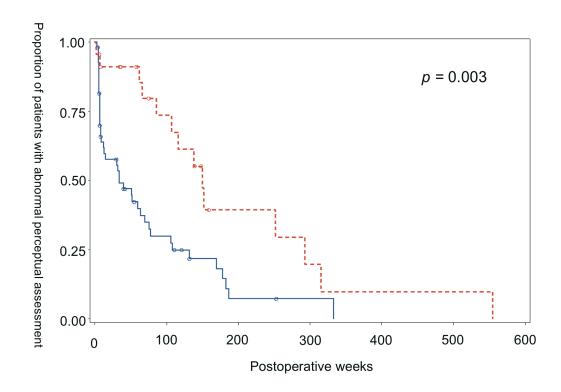


Figure 1

Kaplan-Meier analysis comparing unadjusted probabilities of developing normal resonance on perceptual speech assessment for 22q deletion (*red*) and nonsyndromic (*blue*) patients. Censored data are depicted by open circles. Median times to normal resonance for 22q deletion syndrome and nonsyndromic patients were 150 weeks and 34 weeks, respectively.

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Financial Disclosure

The authors have no commercial associations or financial disclosures to declare.