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Surgical, Speech, and Audiologic Outcomes in Patients with Orofacial Cleft and Van der Woude Syndrome

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Abstract

Objective—The purpose of this study was to investigate the surgical, speech, and audiologic outcomes in patients with Van der Woude syndrome (VWS) and compare them to patients with non-syndromic cleft palate with or without cleft lip (NS-CP±L) treated at the same institution.

Design—Retrospective chart review.

Setting—A single children's hospital at a major academic institution.

Patients—We retrospectively examined the records of 18 patients with VWS who had been treated at a single institution from 1989–2017. 38 patients with NS-CP±L who were also treated at the same institution during that same time frame were selected to closely match sex and date of birth.

Main Outcome Measures—Demographic, clinical, surgical, and speech pathology data were gathered from medical charts.

Result—By age four, 83% of subjects with VWS and 76% of subjects with NS-CP±L ($p = 0.732$) had been, or were actively involved in, speech therapy. By age ten, 100% of remaining subjects with VWS and 58% of remaining subjects with NS-CP±L remained involved in speech therapy ($p = 0.027$).

33% of patients with VWS and 16% of patients with NS-CP±L had a secondary procedure for velopharyngeal dysfunction (VPD) ($p = 0.171$).

Conclusions—The VWS group had more than twice the rate of secondary procedures for VPD repair, and a higher rate of continuing involvement for speech therapy at age ten. No differences were found in the rate of participation in speech therapy at or by age four.

Keywords

Van der Woude; cleft palate; cleft lip; speech pathology

Introduction

Orofacial cleft (OFC) is a frequently occurring developmental deformity, with reported incidences at 1–2/1000^{1–4}. It has been well established that patients with OFC require longitudinal multidisciplinary surgical, dental, speech, audiologic, and psychosocial care^{5–11}. OFC can be classified as syndromic, having additional abnormal clinical characteristics, or non-syndromic, having only isolated OFC¹². A well-known monogenic syndromic form of cleft palate is Van der Woude Syndrome (VWS; OMIM 119300), with most studies reporting an incidence of approximately 2% of all OFC patients^{3,13–17}. VWS is inherited in an autosomal dominant fashion^{14,16,18} and is noted to have high penetrance at 80% to nearly 100%, with high variability in phenotype^{13,19–21}. Patients with VWS are clinically characterized by having paramedian lip pits in addition to cleft lip (CL), cleft palate (CP), or cleft lip and cleft palate (CLP)^{16,19,20,22–24}.

While there have been prior studies investigating paramedian lip pits, dental features, facial features, family histories of VWS, and even wound healing in patients with VWS, the long-term outcome of these patients is not as well reported^{13,15,17,20,22,24–29}. Recently, Reardon et al.³⁰ documented a unique study involving the clinical characteristics and surgical course of 28 patients with VWS/Popliteal Pterygium Syndrome (PPS) at one tertiary care center. While the study by Reardon et al. (2015) is notable, it does not trace patient speech pathology outside of the need for secondary velopharyngeal repair. We therefore aim to investigate patients with VWS at our institution with an increased focus on speech pathology, specifically resonance, intelligibility, and need for speech therapy. The main purpose of this study is to investigate the longitudinal surgical, speech, and audiologic outcomes in patients with VWS and compare them to patients with non-syndromic cleft palate with or without cleft lip (NS-CP±L) treated at the same institution.

Materials and Methods

This study is a retrospective, descriptive analysis of the clinical characteristics, surgical procedures, and speech outcomes of children diagnosed with VWS with cleft palate ± CL. All patients were treated at one tertiary care center from 1989–2017. After obtaining Institutional Review Board approval, the electronic or paper medical records of all patients with VWS registered in the institution's database were reviewed. Candidates under the age of four at the time of the study, and those with medical records that lacked relevant data were excluded. One patient with VWS in the database with records from an outside hospital was excluded for not having received care from the institution of interest.

Demographic and descriptive measures gathered included: age, sex, length of follow up time, family history of OFC, presence of lip pits, and Veau class³¹. Occurrence of various surgical procedures including palatal repair, palatal fistula repair, and secondary surgery for velopharyngeal dysfunction (VPD) were recorded. The medical records showed conflicting

physician assessments on the occurrence and severity of palatal fistulae on several occasions. As such, surgical palatal fistula repairs were recorded and analyzed to track only palatal fistula that were symptomatic. The patient's age at the time of procedure and the repair technique were also recorded when available. In subjects with secondary surgical VPD repair, pre- and post-operative velopharyngeal closure ratios were gathered from videonasopharyngoscopy reports. Clinic evaluation reports nearest in date to the patient's 4th and 10th birthday were reviewed for each patient. These reports were scored on five domains (voice, intelligibility, resonance, language, and fluency) with a score of 0 assigned for absence of disorder/no intervention required, and a score of 1 assigned for disorder requiring intervention. These reports were scored by a single auditor (S.S.K.) with instruction and oversight by a speech pathologist (L.M.G.). Audiograms closest to the 3rd birthday and 10th birthday were analyzed and a pure tone average (PTA) of four frequencies (500Hz, 1000Hz, 2000Hz, and 4000Hz) in the better hearing ear was calculated for both ages.

The same data were also gathered for patients with NS-CP±L treated at the same institution. Patients with NS-CP±L registered in the database were first selected to match the sex of the subjects with VWS. All NS-CP±L patients were chosen by birth date to best match each subject with VWS. NS-CP±L exclusion criteria included physical anomalies besides OFC as well as the loss of follow up prior to age 8, unless the reason for loss of follow up was that the patient had explicitly graduated from cleft team care. Two subjects with NS-CP±L were included for each subject with VWS to increase the statistical power of comparisons between the two groups. We excluded any patients with cleft lip only or submucosal cleft palate, in order to focus more intently on patients with cleft palate with or without cleft lip. This resulted in the exclusion of 2 patients with VWS and 2 patients with NS-CP±L.

Descriptive statistics were performed. Comparisons between patients with VWS and patients with NS-CP±L were performed with Fisher's Exact test for categorical data and Student's t-tests for continuous variables. P values less than 0.05 were considered statistically significant and confidence intervals (CI) for differences in the sample means or proportions between the groups cover the 95% range. Fisher's Exact tests for differences in multinomial proportions were calculated with a programming script³² running in MatLab R2017a (MathWorks, Natick, MA). Power analysis was performed using PS Power and Sample Size Calculations version 3.0³³. All other statistical analysis was performed with IBM SPSS Version 23 (IBM Corp. Armonk, N.Y.).

Results

Demographics

Of 28 candidates with VWS in the database, 18 met the inclusion criteria. 38 patients with NS-CP±L meeting the inclusion criteria were also included. The groups have no statistical difference in terms of current age, proportions of male and female patients, and years of relevant medical records available. All patients with VWS had paramedian lip pits, and none of the patients with NS-CP±L had lip pits (Table 1).

Overall, the composition of Veau class proportions were different between groups. Post-hoc testing showed there were significant differences in the individual proportions of Veau class

III. Differences were not significant for Veau classes I, II, or IV after correcting for multiple comparisons (Figure 1a).

Between groups, there was no statistical difference in the number of patients with cleft palate, the mean age of primary palatal repair, the rate of palatal fistula repair, the rate of VPD repair, or the mean age at VPD repair (Table 2-SDC). Palatal fistula repair was done 12 cases. Of those repairs, 3 (VWS: 1; NSCLP: 2) were prior to the age four speech evaluation. Two of the 12 palatal fistula repairs occurred at the same time as alveolar bone graft. The palatoplasty techniques used in each group are illustrated in Figure 1b. Additionally, the VPD repair types are illustrated in Figure 1c.

Speech Pathology

Speech pathology assessments were gathered at a mean age of 4.9 ± 1.4 years for 17 patients with VWS, after excluding a single outlier. The mean age of speech pathology notes for all 38 patients with NS-CP±L was 4.5 ± 0.9 years ($p = 0.349$, CI [-0.42, 1.13]). At age four in the VWS group, 16 (83%) total patients were identified to have disorders requiring intervention in the domains of voice (1), resonance (3), intelligibility (14), and language (1). 29 (76%) total patients with NS-CP±L of the same age were identified to have disorders requiring intervention in the domains of resonance (5), intelligibility (17), language (5), and fluency (1) (Figure 1d). In both groups, multiple patients required intervention for disorders in multiple domains.

Intelligibility ($p = 0.021$, CI [8%, 58%]) was the only statistically significant subdomain difference between groups. Scores on the domains of voice ($p = 0.321$, CI [-5%, 16%]), resonance ($p = 0.703$, CI [-17%, 24%]), language ($p = 0.652$, CI [-23%, 7%]), and fluency ($p = 1.00$, CI [-8%, 2%]) were all similar between groups. At age four, a similar proportion of subjects with VWS were referred to speech therapy as subjects with NS-CP±L (Figure 1d).

A second set of speech notes was gathered at a mean age of 9.4 ± 0.8 years (VWS, $n=9$) and 9.6 ± 1.0 years (NS-CP±L, $n = 29$) ($p = 0.476$, CI [-1.05, 0.50]). By age 10, need for intervention stemmed from concerns for resonance (VWS: 0; NS-CP±L: 3) ($p = 1.00$, CI [-3%, 62%]) or intelligibility (VWS: 7; NS-CP±L: 14) ($p = 0.148$, CI [-0.03%, 62%]). At this second set of speech notes, speech therapy was recommended for 100% of patients with VWS and 58% of patients with NS-CP±L ($p = 0.027$) (Figure 1d).

In the small subgroup of patients who needed secondary surgical VPD repair, the mean pre- (VWS: 0.6, NS 0.7, $p = 0.671$, CI [-39%, 26%]) and postoperative (VWS: 1.0, NS-CP±L 1.0, $p = 0.735$ CI [-5%, 6%]) velopharyngeal closure ratios were similar in both groups.

Audiology

There was no difference in the mean age at which the audiologic data were gathered, PTA between groups at ages four or ten, or the proportion of patients in each group with abnormal hearing (PTA ≥ 20 dB is considered abnormal) (Table 3).

Discussion

The subjects in this study were evaluated under a number of different speech subdomains. At age four, some patients required intervention based on deficits in voice, language, and fluency subdomains. The majority of the interventions needed came from issues with resonance and intelligibility. Given that VPD has been shown to negatively impact resonance as well as articulation, and is more prevalent among patients with VWS, it was thought that the VWS group would have poorer resonance and intelligibility scores^{30,34–36}. Indeed, the rate of secondary surgical VPD repair in the VWS group (33%) was more than double the rate of the NS-CP±L group (16%). However, the proportions of patients recommended for resonance therapy in the two groups were not statistically different.

Similarly, there was no difference in the number of patients who had already received or were currently receiving speech therapy at age four. In contrast to resonance and overall enrollment in speech therapy, the data at four years of age show the VWS group had higher referrals to speech therapy for intelligibility than the NS-CP±L group. While the age of palatal repair and conductive hearing loss are relevant to speech development, neither of these factors were different between groups^{10,37–43}. A literature review did not find any other reports on the need for speech therapy in this population. While we did not document any other differences relevant to speech outcomes, speech and language development are complex and multifaceted, and it is likely that factors other than those recorded played a role.

Standard practice at our institution is to perform endoscopy as well as SVF studies pre- and post-operatively. However, of the 12 children to receive VPD repair, 8 had pre and post SVF. Several children had pre-operative SVF studies done, but no post-operative SVF studies as they were found to have clinically acceptable resonance by the speech pathologist. While this study is underpowered for full analysis, the VNE data do suggest that the VWS group did not have velar closure ratios consistent with more severe velopharyngeal dysfunction.

Speech pathology reports gathered for subjects at age 10 show that a higher percentage of patients with VWS were still receiving speech therapy compared to patients with NS-CLP. This may be because of a trend towards increased VPD in this group. It may also reflect the different distribution of Veau class between groups in this study. Additionally, the distribution of subdomains responsible for speech therapy recommendations shifted. For example, by age 10 there were no recommendations for speech therapy in either group for voice, language, or fluency deficiencies. All intervention recommendations in this age group stemmed from concerns with resonance and intelligibility.

In fact, the intelligibility scores were markedly inferior to resonance scores at 10 years of age for the population of both cohorts, continuing the trend noticed in the speech evaluations done at 4 years of age. A literature review shows there are not specific data on speech outcomes of a VWS group at age 10. In contrast, there are data on speech outcomes in pre-pubescent and adolescent patients with OFC, with one study reporting nearly half of the parents claimed an articulation disorder was present in their child with OFC at a mean age of 11, while a separate report stated that only 55% of adolescent and teenage patients with OFC

had acceptable speech^{44,45}. It is difficult to directly compare previous studies to ours, given differences in age and methods related to investigating quality of speech. Although there was a significant difference in involvement in speech therapy in this age group for patients with VWS, our data imply a persistent need for speech therapy in both groups.

Our study agrees with previously published work that patients with VWS have different frequencies of Veau class compared to patients with NS-CP±L³⁰. While there were instances of all four Veau classes in both groups, the plurality of patients with VWS had Veau class IV while the most common Veau class for patients with NS-CP±L was Veau Class III. Further, in concordance with previous studies on the phenotypes and penetrance of VWS, all patients with VWS had bilateral lip pits, and the VWS group had a stronger positive family history of OFC than NS-CP±L patients^{13,20–22,24,30}.

In contrast to previously published data, our cases and controls had the exact same rate of palatal fistula repair. Jones et al. (2010) found that patients with VWS had a higher rate of wound complications after cleft repair than patients with NS-CP±L. Furthermore, Reardon et al (2015) reported that patients with VWS/PPS had higher rates of palatal fistula incidence than patients with NS-CP±L. The discrepancy between this study and the literature may result from different palatal repair techniques, different surgeons across institutions, or cleft severity of individual subjects. Without a large scale, multi-site study these effects are difficult to dissect out.

This study has several limitations, most notably small sample size. This may account for the inability of this study to detect hypothetical differences in the proportions of each group requiring secondary surgical VPD repair and need for speech therapy at age 10. Given the proportions obtained in the two groups who received secondary surgical VPD repair, we would need a sample size of roughly 60 patients with VWS and 120 patients with NS-CP±L subjects to have 80% power to detect differences between the groups. In order to detect differences in the proportions of each group requiring speech therapy at age 10, a study would require 37 VWS and 75 NS-CP±L subjects in order to have 80% power to detect differences between the groups. Additionally, missing data decreased the power of statistical analysis.

Variability of recording may pollute the analysis: speech reports were written by two different speech pathologists; some VWS patients received initial repairs at outside hospitals; and several surgeons executed the initial and secondary repairs of the patients. This type of variability is common, and while its impact is uncertain, it is not likely to have had a significant influence on the results.

In conclusion, patients with VWS showed a significantly higher rate of continuing involvement in speech therapy at age ten and they also had more than twice the proportion of patients that needed VPD repair. There was no difference in prior or current involvement in speech therapy at age ten. Future investigations into these groups using magnetic resonance imaging may assist in better understanding the role of potential anatomic differences to clinical outcomes between patients with VWS and patients with NS-CP±L. Presently, this study may help elucidate the clinical and surgical outcomes relevant to speech in this patient

population. In turn, cleft care teams can plan care for the VWS population appropriately, as well as better communicate anticipated risks, costs, and needs to families of children with VWS.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

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Figure 1a.

Veau class frequencies in each group. Overall, difference in Veau class frequencies were different between groups ($p = 0.02$). * $p = 0.87$, CI $[-22\%, 24\%]$; † $p = 0.24$, CI $[-7\%, 30\%]$; ‡ $p = 0.01$, CI $[-62\%, -15\%]$; ¥ $p = 0.04$, CI $[0\%, 52\%]$.

Figure 1b. Palatoplasty repair techniques in each group. 6 patients in the VWS group were excluded because their records were incomplete. Abbreviation: IVV, intravelar veloplasty

Figure 1c. VPD repair techniques in each group

Figure 1d. The proportion of patients in each group who were referred to speech therapy at ages (VWS $n = 17$; NS-CL/P $n = 38$) and 10 (VWS $n = 9$; NS-CL/P $n = 29$).

* $p = 0.0732$, CI $[-15\%, 29\%]$; † $p = 0.027$, CI $[0.23\%, 59\%]$.

Table 1:

Descriptive Statistics of the Group

	VDS	NS-CP±L	<i>P</i>	95% CI*
n	18	38		
Current age (years)	16.00 ± 9.1	17.1 ± 9.2	0.667	(-6.39, 4.12)
Male	10 (55%)	21(55%)	1.00	(-26%, 26%)
Follow-up duration (years)	9.4 ± 5.6	10.6 ± 3.8	0.321	(-3.80, 1.34)

* 95% CI for the difference (VWS – NS-CP±L) between the sample means or the difference between the sample proportions, as appropriate.

Table 3:

Audiologic Data

	VWS	NSCLP	P value	CI
Age	3.4 ± 1.4 (n= 17)	3.9 ± 1.4 (n=38)	0.235	-1.26, 0.32
First PTA	13.2 ± 4.3	16.0 ± 9.9	0.286	-7.97, 2.4
Patients with abnormal hearing *	1 (6%)	8 (21%)	0.247	-32%, 3%
Age	9.5 ± 0.8 (n=8)	9.8 ± 1.1 (n=25)	0.505	-1.2, 0.6
Second PTA	8.6 ± 3.3	8.6 ± 4.3	0.909	-3.4, 3.4

* (Patients with abnormal hearing was decided to be a PTA >20, per audiology.)

Abbreviation: PTA, pure tone average