The Leuven staged supraperiosteal retropositioning repair: long-term velopharyngeal function in non-syndromic cleft palate


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Abstract. The Leuven staged supraperiosteal retropositioning repair: long-term velopharyngeal function in non-syndromic cleft palate. Background: From 1989 to 1993, 113 previously untreated patients were admitted to the Multidisciplinary Cleft Lip and Palate Team of the University Hospitals Leuven. Palate repair was performed in our centre by one surgeon (FO) in 88 patients. Our current surgical technique consists of a single-stage supraperiosteal retropositioning (modified Veau-Wardill-Kilner) for patients with a soft cleft palate only (SCP) or a soft cleft palate with up to 1 cm of the hard palate (HSCPpa). Patients with a larger or complete cleft of the secondary hard palate (HSCP) and patients with unilateral (UCLP) or bilateral (BCLP) cleft lip and palate undergo two surgical stages for palate closure: a supraperiosteal retropositioning is performed around 12 months of age, and a modified Langenbeck closure of the hard palate around 60 months of age.

Aim: To assess velopharyngeal function with speech as outcome measure.

Patients and methods: Velopharyngeal function was assessed in two ways. In one assessment, a “hard” outcome measure was the number of patients undergoing pharyngoplasty following palate repair in our centre (n = 88). In the other assessment, velopharyngeal function was evaluated in a homogeneous sub-population of 44 non-syndromic cleft patients with normal to slight impairment of the following functions: mental development, language development, and hearing. In this group, prospectively collected data about hypernasality and nasal emission were analysed retrospectively using a semi-objective nasality index (NI). Articulation was evaluated using a subjective articulation index (AI) representing articulation errors (retro-articulation, glottal stops and facial grimacing) associated with velopharyngeal insufficiency (VPI). Mean follow-up was 114 months.

Results: Despite rigid assessment by a phoniatrician and speech pathologist, only 1 patient out of 88 patients with soft palate surgery in our institution was thought to need pharyngoplasty. In the sub-cohort of 44 non-syndromic patients, nobody needed a pharyngoplasty. In the latter cohort, at the age of about eight years, 27 patients (61.5%) had undetectable nasality, 13 patients (29.5%) had an NI of 1 or “mild” nasality, and 4 patients (9%) had moderate nasality. At this point in time, articulation errors associated with VPI were noted in 14% of patients.

Conclusion: In this subgroup of cleft palate patients treated following the Leuven protocol, there was no need for secondary pharyngoplasty. Ninety-one per cent of patients had no, or only mild, rhinolalia aperta by the age of eight years, and 84% did not display VPI-related articulation disorders. This suggests that velopharyngeal function in patients treated by this protocol is excellent compared to results in the literature.

Introduction

The aim of the Leuven surgical approach is to achieve the three goals of palate surgery: to achieve velopharyngeal closure on demand, to separate the oral and nasal cavities and to inhibit cranio-facial maxillary growth as little as possible.1 There are two main differences between our technique and the more common techniques (Langenbeck, Veau-Wardill-Kilner push-back, Malek, Bardach two flap palatoplasty).2-4 In order to ensure minimal interference with cranio-facial maxillary outgrowth, we follow a two-stage protocol for palate closure.5 For the same reason,5,6 a supraperiosteal technique is used for soft palate closure at the age of 12 months, leaving the periosteum and the neurovascular bundle with the greater palatine artery intact on the palatal shelves. The hard palate closure is then carried out from 5 years of age onwards using a modified Langenbeck procedure. However, by this time, the remaining anterior hard palate opening decreases in size to only a virtual cleft in many patients.
In order to optimise velopharyngeal function, a maximal retropositioning of the levator muscle sling follows a transsection of the nasal mucosal lining at the dorsal border of the hard palate that proceeds as a back-cut on the lateral pharyngeal wall, inferiorly to the Eustachian tube, providing extra release. Critics hold that the raw surface that is created in this way on the nasal side of the soft palate will contract so much that it will completely undo any retropositioning. However, there is evidence in the wound healing literature that de-epithelialised tissue in a moist environment undergoing secondary healing will contract to about 40-50% of the original raw area, so that there is reason to believe that a substantial retropositioning can be maintained.\(^{10}\)

It is generally accepted that, following cleft palate repair, around 20 to 30% of patients require additional surgical treatment for velopharyngeal insufficiency (VPI).\(^{11}\) It was our clinical impression, even after rigid and systematic evaluation by a phoniatrician and a specialised speech and language therapist, that far less secondary pharyngeal surgery was needed in our patient population. Of about 15 pharyngoplasties performed annually in our department, few were performed in patients who were primarily operated using our current surgical protocol for primary palate repair, these interventions indeed mainly being carried out for secondarily referred cleft lip and palate patients or for non-cleft VPI patients (del22q11, velopharyngeal disproportion appearing following adenoidectomy). This retrospective study was designed to substantiate this clinical impression.

**Patients and methods**

**Patient group**

In the period 1989 to 1993, 113 new and previously untreated patients with cleft pathology were admitted to our cleft lip and palate multidisciplinary programme. Palate repair was performed in our institution in 88 patients. To create a homogeneous subset of patients for the assessment of velopharyngeal function following our current surgical protocol, the following patients were excluded.

Eleven patients were operated, mainly in the beginning of the study period, using a reversed surgical protocol (first hard palate closure, then soft palate closure) and were excluded. Fourteen patients with syndromes were excluded. These included Foetal Alcohol Syndrome (n = 1), Fragile X syndrome (n = 1), Wolf-Hirschhorn syndrome (n = 1), Binder Maxillonasal Dysplasia Malformation (n = 1), del22q11 syndrome (n = 2), Treacher-Collins syndrome (n = 1), Van der Woude syndrome (n = 1), Greig syndrome (n = 1), Opitz G/BBB syndrome (n = 1), Oculodentodigital Dysplasia Syndrome (n = 1), one undefined syndrome (Unilateral Cleft Lip and Palate (UCLP), congenital cataract, microphthalm, mental retardation, epilepsy (n = 1), microcephaly (n = 1), Cleft Palate Lateral Synaecia Syndrome (n = 1). Five patients with serious hearing loss (Fletcher index > 40 dB) at the age of 6 were excluded. Four other patients with serious psychomotor retardation (IQ < 75) were excluded. Finally, 10 patients with missing data for velopharyngeal function were also excluded. These included 3 patients with soft cleft palate (SCP, Veau type I), 5 patients with a soft cleft palate with partial cleft in the hard palate (HSCPpa), and 2 patients with UCLP. They had been operated in our institute, but had subsequently attended the Cleft Team outpatient clinics rarely or not at all.

Of the remaining 44 patients, 20 were patients with cleft palate only (SCP, Veau type I: n = 6, complete Hard/Soft Cleft Palate (HSCP), Veau type II: n = 1, HSCPpa, n = 13). The remaining 24 patients had cleft lip and palate (UCLP n = 17, BCLP n = 7).

**Treatment: surgical technique**

These 44 patients all underwent the current surgical protocol used by the Multidisciplinary Cleft Lip and Palate Team of the University Hospitals Leuven. The technique was developed by one of the authors (FO) and subsequently refined (VV) and combines elements that are also found in other established techniques. The goals are optimal velopharyngeal function by retropositioning and the re-creation of a correctly oriented levator muscle sling,\(^{4,13}\) and the minimisation of maxillary hypoplasia by the prevention of deperistiation\(^{6,9}\) while leaving the hard palate open until about the age of 5.\(^{5,9}\) Under 4x loupe magnification, the first step is closure of the nasal floor using vomerine flaps medially and nasal perosteal flaps laterally. A Wardill incision is then made, with the subsequent development of flaps, including only the mucous membranes and submucous layer with the minor salivary glands. The periostem and the neurovascular bundle remain on the hard palate, thus preventing medial traction on the
periodontal ligament. At the very midline a strip of about 1 to 2 mm of periosteum is retained in the flaps to maintain some strength in the midline where the suture will be placed. A single-stage surgical closure obviously suffices for patients with a soft cleft palate only (SCP) or a soft cleft palate with up to 1 cm of the hard palate (HSCPpa). In patients with a larger or complete cleft of the secondary hard palate (HSCP) and in patients with unilateral (UCLP) or bilateral (BCLP) cleft lip and palate, two-stage supraperiosteal retropositioning was performed.

**Treatment: speech therapy**

In addition to surgical therapy, speech therapy was also provided to attain the final speech result that is the object of this study. Speech therapy in Flanders is typically prescribed for renewable periods of 6 months, during which most patients receive therapy twice a week. By 8 years of age, 9 patients had never had any speech therapy (SCP: n = 5, HSCPpa: n = 2, UCLP: n = 2), 12 patients had received 2 courses of 6 months, 14 patients had received 4 courses of 6 months of speech therapy, 3 patients had received 6 courses of therapy, and the remaining 6 patients had received more than 6 courses. Figure 1 gives an overview of the amount of speech therapy needed to reach the results presented in this study.

**Results assessment**

**Number of pharyngoplasties performed**

The number of pharyngoplasties is subject to a specific threshold, which in turn depends on the rigidity and scrutiny of speech evaluation and the wishes of the patient and his/her environment. However, the number of pharyngoplasties performed in a department can be considered a “hard measure” in so far as evidence of scrutinised analysis and speech results of patients not undergoing secondary pharyngeal surgery are also available for comparison in the population under study.

**Speech evaluation**

The numerical scores resulting from standardised multifactorial speech analysis were reduced to a nasality index (NI) and a compensatory articulation index (AI).

The nasality index consists of a semi-quantitative summary rating of eight items that were systematically rated by the dedicated cleft team speech and language therapists and the otolaryngologist-phoniatrician coordinating the cleft team (WW). Items rated are displayed in Table 1 and include items testing hypernasality (non-nasalised target vowels laal, loel and slightly nasalised lil,) and nasal air emission (patient asked to blow, repeat the words “kapot”, “koude koffie”, “dikke kroket”; and the sustained ls: fricatives). A resulting score from 0-3 is attributed to every item using a semi-objective interpretation of the magnitude of the moist area on the “Czermak cold mirror test”. The scores are totalled for every item, resulting in a score ranging from 0-24, which is subsequently reduced to 0 (completely normal velopharyngeal function), 1-8 (mild VP inadequacy), 9-16 (moderate VPi), and 17-24 (serious VPi). In this way, there is a four-tiered rating system that compares to results of analysis using validated speech analysis systems such as the CAPS (Cleft Audit Protocol for Speech) or the PWSS (University of Pittsburgh Weighted Values for Speech Symptoms associated with VPi).

The Articulation Index was computed following the assessment of the items “facial grimacing” (present = 1; or absent = 0), “retroarticulation” (palatal fricative, mid-dorsal fricative; one of these or a combination present = 1; or absent = 0) and “compensatory articulation” (pharyngeal fricative or glottal stops/fricatives present = 1; or absent = 0) using an extensive, phonetically balanced, word list. For every finding present, one point was attributed, thus resulting in a score ranging from 0 (no VPi-related articulation errors) to 3 (all 3 items present). Minor non-VPi-specific articulation errors (lateralisation, inter- and addentality) were not considered in this assessment.
For all patients, NI and AI were attributed at three points during the follow-up period: at 4 years of age, before closure of the hard palate in our protocol, in patients with a staged hard palate closure (i.e. UCLP, BCLP, and SHCP complete), at about 6 years of age, when pharyngoplasty would normally have been considered in patients with VPI not responding to speech therapy, and at about 8 years of age, when the condition of the patients is expected to be stabilised.

On the basis of differences in the surgical technique used, comparative analysis was conducted, looking separately at patients with SCP, HSCP and three patients with this disorder in association with a cleft lip but with an intact hard palate on one hand, versus UCLP and BCLP patients on the other.

Results

Number of pharyngoplasties performed

In the total group of 88 patients undergoing palate repair, one patient later underwent pharyngoplasty. This patient had Treacher-Collins Syndrome and an isolated soft cleft palate. The primary palate operation was complicated by dehiscence of the muscular sling, scarred healing, and resulting VPI. The pharyngoplasty was performed only seven years later due to irregular attendance at the cleft team clinic.

Speech evaluation

NI as a function of age at follow-up can be seen in Table 2. Figure 2 shows the NI findings and evolution over time as a histogram in 44 patients.

At 4 years of age, 13 patients – 29.5% – had no hypernasality and nasal emission in the test situation, 25 patients or 56.8% had a “mild” nasality index (NI = 1) and 6 patients or 13.6% had a “moderate” result (NI = 2) on NI testing. At 6 years, the rating for VPI had shifted towards lower levels of the NI, with 17 patients or 38.6% without specific VPI (NI = 0), 22 patients or 50% with mild VPI not interfering with intelligibility (NI = 1), and 4 patients or 9.1% remaining with moderate VPI (NI = 2). By 8 years of age, the number of patients with a NI of 0 increased to 27 patients (61.4%) at the expense of the patients with a NI of 1 (n = 13, 29.5%). It was observed that 91% of patients displayed no, or only mild, nasality by the age of 8 years. The two patients with a NI of 2 remained at this level.

The AIs as a function of age of follow-up are shown in Table 3.
A histogram of this set of data for 44 patients can be found in Figure 3. The number of patients with an AI of 0 increases by the age of 8 at the expense of the number of patients with mild and moderate articulation disorders (AD). At 4 years of age, 28 patients or 64% had no specific VPI-related AD, 9 patients or 20% had one typical VPI-related AD (retroarticulation: n = 6, glottal stops n = 3), 6 patients or 14% had a combination of two AD (4 retroarticulation and glottal stops, 2 glottal stops and facial grimacing) and 1 patient or 2% had all three typical VPI-related ADs.

At six years of age, the severity of disorders had shifted towards the lower levels of the AI, with 29 patients or 66% without specific VPI-related AD, 13 patients or 30% with 1 typical VPI-related AD (retroarticulation: n = 7, glottal stops n = 5, facial grimacing n = 1), 1 patient or 2% with a combination of two ADs (retroarticulation and glottal stops) and 1 patient or 2% with all three typical VPI-related ADs. By the age of eight, most VPI-related ADs had disappeared, 38 patients or 86% had no more ADs, with the 6 remaining patients (14%) still displaying retroarticulation (n = 4; 9%), glottal stops (n = 1; 2%) and facial grimacing (n = 1; 2%). This evolution over time can also be found in Figure 3. A comparison with some published and comparable results is made in Figure 4.

When comparing the velopharyngeal function in cleft-palate-only patients (CP; n = 20) to cleft lip and palate patients (CLP; n = 17, BCLP; n = 7), a clearly larger proportion (80%) of patients with CP achieved speech without nasality by the age of eight years, as compared to the CLP patients (45%) (Table 4). These proportions are comparable to the data of Park.16 The rate of moderate to severe nasality was, however, similar in both groups, with 9% displaying moderate nasality and none severe nasality by the age of 8 years. In terms of articulation, CP patients also did better. The six remaining patients with AD at 8 years of age were all CLP patients. These observations are clarified in Figure 5a and Figure 5b.

Discussion

As is generally the case in studies dealing with result evaluation in cleft palate surgery, it is very difficult to make meaningful comparisons with results published by other authors. Differences between surgical techniques are manifold. Even when comparing centres that use “the same” surgical technique, i.e. with the same...
definition, many surgical details will prove to be different when a comparison is actually made. Furthermore, result evaluation as conducted in the patient group we describe is hampered by the fact that this is a retrospective assessment of prospectively gathered data using a local speech evaluation system. In the meantime, internationally accepted and validated tools have become available (e.g., GOS.SP.ASS17). Furthermore, different languages allow for the acceptability of different levels of hypernasality. Nevertheless, in the following discussion, we try to compare our findings to studies that report on their results studying a comparable patient group with a similar evaluation system.

Rate of pharyngoplasty

We had one pharyngoplasty procedure in 88 patients operated for cleft palate and none in the 44 non-syndromic patients. This is a remarkably low rate for secondary pharyngeal surgery. The required number of pharyngoplasties reported in the literature describing speech results following different surgical and speech therapy protocols is generally higher (Table 5). As already stated, this number does not mean a lot in itself. It may mean that our threshold for performing secondary pharyngeal surgery is comparatively high. In order to interpret this finding, a review is required of the speech results (NI, AI) for the patients who did not undergo secondary pharyngeal surgery in our department.

Speech and language parameters

Turning to the NI (hypernasality and nasal emission) by the age of 8 years in our patient group, we found that 9% of patients scored level 2 on a 4-tiered assessment scale, meaning moderate nasality. No patients were classified as “severe”. A comparison with the results of Park et al. can be found in Table 4. Our results are also comparable to the rate reported by Pigott et al. in patients with UCLP: 9.1% of their patients displayed moderate to severe nasal emission/hypernasality. In Pigott’s series, these patients subsequently underwent secondary pharyngeal surgery. It can be argued that our
rate of pharyngoplasty would have attained 9% if we had used the same criterion for our patients with moderate nasality. Tables 6 and 7 compare our findings to the speech results of the Zürich group. This comparison makes sense, particularly because the surgical technique is quite similar (two-stage repair, supraperiosteal dissection). No secondary pharyngoplasty was needed in this group of UCLP patients either. The speech results of the Zürich patients, as evaluated by an independent American observer, are quite comparable to our data. In Table 6, it can be observed that, in both series, about 9 in 10 UCLP patients are considered to have no or mild nasality (with an almost equal distribution between the group displaying “no” versus “mild” nasality). Table 7 shows the NI data for our 44 patients in relation to other series in the literature using a comparable 4-tiered evaluation system. The good results in both the Zürich and the Leuven data might be explained by the common denominator in the surgical technique: the focus on palatal lengthening and radical muscle sling correction. Other authors stressing these points have also achieved very good speech results.[13]

Turning to nasality, one striking difference in our data is the trend towards a higher proportion of patients with no nasality in the SCP-HSCP-HSCPpa group (80%) when compared to the UCLP and BCLP patients (46%) (Figures 5a and 5b; Fisher exact test p = 0.088). The significance of this finding is unclear. Some authors report the same finding,[16,22] other authors found no difference between these cleft type groups,[12] and Timmons et al.[28] found the exact opposite. The fact that only one fully-fledged HSCP patient was included in our group of 20 patients might explain the better outcome, since there are indications in the literature that the complete secondary HSCPs do worse, requiring for example a higher frequency of secondary pharyngeal surgery when compared to SCP en HSCPpa.

Turning to articulation, the results observed in our population in terms of the rate of retroarticulation and the number of patients with glottal stops are also quite comparable to what is observed in other patient series. Table 6 compares the number with the results of authors reporting in a similar way.
Figure 4). There is a clear reduction in these problems during follow-up, which seems to be related to the closure of the anterior hard palate at 6 years and the subsequent speech therapy to reduce the remaining articulatory disorders. Park described a similar decrease over time. The rate of retroarticulation without glottal stops or facial grimacing at 8 years is 9%. The large Japanese series of Park et al.16, using similar selection criteria for the patient sample, found a rate of 11% for retroarticulation at 10 years of age. Glottal stops were observed in 4% in their series, compared to 2% in our patients.

Conclusion

Taking into account the stated reservations, velopharyngeal function in cleft patients treated with surgery and speech therapy according to the Leuven protocol would seem to be at the upper range of results reported in the literature, both in terms of velopharyngeal function (nasal escape, hypernasality) and compensatory articulation. A prospective study with a validated speech assessment tool in combination with objective measurements (nasometer, nasendoscopy) is required for the further substantiation of this impression.

References

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