Cleft Palate Outcomes and Prognostic Impact of Palatal Fistula on Subsequent Velopharyngeal Function – A Retrospective Cohort Study.

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Abstract

Objective: To assess outcomes from cleft palate repair and define the level of impact of palatal fistula on subsequent velopharyngeal function.

Design: A retrospective cohort study.

Setting: A regional specialist cleft lip and palate centre within UK.

Participants: Non-syndromic infants born between 2002 and 2009 undergoing cleft palate primary surgery by a single surgeon with audited outcomes at 5 years of age. Four hundred and ten infants underwent cleft palate surgery within this period and 271 infants met the inclusion criteria.

Interventions: Cleft palate repair including levator palati muscle repositioning with or without lateral palatal release.

Main Outcome Measures: Postoperative fistula development and velopharyngeal function at 5 years of age.

Results: Lateral palatal incisions were required in 57% (156/271) of all cases. The fistula rate was 10.3% (28/271). Adequate palatal function with no significant velopharyngeal insufficiency (VPI) was achieved in 79% of patients (213/271) after primary surgery only. Palatal fistula was significantly associated with subsequent VPI (risk ratio = 3.03, 95% confidence interval: [1.95, 4.69], p-value < 0.001). The rate of VPI increased from 18% to 54% when healing was complicated by fistula. BCLP repair complicated by fistula had the highest incidence of VPI (71%).

Conclusions: Cleft palate repair with levator muscle repositioning is an effective procedure with good outcomes. The prognostic impact of palatal fistula on subsequent velopharyngeal function is defined with a highly-significant three-fold increase in VPI. Early repair of palatal fistula should be considered, particularly for large fistula and in BCLP cases.

Introduction

Palatal fistula is an undesirable complication of cleft palate repair with an a reported incidence of between 2.4% and 24% (Sommerlad, 2003; Inman et al., 2005; Becker and Hansson, 2013; Hosseinabad et al., 2015; Tse and Siebold, 2018) and can result in nasal regurgitation of foods, speech disturbance or both. Some reports (Sommerlad, 2003; Inman et al., 2005) may only include fistulae which required surgical repair whereas others (Becker and Hansson, 2013) include all fistulae regardless of symptoms or need for repair. WhilstBecker and Hansson (2013) found no clear difference in fistula frequency between different cleft types, others including Sommerlad, (2003); Lu et al., (2010); and Hosseinabad et al., (2015) and Tse and Siebold, (2018) have reported higher fistula rates in more severe clefts; such as complete bilateral cleft lip and palate. This may be explained by the increased technical difficulty of repairing wider and more extensive clefts and increased wound tension of the oral aspect. Lateral palatal release incisions (Langenbeck flaps) are sometimes used to facilitate midline closure of cleft palate and reduce wound tension. Indeed Sommerlad (2003) attributed his 15% rate of fistula requiring surgical repair possibly to his attempts to avoid lateral releasing incisions whereas Becker and Hansson (2013) postulated that their lower fistula rate of 5% was a reflection of their more liberal use of lateral releasing incisions in wider clefts.

A partial dehiscence of the palate with subsequent fistula development probably increases the risk of velopharyngeal insufficiency either directly through the fistula or due to the additional scarring, contracture and possible muscle disruption. Velopharyngeal insufficiency rates following cleft palate repair are reported between 20-30% (Clinical Standards Advisory Group - Cleft Lip and/or Palate, 1998; Phua et al., 2008). When considering the potential impact of a palatal fistula on velopharyngeal function it is important to report all patients with VPI and not just those patients who had subsequent velopharyngeal speech surgery. The potential impact of palatal fistula on subsequent velopharyngeal function is unclear. Previous studies have highlighted other factors which impact on velopharyngeal functional outcome. Yang et al. (2013) reported that age (less than 2 years) and cleft type were relevant factors influencing subsequent velopharyngeal function - as in our study, higher rates of VPI were seen in BCLP and complete CPO patients. Other studies

have found that cleft width is associated with higher VPI rates (Lam et al., 2012; Mahoney et al., 2013; Leclerc et al., 2014; Yuan et al., 2016). Hardwicke et al. (2016) also found that Pierre-Robin sequence cleft palate patients had a higher need for secondary VPI surgery compared to matched cleft palate patients without Pierre-Robin sequence.

However no previous study has specifically demonstrated the association or level of risk between palatal fistula following primary surgery and subsequent VPI. Therefore this retrospective cohort study was carried out on infants born with cleft palate between 2002 and 2009 using a standardised surgical protocol and by a single surgeon to determine:

- The incidence of lateral palatal release incisions (Langenbeck flaps)
- The incidence of postoperative fistula (defined as any unintended persistent communication between the mouth and the nose within the surgical site).
- Velopharyngeal functional outcomes
- The prognostic level of impact of a fistula on subsequent velopharyngeal function.

Methods

Four-hundred and ten children born between 2002 and 2009 with a cleft palate with or without cleft lip underwent primary cleft palate repair by a single surgeon (AGS). Patients were excluded from the study if they had an associated syndrome, significant developmental delay, submucous cleft palate or incomplete records at 5 years of age. Infants with isolated cleft palate in association with Pierre-Robin sequence were included. Two-hundred and seventy-one children attended for clinical audit at 5 years of age and were included in this study. The surgical protocol followed for cleft palate only (CP) was a single-stage repair at 8-9 months of age. Repair of unilateral and bilateral cleft lip and palate (UCLP/BCLP) was carried out in two stages with initial lip and anterior palate repair with vomerine flap at 3-4 months; followed by posterior cleft palate repair at 8-9 months. The operating microscope was used routinely in all cleft palate repairs. Lateral palatal incisions to create Langenbeck flaps were made only when considered necessary to achieve adequate

oral closure. Intravelar veloplasty with levator muscle posterior repositioning and repair was performed in all cases as described by Sommerlad (2003). Prophylactic antibiotics were prescribed routinely and arm-splints provided postoperatively for four weeks. Data collection included type of cleft, sex, age at primary repair, need for lateral release incisions, postoperative fistula and palatal functional outcomes as determined by perceptual speech assessment. All palatal fistulae were recorded regardless of size, position, symptoms or need for surgical repair. Speech outcomes were audited at 5 years of age by specialist speech and language therapist consensus listening using the 'Cleft Audit Protocol for Speech

- Augmented (CAPS-A) assessment tool' (John A et al., 2006). Velopharyngeal function was considered adequate when green outcomes (scores 0 or 1) were achieved in all parameters of intelligibility, hypernasality, hyponasality, audible nasal emission, nasal turbulence and no history of secondary palatal or speech surgery.

Statistical Analysis

Data were summarised through frequency with proportion. The incidences of lateral palatal release and palatal fistula and subsequent palatal function at 5 years of age were presented in contingency tables with frequency and proportion, stratified by the type of cleft palate. Chi's-square test and Fisher's Exact test were applied to test the associations between palatal fistula and subsequent velopharyngeal function where appropriate. Risk ratio was reported with 95% confidence interval. All analyses were performed in R version 3.1.2 (https://cran.r-project.org/).

Results

Of the 271 patients with cleft palate, 157 (58%) were CP, 79 (29%) UCLP and 35 (13%) BCLP.

The male to female ratio for CP was 1:1.2 (70 males; 87 females), 2.3:1 for UCLP (55 males; 24 females) and 4.8:1 for BCLP (29 males; 6 females). Mean age at the time of cleft palate repair was 9.2 months (range 7-27 months). The most frequent age at surgery was 8 months (143 cases).

Lateral palatal release/Langenbeck flaps (Table 1)

Lateral palatal release incisions were made in 57% (155/271) of palate repairs and were mostly bilateral (unilateral in 5 patients). The requirement for lateral incisions varied between cleft types being 38% for CP, 78% for UCLP and 97% for BCLP. Further analysis of the CP group revealed that whilst lateral incisions were required in 86% of complete clefts, this reduced to 20% (23/115) for incomplete CP.

Postoperative palatal fistula (Table 1)

Palatal healing was complicated by fistula development in 28 of the 271 (10.3%) cases (28/271; 10.3%). The fistula rate varied between cleft types being highest for BCLP and complete CP (20% and 19% respectively), 10% for UCLP and 4.3% (5/115) for incomplete CP.

Thirteen of the 28 fistulae were symptomatic with a history of nasal regurgitation and/or associated velopharyngeal insufficiency and which required surgical repair. If symptomatic fistulae requiring surgery are considered alone, the fistula rate reduced to 4.8% (13/271). In common with the cleft sub-group overall fistula rates, the BCLP group also accounted for the higher highest proportion of fistulae requiring surgical repair (4/35; 11.4%) in comparison to CPO (6/157; 3.8%) and UCLP (3/79; 3.7%).

Functional Outcomes at Five Years of Age (Table 1)

Seventy-nine per cent (213/271) of patients had adequate palatal function at 5 years of age (green scores on CAPS-A) and no history of secondary palatal or speech surgery. The incidence of adequate palatal function varied between cleft types - CP and UCLP had the highest rates of adequate function (82% and 80% respectively) and BCLP the lowest at 63%.

In the CP sub-group, clefts of the soft palate only had the best functional outcome with 32/36 (92%) children achieving adequate palatal function for speech.

Association between postoperative fistula and subsequent velopharyngeal function (Table 2)

A highly statistically significant association was seen between postoperative fistula and subsequent velopharyngeal insufficiency (VPI). Overall, a three-fold increase in rate of VPI

(Risk ratio [RR]: 3.03, 95% CI: 1.95-4.69, p-value < 0.001) was observed when palate repair was complicated by fistula (VPI - 15/28; 54%) compared to a VPI rate of 18% (43/243) when healing was not complicated by fistula. For CP and UCLP specifically the incidence of VPI increased from 16% and 17% respectively to 46% and 50% when the palate repair was complicated by a fistula. The highest rate of VPI was seen in the BCLP group which increased from 28% without fistula to 71% when a palatal fistula was present (RR: 2.50, 95% CI: 1.18-5.29, p-value = 0.039).

Whilst a strong and statistically significant association was established between palatal fistula development and subsequent speech VPI, no statistical association was apparent between patient sex, cleft type, age at surgery and VPI.

An association was also seen for the full cohort of palate repairs between lateral palatal release incisions (oral flaps) and VPI (RR:1.92, 95% CI: 1.14-3.24, p-value = 0.011), however no statistically significant association was seen for the individual cleft sub-types.

Discussion

The aims of cleft palate repair are to achieve an intact palate with minimal scarring and adequate palatal function for normal speech. It is reasonable to avoid lateral palatal incisions when feasible during cleft palate repair to reduce scar tissue associated with dental cross-bite and maxillary retrusion. However lateral incisions may be required in wider or less favourable clefts to achieve closure. In this study of 271 cleft palate primary repairs, lateral palatal incisions were considered necessary in 57% of cases overall (CP 38%, UCLP 78%, BCLP 97%) which is higher than the 23% overall (CP 16%, UCLP 20%, BCLP 71%) reported by Sommerlad (2003) in his series of 285 cleft palate repairs which had a similar distribution of cleft types. However the use of lateral incisions should not be considered in isolation but preferably correlated with wound complications such as fistula rates and speech outcomes.

The lower incidence of palatal fistula in this series (10.3%; 28/271) and across all three cleft groups (CPO 8%, 13/157; UCLP 10%, 8/79; BCLP 20%, 7/35) compared to the fistula-repair rate of 15% reported by Sommerlad (2003) is possibly a reflection of the more liberal use of

lateral release incisions in our cohort. Furthermore, if only symptomatic fistulae requiring repair are considered then our fistula rate reduces to 4.8% (13/271). This would support the suggestion of a relationship between lateral incisions and palatal fistula however this relationship is complex, as the use of lateral incisions is usually restricted to wider or less favourable clefts.

In this cohort, 79% of patients had adequate palatal function at 5 years of age with no significant hypernasality or other feature of velopharyngeal insufficiency and no history of secondary palatal surgery or speech surgery. National standards for speech outcomes following cleft palate repair have been developed and introduced in the UK (Britton et al., 2014). These standards include a target of 70% of cleft palate patients achieving adequate palatal function at 5 years of age following cleft palate primary repair with no history of previous fistula repair, secondary palatal surgery or speech surgery (national speech standard 2A). Comparison of functional outcomes with other series such as Sommerlad (2003) and Inman et al. (2005) can be difficult as unfavourable outcomes are sometimes reported as the number of patients who underwent secondary velopharyngeal surgery; which is a less accurate measure of palatal functional outcome. Mild presentations of VPI may not be considered sufficiently clinically significant or further surgery may be declined by carers or contra-indicated on medical grounds. Although 58 patients (21%) had some evidence of VPI on perceptual speech assessment, secondary palatal surgery or pharyngoplasty was performed in 36 patients (13.3%). The need for secondary palatal or speech surgery was highest in BCLP group at 22%, followed by CP 13.4% and UCLP 8.9%.

A clear and statistically significant association was found between fistula development and subsequent inadequate palatal function (VPI). The presence of a palatal fistula after cleft palate primary surgery correlated with a highly-significant three-fold increase in subsequent rates of VPI (p-value < 0.001). When palatal healing was uncomplicated by fistula development the incidence of VPI was 18%, however this increased to 54% when palatal healing was complicated by a fistula. This order of increased risk was seen across all three cleft groups. In CP and UCLP the incidence of VPI increased from 17% (without fistula) to 50% when healing was complicated by fistula development. For BCLP, the proportionate risk

of VPI increased from 28% (without fistula) to 71% with fistula.

Although an association was seen between lateral palatal release incisions (oral flaps) and VPI in the full cohort of palate repairs, we are not suggesting that lateral palatal incisions are a direct risk factor for VPI. Lateral release incisions were associated with fistula development which is the risk factor for subsequent VPI. Lateral incisions were only made when necessary to complete palate closure, with the highest need seen in BCLP and complete CP. Therefore lateral release incisions/oral flaps were more often required in wider and complete palatal clefts which are more likely to be complicated by fistula development than narrower or incomplete clefts.

Our study confirms that the presence of a palatal fistula is a significant risk factor for subsequent VPI due to palatal incompetence. Of the 13 patients who subsequently underwent fistula repair, 7 were described as small (pin-hole or slit fistula) which were subsequently repaired as part of a full cleft palate re-repair at 3-4 years of age following confirmation of VPI on full perceptual speech evaluation and lateral videofluoroscopy. Five other patients (2 BCLP, 3 CPO) underwent early fistula repair (between 13 months and 23 months of age) due to significant fistula development/partial wound dehiscence following primary surgery. Whilst one (CPO) of these five patients responded well with no subsequent VPI, the remaining four patients presented with VPI despite an intact palate and required further speech surgery with good outcome (pharyngoplasty in two patients, palate re-repair and buccinator flap lengthening in two patients). A further patient with UCLP and significant fistula development following posterior cleft palate primary repair underwent delayed fistula repair and buccinator flap lengthening at 5 years of age due to co-existing congenital heart disease, again with a good outcome and velopharyngeal competence achieved.

Therefore in CP and UCLP palate repairs complicated by fistula where the risk of subsequent

VPI is approximately 50:50, it may be reasonable to adopt an initial conservative approach and consider secondary velopharyngeal surgery only on for those patients who demonstrate subsequent VPI on full perceptual speech assessment. However with regard to BCLP palate repairs complicated by fistula, early closure of the fistula should be considered due to the

higher risk of subsequent VPI. Although early closure of a palatal fistula in BCLP patients would not necessarily prevent the onset of subsequent VPI, nonetheless an intact palate would assist further speech surgical management.

This data enables professionals involved in cleft-care to better counsel parents and carers of children with cleft palate, according to cleft type; regarding surgical outcomes and quantifies the level of risk of subsequent velopharyngeal insufficiency in CPO, UCLP and BCLP when palatal primary surgery is complicated by fistula development.

Up to 2013, 13.3% (36/271) of patients in this series had undergone secondary surgery for VPI and the most frequent operation was a cleft palate re-repair. Twenty-two further patients (8%) had some evidence of VPI at 5 years of age however this was mild and often inconsistent in nature and further surgery was not performed.

Conclusions

In this retrospective cohort study of 271 cleft palate repairs 79% (213/271) of patients had normal palatal function at 5 years of age with no significant hypernasality or other feature of velopharyngeal insufficiency (green outcomes on CAPS-A) and no history of secondary palatal surgery or speech surgery. Cleft palate only and unilateral cleft lip and palate have favourable palatal outcomes with complete healing in 90% of palate repairs (fistula rate 10%) and adequate palatal function for normal speech in 80% (some evidence of VPI in 20% of patients). However in common with other series, patients with bilateral cleft lip and palate have less favourable outcomes after primary surgery with a higher fistula rate of 20% and some evidence of VPI in around one-third of all BCLP patients. The prognostic impact on palatal function of a complicating postoperative palatal fistula is confirmed and defined with a statistically significant three-fold increase in rate of VPI. The incidence of VPI following cleft palate repair in cleft palate only and unilateral cleft lip and palate increases from 15% without fistula; to 50% with fistula. For bilateral cleft lip and palate VPI rates are higher, increasing from nearly 30% without fistula; to 70% when palatal healing is complicated by fistula. As the risk of subsequent VPI after palate repair complicated by fistula in cleft palate only and unilateral cleft lip and palate patients is 50%, it is suggested that an initial

conservative approach is reasonable (particularly if the fistula is small < 5.0mm) and that the possible need for further surgery will depend upon future speech and language development. However particularly in BCLP palate repairs complicated by fistula where the risk of subsequent VPI is around 70%, consideration should be given to early closure of the fistula. This approach may reduce the risk of subsequent VPI and if required, will assist further speech surgical management with an intact palate.

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Table 1. The incidence of lateral palatal release, palatal fistula and subsequent palatal function at 5 years of age by cleft palate type.

Cleft Palate	**Number (%) of patients**	**Number (%) requiring lateral palatal release**	**Number (%) of postoperative palatal fistula**	**Number (%) with adequate palatal function at 5 years**
All CPO	157 (58%)	60/157 (38%)	13/157 (8%)	128/157 (82%)
-S-	36 (23%)	0/36 (0%)	1/36 (3%)	33/36 (92%)
h1/3S	38 (24%)	4/38 (11%)	1/38 (3%)	34/38 (89.5%)
h2/3S	41 (26%)	20/41 (49%)	3/41 (7%)	29/41 (70.7%)
HSH	42 (27%)	36/42 (86%)	8/42 (19%)	32/42 (76.2%)
UCLP	79 (29%)	62/79 (78%)	8/79 (10%)	63/79 (80%)
BCLP	35 (13%)	34/35 (97%)	7/35 (20%)	22/35 (63%)
All patients	**271 (100%)**	**156/271 (57%)**	**28/271 (10.3%)**	**213/271 (79%)**

(CPO: Cleft Palate only; -S-: cleft of soft palate; h1/3S: cleft of soft palate and posterior one third of hard palate; h2/3S: cleft of soft palate and posterior two-thirds of hard palate; HSH: complete cleft of hard and soft palate; UCLP: unilateral cleft lip and palate; BCLP: bilateral cleft lip and palate).

Table 2. Association between postoperative fistula and subsequent velopharyngeal function by cleft palate type.

Cleft Palate	**Fistula**		**No fistula**	
	VPI	No VPI	VPI	No VPI
CPO (n=157)	6 (46%)	7	23 (16%)	121
UCLP (n=79)	4 (50%)	4	12 (17%)	59
BCLP (n=35)	5 (71%)	2	8 (28%)	20
All patients (n=271)	15 (54%)	13	43 (18%)	200

VPI : velopharyngeal insufficiency; CP : cleft palate only; UCLP : unilateral cleft lip and palate; BCLP : bilateral cleft lip and palate.