

CASE REPORT

Longitudinal outcome of pharyngoplasty

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Abstract Although early complication of airway obstruction following pharyngoplasty is well recognised, there have been few reports of late modifications following this procedure. We retrospectively review cases with late complications which have required either revision or division of an existing pharyngoplasty at the Australian Craniofacial Unit over the last twenty-five years. We assess the outcome of further surgical intervention in each case, with case note and nasendoscopy video review. Fourteen cases were identified where records were complete. There were 12 males and 2 females. The cases are a heterogeneous group of cleft lip and palate patients and include three cases with a diagnosis of Pierre-Robin sequence and one case with a cleft palate as part of an underlying syndrome. Those cases requiring flap division had undergone either superiorly or inferiorly based pharyngeal flaps in contrast to dynamic (Orticochea) pharyngoplasties which required revision. This series of cases demonstrates the need for thorough assessment and planned tailoring of the pharyngoplasty procedure, with ongoing review of speech and airway function. This management philosophy results in the acceptance that a pharyngoplasty may only be required for a limited period of time and ultimately may be redundant.

Introduction

The development and application of high-resolution endoscopy to the study of velopharyngeal function has revolutionized the management of velopharyngeal dysfunction (David and Bagnall, 1990). Prior to its availability standard surgical treatment of velopharyngeal incompetence (VPI) was confined to either flap or sphincter pharyngoplasty. Generally the chosen technique was based on fluoroscopic analysis of velopharyngeal activity and the surgeon's clinical judgement and experience. It was then standard practice to leave the pharyngoplasty untouched except in the case of airway obstruction.

With improved imaging and visualization it has become possible to analyse all components of the velopharyngeal mechanism and tailor surgery to suit the dysfunction (David *et al.*, 1982). Longitudinal experience by clinicians dedicated to achieving excellence has led to an understanding of the need for continued refinement. This is particularly so when initial velopharyngeal surgery is undertaken during childhood before the

completion of growth and maturation of the motor speech system. We present fourteen cleft palate cases that had undergone pharyngoplasty and who have subsequently required either modification or had it taken down. We discuss their management, including the indications for further surgical intervention, and assess its outcome.

Materials and methods

Cases of cleft palate who had undergone revision or division of a pharyngoplasty were identified from the Australian Craniofacial Unit database. Case note review (along with video records of the nasendoscopy studies), were undertaken in all cases.

Results

Fourteen cases were identified where records were complete. Details of the cases presented are summarized in Table 1. There were 12 males and 2 females. Seven cases had isolated cleft palate, 3 had submucous cleft palate, 2 had unilateral cleft lip and palate and 2 had bilateral cleft lip and palate. In addition, three cases had Pierre Robin sequence and 1 case had Stickler syndrome. Four cases had primary pharyngoplasty surgery at other centres. Those who had undergone Orticochea pharyngoplasty

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Table 1: Summary of the cases

Case no.	Sex	Diagnosis	Pharyngoplasty type	Age (Years)	Revision/ Division	Age (Years)	Time to Revision (Years)
1	m	ICP	Sup. Pharyngeal flap	4*	D	10	6
2	f	ICP-Stickler	Sup. Pharyngeal flap	8	D	18	10
3	m	ICP-PR	Sup. Pharyngeal flap	5	D	14	9
4	m	SMCP	Sup. Pharyngeal flap	14	D	19	5
5	m	UCLP	Sup. Pharyngeal flap	10	D	31	21
6	m	ICP	Sup. Pharyngeal flap	5*	D	11	6
7	m	ICP	Sup. Pharyngeal flap	10	R	16	6
8	m	SMCP	Orticochea	4	R	16	12
9	m	BCLP	Orticochea	7	R	18	11
10	m	ICP-PR	Orticochea	4	R	7,10,14	3, 3, 4
11	m	ICP-PR	Orticochea	10	R	17	7
12	f	SMCP	Orticochea	5	R	9	4
13	m	BCLP	Inf. Pharyngeal flap	17*	D	39	22
14	m	UCLP	Inf. Pharyngeal flap	7*	D	17	10

*Pharyngoplasty performed elsewhere

Diagnosis: ICP = Isolated Cleft Palate

SMCP = Submucous Cleft Palate

UCLP = Unilateral Cleft Lip and Palate

BCLP = Bilateral Cleft Lip and Palate

PR = Pierre Robin sequence

(Orticochea, 1968) required revision, while those with superiorly or inferiorly based pharyngeal flaps underwent division in all cases except one.

Discussion

There has been few reported cases about the need for revision following pharyngoplasty. We have identified a group of cases all of which initially benefited from their pharyngoplasty, with no case requiring any intervention in less than three years following primary surgery. This is different from reports from other centres where the initial surgery has failed (Ma *et al.*, 1996) or required revision within two years of the primary surgery (Kasten *et al.*, 1997; Pryor *et al.*, 2006).

The cases presented in this series have had different indications for modification of their pharyngoplasty. The cases are heterogeneous cleft palate population and the only finding of note was that most of isolated cleft palates had the Pierre Robin sequence. However, this heterogeneous population can largely be divided into three groups as to the reason for further interventions: airway obstruction alone, speech difficulties (unresolved VPI), and combined airway obstruction with speech difficulties. Each case will be considered according to the indication for revision.

Group 1. The smallest group consisted of just one subject (case 5) who required modification to the pharyngoplasty due to a compromised airway. This subject had a repaired UCLP and at age 10. He underwent a superior pharyngeal flap following assessment of nasal air escape affecting speech and confirmed by nasendoscopy. He re-presented at age 30 with recent history of sleep apnoea requiring C-PAP. Speech was assessed as mildly to moderately de-nasal with no evidence of VPI or hypernasality. Nasendoscopy demonstrated severe nasopharyngeal

obstruction (Figure 1). The components of the velopharyngeal sphincter were noted to be functioning appropriately during speech and the flap was deemed redundant. He subsequently underwent division of the flap and on reassessment he no longer complained of snoring, did not fall asleep at work and the sleep study was normal.

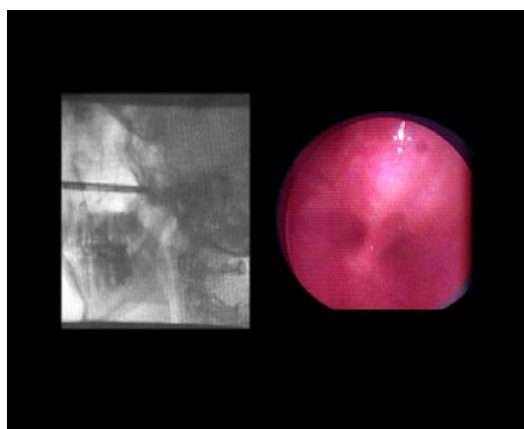


Figure 1: Case 5 Nasendoscopy age 31 demonstrating low attachment of the superior pharyngeal flap at rest prior to division.

Group 2. The largest group which had a total of eight subjects who required surgical revision as part of their management of VPI (cases 4,7,9,10,11,12,13,14).

Case 4 in this group had a superiorly based pharyngeal flap at age 14 after submucous cleft palate repair. This young male had a mild intellectual deficit and considerable debate took place prior to recommending surgery as nasendoscopy demonstrated minimal muscular effort. His poor speech had contributed to his social isolation and severe expressive language difficulties. Eventually it

was agreed that the purpose of the flap would be to partially obturate the isthmus, thus increasing vocal energy and loudness, providing this young man with more intelligible conversational speech. The desired outcome was achieved but the flap was divided four years later at the time of Le fort 3 maxillary advancement. Post-operative assessment including nasendoscopy did not demonstrate any deterioration, which have been previously noted (Harries *et al.*, 1992).

Case 7 had a superiorly based pharyngoplasty following assessment of gross hypernasality and audible nasal air emission supported by observations at nasendoscopy. Post-operative hypernasality was improved and assessed as mild. Six years later after speech deteriorated, the flap was revised. Post-operative assessment demonstrated mild denasality, no hypernasality but minimal nasal air emission through a palatal fistula. The patient declined further surgery.

Three cases (9, 10 and 11) had Orticochea sphincter pharyngoplasties performed at ages 4, 7 and 10 years respectively, with nasal air emission accompanying speech and assessed as ranging from mild to moderate. The surgery resulted in reduced nasal air emission but none was fully competent. Case 9 underwent a superiorly based pharyngeal flap eleven years after the Orticochea in an effort to fine-tune the closure. Case 10 had undergone two posterior pharyngeal wall implants, of which the second time was with pre-operative tattooing (Maegawa *et al.*, 1998), coordinated with therapy for hypernasality. His speech improved and assessed as normal in resonance with effective sphincter closure. Speech assessment four years later demonstrated VPI and recurrence of hypernasality, which appeared to coincide with a period of teenage skeletal and soft tissue growth. Nasendoscopy demonstrated incompetence and he underwent a revision of the Orticochea pharyngoplasty and a short follow-up course of resonance therapy. Review one year later confirmed no nasal air emission or hypernasality. Case 11 underwent nasendoscopy seven years after his Orticochea pharyngoplasty because his speech was noted to be less intelligible. Endoscopy revealed the left flap had become detached. This was surgically re-positioned and his speech clarity improved.

Case 12 was referred for investigation of a speech disorder and subsequently diagnosed with a submucous cleft palate. She underwent Veau-Wardill-Kilner cleft palate repair. Post-operatively nasendoscopy demonstrated a central defect but good lateral pharyngeal wall movement. On-going speech reviews reported limited speech improvement so one year later the patient underwent an intravelar veloplasty and Orticochea pharyngoplasty. The following year increasing velopharyngeal incompetence and hypernasality was noted. A nasendoscopy

found the pharyngoplasty was functioning well but the tonsils were large and inhibiting velar elevation. A tonsillectomy was performed. Post-operatively, although improved, speech featured nasal air emission on the production of high pressure oral phonemes. Two years later nasendoscopy demonstrated maintained function but the velopharyngeal gap was significant and central pharyngoplasty was recommended. This was undertaken 2 weeks later and speech review four months post-operatively found tight velopharyngeal closure on demanding testing, marked improvement in intelligibility but persisting mild assimilative hypernasality. She remains under review.

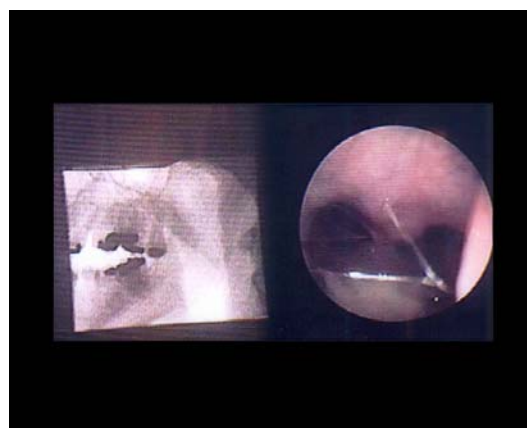


Figure 2: Case 13 Nasendoscopy age 39 years demonstrating the inferior pharyngeal flap at rest with its low attachment prior to division.

Two cases (13 and 14) had inferiorly based central flaps. Both had primary early cleft management and pharyngoplasties in other centres (they first presented at 17 and 39 years of age respectively). Tethering of the velum was observed at nasendoscopy in both cases (Figure 2), and both underwent division of the flap. Post-operative nasendoscopies clearly demonstrated improvement in velar activity with firm or touch closure. Speech assessment supported the finding of nasendoscopy and recommended therapy to decontaminate the speech of hypernasality. While there are a relatively large number of superiorly based pharyngeal flaps in this series (seven), this procedure was commonly undertaken by the department during the period of study. In contrast the two cases of inferior pharyngeal flaps in this series were the only examples managed in the unit during the period of study. That both of these cases required division suggests that this flap could be particularly prone to requiring later modification.

Group 3. The five cases in this final group underwent surgery for combined airway and speech difficulties (cases 1, 2, 3, 6 and 8). Case 1 had a superior pharyngeal flap at age 4 years at another centre and nasal air emission and hypernasality were reportedly eradicated.

Speech assessment at this Unit at age 10 years revealed moderate to severe denasality and snoring. Nasendoscopy demonstrated a wide flap obstructing the nasopharynx and tethering the velum. All components of the mechanism worked well suggesting competence would probably be maintained if the flap was divided. Speech assessment following division revealed no hypernasality, mild denasality and no detectable nasal air emission. Snoring was eradicated.

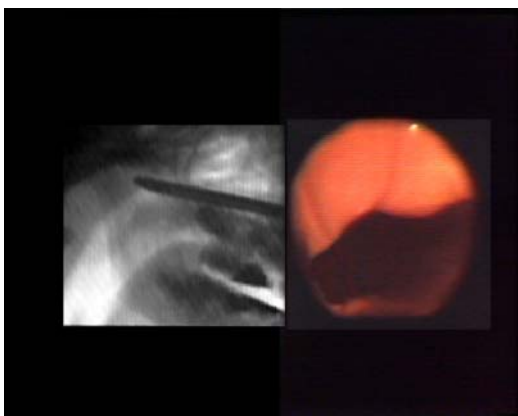


Figure 3: Case 2 Nasendoscopy at age 7 years demonstrating large gap during speech.

Case 2 had a superiorly based pharyngeal flap at age 8 following identification of a velopharyngeal defect during nasendoscopy (Figure 3). Post-operative nasendoscopy was satisfactory. The patient presented at age 18 for investigation of VPI and snoring. Nasendoscopy demonstrated a wide flap with small lateral ports compromising breathing (Figure 4), and speech assessment identified a mixed resonance disorder with denasality and audible nasal air emission accompanying sibilants. The pharyngoplasty was divided and post-operative speech assessment confirmed improvement with a reduction in de-nasal resonance and snoring, and an increased nasal airway for breathing.

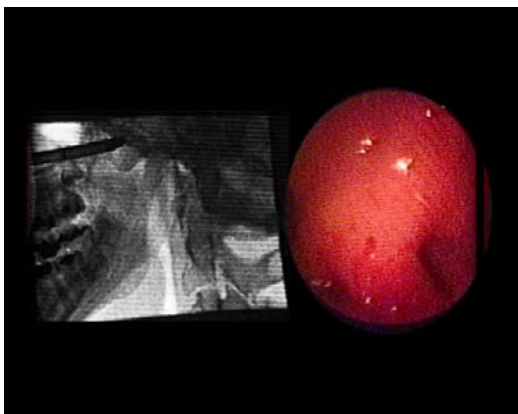


Figure 4: Case 2 Nasendoscopy following pharyngoplasty at age 17 years demonstrating diminutive lateral ports at rest.

Case 3 demonstrates issues inherent in a central flap pharyngoplasty in cases with a diagnosis of Pierre-Robin syndrome. This boy was treated at another center and had his primary palate repaired at age 16 months and a pharyngoplasty due to severe velopharyngeal incompetence at age 5 years on the speech pathologist's recommendation. The surgeon chose to provide a superior pharyngeal flap. Over the next few years his speech improved with eradication of nasal air emission but denasality gradually began to dominate his speech pattern. A speech assessment nine years later in this unit when the patient was aged 14 years identified severe denasality and reports of consistent severe snoring. Nasendoscopy demonstrated small ports and obstruction due to the flap. Five weeks later the flap was divided and post-operative speech assessment at 3 months noted reports of clearer speech, a reduction in snoring, mild denasality and velopharyngeal competence.

Case 8 first presented at age 4 years with an unrepaired submucous cleft palate. He had simultaneous palate repair and Orticochea pharyngoplasty. Six years later he gave a history of increasing denasality and underwent a nasendoscopy. Revision was recommended but not carried out. He presented twice more during childhood and received similar advice. At age 16 he finally underwent pharyngoplasty revision during which the Orticochea flaps were re-positioned superiorly. Post-operative nasendoscopy and speech assessment confirmed improved airway with adequate competence for speech and perceptible improvement to speech intelligibility. He remains under review.

The last case in this group (Case 6) was again initially treated elsewhere. His soft palate cleft was repaired at 6 months of age and the un-repaired hard palate defect was obturated, though not successfully, for speech. At age 5 years he underwent hard palate repair and simultaneous central pharyngoplasty. Reported speech improvement but persisting velopharyngeal incompetence was noted. He attended this Unit at age 10 years for an initial assessment that revealed de-nasal speech, hypernasality and nasal air emission. Nasendoscopy findings confirmed that the superior pharyngeal flap was attached too low to be effective. Given that the patient had good lateral pharyngeal wall movement it was decided to divide the flap at the same time as the palatal fistula was repaired. Speech review ten months later reported no change in hypernasality or nasal air emission but eradication of denasality and reliance on mouth breathing. He is due to undergo nasendoscopy to plan the next phase of management.

Conclusion

A series of heterogeneous group of cleft palate patients who have undergone pharyngoplasty

and a subsequently required surgical modification were reviewed. Results showed that the surgery undertaken, superior (and especially) the inferior, pharyngeal flaps may become redundant and require taking down. However, the Orticochea flaps may require revision of the position of their flaps. Following these surgical interventions the speech has been improved but has not achieved complete resolution of symptoms in all cases.

References

- David DJ and Bagnall AD (1990). Velopharyngeal incompetence. In: McCarthy JG (ed.), *Plastic Surgery: Volume 4, Cleft Lip & Palate and Craniofacial Anomalies*, 1st edn. New York: W.B. Saunders Co., pp. 2903-2921.
- David DJ, White J, Sprod R and Bagnall AD (1982). Nasendoscopy: significant refinements of a direct-viewing technique of the velopharyngeal sphincter. *Plast Reconstr Surg*, **70**(4): 423-428.
- Harries RHC, Moore MH, Bagnall AD and David DJ (1992). Posterior pharyngeal tattooing: localising the implant in velopharyngeal incompetence. *Br J Plast Surg*, **45**(4): 324-326.
- Kasten SJ, Buchman SR, Stevenson C and Berger M (1997). A retrospective analysis of revision sphincter pharyngoplasty. *Ann Plast Surg*, **39**(6): 583-589.
- Ma L, James DR and Sell DA (1996). Failed pharyngoplasty and subsequent management. *Br J Maxillofac Surg*, **34**(5): 348-356.
- Maegawa J, Sells RK and David DJ (1998). Pharyngoplasty in patients with cleft lip and palate after maxillary advancement. *J Craniofac Surg*, **9**(4): 330-335.
- Orticochea M (1968). Construction of a dynamic muscle sphincter in cleft palates. *Plast Reconstr Surg*, **41**(4): 323-327.
- Pryor LS, Lehman J, Parker MG, Schmidt A, Fox L, Murthy AS (2006). Outcomes in Pharyngoplasty: a ten year experience. *Cleft Palate Craniofac J*, **43**(2): 222-225.