Facial growth after different methods of surgical intervention in patients with cleft lip and palate

Gunvor Semb and William C. Shaw

Department of Oral Health and Development, University Dental Hospital of Manchester, Manchester, UK, and University of Oslo, Dental Unit, Department of Plastic Surgery, Rikshospitalet, Oslo, Norway

Semb G, Shaw WC. Facial growth after different methods of surgical intervention in patients with cleft lip and palate. Acta Odontol Scand 1998;56:352–355. Oslo. ISSN 0001-6357.

Studies of the influence of surgery on facial growth in cleft lip and palate must take into account inherent variations in craniofacial form independent of surgery. Primary surgery, the most important iatrogenic influence on facial form, can differ in technique, timing, and sequence, and one of the major challenges in researching the topic is the remarkably varied clinical protocols in current use. Unfortunately, systematic attempts to compare dentofacial outcomes reported in the literature are unlikely to be reliable, as methodologic biases cannot be overcome. Rigorous intercenter studies can improve the dependability of data and provide evidence of the success of cleft services as a whole, but they are still subject to biases introduced by differences in surgical skills and underlying craniofacial form. These shortcomings are finally being overcome through multicenter randomized control trials. □ *Cephalometrics; cleft lip and palate; outcomes; surgery*

Gunvor Semb, Department of Oral Health and Development, Unit of Orthodontics, University Dental Hospital of Manchester, Manchester M15 6FH, UK

Increased interest in the association between cleft surgery and facial growth, and particularly the quest for a 'best method', tends to overshadow important, preexisting variations in the faces of children with cleft lip and palate (CLP). However, surgery is only one of several influences on facial growth (for review, see 1):

1) Congenital dysmorphology. The facial deformity present at birth represents a combination of the initial embryonic defect and subsequent changes occurring in utero. In complete unilateral clefts (UCLP) the features are maxillary asymmetry, an upward tilt of the premaxillary region, and a distortion of the nasal septum. In bilateral clefts (BCLP) the premaxilla can be extremely protruded.

2) Other variations intrinsically associated with the cleft. Comparisons of craniofacial dimensions of adult unoperated patients with CLP and non-cleft patients have shown that patients with CLP have smaller craniofacial dimensions, both jaws are smaller, the posterior face height is reduced, and the lower anterior face height is increased. The mandible has an increased gonial angle, a steep mandibular plane, and a retrusive position (2, 3).

3) Functional adaptations. Patients with CLP have impaired nasal airways, and there is evidence that nasal deficiency is present in utero (4). Hairfield et al. (5) found that 68% of subjects with CLP are predominantly mouthbreathers. It is possible that airway insufficiency is a factor in the mandibular anomalies evident in CLP, in both operated and unoperated subjects.

4) Surgical iatrogenesis. Although attention was drawn to the dramatic effect of surgically induced growth impairment more than 50 years ago (6, 7), only unsubstantiated theories of the mechanism of disturbance are available: scar tissue and tension that distort the action of the functional matrix (though the precise nature and role of the matrix remain obscure), scar tissue that prevents the normal separation of sutures as growth attempts to proceed (under growth influence elsewhere), or damage to primary growth sites or centers (whatever they may be). Whatever the mechanism, maxillary growth impairment becomes progressively apparent as patients reach maturity (Fig. 1). This is superimposed on the underlying facial differences noted above and is mainly confined to the maxillary base and arch. The maxilla is shorter and is posteriorly displaced relative to the cranial base, although it also appears to be deficient in the posterior vertical dimension. The transverse dimension of the basal maxilla does not seem to be affected by surgery, but the dental arches are highly affected. There may be an inward deflection of the dentoalveolar processes, resulting in anterior and transverse crossbites. It is likely that different surgical techniques give rise to malocclusions of different extent without necessarily altering the neighboring structures.

Controversy still exists about whether it is lip or palatal surgery that is the major cause of the growth impairment. Two separate studies of three groups of subjects with UCLPĐone unoperated, one having had lip surgery only, and one having had both lip and palatal surgery (2, 11) show that lip closure alone has a major influence on maxillary development. For example, the length of the maxilla (Ar-ANS) was reduced after lip surgery by 6.3 mm in a Sri Lankan sample (2) and 4 mm in a Brazilian sample (11). With lip and palate surgery the additional mean reduction in maxillary length was only a further 1.0 mm and 0.7 mm, respectively. However, because subjects who have had only palatal surgery are not available, this 'experiment' cannot be considered to be complete.

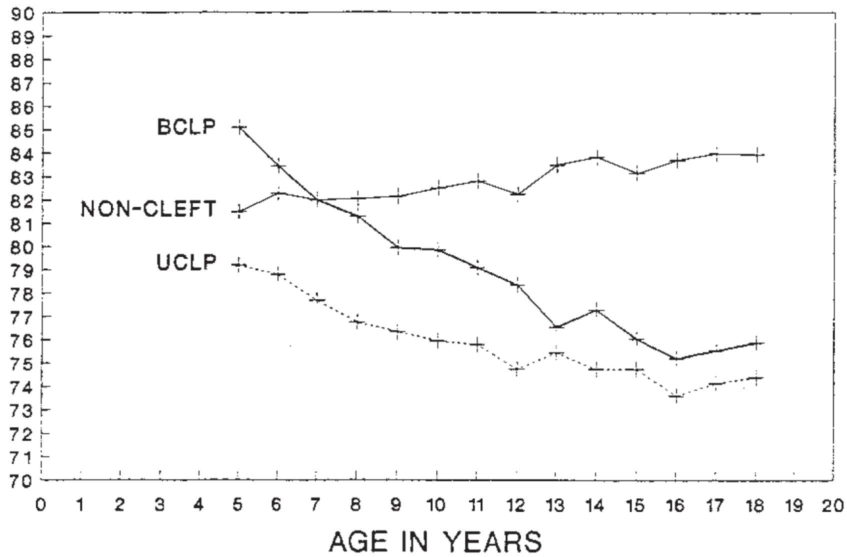

Fig. 1. Changes in maxillary prominence (SNA, s-n-ss) from 5 to 18 years of age for a pooled male and female sample of 257 patients with complete unilateral cleft lip and palate (UCLP) (8) and 90 patients with complete bilateral cleft lip and palate (BCLP) (9), compared with a pooled Bolton sample (non-cleft) (10).

Thus, in the search for optimal surgery, lip and palate surgery deserve equal attention.

What we should be comparing

If we concentrate on primary surgery, undoubtedly the most important influence on late dentofacial development, the key variables are 1) technique, that is, the technical design of the surgical repair, often a specific series of anatomic dissections and unions but modified appropriately for the individual case (and sometimes interpreted with considerable individuality by different surgeons); 2) timing, the age at which operations are done; 3) sequence, the order in which they are done; 4) whether or not the repairs are preceded or followed by orthopedic appliances; and 5) surgical skill, potentially highly variable between one surgeon and the next, and possibly the most important variable of all (12).

Relevant outcomes

The ultimate goal of cleft care is to prepare the patient for a full, integrated social life, unimpeded by physical or psychologic handicap. This important goal and its relation to surgery per se constitute a highly challenging research question that is not yet answerable. For the time being, the choice of intervention has to be a balance between the burden imposed by treatment and the reliability and acceptability of the clinical outcomes achieved. The mix of relevant variables would include, at least, nasolabial appearance, speech, hearing, psychosocial development, and dentofacial development. Although the last is the focus of the present article, it is not necessarily the most important, and balancing trade-offs between outcomes may be an important issue.

The challenge: amazing diversity

The Eurocleft Biomed II Project is working on producing a register of European CLP teams and their surgical protocols. Although the details from all teams are not yet complete, the register currently consists of 178 teams with 171 different protocols for UCLP alone. There are a myriad of different techniques for closing the lip and the hard and soft palate (and often surgeons have their own modification of a named technique). The timing of surgery shows extreme variation: the range for lip closure is from birth to 9 months; for soft palate closure, from birth to 3 years; and for hard palate closure, from birth to 13 years. The sequence of the different surgeries also varies considerably: some start with lip closure only, while others first close the soft palate, leaving the lip closure till a later date. Some teams close the lip and hard palate simultaneously, whereas others close the lip and the soft palate simultaneously. Indeed, all possible combinations seem to be practiced somewhere. Five percent of teams close the whole cleft in one operation, 65% use two operations, 29% use three, and 1% use four. Further, 45% of teams use presurgical orthopedics, 13% do nose surgery together with lip surgery, and 5% use lip adhesion before definitive lip closure. Critics might describe this huge diversity of clinical practices in cleft surgery as clinical chaosÐand a compelling reason for the urgent establishment of an adequate 'evidence base' from which future patients could benefit. Unreliability of comparisons The fundamental difficulty in comparing growth from samples of cases reported in the literature, treated by different methods, is one of potential biases. These include the following:

1) Susceptibility bias. Were the samples truly equivalent, with equivalent prognosis before surgery? That is, were the populations similar in respect of underlying facial pattern and type and severity of cleft?

2) Proficiency bias. Were the operations done by surgeons with precisely the same skill and protocol adherence? For example, if method A is 5% better than method B, but surgeon X is 10% better than surgeon Y, the true relative merits of a protocol will be obscured.

3) Detection bias. Were all cases followed and analyzed, even the bad ones?

4) Exclusion bias. Have the rules for excluding subjects, such as those with other syndromic or 'potentially syndromic' features, been applied in the same way for both groups?

5) Analysis bias. Have the outcomes been assessed according to a standardized protocol ensuring equivalent averaging of age, gender mix, cephalometric technique, and interpretation?

6) Reporting bias. When results are disappointing, are they as likely to be reported as when they are pleasing? Since the above questions cannot be answered in retrospective studies, how can we make the best of the literature?

Making the best of the literature Systematic reviewing is a powerful tool for synthesizing reports in the literature concerning particular therapeutic interventions (13). It involves a formalized strategy for identifying relevant papers, predetermined inclusion/ exclusion criteria, an objective appraisal of the quality of the study and its reporting, and an aggregation of data for two or more comparison groups followed by statistical comparison (possibly meta-analysis). In an attempt to apply these principles to published reports of cephalometric analysis of patients with repaired complete CLP, the search strategy included 1) searching the electronic databases Medline and Embase from 1966 to 1997; 2) hand-searching the Cleft Palate-Craniofacial Journal from 1964 to 1997; 3) searching the Cochrane Trials Register; and 4) following up relevant citations in the papers read.

We included any publication in English concerning samples of Caucasian non-syndromic UCLP and BCLP cases that reported one or more of three common variables: maxillary prominence s-n-ss (SNA), maxillo- mandibular relation sagittally ss-n-sm (ANB), and maxillo-mandibular relation vertically (NSL/ML). Cases with soft tissue band were considered eligible and their presence was listed when reported by the authors. We regarded the following as grounds for exclusion: samples of fewer than 15 cases; samples that included a mixture of UCLP and BCLP, since they are inherently different; and samples in which the actual ages of subjects were dispersed widely on either side of the mean age reported.

In all, 5474 titles were identified, and 189 relevant reports were found. Preliminary analysis suggests that about 20% of the relevant papers are eligible. The reports vary considerably in the reported detail of methodologic rigor. Only some reported plotting or digitizing reproducibility. More significantly, perhaps, it was often unclear whether the data represented consecutive or randomly selected cases, or whether there was an alternative basis upon which cases had been included. Individual reports had varied approaches to determining age, some including a range of 6–33 years (14).

The three papers reporting the highest level of maxillary prominence (SNA) had very different protocols (14–16): 1) 'Lip closed by triangular/rectangular flap at 3 months. The soft and hard palate are closed at 18 months using a push-back technique or von Langenbeck technique. The alveolus is left open.'

2) 'Soft palate closure at 3 months: the muscles are detached from the hard palate and sutured in the midline, the hamuli are fractured and a small mucosal vomerflap is used to close the cleft at the level of the posterior margin of the shelves. The mucoperiosteum is left untouched. Lip, alveolus and hard palate are then closed simultaneously at 6 months. Two mucoperiosteal flaps, (one from the nasal side of the palatal shelf and one from vomer) are used to close the hard palate. The lip is closed using a single or double equilateral triangular flap technique.'

3) 'Lip closure at 3 months using a definitive triangular flap. Then at 12 months the hard palate was closed using a vomerflap [details of alveolar surgery not reported]. At 18 months of age the soft palate was closed using intravelar veloplasty, and either simple midline closure or closure with V-Y lengthening.'

The value of intercenter studies Intercenter studies cannot eliminate susceptibility or proficiency bias (the patients are drawn from different populations and the surgeons are inevitably different). Thus, they are not appropriate for studying individual aspects of surgical or other protocols. However, with appropriate prospective planning they can limit detection, exclusion, analysis, and reporting bias, permitting a reliable indication of the overall level of outcome achieved by the entire service, including protocols and proficiency (12). This, in turn, can be a powerful stimulus for an overhaul of services that have proved deficient (17). Intercenter studies can also provide an opportunity for comparing the costs, complexity, and burden of care. They may promote openness, encourage cooperation (rather than competition), and promote collaborative work on outcome methodology (18–20). Above all, the findings of intercenter studies can be a powerful stimulus for randomized control trials.

Randomized trials of cleft surgery are possible

Randomized control trials are an essential tool for testing the efficacy of different interventions (21). The proper use of randomization provides the best guarantee that there is no bias in the selection of patients for the different treatments and helps considerably to reduce the risk of differences in the clinical environment. Randomized allocation in cleft care is not difficult to implement and enables trial conclusions to be more believable than other forms of treatment allocation (22–24). In a current trial of pharyngeal flap versus pharyngoplasty, all patients (parents) under the Oslo CLP Team requiring surgery for velopharyngeal insufficiency have since 1993 accepted randomization to alternative surgeries. Similarly, all but one patient (parent) with UCLP attending for primary surgery with the teams in Aarhus/Copenhagen (serving all of Denmark), Bergen and Oslo (serving all of Norway), Helsinki (serving all of Finland), Göteborg, Linköping, and Stockholm have accepted randomization for their primary surgery (and so have all of the surgeons) since 1997. The search for optimum surgery in respect of dentofacial growth is valid and deserves the urgent attention of cleft teams. Future work, however, must capture the full range of issues and particularly the burden endured by patients who are subjected to complex and demanding protocols. Accumulation of a reliable 'evidence base' will not be achieved by the traditional methods of the past, but requires instead concerted international cooperation and the adoption of clinical research protocols that meet contemporary standards.

References

1. Semb G, Shaw WC. Facial growth in orofacial clefting disorders. In: Turvey TA, Vig KWL, Fonseca RJ, editors. Facial clefts and craniosynostosis. Philadelphia: W.B. Saunders; 1996. p. 28–56.
2. Mars M, Houston WJB. A preliminary study of facial growth and morphology in unoperated male unilateral cleft lip and palate subjects over 13 years of age. Cleft Palate J 1990;27:7–10.
3. Capelozza L Jr, Taniguchi SM, da Silva OG Jr. Craniofacial morphology of adult unoperated complete unilateral cleft lip and palate patients. Cleft Palate Craniofac J 1993;30:376–81.
4. Siegel M, Mooney MP, Kimes KR, Todhunter JT. Analyses of size variability of the human normal and cleft palate fetus nasal capsule by means of three-dimensional computer reconstruction of histologic preparations. Cleft Palate J 1987;24:190–9.
5. Hairfield MA, Warren DW, Seaton DL. Prevalence of mouth breathing in cleft lip and palate. Cleft Palate J 1988;25:135–8. 6. Graber TM. A cephalometric analysis of the developmental pattern and facial morphology in cleft palate. Angle Orthod 1949;19:91–100.
7. Graber TM. The congenital cleft palate deformity. J Am Dent Assoc 1954;48:375–95.
8. Semb G. A study of facial growth in patients with unilateral cleft lip and palate treated by the Oslo CLP Team. Cleft Palate Craniofac J 1991;28:1–21.
9. Semb G. A study of facial growth in patients with bilateral cleft lip and palate treated by the Oslo CLP Team. Cleft Palate Craniofac J 1991;28:22–39.
10. Broadbent BH Sr, Broadbent BH Jr, Golden WH. Bolton standards of dentofacial developmental growth. St. Louis: C.V. Mosby; 1975.
11. Capelozza Filho L, Correa Normando AD, da Silva Filho OG. Isolated influences of cleft lip and palate surgery on facial growth: comparison of operated and unoperated male adult with UCLP. Cleft Palate Craniofac J 1996;33:51–6.
12. Shaw WC, Asher-McDade C, BrattstroÈm V, Dahl E, Mars M, Mùlsted K, et al. Intercentre clinical audit for cleft lip and palateÐa preliminary European investigation. In: Jackson IT, Sommerlad BC, editors. Recent advances in plastic surgery. No. 4. Edinburgh: Churchill Livingstone; 1992. p. 1–15.
13. Chalmers I, Altman DG. Systematic reviews. London: BMJ Publishing Group; 1995.
14. Ross RB. Treatment variables affecting facial growth in complete unilateral cleft lip and palate. 1. Treatment affecting growth. Cleft Palate J 1987;24:5–23.
15. Ross RB. Growth of the facial skeleton following the Malek repair for unilateral cleft lip and palate. Cleft Palate Craniofac J 1995;32:194–8.
16. Trotman CA, Long RE, Rosenstein SW, Murphy C, Johnston LE. Comparison of facial form in primary alveolar bone-grafted and nongrafted unilateral cleft lip and palate patients: intercentre retrospective study. Cleft Palate Craniofac J 1996;33:91–5.
17. Clinical Standards Advisory Group Cleft lip and/or palate: report of a CSAG committee. London: The Stationery Office; 1998.
18. Friede H, Enemark E, Semb G, Paulin G, A Êbyholm FE, Bolund S, et al. Craniofacial and occlusal characteristics in unilateral cleft lip and palate patients from four Scandinavian centres. Scand J Plast Reconstr Hand Surg 1991;25:269–76.
19. Shaw WC, Dahl E, Asher-McDade C, BrattstroÈm V, Mars M, McWilliam J, et al. A six-center international study of treatment outcome in patients with clefts of the lip and palate. 5. General discussion and conclusions. Cleft Palate Craniofac J 1992;29: 413–8.
20. Enemark H, Friede H, Paulin G, Semb G, A Êbyholm F, Bolund S, et al. Lip and nose morphology in patients with unilateral cleft lip and palate from four Scandinavian centres. Scand J Plast Reconstr Hand Surg 1993;27:41–7.
21. Pocock SJ. Clinical trials. A practical approach. Chichester: John Wiley & Sons; 1983.
22. Marsh JL, Grames LM, Holtman MD. Intravelar veloplasty: a prospective study. Cleft Palate J 1989;26:46–50.
23. Roberts CT, Semb G, Shaw WC. Strategies for the advancement of surgical methods in cleft lip and palate. Cleft Palate Craniofac J 1991;28:141–9.
24. Shaw WC. Ethical issues in the case of surgical repair of cleft palateÐcommentary. Cleft Palate Craniofac J 1995;32:277–80.