B-ENT, 2006, **2**, Suppl. 4, 44-50

Long-term follow-up of UCLP at the Reine Fabiola Children's Hospital

A. De Mey, G. Swennen*, C. Malevez**, M. George*** and A. L. Mansbach****

*Department of plastic surgery, Free University Brussels, **Department of maxillofacial surgery, Free University Brussels, ***Department of ENT surgery, Free University Brussels

Key-words. Long-term follow-up; all-in-one; unilateral cleft lip palate; craniofacial morphology; hearing; speech and velopharyngeal function

Abstract. *Long-term follow-up of UCLP at the Reine Fabiola Children's Hospital.*
Background: The purpose of this retrospective of prospectively acquired data was to evaluate and to compare global evolution in children with complete unilateral cleft lip and palate treated at the Brussels cleft centre following two different surgical treatment protocols.
Methods: A series of forty-four patients operated for non-syndromic complete unilateral cleft lip and palate were included in this study at the age of approximately ten years. Twenty-six children (17 males, 9 females) were treated according to the Malek surgical treatment protocol: the soft palate was closed at a mean age of 3 months, followed by simultaneous repair of the lip and hard palate at a mean age of 6 months. Eighteen children (15 males, 3 females) underwent one-stage "all-in-one" closure of the lip, hard and soft palate at a mean age of 3 months. Craniofacial morphology was evaluated by means of digital lateral cephalometric analysis. Cephalometric data were compared to a control, non-cleft group (n = 40) matched according to age. Data concerning otological status and speech were collected in the same series of children.
Results: Statistical analysis showed that the inclination of the maxillary (MxPVSN) plane to the anterior cranial base was significantly increased ($p < 0.001$) in both cleft groups compared to the non-cleft group and significantly increased ($p = 0.002$) in the Malek cleft group compared to the "all-in-one" cleft group. Otological status was not improved by an early complete closure but by close follow-up and the repeated placement of ventilating tubes.
Speech was found to be satisfactory in the majority of children of both groups at six years after speech therapy. Only 15% needed further surgery with pharyngeal flaps.
Conclusions: There were no significant differences in anteroposterior midfacial morphology between the Malek and "all-in-one" protocols at ten years of age. One-stage "all-in-one" closure resulted in less downward inclination of the maxillary plane to the anterior cranial base compared to the Malek protocol at ten years of age.
Early complete closure of the cleft resulted in no significant change in otological status or the occurrence of nasality. However, early complete closure of the cleft allowed for earlier intelligibility of speech compared to the staged later closure.

Introduction

Many different surgical treatment protocols and adjunctive orthopaedic/orthodontic procedures have been described for the treatment of children with unilateral cleft lip and palate (UCLP).
In the cleft centre of the Reine Fabiola Children's Hospital in Brussels, UCLP children have been treated for more than 20 years by primary lip closure at 6 months, followed by closure of the palate according to Veau Wardill at 18 months. A retrospective study showed bad anteroposterior growth in 66% of patients treated according to the latter protocol. As a result, since 1981, the Malek surgical technique and sequence, which consists of early soft palate closure at 3 months followed by simultaneous closure of the lip and hard palate at 6 months, has been adopted. Based on the experiences with the Malek protocol, one-stage "all-in-one" closure of the lip, hard and soft palate was introduced in 1988 in the Brussels cleft centre to treat UCLP children.

One-stage "all-in-one" closure as a repair for UCLP patients was first described in the literature by Farina[1] in 1958. Other publications by Davies[2] and Kaplan *et al*.[3-5] reported on the long-term results after "all-in-one" closure. The preliminary results of the Brussels cleft centre with this technique were presented at the 7th and 9th International Congresses on cleft palate and related craniofacial anomalies.[6,7]

Other occasional reports on one-stage repair in cleft patients came from Davies *et al*.,[8] Standoli

et al.,[9] Honigmann[10] and Honigmann *et al.*[11]

The purpose of this non-randomised prospective study was to compare craniofacial morphology speech and hearing after one-stage "all-in-one" closure with a cleft group treated according to the Malek surgical treatment protocol at the age of approximately ten years.

Material and methods

The inclusion criteria for this non-randomised study were the following:

1) complete unilateral cleft lip, alveolus palate; patients with small Simonart's bands were included;
2) consecutive cases;
3) no associated syndrome or mental retardation;
4) Caucasian ethnicity;
5) presurgical infant orthopaedics with passive removable appliances;
6) measurement of anterior and posterior cleft margins during initial primary cleft surgery;
7) BERA evaluation at 3 months before the first operation then audiological assessment by audiometry, tympanometry and otoscopy at 3 and 6 years;
8) speech evaluation at 18 months, 3 years and 6 years;
9) lateral cephalometric radiographs at 10 years;
10) all children must have received their primary and secondary surgery at the Reine Fabiola Children's Hospital in Brussels (ULB).

Patients

Between November 1981 and August 1991, a total of 98 children with complete UCLP were operated at the Reine Fabiola Children's Hospital in Brussels (ULB). A total of 44 (45.0%) fulfilled the inclusion criteria. Twenty-six children (17 males, 9 females) were operated in accordance with the Malek protocol; 13 had right and 13 left complete UCLP. Eighteen children (15 males, 3 females) underwent closure of their cleft following the "all-in-one" surgical procedure in the period from November 1987 to August 1991; 8 had right and 10 left complete UCLP. The cleft size was per-operatively measured with a calliper anteriorly at the alveolar cleft margins and posteriorly at the level of the posterior nasal spine. One-stage "all-in-one" closure was only performed when the posterior cleft distance was less than 12 mm; otherwise the "Malek" protocol was adopted.

"Malek" cleft group

The soft palate was closed using the Malek protocol at a mean age of 3.04 +/- 0.20 months. The malpositioned palatal muscles were detached from the palatal shelves and hamulus, correctly repositioned and sutured in the midline. The lip was repaired using the Tennisson-Randall procedure at a mean age of 6.15 +/- 0.67 months and the hard palate was closed using a vomer flap simultaneously with lip closure. Almost all primary surgical procedures (96.2%) were performed by two plastic surgeons; ML (Madeleine Lejour) and ADM performed 42.3% and 53.9% of the primary operations respectively. Children operated from 1985 onwards (n = 15, 57.7%) underwent McComb primary nose correction simultaneously with lip closure. The mean anterior cleft distance was 9 mm, and the mean posterior cleft distance was 10 mm. Tympanotomy was performed at the time of the primary operation depending on the result of the BERA.

"All-in-one" cleft group

One-stage "all-in-one" closure of the lip, hard and soft palate was performed at a mean age of 2.98 +/- 0.16 months. The lip was repaired using the Tennisson-Randall procedure; the hard palate was closed using a vomer flap, while the soft palate was closed with a veloplasty in accordance with Malek. Great care was also taken to reposition the palatal muscles correctly. Most of the surgical procedures were performed by one surgeon (ADM), who operated on 16 (88.9%) children. Primary McComb nose correction was performed in all children simultaneously with "all-in-one" closure. The mean anterior cleft distance was 6 mm, while the mean posterior cleft distance was 8 mm. The same procedure was performed on the ear as in the two-stage procedure.

Orthopaedic / orthodontic treatment

Both cleft groups underwent passive presurgical orthopaedics using a removable acrylic appliance. Presurgical orthopaedics were initiated immediately after birth in the "all-in-one" cleft group and approximately 6 weeks preoperatively in the "Malek" cleft group. Active removable appliances were used during the deciduous and mixed dentition periods to correct small anterior or lateral cross-bites if necessary.

Otological evaluation

Patients were examined preoperatively by BERA under light

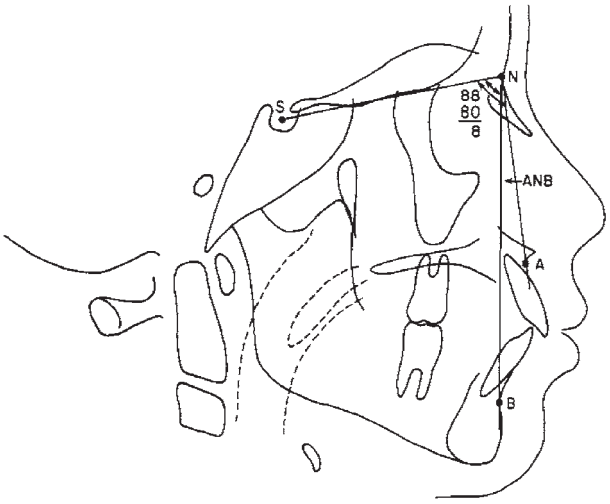

Figure 1
SNA /SNB measurement landmarks

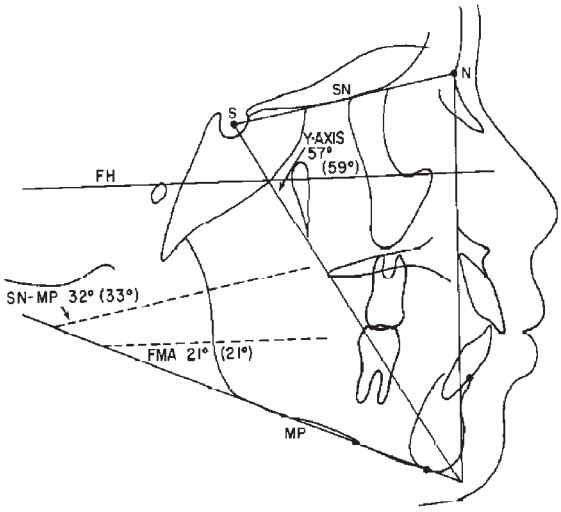

Figure 2
SN /Maxillary and mandibular plane measurement landmarks

sedation and then six weeks postoperatively. Later, complete evaluations were performed with audiometry, tympanometry and otoscopy at the ages of 3 and 6 years.

If necessary, patients found to have a hearing loss or recurrent otitis media were regularly monitored and given active treatment.

Speech evaluation

The first speech evaluation was performed at 18 months in spontaneous speech. Later, at 3 and 6 years, a spontaneous and standardised speech assessment in French or Flemish was performed by one senior speech therapist.

Digital cephalometry

Craniofacial morphology was evaluated using lateral cephalometric radiographs conducted at the age of approximately ten years in both groups (Figures 1,2). The cleft children were compared to a sample of 40 untreated children (20 males, 20 females) at a mean age of 10 years so that there was a reference group with normal skeletal relationships; these patients were randomly selected from the files of the Department of Orthodontics of Hannover Medical University.

Results

Cephalometric craniofacial morphology

The results of the cephalometric analysis are summarised in Tables 1 and 2.

Table 1 compares the "all-in-one" and "Malek" procedures: there are no significant differences for maxillary protrusion at the skeletal, dentoalveolar or dental levels. Anteroposterior maxillary position with respect to the cranial base was better in the "all-in-one" cleft group compared with the control group.

Table 2 compares the inclination of the maxillary plane with respect to the anterior cranial base: it was significantly increased ($p < 0.001$) in both cleft groups compared to the non-cleft group and significantly increased ($p = 0.002$) in the Malek group compared to the "all-in-one" group.

Speech and velopharyngeal function

Three parameters were studied: articulation, nasality and intelligibility.

Seventy per cent of the children in the Malek group and 63% of the "all-in-one" group received speech therapy between the ages of 3 to 6 years.

Articulation

At 3 years of age, 20% of the children presented with major problems (level 3) but these improved with time and 40% of the patients presented with a normal pattern (level 1) at the age of 6 years in both groups (Figures 3A,B).

Nasality

At 3 years of age, about 30% of the children in both groups had no nasality problems (level 1) but about 20% suffered from constant nasality (level 3).

Table 1
Evolution of anterio-posterior growth at 10 years of age

	MALEK	AIO	CONTROL
SNA	76.36	78.64	81.18
SNB	74.50	75.33	77.86
ANB	1.86	3.31	3.34

Table 2
Measurement of the maxillary/mandibular relationship at 10 years of age

	MALEK	AIO	CONTROL
Mx/SN	13.64	11.81	6.49
Md/SN	37.48	35.26	32.26

The situation was significantly improved at 6 years: 40% had no nasality problems at all and less than 10% suffered from constant nasality (Figures 4A,B).

The entire second group underwent a pharyngeal flap procedure.

Intelligibility

There was a significant difference between the two groups in this respect at 3 years of age.

In group I (two-stage closure) 52% of the children presented with a good intelligibility (level 1), 32% with a mild deficiency (level 2) and 18% with a major problem (level 3).

In group II (one-stage closure), 63% had no problems and none had a major problem. The difference is statistically significant ($p = 0.05$).

At 6 years however, no statistical difference was observed between the two groups and intelligibility was satisfactory in about 80% of the patients of both groups (Figures 5A,B).

Otological examination

At 3 months of age, before the first operation, we found only one patient with a threshold inferior to 25 dB, 6 between 25 and 40 dB and 37 over 40 dB.

Tympanic drainage was therefore required in the majority of our patients during the first operation.

During the follow-up, 28% needed only one drainage procedure, 40% two drainage procedures and 24% more than two procedures.

The comparison of audiometry at 3 and 6 years showed a difference between the two groups, with an advantage for the one-stage procedure at 3 years. However, this difference was not statistically significant (Figures 6A,B).

However, we observed significantly improved results between 3 and 6 years of age in a majority of the children with a threshold under 35 dB.

Secondary surgery

A total of eight (30.8%) patients in the Malek cleft group underwent lip correction at a mean age of 4.38 + 2.13 years. No lip revisions were performed in the "all-in-one" cleft group. Four (15.4%) patients in the Malek cleft group underwent a rhinoplasty consisting of the correction of the nostril

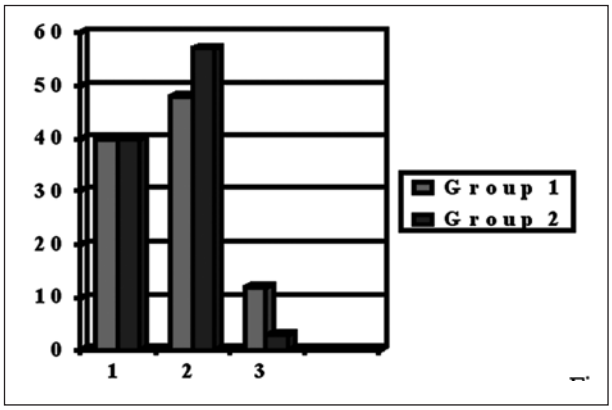

Figure 3A
Articulation at 3 years of age

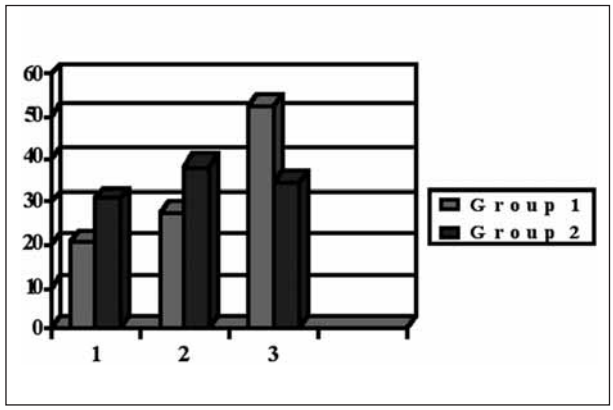

Figure 3B
Articulation at 6 years of age

X axis: Level 1: normal; Level 2: moderate disorder; Level 3: important disorder. Y axis: Percentage of patients in each level.

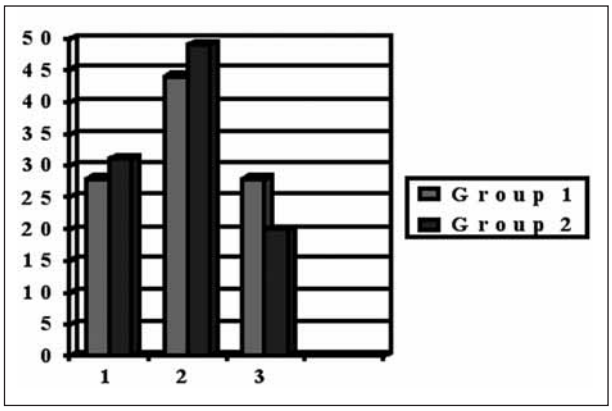

Figure 4A
Nasality at 3 years of age

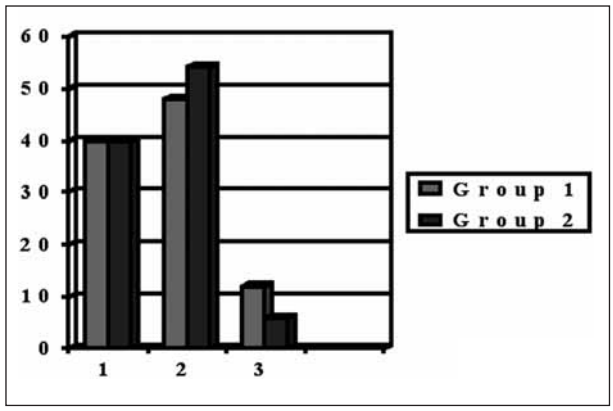

Figure 4B
Nasality at 6 years of age

X axis: Level 1: no nasality; Level 2: inconstant nasality; Level 3: constant nasality.

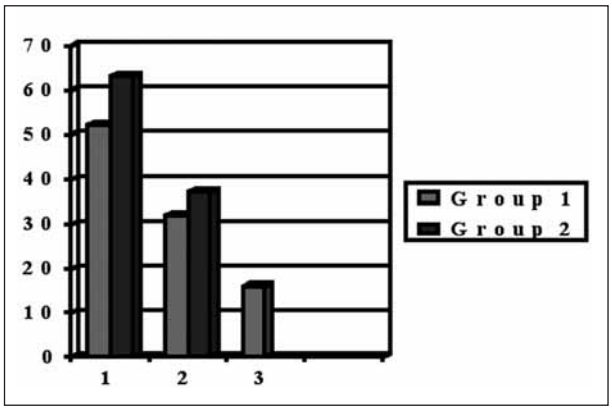

Figure 5A
Intelligibility at 3 years of age

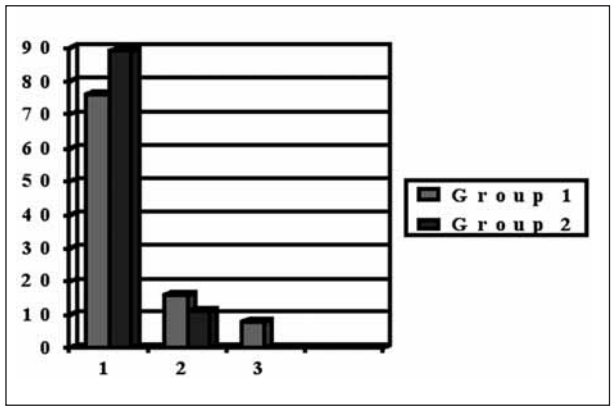

Figure 5B
Intelligibility at 6 years of age

X axis: Level 1: normal; Level 2: mild deficiency; Level 3: major deficiency.

and ala without septoplasty at about five years of age. In the "all-in-one" cleft group, a total of six patients underwent a rhinoplasty at a mean age of five years. This consisted of the correction of the nostril and ala in three patients, and septoplasty in another three patients. Velopharyngoplasty was conducted in three (11.5%) "Malek" cleft patients at a mean age of five years and in one (5.6%) "all-in-one" cleft patient at 5.5 years. In the "Malek" cleft group, a total of 17 fistula (8 oronasal, 9 vestibulonasal) closures were performed in 15 (57.7%) children at a mean age of 3.31 + 1.57 years, while in the "all-in-one" cleft group a total of eight (44.0%) patients had fistulae (7 oronasal, 1 vestibulonasal) that were all closed for phoniatric reasons at a mean age of five years. During the period of the study, secondary alveolar bone grafting was performed in eight (30.8%) patients in the "Malek"cleft group at a mean age of 8.40 + 1.05 years and in seven patients (39.0%) patients in the "all-in-one" cleft group at a mean age of 8.61 + 1.07 years. Another 14 (54.0%) "Malek" and six (33.3%) "all-in-one" patients underwent secondary alveolar bone grafting shortly after the investigation period at 12.3 + 1.66 and 10.6 + 1.31 years respectively.

Discussion

The focus of most reports describing the results of different treatment protocols for cleft lip and palate is on facial growth. However, a complete evaluation of a treatment protocol for cleft

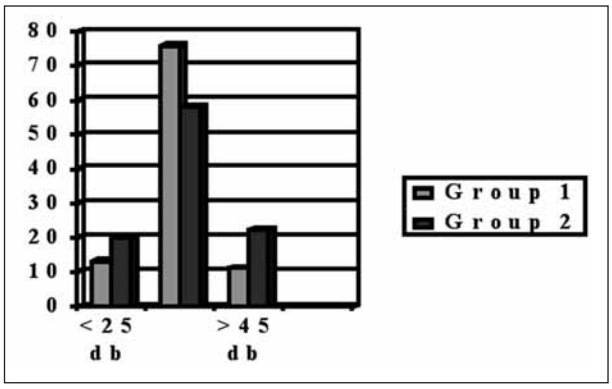

Figure 6A
Audiometry at 3 years of age

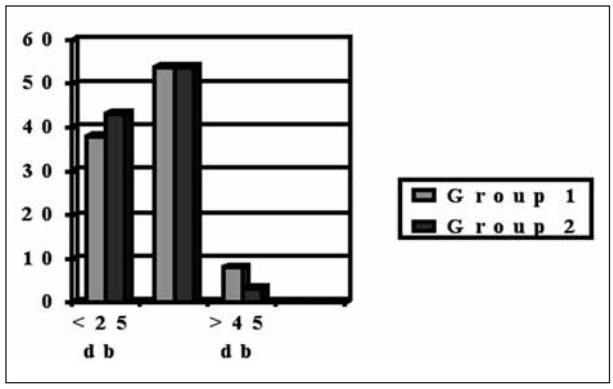

Figure 6B
Audiometry at 6 years of age

X axis: Level 1: threshold inferior to 25 dB; Level 2: threshold between 25 and 40 dB; Level 3: threshold over 40 dB.

patients is essential because it may have a specific influence on the different sequellae of the cleft and their rehabilitation.

One-stage "all-in-one" closure of cleft lip and palate patients was introduced by Farina[1] in 1958. However, since then, only a few long-term clinical studies have been described in the literature. Because none of these studies have presented reliable evidence-based data, Bardach *et al.*[12-14] developed an experimental model in rabbits and beagles to investigate the effect of simultaneous lip and palate repair on craniofacial growth. The results of these experimental studies indicated that simultaneous lip and palate repair caused more growth retardation than lip repair or palate repair performed separately or in sequence.

Our study evaluates and compares craniofacial morphology after one-stage "all-in-one" and two-stage cleft closure in UCLP patients at the age of approximately ten years. A digital lateral cephalometric analysis that was validated for cleft research in a previous study was used to analyse skeletal craniofacial morphology. The "all-in-one"/"Malek" comparison data revealed only one significant difference (MxPI/SN at $p = 0.002$) (Table 1). Although the maxillary plane (Mx/PI) was more open to the anterior cranial base (S-N) in both cleft groups compared to the non-cleft group (Table 2), it was significantly more open in the "Malek" cleft group compared to the "all-in-one" cleft group. This can be explained by the fact that posterior maxillary height (R-PMP) was significantly ($p < 0.001$) less in both cleft groups, whereas anterior maxillary (N-ANS) height was only significantly less in the "all-in-one" cleft group ($p = 0.001$) compared to the control group.

Early complete closure of the palate is considered to improve speech by giving the necessary tools to the child at the time of babbling.[15]

In our series, we observed that nasality was comparable with the two operative sequences.

Comparisons with other series are difficult since many criteria are subjective and different studies look at different types of clefts.

The incidence of pharyngoplasty is similar to that reported by Myklebust *et al.*[16] after closure of the palate at two years but higher than that reported by Malek.[17]

The otological status of cleft patients is frequently overlooked, if not forgotten. Nevertheless, as reported by Robinson *et al.*[18] in his series, the incidence of otitis media with effusion is high and the possibility of acquiring a good level of speech depends directly on the quality of hearing. We observed that an early complete closure does not improve otological status. This conflicts with previous reports in the literature.[19,20] However, careful follow-up, with the repeated placement of ventilating tubes, enabled us to obtain a satisfactory hearing level in most of our patients.

Conclusion

One-stage "all-in-one" closure could provide several substantial advantages for the treatment of CLP patients:

– only one anaesthesia and hospitalisation for primary surgery, with minimisation of potential risks and less psychological stress for the child and parents;

– less growth disturbance due to less scar tissue;
– early functional reconstruction of the palate with the possibility of the earlier acquisition of better speech and hearing;
– good cost/benefit ratio and ideal for countries where the health system cannot afford multi-stage multidisciplinary treatment.

Although this paper presents long-term quantitative data for multidisciplinary outcome after one-stage "all-in-one" closure at 10 years of age, the evaluation of craniofacial morphology at the end of skeletal growth, combined with dental cast analyses, speech and hearing evaluations, are mandatory in the future for a larger number of patients.

References

1. Farina R. Total unilateral harelip: correction of severe deformity of the palate & lips in a single operation; Le Mesurier's cheiloplasty & Veau-Ernst's gnatho-urano-staphyloplasty [in French]. *Ann Chir Plast*. 1958; 3:199-205.
2. Davies D.The one-stage repair of unilateral cleft lip and palate: a preliminary report. *Plast Reconstr Surg*. 1966;38:129-136.
3. .Kaplan I, Dresner J, Gorodischer C, Radin L. The simultaneous repair of cleft lip and palate in early infancy. *Br J Plast Surg.* 1974;27:134-148.
4. Kaplan I, Taube E, Ben-Bassat M, Dresner J, Nachmani A, Rosenbaum M. Further experience in the early simultaneous repair of cleft lip and palate. *Br J Plast Surg.* 1980;33:299-300.
5. Kaplan I, Ben-Bassat M, Taube E, Dresner J, Nachmani A. Ten-year follow-up of simultaneous repair of cleft lip and palate in infancy. *Ann Plast Surg*. 1982;8:227-228.
6. De Mey A, Lejour M, Malevez C. All in one:one stage closure of complete C.L.P.at 3 months. In: *Proceedings of the 7th international congress on cleft palate and related craniofacial anomalies*. Broadbeach, Australia; 1993.
7. De Mey A, Swennen G, Malevez C. All in one closure of unilateral C.L.P.: Results at 10 years. In: *Proceedings of the 9th international congress on cleft palate and related craniofacial anomalies.* Göteborg, Sweden; 2001.
8. Davies D, Fernandes D. The radical one-stage repair of cleft lip and palate in the neonate. In: *Proceedings of the 7th international congress on cleft palate and related craniofacial anomalies.* Broadbeach, Australia; 1993.
9. Standoli L, Cordaro L. One stage repair of unilateral complete clefts of lip and alveolus and palate. A six year follow-up. In: *Proceedings of the 7th international congress on cleft Palate and related craniofacial anomalies*. Broadbeach, Australia; 1993.
10. Honigmann K. One-stage closure of uni and bilateral cleft lip and palate. *Br J Oral Maxillofac Surg.* 1996;34: 214-219.
11. Honigmann K, Scherrer D. One-stage closure of a complete unilateral cleft lip and palate. In: *Proceedings of the 9th international congress on cleft palate and related craniofacial anomalies.* Göteborg, Sweden; 2001.
12. Bardach J, Roberts Dm, Yale R, Rosewall D, Mooney M. The influence of simultaneous cleft lip and palate repair on facial growth in rabbits. *Cleft Palate J*. 1980;17:309-318.
13. Bardach J, Kelly KM, Jakobsen JR. Simultaneous cleft lip and palate repair: an experimental study in beagles. *Plast Reconstr Surg*. 1988;82: 31-39.
14. Bardach J, Kelly KM, Salyer KE. A comparative study of facial growth following lip and palate repair performed in sequence and simultaneously:an experimental study in beagles. *Plast Reconstr Surg*. 1993;91: 1008-1016.
15. Jones CE, Chapman KL, Hardin-Jones MA. Speech development of children with cleft palate before and after palatal surgery. *Cleft Palate Craniofac J*. 2003;40:19-31.
16. Myklebust O, Abyholm FE. Speech results in CLP patients operated on with a Von Langenbeck palatal closure. *Scand J Plast Reconstr Surg Hand Surg*. 1989;23:71-74.
17. Malek R. Multidisciplinary management of cleft lip and palate in Paris, France. In: Bardach J, Morris HL, eds. *Multidisciplinary management of cleft lip and palate*. Saunders, Philadelphia; 1990:Sect 1:1.
18. Robinson PJ, Lodge S, Jones BM, Walker CC, Grant HR. The effect of palate repair on otitis media with effusion. *Plast Reconstr Surg.* 1992;89: 640-645.
19. Too-Chung MA. The assessment of middle ear function and hearing by tympanometry in children before and after early cleft palate repair. *Br J Plast Surg*. 1983;36:295-299.
20. Bluestone CD, Paradise JL, Beery QC, Wittel R. Certain effects of cleft palate repair on eustachian tube function. *Cleft Palate J*. 1972;9:183-193.

Professor A. De Mey
Department of Plastic Surgery
HUDERF
15, Avenue J.J. Crocq
B-1020 Brussels, Belgium
E-mail: albert.demey@chu-brugmann.be