Plastic and Reconstructive Surgery Advance Online Article

DOI: 10.1097/PRS.0000000000010331

Comparison of Internal and External Distraction in Frontofacial Monobloc Advancement:

A Three-Dimensional Quantification

Alexander J Rickart, BDS, MFDS¹, Lara S van de Lande, MD¹, Eimear O' Sullivan, MRes²,

Kevin Bloch, MD³, Eric Arnaud, MD³, Silvia Schievano MEng, PhD¹, Noor ul Owase Jeelani,

FRCS¹, Giovanna **Paternoster**, MD³, Roman **Khonsari**, MD, PhD³, David J **Dunaway**, FRCS¹.

1. UCL Great Ormond Street Institute of Child Health, London, UK & Craniofacial Unit, Great

Ormond Street Hospital for Children, London, UK.

2. Department of Computing, Imperial College London, London, UK.

3. Unité de Chirurgie Cranio-faciale, Service de Neurochirurgie, Centre de Référence Maladies

Rares Craniosténoses et Malformations Craniofaciales (CRANIOST), Hôpital Necker -

Enfants Malades, Assistance Publique – Hôpitaux de Paris; Faculté de Médecine, Université

de Paris; Paris, France.

Corresponding Author: Professor David J Dunaway, FRCS Craniofacial Unit Great Ormond

Street Hospital for Children Great Ormond Street WC1N 3JH London, UK

david.dunaway@gosh.nhs.uk

Financial Disclosure Statement:

This work has been funded by Great Ormond Street Hospital for Children Charity (Grant No.

12SG15), the Engineering and Physical Sciences Research Council (EP/N02124X/1), the

European Research Council (ERC-2017-StG-757923) and the National Institute for Health

Research Biomedical Research Centre Funding Scheme. The views expressed in this publication

are those of the authors and not necessarily those of the NHS, the National Institute for Health

Research or the Department of Health. The funders had no role in study design, collection, analysis and interpretation of the data, decision to publish, or preparation of the manuscript.

Conflict of Interest: The authors have no conflicts of interest to declare.

Short Running Head: Monobloc Distraction - Shape Analysis



Abstract

Introduction

Crouzon syndrome is characterised by complex craniosynostosis and midfacial hypoplasia. Where frontofacial monobloc advancement (FFMBA) is indicated, the method of distraction used to achieve advancement holds an element of equipoise. This two-centre retrospective cohort study quantifies the movements produced by internal or external distraction methods used for FFMBA. Using shape analysis, this study evaluates if the different distraction forces cause plastic deformity of the frontofacial segment, producing distinct morphological outcomes.

Methods

Patients with Crouzon syndrome who underwent FFMBA with internal distraction (*Necker*, *Hôpital Necker - Enfants Malades*, *Paris*) or external distraction (*GOSH*, *Great Ormond Street Hospital for Children*, *London*) were compared. DICOM files of pre- and post-operative CT-scans were converted to three-dimensional bone meshes and skeletal movements were assessed using non-rigid iterative closest point registration. Displacements were visualised using colour maps and statistical analysis of the vectors undertaken.

Results

51 patients met the strict inclusion criteria. 25 underwent FFMBA with external distraction and 26 with internal distraction. External distraction provides a preferential midfacial advancement whereas internal distractors produce a more positive movement at the lateral orbital rim. This confers good orbital protection but does not advance the central midface to the same extent. Vector analysis confirmed this to be statistically significant (p<0.01).

Conclusion

Morphological changes resulting from monobloc surgery differ depending on the distraction technique used. Although the relative merits of internal and external distraction still stand, it may be that external distraction is more suited to addressing the midfacial biconcavity seen in syndromic craniosynostosis.



Introduction

Crouzon syndrome is characterised by multiple craniofacial suture fusions leading to skull deformation and midfacial hypoplasia. Although there is a degree of phenotypic variance, functional problems such as raised intra-cranial pressure, exorbitism, craniovertebral junction anomalies, class III malocclusion, and sleep apnoea often arise.^{1,2}

Surgical management of these patients can be contentious and there is significant variation in

technical approaches globally. Distinct early fronto-orbital remodelling and a sequential Le Fort III procedure later in life were previously preferred, whereas single-stage procedures, frontofacial monobloc advancements (FFMBA), were deemed to carry an unacceptable complication rate.³⁻⁷ However, with the widespread integration of distraction into craniofacial surgery the risk to benefit ratio has become more favourable.⁸⁻¹¹

Where FFMBA is indicated, the method of distraction used to achieve advancement holds an element of equipoise. The choice between internal and external systems is largely down to training and expertise rather than clinical outcomes. External distractors hold the advantage of potentially shorter operating times, precise post-operative control of distraction vector and ease of removal. Conversely, they are bulky and susceptible to accidental knocks, pins can migrate transcranially and there is a psychological burden to wearing the device. 12-15 Internal distractors are more subtle and allow for a longer consolidation period which may, in turn, reduce relapse. 12 Conversely, the operation involves greater subperiosteal dissection, can be technically challenging and there is no option to alter the vector of distraction postoperatively without reoperation. 9,13-15 A recent systematic review comparing outcomes for internal and external midface distraction highlighted the indifference in advancement distance, reoperation and complication rates as well as relapse between both techniques. 12

One of the main unanswered questions regarding internal and external distraction techniques for midfacial advancement is the pattern of advancement obtained. It has been postulated that external distraction allows for a preferential midfacial advancement that could potentially provide a more favourable occlusal relationship and aesthetic outcome. ¹⁶ Internal distractors, owing to their zygomatico-temporal positioning in the midface, produce a positive movement at the lateral orbital rim, conferring good orbital protection. The downside of this is that the distraction forces applied laterally potentially do not provide sufficient drive to address the midfacial biconcavity seen in syndromic craniosynostosis. ^{12,16}

Using three-dimensional quantification, this study aims to compare internal and external distraction techniques for frontofacial monobloc distraction to assess the intricacies of the surgical movements and the subsequent patterns in relapse.

Materials and Methods

Dataset

Ethical approval was gained from the Joint Research and Development Office at the *Great Ormond Street Hospital for Children, London, UK* (GOSH) as well as *Hôpital Necker - Enfants Malades, Paris, France* (Necker). Review of the respective craniofacial databases was undertaken and patients with a clinical diagnosis of Crouzon syndrome who underwent FFMBA between 2004 and 2019 were identified. *FGFR2* mutations were confirmed in all patients. The two centres were directly compared, with internal distraction being undertaken at Necker and external distraction at GOSH. Patients were included if high-resolution computerized tomography (CT)-scans were available for 3D reconstruction at the pre-operative stage, within 90-days following completion of distraction and in the later post-operative phase. CT-scans with ≤1-millimetre slice thickness were required. Those with previous frontofacial surgery were

excluded, although those who had undergone prior posterior vault expansion were considered. Further descriptive demographics were also collected from electronic patient files.

Surgical Technique

Derived from the initial description of FFMBA, the surgical approach when undertaking internal or external distraction are largely similar and have been described previously. ⁷⁻⁹ Both centres utilised a frontal bone flap to facilitate osteotomies in the anterior skull base followed by subsequent use of a pericranial flap and fibrin glue in this region to help prevent cerebrospinal fluid leaks developing.

At GOSH, utilising the rigid external distraction (RED) frame (*KLS Martin, Jacksonville, Fla.*), the principal distraction vector is derived from the positioning of the frame. Polley plates placed at the glabellar region, bilaterally on the frontal bar and either side of the piriform rim connect to the RED frame using wires (*figure 1*). The distraction protocol used at GOSH involves a sevenday latency period followed by 0.5 mm twice daily movements until a satisfactory clinical endpoint is reached.

At Necker, two pairs of internal distraction devices (*KLS Martin, Jacksonville, Fla.*) are placed, with reinforcement of the fragile frontozygomatic sutures using miniplate fixation (*figure 2*). Intra-operative advancement of the distractors by 1-1.5cm is followed by a five-day latency period before daily distraction of 1mm in all four distractors. Distraction is again continued until a sufficient occlusal relationship and ocular protection is achieved, with the devices providing a maximum advancement of 25mm.

Data Processing

Data processing was performed by converting all DICOM data to three-dimensional (3D) bone meshes using Mimics InPrint 3.0 (*Materialise NV*, *Leuven*, *Belgium*). These meshes were then

cleaned using thresholding and isolation techniques to leave the cranium and midfacial bony structures, with the mandible excluded.

Quantification

To objectively quantify movements of the frontofacial segments following surgery, the preoperative meshes were compared with those immediately following completion of distraction or with the later post-operative meshes. A pre-existing workflow detailed by van de Lande *et al.* was used, starting with rigid alignment of meshes on the skull base, in this case using six landmarks. Non-rigid iterative closest point registration (NICP) was then guided by a further twenty-eight manually placed landmarks as specified in *Table 1*, utilising open-source software (*R3DS WRAP v3.4, Voronezh, Russia*). Colour maps were computed to visualise the global displacements across the entire facial skeleton. Point-to-point distances were derived from the landmarks to allow for statistical analysis of the vectors.

Statistical Analysis

Comparison of the mean lateral orbital advancement (landmarks 22-33) with the central landmarks of the nasion and the A-point provide an indication of the deformity of the frontofacial segment relative to where the distraction forces are acting. A ratio derived from isolated z-axis movements in these regions was analysed to describe movements at the upper and lower midface. Data were assessed for normality, with a two-sample t-test used to compare parametric data and Wilcoxon signed-rank test for non-parametric data. Means and standard deviation are used to describe parametric data throughout, with median and inter-quartile range used in non-parametric datasets. Patients were subdivided into four groups according to the age of intervention, early (0-3 years old), early childhood (3-9 years old), before secondary school (9-13 years old) and adolescence (13-19 years old). Comparisons were made between

operative meshes (within 90 days of completion of distraction). The proportional movements were then also calculated between the pre- and later post-operative meshes (6-18 months following surgery).

Reliability

All meshes were landmarked by AJR and intra-observer errors were calculated by choosing twenty landmarked meshes at random to be landmarked for a second time. An interval of at least two weeks between observations was taken to minimise memory bias and standard errors calculated. All statistical analysis was performed using Stata 16.0 (*StataCorp, College Station, Texas, USA*).

Results

Fifty-one patients met the strict inclusion criteria, of which 25 underwent external distraction at GOSH and 26 with internal distraction at Necker. Overall, 26 patients were male with 25 being female. Mean age at operation at GOSH was 9.3 years old, compared with 3.4 years old at Necker. Further details and timings of scans are summarised in *Tables 2* and *3*.

Reliability

All bar one landmark demonstrated an intra-observer error within one standard deviation, with all landmarks giving mean error of less than one millimetre. Outliers were noted in 24 out of 680 repeated landmarks (*figure 3*). One significant error of 5.6 mm was marked at the posterior angle of the zygomatic body in one patient (landmark 24) and on review, this was owing to an unfavourable fracture in this region that had distorted the anatomy significantly. The intra-observer errors are further detailed in *Supplemental Digital Content, Table 1*.

Colour Maps

Although there is relative heterogeneity of the dataset, the colour maps show distinct trends in each group. Representative samples are included for the age groups where direct comparison is possible (figure 4). For rigid external distraction, the universal colour of the frontofacial segment represents a movement more in keeping with a 'true' monobloc advancement. Several patients also demonstrate a slightly darker colour centrally, indicating a preferential central advancement. In comparison, the internal distraction group show a tendency towards preferential advancement at the lateral orbital rims when compared with the maxilla. This is particularly notable at the lower midface, although less pronounced in the upper midface around the supra-orbital rim and nasal bones. Interestingly, this pattern is not noted in all patients, with a small number of patients demonstrating homogenous advancement across the frontofacial segment. Both groups have instances where the frontal bone flap does not advance to the same degree as midfacial area, denoted by a lighter colour in this region.

Pattern of Advancement

A ratio derived from isolated z-axis (antero-posterior) movements both centrally and laterally was used to assess the relative movements at the upper and lower midface. A value of one correlates with a 'true' monobloc advancement, with a ratio greater than one indicating a preferential central advancement and less than one a more pronounced lateral advancement. The pattern of advancement shown in three dimensions by the colour maps is inferred numerically in *Table 4*. Mean proportional movements across the age groups consistently demonstrate higher values in the rigid external distraction group. The difference between the groups holds statistical significance in both the early and late post-operative phase. By comparing the pre-operative imaging with the available early and late post-operative meshes, it is possible to assess overall

patterns of advancement. We are unable to assess individual relapse in this context but can state that the differing morphology produced by the respective techniques persists into the late post-operative stage.

In conjunction with the colour maps, these findings highlight the degree of plastic deformity caused, corresponding to where the distraction forces are applied. For completeness and transparency, mean point-to-point movements at each landmark within 90-days of completion of distraction are included as *Supplemental Digital Content*, *Tables 2* and *3*.

Discussion

This study highlights key morphological differences produced by internal and external distraction techniques when undertaking monobloc advancement. Previously withheld beliefs that the Crouzon face represents a normal morphology in an abnormal position have fallen out of favour. Instead, research has improved our understanding that the Le Fort I subsegment is particularly retrusive compared to the rest of the midface and that this is coupled with orbitozygomatic disproportion. ²⁰⁻²³ This is highlighted by the frequent need for later orthognathic surgery in those who have previously undergone FFMBA or Le Fort III osteotomies and has led to some favouring split-level approaches to correcting the underlying discrepancy. ^{24,25} Bearing in mind the above, the morphological nuances of FFMBA are particularly relevant. This analysis demonstrates the potential additional benefit of external distraction in addressing the midfacial biconcavity seen in syndromic craniosynostosis. It is clear that both internal and external distraction give the means to address raised intra-cranial pressure, provide orbital protection and improve airway volume. 8,9,26 However, a preferential central advancement may infer some advantages in aesthetics, occlusal relationship and in total airway volume increase, although this is speculative. Nevertheless, utilisation of the RED frame in the very young can be

challenging. The weight of the frame necessitates careful support of the head and close monitoring by both medical staff and parents. The young skull is thinner more malleable, increasing the chances of transcranial pin migration. For the same reason, the RED frame can also cause biparietal narrowing.⁸ As ever, the benefits of both internal and external distraction must be balanced against their potential disadvantages, especially in infants.

The authors are keen to highlight that this study purely focusses on shape analysis and is not a case series intended to compare functional outcomes. Again, limitations include the contrasting distraction protocols and differences in age groups, with Necker having a tendency towards operating on younger patients. This is especially relevant as the young skull is more pliable, which may exaggerate the plastic deformity caused by the distraction forces. Additionally, differences in protocols between the two centres resulted in variation in the timings between scans and their subsequent meshes, which is a potential confounding factor. It is possible that the morphometric differences are, at least in part, owing to these key discrepancies. In contrast, the volume of data provided by NICP and dense anatomical correspondence is bolstered by excellent reliability in landmarking which adds strength to the findings. With all points giving mean error of less than one millimetre, this is well within the accepted standard for landmarks placed three-dimensions. ^{27,28}

FFMBA in the very young, where surgery is often a crisis intervention, poses a unique set of challenges. Disjunction at the fragile maxillo-zygomatic (MZ) and fronto-zygomatic (FZ) can occur intra-operatively during mobilisation of the frontofacial segment, or during distraction. This is offset by prophylactic plating at the FZ at Necker. However, in three patients who underwent internal distraction, disjunction at the MZ sutures occurred. Interruption of this structurally important buttress exacerbates the tendency towards preferential lateral

advancement. Previously this has been countered at Necker with the use of a transfacial pin (2.5 mm Kirschner wire) to stabilise the zygomatic body and avoid MZ disjunction. The initial database screening highlighted two patients otherwise meeting the inclusion criteria at the Necker who also had a transfacial pin placed. Unfortunately, owing to the profuse scatter caused by the pin, it was not possible to process these scans accurately for quantification due to distortion of several important landmarks. Using transfacial pins may solve part of the biomechanical issue related to internal distraction in very young patients, but this technique is associated with specific morbidity regarding damage to molar tooth germs; interfering with later orthodontic management and orthognathic surgery.²⁹

Sutural disjunction can also occur during external distraction, although not seen in this cohort at the MZ sutures. Owing to the positioning of central Polley plates, this is less of a setback, although lateral orbital projection may suffer. Instead, a pattern of advancement akin to a Le Fort III with zygomatic repositioning would be seen.^{21,30}

Considering previously published papers from both centres, functional outcomes appear favourable for both techniques and subjective clinical outcomes can be excellent in both instances (*figures 5&6*).^{8,9,23,26} As such, whether the differences highlighted by shape analysis of the two techniques of monobloc distraction translate into a true clinical difference is harder to discern. Comparison of craniofacial meshes to an age standardised 'normal' large-scale morphable model will soon be possible at both a hard and soft-tissue level and will provide considerable objective insight.¹⁹

Conclusion

This study demonstrates that the morphological changes resulting from monobloc surgery differ depending on the distraction technique used and that these changes persist into the late post-operative phase. External distraction provides a preferential midfacial advancement whereas internal distractors produce a more positive movement at the lateral orbital rim. Although the relative merits of internal and external distraction still stand, it may be that external distraction is more suited to addressing the midfacial biconcavity seen in syndromic craniosynostosis.

Acknowledgements

The authors would like to thank Professor Richard Hayward and Mr Robert Evans for their expert insight, help and guidance.

Reference List

- Tessier P. The definitive plastic surgical treatment of the severe facial deformities of craniofacial dysostosis. Crouzon's and Apert's diseases. *Plast Reconstr Surg*. 1971;48(5):419-442.
- Crouzon O. Une nouvelle famille atteinte de dysostose cranio-faciale héréditere. Bull Mem Soc Méd Hôp Paris. 1912;39:231-233.
- 3. Arnaud E, Di Rocco F. Faciocraniosynostosis: monobloc frontofacial osteotomy replacing the two-stage strategy? *Childs Nerv Syst.* 2012;28(9):1557-1564.
- Czerwinski M, Hopper RA, Gruss J, Fearon JA. Major morbidity and mortality rates in craniofacial surgery: an analysis of 8101 major procedures. *Plast Reconstr Surg*. 2010;126(1):181-186.
- 5. Dunaway DJ, Britto JA, Abela C, Evans RD, Jeelani NU. Complications of frontofacial advancement. *Childs Nerv Syst.* 2012;28(9):1571-1576.
- 6. Fearon JA, Whitaker LA. Complications with facial advancement: a comparison between the Le Fort III and monobloc advancements. *Plast Reconstr Surg.* 1993;91(6):990-995.
- 7. Ortiz-Monasterio F, del Campo AF, Carrillo A. Advancement of the orbits and the midface in one piece, combined with frontal repositioning, for the correction of Crouzon's deformities. *Plast Reconstr Surg.* 1978;61(4):507-516.
- 8. Ahmad F, Cobb AR, Mills C, Jones BM, Hayward RD, Dunaway DJ. Frontofacial monobloc distraction in the very young: a review of 12 consecutive cases. *Plast Reconstr Surg.* 2012;129(3):488e-497e.

- 9. Arnaud E, Marchac D, Renier D. Reduction of morbidity of the frontofacial monobloc advancement in children by the use of internal distraction. *Plast Reconstr Surg*. 2007;120(4):1009-1026.
- Bradley JP, Gabbay JS, Taub PJ, et al. Monobloc advancement by distraction osteogenesis decreases morbidity and relapse. *Plast Reconstr Surg.* 2006;118(7):1585-1597.
- 11. Hopper RA, Ettinger RE, Purnell CA, Dover MS, Pereira AR, Tuncbilek G. Thirty Years

 Later: What Has Craniofacial Distraction Osteogenesis Surgery Replaced? *Plast Reconstr*Surg. 2020;145(6):1073e-1088e.
- 12. Bertrand AA, Lipman KJ, Bradley JP, Reidhead J, Lee JC. Consolidation Time and Relapse: A Systematic Review of Outcomes in Internal versus External Midface Distraction for Syndromic Craniosynostosis. *Plast Reconstr Surg.* 2019;144(5):1125-1134.
- 13. Hindin DI, Muetterties CE, Lee JC, Kumar A, Kawamoto HK, Bradley JP. Internal Distraction Resulted in Improved Patient-Reported Outcomes for Midface Hypoplasia. J Craniofac Surg. 2018;29(1):139-143.
- Meling TR, Hogevold HE, Due-Tonnessen BJ, Skjelbred P. Midface distraction
 osteogenesis: internal vs. external devices. *Int J Oral Maxillofac Surg.* 2011;40(2):139-145.
- 15. Pelo S, Gasparini G, Di Petrillo A, Tamburrini G, Di Rocco C. Distraction osteogenesis in the surgical treatment of craniostenosis: a comparison of internal and external craniofacial distractor devices. *Childs Nerv Syst.* 2007;23(12):1447-1453.

- Goldstein JA, Paliga JT, Taylor JA, Bartlett SP. Complications in 54 frontofacial distraction procedures in patients with syndromic craniosynostosis. *J Craniofac Surg*. 2015;26(1):124-128.
- 17. Rickart AJ, van de Lande LS, O'Sullivan E, et al. Maxillary Changes Following Facial Bipartition-A Three-Dimensional Quantification. *The Journal of craniofacial surgery*. 2021.
- 18. van de Lande LS, O'Sullivan E, Knoops PG, et al. Local Soft Tissue and Bone Displacements Following Midfacial Bipartition Distraction in Apert Syndrome-Quantification Using a Semi-Automated Method. *The Journal of Craniofacial Surgery*. 2021.
- 19. Booth J, Roussos A, Ponniah A, Dunaway D, Zafeiriou S. Large scale 3d morphable models. *International Journal of Computer Vision*. 2018;126(2):233-254.
- 20. Britto JA, Evans RD, Hayward RD, Jones BM. From genotype to phenotype: the differential expression of FGF, FGFR, and TGFbeta genes characterizes human cranioskeletal development and reflects clinical presentation in FGFR syndromes. *Plastic and reconstructive surgery*. 2001;108(7):2026-2039; discussion 2040.
- 21. Hopper RA, Kapadia H, Morton T. Normalizing facial ratios in Apert syndrome patients with Le Fort II midface distraction and simultaneous zygomatic repositioning. *Plastic and reconstructive surgery*. 2013;132(1):129-140.
- 22. Khan S, Britto J, Evans R, Nischal K. Expression of FGFR-2 and FGFR-3 in the normal human fetal orbit. *British journal of ophthalmology*. 2005;89(12):1643-1645.

- 23. Visser R, Ruff CF, Angullia F, et al. Evaluating the efficacy of monobloc distraction in the Crouzon-Pfeiffer craniofacial deformity using geometric morphometrics. *Plastic and reconstructive surgery*. 2017;139(2):477e-487e.
- 24. Nout E, Koudstaal M, Wolvius E, Van der Wal K. Additional orthognathic surgery following Le Fort III and monobloc advancement. *International journal of oral and maxillofacial surgery*. 2011;40(7):679-684.
- 25. Posnick JC, Ruiz RL. The craniofacial dysostosis syndromes: current surgical thinking and future directions. *The Cleft palate-craniofacial journal*. 2000;37(5):1-24.
- 26. Witherow H, Dunaway D, Evans R, et al. Functional outcomes in monobloc advancement by distraction using the rigid external distractor device. *Plastic and reconstructive surgery*. 2008;121(4):1311-1322.
- 27. Hajeer MY, Ayoub AF, Millett DT, Bock M, Siebert JP. Three-dimensional imaging in orthognathic surgery: the clinical application of a new method. *The International journal of adult orthodontics and orthognathic surgery*. 2002;17(4):318-330.
- 28. Lagravère MO, Low C, Flores-Mir C, et al. Intraexaminer and interexaminer reliabilities of landmark identification on digitized lateral cephalograms and formatted 3-dimensional cone-beam computerized tomography images. *American journal of orthodontics and dentofacial orthopedics*. 2010;137(5):598-604.
- 29. Sicard L, Hounkpevi M, Tomat C, et al. Dental consequences of pterygomaxillary dysjunction during fronto-facial monobloc advancement with internal distraction for Crouzon syndrome. *Journal of Cranio-Maxillofacial Surgery*. 2018;46(9):1476-1479.

30. Hopper RA, Kapadia H, Susarla SM. Le Fort II distraction with zygomatic repositioning: a technique for differential correction of midface hypoplasia. *Journal of Oral and Maxillofacial Surgery*. 2018;76(9):2002. e2001-2002. e2014.



Figure Legends

Figure 1

Position of Polley plates. These three-hole titanium plates have a central fenestrated screw which allows for a distraction wire to pass to the RED frame.

Figure 2

Three-dimensional reconstruction demonstrating the position of one pair of internal distraction devices.

Figure 3

Intra-observer errors at each landmark. A point was considered an outlier if it was less than (Q1 - 1.5*IQR) or greater than (Q3 + 1.5*IQR).

Figure 4

Colour maps demonstrating movements produced across the facial skeleton following completion of distraction.

Figure 5

Clinical photographs demonstrating pre- and post-operative appearances following FFMBA undertaken with rigid external distraction.

Figure 6

Clinical photographs demonstrating pre- and post-operative appearances following FFMBA undertaken with internal distraction.

Table Legends

Table 1

List of landmarks used to guide both rigid and non-rigid iterative closest point registration.

Table 2

Summary Table – Great Ormond Street Hospital for Children – Rigid External Distraction.

Table 3

Summary Table – Hôpital Necker Enfants Malades – Internal Distraction.

Table 4

Relative Advancement at Lateral Compared to Central Landmarks.

Supplemental Digital Content Legends

Supplemental Digital Content, Table 1

Intra-Observer Errors by Landmark

Supplemental Digital Content, Table 1

Point-to-Point Distances Moved in Millimetres - Pre-Operative to Early Post-Operative Meshes

- Rigid External Distraction

Supplemental Digital Content, Table 3

Point-to-Point Distances Moved in Millimetres - Pre-Operative to Early Post-Operative Meshes

- Internal Distraction

Landmarks

Lanu	marks
0	Basion
1	Opisthion
2,3	Lateral margin foramen magnum
4,5 ⁺	Most anterior point of carotid canal
6	Nasion
7,8	Supra-orbital foramen
9,10	Orbitale
11	A-point
12,13	Antero-lateral margin of piriform rim
14,15	Malar prominence
16,17	Infra-orbital foramen
18,19	Most infero-lateral point of zygomatic arch
20,21	Antero-medial convergence between zygomatic
	process and squamous temporal bone
22,28	Posterior aspect frontozygomatic suture
23,29	Opposes Whitnall's tubercle in axial plane
24,30	Posterior angle formed by zygomatic body
25,31	Anterior aspect frontozygomatic suture
26,32	Whitnall's tubercle
27	Opposes 24 in axial plane
33	Opposes 30 in axial plane

^{*}where there are two landmark numbers, these correspond to

left and right pairs

+ landmarks 0-5 are skull base landmarks used for rigid alignment

GOSH - Rigid External Distraction

			Nur	nber of days betwee	n scan and	
				time of operation	on	
Age Group	Age	Gender	Pre-Op	Early Post-Op*	Late Post-Op	
	0y,11m	M	4	-	365	
	1y,3m	M	12	58	-	
Early	0y,7m	M	7	43	-	
	1y,5m	F	32	71	_	
	0y,8m	M	58	73		
	0y,3m	M	4	134		
	5y,11m	F	35	-	415	
Early	5y,3m	M	209	-	303	
Childhood	8y,5m	M	272	63	234	
	8y,10m	M	47	63	325	
	8y,11m	F	62	-/-	262	
	11y,9m	M	225	-	440	
	12y,8m	F	172	71	-	
Before	11y,7m	F	214	64	-	
Secondary	11y,1m	F	20	42	-	
School	11y,10m	M	151	63	451	
	11y,8m	F	130	-	156	
	10y,2m	F	123	-	479	
		1				
	15y,10m	F	55	62	-	
	17y,8m	F	61	-	182	
	15y,10m	F	110	-	261	
Adolescence	13y,3m	M	174	56	-	
	18y,9m	M	225	89	308	
	14y,11m	M	236	42	318	
	14y,1m	F	202	56	170	
Mean			114	66	311	

^{*} Early post-operative scan defined as within 90 days of completion of distraction

Necker - Internal Distraction

			Nun	nber of days betweer	scan and		
			time of operation				
Age Group	Age	Gender	Pre-Op	Early Pre-Op*	Late Post-Op		
	1y,8m	M	27	33	381		
	2y,2m	F	4	28	254		
	2y,8m	M	326	-	157		
	1y,6m	F	2	36	400		
	0y,11m	F	4	35	170		
	0y,3m	M	2	93	306		
Early	2y,4m	F	4	74	381		
	1y,11m	M	2	-	120		
	2y,2m	M	84	28	373		
	2y,1m	F	8	36	377		
	1y,7m	F	355	30	358		
	2y,7m	M	183	-	155		
	2y,1m	F	12	92	428		
	2y,1m	M	138	-	141		
				-			
	5y,3m	F	138	-	130		
	4y,4m	M	3	42	366		
	4y,9m	M	3	35	437		
	9y,0m	M	1	68	373		
Early	5y,3m	F	6	-	242		
Childhood	3y,3m	F	40	49	295		
	4y,7m	F	1	-	176		
	3y,10m	F	3	21	245		
	5y,10m	M	2	113	302		
	5y,7m	F	3	-	140		
	3y,5m	M	7	-	353		
	6y,11m	M	1	-	128		
Mean			52	51	277		

^{*}Early post-operative scan defined as within 90 days of completion of distraction

Relative Advancement at Lateral Compared to Central Landmarks

Following Completion of Distraction

Relative Advanceme nt*	Early Early Childho		hood	Before A Secondary School		Adole	Be		Difference Setween All age Groups		
		Mea	SD	Mea	SD	Mea	SD	Mea	SD	p-	95%
		n		n		n		n		value	CI
\mathbf{A}	\mathbf{GO}	1.01	0.2	0.93	0.31	1.00	0.04	0.97	0.27	< 0.01	0.88-
Point:Later	SH		3								1.10
al											
Landmarks											
	Nec	0.69	0.2	0.69	0.33	-	- 4	-	- /		0.55-
	ker		5			4					0.83
									~		
Nasion:	GO	1.39	0.1	1.25	0.50	1.09	0.06	1.06	0.08	< 0.00	1.07-
Lateral	SH		5							1	1.30
Landmarks	Nec	0.89	0.1	0.89	0.16	-	-	-	-		0.81-
	ker		4								0.97
				Late	r Post-(Operative	Period				
A	GO	1.07		1.17	0.32	1.10	1.09	0.95	1.00	< 0.05	0.90-
Point:Later	SH										1.24
al											
Landmarks	Nec	0.86	0.2	0.83	0.24	-	-	-	-		0.74-
	ker		8								0.95
		Medi	IQ	Med	IQ	Med	IQR	Med	IQR		Exact
		an	R	ian	R	ian		ian			Confid
Nasion:	GO	1.77	47	1.31	0.11	1.17	0.06	1.16	0.11		ence Level
Lateral	SH	1.//	_	1.31	0.11	1.1/	0.00	1.10	0.11		Level
Landmarks	Nec	1.06	0.1	1.11	0.14	_		_	_	< 0.00	0.84
Landmar K5	ker	1.00	7	1.11	0.17					1	0.07

^{*} A ratio derived from isolated z-axis (antero-posterior) movements both centrally and laterally was used to assess the relative movements at the upper and lower midface. A value of one correlates with a 'true' monobloc advancement, with a ratio greater than one indicating a preferential central advancement and less than one a more pronounced lateral advancement.

Figure 1

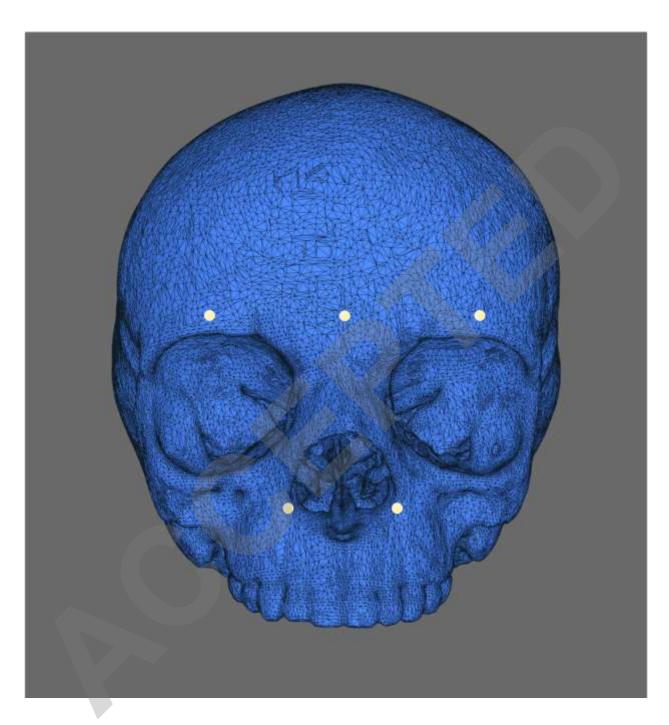


Figure 2



Figure 3

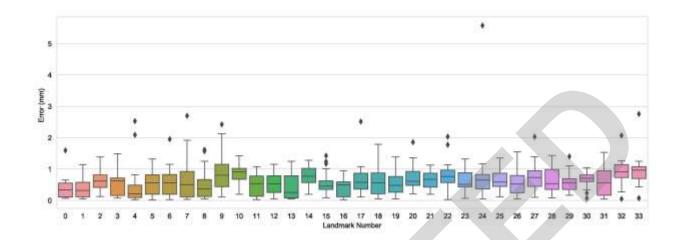




Figure 4

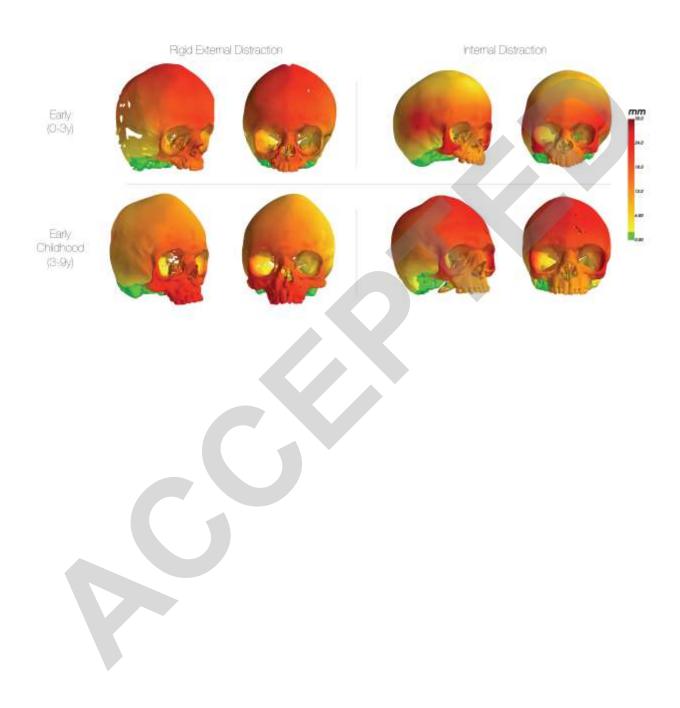


Figure 5











Figure 6



Landmark	Mean Difference	Standard
	(mm)	Deviation
0	0.39	0.35
1	0.39	0.31
2	0.61	0.31
3	0.54	0.37
4	0.48	0.67
5	0.54	0.38
6	0.59	0.49
7	0.65	0.70
8	0.49	0.48
9	0.88	0.61
10	0.85	0.36
11	0.50	0.34
12	0.54	0.35
13	0.44	0.41
14	0.77	0.31
15	0.57	0.37
16	0.43	0.30
17	0.66	0.52
18	0.61	0.43
19	0.54	0.36
20	0.72	0.40
21	0.65	0.27
22	0.81	0.48
23	0.61	0.33
24	0.83	1.16
25	0.63	0.33
26	0.59	0.45
27	0.78	0.46
28	0.63	0.37
29	0.59	0.29
30	0.67	0.25
31	0.64	0.49
32	0.93	0.41
33	0.92	0.55

SDC 1

GOSH – Rigid External Distraction

Point-to-Point Distances Moved in Millimetres - Pre-Operative to Early Post-Operative Meshes

Landmark	Early (n=5)		Early Childhood (n=2)			Before Secondary School (n=4)		Adolescence (n=5)	
	Mean	SD	Mean	SD	Mean	SD	Mean	SD	
6	16.20	3.08	16.43	6.85	14.68	2.17	13.65	2.32	
7	17.61	2.56	16.50	4.35	13.92	2.57	13.34	2.25	
8	16.91	3.32	15.21	6.85	13.33	1.53	14.23	4.16	
9	12.50	3.30	14.51	1.24	15.85	3.49	14.06	4.51	
10	10.72	1.84	12.08	1.47	13.82	0.73	14.33	5.13	
11	11.79	3.13	14.05	1.67	13.74	2.40	12.93	4.99	
12	13.78	4.92	14.49	0.02	14.86	2.33	12.88	3.94	
13	12.73	3.93	13.61	0.05	13.31	2.09	13.07	3.99	
14	12.74	3.50	14.37	4.13	15.26	3.39	13.53	4.18	
15	10.54	4.26	11.40	2.02	13.62	1.24	12.35	4.23	
16	12.99	3.59	14.09	1.42	13.58	3.10	13.27	4.17	
17	12.26	3.23	12.66	1.14	13.77	1.23	12.42	4.97	
22	14.57	4.76	15.41	1.86	14.90	4.47	13.03	2.98	
23	11.82	4.38	15.75	1.24	15.23	3.60	13.68	2.61	
24	12.95	2.50	15.18	1.96	14.37	5.16	12.02	2.71	
25	14.27	4.62	15.31	0.98	14.16	3.55	13.24	2.53	
26	12.21	3.73	14.95	0.27	14.41	3.64	13.37	2.58	
27	11.52	2.18	14.11	1.43	14.27	3.97	12.72	4.53	
28	12.91	3.44	11.95	2.00	12.66	0.96	13.52	2.48	
29	10.26	3.53	11.51	1.36	12.51	1.29	13.70	3.28	
30	8.45	2.51	11.44	1.32	13.17	1.16	13.07	3.86	
31	12.68	4.31	11.98	1.23	12.21	0.85	13.20	2.96	
32	11.01	2.16	11.68	0.55	12.50	0.86	13.09	3.17	
33	10.64	2.25	11.80	0.84	12.29	1.74	13.24	4.81	

Necker – Internal Distraction

Point-to-Point Distances Moved in Millimetres Pre-Operative to Early Post-Operative Meshes

	Early (n=10)		Early Childl	hood (n=6)
Landmark	Mean	SD	Mean	SD
6	16.220	3.269	15.745	5.710
7	16.604	3.473	14.866	4.983
8	16.233	2.933	15.163	5.246
9	23.232	20.105	14.986	6.521
10	22.087	18.714	15.233	6.227
11	12.942	3.313	13.446	7.661
12	13.346	2.756	13.717	6.877
13	13.786	2.540	13.946	6.870
14	16.391	4.094	14.739	7.204
15	15.697	3.032	14.744	7.515
16	13.603	2.576	13.987	6.660
17	13.508	2.825	13.950	7.101
22	18.381	2.590	18.177	4.299
23	19.053	3.320	18.159	4.980
24	18.839	4.256	17.397	5.472
25	17.788	2.051	17.875	3.566
26	16.869	2.329	16.959	5.073
27	16.120	2.669	15.876	5.685
28	18.544	2.461	16.906	5.541
29	16.829	2.046	16.716	5.328
30	18.928	3.361	17.302	4.678
31	18.008	2.636	17.387	4.949
32	16.725	2.550	16.197	5.747
33	16.125	2.195	15.505	6.190