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Review

Distraction osteogenesis in the surgical management of syndromic craniosynostosis: a comprehensive review of published papers

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Abstract

Our aim was to summarise current published evidence about the prognosis of various techniques of craniofacial distraction osteogenesis, particularly its indications, protocols, and complications. Published papers were acquired from online sources using the keywords "distraction osteogenesis", "Le Fort III", "monobloc", and "syndromic craniosynostosis" in combination with other keywords, such as "craniofacial deformity" and "midface". The search was confined to publications in English, and we followed the guidelines of the PRISMA statement. We found that deformity of the skull resulted mainly from Crouzon syndrome. Recently craniofacial distraction has been achieved by monobloc distraction osteogenesis using an external distraction device during childhood, while Le Fort III distraction osteogenesis was used in maturity. Craniofacial distraction was indicated primarily to correct increased intracranial pressure, exorbitism, and obstructive sleep apnoea in childhood, while midface hypoplasia was the main indication in maturity. Overall the most commonly reported complications were minor inflammatory reactions around the pins, and anticlockwise rotation when using external distraction systems. The mean amount of bony advancement was 12.3 mm for an external device, 18.6 mm for an internal device and 18.7 mm when both external and internal devices were used. Treatment by craniofacial distraction must be validated by long-term studies as there adequate data are lacking, particularly about structural relapse and the assessment of function.

Keywords: Syndromic craniosynostosis; distraction osteogenesis; midface; treatment strategies; distraction devices

Introduction

Craniosynostosis is premature fusion of the craniofacial sutures that causes disturbance of craniofacial growth. The function of cranial sutures is to allow deformation of the skull during passage through the birth canal and growth of the calvarium. Craniosynostosis can be an isolated event, which results in non-syndromic craniosynostosis, or it can happen in conjunction with other anomalies in distinct patterns that make up clinically-recognised syndromes. Despite there being a broad range, the most commonly identified syndromes of which craniosynostosis is a part include Crouzon, Apert, Pfeiffer, Muenke, and Saethre-Chotzen syndromes. The condition may develop when a gene mutates, or it may be genetically inherited.

Patients with syndromic craniosynostosis have impaired growth of the skull and facial bones that makes it hard to achieve lasting correction of the appearance and to maintain adequate space in the skull for the growing brain and other vital structures. Affected patients often have serious problems with breathing, hearing, speech, and mastication, and may also be born with anomalies of the limbs that

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give rise to additional functional and operative challenges. Functional assessment involves the evaluation of patients' problems that are associated with structural anomalies in syndromic craniosynostosis, and the main functional issues are caused by raised intracranial pressure as a result of restricted intracranial volume secondary to early fusion of the cranial sutures. Visual functions that can be affected include the optic nerve, which may be injured by the raised intracranial pressure; and inability to close the eyelids as a result of severe exophthalmos, which requires orbital protection. The patient may also have breathing difficulties secondary to the narrow nasopharyngeal airway caused by a severely hypoplastic midface. The most serious problem is the rise in intracranial pressure, which is detrimental to the brain. By using an epidural monitor in children with craniosynostosis, workers have shown that 47% of the children with "multiple-suture fusion" have increased intracranial pressure.[1] This renders those with syndromic craniosynostosis more difficult to care for, and requires a multidisciplinary team to address their needs effectively.

Those with additional midface retrusion, which is one problem of syndromic craniosynostosis, may present with raised intracranial pressure, exophthalmos, malocclusion, respiratory difficulty, and developmental delay.[2] Several techniques that have been developed (including surgical intervention in infancy, the advent of computed tomography, the introduction of rigid (and later resorbable) plating systems, and advances in distraction osteogenesis) now help to solve this problem. The Le Fort III midface advancement technique was introduced by Sir Harold Gillies in 1949 and later refined and popularised by Tessier.[3,4] More recently the development of the application of distraction osteogenesis to the craniofacial skeleton has raised the possibility of a new treatment (Le Fort III distraction osteogenesis), which does not involve bone grafting and has few complications. There are also lower rates of relapse and infection that had previously been caused by the massive frontonasal dead space behind the advancement segment.[5] Le Fort III has been replaced with monobloc or Le Fort III with facial bipartition, depending on the need for cranial expansion.[6]

Distraction osteogenesis was introduced into craniomaxillofacial surgery in 1992 when McCarthy et al successfully used it to lengthen a human mandible.[7] It involves an osteotomy followed by rigid fixation to lengthen the bone, muscles, and soft tissues, and it results in a serious degree of correction over time.[8] After a brief period of latency, the segments of bone are gradually detached at a specific rate and time. After the bone has regenerated, a period of consolidation allows for the mineralisation of the lengthened bone. This is a valuable advance in the treatment of children with mandibular deficiencies, and provides effective correction of micrognathia with associated expansion of soft tissue.[9] Technically, craniosynostosis can be treated more successfully in this way with different distraction devices.[10,11]

Early craniofacial correction (within the first years of life) depends on the neurosurgical and functional indications. During the first six years, reconstruction is focussed on cranial decompression and reshaping procedures associated with it. Reconstruction of craniofacial deformities is recommended after the age of 6 years, and will achieve stable adult dimensions in the cranio-orbitozygomatic regions. Finally, correction of occlusal relations can be achieved after skeletal maturity (Table 1).[12]

Table 1
Timing of distraction of osteogenesis.

Age	Distraction osteogenesis
Less than 6 months/6–12 months	Posterior distraction of the cranial vault (particularly in case of posterior stenosis of the skull)
4–8 or 9–12 years	Le Fort III advancements can combine with distraction osteogenesis when a lack of bone and soft tissue forces preclude a single stage
4–12 years	Monobloc frontofacial advancement
14–18 years	May be needed, may be not

The treatment of syndromic craniosynostosis is still evolving, and there is an increasing number of reports about distraction osteogenesis and the types of device being used.[12] Our aim was to compare the success of this treatment based on published reports (with particular emphasis on clinical indications, surgical techniques, advantages and disadvantages, types of distraction device, and complications). Such a review may alter the management of patients.

Methods

Objective

We have tried to synthesise evidence from current research to assess the effectiveness and prognosis of the different techniques of distraction osteogenesis that have been used in patients with syndromic craniosynostosis, with emphasis on their indications, protocols, and complications. Our question was: what was the effect of different distraction protocols and distraction devices in such patients who required craniofacial distraction, and what were their adverse effects, stability, and complications?

Selection criteria

We included studies of all patients of any age with syndromic craniosynostosis who had Le Fort III, monobloc or bipartition distraction and required one operation or more.

Exclusion criteria

We excluded patients who had been treated conventionally, those with non-syndromic craniosynostosis, and those who had been treated conservatively. Excluded studies were reviews, abstracts, debates between authors, summary

papers, and studies with incomplete data or that involved animals.

Outcome

Review of the data will provide important information about the effect of the technique of distraction osteogenesis on various outcomes: the indication for a particular device; whether a different protocol produces a different result; the mean amount of advancement of bone; and the stability and complications of the procedure.

Protocol of the search

Papers were sought from five resources online (PubMed, ISI Web of Science, Science Direct, Journals@Ovid, and SciFinder), which were searched for all relevant papers published before 2016. The Boolean terms used for the electronic database search were: (syndromic craniosynostosis) AND (distraction osteogenesis) AND (management OR treatment) AND (Le Fort III OR fronto-orbital distraction OR facial bipartition OR monobloc). Criteria for eligibility and exclusion of studies were decided beforehand. We manually searched bibliographies of papers extracted to identify relevant references that we might have missed. The search did not limit inclusion of a study by the year of publication, but was limited to those on humans that were published in English. The process of selection was repeated by two independent reviewers to ensure its transparency and to confirm that the inclusion process worked. The senior author served as a judge, and assessed the full texts of potentially eligible studies for eligibility criteria, type of study, level of evidence, demographic information, methods, and reported outcomes. Disagreements were resolved by discussion and additional review. The search of publications followed the guidelines of the PRISMA statement (Fig. 1).[13]

Assessment of risk of bias (quality)

We made a systematic assessment of the risk of bias in the studies included by analysing the selection bias (for example, the division of participants into exposed and not exposed groups, adequate details of exclusions, and incomplete outcome data in the published report, or selective reporting of outcomes that might affect the results of the study); information bias (for example, differences in the quality of collection and extent of information gathered from various groups, or differences in the numbers lost to follow-up between the groups); and potential sources of misclassification. Two reviewers made the risk-of-bias assessment independently, and disagreements were resolved by discussion (with the involvement of a third review author when necessary).

Table 2
Patients' age at the time of distraction osteogenesis for syndromic craniosynostosis in 298 patients.

Age range	Le Fort III (n = 108)	Monobloc (n = 167)	Bipartition distraction (n = 23)
<6 months	1	5	–
6–11 months	3	2	–
1–3 years, 9 months	11	25	2
4–8 years, 9 months	11	21	2
9–13 years, 9 months	2	20	7
14–18 years	6	6	10
>18 years	1	8	2
Not mentioned	73	80	

Collection and analysis of data

As there were no controlled studies we did not do a meta-analysis. Descriptive statistics were used to report the data. A total of 24 papers were identified. Only full-text papers were retrieved, as abstracts did not provide enough information for detailed review. Data were extracted and summarised. Excel worksheets were designed to list the specific variables for analysis, which included author, year of publication, number of patients, sex, age, diagnosis, indication, and surgical protocol. Data about the technique of distraction osteogenesis included: time between primary reconstruction and the placement of the distractor; details of how the amount of bone was gained; the distraction technique (latency period, active distraction period, the rate of distraction, and consolidation period) and types of distraction devices. These were recorded and compared. Operative and postoperative complications were collected sequentially.

Results

A total of 298 cases treated by Le Fort III, monobloc, or bipartition distraction were reported in 24 papers. The sex and age ranges are summarised in Tables 2 and 3. Most patients had Crouzon syndrome, followed by Apert and Pfeiffer syndromes (Table 4).

Syndromic craniosynostosis is associated with multisutural synostosis and severe midfacial retrusion, which may cause serious functional problems (including obstructive sleep apnoea, feeding difficulties, exorbitism, and raised intracranial pressure). The indications for Le Fort III, monobloc, or bipartition distraction are summarised in Table 5. The associated functional and cosmetic problems were usually treated by Le Fort III (with or without fronto-orbital advancement), monobloc, or bipartition distraction in sequence. The number of cases treated by the different surgical approaches is summarised in Table 6.

Table 3
Profiles of available papers about Le Fort III, monobloc, and bipartition reconstructions.

First author and reference	Year	No. of patients	Mean Age (years)	Sex	Diagnosis	Type of study
Cohen[19]	2001	1	4	F	Crouzon syndrome (n = 1)	Case report
Mavili[20]	2003	1	12	M	Crouzon syndrome (n = 1)	Case report
Satoh[3]	2006	20	9.6	9 M/11 F	Crouzon syndrome (n = 11), Apert syndrome (n = 7), Pfeiffer syndrome (n = 1), Antley-Bixle syndrome (n = 1)	Retrospective study
Jensen[35]	2007	10	5.1	*N/A	Crouzon (n = 6), Pfeiffer (n = 2), Apert syndromes (n = 2)	Cohort study
Witherow[24]	2008a	20	7.8	N/A	Crouzon syndrome (n = 10), Apert syndrome (n = 4), or Pfeiffer syndrome (n = 6)	Retrospective study
Ponniah[25]	2007	10	9	N/A	Crouzon syndrome (n = 7), Apert syndrome (n = 3)	Cohort study
Arnaud[32]	2007	36	5.2	N/A	Crouzon syndrome (n = 19), Pfeiffer syndrome (n = 8), Apert syndrome (n = 8), and Shprintzen-Goldberg syndrome (n = 1)	Cohort study
Witherow[28]	2008b	1	4 months	F	Pfeiffer syndrome (n = 1)	Case report
Xu[14]	2009	11	5.7	N/A	Crouzon syndrome (n = 10), Apert syndrome (n = 1)	Cohort study
Oyama[26]	2009	40	4.5	N/A	Crouzon (n = 22), Apert syndrome (n = 11), Pfeiffer syndrome (n = 7)	Retrospective study
Flores[15]	2009	20	5.7	10 M/10 F	N/A	Retrospective study
Shetye[21]	2010	20	5.7	10 M/10 F	Crouzon syndrome (n = 9), Apert syndrome (n = 7), Pfeiffer syndrome (n = 4)	Retrospective study
Ko[29]	2011	5	9.7	3 M/2 F	Crouzon syndrome (n = 3), Apert syndrome (n = 1), Pfeiffer syndrome (n = 1)	Cohort study
Ahmad[30]	2011	12	18	N/A	Crouzon syndrome (n = 6), Apert syndrome (n = 1), Pfeiffer syndrome (n = 5)	Retrospective study
Shetye[36]	2012	20	5.7	10 M/10 F	Crouzon syndrome (n = 9), Apert syndrome (n = 7), Pfeiffer syndrome (n = 4)	Cohort study
Bender[31]	2012	7	10.7	4 M/3 F	Crouzon syndrome (n = 2), Apert syndrome (n = 4), Pfeiffer syndrome (n = 1)	Retrospective study
Kobayashi[37]	2012	2	7	1 M/1 F	Crouzon syndrome (n = 2)	Cohort study
Greig[34]	2012	17 3	15.2 2.6	10 M/10 F	Apert syndrome (n = 19), Pfeiffer syndrome (n = 1)	Cohort study
Mitsukawa[16]	2013	11	2.5	6 M/5 F	Crouzon syndrome (n = 4), Apert syndrome (n = 4), Pfeiffer syndrome (n = 3)	Case reports
Hariri[27]	2015a	2	3.5	1 M/1 F	Crouzon syndrome (n = 2)	Case reports
Hariri[33]	2015b	1	8 months	M	Crouzon syndrome (n = 1)	Case reports
Bouw[22]	2015	17	14.7	8 M/9 F	Crouzon syndrome (n = 8), Apert syndrome (n = 7), Pfeiffer syndrome (n = 2)	Retrospective study
Dobbs[17]	2014	1	4 months	F	Crouzon syndrome (n = 1)	Case report
Wery[18]	2015	10	14	3 M/7 F	Crouzon syndrome (n = 5), Apert syndrome (n = 3), Pfeiffer syndrome (n = 2)	Retrospective study

* N/A-not available.

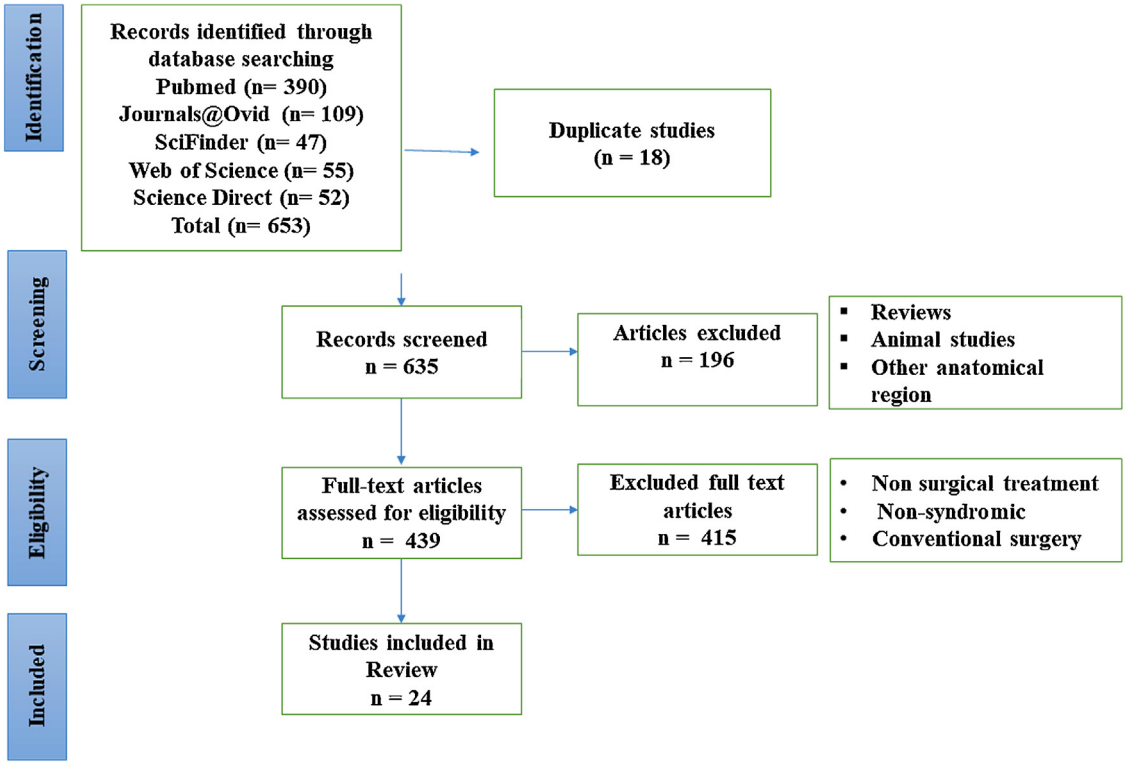

Fig. 1. Algorithm showing the plan of the investigation according to the PRISMA statement.[13]

Table 4
Reported types of syndromic craniosynostosis and number of patients with each.

Syndromic craniosynostosis	Number
Crouzon's syndrome	136
Apert syndrome	91
Pfeiffer syndrome	49
Antley-Bixler syndrome	1
Shprintzen-Goldberg syndrome	1
Not mentioned	20
Total	298

Indications for different devices

Even though the external device was used more than the internal one, the evidence remains the same as the technique allows superior midfacial advancement and opens up the narrow nasopharyngeal airway in children with syndromic craniosynostosis. The system developed by KLS Martin (Tuttlingen, Germany) was the most common system used for distraction osteogenesis among the 24 studies (Table 7).

Table 6
Types of the surgical approach and distraction systems.

Surgical approach and study	No. of patients
Le Fort III distraction[14–16,18–22,35,36]	134
Le Fort III DO + FOD[14,16]	9
Le Fort III DO + staged calvarial expansion remodelling[17]	1
Monobloc distraction[23–33]	131
Facial bipartition distraction[25,34]	23

Table 5
Clinical indications for Le Fort III, monobloc face reconstruction, and facial bipartition distraction.

Clinical indications		
Le Fort III distraction	Monobloc Le Fort III DO	Facial bipartition distraction
• Obstructive sleep apnoea[14–18] • Exorbitism[17–20] • Midfacial hypoplasia[19–22] • Angle Class III malocclusion[20,22]	• Midfacial hypoplasia[23–27] • Raised intracranial pressure[28–31] • Obstructive sleep apnoea[27,28,31,32] • Exorbitism[29,31–33] • Class III malocclusion[31]	• Midface central advancement in Apert syndrome patients[25,34]

Table 7
Types of distraction systems described.

Distraction system and device
External:
RED II device (KLS Martin, Tuttlingen, Germany) and (KLS-Martin, Jacksonville, Fla.)[15,20–22,24,25,28–30,34–36]
Cibei Medical Treatment Appliance (Ningbo, Zhejiang China)[14]
Blue device (Lorenz, Jacksonville, FL, USA)[16]
Atransfacial pinning system (Medical U&A Inc., Osaka, Japan)[16]
External Midface Distractor (Synthes, Zur-ich, Switzerland)[22]
External frame Laminated bone grafts (Modification for prevention of pin displacement)[17]
Internal:
Internal device (Keisei, Tokyo, Japan)[23]
Internal device (W. Lorenz, Jacksonville, FL)[16,23]
Internal device (Synthes Maxillofacial, Paoli,Pa.)[35]
Marchac devices (Martin Medizin, Tuttlingen, Germany)[22]
Arnaud-Marchac device (KLS Martin, Tuttlingen, Germany)[31,32]
2 pairs Marchac devices + Riediger Distractor (Martin Medizin, Tuttlingen, Germany)[26]
Combination:
EMDS (Synthes, Oberdorf, Switzerland)[33]
EMDS [(Synthes, Oberdorf, Switzerland) + a customized headgear][27]
RED System (Martin LP, Jacksonville, FL) + Internal device protype (interlocking with the external device)[37]
Internal Marchac/Arnaud Distraction System (KLS Martin, Tuttlingen, Germany) + RED II (KLS Martin, Tuttlingen, Germany)[31]
Resorbable
Modular Internal Distraction (MID) (MacroPore, San Diego, CA)[19]

Either internal or external devices or both were used in craniofacial distraction. External ones were used in 191 cases: 107 of which were treated by Le Fort III, eight by Le Fort III distraction osteogenesis with fronto-orbital distraction, one by Le Fort III distraction osteogenesis with staged calvarial expansion remodelling, 23 by bipartition, and the other 52 by monobloc. Internal distraction devices were used in 101 cases: 24 by Le Fort III, one by Le Fort III distraction osteogenesis with fronto-orbital distraction, and 76 by monobloc. A combination of external and internal devices was used to treat five cases: two by Le Fort III, and three by monobloc. A resorbable distraction device was used in one case treated by Le Fort III. Most of the papers used the same surgical techniques, except for modifications in some cases to avoid complications. The modifications in technique are summarised in Table 8.

We found that external devices could also be indicated for patients less than 2 years of age if there was consideration of weak zygoma-maxillary union to prevent unnecessary zygomaticomaxillary disjunction, and the risk of fracture of distracted bone during removal.[16,29]

Do different protocols produce different results?

Most of the studies applied similar protocols for distraction, despite the different techniques. Different strategies were used for the latency period, from immediate activation up to seven days. Distraction rates varied from 0.5–1.5 mm/day, and only one study (that used resorbable distraction devices) reported a distraction rate of 2 mm/day.[19] Our analysis also showed that fewer complications were reported when the latency phase was 5–7 days and a distraction rate/day of 1 mm was used, particularly in the prevention of leakage of cerebrospinal fluid.[14,15,21–23,25,27–29,32,33,35,36]

These protocol values were reduced to minimal latency period, can be modified to rapid distraction and had shorter consolidation phase when bone paste was used at the distraction site or resorbable distraction device was indicated. The consolidation period ranged significantly from 1.5 to 6 months, but in general, most studies demonstrated longer period of consolidation when internal distractor systems were used, or massive structural movement was needed.

Mean bony advancement

The mean advancement ranged from 12–27 mm; that for the external device was 12.3 mm,[16,20,21,22,24,25,28,29,34] for the internal device 18.6 mm,[16,22,23,32] and when both external and internal devices were used it was 18.7 mm.[27,33,37]

Stability and complications

Most studies reported their follow-up period, which ranged from 5–132 months. The amount of observation time differed among studies (less than 6 months,[14,19] 6–12 months[15,16,20,21,22–24,35,36] 13–24 months,[24,33,34] and than 24 months.[16,17,23,24,26,29,30,32,34]) A long-term follow-up study of relapse (25 months) with Pfeiffer syndrome only (5/7 patients), reported the poorest reossification of both the cranium and the zygomatic arch.[26] Relapse was noted predominantly at the occlusal level (6/35) and more often in Pfeiffer syndrome, within the mean of 30 months after frontofacial monobloc advancement and quadruple internal distraction (latency of five days and distraction rate of

Table 8
Surgical techniques used in distraction osteogenesis.

First author and reference	Surgical approach	Modification
Cohen[19]	Le Fort III midfacial osteogenesis	Three stages; 1) operation, 2) removal of distraction devices and insertion of tissue expanders., and 3) fronto-orbital remodelling and cranial reconstruction with autogenous bone grafts
Mavili[20]	Le Fort III midfacial osteogenesis	An osteotomy line 1.5 cm inferior to the original lateral wall osteotomy was added to the Le Fort III osteotomy line.
Satoh[23]	Le Fort III midfacial osteogenesis	A newly developed parallel bar adds to the distraction device that attached to the zygoma to obtain parallel setting of the bilateral distraction devices
Jensen[35]	A classic Le Fort III midfacial distraction	No modification
Witherow[24]	Monobloc osteotomy	Passed a long pericranial flap between the osteotomy cuts and sutured the distal part of it through drill holes in the cranial base.
Ponniah[25]	Monobloc distraction	Three with Apert syndrome underwent facial bipartition distraction osteogenesis
Arnaud[32]	Frontofacial monobloc with complete osteotomy	No modification
Witherow[28]	Monobloc	A long-vascularised flap of frontal pericranium was passed to the posterior edge of the skull base. Frontal bone was subdivided "armadillo-fashion" and the components reattached to each other. Distraction frame fixed on to a rectangular sheet of titanium mesh.
Xu[14]	Le Fort III distraction	The procedure included a simultaneous one-stage fronto-orbital advancement
Oyama[26]	Frontofacial monobloc with complete osteotomy	Fibrin glue (bone paste) was added and distraction devices were implanted
Flores[15]	Le Fort III midfacial	No modification
Shetye[21]	Le Fort III	Adding an acrylic and wire splint at maxillary teeth with dental cement
Ko[29]	Monobloc frontofacial	Maxillary arch advancement was conducted to the extension eyelets of the intraoral appliance with distraction screws.
Ahmad[30]	Monobloc frontofacial osteotomies DO	No modification
Shetye[36]	Le Fort III midfacial DO	No modification
Bender[31]	Monobloc DO	No modification
Kobayashi[37]	Le Fort III midfacial DO	Using both external and internal distraction device
Greig[34]	Bipartition distraction	The two bipartition segments wired together in the midline and plates were screwed onto bone and wires were attached to them to avoid the risk of dental morbidity in the pyriform region
Mitsukawa[16]	Le Fort III midfacial DO	No modification
Dobbs[17]	Le Fort III midfacial DO	Mid-face sutural distraction combined with a preliminary sagittal strip craniectomy
Hariri[27]	Monobloc Le Fort III distraction	Using both external and internal distraction device
Hariri[33]	Monobloc Le Fort III distraction	Using both external and internal distraction device + customized headgear
Bouw[22]	Le Fort III midfacial DO	No modification
Wery[18]	Le Fort III DO where the classic osteotomies	No modification

1 mm/day). Other studies reported stability without appreciable relapse within the reported follow up period (Table 9).[17,29]

Most of the studies reported complications associated with the distraction osteogenesis (Table 10 Supplemental data, online only), the most common of which were pin-site infections (n = 7)[14,17,31,34] and counterclockwise rotational movement when using external distraction systems (n = 35).[21,22,37] In addition, there were leaks of cerebrospinal fluid (n = 20) when the minimal latency period (immediate activation and less than 24 hours latency phase), and rapid distraction (2–5 mm) were practised.[28,30,31,34]

Discussion

We have described different protocols for distraction osteogenesis and complications associated with the technique, as well as highlighting the modifications used to avoid the reported complications. We have also reported on the devices used for distraction and their advantages and disadvantages. We have provided objective data to distinguish the technical variables of craniofacial distraction using different devices including the patient's age, optimal vector planning, complications, advantages, and disadvantages, and put forward suggestions

Table 9
Summary of papers available about Le Fort III, monobloc, and bipartition reconstructions.

First author/reference	Latency (day)	Rate (mm/day)	Activation (days)	Mean bone extension (mm)	Consolidation (months)	Follow up (months)
Cohen[19]	4	2	Not mentioned	20	Not mentioned	5
Mavili[20]	Immediately	1	10	14	1.5	7
Satoh[23]	4–5	1	Not mentioned	17	4–6	12–54
Jensen[35]	7	1	7–15	Not mentioned	2	6–12
Witherow[24]	1 and then 7 days	1.5 and 1	Not mentioned	16.4	1.5	6–48
Ponniah[25]	7	1		Monobloc:17.1 (central), 12.1 (lateral) Bipartition: 18.4 (central), 7.0 (lateral)	1.5	Not mentioned
Arnaud[32]	5	1	35	20	5	30
Witherow[28]	Not mentioned	1.5	Not mentioned	25	6	
Xu[14]	5–7	1	Not mentioned	Not mentioned	3	5.4
Oyama[26]	immediate 5-mm and then 5 days	0.5	Not mentioned	Not mentioned	6	25
Flores[15]	5	1	Not mentioned	Not mentioned	1.5	12
Shetye[21]	6	1	Not mentioned	16.1	1.5	12
Ko[29]	7	1–1.5	Not mentioned	17.7 orbital, 22.1 anterior nasal spine, 23.1 A point	4–5	24.6
Ahmad[30]	24 hours and 7 days	1.5 and 0.5	Sufficient advancement	Not mentioned	1.5	25
Shetye[36]	5	1	Not mentioned	Not mentioned	1.5–2	12
Bender[31]	5	5	Not mentioned	Not mentioned	2.8	Not mentioned
Kobayashi[37]	5	1	Not mentioned	15	2	
Greig[34]	5–7	1.4	Not mentioned	Central 13.2 Lateral 4.7	1.5–2	15-84
Mitsukawa[16]	Not mentioned	0.7 (internal)	30 (internal)	15 (internal) 21.8 (external)	5–6 (Internal) 2–3 (external)	12–102 (internal) 12–132 (external)
Dobbs[17]	2	1	21	Not mentioned		120
Hariri[27]	5	1–1.5	2–3	27 and 23	5–6	18
Hariri[33]	5	1	3	13	4	Not mentioned
Bouw[22]	7	1	Not mentioned	12.2	3	7.2
Wery[18]	Few	1	18	Not mentioned	3	Not mentioned

for the timing of the intervention depending on the effects of intracranial hypertension and developmental outcomes.

Indication

The technique of distraction osteogenesis for the gradual lengthening of the craniofacial structure in affected patients, and the timing of the intervention, are shown in Table 5.

Timing of the operation

There are different opinions on the growth potential of the face postoperatively, so decisions about the timing of the operation (at an early age or at skeletal maturity) should be strictly bound by the indications and severity of the condition. There has been much attention focused on obstructive sleep apnoea as one of the causes of sudden death in patients with midface hypoplasia, and severe exorbitism and raised intracranial pressure are absolute indications for advancement at an early age, otherwise the procedure can be done for children aged 5–8 years old or at skeletal maturity.[38] Wery et al showed the usefulness of treatment to solve these problems in early life by classifying treated cases into three phases. The early phase (first years of life) is when advancement of the midface is done only when the patient has obstructive sleep apnoea or exorbitism (with a risk of luxation of the eyeball). The intermediate phase (8–12 years) is when the operation is done at the request of the patient or the parents,[18] and the final phase is when the patients has reached the age of 18, when the intervention has the advantage that growth is complete. This classification was recommended by other studies.[14,17,18,39]

Mitsukawa et al reported that midface distraction was beneficial for young children to reconstruct a sizeable pharyngeal cavity, enlarge the airway, and improve obstructive respiratory disorders.[16] Witherow et al also reported that all the cases treated in early and intermediate life had resolved raised intracranial pressure and obstruction of the airway, together with the protection of the globes in all patients.[24] Despite the fact that treatment in young children has successfully solved most functional problems, it is important to measure the benefits and major complications such as leaks of cerebrospinal fluid and blood loss. Nout et al found an improvement in respiratory outcome after early Le Fort III advancement distraction in most young patients.[39]

Because Le Fort III and monobloc distraction osteogenesis are usually done for growing patients the midface will not grow forward after distraction. As a result, there will be a discrepancy in growth between the midface and the mandible (which will probably require secondary intervention).[29,30] Ko et al reported a patient who had orthognathic surgery to level the maxillary occlusal plane and to advance the mandible after monobloc distraction osteogenesis.[29]

Surgical technique

Le Fort III, facial bipartition, and monobloc distraction osteogenesis are each indicated according to the individual deformity and its associated functional problems. The primary indication for monobloc (combined fronto-orbital and midface advancement), instead of Le Fort III, is raised intracranial pressure together with neurological signs and symptoms.[40] This is reported to have a considerably higher morbidity (including infection such as meningitis).[41,42] Unlike a monobloc procedure, bipartition distraction osteogenesis is primarily indicated for midface central advancement in patients with Apert syndrome.[25,34]

Surgical technique and modification

The surgical technique needed depends on the severity of the condition. The advancement of the midface with the Le Fort III is indicated in those syndromes that include midface hypoplasia. Flores et al found that the nasopharyngeal and velopharyngeal airspaces were appreciably enlarged in 20 patients treated this way, and tracheostomy was avoided in 90% of those with obstructive sleep apnoea.[15] However, Le Fort III may not cure obstructive sleep apnoea in patients who have soft tissue problems such as enlarged adenoids, tonsils, and soft palate (under the uvula level). For them, further treatments such as advancement of the chin, mandibular advancement, or adenotonsillectomy may be recommended.[14,30,39] Preoperative nasoendoscopy, nasopharyngoscopy, hypopharyngoscopy and measurements of airway volume were recommended to identify the level of airway obstruction and incorporate the findings in the planned treatment.

The monobloc technique is indicated for the management of raised intracranial pressure, together with associated orbital and maxillary hypoplasia in patients with severe craniofacial synostosis and midface hypoplasia.[30] The fronto-orbital advancement reduces the internal cranial pressure, the maxillary advancement lessens the obstruction of the airway, and the advancement of the orbital rims relieves severe exorbitism. Ahmad et al used frontofacial monobloc advancement in the treatment of syndromic patients with symptoms that affected the eyes and airway, and so the aesthetic problems were improved simultaneously.[30]

With monobloc frontofacial advancement, reossification in the coronal bone defect is also of great importance, not only in the prevention of relapse but also in protection of the brain. Ko et al reported that treatment with a frontofacial monobloc was effective at different vertical levels of the face with the forehead moving forward by 3.8 mm. This was stable at 24 months' follow up (n = 5).[29] However, in some cases, the relations between the supraorbital area and the frontal bone might not be favourable, as a result of which they required differential advancement (fronto-orbital combined with Le Fort III), which might be difficult to achieve with the monobloc advancement as one bloc.[5,43]

Some factors decrease ossification during osteogenesis, such as older age at operation, history of previous intervention, the position of the osteotomy line, and the type of syndrome. For example, the ossification of the cranium is considerably reduced in patients who have had previous operations, and in older patients.[26] Jensen et al reported that patients who had had Le Fort III procedures had superior bone deposition in the zygomatic arches when the osteotomy was placed at the site of greater bone volume (body of the zygoma), while there was less bony deposition when it was placed in the area of thinnest bone (the middle of the arch).[35] Another study showed that the distraction gap of the monobloc segment influenced the rates of osteogenesis among the syndromes, with patients with Pfeiffer syndrome having the least reossification.[26]

In some circumstances, the surgeon needs to modify his established technique. Dobbs et al reported a young patient with a scaphocephalic skull who required correction of exorbitism and raised intracranial pressure, and they used midface advancement distraction in conjunction with remodelling by staged calvarial expansion rather than fronto-orbital advancement alone, as it is often better at correcting exorbitism. Despite this, they stated that monobloc advancement was better than their technique.[17]

Mavili and Tuncbilek added another osteotomy line (1.5 cm) inferiorly to the original orbital lateral wall in the classic Le Fort III osteotomy to prevent prominence of the inferior part of the lateral orbital rim (in the case of appreciable advancement of the midface). The results were stable, and the contour of the lateral orbital wall was satisfactory.[20] In the case report by Cohen and Holmes, they planned a staged procedure instead of a one-stage operation because of the magnitude of the cranial defects and the fragility of the coronal scar.[19] Mahili and Tuncbileg used a two-level osteotomy (instead of one line as in a Le Fort III) to provide a smooth conjunction between the frontal bone and the Le Fort III segment.[20] A long pericranial flap between the osteotomy cuts sutured at the distal part of the flap has been used to prevent leakage of cerebrospinal fluid by providing a barrier between the nose and the anterior cranial fossa.[28,34]

In the frontofacial monobloc technique, a 5 mm coronal osteotomy bone gap for advancement of the forehead (to accelerate the reduction in intracranial pressure) was filled by bone dust to improve reossification.[26] Bipartition distraction for advancement of the central midface was reported in only two papers as a preferred technique in patients with Apert syndrome.[25,34] The curvature of the frontal bone can be increased by removal of a V-shaped segment from the midline area of the frontal bone, and this allowed medial and upward rotation of the lateral orbits, and increased the amount of projection of the central facial skeleton compared with laterally.[34] Dobbs et al used a laminated bone graft instead of titanium mesh to protect the thin temporal bone from the external distractor after penetration of the pins.[17] Hariri et al used headgear that provided a protective platform at the temporal region, which prevented intracranial perforation of the pins and relapse of the central component.[27]

Selection of the distraction device

As experience with the technique of distraction osteogenesis grows, research is focusing on the development of new internal and external devices that seem to be used at even frequencies. However, selection of a device is often a challenge. Both internal and external devices have their advantages and disadvantages (Table 11, Supplemental data, online only).

Selection of the proper type of distractor is based on many factors such as the age of patients, the amount of distraction required, the rigidity of the zygomatic arch, and the surgeon's technical preference. As shown in Table 11, one of the disadvantages of the external device is that its distraction distance is limited.[44] From a psychological perspective, distraction to the final adult maxillary position is also not recommended.[45]

The disadvantage of the internal device is that it is usually necessary to make a coronal incision in the scalp to remove it, but this can be solved by using biodegradable materials as the foot plates.[19] Because of the plasticity of a young skull there is a tendency for the face to bend around the distraction points, so when the devices are placed, distraction vectors cannot be varied. The skull bones in children younger than 3 years old are fragile and too thin to retain external fixation screws, which makes internal distractors more suitable for young children.[5] However, the internal distraction device is not recommended for patients with developmental hypoplasia because of the weak zygomaticomaxillary union, and the risk of disjunction and fracture of the distracted bone during removal of the appliance. In these cases, external distractors are better.[29]

The combination system has many advantages that may overcome some of these problems. Kobayashi et al showed that it was particularly well-suited for patients (n = 2; Crouzon syndrome) with severe scarring after multiple interventions during follow up, and for patients who required distraction of 25 mm or more. However, the bigger the difference between the vectors of the orbit and point A, the more difficult it was to set cephalometric targets.[37]

It is important to select the most suitable device for any child for early midface distraction. Mitsukawa et al reported two types of external devices – a transfacial pinning system (n = 6) and a halo type (n = 3). The transfacial pinning system was considered to be the most suitable for patients aged less than 2 years, as it is safer and lighter (although scarring of the cheek by the pin was noted in half of the patients). In patients aged 2 years or over, however, the internal device (zygoma-zygoma) was recommended because of the thickness of the temporal bones. However, in cases where fracture of the maxillozygomatic suture was a concern, an external device (halo-type) was recommended, as the device advances the entire maxilla en bloc from its centre rather than laterally.[16]

Titanium mesh applied to the skull beneath the site of the pin has been described as a method of strengthening the construction and allowing application of the rigid external distraction frame at a young age, as the skull bone may be too thin.[28] Ahmad et al[30] reported that when young patients wore a halo-type device, a moulded rectangle of titanium mesh (KLS Martin, Tuttlingen, Germany) was placed on the lower portion of the bilateral parietal bones. Although titanium mesh prevented intracranial injuries from penetration by the pins, it was considered a foreign material that had the potential to migrate or cause long-term problems,[14,16,17] and bone could grow over the titanium mesh.[28,30] A new technique for overcoming penetration of a pin was to use the patient's cranial bone to create laminated bone grafts, which thicken the site of insertion of the pin and secure the stability of fixation of the device.[17] Another solution, reported by Cohen and Holmes, was using resorbable mesh.[19] However, concerns have been raised about its strength, and its ability to prevent such penetration.

A modular internal distraction system is a device that offers benefits over external devices in that it is more acceptable to patients, makes wound care easier, and has superior stability.[46] One advantage is the complete controlled advancement of the entire midface, but it requires substantial operative exposure and can be frustrating.[20] Cohen and Holmes introduced biodegradable plates for internal distractors, which permit internal distraction without the need for complete removal. However, a second operation is still necessary to remove the distractor screw and cable-drive. A biodegradable design was developed in 2001 to replace the remaining metallic components,[19] but more internal biodegradable and multidirectional distraction devices are still needed.

The movement of different levels of the face can be controlled by the rate and duration of activation at different parts of the distraction ports. Jensen et al decided that a more superior point of attachment of the frame of the external distraction device on the nasal bone gave sufficient vector control of the segment, and this solved the problem of counter-clockwise rotation in a patient who had had a standard Le Fort III to increase advancement at the nasofrontal junction. They explained that the "pull" force achieved by the external device is concentrated along the inferior aspect of the Le Fort III segment, and as a result there was compression force at the nasofrontal junction that prevented its advancement.[35] The distraction distance and vector of the orbital and occlusal levels will differ in patients with short facial height (Apert syndrome), so a procedure is recommended that integrates Le Fort III osteotomy, Le Fort I and II osteotomies, and downward facial rotation with the transmaxillary K-wire as the axis, with a splint and elastic traction.[37]

Distraction protocol

Most of the studies that we reviewed followed the standard distraction protocol. The protocol originally introduced by Ilizarov for the extension of tubal bones entailed a latency phase of 5–7 days, a distraction rate of 1 mm/day, and consolidation periods of one to several months. A modified protocol with a minimal latency period, rapid distraction, and reduced time for consolidation have also been described.[47,48] The rationale for this approach – immediate postoperative distraction – was to reduce the chances of the foot plate loosening by immediate expansion of the brain (which may lift the foot plates off the distraction device). In another study, the advancement was stable without relapse or complications seven months postoperatively (distraction was started on the first postoperative day with a distraction rate of 1 mm/day and a consolidation period of 1.5 months).[20]

Oyama et al reported an increase in ossification of the bony defect with no cases of postoperative leakage of cerebrospinal fluid when they placed bone paste in an immediate (5 mm) advancement osteotomy gap.[26] Leakage that resulted from a high rate of distraction with a short latency period was reported in many studies.[24,28,30] In an attempt to prevent it on distraction, the routine that involved distraction of 1.5 mm/day starting 24 hours postoperatively, was changed to one of 1 mm/day starting seven days postoperatively.[24,30]

Complications

Some complications can be managed without difficulty, such as severe diffuse blood loss replaced during and after operation, skin infection at the site of a transcutaneous pin that resolved after removal of the pin, or a subcutaneous abscess and a leak of cerebrospinal fluid managed by lumbar drainage.[31]

The impact of the disjunction force of the osteotomy may cause the developing tooth germs to rotate. A patient reported by Ko et al had rotation of the tooth germ of the upper second molar after monobloc distraction, which had self-corrected 4.5 years later.[29]

Selection of the most suitable distraction device that affords good result with few complications is important. The application of internal devices (zygoma-zygoma) for Le Fort III maxillary distraction is effective, and guarantees a more comfortable life than external devices of the halo-type during long periods of consolidation. However, they do have their complications.[23] Dobbs et al reported that internal devices might have led to lateral, but not medial, distraction of the mid-face, and this limits functional improvement.[17] As an alternative, good results have been obtained with external devices in a salvage procedure in patients with Apert syndrome who developed severe temporal bulging, and those with fractures along the maxillozygomatic suture that were caused by an internal device.[16]

A combination distraction device has the advantages of both external and internal distraction devices, and have been used successfully to control both lengthening distance and vector. This type of device is particularly indicated in patients with severe scarring and those who require more distraction.[37] Another system has been developed to over-

come deficiencies in distraction devices by substituting the metallic fixation plates with biodegradable devices that permit early removal of the distractor screws. This system will provide stability and protect bony regeneration and distraction without the need for complete removal.[19]

Four different movements have been recorded for the location of the vector and the direction of application of the force in Le Fort III procedures: anterior translation, clockwise movement, counter-clockwise movement, and anterocaudal translation.[18,21] Counter-clockwise rotation is the most dominant movement after Le Fort III advancement. Despite the linear anterocaudal translation of the Le Fort III segment achieved in 18 patients, counter-clockwise (external distractors, n = 13) and clockwise (internal distractor, n = 1) movements have been reported that have led to unfavourable anterior open bite and malocclusion. Bouw et al suggested that the counter-clockwise rotation may happen because the vector of the force applied is oriented too far cranially, or the centre of resistance has no fixed location, or there is a difference in the resistance of the soft tissue between the upper and lower parts of the midface.[22]

Figueroa et al suggested that forces were acting both above and below the centre of mass to create rotational control, which prevented the counter-clockwise rotation,[49] while Bouw et al proposed that the distraction forces after Le Fort III osteotomies may be oriented extremely caudally to prevent the counter-clockwise rotation.[22]

Placement of distraction pins can be varied using an external distractor device, which allows application of different distraction forces. Ponniah et al reported more medial advancement and increased volume in the upper airway after monobloc distraction when they used a rigid external distractor with the pins placed medially. This effect was more pronounced after bipartition distraction.[25]

In summary, distraction has proved to be beneficial for complications related to operations for syndromic craniosynostosis. It has shown great potential, movement, and flexibility with both early Le Fort III and repeated Le Fort III procedures after skeletal maturity. However, monobloc frontofacial advancement may be used on younger children if their functional problems require major intervention. Advancement of the midface may be planned in the first years of life for absolute indications such as obstructive sleep apnoea or severe exorbitism.

Previous operations can be an important cause of poor viability of bone, which leads to morbidity and increases the risk of complications.[26,50] If the patient is only mildly affected, elective operations can be postponed until skeletal maturity has been reached after puberty, and may then be done for relative functional and aesthetic reasons. Although the osteotomy of monobloc frontofacial distraction osteogenesis is higher, and away from a tooth-bearing area, the developing tooth germs should be monitored carefully. Improvement in distraction devices is therefore needed.

Most internal devices available nowadays cannot influence the direction of distraction postoperatively. External devices are needed for this. However, infantile skulls are too fragile for fixation of a device. Even using a mesh causes complications on removal, so external devices are used for children over 4 years old. Otherwise, a bone graft laminated to thicken the calvarial bone can potentially prevent penetration by the pins and retain the distraction frame. Devices made with absorbable materials may prove to be successful, but long-term follow up is necessary. We think that grafting with bone paste is extremely effective, particularly as a means of overcoming unfavourable conditions that require reossification when an immediate advancement is done intraoperatively. This may contribute considerably to any increase in the reossification osteotomy gap.[26]

Conclusion

In conclusion, distraction osteogenesis is a potential alternative procedure when a greater degree of advancement is required. Despite the need for a second operation for the removal of the distraction device, it is still a viable option for advancement. For older children whose conditions vary from mild to moderate severity, it may be useful to avoid a bony defect for a longer period. New biodegradable distraction devices are needed to overcome the problems encountered with non-biodegradable devices. We advocate long-term follow up of newly-developed combination and biodegradable distraction devices in the future.

Conflict of interest

We have no conflicts of interest.

Ethics statement/confirmation of patients' permission

Neither was required.

Appendix A. Supplementary data

Supplementary data associated with this article can be found, in the online version, at https://doi.org/10.1016/j.bjoms.2018.03.002.

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