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ORIGINAL PAPER

Posterior cranial vault distraction osteogenesis in craniosynostosis: estimated increases in intracranial volume

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Abstract

Purpose To study distraction osteogenesis of the posterior cranial vault in children requiring increased intracranial volume.

Materials and methods Ten patients were treated with cranial distractors. Five children had previously been operated for scaphocephaly and one child for Saether-Chotzen syndrome. Two patients had bilateral coronal suture synostosis with Muenke syndrome and two patients had Apert syndrome. At surgery, the cranial bones were mobilized, the head was widened during surgery, and the segments fixed to each other with distractors. Further expansion at a rate of 1 mm/day was performed over 2–4 weeks. The cranium was distracted posteriorly from 20 to 30 mm.

Results The patients all tolerated surgery and distraction well. In all cases, the parents were able to perform the distraction at home. There were no technical problems with the distraction devices. Two cases had minor cutaneous problems, where the distractor penetrated the skin. These cases responded to gentle local wound care measures. At the time of distractor removal, ossification had occurred sufficiently in one of these two cases. In the other case, the device was removed and replaced with a resorbable plate, without any harmful effect on the result. In all cases, sufficient expansion was achieved without causing more cosmetic deformity. Ossification occurred in all cases. This method seems effective, as the calculated increase in intracranial volume was a mean of 20.2% (range 10.2–28.5%).

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Conclusions This preliminary series shows that cranial bone distraction is a useful method for cranial expansion with low morbidity in children with craniosynostosis.

Keywords Craniosynostosis · Distraction osteogenesis · Posterior plagiocephaly · Intracranial volume

Introduction

Craniosynostosis results from premature fusion of one or more sutures of the skull resulting in a cascade of disturbances in the normal growth of the brain and skull including increased intracranial pressure (ICP) [1]. Classic presentations of elevated ICP can include headaches, nausea, and vomiting. Softer more subtle signs may manifest as a change in the child's behavior, excessive somnolence, or retarded cognitive development noticed by astute parents or caregivers.

Methods to treat craniosynostosis such as cranioplasty with cranial vault remodeling all seek to increase intracranial volume [2]. The stability of calvarial fragments has been greatly improved by the use of resorbable fixation [3–5]. Increasing the intracranial volume by posterior cranioplasty is inherently unstable. The skin is often tight over the repositioned fragment. When the child sleeps in the supine position, the force on the repositioned fragment of the cranium also drives it back to its original position. In addition, fusion of the sutures following cranioplasty may require multiple secondary revisional cranioplasties.

If the fusing ends of the calvarial bones are kept apart while progressively expanding the skull, the skull volume may be gradually increased in volume so that subsequent fusion might not be of clinical significance. Springs have been used to provide continuous traction on cranioplasty fragments [6, 7]. However, the amount of tension on the fragments is not under the continuous direct control of the clinician so that the method is indirect.

Distraction osteogenesis (DO) is a biological process which promotes bone formation between the cut surfaces of bone segments while traction is applied to separate the bony fragments [8–12]. Modern clinical DO of the craniofacial skeleton began once McCarthy applied the concept to mandibular lengthening [13]. This led to an explosion of clinical and research activity in craniomaxillofacial distraction osteogenesis over the past decade [14, 15]. As in other sites, craniofacial DO involves five distinct periods: osteotomy, latency, distraction, consolidation, and remodeling [14]. The gradual distraction of the osteotomized fragments allows the surgeon unparalleled control in gradually increasing the gap between the cranioplasty fragments, and the consolidation period helps to prevent relapse of the fragments back to their original positions.

Posterior cranial expansion should be considered as an initial procedure particularly for syndromic cases with bilateral coronal synostosis. Operative procedures have been lacking for those cases where the shape of the head is good but with insufficient intracranial volume. Posterior expansion allows for an increase in intracranial volume and with minimal cosmetic disadvantage.

Recently, White et al. described their experience with posterior calvarial vault expansion using DO in six patients [16]. While increases in intracranial volume were attained, five of the six patients in their series sustained hardware loosening complications. The authors suggested that redesigning distractor hardware may address these complications. The purpose of this case series is to report on the current authors experience using miniaturized hardware and removable distractor stems that allow consolidation with the minimum of hardware that might be traumatized during the post-operative period.

Materials and methods

Patients

A total of ten patients, six males and four females with ages between 2.5 months and 7 years were included in this series. Of the ten patients, there were five with scaphocephaly one with Saethre-Chotzen syndrome, two with brachycephaly (Muenke syndrome), and two with Apert syndrome. In all patients 3D CTs, stereolithic skulls and clinical photographs were obtained pre-operatively. CT scans and plain films were taken as required post-operatively at follow-up. The follow-up time was a mean of 1 year, ranging from 6 months to 2 years (Table 1). The main indication for surgery in this group of patients was raised intracranial pressure (ICP) rather than cosmetic indications. Of the ten patients, eight had either measured raised ICP or clinical signs of raised ICP including a change in behavior, nausea, vomiting, or excessive somnolence.

Operative procedure

These operations were carried out through wavy line or zig-zag coronal incisions anterior to the vertex of the skull. The posterior cranium was exposed to allow the planned craniotomies as required. In two patients with scaphocephaly, the cranial fragments were elevated as three segments and then were reconstituted in an expanded arrangement to further increase cranial volume using distractors. These segments were fixated into one solid fragment using resorbable plates of 1.5 mm PLGA (Inion CPS® baby, Tampere, Finland). In the remaining eight cases, the posterior cranial vault was mobilized in one piece. In nine cases, the cranial bone fragments were totally detached from the underlying dura leaving one case where the cranial bone was not totally detached from the underlying dura.

Table 1 Diagnosis and distraction particulars

Case description and age at distraction	Distractor removal (months post-op)	Distraction distance (cm)	Pre-op L (cm)	Pre-op L/2 (cm)	Pre-op H (cm)	Volume of half ellipsoid (cm^3)	V-gain (cm^3)	V-gain (%)
1. (KM) Redo-scapho, 19 months	3	1.2	15.6	7.8	8.6	1,207.6	151	12.5
2. (KT) Redo-scapho, 25 months	4	1.3	17.7	8.9	8.4	1,307.2	149	11.4
3. (RT) Redo-scapho, 60 months	3	1.2	17.6	8.8	7.5	1,036.2	106	10.3
4. (TP) Saethre-Chotzen, 7 years	4	3.0	18.1	9.1	9.2	1,603.48	399	24.9
5. (VK) Brachycephaly, 7 months	4	2.7	15.7	7.9	7.5	948.99	239	25.2
6.(LR) Apert, 10 months	4	2.3	17.1	8.6	9.2	1,511.6	305	20.2
7 (AE) Muenke 14 months	3	2.5	17.6	8.8	8.8	1,426.55	304	21.3
8. (KP) Redo-scapho, 23 months	6	2.6	17.9	8.9	7.7	1,110.82	242	21.8
9. (MM) Redo-scapho, 29 months	4	2.8	16	8.0	8.8	1,296.87	340	26.2
10 (HH) Apert, 17 months	3	3.0	15.8	7.9	9.1	1,369.5	390	28.5

The single vector distraction devices with quick-disconnect distraction rods (Biomet Microfixation 1.5 mm CMF Quick-Disconnect Distractor®, Biomet Microfixation, Jacksonville, Florida, USA) were fixed on either side of the skull osteotomies to the parietal, temporal, or occipital bones to provide a posterior vector of distraction (Fig. 1). The upper part of the devices were fixed to the slightly re-contoured calvarial segments, each device being secured with four 1.5 mm titanium self-drilling screws. The rods of the distractors were bent slightly downwards towards the bone to keep them close to the plane of the cranial bone, in order to minimize skin distension.

Once the distractor rods were attached, the devices were opened to ensure that the vectors were all complimentary and not interfering with each other. The devices were then closed back down with 3 mm of a distraction gap remaining (Fig. 2). The devices were then buried beneath the scalp and the quick-disconnect rods were attached to the distractor devices so they would emerge through the skin through small stab incisions, distant to the zig-zag skin incisions. All incisions were closed. No drains were used. The distraction protocol was started in all ten patients within 5 days of distractor placement (Fig. 3). The distractor rods were removed under general anesthesia once the desired distraction distance had been attained. No additional incisions were necessary. The distractors were removed at a third anesthetic after the consolidation period (Fig. 4). None of the ten patients showed signs of infection through any of the phases of distraction care.

Further expansion of 1 mm/day was performed during 2–4 weeks either posteriorly or upwards, thus gaining cranial expansion in two to three directions. The cranium

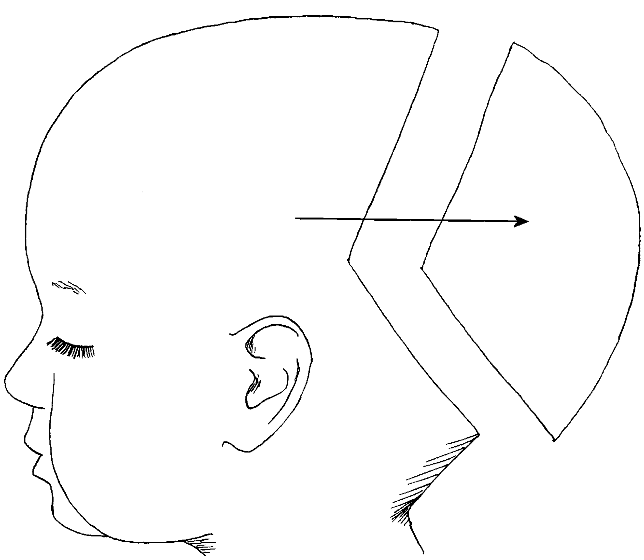


Fig. 1 Diagram illustrating the desired posterior vector of distraction for the ten patients in this series

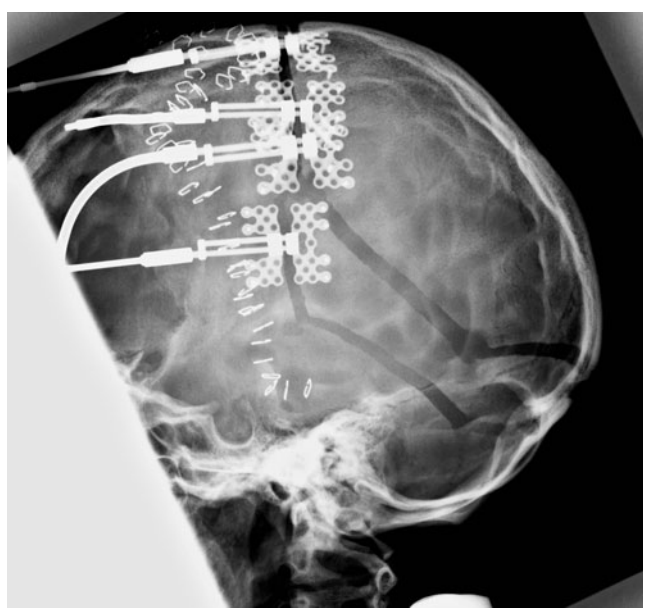


Fig. 2 Lateral skull film at beginning of distraction showing osteotomies and distractors

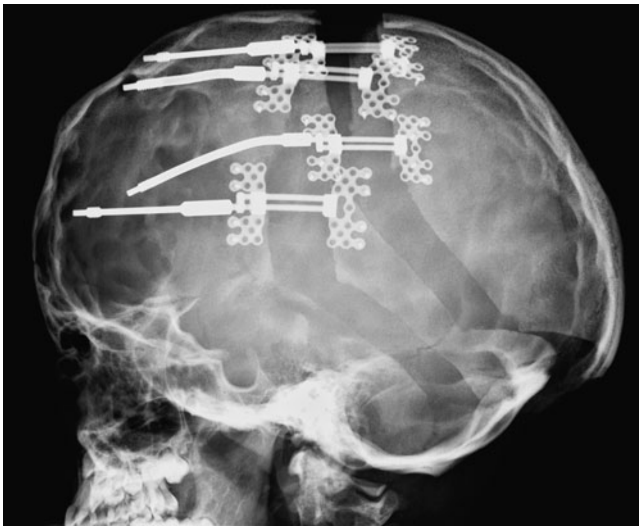


Fig. 3 Lateral skull film at end of distraction showing posterior displacement of the cranial flap

was distracted from 20 to 30 mm (Fig. 5). In all cases, the parents were taught to apply the distraction forces post-operatively. This allowed early discharge of the ten patients home from the hospital. None of the ten parents reported any difficulties with the application of the distraction or the schedule of distraction. They all felt comfortable to be an integral part of their child's treatment.

Results of advancement of the posterior cranial fragment by means of distraction

Osteogenesis

At the beginning of distraction, patient 1 was 19 months of age and had undergone two previous cranioplasties. Patient 2 was 25 months of age and had undergone one previous

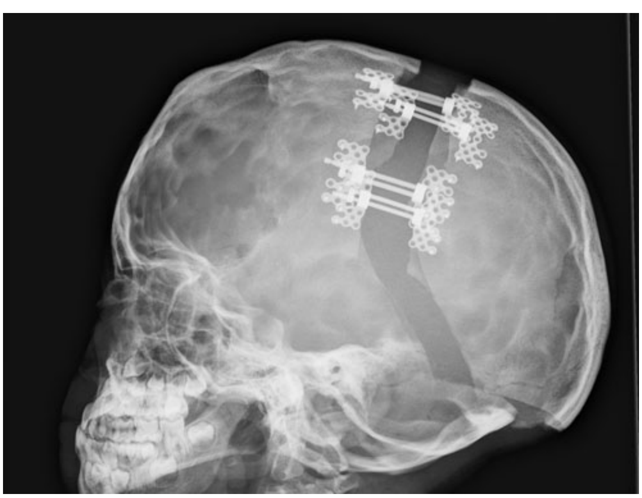


Fig. 4 Lateral skull film at time of distractor rod removal with distractor left in to provide retention during consolidation phase

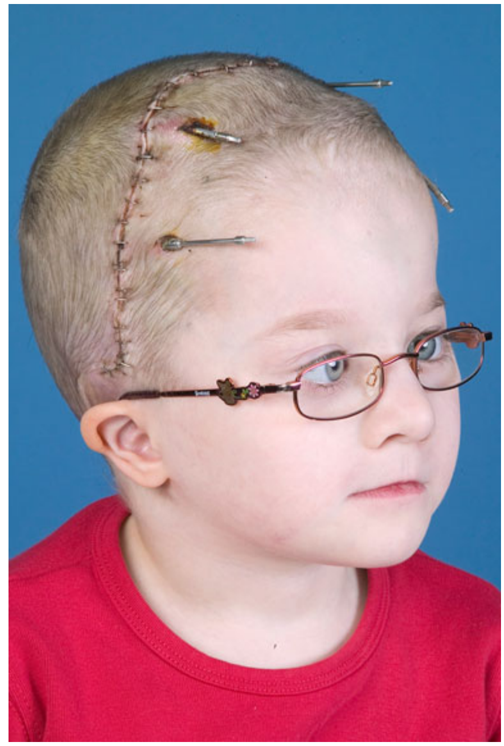


Fig. 5 Photograph of patient 4 with well-tolerated distractor rods and their skin entry sites

cranioplasty. Patient 3 was 60 months having had one previous cranioplasty. Patient 4 was 7 years of age with two prior cranioplasties. Patient 8 was 23 months with two prior cranioplasties. Patient 9 was 29 months with two prior cranioplasties, and patient 10 was 17 months with one prior cranioplasty. Patients 5, 6, and 7 had no prior cranioplasty experience. All re-operations in this series were performed for the indication of raised ICP including the five scaphocephaly patients and the syndromic patients. Clinical signs of raised ICP were present in all eight patients with either measured raised intracranial pressure or those with clinical signs of raised ICP pre-operatively. The results of the ten patients are presented in Table 1.

The following two formulas were applied to pre-operative measurements of the distracted skulls on CT scan and to the distances distracted. An assumption was made by the authors, that an infant skull which has been considered to resemble a sphere, was instead most like an ellipsoid in shape (Fig. 6). Since the area of focus was the upper half of the skull, the volume of half of an ellipsoid can be calculated by using the formula:

$$V = \frac{\frac{4\pi ABC}{3}}{2}$$

where A is half of the length of the skull, B is half of the height of the skull, and C is half of the width of the ellipsoid or skull [17]. The estimated change or gain in

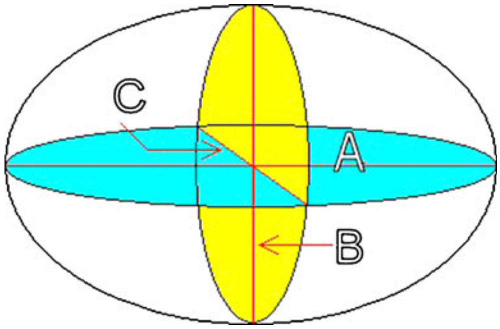


Fig. 6 Representation of the dimensions of an ellipsoid where A is half of the length of the skull, B is half of the height of the skull, and C is half of the width of the ellipsoid or skull

volume (Vc) generated by distraction can be calculated by using the formula:

$$Vc = \frac{\pi r^2 h}{2}$$

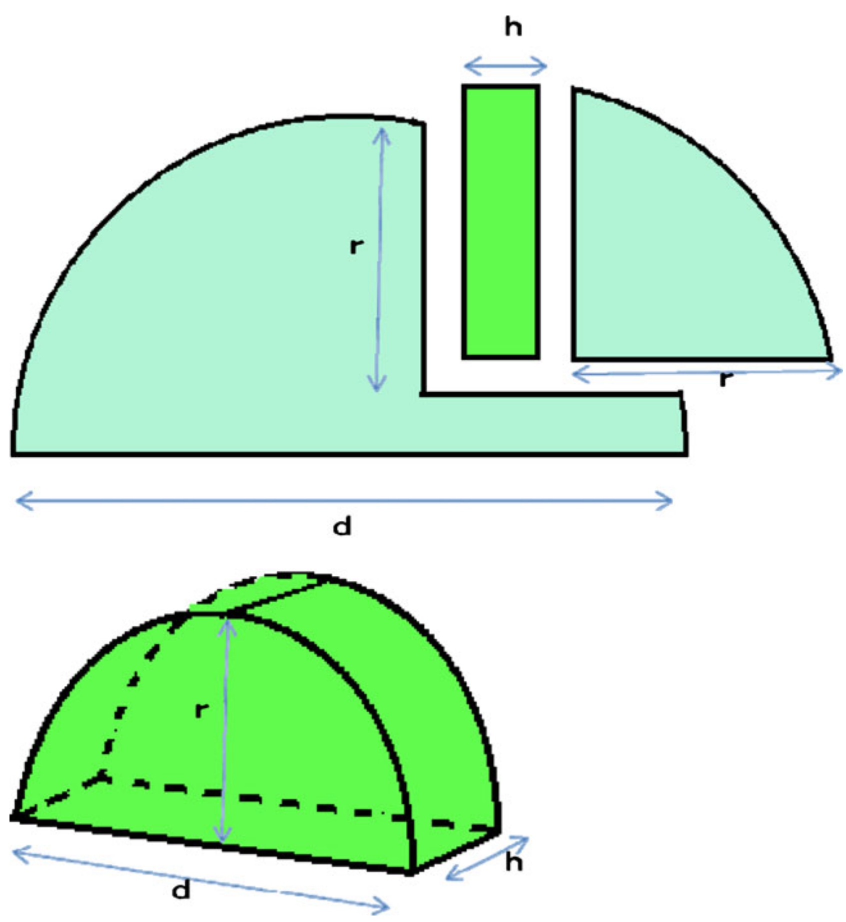
where r is the height of half of the cranium or the radius, and h is the amount of distraction (Fig. 7). The gains in volume ranged from increases of 106–399 cm^3 or 10.2–28.5% with a mean of 20.2%. This method provides an estimation of volume change. To check its reliability, pre- and post-operative 3D CT scans would be necessary. Since the post-operative 3D CT scans would be done for solely research purposes and are not a routine part of clinical care for these patients at our institution, the authors would not have received the necessary ethical approval for that part of the study.

Discussion

The distraction goals were met in all ten patients. There were no cases of hardware loosening in this series. All patients tolerated their devices to the planned end of their consolidation phases without any interruptions. There were no infections involving the peri-distractor tissues or the zig-zag scalp incisions.

Immediately following commencement of distraction, the appearance of the skulls began to show marked improvement in their shape. The length of stay of the distraction patients was no longer than for cranioplasty patients at our center, and the parents were able to perform their child's distraction at home. There were no complications such as premature disconnection of the quick-disconnect distraction stems or dropping out of the

Fig. 7 Diagrammatic representation of volumetric increase with cranial flap distraction, where r is the height of half of the cranium or the radius, and h is the amount of distraction



distraction stems occurred. Distractor stem removal was simple with the quick-disconnect couplings. This occurred at a second short outpatient general anesthethic once the desired amount of distraction had been attained. The remainder of the devices then remained buried in their subcutaneous positions to provide retention during the consolidation phase.

A third short outpatient general anesthetic was necessary for the removal of the titanium distraction devices where a short skin incision was used. The distracted gap had ossified with the same appearance as the surrounding cranial bone.

All ten patients developed normally following the distraction protocol. There were no signs of cognitive delay or behavioral disturbances. Ocular examinations did not show signs of papillary edema nor were there other signs of raised ICP.

There were a few differences between this case series and the report of White et al. [16]. In nine of the ten cases, the cranial bone fragments were totally detached from the underlying dura unlike in the White et al. series. There were no instances of distractor hardware loosening in this group. The current authors believe that placing the distractor stems as flat as possible against the outer layer of the cranial bone is a very important maneuver. This keeps the distractor stem less proud and less likely to sustain trauma. Removal of the distractor stems keeps the devices further away from the risk of traumatic dislodgement. None of these devices came loose, and all patients were able to keep their hardware up to the planned end of their consolidation phases.

One of the major advantages of a distraction technique using miniaturized hardware versus springs is the inherent control that such hardware provides. Springs by their nature are totally buried beneath soft tissue and their control is indirect at best. Miniaturized distractors precisely allow control of the rate of distraction, the amount of distraction, and when to stop distraction.

Posterior calvarial vault expansion using DO is a safe technique. There were no infections despite hardware that was only partly covered by skin. Increases of up to 37.6% of the intracranial volume were possible by this technique. Long-term monitoring of these patients and a larger scale trial will help to further establish the safety of this technique and whether two, three, or four distractors are required to provide the desired increase in intracranial volume in a predictable manner. Future studies to correlate the increase in intracranial volume with the extent of cosmetic improvement and showing cosmetic changes over time with serial photography are also necessary.

Syndromic cases of craniosynostosis were previously treated with frontal advancement, which usually had to be re-operated. Posterior cranial vault distraction results in gains of intracranial volume which are greater than with frontal advancement. Such gains may bring the added benefit that the patients do not need any frontal advancement, which is of benefit if they later need facial or orbital remodeling.

In the syndromic cases with Muenke and Apert syndrome, the posterior expansion gained an increase of cranial volume between 21% and 37%. Future follow-up will show whether this will be sufficient or if the patients will need further frontal advancement. Avoidance of frontal advancement would spare the patients from a major procedure.

In those patients who have been previously operated for plagiocephaly or scaphocephaly whose head has a good shape, but remains too small, we traditionally have difficulties in gaining sufficient expansion without adverse cosmetic outcomes. Posterior cranial vault distraction may overcome such difficulties.

In untreated scaphocephaly, there is an increased antero-posterior dimension. Posterior cranial distraction of untreated scaphocephaly would worsen the cranial index. However, in our recurrent scaphocephaly patients, there was raised ICP with headaches and papilledema. Here, the intracranial volume was felt to be too small, and the authors chose posterior cranial distraction to increase the intracranial volume without an appreciable change in cosmesis. The procedure was not used as a cosmetic procedure in these cases but rather to treat raised ICP in this small group of patients. Further study is required to quantify the cosmetic changes that posterior cranial vault distraction produced in these patients.

While cranial or cephalic index provides one way of quantifying changes in cranial shape in scaphocephaly, it is determined by the use of anterior–posterior and lateral cephalometric X-ray studies. These radiographs were not done as part of this study in order to minimize radiation exposure in these patients. Since these X-rays are not a routine part of clinical care for these patients at our institution, the authors would not have received the necessary ethical approval for that part of the study.

In the future, as resorbable devices become more reliable, hybrid metal and resorbable distractors or completely resorbable distractors [18] will serve to decrease the number of post-operative anesthetics that are necessary. Such reductions of interventions will make this technique more patient- and family-friendly.

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