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# Internal carotid dissection after Le Fort III distraction in Apert syndrome: A case report☆

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*SUMMARY.* **A 10-year-old girl with Apert syndrome underwent a Le Fort III osteotomy with the positioning of internal and external distraction devices. The operation was straightforward with no intraoperative complications. Very soon after completion of surgery an anisocoria (unilateral dilation of a pupil) was noticed. This was followed by intracranial oedema which was fatal. The aetiology was dissection of the right internal carotid artery is reported. The complications of Le Fort osteotomies are discussed regarding patients with complex syndromal craniosynostosis and midface hypoplasia, such as Apert syndrome. © 2010 European Association for Cranio-Maxillo-Facial Surgery**

***Keywords:*** **complication, midface distraction, Apert, cerebral, dissection**

## INTRODUCTION

Midface osteotomies and distraction osteogenisis have become common procedures. The reported rate of complications is low. Most have been described with the Le Fort I osteotomy including haemorrhage, arteriovenous fistula, and ophthalmic symptoms, of which blindness is the most severe (*Bendor-Samuel* et al., 1995; *Tung* et al., 1995; *Girotto* et al., 1998; *Lo* et al., 2002; *Cruz* and *dos Santos*, 2006; *Morris* et al., 2007). Of all midface procedures the Le Fort I osteotomy is the most popular and best documented.

Few complications have been described with the Le Fort osteotomies and distractions performed at a higher level, such as the Le Fort II and III (*Le* et al., 2001; *Rieger* et al., 2001; *Matsumoto* et al., 2003; *Brown* et al., 2006; *Nout* et al., 2006). This is surprising as Le Fort III distraction in a patient with syndromal craniosynostosis including midface hypoplasia is a major operation which occasionally involves serious co-morbidity.

A review of the current literature of this journal produced only 1 case report and 1 oral presentation pertaining to complications with the Le Fort III osteotomy, both reporting only halo-pin related complications (*Van der Meulen* et al., 2005; *Spinelli* et al., 2006).

We present the case of a 10-year-old girl with Apert syndrome, in which a Le Fort III osteotomy with the placing of internal and external distraction devices was complicated by fatal intracranial oedema as a result of internal carotid artery dissection. The complications of Le Fort procedures are discussed including the role of performing this surgery in patients with complex syndromal craniosynostosis and midface hypoplasia such as Apert syndrome.

## CASE REPORT

A 10-year-old girl with Apert syndrome including midface hypoplasia, class III malocclusion and mild exorbitism had a Le Fort III distraction (Fig. 1). Previously the patient had a tracheostomy at the age of 13 months because of severe obstructive sleep apnoea syndrome (OSA). After a monobloc distraction at the age of 15 months she was decanulated at the age of 19 months. Following this there were no clinical signs of OSA and polysomnography prior to the Le Fort III demonstrated no OSA (oxygenation–desaturation index $< 1$). The pre-operative computed tomography (CT) scan showed normal sized ventricles and subdural space.

After an uncomplicated Le Fort III osteotomy bilateral internal midface distractors (Marchac devices, Martin Medizin Tuttlingen, Germany) were positioned with a few millimetres of distraction and the Rigid External Distraction Device-II (RED-II, Martin Medizin Tuttlingen, Germany) was applied. Total operation time was 3

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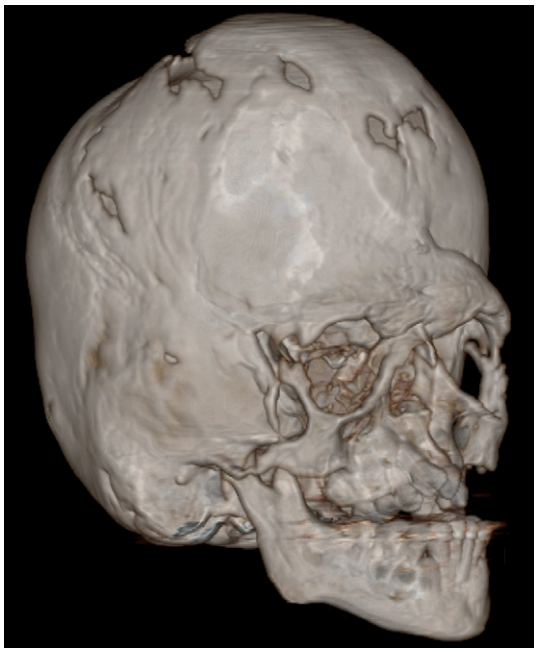

**Fig. 1** – Pre-operative 3D CT scan of 10-year-old patient with Apert syndrome in which a Le Fort III distraction with internal and external devices was planned.

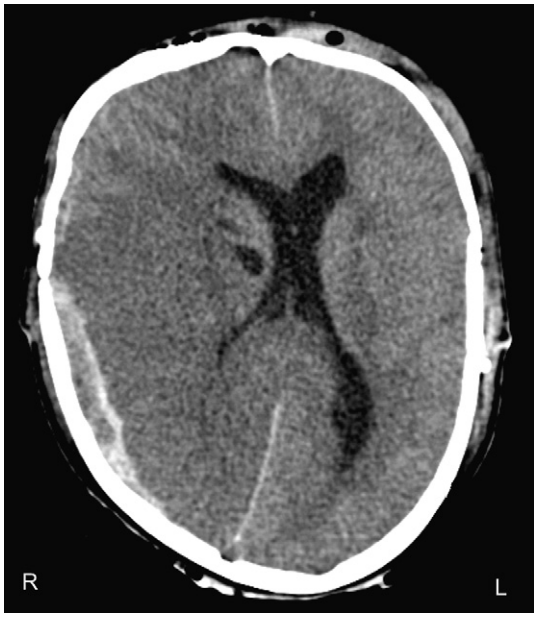


**Fig. 2** – The first postoperative CT scan revealed subdural haemorrhage on the right parietal side, a cerebral midline shift to the left, a swollen, hypodense right hemisphere and uncal herniation.

and a half hours, with a blood loss of 1.5 l. Near the end of the operation a drop in blood pressure and a tachycardia was noticed, with rapid normalisation. At the end of the operation an anisocoria was noticed. A CT scan revealed a subdural haemorrhage in the right parietal region, a cerebral midline shift to the left, a hypodense right hemisphere and uncal herniation (Fig. 2). The parietal subdural haemorrhage itself appeared too small to explain the severe midline shift. Direct neurosurgical intervention was initiated with decompression and the removal of all distraction devices. Two puncture wounds of the dura were found at the site of the internal distractor, without sign of bleeding. After opening the dura a large swelling of the cortex with decreased vascularisation was noticed. There was significant loss of clotted and fresh blood in the temporo-occipital, temporal and frontal areas. The boneflap was buried subcutaneously in the abdomen and a transposition flap of the scalp was performed to cover the exposed brain. A total of 6 l of blood was lost.

At the end of the procedure an intracranial pressure (ICP) monitoring device was inserted. The ICP at this stage was 34 mm Hg (normal value: 7–15 mm Hg). A CT scan demonstrated increasing hypodensity of the right hemisphere, suggestive of ischemia, an increase of the cerebral midline shift and a complete compression of the peripheral subdural space (Fig. 3). Although the neurosurgeon added an external drain the ICP increased to 50 mm Hg.

To investigate the possible cause of the brain ischemia CT-angiography (CTA) was performed (Fig. 4). As an adjunct to the CTA we used the V-Scope volume rendering application in the Erasmus MC I-Space, an immersive virtual reality (VR) system, to provide interactive, three dimensional (3D) images of the CTA. Research has shown that there is additional value in using such a system when studying small details and measuring structures in 3D datasets (*Bol Raap* et al., 2007).

The CTA demonstrated that there was abnormal calibre of the right internal carotid artery. This was diagnosed as a vascular dissection (Fig. 5) by the consultant radiologist. The left internal carotid artery was normal.

In hindsight, hypodense areas, probably old lacunar infarcts with parenchymal loss in the distribution of the right medial cerebral artery could already be seen on a CT scan carried out at 16 months of age, following the monobloc distraction, which unfortunately were previously unrecorded. These infarcts were not present on the first CT scan, prior to the monobloc.

Unfortunately, due to progressive extensive cerebral oedema the patient died 4 days after surgery. Postmortem investigation revealed no fracture of the base of the skull and no problems related to the external and internal distraction devices. No further exploration of the carotid arteries was undertaken.

## DISCUSSION

The Le Fort III osteotomy with distraction osteogenisis is a widely practiced technique to correct midface hypoplasia in patients with syndromal craniosynostosis, such as Apert, Crouzon and Pfeiffer syndrome. Various complications have been described with this procedure, but the incidence seems to be low and appears to be mostly related to the distraction devices and not to the osteotomy itself (*Nout* et al., 2006). Matsumoto et al. described a patient with Crouzon syndrome in which a Le Fort III distraction was performed with the positioning of a RED-II device in whom a lethal haemorrhage occurred from the right posterior maxillary region, most likely resulting

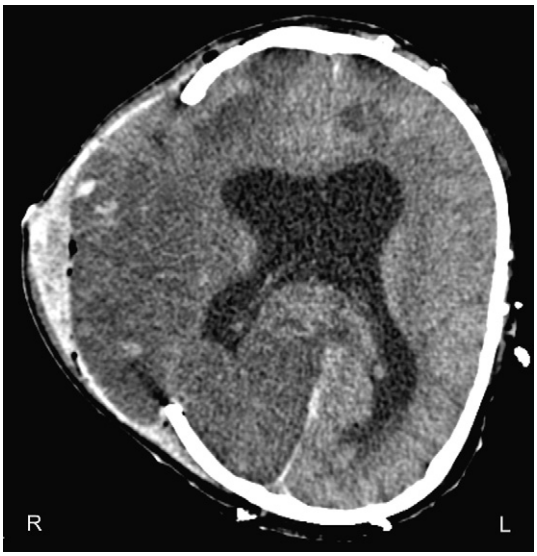


**Fig. 3** – The follow-up CT scan demonstrated an increasing hypodensity of the right hemisphere, suggestive for ischemia, an increase of the cerebral midline shift and a complete compression of the peripheral liquor space.

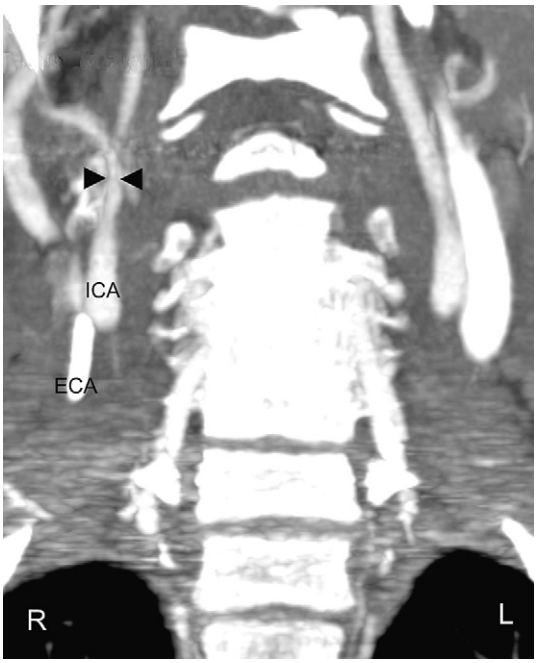


**Fig. 4** – CTA demonstrated a change in calibre of the right internal carotid artery (arrows). This finding is consistent with a dissection. The left internal carotid artery is normal. ICA = internal carotid artery; ECA = external carotid artery.

from a skull base fracture of the middle cranial fossa (*Matsumoto* et al., 2003). Pterygoid maxillary dysjunction and down fracture manipulation was suggested as the most important aetiological factor. These two factors probably result in a vector of force that pushes the pterygoid plates posteriorly. Subsequently, the force is transferred to the skull base through the sphenoid bone. In patients with congenital craniofacial anomalies, such as Apert, Crouzon and Pfeiffer syndrome, besides craniosynostosis and midface hypoplasia, an abnormal skull base can be found (*Tokumaru* et al., 1996).

When studying the pre-operative CT-scans surgeons should pay attention to the individual anatomical appearance of the skull base to enable them to carefully assess the approach to the pterygomaxillary separation. Several technical improvements of this maneuver have been described, but at present most surgeons probably perform the dysjunction with a thin slightly curved Le Fort osteotome. In our craniofacial centre, the correct position of the tip of the osteotome for dysjunction in the Le Fort III procedure is first checked intraorally with a finger. The osteotome is inserted from above, via the temporal fossa, and not from an intraoral approach. The surgeon feels the tip of the osteotome submucosally coming palatally through the bone somewhere around the junction of the hard and soft palate. With this antero-medial direction posterior–superior compression of the pterygoid process is avoided. An alternative approach, utilising a right-angled oscillating saw and an endoscopic approach for pterygomaxillary dysjunction has been described (*Chen* and *Fisher*, 1998). Another critical step and possible pitfall in performing a Le Fort III osteotomy in syndromal patients is the position of the anterior cranial fossa in relation to the nasal bone through which an osteotomy is to be planned. This close relationship is demonstrated on the axial slice of the pre-operative CT scan of an 8-year-old patient with Crouzon syndrome (Fig. 6).

In our centre one of the last surgical steps of Le Fort III osteotomy is the median osteotomy through the nasal septum starting from the osteotomy line through the nasofrontal suture pointing posterior–caudally towards the middle of the most posterior part of the maxilla. This vector must be corrected for an abnormal anteriorly located anterior cranial fossa.

In the patient we present a dramatic cascade of events followed a Le Fort III distraction, probably as a result of dissection of the right internal carotid artery as diagnosed from the CTA. To the best knowledge of the authors this complication has not been described previously.

Although most carotid artery dissections in children are spontaneous and are associated with considerable morbidity, in some case reports it is associated with a traumatic event (*Lequin* et al., 2004).

Keil et al. reported the case of an 8-year-old boy who suffered an internal carotid artery dissection following intraoral soft tissue trauma (*Keil* et al., 2006). He developed cerebral infarction in the vascular territory of the left middle cerebral artery. The patient survived after decompression hemicraniectomy with ICP measurements, but after 5 weeks the patient was still hemiparetic and aphasic.

Even chiropractic manipulation of the neck may cause a traumatic carotid dissection as has been described in an adult patient (*Gamer* et al., 2002; *Keil* et al., 2006).

A Le Fort III down fracture is a major soft tissue trauma event in the head and neck region. Given the monobloc distraction which was carried out 10 years earlier in our case the Le Fort III down fracture was probably even more traumatic, since the procedure was carried

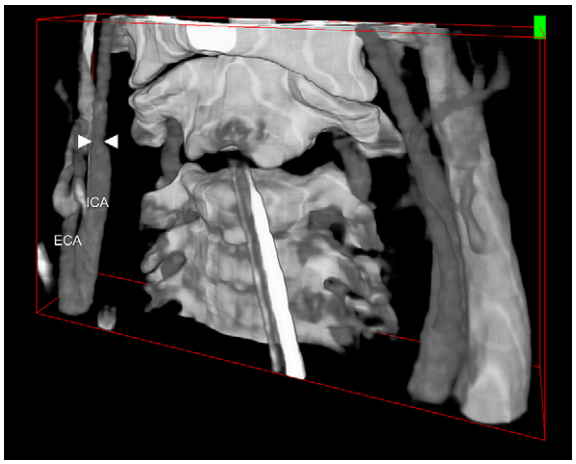


**Fig. 5** – 3D image of the CTA dataset demonstrating the calibre difference between the left and right carotid artery. A marked constriction in the right carotid artery is visible (arrows). ICA = internal carotid artery; ECA = external carotid artery.

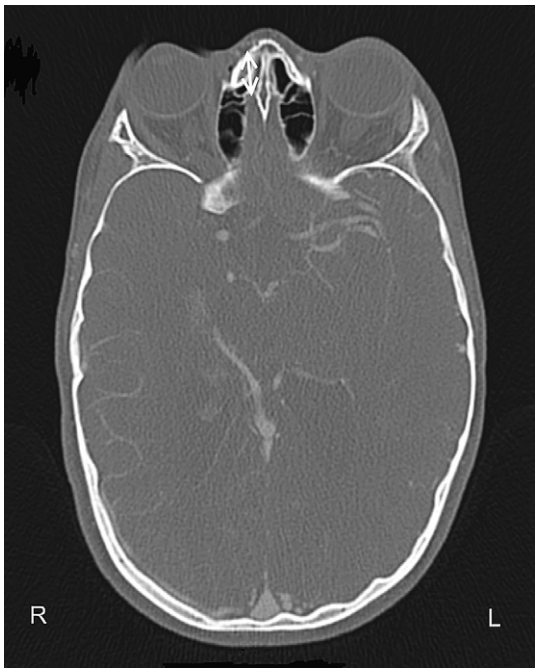


**Fig. 6** – CT scan of an 8-year-old patient with Crouzon syndrome with a relative short distance from the planned osteotomy line through the nasofrontal suture towards the anterior cranial fossa (arrow).

out in area of scar tissue. In addition, the first monobloc procedure seems to have caused a similar vascular incident, given the hypodensities on the postoperative CT scan. At that time, no postoperative neurological changes were noticed in the patient and thus no particular attention was paid to the abnormal findings on the scan. In hindsight, this should possibly have been considered as a warning sign of potential complications following further surgery.

Another possible aetiological co-factor for the complication in this case is that this type of surgery was performed in a patient with Apert syndrome. Marucci et al. analysed a group of 24 cases of Apert syndrome (*Marucci* et al., 2008). It was concluded that there was a high incidence of raised ICP, which can first occur at any age up to 5 years and may recur despite initial successful treatment. Causes of raised ICP include craniocerebral disproportion, venous hypertension, upper airway obstruction from midface hypoplasia, and hydrocephalus. In a retrospective study on 84 patients with Apert, Crouzon, or Pfeiffer syndrome, the prevalence of papilloedema, as a sign of raised ICP was found to be high, not only before cranial decompression but also after cranial vault expansion (*Bannink* et al., 2008). As a consequence, it should be borne in mind that the intracranial mechanisms in Apert syndrome to compensate for a relative minor raise of ICP, i.e. due to surgery, are probably limited, especially in case of a swelling or a relatively small hematoma. Other reports on Apert syndrome seem to concentrate on intracranial anomalies detected by imaging studies. Quintero-Rivera et al. found ventriculomegaly in 76% of the 30 patients with Apert syndrome (*Quintero-Rivera* et al., 2006). Dissection of the internal carotid artery in Apert syndrome has never been reported. At present all patients with syndromal craniosynostosis who are candidates for midface procedures in our craniofacial centre are analysed with CTA. We hope to gain more insight in the intra- and extracranial abnormalities in these complex patients. This will enable us to inform our patients more accurately about possible operative risks.

Finally, several reports have mentioned considerable problems using distraction devices, both internally and externally (*Le* et al., 2001; *Rieger* et al., 2001; *Matsumoto* et al., 2003; *Brown* et al., 2006; *Nout* et al., 2006). Accidental head injury and intracranial migration of halo pins have been described giving rise to serious complications. In addition, the quality of the bony skull in patients with craniofacial anomalies is often compromised due to earlier cranioplasties. In our case no problems with the distraction devices could be observed postmortem.

## CONCLUSION

The Le Fort III distraction is a major extracranial procedure and may result in devastating intracranial complications. For surgical planning pre-operative CTA is justified in order to identify possible vascular anomalies, especially in patients with Apert syndrome. Haemodynamic and neurosurgical parameters should be carefully monitored, with special emphasis on ICP, in order to guide the patient safely through surgery and the initial postoperative period. The management of these patients should be multidisciplinary and focused in specialised centres.

## CONFLICT OF INTEREST

This study has been carried out by the authors only. No external financial sources have been used. There are no relations that could be construed as a conflict of interest.

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