CRANIOFACIAL SURGERY OVER 30 YEARS IN GÖTEBORG

C. Lauritzen, P. Tarnow
Department of Plastic Surgery, Sahlgrenska University Hospital, Göteborg University, Göteborg, Sweden

ABSTRACT
Craniofacial surgery is a new sub-specialty in the field of plastic reconstructive surgery and is dedicated to the treatment of severe cranial and facial malformations. Craniofacial surgery gradually started in Göteborg in the late 1970’s and has been acting as the Scandinavian center since 1983.

Over these 30 years an almost complete change in surgical techniques has evolved. Also profound changes in timing of surgery have followed. Results have been dramatically improved based on critical evaluation of standardized registration of long-term results.

One of the most dramatic developments has been the introduction of implantable stainless steel springs. This has changed the treatment of Craniosynostosis completely and has made midfacial advancement procedures possible without relapse.

Key words: Craniofacial; craniosynostosis; cranioplasty; spring; Le Fort III

INTRODUCTION
In 1954 Ingraham and Matson in their textbook on pediatric neurosurgery for the first time introduced the concept of beauty, or at least cosmetic benefits as an indication for surgery of the cranium (1). At about the same time Sir Harold Gillies from England reported on a successful operation where the lower half of the face had been moved forward in an attempt to correct extreme malocclusion combined with shallow orbits, typical for Crouzon syndrome. In 1967 in Rome, the French plastic surgeon Paul Tessier at the IV International Congress of Plastic and Reconstructive Surgery presented to a spellbound audience his sensational results of treating Apert and Crouzon syndromes (2). Shortly thereafter he also published his dramatic results of correcting orbital hypertelorism by moving orbits together through a transcranial approach (3). In Sweden, the chief of plastic surgery at the Göteborg University, Bengt Johanson soon adopted the new techniques and introduced craniofacial surgery at his unit.

Fundamental aspects of craniofacial surgery entail wide exposure of the craniofacial skeleton often requiring a transcranial approach. This in turn calls for a qualified neurosurgeon. As craniofacial anomalies often also include maxillofacial malformations jaw surgeons were also needed. Special radiological examinations and neuro ophthalmological investigations form the basis for preoperative decisions especially regarding question of elevated intracranial pressure (ICP) etc.

Craniofacial malformations are very rare. In order for a team to be able to accumulate enough experience and dexterity Tessier originally claimed that the referral base of 10–15 million people was necessary for one craniofacial center. However, today simple synostoses are operated in national centres like in the cleft centre in Helsinki, Finland, and more complicated cases in cooperation with Göteborg.

In 1972 the first transcranial correction of orbital hypertelorism was undertaken in Göteborg together with Paul Tessier. From then on craniofacial surgery has continued to develop in Göteborg. For many years thereafter there was a consensus in Sweden and the neighboring Scandinavian countries that cranio-
Craniofacial surgery was to be concentrated to this one center. The result was that the Scandinavian experience of craniofacial surgery soon became considerable and achieved international recognition. In 2001 the International Society of Craniofacial surgery held its biennial congress in Visby under its Swedish president Claes Lauritzen.

The Scandinavian cooperation between Sweden, Iceland and Finland still remains with mutual benefit while Denmark and Norway have withdrawn.

THE FIRST STEPS

In the early years of craniofacial surgery techniques were so new and difficult that completion of the surgery was almost a goal in itself. Results were generally poor and patients were sometimes left in a worse condition than before surgery. This was the same for all start up craniofacial teams throughout the world. Very long procedures paved the way for infections with bone loss and bad sequelae. The use of alloplastic materials were unanimously condemned and reshaping of craniosynostotic skulls were always to be accomplished by letting skull bones be released and left unattached on the dura mater – the floating forehead. The idea was that the growing brain would resume a normal shape when the synostotic bones had been released (Fig. 1).

Craniofacial syndromes such as Apert, Crouzon, Pfeiffer and several others would now be treated by floating forehead cranioplasty plus midfacial advancement. The plan was to combine a dramatic change of appearance with improvement of upper airways. The typical surgical procedure for the midface advancement would be the LeFort III operation where the whole maxilla and zygomatic bones and nasal pyramid were separated with osteotomies from the skull and skull base and advanced forwards. This procedure in unskilled hands is dangerous and yields terrible results. On the international arena initially not more than 10–15 centers performed such surgery. Unfortunately very little scientific long-term follow up was ever presented by anyone. This was also true for the results of orbital movements, canthopexy and long-term survival of bone grafts. Most reports were anecdotal show and tells and of limited value.

Thanks to the many patients from all of Scandinavia it was possible in the Göteborg craniofacial center to be able to critically evaluate the results of large series of treated patients. From the outset cephalometry (X-ray with parallel beams) were performed at standard intervals with the patients asleep. Professional photography and team evaluation at the same time made critical analysis possible. Also at a very early stage 3-D reformatting of CT scans was available in the image-processing laboratory.

One of the biggest early problems was how to standardize methods. It was soon evident that the results of the floating forehead techniques were unsatisfactory and that midface advancements nearly always relapsed, making additional surgery both necessary and slow much more complicated. Orbital movements for hypertelorism relapsed and the canthopexies necessary for a good cosmetic result rarely remained perfect. Repeat surgery for bony irregularities or unacceptable shape of the forehead led to even more bone loss and more irregularities.

DEVELOPMENTS

A strong urge to improve results and techniques has meant that, if compared with the early days, practically every procedure today is different. The preferred age for surgery for various procedures is now based on experience. Long-term plans are usually available for any child born with a craniofacial malformation or syndrome, which makes it possible to make family counseling adequate and thereby consoling.

Certain basic features of the craniofacial surgery remain unchanged despite all other modifications,

- No shaving of the head is done even if an intracranial procedure is planned
- Wide exposure is obtained by coronal, subciliary and/or buccal incisions, leaving no visible scar in the face
- Coronal incisions are made zigzag in order for the hair to not part along the scar
- Alloplastic bone substitutes are frequently used.

At the end of 1980’s a modification of the John Jane’s so called Pt-plasty was introduced for the treatment
of sagittal synostosis i.e. the premature fusion of the sagittal suture, leading to a head shape often referred to as scaphocephaly (boat shaped skull). With the PI-plasty the length of the skull was reduced by active compression while the parietal bones were completely released from the dura mater and green-stick fractured to obtain a rounded shape. The procedure is extensive but yields excellent results and was usually performed at 6 months of age.

The PI-procedure meant a start of new ‘dynamic’ thinking for skull reshaping as opposed to the static (rigid) or ‘floating’ procedures (Fig. 2). Soon thereafter the dynamic cranioplasty for brachycephaly (DCB) was developed by us and introduced for the treatment of the short and wide skull resulting from a premature synostosis of both coronal sutures (4) (Fig. 3). Both the PI- and the DCB procedures yield excellent results, but are limited by the extent of com-
pression that can be allowed on the brain during surgery. Both procedures are also quite extensive. Premature fusion of the metopic suture, leading to the so called trigonocephaly (triangular head) has for the last 10 years been treated by using osteotomies, widening the skull and the forehead and inserting a bone graft between the orbits to help these separate during the subsequent growth. The procedure yields good results. The preferred age for surgery is approximately 6 months and older.

SPRINGS – A REVOLUTIONARY DEVELOPMENT IN CRANIOSYNOSTOSIS SURGERY

Searching for smaller surgery and other means of gently compressing the skull bones into a normal shape after surgical release eventually led to the development of springs. In 1998 the first report on the clinical use of springs in 4 cases was published (5). This was a fundamental new step in the development of lesser surgery and better results for our patients.

SAGITTAL SYNOSTOSIS

Spring assisted cranioplasty for sagittal synostosis is started by making an S-shaped incision above the sagittal suture. A strip craniectomy is performed between the anterior and posterior fontanel and 2 springs across the osteotomy are inserted in a bayonet fashion. This mode of application facilitates spring removal at a later stage, usually 6 months postoperatively. The results of spring assisted strip craniectomies for sagittal synostosis are such that no other technique can be considered provided the child is referred in time. The preferred age for this surgery is 3 months (Fig. 4).

BICORONAL SYNOSTOSIS

As previously mentioned the bicoronal synostosis, leading to brachycephaly (short skull) can be treated with the DCB with good results. The idea of using springs was originally intended for use in the DCB instead of compressing wires. In 4 cases so was done and the results were truly remarkable. It appeared however that this type of compression would create a too high ICP over a longer time and could not be used. Instead a modification of the DCB was developed where springs were placed to push the forehead anteriorly and distend the unoperated normal lambdoid sutures posteriorly. Distending normal sutures in this way has not been undertaken in craniofacial surgery before and is still not done except at our unit where this today is a routine procedure (Fig. 5).
METOPIC SYNOSTOSIS

Surgery for the premature fusion of the metopic suture is performed in a similar way as surgery for the sagittal suture with the addition of a couple of semi-circular concentric osteotomies in the frontal bones. Exposure requires a coronal incision, but the procedure is in every other aspect much smaller than previous techniques. The desired timing of surgery is at 3 months (Fig. 6).

PAN-SYNOSTOSIS

Certain craniofacial dysostosis syndromes, notably Pfeiffer and Crouzon can at birth present with a pan-synostotic skull, where most sutures of the skull are fused. The anterior fontanel and part of the metopic suture are in these cases forced open and the brain shoots up in this location. As both lambdoid sutures are fused there is a strong limitation of room in the posterior cerebral fossa, which creates herniation of the cerebellar tonsils into the greater foramen. This has an effect on tongue movements and breathing. The crowded foramen compromises CSF drainage (hydrocephalus) and the situation is life threatening.

Surgery for this kind of situation is difficult as the immature bone is irregular (copper beaten) from the elevated pressure. The dura mater and the cambium layer of the calvaria are not fully separated making dissection difficult.

Springs have been used in 8 cases in this situation to straddle a simple strip craniectomy with remarkable success and surgery has been adequate and virtually risk free. No other means of operating infants in this situation can be considered today. The ideal age of surgery depends on the condition of the child. A VP shunt is invariably inserted and this buys some extra time, but not longer than 3-4 months (Fig. 7).

SPRINGS – A REVOLUTIONARY DEVELOPMENT IN MIDFACE ADVANCEMENT SURGERY ALSO

As mentioned above midface advancement for improvement of airways and facial esthetics was one of the really new procedures presented by Paul Tessier. Results could be dramatic, but for most surgeons not stable and more or less total relapse could be ex-
Craniofacial surgery over 30 years in Göteborg

expected. For several years these procedures were therefore abandoned in Göteborg and upper airway problems were managed nonsurgically employing the CPAP (Continuous Positive Airway Pressure) mask. This is placed over the nose and/or mouth delivering air under slightly increased pressure and can be extremely helpful in overcoming sleep apnea often seen in children with craniofacial dysostosis syndromes (retruded midface).

In 1997 in a 5-year-old boy with Apert syndrome from a country where neither a CPAP mask could be provided nor a tracheostomy handled a LeFort III procedure was undertaken. In addition 2 strong springs were inserted to keep advancing the face after surgery. Careful cephalometric follow up demonstrated that although no further advancement of the midface was achieved, there was no relapse either, which was sensational. This prompted the surgery of 12 more cases. Therefore midface advancement has again become standard procedure in certain situations, but always together with spring suspension (Fig. 8).

THE SPECTACLES PLASTY

For the same type of cases where no imminent airway obstruction was present and when the exposure of the protruding eyes could be tolerated from a mechanical point of view the so-called Spectacles plasty was introduced in 1983. This surgical technique is based on the idea that for good correction of the malformed face the periorbital bony structures needed transposition in one direction and magnitude, while the rest of the face required different change in a different direction. Thus a staged procedure was introduced where the first step involved osteotomies of the calvaria and the orbits. After healing and finished orthodontic treatment the maxilla would then be operated with a LeFort I operation (separation of the maxilla through the maxillary sinuses). With the addition of bone grafts the short face could be elongated without enlarging the orbits or lengthening of the nose which otherwise would be the case if the problem was to be solved with a LeFort III procedure. By separately moving the maxilla perfect dental occlusion could be achieved. With these techniques it became possible in counseling families with a Crouzon child that if we would be allowed to wait until the early teens, possibly with the aid of the CPAP mask there existed a realistic hope that all trace of the severe facial handicap could be eradicated. Usually these children would need cranial expansion before that time, but in performing that surgery the orbits would not be touched in order to preserve these intact for later already planned surgery.

DISCUSSION

As can be deducted from this review craniofacial surgery has undergone significant development over the last 30 years. This affects every part of the work and cannot be covered in detail in one article only. Two major factors affecting the whole spectrum of craniofacial surgery over the last 30 years can be distinguished however. These two factors are timing of surgery and the introduction of springs.

*Fig. 8. 7-year-old girl with Apert syndrome. Monobloc advancement is performed and supported with springs. Upper row is the preoperative appearance. Lower row 6 months postoperatively. Cephalogram is superimposed on the lower lateral photograph to illustrate position of supporting springs, preventing relapse.*

Photo Copyright: C. Lauritzen
It may seem that the optimal time for surgery would be an easy thing to decide, but that is not always so since at least 10 or even 15 years need to pass until it is possible to find out if surgery was effective, adequate, or even harmful. The modifications that may have been prompted also need long time for evaluation.

Today, compared with the situation 20 years back, the decision on timing of surgery is easier and based on solid experience rather than guessing. We have now followed hundreds of operated children growing up. A family with a newborn baby with a craniofacial syndrome can now be counseled in a better and more comforting way, since we know rather well what is lying ahead and since the prognosis can be determined with reasonable adequacy.

Neither all parents nor all referring doctors do always realize that treatment of a severe craniofacial handicap rarely can be completed in one session only. Naturally parents need in depth information and usually need time to fully understand the impact of the new situation. Mobbing in school is one of the most dreaded expected consequences by most parents. On the other hand, if the parents know that through staged procedures a nearly complete eradication of the handicap may be within reach, problems will be more tolerable.

The introduction of springs in craniofacial surgery in 1997 meant a revolution of the craniofacial surgery at our unit. (This was even acknowledged by Paul Tessier himself in a letter to the senior author, where he described it as a fundamental step in craniofacial surgery). It was suddenly possible to perform considerably smaller operations to reach the same goal. For most of the symmetric craniosynostosis patients complete normalization can be expected with a minimum of blood loss and hospital stay.

The discouraging results after midface advancements with expected relapse for many years made this type of surgery abandoned at our unit. Other means of supporting these children were used, especially the CPAP mask, but eventually some sort of means of supporting these children were used, especially the CPAP mask, but eventually some sort of means of supporting these children were used, especially the CPAP mask, but eventually some sort of means of supporting these children were used, especially the CPAP mask, but eventually some sort of means of supporting these children were used. The change was brought by the one case of the Apert child who had at 4 years of age never been able to leave the hospital for his severe sleep apnea. After his monobloc advancement (i.e. LeFort III plus the orbits and forehead) it appeared that with the help of 2 strong springs there was no relapse and the boy could be sent to his home. The expected spring mediated further facial advancement was never however, but the fact that the problem of relapse could be overcome was a sensational development.

 Springs are usually visible under the skin and are electively removed at various times depending on their function. In our laboratory research is directed at developing springs that do not need to be removed.

PROBLEMS

SCANDINAVIAN COOPERATION

Over the last 3 years several attempts to reestablish the Scandinavian cooperation in craniofacial surgery has been made. Regrettably these attempts have failed. Today cooperation still exists between Iceland, Finland and Sweden with mutual benefit and this has been the case since more than 25 years. Denmark 2 hospitals admit patients for craniofacial surgery and in Norway political decisions block Norwegian patients access to treatment in Coteborg. The reasons for this are not clear but certainly not economical as hundreds of patients each year are sent to Sweden for abdolimnoplasties and breast reductions.

CRANIOFACIAL SURGERY AT DEDICATED CENTERS

Time after time centers with limited experience undertake craniofacial cases. This is objectionable on several grounds and undermines the possibilities for long term follow up analysis, the chance for continuation of training of younger surgeons at craniofacial centers and much more. Appalling examples of this appear from time to time and entail severe injustice to the families.

DIAGNOSIS IN TIME

With the discovery that the risk of SIDS (sudden infant death syndrome) could be reduced if newborns were to sleep in the supine position has led to an enormous increase in skull deformations. The reason for this is that the tradition of supine sleeping position was abandoned a generation ago and therefore knowledge was lost of how to protect these soft skulls from deformation (soft pillows, repositioning of the heads etc.), but the deformation usually had a good prognosis if properly handled. However, since deformed skulls in newborns have become so common children with craniosynostosis tend to pass undetected until much later than before (6). This is unfortunate.

REFERENCES


Received: July 21, 2003