

■ Craniofacial Surgery (*Headlines factsheet*)

■ Introduction

Surgery for craniofacial conditions may be indicated for a variety of reasons. Sometimes it is necessary to correct a functional problem, e.g., to make more space inside the skull for the brain to grow if the pressure here is raised, to provide better protection for the eyes when they are too prominent or to make breathing and/or feeding easier. On other occasions, however, the surgery will be indicated for purely cosmetic reasons. This is not to belittle its importance – but in this situation there is a greater degree of "choice" on behalf of the family as to whether the surgery should or should not be done. This may require careful thought on the part of the parents before coming to a decision. Clearly when there is a severe functional problem these decisions are much easier to make.

■ Surgical procedures

■ Surgical correction of sagittal synostosis

Surgical treatment of this condition varies depending on the age at which the patient is first seen. If the child is very young, i.e. up to 6 months, the bones of the skull are still relatively soft and pliable and the operation can be designed to bend or mould the bones of the skull into a more normal shape. The bones are usually held in their new position with absorbable sutures rather than wires, plates or screws. These sutures take some time to dissolve and will remain in place until the bones are "healed" in their new position. This operation is a relatively small procedure compared with most other craniofacial operations and the child will usually be in hospital for 4 or 5 days.

When treatment is carried out in an older age group then the surgery becomes considerably more major. After 6 months of age it is no longer possible to mould the bones of the skull since they have become much more brittle. It is necessary to remove a large area of bone in order to reshape it or reposition it so that the overall skull shape will be more normal. The exact requirements of any particular individual, of course, will vary but in general this is a relatively major operation. Bones are fixed in their new position usually with wire sutures which remain in position and are not removed unless they cause problems, e.g., are felt through the skin. Occasionally it may be difficult to stabilise the bones in their new position with wires, in which case small metal plates and screws will be used. Patients will usually be in hospital for 7 to 10 days.

■ Frontal remodeling/advancement

Most craniosynostoses which affect the front part of the head are treated by a variety of frontal (forehead) remodeling or advancement procedures. These conditions include metopic synostosis (*trigonocephaly*), unicoronal synostosis (*plagiocephaly*) and bicoronal synostosis (*brachycephaly*).

Generally the surgery is carried out at approximately the age of 10 – 14 months. This may vary considerably if the condition is part of a more generalised syndrome. We have found that when these operations are carried out earlier than 10 months or so then the children tend to "grow out of them" relatively soon afterwards which necessitates a further operation later on. Around about a year seems to be the best compromise between stability of the operation and waiting too long which would necessitate the use of bone grafts to fill gaps behind the remodeled frontal region.

This operation will usually involve moving not only the forehead but also the upper part of the eye sockets (*Corbits*) and reshaping them. In most cases the bone of the forehead is removed completely from the skull and reshaped into a more normal form. The upper part of the eye sockets are released from their attachments and again shaped into a more normal configuration, moved into the position which they should occupy and fixed there using either fine wires or occasionally small plates and screws. The bone of the forehead is then reattached to the upper part of the eye sockets, again with wires, plates or screws. Usually a small gap will be left behind this deliberately to allow further growth to occur as the brain expands before the new bone which will be produced closes the gap. The time allow for this to happen varies but is usually in the region of 6 to

12 months.

Facial advancement

Children who have what are known as craniofacial dysostosis syndromes, e.g. Crouzon syndrome, Apert syndrome, Saethre Chotzen syndrome, Pfeiffer syndrome, Carpenter syndrome, etc., will often require surgery to remove their facial bones forwards because growth of the face in these conditions is not normal (they now, of course, have already undergone an operation on their skull for craniosynostosis).

The timing of the facial surgery is a matter of great debate and will vary considerably from one patient to another and one unit to another. In general, in an ideal world, most of these operations are best left until later adolescence. At this stage growth of the face is more or less complete (16 to 17 years) and we can be reasonably sure that the bones will stay in the position in which we fix them. However, there are frequently circumstances which will dictate that the surgery should be carried out sooner. This may be to correct a functional problem such as feeding or dislocation of the eyes, or it may be to improve the patient's appearance as they grow up, particularly during the difficult phase of adolescence. When the surgery is carried out before growth is complete it is very frequently necessary to carry out a further operation at the end of the adolescent period in order to position the teeth correctly.

Operations to remove the facial bones forward come under the broad heading of *LeFort osteotomies*. Rene LeFort is the name of the French doctor who described a pattern of fractures following injury from which these operations have been devised. The "LeFort" operations are numbered according to the level in the face at which they are done. The LeFort I osteotomy moves the tooth bearing part of the upper jaw only, a LeFort II advancement moves the tooth bearing part of the upper jaw and the nose and the LeFort III advancement moves the whole of the upper jaw, nose, cheek bones and eye sockets forwards. Sometimes a LeFort III type osteotomy is extended into the skull to move the forehead region forward at the same time and this is known as a *"monobloc"* advancement. In some instances when the monobloc advancement is done the face is deliberately split vertically along the line of the nasal bridge in order to bring the eyes closer together, expand the upper jaw and rotate the two halves of the face into a more normal position. This latter procedure is usually reserved for more severe "syndromic" conditions such as Apert syndrome and Pfeiffer syndrome. The procedure is referred to as a *facial bi-partition*.

Generally, before most "LeFort type" osteotomies there is a period of orthodontic preparation in order to put the teeth in the most suitable position for the operation. This generally takes in the region of a year to complete and there is likely to be a period of orthodontics following the surgery also. This is only appropriate in older children who have their permanent adult teeth.

The state of development of the teeth has a significant influence on the timing of these operations. There are "windows" of time which are more suitable than others which generally are at around about the ages of 4, 8, 12 and then the end of development, i.e. 16 to 17 years. General dental care and keeping the teeth in good condition is extremely important to the successful completion of these operations and in some cases considerable dental preparation other than orthodontics may be required.

When the facial bones have been moved they are usually fixed in position by a combination of bone grafts and titanium screws, possibly assisted with small titanium plates. The bone which is used for grafting is usually taken from the hip but sometimes it may be possible to use bone from the skull.

Often the teeth of the upper and lower jaw are fixed together at the time of surgery in order to locate the position of the facial bones accurately and in most instances this fixation will be taken off at the end of the operation so the jaws can move. However, during the recovery period sometimes elastic bands are put between splints on the upper and lower jaw. The orthodontist will manufacture and fix these splints in position before surgery is carried out. Oral hygiene is extremely important both during the period of preparation and post-operative recovery. The hygienist will assist with this but the enthusiastic co-operation of the patient is essential.

Bone grafting

Many craniofacial operations involve taking bone from one place and using it for support or to fill gaps in another place. This is known as *bone grafting*. The patient's own bone is by far the most suitable material for this and certainly much more reliable than any artificial (*prosthetic*) material. It will usually remain in position long term and grow and it has relatively low risk of infection. The most common sites to take bone from for use as a graft are from the skull and the hip. Occasionally the ribs or the tibia are used. When bone is taken from the hip it will of course leave a small scar which is usually about 10 cms long. The hip is usually quite sore afterwards and walking is likely to be uncomfortable for a few weeks depending on the size of the graft which has been taken. In the early time after the operation, a small tube is usually left at the site from which the bone has been taken in to which local anaesthetic can be put to relieve pain. There should be no permanent change either in the appearance of the hip or function of the leg. The gap left from taking of the bone graft will not fill in but surrounding bones are strong enough to provide the necessary support.

In some circumstances it may be preferable to use bone from the skull. In most individuals between the ages of approximately 8 and 50 years the skull bone is, in fact, in two layers known as an inner and outer "table". It is therefore possible to use one layer (usually the outer table) as a bone graft leaving the other layer undisturbed so that no gaps are left in the head. Unfortunately in many children with syndromic craniofacial conditions the usual two layers of the skull do not exist which may make it impossible to use this site for bone grafting.

Bone distraction

Bone distraction is a relatively new technique for the craniofacial Skeleton. It has been developed from limb lengthening operations described by a Russian called Ilizarov. It uses a principle of small pins pushed in the bone which are then moved apart by a connecting bar which incorporates a screw thread. This will, of course, stretch not only bone but also soft tissue as it is moved.

The most frequent application for this technique has been in the *mandible (lower jaw)* for conditions such as hemifacial microsomia, but it is also now being applied to other areas of the facial skeleton such as cheek bones and eye sockets.

Usually two operations are required. In the first operation a cut is made in the bone which is to be stretched, this is known as a *corticotomy*. The pins are passed through the skin and in to the bone on either side of cut. The pins are then connected together by the "*distractor*". The distractor is opened usually approximately by 1mm – 2mm each day by turning a small screw. (It is possible for the parents to be shown how to do this so the procedure can be done at home). An average distraction length is likely to be in the region of two to two and a half cms which will probably therefore take 2 to 3 weeks in total. At the end of the period of distraction the distractor is left in position for 6 to 8 weeks whilst new bone fills the gap which has been created. A second small operation is then required to remove the distractor.

Presently most of these devices are fixed externally. This has the advantage of making them relatively simple to use but the disadvantage that there is some inevitable facial scarring from the site of the pins. A great deal of work is being pursued to develop distractors which can be placed inside the mouth and so reduce or eliminate any resultant facial scar.

Ancillary procedures

For some craniofacial conditions the problems may be solved by a single operation. However, in many, particularly the more complex ones, there may be one or more major operation with smaller procedures being needed towards the end of growth (i.e. 16 to 17 years) to achieve the best possible end result. Common among these are *rhinoplasty (nasal surgery)*, *genioplasty (surgery to the chin)* and *scar revision*.

Rhinoplasty

The aims of surgery will of course be discussed in detail before it is carried out and do vary enormously from one patient to another. This is a common procedure which may be carried out either to assist with

breathing through the nose or to improve the shape of the nose or more commonly both. It is usually done from incisions inside the nose so it leaves no scars on the outside, though this may not always be the case for children with craniofacial problems. In some patients it may be necessary to make an incision on the nasal bridge, particularly when there is an excess of skin in the midline. In a number of children a bone graft may be needed to support the nasal bridge. Nasal passages are widened when necessary and the shape of the bone and the cartilage which supports the nose is altered to provide a pleasing nasal shape. After surgery a splint, which may be of either a plastic material or plaster of Paris, is placed on the nose to support it and left in position for a week or so. Sometimes packs are put inside the nose for 24 hours to help the lining skin adapt to its new shape. In some cases small tubes made of a soft rubber material may be left in the nose when it is necessary to splint it for a prolonged period, usually 5 to 6 days, since the patient will be able to breathe through the tubes.

Genioplasty

This is the name used to describe any surgery to the chin which does not involve the tooth bearing part of the lower jaw. It may be used to move the chin forwards, backwards, upwards or downwards. It is usually done through an incision inside the mouth where the lower lip joins the lower gum. The bone of the chin is cut horizontally below the level of the tooth roots so as not to damage them, moved in to its new position and fixed with either wires or more usually titanium screws. These remain in place and it is not necessary to remove them. The incision is closed with dissolving sutures.

Sometimes some numbness may be experienced on one or other side of the lower lip for a few days after the operation because of bruising of the nerves in the lower region of the chin. This would be expected to recover spontaneously in the majority of instances. It is important to use an antiseptic mouthwash during the recovery period and to avoid the use of a toothbrush for the lower teeth for a week or so until the healing of the gum has occurred.

Scar revision

Scars from operations which have been carried out in childhood may stretch with growth and an improvement in their appearance may be achieved by an operation known as scar revision. Usually this entails removing the existing scar and creating a new one, hopefully of better quality. Whilst this is, of course, usually successful, it is impossible to guarantee the outcome of a scar in any given individual and there is always a small risk that it may stretch again.

Common worries

Swelling and discomfort

Surprisingly, operations on the face and skull are usually accompanied by very little pain. Swelling will settle in a variable length of time but in most cases the majority will go during the first 2 to 4 weeks. However, it will take up to 6 to 12 months for all the swelling to settle down and for a final result to be appreciated.

Incision

Most craniofacial operations involve an incision known as bicoronal incision which is like an Alice band running from the top of one ear to the top of the other ear across the scalp. The incision line may be straight or may be cut in a zigzag pattern depending on individual patients and the surgeon's preferences.

Frequently after the bones of the skull have been moved or reshaped, gaps are left between bone edges. In young children, i.e. under 1 year, new bone will usually form in these gaps over a period of months. In older children, however, gaps will not fill with bone and so bone grafts are usually used to fill them in at the time of surgery. In most instances it is possible to find enough bone for grafting from elsewhere in the skull but occasionally it may be necessary to take further bone from the hip in order to fill a gap. In operations designed to move the facial bones forward, bone grafts will always be used to support the face in its new

position since the gaps which are created will never fill in spontaneously.

Closure of the skin of the scalp is usually done with small clips since these do not damage the hair follicles as much as stitches may. Nevertheless there may be some circumstances in which stitches are preferred. Dressings are sometimes used but again this will vary depending on the particular circumstances of the operation. Usually a small plastic drainage tube will be left beneath the skin of the scalp to remove any fluid or blood which may accumulate, it is taken out after 24 – 48 hours.

Complications

Complications following craniofacial surgery are uncommon but as with any operation, not unknown. Clearly the greater the magnitude of the surgery the higher the complication risk but, nevertheless, statistically even for the most complex craniofacial procedures severe complications are very rare. Those which concern us most are:

(1) Risk of bleeding – This is certainly a very real concern in some craniofacial conditions, particularly where the pressure inside the skull may be raised. However, it is exceedingly unusual for it to be a level which cannot be managed by the surgeon and anaesthetist. Most children undergoing craniofacial surgery will require a blood transfusion though some will not and the volume of blood transfused varies greatly.

(2) Infection – Infection following most craniofacial procedures is very rare as with other operations in the head, neck and facial area. Most children undergoing craniofacial operations will be given antibiotics as a matter of routine as a precaution against infection. When infection does occur, it can usually be treated quickly and efficiently by appropriate antibiotics. Infection is particularly a concern in combined operations involving the head and face where it may be impossible to avoid creating a communication between the nasal cavity and the cavity inside the skull, since the roof of the nose and floor of the skull are essentially the same structure. Of course, every effort is made at the time of the operation to ensure that communications such as this are sealed to reduce the risk of infection as much as possible. Clearly if there is an infection around the brain this is more serious than infection elsewhere. It is known as meningitis though not the same sort of meningitis that one reads about in the newspapers occurring in "epidemic" outbreaks.

(3) Anaesthesia – Anaesthesia for craniofacial operations, particularly the more major ones, is extremely complex and it is most important that the anaesthetists are particularly skilled and trained in surgery of this sort. In the hands of an expert, anaesthesia for this type of surgery is extremely safe. However, this is one of the many reasons that it is important that a child with a craniofacial problem is cared for within a craniofacial centre where all the staff, not just the surgeons, are experienced in the treatment of these conditions.

Other minor complications may occur which can arise after any type of operation. For instance:

Haematoma – a collection of blood or fluid beneath the skin which is usually treated simply by drainage.

Slow wound healing – which may be related to tension on a wound or simply reduced supply of blood to the region of an incision for one reason or another.

Minor wound infections - such as infections around stitches may occasionally arise and are treated by simple dressings and antibiotics. *Scars* in the scalp may stretch as with scars in any other site. Some surgeons believe that making an incision in the shape of a zigzag rather than a straight line make the scars less noticeable because the hair will "fall" more naturally.

Fits, similar to epilepsy, are an exceedingly rare later complication of any operation on or near the brain. Should this occur they would be treated with medication and monitored.

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