



# **Headlines**

## **Craniofacial Support**

### **No 5**

# **The Surgical Treatment of Hand Anomalies**

**associated with**

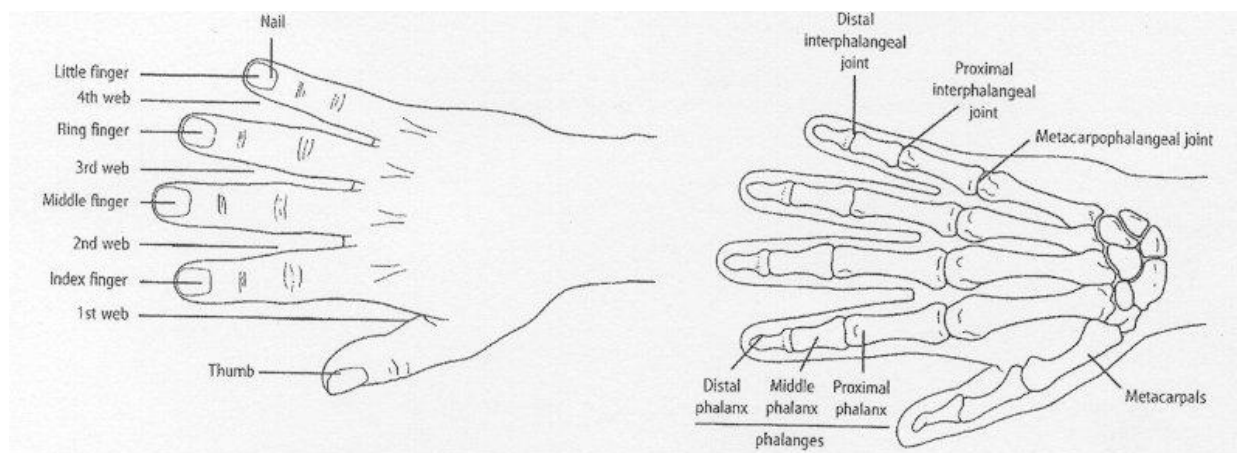
# **Craniofacial Conditions**

**April 2002**

**Reg Charity No 1058461**  
**[www.headlines.org.uk](http://www.headlines.org.uk)**

## Introduction

This leaflet contains information on the actual hand anomalies associated with some craniofacial conditions. It also describes the surgical treatment undertaken to improve the function of the hand.



## Apert Syndrome

Anomalies seen:- Syndactyly of the hands and deviation (curving outwards) of the thumb.

**Deviation of the thumb** is caused by a broad, short, triangular shaped bone. You may hear it referred to as a *delta phalanx*. This causes mal-alignment of the joint surfaces and gives rise to the thumb deviating towards the wrist. This hinders function, as a straight thumb is required to allow pinch with the other digits (fingers) to provide a strong grip.

**Syndactyly** is the failure of two or more neighbouring digits to separate. This may be simple or complex.

**Simple** - where only the soft tissue is joined, with the bones to the neighbouring digits being uninvolved. Simple syndactyly may be **complete** to the tips of the involved digits, or **incomplete** where fusion stops before the tip of the digits. This type commonly involves the thumb and little finger.

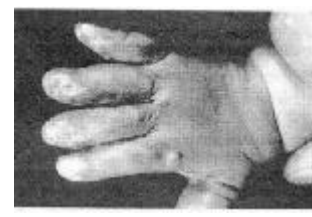
**Complex** - the complex form consists of bony fusion as well as soft tissue, and commonly involves the index, middle and ring fingers (central digital mass).

The Apert's hand can be classified into three types, depending on the severity and complexity of the structures of the hand.

### Type 1

This is sometimes referred to as "**spade**" like or "**paddle**" hand.

In this arrangement the thumb is separated from the central digital mass by a shallow web space. The central mass is flat and the digits (fingers) lie side by side. It is often possible to see the definition (outline) of the individual digit borders (edges). Bony fusion, if any, usually occurs at the distal phalanx (small bone at the end of finger) level. Although partially fused at this level, the individual digits have their own distal phalanx. Simple syndactyly may occur in the fourth web space (space between the ring and little finger, see diagram). The proximal interphalangeal joint (joint between the bone of the finger nearest the hand and the middle bone, see diagram) may be present in the little finger and is the only interphalangeal joint in the hand which moves. It does not occur in the other digits due to *symphalangism* (stiffness) of the joints.



Type 1, before surgery

Type 1, after surgery

## Type 2

This is sometimes referred to as the "mitten".

The thumb is syndactylised to the central digital mass in the simple form. The more severe bony union of the central mass at the distal phalangeal level, causes the bones in the palm and the base of the fingers to splay out, which results in a large concave (scoop like) palm. Simple syndactyly seen in the fourth web space is usually complete.



Type 2, before surgery



Type 2, after surgery



## Type 3

This is sometimes referred to as the "rosebud".

Most severe of the hand types. The thumb is joined to the fingers in a bony or cartilaginous (strong tissue) union. The nails are usually joined. The little finger is joined to the rest of the hand with a simple complete syndactyly. It is interesting to note that, not only do the structures and appearance of the hands differ between each child, but may also from left to right hand.



Type 3, before surgery



Type 3, after surgery

## When to operate

Due to the knowledge and experience now available in plastic surgery, children with craniofacial conditions face many procedures for their craniofacial anomalies. Therefore their main priority of hand surgery is to provide the child with as fully functional a hand in one operation as possible. Commencement of surgery on the child's hand can be carried out from four months of age. Timing obviously depends on the child's overall condition in regards to intracranial pressure, their airway and size. The younger the child, the more likely the surgery is to be carried out on both hands at once. If surgery is performed on the hands separately, operations can be carried out at approximately three-monthly intervals.

## Surgery undertaken

Surgical approach to these hands obviously differs from surgeon to surgeon, but their main aim is for the child to have good functioning hands, with as many separated digits as possible.

A one stage approach can be undertaken in types "1 and 2", consisting of release of 1st, 2nd and 4th web spaces and *osteotomy* (cutting of the bone and realignment of the two ends) of the thumb.

The 1st web space is released by a rotation flap (a piece of tissue still containing its blood and nerve supply which is partially removed and turned and placed in a new position) combined with a *z-plasty* (z shaped cuts in skin). The small flaps produced with zig-zag shaped incisions are rearranged and sutured together. This type of incision prevents contracture deformities, potentially produced by scar tissue of straight line incisions. A curved (closed) or (open) wedge *osteotomy* is performed on the thumb, depending on the surgeon concerned. This allows straightening of the thumb by realignment of the phalanges and joint surfaces in the thumb. The bones in the thumb are held together in a straight and stable position to allow healing, by a *K-wire* (K stands for Kirschner, the man who invented it). This wire remains in the thumb for approximately six weeks, when it usually becomes loose and is removed.

The 2nd and 4th web spaces are released simultaneously.

The 2nd web space can be released using a straight line incision due to the presence of symphalangism in the central digital mass proximal interphalangeal joints. As the digits are stiff and do not bend, due to the bony and joint deformities, there is no worry of a straight-line incision scar contracture deformity. A full-thickness skin graft from the groin is used to cover the raw defects on either side of the web space. This is necessary due to the deficiency of skin available, as the circumference of two separated digits is more than joined ones. The incision lines in the groin becomes less visible as they mature and are in a natural skin crease. The base of the web space is created and lined using "V" shaped flaps from the front and back of the web.

The 4th web space, as it often has a functional proximal interphalangeal joint, is released using zig-zag incisions as in the 1st web space, for the same reasons.

Due to the complex abnormal structures seen in these hands, usually only one side of a digit is released at a time. This is to preserve the digit, as the blood supply to the digit may be impaired (altered or lost). Just because the child at two years old only has four digits does not mean by the age of five they will not have five.

The 3rd web space may be released, depending on the bony structure and size, at the age of three to five. Waiting allows the bones to grow making separation, if possible, easier and more successful. Unless evidently possible both clinically and radiologically, too early release of the 3rd web space may lead to one of the digits being abnormally thin and weak, and becoming more of a hindrance than a help. It is obviously very important for the children to obtain five-digit hands if at all possible, as it helps them to have more normal hand appearance like their peers.

In type 3 the achievement of a functional hand is a longer process. Often initial separation of the nail beds is required to alleviate the occurrence of nail sepsis (infection). The thumb is then usually separated from the rest of the digits and a skin graft is required. The release of the other web spaces is then usually undertaken as in types "1 and 2".

### **After surgery**

After surgery the hands are covered in large bulky dressings, so loose fitting, sleeved clothes such as tee-shirts or sleeveless garments make dressing and undressing easier. It also prevents other clothes needing to be cut up.

For the first 48 hours the hand/ hands should be kept above the level of the heart, day and night, and then during the day for another 3-5 days. This helps to reduce pain, prevent swelling and reduce bleeding as blood finds it more difficult to travel uphill. Bleeding under the skin grafts can lead to loss of graft take, which may require further grafting, or if left spontaneous healing will be prolonged.

Two weeks following surgery the dressing is changed under a general anaesthetic due to fragility of the skin grafts. Once the grafts have become stable the dressings are performed on the ward, at one to two weekly intervals until completely healed. The hand takes approximately 4-6 weeks to heal. Once the web spaces have healed the incision lines should be massaged twice a day until the scars have matured at 12-18 months, with moisturising cream **without** Vitamin E (which alters the scar healing process), such as Nivea. A small amount of cream is applied to the scar and massaged in using small circular movements. This helps to soften and flatten the scar and produce a good final result. The child is then seen by a physiotherapist/ occupational therapist for measurement of a pressure garment (glove) which helps flatten the scars and maintain the depth of the web space achieved at surgery. The gloves are worn for 23 out of 24 hours a day. The children adapt to these gloves very quickly, and usually begin to use their hands within a week. If osteotomies have been performed on the thumbs, support splints are made for the 1st web space to support the thumbs. The splints are made of a plastic type material which is moulded to fit your child's hand. This material becomes soft when put in hot water. The splint is then made and attached to your child's hand using velcro straps or a bandage. These children are very adept (quick) at using their hands very soon after surgery. They do very well despite

their inability to bend their fingers. In fact people not in the know often do not notice. Fine movements such as doing up buttons will always be difficult, and encouragement in these areas is very helpful, although children often find their own way of performing tasks. Practical measures such as velcro fastenings or curly shoe laces often help with their independence and gives them confidence in the presence of their peers.

## **Pfeiffer's syndrome**

**Anomalies seen:-** Broad and stocky phalanges and deviation of the thumb. Simple incomplete syndactyly of the 2nd web space may be seen.

### **Surgery undertaken**

Like in the Apert thumb, deviation is seen here, making pinch grip difficult and weak. Fine manipulative skills can be improved and made easier and cosmetic appearance more acceptable by an osteotomy as described previously. The syndactyly would be released as in the 4th web space of Apert's syndrome.

## **Saethre Chotzen syndrome**

**Anomalies seen:-** Simple incomplete syndactyly, most commonly seen in the 2nd web space. Mild to moderate **brachydactyly** (relatively short and stubby fingers), and **clinodactyly** (inturned) little fingers.

### **Surgery undertaken**

The syndactyly would be released as in the 4th web space of Apert's syndrome. The little fingers, unless affecting function of the hand, are left untreated, as clinodactyly is seen in many people with no other anomalies.

**Written by Rachel Hall, Clinical Nurse Specialist for Hand Anomalies, Great Ormond Street Hospital for Children NHS Trust**

This leaflet is for information purposes only.

**Other leaflets are available from  
Headlines-Craniofacial Support  
Please contact Group Administrator  
Gil Ruff on 01454 850557  
for details on how to obtain copies**