Treatment options for symbrachydactyly

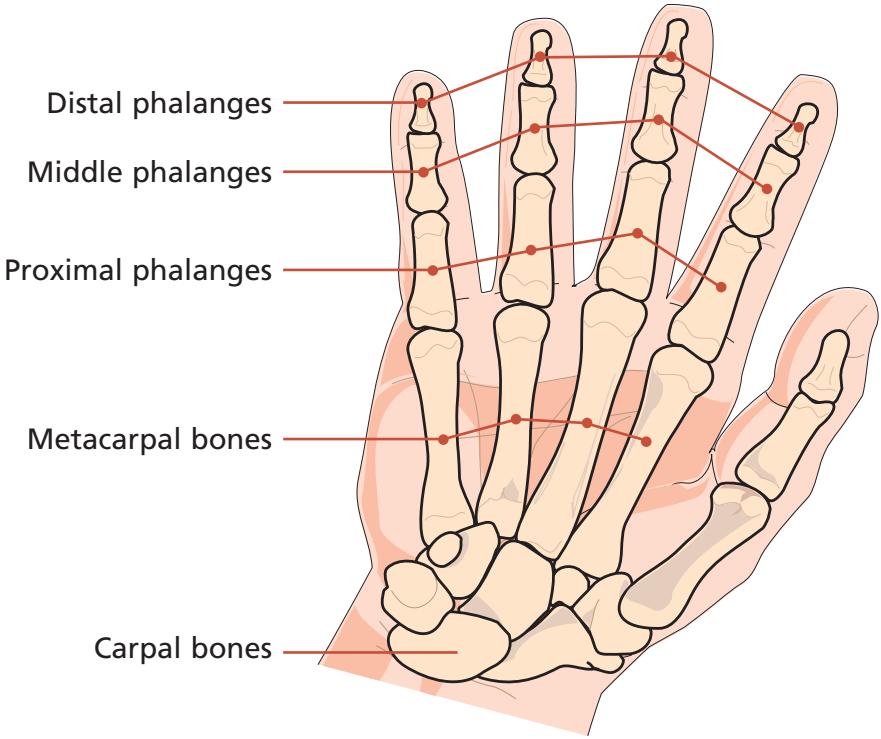
Information for families

Great Ormond Street Hospital for Children NHS Foundation Trust

This information sheet explains about the hand anomaly symbrachydactyly and how it can be corrected.

What is symbrachydactyly?

Symbrachydactyly is a congenital (present at birth) hand anomaly, which affects a single upper limb. It is not inherited. It is characterised by short, stiff, webbed or missing fingers. The underlying muscles, tendons, ligaments and bones are all affected.



Symbrachydactyly is graded from mild to severe but within each grade, the degree of severity can vary enormously.

- At the milder end of the spectrum, the hand has slightly short, mobile fingers with minor webbing. The hand bones (metacarpals) and some of the finger bones (phalanges) and the thumb are present.

Figure 1: Mild symbrachydactyly before treatment

- In moderate cases most or all of the finger bones are missing and small nubbins of skin and soft tissue are present where the fingers would have developed. The forearm and hand may also be shorter. The thumb is usually present, but may or may not be shorter than usual.

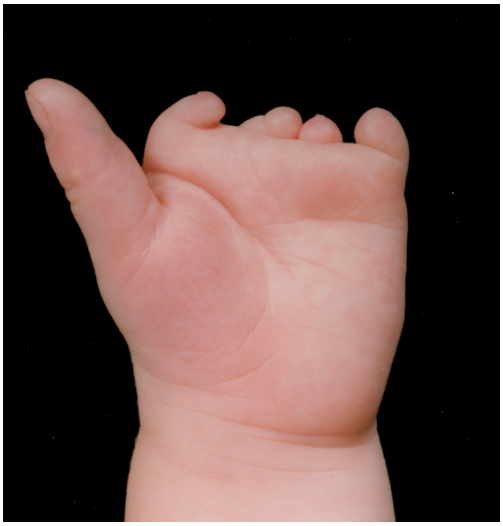


Figure 2: Moderate symbrachydactyly before treatment

- In the severe cases there is either a partial thumb or no thumb and no fingers.

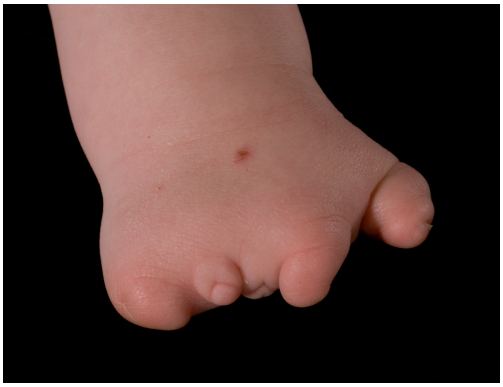


Figure 3: Severe symbrachydactyly before treatment

What causes symbrachydactyly?

The cause of symbrachydactyly is unknown. One possible cause might be an interruption of the blood supply to the developing arm at four to six weeks of pregnancy. There is no link to anything the mother did or did not do during pregnancy. There is also no increased risk of having another child with the same condition or that your child will pass the condition on to his or her children.

How common is symbrachydactyly?

It occurs in approximately 1 out of 32,000 births.

What is hand function like for children with symbrachydactyly?

Without any surgical treatment, overall hand function is usually good because symbrachydactyly affects one hand with the other being normal.

Most children use their unaffected hand as their dominant hand with the affected hand assisting. Surgery therefore aims to upgrade performance in the affected 'assisting' hand. In many cases, children can still grip with the affected hand although fine motor skills might be a problem. Also, children are very good at adapting to their condition, and can usually achieve most tasks. There are some things that can be more difficult, particularly with a more severely affected hand. These include some activities that need both hands, for example, hooking up a zip, managing brakes on a bike, using a knife and a fork together, holding paper when cutting with scissors, and playing some musical instruments. Some of these tasks can be managed with special equipment. You can get advice about this from your occupational therapist and through the organisation REACH (contact details at the end of the information leaflet).

How can symbrachydactyly be treated?

Symbrachydactyly can be treated with surgery, with the aim of improving hand function and appearance. However, it cannot give your child a normal looking hand.

The surgical options for reconstruction will depend on the severity of the condition. Some mild forms do not need any treatment. In some situations, the surgeon may present a choice of two options for surgical reconstruction but for others, only one type of surgery may be suitable. At the most severe end of the spectrum surgery may not be possible or worthwhile.

As well as surgery, there is also the option of prosthetics (an artificial hand or fingers). They can be used in conjunction with surgery or independently. These are made by your local Artificial Limb and Appliance Centre and can involve a complete false hand or individual false fingers. Your family doctor (GP) will need to refer you.

Figure 4: Example of hand prosthesis

Prostheses are used either for cosmetic purposes, or for specific functional purposes, for example, to help hold something such as a golf club. High quality prostheses give a near normal appearance, but this may be at the cost of function, since they prevent the use of touch sensation. More information about prosthetics is available in the REACH guide to artificial limbs, which is available online at www.reach.org.uk/ReachCMS/assetmanager/images/files/guidetoartificialarms.pdf

What are the options for treatment?

There are two types of surgery used to correct symbrachydactyly: distraction augmentation manoplasty and toe to hand transfer. More detail about each type follows.

Distraction augmentation manoplasty

This involves a series of operations. It aims to produce a hand with four short, stiff fingers, which can move at the knuckle joint only. This procedure is often used for moderate symbrachydactyly, where the metacarpal bones are all present and of reasonably normal length, the thumb is present and there are skin sacks with sufficient space to allow new bone to be placed in them.

The **first stage** of this surgery is called a **free phalangeal transfer**. This involves removing one bone from a toe and transferring it to the finger sack. This is repeated for each finger, taking a bone from another toe each time. The aim of surgery is to increase the length of the fingers and allow the thumb to work against them to produce a useful, gross grip. The amount of movement at the knuckle joints varies. Sometimes they are stiff and need stretches to maximise movement and sometimes movement can be almost normal. The fingers do not have joints so will not bend and straighten.

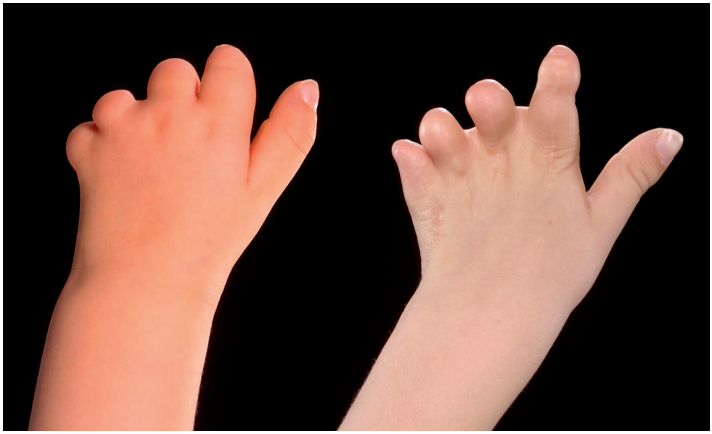
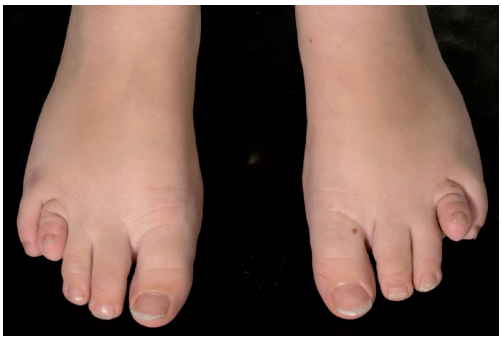


Figure 5:
Before and after free phalangeal transfer



The toes from which the bones are removed are shorter than normal, and occasionally do not sit in alignment. They are often floppy, but this does not alter the child's ability to walk or run. Usually the first bones of the third and fourth toes on each foot are used. The best time to undertake this procedure is before the age of two years, as the transferred bones have better chance of growing when a child is young. Some patients and their families choose to remain at this stage and do not proceed to the second stage.

The **second stage** is called **distraction lengthening**. Surgery aims to lengthen the finger bones to approximately the length of the first finger joint. This allows the child to hold larger objects. It is a time-consuming process, which requires a number of operations and there can often be complications post-operatively. The child is usually at least seven years old. This ensures that the transferred toe bones are big enough to have a distractor device applied, and the child is old enough to understand and want the procedure, and can comply with post-operative treatment.

A distractor is a device with two metal pins, which are inserted through the upper and lower portions of the finger bones, connected by rods. The bone is then cut between the pins and the rods are distracted away from each other to produce a gap. The process of distraction takes 8 to 12 weeks.

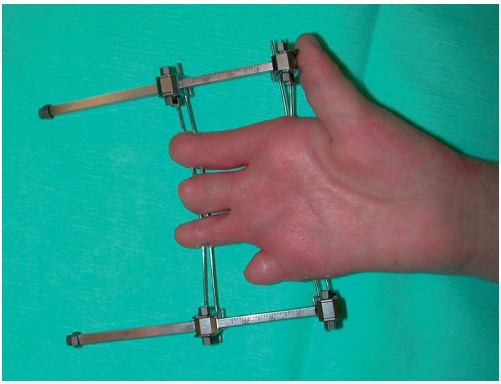


Figure 6: Distraction lengthening

While the distractor is in place it requires daily attention. Parents or carers will need to distract the bones twice daily by turning small screws attached to the distractor device. It is also important to keep the skin and the distractor clean. The pin sites should be cleaned daily before dressings and a bandage are applied. You will be shown how to do this following surgery before your child is discharged home. Your child can be referred to a community children's nurse who can support you at home. However they are unlikely to be able to visit daily so you will have to carry out most of the home care yourselves.

Your child will need to attend the hospital every one to two weeks during distraction to check progress and ensure any complications are identified as soon as possible. Complications at this point are: wound infections around the pin site area, bone fractures, displacement of the pins requiring surgery to re-site them, and pin breakages.

When distraction has achieved the correct length, further surgery is then needed to remove the distractor and graft the gap with bone taken from the hip or foot area. The bone of each finger is fixed together with a wire and left for a number of weeks to heal. The wires extend outside the fingers so that they can be removed when the bone has healed together. During this time the hand will need to be protected with dressings and a splint. Exercises should be done every day to encourage bending of the knuckle joints. Complications at this point are: infections around the wire; poor bony healing, stiff knuckle joints and finger dislocation, where the finger does not sit in the correct position. The wire will be removed once the bones have healed.

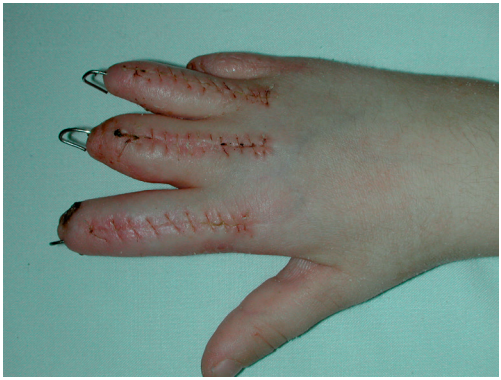
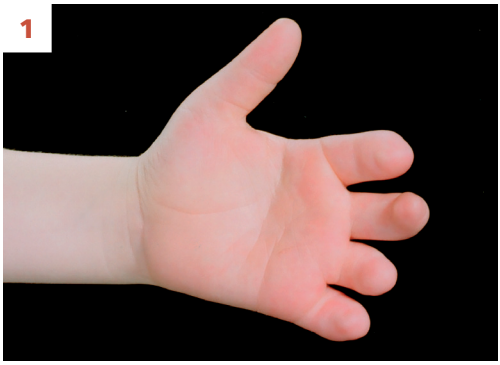


Figure 7: K-wires in place

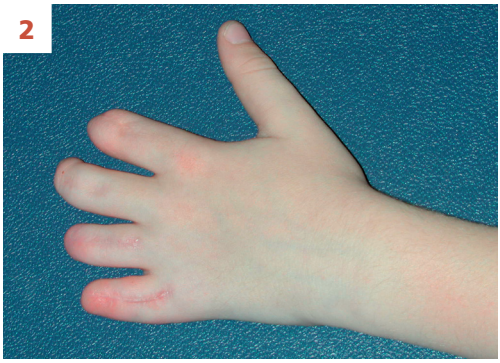
After the second stage is complete there is a **surgery-free period** to allow your child to use the hand so that he or she can gain movement, strength and function. During this time it is important to avoid injuring the hand and your child will be asked to wear a splint during sports activities. Contact sports should be avoided. This may be very difficult for some children who enjoy physical activity. Common complications at this stage involve fracturing the fingers, and occasionally stiffness at the knuckle joints, which will require a programme of daily stretches or surgical release if this is not successful. Sometimes the digits slip off the head of the metacarpal bones and require surgery to re-align them.

The **third stage** is to deepen the web spaces between the fingers. This involves surgery with skin grafts and allows better spreading of the fingers.

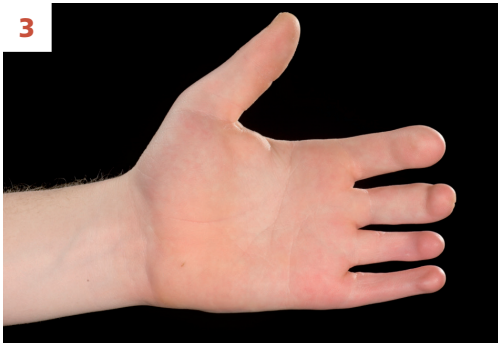
Figure 8: Child with mild symbrachydactyly before free phalangeal transfer (1),



after free phalangeal transfer (2)



and after distraction (3)



Toe to hand transfer

This procedure involves the removal of one or two toes (the second toes from each foot), which are then transferred to the hand to allow the thumb to have something to pinch against. It produces a hand with one or two digits. The digits have nails, and joints which can bend and straighten, although not as much as finger joints. Toe to hand transfer involves complicated micro-vascular surgery, which is usually done after the age of four years, when the blood vessels of the toes and those in the hand are big enough to allow transfer reliably. This surgery is carried out when the affected hand has metacarpals of a reasonable length present. The thumb may not be normal, but will need to be present. It will take about six months before sensation is restored after surgery and the hand is functioning well. Other minor procedures may be needed to free the tendons from scars to improve motion and/or to improve appearance.

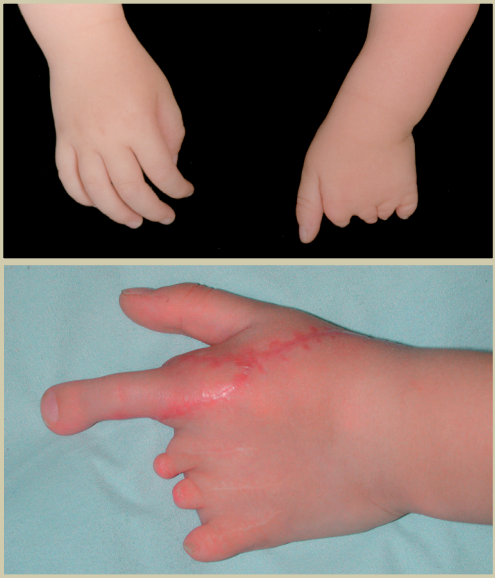


Figure 9: Before and after toe to hand transfer

The donor site leaves a foot reduced in width, with four toes present. The gap where the toe was removed is closed. Removal of the second toe does not have any functional impact on walking, running or jumping.

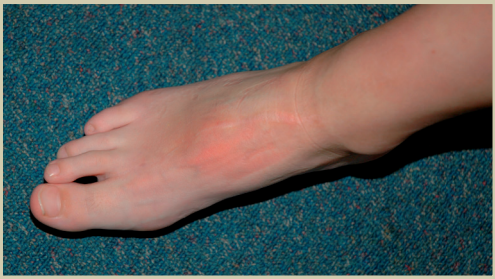


Figure 10: After toe to hand transfer

Does surgery improve the cosmetic appearance of the hand?

Often one of the expectations of undergoing surgery is that it will improve the way the hand looks as well as upgrade hand function. Surgery may improve the hand's appearance, but it will never achieve a normal looking hand, and does this at a cost to the appearance of the feet.

As children grow older they will become more aware of the how different the affected hand looks. This can sometimes lead to self-consciousness and a worry about being stared at or teased at school.

It may be helpful to discuss these issues with the clinical psychologist attached to the team, who will be able to help with strategies to deal with teasing and self-consciousness and help with decision-making. Our booklet *Bringing up a child whose face looks different* was written for families of children with birthmarks but contains some useful strategies for dealing with unwanted attention to any sort of visible difference. It is available on our website or ask for a copy at the Patient Advice and Liaison Service (Pals) office.

Further information about symbrachydactyly

If your child has symbrachydactyly and you would like to know more about the condition and how it can be treated surgically, you will need a referral to the Congenital Hand Anomaly team.

Contact **REACH**, www.reach.org.uk, the support group for children with all sorts of arm and hand problems, by visiting their website or telephoning 0845 130 6225.

Notes

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Compiled by the Congenital Hand Anomaly team
in collaboration with the Child and Family Information Group

Great Ormond Street Hospital for Children NHS Foundation Trust
Great Ormond Street
London WC1N 3JH

www.gosh.nhs.uk