



Great Ormond Street Hospital for Children NHS Foundation Trust

Monobloc advancement with or without using a rigid external distraction (RED) frame: information for families

Monobloc advancement is an operation to reshape the front portion of the skull and face including the upper jaw to correct an abnormal head shape. This information sheet from Great Ormond Street Hospital (GOSH) explains about the operation called monobloc advancement with or without rigid external distraction (RED) frame, which is used to treat craniofacial disorders. It explains how to prepare your child for surgery as well as what to expect in hospital afterwards. An Easy Read information sheet is also included for your child.

Reshaping and moving the front portion forwards means that the eyes sit deeper in the skull so are less likely to be damaged. Airway problems may also be improved as well as increasing the space within the skull to allow the brain to grow and develop.

Monobloc advancement can be carried out at any stage in childhood. Children with the following types of craniosynostosis may benefit from this operation:

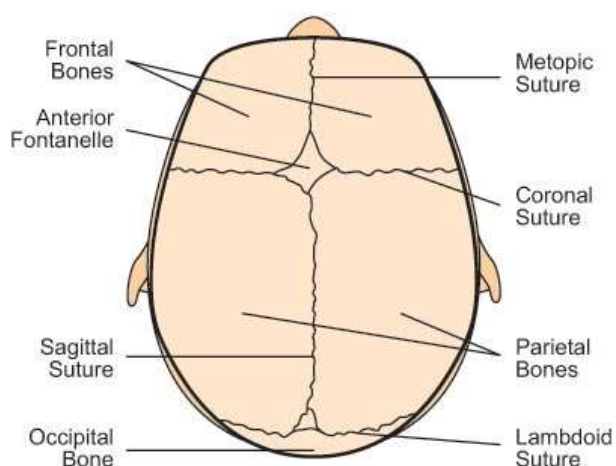
- Apert syndrome
- Crouzon syndrome
- Pfeiffer syndrome

It may be the only form of treatment needed or it may be just one in a series of operations carried out throughout childhood and adolescence.

Monobloc advancement may use a rigid external distraction (RED) frame after the operation. The RED frame is made of pins inserted into the skull bone attached to a frame with screws, which are moved each day gradually stretching the bone and soft tissue of the face to bring it forwards. New bone tissue forms in the gap created. When enough distraction has occurred and the bone is healing well, the frame is removed in another short operation.

Sometimes, bone grafting is used instead of a RED frame. This involves moving bone from another

Normal Skull of the Newborn



part of the skull and fixing it in place with plates and screws.

Getting ready for the operation

Pre-admission clinic

Preparing for a planned operation, test or procedure before coming in to hospital avoids delays and reduces the risk of cancellation. The results of any tests and investigations are available in plenty of time and can also be re-checked if they are not within the normal range. Your child may need various blood tests before the operation – this depends on your child's medical condition and the nature of the surgery that is planned.

The doctors and/or nurses will meet you and your child to take down their medical history and any other information needed before your child is admitted to hospital. The nurses will explain about any care your child will need before and after the operation. If your child has any medical problems, particularly allergies, please tell the doctors about these. Please also bring in any medicines your child is currently taking.

You may be seen by one of the team carrying out your child's operation and be asked to give permission for the procedure by signing a consent form. If you give your consent at the pre-admission appointment, you will need to confirm that you still agree to the procedure on the day of admission.

One of the team will explain about the types of anaesthesia that are used at the hospital, and also about options for pain relief after the operation, test or procedure. If there are any questions or concerns about your child's anaesthesia, an anaesthetist may come to see your child in the pre-admission clinic.

The night before surgery

You will be asked to give your child a bath or shower and hair wash before surgery.

It is important that your child does not eat or drink anything for a few hours before the operation. This is called 'fasting' or 'nil by mouth'. Fasting reduces the risk of stomach contents entering the lungs during and after the procedure.

You will be informed the night before the procedure of the time that your child should be 'nil by mouth' – in other words, have nothing to eat or drink before the anaesthetic.

It is equally important to keep giving your child food and drink until those times to ensure they remain well-hydrated and get adequate nutrition. This may involve waking your child in the night to give them a drink which we recommend.

On the day of surgery

Please come to Woodpecker Ward at the time stated in your admission letter. One of the nurses will check that your child is well enough for the operation, complete some paperwork with you and take some baseline observations of their temperature, heart rate and breathing. They will also put an identification wristband on your child.

If you did not give your consent for the operation at the pre-admission appointment, a member of the surgical team will visit you to explain about the operation and ask you to sign a consent form.

Important

The person bringing your child for the operation should have 'Parental Responsibility' for them. Parental Responsibility refers to the individual who has legal rights, responsibilities, duties, power and authority to make decisions for a child. If the person bringing your child does not have Parental Responsibility, we may have to cancel the operation.

All children are seen by the anaesthetist on the day of the operation.

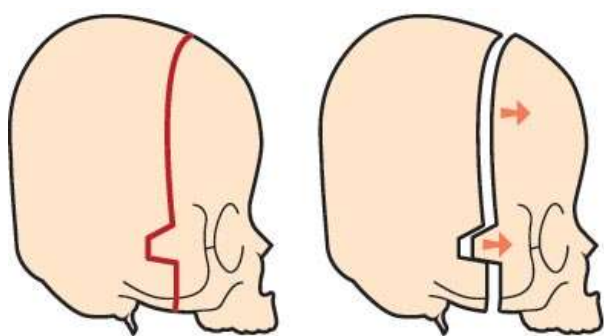
What anaesthetic is given?

Your child will be given a general anaesthetic by an anaesthetist who specialises in giving anaesthetics to babies and children. Both parents will be able to go with your child to the anaesthetic room and stay until they are asleep.

This usually involves your child breathing some anaesthetic gas. Later, a tube is passed into the airway (trachea) to safeguard breathing. A cannula (thin, plastic tube) is put in a vein and usually left in place for a short time after the operation. Fluids can be given to your child through this tube during the operation and afterwards if necessary.

What does the operation involve?

When your child is under general anaesthetic, the surgeon will clip your child's hair just over the incision site and fix the rest of their hair out of the way. They will make an incision over the top of your child's head from ear to ear, between the coronal sutures and the hair line. They will then pull the skin and soft tissues over the forehead downwards to expose the skull.



The surgeon will adjust the position of the forehead and midface. The forehead may need to be re-shaped. The bone is pulled forwards to leave a small gap between the cut edges.

If bone grafting is being used, small pieces of bone will be taken from the skull and used to fill the

gap. When the bones are in place and the surgeon is happy with the result, the bone grafts will be fixed in place with strong stitches that dissolve very gradually over the next few months while the bone heals. Sometimes plates and screws are used to hold the bone grafts in place. The skin is then closed over the incision site and held in place with dissolvable stitches.

If a RED frame is being used, the surgeon will insert pins through the skin into the skull bone in several places either side of the gap. The pins are then attached to a frame and screws.

Some children have a drain inserted which will be left in place to drain off any fluid that collects after surgery – this will be removed a day or two later when no longer needed. Finally, the surgeon will put a head bandage over the operation site. Your child will then be taken to the Recovery area to start to wake from the anaesthetic.

Are there any risks?

Healthy children usually cope well with the anaesthetic, but the risk increases if your child has other problems.

All surgery carries a small risk of infection or bleeding. To reduce the risk of infection, your child will be given an initial dose of antibiotics during the operation. This will continue as an infusion into a vein (intravenously) for seven days after surgery.

Very rarely, the cerebrospinal fluid (CSF), which is a watery liquid that surrounds the brain and spinal cord acting as a 'cushion', also becomes infected, causing meningitis. This will need additional treatment with intravenous antibiotics.

This surgery requires separating the skull bone from the protective layer covering the brain (the dura), a process which can, in a tiny proportion of cases, cause brain injury or bleeding inside.

Therefore, any craniofacial operation carries a very small chance of causing serious complications

such as seizures (fits) or stroke (brain damage), which may in some very rare cases be life-threatening. The overall risk of a major neurological event or death is much less than one per cent (less than a 1 in 100 chance).

Sometimes small holes can be made in the dura during the operation, which do not pose any risk to the brain, but which can occasionally be the source of a leak of cerebro-spinal fluid (CSF; a clear, watery fluid that surrounds and cushions the brain). The surgeons usually identify and repair (stitch) any holes during the operation. Despite this, sometimes children develop a CSF leak after surgery, which may show up as a fluid-filled swelling at the operation site or a leak of clear, watery fluid from the wound. It will be explained to you what to do if you notice these problems after discharge from hospital, as sometimes further procedures may become necessary to stop the leakage if one occurs.

Your child will be monitored very closely during and after the operation to identify any blood loss. A blood transfusion is sometimes required but the surgeon will have ensured that donated blood of the correct type is available if needed.

Your child's head and face will look swollen and bruised after the operation. This particularly affects the eyes following monobloc advancement and the nurses will clean them carefully in the days following the operation. Swelling tends to get worse for the first two to three days and then start to improve. The nurses will check your child's head bandage every few hours and re-apply it if it is getting tight.

The incision site will start to heal and will eventually fade until it can hardly be seen. Occasionally scars can become wide or red and bumpy. The hair will also start to re-grow in the days following surgery. Some children develop velopharyngeal insufficiency after the operation. This is a problem with the soft palate and back of the nose, which may cause nasal sounding speech

and problems with eating and breathing. Occasionally further surgery may be needed.

Bone grafting carries some specific risks. There is a chance that the skin and soft tissue may not be able to stretch over the portion of skull bone that has been moved forwards. This may delay healing as the skin and soft tissue are under tension. If the bone grafts have been fixed in place with plates and screws, there is a chance that the screws could become buried in the bone or noticeable under the skin. This does not cause any problems immediately but could make future surgery more complicated.

The use of a RED Frame also carries some specific risks. There is a chance that the equipment could become dislodged or the pins may fail. This is not necessarily harmful but a further operation to repair or re-insert the pins may be needed. As the pins pass through the skin to the bone, infection can be a problem. This risk can be reduced by good hygiene but sometimes infections develop even with good hygiene.

The process of distraction is not painful but may cause some discomfort. Giving regular pain relief is usually enough to deal with any discomfort. Tiny scars will be left when the pins are removed but these will be hidden by the hair.

Are there any alternatives?

Monobloc advancement is the only operation that can reshape the front portion of the skull, face and upper jaw to correct an abnormal facial appearance all in one operation. Occasionally multi-staged procedures are a better choice.

What happens after the operation?

Once your child has started to recover from the anaesthetic, they will be brought back to Koala Ward to continue recovering. At the end of the day, the doctors will visit you to talk about the operation. They will spend the first few nights in

the High Dependency Area on the ward, where a nurse will monitor them closely to watch for any signs of bleeding and fluid imbalance. They will carry out regular observations of their breathing, heart rate and temperature throughout the rest of the day and night.

Your child may feel sick after the operation, but the doctors will give them medicines to reduce this. The sickness should pass within a couple of days. Your child will be able to start eating and drinking as soon as they feel like it.

The nurses will also assess your child's pain and give them regular pain relief for the first few days – intravenously to start with and then by mouth when they are eating and drinking. The intravenous infusion of antibiotics will continue for seven days.

Your child's eyes will be very swollen after the operation so the nurses will clean them gently to make them more comfortable. Your child should sit and lie in as upright position as possible to reduce any swelling. The swelling is temporary and will start to improve in a few days. The nurses will remove the head bandage and drains when they are no longer needed.

At various points during your child's stay, the nurses will wash your child's hair gently with a mild shampoo and show you how to do it safely at home.

Over the next few days, your child will be able to start moving around.

If your child has a RED frame in place, the nurses will start to teach you and your child how to look after it. They will also give you an information booklet explaining what to do and how other families have coped. While the RED frame is in place, your child will need to eat a soft, sloppy diet with no chewing. The nurses will advise you on what to feed your child once you get home.

Going home

Your child is likely to be in hospital for up to three weeks but can go home sooner if you are confident in distracting the RED frame.

The nurses will also give you a copy of our discharge information which explains how to look after your child and what signs to look out for over the next few weeks.

Follow up appointments

Your child will have a series of appointments after the operation – we will give you details before you go home.

Removal of the RED frame

If your child has a RED Frame, distraction will continue for two to three weeks after the operation. The frame is then left in place without any distraction for a further six to eight weeks while the bone heals. Your child will then need to come back to GOSH to have the frame and pins removed in a short operation under general anaesthetic. In most cases, your child will be able to go home later the same day.

What is the outlook for children who have had monobloc advancement with or without rigid external distraction?

The monobloc advancement operation is successful in most cases and results are usually good. This operation provides a firm basis for future surgery to improve head shape. Overall most parents are happy with the results. The lower jaw is not affected with this operation. Further surgery may be required in late adolescence.

If you have any questions, please telephone the Craniofacial Clinical Nurse Specialists on 07768 526 449 (Monday to Thursday from 9am to 5pm). Out of hours, call Koala Ward on 020 7829 8826. You can also seek medical advice out of hours by telephoning the GOSH switchboard on 020 7405 9200 and asking them to page the craniofacial doctor on call.