

Little Hearts Matter



# The Fontan procedure

or Total Cavopulmonary Connection  
(TCPC)





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# Introduction

Children born with only one working pump in their heart (single ventricle heart disease) have a series of operations that aim to make the best use of the one pump that they have. The Fontan procedure, completion of the Fontan or Total Cavopulmonary Connection is one, and for many the last of the children's planned operations. (For ease of description, all of the above names for surgery will be referred to as the Fontan).

Most children have their initial surgery in their first year of life. The Fontan procedure is usually offered when a child is older, with the timing planned for each child on an individual basis, most commonly between the ages of three and six years.

The aim of the Fontan procedure is to give a child as much energy as having only half a pumping heart will allow. If surgery has gone to plan the children will gradually recover from the operation and go on to gain far more energy than they had before.

## The history of the Fontan procedure

Francis Fontan, a French cardiac surgeon, developed the first Fontan procedure in 1971 as a treatment for Tricuspid Atresia.

Gradually, more surgeons around the world developed the surgery, with Professor Marc de Leval from Great Ormond Street Hospital for Children expanding the use of the surgery in the UK using the operation to support other right-sided heart conditions.

In the late 1980s the Fontan was developed as one of the stages of treatment for Hypoplastic Left Heart Syndrome.

In 1990, Salvatore Giannico and Carlo Marcelletti developed the External Fontan (Total Cavopulmonary Connection) as a way to avoid some of the complications that arise from the original operation, especially unusual heart beats (arrhythmias).

# The Fontan procedure

## What are the signs that it is time for surgery?

Whilst a child is growing, eating and participating with their friends in play and physical activity, they are still making the most of their previous surgery.





Once a child starts to physically slow down and is increasingly unable to keep up with their friends during play, or they are sleeping for longer or needing short naps during the day, they are showing signs that their heart function is changing.

They may become more breathless and/or bluer (more cyanosed), they may eat less and they may stop growing.

## What do I tell the doctor?

These signs become part of the assessment process. The information given to the cardiologist at an outpatient visit helps them to understand what life is like at home so children and their parents need to plan what they are going to say to the doctor at each outpatient visit.

They need to know about:

-  climbing the stairs
-  playing with friends
-  sleep patterns
-  when the child is breathless and how long it takes them to recover from a breathless episode.

The cardiologist will also be looking for signs within the heart, initially at an outpatient visit by performing an echo or Magnetic Resonance Imaging (MRI) scan. These tests will look at the structure of the chambers and the blood vessels in the heart, and the flow of blood through the heart so the doctors can identify any narrowings or blockages to the flow of blood. They will also be looking at how strongly the heart is pumping.

If they begin to see that the heart's function is changing they may arrange for a cardiac catheter. This is often planned within the six months before they think that the Fontan may be needed. Some units may choose to combine the admission for cardiac catheter and then surgery if the treatment plan is clear from outpatient tests. The information gained during cardiac catheterisation will confirm any narrowings or blockages of major blood vessels within the heart. It will also give information about the pressure of the blood in different parts of the heart and lungs. This information is extremely important as it helps the cardiologists and cardiac surgeons to plan the timing of surgery.

## Who decides when surgery is necessary?

At each of the UK's congenital cardiac surgical hospitals a meeting called a multidisciplinary team (MDT) meeting is held once or twice per week. All of the doctors who work within the cardiac team attend: cardiologists, surgeons, cardiac anaesthetists and intensive care doctors as well as specialist nurses and clinical nurse specialists. The aim of these meetings is for the cardiologist to present information collected about a child that they feel needs further interventional treatment, either surgery or interventional cardiac catheterisation. All the members of the team discuss the pros and cons of treatment and they agree on the best plan for each individual child. They will also agree the best timing of surgery.

The timing of the Fontan operation will be different for every child. In most cases it will be performed between three and six years of age. The operation is planned because the child is showing signs that they need more oxygen-filled blood flowing around their body. This added oxygen will give them more energy for growth, play and learning.

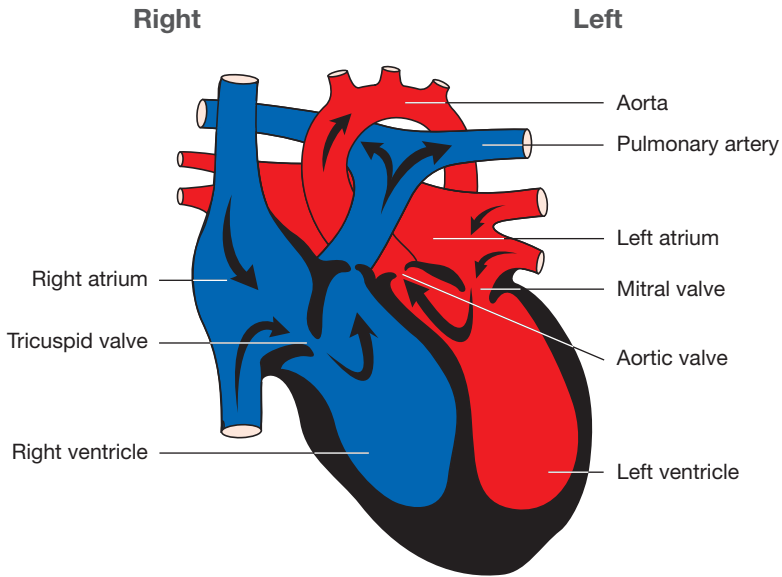
Following agreement on the treatment plan and timing, parents will receive a letter from the hospital or a phone call confirming the doctors' plan.

In some cases the medical team may suggest a delay in surgery while they change medication that may improve a

child's condition before surgery is performed. For example, if a child has a high blood pressure in their lung blood vessels they will struggle to cope with the new blood flow after a Fontan so they may be started on medication to bring the lung (pulmonary) blood pressure down.

Before we look at what the Fontan operation does, it is helpful to look again at what a normal heart does.

## The normal heart



The heart's job is to pump blood around the body. There are two separate circulations: the first takes blood to the lungs (the right side of the heart), and the second takes blood to the body (the left side of the heart).

This is how the journey begins: blood returns from the body, via veins, to the right side of the heart into a collecting chamber called the right atrium. This blood has a bluish tinge (blue blood) because the body has extracted oxygen from it (deoxygenated blood).

The blood is then passed through a valve (tricuspid valve) to a pumping chamber (right ventricle), which pumps the blood to the lungs via the lung arteries (pulmonary arteries).

The blood picks up oxygen as it passes through the lungs which turns the blood a red colour (oxygenated blood). This blood then returns to the left collecting chamber (left atrium) and it then passes through a valve (mitral) to the left pumping chamber (left ventricle). The left ventricle then pumps blood to the body through a valve (aortic valve) and to the body via the body artery (aorta). The body uses the oxygen from the blood, turning it blue again. And the journey starts again.

In a normal heart the right side pumps used blood to the lungs and the left side pumps oxygen-filled blood to the body. The blood on the left and right side do not mix.

A child born with only one of the two pumps (ventricles) has a mixture of blood pumping within their heart and body because when they are small this is the safest way to make sure that the body gets some oxygen.

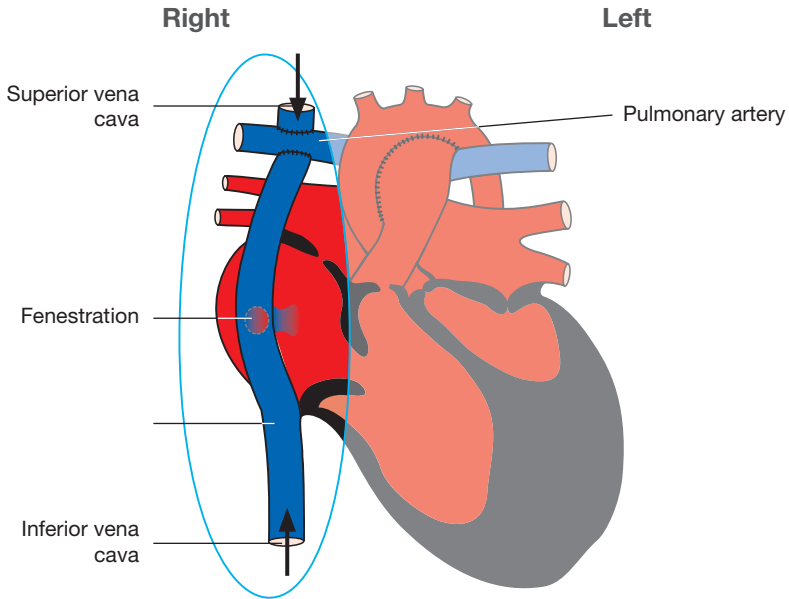
The Fontan operation finally separates the flow of blue (deoxygenated) blood returning to the lungs from the red (oxygenated) blood leaving the heart for the body. The surgery uses the one working pump (ventricle) available to do the hardest job of the heart which is to pump oxygen-filled blood around the body.

All blood returning to the lungs is re-directed to the lung blood vessels without the use of a pump.

There are two different styles of surgery, the External Fontan (Total Cavopulmonary Connection (TCPC)) or the Internal Fontan. Although the operations are slightly different the aims and the outcomes are exactly the same. The choice of operation is determined by the surgeon's preference or the type of heart structure a child has.



## External Fontan - Total Cavopulmonary Connection

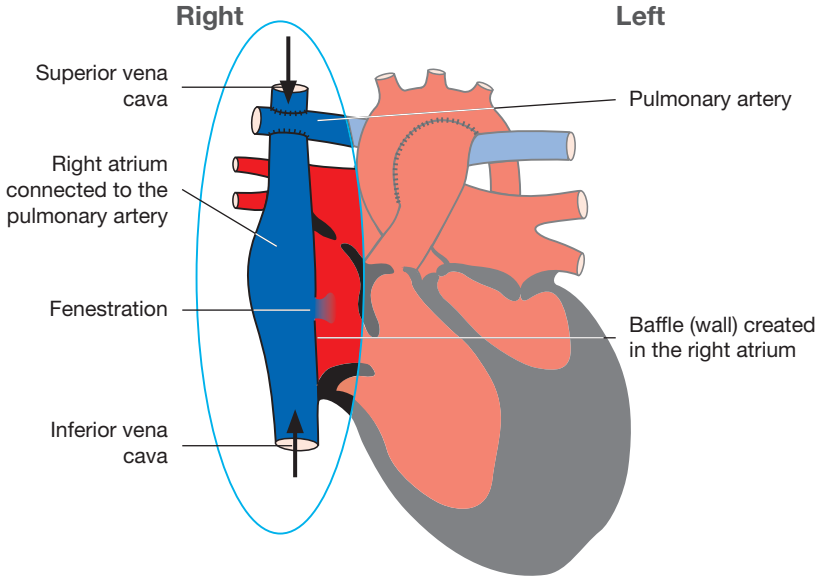


This operation is done by attaching a tube of a special plastic (a conduit of Gore-Tex®) from the lower body vein (inferior vena cava) to the base of the lung artery (pulmonary artery), diverting blue (deoxygenated) blood away from the heart straight to the lungs.

A hole (fenestration) (see page 11) may be created between the tube and the right collecting chamber (right atrium). As with the Internal Fontan (see page 10) there can be a rise in pressure in the lung arteries after surgery and the hole acts as a pressure valve.

Children may be in hospital for some weeks after the third operation as it is important to give them time to adjust to their new circulation.

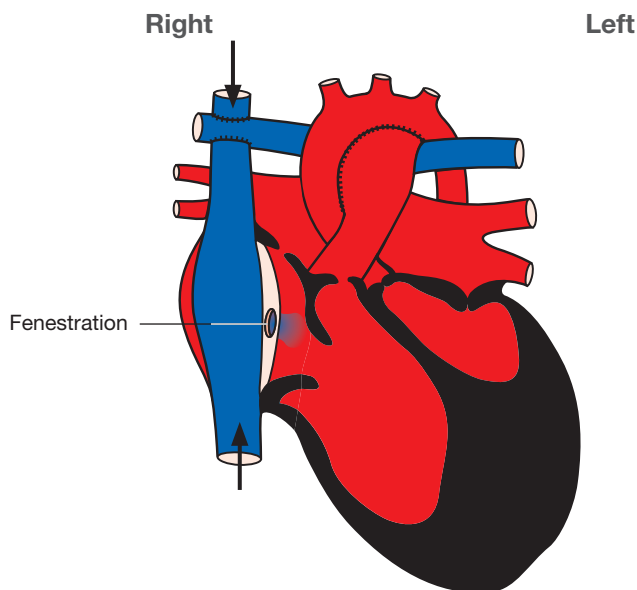
## Internal Fontan



This is done by creating a wall (baffle) out of Gore-Tex® in the right collecting chamber (right atrium) and then attaching the chamber to the base of the lung artery (pulmonary artery). This may have been partially completed at stage two, the Hemi-Fontan.

All the returning blue (deoxygenated) blood will now be flowing to the lungs, without a pump behind it. This causes an increase in pressure within the lung blood vessels, so, to aid circulation, a small hole (fenestration) (see page 11) may be created in the wall (baffle). This acts as a pressure release valve whilst the child's body adjusts to the new circulation.

## Fenestration



Some children have a fenestration created during surgery. A fenestration (window or hole) is routed between the blue blood flow from the body to the lungs and the red blood flow through the heart going to the body.

This hole is created as a pressure valve. If the blood flowing to the lungs is moving slowly because the blood pressure in the lungs is high it can create a back-flow down through the veins in the body. To prevent a build-up of pressure a small hole is created which pressured blood can pass through, a little bit like the pressure valve in a boiler.

This hole prevents too much pressure building up within the body's veins and organs, helping to keep the circulation moving smoothly. As a child grows and becomes more able to manage the change in heart pressures the fenestration may no longer be needed.

The fenestration may gradually close as the child grows or may be closed, if no longer needed, when the child is older. This is done during a cardiac catheter procedure.

# How does the Fontan operation work?

The Fontan procedure will only work if three things are in place in the heart:

- The one heart pump available has to be strong and pumping well.
- The flow of the blood through the heart and blood vessels has to be smooth, not blocked or restricted.
- The blood pressure in the lungs has to be lower than the body's blood pressure.

The Fontan operation completes the aims of the two or three operations started earlier in childhood. Oxygen-filled blood is routed through the heart to the one pump available so that the heart can push the red, oxygen-filled blood out to every part of the body.

The body uses that oxygen to make energy and then sends the now blue oxygen-empty blood back towards the lungs to pick up more oxygen. The pump that pushed the blood out of the heart creates the push that helps it back to the lungs. It is helped to flow forward by very small one-way opening valves, which are naturally in the veins which stop slow-moving blood from going backwards.

## Vein valves

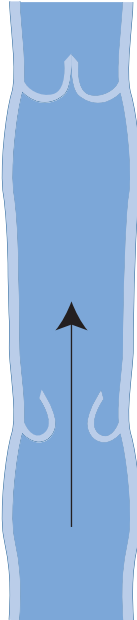
The blood passes from the lower body to the lungs either through the newly formed external tube (conduit) or through the passage formed in the collecting chamber in the right side of the heart (see Fontan diagrams on pages 9 and 10).

The flow of the blood through the heart and blood vessels has to be smooth, not blocked or restricted.

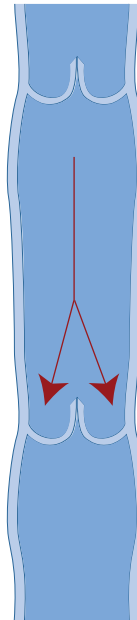
The blood from the head and neck is already flowing to the lungs as that operation was performed earlier in the child's life, the Cavopulmonary Connection, Glenn Shunt or Hemi-Fontan, for some children their second-stage operation. During the last stage of surgery the major blood vessel bringing blood up from the body back up to the heart, the inferior vena cava (lower body vein) is attached directly to the lungs using a tube or wall. For the blood to flow smoothly to the lungs, with no pump behind it, the blood pressure in the body must be higher than the blood pressure in the lungs as fluid flows easily from an area of high pressure to one of low pressure. The blood pressure in the lungs has to be lower than the body's blood pressure.

## Vein valves

Blood  
flowing  
to heart



Healthy valve  
prevents reverse  
blood flow



# How long does the Fontan operation take?

As there are two styles of surgery used to create a Fontan circulation the operation takes different lengths of time.

The External Fontan surgery is done outside the heart and the need to stop heart blood flow is reduced.

The Internal Fontan involves surgery within the heart and may take a little longer.

It is important to remember that a great deal of the operation time is spent setting up the equipment needed to keep your child safe during surgery. Following the operation, time is taken to make your child safe before moving them to the Paediatric Intensive Care Unit (PICU).

Ask your doctor to give you an approximate time that your child's operation might take.

# What happens next?

Following the doctors' multidisciplinary team meeting, parents will receive a letter or a phone call telling them about the plan for surgery and the timing of the operation. In most cases it will be suggested that surgery should be done within six months. Occasionally surgery will be planned sooner.

## When should parents start to prepare their child to go into hospital?











The most important part of preparing your child for surgery is to make sure that you are prepared yourself. Make sure that you understand what the surgeons plan to do and what care will be like after surgery. Once you are confident that you know what the plan is, it will be easier to prepare your child with confidence.

LHM have a booklet *Preparation for hospital* and some leaflets on useful books and toys.

Some hospitals hold a pre-admission clinic or visit. Tests may be done in preparation for the child's admission - chest x-rays and blood tests for example. In some hospitals the children have the chance to explore their forthcoming admission through play. This can be a really helpful way to understand how your child is feeling about going into hospital.

Overleaf there are a few tips for preparing your child for a hospital visit.

## Hospital preparation tips

-  Use play to explore going into hospital with your child.
-  Talk to the specialist nurse if you have questions about the admission, operation or aftercare. It is easier to plan for what might happen rather than worry about an issue that may not arise.
-  Call the hospital to find out where you will be sleeping if you are staying with your child.
-  Confirm long-term hospital parking if parking is possible.
-  In the week leading up to surgery help your child understand that drinks might be smaller than normal by offering only half a cup of fluid when they ask for a drink. However, don't deny them a drink if they are really thirsty. See page 23 for more details.
-  Get your child and their siblings to pack a bag full of their favourite toys and pack favourite pillows or duvet covers that can be used on the ward (you may need to ask if your ward allows this).
-  Pack your own survival kit: cereal, comfortable clothes, chocolate, coffee and tea.
-  Pack your phone charger.
-  Give all your close family a small job so that they do not feel left out of the support and planning. Give brothers and sisters the role of toy provider; they can change the toys from home so that the toys in hospital don't become boring.
-  Organise a 'communication tree' or create a special Facebook page so messages about how the stay is going reach all of your friends with one message.



## The day of admission

Each hospital stay will be slightly different but in most cases a child being admitted for major heart surgery will need to be at the hospital the day before their operation. This gives the medical and nursing teams a chance to ask questions about the most recent medical history and find out about your child's likes and dislikes. They can also undertake any tests that have not been done at a pre-admission visit.

There will also be an opportunity to meet again with the surgeon and discuss the plan for surgery. This is when parents will be asked to give consent for their child to have the operation. The anaesthetist will come to examine your child and chat to you before the operation. They will ask questions about the current health of your child and also prescribe any medication that they think will be helpful in settling the child just before the operation, the pre-med.

## How will my child travel from the ward to the operation?

If your child has had some pre-surgery medication to settle them, they may be very drowsy when they travel to the operating theatre so they normally travel in their bed or on a theatre trolley. Occasionally a parent can carry them to theatre if the child is distressed or clingy but they will need to be in their bed for the full anaesthetic. Both parents can travel down to the operating theatre entrance but sometimes only one of them can actually go into the anaesthetic room where they can stay with their child until they are asleep.

Some anaesthetists are happy to give an injection into the hand before adding tubes, others give the child a whiff of anaesthetic gases. They will ask pre-operatively if your child has a preference.

The main aim during this time is to keep the child as calm as possible. Gentle chatter until they are asleep, counting sheep or other animals or trying to count backwards helps the child to concentrate until they fall asleep.

A game to play at home before admission is counting backwards with small bets on how far your child can count when they have their anaesthetic.

## What do parents do during surgery?

Being a mum or dad is never easy but it is an extremely stressful time during surgery.

Most parents are encouraged to get out of the hospital for a while to get a break from the ward, where they may be staying for some time after the operation.

They are encouraged to have a proper meal because once their child is back from their operation it is sometimes difficult to fit in more than a short break away from the bedside.

If parents can have a sleep or at least a rest it is a good thing at this time because they may spend many restless nights beside their child's bed over the next few nights.

Parents will be updated about the progress of their child's operation by the ward-based nursing team by ringing or texting them on their mobile phone.

Once their child's operation is complete they will be asked to return to the ward where the surgeon may visit them or they may be taken to the PICU where they will meet the doctors and, once their child is settled, they will be able to join their child.

# After the operation

Immediately after the Fontan operation children are cared for in the PICU.

## Breathing






As with earlier surgery, immediately after the operation, the children are ventilated, their breathing is done by a machine.

However, as soon as the medical team are happy that the child is coping with the operation they will remove the ventilator.

For the Fontan procedure to support a good circulation the blood pressure within the lungs needs to be as low as possible. The ventilator causes a rise in blood pressure so the doctors will remove it as soon as it is safe to do so.

Once the ventilator has been removed and the intensive care team and surgeons are happy with the heart's function they will send the child back to the ward for further monitoring and care.

### Ventilation is done for a number of reasons

-  It gives a child the opportunity to rest as much as possible which takes strain away from the heart.
-  It gives the medical staff time to assess how the child is coping with surgery and the new blood circulation.
-  It gives the surgeons and intensive care staff the opportunity to balance medications and the heart's function.
-  If the medical team have to re-operate it can be swift.
-  The child can be kept comfortable and stress-free.



## Colour

The job of the Fontan procedure is to separate the mixed blue and red blood within the heart and circulation. After surgery, for the first time, most blood circulating around the body will be red. Consequently, children look very pink after surgery. Even children whose parents have not thought that they have a blue colour are astonished at how pink their child is.

Oxygen saturations (levels of oxygen in the blood) will be measured throughout the child's stay in hospital.



It was a shock to see Charlie back on a ventilator on PICU after so many years, but it was amazing to see him so wonderfully pink. I had never realised that his lips could be such a beautiful ruby red colour!



## Scar

The surgeon will use the same scar line that was created during earlier surgery, down the centre of the chest, a sternotomy.

It is rare that the scar line is left open after this operation, but if the surgeon feels that the heart needs a little time to settle following surgery they may cover the wound with a large dressing until they are happy to close the wound.

The scar will be closed with stitches that will dissolve as the wound heals. The breast bone (sternum) will be closed with wire which will remain in the chest. This is normal following heart surgery.

If the wound becomes red or begins to leak fluid or pus, tests will be done to investigate the problem and then antibiotics may be prescribed.

The scar wound will heal over six weeks and fade over six months.





## Fluid balance

Straight after surgery the children will be given only small amounts of fluid in the way of drinks and the fluid given into the veins. This helps to reduce the heart's workload.

Everything that we drink, or are given by drip (infusion), ends up in our blood stream. The heart has to pump this blood around the body. If the fluid intake is restricted the amount of fluid in the blood is less so the heart does not have to work as hard.

Initially all fluid will be given by drip (infusion). Gradually, very small amounts of fluid will be given as drinks.

This can be very frustrating for the children as they often want a long drink. One possibility is to offer the fluid as frozen ice pops as these last longer and are more refreshing than a small sip of fluid. The nurses will record all types of fluid and ensure that there is a balance between the fluid that goes in and the fluid that is passed out either as urine or from the chest drains.

As the heart recovers the amount of fluid given by mouth will be increased.



Surprisingly, Charlie's main concern post-op was his thirst, rather than pain from his scar or chest drains. Ice pops and pieces of cucumber were real lifesavers until his fluid limit was increased!



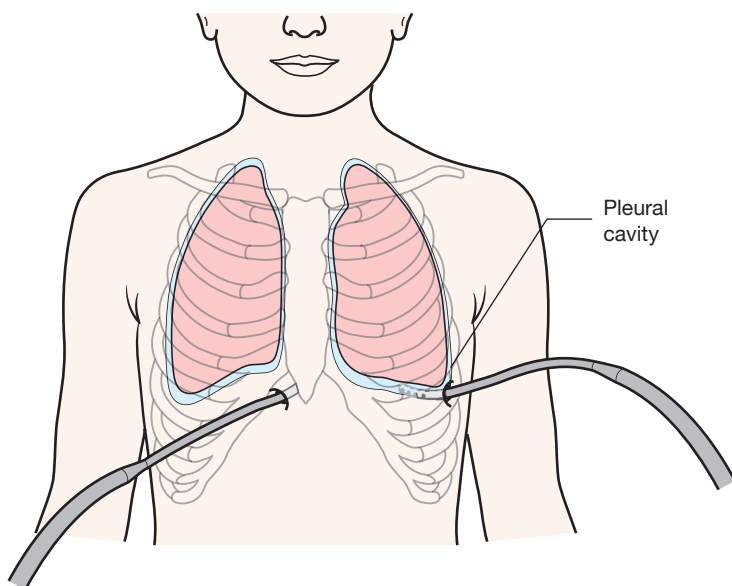
## Chest drains

As with all chest operations there will be a number of chest drains placed in the chest to help remove any fluid (blood, plasma and/or lymph) that collects around the operation site and to help re-inflate the lungs (after chest surgery the lungs collapse and need to be re-inflated, this is normal).

## Why do you have to remove the fluid?

There should be no air and no fluid in the space around the lungs. This empty space (vacuum) is there so that there is no resistance when the lungs fill with air. If there was a lot of fluid in the lung space or even air, the lungs would be pushing against the resistance because they are in an enclosed space and the lungs would be competing with the air or fluid for the same room.

This is why the drainage bottle contains water and the drains are often clamped if the child is moving around to help prevent the movement of air into the lung space.



## Why does fluid collect in the chest?

After all types of surgery there will be some bleeding so blood will form part of the chest drainage. The Fontan procedure (TCPC), dramatically changes the blood circulation. It takes time for the heart and the body to get used to the new way that the blood is pumped around the body.







This high back-pressure (described on page 11) causes clear fluid from the blood to ooze out into the chest cavity (the pleural space) around the lungs. It can take a number of days, if not weeks, for the oozing to settle down so the chest drains will stay in until little or no fluid drains from them.

This does not stop the children from getting up and playing carefully around the ward.

Sometimes the fluid draining from the chest contains fat (lymph). Throughout the chest there are little tubes like veins that carry fat around the body, the lymph system. If the pressure within the chest area is high after surgery these vessels can leak fat into the chest, a chylothorax.

If this problem happens the children will be started on a strict reduced-fat diet. In some hospitals the children are started on this special diet routinely after surgery to help prevent the fat from leaking. For more information on chylothorax and the MCT diet see LHM's *MCT diet* booklet.

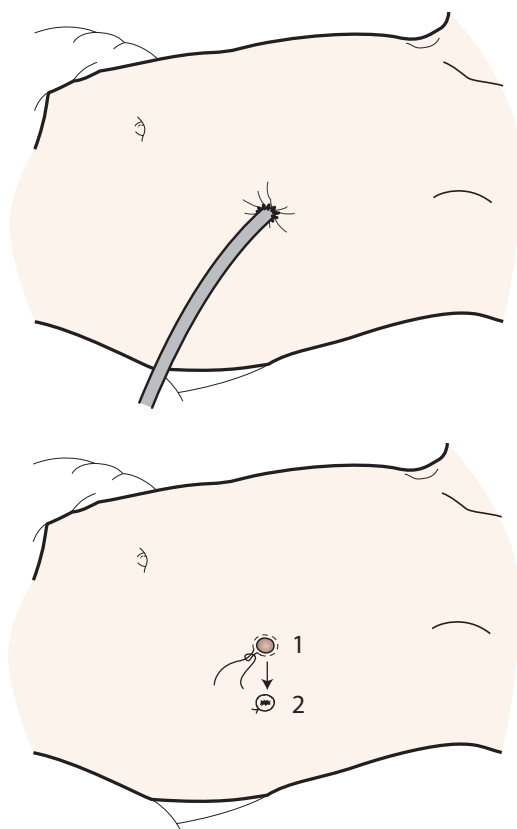
As soon as the medical team are happy that the chest fluid has gone and that there is no more drainage they will remove the chest drains.

## Chest drain removal

Chest drains are taken out on the ward by senior nurses or a doctor. Added care must be taken over the removal of these drains because they are in the chest cavity which should be completely empty.

To protect the empty lung space from being filled with air the surgeon will have placed a special set of stitches around the chest drain, a purse string suture. When the team take the chest drain out they also pull the purse string suture tight to close off the hole.

### Purse string suture



The children may be given some pain relief or mild sedation to help keep them calm through the drain removal but it is also useful for a parent to keep them distracted by chatting through the procedure. Ideas about what games can be played once the drains are removed helps to make the procedure more positive.

The nurses will observe the chest drain wounds. If there is any sign of redness, or the wound leaks, tests will be done and treatment, normally with antibiotics, will be started.

Stitches will be removed after six days but the timing depends on how well the wound has healed.

## Pacing wires

Whenever surgery is done on the heart there is a possibility that the rhythm, or beat of the heart will be disrupted, causing an irregular, fast or slow beat (an arrhythmia). So that any problems can be treated quickly the surgeons place two pacing wires into the heart during surgery. These blue wires can be used to pass information to the heart from a pacing box that is outside the chest (external pacing). This means that any problems with the heart rhythm can be treated swiftly without the need for further surgery.

Most problems with the heart beat (arrhythmias) settle as the heart recovers.

The pacing wires will be removed as soon as the heart rhythm is steady.

## Naso-gastric tube

Immediately after surgery there will be a tube passed down through the nose into the stomach. This tube gives the nurses access to the content of the stomach. This is important because although the child will not be eating when they first

return from theatre their stomach will still be making acid. To prevent the child being sick it is important that the nurses can remove the acid through the tube.

Once the child is fully awake and they have begun to take sips of water the naso-gastric tube will be removed and small amounts of food will be offered.

# Medications after surgery

## Pain relief

It is extremely important that a child who has had chest surgery has as little pain as possible after the operation.

Children who have pain will not settle because they are unhappy; this affects sleep and recuperation. Pain also affects the way that the children move around after the operation. It is very important that they sit up, breathe easily and cough when asked to. It is also important for them to have confidence that they can get out of bed and walk. All of this movement helps them recover, creates better circulation of blood and can also help prevent chest infection.

Immediately after the operation most children will have a Morphine infusion. This is a drip that very slowly gives a small dose of Morphine to the child. This keeps them pain-free but can leave them feeling sleepy and sometimes a little sick. They may be prescribed a medication to settle their stomach (an anti-emetic).

Gradually, as they recover, the medication will be reduced (weaned) so that the child takes medication by mouth, either Codeine or Paracetamol.

## Medications that support the heart

When the children are moved from the operating theatre to the PICU they will have been started on many infusions of drugs to support the heart.

After most operations you would rest the part of the body that had been operated on, gradually making it work harder and harder as it recovers. You can't rest the heart in the same way because you need it to beat so it is rested by supporting its work with medications. Dobutamine, Dopamine, Milrinone and Adrenaline are some examples.

These drugs are gradually weaned down as the heart function stabilises and strengthens.

The children may also be on medication to help regulate how much fluid they have circulating. Diuretics like Furosemide may be given to help the kidneys work more efficiently and pass out more urine. These medications will be used in partnership with the fluid restrictions described on page 23.

A very careful balance will be kept between keeping fluid levels low so that the heart function is not strained and the child's blood pressure being too low because there is not enough liquid in the blood.

Some medications may be needed to help stabilise the heart's rhythm as it settles after the operation. These medications are used at the same time as the pacing wires.




## Antibiotics

It is very important that children who are having surgery are protected from infection as much as possible. They will be given antibiotics during their recovery period.

## Anticoagulation

The Fontan surgery creates a new passage for blood to pass from the body to the lungs. The surgery involves a great many stitch lines. These create areas in the heart where blood cells can stick and make clots within the heart or blood vessels. To ensure that the blood flows smoothly through the heart and blood vessels, the children are prescribed anticoagulant therapy. Anticoagulants alter the way that the blood clots by slowing the clotting process.

This process will be started via veins (intravenously) with Heparin. Once the right level of anticoagulation has been reached the children will be changed onto an oral medicine. This could be:

-  Warfarin permanently.
-  Warfarin for a short period and then changed to Aspirin.
-  Just Aspirin.

For more information on these medications see the LHM *Living with anticoagulation* booklet.



# Observations done in PICU and beyond

When a child first returns to PICU after an operation the doctors and nurses will be carefully monitoring a number of things so that they can balance medications, ventilation and fluid.

Most measurements can be taken through a special arterial line (central line) that is passed into the heart and secured there during surgery. The arterial line measures the pressures within the heart and the pulse rate.

Oxygen saturations are measured both with a monitor and by checking the levels of gas in the blood.

Urine is measured by recording all input and output so the children have a urinary catheter passed into their bladder.

Temperature is measured with a temperature probe. An increase in temperature may be a sign of infection.

They will also have post-surgery echos (scans) to check that the blood is circulating correctly and that the heart muscle is recovering.

The nurses and surgeons will look at the scar to make sure that it is healing well and that there are no signs of infection.

Gradually, as the child shows that they are recovering, they will have these measuring devices removed.

Following a Fontan procedure the children do not stay in the PICU for very long, unless they have had a problem. Once the ventilator is removed and the heart is showing that it no longer needs the intravenous medications to support the heart function, the drips and the arterial monitoring line will be removed. Most children are transferred back to the ward the day after surgery.





# Time on the ward

The time in the PICU may be short but it may take a little longer to recover on the ward.

Every child will be nursed in the high dependency area of the ward after they are discharged from the PICU. Regular monitoring will continue, as will strong pain relief and fluid restrictions.

In many units the chest drains may stay in place for a minimum of five days after surgery because it takes time for the fluid in the chest to drain away.

It is really important that the children get out of bed and move around the ward as early as possible. As long as their pain relief is good and their heart is recovering there is no reason that they should not gradually get up and about, doing a little more every day.

Gradually the monitoring of the heart and the pain medication will be reduced. The children will be able to drink more, drips (infusions) will be removed and the child will be happy to eat more.

Once the doctors are happy that the chest drainage has reduced the child will have a chest x-ray and if all the chest fluid has gone the chest drains will be removed. If the chest fluid is still collecting the drains will remain in place.







Some children take some weeks before the fluid levels reduce. In some units the doctors will do a cardiac catheter to investigate an ongoing fluid collection. Samples of the fluid will be tested to check to see if it contains any fat; if it does then a diet low in fat may be prescribed. For more information on the MCT diet see LHM's *MCT diet* booklet.

## Keeping the children happy on the ward

Most children have their Fontan operation between the ages of three and six. Although they will be able to understand some of the information they were given about being in hospital before they had the operation, they may not remember what it is like to have drips and drains and be uncomfortable in bed because last time they had surgery they were probably a baby.

Keeping the children happy and occupied is a parent's most challenging job.

Planning the job before going into hospital can be a really helpful thing.

-  Pack up boxes of toys and games that can be swapped when visitors come into the hospital.
-  Bring a selection of DVDs and computer games that can be used when the children are tired.
-  Pack favourite books and stories - sometimes it is helpful to bring ones that the child has just grown out of as it is often comforting to hear a well-loved story.
-  Plan hospital visitors who can give parents a break from the bedside to have a meal or a shower. Grandparents are invaluable here as they will read to a child and play games with them as they would do at home.
-  Plan small treats that can be given every day - these do not have to be big, e.g. a small packet of sweets, a small car, paper plane kits or sewing. If these are presented all wrapped up as a pick and mix, it is exciting for the child.
-  Plan what presents visitors bring - games, books and colouring are better than an active toy that the child may not be able to play with yet.





We brought a lucky-dip bag with lots of small, wrapped presents with us to the hospital.

Little things like a whoopee cushion or action figures, and every time Charlie was brave (or fed up!) we'd let him have a dip in the bag. It really helped to keep him focused when he was reaching the end of his tether.

We also took books and DVDs with us - some of which were normally too young for him, but the familiarity of the old programmes was comforting when he was still very dozy on Morphine. We knew he was on the mend once he wanted Dr Who and Harry Potter instead of Tom and Jerry.



If a child is in hospital for a number of weeks the hospital school and play team will organise as much activity as possible to keep them busy. They can cook, paint, build and explore with drips and drains attached.

Once they are free of drains and drips they will be able to visit play departments and even go outside for short periods.








While Charlie was recovering, we found that short bursts of different activities were ideal to keep him entertained. During the week, the play team at the hospital organised baking, painting, singing and games, which he could join in with as and when he wanted.



## How long will we be in hospital?

Every child's length of hospital stay will be different. It is always wise to plan to be in for about three weeks. Some children will be discharged home before then but others may need to be in for longer, especially if they continue to have fluid drainage into their lung space.

Children will be discharged home once the doctors are happy that their:

-  heart function is good and recovering
-  fluid balance is controlled
-  pain has gone and they are moving with ease
-  anticoagulation is stable and the measurement of the level of medication is balanced
-  wound is healing and not infected.





# Going home

## Medications to take home

Each hospital chooses to use different medications for each child but most children are on some medication when they go home.

### Anticoagulation

Nurses and the haematology team will explain about anticoagulation and the measurement of drug blood levels before discharge.

### Heart medication

Many children are on ACE inhibitors like Captopril or Lisinopril, medications that relax the blood vessels so that the heart has to work less hard to push blood around the body.

### Diuretics

Medications that help the body pass out urine; this takes strain away from the heart by reducing the amount of fluid in the blood.

### Anti-arrhythmics

Will be prescribed to any child who needs support to keep the heart in rhythm.

## What should I be looking for when I take my child home?

Parents should be reassured that if their child is discharged home, the doctors are happy that their heart is recovering well and that they can cope with the journey home.

There are some things that parents are told to watch for at home.

### Fatigue

When your child gets home from hospital they will probably try to do too much because they are delighted to be in their own



safe environment. This, mixed with the exhaustion created by surgery and the journey home, may make them more tired than when they were in the hospital. Gradually they will have more energy and be able to do more and eventually they will be able to take part in more physical activity than before the surgery. If they do not show signs of gradual recovery, ask your specialist nurse for advice.

## Colour

Children usually have a much pinker colour after the Fontan operation. Some children who have had a fenestration during surgery may have a slightly dusky look because they still have a mixture of blue and red blood circulating around their body. If the child becomes cold or if they feel ill they may have a bluer colour than normal for a short time. However, if your child becomes very blue and does not return to the new pink colour quickly, you should ask their medical team for advice through your specialist nurse or cardiologist. If your child is very blue and breathless, phone for an ambulance.

## Eating and drinking

Gradually the children should start to eat and drink as normal. Some children find that the improved energy levels created by the operation mean that they want to eat more but it would be unusual for them to want very large meals. Small meals spread throughout the day help them to maintain their new-found energy. If your child does not recover their appetite over the first few weeks at home then speak to the doctor at the first hospital appointment after discharge.

## Wound

Gradually the wound/scar will begin to heal. The stitches from the main scar will gradually dissolve as the wound heals. The stitches from the chest drains will be removed at about six days after the drains were removed. All of the scars will gradually fade over six months. If the wound gapes or is leaking fluid, ask your GP for advice or ring the specialist nurse.

## Breathlessness

Although the children will gradually gain more energy and be able to take part in many more activities, they will never be able to keep up with other children of their age as they still only have half a working heart. Short periods of breathlessness during and immediately after exercise will be normal. Parents should only become worried if they see that their child does not recover their breath or that they are unable to maintain a conversation because they are breathing with difficulty. If these episodes happen occasionally it will be important to raise the matter at the next outpatient visit. If breathlessness is happening all the time and the child is getting blue and sweaty, seek SWIFT medical support either by taking your child to the local hospital or, if very serious, by calling an ambulance.

## When will we see the doctors again?

How well your child was when they were discharged from hospital determines when the doctor will want to see them again at the hospital.

If they have had a slow recovery they may need a check-up a week later; if they have recovered very quickly then they may not be seen for a few weeks.

If the children have been prescribed Warfarin then they will need regular blood tests to measure if the medicine is working the way that the doctors have planned. This is usually done at the local hospital or on some occasions at the GP practice. Initially these tests will happen more than once a week but gradually as the medication levels settle they will be done once a week and then less frequently. Parents can be trained to test the blood levels at home using an INR machine. See LHM's *Living with anticoagulation* booklet.

## When can the children go back to school?

This is a question to ask your doctor before discharge and again at the first clinic appointment after the operation.



Some children return to school three weeks to a month after they have gone home from the hospital. It can be really useful if they start to make short visits back to their school with a parent or relative, perhaps to take part in storytime or a free play session.

Once the doctors are happy that their wound has healed and their heart is working well they will allow the child to gradually start up school again. Some children have so much more energy than they had before the operation that they return to school with great enthusiasm. Others need to gradually take part in school life again - perhaps short days to start with, gradually building towards full days over a couple of weeks.

## Life after a Fontan

The aim of the Fontan procedure is to give a child as much energy as having only half a pumping heart will allow. If surgery has gone as planned the children will gradually recover from the operation and go on to gain far more energy than they had before.

They will:



enter into playground games with far more energy



eat with more gusto



take part in the skills part of a PE lesson



have more confidence with their friends; and



they will look pink for most of the time.



# Key terminology

## The Fontan operation

The Fontan operation has three potential names:

- The Internal, Classic Fontan
- The External, Conduit Fontan also known as the Total Cavopulmonary Connection or TCPC
- The Hemi-Fontan is the name given to one type of stage two surgery. It is a stepping stone to the full Internal Fontan procedure.

## The Fontan circulation

The name given to the way that blood circulates around the body after a Fontan operation.

## Fenestration

A hole created to relieve back-pressure within the Fontan circulation (see page 11).

## Back-pressure

Blood returning from the head and neck and body to the lungs to pick up oxygen does not have a pump behind it so travels slowly. Sometimes this creates a pressure build-up within the blood vessels, creating pressure on the organs further down the body, for example, the liver and gut.

## Ventilator

The machine connected to a patient who needs to be ventilated, i.e. have the breathing done for them. This is a very common way to rest a patient after an operation.

## Echo - Echocardiogram

A diagnostic test that uses sound waves to show organs in the body.

## MRI - Magnetic Resonance Imaging

A scan that uses magnetic impulses to show organs in the body.

## Gore-Tex®

A waterproof, breathable fabric membrane created in patches and tubes.

# Further information

## Little Hearts Matter

Other booklets available from LHM which may be useful:

- Preparation for Hospital
- MCT Diet - information booklet for parents
- Living with Anticoagulation

Tel: 0121 455 8982

[www.lhm.org.uk](http://www.lhm.org.uk)



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It is hard to express just how big a difference Charlie's Fontan has made to his life. He no longer needs his buggy and can walk (and walk!) without getting puffed and blue. He eats SO much more quickly - and for the first time in his life complains about being hungry! He is aware of the difference in himself since his op, and takes great pleasure in being able to join in everything with his friends.  
We are SO proud of him.

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# Acknowledgements

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David Beattie M.Phil.; MITI
- Medical illustrations by: Jane Fallows  
Medical Illustrator
- Disclosure: Dr David Crossland has no conflict of  
interest

We would like to thank members of our reader panel for reviewing this publication and sharing their experiences.

We are very grateful to Charlie Turner and his parents Alison and Pete for sharing their Fontan experience with us in this publication.

References are available from the LHM office on request.





## Little Hearts Matter

Supporting every step of the half a heart journey

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LHM FPB 0819

Review date 0821