Great Ormond Street Hospital for Children NHS Foundation Trust: Information for Families

Central venous access devices for children with lysosomal storage disorders

This information explains about central venous access devices, particularly for children with lysosomal storage disorders. Some lysosomal storage disorders can be treated with enzyme replacement therapy. This needs to be given directly into the blood stream. To do this, we will put a central venous access device into one of your child's veins. This gives easy access to your child's bloodstream so that your child can have enzyme replacement therapy.

What is a central venous access device?

A central venous access device (CVAD) is made from a non-irritating material such as silicon and titanium, which means that it can be left in place in a vein for long periods of time.

What are the advantages of having a CVAD?

Whatever the type of CVAD, it can be used to give your child's treatment. It may also be used to take blood samples, although some blood samples will still need to be taken directly from another vein.

What are the disadvantages of having a CVAD?

With any CVAD there is a risk of infection. which may cause a blockage. There is also a risk of a clot forming around the tip of the device. There may also be times when it is not possible to take blood samples from the CVAD.

What types of CVAD are available?

There are three main types:

- An implanted port
- A peripherally inserted central catheter (PICC)
- A skin-tunnelled catheter

Please note: Skin tunnelled catheters are not used to give enzyme replacement therapy so are not covered in this booklet.

Are there any risks involved in inserting a CVAD?

Occasionally there can be complications when the CVAD is inserted. The needle or guide wire used to insert the catheter could scratch the top of the lung causing a pneumothorax (air pocket). This shows up on an x-ray and would be treated immediately, but would not cause any long-term problems. The catheter may thread into the wrong position, but the position of the catheter is checked with an x-ray during or after insertion. Your child may have some bruising and feel a little sore where the CVAD was inserted but this soon improves.

Recognising early signs and symptoms of complications

It is very important that you learn to recognise the early signs and symptoms of any complications.

They are as follows:

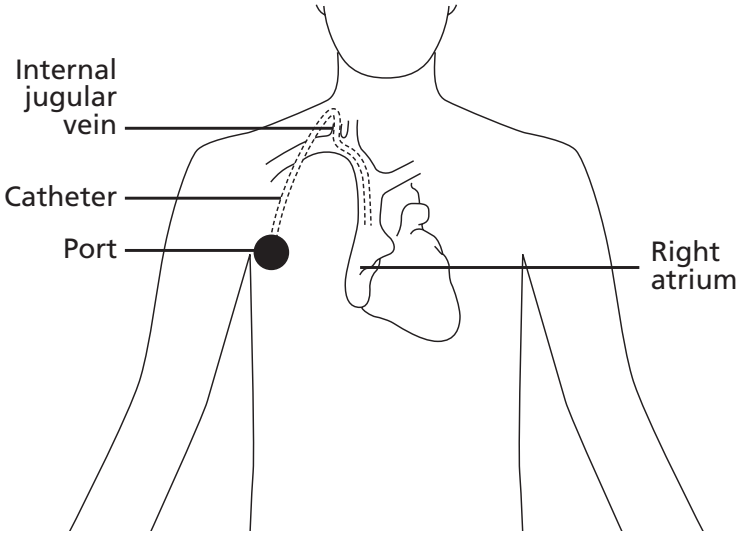
- Your child develops a fever, chills or flu-like symptoms – this could be an infection
- Your child has pain or a burning sensation while injecting
- Your child's arm, neck or shoulder is painful and swollen – this could be a sign of a blood clot
- Your child has a sudden onset of chest pain or difficulty in breathing
- You cannot flush your child's catheter easily or you feel resistance – this could be a sign that the catheter is blocked
- You think that your child's CVAD site looks red and inflamed and/or there is any discharge and/or redness tracking across the chest (implanted port) or up the arm (PICC)
- Your child's PICC is leaking under the dressing
- Your child's PICC is partially pulled out – do not try to push it back in as the tip will not be in the correct place
- Your child's PICC is completely pulled out – apply pressure to the area and take your child to the nearest Accident and Emergency (A+E) department

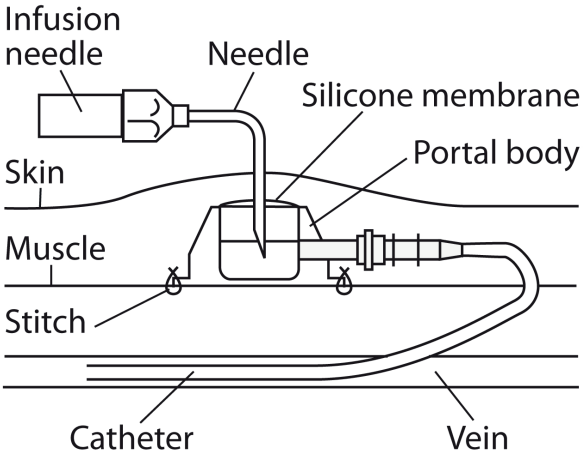
When should I call the homecare company, local hospital or GOSH?

You must contact a healthcare professional if you have any concerns whatsoever. It is better to talk them over with us just in case than have your child rushed to hospital as an emergency.

Useful numbers

GOSH Clinical Nurse Specialists for Lysosomal Storage Disorders	020 7405 9200 ext 0366
Homecare Nurses	
Family Doctor (GP)	
Local hospital children's ward	
Accident and Emergency (A+E) department	





Implanted port

What is an implanted port?

An implanted port is a type of CVAD that is inserted under the skin, usually on the chest. The port is made up of a portal body, which is connected by a catheter inserted into one of the veins. The port can be felt through the skin. The port is accessed by inserting a special type of needle called a gripper needle through the skin and silicon membrane into the portal body.

Inserting the needle is similar to pricking the skin with a pin. Naturally it takes some time to get used to but local anaesthetic cream can be used to numb the skin if needed.

What are the advantages of an implanted port?

The gripper needle only needs to be inserted for each treatment. The needle is removed between treatments and no dressing is required. The port is not visible. Normal activities, except contact sports, are not restricted.

What are the disadvantages of an implanted port?

Your child will need to have a gripper needle inserted for each treatment. Inserting and removing the port has to be done in the operating theatre under general anaesthetic. There will be a small scar where the port was inserted and removed. It is not possible to take some blood samples from a port. For example, samples to check clotting as a heparin solution is used to flush the port, which causes abnormal test results.

How is the port inserted?

Your child will need to come into hospital for an overnight stay. A surgeon inserts the port usually, under general anaesthetic. Two small cuts are made, one forming the 'pocket' where the port sits and the other for the catheter to be inserted. The stitches over the pocket are dissolvable as are the stitches at the catheter site.

How do I care for the port?

No special care is needed for a port. The needle is removed in between treatments and your child will not need any dressings. The only restriction on activities is contact sports, such as rugby, which should be avoided in case the port is damaged.

How do I access the port?

Only when trained to do so.

If you are using local anaesthetic cream, apply it to the skin over the port as you have been taught and leave for 30 minutes to an hour to numb the area.

You will need:

- Dressing pack
- Dressing towel
- Sterile gloves
- Gauze
- Chlorhexidine solution
- Disposable pot
- Gripper needle
- Normal saline solution
- Transparent dressing

1 Collect together all the equipment needed on a clean surface
2 Wash your hands thoroughly with soap and water
3 Open the dressing pack
4 Wipe off the local anaesthetic cream (if used)
5 Pour some chlorhexidine solution into the disposable pot
6 Place the dressing towel over your child's chest and put on the sterile gloves
7 Using the non-touch technique, carefully clean around the area with gauze dipped in chlorhexidine solution. Use a circular motion from the port outwards and allow the area to dry. Repeat twice more.
8 With your non-dominant hand (left hand if you are right handed and right hand if you are left handed), feel for the edges of the port
9 With your other hand, insert the gripper needle into the centre of the port as you have been shown
10 Use 4ml of normal saline solution to check that you can access the port
11 Press a transparent dressing over the gripper needle to keep it in place during treatment.
12 Start treatment.

Flushing the port

Only when trained to do so.

The port must be kept clear by injecting it first with normal saline and then 2ml (100iu/ml) strength heparin solution. After every treatment, you will use 5ml normal saline and 2ml heparin solution.

You will need:

- Two syringes and green needles
- Ampoule of heparin solution
- Ampoule of normal saline
- Dressing towel
- Sterile gloves
- Large chlorhexidine wipe
- Gripper needle
- Sharps bin

1 Collect together all the equipment needed on a clean surface
2 Wash your hands thoroughly with soap and water
3 Using the non-touch technique fit a green needle onto the end of each syringe
4 Break the heparin ampoule as instructed
5 Draw up the heparin into one syringe and push the plunger a little to get rid of any air in the syringe
6 Draw up the normal saline into the other syringe and get rid of any air as above
7 Place the dressing towel over your child's chest and put on the sterile gloves
8 Clean the connection between the treatment line and gripper needle with the chlorhexidine wipe and allow it to dry

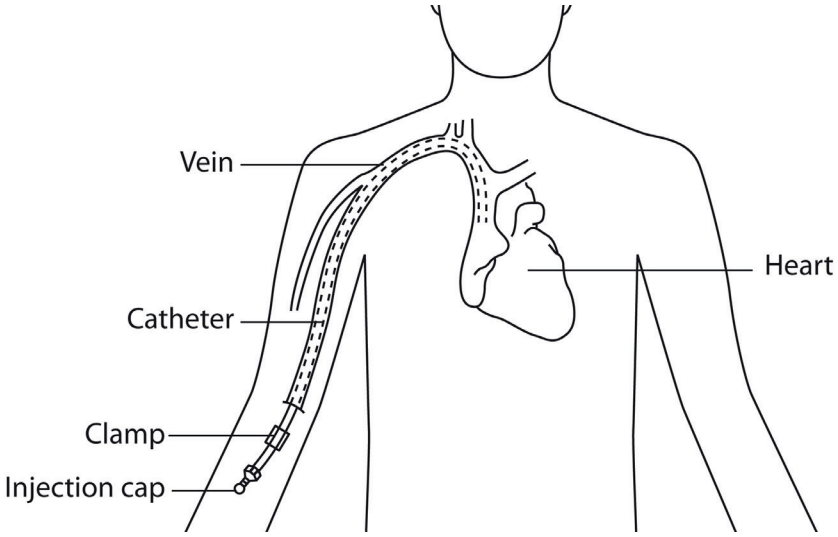
9 Clamp the gripper needle line and disconnect from the treatment line
10 Attach the normal saline-filled syringe to the gripper needle and inject slowly, 1ml at a time
11 Disconnect this syringe keeping pressure on the plunger
12 Attach the heparin-filled syringe and inject slowly, 1ml at a time
13 Disconnect this syringe keeping pressure on the plunger and re-clamp the gripper needle line
14 Peel back the transparent dressing and remove the gripper needle, holding the port in place
15 Dispose of the used needles and syringes in the sharps bin

What equipment and training will I receive?

All the equipment you require will be provided by the homecare company, as well as any training you need according to an approved training plan.

How is the port removed?

Your child will need to come to hospital as a day case as it is removed under general anaesthetic.



Peripherally inserted central catheter

What is a peripherally inserted central catheter (PICC)?

A peripherally inserted central catheter (PICC) is a tube inserted into a vein in your child's arm, at or above the bend in the elbow. It is then fed through the vein into a larger vein leading to your child's heart.

What are the advantages of a PICC?

The PICC can be inserted under a local anaesthetic so your child may not need to go to the operating theatre. The PICC will not leave any scars when it is removed. There is a lower risk of complications when the PICC is inserted compared with other types of CVAD.

What are the disadvantages of a PICC?

The dressing covering the PICC needs to be changed once a week. The PICC is visible so your child might feel self-conscious. Your child will not be able to go swimming with the PICC in place and it may restrict vigorous sporting activities.

How is the PICC inserted?

A PICC may be inserted under a local anaesthetic, or he or she could have it inserted under general anaesthetic as a day case. A nurse will put local anaesthetic cream on the skin over the vein for 30 minutes to an hour and then wipe it away. The cream makes the skin over the vein numb so inserting the needle will be less painful. An introducer needle is inserted into the vein and the PICC is threaded through it until the tip of the catheter is in the correct position. The needle is then removed leaving the catheter in place in the vein. It is sometimes stitched in place, then covered with sterile tape and a dressing. Extra padding is put over the area for the first 24 hours to reduce any bleeding.

How do I care for the PICC?

Initially, the dressing should be changed by a healthcare professional within the first 48 hours. After this, either a nurse can change the dressing every week, or we can teach you to do it. The transparent dressing over the exit site, where the catheter comes out of the skin is waterproof. However, your child should avoid getting the dressing wet, as it needs to stay clean, dry and sticking firmly to the skin. When bathing or showering, your child should keep the site covered, either with a plastic bag or cling film. During the first week after the catheter insertion, the exit site may look red, inflamed and a little sore. This should ease after the first week but if it continues, please contact us.

Changing the PICC dressing – only when trained to do so

You will need:

- Dressing pack
- Dressing towel
- Chlorhexidine solution
- Disposable container
- Gauze

1 Collect together all the equipment needed on a clean surface
2 Wash your hands thoroughly with soap and water
3 Place the dressing towel under your child's arm
4 Pour some chlorhexidine solution into the disposable pot
5 Carefully peel off the old transparent dressing and dispose of it
6 Wash your hands thoroughly with soap and water again
7 Using the non-touch technique, carefully clean around the catheter with gauze dipped in chlorhexidine solution. Use a circular motion from the catheter outwards and allow the area to dry. Repeat if necessary.
8 Open the transparent dressing
9 Straighten your child's arm and put the dressing over the exit site, pressing it into place
10 Put a piece of gauze on the skin above the exit site and curl the remainder of the catheter on top of it to stop it digging into your child's skin
11 Put another piece of gauze on top and cover with a bandage, taking care not to cover the transparent dressing with the bandage.

Flushing the PICC – only when trained to do so

The PICC must be kept clear by injecting it with either heparin solution or normal saline once a week after the dressing change. The type of solution to be used is dependent on the type of tube inserted. In addition, the needle free cap on the end of the PICC will need to be changed every week.

You will need:

- Syringe and green needle
- Ampoule of heparin solution or normal saline
- Large chlorhexidine wipe
- Needle free cap
- Sharps bin

1 Collect together all the equipment needed on a clean surface
2 Wash your hands thoroughly with soap and water
3 Using the non-touch technique fit the green needle onto the end of the syringe
4 Break the heparin or normal saline ampoule as instructed
5 Draw up the heparin or normal saline into the syringe and push the plunger a little to get rid of any air in the syringe
6 Clean the connection between the cap and catheter with the chlorhexidine wipe and allow it to dry
7 Attach the new needle free cap to the end of the catheter

8 Insert the heparin- or normal saline-filled syringe into the cap and inject slowly, 1ml at a time

9 Remove the syringe, keeping pressure on the plunger

10 Dispose of the used syringe in the sharps bin.

What equipment and training will I receive?

All the equipment you require will be provided by the homecare company, as well as any training you need according to an approved training plan.

How is the PICC removed?

Your child will be able to have the PICC removed during a visit to hospital, but special arrangements do not usually need to be made as it is similar to having a cannula removed. The catheter is gently pulled out of the vein and a pressure dressing is put over the area to reduce bleeding and bruising. This dressing can usually be removed after 24 hours.

Compiled by the Metabolic Clinical Nurse Specialist for Lysosomal Storage Disorders
in collaboration with the Child and Family Information Group

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