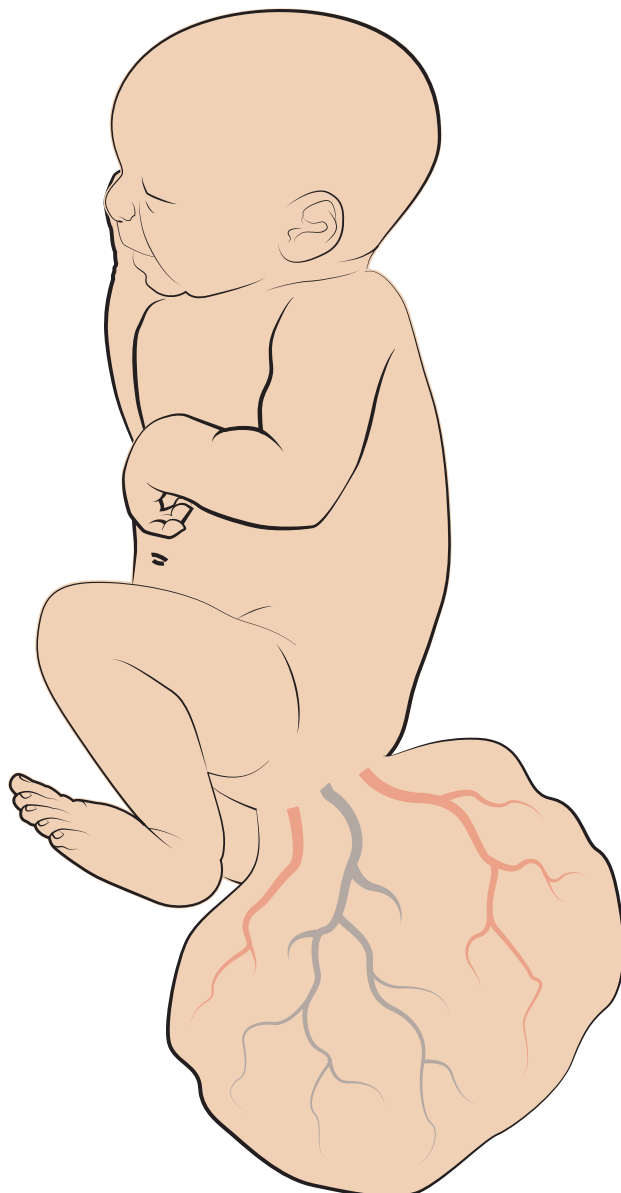




Great Ormond Street Hospital for Children NHS Foundation Trust: Information for Families

Sacroccygeal teratoma removal in children

This information sheet from Great Ormond Street Hospital (GOSH) explains about sacroccygeal teratomas (SCT) in children, the operation to remove them and what to expect when your child comes to GOSH for treatment.



What is a sacroccygeal teratoma?

A sacroccygeal teratoma (SCT) is a congenital (present at birth) growth or tumour that develops at the base of the spine just above the buttocks. It is the most common neonatal (newborn) tumour, affecting about 1 in every 40,000 babies born. An SCT can grow to quite a large size while the baby is in the womb, sometimes a similar size to the baby.

An SCT can be made up of fluid-filled cysts (sacs) or solid tissue or a mixture of both with its own blood supply. The growth is usually covered by skin but occasionally it might only be covered by a thin, see-through membrane.

In the majority of cases, SCTs are benign (not cancerous) but in a small number, they may be found to be cancerous (malignant) when examined in a laboratory. SCTs can sometimes be associated with other congenital problems.

There are four types of SCT, depending on the location of the growth:

- Type I is where nearly all the tumour is outside the body
- Type II is where the tumour is mainly outside the body but a small part may be in the pelvis or abdomen
- Type III is where the tumour is mainly inside the pelvis or abdomen but a small part may be outside the body
- Type IV is where the entire tumour is inside the pelvis or abdomen

All forms can be removed surgically, but treatment may be more complicated in types III and IV SCT or growths that have grown very large.



What causes a sacrococcygeal teratoma?

A teratoma is a type of tumour that arises from stem cells. Stem cells are the 'blank' cells that can turn into any type of specialised tissue within the body. This means that teratoma tumours can be made up of any type of specialised tissue from anywhere on the body, such as skin, hair and teeth. We do not know what causes the stem cells to develop abnormally and at a faster rate than usual. SCTs seem to be more common in females than males but we do not know why. Doctors do not think it was caused by anything you did or did not do during pregnancy.

How is a sacrococcygeal teratoma diagnosed?

A sacrococcygeal teratoma might be suspected first if it shows up on a routine prenatal ultrasound scan. Occasionally, additional more detailed scans might be suggested if the uterus (womb) is larger than expected for the stage in pregnancy or if there is more amniotic fluid surrounding the baby in the womb. Once an initial scan has raised suspicion of an SCT, regular scans will be carried out during pregnancy to monitor the baby's health as well as the size of the SCT.

What are the signs and symptoms of a sacrococcygeal teratoma?

Early in pregnancy, there might not be any specific symptoms. Later on, the mother's bump may be larger than expected for the stage in pregnancy due to the increased size of the baby and an increase in the amount of amniotic fluid surrounding the baby in the womb. As the teratoma has a blood supply, the baby's heart may have to work harder than usual to pump blood around the body, which can lead to hydrops or an accumulation of fluid in the baby's body. This can quickly become serious and require early delivery of the baby. Sometimes the hydrops is so severe that the baby cannot survive and is stillborn.

After the baby is born, in most cases, the teratoma will be immediately obvious. The growth can be quite large, sometimes as large as the baby itself, and covered in skin or a see-through membrane. The growth may feel hard to soft depending on whether it is made up of cysts or solid tissue or a mixture of both. A baby's heart may be tired if it has had to pump harder than usual during pregnancy so feeding may be difficult.

If the teratoma is entirely inside the pelvis or abdomen, it might not immediately obvious but suspected when the baby does not pass urine or faeces as expected due to the pressure of the growth on the internal organs.

How is a sacrococcygeal teratoma treated?

The delivery of a baby with a sacrococcygeal teratoma will need to be planned carefully ahead of the due date. In most cases, babies who are due to be treated at GOSH tend to be delivered at University College London Hospital (UCLH) and transferred here in the first few hours after birth. The best option for delivery may be a caesarean section to decrease the risk of damage or bleeding from the tumour. Your midwife and obstetric team will discuss this fully with you before birth.

Once the baby is born, we will transfer them to our Neonatal Intensive Care Unit (NICU) at GOSH. To begin with, your child will be nursed in an incubator and will have a naso-gastric (NG) tube passed through their nose into the stomach. This will drain off the contents of the stomach and stop your child feeling and being sick. It also releases any excess air from the stomach, which could make your child uncomfortable. They will also have an intravenous infusion (drip) of fluids and medicines.

The surgeon may ask for imaging scans such as an ultrasound scan or CT or MRI scan to see the teratoma and its blood supply in more detail. The results of these scans will help to plan the operation to remove the teratoma. Blood tests looking for levels of specific proteins (AFP and BHFG) that may be secreted by the teratoma will be measured.



What happens before the operation?

When they are stable, your child will have an operation under general anaesthetic to remove the teratoma. The surgeon will explain about the operation in more detail, discuss any worries you may have and ask you to sign a consent form giving permission for your child to have the operation. An anaesthetist will also visit you to explain about the anaesthetic.

What does the operation involve?

When your child is under anaesthetic, the surgeon will make an incision across the buttocks around the teratoma and free the tumour from the surrounding tissue. If the teratoma is a type III or IV, they may need to make an incision on the abdomen too.

The surgeon will seal off the blood vessels feeding the teratoma as they are separating it from the normal tissue. When the teratoma is clear from the surrounding tissue, the surgeon will remove it along with the coccyx bone from the base of the spine. The teratoma will be sent to the laboratory for examination under a microscope.

When as much of the teratoma as possible has been removed, the surgeon will close the incision(s) with stitches and cover the area with a dressing.

Are there any risks?

For this operation, the main risk is bleeding so all children will have a blood test to 'cross match' their blood beforehand so that donated blood of the right type is available during the operation if it is needed. During the operation, the surgeon will minimise any bleeding by sealing off the blood vessels affected.

There is a chance that nearby structures in the abdomen could be damaged during surgery especially the nerves and muscles controlling the bladder and bowel. The surgeon will aim to remove as much of the teratoma as possible without damaging surrounding structures. The tumour itself may have already affected the nerves in this area and this will be monitored and reviewed during follow up throughout childhood.

Sometimes a small area of teratoma may be left behind and the treatment of this will vary from child to child.

Every anaesthetic carries a risk of complications, but this is very small. For more information about your child's anaesthetic, please ask your anaesthetist for a copy of our information sheet. Your child's anaesthetist is a very experienced doctor who is trained to deal with any complications.

What happens afterwards?

Your child will be transferred back to NICU for a few days to recover. They will continue to have breathing support from a ventilator but this will be gradually reduced over the next few days. The drips giving pain relief and feed will also remain for the next few days but will be removed when they are no longer needed. If the teratoma was very large and situated on your baby's back, the wound site and surrounding skin will be delicate and may need further dressings. While the area is healing, avoid putting pressure on the area so do not sit them up or keep them in the same position for too long. Ask for advice from your nurse if you have any concerns. The surgeons will let you know when you can start feeding your baby again. When your child is feeding well and gaining weight they could be transferred back to your local hospital to continue recovering or straight to home.



What is the outlook for children who have had a sacrococcygeal teratoma removed?

In the majority of cases, the teratoma is benign (non-cancerous) so no further treatment will be required. If any cancerous cells are found, our oncology (cancer specialist) team will be involved in further management and follow up. However, the surgical team will continue to follow up your child with blood tests and ultrasound scans. Initially this will be every three to six months and then less frequently. The risk of recurrence is greatly reduced by removing the coccyx at the time of surgery. Some children have ongoing bladder and bowel problems which may require treatment in the future. The scars from the surgery will fade gradually over time but will grow in proportion as your child grows. Cosmetic revision of the scars may be offered when your child is older if needed. Your child will need to come back to GOSH for regular check-ups throughout childhood and adolescence.

Further information and support

There is no support group in the UK specifically for people affected by a sacrococcygeal teratoma but the umbrella organisation Contact a Family may be able to put you in touch with another family. Call their helpline on 0808 808 3555 or visit their website at www.cafamily.org.uk