

# Cloacal Exstrophy

This leaflet is for parents or carers whose baby has recently been diagnosed with Cloacal Exstrophy. We understand that you may be confused and worried about what will happen. This information leaflet aims to support you by covering some commonly asked questions.

## What is Cloacal Exstrophy?

Cloacal Exstrophy is part of a spectrum of birth defects that affect the bladder called the Bladder Exstrophy & Epispadias complex, the urethra, the genitals and the pelvic bone. Epispadias is at the minor end of the spectrum with cloacal exstrophy being the most severe form, and Cloacal Exstrophy additionally affecting the bowel.

Cloacal Exstrophy is a very rare and complex abnormality and is estimated to occur in approximately 1 in every 200,00-400,000 live births. A baby born with Cloacal Exstrophy is born with the bladder and bowel on the outside, and the pelvic bones are open like a book. The anus is often not formed (also known as an imperforate anus) and the intestine can be short. All babies born with Cloacal Exstrophy will also be born with abnormal genitalia, which in boys means the penis is short, open and flat or in girls, the labia and clitoris are split, and occasionally two vaginal openings are present.

In approximately 75% of Cloacal Exstrophy cases, there are other birth defects such as Spina Bifida, Omphalocele (where the intestines are covered by only a thin layer of tissue) or kidney abnormalities.

## Why do I have to come to Manchester or London?

There are two specialist centres for children born with Cloacal Exstrophy in the United Kingdom, which are Royal Manchester Children's Hospital and Great Ormond Street Hospital in London. We have a vast understanding of the condition and experience in caring for these children. Our specialist team is made up of:

- Consultant Paediatric Urologists
- Consultant Paediatric Anaesthetists
- Clinical Nurse Specialists
- Consultant Orthopaedic Surgeons
- Clinical Psychologists
- Play Specialists

## What is Bladder Exstrophy, Epispadias and Cloacal Exstrophy?

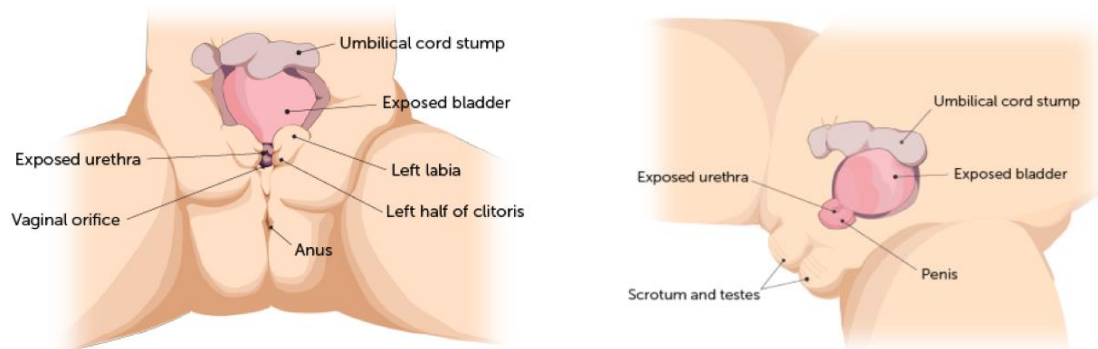
These conditions are part of a spectrum of birth defects, with Epispadias being at the minor end of the spectrum, and Cloacal Exstrophy being the most severe form. Approximately 40% of Bladder and Cloacal Exstrophy defects are detected on ante-natal scan, but sometimes Cloacal Exstrophy can be mistaken for Bladder Exstrophy. Therefore, if there is any suspicion of Cloacal Exstrophy, then a further scan called a “foetal MRI” is carried out whilst baby is still in utero to confirm the diagnosis.

**Classic Bladder Exstrophy** is an abnormality of the lower part of the tummy where the bladder, urethra (the tube that takes urine to the outside of the body), the pelvic bone and the genitals are affected. The bladder is seen on the outside of the baby’s tummy. The urethra is open along the top and is not formed into a cylindrical tube as it should be, and the tummy button is in a lower position. The pelvic bones do not come together completely at the front (this is known as pubic diastasis). In boys, the penis is tilted backwards and is often shorter and wider than normal. In girls the clitoris is split.

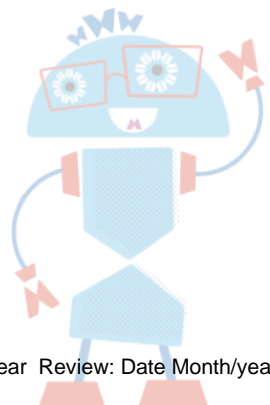
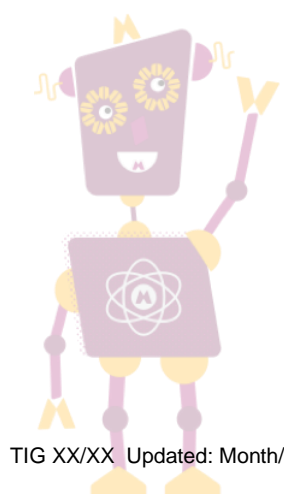
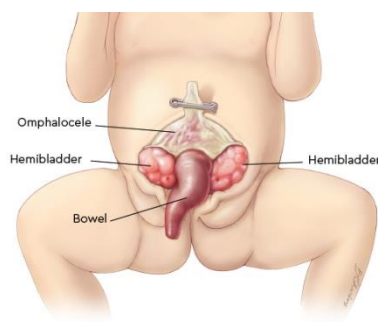
**Epispadias** is a less extensive form of the condition. The bladder is not visible outside the tummy but the urethra (the tube that takes urine to the outside of the body), is open on the top side and is not formed into the cylindrical tube shape as it should be. In girls, the clitoris and labia are split. Epispadias is always present in Bladder Exstrophy, however it is possible to see Epispadias on its own (without Bladder Exstrophy or Cloacal Exstrophy).

In **Cloacal Exstrophy**, the bowel and bladder are both seen on the outside of the baby’s tummy.

### Diagram showing female and male Bladder Exstrophy & Epispadias



### Diagram showing Cloacal Exstrophy



## Why has this happened?

It is not known what causes Cloacal Exstrophy. The problem occurs between the 4<sup>th</sup>-10<sup>th</sup> week of pregnancy when various organs, tissues and muscles begin to form. Cloacal Exstrophy **does not** occur because of anything the mother or father did or did not do during pregnancy.

Our specialists are working in conjunction with geneticists and researchers across the UK to try to ascertain if there is a genetic link that makes it more likely for a baby to be born with Cloacal Exstrophy, but so far results have been inconclusive. You and your baby may be asked to enrol in the study and we may ask permission to take blood samples for our data. If you are a suitable candidate for this study, this will be discussed with you by a member of the medical and genetics team when you come to clinic, and it is entirely your choice if you choose to enrol. Not enrolling in the study does not affect the level of care you and your child receive from our team.

## So what happens first?

Cloacal Exstrophy is corrected in a series of operations over the first years of life. The overall aim of treatment is to protect the kidneys, allow your baby to feed and grow normally, reconstruct the tummy wall and close the bladder and pelvic bones. When your child is older we consider the different options which will enable the child to be clean and dry. The reconstruction of the genitalia in boys is more challenging and is usually left until the child is older.

If your baby has been born with **Cloacal Exstrophy**, an operation to ensure that your baby can pass faeces will usually be performed immediately, or within a few weeks of life. During this operation, the Doctors will put the bowel back inside and make a small hole on the tummy (called a stoma) through which your baby will pass faeces (poo). You will meet with the Consultant Paediatric Urologist and our specialist team who will explain the operation. The anatomical abnormalities to the urethra, genitalia, pelvis and bladder will not be corrected within the same operation.

The operation to close the bladder and tummy and reconstruct the pelvis is done once your baby is a little bit older, usually around 9-12 months of age. This gives your baby the time to grow and mature, making surgery much safer and more successful. You will be shown how to take care of your baby's tummy and stoma before you are discharged home.

In girls the genitalia will look close to normal after the 2nd operation to close the bladder and the pelvic bones. When girls are older our gynaecologist, will be involved to ensure girls have normal periods and when older they may need adjustment of the vagina to enable sexual relationships. The reconstruction of genitalia in boys with cloacal exstrophy can be very challenging and penile reconstruction is best left until the boy is older or even till adulthood. It will be important to work closely with our psychology team to ensure that the children have a healthy body image and self-esteem as they grow up.

Cloacal Exstrophy is obvious at birth, and your baby will need to go to the NICU (neonatal intensive care unit) or SCBU (special care baby unit) so they can receive the right medical and nursing care. Where possible, delivery is planned for St Mary's Hospital in Manchester, as this is attached to Royal Manchester Children's Hospital, and allows us to review the baby soon after birth, assess them and plan timing of the surgery. Sometimes, babies are born at different hospitals around the country and, if this is the case, we will arrange for your baby to be transferred to Manchester.

Your baby will be assessed by a neonatal or paediatric doctor when they are born, to check for other birth defects and to ensure baby is otherwise healthy. The urology team will then come to assess your baby and we will usually check for any other abnormalities (involving the kidneys for example) by doing

some simple tests including an ultrasound scan and blood test. We may also want to carry out a special scan of your baby's spine, called an MRI, as often babies born with Cloacal Exstrophy have some degree of Spina Bifida. Our expert neurosurgeon will review the MRI scan and keep a careful eye on your child's development in terms of walking and the spine as they grow up.

We will ask your permission to take a medical photograph (medical illustration) of your baby's tummy, so we can document how it looked when they were born, and document how it changes as we perform the various operations and baby grows and develops. Medical illustrations are kept confidentially, but we sometimes use them to teach other centres about the condition, keeping patient details anonymous. We will ask you to sign a consent form for this at the time and will explain how we store and use photographs.

## What happens for the first operation, to make the stoma?

Once your baby is well enough to have the operation, your baby will be put to sleep (often using a special anaesthetic gas that they breathe in) whilst we make the stoma. This is a major operation and baby will be in theatre most of the day.

When your baby returns from theatre, they will have a stoma on their tummy through which they are able to poo, and we will attach a small bag over the stoma to catch their poo as they pass it. You will be taught how to care for the stoma and empty and change the bag by our stoma nurse specialist.

The bladder will look like a pink dome on the tummy (Bladder Exstrophy), and their genitalia will look as it did at birth. A special dressing is placed over the bladder to keep it protected from the nappy until the operation to close it is done at a later date. You will be taught how to change the dressing and perform nappy cares by the Urology Nurse Specialist or ward nursing staff.

As babies born with Cloacal Exstrophy often have a shorter gut than other babies, their body has less time to absorb all the water and nutrients before it is passed out through the stoma. This means the faeces can look watery and your baby is more likely to lose fluid, salts and electrolytes through their faeces. In the first few weeks after the operation, we will closely monitor your baby's weight, feeding, poo and urine, so we can ensure that we give extra water and salt if they need it. At first, we may choose to give your baby a special type of intravenous (via the vein) food called TPN (Total Parental Nutrition) and ask that they don't feed via their mouth straight away. This is because TPN is absorbed in a different way to milk that you drink, and the TPN can help your baby gain weight whilst their body adapts to their new stoma. Sometimes we need to give some medicines to help the poo become thicker and replace salts that your baby can lose through their poo.

Your baby will likely be on the Intensive Care Unit or High Dependency Unit for some time after the operation. Once they are well enough, we will transfer them to the Children's ward, where they will stay until they are well enough to go home.

Your baby will then be closely monitored in the community, and the health visitor or community nurse will visit, initially every week, to check your baby is gaining weight and not losing too much salt (this is done by checking the salt level in the urine). Often, you will also have the support of the local stoma nursing team, and our urology nurse will be contactable via phone. You will be sent an out-patient clinic appointment to see us within a few months.

## What happens in the next operation, to close the bladder?

Once the Doctors feel your baby is ready for their next operation and a date has been scheduled, you will receive a letter by post asking for you to bring your baby to the hospital for a pre-operative appointment, this is usually the week before the operation.

During this admission the doctors will make sure that all the blood tests have been completed, scans have been done and you will be given the opportunity to meet the anaesthetist who will look after your baby throughout the operation.

In order for surgery to be performed your baby will receive a general anaesthetic. General anaesthesia is frequently used in new-borns and children. The anaesthetist will monitor your baby continuously throughout the procedure.

The anaesthetist will talk to you about the general health of your baby and any relevant medical history, particularly in relation to heart and lung disease. The anaesthetist will also need to know any significant family history related to anaesthetics. If there is any family history of heart or muscle disease, these may need further investigation.

If your child has an allergy to any medication, you must bring this to the attention of the anaesthetist. There is evidence that sensitivities to Latex may develop in Exstrophy patients as a result of the early, intense and constant exposure to rubber products through surgery, diagnostic tests, examinations and bladder and bowel interventions. It is therefore common practice to avoid exposure to Latex in these children and any procedure will be performed following 'Latex precautions'.

The poor development of the bony pelvis is an important aspect of Exstrophy and there will be a wide separation of the pubic bones (bones at the front of the pelvis) called "diastasis". The Consultant Orthopaedic Surgeon will explain how he will repair your baby's pelvis & will talk to you about the metal frame that he will attach to your baby's pelvis following surgery.

## Anaesthetic

General anaesthesia is inducted using either anaesthetic gases through a face mask or by using anaesthetic medicines which are given through a small flexible tube called a cannula. The cannula is placed in a vein in your baby's arm. Once your baby is asleep, anaesthesia will be maintained using the anaesthetic gases.

In addition, to help monitor your child's blood pressure continuously, it may be necessary to place a cannula (small tube) into an artery, as well as a cannula in your baby's neck which helps us measure how well your baby is hydrated after surgery, and allows us to take blood samples without needing to use a needle. These procedures independently have risk of bleeding, infection and injury to other organs but are important to monitor your baby. These procedures will be undertaken with due care and under ultrasound guidance under general anaesthesia.

During the operation your child will be given fluids through the cannula and a blood transfusion is often required.

## Pain Relief

Once your child is asleep the Anaesthetist will ensure that your child is pain free. Pain relief during the procedure will be provided using medicines given either through the cannula (intravenously), a regional

anaesthetic technique (described below) or a combination of the two. These can then be continued in the post-operative period as an infusion.

For the first few days after the operation, your baby will be kept asleep with a special medicine. This ensures that your baby does not experience any pain whilst they recover from the surgery. Whilst they are asleep, they will be attached to a machine called a ventilator and will have a small tube in his/her throat, which will help them breathe whilst they are sedated.

After the first few days, the team can begin to think about when the right time is to wake your baby up and take them off the ventilator. It is important this is timed right, as we want to keep your baby as comfortable as possible after the operation.



## Bony pelvis repair – pelvis osteotomies and external fixator

The pelvis osteotomies (re-adjusting the position of the pelvic bones) are performed to help repair the tummy and reduce the pubic diastasis and will heal usually within 4-6 weeks. External fixation using pins and rods maintain the alignment of the bones during the critical initial weeks of healing. During this period, your child will have to lie flat on his/her back and handling is limited; you will not be able to pick up and hold your baby until after the pins are removed. After approximately 4 weeks, an x-ray is performed and the pins are removed when the bones have healed with new bone formation. Removal of the pins can be done on the ward or in the out-patient department. This does not require a general anaesthetic and pain relief will be given by the ward nursing team before the procedure.



## Care of the pin sites and external fixator

The pin sites require regular cleaning and are usually cleaned on the ward at least once a day. You may wish to be involved in the care of the wound and if so, you will be shown how to do it. All pin sites will ooze a little to start with and this is perfectly normal. As the pin sites settle, the leakage from them will become less and eventually stop.

Your baby will also have their legs bandaged together from the ankle to just above the knee, these are also known as mermaid bandages. This reduces movement of the pelvis whilst the bones are healing, and the bandages remain in place for a further 2 weeks after the pins are removed. The bandages are changed at least once a day, and you will be shown by the ward nursing staff how to do this. You will

be given a supply of bandages to take home with you when you get discharged so you can continue to apply them until they have been on for the 2 weeks after the pin removal.

## Further care of the bladder repair

During the operation, the bladder will be repaired with stents placed in the ureters (the tubes that connect the kidneys to the bladder). The stents are designed to ensure all the urine is able to drain down from the kidneys without any obstruction whilst everything is healing. These stents will be removed after approximately 4 weeks. A kidney, ureter and bladder (KUB) ultrasound scan will then be performed after 24 hours. If the scan is satisfactory, then we will clamp the suprapubic catheter (a tube placed in the tummy to drain the newly repaired bladder) and repeat the scan 24 hours later. If this scan shows no problems the catheter will be removed.

Sometimes, before you are discharged, your baby will have another short anaesthetic to check inside the bladder with a special camera and make sure the urethra (the tube and opening where urine drains out of the bladder) is working well. Occasionally, if the urethra is a little bit narrow, we will leave a small catheter (tube) in the urethra for a further 2 weeks and arrange for you to come back for it to be removed. This helps the urethra stay open whilst it continues to heal and gives your baby a better chance of emptying their bladder more effectively.

Before you go home (or when you come back in 2 weeks if we've left a catheter in), we will teach you the technique for intermittent urethral catheterisation, which is to be performed twice a day (morning and night). This is a very important aspect of caring for the bladder and kidneys after the surgery, as it ensures that the tube the urine drains through (urethra) remains patent, and checks that no urine is being left behind throughout the course of the day, which can cause infections and make your baby feel poorly.

## Other Teams who might help look after your baby

Cloacal Exstrophy is a rare and complex medical condition, and there are often other associated abnormalities, a common one being Spina Bifida. We understand that many families need to travel from across the U.K to see the team and attend appointments, so your baby will be seen in a special Cloacal Exstrophy Clinic, to allow you to see different teams all at the same time. The urology team will be in the clinic appointment, as well as a member of the psychology team. There will also be a neurosurgical consultant; it is their job to look after your baby's spine and monitor their development.

Despite the pelvic reconstruction that is performed when we close your baby's bladder, most patients do not need long term follow up from the orthopaedic team. However, sometimes children born with Cloacal Exstrophy can have other associated abnormalities such as Talipes, in which case you may be sent separate appointments for the orthopaedic Doctors.

## Psychological care at Royal Manchester Children's Hospital

At Royal Manchester Children's Hospital there is an experienced team of doctors, nurses, play specialists and clinical psychologists committed to ensuring your child gets the best possible care and treatment both physically and emotionally.

A clinical psychologist is someone who has studied behaviour and feelings. They use this knowledge to try to understand the problems that children and their families may be having and suggest different ways of trying to help.

## Psychological aspects of living with Cloacal Exstrophy

Living with any kind of physical health problem can present challenges, even if only from time to time. Cloacal Exstrophy are conditions that tend to present different challenges for children and their families at different ages and developmental stages. These hurdles can be overcome, and with support and encouragement, children can grow up to feel good about themselves and in charge of their lives.

### When might I see the clinical psychologist?

A clinical psychologist will try to meet you on the ward when your child is an in-patient as part of our routine package of support. You might also see the clinical psychologist in clinic when your child comes to an out-patient appointment to see the doctor. Your child's doctor or nurse may suggest you see the psychologist to help with a particular problem, or you can contact the psychologist directly if you would like some support. The clinical psychologist is a regular part of the hospital team and is there to help you and your child manage and cope with his/her condition.

### How can the clinical psychologist help?

Children who have physical health problems, and the families who care for them, often face problems from time to time. These can be distressing, and sometimes families need help with these problems. Clinical psychologists can help with difficulties children face about their health or treatment. They can also help with other emotional, behavioural and relationship problems. Clinical psychologists aim to help parents and children understand their problems and find ways of solving them.


Research is clear – with good support from family, teachers and friends, children with Exstrophy and related conditions do extremely well. But the process of learning to cope can be hastened by timely intervention by someone like a psychologist.

All of us need help from time to time and a psychologist can provide:

- A listening ear
- Understanding to help piece things together
- Support to achieve things that are proving difficult
- Ideas of how to face/handle problems

### Contact

If you would like to discuss any of the information in this leaflet further, please contact a member of the team.

 In case of problems or advice please call:

Debra Collins (secretary)

(0161) 701 1636 (appointment queries)

Jenny Powell (Clinical Nurse Specialist)

(0161) 701 7707 (for clinical queries)

Ruth Hurrell (Clinical Psychologist)

(0161) 701 4514 (for psychology queries)

James Devine (Theatre Scheduler)

(0161) 701 0779 (for surgery date queries)

Ward 77 (urology ward)

(0161) 701 7700 (for urgent advice)