Manchester University
NHS Foundation Trust

Royal Manchester Children's Hospital

Information for Patients

Bladder Exstrophy & Epispadias

This leaflet is for parents or carers whose baby has recently been diagnosed with Bladder Exstrophy & Epispadias. 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 Bladder Exstrophy & Epispadias?

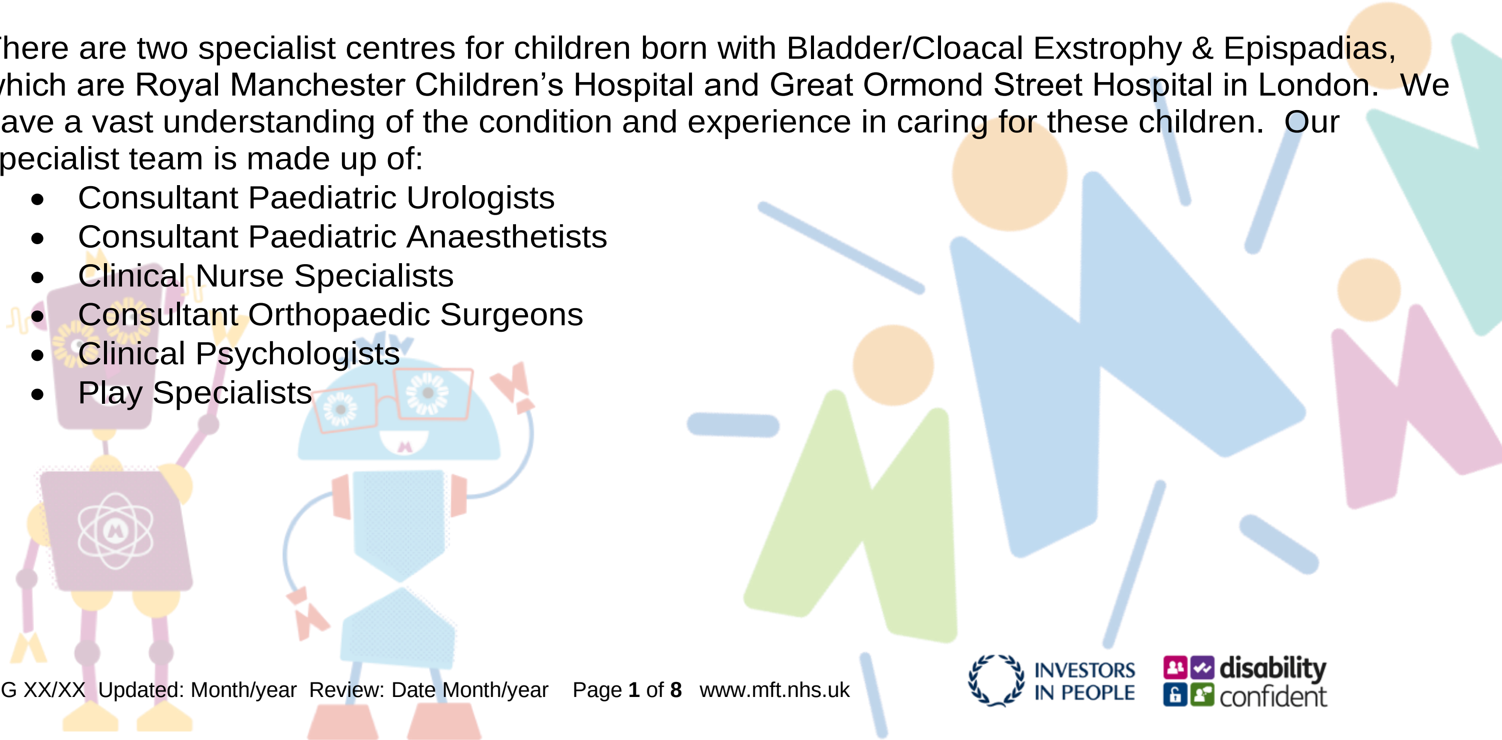
The Bladder Exstrophy & Epispadias complex is a birth defect that affects the bladder, the urethra, the genitals and the pelvic bone. It affects approximately 1 in 40,000 live births. Epispadias is at the minor end of the spectrum with cloacal exstrophy being the most severe form.

Bladder Exstrophy & Epispadias is a surgically correctable birth defect, but each child is unique and therefore their journey with Bladder Exstrophy & Epispadias and the care they receive is specific to individual needs.

Why do I have to come to Manchester or London?

There are two specialist centres for children born with Bladder/Cloacal Exstrophy & Epispadias, 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?

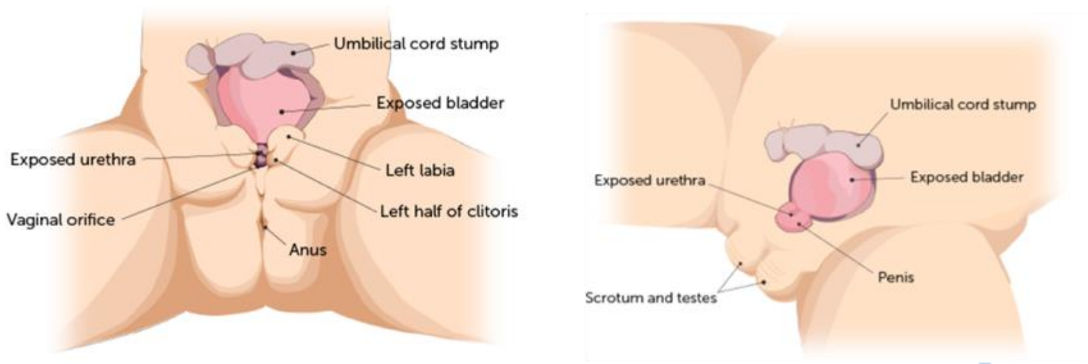
These conditions are part of a spectrum of birth defects, with Epispadias being at the minor end of the spectrum, and Cloacal Exstrophy being the most severe form. Approximately 40% of Bladder and Cloacal Exstrophy defects are detected on ante-natal scan, but the condition can often be missed due to timings of scans and positioning of the foetus, making it harder to identify on a routine ante-natal scan.

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 boys, the penis may be shorter and wider than normal. In girls, the clitoris and labia are split. Epispadias is always present in both Bladder Exstrophy and Cloacal Exstrophy, however it is possible to see Epispadias on its own (without Bladder Exstrophy or Cloacal Exstrophy).

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.

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



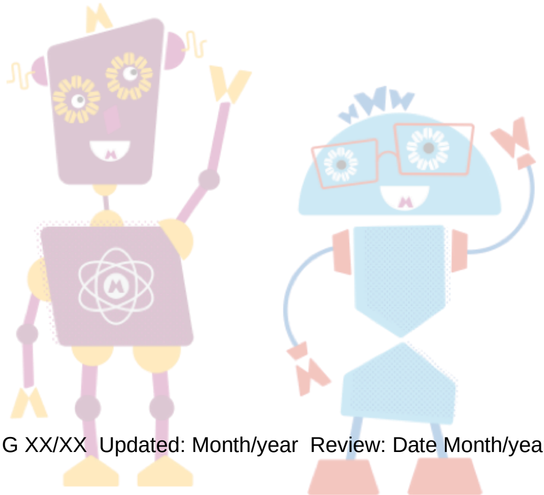
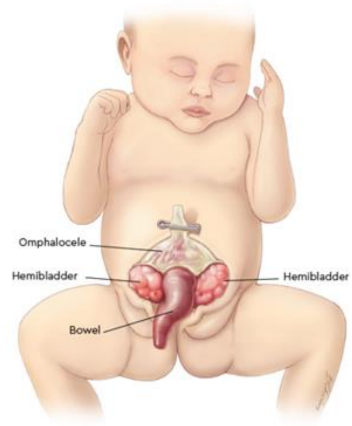


Diagram showing Cloacal Exstrophy



Why has this happened?

It is not known what causes Bladder Exstrophy & Epispadias defects. The problem occurs between the 4th-10th week of pregnancy when various organs, tissues and muscles begin to form. Bladder Exstrophy & Epispadias **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 Bladder Exstrophy & Epispadias, 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?

Bladder Exstrophy & Epispadias are corrected in a series of operations over the first years of life. The overall aim of treatment is to protect the kidneys and correct the abnormalities, so that your child's urinary system and genitals work properly and look as normal as possible.

If your baby has been born with **Cloacal Exstrophy & Epispadias**, an operation to ensure that your baby can pass faeces will usually be performed within the first week of life. You will meet with the Consultant Paediatric Urologist and our specialist team who will explain the operation. The anatomical abnormalities to the urethra, genitals and bladder will not be corrected within the same operation but will thereafter follow a similar structure and timing of surgeries as those born with Bladder Exstrophy & Epispadias.

If your baby has been born with **Bladder Exstrophy & Epispadias**, the operation to close the tummy is often done around 4-6 months after birth. This gives your baby the time to grow and mature, making surgery much safer. You will be shown how to take care of your baby's tummy after birth, and can usually be discharged home after a few days, providing baby is otherwise fit and well. We believe that having the opportunity to get to know your new baby and spend time together in your own environment is of benefit for the whole family. Our clinical evidence shows that closing the tummy when baby is a little bit older has no impact on their overall outcome and continence.

The **Epispadias**, which affects the urethra and genitals, is repaired in girls during the operation to close the bladder. In boys, the surgery is delayed until they are approximately 1-2 years old as the surgery is complex so cannot be done in one operation.

Your baby will need to have some blood tests, a medical photograph and an ultrasound scan of the kidneys before they are discharged home. This checks that your baby's kidneys are working well, and documents how their tummy looked when they were born. Your baby will also need to have some special scans of the pelvis (CT/MR) but this is often done once a date for closing the tummy has been scheduled. Your baby may need to be given medicines to keep them still whilst the scans are completed.

Once our team at Royal Manchester Children's Hospital have been informed of the birth of your baby and their diagnosis, we will arrange an out-patient appointment for you to attend our clinic to meet the team and will be provided telephone support by our clinical nurse specialist. If your baby was diagnosed ante-natally (on a scan), then you may be offered to meet us in clinic before baby is born. When we all agree baby is ready for the surgery, we will list them and a date for them to come in will be allocated by the theatre scheduler.

What happens when an operation date is available?

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 you 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 and he 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 trough 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.

Regional anaesthesia involves placing local anaesthetics so that they block the painful nerve impulses and provide pain relief. An epidural or caudal are examples of such a technique. The Anaesthetist will site a fine plastic tube (catheter) into your child's back. Local anaesthetics can then be injected through this catheter. The local anaesthetic will block the nerve messages and cause numbness at the surgical site.

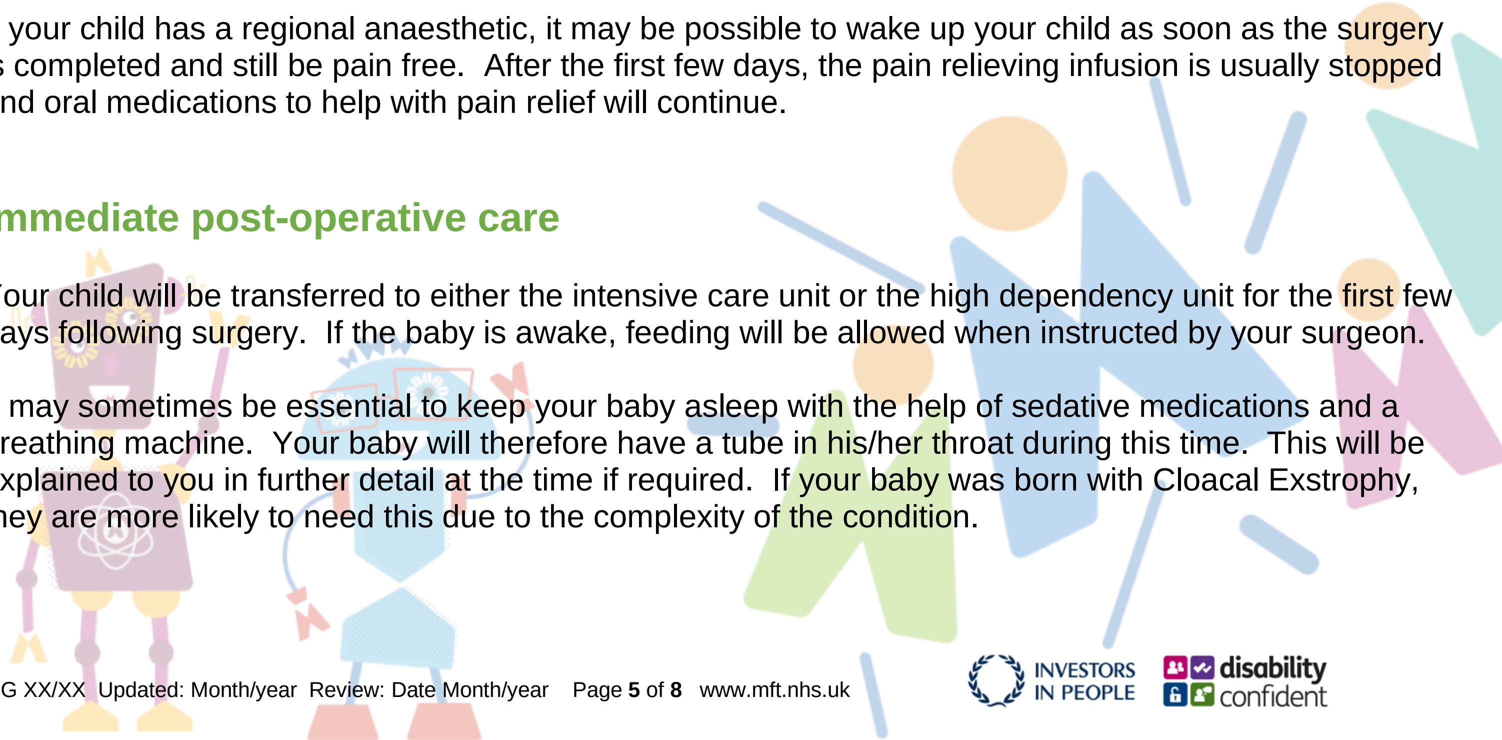
An infusion pump allows local anaesthetic to be given continuously in prescribed amounts through the catheter following the surgery. The nursing team will monitor your baby at regular intervals. Specialist nurses from the Pain Team will also visit and check that the caudal-epidural is working properly. If this is considered inadequate, intravenous pain relief can be added to ensure your child is pain free.

If your child has a regional anaesthetic, it may be possible to wake up your child as soon as the surgery is completed and still be pain free. After the first few days, the pain relieving infusion is usually stopped and oral medications to help with pain relief will continue.

Immediate post-operative care

Your child will be transferred to either the intensive care unit or the high dependency unit for the first few days following surgery. If the baby is awake, feeding will be allowed when instructed by your surgeon.

It may sometimes be essential to keep your baby asleep with the help of sedative medications and a breathing machine. Your baby will therefore have a tube in his/her throat during this time. This will be explained to you in further detail at the time if required. If your baby was born with Cloacal Exstrophy, they are more likely to need this due to the complexity of the condition.



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.

You 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.

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 Bladder Exstrophy & Epispadias

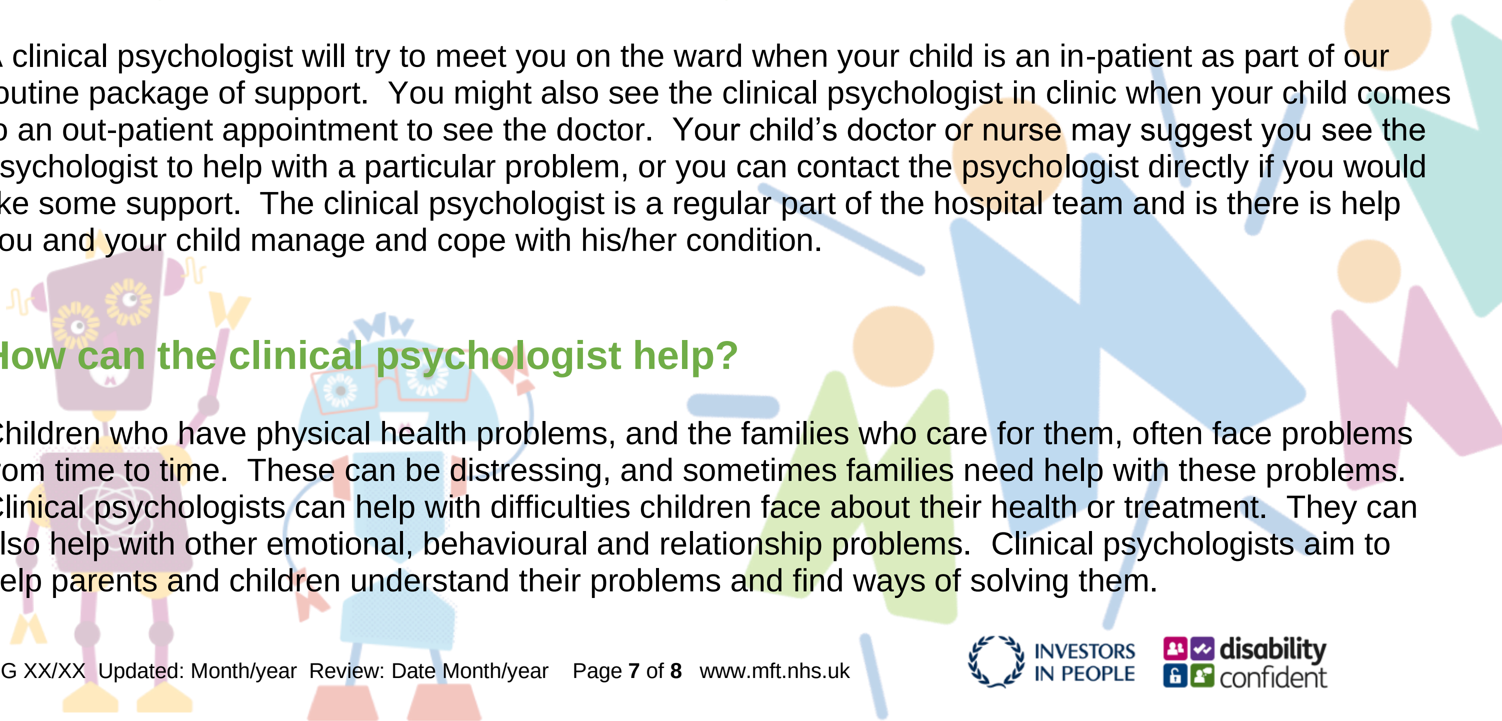
Living with any kind of physical health problem can present challenges, even if only from time to time. Bladder Exstrophy & Epispadias are conditions that tend to present different challenges for children and their families at different ages and developmental stages. These hurdles can be overcome, and 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 is 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)
Ward 76 (day case)	(0161) 701 7600 (for urgent advice)