ConvaTec

Cloacal Exstrophy

What is it?

This is a rare and complicated birth defect that affects the midline of the body.

- Diagnosis is usually made at birth can sometimes be diagnosed prenatally.
- What causes Cloacal Exstrophy is unknown.

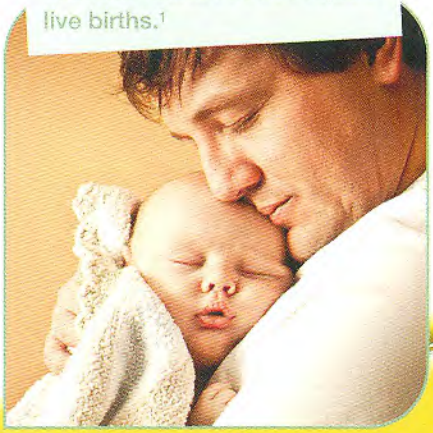
Signs and symptoms

Your baby will have some of the following:

- the bowel is exposed outside of the body
- the bladder is exposed outside of the body and may be in two halves
- imperforate anus – the anus (the outlet of the back passage) has not been formed and is not open in the normal place but may communicate with the bladder
- spinal defects – these may be major or minor. Many children born with Cloacal Exstrophy also have varying degrees of spina bifida
- the pelvic bones will be widely separated at the front (pubis)
- genital abnormalities - penis absent or divided in two/ vagina may be absent
- renal abnormalities

The severity of each of these anomalies will determine the plan of action for the management of your child's condition. Each child's situation will be different because of the multiple defects that have occurred together or separately. Because of these defects it may make it difficult to determine which sex your child is. Your child will be assessed as an individual and the appropriate counselling/support will be given.

Cloacal Exstrophy is a rare disorder found in 1 in 145,000 live births.[1]



Reference:
1. EMSYCAR (The East Midlands and South Yorkshire Congenital Anomalies Register) http://www2.le.ac.uk/departments/health-sciences/research/ships/timms/projects/car. Period covered 2000 - 2008 inclusive.

Investigations may include:

- ultrasound of kidneys at regular intervals
- I.V.P.
- DMSA Scan
- urodynamics
- CT Scans of the pelvis
- genetic tests

Frequent hospital visits will be necessary for your child to be assessed.

Treatment / surgery

Your child will have a series of operations throughout life. Some children may have more operations than others depending on the health of the child and the preferred management by individual consultants.

Sometimes more than one thing can be accomplished during one operation.

- Soon after your child is born an operation on the bowel will be necessary to get it back inside the tummy and to make an opening (stoma) for your child to be able to get rid of waste into a bag. This may be a permanent stoma if your child does not have enough colon (large bowel).
- The bladder will also be repaired at this time and your baby may require a bag to collect the urine (wee) but this is not likely to be permanent.
- Any spinal defects are likely to be repaired at the same time.
- If there is an adequate amount of large bowel, the stoma may be closed at a later date and the bowel brought down to the bottom and a new opening made (a pull-through operation).

To try and achieve continence, major urinary surgery will be performed at a later date. The various options will be discussed with you, but some of the following may be necessary:

1. The bladder may be very small, therefore an operation may be needed to make this bigger (augmentation).
2. Most children will need to use a small tube (catheter) to empty the bladder, several times per day. This is called intermittent catheterisation and will be taught by a specialist nurse.
3. To be able to pass the catheter to perform intermittent catheterisation it may be necessary to make an opening on the tummy from the bladder (Mitrofanoff).
4. Some children may not wish to use a catheter to empty the bladder and they will be able to have an opening (stoma) made on the tummy to be able to collect the wee in a special bag.
5. A vagina may also need to be created.
6. Some children may have problems with urine flowing back up the ureters to the kidneys (reflux). This can cause serious damage to the kidneys and may have to be treated with an operation called 're-implantation of ureters'.

Possible problems

- Urinary tract infections.
- Latex allergy – occasionally some children may have an allergic reaction to latex that is contained in surgical gloves. As with any allergy this may be a minor reaction or it may be more serious. If you have any worries contact your doctor.
- Urinary incontinence requiring further surgery to gain continence.

Child's health and development

- The birth defect that your child has is not obvious to the public. Most people will not know unless you tell them.
- Your child will require frequent visits to hospital and may still be incontinent when starting school. This can affect education, however, there is no evidence to suggest that children with Cloacal Exstrophy have a lower IQ than the rest of the population.
- Most girls will be able to have sexual intercourse using their own natural vagina. However, some may require a surgically-made vagina.
- If your child has ovaries and a uterus, it is very likely that they will be able to have children.

What will happen to us emotionally?

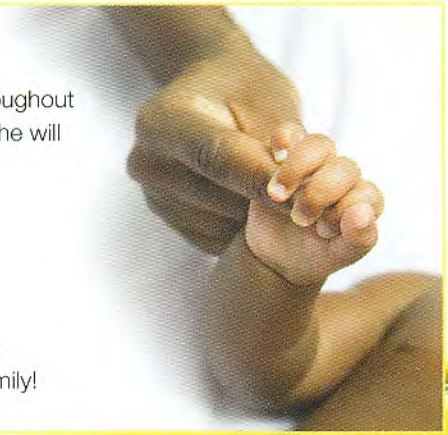
- You will have to deal with questions from family and friends when your child is born.
- When your child reaches school age you will have to deal with questions from other people.
- Social issues with your child, other children, and adults may be more difficult.
- Your life will be different and probably more difficult than those around you.
- Support will be available to help you deal with these situations.

Support

Support and counselling will be given throughout by your Clinical Nurse Specialist and he/she will give you relevant information for:

- support groups – parent led groups
- financial support
- educational issues

Please don't hesitate to ask for advice on anything which is worrying you or your family!



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