Cloacal Exstrophy

BIRTH DEFECT RESEARCH FOR CHILDREN

What is Cloacal Exstrophy?

Cloacal Exstrophy is a serious congenital (present at birth) abnormality. The bladder is divided into two halves, part of the bowel is exstrophied (turned inside out) and located between the bladder halves, and the bones of the pelvis are separated in the front. In males, the penis is split into two halves. In females, the clitoris is split into two halves. Associated conditions often include abnormalities of the kidneys, cardiovascular system, and spine (Spina Bifida). Often there is an omphalocele which is a sac containing abdominal organs protruding through a defect to the outside of the body. Females may also have two vaginal openings or the vagina may be absent.

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How many children have Cloacal Exstrophy?

Cloacal Exstrophy is very rare occurring in one out of 200,000-400,000 live births.

How do you know if your child has Cloacal Exstrophy?

Cloacal Exstrophy can be detected prenatally by ultrasound; however, it is usually diagnosed at birth.

What causes Cloacal Exstrophy?

The cause of Cloacal Exstrophy is not known.

How can you help a child with Cloacal Exstrophy?

The treatment and timing are individualized depending on the health of the child and the extent of the defects. It is recommended that a multidisciplinary medical team be utilized at a large facility capable of handling complex birth defects. The usual treatment is staged reconstructive surgery. The basic surgeries are usually performed at three separate times. The first stage of surgery is usually performed soon after birth. This surgery focuses on the bladder, bowel, and pubic bones. The pubic bones are brought together, the bladder is closed, and a way is created for the infant to eliminate waste. Penis malformations are usually too extensive to correct, so most males undergo gender reassignment surgery. The second stage of surgery is usually performed around age 3-5 and involves additional reconstruction of the bladder. The third stage of surgery is performed at the time of puberty and involves reconstruction of the vagina.

What's in the future for a child with Cloacal Exstrophy?

With aggressive reconstructive surgery, the chances for survival are around 80%. Children are usually not affected intellectually; however, their quality of life will depend on the severity of their abnormalities and whether they can be treated.

Fact Sheet by:

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