

Neuronal intestinal dysplasia

The large intestine massages waste along its length to the rectum by rhythmic, muscular contractions of its walls (peristalsis). This activity is controlled by nerves of the enteric nervous system (ENS). Neuronal intestinal dysplasia (NID) is characterised by the reduced motility of the large intestine, caused by abnormalities of the enteric nerves. The unusually slow passage of waste through the large intestine leads to chronic problems, such as constipation and uncontrollable soiling. There is no cure. Treatment options include laxatives and surgery.

Symptoms

The most common symptoms of NID include:

- Passing bowel motions infrequently
- Constipation
- Uncontrollable soiling
- Abdominal pains
- Nausea
- Poor appetite.

Less common symptoms include:

- Blood in the stools
- Haemorrhoids
- Diarrhoea.

NID can be diagnosed soon after birth

The first bowel motion after birth isn't true faeces, but a dark, gluey substance known as meconium. Most of this substance comprises the mucus that layered and protected the bowel lining while the baby was in utero. A newborn with NID may not pass their meconium until 24 hours or more after being born. The delayed passage of meconium may lead doctors to investigate further.

NID is a neuromuscular problem

The nerves communicate with muscle fibres by releasing chemical messages called neurotransmitters. These messages are picked up by special receptors in the muscle tissue. If enough receptors are stimulated, the result is muscular contraction. Some studies have suggested that children with NID have abnormal neurotransmitters in the muscular layer of their intestinal walls. These abnormalities include a deficiency of a peptide known as substance P, which is thought to contribute to peristalsis. Research also indicates that the nerve cells of the bowel may be abnormal in number, position or appearance.

Quality of life can be severely affected

Many children with NID have emotional and behavioural problems, including:

- Constant anxiety about losing control of their bowels in public.
- Fear and embarrassment about their condition.
- Withdrawal from social situations.
- Depression and loss of self-esteem, especially if they are teased by other children at school.

NID may mimic or coincide with Hirschsprung's disease

Hirschsprung's disease is characterised by the congenital lack of nerve cells in the rectum, large intestine or both. This means that peristalsis of the rectum or large intestine is absent, so that waste simply stalls and can't be expelled via the anus. The symptoms include severe constipation and obstruction. This disease is similar in many ways to NID, which can make diagnosis difficult. A further challenge to diagnosis is that NID and Hirschsprung's disease can sometimes occur together.

Diagnosis methods

NID is diagnosed using a number of tests, including:

- **Physical examination** - a child with NID may have a distended abdomen and obvious discomfort.
- **X-ray** - to see if the bowel is distended with excess faeces.
- **Barium enema** - a special contrasting liquid is flushed into the bowel via the anus, then x-rays are taken.
- **Transit study** - the child swallows a radioactive dose. Its passage through the bowels is then tracked.
- **Biopsy** - a small tag of muscle tissue from the large intestine is removed and examined in a laboratory.

Treatment options

Treatment options may include:

- Drugs to improve bowel motility.
- Regular enemas to flush the rectum of faeces.
- Surgery to remove affected portions of bowel.
- Sometimes, the appendix is used to create a small opening into the bowel on the outside of the abdomen.

A range of surgery options

The type of surgery chosen depends on the location and extent of affected bowel, and the health and age of the child. Options include:

- The affected parts of the bowel are removed, and the healthy section is attached directly to the rectum ('pull-through' operation).
- A colostomy may be performed, where the bowel is re-routed through an artificial hole in the abdominal wall, and a colostomy bag fitted. Sometimes, a temporary colostomy is performed; once the child has regained strength, they undergo the pull-through operation.
- The appendix may be brought to the surface to create a tiny stoma (opening). This can be done using a laparoscope (telescopic surgery).
- Enemas can be given regularly directly into the stoma or appendix.
- It is not always possible to treat NID with surgery, as too much of the bowel may be affected.

Coping strategies

Strategies that may help your child to cope with NID include:

- Don't ever chastise your child for soiling. Remember that their bowels are difficult to control.
- Offer pull-up disposable nappies or other continence aids appropriate to your child's age.
- Allow your child to talk about their feelings.
- Make sure you educate your child about NID, so they realise their bowel control problems are not their fault.
- Devise an 'action plan' in consultation with your child's school, that includes a trusted teacher's cooperation at toilet time, and easy access to toilets and a shower.
- Join a support group such as NidKids.
- Professional counselling for the child and family members may be helpful.

Where to get help

- Your doctor
- Gastroenterologist
- NID Clinic at Royal Children's Hospital, Melbourne VIC

- Paediatric Continence Association of Australia Tel: 1 300 885 209

Things to remember

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- The unusually slow passage of waste through the large intestine leads to chronic problems, such as constipation and uncontrollable soiling.
- Treatment options include laxatives and surgery.

This page has been produced in consultation with, and approved by:

Paediatric Continence Association

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