

Total Colonic Aganglionosis

Total colonic aganglionosis is a birth defect in which nerve cells known as ganglion cells are lacking in the large intestine. As a result, the large intestine cannot move stool through and becomes blocked, causing the abdomen to swell.

Treatment

Treating total colonic aganglionosis by surgically removing the colon and reconnecting the lower end of the small intestine to the beginning of the anal canal produces many bowel movements and provokes a terrible diaper rash. Because colon tissue without ganglion cells (aganglionic tissue) absorbs water, some surgeons theorized that using a patch of aganglionic cells in the reconnection (anastomosis) would take advantage of the water absorption capacity of aganglionic tissue and reduce the number of bowel movements following surgery.

While using the patch of aganglionic cells sometimes produces the intended results, many other times the patch causes a significant number of problems. These include irritation and inflammation of the intestines, retention of secretions and stool in the patch, and diarrhea from the secretions in the patch itself and in the rest of the intestines. In some cases, the diarrhea is so severe that the patch has to be removed. In our practices, we have had to do this for several patients who had received the patches in operations performed by other surgeons. This included a patient that lost a large volume of fluid, so large that it required eight liters of electrolyte solution intravenously everyday to compensate for the losses. The moment we resected the patch, the patient improved dramatically.

Because of these complications, we currently use a straightforward anastomosis between the end of the small intestine and beginning of the anal canal to treat patients with total colonic aganglionosis.

Timing of Surgery

The optimal timing for the surgery is controversial. While surgeons have demonstrated that the operation can be performed very early in life, this may not be best for the patient in the long run. In our opinion, the surgery should be performed when the patient is totally continent for urine (able to exercise total control over urination). Doing the surgery when the patient is continent will allow the



patient to become toilet trained for feces much faster and means the patient will suffer much less diaper rash.

Contact the Colorectal Center at Cincinnati Children's

For more information or to request an appointment for the Colorectal Center at Cincinnati Children's Hospital Medical Center, please [contact us](#).