

Patient Care > Imperforate Anus

Introduction

Congenital malformation of the anus occurs approximately once in every 5000 live births. There are various forms of "imperforate" anus including anal stenosis (tight opening), anal membrane, low agenesis (absence), high agenesis, and colonic atresia (absence of continuity). The most common forms are the low and high agenesis where there are no skin openings where the anus should be.

Embryology/Pathology

In the normal individual, the anorectal (lowest portion) segment of the intestine passes through a sling of muscle in the pelvis which provides involuntary (not in your control) continence. When stool distends the bowel this muscle automatically pulls up and prevents stool from progressing further. The distended bowel sends a signal to your brain telling you that you need to use the toilet. The bowel at the anus passes through two other muscles which we can control. By contracting these muscles we can further delay a bowel movement.

In low agenesis, the bowel does not connect with the skin but it passes through the sling of muscle providing involuntary control. In high agenesis the bowel terminates above this sling of muscle.

Low agenesis occurs predominantly in girls and there may be a fistula (tract) with an opening at the lower part of the vaginal opening. Boys, predominantly, have high agenesis. They too may have a fistula which cannot be seen and connects the end of the bowel to the urethra (tube from the bladder exiting at the penis).

Anal malformations may be associated with other anomalies, including the esophagus (swallowing tube), heart, and kidneys.

Sign/Symptoms

In anal stenosis and colonic atresia an anal opening is present but there is constipation and no bowel movement, respectively. Abdominal distension may occur and may progress to vomiting.

Anal membrane as the name implies presents with a skin covering which prevent defecation. Abdominal distension and vomiting may occur.

Both low and high agenesis will have an absent anal opening. Low agenesis may have a fistula (more in girls than boys).

Diagnosis

Anal stenosis usual can be determined by the calibration of the anal opening by dilators. A barium contrast enema is usually performed and a rectal suction biopsy may be obtained to rule out the absence of nerve cells (see [Hirschprung's Disease](#)).

Colonic atresia is diagnosed by a barium contrast enema.

The diagnosis of anal membrane, low or high agenesis is made by visual inspection. Low and high agenesis may require further investigations in an attempt to determine whether the bowel ends below or above the muscle sling referred to earlier. This may involve ultrasound, contrast, computerized tomography (CAT scan) studies. The location of the end of the bowel often dictates the surgical approach as well as the long term prognosis.

Treatment

Anal stenosis usually only requires anal dilation for a few months. Occasionally surgery is necessary if dilations are not successful to prevent constipation.

Anal membrane requires excision of the membrane and suture of the anal lining to the skin. The anal suture line will have scarring and contraction with healing. Therefore, after healing, a period of anal dilation will be necessary.

Colonic atresia will require abdominal surgery to remove the obstructed segment and to reconnect the colon. The repair may be performed by laparoscopy or by the "open" method, a choice dictated by the anatomy and your surgeon.

Low agenesis can usually be surgically approached through the fistula tract or the perineum. In girls where there is a fistulous opening near the vaginal opening; often only a "cut-back" procedure is needed. This provides for an adequate opening to prevent constipation. Post-operative dilations will be necessary. If there is no fistulous tract, exploration of the perineum to find the end of the bowel is performed. The bowel may be sewn to the skin followed by healing and dilations.

High agenesis may require an abdominal, sacral, or perineal approach. The route of repair is dictated by the anatomy and the presence of a fistulous tract. The abdominal approach may be performed laparoscopically or "open" (although laparoscopy is typically preferred). The sacral approach is through an incision from the coccyx (tail bone) down to where the anal opening should be. In either case, the bowel is pulled through the muscle sling and the external muscles and sutured to the skin. This is a complex procedure and your surgeon will describe in detail the anatomy and why he or she has chosen this particular approach.

Occasionally, a newborn has other life-threatening problems which take precedence over the anal malformation. To temporize, a colostomy may be performed. The definitive surgery may then be done later.

Post-operative and Long-term Complications

If an operation is performed under general anesthesia, there are risks of anesthesia, bleeding, infection, and post-operative intestinal obstruction. In any pull through procedure there is always the risk of vascular (blood supply) compromise to the intestine requiring reoperation.

In anal stenosis and anal membrane, the usual problem is that of constipation. The dilations which may continue for three to six months must be performed regularly. There is nearly uniform success with normal bowel movement and control.

In colonic atresia, problems usually stem from associated anomalies rather than the operation or the atresia. Since the defect does not involve the anus, bowel function is rarely a problem.

Patients with low agenesis have greater than 90% success without constipation and with good bowel control. Unfortunately, some do have difficulties with both constipation and control. Diet, laxatives, and enemas alleviate most of these problems.

Of patients with high agenesis, unfortunately, only approximately 75-80% have no constipation and have good bowel control. Another 10-15% with bowel control and constipation problems may be managed medically with diet, laxatives, and enemas. But about 5% will have persistent difficulties with constipation or lack of bowel control to the point that it adversely affects their life style and social development. A few may require a permanent colostomy diverting the stool to the skin and a collection device.

Any newborn with neurologic impairment or Down's syndrome with low or high agenesis will have a much more difficult time with constipation and bowel control. These can usually be treated with medical means.

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