Hirschsprung's disease (HD) is characterized by a lack of nerve structures in the intestine. In about 75% of all cases affected by this abnormality, the aganglionosis is confined to the rectum and sigma. About 17% of all patients show an extended disease and in about 5-8% the absence of ganglia is present in the total large bowel and terminal ileum. Only a small number of patients exist with extended aganglionosis including a large portion of the small bowel and upper GI tract. Besides the classical HD patient, syndromic Hirschsprung disease and rare neurocristopathy exist. Ondine syndrome or Waardenberg syndrome are part of this entity. Finally there is a clear increased incidence of extended Hirschsprung's disease in patients affected by Down's syndrome. Aganglionosis is more frequent in males than females (4:1) in the "classic HD group". In syndromic and long-segment disease the ratio between male and female is 1.5-2:1 (1).

The typical clinical signs of HD are delayed meconium passage, abdominal distension, vomiting and enterocolitis. More than 80% of all Hirschsprung cases present symptoms in the neonatal period. Only a few of these are having a prenatal diagnosis (mostly performed by intrauterine MRI and ultrasound). Entero-colitis is present in one third of babies and toddlers with HD and associated with diarrhea. Enterocolitis is still the most common cause of death in HD and especially in Down's syndrome a severe hazard for the patient's life (2).

Historically the contrast study was one of the most important exams. The typical proximal dilatation above the distal narrow segment is usually always present in elder children. In the newborn the dilatation can be absent, or already not present. The histological exam to confirm HD, is showing the absence of ganglia structures in a rectal biopsy. Nowadays histochemical staining technique for the detection of acetylcholineesterase activity in rectal suction biopsy is a reliable and simple method for diagnosing HD. Nevertheless in some cases this diagnostic technique is not always reliable in the young and immature patient. Many pathologists applied prefer to repeat the exam to wait for a "maturation" of nerve structures avoiding the risk a false positive diagnosis. Manometry can be performed but it is only an adjunctive diagnostic procedure.

The surgical strategy should include the determination and extension of the disease (frozen section), the removing of the affected bowel segment and the preservation of the sphincter complex. Usually a primary anastomosis is performed. In critical ill babies with a septic situation or a long aganglionosis an enterostomy can be a life saving procedure postponing the definitive diagnostic and surgery to a later date (3).
Torre technique. For long and total HD many modifications of the above mentioned techniques were described and applied. All techniques follow the above-mentioned principles to bring gangli- onotic, normal bowel as close as possible to the sphincter complex. The resection at the level of the rectum is in all cases different due to technical details. The Swenson procedure is clearly the most radical approach, doing a total resec- tion right above the sphincter complex with an end-to-end anastomosis. Soave’s techniques end up with a similar ana- stomosis, but leaves the cuff of aganglio- notic muscle inside, which is divided and opened. The Duhamel leaves the rectum with aganglionic muscle in place, doing a lateral posterior anastomosis. Rehbein did in his approach an extremely deep resection of the rectal part, leaving a short distance to the sphincter complex. The modifications of De la Torre and the Georgeson approach repeat a Soave-like technique, but starting the mucosectomy from the anus, preparing the rectum from below. Each technique has its own technical difficulties and risk, as well as surgical complications which might lead to the known consequences and pro- blems of surgery for Hirschsprung’s di- sease. Therefore the aim of any surgery should be a patient who is having regular stool frequency and bowel function, no more enterocolitis and stool retention and faecal continence. Surgery should be performed ideally as early as possible in life, using a minimal-invasive technique for short post-op course and excellent cosmetically results. The risk for surgi- cal early and late surgical complications should be minimized. All this factors fi- nally contribute to a good quality of life. The purpose of this article is to empha- size the accuracy for a correct diagnostic approach before surgery, to discuss early and late complications in HD treatment and to elucidate the treatment options in case of complications (4-6).

Diagnostic Pitfalls

The diagnostic approach for Hirschsprung’s disease is represented by clear and precise algorithms. This can be described as a diagnostic triad for HD including x-ray contrast studies, mano-}

Figure 1

Extramucosal laparoscopic biopsies taken with 3 mm instruments before starting a laparoscopic assisted pull through.

Figure 2

4 months after De la Torre pull through. Still unrecognized total hypoganglionosis on histological exam. Patient was operated twice, ending up with a near total colectomy.

Table 1

Algorithm for diagnostic and therapeutic approach to Hirschsprung’s disease.

X-ray studies should always be per- formed without prior cleaning of the co- lo-rectum and with sufficient contrast media to demonstrate not only the recto- sigmoid region but also the more prox- imal part of the colon. In some instances the real extension of the aganglionic zone is not seen, because of an incom- plete study of the intestine. Especially in long HD the typical signs of a calibre difference and the proximal dilatation might be absence. Finally the exam shou- ld always include a defecation without rectal tube inside the bowel. In about 80% an experienced radiologist is able to make a correct diagnosis. The second exam should be a mano- metry and finally the histochemical and histological work up. X-ray studies should always be per- formed without prior cleaning of the co- lo-rectum and with sufficient contrast media to demonstrate not only the recto- sigmoid region but also the more prox- imal part of the colon. In some instances the real extension of the aganglionic zone is not seen, because of an incom- plete study of the intestine. Especially in long HD the typical signs of a calibre difference and the proximal dilatation might be absence. Finally the exam shou- ld always include a defecation without rectal tube inside the bowel. In about 80% an experienced radiologist is able to make a correct diagnosis. The second exam should be a mano- metry and finally the histochemical and histological work up. X-ray studies should always be per- formed without prior cleaning of the co- lo-rectum and with sufficient contrast media to demonstrate not only the recto- sigmoid region but also the more prox- imal part of the colon. In some instances the real extension of the aganglionic zone is not seen, because of an incom- plete study of the intestine. Especially in long HD the typical signs of a calibre difference and the proximal dilatation might be absence. Finally the exam shou- ld always include a defecation without rectal tube inside the bowel. In about 80% an experienced radiologist is able to make a correct diagnosis. The second exam should be a mano-
helpful to reduce the amount of bile salts. In some instances is might be using barrier creams can resolve these more frequent and also difficult to treat.

colon resection, and especially in total surgery. In long HD after extensive in at least one third of HD patients af -
tal colon and might cause the occlusion. can slip under the mesentery of the dis -
tal colon and might cause the occlusion. can slip under the mesentery of the dis -

In difficult situations a local revision of the stoma might be necessary or the de-
mulated segments. This can be perfor -

The choice of the re-do technique is

Post op complications
In the post op period all general complications after abdominal surgery can occur (haemotoma, infection, bleed -

Post-op bowel occlusion seems to be less frequent after laparoscopic and trans anal pull through surgery. In cases of prior severe enteroctis and previous surgery the incidence of bowel occlusion due to adhesions is described in the literature by close to 3%. To avoid vas-
cular damage and perforation an early reoperation should be considered in the -

A difficult condition described after transanal pullthrough is incontinence. Basically two major reasons are known. Damage to the sphincter complex can be due to dilatation or a direct damage of the sphincter during the initial prepara-
tion of the muscular cuff. If the inci-
sion is not respecting the natural plane between mucosa and muscle, the damage to the sphincter structures can occur. We would like to emphasize like other au-
ters that using saline submucosal injec-
tion with or without adrenaline is very helpful to discriminate the mucosa from the underlying muscular structure. This dissection can be very difficult after previ-
ous full thickness biopsies and scoring. Incontinence due to sphincter damage is extreme difficult to treat and might beco-

in a lifelong handicap (15). The second condition leading to incontinence is an ana stomosis distal of the dentate line (16). This occurs, if the initial circular incision is not performed above the den-
tate line, but to distally. This problem is also well known in patients operated for inflammatory bowel disease with a total colectomy and ileo-anal anastomosis. In this condition the sensitive continence organ is destroyed and patients are not able to control especially losses of liqu-

The bowel contains large masses of stool and might create very often a complicated condition leading to inconti-
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In many cases these problems are functional and temporary, but in all cases the first approach, before a symp-
tomatic treatment is started, should be a research on complications and pitfalls during the different steps leading to the treatment of HD.

Discussion
In our experience and as described by others, the successful initial approach can guarantee in most cases a good result for the future quality of life for the pa-
tent. Some complications during diagnosis (missed aganglionosis or hypogangli-
onosis) or during the surgical approach (vascular damage, death to the sphinc-
ter, fistula, stenosis, abscess formation) might create very often a complicated situation for the patient, which leads to a life long handicap. Surgery, resulting in incontinence and soiling for the patient a severe and disastrous condition (19).

Table 2
Algorithm for diagnostic and therapeutic approach in children with post op obstruction

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KOMPLIKACIJE HIRSCHPRUNGOVE BOLESTI

J. Schleef, D. Olenik


Deskriptori: HIRSCHPRUNGOVA BOLEST, DIJAGNOSTIČKA PROCEDURA, KOMPLIKACIJE, KIRURŠKI PRISTUPI

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