Paediatr Croat. 2013; 57 (Supl 1): 133-138

Pregled
Review

COMPLICATIONS IN HIRSCHSPRUNG'S DISEASE

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Hirschsprung's disease (HD) is a rare disease affecting in most cases the distal part of the large bowel. Different surgical techniques are described in the literature. The post operatively course can be complicated by surgical and non surgical complications. These problems are discussed and described. Pitfalls and errors in diagnostic and therapy can lead to severe complications like incontinence. A correct diagnostic approach, including in all cases a reliable histochemical work up and a standard surgical approach can avoid many problems and complications in these patients. If redo surgery is necessary, the Duhamel approach is considered as a safe and valuable technique. Nevertheless in some patients also other techniques for special indications like fistulas or stenosis can be very helpful.

Descriptors: HIRSCHSPRUNG'S DISEASE, DIAGNOSTIC PROCEDURE, COMPLICATION, SURGICAL STRATEGIES

Introduction

Hirschsprung's disease (HD) is characterized by missing of ganglia 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. Beside 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). Enterocolitis 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 acetylcholinesterase 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).

Different surgical procedures (Swenson, Duhamel, Soave, Rehbein, Boley) are described in the literature and performed. In the last 20 years different authors introduced laparoscopic assisted techniques, made popular by Keith Georgeson and others. The most recent modification was the introduction of the transanal approach, known as the de la

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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 ganglionotic, 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 resection right above the sphincter complex with an end-to-end anastomosis. Soave's techniques ends up with a similar anastomosis, but leaves the cuff of aganglionotic muscle inside, which is divided and opened. The Duhamel leaves the rectum with aganglionotic 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, preparating 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 problems of surgery for Hirschsprung`s disease. 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 surgical early and late surgical complications should be minimized. All this factors finally contribute to a good quality of life. The purpose of this article is to emphasize 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).

Figure 1
Extramucosal laparoscopic biopsies taken with 3 mm instruments before starting a laparoscopic assisted pull through.

Diagnostic Pitfalls

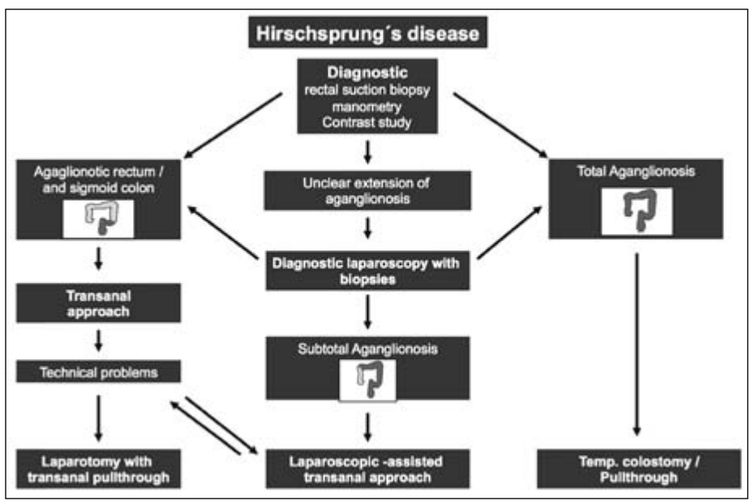
The diagnostic approach for Hirschsprung's disease is represented by clear and precise algorithms. This can be described as a diagnostic triad for HD including x-ray contrast studies, manometry and finally the histochemical and histological work up.

X-ray studies should always be performed without prior cleaning of the colo-rectum and with sufficient contrast media to demonstrate not only the recto-sigmoid region but also the more proximal part of the colon. In some instances the real extension of the aganglionotic zone is not seen, because of an incomplete 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 should 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 manometry. In smaller children, without rectal dilatation, a repeated manometry can be diagnostic, verifying the absence of a relaxation reflex in the internal sphincter. It has to be sad, that a manometry study alone is not sufficient to verify or to exclude HD.

The third column of HD diagnostic is the bioptic study of tissue gained by rectal suction biopsy and segmental, extramucosal biopsy. HD is part of a group of so called intestinal dysganglionoses being part of a heterogeneous group of enteric nervous system anomalies including also intestinal neuronal dysplasia (IND), internal anal sphincter achalasia and different forms of hypoganglionosis. The most important diagnostic features for Hirschsprung's disease are the combination of hypertrophic nerve trunks (acetylcholinesterase staining) in the in lamina propia mucosae and aganglionosis in adequate specimens (absence of

Table 1
Algorithm for diagnostic and therapeutic approach to Hirschsprung's disease.



gangliar structures). Moreover it has to be stressed that a precise determination of the extension of the aganglionosis should be achieved before starting surgery by fresh frozen section. The extension of aganglionosis and proximal dys-or hypoganglionosis and the so-called transition zone is not always correlating with x-ray studies (2, 7). In cases with unclear and untypical extension a prior diagnostic laparoscopy to get histological specimen for further studies might be a choice (Figure 1). The Table 1 gives a picture of the algorithm for diagnosing and treating HD patients in our experience.

The x-ray exam (Figure 2) shows a persistent constipation in a boy after De la Torre pull through. The histological work-up finally showed a near total severe hypoganglionosis of the colon, which was not diagnosed during frozen section exam and not diagnosed during the histological exam of the specimen after surgery. The child was reoperated by an extended resection of the colon after repeated laparoscopic and transanal biopsies. Biopsies are part of every diagnostic work up for HD and are the Gold standard for the diagnostic process. In cases with unclear extension and unclear histological findings, repeated biopsies should be performed. The help of an experienced pathologists is extremely important to avoid mistakes or pitfalls prior or during the surgery (6).

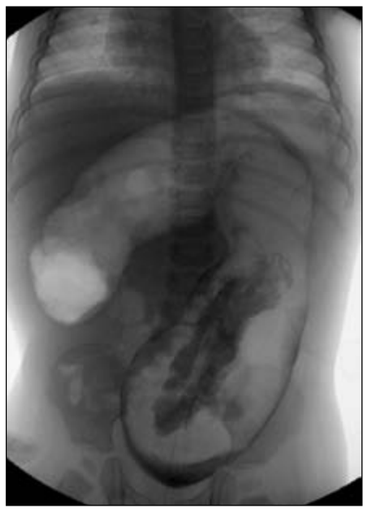


Figure 2
4 months after De la Torre pull through. Non recognized total hypoganglionosis on histochemical exam. Patient was operated twice, ending up with a near total colectomy.

Intraoperative complications

As in any other surgical procedure, intraoperative complications might occur and can be responsible for tragic post-op courses and reoperations. Many of these are general complications like bleeding, injury to other organs or not recognized damage to the bowel remnant, left in place. All minimal-invasive-techniques, as well as the transanal approach have the advantage of reduced tissue trauma, less stress due to surgery and better cosmetically results. But it should be emphasized that not all patients are suitable for these procedures. A transanal approach in elder children with a massive dilated colon can be extremely difficult, leading to damage of the sphincter muscle complex due to stretching (8).

Pelvic abscess formation, observed by some authors, is usually due to bleeding, haematoma and consecutive infection. This bleeding might occur from the incision of the muscle cuff or from the mesentery. In these cases a thorough control of the resection margins should be performed and these might be secured with haemostatic stiches. In some instances cutting and sealing devices might not be secure if the tissue is divided under tension. If there is any doubt, we will recommend in all these cases a laparoscopic control of the pelvic region. Also a mucosa remnant can be the cause of e mucocele and pelvic abscess formation (9). If the preparation of the anterior part of the mucosectomy is difficult, especially in small girls a vaginal damage and a consecutive fistula can be overseen. Figure 3 shows a case of a vaginal fistula in an 18 months old girl with Down-syndrome. The fistula was repaired by extensive mobilization, re-pull through and reconstruction of the perineum. A protective colostomy was necessary.

Another important intraoperative complication is the twisting of the bowel, performing a pullthrough. In a standard transanal de la Torre approach a torsion of the bowel can be possible. If the anatomy is unclear, the bowel dilated or the perfusion of the bowel unsecure, we recommend a laparoscopy during the pullthrough. In long HD, with the necessity of the mobilization and dissection of the transverse colon we would recommend also a combined laparoscopic approach. Many vascular problems, causing severe anastomotic complications with dehiscence and stenosis are due to insufficient mobilization of the mesentery and subsequent traction on the anastomosis and possible retraction of the neorectum (10).

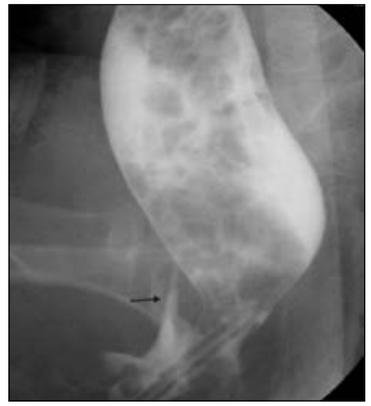


Figure 3
Rectovaginal fistula in a girl with Down's syndrome. The Defect was repaired by a redo-Soave pull through with a protective colostomy.

In some cases, segmental extramucosal biopsies without opening of the lumen are performed. An experienced laparoscopic surgeon should have the capacity to suture the defect in case of opening the bowel. An overseen opening of the bowel might lead to peritonitis and severe septic complications. A good alternative is the technique, described by Langer, bringing the bowel through the umbilical incision and performing the biopsy under open vision.

A comment should be made on enterostomies. In some instances, especially very small and septic children with enterocolitis and bad conditions, an enterostoma might be a fast and live saving procedure. Nevertheless, in these cases some important aspects should be considered. In unclear extension and suspected long extension, an ileostomy is the

safest choice to avoid a stoma in an aganglionotic segment. This can be performed as a double lumen stoma or a loop ileostomy. A biopsy should always be taken at the stoma site for histology. Colostomies, primary or protective should be performed regarding the recommendations of the literature. Stenotic stomas, retracted colostomies or a stoma under tension with a vascular damage can cause severe complications and problems, leading in many cases to the necessity of a reoperation to resolve stoma complications. Especially in small children a stoma prolapse is a frequent problem. Many recommendations in the literature can be found, but unfortunately a stoma prolapse cannot be avoided in all cases. In difficult situations a local revision of the stoma might be necessary or the decision of a definitive surgery might be anticipated (7, 8).

Post op complications

In the post op period all general complications after abdominal surgery can occur (haematoma, infection, bleeding). Post-op bowel occlusion seems to be less frequent after laparoscopic and trans anal pullthrough surgery. In cases of prior severe enterocolitis and previous surgery the incidence of bowel occlusion due to adhesions is described in the literature by close to 2%. To avoid vascular damage and perforation an early reoperation should be considered in these cases. The Figure show the x-ray of a boy, 10 days after laparoscopic assisted pullthrough. A bowel occlusion was found during the subsequent surgery. In some cases, a part of the small bowel can slip under the mesentery of the distal colon and might cause the occlusion. This problem can be resolved by stiches, anchoring the neorectum to the presacral fascia structures (11).

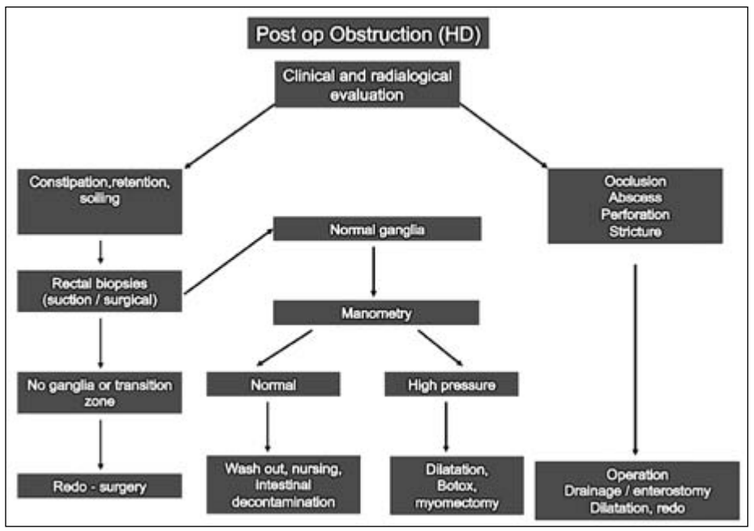
Perianal excoriation can be seen in at least one third of HD patients after surgery. In long HD after extensive colon resection, and especially in total colonic resection this complication is more frequent and also difficult to treat. Using barrier creams can resolve these problems. In some instances is might be helpful to reduce the amount of bile salts using cholestyramine. Barrier creams, used by stomal therapists can be very helpful. (12) Usually the problem of these excoriations are self limiting, since the bowel is adopting and resorbing more and more liquids and the stool is getting a normal consistence. If the stool is continuing to be liquid enterocolitis should be suspected and treated by bowel washing and antibiotic therapy including an oral decontamination of the bowel (13).

Long term problems

Some patients with HD are continuing having some kind of obstructive, functional or inflammatory complication after surgery. These problems can occur after a period of well being or are initially of minor importance but can be affecting the long term quality of life. Not only obstipation but incontinence or pseudo-incontinence can occur and are extreme difficult to cure (14).

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 preparation of the muscular cuff. If the incision 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 authors, that using saline submucosal injection with/or without adrenaline is very helpful to discriminate the mucosa from the underlying muscular structure. This dissection can be very difficult after previous full thickness biopsies and scaring. Incontinence due to sphincter damage is extreme difficult to treat and might become a lifelong handicap (15). The second condition leading to incontinence is an anastomosis distal of the dentate line (16). This occurs, if the initial circular incision is not performed above the dentate 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 these condition the sensitive continence organ is destroyed and patients are not able to control especially losses of liquid stool. The line of the incision of the mucosa for preparing the mucosal cuff should be above the dentate line, ending with anastomoses above the dentate line within the anal canal. The distance from the dentate line should be between 0.5 cm in small children and at least 1 cm in elder children. An anastomosis between anal skin and mucosa creates a situation of permanent incontinence with continuous leakage and uncontrolled loss of

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



liquids and stool. This soiling is extremely disturbing for the patient (17). In some cases these patients might need a bowel diversion or an ACE approach.

In other cases soiling and pseudo incontinence are due to chronic constipation and is a secondary phenomena. The bowel contains large masses of stool and faecalomas bypassed by liquid stool components. This phenomena of so called "overflow incontinence" is well-known in patients with severe constipation, independent of the underlying pathology. In HD cases this phenomena con by transitorily, functional and has to be treated according to guidelines in constipated patients, or might be in selected cases due to patho-anatomical problems like stenosis, twisted bowel, incomplete resection or persistence of a megacolon. All these patients need a thorough follow up according to guidelines (Table 2). Part of this treatment can be in selected cases the use of botolinum toxin, injected into the sphincter muscle area. The effect is usually transient, but might be a helpful tool in the treatment of sphincter hypertonus or a suspected achalasia of the sphincter muscle (18).

In many cases these problems are functional and temporarily, but in all cases the first approach, before a symptomatic 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 patient. Any complication during diagnosis (missed aganglionosis or hypoganglionosis) or during the surgical approach (vascular damage, damage to the sphincter, 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, is usually for the patient a severe and disastrous condition (19).

The choice of the re-do technique is not uniform in the literature and different authors are describing different solutions. Lewitt is describing a sagittal posterior approach for some special indications (stenosis, strictures) and especially in cases of multiple surgery. In all cases the authors recommend a protective colectomy (16). We had the experience, that in patients with incomplete resection the Duhamel approach can be easily performed and is possible in most cases. If this surgery is without major problems, a colostomy might not be necessary. In patients, operated according to De la Torre, even a second transanal pull through is possible. The transanal approach can also be performed as redo laparoscopically assisted. In cases with fistulas and vaginal lesions we prefer a secondary bowel pull through, but in all cases accomplished by a protective colostomy. This kind of surgery needs a surgical team, having experience with different kind of HD techniques and a center, which is able to have always a frozen section diagnostic during surgery available.

The long term results show, that many other problems not directly related to surgical pitfalls (enterocolitis, constipation, soiling) will usually diminish and in some cases disappear, since the patient is getting older (20).

The surgeon who is dealing with HD patients should be able to deal with all these conservative treatment options as well. Patients should be controlled and followed even as adolescents. A transition of these patients to adult physicians is sometimes very difficult due to the poor knowledge of the disease under adult general surgeons and gastroenterologists.

Autori izjavljuju da nisu bili u sukobu interesa.
Authors declare no conflict of interest.

Acknowledgements

We wish to thank Dr. Elisabetta Cattaruzzi and Dr. Flora Maria Murru (Paediatric Radiology, IRCCS Burlo Garofolo, Trieste, Italy) for their help selecting and preparing the x-ray images.

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Sažetak

KOMPLIKACIJE HIRSCHPRUNGOVE BOLESTI

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Hirschprungova bolest (HD) je rijetko oboljenje koje najčešće zahvaća distalni dio debelog crijeva. U literaturi su opisane različite kirurške tehnike. Poslijeoperacijski tijek može imati kirurške i nekirurške komplikacije. U ovom radu su ti problemi raspravljeni i opisani. Zamke i pogreške u dijagnostici i liječenju mogu dovesti do ozbiljnih komplikacija poput inkontinencije. Ispravan dijagnostički pristup koji uključuje u svim slučajevima pouzdanu histokemijsku analizu te standardni kirurški pristup mogu spriječiti mnogo problema i komplikacija u ovih bolesnika. Ako je potrebna reoperacija, Duhamelova procedura se smatra pouzdana i vrijedna tehnika. Ipak u nekih bolesnika s posebnim indikacijama poput fistula ili stenoza druge tehnike mogu biti od velike pomoći.

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

Primljeno/Received: 15. 3. 2013.
Prihvaćeno/Accepted: 2. 4. 2013.