

Iraqi Experience Of Performing Rehbein S Procedure As A Surgical Treatment Of Hirschsprung S Disease At Central Child Hospital

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Summary:-

Background: Since Hirschsprung s disease is a complicated and frightful congenital anomaly to the parents , every effort should be taken to reach the optimal result that is beneficial to the patient, parents and surgeon also . Its incidence is 1; 5000, more common in male. The principle for treatment by Rehbein s procedure is to remove the aganglionic narrow segment including the dilated sigmoid colon by dissection of the upper rectum deep down into the pelvic cavity about 2 cm from peritoneal reflection and to elimination of achalasia of internal anal sphincter by vigorous dilatation .

Objective:- Assessment of the advantage of the Rehbein procedure in treatment of Hirschsprung disease

Patients and methods:- 50 cases (44 male and 6 female) with Hirschsprung s disease presented with positive history of delayed passing meconium , clinical symptoms and Ba-enema (different level of narrow segment) treated by Rehbein s procedure are presented at central teaching hospital for pediatrics between Aug 1998-april 2005

In children failed to thrive, colostomy was performed until their development had progressed well and corresponding to their age Rehbein s procedure was done primarily in 44 cases at the age of 2 months to 3.5 years (88%).

Children who were well corresponding to their age were treated with abdominal resection plus protective loop transverse colostomy at the same time 6 cases (12%)

Results:-39 cases classified as very good (78%); 8 cases classified as good (16%); 3 cases classified as satisfactory or unsatisfactory (6%); No disorders of urinary evacuation such as retention or incontinence were seen. We never observed anal incontinence. In 12 cases(24%), an anastomotic line stenosis was treated by Hegars dilator under general anesthesia and continued at home by the family, No anastomotic leak was found . 8 cases(16%) needed frequent anal dilatation under general anesthesia at intervals. 1 case needed sphincterotomy.

Conclusions:- Hirschsprung disease is not an uncommon disease. Its treatment needs experience and surgery can have uncorrectable complications. I found that this procedure is efficient, easy to perform, and less time- consuming and carries few complications and finally keeps good relation with the families of the children

Keywords ;Rehbein s procedure, Hirschsprung disease

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Introduction:-

Hirschsprung s disease is a congenital anomaly of the colon affecting about 1:5000 live births(1,2). Harald Hirschsprung in1886 gave the first details of congenital megacolon. The term megacolon was introduced by Mya in1894. In 1901,Tittlel described the absence of intramural ganglion cells in the rectum. Ehrenpreis 1946 doctoral thesis, an exhaustive review of the etiology and pathogenesis of Hirschsprung s disease, point out that the diagnosis could be made in the neonate(3,4,5). The aganglionosis lead to sustained contraction of the diseased colon with secondary dilatation and hypertrophy of the more proximal segment.

Congenital megacolon was named after this secondary change(6). In 1948, Swenson and Bill presented a curative operative technique. Professor Fritz Rehbein developed a procedure for hirschsprung s disease that bears his name in 1953. During his career he successfully used this operation on nearly 400 children. His procedure remains the operation of choice in many pediatric surgical centers worldwide(3)

Duhamel in 1956 proposed partially bypassing the rectum and performing the anastomosis with the use of crushing clamps. Soave in 1962 applied the endorectal pull- through procedure for Hirschsprung s disease(7,8,9,10) .

The principle for the treatment of Hirschsprung s disease is to remove the aganglionic narrow segment including the dilated sigmoid and to eliminate the achalasia of the internal sphincter(4). The purpose of the present study was to re-

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emphasize the author's modification of the Rehbein's procedure and report the result of treatment with Rehbein's procedure in central teaching hospital for pediatric.

Patients and methods:-

50 patients have been diagnosed as cases of hirschsprung's disease either by full thickness rectal biopsy (3 cases) or by barium enema (47 cases). All of these patients have positive past history of delay in passing meconium in the first 24 hours after delivery.

After diagnoses and confirmation, the principle step was establishing a defunctioning of the affected colon by making a loop colostomy performed in the transverse colon together with employing a laparotomy operation for taking multiple seromuscular biopsies from different parts of the sigmoid, colosigmoid junction and the colostomy site (First. Stage operation).

The patients then were left for gaining acceptable weight then the patient will be ready to undergo a surgical correction of his anomaly which include resection of the affected (aganglionic) colon together with part of the proximal dilated, nonfunctioning colon and anastomosing the rest of the colon to the rectum (deep anterior resection – colorectal anastomosis) using Rehbein's procedure (2nd stage operation). Then after one month the anastomotic line checked by PR, If it is good and no leaking is present, the colostomy is ready to close (3rd stage operation) 44 cases (88%) out of 50 underwent three stages operation. The other 6 (12%) cases had been treated by 2 stage operation either

A- When the sigmoid is hugely dilated, sigmoidectomy done plus the descending colon delivered as an end colostomy until the child were left for gaining acceptable weight then ready to undergo a surgical correction by Rehbein's procedure (. 4 cases 8%). OR

B- Rehbein's procedure plus protective loop transverse colostomy done at the same time then followed by closure of colostomy (2cases 4%)

Surgical procedure

Anesthesia

We used general anesthesia without pre medication

Position

Supine with slight extension of the back using sand bags

Incision

Most of the patients operated upon through lower left Para median incision

A-For 3 stage operation (colostomy + multiple colonic biopsy followed by rehbein's procedure then closure of colostomy) this done for child who

presented with failure to thrive, malnourished, under weight corresponding to his age

In these cases previously known to be resected at the level of colosigmoid junction

(Depending on Ba –enema), no patient preparation is needed to be done preoperatively, the preparation is done peroperatively as follow

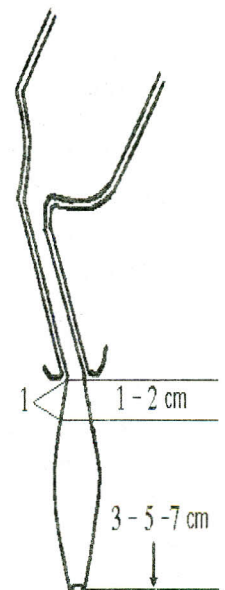
1- To start with cleaning the lower rectum and anal canal by mopping it with Betadin swabs leaving a small pack of gauze inside (about 5 cm from the anal canal).

2- After resection of sigmoid and aganglionic segment, the lower segment of the rectum and the anal canal also mopped by betadin swabs after removal of the gauze pack that has been left previously. The proximal colon which is arranged to be anastomosed with the lower rectum is also washed by normal saline and betadin swabs several times until the rest of the colon is clean completely from the colostomy site to the site of resection

If the colostomy site is arranged to be anastomosed with lower rectum, patient in this case need to be prepared by oral manitol with control of fluid suppletion

B-In case of two stage operation (Rehbein's procedure + protective loop transverse colostomy at the same time followed by closure of colostomy)

The preparation will be the same as above except that keeping the child on fluid diet and colonic enemas for 3 days duration, all the cases received a preoperative oral antibiotics which were trimethoprim, metronidazole according to their weight with the use of one injectable dose of cefotaxim preoperatively.



A segment 1 to 2 cm is dissected

free below the peritoneal reflection (1).

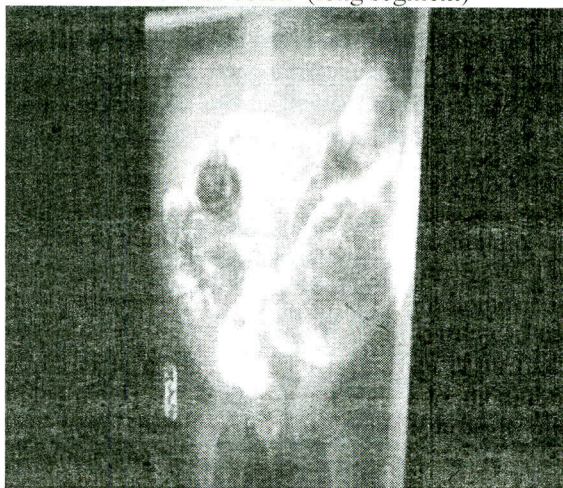
The distance of the cut level from the anal orifice varies according to the child's age .

It is about 3 cm in infant and somewhat greater in old children .

After dissection of peritoneum as shown in the diagram above the rectum now resected with narrow aganglionic segment +dilated sigmoid up to the level of aganglionic presence al ready marked in the first operation then fallowed by anastomosis in one layer by long absorbable suture starting in the posterior layer then anterior layer then drain fixed in the pelvis, we used to put a wide bore tube from the anus passing up to colon through the anastomosing line

Aganglionic segment level

- 42 cases(84%) were close to colosigmoid junction
- (short segment)
- 8 cases (18%) were sigmoid, left colon and left
- transverse colon (long segment)



Complication:-

- 1- Skin infection 3 cases (6 %).
 - 2- Wound dehiscence, 2 cases (4 %).
 - 3- Anastamotic line stenosis .12 cases (24%) were treated by dilatation under G.A. by Hegar's dilator then continue at home by family.
 - 4- No anastamotic leak.
 - 5- Recurrent obstruction: - 8 cases (16%) need frequent dilation for anal sphincter under GA after certain intervals In Hirschsprung s disease 2 components are blamed as cause of symptoms
 - 1- agangloinc segment
 - 2- achalasia of the internal anal sphincter,
- Professor Rhebien regard the main problem after surgical treatment of Hirschsprung s disease to be the gradual recurrence of constipation. The reappearance of disorders of stool evacuation cannot attributed to inadequate resection of the narrow segment, leaving in aganglionic rectal tissue, but is due to exclusively to the return of tenacious sphincter achalasia.

Rehbein was convinced that the central problem in the treatment of Hirschsprung s disease after the removal of narrow segment was the permanent elimination of the sphincter achalasia(4)

. A special collagenous fibrous layer called the adventitia rectalis was found to limit the free expansion of the rectal wall. This tissue plays an important role in bowel movement by defining a threshold of intrarectal pressure in the bowel reflex above of defecation and by transmitting the expulsive force from the abdomen above down to the anal opening. This discovery may explain why in Hirschsprung s disease, the preservation of the rectal wall in Rehbein s procedure will preserve better bowel function(11) (5) Out of (8) required no more than one dilatations. 2 patients required, tow dilatations, in only 1 patient internal sphincterotomy was required after failer of three time dilatations

Results

1-The evaluation of patients after surgery include 4 categories :

A- Very Good " implies that stool evacuation occurs without difficulty and with no auxiliary , in otherwords. Essentially normal , (39 cases 78%) ,

B- Good " an occasional laxative was necessary (8cases,16%)

C- Satisfactory "had bowel movement without difficulty when laxatives given regularly (3cases6%).

D- Unsatisfactory,, indicate that the patient had no adequate bowel evacuation despite the measure indicated above (2cases 4%)

When the patients with very good and good results are combined, the resulting figure is 47 (94%)

One case needed internal sphincterotomy in spite of frequent dilatations.

2-Mortality_isZero.

3-No disorder of urinary evacuation in the sense of retention or incontinence.

4-Never found symptoms of anal incontinence.

Discussion

Since Hirschsprung disease is a complicated and frightful congenital anomaly to the patient, every effort should be taken to reach the optimal result that is beneficial to the patient, parents and the surgeon.

So we found the fallowing points are support us to use Rehbein s procedure for the treatment of Hirschsprung s disease

1- The autonomic nerves to the anal sphincter, bladder and genital organs which run in small pelvis are not endanger.

2- There is no contact with the anal sphincter and thus anal incontinence is avoided.

3- Technical performance of the operation is simple.

4 - Position of the patient is restful to the surgeon (only abdominal).

5- No need to use Foley s catheter during the operation but if we put it, can be removed next morning after operation.

6- Less time consuming and carries less complication.

7- Preparation can be done at operation.

Conclusion

Hirschsprung s disease is one of the complicated congenital anomaly which is not uncommon in the population, since 1886 till now. Many procedures used for correction this anomaly. Each one carries many complications and some difficulties in performance. From the study above in using Rehbein s procedure I have found that this procedure is simple, carries less complication. in comparison to other procedure and give a guarantee for anal and urinary continent which not get it in other procedure like Swenson because of there is no contact with anal sphincter and the autonomic nerves to the anal sphincter , bladder and genital organs which run in a small pelvis is not endanger for that no need to use Foley s and if use it can be removed next morning after operation .Finally because of the preparation of the bowel can be done at operation so the patient can admitted to the hospital at the night or morning before operation and no need to admit him 2-3 days before operation for bowel wash

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