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GOOD LESSONS FROM BAD EXPERIENCE. MANAGING CHILDREN WITH HIRSCHSPRUNG'S DISEASE AND ANO-RECTAL MALFORMATIONS

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Abstract: *Surgical management for both Hirschsprung's Disease (HD) and ano-rectal malformations (ARM) is in continuing progress and new techniques are developed, based on the classical ones, but adapted for a minimally invasive strategy. We are presenting a few examples from our series of HD and ARMs patients, the ones with initial unorthodox and unexpected events from different reasons, eventually corrected and with good final outcomes. For some of our patients the immediate outcome included incidents and complications due to a series of misjudgments or technical problems. The incidents are discussed as well as their consequences and the "good lessons" are pointed out for each of these events.*

Key words: *new born, Hirschsprung's, anorectal, management.*

1. Introduction

"Good judgment comes from experience. Experience comes from bad judgment" (Moneer K. Hanna, MD, FRCS, FACS, Clinical Professor of Urology, New York Hospital Cornell University, New York, International Symposium, Mannheim, Germany, 2007).

Surgical management for both Hirschsprung's Disease (HD) and ano-rectal malformations (ARM) is in continuing progress and new techniques are developed, based on the classical ones, but adapted for a minimally invasive strategy [5].

For HD, modern techniques derive from the basic principles of the endorectal pull-through, described by Soave in 1964, and then modified by Boley who performed the coloanal anastomosis during the pull through [1, 3, 6, 10-11-12].

Combined techniques have been described, laparoscopic and endorectal (Georgeson et al) or just endorectal pull-through in newborns without a previous colostomy (So et al). But the first ones to report HD patients managed by an exclusive transanal approach were De la Torre-Mondragón and Ortega-Salgado in 1998, who performed mucosectomy, colectomy and pull-through in this manner on several cases [2, 4, 9].

As for the ***ARMs***, since 1982, when De Vries and Pena exposed their new anatomical concept, developed and popularized later by Pena through the separate protocols for the management of male and female patients and his posterior

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sagital approach, most surgeons around the world adopted them [7-8].

2. Material and Methods

The paper presents a single surgical team experience (Pediatric Surgery Department, Children's Hospital of Brasov). Since 1990, a series of 42 patients with Hirschsprung's Disease (HD) and 75 anorectal malformations (ARM) have been surgically managed by the same surgical team. For HD the preferred method until 2000 was the Duhamel pull-through procedure with crushing clamp for the colorectal septum, while after 2000 the same procedure had been performed using a single stage procedure thanks to the availability of a modern stappling device, the longitudinal stappler. Since 2000 we also performed several endorectal Soave pull-through procedures with encouraging results.

As for the ARM, since 1994 our preferred approach was the Posterior Sagital Anorecto Plasty (PSARP), popularized by Pena [7], both for male and female patients, the great majority with an initial sigmoid double-ends colostomy, as indicated and described by the same surgeon. Until 1994 our approach inconstantly included an initial colostomy, but some "bad" experience taught us the "good lesson" for ARMs.

Four representative cases have been selected from the above mentioned patients and the "bad experiences" were exposed as Results and Discussion, followed by the "good lessons" we learned, stated in the Conclusions chapter.

3. Results and Discussions

The "Bad" Experience Nr.: AN., female new born baby, 24 hours after delivery, admitted for: very distended abdomen, bilious vomiting since 5 hours and no meconium discharge. Upon admission a KUB (plain abdominal X-ray) revealed very distended loops on the whole abdominal area, while a contrast enema confirmed long segment Hirschsprung's disease (transition zone by the middle of the descending colon).

The initial 2 weeks were managed with "nursing". At 14 days a Duhamel PT was performed with resection of the aganglionic segment and application of a crushing clamp on the colorectal septum. At 48 hours postoperatively: suspect discharge through the abdominal drainage catheter was noticed and a second look laparotomy had to be performed: colorectal septum earlier and completely crushed, but no sealing of the lateral margins. The posterior wall of the split rectum had to be repaired and an end colostomy on the colon was performed at the end of the operation. The immediate follow up was eventless.

At age 8mo. (10kg) a second PT, a Soave-Boley ERPT using the left colon (mobilization beyond splenic flexure). At present she is in her 4th grade of Primary School, continence much improved (used to soil her underwear from time to time with small amounts of feeces).

The "Bad" Experience Nr. 2: B.S., 13 mo.old baby boy, presenting with constipation since he's been born and empiric treatment until 6mo. GP's nursing recommendations followed up to 12 ½ months when first seen in a Pediatric Unit. Radiology investigations (barium enema): confirm HD with hugely dilated bowel above the narrow aganglionic sigmoid segment.

Referred to our Surgical Dept., intensive nursing for two weeks: rectal washouts twice a day, then daily, lactulose and mineral oil (paraffin) also. Scheduled for operation after two weeks of intensive conservative management, bowel preparation done on table with saline and Betadine^{R}. At operation, much dilated sigmoid and ascendent colon above a narrow recto-sigmoid segment was noticed and a very large descending colon with thick wall (hypertrophy) with a transition zone very well marked.

Next day after surgery everything seemed fine, but 48 hours after surgery, suspect discharge comes through the catheter left on site. A second look laparotomy was decided and a small amount of brownish fluid on the sides of the pulled colon, deep into the pelvis was noticed, but the rectal stump was leak proof. Leakage from the thick septum stappling was suspected. Bilateral pararectal drainage catheters and a protection transvers colostomy was performed during the same operation.

At six weeks a massive prolapse on the distal end determined the parents to bring the child back to the hospital and a modified Tiersch procedure for prolapse contention was performed under general anesthesia, which partially failed after 4 days and the prolapse recurred. A contrast study of the rectum, both from below and above revealed no intra-abdominal or para-rectal leakage present. Eventless follow up, BMs after 24 hours, oral feeding normal. Discharged with mild perineal irritation due to 4-5 softer stools/day.

The "Bad" Experience Nr. 3: D.A., new born baby girl, admitted for ARM: recto-vestibular fistula. No previous colostomy performed. A rectal transposition performed at 24 hours, no complications after surgery, until 48 hours later when stools were present: contaminated wound, infection, sutures disruption. A sigmoid colostomy was urgently needed.

After 10 days, when wound infection cleared out, re-suturing of the perineal wound was possible and the child was discharged after another 7 days, with dilatations program for a few months. Now, she is 10 years old, very rarely presenting minor soiling when very soft stools.

The "Bad" Experience Nr. 4: P.G., 3 days old baby boy, admitted for ARM, apparently a low malformation. On a W.R. invertogram, a very large rectum with blind end apparently close to the perineal skin was noticed. No meconium came with urine, no gas in the bladder noticed on the X-ray. No previous colostomy was considered and a standard proctoplasty was performed with no immediate incident.

Next day after initial surgery, irritation of the perineal skin occured and urine leaking through the newly made anus was noticed from fistula with the urinary tract not seen before or during the initial surgery. Urgent surgery was needed to protect the proctoplasty: a colostomy on the descendent colon and a bladder suprapubic catheter.

In spite of the protective measures, the initial procedure had been compromised. A contrast examination via the distal end of the colostomy (colostogram) revealed the real anatomy of the malformation: ano-rectal agenesis with a recto-bulbar urethra fistula. The dilated end of the rectum close to the skin on the initial X-ray represented the dilated fistula (due to late presentation with huge rectal distension).

This type represented a high ARM with rectum at the levator level. PSARP was performed at four months of age (over 6 kg), after which an anal stricture developed and dilatations were not successful after two months. Thus, a modified "Z" plasty was performed at age 12 months and a colostomy closure after 4 weeks from the last procedure.

Dilatations continued based on Pena's recommendations for two years, and at 7 years the perineal aspect is almost normal, the boy goes to school, presenting little soiling if not able to go to the toilet soon after urgency was perceived.

4. Conclusions

In case there are several authors, there will be written the first 2 authors followed

The Good Lessons for H.D.:

- Do not dare a Duhamel PT in the new born or young infants unless you have adequate technical conditions.
- The Soave-Boley ERPT is more suitable for small infants (2 weeks or older) with long narrow segments of aganglionosis.

- TEPT (Mondragon&Salgado) is not suitable for long segments, unless long experience and frozen sections available.
- For patients presenting with very large, hypertrophied dilated segments, do not hesitate to perform a previous colostomy to deflate the dilated colon and make a much easier and safer job during the PT.
- Always place a drainage catheter after the PT.
- Prefer a separate, double ends colostomy – double barrel colostomy much prone to a prolapse, more difficult nursing.

The Good Lesson for ARM:

1. Always perform an *initial colostomy* for any ARM, regardless sex of patient or type of anomaly. Advantages of an initial colostomy:
   - Offers the opportunity to diagnose /treat other prioritary conditions: associated anomalies, prematurity, infections, etc.
   - Best way to investigate real anatomy of the malformation - *colostogram* and adopt the right strategy for the PT.
   - Best protection after major surgery or unexpected complications
2. For ARMs always *a high sigmoid colostomy*, as described by Pena [7]:
   - Easy to care, either with bags or diapers.
   - Easy cleaning of the rectal pouch
   - Enough distal segment length for comfortable PT.

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