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REVIEW ARTICLE

Update on the management of anorectal malformations

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Abstract Thirty-three years ago, on 10 August 1980, in Mexico City, the first patient with an anorectal malformation was operated on using the posterior sagittal approach. At that time it was not obvious that we were actually opening a "Pandora's box" that continues to give many positive surprises, a few disappointments, and the constant hope that each day we can learn more about how to improve the quality of life of children born with all different types of anorectal malformations. In November 2012, patient number 3000 in our database was operated in the city of Cochabamba, Bolivia; during one of our International Courses of Anorectal Malformations and Colorectal Problems in Children. The goal of this article is to give a brief update on the current management of patients with anorectal malformation, based on the multiple lessons learned during this period.

Keywords Anorectal malformation · Imperforate anus · PSARP · PSARVUP

Thirty-three years ago, on 10 August 1980, in Mexico City, the first patient with an anorectal malformation was operated on using the posterior sagittal approach. In January 1982, the first patient with a cloacal anomaly was repaired using the posterior sagittal approach, in the city of Monterrey, Mexico (Fig. 1). At that time it was not obvious that we were actually opening a "Pandora's box" that continues to give many positive surprises, a few disappointments, and the constant desire and hope that each day we can learn more about how to improve the quality of life of children born with all different types of anorectal malformations.

In November 2012, patient number 3000 in our database was operated in the city of Cochabamba, Bolivia; during one of our International Courses of anorectal malformations and colorectal problems in children (Fig. 2).

The goal of this article is to give a brief update on the current management of patients with anorectal malformation, based on the multiple lessons learned during this period.

Prenatal diagnosis [1–4]

Currently, the most complex anorectal malformations are the ones that can be most often diagnosed prenatally. The reason for this is the fact that the higher the malformation (recto-bladderneck fistula in males, cloaca in females) the higher is the presence of associated anomalies, and many of these associated defects can be seen in utero.

During the prenatal imaging study, one important clue to suspect an anorectal malformation is the finding of multiple systems with abnormalities (digestive, vertebral, genitourinary).

The advantages of having a prenatal diagnosis include: giving the parents some information about the type of anomaly that the patient will be born with and also giving them the opportunity to make arrangements for the baby to be delivered in a specialized center that is familiar with the neonatal management of patients born with these conditions.

Images that can be seen prenatally and should raise suspicious for an anorectal malformation include: dilated and or calcified bowel, lack of meconium at the expected rectal level, hydronephrosis, absent kidney, neural tube defects, tethered cord, hydrocolpos, vertebral anomalies,

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Fig. 1 First cloaca operation done posterior sagittally. Monterrey, Mexico; January 1982

Fig. 2 Case number 3000. Cochabamba, Bolivia; November 2012

absent radius, and omphalocele in the absence of bladder visualization.

Neonatal management

During the first 24 h of life it is important to rule out associated malformations that might be life-threatening. With an echocardiogram the physician will rule out cardiac conditions, a naso-gastric tube should be passed to rule out esophageal atresia, an abdominal radiograph should rule out duodenal atresia, a kidney ultrasound should rule out severe hydronephrosis, and a pelvic ultrasound in females born with a cloaca should rule out a hydrocolpos. A sacral radiograph film in antero-posterior and lateral views will allow for the calculation of the sacral ratio (Fig. 3), which is an important tool to predict the future prognosis for bowel control. A spinal ultrasound should be ordered to rule out tethered cord.

During the first 24 h the surgeon will also have enough information to decide between a primary repair or a colostomy. This decision should take into consideration the experience of the surgeon, the condition of the baby, and the circumstances surrounding the patient and the surgeon.

Common indications for a colostomy include: flat perineum, meconium in the urine, distal gas on the cross table lateral film taken after 24 h of life above the coccyx or high in the pelvis, and cloaca.

We firmly believe that following these principles and with a good index of suspicion the surgeon can establish the future functional prognosis of the baby, which is extremely important to adjust the parents' expectations.

Colostomy and hydrocolpos drainage [5, 6]

In anorectal malformations the ideal colostomy must be completely diverting, leaving enough distal bowel to allow for the future pullthrough. Both stomas must be separated enough to accommodate a stoma bag that only covers the proximal stoma.

Our recommendation is a descending colostomy taking advantages of the peritoneal attachments of the descending colon to avoid prolapse of the proximal stoma and making the mucous fistula as tiny as possible to avoid prolapse of the distal stoma (Fig. 4). During the colostomy opening, the distal bowel should be irrigated with large amounts of saline solution to clear it from any distal meconium.

Loop, Hartman pouch, and transverse colostomies are contra-indicated.

In patients with cloaca, during the first 24 h of life a pelvic ultrasound should be ordered, specifically looking for a pelvic cystic mass behind the bladder. If a hydrocolpos is diagnosed, it should be drained at the time of colostomy opening with a trans-abdominal indwelling tube that should be left in place until the time of the main repair (when the patient will have a vaginal opening created).

High-pressure distal colostogram [7, 8]

The high-pressure distal colostogram is the most valuable diagnostic study to determine the specific type of anorectal malformation in male patients (the precise location of the fistula), the length of bowel available for the pullthrough, and the relationship between the sacrum, the coccyx, and the rectum. All this information is vital to plan the operation (laparotomy, laparoscopy or posterior sagittal approach). In addition, it allows for the determination of the future functional prognosis.

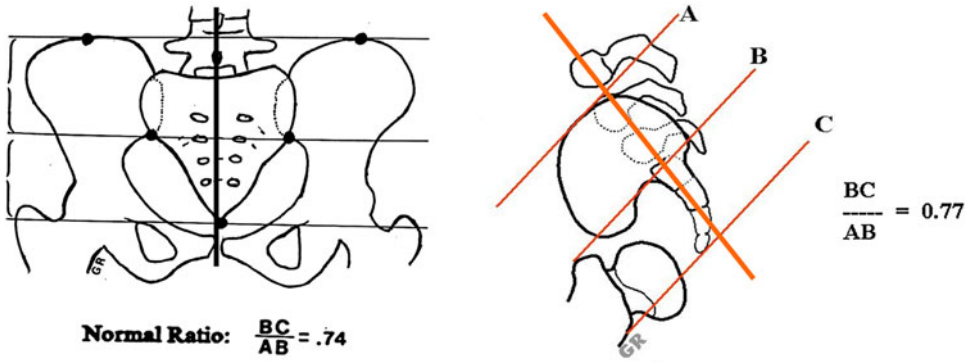

Fig. 3 Sacral ratio calculation in antero-posterior and lateral views

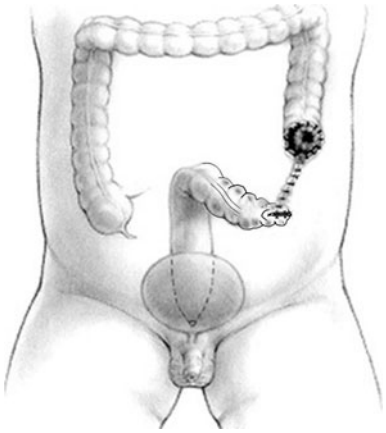

Fig. 4 Descending colostomy with mucous fistula

Surgical repair [9–16]

(a) *Posterior sagittal anoplasty for rectoperineal fistula*: the repair of these defects consists of a small posterior sagittal incision with enough mobilization of the rectum, sufficient to be transposed and placed within the limits of the sphincter. This is a meticulous operation and can be done during the neonatal period without a colostomy. The most common complication during the repair of this defect in male patients is a urethral injury, which can be avoided by placing a urethral catheter and taking particular care during the dissection of the anterior rectal wall.

(b) *Posterior sagittal anorectoplasty for recto-vestibular fistula, recto-urethral bulbar fistula, recto-urethral prostatic fistula, and imperforate anus without fistula*: the key anatomic characteristics that should be kept in mind are that in recto-vestibular fistulas the rectum shares a common wall with the vagina, and in recto-urethral fistulas and imperforate anus without fistula the rectum shares a common wall with the urethra. The surgeon has to make two walls out of one with a careful and meticulous separation of these structures. The posterior sagittal incision in these cases should be long enough to allow for adequate rectal mobilization. The posterior rectal wall should be identified, the lateral walls dissected, and then the surgeon should concentrate on the most delicate portion of the operation: the separation of the anterior rectal wall, without damaging the urethra in males and the vagina in females.

(c) *Laparoscopic assisted posterior sagittal anorectoplasty*: in 10 % of the male patients, the abdominal cavity has to be entered either through laparoscopy or laparotomy to repair the anorectal malformation. We consider the recto-bladderneck fistulas the ideal indication for laparoscopy as well as some selected recto-urethral prostatic fistulas. Laparoscopy is contra-indicated in recto-perineal fistula, recto-vestibular fistula, imperforate anus without fistula, and recto-urethral bulbar fistula, as it risks injury to the urethra or leaving behind the distal rectum which will form a posterior urethral diverticulum, in male patients.

(d) *Posterior sagittal anorectal–vaginal–urethral plasty with total urogenital mobilization for cloaca with a common channel length of <3 cm*: before undertaking repair of a cloaca, the surgeon should perform an endoscopy with the specific purpose of determining the length of the common channel. Cloacas with a common channel length of <3 cm, represent more than 50 % of all cases.

After opening posterior sagittally usually the first structure to be identified is the rectum and it should be separated from the vagina in the same way as in cases of recto-vestibular fistula. The total urogenital mobilization consists of mobilization of both the vagina and urethra as a unit without separating them, by dividing the suspensory ligament of the urethra and bladder. In this type of cloaca a laparotomy is usually not needed.

(e) *Posterior sagittal anorectal–vaginal–urethral plasty with laparotomy for cloaca with a common channel length of more than 3 cm*: the repair of these complex defects requires the implementation of a rather

complicated decision making algorithm. When the total urogenital mobilization is not enough for the urethra and vagina to reach the perineum, carving the pubic cartilage and making a Heineke-Mikulicz maneuver in the vagina may give extra millimeters, when that is not enough, an extended trans-abdominal total urogenital maneuver is performed. If the structures still do not reach the perineum, the most challenging maneuver should be done and it consists in the separation of the vagina from the bladder. To do that, the bladder must be open and catheters have to be inserted into the ureters. At this stage, if the vagina still do not reach, depending on the anatomy, a vaginal switch maneuver can be performed or a partial vaginal replacement using rectum or colon.

Reoperations [12, 15, 17]

Reoperations in anorectal malformations are divided in those performed to repair an anatomic problem consecutive to a surgical complication. These complications include: anal stricture, acquired anorectal atresia, anal mislocation, rectal prolapse, posterior urethral diverticulum, and persistent urogenital sinus in patients with cloaca.

Posterior urethral diverticulum was previously seen when abdomino-perineal operations were the standard of care in anorectal malformation, and recently we have seen an increase in the number of cases again due to the use of laparoscopy to repair patients with recto-urethral bulbar fistula.

These kinds of reoperations should not be offered in an attempt to improve bowel control, since we do not know the degree of damage done in the original operation.

Reoperations to try to improve bowel control are currently done by us only in patients that have the following characteristics: normal sacrum, no tethered cord, intact anal sphincter with a completely mislocated anus, and that the rectum has not been resected. Only about 50 % of patients reoperated following these indications reported a significant improvement in fecal control. This emphasizes the importance of having the first surgical procedure done correctly.

Colostomy closure [18]

Colostomy closure is a procedure that is frequently done in pediatric surgery, and carries a high morbidity rate. However, we believe that those complications can be avoided by observing a meticulous surgical technique.

We have published our technique for colostomy closure that consists in packing of the proximal stoma, using a plastic drape to immobilize the surgical field, multiple silk sutures in the muco-cutaneous junction of the stomas to provide uniform traction that allows the surgeon to identify the correct dissection plane, remaining as close as possible to the bowel wall, careful hemostasis, emphasis in avoiding contamination, cleaning the edge of the stomas to allow a precise anastomosis; a two-layer, end-to-end anastomosis with separated long-term 6-0 absorbable sutures, generous irrigation of the peritoneal cavity and subsequent layers with saline solution, closure in layers to avoid dead space, avoidance of hematomas, and wound coverage with Dermabond®.

Management of the sequelae (constipation and fecal incontinence) [19–25]

(a) *Before toilet training age*: before toilet training it is important to avoid constipation by treating it adequately with laxatives, and to try to achieve regularity in bowel movements, as much as possible, by having meals at the same time each day.

(b) *Treatment of constipation*: patients with good prognosis for bowel control (recto-perineal fistula, recto-vestibular fistula, recto-urethral bulbar fistula, imperforate anus without fistula, with normal sacrum, and no tethered cord) are the ones that suffer from the most severe type of constipation. We consider constipation incurable but manageable. It is important that parents learn about this condition early in life and that doctors teach them how to appropriately manage it adequately with the correct amount of laxatives. These patients usually require laxative dosages much higher than what is conventionally recommended. The dosage of laxative must be individually determined and confirmed to be adequate by taking an abdominal radiograph film to monitor the amount of stool in the colon.

(c) *Bowel Management for fecal incontinence*: patients with poor prognosis for bowel control (recto-bladder-neck fistulas, cloaca with common channel more than 3 cm in length, sacral ratio <0.4, and tethered cord) should be kept artificially clean in the underwear with a daily enema, (after the age of 3 years) at the same age as normal children are out of diapers.

An enema recipe is created, by trial and error, during a 1-week period, specially designed for each patient taking into consideration the characteristics of the colon on the contrast enema (dilated or non-dilated). Rectal administration of this daily enema allows the patient to be clean of stool in the

underwear for a 24 h period, until the time for the next enema.

(i) *Appendicostomy (Malone procedure)*: the appendicostomy or neo-appendicostomy offers a different route (antegrade) for enema administration. We only offer this operation, when the patient has been subjected to a successful bowel management and is old enough to collaborate with the management.

Long-term follow-up [26–29]

(a) *Urologic*: every patient with anorectal malformation is a potential urological patient, since the urinary tract is commonly affected in these patients. In an almost linear way, the frequency of urologic defects increases the higher the malformation (30 % in recto-urethral bulbar fistula, 50 % in recto-urethral prostatic fistula, 90 % in recto-bladderneck fistula and cloaca). The most common anatomic defect associated with an anorectal malformation is an absent or dysplastic kidney, followed by hydronephrosis. The most common functional defect is vesico-ureteral reflux.

In addition to the congenital urologic abnormalities associated with anorectal malformation, the iatrogenic problems, mainly neurogenic bladder are significant. Every patient with anorectal malformation should have a kidney ultrasound as an initial evaluation early in life. If the kidney ultrasound shows abnormalities, the patient deserves a full urologic evaluation.

Even when the initial evaluation shows normal kidneys, all patients must be followed for life to be sure that the kidney function does not deteriorate over time. Patients with anorectal malformations' most significant morbidity is related to urologic problems.

(b) *Gynecologic*: a new chapter will be soon written related with reproductive and obstetric concerns in females born with anorectal malformations. 6 % of patients with vestibular fistula have a vaginal septum (two hemi-vaginas, two hemi-uteri), 60 % of patients with cloaca suffer from this same problem. The presence of two hemi-uteri should alert the surgeon during the time of menstruation for possible obstruction of one of the Mullerian structures with retained menstrual blood causing severe abdominal pain. These patients also carry a higher risk of suffering from a miscarriage and premature labor and should be followed by a specialized obstetrician.

Patients that had a type of vaginal reconstruction (vaginal pullthrough, introitoplasty, or vaginal replacement) should be examined prior to starting sexual activity to insure that the introitus is of adequate size for intercourse.

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