Review
The antegrade continence enema procedure and total anorectal reconstruction

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Patients may present with anal incontinence (AI) following repair of a congenital anorectal anomaly years previously, or require total anorectal reconstruction (TAR) following radical rectal extirpation, most commonly for rectal cancer. Others may require removal of their colostomy following sphincter excision for Fournier’s gangrene, or in cases of severe perineal trauma. Most of the data pertaining to antegrade continence enema (the ACE or Malone procedure) comes from the pediatric literature in the management of children with AI, but also with supervening chronic constipation, where the quality of life and compliance with this technique appears superior to retrograde colonic washouts. Total anorectal reconstruction requires an anatomical or physical supplement to the performance of a perineal colostomy, which may include an extrinsic muscle interposition (which may or may not be ‘dynamized’), construction of a neorectal reservoir, implantation of an incremental artificial bowel sphincter or creation of a terminal, smooth-muscle neosphincter. The advantages and disadvantages of these techniques and their outcome are presented here.

Keywords: anal incontinence; antegrade continence enema; malone procedure; total anorectal reconstruction

INTRODUCTION

Anal (fecal) incontinence (AI) is characterized by uncontrollable episodes of an involuntary loss of stool at inappropriate times and in socially unacceptable circumstances \cite{1, 2}. Although the incidence varies worldwide, there is a standard reported prevalence of AI in between 4% and 7% of the general population, with a higher estimate (up to 20%) recorded in patients who reside in nursing homes \cite{3, 4}. Evidence would suggest that this symptom seriously impacts on patient-reported standardized quality of life and many aspects of healthy existence where—frequently because of embarrassment—most patients fail to seek specific medical help \cite{5–7}. As a result of these decisions, there is a significant national, annual economic cost of conservative (i.e. nonsurgical) care of these patients \cite{8}, part of which is influenced by the effect AI has on elderly patient institutionalization \cite{9}, as well as the inherent additional costs of anti-diarrheal drugs, healthcare visits, intermittent hospitalizations and patient payment for protective materials and pads. The additive costs of surgical therapies are significant and are impacted by their long-term success rates, the economic impact of procedure-related complications (which are considerable with some of the newer therapies) and the incidence of revisional operative procedures \cite{10}. This article assesses the use and clinical results of antegrade continence enemas, either alone or in combination with total anorectal reconstruction following complete rectal extirpation, as valid surgical alternatives in the management of selected cases of AI.

THE ANTEGRADE CONTINENCE ENEMA OR ‘MALONE’ PROCEDURE

An alternative for the management of various cases of AI that have resisted other more conventional forms of
treatment is the operative technique of antegrade continence enema (ACE), rediscovered by Malone [11]. This approach is more popular in Europe, where it was originally reported in 1905 [12]. The basic technique was adapted from the Mitrofanoff procedure used for a continent catheterizable stoma leading to the bladder [13], but using an appendicostomy for antegrade colonic irrigation. Originally, it was used as a cleansing treatment in spina bifida patients presenting with incontinence. The original operative description used the appendix as a continent, catheterizable, abdominal stoma, which was reversed and placed in a submucosal tunnel of the cecum to form a non-refluxing channel. This was modified to a simpler, non-reversed design with or without creation of a definitive anti-reflux mechanism [14]. The Malone procedure may be carried out on the right iliac fossa, employing a V–Y cutaneoplasty with intermittent catheterization, using a Foley's catheter for creation of a continent, usable conduit under the skin (Figure 1). In the event that there has been a prior appendectomy, or where the appendix is atrophic, the cecal wall can be used as a flap, or a flap may be constructed from the terminal ileum, with the latter being the preferred method overall; this is performed by transecting the ileum about 15 cm from the ileocecal valve and turning the vascularized segment outwards as a buried stoma with neo-ileo-cecal anastomosis (Figure 2) [15–18]. Latterly, part of this procedure may have been laparoscopically assisted [19].

Usually after a two-week waiting period—which allows the system to heal and mature—enemas are then progressively increased in volume up to 1 L, with the final regimen determined by trial and error, as well as by patient tolerance. Results, in both children and adults who are motivated, appear to be acceptable in both the short- and the long terms [20–40]. Long-term quality-of-life data is sparse, where it has been shown that motivation and usage diminishes over time [41]. The results overall appear better in those with neurogenic bowel disability [30, 36]. The principal morbidity of the procedure includes stomal complications, such as stenosis in between one-quarter and one-half

Figure 1. V–Y ACE procedure. The skin flap is sutured to the wall of either the appendix or a fashioned ileal conduit, with formation of a skin tunnel which covers the stoma. (Reprinted with permission from Christensen P, Laurberg S. The Malone procedure and its variants. In Reconstructive Surgery of the Rectum, Anus and Perineum AP Zbar, RD Madoff and SD Wexner Eds. Springer 2013:273–282).

Figure 2. An ileocolic anastomosis is fashioned from the proximal ileum and the ascending colon with production of a small-caliber orifice for the stoma preserving the ileo-cecal valve. (Reprinted with permission from Christensen P, Laurberg S. The Malone procedure and its variants. In Reconstructive Surgery of the Rectum, Anus and Perineum AP Zbar, RD Madoff and SD Wexner Eds. Springer 2013:273–282).
of cases during follow-up, and stomal irritation due to refluxing mucus discharge [42]. The complication rate is high but often relatively minor in nature, with fewer problems if the tapered neo-ileal conduit design is used [43].

Latterly the ACE procedure has been performed as a percutaneous, endoscopic colostomy which was originally used in the treatment of intermittent sigmoid volvulus [44, 45]. The comparative functional results appear excellent, although there is a considerable morbidity which, in a small percentage, can be life-threatening [46, 47]. Norman Williams and his group from the London Hospital have used an alternative here, describing a continent colonic conduit with a full-thickness intussuscepted valve, similar to a Kock continent ileostomy [48], with others describing a retubularized ileal segment for this purpose [49]; still others using a retubularized stomach segment [50]. It would appear that antegrade irrigation provides better results than retrograde irrigation [26, 51, 52], although patients should be warned that some symptoms such as bloating—and nausea if there is coincident constipation—may be essentially unaffected.

A range of fluids may be used for irrigation purposes, including phosphate solution, tap water, saline, phospho-soda, polyethylene glycol, liquorice root solution or arachis oil. Caution is advised in small children and fragile, elderly patients, as well as in those with chronic renal failure [53, 54]. Table 1 shows the reported outcomes of ACE-related procedures in a range of disorders that were combined with primary AI.

### TOTAL ANORECTAL RECONSTRUCTION

Total anorectal reconstruction (TAR) is a method of neorectal reconstruction following complete rectal and sphincter excision. The concept was first proffered in 1930 by Chittenden, who performed a continent perineal colostomy using a flap of the gluteus maximus as a neosphincter [55], with Margottini reporting a large series of this approach in 1950 [56]. The coincident surgical developments of muscle transfer procedures, techniques of dynamization through electrical field stimulation, artificial implants and myogenic sphincter augmentation techniques have been applied to this approach in the development of TAR. The design makes no real attempt to restore those normal functions that are lost, including an adaptable neorectal reservoir, capability of storage and intermittent discharge, a complex closure (sphincteric) mechanism and a discriminatory sensory apparatus, the arms of which are part of normal continence and, as such, full continence cannot be guaranteed for patients undergoing a TAR.

TAR has been made technically feasible in selected cases by the creation of a neorectal reservoir, along with supplementation using autologous muscle or an artificial sphincter. An additional supplement would be the use of an appendicostomy (or an ileal/colonic conduit) for antegrade (ACE) irrigation, as described above, with the result of a ‘pseudo-continent’ status in the patient [57]. Substitution for the rectal functions of storage and sensibility can further be achieved with a segment of descending colon, which has a propulsive function and limited storage capacity, although there is extensive evidence to show that many patients (at least 50%) have a significant ‘low anterior resection syndrome’ after low restorative proctectomy, characterized by an increase in the number of daily bowel motions, nocturnal urgency, stool fragmentation, irregular/incomplete defecation and even frank tenesmus and incontinence [58]. In those who have undergone total rectal excision, the lack of sensory receptors in a peri-anal colostomy results in universal passive incontinence whereas, in those with some area of rectal sensation remaining, data from the creation of various forms of neorectal reservoir (such as the colonic ‘J’ pouch, the side-to-end Baker anastomosis or the coloplasty) suggests that improvements in function over time are due to the reduced action of neorectal motor activity, rather than its role (and capacity) as a true reservoir [59].

In the specific circumstance of TAR, if a pouch is constructed, the shape of the neorectum needs to conform to the anatomical type of reconstruction, where the distal 3–4 cm of the colon will be surrounded by a neosphincter. For this purpose, ‘J’ pouch construction has been combined with a gracilis neoanal sphincter in dogs [60], as well as in humans [61–63]. Geerdts et al. [64] described a pouch placed just proximal to a gracilis wrap, opening the colon

| Author [Ref] | Indication | Number | Success | Complications |
|--------------|-----------|--------|---------|---------------|
| Hill [20]    | Slow transit | 6 | 6 | 50% |
| Christensen [26] | Neurogenic | 8 | 7 | 38% |
| Rongen [27]  | Slow transit | 12 | 8 | 83% |
| Teichman [30] | Neurogenic | 6 | 5 | 67% |
| Lees [31]    | Slow transit | 32 | 15 | 88% |
| Hirst [32]   | Obstructed defecation syndrome | 20 | 13 | 85% |
| Portier [33] | Mixed | 28 | 28 | 50% |
| Lefevre [35] | Mixed | 22 | 18 | 20% |
| Poirier [36] | Mixed | 18 | 14 | 56% |
| Altmare [37] | Mixed | 11 | 8 | – |
| Koivusalo [38] | Mixed | 27 | 24 | 63% |
| Worsoe [40]  | Mixed | 69 | 51 | 38% |

ODS = obstructed defecation syndrome
anti-mesenterically over a length of 15 cm and covering the
defect with an isolated patch segment of distal ileum. As an
alternative, Williams et al. used a triplicated ileal pouch as
15 cm limbs, combined with a stimulated graciloplasty, for
the same purpose [65]. Both of these complex techniques
were not, however, associated with particularly good func-
tion. A simpler approach is to translate a 6–7 cm long colo-
plasty above a colo-anal anastomosis, as advocated by
Fazio and colleagues [66], or by Devesa, who performed a
longitudinal colonic myotomy proximal to a neosphincter,
designed to diminish the peristaltic activity of the descend-
ning colonic segment [67].

The second approach is that of sphincter substitution,
where it is increasingly understood that IAS damage leads
to serious continence disturbance in some cases. The issue
of IAS implantation and augmentation is discussed else-
where in this special edition. In this respect, Torres et al.
originally described a neo-internal anal sphincter [68],
which was wrapped in a spiral configuration around a col-
onic pull-through similar to that described by others [69,
70]. In this technique, 3–4 cm of distal colon is freed from
the pericolic fat and the seromuscular layer is dissected
away from the mucosa, creating a smooth-muscle sleeve
which is then incised in a spiral fashion. The effect is to
construct a pedunculated muscular flap, 1.0–1.5 cm wide
and 5–7 cm in length, which is then wrapped around the
bowel and fixed to its wall. This creates a cone-shaped
smooth muscle cuff attached to the terminal part of the
colon. Results of this technique have been variably reported
[71–75], with Lorenzi et al. modifying this approach by de-
nuding the mucosa and then evertting the last 1.5 cm of the
colonic end, which is then anastomosed to the neo-anus in
the perineal skin [76]. Physiological studies have shown that
these areas distally develop a high pressure zone and a
passage pressure gradient. The role of this added approach
is unclear, where free grafts obviously lack intrinsic and
extrinsic innervation and where they may function more
as a biological peri-anastomotic sling and as a barrier to
evacuation, than as a true functional neosphincter.

A variety of muscles have been used as translation, for
the management of AI, to those patients undergoing TAR,
including the gluteus maximus, the adductor musculature
and the gracilis.

This technique has been supplemented by Farid of Egypt
with fascia lata in very specialized AI patients after recon-
struction of congenital anorectal anomaly [77], although
the use of a gluteoplasty in adult TAR data is limited [78].
Yuri Shelygin’s Moscow group has described success in 82%
of patients treated with an adductor longus reconstruction
TAR in the only report available [79]. Jacob and colleagues
first used a static (adynamic) graciloplasty for the purposes
of TAR for a congenital anomaly [80], with Simonsen et al.
using the technique after rectal cancer excision [81]. The
data here are limited [82–86]; however, the largest series
of dynamic graciloplasties for TAR reported by Cavina et al.
showed an 87% success rate in 98 patients after 55 months of
follow-up, although there was significant morbidity in one-third of cases [83]. The dreaded complication is necrosis
of the neo-anus, which appears to occur particularly in the
TAR cases [87].

Another approach, by Romano et al., is formal sphincter
reinforcement with an artificial anal sphincter with transla-
tion to those specialized patients after abdomino-perineal
excision [88]. The initially good results seen in his eight
cases prompted similar work by Devesa et al. in a small
number of cases, but the high rate of complications and
the need for explants (as in those patients treated primarily
for AI) did not result in extensive use of this technique [67].
The use of an anal sling as a supplement to TAR (a subject
covered elsewhere for the management of AI in this special
edition) has not been reported.

Others have reported the use of an antegrade continen-
tence enema technique for specific use in TAR cases. Chiotasso et al. first reported its use in conjunction with a
perineal colostomy [89], where Farroni and colleagues com-
pared the quality-of-life parameters of those with a perine-
al colostomy and an appendicostomy with those with an
abdominal colostomy, concluding that the perineal colos-
tomy with appendicostomy for was a viable option [90]. As
per the standard ACE procedure, if the appendix is not
available, an ileal neo-appendicostomy, cecal flap or colonic
conduit may be fashioned. The advantage of providing
‘pseudo-continence’ in these patients is the secondary
avoidance of fecal impaction, which can be a very dis-
abling symptom after TAR, particularly where an exter-
nal sphincter recreation or substitution has also been
performed.

Much of the available literature in this specialist group of
patients is difficult to interpret, where congenital anomali-
ies that have been reconstructed are mixed with cases
where radical rectal extirpation for cancer has been carried
out, and where the procedures performed are heteroge-
neous and combined. Apart from comparing quality-of-life
parameters, another way of expressing satisfaction with
the procedure might be the comparison of patients’ quality
of life scores between those with an abdominal stoma and
those in whom there is reconversion to a perineal stoma
[91]. Such an approach requires a revision of the way in
which we assess quality of life in incontinent patients fol-
lowing reconstructive surgery.

Table 2 shows the outcomes of dynamic and adynamic
gracioplasty alone for TAR. In this group there is a high
morbidity and surgical revision rate, with normal conti-
nence reported in only 20% of evaluable patients. At
least one year is required to achieve acceptable continence
in these cases. There does not appear to be any advantage
in ‘dynamizing’ the graciloplasty in some series [81, 84, 92],
suggesting that the functional results of graciloplasty
would be more attributable to the biological ‘cerclage’ effect with the gracilis, rather than to the stimulation itself. If this is true, then most of the perineal stomas treated by explantation of the stimulator would have either undergone re-implantation or been reconverted to an abdominal stoma. Further, severe constipation after graciloplasty has almost always been a feature of stimulated cases [93].

Table 3 shows the outcomes of perineal colostomy with a colonic smooth-muscle wrap and colonic irrigations as part of TAR. As patients do better with colonic irrigation, the value of a neosphincter remains somewhat questionable. Table 4 shows the outcomes if an artificial implanted sphincter device is used in TAR, and Table 5 shows the functional outcome if TAR incorporates an ACE procedure in the management. In this latter group, ileal/cecal/colonic conduit procedures are technically more complex and carry a higher morbidity rate than a simple appendicostomy. Late complications are usually related to stomal stenosis, which can be easily managed by a temporary catheter at night or by a surgical V–Y plasty. Stomal leakage and reflux may be prevented by a cecal imbrication—somewhat akin to a Nissen fundoplication [94]. This approach particularly appears very viable for young patients with AI and prior congenital anorectal anomalies [95]. Overall, the ACE procedure contributes to the avoidance of constipation after TAR when external sphincter reconstruction or substitution has been performed, and where it would appear that in all procedures in which ACE was associated, the good functional results are due to colonic irrigation rather than the other more complex aspects of the technique.

In summary, the role for TAR (and its preferred technique) is currently unclear. In its use, patients and their families need to be informed that continence will effectively never be perfect. The two main candidate groups for this procedure include those with imperfect continence after surgery for a congenital anomaly as children or infants, and those who request reconstruction after radical rectal extirpation for cancer. Patients must understand the morbidity of any proposed procedure and the reported likelihood of subsequent revisional surgery over time.

### Table 2. Dynamic and adynamic graciloplasty as a supplement to total anorectal reconstruction

| Author [Ref] | Number | Dynamic/ adynamic | Complications | Function          |
|--------------|--------|-------------------|---------------|-------------------|
| Santoro [92] | 14     | 0/14              | 1 converted   | 73% pseudocontinuous |
| Mander [61]  | 10     | 10/0              | 80%           | All wore pads      |
| Geerdes [64] | 16     | 16/0              | 4 reconverted | 30% continent      |
| Cavina [83]  | 98     | 98/0              | 37%           | 87% continent      |
| Rullier [84] | 15     | 0/15              | 73%           | 78% continent      |
| Ho [86]      | 17     | 17/0              | 40%           | 45% continent      |
| Simonsen [81]| 24     | 0/24              | 65%           | 77% continent      |
| Violi [85]   | 23     | 15/8              | 37%           | 75% continent      |

### Table 3. Data pertaining to smooth muscle neosphincters combined with colonic irrigation for total anorectal reconstruction

| Author [Ref] | Number | Complications | Functional status |
|--------------|--------|---------------|-------------------|
| Lasser [73]  | 40     | 55%           | 11% continent     |
| Gamagami [74]| 63     | 65%           | 39% satisfactory  |
| Portier [75] | 18     | 33%           | No reconversions  |
| Pocard [96]  | 12     | Not stated    | 92% pad use       |
| Hirche [97]  | 44     | 40%           | 50% continent     |

### Table 4. Artificial bowel sphincter use in total anorectal reconstruction

| Author [Ref] | Number | Complications | Functional result |
|--------------|--------|---------------|-------------------|
| Romano [88]  | 8      | –             | 87% continent     |
| Lirici [98]  | 3      | All explanted | All continent     |
| Devesa [67]  | 1      | Explanted     | Improved          |
| Ocares [99]  | 1      | Explanted     | Not evaluable     |
In cancer, after exclusion of recurrent disease, other factors such as obesity, intra-abdominal adhesions, comorbidity, prior perineal irradiation and even age may be precluding conditions for TAR. The issue of immediate TAR vs delayed TAR is also controversial and somewhat akin to the argument of immediate vs delayed breast reconstruction after mastectomy. It would seem feasible to perform a perineal colostomy and an appendicostomy for ACE at the initial rectal excision in motivated cases, and this may be associated with quite minimal perineal morbidity in early selected cases (those with T1-2N0 tumors) when compared with the known perineal morbidity of primary perineal closure.

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