

# Laparoscopically inserted button colostomy as a venting stoma and access port for the administration of antegrade enemas in African degenerative leiomyopathy

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## Summary

African degenerative leiomyopathy (ADL) is a rare incurable disorder seen in African children, predominantly in southern and south-eastern Africa. ADL presents as chronic intestinal pseudo-obstruction. Management is traditionally conservative, with surgery restricted to the management of complications. We have placed Malone antegrade continence enema (MACE) stomas in the grossly dilated colon to vent accumulated gas and administer antegrade bowel enemas. This is done mainly for relief of gaseous distension and constipation in an attempt to provide symptomatic relief and improve quality of life. In this article, we present our preliminary results of laparoscopically assisted technique to insert a Mic-Key gastrostomy device as a 'button colostomy' in 8 patients over the past 6½ years.

African degenerative leiomyopathy (ADL) is a rare disorder of unknown aetiology seen in African children, predominantly in southern and south-eastern Africa. The pathology involves degeneration of smooth muscle with replacement by fibrous tissue. Predominantly affecting the rectum and colon, ADL presents as chronic intestinal pseudo-obstruction, and anorexia and malnutrition are hallmarks of the disease. The disease is universally fatal in the second or third decade of life. Management is traditionally conservative, with surgery restricted to the management of complications. Over the past 15 years we have placed Malone antegrade continence enema (MACE) stomas in the grossly dilated colon to vent accumulated gas and administer antegrade bowel enemas. This is done mainly for relief of gaseous distension and constipation in an attempt to provide symptomatic relief and improve quality of life. Our operative strategy evolved from an open modified MACE procedure to currently using a laparoscopically assisted technique to insert a Mic-Key gastrostomy device as a 'button

colostomy'. We present our preliminary results of this procedure in 8 patients over the past 6½ years with follow-up of 6 months to 6 years.

## Materials and methods

Eight patients with histologically confirmed ADL, who failed to improve and remained symptomatic despite optimal medical management, were assessed for placement of 'button colostomy'. After appropriate work-up, informed consent was obtained.

The proposed insertion site was marked. A 5 or 10 mm port was placed through the umbilicus using an open technique, and pneumoperitoneum established. A 30° telescope was inserted and the abdominal cavity inspected. Two additional 3.5 mm ports were inserted into each iliac fossa (Fig. 1) and a suitable loop of dilated colon, preferably transverse or sigmoid, was selected. A small transverse incision was made on the anterior abdominal wall at the proposed insertion site and the selected loop of colon brought out via the incision under vision. The colon was opened on the antimesenteric border and an 18F Mic-Key device (Fig. 2) was inserted, after which the colon was returned to the abdominal cavity and secured to the anterior abdominal wall with externally inserted sutures (Fig. 3). The balloon device was then inflated and the sutures tied prior to completing the procedure. Sutures were removed after 5 days. The caregivers and patients were taught by the nursing staff to insert an 8F feeding tube through the Mic-Key device for regular (usually 3 times a day) deflation and antegrade enemas (once or twice a day). The patients were discharged as soon as they were capable of using the button colostomy. Prokinetic agents and laxatives were continued as baseline medical treatment after discharge. All patients were followed up regularly. Assessment included presence of recurrent symptoms, problems related to the use of the button colostomy, and any complications. In addition the dietitians subjectively assessed improvement in the patients' symptoms and objectively measured their nutritional

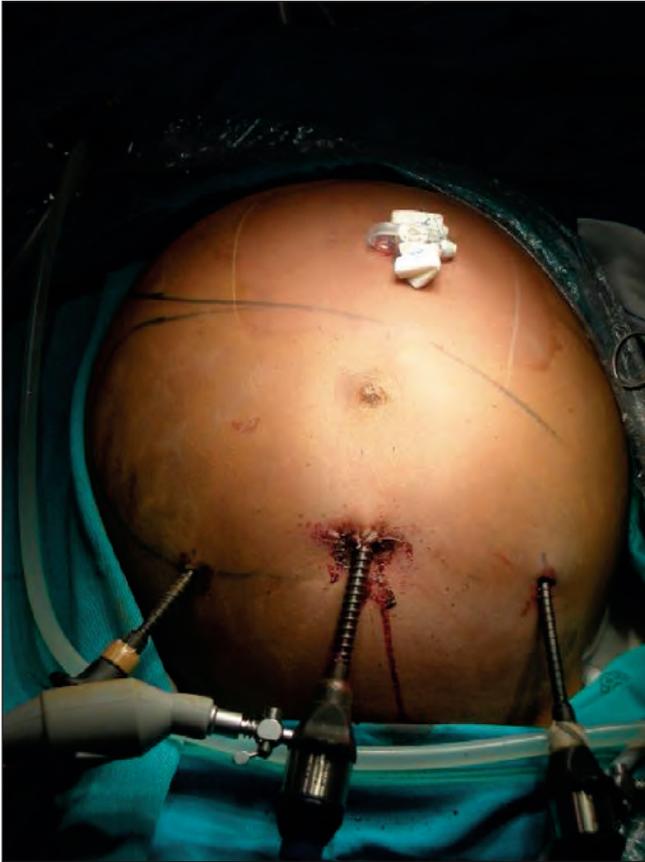


Fig. 1. Photograph showing port positions and the Mic-Key device in place.

parameters as part of an ongoing prospective study. There was no mortality. Two patients had minor skin excoriation due to leakage of stool around the button device that was managed conservatively. The button device needed to be changed after a minimum period of 6 months and a maximum period of 2 years in each of the 8 patients. All the caregivers and the patients were comfortable in the daily use of the button colostomy.

## Results

The results of the use of laparoscopically assisted button colostomy in ADL are set out in Table I.



Fig. 2. The Mic-Key skin-level gastrostomy device.

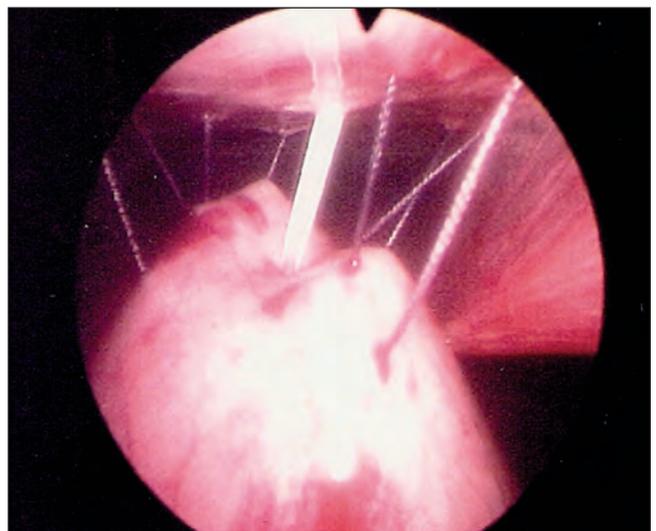


Fig. 3. Photograph showing the Mic-Key device in place and being secured by 3:0 Vicryl sutures under laparoscopic vision.

## Discussion

ADL is seen only in young African children from southern and south-eastern Africa and was originally known as 'Bantu pseudo-Hirschsprung's disease'.<sup>1,2</sup> It is a rare incurable disorder

**TABLE I. THE RESULTS OF THE USE OF LAPAROSCOPICALLY ASSISTED BUTTON COLOSTOMY IN AFRICAN DEGENERATIVE LEIOMYOPATHY**

<b>Total patients</b>	<b>8</b>
<b>Males</b>	<b>4</b>
<b>Females</b>	<b>4</b>
<b>Age</b>	<b>7 - 14 years</b>
<b>Follow-up</b>	<b>6 months - 6 years</b>
<b>Procedure-related mortality</b>	<b>None</b>
<b>Morbidity: skin excoriation due to peristomal leak</b>	<b>2</b>
<b>Time for replacement</b>	<b>6 months - 2 years</b>
<b>Subjective improvement in symptoms</b>	<b>All 8 patients</b>
<b>Objective nutritional improvement</b>	<b>All 8 patients</b>

of unknown aetiology and is universally fatal in the second or third decade of life. Pathologically it is an acquired visceral myopathy that involves degeneration of the smooth muscles and their replacement by fibrous tissue. The rectum and colon are predominantly affected, but it is a progressive disorder and tends to affect the rest of the

gastro-intestinal tract and other organs and structures that have smooth muscles, including the genito-urinary system, extra-hepatic biliary system and systemic vasculature.

ADL commonly presents in the second or third year of life with features of chronic intestinal pseudo-obstruction such as chronic constipation, inability to evacuate flatus, and progressive abdominal distension. Persistent anorexia and ongoing nutritional depletion are significant problems. The use of laxatives, prokinetic agents and regular bowel wash-outs are the mainstay of treatment. Surgical management of the grossly dilated bowel by either diversion or resection is not curative and does not provide sustained symptomatic relief. The role of surgery has therefore traditionally been restricted to the treatment of complications such as volvulus. We have previously described the successful application of the MACE principle, in the form of a tubularised colostomy, for symptomatic relief of abdominal distension and chronic constipation in patients with ADL.<sup>3</sup> In an ongoing prospective study we have observed objective evidence of nutritional improvement and subjective improvement in their symptoms.

Our attempt to provide symptomatic relief to patients with ADL using simple surgical procedures has evolved over the past 15½ years. After 2 of the 14 patients who had a tubularised colostomy developed skin-level strictures due to non-use, we decided to use the Mic-Key gastrostomy device in the colon to prevent the problem of stomal stricture. Its application for

antegrade enemas has been described previously.<sup>4</sup> Initially we inserted the button device using an open technique. In the past 6½ years we have changed to a laparoscopically assisted technique with minimum morbidity. All patients have been closely followed up with special attention to the ease of use of the device and any complications. In an ongoing prospective study these patients are independently assessed by the staff of the dietetics department, objectively with regard to their nutritional status and subjectively for symptom recurrence.

## Conclusions

In this short series of 8 patients with ADL, we found that the simple technique of laparoscopically placing a button colostomy, using a Mic-Key skin-level gastrostomy device, is useful in providing symptomatic relief and improving quality of life. We are aware of the limitations of this report, namely small patient numbers, short follow-up, and lack of objective assessment of quality of life scores. Despite this, acceptance by the patients and their parents, substantial symptomatic relief, nutritional improvement and significant improvement in quality of life have encouraged us to continue using this procedure in patients with ADL. The issue of nutritional improvement is being studied and the results will be published in due course.

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