INTRODUCTION

A primary concern of any surgeon is the patient’s recovery from a particular surgical procedure. In recent decades, physicians have taken great care to reduce the impact of major surgery by improving their techniques, but equally important to the effectiveness of treatment is the environment in which the patient recovers. Even the best surgeons cannot cure an ailment if the patient is allowed to come into contact with unsanitary or dangerous conditions during the healing process. The course of the recovery process for an individual is thus a function of two variables: the physician’s expertise and post-operative care.

Compared to more affluent countries, the Third World falls unquestionably behind in terms of the care it can offer its patients, and naturally we expect that patients will exhibit a discrepancy in recovery based on where they receive care. To determine the extent to which either the operative or post-operative care environments are responsible for this discrepancy, we must standardize one of these variables and study the other. Specifically, if the quality level of the operative medical care in Haiti and the United States could be made equal, then any discrepancy between the nations’ recovery patterns can only be attributed to differences in the post-operative environments of both countries.

L’Hôpital Albert Schweitzer is an American-style hospital established in 1956 in the Artibonite district of Haiti that provides such conditions. The H.A.S. medical staff is comprised not only of Haitian physicians, but is supplemented by physicians from the United States, Canada, and Europe. Visiting physicians volunteer from two weeks to several years, living on the hospital campus and practicing modern techniques in primary care and surgical situations. One of the smaller departments is pediatric surgery, where the most sophisticated surgical cases must wait until Dr. Michael Curci, a pediatric surgeon from Portland, Maine, makes his annual two-week visit every spring. During these visits, Dr. Curci performs advanced surgical procedures on dozens of pediatric patients, thus providing a level of care that is nearly equal to the care he provides patients in the United States.

One of the newborn anomalies that Dr. Curci frequently treats in Haiti is a birth defect called Hirschsprung’s disease, in which the nerve tissue of part of the colon is absent. Dr. Curci’s preferred surgical remedy is to remove the affected segment of the colon in a procedure known as the Swenson pullthrough.

The purpose of this research was to determine the patterns of recovery and patterns of intestinal function in Hirschsprung’s patients who have undergone the Swenson pullthrough procedure and to compare the results with data published on Hirschsprung’s recovery in the West. The findings from this research will ideally enable physicians at H.A.S. to decide whether the care they are providing at the hospital is truly effective in this subset of patients. That is, are the efforts of the hospital staff actually working to permanently restore bowel motility to a Hirschsprung’s patient?

Hirschsprung’s Disease: Pathophysiology, Diagnosis, and Treatment

Hirschsprung’s disease involves absence of ganglion nerve cells in the smooth muscle tissue of a portion of the colon and, consequently, the
inability to stimulate coordinated smooth muscle contractions and efficiently pass stool. The condition usually manifests itself immediately after birth, but in some cases clinical onset may be delayed for up to two years. Surgeons advise that when the passage of the first bowel movement is delayed beyond 48 hours in an otherwise healthy infant, Hirschsprung’s should be suspected (Greenfield, 1997). Abdominal distention is often present after several days, accompanied by diarrhoea and/or constipation. If not treated, Corman (1998) reports that mortality may be as high as 30% in the United States, mostly due to enterocolitis. It is highly probable that in Haiti, where nearly all children are born in the home, the majority of Hirschsprung’s cases may go unnoticed or untreated, and mortality may be even higher.

The patient will often be given a saline-barium enema, which allows the physician to study the colon on X-ray (see Figure). Such X-rays typically indicate a tapering “transition zone” between a distal colon of normal diameter and the widened segment of colon above it. The transition zone is the boundary between ganglionic and aganglionic intestinal tissue; it is the beginning of the segment affected by Hirschsprung’s disease. If Hirschsprung’s is suspected, a rectal biopsy is taken for confirmation. Smooth muscle tissue from a healthy colon contains ganglion nerve cells visible under a microscope. In Hirschsprung’s disease, these cells are absent.

At l’Hôpital Albert Schweitzer, most of this diagnostic protocol is done by staff physicians and general surgeons. Depending on the severity of the patient’s symptoms, the patient is typically managed by medical measures such as diet modification, or surgical measures such as a temporizing colostomy. Once stabilized, the patients are sent home and given appointments to come back to the hospital in the spring. When he arrives each April, Dr. Curci evaluates and selects candidates for the Swenson pullthrough.

The definitive operation involves a large abdominal incision and major dissection of the tissues that suspend the lower intestine. The affected portion of the colon is identified by visual criteria. Using a second set of surgical instruments, the assisting surgeon manipulates the colon and inverts the aganglionic segment outside the anus. When the healthy colon and the sphincters are aligned, the surgeon anastomoses the colon and removes the unhealthy portion externally through the anus. Thus, the contact between the contaminated interior and clean exterior of the colon is minimized. The surgery is curative; once the affected colon tissue is removed, Hirschsprung’s disease will not recur.

Recovery rates from this procedure are high in the United States; only ten to twenty percent of all cases present long-term complications. Dr. Curci performs the Swenson pullthrough for most of his patients with Hirschsprung’s disease, both in Maine and in Haiti, under nearly identical circumstances. Therefore, differences in recovery are controlled up to the point when the patient leaves the surgery. Beyond the operating room, the differences must be due to the recovery environment.

METHODOLOGY

The names of eighteen patients who had undergone the Swenson procedure for Hirschsprung’s disease at H.A.S. were obtained. Over a period of eight weeks, as many of these patients or their families as possible were located and interviewed. The primary goal was to complete a list of survey questions for as many patients as possible to develop an accurate assessment of their recovery status. In a country with 65% illit-
eracy and almost no organized postal system, the surveys had to be completed orally and in person.

The first and most difficult objective was to locate the patients. The H.A.S. Medical Records Department was able to provide limited location data for sixteen patients on the list. Dossiers and location information for two patients were missing completely.

The second objective was to interview each patient regarding his or her recovery from surgery. The survey, consisting of twenty questions, was designed to help characterize the patient's bowel patterns and post-operative recovery. One group of questions assessed the recovery directly following the operation; these questions were designed to develop a general picture of recovery over the course of time. Patients were asked how much time it took following the surgery for the patient to resume normal physical activity and how much time passed before the child could eat normal foods. Patients were also asked about any infection in the surgical wound shortly after discharge from the hospital, as this tends to present complications in Haiti. The responses to these questions assessed the degree of post-operative morbidity: the more quickly the child assumed normal eating and activity patterns, the less severe the post-operative effects.

A second group of questions assessed the patient's present condition. These were the questions that most interested Dr. Curci, since they assessed the long-term success of the operation. Patients were asked about stool frequency, stool diameter and consistency, occurrence of infection, and distention of the abdomen. A characteristically normal bowel pattern as determined by these questions indicates restored bowel motility. Abnormal responses to these questions may be indicative of complications associated with either the disease or the surgery. Patients also commented on any abdominal pains they had experienced following surgery. Though it would not be possible to determine if such pains were associated with Hirschsprung's, chronic pain is sometimes an indication of enterocolitis, a dangerous complication sometimes associated with the disease and the curative Swenson procedure.

A third line of questions tried to determine some of the more sociological ramifications of surgery; in particular the amount of time the child has missed from school, chores, and recreation with friends as a result of the operation.

In developing the questionnaire a “red herring” question was included which was designed to detect “yes-man syndrome.” Several veteran clinical researchers advised that if patients think they will receive more medical care for providing embellished or exaggerated information, they will do so. Each patient was asked about the presence of blood in the their stool, a symptom that is virtually never observed in Hirschsprung's patients more than a few months post-operatively. Whether the patient claimed to have blood in his stool allowed the patient's credibility to be determined with regard to the remainder of the survey.

RESULTS

By the end of eight weeks eleven of the sixteen patients whose files were available had been located. The remaining five would likely not have been found even with more time; the information on their hospital dossiers was either absent or so vague that locating them was virtually impossible. Given the limitations of the infrastructure in rural Haiti, the project was successful in gathering data from nearly 65 % of Dr. Curci's former Hirschsprung's patients.

The primary objective of this project was to determine whether Dr. Curci's patients were recovering bowel motility to the same extent that they would in the United States following surgical treatment for Hirschsprung's disease. General information about the patients as a group will first be presented, followed by case studies of five patients who typified the various outcomes of the Swenson Pullthrough procedure.

All of the patients had lived in Haiti all their lives, and the majority ranged in age from seven to fourteen years. One patient was four; and one was seventeen. Among the surveys completed there were ten males and one female; Hirschsprung's affects males more often than females and according to some studies (Corman, 1998; Greenfield, 1997) may be a sex-linked trait. Most patients were diagnosed near the time of birth. The definitive surgical procedure for Hirschsprung's, most often the Swenson Pullthrough but with one exception, was performed on all patients by Dr. Curci. The median age for undergoing the procedure was four; and ranged from twelve months to twelve years. The majority of patients were at least four years post-operative, and two patients were in their second post-op year or earlier.

Following the interviews, the surveys were organized according to relative recovery patterns. Five patients recovered complete bowel motility and expressed no post-operative complaints about
the surgery. They exhibited normal healthy stool patterns (defined as one to two per day) and did not complain of any swelling of the abdomen. This group included four males and one female, between the ages of seven and eleven.

Four patients showed moderate irregularity in their bowel function, and complained of having periods of extreme discomfort and constipation accompanied by frequent diarrhea, five to six times daily in each case. Two of these cases, it should be noted, had a history of kwashiorkor (malnutrition). It may be that in the absence of a good diet these two malnourished patients were particularly prone to constipation, which in the case of a post-Hirschsprung's individual can be dangerous. This group averaged twelve years of age, with one male aged seventeen at the time of the interview.

Two of the Hirschsprung's patients died before my arrival to H.A.S. in September of 1999. Their deaths occurred under unique circumstances, yet both are indicative of critical deficiencies in the Haitian health care system which have a direct impact on recovery from surgical care.

**CASE STUDIES**

Each of the case studies below was chosen as an example of one type of recovery pattern. They contain information compiled both from the interview and the patient's H.A.S. medical dossier. Case I is a classic successful Hirschsprung's case and exhibits normal bowel motility. Case II is illustrative of the group which showed evidence of chronic bowel problems. Case III is a more extreme example from that same group, whose economic and geographic situation made him something of a special case. Finally, Cases IV and V are the two recorded deaths in the survey group, each of which is specific enough to merit separate attention.

**Case I: Full recovery of bowel motility.**

LJ was born in 1988 and lives with his family in the town of Dessalines, which is outside the district normally serviced by the hospital. Hirschsprung's was not diagnosed until his second birthday, whenupon a temporizing sigmoid colostomy was performed. The pullthrough operation took place in 1992, after which the patient was hospitalized for three weeks.

LJ's recovery was fairly uneventful. The hospital staff made sure LJ was able to stand and walk before discharging him, so within only a few days of returning home his normal activity was allegedly restored. After two weeks of liquids, he resumed a normal Haitian diet of rice and beans. Of his present health he claims he has had no pain, illness, diarrhea, constipation, or abdominal distention. His stool frequency is once per day and is of normal diameter.

LJ's father says he started school one year later than normal. However, other than the scar on his belly, none of his friends or peers notices anything different about him compared to anyone else. He is active in neighborhood soccer and says there is nothing that his condition prevents him from doing.

**Case II: Persistent bowel motility problems.**

GA was born in 1986 and lives with his mother in Petite Riviere, the largest town in the hospital district. He received a colostomy for Hirschsprung's disease before his third birthday, and Dr. Curci performed the pullthrough procedure when GA was four. He was hospitalized three weeks. Two months post-operatively, he was admitted for infection of the surgical wound and an obstruction in the small bowel which his dossier did not characterize further. In 1997, GA was admitted for malnutrition and malaria.

After GA was discharged following his pullthrough operation, his mother reported that he stayed in bed for about one week. It was several weeks before he began to walk and act normally, though he still does not exert himself.

GA complains of frequent loose stools, sometimes six or more per day. He claims his diarrhea comes in irregular episodes; usually every week to two weeks the loose stools recur and last a period of several days. He says his abdomen is often swollen and painful during these episodes. Abdominal pain was described as sharp at times, but the location of the pain was not specific. GA reports no blood in his stool. Because of his physical status his family does not allow him to lift heavy objects or perform strenuous chores such as carrying water. When his family needs to travel, they must pay for a taxi rather than walk so as to minimize GA's activity.

The recovery from surgery required GA to start school two years late. He says at first his school-teachers would become frustrated with his frequent need to be excused to use the bathroom, and would forbid him to leave. He added that after his mother explained GA's condition to his teachers, they excused him from class as necessary. He says he enjoys playing soccer although he is not able to play as aggressively as his peers.
Case III: Special case.

Because of the unusual isolation in which DL lives relative to the other ten patients, this particular patient's case is somewhat atypical and nonrepresentative of a normal recovery pattern in Haiti. However, the conditions in which he was forced to recover from surgery are worth noting as a special case scenario and as a testimony to the disorganization of health care in Haiti.

DL was the only patient who had serious bowel motility problems. This boy lived in one of the most remote parts of Haiti, on a large isolated island ten miles offshore. The entire island, except for one main town closest to the mainland, lacked electricity and drinking water. Rice and beans, the staple diet elsewhere in Haiti, could not grow there because of the mountainous terrain.

DL was born in 1982 and was immediately diagnosed with Hirschsprung's disease at the Wesleyan Mission Hospital near his locality, about 150 miles from l'Hôpital Albert Schweitzer. He was referred to H.A.S. for treatment, but in Dr. Curci's absence underwent a temporizing colostomy and was given a rendezvous appointment for the following spring. However, the family's extreme poverty made it unable for them to afford the costs of surgery, which amounted to about $U.S. 100. For the first twelve years of his life, DL lived at home with an open colostomy, rarely staying in contact with a doctor. In 1994 at the time of the pullthrough surgery he weighed 67 pounds. Since then he claims he has had episodic abdominal pains on an almost weekly basis. His stool frequency varies from once daily to five to ten times daily, and more often than not his stools are loose and low-volume. He says he must defecate in the corn field next to his house, since there are no toilets in his locality. Epigastric abdominal pains accompany his episodes of diarrhea. His family does not allow DL to exert himself; when I asked whether he plays sports with his friends, he was the only one of my sixteen patients who admitted that he could not engage in any exercise because of his condition.

Case IV: First fatality.

EM was born in June 1996 and lived in the town of Verrettes about three miles from the hospital. At twenty months, he was admitted for malnutrition. One month later he was diagnosed with Hirschsprung's disease and the following spring (1999) underwent the Swenson pullthrough. The physical examination prior to and following surgery noted repeatedly that EM was small for a three-year-old.

Two months postoperatively EM presented with chronic constipation, for which he was readmitted to the hospital on two occasions. Over the course of the summer, he was seen for fever and constipation three more times. Doctors' notes in the dossier indicate severe dehydration. According to his mother, during his final stay at the hospital he was put on a carrot juice diet, but each day consumed less and less until he died August 9.

EM's mother agreed to answer some questions about her son. She indicated that he had experienced abdominal pains and that his bowel pattern included periods of constipation and diarrhea. EM was able to walk only with considerable effort and never resumed full normal walking activity.

Case V: Second fatality.

CL was born in 1994 and lived in Petite Riviere. He presented with Hirschsprung's less than a week after birth and was given a right transverse colostomy. In April of 1995, Dr. Curci performed a pullthrough, the colostomy was closed three months later, and CL was sent home. Following the closure of the colostomy, his mother reported that his stools were frequent and loose.

In the HAS system, patients are initially seen at one of ten outlying dispensaries. Based on that evaluation (often done by a nurse), a patient is either treated directly or referred in writing to the central hospital. The goal is to refer patients to the hospital who require a doctor's care, and to limit the number of unnecessary referrals and thus improve efficiency. Unfortunately, as was the case for CL, the system is not perfect. The patient's mother contests that her son became constipated and sick very frequently throughout subsequent years, and it was difficult to convince the supervisor of her local dispensary to refer CL to the hospital. The dispensary supervisor did not agree that CL's symptoms required a hospital clinic visit.

CL's last admission to H.A.S. was in June of 1996 for enterocolitis, for which he stayed eight days. In the year that followed, his mother would take him to the dispensary every two to three weeks when he exhibited recurring signs of abdominal pain, inflammation, diarrhea, and constipation. Each time, the dispensary would not sign a referral to H.A.S. Finally in September of 1997, the dispensary staff endorsed a referral, and CL's mother began the eight-mile walk to H.A.S. with her son. She alleges that on the way to the hospital, CL began vomiting food and water and became unconscious, finally dying of dehydration on the roadside only a few miles from the hospital.
DISCUSSION

The five case studies presented here clearly represent a wide variance in recovery patterns from surgical treatment of Hirschsprung’s in Haiti. Among the eleven patients surveyed, the incidence of full recovery is remarkably low relative to United States standards. The current standards of comparison for the United States and Europe, published in Corman 1998, estimate that roughly eighty percent of Hirschsprung’s patients in the United States have no post-operative complications and have normal bowel patterns. This study finds that complete bowel motility was restored in five of the eleven patients, as typified by Case Study I and four patients showed evidence of significant motility problems postoperatively, as in Case Studies II and III.

Although it is unfair to compare statistics given the small sample size, in the U.S. we would expect far fewer of these patients to present serious complications. Indeed, many of the patients interviewed for this research exhibited symptoms that could be easily alleviated with proper medication, diet, or surgical revision. Judging from their present conditions, the four patients with motility problems would most likely have recovered full motility had they been allowed to recover with better post-operative follow-up and care. Because the patients’ post-operative health problems cannot be effectively managed in Haiti, many Hirschsprung’s patients suffer higher morbidity than they would in the United States.

Although it is impossible to say with certainty, the evidence suggests that the two patients who died would most likely have survived under the U.S health care system. In Case Study IV, the patient’s condition was aggravated by recurrent malnutrition, which significantly diminished the patient’s chances for recovery. In a patient such as this who presents with multiple health problems, chances for survival are low regardless of the complications from Hirschsprung’s, thus Hirschsprung’s should not be considered the singular cause of death. Malnutrition in the U.S. is virtually non-existent relative to Haiti, so such a rare case of Hirschsprung’s would most likely not present such problems in the developed world. In Case Study V, it is possible that the patient died because of inappropriate triage and delays in transfer. The organization of the health care system in Haiti is in a period of slow transition. Improved health care requires better roads, infrastructure, education, and community development. While the Haitian political system is only marginally stable, Haitian health organizations such as H.A.S., which have financial bases in the developed world, have begun to take the initiative in creating an efficient, egalitarian plan to provide health services in Haiti. But even this system, as was observed in Case Study V, leaves considerable room for improvement.

Many physicians are beginning to show involvement in international medicine in the developing world, if not by volunteering their skills, then by donating money or equipment to hospitals. But providing American-style technology is not necessarily the right solution to Haiti’s health problems. Instead, more attention needs to be given to improving post-operative recovery care and other facets of the larger health care system that technology cannot improve directly. Haiti cannot use, for example, a heart-lung machine to perform open-heart surgery unless it has the ability to care for the patient post-operatively to the same level of attention that hospitals provide in the United States. American medical companies continue to donate what they assume Haiti needs, when in fact, a Haitian hospital cannot effectively put to use much of the technology it receives (D’Amico, 1998). The challenge when practicing medicine in Haiti is to provide First World quality care in a Third World environment. Research similar to this Hirschsprung’s project is invaluable in this regard because it helps demonstrate what health care in underdeveloped nations needs and does not need from volunteers and benefactors in the developed world.

On similar lines, there is a question as to whether it is appropriate to perform sophisticated pediatric abdominal surgery, such as the Swenson pullthrough, in a society that does not yet have the resources to follow up such cases with the necessary care. Treating Hirschsprung’s disease with surgery requires a substantial investment of time, skill, and resources even in the United States. The ideal surgical remedy for Hirschsprung’s disease in Haiti would be a single “magic” procedure that would eliminate the problem and heal quickly, so as to allow the child to resume normal bowel motility as soon as possible with minimal dependence on follow-up care. For the Hirschsprung’s patients in this study who presented with postoperative complications, development of such a procedure would unquestionably improve the odds of restoring bowel motility. This study demonstrated that it is the unavailability of follow-up care that
Table. Summary of patients’ recovery patterns from Hirschsprung’s disease. In terms of recovery pattern “A” indicates no problems, “B” indicates moderate to severe complications, and “C” indicates deceased. Ages of the deceased cases are given as age at time of death.

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age / Sex</th>
<th>Years since Swenson procedure</th>
<th>In / Out District</th>
<th>Recovery Pattern</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>LC</td>
<td>11yo / M</td>
<td>6</td>
<td>In</td>
<td>A</td>
<td>Occasionally uses laxatives to maintain a regular bowel pattern of one stool per day.</td>
</tr>
<tr>
<td>MF</td>
<td>5yo / F</td>
<td>3</td>
<td>Out</td>
<td>A</td>
<td>No complications, 2-3 stools per day.</td>
</tr>
<tr>
<td>PB</td>
<td>11yo / M</td>
<td>2</td>
<td>In</td>
<td>A</td>
<td>Required more hospitalization than most other patients, but did not complain of complications.</td>
</tr>
<tr>
<td>IC</td>
<td>7yo / M</td>
<td>5</td>
<td>In</td>
<td>A</td>
<td>No complications, 1 stool per day.</td>
</tr>
<tr>
<td>LJ</td>
<td>11yo / M</td>
<td>7</td>
<td>Out</td>
<td>A</td>
<td>No complications, 1 stool per day. (See Case I)</td>
</tr>
<tr>
<td>FE</td>
<td>13yo / M</td>
<td>9</td>
<td>In</td>
<td>B</td>
<td>4-5 stools daily. Received saline enema following interview to relieve constipation.</td>
</tr>
<tr>
<td>KC</td>
<td>13yo / M</td>
<td>7</td>
<td>Out</td>
<td>B</td>
<td>5-6 stools daily. Often has painful abdominal swelling. Reported blood in stool (reports unconfirmed).</td>
</tr>
<tr>
<td>GA</td>
<td>13yo / M</td>
<td>9</td>
<td>In</td>
<td>B</td>
<td>6+ stools daily. Recurrent painful abdominal swelling, about every two weeks. (See Case II)</td>
</tr>
<tr>
<td>DL</td>
<td>17yo / M</td>
<td>5</td>
<td>Out</td>
<td>B</td>
<td>5+ stools daily. Says he is “usually” sick. Recurrent pain and abdominal swelling. Very low physical activity. (See Case III)</td>
</tr>
<tr>
<td>EM</td>
<td>3yo / M</td>
<td>0.5</td>
<td>In</td>
<td>C</td>
<td>Hirschsprung’s symptoms were complicated by acute malnutrition. (See Case IV)</td>
</tr>
<tr>
<td>CL</td>
<td>3yo / M</td>
<td>2.5</td>
<td>Out</td>
<td>C</td>
<td>Frequent loose stools. Unable to obtain follow-up care at H.A.S. (See Case V)</td>
</tr>
</tbody>
</table>

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REFERENCES


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