

**JOURNAL CLUB SUMMARY**

**11/26/03 – HIRSCHSPRUNGS DISEASE**

**(attendees: Breuer, Moss, Touloukian, Seashore, Tashjian, Chen, Yoo, Jose, Henry, Goyal, Silverman)**

- I. Sherman JO, Snyder ME, Weitzman JJ, Jona JZ, Gillis DA, O'Donnell B, Carcassonne M, and Swenson O. A 40-year multinational retrospective study of 880 Swenson procedures. J Ped Surg 24(8):833-838, 1989.

Presented by Marion Henry, MD

Reported	Not App.	REPORTING DETAIL – METHODS SECTION	Comments
yes		A clear description of study design	A retrospective multi-institution case series without comparison group with a goal of long-term follow-up
yes		The number and practice type of institutions where cases were performed	Seven centers in North America and Europe with pediatric surgeons
No		Number of surgeons who actually operated in study	Not specified
	X	A statement as to whether the same surgeon operated on pts from different treatment groups or just one group	No comparison group
yes		The precise timeline during which patients were treated	1947-1987
No		A clear description of how patients were selected into study	NO. Pts who had Swenson procedure but unclear how many other pts received other operations, how these pts were chosen for Swenson procedure
no		The number of eligible patients at the study sites excluded during the study	
no		A clear description of the study population	
no		A clear description of the relevant diagnostic criteria used to identify cases	
no		A clear description of critical aspects of operative technique and peri-operative care	Only says "Swenson procedure", no other details of operative technique or perioperative care
	X	Statement as to whether any attempts were made to standardize operative technique or peri-operative care	Retrospective review
		<b>RESULTS</b>	
yes		Range and mean of all relevant demographic and baseline variables	See summary
yes		Range and median for length of follow-up reporting	"
yes		Relevant outcome variables are presented with appropriate measures of range and variability	
yes		Methods for measuring outcomes of interest are clearly described	
no		Statement is made whether any data is missing (and how missing data is addressed)	
yes		Number and appropriate details regarding all complications	

Summary: A retrospective multi-center case series covering seven centers from 1947 to 1987. This is the largest study with the longest follow up period for patients treated for Hirschsprungs disease. The charts of 880 patients who underwent a Swenson procedure were reviewed by individual surgeons at each center.. There was no comparison group for this study.

Result summary:

Demographics:

Race:	88% white	7% black	4% other
Sex:	81% boys	19% girls	
Birthwt.	900-5955 gms		
Comorbidities:	4% Downs	2.5% mentally retarded	
Age at OR	4 d to 50 yrs (mean 3.7 yrs)		

- Table 1: Length of aganglionic bowel (75% at or below sigmoid colon)  
 22% blacks had rectal aganglionosis vs 20.5% others and 8.2% whites  
 None of the nonwhite, Downs, mental retardation or low birth weight infants had total colon aganglionosis
- Table 2: Accuracy of barium enema diagnosis  
 Length of aganglionic section influenced ability to correctly diagnose Hirschsprungs radiographically
- Table 3: Patient age at time of resection  
 57% were between 5 mo and 1 yr. (62% under one year)
- Table 4: Summary of patients without complications (47.5% had NO complications)
- Table 5: Early and late postoperative complications  
 Early: 6.5% wound infection, 1.7% dehiscence, 5.6% anastomotic leak, 2.4% sepsis  
 Late: 7.6% stricture
- Table 6: Postoperative mortality – total mortality 2.4% (21/880)
- Table 7: Enterocolitis  
 Postoperative enterocolitis – 11.5%  
 Late enterocolitis – 22.5%

Discussion: The strengths of this paper lie in the fact that it is a large series with a very long term follow-up of its patients. Given this fact, it is a significant series in pediatric surgery, which reveals a lot of information about the outcomes of patients receiving the Swenson procedure for Hirschsprungs disease. Consideration must be paid, however, to the fact that it is retrospective data collection, received from the operating centers, and as such subject to significant information bias and selection bias. There were no criteria given for why these patients received this procedure and how they may have differed from any other patient being treated for Hirschsprungs at this time at these centers. The data collection came from the centers themselves. Follow up occurred by telephone survey and had a low rate of contact (27%). All of these facts make the paper subject to recall bias. There was no report of pre-determined study questions, and thus the associations observed and analyzed by chi-square statistics need to have the issue of multiple comparisons addressed in the statistical analysis in order to have valid p-values. As these were not presented, these p-values are overestimations of the associations and cannot be accepted at face value as statistically significant.

This study by Sherman is a very large, multi-center case series with long-term results on the Swenson procedure as performed on 880 patients. It is valuable for showing what outcomes these seven centers have had and it does support the desire of the authors to dispel the myth that only Swenson and his co-workers can have consistently good results. There is no evidence, given the lack of a true comparison group, however, to accept the authors concluding sentence that “it seems physiologically correct to remove all of the distal aganglionic bowel rather than just part of it.”

- II. Teitelbaum DH, Cilley RE, Sherman NJ, Bliss D, Uitvlugt ND, Renaud EJ, Kirstioglu I, Bengston T and Coran AG. A decade of experience with the primary pull-through for Hirschsprung Disease in the newborn period. *Ann Surg* 232(3):372-380, 2000.

Presented at the American Surgical Association annual meeting, April 2000.

Presented by Jasmin Jose, PA

Reported	Not App.	<b>REPORTING DETAIL – METHODS SECTION</b>	Comments
yes		A clear description of study design	A retrospective case series with a historical control group
yes		The number and practice type of institutions where cases were performed	4 sites
no		Number of surgeons who actually operated in study	
yes		A statement as to whether the same surgeon operated on pts from different treatment groups or just one group	Control group only from 1 of the 4 centers
yes		The precise timeline during which patients were treated	May '89 – Sep '99
no		A clear description of how patients were selected into study	
no		The number of eligible patients at the study sites excluded during the study	
esy		A clear description of the study population	
yes		A clear description of the relevant diagnostic criteria used to identify	

		cases	
yes		A clear description of critical aspects of operative technique and peri-operative care	
yes		Statement as to whether any attempts were made to standardize operative technique or peri-operative care	“using the same surgical technique”
		<b>RESULTS</b>	
yes		Range and mean of all relevant demographic and baseline variables	See summary
		Range and median for length of follow-up reporting	
yes		Relevant outcome variables are presented with appropriate measures of range and variability	
yes		Methods for measuring outcomes of interest are clearly described	
yes		Statement is made whether any data is missing (and how missing data is addressed)	
yes		Number and appropriate details regarding all complications	
		<b>IF MORE THAN ONE TREATMENT GROUP</b>	
yes		Mean and range for all relevant demographic and baseline variables for all treatment groups	
no		The range and median for length of follow-up reporting for each treatment group	
yes		A precise timeline during which all patients were treated for each group	
no		Outcome variables being compared between groups are presented with appropriate measures of variability (eg standard deviation)	
yes		Measure of type I error: (p-values) for comparison statistics are presented with actual values if p=0.01 or larger (eg. P=NS and p<0.05 are not acceptable).	
no		Type I error cont: Confidence interval presented	
no		Measures of type II error should not be >0.2 (study should have at least 80% power)	
yes		A description of how patients were selected into each treatment group	
yes		A statement is made as to whether the same surgeons operated on patients from different treatment groups	

Summary: A retrospective case series report with historical control group covering four surgical centers over a ten year period. 78 children were treated by primary endorectal pull-through as part of the case series and 103 children treated by a two-stage approach during the past fifteen years (from a database of 131) served as the historical controls.

Result summary:

The demographics of the treatment and control group were similar in sex distribution (69% boys) and incidence of associated anomalies (31%). Length of aganglionosis was also similar between the two groups. However, the control group was older at time of diagnosis (8.8 months versus 13.5 days), mean age at pull-through was also older (19.6 months vs 17.8 days) and the incidence of preoperative enterocolitis was higher (22.7% vs 5%) in the control group. All of these differences must be carefully considered when examining the results of the study.

Table 1: Stooling grading sheet used during telephone interview

Table 2: Percent of early complications in the two groups

One significant difference (p value of <0.001) for anastomotic dehiscence which was higher in the staged pull-through group than the primary pull-through group (9.7% vs 2.6%) Bear in mind the differences between the two groups in incidence of preoperative enterocolitis when considering this outcome.

Table 3: Percent of late complications in the two groups

One significant outcome in the incidence of enterocolitis. Enterocolitis occurred more frequently in the primary pull-through group compared to the staged pull-through group (42.3% vs 22%). Near significant outcome in the rate of anastomotic stricture between the two groups (26.2% in the staged pull-through group vs 15.4% in the primary pull-through group).

Discussion: The major problem with this study is the comparison group. The comparison group data does not include information about how and why patients were assigned to each group. These facts question the relevance of the comparison group to the control group. The patients for the treatment group were “carefully selected” as the authors discussed. The value of this study is in showing that in this carefully selected group of newborns, the primary pull-through procedure can be carefully and successfully done.

- III. Langer JC, Durrant AC, de la Torre L, Teitelbaum DH, Minkes RK, Caty MG, Wildhaber BE, Ortega SJ, Hirose S and Albanese CT. One-stage transanal soave pullthrough for Hirschsprung disease: a multicenter experience with 141 children. *Annals of Surgery* 238(4):569-576, 2003.

Presented by Peter Yoo, MD

Reported	Not App.	<b>REPORTING DETAIL – METHODS SECTION</b>	Comments
yes		A clear description of study design	A retrospective case series
yes		The number and practice type of institutions where cases were performed	6 sites
no		Number of surgeons who actually operated in study	
	X	A statement as to whether the same surgeon operated on pts from different treatment groups or just one group	No control group
yes		The precise timeline during which patients were treated	1995-2002
no		A clear description of how patients were selected into study	
no		The number of eligible patients at the study sites excluded during the study	
yes		A clear description of the study population	“all children who had a 1-stage transanal Soave pullthrough procedure”
no		A clear description of the relevant diagnostic criteria used to identify cases	
yes		A clear description of critical aspects of operative technique and peri-operative care	
yes		Statement as to whether any attempts were made to standardize operative technique or peri-operative care	“technique was basically the same although minor variations were present”
		<b>RESULTS</b>	
yes		Range and mean of all relevant demographic and baseline variables	See summary
yes		Range and median for length of follow-up reporting	
no		Relevant outcome variables are presented with appropriate measures of range and variability	
no		Methods for measuring outcomes of interest are clearly described	
yes		Statement is made whether any data is missing (and how missing data is addressed)	
yes		Number and appropriate details regarding all complications	Table 3 in paper

Summary: A retrospective case series of 6 north American centers from 1995-2002 including 141 patients. There was no comparison group for this series. This paper represents a description of the experience of these centers. Although this is a large study, the size can lend precision to a study but does not imply validity. The large size and multiple centers does help support the authors’ claim that the procedure can be done safely and efficaciously by a variety of surgeons and centers.

Result summary: 141 children. 113 (80.1%) boys, 28 (19.9%) girls

Avg gestational age: 39 +/- 0.78 weeks

Mean birth weight 3.27 +/- 0.19 kg

Mean time until surgery 31.9 +/- 19.8 days

77% rectosigmoid transition zone

mean time to oral feeding 36 +/- 19.3 hours

mean postoperative hosp stay 81.3 +/- 30.8 hours  
 early postop complications 38 pts (27%)  
 postop enterocolitis 9 pts  
 mean length of follow-up 20.2 +/- 9.2 months

Table 1: 6 participating centers

Table 2: Effect of identifying the pathological transition zone before beginning the anal dissection – no effect on time to oral feeds, precent narcotic use, hours of hospitalization

Table 3: Table of complications by center

Enterocolitis 6%, Perianal excoriation 11%, Stricture 4%, Death 0.7%, Other 5%

Discussion: This study is a large study describing the experience at 6 centers with a one-stage transanal soave pull-through for Hirschsprung’s disease. The variability of patient populations and patient care routines from multiple centers lends generalizability to this study. There was no comparison group in this study against which to compare the results, thus complications rates were compared to those published in the literature. Overall, the authors show that his procedure can be done safely with few complications in selected patients. However, the selection criteria for the patients who received this procedure was not discussed so it is unclear which specific group of patients this study might represent.

- IV. Georgeson KE, Cohen RD, Hebra A, Zona JZ, Powell DM, Rothenberg SS, Tagge EP. Primary laparoscopic-assisted endorectal colon pull-through for Hirschsprungs disease: a new gold standard. *Annals of Surgery* 229(5):678-583, 1999.

Presented by David Tashjian, MD

Reported	Not App.	<b>REPORTING DETAIL – METHODS SECTION</b>	Comments
No		A clear description of study design	
yes		The number and practice type of institutions where cases were performed	
no		Number of surgeons who actually operated in study	
	X	A statement as to whether the same surgeon operated on pts from different treatment groups or just one group	
no		The precise timeline during which patients were treated	
yes		A clear description of how patients were selected into study	
no		The number of eligible patients at the study sites excluded during the study	
no		A clear description of the study population	
no		A clear description of the relevant diagnostic criteria used to identify cases	
yes		A clear description of critical aspects of operative technique and peri-operative care	
no		Statement as to whether any attempts were made to standardize operative technique or peri-operative care	
		<b>RESULTS</b>	
no		Range and mean of all relevant demographic and baseline variables	See summary
no		Range and median for length of follow-up reporting	
no		Relevant outcome variables are presented with appropriate measures of range and variability	
no		Methods for measuring outcomes of interest are clearly described	
no		Statement is made whether any data is missing (and how missing data is addressed)	
no		Number and appropriate details regarding all complications	

Summary: This study is a multi-center, case series discussing the outcomes after primary laparoscopic pull-through in 80 patients at 6 sites over five years.

Result Summary: 86% had transition zone in rectum or sigmoid  
 87.5% were under 6 months of age at the time of the procedure

74 (92.5%) patients had a bowel movement within 24 hours  
Average time to ad lib feedings: 28 hours  
Mean time to discharge was 3.7 days  
10 (12.5%) readmissions to the hospital  
4 required postoperative GI diversion on readmission – 1 for severe enterocolitis, 2 for anastomotic leak,  
and 1 for a congenital syndrome  
6 patients had frequent stools for more than 6 months

Table 1: Participating centers

Table 2: Early complications after procedure

Enterocolitis 6 (7.5%)	Bleeding 1 (1%)
Chronic diarrhea 6 (7.5%)	Recurrent constipation 1 (1%)
Anastomotic leak 2 (2.5%)	Conversion to open 2 (2.5%)

Discussion: This article is a good discussion of the experience of these authors with a new technique and their opinions about this procedure. There are no criteria indicated for the selection of patients and no demographics reported about the patients in this series. Because of this fact, it is not possible to determine in which patients this procedure may be best applied. In the discussion section the authors link their new technique to their conclusions as to why it is a better procedure, however, there is no evidence presented during the paper to prove these connections. This study does not include a comparison group but implies comparison to those previously reported results in their statement that this procedure “appears to reduce the postoperative recovery time and perioperative complications dramatically when compared with open pull-through procedures.” This statement is made in the absence of a description of this patient study group and without a valid comparison group. Instead of being considered “a new gold standard,” this paper may have been better titled “a preliminary presentation of promising results.”