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Seeking Enteral Autonomy with **Teduglutide**







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Patients with short bowel syndrome (SBS) often struggle to maintain nutrition and hydration status. Using a combination of diet and pharmacotherapies, partial or full enteral autonomy may be achievable over time as the bowel adapts. There are select groups that cannot achieve autonomy with conventional therapy, but with augmentation of the absorptive capacity of their existing small bowel, autonomy may be feasible and quality of life significantly improved. With the approval of the GLP-2 analog intestinotrophic agent teduglutide, adaptation and intestinal absorption enhancement is possible. The following describes the course of 7 patients on maximal conventional SBS therapy who achieved either enteral autonomy or significant improvements in nutritional status, stool and urine output, and quality of life after initiation of teduglutide.

INTRODUCTION

hort bowel syndrome (SBS) causes severe malabsorption most often due to significant resection or defunctionalized segments of the small intestine. It presents an extremely difficult challenge for clinicians and is a life altering condition for patients. Despite aggressive management of SBS with dietary modification and gut slowing agents, the use of parenteral nutrition (PN) is often required to maintain adequate nutrition and hydration. Resource utilization and healthcare costs for the necessary care of short bowel patients are high and our current healthcare

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system does not lend itself to the time needed in caring for these patients.

The goal of autonomy from PN facilitates avoiding both complications of SBS as well as those of PN such as line infections and vascular thrombosis. Successful enteral autonomy is achieved by enlisting a multifaceted approach including: dietary modification to initiate and stimulate intestinal adaptation, fluid management, and pharmacotherapy. The goal of this approach is to gradually decrease (or eliminate) a patient's dependence on intravenous fluids and PN. The introduction of intestinal mucosal trophic agents such as the glucagon-like peptide 2 (GLP-2) analog, teduglutide, has added another facet to this approach. Incorporating the strategic use of trophic agents such as teduglutide may reduce or eliminate PN requirements in previously PN-dependent patients.

Endogenous GLP-2 is a locally active hormone secreted from the terminal ileum/proximal colon

promoting crypt cell growth and reducing enterocyte apoptosis, while also stimulating intestinal blood flow. Early research demonstrated that exogenous GLP-2 stimulated growth of the intestinal mucosa, which led to enhanced fluid and nutrient absorption. Clinical trials of the GLP-2 analog, teduglutide, demonstrated improved intestinal function in short bowel patients. The first randomized control trial performed to evaluate the efficacy of teduglutide in SBS showed a significant decrease in dependence on parenteral support over a 24week period. The 28-week extension of this trial (52 total weeks of treatment) demonstrated that over 50% of patients had a 20% or more decrease in PN dependence.² In a subsequent 2 year open-label extension trial, an even more significant reduction in total PN volume was described in patients treated with teduglutide with 15% (13/88) patients enrolled achieving full enteral autonomy.3

The following illustrates our institution's experience with 7 cases where strategic use of teduglutide was utilized for short bowel patients on otherwise maximal therapy. Patient initials have been changed to protect patient confidentiality.

Case 1

WS is a 66 y/o female with a history of SBS as a result of mesenteric ischemia. Her GI anatomy consists of 105cm of normal proximal small bowel terminating in an end jejunostomy. She was unable to maintain adequate hydration status 18 months post-op from bowel resection with significant ostomy output making enteral feeding quite difficult. She was initiated on teduglutide after maximizing the use of high dose codeine. She was long

past the hypersecretory phase. Over the initial 3 months of teduglutide therapy, she successfully transitioned from nightly IV hydration fluids to nocturnal oral rehydration therapy infused via a gastrostomy tube. Her urine output increased significantly during this time indicating improved hydration and her stool output became much more manageable. Over the first year of therapy with teduglutide, WS was able to gradually reduce nightly hydration via gastrostomy; 13 months after initiating teduglutide she was succeeding with oral nutrition and hydration alone. She now has enteral autonomy, her gastrostomy is removed, and she relies on careful management of her PO diet in addition to gut slowing agents, which we were also able to wean. She now volunteers at the hospital and exercises at the gym 3 times a week.

WS stands as an example for the potential for using intestinal growth factors such as teduglutide to free patients from the use of PN, IV fluids, and augmentation of nutrient absorption via gastrostomy. She also highlights the usefulness of gastrostomy tubes in this patient population. With careful management, many patients with significant malabsorption can be managed quite well by a combination of enteral feedings (or oral rehydration) instilled slowly through a nasogastric or gastrostomy tube via a pump. This can be a very effective tool in patients who are unable to wean from PN or IV fluids, and/or tolerate sufficient PO intake. See Box 1

Case 2

LP is a 31 y/o gentleman with a history of SBS as a result of necrotizing enterocolitis as a child. His GI

Box 1

Parameter	Pre-teduglutide	Post teduglutide (6mo)	Post teduglutide (12mo)
Weight (goal 77kg)	76.7kg	77.1kg	82.9kg
Stool Output	2500mL	1000-1750mL	1000mL or less
Urine Output	880mL	1300-1750mL	>1000mL
IV fluids	1500mL	Off 3 months prior	None
PN	None	None	None
ORT/Tube Feed	470mL daily	1250mL per night by PEG	None
SBS diet	+	+	+
Anti-diarrheal agents	Codeine 45mg TID +60mg QHS (crushed)	Codeine 45mg BID am, 60mg BID (crushed)	30-45mg Codeine TID (no longer needs to be crushed)

Box 2

Parameters	Pre- Teduglutide	Post Teduglutide (6mo)	Post Teduglutide (12mo)	Post Teduglutide (1.5yrs)	Post Teduglutide (2yrs)	Post Teduglutide (3yrs)
Weight (goal 52 kg)	44.9kg	53kg	53kg	51.4kg	48.7kg	50.7kg
Stool Output	2500mL	1500mL	2000mL	2000mL	2000mL	2200mL
Urine Output	<1050mL	1100mL	800mL	2400mL	1700mL	1275mL
IV fluids	3L IV fluids QHS	3L IV fluids QHS	3L IV fluids QHS	3L IV fluids QHS	2L IV fluids QHS	2L IV fluids QHS
PN	None	None	None	None	None	None
ORT/Tube Feed (via PEG)	Peptamen 1.5 (1000mL) QHS	Peptamen 1.5 (1500mL) QHS	Peptamen 1.5 (1000mL) QHS	Peptamen 1.5 (750mL) QHS	Peptamen 1.5 (750mL) QHS + 1L Pedialyte 4 days /week	Peptamen 1.5 (1500mL) QHS +1L Pedialyte 4/days/week
SBS Diet	+	+	+	+	+	+
Anti-diarrheal agents	Imodium 4mg QID	Imodium 4mg QID	4mg Imodium QID	6mg Imodium TID	4mg Imodium QID	4mg Imodium QID

anatomy consists of 30cm of normal proximal small bowel anastomosed to 50cm of distal colon. Prior to starting teduglutide, he was transitioned from PN to a regimen of nocturnal enteral feeding via PEG in addition to nightly IV fluids. This was sufficient for him to maintain a marginal weight and urine output. Escalation of gut slowing agents was somewhat limited by a history of bowel obstruction. After initiation of teduglutide at a standard dose, he had a significant improvement in his weight over the initial 6 months of therapy. While his weight stabilized at his goal, he was able to decrease the amount of tube feeding required to maintain this weight in addition to oral intake. His urine output gradually improved to over 1L daily with a decreased IV fluid requirement. He has been maintained on teduglutide for 3 years and continues on a stable regimen at or around his goal weight. As his strength increased, he began working longer hours and started an exercise program. We had to increase his enteral feedings overnight to compensate for his higher energy expenditure.

LP experienced no significant side effects from initiation of teduglutide. Three years into therapy, he was admitted for 7 days for symptomatic small bowel obstruction. This was managed conservatively and required no surgical intervention. He was subsequently

restarted on ½ dose teduglutide 2 weeks after discharge, which he tolerated well and then progressed back to full dose. During this brief period, he experienced increased stool output and mild weight loss, so his nutrition support was adjusted as needed. See Box 2.

Case 3

JF is a 48 y/o female with a distant history of Roux-en-Y gastric bypass now with SBS as a result of a volvulus followed by significant bowel ischemia. Her GI anatomy consists of Roux-en-Y gastric bypass anatomy to small bowel to colon via an ileocolic anastomosis. She had been on PN for 7 years. However, because of a history of multiple line infections, it was decided to hold her PN, optimize gut slowing, and enlist intense diet and hydration instruction. She did well at first, but difficulty with adherence ultimately resulted in precipitous weight loss and significant fat-soluble vitamin deficiency. We then admitted her for a nocturnal enteral feeding trial with concurrent 72 hour fecal fat collection to determine if she had sufficient absorptive capacity. However her fecal fat wasting was profound so further enteral feeding was abandoned. She was restarted on PN and gradually made improvements in both her weight and hydration status. She was then started on teduglutide. She had a history of lower extremity edema prior to teduglutide

Box 3

Parameters	Pre-Teduglutide	Post Teduglutide (3months)	Post Teduglutide (5months)
Weight (goal 68kg)	56.6kg	66.2kg	74kg
Stool Output	2300mL	2100mL	1300-1450 mL
Urine Output	800mL	2500-3025mL	1350mL
IV fluids	None	None	None
PN	1200kcal in 1440mL	1200kcal in 1440mL	830kcal in 1440mL
ORT/Tube Feed	Failed nocturnal enteral trial		
SBS Diet	+	+	+
Anti-diarrheal agents	Chronic narcotics for pain (dilaudid)	Chronic narcotics for pain (dilaudid)	Chronic narcotics for pain (dilaudid)

initiation, but did not require titration of her diuretics during the first few months of therapy. She currently has manageable stool output and is on chronic narcotics for pain, but no additional gut slowing agents.

Initiation of teduglutide has resulted in improved weight gain and hydration status over the first 5 months of therapy and resolution of her lower extremity edema. This case is significantly complicated by the pre-existing Roux-en-Y gastric bypass anatomy, which requires particular attention to the risks of vitamin deficiency. Non-adherence played a partial role as well. Fluid retention is a known risk of teduglutide and may require careful observation for rapid weight gain and/or edema during the initiation weeks to months after starting therapy in those who are sensitive to fluid overload. As of this writing, she has reached her goal

weight, and we are planning to decrease PN from 7 to 6 days, giving her a night off PN every week. See Box 3.

Case 4

MS is a 47 y/o gentleman with a history of SBS as a result of mesenteric ischemia secondary to thrombosis. His GI anatomy consists of approximately 200cm of small bowel terminating in an end-ileostomy. There is a suggestion of dilated, defunctionalized bowel on small bowel imaging. Although he has a significant amount of colon, it is not in continuity. Prior to starting teduglutide, he attempted discontinuation of his PN with dramatic weight loss and high ostomy output with attempts at increased PO. He was admitted for a combination SBS diet and nocturnal enteral feeding trial using a nasogastric tube concurrent with a 72-hour fecal fat

Box 4

Parameter	Pre-Teduglutide	Post Teduglutide (4 months)
Weight (goal 80.7kg)	66.9kg	77.4kg
Stool Output	1500mL	1350mL
Urine Output	1160mL	1325mL
IV fluids	As needed	As needed
PN	1200kcal in 2500mL	Unchanged
ORT/Tube Feed	Failed nocturnal enteral trial	
SBS Diet	+	+
Anti-diarrheal agents	2mL Tincture of Opium q6h	Stopped

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collection and demonstrated significant malabsorption with high ostomy output despite high dose gut slowing agents. He had also experienced stomal stenosis, which required dilation. He was initiated on a halfdose of teduglutide because of the stomal stenosis and demonstrated a robust response while on a combination of PO intake with no change in his PN solution. His weight improved over 3 months nearing his goal weight and his ostomy output required decreasing doses of gut slowing agents. Over the subsequent few months, he experienced more discomfort around the stenosis of his end ileostomy altering his oral intake and worsening his ostomy output due to outflow diarrhea. A decision was made to revise this site with plan for placement of the colon back in continuity with the small bowel in an effort to facilitate improved enteral absorption and fluid management and possibly permit weaning of PN. See Box 4.

Case 5

CR is a 50 y/o female with a history of SBS as a result of surgery and radiation therapy for cervical cancer. Multiple surgeries for mechanical bowel obstruction resulted in her GI anatomy consisting of 45cm of viable small bowel and an additional 35cm of irradiated, defunctionalized bowel anastomosed to the colon with the sigmoid colon terminating in an end ostomy. Prior to starting teduglutide, CR was unable to reach her goal weight or control her stool output despite PN for

nutrition support and attempts at gut slowing. CR has had a great deal of difficulty adhering to medication/ nutrition regimens and tracking her urine and stool outputs. Her clinical course has also been complicated by line infection. After initiation of teduglutide, her weight has remained relatively stable and she has tolerated a gradual tapering of her PN from 4 days/ week to 1L of IV fluids plus electrolytes and vitamins 3 days weekly. While she has not reached her goal weight, her current nutrition support strategy with IV fluids is lower risk relative to PN, and the current regimen, in combination with gut slowing and SBS diet, has been sufficient to maintain hydration and meets her preferences. See Box 5.

Case 6

KJ is a 57 y/o male with longstanding Crohn's disease with prior small bowel resections in 1991 and 2001 with approximately 50% of the small bowel remaining. The patient also underwent liver transplantation in 2008 due to nodular regenerative hyperplasia. In 2012, he developed increasing GI symptoms in the setting of colonic pneumatosis. While the etiology of pneumatosis was never identified, and despite resolution on subsequent imaging, his GI symptoms did not return to baseline. PN and IV fluids were required to maintain hydration and adequate urine output. Attempts at tapering IV fluids led to reduced urine output and worsening renal function and 2 episodes of nephrolithiasis. The patient was started on teduglutide

Box 5

Parameters	Pre-Teduglutide	Post Teduglutide (6mo)	Post Teduglutide (12mo)	Post Teduglutide (1.5yrs)	Post Teduglutide (2yrs)
Weight (goal 45.3 kg)	41.7kg	41.3kg	41.8kg	42.3kg	41.7kg
Stool Output	>2L day	"a lot"	1500mL	Unrecorded	"a lot"
Urine Output	1000mL	"good"	Unrecorded	1500	"a lot"
IV fluids	None	None	None	1L + electrolytes MWF	1L + electrolytes MWF
PN	4 days/week	4 days/week	4 days/week	Off	Off
ORT/Tube Feed (via PEG)	Failed enteral trial	Failed enteral trial	None	None	none
Diet	SBS	SBS	SBS	SBS	SBS
Anti-diarrheal agent	4mL tincture of opium QID	4mL tincture of opium QID	4mL tincture of opium QID	4mL tincture of opium QID	4mL tincture of opium QID

Box 6

Parameter	Pre-Teduglutide	Post-Teduglutide (4 months)	Post-Teduglutide (8 months)	Post-Teduglutide (16 months)
Weight (goal 70.5kg)	66kg	65.3kg	68.1kg	70.2kg
Stool Output	1500mL	200mL	300-500mL	300-500mL
Urine Output	1250mL	1275mL	2400mL	2500mL
IV fluids	2 liters daily	1.5 liter daily	1.5 liter daily	Off
PN	none	none	none	none
ORT/Tube Feed	ORT; declined enteral trial	Oral diet	Oral diet	Oral diet
Diet	Relative SBS, low oxalate	Relative SBS, low oxalate	Relative SBS, low oxalate	Relative SBS, low oxalate
Anti-diarrheal agents	Codeine 30-45mg QID	Loperamide prn	Loperamide prn	Loperamide prn

in June 2013. The patient had a significant reduction in stool volume after initiation; slower improvement in weight and urine output followed, and IV fluids were reduced 4 months later. By month 16 the patient was off IV fluids and was maintaining adequate hydration with oral intake alone. No changes in post-transplant medications were necessary, and he has enjoyed traveling both nationally and internationally without incident. See Box 6.

Case 7

NN is a 54 y/o male with SBS related to Crohn's disease with multiple prior small bowel and colon resections with resultant transverse colostomy. His last surgery was in March 2016 at which time he underwent an

ileocolic resection due to anastomotic stricturing. Postoperatively he struggled with high ostomy output, weight loss, and difficulty with hydration. Small bowel imaging showed approximately 100cm of small bowel remaining, and stool testing showed significant malabsorption with 97g of fat per 24 hours (normal 2-7g). He was initiated on PN in December 2016, but developed a line-related infection shortly after initiation. Teduglutide was initiated in February 2017; weight increased significantly after initiation and he was able to wean off parenteral support in July 2017. See Box 7.

DISCUSSION

Living with SBS is extremely challenging for patients, not only as a result of the short bowel and associated

Box 7

Parameter	Pre-Teduglutide	Post Teduglutide (5 months)
Weight (goal 72kg)	65 kg	71.4 kg
Stool Output	2300 mL	1150 mL
Urine Output	1500 mL	1225 mL
IV fluids	2000 mL	Off
PN	Off	Off
ORT/Tube Feed	Failed enteral trial	Oral diet
SBS Diet	+	+
Anti-diarrheal agents	Codeine 60mg QID	Codeine 60mg BID-TID

diarhea in particular, but also from the labor-intensive regimens that include careful meal planning, medication adherence, coordination of PN/IV fluids and enteral feedings from preparation to delivery to administration. All of these factors have a great impact on quality of life for patients with SBS. Anything clinicians can do to ease this burden is a worthy goal. This requires focusing on issues that are most important to patients. Interventions that decrease side effects of SBS are extraordinarily meaningful to patients—for example, one of our patients reported the following after initiating codeine for gut slowing instead of the loperamide he was previously using:

"I awoke at 4:30am and went fishing with the TPN hanging on my back. Bathroom once on the way there and once again at 2:30! That's incredible!" and later, "with the decrease in my diarrhea on the codeine, I was able to take 2 hours off my usual 9 hour trip up to Virginia as I did not have to find a bathroom nearly as often!!"

The use of intestinotrophic agents such as

teduglutide can play a vital role in improving patient quality of life by helping to manage core issues in SBS to include improving nutritional status, limiting stool/ostomy output, and decreasing dependence on parenteral support.

Careful patient selection is essential to maximizing the benefits of trophic agents while minimizing the risks. Gastrointestinal dysplasia and cancer are contraindications to teduglutide therapy given concern for the trophic effects on a potential malignancy. A history of biliary disease or pancreatitis is another. It should be clear that patients should maximize other potential therapies⁵⁻⁸ before initiating teduglutide given the significant cost. Following this, a careful discussion with the patient regarding the risks and potential benefits should be undertaken and reasonable goals set. It may not be possible for a short bowel patient to discontinue PN due to teduglutide therapy, but it may be reasonable to hope for fewer days of PN each week or significantly shorter infusion times, less diarrhea, and an increase in overall well-being.

Finally, it is crucial to keep in mind (and express

Table 1. Considerations When Weaning Short Bowel Patients Off Parenteral Support (continued on page 50)

Parameter	Considerations
Weaning PN/IV Solution	 Reduce PN/IV solution in stepwise fashion in 2-4-week intervals Consider need for escalation of bowel slowing agents such as Imodium or codeine—may need to crush tablets to increase efficacy; avoid capsules Consider enteral feeding or hydration as a tool to continue nutrition support while weaning PN.
Weight	 Determine and maintain goal weight range Patient weighs 2-3 times per week
24 hour Stool/ostomy Urine	 ◆ 2-3 days per week ◆ Ideally < 1500mL/day ◆ Ongoing maintenance of daily urine output of > 1200mL; 1500mL if known nephrolithiasis
Labs	♦ Weekly as appropriate per individual patient
Sodium ^{9,10}	 ◆ 24-hour urine for sodium if depletion suspected (end ileostomy/jejunostomies at risk for this) ◆ Maintain urinary sodium concentration > 20mEq/L
Vitamin & Mineral Supplementation	 ◆ Start therapeutic vitamin & mineral supplement when PN requirements drop to 5 nights or less per week. ◆ Do not use capsules (often pass through whole) ◆ Chewable therapeutic vitamin/mineral tablet either daily, or ½ tab BID ◆ Gummies—but verify content is complete therapeutic and watch for number pills needed per day to meet 100% needs ◆ Liquid version also good alternative—be aware zinc content is < tablet & may need to supplement over that amount if zinc deficient

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Table 1. Considerations When Weaning Short Bowel Patients Off Parenteral Support (continued from page 48)

Parameter	Considerations
Vitamin D ¹¹⁻¹³	 ♦ Vitamin D is notoriously low in these patients, therefore larger doses are needed to achieve normal serum levels. ♦ Check 25 OH vitamin D every 2-3 months until stable, then twice per year. ♦ Check intact PTH at baseline, then as needed after. ♦ Check baseline DEXA scan, then as needed. ♦ Note: Standard 10mL MVI has only 200 units vitamin D2. ♦ Avoid 50,000 weekly dose • This limits patients to one opportunity per week to get some vitamin D—if they lose the tablet in their stool that day, game over. • Daily dosing increases the chances that some will be absorbed. • If dose does not increase level, double the dose and recheck in 2-3 months; if no change, double the dose again and consider giving half BID. ◆ Give daily dose as tablet (even crushed), or liquid (not capsule) ◆ Use tablets crushed or liquid form—if insurance does not cover liquid, have patient obtain liquid from store such as the Vitamine Shoppe, GNC, etc. ◆ Other options available: • Superior Source—microlingual technology available in 400, 1000, 5000, 10000units. • http://superiorsourcevitamins.com/microlingual-tablets/a-d-k-vitamins • Bio-D-Mulsion Forte Drops—absorbed on the tongue. • http://www.webmd.com/drugs/2/drug-152707/bio-d-mulsion-forte-oral/details. • UV Light • Sun exposure—If you're fair skinned, 10 minutes in the midday sun in shorts and a tank top with no sunscreen—will give you enough radiation to produce about 10,000units of the vitamin. • In winter, if you live north of Atlanta, it is impossible to produce adequate vitamin D from the sun because the sun never gets high enough in the sky for the UV B rays to penetrate the atmosphere. • Sperti lamp—UV light—average Vitamin D Lamp sessions are 3-5 minutes, only 2 or 3 times per week.
Calcium	 Determine approximate usual daily intake Ensure 1000-1500mg daily Keep each dose at 250-500mg at a time, either BID-QID
B12 1000mcg oral daily tablet 1000mcg SQ or IM monthly Sublingual	 B12 SQ vs. IM (SQ less painful) In patients with known small bowel bacterial overgrowth: check both serum B12 and methylmalonic acid if on oral supplementation. When transitioning patient to oral from monthly IM or SQ dosing, check B12 and methylmalonic acid every 6 months until stable to ensure plan works for that individual patient. Stop checking B12 & methylmalonic acid status if patient on 1000mcg monthly SQ or IM injections Unless non-adherence a concern.
Iron	 May need liquid form (Fer-in-sol = 1 mL = 15mg elemental iron) – BID-QID as needed. Have patient take with 50-60mg Vitamin C such as Nature-made VitaMelts™. Do not use sustained release (SR), controlled release (CR), or extended release (EC) iron supplements in SBS patients
Essential Fatty Acids	 Triene:tetraene ratio < than 0.4 = deficiency Annually if deficiency suspected Try to incorporate high EFA fats in patient's diet such as flaxseed, sunflower, walnut, soybean oils

Table 2. Resources For Clinicians

For Clinicians

Professional Texts

Short Bowel Syndrome: Practical Approach to Management. Eds. DiBaise JK, Parrish CR, Thompson JS. CRC Press; Taylor & Francis Group, Boca Raton, FL, 2016.

Recent 6-part series on Managing Short Bowel Syndrome in Practical Gastroenterology

- 1. DiBaise J, Parrish CR. Part 1: Short Bowel Syndrome in Adults Physiological Alterations and Clinical Consequences. Practical Gastroenterology 2014;Aug(8):30.
- 2. Parrish CR, DiBaise J. Part II: Nutrition Therapy for Short Bowel Syndrome in the Adult Patient. Practical Gastroenterology 2014;Oct(10):40.
- 3. Parrish CR, DiBaise J. Part III: Hydrating the Adult Patient with Short Bowel Syndrome. Practical Gastroenterology 2015;Feb(2):10.
- 4. Chan LN, DiBaise J, Parrish CR. Part IV-A: A Guide to Front Line Drugs Used in the Treatment of Short Bowel Syndrome. Practical Gastroenterology 2015;March(3):28.
- 5. Chan LN, DiBaise J, Parrish CR. Part IV-B: A Guide to Front Line Drugs Used in the Treatment of Short Bowel Syndrome. Practical Gastroenterology 2015;April(4):24.
- 6. DiBaise J, Parrish CR. Part V: Short Bowel Syndrome in Adults Part 5: Trophic Agents in the Treatment of Short Bowel Syndrome. Practical Gastroenterology 2015;May(5):56.

Extensive Professional & Patient Education Materials for Short Bowel Syndrome

- University of Virginia Health System GI Nutrition Website:
 - ◆ Under Patient Education Materials link
 - Extensive written short bowel diet education and hydration materials

For Patients

Patient Educational Guidebooks:

Parrish, CR. A Patient's Guide to Managing a Short Bowel, 4th Edition; June 2016:1-66. Available at no cost to patients and clinicians at: http://www.shortbowelsyndrome.com/sign-up

Olev Foundation

www.oley.org; (800/776-0LEY)

Short Bowel Syndrome Foundation

www.shortbowelfoundation.org; (888-740-1666)

to patients) that trophic agents such as teduglutide are not a substitute for a comprehensive SBS program, but rather an adjunct to this program.

Weaning from PN

The weaning of parenteral support can be a daunting task for short bowel patients, particularly those who have struggled in the past to maintain weight and hydration. We typically begin this process once a patient has stabilized at or near their goal weight, unless clinical course accelerates our decision (multiple septic episodes, unwieldy stool output on maximum anti-diarrheal/anti-secretory agents, etc.).

For patients on PN, we make stepwise decreases in total fluid volume and macronutrient content (Table 1). Weekly labs are followed closely as are patient's weights, stool output, and urine output, particularly in patients who attempt to increase their PO intake to compensate for decreased PN support, which may make stool output increasingly difficult to manage. This frequently requires the escalation or addition of anti-diarrheal agents such as loperamide or codeine. A subset of patients is unable to increase their PO intake successfully without uncontrolled stool output. Our experience has suggested that many of these patients can still work toward enteral autonomy through the

use of a gastrostomy tube using a pump for nightly feedings at a decreased, but continuous rate for 8-12 hours. However, prior to placing permanent access, a nasogastric nocturnal feeding trial with concurrent 72hour fecal fat collection is undertaken to ensure we do not drive stool output higher than manageable and to ensure absorption is adequate. Similarly, some patients succeed with an enteral backpack for daytime use to infuse enteral feeding or oral rehydration solutions. The portability of the enteral backpack is often a key to success and quality of life in these patients. A small subset of patients is unable to achieve enteral autonomy. This is most commonly the case in patients with severely foreshortened bowel, defunctionalized remaining bowel, or short bowel in the context of bariatric surgeries. Rather than a change in the above process, these patients often require a change in goals of weaning. For many patients, even one or two nights off of PN per week is very meaningful for their quality of life and often helps facilitate vacations and social functions. A similarly worthwhile alternative goal might be attempting to reduce total weekly PN in favor of IV fluids to support hydration, which has a more favorable risk profile particularly with regards to line infection.

CONCLUSION

Successful management of the challenges SBS patients face requires systematic and strategic intervention. It is important to avoid "throwing the kitchen sink" at patients to prevent overtreatment (which can act to drive stool output further), but also result in unnecessary healthcare costs. This article demonstrates how diet, hydration, and medication selection, including the judicious use of intestinotrophic agents, all play an integral role in the complex management of SBS. See Table 2 for additional resources for clinicians.

References

- 1. Jeppesen PB, Gilroy R, Pertkiewicz M, et al. Randomized placebo-controlled trial of teduglutide in reducing parenteral nutrition and/or intravenous fluid requirements in patients with short bowel syndrome. Gut 2011;60:902e914.
- 2. O'Keefe SJ, Jeppesen PB, Gilroy R, et al. Safety and efficacy of teduglutide after 52 weeks of treatment in patients with short bowel intestinal failure. Clin Gastroenterol Hepatol. 2013;11(7):815-23.e1-3.
- 3. Schwartz LK, O'Keefe SJ, Fujioka K, et al. Long-Term Teduglutide for the Treatment of Patients with Intestinal Failure Associated with Short Bowel Syndrome. Clinical and Translational Gastroenterology (2016) 7, e142.
- Winkler MF, Hagan E, Wetle T, et al. An exploration of quality of life and the experience of living with home parenteral nutrition. JPEN J Parenter Enteral Nutr. 2010;34(4):395-407.
- 5. Parrish CR, DiBaise J. Part II: Nutrition Therapy for Short Bowel Syndrome in the Adult Patient. Practical Gastroenterology 2014;Oct(10):40.
- Parrish CR, DiBaise J. Part III: Hydrating the Adult Patient with Short Bowel Syndrome. Practical Gastroenterology 2015;Feb(2):10.
- Chan LN, DiBaise J, Parrish CR. Part IV-A: A Guide to Front Line Drugs Used in the Treatment of Short Bowel Syndrome. Practical Gastroenterology 2015;March(3):28.
- 8. Chan LN, DiBaise J, Parrish CR. Part IV-B: A Guide to Front Line Drugs Used in the Treatment of Short Bowel Syndrome. Practical Gastroenterology 2015;April(4):24.
- Lauverjat M, Hadj Aissa A, Vanhems P, et al. Chronic dehydration may impair renal function in patients with chronic intestinal failure on long-term parenteral nutrition. Clin Nutr. 2006;25(1):75-81.
- O'Neil M, Teitelbaum DH, Harris MB. Total body sodium depletion and poor weight gain in children and young adults with an ileostomy: a case series. Nutr Clin Pract. 2014;29(3):397-401.
- Napartivaumnuay N, Gramlich L. The Prevalence of Vitamin D Insufficiency and Deficiency and Their Relationship with Bone Mineral Density and Fracture Risk in Adults Receiving Long-Term Home Parenteral Nutrition. Nutrients. 2017 May 10;9(5).
- 12. Fan S, Ni X, Wang J, et al. High Prevalence of Suboptimal Vitamin D Status and Bone Loss in Adult Short Bowel Syndrome Even After Weaning Off Parenteral Nutrition. Nutr Clin Pract. 2017;32(2):258-265.
- 13. Grenade N, Kosar C, Steinberg K, et al. Use of a Loading Dose of Vitamin D for Treatment of Vitamin D Deficiency in Patients with Intestinal Failure. JPEN J Parenter Enteral Nutr. 2017;41(3):512-516.