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Short Bowel Syndrome: The Patient Journey

Announcer:
Welcome to GI Insights on ReachMD. This medical industry feature, titled “Short Bowel Syndrome: The Patient Journey,” is sponsored by Takeda Pharmaceuticals U.S.A., Incorporated.

Here’s your host, Dr. Charles Turck.

Dr. Turck:
Short bowel syndrome is a serious and chronic malabsorption disorder that can have a substantial impact on a person’s health and well-being. For those who are living with this disorder, it’s important to have access to the right information and resources, which is why it’s critical for us clinicians to have a full understanding of this disease.

This is GI Insights on ReachMD and I’m Dr. Charles Turck. Joining me to explore the causes and various management strategies for short bowel syndrome are Drs. Paul Wales and Kelly Tappenden. Dr. Wales is a Professor of Surgery at the University of Cincinnati. Dr. Wales, it’s great to have you here today.

Dr. Wales:
Thanks so much for having me.

Dr. Turck:
And Dr. Tappenden is a Dean Professor at the College of Health at the University of Utah Health. Dr. Tappenden, welcome to you.

Dr. Tappenden:
It’s great to be here. Thank you.

Chapter 1: SBS Pathophysiology and Diagnosis

Dr. Turck:
So, Dr. Wales, can you start by giving us some background on short bowel syndrome and how it can develop?

Dr. Wales:
Well, short bowel syndrome is one category of intestinal failure and intestinal failure occurs when the absorptive capacity of the bowel is inadequate to absorb enough nutrients or fluids to support survival and hydration[1] and in the case of children, growth.[2] In many patients that have lost their bowel, they lose the enteroendocrine cells, the L-cells, in the distal ileum and colon, which are responsible for producing trophic peptides, like glucagon-like peptide 2.[3] GLP-2 is partially responsible for natural nutritional absorption and motility in patients with normal gut health.[3] After the bowel is lost, the bowel that remains is going to try to compensate, it’s going to try to go through this process of adaptation and patients, there’s a spectrum of malabsorption that they experience, which can lead to dehydration and electrolyte disturbances as well as malnutrition.[1,4,5] The patients with short bowel syndrome are very heterogenous because of the large variations in intestinal function and remnant bowel anatomy.[6] There’s limited studies available on the prevalence of SBS due to lack of population-based data.[7] In the United States approximately between and 10 and 20,000 people have this disorder.[8]

Dr. Tappenden:
Excellent stats there for us, Dr. Wales. I just want to add for clinicians who aren’t familiar with seeing this that the level of malabsorption and diarrhea[9] that these individuals suffer is life-threatening,[9] unremitting in many cases, which is why we have to require parenteral

nutrition and fluids in many of them.[9] It is keeping them home bound in many cases because of the liters of malabsorption, diarrhea, if they have a colon that they're experiencing and it just has a huge impact on their ability to go through their daily lives.[9,10]

Dr. Turck:
And as a quick follow-up to that, Dr. Wales, what underlying etiologies can lead to SBS and what does treatment look like if your patient is diagnosed?

Dr. Wales:
The causes of SBS vary between children and adults. In the pediatric space, usually short bowel syndrome is related to neonatal disorders, whether they're congenital or acquired.[11-13] And that would include congenital anomalies of the GI tract, including intestinal atresia, gastroschisis, Hirschsprung's disease or volvulus. And acquired diseases of the neonate, usually premature, necrotizing enterocolitis. For adults, possible diseases or conditions that can lead to SBS include Crohn's disease, complications of bariatric trauma, intestinal volvulus,[14,15] strangulated hernia,[7] vascular disease,[4,7] or small bowel fistula.[7]

If your patient is diagnosed with SBS, the treatment is complex and requires a multidisciplinary approach,[16] which includes many care providers, physicians, surgeons, nurses, dieticians, and all other allied health such as pharmacists, social workers, physiotherapists,[17] occupational therapists.[18]And really, the advent of multidisciplinary care has really been transformational, especially in pediatrics over the last two decades. Presumably the creation of multidisciplinary teams brings together experts with different perspectives to provide better coordination of care and continuity of care, which complex patients really benefit from.[16,17,19-21] And one of the major roles of a multidisciplinary team, not only the optimal medical management, is really to help educate, support,[16,17] and empower the patients and their caregivers, their families.[21]

When we approach patients it's a two-pronged approach. On one hand, we're trying to promote adaptation, trying to optimize intestinal function so that we can minimize or decrease parenteral support, and we have three tools in our toolbox to do this. We can use nutrients, nutrition, and we look at nutrition as a therapy, not just as a supportive measure. What we feed, how we feed, when we feed has therapeutic benefit depending on the remnant anatomy that the patient has.[22] And then we have surgery as well where we'll do reconstructive intestinal procedures to try and optimize function.[22] In addition to that prong the other prong of care that a team must demonstrate, is the mitigation of complications. While we're trying to get these patients to adapt, they're partially or totally dependent on parenteral support and as a result of long-term parenteral support with a central venous catheter, they're at risk for many complications. So, a good multi-D team should be involved in surveying and trying to prevent or mitigate the development of these complications over time.[16]

Chapter 2: A Different SBS Journey for Children

Dr. Turck:
And if we come back to the underlying conditions that can lead to SBS, Dr. Wales, you mentioned that the journey is different for pediatric patients than for adults. So, can you explain what those differences are?

Dr. Wales:
Yeah. Thank you. The big difference between pediatrics and adults is in the pediatric space, we have more than one patient. We have the patient, the actual patient, the child suffering from the condition, and then we have their care providers, which in the majority of cases is their family, their parents.[23]

The other big difference is related to adaptive potential. It is believed that children have a greater adaptive potential than adults do to achieve enteral autonomy. Part of the reason for this is that there's inherent gut growth that takes place in the first several years of life.[12] So, when a term baby is born they're born with about 160 cm of small intestine, and this essentially triples over the first five years of life to about 425 cm.[24] As opposed to an adult who loses a significant amount of their gut, has short bowel syndrome, their adaptive potential is more limited.[12] They don't have that same gut-lengthening potential that a child has.

The other difference is the implications of growth. We have to provide adequate nutritional support, not only to maintain hydration and survival, but children are growing and so we need to make sure that they meet their growth potential, not only with weight but also linear growth.[11,25-27]

Dr. Turck:
Thanks for breaking down those differences Dr. Wales. And if we turn to you now Dr. Tappenden, are there any consequences to long-term parenteral support in children?

Dr. Tappenden:
Oh, they sure are, for children and adults, but children in particular have higher consequences. Now, parenteral nutrition is being credited with being the most important factor responsible for saving the lives of patients with short bowel syndrome, since it's widespread introduction in about 1980 or so,[28] but we can't overlook the fact that there are long-term consequences.[29] Liver disease has been a very big problem.[29] Vascular access is something that we struggle with over time.[29] Metabolically, there are many, many issues that develop from the use of parenteral nutrition. Metabolic bone disease can be a problem.[3] Regulation of glycemic control is often an issue.[19] So, all the more reasons why we need to get these individuals to a multidisciplinary team that really has dialed in the best protocols for managing these issues.[16]

The other interesting thing that happens with long-term parenteral support is that the gut is a little bit like a muscle in that you use it or lose it. If we provide parenteral nutrition and lack the stimulation that oral or enteral nutrients provide to the gut, the mucosa will atrophy and it's exactly the opposite of what we want to happen in these patients.[29] We want growth of the mucosa and increased function, but when we feed parenterally, we aren't providing the stimuli to the gut to cause the expansion of the villus height and crypt depth really cause epithelial cell numbers in the mucosa to grow.[29] So, that's counterproductive to what we want, too. And in kids in particular, if we don't start that normal developmental process of oral nutrition, they can develop what's called oral aversions, and that is something that can stay with them lifelong wherein they simply don't have the ability to eat and will reject oral nutrients. But it can also cause aspiration because they haven't developed that swallowing reflex. It can result in fatal airway obstruction.[18] So that is aggravating this whole process and them developmentally.[30] So, we really want to try and start with oral stimuli and nutrients as much as possible in patients.[30] I find working with occupational therapists to try and avoid oral aversions important,[30] but it also then promotes growth of the intestine and intestinal adaptation and overall minimizes, then, some of those other long-term complications if we can make sure that we're providing the oral and enteral nutrients as much as possible, too.[30]

Dr. Turck:
You're listening to GI Insights on ReachMD and I'm Dr. Charles Turck. Joining me today to talk about the challenges of short bowel syndrome are doctors Paul Wales and Kelly Tappenden.

So, Dr. Wales, now that we've examined the unique SBS journey in children, let's extend this to adolescents and teens. How does SBS evolve as these patients grow up?

Dr. Wales:
Well, early in life, usually the caregivers were likely managing the patient's TPN and other medications.[31] In my opinion, as a child grows up or gets into adolescence, it's important that they play a much larger role in their own care and nutrition.[31] It's really a graduated responsibility. We want them to take ownership over their own care and that includes their central line management, and also paying attention to their own hydration and their diet. The adolescent years are really no different in high school, and especially in college, teens and their caregivers should start talking to their healthcare team as soon as possible about making the seamless transition from their pediatrician, or pediatric gastroenterologist, to a team experienced in adult care.[31] Most multidisciplinary programs should aspire to having a relationship with an adult program.[31] I will say however, that this is a real challenge in most places globally, there is a challenge around transition to adult care. It's not completely isolated to intestinal failure, we do see that with other complex pediatric illnesses as well. But this can sometimes be a delay in transitioning to adult care. Ideally you have a close working relationship with an adult program and you start to talk about transition a few years in advance.[31] If you have the luxury of having shared clinic visits with your adult partners or an overlap, that really, I think, helps make that transition. The families they still have the physicians and nurses and team that they're used to as you introduce new faces and I think it really helps optimize that handover.

Chapter 3: Understanding the Role of Parenteral Support

Dr. Turck:
Thank you, Dr. Wales. And now I'd like to switch gears a bit and focus a little more on the role of parenteral support, because, from what's been said, it sounds like it can be a heavy burden for all SBS patients, not just children. So, coming back to you Dr. Tappenden, how do you manage this disorder to help patients achieve their goals?

Dr. Tappenden:
Well, you're right, and let's not forget the burden for caregivers as well, especially those who care for children with SBS.[23,32] The burden of SBS in patients who are dependent on parenteral support is substantial.[9] Not only can long-term parenteral support increase the likelihood of life-threatening conditions like liver disease, infections that can lead to sepsis, and fluid and electrolyte imbalances,[33] but it

really reduces a patient's quality of life by causing any number of physical problems, like pain and fatigue as well as social and psychosocial distress.[9,10] But what's more, the high cost associated with parenteral support can lead to a loss of income, insurance challenges, large out-of-pocket expenses and hospital costs.[9,22,34,35] The good news is that we have treatment options to help manage symptoms of SBS and reduce the frequency and volume of parenteral support. Conventional medications for symptoms relief mainly include anti-secretory and anti-motility drugs to control gastric hypersecretion and high-volume diarrhea,[36] and then there are glutamine and intestinotrophic agents that are used to help patients with SBS reduce the need for parenteral support.[36]

Dr. Turck:
And we're almost out of time, Dr. Tappenden, but before we close, what would you like to leave with our audience today?

Dr. Tappenden:
Thank you. You know, I just want to leave with emphasizing the fact that short bowel syndrome really is a rare disease,[8,37] but chronic and life-threatening for these patients.[37] So, when it is present, we need to be able to identify and diagnose it and get these patients to appropriate management, which in 2023 is not just the fundamentals of providing intravenous nutrition and fluid support and anti-symptomatic agents, but really trying to optimize that oral nutrition that they get with an individualized diet and then the potential use of intestinotrophic agents to stimulate intestinal adaptation and rehab that gut as much as possible, because what that's going to allow is for us to reduce the dependency that these patients have on parenteral support, and thereby reduce the long-term complications that they have.[22,26] Let's not forget that SBS is a chronic condition and it may require ongoing management, but fortunately, we've found ways to manage SBS symptoms, reduce dependence on parenteral support, and improve quality of life.[36] And that's where dialing in those long-term management goals of intestinal rehab are really critical moving forward.

Dr. Wales:
I agree. I just want to reiterate that I think it's imperative that these patients are managed by a multidisciplinary program.[16,17]

Dr. Turck:
Those are great practical takeaways to consider as we end today's program. And I want to thank my guests Dr. Paul Wales and Dr. Kelly Tappenden for helping us better understand the SBS burden and the symptom management strategies for these patients.

Dr. Wales, Dr. Tappenden, it was great speaking with you both today.

Dr. Wales:
Thank you. I was happy to be involved. Thank you very much.

Dr. Tappenden:
Thanks from me also to you and our listeners.

Announcer Close
This program was sponsored by Takeda. If you missed any part of this discussion, visit ReachMD.com/GI Insights. This is ReachMD. Be part of the knowledge.

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