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INVITED REVIEW

Advocating for a patient- and family centered care approach to management of short bowel syndrome

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Abstract

Patient- and family centered care (PFCC) is a model of providing healthcare that incorporates the preferences, needs, and values of the patient and their family and is built on a solid partnership between the healthcare team and patient/family. This partnership is critical in short bowel syndrome (SBS) management since the condition is rare, chronic, involves a heterogenous population, and calls for a personalized approach to care. Institutions can facilitate the practice of PFCC by supporting a teamwork approach to care, which, in the case of SBS, ideally involves a comprehensive intestinal rehabilitation program consisting of qualified healthcare practitioners who are supported with the necessary resources and budget. Clinicians can engage in a range of processes to center patients and families in the management of SBS, including fostering whole-person care, building partnerships with patients and families, cultivating communication, and providing information effectively. Empowering patients to self-manage important aspects of their condition is an important component of PFCC and can enhance coping to chronic disease. Therapy nonadherence represents a breakdown in the PFCC approach to care, especially when nonadherence is sustained, and the healthcare provider is intentionally misled. An individualized approach to care that incorporates patient/family priorities should ultimately enhance therapy adherence. Lastly, patients/families should play a central role in determining meaningful outcomes as it relates to PFCC and shaping the research that affects them. This review highlights needs and priorities of patients with SBS and their families and suggests ways to address gaps in existing care to improve outcomes.

KEYWORDS
caregivers, nutrition support, parenteral nutrition, patient compliance, patient-centered care, short bowel syndrome

INTRODUCTION

Healthcare practitioners who are trained to manage patients with short bowel syndrome (SBS) possess a certain expertise. They understand the pathophysiology of the condition and the evidence that supports the use of available therapies, as well as having the experiential insight to guide practice. Patients with SBS also develop unique and valuable expertise on the condition. The experience of living with the condition makes them intimately familiar with what it takes to incorporate associated therapeutic measures into their daily lives. This also applies to family members of patients with SBS, especially when the patient is a child. A solid partnership between the patient, family, and practitioner ideally brings together the expertise from both perspectives to achieve optimal outcomes. This partnership is critical in SBS management since the condition is rare, chronic, and involves a heterogeneous population that requires a personalized approach to care, particularly given that there is often more than a single reasonable course available.

Patient- and family centered care (PFCC) is a model of providing care in which a partnership is forged between the care team and the patient/family to provide healthcare that respects and responds to the preferences, needs, and values of the individual patient throughout all healthcare decisions.[1] Both patient-centered care (PCC) and PFCC are terms used to describe this partnership. PFCC explicitly acknowledges the vital role family members play in the patient's care, which is particularly important in the pediatric setting.[2] Leading national organizations, including the Institute of Medicine[1] and the Picker Institute,[2] have advocated for the adoption of a healthcare approach that centers on the patient and their family as an essential foundation for quality and patient safety. Although PFCC continues to evolve, healthcare organizations as well as individual healthcare providers remain challenged by effective implementation of this care model across the continuum of care.

Several conceptual frameworks of PCC and PFCC have been proposed.[3–7] One comprehensive review of the literature identified common themes and classified them into three domains: structure, process, and outcome.[8] The *structure* domain encompasses the necessary materials, healthcare resources, and organizational characteristics needed at a system level to provide a patient-centered model of care. Interactions between patients, families, and healthcare providers fall into the *process* domain, whereas the value achieved from the implementation of a PCC or PFCC approach is classified as *outcomes*. This framework can serve as a roadmap to guide healthcare systems and practitioners in providing PFCC.[2,8,9] The PCC approach has previously been applied to the management of patients with other chronic conditions, including chronic kidney disease[10] and interstitial lung disease.[9]

This review applies the domains of structure, process, and outcome to advocate for a PFCC approach in SBS management. It highlights needs and priorities of patients with SBS and their families and suggests ways to address gaps in existing care to improve outcomes. Notably, the writing of this article reflects multiple perspectives by bringing together the voices of a healthcare practitioner who manages adult patients with SBS as part of an intestinal failure (IF) program based in the United States, and two US-based researchers with personal connections to the condition (an adult patient with SBS since birth and the parent of a child with SBS since infancy).

ORGANIZATIONAL STRUCTURE TO SUPPORT THE PROVISION OF PFCC

The implementation of a PFCC model in the management of patients with SBS, whose medical backgrounds are often highly complex, requires a coordinated effort within the entire healthcare system and across the continuum of care. An organizational structure that promotes PFCC processes is the foundation of an environment in which clinicians, patients, and family members can collaborate to achieve optimal outcomes. Institutions must lay the groundwork for this foundation by bringing the voices and values of patients and families into the development of organizational structures. It starts by incorporating patient-directed core values into its vision, mission, and common language. Core values include recognition of dignity and mutual respect, building a caring and trusting relationship, and addressing power differentials between patient/family and healthcare practitioner. Another core value is nurturing a culture of transparency by openly and candidly providing information and sharing knowledge with patients and their families.

Institutions can facilitate the practice of PFCC by supporting a teamwork approach to care. Healthcare teams should consist of qualified and skilled healthcare providers who are supported with the necessary resources and budget to practice this model of care. One of the primary barriers to promoting and practicing PFCC in the United States is the reimbursement structure of the healthcare system. Requirements set by healthcare institutions to meet established thresholds for billable services can significantly impact the ability to

spend sufficient time with patients and their family. Quality of care as perceived by patients is often given less priority. Multiple international professional nutrition support societies have endorsed the involvement of an interdisciplinary team of qualified clinicians as critical to reducing complications and improving quality of life in patients with chronic IF.[11–14] Such teams should ideally include a gastroenterologist, surgeon, advanced practitioner (nurse practitioner and/or physician assistant), nurse, dietitian, and pharmacist.[11] The reality is that nutrition support teams have greatly evolved over the past several decades; in the US context, this is primarily due to changes in funding.[15] Functions conducted by these core team members have adapted over time by transitioning responsibilities among team members that were traditionally discipline specific. Nutrition support functions may also be delegated to other healthcare teams within an institution or outside the institution, such as with the home infusion provider. Regardless of structure, an advanced level of training and experience is required of team members to provide competent care. In addition to dedicating the resources to create and maintain a competent team, healthcare organizations should provide the comprehensive services required to manage patients with SBS, such as access to a mental health provider, social worker, and interventional radiology services with expertise in vascular access. Healthcare providers who do not practice within a comprehensive IF program should be encouraged to refer patients with SBS to one of these programs when available, especially for those patients requiring home parenteral nutrition (HPN) therapy.

PROCESSES UNDERLYING THE PROVISION OF PFCC

Clinicians can engage in a range of processes to center patients and families in the management of SBS. Select themes, including fostering whole-person care, building partnerships with patients and families, cultivating communication, and providing information effectively, are highlighted below.

Fostering whole-person care

Whole-person, or person-centered care, acknowledges the patient/family as a person first and values their context, preferences, needs, and beliefs based on accumulated knowledge over time.[8,16] Understanding a patient as a person, their context, and where they are in their journey takes time and often requires a long-term relationship that develops beyond the initial medical encounter. Whole-person care respects the physical, behavioral, and emotional needs of the patient.[8] When patients and families are appropriately involved in designing their care, they feel respected and have a voice in getting their needs met, which positively impacts healthcare outcomes.[8] In SBS management, this occurs when patients and families have a voice in determining the team of specialists and nonspecialists involved in their care. Since it is typical for providers to change throughout the life span of a patient with SBS because of the patient moving, provider leaving, changes in patient needs, or other patient milestone life transitions, the role of a comprehensive IF program becomes even more important in providing oversight to ensure overall medical needs are being addressed.

There are person-centered tools available to help address gaps in the management of SBS in the context of whole-person care. Winkler and colleagues developed an HPN Patient-Reported Outcome Questionnaire (PROQ) to recognize the experience of living with HPN, the various disease influences that can come with requiring HPN, and the extent to which individuals cope to find normalcy.[17] The HPN-PROQ provides a tool for healthcare providers to engage in more meaningful and personalized conversations with patients and their families. Another option that may be helpful in addressing whole-person care is referral to a palliative care service to assist with symptom control and providing relief of stress associated with chronic disease. Given the complexities of SBS management, palliative care teams may offer a valuable perspective by engaging meaningful conversations with patients, caregivers, supportive loved ones, and clinicians that focus on the delivery of whole-person care.

Patients and families as partners

At the core of the PFCC approach is the partnership between clinicians and patients/families. This partnership model assumes that (1) patients with chronic disease and their families are responsible for a significant amount of self-care and (2) experiential knowledge and competencies develop through living with and caring for the condition that complements the scientific knowledge of clinicians.[18] The model further stipulates that healthcare decisions should draw on both kinds of knowledge and consider the patient's and family's life goals and preferences. Valuing the knowledge that patients and families bring to the table in medical encounters is essential in building meaningful clinician-patient/family partnerships.

Recognizing patient and family expertise

The development of patient/family expertise is especially relevant in the context of rare disease, which is characterized by a low prevalence and an associated lack of widespread clinician knowledge about and experience with the condition.[19–21] This phenomenon may further be compounded by heterogeneity in terms of disease presentation; in the case of SBS, no patient is alike, with specialized nutrition needs, symptoms, and SBS-related comorbidities dependent on their unique gastrointestinal anatomy and functional status of remaining intestine.[22] In this context, patients or family caregivers may bear an especially large responsibility for developing experiential (and in many cases technical) knowledge related to their condition.[23,24] They have spent countless hours caring for complex needs at home, including preparing and administering HPN or tube feeding, keeping to a medication schedule, and monitoring input and output. They are asked to constantly watch for symptoms associated with development of a central line–associated bloodstream infection or other illness. Through their SBS journey, they have developed considerable knowledge, processes, and intuition related to the condition.

Unfortunately, patient/family knowledge and experience have traditionally been an untapped resource, despite their potential to benefit the quality of care.[25] Considering patients and their families as equal partners in the provision of care allows clinicians to draw on this invaluable experiential and learned knowledge. Recognizing, incorporating, and supporting the acquisition of knowledge is essential to increasing patient/family self-efficacy and may lead to innovations in disease management.[26] This partnership extends to the decision-making process when patient and family perspectives, preferences, goals, and ideas are actively solicited and considered.

A personal reflection from the mother of a 4-year-old child living with SBS since birth due to gastroschisis:

My son with SBS will depend on HPN indefinitely. At home, we have developed diligent central line–care processes and have kept the number of people who manipulate his central line to an absolute minimum. This strict routine helps us feel more in control and in charge of the ever-present risk of central line infections, which we have fortunately been able to avoid thus far. In the inpatient setting, however, this sense of control is replaced with feelings of anxiety and disempowerment. Suddenly, we no longer have a say over who accesses our child's central line and the protocols they use. Our only option is to carefully observe every time his line is accessed and speak up when necessary. In an already emotionally laden context, this places an additional burden on me as a parent to constantly monitor and advocate without stepping on the toes of those who are ultimately responsible for my son's care.

At one of his hospitalizations, my son's nurse pulled me aside shortly after our arrival on the pediatric floor. She told me:

"Please tell me how you care for your child's central line at home. You are clearly doing a great job, and we want to do things exactly how you do them to make you feel comfortable."

This simple interaction was incredibly powerful. It changed the dynamic from one that was system- or clinician-centered to one that centered my son, and me as his parent. By being recognized and drawn into the team of professionals caring for my son, I felt empowered. Not only had this nurse recognized me as an important source of information and expertise, but she had also established communication that was patient- and family-centered. I felt ***heard****.*

Shared decision-making

When patients and families are recognized as partners, they are naturally drawn into the shared decision-making process. Encouraging and supporting active participation by patients and families in decision-making at the level they choose is one of the core concepts of PFCC.[27] In shared decision-making, decisions are made collaboratively between patient/family and the clinician(s) based on discussions of available options, which include consideration of available evidence, potential benefits and harm, and patient/family preferences.[28] Listening to patient and family perceptions of benefit, harm, gain, or loss in regards to their journey when discussing available options is an important component of shared decision-making. Patient and family involvement in decision-making can be guided by three tasks. The task of *information seeking* involves a process in which the clinician(s) and the patient/family seek and share their respective viewpoints; for the patient, these may include their knowledge about the condition and their needs, questions, preferences, and

concerns. This initial task is followed by *deliberation*, in which differences in viewpoints are reconciled before a *final decision supported by available evidence* is made.[29] Importantly, the level of desired involvement in making final decisions will vary across patients/families and over time. Thus, it is involvement in the process more so than sharing in making the final decision that matters to patients and their families.[29] In rare disease, shared decision-making presents opportunities to increase healthcare system efficiency, foster innovation, and improve patient satisfaction.[24,26]

Enabling patient self-management

Fostering the ability to self-manage is an extension of patient and family involvement in the decision-making process.[8] Self-management can be facilitated by comprehensive patient and family education. Techniques for self-monitoring and recognition of potential complications of SBS and associated therapies should be incorporated into training at the time of SBS diagnosis or initiation of therapy and routinely reinforced. Self-monitoring guidelines should incorporate specific thresholds on when and how to contact their healthcare providers. Training should be provided by qualified clinicians[11] and tailored to meet the assessed needs, abilities, and readiness of the patient, caregiver, and all supportive family members. It should start prior to hospital discharge for patients discharged home with HPN as a new therapy. These patients will typically depend on skilled nursing visits from a home health agency initially but should be encouraged to achieve independence with self-management as soon as possible.[30] For example, selection of a tunneled central venous catheter in a location accessible by the patient when the long-term need for HPN is anticipated can assist with the transition to self-care by allowing patients the ability to self-administer HPN and perform their own site care and catheter dressing change. In addition, adult patients who are no longer homebound should be encouraged to increase autonomy by transitioning from home health visits to an outpatient laboratory facility for required laboratory monitoring.

Patient/family self-management includes finding information, coping with symptoms and the effects of treatment, and seeking appropriate care when indicated.[29] These skills are linked with improved health-related quality of life, self-reported health status, clinical outcomes, and a reduction in healthcare utilization.[29,31] In fostering patient and family ability to self-manage, clinicians should ensure that care plans can be accessed by patients/families.[8] Clinicians can also support patient autonomy by providing access to resources, advocating for the patient and their family, and helping them navigate the healthcare system.[29] Perhaps, particularly so in the rare disease context, clinicians should support patients in developing the motivation and confidence to use the knowledge and skills they have to take effective control over living with their disease.[19] Empowering patients to self-manage important aspects of their condition builds on the partnership model and can enhance coping to chronic disease.

Engaging with concerns, emotions, and uncertainties

To successfully build a partnership with patients and their families, communication during medical interactions must become patient- and family centered.[8] In the context of a complex and rare disease like SBS, patients and families are likely to have many questions and concerns and develop their own understanding of their condition. They may have experienced trauma associated with the medical setting, fear, isolation, and uncertainty about what the future holds. Engaging with patients and families about their experiences, beliefs, values, and concerns is an opportunity to build trust and rapport and strengthen the clinician-patient/family relationship.

Authentic, active listening

Actively listening is a core characteristic of PFCC.[8,29] Listening on the part of the clinician is essential to both patients and caregivers. It can serve multiple purposes: to support the gathering of data needed to make a diagnosis, to serve as a "healing or therapeutic agent," and to foster and strengthen the patient/family-clinician relationship.[32] This type of listening can be accomplished by asking patients and families what items (concerns, questions, views, understandings, priorities) they want to discuss and actively listening to their responses.[8] Importantly, beyond engaging in authentic and active listening during clinical encounters, clinicians must hear the patient/family and show it with eye contact and engaged attention. Clinicians should work to understand where the patient is coming from, how they perceive their condition, and acknowledge patient and caregiver perspectives, values, and context.[29] A narrative medicine approach, which prompts patients to share stories of their health through guided conversations and personal writing, may present an insightful way for clinicians to engage more deeply with the underlying meanings of patient and family situations and experiences.[33] It may

help patients and clinicians understand their illness and journey in the context of their life.

Responding to emotions

Receiving the diagnosis of—and living with—a serious, chronic, and rare condition and its associated symptoms and therapies can result in a range of emotions, including anger, resentment, fear, sadness, and anxiety. Mental health–related issues, including social isolation, anxiety, and depression, have been reported for patients with SBS and their caregivers.[34,35] Recognizing and responding to the emotional states of patients and family members can be accomplished by legitimizing (ie, "it's only natural to feel that...") validating (ie, "this is an anxiety-provoking time for you") verbally expressing empathy (ie, "this is making you worried and sad, is that right?"), and offering tangible help (ie, "I think I can help by..."). [29] This affective communication can, in turn, foster a sense of being understood and a healing clinician-patient/family relationship.

Managing uncertainty

Illness-related uncertainty occurs when an individual perceives their illness, treatment, or recovery as ambiguous, complex, and unpredictable.[36] This type of uncertainty is associated with poorer adjustment and coping with the condition and higher rates of depression.[37,38] However, it is notable that, for some patients or caregivers, the maintenance of uncertainty may be a protective strategy that allows them to make space for hope.[29] The rare disease context is especially likely to be characterized by uncertainty for clinicians and patients/families.[39] For patients and families affected by SBS, uncertainty may relate to long-term outcomes and available treatment paradigms and is compounded by the complexity and rarity of the condition. In this context, a goal is not only to attempt to reduce patient/caregiver uncertainty but also to promote uncertainty management by acknowledging and openly discussing uncertainties.[8,31]

A personal reflection from a 33-year-old living with SBS since birth due to atresia:

At the age of 10, I was sitting in the pediatric endocrinologist outpatient clinic. He was just one of the many specialists I had to see regularly. It was my first time to see him. He walked in, sat on the exam table, put my chart to his side, and did not open it. He instead leaned forward toward ME, not my Dad sitting next to me, made eye-contact with me, and said: "What is your goal? What do you want to get out of this, and how can I help you get there?" Unfortunately, I did not have an answer. I was 10. I was shocked. So much so I remember this interaction today 23 years later. He cared. This doctor wanted to meet me where I was. He knew this was not the only time I was going to see him. This is a chronic disease. This is a chronic relationship. He was willing to empower me. He wanted to show and tell me he cared.

Now, I can answer his question. My goal is to LIVE with IVs and/or tubes, not to SURVIVE without them. My goal is to be grateful for where I am and appreciate my chronic disease and the journey I have had achieving, life, health, and milestones while living with the disease. Not take it away. Goals that have evolved drastically since I was a child.

Providing information effectively

The exchange of information is another important communication process linked to the provision of PFCC. Rather than being a one-way flow of information from clinicians to the patient/family, effective information exchange is a bidirectional process that includes the assessment of information needs, understanding patient/family health beliefs, communicating clinical information, and providing guidance and resources.[29]

Assessing information needs

Clinicians should engage with patients/family members to help identify their information needs and to personalize the content and communication approach. This should be a continuous process as information needs are likely to change over time and patients vary in their health literacy and ability to ask questions when they arise.[29] Clinicians should encourage patients and families to ask questions, offer opportunities to reflect on information gaps, and offer ways to follow up if questions arise later. For example, asking "what matters to you?" or "what does a good day look like for you?" and using perceptual skills to know when to ask "what is the matter?" are effective ways to engage patients to speak openly about their needs.[40]

Understanding patient/family health beliefs

An important part of effective information exchange is uncovering how patients/families understand their illness, known as "illness representations."[29] The dimensions of illness representations describe beliefs and expectations about identity, duration of illness, impact on life, causality, and control.[29] Illness representations are how patients/families make sense of their disease and can influence coping, adherence, and health outcomes. Thus, learning about patient/family illness representations can help clinicians personalize care, address misperceptions, and provide patients/families with a sense of being understood and validated. The dimensions of illness representations are influenced by factors including cultural background, social determinants of health, media, Internet, family, friends, and coworkers.[29] Incongruence between clinician and patient/family illness representations can lead to misunderstandings and poor quality care.[29] It is therefore important not to assume patients/families have the same understanding and interpretation of clinical information as clinicians, regardless of health literacy. Instead, seeing the patient beyond their disease and understanding how patients and families process and cope with their condition is an important undertaking that influences the effectiveness of the information exchange.[29]

Communication of clinical information

Providing information to patients is linked with greater patient satisfaction, makes it easier for patients/families to participate in medical interactions, and helps them adapt and cope.[29] Effective PFCC of clinical information should be thorough, accurate, unbiased, timely, and delivered in a way the patient/family understands based on their information needs and underlying health knowledge.[8,27] A personalized style of communication should be tailored to the needs of the patient/family and incorporate appropriate teaching methods and written materials (ie, audio recordings, resources that explain complexities of the disease and the care plan, guidance on where to find additional information, and how to identify trusted resources).[29] To help clinicians manage and tailor the delivery of clinical information to patients/families, the following skills are suggested: (1) use everyday language to provide clear explanations; (2) repeat and summarize; (3) ask patients to restate information in their own words to maximize comprehension; (4) encourage patients and family members to ask questions; (5) engage in active listening; (6) allow adequate time for discussion; (7) be honest and giving realistic hope.[29]

Providing guidance and access to resources

Clinicians should integrate clinical recommendations with instruction, advocacy, and support.[29] Regardless of a patient and family's health literacy and self-empowerment, the guidance they require may change over time based on where they are in their SBS journey. For example, an SBS patient restarting tube feeding or HPN after a hiatus may need clear instructions but delivered with respect to the patient's level of experience and underlying knowledge gaps. To encourage self-autonomy but not overwhelm, it is important to regularly ask patients and families if they feel competent with required skills and have the resources to follow through with a mutually agreed upon plan. Instructions can include suggested therapy schedules that accommodate the patient's lifestyle, pain management techniques, nonnegotiables in care management, and the interpretation of laboratory results. It is also important to provide specific self-monitoring parameters that include when, who, and how to contact care team members when complications occur.

Additional strategies to foster autonomy and empowerment is guidance on advocacy and finding support through community. Early in the disease process, clinicians can direct patients to educational programs that help guide and inform self-management techniques and foster safe SBS community connections.[29] This includes patient support organizations, advocacy toolkits, educational webinars, books, podcasts, peer-to-peer support, patient and family community-driven resources illustrating coping with day-to-day life, and advice on how to effectively find additional resources online from trusted resources across the Internet and social media platforms.

CHALLENGES RELATED TO THERAPY NONADHERENCE

Therapy nonadherence is a complex challenge that often confronts clinicians who manage patients with SBS. Patients with SBS are often asked to take multiple medications and dietary supplements with complex dosing schedules and comply with challenging oral fluid/dietary restrictions. HPN and/or home intravenous (IV) fluid support may be required, which is time- and labor-intensive, involves meticulous care of a central venous catheter, and requires commitment to a detailed monitoring regimen. Therapy nonadherence and dietary indiscretion from time to time should not come as a surprise. But the situation becomes a problem if therapy nonadherence is sustained, particularly if the healthcare

provider is intentionally misled. This represents a breakdown in the core values of mutual trust and transparency. Other examples of nonadherence include improper catheter care, failure to perform requested laboratory monitoring, failure to return phone calls, and failure to attend scheduled clinic visits. This clinical dilemma was addressed by a large US-based HPN program and describes guidelines to help identify nonadherence of HPN therapy and explores the legal and ethical implications when addressing HPN nonadherence.[41] Ultimately, consistent nonadherence to therapy represents a breakdown in the patient-centered approach to care.[42]

Reasons for therapy nonadherence are generally more complicated than unintentional forgetfulness. A patient-centered model of care that promotes adherence to a prescribed regimen should include three basic principles: the patient/caregiver must understand what to do, want to do it, and have the means to carry out their intentions.[43] Helping patients and their families understand the reasons for recommendations and acknowledging concerns are fundamental to developing mutually agreeable treatment plans. For patients with SBS who are often prescribed complex therapies, clinicians should provide proper education and detailed verbal and written instructions to help alleviate anxiety related to performing these tasks and promote adherence. Taking measures to simplify the regimen whenever possible, such as eliminating medications that offer minimal benefit and reducing the frequency of laboratory monitoring when stabilized, can help alleviate some of the treatment burden.

Perhaps most importantly for therapy adherence, the prescribed regimen should be integrated with existing patient lifestyle and habits (eg, accommodating an HPN cycled infusion with work/school schedule or determining a pump-assisted vs gravity infusion for administration of an IV fluid bag). Patient motivation to comply with therapies related to SBS can be enhanced by the recognition of patient priorities. For example, patients who require frequent hospitalization for complications associated with SBS may be highly motivated to adhere to regimens that keep them home and out of the hospital. Patients who want to spend time with family but are afraid to leave home because of uncontrolled stool output may be motivated to adhere to antidiarrheal regimens that effectively decrease stool output.

Although all healthcare team members should communicate a consistent message regarding treatment plans, the reality is that many patients with SBS and their families are likely to receive conflicting information from their providers regarding their care. This certainly complicates therapy adherence. We need to prepare patients and their families that this may occur and empower them to speak up and seek clarification when they have questions. Conversely, many healthcare providers are likely to encounter patients who misrepresent their medical condition. Even though it may not be realistic to achieve full therapy adherence among all patients, an individualized approach to care that incorporates patient and family priorities will ultimately enhance adherence to therapy.

PFCC OUTCOME AND RESEARCH

Outcomes derived from a patient- and family centered approach to care need to be measured in terms of what is meaningful and valuable to the individual patient and family. Patients and families should play a central role in determining meaningful outcomes and shaping the research that affects them. Patient-centered outcomes research (PCOR) fundamentally assumes that patients (and caregivers) have important and unique perspectives that can contribute to and improve research.[44] Community engagement in research can be conceptualized as a "a process of inclusive participation that supports mutual respect of values, strategies, and actions for authentic partnership" with a focus on issues that affect the wellbeing of the community of interest.[45] This engagement can take a number of forms, including involvement in defining research questions and selecting outcomes, the provision of input into a study's conception and design, coauthoring a research study, and assistance with the dissemination of study findings.[46] Importantly, stakeholder engagement must take place early enough in the decision-making process to be meaningful, and compensation should be considered to allow for patient/caregiver involvement.[46]

Because research is shaped by the worldview of those who conduct it, PCOR allows space for the worldviews of patients and caregivers, whose experiences with and perspectives about the condition of interest fundamentally differ from those of clinicians and researchers.[44] Meaningful patient and caregiver engagement in research may also help address some of the common barriers to conducting human-subjects research, including distrust of the research enterprise by communities of interest, study enrollment in sufficient numbers,[47] and poor dissemination and adoption of research findings in said communities. In the rare disease setting, an additional barrier to research includes low disease prevalence and geographic dispersion of patients. In this context, patients and caregivers develop a level of expertise and can thus provide a nuanced and deep understanding of their condition that can help to inform and advance research.[48] Meaningful engagement of

community stakeholders can thus help to identify community priorities and relevant research questions as well as patient-centered outcomes in rare disease.[49]

The mandate for PCOR has gained momentum in recent years, particularly with the establishment of the Patient-Centered Outcomes Research Institute in 2010.[46] In the setting of SBS, a patient/family centered approach to research was performed to investigate quality of life and family management for children with SBS and their families.[50] The authors argue that their community-driven research on quality of life contributes nuance to the narrative of quality of life for children with SBS and their families. Future research related to SBS should explicitly engage patients and caregivers as partners in knowledge production at every stage of the research process.

CONCLUSION

The shift to a PFCC approach can improve the delivery of healthcare in patients with SBS. Prioritizing this approach calls for a systematic review of existing structural, process, and outcome domains. Clinicians should work within their healthcare organization to seek the necessary resources for managing patients with SBS, including the creation and maintenance of a qualified interdisciplinary care team. PFCC techniques should be used to determine the values and preferences of patients and their families and promote shared decision-making and partnership. Meaningful engagement of patients and families in research is needed to foster innovation and ensure alignment with SBS community priorities. All relevant stakeholders—patients, their families, healthcare providers, and healthcare systems—must bring their perspectives, knowledge, and experience to the table and work together to improve health quality through PFCC.

AUTHOR CONTRIBUTIONS

Vanessa Kumpf, Marie Neumann, and Swapna Kakani equally contributed to the conception and design of the manuscript. All authors critically revised the article, agree to be fully accountable for ensuring the integrity and accuracy of the work, and for reading and approving the final article.

CONFLICT OF INTEREST STATEMENT

Author conflict of interest disclosures are as follows: Vanessa Kumpf is a Consultant for Takeda Pharmaceuticals, Fresenius Kabi, Zealand Pharma. Marie Neumann: none. Swapna Kakani is a Speaker and Consultant for Takeda Pharmaceuticals, VectivBio.

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