Seminars in Pediatric Surgery 29 (2020) 150990

Contents lists available at ScienceDirect

Seminars in Pediatric Surgery

journal homepage: www.elsevier.com/locate/sempedsurg

Importance of education and the role of the patient and family in the care of anorectal malformations

Greg Ryan[a,*], Stephanie Vyrostek[b], Dalia Aminoff[c], Kristina Booth[d], Sarah Driesbach[e], Meghan Fisher[f], Julie Gerberick[g], Michel Haanen[h], Chelsea Mullins[i], Lori Parker[j], Nicole Schwarzer[k]

[a] ARM Adult, Founder of ONE in 5000 Foundation
[b] Program Manager, Center for Colorectal and Pelvic Reconstruction, Nationwide Children's Hospital, Columbus, OH, USA
[c] AIMAR, Italy Patient and Parent Association
[d] APN Program Coordinator, Center for Colorectal and Pelvic Reconstruction, Nationwide Children's Hospital, Columbus, OH, USA
[e] Center for Colorectal and Pelvic Reconstruction, Nationwide Children's Hospital, Columbus, OH, USA
[f] Nurse Clinician, Center for Colorectal and Pelvic Reconstruction, Nationwide Children's Hospital, Columbus, OH, USA
[g] Nurse Clinician, Center for Colorectal and Pelvic Reconstruction, Nationwide Children's Hospital, Columbus, OH, USA
[h] ARM Adult – VA, Netherlands Patient and Parent Association, Netherlands
[i] ARM Adult, Pull-Thru Network – USA Patient and Parent Association, USA
[j] Pull-Thru Network, USA Patient and Parent Association, USA
[k] SOMA, Germany Patient and Parent Association, Germany

A B S T R A C T

During this early part of the 21st century online technology has prompted many major advances in medical care. In this section we argue that this is particularly evident in the treatment and care of patients born with Anorectal Malformation (ARM) and Hirschsprung's Disease (HD). Our stories show that anyone born with these complex colorectal conditions in the 20th century was destined to a life of isolation and stigma. Here we explore the lack of understanding and recognition of the psychological effects on children and families which has characterised this period. We show that advances in clinical practice has been supported by developing social media platforms.

There has been a rapid creation of online support groups for patients and families which has enabled survivors' greater access to patient and parent organizations across the globe and thereby stimulated a sense of belonging and solidarity. Online technology and social media platforms have also opened up the opportunity for pediatric medical professionals to provide a greater level of patient education.

There is no doubt families have become much more aware of the complexities of ARM & HD and achieved greater comfort and understanding of their needs. We have generated "lightbulb moments" for pediatric providers with adult ARM & HD patients, enabling them to share their lived experiences in a therapeutic exchange. In the past survivors felt they were abandoned by the adult healthcare system. We are seeing evidence-based research of major psychosocial issues experienced by adult patients and, as a result, improved understanding of how to treat ARM & HD survivors across their whole of life journey.

The winds of change continue to direct our cohorts to a mature approach based on improving levels of interactive communication and education. We argue that this maturity has mostly been facilitated by the use of online technology and the ensuing collaboration between providers and patient and parent organizations.

Introduction

Various teaching strategies and methods of delivery for patient education can be found in the medical literature. Included in the strategies are computer-based information specific to their own situation, DVDs, audiotapes of the teaching performed for recall of verbal teaching, tailor- written materials at an appropriate reading level, verbal instructions in conjunction with another teaching method, demonstration of a skill with teach-back approach, and illustrations with text description. The methods of delivery should be patient specific and involve multiple teaching strategies with sensitivity to cultural issues.

Anorectal malformations (ARM)

Case 1

A 12-year-old patient who underwent a previous repair of an anorectal malformation, has both urinary and fecal incontinence. He is followed by a urologist who recommends that the patient

* Correspoding author.
E-mail address: greg.ryan@onein5000.org (G. Ryan).

https://doi.org/10.1016/j.sempedsurg.2020.150990
1055-8586/

starts on clean intermittent catheterization (CIC) and the colorectal team recommends that he be started on rectal enemas.

What do you think is the best option for families to understand what treatment plan their child would be exposed to?

A Send them an e-mail with the instructions
B Have a conversation in a multidisciplinary facility including a specialized urology and colorectal nursing team
C Call them on the phone to explain how they will be doing CIC and enemas
D Send them a booklet with a step-by-step explanation
E All the above could be utilized

Answer: E

Patients who have colorectal diagnoses often require tailored education as these diagnoses are often unfamiliar to parents and even medical professionals. These patients and families require general education about the diagnoses and long-term care outcomes including the importance of routine bowel and urological management, and gynaecological needs. In addition, patients with colorectal diagnoses may require education about various medical treatments that families will perform in the home setting. These may include treatments such as anal dilations, rectal enemas, rectal irrigations, antegrade enemas, and clean intermittent catheterization.

Every day in hospitals across the world children are seen with a new diagnosis of an anorectal malformation (ARM). The most common question families have is "Will my child poop normally?" This unknown haunts parents whose babies are born "different." It is the job of the provider to help put the information known about the child into clear explanations of how their malformation leads them to continence. This is how the Anorectal Malformation Continence Predictor Index was created:

** *Continence predictor*** *JPEG Attached*

Every patient seen should get one of these as a handout for visual learners and have it reviewed verbally with their provider for auditory learners. The provider should review each section and how it pertains to the specifics of each child's case. This continence predictor helps guide the conversation about what parents can expect as the child grows and will help them understand the patient's potential for continence. The discussion with the family should also focus on the malformation the child has and how that falls into the scoring system of this tool.

The Anorectal Malformation Continence Predictor Index handout is a good tool to utilize for each patient to review the specifics of their child's anorectal malformation, spine, and sacrum. This allows the families to follow along on a handout during the discussion and they can visually see where the child falls in the spectrum of all children.

The sections discussed in the index are: ARM type, quality of spine, and quality of sacrum. These sections are broken down into three subcategories. These subcategories are then tabulated using a scoring system. The scoring system adds all three sections together to then predict the child's potential for continence.

ARM type

The ARM type is the largest of the three sections as there are many different variations of malformations. Based on the malformation a score is given. Those with good potential for continence (perineal fistula, anal stenosis, rectal atresia, rectovestibular fistula, rectobulbar fistula, and ARM without fistula) are awarded 1 point. Fair potential (cloaca with <3cm common channel, rectoprostatic fistula, and rectovaginal fistula) are awarded 2 points. The last subcategory of poor potential for continence (rectobladderneck fistula, cloaca >3cm common channel, and cloacal exstrophy) are given 3 points. The proper diagnosis of the original malformation is vital in identifying where the child falls on the spectrum and can impact the overall score of the index, and therefore impact the kind of discussion you are having with the family.

Spine

Spinal anomalies can impact the potential for continence as well. Many children with anorectal malformations have spinal anomalies, so it is very important to screen all ARM patients As with ARM type, there is a spectrum of spinal abnormalities that can affect the child's potential for continence. The spine that appears "normal" (termination of conus at L1-L2, and normal filum appearance) has a good potential for continence and is awarded 1 point. Abnormally low termination of the conus (below L3) and abnormal thickening of the fatty filum have been shown to impact the potential for continence and is noted as such with 2-points. Those who have myelomeningocele have the poorest potential for continence in this group of patients and are given 3 points.

Sacrum

The sacrum is evaluated based on the sacral ratio and any anomalies associated with the formation of the sacral bones. This section is also rated using a scoring system. A sacral ratio equal to or greater than 0.7 is awarded 1 point, as that represents a normal sacrum. Ratios between 0.69 and 0.4, hemisacrum, sacral hemivertebrae, or presacral mass, is given 2 points. Finally, a sacral ratio less than 0.4 gets 3 points.

Overall score

The goal of the evaluation and interpretation of this data and scoring system is to get to a final number which is explained to family. It resembles the game of golf, the lower the number the better the potential for continence.

Once a prediction has been made, it is essential to discuss with families there are always exceptions to every rule, and many children rely on a formalized bowel management regimen, no matter what the index has predicted. Making sure to keep the family informed is critical in the process as the clinician manages their expectations of what they can expect for their child's continence.

Hirschsprung disease (HD)

Case 2

A 6-year-old patient with Hirschsprung disease is referred to your institution as a new patient due to severe constipation and obstructive symptoms. His family have never performed irrigation before, only rectal enemas with glycerin.

What is the best method to teach this family proper irrigation techniques?

A Discuss how they are going to do the irrigation at home while providing equipment as visual aids.
B Show them how to do the irrigation while the child and family are in clinic, and have them demonstrate the proper technique in front of the healthcare provider.
C Have them watch a video of how to do an irrigation
D Give them a handout with steps on how to do a proper irrigation
E All of the above

Answer: E

Hirschsprung disease (HD), like many colorectal diagnoses, requires diagnosis education and surgical intervention in a child's

early years of life, and additional treatments may need to be continued throughout their lives. For many families, this may start within the first few days of life as their child's symptoms prompt testing of the disease process. Key moments for education include at the initial diagnosis, at the time of their pull through, postoperative follow-up, and at subsequent follow-up visits. Families and patients need to understand that HD is a disease that requires long-term attention including enterocolitis prevention and bowel management for hyper- or hypomotility.

Throughout all these intervals of care, learning materials need to be written at an appropriate reading level and that discussions are held understanding that the family may have minimal to no medical knowledge. Education for families on the diagnosis itself should be presented in various forms—in person discussion, written handouts, and if possible, video education.

Education and discussion on initial testing to confirm the diagnosis (contrast enema, rectal biopsy) and how that information helps to guide treatment are vital. For patients who require an ostomy placement as their first intervention, it is crucial to provide the family with hands-on ostomy education and management. Families must also start to understand the process and medical concern regarding the heightened risk of enterocolitis. Hands-on teaching on how to perform a rectal irrigation and discussion regarding prevention could be held at the bedside or within the clinical setting. Consider written education with visual images reviewing step by step of how to perform a rectal irrigation at home. This should also include education on the common signs and symptoms of enterocolitis, and when to contact their medical team.

Between the overwhelming news that their child has Hirschsprung disease and will require surgery, it may be hard for families to absorb the information and education you are trying to deliver. While the patient and parent are in the hospital for their pull-through operation, take advantage of the opportunity to teach them key post pull through concepts like enterocolitis prevention, when to do a rectal irrigation, and what their long-term follow-up plan will include.

For those who are not diverted with a stoma or who have just had their ostomy taken down, skin care prevention education is vital. Provide handouts or written instructions on what products to use for routine prevention, mild, and severe skin breakdown. Include pictures of the products and where they can order or buy them. These patients typically require specific bowel management medications within the first month to protect their anastomosis from hard stool; it is important to review the purpose and importance of those medications.[2]

Bowel management

Case 3

A 6-year-old patient with a past surgical history of rectovestibular fistula, and a tethered cord release presents to your bowel management clinic. You've decided to enrol her in a 1-week bowel management program with rectal enemas.

How do you educate families prior and during the treatment program?

A Provide family with information regarding the program prior their clinic visit so they have basic understanding of their expectations during and after the program
B Have an informed discussion with family about their child's diagnosis and expectations based on their ARM index continence score
C Provide family with handouts and education on how to troubleshoot issues that may arise with the enema.
D All the above

Answer: D

Patient education regarding bowel management's purpose, strategies for care, and areas of concern are key to long-term success in patients with colorectal anomalies. No matter the program, oral medications versus rectal enemas/antegrade enemas, patients and their caretakers must receive education on the importance of routine bowel management. This education must start early and continue as the child grows with the goals being socially continent and eventually independent in their bowel routine.

A formal bowel management program requires dedication from the medical team, patient, and family. Within a program, there are several opportunities to provide education sessions and materials which families can utilize throughout the program and after they are home. Multiple learning formats should be utilized when educating patients and families, which include written materials, hands-on sessions, and visual aids.

A program booklet given to each family with an overview of information about the program and additional education materials provides structure to the program. This booklet serves as a resource throughout the bowel management week, as well as reference guide in the future. In this booklet, it is important to review the basics of bowel management including how continence works, why patients soil, and an outline of what their bowel management day-to-day schedule will look like. Sections on each specific program (oral medication, rectal enema/antegrade enemas) and the goals for each are helpful to include, as is a detailed report sheet for them to fill out after each treatment. This will allow families to focus on key pieces of the program which are helpful for the medical team to note when determining if a medication change is needed.

For patients and caregivers who are visual or hands-on learners, opportunities for them to hear the basic material in person should be given. This can be done as a formal bowel management talk with the patients and families in attendance. This talk not only can reiterate the basics reviewed in the booklet, but also allow for more open discussion with families and offers an opportunity to ask real-time questions. The content is best kept generic, irrespective of the diagnosis, with the goal for the family to understand the basic concepts of the program.

For many patients and families, they may be entering a new phase of treatment with rectal enemas. This can be a scary time for them as they may not be familiar with the process of a rectal enema. Create time after the talk to provide group teaching on how to give an enema. In their clinic appointment, one-on-one teaching can also be completed with individuals on how to perform an enema. Consider the importance of the teach-back method in this process. Ensure the family is able to follow the instructions and can adequately perform the treatment themselves.

With several different components of bowel management, it is helpful to have one-page handouts with pictures and visual references. For processes like giving an enema, families and patients will likely not be able to remember each step even with hands-on education. A step-by-step visual guide with associated steps written out will allow families the assurance that they are doing the treatment correctly. For patients new to laxative therapy, consider a handout with pictures of the types of oral medications used for bowel management. Many of these medications are over the counter and it can be confusing which medication to purchase. For those starting on fiber therapy, a chart indicating what types of fiber are recommended and where they can be purchased would help ensure the family gets the right product and can properly tell what dose they are giving. For programs recommending dietary changes such as following a laxative or constipating diet, a list of foods within that category, or those to avoid would provide a guide for meals.

Laxative and Constipating Diet JPEG Attached*

Surgical antegrade options

Case 4

A 10-year-old patient had a Malone appendicostomy procedure 1 month ago. His Malone catheter is still in place when he arrives to clinic.

How would you educate families on how to perform Malone catheterization before and during their post-operative clinic appointment?

A Have the surgeon or specialized nurse perform the first Malone catheterization. Require the caregiver to return demonstrate their ability to pass the catheter
B Have the family watch a video and then will be able to it on their own at home without the need for education at their clinic appointment.
C In clinic, provide family with handouts and instruction on what to do if they have trouble with their Malone flush at home
D Support to child with the use of medical play to prepare them for what to expect during the Malone catheterization

Answer: A, C & D

Patients with colorectal diagnoses and associated fecal incontinence may benefit from surgical antegrade enema options including a Malone appendicostomy or a cecostomy tube. These patients benefit from a one-on-one session with the colorectal surgeon and colorectal nurse discussing surgical options and management after the antegrade access is in place. Written handouts about these surgeries can also be utilized and include information such as the preoperative care, surgical procedure details, and postoperative care. It can also include important details about the care of the surgical site during the postoperative period and step-by-step instructions on how to give an antegrade enema.

When it is identified that the patient should start catheterizing the appendicostomy or using the cecostomy tube after surgery, education should be provided using multiple teaching strategies. These should include hands-on teaching with step-by-step instructions on how to catheterize the channel or use the cecostomy device, and how to give the antegrade enema. Written instructions should also be given to the family as a supplement to the verbal instructions provided. Return demonstration is key to identify any pitfalls and improve the patient and family confidence. If available a doll or medical model can be used to help with the return demonstration. It is also important to educate patients and families on care of catheters and enema supplies during this time.

** *Malone doll photo*** *JPEG Attached*

Urology

Case 5

A patient with a history of an anorectal malformation – a recto-prostatic fistula, as well as prior tethered cord release and neurogenic bladder is asked to start clean intermittent catheterization (CIC)?

A The provider should discuss with the patient and family the reason for starting CIC and the urological goals
B The nurse specialized in urology should have an appointment with the patient and family and provides verbal instructions on CIC
C The patient and family should be provided with an opportunity to practice CIC on a medical model
D The patient and family should watch a video on CIC
E All of the above

Answer: E

Patients with colorectal diagnoses can have associated urological anomalies or complaints. Education on associated urological anomalies should be provided to patients and families early after colorectal diagnosis is made and appropriate follow-up arranged.

Some patients with colorectal diagnoses may also require clean intermittent catheterization (CIC). This can often be scary and challenging for both the patient and the family, especially in patients who have sensation. Appropriate education and follow-up is the key to success. When implementing CIC it is important to provide education about a patient's specific urological plan and how to perform the catheterization, including an opportunity for return demonstration. It is vital that the patient and family understand why CIC is being started and the goals of catheterization. A one-on-one discussion about how to perform the catheterization, along with practice using a medical model are ideally included in the education process. Handouts with pictures and step-by-step instructions also supplement this teaching. Return demonstration is key and can help identify any challenges or troubles with catheterization. After a patient/family is taught catheterization there should be a follow-up in place. This can be a clinic visit or telephone call by the nursing team to get an update and help troubleshoot any concerns.

In select patients, surgical intervention with an appendicovesicostomy may be considered. A one-on-one discussion with the urologist about these surgical procedures is helpful to determine if this procedure would be of benefit. Written handouts can also be utilized and include information including details about the surgical procedure and the typical postoperative course. Other visual aids can also be used such as videos. Often it is helpful for both the patient and the family to see a video of another patient with an appendicovesicostomy and how the channel is catheterized. When it is time for the patient and/or family to start catheterization the appendicovesicostomy in the postoperative period education is provided to the family about catheterizations. This information is presented using multiple teaching strategies including verbal discussion, written instructions, and return demonstration with the patient and/or family catheterizing the appendicovesicostomy channel.

Education on the in-patient side of care

Education for the family/caregivers/guardians of a child born with an anorectal malformation needs to begin at birth or shortly after the malformation has been identified. This can be a very scary and difficult time for families. Most likely, they did not know that their child would be born with this malformation, and in most cases never even heard that this was something that could happen. Some families may be looking into the adoption of a child with an anorectal malformation and have many questions. The child may require extensive testing and expert assessment to identify the specific type of malformation. Diagnoses can range from mild to very complex and limiting. Families may look to social media for information and try to interact with other families who have children with the same diagnosis. Accurate information and education are crucial for best medical and social outcomes for these children and their families.

Healthcare providers need to be supportive and provide this accurate information to the family. The primary care providers may not have previously cared for a child with an anorectal malformation and need to identify resources for families. It is imperative that the family receive information and education on the diagnosis and care of their child. Families may have to travel from their homes and away from their usual coping and support systems to go to a colorectal center which can provide expert care for their child. These experts can provide much needed information and ed-

ucation for the family on the specific diagnosis and specific care needs of their child.

Once the diagnosis has been determined, families and children, if age appropriate, will need information on the necessary diagnostic and surgical procedures required. When available, hospital support systems such as child life specialists, psychology and social work should be included in this care. Initial education for families may range from medication administration to skills such as anal dilations, colostomy, vesicostomy, or vaginostomy care. This will ensure the child can stool and urinate safely and adequately until more definitive procedures can be completed.

Healthcare providers who care for families and patients with anorectal malformations need to have an understanding of the diagnosis and surgical procedures in order to anticipate the child's care needs and plan for the family's education needs. Pre-operative care may include education on bowel preparation prior to surgery, appropriate diet, and when to stop eating and drinking, lab work and antibiotics. Post-operative care may include focused assessment skills, pain management, intravenous fluids and antibiotics, surgical incision care, care of tubes and drains, stoma care, bowel management, perineal suture and skin care, importance of ambulation, diet, self-care as age appropriate, and plan for ongoing follow up. The family will need to know who to contact with questions or concerns.

Knowledge is critical for these children and their families. The family will be the child's best advocates when they return to their everyday lives surrounded by those who do not understand the diagnosis of anorectal malformation and the care needs associated with these malformations. Informed children will grow into knowledgeable teenagers and adults who are able to care for themselves and achieve their life goals and dreams.[1]

Online resources for families and burden of therapy

Burden of therapy

As the burdens accumulate some patients are overwhelmed, and the consequences are likely to be poor health-care outcomes for individual patients, increasing strain on caregivers, and rising demand and costs of health-care services. In the face of these challenges we need to better understand the resources that patients draw upon as they respond to the demands of both burdens of illness and burdens of treatment, and the ways that resources interact with health-care utilization.

Burden of Treatment Theory is oriented to understanding how capacity for action interacts with the work that stems from health care. Burden of Treatment Theory is a structural model that focuses on the work that patients and their networks do. It thus helps us understand variations in health-care utilization and adherence in different health-care settings and clinical contexts.

Case 1

As a colorectal surgeon who treats conditions such as anorectal malformations and Hirschsprung disease, what are the main points to discuss with patients and family from a family support point of view?

A Quality of life
B Burden of therapy
C Support forums
D Annual family conferences
E All the above

Answer: E

Learning points

As an adult or parent of a child affected by ARM, the writers understand the burden of therapy as a concept and a reality. The phrase describes the demands placed on many ill people suffering from chronic health problems. The burden is all aspects of self or childcare handled in private and sometimes in secret. The burden is experienced as a complex tapestry of surgery, monitoring, management, emotional self-protection, and consequential social, emotional, and economic effects. For most of the individuals affected with ARM, this burden will last a lifetime.

Research on the topic of burden of treatment is ongoing. There is still a significant need for validated tools to help assess patients' level of burden in order for caregivers to weigh the risks and benefits of the treatment. Consequences of a significant burden of treatment include poor adherence to treatment regimens and poor overall health and well-being in addition to the socioeconomic consequences mentioned above. Providers for patients who have anorectal malformations and other lifelong colorectal diagnoses should be cognizant of the burden of treatment they are imposing and modify treatment plans so that patients are able to have optimal quality of life. Colorectal diagnoses do not exist in a vacuum, and theoretical "best care" often differs from the care that provides the best overall quality of life for the patient.

Because of historical and cultural perceptions, the anorectal area is viewed as a private domain overlayed with a sense of shame and stigma. Ignorance and fear are attached to the topic of malfunctions, adding an extra layer of anxiety to patients and families. Learning how to deal with physical and mental realities across a lifetime has become the primary task for family and sufferers. Learning how to relate and relay emotional and psychological understandings is even more complex in an area which is seen to be on the dark side of human experience. In contrast, a child born with a congenital heart condition is not treated as a "silent" matter and the awareness, empathy, and understanding in the wider community provide such families with a sense of comfort and support that is not available to ARM families. This difference in perception between a heart defect and an anorectal defect begins the burden of therapy for parents and families. At birth, the parents are faced with the decision of "who to tell" and "what to tell." Many families find it difficult to share information about their child's anorectal and/or urogenital defects with close family, let alone friends. Thus, begins the early stages of isolation. In addition, parents, mothers particularly, wonder if they did something to cause this defect to occur, adding to their burden of therapy.

When a child is born with ARM, it is almost always the first time those parents have heard the words "imperforate anus." These parents are told that their child will require surgery, or a series of surgeries, to correct the anorectal malformation. The parents share this information with family and friends, who then incorrectly assume that the child is "fixed" after these surgeries. It's generally after these surgeries that the family learns of "bowel management" and "incontinence" and the therapies required to keep their child "clean." Friends and family don't understand why the child still requires diapers past the "normal" toilet-training years. This is when it is vital for families dealing with the challenges of ARM to find other families living with the same challenges.

Patient and parent organizations

The global patient and parent organizations play a crucial and influential role within the ARM community. The emotional sustenance they offer ARM and HD survivors and their families is significant, including the importance of not feeling alone and the knowledge of feeling comfortable in discussing issues in a safe and trusting environment. We feel it is important that these agencies are recognized with the following contributions from long term and influential patient organizations from across the world.

AIMAR – Italy (www.aimar.eu)

"Aimar – Italian Association for Anorectal Malformations is a national non-profit organization formed by and for families of children born with anorectal malformations (imperforate anus, cloaca, cloacal exstrophy, VATER/VACTERL Associations, urogenital sinus etc..), colorectal disease and any associated defects.

At present we gather around 850 members all over Italy out of them 300 are adult patients. Through AIMAR, members find support among other families with similar issues and are connected based on geographic areas, thus avoiding isolation. AIMAR is trying to help families and their children working in close cooperation with surgeons, physicians, nurses and health care specialists involved in the management of anorectal malformation and associated defects".

SoMA e.V. – Germany & Austria (www.soma-ev.de)

"SoMA e.V. – the German Association for Anorectal Malformation is a non-profit organization founded in 1989 by families of children born with anorectal malformations (ARM). Today SoMA supports also patients with cloacal exstrophy, or Hirschsprungś disease, as well as Vacterl patients. Plus, there is our sub-group SoMA Austria – for parents and patients in Austria.

Currently, SoMA Germany has 1,100 members, 380 of which are adult patients, and 100 are supporting members. Becoming a member of SoMA means that one doesn't have to feel alone any longer. At the beginning of this process, parents get in touch with other parents, which then leads to patients directly contact other patients with the same or similar problems and experiences".

Association Anusatresie (VA) – Netherlands (www.anusatresie.nl)

"The Association Anusatresie (VA) was established in 1993 and has nearly 400 members in the Netherlands and Belgium. One of the main aims of the VA is to represent the interests of members by advocating for and promoting the interests of its members with care providers, health insurers and the government.

The association has extensive medical, technical and psychosocial information available for those directly involved (patients, parents, care providers, health insurers). We organize meetings aimed at the mutual exchange of experiences to share contact with fellow sufferers and the creation of solidarity, and have a close link to other patient organizations at home and abroad".

Pull-Thru Network – USA (www.pullthrunetwork.org)

"Pull-thru Network (PTN) is a 501(c)3 non-profit support organization for parents, children, teens and adults who are living with the challenges of congenital anorectal, colorectal, or urogenital disorders and the associated diagnoses. Members of the Pull-thru Network live across the United States and around the world. They represent a wide variety of initial diagnoses and related disorders. PTN also maintains a large database of professional members, including doctors, nurses, and therapists who are interested in the care of these affected individuals. In addition, Pull-thru Network provides support and advocacy for its members. Member support has been central to PTN's mission since its establishment in 1988. Support is provided through web-based forums, including a Yahoo list-serv and Facebook public and private groups, as well as their in-person events".

ONE in 5000 Foundation – Australia & Global(www.onein5000foundation.org)

"The ONE in 5000 Foundation was established in 2017 and is fully registered with the Australian Charities and Not-for-profits Commission. The purpose of our organization is to provide a global resource for the children, adolescents, adults, parents, families, and medical professionals associated within the Anorectal Malformation (ARM) community. Our organization has a very active and influential online social media presence (Facebook, Instagram, Twitter and Instagram), as well as a comprehensive website. We are passionate about promoting awareness, understanding and support within the ARM community, as well as educating the general public. To achieve this objective, we are committed to the following four outcomes, Awareness; Information; Medical; Support, which we call our A.I.M.S.".

Each organization has their own specific vision which can be accessed through their respective websites, but to summarize what they offer members please refer to the following:

- Provide information, support and advocacy for members
- Publish newsletters (printed and online)
- Collaborate with surgeons, nurses, and health care specialists
- Provide psychosocial support and information
- Provide sexual health (ARM adults and adolescents)
- Share personal stories (ARM adults, adolescents and parents of ARM children)
- Conduct seminars and conferences
- Transition of care information and support for adolescents
- Promote the latest medical advancements
- Update scientific research
- Host social media platforms and website

There are other established ARM patient organizations in their respective countries which all are also represented on Facebook, under the following names:

- Panhellenic Association for individuals and parents of children with congenital Anorectal Malformation – (Greece)
- NFA - Norsk foerning for analatresi – (Norway)
- ONE in 5000 Support Groups – (Australia; United Kingdom; Canada; France; New Zealand; Philippines; Hong Kong; Bangladesh and The Balkans)
- AH-Potilaat ry – (Finland)
- AEMAREH - Asociación Española de Malformaciòn Anorrectal y Enfermedad de Hirschsprung (Spain)
- Max's Trust – (United Kingdom)
- Vivencia de vida de ostomizados con ano imperforado – (South America: Argentina, Chile, Mexico, Paraguay, Peru, Venezuela)
- Bebes Con ano imperforado (malformacion congenita) – (Colombia)

Initial discussions have taken place in regard to establishing an association where patient organizations can come together in a sign of unity for the worldwide ARM community.

Social media

The advent of online social media (Facebook, Twitter, Instagram, etc.) has changed the lives of ARM patients and families in an overwhelmingly positive way. We have been able to reach out across the globe to the few individuals who share our physical symptoms and emotional needs. These informal networks have enabled us to undertake a level of collective emotional and practical self-care that previously did not exist. For example, almost universally the daily task of performing anal dilatations by parents is an incredibly emotional responsibility, both from the thought of causing pain to the child and the pervasive fear of the moral cloud that hangs over such an intrusive procedure. Being able to make human contact with someone else undergoing the same isolating experience has been hugely important. Connecting with other patients and families who are living with a colorectal diagnosis often provides an immediate sense of relief. Simply connecting with a community who has had similar experiences and struggles helps normalize the diagnosis and helps alleviate the sense of extreme isolation associated with these complex problems.

However, not all online information is accurate or suitably general and, in some cases, may be detrimental to individuals and families. There is much anecdotal evidence of increased anxiety and stress being caused by a bombardment of information, especially at the early stages of treatment. Furthermore, the subtle differences in each person's needs may not be examined and another person's anecdotal examples may be useless or in fact harmful.

From a provider's perspective, incorrect information presented on social media platforms presents a unique challenge to patient care. Considering that many patients and families with colorectal diagnoses have sought care from multiple providers and may have had unpleasant or traumatic relationships with past providers, it is understandably difficult to gain trust and "debunk" some of the misconceptions presented in online communities. Patients often have a strong sense of trust in information they are getting from within their own community, and providers should be aware of this and provide empathetic guidance toward evidence-based treatment while still respecting the patient's past experiences and feelings of trepidation.

Navigating social media is notoriously difficult and in the case of health management, the stakes are very high. The caregiver ideally needs to provide a direct or online pathway that is sensitive to the individuals' specific diagnosis. Additional resources aimed at providing emotional support must directly help deal with the burden of therapy that families and individuals undertake across their lifetime.

Across the world the perception is that surgery "fixes" the medical issue and ongoing selfcare defines the emotional outcome. In truth, for most ARM-related problems the issues will be lifelong, and the surgery is a maintenance rather than curative activity. The advent of social media support groups has created support and also a different dialogue. Both the surgical profession and the ARM community of families and individuals are now looking at a four-strand strategy that ties continually improving medical intervention with greater community awareness, clear and accurate information provision, and social/emotional support services. The innovative partnership arrangements between major hospitals and family support networks in both the United States and Australia are particularly encouraging in this regard.

Transition to adulthood

An interesting dyad in the treatment of children and adults has been obvious for many years but is rarely discussed. Pediatric surgical care has made huge advances and many young lives have been saved and improved as techniques advanced. Sadly, once adulthood is reached it has been expected that the individual must find his or her own way. Due to the lack of transitional care into adulthood, if someone had not made a connection with another ARM patient or family during their time under pediatric care, there is only an extremely slim chance of meeting a supportive other. Adding the ARM issues into the mix of finding a supportive workplace environment, building and maintaining an understanding intimate relationship while maybe dealing with sexual function issues and incontinence has been a massive mental health challenge for these affected adults.

It is widely acknowledged now by the medical profession that the mental health well-being of ARM children in the past has been inadequately serviced. With the incredibly invasive procedures and intrusive examinations a child must endure, and the stigma attached to having "bowel accidents," be it at home, school, or in sporting endeavours, the burden this imparts on a patient's quality of life from a mental health perspective is highly significant. Lack of resources for our relatively rare congenital issues means that there is no blame that can be apportioned to the medical profession for these omissions.

However, the gap has now become obvious to most professionals and others from the ARM community. Later as that child moves to adulthood the patient logically must leave the pediatric care system. Due to the absence of adult or transitional care programs for the overwhelming majority of ARM patients worldwide until very recently, there has been no real quantifiable information which has highlighted this crisis in the broad community.

Continence Predictor Index

1. ARM type –	
Rectal perineal fistula	
Rectal stenosis	
Rectal atresia	
Recto vestibular fistula	
Imperforate anus without fistula	
Recto bulbar fistula	
Cloaca <3 cm common channel	
Recto prostatic fistula	
Recto vaginal fistula	
Recto bladder neck fistula	
Cloaca >3 cm common channel	
Covered exstrophy	
Cloacal exstrophy	
2. Spine –	
Termination (end) of the conus	
Normal	
Abnormally low termination (>L2)	
Myelomeningocele	
Yes	
No	
Filum appearance	
Normal	
Abnormal - fatty thickening	
Lipoma of Cord	
3. Sacrum –	
Sacral Ratio	
Great than 0.7	
Between 0.4 and 0.69	
Less than 0.4	
Hemisacrum	
Presacral mass	
Sacral hemivertebra	
Spinal hemivertebra	

Fig. 1. Continence predictor index.

Mental and financial costs arising from this gap are huge. The coalitions that are forming across the globe to address this are demanding a more integrated and systematic approach to all aspects of the care. The gulf between medical intervention and social/emotional/psychological support is beginning to be addressed. Nevertheless, we see many holes yet to be filled.

One of the most momentous and life-changing issues faced by ARM adults is the lack of understanding of the implications of being able to have children. In most cases this is not addressed satisfactorily (if at all) whilst in pediatric care. This is understandable in one sense, but nonetheless an important issue that must be tackled during adolescence by medical professionals and our support community. There are numerous examples of female ARM adults who have endured great pain and heartache in their endeavours of becoming pregnant and sustaining that pregnancy due to the com-

LAXATIVE versus CONSTIPATING FOODS

Laxative foods help to loosen bowel movements allowing stool to move through the bowel faster. These foods can be helpful in patients who have hypomotility or a slower moving colon. Constipating foods assist to control watery stools and help to slow down the bowel.

Food Group	Laxative Foods	Constipating Foods
Milk	High-fat dairy products	All milk products allowed, but limit to 16 oz. total per day
Vegetables	All vegetables – especially raw	Vegetable juice without pulp, well cooked vegetables , green beans, spinach, pumpkin, eggplant, potatoes without skin, asparagus, beets, carrots
Fruits	Fruit, fruit juice with pulp, canned pineapple, prunes, dried fruit, jam, marmalade, dried fruits	Applesauce, apples (without skin), banana, melon
Starch, Grains	Whole-grain or seeded breads, whole grain pasta, brown rice, oatmeal, bran cereal, whole-grain cereal	Bread, crackers, cereals made from refined flours, pasta or noodles made from white flour, white rice, pretzels, white potatoes (without skin), dry cereal
Meat, Seafood, Legumes	Beans, fried or greasy meats, cold cuts, hot dogs, meat substitutes	Baked/broiled/grilled meats, poultry or fish, lean deli meats, eggs
Fats, Oils	Butter, margarine, oils, fried foods	Very limited amounts of all oils, margarine, butter, mayonnaise
Sweets	Chocolate (especially dark). Sugar, marshmallows, angel food cake	Sugar-free gelatin, popsicles, jelly, or syrup, rice-milk ice cream
Beverages	Water, Gatorade, Kool-Aid, Soda	Water, sugar-free Gatorade, sugar free Crystal Light, sugar free Kool-Aid, Pedialyte

Fig. 2. Constipating and Laxative Foods.

plexity of their ARM. The lack of knowledge of ARM in the adult gynaecological profession continues to exacerbate the already difficult situation.

There have been cases where the female has been told that ARM plays no part on childbirth which has resulted in extremely painful births and major ramifications for the person's ARM issues. These issues could have been avoided with a basic understanding of an ARM female patient. Similarly, it has often been found that the female ARM patient was unable to have children due to the complexity of their ARM but had not been made aware of this devastating news until later in their adult life. For male patients there have been analogous issues regarding those who have associated urological and genital issues which are distressing. The need for reproductive health to be discussed in adolescence must become mandatory in the ARM medical community.

For practically every ARM patient in the past, once they are old enough to be ineligible for the care of the pediatric system, they feel abandoned by the medical profession. The main problem is that ARM is viewed primarily as a "pediatric condition" by the adult medical profession. This means that the expertise or experience to deal with such a complex condition is simply not available in the adult health-care system. We have many anecdotal tales of ARM community members presenting at medical clinics or hospitals and finding that the doctors had no knowledge of the issue. Even more disturbing has been the misdiagnoses and inappropriate medical service as a result of this ignorance.

When there is little general awareness and no transition to an adult colorectal specialist available, the patients must rely on their local general practitioners for "maintenance." When those doctors aren't able to cater to the needs of the ARM patient due to its complexity and rarity, the feeling of abandonment and isolation is emphasized. It is now widely acknowledged that the result of this lack of transitional care has manifested itself into a major crisis regarding the mental health of many ARM patients. The only salvation some have been able to receive is by finding solidarity and information from others with similar issues.

For adult ARM community members that informal help has most often emerged through social media communication. The growth of online support networks worldwide over the last five years, especially a private social media support group on Facebook called "Adults living with IA/ARM," is providing this salvation. This is a "secret" group which ensures people's privacy because of the continued immense shame and embarrassment the majority of adults feel who have to live with the condition.

The feeling people have when they "find" others and become a part of this support group can be incredibly overwhelming. Because most have never met anyone else with the condition in their lifetime, the emotional effect can be substantial. The overriding

feeling is that of relief that "I'm not alone anymore." There is also an immense sense of validation when members are able to share experiences openly in a totally safe environment where others automatically understand context and detail. There are countless examples of people expressing that the day they found this group was "the day my life changed." We have made empathetic friends; instigated and supported each other's projects and begun to feel that the burden of this therapy can be shared and made lighter.

An example of the positive effects social media has had on ARM adults, is the story of one of our co-authors, Chelsea Mullins, 34 years old from USA, who was born with Cloaca:

"Five years ago, I searched Imperforate Anus on "Google'. I hadn't had many opportunities to meet others born with the same condition, other than a few chance meetings up to that time in my life. I never felt the sense of "community" that could understood my day to day struggles. That "Google" search changed everything!

The Pull-Thru Network popped up and my world was forever changed. This led me to not only meet others in our community personally, but also discover a network that created support and friendships that I will cherish forever.

Not only did this provide me with the support I needed, it showed that my journey could help other families. It opened up the opportunity for me to be elected as a Pull-Thru Network board of director, as well as being involved in the creation of the "Adults living with IA/ARM" Facebook support group.

Most importantly this has given me an ability to speak to adults and families from all across the world, allowing them to know that we are all in this together".

The positive effect has been twofold. First, it has provided much needed clinical information to adults who had not been exposed to new medical advancements such as the Malone Procedure and other bowel management regimes. In addition, the shared advice on new colorectal centres that will care for adult patients has given the opportunity for ARM adults to seek life-changing medical procedures. Second, the groups have provided adults with new levels of support for the mental health issues with great consequence to their quality of life.

It has become undeniable due to the overriding evidence of adults' shared experiences that depression, anxiety, panic attacks, and body image issues have been evident in the overwhelming majority of the adult ARM community. The collective experience is beginning to mitigate some of the worst aspects of this alienation.

Over the years, the surgical options for ARM have greatly advanced in both technique and success rates. In addition, the ARM Continence Index has been a wonderful initiative for ARM children and families that provide a good indicator of future continence. Despite these medical advances, the greatest issue for adolescents and adults born with ARM continues to be the mental health consequences of what they experienced physically in childhood. There is a lack of mental health support in the formative years and a lack of transitional care programs to adulthood which contribute largely to this mental health issue.

Fortunately, we live in a newly enlightened era in which this burden is better understood by the medical fraternity. Consequently, we see this burden as being shared in a mutually interactive way between anorectal medical specialists and the families and informal caregivers who make up the ARM community. The isolation previously felt by many sufferers in managing their own therapies for both physical and mental problems is being acknowledged and recent innovations, studies, and activities have thankfully begun to see the tide turn.

Fig. 3. Malone cath doll.

In summary the burden of therapy needs to be shared better. The good news is that many of us in the ARM community have begun to step away from our embarrassment and shame and speak up for ourselves and our children. We are offering services to each other. We are talking more clearly with our medical professionals. Our words are being heard and changes are occurring. Our call is for the burden to be identified and for a partnership of the medical fraternity and the ARM community to be reinforced and financially supported.

We must ensure that the congenital issues are identified and discussed. Privacy must be protected but medical services need to be enhanced, communities must be encouraged, and better information should be provided by respectable and authenticated sources. Much has been learned by the ARM community over the past 50 years, yet there is much still to be done. It is necessary for the entire colorectal medical system, both pediatric and adult care, to acknowledge and integrate the distinct information for these therapies for the benefit of the current and future generations (Figs. 1–3).

References

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