A PATIENT'S VIEW : WHERE IS THE EXIT?

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In de *erVAring* van maart 2008 is een verkorte versie opgenomen van de presentatie die Muriël Schipper hield tijdens de Workshop Anorectal Malformation 2008 in het UMCN St. Radboud te Nijmegen.
De integrale tekst van de presentatie is hieronder te lezen. Een Nederlandse vertaling van de tekst wordt gepubliceerd in de *erVAring* van december 2008.

To live with an anorectal malformation

Good afternoon ladies and gentlemen. My name is Muriel Schipper. I am an adult patient born with an imperforate anus and a board member of the Dutch patient organization Vereniging Anusatresie. You can see our logo on the screen. The rectangular shape symbolizes the large intestine and the dot of the question mark is where the anus should be. Our motto is: Where is the exit? Of course, this literally refers to babies born with an imperforate anus, but to me, it also refers to the main theme of my presentation: the transition of care.

It has long been thought that surgery correcting congenital defects was the ending point for treatment. Surgery done, problem solved. Today, we know better. We now realize, as patients grow older, that surgery is just a beginning. After surgery, a life long process starts of adapting, learning to live with an anorectal malformation. Of accepting and dealing with the limitations that may be associated with the defect. We patients appreciate all research and treatments and surgeries that we undergo as babies. They help us to survive, enable us to grow up and even grow old.
Until the late eighties, emphasis has been on cure and care, and this made the development of advanced surgical techniques, such as Dr. Pena's PSARP surgery, possible. Clinical trials have been focused on understanding how the defect develops in the womb, on perfection of surgical techniques and hospital treatments and on genetics.

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For a long time, it has been unclear what happened as the patient matured into adulthood and outgrew the pediatric surgeon. Not only regarding treatment and hospital care, but also the psychosocial aspects of a ARM.
Parents and patients organized themselves in patient organizations such as: Soma in Germany, Aimar in Italy, the Pullthru Network in the USA, even patient organizations in Israel and Taiwan. They did this out of a need for sharing information on basic day to day care and psychosocial support from peers. Gradually mature patients joined as members and with them came the need for new kinds of information and support. This also changed the role of the patient organization. Today, the Dutch patient organization deals with many themes in patient treatment and care. While the emphasis historically was on the young patient, we find ourselves challenged with the questions that are raised by the senior patients. Today, we are dealing with two main themes.

- Transition of care
- Psycho social aspects of living with ARM

But first I need to reflect on the role of patient organizations. What is their reason for existence and why should you (pediatric surgeons) have anything to do with them?

The role of patient organizations

The role of patient organizations can be divided into 3 dimensions. These are: education and provision of information, patient-to-patient support, and advocacy. In first instance, education and information is aimed at patients (and their parents) themselves. As a patient organization we would like to see that parents are referred to us by the children's hospitals. In an ideal world this would be standard practice. We believe that the importance of patient support groups to patients and their parents is underestimated by the medical community. Pediatric surgeons should be aware that while they are busy treating the newborn child, psychosocial support to the parents is also very necessary. For example, the period in which parents have to dilate their children is often experienced as very traumatic by the parents. They feel they are hurting their baby's and the dilations require actions that in another context could send people to prison. Parents experience feelings of guilt and shame and are afraid their children will remember it for the rest of their lives.

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A support group is the ideal place for parents to get support from people who have had the same experiences. Older patients might reassure the parents that they have no recollection from the dilation period. Support groups bring patients and parents together to share information and give and receive support.
Support groups can also benefit the pediatric surgeons. On our symposia and gatherings, education about treatments can be given efficiently to dozens of people at once. And they are the ideal place to recruit people to participate in research programs. Therefore, the more members a patient organization has, the better. In The Netherlands, the VA collaborates with researchers. We are engaged in research design, and assist in interpreting the results.

Two great examples of this collaboration are the NAHO research and the research 'An adult life with ARM' which is taking place right now. The aim of the NAHO (National Anal Atresia and Hirschsprung's Disease Research) project was to examine changes in quality of life, disease-specific functioning, and psychosocial competences of children and adolescents (aged 8–16) with anorectal malformations or Hirschsprung's disease and to identify predictors of change in quality of life. These results can be read in the thesis of Esther Hartman. Mr. Nic Looijaard, an adult patient with ARM was one of the initiators of this research and was involved in all stages.

Our patient organization is witness to the need of educating the general public. Parents for instance, experience difficulties getting their child with ARM admitted on primary schools. Senior patients experience difficulties on the work floor because of misconceptions of their employer. Gathering and developing sources of information for both the general public as concerned parties such as schools, employers and house doctors is a key task for patient organizations as well as the medical community. And we do not want to limit ourselves to education only. We also aim for advocacy directed towards the government and medical insurance companies, where we act as spokesman for our members.

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Transition of care

But we are also advocates for our cause towards the medical community. In this regard I want to talk about what's currently the most important theme within our organization: the transition of care. I will start with an example:
I once met an older woman who had just recently learned she had been born with ARM. She had experienced problems with incontinence all her life, but had no idea what the reason was. You may find this unbelievable, but she is not the only senior patient who learned of ARM at such a late moment in life. And finding out is too often a matter of coincidence. An article in a magazine for instance. Or by pure chance meeting a fellow patient. We must bear in mind that patient-doctor relationships have just recently changed from top-down care with little communication, to patient-doctor collaboration with extensive communication. Until late in the seventies, people didn't talk about defects below the belt. And the medical community thought it wiser not to confuse the patient and his parents with too much information. Patient files of these senior patients are not always available anymore. In the case of the woman I just described, the hospital that had performed her surgery had been bombed during the Second World War.

We estimate that there are in total 2000 patients born with ARM in the Netherlands. Of these 2000, we estimate that about 900 are 25 years and older. We don't know how many patients of 50 years and older there are. The patients in this last group come from a generation where you don't hang out your dirty laundry as we say in Holland (and we may take this literally in this case). We wonder where they go when they experience medical or emotional problems due to ARM. Many of my fellow senior patients have come to a point where daily irrigation routines don't work anymore. Not much is known about the options these patients may have. Most of them now have a stoma. And although they are very content with their stoma, I dread the day that a stoma is my only alternative.

Most of the younger patients I know (until their mid thirties), know what they have, and are still seeing a pediatrician or pediatric surgeon for their check-ups and treatments. A silly situation of course, but we have no clue which specialist to see after we turn 18. Should we see a gastro-enterologist? Technically, ARM is not necessarily related to colon disorders. And of course, a children's hospital has its

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advantages. Where adult patient care is highly specialized around specific disorders like little islands, pediatric care is (in the Netherlands) often organized in a multi-disciplinary approach which ensures the involvement of physiotherapists, psychologists, and any other –ist that may be required. This difference in 'culture' if you will makes transition from pediatric care to adult patient care a scary step for most patients. To be honest, I would prefer to remain under the care of the pediatric team that treated me since birth.

The good news is that, at least in The Netherlands, the pediatric community is well aware of the transition problem. We have the advanced treatment of cystic fibrosis to thank for that. Patients with Cystic Fibrosis are getting older and pediatricians saw themselves faced with an ever growing patient population. The adult patients didn't leave their ward, because they had nowhere else to go. This initiated transition projects in which adolescent patients are gradually introduced to lung specialists and adult patient care facilities. This has inspired the starting up of projects that relate to other disorders such as Crohn's Disease. The discussion about transition has started in relation to ARM as well. Pilot projects were started in Rotterdam by Dr. Langemeijer and in Amsterdam by Dr. Kuijper. Other initiatives for adult care include the 'poop'ward in Nijmegen and bowel management programs in other hospitals.

However, in our country, there is no consensus of what transition should look like. Should patients be passed on to adult care facilities? Should patients remain under the care of their pediatric team? The medical community should not be afraid to ask for input from the patient organizations. We, on our part, will keep harassing you on this subject until we are satisfied with the solution, I promise!

Psychosocial consequences of living with ARM

I have already talked about the importance of psychological support to parents of patients born with ARM. But this is important for all patients born with ARM. Until my seventeenth, I was living a fairly normal life. I paid as little attention to my condition as possible. I had no enema- or irrigation routine, and I had gotten very handy in hiding my accidents which occurred very frequently. In short, I was able to pretend I was a normal kid. But seventeen is an age where you start to figure out

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who you are and what your place is in life. And I got to a point where I could not deny it to myself any longer. I was forced to acknowledge that I was incontinent and that I would be for the rest of my life. Until then I had been able to hide this, not just from my social surroundings, but from myself as well. All of a sudden this didn't work anymore. I stopped eating; reasoning that whatever you don't put in your mouth doesn't need to come out either.

To cut a long story short, it took me many years to make peace with myself and to no longer feel betrayed by my body. And I must admit that I still have some work to do. Over the years I have come to realize that most of my limitations are inside my own head. Whenever I talked to a lay person about ARM or my incontinence, I never ever had any negative reaction. People felt sorry for me, or were surprised because they had never suspected anything, but no one ever turned me away. But you should all know that my experience with telling other people is quite unique. I know many fellow patients who did experience negative reactions from their social surroundings.

According to the NAHO research I mentioned earlier, psychosocial functioning was the most important predictor of the quality of life of patients with ARM or HD, whereas faecal incontinence and constipation had almost no effect on patient QoL. One of the conclusions of the research was that "QoL of children and adolescents with ARM or HD might be improved by enhancing self-esteem and promoting a more positive school attitude. In general, to improve and maintain an optimal level of children and adolescents' QoL, it is important to direct treatment both to reducing symptoms and improving psychosocial competencies".

To summarize

Both the medical community and the patients should discuss what should happen when a patient turns eighteen. We must reach consensus on how transition of care should be dealt with and support transition projects.

ARM becomes more an issue as a patient grows older; physically and psychosocially. Transition wards should take this into account and organize care not only based on treatment of disorders but also on psychosocial support.

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My last appeal to the medical community is: make use of the patient organizations. We are not here for patients alone, but we can help you get access to our patient population for research and education. And make us one of your sparring partners in research design and in discussions on transition and other themes that are important.

I am not the only board member of a patient organization to walk around here. On the screen behind me you can see our sister organizations. Many of them are present on this conference. Please feel free to approach us.

Thank you.

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