

International Camps for Children with Lymphedema and Lymphatic Anomalies: When Education Links with Psychosocial Research

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Abstract

Background: Lymphedema in children and adolescents is a rare and chronic condition. The management of their lymphedema is mainly driven by the adaptation of treatments used in adults. The aim of our study was to explore the needs and challenges the children and adolescents face during their management with the aim of finding ways to satisfy these needs and organize an hospital-based centre accordingly with an educational program.

Methods and Results: Patients and their families were given the opportunity to meet other patients, their families and professionals during social activities organised annually and during two international camps. They were invited to take part in different semi structured focus groups and interviews. All patients and families described a long journey and relief when the diagnosis was obtained followed by the shock of being told that it was a chronic condition. Meeting other children with the condition was a relief. The impact of lymphedema on body shape and genitals was a source of distress. Rejection of the compression was part of journey. Lymphedema management had an impact on all the family members including siblings. Parents were responsible for their child self-management in young children which was described as demanding. It was followed by a complex transition phase to self-management. The impact was not the same according to the age the lymphedema had started.

Conclusion: Acceptance and management of lymphedema is complex and invades many aspects of families' life. Self-management is demanding. Based on these results, the management of lymphedema in the centre included meeting other children and families and an educational program based on individual needs and follow-up.

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Keywords: lymphedema, children, health care organization, education, self-management, self-efficacy, psychosocial research

Introduction

LYMPHEDEMA IS A RARE chronic condition in children. Its clinical outcome differs considerably between genetic backgrounds and etiologies, with cancer and filariasis the acquired cause in most cases over the world. The recent discovery of causal genetic mutations has led to the identification of many different conditions grouped together under the umbrella term of “lymphedema.” However, this breakthrough has been translated into specific management for only a minority of children with lymphedema. For example, close follow-up of hematologic disorders can be done in cases of *GATA2* mutation underlying Emberger syndrome associated with lymphedema.^{1,2} The identification of *VEGFR3*

mutations as the main cause in the Milroy lymphedema has allowed to identify it is limited to the lower legs and below the knee.³ Such understanding is important for determining prognosis, for parental acceptance of the condition, and for the personalization of treatment. However, mutations are still identified in only a minority of patients with lymphedema.^{4,5}

The management of lymphedema has never been specifically designed for children, and pediatric treatment is instead driven essentially by the adaptation of treatments used in adults with the disease in specialized centers and children are followed locally.⁶

Only a few teams worldwide have both the knowledge and skills to treat lymphedema in children. Most of these teams are also specialized in vascular malformations, and

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lymphedema cases are the most rarely referred to these centers. Conversely, the development of skills in lymphedema management mostly occurs in the context of the treatment of adults. As a result, children with lymphedema are doubly disadvantaged.

In this context, we decided to perform systematic observations of the natural history of lymphedema in all children referred to us, focusing on the needs of the children and their families, to make it possible to adapt and personalize management based on a holistic assessment. This process was the starting point in the development of the specific organization of a center dedicated to the management of lymphedema in children.

We report here the key observations that drove our decisions relating to the specific organization of the vascular medicine department for children with lymphedema. Education and research projects were developed with the families during 3-day overlapping weekends each year. The subsequent organization of camps, bringing together patients, families, and health care professionals from different countries, cultures, and languages, has facilitated international collaborative research and helped to raise awareness.

Methods

Organization of the Montpellier Center for Rare Vascular and Lymphatic Diseases

IRB approval was not needed for this study. The Department of Vascular Medicine is located at Saint Eloi Hospital, the university hospital in Montpellier. The center for rare disease is dedicated to the diagnosis and management of rare vascular and lymphatic diseases and has been recognized by the French National Plan for Rare Diseases since 2014, which is linked to the European Network for Rare Disease European Reference Network for Rare Vascular Disease (VASCERN). Both children and adults with rare vascular and lymphatic diseases are treated at this center. For children, pediatricians are involved in diagnosis and management, working in direct collaboration with the specialists at the center. This center was recently recognized as a center of excellence by Lymphatic Education & Research Network (LERN).

In 2009, it was decided to develop a specific organization of the center for the management of children and young adults with lymphedema. All children, adolescents, and young adults were initially seen by a physician from the center, at a consultation. About 60% of these patients came from outside the regional health care area. Their medical assessment and management were initiated, and they were then asked to join other children and their families for a follow-up assessment on a Friday, followed by 1 or 2 days dedicated to the sharing of knowledge and trying to determine the needs of these patients and their families, with the aim of finding ways to satisfy these needs and thus optimize their ability to self-manage. Patients and their families were thus given the opportunity to meet other patients and their families, to attend talks given by experts, and to take part in different focus groups, with the aim of constructing a specific education program based on their needs. These actions were launched in 2007 and 2008, and the education program called "Montpellier Week-ends" began in 2009 and has been repeated annually ever since. The education program was fully

recognized as an educational tool by health care agencies in 2014 and is partly funded by the regional health care agency.

Interviews and focus groups

Consent was obtained from participants, and the focus groups were always semistructured and conducted with different groups of patients, parents, and professionals. Different aspects of the patients' condition, their everyday lives and that of their families were explored, together with management issues. The participants were first introduced to each other and allowed to get to know the other members of the group. The initial questions were followed by discussions, including descriptions of facts, and individual experiences, and also beliefs and types of behavior relating to the questions raised. The questions dealt with in the general discussion responded to these prior exchanges and were tailored to each individual group. Finally, the main points raised were summarized. Participants were asked whether their thoughts had been summed up adequately and whether anything had been missed. Audio recordings were made of all focus group sessions, which lasted between 1 and 1.5 hours. If participants became distressed during the focus groups, they were offered time out and asked whether they wanted to continue. Emotional support and counseling were available to all participants after each event. These focus groups were conducted each year from 2007 to 2011 in Montpellier, and then in 2012 during the first international camp in Montpellier, which was organized at the same time as the International Lymphoedema Framework (ILF) conference in the town, and in 2017 in Candia Canavese, during the second ILF international camp. The participants were children of a suitable age who were willing to take part, and the groups were organized according to age, and the participation of other family members attending, including siblings. The health care professionals responsible for patient care were invited to attend the weekends and the camps.

Results

Identification of the difficulties and needs of patients and their families

The journey to diagnosis. As for many rare diseases, the road to obtaining a diagnosis appeared to be very long. Furthermore, obtaining a diagnosis was associated with the difficulty of knowing that there is no cure for the disease. Patients and their families often said that relief at diagnosis was followed by the shock of being told that the condition could not be cured. Some patients described rejection by health care professionals and various types of inappropriate behavior. Finding the right health care professional was a relief for all of them.

Psychosocial impact on children and adolescents

All the focus groups indicated that lymphedema affected the lives of children and their families, with the greatest difficulties occurring during adolescence. The impact of lymphedema on body shape was a source of distress and isolation for girls quite early on, before adolescence. The involvement of the genitals was a source of distress that was very often difficult to address, and affected girls and boys equally. Meeting other children with the same condition was a relief

for children and young adults. The feeling of being different, and of having to deal with their differences alone, was attenuated by discovering that there were other children with the same condition. Some of the children were afraid of complications, mostly of infections. Some of the older patients expressed worries about their future. Different children reacted differently to the use of compression. Some found it difficult to cope with, whereas others were happy because it concealed their lymphedema.

Psychosocial impact on parents and family life

Parents were very anxious before diagnosis. The most common difficulties faced were dealing with the uncertainty of health care professionals, administering daily, sometimes time-consuming, treatment at home, and trying to find much-needed care resources close to home. For many families, particularly in cases of mild localized lymphedema, the goal was to achieve “normality” or to cope with the condition in a positive manner, to allow their children to embrace their future fully. Some parents said that they saw no improvement with the treatment, but that they nevertheless followed the recommendations. The fear of complications, such as cellulitis, sometimes led to restrictions on the daily life of the children, decreasing their confidence. All the siblings questioned said that lymphedema had had an impact on them and on their role within the family.

Impact of self-management

Lymphedema management had such a large impact on the children and on family life that we decided to conduct a specific study of self-management and self-efficacy during the 2017 Camp. The limited studies performed until then had focused on the impact of quality of life but had not attempted to compare or contrast the experiences of parents from different countries with different health care systems and treatment regimens. Moreover, recommendations for effective self-management in lymphedema have not been established for either adults or children, and this remains a topic of intense international debate.

Organization of the center for children with lymphedema

Following identification of the difficulties and needs of the patients and their families, we modified the organization of the center.

Medical pathway

The first appointment was retained, as in the existing care pathway, but the time reserved for each patient was increased from 30 minutes to 1 hour. Both parents were asked to attend. Great efforts were made to ensure that these children and their families were seen early in the clinic, so that they would not have a long wait in any delays built up during the day. The diagnosis was explained or excluded and a treatment was prescribed when needed according to the standards of care for lymphedema in our country.⁷ Information was given on complications⁸ and how to prevent them as much as possible. We then performed an initial assessment of expectations and ability to cope with the diagnosis and management. Contact with a patient support group and participation in the education

program were proposed. The parents were provided with a telephone number that they could call if they faced any difficulties at home. Contact with local health care professionals was established where possible.

“Montpellier weekends” education program

This program was developed according to the issues and needs identified during the 3 years of focal groups. It was thus the result of an interaction between families and professionals. The patient support group, AVML (for “Association Vivre Mieux mon Lymphoedème”), was responsible for organizing the social aspects of the weekends. The name “Montpellier weekends” was proposed, deliberately linking the education program to social life.

The program is a mixture of personal and group activities prescribed according to the specific needs of each individual. It provides knowledge about what lymphedema is, healthy eating, physical activity, cellulitis, skin care, and compression. It also offers parents and young adults training in skin and nail care, self-drainage, and self-bandaging.

The 1st day of each weekend, a Friday, is dedicated to individual care and management. Each child or young adult is seen by the referral specialist at the clinic, and specific treatments, such as lymphatic drainage, nail care, specific compression, and consultations with a psychologist, are organized. This time dedicated to the patients is important, to ensure the optimization of treatment and to allow patients to obtain answers to their questions, so that they can then take part in the collective activities the next day without being worried about personal issues.

During the 2nd day, collective activities are organized around a swimming pool. Water is very symbolic of lymph. Swimming and physical exercise in water are performed in groups, with explanations so that patients can continue the activities in everyday life. The physical activity was adapted from an already existing program for children.⁹ Last, but not least, wearing a swimming costume, which exposes the lymphedema, can be very difficult for some patients, and psychological help is provided when needed. In the morning, a talk is given to pass on scientific news and to share information about projects in which patients and their families can participate, with or without the support group. New tools are often developed or tested during these weekends, such as the device a father made for rolling bandages.

In addition to the activities of the training program, social activities are organized by support groups, and families are given time together to help them to establish links.

International camps and research

Raising awareness. The first international camp was organized to highlight the cause of children with lymphedema internationally, as it was deliberately organized to take place during an international ILF and Partenariat français du lymphoedème conference in 2012. Children and adolescents from Canada, Denmark, France, and Sweden took part in two films created by professionals from the film industry. The first showed the highlights of the camp and was a very simple and happy film. This film was shown during the conference and had a great success with a long-standing ovation. The second film was the first educational film to be produced on this subject. It had three parts and was made in collaboration with

a film industry professional and an American actor. The first part was filmed in the swimming pool and introduced the children in the swimming pool. The second part was a cartoon, in which the machinery of the swimming pool was used to provide a simple explanation of what lymphedema is. In the last part of the film, the children related their experiences of the diagnosis and consequences of lymphedema.

Exploring quality of life, self-management, and self-efficacy

When faced with a chronic illness, such as lymphedema, parents and children must address the issues of engagement with treatment, to help control swelling and to reduce complications, such as cellulitis, while trying to ensure the patient's well-being and normal childhood development. "Self-efficacy" is the subjective judgment, by individuals and families, of their own capacity and ability to undertake a course of action to achieve a specific goal. People's expectations of treatment depend on many factors, but are significantly influenced by their own judgment of how well they will be able to perform activities in different situations and to solve problems.¹⁰

As this area remained largely unexplored in children with lymphedema, a specific study was performed in 2017, at the Candia Canavese camp, near Turin in Italy by the Center of Research of Immunopathology and Rare Diseases of Piedmont and Aosta Valley. This study explored the enabling factors and barriers to self-management in children, adolescents, parents, and professionals in different health care systems, cultures, and languages. It was divided into three streams. One explored the parental experience of caring for a child or adolescent with lymphedema and the daily challenges of self-management. Another made use of visual art methods to explore how children and adolescents perceived their lymphedema and to conceptualize the barriers and enablers in self-management. The final stream explored the experiences of health care professionals caring for children and adolescents with lymphedema, investigating their understanding and implementation of self-management strategies, and the influence of their own self-efficacy beliefs on this process. The role of educational camps in promoting self-management and self-efficacy was explored in all groups. The results have been published and are available online.¹¹⁻¹³

Along with this study, the validation of a quality-of-life scale for children with lymphedema, initially developed in France (NCT01922635), was extended during this camp, with face-to-face meetings in three different languages and forward and back translations into four other languages (Danish, Dutch, German, and Turkish), with a Spanish translation pending through an international research study led by the Nottingham University and promoted by the Montpellier Hospital.

Discussion of the Results

All focus groups indicated that lymphedema had an impact on the lives of children and their families, with the greatest difficulties occurring during adolescence.¹¹ Chronic rare diseases, such as lymphedema, are challenging for patients, their families, and health care professionals. We have developed a specific professional organization and link this health care organization with psychosocial activities and clinical research based on the needs of patients and families we

identified. This creates an integrated pathway of holistic management of lymphedema in children and young adults.

The lack of dedicated lymphedema services and the poor evidence base for treatment are major factors with a negative impact on the experiences of parents. A lack of reliable information led parents whose children were not treated by a dedicated multiprofessional service to search continually for answers and to develop a mistrust of the medical profession. This has already been shown for lymphedema in the United Kingdom¹⁴ and for other chronic rare diseases, such as cystic fibrosis.¹⁵ Our results, confirming these previous findings, indicate that this situation is not culture- or health care system-specific.

Uncertainty was also a major factor for all parents but was most severe in those searching for expert care. The magnitude of uncertainty in the management of a chronic disease is accentuated if the condition concerned is an orphan illness not clearly covered by one particular medical specialty, or for which there is no access to specialist care, a common problem for lymphedema.¹⁶

Two changes in our organization had an impact on the anxiety of parents and children. The first of these changes was the provision of a telephone number, to enable parents to contact the team if they encountered any difficulties with the carers or the health care professionals caring for their children. The second was allowing children and adolescents to spend time with other children and adolescents with lymphedema during their time at the center, facilitating the establishment of a social life. Any health care professional caring for children with lymphedema could provide these aspects within their own organization.

The camps afforded professionals an opportunity to express their own distress when faced with a child with lymphedema, and to address their own perception of self-management and self-efficacy in this context. We did not expect to identify such high difficulty in professionals working at the community level mainly because of their own uncertainty about the appropriateness of their management. This proved to be a reverse knowledge-transfer experience. "Reverse knowledge transfer" is a concept developed in business, specifically in the context of multinational corporations. It has been suggested that the determinants of conventional knowledge transfers from the parental companies of multinational corporations to subsidiaries and reverse knowledge transfers from the subsidiaries to the parental companies are based on different transfer logics. Knowledge transfer is a process in which an organization recreates a complex, causally ambiguous set of routines in new settings and keeps the routines functioning. These routines appear in the form of know-how and are often tacit ones that do not necessarily flow easily.¹⁷ This applies to the camp situation, if you consider that patients and their physiotherapists are subsidiaries with respect to a parental expert center and that knowledge transfer aims to recreate a set of routines in a new setting and to keep them functioning. Difficulties in knowledge transfer were clearly expressed by professionals working on their own and have to be specifically addressed.

Finally, the involvement of patients in the organization of the center and in identifying the questions to be addressed by specific research has led to significant research advances. As an example, the LymphoRAC study was designed following the patients request and has included patients with primary

lymphedema.¹⁸ It can also lead to innovation from the patients, with one father, for example, creating a bandage roller from a well-known children's game. He now provides other parents with a guide to allow them to produce a similar device for themselves.

The role of support groups and organizations linking patients and professionals at the national and international level such as the ILF, LERN, and VASCERN, is vital, to increase awareness and ensure the establishment of sound organizations of health care.

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