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Reseach

Comprehensive and disease-specific quality of life in Japanese young people with primary lymphoedema: a case series

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ABSTRACT

Primary lymphoedema is rare, and the quality of life (QOL) of children and young people with lymphoedema has received little attention. This study aimed to describe the comprehensive and disease-specific QOL of children and young people with primary lymphoedema in Japan. The Japanese version of the KIDSCREEN-27 was used to describe the comprehensive QOL of these children and young people. The English version of the lymphoedema-specific QOL tool was translated into Japanese (J-LYMPHOQOL) and used. Six young people aged 15-21 years participated in this study. Regarding comprehensive QOL, young people with primary lymphoedema had high scores on *Social Support and Peers* and moderate scores on *Physical Well-Being and Psychological Well-Being*. Regarding disease-specific QOL, all young people with primary lymphoedema had a J-LYMPHOQOL score of 80 in the dimension *At School/College/Work*. Furthermore, 5 of 6 young people scored 80 or higher for the *At Home*, and *You and Your Feeling* dimensions. In contrast, zero and two respondents, respectively, scored 80 or higher in the dimensions *You and Other Children/Peers*, and *Treatment Considerations*. In conclusion, physical changes may affect self-acceptance and self-esteem, suggesting that fashion-related support is vital in addition to psychological support.

KEY WORDS: case series, disease-specific QOL, primary lymphoedema, radar chart, young people

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Introduction

Lymphoedema is chronic edema caused by the stagnation of the lymphatic circulation.[1)][2)] Lymphoedema can be classified into two types: primary lymphoedema, a disorder of lymphangiogenesis, and secondary lymphoedema, which is caused by lymphatic tissue damage due to lymph node dissection or radiation therapy as part of cancer treatment.

Primary lymphoedema is caused by developmental abnormalities in the lymphoid system and is prevalent in children.[3)][4)] It is important to note that the quality of life (QOL) of patients with primary lymphoedema may be impaired by the physical, psychological, and social effects of oedema,[5)] which can occur in all parts of the body, including the extremities. Particularly in children and young people, the physical, psychological, and social effects of oedema may affect their growth and development. Therefore, understanding the QOL of children and young people with lymphoedema and managing it appropriately are crucial.

To date, the QOL measurement for limb lymphoedema (LYMQOL) survey form, which is a reliable, validated,[6)][7)] and Japanese-translated method for assessing QOL,[8)] has been used to assess the QOL of adult patients with lymphoedema. Furthermore, a cross-sectional survey of patients with primary lymphoedema have been conducted using comprehensive QOL tool.[9)] However, this evaluation tool is only used in adults. Indeed, there is a health-related QOL scale called KIDSCREEN.[10)] This tool is conceptually based on the definition of QOL as a multidimensional construct with physical, emotional, mental, social, and behavioral components of well-being and functioning as perceived by children and young people. Unfortunately, this is a comprehensive tool for measuring QOL and cannot assess the impact of lymphoedema on QOL. Thus, no measurement tool is available in Japan or overseas for evaluating disease-specific QOL in children and young people with lymphoedema at different developmental stages. A disease-specific QOL tool can help healthcare professions understand the impact of lymphoedema on children's and young people's unique QOL or developmental issues, for example, not wanting to go to school because of swollen limbs, and provide appropriate care for these children and young people.

In 2021, the International Lymphoedema Framework (ILF), an academic organization specializing in oedema, developed the lymphoedema-specific QOL tool (LMYPHOQOL, a version for children & young people), which is a QOL assessment tool for children and young people with lymphoedema. Furthermore, a research project was conducted to verify its reliability and validity in several countries and to compare the actual status of QOL internationally. The authors participated in this research project in Japan and developed a Japanese version of the LYMPHOQOL (J-LYMPHOQOL) for children and young people with lymphoedema.

This study aimed to describe previously unknown QOL in children and young people with lymphoedema in Japan. Future care and treatment can be evaluated by using the J- LYMPHOQOL for children and young people. Furthermore, the same LYMPHOQOL for children and young people survey allows for international comparisons and provides evidence for managing lymphoedema in these persons.

Materials and Methods

1. Study design

A case series study design was used to describe QOL.

2. Participants

This study included children and young people with primary lymphoedema. The inclusion criteria were follows: persons aged between 5-21 years with a diagnosis of primary lymphedema made by a pediatrician or a lymphoedema specialist. The exclusion criteria were as follows: persons who had difficulty understanding the questionnaire even with medical or family follow-up and those who the physician deemed unable to participate in the study.

We were referred by a physician at a hospital that manages lymphoedema in children or young people and recruited the target population using the snowball method. Furthermore, we distributed e-mails to directors and other members of the ILF Japan or directors of the Japanese Society for Lymphedema Therapy and sent e-mails to certified nurses in wound ostomy continence care and persons qualified for lymphoedema management. Furthermore, we distributed information through our website and social networking service

(SNS). In addition, the survey was distributed via a website and SNS to recruit facilities and medical professionals who agreed to participate in the survey and recruit the target population.

3. Data items

A self-administered questionnaire survey on comprehensive QOL and disease-specific QOL, present age, age at onset, oedema location, history, and complications was administered between April 2022 and June 2023. If a person failed to complete the form, their parent proxy answered the questionnaire.

The Japanese version of the KIDSCREEN-27 (J-KIDSCREEN-27)[11] was used to comprehensively describe QOL. This tool has five dimensions: *Physical Well-Being* (5 items), *Psychological Well-Being* (7 items), *Parent Relation & Autonomy* (7 items), *Social Support & Peers* (4 items), *School Environment* (4 items). Responses were scored on a 5-point Likert-type scale assessing frequency: never (1), seldom (2), quite often (3), very often (4), always (5), or intensity: not at all (1), slightly (2), moderately (3), very (4), extremely (5), with a 1-week recall period. The reliability and validity of this tool have been established.[11] Permission to use for this study was obtained from the originator of the J-KIDSCREEN-27.

The J-LYMPHOQOL for children and young people with lymphoedema was used to describe the disease-specific QOL. The original version is divided into developmental stages for children aged 5–12 years and for young people aged 13–21 years. Each version comprises seven dimensions: *You and Other Children/Peers* (7 items), *You and Your Physical Activity* (3 items), *At School/College/Work* (6 items), *At Home* (6 items), *You and Your Feeling* (10 items), *You and Your Free Time* (3 items), and *Treatment Considerations* (10 items). Responses are marked on a 5-point Likert-type scale assessing frequency: never (1), sometimes (2), often (3), very often (4), and always (5), with high scores indicating high QOL. Three open-ended questions for qualitative QOL assessment then followed after the selected-response questions: *What good treatment means for you?*, *What makes living with lymphoedema better or worse for you?* and *How good your life is overall?*.

Permission for the translation into Japanese was obtained from the ILF President. Forward and backward translations were performed and the J-LYMPHOQOL for children and young people was completed (**Appendix**).

4. Data collection process

The survey request was explained to the participants, and consent was obtained at each facility from the patients or families with the consent of the physician-in-charge at each facility. The survey explanation and consent forms used for the survey were prepared by the researcher. If the participant consented to participate in the survey, the physician-in-charge provided the survey form and returned the envelope to the participant or their proxy. If a participant was not enrolled in a school, the parent or proxy explained the questionnaire and provided the answers of the participant.

After obtaining responses, the survey form was returned to the research office by mail.

5. Analysis

Although the scoring methods for the J-KIDSCREEN-27 has been established,[11] we used a radar chart to describe the comprehensive and disease-specific QOL of each participant in the same style. Scores in the J-KIDSCREEN-27 and J-LYMPHOQOL for children and young people were converted into the following format for each dimension: from 0 to 100. A radar chart was established based on the converted values.[12] The overall average radar chart was added to the individual radar charts. A radar chart in Microsoft Excel (version 2403) was used to visually display each dimension of the J-KIDSCREEN-27 and J-LYMPHOQOL for children and young people. Open-ended responses for qualitative QOL description were provided by the participants.

6. Ethical consideration

This study was conducted according to the guidelines of the Declaration of Helsinki and it was approved by the Medical Research Ethics Review Committee of Fujita Medical College (Approval No. HM-21-466, 22 April, 2022).

Results

Of the 10 facilities that were requested to participate in the study, eight agreed to participate, and nine participants were referred. Among them, six partici-

付録　LYMPHOQOL 日本語版（13ー 21 歳用）

周りの人々のとの関わり

リンパ浮腫について容易に話すことができますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

あなたは他の子供／若い人がリンパ浮腫を見ることを心配に感じますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

リンパ浮腫が原因で、他の子供／若者からほかの人とは異なる扱いを受けることはありますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

履きたい靴を見つけられますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

着たい服を見つけることができますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

他の子供／若者がリンパ浮腫のことであなたをいじめることがありますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

他のリンパ浮腫を持った子供や若者と一緒にいる時間は、過ごしやすいですか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

身体活動

リンパ浮腫が原因でできないスポーツや身体活動はありますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

リンパ浮腫が原因でスポーツや身体活動を避けていますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

身体活動に参加するのは楽しいですか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

学校／大学／職場で

リンパ浮腫が原因で教師／講師／マネージャーから他の子供／若者とは違う扱いを受けますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

友達はあなたにリンパ浮腫があることについて、学校／大学／職場であなたを守ってくれますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

リンパ浮腫が原因で学校／大学／仕事に行けなくなったことはありますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

リンパ浮腫が原因で学校／大学／仕事に行けなくなることは悲しいですか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

リンパ浮腫が原因で学校／大学／仕事のことを行うのが困難であると感じますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

リンパ浮腫が原因で物事に集中するのが難しいと感じるときはありますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

自宅で

両親はリンパ浮腫が原因であなたが何か特定のことをするのを止めますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

リンパ浮腫のケアをすることは家族の時間に影響しますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

あなたはリンパ浮腫があることによってあなたの兄弟姉妹と異なった扱いを受けていますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

リンパ浮腫が原因で、服を着たり脱いだりするのが難しいと思いますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

リンパ浮腫が原因で眠りにくかったことはありますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

あなたはリンパ浮腫があることによって、特別なことをすることは許されていますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

気持ち

リンパ浮腫があることで悲しさを感じることがありますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

リンパ浮腫があることで他の人と違うと感じることがありますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

リンパ浮腫があることで恥ずかしい思いをすることがありますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

リンパ浮腫のために将来が心配と感じることがありますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

リンパ浮腫が悪化するのではないかと心配することがありますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

感染や怪我をするのではないかと心配することがありますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

リンパ浮腫があることで特別な気分になりますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

リンパ浮腫のために孤独を感じることがありますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

リンパ浮腫のために怒りを感じることがありますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

あなたはリンパ浮腫があることで、世話をされていると感じますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

自由時間

リンパ浮腫によってあなたが何か物事をする際に妨げられたと感じることはありますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

リンパ浮腫のために注意深く見守られすぎていることはありますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

あなたはリンパ浮腫を持っていない他の子供／若者と同じことをすることができますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

治療と自己管理

誰かから、リンパ浮腫とは何か説明されたことはありますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

推奨される治療法に従うのは難しいと感じますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

治療をすることで元気になりますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

リンパ浮腫が悪化した場合、あなたはリンパ浮腫のケアをすることができますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

あなたは勧められた治療に専念することができますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

あなた自身でリンパ浮腫のケアをすることに対して自信はありますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

頑張ったらリンパ浮腫の問題を解決できると感じますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

リンパ浮腫に関して何か問題が発生したときに、落ち着いて過ごすことができますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

治療をやめたいと思うことがありますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

あなたの治療は他の活動の邪魔になりますか？

□全くない　□ときどき　□頻繁に　□とても頻繁に　□いつも

生活の質について

あなたにとって良い治療とは：

リンパ浮腫とともに生活していく上で良いこと、または悪いこと：

全体的にあなたの人生はどのくらい良いと感じますか：

あなたとあなたのリンパ浮腫について何か知りたいことはありますか？

For permission to use contact：国際リンパ浮腫フレームワーク・ジャンパン研究協議会事務局

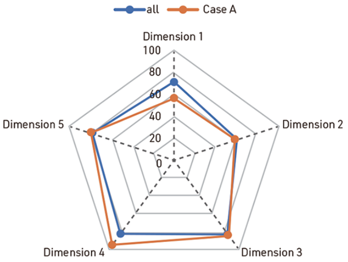
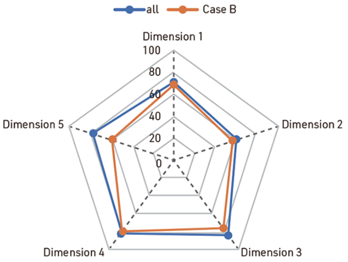
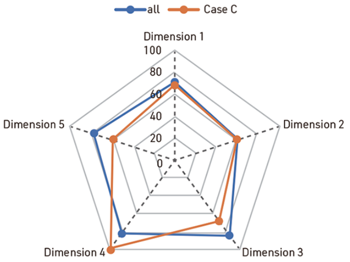
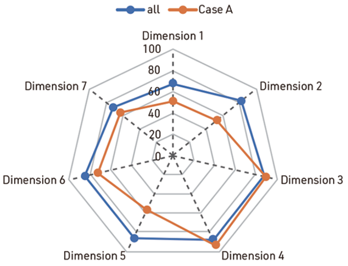
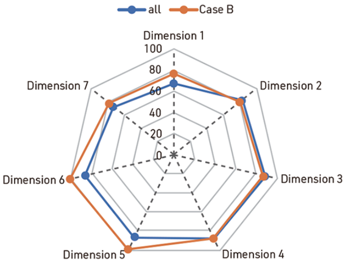
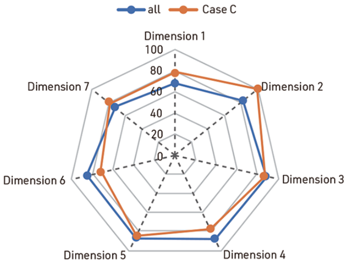
pants 15–21 years agreed to participate. The demographic data, medical history, edema location and QOL of the participants are presented **Table 1**. All responses to the survey form were provided in person.

1. Comprehensive and disease-specific QOL

Case A was a 15-year-old female with oedema in both feet. She was diagnosed with primary lymphoedema at the age of 13 years.

J-KIDSCREEN-27 scores in each dimension were similar to the overall average. QOL scores in *Physical Well-Being* and *Psychological Well-Being* were less than 80. Scores for each dimension of the J-LYMPHOQOL for young people were lower than the overall average. Two dimensions exceeded 80: *At School/College/Work*, and *At Home*. In contrast, *You and Other Children/Peers*, *You and Your Physical Activity*, and *You and Your Feeling* were lower than the other dimensions, ranging from 51 to 56. She has never had a patient with primary

Table 1 Participants' chracteristics and QOL radar charts

	Case A	Case B	Case C
Age and gender	15-year-old female	15-year-old female	17-year-old female
Age on set	13	as an infant	15
Location of oedema	both feet	left forearm and left hand	both feet
Complications	none	none	none
J-KIDSCREEN-27			
J-LYMPHOQOL			

The orange radar chart showed each case's QOL score. The blue radar chart showed the average QOL score for all cases.
J-KIDSCREEN-27, Dimension 1: Physical well-being, Dimension 2: Psychological well-being, Dimension 3: Parent relation & autonomy, Dimension 4: Social support & peers, Dimension 5: School eviroment
J-LYMPHOQOL, Dimension 1: You and other children/peers, Dimension 2: You and your physical activiy, Dimension 3: At school/college/work, Dimension 4: At home, Dimension 5: You and your feelings, Dimension 6: You and your free time, Dimension 7: Treatment considerations

lymphoedema before.

In her free responses, wearing an elastic stocking was a good treatment for herself, and sitting with her lower extremities down made her symptoms worse. Overall, she felt life was not so good and wanted to know about which treatment was best for her lymphoedema.

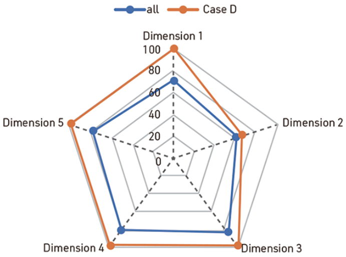
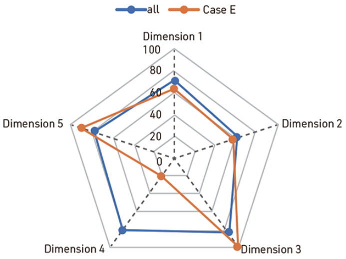
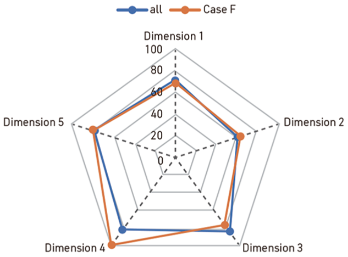
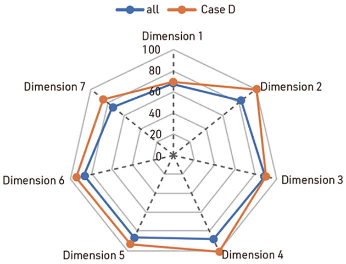
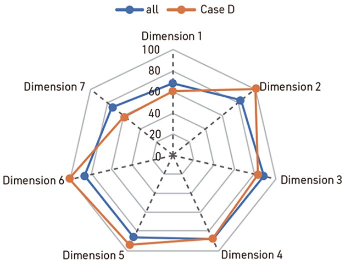
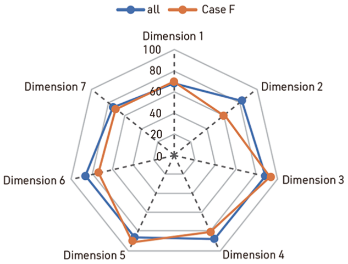
Case B was a 15-year-old female with oedema of the left forearm and hand. She was diagnosed with primary lymphoedema as an infant.

J-KIDSCREEN-27 scores in each dimension were lower than the overall average. QOL scores in *Physical Well-Being*, *Psychological Well-Being* and *School Environment* were less than 80. Scores for the J-LYMPHOQOL for young people in each dimension were higher than the overall average. Five dimensions exceeded 80: *You and Your Physical Activity*, *At School/College/Work*, *At Home*, *You and Your Feelings*, and *You and Your Free Time*. The scores for *You and Your Free Time*, and *You and Your Feeling* dimensions were high at 100 and 98, respectively. She did not answer the open-ended questions.

Case C was a 17-year-old female with oedema in both feet. She was diagnosed with primary lymphoedema at the age of 15 years.

J-KIDSCREEN-27 scores in each dimension were lower than the overall average, except for *Social Support & Peers*. Scores for *Physical Well-Being*, *Psychological Well-Being*, *Parent Relation & Autonomy* and *School Environment* were lower than 80. Scores for the J-LYMPHOQOL for young people in each dimension were higher than the overall average except for *At Home*, and *You and Your Free Time*. Four dimensions exceeded 80: *You and Your Physical Activity*, *At School/College/Work*, *You and Your Feeling*, and *Treatment Considerations*. The score obtained for *You and Your Physical Activity* was high at 100.

Table 1 (Continued)

	Case D	Case E	Case F
Age and gender	19-year-old female	19-year-old male	21-year-old male
Age on set	2	Klippel-Trenaunay-Webber Syndrome diagnosed as an infant	as an infant
Location of edema	right foot and right toes	both legs, feet, anf toes	head, face, scrotum, right upper arm, right forearm, right hand, right fingers, right leg, right feet, and right toes
Complications	cellulitis	cellulitis	repeated punctures for pleural and ascitic effusions malnutrition, respiratory distress
J-KIDSCREEN-27			
J-LYMPHOQOL			

The orange radar chart showed each case's QOL score. The blue radar chart showed the average QOL score for all cases.
J-KIDSCREEN-27, Dimension 1: Physical well-being, Dimension 2: Psychological well-being, Dimension 3: Parent relation & autonomy, Dimension 4: Social support & peers, Dimension 5: School evironment
J-LYMPHOQOL, Dimension 1: You and other children/peers, Dimension 2: You and your physical activiy, Dimension 3: At school/college/work, Dimension 4: At home, Dimension 5: You and your feelings, Dimension 6: You and your free time, Dimension 7: Treatment considerations

In her free responses, she would like to know what treatment would be good for her. The good thing about living with lymphoedema was that she understood the pain of the disease. The bad thing about living with lymphoedema was that she had heavy legs. Overall, she responded that life was very good.

Case D was a 19-year-old female with oedema in the right foot and toes, which was complicated by cellulitis; she was diagnosed with primary lymphedema at the age of 2 years.

J-KIDSCREEN-27 scores in each dimension were higher than the overall average. The score in *Psychological Well-Being* was lower than 80. Scores for the J-LYMPHOQOL for young people in each dimension were higher than the overall average. Six dimensions except for *You and Other Children/Peers* exceeded 80. Both *You and Your Physical Activity*, and *At Home* showed high scores of 100.

In her free responses, the best treatment for herself was a steady, step-by-step healing process. The bad things in her life were not being able to pick out shoes to wear, feeling sluggish all over her oedema feet, and not being able to take off her socks easily. Overall, she stated that her life was very good. She would like to know how many patients with primary lymphoedema there were and if primary lymphoedema was a rare disease.

Case E was a 19-year-old male with oedema in both legs, feet, and toes, whose condition was complicated by cellulitis. The cause of the lymphoedema was Klippel-Trenaunay-Webber Syndrome. He was diagnosed with primary lymphoedema as an infant.

J-KIDSCREEN-27 scores in each dimension were comparable to the overall average score, except for *Social Support & Peers*. Score of *Social Support & Peers* was less than 20.

Scores for the J-LYMPHOQOL for young people in each dimension were comparable to the overall average score, except for *Treatment Considerations*. Five dimensions exceeded 80: *You and Your Physical Activity*, *At School/College/Work*, *At Home, You and Your Feelings*, and *You and Your Free Time*. In contrast, *You and Other Children/Peers*, and *Treatment Considerations* were lower than those of the other dimensions with 60 and 58, respectively.

In his free responses, the best treatment for him was to proceed with the treatment while discussing it thoroughly with his doctor. He was not particularly concerned about lymphoedema because it was a disease had been born with and was natural to him. Overall, he responded that life was very good.

Case F was a 21-year-old male with oedema in the head, face, scrotum, right upper arm, forearm, hand, and fingers. Furthermore, he had oedema in the right leg, feet, and toes and had previously undergone repeated punctures for pleural effusions and ascites. He had decreased nutritional status and respiratory distress and was diagnosed with primary lymphoedema as an infant.

J-KIDSCREEN-27 scores in each dimension were comparable to the overall average score, except for *Social Support & Peers*. Score of *Social Support & Peers* was 100. Scored for the J-LYMPHOQOL for young people in each dimension were comparable to the overall average score, except for *You and Your Physical Activity*, and *You and Your Free Time*. Three dimensions exceeded 80: *At School/College/Work*, *At Home* and *You and Your Feeling*.

In his free responses, a good treatment for him was the material of the elastic bandage, which did not cause any discomfort during use in daily life. The oedema was so severe that he lost confidence in his ability to get stronger and had a hard time. Overall, 90% of life was good, and the remaining 10% was fulfilled when lymphoedema was fully cured.

Discussion

The QOL of young people with primary lymphoedema aged 15-21 years was described by using a comprehensive QOL tool, J-KIDSCREEN-27, and a disease-specific QOL tool, J-LYMPHOQOL for young people. Our study results revealed that young people with primary lymphoedema had high scores on *Social Support and Peers* and moderate scores on *Physical Well-Being* and *Psychological Well-Being* in terms of comprehensive QOL. In contrast, regarding disease-specific QOL, all participants had a J-LYMPHOQOL for young people score of 80 on *At School/College/Work*. In addition, 5 out of 6 young people scored 80 or higher for *At Home*, and *You and Your Feeling*. In contrast, zero and two young people respectively scored 80 or higher for *You and Other Children/Peers*, and *Treatment Considerations*. These results suggest that involvement in *You and Other Children/Peers*, and *Treatment Considerations* are crucial for improving the QOL of young people with primary lymphoedema.

1. Disease-specific QOL

Okajima et al. measured the QOL of adult patients with primary lymphoedema using the SF-36.[9] They reported that scores in the general health perception domain were lower than those of the healthy adult population; however, scores for the social functioning, role emotion, mental health, and vitality domain were similar to those of healthy adults. Although the comparison of results was challenging because of differences in ages and QOL tools, the results for role emotion and *You and Your Feeling* were similar. In contrast, the results for social functioning and *You and Other Children/Peers* were different. We believe that this difference was influenced by the age of the study participants. The mean age of the 83 participants in the previous study was 42.3 years, and their SF-36 emotional role scores were 49.6, which was similar to those of healthy adults. In contrast, the participants in this study ranged in age from 15 to 21 years, and none of their *You and Other Children/Peers* scores were above 80. The age group of 15-21 years is a time when ties to peer groups become stronger, and we believed that developmental issues, rather than disease, may have influenced the scores of the participants.

The distribution of scores for all participants in the dimension *You and Other Children/Peers* ranged from 51 to 77. This dimension includes two unique items related to clothing and shoes such as, "Can you find shoes that you like to wear?" We believe that the participants' oedematous feet and extremities contributed to their low scores. Furthermore, shoes and clothing for young people serve as a means of showing their sense of value to others.[13] In the future, building a collaborative partner for shoes and clothing for young people with primary lymphoedema will be imperative.

Lymphoedema is a disease in which the symptoms can be seen and touched directly by others, and by the patient. This characteristic of lymphoedema affected the scores for *You and Other Children/Peers* and may have influenced developmental issues such as stronger ties in peer groups than at home or school. Changes in the appearance of females with secondary lower extremity lymphoedema have been reported to decrease body image.[14-17] In this study, the same was suggested for young people with primary lymphoedema. Therefore, active involvement in this area was required.

The distribution of scores for the four participants in *Treatment Considerations* ranged from 58 to 78. Moreover, three of them had disease onset in infancy. This suggests that a longer disease duration does not improve scores in *Treatment Considerations*. Lymphoedema that develops in infancy is considered to be the result of congenital abnormalities, such as hypoplasia or hyperplasia of the lymphatic vascular system.[3] Although several genetic abnormalities have been identified as causes of this condition, identifying them is time-consuming and costly for patients and their families. Therefore, it was speculated that some young persons were treating and self-managing lymphoedema of unknown pathogenesis.

From the free responses, it was clear that their relationship with their doctors, knowledge about lymphoedema, the heaviness of the legs derived from oedema, and material of elastic garment affected the QOL of young people with primary lymphoedema.

The LYMPHOQOL for children and young people is still undergoing reliability and validation studies. Therefore, details of the scoring method are not yet available. We devised an original method known as the radar chart.[12] Radar chart visualization helps healthcare providers understand the dimensions of the J-LYMPHOQOL for children and young people that require intervention. Sharing this information with other healthcare professions will encourage healthcare team activity. Furthermore, it could be a useful communication tool for young people with primary lymphoedema. This radar chart could also be used to encourage young people themselves participate in lymphoedema management planning.

2. Comprehensive QOL

Young people with primary lymphoedema had high mean scores of 95 for *Social Support and Peers* and moderate scores for *Physical Well-Being* and *Psychological Well-Being* (mean scores of 70.6, and 60.5, respectively). In a J-KIDSCREEN-27 survey of 12-18-year-persons attending school in Japan, the mean *Social Support and Peers* scores were 49.1 for boys and 53.9 for girls. Furthermore, the mean *Physical Well-Being* scores were 49.7 for boys and 49.0 for girls, and the mean *Psychological Well-Being* scores were 46.9 for boys and 48.8 for girls.[11] These scores were transformed into T scores with a mean of 50 derived from a European study and a standard deviation of 10. T scores are standardized individual scores used for international comparisons. Although the scores were calculated differently, the *Social and Peers* scores of young people with primary lymphoedema suggested to be not lower than those of healthy young people. In contrast the *Physical Well-Being* and *Psychological Well-Being* scores of young people with primary lymphoedema were suggested to be lower than those of healthy young people.

3. Usability of J-LYMPHOQOL

Because an important reason to assess QOL in growing children and young people is to facilitate communication, we investigated the usability of the J-LYMPHOQOL for young people. Four of five participants responded positively to all usability items of the J-LYMPHOQOL. In contrast, one participant gave a negative evaluation for the three items: *Easy to Use, Helpful*, and *Useful* (**Table 2**). The reasons for the low

Table 2　Usability of J-LYMPHOQOL

Item	case A	case B	case C	case D	case E	case F
Easy to use	3	3	5	4	2	4
Helpful	4	3	5	5	2	4
Meaningful	4	3	5	5	4	5
Thought provoking	3	3	5	5	3	5
Useful	3	3	5	5	2	4

5: extremely, 4: very, 3: moderately, 2: not very, 1: not at all

ratings were not confirmed in this study. To improve the usability of this tool in the future, clarifying the reasons for low ratings through qualitative research will be necessary.

4. Future research

The development of a case-counting system is necessary to verify the reliability and validity of the J-LYMPHOQOL for children and young people because primary lymphoedema is a rare disease in Japan.

Children and young people with lymphoedema have not been evaluated using QOL scales for their generation. Therefore, when interpreting individual disease-specific QOL scores in this study, the mean of the study participants was used as a relative rating. In addition, 80 point was used as an absolute rating. This cutoff score for interpreting disease-specific QOL was 80 tentatively; however, there was no evidence for this cutoff value. Thus, scoring criteria for clinically meaningful interpretation should be determined.

5. Limitations

A limitation of this study was the small number of participants. This was because there were few medical institutions in Japan that were specialize in primary lymphoedema. Furthermore, the study included young people who were referred by their physicians, which may have led to a selection bias. Moreover, children under 15 years of age did not participate in this study. Therefore, the generalizability of the results remains an issue.

Case F was a person with generalized oedema with pleural effusions and ascites, which might differ from generalized primary lymphoedema. Determining whether the case F met the inclusion criteria of this study was difficult because the exact pathophysiology could not be determined using the self-administered questionnaire alone.

Conclusions

The QOL of young people with primary lymphoedema was low in the *You and Other Children/Peers* and *Treatment Considerations*. Physical changes due to oedema of the limbs may negatively affect self-acceptance and self-esteem, suggesting that fashion-related support is essential in addition to psychological support.

Primary lymphoedema is rare, and the QOL of children and young people with lymphoedema has received little attention. In the future, the J-LYMPHOQOL for children and young people will enable healthcare teams to intervene with awareness of the QOL of children and young people.

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Conflicts of Interest

The authors declare no conflicts of interest.

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日本における原発性リンパ浮腫を有する若年者の包括的ならびに疾患特異的QOL：ケースシリーズ

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要　旨

原発性リンパ浮腫はまれであり、リンパ浮腫を有する小児および若年者のQOLはほとんど注目されていない。本研究は、日本における原発性リンパ浮腫の小児および若年者の包括的および疾患特異的QOLを記述することを目的とした。これらの小児および若年者の包括的QOLを記述するために、KIDSCREEN-27の日本語版を用いた。リンパ浮腫に特化したQOLツールの英語版を日本語に翻訳して使用した（J-LYMPHOQOL）。本研究には、15～21歳の6人の若年者が参加した。包括的QOLに関しては、原発性リンパ浮腫の若年者は、「社会的な支えと仲間」で高得点を示し、「身体的幸福感」、「心理的幸福感」で中程度の得点を示した。疾患特異的QOLに関しては、原発性リンパ浮腫の若年者は全員、「学校/大学/職場で」という次元でJ-LYMPHOQOLスコアが80点であった。さらに、6人中5人が「自宅で」、「気持ち」の次元で80点以上であった。対照的に、「周りの人々との関わり」、「治療と自己管理」という次元で80点以上を獲得した回答者は、それぞれ0人と2人であった。結論として、身体的変化は自己受容と自尊心に影響を及ぼす可能性があり、心理的支援に加えてファッション関連の支援が不可欠であることが示唆された。

キーワード：ケースシリーズ、疾患特異的QOL、原発性リンパ浮腫、レーダーチャート、若年者