Living with a Rare Vascular Liver Disease

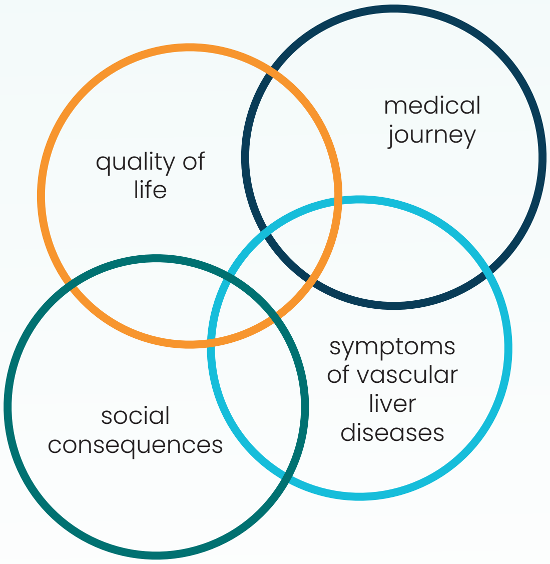
Peer Research on Quality of Life

Portal vein thrombosis (PVT) and Budd–Chiari syndrome (BCS) are rare vascular liver diseases that can significantly affect daily functioning and quality of life (QoL). Despite their medical complexity and their impact on both physical and mental well-being, little is known about how patients experience life with these conditions and which care needs may remain unmet.

Through a peer research study, insights were gained into the quality of life of individuals living with PVT or BCS, and unmet care and support needs were identified. These insights are essential for improving patient-centered care and providing appropriate support.

Method

Data were collected through in-depth interviews conducted by peer researchers (experts by experience) between March and April 2024. These peer researchers were involved from the start of the project and actively contributed to developing the topic guide. The interviews (n=4) were conducted face-to-face, allowing for open conversations and deeper exploration of the participants' personal experiences and needs.



Important themes

The interviews revealed four central themes that are closely interconnected and collectively shape the lives of people with PVT or BCS. These themes are presented in the figure on this page.

Symptoms of Vascular Liver Diseases

The direct symptoms, complaints, and medical aspects associated with PVT or BCS.

Quality of Life

The overall impact of the condition on patients' physical, mental, and emotional well-being.

Medical journey

Patients' experiences with diagnosis, treatment, and follow-up within the healthcare system.

Social consequences

The effect of the disease on relationships, work, social functioning, and participation in society.

The overlapping areas in the figure highlight how these themes influence one another. For example, the medical journey can affect the perceived quality of life, while social consequences may be intensified by the symptoms caused by the liver disease. This interconnection underscores the importance of an integrated approach to care and support for these people.

Symptoms of Vascular Liver Diseases

People with a vascular liver disease mainly experience fatigue and difficulty processing stimuli. Symptoms are often unpredictable, causing patients to remain constantly alert and plan ahead in an effort to stay ahead of potential flare-ups. This requires a great deal of energy and affects their ability to function in daily life.

> “I don’t feel ill, but I do feel limited”

Care or support needs

- Coping with fatigue and sensory overstimulation.
- Support in dealing with the unpredictability of symptoms.
- Guidance in learning to anticipate potential symptoms.
- Assistance in functioning as well as possible in daily life despite the symptoms.

Social consequences

The social impact of PVT and BCS extends across multiple layers of life. In the immediate environment (*inner circle*), partners, children, and close friends play a crucial role in recognizing symptoms, providing support, and “fighting alongside” the patient. At the same time, patients struggle with the ideal of being a “good parent” or “good partner” and the pressure to remain strong. In the broader social circle (*extended circle*), which includes family, friends, and acquaintances, understanding is often limited, and openness or transparency about the disease is not always self-evident. Professional life also comes under strain: the condition affects career opportunities, income, and job applications, and sharing the diagnosis is not always easy. Finally, patients face barriers at the societal level as well, such as difficulties with insurance companies and government agencies, which increases their vulnerability in everyday life. All respondents reported that they had, in a sense, “lost the normalcy” of everyday life. They constantly have to think about what they can or cannot do, and whether or not to disclose their illness, for example, at work or in personal relationships.

Care or support needs

- Support and recognition for partners and children in their roles and the burdens they face.
- Assistance for parents struggling with the ideal of being a “good parent.”
- Greater understanding and awareness within the broader social circle.
- The opportunity to be open and transparent without feelings of guilt.
- Support in dealing with the negative effects on career, income, and job applications.
- Guidance in navigating dilemmas around disclosure in the workplace.
- Removal of obstacles related to insurance and government institutions.

> “I don’t want to disappoint my child, so instead of telling the truth, I make up an excuse that the activity is too expensive”

> “I know I have the ability to take on a higher position, but that would mean I’d have to be mentally ‘on’ all the time”

Medical journey

A well-organized and clearly structured medical journey is crucial for patients with a vascular liver disease. It begins with timely referral to the appropriate specialist and is strengthened by clear communication, transparency about treatment, and a well-defined care pathway. Patients place great value on having a designated point of contact, short lines of communication with healthcare providers, and building trust with their physician. In addition, aftercare plays an important role, because knowing what to expect and what to watch out for, provides a sense of reassurance. Finally, contact with peers is viewed as a valuable complement to professional medical care.

> “The effort the doctor made to sit next to me at eye level meant a lot to me”

Care or support needs

- Timely and accurate referral to a specialist.
- Clear communication and transparency throughout the entire care pathway.
- A designated point of contact and short lines of communication with healthcare providers.
- Building trust within the doctor–patient relationship.
- Clear aftercare: knowing what to expect and what to watch out for.
- Access to peer support as a complement to medical care.

Quality of life

For patients, quality of life is not only about physical health, but also about being seen and acknowledged as a person beyond the disease. Identifying as a patient provides a sense of stability and helps them understand their own situation. At the same time, recognition from healthcare professionals and the surrounding social environment plays an important role. Feeling understood and taken seriously greatly contributes to a sense of well-being. Despite the impact of the condition, many patients search for a “silver lining”, a positive aspect or deeper meaning in their experience, which strengthens their resilience and enhances their quality of life.

> “For me, quality means getting through the day without thinking: ‘I can’t make it’”

Care or support needs

- Being seen and acknowledged as a person, not only as a patient.
- Experiencing understanding and being taken seriously by healthcare professionals and one’s social environment.
- Finding reassurance in the diagnosis and gaining a better understanding of one’s own situation.
- Having space to find meaning or a positive element (a ‘silver lining’) in the experience.

> “I feel that the people around me live their lives more unconsciously, that many things just continue on autopilot”

Conclusion

People living with a rare vascular liver disease experience profound effects on their daily lives that extend far beyond the medical diagnosis. Their quality of life is influenced by physical symptoms, social consequences, and experiences within the healthcare system. Findings from the peer research show that recognition, open communication, and attention to the person behind the disease are crucial. By focusing in healthcare not only on the liver condition itself but also on the individual's personal situation, close relationships, and overall quality of life, care can become more appropriate, holistic, and truly patient-centered.

> “*I hope something comes out of this that makes other patients think: ‘Hey, I'm not alone in this, I'm not the only one, there are others like me*”

> “*We are more than our diagnosis*”

Recommendations for healthcare professionals

- See the person behind the diagnosis. Look at the patient's personal situation, not just their medical data.
- Actively involve close relatives. Ask about partners, children, and caregivers, and acknowledge their roles and burdens.
- Continuously re-evaluate information and support needs. Recognize that the needs of both patients and caregivers change over time, and initiate repeated conversations to provide tailored guidance.
- Actively inquire about quality of life. During consultations, discuss aspects such as energy, pain, mood, work, relationships, and finances, not only clinical findings.
- Foster open and equal dialogue. Acknowledge uncertainties, listen to personal goals, and encourage the patient's own input.

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Funded by
the European Union

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