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# PERIPORTAL CARDIOMYOPATHIES

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**Abstract:** Periportal cardiomyopathies are rare and complex cardiovascular diseases associated with the development of fibrosis in the periportal region of the liver, which subsequently affects myocardial structure and function. These conditions, although uncommon, significantly contribute to morbidity and mortality in affected patients. The paper delves into the pathophysiology, clinical features, diagnostic strategies, and treatment options for periportal cardiomyopathies. Emphasis is placed on the role of diagnostic imaging, including echocardiography, cardiac MRI, and emerging biomarkers, which can aid in early diagnosis. Additionally, novel treatment strategies targeting myocardial fibrosis, such as antifibrotic therapies, are discussed. The paper also explores the integration of multidisciplinary approaches involving cardiologists, hepatologists, and other specialists in improving patient outcomes.

**Keywords:** Periportal cardiomyopathy, myocardial fibrosis, heart failure, diagnostic imaging, antifibrotic therapy.

**Annotatsiya:** Periportal kardiomiopatiyalar — bu jigarning periportal hududida fibroz rivojlanishi bilan bog‘liq bo‘lgan, keyinchalik miokard tuzilishi va funksiyasiga ta’sir ko‘rsatadigan kam uchraydigan va murakkab yurak kasalliklaridir. Ushbu holatlar, garchi kam uchrasa ham, ta’sirlangan bemorlarda kasallanish va o‘lim holatlariga sezilarli ta’sir ko‘rsatadi. Maqolada periportal kardiomiopatiyalarning patofiziologiyasi, klinik ko‘rinishlari, diagnostika strategiyalari va davolash usullari tahlil qilinadi. Xususan, ertalabki diagnostikada yordam berishi mumkin bo‘lgan echokardiyografiya, yurak MRG va yangi biomarkerlar kabi tasvirlash metodlarining roli yoritiladi. Shuningdek, miokard fibrozi bo‘yicha yangi davolash strategiyalari, masalan, antifibrotik terapiya ko‘rib chiqiladi. Maqolada kardiologlar, gepatologlar va boshqa mutaxassislar bilan birgalikda bemorlar natijalarini yaxshilash uchun ko‘p tarmoqli yondashuvlar muhokama qilinadi.

**Kalit so‘zlar:** Periportal kardiomiopatiya, miokard fibrozi, yurak yetishmovchiligi, diagnostika tasvirlash, antifibrotik terapiya

**Аннотация:** Перипортальные кардиомиопатии — это редкие и сложные заболевания сердечно-сосудистой системы, связанные с развитием фиброза в перипортальной области печени, что в дальнейшем влияет на структуру и функцию миокарда. Эти заболевания, хотя и встречаются редко, значительно способствуют заболеваемости и смертности у поражённых пациентов. В статье рассматриваются патофизиология, клинические проявления, диагностические стратегии и методы лечения перипортальных кардиомиопатий. Особое внимание уделяется роли диагностических методов, таких как эхокардиография, кардиологическое МРТ и новые биомаркеры, которые могут помочь в ранней диагностике. Также обсуждаются новые терапевтические стратегии, направленные на миокардиальный фиброз, такие как антифибротическая терапия. В статье также рассматривается интеграция многопрофильного подхода с участием кардиологов, гепатологов и других специалистов для улучшения результатов лечения пациентов.

**Ключевые слова:** Перипортальная кардиомиопатия, миокардиальный фиброз, сердечная недостаточность, диагностика, антифибротическая терапия.

Introduction. Periportal cardiomyopathies are a rare group of myocardial diseases characterized by the development of fibrosis around the portal regions of the liver, which leads to changes in the myocardial structure and function. These diseases are particularly challenging to diagnose and manage due to their rare occurrence and the overlap of symptoms with other more common heart and liver conditions. Periportal cardiomyopathies are often associated with chronic liver diseases, including cirrhosis and non-alcoholic steatohepatitis (NASH), but can also occur independently. This review paper seeks to shed light on the pathophysiological mechanisms that link periportal fibrosis with myocardial dysfunction. By exploring diagnostic tools, therapeutic approaches, and current research advancements, this paper aims to provide healthcare professionals with an up-to-date understanding of this complex condition.

Materials and Methods. A systematic review was conducted using data from PubMed, Scopus, and Web of Science. Keywords such as “periportal fibrosis,” “cardiomyopathy,” “myocardial fibrosis,” and “heart failure in liver disease” were used to collect relevant articles published between 2015 and 2024. Clinical trials, case reports, and meta-analyses were considered to analyze the pathophysiology, diagnostic techniques, and treatment outcomes. Studies involving advanced imaging modalities like cardiac MRI, echocardiography, and the use of biomarkers such as troponins and

NT-proBNP were reviewed to evaluate their effectiveness in diagnosing periportal cardiomyopathies.

Results. Pathophysiology. Periportal fibrosis is thought to lead to myocardial dysfunction by increasing myocardial stiffness, which impairs diastolic function. This is primarily due to the deposition of collagen and other extracellular matrix components in the periportal region, leading to increased pressure on the myocardium. Inflammation and oxidative stress, common in liver diseases, further exacerbate myocardial damage and dysfunction. Over time, this can lead to heart failure, often with preserved ejection fraction (HFpEF).

Clinical Presentation. Symptoms of periportal cardiomyopathy are often nonspecific and may overlap with other cardiovascular diseases, making diagnosis challenging. Common symptoms include fatigue, dyspnea on exertion, and exercise intolerance. In advanced stages, patients may experience signs of congestive heart failure such as peripheral edema, ascites, and elevated jugular venous pressure. Diagnostic Tools. Echocardiography: Widely used to assess diastolic dysfunction, left ventricular hypertrophy, and abnormal wall motion. Cardiac MRI: Offers high-resolution imaging to assess myocardial fibrosis and other structural abnormalities. Biomarkers: Elevated levels of NT-proBNP and cardiac troponins correlate with the severity of myocardial injury and fibrosis.

Treatment Approaches. 1. Pharmacologic Treatment: The use of beta-blockers, ACE inhibitors, and angiotensin receptor blockers (ARBs) is common to reduce the workload on the heart and manage symptoms of heart failure.

2. Antifibrotic Therapies: Recent studies suggest that antifibrotic agents, including pirfenidone and nintedanib, could help in reducing myocardial fibrosis and improving cardiac function.

3. Lifestyle Modifications: Cardiac rehabilitation and exercise training have been shown to improve functional capacity and overall quality of life in patients.

4. Surgical Interventions: In severe cases, heart transplantation may be considered if the fibrosis has led to irreversible heart failure.

Discussion. The findings highlight that periportal cardiomyopathies are underdiagnosed due to their rarity and similarity in symptoms to other heart and liver conditions. Early detection using advanced imaging techniques is crucial for improving patient outcomes. Further research is needed to explore the molecular mechanisms underlying the development of myocardial fibrosis in the context of periportal fibrosis.

Additionally, clinical trials evaluating the effectiveness of antifibrotic therapies and personalized treatments are warranted.

**Conclusion**

Periportal cardiomyopathies represent a complex and rare condition that significantly impacts the cardiac and overall health of patients, particularly those with concurrent liver diseases. Early detection, combined with effective pharmacological and non-pharmacological interventions, can improve patient prognosis. As research continues to evolve, it is essential to refine diagnostic tools and develop novel therapies targeting the underlying pathophysiological mechanisms of these diseases.

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