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# Chronic exertional compartment syndrome in a patient with restrictive cardiomyopathy and portal hypertension

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**Abstract:** Chronic exertional compartment syndrome (CECS) is a condition that produces exercise-induced pain and swelling due to a transient increase in compartment pressures. It is thought to be due to muscle hypertrophy and is classically associated with young athletes under 30, overtraining, anabolic steroid use, and aberrant running biomechanics. We present a unique case of CECS in a patient without the traditional risk factors but rather diagnosed with cardiac cirrhosis and portal hypertension. This patient's exercise-induced bilateral leg pain met the CECS criteria for leg compartment pressure testing and was attributed to fluid retention secondary to his comorbidities. His symptoms significantly improved after initiating diuretic pharmacotherapy. Based on our literature review, there is a dearth of literature associating CECS with specific chronic cardiac or hepatic conditions as well as describing its incidence in these conditions.

**Keywords:** cardiomyopathy; cirrhosis; musculoskeletal; pain; sports medicine.

Chronic exertional compartment syndrome (CECS) is a condition that is classically associated with young adult athletes undergoing a rigorous training regimen particularly involving running [1, 2]. In this population, CECS is thought to be caused by muscle hypertrophy and transient increases

in muscle volume that occurs during exercise against noncompliant fascial tissue. This in turn elevates intra-compartment pressure, decreases arterial and venous blood flow, and results in an inability to meet metabolic requirements, leading to ischemia and pain [2, 3]. Another potential cause of elevated intra-compartment pressure is fluid accumulation, such as hemorrhage from trauma, which can lead to acute compartment syndrome [4]. However, although previously not well described, CECS can also present in patients with pre-existing poor vascular circulation. In this report, we present a unique case of CECS in a sedentary individual secondary to fluid retention from cardiac cirrhosis and portal hypertension. Based on our literature review, there is a dearth of literature associating CECS with specific chronic cardiac or hepatic conditions as well as describing its incidence in these conditions.

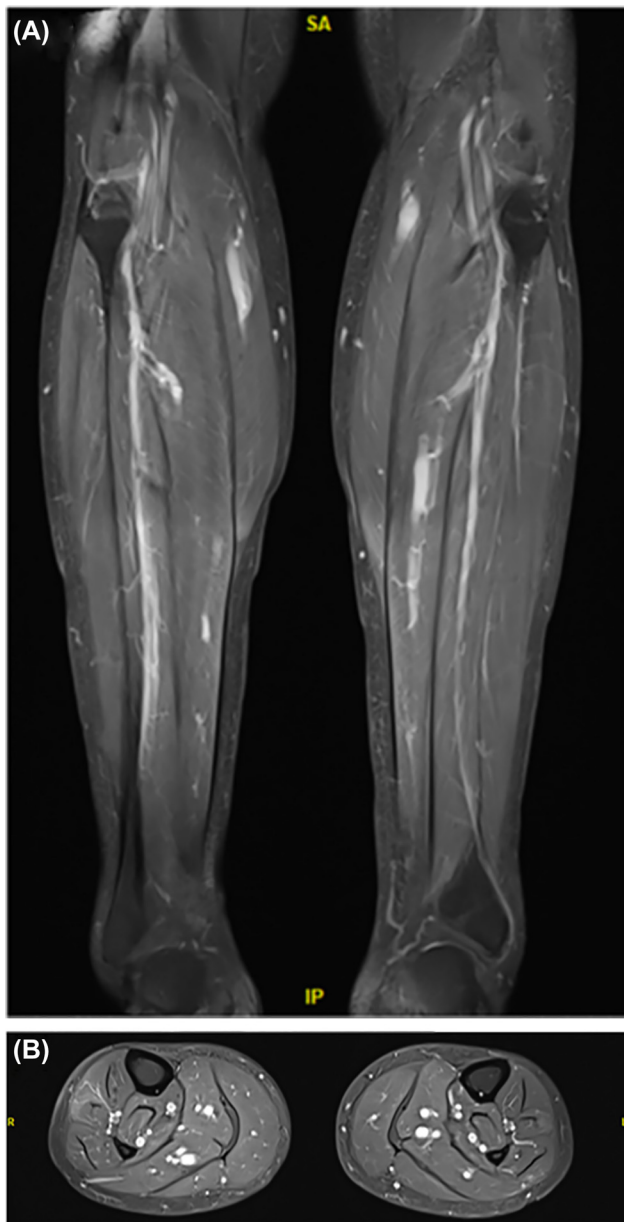
## Case description

A 28-year-old man was referred to the Physical Medicine and Rehabilitation (PM&R) clinic in October 2020 with a 4-month history of bilateral lower extremity pain. The patient had a suspected diagnosis of peripheral neuropathy. His medical history was significant for portal hypertension and idiopathic restrictive cardiomyopathy status-post heart transplant. His symptoms began without any inciting events and were exacerbated by walking. The pain was localized to the anterior and lateral legs without involving the feet or thighs. There was no associated paresthesia or weakness. On examination, the patient demonstrated full strength and range of motion in the upper and lower extremities. Sensation was intact throughout. There was no significant peripheral edema. Peripheral pulses were 2+ at the dorsal pedis bilaterally. Evaluation of the lumbar spine exam was unremarkable. Homan's sign was negative. Patellar and Achilles reflexes were unable to be elicited. The patient was able to ambulate a short distance in-office under observation, without assistive devices or onset of symptoms.

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**Figure 1:** MRI of the patient's bilateral lower extremities. (A) Coronal view, T2-weighted. (B) Axial view, T1-weighted with contrast. The MRI did not show any evidence of osteomyelitis. There was a diffuse muscle edema-like signal involving multiple compartments, for which differential considerations remained broad and included inflammatory, medication-related, and autoimmune etiologies in addition to possible compartment syndrome.

The patient underwent an extensive battery of diagnostic testing in vascular surgery, PM&R, and rheumatology clinics. Laboratory workup for various rheumatological conditions, lower extremity x-rays and Doppler ultrasound, ankle-brachial index, and nerve conduction studies/electromyography (NCS/EMG) were all unremarkable.

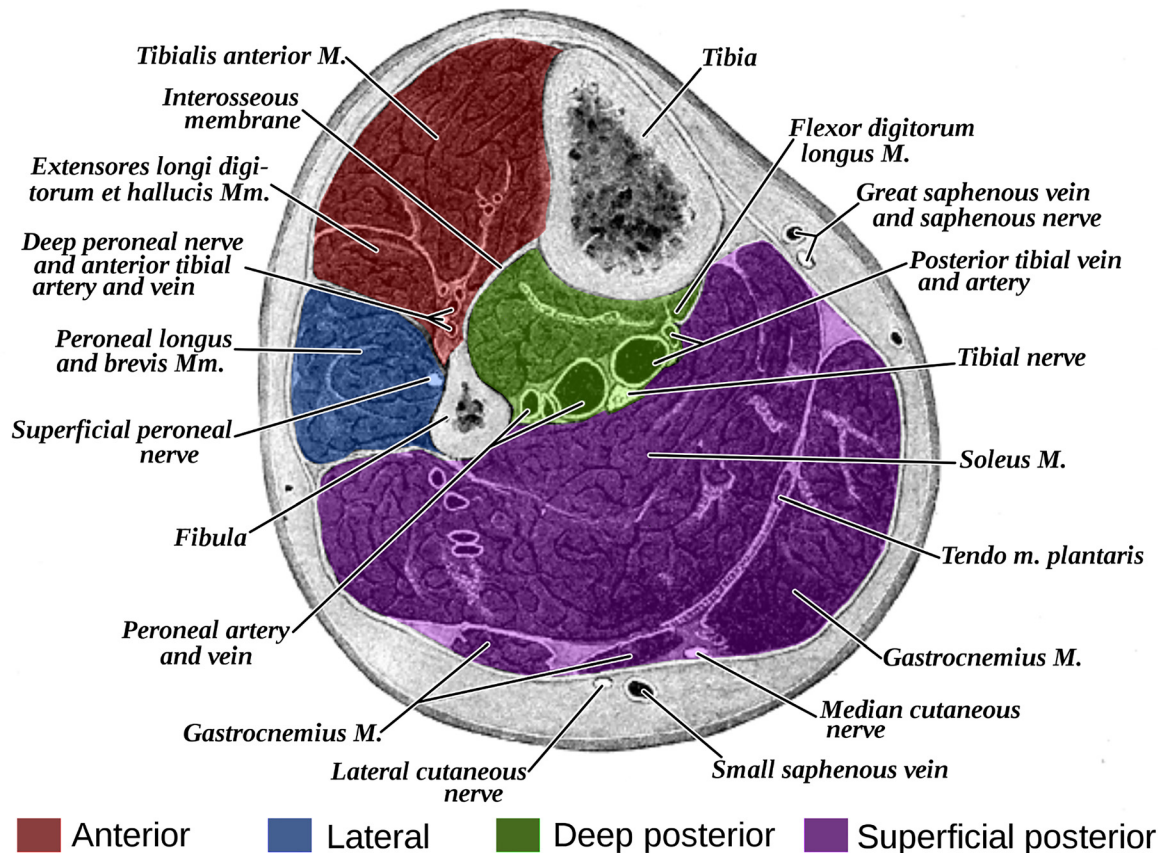
MRI of the right fibula and tibia demonstrated no evidence of osteomyelitis, but it did show diffuse muscle edema in all compartments, as seen in Figure 1, which could represent compartment syndrome; however, this nonspecific finding could also be attributed to inflammatory, medication-related, or autoimmune etiologies. Statin-induced myopathy was considered but thought to be unlikely because the patient had been on a stable dose without any recent addition of new medications. Due to the exertional nature of his pain, CECS was suspected, and compartment pressure testing was performed in the PM&R clinic. Pre-exertional measurements revealed the following pressures: Left Anterior 14 mmHg, Left Lateral 30 mmHg, Right Anterior 39 mmHg, and Right Lateral 35 mmHg. Detailed anatomy of the distal lower-extremity compartments can be seen in Figure 2.

Diagnosing CECS is often based on meeting one of the three Pedowitz criteria: (1) pre-exertional pressure >15 mmHg; (2) postexertional pressure (1 min) > 30 mmHg; or (3) postexertional pressure (5 min) >20 mmHg [5]. The patient had elevated pressures in the left lateral, right anterior, and right lateral compartments, consistent with CECS, which impacts the anterior or lateral compartments in 95% of cases [3]. Based on these criteria, the patient's pre-exertional compartment pressures alone met the criteria for CECS without the need for further post-exertional pressure testing.

The patient's underlying portal hypertension was thought to be related to his CECS. He was subsequently evaluated in the heart transplant clinic, where he was diagnosed with cardiac cirrhosis as the cause of the portal hypertension. He was started on a low-sodium diet and a diuretic regimen of torsemide 20 mg daily and eplerenone 25 mg daily. Over the next several weeks, he reported a 7 lb weight loss. This eventually led to complete resolution of his symptoms, which allowed him to return to his normal exercise activities.

## Discussion

Fluid shifts from the intravascular to extravascular space is a well-studied phenomenon in patients with restrictive cardiomyopathy or cirrhotic patients with portal hypertension. Restrictive cardiomyopathy can increase inferior vena cava pressures, resulting in increased hydrostatic pressure and fluid extravasation [6, 7]. Cirrhotic patients have increased salt and water retention due to activation of the renin-angiotensin-aldosterone system and decreased albumin production



**Figure 2:** Anatomy of the distal lower extremity compartments. Cross-section through the middle of the leg (with permission from Gray, Henry, and Warren H. Lewis. *Anatomy of the Human Body*. Plate 440, Lea & Febiger, 1918).

[8]. We hypothesize that increased extravascular volume in conjunction with noncompliant fascia elevated this patient's compartment pressures. As the patient was diuresed, this extravascular fluid was likely reduced, leading to decreased compartment pressures and an improvement in symptoms.

Standard management of classic CECS involves both nonsurgical and surgical approaches. Nonsurgical modalities include physical therapy, orthotics, and limiting physical activity. Gait modification and botulinum A injections may also be considered [1, 9]. Management with surgical fasciotomy appears to have better outcomes compared to nonsurgical approaches, especially if a patient wishes to return to their prior level of activity [1–3]. Open fasciotomy is the traditional approach, although surgeons may opt to perform endoscopy-assisted and single minimal-incision techniques.

In this patient, diuretic management was sufficient for improving his exercise-induced lower extremity pain given his chronic comorbidities. In the United States, there are an estimated 4.5 million adults with chronic liver disease and cirrhosis, and an estimated

6.2 million adults with heart failure [10, 11]. These patients are frequently encountered in clinics. Having an appropriate understanding of the pathophysiology of these chronic conditions can allow for appropriate conservative management without requiring more interventional approaches.

One limitation of this case report is that postdiuretic compartment pressure testing was not performed. The patient demonstrated complete resolution of his symptoms with diuretic therapy. Compartment pressure testing is an invasive and painful test for the patient. As such, the decision was made that repeat compartment pressure testing would cause unnecessary discomfort for the patient.

## Conclusions

To our knowledge, CECS secondary to fluid retention from cardiac cirrhosis and portal hypertension has not been described previously in the literature. Although

the prevalence of CECS in a population with underlying cardiovascular and hepatic conditions is not well elucidated, it should be considered in patients presenting with exertional lower extremity pain and these comorbidities. Pharmacologic management can contribute significantly to improving symptoms and overall quality of life without requiring more invasive procedures. Ultimately, this case emphasizes the importance of understanding the pathophysiology of these chronic conditions seen in a PM&R clinic and the importance of coordinated multidisciplinary care.

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**Author contribution:** All authors provided substantial contributions to conception and design, acquisition of data, or analysis and interpretation of data; all authors drafted the article or revised it critically for important intellectual content; A.J.D.L. gave final approval of the version of the article to be published; all authors agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

**Competing interests:** None reported.

**Disclaimers:** Dr. De Luigi, who is a Section Editor for *Journal of Osteopathic Medicine*, was involved neither in the peer review of this manuscript nor the decision to publish it.

**Informed consent:** The patient described in this report provided written informed consent.

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