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Original Article

Compartment syndrome in patients with haemophilia

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ABSTRACT

Background: Acute compartment syndrome (ACS) is an uncommon but potentially devastating condition.

Methods and results: There are scattered case reports and case series in the literature of ACS in persons with haemophilia (PWH), and even fewer in PWH and inhibitors. The management of compartment syndrome in these scenarios is controversial and often anecdotal. In addition haematological outcomes are frequently quoted but functional outcomes are generally overlooked.

This article aims to provide an overview of ACS and its contemporary management. We also review the literature and outcomes of patients with haemophilia who develop ACS in an effort to assess the best treatment modality.

Conclusion: In the majority of cases ACS settles with normalisation of the clotting cascade. Specialist haematological input is mandatory before surgical intervention should be considered, especially in PWH and inhibitors.

1. Introduction

Richard von Volkmann first described compartment syndrome in 1881.[1] He suggested that paralysis and contracture came on simultaneously as result of an interruption to the blood supply of the affected muscles. The first surgeon to reproduce ischaemic contracture in animals was Paul Jepson in 1924[2] whilst working at the Mayo Foundation. He also demonstrated that prompt surgical decompression could prevent these contractures.

Acute compartment syndrome (ACS) is defined as a critical pressure increase within a confined compartmental space causing a decline in the perfusion pressure to that compartment's tissues.[3–6] It can occur with any elevation in interstitial pressure in a closed osseo-fascial compartment. This causes microvascular compromise and a reduction in the perfusion gradient to below a critical value, leading to ischaemia of the tissue within that compartment. ACS is now considered a surgical emergency warranting prompt evaluation and treatment.

Any internal or external event that increases intracompartmental pressure can potentially cause a compartment syndrome. The incidence is thought to be 3.1 per 1,00,000 population, with males ten times more commonly affected than females. By far the commonest cause is trauma, and in particular tibial shaft fractures (which are more common in men). Bleeding disorders, and in particular

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haemophilia, are recognised, albeit rarer causes, which put these patients uniquely at risk of ACS often with only trivial trauma. Indeed eighty five percent of bleeding in patients with haemophilia occurs within musculoskeletal tissue. In addition a recent systematic review showed that 21% of patients with acute compartment syndrome of the thigh had coagulation defects.[7]

1.1. *Overview of compartment syndrome*

ACS is characterised by a critical pressure increase within a confined compartmental space causing a decline in the perfusion pressure to the compartment tissue.[3–6] It can occur with any elevation in interstitial pressure in a closed osseofascial compartment. This causes microvascular compromise and a reduction in the perfusion gradient to below a critical value, leading to ischaemia of the tissue within that compartment. Fig. 1 below displays the cycle of events and the development of acute compartment syndrome.

Approximately 40% of all ACSs occur after fractures of the tibial shaft[8] with an incidence in the range of 1%–10%.[9] The next commonest location is in the forearm, but almost any compartment can be affected: arm,[10] thigh,[11] foot,[12] buttock,[13] hand,[14] and abdomen.[15] Any internal or external event that increases intra-compartmental pressure can cause a compartment syndrome. Table 1 below shows some of the more common causes.

Rorabeck reported almost complete recovery of limb function if fasciotomy was performed within six hours.[16] When fasciotomy was performed within twelve hours normal limb function was regained in only 68% of patients; after twelve hours only 8% regained normal function.[17]

With late diagnosis irreversible tissue ischaemia develops causing potentially disastrous neurological deficits, muscle necrosis, ischaemic contracture, infection, chronic pain, delayed fracture union,[18] rhabdomyolysis,[17] amputation and even death.

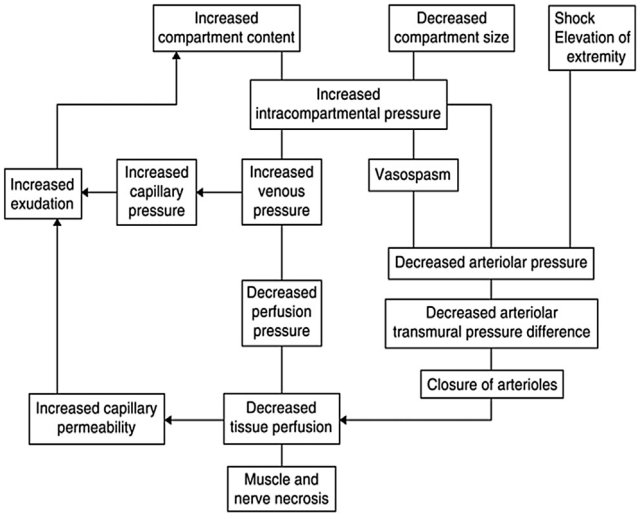

Fig. 1 – Pathophysiology of compartment syndrome. ***Reproduced with permission: Prasarn M, Oulette E. J Am Acad Orthop Surg 2011; 19:49–58.***

Table 1 – Common causes of ACS.

Fracture	Burns
Crush injury	Infection
Injection injury	Bleeding disorders
Penetrating trauma	Arterial injury
Constrictive dressings	Reperfusion
Casts	Extravasation of drugs

ACS is a clinical diagnosis; the most important determinant of outcome is early recognition and expeditious surgical intervention. Where doubt remains the intra-compartmental pressures can be measured. Marginal pressure readings should be repeated with further compartment pressure readings and repeat physical exam. A compartment pressure of 30 mmHg less than the diastolic blood pressure is often quoted beyond this capillary pressure is insufficient to maintain muscle blood flow and the fascia has reached its maximum limitation of stretch.[19]

The treatment of ACS is immediate removal of all circumferential dressings down to the skin, usually followed by open fasciotomy. The limb should be kept at the level of the heart and not be elevated in the interim (or if the diagnosis is in doubt), as this decreases arterial flow and narrows the arterial-venous pressure gradient.[20]

2. Compartment syndrome in haemophilia

Compartment syndrome is well described in persons with haemophilia (PWH) and remains a significant challenge for the haematologist and the orthopaedic surgeon. The lack of a clear aetiology or conspicuous traumatic event can contribute to the diagnostic difficulty, whilst the possibility of catastrophic bleeding makes fasciotomy potentially disastrous.

Haemorrhages located within the calf and anterior forearm represent the highest risk for the development of compartment syndrome.[21] In patients without bleeding disorders the treatment of choice is fasciotomy, even when in doubt to prevent the establishment of motor and sensory loss, contracture and severe extremity impairment. The role of fasciotomy in haemophilia has been downplayed and care largely focuses on haemostatic manoeuvres.[22]

The first step in the management of a suspected compartment syndrome should be sufficient substitution of clotting factors, which may help lower the compartment pressure.[23] Bleeding will typically tamponade before the patient experiences a substantial drop in haemoglobin. However, raised compartment pressures can cause significant morbidity in the extremities. Given the short window between symptomatic onset and tissue necrosis, early intervention is crucial and may prove to be not only limb-saving but also life-saving.

If manoeuvres to normalise the clotting derangement fail, fasciotomy is often performed to prevent muscle necrosis, joint contracture and long-term sequelae.

There are no specific pre-operative guidelines for factor replacement levels during fasciotomy, but the authors recommend 100% of normal. Infection is a significant complication usually related to persistent bleeding.