

# Multiple Limb Compartment Syndromes in a Recruit With Sickle Cell Trait

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### Abstract

**Sickle cell trait, as opposed to the disease, is a rare condition with fewer medical complications. We present a case of a 24 year old Army recruit, who required multiple fasciotomies for limb compartment syndrome, associated with sickle cell trait. We discuss the management, complications and screening programmes of the condition and make suggestions for the training of sickle cell trait personnel in planning a career in the Armed Forces.**

### Introduction

Sickle Cell Anaemia (SCA) is a widely known disease with significant morbidity and a reduced life expectancy [1]. Sickle cell trait (SCT) is often not perceived as a disease *per se* due to its milder form, with few complications [2]. When complications do occur they are significant and there are reports of rhabdomyolysis [3], compartment syndrome and even sudden death from SCT[4]. We present a case of exercise induced upper and lower limb compartment syndrome, in a recruit of Afro-Caribbean origin from previously undiagnosed SCT.

### Case Report

A 24 year old recruit, from Grenada, collapsed with bilateral thigh pain during a 1.5 mile training run, in his first week of Army recruit training. On arrival at the medical centre he was tachycardic (185 beats/minute), tachypnoeic (24 respirations/minute) and hypotensive (97/53mmHg). The differential diagnosis on transfer to a regional hospital included heat exhaustion and hypertrophic obstructive cardiomyopathy. On examination he had a tender tense right thigh, with good distal pulses and frank haematuria. Following fluid resuscitation his observations had improved to a heart rate of 123 beats/min, blood pressure of 124/66mmHg, a respiratory rate of 20/minute; he was acidotic (pH7.23) with a raised white cell count of 18 and a creatinine kinase (CK) of 198 units. He had normal renal function and a normal echocardiogram. Compartment pressures were recorded by the orthopaedic team to rule out compartment syndrome as he had tense thigh compartments. He had raised compartment pressures of 50mmHg in vastus lateralis and 60mmHg in biceps femoris, confirming the diagnosis of acute compartment syndrome.

He was taken urgently to the operating theatre and underwent bilateral thigh and gluteal compartment fasciotomies. A curved incision from the posterior superior iliac spine to greater trochanter on either side was used to expose and decompress the gluteal compartment. The same incision was extended distally down to the lateral femoral epicondyle and the iliotibial band was divided along the line of the skin incision. The anterior compartment was

under tension and the muscles were still viable and bulged out on dividing the fascia lata. Vastus lateralis was reflected off the lateral intermuscular septum, which was also divided along the length of skin incision, thus decompressing both anterior and posterior compartments of the thigh. The medial compartment of the thigh was not decompressed. A vacuum assisted closure (VAC pump) device was applied to both thigh wounds and he was transferred to the Intensive Therapy Unit (ITU). His urine was positive for myoglobin, and the CK had risen to 9,970 units. On the advice of a haematologist he had sickle cell screening and electrophoresis and was diagnosed to have sickle cell trait (SCT).

On the first post operative day, CK rose further to 163,400 units and he began to complain of left forearm and wrist pain. Examination revealed an acute compartment syndrome of his forearm and he underwent urgent hand and forearm compartment decompression, however this was extended proximally intra-operatively to include the upper arm, due to increased pressure noted in that compartment. He was managed in ITU for six days following his initial presentation, requiring aggressive fluid replacement and treatment for mild renal impairment, although this quickly rectified without the need for more invasive treatment or renal replacement therapy.

Over the following week, he underwent two further compartment inspections and wound closures in theatre (Figures 1 and 2). His CK dropped rapidly and he made a full recovery and was discharged within two weeks of initial presentation.



**Figure 1.** Closure of the wounds on dorsum of the left hand using clips and sutures

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**Figure 2.** Progressive delayed closure of the left forearm wound with a bootlace technique.

At his latest follow up, six months post fasciotomy he had had an unremarkable recovery without neurological deficit or contractures. He decided to leave Army training and returned to the Caribbean.

### Discussion

Compartment syndrome is defined as an elevation of the interstitial pressure in a closed anatomical space, leading to muscle and nerve ischaemia. The diagnosis is a clinical one, although can be assisted using devices such as a compartment pressure monitor. Immediate surgical decompression is essential to prevent rhabdomyolysis and acute renal failure (ARF) in the acute setting and contractures in the longer term. Rhabdomyolysis is the rapid breakdown of skeletal muscle due to injury to muscle tissue. This leads to a release of breakdown products such as myoglobin, which is nephrotoxic. Hypovolaemia/dehydration and aciduria are important contributing factors in the development of myoglobinuric ARF [5]. Three mechanisms predominate in haem protein toxicity: renal vasoconstriction with diminished renal circulation, intraluminal cast formation and direct haem protein-induced cytotoxicity. In the absence of hypovolaemia and aciduria, effects are minimal but become significant in their presence [6]. The most sensitive laboratory test is the elevation of CK, which rises within 12 hours of the onset of myocyte injury, peaks at 1-3 days and declines 3-5 days after the cessation of muscle injury. A peak CK level (>6000IU/L) may be predictive of the development of renal failure [5] and persistently elevated CK level should raise the suspicion of ongoing muscle injury.

An important feature of this case is the need to exclude compartment syndrome at other sites than that first identified, especially if there is a systemic/generalised cause for compartment syndrome. This requires particular attention in the unconscious or ventilated patient. We also would like to emphasise that acute compartment syndrome should be considered in any sportsperson who presents with an acutely swollen and very painful limb without a traumatic fracture [7,8].

The risk in athletes and military personnel, with SCT, of sudden death has been documented [4], although there are few reports of compartment syndromes from SCT [3,9]. With a prevalence rate of 8% of the haemoglobin AS gene (SCT) in African Americans, practice in the US military is variable: the US Army stopped screening for the sickle gene in 1996; the US Marine Corps screen all individuals but do not alter the routine of those with SCT; the US Air Force screen everyone and offers the option to decline service to each recruit positive for SCT and the

US Navy screens recruits and identifies those with SCT by a neck tag and a red belt during strenuous exercise drills [10]. The US National Collegiate Athletic Association (NCAA) is the largest collegiate athletic association in the world and it recommends that all member institutions screen its athletes for SCT [11]. It is known that people with SCT are at greater risk of complications with dehydration, exercise and at altitude. This should be borne in mind when considering the training of both recruits and exercising military personnel. This case, therefore, raises the question as to whether there should be a screening programme for SCT in Afro-Caribbean personnel in the UK military, similar to that of the US Navy. A recruit with SCT in training can then be highlighted to both the training and medical personnel in order to reduce the risk of either compartment syndrome or sudden death.

### Conclusion

Compartment syndrome in the upper limb or gluteal/thigh regions can be easily overlooked due to its infrequent presentation and the size of compartment, in the absence of overt trauma. It is imperative to consider this diagnosis in patients with SCT, even in the absence of a fracture, to avoid potential devastating consequences. It is also important to assess other compartments if there is a systemic/generalised cause for compartment syndrome.

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