Letter to the Editor re: Clinical Significance of Mottling Rashes in Diving Decompression Sickness

Dear Editor:

A review by Breen et al. identifies a frequent (>50%) association of the cutis marmorata (CM) skin rash with more serious neurological manifestations in decompression sickness (DCS).[1] On this basis they advocate "aggressive and immediate medical attention" for CM, by which they appear to mean "immediate evacuation and recompression," even if CM is the only presenting symptom. This contradicts the advice of two international consensus guidelines that classify rash (including CM), of itself, as a "mild" symptom and legitimize treatment without recompression provided more serious manifestations are absent.[2,3] Those guidelines stipulate caveats, such as strong advocacy for a competent neurological examination, before designating a case as "mild," which helps ensure that serious manifestations are not overlooked.

The potential for CM to appear in association with serious neurological symptoms of DCS is not new knowledge and is unsurprising given common elements of pathophysiology (right-to-left shunting of venous bubbles). This was well understood by the experts who contributed to the present guidelines.[2,3] Equally, those experts also recognized that CM frequently occurs with no serious DCS symptoms. It is difficult to understand the present suggestion to designate the latter cases as a medical emergency, particularly since this would, for example, mandate a hugely expensive and potentially hazardous aeromedical evacuation of a patient from a very remote location for treatment of a rash. Isolated CM usually responds extremely well to treatment with surface oxygen. Indeed, all isolated cases reported by Breen et al. who received only surface oxygen (or no treatment whatsoever) completely recovered.

There are several counterarguments I wish to preempt. First, one could argue that delaying evacuation after noticing CM to see if serious symptoms develop might compromise outcome. However, latency in serious neurological DCS is very short (50% of cases appear with 10 min and 98% within 1 h),[4,5] and CM would therefore rarely be the first symptom noticed. Moreover, there is mounting evidence that once recompression latency exceeds 3–6 h, further delay makes little difference to the final outcome.[6] The duration of evacuations from remote locations often vastly exceeds this threshold, so small delays confirming the presence of serious symptoms are unlikely to be important. Second, as evidence of severity, Breen et al. cite the hypothesis that CM could arise as a neurogenic response to cerebral arterial gas embolism, but this is highly implausible in the absence of catastrophic neurological symptoms and would, therefore, not explain an isolated rash.[7]

The mild DCS guidelines[2,3] and other recent expert reviews[8,9] stipulate that divers with mild symptoms (including rash) should ideally be recompressed if a suitable chamber is readily accessible, for example, by road transport. However, the alternative option not to recompress mild DCS where chamber access is difficult has profoundly simplified management of remote cases for 20 yr now with no literature signal suggesting this approach is flawed. Breen et al. provide a valuable reminder that serious neurological manifestations may accompany CM, but their findings don't justify change to international expert consensus guidelines on managing DCS with CM but without serious symptoms.

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In Response:

We appreciate Professor Mitchell's thoughtful comments regarding our paper.

Professor Mitchell points out that prior guidelines do not regard cutaneous diving decompression sickness (DCS) as a severe symptom. We call those guidelines into question, based on the shifting paradigms as we note in our manuscript.

Professor Mitchell asserts that isolated cutis marmorata (CM) generally responds well to treatment with surface oxygen. While historically this may be true, the clinical picture is muddied by a lack of clear distinction in the field between CM and its close benign mimickers livedo reticularis and aquagenic or cold-induced urticaria. This lack of clinical diagnostic rigor may lead to underestimation of the severity of CM.

Unfortunately, that clinical ambiguity makes it difficult to interpret reporting. Despite that, in our systematic review, we identified a strong association between right-to-left shunts and neurological sequelae in patients exhibiting signs of cutaneous DCS or CM. Of the patients with documented workup, 84% showed evidence of right-to-left shunts with CM. Of the patients with documented neurological evaluations, 57% experienced both CM and neurological DCS manifestations.[1]

Accordingly, we called for a reappraisal of its clinical significance to rethink this clinical sign and consider potential reclassification. While we recommended aggressive and immediate medical attention, we did not intend this to mean immediate evacuation and recompression, but rather intensive monitoring and potential evacuation depending on clinical exam and context.

Professor Mitchell points out that all cases reported in our study who received only surface oxygen (or no treatment whatsoever) recovered. However, it is critical to note that there were fatalities among patients with cutaneous DCS who went on to develop severe neurological or cardiopulmonary complications. There were also cases of cutaneous DCS who improved with hyperbaric oxygen therapy. While all cases of isolated CM (not otherwise clarified) mentioned in our review recovered with surface oxygen or spontaneously, it must be acknowledged that there are conflicting data regarding the importance and influence of early recompression in Type II DCS on outcomes (Type II DCS is the severe class of DCS).[2–4] Therefore, while some patients with CM may improve spontaneously or with minimal intervention, that is also true with patients having more severe Type II DCS.

In conclusion, we appreciate Professor Mitchell's meaningful points. We also recognize that the reviewed literature data indicate merely a high co-association with other severe DCS symptoms and CM, and we readily acknowledge that more research is needed to delineate its significance. While we agree that changes to international consensus guidelines may not be justified at this juncture based on novel data from a literature review, the data would suggest clinical vigilance and heightened preparedness and deliberate diagnostic precision in assessment of CM. Accurate assessment of cutaneous findings may be critically important for patient management. Hartig et al. recently suggested that CM be treated as a life-threatening systematic vascular disorder based on their theories of the pathomechanism, venturing farther to state that such a vascular disorder "should always be treated with hyperbaric oxygen therapy."[5]

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