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

Dear Editor:

Breen and colleagues conducted a systematic review of reports of cutaneous decompression sickness (DCS) to investigate its relationships with right-to-left shunts (RLS) and with neurological DCS.[1] They found association with RLS. They concluded that the findings support the hypothesis that the pathogenesis involves paradoxical gas embolism. These are not new findings. My colleagues and I were the first to suggest paradoxical gas embolism in the causation of both neurological and cutaneous DCS.[2–6]

I have a number of concerns about the paper by Breen et al. The first is that Breen et al. did not identify all relevant publications that reported cutaneous DCS. Breen et al. identified 128 patients with cutaneous DCS.[1] Their review is critically dependent on a case-control study by my colleagues and me, which provided 61 of the 128 patients.[5]

In our paper,[5] we referred to our earlier original reports of the role of RLS in both neurological and cutaneous DCS.[3,4] We stated: "We report here the findings from a consecutive series of a further 61 individuals with cutaneous decompression illness referred to us for investigation since our earlier report of 14 cases (raising the total number of cases reported by us to 75)."[5] The 14 divers reported earlier had more than 29 episodes of cutaneous DCS and 12 of the 14 divers had an RLS.[4] The failure to include the data from our earlier report, despite it being mentioned clearly in the paper cited by Breen et al. makes me wonder how carefully the authors read our paper.

In addition, Breen et al. did not mention our subsequently reported 200 divers who had transcatheter closure of an atrial shunt after one or more episodes of DCS, of whom 99 had cutaneous DCS.[7] I wonder how many reports of cutaneous DCS by other investigators were missed by Breen et al.

My second concern is that Breen et al. have misinterpreted data reported, which has caused them to overemphasize the temporal relation between cutaneous and neurological DCS. For example, in response to the letter from Mitchell,[8] Breen responded: "Of the patients with documented neurological evaluations, 57% experienced both cutis marmorata and neurological DCS manifestations."[8] That gives the misleading impression that 57% of divers who had cutaneous DCS had neurological DCS at the same time.

Our report on 61 divers who had cutaneous DCS described the results of bubble contrast echocardiography and noncutaneous manifestations of DCS in all the divers.[5] We reported that 29 divers had cutaneous DCS on a single occasion and 32 divers had multiple episodes, including 16 divers that had 4 or more episodes.[5] We also reported that 35 of the divers (29 with a shunt) had neurological DCS.[5] However, we clearly stated that cutaneous DCS and noncutaneous DCS often occurred on separate occasions.[5]

The other 67 patients in the systematic review by Breen et al. were reported in 5 smaller case series and 25 single case reports.[1] For only 11 of the 67 patients were both the results of tests for an RLS and occurrence of noncutaneous DCS reported.[1] Each of the 11 patients had an RLS and 7 had neurological DCS.[1] As we found, cutaneous and neurological DCS did not always occur after the same dive. For example, Schwob et al. reported a diver who had multiple episodes of cutaneous DCS over 8 yr and had a single episode of concurrent cutaneous DCS with dizziness, blurred vision, and headache, which may have been neurological DCS.[9] Azzopardi et al. and Kemper et al. each reported divers with multiple episodes of cutaneous DCS but no neurological manifestations.[10,11] From all these reports, it should be clear that far fewer than 57% of episodes of cutaneous DCS were associated with concurrent neurological DCS.

Of the 38 patients for whom there was no information about an RLS, 21 had neurological DCS.[1] Therefore, our report also provided 35 of the 63 patients who had neurological DCS and 17 of the 18 patients who had transcatheter closure of an atrial shunt included in the review by Breen et al.[5]

In more than 35 yr since our first reports of association between DCS and RLS, I have seen many hundreds of divers who had many thousands of episodes of DCS; several hundreds have had transcatheter closure of atrial RLS. Several amateur divers who I have seen had more than 10 episodes of cutaneous

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DCS before seeking referral, and some of those only sought medical advice after their first episode of neurological DCS.

In addition, my colleagues and I reported that 47 of 61 (77%) divers with one or more episodes of cutaneous DCS had an RLS compared with 34 of 123 (27.6%) control divers ($P < 0.001$).[5] The size of the shunts in the divers with cutaneous DCS was significantly greater than in the controls.[5] Cutaneous DCS occurred after dives that were provocative or deep in divers without shunts, but after shallower and nonprovocative dives in those with shunts.[5]

Breen et al. reported that Hartig et al. demonstrated a 100% RLS prevalence in a study of 18 divers with cutaneous DCS.[12] However, that was likely the result of the selection criteria for their retrospective review of cases, because Hartig et al. excluded all divers whose illness occurred after dives with possible deviations from safe decompression procedures.[12] Had they included all divers who had cutaneous DCS, I suspect that the majority of those who had cutaneous DCS after unsafe dive profiles would not have had an RLS, as we found. Breen et al. also stated that a study by Germonpre and colleagues implicated patent foramen ovale in nearly 100% of cases of cutaneous DCS when the paper was a hypothesis paper that provided no numerical data on the link between patent foramen ovale and cutaneous DCS.[13]

My third concern is that a number of statements by Breen et al. suggest a lack of understanding of diving physiology and gas nucleation. For example, Breen et al. wrote: "Echocardiography and transcranial Doppler studies used to search for right-to-left shunts carry high false negative results, since high intrathoracic pressures in hyperbaric conditions cannot be replicated with simple induced Valsalva during testing, further underestimating the prevalence."[1] That comment makes no sense because although pressures are increased when underwater, right and left atrial pressures are increased equally.

They also wrote: "Valsalva to equalize Eustachian pressures during ascent may contribute to transient right-to-left shunt."[1] If maneuvers are required to equalize middle ear pressure, it is virtually always on descent, when there is no risk of bubble formation, rather than during ascent. Irrespective of that, for most dives venous bubble nucleation occurs after surfacing so that risk of paradoxical gas embolism is also after surfacing.

However, I believe that if a diver has either cutaneous or neurological DCS, the possibility that there is an RLS should be excluded by appropriate testing.[14]

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In Response:

We appreciate Dr. Wilmshurst's experience in the domain of paradoxical embolism and his comments regarding our publication.[1] The critical point raised by our work is that cutaneous decompression sickness (DCS) (livedo racemosa) must be distinguished both clinically and mechanistically from its close benign dermatological mimickers, livedo reticularis and aquagenic, or cold-induced, urticaria.

Dr. Wilmshurst suggests additional articles for consideration in our systematic review.[2–4] We conducted a PRISMA-compliant systematic review in collaboration with the Department of Library Sciences at Mayo Clinic to develop keywords and medical subject headings (MeSH) to optimize search results and maximize power of the study. One of these studies did not meet inclusion criteria, as presented data regarding DCS episodes did not stratify by presence of cutaneous manifestations.[4] Two of the studies were not captured by the search strategy.[2,3] The second study[3] incorporates data presented from the earlier study.[2] We have integrated the suggested data into our review and updated the results in **Table I**.

Table I. Interrelationships Among Right-to-Left Shunt, Neurological Symptoms, and Cutaneous DCS.

STUDY	STUDY TYPE (NO. OF PATIENTS)	CLINICAL PRESENTATION	RIGHT-TO-LEFT SHUNT	NEUROLOGICAL EVENTS	RECOVERY
Avivi (2022)	Case report (1)	Skin rash, portal venous gas emboli	Unknown	Yes	Significant after hyperbaric oxygen therapy
Azzopardi (2021)	Case report (1)	Skin rash only	Yes	No	Full after hyperbaric O_2
Dziewiatowski (2021)	Case report (1)	Skin rash, numbness in limb, vertigo	Yes	Yes	Full after hyperbaric O_2
Magri (2021)	Case series with matched controls (7)	Skin rash, neurological involvement (2/7), multisystem involvement (3/7)	Unknown	Yes 2/7	Full after hyperbaric O_2
Arjomand (2020)	Case report (1)	Loss of consciousness, shock, skin rash, severe hypoxia	Unknown	Yes	Residual DCS-related neurological deficits after hyperbaric O_2
Selesny (2020)	Case report (1)	Skin rash, lightheadedness	Unknown	No	Full with hyperbaric O_2
Yu (2020)	Case report (1)	Skin, cardiac, joint, abdominal, respiratory, cardiovascular	Unknown	No	Full after hyperbaric O_2
Hartig (2020)	Case series (18)	Skin	Yes 18/18	Unknown	Unknown
Schwob (2020)	Case report (1)	Skin rash	Yes	Yes	Unknown
Garcia (2020)	Case series (4)	Skin rash only	Yes 4/4	Yes 4/4	Full after hyperbaric O_2
Yount (2019)	Case report (1)	Skin, neurological, cardiovascular, renal	Unknown	Yes	Full after hyperbaric O_2 and ICU supportive care
Lau (2019)	Case report (1)	Skin rash only	Unknown	No	Full
Kerut (2019)	Case report (1)	Skin rash only	Yes	No	Full after hyperbaric O_2
Strauss (2019)	Case report (1)	Skin rash only	Unknown	Yes	Full after high flow 100% O_2
Rozenblat (2018)	Case report (1)	Skin rash only	Unknown	No	Full (spontaneously, no intervention)
Mendez (2017)	Case report (1)	Skin, neurological, cardiopulmonary	Unknown	Yes	Fatality
Sun (2017)	Case report (1)	Skin, portal venous	Unknown	No	Full after hyperbaric O_2
Perez-Lopez (2015)	Case report (1)	Skin rash only	Yes	No	Full (spontaneously, no intervention)
Jitsuiki (2015)	Case report (1)	Skin, neurological, cardiopulmonary	Unknown	Yes	Fatality
Kemper (2015)	Case report (1)	Skin rash only	Yes	No	Full after 100% O_2
Tasios (2014)	Case report (1)	Skin rash only	Unknown	No	Full after hyperbaric O_2
Modell (2014)	Case report (1)	Skin, neurological, lymphatic	Yes	Yes	Full after hyperbaric O_2
Wen (2013)	Case report (1)	Skin, cardiopulmonary, neurological	Unknown	Yes	Fatality
Oode (2013)	Case report (1)	Skin, cardiopulmonary, neurological	Unknown	Yes	Fatality
Akca (2012)	Case report (1)	Skin, neurological	Unknown	Yes	Full after hyperbaric O_2
Sundal (2011)	Case series (10)	Skin, joints, neurological	Unknown	Yes 7/10	Unknown
Mutluoglu (2011)	Case report (1)	Skin, joints, neurological	Unknown	Yes	Full after hyperbaric O_2
Bledsoe (2011)	Case report (1)	Skin, musculoskeletal	Unknown	No	Full after hyperbaric O_2
Kalentzos (2010)	Case report (1)	Skin, neurological	Unknown	Yes	Full after hyperbaric O_2
Wilmshurst (2001)	Retrospective case-control (61)	61/61 skin, 35/61 neurological, 3/61 cardiovascular, 12/61 musculoskeletal	Yes 47/61	Many of the divers had neurological symptoms but not necessarily concurrent with the dive manifesting cutaneous symptoms and therefore numeric data cannot be presented	Full
Li (2001)	Case series (3)	Skin, joints, neurological	Unknown	Yes	Full after hyperbaric O_2
Wilmshurst (1990)	Retrospective case-control (14)	Skin, among other manifestations	Yes 12/14	Many of the divers had neurological symptoms but not necessarily concurrent with the dive manifesting cutaneous symptoms and therefore numeric data cannot be presented	

DCS: decompression sickness.

The added data did not materially change the results or analysis of our study, nor did they change the conclusion or central message of our publication. The addition of the study[3] increased the pooled total of patients from 128 to 142. In total, 88 patients had right-to-left shunt (RLS), while 16 had no shunt and 38 were unknown. Of the patients with documented RLS workup (N = 104), 85% (88/104) showed evidence of RLS in addition to DCS skin manifestations. Our original results reported that in patients with documented RLS workup (N = 90), 84% (76/90) showed evidence of RLS in addition to DCS skin manifestations.[1]

We also appreciate Dr. Wilmshurst's clarification about the 61 divers discussed in his 2001 publication[5]: that cutaneous DCS and noncutaneous DCS events often occurred on separate occasions. This clarification does not change our results nor interpretation in our study. The data analysis was updated to account for this clarification: of the patients with documented neurological evaluations (N = 49), 57% (28/49) experienced skin and neurological DCS manifestations. Our original results reported that in patients with documented neurological workup (N = 110), 57% (63/110) experienced both skin and neurological DCS manifestations.[1] As we stated, "The reported instances of RLS and neurological symptoms were frequently coincidental (see **Table I**); the exact percentages of overlap cannot be stated due to lack of data availability."[1]

Dr. Wilmshurst also makes the point that in patients with multiple DCS episodes, cutaneous and neurological manifestations were not always concurrent; we agree and do not dispute this. Our data analysis was based on the concurrent presence of neurological and cutaneous symptoms from DCS during one episode, not over a diver's entire DCS history. We agree this would be a valuable analysis, but the nature of the data precluded such analysis.

The Germonpre et al. publication stated, "It has been observed that the occurrence of cutis marmorata is frequently associated with the presence of a patent foramen ovale (PFO) of the heart, and indeed, with a properly executed contrast echocardiographic technique, these patients have an almost 100% prevalence of PFO."[6] This citation was the reference for our statement: "Another study implicated patent foramen ovale (PFO) in nearly 100% of CM/LRC cases."[6]

We stated in our paper that "echocardiography and transcranial Doppler studies used to search for right-to-left shunts carry high false negative results, since high intrathoracic pressures in hyperbaric conditions cannot be replicated with simple induced Valsalva during testing, further underestimating the prevalence."[1] This concept has been cited in literature.[7] Factors influencing diving physiology, such as immersion, water pressure, and vertical position of the diver, are difficult to reproduce and Valsalva maneuver may not result in the same intrathoracic pressure as that underwater.[7] Nevertheless, we appreciate the physiological basis of Dr. Wilmshurst's assertion that "although pressures are increased when underwater, right and left atrial pressures are increased equally." The statement "Valsalva to equalize Eustachian pressures during descent may contribute to transient right-to-left shunt" has been corrected.

We concur with Dr. Wilmshurst's last statement "that if a diver has either cutaneous or neurological DCS, the possibility that there is a right-to-left shunt should be excluded by appropriate testing." We add with emphasis that it is also critical clinically to distinguish true cutaneous DCS (livedo racemosa) from its benign clinical mimickers, livedo reticularis and aquagenic urticaria.

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