Relationship between right-to-left shunts and cutaneous decompression illness

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ABSTRACT

The presence of a large right-to-left shunt is associated with neurological decompression illness after non-provocative dives, as a result of paradoxical gas embolism. A small number of observations suggest that cutaneous decompression illness is also associated with a right-to-left shunt, although an embolic aetiology of a diffuse rash is more difficult to explain. We performed a retrospective case–control comparison of the prevalence and sizes of right-to-left shunts determined by contrast echocardiography performed blind to history in 60 divers and one caisson worker with a history of cutaneous decompression illness, and 123 historical control divers. We found that 47 (77.0%) of the 61 cases with cutaneous decompression illness had a shunt, compared with 34 (27.6%) of 123 control divers ($P < 0.001$). The size of the shunts in the divers with cutaneous decompression illness was significantly greater than in the controls. Thus 30 (49.2%) of the 61 cases with cutaneous decompression illness had a large shunt at rest, compared with six (4.9%) of the 123 controls ($P < 0.001$). During closure procedures in 17 divers who had cutaneous decompression illness, the mean diameter of the foramen ovale was 10.9 mm. Cutaneous decompression illness occurred after dives that were provocative or deep in subjects without shunts, but after shallower and non-provocative dives in those with shunts. The latter individuals are at increased risk of neurological decompression illness. We conclude that cutaneous decompression illness has two pathophysiological mechanisms. It is usually associated with a large right-to-left shunt, when the mechanism is likely to be paradoxical gas embolism with peripheral amplification when bubble emboli invade tissues supersaturated with nitrogen. Cutaneous decompression illness can also occur in individuals without a shunt. In these subjects, the mechanism might be bubble emboli passing through an 'overloaded' lung filter or autochthonous bubble formation.

INTRODUCTION

There is a relationship between neurological decompression illness and right-to-left shunts [1–4]. The possibility that paradoxical gas embolism may be the aetiological mechanism draws comparisons with paradoxical thromboembolism as a cause of stroke [5]. In a study to determine the relationship between right-to-left shunts and neurological decompression illness in divers, we observed that three out of five divers who had cutaneous decompression illness had a shunt [2]. The next nine divers that we saw with a history of cutaneous decompression illness each had a right-to-left shunt [6]. These observations combined suggested an 86% prevalence (12 of 14) of right-to-left shunts in divers with cutaneous decompression illness, compared with a 24%

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prevalence (26 of 109) in control divers who had never had decompression illness ($P < 0.05$) [6].

However, many investigators are sceptical that paradoxical gas embolism could be involved in the aetiology of the widespread skin rash of cutaneous decompression illness. It has been suggested that our earlier observations of an association between cutaneous decompression illness and right-to-left shunts was the result of a small sample size [7]. 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).

METHODS

The cases studied consisted of 60 divers (57 amateur and three professional; 21 female) and one male caisson worker who, on one or more occasion, had a skin rash after decompression from a dive or caisson exposure which was diagnosed by a treating doctor as cutaneous decompression illness. The referral and assessment of each amateur diver was in accordance with the Medical Standards of the United Kingdom Sport Diving Medical Committee. These require enquiry into the circumstances of the incident, including the dive profile, and investigations to exclude the presence of physical predisposition (i.e. to exclude a right-to-left shunt and lung disease). The professional divers and the caisson worker were referred for the same reason, although there is no formal requirement. The referrals came from recompression facility doctors, diving medical referees and general practitioners.

In all cases, transthoracic contrast echocardiography was performed to detect the presence of a right-to-left shunt and for semi-quantitative assessment of the size of any shunt detected, as described previously [4]. We always perform contrast echocardiography before reading the referral letter and blind to history and physical findings, to avoid bias in interpretation. Blinding was achieved by requesting that referring doctors supplied a brief covering letter containing no clinical details, and by booking the referred divers who had had decompression illness on the same echocardiography lists as patients with other clinical conditions, divers who had not had decompression illness and prospective divers who wished to be screened for a shunt.

The heart was imaged (apical four-chamber view) with a Hewlett Packard Sonos 2000 machine. Bubble contrast was produced by pushing approx. 5 ml of sterile saline (0.9% NaCl), 0.5 ml of the subject's blood and 0.5 ml of air back and forth between two syringes connected by a three-way tap until there were no visible bubbles. This mixture was injected through a 21 gauge butterfly needle into a left antecubital vein. The first contrast injection was performed with the subject resting and breathing normally. If no shunt was seen with the first contrast injection, up to five subsequent injections were performed with Valsalva manoeuvres, with the operator causing sudden release of the manoeuvre, as described previously [8]. The size of the shunt was graded according to the maximum number of bubbles seen in the left heart in a single frame on frame-by-frame analysis: small shunts had fewer than six bubbles, medium shunts had between six and 20 bubbles, and large shunts had more than 20 bubbles [5]. On the basis of our previous observations, we believe that small shunts represent a low risk of predisposing to decompression illness, and in the present study they were prospectively considered to be clinically irrelevant [4]. The late appearance of bubbles in the left heart was taken to indicate a pulmonary shunt. Such shunts usually appear as a constant stream of bubbles in the left heart, whereas atrial shunts usually appear as groups of bubbles. The sizes of shunts in the divers with decompression illness were compared with the results obtained by the same method in 123 historical control divers who had not had decompression illness [4,9]. The series of dives or caisson pressure exposures leading up to the episode of decompression illness were analysed for provocative events, as described previously [4].

Statistical comparisons were carried out using the Chi-squared test. Results were considered significant if $P < 0.05$.

All patients were specifically referred to one of us (P. W.) for investigation by doctors outside his hospital, and testing was performed in accordance with national recommendations as part of a clinical service. Contrast echocardiography was performed on cases to enable counselling on the risk of decompression illness in the future, after which some divers went on to closure of their atrial shunts [10]. The chairman of the local ethics committee felt that retrospective review and analysis of the clinical data from patients did not require ethics committee approval. The historical control subjects used for comparison of shunt size were part of a study which had ethics committee approval, and all gave written consent [9].

RESULTS

Table 1 shows the prevalence and sizes of right-to-left shunts at rest and with Valsalva manoeuvres in the individuals who had cutaneous decompression illness and the controls. We found that 47 (77.0%) of the 61 cases with cutaneous decompression illness had a shunt, compared with 34 (27.6%) of the 123 control divers ($P < 0.001$). The sizes of the shunts in the two groups differed

Table 1 Prevalence and sizes of right-to-left shunts in individuals with cutaneous decompression illness and control divers

	Cases with cutaneous decompression illness	Control divers
Large shunts		
At rest	30 (49.2%)	6 (4.9%)
On Valsalva	11 (18%)	3 (2.4%)
All large	41 (67.2%)	9 (7.3%)
Medium shunts		
At rest	4 (6.6%)	2 (1.6%)
On Valsalva	1 (1.6%)	4 (3.3%)
All medium	5 (8.2%)	6 (4.9%)
Small shunts		
At rest	1 (1.6%)	6 (4.9%)
On Valsalva	0	13 (10.6%)
All small	1 (1.6%)	19 (15.4%)
No shunt	14 (23%)	89 (72.4%)
Total	61	123

markedly. Thus 19 of the control divers with shunts had small shunts (which we prospectively considered to be clinically irrelevant), but only one diver with cutaneous decompression illness had a small shunt. However, 30 (49.2%) of the 61 cases with a history of cutaneous decompression illness had a large shunt at rest. Unfortunately, the size grading used concealed the finding that many of the cases had a massive shunt, with hundreds or thousands of bubbles shunting from right to left. This appearance was not seen in control divers. However, even using the predefined grading, the number of cases with large shunts at rest was significantly greater than in the control group [6 (4.9%) of 123] ($P < 0.001$). Whether one compares the prevalence of shunts in the groups using large shunts at rest or at rest and on Valsalva, or whether one compares large and medium shunts at rest or at rest and on Valsalva, the differences between cases and controls were always significant ($P < 0.001$). In five individuals with cutaneous decompression illness, the shunt had the characteristics of a pulmonary shunt (three large, one medium and one small).

Cutaneous decompression illness occurred on a single occasion in 29 divers (16 with large shunts, three with medium shunts, one with a small shunt and nine without a shunt), on two or three occasions in 16 divers (12 with large shunts, two with medium shunts and two without a shunt), and on more than three occasions in the caisson worker and in 15 divers (13 with large shunts and two without a shunt). The caisson worker was affected after standard pressure profiles which did not cause symptoms of decompression illness in other workers on the same shifts, but he had no shunt. When cutaneous decompression illness occurred in a diver with a right-to-left shunt, it was after a dive (depth–time) profile considered to be safe. When it occurred in a diver without a shunt, it usually followed an unsafe profile; usually a long-duration deep dive with a maximum depth greater than 50 m in most cases, and frequently followed a rapid ascent or missed decompression stops.

The rashes were usually confined to parts of the body with significant subcutaneous fat (such as the trunk or thighs). Repeat episodes in an individual usually affected the same region each time.

It was found that 44 of the 60 divers had manifestations of decompression illness on the same or a different occasion other than their cutaneous symptoms. Thus 35 of the divers (29 with a shunt) had neurological decompression illness and three (two with a shunt) had cardiorespiratory decompression illness. In addition, 12 divers (four with a shunt) had limb pain, but in five (three with a shunt) the pain was in a shoulder over which the skin had a marbled/mottled purple cutaneous rash of cutaneous decompression illness. One individual with a shunt had pain and tenderness in a cutaneous lipoma associated with cutaneous and neurological decompression illness after a non-provocative dive.

Of the divers with cutaneous decompression illness and a right-to-left shunt, 17 have had closure of a patent foramen ovale to permit resumption of diving [10]. The mean size of the foramen ovale at the time of the closure procedure was 10.9 mm (median 9 mm; range 7–16 mm). None of these divers have reported the recurrence of cutaneous decompression illness when diving after the closure procedure.

DISCUSSION

These data confirm the finding in a small number of cases that most divers who suffer cutaneous decompression illness also have a right-to-left shunt [6]. The shunt is usually across a foramen ovale, but some have pulmonary shunts. In affected divers, the size of the shunt is considerably greater on contrast echocardiography than that of shunts found in control divers. In the divers who had shunt closure, the large shunts on contrast echocardiography correlated with patent foramen ovale with a mean diameter of 10.9 mm. Foramen ovale of 10 mm diameter or greater were found in only 1.3% of the population in a post-mortem study [11].

Divers who had a right-to-left shunt developed cutaneous decompression illness after dives that would have liberated venous bubbles, but the dives were shallower and more conservative than dives causing cutaneous decompression illness in the divers and the caisson worker who had no shunt. In those without a shunt, the mechanism might be bubble emboli passing through an 'overloaded' lung filter or autochthonous bubble formation. The marbled/mottled appearance of rashes is

consistent with vascular occlusion, but it is not possible to explain the distribution of the shunt by paradoxical gas embolism alone. No individual with a shunt had cutaneous decompression illness after contrast echocardiography. The areas of skin affected were too large to be explained by occlusion of a few adjacent skin vessels. Instead, we believe that the large shunts in affected divers result in masses of microbubble emboli entering the arterial circulation, where their effects are determined largely by the susceptibility of tissues at the time embolism occurs. This susceptibility is determined by the nitrogen contents of different tissues at the time of gas embolism, which in turn determine whether gas emboli dissolve in a tissue with a low dissolved nitrogen content or are amplified as gas passes down the concentration gradient from supersaturated tissues. Nitrogen is highly lipid-soluble, so after a dive the greatest amounts of dissolved nitrogen are in lipid-rich tissues. Some dive profiles cause maximal nitrogen loading of neurological tissues and cause early liberation of venous bubbles, so that paradoxical gas embolism results in bubble amplification in neurological tissues and hence neurological decompression illness [4]. Other dive profiles may cause maximal priming of subcutaneous tissues with nitrogen, to be amplified by later liberation of venous bubbles which pass across the shunt and result in cutaneous decompression illness. Often an episode of cutaneous decompression illness in a diver with a shunt heralded neurological decompression illness after a safe but different dive profile.

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