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Fragmented Health Care Delivery in Ichthyosis[☆]



Dispersión en la atención médica de los pacientes con ictiosis

Congenital ichthyosis is a very rare genetic disease—an estimated 294 patients in Spain are thought to be affected.¹ The disease usually has a large impact on patients' quality of life because, regardless of the type of ichthyosis, patients may experience intractable pruritus, hyperhidrosis, failure to thrive during childhood, cicatricial alopecia, repeated keratitis, conductive hearing loss, and social problems in addition to cutaneous manifestations. As a result, in addition to dermatologists, most patients need to be attended by other specialists such as ophthalmologists, ear-nose-throat specialists, endocrinologists, nutrition experts, and psychologists. If it is rare for many dermatologists to see a patient with ichthyosis, it is easy to imagine the limited experience that many of these other specialists have.

During the first quarter of 2014, we conducted a survey of the members of the Spanish Ichthyosis Association (abbreviated as ASIC in Spanish) to find out the name of the attending physicians. Our aim was to determine whether dermatologists, ophthalmologists, ear-nose-throat specialists, and endocrinologists had experience in ichthyosis. Seventy-two ASIC members responded; in addition, ASIC has data on the dermatologists who attended a further 35 patients. In total, the ASIC members mentioned 70 different dermatologists. Of these, only 3 were mentioned by more than 5 different patients, 5 were the regular

dermatologist for 3 or 4 patients, 9 saw at most 2 patients regularly, and the remaining dermatologists (up to a total of 53) were mentioned by a single patient. A similar picture emerged with the other specialists, who were mentioned in the survey by at least half of the patients: 4 ophthalmologists were mentioned by 2 different patients, only 1 ear-nose-throat specialist was mentioned by more than 1 patient, and no endocrinologist of the 6 mentioned had more than one patient with ichthyosis (Table 1). Interestingly, several members highlighted that although they regularly attended appointments with these specialists, they were not usually seen by the same physician. In summary, our survey shows that very few physicians have experience with a significant number of patients with ichthyosis, and that it is likely that most patients are not attended regularly by other specialists who could help them with some of their possible comorbidities.

It is important to highlight possible biases in our survey. On the one hand, not all patients with ichthyosis belong to the ASIC (in fact, we have the names of dermatologists who attend approximately one third of all Spanish patients), and so the data only partially reflect the true situation. On the other, it is logical to think that dermatologists who recommend joining the ASIC are those who are more strongly represented in the survey. In any case, the proportion of dermatologists, ophthalmologists, and ear-nose-throat specialists who attended a single patient is noteworthy and probably reflects the true situation: most patients with ichthyosis lack a multidisciplinary follow-up, and are also seen by specialists with few other similar patients.

Table 1 Proportion of Patients With Ichthyosis Who Were Attended by the Same Specialist, According to the April 2014 Survey of Members of the Spanish Ichthyosis Association (ASIC).

	Dermatologists n = 70	Ophthalmologists n = 27	Ear-Nose-Throat Specialists n = 19	Endocrinologists n = 6
Attend 7 patients	1	–	–	–
Attend 6 patients	1	–	–	–
Attend 5 patients	1	–	–	–
Attend 4 patients	3	–	–	–
Attend 3 patients	2	–	–	–
Attend 2 patients	9	4	2	–
Attend 1 patient	53	23	18	6
Total Patients	107 (72 + 35 ^a)	31/72	22/72	6/72

^a Members of the ASIC who did not respond to the survey but whose information was available.

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In rare diseases, to become an expert requires experience, but this can only be acquired if professionals attend a significant number of patients. The creation of official reference centers would help referral of patients and group a significant number of them together. This in turn could help to improve the care that they receive.²

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Skin Rash as the Only Manifestation of Mild Decompression Sickness[☆]



Erupción cutánea como única manifestación de enfermedad descompresiva leve

To the Editor:

Decompression sickness is a clinical condition characterized by the formation of bubbles of inert gas in different parts of the body; these bubbles are caused by changes to the solubility of gases triggered by pressure changes during a dive.¹

We report the case of a 55-year-old man with no personal or family history of interest who presented with a slightly pruritic rash that had appeared several hours after a recreational dive. The examination revealed a purpuric-violaceous macular rash on the trunk that was more pronounced in the supine decubitus position (Fig. 1). No rales could be heard in the affected area. Cutaneous ultrasound of the area showed no significant findings in the dermis or hypodermis. With a suspected diagnosis of decompression sickness, the patient underwent an echocardiogram, which revealed a patent foramen ovale measuring 9 mm in diameter; this defect is one of the predisposing factors for decompression sickness. In view of the ultrasound findings and the absence of other symptoms or significant findings in the patient's history, a diagnosis of mild decompression sickness was established. As the patient was otherwise asymptomatic, refused to undergo further tests, and showed progressive clinical improvement, we decided not to perform a skin biopsy and to administer symptomatic treatment



Figure 1 Purpuric-violaceous macular rash with a reticular appearance on the abdomen. Photograph taken 24 hours after the dive.

and take a watch-and-wait approach. At the time of writing, 2 weeks after the diagnosis, the patient is free of symptoms.

Decompression sickness is caused by an increase in ambient pressure during submersion that causes an increase in the partial pressure of inhaled gases (Dalton's Law). This increase, in turn, gives rise to a pressure gradient that results in the accumulation of gases, and nitrogen in particular, that remain dissolved in the body.¹ When the diver returns to the surface, the pressure gradient is reversed, causing oversaturation with gases. Above a certain level, this oversaturation gives rise to the formation of bubbles in different parts of the body. The bubbles trigger a series of responses, such as increased platelet aggregation, capillary permeability and vasoconstriction, that complicate the elimination of gas even further. Predisposing factors

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