RECOGNIZING BASCULE SYNDROME: A REPORT OF FOUR CASES

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Abbreviations **BASCULE** = Bier anemic spots, (acro)cyanosis, and urticaria-like eruption; **POTS** = postural orthostatic tachycardia syndrome.

Case reports. We describe four cases of BASCULE syndrome in adolescent patients aged 13 to 17 years who presented to our department between 2022 and 2025. One additional patient did not provide consent for the publication of his data.

Case 1. A 13-year-old female presented with episodic, pruritic lesions on the lower extremities on a background of transient acrocyanosis. These lesions had occurred episodically for 3 years and were triggered by prolonged standing. The patient reported a burning sensation after a couple of minutes in the shower; the lesions typically resolved within 10 to 15 minutes. Physical examination in the standing position revealed anemic spots on the lower legs, surrounded by orange-red halos against a background of acrocyanosis. The patient reported a mild pruritic and burning sensation, with complete resolution upon sitting. No concomitant systemic symptoms were reported. Her past medical history was notable for two episodes of pre-syncope. Extensive serological testing performed prior to presentation was unremarkable. Cardiological evaluation revealed no autonomic dysfunction or electrocardiographic abnormalities. Based on the clinical findings, a diagnosis of BASCULE syndrome was made. Treatment with high-dose bilastine (20 mg, 2 to 4 times daily) yielded mild improvement at the 3-month follow-up. Given the excellent tolerability of bilastine, continuation of oral antihistamine therapy was recommended.

Case 2. A 16-year-old female presented with pruritic, urticaria-like lesions associated with mottled skin changes (livedo) on her lower extremities. The initial complaint was a marked blue-violet discoloration of the lower legs, feet, and toes. Symptoms began at 10 years of age and were consistently triggered by prolonged standing. No systemic symptoms were reported. During clinical examination in the standing position, mild erythema appeared on the lower limbs along with Bier spots and urticaria-like lesions, thereby confirming the diagnosis of BASCULE syndrome. No other cutaneous signs were observed. The patient showed no signs of autonomic dysfunction. Given the mild nature of the pruritus, initial management focused on trigger avoidance and a watchful waiting approach. Follow-up over 2.5 years demonstrated stable episodic activity.

Case 3. A 14-year-old male presented with a burning rash on his lower legs upon standing. He reported a discoloration of both feet and lower legs that typically lasted a couple of minutes, with complete resolution after sitting down. This rash had been occurring episodically for several years. Clinical examination in the standing position confirmed the characteristic clinical triad of BASCULE syndrome on the lower extremities, with subsequent disappearance of symptoms upon sitting (Figs. 1-4). There were no signs of autonomic dysfunction. Prior to our evaluation, the patient had already been assessed by a cardiologist and a vascular surgeon, both of whose consultations were reassuring. Given

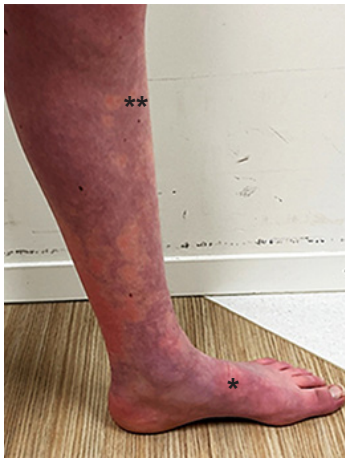

Fig. 1

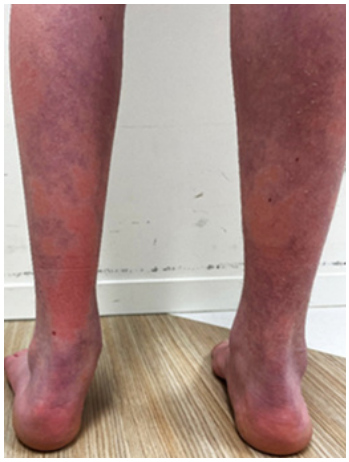

Fig. 2

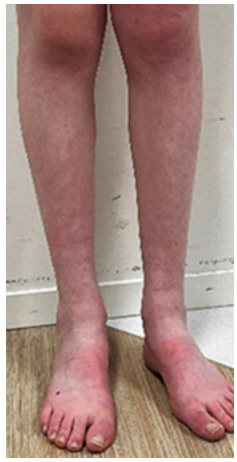

Fig. 3

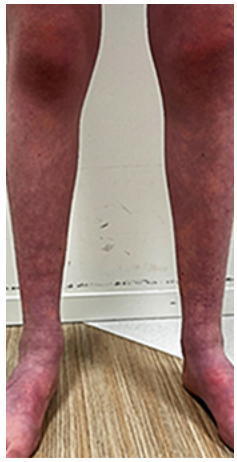

Fig. 4

Fig. 1, 2, 3, 4: BASCULE syndrome in patient 3 with acrocyanosis, Bier's spots*, and urticarial lesions**. In Figs. 3 and 4, patient 3 shortly after standing, showing no abnormalities (Fig. 3). After standing for a couple of minutes (Fig. 4), patient 3 begins to show acrocyanosis and initial Bier's spots on the lower extremities.

the mild, episodic burning sensation, we initiated therapy with desloratadine 5 mg twice daily, with an additional 10 mg as needed. The patient experienced mild improvement at his 3-month follow-up visit; he was subsequently lost to follow-up.

Case 4. A 13-year-old female presented with a 6-month history of episodic, pruritic, and burning lesions on her feet and her upper and lower legs. These lesions appeared exclusively while showering, a few minutes after standing, and resolved spontaneously upon sitting or lying down. No history of recurrent infections, arthritis, or fever was reported. Two months prior to the onset of cutaneous symptoms, the patient had suffered a traumatic fall from a horse resulting in a minor fracture of the T6 vertebra, which was managed with a corset and ibuprofen. On physical examination, the patient was seated and exhibited no abnormal cutaneous findings, with the exception of reversible livedo reticularis on the lower limbs. Given that water was a consistent trigger, we had the patient place her feet in a bucket of warm water, after which cyanosis of the toes and feet rapidly emerged; a few minutes later, both Bier spots and urticaria-like lesions were observed. After standing for 5 minutes, the patient experienced pre-syncope, and a diagnosis of orthostatic hypotension was made. The patient did not undergo the recommended cardiological evaluation and failed to attend her follow-up appointment.

Discussion. In this case series, we describe four adolescents diagnosed with BASCULE syndrome at a single institution within a Belgian tertiary care center, thereby contributing to the limited existing literature and raising awareness of this rare condition. The demographic characteristics of our patients (e.g., age, female predominance) are consistent with previously reported cases.

The underrecognition of BASCULE syndrome has been cited by multiple authors as a primary cause of delayed diagnosis and unnecessary diagnostic investigations. This is highly consistent with our observations. Indeed, all of our patients had been experiencing symptoms for several years and had already consulted multiple physicians for the same complaints. Two of them had undergone extensive diagnostic testing. Baurens et al. reported a case of BASCULE syndrome while also conducting a national French survey to assess awareness of this condition among a group of dermatologists, pe-

diatric rheumatologists, and vascular specialists. Among 95 physicians surveyed, 65% reported having encountered patients with similar clinical findings, yet only 30% were familiar with BASCULE syndrome (8). Consequently, extensive investigations could have been avoided through simple clinical recognition, thereby lowering healthcare costs, reducing invasive procedures (e.g., skin biopsies in a pediatric or adolescent population), and minimizing various specialist referrals. We emphasize that BASCULE syndrome remains a benign vasomotor condition with a chronic yet transient nature, though the exact timeline for resolution remains unknown. Reassurance of patients and parents remains of vital importance.

However, several case series have reported an association between BASCULE syndrome and autonomic dysfunction, particularly postural orthostatic tachycardia syndrome (POTS) and orthostatic hypotension. One of our patients demonstrated clinical orthostatic hypotension; no other signs of autonomic dysfunction were observed. In light of the retrospective studies by Reinhart et al. (2) and Bessis et al. (3), which reported both autonomic abnormalities and electrocardiographic changes in a subset of patients, referral to a cardiologist may be warranted. We recommend remaining vigilant and advise targeted evaluation in patients presenting with symptoms of autonomic dysfunction.

The management of BASCULE syndrome remains empirical and based on case reports and small case series, given that no guidelines currently exist. Oral antihistamines such as bilastine, desloratadine, and fexofenadine have been reported to provide symptomatic relief in a subset of patients. High-dose bilastine and desloratadine improved symptoms in two of our patients. Treatment with propranolol has also been described in the literature. However, evidence regarding long-term outcomes and treatment strategies remains limited. Moreover, several authors, including ourselves, underscore the self-limiting course of this condition along with the often mild symptoms experienced by patients, raising questions regarding the necessity of pharmacological treatment in all patients (2, 8).

Conclusion. The present study describes four cases of BASCULE syndrome and highlights the importance of recognizing this benign, transient vasomotor skin disorder in order to avoid unnecessary investigations and specialist referrals, except in cases where autonomic dysfunction or electrocardiographic abnormalities are suspected.

Conflicts of interest

The authors declare that they have no conflicts of interest.

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