

## Greater Detection of Small Caliber Blood Vessels in Port Wine Stain with X Mode Than With Conventional Doppler: A Pilot Study<sup>☆</sup>



### Superioridad en la detección de vasos de pequeño calibre con modo X flow frente al doppler convencional en la mancha en vino de Oporto. Estudio piloto

Dear Editor:

Port wine stain (PWS) is a type of capillary malformation than can be disfiguring. Although diagnosis is based on clinical findings,<sup>1</sup> the availability of an objective method for evaluating the density and depth of the vessels can help to predict the response to treatment and monitor the stains.<sup>2</sup> Therefore, we designed a pilot study with 2 objectives: (1) to compare the ultrasound characteristics of skin with PWS using high-sensitivity Doppler for the detection of slow-flow vessels (X flow) and contralateral healthy skin; (2) to evaluate whether the blood vessels in PWS can be better visualized with X flow than with conventional Doppler.

We included 20 patients with PWS (10 men and 10 women) who were evaluated in 2015. Sixteen of the 20 patients had previously received treatment with pulsed-dye laser (MultiPlex R, Cynergy) (Table 1). The affected skin and the healthy contralateral skin were evaluated using conventional Doppler ultrasound and X flow. We used an ultrasound device with a 22-MHz transducer (MyLab, Esaote). The images were reviewed by 2 blinded observers, who reported which of the images revealed greater vessel density, with both conventional Doppler and X flow. Clinical and ultrasound images were also recorded (Fig. 1).

The results are summarized in Table 2. Observer 1 reported greater Doppler flow in the PWS than in the healthy skin in 40% of cases and equal Doppler flow in 55%. X flow mode revealed greater flow in the PWS than in healthy skin in 15 patients (75%) and equal flow in 5 (25%), with no cases of greater flow in healthy skin than in affected skin (0%). Observer 2 reported similar findings, namely, greater Doppler flow in the PWS than in healthy skin in 8 cases (40%) and equal flow in 12 cases (60%). X flow mode revealed flow to be greater in the PWS than in healthy skin in 15 patients (75%) and equal in 5 (25%), with no evidence of greater flow in healthy skin than in affected skin (0%).

At present, the treatment of choice for PWS is pulsed-dye laser, which enables the lesion to be treated without damaging the surrounding tissue.<sup>1</sup> However, the results are not satisfactory: complete resolution is achieved in only 10% of cases, and 20%-30% of cases are completely refractory.<sup>2</sup>

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Table 1 Patient Characteristics.

Sex	Age, y	Location	Previous Treatment With PDL
Female	22	Left cheek	5
Female	38	Nasal dorsum	6
Male	55	Upper back	1
Male	43	Left forehead	4
Male	43	Left neck	0
Male	47	Left hip	4
Male	54	Right cheek	0
Male	52	Left V2	0
Female	25	Right leg	3
Male	38	Left V3	0
Male	51	Scalp	0
Male	7	Left leg	0
Female	46	Left V2	3
Female	57	Left V2	3
Female	42	Left V2	3
Female	38	Right V2	1
Female	24	Left V3	1
Female	14	Left hand	1
Female	52	Right V2	1
Male	57	Left chest	1

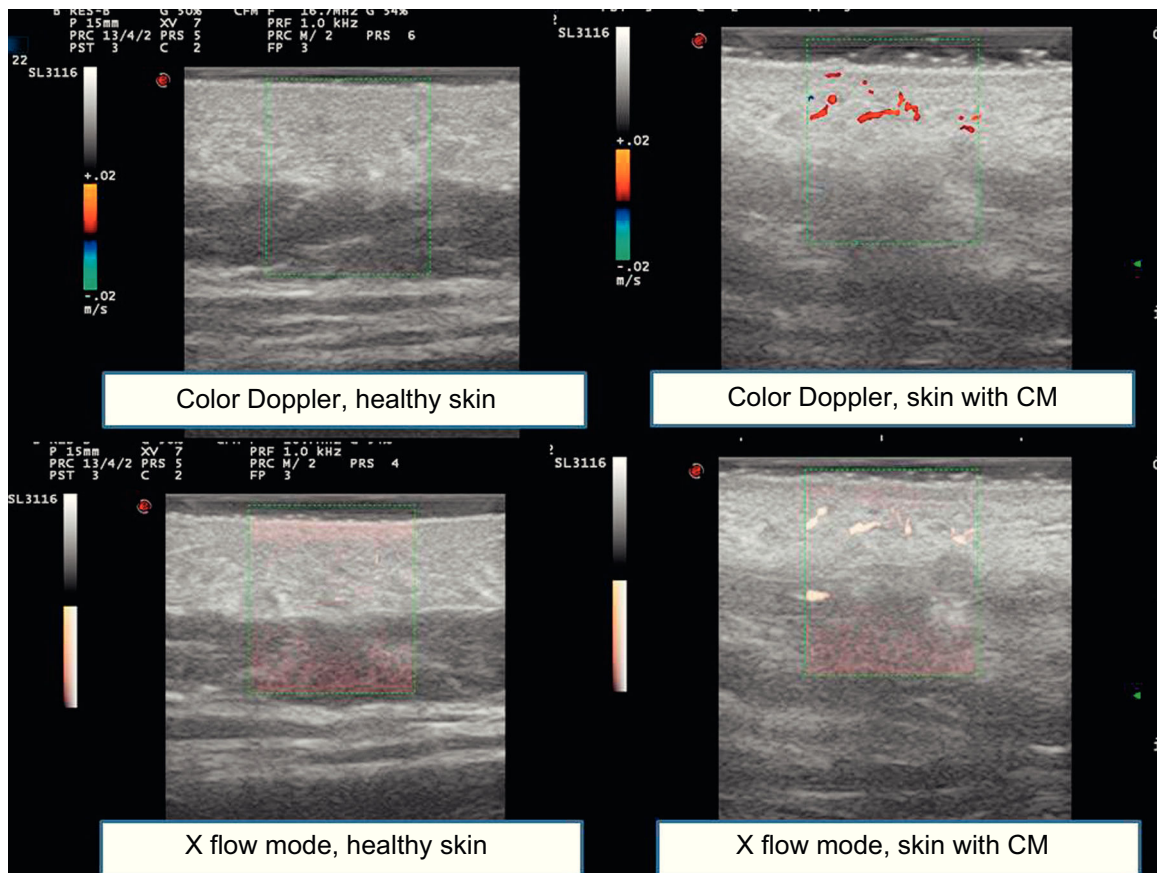
Abbreviation: PDL, pulsed-dye laser.

Very few studies have assessed PSW using ultrasound. Troilus et al.<sup>3</sup> analyzed 55 cases in which ultrasound was used to assess the depth of the PWS and to establish an association with response to pulsed-dye laser treatment. This is of interest, since the penetration of pulsed-dye laser ranges from 0.65 mm according to Hohenleutner et al.<sup>2</sup> to 1.2 mm according to Tan et al.<sup>4</sup> Therefore, the thickness of the PWS and the depth and diameter of the vessels play a key role in the response to treatment.

Ultrasound is a noninvasive method of obtaining objective information on PWS. B mode reveals hypoechogenicity in the dermis, which is more evident in thicker PWS, although this should not be confused with solar elastosis (which requires monitoring of healthy skin). With color Doppler, we find increased vascularization in the dermis; pulsed Doppler reveals neither veins nor arteries.<sup>5</sup> Of note, Troilus et al.<sup>3</sup> reported that a high percentage of PWS (10%) are not visible using ultrasound; in our experience, this percentage could be even higher (20%).

Modes such as X flow (Esaote, MyLab) and B mode (General Electric) enable real-time evaluation of red cells that move within the vascular lumen. The display is obtained by transmitting digitally coded broadband pulses, which are reflected by mobile cells. The echoes that return are decoded and filtered, thus amplifying the weak echoes of the blood cells and eliminating or attenuating echoes in the surrounding tissue. These methods are visual—not quantitative—and make it possible to detect smaller or low-flow vessels. In addition, one of their advantages is the reduced dependency on the angle and the ability to work in real time more dynamically than with conventional Doppler.

A notable limitation is the fact that most patients had previously received laser treatment. It would therefore be interesting to perform a prospective study to compare X



**Figure 1** Ultrasound images comparing color Doppler in healthy skin and skin with capillary malformations and X flow in healthy skin and skin with capillary malformations. CM indicates capillary malformations.

**Table 2** Results.

	Doppler Observer 1	X Flow Observer 1	Doppler Observer 2	X Flow Observer 2
1	A < B	A < B	A < B	A < B
2	A < B	A < B	A < B	A < B
3	A < B	A < B	A < B	A < B
4	A = B	A < B	A = B	A = B
5	A = B	A = B	A = B	A = B
6	A < B	A < B	A < B	A < B
7	A < B	A < B	A < B	A < B
8	A < B	A < B	A < B	A < B
9	A = B	A = B	A = B	A < B
10	A = B	A < B	A = B	A < B
11	A < B	A < B	A < B	A < B
12	A = B	A = B	A = B	A = B
13	A = B	A < B	A = B	A < B
14	A = B	A < B	A = B	A < B
15	A = B	A < B	A = B	A < B
16	A < B	A < B	A < B	A < B
17	A = B	A < B	A = B	A < B
18	A = B	A < B	A = B	A < B
19	A > B	A = B	A = B	A = B
20	A = B	A = B	A = B	A = B

Abbreviations: A, healthy skin; B, port wine stain.

flow images before and after treatment as a complement to clinical observation.

The present study revealed considerable agreement between the 2 observers, which in the case of X flow was 100%. Therefore, we believe that X flow mode provides a better evaluation of PWS, since it is more efficacious for detecting small-caliber, low-flow vessels.

### Conflicts of Interest

The authors declare that they have no conflicts of interest.

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## A Simple and Effective Method for Treating Cyndromas in Brooke-Spiegler Syndrome<sup>☆</sup>



### Método simple y efectivo para el tratamiento de los cilindromas en el síndrome de Brooke-Spiegler

Dear Editor:

The genetic skin disease known as Brooke-Spiegler syndrome (BSS) is a rare autosomal dominant condition with more than 50 cases reported to date.<sup>1</sup> BSS predisposes to benign adnexal tumors that generally first appear in adulthood and involve the folliculo-sebaceous-apocrine unit. It is characterized by multiple cyndromas, trichoepitheliomas, and, occasionally, spiradenomas.<sup>2</sup> The tumors appear simultaneously, as in BSS, or in isolation, as in familial cyndromatosis and multiple familial trichoepithelioma. Consequently, they seem to have a common origin in a pluripotent cell in the folliculo-sebaceous-apocrine unit, with several phenotypic variants.<sup>3,4</sup>

Various mutations have been identified in the tumor suppressor gene *CYLD* on chromosome 16q12-913, thus indicating an association with BSS. *CYLD* inhibits the tumor necrosis factor  $\alpha$  pathway, diminishing expression of nuclear factor  $\kappa$ B (NF- $\kappa$ B), a transcription factor that in turn regulates various antiapoptotic genes involved in the

proliferation of adnexa. A loss of the suppressive function of *CYLD* leads to greater resistance to apoptosis and, consequently, the appearance of these tumors.<sup>3–5</sup>

Clinically, cyndromas present as pink nodules on the scalp and behind the ears (Fig. 1). Trichoepitheliomas appear as papules or small translucent or flesh-colored nodules and are located mainly in the center of the face, especially in the nasal region. Spiradenomas, on the other hand, usually present as solitary bluish or flesh-colored nodules that are often painful.<sup>2</sup>

The emotional impact of recurrence of these lesions on the face and scalp makes treatment essential. Our literature search revealed various options, such as electrocautery, dermabrasion, radiofrequency, cryotherapy, CO<sub>2</sub> laser,<sup>6</sup> radiotherapy (in special cases),<sup>7</sup> and, recently, enucleation.<sup>8</sup> The treatment recommended for solitary cyndromas is surgical removal, whereas partial or total skin grafting has been proposed for multiple lesions.<sup>9</sup> NF- $\kappa$ B



Figure 1 Cyndroma on the scalp.

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