SHORT REPORT

Pulsed dye laser for Sturge–Weber syndrome

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Port wine stain of Sturge–Weber syndrome represents a cosmetic prejudice with social consequences. We have treated eight patients with a 585 nm pulsed dye laser. According to our experience, the treatment is not risky provided that adequate care is taken; the cosmetic result on the V1 port wine stain component is satisfactory.

Sturge–Weber syndrome (SWS) is a neurocutaneous disorder characterised by a facial port wine stain (PWS) of the ophthalmic trigeminal dermatome (V1), associated with leptomeningeal angioma and glaucoma. The aims of the present study were twofold: (1) to assess the safety of the flashlamp pulsed dye laser (PDL) in patients with SWS; and (2) to evaluate its efficacy.

PATIENTS AND METHODS

Eight patients with SWS asked for a laser treatment in our department. The eight patients ranged in age from 6 months to 19 years at onset of treatment (see table 1); all had been diagnosed as having SWS very early and had seizures during their first year of life. Associated leptomeningeal angioma was confirmed by magnetic resonance imaging; seven patients were receiving anticonvulsant therapy during PDL treatment (carbamazepine, stiripentol, and benzodiazepines). Glaucoma was present in five patients; four have been operated on.

The eight patients had complete involvement of the V1 territory including involvement of the forehead and superior eyelid. In five another trigeminal dermatome was involved: V2 in one case and V2+V3 in four cases. We used a PDL (Cynosure, Photogenica V) emitting 450 ms pulses at 585 nm. An average of four complete PDL treatments were performed at 4–6 month intervals (spot size 3–7 mm, energy fluences 5–7 J/cm²). Six patients were treated with EMLA cream for tests or partial treatments; all eight patients were subsequently treated under repeated general anaesthesia with 2–7 sessions.

The cosmetic result was evaluated on the V1 component of PWS with photographs taken before and after each treatment: a good result corresponded to a 50–70% decrease of colour of the PWS; an average result was a 30–50% decrease; and a poor result was less than 30% fading.

RESULTS

The patients tolerated anaesthesia well. During treatment sessions, no seizure was noted under local anaesthesia or during and after general anaesthesia.

On the V1 PWS component, a good cosmetic result was noted for five patients and an average result in three patients (see table 1 and figs 1 and 2). The cosmetic result was always poor on the concomitant V3 involvement, and average on the V2.

DISCUSSION

Children with SWS are at risk of psychological problems, especially in the case of lower levels of intellectual functioning and frequent seizures.¹ Seizures occur in 55–97% of SWS patients,¹ the majority starting during the first year of life. In rare cases, SWS patients may have other vascular abnormalities (Klippel–Trenaunay or Parkes–Weber syndrome) or other neurocutaneous syndromes, especially phakomatosis pigmentovascularis. Psychological problems may be exacerbated by the social consequences of the PWS. It has been shown that large facial PWSs are associated with an increase in mood and social problems in children older than 10 years¹; early treatment of the PWS is therefore desirable.

According to our experience, there is no risk associated with use of PDL in a child with SWS provided that anticonvulsant therapy is maintained and that adequate care is taken. The level of pain associated with the laser treatment is variable. EMLA cream appeared inefficient on large surfaces, and was difficult to apply, especially in the periocular area and eyelids; general anaesthesia has been proposed, with only minimal sequelae²—its benefits outweigh the risks. The anaesthetic management should be carefully planned in such a way as to minimise secondary effects. An increase of both intraocular and intracranial pressures may occur with

Abbreviations: PDL, pulsed dye laser; PWS, port wine stain; SWS, Sturge–Weber syndrome

Table 1 Summary of patients with SWS

Sex, age (y)	Age at 1st treatment (y)	Neurological involvement	Ocular involvement	Anticonvulsant treatment	Number of laser sessions*	Cosmetic result†
M, 1	0.5	Yes	Unknown‡	Carbamazepine	2	Average
F, 6	1	Yes	No	Clonazepam stiripentol, sodium valproate	6	Good
M, 8	7	Yes	No	Carbamazepine	2	Good
M, 11	5	Yes	Yes	Sodium valproate	6	Good
F, 17	16	Yes	Yes	Carbamazepine, epitomax	3	Good
F, 21	13	Yes	Yes	Carbamazepine, clonazepam	2	Average
F, 24	14	Yes	Yes	No	5	Good
M, 29	19	Yes	Yes	Sodium valproate	7	Average

*A session corresponded to a complete PWS treatment.

†Cosmetic result was determined on V1 component of PWS.

‡Ocular involvement could not be excluded because of patient's young age.

Figure 1 Example of a good cosmetic result after six PDL treatments—before.

Figure 2 Example of a good cosmetic result after six PDL treatments—after.

some anaesthetic agents, and in during straining, bucking, and obstructed airways during induction or emergence.³ Difficulties may occur with tracheal intubation because of subsequent hypertrophy of overgrowth angiomatous soft tissues (lips, tongue, etc). It should be performed with soft, non-stylleted, and well lubricated endotracheal tubes to avoid trauma of the vascular lesion.³ The benefit of general anaesthesia is that a large surface area of the PWS can be treated in one session, resulting in less visits and shorter treatment times. In addition, overlapping of pulses is limited, leading to less scarring; also, insertion of stainless steel eye-shields, necessary to protect the eye from the laser beam, is easier.

PDL remains the treatment of choice for the majority of children with PWS; however, it is now widely accepted that the percentage of children achieving complete clearance of their PWSs is considerably less than initially reported.^{4,5} It is essential to inform the patient and parents of a possible failure of treatment, to avoid excessive hope. Indeed, the cosmetic result of PDL on the V1 PWS component in SWS patients is satisfactory, but the result is usually less satisfactory when there is concomitant involvement of V2 and V3 skin territories.⁵ Many factors are involved in the clearance of a PWS by PDL. Both the depth and the diameter of the ectatic blood vessels influence the response to PDL, and as large PWSs consist of varying sized capillaries at differing depths, it would not be surprising if some PWSs failed to respond to the fixed wavelength and pulse duration of the PDL.⁶ A poor response of the PWS is predictable in the case of pink coloration because of small vessel size and deep location, and in the case of a purple PWS formed from

large diameter vessels.⁶ Furthermore, variation of vessels size with age, may explain the better results observed in patients who begin PDL treatment before 1 year of age.

Despite a good result in five cases, none of our SWS patients had a complete clearance of their PWS. The development of new laser technologies is still needed to improve this distressing disorder.

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REFERENCES

- 1 **Chapieski L**, Friedman A, Lachar D. Psychological functioning in children and adolescents with Sturge-Weber syndrome. *J Child Neurol* 2000;**15**:660–5.
- 2 **Grevelink JM**, White VR, Bonoan R, *et al*. Pulsed laser treatment in children and the use of anesthesia. *J Am Acad Dermatol* 1997;**37**:75–81.
- 3 **Batra RK**, Gulaya V, Madan R, *et al*. Anaesthesia and the Sturge-Weber syndrome. *Can J Anaesth* 1994;**41**:133–6.
- 4 **Tan OT**, Sherwood K, Gilchrist BA. Treatment of children with port-wine stains using the flashlamp-pulsed tunable dye laser. *N Engl J Med* 1989;**320**:416–21.
- 5 **Taïeb A**, Touati L, Cony M. Treatment of port-wine stains with the 585-nm flashlamp-pulsed tunable dye laser: a study of 74 patients. *Dermatology* 1994;**188**:276–81.
- 6 **Lanigan SW**. Port-wine stains unresponsive to pulsed dye laser: explanations and solutions. *Br J Dermatol* 1998;**139**:173–7.