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Lasers, Birthmarks, and Sturge-Weber Syndrome: A Pilot Survey

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Background and Objectives: There is limited or no data on the experience of patients with Sturge-Weber Syndrome (SWS) and/or their parents who seek out laser treatments for their port-wine birthmark (PWB). Our study aimed to develop a survey to understand patient perspectives on laser therapy and subsequent behaviors while focusing on three elements of the health belief model: perceived susceptibility, perceived benefits, and perceived barriers.

Study Design/Materials and Methods: A 31 item survey was developed and sent to 650 members of the Sturge-Weber Foundation. The survey included questions regarding four domains: demographics, disease severity, laser experience, and perceptions and behaviors. Logistic regression models and bivariate analyses were conducted to analyze the results.

Results: Among the 123 respondents, earlier initiation of laser treatments was associated with greater birthmark satisfaction. Overall satisfaction increased up to 20 treatments and then leveled off. Color satisfaction increased up to 100 laser treatments; however, the flatness of the PWB was not associated with the number of laser treatments. The perceived benefits of lasers were not associated with the number of lifetime treatments or with spending. However, the perceived susceptibility was associated with greater spending on travel and medical expenses.

Conclusion: The results of our survey provide insight into SWS patient experiences, beliefs, and behaviors regarding laser therapy. Further exploration of these variables may allow for the improvement of the care experience. Lasers Surg. Med. © 2020 Wiley Periodicals LLC

Key words: Sturge-Weber syndrome; port-wine birthmark; laser treatment; health belief model

INTRODUCTION

Sturge-Weber Syndrome (SWS) is a congenital, neurocutaneous disorder classically characterized by two or three of the following: a facial port-wine birthmark (PWB), early-onset glaucoma, and variable degrees of brain involvement [1]. Treatment of the PWB is primarily

with light-based therapies. The rationale for treating the PWB is to improve color, prevent potential future soft tissue hypertrophy, nodularity, bleeding, or facial disfigurement, and reduce the negative psychological impact [2]. Thus, treatment is important for both cosmetic and psychosocial well-being [3,4].

There is a paucity of data regarding perceived beliefs and consequent behaviors of patients with SWS. An investigation of these factors is important to understand patient experiences and determine how this population can be better served. Specifically, measuring patient experience by way of surveys is a means of learning from patients and their families in order to facilitate improvement in their care [5]. Additionally, surveys of patient experience directly evaluate the degree to which care is patient-centered, which is an important dimension of care quality [6].

We performed a pilot survey of the entire membership of the Sturge-Weber Foundation (SWF) to understand how perceptions and behaviors of patients are related to birthmark satisfaction and outcomes. An assessment of these behavioral elements could aid in the development of practical, evidence-based guidelines to approach problems and to provide practical direction for research questions.

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METHODS

The community-based participatory research model was used to develop research questions, create and administer a survey instrument, and interpret the resulting data [7]. Two SWS advocates who are also parents of patients with SWS (JT and KB) collaborated with the investigative team (CB, KT, KK, and SS) as well as the Odum Institute for qualitative research at the University of North Carolina (UNC) to develop research questions, create a survey, ensure readability and understandability, and administer the survey. Two main hypotheses were developed (i) More laser therapy is associated with better outcomes, and (ii) SWF members' behavior is guided by their beliefs about lasers. Based on these hypotheses, survey questions were developed, modified, and narrowed by the entire team over three sessions resulting in thirty-one questions covering four domains (demographics, disease severity, laser experience, and perceptions and behaviors). A group of 5–10 volunteers who had SWS or were caregivers of patients with SWS piloted the survey after each session to ensure reliability. Data were presented to SWF patients and families and discussed to evaluate content validity. We used the health belief model (HBM) to investigate the perceptions and self-reported assessments to investigate outcomes. The HBM has been extensively researched and provides a framework for predicting health behavior [8,9]. The survey was emailed to 650 SWF members and was filled out by the patient and/or parent.

Statistical analysis of data was performed using STATA version 16.4 (StataCorp LLC, College Station, Texas). A predictive logistic regression model was used to investigate the association between birthmark location, type of laser used, initial treatment timing, and the total number of treatments, in relation to the overall satisfaction with birthmark appearance, with the overall satisfaction of 1 being "extremely satisfied" and 7 being "extremely dissatisfied." To assess perceived susceptibility, regarding how stopping laser treatment would affect birthmark appearance, we defined "not concerned" as those who selected "not change," "much less noticeable," and "slightly less noticeable." We defined "concerned" as those who selected "slightly more noticeable" or "much more noticeable." Bivariate analysis was used to investigate relationships between individual variables such as birthmark location and overall satisfaction. A χ^2 test was used to investigate the overall relationships of each independent variable with the dependent variable of birthmark satisfaction.

RESULTS

Demographics

The survey yielded 123 responses and the average age of respondents was 48 years old (range: 8–82 years old). Of the respondents, 80% were the SWS patient and 20% were the caregiver. The majority of respondents were white (83%) and lived in the United States (84%). Most respondents had health insurance, of which 43% were insured by their employer, 17% under Medicare, 12% under

Medicaid, and 7% were uninsured, while the remaining participants did not respond. A little under half of the respondents (48%) were satisfied with their birthmark's appearance. The following PWB locations were reported, and respondents selected as many locations that pertained to their birthmark: 70% eyelid, cheek, and forehead; 60% lip and nose; 40% temple; 25% chin, neck, ear and other; 22% bilateral (Fig. 1).

The following associated diseases were reported and respondents were able to select multiple: 66% medical eye problems and 45% surgical eye problems, 38% migraines, 38% depression, 36% seizures (treated with medications), 35% anxiety, 28% learning disability, 8% attention deficit hyperactivity disorder, 8% limb paralysis, and 7% seizures (treated with surgery). The lifetime number of laser treatments varied, but the majority (63%) reported having 20 or fewer treatments.

Clinical Characteristics and Birthmark Satisfaction

What clinical characteristics determine birthmark satisfaction? Among all respondents, 48% of patients were satisfied with the overall appearance of their PWB. Respondents were more satisfied with the overall appearance of their PWB if the lesion was flat (odds ratio [OR], 4.3 [95% confidence interval [95% CI], 1.50–12.3]) and the same color as the surrounding skin (OR, 5.0 [95% CI, 1.81–13.8]). Individuals with PWB involving the eyelid, forehead, or temple were more likely to describe the birthmark as raised.

Respondents were more satisfied with the overall appearance of their PWB if the lesion was located on the lateral face (cheek, temple, or ear), but less satisfied if the lesion was located on the central face or was bilateral. However, an increased number of sites involved with PWB was not associated with less patient satisfaction with the overall appearance.

Health Behaviors and Birthmark Satisfaction

Are more laser treatments associated with more satisfaction? The majority of respondents (23) had 0–5 treatments, followed by 11–20 treatments as the next popular response (18), and then 21–50 (16), 6–10 (11), 51–100 (11), and >100 treatments (6) (Fig. 2).

The highest percent satisfaction (85%) occurred in those who had >100 treatments; however, only six individuals were in this category. Overall, there was an increase in satisfaction with increased numbers of treatments that leveled off around 20 treatments. However, logistic regression modeling failed to show a statistically significant relationship between the number of treatments and overall satisfaction with the birthmark.

Greater color match with the surrounding skin trended toward more perceived color match with more treatments. Specifically, there was 13% color match with 0–5 treatments; 17% with 6–10; 21% with 11–20; 25% with 21–50; 30% with 51–100; and 17% with >100. However, logistic regression modeling failed to show a statistically significant relationship between the number of treatments

Fig. 1. Most commonly reported port-wine birthmark locations.

and the perceived color match. More treatments were not associated with the greater flatness of the PWB in this sample.

Are earlier laser treatments associated with more satisfaction? Laser treatment initiated between the age of 0 and 1 had the highest percentage satisfaction (65%) compared with satisfaction when treatments were started after the age of 1 (<60%). Additionally, of those who were mildly satisfied or extremely satisfied with the overall appearance of the PWB, 100% of them had ≥ 5 treatments before the age of 2.

Perceived benefits. The results indicated that the majority of people are optimistic about lasers, with 71% believing more lasers will make their PWB less noticeable. We defined optimistic as those who think lasers will make PWB less noticeable and pessimistic as those who think it will not change the PWB. Generally, a greater number of treatments was associated with slightly less optimism, with a drop in optimism after 20 treatments (Fig. 3).

Fig. 2. Number of treatments reported by the respondents.

Fig. 3. Percentage of respondents who believed more laser treatments would make their port-wine birthmark (PWB) less noticeable.

Generally, a greater amount of spending on travel for their last laser treatment was associated with slightly less optimism. Among those who spent \$0–\$20 on their last laser treatment, 70% believed more lasers would make their PWB less noticeable, followed by 78% in those who spent \$21–\$50, 57% in those who spent \$51–\$100, 62% in those who spent \$101–\$500, 67% in those who spent \$501–\$1000, and 67% in those who spent >\$1000. The OR did not show an association with being satisfied with PWB appearance and spending over \$100 on travel (OR, 0.9) or over being satisfied with PWB appearance and spending over \$1000 on out-of-pocket expenses (OR, 1.0).

Overall, the perceived benefits of lasers were not associated with the number of lifetime treatments or with spending.

Perceived susceptibility. Respondents were asked if stopping lasers would make their birthmark less noticeable, slightly less noticeable, unchanged, slightly more noticeable, and much more noticeable. A little over half (56%) of the sample did not think stopping lasers would make their PWB more noticeable. Only 18% believed that stopping lasers will make their PWB much more noticeable.

The number of treatments was not associated with the percentage of those who were worried that stopping lasers will make the PWB more noticeable. The amount of spending on travel for their last treatment was associated with a greater percentage of people who believed that stopping lasers would make the PWB more noticeable. However, the odds overall satisfied if they spent >\$100 on travel costs was 0.9. Total out-of-pocket medical costs in the last year were associated with a greater percentage of people who believed that stopping lasers would make the PWB more noticeable.

Overall, perceived susceptibility was not associated with the number of lifetime treatments. However, perceived susceptibility was associated with spending on travel and medical expenses.

Perceived barriers. The majority of respondents (45%) traveled 11–50 miles to their last laser

appointment, followed by 21% traveling 0–10 miles, 16% traveling 51–100 miles, and 18% traveling >100 miles.

Interestingly, over a third of respondents (37%) are willing to travel >100 miles to laser appointments. Nevertheless, about 20% of respondents have had to skip laser treatment due to travel costs.

A greater number of treatments was associated with more out-of-pocket medical costs. The percentage of those who paid >\$1000 out of pocket increased after 20 treatments (on average, 29% who had less than 20 treatments vs. 50% of those who had over 20 treatments). Therefore, out-of-pocket medical costs may be a significant barrier to receiving more laser treatments.

DISCUSSION

The purpose of this research was to relate perceptions and behaviors with outcomes deemed important to SWS patients and families.

A large percentage of associated diseases reported by the sample included eye problems and seizures, as expected. Interestingly, anxiety and depression were high, but are nevertheless in accordance with the general population who visit SWS-related specialties, including head and neck surgery, dermatology, neurology, and ophthalmology (weighted average = 39%) [10]. The lifetime number of laser treatments varied, but the majority had 20 or fewer treatments. This finding is consistent with what is typically taught to patients. Four to twelve treatments are generally needed, and in about half of the cases, the PWB is partially resistant to treatment depending on various factors [11]. Overall, our data and demographics reflect the general SWF population.

Patients were more likely satisfied if the birthmark was located on the lateral face, was flat and level, unilateral, and was a color match with the surrounding skin. Patients were less likely satisfied if the birthmark was located on the central face, had no color match, was raised, bilateral, and involved classic SWS sites. Interestingly, the number of sites affected by the birthmark was not associated with PWB satisfaction. Overall satisfaction increased up to 20 treatments and then leveled off. Color satisfaction increased up to 100 laser treatments. It is possible that those who have more resistant lesions may undergo more laser treatments, so this subset of patients may have numerous treatments with less or unimproved efficacy, and therefore less overall satisfaction. Other factors besides lesion characteristics, such as laser type or age of the patient, may also influence therapeutic efficacy and overall satisfaction. Surprisingly, the flatness of the PWB was not associated with the number of laser treatments. It is possible that lasers may offer too small of an effect for this benefit to be noticed. Alternatively, lasers may have prevented worse elevation and a “flat” or “raised” descriptor may not capture this kind of dose-response relationship.

Earlier initiation of lasers was associated with higher overall satisfaction. Initiation of lasers before age 1 was associated with color satisfaction and describing the PWB

as flat. Having ≥ 5 treatments before age two was associated with describing the PWB as flat, but not with color satisfaction. Generally, it is known that PWBs can become darker and thicken with time if left untreated [12]. Consequently, the earlier treatment produces more optimal results [13]. Early treatment also decreases the psychosocial effects associated with facial lesions [14]. Therefore, this finding is consistent with the advantages of initiating early PWB treatment.

Perceived susceptibility was not associated with the number of lifetime treatments but was associated with spending on travel and medical expenses. A little over half of our sample did not think stopping lasers would make their PWB more noticeable. Appropriately, patients should be provided more information about the progression of the PWB and the potential to result in hypertrophy and nodularity if patients fail to follow through with laser treatments [15]. However, studies have shown that many PWB will demonstrate some recurrence or re-darkening [16,17], so we should also instruct patients about this recurrence. Unsurprisingly, individuals who paid higher total out-of-pocket costs were more likely to think that stopping lasers will make their PWB more noticeable. However, their lesions were no less flat than people who paid less medical expenses. It is possible that people may be seeking experts (with increased cost) mostly out of fear and not hope.

Regarding perceived barriers, individuals are very willing to travel long distances for laser therapy. This is in accordance with another study, which found that the majority of their respondents were willing to travel 3–6 hours for a treatment that would remove their PWB within five treatment sessions [18]. Although our findings demonstrated that travel costs are a significant barrier for a large portion of the PWB population, especially those who receive more treatments. Overall, people who have more treatments, pay more for travel, and hence end up paying more for their treatment. Since current treatment modalities are limited in their ability to clear all PWB, new and improved therapies with enhanced efficacy are needed [18]. This could decrease the number and costs of treatments in a considerable proportion of patients.

These findings demonstrate that our study population is well-informed on the benefits of lasers; however, our sample would appreciate a better understanding of their perceived susceptibility. More effort through research and communication is needed to define who needs more lasers and who needs to seek more specialized care. In particular, it is important to better define levels of risk and tailor this information to each patient.

The current literature provides insight on the risk stratification of patients with PWB and has discussed various treatment regimens depending on the characteristics of each patient's PWB. PDL treatments are generally effective and have demonstrated a 70% improvement in 50% of patients. Superficial PWB located in V1 and V3 dermatomes or truncal distribution tend to be more responsive to PDL, whereas large capillary malformations involving the V2 dermatome, distal extremities,

deep dark, and hypertrophic PWB respond less to PDL [15,19]. Another “risk” factor is related to the age of the patient at the time of the treatment. PWB in adults tends to be larger compared with infants with possible development of nodularity and hypertrophy [14]. Other light-based therapies with longer wavelengths and deeper penetration can be employed to treat resistant and hypertrophic PWB [15]. The above information should be communicated with each patient while keeping in mind the goals and desires of the patients and families as part of a shared decision-making process. Continued support to alleviate potential barriers to care, primarily cost, should be provided by the SWF, and providers should also consider financial aspects when determining the optimal treatment strategy for each patient.

Several limitations exist in our pilot study. The response rate was low, which may be due to the email method of survey delivery or length of the survey. Due to our low response rate, definitive conclusions based on our data cannot be made, and this pilot survey can only provide preliminary insight. In the future, strategies to increase response rates are necessary. Furthermore, parents were allowed to respond for their children with SWS, which may not provide an accurate representation of the patient experience. Lastly, those involved with the SWF may be more likely to seek treatment; therefore, our survey is not necessarily representative of all patients with PWB. Despite these limitations, we provide a description of the characteristics, perceptions, and behaviors of a representative cohort of patients with SWS.

CONCLUSION

The present research expands our understanding of the SWF member experience in a sample of patients with SWS using the HBM as a framework. Comprehensive understanding of variables related to patient experience, beliefs, and behaviors will assist families and medical providers to deliver best care practices and improve patient outcomes. Our findings also support the need for further research on risk stratification and improved communication regarding the perceived susceptibility of these patients. Such information could help SWS patients, advocates, and providers develop strategies to improve the care experience.

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